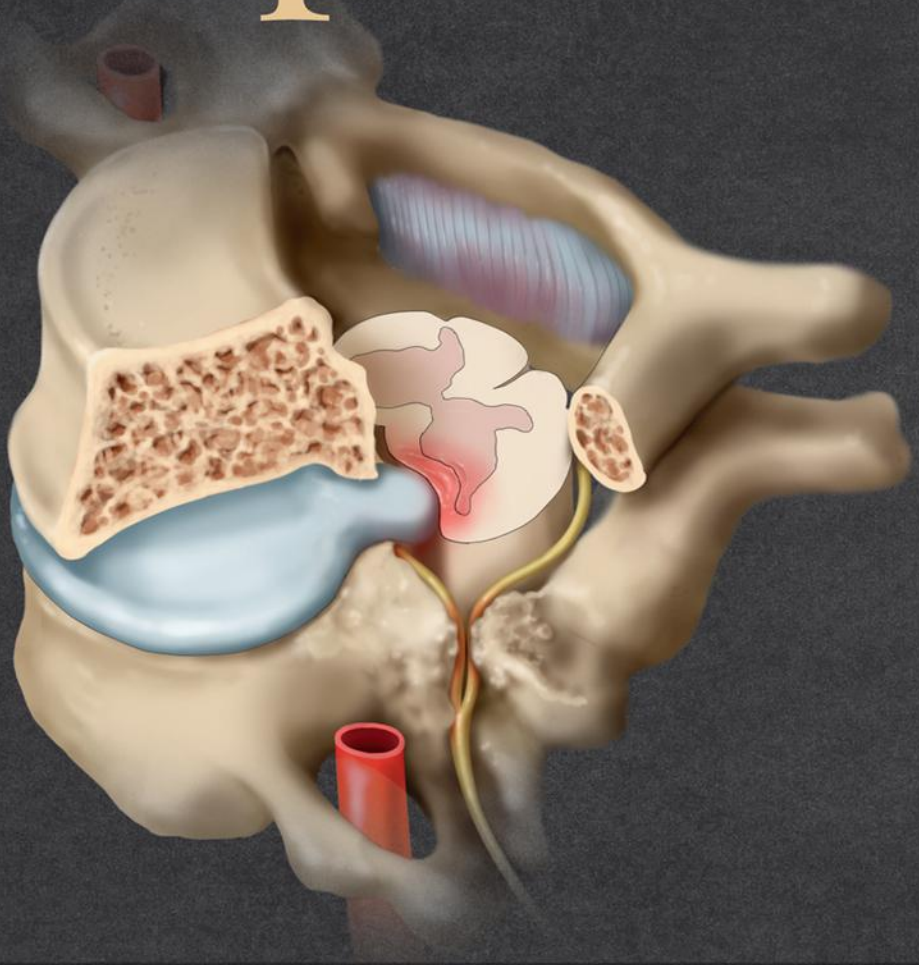
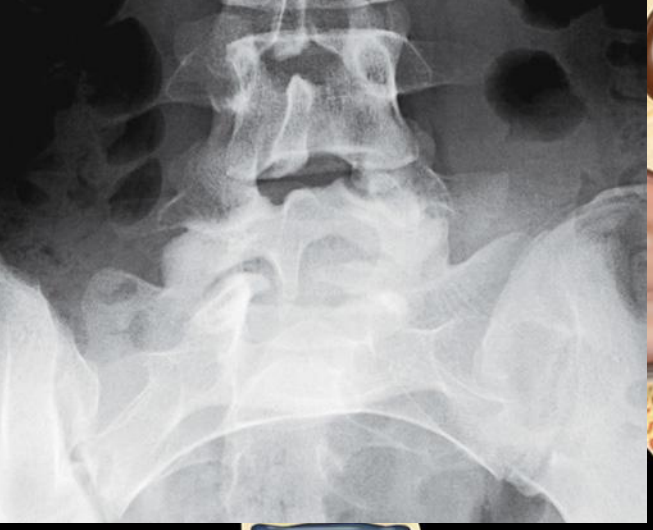
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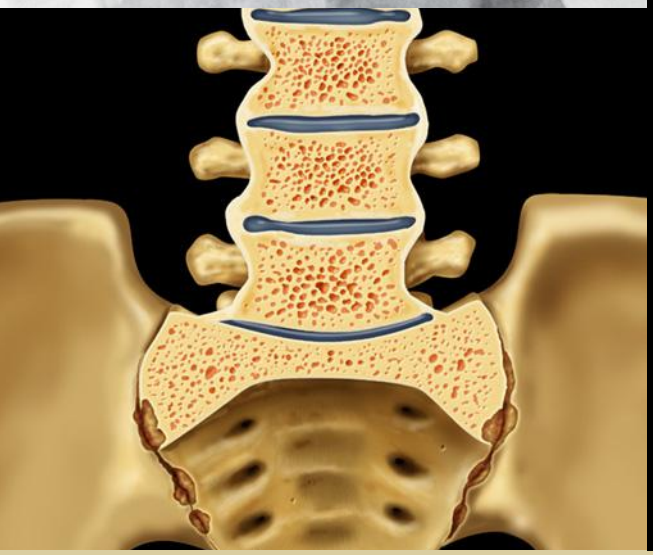
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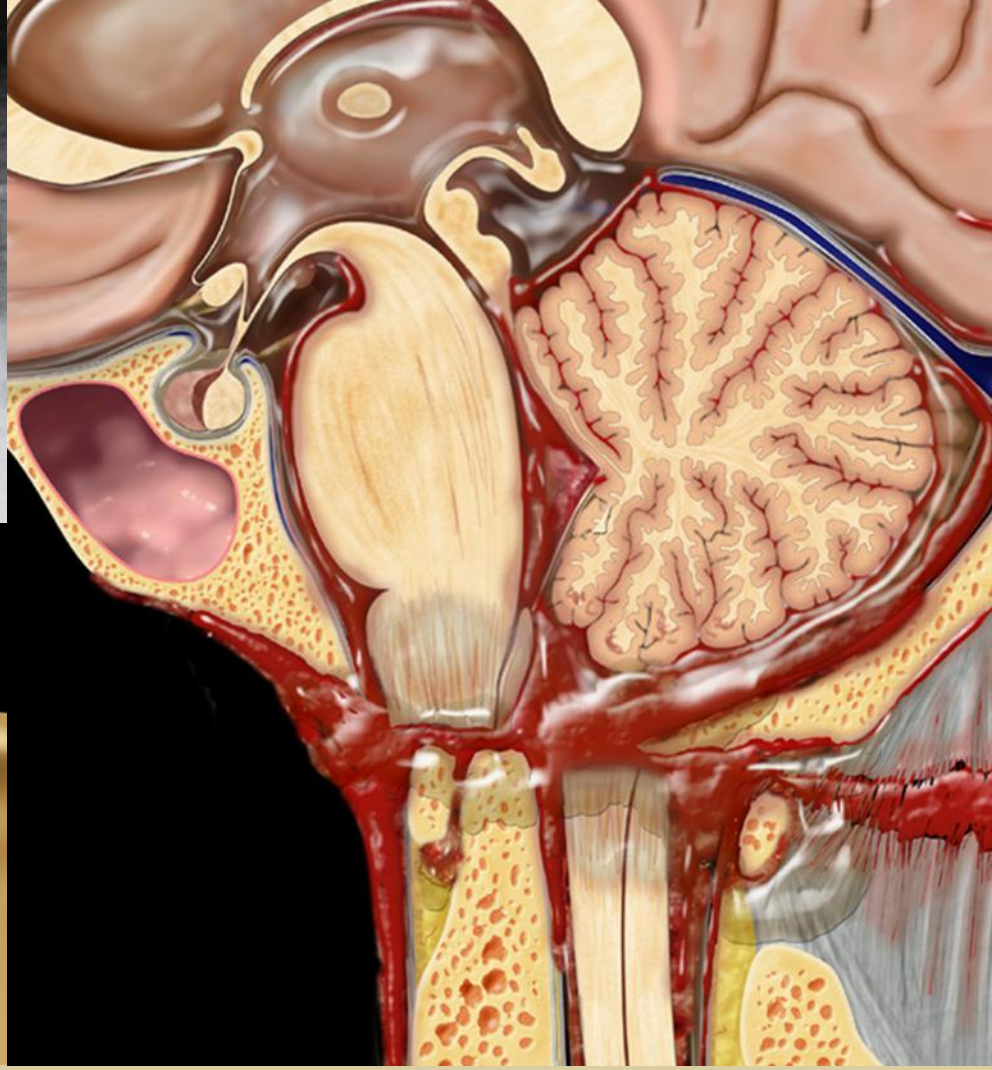


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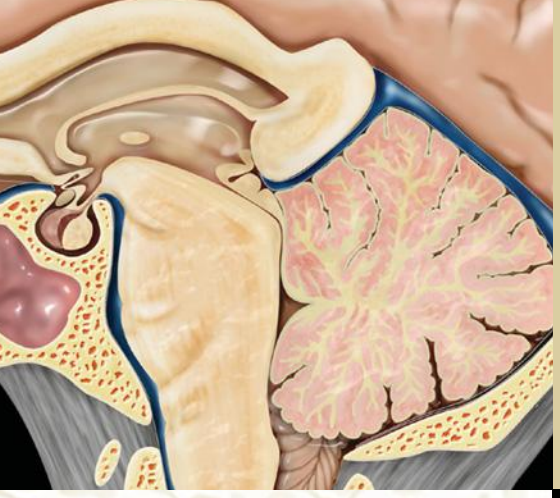


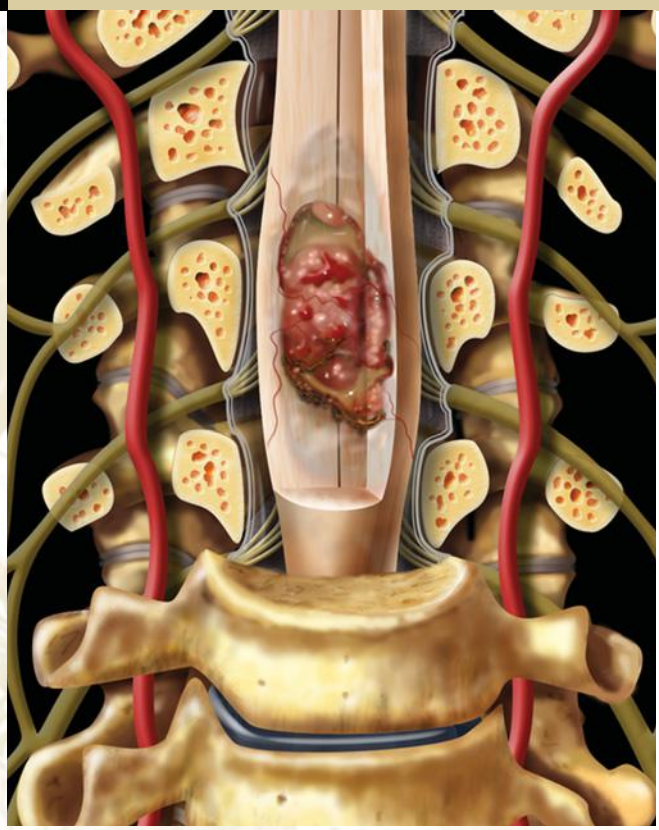




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Jeffrey S. Ross, MD
Neuroradiology
Barrow Neurological Institute
St. Joseph's Hospital
Phoenix, Arizona

Kevin R. Moore, MD
Pediatric Neuroradiology
Intermountain Pediatric Imaging
Primary Children's Hospital
Salt Lake City, Utah

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1600 John F. Kennedy Blvd.
Ste 1800
Philadelphia, PA 19103-2899

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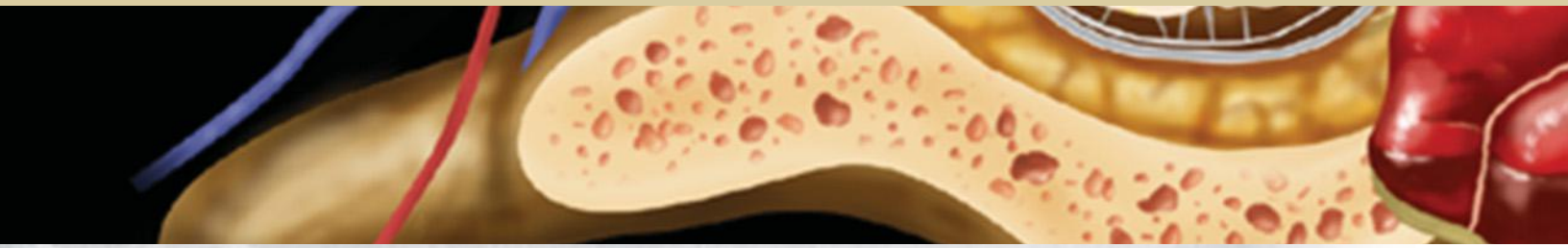
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"Let the wise hear and increase in learning, and the one who understands obtain guidance."
Proverbs 1:5

JSR

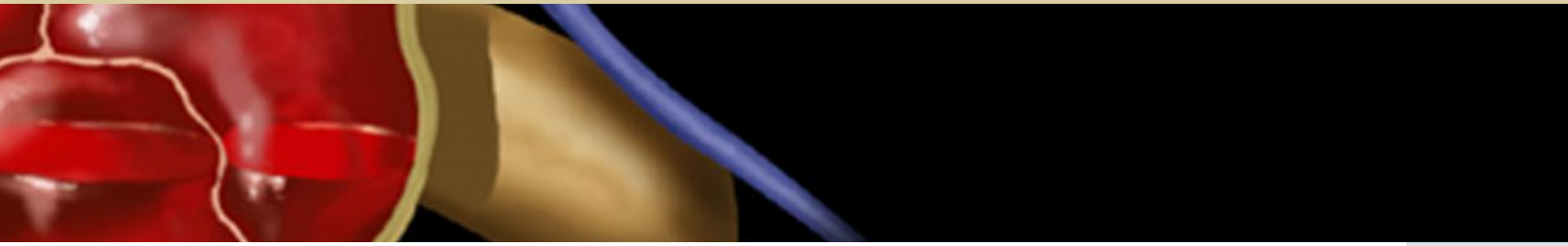
A project of this magnitude reflects the hard efforts of many underrecognized people. I extend my deepest gratitude to all of you – thank you!

KRM





Contributing Authors



Lubdha M. Shah, MD
Associate Professor of Radiology
Division of Neuroradiology
University of Utah School of Medicine
Salt Lake City, Utah

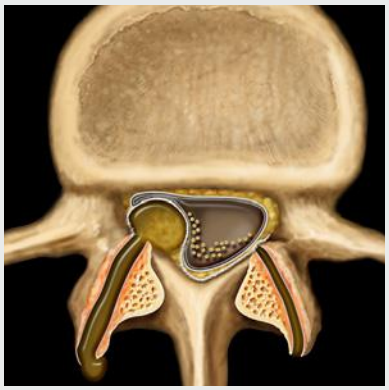
Bryson Borg, MD
Fairfield, California

Julia Crim, MD
Chief of Musculoskeletal Radiology
Professor of Radiology
University of Missouri at Columbia
Columbia, Missouri

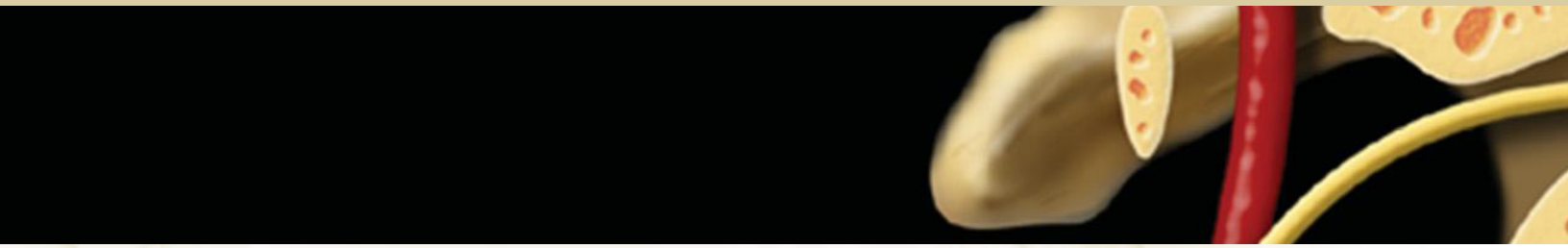
Cheryl Petersilge, MD, MBA
Clinical Professor of Radiology
Cleveland Clinic Lerner College of Medicine
Case Western Reserve University
Cleveland, Ohio

A. Carl Merrow, MD
Corning Benton Chair for Radiology Education
Cincinnati Children's Hospital Medical Center
Associate Professor of Clinical Radiology
University of Cincinnati College of Medicine
Cincinnati, Ohio









Preface



Welcome to the third edition of *Diagnostic Imaging: Spine*. Five years have passed since the second edition, and eleven years have flown by since the first edition was published. This edition is a refresh with new images, new diagnoses, and updated text and references. The same excellent Amirsys formatting is present, with individual diagnoses capable of standing alone but with a logical integration within the larger sections. The Key Facts box retains its visual prominence at the beginning of each diagnosis, allowing for a quick scan of the most important bullet points when time is short (and when is it not?). The text format remains in the hallmark Amirsys bulleted form that allows a large amount of important information to be displayed in an easy-to-use and inviting layout. Prose text chapters are included for the introduction to major sections, which are color coded, and the use of tables allows quick scanning for important data and measurements.

Our coauthors and the staff at Amirsys/Elsevier have been amazing to work with, and we have been very fortunate to be able to interact with and learn from such a fantastic team. We hope you find this edition useful not only as a reference, but also as an integral assistant in your daily practice.

Jeffrey S. Ross, MD

Neuroradiology
Barrow Neurological Institute
St. Joseph's Hospital
Phoenix, Arizona

Kevin R. Moore, MD

Pediatric Neuroradiology
Intermountain Pediatric Imaging
Primary Children's Hospital
Salt Lake City, Utah



Acknowledgements

Text Editing

Dave L. Chance, MA, ELS
Arthur G. Gelsinger, MA
Nina I. Bennett, BA
Sarah J. Connor, BA
Tricia L. Cannon, BA
Terry W. Ferrell, MS
Lisa A. Gervais, BS

Image Editing

Jeffrey J. Marmorstone, BS
Lisa A. M. Steadman, BS

Medical Editors

Michal Klysik, MD
Karthik C. Kumar, MD

Illustrations

Richard Coombs, MS
Lane R. Bennion, MS
Laura C. Sesto, MA

Art Direction and Design

Tom M. Olson, BA
Laura C. Sesto, MA

Lead Editor

Nina I. Bennett, BA

Production Coordinators

Angela G. Terry, BA
Rebecca L. Hutchinson, BA

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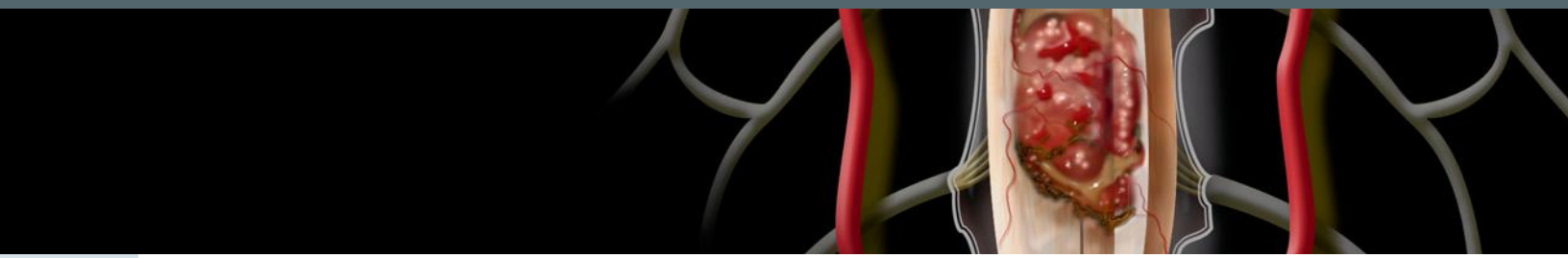
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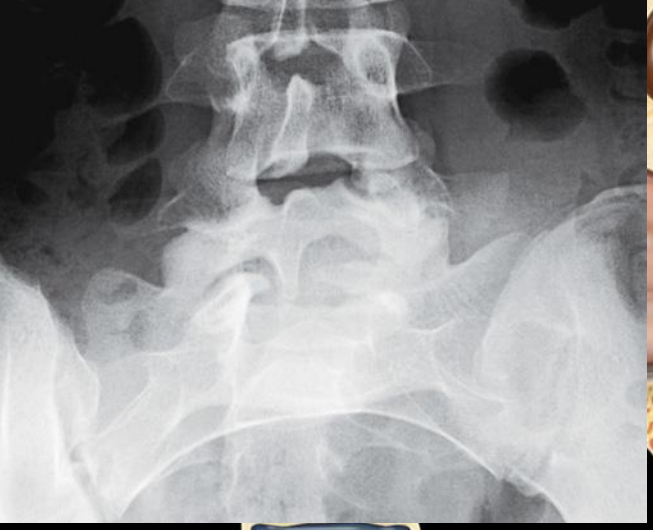
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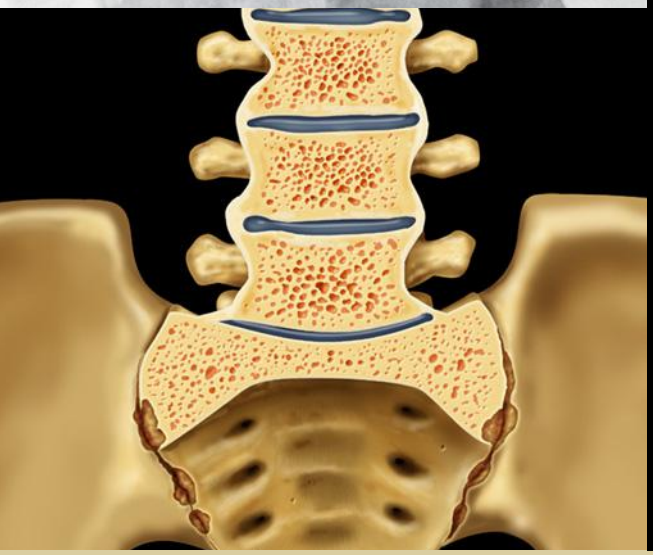
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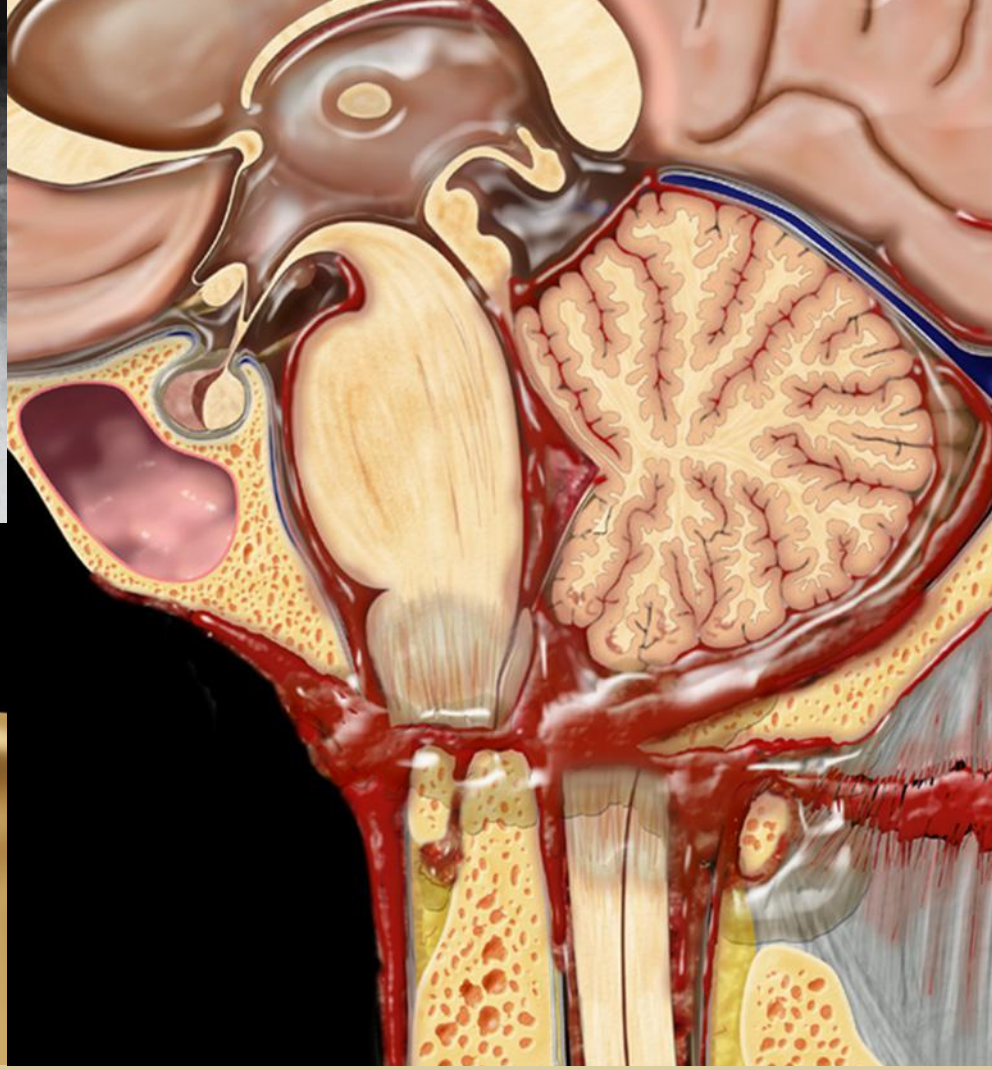
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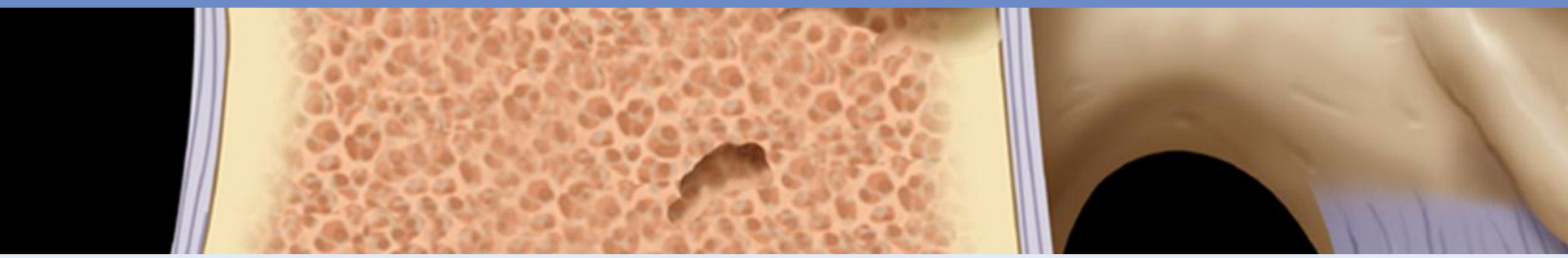


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Spine

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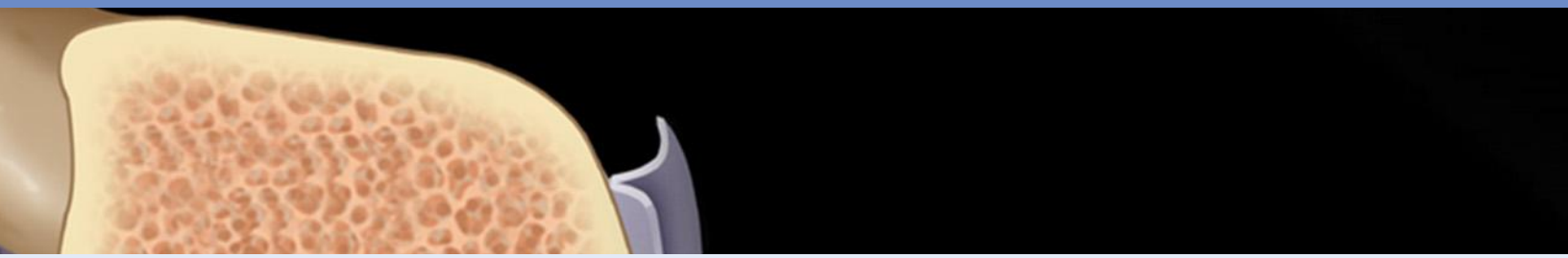
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Imaging Anatomy

There are **33** spinal vertebrae, which comprise two components: A cylindrical ventral bone mass, which is the vertebral **body**, and the dorsal **arch**.

7 cervical, 12 thoracic, 5 lumbar bodies

- 5 fused elements form the sacrum
- 4-5 irregular ossicles form the coccyx

Arch

- 2 pedicles, 2 laminae, 7 processes (1 spinous, 4 articular, 2 transverse)
- Pedicles attach to the dorsolateral aspect of the body
- Pedicles unite with a pair of arched flat laminae
- Lamina capped by dorsal projection called the spinous process
- Transverse processes arise from the sides of the arches

The two articular processes (zygapophyses) are diarthrodial joints.

- (1) Superior process bearing a facet with the surface directed dorsally
- (2) Inferior process bearing a facet with the surface directed ventrally

Pars interarticularis is the part of the arch that lies between the superior and inferior articular facets of all subatlantal movable elements. The pars are positioned to receive biomechanical stresses of translational forces displacing superior facets ventrally, whereas inferior facets remain attached to dorsal arch (spondylolysis). C2 exhibits a unique anterior relation between the superior facet and the posteriorly placed inferior facet. This relationship leads to an elongated C2 pars interarticularis, which is the site of the hangman's fracture.

Cervical

The cervical bodies are small and thin relative to the size of the arch and foramen, with the transverse diameter greater than the AP diameter. The lateral edges of the superior surface of the body are turned upward into the uncinate processes. The transverse foramen perforates the transverse processes. The vertebra artery resides within the transverse foramen, most commonly starting at the C6 level.

C1 has no body and forms a circular bony mass. The superior facets of C1 are large ovals that face upwards and the inferior facets are circular in shape. Large transverse processes are present on C1 with fused anterior and posterior tubercles.

The C2 complex consists of the axis body with dens/odontoid process. The odontoid embryologically arises from the centrum of the first cervical vertebrae.

The C7 vertebral body shows a transitional morphology with a prominent spinous process.

Thoracic

- Thoracic bodies are heart-shaped and increase in size from superior to inferior
- Facets are present for rib articulation and the laminae are broad and thick
- Spinous processes are long, directed obliquely caudally
- Superior facets are thin and directed posteriorly
- The T1 vertebral body shows a complete facet for the capitulum of the first rib, and an inferior demifacet for capitulum of second rib
- The T12 body has transitional anatomy, and resembles the upper lumbar bodies with the inferior facet directed more laterally

Lumbar

The lumbar vertebral bodies are large, wide and thick, and lack a transverse foramen or costal articular facets. The pedicles are strong and directed posteriorly. The superior articular processes are directed dorsomedially and almost face each other. The inferior articular processes are directed anteriorly and laterally.

Joints

Synarthrosis is an immovable joint of cartilage, and occurs during development and in the first decade of life. The neurocentral joint occurs at the union point of two centers of ossification for two halves of the vertebral arch and centrum.

Diarthrosis is a true synovial joint that occurs in the articular processes, costovertebral joints, and atlantoaxial and sacroiliac articulations. The pivot-type joint occurs at the median atlantoaxial articulation. All others are gliding joints.

Amphiarthroses are nonsynovial, movable connective tissue joints. **Symphysis** is a fibrocartilage fusion between two bones, as in the intervertebral disc. **Syndesmosis** is a ligamentous connection common in the spine, such as the paired ligamenta flava, intertransverse ligaments, and interspinous ligaments. An unpaired syndesmosis is present in the supraspinous ligament.

Atlantooccipital articulation is composed of a diarthrosis between the lateral mass of atlas and occipital condyles and the syndesmoses of the atlantooccipital membranes. Anterior AO membrane is the extension of the ALL. The posterior AO membrane is homologous to the ligamenta flava.

Atlantoaxial articulation is a pivot joint. The transverse ligament maintains the relationship of the odontoid to the anterior arch of atlas. Synovial cavities are present between the transverse ligament/odontoid and the atlas/odontoid junctions.

Disc

The intervertebral disc is composed of three parts: The **cartilaginous endplate**, the **anulus fibrosis**, and the **nucleus pulposus**. The height of the lumbar disc space generally increases as one progresses caudally. The anulus consists of concentrically oriented collagenous fibers which serve to contain the central nucleus pulposus. These fibers insert into the vertebral cortex via Sharpey fibers and also attach to the anterior and posterior longitudinal ligaments. Type I collagen predominates at periphery of anulus, while type II collagen predominates in the inner anulus. The normal contour of the posterior aspect of the anulus is dependent upon the contour of its adjacent endplate. Typically, this is slightly concave in the axial plane; although, commonly at L4-L5 and L5-S1 these posterior margins will be flat or even convex. A convex shape on the axial images alone should not be interpreted as degenerative bulging.

The nucleus pulposus is a remnant of the embryonal notochord and consists of a well-hydrated, noncompressible proteoglycan matrix with scattered chondrocytes. Proteoglycans form a major macromolecular component, including chondroitin 6-sulfate, keratan sulfate, and hyaluronic acid. Proteoglycans consist of protein core with multiple attached glycosaminoglycan chains. The nucleus occupies an

eccentric position within the confines of anulus and is more dorsal with respect to the center of the vertebral body. At birth, approximately 85-90% of the nucleus is water. This water content gradually decreases with advancing age. Within the nucleus pulposus on T2-weighted sagittal images, there is often a linear hypointensity coursing in an anteroposterior direction, the intranuclear cleft. This region of more prominent fibrous tissue should not be interpreted as intradiscal air or calcification.

Anterior Longitudinal Ligament (ALL)

The ALL runs along the ventral surface of the spine from the skull to the sacrum. The ALL is narrowest in the cervical spine and is firmly attached at the ends of each vertebral body. It is loosely attached at the midsection of the disc.

Posterior Longitudinal Ligament (PLL)

The PLL runs on the dorsal surface of bodies from the skull to the sacrum. The PLL has a segmental denticulate configuration and is wider at the disc space, but narrows and becomes thicker at the vertebral body level.

Craniocervical Ligaments

The craniocervical ligaments are located anteriorly to spinal cord and occur in three layers: Anterior, middle, and posterior. Anterior ligaments consist of the odontoid ligaments (apical and alar). The apical ligament is a small fibrous band extending from dens tip to basion. Alar ligaments are thick, horizontally directed ligaments extending from the lateral surface of dens tip to anteromedial occipital condyles. The middle layer consists of the cruciate ligament. The **transverse ligament** is a strong horizontal component of the cruciate ligament extending from behind the dens to the medial aspect of C1 lateral masses. The craniocaudal component consists of a fibrous band running from the transverse ligament superiorly to the foramen magnum and inferiorly to C2. Posteriorly, the tectorial membrane is the continuation of PLL and attaches to anterior rim of the foramen magnum.

Vertebral Artery

The vertebral artery arises as the first branch of the subclavian artery on both sides. The vertebral artery travels cephalad within the foramen transversarium (transverse foramen) within the transverse processes. The first segment of the vertebral artery extends from its origin to the entrance into the foramen of the transverse process of the cervical vertebrae, usually the 6th. The most common variation is the origin of the left vertebral from the arch, between the left common carotid and the left subclavian arteries (2-6%). The vertebral artery in these variant cases almost always enters the foramen of the transverse process of C5. The second segment runs within the transverse foramen to the C2 level. Nerve roots pass posterior to the vertebral artery. The third segment starts at the C2 level where the artery loops and turns lateral to ascend in the C1 transverse foramen. It then turns medial crossing on top of C1 in a groove. The fourth segment starts where the artery perforates the dura and arachnoid at lateral edge of posterior occipitoatlantal membrane, coursing ventrally on the medulla to join with the other vertebral to make the basilar artery.

Vertebral Column Blood Supply

Paired segmental arteries (intercostals, lumbar arteries) arise from the aorta and extend dorsolaterally around the middle of the vertebral body. Near the transverse process the segmental artery divides into lateral and dorsal branches. The lateral branch supplies dorsal musculature and the dorsal branch passes lateral to the foramen, giving off branch(es) and providing major vascular supply to bone and vertebral canal contents. The posterior central branch supplies disc and vertebral body, while the prelaminal branch supplies the inner surface of the arch, ligamenta flava, and regional epidural tissue. The neural branch entering the neural foramen supplies pia, arachnoid, and cord. The postlaminar branch supplies musculature overlying lamina and branches to bone.

Nerves

- Spinal nerves are arranged in 31 pairs and grouped regionally: 8 cervical, 12 thoracic, 5 lumbar, 5 sacral, 1 coccygeal
- Ascensus spinalis is the apparent developmental rising of the cord related to differential spinal growth
- Course of nerve roots becomes longer and more oblique at lower segments
- C1 nerve from C1 segment and exits above C1
- C8 nerve from C7 segment and exits at C7-T1
- T6 nerve from T5 segment and exits at T6-T7
- T12 nerve from T8 segment and exits at T12-L1
- L2 nerve from T10 segment and exits at L2-L3
- S3 nerve from T12 segment and exits at the S3 foramen

Meninges are divided into dura, arachnoid, and pia

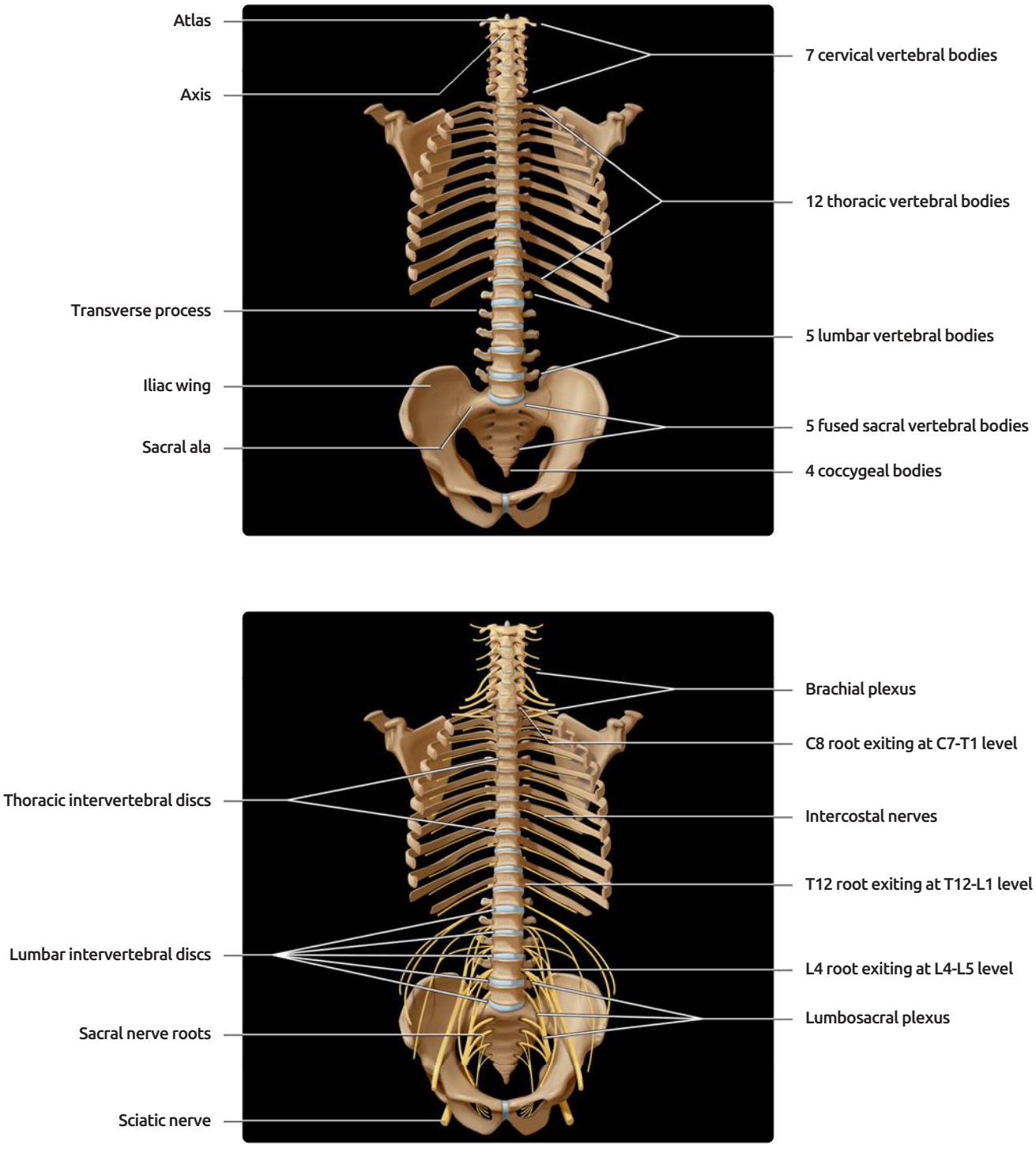
Dura is a dense, tough covering corresponding to the meningeal layer of the cranial dura. The epidural space is filled with fat, loose connective tissue, and veins. The dura continues with spinal nerves through the foramen to fuse with the epineurium. Cephalic attachment of the dura is at the foramen magnum and the caudal attachment at the back of the coccyx.

Arachnoid is the middle covering, which is thin, delicate, and continuous with cranial arachnoid. The arachnoid is separated from the dura by the potential subdural space.

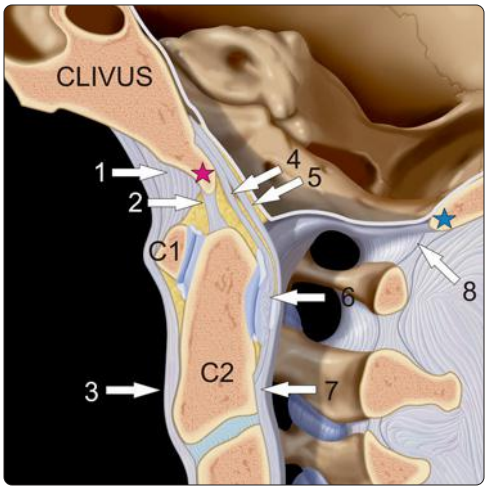
Pia is the inner covering of delicate connective tissue closely applied to the cord. Longitudinal fibers are laterally concentrated as denticulate ligaments lying between posterior and anterior roots, and attach at 21 points to dura. Longitudinal fibers are concentrated dorsally as the septum posticum attaching the dorsal cord to the dorsal midline dura.

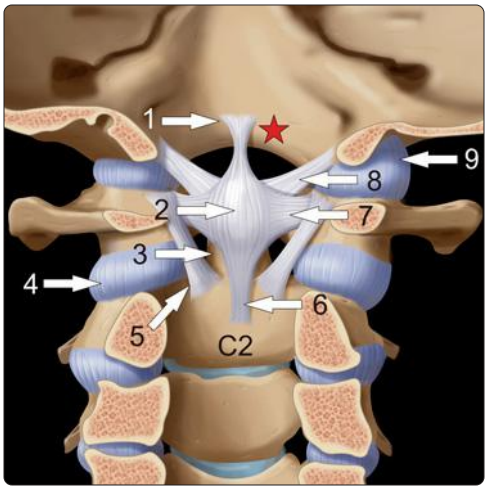
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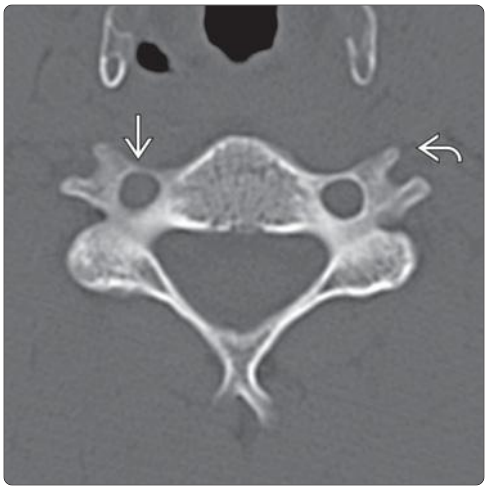
(Top) *Coronal graphic of the spinal column shows relationship of 7 cervical, 12 thoracic, 5 lumbar, 5 fused sacral, and 4 coccygeal bodies. Note cervical bodies are smaller with neural foramina oriented at 45° and capped by the unique C1 and C2 morphology. Thoracic bodies are heart-shaped, with thinner intervertebral discs, and are stabilized by the rib cage. Lumbar bodies are more massive, with prominent transverse processes and thick intervertebral discs.* **(Bottom)** *Coronal graphic demonstrates exiting spinal nerve roots. C1 exits between the occiput and C1, while the C8 root exits at the C7-T1 level. Thoracic and lumbar roots exit below their respective pedicles.*



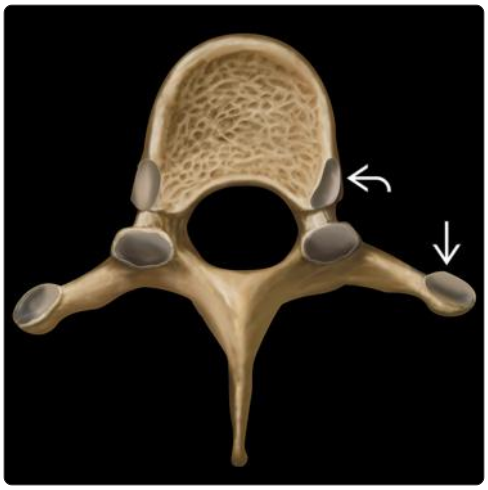


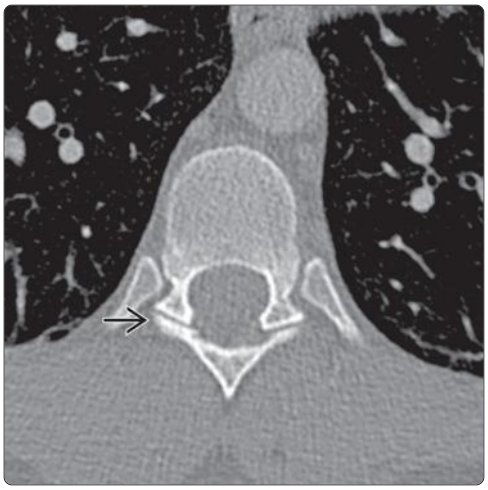
(Left) *Sagittal graphic of CVJ shows: 1) Ant. atlantooccipital membrane, 2) apical lig., 3) ALL, 4) cruciate lig., 5) tectorial membrane, 6) transverse lig., 7) post. longitudinal lig., and 8) post. atlantooccipital membrane. Red star is the basion; blue star is the opisthion.* **(Right)** *Posterior view of CVJ shows: 1) Sup. cruciate ligament, 2) cruciate lig., 3) odontoid ant. to cruciate lig., 4) atlantoaxial jt., 5) accessory atlantoaxial lig., 6) inf. cruciate lig., 7) transverse lig., 8) alar lig., and 9) atlantooccipital jt. Red star = basion.*





(Left) *Graphic shows cervical vertebra from above. Vertebral body is broad transversely, central canal is large and triangular in shape, pedicles are directed posterolaterally, and laminae are delicate. Lateral masses contain the vertebral foramen for passage of vertebral artery and veins.* **(Right)** *Mid-C5 body at the pedicle level: Transverse foramina are prominent, encompassing the vertical course of vertebral artery. The anterior and posterior tubercles give rise to muscle attachments in neck.*

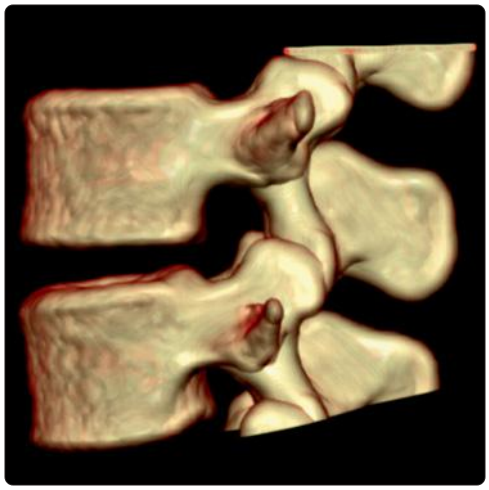




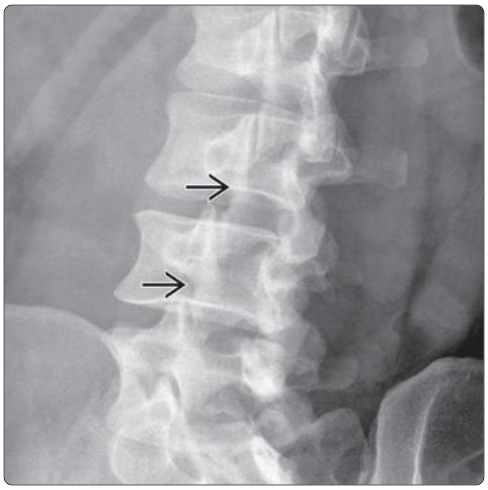
(Left) *Graphic shows thoracic vertebral body from above. Thoracic bodies are characterized by long, spinous processes and transverse processes. Complex rib articulation includes both costotransverse joints and costovertebral joints. Facet joints are oriented in the coronal plane.* **(Right)** *Image through the pedicle level of the thoracic spine: The coronal orientation of the facet joints are well identified in this section. The pedicles are thin and gracile, with adjacent rib articulations.*

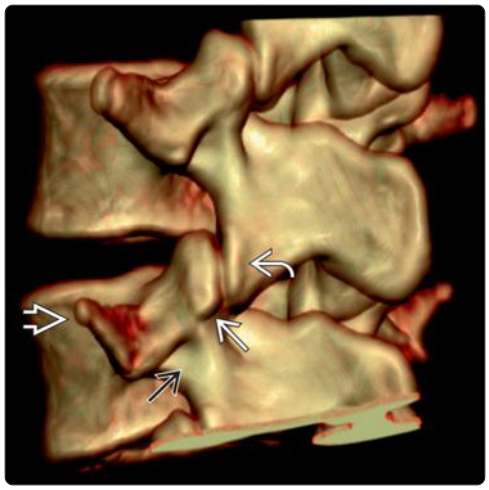
(Left) *Graphic shows lumbar body from above. Large, sturdy lumbar bodies connect to thick pedicles and transversely directed transverse processes. Facets maintain oblique orientation favoring flexion/extension.* **(Right)** *Lateral 3D scan of the lumbar spine shows large bodies joined by thick posterior elements with the superior and inferior articular processes angled in lateral plane. Transverse processes jut out laterally for muscle attachments. Pars interarticularis form a junction between articular processes.*



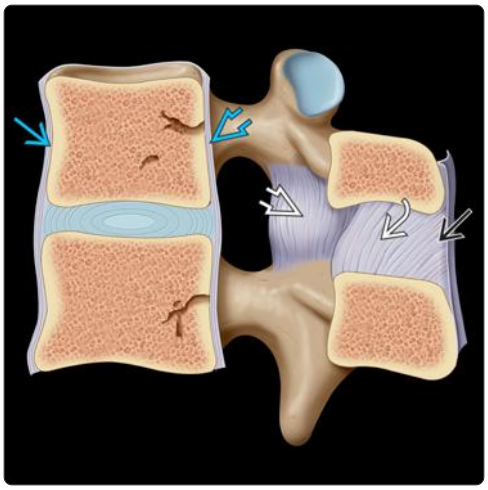


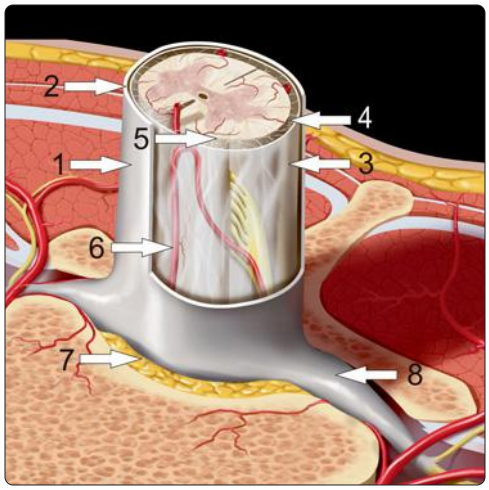
(Left) *Coronal oblique view of the lumbar spine shows the typical "scotty dog" appearance of the posterior elements. The neck of the "dog" is the pars interarticularis.* **(Right)** *Oblique 3D exam of lumbar spine shows surface anatomy of "scotty dog": Transverse process (nose), superior articular process (ear), inferior articular process (front leg), and intervening pars interarticularis (neck). Pedicle that forms "eye" on CT reconstructions is obscured.*

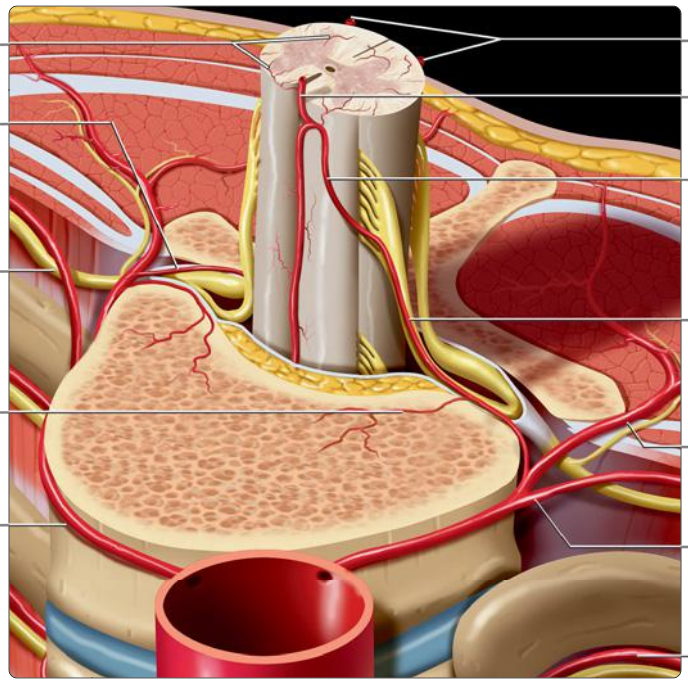


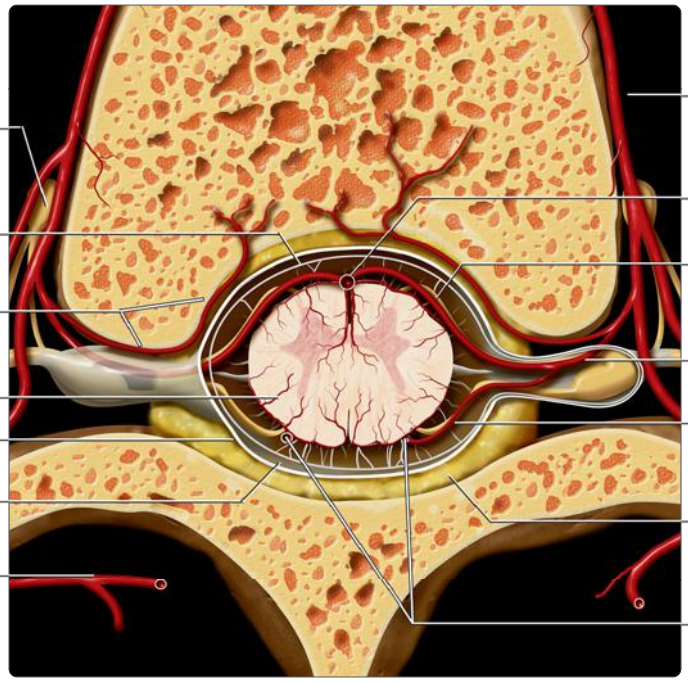


(Left) *Cut-away graphic shows lumbar vertebral bodies joined by disc & anterior & posterior longitudinal ligaments. Paired ligamentum flavum & interspinous ligament join posterior elements, with midline supraspinous ligament.* **(Right)** *Graphic shows spinal cord and coverings: 1) dura mater, 2) subdural space, 3) arachnoid mater, 4) subarachnoid space, 5) pia mater, 6) ant. spinal artery, 7) epidural space, and 8) root sleeve.*









(Top) *Oblique axial graphic of the thoracic spinal cord and arterial supply at T10 shows segmental intercostal arteries arising from the lower thoracic aorta. The artery of Adamkiewicz is the dominant segmental feeding vessel to the thoracic cord, supplying the anterior aspect of the cord via the anterior spinal artery. Adamkiewicz has a characteristic "hairpin" turn on the cord surface as it first courses superiorly, then turns inferiorly.* **(Bottom)** *Axial graphic shows the anterior and posterior radiculomedullary arteries anastomosing with the anterior and posterior spinal arteries. Penetrating medullary arteries in the cord are largely end-arteries with few collaterals. The cord "watershed" zone is at the central gray matter.*

Terminology

Radiographic measurement techniques, skull base craniometry, skull base lines

Pathology-Based Imaging Issues

This chapter provides a broad summary of the varied measurement techniques used for evaluating the spine. The main focus for the reader should be the tables and the multiple schematics that define the variously named lines and angles. These summarize the classic measurement techniques for the skull base, rheumatoid disease, and some of the most commonly used measurements for assessing trauma. The rest of the measurements defined below are a mixture of miscellaneous measurements and those that do not translate well into a table (i.e., equations).

Torg-Pavlov Ratio

- Diameter of canal to width of vertebral body (initially defined on plain radiographs of the subaxial spine)

The practical utility of this measurement is controversial. Less than 0.80 as seen on the lateral view is considered to be cervical stenosis, and such a small canal potentially increases risk for cord injury.

Maximum Canal Compromise (%)

- = 1-(**Di**/[(**Da**+**Db**)/2]) x 100%

AP canal diameter at the normal levels (immediately above and below the level of injury) and at the level of maximum compromise are defined. The measurement of normal levels is taken at the midvertebral body level. **Di** is the anteroposterior canal diameter at the level of maximum injury, **Da** is the AP canal diameter at the nearest normal level above the level of injury, and **Db** is the AP canal diameter at the nearest normal level below the level of injury.

In spinal cord injury patients, midline T1 and T2 images provide an objective, quantifiable, and reliable assessment of cord compression that cannot be defined by CT alone.

Maximum Cord Compression (%)

- = 1-(**di**/[(**da**+**db**)/2]) x 100%

AP cord diameter at the normal levels immediately above and below the level of injury and at the level of maximum cord compression is defined. **di** is the anteroposterior cord diameter at the level of maximum injury, **da** is the AP cord diameter at the nearest normal level above the level of injury, and **db** is the AP cord diameter at the nearest normal level below the level of injury. If cord edema is present, then measurements are made at the midvertebral body level just above or below the extent of the edema where cord appears normal.

Cobb Measurement of Kyphosis

Lines are drawn to mark the superior endplate of the superior next unaffected vertebral body and the inferior endplate of the inferior next unaffected vertebral body, which are then extended anterior to the bony canal. Perpendicular lines are then extended and the angle between the 2 perpendicular lines is measured.

Tangent Method for Kyphosis

Lines are drawn along the posterior vertebral body margin on the lateral view of the affected body and the next most superior body that is unaffected. The angle between these 2 vertically oriented lines is measured.

Centroid

Also called the geometric center of the vertebral body, this measurement is defined by drawing diagonal lines between opposite corners of the body, with the centroid at the intersection.

Apical Vertebral Translation (AVT)

Lateral displacement of the apex of the coronal curve is relative to the **center sacral vertical line** (CSVL) on AP plain film. The AVT is the horizontal distance between the centroid of the apical body and the CSVL.

Sagittal Balance

Sagittal alignment is defined on the lateral view using a C7 plumb line. The distal reference point is the posterior superior aspect of the sacrum. There is a positive number if C7 plumb line falls anterior to the reference point, and a negative number if it falls posterior to the reference.

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Common CV Junction Measurements

Measurement	Definition	Normal	Abnormal
Chamberlain (palatooccipital) line	Posterior hard palate to opisthion	< 2.5 mm of dens above line	Dens > 2.5 mm above line
McGregor (basal) line	Posterior hard palate to lowest point of occipital bone	Tip of dens < 4.5 mm above line	Tip of dens > 4.5 mm above line
McRae line	Basion to opisthion	Entire dens below line	< 19 mm
Wackenheim clival line	Dorsum sellae to tip of clivus	Entire dens ventral to line	Dens bisects line
Fischgold digastric line	Connects the 2 digastric fossae	Entire dens below line	Dens bisects line
Fischgold bimastoid line	Connects tips of mastoid processes	Tip of dens 3 mm below to 10 mm above line	Tip of dens > 10 mm above line
Anterior atlantodental interval	Posterior aspect of anterior C1 arch to anterior margin odontoid process	Plain films in children: < 4-5 mm; plain film in adults: Males < 3.0 mm, females < 2.5 mm; sagittal CT reformats in children: < 2.6 mm; sagittal CT in adults: Both males and females < 2.0 mm	Plain films in children: > 4-5 mm; plain film in adults: Males > 3.0 mm, females > 2.5 mm; sagittal CT reformats in children: > 2.6 mm; sagittal CT in adults: Both males and females > 2.0 mm
Atlantooccipital joint space	Line from midpoint of occipital condyle to C1 condylar fossa	Plain films in children: < 5 mm; CT in children: < 2.5 mm	Plain films in children: > 5 mm; CT in children: > 2.5 mm
Summed condylar distance	Sum of bilateral distances between midpoint of occipital condyle and C1 condylar fossa	CT in adults: < 4.2 mm	
Atlantoaxial joint space	Line defining midpoint of C1-C2 joint on coronal view	CT in adults: < 3.4 mm; CT in children: < 3.9 mm	CT in adults: > 3.4 mm; CT in children: > 3.9 mm

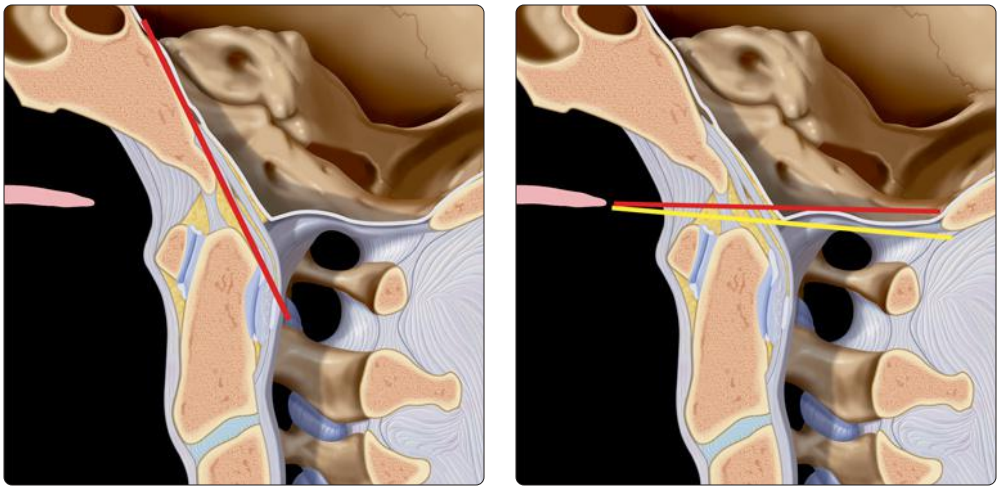
Rheumatoid Arthritis Measurements

Measurement	Definition	Normal	Abnormal
Ranawat	Distance between center of C2 pedicle and transverse axis of atlas measured along axis of odontoid process	< 15 mm in males, < 13 mm in females	≥ 15 mm in males, ≥ 13 mm in females
Redlund-Johnell line	Distance between McGregor line and midpoint of caudal margin C2	< 34 mm in males, < 29 mm in females	≥ 34 mm in males, ≥ 29 mm in females
Clark stations	Dividing odontoid process into 3 parts in sagittal plane	Anterior ring of atlas is level with 1st station	Anterior ring of atlas is level with middle 1/3 (2nd station) or caudal 1/3 (3rd station)
Posterior atlantodental interval (PADI)	Horizontal distance from posterior dens to anterior aspect of C1 lamina or ring		Smaller is worse and relates to potential neurologic deficit
Space available for cord (SAC)	AP diameter on sagittal MR study between dorsal and ventral dura	> 13 mm	SAC < 13 mm then decompression in RA patients considered
Spinal canal diameter (SCD)	Horizontal measurement from posterior vertebral body to spinolaminar line		On plain films < 14 mm, canal stenosis warrants MR for further evaluation

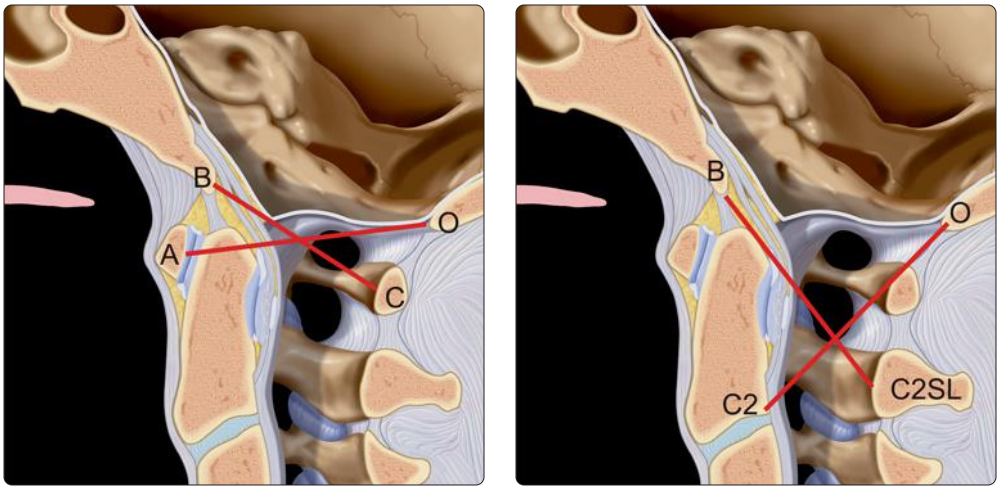
CV Junction Trauma Measurements

Measurement	Definition	Normal	Abnormal
Basion dental interval (BDI)	Basion to superior aspect of odontoid process	< 12-12.5 mm in children on plain films, < 10.5 mm in children by sagittal CT, < 8.5 mm in adults	> 12 mm (Harris measurement)
Basion axial interval (BAI)	Distance between basion and a line drawn along posterior cortical margin of C2	0-12 mm on plain films	Highly variable and not recommended as primary diagnostic method
Powers ratio	Ratio of distance between basion and C1 posterior arch divided by distance between opisthion and midpoint of posterior aspect of anterior C1 arch (BC/OA)	< 1.0	> 1.0 (anterior dislocation only) posterior dissociation or vertical distraction could be missed with normal value

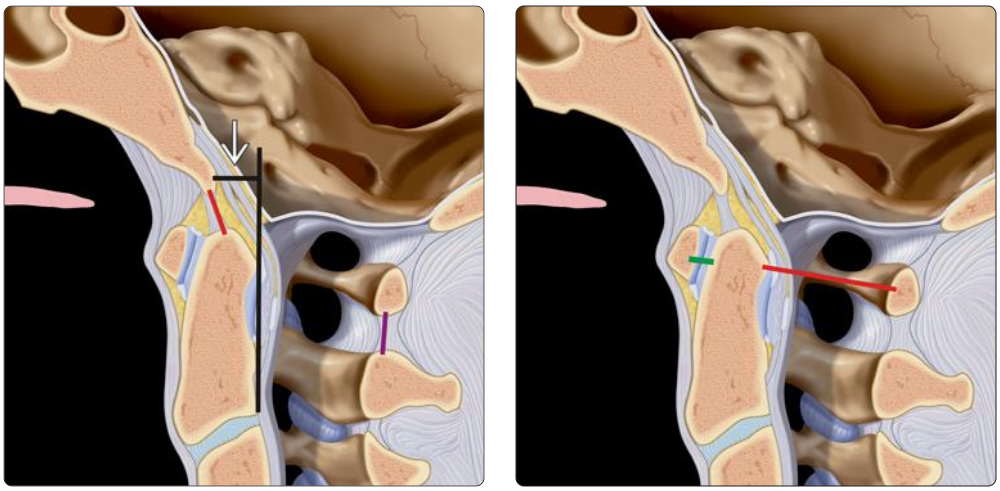
(Left) *Sagittal graphic of craniocervical junction shows Wackenheim clival line (red) extending tangent to the normal odontoid position. The line is drawn from the dorsum sellae to the tip of clivus.*
(Right) *Sagittal graphic of the craniocervical junction shows Chamberlain line (red), drawn from the posterior hard palate to opisthion, and McGregor line (yellow), drawn from the posterior hard palate to the lowest point of the occipital bone.*



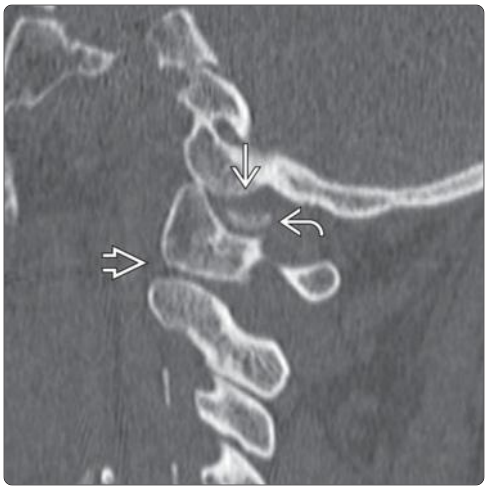
(Left) *Sagittal graphic of craniocervical junction shows lines comprising the Powers ratio (BC/OA), where normal is < 1. BC = basion to C1 posterior arch, OA = opisthion and midpoint of the posterior aspect of anterior C1 arch.*
(Right) *Sagittal graphic shows lines comprising the Lee method. BC2SL and C2O should just intersect tangentially with the posterosuperior aspect of the dens and the highest point on the atlas spinolaminar line respectively in normal state. Deviation suggests AOD.*



(Left) *Sagittal graphic shows basion dental interval (BDI) in red, which should be < 12-12.5 mm in children on plain films and < 8.5 mm in adults on CT. Black lines define basion axial interval (BAI) ➡ extending from basion to line extended along posterior margin of C2, which should be < 12 mm on plain films. C1-C2 spinolaminar line is shown in purple (< 8 mm in adults).*
(Right) *Sagittal graphic shows atlantodental interval (green) and spinal canal diameter (red).*





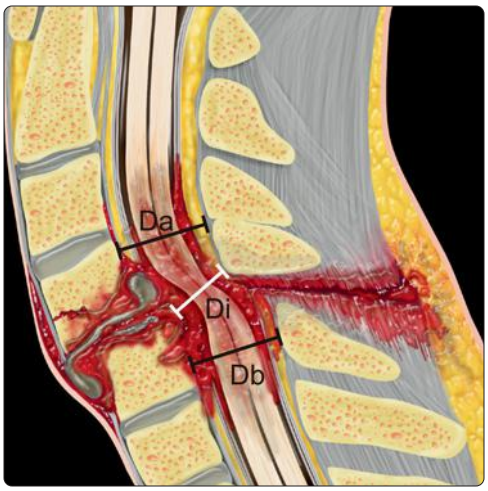


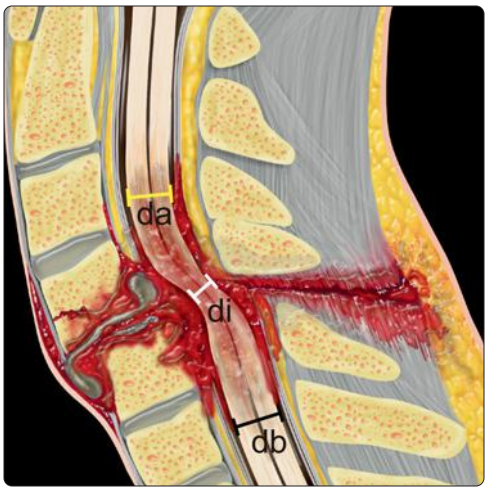
(Left) *Sagittal CT reconstruction in AOD shows widening ➡ of the BDI > 8.5 mm in this adult. Note the normal Wackenheim line relationship.* **(Right)** *Sagittal CT reconstruction in AOD shows widening of the C0-C1 junction ➡ with anterior subluxation of the condyle. Condylar fragment is present in the joint space ➡. Note the normal C1-C2 relationship ➡.*





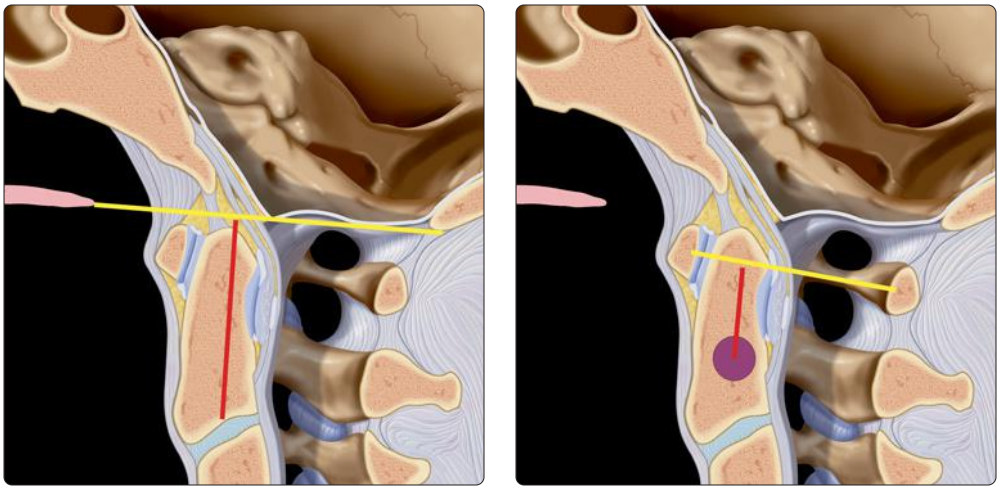
(Left) *Sagittal STIR MR in AOD shows widening of BDI with ↑ T2 signal ➡ from alar and apical ligament rupture. Note also prevertebral edema ➡ and posterior interspinous ligament disruption ➡. Posterior epidural hemorrhage ➡ contributes to subarachnoid space narrowing.* **(Right)** *Coronal CT reconstruction in AOD shows marked widening of the C0-C1 joint space ➡, which should be approximately 2 mm. Bilateral symmetric avulsion fractures off of the condyles are present ➡.*



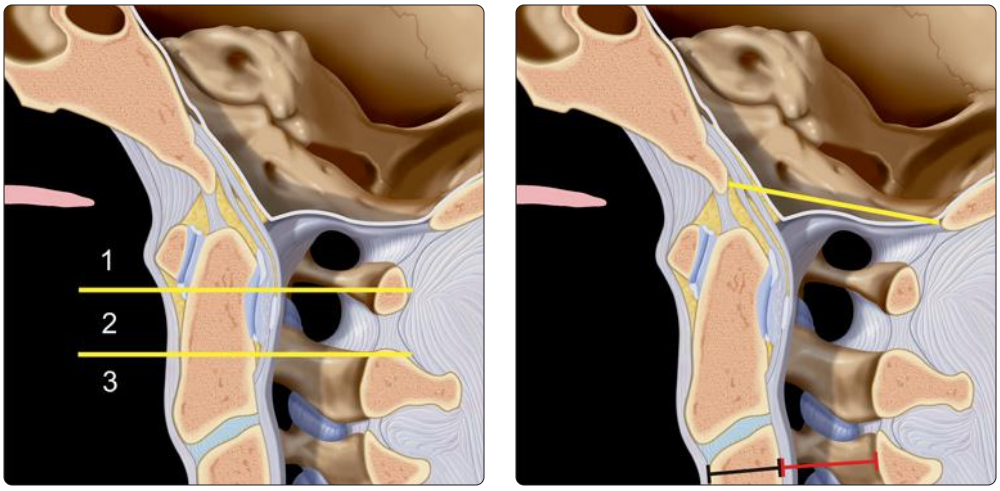


(Left) *Sagittal graphic shows the maximum canal compromise measurement. Di is the anteroposterior canal diameter at the level of maximum injury, Da is the AP canal diameter at the nearest normal level above the level of injury. Db is AP canal diameter at nearest normal level below level of injury.* **(Right)** *Sagittal graphic shows maximum cord compression measurement. di is AP cord diameter at level of maximum injury. da is AP cord diameter at nearest normal level above injury. db is AP cord diameter at nearest normal level below injury.*

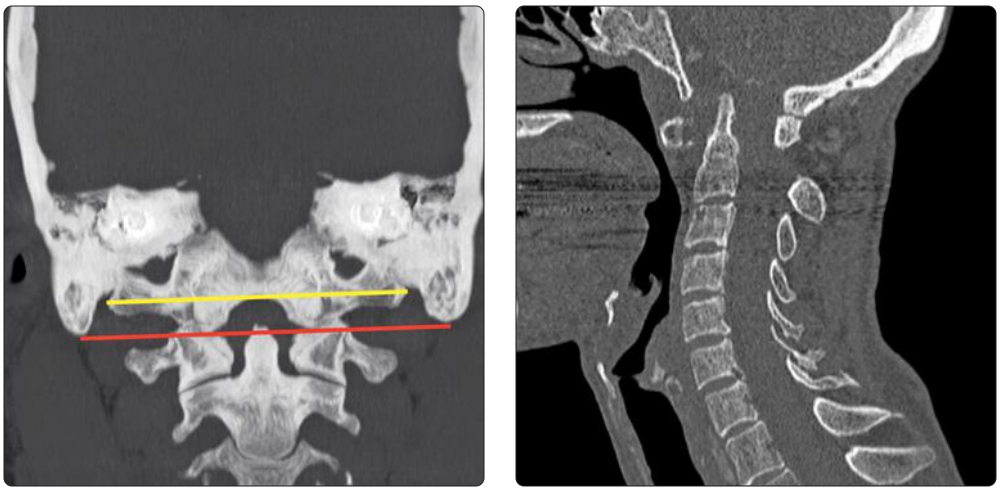
(Left) *Sagittal graphic shows Redlund-Johnell line (red) defined by distance between McGregor line (yellow) and midpoint of caudal margin of C2 body.* **(Right)** *Sagittal graphic shows Ranawat measurement as the distance between the center of C2 pedicle (purple) and transverse axis (yellow) of the atlas measured along the axis of the odontoid process.*

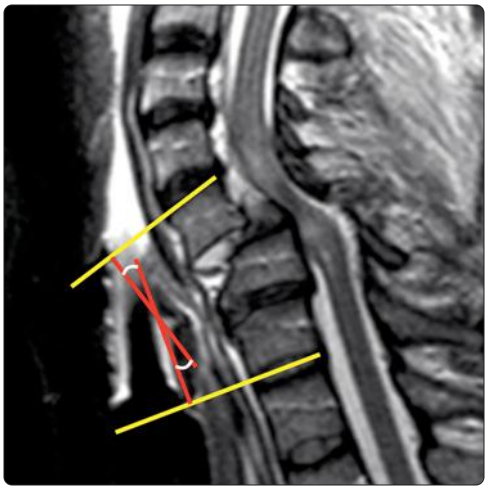


(Left) *Graphic shows the 3 Clarke stations for measuring basilar impression in RA. If anterior ring of C1 is level with the 2nd or 3rd station, then basilar impression is present.* **(Right)** *Sagittal graphic shows 2 different measurements: McRae line (yellow) is defined from basion to opisthion, and the dens should be below this line. Length of McRae should be > 19 mm. Lower measurement shows measurements of Torg-Pavlov ratio, which is the diameter of canal (red) to width of vertebral body (black). Normal is > 0.8.*



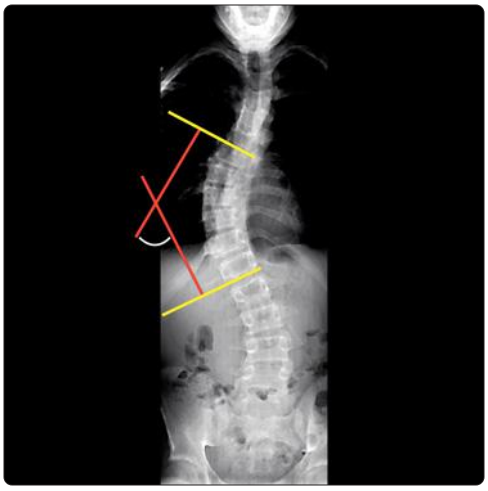
(Left) *Coronal CT shows Fischgold digastric line (yellow) connecting the 2 digastric fossae (normal when dens below the line), and Fischgold bimastoid line (red) connecting the mastoid processes (abnormal if tip of dens > 10 mm above line).* **(Right)** *Sagittal CT shows upward translocation of odontoid with Wackenheim clival line and Chamberlain line grossly abnormal in this patient with RA. Odontoid is eroded with thinned pencil-tip appearance. Note markedly increased atlantodental interval.*







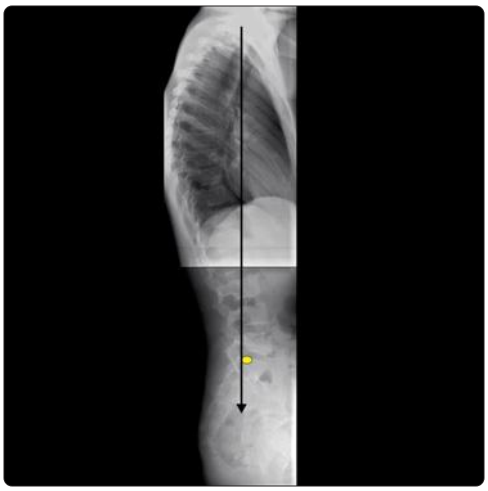
(Left) *Sagittal graphic shows Cobb angle method for cervical kyphosis. Lines are drawn along superior endplate of cephalad unaffected body and inferior endplate of caudal unaffected body (yellow). The angle of perpendiculars (red) from these lines is considered Cobb angle (white).* **(Right)** *Sagittal graphic shows posterior vertebral body tangent method. Lines extend from the posterior body at fractured level and superior unaffected level. Distance can be measured (red) or angle of lines determined.*





(Left) *AP view shows Cobb measurement. Lines are drawn (yellow) along end vertebrae, which are upper and lower limits of curve tilting most severely toward concavity. Perpendiculars are drawn from the endplate lines (red), with angle measurement (white).* **(Right)** *Anteroposterior radiograph shows measurement of overall coronal plane balance by the distance (yellow) from C7 plumb line (white) to central sacral vertical line (CSVL) (black). Positive displacement is to the right, negative to the left.*





(Left) *Anteroposterior radiograph shows measurement of apical vertebral translation (AVT). Distance (arrowed line) from centroid of apex of curve (yellow) is measured relative to CSVL (black).* **(Right)** *Lateral radiograph shows measurement of sagittal balance. Horizontal displacement measured from plumb line extended from centroid of C7 (black line) to posterior superior margin of the sacrum (yellow). Positive balance when line is anterior to reference point.*

KEY FACTS

TERMINOLOGY

- MR imaging artifacts that simulate disease

IMAGING

- Artifactual "pseudolesion" is usually bizarre or nonanatomic in appearance or distribution
  - May be detected anywhere in spine
- Common artifacts
  - Truncation ("Gibbs") artifact
  - Phase ghosting artifact
  - Motion artifact
  - CSF flow artifact
  - Chemical shift artifact
  - Wraparound artifact (aliasing)
  - Susceptibility artifact
  - Zipper artifact
  - Gradient warping artifact
  - Fat-saturation failure ± inappropriate water saturation
  - Normal marrow T1 hypointensity at high field strength (≥ 3.0 tesla)

TOP DIFFERENTIAL DIAGNOSES

- Syringohydromyelia
- CSF drop metastases
- Aneurysm or arteriovenous malformation
- Spinal cord hemorrhage
- Marrow infiltration or replacement

CLINICAL ISSUES

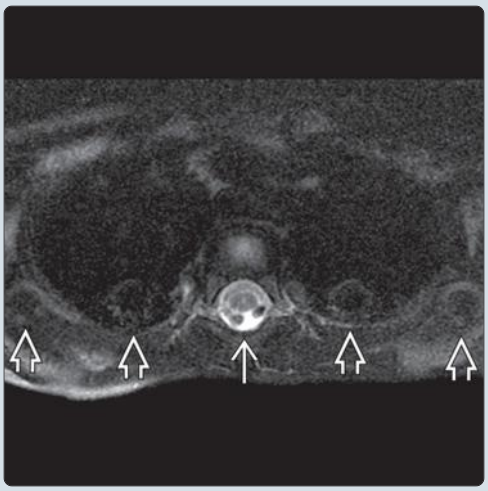
- Artifact location often unrelated to clinical findings

DIAGNOSTIC CHECKLIST

- Always consider MR artifacts when confronted with bizarre imaging findings

(Left) *Sagittal STIR MR demonstrates linear T2 hyperintensity ➡ within the lower thoracic spinal cord extending to the conus, representing motion artifact from abdominal wall movement. The appearance is suspicious for artifact, because it continues to the conus tip, unusual with true syringomyelia.* **(Right)** *Axial T2WI FS MR demonstrates prominent periodic ghosting artifact ➡ of the thecal sac ➡ propagated across the image in the phase direction.*





(Left) *Sagittal T2WI MR obtained for CSF drop metastasis surveillance shows bizarre intradural T2 hypointensity ➡, representing marked CSF pulsation artifact in the thoracic spine.* **(Right)** *Axial T2WI MR (imaging surveillance for CSF drop metastases) confirms intradural low signal intensity CSF pulsation artifact ➡. Correct interpretation as artifact is possible by recognizing that this low signal intensity does not resemble either a normal anatomic structure or typical drop metastasis.*





TERMINOLOGY

Abbreviations

- Specific absorbed radiation (SAR), cerebrospinal fluid (CSF), radiofrequency (RF)

Synonyms

- "Phase ghosting," "Gibbs" artifact, "blooming" artifact

Definitions

- MR imaging artifacts that simulate disease

IMAGING

General Features

- Best diagnostic clue
  - Artifactual "pseudolesion" is usually bizarre or nonanatomic in appearance or distribution
- Location
  - May be detected anywhere in spine
    - Appearance depends on artifact type
- Size
  - Variable
- Morphology
  - Bizarre appearance that seems nonphysiological or nonanatomical
    - MR is very prone to imaging artifacts
    - Fortunately, most are pretty unique in appearance, readily identified if reader is aware of their existence
  - Compare with CT motion artifacts, which often simulate spine fractures or congenital anomalies on reformatted images

MR Findings

- Truncation ("Gibbs") artifact
  - Mathematical artifact related to truncation of computational series used during Fourier transformation of K-space data
    - Not possible to calculate infinite series when reconstructing raw data
    - Mathematical equation is "truncated" or shortened to reflect practical necessity of sampling finite, rather than infinite, number of frequencies
  - May be detected in both phase and encoding directions, although more conspicuous in phase direction, because fewer samples usually obtained
  - Occurs at high contrast interfaces, producing alternating light and dark bands that may mimic lesions
  - In spine, may artifactually widen or narrow spinal cord or mimic syrinx
  - Decrease artifact conspicuity by ↑ number of phase encoding steps or ↓ field of view (FOV)
- Phase ghosting artifact
  - Periodic replication of structure along line in phase encoding direction
  - Ghost artifacts, in particular, occur whenever there is periodic motion within field of view FOV
    - Examples include blood flow in vessels, CSF pulsations, cardiac, and respiratory motion
  - Like many artifacts, phase ghosting is more conspicuous in phase encoding direction, because sampling times are much slower than in frequency encoding direction
    - Thus, these artifacts are propagated in phase direction regardless of motion direction
- Motion artifact
  - Voluntary or involuntary patient movement (random) or pulsating flow in vessels (periodic)
  - Detected in phase encoding direction
  - May simulate pathologic intramedullary or intradural lesion
    - Usually recognizable as artifact, but may render study limited or nondiagnostic for cord pathology
  - If motion is nonperiodic (e.g., bowel peristalsis), ghosting will not occur, but generalized image degradation will be apparent
  - Address with patient instruction, respiratory compensation, sedation, faster scanning
- CSF flow artifact
  - Subset of periodic motion artifact
  - Dephasing of protons due to CSF motion may simulate intradural blood, disc herniation, CSF metastasis, or intramedullary lesion
  - Flow compensation techniques can decrease artifact conspicuity
- Chemical shift artifact
  - Protons precess at slightly different frequencies in fat compared to water
    - Difference in proton precessional frequency between water and fat at 1.5T is 220 Hz
  - Spatial misregistration of fat and water produces overlap that causes additive bright band at lower frequency range and subtractive dark band at higher frequency range
  - Can be useful to confirm presence of macroscopic fat
- Wraparound artifact (aliasing)
  - Occurs when dimensions of imaged object exceed FOV
  - Seen in both frequency and phase encoding directions, although much more conspicuous in phase encoding direction
  - May also be identified in slice select direction on 3D sequences, because of additional phase encoding direction
  - Increasing FOV and number of phase encoding steps helps reduce artifact in phase direction, as does use of "no phase wrap" software
  - Aliasing in frequency direction may be abolished by oversampling above Nyquist frequency
- Susceptibility artifact
  - Metal or blood products disrupt local magnetic field homogeneity, produce signal void or image distortion
  - Common in postoperative instrumented spine
  - New metal-insensitive pulse sequences help reduce artifact
  - Can be used to advantage to diagnose hemorrhagic or calcified lesions
    - e.g., cavernous malformations, spinal cord hemorrhage
- Zipper artifact
  - Family of similar appearing artifacts most commonly occurring in phase encoding direction

- Produced by radiofrequency noise related to scanner equipment, scan parameters, or extrinsic noise (TV or radio channel, flickering fluorescent room light, patient monitor, etc.)
- Usually easy to identify as artifact, but may obscure important findings or create confusing pseudolesion

- Gradient warping artifact
  - Image distortion at edges of large FOV studies (> 30 cm) caused by gradient distortion
  - May be be addressed with gradient distortion correction (GDC)
- Fat-saturation failure ± inappropriate water saturation
  - Frequency selective RF ("fat-saturation") pulse frequency chosen to saturate fat spectral peak center frequency
  - Magnetic field inhomogeneity alters fat center frequency so that fat-saturation pulse frequency no longer overlaps fat spectral peak → fat-saturation failure
  - If fat-saturation pulse frequency inadvertently overlaps water spectral peak → inappropriate water suppression
- Normal marrow T1 hypointensity at high field strength (≥ 3.0 T)
  - Problem arises because of technical parameter changes needed to acquire T1-weighted images at high field in reasonable time without SAR issues
  - Inversion recovery T1WI sequences less SAR intensive than T1 SE sequence, commonly used in 3.0T spine imaging
    - Marrow signal relatively hypointense compared to appearance on SE T1WI, simulates pathological marrow infiltration

Imaging Recommendations

- Protocol advice
  - Minimize artifacts using appropriate parameters, flow compensation, saturation bands, adequate sedation, or comfort measures, etc.

DIFFERENTIAL DIAGNOSIS

Syringomyelia

- True dilation of central spinal cord canal, without (hydromyelia) or with (syringomyelia) underlying cord injury and myelomalacia, eccentric cavitation
- Does not usually extend all the way to conus; may be sacculated
- Simulated by truncation artifact or phase ghosting

CSF Drop Metastases

- Will usually be detectable in at least 2 planes
- Mimicked (or obscured) by CSF pulsation artifact
- Swap phase and frequency if needed to confirm as not an artifact

Aneurysm or Arteriovenous Malformation

- Phase ghosting permits diagnosis of patent flow within vessels or tissue based on periodic ghost artifact
- Absence of this artifact in technically satisfactory MR exam indicates slow flow or thrombosed lesion

Spinal Cord Hemorrhage

- Cavernous malformation, post-traumatic spinal cord hematoma, neoplasm
- Mimicked by phase ghosting, RF leak, motion artifact
- Use gradient recalled echo imaging (GRE) to capitalize on susceptibility artifact, confirm blooming of hypointense signal in hemorrhagic lesion

Marrow Infiltration or Replacement

- Marrow replacement or ablation, fibrosis
- Hematopoietic neoplasms, marrow replacement, or ablation diseases, osteopetrosis
- Mimicked by use of T1 FLAIR technique for T1WI at higher field strengths (≥ 3.0T)
- Results in lower normal marrow signal intensity than seen on spin-echo T1WI sequences

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Artifact location often unrelated to clinical findings
  - Exception is susceptibility artifact in cases of hemorrhage, metallic foreign body, or medical devices

Natural History & Prognosis

- Not applicable

Treatment

- Not applicable

DIAGNOSTIC CHECKLIST

Consider

- Artifacts often have characteristic appearance, recognizable if imager is aware of and considers artifact

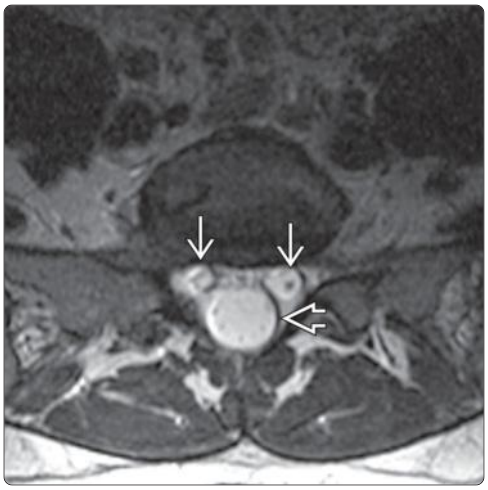
Reporting Tips

- Always consider MR artifacts when confronted with bizarre imaging findings
  - If MR artifact cannot be excluded, consider true pathology
  - Clinical context always crucial to avoid failure to consider important pathologic differential considerations

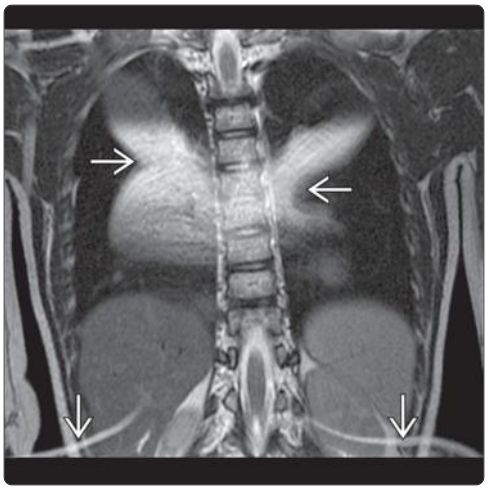
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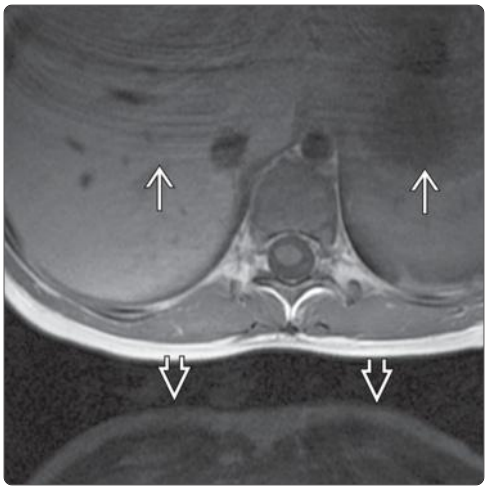
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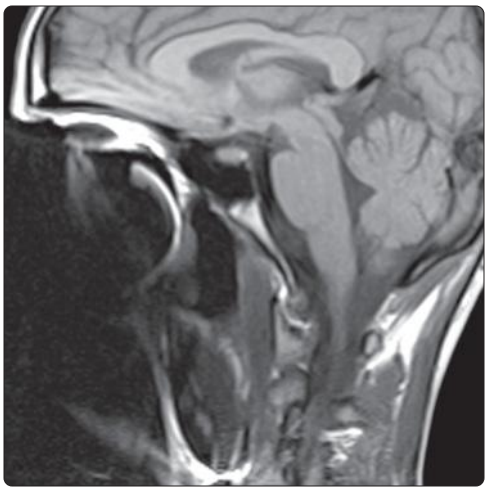
(Left) *Axial T2WI MR in a patient with tethered spinal cord shows classic chemical shift artifact ➡, confirming presence of fat within the filum. Pattern of alternating hyper- and hypointensity confirms that frequency direction is AP.* **(Right)** *Axial T2WI MR depicts prominent chemical shift artifact in the thecal sac ➡ and nerve root sleeves ➡ reflecting presence of water (CSF in root sleeves) and fat (epidural) within adjacent voxels. Frequency direction is right to left.*





(Left) *Coronal T2WI MR demonstrates artifactual wrap of back fat ➡ onto the thoracic spine. Such artifacts could be diminished by increasing the field of view (FOV), at the cost of spatial resolution, unless the number of phase encoding steps is also increased.* **(Right)** *Axial T1WI MR depicts propagation of respiratory motion ➡ through the abdomen and spine, as well as phase wrap of the abdomen behind the back ➡.*





(Left) *Sagittal T2WI MR shows marked signal loss and image degradation in the upper thoracic spine, reflecting attempted MR imaging with the incorrect spine coil elements turned on. There is also a prominent zipper artifact ➡ running in the craniocaudal direction.* **(Right)** *Sagittal T1WI MR reveals marked distortion of the inferior frontal lobes and facial soft tissues reflecting severe metallic dental brace artifact.*

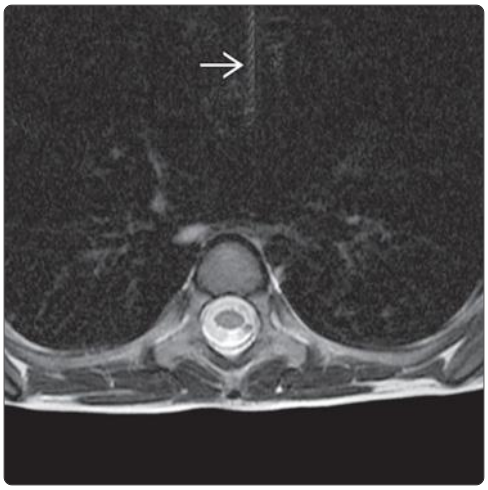
(Left) *Sagittal T1WI MR (suprasellar craniopharyngioma, postoperative) performed for spinal surveillance with known CSF craniopharyngioma dissemination shows a metastatic lesion ⇒ anterior to the pons.* **(Right)** *Sagittal T1WI C+ FS MR (suprasellar craniopharyngioma, known CSF metastases) demonstrates a prominent band of susceptibility artifact ⇒ 2° to inhomogeneous fat saturation, unfortunately nearly completely obscuring known CSF metastasis ⇒.*



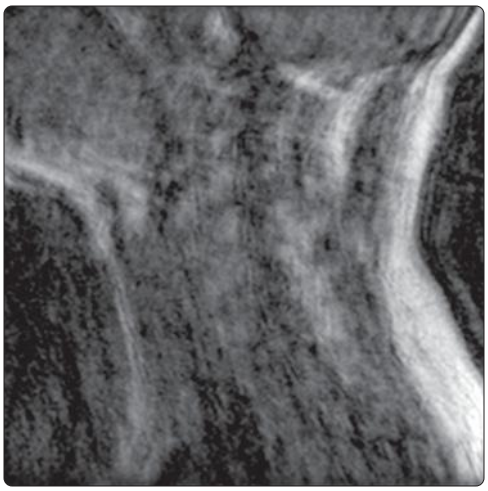


(Left) *Sagittal T1WI MR shows severe radiofrequency interference artifact creating horizontal stripes across the image producing mild image degradation. Note also the incomplete zipper artifact ⇒ at the inferior margin of the image center.* **(Right)** *Axial T2WI MR of the thoracic spine demonstrates a prominent zipper artifact ⇒ in the upper center of the image. Like most zipper artifacts, this does not seriously compromise the diagnostic quality of the exam.*

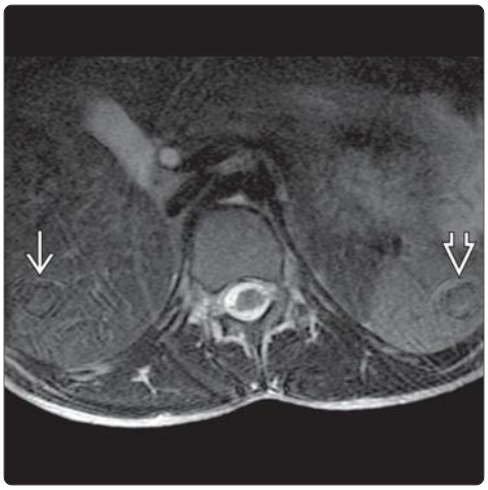


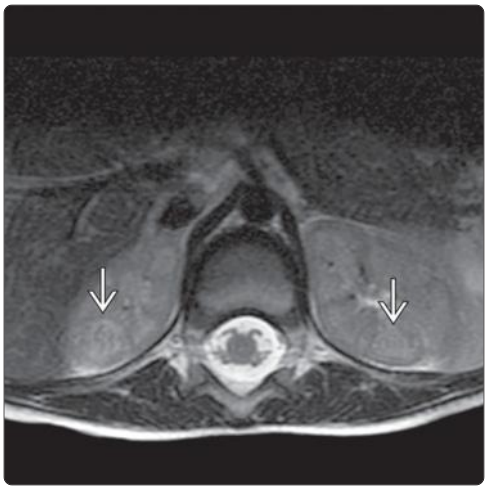


(Left) *Sagittal T1WI MR in an uncooperative, delirious patient shows severe motion artifact that markedly degrades the image quality and renders it nondiagnostic. It is important not to try to use these images in a diagnostic attempt, because artifact may either simulate or obscure a lesion.* **(Right)** *Axial T1WI C+ FS MR shows severe motion artifact running in the AP direction reflecting respiratory movement of the abdominal wall. There is also periodic phase ghosting of the aorta ⇒.*







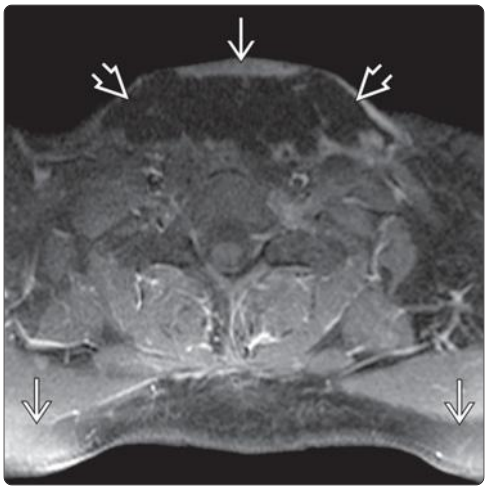


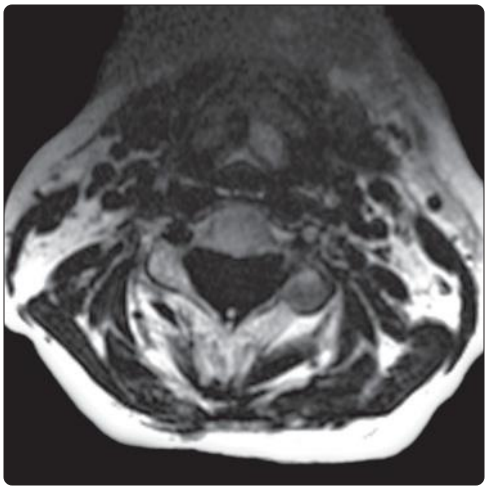
(Left) *Axial T2WI MR depicts periodic phase encoding artifact from the thecal sac simulating lesions in the liver ⇨ and spleen ⇨. The spacing between the thecal sac and the artifactual ghosts is precisely the same distance, a characteristic finding with periodic phase ghosting artifact. Recognizing this assists in making the diagnosis of artifact.* **(Right)** *Axial T2WI MR shows periodic phase ghosting from the thecal sac and spinal cord simulating bilateral renal lesions ⇨.*





(Left) *Sagittal T1WI MR demonstrates gradient warping artifact of the lower thoracic spine. This artifact is characterized by image distortion ⇨ at the edges of large FOV studies (esp. > 30 cm) caused by gradient distortion.* **(Right)** *Sagittal T2WI MR depicts unacceptable phase ghosting artifact ⇨ that mimics cervical syringohydromyelia in this case arising from failure to place an anterior saturation band in front of the cervical spine.*





(Left) *Axial T1WI C+ FS MR shows inhomogeneous fat suppression using chemical fat-saturation technique, with bright areas of fat-suppression failure ⇨, as well as dark areas of inappropriate water saturation ⇨.* **(Right)** *Axial T1WI C+ FS MR shows complete failure of chemical fat-saturation technique, compounded by unwanted inappropriate water saturation that nullifies all water signal. This artifact is commonly encountered when imaging complex shapes such as the cervicothoracic junction.*

KEY FACTS

TERMINOLOGY

- Normal anatomical variations that simulate pathological conditions

IMAGING

- Multiplanar MR best for soft tissue evaluation
- Radiography, bone CT most useful for bone anatomy

TOP DIFFERENTIAL DIAGNOSES

- Traumatic or degenerative vertebral subluxation
- Transverse process fracture
- Vertebral segmentation and formation anomalies
- Vertebral hemangioma
- Intervertebral disc herniation
- Nerve sheath tumor
- Posterior spinal dysraphism
- Caudal regression spectrum

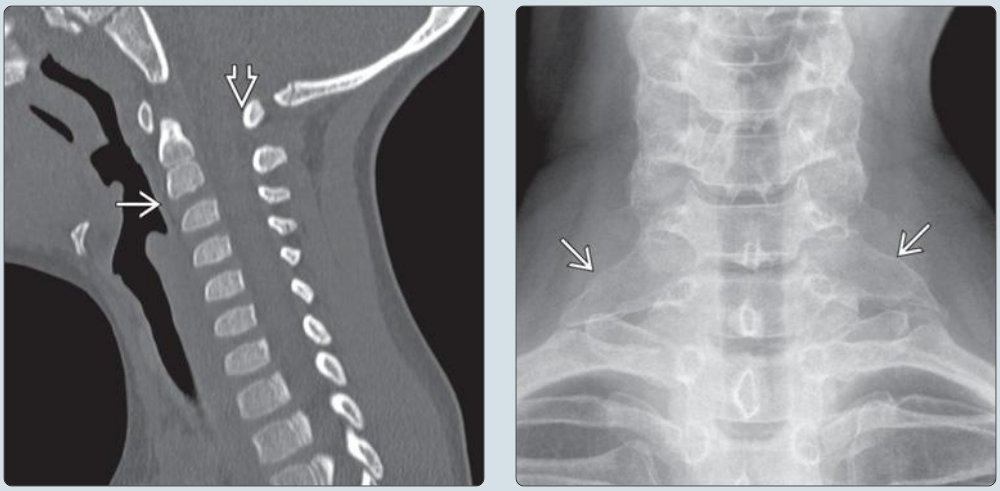
CLINICAL ISSUES

- Discovered incidentally when patient is imaged for other indications
  - Patient asymptomatic, or presenting symptoms do not match location of finding
- Normal life expectancy, no incremental morbidity
- May lead to unnecessary diagnostic tests or treatment if not recognized as normal variant

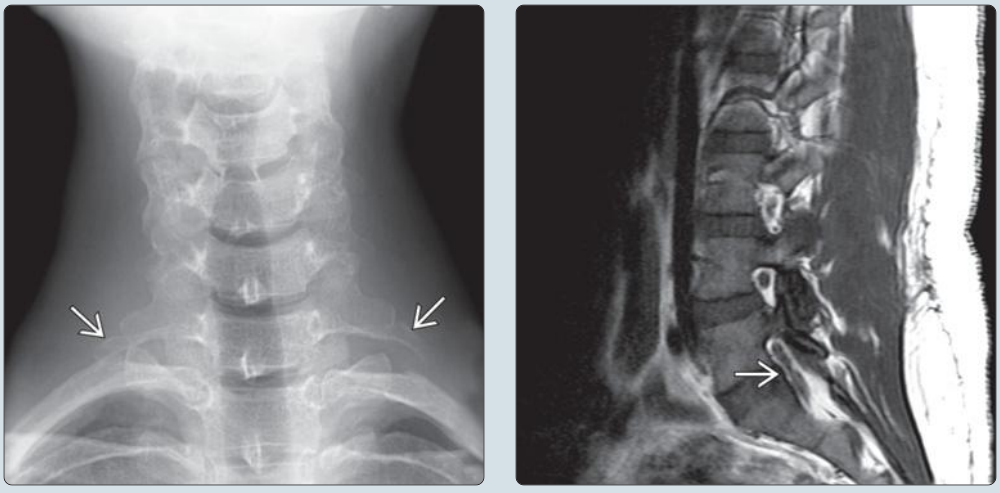
DIAGNOSTIC CHECKLIST

- Many normal variants are common and readily recognized by experienced observers
- Some are uncommon and may not be recognized as normal variant, require high index of suspicion to correctly diagnose
- Consider normal variant within differential diagnostic considerations when unexpected finding is detected

(Left) *Sagittal bone CT in a young pediatric patient demonstrates slight apparent subluxation of C2 on C3 ➡. All other spinal lines are normal, particularly the posterior spinolaminar line ➡ from C1 to C3, confirming pseudosubluxation.* **(Right)** *Anteroposterior cervical radiograph shows unusual elongation of the C7 transverse processes ➡. In some patients, draping of the lower brachial plexus trunk over the C7 transverse process may produce a clinical brachial plexopathy.*



(Left) *Anteroposterior cervical radiograph demonstrates small bilateral rudimentary C7 cervical ribs ➡. Downward angulation of the adjacent transverse processes distinguishes these from hypoplastic T1 thoracic ribs.* **(Right)** *Sagittal T1WI MR along the plane of the medial aspect of the L5 pedicle shows a vertically oriented root ➡ spanning the L5 disc level due to a conjoined nerve root. This vertical orientation of the exiting inferior root is typical, along with the more horizontal course of the superior root.*



TERMINOLOGY

Definitions

- Normal anatomical variations that simulate pathological conditions

IMAGING

Cervical Vertebral Pseudosubluxation

- Classically most conspicuous at C2/3 level, with apparent anterior subluxation of C2 on C3 with head in flexed position
- Observed in younger pediatric patients with incomplete ossification of upper cervical spine
  - Prevalence in older patients controversial, may represent true ligamentous injury
- Absence of spinolaminar line disruption and patient age keys to correct recognition

Incomplete C1 Ring

- C1 ring is incompletely ossified
- In absence of neurological abnormalities or documented instability, asymptomatic normal variant detected during imaging for other reasons

Cervical Ribs

- Small rudimentary ribs at C7
  - Elongation of C7 transverse processes is close variant with similar clinical findings
- Usually asymptomatic but may produce brachial plexopathy or thoracic outlet symptoms
- Orientation of transverse processes key to distinguishing cervical from thoracic ribs
  - Cervical transverse processes point caudal, while thoracic transverse processes point rostral

Transitional Vertebral Anatomy

- Variant osseous anatomy at thoracolumbar or lumbosacral transitions
- Common variants include S1 "lumbarization," L5 "sacralization," rudimentary L1 ribs, hypoplastic T12 ribs
- Confound correct counting of vertebral levels
  - Usually not clinically significant issue if counting method well described in imaging report
- May predispose to accelerated degenerative changes at mobile segments above or below variant levels

Focal Fatty Marrow

- Focal fat conglomeration within vertebral marrow
- Follows fat signal and density on all sequences
  - Fat-saturation MR sequences helpful to confirm diagnosis
- Primary clinical impact is mimicry of vertebral hemangioma

Ectopic Kidney

- Pelvic kidney lower than orthotopic location, more midline than expected
- May mimic prevertebral neoplasm
- Correct identification of corticomedullary reniform architecture critical to avoid unnecessary biopsy or resection

Conjoined Nerve Root

- 2 adjacent spinal nerve roots exit through a common aberrant nerve root sleeve
- Nerves subsequently either exit individually through correct-numbered neural foramina or both through a single foramen
- Mimics nerve sheath tumor or intervertebral disc herniation on imaging

Incomplete Posterior Lumbar Elements

- Incomplete fusion of L5 or S1 posterior elements
  - Term "spina bifida occulta" outdated and use is discouraged
- Commonly observed in asymptomatic population
- Should be considered normal variant in absence of cutaneous abnormalities or referable neurological abnormalities

High Thecal Sac Termination

- Normal thecal sac termination usually inferior S2 level
  - Range from lower 1/3 of L3 → upper 1/3 of S5
  - < 5% terminate above S1 level
- Normal neurological examination expected with incidental anatomic variant
- Spinal cord anomalies or sacral dysplasia prompts consideration of caudal regression spectrum

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR best for soft tissue evaluation
  - Plain radiographs, bone CT most useful for working out bone anatomy

DIFFERENTIAL DIAGNOSIS

Traumatic or Degenerative Vertebral Subluxation

- Mimics cervical pseudosubluxation
- True listhesis secondary to trauma or degenerative changes
- Characteristically demonstrates spinolaminar line disruption, in contradistinction to cervical pseudosubluxation
- Concurrent fractures or degenerative osseous changes common
- Patients generally much older than pseudosubluxation age range

Transverse Process Fracture

- Mimics cervical rib or hypoplastic thoracic or lumbar rib
- Sharp, noncorticated edge at fracture site
- ± paraspinal hematoma, other injuries

Vertebral Segmentation and Formation Anomalies

- True failure of normal vertebral formation or segmentation
  - Butterfly and hemivertebra, block vertebra, posterior element fusion
- Mimics variant transitional thoracolumbar or lumbosacral anatomy
- Consider syndromal associations, concurrent visceral anomalies

Vertebral Hemangioma

- Mimics focal fatty vertebral marrow

- Most are asymptomatic and incidentally discovered on imaging for other indications
  - Small percentage have atypical vascular (nonfatty) stroma, may extend beyond vertebral body, pathological fracture
  - Primary clinical importance has resemblance to primary or metastatic vertebral malignancy
- Round shape, coarsened vertebral trabeculae help confirm benign hemangioma diagnosis

Intervertebral Disc Herniation

- Mimics conjoined nerve root, nerve sheath tumor
- Protean imaging appearances
  - Disc fragment does not homogeneously enhance, helping to distinguish from a nerve sheath tumor or normal dorsal root ganglion
  - May resemble conjoined nerve root; look for increased number of nerves in root sleeve, aberrant nerve sheath origin

Nerve Sheath Tumor

- Mimics conjoined nerve root, intervertebral disc herniation
- Neurofibroma, schwannoma most common spinal nerve neoplasms
- Enhancement pattern helps distinguish from normal or conjoined nerve root, herniated intervertebral disc

Posterior Spinal Dysraphism

- Mimics incidental variant incomplete posterior element fusion
- May be open or closed (skin-covered)
- Frequently coexists with low-lying spinal cord and filum anomalies ± lipoma

Caudal Regression Spectrum

- Mimics isolated incidental high thecal sac termination
- < 5 sacral segments + coccyx by definition
- Spinal cord either wedge-shaped, high termination (type 1) or elongated and low-lying (type 2)
- Association with pelvic visceral anomalies, other congenital spinal malformations

PATHOLOGY

General Features

- Etiology
  - Normal variant

Gross Pathologic & Surgical Features

- Concordant with imaging features

Microscopic Features

- Characteristic of tissue composition

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Discovered incidentally when patient is imaged for other indications
  - Patient asymptomatic or presenting symptoms that prompt imaging does not match location of findings

Demographics

- Age
  - Detection possible throughout entire life span
- Epidemiology
  - Variable

Natural History & Prognosis

- Normal variant; does not alter normal life expectancy or produce morbidity
- May lead to unnecessary diagnostic tests or treatment if not recognized as normal variant

Treatment

- None indicated

DIAGNOSTIC CHECKLIST

Consider

- Many normal variants are common
  - Readily recognized by experienced observers
- Some are uncommon enough that may not be recognized as normal variant
  - Require high index of suspicion to correctly diagnose
  - Consider normal variant within differential diagnostic considerations when unexpected finding is detected

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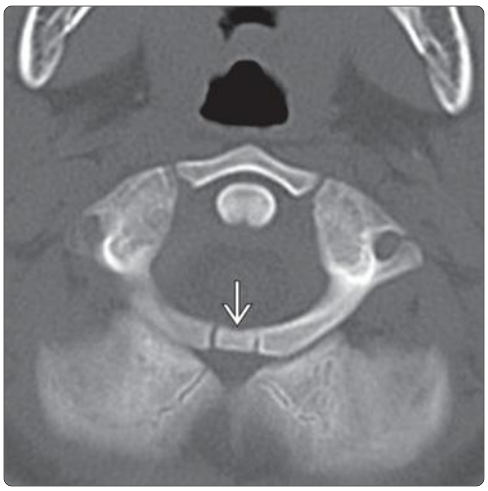


(Left) *Sagittal T1WI MR of the thoracic spine demonstrates focal, well-circumscribed hyperintense signal intensity within a midthoracic vertebral body. There is no disruption of the posterior vertebra. The differential is focal fatty marrows vs. a small hemangioma. Both are of no clinical consequence.* **(Right)** *Anteroposterior radiograph of the lumbar spine obtained for low back pain reveals incidental incomplete posterior fusion of the S1 lamina in the midline.*





(Left) *Anteroposterior radiograph shows incorporation of the L5 transverse processes into the sacral ala bilaterally ("sacralization of L5").* **(Right)** *Axial T1WI MR of the sacral spine depicts 2 ovoid masses within the right S1 foramen representing conjoined nerve roots.*





(Left) *Axial bone CT shows a variant accessory ossification center in the midline of the posterior C1 arch. The anterior C1 ring shows conventional anatomy with a single ossification center. This uncommon variant is asymptomatic and stable.* **(Right)** *Sagittal T1WI MR (normal neurological examination) demonstrates incidentally detected high termination of the thecal sac at the mid-L5 level with a small perineural cyst. The conus terminates at the normal L1 level, and the filum terminale is not thickened.*

KEY FACTS

TERMINOLOGY

- Anomalous or variant craniovertebral junction anatomy

IMAGING

- Flattening or malformation of clivus/occipital condyles, anterior C1 ring, or dens
- Variable instability during dynamic flexion, extension
- ± enhancing granulation tissue mass ("pannus")
  - Reducible abnormalities have largest pannus

TOP DIFFERENTIAL DIAGNOSES

- Developmental CVJ anomalies
  - Achondroplasia, Down syndrome
- Inflammatory and degenerative arthritides
- Acquired basilar impression
- Trauma

PATHOLOGY

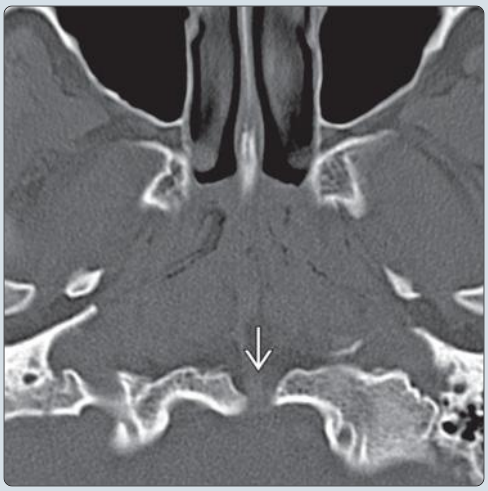
- Congenital insult to developing CVJ neural and osseous tissue between 4th and 7th intrauterine weeks
- Solid ankylosis or fibrous union in many irreducible anomalies
- Granulation tissue proliferation around motion areas in unstable or reducible anomalies

CLINICAL ISSUES

- Occipital headache or suboccipital neck pain exacerbated by flexion/extension
- Symptomatic presentation following mild trauma common

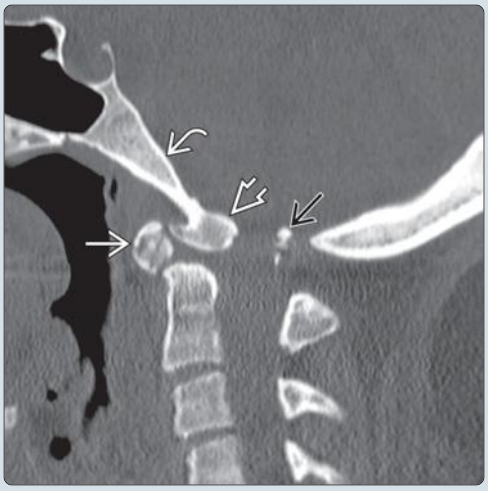
DIAGNOSTIC CHECKLIST

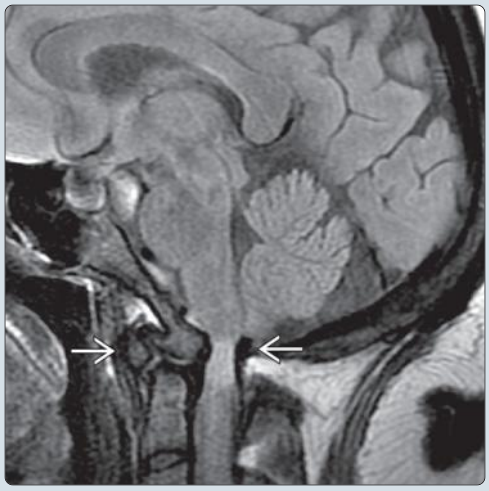
- Dynamic flex-extend imaging determines stability, reducibility of abnormality





(Left) *Axial bone CT (platybasia) reveals an aberrant, flattened clivus with unusual coronal orientation. Also demonstrated is a prominent midline bone cleft defect.* **(Right)** *Sagittal bone CT demonstrates coexisting condylus tertius and os odontoideum in a patient with atlantoaxial instability. Note the osseous protrusion from the clivus tip representing the condylus tertius. The os is dystopic, and the anterior C1 ring is enlarged.*





(Left) *Sagittal bone CT (os avis) reveals abnormal fusion of the os to the clivus. Remodeling changes are also noted in the anterior C1 ring and os. The posterior C1 ring is hypoplastic and resides within the spinal canal, just below the foramen magnum.* **(Right)** *Sagittal FLAIR (os avis, same patient) reveals abnormal T2 signal within the cervical spinal cord at the C1 level, reflecting injury related to atlantoaxial instability and mild C1 ring hypoplasia.*

TERMINOLOGY

Abbreviations

- Craniovertebral junction (CVJ), craniocerebral junction (CCJ)

Synonyms

- CVJ anomalies, CCJ variants or anomalies

Definitions

- Anatomical variations of skull base-upper cervical spine articulation

IMAGING

General Features

- Best diagnostic clue
  - Flattening or malformation of clivus/occipital condyles, anterior C1 ring, or odontoid process

Radiographic Findings

- Radiography
  - Platybasia, basilar invagination, C1 ring assimilation, asymmetrical C1/2 articulation, backward tilt, hypoplasia/aplasia, os odontoideum of odontoid process, C2/3 fusion (Klippel-Feil)

Fluoroscopic Findings

- Variable instability of C0/1, C1/2 articulations with dynamic flexion and extension

CT Findings

- CECT
  - ± enhancing granulation tissue mass ("pannus")
    - Patients with reducible abnormalities usually have largest pannus
- Bone CT
  - Findings similar to plain radiographs; sagittal and coronal reformats minimize effect of overlapping structures

MR Findings

- T1WI
  - Osseous anomalies ± soft tissue granulation tissue, cord compression, hindbrain anomalies
- T2WI
  - Similar to T1WI; best for evaluating spinal cord
- T1WI C+
  - ± enhancing "pannus"

Imaging Recommendations

- Protocol advice
  - Dynamic flex-extend plain films to demonstrate biomechanics, uncover instability
  - Multiplanar T1WI, T2WI to evaluate cord, soft tissues; flex-extend MR optimally delineates effect of position on neural compression
  - Bone CT with multiplanar reformats to evaluate osseous structures for surgical planning

DIFFERENTIAL DIAGNOSIS

Developmental CVJ Anomalies

- CVJ stenosis or atlantoaxial instability 2° to
  - Achondroplasia
  - Mucopolysaccharidosis
  - Down syndrome
  - Inborn errors of metabolism

Inflammatory and Degenerative Arthritides

- ± CVJ fusion, anterior displacement of C1 ring from dens, basilar invagination → possible cord compression 2° to canal stenosis
  - Rheumatoid arthritides
  - Reactive arthritides
  - Psoriatic arthritides
  - Ankylosing spondylitis
  - Degenerative arthritis (osteoarthritis)
- Clinical manifestations include myelopathy, pain, extremity deformity
  - Elicit characteristic clinical, historical, and laboratory abnormalities to confirm diagnosis

Acquired Basilar Impression

- Upward displacement of occipital condyles above plane of foramen magnum, radiological protrusion of dens tip above Chamberlain line
- 2° to bone softening
  - Paget disease
  - Osteogenesis imperfecta
  - Rickets
  - Rheumatoid arthritis
  - Hyperparathyroidism

Trauma

- CVJ injuries relatively uncommon but high morbidity/mortality
- Fracture &/or ligamentous injury
- Sharp, noncorticated margins argue against congenital anomaly

PATHOLOGY

General Features

- Etiology
  - Congenital insult to developing CVJ neural and osseous tissue between 4th and 7th intrauterine weeks → hypoplasia, segmentation/fusion anomalies, CVJ ankylosis
- Associated abnormalities
  - Dwarfism, jaw anomalies, cleft palate, congenital ear deformities, short neck, Sprengel deformity, funnel chest, pes cavus, and syndactyly
- Congenital CVJ abnormalities are relatively uncommon
  - Severity ranges from benign, asymptomatic to potentially fatal instability → cord/brainstem compression
- Type and severity of anomaly determined by anatomy relative to 1 or more standard "lines of reference"
  - Chamberlain line: Line drawn from opisthion to dorsal margin of hard palate
  - McGregor line: Line drawn from upper posterior hard palate to most caudad point of occiput

Staging, Grading, & Classification

- Occipital sclerotome malformations

- Most occiput anomalies associated with ↓ skull base height ± basilar invagination (odontoid tip > 4.5 mm above McGregor line)
  - Condylus tertius, condylar hypoplasia, basiocciput hypoplasia, atlantooccipital assimilation, bifid clivus
  - Platybasia = congenital flattening of craniocervical angle > 135°
  - Associated with hindbrain herniation, syringomyelia (≤ 30%)
- C1 ring anomalies
  - C1 assimilation ("occipitalized C1"): Segmentation failure → fibrous or osseous union between 1st spinal sclerotome and 4th occipital sclerotome
    - ± occipitocervical synostosis; most C1 ring assimilations asymptomatic, more likely to be symptomatic if retroodontoid AP canal diameter < 19 mm
  - C1 malformation: Aplasia, hypoplasia, cleft C1 arch, "split atlas" (anterior and posterior arch rachischisis)
  - Association with Klippel-Feil, basilar invagination, Chiari 1 malformation
- C2 anomalies: C1/2 segmentation failure, dens dysplasia
  - Majority confined to odontoid process; partial (hypoplasia) → complete absence (aplasia), ossiculum terminal persistens, os odontoideum
    - Ossiculum terminale persistens: Ossification failure of terminal ossicle → incidental "notch" in dens tip
    - Os odontoideum: Well-defined ossicle at dens tip + anterior C1 arch enlargement
  - Odontoid anomalies in association with ligamentous laxity → atlantoaxial instability
    - Incompetence of cruciate ligament → C1/2 instability, possible neurologic deficit, or death
    - Most common in Down syndrome, Morquio syndrome, Klippel-Feil syndrome, & skeletal dysplasias

Gross Pathologic & Surgical Features

- Solid ankylosis or fibrous union in many irreducible anomalies
- Granulation tissue proliferation around motion areas in unstable or reducible anomalies

Microscopic Features

- Variable components of histologically normal bone, fibrous tissue, & granulation tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Suboccipital neck pain (85%); may clinically mimic "basilar migraine"
  - Posterior occipital headache exacerbated by flexion/extension
- Other signs/symptoms
  - Myelopathy, brainstem/cranial nerve deficits, weakness, lower extremity ataxia
  - Vascular symptoms (15-20%); TIA, vertigo, visual symptoms with rotation or head manipulation
- Clinical profile
  - Usually normal clinical appearance; obvious clinical dysmorphism implies syndromal association
  - Symptomatic presentation following mild trauma common

Demographics

- Age
  - Infancy → late adulthood depending on severity
- Gender
  - M = F
- Epidemiology
  - Relatively uncommon: 0.14-0.25% pediatric population

Natural History & Prognosis

- Usually gradual onset of neurological symptoms with localizing signs; some patients asymptomatic throughout life
- Occasionally, neurological presentation is fulminant → quadriplegia, sudden death
- Undetected anomalies at risk for injury during minor trauma, anesthesia
- Early diagnosis permits treatment before symptoms or permanent neurological sequelae

Treatment

- Conservative approach initially unless unstable or neural deficits present
  - Traction, cervical orthosis, activity restriction
- Symptomatic, refractory to conservative management
  - Skeletal traction to distinguish reducible from irreducible abnormalities, relieve symptoms preoperatively
  - Correction of underlying biomechanical abnormality with decompression ± fusion

DIAGNOSTIC CHECKLIST

Consider

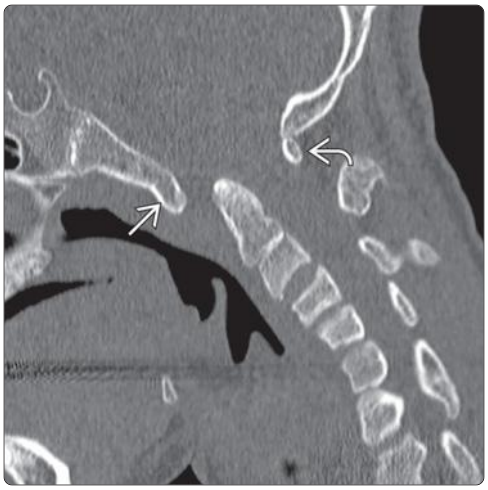
- Look for combinations of anomalies based on known association patterns
- Impact of diagnosis must be customized to individual patient to develop best treatment approach

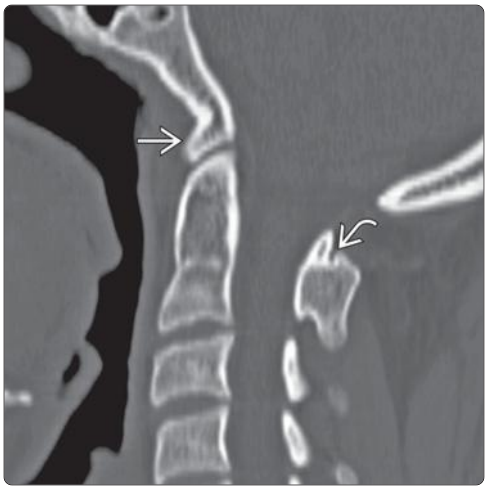
Image Interpretation Pearls

- Dynamic flex-extend imaging determines stability, reducibility of abnormality
- CT with reformats valuable for evaluating osseous abnormalities

SELECTED REFERENCES

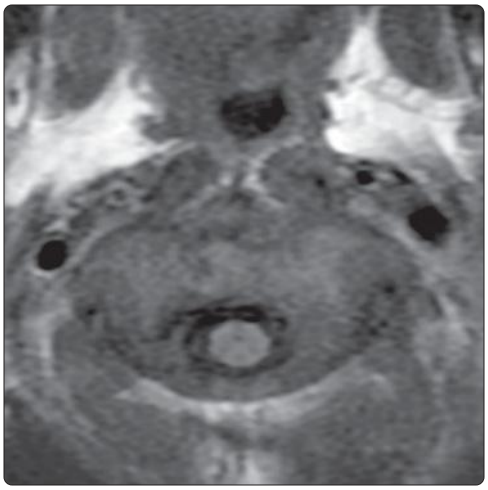
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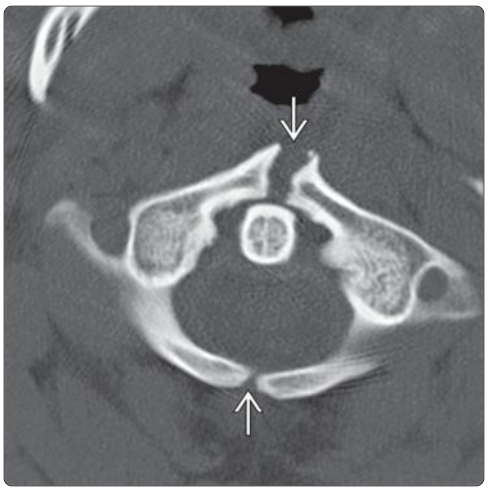


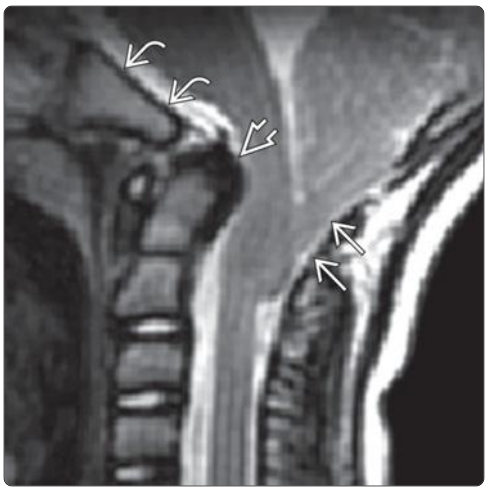
(Left) *Sagittal bone CT shows assimilation of anterior C1 ➡ with basion and partial assimilation of posterior C1 arch ➡ with opisthion. There is widened atlantodental interval and mild basilar invagination.* **(Right)** *Sagittal bone CT demonstrates congenital assimilation of anterior C1 ring ➡ with clivus as well as fusion of the posterior C1 ring to the C2 spinous process ➡. This CVJ anatomical variation would be expected to produce reduced CVJ dynamic motion.*





(Left) *Sagittal T2WI MR (VACTERL) shows moderately severe central canal stenosis at C1 ➡ secondary to C1 ring hypoplasia. There is mild T2 hyperintense signal in the cord at that level indicating myelomalacia. Also incidentally demonstrated is a rudimentary intervertebral disc space at C2-C3, reflecting congenital segmentation failure.* **(Right)** *Axial T1WI (VACTERL) MR confirms moderately severe central canal stenosis at C1 secondary to C1 ring hypoplasia.*





(Left) *Axial bone CT depicts congenital fusion failure of the C1 ring, with both anterior and posterior ring clefts ➡. Despite its bizarre appearance, this constellation of findings does not necessarily predispose to cervical instability.* **(Right)** *Sagittal T2WI MR reveals flattening of the clivus ➡ angle and aberrant odontoid ➡ retroflexion with adjacent hypointense pannus. In addition, MR imaging discloses a previously unsuspected Chiari 1 malformation with cerebellar tonsillar ectopia ➡.*

Ponticulus Posticus

KEY FACTS

TERMINOLOGY

- Latin: "Little posterior bridge"
- Synonyms: Arcuate foramen, sagittal foramen, canalis vertebralis, retroarticular canal, retroarticular/retrocondylar vertebral artery ring, upper retroarticular foramen, and atlantal posterior foramen

IMAGING

- Osseous roof along superior C1 arch covers C1 vertebral artery foramen
  - Vertebral artery passes through osseous tunnel
- May be partial or complete
- Unilateral or bilateral

TOP DIFFERENTIAL DIAGNOSES

- Broad C1 posterior arch

PATHOLOGY

- Postulated to arise from ossification of lateral segment of posterior atlantooccipital ligament or joint capsule
- Morphologically normal bone

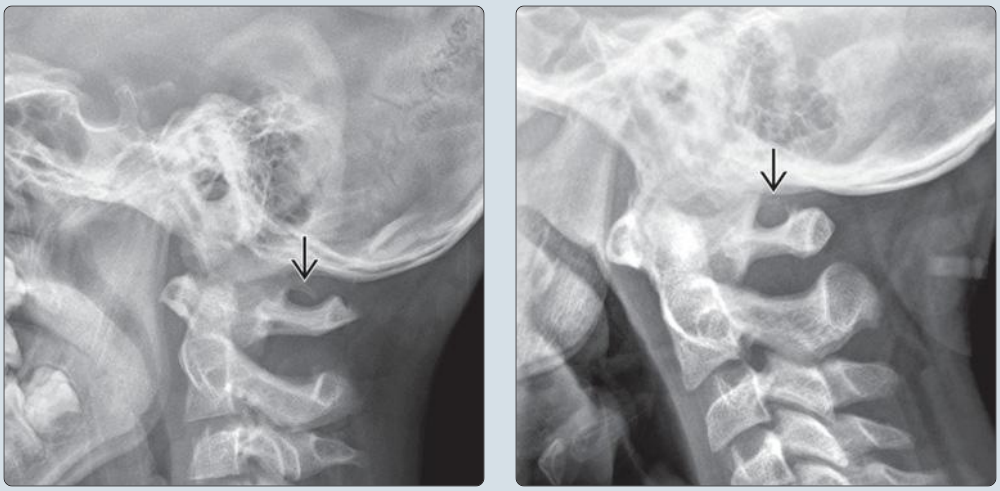
CLINICAL ISSUES

- Most common: Asymptomatic
- Other symptoms
  - Vertebrobasilar ischemia or infarction
  - Vertigo
  - Headache
  - Neck pain

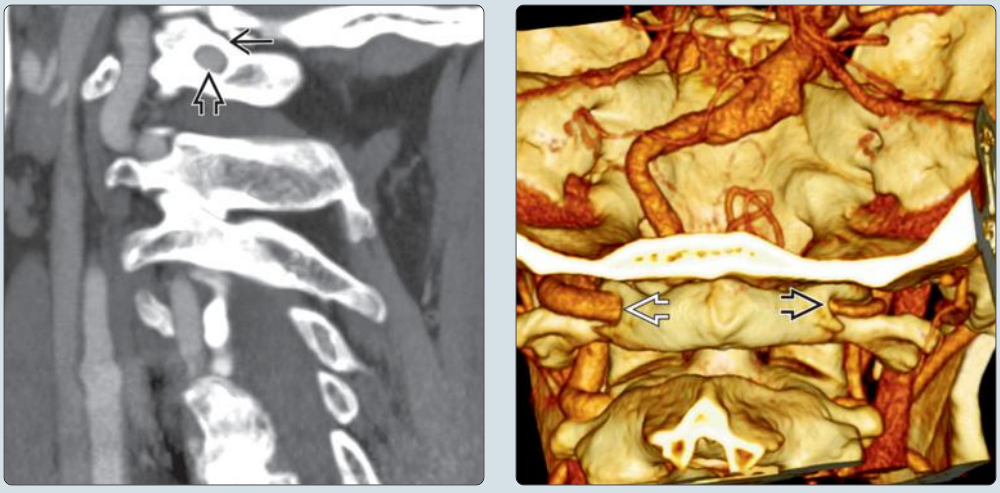
DIAGNOSTIC CHECKLIST

- Vertebral arteries predisposed to surgical injury during lateral mass screw placement
- Multiplanar bone CT with 3D reformats best demonstrates ponticulus posticus

(Left) *Lateral radiograph of the upper cervical spine demonstrates a partial osseous roof ⇨ over the C1 vertebral artery foramen, characteristic of ponticulus posticus (incomplete variant).* **(Right)** *Lateral radiography of the upper cervical spine reveals a complete osseous roof ⇨ over the C1 vertebral artery foramen, typical of classic ponticulus posticus (complete variant).*



(Left) *Sagittal CTA depicts a complete C1 arch, with a robust osseous covering (complete ponticulus posticus) ⇨ over the vertebral artery ⇨ along the superior aspect of the C1 arch.* **(Right)** *Anteroposterior view of a CTA 3D reformat shows a dominant left vertebral artery ⇨ and smaller (nondominant) right vertebral artery ⇨. Both are covered by a complete osseous bridge along the superior aspect of the C1 arch.*



TERMINOLOGY

Synonyms

- Arcuate foramen, sagittal foramen, canalis vertebralis, retroarticular canal, retroarticular/retrocondylar vertebral artery ring, upper retroarticular foramen, and atlantal posterior foramen

Definitions

- Osseous arch connecting retroglenoid tubercle with posterior C1 arch
- Latin: "Little posterior bridge"

IMAGING

General Features

- Best diagnostic clue
  - Osseous "bridge" along superior C1 arch covers vertebral artery
- Location
  - C1 arch vertebral foramen
  - Unilateral or bilateral
- Morphology
  - May be partial or complete

Radiographic Findings

- Radiography
  - Partial or complete osseous roof over C1 vertebral artery foramen

CT Findings

- Bone CT
  - Complete or incomplete osseous "bridge" covers C1 vertebral foramen
- CTA
  - Vertebral artery passes through osseous tunnel formed by ponticulus posticus

MR Findings

- Partial or complete osseous roof over C1 vertebral artery foramen
- MRA
  - Contains ipsilateral vertebral artery

Imaging Recommendations

- Best imaging tool
  - Multiplanar bone CT with 3D reformats

DIFFERENTIAL DIAGNOSIS

Broad C1 Posterior Arch

- Mistaking ponticulus posticus for broad posterior C1 arch → vertebral artery injury during C1 lateral mass screw placement

PATHOLOGY

General Features

- Etiology
  - Postulated to arise from ossification of lateral segment of posterior atlantooccipital ligament or joint capsule
- Associated abnormalities
  - None

Staging, Grading, & Classification

- Full: Complete osseous ring
- Incomplete: Portions of osseous ring absent
- Calcified: Linear or amorphous calcification

Gross Pathologic & Surgical Features

- Morphologically normal bone
- May mimic posterior C1 arch

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
- Other signs/symptoms
  - Vertebrobasilar ischemia or infarction
  - Vertigo
  - Headache
  - Neck pain
  - Shoulder/arm pain

Demographics

- Age
  - No association
- Gender
  - F > M
- Epidemiology
  - 3-38%

DIAGNOSTIC CHECKLIST

Consider

- Vertebral arteries predisposed to surgical injury during lateral mass screw placement
- Multiplanar bone CT with 3D reformats best demonstrates ponticulus posticus

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Ossiculum Terminale

KEY FACTS

TERMINOLOGY

- Synonym: Ossiculum terminale persistens
- Persistence of unfused odontoid tip ossification center into adulthood

IMAGING

- Separate ossicle with intact cortical margin positioned above normal-sized odontoid process in adolescent or adult
  - May be dystopic or orthotopic
- Soft tissue pannus suggests atlantoaxial instability
- Myelomalacia, brainstem compression in rare cases

TOP DIFFERENTIAL DIAGNOSES

- Os odontoideum
- Type I or II odontoid fracture
- Normal unfused odontoid tip synchondrosis
- Degenerative remodeling of odontoid process

PATHOLOGY

- Terminal ossicle normally appears by 3 years of age, fuses with odontoid body by 12 years of age
- Terminal ossicle located above transverse ligament
  - Atlantoaxial instability less common than with os odontoideum
  - Unstable ossiculum terminale usually dystopic

CLINICAL ISSUES

- Usually asymptomatic
- Uncommonly, neck pain or myelopathy related to CVJ instability

DIAGNOSTIC CHECKLIST

- Consider ossiculum terminale in patients > 12 years with persistent terminal ossicle at odontoid tip
- Evaluate for atlantoaxial instability if dystopic ossicle, excessive soft tissue pannus, or trisomy 21 patient

(Left) *Sagittal bone CT demonstrates a small ossiculum terminale ➡ located adjacent to the odontoid tip. The ossicle is in orthotopic position, and CVJ alignment is normal.* **(Right)** *Coronal bone CT confirms orthotopic placement of a small orthotopic ossiculum terminale ➡. Note that there is minimal flattening of the odontoid tip adjacent to the ossicle, supporting classification as a persistent terminal odontoid ossicle.*





(Left) *Sagittal bone CT reveals a small dystopic ossiculum terminale ➡ positioned between the odontoid tip and clivus. The odontoid tip has remodeled.* **(Right)** *Sagittal bone CT shows subluxation of the anterior C1 ring ➡ relative to the nearly normal-sized odontoid process ➡. There is a large dystopic ossiculum terminale ➡. Although debatable, the nearly normal size of the odontoid process favors classification as an ossiculum terminale rather than os odontoideum.*





TERMINOLOGY

Synonyms

- Ossiculum terminale persistens

Definitions

- Persistence of unfused odontoid tip ossification center into adulthood

IMAGING

General Features

- Best diagnostic clue
  - Separate ossicle with intact cortical margin positioned above normal-sized odontoid process in adolescent or adult
- Size
  - Variable (several mm to > 10 mm)
- Morphology
  - Rounded ossicle at odontoid tip

Radiographic Findings

- Radiography
  - Separate ossicle with intact cortical margin positioned above remainder of odontoid process
  - Flexion/extension radiographs may show ossicle movement relative to odontoid process if unstable

CT Findings

- Bone CT
  - Separate rounded ossicle at odontoid tip
    - Circumferential sclerotic cortical margins
    - May be dystopic or orthotopic

MR Findings

- Remnant of synchondrosis may persist at junction between ossiculum and odontoid proper
  - Cartilage may be visible between ossicle, odontoid
- Soft tissue pannus suggests atlantoaxial instability
- Myelomalacia, brainstem compression in rare cases

Imaging Recommendations

- Best imaging tool
  - Multiplanar bone CT

DIFFERENTIAL DIAGNOSIS

Os Odontoideum

- Rounded, corticated os margins
- Dystopic or orthotopic os position
- Odontoid process dysplastic, hypoplastic
- Atlantoaxial instability more common than with persistent ossiculum terminale

Type I or II Odontoid Fracture

- Inferior margin of odontoid fragment not corticated
- Soft tissue swelling, trauma clinical history helpful

Normal Unfused Odontoid Tip Synchondrosis

- Small terminal ossicle lying within "V" of odontoid tip
- Young patient (< 12 years)

Degenerative Remodeling of Odontoid Process

- Osteophytes, degenerative pannus
- Much more common, especially in adults, than persistent ossiculum terminale

PATHOLOGY

General Features

- Etiology
  - Atlas ossifies from 5 primary ossification centers
    - Odontoid process begins to ossify between 1st and 5th gestational months
  - Odontoid tip arises from a separate secondary ossification center
    - Derived from 4th occipital sclerotome
    - Terminal ossicle normally appears by 3 years of age fuses with odontoid body at around 12 years of age

Gross Pathologic & Surgical Features

- Terminal ossicle is located above transverse ligament
  - Ossiculum may be mobile if there is CVJ instability
  - Atlantoaxial instability less common than with os odontoideum

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
- Other signs/symptoms
  - Neck pain or myelopathy related to CVJ instability

Natural History & Prognosis

- Most patients asymptomatic
- True atlantoaxial instability requiring surgical intervention very rare
  - Unstable ossiculum terminale usually dystopic
  - Presents later in life or in association with trisomy 21

Treatment

- Usually none required
- CVJ instability may require surgical fusion

DIAGNOSTIC CHECKLIST

Consider

- Consider ossiculum terminale in patients older than 12 years with persistent terminal ossicle
- Evaluate for atlantoaxial instability if dystopic ossicle, excessive soft tissue pannus, or trisomy 21 patient

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KEY FACTS

TERMINOLOGY

- Conjoined nerve root (CNR), composite nerve root sleeve
- Asymmetric anomalous origin of enlarged nerve root sleeve containing 2 nerve roots

IMAGING

- Enlarged root sleeve containing 2 roots originating midway between expected positions of 2 contributing nerve roots
  - Roots usually exit separately at expected neural foraminal levels
  - Occasionally both roots exit through same (usually lower) foramen

TOP DIFFERENTIAL DIAGNOSES

- Nerve sheath tumor (NST)
- Intervertebral disc herniation
- Synovial cyst
- Perineural root sleeve (Tarlov) cyst

PATHOLOGY

- Composite root sleeve bigger than normal, originates from dural sac halfway between expected positions of 2 sleeves it replaces
- Most commonly unilateral

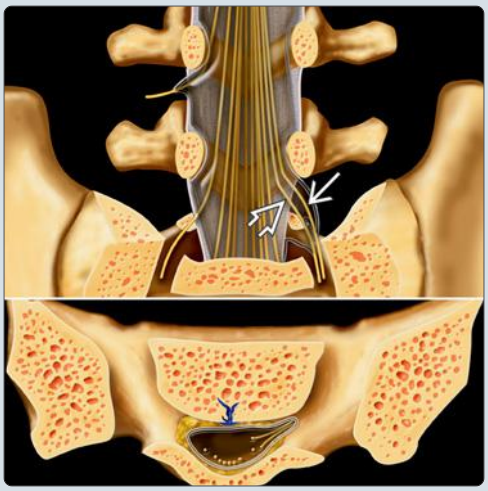
CLINICAL ISSUES

- Usually asymptomatic
- Symptomatic patients present with radiculopathy

DIAGNOSTIC CHECKLIST

- More commonly recognized at autopsy or during surgery than on imaging studies
- Important anatomical variant to identify prior to spinal surgery to prevent inadvertent nerve injury
- Exclude differential diagnoses, such as nerve sheath tumor, disc herniation, or synovial cyst

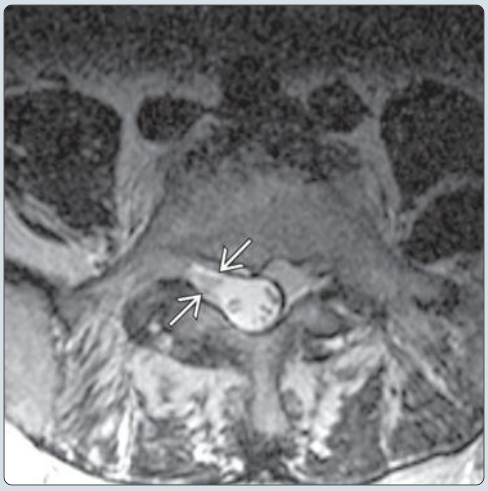
(Left) *Composite graphic shows variant nerve root anatomy, with 2 nerve roots (left L5 ⇨ and S1 ⇨) exiting the dural sac through a single composite dural sleeve and neural foramen (left S1 level) instead of the more common variant with each nerve exiting at its named level.* **(Right)** *Anteroposterior myelography demonstrates the typical conjoined root configuration, with an aberrant enlarged root sleeve ⇨ containing 2 roots (L5, S1) that will each exit at their named level.*





(Left) *Sagittal T1WI MR (conjoined L5 ⇨ and S1 ⇨ nerve roots) demonstrates exit of the nerve roots independently through their appropriately named neural foramina. This is the most common variation of conjoined nerve root anatomy.* **(Right)** *Axial T2WI MR (conjoined right L5, S1 nerve roots) demonstrates typical conjoined nerve root anatomy, with an aberrant large right nerve root sleeve ⇨ arising rostral to expected level containing 2 different nerve roots.*





TERMINOLOGY

Abbreviations

- Conjoined nerve root (CNR)

Synonyms

- Composite nerve root sleeve

Definitions

- Asymmetric anomalous origin of enlarged nerve root sleeve containing 2 nerve roots

IMAGING

General Features

- Best diagnostic clue
  - Enlarged root sleeve containing 2 roots originating midway between expected positions of 2 contributing nerve roots
- Location
  - Lumbar spine > > cervical, thoracic spine
    - L4/5 > L5/S1 > L3/4
- Size
  - Conjoined root sleeve larger than normal root sleeve
  - Nerve roots normal size (unless inflamed)
- Morphology
  - Asymmetric anomalous origin of enlarged nerve root sleeve
    - Roots usually exit separately at expected neural foraminal levels
    - Occasionally both roots exit through same (usually lower) foramen
    - → CSF-filled foramen without dorsal root ganglion (DRG) at 1 level, 2 enhancing DRGs within other foramen

Radiographic Findings

- Radiography
  - Enlarged neural foramen
  - Ipsilateral hypoplastic or absent pedicle

CT Findings

- CECT
  - Enlarged neural foramen, ± hypoplastic or absent pedicle
  - Asymmetric mild dilatation of ipsilateral osseous lateral recess
    - Not typically observed with extruded free intervertebral disc fragments
  - DRG enhancement

MR Findings

- T1WI
  - Enlarged neural foramen, ± hypoplastic or absent pedicle
  - Nerve roots easily visualized in foramen surrounded by hyperintense fat
- T2WI
  - Similar osseous findings to T1WI MR
  - Supranumerary nerve roots identified within asymmetric enlarged root sleeve
- T1WI C+
  - Normal DRG enhancement
  - Remainder of nerve is nonenhancing (postoperatively, nerves may enhance)

Ultrasonographic Findings

- Grayscale ultrasound
  - 2 nerve roots within single root sleeve

Nonvascular Interventions

- Myelography
  - Supernumerary nerve roots on CT myelography
    - 2 nerve roots exit through single contrast-filled sleeve midway between sleeve exit levels of contralateral side
    - Roots either exit separately through appropriate foramina or remain joined and exit through a single foramen
  - Most cases demonstrated equally well using MR imaging
    - Reserve myelography for problem solving, patients with MR imaging contraindications

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal and axial MR imaging
    - Both T1WI and T2WI MR needed
      - Sagittal slice prescription must extend lateral to neural foramina
      - Contrast useful to locate DRG
  - CT myelography demonstrates relationship of root sleeve to osseous structures well
    - Use for inconclusive MR imaging or when MR contraindications exist

DIFFERENTIAL DIAGNOSIS

Nerve Sheath Tumor (NST)

- Conjoined nerve root mimics appearance of schwannoma or neurofibroma
- Larger tumors show dumbbell configuration
- Entire NST usually enhances avidly; only DRG of conjoined nerve root normally enhances

Disc Herniation

- Rim enhancement of disc fragment, contiguity with disc space
- Neural sleeves exit dural sac at appropriate levels

Synovial Cyst

- Severe degenerative facet disease
- Neural sleeves exit dural sac at appropriate levels

Perineural Root Sleeve Cyst

- Nerve root sleeve cyst follows nerve course into foramen
- Cyst usually displaces nerve DRG peripherally

PATHOLOGY

General Features

- Etiology
  - Unknown
- Associated abnormalities
  - Occasionally developmental lumbar spinal stenosis

- More commonly recognized at autopsy or during surgery than on imaging studies
- May produce sciatica without additional compressive impingement from disc herniation, spondylolisthesis, or lateral recess stenosis
- Symptomatic (but undiagnosed) conjoined nerve roots are 1 described cause of failed back syndrome

Gross Pathologic & Surgical Features

- Most commonly unilateral
- Composite root sleeve originates from dural sac at point halfway between expected positions of the 2 sleeves it replaces, and is bigger than normal sleeve
  - Conjoined nerve roots commonly leave spinal canal through separate neural foramina
  - Less commonly, 2 roots exit through single foramina (usually that of lower root)
  - Failure to correctly appreciate this anatomical variant → misdiagnosis as nerve root tumor or operation at wrong level for clinical radiculopathy

Microscopic Features

- Normal nerve, dural histology

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
  - Symptomatic patients present with radiculopathy
    - Conjoined nerves prone to compression by discs; lateral bony stenosis along lateral recess
    - Herniated disc beneath bifid root → pain, disability, signs of entrapment 2° to fixation of conjoined root within lateral recess, between pedicles
- Clinical profile
  - Patient presents with cervical or lumbar radiculopathy

Demographics

- Age
  - No age predilection
- Gender
  - M = F
- Epidemiology
  - Prevalence 0.3-2%; some studies as high as 10.6%

Natural History & Prognosis

- Asymptomatic patients require no treatment
- Rare symptomatic patients referable to conjoined root level may require surgical therapy
- Symptomatic undiagnosed conjoined roots are 1 cause of failed back syndrome
  - Nerve root anomalies should be searched for in cases of failed disc surgery

Treatment

- Conservative nonoperative observation unless symptomatic
- Surgical decompression in selected patients with documented concordant radiculopathy
  - Hemilaminectomy with foraminotomy, pediculectomy, and discectomy best alleviate symptomatology

DIAGNOSTIC CHECKLIST

Consider

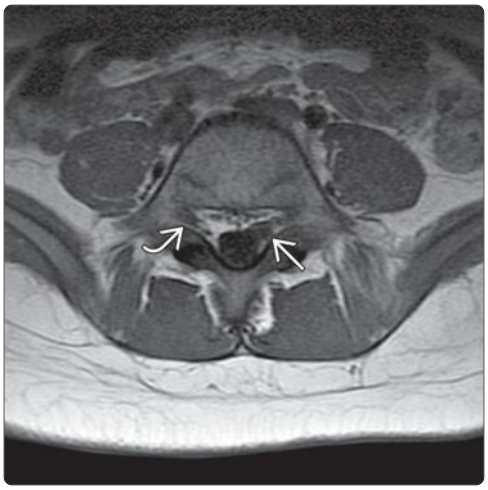
- Important anatomical variant to identify prior to spinal surgery to prevent inadvertent nerve injury
- Exclude differential diagnoses, such as nerve sheath tumor, disc herniation, or synovial cyst

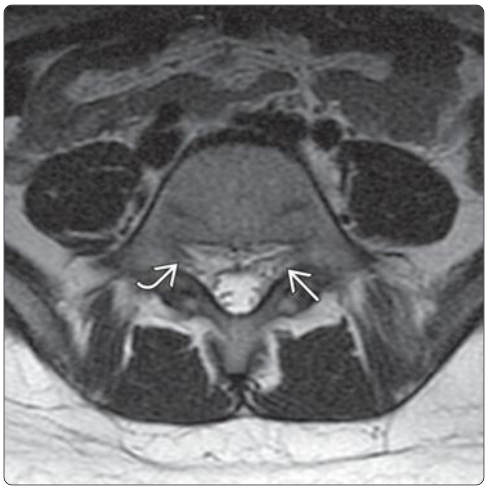
Image Interpretation Pearls

- Dorsal root ganglia contrast enhancement and enlargement of foraminal neural structures may lead to misdiagnosis as nerve sheath tumor

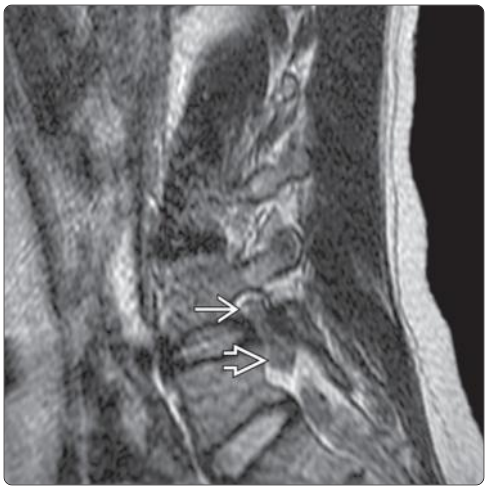
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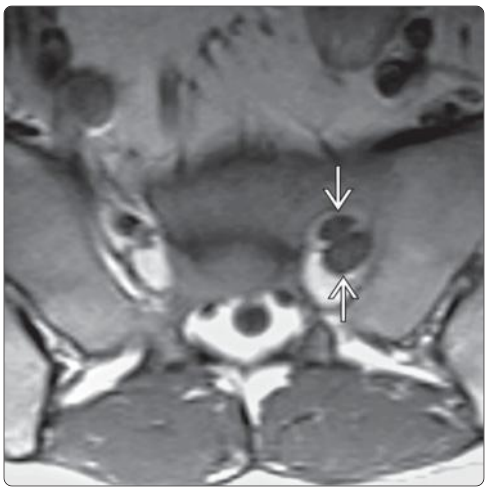
(Left) *Axial T1WI MR demonstrates exit of both L5 and S1 nerve roots from the left dural sac through a single nerve root sleeve. Conversely, the right L5 root exits conventionally from the lateral recess toward its named L5 neural foramen.* **(Right)** *Axial T2WI MR reveals the conjoined left L5 and S1 nerve roots exiting the dural sac through a single enlarged, aberrant nerve root sleeve. In contrast, the right L5 nerve root exits from the lateral recess toward the L5 neural foramen.*





(Left) *T2WI right parasagittal MR depicts the right L4 and L5 nerve roots exiting through a common aberrant root sleeve and subsequently diverging to transit and exit at their appropriately named foraminal levels.* **(Right)** *Axial T1WI C+ MR reveals that the conjoined right L4 and L5 nerve roots exit through a common aberrantly large root sleeve, and then transit towards their appropriately named neural foramina.*





(Left) *Sagittal T1 C+ FS MR shows normally enhancing dorsal root ganglia at all levels except left L5/S1. Conversely, 2 enhancing DRGs are seen within the S1 foramen representing DRGs for both L5 and S1 within the S1 neural foramina, an uncommon conjoined root variation.* **(Right)** *Axial T1WI MR of conjoined left L5 and S1 nerve roots reveals both DRGs within the left S1 neural foramen. No DRG was identified in the left L5 neural foramen (not shown).*

Limbus Vertebra

KEY FACTS

TERMINOLOGY

- Limbus vertebra (LV), rim apophysis (RA)
- Variation of vertebral cartilaginous node formation

IMAGING

- Children: Lucent defect in anterosuperior corner of vertebral body
- Adults: Triangular corticated bone fragment
  - LV roughly matches size of corresponding donor site defect in adjacent vertebral body
- Mid-lumbar > midcervical, anterior > > posterior

TOP DIFFERENTIAL DIAGNOSES

- Acute vertebral fracture
- Schmorl node
- Anterior osteophyte fragment
- Calcified disc herniation

PATHOLOGY

- RA separated from vertebral body by thin cartilage layer until apophysis fuses to vertebral body (age 18-20 years)
  - Relative weak point in disc/vertebral body until fusion complete
- LV occurs when nucleus pulposus herniates between RA and vertebral body before osseous fusion, permanently isolating small segment of vertebral rim

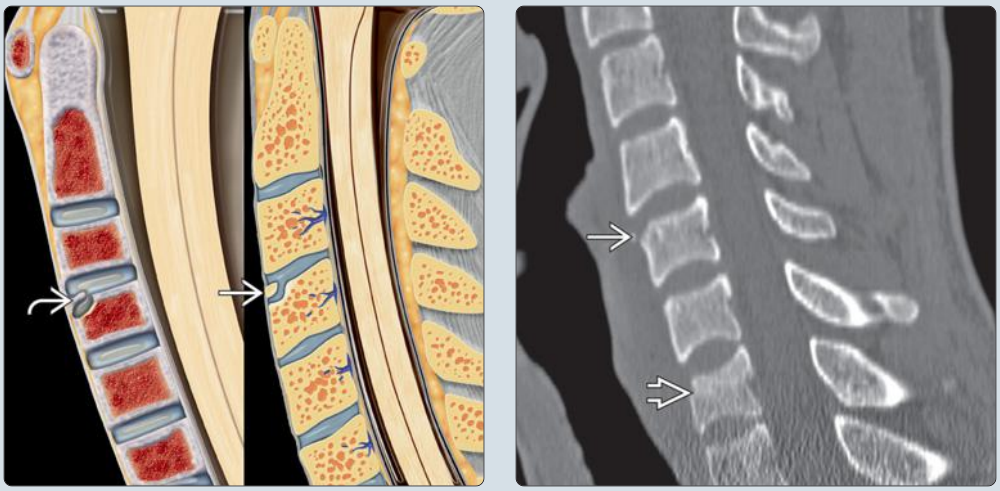
CLINICAL ISSUES

- Usually detected during imaging for chronic back pain
- Occasionally incidental finding
- Inciting event rarely recognized or remembered

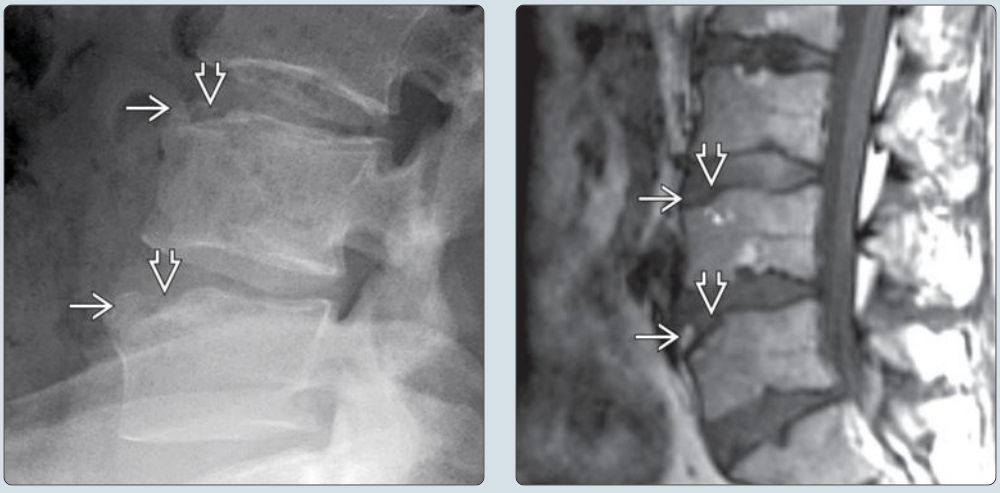
DIAGNOSTIC CHECKLIST

- LV fragment size roughly matches vertebral defect

(Left) *Sagittal graphic demonstrates age-related appearance of cervical limbus vertebra. The disc herniation underneath the nonossified apophysis in a pediatric patient (left) mimics a vertebral "defect," while subsequent ossification (right) produces a typical adult LV appearance.* **(Right)** *Sagittal bone CT depicts a typical C5 LV. Additionally, anterior C7 wedging, with anterior defect and dense trabecular compression, is compatible with clinical acute fracture.*



(Left) *Lateral radiography demonstrates limbus fragments at both L4 and L5. Additionally, at both levels, the "divot" in the anterosuperior vertebral body is larger than the limbus fragment because of concurrent Schmorl nodes behind the corticated limbus fragments.* **(Right)** *Sagittal T1WI MR confirms LV at both L4 and L5. In addition, there is the anatomic variation of concurrent typical Schmorl nodes located posterior to the typical osseus defect of LV.*



TERMINOLOGY

Abbreviations

- Limbus vertebra (LV), rim apophysis (RA), nucleus pulposus (NP)

Synonyms

- "Slipped vertebral apophysis"

Definitions

- Variation of vertebral cartilaginous node formation
  - Intraosseous disc penetration at junction of cartilaginous endplate, developing rim apophysis

IMAGING

General Features

- Best diagnostic clue
  - Small corticated bone fragment matching osseous defect at anterosuperior vertebral margin
- Location
  - Midlumbar > midcervical
  - Anterior > > posterior
- Size
  - Small (~ 1-3 mm)
- Morphology
  - Small unfused apophyseal osseous fragment at vertebral margin
    - Lumbar LV: Anterosuperior > > posterior
    - Cervical LV: Anteroinferior > anterosuperior

Radiographic Findings

- Radiography
  - Children: Lucent defect in anterosuperior corner of vertebral body
    - Fragment may not be visible (nonossified)
  - Adults: Triangular corticated bone fragment
    - Roughly matches corresponding donor site defect in adjacent vertebral body

CT Findings

- Bone CT
  - Osseous vertebral defect roughly matching corticated limbus fragment
    - Fragment and donor site margins usually sclerotic when subacute/chronic
    - Limbus fragment may not be visible (nonossified) in children
  - ± degenerative spine manifestations
    - Mild kyphosis
    - Variable disc height loss

MR Findings

- T1WI
  - Hypointense bone fragment often blends in with anterior spinal ligaments
    - Limbus fragment subtle but may be detectable on careful inspection of sagittal images
    - ± mild kyphosis
    - ± disc height loss
  - Mild hypointense marrow edema (if acute) in adjacent vertebral body
- T2WI
  - Hypointense bone fragment usually blends in with anterior spinal ligaments
  - T2 hyperintense disc extends between fragment and vertebral body
    - Caveat: Disc protrusion may be hypointense in adults with degenerative disc dehydration but usually resembles intrinsic signal intensity within remainder of intervertebral disc
- STIR
  - Mild hyperintense marrow edema (if acute) in adjacent vertebral body
  - Other findings similar to T2WI

Nonvascular Interventions

- Discography: Intradiscal contrast flows between fragment and vertebral body, confirming diagnosis

Imaging Recommendations

- Best imaging tool
  - Lateral plain radiography usually diagnostic
- Protocol advice
  - Plain radiographs establish diagnosis
  - MR helpful (if necessary) to assess for edema or inflammation in context of acute symptoms

DIFFERENTIAL DIAGNOSIS

Acute Vertebral Fracture

- LV commonly misdiagnosed as acute vertebral fracture
- Look for trabecular smudging, soft tissue swelling around fragment in acute fracture

Schmorl Node

- Schmorl node herniates more centrally into vertebral endplate compared to more peripheral location of LV

Anterior Osteophyte Fragment

- Degenerative osteophyte fractures off, mimicking LV
- No matching donor site on adjacent vertebral body

Calcified Disc Herniation

- Location usually not characteristic of LV
- No matching vertebral body defect

PATHOLOGY

General Features

- Etiology
  - Superior and inferior surfaces of developing vertebral body covered by thin, peripherally thickened cartilage plate → cartilaginous marginal ridge
  - Endochondral ossification of marginal ridge begins at age 7-9 years → forms RA
  - Outermost fibers of anulus fibrosus (Sharpey fibers) embedded into RA fasten disc to vertebral column
  - RA separated from vertebral body by this thin cartilage layer until apophysis fuses to vertebral body (at age 18-20 years)
    - Relative weak point in disc/vertebral body until fusion complete

  - LV occurs when nucleus pulposus herniates between RA and vertebral body before osseous fusion, permanently isolating small segment of vertebral rim
- Associated abnormalities
  - Scheuermann disease
  - Schmorl nodes
- Caused by herniation of nucleus pulposus beneath ring apophysis (junction of cartilaginous endplate, bony rim)
  - Occurs prior to fusion of apophysis to vertebral body
  - Apophysis remains separate from vertebral body
- Anterosuperior vertebral margin may be more frequently affected because of adjacent vertebral body size difference (superior < inferior)
  - During flexion, anterior disc forced down into superior endplate of larger inferior vertebra
  - Pushes nucleus pulposus through cartilage plate, isolating apophysis from main vertebral body

Gross Pathologic & Surgical Features

- Small corticated bone fragment, separated from vertebral body by intervening disc material

Microscopic Features

- Cancellous bone, hyaline cartilage, and acellular hyaline tissue (disc)
- Basophilic degeneration, hemorrhagic foci in hyaline cartilage common

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic back pain
  - Occasionally incidental finding
- Clinical profile
  - Variable symptomatology depending on location
    - Anterior defect → symptoms usually minor; stiffness or loss of physiologic lordosis
    - Posterior defect → symptoms mimic nerve root (lumbar) or spinal cord compression (cervical)
  - Inciting event rarely recognized or remembered
  - Physical exam shows decreased range of motion, ± kyphosis, or pain on spinous process palpation

Demographics

- Age
  - Adults > children, adolescents
- Gender
  - M > F
- Epidemiology
  - Common radiologic finding in adults; relatively less common in children
  - Trauma may be responsible for many cases
    - LV more common in active, athletic children; traumatic insult may not be remembered

Natural History & Prognosis

- No further investigative procedures necessary following diagnosis
  - Symptoms usually resolve over months to years
- LV may evolve into anterior Schmorl node in some patients

Treatment

- Conservative management
  - Analgesics
  - Physical therapy
  - Limitation of physical activity during acute phase
- Surgical management not indicated

DIAGNOSTIC CHECKLIST

Consider

- LV is sequel of remote injury in immature skeleton
- Important to exclude other more important causes of back pain in patient with LV

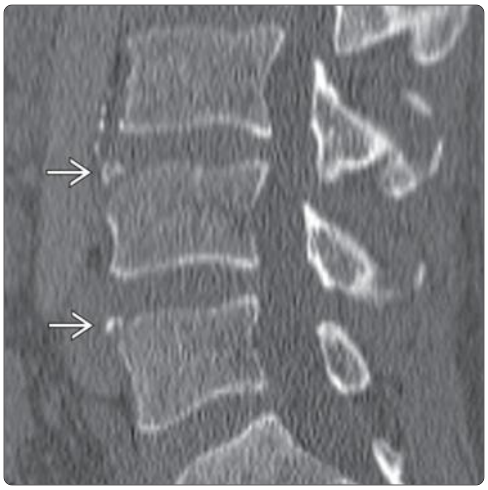
Image Interpretation Pearls

- Diagnosis of LV less difficult in adults than children
  - Detection of characteristic, well-formed triangular osseous fragment
- LV fragment roughly matches size of vertebral body defect
  - Helps distinguish LV from other differential considerations, such as calcified disc or fractured vertebral osteophyte

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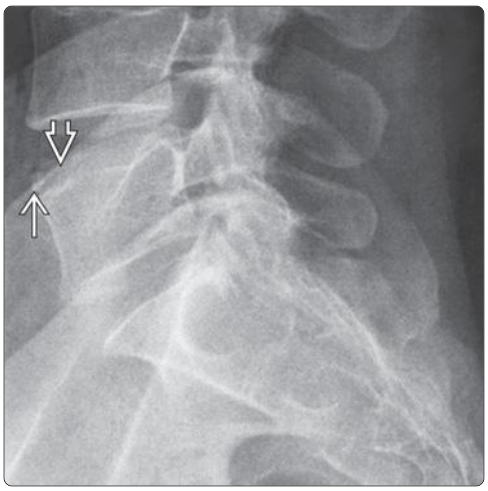
(Left) *Lateral radiograph depicts the typical appearance of a large, well-corticated L5 limbus fragment* ➡. **(Right)** *Sagittal bone CT demonstrates well-defined bony fragments* ➡ *along the superior anterior vertebral body margins of L4 and L5, a typical appearance for ununited ring apophysis (limbus vertebra). The anterosuperior corner of a single vertebral body is the most common presentation. Inferior and posterior margins are less frequently identified.*





(Left) *Lateral plain radiograph demonstrates a classic-appearing limbus vertebra* ➡ *at L3. The triangular-shaped limbus fragment nicely matches the osseous defect in the anterosuperior L3 vertebral body.* **(Right)** *Sagittal bone CT reveals typical-appearing limbus vertebra fragments* ➡ *at both L4 and L5, a less common presentation than identification of a single level LV. Note that the limbus fragments roughly match the bone defect in the anterosuperior L4 and L5 vertebral bodies.*





(Left) *Lateral plain radiograph shows a small, well-corticated limbus fragment* ➡ *that neatly fits into the "divot" in the anterosuperior L5 vertebral body.* **(Right)** *Variant case shows a limbus vertebra in conjunction with anterior Schmorl node. Lateral radiograph demonstrates a small, well-corticated limbus fragment* ➡, *smaller than the adjacent vertebral defect, created by superimposed limbus vertebra in addition to a large Schmorl node behind the limbus fragment* ➡.

Filum Terminale Fibrolipoma

KEY FACTS

TERMINOLOGY

- Synonyms: Fibrolipoma of filum terminale, fatty filum terminale, "fat in filum"
- Asymptomatic presence of fat within otherwise normal-appearing filum terminale

IMAGING

- Linear fat signal/density within filum terminale
- Conus termination at normal position
- No additional tethered cord imaging stigmata

TOP DIFFERENTIAL DIAGNOSES

- Intraspinal lipoma
- Tethered spinal cord
- Lipomyelocele
- Epidermoid/dermoid cyst
- Subarachnoid hemorrhage
- Tumor with paramagnetic effects

PATHOLOGY

- Small focus of fat detected within otherwise normal filum terminale
  - Typical adipose tissue on gross pathology, microscopy
  - Incidentally detected in 4-6% of autopsy subjects
- No cutaneous stigmata on lower back with asymptomatic filum fibrolipoma

CLINICAL ISSUES

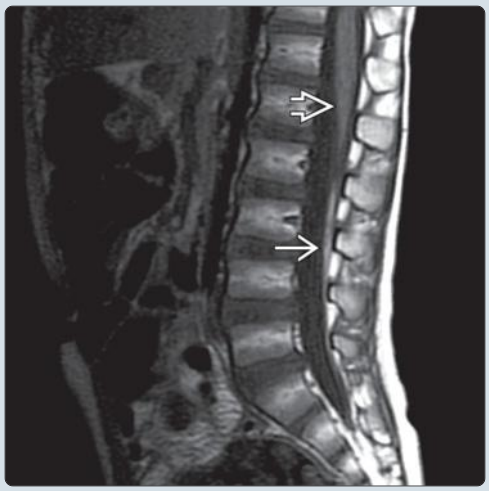
- Asymptomatic, incidental finding by definition
- Incidental finding in 4-6% of autopsy subjects

DIAGNOSTIC CHECKLIST

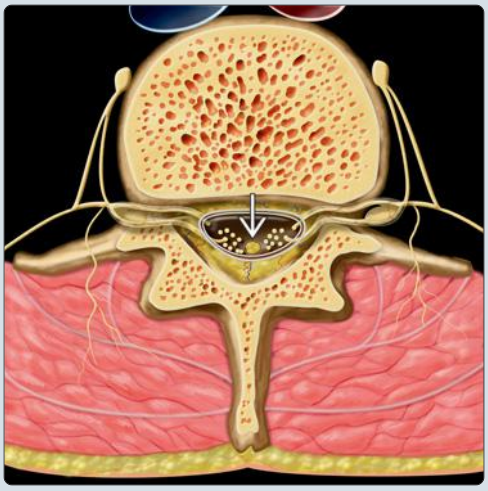
- Symptomatic patient implies diagnosis of intraspinal lipoma, not asymptomatic fibrolipoma
- Consider intraspinal lipoma if larger lesion, symptomatic, filum > 2 mm in thickness

(Left) *Sagittal graphic of the lumbosacral spine demonstrates extensive fatty infiltration ⇨ of the filum terminale. The spinal cord conus ⇨ terminates at the normal L1-L2 level.* **(Right)** *Sagittal T1WI MR depicts fatty infiltration of the filum terminale ⇨. The unremarkable conus ⇨ terminates at normal L1-L2 level. In this case, the filum fibrolipoma was thought to be incidental rather than causal due to the patient presenting with gait abnormality.*





(Left) *Axial graphic of the lumbar spine shows the characteristic appearance of the fatty filum ⇨ within the thecal sac. As depicted in this graphic, the fatty filum is dorsally located within the dural sac and produces no mass effect on the cauda equina.* **(Right)** *Axial T1WI MR confirms fatty infiltration ⇨ of the filum terminale. The conus terminated at the normal L1/2 level (not shown). In general, a fatty filum is most easily detected on axial T1WI MR.*





TERMINOLOGY

Synonyms

- Fibrolipoma of filum terminale, fatty filum terminale, "fat in filum"

Definitions

- Asymptomatic presence of fat within otherwise normal-appearing filum terminale
- Conus terminates at normal level without imaging stigmata of tethered spinal cord

IMAGING

General Features

- Best diagnostic clue
  - Linear fat signal/density within filum terminale
- Location
  - Filum terminale (conus level to sacrum)
- Size
  - Variable length, 1-5 mm transverse
    - Label as "lipoma" if > 2 mm transverse

CT Findings

- NECT
  - Punctate fat attenuation in dorsal aspect of lumbar thecal sac
  - No associated spinal dysraphism
- CECT
  - No enhancement

MR Findings

- T1WI
  - Linear high signal oriented in craniocaudal direction on sagittal images
    - May occur anywhere from below conus to sacrum
  - Located in dorsal 1/2 of thecal sac in position of filum terminale
  - Signal loss on fat-suppressed sequences
- T2WI
  - Tracks fat signal
- T1WI C+
  - No enhancement

Imaging Recommendations

- Best imaging tool
  - T1WI MR shows typical fat appearance, normal conus position, and morphology
- Protocol advice
  - Fat-suppressed T1WI to define hyperintensity

DIFFERENTIAL DIAGNOSIS

Intraspinal Lipoma

- Larger lipomatous mass (> 5 mm)
- Thickening of filum terminale (> 2 mm)
- Low-lying or indistinct conus

Tethered Cord

- Thick filum, indistinct conus termination, with smooth transition to filum

Lipomyelocele

- Dorsal dysraphism

Epidermoid/Dermoid Cyst

- Complex intradural mass, variable fat suppression

Subarachnoid Hemorrhage

- Fluid-fluid level in thecal sac

Tumor With Paramagnetic Effects

- Melanoma, melanotic meningioma, or schwannoma may show high signal on T1WI

PATHOLOGY

General Features

- Etiology
  - Congenital
- Associated abnormalities
  - No cutaneous stigmata on lower back with asymptomatic filum fibrolipoma
- Fatty infiltration of filum

Gross Pathologic & Surgical Features

- Small focus of fat within otherwise normal filum terminale

Microscopic Features

- Typical adipose tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic by definition, incidental finding

Demographics

- Age
  - Children and adults
- Gender
  - M = F
- Epidemiology
  - Incidental finding in 4-6% of autopsy subjects

Natural History & Prognosis

- Normal variant

Treatment

- None required for asymptomatic lesion

DIAGNOSTIC CHECKLIST

Consider

- Symptomatic patient implies diagnosis of intraspinal lipoma, not asymptomatic fibrolipoma
- Intraspinal lipoma if larger lesion, symptomatic, and filum > 2 mm in thickness

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2. Thompson EM et al: Clinical significance of imaging and histological characteristics of filum terminale in tethered cord syndrome. J Neurosurg Pediatr. 13(3):255-9, 2014

Bone Island

KEY FACTS

TERMINOLOGY

- Synonyms: Enostosis, sclerotic island, calcified island, compact island

IMAGING

- Single or multiple small focal areas of sclerosis in cancellous bone
- Distinctive radiating bony streaks ("thorny radiation") blend with trabeculae of host bone
- Focal low signal intensity on all MR imaging sequences

TOP DIFFERENTIAL DIAGNOSES

- Metastatic disease
- Schmorl node
- Degenerative endplates
- Osteoid osteoma
- Osteopoikilosis
- Bone infarct

PATHOLOGY

- Developmental or congenital failure of bone resorption during endochondral ossification
- Compact bone connected to adjacent bony trabeculae without cartilage or fibrous component

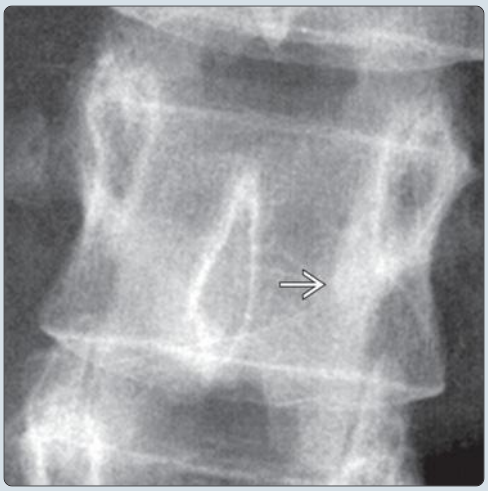
CLINICAL ISSUES

- Asymptomatic incidental finding in patient imaged for other reasons
- Stable lesions; rarely change size over time
- No treatment necessary

DIAGNOSTIC CHECKLIST

- Characteristic density, margins, absence of destruction or soft tissue mass reassuring for enostosis

(Left) *Thoracolumbar anteroposterior radiograph demonstrates sclerotic focus ➡ within the lower thoracic vertebral body, adjacent to the pedicle. There is characteristic nonaggressive appearance with sclerotic density and brush-like margins that suggest bone island.* **(Right)** *Axial bone CT of the thoracic spine reveals a focal sclerotic bone lesion ➡ with characteristic dense sclerosis and irregular, brush-like margins of the vertebral body bone island.*





(Left) *Sagittal T1WI MR demonstrates a small focal area of very low signal intensity within the anterior L4 body ➡, without reactive marrow change or osseous destructive changes to imply an aggressive lesion.* **(Right)** *Sagittal T2WI MR displays a small focal area of very low signal within the anterior L4 body ➡, without reactive marrow change or other worrisome marrow abnormality. Lack of destructive change and soft tissue mass favor bone island.*





TERMINOLOGY

Synonyms

- Enostosis, sclerotic island, calcified island, compact island

Definitions

- Asymptomatic focal areas of bony sclerosis

IMAGING

General Features

- Best diagnostic clue
  - Small focal areas of sclerosis, with feathered or brush-like margins, in cancellous bone
- Location
  - Found in any skeletal bone
    - Most common in pelvis, femur, ribs, long bones
    - Less common in spine; may involve vertebral body or posterior elements
- Size
  - 1 mm to several cm
- Morphology
  - Round, well-defined margins

Radiographic Findings

- Radiography
  - Single or multiple areas of focal homogeneously dense sclerotic foci in cancellous bone
  - Distinctive radiating bony streaks ("thorny radiation") blend with trabeculae of host bone

CT Findings

- NECT
  - Findings similar to plain radiography, without demonstrable soft tissue component

MR Findings

- T1WI
  - Focal low signal intensity
- T2WI
  - Focal low signal intensity
- T1WI C+
  - No enhancement

Nuclear Medicine Findings

- Bone scan
  - Usually normal but may show ↑ uptake, particularly in larger lesions

Imaging Recommendations

- Best imaging tool
  - Evaluation of solitary indeterminate lesion: CT
  - Evaluation of multiple lesions: MR

DIFFERENTIAL DIAGNOSIS

Metastatic Disease

- Multiple lesions, epidural extension

Schmorl Node

- Irregular undulations of vertebral endplate 2° to intervertebral disc herniation

Degenerative Endplates

- Adjacent to endplate, ± enhancement

Osteoid Osteoma

- Bone expansion, central nidus

Osteopoikilosis

- Multiple bone islands

Bone Infarct

- More ill-defined or patchy signal abnormality, ± enhancement

PATHOLOGY

General Features

- Etiology
  - Developmental or congenital failure of bone resorption during endochondral ossification

Gross Pathologic & Surgical Features

- Dense osseous lesion

Microscopic Features

- Compact bone connected to adjacent bony trabeculae
- No cartilage or fibrous component

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
- Clinical profile
  - Incidental finding in patient imaged for other reasons

Demographics

- Age
  - All age groups
- Gender
  - M = F
- Epidemiology
  - Spine involvement in up to 14% of patients

Natural History & Prognosis

- Stable lesions that rarely change size over time

Treatment

- None necessary

DIAGNOSTIC CHECKLIST

Consider

- Blastic metastatic disease (prostate or breast) with multiple small foci of low signal on MR

Image Interpretation Pearls

- Characteristic density, margins, and absence of destruction or soft tissue mass reassuring for enostosis

SELECTED REFERENCES

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2. Flemming DJ et al: Primary tumors of the spine. Semin Musculoskelet Radiol. 4(3):299-320, 2000

Ventriculus Terminalis

KEY FACTS

TERMINOLOGY

- Synonyms: Terminal ventricle, 5th ventricle

IMAGING

- Mild cystic dilatation of distal spinal cord canal
- No gliosis or myelomalacia
- No marginal nodularity or pathological enhancement
- Conus terminates at normal level (T12 → L2)

TOP DIFFERENTIAL DIAGNOSES

- Transient dilatation of central spinal cord canal
- Hydrosyringomyelia
- Cystic spinal cord neoplasm
- Myelomalacia

PATHOLOGY

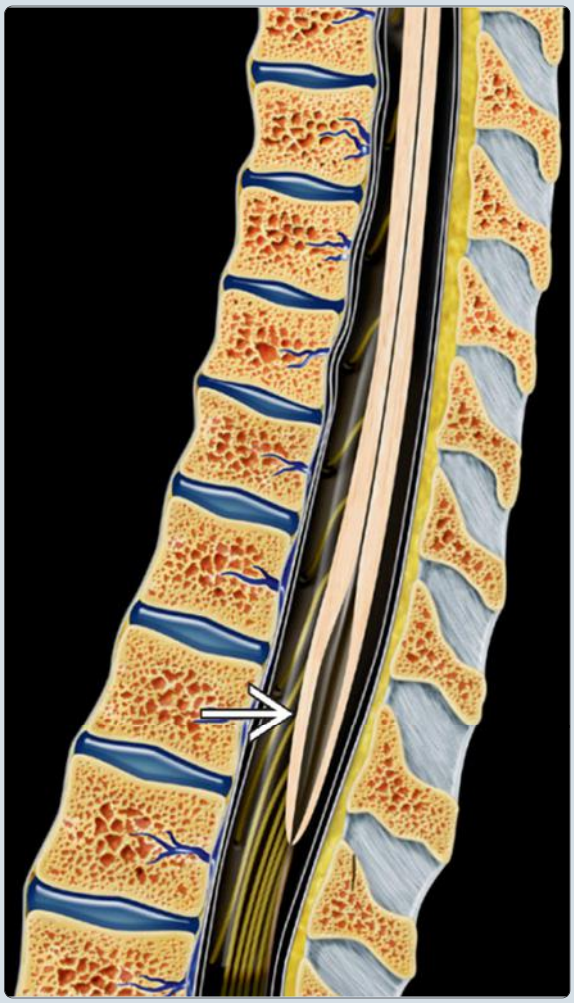
- Forms during embryogenesis (9th week) of caudal spinal cord by secondary neurulation
- Simple CSF-filled cavity of conus medullaris enclosed by ependymal tissue

CLINICAL ISSUES

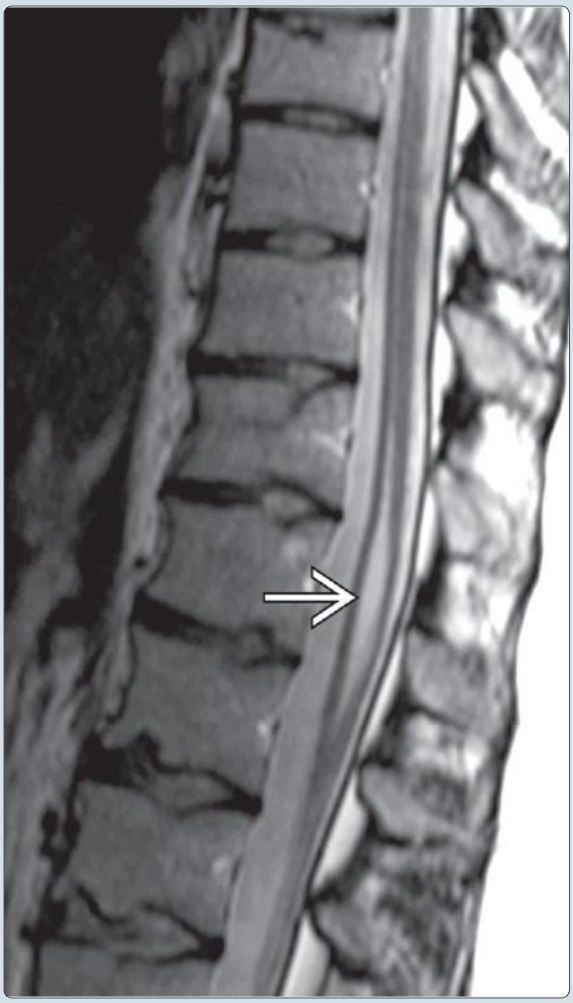
- No treatment indicated for classic incidental finding in asymptomatic patient
- Surgical decompression and management of associated abnormalities in rare symptomatic patients with cystic expansion

DIAGNOSTIC CHECKLIST

- Isolated mild dilatation of distal central canal in normally located conus is nearly always incidental normal finding
- Most important imaging goal is to distinguish from cystic cord neoplasm or syrinx
  - Calcification, septations, nodules, enhancement, or eccentric location all argue against ventriculus terminalis and prompt further evaluation
- Exclude other unsuspected abnormalities predisposing to syrinx or cord tethering



Sagittal graphic demonstrates the classic appearance of ventriculus terminalis ⇒ with uncomplicated fusiform dilatation of the distal central cord canal confined to the conus.



Sagittal T2WI MR shows the classic imaging appearance of ventriculus terminalis ⇒ with smooth dilation of the distal spinal cord canal in the conus. Multiple vertebral endplate Schmorl nodes are an unrelated finding.

TERMINOLOGY

Synonyms

- Terminal ventricle, 5th ventricle

Definitions

- CSF signal/density within central spinal cord with central canal expansion at conus/proximal filum level

IMAGING

General Features

- Best diagnostic clue
  - Mild cystic dilatation of distal central spinal cord canal without cord signal abnormality or enhancement
- Location
  - Distal spinal cord and conus medullaris
- Size
  - 2-4 mm (transverse); rarely exceeds 2 cm in length
- Morphology
  - Central intramedullary CSF signal/density dilatation with smooth regular margins and no mass
  - Conus terminates at normal level (T12 → L2)

CT Findings

- CECT
  - CSF density dilatation of distal spinal cord central canal
  - No mass lesion or enhancement

MR Findings

- T1WI
  - Hypointense CSF signal intensity dilatation of distal cord central canal
    - Beware of phase ghosting or truncation (Gibb) artifact imitating central canal dilatation
  - Conus terminates at normal level (T12 to L2)
  - No filum terminale thickening or lipoma
- T2WI
  - Hyperintense CSF signal intensity dilatation of distal cord central canal
  - No septation of dilated central canal
- T1WI C+
  - No associated mass lesion or enhancement
    - Differentiates from cystic cord neoplasm

Ultrasonographic Findings

- Grayscale ultrasound
  - Mild anechoic nonseptated central canal dilatation within normally situated conus
  - Normal nerve root and conus pulsation
  - No filum terminale thickening (< 2 mm normal) or lipoma
- M-mode
  - Useful to confirm normal nerve root and conus pulsation

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging
- Newborns
  - Ultrasound to screen for congenital anomalies
    - Distinguish ventriculus terminalis from syrinx or cord neoplasm
    - Abnormal findings should be confirmed with MR imaging
- Children, adults, and infants (with positive ultrasound studies)
  - Thin section sagittal T1WI and T2WI MR (3 mm slice thickness)
  - Axial T1WI and T2WI (3-4 mm slice thickness) distal cord to sacrum
    - Best to exclude occult dysraphism, lipoma, or thick filum
  - T1WI C+ MR in sagittal, axial planes to exclude mass

DIFFERENTIAL DIAGNOSIS

Transient Dilatation of Central Spinal Cord Canal

- Normal variant
- Slight dilatation of central canal in newborn
- Disappears in 1st weeks of life

Hydrosyringomyelia

- Cystic expansion of distal 1/3 (or more) of spinal cord
- Isolated finding or associated with congenital spine anomalies (up to 30% of patients)

Cystic Spinal Cord Neoplasm

- Astrocytoma
- Ependymoma
- Hemangioblastoma
- Differentiated by cord signal abnormality and expansion, contrast enhancement in solid portions

Myelomalacia

- History of trauma, vascular accident, or other cord insult
- Cord atrophy, ± T2 hyperintensity

PATHOLOGY

General Features

- Etiology
  - Forms during embryogenesis (9th week) via secondary neurulation (formerly canalization and retrogressive differentiation) of caudal spinal cord
    - Represents point of union between central canal portion formed by primary neurulation and portion formed by secondary neurulation
    - Central canal size usually regresses during 1st weeks after birth; persistence leads to identification in children or adults
- Associated abnormalities
  - Occasionally identified in conjunction with caudal regression or tethered cord

Staging, Grading, & Classification

- Proposed classification systems for cystic ventriculus terminalis stratifies to surgical therapy based on symptoms

Gross Pathologic & Surgical Features

- Simple CSF-filled cavity of conus medullaris enclosed by ependymal tissue
  - Normally present as either virtual cavity or ependymal residue
  - Small minority of patients demonstrate cystic expansion
- No gliosis or neoplasm

Microscopic Features

- Normal cord microscopic histology
  - Cystic cavity lined by ependymal cells
  - No gliosis or neoplasm

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Incidental finding on imaging performed for unrelated indications
    - Usually identified in adults during sciatica work-up
- Other signs/symptoms
  - Rarely becomes abnormally dilated and symptomatic, necessitating treatment
    - Bilateral sciatica
    - Cauda equina syndrome
    - Lower extremity weakness
    - Urinary retention
- Clinical profile
  - Patient usually asymptomatic or presents with nonspecific neurological symptoms

Demographics

- Age
  - Most commonly identified < 5 years old; may occur at any age
- Gender
  - M = F
- Epidemiology
  - 2.6% of normal children (< 5 years old) have visible ventriculus terminalis on MR
  - Less commonly identified in adults; primarily autopsy curiosity before widespread availability of MR imaging

Natural History & Prognosis

- Size variable throughout life; smallest in middle age, largest in early childhood and old age
- No effect on mortality or morbidity
  - Most commonly either stable in size or regresses; size progression not reported
- Rare symptomatic surgical cases show symptom improvement postoperatively

Treatment

- No treatment indicated for classic incidental finding in asymptomatic patient
  - MR follow-up if deemed necessary based on clinical findings
- Surgical decompression and management of associated abnormalities in rare symptomatic patients with cystic expansion
  - Cyst fenestration ± cyst shunting to subarachnoid space, pleural cavity, or peritoneal cavity

DIAGNOSTIC CHECKLIST

Consider

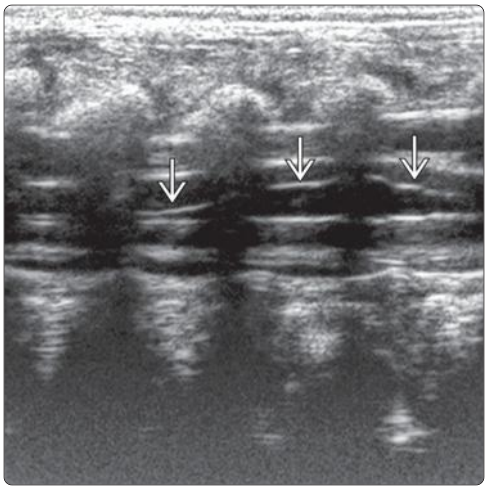
- Most important imaging goal is to distinguish from cystic cord neoplasm or syrinx
- Asymptomatic patients require no further imaging evaluation
- Symptomatic patients should be monitored using clinical and MR follow-up unless degree of cyst expansion prompts surgical drainage

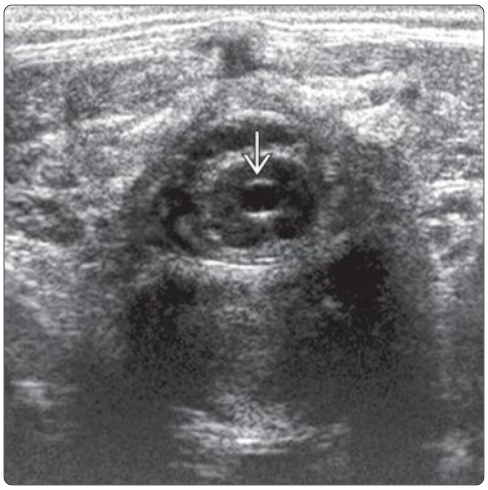
Image Interpretation Pearls

- Isolated mild dilatation of distal central canal in normally located conus is nearly always incidental normal finding
  - Calcification, septations, nodules, enhancement, or eccentric location all argue against ventriculus terminalis and prompt further evaluation
  - Beware of truncation or phase ghosting artifact mimicking dilated terminal ventricle or syrinx
- Exclude other unsuspected abnormalities that may predispose to syrinx or cord tethering before attributing finding to incidental dilatation of ventriculus terminalis

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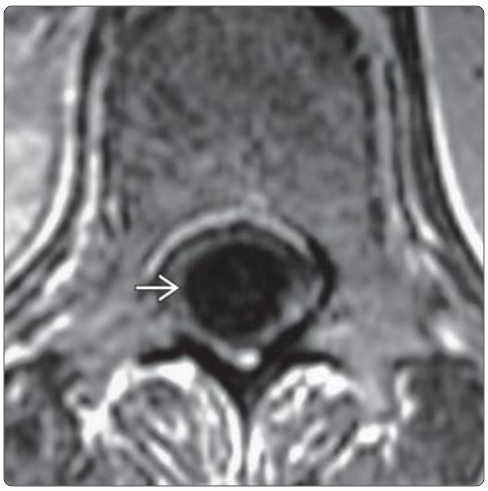
(Left) *Longitudinal ultrasound demonstrates smooth fusiform dilatation of the distal central canal ➡ localized to the conus.* **(Right)** *Transverse ultrasound reveals smooth dilation of the distal spinal cord central canal ➡ at the conus. Note the central location without eccentric cavitation or nodularity, findings that would exclude simple ventriculus terminalis.*





(Left) *Sagittal T1WI C+ FS MR demonstrates fluid signal intensity dilatation of the central spinal cord canal ➡ at the conus only. No incremental abnormal enhancement is present. Imaging of the thoracic and cervical spine was normal (not shown).* **(Right)** *Axial T2WI FS MR in an asymptomatic patient imaged for a cutaneous vascular malformation reveals incidental CSF signal dilatation of the conus central spinal cord canal ➡.*





(Left) *Sagittal T2WI MR depicts cystic expansion of the ventriculus terminalis, or "5th ventricle" ➡. The dilatation is centered within the central canal and confined to the conus only.* **(Right)** *Axial T1 C+ MR shows no incremental abnormal enhancement within the cystic expanded ventriculus terminalis ➡. The cystic dilatation is centered within the central canal and confined to the conus only, helping to make the diagnosis of variant ventriculus terminalis.*

Chiari 0

KEY FACTS

TERMINOLOGY

- Synonym: Chiari type 0

IMAGING

- Syringomyelia + low obex position, no cerebellar tonsillar ectopia
- MR cine: ± aberrant CSF flow at foramen magnum

TOP DIFFERENTIAL DIAGNOSES

- Chiari 1
- Complex Chiari
- Chiari 2

PATHOLOGY

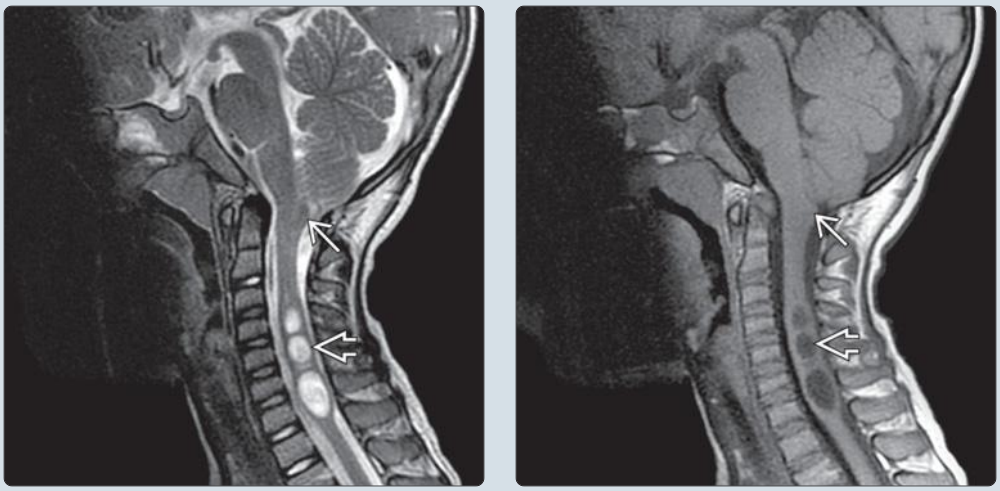
- No cerebellar tonsillar ectopia
- Caudal brainstem descent
- Small posterior fossa + AP foramen magnum enlargement
  - Crowded foramen magnum, arachnoid adhesions, 4th ventricular arachnoid veil
  - Cisterna magna impacted by tonsils ("tight cisterna magna")

CLINICAL ISSUES

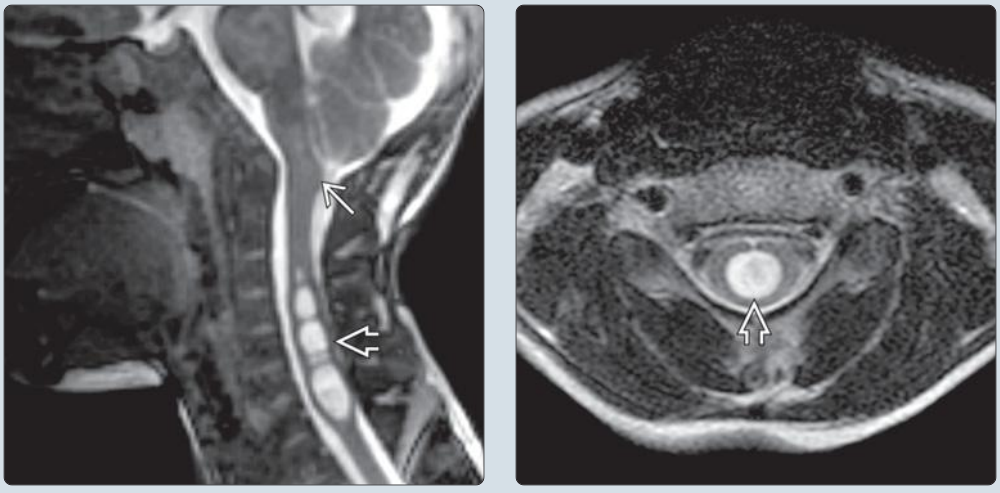
- Most common symptoms
  - Chiari-like headache
  - Neck or back pain
  - Scoliosis
- Carefully selected patients benefit from posterior fossa decompression + duraplasty
  - Syrinx frequently improves or resolves without syrinx drainage catheter placement

DIAGNOSTIC CHECKLIST

- CM 0 rare; diagnosis made only after other syrinx etiologies excluded
- Existence as separate diagnosis is controversial



(Left) *Sagittal T2WI MR reveals syringohydromyelia ⇨ of the midcervical spinal cord in conjunction with low obex ⇨ position. The cerebellar tonsils are rounded without abnormal caudal descent.* **(Right)** *Sagittal T1WI MR confirms midcervical syringohydromyelia ⇨. No cerebellar tonsillar ectopia is present to explain syringohydromyelia, although the obex ⇨ is low, prompting clinical consideration of Chiari 0 malformation.*



(Left) *Sagittal 3D FIESTA image (from a cine MR clip) demonstrates moderate cervical syringohydromyelia ⇨ and low obex ⇨ without abnormal cerebellar tonsillar ectopia. In this patient, the cine MR clip did not confirm suspected aberrant CSF flow at the foramen magnum.* **(Right)** *Axial T2WI MR in a patient clinically suspected to have Chiari 0 malformation shows moderate cervical syringohydromyelia ⇨.*

TERMINOLOGY

Abbreviations

- Chiari 0 (CM 0)

Synonyms

- Chiari type 0

Definitions

- Syringomyelia without cerebellar tonsillar herniation
- Headache, typical Chiari 1 malformation (CM1) symptoms

IMAGING

General Features

- Best diagnostic clue
  - Syringomyelia + low obex position, no cerebellar tonsillar ectopia
- Morphology
  - Obex below normal level
  - ↑ anteroposterior foramen magnum diameter

CT Findings

- Normal cerebellar tonsil position
- Syringohydromyelia

MR Findings

- Normal cerebellar tonsil position + low obex
- Syringohydromyelia
- MR cine: ± aberrant CSF flow at foramen magnum

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR with sagittal cine imaging

DIFFERENTIAL DIAGNOSIS

Chiari 1

- Cerebellar tonsillar ectopia ± low obex
- ± syringohydromyelia

Complex Chiari

- Cerebellar tonsillar ectopia + low obex
- ± syringohydromyelia
- Platybasia, CVJ anomalies more prevalent than CM1

Chiari 2

- Characteristic tectal beak, tonsillar/vermian herniation, medullary kink
- Nearly 100% of patients have concurrent myelomeningocele

PATHOLOGY

General Features

- Small posterior fossa + AP foramen magnum enlargement
- No cerebellar tonsillar ectopia
- Caudal brainstem descent (measured using obex/nucleus gracilis position)
- Etiology
  - Putative causes of disrupted CSF flow include 4th ventricle arachnoid veil, foramen of Magendie arachnoid adhesions
  - Syringohydromyelia may represent communicating central canal dilation

Gross Pathologic & Surgical Features

- Physical barrier to CSF flow may be discovered at surgery → abnormal CSF flow at posterior fossa, foramen magnum
  - Crowded foramen magnum, arachnoid adhesions, 4th ventricular arachnoid veil
  - Cisterna magna impacted by tonsils ("tight cisterna magna")

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chiari-like headache
  - Neck or back pain
  - Scoliosis
- Other signs/symptoms
  - Torticollis
  - Gait disturbance
  - Paresthesias

Demographics

- Epidemiology
  - Very small cohort within Chiari malformations category
    - 3.7% of syringomyelia patients at 1 large children's hospital

Treatment

- Carefully selected patients benefit from posterior fossa decompression + duraplasty
  - Syrinx frequently improves or resolves without syrinx drainage catheter placement

DIAGNOSTIC CHECKLIST

Consider

- CM 0 very small subgroup of syringohydromyelia patients
  - Diagnosis made only after other syrinx etiologies have been excluded

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KEY FACTS

TERMINOLOGY

- Synonyms: CM1, Arnold-Chiari 1 malformation (AC1)

IMAGING

- Pointed cerebellar tonsils extend ≥ 5 mm below foramen magnum ± syringohydromyelia, scoliosis, hydrocephalus

TOP DIFFERENTIAL DIAGNOSES

- Normal low-lying tonsils below foramen magnum
- Acquired tonsillar herniation ("acquired Chiari 1")

PATHOLOGY

- Etiology not fully understood
  - Postulated mechanisms include hydrodynamic theory and posterior fossa underdevelopment theory
- May present as isolated finding or in conjunction with syndromic (4th occipital sclerotome) or nonsyndromic skull base and CVJ anomalies

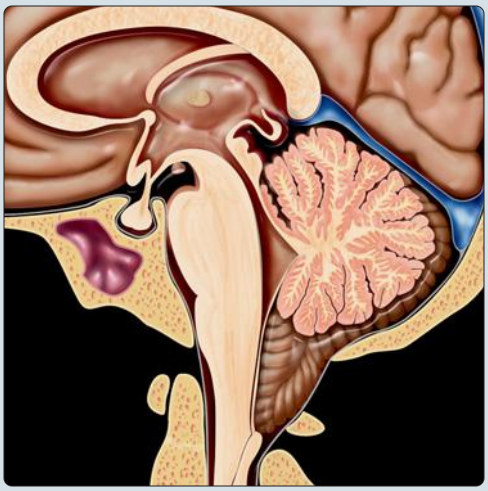
CLINICAL ISSUES

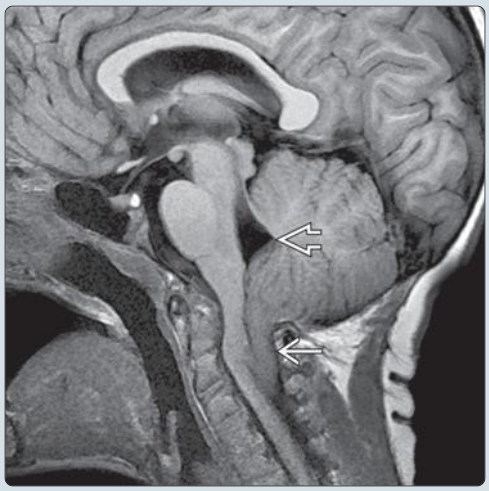
- Up to 50% of CM1 asymptomatic
  - Surgical treatment for asymptomatic patients controversial
- Symptomatic patients
  - Surgical goal is restoration of normal CSF flow at foramen magnum
  - PF decompression and resection of posterior C1 arch ± duraplasty, cerebellar tonsil resection

DIAGNOSTIC CHECKLIST

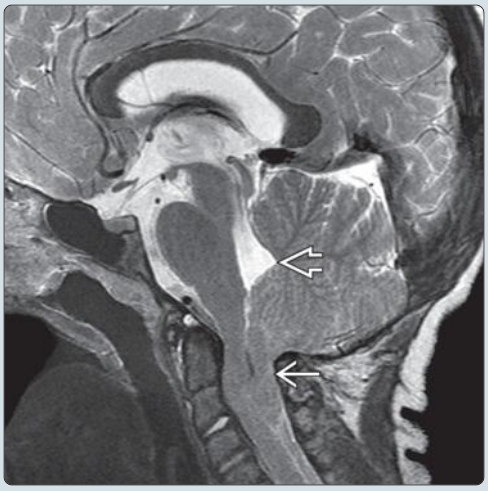
- Tonsillar herniation > 12 mm usually symptomatic
- Probably not clinically significant CM1 unless tonsils > 5 mm &/or pointed

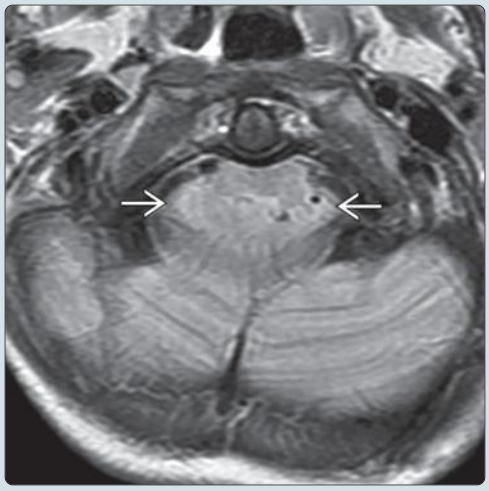
(Left) *Sagittal graphic demonstrates pointed peg-like tonsils extending below the foramen magnum, elongating the normally positioned 4th ventricle.* **(Right)** *Sagittal T1WI MR shows the normal position and appearance of the 4th ventricle. The fastigium ⇨ is in normal position, helping to distinguish from Chiari 2 malformation. There is inferior displacement of the ectopic cerebellar tonsils ⇨ through the foramen magnum with ventral spinal cord displacement.*





(Left) *Sagittal T2WI MR shows the normal position and appearance of the 4th ventricle. The fastigium ⇨ is in normal position, helping to distinguish from the Chiari 2 malformation. Note inferior displacement of the ectopic elongated cerebellar tonsils ⇨ through the foramen magnum that produces mass effect on the upper cervical spinal cord.* **(Right)** *Axial T2WI MR shows inferior displacement of the ectopic cerebellar tonsils ⇨ through the foramen magnum with crowding of the foramen magnum.*





TERMINOLOGY

Abbreviations

- Chiari 1 malformation (CM1)

Synonyms

- Arnold-Chiari 1 malformation (AC1)

Definitions

- Elongated, peg-shaped cerebellar tonsils extend below foramen magnum into cervical spinal canal

IMAGING

General Features

- Best diagnostic clue
  - Pointed cerebellar tonsils ≥ 5 mm below foramen magnum ± syringohydromyelia (14-75%)
- Location
  - Craniovertebral junction (CVJ)
- Size
  - Tonsils > 5 mm below foramen magnum
- Morphology
  - Low-lying, pointed peg-like tonsils with oblique vertical sulci, elongated but normally located 4th ventricle

Radiographic Findings

- Radiography
  - Either normal or short clivus, CVJ segmentation/fusion anomalies, small posterior fossa (PF)

CT Findings

- NECT
  - Small PF → low torcular, "crowded" foramen magnum, tonsillar ectopia, flattened spinal cord
- Bone CT
  - Often normal; abnormal cases → short clivus, CVJ segmentation/fusion anomalies, small PF

MR Findings

- T1WI
  - Pointed (not rounded) tonsils ≥ 5 mm below foramen magnum
  - "Tight foramen magnum" with small/absent cisterns
  - ± elongation of 4th ventricle, hindbrain anomalies
- T2WI
  - Oblique tonsillar folia (like "sergeant's stripes")
  - ± short clivus → "apparent" descent of 4th ventricle, medulla
  - ± syringohydromyelia (14-75%)
- MR cine
  - Cine phase contrast (PC) MR: Disorganized CSF pulsation, ↑ brainstem/cerebellar tonsil motion → ↑ peak systolic velocity, ↓ flow through foramen magnum

Nonvascular Interventions

- Myelography
  - Low-lying tonsils efface CSF at foramen magnum

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Multiplanar T1WI, T2WI of spinal axis and posterior fossa, PC MR CSF flow study

DIFFERENTIAL DIAGNOSIS

Normal Tonsillar Displacement Below Foramen Magnum

- Tonsils may normally lie below foramen magnum
- Unless tonsils > 5 mm &/or pointed, probably not clinically significant Chiari 1 malformation

Acquired Tonsil Herniation ("Acquired Chiari 1")

- Acquired basilar invagination → small posterior fossa
  - Osteogenesis imperfecta
  - Paget disease
  - Craniosynostosis
  - Rickets
  - Achondroplasia
  - Acromegaly
- "Pull from below" (LP shunt, CSF leak) 2° to intracranial hypotension
  - "Sagging" brainstem, tonsillar herniation, smooth dural enhancement, dilated epidural plexus, retrospinal C1/2 fluid collection, spinal hygroma
- "Push from above"
  - Chronic VP shunt; thick skull, premature sutural fusion, arachnoidal adhesions
  - ↑ intracranial pressure (ICP), intracranial mass

PATHOLOGY

General Features

- Etiology
  - Hydrodynamic theory
    - Systolic piston-like descent of impacted tonsils/medulla → plugging of CSF pathway at foramen magnum
    - During diastole, rapid recoil of brain stem/tonsils disimpacts foramen magnum, permits normal CSF diastolic pulsation
  - Posterior fossa underdevelopment theory
    - Underdevelopment of occipital somites of paraaxial mesoderm produce small posterior fossa → 2° tonsillar herniation
    - Not all CM1 patients have small posterior fossas
- Genetics
  - Autosomal dominant inheritance with reduced penetrance or autosomal recessive inheritance
  - Syndromic/familial associations
    - Velocardiofacial/microdeletion chromosome 22, Williams syndrome, craniosynostosis, achondroplasia, Hajdu-Cheney syndrome, and Klippel-Feil syndrome
- Associated abnormalities
  - 4th occipital sclerotome syndromes (50%): Short clivus, craniovertebral segmentation/fusion anomalies
  - Osseous skull base/skeletal anomalies (25-50%)
    - Scoliosis ± kyphosis (42%); left thoracic curve
    - Retroflexed odontoid process (26%)
    - Platybasia, basilar invagination (25-50%)
    - Klippel-Feil syndrome (5-10%)
    - Incomplete C1 ring ossification (5%)
    - Atlantooccipital assimilation (1-5%)

- Syringomyelia (30-60%) → 60-90% in symptomatic CM1 patients
  - Most common C4-C6; holocord hydrosyringomyelia, cervical/upper thoracic syrinx, syringobulbia uncommon
- Hydrocephalus (11%)
- Underdeveloped occipital enchondrium → small posterior fossa vault, downward hindbrain herniation
- Foramen magnum obstruction → decreased communication between cranial, spinal CSF compartments

Staging, Grading, & Classification

- Diagnostic criteria: Herniation of at least 1 cerebellar tonsil > 5 mm or herniation of both tonsils ≥ 3-5 mm below a line connecting basion with opisthion
  - Herniation of both tonsils ≥ 3-5 mm below foramen magnum + syrinx, cervicomedullary kinking, 4th ventricular elongation, or pointed tonsils → congenital CM1
  - Tonsil herniation ≤ 5 mm does not exclude CM1

Gross Pathologic & Surgical Features

- Herniated, sclerotic tonsils grooved by opisthion
- Arachnoidal scarring and adhesions at foramen magnum

Microscopic Features

- Tonsillar softening or sclerosis with Purkinje/granular cell loss

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Up to 50% asymptomatic (especially if ≤ 5 mm caudal displacement)
  - Symptomatic patients present with constellation of findings
    - Sudden death (rare)
    - Suboccipital headache, cranial nerve palsy, ocular disturbances, otoneurologic dysfunction, cord motor or sensory abnormalities, gait disturbance, neuropathic joint
    - Tonsillar herniation > 12 mm nearly always symptomatic; ~ 30% with tonsils 5-10 mm below foramen magnum asymptomatic
    - CM1 patients with syrinx nearly always present with symptoms referable to syrinx; if syrinx extends into medulla, bulbar symptoms predominate
  - Trauma is common precipitating event for symptom onset (24%)
- Clinical profile
  - Clinical CM1 syndrome: Headache, pseudotumor-like episodes, Ménière disease-like syndrome, lower cranial nerve and spinal cord signs

Demographics

- Age
  - 10 months to 65 years; syrinx, congenital CVJ anomalies hasten presentation
- Gender
  - F > M (3:2)
- Epidemiology
  - Incidence 0.01-0.6% all age groups, 0.9% pediatric patient groups

Natural History & Prognosis

- Natural history not clearly understood
  - Many patients asymptomatic for prolonged periods
  - Increasing ectopia + time → ↑ likelihood symptoms
- Children respond better to treatment than adults
- Degree of tonsillar herniation correlates with clinical severity

Treatment

- Treating asymptomatic patients is controversial
  - "Radiographic" CM1 (no syrinx), no corresponding clinical signs/symptoms → conservative management
  - Asymptomatic Chiari 1 + syrinx → consider decompression
- Symptomatic patients: PF decompression/resection of posterior arch C1, ± duraplasty, cerebellar tonsil resection
  - > 90% ↓ brainstem signs
  - > 80% ↓ hydrosyringomyelia
  - ± scoliosis arrest
- Direct shunting of symptomatic syrinx obsolete; goal to restore normal CSF flow at foramen magnum

DIAGNOSTIC CHECKLIST

Consider

- Unless tonsils > 5 mm &/or pointed, probably not clinically significant Chiari 1 malformation

Image Interpretation Pearls

- Tonsillar herniation > 12 mm nearly always symptomatic

SELECTED REFERENCES

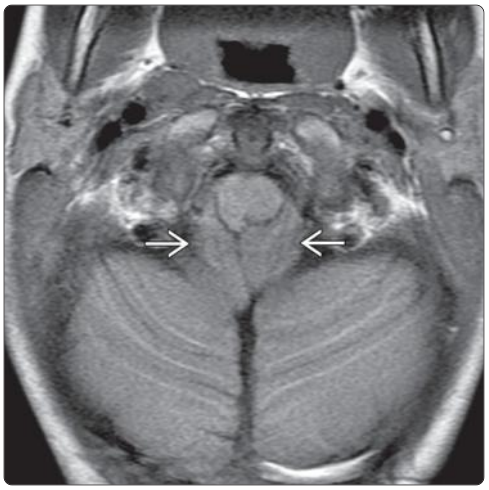
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(Left) *Sagittal T2WI MR in a case of asymptomatic CM1 demonstrates severe cerebellar tonsillar ectopia producing deformation of the upper cervical spinal cord with abnormal cord T2 prolongation from edema and myelomalacia.* **(Right)** *Axial T2WI MR in a case of asymptomatic CM1 reveals caudal extension of the ectopic cerebellar tonsils into the foramen magnum, completely effacing the basilar cisterns and displacing the adjacent cord.*





(Left) *Sagittal T2WI MR depicts mild abnormal pointed configuration of the ectopic cerebellar tonsils. The 4th ventricle is in normal position. There is also midcervical syringohydromyelia producing spinal cord expansion.* **(Right)** *Axial T1WI MR demonstrates crowding of the ectopic cerebellar tonsils at the foramen magnum, with distortion of the adjacent cervical spinal cord. It is important to exclude cervical syringohydromyelia when ectopic tonsils are identified.*





(Left) *Sagittal T2WI MR depicts marked cerebellar tonsillar ectopia with normal tectum and 4th ventricular position. The clivus is also mildly foreshortened. There is central intramedullary edema in the cervical spinal cord without frank syringomyelia, a finding that has been described as reflecting a "presyrinx" state.* **(Right)** *Axial T2WI MR confirms displacement of the cerebellar tonsils through the foramen magnum.*

KEY FACTS

TERMINOLOGY

- Complex Chiari malformation (CCM), Chiari 1.5

IMAGING

- Cerebellar tonsillar herniation with low obex, dorsal medullary "bump"
  - "Bump" thought to represent aberrant clava, used as marker for cervicomedullary junction
  - Abnormal tonsillar "pistoning" motion, reduced CSF flow around foramen magnum and cerebellar tonsils
- ± syringohydromyelia, ventral cervicomedullary compression
- ± odontoid retroflexion, small posterior fossa, clival anomalies, platybasia

TOP DIFFERENTIAL DIAGNOSES

- Chiari 1 malformation
- Chiari 2 malformation

PATHOLOGY

- Medullary pyramid decussation anatomic border between spinal cord, medulla oblongata
  - Normal obex level is 10-12 mm above foramen magnum
  - Obex displaced to or below foramen magnum level in CCM patients

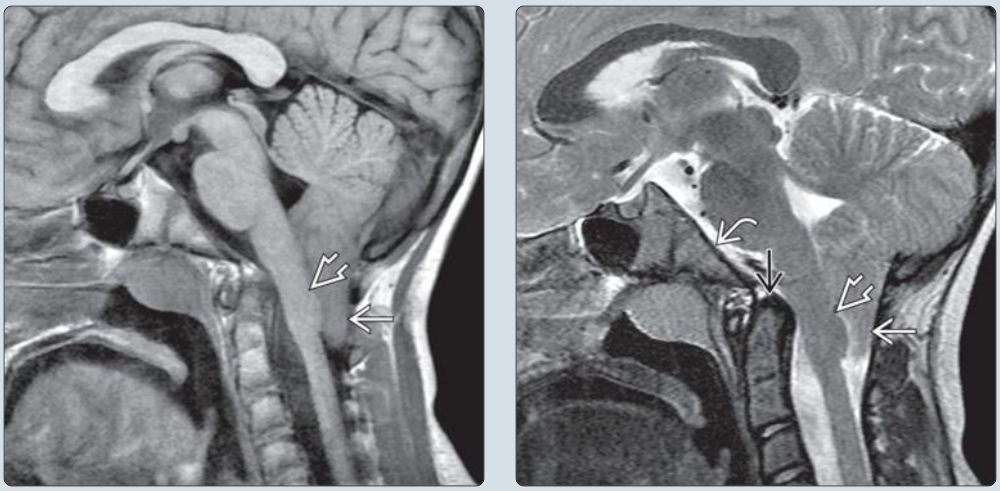
CLINICAL ISSUES

- Headaches, myelopathy, bulbar symptoms, lethargy, failure to thrive

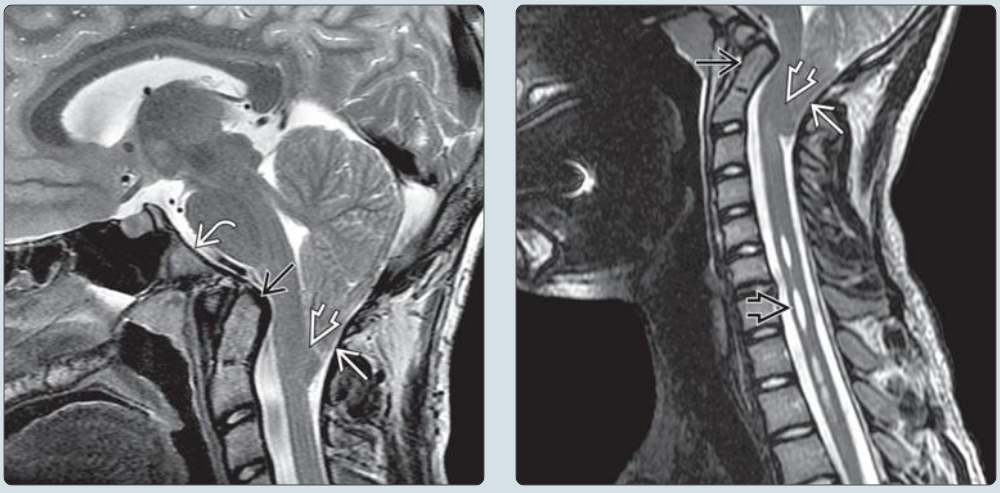
DIAGNOSTIC CHECKLIST

- Consider CCM in context of cerebellar tonsillar herniation + low obex, dorsal medullary "bump"
- CCM considered subgroup of Chiari 1 malformation with more severe clinical phenotype

(Left) *Sagittal T1WI MR shows classic features of a complex Chiari malformation, including marked cerebellar tonsil ectopia and inferior displacement of the obex. The 4th ventricle is in normal position.* **(Right)** *Sagittal T2WI MR reveals characteristic tonsillar ectopia and inferior displacement of the cervicomedullary junction, marked by position of the obex below the foramen magnum. There is also mild retroflexion of the odontoid process and mild clivus foreshortening.*



(Left) *Sagittal T2WI MR (different patient) shows a prominent obex and pointed, ectopic cerebellar tonsils. The odontoid process is retroflexed and the clivus is foreshortened, with a reduced craniocervical angle with platybasia.* **(Right)** *Sagittal T2WI MR (different patient) reveals striking odontoid retroflexion. Note also severe cerebellar tonsillar ectopia extending inferiorly to the C3 level and cervical syringohydromyelia. The prominent, inferiorly displaced obex confirms a complex Chiari malformation.*



TERMINOLOGY

Abbreviations

- Complex Chiari malformation (CCM), Chiari 1.5

Definitions

- Caudal cerebellar tonsil and brainstem herniation with dorsal cervicomedullary "bump"
- Frequently associated craniovertebral junction (CVJ) anomalies

IMAGING

General Features

- Best diagnostic clue
  - Cerebellar tonsillar herniation with low obex, dorsal medullary "bump"
- Location
  - Posterior fossa, craniovertebral junction
- Morphology
  - Dorsal medullary "bump" thought to represent aberrant clava, used as marker for cervicomedullary junction

CT Findings

- Bone CT
  - May show odontoid retroflexion, small posterior fossa, clival anomalies, platybasia

MR Findings

- T1WI
  - Cerebellar tonsillar ectopia + low obex, dorsal medullary "bump"
    - Obex descends to or below foramen magnum
  - ± retroflexed odontoid, foreshortened clivus
- T2WI
  - Same as T1WI ± syringohydromyelia, ventral cervicomedullary compression
- MR cine
  - Abnormal tonsillar "pistoning" motion, reduced CSF flow around foramen magnum and cerebellar tonsils

DIFFERENTIAL DIAGNOSIS

Chiari 1

- Cerebellar tonsillar ectopia ± low obex position
  - If obex inferiorly displaced, more modest than CCM and no dorsal "bump"
  - ± syringomyelia, retroflexed odontoid process, platybasia, CVJ anomalies
- Generally less symptomatic than CCM patients

Chiari 2

- Tectal beaking, cerebellar tonsil/vermis ectopia, medullary kink, other characteristic brain findings
- Associated with myelomeningocele

PATHOLOGY

General Features

- Associated abnormalities
  - Platybasia, occipital sclerotomal anomalies, Klippel-Feil spectrum

Gross Pathologic & Surgical Features

- Medullary pyramid decussation anatomic border between spinal cord, medulla oblongata
  - Pyramidal decussation difficult to image; obex position used as surrogate imaging landmark for cervicomedullary junction
  - Obex is thin band of neural tissue between area of postrema nuclei
    - Forms dorsal roof of central canal opening
    - Apex defines inferior margin of foramen of Magendie
  - Normal obex level is 10-12 mm above foramen magnum
  - Obex displaced to or below foramen magnum level in CCM patients

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Headaches
- Other signs/symptoms
  - Myelopathy
  - Bulbar symptoms
  - Lethargy, failure to thrive

Natural History & Prognosis

- Progressively severe symptoms

Treatment

- Some patients (rarely) have minimal or no symptoms and require only observation
- Symptomatic patients generally require > 1 operation
  - Standard Chiari decompression procedure with dural patch
  - May also require dens resection, CVJ fusion to address platybasia, cervicomedullary compression

DIAGNOSTIC CHECKLIST

Consider

- CCM in context of cerebellar tonsillar herniation + low obex, dorsal medullary "bump"
- Growing repudiation of term "Chiari 1.5 malformation"
  - Incorrectly implies that patients reside along spectrum between Chiari 1, Chiari 2 malformations
  - Complex Chiari malformation is best term to refer to these patients

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KEY FACTS

TERMINOLOGY

- Synonyms: Chiari malformation 2 (CM2), Chiari II
- Complex hindbrain malformation
- Virtually 100% associated with neural tube closure defect (NTD), usually lumbar myelomeningocele (MMC)

IMAGING

- Cerebellum "wraps" around medulla and "towers" through incisura, with "beaked" tectum and heart-shaped midbrain
- Virtually 100% have neural tube closure defect (NTD)

TOP DIFFERENTIAL DIAGNOSES

- Chiari 1 malformation
- Chiari 3 malformation
- Intracranial CSF hypotension
- Severe, chronic shunted hydrocephalus (congenital)

PATHOLOGY

- Secondary to sequelae of CSF leakage through open spinal dysraphism during gestation (4th fetal week)
- Methylenetetrahydrofolate reductase (*MTHFR*) mutations → abnormal folate metabolism
- Spine and brain/skull associated anomalies common

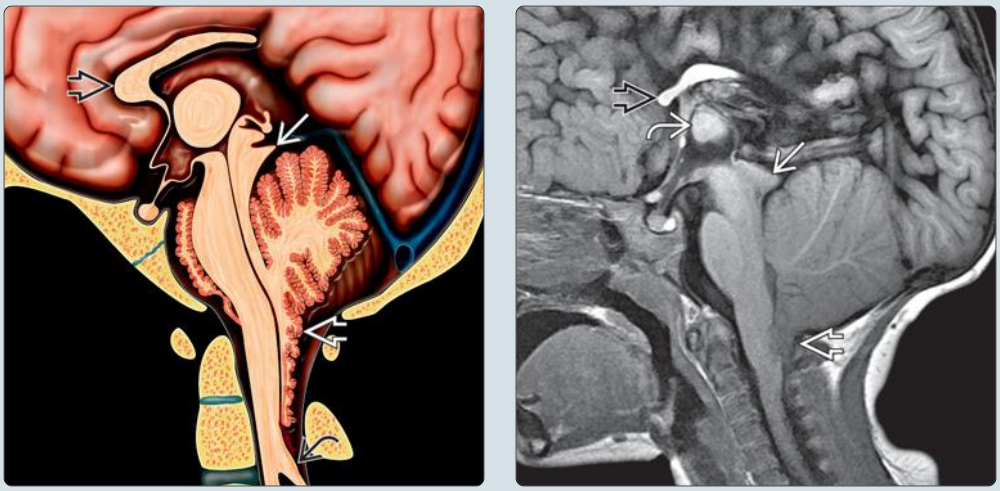
CLINICAL ISSUES

- Chiari 2 malformation most common cause of death in myelomeningocele patients
  - Brainstem compression/hydrocephalus, intrinsic brainstem "wiring" defects

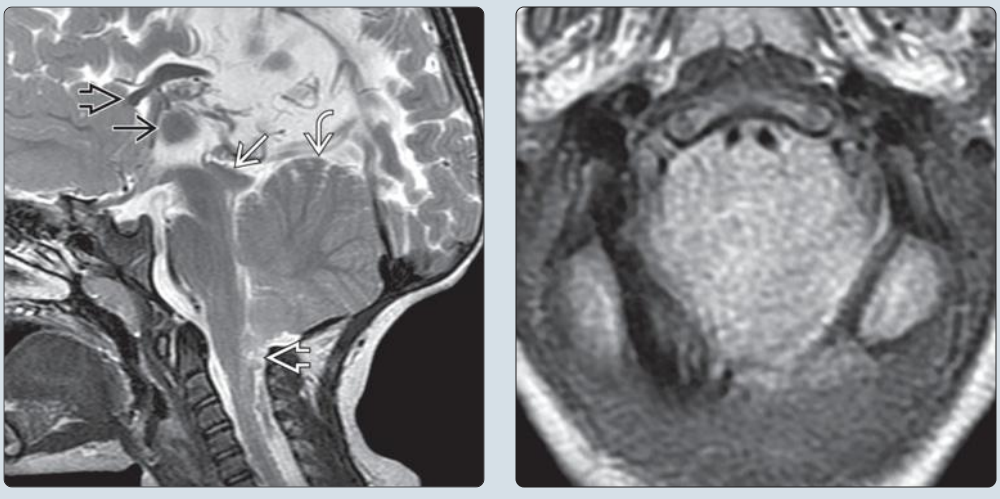
DIAGNOSTIC CHECKLIST

- Towering cerebellum, downward vermian displacement, ± brainstem compression diagnostic for Chiari 2

(Left) *Sagittal graphic of the posterior fossa and upper cervical spine demonstrates characteristic findings of Chiari 2 malformation, including callosal dysgenesis, tectal beaking, small posterior fossa, vermian ectopia, and medullary kinking.* **(Right)** *Sagittal T1WI MR reveals characteristic Chiari 2 malformation findings. Note tectal beaking and vermian displacement through the foramen magnum, large massa intermedia, and dysplastic corpus callosum.*



(Left) *Sagittal T2WI MR confirms characteristic findings of Chiari 2 malformation including tectal beaking, vermian displacement through the foramen magnum, "towering cerebellum", large massa intermedia, and dysplastic corpus callosum.* **(Right)** *Axial T2WI MR shows characteristic posterior fossa crowding at the foramen magnum, reflecting small dimensions of the posterior fossa combined with cerebellar ectopia and vermian displacement through the foramen magnum.*



TERMINOLOGY

Synonyms

- Chiari 2 malformation (CM2), Chiari type II

Definitions

- Complex hindbrain malformation
  - Virtually 100% associated with neural tube closure defect (NTD), usually lumbar myelomeningocele (MMC)
    - Rare reports in closed spinal dysraphism (probably misinterpreted Chiari 1)

IMAGING

General Features

- Best diagnostic clue
  - Cerebellar downward herniation with concurrent myelomeningocele
- Location
  - Posterior fossa, upper cervical spine; syrinx may involve entire cord
- Size
  - Posterior fossa smaller than normal
- Morphology
  - Cerebellum "wraps" around medulla and "towers" through incisura, with "beaked" tectum and heart-shaped midbrain

Radiographic Findings

- Radiography
  - Lateral skull → low torcular Herophili ± "lacunar" skull (universal at birth, largely resolves by 2nd year)

CT Findings

- NECT
  - Crowded posterior fossa, widened tentorial incisura, tectal beaking, and inferior vermian displacement
- Bone CT
  - Small posterior fossa (PF)
    - Low-lying tentorium/torcular inserts near foramen magnum
    - Large, funnel-shaped foramen magnum with "notched" opisthion
    - "Scalloped" posterior petrous pyramids, clivus
  - Posterior C1 arch anomalies (66%), enlarged cervical canal

MR Findings

- T1WI
  - "Cascade" or "waterfall" of cerebellum/brainstem downward
    - Uvula/nodulus/pyramid of vermis → sclerotic "peg"
    - Cervicomedullary "kink" (70%)
    - "Towering" cerebellum → compresses midbrain, associated beaked tectum
    - 4th ventricle elongated with no posterior point (fastigium)
  - Open spinal dysraphism, MMC ~ 100% (lumbar > > cervical)
  - Hydrosyringomyelia (20-90%)
- T2WI
  - Similar to T1WI + hyperintense, gliotic cerebellar tissue
  - ± 4th ventricular lesions (rare)
    - Roof of 4th ventricle adjacent/within choroid plexus
    - Glial or arachnoidal cysts, glial or choroidal nodules, subependymoma
- MR cine
  - Phase-contrast cine MR → restricted CSF flow through foramen magnum
- Diffusion tensor imaging (DTI)
  - Fractional anisotropy (FA) map and quantitative analysis defines callosal dysgenesis, confirms abnormal white matter architecture

Ultrasonographic Findings

- Grayscale ultrasound
  - Fetal obstetrical ultrasound pivotal for early diagnosis
    - MMC may be identified as early as 10 weeks
    - Characteristic brain findings ("lemon" and "banana" signs) seen as early as 12 weeks

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR for initial brain, spine evaluation
  - Follow-up brain CT or MR to assess hydrocephalus
  - Cervical spine MR for progressive brainstem or spinal symptoms

DIFFERENTIAL DIAGNOSIS

Chiari 1

- No association with myelomeningocele
- Tonsillar herniation (not vermis)

Chiari 3

- Chiari 2 malformation + cervical dysraphism, cephalocele

Intracranial Hypotension

- Symptomatic expression of low CSF pressure; distinguishable by clinical onset and symptoms
- Slumping posterior fossa with pons compressed against clivus, dural thickening/enhancement

Severe, Chronic Shunted Hydrocephalus (Congenital)

- May cause collapsed brain, upwardly herniated cerebellum

PATHOLOGY

General Features

- Etiology
  - Secondary to sequelae of CSF leakage through open spinal dysraphism during gestation (4th fetal week)
    - Abnormal neurulation → CSF escapes through NTD → failure to maintain 4th ventricular distention → hypoplastic PF chondrocranium → displaced/distorted PF contents
    - Exceedingly rare cases of closed spinal dysraphism with Chiari 2 malformation may contradict this theory
  - Alternative theory proposes association between Chiari 2 malformation and myelomeningocele is due to rostral and caudal neural tube dysgenesis
- Genetics
  - Methylenetetrahydrofolate reductase (*MTHFR*) mutations associated with abnormal folate metabolism

  - *MTHFR* mutations **plus** folate deficiency → ↑ risk NTD → Chiari 2
  - 4-8% risk of 2nd affected child
- Associated abnormalities
  - Spine
    - Open dysraphism (MMC) ~ 100% (lumbar > > cervical)
    - Posterior arch C1 anomalies (66%)
    - Syringohydromyelia (20-90%)
    - Diastematomyelia (5%)
    - Klippel-Feil syndrome
    - Cervical myelocystocele
  - Brain/skull
    - Corpus callosum (CC) dysgenesis (90%), aqueductal stenosis, rhombencephalosynapsis, gray matter malformations, absent septum pellucidum, fused forniceal columns
    - Lacunar skull (Lückenschädel)
- Hydrocephalus and severity of brain malformation relate to size of PF, degree of caudal hindbrain descent

Gross Pathologic & Surgical Features

- Small PF → contents shift down into cervical spinal canal
  - Cerebellar hemispheres/tonsils "wrap" around medulla
  - Pons/cranial nerve roots often elongated
  - Compressed/elongated/low 4th ventricle → pouch in cervical canal
  - Medullary kink
  - ± syringohydromyelia

Microscopic Features

- Purkinje cell loss, sclerosis within herniated tissues

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neonate: MMC, enlarging head circumference ± hydrocephalus symptoms
  - Older child/adult: Clinical hydrocephalus, symptoms referable to tethered cord (MMC repair)
  - All age groups: Varying degrees of lower extremity paralysis/sphincter dysfunction/bulbar signs
- Clinical profile
  - Usually presents within context of known MMC
    - Infants: Enlarging head circumference
    - Child/adult: Known Chiari 2 malformation, signs of hydrocephalus/shunt failure ± bulbar symptoms
- Laboratory
  - Fetal screening: ↑ alpha-fetoprotein

Demographics

- Age
  - Usually presents at birth with MMC ± hydrocephalus
- Gender
  - M = F
- Epidemiology
  - Incidence: 0.44 per 1,000 births, ↓ with folate replacement therapy

Natural History & Prognosis

- Chiari 2 malformation most common cause of death in MMC
  - Brainstem compression/hydrocephalus, intrinsic brainstem "wiring" defects
- Progression of spinal neurological deficits is rare; suspect hydrocephalus, associated undiagnosed spinal deformity (diastematomyelia), tethered cord
- Cerebellar tonsil/vermian ectopia may "improve" (ascend) following postnatal repair

Treatment

- Folate supplement for pregnant mothers (preconception → 6 weeks postconception) significantly decreases MMC risk
- Surgical management
  - Chiari decompression with resection of posterior foramen magnum, C1 ring
  - CSF diversion/shunting
  - Fetal MMC repair in selected patients may ameliorate Chiari 2 severity

DIAGNOSTIC CHECKLIST

Consider

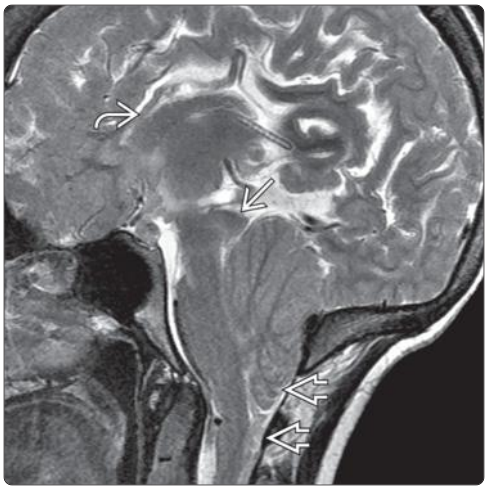
- Brain/spinal axis MR to detect presence of Chiari 2, assess severity, look for complications

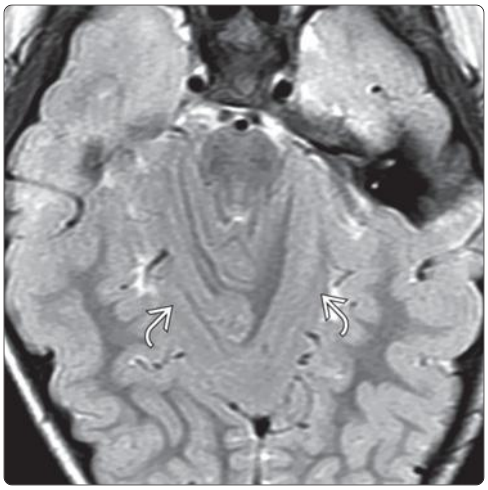
Image Interpretation Pearls

- Low torcular Herophili indicates small posterior fossa
- CT or MR showing towering cerebellum, downward vermian displacement, ± brainstem compression diagnostic of Chiari 2

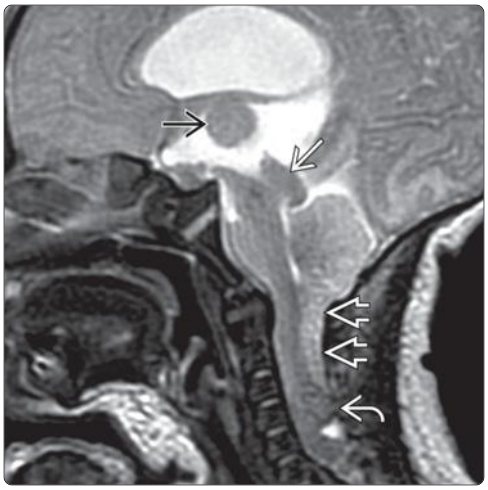
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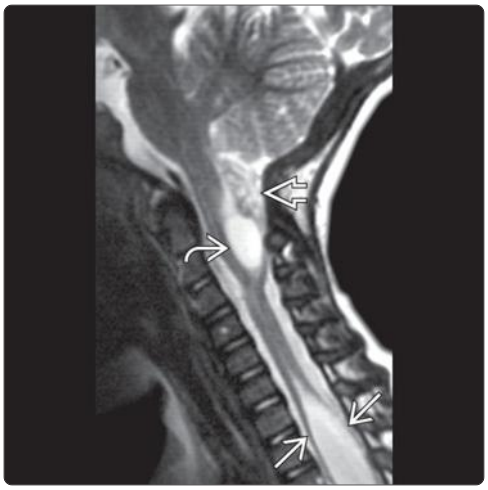
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(Left) *Sagittal T2WI MR of the brain shows characteristic Chiari 2 findings with very small posterior fossa size, prominent tectal beak, and vermian/tonsillar extension through the foramen magnum. The corpus callosum is severely dysplastic and the ventricles are small following CSF shunting.* **(Right)** *Axial T2WI MR through the posterior fossa demonstrates the "towering" cerebellum extending through the wide tentorial incisura.*





(Left) *Sagittal T2WI MR depicts marked vermian ectopia and a prominent medullary kink positioned lower than typically seen at the C4 level. Note also the dysplastic "beaked" tectum and prominent massa intermedia.* **(Right)** *Sagittal T2WI MR shows crowded posterior fossa with vermian ectopia and a large cervicothoracic cord syrinx as well as a sizable focal cervicomedullary syrinx (syringobulbia). Note hyperintense gliotic changes of scarred vermis.*





(Left) *Sagittal T2WI MR of the cervical spine reveals a very small size of the posterior fossa with vermian/tonsillar extension through the foramen magnum. Also present is vertebral segmentation failure at C7 and T1, with fusion of the spinous processes.* **(Right)** *Sagittal T1WI MR in a different patient demonstrates typical vermian ectopia. Also depicted is variant dorsal cervical dysraphism with meningocele and a dermoid cyst with an associated dermal sinus tract.*

Chiari 3

KEY FACTS

TERMINOLOGY

- Chiari 3 malformation (CM3)
- Synonyms: Chiari III, rhombencephalocele

IMAGING

- Low occipital or high cervical meningoencephalocele containing cerebellum ± brainstem, meninges, vessels, CSF
- Midline bone defect within supraoccipital bone, opisthion

TOP DIFFERENTIAL DIAGNOSES

- Isolated occipital encephalocele
- Other occipital encephaloceles
  - Iniencephaly
  - Syndromic occipital encephalocele

PATHOLOGY

- Severity classified by sac contents
- Cephalocele contents: Meninges, cerebellum, brainstem ± cervical cord, occipital poles, vasculature
  - Disorganized (neuronal migration anomalies, cortical dysplasias) and gliotic brain tissue
  - Lining of sac may show gray matter heterotopias
- Associated abnormalities: Corpus callosum anomalies, gray matter heterotopia, syringohydromyelia, tethered cord

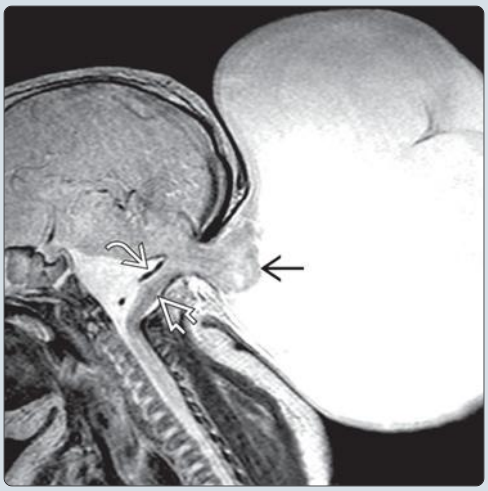
CLINICAL ISSUES

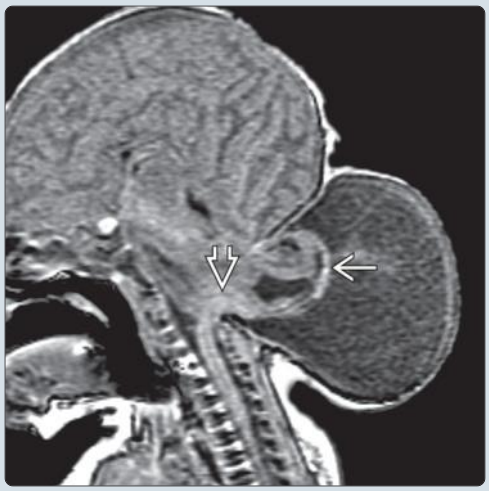
- Microcephaly, severe developmental delay, spasticity, hypotonia, seizures
- Mechanical brainstem traction, respiratory deterioration, lower cranial nerve dysfunction

DIAGNOSTIC CHECKLIST

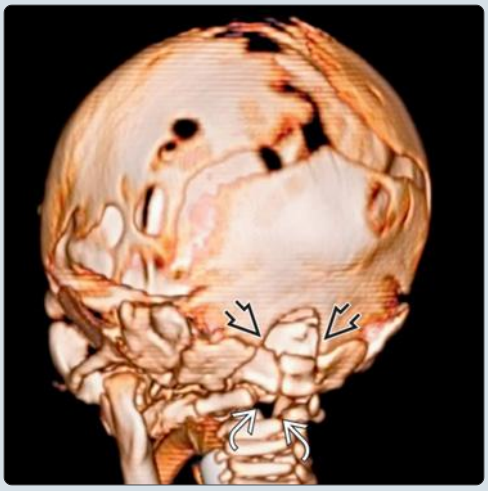
- Occipitocervical cephalocele containing cerebellum ± brainstem in conjunction with C1-C2 spina bifida = Chiari 3 malformation
- Distinct malformation; not just a Chiari 2 malformation with encephalocele

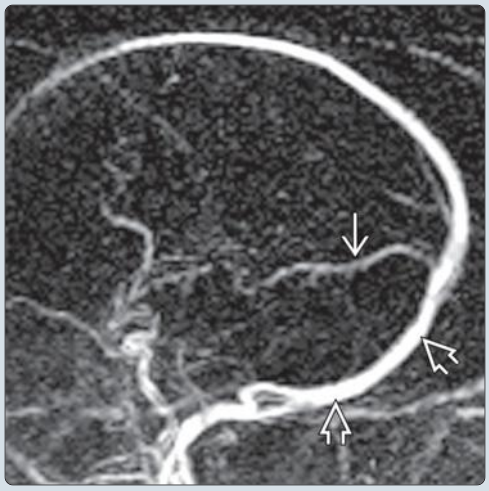
(Left) *Sagittal T2WI MR reveals a large defect in the ventral chondral portion of the supraoccipital bone and opisthion of the foramen magnum. Gliotic cerebellar tissue ⇨ protrudes into a large sac. Note displacement of the brainstem ⇨ and basilar artery ⇨.* **(Right)** *Sagittal T1WI MR shows a large meningoencephalocele composed of meninges, CSF, cerebellum ⇨, brainstem ⇨, and upper cervical spinal cord herniated through a bone defect in the lower occiput and upper cervical spine.*





(Left) *Bone CT 3D shaded surface rendering of the posterior calvarium/upper cervical spine in a Chiari 3 malformation (CM3) patient shows a large defect ⇨ of the ventral chondral and squamous supraoccipital bones, in conjunction with upper cervical spina bifida ⇨.* **(Right)** *Sagittal MRV demonstrates typical venous abnormalities of Chiari 3. The straight sinus ⇨ and vein of Galen is severely hypoplastic. Large occipital sinuses ⇨, rather than transverse sinuses, are present.*





TERMINOLOGY

Synonyms

- Chiari III, rhombencephalocele

Definitions

- Combined cephalocele + myelocele herniating through high cervical ± low occipital dysraphic defect

IMAGING

General Features

- Best diagnostic clue
  - Skin-covered upper cervical meningoencephalocele containing cerebellum

CT Findings

- NECT
  - Midline posterior cephalocele containing cerebellum
  - Small posterior cranial fossa ± scalloped clivus, lacunar skull
- Bone CT
  - Opisthion, upper cervical osseous dysraphic bone defect
- CTA
  - Basilar artery "pulled" into defect along with brainstem into cephalocele sac
  - ± veins/dural sinuses within cephalocele sac
  - Anomalous &/or ptotic veins, dural sinuses

MR Findings

- T1WI
  - High cervical cephalocele sac containing meninges and cerebellum ± brainstem, upper cervical cord
  - Cisterns, 4th ventricle, dural sinuses may extend into cephalocele (50%)
- T2WI
  - Tissues in cephalocele sac may be bright (gliosis), strand-like (necrotic), or hypointense (hemorrhagic)

Imaging Recommendations

- Best imaging tool
  - Multiplanar brain MR + MRV to characterize occipitocervical encephalocele, vessels
  - Multiplanar bone CT to evaluate osseous defects

DIFFERENTIAL DIAGNOSIS

Isolated Occipital Encephalocele

- Spares foramen magnum, lacks intracranial Chiari 2 findings

Other Occipital Encephaloceles

- Iniencephaly
- Syndromic occipital encephalocele
  - Meckel-Gruber, Goldenhar-Gorlin, MURCS (müllerian, renal, cervical spine), Walker-Warburg, amniotic band

PATHOLOGY

General Features

- Genetics
  - 677C → T mutation on methylenetetrahydrofolate reductase (*MTHFR*) gene (≤ 50%)
- Associated abnormalities
  - Corpus callosum anomalies, gray matter heterotopia, syringohydromyelia, tethered cord
  - Previously described in combination with intracranial CM2 manifestations
    - Now thought that tectum, lower brainstem findings actually reflect distortion related to cerebellar displacement into sac

Gross Pathologic & Surgical Features

- Cephalocele contents: Meninges, cerebellum, brainstem ± cervical cord, ± occipital poles, ± vasculature

Microscopic Features

- Disorganized (neuronal migration anomalies, cortical dysplasias, gliotic) brain tissue within sac

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Occipital/upper cervical cephalocele, microcephaly
  - Discovered by fetal ultrasound/MR or surprise at birth
- Other signs/symptoms
  - Mechanical brainstem traction, respiratory deterioration, lower cranial nerve dysfunction
- Clinical profile
  - Severe developmental delay, spasticity, hypotonia, seizures

Demographics

- Age
  - Newborn
- Gender
  - F > M (as in all neural tube defects [NTDs]) in most series
- Epidemiology
  - Rare; 1-4.5% of all Chiari malformations

Natural History & Prognosis

- Dependent on amount, type of herniated tissue
- Prognosis usually dismal; severe disability, early death

Treatment

- Surgical resection, encephalocele repair
  - Resect or repair sac (most structures in sac are nonfunctioning)
  - If amount of CNS tissue in sac > intracranial → not surgical candidate
- Cerebrospinal fluid diversion for hydrocephalus

DIAGNOSTIC CHECKLIST

Consider

- Chiari 3 in newborn presenting with low occipital encephalocele

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Terminology

Definitions

Morphogenetic fields

- Embryological cellular regions with potential for development into discrete anatomic structures

Gradient fields

- Describe gradually varying potential to form specific individual bones and organs within a morphogenetic field

Homeobox genes

- Gene family complexes that regulate early embryologic differentiation
- Located on separate chromosomes
- Arranged linearly in same order as expression along craniocaudal axis

Imaging Protocols

Multiplanar magnetic resonance imaging best evaluates soft tissues, neural structures, and ligaments. Multidetector axial CT with sagittal and coronal reconstruction optimally defines bone anatomy.

Embryology

The spinal axis develops following a (mostly) orderly progression of steps, with the vertebral axis and spinal cord developing synchronously. Embryological spine formation begins in the occipital region, then subsequently commences at multiple other sites simultaneously. These sites progress at different rates, and thus will be at different developmental stages at any given point in time. The cephalic spine (to about the conus level) forms by the process of primary neurulation, while the caudal spine forms separately by secondary neurulation (also called canalization and retrogressive differentiation). Most spinal anomalies may be explained by one or more events going awry during these steps.

Primary Neurulation

Embryonic spinal formation commences at the end of the second gestational week with formation of the Hensen node and at the beginning of the third week with the appearance of the neural plate during gastrulation (two germ cell layer embryo → three germ cell layer embryo). The notochordal process forms at day 16-17, with transient communication of the amnion through notochordal canal to the yolk sac through neurenteric canal of Kovalevsky. Abnormalities during this stage produce the neurenteric cyst spectrum. The neural tube folds and closes at the end of third week, leaving temporary cranial and caudal openings called neuropores. The neural tube normally closes by day 25-27, signaling the end of primary neurulation. A closed neural tube is a prerequisite for normal development of the posterior neural arch.

During primary neurulation, the neural tube separates from the overlying ectoderm during the process of dysjunction. If dysjunction occurs prematurely (premature dysjunction), perineural mesenchyme is permitted access to the neural groove and ependymal lining, differentiates into fat, and prevents complete neural tube closure, leading to the lipomatous malformation spectrum. Conversely, if dysjunction fails to occur (nondysjunction), an ectodermal-neuroectodermal tract forms that prevents mesenchymal migration, leading to focal or widespread dysraphism and the open neural tube defect spectrum, including myelomeningocele (MMC), dorsal dermal sinus, and myelocystocele. It is important to note that nondysjunction and premature dysjunction anomalies can coexist in combination simultaneously.

Premature Dysjunction

Premature dysjunction of the neural tube from the overlying ectoderm allows the perineural mesenchyme to access the neural groove and differentiate into fat, physically preventing completion of neural tube closure and resulting in a spinal lipoma, with or without closed posterior spinal dysraphism. The most commonly premature dysjunction anomalies are lipomyelomeningocele (LMMC) and intradural (juxtamedullary) lipoma. LMMC is a skin-covered, closed dysraphism anomaly where the neural placode is complexed with a lipoma contiguous with the subcutaneous fat through a posterior dysraphic defect. In all cases, the lipoma attaches to and tethers the spinal cord. LMMC accounts for 20-56% of occult spinal dysraphism and 20% of skin-covered lumbosacral masses. Spinal lipoma is subdivided into intradural (juxtamedullary, subpial) and terminal lipomas. Intradural lipomas most commonly occur near the conus. Conversely, terminal lipoma is variably considered within both the premature dysjunction and abnormal caudal cell mass differentiation spectra, with findings overlapping both mechanisms. Some authorities ("lumpers") combine all of the premature dysjunction disorders featuring lipomas into a lipomatous malformation spectrum for simplicity, recognizing that there is considerable overlap between the LMMC and lipoma malformations.

Nondysjunction

In contrast, anomalies resulting from nondysjunction related to teratogens or faulty notochord induction occur when the neural tube fails to dissociate from adjacent cutaneous ectoderm over either a focal or long interval. The least extensive variation is the dorsal dermal sinus, which occurs when a single connection remains forming a fibrous cord from skin to conus or central spinal cord canal. The dermal sinus tract usually has an atypical dimple at the ostium that is remote (> 2.5 cm) from the anus, often combined with other cutaneous anomalies. They are most common in the lumbosacral spine followed by the occipital region, and the sinus opening dermatomal level correlates with the metameric level of spinal cord attachment. All have some degree of focal dysraphism, which may be as subtle as a bifid spinous process. More extensive nondysjunction produces the open MMC lesion associated with maternal folate deficiency. In this entity, the infant presents with an open, red, weeping skin defect. Lumbosacral and thoracolumbar MMC are most common, but cervical and thoracic MMC are observed rarely. Their association with the Chiari 2 malformation is well described.

Important mimics of the dorsal dermal sinus are the low sacral (coccygeal) dimple and pilonidal sinus. The low sacral or coccygeal dimple is common, and features a low skin dimple affixed to the coccyx by a "spot weld." These are found within the intergluteal cleft, never communicate with the spinal canal, and require no treatment. A pilonidal sinus has a low ostium, does not communicate with the spinal canal, and may present later with cutaneous infection.

Secondary Neurulation (Canalization and Retrogressive Differentiation)

By day 30, neural tube development below the caudal neuropore has commenced within the undifferentiated caudal cell mass of the primitive streak, which subsequently

regresses. This process is inherently less precise than primary neurulation, which leads to more bizarre anomalies, and is characterized by the development of cysts in the caudal cell mass that coalesce into an ependymal-lined tubular structure that subsequently unites with the rostral neural tube. At day 48, a transient ventriculus terminalis appears in the future conus. If this persists after birth, it is noted incidentally as a persistent ventriculus terminalis or fifth ventricle, usually of no clinical significance. Failure of proper canalization and retrogressive differentiation leads to caudal regression spectrum, terminal myelocystocele, anterior sacral meningocele, tethered cord spectrum, or sacrococcygeal teratoma. By the third gestational month, the spinal cord extends the entire length of the developing spinal column. It is the more rapid elongation of the vertebral column and dura relative to the spinal cord that produces the apparent cord ascent during gestation. The conus should be at adult level soon after birth, and conus persistence below L2 after the first month of life in a full-term infant is probably abnormal.

Caudal cell mass anomalies are a diverse group of anomalies with a significant range of severity resulting from aberrant differentiation and retrogressive differentiation of the caudal cell mass. The etiology is postulated to be a metabolic or toxic insult to the caudal cell mass prior to the fourth gestational week. Most cases are sporadic, although a dominantly inherited defect in *HLBX9* has been recently described. 15-20% are infants of diabetic mothers, while 1% of diabetic mother offspring are afflicted. There is an association with vertebral, anal, cardiac, tracheal, esophageal, renal, and limb (VACTERL) anomalies, omphalocele, bladder extrophy, imperforate anus, and Currarino triad.

Hypogenesis or agenesis of the caudal cell mass produces the caudal regression syndrome (CRS). Two types are described: The more severe type 1 CRS shows a foreshortened terminal vertebral column with high-lying, wedge-shaped conus termination and severe associated anomalies, while the less severe type 2 CRS demonstrates a low-lying, tethered cord with milder associated malformations. In general, the higher the cord termination, the more severe the sacral anomalies. The most severe CRS presentations are lumbosacral agenesis, where the spine terminates at the lower thoracic level, and severe sacral dysgenesis with fused lower extremities in a so-called "mermaid" configuration (sirenomelia). In contrast, the mildest CRS cases manifest only a missing terminal sacral segment and are clinically asymptomatic. CRS is associated with myriad other congenital abnormalities, including renal/pulmonary hypoplasia and anorectal malformations. Other congenital spinal malformations, such as open dysraphism, segmentation and fusion anomalies, and split cord malformations, are also commonly associated, and imaging should include a search to exclude those entities as well.

A rarer presentation of caudal cell mass dysplasia than CRS is the anterior sacral meningocele (ASM), which is a large meningocele traversing an enlarged sacral foramen producing a presacral cystic mass. Most are sporadic, with a minority showing an inherited predisposition within the Currarino triad or in syndromes featuring dural dysplasia (NF1, Marfan syndrome). As with other caudal cell mass dysplasias, anorectal malformations, caudal dysgenesis, and dermoid/epidermoid tumors may be found as well.

Terminal myelocystocele is very rare, and manifests as a hydromyelic spinal cord traversing a meningocele terminating in a skin-covered myelocystocele. Associated anorectal and other visceral anomalies are common and drive the early management as well as produce much of the morbidity and mortality.

Perhaps the most common lesion in the caudal cell mass dysplasia spectrum is tethered cord syndrome (TCS). Semantically speaking, TCS refers only to patients with a low-lying cord and thickened filum and not patients with other spinal anomalies and cord abnormalities, even though those patients may look "tethered" and clinically present similarly. It is important to consider TCS as a clinical diagnosis, with imaging relegated to the role of preoperative planning rather than for establishing the primary diagnosis. On imaging, TCS manifests either as a taut spinal cord without definitive conus or low-lying conus with thickened, shortened filum and variably an associated lipomatous malformation.

Finally, if the primitive streak incompletely regresses and leaves a caudal remnant, a sacrococcygeal teratoma (SCT) may arise from the residual totipotential cell rests (Hensen node). SCTs demonstrate tissue derived from all three cell layers and contain varying proportions of mature and immature elements.

Vertebral Formation and Segmentation

Vertebral body formation occurs simultaneously with spinal cord development. During neurulation, the notochord induces the surrounding paraxial mesoderm derived from primitive streak to form paired somite blocks (myotomes, sclerotomes). The myotomes form the paraspinal muscles and skin cover, while the sclerotomes divide into medial and lateral formations to produce the vertebral body, intervertebral disc, meninges, and spinal ligaments (medial) and posterior spinal elements (lateral). Failure of correct notochordal induction leads to incomplete splitting of the neural plate from the notochord and produces the split notochord syndromes (neurenteric cyst and diastematomyelia). At day 24, a remarkable process known as sclerotomal resegmentation commences and continues until the fifth week. During sclerotomal resegmentation, a horizontal sclerotomal cleft appears in the vertebra, and the caudal half of one vertebra combines with the rostral half of the vertebra below to form a "new" vertebral body. The notochord within the vertebral body subsequently degenerates, and the intervertebral notochordal remnant becomes the intervertebral disc nucleus pulposus. Between day 40-60, the vertebral body and neural arches undergo chondrification and then subsequent ossification continuing past birth and into young adulthood. Ossification begins in the lower thoracic and upper lumbar regions and diverges cranially and caudally. In the cervical region, the vertebral primary ossification centers appear after the neural arch centers, beginning in lower cervical spine (C6, C7) and commencing rostrally. Cells of sclerotomal origin migrate dorsally to form the neural arch and condense to produce the spinous processes. The neural arches are situated intersegmentally with respect to the myotomes, so that each neural arch is associated with two successive myotomal segments. Absence of a ganglion at a vertebral level is postulated to allow unilateral arch fusion (congenital osseus bar).

Anomalies of vertebral formation and segmentation arise from aberrancies in vertebral column formation. These are generally divided into anomalies resulting from failure (partial or total) of vertebral formation (FVF) and failure to correctly segment after vertebral formation (vertebral segmentation

failure [VSF]). The abnormal vertebra may be supernumerary or replace a normal vertebral body. As a rule of thumb, the more severe the segmentation and fusion anomalies, the higher the incidence of concurrent visceral organ or other neuraxis anomalies. Abnormal *PAX1* expression is postulated to be etiological in the development of the segmentation anomalies, and other visceral and neuraxis anomalies are commonly identified as well. With FVF, the degree and location of vertebral formation failure predicts morphology; unilateral chondral center deficiency and failure of ossification produces a hemivertebra, whereas central failure of ossification centers to unite produces a "butterfly" vertebra. Conversely, vertebral segmentation failure presents with composite or "block" vertebra and posterior element fusions. Not surprisingly, block vertebra frequently coexist with FVF hemivertebra and butterfly vertebra, leading many to lump them together into the working term "segmentation and fusion anomalies" (SFAs). Many clinical syndromes prominently feature SFAs, including Klippel-Feil and Jarcho-Levin (spondylothoracic dysplasia). Therefore, SFAs are not a finding that implicates a specific disorder, but rather function more as an imaging marker for a possible syndromic process.

Spinal Cord

Spinal cord formation occurs in conjunction with vertebral formation. Within the spinal cord, the neuroepithelial cells (neuroblasts) around the central tube form the mantle layer, which produces the spinal cord gray matter. The outermost layer forms the marginal layer, which myelinates to produce the spinal cord white matter. The central neuroepithelial cells differentiate into ependymal cells along the central canal. Neural crest cells along each side of the neural groove form the dorsal root ganglia (DRG), autonomic ganglia, Schwann cells, leptomeninges, and adrenal medulla.

Notochordal Anomalies

Notochordal anomalies, primarily neurenteric cyst and diastematomyelia, produce some of the most bizarre congenital spinal anomalies. Neurenteric cysts, consisting of an intraspinal cyst lined by enteric mucosa, are most common in the thoracic spine followed by the cervical spine. They arise from a putative abnormal persistent connection between the primitive endoderm and ectoderm during the 3rd embryonic week. Whereas normally the notochord separates the ventral endoderm (foregut) and dorsal ectoderm (skin, spinal cord) during embryogenesis, in a neurenteric cyst there is failure of separation that "splits" the notochord and hinders development of mesoderm, trapping a small piece of the primitive gut within the developing spinal canal. This gut remnant may become isolated, forming a cyst, or maintain connections with the gut, skin, or both, producing the spectrum of fistulae and sinuses comprising the dorsal-enteric spinal anomalies spectrum. The most severe cases maintain communication through a primitive vertebral canal of Kovalevsky, but even mild cases usually show some kind of vertebral segmentation anomalies on close inspection.

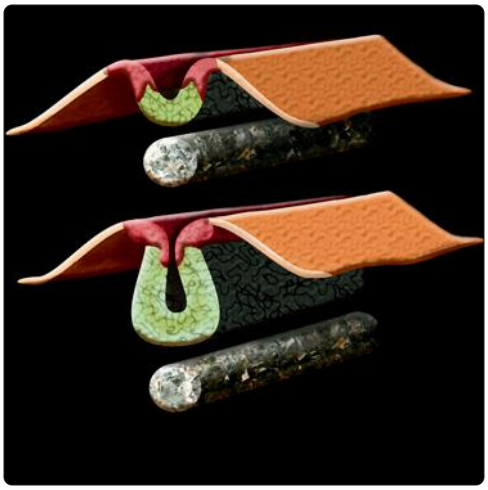
Diastematomyelia (DSM) arises from a similar aberrant process to neurenteric cyst, resulting in a splitting of the spinal cord into two hemicords, each with one ventral and one dorsal root. The hemicords may be symmetric or asymmetric (partial diastematomyelia), and one or both may feature hydromyelia or tethering. Since the notochord influences vertebral development, vertebral segmentation anomalies are very commonly associated with diastematomyelia. The presence (type 1) or absence (type 2) of an osseous or fibrous spur and whether the cords reside in separate or a single dural tube is important for preoperative planning. Occasionally, the nerve root(s) will become adherent to the dura and tether the spinal cord, producing the so-called "meningocele manqué." DSM may be isolated or found in conjunction with other spinal anomalies, particularly myelomeningocele, and it is thus critical to search for DSM prior to operative repair of a spinal anomaly or scoliosis correction.

Congenital and Developmental Anomalies of Unknown Etiology

Other uncommon but important anomalies where the etiology is not definitively known include the simple dorsal meningocele and the lateral meningocele. Dorsal meningocele by definition occurs dorsally, often in the lumbosacral spine, and strictly defined features a skin-covered meningocele devoid of neural elements protruding through a posterior dysraphic defect. In practice, however, it is not uncommon to find pieces of dysplastic nerve root or other neural tissue within the meningocele. Lateral meningoceles present as CSF-filled paraspinal masses contiguous with the thecal sac extending through a neural foramen with adjacent pedicular and foraminal osseous remodeling.

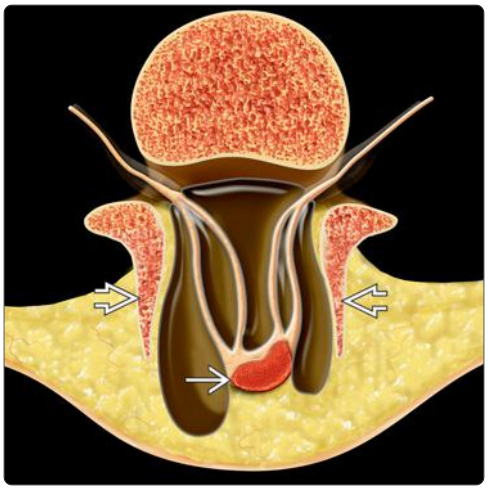
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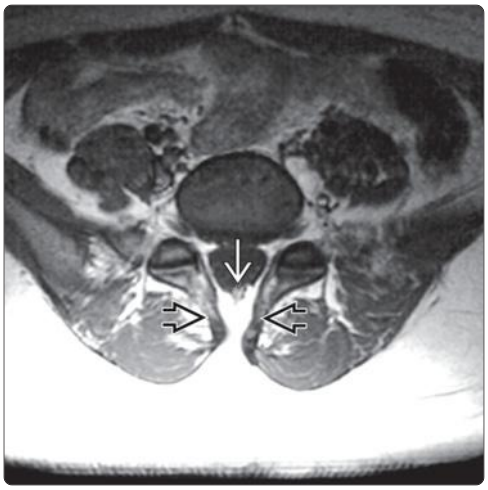
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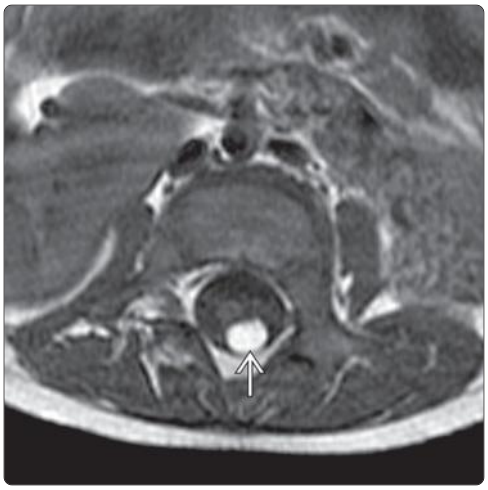
(Left) *3D graphic of neural tube embryology demonstrates normal formation of neural plate (top) and neural groove (bottom). Note the cutaneous epithelium (orange), neural crest (red), neural ectoderm (green), and notochord (gray).* **(Right)** *3D graphic of neural tube formation depicts normal neural tube closure (top) and subsequent dysjunction (bottom). Note the cutaneous epithelium (orange), neural crest (red), neural ectoderm (green), and notochord (gray).*





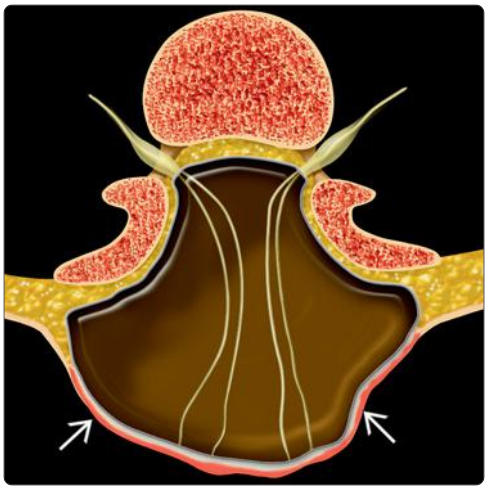
(Left) *Axial graphic illustrates premature dysjunction with a classic lipomyelomeningocele. The neural placode ⇨ is displaced through the dysraphic unfused posterior elements ⇨ into a meningocele sac.* **(Right)** *Axial T1WI MR (lipomyelomeningocele) shows the distal spinal cord ⇨ inserting into a lipoma merging imperceptibly with lumbosacral subcutaneous fat through dysraphic posterior elements ⇨.*

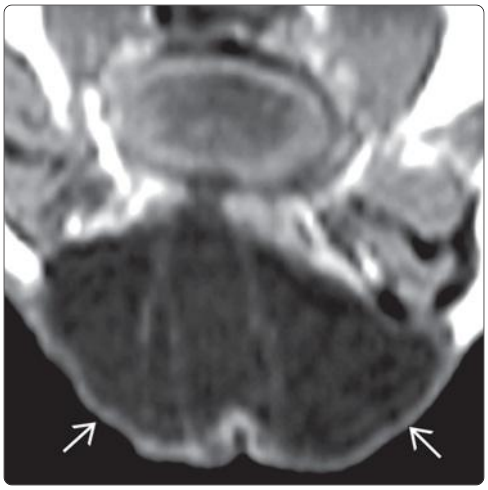




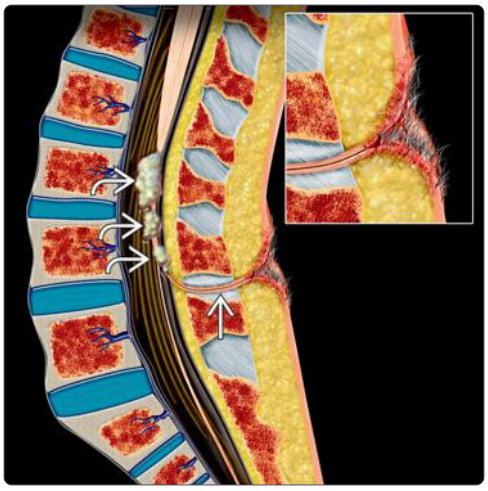
(Left) *Sagittal T1WI MR demonstrates the typical hyperintense appearance of a juxtamedullary or subpial conus lipoma ⇨. The conus is low-lying at L2/L3. These lipomas classically arise from premature dysjunction.* **(Right)** *Axial T1WI MR confirms the lipoma's ⇨ intradural location and close relationship with the dorsal conus surface. Additionally, the misshapen right kidney and empty left renal fossa was proven to represent crossed fused renal ectopia.*

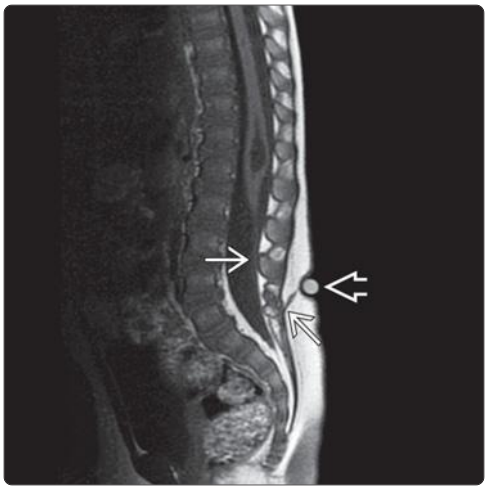
(Left) *Axial graphic (nondysjunction) shows a classic myelomeningocele, with the nerve roots traversing a large meningocele sac from the exposed neural placode ⇒, which merges into adjacent cutaneous fat and skin.* **(Right)** *Axial T1WI MR (unrepaired lumbar myelomeningocele) demonstrates nondysjunction, characterized by open dysraphism with lack of skin covering over the neural placode ⇒. The neural elements are in direct contact with the outside environment.*



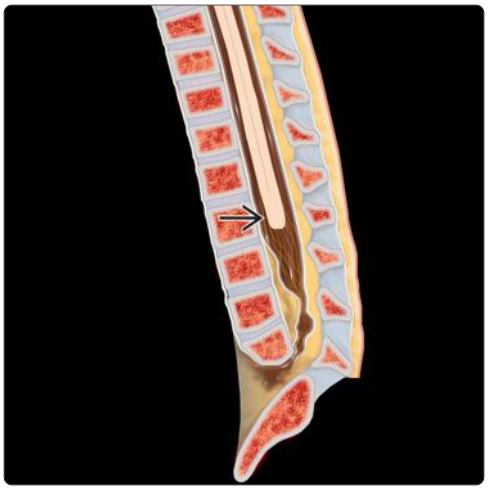


(Left) *Sagittal graphic illustrates focal nondisjunction, with a dorsal dermal cutaneous sinus tract ⇒ extending from skin surface to conus. A hairy vascular cutaneous lesion marks the sinus ostium. Note the epidermoid deposits ⇒ along the sinus tract.* **(Right)** *Sagittal T1WI MR reveals a hypointense dermal sinus ⇒ extending through the L5 posterior elements and coursing intradural to tether the low-lying conus at L2/L3. A vitamin E capsule has been placed to mark the skin opening of the sinus ⇒.*



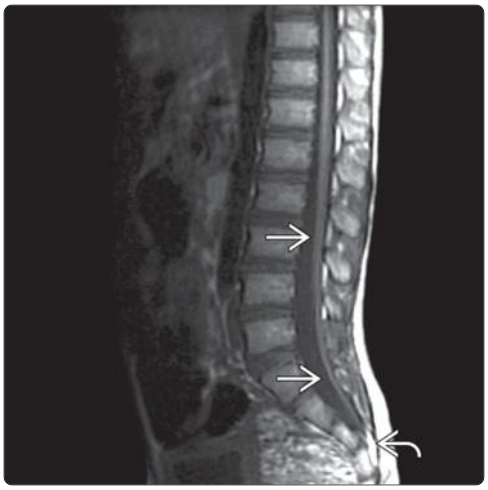


(Left) *Sagittal graphic depicts aberrant secondary neurulation resulting in severe caudal regression and vertebral column termination at the lower lumbar spine. The blunt, abnormally high conus ⇒ is characteristic of type 1 CRS.* **(Right)** *Sagittal T1WI MR (caudal regression) reveals abnormal vertebral column termination at the low lumbar level. The abnormal conus ⇒ shows a characteristic blunt wedge shape. Sacral absence permits medial migration of the iliac wings ⇒.*







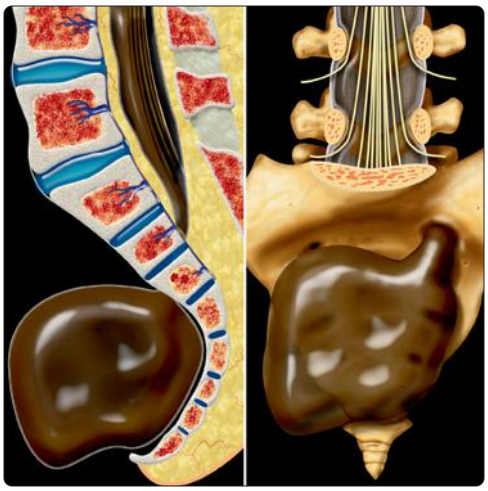


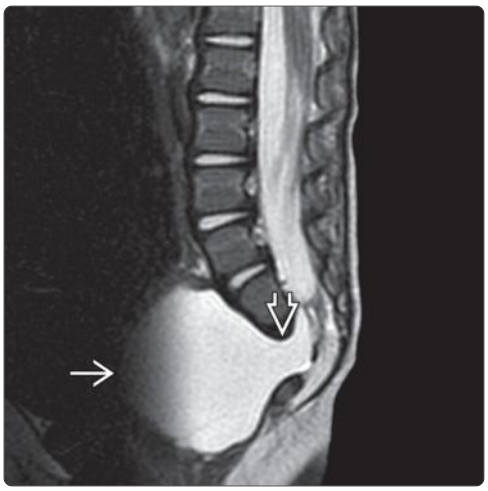
(Left) *Sagittal graphic (tethered spinal cord) demonstrates abnormal elongation of the low-lying spinal cord, which inserts into a terminal lipoma at the end of a thick, shortened fatty filum.* **(Right)** *Sagittal T1WI MR (tethered spinal cord) reveals an abnormally elongated spinal cord extending to at least L5, subtly transitioning into a thickened filum terminale that terminates in a small terminal lipoma. No syrinx or hydromyelia is present.*





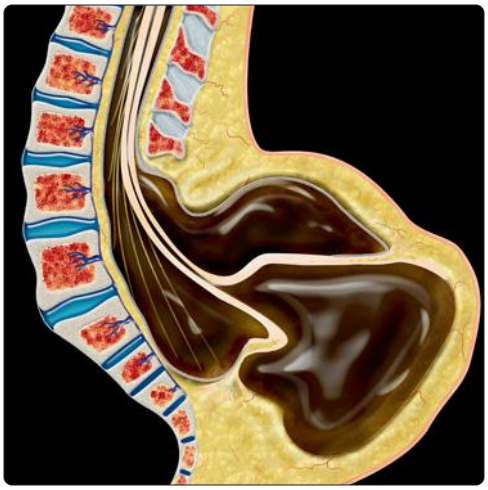
(Left) *Sagittal graphic (AAP II sacrococcygeal teratoma) shows a heterogeneous necrotic neoplasm located ventral to the sacrum. Approximately equal external and internal components classify the tumor as AAP type II. Absence of sacral invasion is characteristic.* **(Right)** *Sagittal T2WI MR (AAP II sacrococcygeal teratoma) depicts a mixed cystic and solid pelvic mass. The internal portion is predominately solid, while the external portion is more cystic.*

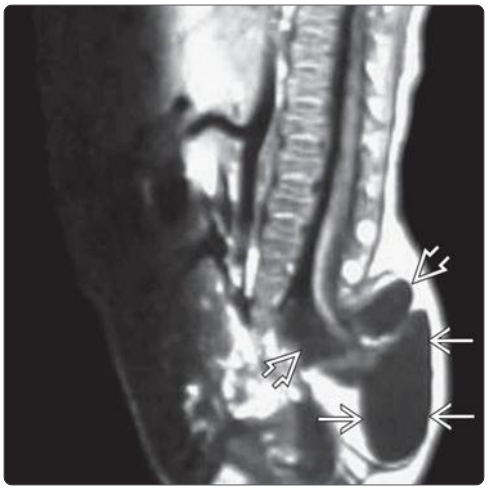




(Left) *Sagittal and coronal graphics demonstrate a classic anterior sacral meningocele. The sac enters the presacral space through an enlarged sacral neural foramina.* **(Right)** *Sagittal T2WI MR (anterior sacral meningocele) imaged parasagittal off-midline demonstrates continuity of the ventral meningocele sac with the thecal sac through an abnormally enlarged sacral foramen.*

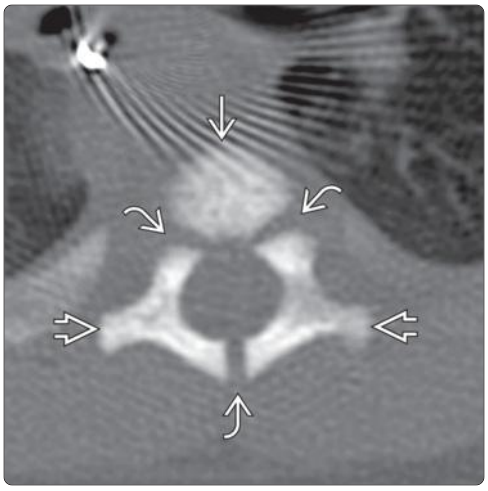
(Left) *Sagittal graphic demonstrates a typical terminal myelocystocele, with posterior spinal dysraphism and classic "cyst within a cyst" configuration.* **(Right)** *Sagittal T1WI MR shows the classic appearance of terminal myelocystocele, with a low-lying tethered spinal cord with distal hydromyelia traversing a meningocele produced by obstruction of the subarachnoid space. Also note the hypoplastic sacrum.*



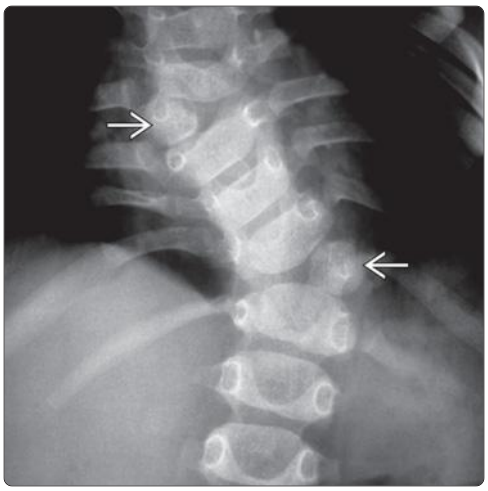


(Left) *Sagittal bone CT of the sacrum and coccyx in a 24 month old demonstrates normal ossification of the 5 sacral vertebra and first 3 coccygeal vertebra. The underlying cartilaginous model is faintly visible as soft tissue density containing the ossified centra.* **(Right)** *Axial bone CT in a 3-day-old infant demonstrates normal CT imaging appearance of the vertebral centrum and neural arch, primary ossification centers separated by cartilaginous synchondroses.*

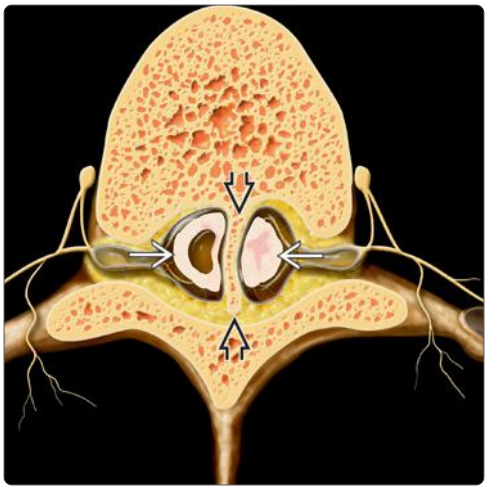


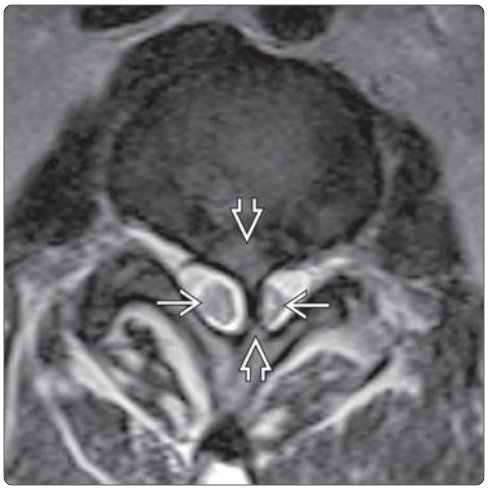


(Left) *Anteroposterior radiograph demonstrates 2 thoracic hemivertebra. These hemivertebra are on opposite sides and "cancel" to limit spinal curvature. This "balanced" configuration is less likely to show progressive curvature than "unbalanced" hemivertebra.* **(Right)** *Coronal T1WI MR shows thoracic dextroscoliosis with extensive vertebral segmentation failure. Low signal intensity between the dysplastic vertebral elements represents nonossified cartilage in this young, skeletally immature patient.*

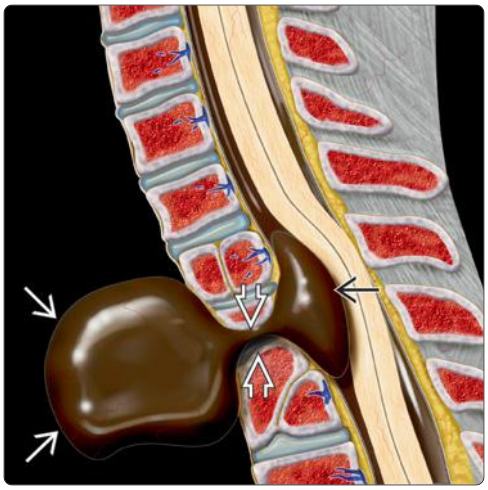


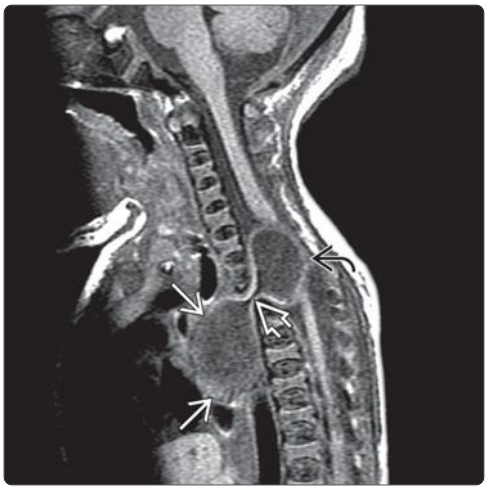




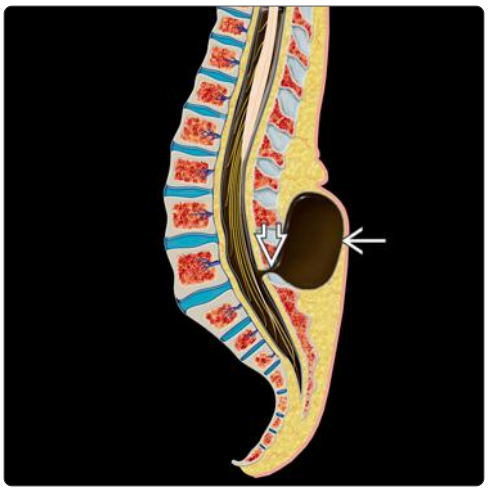


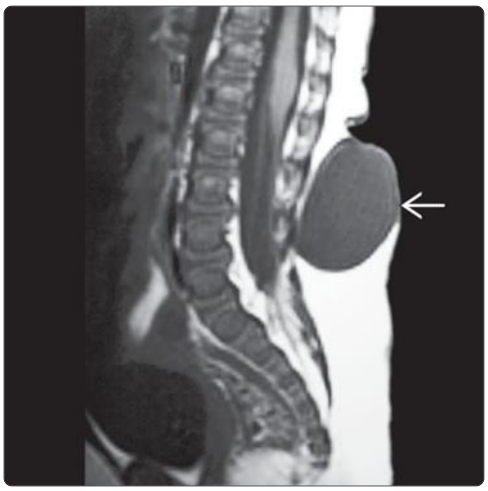
(Left) *Axial graphic demonstrates type 1 diastematomyelia, with a large osseous spur ⇨ connecting the vertebral body to posterior elements, dividing the spinal cord into 2 hemicords ➡, each within a separate dural tube. The right hemicord has a syrinx.* **(Right)** *Axial T1WI MR (split notochord syndrome) depicts classic type 1 diastematomyelia, with an ossified spur ➡ dividing the dural sac and spinal cord into 2 separate hemicords ➡, each within their own dural tube.*





(Left) *Sagittal graphic (split notochord syndrome) shows upper thoracic vertebral segmentation anomalies with a persistent canal of Kovalevsky ➡ permitting communication of the mediastinal enteric cyst ➡ with the extradural neurenteric cyst ⇨.* **(Right)** *Sagittal T1WI MR demonstrates a large, dumbbell-shaped neurenteric cyst. The mediastinal enteric cyst ➡ extends into the ventral spinal canal through the canal of Kovalevsky ➡ to produce an intracanalicular neurenteric cyst ⇨.*





(Left) *Sagittal graphic demonstrates a dorsal spinal meningocele, a malformation of unknown etiology. The meningocele sac ➡ communicates with the dural sac by a thin isthmus ➡. There are no neural elements within the meningocele sac.* **(Right)** *Sagittal T1WI MR shows the classic appearance of a dorsal spinal meningocele. The meningocele sac ➡ extends through the posterior elements via a thin isthmus, representing mild focal closed spinal dysraphism.*

Myelomeningocele

KEY FACTS

TERMINOLOGY

- Posterior spinal defect lacking skin covering → neural tissue, CSF, and meninges exposed to air
- Synonyms: Meningomyelocele (MMC), open spinal dysraphism (OSD), spina bifida aperta, spina bifida cystica

IMAGING

- Lumbosacral (44%) > thoracolumbar (32%) > lumbar (22%) > thoracic (2%)
- Preoperative: Posterior spinal defect lacking skin covering → neural tissue, CSF, and meninges exposed to air
- Postoperative: Dysraphism, low-lying cord/roots, postoperative skin closure changes

TOP DIFFERENTIAL DIAGNOSES

- Dorsal meningocele
- Closed (occult) spinal dysraphism
- Postoperative pseudomeningocele

PATHOLOGY

- Failure of neural tube closure
  - Placode may be segmental or terminal
- Association with maternal folate deficiency or abnormal folate metabolism
- Usually accompanied by multiple neurological and orthopedic complications

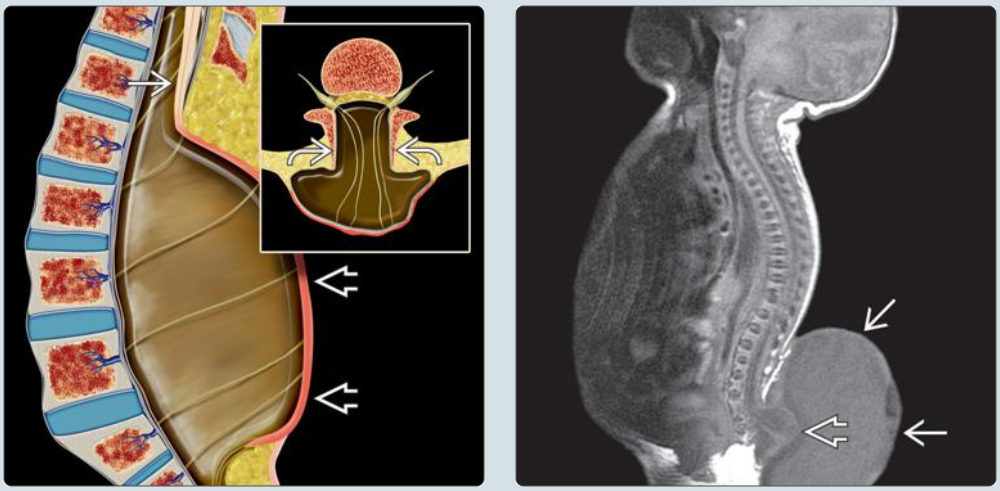
CLINICAL ISSUES

- Stable neurological deficits expected following closure
- Subsequent neurological deterioration prompts imaging evaluation for tethered cord, dural ring constriction, cord ischemia, or syringohydromyelia

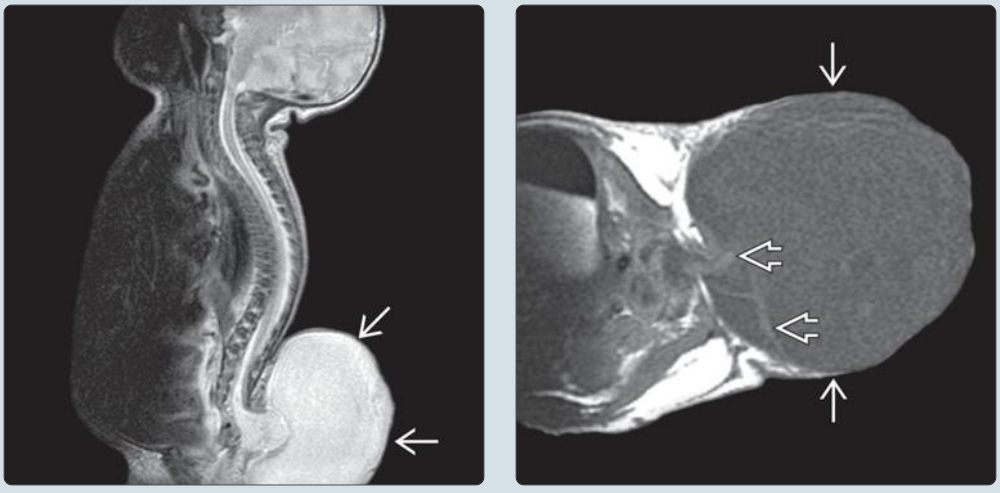
DIAGNOSTIC CHECKLIST

- Cord retethering is most common spinal cause of delayed deterioration
- Low-lying cord on MR imaging does not always equate to clinical tethering

(Left) *Sagittal graphic shows ballooning of meninges through dysraphic spinal defect, with low-lying cord ⇨ terminating in the red neural placode ⇨. Axial insert shows origin of spinal roots from ventral placode and protrusion of meninges and placode through dysraphic posterior elements ⇨.* **(Right)** *Sagittal T1WI MR shows a large unrepaired lumbosacral myelomeningocele sac ⇨ protruding through a posterior dysraphic defect. Neural elements are seen protruding into the sac ⇨.*



(Left) *Sagittal T2WI MR shows typical posterior fossa Chiari 2 malformation changes. The large myelomeningocele lumbosacral sac ⇨ has not been surgically repaired, and it protrudes dorsally through a large posterior dysraphic defect.* **(Right)** *Axial T1WI MR of unrepaired lumbosacral myelomeningocele confirms an exophytic meningeal sac ⇨ and extension of dysplastic neural elements ⇨ through the osseous spinal dysraphic defect into the myelomeningocele.*



TERMINOLOGY

Synonyms

- Meningomyelocele (MMC), open spinal dysraphism (OSD), spina bifida aperta, spina bifida cystica

Definitions

- Posterior spinal defect lacking skin covering → neural tissue, CSF, and meninges exposed to air

IMAGING

General Features

- Best diagnostic clue
  - Wide osseous dysraphism, low-lying cord/roots, postoperative skin closure changes
- Location
  - Lumbosacral (44%) > thoracolumbar (32%) > lumbar (22%) > thoracic (2%)
- Size
  - Small → large, depending on extent of neural tube defect
- Morphology
  - Exposed CSF sac + neural elements protrude through wide dorsal dysraphism

Radiographic Findings

- Radiography
  - Posterior spina bifida with wide eversion of lamina
  - Most rostral normal lamina = superior margin of dysraphic defect

CT Findings

- NECT
  - Wide posterior osseous dysraphism, skin-covered CSF sac (postoperative)
  - Associated anomalies, postoperative complications
    - Spine CT: Diastematomyelia spur, dural constriction, or cord ischemia sequelae (abrupt cord termination)
    - Head CT: Hydrocephalus from VP shunt failure

MR Findings

- T1WI
  - Wide spinal dysraphism, flared laminae, low-lying cord/roots; skin-covered CSF sac (postoperative)
  - Loss of normal posterior epidural fat segmentation at anomaly level (sagittal imaging)
    - Epidural fat contiguous on 2 or more adjacent levels → suspicious for dysraphism
- T2WI
  - Nerve roots originate from ventral placode surface; ventral roots exit medial to dorsal roots

Ultrasonographic Findings

- Grayscale ultrasound
  - Obstetrical ultrasound → antenatal diagnosis
    - Open neural arch, flared laminae, protruding myelomeningocele sac, and Chiari 2 brain findings ("lemon" sign, "banana" sign, hydrocephalus)

Nonvascular Interventions

- Myelography
  - ± CSF loculations, absence of CSF between placode and dura, low-lying cord

Other Modality Findings

- Sagittal cine phase contrast MR → diminished conus pulsation may indicate tethering

Imaging Recommendations

- Best imaging tool
  - MR imaging
- Protocol advice
  - Obstetrical ultrasound: Initial MMC diagnosis, delivery planning (cesarean section), triaging for possible fetal surgery
  - Head CT: Hydrocephalus evaluation
  - MR: Sagittal and axial T1WI and T2WI; must include entire sacrum

DIFFERENTIAL DIAGNOSIS

Dorsal Meningocele

- Meninges protrude through dorsal dysraphism into subcutaneous fat
- Skin-covered, usually does not contain neural elements

Closed (Occult) Spinal Dysraphism

- Dorsal osseous dysraphism; cord may be low-lying
- Skin or other cutaneous derivative (e.g., lipoma) covers neural elements

Postoperative Pseudomeningocele

- History, clinical exam permit distinction
- Look for surgical laminectomy defect, absence of spina bifida osseous changes

PATHOLOGY

General Features

- Etiology
  - Lack of complex carbohydrate molecule expression on neuroectodermal cell surface → failed neural tube closure
  - Neural tube defect (NTD) deficits worsened by chronic mechanical injury, amniotic fluid chemical trauma
- Genetics
  - *Pax3* paired box gene derangements
  - Methylene-tetra-hydrofolate-reductase (MTHFR) mutations associated with abnormal folate metabolism
    - MTHFR mutations + folate deficiency → increased risk of NTD
  - Trisomy 13, 18 (14% of NTD fetuses)
- Associated abnormalities
  - Kyphoscoliosis: Neuromuscular imbalance ± vertebral segmentation anomalies
    - Developmental (65%): Neuromuscular imbalance, anterior displacement of spinal extensor muscles
    - Congenital (30%): Congenital osseous anomalies (hemivertebrae, bony bar)
  - Diastematomyelia, dermal sinus (31-46%)
    - Cord split above (31%), below (25%), or at (22%) MMC level
    - Hemimyelocele variant (10%) → asymmetric deficits
  - Syrinx (30-75%)

- Chiari 2 malformation (≈ 100%), hydrocephalus requiring shunting (80%)
- Orthopedic abnormalities (80%); 2° to muscular imbalance

- Dysjunction = normal neural tube separation from overlying ectoderm during neural tube closure
- Open NTDs arise from dysjunction failure → neural placode
  - Anterior placode surface → external pia mater cord surface
  - Posterior placode surface → internal neural tube ependyma
- Placode may be segmental or terminal
  - Segmental (lumbar, thoracolumbar, thoracic) → spinal cord continues distally
  - Terminal (lumbosacral, sacral) → placode at end of spinal cord

Gross Pathologic & Surgical Features

- Red, exposed neural placode leaking CSF protrudes through osseous midline defect
- Spinal cord always physically tethered; ± clinical neurologic deterioration

Microscopic Features

- Purkinje cell loss, sclerosis of herniated posterior fossa tissues related to Chiari 2 malformation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Stable neurological deficits expected following closure
    - Neurological deterioration → imaging evaluation for tethered cord, dural ring constriction, cord ischemia, or syringohydromyelia
- Clinical profile
  - Newborn: Midline raw, red, exposed neural placode
    - Lesion level determines severity of neurological deficits
    - Hydrocephalus 2° to Chiari 2 malformation
  - Postoperative: Fixed paraparesis and sensory deficits concomitant with defect level, large heads (hydrocephalus), neurologically-induced orthopedic disorders, neurogenic bladder (90%), ± kyphoscoliosis
- Laboratory findings: Elevated maternal serum alpha fetoprotein (AFP)

Demographics

- Age
  - Always present at birth
- Gender
  - M < F = 1:3
- Ethnicity
  - ↑ frequency in Irish/Welsh populations (4-8x), families with other affected children (7-15x)
- Epidemiology
  - Incidence 0.17-0.6/1,000 live births
  - Maternal folate deficiency, obesity, antiepileptic therapy (lowers folate bioavailability)
  - Declining prevalence of myelomeningocele worldwide attributed to folic acid fortification, prenatal diagnosis with termination of affected fetuses, and other unknown factors

Natural History & Prognosis

- Stable postoperative deficit expected → best possible outcome
  - Hydrocephalus and tethered cord determine prognosis for deterioration
  - Neurological deterioration suggests complication
    - Tethering by scar or 2nd (unrecognized) malformation → most common
    - Constricting postoperative dural ring
    - Cord compression by epidermoid/dermoid tumor or arachnoid cyst
    - Cord ischemia
    - Syringohydromyelia (29-77%)
- Chiari 2 malformation most common cause of death in MMC patients

Treatment

- Folate supplementation to pregnant/conceiving women
- MMC closure < 48 hours to stabilize neural deficits, prevent infection
  - Some tertiary centers perform in utero surgical repair; may lessen Chiari 2, neurological deficit severity, and need for ventriculoperitoneal shunting
  - Currently evidence inconclusive that prenatal repair of MMC improves future neurologic function
- Subsequent management revolves around treating postoperative complications
  - Cord untethering, hydrocephalus management, treatment of kyphoscoliosis

DIAGNOSTIC CHECKLIST

Consider

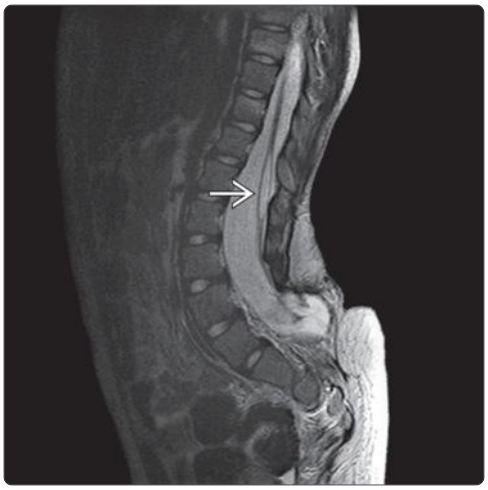
- Imaging untreated MMC rarely indicated
- MMC patients frequently have other CNS abnormalities; neurological deterioration requires assessment of entire craniospinal axis
- Cord retethering = most common spinal cause of delayed deterioration

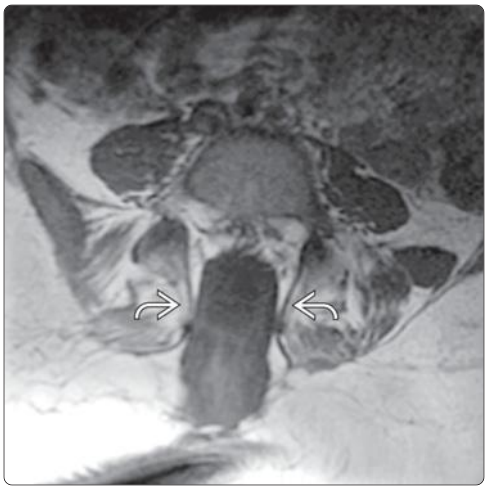
Image Interpretation Pearls

- Low-lying cord on MR imaging does not always equate to clinical tethering

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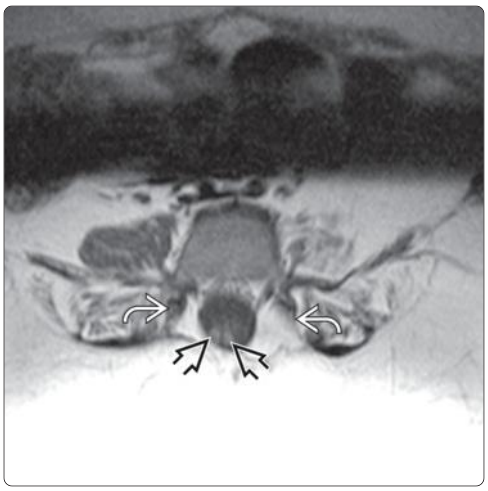
(Left) *Sagittal T2WI MR shows the typical appearance of patulous distal thecal sac and posterior soft tissues after myelomeningocele closure. The attenuated elongated spinal cord typically inserts into the surgical closure, with detection of small terminal syrinx.* **(Right)** *Axial T1WI MR (repaired myelomeningocele) shows the expected postoperative appearance at the surgical closure site. Note parallel orientation of the lamina, characteristic of spinal dysraphism.*





(Left) *Axial bone CT (surgically-repaired myelomeningocele) shows posterior spinal dysraphism, with characteristic parallel orientation of the dysraphic unfused lamina and spinous processes.* **(Right)** *Axial T2WI MR (repaired myelomeningocele) obtained to evaluate delayed neurological deterioration after a period of static neurological deficit shows splitting of the thoracic spinal cord into 2 hemicords, reflecting previously undiagnosed type 2 diastematomyelia.*





(Left) *Sagittal T1WI (repaired segmental lumbar myelomeningocele) demonstrates an elongated, attenuated spinal cord with segmental adhesion to the myelomeningocele closure site. The spinal cord continues past the neural placode to terminate in a small distal terminal lipoma.* **(Right)** *Axial T1WI MR (repaired segmental lumbar myelomeningocele) shows widely dysraphic posterior elements but also reveals a previously undiagnosed type 2 diastematomyelia with 2 hemicords.*

Lipomyelomeningocele

KEY FACTS

TERMINOLOGY

- Lipomyelomeningocele (LMMC), lipomyelocele (LMC), spinal lipomatous malformation

IMAGING

- Subcutaneous fatty mass contiguous with neural placode/lipoma through posterior dysraphism
  - Size varies from nearly imperceptible to large
- Posterior spinal dysraphism, enlarged canal at placode level
- Tethered, low-lying spinal cord ± meningocele inserts into lipoma through dysraphic defect

TOP DIFFERENTIAL DIAGNOSES

- Terminal lipoma
- Intradural (juxtamedullary) lipoma
- Dorsal meningocele
- Myelocele/myelomeningocele

PATHOLOGY

- Premature disjunction of neural ectoderm from cutaneous ectoderm → induction of mesenchyme to form fat (lipoma)
- Spinal cord always tethered
- Association with vertebral segmentation anomalies, sacral dysgenesis, anorectal and genitourinary abnormalities

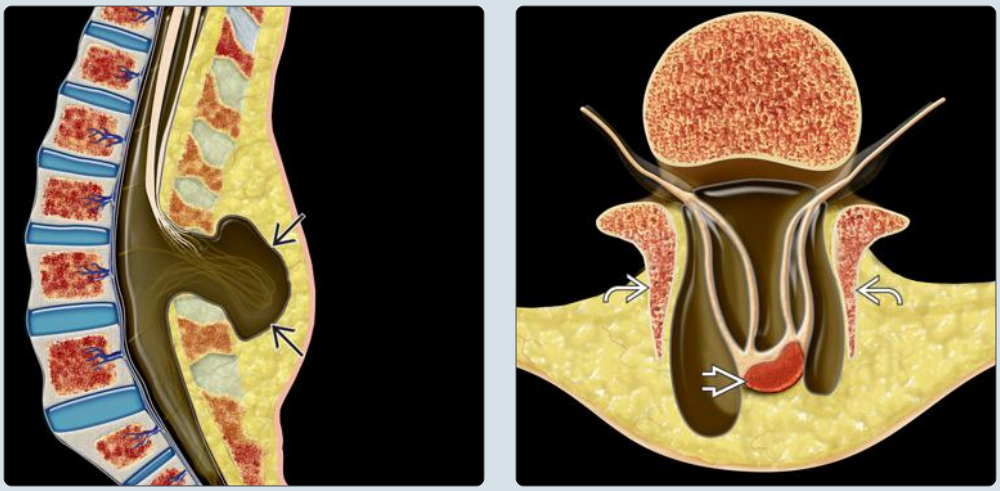
CLINICAL ISSUES

- Soft midline or paramedian skin-covered mass above buttocks
- Back/leg pain, scoliosis, lower extremity paraparesis, bladder/bowel dysfunction

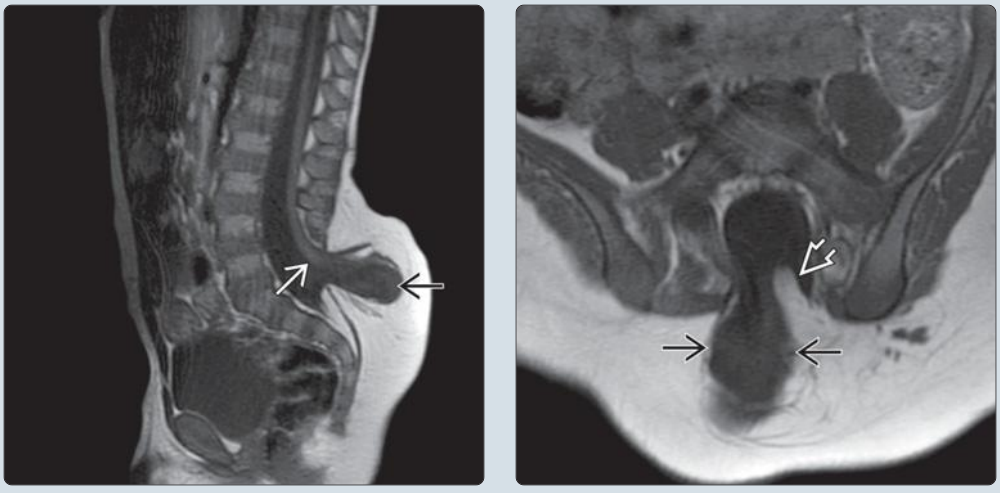
DIAGNOSTIC CHECKLIST

- Diagnosis of postoperative retethering primarily clinical; use imaging to search for complications

(Left) *Sagittal graphic of the lumbosacral spine demonstrates classic lipomyelomeningocele anatomy. The low-lying spinal cord and cauda equina nerve roots protrude through the dysraphic posterior elements into a dorsal meningocele sac ⇨ covered by skin and subcutaneous fat.* **(Right)** *Axial graphic of the lower lumbar spine shows protrusion of the nerve roots and neural placode ⇨ through the dysraphic posterior elements ⇨ into a skin-covered sac.*



(Left) *Sagittal T1WI MR shows the low-lying tethered spinal cord ⇨ extending through a dorsal dysraphic defect into the subcutaneous fat to terminate in a skin-covered cyst ⇨.* **(Right)** *Axial T1WI MR demonstrates protrusion of the lipomyelomeningocele sac ⇨ with eccentric lipoma ⇨ through the parallel-oriented dysraphic posterior elements into the subcutaneous fat (clinically evident as a skin-covered fatty mass).*



TERMINOLOGY

Abbreviations

- Lipomyelomeningocele (LMMC)

Synonyms

- Lipomyelocele (LMC), lipomyeloschisis, lipomatous malformation

IMAGING

General Features

- Best diagnostic clue
  - Subcutaneous fatty mass contiguous with neural placode/lipoma through posterior dysraphism
- Location
  - Lumbosacral
- Size
  - Subcutaneous mass varies from nearly imperceptible to large
- Morphology
  - Tethered low-lying spinal cord inserts into lipoma through dysraphic defect

Radiographic Findings

- Radiography
  - Multilevel dorsal spinal dysraphism ± hypodense soft tissue mass

CT Findings

- NECT
  - Ventral neural placode/tethered cord ± terminal hydromyelia, cord myeloschisis
  - Hypodense dorsal lipoma is contiguous with subcutaneous fat through posterior spina bifida
- Bone CT
  - Multilevel dysraphism, enlarged canal at placode level

MR Findings

- T1WI
  - Hyperintense lipoma contiguous with subcutaneous fat, tethered cord/placode
    - Herniation of placode-lipoma complex immediately inferior to last intact lamina above dorsal defect
    - Lipoma may be rotated (40%)
    - ± intramedullary, intradural, or extradural lipoma; actually occult extension from LMMC rather than discrete 2nd lesion
  - Cord tethering ± myeloschisis, terminal hydromyelia
- T2WI
  - Hyperintense lipoma; neural elements isointense on background of hyperintense CSF
- STIR
  - Lipoma → hypointense, confirming fat composition

Ultrasonographic Findings

- Grayscale ultrasound
  - Echogenic intradural spinal mass contiguous with tethered cord through dorsal dysraphism

Nonvascular Interventions

- Myelography
  - Dorsal dysraphism + dilated dural sac; low-lying conus inserts into hypodense lipoma

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - STIR or chemical fat-saturated techniques confirm fat composition

DIFFERENTIAL DIAGNOSIS

Terminal Lipoma

- Lipoma tethers low-lying conus, stretching cord; merges through sacral spina bifida to join subcutaneous fat
- No spinal canal enlargement
- Rotation of lipoma, vertebral segmentation, and fusion anomalies (SFA) rare

Intradural (Juxtamedullary) Lipoma

- Enclosed by intact dura; cutaneous manifestations unusual
- Posterior elements not widely dysraphic
- Cervical, thoracic spine most common

Dorsal Meningocele

- Skin covered, no lipomatous elements
- More limited dysraphic defect

Myelocele/Myelomeningocele

- Clinically obvious open dysraphism; no skin or subcutaneous fat covering, no lipoma

PATHOLOGY

General Features

- Etiology
  - Neural tube normally forms by infolding and closure of neural ectoderm as it separates from cutaneous ectoderm during 3rd and 4th week → neurulation and dysjunction
    - In LMMC, premature disjunction of neural ectoderm from cutaneous ectoderm → mesenchymal tissue direct access to incompletely closed neural tube
    - Mesenchyme incorporated between neural folds
    - Neural folds remain open, forming neural placode at site of premature disjunction
    - Ependymal lining of primitive neural tube induces mesenchyme to form fat → lipoma
- Associated abnormalities
  - ± intramedullary, intradural, or extradural lipoma → occult extension of LMMC
  - Butterfly vertebrae, hemivertebrae, fused vertebrae (≤ 43%)
  - Sacral abnormalities (≤ 50%); confluent foramina and partial sacral dysgenesis
  - Anorectal and GU abnormalities (5-10%); if concurrent sacral anomalies → 90%
  - Terminal diastematomyelia (≤ 10%)
  - (Epi)dermoid tumor, dermal sinus, angioma, arachnoid cyst (rare)
  - Arteriovenous malformation (very rare)

- LMC and LMMC are analogous to myelocele and myelomeningocele, respectively, except lipoma is attached to dorsal placode surface and intact skin overlies lesion
- Spinal cord always tethered

Staging, Grading, & Classification

- Lipomyelocele = neural placode-lipoma complex contiguous with subcutaneous fat through dysraphic defect, attaching to and tethering spinal cord
- Lipomyelomeningocele = lipomyelocele + meningocele, enlargement of subarachnoid space with displacement of neural placode outside of spinal canal
- Newly proposed spinal lipomatous malformation classification divides spinal lipomas into 2 groups based on presence or absence of dural defect
  - Addresses pertinent differences in embryology, clinical presentation, and prognosis

Gross Pathologic & Surgical Features

- Canal enlarged at placode, spina bifida level
- Lipoma may envelop both dorsal and ventral nerve roots, dorsal nerve roots only, or filum terminale/conus
- Dura deficient at spina bifida zone → does not close neural tube
  - Attached laterally to neural placode, posterior to dorsal nerve roots as they emerge from cord
- Lipoma may be asymmetric (40%), rotating placode and elongating roots on 1 side, shortening on other
  - Predisposes to operative injury, incomplete tether release

Microscopic Features

- Dorsal placode surface adjacent to lipoma has no ependymal lining; covered by connective tissue mixed with islands of glial cells, smooth muscle fibers

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back/leg pain, scoliosis, lower extremity paraparesis, sacral sensory loss, bladder/bowel dysfunction
- Other signs/symptoms
  - Limb atrophy, orthopedic foot deformity
- Clinical profile
  - Soft midline or paramedian skin-covered mass above buttocks
    - Lumbosacral mass: Clinical attention usually within 6 months
    - No mass: Present when neurological deficits manifest (5 years → adulthood)
  - Cutaneous stigmata (50%); hemangioma, dimple, dermal sinus, skin tag, hairy patch

Demographics

- Age
  - Infancy (most common) → adulthood
- Gender
  - M < F
- Epidemiology
  - 20-56% of occult spinal dysraphism; 20% of skin-covered lumbosacral masses
  - Incidence not impacted by folate supplementation to pregnant women (unlike MMC)

Natural History & Prognosis

- Potentially irreversible progressive neurological impairment (cord tethering, enlarging lipoma)
  - Lipoma grows with infant
  - Bladder dysfunction usually persists if not operated early
- ≤ 45% of children neurologically normal at diagnosis
  - 16-88% develop neurological symptoms if left untreated
  - Most symptomatic patients progress if untreated
- Neurologically intact patients at surgery usually remain intact on long-term follow-up
- Postoperative exam should not deteriorate with longitudinal growth; neurological decline raises suspicion of retethering, warrants repeat imaging
- Symptomatic retethering common; weeks to years after initial surgery
  - Median time between initial procedure and reoperation for retethering → 52 months

Treatment

- Timing of intervention currently under reconsideration
  - Most advocate early prophylactic surgery (< 1 year of age) to untether cord, resect lipoma, and reconstruct dura
  - Some advocate more conservative management, delaying 1st operation until symptomatic

DIAGNOSTIC CHECKLIST

Consider

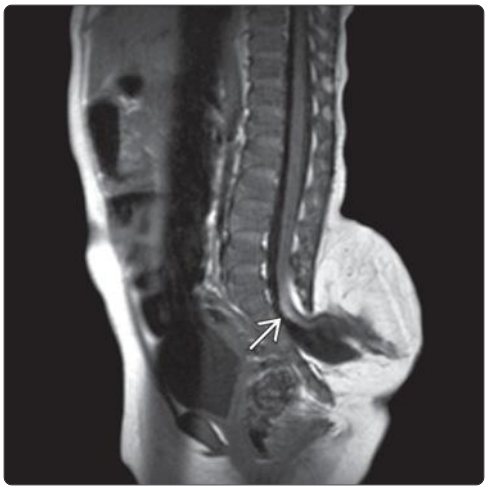
- Presence/absence of placode rotation important preoperative information for surgeon

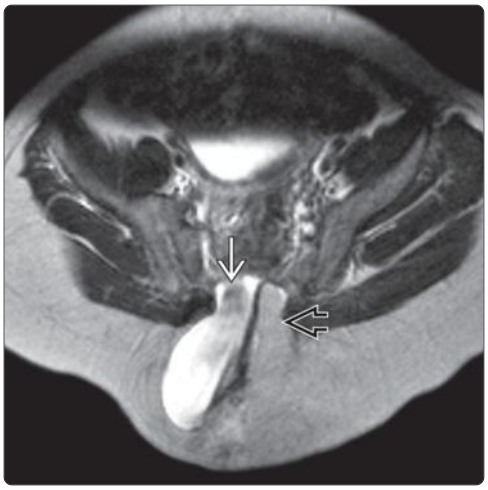
Image Interpretation Pearls

- Determination of retethering following surgery primarily clinical diagnosis; use imaging to search for complications

SELECTED REFERENCES

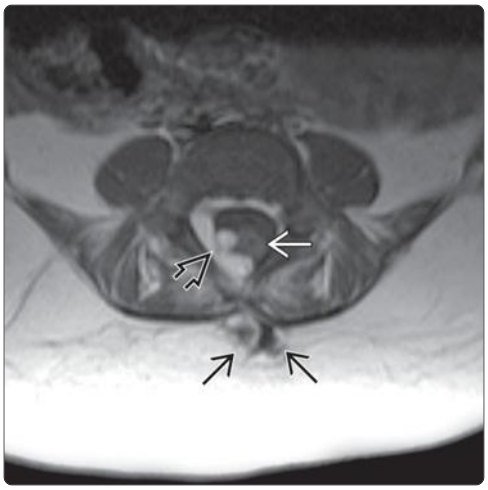
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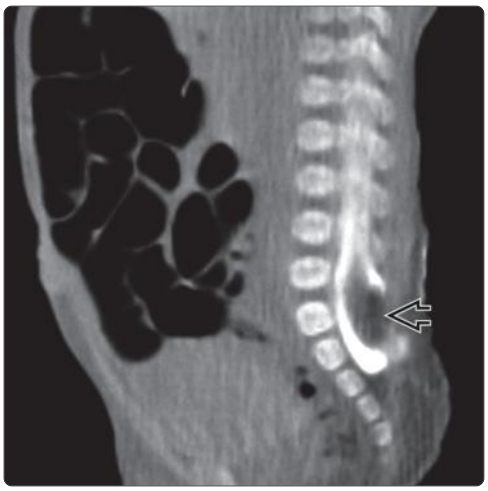
(Left) *Sagittal T1WI MR demonstrates low-lying spinal cord ➡ inserting into a large lipomatous malformation that is contiguous with subcutaneous fat through a posterior dysraphic defect. The most inferior level of complete posterior elements is at the S1 level.* **(Right)** *Axial T2WI MR at the upper sacral level demonstrates the relationship of the low-lying cord/placode ➡ with the lipoma ⇨ component of the lipomyelomeningocele. Chemical shift artifact in the lipoma confirms fat content.*





(Left) *Sagittal T1WI MR demonstrates low-lying spinal cord placode ➡ inserting directly into a lumbosacral lipomatous mass ⇨ that extends into contiguous large subcutaneous lipoma. The distal cord also demonstrates syringohydromyelia ⇨.* **(Right)** *Axial T1WI MR depicts the insertion of the distal cord ➡ into the rotated neural placode and lipoma ⇨. The lipomyelomeningocele protrudes through the posterior dysraphic defect into the subcutaneous lipomatous mass ⇨.*





(Left) *Anteroposterior myelography depicts splitting ("myeloschisis") of the low-lying spinal cord ➡ within the dilated distal dural sac.* **(Right)** *Sagittal NECT following intrathecal myelographic contrast injection shows a dilated dural sac and low-lying cord terminating in a large hypodense lipoma ⇨. Subcutaneous fat is contiguous with the lipoma through the dysraphic posterior elements.*

Lipoma

KEY FACTS

TERMINOLOGY

- Intradural (juxtamedullary, subpial) or terminal lipoma

IMAGING

- Lipoma intimately associated with spinal cord (intradural) or distal cord/filum insertion (terminal)
- Lipoma follows fat signal intensity, density, and echogenicity

TOP DIFFERENTIAL DIAGNOSES

- Lipomyelocele/lipomyelomeningocele
- Filum fibrolipoma
- Dermoid cyst

PATHOLOGY

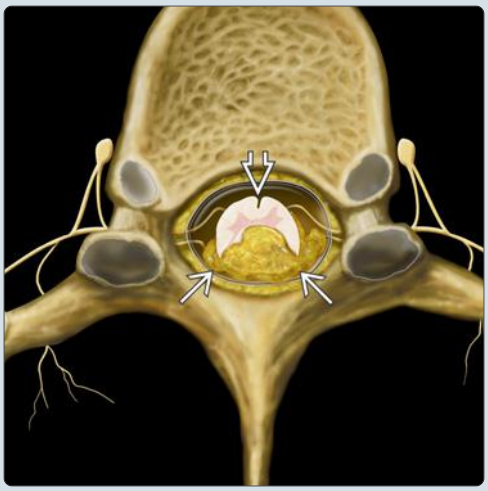
- Follows premature separation (premature disjunction) of cutaneous ectoderm from neuroectoderm during neurulation
  - Surrounding mesenchyme enters ependyma-lined central spinal canal, impedes neural tube closure open placode
  - Mesenchyme differentiates into fat
- Skin closed over malformation (closed neural tube defect)

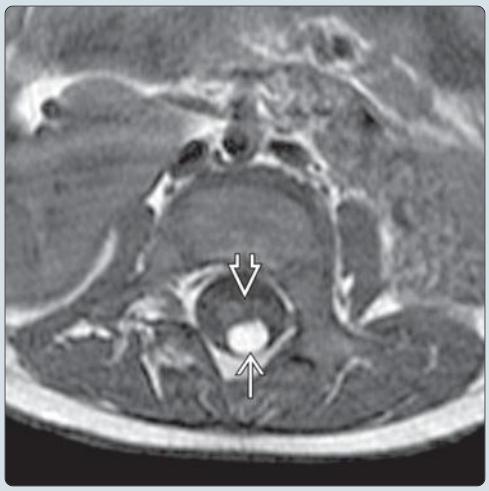
CLINICAL ISSUES

- Symptoms referable to lipoma level, presence of spinal cord compression
- Small lipomas may grow dramatically during infancy

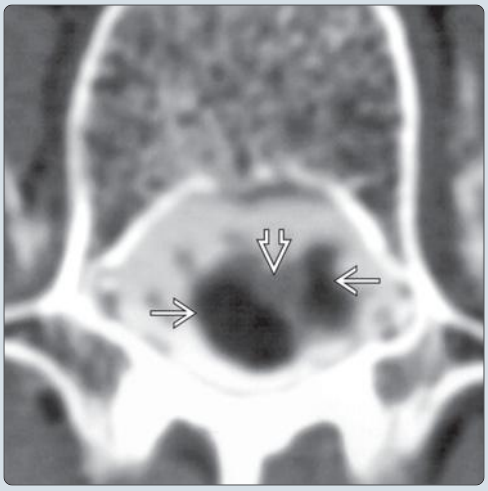
DIAGNOSTIC CHECKLIST

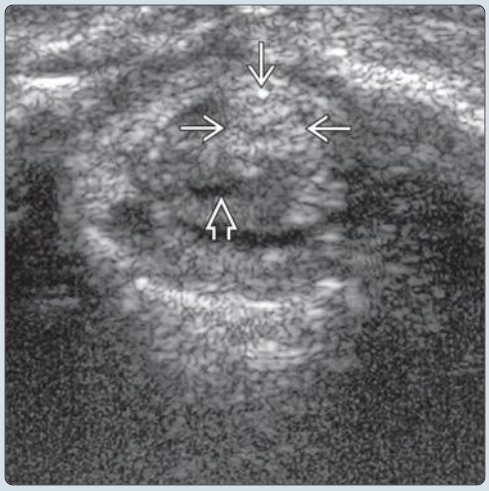
- Profound hypodensity on CT myelography and T1WI hyperintensity distinctively characteristic of lipoma
- Use chemical fat-saturation or inversion recovery MR technique to confirm fat content





(Left) *Axial graphic of the thoracic spine demonstrates incomplete closure of the dorsal spinal cord ⇨ around a dorsal juxtamedullary conus lipoma ⇨, encompassing the dorsal spinal nerve roots.* **(Right)** *Axial T1WI MR reveals the typical hyperintense appearance of a conus juxtamedullary (subpial) lipoma ⇨. Note the intradural location and close relationship with the dorsal conus ⇨ surface.*





(Left) *Axial NECT following intrathecal myelography reveals a typical markedly hypodense dorsal juxtamedullary (subpial) lipoma ⇨. The lipoma encircles and distorts the spinal cord conus tip ⇨. Note mild scalloping of the posterior vertebral body.* **(Right)** *Transverse grayscale real-time sonography demonstrates a hyperechoic intradural juxtamedullary lipoma ⇨ located within the dural sac. The lipoma is contiguous with the dorsal conus tip ⇨.*

TERMINOLOGY

Synonyms

- Intradural (juxtamedullary, subpial) or terminal lipoma

Definitions

- Spinal lipoma intimately associated with spinal cord (intradural) or distal cord/filum insertion (terminal)

IMAGING

General Features

- Best diagnostic clue
  - Hyperintense (T1WI) intradural mass
- Location
  - Intradural
    - Thoracic (30%) > cervicothoracic (24%) > cervical (12%) > lumbosacral spine
    - Dorsal (73%) > lateral/anterolateral (25%) > anterior (2%)
  - Terminal
    - Lumbosacral
- Size
  - Range: Tiny → huge
- Morphology
  - Wide spectrum of lesions ranging from simple filum lipomas → complex malformations

Radiographic Findings

- Radiography
  - Intradural lipoma
    - Hypodense mass ± dysraphism; posterior elements generally intact but show focal canal widening 2° to bony erosion
  - Terminal lipoma
    - Hypodense mass ± posterior dysraphism

CT Findings

- NECT
  - Intradural lipoma: Focal lobulated hypodense intradural mass ± central canal, neural foraminal widening at lipoma level
  - Terminal lipoma: Elongated hypodense mass at filum termination; may extend into subcutaneous fat through dysraphic defect

MR Findings

- T1WI
  - Intradural lipoma
    - Lobulated ovoid/rounded intradural hyperintense mass intimately associated with cord
    - ± canal widening, local dysraphism
    - ↓ signal intensity on fat-saturation sequences
  - Terminal lipoma
    - Hyperintense mass attached to distal cord/filum; extends through lumbosacral dysraphism → subcutaneous fat
    - Thin, "stretched" cord usually tethered ± syrinx
- T2WI
  - Similar signal intensity, imaging appearance to T1WI
  - ± spinal cord compression (intradural) → hyperintense cord signal
- STIR
  - ↓ signal intensity confirms fat
- T1WI C+
  - No lipoma enhancement
- MR cine
  - Decreased conus motion → tethered cord

Ultrasonographic Findings

- Grayscale ultrasound
  - Echogenic intraspinal mass ± reduced conus motion

Nonvascular Interventions

- Myelography
  - Intradural or terminal hypodense mass partially surrounded by hyperdense contrast
  - Large tumors may produce spinal block

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging
- Protocol advice
  - Ultrasound in infants for screening; confirm with MR if positive
  - Sagittal, axial T1WI to define lipoma(s) extent and relationship to neural placode, adjacent tissues

DIFFERENTIAL DIAGNOSIS

Lipomyelocele/Lipomyelomeningocele

- Skin-covered (closed) neural placode-lipoma complex contiguous with subcutaneous fat through dysraphic defect
- Mass often palpable ± cutaneous stigmata

Filum Fibrolipoma

- Common; 4-6% population, mostly asymptomatic but symptomatic patients present with "tethered cord"
- Hyperintense/hypodense mass in filum ± tethering, low-lying conus

Dermoid Cyst

- Mixed density/signal intensity mass; lack of homogeneous hyperintensity ± dermal sinus help to distinguish

PATHOLOGY

General Features

- Etiology
  - Arise from premature separation (disjunction) of cutaneous ectoderm from neuroectoderm during neurulation
    - Surrounding mesenchyme enters ependyma-lined central spinal canal, impeding closure of neural folds → open placode
    - Mesenchyme differentiates into fat
    - Similar mechanism (nondisjunction) → dermal sinus tract; explains their frequent association
- Associated abnormalities
  - Intradural lipoma: ± localized dysraphism at lipoma level; segmentation anomalies rare

- Terminal lipoma: Sacral hypogenesis, anorectal malformations, genitourinary (GU) malformations (5-10%), terminal diastematomyelia, epidermoid, dermal sinus, angioma, arachnoid cyst
  - Sacral anomalies much more likely if GU, anorectal malformations present (≥ 90%)

Staging, Grading, & Classification

- Traditional classification divides into intradural and terminal lipomatous malformations
  - Ambiguous and confusing; many lipomatous malformations fall along spectrum between lipoma and lipomyelomeningocele
  - Intradural lipoma
    - Partially encapsulated sessile (55%) or exophytic (45%) juxtamedullary fatty mass entirely enclosed within dural sac
    - Midline spinal cord "open"; subpial lipoma nestled between open lips
  - Terminal lipoma
    - Delicately encapsulated fatty mass attached to cord/filum; frequently contiguous with subcutaneous fat through dorsal lumbosacral dysraphism
    - Cord almost always tethered; stretched and thinned ± hydrosyringomyelia (20%)
- Newly proposed (2009) classification scheme proposes dividing lipomas into 2 groups of lipomatous malformations based on presence or absence of dural defect
  - Better accounts for differences in embryology, clinical presentation, operative findings, complications, and prognosis than traditional classification
  - Lipomas without dural defect
    - Filum lipoma, caudal lipoma without dural defect, intramedullary lipoma
  - Lipomas with dural defect
    - Dorsal lipoma, caudal lipoma with dural defect, transitional lipoma, lipomyelocele, lipomyelomeningocele

Gross Pathologic & Surgical Features

- Composed of normal fat
  - Fat cells ↑ in size considerably during infancy; tiny lipomas in neonates may grow substantially during infancy
  - Conversely, lipomas ↓ in size if patient loses weight

Microscopic Features

- Homogeneous mass of mature fat separated into globules by strands of fibrous tissue
  - ± calcification, ossification, muscle fibers, nerves, glial tissue, arachnoid, ependyma

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Cervical, thoracic intradural lipoma: Slow ascending mono- or paraparesis, spasticity, cutaneous sensory loss, deep sensory loss
  - Lumbosacral intradural lipoma: Flaccid lower extremity paralysis, sphincter dysfunction
  - Terminal lipoma: Bowel/bladder dysfunction, lower extremity weakness/sensory abnormality, foot deformity, scoliosis
- Other signs/symptoms
  - Symptoms may be exacerbated by pregnancy
- Clinical profile
  - Intradural lipoma: Patient complains of weakness and sensory anomalies referable to lesion level
    - Overlying skin usually looks normal; no cutaneous stigmata
  - Terminal lipoma: Patient presents with clinical appearance of "tethered cord" syndrome and (frequently) cutaneous stigmata

Demographics

- Age
  - 3 age peaks for presentation
    - < 5 years (24%)
    - 2nd → 3rd decades (55%)
    - 5th decade (16%)
- Gender
  - Intradural: M ≤ F
  - Terminal: M < F
- Epidemiology
  - Intradural lipoma (4%)
    - < 1% of primary intraspinal tumors
  - Lipomyelo(meningo)cele (includes terminal lipoma) (84%)
  - Filum lipoma (12%)

Natural History & Prognosis

- Small lipomas may grow dramatically during infancy
- Symptomatic patients unlikely to improve spontaneously without intervention

Treatment

- Surgical resection, untethering of cord (if applicable)

DIAGNOSTIC CHECKLIST

Consider

- Follow-up even small lipomas in neonates; they may grow significantly!

Image Interpretation Pearls

- Profound hypodensity on CT myelography and T1WI hyperintensity distinctively characteristic of lipoma
- Use chemical fat-saturation or inversion recovery MR technique to confirm fat content

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(Left) *Sagittal T1WI MR demonstrates hyperintense signal intensity within a residual intradural dorsal conus subpial lipoma in this patient who underwent prior partial lipoma resection.* **(Right)** *Sagittal T1 FS MR reveals focal low signal intensity in a small conus lipoma, confirming the fatty nature of the lesion. A T1 hyperintense epidermoid cyst or subacute (methemoglobin) hemorrhage would not demonstrate this suppression, confirming the lipoma diagnosis.*





(Left) *Sagittal T1WI MR reveals a classic intradural subpial lipoma adherent to the dorsal cervical spinal cord. Note that there is very subtle remodeling and enlargement of the osseous spinal canal at the lipoma level.* **(Right)** *Sagittal T1WI MR depicts a large intradural thoracic lipoma that invaginates into the dorsal spinal cord surface. Note also remodeling of the dorsal osseous spinal canal indicating the longstanding nature of this lesion.*





(Left) *Sagittal T1WI MR reveals a terminal lipoma tethering the low-lying spinal cord at the S1 level. There is also associated terminal syringohydromyelia. A vitamin E capsule placed at the skin opening marks clinically detected dermal sinus ostium that prompted imaging.* **(Right)** *Sagittal T1WI MR demonstrates a low-lying spinal cord with long hyperintense filum lipoma that inserts into a heterogeneous intraspinal sacral dermoid tumor.*

KEY FACTS

TERMINOLOGY

- Synonyms: Dermal sinus tract (DST)
- Midline/paramedian stratified squamous epithelial-lined sinus tract
- Extends inward from skin surface for variable distance

IMAGING

- Sinus tract easily identified superimposed on background of cutaneous fat
- Terminus usually conus medullaris (lumbosacral) or central spinal canal (cervical, thoracic)

TOP DIFFERENTIAL DIAGNOSES

- Low coccygeal midline dimple
- Pilonidal sinus
- (Epi)dermoid tumor without dermal sinus

PATHOLOGY

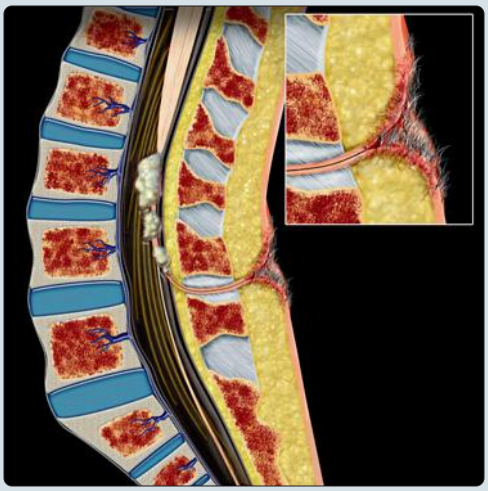
- Focal incorporation of cutaneous ectoderm into neural ectoderm during disjunction at circumscribed point only → focal segmental adhesion
- Spinal cord ascends relative to spinal canal, stretches adhesion into long, tubular tract

CLINICAL ISSUES

- Infancy → 3rd decade
- Presentation either asymptomatic (incidentally noted skin dimple) or infection, neurological deficits 2° to cord tethering or compression

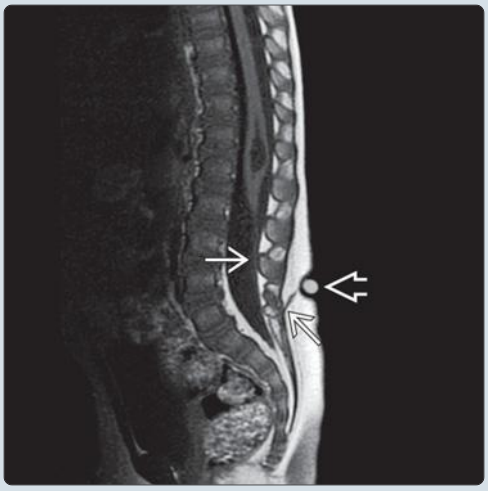
DIAGNOSTIC CHECKLIST

- Must differentiate DST from simple sacral dimple or pilonidal sinus
- Identify sinus course, termination for surgical planning





(Left) *Sagittal graphic shows a dermal sinus extending from the skin surface into the spinal canal to terminate at conus with epidermoid cysts. A skin dimple with capillary angioma and hairy tuft (cutaneous marker) indicates sinus opening.* **(Right)** *Sagittal T2WI MR shows hypointense dermal sinus ➡ extending through L5 posterior elements and coursing intradural to tether the low-lying hydromyelic conus ➡ at L2-L3. A vitamin E capsule has been placed to mark the skin opening of the sinus ➡.*





(Left) *Sagittal T1WI MR reveals hypointense dermal sinus ➡ extending through L5 posterior elements and coursing intradural to tether the low-lying conus at L2-L3. A vitamin E capsule marks skin opening of the sinus ➡.* **(Right)** *Sagittal T1WI C+ FS MR confirms mild DST enhancement ➡ without abscess. Note that the terminal cord central canal is dilated ➡, probably representing syringohydromyelia rather than ventriculus terminalis because of low-lying cord and presence of dermal sinus.*

TERMINOLOGY

Synonyms

- Dermal sinus tract (DST)

Definitions

- Midline/paramedian stratified squamous epithelial-lined sinus tract extending inward from skin surface for variable distance

IMAGING

General Features

- Best diagnostic clue
  - T1W hypointense sinus tract superimposed on hyperintense subcutaneous fat
- Location
  - Lumbosacral (60%) > occipital (25%) > thoracic (10%) > cervical (1%)
- Size
  - Thin tract (several mm); length variable
- Morphology
  - Subcutaneous tract ± dysraphism
  - Length of tract varies short → long; may end in subcutaneous tissue or extend to terminus
    - Terminus usually conus medullaris (lumbosacral) or central spinal canal (cervical, thoracic)

Radiographic Findings

- Radiography
  - ± dysraphism, laminar defect

CT Findings

- CECT
  - ± (epi)dermoid, ± ring enhancement (abscess, arachnoiditis), or nerve root clumping (adhesive arachnoiditis, prior infection, ([epi]dermoid rupture)
- Bone CT
  - Osseous findings range from normal → groove in lamina/spinous process → multilevel dysraphism

MR Findings

- T1WI
  - Sinus tract hypointense to subcutaneous fat
    - Extraspinal tract passes inferiorly and ventrally to lumbodorsal fascia, turns upward to ascend within spinal canal
  - Dorsal dural tenting indicates → dural penetration
    - Intradural sinus course nearly impossible to follow; indistinguishable from cauda equina, filum
  - ± (epi)dermoid cyst
    - Dermoid: Hypo- → hyperintense (fat)
    - Epidermoid: Hypointense
    - Extradural lesions may be subtle; look for nerve root or cord displacement
    - Ruptured (epi)dermoid difficult to detect; look for nerve clumping, CSF "smudging"
  - ± tethered cord
    - Lumbosacral sinus → tethered cord, low-lying conus
    - Thoracic, cervical sinuses → normal conus position
- T2WI
  - Sinus tract hypointense to subcutaneous fat unless filled with fluid
  - ± hyperintense (epi)dermoid cyst
  - ± nerve root clumping (adhesive arachnoiditis)
- DWI
  - ± hyperintense epidermoid cyst
- T1WI C+
  - ± intra-/extramedullary abscess, infectious or chemical arachnoiditis
- MR cine
  - Increased contrast between free-flowing CSF, solid epidermoid tumor
  - Loss of normal conus pulsation

Ultrasonographic Findings

- Grayscale ultrasound
  - Shows entire length of tract from skin to spinal cord
    - Subcutaneous tract slightly hypoechoic, hard to detect
    - Echogenic subarachnoid tract clearly demonstrated within anechoic CSF
  - ± low-lying conus, thick filum, ↓ nerve mobility, intrathecal mass

Nonvascular Interventions

- Myelography
  - Dorsal dural tenting ± (epi)dermoid, nerve root clumping

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Sagittal and axial T1WI, T2WI
    - Adjust window/level to best delineate subcutaneous tract
  - Ultrasound supplements MR in infants < 6 months; use MR to confirm positive ultrasound study

DIFFERENTIAL DIAGNOSIS

Low Coccygeal Midline Dimple

- 2-4% of infants
- Small (< 5 mm), low (< 2.5 cm from anus), extend inferiorly or horizontally toward coccyx
- Usually no associated masses, other cutaneous stigmata

Pilonidal Sinus

- Common; nearly always incidental
- Low ostium, do not enter spinal canal

(Epi)dermoid Tumor Without Dermal Sinus

- No skin stigmata or sinus tract

PATHOLOGY

General Features

- Etiology
  - Localized incorporation of cutaneous ectoderm into neural ectoderm during disjunction → focal segmental adhesion
    - Spinal cord ascends relative to spinal canal, stretches adhesion into long, tubular tract
- Associated abnormalities

- (Epi)dermoid tumor (30-50%)
  - Midline sinus ostia → usually dermoid
  - Paramedian ostia → epidermoid more common
  - May be multiple; most common at conus
- Epidural/subdural abscess, meningitis, or intramedullary abscess 2° to staphylococcal or coliform bacteria
- Lipoma (15-20%)
- Cutaneous stigmata; angioma, pigment abnormalities, hypertrichosis, lipoma, skin tag, tail, or (rarely) supranumerary sinus tracts

- 3 clinically important types of sinus tracts
  - Low sacral or coccygeal dermal sinuses form differently embryologically; always terminate in sacral or coccygeal fascia and never extend into subarachnoid space
  - Pilonidal sinus: Low ostium, do not enter spinal canal
  - Congenital dorsal dermal sinus + atypical dimple (large [> 5 mm], remote from anus [> 2.5 cm], combined with other lesions)
- Midline dimple/pit is one of most common referrals to pediatric neurosurgeons
  - Regardless of depth, dimples below top of intergluteal crease end blindly and never extend intraspinally
  - Sinus opening dermatomal level correlates with metameric cord level of attachment

Gross Pathologic & Surgical Features

- Sinus tract course varies short → long; may terminate extraspinal
- Intraspinal extension of sinus tract ≥ 50%
  - May terminate in subarachnoid space, conus medullaris, filum terminale, nerve root, fibrous nodule on cord surface, or (epi)dermoid cyst
- Palpable sinus tract
- (Epi)dermoid tumor
  - ± cheesy, oily material (dermoid) or discrete pearly white (epidermoid) tumor
  - Capsule often adherent to surrounding neural structures

Microscopic Features

- Sinus tract lined by stratified squamous epithelium
- Epidermoid: Desquamated epithelium
- Dermoid: Skin adnexa

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic; incidentally noted skin dimple
  - Neurological deficits below level of tract secondary to tethering or cord compression
  - Other signs/symptoms
    - Meningitis, intraspinal abscess (retrograde passage of pathogens)
- Clinical profile
  - Atypical sacral dimple above intergluteal cleft, pinpoint ostium, ± cutaneous stigmata

Demographics

- Age
  - Infancy 3rd decade
- Gender
  - M = F
- Epidemiology
  - Low sacrococcygeal dimple: 2-4% of all infants
  - Pilonidal sinus: Common
  - DST: Uncommon, midline > paramedian ostium

Natural History & Prognosis

- Progressive neurological deterioration from cord tethering, (epi)dermoid enlargement, mass effect on spinal cord or cauda equina, sequelae of meningitis/abscess
  - Early surgical intervention → normal neurological development possible
- Most important factor influencing outcome is total excision of sinus before development of infection, neural compression

Treatment

- Surgical excision of sinus tract, tethered cord release, treatment of complications
- Long-term antibiotics (if infected)

DIAGNOSTIC CHECKLIST

Consider

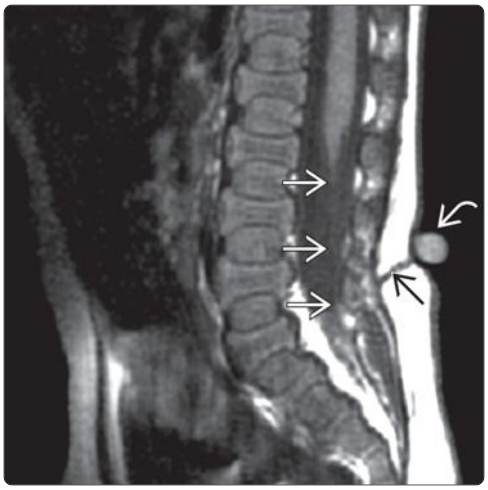
- Dorsal dermal sinus must be differentiated from simple sacral dimple or pilonidal sinus
- Regardless of depth, dimples below top of intergluteal crease end blindly and never extend intraspinally
- Maintain high index of suspicion for all dimples above intergluteal fold

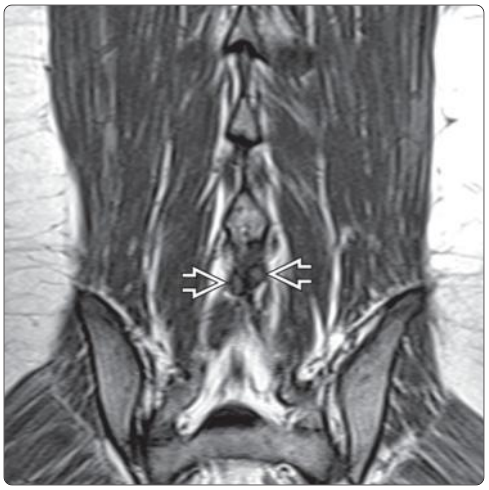
Image Interpretation Pearls

- Critical to identify dermal sinus course and termination for surgical planning
- Up to 50% of dermal sinuses associated with (epi)dermoid tumor
- Dural "nipple" on sagittal image indicates dural penetration

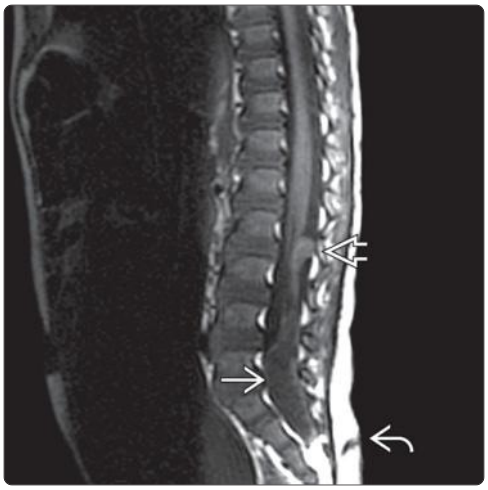
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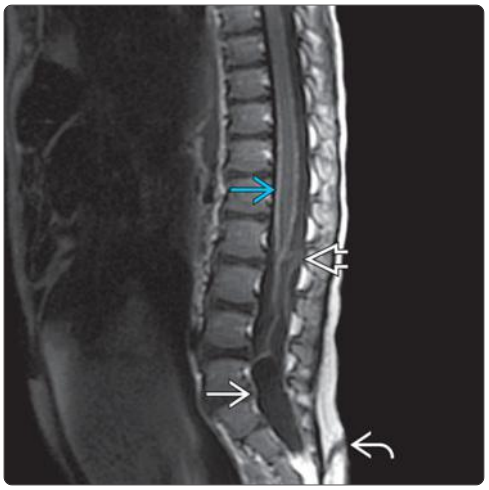
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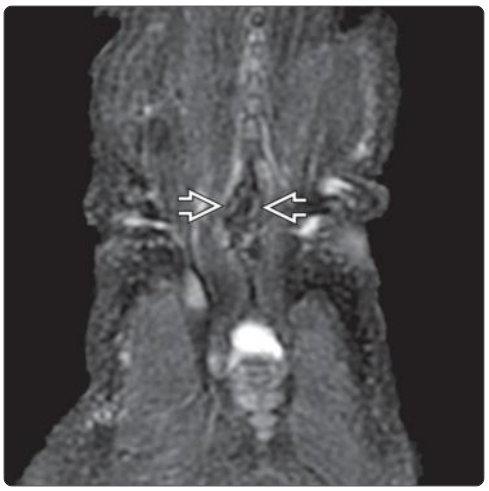
(Left) *Sagittal T1WI MR shows a typical dermal sinus tract entering the dural sac at L4-L5 and coursing cranially, terminating at the low-lying conus. A vitamin E capsule marks the skin opening.* **(Right)** *Coronal T1WI MR in a patient with lumbar dermal sinus tract demonstrates a bifid L5 spinous process. This mild dysraphic defect represents the point of spinal entry for the dorsal dermal sinus.*





(Left) *Sagittal T1WI MR shows low to isointense signal intradural masses (epidermoid cysts) at L2-L3 and L5-sacrum. There is a dermal sinus tract extending dorsally from the low sacral region.* **(Right)** *Sagittal T1WI C+ MR in patient with dorsal dermal sinus tract and clinical meningitis demonstrates diffuse abnormal pial and cauda equina enhancement. Note minimal rim enhancement of the sacral and lumbar epidermoid cysts.*





(Left) *Sagittal T1WI MR depicts a lumbar lipomyelomeningocele with additional dermal sinus tract extending from the skin opening into the lipomatous mass. The spinal cord termination within the lipomatous mass is low lying and reflects radiographic cord tethering.* **(Right)** *Coronal T2 STIR MR confirms a bifid L5 spinous process. This finding represents a subtle posterior dysraphism that facilitates passage of the dermal sinus tract into the spinal canal.*

Simple Coccygeal Dimple

KEY FACTS

TERMINOLOGY

- Synonym: Sacral dimple

IMAGING

- Low sacral dimple connecting to coccyx by fibrous tract
  - Usually resides within intergluteal cleft
  - No intradural extension by definition
- Variable size; deeper dimples usually evoke more physician and parental concern

TOP DIFFERENTIAL DIAGNOSES

- Dorsal dermal sinus
- Pilonidal sinus

PATHOLOGY

- Congenital
- Tract usually atretic; may occasionally be patent with fluid in lumen

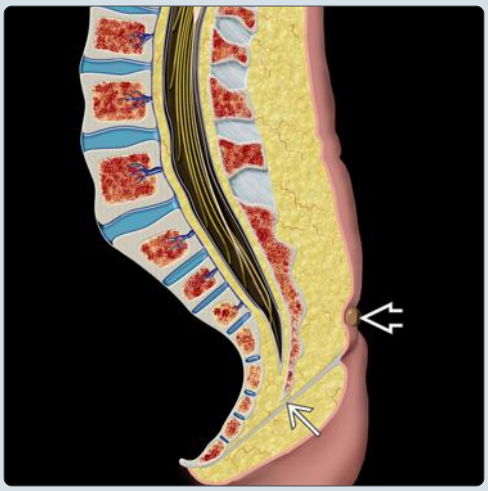
CLINICAL ISSUES

- Often asymptomatic discovery by parents during diaper change or bathing
  - Dimple becomes less conspicuous as patient grows
  - No specific treatment indicated; reassure parents
- Occasionally present with acute inflammation or purulent discharge

DIAGNOSTIC CHECKLIST

- Important to distinguish from dorsal dermal sinus, which requires surgical excision
  - Low dimple opening within intergluteal cleft usually but not always coccygeal dimple
  - High dimple position, leaking of fluid more likely dorsal dermal sinus
- Look for hypointense tract surrounded by bright fat on MR
- Always mark skin dimple ostium with MR visible marker

(Left) *Sagittal graphic of the lumbosacral spine demonstrates a low sacral dimple (marked by vitamin E capsule ⇨), which is connected to the coccyx tip by a fibrous tract ⇨. There is no intradural extension, and the conus/intradural structures are normal.* **(Right)** *Sagittal T2WI MR demonstrates a deep low sacral dimple with vitamin E marker capsule ⇨ placed. The conus terminates at normal L1 level. The dimple and tract ⇨ connect directly to the coccyx ⇨ with no intradural extension.*





(Left) *Sagittal T1WI MR reveals a deep sacral dimple marked for imaging with a vitamin E capsule ⇨. The conus terminates at normal L1 level. The dimple shows typical "spot-welded" proximity to the coccyx via a short fibrous tract ⇨.* **(Right)** *Axial T1WI MR confirms that the sacral dimple, marked with vitamin E capsule ⇨, and tract ⇨ connect directly to the coccyx ⇨ without intradural extension.*





TERMINOLOGY

Synonyms

- Sacral dimple

Definitions

- Low sacral dimple connecting to coccyx by fibrous tract
- No intradural extension by definition

IMAGING

General Features

- Best diagnostic clue
  - Subcutaneous tract extending between surface skin dimple and coccyx
- Location
  - Low; usually resides within intergluteal cleft
- Size
  - Variable; deeper dimples usually evoke more physician and parental concern
- Morphology
  - Tract usually atretic; may occasionally be patent with fluid in lumen

CT Findings

- CECT
  - Limited role for diagnostic evaluation because of poor tract delineation

MR Findings

- T1WI
  - Hypointense tract extends from dimple to coccyx tip
  - Tract conspicuous because surrounded by hyperintense fat
- T2WI
  - Hypointense tract extends from dimple to coccyx tip
  - Occasionally tract lumen is patent → T2-hyperintense fluid signal
    - These patients thought to have greater infection risk for skin abscess
- T1WI C+ FS
  - Tract and subcutaneous tissues may enhance, especially if inflamed

Ultrasonographic Findings

- Grayscale ultrasound
  - Imaging over dimple reveals coccyx tip; may be able to follow tract to coccyx
  - Useful to confirm normal conus position, periodic motion in low-risk dimple
  - Generally most helpful within 1st 3 months of life, before posterior elements are ossified

Imaging Recommendations

- Protocol advice
  - Always mark skin dimple opening with MR visible marker
  - T1WI MR usually most helpful pulse sequence

DIFFERENTIAL DIAGNOSIS

Dorsal Dermal Sinus

- Ostium usually higher than coccygeal dimple; may be off midline or have cutaneous stigmata
- Conus often low and may appear tethered
- Increased risk for meningitis if sinus lumen patent; surgical excision indicated

Pilonidal Sinus

- Generally no imaging role except for detection of subcutaneous abscess
- Unclear if etiologically related to simple coccygeal dimples

PATHOLOGY

General Features

- Etiology
  - Congenital
- Associated abnormalities
  - None

Gross Pathologic & Surgical Features

- Fibrous tract with atretic lumen
- No intradural extension

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often asymptomatic discovery by parents during diaper change or bathing
- Other signs/symptoms
  - Sometimes present with acute inflammation or purulent discharge

Natural History & Prognosis

- Dimple becomes less conspicuous as patient grows

Treatment

- No specific treatment indicated; reassure parents

DIAGNOSTIC CHECKLIST

Consider

- Coccygeal dimples never communicate with thecal sac
- Important to distinguish from dorsal dermal sinus, which requires surgical excision

Image Interpretation Pearls

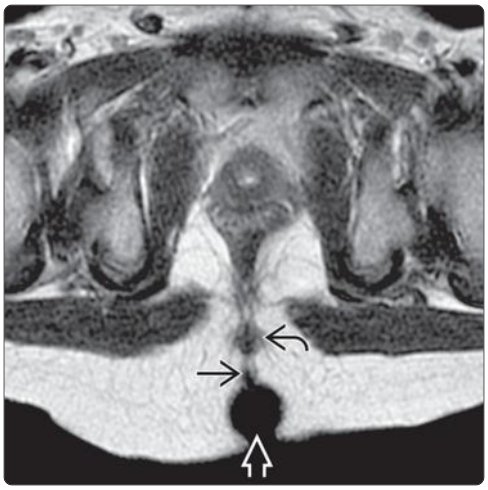
- Look for hypointense tract surrounded by bright fat on MR

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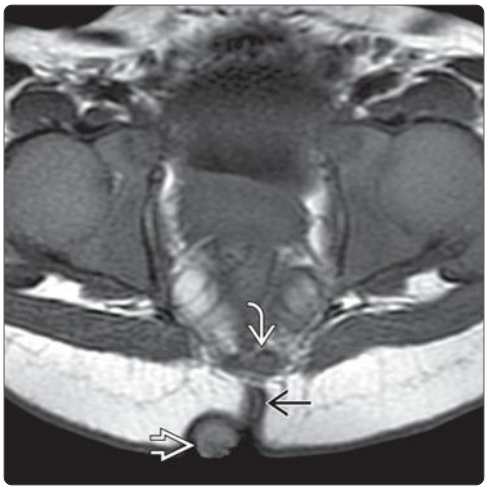
(Left) *Sagittal T1WI MR demonstrates a deep dimple with fibrous tract connecting to coccyx. Note a vitamin E capsule was placed within the dimple to mark the skin opening. There is no communication with the thecal sac.* **(Right)** *Axial T2WI MR confirms connection of low sacral dimple marked by hypointense vitamin E capsule with the coccyx by a short fibrous tract. Dimple location in gluteal cleft and contiguity with the coccyx are characteristic of the simple coccygeal dimple.*





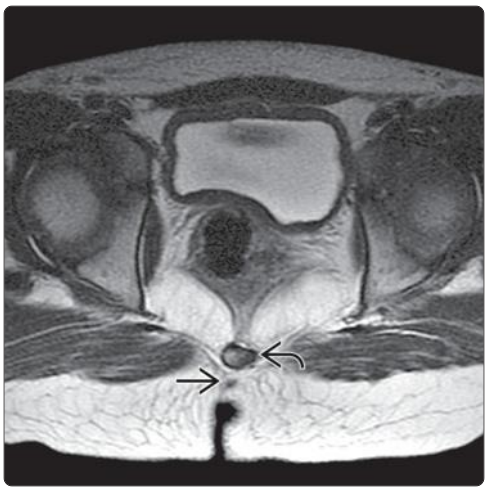
(Left) *Parasagittal T1WI MR demonstrates normal conus termination at L1 without imaging stigmata of tethered spinal cord. A vitamin E capsule has been placed to mark the skin opening of the dimple contiguous with fibrous tract.* **(Right)** *Axial T1WI MR depicts tract extension from the dimple marker to the coccyx. The low position of the dimple and connection to the coccyx are typical of a simple coccygeal dimple, which never communicates with the thecal sac.*

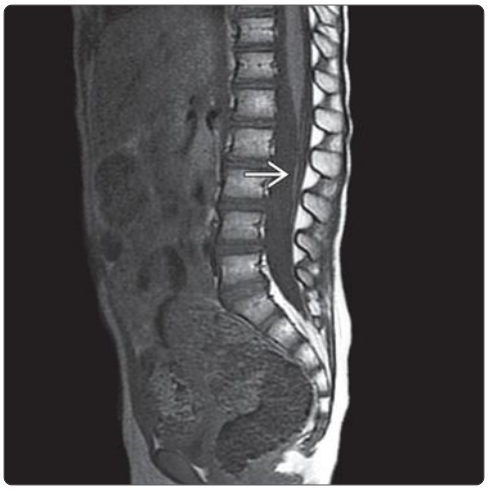


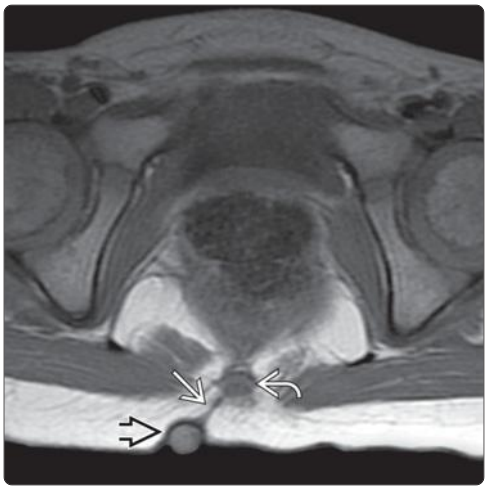


(Left) *Sagittal T1WI MR obtained off midline shows normal sacral development. The deep sacral dimple is marked with a vitamin E capsule to facilitate its identification on MR imaging. A short fibrous tract extends to the coccyx tip.* **(Right)** *Axial T2WI MR shows a deep sacral dimple. The hypointense tract is conspicuously surrounded by hyperintense fat along its course to the coccyx.*



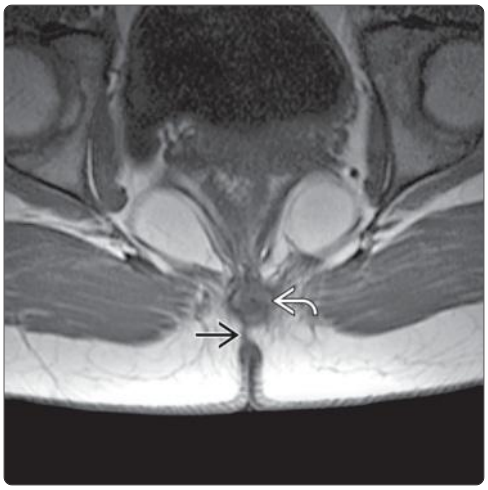






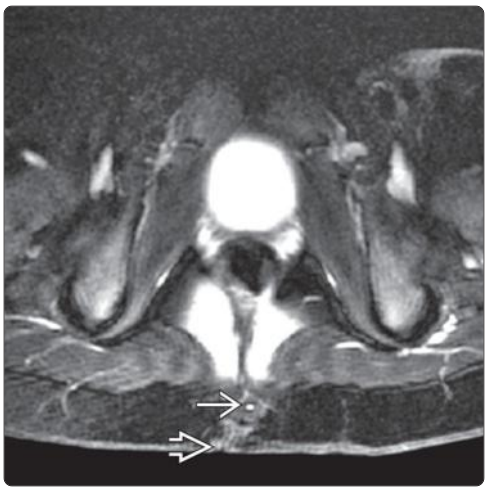
(Left) *Sagittal T1WI MR shows normal conus termination and lumbosacral anatomy. Incidentally noted is minimal fatty infiltration of the filum. This image does not reveal the clinically obvious deep sacral dimple, reiterating the importance of marking skin dimple ostium.* **(Right)** *Axial T1WI MR shows that the dimple, marked by vitamin E capsule, is slightly off midline, and connects to the coccyx by a short fibrous tract.*





(Left) *Sagittal T1WI MR in a patient with a low sacral dimple demonstrates a distended sinus tract to the coccyx tip filled with debris. The cutaneous dimple (not shown) was fairly small.* **(Right)** *Axial T1WI MR shows a fibrous tract extending from the skin to the coccyx. The more rostral tract (not shown) was filled with debris, a much less common appearance than fibrous atretic tract, indicating a persistently patent tract lumen.*





(Left) *Sagittal T1WI MR shows a cutaneous skin defect with fibrous scarring in the gluteal cleft corresponding to the clinically detected dimple. The hypointense sinus tract leading to the coccyx tip is short. Low dimples within the gluteal cleft may become infected and present with local abscess, as in this case, but they never communicate with the thecal sac.* **(Right)** *Axial T2WI FS MR reveals an off-midline dimple with patent fluid-filled tract surrounded by fibrous tissue.*

Dermoid Cysts

KEY FACTS

TERMINOLOGY

- Synonyms: Dermoid tumor, "dermoid"
- Benign spinal mass composed of cells embryologically comprising skin and its appendages

IMAGING

- Lumbosacral or cauda equina CSF isointense/isodense mass ± interspersed fat signal intensity/density
- Focal osseous erosion, spinal canal widening, flattening of pedicles and laminae at spinal level of mass
- Less likely to show diffusion restriction than epidermoid cyst

TOP DIFFERENTIAL DIAGNOSES

- Arachnoid cyst
- Neurenteric cyst

PATHOLOGY

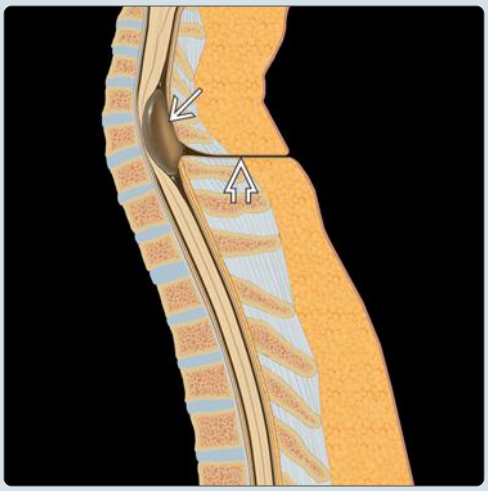
- Congenital or acquired origin
  - Congenital cysts arise from dermal rests or focal expansion of a dermal sinus
- Cyst filled with thick cheesy, buttery, yellowish material (desquamated keratin, lipids)
- Associated anomalies include dermal sinus, vertebral segmentation anomalies, closed dysraphism

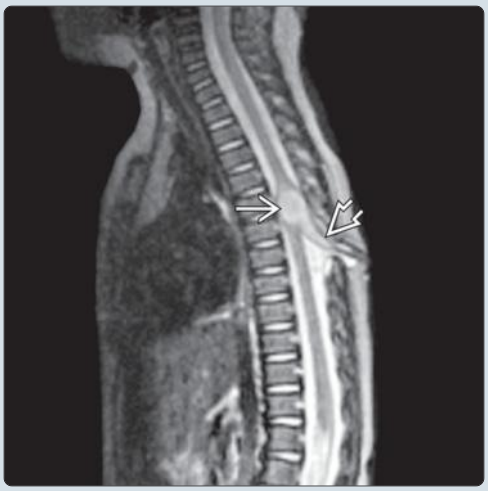
CLINICAL ISSUES

- Most commonly asymptomatic or presentation with slowly progressive compressive radiculopathy/myelopathy
- Infectious meningitis in association with dermal sinus
- Acute chemical meningitis 2° to rupture, discharge of inflammatory cholesterol crystals into CSF

DIAGNOSTIC CHECKLIST

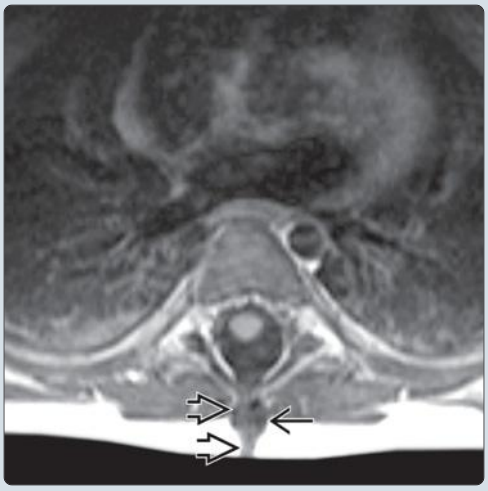
- Often difficult to diagnose on CT and MR; presence of fat helpful to suggest diagnosis





(Left) *Sagittal graphic of the cervicothoracic spine demonstrates a large dorsal dermoid cyst ➡ in association with a dermal sinus tract ➡. There is marked spinal cord compression.* **(Right)** *Sagittal T2WI MR (myelopathy) demonstrates a hyperintense dermoid tumor with an intramedullary component ➡ contiguous with an extramedullary component following a dermal sinus tract ➡ to the skin surface.*





(Left) *Axial T1WI MR (myelopathy) depicts a dermoid cyst ➡ with prominent intramedullary component entirely within the spinal cord substance.* **(Right)** *Axial T1WI MR (myelopathy) shows contiguous extension of dermoid cyst along the dermal sinus tract ➡ to the skin surface. There is a small dermoid cyst ➡ within the dermal sinus tract as well.*

TERMINOLOGY

Synonyms

- Dermoid tumor, "dermoid"

Definitions

- Benign spinal mass
- Composed of cells that embryologically comprise skin and its appendages (hair follicles, sweat glands, and sebaceous glands)

IMAGING

General Features

- Best diagnostic clue
  - Lumbosacral or cauda equina CSF isointense/isodense mass ± interspersed fat signal intensity/density
- Location
  - Lumbosacral (60%), cauda equina (20%)
    - 40% intramedullary, 60% extramedullary; extradural rare
  - Rare in cervical and thoracic spine
- Size
  - Range: Tiny subpial masses → huge growths
- Morphology
  - Uni- or multilobular round/ovoid mass

Radiographic Findings

- Radiography
  - Focal vertebral osseous erosion
  - Spinal canal widening
  - Flattening of pedicles and laminae
  - ± dysraphic posterior elements

CT Findings

- CECT
  - Well-demarcated isodense mass ± regions of fat hypodensity, calcification
    - ± minimal enhancement
- Bone CT
  - Focal osseous erosion, spinal canal widening, flattening of pedicles and laminae at spinal level of mass

MR Findings

- T1WI
  - Hypo- to hyperintense signal intensity mass
    - Hypointensity may reflect increased water content from sweat gland secretions
    - Fat hyperintensity is most specific for dermoid, but least common appearance
      - Intrinsic T1 shortening permits differentiation of dermoid from epidermoid cyst
- T2WI
  - Hyperintense T2 signal intensity mass
- FLAIR
  - Mild hyperintensity to CSF may help detect occult dermoid tumor, differentiate from arachnoid cyst
- DWI
  - Less likely to show diffusion restriction than epidermoid cyst
- T1WI C+
  - ± mild ring enhancement; more avid surrounding contrast enhancement if infected

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic mass lesion with internal echoes, focal areas of hyperechogenicity (if fat present)

Nonvascular Interventions

- Myelography
  - CSF or fat-density mass adjacent to bright CSF
  - Myelographic block common if symptomatic
  - Most useful in conjunction with CT (CT myelography)
  - Largely replaced by MR imaging
    - Primarily of utility in patients with MR contraindications (pacemaker, devices, etc.)

Imaging Recommendations

- Best imaging tool
  - MR; reserve CT myelography for patients with MR contraindications or inconclusive MR studies
- Protocol advice
  - Sagittal and axial T1WI and T2WI MR to include entire conus and cauda equina to coccyx tip

DIFFERENTIAL DIAGNOSIS

Arachnoid Cyst

- Follows CSF signal intensity/density on all sequences
- DWI MR useful to distinguish arachnoid cyst (isointense to CSF) from dermoid with restriction (hyperintense)

Neurenteric Cyst

- Intradural cyst; usually ventral to cord but may be dorsal or intramedullary
- ± vertebral anomalies, hyperdense/hyperintense proteinaceous content
- Definitive diagnosis is pathological

PATHOLOGY

General Features

- Etiology
  - Congenital
    - Arise from dermal rests or focal expansion of a dermal sinus
  - Acquired
    - Iatrogenic lesion
    - Results from implantation of viable dermal and epidermal elements following lumbar puncture (nontrocar spinal needle) or following surgery (myelomeningocele closure)
    - Cells slowly grow until large enough to cause symptoms
    - Link between lumbar puncture and subsequent epidermoid or dermoid cyst is particularly strong in neonatal period
- Associated abnormalities
  - Dermal sinus (20%)
  - Vertebral abnormalities (diastematomyelia, hemivertebra, scoliosis)
  - Closed dysraphism (anterior sacral meningocele, "spina bifida occulta"): Rare

- Benign "tumor"
- Arises from cells that produce skin and its appendages (hair follicles, sweat glands, sebaceous glands)

Gross Pathologic & Surgical Features

- Well-demarcated, smooth uni- or multilocular mass
- Cyst wall may be thickened by skin appendages or calcifications
- Cyst filled with thick, cheesy, buttery, yellowish material

Microscopic Features

- Outer connective tissue capsule lined with stratified squamous epithelium containing hair follicles, sebaceous glands, and sweat glands
- ± inflammation (if ruptured)
- Centrally contains desquamated epithelial keratin, lipid material

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Slowly progressive compressive radiculopathy/myelopathy
- Other signs/symptoms
  - Cauda equina syndrome
  - Infectious meningitis; most common in association with dermal sinus
  - Acute chemical meningitis secondary to rupture and discharge of inflammatory cholesterol crystals into CSF

Demographics

- Age
  - Usually symptomatic before 20 years of age
- Gender
  - M = F
- Epidemiology
  - (Epi)dermoid comprise 1-2% of all spinal cord tumors, up to 10% of spinal cord tumors < age 15 years
  - Dermoid and epidermoid occur roughly equally in spine; ~ 40% single epidermoid, 35% single dermoid, and 5% multiple dermoid or epidermoid

Natural History & Prognosis

- Symptoms slowly progress if untreated
- Complete surgical resection offers best opportunity for good neurologic outcome
  - Incomplete resection frequently recurs; malignant transformation extremely rare

Treatment

- Standard treatment is complete surgical excision

DIAGNOSTIC CHECKLIST

Consider

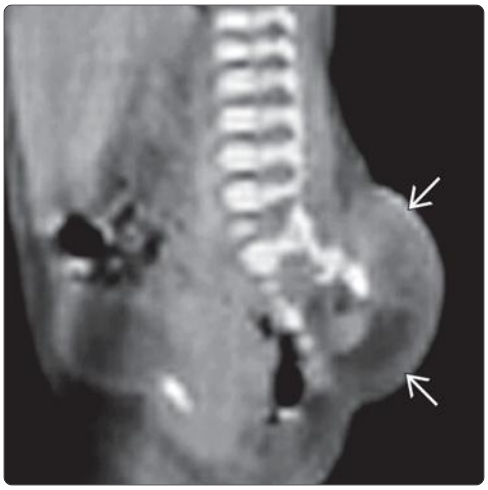
- Congenital lesion that usually presents during childhood/adolescence

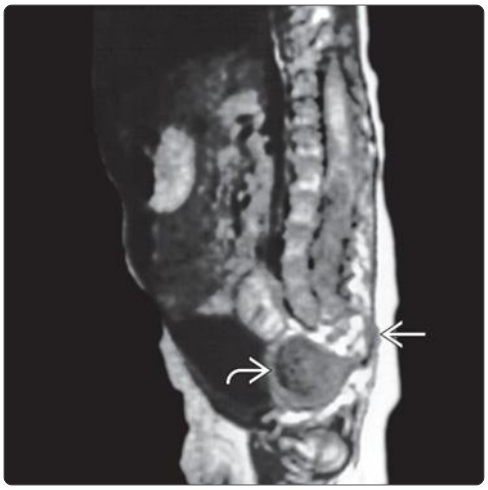
Image Interpretation Pearls

- Often difficult to diagnose on CT and MR; presence of fat helpful to suggest diagnosis
  - Requires high index of suspicion; look for mass effect on regional structures

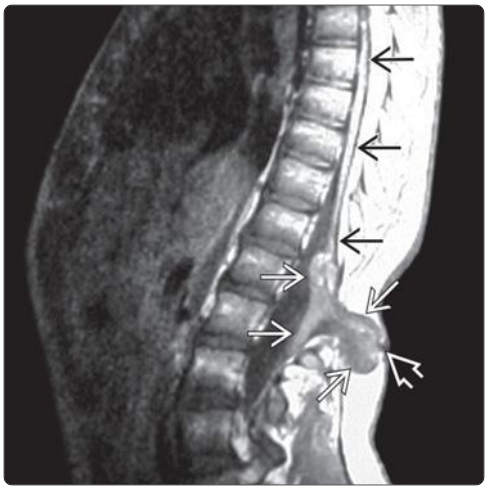
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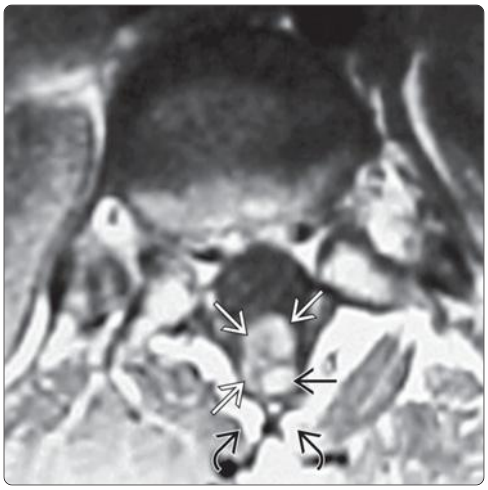
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(Left) *Sagittal NECT in an infant with Currarino triad demonstrates caudal regression and a mixed density dermoid "tumor" ➡ contiguous with the spinal canal through a wide posterior dysraphic defect.* **(Right)** *Sagittal T1WI MR in an infant with Currarino triad shows a small mixed signal intensity dermoid "tumor" ➡ as well as a characteristic associated anterior sacral meningocele ➡.*





(Left) *Sagittal T1WI MR demonstrates a large, lobulated, mildly heterogeneous dermoid tumor ➡ filling a dermal sinus tract from its cutaneous origin ➡ to the conus. The elongated spinal cord ⇨ inserts directly into the dermoid.* **(Right)** *Axial T1WI MR delineates the relationship of the distal tethered spinal cord ⇨ to an extensive dermoid tumor ➡ that fills much of the caudal spinal canal. The posterior elements are dysraphic ⇨, reflecting presence of a dermal sinus tract.*





(Left) *Sagittal T1WI MR reveals a mixed signal intensity extradural sacral dermoid cyst ➡ with intrinsic heterogeneous signal intensity. The low-lying spinal cord ➡ is tethered by a long fatty filum ➡.* **(Right)** *Sagittal STIR MR (sacral dermoid cyst) confirms signal loss within the lipomatous portions of the dermoid ➡. There is also expected suppression of fat signal within the fatty filum ➡.*

Epidermoid Cysts

KEY FACTS

TERMINOLOGY

- Benign nonneoplastic spinal mass embryologically derived from epidermal (skin) elements

IMAGING

- Lumbosacral or cauda equina CSF isointense/isodense mass
  - 40% intramedullary, 60% extramedullary
  - Acquired epidermoid cysts nearly always occur at cauda equina

TOP DIFFERENTIAL DIAGNOSES

- Arachnoid cyst
- Neurenteric cyst

PATHOLOGY

- Congenital (60%)
  - Arise from epidermal rests or dermal sinus
- Acquired (40%)
  - Iatrogenic, follows implantation of viable epidermal elements after lumbar puncture or surgery (myelomeningocele closure)
- Striking white, pearly sheen capsule containing creamy, waxy, pearly material

CLINICAL ISSUES

- Asymptomatic or slowly progressive compressive radiculopathy/myelopathy
- Symptoms slowly progress if untreated
- Complete surgical resection offers best opportunity for good neurologic outcome

DIAGNOSTIC CHECKLIST

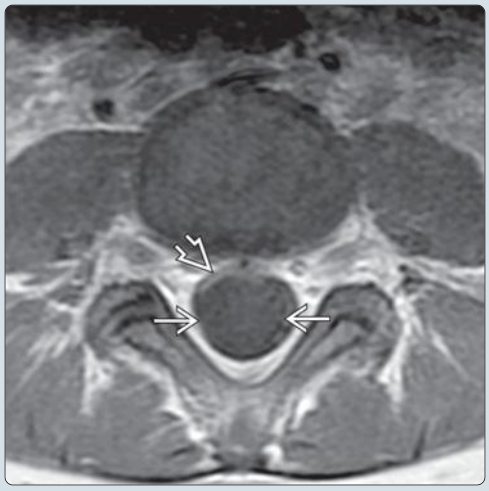
- Epidermoid cysts may be congenital or acquired
- Acquired epidermoid cysts nearly always occur in cauda equina





(Left) *Sagittal graphic of the thoracolumbar spine shows a pearly white epidermoid cyst ➡ located within the cauda equina at the conus, a typical location for post lumbar puncture acquired epidermoid cyst.* **(Right)** *Sagittal T1WI MR demonstrates a hypointense intradural extramedullary mass ➡ within the cauda equina in a patient who previously had an LP as an infant. Signal intensity is slightly hyperintense to CSF, and there is no hyperintense lipid content to indicate dermoid cyst is conspicuous.*





(Left) *Sagittal T2WI MR in a patient who had an LP as an infant reveals a well-circumscribed intradural extramedullary mass ➡ within the cauda equina. The cauda equina is distorted and anteriorly displaced by the cyst. Signal intensity is slightly hypointense to CSF.* **(Right)** *Axial T1WI MR demonstrates that the epidermoid cyst ➡ nearly fills the spinal canal and displaces the cauda equina ➡ anteriorly. This location is typical of a post lumbar puncture acquired epidermoid cyst.*

TERMINOLOGY

Synonyms

- Epidermoid tumor

Definitions

- Benign nonneoplastic spinal mass composed of cells embryologically derived from epidermal (skin) elements

IMAGING

General Features

- Best diagnostic clue
  - Lumbosacral or cauda equina CSF isointense/isodense mass
- Location
  - 40% intramedullary, 60% extramedullary; extradural rare
    - Upper thoracic (17%), lower thoracic (26%), lumbosacral (22%), cauda equina (35%)
  - Acquired epidermoid cysts nearly always occur at cauda equina
- Size
  - Range: Tiny subpial masses → huge growths
- Morphology
  - Uni- or multilobular round/ovoid mass

Radiographic Findings

- Radiography
  - Focal vertebral osseous erosion, spinal canal widening, flattening of pedicles and laminae at spinal level of epidermoid cyst

CT Findings

- CECT
  - Well-circumscribed hypodense mass with attenuation similar to CSF ± calcification (rare)
    - Rarely hyperdense on nonenhanced images reflecting high protein content, hemorrhage, or cellular debris
    - Minimal to no enhancement (unless infected)
- Bone CT
  - Focal osseous erosion, spinal canal widening, flattening of pedicles and laminae at spinal level of epidermoid cyst

MR Findings

- T1WI
  - Usually isointense to CSF
  - Occasionally mildly hyperintense to CSF, reflecting protein or cellular debris (white epidermoid)
- T2WI
  - Hyperintense signal intensity to CSF
- FLAIR
  - Mildly hyperintense to CSF
    - May help detect occult epidermoid tumor, differentiate from arachnoid cyst
- DWI
  - Hyperintense diffusion restriction with concordant hypointensity on ADC map
    - Distinguishes epidermoid from arachnoid cyst (isointense to CSF)
- T1WI C+
  - ± minimal ring enhancement
  - May enhance avidly if infected

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic mass lesion with subtle internal echoes

Nonvascular Interventions

- Myelography
  - CSF density mass conspicuous adjacent to bright CSF
    - Most useful in conjunction with bone CT
  - Myelographic block common in symptomatic patients
  - Has been largely replaced by noninvasive MR imaging

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
    - Reserve CT myelography for patients with MR contraindications, inconclusive MR studies
- Protocol advice
  - Sagittal and axial T1WI and T2WI MR to include entire conus and cauda equina → coccyx
  - DWI to evaluate for diffusion restriction

DIFFERENTIAL DIAGNOSIS

Arachnoid Cyst

- Follows CSF signal intensity/density on all sequences
- DWI MR useful to distinguish arachnoid cyst (isointense to CSF) from epidermoid (hyperintense)

Neurenteric Cyst

- Intradural cyst; usually ventral to cord but may be dorsal or intramedullary
- ± vertebral anomalies, hyperdense/hyperintense proteinaceous content
- Definitive diagnosis is pathological

PATHOLOGY

General Features

- Etiology
  - Congenital (60%)
    - Arise from epidermal rests or focal expansion of a dermal sinus
  - Acquired (40%)
    - Iatrogenic lesion resulting from implantation of viable epidermal elements following lumbar puncture (nontrocar spinal needle) or surgery (myelomeningocele closure)
    - Cells slowly grow until large enough to produce symptoms
    - Link between lumbar puncture and subsequent epidermoid cyst development is strongest in neonatal period
- Associated abnormalities
  - Dermal sinus (20%)
  - Vertebral abnormalities (diastematomyelia, vertebral segmentation anomalies, scoliosis)
  - Closed spinal dysraphism (anterior sacral meningocele, spina bifida occulta): Rare

Gross Pathologic & Surgical Features

- Striking white, pearly sheen capsule; may be smooth, lobulated, or nodular
- Cyst filled with creamy, waxy, pearly material

- May be either easy to shell out or firmly affixed to regional structures (result of local inflammation)

Microscopic Features

- Outer connective tissue capsule lined with stratified squamous epithelium; calcification rare
- Centrally contains desquamated epithelial keratin, cholesterol crystals; positive staining with antibodies to EMA and cytokeratin

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Slowly progressive, compressive radiculopathy/myelopathy
- Other signs/symptoms
  - Infectious meningitis; most common in association with dermal sinus
  - Chemical meningitis secondary to rupture and discharge of inflammatory cholesterol crystals into CSF
  - Cauda equina syndrome

Demographics

- Age
  - Slower growing than dermoids; symptoms usually manifest in early adulthood (3rd-5th decades)
- Gender
  - M > F
- Epidemiology
  - (Epi)dermoid cysts comprise 1-2% of all spinal cord masses, up to 10% of spinal cord masses < 15 years of age
  - Dermoid and epidermoid cysts occur roughly equally in spine; ~ 40% single epidermoid, 35% single dermoid, and 5% multiple dermoid or epidermoid

Natural History & Prognosis

- Symptoms slowly progress if untreated
- Complete surgical resection offers best opportunity for good neurologic outcome
  - Incomplete resection frequently recurs; malignant transformation extremely rare

Treatment

- Standard treatment is complete surgical excision
- Radiotherapy not established for treatment of epidermoid cysts
  - May be alternative to palliative operation or for patients who cannot undergo surgery

DIAGNOSTIC CHECKLIST

Consider

- Epidermoid cysts may be congenital or acquired
  - Acquired epidermoid cysts nearly always occur in cauda equina
- Usually diagnosed later than dermoid (3rd-5th decades)

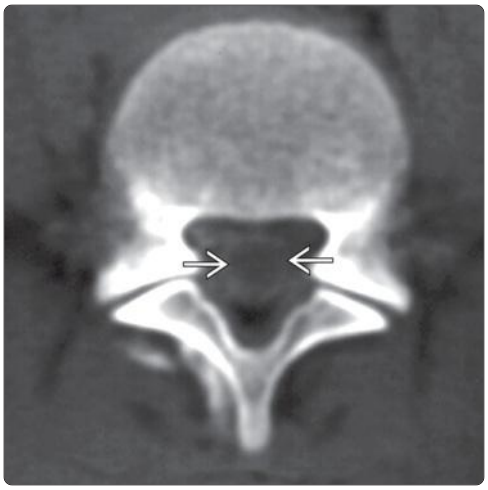
Image Interpretation Pearls

- Frequently difficult to identify using CT or MR
  - Diagnosis requires high index of suspicion; look for mass effect on regional structures
  - FLAIR and DWI may distinguish epidermoid cyst from CSF, arachnoid cyst

SELECTED REFERENCES

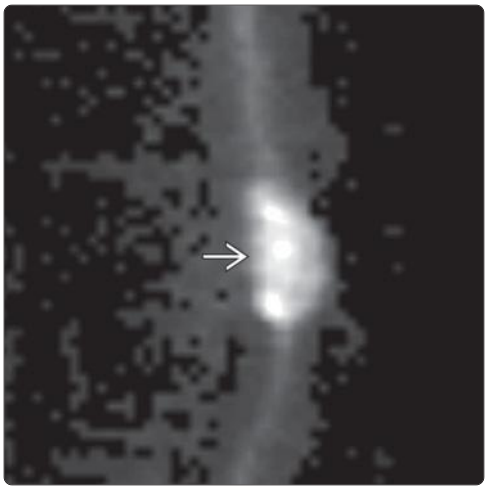
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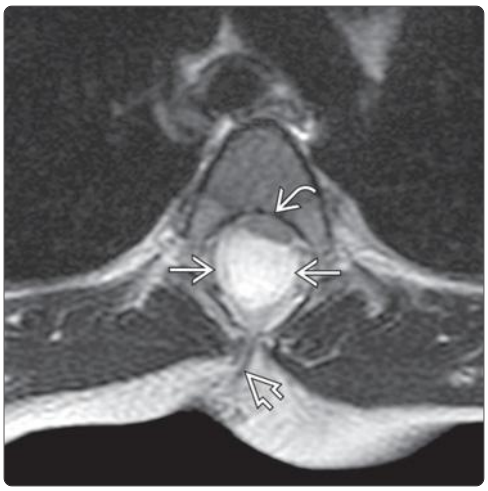
(Left) *Anteroposterior myelography demonstrates a hypodense intradural mass, representing an epidermoid cyst nestled within the cauda equina.* **(Right)** *Axial CT myelography confirms a rounded hypodense epidermoid cyst in a typical intradural location.*





(Left) *Sagittal T1WI MR demonstrates an inhomogeneous mixed signal intensity intramedullary lesion within the swollen thoracic spinal cord.* **(Right)** *Sagittal DWI MR reveals a high signal intensity intramedullary epidermoid cyst with avid diffusion restriction. ADC map (not shown) showed low ADC values, confirming true restricted diffusion. At surgery, an intramedullary epidermoid cyst was confirmed.*





(Left) *Sagittal T1WI MR shows a thoracic dermal sinus tract extending from the skin surface into the spinal canal, contiguous with a large intradural epidermoid cyst. The epidermoid cyst displaces the spinal cord.* **(Right)** *Axial T2WI MR reveals a dermal sinus tract extending from the skin surface into the spinal canal through a bifid thoracic spinous process, connecting to an intradural epidermoid cyst. Note ventral displacement and distortion of the spinal cord.*

KEY FACTS

TERMINOLOGY

- Synonyms: Tethered cord syndrome (TCS), tight filum terminale syndrome (TFTS)

IMAGING

- Stretched, thinned cord with low-lying conus, thickened filum
- ± fibrolipoma/terminal lipoma, dysraphism, vertebral segmentation anomalies
- ↓ spinal cord motion

TOP DIFFERENTIAL DIAGNOSES

- Normal variant low-lying conus
- Open or closed spinal dysraphism
- Postsurgical low-lying conus

PATHOLOGY

- Tethering stretches nerve fibers, arterioles, and venules → impairs oxidative metabolism of conus and nerve roots → syringohydromyelia, myelomalacia
- Tethered filum histologically abnormal, even if conus terminates at normal level

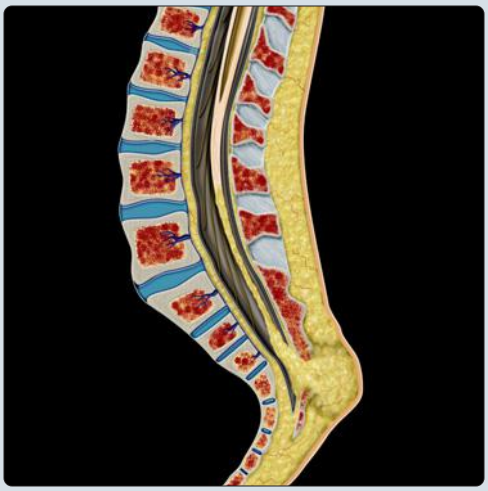
CLINICAL ISSUES

- Low back and leg pain, gait and sensory abnormalities, urinary bladder dysfunction
- Symptomatic presentation most common during rapid somatic growth (adolescent growth spurt, school age 4-8 years), or 2° to kyphosis (elderly)

DIAGNOSTIC CHECKLIST

- TCS is clinical diagnosis; imaging role is detection of low-lying conus/thick filum, associated anatomic abnormalities for surgical decision making
- Clinical tethering may be present despite normal conus level
- Measure filum thickness at L5/S1; stretching at more rostral levels may erroneously thin filum into "normal" size range

(Left) *Sagittal graphic of the lumbosacral spine depicts composite tethered cord syndrome (TCS) findings of low-lying, hydromyelic tethered cord with thickened filum and fibrolipoma inserting into a terminal lipoma that is contiguous with subcutaneous fat through dorsal dysraphism.* **(Right)** *Sagittal T2WI MR (clinical TCS) demonstrates an elongated low-lying spinal cord extending to the S2 level, where it ends in a small terminal lipoma. Focal sacral posterior dysraphism is also present.*





(Left) *Sagittal T1WI MR (clinical TCS symptoms) confirms an elongated low-lying spinal cord extending to the S2 level and ending in a small terminal lipoma. Focal sacral posterior dysraphism is also conspicuous. In general, lipomas are considerably more conspicuous on T1WI than T2WI.* **(Right)** *Axial T1WI MR at the lumbosacral transition reveals that the abnormal elongated, low-lying spinal cord continues to the sacral level.*





TERMINOLOGY

Synonyms

- Tethered cord syndrome (TCS), tight filum terminale syndrome (TFTS)

Definitions

- Symptoms and imaging findings related to low-lying conus medullaris tethered by short, thick filum terminale

IMAGING

General Features

- Best diagnostic clue
  - Conus ends below L2 inferior endplate; tethered by thickened filum ± fibrolipoma, terminal lipoma
- Location
  - Thoracolumbar junction → sacrum
- Size
  - Filum thickened (> 2 mm at L5-S1, axial MR)
- Morphology
  - Attenuated conus terminates lower than normal

Radiographic Findings

- Radiography
  - May be normal but frequently shows localized dysraphism or incomplete posterior element fusion
  - ± scoliosis (20%)

CT Findings

- NECT
  - Stretched, thinned cord with low-lying conus, thickened filum ± fibrolipoma, dysraphism, vertebral segmentation, and fusion anomalies

MR Findings

- T1WI
  - Thickened filum ± hyperintense lipoma
    - Filum > 2 mm (L5-S1, axial MR)
  - ± low-lying conus; may be difficult to distinguish transition from thickened filum
  - Dorsal positioning of conus medullaris, filum terminale in thecal sac
    - Noted even in prone position; normally cord falls into anterior 2/3 of canal when prone
- T2WI
  - Findings similar to T1WI
  - ± hyperintense dilatation of conus central canal 2° to syringohydromyelia or myelomalacia (25%)
  - Fatty filum → chemical shift artifact
  - Dural sac widened; dorsal dura tense, tented posteriorly by thickened filum
- MR cine
  - ↓ spinal cord motion; ≤ 1/3 return to normal cord motion after untethering, even if symptoms resolve

Ultrasonographic Findings

- Grayscale ultrasound
  - ± low-lying conus, thickened filum terminale, reduced or absent spinal cord movement

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Ultrasound < 6 months old; confirm positive study with MR
  - Thin-section sagittal, axial T1WI and T2WI, phase contrast cine MR; extend axial slices to thecal sac termination

DIFFERENTIAL DIAGNOSIS

Normal Variant Low-Lying Conus

- Asymptomatic patient with normal filum thickness

Open or Closed Spinal Dysraphism

- Spinal lipoma
- Myelomeningocele
- Meningocele
- (Epi)dermoid
- Diastematomyelia
- Dermal sinus tract

Postsurgical Low-Lying Conus

- Imaging shows low-lying conus irrespective of symptoms; cannot exclude retethering by imaging alone
  - Ultrasound, post-contrast MR helpful to evaluate conus motion
  - Diagnosis of retethering must be made using clinical criteria

PATHOLOGY

General Features

- Etiology
  - Tethering stretches nerve fibers, arterioles, and venules → impairs oxidative metabolism of conus and nerve roots → syringohydromyelia/myelomalacia
  - CSF shows elevated lactate and other metabolites → normalize following successful release
- Associated abnormalities
  - Cutaneous stigmata (50%)
  - Lumbosacral hypogenesis, VACTERL syndrome
  - Open or closed spinal dysraphism, incomplete posterior fusion (up to 100%)
    - Diastematomyelia, spinal lipoma, intrasacral or dorsal meningocele, lipomyelomeningocele, myelomeningocele
  - Scoliosis (functional adaption to ↓ length of spinal cord course, ↓ intramedullary tension)
  - Syringohydromyelia/myelomalacia (25%)
- Incomplete retrogressive differentiation with failure of terminal cord involution or failure of filum terminale to lengthen
- Conus normally terminates at or above inferior L2 in ≥ 98% of normal population
  - Conus should be at normal position by birth → 2 months old
  - Conus terminating below L2/3 always abnormal at any postnatal age
- May not be distinct filum; spinal cord sometimes elongated, ends directly in small terminal lipoma

Gross Pathologic & Surgical Features

- Thickened fibrotic filum (55%), small fibrolipoma within thickened filum (23%), or filar cyst (3%)

- Widened dural sac; thickened filum closely applied to or indistinguishable from dural sac

Microscopic Features

- Tethered filum histologically abnormal, even if conus terminates at normal level
  - Normal filum → mainly collagen fibers
  - TCS filum → ↑ connective tissue with dense collagen fibers, hyalinization, and dilated capillaries

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back and leg pain; worst in morning, exacerbated by exertion
  - Gait spasticity, weakness, muscular atrophy
  - ↓ sensation, abnormal lower extremity reflexes
  - Urinary bladder dysfunction
- Other signs/symptoms
  - Cutaneous stigmata (≤ 50%); simple dimple, hairy tuft, or hemangioma
  - Orthopedic foot abnormalities (usually clubfoot)
- Clinical profile
  - Adults and children present differently
    - Adults: Pain 1st (2° to degenerative changes), followed later by weakness ± incontinence
    - Children: Incontinence, scoliosis, weakness
  - Tight filum terminale syndrome (TFTS): Clinical findings of tethered cord with normal conus position

Demographics

- Age
  - Symptomatic presentation most common during rapid somatic growth (school age 4-8 years and adolescent growth spurt), or 2° to kyphosis (elderly)
  - Prevalence in adults probably underestimated
- Gender
  - M = F
- Epidemiology
  - Prevalence unknown; probably more common than appreciated

Natural History & Prognosis

- Progressive, irreversible neurological impairment
  - Majority of patients show improvement or stabilization of neurological deficits following surgical untethering; motor weakness (12-60%), sensory dysfunction (40-60%), pain (50-88%), bladder dysfunction (19-67%)
  - Better outcome if symptom duration shorter or conus moves to more normal level following surgery
- Postoperative symptom recurrence rare; prompts consideration for re-tethering

Treatment

- Symptomatic patients: Early prophylactic surgery
  - Resect tethering mass, release cord, and repair dura
- Asymptomatic patients with radiographic tethering: Management controversial
  - Some advocate prophylactic surgery → low morbidity; prognosis for asymptomatic patients better than symptomatic
  - Others advocate prophylactic surgery only for asymptomatic adults who lead physically active lifestyles

DIAGNOSTIC CHECKLIST

Consider

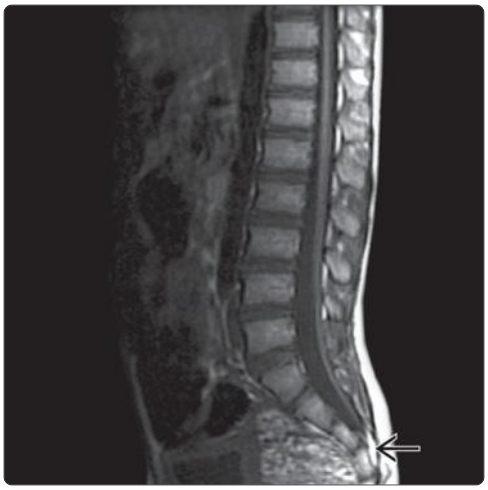
- Clinical tethering may be present despite normal conus level
  - TCS is clinical diagnosis; imaging role is detection of low-lying conus/thick filum, associated anatomic abnormalities for surgical decision making
- Distinguish cord tethering 2° to tight/thickened filum terminale from tethering 2° to other lesions

Image Interpretation Pearls

- Measure filum thickness at L5/S1; stretching at more rostral levels may erroneously thin filum into "normal" size range
- Determine conus level using axial images; cauda equina may obscure conus tip on sagittal images → imitate elongated conus

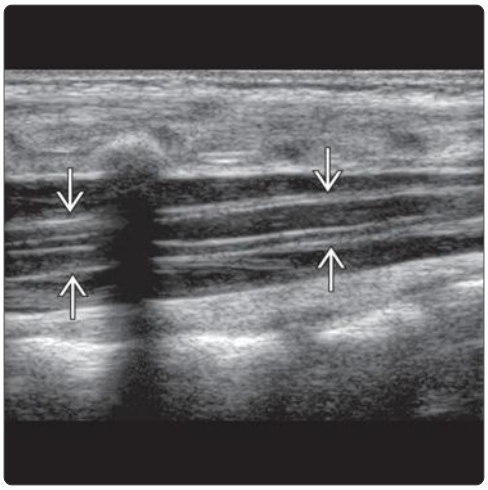
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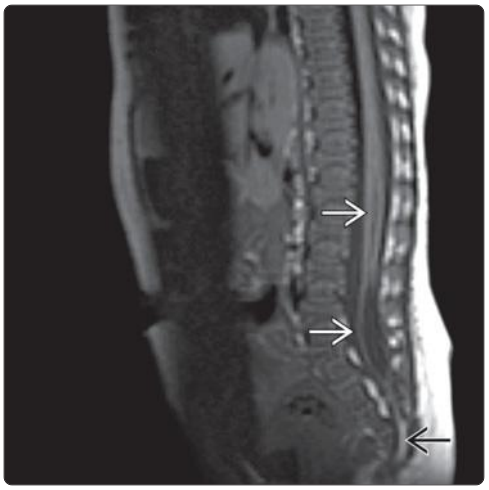
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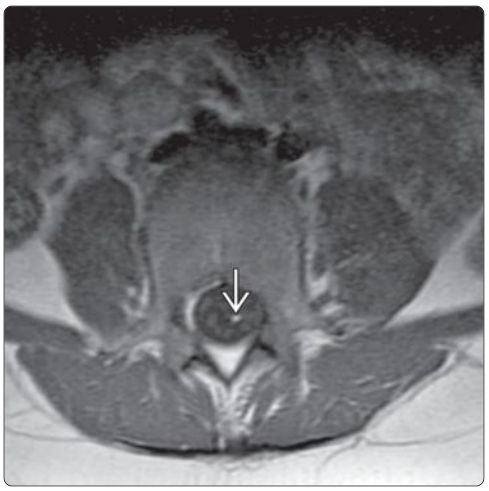
(Left) *Sagittal T1WI MR (clinical TCS) demonstrates an abnormal elongated spinal cord extending to at least L5, subtly transitioning into a thickened filum terminale that inserts into a small terminal lipoma. No associated syringohydromyelia is present.* **(Right)** *Axial T2WI MR (clinical TCS) reveals an otherwise normal-appearing low-lying spinal cord at the lower lumbar spine level.*





(Left) *Longitudinal ultrasound (neonate with suspected tethered cord) demonstrates a low-lying, hydromyelic spinal cord extending to the S1 vertebral level. Sonography shows syrinx contiguity with central spinal cord canal.* **(Right)** *Sagittal T1WI MR (neonate with suspected tethered cord) confirms abnormal-appearing, low-lying spinal cord with syringohydromyelia and short thickened filum terminale inserting into a distal thecal sac terminal lipoma.*





(Left) *Sagittal T2WI MR (vertebral anomalies, clinical tethered cord) shows congenital segmentation failure of L1-L4 producing focal lumbar kyphosis. The spinal cord ends at L1-L2 level but is abnormally pointed with thickened, tense-appearing filum.* **(Right)** *Axial T1WI MR (segmentation anomalies, clinical tethered spinal cord) confirms fat infiltration of the filum terminale, which is abnormally positioned within the central thecal sac.*

Segmental Spinal Dysgenesis

KEY FACTS

TERMINOLOGY

- Segmental spinal dysgenesis (SSD), caudal regression syndrome (CRS)

IMAGING

- Localized segmental lumbar or thoracolumbar vertebral, spinal cord dysgenesis or agenesis
- Congenital acute angle kyphosis or kyphoscoliosis
- Distal spinal osseous architecture usually normal (unless concurrent CRS)

TOP DIFFERENTIAL DIAGNOSES

- Multiple vertebral segmentation disorders (MVSD)
- Congenital vertebral displacement (CVD)
- Medial spinal aplasia (MSA)
- Caudal regression syndrome

PATHOLOGY

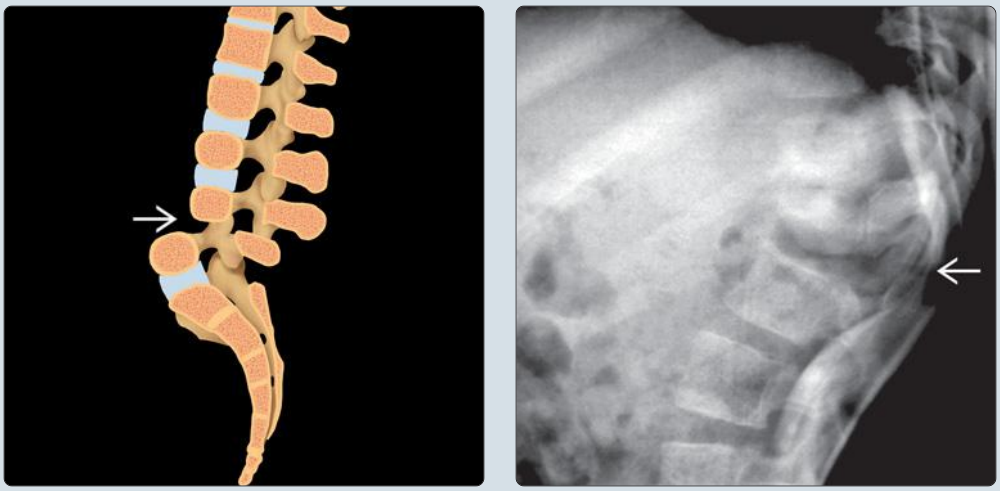
- Characteristic segmental vertebral, cord anomalies
  - Normal upper spinal cord
  - Hypoplastic or absent spinal cord, vertebral dysgenesis at gibbus apex
  - Bulky, thickened, low-lying cord segment within spinal canal below dysgenesis

CLINICAL ISSUES

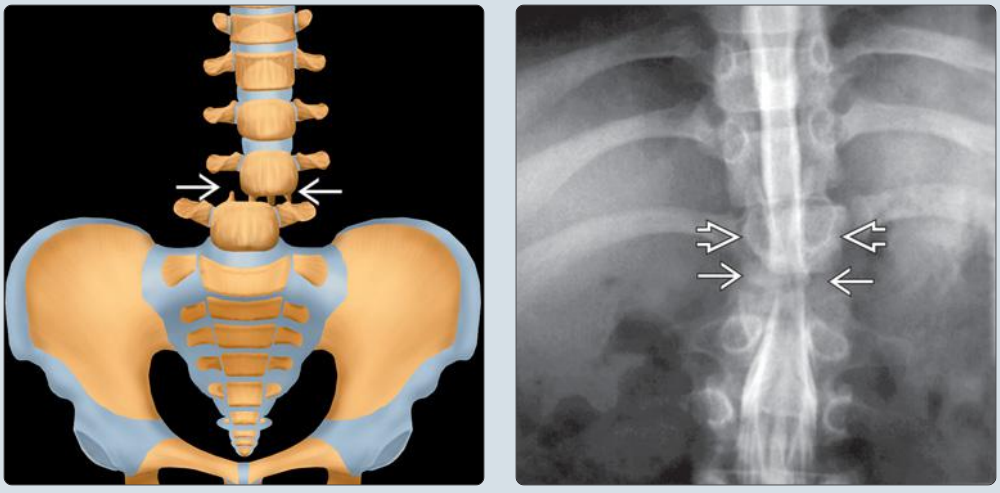
- Thoracic or lumbar kyphosis
- Palpable bone spur at gibbus apex
- Spastic paraparesis or paraplegia
- Rarely present with normal or mildly impaired lower extremity function → subsequent deterioration

DIAGNOSTIC CHECKLIST

- SSD and CRS probably represent 2 different phenotypes along single malformation spectrum
- Morphologic severity correlates with residual spinal cord function, severity of clinical deficit



(Left) *Sagittal graphic demonstrates lower lumbar segmental spinal dysgenesis (SSD), with posterior subluxation of the upper lumbar spine relative to the segment below the dysgenetic level. The spine is dysraphic at the dysgenetic level, but kyphosis has not yet developed.* **(Right)** *Lateral myelography of the thoracolumbar spine shows relatively mild segmental spinal dysgenesis, with focal kyphosis and severe thecal sac narrowing at the hypoplastic level.*



(Left) *Coronal graphic demonstrates lower lumbar segmental spinal dysgenesis with lateral subluxation of the upper spine relative to the lumbosacral spine caudal to the dysgenetic level.* **(Right)** *Anteroposterior myelography shows hypoplasia of the lower thoracic vertebral body above the dysgenetic segment, with narrowing of the myelographic column secondary to spinal canal stenosis. There is also mild focal scoliosis, probably related to segmental instability.*

TERMINOLOGY

Abbreviations

- Segmental spinal dysgenesis (SSD)

Definitions

- Complex congenital spinal malformation
  - Localized segmental agenesis or dysgenesis of lumbar or thoracolumbar spine, spinal cord
  - Distal spinal osseous architecture usually normal (unless concurrent caudal regression syndrome [CRS])

IMAGING

General Features

- Best diagnostic clue
  - Localized segmental spinal agenesis or dysgenesis with severe congenital kyphosis or kyphoscoliosis, segmental spinal cord attenuation
- Location
  - Thoracolumbar, lumbar > lumbosacral
- Size
  - Extent variable
- Morphology
  - SSD is morphologic continuum rather than single appearance
    - Severe: Acute-angle gibbus kyphosis or kyphoscoliosis, indiscernible spinal cord at dysgenetic level, palpable spur at apex
    - Mild: Less severe kyphosis, segmental spinal cord thinning, hypoplastic spinal canal contiguous across dysgenetic segment
  - Classic spinal cord morphology
    - Upper spinal cord normal
    - Spinal cord thinned or atretic, absent nerve roots at dysgenetic level
    - Lower cord below dysgenetic level thickened, low lying
      - Nerve roots directed cephalad

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Sagittal and coronal MR imaging most helpful for evaluating spinal cord
  - Multiplanar and 3D bone CT useful for characterizing vertebral anomalies, surgical planning

Radiographic Findings

- Radiography
  - Moderate to severe congenital kyphosis or kyphoscoliosis
    - Gibbus apex marks dysgenetic level of vertebral malformation
  - Segmental osseous anomalies at dysgenetic level
    - Focal canal stenosis, hypoplastic or aplastic vertebrae, spinal column subluxation (implies instability)
  - Marked stenosis or interruption of spinal canal at malformation level
    - Spinal canal superficially located at gibbus apex
  - ± costal abnormalities (bifid, fused, or absent ribs)
  - ± vertebral formation and segmentation anomalies at other levels

CT Findings

- Bone CT
  - Similar to radiography, but easier to characterize vertebral anomalies

MR Findings

- T1WI
  - Significant narrowing of thecal sac, abnormal spinal cord, absent nerve roots at lesion level
  - Spinal cord always abnormal
  - Low-lying conus medullaris terminates at or below L3
  - Hypoplastic cord segment blends with filum terminale in lumbosacral SSD
- T2WI
  - Same as T1WI

Ultrasonographic Findings

- Grayscale ultrasound
  - Similar to MR findings
  - Diagnostic role limited; may be useful for neonates unable to undergo MR imaging

DIFFERENTIAL DIAGNOSIS

Multiple Vertebral Segmentation Disorders (MVSD)

- Short-trunk dwarfism, variably associated with costal abnormalities
  - Jarcho-Levin syndrome proper, spondylothoracic dysostosis, and spondylocostal dysostosis
- Entire spine abnormal rather than focal dysgenesis

Congenital Vertebral Displacement (CVD)

- Dislocation at single vertebral level → abrupt spinal canal offset
- Vertebral pedicles present in CVD, absent in SSD
- Spinal cord stretched/compressed but otherwise normally developed
- Closed spinal dysraphism unusual
- Neurologic deficits often mild, 2° to spinal instability ± cord compression

Medial Spinal Aplasia (MSA)

- Isolated absence of spinal segments (mainly lower thoracic, lumbar)
- Congenital kyphoscoliosis + paraplegia
- Probably represents severe SSD; use of term "MSA" discouraged

Caudal Regression Syndrome

- Malformation resulting from disturbed development of caudal cell mass
- Dysgenesis or absence of caudal spine, spinal cord
- Isolated or in conjunction with SSD

PATHOLOGY

General Features

- Etiology
  - Cause not definitively known, putative mechanisms include

- Association with maternal diabetes, medications, toxins
- Related to aberrant segmental vascular supply
- Embryologically, may represent inappropriately induced activation of positional apoptosis at dysgenetic level during gastrulation
  - Wrongly specified cells eliminated → few or no prospective neuroectodermal cells induced to form neural plate during primary neurulation
  - Abnormal primary neurulation → hypoplastic or aplastic spinal cord segment
    - Missing nerve roots at abnormal segment may be secondary to apoptotic elimination of somatic motoneurons
  - Neurulation in neural plate proceeds below malformation level → lower cord segment usually present
- SSD and CRS probably represent different phenotypes along single spectrum of segmental spine/spinal cord malformations
  - Segmental location(s) of derangement along embryo longitudinal axis determines malformation type
    - Intermediate segment → SSD
    - Caudal segment → CRS
    - 2 abnormal segments → double level SSD or SSD + CRS

- Associated abnormalities
  - Vertebral formation and segmentation anomalies
  - Closed spinal dysraphism (diastematomyelia, dermal sinus tract, terminal myelocystocele, lipoma, thickened filum terminale)
  - Visceral anomalies (renal, cardiac, neurogenic bladder)
  - Orthopedic anomalies (equinocavovarus feet, lower extremity deformities)
  - Cutaneous stigmata (angiomas, dermal sinus, hairy tuft)

Staging, Grading, & Classification

- Diagnostic criteria
  - Lumbar or thoracolumbar vertebral dysgenesis with kyphosis
  - Focal spinal cord hypoplasia or absence without exiting roots
  - Lower extremity deformity with paraplegia or paraparesis

Gross Pathologic & Surgical Features

- Normal upper spinal cord
- Spinal cord, vertebral malformation located at kyphotic gibbus apex
  - Vertebrae present above and below anomaly
- Bulky, thickened, low-lying cord segment within caudal spinal canal below dysgenesis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Thoracic or lumbar kyphosis with palpable bone spur to gibbus apex
  - Spastic paraparesis or paraplegia
  - Neurogenic bladder
- Other signs/symptoms
  - Rarely present with normal or mildly impaired lower extremity neurological function → subsequent deterioration
- Clinical profile
  - Infant with severely hypotrophic, deformed lower limbs, hip joint flexion-abduction, knee flexion equinocavovarus feet → "Buddha" sitting position
  - Moderate to severe motor impairment

Demographics

- Age
  - Diagnosed during infancy
- Epidemiology
  - Very rare

Natural History & Prognosis

- Progressive kyphosis
- Degree of neurologic deficit dependent on severity of hypoplasia, functionality of residual cord
  - Nearly all patients eventually progress to severe motor impairment

Treatment

- Goals include maintenance of spinal stability, arrest of progressive kyphosis
  - Early spinal fusion ± cord decompression enables patient to sit up without support
  - Bracing less effective than surgical stabilization for arresting neurological decline
- Untethering procedures of limited utility
  - Neurologic dysfunction attributed to segmental hypoplasia or absent spinal cord, roots rather than spinal cord tethering

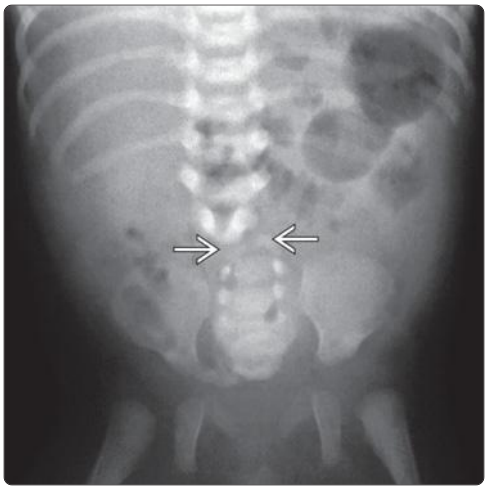
DIAGNOSTIC CHECKLIST

Consider

- Severity of morphologic derangement correlates with residual spinal cord function, severity of clinical deficit
- SSD and CRS probably represent 2 different phenotypes along single malformation spectrum

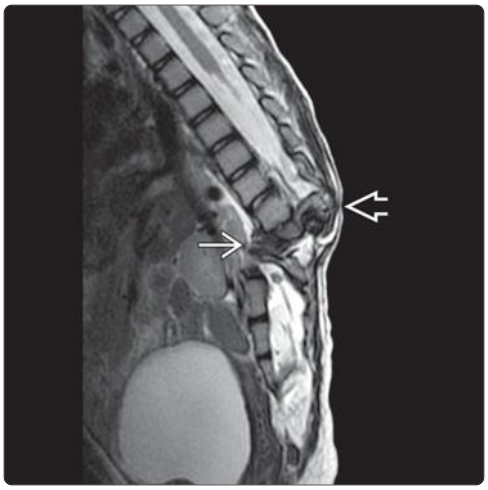
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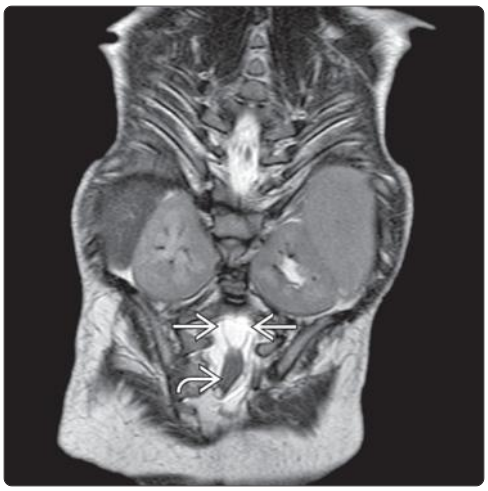


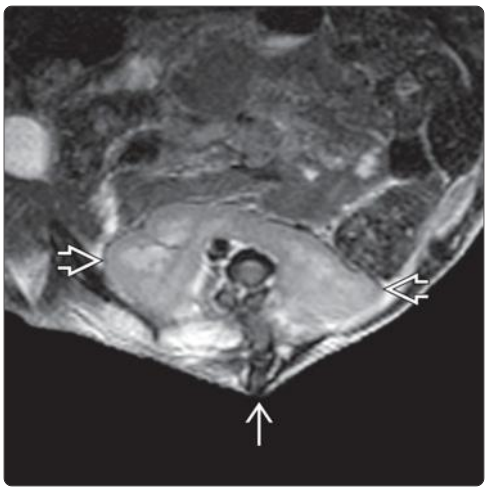
(Left) *Anteroposterior radiograph (lumbosacral SSD) shows lateral subluxation of the upper relative to lower spinal segments related to instability at the dysgenetic level.* **(Right)** *Lateral radiograph of the lumbosacral spine shows posterior subluxation of the upper lumbar spine above the dysgenetic spinal segment relative to the sacrum. The transitional vertebra at the dysgenetic site are hypoplastic and malformed.*





(Left) *Sagittal T2WI MR shows complete disconnection of the spinal canal at the dysgenetic level, with pronounced kyphosis and attenuation of the proximal spinal cord. A clinically palpable spur is conspicuous at the kyphotic apex.* **(Right)** *Sagittal T2WI MR reveals complete disconnection of the spinal canal at the dysgenetic level. There is pronounced kyphosis and prominent bone spur at the gibbus apex. The bulky distal cord is separate from the thoracic cord.*





(Left) *Coronal T2WI MR (severe lumbosacral SSD) shows separation of the distal spinal cord component from the thoracic spinal cord (not shown) above the dysgenetic level. The cephalad-directed nerve roots are commonly observed in lumbosacral SSD.* **(Right)** *Axial T2WI MR (severe lumbosacral SSD) depicts an associated anomalous horseshoe kidney. The prominent midline bone spur was clinically obvious and projects dorsally at the kyphotic gibbus apex.*

KEY FACTS

TERMINOLOGY

- Caudal regression syndrome (CRS), sacral agenesis, lumbosacral hypogenesis

IMAGING

- Constellation of caudal developmental growth abnormalities and associated soft tissue anomalies
- Spectrum ranges in severity from absent coccyx to lumbosacral agenesis
- 2 main types
  - Group 1: Distal spinal cord hypoplasia, severe sacral osseous anomalies
  - Group 2: Tapered, low-lying, distal cord elongation with tethering, less severe sacral anomalies

TOP DIFFERENTIAL DIAGNOSES

- Tethered spinal cord
- Closed spinal dysraphism
- Occult intrasacral meningocele (OIM)

PATHOLOGY

- Group 1: More severe caudal dysgenesis with high-lying, club-shaped cord terminus (decreased number of anterior horn cells)
- Group 2: Less severe dysgenesis with low-lying, tapered, distal cord tethered by tight filum, lipoma, lipomyelomeningocele, or terminal myelocystocele

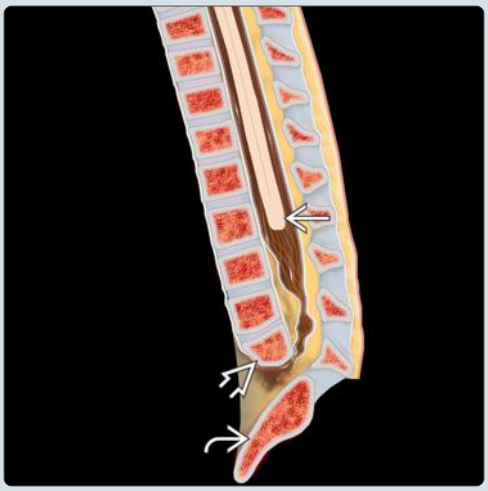
CLINICAL ISSUES

- Clinical spectrum ranges from neurologically normal → severely impaired
- Symptomatic patient presentation spans mild foot disorders → complete lower extremity paralysis and distal leg atrophy

DIAGNOSTIC CHECKLIST

- Look for caudal spine anomalies in patients with genitourinary or anorectal anomalies

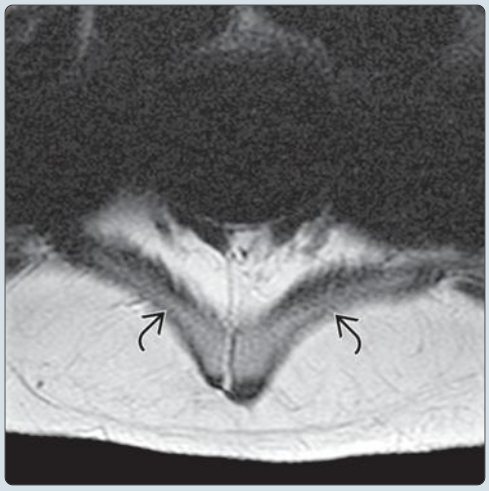
(Left) *Sagittal graphic of the lumbosacral spine in severe group 1 caudal regression syndrome (CRS) shows high truncation of the sacrum and medial position of the iliac wing. The conus terminates abnormally high in a blunt wedge shape.* **(Right)** *Sagittal T2WI MR (group 1 CRS) reveals severe truncation of the sacrum and abnormally high termination of the spinal cord at the T12-L1 vertebral body level with a typical blunted, wedge-shaped conus.*





(Left) *Axial T2WI MR (group 1 CRS) obtained immediately below the conus level demonstrates abrupt conus termination with relatively normal-appearing ventral and dorsal cauda equina nerve roots.* **(Right)** *Axial T2WI MR of the pelvis (severe group 1 CRS) demonstrates bilateral abnormal hypoplastic iliac wings closely approximated in the midline secondary to complete absence of a normal sacrum at this level.*





TERMINOLOGY

Abbreviations

- Caudal regression syndrome (CRS)

Synonyms

- Sacral agenesis, lumbosacral hypogenesis

Definitions

- Constellation of caudal developmental growth abnormalities and associated soft tissue anomalies

IMAGING

General Features

- Best diagnostic clue
  - Incomplete lumbosacral spine formation
  - Abnormal distal spinal cord
- Location
  - Lumbosacral spine
- Size
  - Variable diminution of caudal spine
- Morphology
  - Spectrum ranging in severity from absent coccyx to lumbosacral agenesis
    - Partial or total unilateral dysgenesis with oblique lumbosacral joint
    - Bilateral total lumbosacral hypogenesis; vertebral column terminates in thoracic spine
  - Caudal vertebral bodies are often fused
  - Severe canal narrowing rostral to last intact vertebra
    - Osseous vertebral excrescences, fibrous bands connecting bifid spinous processes, or severe distal dural tube stenosis

Radiographic Findings

- Radiography
  - Lumbosacral osseous hypogenesis

CT Findings

- CECT
  - Lumbosacral dysgenesis with distal spinal stenosis
  - ± prominent nerve/dorsal root ganglion (DRG) enhancement

MR Findings

- T1WI
  - Vertebral body dysgenesis/hypogenesis
  - Group 1: Distal spinal cord hypoplasia (wedge-shaped cord termination), severe sacral osseous anomalies
    - ± dilated central canal, conus CSF cyst
  - Group 2: Tapered, low-lying, distal cord elongation with tethering, less severe sacral anomalies
- T2WI
  - Same findings as T1WI; best for depicting dural stenosis
- T1WI C+
  - Hypertrophied DRG/nerve roots may enhance

Ultrasonographic Findings

- Grayscale ultrasound
  - Group 1: Blunt spinal cord termination above L1; central canal may be prominent
  - Group 2: Elongated conus with thick filum ± intraspinal lipoma

Nonvascular Interventions

- Myelography
  - Caudal hypogenesis ± dural stenosis; most useful in conjunction with CT

Imaging Recommendations

- Protocol advice
  - Consider US for infant screening if MR is not possible
  - MR to confirm US findings, treatment planning
    - Sagittal MR to demonstrate extent of lumbosacral deficiency, distal spinal cord morphology, and presence/absence of tethering/lipoma
    - Axial MR to detect osseous spinal narrowing, hydromyelia, and associated lesions

DIFFERENTIAL DIAGNOSIS

Tethered Spinal Cord

- Low-lying spinal cord ± thickened or fatty filum, no caudal dysgenesis
- Difficult to clinically discern from mild sacral dysgenesis
  - ± associated imaging abnormalities that may help distinguish

Closed Spinal Dysraphism

- Dorsal dysraphism without vertebral column hypogenesis (e.g., lipomyelomeningocele)

Occult Intrasacral Meningocele (OIM)

- Sacrum is thinned and remodeled, sometimes imitating caudal regression
- Currently considered an extradural arachnoid cyst rather than true meningocele

PATHOLOGY

General Features

- Etiology
  - Normal caudal spine development → canalization and retrogressive differentiation (secondary neurulation)
    - Anorectal and genitourinary structures form contemporaneously in close anatomic proximity
  - Insult prior to 4th gestational week → caudal cell mass developmental abnormalities
    - Hyperglycemia, infectious, toxic, or ischemic insult postulated to impair spinal cord and vertebral formation
    - Signaling defects by retinoic acid and sonic hedgehog during blastogenesis and gastrulation
    - Abnormal neural tube, notochord development → impaired migration of neurons and mesodermal cells
- Genetics
  - Most cases sporadic
  - Dominantly inherited form recently described; defect in *HLXB9* (*MNX1*) homeobox gene (chromosome 7)
    - *HLXB9* (*MNX1*) also expressed in pancreas → possible association between diabetes hyperglycemia and caudal regression
- Associated abnormalities
  - Tethered cord

  - ~ 100% of CRS patients with conus terminating below L1
  - Thickened filum (65%) ± dermoid or lipoma
- Other spinal anomalies
  - Vertebral anomalies (22%), diastematomyelia, terminal hydromyelia (10%), myelomeningocele (35-50%), lipomyelomeningocele (10-20%), terminal myelocystocele (15%), anterior sacral meningocele, terminal lipoma (< 10%)
- Congenital cardiac defects (24%), pulmonary hypoplasia
- Genitourinary abnormalities (24%)
  - Renal agenesis/ectopia, hydronephrosis, müllerian duct malformations, urinary bladder malformation
- Anorectal anomalies (particularly anal atresia)
  - Higher level of anal atresia → more severe lumbosacral dysgenesis, genitourinary anomalies
- Currarino triad: Partial sacral agenesis with intact 1st sacral vertebra, presacral mass, anorectal malformations
- Orthopedic abnormalities; extreme cases → lower extremity fusion (sirenomelia)

- Sequela of caudal cell mass dysplasia with spectrum of severity
- Lower extremity deformities, lumbosacral agenesis, anorectal abnormalities, renal/pulmonary hypoplasia characteristic
  - Most severe cases → sirenomelia ("mermaid syndrome")
  - 20% → tethering subcutaneous lesions (group 2)

Staging, Grading, & Classification

- Group 1: More severe caudal dysgenesis with high-lying, club-shaped cord terminus (↓ number of anterior horn cells)
  - Distal cord hypoplasia with wedging is seen in all patients with partial or complete dysgenesis and termination of spinal cord above L1
  - Termination of conus above L1 is highly correlated with sacral malformations ending at S1 or above
- Group 2: Less severe dysgenesis with low-lying, tapered, distal cord tethered by tight filum, lipoma, lipomyelomeningocele, or terminal myelocystocele
  - Conus termination below L1 highly correlated with sacral malformations ending at S2 or below
  - Cord tethering more common in milder sacral dysgenesis

Gross Pathologic & Surgical Features

- Severity of vertebral dysgenesis, presence/absence of tethering, and osseous canal diameter impact surgical planning

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neurogenic urinary bladder dysfunction (nearly all patients)
  - Sensorimotor paresis (group 2 > group 1)
    - Severity of motor deficit > sensory
    - Sacral sensory sparing common, even in severe cases
- Other signs/symptoms
  - Neurologically asymptomatic (group 1 > group 2)
- Clinical profile
  - Broad spectrum ranging from neurologically normal to complete lumbosacral agenesis and fusion of lower extremities (sirenomelia)
    - Always accompanied by narrow hips, hypoplastic gluteal muscles, shallow intergluteal cleft
  - Symptomatic patient presentation ranges from mild foot disorders to complete lower extremity paralysis and distal leg atrophy
    - Motor level usually higher than sensory level
    - Level of vertebral aplasia correlates with motor but not sensory level

Demographics

- Age
  - Severe cases identified in utero (obstetrical US) or at birth
  - Mild cases may not be identified until adulthood
- Gender
  - M = F
- Epidemiology
  - 1/7,500 births (milder forms > severe forms)
  - 15-20% are infants of diabetic mothers; 1% of offspring from diabetic mothers affected
  - Association with VACTERL (10%), omphalocele, exstrophy bladder, imperforate anus, spinal anomalies (10%), and Currarino triad syndromic complexes

Natural History & Prognosis

- Variable depending on severity

Treatment

- Surgical untethering if clinically symptomatic
- Surgical release/duraplasty may improve neurological function in patients with distal spinal canal stenosis
- Orthopedic procedures to improve lower extremity functionality

DIAGNOSTIC CHECKLIST

Consider

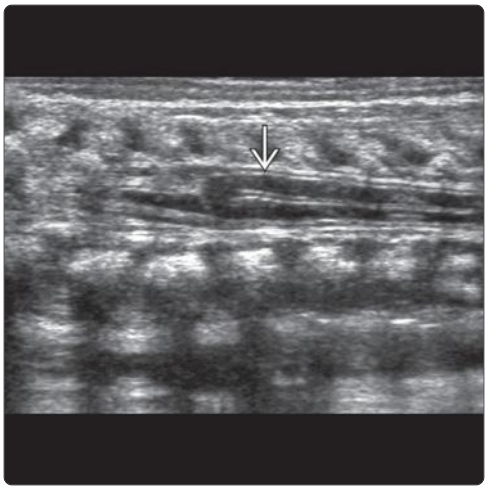
- MR most useful imaging tool for characterizing abnormalities and surgical planning

Image Interpretation Pearls

- Caudal spine anomalies should be sought out in patients with genitourinary or anorectal anomalies and vice versa

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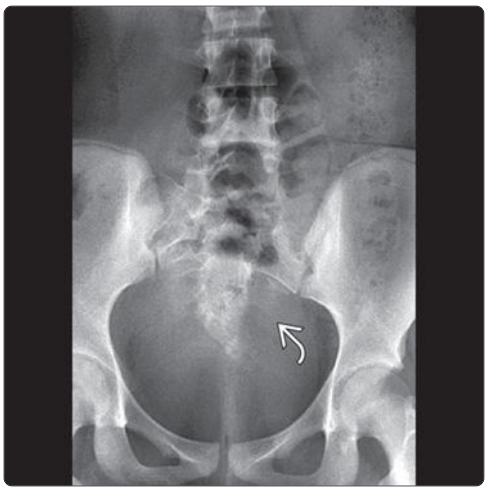
(Left) *Longitudinal spinal ultrasound (group 1 CRS) shows mild distal spinal cord thinning with blunt wedge- or club-shaped termination ➡ of the spinal cord (at T1 level, confirmed with radiography).* **(Right)** *Anteroposterior abdominal radiograph (group 1 CRS) reveals symmetric hypoplasia of the iliac wings ➡ and S1 vertebra with absence of the remaining caudal sacral vertebra. Note also the sternal wire following surgery for congenital heart disease, an associated anomaly.*





(Left) *Sagittal T2WI MR (group 2 CRS) demonstrates a milder variant of CRS. In this case, there are 4 sacral vertebra with absence of S5 and the coccyx. The spinal cord ➡ is low-lying and taut, features suggesting that it is probably tethered.* **(Right)** *Sagittal T1WI MR (group 2 CRS) depicts mild sacral dysgenesis and posterior spinal dysraphism, with the abnormal low-lying tapered spinal cord terminating ➡ into a large terminal lipoma ⇨.*





(Left) *Sagittal T2WI MR (group 2 CRS) reveals sacral dysgenesis with barely 2 sacral vertebra. The spinal cord ➡ is low-lying with prominent conus syrinx. Also confirmed was a parasagittal presacral cystic mass (anterior sacral meningocele) ➡ with a thin, fluid-filled tract ⇨ communicating to the thecal sac.* **(Right)** *Anteroposterior radiograph of the pelvis demonstrates partial absence of the left sacral ala ➡ in a patient with hemisacral agenesis variant of caudal regression.*

KEY FACTS

TERMINOLOGY

- Synonym: Terminal syringocele

IMAGING

- Complex spinal malformation → closed spinal dysraphism, large skin-covered back mass
- Hydromyelic, low-lying tethered spinal cord traverses dorsal meningocele and terminates in dilated terminal cyst (myelocystocele)
- Multiplanar MR imaging best demonstrates constellation of abnormalities

TOP DIFFERENTIAL DIAGNOSES

- Anterior sacral meningocele
- Simple dorsal meningocele
- Sacrococcygeal teratoma
- Myelomeningocele

PATHOLOGY

- Results from deranged secondary neurulation of caudal cell mass
- Associated malformations: Cloacal exstrophy, imperforate anus, omphalocele, pelvic deformities, equinovarus, ambiguous hypoplastic genitalia, and renal abnormalities

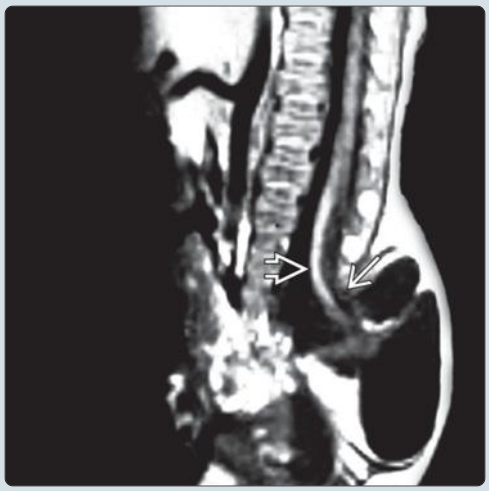
CLINICAL ISSUES

- Presents at birth with large skin-covered back mass
- Usually neurologically intact at birth; may later develop lower extremity sensorimotor deficits

DIAGNOSTIC CHECKLIST

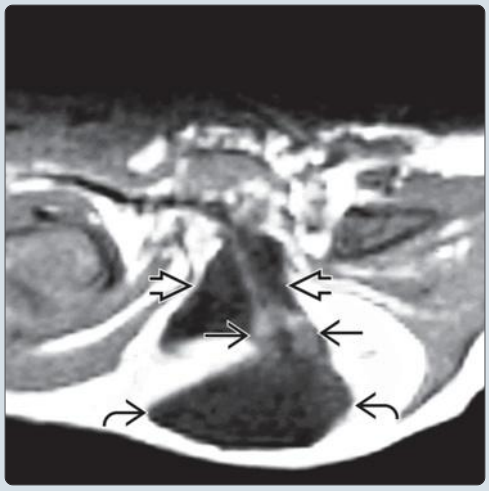
- Early diagnosis and surgery → best chance for normal neurological outcome
- Nonneurological prognosis largely linked to severity of associated anomalies





(Left) *Sagittal graphic displays a low-lying, hydromyelic spinal cord piercing an expanded subarachnoid space (meningocele), terminating in a myelocystocele.* **(Right)** *Sagittal T1WI MR depicts a low-lying hydromyelic spinal cord traversing a meningocele and expanding into a large terminal cyst. Note the dorsal fibrous band. Note that the back mass is caused by both the meningocele and myelocystocele in this case.*





(Left) *Coronal T1WI MR demonstrates the meningocele component of the terminal myelocystocele malformation. Note also the solitary hypertrophic right kidney. The coronal imaging plane is particularly useful to evaluate for associated visceral anomalies.* **(Right)** *Axial T1WI MR shows the passage of bifid hydromyelic, low-lying cord through the arachnoid-lined meningocele to terminate in a myelocystocele. Note classic "trumpet flaring" of the terminal spinal cord.*

TERMINOLOGY

Synonyms

- Terminal syringocele

Definitions

- Complex spinal malformation → hydromyelic, low-lying tethered spinal cord traverses meningocele to terminate in myelocystocele

IMAGING

General Features

- Best diagnostic clue
  - Hydromyelic tethered cord traversing dorsal meningocele to terminate in dilated terminal ventricle cyst
- Location
  - Sacrum/coccyx
- Size
  - Mass varies from small to large (> 10 cm diameter)
- Morphology
  - Closed spinal dysraphism, large skin-covered mass, terminal cord cyst traversing meningocele

Radiographic Findings

- Radiography
  - Lumbosacral dysraphism ± soft tissue mass
  - ± pubic diastasis (usually + bladder exstrophy)

CT Findings

- NECT
  - Lumbosacral dysraphism
  - Hydromyelic cord penetrates meningocele sac, ends in caudal terminal cystic sac

MR Findings

- T1WI
  - Hypointense cephalic dorsal meningocele → back mass
  - Tethered distal cord shows trumpet-like flaring into hypointense caudal terminal cyst
- T2WI
  - Hypointense dorsal fibrous band at cephalic margin of meningocele constrains spinal cord
  - Fibrolipomatous tissue surrounds both sacs

Ultrasonographic Findings

- Grayscale ultrasound
  - Sagittal plane: Spinal cord passes through hypoechoic, dilated subarachnoid space (meningocele), terminates in cord cyst
  - Axial plane: Bifid hydromyelic dorsal spinal cord within meningocele

Nonvascular Interventions

- Myelography
  - Hydromyelic cord passes through meningocele, terminates in separate sac caudal to contrast-filled meningocele
  - Delayed imaging → ± contrast imbibition into hydromyelic cord/terminal cyst

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Sagittal MR for diagnosis and estimating length of hydromyelia, sizing cysts, and identifying associated abnormalities
  - Axial MR to clarify extent of rachischisis, evaluate associated anomalies

DIFFERENTIAL DIAGNOSIS

Anterior Sacral Meningocele

- Anterior meningocele protrudes thorough enlarged sacral neural foramen into pelvis

Simple Dorsal Meningocele

- Dorsal meningocele protrudes thorough focal dysraphism
- Cord rarely tethered or hydromyelic

Sacrococcygeal Teratoma

- Similar clinical appearance of skin mass
- Distinguish by presence of solid tumor elements within cysts, calcifications

Myelomeningocele

- Open spinal dysraphism; no skin covering, clinically obvious

PATHOLOGY

General Features

- Etiology
  - Postulated to result from deranged secondary neurulation of caudal cell mass; typically associated with other caudal tail-fold malformations
    - CSF unable to normally exit from early neural tube → terminal ventricle balloons into cyst → disrupts overlying dorsal mesenchyme but not superficial ectoderm
    - Posterior elements do not form normally → spina bifida with intact skin
    - Terminal ventricle dilatation → distends arachnoid cord lining → meningocele
    - Cyst bulk prevents cord ascent → tethered cord
    - Progressive distention of distal cord causes caudal bulge below end of meningocele into extraarachnoid space and results in flaring of distal spinal cord
    - Disruption of caudal motor segments → progressive symptoms that present at birth or appear later
  - Some speculate → most severe manifestation of persistent terminal ventricle (ventriculus terminalis) spectrum
- Associated abnormalities
  - Cloacal exstrophy, imperforate anus, omphalocele, pelvic deformities, equinovarus, ambiguous hypoplastic genitalia, and renal abnormalities
  - Syndromal associations
    - Caudal regression syndrome
    - OEIS syndrome constellation (omphalocele, exstrophy of bladder, imperforate anus, and spinal anomalies)
  - Chiari 1 and 2 malformations, hydrocephalus, and vertebral segmentation anomalies (rare)
- Classic pathological triad

- Skin-covered lumbosacral dysraphism
- Arachnoid-lined meningocele directly contiguous with spinal subarachnoid space
- Low-lying hydromyelic cord traverses meningocele, expands into large contiguous ependymal-lined terminal cyst

Gross Pathologic & Surgical Features

- Lumbosacral dysraphism with hypoplastic, widely everted laminae
- Proximal (smaller, rostral) sac resembles typical meningocele, with inner surface lined by arachnoid and thick fibrous layer
- Distal spinal cord herniates under fibrous band between medial ends of most cephalic widely bifid lamina, traverses meningocele → terminal cyst (larger, caudal)
  - Cord narrowed by fibrous band where it exits spinal canal, then widens distally due to concurrent hydromyelia
  - Distal spinal nerve roots arise from ventral surface of intraarachnoid segment of spinal cord, traverse meningocele, and reenter spinal canal before exiting at their root sleeves or via bony clefts
- Terminal cyst does not communicate directly with subarachnoid space

Microscopic Features

- Meningocele lined by arachnoid, thick fibrous layer
- Terminal cyst lined by ependyma, dysplastic glia; directly contiguous with central cord canal
- Outer cord surface pia, arachnoid continuous with meningocele

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Presents at birth with large skin-covered back mass
    - Skin appears normal or exhibits hemangioma, nevus, or hypertrichosis
    - Rare patients without back mass present later with progressive neurological deficits
  - Majority neurologically intact at diagnosis
- Other signs/symptoms
  - Later presentation or untreated lesion may develop progressive lower extremity paresis
- Clinical profile
  - Usually neurologically intact at birth, but may present with lower extremity sensorimotor deficits
  - Large mass obliterates intergluteal cleft, extends upward from perineum for variable distance
  - ± concurrent midline cecal and paramedian bladder exstrophy, other visible anomalies

Demographics

- Age
  - Infancy
- Gender
  - F > M
- Epidemiology
  - Rare: 1-5% of skin-covered lumbosacral masses
    - Much more common in cloacal exstrophy patients
  - Sporadic; no familial incidence
  - Postulated associations with teratogens retinoic acid, hydantoin, loperamide hydrochloride
  - No known association with diabetes mellitus (unlike caudal regression syndrome)

Natural History & Prognosis

- Normal intellectual potential
- Mass size and deficits tend to progress with time, may be partially or totally reversible with prompt surgical repair
  - Main goals of neurosurgical intervention are to reduce mass size and untether cord
  - Operation soon after diagnosis indicated to prevent progression of neurological abnormalities, cyst growth
  - Persistent neurological deficits usually permanent
- Overall prognosis mainly related to other associated anomalies (OEIS constellation)
  - Patients without abdominal wall defects have more favorable neurological outcome than those with ventral wall defects

Treatment

- Early diagnosis and surgical repair maximizes probability of normal neurological outcome
- Delayed recognition and operation increase odds of onset, progression of lower extremity paresis

DIAGNOSTIC CHECKLIST

Consider

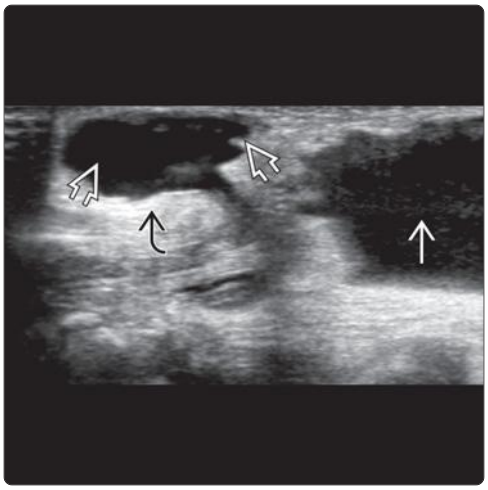
- Early diagnosis and surgery → best chance for normal neurological outcome
- Nonneurological prognosis largely linked to severity of associated anomalies

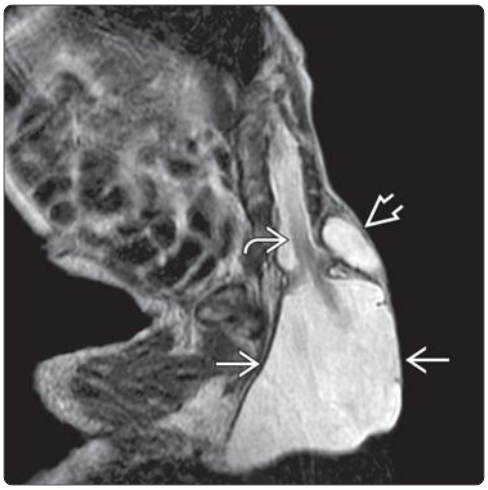
Image Interpretation Pearls

- Tethered hydromyelic cord traversing meningocele to terminate in separate caudal cyst is unique imaging finding diagnostic of terminal myelocystocele

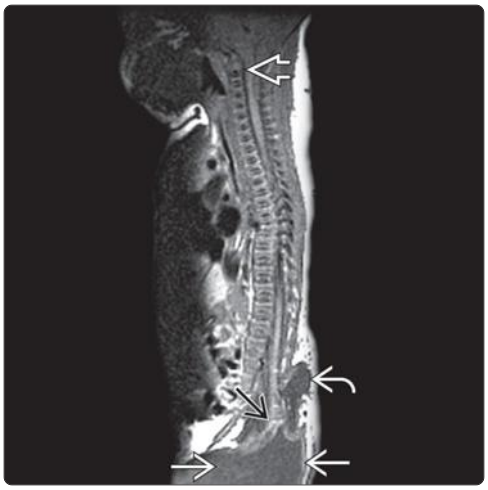
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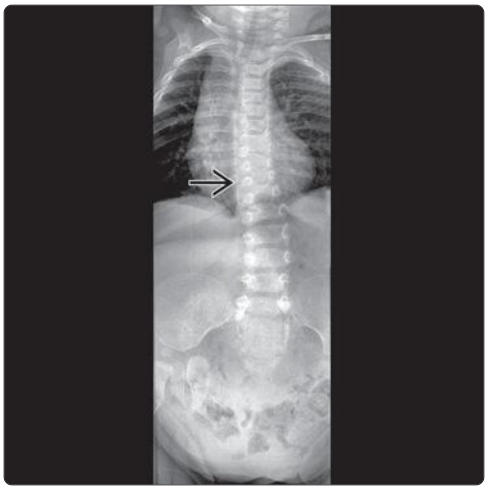
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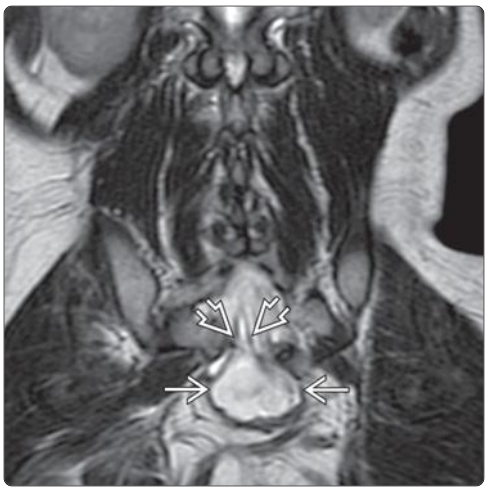
(Left) *Longitudinal ultrasound shows the low-lying spinal cord ending in a terminal cyst that is contiguous with the central spinal cord canal. The spinal cord passes through a dilated subarachnoid space representing the meningocele component.* **(Right)** *Sagittal T2WI MR demonstrates the low-lying spinal cord crossing the meningocele to terminate in a large myelocystocele cyst, the primary contributor to the clinically obvious skin-covered back mass in this case.*





(Left) *Sagittal T1WI MR shows central canal of low-lying cord traversing the meningocele before expanding into a large terminal cyst. Note "trumpet" splaying of the terminal spinal cord. This infant also has an associated Chiari 1 malformation.* **(Right)** *Anteroposterior radiograph in a patient with terminal myelocystocele shows associated sacral dysgenesis, wide symphysis pubis (bladder exstrophy), unfused posterior sacral elements, and a midthoracic hemivertebra.*





(Left) *Sagittal FIESTA cine MR demonstrates truncation of the sacrum. The spinal cord is abnormally low-lying with caudal extension of the attenuated cord through a sacral cyst (meningocele). The terminal spinal cord flares into a second terminal cyst within the meningocele ("cyst in a cyst") completing the terminal myelocystocele malformation.* **(Right)** *Coronal T2WI MR demonstrates flaring of terminal spinal cord and transition of central canal into terminal myelocystocele.*

Anterior Sacral Meningocele

KEY FACTS

TERMINOLOGY

- Anterior sacral meningocele (ASM)
- Sacral meninges herniate anteriorly into pelvis through focal erosion or hypogenesis of sacral ± coccygeal vertebral segments

IMAGING

- Presacral cyst, contiguous with thecal sac through anterior osseous defect
- Deficient sacrum ± curved (scimitar) shape

TOP DIFFERENTIAL DIAGNOSES

- Sacrococcygeal teratoma (SGT)
- Sacral chordoma
- Neurenteric cyst
- Cystic neuroblastoma
- Ovarian cyst

PATHOLOGY

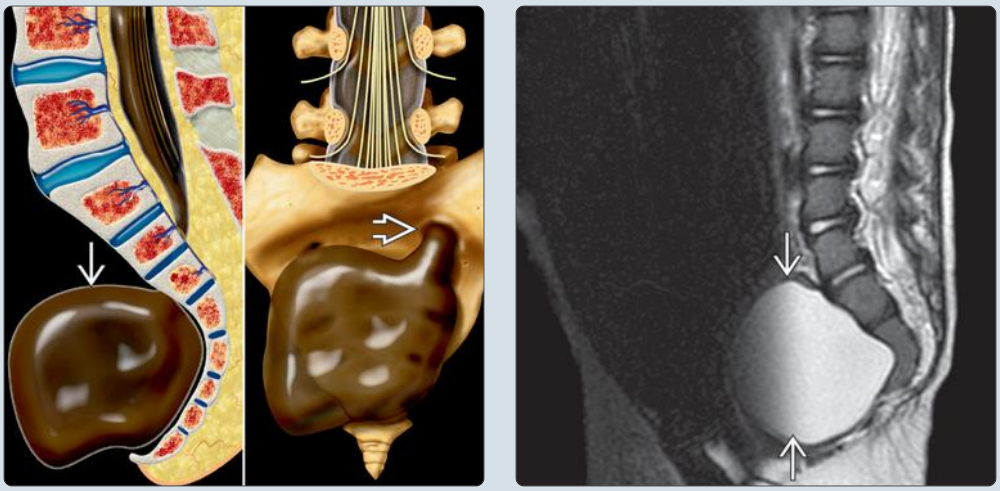
- Currarino triad: Anorectal anomalies, caudal regression syndrome, epidermoid/dermoid tumor or other tethering lesion

CLINICAL ISSUES

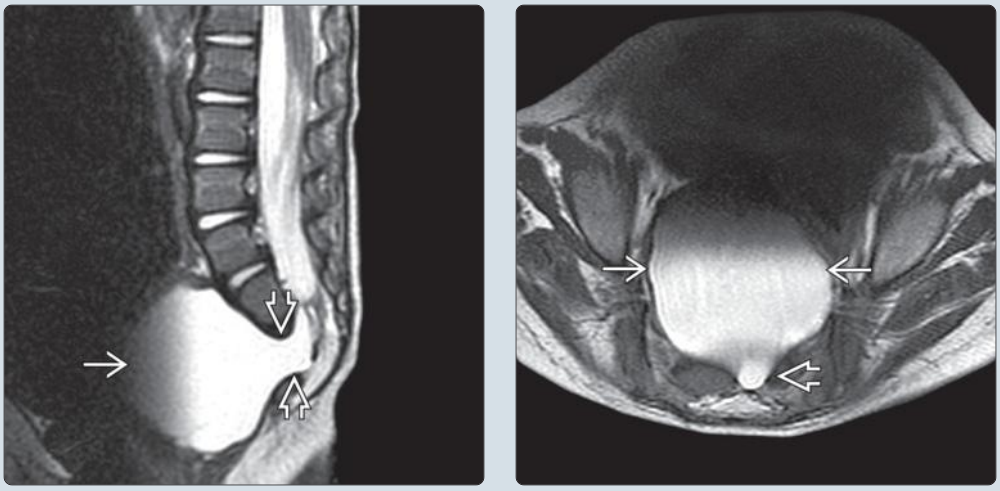
- Constipation, urinary frequency, incontinence, dysmenorrhea, dyspareunia, low back/pelvic pain

DIAGNOSTIC CHECKLIST

- Continuity of cyst with thecal sac necessary to ensure ASM diagnosis
- Soft tissue mass or calcification implies tumor
- Imaging recommendations
  - Ultrasound for NICU infant screening
  - MR to confirm presacral cyst location and characterize contents



(Left) *Sagittal graphic (left) depicts the characteristic anterior sacrum "scimitar" remodeling by a large anterior meningocele cyst. Coronal graphic (right) shows an anterior sacral meningocele cyst origin through an enlarged neural foramen.* **(Right)** *Midline sagittal T2WI MR demonstrates the classic relationship of characteristic "scimitar sacrum" configuration with the adjacent presacral cyst (anterior sacral meningocele [ASM]).*



(Left) *Sagittal T2WI MR obtained off-midline parasagittal shows a large anterior presacral cyst contiguous with the meninges, extending from the thecal sac into the pelvis through an enlarged neural foramen.* **(Right)** *Axial T2WI MR confirms unilateral left sacral neural foraminal enlargement permitting ventral herniation of the meninges into the pelvis to form a classic ASM.*

TERMINOLOGY

Abbreviations

- Anterior sacral meningocele (ASM)

Definitions

- Sacral meninges herniate anteriorly into pelvis through focal erosion or hypogenesis of sacral ± coccygeal vertebral segments

IMAGING

General Features

- Best diagnostic clue
  - Presacral cyst, contiguous with thecal sac, protruding through anterior osseous defect
- Location
  - Sacrum/coccyx
- Size
  - Variable
- Morphology
  - Uni- or multiloculated cyst; usually devoid of neural tissue but may have traversing nerve roots
  - Sacral defect 2° to neural foraminal widening
    - May be unilateral or bilateral, symmetrical or asymmetrical, single or multiple level

Radiographic Findings

- Radiography
  - Widened sacral canal and neural foramina
  - Scalloping of anterior sacrum wall
    - Curved "scimitar" sacrum on lateral projection
  - ± scoliosis

CT Findings

- CECT
  - Deficient sacrum with variably sized anterior cyst
    - ± nerve roots traversing sacral defect
  - ± lipoma/dermoid (hypodense)
  - Absence of enhancement

MR Findings

- T1WI
  - Deficient sacrum with variably sized presacral cyst
    - Sagittal images confirm cyst/thecal sac continuity
  - ± cord tethering, lipoma/dermoid (hyperintense)
- T2WI
  - Homogeneous, hyperintense CSF signal intensity cyst (similar to CSF in thecal sac)
  - Best sequence to identify nerve roots traversing sacral defect
- T2* GRE
  - No hypointensity (calcification) within cyst wall
    - Helps distinguish from sacrococcygeal teratoma
- DWI
  - Hypointense signal (↑ diffusivity) confirms CSF composition, excludes epidermoid cyst (hyperintense)
- T1WI C+
  - Absence of enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Cystic hypoechoic pelvic mass anterior to sacrum
  - Complex internal echoes if prior/concurrent inflammation or infection

Nonvascular Interventions

- Myelography
  - Thecal sac communicates with cyst
    - Most useful in conjunction with CT post intrathecal contrast
  - Rarely used unless MR contraindicated or nondiagnostic

Imaging Recommendations

- Best imaging tool
  - MR confirms cyst contiguity with thecal sac
    - T1WI shows epidermoid, lipoma/dermoid if present
    - T2WI demonstrates ± entrapped neural tissue
  - CT imaging best depicts osseous defect, absence of rim calcification (that would imply sacrococcygeal teratoma)
- Protocol advice
  - Ultrasound for initial screening in NICU infants
  - Sagittal and axial MR to confirm positive ultrasound studies, preoperative planning, and postoperative surveillance

DIFFERENTIAL DIAGNOSIS

Sacrococcygeal Teratoma

- Cystic sacrococcygeal teratoma may be difficult to distinguish from ASM
- Look for soft tissue mass, enhancement, calcification

Sacral Chordoma

- Mixed solid/cystic destructive sacral mass
  - No communication with thecal sac
- Rare in children; peak incidence: 5th-6th decade
- Bone CT shows destructive margins

Neurenteric Cyst

- Cyst usually within spinal canal ± rachischisis, vertebral formation anomalies
- Often proteinaceous; T1 hyperintense

Cystic Neuroblastoma

- Search for calcifications, metastatic lesions

Ovarian Cyst

- Ultrasound may reveal surrounding ovarian tissue that clinches diagnosis

PATHOLOGY

General Features

- Etiology
  - Embryogenesis not definitively known; classified within caudal mass anomalies spectrum
    - Erosion or dysgenesis of sacral/coccygeal segments permits herniation of meningeal sac into anterior pelvis
    - 5% of retrorectal tumors
  - Possible associations
    - Simple form: Marfan, NF1
    - Complicated form: Familial, partial sacral agenesis, imperforate anus or anal stenosis, and tethered cord

- Genetics
  - Sporadic (most patients)
  - Minority show inherited predisposition
    - Currarino triad: Autosomal dominant with variable penetration (*HLXB9* [*MNX1*] gene, chromosome 7q36)
    - Conditions where dural ectasia is prominent (NF1, Marfan, homocystinuria)
- Associated abnormalities
  - Currarino triad: Anorectal anomalies, caudal regression syndrome, epidermoid/dermoid tumor or other tethering lesion
    - 1st described in 1837

Gross Pathologic & Surgical Features

- Osseous sacral remodeling in large meningoceles → classic "scimitar" shape
- Widened sacral dural sac communicates with pelvic cyst through narrow neck within sacral defect
- ± neural tissue within cyst sac

Microscopic Features

- Characteristic features of dural sac tissue
- May reveal signs of prior inflammation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Constipation, urinary frequency, incontinence, dysmenorrhea, dyspareunia, low back/pelvic pain
    - 2° to pressure on pelvic viscera
- Other signs/symptoms
  - Sciatica, diminished rectal/detrusor tone, numbness/paresthesias in lower sacral dermatomes (nerve root pressure)
  - Intermittent positional high- or low-pressure headaches due to fluid shifts between ASM and spinal subarachnoid space
  - Superinfection ± meningitis (uncommon)
- Clinical profile
  - Most present with urinary or bowel complaints; ASM is discovered during imaging evaluation
  - Rare finding in (low-pressure) headache patient

Demographics

- Age
  - Onset of symptoms in 2nd-3rd decade
- Gender
  - M = F (children)
  - M < F (adults)
- Epidemiology
  - Rare; less common than dorsal meningoceles

Natural History & Prognosis

- Good prognosis following successful surgical repair

Treatment

- Open posterior transsacral approach, patching of meningocele-dural sac connection
  - Easier if cyst does not contain neural elements, permitting simple ligation of cyst neck
- Endoscopic approach → lower morbidity

DIAGNOSTIC CHECKLIST

Consider

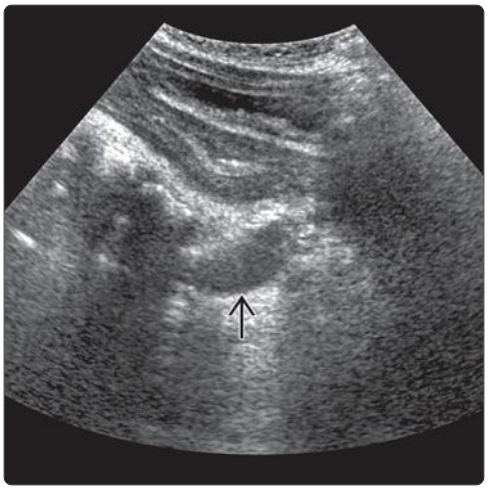
- Ultrasound for infant screening
- MR to identify and characterize cyst location and contents, confirm abnormal ultrasound findings
- Bone CT to demonstrate characteristic osseous findings that permit specific diagnosis

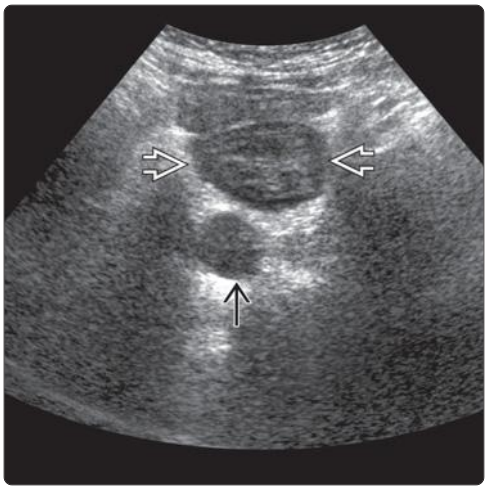
Image Interpretation Pearls

- Continuity of cyst with thecal sac necessary to ensure diagnosis
- Presence/absence of neural tissue within cyst important for surgical planning
- Soft tissue mass or calcification implies tumor

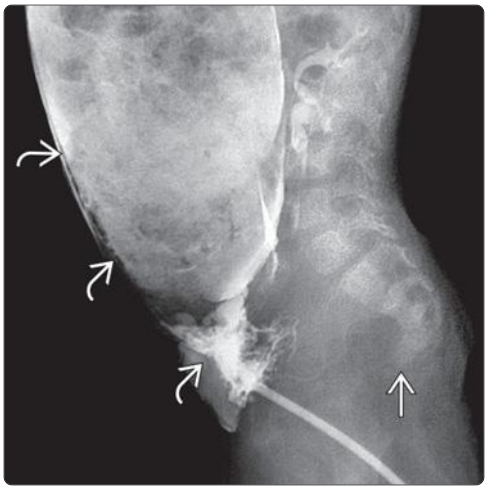
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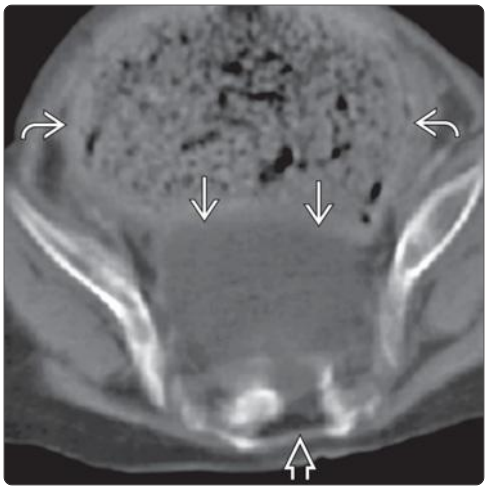
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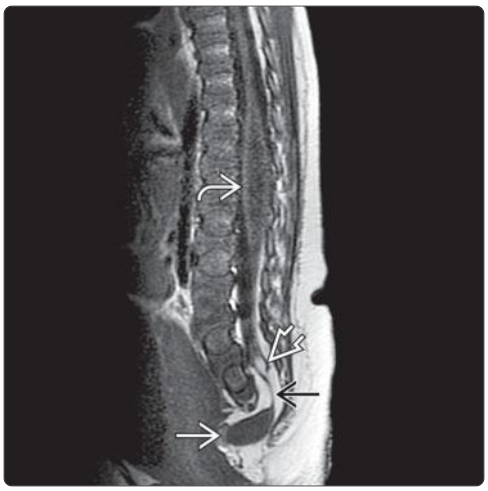


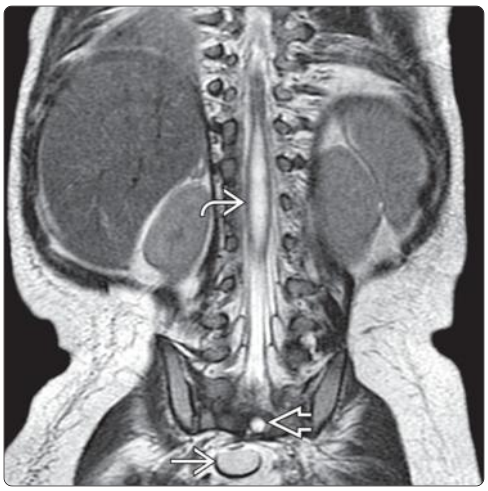
(Left) *Longitudinal ultrasound (performed in supine position to exclude intussusception in clinical context of bloody stool) reveals an unexpected cystic extraperitoneal presacral fluid collection ⇨ representing an ASM behind the rectum and below the peritoneal reflection.* **(Right)** *Transverse ultrasound (in the same patient and performed to exclude intussusception in clinical context of bloody stool) confirms a presacral ASM ⇨ located behind the rectum ➡ and below peritoneal reflection.*





(Left) *Lateral radiography obtained following barium enema (performed for severe constipation) reveals the characteristic "scimitar" sacrum ➡ shape associated with anterior sacral meningocele. Note the huge obstructed large bowel ➡ opacified by barium.* **(Right)** *Axial bone CT shows a large anterior sacral cyst ➡ contiguous with a unilateral enlarged left sacral neural foramen ➡. Note the anterior displacement of the markedly dilated rectum ➡ in this patient with severe constipation.*





(Left) *Sagittal T1WI MR detects sacral dysgenesis with 2 sacral vertebra. The low-lying spinal cord shows a syrinx ➡ and terminal lipoma ➡. Also confirmed was a parasagittal presacral cystic mass ➡ with thin fluid-filled tract ⇨ communicating with the thecal sac through an enlarged sacral foramina.* **(Right)** *Coronal T2WI MR confirms a presacral cystic mass ➡ communicating with the thecal sac through an enlarged left S2 neural foramen ➡. Note also the terminal conus syrinx ➡.*

Sacral Extradural Arachnoid Cyst

KEY FACTS

TERMINOLOGY

- Spinal extradural arachnoid cyst (AC) located within sacrum, caudal to thecal sac termination
  - Sacral meningeal cyst, type IB meningeal cyst
- Synonym: Occult intrasacral meningocele (outdated term)

IMAGING

- Smooth enlargement of sacral spinal canal
  - Expands but does not transgress sacrum margins
  - No expansion or remodeling of neural foramina
- Posterior sacral vertebral scalloping characteristic
- No cyst wall enhancement

TOP DIFFERENTIAL DIAGNOSES

- Tarlov cyst
- Dorsal spinal meningocele
- Dural dysplasia

PATHOLOGY

- Diverticulum of sacral subarachnoid space expands into sacral cyst with secondary remodeling of sacral canal
- Extradural arachnoid cyst; no meningeal herniation (not true meningocele)
- Cyst is connected to thecal sac by thin pedicle
- No neuronal elements within cyst

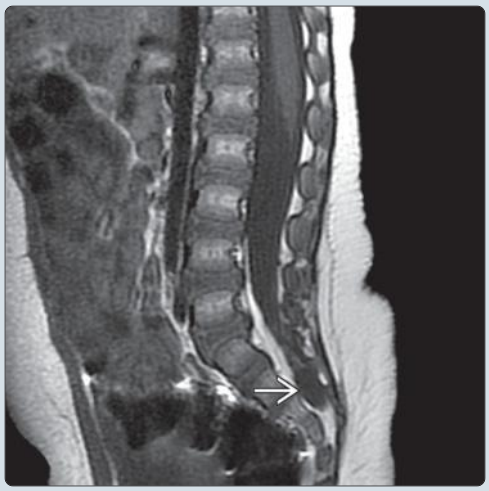
CLINICAL ISSUES

- Usually asymptomatic; incidental discovery on MR
  - Asymptomatic patients need no specific treatment
  - Indications for operation include ↑ cyst size on serial exams, onset of symptoms referable to cyst

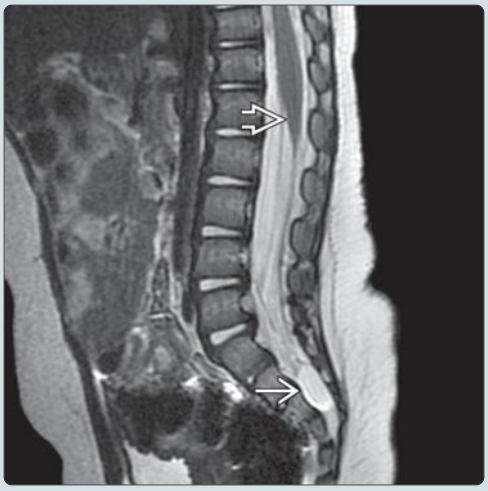
DIAGNOSTIC CHECKLIST

- AC centered in midline; cyst center over neural foramen implies Tarlov cyst





(Left) *Sagittal graphic of the lumbosacral spine demonstrates an extradural cyst ➡ within the sacral spinal canal below the thecal sac termination at S2. Note osseous remodeling of the sacral spinal canal margins.* **(Right)** *Sagittal T1WI MR demonstrates a small fluid signal extradural cyst in the sacral spinal canal ➡. The location is typical for occult sacral meningocele, now designated extradural arachnoid cyst. No solid tissue or fat is detected within the cyst.*





(Left) *Sagittal T2WI MR demonstrates a small extradural sacral arachnoid cyst ➡ at the caudal end of the spinal canal. The conus termination ➡ is normal. Note that the cyst signal intensity is slightly brighter than CSF within the dural sac, reflecting increased proteinaceous content.* **(Right)** *Axial T2WI MR through the sacrum at the level of the arachnoid cyst ➡ confirms mild expansile remodeling of the caudal osseous spinal canal by the cyst at the sacral level.*

TERMINOLOGY

Synonyms

- Sacral meningeal cyst, type IB meningeal cyst

Definitions

- Spinal extradural arachnoid cyst (AC) localized to sacrum

IMAGING

General Features

- Best diagnostic clue
  - Smooth, cystic enlargement of sacral spinal canal
- Location
  - Sacrum
- Size
  - Expands but does not transgress margins of sacrum
- Morphology
  - Sacral extradural meningeal cyst; no internal neural elements

Radiographic Findings

- Radiography
  - Sacral canal enlargement and posterior vertebral scalloping

CT Findings

- NECT
  - CSF-density cyst enlarges sacral canal
  - Nerve roots displaced by extradural cyst
- CECT
  - No cyst wall enhancement
- Bone CT
  - Smooth remodeling, enlargement of sacral canal
  - No expansion or remodeling of neural foramina

MR Findings

- T1WI
  - CSF signal intensity extradural sacral cyst adjacent to distal thecal sac
- T2WI
  - Cyst follows CSF signal intensity
  - No neural elements within cyst
- DWI
  - Hypointense signal intensity excludes epidermoid tumor
- T1WI C+
  - No cyst enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic intrasacral cyst without intrinsic neural elements

Nonvascular Interventions

- Myelography
  - Enlargement of distal sacral canal, extradural compression of distal contrast-filled thecal sac
    - ± contrast opacification of cyst (if isthmus large enough)

Other Modality Findings

- CSF cine phase contrast flow sensitive imaging may help define cyst walls by depicting differential CSF pulsation

Imaging Recommendations

- Best imaging tool
  - MR imaging best modality for initial diagnosis
  - CT myelography may help reveal connection between cyst and subarachnoid space
    - Delayed post myelographic imaging may show additional cyst filling
- Protocol advice
  - Sagittal and axial T1WI and T2WI to identify cyst, clarify relationship to adjacent structures

DIFFERENTIAL DIAGNOSIS

Tarlov Cyst

- Etiologically similar to sacral extradural arachnoid cyst; congenital dilatation of nerve root meningeal sleeves
- Large cysts may remodel sacrum, but will be eccentrically centered over neural foramen
- Frequently multiple

Dorsal Meningocele

- True meningocele; protrudes through dorsal dysraphism into subcutaneous tissues

Dural Dysplasia

- Vertebral scalloping usually present in lumbar spine as well as sacrum, ± lateral meningocele
- Search for characteristic imaging and clinical stigmata of etiological disorder

PATHOLOGY

General Features

- Etiology
  - Diverticulum of sacral subarachnoid space that expands into sacral cyst 2° to valve-like mechanism → secondary remodeling of sacral canal
- Associated abnormalities
  - Tarlov cyst(s)
  - Posterior spinal dysraphism
  - Tethered cord syndrome
- Extradural arachnoid cyst
  - No herniation of meninges, hence not true meningocele
  - No neuronal elements within cyst
- Cyst is connected to thecal sac by pedicle that permits contiguous CSF flow
- CSF pulsation ± raised intraspinal pressure (across stenotic pedicle) pressure erodes and remodels sacral canal

Staging, Grading, & Classification

- Nabors classification: Type IB meningeal cyst

Gross Pathologic & Surgical Features

- Sacral laminectomy → thinned sacral vertebral laminae
  - Cyst may be attached to distal thecal sac by narrow pedicle that permits 1-way (mostly) CSF flow into cyst
  - Symptomatic cysts less likely to communicate with subarachnoid space than asymptomatic cysts

Microscopic Features

- Cyst is lined by fibrous connective tissue ± single inner layer of arachnoid membrane

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic; incidental discovery on MR
  - Symptomatic: Chronic low back pain, sciatica, perineal paresthesias, and bladder dysfunction
  - Less common signs/symptoms
    - Intermittent, severe lower back pain
    - Atypical bowel symptoms, severe constipation, and stool incontinence
    - Tethered cord syndrome
- Clinical profile
  - Specific symptoms are referable to sacral root compression
  - May be exacerbated by positional change or Valsalva maneuver

Demographics

- Age
  - Teen → elderly
  - Rarely diagnosed in children
- Gender
  - Some series report M > F, others M < F
- Epidemiology
  - Spinal meningeal cysts are uncommon
    - Comprise 1-3% of all spinal tumors
  - Prevalence of occult sacral meningocele uncertain, but < prevalence of Tarlov cyst

Natural History & Prognosis

- Most patients are asymptomatic and need no specific treatment
  - Asymptomatic cysts are most commonly identified and referred to specialists following incidental discovery on MR
- Symptomatic patients or very large cysts may require surgery
  - Indications for operative intervention include increased cyst size on serial exams or onset of symptoms referable to cyst
  - Good prognosis for recovery following surgery

Treatment

- Conservative approach recommended for asymptomatic cysts, especially when small
- Symptomatic cysts may require treatment
  - Percutaneous cyst aspiration may relieve symptoms temporarily
    - May be used as diagnostic test prior to definitive therapy
    - Percutaneous cyst aspiration with fibrin glue therapy may produce definitive long-lasting symptom reduction
  - Operative therapy → sacral laminectomy to expose, resect cyst
    - May not be necessary to completely resect entire cyst
    - Primary goal is to close dural defect → eradicate 1-way valve communication, prevent cyst recurrence
    - If adhesions prevent full excision, may partially resect posterior wall or marsupialize cyst to subarachnoid space

DIAGNOSTIC CHECKLIST

Consider

- MR best modality for diagnosis of sacral cyst, preoperative planning
- Definitive cyst characterization based on operative inspection, histological examination

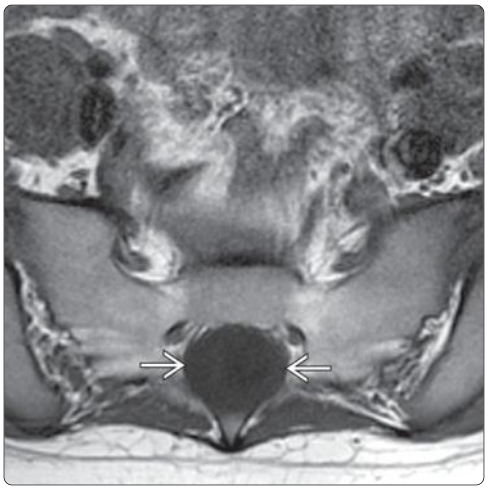
Image Interpretation Pearls

- Classic appearance is intrasacral cyst producing smooth sacral canal expansion with outward displacement of nerve roots
- AC centered in midline; center over neural foramen implies Tarlov cyst

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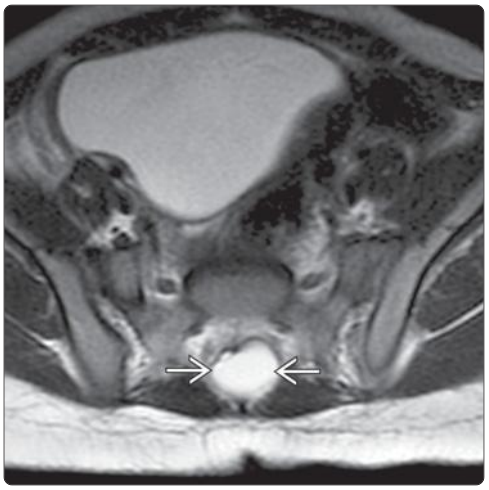
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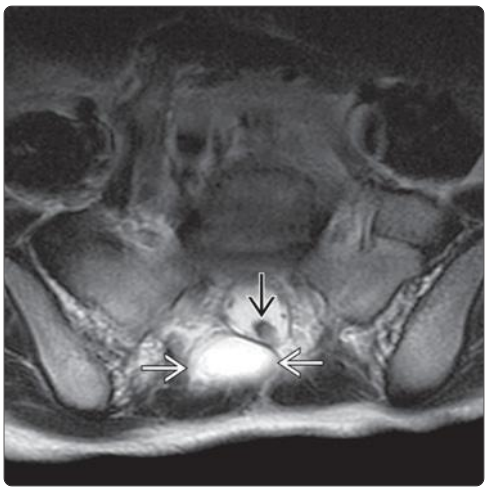
(Left) *Sagittal T1WI C+ MR demonstrates an extradural cyst ➡ located within the spinal canal at S2 and S3. It is extradural and distal to the thecal sac termination at S2. Note mild upward mass effect on the terminal thecal sac, which normally terminates at S2-S3.* **(Right)** *Axial T1WI MR confirms central arachnoid cyst ➡ location within the sacral spinal canal. There is remodeling and expansion of the adjacent vertebral body and overlying posterior elements.*





(Left) *Sagittal T1WI MR performed to evaluate clinically obvious but unrelated sacral dimple (marked with vitamin E capsule ➡) reveals an incidental small extradural arachnoid cyst ➡ within the spinal canal at the upper sacral level.* **(Right)** *Axial T2WI MR confirms extradural location of the arachnoid cyst ➡ with smooth expansion and remodeling of the central spinal canal and displacement of adjacent epidural fat.*





(Left) *Sagittal T1WI MR in a Klippel-Feil patient with diastematomyelia ➡ demonstrates a low-lying, hydromyelic tethered spinal cord in addition to a fluid signal cyst ➡ in the terminal sacral canal. The cyst results in remodeling and enlargement of the sacral central canal.* **(Right)** *Axial T2WI MR reveals a low-lying, tethered spinal cord ➡ within the thecal sac, which is anterior and superior to the sacral extradural arachnoid cyst ➡ located eccentrically within the canal.*

Sacrococcygeal Teratoma

KEY FACTS

TERMINOLOGY

- Congenital sacral tumor containing elements of all 3 germ layers

IMAGING

- Large, heterogeneous sacral mass in infant
  - Variably contains calcifications, mixed solid and cystic components, fat-debris levels, bone, hair, teeth, or cartilage
- Usually large at diagnosis

TOP DIFFERENTIAL DIAGNOSES

- Anterior sacral meningocele (ASM)
- Chordoma
- Dermoid tumor
- Exophytic rhabdomyosarcoma

PATHOLOGY

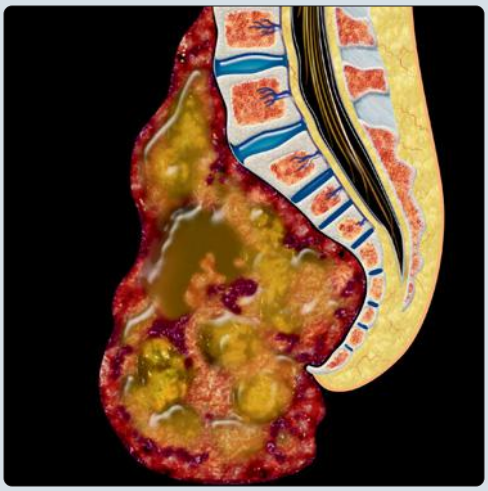
- Tumor originates from totipotential cell rests at caudal spine/notochord (Hensen node)
- Altman/AAP classification: 4 surgical subtypes based on relative proportion of internal and external components

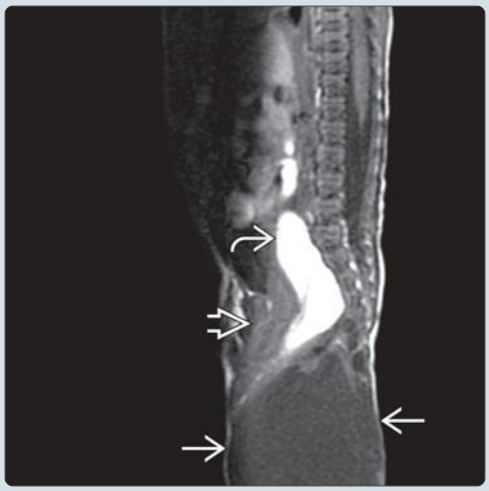
CLINICAL ISSUES

- Back/pelvic mass in newborn
  - Exophytic masses (AAP types I, II) easily diagnosed, but internal (types III, IV) occult → delayed diagnosis
- In utero presentation
  - Polyhydramnios, high-output cardiac failure with hydrops, hepatomegaly, placentomegaly

DIAGNOSTIC CHECKLIST

- Heterogeneous sacral tumor ± calcification, cysts, hemorrhage in infant strongly suggests diagnosis
- AAP type influences prognosis and treatment approach





(Left) *Sagittal graphic (AAP II) depicts a typical large, heterogeneous, partially cystic sacrococcygeal teratoma located anterior to the sacrum, with both internal and external components.* **(Right)** *Sagittal T1WI MR (AAP type I) demonstrates a predominately cystic sacrococcygeal teratoma contiguous with the coccyx tip. The tumor is nearly entirely external. The urinary bladder is not displaced. Hyperintense rectal contents probably reflect meconium.*





(Left) *Sagittal T2WI MR (AAP type II) reveals a large, heterogeneous exophytic sacral mass with relatively equal internal and external distribution. The mass displaces the urinary bladder anteriorly and superiorly.* **(Right)** *Sagittal STIR MR (AAP type III, older female infant) reveals a heterogeneous, predominantly internal pelvic mass. The tumor anteriorly and upwardly displaces the rectum and urinary bladder. No sacral osseous destruction is apparent.*

TERMINOLOGY

Synonyms

- Sacral germ cell tumor (SGT), germ cell tumor of coccyx

Definitions

- Congenital sacral tumor containing elements of all 3 germ layers

IMAGING

General Features

- Best diagnostic clue
  - Large, heterogeneous sacral mass in infant
- Location
  - Sacrum/coccyx
- Size
  - Usually large at diagnosis
    - Mature teratomas ~ 7.5 cm
    - Immature teratomas ~ 11.6 cm
- Morphology
  - Heterogeneous mass variably contains calcifications, mixed solid and cystic components, fat-debris levels, bone, hair, teeth, or cartilage

Radiographic Findings

- Radiography
  - Soft tissue mass, calcifications (≤ 60%)

CT Findings

- CECT
  - Complex mixed attenuation cystic/solid pelvic mass with enhancement of solid portions
    - Calcifications (≤ 60%); small punctate foci → well-formed teeth, bones
    - Calcium (hyperdense), fluid (hypodense), and fat (markedly hypodense)

MR Findings

- T1WI
  - Heterogeneous mixed signal intensity; fat (hyperintense), soft tissue (isointense), cyst fluid (hypointense), and calcium (markedly hypointense)
- T2WI
  - Heterogeneous signal intensity; fat (hyperintense), soft tissue (isointense), cyst fluid (hyperintense), and calcium (markedly hypointense)
- STIR
  - Depicts lobulated, sharply demarcated tumor margins against dark marrow background
- T2* GRE
  - Shows calcifications best; hemorrhage presents similar imaging appearance
- T1WI C+
  - Heterogeneous enhancement of solid portions

Ultrasonographic Findings

- Grayscale ultrasound
  - Complex mixed hypo-/hyperechoic sacral mass
    - Obstetrical: In utero diagnosis → cesarean section, possible fetal surgery or radiofrequency ablation (RFA)
    - Postnatal: Visualization hampered by fat/calcium shadowing, posterior bone elements, large size

Imaging Recommendations

- Best imaging tool
  - Prenatal: Obstetrical ultrasound ± fetal MR
  - Postnatal: MR
- Protocol advice
  - MR ± CT to determine full extent of mass, treatment planning
    - Sagittal and axial MR imaging for surgical planning
    - CT with oral/IV contrast to delineate extent of bone destruction, calcification

DIFFERENTIAL DIAGNOSIS

Anterior Sacral Meningocele (ASM)

- Cystic mass; no solid component or enhancement
- Enlarges with Valsalva maneuver

Chordoma

- Mixed solid/cystic destructive sacral mass
- Rare in children; peak incidence: 5th-6th decade
- T2WI → markedly hyperintense
- Bone CT → destructive margins

Dermoid Tumor

- Isointense/hyperintense (T1WI) → hyperintense (T2WI)
- Smaller, more homogeneous than SGT

Exophytic Rhabdomyosarcoma

- Aggressive-appearing tumor without calcium, bone, or hair

PATHOLOGY

General Features

- Etiology
  - Originates from totipotential cell rests at caudal spine/notochord (Hensen node)
    - Degree of totipotential cell differentiation → tumor type
    - Undifferentiated cells → more malignant tumors
  - Alternative hypothesis: "Twinning" accident → incomplete separation during embryogenesis
- Genetics
  - Most cases are sporadic; some syndromic associations (Currarino triad)
  - Scattered reports of familial parent-child associations (autosomal dominant)
    - Congenital sacral deformities, anorectal stenosis, vesicoureteral reflux, retrorectal abscess, and skin dimple
- Associated abnormalities
  - Currarino triad, anorectal or genitourinary abnormalities (10%)
- Most common newborn tumor
  - 80% mature, 20% immature; immature teratomas have greater risk of malignancy
- Malignancy risk 10% at term; ↑ to 65-90% if diagnosed > 2 months
  - Malignant risk ↑ with age at diagnosis, higher surgical subtype, male gender, presence of necrosis or hemorrhage

Staging, Grading, & Classification

- 4 surgical subtypes (Altman/AAP classification)
  - Type I: Primarily external (47%) → best prognosis
  - Type II: Dumbbell shape, equal external/internal portions (34%)
  - Type III: Primarily internal within abdomen/pelvis (9%)
  - Type IV: Entirely internal; no visible external component (10%) → worst prognosis

Gross Pathologic & Surgical Features

- Benign teratomas (83%)
  - Heterogeneous, noninvasive, partially cystic, calcified soft tissue mass; rarely invade spinal canal
  - Multiple tissue types show varying stages of differentiation, maturation
- Malignant teratomas (17%)
  - More likely intrasacral, aggressive local behavior
  - Malignancy postulated to arise from differentiated cell lines within teratoma → neuroblastoma, adenocarcinoma, rhabdomyosarcoma, embryonal cell carcinoma, or anaplastic carcinoma

Microscopic Features

- Elements derived from all 3 germ layers (endoderm, mesoderm, and ectoderm)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back/pelvic mass in newborn
    - Exophytic masses (AAP types I, II) easily diagnosed, but internal (types III, IV) occult → delayed diagnosis
  - In utero presentation
    - Polyhydramnios, high-output cardiac failure with hydrops, hepatomegaly, placentomegaly
- Other signs/symptoms
  - Urinary retention, constipation (larger tumors)
- Clinical profile
  - Most common: Neonate with large benign sacral tumor noted prenatally or at delivery
  - Less common: Infant with buttock asymmetry or delayed presentation at > 1 month with presacral tumor
- Laboratory: Serum α-fetoprotein (AFP), β-human chorionic gonadotropin (HCG) levels postoperative tumor markers

Demographics

- Age
  - 50-70% diagnosed in utero or during 1st days of life, 80% by 6 months, and < 10% beyond 2 years
- Gender
  - M < F (1:4)
- Epidemiology
  - 1:35,000-40,000 births

Natural History & Prognosis

- Prognosis excellent in benign tumors
  - Mature or mostly cystic → good prognosis
  - Immature teratomas → greater risk of malignancy if not totally resected
- Malignant tumors have variable prognosis; later diagnosis (> 1st birthday), internal location → worse prognosis
- Fetal mortality rate 20-65% (2° to polyhydramnios, tumor hemorrhage, high-output cardiac failure)
  - Hydrops fetalis portends grim prognosis; > 30 weeks gestation → 25% mortality, < 30 weeks gestation → 93% mortality
- Postpartum morbidity attributable to associated congenital anomalies, tumor mass effects, recurrence, and intraoperative/postoperative complications

Treatment

- Surgery alone curative if entire benign tumor and coccyx removed; recurs if coccyx not resected
- Cytoreduction surgery + radiation and chemotherapy may be palliative for malignant tumors
  - Early diagnosis, resection best chance for cure
- Fetal surgery, (RFA) offered at some specialized centers; good outcome requires careful patient selection

DIAGNOSTIC CHECKLIST

Consider

- Imaging goals: Imperative to confirm diagnosis, determine intra-/extrapelvic extent and size, relationship to adjacent structures, and presence/absence of metastatic disease
- AAP type influences prognosis, treatment approach

Image Interpretation Pearls

- Heterogeneous sacral tumor ± calcification, cysts, hemorrhage suggests diagnosis

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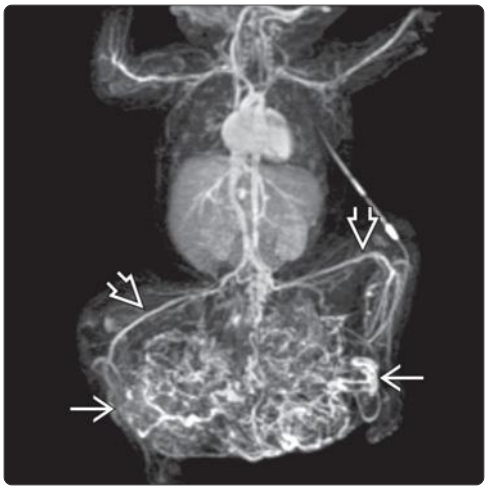
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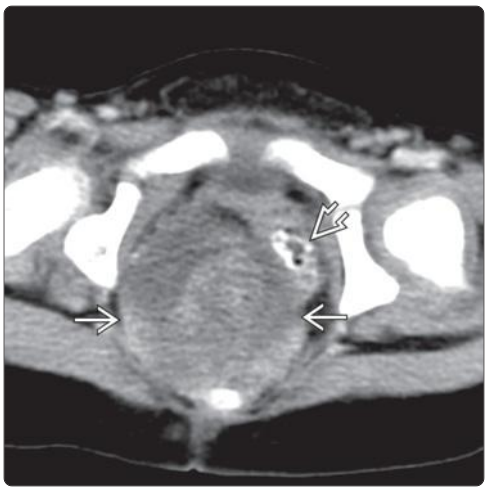
(Left) *Anteroposterior radiograph shows a large calcified pelvic mass (note calcifications ⇨) originating from the perineum. No osseous sacral anomalies are visible. Surgical resection confirmed sacrococcygeal teratoma (AAP type I).* **(Right)** *Coronal T1WI MR (AAP type I) demonstrates a cystic exophytic pelvic mass ➡ with relatively small connection to the perineum and no significant internal component. There is a small solid tumor nodule ➡ along the superior tumor margin.*





(Left) *Sagittal T1WI C+ FS MR (AAP type II) reveals a large, heterogeneous, exophytic sacral mass ➡ with relatively equal internal and external distribution. There is avid enhancement of the solid tumor elements.* **(Right)** *Coronal contrast-enhanced TRICKS MRA (AAP type II) demonstrates a large vascular sacral tumor ➡ with arterial vascular supply from large internal iliac artery branches ➡ feeding numerous tortuous abnormal tumor vessels.*





(Left) *Sagittal CECT (AAP type III, older female infant) depicts a heterogeneous, mildly enhancing predominately intrapelvic mass ➡ located along the ventral sacrum and coccyx. The presacral fat plane is obliterated, but no osseous invasion is apparent.* **(Right)** *Axial CECT (AAP type III, older female infant) shows a heterogeneous, mildly enhancing pelvic mass ➡ producing anterior and leftward rectal displacement ➡. There is not a well-developed fat plane between the rectum and mass.*

Terminology

Abbreviations: Craniovertebral junction (CVJ).

Anatomy-Based Imaging Issues

Development of the skull base, atlas (C1), and axis (C2) is embryologically distinct compared to the subaxial cervical spine and therefore merits separate discussion. Composed of the occiput, atlas (C1), and axis (C2), with the adjacent muscles, soft tissues, and ligaments, the craniovertebral junction serves as the articulation between the skull base and the spine.

Imaging Protocols

Multiplanar magnetic resonance (MR) imaging provides superior contrast resolution to other imaging modalities and is ideal for evaluation of soft tissues, neural structures, and ligaments. Conversely, multidetector computed tomography (CT) with orthogonal reconstructions optimally defines CVJ osseous anatomy and pathology.

Embryology

Occipital Bone and Posterior Skull Base

Occipital bone and skull base development is complex and derives from four occipital sclerotomes. The 1st and 2nd occipital sclerotomes form the basiocciput, which comprises the inferior half of the clivus. The sphenooccipital synchondrosis separates the basiocciput from the basisphenoid portion of the sphenoid bone. The 3rd occipital sclerotome forms the exoccipital bone and jugular tubercles. The 4th occipital sclerotome contributes to the anterior clival tubercle, while the anterior margin of the foramen magnum and occipital condyles are formed by the ventral segment of the proatlas neural arch.

Embryologically, the occipital bone has both enchondral and membranous origins. Hypoplasia or aplasia of the occipital and 1st cervical somite results in congenital skull base and atlas anomalies (atlantooccipital assimilation).

Atlas (C1)

The 4th occipital somite combines with the 1st cervical somite to form the anterior C1 arch (C1 centrum) and contiguous supporting ligaments. The atlas forms from three primary ossification centers (one centrum, two neural arches), separated by cartilaginous neurocentral synchondroses. Development of the posterior ossification centers normally occurs before the anterior ossification center.

Axis (C2)

The axis is derived from both the 1st and 2nd cervical somites. The 1st cervical sclerotome forms the odontoid process, while the axis body develops from the 2nd cervical sclerotome. The C2 facets and posterior arch also derive from the 2nd cervical sclerotome. The axis develops from five primary and two secondary ossification centers. The odontoid is separated from the axis body by the neurocentral synchondrosis. The terminal dens ossicle is not ossified at birth but usually is visible by age 3 years and fuses with odontoid process by age 12 years. However, an odontoid cleft at the tip may normally persist into adolescence. The 1st cervical somite forms the anterior C1 arch (C1 centrum), and failure of the C1 centrum to separate into the anterior C1 arch and the odontoid process results in either odontoid hypoplasia/aplasia or fusion of C1 and the dens.

Pathology-Based Imaging Issues

Occipital Bone & Skull Base Anomalies

Occipital sclerotome segmentation failure may result in **basilar invagination**, with severity best assessed using CVJ craniometry. Platybasia (flattening of skull-base angle to the upper cervical spine) is variably present in patients with basilar invagination.

Basiocciput hypoplasia is characterized by clival foreshortening ("short clivus") and abnormal craniovertebral relations; it is commonly identified in conjunction with occipital condyle hypoplasia. Some patients additionally demonstrate concurrent basilar invagination, with severity variable depending upon the number of affected occipital sclerotomes.

Occipital condyle hypoplasia occurs secondary to anomalous hypoplastic development of the 3rd occipital sclerotome. In this malformation, partial hypoplasia of the proatlas is frequently associated with hypoplastic exoccipital bone and jugular tubercles resulting in decreased skull base height and basilar invagination. Condylar hypoplasia may be either unilateral or bilateral and symmetric or asymmetric.

Segmentation failure of the 4th occipital sclerotome and 1st cervical sclerotome results in **atlantooccipital nonsegmentation** or assimilation ("occipitalization of the atlas"). Assimilation may be limited to the anterior arch, posterior arch, lateral masses, or a combination. Nonsegmentation between the anterior C1 arch and the anterior foramen magnum lip produces a characteristic comma-shaped configuration of the clivus tip and decreased foramen magnum AP diameter. A rarer variant of atlantooccipital nonsegmentation demonstrates occipitalization of the anterior C1 arch and nonsegmentation of the posterior C1 arch from the C2 vertebral posterior elements ("block atlas"). Additionally, up to 70% of atlantooccipital nonsegmentation patients may also have partial or complete C2/C3 nonsegmentation.

Atlas (C1) Anomalies

Premature fusion of the cartilaginous C1 neurocentral synchondrosis produces an abnormally small C1 ring diameter, known as **hypoplastic complete posterior arch**. The resultant osseous spinal canal narrowing may manifest clinically significant spinal stenosis requiring surgical decompression.

The majority of atlas anomalies reflect **incomplete posterior arch formation**. Incomplete fusion of the posterior C1 arch is relatively common (4%) and usually asymptomatic. The majority (97%) of osseous defects are midline, with lateral clefts accounting for the remaining 3%.

Incomplete anterior arch fusion is comparatively rare. The anterior C1 arch usually forms from a single ossification center, producing an anterior tubercle with paramedian synchondroses between the two neural arches. If the anterior arch instead forms from paired paramedian ossification centers with subsequent chondrogenesis failure, anterior arch fusion failure results. If the posterior arch is also incomplete, a "split atlas" configuration results. Isolated incomplete C1 arch is often asymptomatic, while association with meningeal or spinal cord anomalies or occurrence in conjunction with posterior arch fusion anomalies may be clinically significant. In these cases, it is important to assess for dynamic instability.

Axis (C2) Anomalies

Persistent ossiculum terminale, a variant of normal anatomy that is stable and of little clinical significance, results from failure of the proatlas to fuse to the rest of the odontoid process. The terminal ossicle is well corticated and the remainder of the odontoid is normal size, helping to distinguish from os odontoideum.

The "os" of **os odontoideum** is a round, well-corticated ossicle positioned between the basion and C2 body in place of a normal dens. The axis body shows a smooth, rounded upper margin representing the residual hypoplastic odontoid process, and the anterior C1 arch is usually enlarged. The exact etiology of os odontoideum is debated, with both developmental and post-traumatic etiologies championed. Because the gap between the os and C2 body extends above the level of the superior C2 articular facets, cruciate ligament incompetence and atlantoaxial instability are common. Assessment for associated congenital CVJ anomalies is important because there is an increased incidence of os odontoideum in association with many congenital conditions including Down syndrome, Morquio syndrome, spondyloepiphyseal dysplasia, and Klippel–Feil spectrum.

Odontoid hypoplasia is an uncommon anomaly that may be isolated, although it is often described as part of skeletal dysplasia syndromes (spondyloepiphyseal dysplasia, mucopolysaccharidoses, metatropic dwarfism). Complete odontoid aplasia is very rare and features an "excavated" appearance of upper C2 body. The attachments for the apical and alar ligaments are abnormal in more severe cases, predisposing to atlantoaxial instability.

Clinical Implications

Klippel-Feil Syndrome (KFS)

The classic clinical triad described by Klippel and Feil in 1912 includes short neck, low hairline, and limited neck mobility, although only a minority of patients feature all three findings. Three KFS types are described: Type I KFS is the classic variety featuring extensive cervical and thoracic segmentation anomalies; type II KFS is most common and demonstrates segmentation anomalies of one or two pairs of cervical vertebrae (usually C2/C3 and C5/C6); and type III KFS consists of either type I or II findings in conjunction with lower thoracic or lumbar segmentation anomalies. CVJ anomalies including basilar invagination, odontoid hypoplasia, and atlantooccipital assimilation are common. Additionally, hypermobility from accelerated cervical spondylosis at levels adjacent to nonsegmented vertebra predisposes KFS patients to cervical spinal cord or vertebral artery injuries following relative minor trauma.

Down Syndrome (Trisomy 21)

Atlantooccipital instability (AOI), atlantoaxial instability (AAI), or a combination of both are commonly observed in trisomy 21 patients. The prevalence of AAI in children with Down syndrome is estimated between 9-30%, while the prevalence of AOI may be as high as 61%. Other CVJ anomalies are also more common in children with trisomy 21 than the general population.

Achondroplasia

Achondroplasia is the most common congenital short-limbed dwarf syndrome and is characterized by abnormal enchondral bone formation. Consequently, CVJ abnormalities occur within regions where bone growth occurs primarily through enchondral ossification. Therefore, the skull, facial bones, skull base, and foramen magnum are underdeveloped while the calvarium (membranous origin) develops normally. Foramen magnum stenosis is common in achondroplasia patients and often clinically significant. Other associated CVJ anomalies include basiocciput hypoplasia, platybasia, and odontoid hypoplasia.

Mucopolysaccharidoses (MPS)

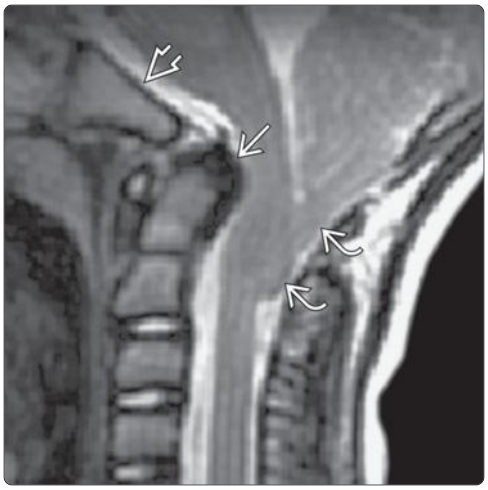
Mucopolysaccharidoses (MPS) are inherited lysosomal storage disorders caused by enzyme deficiency for degradation of specific glycosaminoglycans. CVJ anomalies are a major cause of morbidity and mortality, especially in the Morquio (type IV) and Hurler (type I) variants, and include odontoid hypoplasia, ligamentous instability, soft tissue pannus at the dens, and spinal cord compression by the posterior C1 arch.

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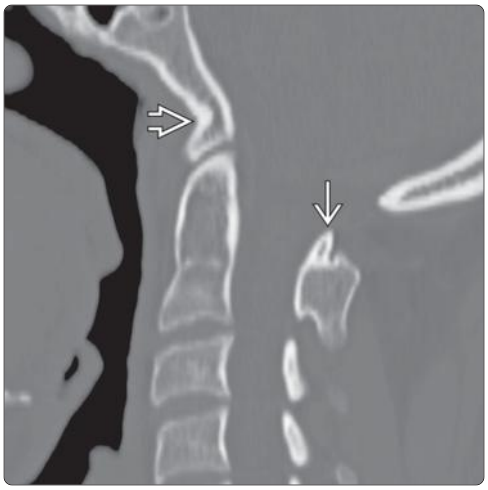
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(Left) *Axial bone CT demonstrates an anomalous clival cleft* ➡ *in conjunction with aberrant flattened coronal clivus orientation.* **(Right)** *Sagittal T2WI MR reveals anomalous flattening of the clivus* ➡ *relative to the cervical spine (platybasia) in conjunction with pronounced odontoid process retroflexion* ➡*. An important additional finding demonstrated is previously unsuspected Chiari 1 malformation* ➡ *with cerebellar tonsillar ectopia.*



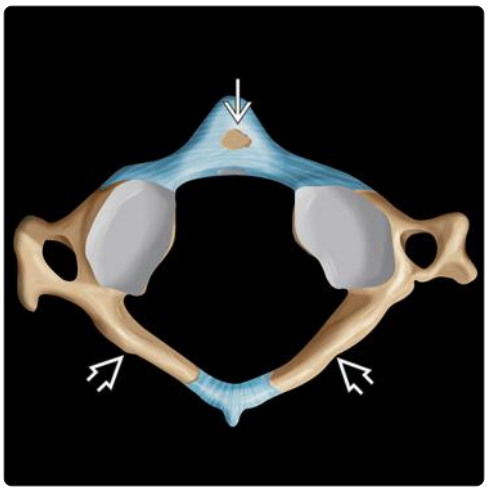


(Left) *Sagittal bone CT reformat shows assimilation of the anterior C1 ring and clivus* ➡*, producing a "comma-shaped" clivus. Note also fusion of the posterior C1 ring* ➡ *with the C2 spinous process ("block atlas") resulting in mild canal stenosis.* **(Right)** *Coronal bone CT of a patient with atlantooccipital instability shows congenitally abnormal flat contour of both occipital condyles* ➡*, an anatomic variant predisposing to abnormal CVJ mechanics and atlantooccipital subluxation.*

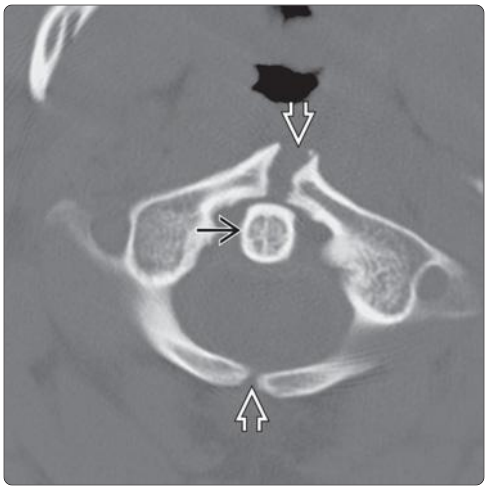




(Left) *Axial graphic of the atlas shows normal embryological development of the paired neural arch primary ossification centers* ➡ *and single anterior C1 (centrum) ossification center* ➡*. Nonossified cartilage (blue) joins the ossification centers.* **(Right)** *Axial bone CT in a 2-week-old infant demonstrates the 3 atlas 1° ossification centers. Much of the atlas is unossified cartilage at this age. The paired odontoid process ossification centers* ➡ *are posterior to the C1 anterior arch* ➡*.*

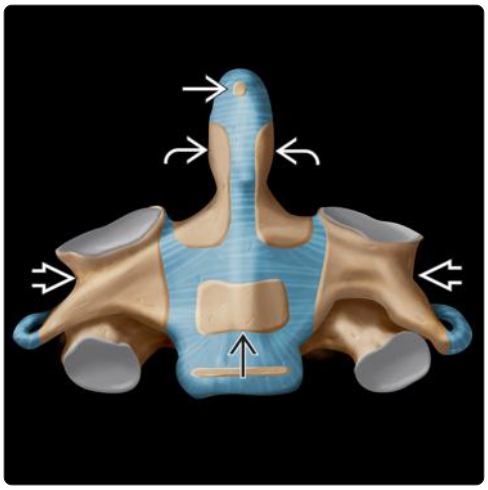


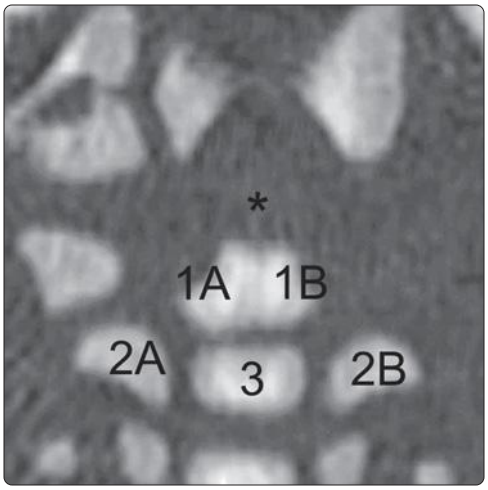




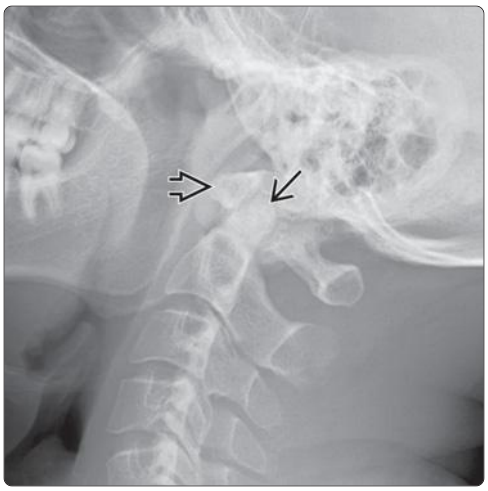


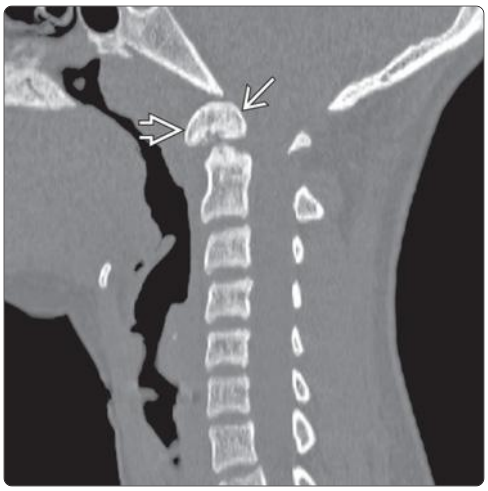
(Left) *Axial bone CT demonstrates incomplete fusion of both anterior and posterior C1 rings with prominent midline clefts. Normal alignment with the odontoid process is maintained.* **(Right)** *Axial bone CT confirms presence of both C1 lateral masses and complete anterior C1 ring. Posterior ring formation is anomalous, with the right posterior C1 ring mostly present, but the left C1 posterior ring shows only a small ossified remnant.*





(Left) *Coronal graphic of the skeletally immature axis shows normal appearance of the 5 primary C2 ossification centers (centrum, lateral masses and neural arches, and odontoid process). The secondary terminal ossicle has also commenced ossification.* **(Right)** *Coronal bone CT in a 2-week-old infant shows 5 C2 vertebra 1° ossification centers (1-3). Odontoid and lateral mass centers are paired. Dens tip (*) is cartilaginous at this developmental stage.*





(Left) *Lateral radiograph shows a congenital hypoplastic odontoid process with normal C1/C2 alignment. The anterior C1 ring is mildly enlarged in compensation. Dynamic flex-extend radiographs (not shown) confirmed no dynamic instability.* **(Right)** *Sagittal bone CT of the cervical spine shows congenital fusion of anterior C1 ring and os odontoideum. Irregular margination of the inferior os and the odontoid base may indicate sequelae of unrecognized remote trauma.*

KEY FACTS

TERMINOLOGY

- Paramastoid process, paroccipital process, jugular process, parajugular process
- Enlarged bony process of cranial base projecting caudally to transverse process of atlas (C1)
- Uncommon variant (approximately 0.5%)

IMAGING

- Many variants ranging from small hump of bone (paracondylar tuberculum) to a free-ended process or jointed with transverse process of C1
  - If not attached to occipital bone, process is isolated rod-like bony element called massa paracondylica
  - Portions may be pneumatized from mastoid air cells

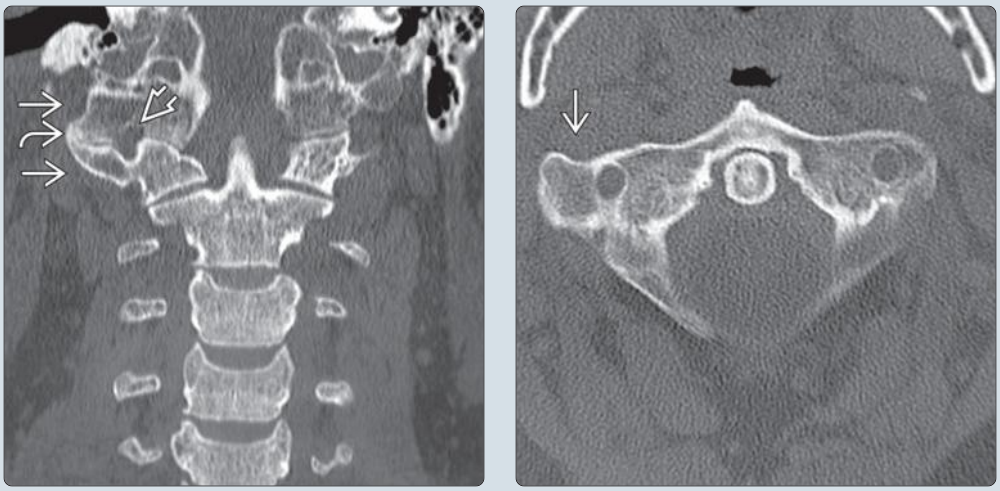
TOP DIFFERENTIAL DIAGNOSES

- Assimilation of C1 into occiput
  - Bony fusion across occipital condyles and lateral masses of C1
- Klippel-Feil syndrome
  - Variable fusions of multiple cervical vertebral bodies and posterior elements
- Proatlas segmentation abnormality
  - Shelf-like bony projection arising from ventral foramen magnum or occipital condyle

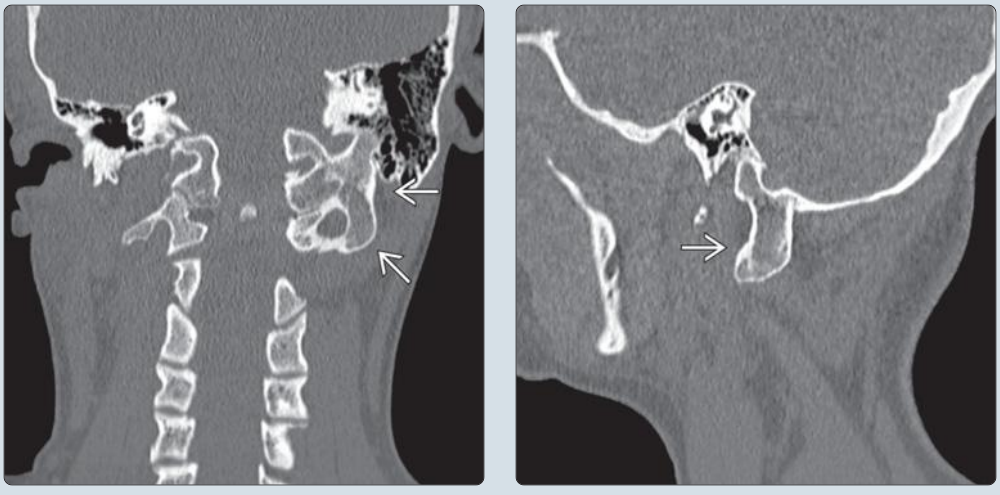
CLINICAL ISSUES

- Typically asymptomatic
- May cause limitation of neck movement and restricted range of motion
- Rare cases of chronic headache pain relieved by surgical resection of paracondylar process
- Rare reports of associated vertebral stenosis

(Left) *Coronal NECT shows a large bony mass projecting off the right occipital condyle and merging with an enlarged right transverse process of C1 with a neoarticulation. The vertebral artery has a well-defined canal through the bony mass.* **(Right)** *Axial NECT shows the junction of the right paracondylar process with the markedly enlarged right transverse process of C1. The paracondylar process shows considerable variation, ranging from a small tubercle to a large bony process.*



(Left) *Coronal NECT shows a large, well-defined and corticated bony fusion that has occurred on a congenital basis connecting the mastoid region to the transverse process of C1. This is usually asymptomatic, although there are rare reports of associated vertebral artery stenosis.* **(Right)** *Sagittal NECT shows the paracondylar process as a large, cone-shaped, corticated bony mass extending inferiorly from the occipital condylar region.*



TERMINOLOGY

Synonyms

- Paramastoid process, paroccipital process, jugular process, parajugular process

Definitions

- Enlarged bony process of cranial base projecting caudally to transverse process of atlas (C1)
- Epitransverse process is mirror image process and arises from transverse process of atlas and projects cephalad to occipital condyle
- Uncommon variant (approximately 0.5%)

IMAGING

General Features

- Best diagnostic clue
  - Broad osseous corticated process projecting down from lateral aspect of occipital condyle to transverse process of C1
  - At location of insertion of lateral rectus capitis muscle, which may be small or absent
- Location
  - Lateral skull base to transverse process of C1
- Size
  - Variable from small tubercle to large bony process with synovial joint
- Morphology
  - Corticated, normal bone
    - ± joint or neoarticulation
  - Many variants ranging from small hump of bone (paracondylar tuberculum) to a free-ended process or jointed with transverse process of C1
    - If not attached to occipital bone, process is isolated rod-like bony element called massa paracondylica
    - Portions may be pneumatized from mastoid air cells

CT Findings

- NECT
  - Corticated, cone-shaped bony projection extending to transverse process of C1, ± true or pseudojoint formation

DIFFERENTIAL DIAGNOSIS

Assimilation of C1 Into Occiput

- Bony fusion across occipital condyles and lateral masses of C1
- Up to 3% of general population
- Ranges from complete incorporation of atlas into occiput to discrete osseous bridges between the 2
- Lateral process assimilation seen in 17% of C0-C1 assimilations

Klippel-Feil Syndrome

- Variable fusions of multiple cervical vertebral bodies and posterior elements

Proatlas Segmentation Abnormality

- Shelf-like bony projection arising from ventral foramen magnum or occipital condyle
- May cause cord compression, cranial nerve palsies

PATHOLOGY

General Features

- Etiology
  - Forms from maldevelopment of 1st cervical sclerotome around 4th week in utero
    - Occipital bone derived from basioccipital, exoccipital, and supraoccipital portions surrounding foramen magnum
    - Basioccput develops 4 occipital somites
    - Caudal portion of 4th occipital somite fuses with cranial portion of 1st cervical somite to form proatlas
    - Proatlas is assimilated into occiput to form articular condyles and tip of odontoid
    - Caudal 1/2 of 1st cervical somite fuses with cranial portion of 2nd cervical somite to form atlas and odontoid of axis
      - Paracondylar process represents vestiges of cranial half of 1st cervical somite

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Typically asymptomatic
  - May cause limitation of neck movement and restricted range of motion
  - Rare cases of chronic headache pain relieved by surgical resection of paracondylar process
  - Rare reports of associated vertebral stenosis
- Other signs/symptoms
  - May be associated with other abnormalities
    - Assimilation of anterior arch of C1 with anterior foramen magnum
    - C2-C3 fusion
    - Bipartite hypoglossal canal
  - May predispose to accelerated degenerative change of C1-C2 level

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KEY FACTS

TERMINOLOGY

- Synonym: Split C1

IMAGING

- Anterior, posterior C1 arch defects → 2 C1 hemirings
  - Defects usually midline but may be paramedian
  - Wider gaps suggest independent lateral migration of C1 hemirings, instability
- ± hypoplastic ring → spinal canal stenosis, cord compression, myelomalacia, syrinx

TOP DIFFERENTIAL DIAGNOSES

- Jefferson C1 fracture
- Isolated anterior C1 arch defect
- Isolated posterior C1 arch defect

PATHOLOGY

- Simultaneous hypoplasia of C1 hypochordal bow, lateral C1 sclerotomes → combined anterior, posterior C1 arch defects
  - Ring defects usually contain fibrous connective tissue, not cartilage
  - Connective tissue more fragile than cartilage → hemirings may split
- Bifid anterior and posterior C1 rings without hypoplasia or significant gap usually stable

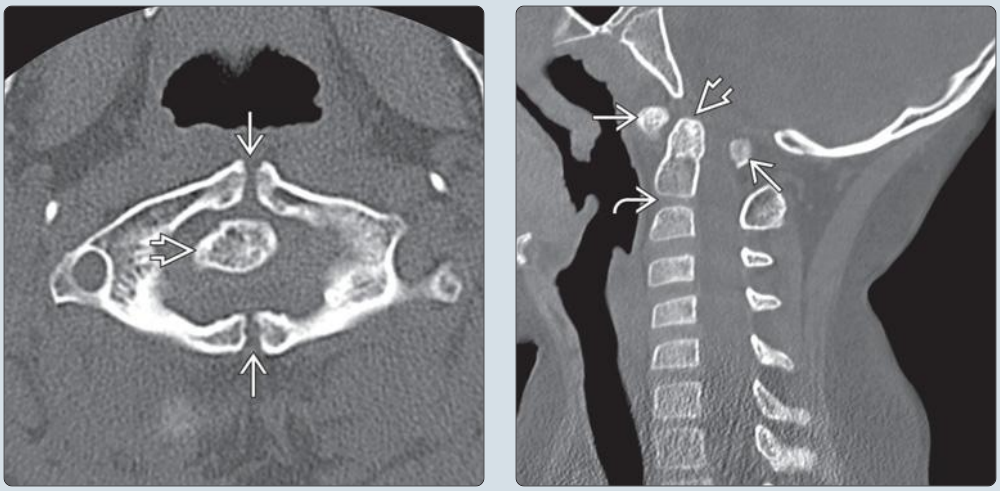
CLINICAL ISSUES

- Prognosis largely depends on severity of ring defects, CVJ stability, C1 ring hypoplasia

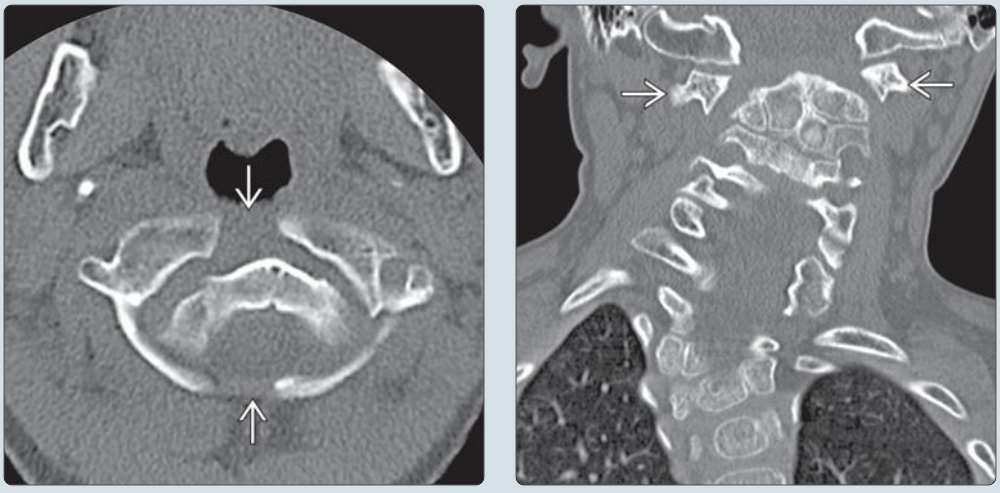
DIAGNOSTIC CHECKLIST

- Split atlas is rare congenital anomaly
  - Stability depends on anomaly severity, transverse ligament integrity
- Search for associated skull base, CVJ anomalies

(Left) *Axial bone CT demonstrates short AP dimension of the hypoplastic C1 ring as well as a "split ring" configuration characterized by midline defects ➡ in both anterior and posterior rings. The dens ➡ is thickened and dysplastic.* **(Right)** *Sagittal bone CT in patient with a split C1 ring shows mildly dysplastic dens ➡ formation and abnormal hypoplastic C1 ring ➡, resulting in severe spinal canal stenosis. Note the associated C2/3 segmentation failure ➡.*



(Left) *Axial bone CT reveals large midline osseous defects ➡ in both the anterior and posterior C1 rings. The wide gaps suggest instability.* **(Right)** *Coronal bone CT shows congenital scoliosis and extensive cervicothoracic vertebral segmentation failure. The C1 lateral masses ➡ are independently displaced laterally in opposite directions, permitted by the split atlas configuration. This is a very unstable configuration necessitating surgical fusion.*



TERMINOLOGY

Synonyms

- Split C1

Definitions

- Anterior, posterior C1 arch defects → 2 C1 hemirings

IMAGING

General Features

- Best diagnostic clue
  - Midline gaps in anterior and posterior C1 arches
- Location
  - Craniovertebral junction
- Size
  - Cleft gap ranges from narrow to wide
- Morphology
  - Anterior and posterior C1 arch defects
    - Defects usually midline but can be paramedian

CT Findings

- Bone CT
  - Left and right C1 hemirings
    - ± hypoplastic ring → spinal canal stenosis, cord compression
    - Wider gaps suggest independent lateral migration of C1 hemirings, instability

MR Findings

- T1WI
  - Soft tissue in gaps usually connective tissue, not cartilage
- T2WI
  - Transverse atlantal ligament (TAL) best seen on thin axial T2WI
  - ± cord compression, myelomalacia, syrinx

DIFFERENTIAL DIAGNOSIS

Jefferson C1 Fracture

- Noncorticated osseous fracture margins
- Trauma history, detection of soft tissue injury helpful

Isolated Anterior C1 Arch Defect

- Variable size of C1 ring cleft
- May be associated with clivus anomalies

Isolated Posterior C1 Arch Defect

- Posterior C1 arch rachischisis
- 10x more common than anterior arch defects

PATHOLOGY

General Features

- Etiology
  - Atlas normally forms from 3 primary ossification centers
    - Anterior ossification center → anterior tubercle
    - 2 lateral centers → lateral masses, posterior arch, posterior tubercle (~ 4th year)
    - 4th ossification center (variant, 2% of population) forms posterior tubercle between 2 neural arches
  - Simultaneous hypoplasia of C1 hypochordal bow, lateral C1 sclerotomes → combined anterior, posterior C1 arch defects
- Associated abnormalities
  - Skull base anomalies
  - Klippel-Feil spectrum

Gross Pathologic & Surgical Features

- Anterior and posterior ring defects usually contain fibrous connective tissue, not cartilage
  - Connective tissue more fragile than cartilage → hemirings may split
- Bifid anterior and posterior C1 rings without hypoplasia or significant gap usually stable
  - Anterior arch defect small, insertion tubercles and TAL usually intact
  - Arch gaps usually remain open into adulthood
- If anterior arch defect large or unilateral C1 lateral mass aplastic, insertion tubercle/TAL disrupted → unstable
- Simultaneous C1 ring hypoplasia → spinal cord compression, myelomalacia secondary to canal stenosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Simple bifid atlas (minimal gaps) usually asymptomatic, discovered incidentally
  - Split atlas with larger arch defects more likely to be unstable → neck pain, torticollis, myelopathy
- Other signs/symptoms
  - Cervicomedullary ischemia, post-traumatic quadriplegia

Demographics

- Epidemiology
  - 0.1% of autopsy cases show split atlas

Natural History & Prognosis

- Largely depends on severity of ring defects, CVJ stability, C1 ring hypoplasia

Treatment

- Asymptomatic patients → observation
- Unstable, symptomatic patients → surgical stabilization

DIAGNOSTIC CHECKLIST

Consider

- Split atlas is rare congenital anomaly
  - Stability depends on anomaly severity, TAL integrity
- Search for associated skull base, CVJ anomalies

SELECTED REFERENCES

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KEY FACTS

TERMINOLOGY

- Synonyms: Klippel-Feil syndrome (KFS)
- Congenital spinal malformation characterized by segmentation failure of ≥ 2 cervical vertebrae ± thoracic, lumbar segmentation failure

IMAGING

- Single- or multiple-level congenital cervical segmentation and fusion anomalies
- C2-3 (50%) > C5-6 (33%) > CVJ, upper thoracic spine
- Vertebral bodies usually smaller than normal
- Vertebral body narrowing ("wasp waist") at fused rudimentary disc space ± fusion of posterior elements

TOP DIFFERENTIAL DIAGNOSES

- Juvenile idiopathic arthritis
- Surgical fusion
- Chronic sequelae of discitis
- Ankylosing spondylitis

PATHOLOGY

- No universally accepted etiology; embryonic insult postulated between 4th and 8th weeks
- Sporadic; familial genetic component with variable expression identified in many patients

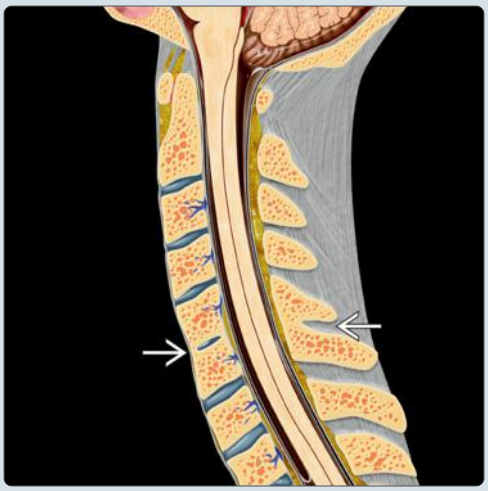
CLINICAL ISSUES

- Classic triad (33-50%): Short neck, low posterior hairline, and limited cervical motion
- In practice, however, wide variation in clinical, anatomical expression

DIAGNOSTIC CHECKLIST

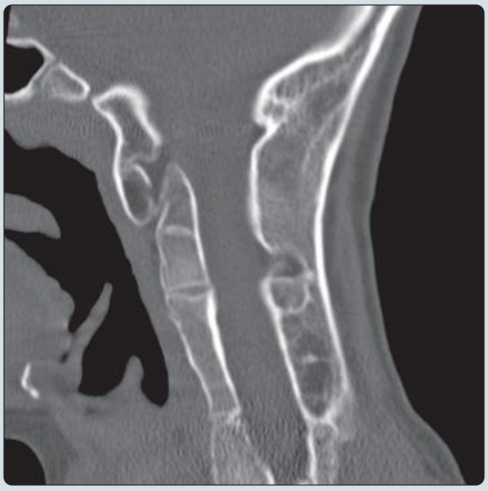
- Much KFS morbidity and nearly all mortality related to visceral system dysfunction
- Look for instability, progressive degenerative changes, cord/brainstem compression

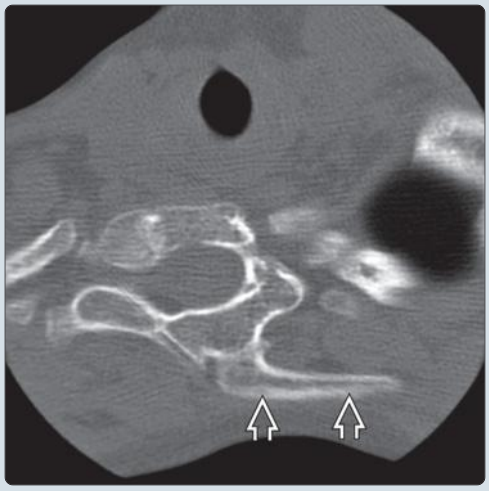
(Left) *Sagittal graphic (KFS 2) shows congenital fusion ➡ of the C5-C6 vertebrae and spinous processes, with characteristic rudimentary disc space and distinctive "waist" typical of congenital fusion.* **(Right)** *Lateral radiograph (KFS type 2) demonstrates typical C2/3 congenital segmentation failure ("fusion") with characteristic rudimentary disc space ➡ and fusion of the facets and spinous processes. The disc space appearance helps distinguish from postcervical fusion.*





(Left) *Sagittal bone CT (KFS 1) reveals extensive fusion of all cervical vertebrae and posterior elements, with the characteristic hypoplastic appearance of the vertebrae and intervertebral disc spaces. In this patient, there is additionally incorporation of C1 into the skull base ("occipitalization of the atlas").* **(Right)** *Axial bone CT (KFS 1, severe segmentation anomalies) demonstrates the vertebral articulation of a unilateral left omovertebral bone ➡.*





TERMINOLOGY

Synonyms

- Klippel-Feil syndrome (KFS)

Definitions

- Congenital spinal malformation characterized by segmentation failure of ≥ 2 cervical vertebrae ± thoracic, lumbar segmentation failure

IMAGING

General Features

- Best diagnostic clue
  - Single- or multiple-level congenital cervical segmentation and fusion anomalies
- Location
  - C2-3 (50%) > C5-6 (33%) > craniocervical junction, upper thoracic spine
- Size
  - Vertebral bodies < normal size with tapered contour at fused disc space
  - Rudimentary disc space reduced in height, diameter
- Morphology
  - Vertebral body narrowing ("wasp waist") at fused rudimentary disc space ± fusion of posterior elements

Radiographic Findings

- Radiography
  - 1 or more fused vertebral levels with rudimentary, narrow disc space(s)
    - Disc space always abnormal; frequently fused facets, spinous processes as well
    - Adjacent disc spaces at mobile levels ± degenerative changes
  - ± omovertebral bone
  - Flexion/extension films → lack of motion between fused segments, increased mobility at nonfused levels

CT Findings

- Bone CT
  - Typical osseous findings ± degenerative changes
  - Sagittal, transverse spinal canal diameter usually normal
    - Narrowing reflects secondary degenerative changes adjacent to fused segments
    - Canal enlargement → consider syringomyelia

MR Findings

- T1WI
  - Cervical fusion(s); vertebral bodies ± facets, posterior elements
  - ± degenerative changes; spondylosis, disc herniations common (especially lower cervical spine)
  - ± CVJ osseous anomalies, Chiari 1 malformation
- T2WI
  - Osseous findings same as T1WI; normal marrow signal
  - ± cord or nerve root compression, syringomyelia, brainstem abnormalities, myeloschisis

Imaging Recommendations

- Best imaging tool
  - Radiography to evaluate and follow instability, degenerative changes
  - MR to exclude cord compression, detect degenerative changes
- Protocol advice
  - Serial neutral and flex/extend plain radiographs to detect progressive instability, degenerative disease
  - Multiplanar MR to evaluate canal compromise, cord compression, soft tissue degenerative changes
  - Ultrasound or CECT to detect and characterize associated visceral organ abnormalities

DIFFERENTIAL DIAGNOSIS

Juvenile Idiopathic Arthritis

- Difficult to distinguish from cervical block vertebra
- Search for other affected joints, appropriate history

Surgical Fusion

- No disc space "waist," facets infrequently ankylosed
- Surgical history key to making diagnosis

Chronic Sequelae of Discitis

- Irregular endplate margins, no "waist," ± kyphosis
- Confirm history of prior spinal infection

Ankylosing Spondylitis

- Delicate contiguous syndesmophytes ("bamboo spine") + symmetric SI joint disease
- HLA-B27 positive (95%)

PATHOLOGY

General Features

- Etiology
  - No universally accepted etiology; embryonic insult postulated between 4th and 8th weeks
    - Putative environmental causative factors include teratogens, maternal alcoholism
    - Association with other syndromes (fetal alcohol, Goldenhar, Wildervanck: Cervico-occulo-acoustic)
- Genetics
  - Sporadic; familial genetic component with variable expression identified in many patients
    - C2/3 fusion (type 2) → autosomal dominant with variable penetrance
    - C5/6 fusion (type 2) → autosomal recessive
  - *SGMI* (chromosome 8) → 1st Klippel-Feil gene identified; gene expression overlaps all 3 KFS types
- Associated abnormalities
  - Hemivertebrae, butterfly vertebrae, spina bifida
  - Scoliosis (usually congenital) ± kyphosis (60%)
  - Odontoid dysplasia, basilar impression, C1 assimilation, occipitocervical instability
  - Syringomyelia, diastematomyelia (20%), Chiari 1 malformation (8%), neurenteric cyst or dermoid (rare)
  - Cervicomedullary neuroschisis ± synkinesis (20%)
  - Sprengel deformity ± omovertebral bone (15-30%); unilateral or bilateral
  - Sensorineural hearing loss (30%), external ear anomalies, GU tract abnormalities (35%), congenital heart disease (14%), upper extremity deformity, facial anomalies
- "Klippel-Feil syndrome" often colloquially used for all patients with cervical congenital fusion anomalies regardless of extent

- Congenital cervical fusion 2° to failure of normal segmentation of cervical somites (3rd-8th weeks)

Staging, Grading, & Classification

- Type 1 (9%): Massive fusion of cervical, upper thoracic spine → severe neurological impairment, frequent associated abnormalities
- Type 2 (84%): Fusion of ≥ 1 cervical vertebral interspaces
- Type 3 (7%): Fusions involve cervical and lower thoracic/lumbar vertebra

Gross Pathologic & Surgical Features

- Surgical observations correlate to imaging findings

Microscopic Features

- Histologically normal bone, disc

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Cosmesis complaints, neck or radicular pain, slowly progressive or acute myelopathy
    - Massive fusions often noted in infancy/early childhood 2° to cosmetic deformity
  - Neurologic problems in infancy, childhood usually 2° to CVJ abnormalities
  - Lower cervical fusions (unless massive) usually present ≥ 3rd decade when degenerative changes or instability of adjacent segments manifests
- Other signs/symptoms
  - Vocal impairment (usually > 1 level fusion)
  - Synkinesia (mirror movements): 20%, upper > lower extremity, diminish with time
- Clinical profile
  - Classic triad (33-50%): Short neck, low posterior hairline, and limited cervical motion
  - Wide variation in clinical and anatomical expression
    - Many patients have normal clinical appearance despite severity of involvement
    - Cervical motion limitation is most consistent clinical finding

Demographics

- Age
  - 2nd-3rd decade; spans entire range of life expectancy
- Gender
  - M ≤ F
- Epidemiology
  - 1/42,000 births

Natural History & Prognosis

- Progressive accelerated degenerative changes adjacent to fused segments
- 3 patterns at greatest risk for future instability
  - C0 → C3 fusion with occipitocervical synostosis
  - Long cervical fusion + abnormal C0/1 junction
  - Single open interspace between 2 fused segments
- ↑ risk of neurological injury following minor trauma 2° to hypermobility of cervical segments
  - High-risk patients: 2 sets of block vertebrae, occipitalization of atlas + basilar invagination, and cervical fusion + spinal stenosis

Treatment

- Avoidance of contact sports, occupations, and recreational activities at risk for head or neck trauma
- Activity modification, bracing, and traction may reduce symptoms
- Neurological lesion, significant pain despite conservative therapy, or progressive instability → decompression ± spinal fusion

DIAGNOSTIC CHECKLIST

Consider

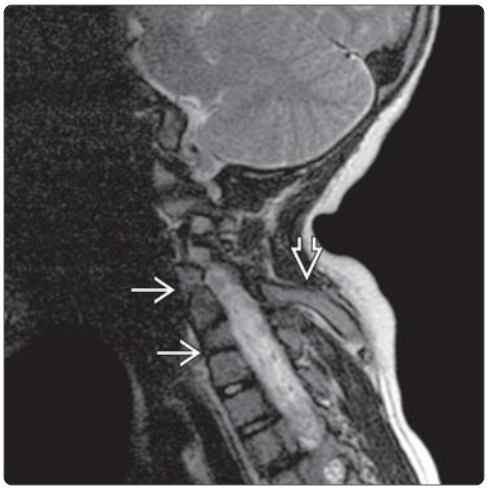
- Much KFS morbidity and nearly all mortality related to visceral system dysfunction

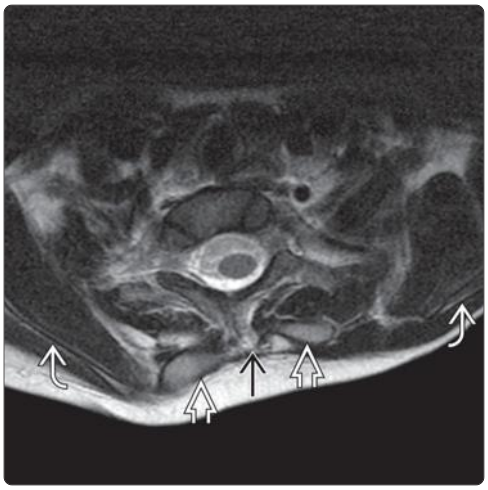
Image Interpretation Pearls

- Look for instability, progressive degenerative changes, cord/brainstem compression

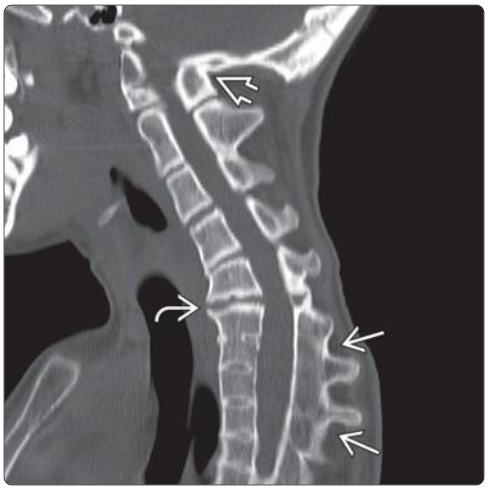
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(Left) *Sagittal T2WI MR (KFS 1, bilateral omovertebral bones) shows abnormal straightening of cervical lordosis and multiple vertebral segmentation anomalies. The omovertebral bone resides within the dorsal subcutaneous soft tissues and articulates with the posterior elements.* **(Right)** *Axial T2WI MR (KFS 1) depicts the course of bilateral omovertebral bones from the posterior spinal elements to the scapulae.*





(Left) *Sagittal reformatted bone CT of the cervicothoracic spine shows lack of segmentation of the cervical and thoracic spinal column. Note C1 spinous process fused to the occiput and fusion of bodies and posterior elements of C6-T4. Note accelerated disc degeneration at C5-C6, which is not fused.* **(Right)** *Sagittal T2WI MR (KFS 3) of the lumbar spine confirms that anomalous segmentation failure involves the lumbar spine as well as cervical and thoracic spine (not shown).*





(Left) *Sagittal T2WI MR (KFS 1) demonstrates multilevel severe cervical vertebral segmentation failure with hypoplastic vertebra and intervertebral discs as well as apparent thinning of the upper cervical spinal cord, which was confirmed on axial imaging to represent partial volume averaging of cervical diastematomyelia between the hemicords.* **(Right)** *Axial T2* GRE MR (KFS 1) confirms cervical diastematomyelia (type 2, no septum) as an associated finding.*

KEY FACTS

TERMINOLOGY

- Vertebral dysplasia, segmentation and fusion anomaly (SFA), "disorganized spine"
- Partial or complete failure of vertebral formation

IMAGING

- Sharply angulated, single curve, or focal (kypho)scoliosis
- Hemivertebra, butterfly vertebra generally smaller than normal vertebra

TOP DIFFERENTIAL DIAGNOSES

- Vertebral fracture, history critical
- Inherited spinal dysplasias

PATHOLOGY

- Deranged *PAX1* gene expression in developing vertebral column
- Many syndromes manifest vertebral dysplasia
- Associated anomalies include dysraphism, split notochord syndromes, visceral anomalies (61% of congenital scoliosis patients)

CLINICAL ISSUES

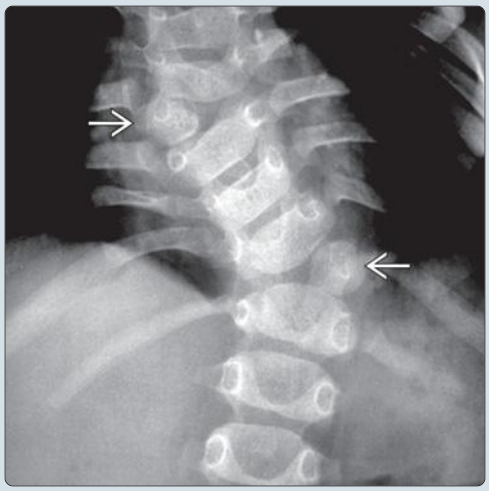
- Many asymptomatic or detected during scoliosis evaluation
- Syndromal patients usually detected in infancy
  - Abnormal spine curvature ± neural deficits, limb or visceral abnormalities
  - Respiratory failure (impeded chest movement 2° to fused ribs, kyphoscoliosis)

DIAGNOSTIC CHECKLIST

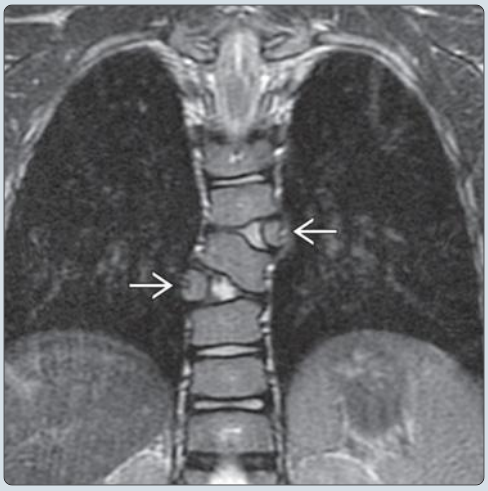
- Important to look for and characterize associated visceral anomalies
- Type of deformity determines propensity for scoliosis progression

(Left) *Coronal graphic of the thoracolumbar spine demonstrates several different types of vertebral formation failure, with segmented "balanced" L1 and L4 hemivertebrae ➡ and a T11 butterfly vertebra ➡.* **(Right)** *Anteroposterior radiograph shows segmented right T7 and left T11 hemivertebrae ➡ producing focal scoliotic curves. Because they mostly cancel each other's curve, this configuration is considered "balanced" for the purposes of prognostic and treatment planning.*





(Left) *Coronal T2WI MR demonstrates left T6 and right T8 hemivertebrae ➡. There is only minimal resultant leftward curvature because the 2 hemivertebrae "balance" each other, and this curvature would not be expected to be rapidly progressive.* **(Right)** *Axial T2WI MR depicts the abnormal intervertebral disc space appearance in the presence of a hemivertebra. Diagnosing hemivertebra is most straightforward in the coronal plane, and often difficult in the sagittal and axial planes.*





TERMINOLOGY

Synonyms

- Vertebral dysplasia, segmentation and fusion anomaly (SFA), "disorganized spine"

Definitions

- Partial or complete failure of vertebral formation
  - Partial formation failure → wedge vertebra
  - Complete formation failure → vertebral aplasia, hemivertebra, butterfly vertebra

IMAGING

General Features

- Best diagnostic clue
  - Sharply angulated, single curve, or focal scoliosis with deformed vertebral bodies
- Location
  - Thoracolumbar most common
- Size
  - Hemivertebra, butterfly vertebra generally smaller than normal vertebra
- Morphology
  - Incompletely formed vertebra; may be missing front, back, side, or middle of vertebral body

Radiographic Findings

- Radiography
  - Vertebral formation failure anomalies
  - ± scoliosis
    - Paired right and left hemivertebra → "balanced," scoliotic curves cancel out
    - 1 or more unilateral hemivertebra → "unbalanced," uncompensated scoliotic curve
  - Best modality for "counting" vertebral levels, determining presence and severity of scoliosis

CT Findings

- Bone CT
  - Sagittal or coronal vertebral clefts, hemivertebra, butterfly vertebra
    - Abnormal vertebra difficult to evaluate in axial plane; sagittal, coronal reformats helpful
  - ± posterior element dysraphism, fusion anomalies
    - Most readily evaluated in axial plane
  - 3D CT reformatted images best demonstrate vertebral anomalies for surgical planning

MR Findings

- T1WI
  - Vertebral formation failure anomalies
  - Normal marrow, disc signal intensity
- T2WI
  - Similar findings to T1WI
  - ± lipoma, tethered cord, diastematomyelia, spinal cord compression

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Long-cassette weightbearing radiographs → quantitate scoliosis, "counting" to definitively localize abnormal vertebral level
  - Multiplanar T1WI, T2WI MR → identify vertebral anomalies, evaluate spinal cord and soft tissues
    - Most vertebral and spinal cord anomalies seen best in coronal, sagittal planes
  - 3D bone CT useful to characterize scoliosis and vertebral anomalies for preoperative planning

DIFFERENTIAL DIAGNOSIS

Vertebral Fracture, History Critical

- Pathologic
- Traumatic
- 2 pedicles per level
- Noncorticated irregular fracture margins ± soft tissue edema, cord injury

Inherited Spinal Dysplasias

- Mucopolysaccharidosis
- Achondroplasia
- Distinguish by characteristic imaging, clinical, and laboratory findings

PATHOLOGY

General Features

- Etiology
  - Total aplasia: Both chondral centers fail to develop early in development
  - Lateral hemivertebra: 1 chondral center does not develop; ossification center subsequently fails to develop on that side
  - Sagittal cleft (butterfly) vertebra: Separate ossification centers form (but fail to unite) in each paired paramedian chondrification center
  - Coronal cleft vertebra: Formation and persistence of separate ventral and dorsal ossification centers
  - Posterior hemivertebra: Later failure during ossification stage
- Genetics
  - Deranged *PAX1* gene expression in developing vertebral column
  - Many syndromes manifest vertebral dysplasia
- Associated abnormalities
  - Dysraphism, split notochord syndromes
    - Diastematomyelia, syrinx, tethered cord/fatty filum, congenital tumor, visceral organ anomalies
  - Other developmental vertebral abnormalities
    - Partial duplication (supernumerary hemivertebra)
    - Segmentation failure (block vertebra, posterior element dysraphism, pediculate bar, neural arch fusion)
  - Visceral abnormalities; 61% of congenital scoliosis patients
- Normal vertebral formation occurs over 3 sequential periods
  - Membrane development: Segmental formation of medial sclerotome (vertebral bodies) and lateral myotome (paraspinal muscles)

- Chondrification: Sclerotomes separate transversely and join with adjacent sclerotomal halves → paired chondrification sites develop in vertebral bodies and neural arches
- Ossification: Chondral skeleton ossifies from single ossification center

- Segmentation and fusion anomalies result from aberrant vertebral column formation
  - Abnormal vertebrae may be supranumerary or replace normal vertebral body
  - More severe SFA → higher likelihood of associated visceral anomalies
- Imaging appearance of vertebral formation failure determined by deficient vertebral body portion
  - Anterior formation failure (common) → sharply angulated kyphosis
  - Posterior formation failure (rare) → hyperlordotic curve
  - Lateral formation failure (common) → classic hemivertebrae of congenital scoliosis
- Hemivertebra variants subclassified as incarcerated, nonincarcerated, segmented, nonsegmented, or semisegmented
  - Incarcerated: Vertebral bodies above and below shaped to accommodate hemivertebrae → do not generally produce scoliosis
  - Nonincarcerated: Found at scoliosis apex, curve magnitude depends on size of wedged segment
  - Segmented ("free"): Normal discs above and below hemivertebra → progressive scoliosis due to unbalanced growth
  - Nonsegmented: Lack disc spaces between wedged, normal adjacent vertebral bodies
  - Semisegmented: Normal disc space on 1 side, nonsegmented on other side
- Segmental spinal dysgenesis (SSD) = rare congenital abnormality in which 1 segment of vertebral column/spinal cord fails to develop properly
  - Spinal cord thinned or indiscernible at abnormality level; bulky, low-lying cord segment caudad to focal abnormality in most cases
  - Severity of morphologic derangement correlates with residual spinal cord function, severity of clinical deficit

Gross Pathologic & Surgical Features

- Normal bone density unless concurrent metabolic abnormality
- Surgical observations of vertebral configuration matches imaging findings

Microscopic Features

- Normal bone histology unless concurrent metabolic abnormality

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Abnormal spine curvature ± neural deficits, limb or visceral abnormalities
- Other signs/symptoms
  - Respiratory failure (impeded chest movement 2° to fused ribs, kyphoscoliosis) → rare
- Clinical profile
  - Most are asymptomatic or detected during scoliosis evaluation
  - Syndromal patients usually detected in infancy

Demographics

- Age
  - Usually diagnosed in infancy → early childhood
  - Mild cases may present in adulthood
- Gender
  - M ≈ F
- Epidemiology
  - Isolated or syndromal, singular or multiple
  - Incidence increased with parental consanguinity, concurrent multisystem anomalies

Natural History & Prognosis

- Scoliosis is frequently progressive
  - Expectant watching, early intervention if warranted to prevent development of severe deformities

Treatment

- Conservative in mild cases (orthotics, observation)
- Surgical resection &/or fusion to arrest/reverse kyphoscoliosis in moderate to severe cases

DIAGNOSTIC CHECKLIST

Consider

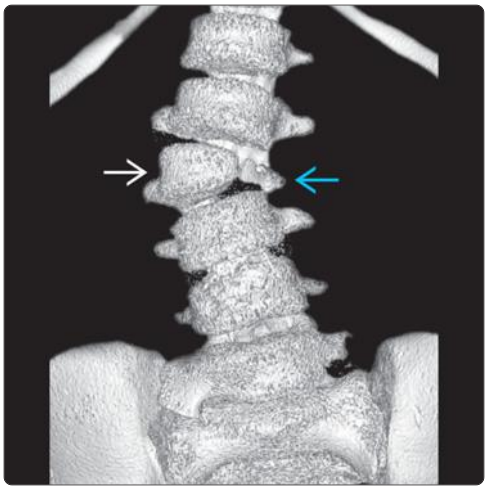
- Syndromal origin or association → important to look for and characterize visceral anomalies

Image Interpretation Pearls

- Look for concurrent segmentation failure, other neural and visceral anomalies
- Type of deformity determines propensity for scoliosis progression

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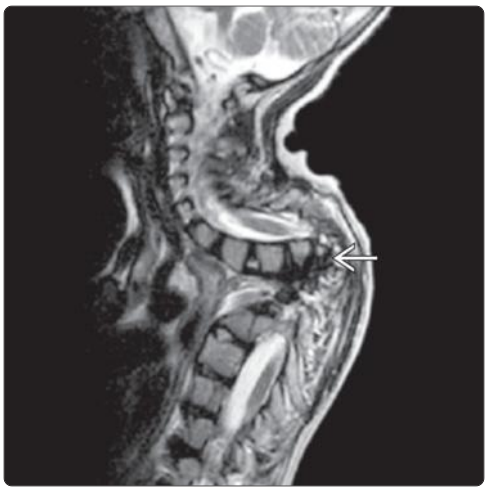


(Left) *Coronal 3D bone CT demonstrates an unbalanced right L3 hemivertebra resulting in focal convex right scoliotic curvature. In this instance, there are rudimentary left L3 pedicle and posterior elements.* **(Right)** *Coronal T2WI MR of the thoracolumbar spine obtained for scoliosis evaluation reveals a segmented, unbalanced right L3 hemivertebra. The unilateral unbalanced configuration produces convex right focal congenital scoliosis.*





(Left) *Coronal T1WI MR (myelomeningocele, scoliosis) reveals convex right thoracic curvature, with multiple abnormally formed vertebra in the mid and lower thoracic spine. This patient also had associated diastematomyelia (not shown).* **(Right)** *Axial T2WI MR (myelomeningocele, diastematomyelia) at the T10 level shows a bizarre, abnormally widened vertebra representing a butterfly vertebra with abnormal intervertebral disc. Note that the right hemicord shows syringomyelia.*





(Left) *Sagittal T2WI MR demonstrates severe acute angle upper thoracic kyphosis. There is markedly hypoplastic anterior hemivertebra at the curve apex as well as jumbled appearance of other vertebra indicating multiple vertebral formation and segmentation anomalies.* **(Right)** *Axial T1WI MR (severe kyphoscoliosis) reveals a normal spinal cord with adjacent osseous spinal canal narrowing related to multiple congenital vertebral formation anomalies.*

Vertebral Segmentation Failure

KEY FACTS

TERMINOLOGY

- Synonyms: Segmentation anomaly, segmentation and fusion anomaly (SFA), "block vertebra"

IMAGING

- Sharply angulated focal scoliotic curvature with abnormal "fused" vertebra
  - Ranges single level → extensive multilevel involvement
  - Lumbar > cervical > thoracic
- May have scoliosis, kyphosis, cord compression
- Search for related fused pedicles, ribs, posterior elements

TOP DIFFERENTIAL DIAGNOSES

- Juvenile chronic arthritis
- Surgical vertebral fusion
- Chronic sequelae of discitis
- Ankylosing spondylitis

PATHOLOGY

- Deranged *PAX1* gene expression → abnormal notochord signaling in developing vertebral column
- Many syndromes associated with SFA
- Associations include other neuraxis anomalies, renal, gastrointestinal, congenital cardiac defects

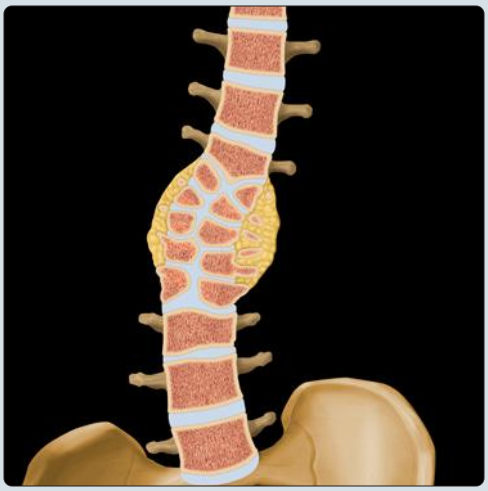
CLINICAL ISSUES

- Usually asymptomatic or present with kyphoscoliosis
- Less commonly neural deficit, limb or visceral anomalies, respiratory failure
- Scoliosis frequently progressive

DIAGNOSTIC CHECKLIST

- Clinical manifestations variable, determined by type of SFA and syndromal association
- Block vertebra usually larger than single normal vertebral body

(Left) *Coronal graphic of the thoracolumbar spine demonstrates multilevel failure of vertebral segmentation producing congenital scoliosis. Space between the dysplastic vertebra is filled with cartilage and aberrant disc material.* **(Right)** *Coronal bone CT (congenital scoliosis) shows multiple examples of vertebral segmentation failure with several right-sided hemivertebra ⇨ that have failed to successfully segment, producing a jumble of malformed vertebra and multiple curve scoliosis.*





(Left) *Coronal 3D bone CT reformat shows multiple curve scoliosis with convex right thoracic and convex left thoracolumbar kyphoscoliosis resulting from multiple levels of vertebral segmentation failure. 3D CT is best for fully characterizing contribution of the various anomalous vertebra to scoliosis and kyphosis for treatment planning.* **(Right)** *Sagittal 3D CT reformat demonstrates focal kyphosis at the thoracolumbar junction ⇨ related to multilevel vertebral segmentation failure.*





TERMINOLOGY

Synonyms

- Segmentation anomaly, segmentation and fusion anomaly (SFA), "block vertebra," "disorganized spine"

Definitions

- Vertebral column malformations (block vertebra, neural arch fusion, pediculate bar) resulting from deranged embryological development → failure of normal segmentation

IMAGING

General Features

- Best diagnostic clue
  - Sharply angulated focal scoliotic curvature with abnormal "fused" vertebra
- Location
  - Lumbar > cervical > thoracic
- Size
  - Ranges from single level → extensive multilevel involvement
  - Block vertebra usually larger than a single normal vertebral body
- Morphology
  - Incomplete vertebral body segmentation, fused posterior elements, and large misshapen vertebral bodies incorporating 1 or more levels

Radiographic Findings

- Radiography
  - Deformed, fused vertebra ± scoliosis, kyphosis
  - Upright images with lateral and AP bending assess weightbearing effect on scoliosis

CT Findings

- Bone CT
  - Incomplete vertebral body segmentation
    - Large, misshapen vertebral bodies incorporating 1 or more levels
    - ± fused pedicles, ribs, posterior elements

MR Findings

- T1WI
  - Deformed, fused vertebral bodies and posterior elements
  - ± scoliosis
  - Normal marrow signal intensity
- T2WI
  - Findings similar to T1WI
  - ± scoliosis, cord compression

Imaging Recommendations

- Best imaging tool
  - Weightbearing plain radiographs to evaluate scoliosis, "count" abnormal vertebral levels
  - MR imaging
    - Multiplanar T1WI to evaluate vertebral anatomy
      - Vertebral anomalies seen best in coronal, sagittal planes
    - T2WI to evaluate spinal cord pathology, compression
  - 3D CT reformats to characterize osseous structures, preoperative planning

DIFFERENTIAL DIAGNOSIS

Juvenile Chronic Arthritis

- Difficult to distinguish from cervical block vertebra
- Search for other affected joints, appropriate clinical history

Surgical Fusion

- No "waist" at disc space level; facets infrequently ankylosed
- Surgical history key to making diagnosis

Chronic Sequelae of Discitis

- Cortical endplate margins irregular, "waist" absent
- Search for history of prior spinal infection

Ankylosing Spondylitis

- Delicate contiguous syndesmophytes ("bamboo spine") + symmetric SI joint disease
- HLA-B27 positive (95%)

PATHOLOGY

General Features

- Etiology
  - Segmentation and fusion anomalies result from aberrant vertebral column formation
  - Mildest (and most common) form is indeterminate (transitional) vertebrae at thoracolumbar, lumbosacral transition
- Genetics
  - Deranged *PAX1* gene expression → abnormal notochord signaling in developing vertebral column
  - Many syndromes associated with SFA
    - Klippel-Feil (cervical SFA): Common, gene locus 8q22.2
    - Spondylothoracic dysplasia (Jarcho-Levin): Uncommon, thoracic spine fusions in crab-like array with multiple rib fusions
- Associated abnormalities
  - Other neuraxis anomalies (40%)
    - Dysraphism, split notochord syndrome
    - Partial or complete failure of formation (vertebral aplasia, hemivertebra, butterfly vertebra)
    - Partial duplication (supernumerary hemivertebra)
  - Scoliosis
  - Renal, gastrointestinal, congenital cardiac defects
- Normal embryology: Vertebral formation occurs over 3 sequential periods
  - Membrane development: Segmental formation of medial sclerotome (vertebral bodies) and lateral myotome (paraspinal muscles)
  - Chondrification: Sclerotomes separate transversely and join with adjacent sclerotomal halves → paired chondrification sites develop in vertebral bodies, neural arches
  - Ossification: Chondral skeleton ossifies from single ossification center

Staging, Grading, & Classification

- Block vertebra: Failure of ≥ 2 vertebral somites to segment
  - Combined vertebrae may be normal height, short, or tall
  - Disc space frequently rudimentary or absent

- Frequent association with hemivertebra/absent vertebra above or below block level, posterior element fusion
- Posterior neural arch anomalies
  - Failure to unite in midline → dysraphism (± unilateral pedicle aplasia/hypoplasia)
  - Unfused spinous processes; L5, S1 > C1 > C7 > T1 > lower thoracic spine
  - Multiple level posterior fusion → congenital vertebral bar

Gross Pathologic & Surgical Features

- Normal bone density unless concurrent metabolic abnormality
- Surgical observations of vertebral configuration matches imaging findings

Microscopic Features

- Normal bone histology unless concurrent metabolic abnormality

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Kyphoscoliosis
- Other signs/symptoms
  - Neural deficits (usually myelopathic), limb or visceral anomalies
  - Respiratory failure (rare, 2° impeded chest movement due to severe scoliosis, rib cage fusion)
- Clinical profile
  - Asymptomatic incidental detection or presents for evaluation of abnormal spine curvature

Demographics

- Age
  - Severe cases detected during infancy/childhood; mild cases may present as adults
- Gender
  - M ≈ F; dependent on syndromal association
- Epidemiology
  - Isolated or syndromal, singular or multiple
  - SFAs account for 18% of scoliosis; incidence higher with multisystem abnormalities, parental consanguinity

Natural History & Prognosis

- Scoliosis frequently progressive
  - Unilateral unsegmented bar with contralateral hemivertebrae → rapidly progressive, severely deforming congenital scoliosis
  - Isolated block vertebra rarely produce scoliosis
- Abnormal segments may continue to fuse

Treatment

- Conservative in mild cases (orthotics, observation)
- Surgical fusion to arrest/reverse kyphoscoliosis in moderate to severe cases

DIAGNOSTIC CHECKLIST

Consider

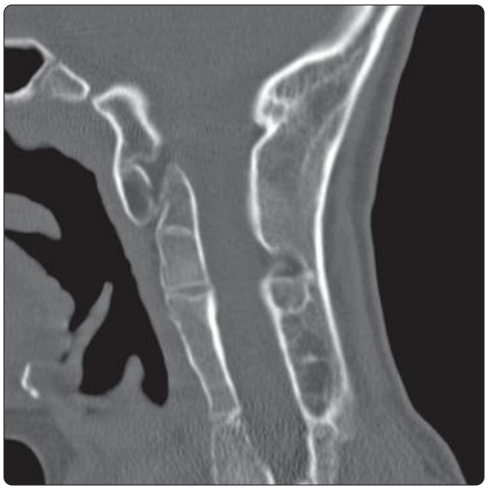
- Clinical manifestations variable, determined by type of SFA and syndromal association

Image Interpretation Pearls

- Coronal MR, AP radiography best for detecting and characterizing SFAs, "counting" abnormal vertebral levels

SELECTED REFERENCES

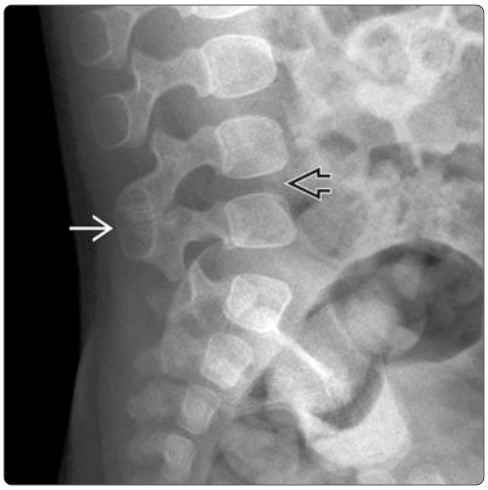
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(Left) *Sagittal bone CT (severe Klippel-Feil syndrome) reveals fusion at all cervical levels, as well as incorporation of C1 into the skull base. The narrow vertebra are typical of congenital segmentation failure.* **(Right)** *Coronal bone CT (severe Klippel-Feil spectrum) show dens hypoplasia and fusion of the right occipital condyle, C1 lateral mass, and C2 lateral mass. The left C1 lateral mass is not incorporated into either the occipital condyle or C2 lateral mass.*





(Left) *Sagittal T2WI MR (Chiari 2 malformation) shows characteristic posterior fossa malformation findings with vermian extension ⇨ into the upper cervical spinal canal and vertebral segmentation failure of C7 and T1 ⇨ with fusion of the spinous processes and facets.* **(Right)** *Lateral radiograph demonstrates single level segmentation failure of the L4 and L5 spinous processes and lamina ⇨. The L4/5 intervertebral disc space ⇨ is also abnormally narrow and hypoplastic.*





(Left) *Sagittal T2WI depicts congenital segmentation failure of L4 and L5 ⇨ with abnormal straightening of lumbar lordosis and mild focal kyphosis. The remaining lumbar vertebra show normal appearance.* **(Right)** *Sagittal T1WI shows congenital segmentation failure at L1-L4 ⇨. There are rudimentary residual disc spaces and abnormal lumbar kyphosis. The spinal cord ends at L1-L2 but is tethered by a thickened fatty infiltrated filum terminale ⇨.*

KEY FACTS

TERMINOLOGY

- Synonyms: Split cord malformation (SCM), "diastem"

IMAGING

- Sagittal division of spinal cord into 2 hemicords, each with 1 central canal, dorsal horn, and ventral horn
  - Hemicords usually reunite above and below cleft
  - ± fibrous or osseous spur
- Frequently associated vertebral segmentation anomalies
- Imaging evaluation
  - Consider ultrasound to screen infants with skin dimple or cutaneous marker
  - MR imaging most definitive for characterization
  - Supplement with bone CT ± myelography to optimally define spur anatomy for surgical planning

TOP DIFFERENTIAL DIAGNOSES

- Duplicated spinal cord (diplomyelia)

PATHOLOGY

- "Split notochord syndrome" is spectrum of diastematomyelia, dorsal enteric fistula/sinus, and dorsal enteric cysts/diverticula
- Spinal cord split into symmetric or asymmetric hemicords
- Either 1 (type II) or 2 (type I) dural tubes

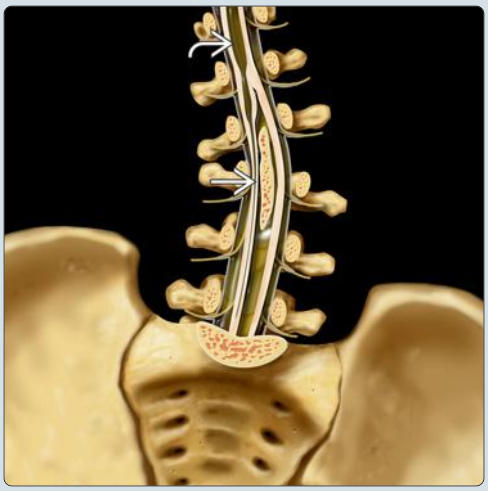
CLINICAL ISSUES

- May be clinically indistinguishable from other causes of tethered spinal cord in absence of cutaneous stigmata
  - Cutaneous stigmata indicate diastematomyelia level (> 50%); "fawn's tail" hair patch most common

DIAGNOSTIC CHECKLIST

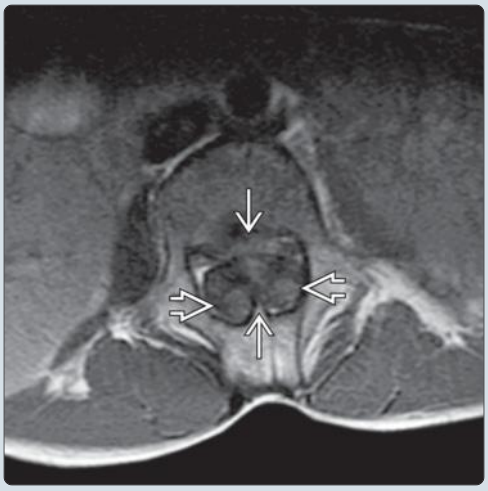
- Search for DSM in patients with cutaneous stigmata, intersegmental fusion of posterior elements, clinical tethered cord
- Presence of spur = type I SCM; more severe symptoms and anomalies, worse prognosis

(Left) *Coronal graphic of the lumbar spine demonstrates a type II SCM, with osseous spur ➡ splitting the low-lying syringomyelic ➡ spinal cord. Both hemicords continue into the sacrum.* **(Right)** *Sagittal T1WI MR (type I SCM) shows a large osseous spur ➡ extending from the L2 vertebral body to the dysplastic posterior elements. The spinal cord is low-lying and tethered by a lipoma ➡ and extradural arachnoid cyst ➡. Note solitary pelvic kidney ➡ anterior to the sacrum.*





(Left) *Axial T1WI MR demonstrates "splitting" of the spinal cord into 2 symmetric hemicords ➡ traversing around a large osseous spur ➡.* **(Right)** *Axial T2WI MR confirms characteristic "splitting" of the abnormal spinal cord into 2 symmetric hemicords ➡. The prominent osseous spur ➡ classifies this patient as a type I SCM.*





TERMINOLOGY

Synonyms

- Split cord malformation (SCM), "diastem"

Definitions

- Sagittal division of spinal cord into 2 hemicords, each with 1 central canal, dorsal horn, and ventral horn

IMAGING

General Features

- Best diagnostic clue
  - Fibrous or osseous spur splits spinal cord into 2 hemicords
    - Split cord and spur often occur in conjunction with intersegmental fusion
- Location
  - Thoracolumbar cleft (85% between T9 and S1) > > upper thoracic, cervical clefts
- Size
  - Range focal → extensive
- Morphology
  - Hemicords usually reunite above and below cleft
  - ± spur (fibrous, osteocartilaginous, or osseous), thickened filum, cord tethering

Radiographic Findings

- Radiography
  - Quantify kyphoscoliosis, "count" level of vertebral segmentation anomalies
  - Detects spur in < 50% of cases

CT Findings

- NECT
  - Osseous spur often visible; fibrous spurs frequently occult
  - Vertebral segmentation anomalies

MR Findings

- T1WI
  - 2 hemicords ± syringohydromyelia (50%)
  - ± isointense (fibrous) or hyperintense (osseous) spur
- T2WI
  - 2 hemicords ± syringohydromyelia (50%) in 1 or both cords, surrounded by bright CSF
  - Hypointense (fibrous or osseous) spur; fibrous spur best seen in axial or coronal plane
- T2* GRE
  - Myelographic effect produces bright CSF adjacent to dark bones, highlights hypointense spur

Ultrasonographic Findings

- Grayscale ultrasound
  - Obstetrical ultrasound → "extra" posterior echogenic focus, splaying of fetal posterior spinal elements
  - Postnatal imaging → spur, tethered cord

Nonvascular Interventions

- Myelography
  - Split cord; 1 or 2 dural tubes
  - Excellent delineation of spur location and meningocele manqué (if present); most useful in conjunction with CT

Imaging Recommendations

- Best imaging tool
  - MR imaging
- Protocol advice
  - Consider ultrasound to screen infants with skin dimple or cutaneous marker
  - MR imaging most definitive
    - Coronal, axial images best demonstrate hemicords, spur
    - T1WI to evaluate for filum lesions (e.g., fibrolipoma), vertebral anomalies
    - T2WI to determine number of dural sacs, ± syringohydromyelia
    - T2* GRE to detect spur
- Supplement with bone CT ± myelography to optimally define spur anatomy for surgical planning
  - Sagittal, coronal reformats excellent for depicting osseous anatomy, extent of spur

DIFFERENTIAL DIAGNOSIS

Duplicated Spinal Cord (Diplomyelia)

- 2 complete spinal cords, each with 2 anterior and 2 posterior horns and roots
  - Exceedingly rare, seen only in presence of spinal canal duplication; many authors dispute its true existence, believe it represents a severe form of diastematomyelia

PATHOLOGY

General Features

- Etiology
  - "Split notochord syndrome" spectrum
    - Congenital splitting of notochord → "split notochord syndrome" spectrum; diastematomyelia, dorsal enteric fistula/sinus, and dorsal enteric cysts/diverticula
  - Notochord directly influences vertebral body formation → segmentation anomalies
    - Lateral notch produces hemivertebra
    - Cleft produces butterfly vertebra
- Genetics
  - Sporadic; rare familial cases described
- Associated abnormalities
  - Other split notochord syndromes (20%)
  - Congenital spinal deformities (85%)
    - Segmentation and fusion anomalies (SFA)
    - Intersegmental laminar fusion (60%); virtually pathognomonic for diastematomyelia
    - Spinal dysraphism (myelocele/myelomeningocele 15-25%, hemimyelocele 15-20%)
    - Tethered spinal cord (75%); thickened filum terminale (40-90%)
    - Syringohydromyelia (50%) 1 or both hemicords, usually above diastematomyelia
    - Congenital scoliosis (79%)
    - 15-20% of Chiari 2 malformations
    - Spinal lipoma (26%)
    - Dermoid cyst (13%)
- Septum inhibits normal spinal cord movement during activity → symptom progression

- Spinal cord damage occurs via direct pressure and by traction → cord ischemia
- Cleft splits cord, with single cord above and below split
  - Most (91%) hemicords reunite caudally to terminate in a single filum terminale
- Roots may adhere to dura → "meningocele manqué"

Staging, Grading, & Classification

- Pang type I SCM
  - Separate dural sac, arachnoid space surrounds each hemicord
  - Osseous/fibrous spur
  - More commonly symptomatic than type II
- Pang type II SCM
  - Single dural sac, arachnoid space
  - No osseous spur; ± adherent fibrous bands tether cord
  - Rarely present with symptoms unless hydromyelia, tethering

Gross Pathologic & Surgical Features

- Sagittal spinal cord split into symmetric or asymmetric hemicords, residing in either 1 or 2 dural tubes
  - Symmetric: Each hemicord contains 1 central canal, dorsal horn/root, and 1 ventral horn/root surrounded by pial layer
  - Asymmetric: Division of anterior or posterior hemicord ("partial diastematomyelia")
- 2 dural tubes, each with own pial, arachnoid, and dural sheaths for several spinal segments (50%)
  - Bony or cartilaginous spur at inferior cleft, originating from lamina or vertebral body
  - Vertebral anomalies more severe (block or butterfly vertebrae, hemivertebrae, and posterior spina bifida)
  - Hydromyelia common
- Single dural tube, subarachnoid space (50%)
  - No bony spur; nearly always have fibrous band coursing through inferior cleft inserting into dura → tethers cord
  - Vertebral anomalies less severe (usually butterfly vertebrae)
  - Nerve roots may become adherent to dura and tether cord → "meningocele manqué"

Microscopic Features

- Single, ependymal lined central canal, ventral horn/root, and dorsal horn/root per hemicord
- Septal spur may be composed of mainly cortical bone (most common), mainly cancellous bone, or bone + soft tissues

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Clinically indistinguishable from other causes of tethered cord
  - Cutaneous stigmata high on back indicates diastematomyelia level (> 50%); "fawn's tail" hair patch most common
  - Other signs/symptoms
    - Progressive kyphoscoliosis in older children, adults
    - Orthopedic foot problems (50%); especially clubfoot
    - Urologic dysfunction
- Clinical profile
  - Mild cases normal ± cutaneous stigmata
  - Severe cases → kyphoscoliosis, neurological and musculoskeletal abnormalities

Demographics

- Age
  - Diagnosis in childhood; adult presentation uncommon
- Gender
  - Pediatric: F > > M
  - Adult: M < F (1:3.4)
- Epidemiology
  - 5% of congenital scoliosis

Natural History & Prognosis

- Stable or progressive disability if untreated
  - Late onset or previously stable patients may become symptomatic following relatively minor back injury or surgery requiring spinal manipulation
- ≥ 90% of patients stabilize or improve following surgery
  - Caveat: Scoliosis rarely affected by surgical untethering

Treatment

- Surgical tethered cord release, spur resection, and dural repair for progressive symptoms, prophylactic precursor to scoliosis surgery (particularly type I SCM)

DIAGNOSTIC CHECKLIST

Consider

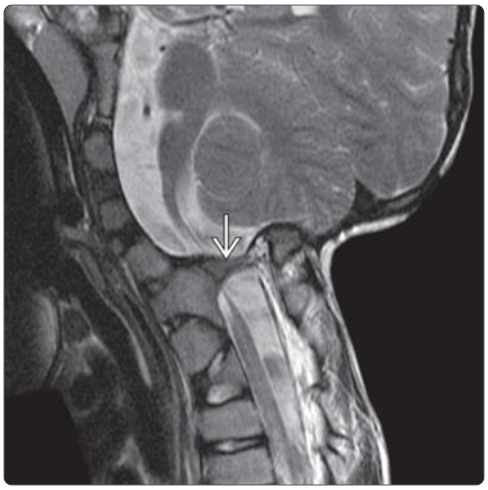
- Scrutinize images for presence of spur; type I DSM generally has more severe symptoms and anomalies, worse prognosis
- Search for DSM in patients with cutaneous stigmata, intersegmental fusion of posterior elements, clinical tethered cord

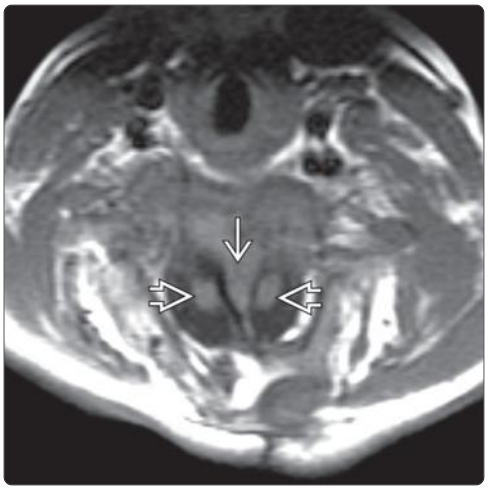
Image Interpretation Pearls

- Segmentation anomalies with intersegmental laminar fusion virtually pathognomonic for diastematomyelia

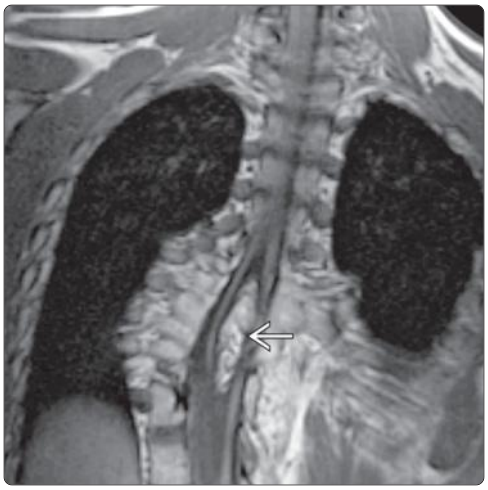
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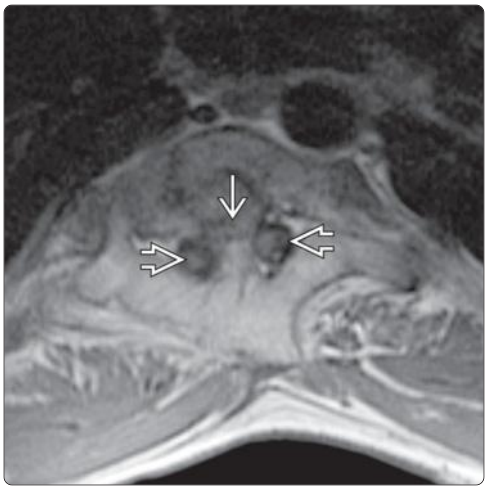
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(Left) *Sagittal T2WI MR (type I SCM) demonstrates severe craniocervical vertebral segmentation failure and a prominent osseous spur ➔ that pierces the high cervical dural tube. The 2 hemicords are laterally displaced out of this slice.* **(Right)** *Axial T1WI MR (type I SCM) reveals a prominent cervical osseous spur ➔ piercing the dural sac and splitting the 2 cervical hemicords ➔.*





(Left) *Coronal T1WI MR (type I SCM) reveals multiple vertebral segmentation anomalies, with a large midline osseous spur ➔ that splits the thoracic spinal cord into 2 hemicords. Note associated multiple posterior element and rib fusions.* **(Right)** *Axial T1WI MR (severe type I SCM) confirms a thick midline osseous spur ➔ that divides the spinal cord into 2 hemicords ➔, each contained within their own dural tube.*





(Left) *Sagittal T1WI MR (type II SCM, myelomeningocele closed at birth) demonstrates expected postoperative findings of the dorsal lumbar spine following myelomeningocele closure, with patulous distal thecal sac and adherence of the elongated spinal cord placode ➔ to the dorsal thecal sac closure at L4-L5.* **(Right)** *Axial T2WI MR (type II SCM, myelomeningocele) shows splitting of the thoracic spinal cord into 2 hemicords ➔ without intervening osseus or fibrous spur.*

Partial Vertebral Duplication

KEY FACTS

TERMINOLOGY

- Partial duplication of vertebral column → 1 or more supranumerary ("extra") vertebra

IMAGING

- Atypical scoliosis (sharply angulated, single curve, or focal) with 1 or more "extra" hemivertebra
- "Extra" hemivertebra frequently associated with focal scoliotic curvature

TOP DIFFERENTIAL DIAGNOSES

- Butterfly vertebra
- Vertebral fracture

PATHOLOGY

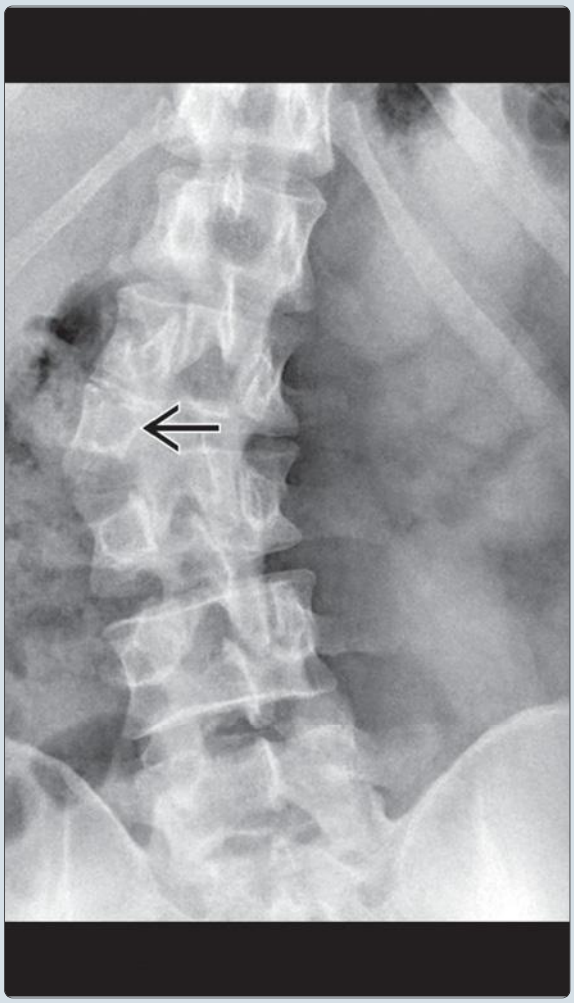
- Derangement of normal vertebral ossification leads to hemivertebra
- Cervicothoracic or thoracolumbar junction segmentation variation → "extra" counted hemivertebra
- Supernumerary hemivertebra may be incorporated into adjacent vertebra producing unbalanced block vertebra, focal scoliosis

CLINICAL ISSUES

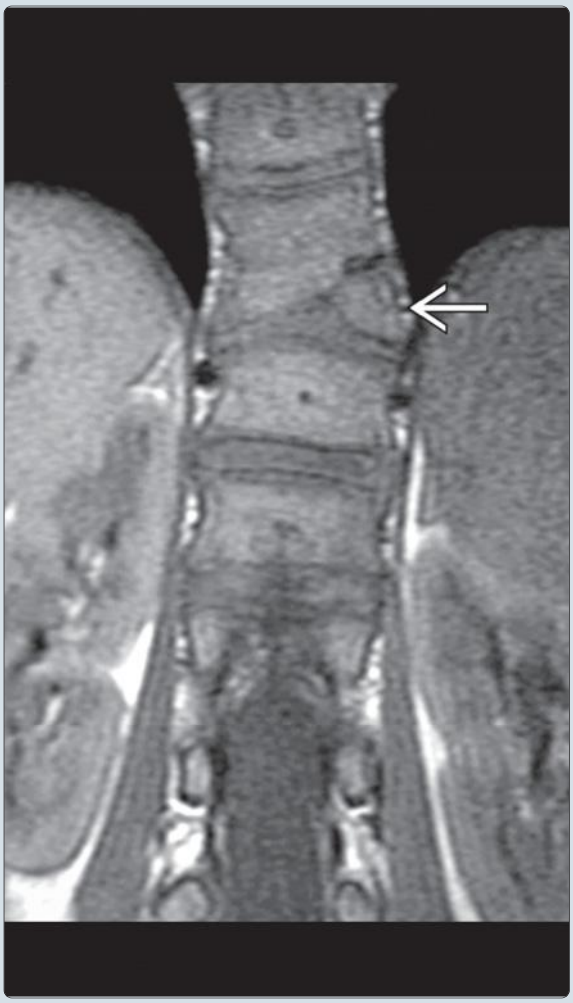
- May be asymptomatic or present with neuromuscular scoliosis
- Respiratory failure 2° to impeded chest movement from severe scoliosis (rare)

DIAGNOSTIC CHECKLIST

- Patient may either appear normal or present with scoliosis ± stigmata of congenital disorder
- Supranumerary hemivertebra demonstrates "missing" pedicle on hypoplastic side; distinguishes from fracture



Anteroposterior radiography depicts a typical supernumerary hemivertebra ⇨ positioned between the right L2 and L3 vertebral levels, resulting in convex right scoliotic curvature.



Coronal T1WI MR depicts a small left thoracic supernumerary hemivertebra ➡ producing minimal convex left lower thoracic scoliosis.

TERMINOLOGY

Synonyms

- Supranumerary vertebra

Definitions

- Partial duplication of vertebral column → 1 or more supranumerary ("extra") vertebra

IMAGING

General Features

- Best diagnostic clue
  - Atypical scoliosis (sharply angulated, single curve, or focal) with 1 or more "extra" hemivertebra
- Location
  - Thoracolumbar > cervical

Radiographic Findings

- Radiography
  - Supranumerary lateral hemivertebra with scoliosis

CT Findings

- Bone CT
  - Supranumerary lateral hemivertebra

MR Findings

- T1WI
  - Supranumerary lateral hemivertebra ± dysraphism, spinal cord anomalies
- T2WI
  - Same as T1WI

Imaging Recommendations

- Best imaging tool
  - Anteroposterior plain radiograph
- Protocol advice
  - AP plain film for "counting" to localize abnormal vertebral level
  - Long-cassette standing weight-bearing radiograph to quantitate scoliosis
  - Multiplanar MR imaging to evaluate vertebral anatomy, assess for associated neurological abnormalities

DIFFERENTIAL DIAGNOSIS

Butterfly Vertebra

- 2 pedicles; looks like "bilateral hemivertebra" with central cleft
- Separate bilateral ossification centers fail to unite

Vertebral Fracture

- Pathologic or traumatic (history critical)
- Noncorticated irregular margins ± soft tissue edema, spinal cord injury
- 2 pedicles at abnormal level (unlike hemivertebra)

PATHOLOGY

General Features

- Etiology
  - Derangement of normal vertebral ossification → hemivertebra
  - Cervicothoracic or thoracolumbar junction segmentation variation → "extra" counted hemivertebra
- Associated abnormalities
  - Dysraphism, split notochord syndromes, caudal regression, other vertebral segmentation anomalies, visceral organ anomalies

Gross Pathologic & Surgical Features

- Hemivertebra may be incorporated into adjacent vertebra → unbalanced block vertebra

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Neuromuscular scoliosis; usually progressive
- Other signs/symptoms
  - Respiratory failure 2° to impeded chest movement from severe scoliosis (rare)
  - Neural deficits, limb/visceral defects

Demographics

- Age
  - More severe cases detected in infancy or childhood
  - Milder cases detected in adolescence during school scoliosis checks, by pediatrician, or incidentally during imaging
- Gender
  - M = F
- Epidemiology
  - Isolated or syndromal, singular or multiple
    - Many syndromes associated with segmentation and fusion anomalies (SFA)
    - Increased incidence when multisystem congenital anomalies, parental consanguinity present

Natural History & Prognosis

- Variable: Scoliosis may progress with somatic growth, require treatment

Treatment

- Conservative management in mild cases
- Surgical management to arrest/reverse scoliosis in more severe cases

DIAGNOSTIC CHECKLIST

Consider

- Exclude associated abnormalities

Image Interpretation Pearls

- Supranumerary hemivertebra "missing" pedicle on hypoplastic side; distinguishes from fracture

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KEY FACTS

TERMINOLOGY

- Spinous process/lamina unfused in midline without underlying neural or dural abnormality
- "Spina bifida occulta" outdated term that should be abandoned

IMAGING

- Spinous process/lamina fail to fuse in midline
  - No abnormal neural, dural, or lipomatous tissue within posterior osseous defect
  - Normal dural sac, conus position, and filum thickness
- Lumbosacral junction (L5 > S1) > > cervical (C1 > C7 > T1), thoracic

TOP DIFFERENTIAL DIAGNOSES

- Closed spinal dysraphism (CSD)
- Surgical laminectomy defect
- Normal progression of incomplete ossification

PATHOLOGY

- Etiology unknown
  - Probably not linked to abnormal neurulation process

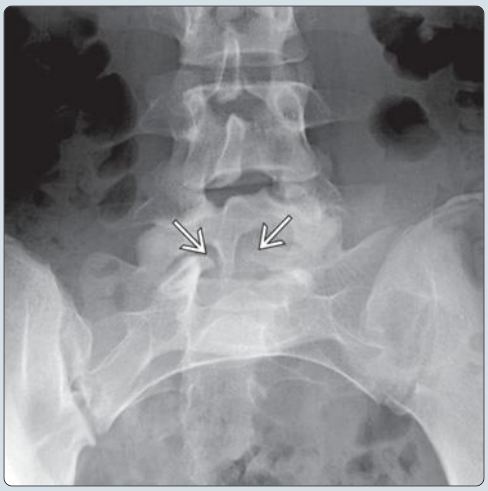
CLINICAL ISSUES

- Most commonly, patient presents with low back or leg pain; finding is incidentally noted during imaging
- Occasionally, skin dimple observed on back
- Usually asymptomatic if no cutaneous stigmata

DIAGNOSTIC CHECKLIST

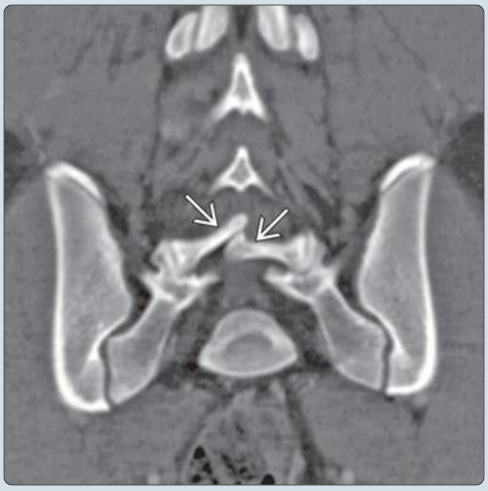
- Incomplete posterior fusion is incidental finding that is very rarely of neurological significance
  - Plain films economical screening tool, but positive finding rarely significant
  - MR best for exclusion of significant underlying abnormality, but yield is exceedingly low in absence of cutaneous stigmata or neurological deficits

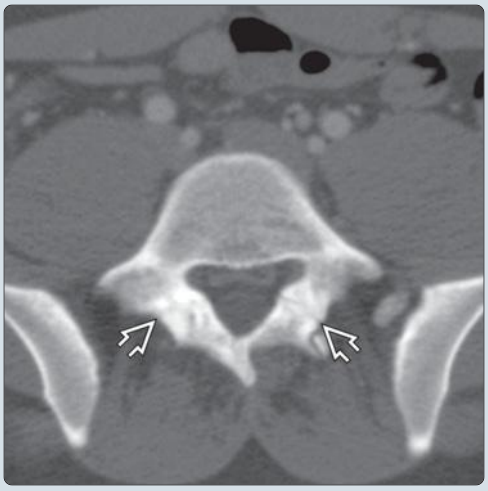
(Left) *Anteroposterior radiograph of the lumbosacral spine (imaging performed for low back pain) demonstrates incidental incomplete fusion of the S1 lamina ➡.* **(Right)** *Axial T1WI MR reveals typical incomplete fusion of the posterior elements ➡ in the midline. This finding was incidentally observed during MR imaging for low back pain.*





(Left) *Coronal bone CT demonstrates oblique asymmetric rotation of the L5 lamina ➡ with overlap, a configuration thought to produce rotational stresses leading to premature facet degenerative changes.* **(Right)** *Axial bone CT at the lumbosacral junction in a patient with oblique asymmetric rotation and incomplete fusion of the posterior elements additionally demonstrates nondisplaced L5 pars defects ➡, possibly related to rotational stress on the pars interarticularis.*





TERMINOLOGY

Synonyms

- "Spina bifida occulta" outdated term

Definitions

- Spinous process/lamina unfused in midline without underlying neural or dural abnormality

IMAGING

General Features

- Best diagnostic clue
  - Incomplete lumbosacral posterior element fusion
- Location
  - Lumbosacral junction (L5 > S1) > > cervical (C1 > C7 > T1), thoracic
- Size
  - Defect usually small
- Morphology
  - Posterior elements do not fuse in midline; margins are rounded, well corticated, and may overlap

Radiographic Findings

- Radiography
  - Unfused spinous process/lamina approximate at midline

CT Findings

- Bone CT
  - Unfused spinous process/lamina

MR Findings

- T1WI
  - Normal dural sac, conus position, and filum thickness
  - No abnormal neural, dural, or lipomatous tissue within posterior osseous defect
- T2WI
  - Same as T1WI

Ultrasonographic Findings

- Grayscale ultrasound
  - Posterior element defect; normal conus level and periodic motion

Imaging Recommendations

- Protocol advice
  - Ultrasound for infants with low-risk sacral dimples
  - Multiplanar MR for tethered cord, neurological abnormalities + cutaneous stigmata

DIFFERENTIAL DIAGNOSIS

Closed Spinal Dysraphism (CSD)

- Lipoma, tethered cord, lipomyelomeningocele, dorsal meningocele, dorsal dermal sinus

Surgical Laminectomy Defect

- Surgical history present; look for incision scar, denervation of paraspinal muscles, laminectomy defect

Normal Progression of Ossification

- Normal L5 laminae may remain unfused until 5-6 years of age

PATHOLOGY

General Features

- Etiology
  - Unknown; probably not linked to neurulation process aberrations

Gross Pathologic & Surgical Features

- Nonfused spinous processes and lamina overlie normal dural sac

Microscopic Features

- Histologically normal corticated bone in lamina

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic if no cutaneous stigmata
  - Occasionally, skin dimple on back
    - Dimples within gluteal cleft need no further evaluation
    - Dimples above gluteal cleft have ↑ incidence of associated abnormalities; merit further evaluation with MR
- Other signs/symptoms
  - None
- Clinical profile
  - Patient presents with low back or leg pain; finding is incidentally noted during imaging

Demographics

- Age
  - All ages
- Gender
  - F > M
- Epidemiology
  - Up to 30% of USA population

Natural History & Prognosis

- Usually incidental finding of no clinical significance

Treatment

- Conservative

DIAGNOSTIC CHECKLIST

Consider

- Incomplete posterior fusion is incidental finding that is very rarely of neurological significance

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Neurenteric Cyst

KEY FACTS

TERMINOLOGY

- Synonyms: Spinal enterogenous cyst, spinal enteric cyst, spinal dorsal enteric cyst

IMAGING

- Intraspinal cyst ± vertebral abnormalities (persistent canal of Kovalevsky, vertebral anomalies)
- Thoracic (42%) > cervical (32%) > > lumbar spine, intracranial/basilar cisterns (rare)

TOP DIFFERENTIAL DIAGNOSES

- Arachnoid cyst
- (Epi)dermoid cyst
- Anterior thoracic meningocele

PATHOLOGY

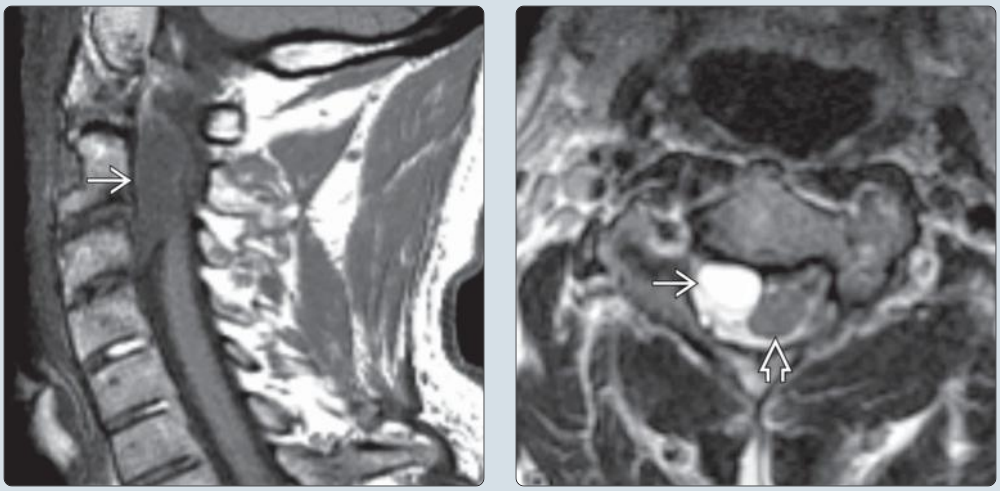
- Subgroup of split notochord syndrome spectrum
  - Sporadic or syndromic (Klippel-Feil, VACTERL, OEIS syndromes)
  - Association with vertebral anomalies, diastematomyelia, lipoma, dermal sinus tract, and tethered spinal cord

CLINICAL ISSUES

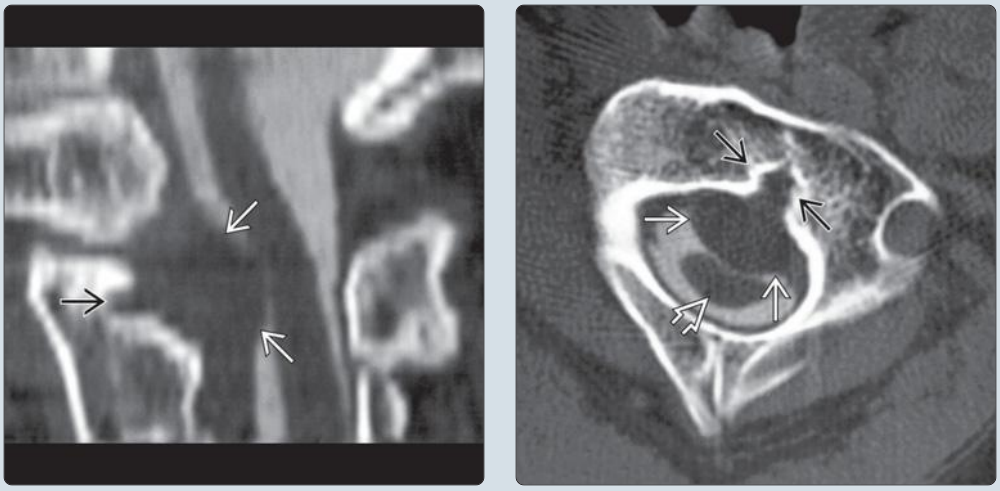
- Most common symptoms: Back/radicular pain, paraparesis/paresthesias, gait disturbance, meningitis
  - Children usually present with cutaneous stigmata, spinal dysraphism symptoms
  - Adults present primarily with pain, myelopathy
- Some asymptomatic, but most show progressive neurological deterioration

DIAGNOSTIC CHECKLIST

- Imaging appearance reflects cyst composition
- Look for associated mediastinal or abdominal cysts, connecting fistulae, or vertebral anomalies



(Left) *Sagittal T1WI MR demonstrates a large ventral extradural cystic mass (neurenteric cyst) producing spinal cord compression. CVJ segmentation anomalies are better demonstrated on other slices (not shown). (Courtesy M. Brandt-Zawadzki, MD.)* **(Right)** *Axial T2WI MR confirms a ventral, extradural neurenteric cyst that displaces the spinal cord and dura to the left, resulting in mild spinal cord compression. (Courtesy M. Brandt-Zawadzki, MD.)*



(Left) *Sagittal bone CT (Klippel-Feil syndrome) after myelography shows a sagittal vertebral cleft in conjunction with a ventral extradural neurenteric cyst. The cyst produces mild spinal cord displacement.* **(Right)** *Axial bone CT (Klippel-Feil syndrome) following myelography shows a small, well-corticated blind vertebral cleft that communicates with a ventral extradural neurenteric cyst. The cyst produces mild compression of the ventral spinal cord.*

TERMINOLOGY

Synonyms

- Spinal enterogenous cyst, spinal enteric cyst, spinal dorsal enteric cyst

Definitions

- Intraspinal cyst lined by enteric mucosa

IMAGING

General Features

- Best diagnostic clue
  - Intraspinal cyst + vertebral abnormalities (persistent canal of Kovalevsky, segmentation and fusion anomalies)
- Location
  - Thoracic (42%) > cervical (32%) > > lumbar spine, intracranial/basilar cisterns (rare)
- Size
  - Range: Small → large
- Morphology
  - Intraspinal or dumbbell-shaped abdominal or mediastinal enteric/spinal cyst; ventral > dorsal, extramedullary (80-85%) > intramedullary (10-15%), midline > paramedian

Radiographic Findings

- Radiography
  - Enlarged spinal canal, widening of interpedicular distance
  - ± vertebral segmentation/fusion anomalies, midline circular vertebral defect (canal of Kovalevsky)

CT Findings

- CECT
  - Hypodense intraspinal cyst with minimal to no enhancement
- Bone CT
  - Focal osseous canal enlargement, widening of interpedicular distance
  - Vertebral anomalies (≤ 50%, usually pediatric patient); vertebral clefts, butterfly vertebra, segmentation anomalies

MR Findings

- T1WI
  - Well-circumscribed, fluid intensity lesion ± vertebral anomalies
    - Iso- → hyperintense (to CSF) depending on protein/mucin content
- T2WI
  - Well-circumscribed, fluid intensity cystic lesion
    - Hypo- → isointense (to CSF) depending on protein/mucin content
  - ± cord myeloschisis, focal cord atrophy
- T1WI C+
  - Mild to no rim enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic intraspinal cyst

Nonvascular Interventions

- Myelography
  - Focal spinal canal enlargement, contrast filling defect
    - Invaginating cyst mimics intramedullary lesion

Nuclear Medicine Findings

- Tc-99m cyst uptake (gastric mucosa) confirms diagnosis

Imaging Recommendations

- Best imaging tool
  - Multiplanar T1WI, T2WI to assess for vertebral anomalies, cord compression, cyst relationship to adjacent structures
  - Bone CT/3D CT to characterize osseous anomalies, surgical planning

DIFFERENTIAL DIAGNOSIS

Arachnoid Cyst

- CSF density/intensity on all pulse sequences
- Dorsal > ventral spinal canal
- Vertebral anomalies uncommon

Epidermoid Cysts

- Uncommon at CVJ
- Most common at conus/cauda equina level
- ± sinus tract, cord tethering, skin dimple

Anterior Thoracic Meningocele

- Anterior dumbbell shape, contiguous with dural sac
- Associated with ventral osseous defect

PATHOLOGY

General Features

- Etiology
  - Putative derivation from abnormal connection between primitive endoderm and ectoderm during 3rd embryonic week
    - Notochord normally separates dorsal ectoderm (skin and spinal cord) and ventral endoderm (foregut)
    - Failure of separation → "split" notochord or notochord deviated to side of adhesion
    - Incomplete separation of notochord layer from endoderm (primitive foregut) layer hinders development of mesoderm; small piece of primitive gut becomes trapped in developing spinal canal
    - Embryo growth, adhesion may lengthen or partially obliterate; enteric and spinal structures connected through persistent canal of Kovalevsky in severe cases
- Associated abnormalities
  - Vertebral anomalies (≤ 50%), anterior or posterior dysraphism, small dysplastic vertebral bodies, vertebral fusion, butterfly or hemivertebra
    - Implies earlier developmental "error" than cases without vertebral anomalies
    - More common if mediastinal or abdominal cysts
  - Diastematomyelia (31%), lipoma (31%), dermal sinus tract, or tethered spinal cord (23%)
  - Klippel-Feil syndrome
  - Cutaneous stigmata
  - Alimentary duplication or fistulae, VACTERL syndrome
- Enteric cysts not limited to spinal column; may be found within brain, mediastinum, abdomen, pelvis, subcutaneous tissues

- Neurenteric cyst is subgroup of split notochord syndrome spectrum
  - Dorsal enteric diverticulum: Diverticulum from dorsal mesenteric border of bowel
  - Dorsal enteric enterogenous cyst: Prevertebral, intraspinal, postvertebral, mediastinal, or mesenteric location
  - Dorsal enteric sinus: Blind-ending tract with opening on dorsal skin surface
  - Dorsal enteric fistula (most severe): Connects intestinal cavity with dorsal skin surface, traversing through soft tissues and spine

Staging, Grading, & Classification

- World Health Organization (WHO) classification: Other malformative tumors and tumor-like lesions
- 3 types based on histology (Wilkins and Odum)
  - Type A: Single layer or pseudostratified epithelium resembling respiratory (17%) or gastrointestinal (50%) epithelium, or both (33%)
  - Type B: Type A + mucous or serous glands, smooth muscle connective tissue, lymphoid tissue or nervous tissue
  - Type C: Type A + ependymal or other glial elements

Gross Pathologic & Surgical Features

- Single smooth unilocular (rarely multilocular) cyst containing clear or proteinaceous fluid (milky, cream-colored, yellowish, xanthochromic)
  - Demonstrable cyst connection with spinal cord, vertebrae, or both
  - Dorsal spinal-enteric tract traverses cartilage-lined canal of Kovalevsky through small dysplastic vertebra

Microscopic Features

- Thin-walled cyst lined by simple, pseudostratified, stratified cuboidal, or columnar epithelium ± ciliated epithelium, goblet cells
- Ultrastructurally similar to brain Rathke cleft cyst, colloid cyst

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck/radicular pain
  - Progressive quadriparesis/paresthesias
  - Gait disturbance
  - Meningitis
    - Mollaret → rupture into CSF
    - Bacterial → seeding by enteric organisms
- Other signs/symptoms
  - Chronic fever and myelopathy 2° to secretion of TNF-α (usually infants)
  - Congenital mirror movements of hands (very rare, seen in conjunction with diastematomyelia)
- Clinical profile
  - Children present more commonly with cutaneous stigmata, spinal dysraphism symptoms
  - Adults present primarily with pain, myelopathy

Demographics

- Age
  - 2nd → 4th decades (range: 8 days → 72 years)
    - Cysts with associated malformations present earlier than isolated cysts
- Gender
  - M:F = 2-3:1-2
- Epidemiology
  - Rare: 0.3-0.5% of spinal "tumors"

Natural History & Prognosis

- Some asymptomatic → diagnosis at autopsy
- Most show progressive neurological deterioration
- Significant symptomatic improvement following resection in many patients

Treatment

- Primary treatment goal is complete surgical excision
  - Drainage, partial resection if complete excision not possible
  - Subtotal excision → recurrence

DIAGNOSTIC CHECKLIST

Consider

- Degree of cord compression, size of cyst, and severity of associated anomalies determine disease course and prognosis

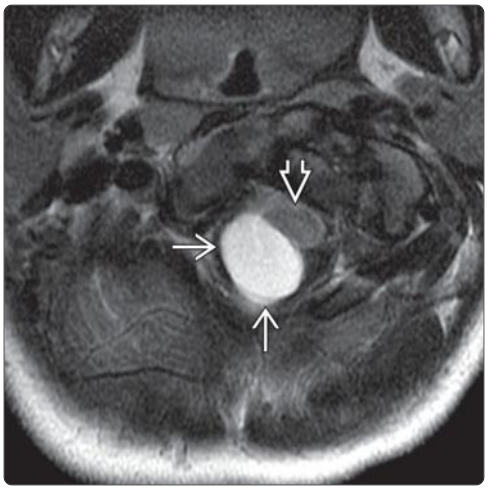
Image Interpretation Pearls

- Imaging appearance reflects cyst composition
- Look for associated mediastinal or abdominal cysts, connecting fistulae, or vertebral anomalies

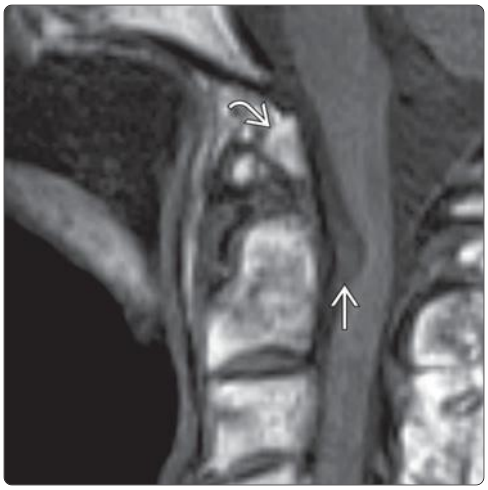
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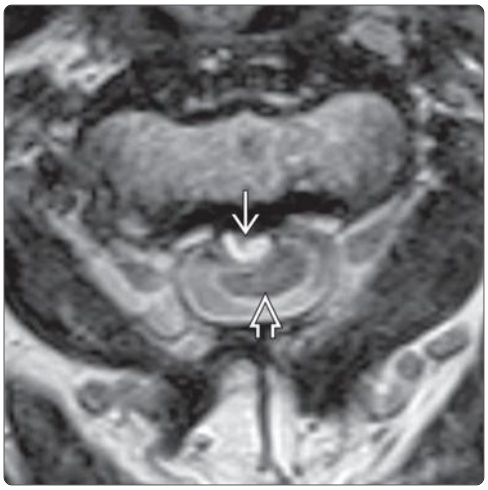
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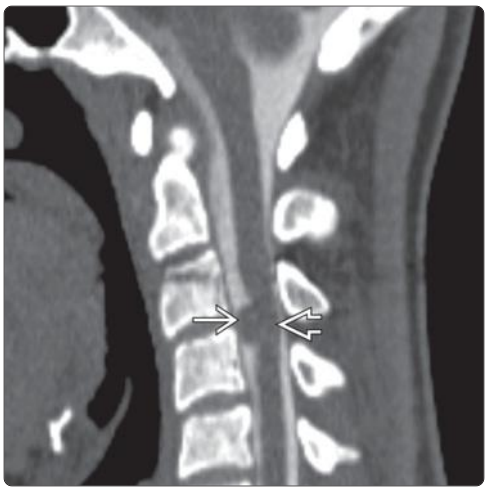


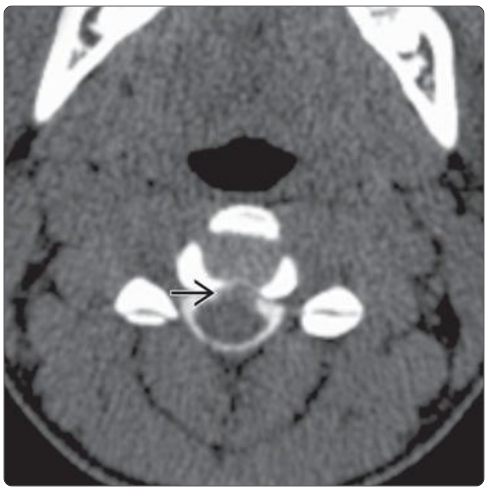
(Left) *Sagittal T1WI C+ FS MR demonstrates a nonenhancing posterior extradural cystic mass ➡ at the craniovertebral junction resulting in adjacent spinal cord ➡ compression. Note that there are congenital vertebral anomalies at the craniovertebral junction as well.* **(Right)** *Axial T2WI confirms posterior and right placement of the neurenteric cyst ➡ within the spinal canal, producing ventral and leftward displacement of the cervical spinal cord ➡.*





(Left) *Sagittal T1WI MR (Klippel-Feil syndrome) shows a small ventral intradural extramedullary neurenteric cyst ➡ that compresses the ventral spinal cord. There is an associated os odontoideum ➡.* **(Right)** *Axial T2WI MR in a patient with Klippel-Feil syndrome reveals a small hyperintense intradural, extramedullary neurenteric cyst ➡ that produces distortion and posterior displacement of the cervical spinal cord ➡.*





(Left) *Sagittal NECT following myelographic contrast administration reveals a ventral intradural extramedullary mass ➡ at the C3-C4 level, producing mild spinal cord mass effect ➡. (Courtesy M. Brandt-Zawadzki, MD.)* **(Right)** *Axial NECT after intrathecal myelographic contrast administration demonstrates a ventral intradural mass ⇨ representing a neurenteric cyst. No vertebral segmentation anomalies are present. (Courtesy M. Brandt-Zawadzki, MD.)*

Os Odontoideum

KEY FACTS

TERMINOLOGY

- Synonym: Os
- Corticated oval or round ossicle (os) at odontoid process tip

IMAGING

- Well-defined round or oval ossicle at dens tip with smooth, uniform cortex
- ± hypertrophy of anterior C1 (atlas) arch
- ± spinal cord contusion, myelomalacia (if craniocervical instability)

TOP DIFFERENTIAL DIAGNOSES

- Ossiculum terminale
- Odontoid C2 fracture (type II)

PATHOLOGY

- 2 types based on os position
  - Orthotopic: Os in anatomic position
  - Dystopic: Os in any position other than orthotopic (considered less stable)
- Incompetence of transverse atlantal ligament (TAL) → C1/2 instability

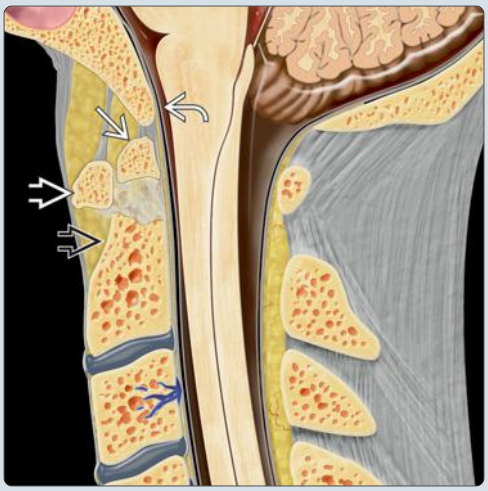
CLINICAL ISSUES

- Commonly asymptomatic
  - Os incidentally detected on imaging obtained for other reasons (e.g., trauma)
- Symptomatic patients
  - Local mechanical neck pain, torticollis, headache, or other neurological symptoms
- Prognosis variable; depends in large part on CVJ ligamentous stability

DIAGNOSTIC CHECKLIST

- Evaluate dynamic CVJ stability when os odontoideum is identified

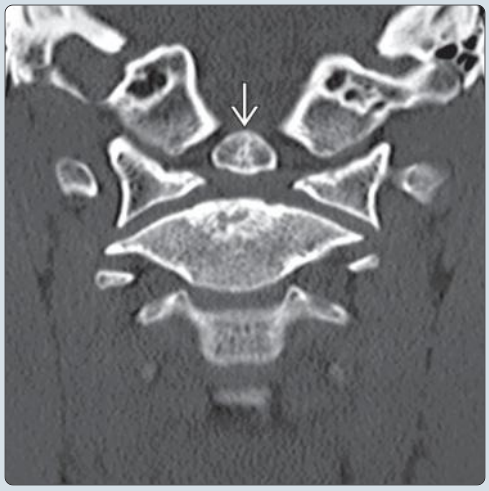
(Left) *Sagittal graphic of the cervical spine demonstrates an orthotopic os odontoideum ➡. There is mild separation of the os away from the odontoid process ⇨ toward the clivus ➡. Note the maintenance of normal os distance to the mildly enlarged anterior C1 ring ➡.* **(Right)** *Sagittal T2WI MR of the craniovertebral junction depicts an orthotopic os odontoideum ➡, with close os approximation to the anterior C1 arch ➡. The cervical spinal cord is normal.*





(Left) *Sagittal bone CT reveals enlargement of the C1 anterior ring ➡ in conjunction with mildly dystopic os odontoideum ➡ that is discontiguous with the dysplastic, rounded odontoid process ➡. Note maintenance of os alignment with C1 and the clivus.* **(Right)** *Coronal bone CT confirms discontinuity of the rounded os odontoideum ➡ with the C2 vertebral body. The os is mildly displaced to the left. Alignment of the occipital condyles and C1 lateral masses is normal.*





TERMINOLOGY

Synonyms

- Os

Definitions

- Corticated oval or round ossicle (os) at odontoid process tip
- 2 types described based on position of os
  - Orthotopic: Os in normal position of odontoid tip
  - Dystopic: Os in any other location
    - Most commonly os located near foramen magnum, may fuse with clivus (os avis) or anterior C1 ring

IMAGING

General Features

- Best diagnostic clue
  - Ossicle at dysplastic odontoid tip + anterior C1 arch enlargement
- Location
  - CVJ, near dens tip
- Size
  - Variable; typically smaller than normal dens
- Morphology
  - Round or oval ossicle with smooth, uniform cortex
    - Separate from C2 base and odontoid process
    - Ossicle border does not directly match with axis body
  - ± hypertrophy of anterior atlas arch

Radiographic Findings

- Radiography
  - Well-defined ossicle at dens tip + anterior C1 arch enlargement
  - ± atlantoaxial instability on flexion/extension

CT Findings

- Bone CT
  - Well-defined ossicle at dens tip + anterior C1 arch enlargement
  - Os may be fused to clivus or C1 anterior arch

MR Findings

- T1WI
  - Well-defined ossicle at dens tip + anterior C1 arch enlargement
  - Normal marrow signal in os, anterior C1 ring
- T2WI
  - Same as T1WI
  - ± spinal cord contusion, myelomalacia if craniocervical instability

Imaging Recommendations

- Best imaging tool
  - Multiplanar bone CT for osseous anatomy
  - Multiplanar MR for spinal cord, soft tissue evaluation
- Protocol advice
  - Axial bone CT with sagittal, coronal reformats
  - Sagittal, axial, coronal T2WI most helpful
    - May be performed in flexion and extension to assess stability, effect on spinal cord

DIFFERENTIAL DIAGNOSIS

Ossiculum Terminale

- Variant ossification failure of terminal ossicle → incidental "notch" in dens tip, adjacent round ossicle
- Ossicle of ossiculum terminale usually much smaller than os odontoideum; lies at level of C1 ring above transverse atlantal ligament
- Rarely associated with clinical instability

Odontoid C2 Fracture (Type II)

- Dens fracture nonunion
- Normal dens size and shape
- Typically narrow gap between axis base, dens
- Anterior C1 arch normal size

PATHOLOGY

General Features

- Etiology
  - Controversial and actively debated; several theories proposed
    - "Congenital" theory
      - Aberrant embryogenesis of occipital sclerotome, proatlas, and C1 resegmented sclerotome
      - Excessive cartilaginous dens movement at time of ossification → congenital failure of dens fusion to axis body
      - Explains detection of os odontoideum in very young
    - "Traumatic" theory
      - Sequela of prior fracture of odontoid synchondrosis before closure (age 5-6 years) → gradual os odontoideum development
      - With growth, alar ligaments carry dens fragment away from axis base
      - Cranial portion of dens fragment continues to receive blood supply from apical vessels
      - Avascular caudad portion resorbs → typical rounded ossicle
      - Explains os odontoideum detection in patients with previously normal cervical radiographs
- Associated abnormalities
  - ↑ frequency reported in Morquio syndrome, multiple epiphyseal dysplasia, Klippel-Feil syndrome, Down syndrome
- Pertinent embryology of cervicocranium (occiput-C3 sclerotomes) for os odontoideum
  - 4th occipital sclerotome (proatlas) divides into ventral rostral and dorsal caudal portions during normal embryogenesis
    - Ventral proatlas component → anterior foramen magnum, occipital condyles
      - Part of ventral proatlas also detaches to incorporate with pleuricentrum of atlas vertebra → dens
      - Core of proatlas centrum → apical dens ligament
    - Atlas, transverse atlantal ligament (TAL) derived from proatlas
      - Odontoid process represents atlas centrum following separation from C1 ring during 6th-7th gestational weeks, fusion to axis body

- Osseous elements develop through enchondral ossification
  - Dens base ossifies just prior to birth
  - Axis body fuses with odontoid process ~ age 4 years
  - Dens tip fuses to odontoid process ~ age 12 years

Staging, Grading, & Classification

- Orthotopic: Os in anatomic position
- Dystopic: Os in any position other than orthotopic
  - Most commonly os located near foramen magnum, may fuse with clivus or anterior C1 ring
  - Potentially less stable than orthotopic os
- Plain radiograph metrics
  - Anterior atlantodens interval (AADI)
    - Used clinically to measure C1/2 instability
    - Caveat: Os fragment often moves with atlas, even with gross C1/2 instability
  - Posterior atlantodens interval (PADI)
    - Distance between posterior border of dens and anterior border of posterior C1 ring
    - Indirectly reflects space available for cord (SAC)
      - Soft tissue proliferation (synovial cyst, pannus) ↓ SAC but occult on plain radiographs
    - Surrogate for MR or CT, where actual space for spinal cord can be measured
      - MR measurement of cord compression may be more predictive of symptomatology
    - PADI < 13 mm associated with progressive neurologic decline
  - PADI instability index
    - Change in SAC between flexion and extension
  - Direct measurement of C1 motion of C1 on C2 body
    - Space between line superiorly from anterior C2 body and line inferiorly from posterior border of anterior C1 arch
    - > 3 mm separation pathologic

Gross Pathologic & Surgical Features

- Os odontoideum is potentially unstable
  - TAL normally straps around basal dens segment
  - May not normally attach in os odontoideum
    - Incompetence of TAL → C1/2 instability

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Commonly asymptomatic
    - Os incidentally detected on imaging obtained for other reasons (e.g., trauma)
  - Symptomatic patients report local mechanical neck pain, torticollis, headache
    - Because asymptomatic os is common, may be difficult to determine relation of os odontoideum to presenting symptoms
- Other signs/symptoms
  - Transitory or persistent quadriparesis following trauma
  - Progressive myelopathy
  - Weakness
  - Ataxia
  - Lower cranial neuropathies
  - Vertebral artery compression → cervical cord, brainstem ischemia
  - Sudden death (rare)

Demographics

- Age
  - Variable
- Epidemiology
  - Exact prevalence, incidence unknown

Natural History & Prognosis

- Variable; depends in large part on CVJ ligamentous stability
- AADI and PADI prognostic indices for stability

Treatment

- Nonoperative management recommended for most patients incidentally diagnosed with os odontoideum
  - Asymptomatic patients without radiographic evidence for significant instability → observation
  - Symptomatic patients with mechanical symptoms → medical management
    - Cervical traction, physical therapy, firm collar, activity limitation, anti-inflammatory medications
- Surgical stabilization for spinal instability, neurologic decline, or intractable pain

DIAGNOSTIC CHECKLIST

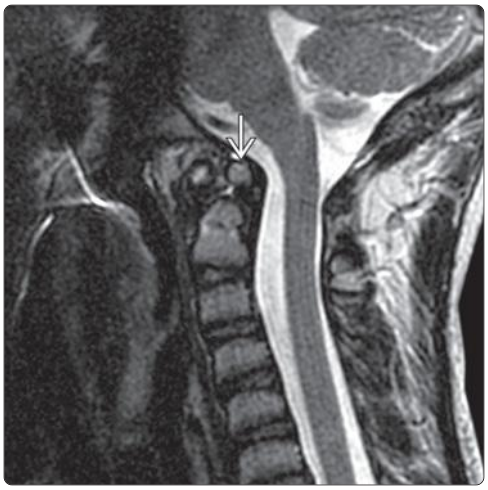
Consider

- Evaluate dynamic CVJ stability with flexion/extension radiographs when os odontoideum identified
- Dystopic os considered more likely to be unstable than orthotopic os

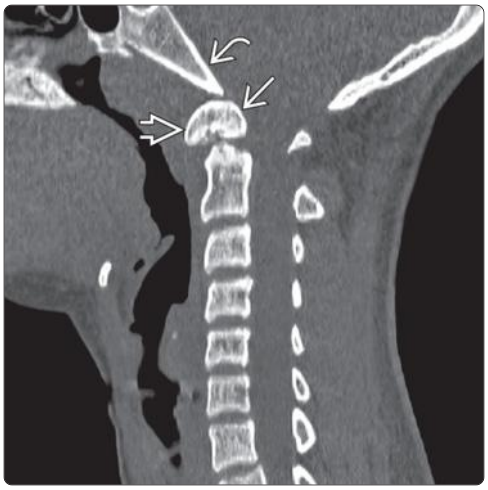
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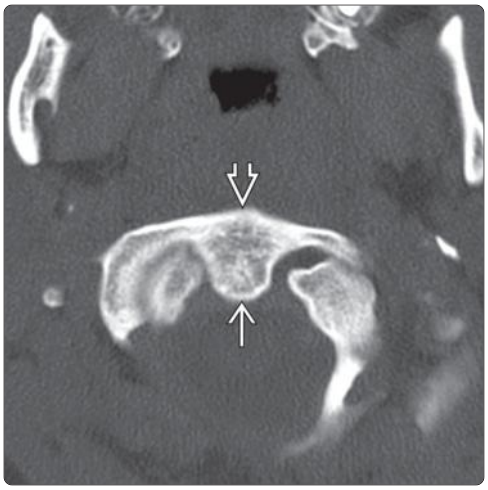
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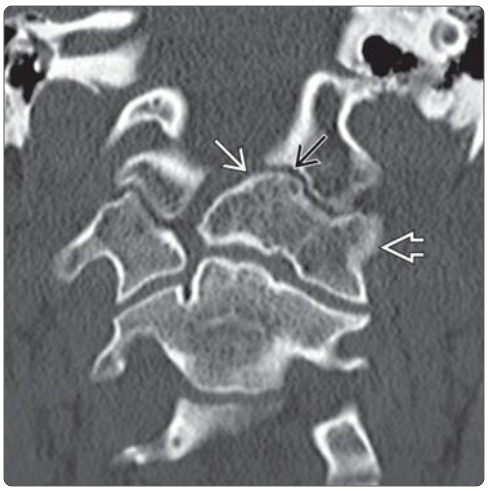
(Left) *Sagittal T2WI MR obtained in flexion shows anterior positioning of the dystopic os relative to C2. The normal distance between the anterior C1 ring and the os is preserved, but there was a change in os position relative to C2 (atlantoaxial instability) compared to neutral and extension sagittal images.* **(Right)** *Sagittal T2WI MR obtained in extension shows normal os position. Flexion sagittal T2WI MR showed anterior os translation, indicating atlantoaxial instability.*





(Left) *Sagittal bone CT of the cervical spine shows congenital fusion of the anterior C1 ring and a large os odontoideum. Normal clivus relationship to the os is maintained. The subaxial cervical spine was otherwise normal, and no additional vertebral segmentation anomalies were detected.* **(Right)** *Axial bone CT confirms solid congenital fusion of an orthotopic os to the midline anterior C1 ring. The posterior C1 ring was complete (not shown).*





(Left) *Sagittal bone CT demonstrates fusion of a large dystopic os odontoideum to the anterior C1 arch. The clivus tip is minimally remodeled (flattened). The short, rounded odontoid process is dysplastic.* **(Right)** *Coronal bone CT depicts eccentric fusion of a dystopic os odontoideum to the left C1 anterior arch and lateral mass. Note also the abnormal pseudoarticulation between the os and the left occipital condyle.*

Lateral Meningocele

KEY FACTS

TERMINOLOGY

- Lateral meningocele (LM), lateral thoracic meningocele (LTM), lateral lumbar meningocele (LLM)

IMAGING

- CSF-filled dural/arachnoidal sac protrudes laterally through neural foramen
- Pedicular erosion, foraminal enlargement, dural dysplasia
- Bilateral meningoceles: Consider NF1, Marfan syndrome

TOP DIFFERENTIAL DIAGNOSES

- Nerve sheath tumor
- Radicular (meningeal) cyst
- Chronic inflammatory demyelinating polyneuropathy
- Foregut duplication cyst

PATHOLOGY

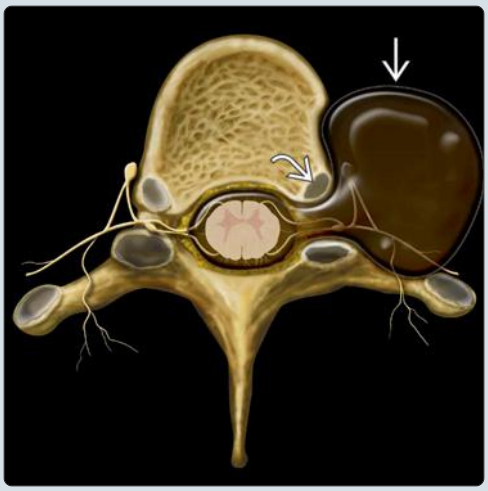
- Etiology secondary to primary meningeal dysplasia
- Scalloping of pedicles, laminae, and vertebral bodies adjacent to meningocele
- Enlarged central spinal canal, neural foramina

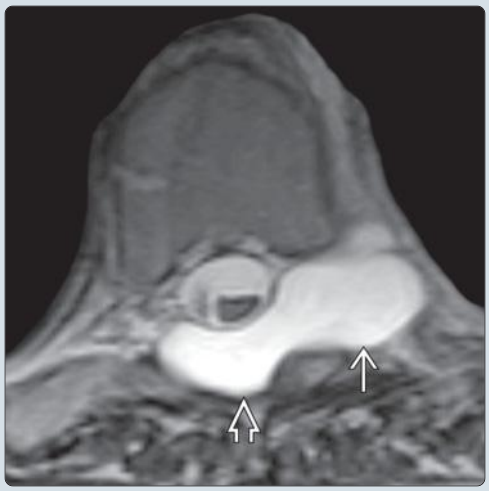
CLINICAL ISSUES

- Asymptomatic (most common) or nonspecific motor or sensory symptoms referable to cord/nerve root compression
- Most remain asymptomatic unless very large or scoliosis causes symptoms
- Very large meningoceles may → respiratory embarrassment (meningocele fills hemithorax)

DIAGNOSTIC CHECKLIST

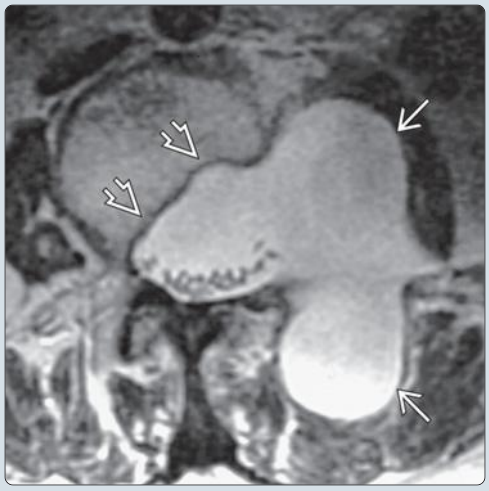
- Lateral meningocele prompts search for history/stigmata of NF1 or connective tissue disorder





(Left) *Axial graphic depicts a large left lateral thoracic meningocele ➡ producing pedicular erosion ➡, transverse process remodeling, and widening of the neural foramen.* **(Right)** *Axial T2WI MR (Marfan syndrome) demonstrates a large left lateral thoracic meningocele ➡ extending through an enlarged, remodeled neural foramen. The contiguous intraspinal extradural component of the meningocele ➡ displaces the thecal sac anteriorly.*





(Left) *Axial NECT following myelography (neurofibromatosis type 1) reveals a large left lateral lumbar meningocele ➡ and extensive vertebral scalloping from dural dysplasia ➡, resulting in marked left pedicular erosion and enlargement of the ipsilateral neural foramen.* **(Right)** *Axial T2WI MR (neurofibromatosis type 1) shows a large left lateral lumbar meningocele ➡ in conjunction with extensive dural dysplasia, vertebral remodeling ➡, and marked left pedicular erosion.*

TERMINOLOGY

Synonyms

- Lateral meningocele (LM), lateral thoracic meningocele (LTM), lateral lumbar meningocele (LLM)

Definitions

- Meningeal dysplasia → CSF-filled dural/arachnoidal sac protrudes laterally through neural foramen

IMAGING

General Features

- Best diagnostic clue
  - CSF signal/density meningeal protrusion through neural foramen into adjacent intercostal/extrapleural space
- Location
  - Thoracic > lumbar spine
  - R > L; 10% bilateral
    - Bilateral meningoceles usually associated with NF1 but may be seen in Marfan syndrome
- Size
  - Typical size 2-3 cm; range from tiny to huge
- Morphology
  - CSF signal/density "cyst" adjacent to spine
    - Contiguous with neural foramen
    - ± sharply angled scoliosis at meningocele level

Radiographic Findings

- Radiography
  - Pedicular erosion ± neural foraminal enlargement
    - Often accompanied by scalloping of posterior vertebral bodies (dural ectasia)
    - ± sharply angled kyphosis/scoliosis (meningocele is near apex of deformity on convex side)

CT Findings

- CECT
  - CSF density mass extends through enlarged neural foramen
  - No enhancement; useful to distinguish from nerve sheath tumor, nerve inflammation (CIDP)
- CTA
  - ± aortic aneurysm, dissection in context of systemic connective tissue disorder
- Bone CT
  - Wide neural foramen; ± pedicular thinning, posterior vertebral scalloping (usually)
  - Reformatted images may show focal scoliosis (coronal plane) and dural ectasia (sagittal plane)

MR Findings

- T1WI
  - CSF signal intensity (hypointense) mass in contiguity with thecal sac
  - Pedicular thinning, neural foraminal widening ± posterior vertebral scalloping
- T2WI
  - CSF signal intensity (hyperintense) mass in contiguity with thecal sac; rarely see neural elements within meningocele
- T1WI C+
  - No enhancement; distinguishes from nerve sheath tumor or inflammation (CIDP)

Ultrasonographic Findings

- Grayscale ultrasound
  - Posterior mediastinal or lumbar hypoechoic paraspinal cystic mass contiguous with expanded spinal canal
    - Displaces and compresses adjacent spinal cord
  - Ultrasound is primary diagnostic tool in utero, screening newborn infants
- Pulsed Doppler
  - No vascular flow pattern
- Color Doppler
  - Avascular hypoechoic mass

Nonvascular Interventions

- Myelography
  - Intrathecal contrast fills cyst from dural sac through enlarged neural foramen
    - Confirms contiguity with thecal sac
    - Delayed imaging may be required
    - Consider placing patient with meningocele(s) dependently to improve contrast filling

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Consider sonography for newborn screening; follow-up with MR to clarify positive ultrasound study
  - MR imaging for diagnosis, preoperative planning
  - Bone CT to evaluate pedicles, vertebral bodies (particularly if surgery is contemplated)

DIFFERENTIAL DIAGNOSIS

Nerve Sheath Tumor

- Less hyperintense than CSF on T2WI, higher signal intensity than CSF on T1WI
- Contrast enhancement implies tumor
  - Caveats: Some schwannomas may appear cystic, and some neurofibromas minimally enhance

Radicular (Meningeal) Cyst

- CSF signal intensity/density cyst within neural foramen
  - Cyst separate from dural sac, unlike meningocele
- Nerve root definable as discrete structure within or adjacent to cyst

Chronic Inflammatory Demyelinating Polyneuropathy

- Solid fusiform nerve root enlargement, enhancement
- Clinical and laboratory findings characteristic

Foregut Duplication Cyst

- Bronchogenic most common; may contain gastrointestinal (GI) mucosa
  - Seldom contiguous with neural foramen
- Proximity to spinal canal ± vertebral anomalies = neurenteric cyst

PATHOLOGY

General Features

- Etiology
  - Meningocele 2° to primary meningeal dysplasia
    - Meningeal weakness permits dural sac to focally stretch in response to repetitive CSF pulsation → enlarged neural foramina
    - Secondary osseous remodeling permits further herniation
  - Posterior vertebral scalloping with dural dysplasia → same etiology
- Genetics
  - Strong association with neurofibromatosis type 1 (NF1) (85%)
    - Most common posterior mediastinal mass
  - Less common with Ehlers-Danlos, Marfan syndromes
- Associated abnormalities
  - Occasionally isolated finding
  - ± coexistent lumbar and thoracic lateral meningoceles
  - ± findings specific to hereditary disorder
    - NF1: Dural ectasia, nerve sheath tumors, CNS neoplasms, pheochromocytomas, interstitial pulmonary fibrosis, skin, and subcutaneous neurofibromas
    - Marfan syndrome: Dural ectasia, vascular dissection/aneurysm, lens dislocation, joint laxity
- Dural sac diverticulum, pedicular erosion, neural foraminal widening, and posterior vertebral scalloping

Gross Pathologic & Surgical Features

- Scalloping of pedicles, laminae, and vertebral bodies adjacent to meningocele
- Enlarged central spinal canal, neural foramina
- Cord position variable; usually displaced away from meningocele
- Scoliosis convex toward meningocele

Microscopic Features

- Dura/arachnoid lined outpouching of thecal sac

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic (most common)
  - Nonspecific motor or sensory symptoms referable to cord/nerve root compression
- Other signs/symptoms
  - Respiratory embarrassment (neonates); very large meningocele fills thoracic cavity
- Clinical profile
  - Asymptomatic patient (incidental discovery)
  - ± scoliosis evaluation
  - NF1: Cutaneous café au lait spots, cutaneous and subcutaneous neurofibromas, ± kyphoscoliosis
  - Connective tissue disorder: Frequently tall, joint hypermobility, lens dislocation, ± normal intelligence, ± scoliosis

Demographics

- Age
  - Most commonly present during 4th-5th decades of life
- Gender
  - M = F
- Epidemiology
  - Uncommon in NF1 and inherited connective tissue disorders but substantially more common than occurrence as isolated lesion

Natural History & Prognosis

- Most remain asymptomatic unless very large or scoliosis causes symptoms
- Most static in size; occasionally grow slowly
- May disappear after hydrocephalus shunting
- Excellent prognosis after surgical resection

Treatment

- Options, risks, complications
  - Surgical ligation of dural sac neck, resection of meningocele
  - Correction/stabilization of scoliosis

DIAGNOSTIC CHECKLIST

Consider

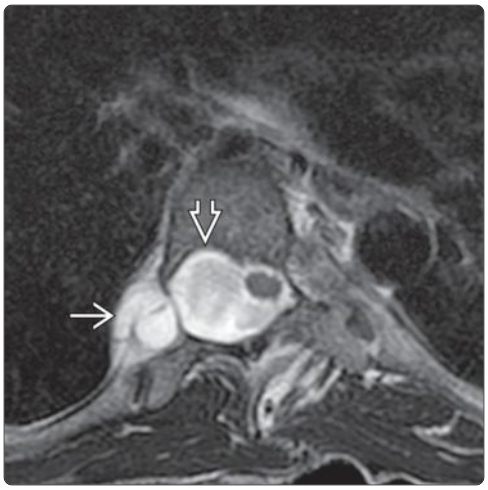
- Lateral meningocele prompts search for history/stigmata of NF1 or connective tissue disorder

Image Interpretation Pearls

- MR shows nonenhancing CSF signal intensity/density mass extending through enlarged neural foramen

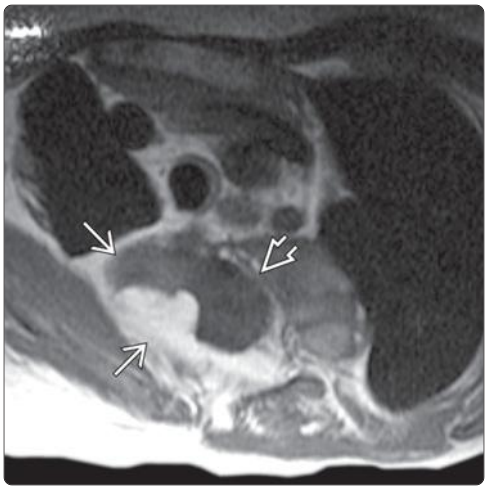
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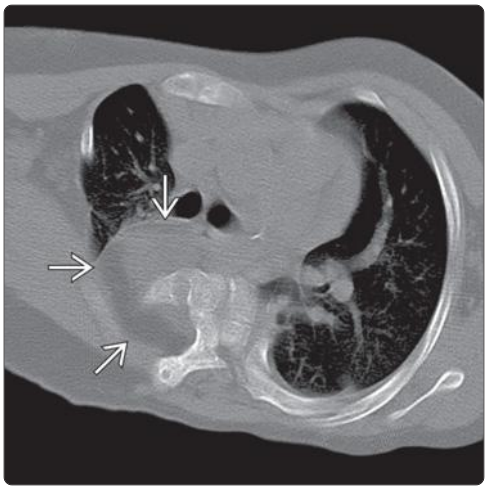
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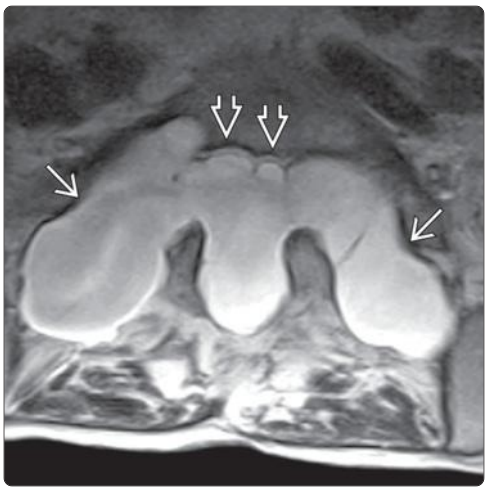
(Left) *Axial T2WI MR (neurofibromatosis type 1) demonstrates characteristic osseous remodeling of dural ectasia involving the midthoracic thecal sac* ➪ *with right lateral meningocele* ➪ *projecting into right the paraspinal soft tissue through an enlarged neural foramen.* **(Right)** *Axial T1WI C+ MR (neurofibromatosis type 1) reveals extension of a lateral thoracic meningocele* ➪ *through an enlarged neural foramen. No abnormal tissue enhancement to indicate neurofibroma is demonstrated.*





(Left) *Axial T1WI MR demonstrates lateral extension of a complex fat-containing lateral thoracic meningocele* ➪ *into the right paraspinal tissues. Note scalloping of the posterior vertebral body* ➪ *and absence of the right pedicle.* **(Right)** *Axial bone CT shows lateral extension of a complex fat-containing lateral thoracic meningocele* ➪ *around the vertebral body, with enlargement of the neural foramen and absence of the right thoracic pedicle at the meningocele level.*





(Left) *Sagittal T2WI MR (Marfan syndrome) demonstrates bilateral large lateral lumbar meningoceles* ➪*. There is also characteristic posterior vertebral scalloping* ➪ *related to severe dural dysplasia.* **(Right)** *Axial T2WI MR (Marfan syndrome) demonstrates extensive large bilateral lumbar lateral meningoceles* ➪ *extending into the adjacent paraspinal tissues. Note also prominent vertebral scalloping* ➪ *reflecting concurrent severe dural dysplasia.*

Dorsal Spinal Meningocele

KEY FACTS

TERMINOLOGY

- Synonyms: Simple meningocele, simple spinal meningocele, posterior meningocele

IMAGING

- Skin-covered dorsal dural sac protruding thorough posterior osseous defect
  - Dorsal herniation of dura, arachnoid, and CSF into spinal subcutaneous tissue
  - Occasional herniation of filum or nerve roots into defect
- Conus medullaris low or normal in position

TOP DIFFERENTIAL DIAGNOSES

- Lipomyelomeningocele
- Terminal myelocystocele
- Myelomeningocele

PATHOLOGY

- ± hydromyelia, tethered cord, diastematomyelia
- Meningocele always skin covered; skin may be dysplastic, ulcerated

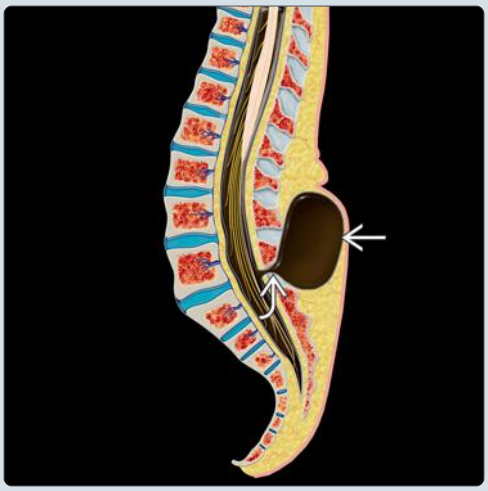
CLINICAL ISSUES

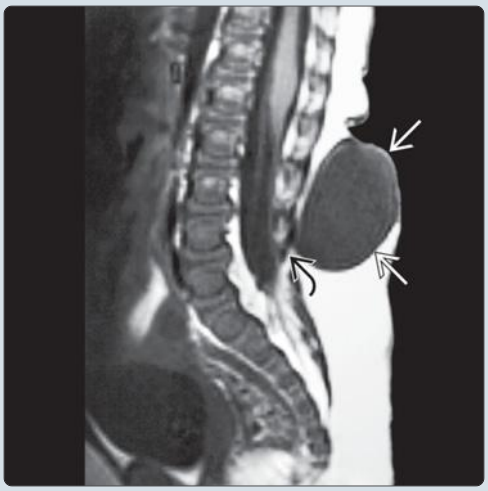
- Palpable skin-covered mass or incidental discovery during imaging for other indications
  - Patients usually neurologically normal
- Additional presenting signs/symptoms include back pain, meningitis, spinal headache
- Cervical, thoracic meningoceles more likely to be symptomatic than lumbar meningocele

DIAGNOSTIC CHECKLIST

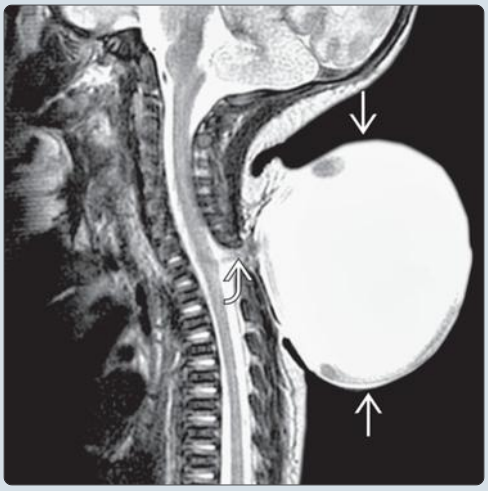
- Imaging goals are lesion detection, determination of simple or complex morphology, and exclusion of other spinal anomalies
  - Important to determine whether neural elements reside within sac prior to surgery

(Left) *Sagittal graphic of the lumbosacral spine shows a typical dorsal spinal meningocele sac protruding through the dysraphic posterior elements, with a thin isthmus connecting to the thecal sac.* **(Right)** *Sagittal T1WI MR reveals a borderline low-lying conus at L2 and a large, skin-covered CSF-signal mass in the subcutaneous tissues. The mass (dorsal meningocele) communicates with the thecal sac via a very thin fluid signal pedicle traversing the posterior elements.*





(Left) *Sagittal T2WI MR demonstrates a large cervicothoracic dorsal meningocele extending into the subcutaneous tissues through a thin dysraphic isthmus. There is a hypointense CSF flow jet through the isthmus into the sac.* **(Right)** *Axial T2WI MR shows a large cervicothoracic dorsal meningocele extending into the subcutaneous tissues through a thin dysraphic isthmus. Conforming to strict definition, the sac contains no neural elements, and the cervical spinal cord is normal.*





TERMINOLOGY

Synonyms

- Simple meningocele, simple spinal meningocele, posterior meningocele

Definitions

- Dorsal herniation of dura, arachnoid, and CSF into spinal subcutaneous tissue

IMAGING

General Features

- Best diagnostic clue
  - Skin-covered dorsal dural sac protruding thorough posterior osseous defect
- Location
  - Anywhere along dorsal spinal canal; lumbosacral junction, sacrum > > cervical, thoracic
- Size
  - Small and localized to large, encompassing multiple spinal levels
- Morphology
  - Sessile or pedunculated CSF intensity/density sac with spinal dysraphism
  - May (complex) or may not (simple) contain neural tissue

Radiographic Findings

- Radiography
  - Dorsal dysraphism; defect usually confined to 1-2 vertebrae
  - Widening of spinal canal, increased interpedicular distance

CT Findings

- NECT
  - Hypodense CSF dural sac
  - Overlying skin intact; may be ulcerated
- Bone CT
  - Dysraphism ± spinal canal widening, increased interpedicular distance
    - Mild cases may show only absent spinous process or localized spina bifida
    - More severe cases show multisegmental spina bifida, spinal canal enlargement

MR Findings

- T1WI
  - Skin-covered hypointense dural sac
  - Conus medullaris low or normal in position
  - Occasional herniation of filum or nerve roots into defect
- T2WI
  - Hyperintense dural sac
  - Low or normal conus medullaris position
  - ± filum terminale or nerve roots within defect

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic CSF-filled sac protrudes through posterior defect ± entrapped nervous tissue
  - Demonstrates conus termination level and periodic motion
  - Primary utility for in utero or neonatal screening
    - Obstetrical ultrasound may detect large meningocele in utero → altered delivery planning
    - Less useful in older children/adults due to spinal column ossification → posterior acoustic shadowing

Nonvascular Interventions

- Myelography
  - Dorsal dysraphism, widening of spinal canal, increased interpedicular distance
  - Dural sac herniates through dysraphic posterior elements
    - Dural sac communicates with subarachnoid space
      - May change size with position, Valsalva maneuver
    - ± neural elements, filum terminale within defect
  - Myelography mostly replaced by MR
    - Primary utility is for patients with MR contraindications and to answer discrepancies between MR and clinical findings

Imaging Recommendations

- Best imaging tool
  - MR imaging best shows dural sac characteristics, associated vertebral or spinal cord anomalies
- Protocol advice
  - Sagittal and axial T1WI and T2WI
    - Sagittal images useful to evaluate cord
    - Axial T1WI most helpful to evaluate size of dysraphic defect and to exclude lipoma
    - Axial T2WI best for detecting nervous tissue within sac

DIFFERENTIAL DIAGNOSIS

Lipomyelomeningocele

- Distinguish by presence of neural tissue and fat within defect in addition to cyst

Terminal Myelocystocele

- Hydromyelic, low-lying cord protrudes through meningocele

Myelomeningocele

- Open spinal dysraphism; clinical appearance is diagnostic

PATHOLOGY

General Features

- Etiology
  - Unknown
    - No universally accepted unifying theory explains pathogenesis
- Associated abnormalities
  - ± hydromyelia, tethered cord, diastematomyelia
  - Chiari 2 malformation; much less common than with myelomeningocele
- Classically, simple meningoceles is considered an isolated abnormality with normal conus termination level
  - However; some authors report low conus termination and other spinal anomalies with meningocele and question whether meningocele is ever truly an isolated lesion
- Meningocele is always skin covered
  - Skin may be dysplastic or ulcerated

- Cervical lesions considered within cervical myelocystocele spectrum

Gross Pathologic & Surgical Features

- Nearly always contain aberrant nerve roots, ganglion cells, &/or glial nodules at surgery, gross pathology
- Herniated meningocele sac contains both dura and arachnoid
- Overlying tissue intact unless secondary skin ulceration

Microscopic Features

- Meningocele lined by arachnoid and thin-walled blood vessels
  - Arachnoidal adhesions may obstruct sac neck
- Overlying skin shows atrophic epidermis, lacks rete pegs and normal skin appendages

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Palpable skin-covered mass
    - May be incidental discovery during imaging for other indications
- Other signs/symptoms
  - Back pain
  - Meningitis (ruptured or leaking meningocele)
  - Headache, other signs/symptoms of alternating high and low intracranial pressure
- Clinical profile
  - Patients are usually neurologically normal and present for imaging of palpable back mass
  - Cervical, thoracic meningoceles more likely to be symptomatic than lumbar meningocele

Demographics

- Age
  - In utero to adult life
- Gender
  - M = F
- Epidemiology
  - 1 per 10,000 live births

Natural History & Prognosis

- Variable; depends on cyst size and contents, status of overlying skin, presence or absence of meningitis
- Skin covering permits more "elective" surgical correction
  - Most newborns require an operation before release from nursery to home

Treatment

- Asymptomatic patients may warrant conservative observation
- Symptomatic patients require surgical resection, repair of dural defect

DIAGNOSTIC CHECKLIST

Consider

- Ultrasound in infants to determine cord level, presence or absence of neural elements in meningocele
- MR is best modality to definitively characterize meningocele, evaluate for other spinal abnormalities
  - Use steady state technique (CISS/FIESTA) to best visualize nerve roots/adhesions/masses

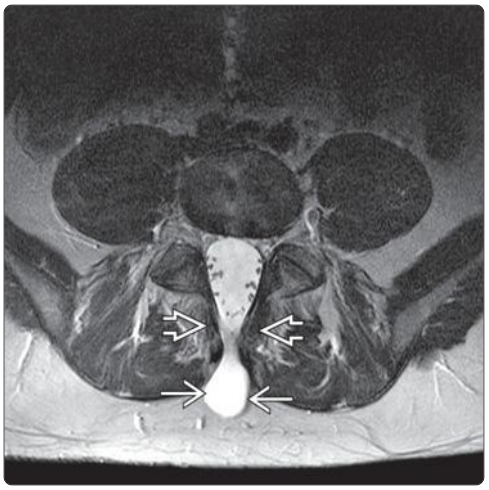
Image Interpretation Pearls

- Imaging goals are lesion detection, determination of simple or complex morphology, and exclusion of other spinal anomalies
  - Important to determine whether neural elements reside within sac prior to surgery
  - Meningocele may be most obvious abnormality that masks other less obvious but clinically more important lesions

SELECTED REFERENCES

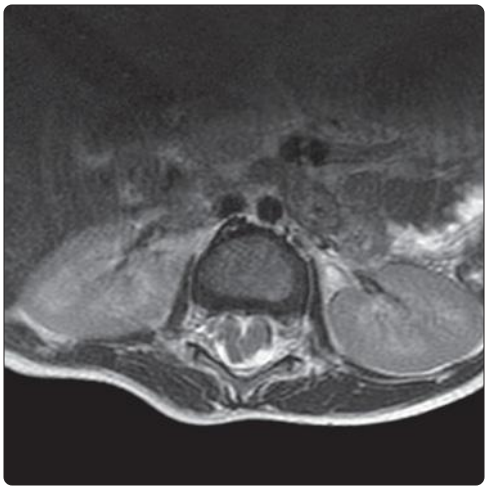
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(Left) *Sagittal T1WI MR depicts a small dorsal spinal meningocele extending through an L4-L5 level dysraphic defect into the posterior subcutaneous fat. The distal osseous spinal canal is mildly expanded, and focal fat is present within the thickened filum terminale, which is tethered at the neck of the herniation sac.* **(Right)** *Axial T2WI MR reveals a single-level posterior spinal element dysraphic defect, with protrusion of the meningocele sac into the subcutaneous fat.*





(Left) *Sagittal T2WI MR shows a skin-covered cystic back mass at the lower lumbar level. The spinal cord is low lying, with the conus terminating at the L5 level. An obliquely oriented hypointense fibrous band traverses the spinal cord and enters the cystic back mass.* **(Right)** *Axial T2WI MR reveals spinal cord distortion as the fibrous band traverses the cord parenchyma above the meningocele level, representing a short segmental diastematomyelia (surgically proven).*





(Left) *Sagittal T1WI MR shows abnormal low-lying conus position at L4. There is also mild distortion and dorsal positioning of the conus, with inclusion of dorsal nerve roots in the meningocele sac that protrudes through a posterior dysraphic defect into the subcutaneous soft tissues.* **(Right)** *Axial T2WI MR shows variant inclusion of neural elements extending into a skin-covered dorsal meningocele sac. The posterior elements are widely dysraphic.*

Dural Dysplasia

KEY FACTS

TERMINOLOGY

- Synonym: Dural ectasia

IMAGING

- Smooth remodeling of posterior vertebral body, expansion of osseous spinal canal, ± kyphoscoliosis
  - Pedicular attenuation, widened interpediculate distance, erosion of anterior and posterior elements
- Patulous, expansile dural sac enlargement
- Easiest to appreciate on sagittal images

TOP DIFFERENTIAL DIAGNOSES

- Congenital vertebral dysplasia
- Spinal tumor or syrinx
- Cauda equina syndrome of ankylosing spondylitis (CES-AS)

PATHOLOGY

- Etiologies include NF1, connective tissue disorders (Marfan syndrome, Ehlers-Danlos syndrome, Loey-Dietz syndrome, homocystinuria), idiopathic
- Association with lateral thoracic or lumbar meningocele, anterior sacral meningocele, kyphoscoliosis, joint hypermobility, lens abnormalities, aneurysm, arterial dissection, and peripheral or central neoplasms
- Dura in ectatic areas is extremely thin, fragile

CLINICAL ISSUES

- Most commonly presents with back pain ± radiculopathy
- Other complaints include headache, incontinence, pelvic symptoms
- May present at any age depending on severity, underlying etiology

DIAGNOSTIC CHECKLIST

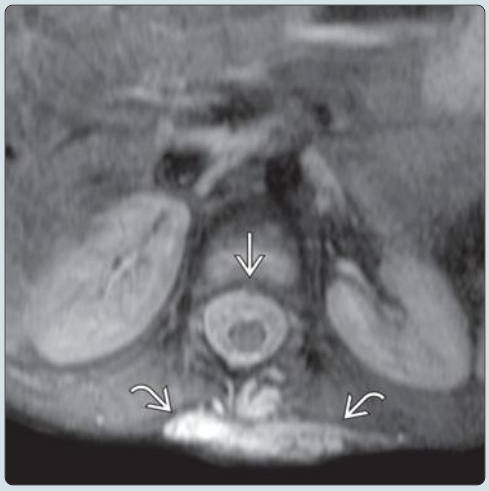
- Recognition of specific imaging clues and integration of available clinical data permits a more specific diagnosis
- Important to determine underlying disorder for treatment planning, genetic counseling, and determining prognosis





(Left) *Sagittal graphic of the lumbosacral spine demonstrates scalloping of the posterior vertebral bodies and central spinal canal enlargement. Note that the dural sac generally fills the concavity produced in the posterior vertebral body. Also conspicuous (inset) are bilateral lumbar lateral meningoceles.* **(Right)** *Sagittal bone CT (neurofibromatosis type 1) reveals characteristic posterior vertebral remodeling and scalloping ➡ with resultant widening of the central spinal canal.*





(Left) *Sagittal T2WI MR (neurofibromatosis type 1) depicts characteristic posterior vertebral body scalloping and patulous dural sac from L2 through S2. No intradural or foraminal neurofibromas are present.* **(Right)** *Axial STIR MR (neurofibromatosis type 1) demonstrates posterior vertebral body remodeling ➡ related to dural dysplasia. Note also the typical appearance of a hyperintense dorsal lumbar subcutaneous infiltrative plexiform neurofibroma ➡, the clinical indication for imaging.*

TERMINOLOGY

Synonyms

- Dural ectasia

Definitions

- Patulous dural sac with posterior vertebral scalloping

IMAGING

General Features

- Best diagnostic clue
  - Smooth C-shaped scalloping of posterior vertebral bodies with patulous dural sac
- Location
  - Lumbar > cervical, thoracic
- Size
  - Mild → extensive deformity
- Morphology
  - Expansile dural sac, spinal canal remodeling with posterior vertebral scalloping

Radiographic Findings

- Radiography
  - Smooth remodeling of posterior vertebral body, expansion of osseous spinal canal, ± kyphoscoliosis
  - Osteopenia (homocystinuria)

CT Findings

- CECT
  - Posterior vertebral scalloping → spinal canal enlargement
    - Easiest to appreciate on sagittal images
  - Pedicular attenuation, widened interpediculate distance, erosion of anterior and posterior elements, patulous CSF density dural sac
- CTA
  - ± arterial dissection or aneurysm; implicates Marfan or Ehler-Danlos syndrome as underlying etiology

MR Findings

- T1WI
  - Posterior vertebral scalloping, expansion of osseous spinal canal, patulous dural sac, ± kyphoscoliosis
  - ± pedicular thinning, lateral meningocele(s)
- T2WI
  - Similar findings to T1WI
  - Best evaluates position of neural elements relative to dural ectasia
- MRA
  - ± arterial dissection or aneurysm (Marfan, Ehler-Danlos syndrome)

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic patulous dural sac, widening of spinal canal

Angiographic Findings

- Conventional
  - Not useful for imaging spine deformity; primary utility is detecting associated vascular anomalies
  - Ascending aortic aneurysm ("tulip bulb configuration") suggests Marfan syndrome

Nonvascular Interventions

- Myelography
  - Posterior vertebral scalloping, contrast fills enlarged dural sac, ± lateral meningocele(s)

Imaging Recommendations

- MR shows osseous abnormalities well
- Additionally, MR most useful modality to exclude syrinx or tumor as a cause of canal enlargement before attributing to dural ectasia

DIFFERENTIAL DIAGNOSIS

Congenital Vertebral Dysplasia

- Achondroplasia
- Mucopolysaccharidosis
- Osteogenesis imperfecta (tarda)
- Search for appropriate family history, clinical stigmata

Spinal Tumor or Syrinx

- Astrocytoma
- Ependymoma
- Nerve sheath tumor
- Syrinx
- Characteristic imaging findings lead to correct diagnosis

Cauda Equina Syndrome of Ankylosing Spondylitis (CES-AS)

- Irregular lumbar canal expansion
- Proposed etiology for dural ectasia; proliferative inflammatory synovium → cauda equina symptoms
- Imaging and clinical stigmata of AS typically present

PATHOLOGY

General Features

- Etiology
  - Genetic disorder
    - NF1: Primary mesenchymal disorder
    - Marfan syndrome: Primary connective tissue defect unknown
    - Ehlers-Danlos: > 10 different types of collagen synthesis defects
    - Homocystinuria: Cystathionine β-synthetase deficiency
  - Idiopathic
- Genetics
  - NF1: Autosomal dominant (chromosome 17q12)
  - Marfan: Autosomal dominant
  - Homocystinuria: Autosomal recessive
  - Ehler-Danlos: Autosomal dominant
- Associated abnormalities
  - Lateral thoracic or lumbar meningocele, anterior sacral meningocele
  - Kyphoscoliosis
  - Joint hypermobility, lens abnormalities, aneurysm, arterial dissection (connective tissue disorders)
  - Peripheral and central neoplasms (NF1)
- Genetic predisposition → primary meningeal dysplasia → weakness in meninges → expansion, secondary remodeling of posterior vertebral body and pedicular thinning → further dural sac expansion

Gross Pathologic & Surgical Features

- Enlarged CSF thecal sac, remodeled posterior vertebral bodies
- Dura in ectatic areas is extremely thin, fragile

Microscopic Features

- Dural thinning in ectatic areas

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain → radiculopathy
    - Moderate to severe back pain in > 50% of Marfan patients; presence, degree of dural ectasia associated with back pain
    - High prevalence of dural ectasia (41%) in Marfan patients without back pain; however, mere presence of dural ectasia does not necessarily mean patient is symptomatic
  - Other signs/symptoms
    - Headache
    - Incontinence, pelvic symptoms
- Clinical profile
  - NF1: Plexiform neurofibromas, kyphoscoliosis, optic nerve gliomas and other astrocytomas, café au lait spots, axillary freckling, extremity pseudoarthrosis
  - Marfan syndrome: Tall, joint hypermobility, arachnodactyly, kyphoscoliosis, joint and lens dislocations
  - Homocystinuria: Tall, arachnodactyly, scoliosis, mental retardation, seizures, lens dislocations
  - Ehlers-Danlos: ± tall, thin hyperelastic skin, hypermobile joints, fragile connective tissue

Demographics

- Age
  - May present at any age depending on severity
- Gender
  - M = F
- Ethnicity
  - NF1: All ethnicities; higher in Arab-Israeli subpopulations
  - Marfan: All races, ethnicities
  - Homocystinuria: Northern European descent
  - Ehlers-Danlos: Caucasian, European descent
- Epidemiology
  - NF1: 1/4,000, 50% new mutations; dural ectasia common
  - Marfan: 1/5,000 (USA); dural ectasia present in > 60% of patients
  - Homocystinuria: 1/344,000 worldwide; dural dysplasia less common than Marfan
  - Ehler-Danlos: 1/400,000 (worldwide); dural dysplasia less common than Marfan

Natural History & Prognosis

- Variable; dependent on underlying etiology
- Morbidity and mortality primarily related to vascular pathology
  - Vascular fragility → predisposition to arterial dissection or aneurysm → premature death

Treatment

- Treatment directed toward addressing underlying etiology
- Meningocele repair, scoliosis surgery for more severe cases

DIAGNOSTIC CHECKLIST

Consider

- 3 disease categories produce posterior vertebral scalloping
  - Dural ectasia
  - Increased intraspinal pressure
  - Congenital vertebral dysplasia
- Important to determine underlying disorder for treatment planning, genetic counseling, and determining prognosis

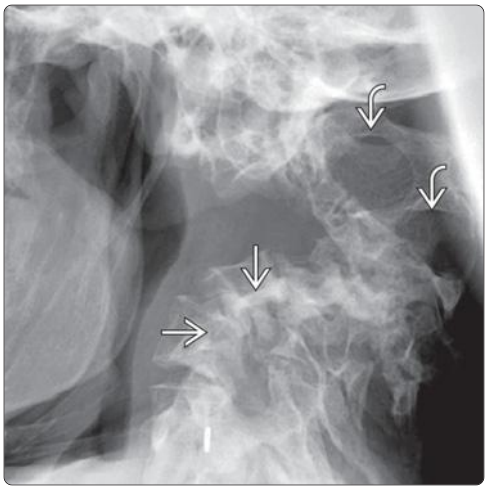
Image Interpretation Pearls

- Recognition of specific imaging clues and integration of available clinical data permits more specific diagnosis
- Look for imaging stigmata of etiological diseases
  - "Tulip bulb" aortic aneurysm → Marfan syndrome
  - Osteoporosis → homocystinuria
  - Pseudoarthrosis, CNS/PNS tumors → NF1

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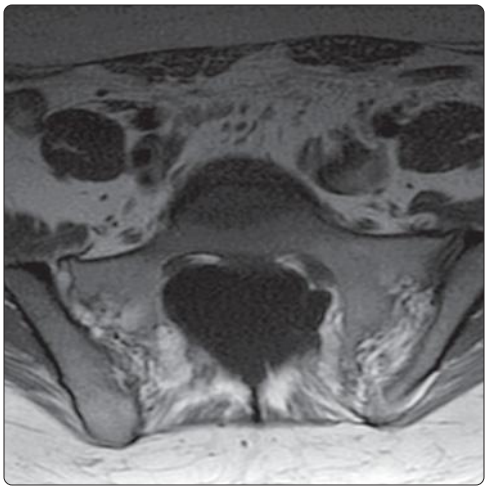
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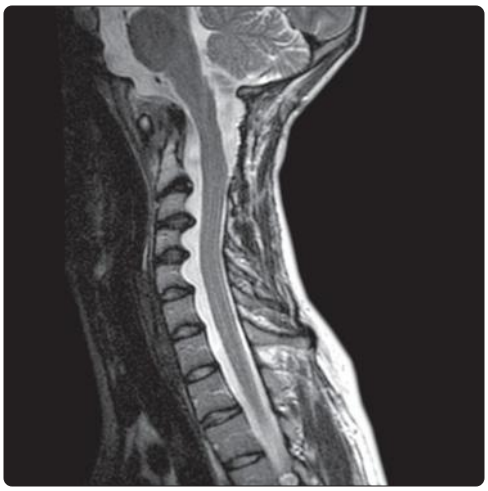


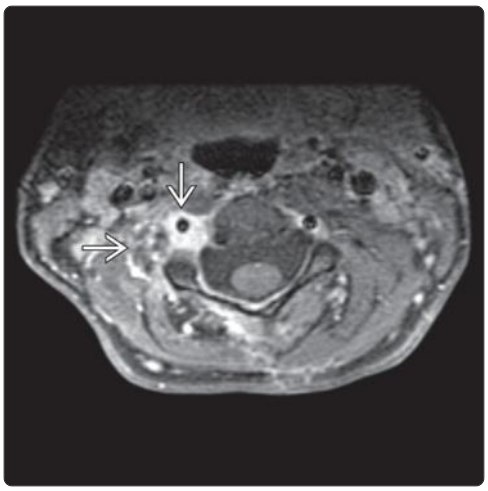
(Left) *Lateral plain radiograph (neurofibromatosis type 1) demonstrates curved posterior vertebral body remodeling* ⇒ *at multiple lumbar vertebral levels characteristic of dural dysplasia. Vascular clips indicate site of prior pheochromocytoma resection.* **(Right)** *Lateral radiograph (neurofibromatosis type 1) shows striking posterior vertebral scalloping* ⇒*, extreme focal kyphosis, and marked neural foraminal enlargement* ⇒*.*





(Left) *Sagittal T2WI MR of the lumbosacral spine (scoliosis, Loey-Dietz connective tissue syndrome) reveals prominent posterior vertebral scalloping and a patulous thecal sac, with marked thinning and posterior scalloping of the sacrum.* **(Right)** *Axial T1WI MR (scoliosis, Loey-Dietz connective tissue syndrome) confirms patulous thecal sac enlargement with characteristic sacral osseous spinal canal remodeling of dural dysplasia.*





(Left) *Sagittal T2WI MR (neurofibromatosis type 1) demonstrates striking posterior vertebral scalloping (C2 through C6) related to dural dysplasia, producing widening of the anteroposterior canal diameter. No intradural or foraminal neurofibromas are identified at the affected levels.* **(Right)** *Axial T1WI C+ FS MR confirms AP canal enlargement as well as an infiltrating, enhancing plexiform neurofibroma* ⇒ *diagnostic of neurofibromatosis type 1.*

KEY FACTS

TERMINOLOGY

- Synonyms: von Recklinghausen disease, peripheral neurofibromatosis
- Mesodermal dysplasia with neurofibromas, spinal deformity, neoplastic and nonneoplastic brain lesions, and cutaneous stigmata

IMAGING

- Kyphoscoliosis ± multiple nerve root tumors, plexiform neurofibroma, dural ectasia/lateral meningocele
- Tumors range from tiny to very large

TOP DIFFERENTIAL DIAGNOSES

- Neurofibromatosis type 2 (central neurofibromatosis)
- Chronic inflammatory demyelinating polyneuropathy (CIDP)
- Congenital hypertrophic polyradiculoneuropathies

PATHOLOGY

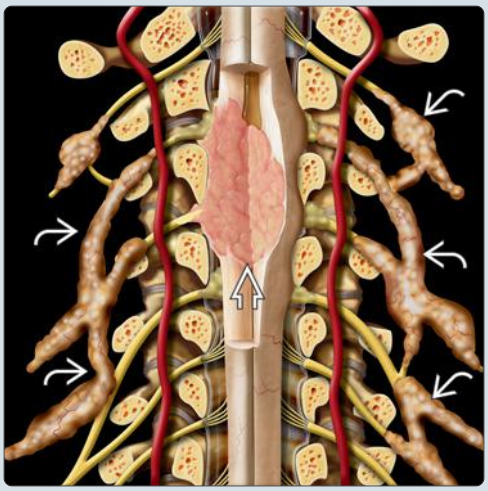
- Autosomal dominant
- Characteristic lesion is plexiform neurofibroma, although 3 types of spinal NF seen in NF1
  - Localized NF (90% of all NF)
  - Diffuse NF
  - Plexiform NF (pathognomonic for NF1)

CLINICAL ISSUES

- Pigmentation anomalies (café au lait, axillary freckling, Lisch nodules)
- Focal or acute angle kyphoscoliosis ± myelopathy
- Palpable spinal or cutaneous masses

DIAGNOSTIC CHECKLIST

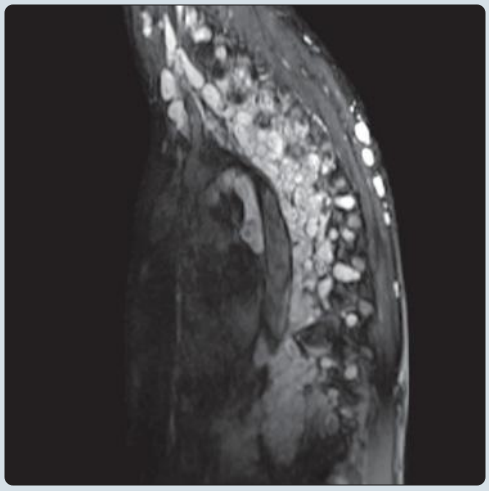
- Multiple nerve sheath tumors, ≥ 1 neurofibroma, bizarre kyphoscoliosis with deformed vertebra → consider NF1
- Absence of visible stigmata does not exclude NF1
- Characteristic PNF imaging appearance best displayed using fat-saturated T2WI or STIR MR imaging





(Left) *Coronal graphic of the cervical spine demonstrates multiple manifestations of neurofibromatosis type 1, including a large intramedullary spinal cord tumor and bilateral brachial plexus plexiform neurofibromas.* **(Right)** *Sagittal T2WI MR demonstrates a large expansile intramedullary primary spinal cord neoplasm expanding into the brainstem. A large neoplastic syrinx is present. Note also cervical lordosis reversal, probably secondary to laminectomy for cord biopsy.*





(Left) *Coronal STIR MR reveals extensive plexiform neurofibromas involving the bilateral paraspinal sympathetic chains as well as involvement of multiple intercostal nerves.* **(Right)** *Sagittal STIR MR obtained off-midline demonstrates innumerable T2 hyperintense plexiform neurofibromas involving the spinal nerves, sympathetic chains, bilateral brachial plexus, as well as multiple cutaneous nerves.*

TERMINOLOGY

Abbreviations

- Neurofibromatosis type 1 (NF1), nerve root neurofibroma (NF), plexiform neurofibroma (PNF), malignant peripheral nerve sheath tumor (MPNST)

Synonyms

- von Recklinghausen disease, peripheral neurofibromatosis

Definitions

- Autosomal dominant mesodermal dysplasia characterized by plexiform and nerve root neurofibromas, spinal deformity, neoplastic and nonneoplastic brain lesions, and cutaneous stigmata

IMAGING

General Features

- Best diagnostic clue
  - Kyphoscoliosis ± multiple nerve root tumors, plexiform neurofibroma, dural ectasia/lateral meningocele
- Location
  - Entire craniospinal axis
- Size
  - Tumors range from tiny to very large
- Morphology
  - Kyphosis/kyphoscoliosis often severe and bizarre
  - Neurogenic tumors localized to nerve roots as well as within plexiform nerve masses, cutaneous lesions

Radiographic Findings

- Radiography
  - Kyphosis/scoliosis, scalloped vertebra, hypoplastic pedicles and posterior elements, "ribbon" ribs

CT Findings

- NECT
  - Hypodense fusiform or focal nerve root enlargement ± heterogeneous spinal cord expansion (glial tumor)
  - Dural ectasia ± CSF density lateral meningocele(s)
- CECT
  - Variable mild/moderate tumor enhancement
- Bone CT
  - Vertebral findings similar to radiography; canal, foraminal widening 2° to dural ectasia ± spinal cord tumor

MR Findings

- T1WI
  - Nerve root neurofibroma: Intramedullary glial cord tumors hypo- → isointense to normal spinal cord, nerve roots, muscle
- T2WI
  - Nerve root neurofibroma: Cord tumors hyperintense to normal spinal cord, nerve roots
    - Nerve root neurofibroma: "Target" sign (hyperintense rim, low/intermediate signal intensity center) suggests neurogenic tumor; PNF > NF > MPNST
- STIR
  - Nerve root neurofibroma: Cord tumors hyperintense to normal nerve root, cord, muscle
- T1WI C+
  - Variable mild to moderate NF, cord tumor enhancement

Nuclear Medicine Findings

- PET
  - FDG standard uptake value (SUV) MPNST > benign tumors

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Radiography to quantitate and follow kyphosis, scoliosis
  - Multiplanar enhanced MR (especially STIR, fat-saturated T2WI, and T1 C+ MR) to evaluate cord, nerve pathology
  - Bone CT to optimally define osseous anatomy for surgical planning

DIFFERENTIAL DIAGNOSIS

Neurofibromatosis Type 2 (Central Neurofibromatosis)

- Multiple intracranial schwannomas and meningiomas, spinal schwannomas and meningiomas
- Spinal deformity uncommon
- Clinical, laboratory, and genetic testing findings distinguish from NF1

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Repeated episodes of demyelination, remyelination → "onion skin" spinal, peripheral nerve enlargement
- Mimics PNF on imaging studies
- No cutaneous stigmata of NF1

Congenital Hypertrophic Polyradiculoneuropathies

- Charcot-Marie-Tooth, Dejerine-Sottas disease
- Nerve root enlargement mimics PNF on imaging studies
- No cutaneous stigmata of NF1

PATHOLOGY

General Features

- Etiology
  - Postulated that NF1 tumor suppression gene "switched off" → tissue proliferation, tumor development
- Genetics
  - Autosomal dominant; chromosome 17q12, penetrance → 100%
    - NF gene product (neurofibromin) is tumor suppressor
  - ~ 50% new mutations (paternal germ line; paternal age 35 years → 2x ↑ in new mutations)
- Associated abnormalities
  - Brain abnormalities: Macrocephaly, focal areas of signal abnormality (FASI), sphenoid wing dysplasia, glial tumors, intellectual handicap, epilepsy, hydrocephalus, aqueductal stenosis
  - ↑ risk of other neuroendocrine tumors (pheochromocytoma, carcinoid tumor), chronic myelogenous leukemia (CML)
  - Congenital bowing, pseudoarthrosis of tibia and forearm, massive extremity overgrowth
  - ↑ fibromuscular dysplasia, intracranial aneurysms, multiple sclerosis

- Plexiform neurofibroma is hallmark of NF1
- Kyphoscoliosis is most common NF1 osseous abnormality; variable severity mild, nonprogressive → severe curvature
  - Dystrophic scoliosis: Short segment, sharply angulated, < 6 spinal segments, tendency → severe deformity
  - Nondystrophic scoliosis: Similar to adolescent idiopathic curvature, usually 8-10 spinal segments, right convex
  - Severe cervical kyphosis highly suggestive of NF1
- Dural ectasia: 1° bone dysplasia, some cases 2° pressure erosion from intraspinal tumors
- "Ribbon" ribs 2° to bone dysplasia ± intercostal NF

Staging, Grading, & Classification

- Consensus Development Conference on Neurofibromatosis (NIH, 1987)
  - 2 or more of the following criteria
    - > 6 café au lait spots measuring ≥ 15 mm in adults or 5 mm in children
    - ≥ 2 neurofibromas of any type or ≥ 1 plexiform neurofibroma
    - Axillary or inguinal freckling
    - Optic glioma
    - 2 or more Lisch nodules (iris hamartomas)
    - Distinctive osseous lesion (sphenoid wing dysplasia, thinning of long bone ± pseudoarthrosis)
    - 1st-degree relative with NF1

Gross Pathologic & Surgical Features

- 3 types of spinal NF recognized in NF1
  - Localized NF (90% of all NF)
    - Most common NF in both NF1, non-NF1 patients
    - Cutaneous and deep nerves, spinal nerve roots
    - NF1: Larger, multiple, more frequently involve large deep nerves (sciatic nerve, brachial plexus)
    - Malignant transformation rare
  - Diffuse NF
    - Infiltrating subcutaneous tumor; rarely affects spinal nerves, majority (90%) unassociated with NF1
  - Plexiform NF (pathognomonic for NF1)
    - Diffuse enlargement of major nerve trunks/branches → bulky rope-like ("bag of worms") nerve expansion with adjacent tissue distortion
    - Commonly large, bilateral, multilevel with predilection for sciatic nerve, brachial plexus
    - ~ 5% risk malignant degeneration → sarcoma

Microscopic Features

- Neoplastic Schwann cells + perineural fibroblasts grow along nerve fascicles
  - Collagen fibers, mucoid/myxoid matrix, tumor, nerve fascicles intermixed
  - S100 positive, mitotic figures rare unless malignant degeneration

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Skeletal deformity common (25-40%)
    - Focal or acute angle kyphoscoliosis ± myelopathy
    - Extremity bowing or overgrowth
  - Palpable spinal or cutaneous mass
  - Pigmentation anomalies (café au lait, axillary freckling, Lisch nodules) ≥ 90% NF1 patients
- Clinical profile
  - Severity of clinical appearance highly variable
  - Classic NF1 triad: Cutaneous lesions, skeletal deformity, and mental deficiency

Demographics

- Age
  - Childhood diagnosis; minimally affected patients may be diagnosed as adults
- Gender
  - M = F
- Ethnicity
  - ↑ frequency in Arab-Israeli populations
- Epidemiology
  - Common (1:4,000)

Natural History & Prognosis

- Kyphosis, scoliosis frequently progressive
- NF growth usually slow; rapid growth associated with pregnancy, puberty, or malignant transformation

Treatment

- Conservative observation; intervention dictated by clinical symptomatology, appearance of neoplasm
- Surgical resection of symptomatic localized NF, spinal cord tumors
- PNF invasive, rarely resectable; observation ± biological or chemotherapeutic (thalidomide, antihistamines, maturation agents, antiangiogenic drugs) intervention
- Spinal fusion reserved for symptomatic or severe kyphoscoliosis

DIAGNOSTIC CHECKLIST

Consider

- Multiple nerve sheath tumors, ≥ 1 neurofibroma, bizarre kyphoscoliosis with deformed vertebra → consider NF1
- Absence of visible stigmata does not exclude NF1

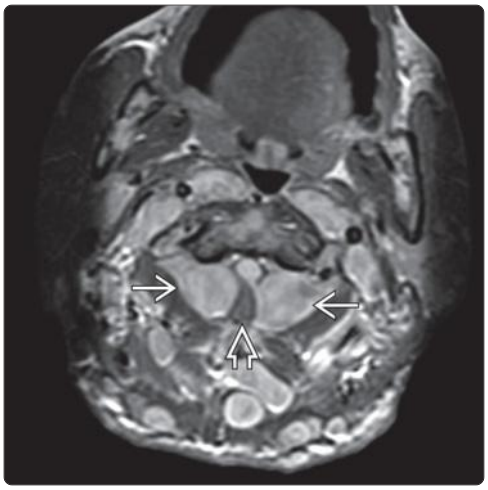
Image Interpretation Pearls

- Characteristic PNF imaging appearance best displayed using fat-saturated T2WI or STIR MR imaging

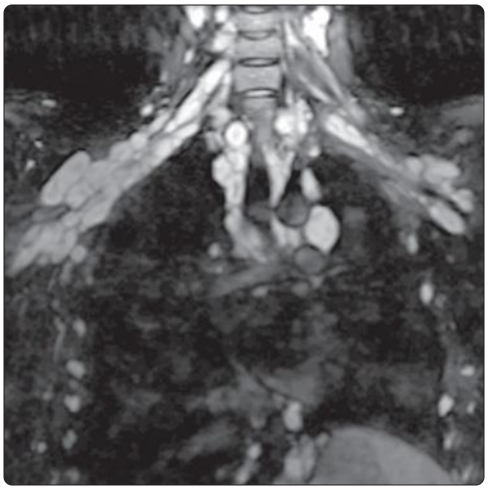
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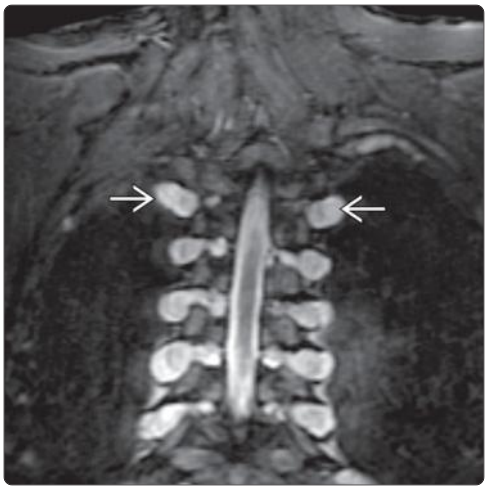
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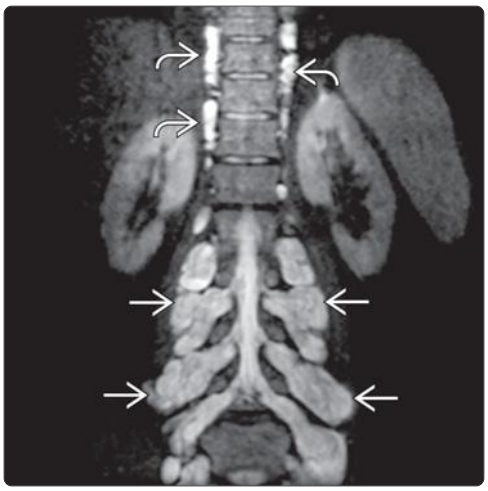


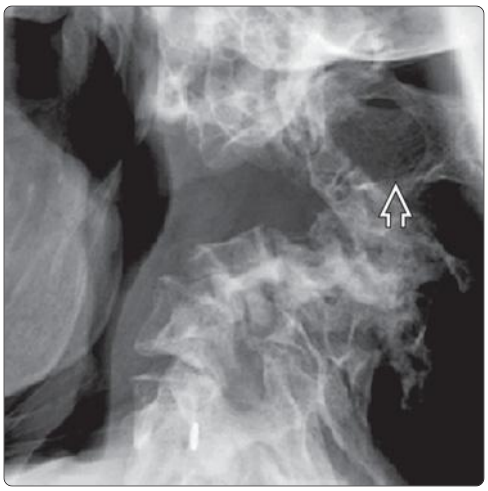
(Left) *Sagittal T1WI MR shows extensive intradural extramedullary neurofibromas ⇨ located ventral to the spinal cord, displacing the cord posteriorly. Numerous subcutaneous soft tissue neurofibromas are also present posteriorly (several indicated with ⇨).* **(Right)** *Axial T1 C+ MR of the upper cervical spine demonstrates marked extent of involvement by innumerable soft tissue and spinal neurofibromas. Note severe spinal cord ⇨ compression produced by bilateral C2 neurofibromas ⇨.*





(Left) *Coronal STIR MR reveals innumerable plexiform neurofibromas involving the bilateral spinal nerves, sympathetic chains, and bilateral brachial plexus, as well as extensive involvement of multiple intercostal nerves.* **(Right)** *Coronal STIR MR depicts bilateral thoracic nerve root neurofibromas ⇨, extending through the neural foramina into the paraspinal tissues. Neurofibromas are moderately hyperintense on STIR MR imaging.*





(Left) *Coronal STIR MR demonstrates multiple bilateral lumbar nerve root neurofibromas ⇨ extending through the neural foramina into the adjacent paraspinal soft tissues. Small plexiform neurofibromas ⇨ of the sympathetic chain are also detected.* **(Right)** *Lateral radiograph of the cervical spine depicts bizarre acute angle cervical kyphosis with marked dural ectasia and neural foraminal enlargement ⇨, highly characteristic of neurofibromatosis type 1.*

Neurofibromatosis Type 2

KEY FACTS

TERMINOLOGY

- Rare autosomal dominant disease from chromosomal 22 defect in which all patients develop CNS tumors
- Mnemonic for NF2 tumors: **M**ultiple **i**nherited **s**chwannomas, **m**eningiomas, and **e**pendymomas (MISME)

TOP DIFFERENTIAL DIAGNOSES

- Metastases
- Hemangioblastomas
- Nonsyndromic schwannoma
- Nonsyndromic meningioma
- Nonsyndromic ependymoma
- Lymphoma

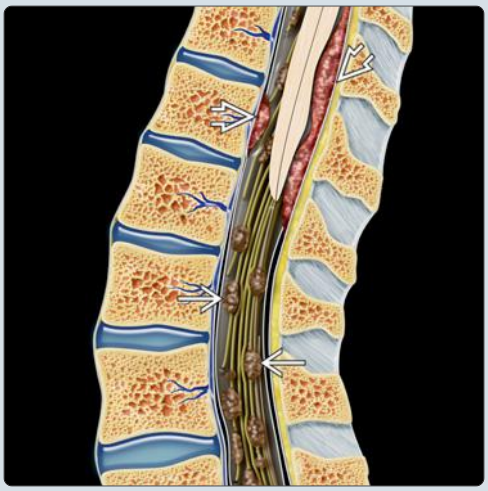
PATHOLOGY

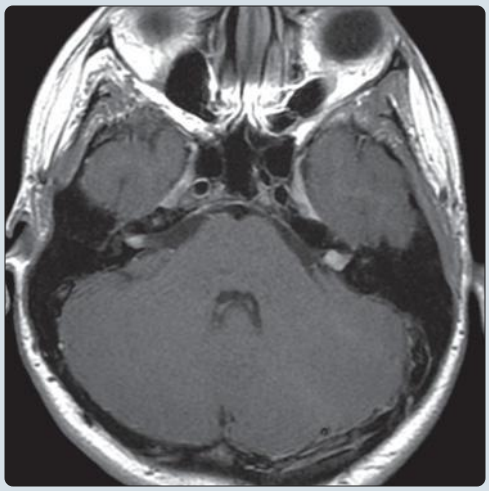
- 22q12 deletion correlates with loss of *NF2* gene product "merlin" (a.k.a. schwannomin)
- Definite diagnosis of NF2
  - Bilateral CN8 (vestibular) schwannomas
  - 1st-degree relative with NF2 and either unilateral early onset vestibular schwannoma (age < 30 years) or any 2: Meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity
- Presumptive diagnosis of NF2
  - Early onset unilateral CN8 schwannomas (age < 30 years) and 1 of the following
  - Meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity
  - Multiple meningiomas (> 2) and unilateral vestibular schwannoma
  - Or 1 of these: Glioma, schwannoma, juvenile posterior subcapsular lenticular opacity

DIAGNOSTIC CHECKLIST

- Screen using MR C+ of brain and entire spine
- Imaging follow-up of patients with spinal tumors should be based on knowledge of tumor location, number, and suspected histologic type

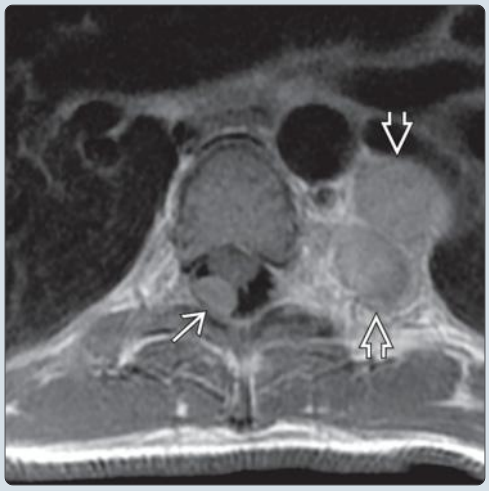
(Left) *Sagittal graphic illustrates multiple rounded schwannomas (brown) along the cauda equina, as well as flat dural-based meningiomas (red) impinging the conus.* **(Right)** *Axial T1WI C+ MR shows bilateral cerebellopontine angle masses due to vestibular schwannomas in this patient with NF2.*





(Left) *Sagittal T1WI C+ MR demonstrates multiple intramedullary enhancing ependymomas. The largest is at the C3-C5 level, with associated inferior cyst, with a smaller 2nd tumor at C7-T1 level.* **(Right)** *Axial T1 C+ MR shows heterogeneous enhancement of paraspinal schwannomas, as well as an intradural extramedullary tumor with a broad dural base consistent with meningioma.*





TERMINOLOGY

Abbreviations

- Neurofibromatosis type 2 (NF2), nonsyndromic (NS)

Synonyms

- Bilateral acoustic neurofibromatosis, central neurofibromatosis (both obsolete)

Definitions

- Autosomal dominant disease from chromosomal 22 defect in which all patients develop CNS tumors
- Mnemonic for NF2 tumors: **M**ultiple **i**nherited **s**chwannomas, **m**eningiomas, and **e**pendymomas (MISME)

IMAGING

General Features

- Best diagnostic clue
  - Multiple spinal tumors of various histologic types

MR Findings

- T1WI
  - Schwannomas
    - Well-delineated, rounded, intermediate signal mass
    - May undergo hemorrhage or cystic degeneration
    - Uncommon intramedullary, centrally located
  - Meningiomas: Mid to low intensity
  - Ependymomas
    - Iso- to slightly hyperintense
    - Centrally located with cord expansion
- T2WI
  - Schwannomas
    - Well-delineated, rounded, hyperintense mass
    - May undergo hemorrhage or cystic degeneration
  - Meningiomas
    - May be heterogeneous
    - Best defines high occurrence of cord compression
  - Ependymomas: Hyperintense; centrally located with cord expansion
    - Ill-defined hyperintense lesion may be low-grade astrocytoma
    - Any intramedullary lesion may have syrinx
- T1WI C+
  - Schwannomas: Enhance intensely; homogeneously when small, heterogeneously when large and cystic
  - Meningiomas: Enhance intensely (often to lesser degree than schwannomas) and homogeneously
  - Ependymomas: Enhancing, centrally located mass

Nonvascular Interventions

- Myelography
  - Intradural filling defects &/or cord expansion
  - May have complete block

Imaging Recommendations

- Best imaging tool
  - MR + gadolinium
- Protocol advice
  - Contrast-enhanced imaging is best method for detecting lesions regardless of size

DIFFERENTIAL DIAGNOSIS

Metastases

- Cord lesions tend to have extensive edema
- Eccentrically placed, rarely within center of cord

Hemangioblastomas

- Often associated with von Hippel-Lindau
- Originate from leptomeninges, associated cyst

Nonsyndromic Schwannoma

- Focal, usually solitary lesion

Nonsyndromic Meningioma

- Isolated dura-based mass

Nonsyndromic Ependymoma

- Usually solitary, with imaging identical to syndromic tumors

Lymphoma

- May coat spinal cord surface

PATHOLOGY

General Features

- Etiology
  - Chromosomal 22 deletion with eventual inactivation of merlin functionality
    - 1st event is 22 chromosomal loss
    - "2nd hit theory": Remaining single *NF2* copy is mutated; vast majority are null mutations
    - Results in truncated, poorly functioning, or nonfunctional merlin protein
- Genetics
  - Inherited autosomal dominant syndrome
  - 22q12 deletion correlates with loss of *NF2* gene product "merlin" (a.k.a. schwannomin)

Staging, Grading, & Classification

- Definite diagnosis of NF2
  - Bilateral CN8 (vestibular) schwannomas
  - 1st-degree relative with NF2 and either unilateral early onset vestibular schwannoma (age < 30 years) or any 2 of the following
    - Meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity
- Presumptive diagnosis of NF2
  - Early onset unilateral CN8 schwannomas (age < 30 years) and 1 of the following
    - Meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity
  - Multiple meningiomas (> 2) and unilateral vestibular schwannoma or 1 of the following
    - Glioma, schwannoma, juvenile posterior subcapsular lenticular opacity
- Germline study findings support genotype-phenotype correlation of tumor grading
  - Nonsense and frameshift mutations
    - Usually associated with severe disease phenotype
    - Higher percentage of these patients have intramedullary tumors
    - Have higher numbers of all spinal tumors; intramedullary and extramedullary

  - Younger at onset of symptoms, diagnosis of NF2, and presentation for imaging
- Missense mutations and large gene deletions are found with mild phenotype
- Splice site mutations; severe or mild phenotype
  - Dependent on intron involved and its affect on protein functionality
- 3 features of intramedullary ependymomas
- Central location within cord parenchyma
- Intense enhancement
- Multiplicity, often too many to count

Gross Pathologic & Surgical Features

- Multiple tumors of different histiologic types

Microscopic Features

- Merlin associates at cell cytoskeleton near plasma membrane and inhibits cell proliferation, adhesion, and migration
- Merlin monoclonal antibody immunohistochemistry exhibits consistent immunostaining of Schwann cells
- NF2 histopathology no different than that of nonsyndromic tumors; exception is multiplicity

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
- Nearly half initially present with hearing loss
- Up to 45% with extramedullary tumors exhibit signs/symptoms of cord compression
  - Varies depending on location
  - Weakness and sensory loss at or below level
  - Spasticity, pain, loss of bowel/bladder control
- Clinical profile
- Detection rates for genetic testing ~ 65%
- Juvenile subcapsular lens opacities common
- Retinal and choroidal hamartomas in 10-20%
- Café au lait spots < 50%
- Minimal to no cutaneous neurofibromas
- Cutaneous schwannomas in 2/3

Demographics

- Age
- Genetic disease present at conception
- Become symptomatic 2nd-3rd decades
- Gender
- M = F
- Ethnicity
- No racial predilection
- Epidemiology
- 1 in 50,000 live births worldwide
- Intradural spinal tumors are present in up to 65% of patients at initial presentation for imaging
  - 84% have intramedullary tumors
  - 87% have intradural extramedullary tumors

Natural History & Prognosis

- Many have relatively normal lifespans
- Few patients require therapeutic intervention for intramedullary tumors, which often remain quiescent
- Intradural extramedullary tumors frequently lead to surgical intervention
- Percentage of patients with extramedullary tumors who undergo surgery is about 5x higher than percentage of patients with intramedullary tumors
- Higher surgical rate is result of high number of tumors and frequent occurrence of cord compression
- Schwannomas are present more often and in higher numbers than meningiomas and have more surgical procedures overall
- Meningiomas account for a disproportionate number of symptomatic lesions
  - Meningiomas comprise ~ 12% of extramedullary tumors, yet account for 37% of extramedullary tumors requiring excision
  - Suggests NF2 meningiomas are more aggressive

Treatment

- Tumor resection is mainstay of NF2 treatment
- Intramedullary tumors: Monitor with regular imaging and appropriate clinical correlation
- Relatively indolent course of most intramedullary tumors and anticipated burden from other spinal/intracranial tumors must be considered
- Standard aggressive management may not be warranted, even in symptomatic patient
- Intradural extramedullary tumors: Early removal of rapidly growing or symptomatic tumors
- 1st-degree relatives need to be evaluated

DIAGNOSTIC CHECKLIST

Consider

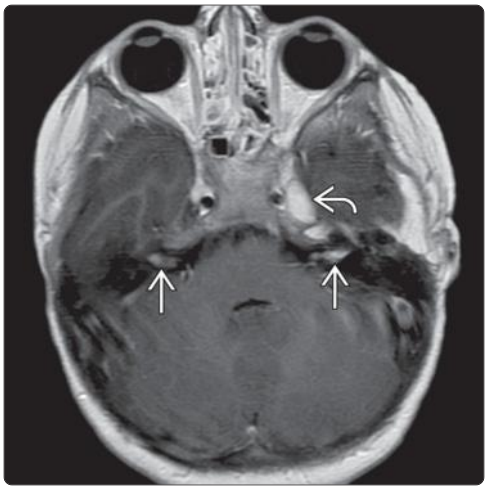
- Presence of multiple and different pathologic types of spinal tumors is highly suggestive of NF2

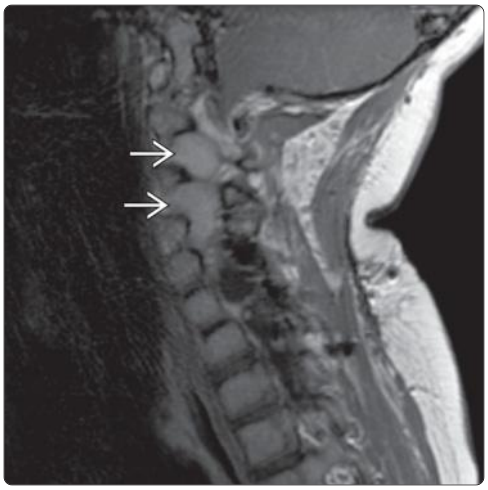
Image Interpretation Pearls

- Screen using MR C+ of brain and entire spine
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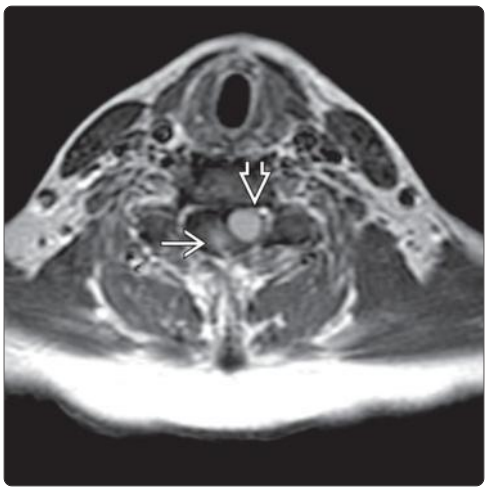


(Left) *Axial T1WI C+ MR of the brain reveals bilateral vestibular schwannomas and a left trigeminal schwannoma, confirming the diagnosis of NF2.* **(Right)** *Parasagittal T1WI C+ MR of the cervical spine shows enhancing foraminal schwannomas.*





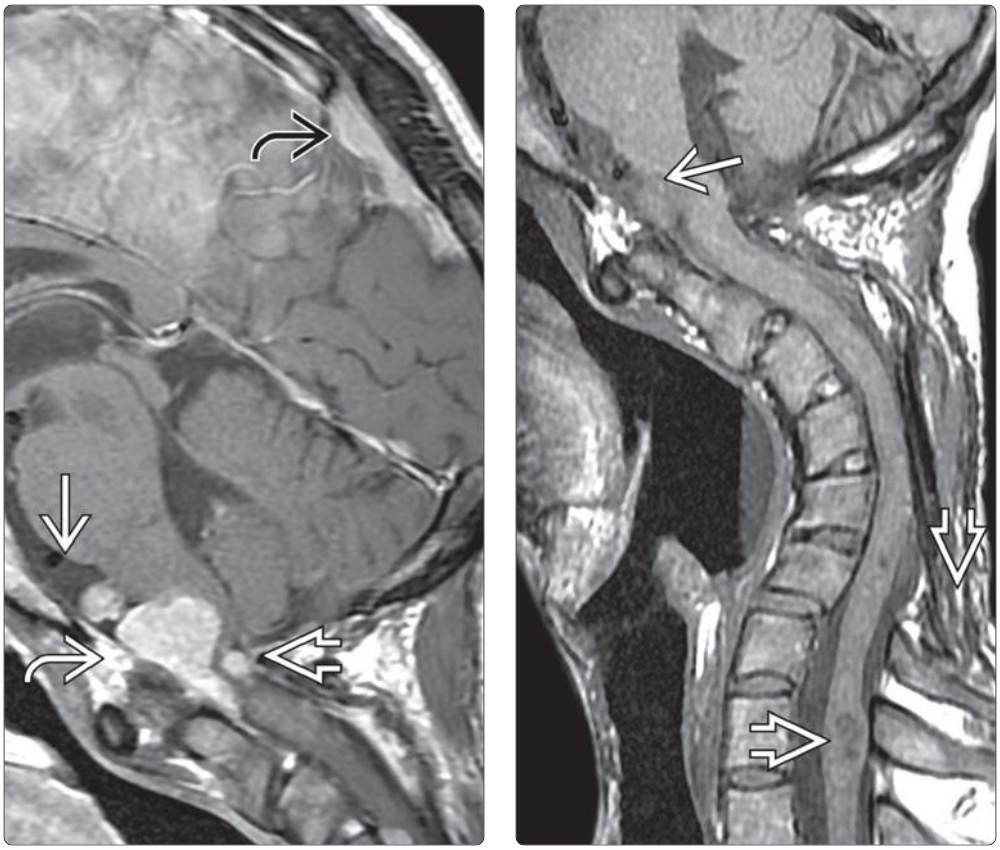
(Left) *Sagittal T1 C+ MR of the lumbar spine in a typical case of spinal neurofibromatosis type 2 demonstrates numerous nodules arising within the cauda equina with avid tumor enhancement, characteristic of schwannoma.* **(Right)** *Sagittal T1WI C+ MR of the lumbar spine shows enhancement of multiple intradural masses scattered along the cauda equina, characteristic of schwannoma.*



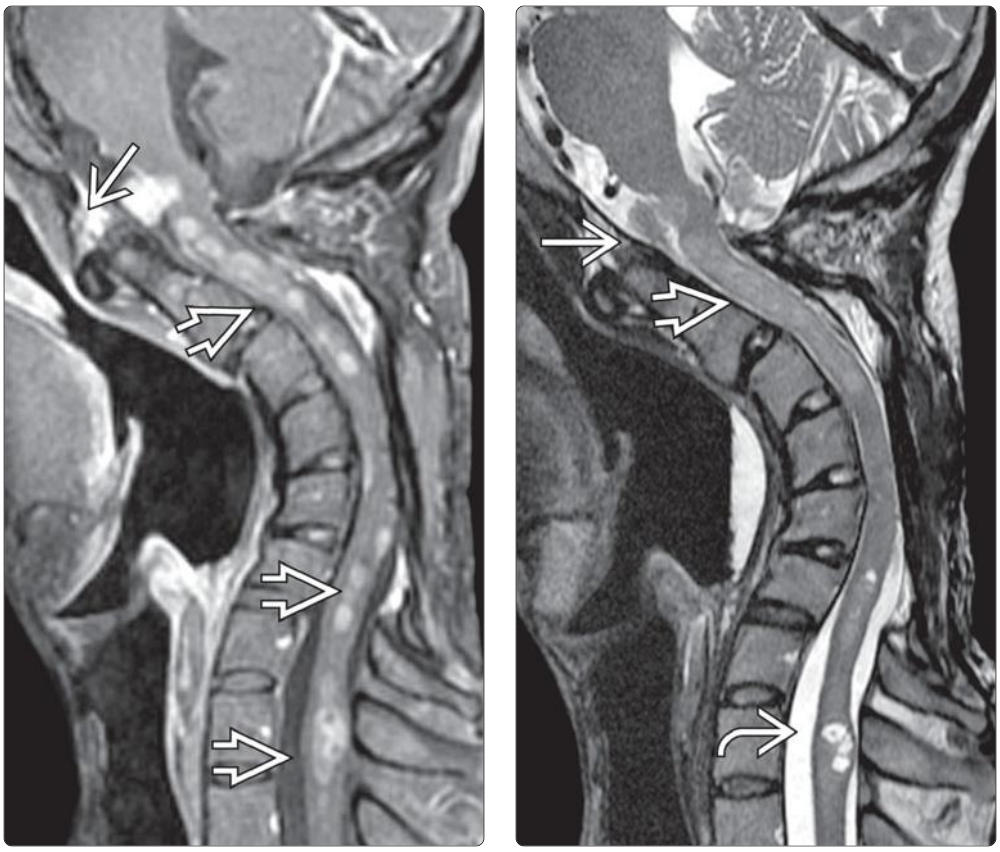


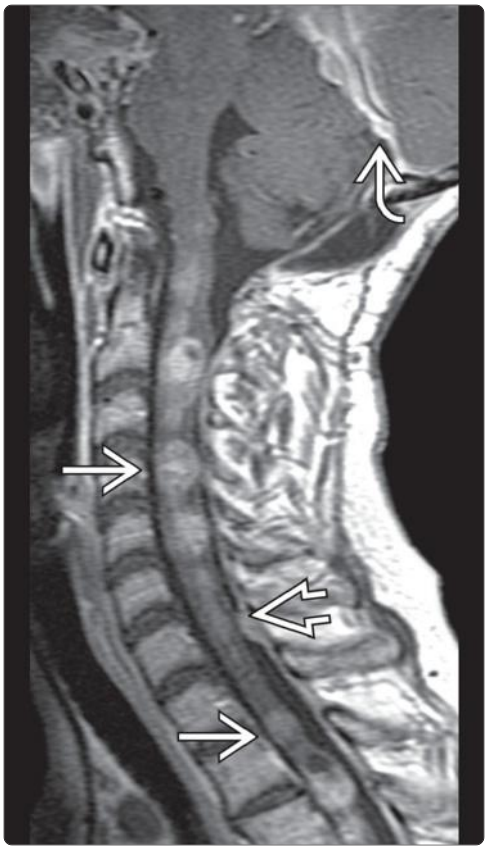
(Left) *Axial T1WI C+ MR of the cervical spine shows both intramedullary ependymoma enhancing and an intradural extramedullary lesion, which is a schwannoma.* **(Right)** *Axial T1WI C+ MR of the lumbar spine demonstrates the origin of the schwannomas to be from the cauda equina nerve roots.*

(Left) *Sagittal T1WI C+ MR shows multiple intradural extramedullary masses at the cervicomedullary junction, reflecting a mixture of meningioma and schwannomas. There is also an enhancing intramedullary ependymoma. Recurrent meningioma seen at the parietooccipital junction.* **(Right)** *Sagittal T1WI MR shows a "swan neck" deformity from prior multilevel laminectomy. Multiple intramedullary lesions within the spinal cord are biopsy-proven ependymomas, vaguely seen as slightly diminished signal. Meningioma is also present at skull base.*



(Left) *Sagittal T1WI C+ MR shows multiple enhancing intramedullary masses throughout the cervical and thoracic cord due to ependymomas in this patient with NF2. Note the enhancing meningioma at the foramen magnum. Patient shows severe "swan neck" deformity as complication of prior multilevel laminectomies for tumor resection.* **(Right)** *Sagittal T2WI MR shows variable signal intensity from the multiple intramedullary tumor throughout the cervical and thoracic cord in this patient with NF2. These vary from near isointense to hyperintense signal. Meningioma is also present at the foramen magnum, seen as a near-cord isointense signal. "Swan neck" deformity present from prior multilevel laminectomy with significant narrowing of subarachnoid space at the laminectomy site.*

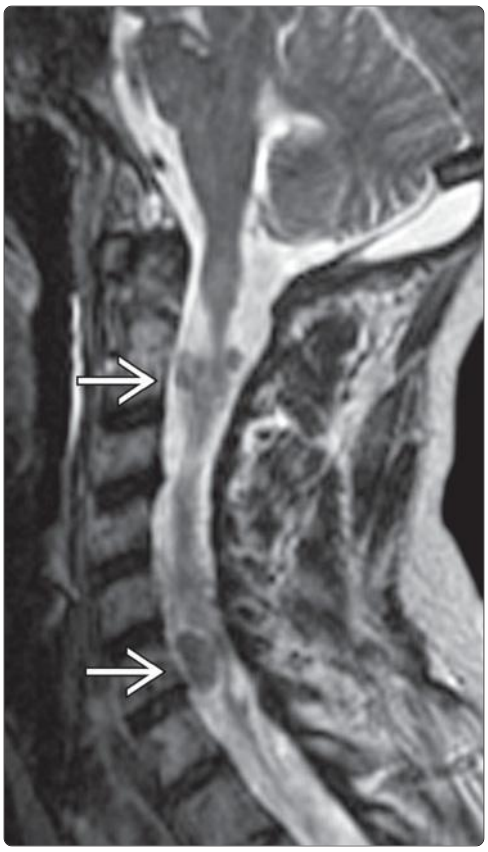






(Left) *Sagittal T1WI C+ MR shows typical severe NF2 cord involvement with multiple intramedullary ependymomas ➡ and multiple intradural extramedullary schwannomas and meningiomas ➡. Enhancing intradural extramedullary lesions and enhancing ependymomas are also seen. Note the intracranial diffuse meningiomas ➡ along the tentorium and juxta sellar region.* **(Right)** *Sagittal T2WI MR shows the intrinsic focal cord lesions at multiple levels consistent with ependymomas ➡. The lesions show well-defined margins with little or no associated cord edema. Scattered dural-based meningiomas are also present ➡. This patient has had prior suboccipital craniectomy for posterior fossa meningioma removal.*





(Left) *Sagittal T1WI C+ MR shows multiple enhancing intradural extramedullary lesions ➡ reflecting both schwannomas and meningiomas. Note the intracranial meningiomas ➡. The presence of multiple and different pathologic types of spinal tumors is highly suggestive of neurofibromatosis type 2.* **(Right)** *Sagittal T2WI MR shows the ID/EM lesions (schwannomas, meningiomas) ➡ as relatively low signal near isointense to the cord parenchyma. Meningiomas comprise 12% of extramedullary tumors, yet account for 37% of extramedullary tumors requiring excision in this population.*

Achondroplasia

KEY FACTS

IMAGING

- Shortened vertebral pedicles
  - Decreasing interpediculate distance toward lower levels of lumbar spine
- Mildly flattened &/or anteriorly wedged vertebral bodies
- Thoracolumbar kyphosis
- Lumbar hyperlordosis
- Small foramen magnum
- Other
  - Growth disturbance more obvious in proximal limbs (rhizomelic dwarfism)
  - "Champagne glass" pelvis: Pelvic inlet is flat and broad
  - Squared iliac wings
  - Short ribs
  - Enlarged skull with constricted skull base

TOP DIFFERENTIAL DIAGNOSES

- Pseudoachondroplasia
- Hypochondroplasia
- Diastrophic dysplasia
- Spondyloepiphyseal dysplasia
- Thanatophoric dysplasia
- Osteogenesis imperfecta

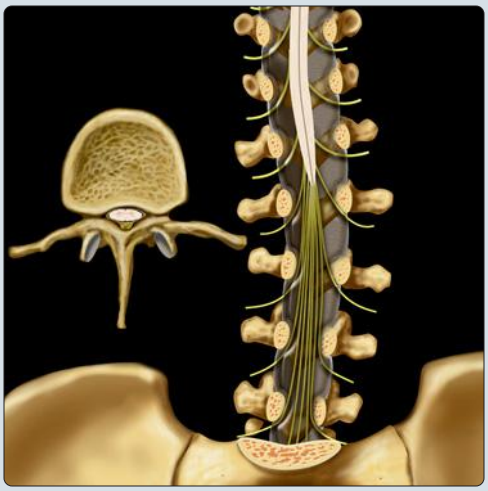
PATHOLOGY

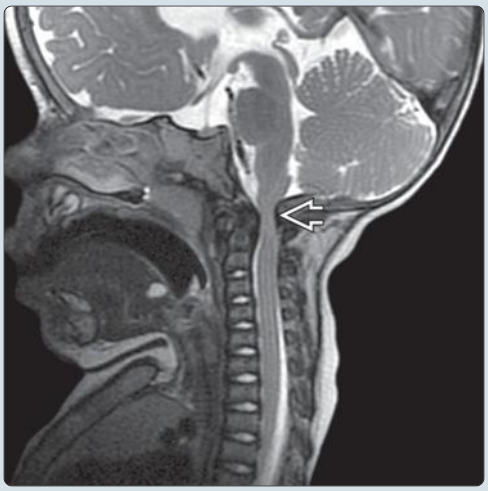
- Defect in *FGFR3*, responsible gene mapped to 4p16.3
- Usually spontaneous mutation (80%)
- Results in defective enchondral bone formation
- Autosomal dominant transmission

CLINICAL ISSUES

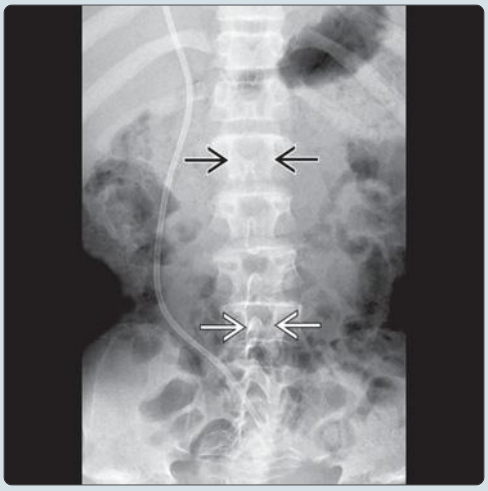
- Most common nonlethal skeletal dysplasia
- High morbidity from spinal stenosis
  - Surgical correction of progressive/unresolving kyphosis
  - Surgical decompression of foramen magnum in severe cases; usually symptoms resolve

(Left) *Graphic shows progressive narrowing of interpediculate distance in caudad direction. Axial insert image shows spinal stenosis related to short pedicles & decreased interpediculate distance.* **(Right)** *Sagittal T2WI MR shows constricted skull base relative to the visualized cranial vault. Stenosis of the foramen magnum compresses the cervicomedullary junction, with mildly increased signal in the upper cervical cord due to myelopathic changes.*





(Left) *AP radiograph of the lumbar spine shows narrowing of the interpediculate distance and progressive interpedicular narrowing between L1 and L4, causing narrowing of the lumbar canal in the transverse dimension.* **(Right)** *Sagittal T2WI MR shows a diffusely narrowed AP dimension of the lumbar spinal canal, reflecting shortened vertebral pedicles. Congenital canal stenosis is further narrowed by a small disc protrusion at L2-L3.*





TERMINOLOGY

Synonyms

- Achondroplastic dwarfism

Definitions

- Autosomal dominant dwarfism affecting spine and extremities
- OMIM #100800

IMAGING

General Features

- Best diagnostic clue
  - Flattened vertebral bodies with short pedicles
  - Interpediculate distance of lumbar spine decreases in caudal direction (reversal of normal relationship)
- Location
  - Characteristic skeletal changes in spine, skull, pelvis, extremities
- Morphology
  - Severe dwarfism involves trunk and extremities
    - Growth disturbance more obvious in proximal limbs (rhizomelic dwarfism)
  - Vertebral bodies mildly flattened
  - Shortened pedicles
  - Lumbar hyperlordosis
  - Thoracolumbar kyphosis

Radiographic Findings

- Radiography
  - Thickened, short lumbar pedicles
    - Decreasing interpediculate distance toward lower levels of lumbar spine
  - Dysmorphic vertebral bodies
    - Bullet-shaped (anteriorly blunted) in childhood
    - Mildly flattened &/or mildly anteriorly wedged
  - Thoracolumbar kyphosis
    - Initially flexible deformity, located T10 through L4
    - Resolves as child begins to walk independently
  - Lumbar hyperlordosis
  - Mild scoliosis may be present
  - C1-C2 instability rare
  - Extraspinal findings
    - "Champagne glass" pelvis: Pelvic inlet is flat and broad
    - Squared iliac wings
    - Shortened long bones, most prominent in proximal limbs ("rhizomelic")
    - "Trident hand": 2nd, 3rd, and 4th digits equal in length
    - Enlarged skull with constricted skull base
  - Bone mineral density normal

CT Findings

- NECT
  - Small foramen magnum
  - Mildly flattened (adult) or bullet-shaped (child) vertebral bodies
  - Shortened vertebral pedicles with narrowing of interpediculate distances
    - Predisposition to significant canal and neural foraminal stenosis with otherwise mild degenerative change
  - Posterior vertebral body scalloping
  - Deformity may lead to stress fractures

MR Findings

- Morphologic changes as described for CT
- Disc herniations common
- T2WI/STIR
  - Compression of cervicomedullary junction, spinal cord, nerve roots → myelopathic signal changes (hyperintensity) in spinal cord

Nonvascular Interventions

- Myelography
  - Spinal stenosis; spinal cord and nerve root compression
  - Disc herniations common

Imaging Recommendations

- Best imaging tool
  - Single AP "babygram" at birth shows characteristic skull, spine, pelvic abnormalities
- Protocol advice
  - Axial, sagittal MR through foramen magnum in all infants and children to assess for stenosis

Ultrasonographic Findings

- Homozygous form can be diagnosed by 2nd trimester
- Growth disturbance evident in 3rd trimester ultrasound

DIFFERENTIAL DIAGNOSIS

Pseudoachondroplasia

- Facial features and skull normal
- Usually detected in early childhood rather than at birth
- Vertebral body flattening variably seen

Hypochondroplasia

- Midface hypoplasia
- Similar to but milder than achondroplasia: Narrowed foramen magnum, decreased interpediculate distances

Diastrophic Dysplasia

- Facial features marked by micrognathia
- Flattened vertebral bodies; narrowed interpediculate distances
- Scoliosis and kyphosis
- C1-C2 instability
- Subluxations in extremities ("hitchhiker's thumb")
- Clubfoot

Spondyloepiphyseal Dysplasia

- Facial features and skull normal
- Severe vertebral body flattening
- C1-C2 instability and odontoid hypoplasia
- Delayed ossification and flattening of epiphyses
- Shortened long bones
- Absent pubic ossification in infants

Thanatophoric Dysplasia

- Lethal dwarfism
- Severe platyspondyly

- Long, narrow trunk
- Bell-shaped thorax

Osteogenesis Imperfecta

- Infantile form has short, thick bones, multiple fractures
- Milder forms present in childhood or adulthood with short stature, platyspondyly, osteopenia, fractures, bone deformities

PATHOLOGY

General Features

- Etiology
  - Fibroblast growth factor receptor 3 (FGFR-3) abnormality
  - Defect in enchondral bone formation
  - Early fusion of pedicles to vertebral bodies
  - Short, thickened pedicles; reduction of spinal canal diameter in transverse and anteroposterior dimensions
    - Symptomatic spinal stenosis, common by 3rd or 4th decade of life
  - Neural foramina often narrowed
- Genetics
  - Gene mapped to chromosome 4p16.3
    - Same chromosome as hypochondroplasia, thanatophoric dwarfism
  - Usually spontaneous mutation (80%)
  - Autosomal dominant transmission
  - Homozygous form rare, much more severe than heterozygous
- Associated abnormalities
  - Sleep apnea, sudden infant death
    - Secondary to compression of brainstem by craniovertebral junction stenosis
  - Chest wall deformity may lead to respiratory difficulties
  - Limb lengthening procedures can lead to neurologic symptoms

Gross Pathologic & Surgical Features

- Spinal stenosis
  - Short pedicles, thickened laminae
  - Disc bulges and herniations contribute to stenosis
  - Can occur at all spinal levels
  - Significant cause of morbidity in adults
- Foramen magnum stenosis
  - Found in almost all children with achondroplasia
  - Often resolves functionally as child grows
- Thoracolumbar kyphosis
  - Related to hypotonia in infants
  - Often resolves as child begins independent ambulation and trunk muscles strengthen
  - Up to 94% prevalence in childhood, decreases to 11% by 10 years of age, then increases to 35% at 4th or 5th decade

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Dwarfism, characteristic facies evident at birth
  - Other signs/symptoms
    - Infantile hypotonia
    - Sleep apnea in infants and children due to compression of cord at foramen magnum
    - Thoracolumbar kyphosis
    - Spinal stenosis
- Clinical profile
  - Rhizomelic dwarfism apparent at birth
  - Frontal bossing and depressed nasal bridge characteristic
  - Intelligence normal
  - Obesity common clinical problem
  - Clinically overdiagnosed in infants as cause of skeletal dysplasia

Demographics

- Age
  - Congenital; diagnosis usually made in infancy
- Gender
  - M = F
- Epidemiology
  - Most common nonlethal skeletal dysplasia (1:26,000 live births)

Natural History & Prognosis

- High morbidity from spinal stenosis

Treatment

- Options, risks, complications
  - Infantile kyphosis may be treatable with sitting modification ± thoracolumbar bracing
  - Surgical correction of progressive/unresolving kyphosis
  - Surgical decompression of foramen magnum in severe cases; usually symptoms resolve
  - Surgical decompression for spinal stenosis
  - Growth hormone to lengthen long bones
  - Ilizarov limb lengthening procedure controversial

DIAGNOSTIC CHECKLIST

Consider

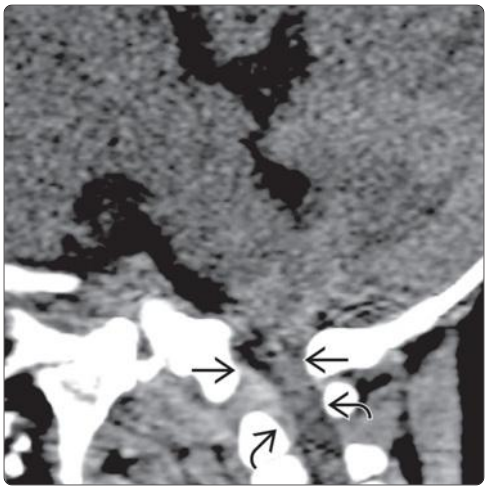
- Most likely cause of short-limbed dwarfism with normal ossification detected in 3rd trimester

Image Interpretation Pearls

- Spine imaging must include foramen magnum to evaluate for stenosis

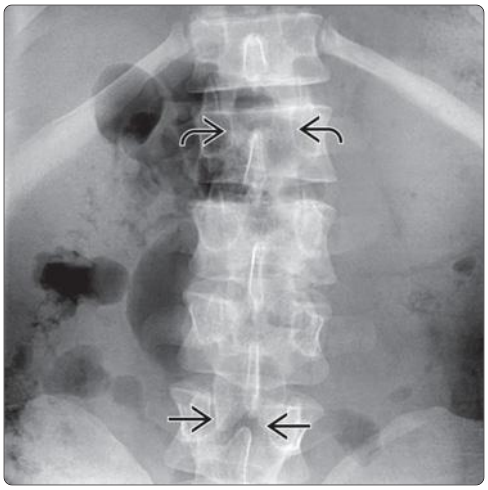
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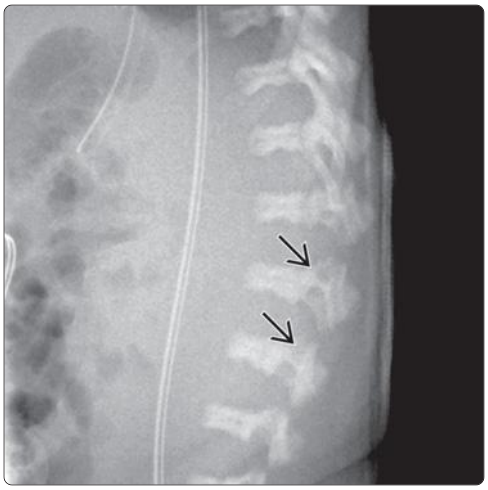
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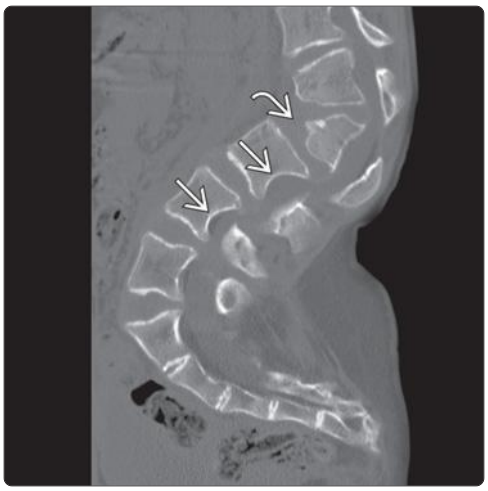


(Left) *Sagittal reconstructed NECT shows narrowed foramen magnum ⇨ and small osseous canal at C1 ⇨ resulting in craniovertebral junction stenosis. Note loss of CSF spaces around the upper cord and cervicomedullary junction.* **(Right)** *Sagittal T1WI shows diffuse, congenital narrowing of the spinal canal. Note posterior vertebral body scalloping ⇨. There is a stenotic foramen magnum, contributing to compression of the cervical medullary junction ⇨.*





(Left) *Anteroposterior radiograph shows decreased interpediculate distance, worsening between L1 ⇨ and L4 ⇨, narrowing the spinal canal in the transverse dimension.* **(Right)** *Lateral radiograph shows platyspondyly in a neonate with achondroplasia. Shortened vertebral pedicles ⇨ cause narrowing of the thoracolumbar canal. Endplates are notched, an abnormality that resolves in childhood.*





(Left) *Sagittal reconstructed NECT shows anteriorly hypoplastic L2 vertebral body ⇨ resulting in severe, focal gibbus deformity. AP diameter of the lumbar canal is diffusely narrowed due to short pedicles. Vertebral scalloping ⇨ is also noted.* **(Right)** *Lateral radiograph shows anteriorly hypoplastic "bullet" vertebrae ⇨ at the thoracolumbar junction. The lumbar canal is narrowed in the AP dimension due to congenitally short pedicles. There is also posterior vertebral body scalloping ⇨.*

KEY FACTS

TERMINOLOGY

- Mucopolysaccharidoses (MPS) are inherited lysosomal storage disorders

IMAGING

- Craniocervical spine
  - CVJ stenosis, dens hypoplasia, ligamentous laxity, atlantoaxial instability, thickened dural ring at foramen magnum
- Thoracolumbar spine
  - Kyphoscoliosis, platyspondyly, anterior vertebral beaking, ± thoracolumbar gibbus deformity

TOP DIFFERENTIAL DIAGNOSES

- GM1 gangliosidosis
- Mucolipidosis III (pseudo-Hurler polydystrophy)
- Achondroplasia
- Trisomy 21 (Down syndrome)
- Spondyloepiphyseal dysplasia

PATHOLOGY

- Inherited lysosomal enzyme deficiency → storage disorder
  - Autosomal recessive (except MPS II Hunter; X-linked recessive)
- Glycosaminoglycan (GAG) accumulates in organs and ligaments

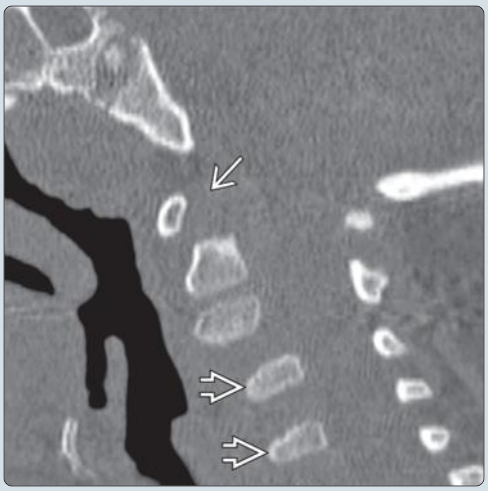
CLINICAL ISSUES

- Gradual progressive myelopathy
- Clinical neurologic symptoms attributable to brain GAG deposition, myelination abnormalities, spinal deformities, peripheral nerve entrapment

DIAGNOSTIC CHECKLIST

- Successful diagnosis requires combination of clinical, imaging, and genetic/biochemical information

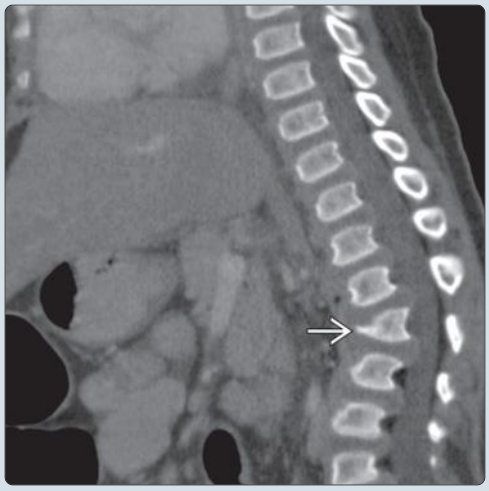
(Left) *Sagittal bone CT (MPS IV, Morquio syndrome) demonstrates nonossification of the odontoid process ⇨, as well as mild hypoplasia of the C3 and C4 vertebral bodies ⇨.* **(Right)** *Sagittal T1WI MR (MPS IV, Morquio syndrome) confirms presence of the unossified odontoid process ⇨ without platybasia or basilar invagination. Characteristic hypoplasia of the subaxial vertebral bodies ⇨ is also demonstrated.*





(Left) *Sagittal T2WI MR (MPS IV, Morquio syndrome) demonstrates hypoplasia and flattening of all thoracic vertebral bodies, as well as focal kyphosis ⇨ and mild spinal canal narrowing at the thoracolumbar junction. The kyphosis results from focal anterior vertebral hypoplasia at the thoracolumbar junction.* **(Right)** *Sagittal bone CT (MPS VI, Maroteaux-Lamy syndrome) reveals mild vertebral hypoplasia and inferior beaking ⇨ at the thoracolumbar junction producing focal kyphosis.*





TERMINOLOGY

Abbreviations

- Mucopolysaccharidosis (MPS)
- Morquio IVA = Morquio A

Synonyms

- "Gargoylism" (not politically correct terminology)

Definitions

- Multiple inherited lysosomal storage disorders
  - Specific enzyme deficiency → inability to breakdown specific glycosaminoglycans (GAG)
  - Failure to break down GAG → intracellular accumulation and toxicity

IMAGING

General Features

- Best diagnostic clue
  - Dens hypoplasia, craniovertebral junction (CVJ) stenosis, and thickened dural ring at foramen magnum
- Location
  - Spine: CVJ, thoracolumbar spine, pelvis
  - Extraspinal: Brain, visceral organ deposition
- Size
  - Odontoid soft tissue mass varies from small to large; larger masses usually found in older patients
- Morphology
  - Craniocervical spine
    - Skull base thickening, occipital hypoplasia, short posterior C1 arch, odontoid hypoplasia ± os odontoideum, ligamentous laxity, dural sac stenosis, and atlantoaxial instability
  - Thoracolumbar spine
    - Kyphosis, kyphoscoliosis
    - Platyspondyly, anterior beaking + thoracolumbar gibbus deformity (MPS I-H and IV)

Radiographic Findings

- Radiography
  - Odontoid dysplasia ± atlantoaxial subluxation
  - Dysplastic blunted spinous processes, wedged vertebral bodies, and spinal canal stenosis
  - Thoracolumbar inferior (MPS I-H) or central (MPS IV) vertebral beaking, gibbus deformity

Fluoroscopic Findings

- ± dynamic CVJ instability on flexion/extension

CT Findings

- CECT
  - CVJ central and foraminal narrowing, marked dural thickening without abnormal enhancement
- Bone CT
  - Abnormal dens ossification, marked laminar thickening, enlargement of medullary cavity

MR Findings

- T1WI
  - Hypo- to isointense periodontoid soft tissue mass, hypointense thickened dura
- T2WI
  - Hypoplastic dens and periodontoid soft tissue mass
  - Hypointense, thickened dura ± cyst formation due to meningeal thickening (MPS 1-H, II)
  - ± cord compression with hyperintense signal abnormality
- T1WI C+
  - No abnormal enhancement, even if mass present
- MRS
  - Brain proton MRS may show diminished NAA/choline ratio, elevated glutamine/glutamate and inositol peak areas

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Spine MR imaging to elucidate cause/site of cord compression
  - Plain radiographs to characterize osseous spine and limb abnormalities
  - Flexion/extension radiographs or fluoroscopy to detect craniovertebral instability

DIFFERENTIAL DIAGNOSIS

GM1 Gangliosidosis

- Shares features of vertebral beaking, upper lumbar gibbus, and dens hypoplasia
- Distinguish on clinical, genetic criteria

Mucolipidosis III (Pseudo-Hurler Polydystrophy)

- Shares features of vertebral beaking, upper lumbar gibbus, and dens hypoplasia
- Distinguish on clinical, genetic criteria

Achondroplasia

- Autosomal dominant disorder of enchondral bone formation
- Short broad pedicles and thickened laminae → spinal stenosis
- Distinguish on genetic, clinical criteria

Down Syndrome

- ± dens hypoplasia without soft tissue dens mass or marrow deposition features
- Distinguish using genetic and clinical information

Spondyloepiphyseal Dysplasia

- Flattening of vertebral bodies, dens hypoplasia, scoliosis present at birth
- Minimal hand and foot involvement

PATHOLOGY

General Features

- Etiology
  - Inherited lysosomal enzyme deficiency
- Genetics
  - Autosomal recessive (except MPS II Hunter; X-linked recessive)
- GAG accumulates in most organs and ligaments
  - Coarse facies (hence the name "gargoylism")
  - Hepatosplenomegaly, umbilical hernia
  - Skeletal dysostosis multiplex, joint contractures

- Arterial wall (midaortic stenosis), cardiac valve thickening
- Upper airway obstruction (38%); very difficult intubations

- Odontoid hypoplasia, GAG deposition, ± ligamentous laxity with reactive change produce soft tissue mass around dens
- Biconvex ovoid, bullet-shaped or rectangular vertebral bodies, vertebral beaking, posterior vertebral slip (MPS IV), large disc protrusions (MPS VI)

Staging, Grading, & Classification

- Hurler (MPS I-H)(OMIM #607014), Hurler-Scheie (MPS IH/S): α-L-iduronidase (4p16.3)
- Hunter (MPS II)(OMIM #309900): Iduronate 2-sulfatase (Xq28)
- Sanfilippo (MPS IIIA)(OMIM #252900): Heparin N-sulfatase (17q25.3)
- Morquio (MPS type IVA)(OMIM #253000): Galactose 6-sulfatase (16q24.3)
- Scheie (MPS IS)(MPS V, formerly)(OMIM #607016): α-L-iduronidase (4p16.3)
- Maroteaux-Lamy (MPS VI)(OMIM #253200): Arylsulfatase B (5q11-q13)
- Sly (MPS VII)(OMIM #253220): β-glucuronidase (7q11.21)

Gross Pathologic & Surgical Features

- CVJ stenosis → neurovascular compression, altered CSF dynamics → hydrocephalus, hydrosyringomyelia
- Dilated enlarged laminae medullary cavities
- Thickened dura may appear normal at surgery

Microscopic Features

- Epidural/dural mucopolysaccharide deposition with elastic and collagenous proliferation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Gradual subtle progressive myelopathy (often falsely attributed to lower extremity deformities)
    - Common with MPS I-H, II, III, and VII; uncommon with MPS IV, I H/S, VI unless associated with musculoskeletal deformities
    - Reduced exercise tolerance; may be earliest symptom
  - Clinical neurologic symptoms attributable to brain GAG deposition, myelination abnormalities, spinal deformities, and peripheral nerve entrapment
- Clinical profile
  - Coarse facies with macroglossia, bushy eyebrows, flat nasal bridge (mild in MPS VI, VII)
  - Corneal clouding (except MPS II)
  - Significant mental retardation (except in MPS I H/S, IIb, IV)
  - Joint contractures, dysostosis multiplex (dominates in MPS IV, VI)
- Laboratory diagnosis: Measurement of specific urinary glycosaminoglycans
- Clinical diagnostic algorithm
  - Age, IQ, ± corneal clouding, urinary GAG excretion, and specific clinical findings

Demographics

- Age
  - Usually diagnosed in childhood; occasional mild cases diagnosed as adults
- Gender
  - M = F, except MPS II (boys only → X-linked)
- Epidemiology
  - MPS I-H (1:10,000 births), MPS IV (1:40,000 births) most common

Natural History & Prognosis

- Premature death is the rule; rate of deterioration depends on specific enzymatic deficiency
- Slowly progressive cord compression → quadriparesis, sensory loss without treatment
  - Surgical results poor when performed late in disease course
- High spinal cord compression is major spinal cause of MPS complications and death
  - Apnea and sudden death may follow relatively minor trauma

Treatment

- Conservative (mild symptoms): External spinal bracing
- Surgical (more severe symptoms)
  - Posterior occipitocervical decompression/stabilization
  - Transoral odontoid resection/posterior stabilization for symptomatic odontoid mass effect
- Bone marrow transplant (BMT) or intravenous recombinant human enzyme
  - Decreases GAG accumulation in organs; ameliorates some, but not all manifestations

DIAGNOSTIC CHECKLIST

Consider

- Successful diagnosis requires combination of clinical, imaging, and genetic/biochemical information

Image Interpretation Pearls

- Dens hypoplasia with mass and dural thickening suggests MPS syndrome
- Vertebral beaking pattern may permit specific diagnosis

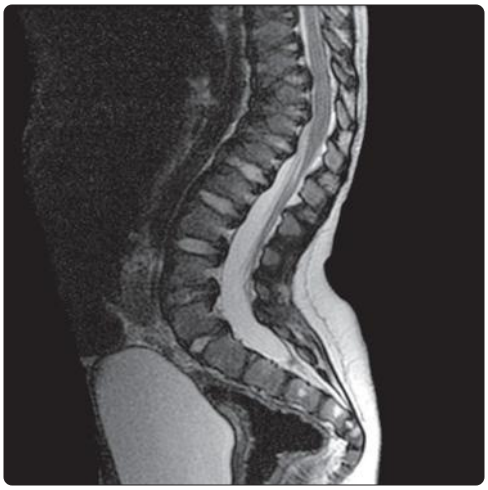
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(Left) *Lateral plain radiograph of the cervical spine (Hunter syndrome, MPS II) shows mild anterior inferior vertebral beaking. As a general rule, the imaging (and clinical) findings of Hunter syndrome are similar to Hurler MPS I-H, but usually less severe and slower to progress.* **(Right)** *Lateral radiograph of the thoracolumbar spine (Hunter syndrome, MPS II) reveals inferior vertebral beaking, as well as a hypoplastic L1 vertebra ⇨ producing focal kyphosis.*





(Left) *Sagittal T2WI MR (MPS IV, Morquio syndrome) demonstrates characteristic thoracolumbar vertebral anomalies; although, the lower lumbar vertebra are less severely affected than the thoracolumbar vertebra. Prominent kyphosis at the thoracolumbar junction related to vertebral hypoplasia is commonly observed.* **(Right)** *Axial T2WI MR (MPS IV, Morquio syndrome) of the lumbar spine confirms coarse thickening of the posterior spinal elements without spinal stenosis.*





(Left) *Sagittal T2WI MR (MPS VI) demonstrates mildly abnormal vertebral body morphology. In addition, there is segmental syringohydromyelia ⇨ extending from C5 to T1.* **(Right)** *Sagittal T2WI MR (MPS VI) reveals mild lumbar kyphosis centered at the hypoplastic T12 vertebra ⇨, with multiple other abnormal anteriorly beaked vertebral bodies. Note also moderate narrowing of the thoracolumbar spinal canal at the apex of the kyphosis and abnormal spinal cord T2 signal intensity ⇨.*

KEY FACTS

IMAGING

- Spine involvement in 43-70% of patients
- Bone infarction
  - Patchy or diffusely sclerotic bones
  - Central endplate compression deformities in spine, resulting in H-shaped vertebral bodies
- Osteopenia
  - Osteoporotic compression fractures
- Tower vertebra
  - Compensatory vertical growth of vertebral body adjacent to infarcted/collapsed vertebra
- Osteomyelitis
  - Rare in spine

TOP DIFFERENTIAL DIAGNOSES

- Thalassemia
- Renal osteodystrophy
- Osteopetrosis
- Spondyloepiphyseal dysplasia (SED)
- Diffuse osseous metastases

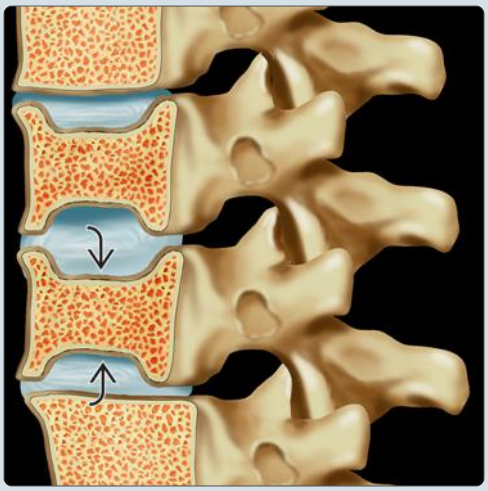
PATHOLOGY

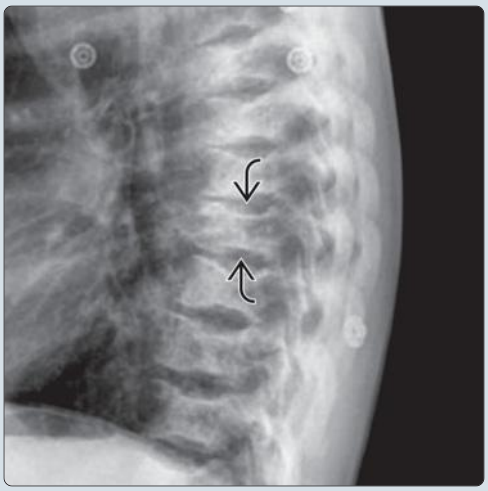
- Autosomal recessive inheritance
  - African, Middle Eastern, and Eastern Mediterranean
- Associated abnormalities
  - Moyamoya disease, multiple infarcts
  - Femoral head and humeral head infarcts
  - Growth disturbance
  - Cholelithiasis (bilirubin stones)
  - Hemosiderosis, due to multiple transfusions

CLINICAL ISSUES

- Sickle cell crisis: Oxygen, hydration, pain management, blood transfusion
- High incidence of infections
  - Prophylactic vaccinations to prevent infection

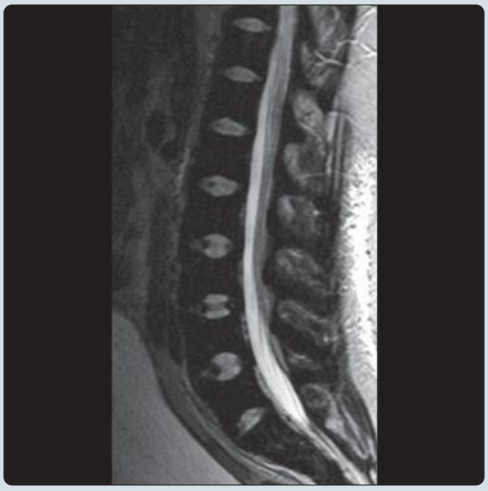
(Left) *Sagittal graphic shows central collapse of the vertebral endplates due to bone infarcts, which tends to affect the central portion of the vertebral body. Peripherally, height is preserved, yielding an H-shaped vertebra.* **(Right)** *Lateral radiograph shows patchy sclerosis in the visualized osseous structures and central endplate compression deformities resulting in characteristic H-shaped vertebral bodies.*





(Left) *Axial NECT shows diffuse, patchy sclerosis in a lower thoracic vertebra due to chronic bone infarcts in a patient with sickle cell disease.* **(Right)** *Sagittal T2WI MR of the lumbar spine shows markedly hypointense marrow signal, reflecting iron deposition in a patient requiring multiple transfusions.*





TERMINOLOGY

Synonyms

- Sickle cell anemia, HbSS disease

Definitions

- Hereditary hemoglobinopathy resulting in anemia and red blood cells that deform (sickle) and occlude blood vessels
- Homozygous: HbSS (sickle cell anemia)
- Heterozygous: HbSA (sickle cell trait, asymptomatic), HbSC (less severe form)
- Sickle cell crisis: Acute episode of severe bone, abdomen, chest pain

IMAGING

General Features

- Best diagnostic clue
  - H-shaped vertebral bodies (on sagittal imaging) due to central endplate compression deformities
- Location
  - Spine involvement in 43-70% of patients
  - Bone infarcts much more commonly involve long bones of appendicular skeleton

Radiographic Findings

- Radiography
  - Bone infarction
    - Patchy or diffusely sclerotic bones due to chronic, repeated bone infarcts
    - Central endplate compression deformities in spine, resulting in H-shaped vertebral bodies
  - Osteopenia
    - Osteoporotic vertebral compression fractures, with biconcave or wedged morphology
  - Tower vertebra
    - Compensatory vertical growth of vertebral body adjacent to infarcted/collapsed vertebra
  - Osteomyelitis
    - Rare in spine

CT Findings

- NECT
  - Mottled sclerosis in medullary spaces of axial skeleton
  - Central endplate compression deformities (on sagittal or coronal reconstructed images), resulting in H-shaped vertebral bodies
  - Osteoporotic compression fractures

MR Findings

- T1WI
  - Low T1 signal intensity marrow due to: Hyperplastic marrow, sclerosis due to chronic bone infarcts, infection, iron deposition with transfusion-dependent HbSS disease
  - Round, paravertebral soft tissue masses reflect extraosseous hematopoiesis; may be seen with severe, chronic anemia
- T2WI, STIR
  - Hyperplastic (hematopoietic) marrow: Intermediate to low T2 signal intensity
  - Iron deposition with transfusion-dependent HbSS disease: Very low T2 signal intensity
  - Acute bone infarct: Diffuse area of hyperintense marrow signal
  - Osteomyelitis: Diffuse area of hyperintense marrow signal
  - Round, paravertebral soft tissue masses reflect extraosseous hematopoiesis; may be seen with severe, chronic anemia
- T1WI C+
  - Bone infarct: Thin, serpentine rim of enhancement
  - Osteomyelitis: Diffuse enhancement or coarse peripheral rind of enhancement
    - Adjacent soft tissue enhancement with focal cortical defect/enhancement

Imaging Recommendations

- Best imaging tool
  - Radiography

Nuclear Medicine Findings

- Bone scan
  - Bone infarct: Acutely photopenic, with increased activity during healing and revascularization
  - Osteomyelitis: Increased activity
- Tc-99m sulfur colloid
  - Acute bone infarct: Diminished or absent marrow uptake
  - Osteomyelitis: Normal or decreased marrow uptake
    - Marrow uptake may be normal in very early phase (< 1 week) of infection
- Ga-67 scintigraphy
  - Acute bone infarct: Congruent uptake relative to Tc-99m sulfur colloid marrow scan
  - Osteomyelitis: Increased uptake or incongruently increased relative to Tc-99m sulfur colloid marrow scan
- Labeled leukocyte scintigraphy
  - Osteomyelitis: Increased activity without corresponding activity on Tc-99m sulfur colloid marrow scan

DIFFERENTIAL DIAGNOSIS

Thalassemia

- Avascular necrosis less common than in sickle cell anemia

Radiographic Mimics

- Renal osteodystrophy
  - Thickened, dense vertebral endplates
- Osteopetrosis
  - Diffuse bone sclerosis
  - "Bone-in-bone" sign
- Spondyloepiphyseal dysplasia (SED)
  - Vertebrae with cup-shaped endplates
- Diffuse osseous metastases
  - Diffuse low signal marrow on T1WI MR, intermediate to hyperintense T2WI
  - Leukemia, lymphoma
  - Neuroblastoma

PATHOLOGY

General Features

- Etiology

- Abnormal B-chain of hemoglobin tetramer
- Tendency for red blood cells to deform ("sickle") on deoxygenation
  - Decreased RBC pliability → capillary obstruction → ischemia and infarction
- Splenomegaly (RBC sequestration, destruction) → atrophy (autoinfarction)
  - Autoinfarction leads to decreased host resistance to encapsulated bacteria
    - *Salmonella typhi*, *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Neisseria meningitidis*, *Klebsiella pneumoniae*
  - Pneumonia
  - Osteomyelitis
  - Meningitis
  - Sepsis
- H-shaped vertebral bodies
  - Central endplate bone vascular supply from long branches of vertebral nutrient artery
    - More susceptible to vasoocclusive effects of sickled red blood cells
  - Peripheral portion of endplate supplied by short perforating branches of periosteal vessels
  - Central osteonecrosis → central collapse → H-shaped vertebral body in sagittal plane
- High prevalence of abnormal bone mineral density (BMD), especially in lumbar spine
  - Increased BMD due to healed bone infarcts
  - Decreased BMD due to chronic, severe anemia
    - Hyperplasia of marrow elements causes decreased trabecular bone and osteopenia
    - Osteonecrosis
  - One series reported 79.6% incidence of low BMD in sickle cell disease, irrespective of age or gender
  - Osteoporotic vertebral compression fractures
  - Other insufficiency fractures

- Genetics
  - Autosomal recessive inheritance
  - Structural defect in hemoglobin HbS: Glutamic acid in position 6 substituted with valine
- Associated abnormalities
  - Moyamoya disease, multiple infarcts
  - Femoral head and humeral head infarcts
  - Growth disturbance
    - Limb length discrepancy
  - Cholelithiasis (bilirubin stones)
  - Hemosiderosis, due to multiple transfusions

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sickle cell crisis
    - Painful episode due to vasoocclusive crisis, most commonly involving bone
  - Hemolytic anemia
- Clinical profile
  - Recurrent pain crises, jaundice, growth retardation, stroke
  - High incidence of infections
    - Pneumococcal septicemia, meningitis
    - Osteomyelitis usually seen in children

Demographics

- Age
  - Children protected during 1st 6 months by elevated levels of fetal Hb (HbF)
- Ethnicity
  - African, Middle Eastern, and Eastern Mediterranean ethnic heritage
- Epidemiology
  - 1% of African Americans homozygous HbSS; 8-13% are heterozygous HbSA (asymptomatic)
  - 3% of African Americans are HbS carriers
  - In one series, 26% of all sickle cell bony crises resulting in hospital admission involved spine
    - 2nd most common site of bony involvement (after lower extremity)

Natural History & Prognosis

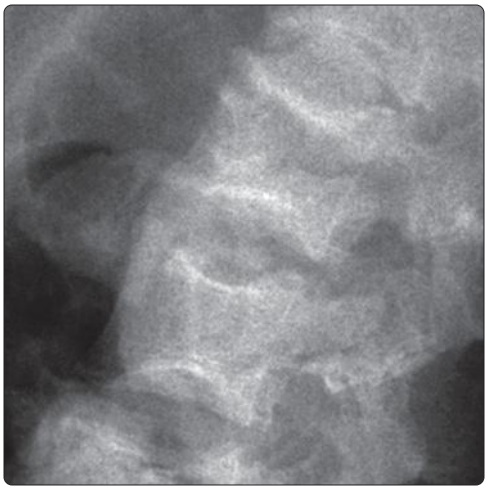
- Death < 50 years in homozygous form

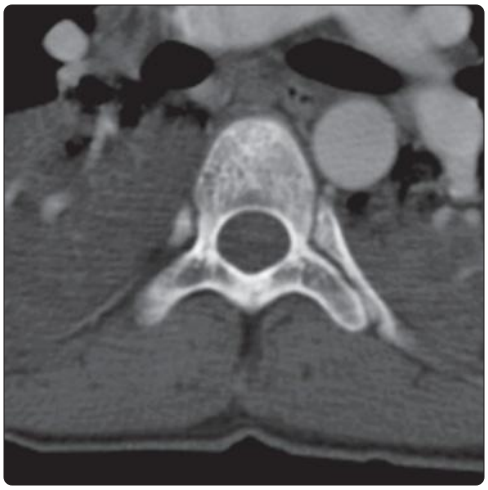
Treatment

- Sickle cell crisis: Oxygen, hydration, pain management, blood transfusion
- Prophylactic vaccinations to prevent infection

SELECTED REFERENCES

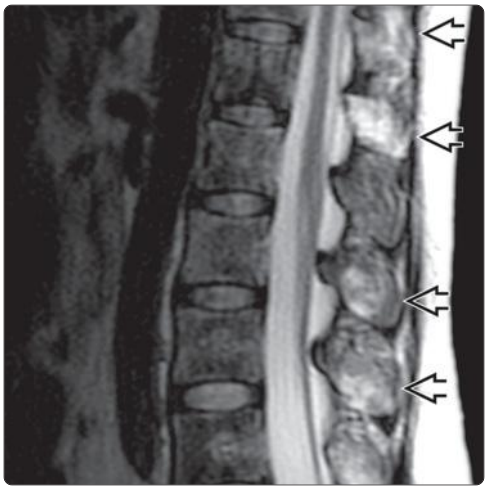
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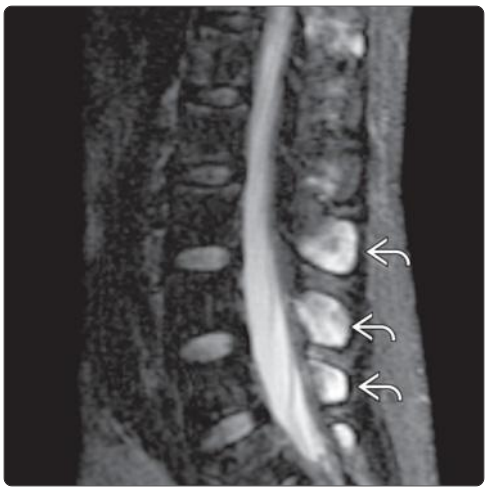


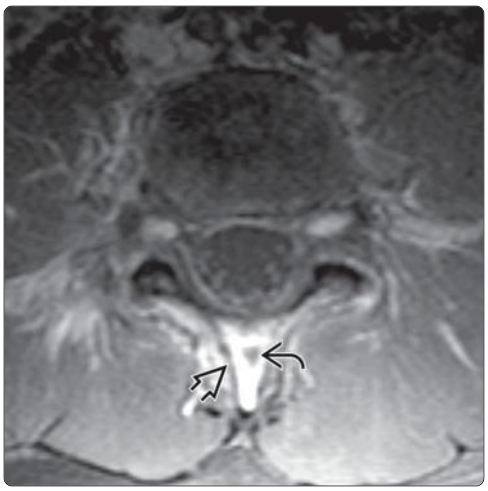
(Left) *Lateral radiograph shows the classic spine involvement of sickle cell anemia, with central depression of the endplates sparing the periphery of the vertebral bodies.* **(Right)** *Axial CECT shows diffuse ground-glass sclerosis of the vertebral body. Note also extensive consolidation of the lower lobes of both lungs from severe bilateral pneumonia, which can occur in sickle cell anemia patients due to impaired immunity following splenic infarction.*





(Left) *Sagittal T1WI MR shows hypointense marrow signal, lower than expected than for erythroid hyperplasia and more characteristic of hemosiderin deposition due to multiple transfusions. There are also central endplate compression deformities, resulting in the characteristic H-shaped vertebral bodies.* **(Right)** *Sagittal STIR MR displays low signal in vertebral bodies due to marrow hyperplasia. High signal in multiple spinous processes is consistent with bone infarctions.*





(Left) *Sagittal STIR of the lumbar spine shows hyperintense signal in the spinous processes of L3, L4, and L5, ultimately determined to be due to acute bone infarcts.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows peripheral enhancement around a tiny nonenhancing infarct in a lumbar spinous process. Circumscribed margins of the enhancement and lack of adjacent soft tissue changes favor infarct over osteomyelitis.*

Osteogenesis Imperfecta

KEY FACTS

TERMINOLOGY

- Genetic disorder of type I collagen resulting in bone fragility
- Classified into 4 types based on clinical, genetic, and radiographic criteria

IMAGING

- Severe osteopenia
- Vertebral fractures, kyphoscoliosis
- Multiple long bone, rib fractures
- Enlarged epiphyses, "popcorn" metaphyseal calcifications
- Medullary cavity nearly entirely filled with fat
  - Primary trabeculae sparse but normally oriented
  - Secondary trabeculae nearly absent

TOP DIFFERENTIAL DIAGNOSES

- Nonaccidental trauma (NAT)
- Congenital dwarfism
- Osteoporosis

PATHOLOGY

- Numerous type I collagen mutations → "brittle" bone
  - Most autosomal dominant
  - Inherited or spontaneous mutation
- Associated anomalies include blue sclerae, early hearing loss, brittle teeth, thin fragile skin, joint laxity

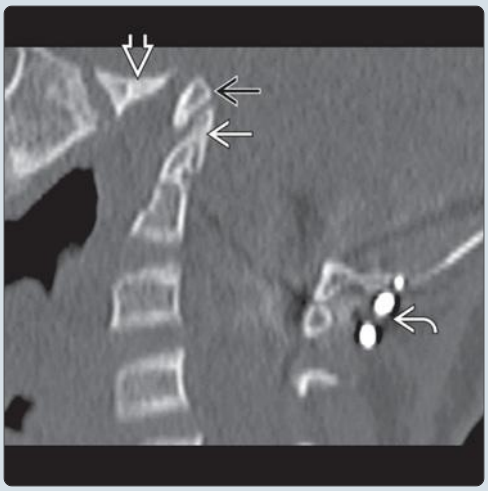
CLINICAL ISSUES

- Short stature secondary to multiple spinal and extremity fractures, kyphoscoliosis, growth plate abnormalities
- Diagnosis suggested by radiographs, confirmed with ancillary testing

DIAGNOSTIC CHECKLIST

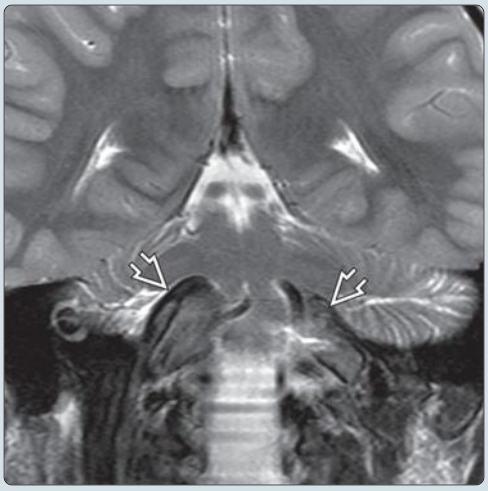
- Important to differentiate from nonaccidental trauma
- Basilar impression and other spinal complications may be difficult to detect on radiographs
  - Consider MR or CT

(Left) *Sagittal bone CT demonstrates platybasia and severe basilar impression, with upward displacement of the remodeled odontoid process and anterior C1 ring through the foramen magnum. Metallic posterior spinal hardware has been placed to arrest basilar impression.* **(Right)** *Sagittal T2WI MR reveals severe cranial settling and platybasia with basilar impression of the odontoid process into the foramen magnum, producing ventral cervicomedullary compression.*





(Left) *Coronal T2WI MR confirms upward sloping of the medial skull base related to bone softening and severe basilar impression. This results in compression of the brainstem and cerebellar hemispheres.* **(Right)** *Axial T2WI MR confirms medullary distortion by the clivus related to cranial settling. Metallic susceptibility artifact reflects spinal surgical fusion hardware intended to arrest cranial settling.*





TERMINOLOGY

Synonyms

- "Brittle bone disease"

Definitions

- Genetic disorder of type I collagen resulting in bone fragility
- Classified into 4 types based on clinical, genetic, and radiographic criteria
  - Severe forms: Bones short, thickened due to multiple fractures in utero and early childhood
  - Milder forms: Bones thin and gracile

IMAGING

General Features

- Best diagnostic clue
  - Severe osteopenia with multiple fractures, kyphoscoliosis
- Location
  - Entire axial and appendicular skeleton
- Morphology
  - Multiple skeletal fractures common

Radiographic Findings

- Radiography
  - Osteoporosis
    - Cortical thinning, resorption of secondary trabeculae with prominent primary trabeculae
  - Kyphoscoliosis
  - Vertebral fractures, vertebra plana, "codfish vertebrae"
    - Codfish vertebra: Cupping of superior and inferior vertebral body endplates
  - Multiple fractures of long bones and ribs with increased anteroposterior chest diameter
    - Bowing deformity due to microfracture, as well as radiographically evident fractures
  - Enlarged epiphyses
  - "Popcorn" metaphyseal calcifications
    - Calcified cartilage nodules due to fragmentation of growth plate from trauma
  - Pelvis: Protrusio acetabulae, coxa vara

CT Findings

- Bone CT
  - Thin bony cortices
  - Medullary cavity nearly entirely filled with fat
    - Primary trabeculae sparse, but normally oriented
    - Secondary trabeculae nearly absent
  - Kyphoscoliosis
  - Basilar impression
  - Otospongiosis (temporal bone)

MR Findings

- T1WI
  - Kyphoscoliosis
  - Abnormal marrow signal intensity related to fractures, fatty infiltration
- T2WI
  - Same as T1WI
  - ± spinal cord compression, syringohydromyelia

Ultrasonographic Findings

- Grayscale ultrasound
  - Poorly ossified skull, short ribs, short deformed limbs
  - Antenatal diagnosis possible during 2nd trimester

Nuclear Medicine Findings

- Bone scan
  - Increased radiotracer uptake at fracture sites

Other Modality Findings

- ↓ bone density on dual energy x-ray absorptiometry (DEXA) or CT
  - Used to aid diagnosis (mild forms), follow response to medical treatment

Imaging Recommendations

- Best imaging tool
  - Radiography for diagnosis, kyphoscoliosis characterization
  - MR best to evaluate for spinal cord compression, basilar impression

DIFFERENTIAL DIAGNOSIS

Nonaccidental Trauma (NAT)

- Normal bone mineral density
- Fractures may otherwise appear identical to OI
- Careful history, family evaluation required
- Genetic testing may be necessary

Congenital Dwarfism

- Loss of vertebral body height
- Short stature
- Scoliosis
- Bone density variable
- Common etiologies
  - Achondroplasia
  - Thanatophoric dwarfism
  - Spondyloepiphyseal dysplasia

Osteoporosis

- Thinned bone cortices, accentuation of primary trabeculae, resorption of secondary trabeculae
- Radiography insensitive for diagnosis; better evaluated by bone densitometry
- "Codfish vertebrae," compression fractures common
- Myriad etiologies

PATHOLOGY

General Features

- Etiology
  - Numerous type I collagen mutations → abnormal type I collagen, brittle bone
    - Type I collagen found in bone, skin, sclerae
- Genetics
  - Most autosomal dominant
  - Inherited or spontaneous mutation
- Associated abnormalities
  - Blue sclerae
  - Early hearing loss
  - Brittle teeth

- Thin, fragile skin
- Joint laxity
- Respiratory, cardiac problems

Staging, Grading, & Classification

- Type I: Most common
  - Thin, gracile long bones
  - High fracture risk in childhood, decreases after puberty
  - Kyphoscoliosis, wormian bones
  - Blue sclerae
- Type II: Often lethal early in life
  - Short, broad, deformed bones at birth due to multiple fractures
- Type III: Autosomal recessive; rare
  - Fractures at birth, kyphoscoliosis
- Type IV: Similar to type I
  - Kyphoscoliosis, wormian bones
  - Sclerae usually normal in adulthood, may be blue in childhood
- Recently types V-XII have been proposed; not distinguishable radiographically
  - These types do not localize to genes for type I collagen; etiology unknown
- Terms "congenita" and "tarda" no longer used

Gross Pathologic & Surgical Features

- Thin, eggshell-like bone cortices
- Decreased medullary trabeculae
- Recent or healed fractures

Microscopic Features

- Lack of organized trabeculae
- Prominent osteoid seams
- Increased numbers of morphologically normal osteoblasts
- Growth plate fragmentation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Multiple spinal and extremity fractures
  - Kyphoscoliosis
- Other signs/symptoms
  - ± deafness, blue sclerae
- Clinical profile
  - Short stature secondary to fractures, kyphoscoliosis, growth plate abnormalities
  - Diagnosis suggested by radiographs, confirmed with ancillary testing
    - Skin biopsy
    - Genetic testing
    - Caveat: Both tests may be false-negative

Demographics

- Age
  - Findings often evident at birth
  - Mild cases may present in adulthood
- Gender
  - M = F
- Epidemiology
  - 4/100,000 births

Natural History & Prognosis

- Severe forms, previously lethal, now may survive to adulthood
- Growth retardation varies with severity of disease
- Fractures less common after puberty

Treatment

- Conservative management
  - Bisphosphonates variably successful for medical therapy
  - Prolonged immobilization avoided because of resultant worsened osteoporosis
- Surgical management
  - Spinal fusion for kyphoscoliosis
    - Aggressive surgical decompression stabilization procedures may halt basilar impression progression and facilitate good long-term functional outcome
    - High risk of hardware mechanical failure
    - May be future role for kyphoplasty as alternative management
  - Intramedullary rod placement for long bone fractures
    - Least invasive internal fixation for rapid mobilization
    - Helps prevent progression of deformity, further fractures

DIAGNOSTIC CHECKLIST

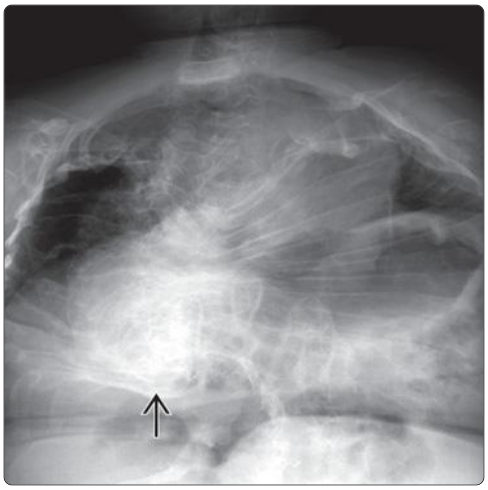
Consider

- Important to differentiate from nonaccidental trauma
- Basilar impression and other spinal complications may be difficult to detect on radiographs
  - Consider MR or CT

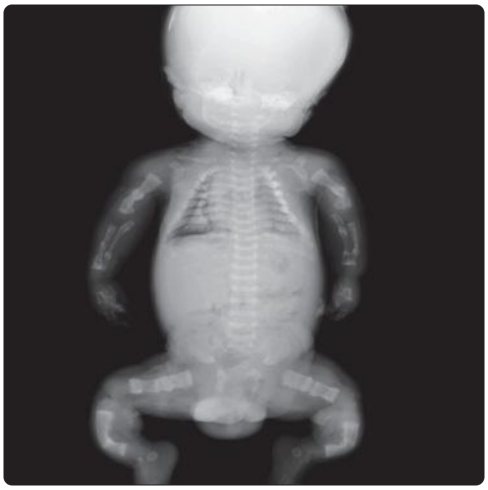
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(Left) *Anteroposterior radiograph of the thoracic spine shows convex right neuromuscular scoliosis with spinal rods. There is also osteopenia and ribbon-like ribs, characteristic findings of OI.* **(Right)** *Anteroposterior radiograph demonstrates osteopenia and severe scoliosis, with a > 130° curvature at the thoracolumbar junction. This, in addition to multiple compression fractures, results in marked shortening of the trunk. Note also multiple healing rib fractures.*





(Left) *Frontal radiograph of a fetus obtained during fetal imaging autopsy demonstrates numerous fractures of all extremities and the ribs in varying states of repair, as well as characteristic platyspondyly and underossification of the calvaria.* **(Right)** *Coronal bone CT of the cervical spine performed to evaluate severe headaches (type I OI) confirms diffuse severe osteopenia. No collapse of the occipital condyles or C1 lateral masses is confirmed.*





(Left) *Sagittal bone CT (neonatal OI) shows abnormal "swan neck" deformity of the cervical spine. The anterior predental space is mildly widened, suggesting atlantoaxial instability. No basilar impression or platybasia is present.* **(Right)** *Axial bone CT shows generalized osteopenia with retained bone density in the otic capsule, normally the densest bone in the human body. There is also bilateral dysplasia of the cochlea and vestibular apparatus.*

KEY FACTS

TERMINOLOGY

- Inherited tumor disorder with multiorgan hamartomas

IMAGING

- Best diagnostic clue
  - Brain: Calcified subependymal nodules (hamartomas)
  - Spine: Multiple foci of bony sclerosis (bone islands)
- Radiography/CT spine
  - Regions of bony sclerosis, which may be round, oval, or flame-shaped
    - Homogeneously dense or mottled
    - Should not expand bone or transgress bony margins
  - Bone cysts are other skeletal manifestation
    - Often in phalanges of hand
- MR spine: Sclerosis seen as low signal foci on all sequences in multiple bones

TOP DIFFERENTIAL DIAGNOSES

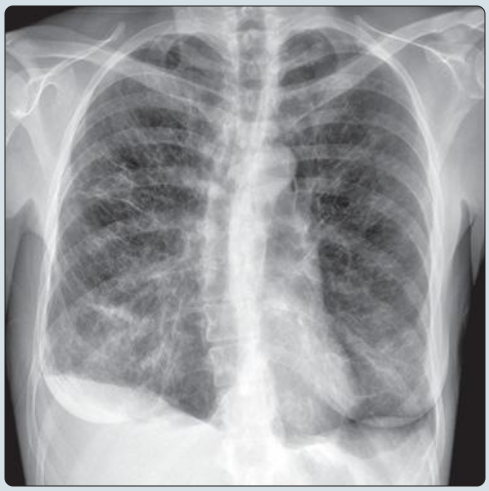
- Sporadic lymphangioleiomyomatosis (LAM)
- Blastic metastatic disease
- Paget disease
- Pycnodysostosis
- Hypervitaminosis A or D
- Myelofibrosis
- Osteopetrosis
- Bone island

PATHOLOGY

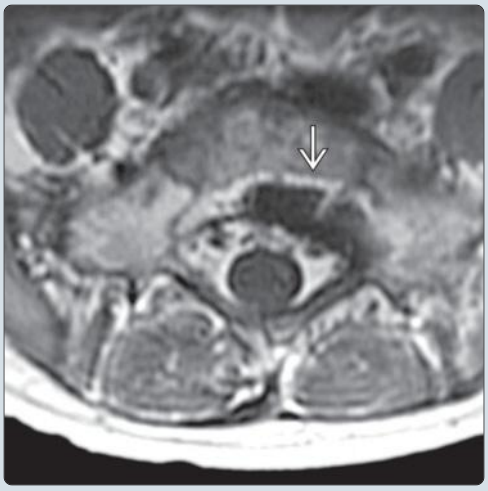
- Mutations in TSC tumor suppressor genes cause abnormal cellular differentiation, proliferation
- Sclerotic bone lesions on CT scans are more common and numerous in patients with TSC or TSC/LAM than in patients with sporadic LAM

(Left) *Axial FLAIR MR shows typical brain parenchymal changes of TSC with subependymal nodules, streaky linear or wedge-shaped hyperintensities (which occur along radial migration lines from ventricle to cortex), and cortical and subcortical tubers.* **(Right)** *Anteroposterior radiograph of TSC/LAM patient shows increased lung volumes with diffuse bilateral symmetric reticular opacities and bilateral pleural effusions. Only 5% of TSC patients have clinically progressive lung disease.*





(Left) *Axial T1WI MR shows spinal manifestation of TSC with large focus of sclerosis seen as low signal involving the posterior vertebral body and extending to pedicle.* **(Right)** *Coronal oblique dynamic enhanced aortic MRA shows extensive aneurysmal dilatation of the thoracic and abdominal aorta, with long segments of stenosis. Intracranial aneurysms are the most common vascular anomaly in TSC besides angiomyolipomas.*





TERMINOLOGY

Abbreviations

- Subependymal giant cell astrocytoma (SGCA), subependymal nodule (SEN), lymphangioleiomyomatosis (LAM)

Synonyms

- Tuberous sclerosis complex (TSC), Bourneville-Pringle syndrome

Definitions

- Autosomal dominant disorder with multiorgan hamartomas

IMAGING

General Features

- Best diagnostic clue
  - Brain: Calcified subependymal nodules (hamartomas)
  - Spine: Multiple foci of bony sclerosis (bone islands)
- Size
  - Few mm to several cm
- Morphology
  - Round, ovoid, flame-shaped sclerosis

Radiographic Findings

- Radiography
  - Regions of bony sclerosis, which may be round, oval, or flame-shaped
    - Size from few mm to cm in diameter
    - Homogeneously dense or mottled
    - Should not expand bone or transgress bony margins
  - Undulating periosteal new bone occasionally seen

CT Findings

- NECT
  - Brain: SEN along caudothalamic groove > atrial > > temporal
    - 50% calcify (progressive after 1 year)
  - Spine: Multiple foci of bony sclerosis involving bodies and posterior elements with characteristics similar to plain films

MR Findings

- FLAIR
  - Brain: Streaky linear or wedge-shaped white matter hyperintensities (along radial migration lines from ventricle to cortex)
  - Spine: Low signal foci on all sequences in multiple bones
- T1WI C+
  - Brain
    - SEN enhancement more visible on MR than on CT
    - 30-80% enhance (enlarging SEN at foramen of Monro = SGCA)
    - 12% cortical/subependymal tubers enhance
  - Spine: Sclerotic or osteoblastic lesions do not enhance
- MRA
  - Stenosis, occlusions, and large vessel aneurysms
    - Intracranial, thoracic, and abdominal aortic aneurysms
    - Likely related to vascular hamartomas disrupting vasa vasorum

Imaging Recommendations

- Best imaging tool
  - CT for bony lesions
  - MR C+ for brain evaluation

Angiographic Findings

- Conventional DSA/MRA: Vascular dysplasia (rare = moyamoya, aneurysm)

DIFFERENTIAL DIAGNOSIS

Sporadic LAM

- Identification of 4 or more sclerotic bone lesions in patients with LAM markedly increases likelihood of associated TSC

Blastic Metastatic Disease

- Prostate > breast > colon; may be widespread

Paget Disease

- Thickened, irregular trabeculae

Pycnodysostosis

- Dense bones; dwarfism

Hypervitaminosis A or D

- Dense, brittle bones

Myelofibrosis

- Spine sclerosis, but peripheral bones spared

Osteopetrosis

- Thickened bone cortex, "bone within bone" appearance

Bone Island

- Few small foci of bony sclerosis

PATHOLOGY

General Features

- Etiology
  - Inherited tumor disorder with multiorgan hamartomas
- Genetics
  - Approximately 50% of TSC cases inherited
    - De novo: Spontaneous mutation/germline mosaicism
    - Autosomal dominant, high but variable penetrance
  - Mutations in TSC tumor suppressor genes cause abnormal cellular differentiation, proliferation
  - 2 distinct loci: *TSC1* (9q34.3) encodes "hamartin"; *TSC2* (16p13.3) encodes "tuberin"
- Associated abnormalities
  - Renal: Angiomyolipoma and cysts (40-80%)
  - Cardiac: Rhabdomyomas (50-65%); majority involute over time
  - Lung: Cystic lymphangiomyomatosis/fibrosis
  - Solid organs: Adenomas, leiomyomas
  - Skin: Ash-leaf spots (majority), including scalp/hair; facial angiofibromas; shagreen patches 20-35% postpubertal
  - Extremities: Subungual fibromas 15-20%; cystic bone lesions; undulating periosteal new bone formation
  - Ocular: "Giant drusen" (50%); retinal astrocytomas (which may regress)
  - Dental pitting permanent teeth in most adults with TSC

Staging, Grading, & Classification

- **Revised diagnostic criteria (Tuberous Sclerosis Alliance)**
  - Definite TSC = 2 major or 1 major + 2 minor
  - Probable TSC = 1 major and 1 minor
  - Possible TSC = 1 major or 2 or more minor
- **Major**
  - Cortical tuber
  - Subependymal nodule
  - Subependymal giant cell astrocytoma
  - Cardiac rhabdomyoma, single or multiple
  - Lymphangiomyomatosis
  - Renal angiomyolipoma
  - Facial angiofibromas or forehead plaque
  - Hypomelanotic macules (more than 3)
  - Ungual or periungual fibroma
  - Shagreen patch (connective tissue nevus)
  - Multiple retinal nodular hamartomas
- **Minor**
  - Bone cysts
    - Often in the phalanges of the hand
    - Not reported involving axial skeleton
  - Dental enamel pits
  - Hamartomatous rectal polyps
  - Cerebral white matter migration lines
  - Gingival fibromas
  - Nonrenal hamartoma
  - Retinal achromic patch
  - "Confetti" skin lesions
  - Multiple renal cysts
- **TSC/LAM vs. sporadic LAM**
  - May be indistinguishable on body CT studies
  - Both may have lung cysts, renal &/or hepatic angiomyolipomas, and lymphangioleiomyomas
  - Revised criteria notes that "when both lymphangiomyomatosis and renal angiomyolipomas are present, other features of TSC should be present before a definitive diagnosis is assigned"
  - Sclerotic bone lesions on CT scans are more common & numerous in patients with TSC or TSC/LAM than in patients with sporadic LAM

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Classic clinical triad
    - Facial angiofibromas (90%), mental retardation (MR) (50-80%), seizures (80-90%)
    - All 3 ("epiloia"): 30%
- Clinical profile
  - Seizures (infantile type spasms in very young), facial angiofibroma, hypopigmented skin lesions
    - Infant/toddler: Infantile spasms (65%), autism (50%) have poor prognosis
    - Infantile spasms occur before development of facial lesions, shagreen patches

Demographics

- Age
  - Diagnosed at any age
    - 1st year of life if infantile spasms or surveillance for positive family history
    - Child: Autistic-like behavior, mental retardation, seizures, or skin lesions
    - Adult diagnoses reported with demonstration of symptomatic SGCA on brain imaging

Treatment

- Treat seizures, infantile spasms respond to vigabatrin
- Resect isolated tubers if seizure focus or if able to identify seizure focus among many tubers
- SGCAs resected if obstructing foramen of Monro
- Oral rapamycin reported to cause regression of SGCA (may also spontaneously regress)

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Evaluating patients with LAM at CT, bone windows should be examined to search for sclerotic or lytic bone lesions

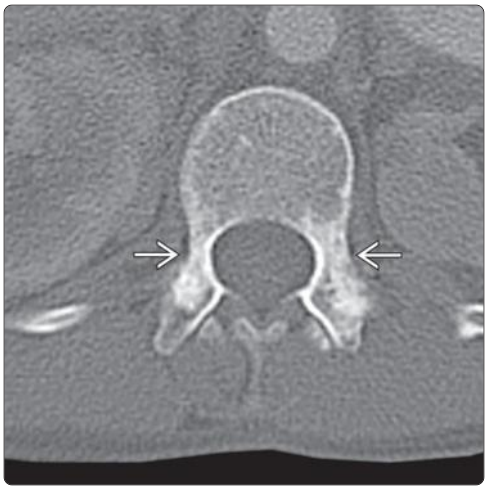
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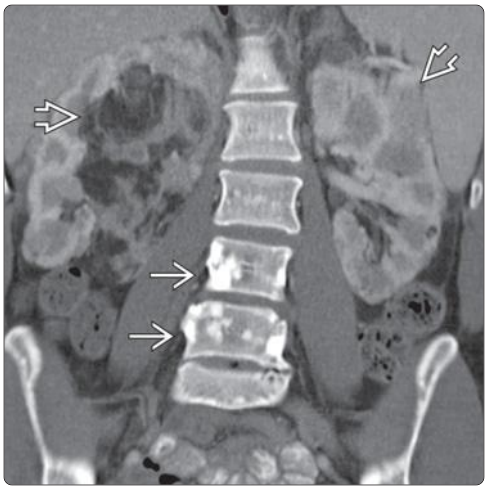
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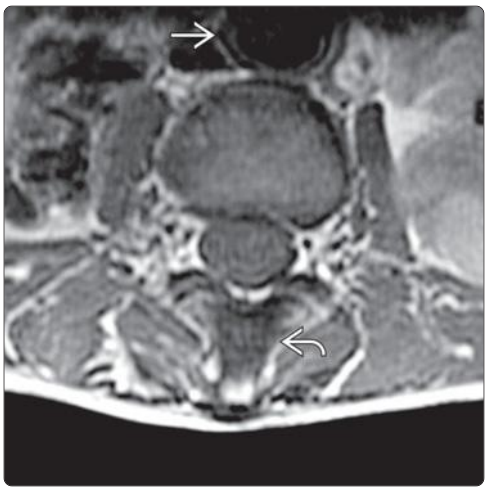
(Left) *Sagittal bone CT shows multiple foci of bony sclerosis involving the posterior aspects of the vertebral bodies in this patient with TSC.* **(Right)** *Coronal bone CT shows multiple sclerotic foci involving the iliac wings, sacral ala, and vertebral bodies. There is a large angiomyolipoma involving the right kidney with central fat attenuation and small AML in the superior left kidney. 75% of children with TSC have AMLs.*





(Left) *Axial bone CT shows focal bony sclerosis involving the posterior vertebral body and both pedicles. Spinal bony sclerosis in TSC has a predilection for the posterior aspects of the body/pedicles. Sclerosis presumably reflects hamartomas.* **(Right)** *Coronal bone CT shows focal bony sclerosis involving multiple vertebral bodies in this patient with TSC. Bilateral AMLs are present. 80% of patients with AML do not have TSC. Sporadic AMLs are usually single, unilateral, and in women.*





(Left) *Sagittal T1WI MR shows multiple focal areas of bony sclerosis as geographic low signal involving the posterior vertebral bodies and pedicles.* **(Right)** *Axial T1WI MR shows a bony hamartomatous sclerotic lesion involving the lamina with focal low signal. Note the thickening of the abdominal aortic wall in this child with extensive vascular dysplasia from TSC.*

Osteopetrosis

KEY FACTS

TERMINOLOGY

- "Marble bone disease"
- Heterogeneous grouping of hereditary osteoclast disorders

IMAGING

- Diffuse ↑ bone density, severe cortical thickening, bowing deformity of bones, frequent fractures
- ± extramedullary hematopoiesis

TOP DIFFERENTIAL DIAGNOSES

- Normal newborn
- Renal osteodystrophy
- Blastic metastatic disease
- Paget disease
- Pycnodysostosis
- Hypervitaminosis A or D
- Myelofibrosis

PATHOLOGY

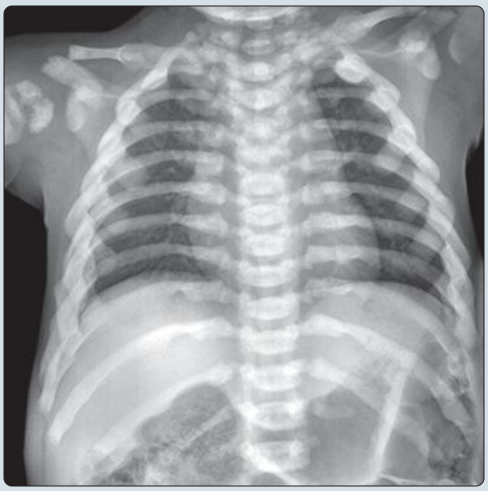
- Heterogeneous grouping of genetic defects with similar appearance
  - Infantile presentation ("congenita")
  - Intermediate presentation
  - Adult presentation ("tarda")
- All feature abnormal osteoclastic activity

CLINICAL ISSUES

- Fractures, pain, neurologic symptoms
- Growth disturbance in infantile forms

DIAGNOSTIC CHECKLIST

- Osteopetrosis in differential diagnosis list for dense bones
- "Bone-within-bone" appearance not pathognomonic; may also be seen in young children during growth spurts





(Left) *Anteroposterior radiograph of the thoracic spine and chest (infantile osteopetrosis) demonstrates marked diffuse sclerotic density within all bones. Note also abnormal thickened rib shape and multiple healing rib and clavicle fractures.* **(Right)** *Lateral radiograph of the cervical spine (infantile osteopetrosis) reveals marked sclerotic bone density. Note also C2 pars fracture ⇨ related to brittle bone with abnormal anterior angulation at C2/C3.*





(Left) *Sagittal T1WI MR (infantile osteopetrosis) shows universally abnormal low marrow signal intensity in all bones reflecting a combination of diffuse marrow replacement and sclerosis. The cerebellar tonsils ⇨ are ectopic, probably acquired secondary to bone changes in the posterior fossa.* **(Right)** *Sagittal T2WI MR (infantile osteopetrosis) reveals extensive abnormal low marrow signal intensity in all bones, including spine, skull base, sternum from marrow replacement, and osseous sclerosis.*

TERMINOLOGY

Synonyms

- "Marble bone disease"

Definitions

- Heterogeneous grouping of hereditary osteoclast disorders, multiple autosomal dominant and recessive forms

IMAGING

General Features

- Best diagnostic clue
  - Diffuse increase in bone density
- Location
  - Entire skeleton

Radiographic Findings

- Radiography
  - Bowing deformity of bones, frequent fractures
  - Infantile form: Dense bones, marrow space obliteration
  - Delayed form: Thickened bone cortex, "bone-within-bone" appearance

CT Findings

- Bone CT
  - Marked cortical thickening

MR Findings

- Sclerotic bone low signal intensity on T1WI, T2WI
- ± extramedullary hematopoiesis

Imaging Recommendations

- Best imaging tool
  - Radiography

DIFFERENTIAL DIAGNOSIS

Normal Newborn

- "Sandwich" appearance of sclerotic endplates

Renal Osteodystrophy

- "Rugger jersey" spine, soft tissue calcification

Blastic Metastatic Disease

- Prostate > breast > colon; may be widespread

Paget Disease

- Thickened, irregular trabeculae, "big vertebra"

Pycnodysostosis

- Dense bones, dwarfism, characteristic jaw deformity

Hypervitaminosis A or D

- Dense, brittle bones

Myelofibrosis

- Spine sclerosis with peripheral bone sparing

PATHOLOGY

General Features

- Etiology
  - Abnormal osteoclastic activity
- Genetics
  - Infantile types: Autosomal recessive
  - Intermediate, delayed types: Autosomal dominant
- Associated abnormalities
  - Renal tubular acidosis, hepatosplenomegaly, cranial neuropathy (cranial foraminal narrowing), cerebral calcifications ("marble brain syndrome")
- Heterogeneous grouping of genetic defects with similar appearance

Staging, Grading, & Classification

- Infantile presentation ("congenita")
- Intermediate presentation
- Adult presentation ("tarda")

Gross Pathologic & Surgical Features

- Dense, brittle bones with increased fracture risk

Microscopic Features

- Disorganized, dense bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Growth disturbance in infantile forms
- Other signs/symptoms
  - Fractures, pain, neurologic symptoms

Demographics

- Age
  - Severe forms present in infancy; mild forms may be asymptomatic until incidental detection
- Epidemiology
  - Up to 5.5/100,000 births

Natural History & Prognosis

- Infantile forms lethal
- Milder forms have ↑ fracture risk

Treatment

- Medical: Calcium restriction, calcitriol, steroids, parathyroid hormone
- Bone marrow transplantation (infantile form)
  - Radiographic abnormalities regress 1-2 years following successful transplantation

DIAGNOSTIC CHECKLIST

Consider

- Osteopetrosis in differential for dense bones

Image Interpretation Pearls

- "Bone-in-bone" appearance not pathognomonic; may also be seen in young children during growth spurts

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KEY FACTS

TERMINOLOGY

- Lipid storage disease
- Deficiency of lysosomal glucocerebrosidase → glucocerebroside deposition in macrophages (Gaucher cells)

IMAGING

- Hepatosplenomegaly
- Bone lesions
  - Vertebral endplate collapse, femoral Erlenmeyer flask deformity, AVN, pathologic fractures
- ↓ marrow signal 2° to glucocerebroside deposition

TOP DIFFERENTIAL DIAGNOSES

- Niemann-Pick disease (NPD)
- Sickle cell disease, spine
- Legg-Calvé-Perthes

PATHOLOGY

- Glucosylceramide accumulation in liver, spleen, bone marrow, lung macrophages (Gaucher cells)

CLINICAL ISSUES

- All 3 types
  - Hepatosplenomegaly
  - Acute bone crises 2° to infarcts
  - Pancytopenia
- Types 2, 3
  - Same as type 1 + developmental delay, oculomotor abnormalities
- Treatment with recombinant enzyme replacement therapy (ERT)

DIAGNOSTIC CHECKLIST

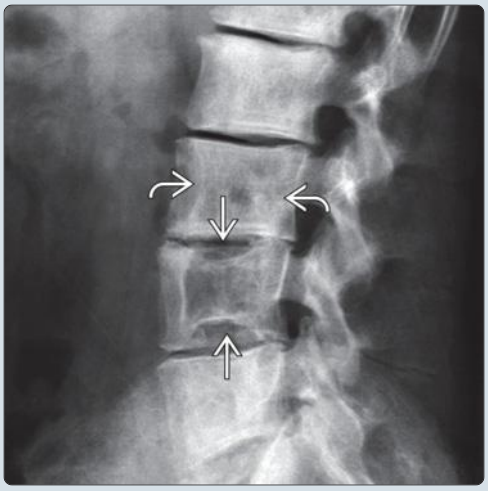
- Clinical response to ERT more rapid than radiographic response

(Left) *Sagittal T2WI MR demonstrates typical spine findings of untreated Gaucher disease, with endplate changes and mild "ballooning" of discs. Marrow signal loss is generally greater on T2WI than T1WI. No other spinal abnormalities were demonstrated. (Courtesy S. Blaser, MD.)* **(Right)** *Axial T1WI MR shows typical mild marrow T1 signal loss in a patient with untreated Gaucher disease. (Courtesy S. Blaser, MD.)*





(Left) *Lateral radiograph of the lumbar spine shows central sclerotic, serpentine lines representing bone infarcts surrounding regions of mottled density. Similar findings are demonstrated at other levels. The infarct has progressed to central L4 vertebral endplate collapse.* **(Right)** *Anteroposterior radiograph shows a widened metaphysis and cortical thinning in the distal femur; this is the classic "Erlenmeyer flask" deformity. Serpentine lines of bone sclerosis represent bone infarcts.*





TERMINOLOGY

Abbreviations

- Enzyme replacement therapy (ERT), avascular necrosis (AVN)

Definitions

- Lipid storage disease resulting from deficiency of lysosomal glucocerebrosidase → glucocerebroside deposition in macrophages (Gaucher cells)

IMAGING

General Features

- Best diagnostic clue
  - Erlenmeyer flask deformity of distal femora

Radiographic Findings

- Radiography
  - Vertebral endplate collapse, femoral Erlenmeyer flask deformity, AVN, pathologic fractures

CT Findings

- CECT
  - Hepatosplenomegaly

MR Findings

- T1WI
  - Early hip AVN
  - ↓ marrow signal 2° to glucocerebroside deposition
  - "Ballooning" of intervertebral discs 2° to vertebral endplate collapse
- T2WI
  - Same as T1WI, except marrow signal loss more conspicuous

DIFFERENTIAL DIAGNOSIS

Niemann-Pick Disease (NPD)

- Lipid storage disorder 2° to deficiency of lysosomal sphingomyelinase
- Failure to thrive, hepatosplenomegaly, rapidly progressive neurodegenerative decline → death by age 2-3 years

Sickle Cell Disease, Spine

- Hereditary hemoglobinopathy
- H-shaped vertebra, bone infarcts, osteomyelitis

Legg-Calvé-Perthes

- Hip avascular necrosis common
- No spinal manifestations

PATHOLOGY

General Features

- Etiology
  - 3 clinical subtypes characterized by abnormal glycolipid storage in Gaucher cells
    - Glucosylceramide accumulation in liver, spleen, lungs
    - Bone marrow infiltration
  - Neurologic abnormalities in types 2, 3 attributed to cytotoxic glycolipid accumulation in brain
- Genetics
  - Autosomal recessive
- Associated abnormalities
  - ↑ incidence of Parkinson disease in patients, carriers

Staging, Grading, & Classification

- 3 clinical subtypes
  - Type 1: Nonneuronopathic form
  - Type 2: Acute neuronopathic form
  - Type 3: Chronic neuronopathic form

Microscopic Features

- Gaucher cells (glycolipid-laden macrophages) pathologic hallmark
  - "Wrinkled paper" appearance, displaced nuclei 2° to intracytoplasmic substrate deposition

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Hepatosplenomegaly
  - Acute bone crises 2° to infarcts
- Other signs/symptoms
  - Pancytopenia
  - ± developmental delay, oculomotor abnormalities (types 2, 3)

Demographics

- Age
  - Type 1: Variable presentation age (children → elderly)
  - Types 2, 3: Presentation in early childhood
- Gender
  - M = F (all 3 types)
- Epidemiology
  - Type 1 more common in Eastern European Jewish populations
    - Most prevalent genetic disorder in Ashkenazi Jews
  - Type 3: Most common in Norrbottnian region of Sweden (1/50,000 population)

Treatment

- Enzyme replacement therapy (ERT) with recombinant enzyme (imiglucerase, Cerezyme)
- Hip replacement, other orthopedic procedures to treat skeletal disease

DIAGNOSTIC CHECKLIST

Consider

- Clinical response to ERT more rapid than radiographic response

SELECTED REFERENCES

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Ochronosis

KEY FACTS

TERMINOLOGY

- Synonyms: Alkaptonuria, alcaptonuria
- Abnormal pigmentation secondary to tissue deposition of homogentisic acid

IMAGING

- Severe degenerative intervertebral disc changes
- Linear intervertebral disc calcification
- Premature osteoarthritis of appendicular large joints

TOP DIFFERENTIAL DIAGNOSES

- Degenerative disc disease
- Ankylosing spondylitis
- Osteoarthritis
- Gout
- Hemodialysis arthropathy
- Calcium pyrophosphate deposition disease (CPPD)

PATHOLOGY

- Homogentisic acid oxidase enzyme deficiency
- Arthritis caused by tissue accumulation of homogentisic acid
  - Brittleness, degeneration of cartilage

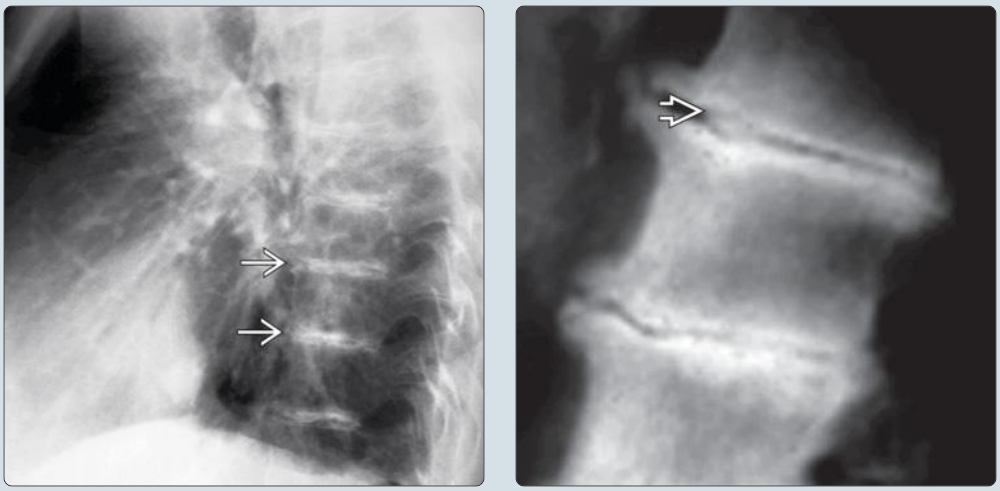
CLINICAL ISSUES

- Dark urine
- Back pain
- Accelerated premature degenerative arthritis of large peripheral joints (hips, knees, shoulders)

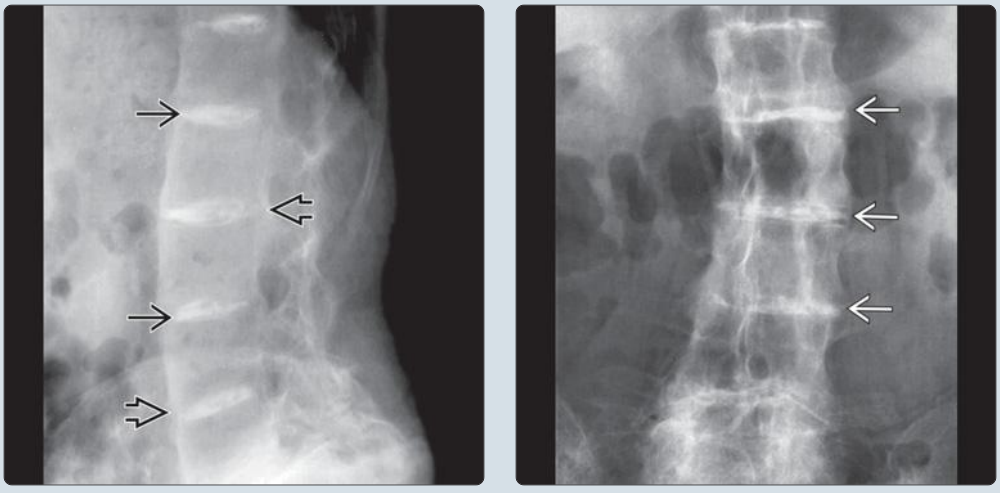
DIAGNOSTIC CHECKLIST

- Extremely rare; consider other causes of disc calcification, severe degenerative spine before ochronosis
- Consider urinalysis for homogentisic acid in younger patients with severe spinal degenerative changes

(Left) *Lateral radiograph of the thoracic spine demonstrates characteristic calcifications within the intervertebral discs at every level. Note 2 of the calcified discs ⇨, as well as the severe degenerative endplate changes present at every spinal level.* **(Right)** *Lateral radiograph of the spine depicts calcification within the intervertebral disc ⇨ in conjunction with severe degenerative intervertebral disc space narrowing.*



(Left) *Lateral radiography of the lumbar spine reveals marked narrowing and calcification of the intervertebral discs ⇨. The anterior and posterior longitudinal ligaments are also ossified ⇨, mimicking ankylosing spondylitis.* **(Right)** *Anteroposterior radiograph of the lumbar spine demonstrates marked narrowing and calcification of the intervertebral discs ⇨ in addition to convex left degenerative scoliotic curvature.*



TERMINOLOGY

Synonyms

- Alkaptonuria, alcaptonuria

Definitions

- Ochronosis: Absence of homogentisic acid oxidase enzyme → abnormal black tissue pigmentation
- Alkaptonuria: Detection of homogentisic acid in urine

IMAGING

General Features

- Best diagnostic clue
  - Calcified intervertebral discs, severe degenerative changes
- Location
  - Lumbar spine > thoracic > cervical

Radiographic Findings

- Radiography
  - Spinal axis
    - Calcified intervertebral discs, premature degenerative disc disease
    - Ankylosis of spine
    - Scoliosis, kyphosis
    - Sacroiliac osteoarthritis
  - Extraspinal findings
    - Premature osteoarthritis of large joints
    - Calcification of pubic symphysis fibrocartilage

CT Findings

- Bone CT
  - Calcified intervertebral discs
  - Premature degenerative disc disease

MR Findings

- T1WI
  - Severe chronic disc, marrow degenerative changes
- T2WI
  - Same as T1WI

DIFFERENTIAL DIAGNOSIS

Degenerative Disc Disease

- Discogenic vertebral body sclerosis
- Degenerated discs may calcify without ochronosis

Ankylosing Spondylitis

- Calcification of annular ligament (not disc) + facet ankylosis

Osteoarthritis

- May have identical appearance to ochronosis in appendicular skeleton large joints

Gout

- Rare in spine; positive cases show endplate destruction, tophi

Hemodialysis Arthropathy

- Deposition of crystals &/or amyloid
- Endplate destruction, peridiscal calcifications
- Usually involves cervical spine

Calcium Pyrophosphate Deposition Disease (CPPD)

- Calcification of hyaline and fibrocartilage, periarticular structures
- Often spares spine; most commonly involves hips, pubic symphysis, shoulder, knees, wrists

PATHOLOGY

General Features

- Etiology
  - Homogentisic acid oxidase enzyme deficiency
- Genetics
  - Autosomal recessive
- Associated abnormalities
  - Large joint arthritis
    - Accumulation of homogentisic acid → cartilage brittleness, degeneration
    - Not all alkaptonuria patients develop ochronotic arthritis
  - Cardiac ochronosis → accelerated coronary disease

Gross Pathologic & Surgical Features

- Black intervertebral disc material identified at surgery
- Brown or black pigmentation of skin, mucosa, sclerae, connective tissues, sweat, urine

Microscopic Features

- Intracellular pigment
- Cartilage pigmentation, degeneration

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Dark urine, back pain, large joint arthritis

Natural History & Prognosis

- Accelerated degenerative disease of spine, hips, knees, shoulders
- Coronary artery disease negatively impacts mortality

Treatment

- Nitisinone reduces homogentisic acid production
  - Long-term efficacy unknown

DIAGNOSTIC CHECKLIST

Consider

- Extremely rare; consider other causes of disc calcification, severe degenerative spine before ochronosis

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KEY FACTS

TERMINOLOGY

- Group of congenital musculoskeletal disorders with similar imaging findings

IMAGING

- Tremendous overlap in imaging appearance of connective tissue disorders
  - Rely on physical examination, other organ system findings to achieve specific diagnosis
  - Most common imaging findings are dural ectasia ± scoliosis, lateral meningocele(s)
- Lumbar spine, sacrum > thoracic

TOP DIFFERENTIAL DIAGNOSES

- Neurofibromatosis type 1
- Nerve sheath tumor
- Tarlov cyst
- Osteogenesis imperfecta
- Seronegative spondyloarthropathy

PATHOLOGY

- Variety of genetic defects in connective tissue disorders
  - Marfan, Ehlers-Danlos, and Stickler syndromes frequently share characteristics
- Associated anomalies commonly involve multiple organ systems

CLINICAL ISSUES

- High prevalence of back pain
- Lumbar dural ectasia often asymptomatic but may produce cord compression in thoracic spine
- Premature death from cardiac disease common
- Bone and joint injuries common because of hypermobility

DIAGNOSTIC CHECKLIST

- Check for aortic dilation on all thoracic spine MR and CT obtained for scoliosis
- "Marfanoid" body habitus not specific to Marfan syndrome

(Left) *Sagittal T2WI MR (Marfan syndrome) demonstrates relatively mild scalloping of the posterior lumbar vertebral bodies. Relatively mild findings are more common in younger patients with only mild dural ectasia &/or scoliosis.* **(Right)** *Axial T2WI MR (Marfan syndrome) reveals enlargement and remodeling of the osseous spinal canal in a heart-shaped configuration of the spinal canal frequently seen with dural ectasia.*





(Left) *Sagittal T2WI MR in a scoliotic patient with Marfan syndrome shows markedly increased AP canal diameter secondary to bone remodeling. There is scalloping of the posterior vertebral bodies and development of nerve root sleeve cysts ⇨ extending into the spinal canal.* **(Right)** *Coronal T2WI MR (Marfan syndrome) shows convex right scoliosis with widening of the transverse spinal canal related to pedicular thinning. Note formation of bilateral sacral nerve root sleeve cysts ⇨.*





TERMINOLOGY

Definitions

- Group of congenital musculoskeletal disorders with similar imaging findings
  - Most common types are Marfan, Ehlers-Danlos (EDS), Loey-Dietz syndrome, Stickler syndrome
  - Ehlers-Danlos also known as cutis hyperelastica

IMAGING

General Features

- Best diagnostic clue
  - Dural ectasia + scoliosis, lateral meningocele
- Location
  - Lumbar spine, sacrum > thoracic
- Size
  - Meningoceles, ectatic dura may be large

Radiographic Findings

- Radiography
  - Tremendous overlap in appearance of connective tissue disorders
    - Rely on physical examination and other organ system findings to achieve specific diagnosis
  - Some findings more suggestive of specific diagnosis
    - Marfan syndrome
      - Increased anterior translation C1-C2
      - Rotary subluxation C1-C2
      - Increased height of odontoid process
      - Basilar invagination
      - Biconcave vertebrae
    - Ehlers-Danlos syndrome
      - Platyspondyly
  - Dural ectasia
    - Scalloping posterior margins of vertebra
    - Widened interpediculate distance
    - Thinned, eroded pedicles
    - Erosion of sacrum
  - Scoliosis
    - Usually S-curve, same appearance as idiopathic scoliosis
    - Sometimes complex curve
    - Kyphosis often present
  - Osteopenia
    - Often occult in Marfan syndrome but documented by bone densitometry

CT Findings

- Bone CT
  - Dural ectasia
    - Width of dural sac at L5 > L4
    - Scalloping posterior margins of vertebral bodies
    - Anterior sacral meningocele
    - Perineural cysts
- CT myelogram
  - Delineates extent of ectasia, evaluates for spinal cord compression
  - Contrast diluted by large CSF space; may not have homogeneous filling of presacral meningocele or diverticula

MR Findings

- T1WI
  - Enlarged CSF spaces (dural ectasia)
  - Bone erosion, remodeling
  - Presacral meningocele may be difficult to see (mimicking bowel)
- T2WI
  - Enlarged CSF spaces
  - May have some heterogeneity of signal due to flow artifact
  - May see thoracic spinal cord compression 2° to diverticula or cysts posterior to spinal canal
  - Presacral meningocele better seen than on T1WI
- STIR
  - Similar to T2WI
- T1WI C+
  - No abnormal enhancement

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Include entire sacrum on sagittal, axial images

DIFFERENTIAL DIAGNOSIS

Neurofibromatosis Type 1

- Scoliosis
- Dural ectasia
- Nerve sheath tumors
- Gracile bones

Nerve Sheath Tumor

- Posterior vertebral body scalloping
- Thinning of pedicles
- Heterogeneous SI on T2WI
- Enhances with gadolinium

Tarlov Cyst

- Perineural cyst in sacrum
- Usually incidental finding on MR or CT

Osteogenesis Imperfecta

- Scoliosis
- Vertebral deformities and fractures
- Short body habitus

Seronegative Spondyloarthropathy

- Rarely see scalloping of posterior vertebral bodies
- Vertebral erosions progress to ankylosis

PATHOLOGY

General Features

- Etiology
  - Variety of genetic defects in connective tissue disorders
    - Marfan: Fibrillin-1 protein important component of elastic + nonelastic connective tissues
    - EDS I-III linked to collagen type 5 mutations (*COL5A1, COL5A2, COL5A3* genes)
    - EDS IV linked to collagen type 3 abnormality
    - EDS VII linked to collagen type 1 abnormality

- Stickler syndrome: Most involve mutations of *COL2A1* gene
- Genetics
  - EDS: Heterogeneous group of genetic-based disorders with different modes of inheritance
    - EDS I, II (classic type): Autosomal dominant, 80% of cases
    - EDS III (hypermobility type): Autosomal dominant
    - EDS IV (vascular type): Dominant and recessive forms
    - EDS VII (arthrochalasia type): Autosomal dominant
  - Marfan: Autosomal dominant with wide range of clinical severity
    - Mutations in fibrillin-1 gene (*FBN1*) on chromosome 15q21
    - > 90 different mutations of *FBN1*
    - > 25% of cases are new mutations
  - Stickler syndrome
    - Autosomal dominant, osteoarthritis, joint hypermobility, mild spondyloepiphyseal dysplasia
- Associated abnormalities
  - Marfan, Ehlers-Danlos, and Stickler syndromes share following characteristics
    - Aortic root dilation
    - Mitral valve prolapse
    - Unusually tall, slender body habitus
    - Joint hypermobility
    - Arachnodactyly
    - Blue sclerae
  - Marfan
    - Ectopia lentis
    - Retinal detachment
    - Pectus carinatum or excavatum
    - Protrusio acetabula
    - Leg length discrepancy
  - Ehlers-Danlos syndrome
    - Hyperelastic, fragile skin
    - Fragile blood vessels
    - Aneurysms of multiple vessels
    - Peripheral neuropathy
    - Triphalangeal thumbs
    - Radioulnar synostosis
    - Some types have ectasia, perforation of bowel
  - Homocystinuria
    - Scoliosis
    - Osteopenia
    - Arachnodactyly
    - Ligamentous laxity
    - Mental retardation
    - Ectopia lentis
  - Stickler syndrome
    - Midface hypoplasia, micrognathia

Staging, Grading, & Classification

- Many types of Ehlers-Danlos

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Dural ectasia often asymptomatic
  - Cord symptoms with thoracic dural ectasia
  - High prevalence of back pain
- Other signs/symptoms
  - Headaches, sciatica
- Clinical profile
  - "Marfanoid" body habitus not specific to Marfan syndrome

Demographics

- Age
  - Usually becomes apparent in early childhood
  - May not present until adulthood
- Gender
  - Marfan: M = F
  - Ehlers-Danlos: M > F
- Epidemiology
  - Ehlers-Danlos: 1:20,000
  - Marfan: 1:10,000 → 20,000

Natural History & Prognosis

- Premature death from cardiac disease common
  - Average life expectancy < 40 years if untreated
- Bone and joint injuries common because of hypermobility
  - Injuries can be reduced by lifestyle alteration to avoid overly stressing joints

Treatment

- Surgery for scoliosis has increased complication rate compared to idiopathic scoliosis
- Surgical treatment of aortic dilation permits near normal life expectancy
  - Lifespan for Marfan syndrome ↑ from 32 years in 1972 to 61 years in 1996
- Surgical treatment for Ehlers-Danlos complicated by skin and connective tissue fragility

DIAGNOSTIC CHECKLIST

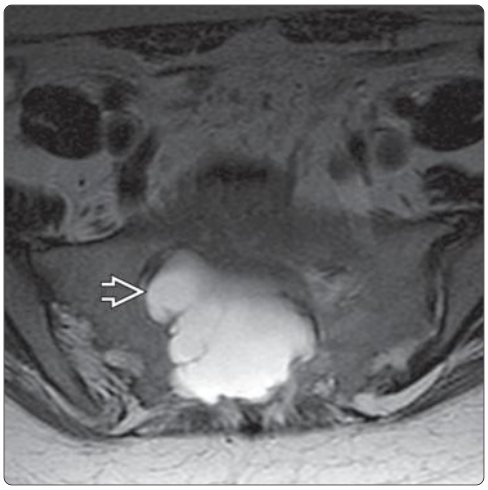
Consider

- Check for aortic dilation on all thoracic spine MR and CT obtained for scoliosis

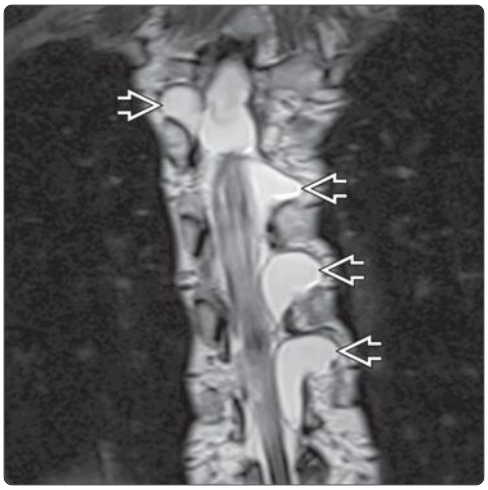
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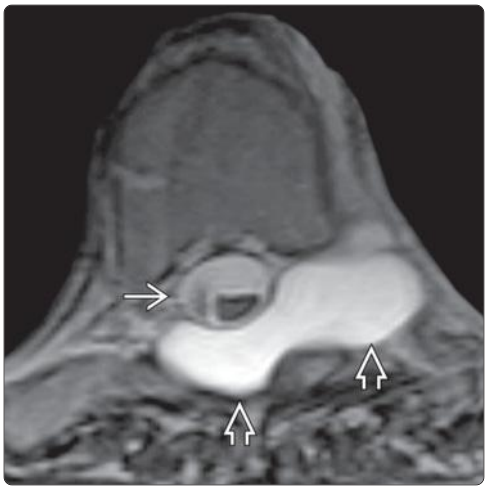
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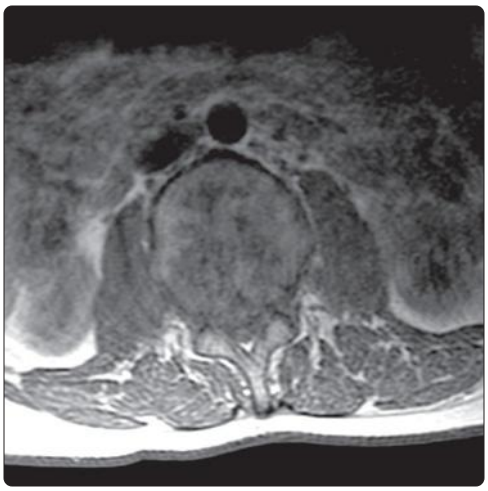
(Left) *Sagittal T2WI MR (Loey-Dietz syndrome) of the lumbosacral spine demonstrates posterior vertebral scalloping and patulous terminal thecal sac. Note in particular the marked remodeling and erosion of the sacrum.* **(Right)** *Axial T2WI MR (Loey-Dietz syndrome) confirms a markedly patulous thecal sac with dural diverticulum extending into the adjacent sacral neural foramen. Note also striking osseous remodeling of the posterior vertebral body and osseous spinal canal.*





(Left) *Coronal T2WI MR (Marfan syndrome) shows formation of bilateral dural diverticula (lateral meningoceles) extending into the neural foramina and displacing the spinal cord.* **(Right)** *Axial T2WI MR (Marfan syndrome) shows displacement of the thecal sac by a large lateral thoracic meningocele that has extended dorsally into the osseous spinal canal. Note significant osseous remodeling, which has enlarged the spinal canal and mitigated cyst compressive effects.*





(Left) *Sagittal T2WI MR (homocystinuria) demonstrates relatively mild vertebral remodeling of dural dysplasia as well as marked degenerative intervertebral disc changes that produce severe central spinal canal stenosis.* **(Right)** *Axial T1WI MR (homocystinuria) through a lumbar intervertebral disc level confirms severe central spinal stenosis secondary to marked degenerative changes that outweigh the effects of dural dysplasia, which generally produce widening of the canal.*

KEY FACTS

TERMINOLOGY

- Group of generalized skeletal dysplasias primarily involving vertebrae, proximal epiphyseal centers
- Affected patients demonstrate short trunk, neck, and limbs with normal hand and foot size

IMAGING

- Platyspondyly, vertebral hypoplasia & underossification, abnormal epiphyses
- ± pannus at C1/C2, os odontoideum
- Delayed ossification of capital femoral epiphysis → femoral head flattening, premature osteoarthritis

TOP DIFFERENTIAL DIAGNOSES

- Spondylometaphyseal dysplasia
- Spondyloepimetaphyseal dysplasia
- Multiple epiphyseal dysplasia

PATHOLOGY

- SED congenita
  - Abnormal type II collagen synthesis
- SED tarda
  - SEDL gene mutation (vesicular transport protein)

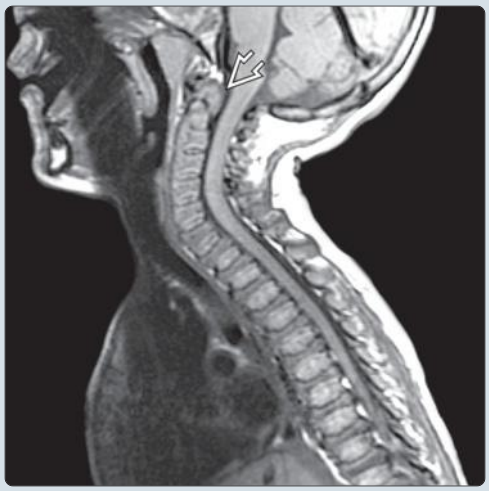
CLINICAL ISSUES

- SED congenita: Diagnosed at birth, short proximal limbs with normal hand, foot size
- SED tarda: Normal appearance at birth, subsequent identification of disproportionately short stature in adolescence or adulthood

DIAGNOSTIC CHECKLIST

- Consider SED congenita for platyspondyly, dysplastic epiphyses
- Consider SED tarda in adults with short trunk, early symmetric large joint osteoarthritis





(Left) *Lateral cervical radiograph (kyphosis, myelopathy) obtained in extension shows platyspondyly and delayed ossification status. The odontoid process is hypoplastic or underossified, rendering evaluation for atlantoaxial subluxation difficult.* **(Right)** *Sagittal T1WI MR (kyphosis, myelopathy) of the cervical spine reveals characteristic abnormal vertebral shape as well as a large odontoid process with delayed tip ossification. No spinal cord compression is demonstrated.*





(Left) *Lateral radiograph of the thoracolumbar spine (kyphosis, myelopathy) demonstrates characteristic platyspondyly in addition to focal kyphosis at the thoracolumbar junction secondary to a hypoplastic L2 vertebra.* **(Right)** *Sagittal T2WI MR (kyphosis, myelopathy) of the thoracolumbar spine confirms significant stenosis of the lower thoracic spinal canal producing focal spinal cord compression and abnormal spinal cord T2 hyperintensity.*

TERMINOLOGY

Abbreviations

- Spondyloepiphyseal dysplasia (SED)

Definitions

- Group of generalized skeletal dysplasias primarily involving vertebrae and proximal epiphyseal centers
  - Affected patients demonstrate short trunk, neck, and limbs with normal hand and foot size
- Classified as short-trunk dwarfing syndrome (trunk involvement > limbs)
- 2 major SED types primarily seen: SED congenita and SED tarda
  - Multiple rare uncommon subtypes also reported

IMAGING

General Features

- Best diagnostic clue
  - Platyspondyly, abnormal epiphyseal centers
- Location
  - Spine, large joints
- Morphology
  - Extent of osseous abnormalities diffusely distributed throughout skeleton
    - Severity of osseous abnormalities is variable

Imaging Recommendations

- Best imaging tool
  - Plain radiographs for screening
  - Multiplanar CT or MR for further evaluation, preoperative planning

Radiographic Findings

- Platyspondyly, vertebral hypoplasia, and underossification
- ± os odontoideum
- Delayed ossification of capital femoral epiphysis → femoral head flattening, premature osteoarthritis

CT Findings

- CECT
  - ± pannus at C1 to odontoid articulation
- Bone CT
  - Platyspondyly, vertebral hypoplasia, and underossification
  - ± os odontoideum

MR Findings

- T1WI
  - Same as bone CT
  - Improved detection of spinal cord and soft tissue (pannus) abnormalities
- T2WI
  - Same as T1WI
  - Better detection of spinal cord myelomalacia or syringohydromyelia

DIFFERENTIAL DIAGNOSIS

Spondylometaphyseal Dysplasia

- Generalized skeletal dysplasia featuring significant vertebral involvement
- Affects metaphyseal rather than epiphyseal portions of long bones

Spondyloepimetaphyseal Dysplasia

- Generalized skeletal dysplasia featuring significant vertebral involvement
- Affects both metaphyseal and epiphyseal regions of long bones

Multiple Epiphyseal Dysplasia

- Primarily affects (multiple) epiphyses
- Relatively mild clinical signs and symptoms
  - Joint pain; most commonly affects hips and knees
  - Early onset arthritis, abnormal waddling gait
- Some adults show mildly short stature, but most are normal height

PATHOLOGY

General Features

- Etiology
  - SED congenita
    - Abnormal synthesis of type 2 collagen (α-1 chain)
      - Type 2 collagen is primary matrix protein of physeal and epiphyseal cartilage, major component of nucleus pulposus, vitreous (eye)
      - Other skeletal dysplasias affected by collagen 2 include achondrogenesis type 2, hypochondrogenesis, Kniest dysplasia, Stickler dysplasia, autosomal forms of SED tarda, and spondylometaepiphyseal (Strudwick) dysplasia
  - SED tarda
    - *SEDL* gene mutation encoding a vesicular transport protein
- Genetics
  - SED congenita
    - *COL2A1* gene mutation, mapped to long arm of chromosome 12 (12q14.3)
      - Most cases result from random new mutation
    - Autosomal dominant (M = F)
    - Rare autosomal recessive cases reported
  - SED tarda
    - Genetically distinct from SED congenita
    - Most commonly X-linked recessive (M > > F)
    - Rare autosomal forms described
    - Caused by mutation in *SEDL* (SED late) gene (Xp22)
      - Encodes vesicular transport protein
- Associated abnormalities
  - Cervical instability, spinal curvature (scoliosis, kyphosis, lordosis), vision (myopia, retinal detachment), hearing loss, extremity (coxa vara, genu valgum, equinovarus foot, 2° large joint degenerative disease), nephrotic syndrome

Staging, Grading, & Classification

- International Nomenclature and Classification of Osteochondrodysplasias
  - SED congenita, tarda most common
  - Other rare SED variants
    - SED Maroteaux type: Musculoskeletal system only
    - SED tarda Toledo type: Musculoskeletal + corneal opacification

- SED tarda (Wynne-Davies): Progressive arthropathy (similar to juvenile inflammatory arthritis)
- SED with brachydactyly
- SED tarda Namaqualand type (NSED)
- Pseudo-Morquio disease
- Pseudoachondroplasia SED

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - SED congenita
    - Short proximal, middle limbs with normal hand, foot size
    - Normal intelligence
    - Cleft palate common
    - Short neck ± pectus carinatum
    - Scoliosis, kyphoscoliosis
    - Lumbar lordosis, hip flexion contractures in newborn
    - Coxa vara → characteristic waddling gait
  - SED tarda
    - Normal appearance at birth
      - Subsequently, disproportionate trunk shortening noticed
      - May be occult until adolescence or adulthood, when hip pain or scoliosis manifests
    - Normal achievement of motor, cognitive milestones
    - Epiphyseal involvement 1° in shoulders, hips, and knees
    - Hip findings may mimic bilateral Legg-Calvé-Perthes disease
- Other signs/symptoms
  - SED congenita
    - Buphthalmos, secondary glaucoma, strabismus
    - Myopia + retinal detachment (> 50%), cataracts
  - SED tarda
    - Atlantoaxial instability 2° to odontoid hypoplasia, os odontoideum
    - Nephrotic syndrome
- Clinical profile
  - SED congenita
    - Short proximal and middle limbs with normal hand and foot size, protruding abdomen, flat face, wide-set eyes, waddling gait
  - SED tarda
    - Normal appearance at birth
    - Subsequent development of disproportionately short stature in adolescence or adulthood with relatively short trunk, barrel-shaped chest

Demographics

- Age
  - SED congenita diagnosed at birth
  - SED tarda later onset, normal appearance at birth
    - Clinical manifestations recognized around puberty
    - Radiographic abnormalities apparent earlier
- Gender
  - SED congenita: M = F
  - SED tarda: M > > F
- Ethnicity
  - No racial predilection
- Epidemiology
  - Rare
    - International prevalence of SED congenita ~ 3.4 per million population
    - International incidence ~ 1 per 100,000 live births

Natural History & Prognosis

- No increased mortality
- Osteoarthritis nearly inevitable 2° complication of skeletal dysplasia

Treatment

- Address musculoskeletal deformities, secondary complications

DIAGNOSTIC CHECKLIST

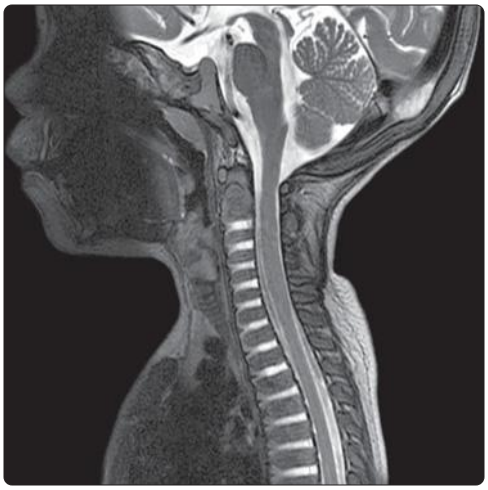
Consider

- SED congenita in context of platyspondyly, dysplastic epiphyses
- SED tarda in adults with short trunk and barrel chest in conjunction with early symmetric large joint osteoarthritis

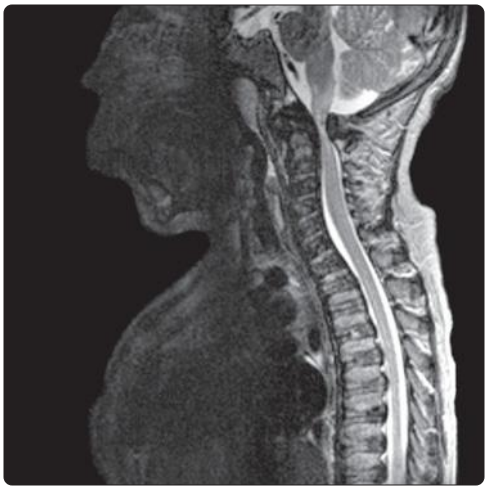
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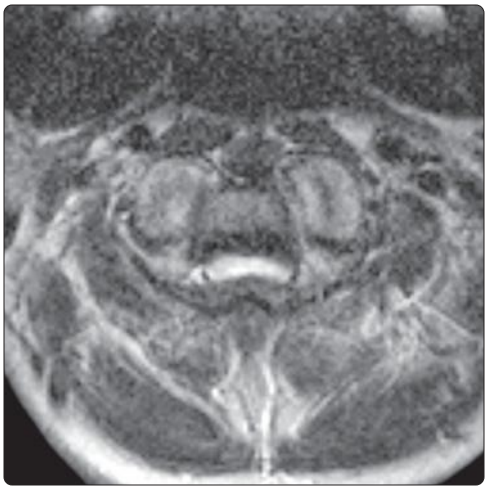
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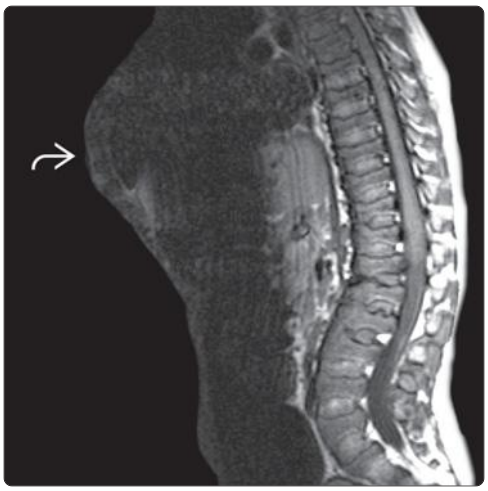


(Left) *Lateral cervical radiograph demonstrates mild platyspondyly and delayed ossification of the cervical vertebra and odontoid process (confirmed by MR, not shown). Initial interpretation of the radiograph mistakenly postulated that the incompletely ossified odontoid process represented an os odontoideum.* **(Right)** *Sagittal T2WI MR confirms platyspondyly and confirms absence of os odontoideum or craniovertebral junction subluxation.*





(Left) *Sagittal T2WI MR demonstrates marked platyspondyly. There is moderate narrowing of the central osseous spinal canal except at the foramen magnum, where there is severe stenosis with spinal cord compression and abnormal intramedullary T2 signal abnormality.* **(Right)** *Axial T2WI MR confirms severe anteroposterior (AP) canal stenosis and cord compression. Note relatively modest canal narrowing in transverse plane compared to severe AP canal stenosis.*





(Left) *Sagittal T1WI MR shows marked platyspondyly and relatively mild kyphosis at the thoracolumbar junction. Note also prominent pectus carinatum sternal configuration.* **(Right)** *Sagittal T2WI MR reveals marked platyspondyly with relatively mild kyphosis at the thoracolumbar junction. Clinically, this barrel-chested patient had a prominent pectus carinatum.*

Thanatophoric Dwarfism

KEY FACTS

TERMINOLOGY

- Thanatophoric dysplasia (TD)

IMAGING

- Severe dwarfism with narrow chest, short ribs
- Flattening of vertebral bodies (platyspondyly)
- Limb shortening
- "Cloverleaf" skull (variable)

TOP DIFFERENTIAL DIAGNOSES

- Achondroplasia
- Spondyloepiphyseal dysplasia
- Spondylocostal dysostosis
- Osteogenesis imperfecta congenita
- "Asphyxiating" thoracic dysplasia

PATHOLOGY

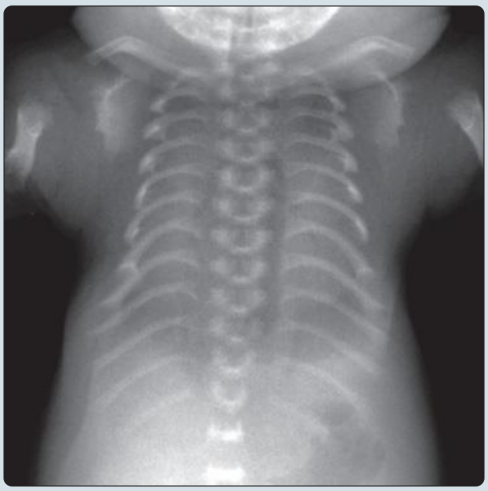
- Abnormal fibroblast growth factor receptor
- Allelic to same gene as achondroplasia and hypochondroplasia (chromosome 4p)
- Cerebral cortex malformations
  - Temporal lobe enlargement, deep temporal lobe sulci, hippocampal dysplasia

CLINICAL ISSUES

- Respiratory distress in neonate with severe dwarfism
- Most common lethal neonatal skeletal dysplasia
- Congenital presentation
- Usually fatal shortly after birth; may survive several years

DIAGNOSTIC CHECKLIST

- "Cloverleaf" skull can occur with wide variety of dysplasias or as isolated anomaly
- Recognition of characteristic axial, appendicular, and brain findings permits specific diagnosis





(Left) *Anteroposterior postmortem chest radiograph demonstrates characteristic TD skeletal deformities, including short ribs, narrow thorax, and flattened vertebral bodies.* **(Right)** *Lateral radiograph of the chest demonstrates characteristic short ribs and a hypoplastic sternum. Platyspondyly is also diffuse, although vertebral morphology may be difficult to visualize on a lateral chest radiograph because of obscuration by pulmonary opacities, as in this case.*





(Left) *Anteroposterior radiograph "babygram" shows a typical narrow thorax with resultant pulmonary hypoplasia and diffusely flattened vertebral bodies (representative level). The marked bowing deformities of both femurs has been called the "telephone receiver" configuration.* **(Right)** *Lateral radiograph of the thoracolumbar spine shows diffusely flattened vertebrae (platyspondyly, representative vertebra) and very short ribs resulting in a small thoracic cavity.*

TERMINOLOGY

Abbreviations

- Thanatophoric dysplasia (TD)

IMAGING

General Features

- Best diagnostic clue
  - Severe dwarfism with narrow chest, short ribs
- Morphology
  - Severe flattening of vertebral bodies (platyspondyly)

Radiographic Findings

- Radiography
  - Flattened vertebral bodies
  - Short ribs
  - Severe limb shortening
  - "Cloverleaf" skull (variable)

Ultrasonographic Findings

- Grayscale ultrasound
  - Diagnosis often possible in 2nd trimester
    - Small, narrow thorax
    - Enlarged skull ± "cloverleaf" appearance
    - Short femurs may be curved or straight

Imaging Recommendations

- Best imaging tool
  - Antenatal: Obstetrical ultrasound
  - Post partum: Plain radiography

DIFFERENTIAL DIAGNOSIS

Achondroplasia

- Similar deformities to TD but less severe

Spondyloepiphyseal Dysplasia

- Barrel-shaped thorax
- Delayed epiphyseal ossification

Spondylocostal Dysostosis

- Vertebral anomalies
- Thin, elongated bones of extremities

Osteogenesis Imperfecta Congenita

- Short, thick extremity bones
- Fractures detected at birth

"Asphyxiating" Thoracic Dysplasia

- Jeune syndrome
- Short ribs, narrow chest, "trident" appearance of iliac bones

PATHOLOGY

General Features

- Etiology
  - Abnormal fibroblast growth factor receptor (*FGFR3*)
- Genetics
  - Autosomal dominant
  - Most have de novo mutation in *FGFR3*
  - Allelic to same gene as achondroplasia, hypochondroplasia (chromosome 4p)
- Associated abnormalities
  - Maternal polyhydramnios
  - Hydrocephalus (foramen magnum stenosis)
  - Cerebral cortex malformations
  - Cervical spinal cord compression (spinal stenosis)
  - Genitourinary abnormalities

Staging, Grading, & Classification

- Type 1: Without "cloverleaf" skull
- Type 2: With "cloverleaf" skull

Gross Pathologic & Surgical Features

- Large skull, severe dwarfism

Microscopic Features

- Disruption of growth plate enchondral ossification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Respiratory distress in neonate with obvious severe dwarfism
- Other signs/symptoms
  - Short limbs

Demographics

- Gender
  - M = F
- Epidemiology
  - 1.7-3.8/100,000 births
  - Most common lethal neonatal skeletal dysplasia

Natural History & Prognosis

- Usually fatal shortly after birth; rarely may survive several years

Treatment

- Supportive care
- Tracheostomy
- Ventriculoperitoneal shunt for hydrocephalus

DIAGNOSTIC CHECKLIST

Consider

- "Cloverleaf" skull may occur with wide variety of dysplasias or as isolated anomaly
- Molecular genetic testing of *FGFR3* clinically available

Image Interpretation Pearls

- Recognition of characteristic axial, appendicular, and brain findings permits specific diagnosis

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PART I
SECTION 2

Scoliosis and Kyphosis



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Terminology

Scoliosis is considered to be present when there is a coronal plane curvature of the spine measuring at least 10°. However, treatment is not generally instituted unless the curvature is > 20-25°. The curvature may be balanced (returning to midline) or unbalanced. The vertebrae at the ends of the curve are designated the terminal (or end) vertebrae, while the apical vertebra is at the curve apex.

Curvatures are described by the side to which they deviate. A dextroscoliosis is convex to the right, with its apex to the right of midline. A levoscoliosis is convex to the left, with its apex to the left of midline.

Curvatures can be categorized as flexible (normalizing with lateral bending toward the side of the curve) or structural (failing to correct).

Most scoliotic curvatures are associated with abnormal curvature in the sagittal plane. These are described as kyphosis (apex dorsal) or lordosis (apex ventral).

Morphology of the Curvature

Scoliosis due to fracture, congenital anomaly, or infection typically has an angular configuration. Other causes of scoliosis tend to have a smooth curvature. Scoliosis most commonly involves the thoracic spine, followed by the thoracolumbar spine. In the past, curves were categorized as primary and secondary (compensatory), but it is often difficult to make the distinction and so these designations are no longer commonly used.

Measurement of Scoliosis

The Cobb method is most commonly used to measure scoliosis. The vertebrae at each end of the curve (the terminal vertebrae) are chosen. These are the endplates with the greatest deviation from the horizontal. The curvature is the angle between a line drawn along the superior endplate of superior terminal vertebra and a line along the inferior endplate of the inferior terminal vertebra. In severe curvatures, the endplates are often difficult to see. In that case, the inferior cortex of the pedicle can be used as the landmark for making the measurement. If measurements are made on hard copy radiographs, it is usually necessary to draw lines perpendicular to the endplates and measure the angle between the perpendicular lines. On most PACS, the measurements can be made directly from the endplates.

The Ferguson method is another way to measure scoliosis. In this method, lines are drawn from the center of the apical vertebra to the center of each terminal vertebrae. The angle of the scoliosis is the angle between these 2 lines.

Scoliosis is almost always associated with abnormal curvature in the sagittal plane. The most common finding is loss of normal thoracic kyphosis. The Cobb method can be used to determine sagittal plane deformity. Rotational deformity is often present but can only be grossly assessed on radiographs. It can be measured on CT scan by superimposing the apical and terminal vertebrae.

Normally, the T1 vertebra is centered over the L5 vertebra in both the coronal and sagittal planes. Coronal or sagittal plane imbalance can be measured as the horizontal distance between the center of the L5 vertebral body and a plumb line drawn through the center of the T1 vertebral body.

Rotational deformity is present in most types of scoliosis. This is difficult to measure. Measurement is most easily made by superimposing axial images of the terminal and apical vertebrae or by using 3D CT.

Risser Index

Since idiopathic scoliosis tends not to progress after skeletal maturity, it is useful to assess how close the young patient is to skeletal maturity. This is commonly assessed by the Risser method, based on the appearance of the apophysis of iliac wing. However, this has been shown to be less accurate than skeletal age assessed on hand radiographs.

- Apophysis not present = stage 0
- Apophysis covers lateral 25% of iliac wing = stage 1: Bone age 13 y, 8 mo (F); 14 y, 7 mo (M)
- Apophysis covers lateral 50% of iliac wing = stage 2: Bone age 14 y, 6 mo (F); 15 y, 7 mo (M)
- Apophysis covers 75% of iliac wing = stage 3: Bone age 15 y, 2 mo (F); 16 y, 2 mo (M)
- Apophysis covers entire iliac wing = stage 4: Bone age 16 y, 2 mo (F); 17 y, 0 mo (M)
- Apophysis fused = stage 5: Bone age 18 y, 1 mo (F); 18 y, 6 mo (M)

Radiology Reporting of Scoliosis

The radiology report should include measurements of all coronal and sagittal plane curvatures, using the method of Cobb. If bending or supine films are obtained, the radiologist should report the change in the curvature from the upright film. Spondylolysis is a frequent finding below the scoliosis, and routine search pattern should include evaluation for spondylolysis.

The radiographs should be evaluated for atypical findings: Are there any vertebral anomalies? Is the patient osteopenic and are fractures visible? Is the thoracic curve to the right (typical) or to the left (atypical)? Are the curves balanced in the coronal and sagittal planes, i.e., is the spine centered over L5? Are the ribs, cardiac silhouette, and paraspinous soft tissues normal?

Role of Advanced Imaging

MR or CT are performed when there is concern for an underlying abnormality, such as syrinx, tethered spinal cord, congenital bony abnormality, or tumor.

Suspicion of syrinx is raised when the thoracic curve is convex to the left or when the thoracic curve does not exhibit lordosis at its apex. Tethered cord usually presents in childhood but can be seen in early adulthood, where it presents with lower extremity symptoms or bowel and bladder dysfunction.

Treatment of Scoliosis

Juvenile scoliosis tends not to progress after skeletal maturity unless it is severe (> 50-60°). Therefore, minimal scoliosis is often treated just with observation, especially if the patient is near skeletal maturity. Mild degrees of scoliosis (< 40°) are treated with bracing, which has a high rate of success in compliant patients who wear a brace > 16 hours per day. More severe curvatures are usually treated with fusion, and techniques are constantly evolving. Currently, most surgeons use paired posterior rods and transpedicular screws. Alternatively, an anterior, transthoracic approach with fusion of the vertebral bodies and a lateral fusion rod is also common. Harrington rods, which have hooks at the ends but no pedicle screws, are rarely used today.

Postoperative Imaging

Imaging is performed after scoliosis surgery to evaluate the success of the operation. In general, it is irrelevant for the radiologist to name the brand of hardware used, and simple description is preferable. Postoperative radiographs must be evaluated first for correction of deformity in the coronal and sagittal planes. Then the hardware is assessed. On digital radiography, a thin lucent rim is typically seen surrounding screw threads and is not a sign of loosening. However, lucencies of > 1 mm suggest screw loosing. Screws may also back out from the vertebral body (in which case the screw heads are described as "proud"). Laminar hooks may become dislodged from the bone or detached from the fixation rod.

Hardware placed for correction of scoliosis provides rigid fixation as the bone fusion matures. If there is failure of bony fusion, the hardware will fail. CT scans with reformatted coronal and sagittal images are the most reliable method for assessing bony union. CT scan will show gradual coalescence of bone graft over a several month period postoperatively. By the end of 6 months, the graft should have developed a confluent mass with a well-defined cortex surrounding trabecular bone. Facet joint fusion is seen as loss of joint space and bridging trabeculae. Intervertebral body fusion will also show confluent bone.

If there is failure of bony fusion and consequent hardware failure, there may be loss of surgical correction of the scoliotic curvature. Crankshaft phenomenon results when the ends of the curvature are fixed, but the intermediate portions are not, and so the curve apex may continue to migrate.

When reviewing postoperative imaging, a useful pattern is to evaluate the hardware and bone at the fused levels and then look for abnormalities above and below the fused levels. In addition to hardware failure, common problems include adjacent segment degeneration, infection, and insufficiency fractures.

Imaging Protocols

Radiographs are performed to include the entire thoracic and lumbar spine. If the patient has unequal limb lengths, a lift is used under the shorter limb. Frontal radiographs are usually obtained PA instead of AP, in order to minimize radiation dose to the breasts.

CT scan should always be performed with reformatted images. Angled reformatted images and 3D reformations are often useful in assessment of severe curvatures.

Some physicians find it useful to obtain both SPECT and CT images of degenerative scoliosis. An area of arthritis on CT scan, which shows increased uptake on SPECT, is probably a pain generator.

MR can be difficult to interpret when scoliosis is severe. Angled axial images should be obtained based on both sagittal and coronal scout images and angled along the plane of the vertebral endplate on both scouts. Sagittal images should be angled along each segment of the curvature. The coronal plane is often the most useful for evaluating bony anomalies, spondylolysis, or degeneration of the discs and facet joints.

Differential Diagnosis

Idiopathic scoliosis is the most common type and may present in infancy, childhood, or adolescence. It may have a single curve or a balanced, S-shaped curve. The thoracic curvature is usually to the right. The vertebral bodies near the apex of the curve are often slightly wedged due to asymmetric stress during growth, but no anomalies are present.

Congenital scoliosis is scoliosis due to an abnormality of vertebral segmentation. It is associated with hemivertebrae, block (unsegmented) vertebrae, &/or fusions of the posterior elements. It often occurs as part of the VACTERL association: **V**ertebral anomalies, **a**nal atresia, **c**ardiac anomalies, **t**racheo-**e**sophageal fistula, **r**enal and other genitourinary anomalies, and **l**imb anomalies.

Congenital syndromes may cause scoliosis without vertebral anomalies. Collagen vascular disorders, neurofibromatosis, and osteogenesis imperfecta are the most common syndromes causing scoliosis. The curvatures are variable in appearance.

Neuromuscular scoliosis is seen in a variety of disorders, including muscular dystrophy and cerebral palsy. The curvature is most often a long, thoracolumbar C-shaped curve. It has a tendency to progress and can be very difficult to treat.

Osteoid osteoma and osteoblastoma may cause scoliosis. These tumors are very closely related, distinguished primarily by size and host response. Osteoid osteoma is < 1 cm in size and surrounded by dense reactive bone. Both tumors secrete prostaglandins, which stimulate a short-curve scoliosis, with the tumor on the concave side of the curvature. Prostaglandin release also often causes bone marrow edema involving bones not involved with tumor, pleural effusion, or soft tissue edema. Importantly, scoliosis due to osteoid osteoma/osteoblastoma is painful, while idiopathic scoliosis is not.

Adult scoliosis is divided into 3 types. First is degenerative scoliosis, which usually involves the lumbar spine. Degenerative scoliosis often develops above a surgical fusion, especially if there is a mild preexisting curvature above the fused levels. The second type is juvenile scoliosis, which continues to progress after skeletal maturity. The third type is scoliosis due to an underlying abnormality, such as limb length inequality, asymmetric variants at the lumbosacral junction, or osteoporosis.

Trauma, infection, failure of surgical instrumentation, or neuropathic arthropathy may cause scoliosis. In each of these cases the curvature is short, and the bony abnormality causing the scoliosis is visible on radiographs.

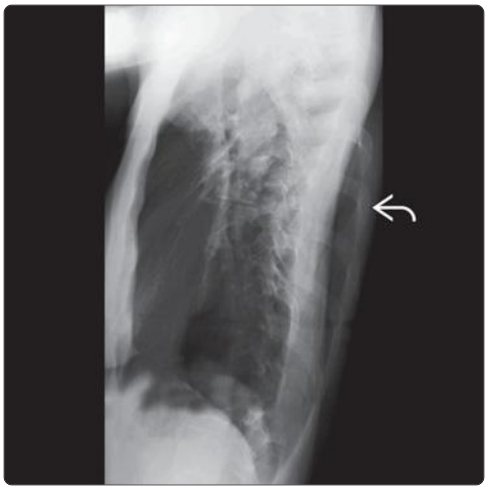
Chest wall anomalies or thoracic surgery in childhood are an uncommon cause of scoliosis. In the past, radiation therapy for Wilms tumor was an important cause of scoliosis, but changes in radiation therapy technique have rendered that a rare occurrence.

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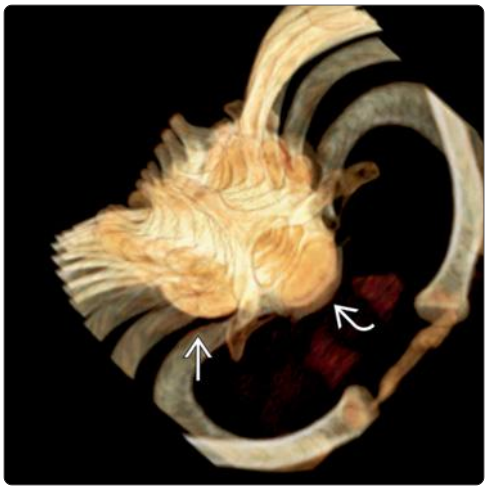
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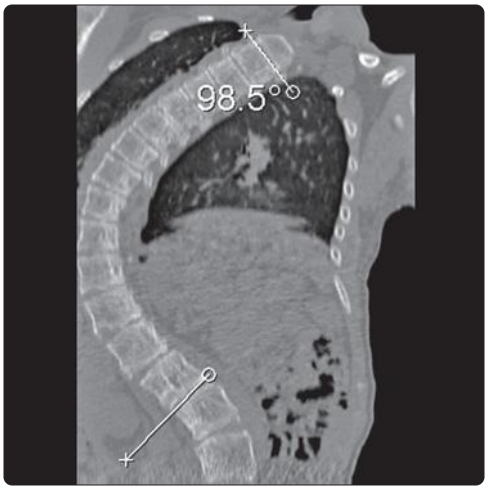
(Left) *Anteroposterior radiograph shows idiopathic thoracic dextroscoliosis. Apical vertebra ⇄ is the most displaced from the midline. Terminal vertebrae ⇄ show the greatest deviation of the endplates from the horizontal.* **(Right)** *Lateral radiograph in the same patient shows reversal of usual thoracic kyphosis. Rotational component of the scoliosis can be grossly assessed by the degree of rotation of the ribs ⇄.*





(Left) *Axial 3D reformation allows measurement of the rotational component of a scoliosis. This is the angle between the terminal vertebra ⇄ and the apical vertebra ⇄.* **(Right)** *Coronal CT reconstruction shows measurement of a severe neuromuscular scoliosis. The scoliotic curvature is the angle between the vertebrae with the greatest degree of inclination from the horizontal.*

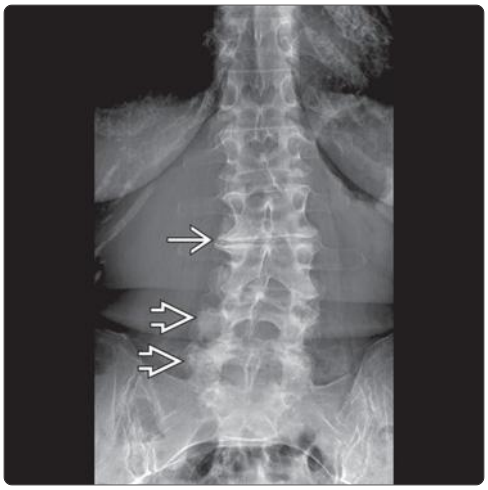




(Left) *Anteroposterior radiograph shows a single, long curve thoracolumbar scoliosis typical of neuromuscular scoliosis. No vertebral anomalies are present.* **(Right)** *Anteroposterior radiograph in the same patient shows only slight improvement in the curve when the patient bends to the right. Lateral bending films or supine films are used to assess the flexibility of a scoliotic curvature. Rigid curves are less amenable to reduction.*

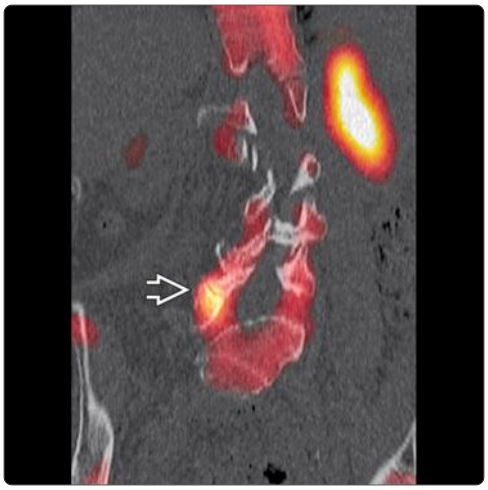


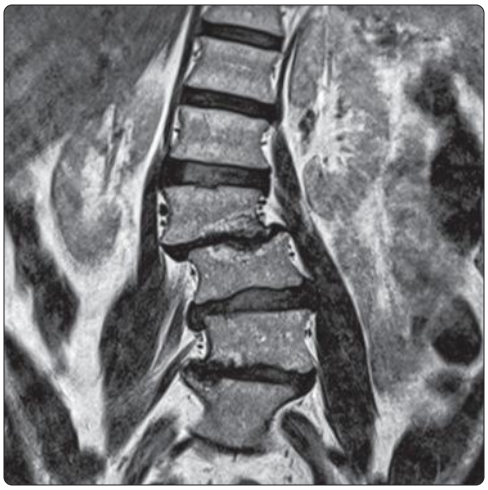




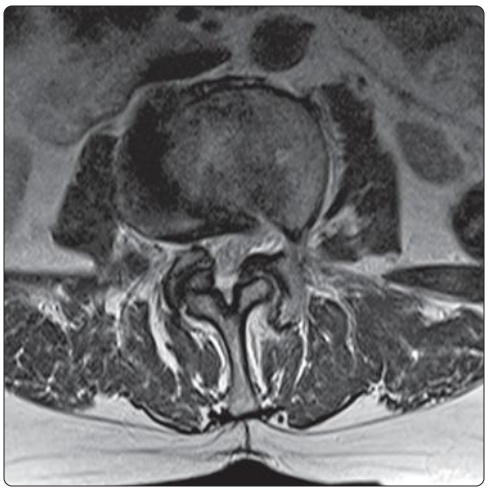


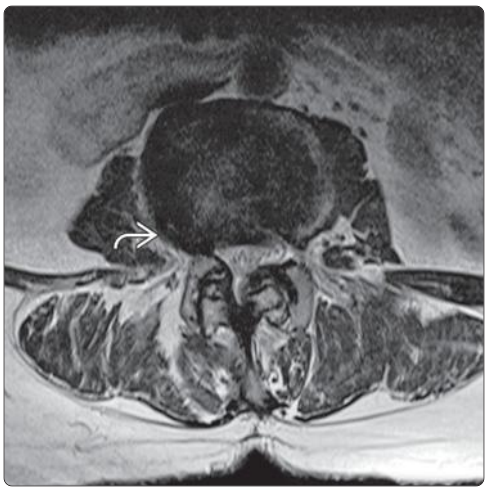
(Left) *Anteroposterior radiograph shows multilevel degenerative disc disease and only minimal leftward curvature in a 50-year-old woman. Disc disease is slightly asymmetric at the apex of the curve ⇨, and facet osteoarthritis ⇨ is more severe on the right (concave) side.* **(Right)** *Anteroposterior radiograph in the same patient 4 years later shows significant curve progression and progression of the asymmetric degenerative disc disease ⇨ and facet osteoarthritis.*





(Left) *Coronal CT reconstruction fused with SPECT bone scan shows abnormal uptake ⇨ at facet joint at concave aspect of a degenerative scoliosis. Fusion of CT and SPECT is sometimes useful to pinpoint pain generators and guide joint injections.* **(Right)** *Coronal T2WI MR is useful in evaluating disc and facet abnormalities in degenerative scoliosis. In addition, it can be used as a scout to set angled images through the intervertebral discs.*





(Left) *Axial T2WI MR at the L2-L3 level was angled based on sagittal scout, but not in reference to sagittal images. The disc is obliquely imaged, and even though the scoliosis was mild, the image appears distorted.* **(Right)** *Axial T2WI MR at the same level, but angled with the disc orientation on the coronal image, shows an annular fissure ⇨ on the right. Spinal stenosis appears more severe because the image is perpendicular to the canal rather than oblique as in the preceding image.*

Scoliosis

KEY FACTS

TERMINOLOGY

- General term for any lateral curvature of spine

IMAGING

- Lateral curvature of spine
  - Returns to midline at ends of curve
  - Rotational component common
- Most commonly thoracic or thoracolumbar
- Imaging approach
  - Radiography for initial diagnosis
  - Multiplanar MR to screen for bone, cord abnormalities
  - CT for surgical planning, complications

TOP DIFFERENTIAL DIAGNOSES

- Idiopathic scoliosis
- Neuromuscular scoliosis
- Congenital scoliosis
- Scoliosis due to congenital syndromes without vertebral anomalies
- Degenerative scoliosis
- Scoliosis due to infection
- Scoliosis due to tumor
- Scoliosis due to trauma
- Curvature related to limb length inequality
- Positional scoliosis

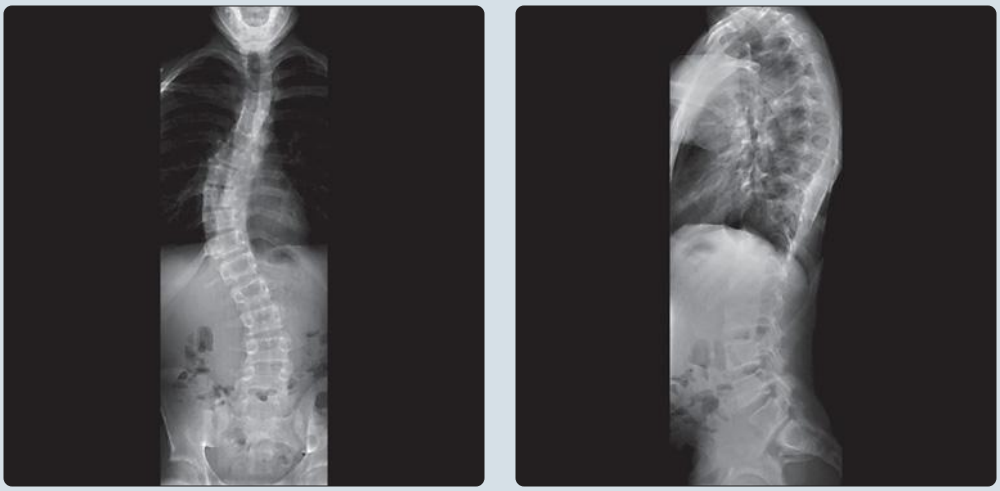
CLINICAL ISSUES

- Usually presents in childhood or adolescence
- Idiopathic scoliosis usually asymptomatic
  - Painful scoliosis indicates underlying abnormality
- Surgical fusion for rapidly progressive curves, curves > 40°

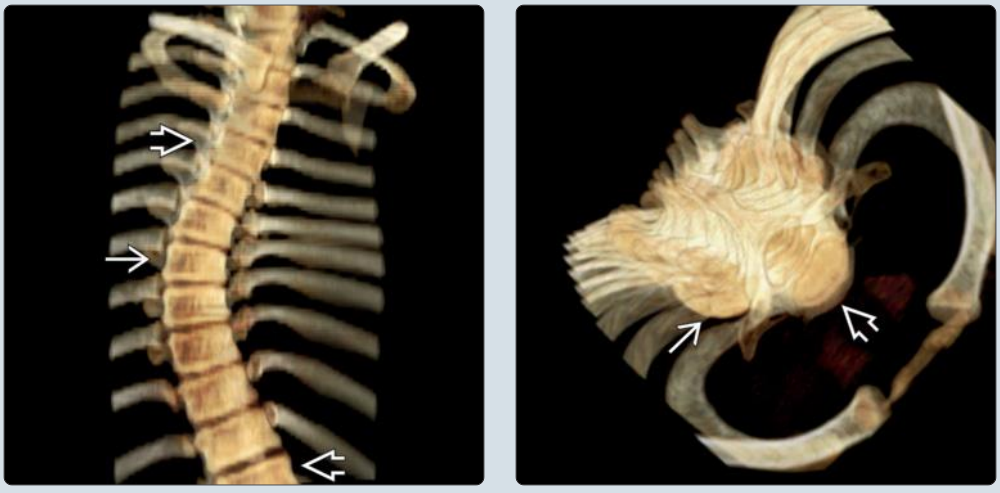
DIAGNOSTIC CHECKLIST

- Short-curve or painful scoliosis usually has underlying abnormalities
- Curve may progress rapidly, especially during growth spurts

(Left) *Anteroposterior radiograph in an idiopathic scoliosis patient shows the typical curve of idiopathic scoliosis that is convex to the right in the thoracic spine and convex to the left in the lumbar spine. No vertebral anomalies are present, excluding congenital scoliosis.* **(Right)** *Lateral radiograph (idiopathic scoliosis) depicts the rotational component of scoliosis, leading to posterior position of the ribs on the left. Thoracic kyphosis is diminished, another characteristic finding.*



(Left) *Anteroposterior 3D reformatted bone CT of idiopathic scoliosis shows rotation as well as scoliosis to be present on the apical vertebra ⇨ and terminal vertebrae ⇨.* **(Right)** *Axial bone CT 3D reformation demonstrates the rotation of the apical vertebra ⇨ compared to the terminal vertebrae ⇨. 3D CT is valuable in surgical planning to evaluate severity of rotation.*



TERMINOLOGY

Definitions

- General term for any lateral spine curvature
- Dextroscoliosis: Curve convex to right
- Levoscoliosis: Curve convex to left
- Kyphoscoliosis: Scoliosis with component of kyphosis
- Rotoscoliosis: Scoliosis which includes rotation of vertebrae
- S-curve scoliosis: 2 adjacent curves, 1 to right and 1 to left
- C-curve scoliosis: Single curve
- Terminal vertebra: Most superior or inferior vertebra included in curve
- Transitional vertebra: Vertebra between 2 curves
- Apical vertebra: Vertebra with greatest lateral displacement from midline
- Primary curvature: Curvature with greatest angulation
- Secondary or compensatory curvature: Smaller curve which balances primary curvature

IMAGING

General Features

- Best diagnostic clue
  - Lateral curvature of spine which returns to midline at ends of curve
- Location
  - Most commonly thoracic or thoracolumbar
- Size
  - Curve > 10°
  - May be > 90°
- Morphology
  - S-curve scoliosis
    - Idiopathic
    - Congenital
    - Syndromic
  - C-curve scoliosis
    - Neuromuscular
    - Neurofibromatosis
    - Scheuermann disease
    - Congenital
    - Syndromic
  - Short-curve scoliosis
    - Tumor
    - Trauma
    - Infection
    - Radiation
    - Congenital
    - Neuropathic

Radiographic Findings

- Radiography
  - Standing PA radiograph of full thoracic and lumbar spine on single cassette
    - PA projection gives lower radiation dose to breasts than AP
    - Lifts used to equalize limb lengths if needed
  - Method of Cobb is standard for measuring scoliosis
    - Draw lines parallel to endplates of terminal vertebrae
    - If endplates difficult to see, use pedicles as landmarks
    - Cobb angle is between terminal endplates
    - Can also measure angle between 2 lines drawn perpendicular to endplates
    - 2nd method is easier with small curves
  - Choosing correct vertebrae to measure scoliosis is critical to accuracy and monitoring
    - Terminal vertebra is one with greatest angle of endplate from horizontal
    - Rotoscoliosis: Terminal vertebra spinous process returns to midline
    - Interobserver variability 7-10°
  - Coned-down radiographs for better definition of vertebral abnormalities
  - Lateral radiograph to show sagittal plane abnormalities
    - Usually alters normal thoracic kyphosis, lumbar lordosis
  - Estimate rotational deformity by rib displacement on lateral radiograph

CT Findings

- Bone CT
  - Shows congenital bone anomalies, tumor, infection, postoperative complications
  - Not as useful for measuring curves compared to radiography

MR Findings

- Shows bone and spinal cord anomalies, syrinx, tumor, infection

Imaging Recommendations

- Best imaging tool
  - Radiography for initial diagnosis
- Protocol advice
  - Multiplanar MR to screen for bone, cord abnormalities
    - Coronal and sagittal T1WI and T2WI
    - Include craniocervical junction
    - Axial T2WI through areas of suspected abnormality
    - Axial T2WI through conus
  - CT for surgical planning
    - 1-3 mm multidetector CT with reformatted images
    - 3D helpful
  - CT for surgical complications
    - Thin, overlapping sections minimize artifact
    - Bone and soft tissue windows

DIFFERENTIAL DIAGNOSIS

Idiopathic Scoliosis

- Classic S-curve scoliosis

Neuromuscular Scoliosis

- Usually C-curve
- Neurologic disorders
- Muscular dystrophies

Congenital Scoliosis

- Morphology of curve highly variable
  - Focal short curves common
- Due to abnormal vertebral formation and segmentation

Scoliosis Due to Congenital Syndromes Without Vertebral Anomalies

- Often complex curvatures

- Neurofibromatosis
- Marfan
- Osteogenesis imperfecta
- Diastrophic dwarfism
- Ehlers-Danlos syndrome

Scoliosis Due to Infection

- Usually short curve
- Painful
- Systemic signs may be absent
- Pyogenic bacteria, tuberculosis, fungi

Scheuermann Disease

- 15% have scoliosis as well as kyphosis
- Scoliosis usually mild compared to kyphosis

Scoliosis Due to Tumor

- Short curve
- Painful
- Tumor may be occult on radiography
- Multiplanar MR most helpful for evaluation

Scoliosis Due to Trauma

- Usually see post-traumatic deformity
- Stress fracture may be occult cause

Scoliosis Due to Radiation

- Usually avoided today by radiation port placement
- Radiation to entire vertebra rather than portion preferred

Degenerative Scoliosis

- Develops in adults
- Degenerative disc disease, facet arthropathy seen
- Can also have secondary degenerative disease from idiopathic scoliosis

Neuropathic Spine

- Rapidly developing spine deformity
- Bony destruction seen on radiography

Compensatory Scoliosis

- Due to limb length inequality
- Can be diagnosed on frontal spine radiographs by position of iliac crests

Positional Scoliosis

- Poor positioning by radiology technologist
- Present on supine radiographs
- Resolves on upright radiographs

Iatrogenic Scoliosis

- Rib resection
- Level above lumbar fusion
- Hardware failure

PATHOLOGY

General Features

- Etiology
  - Variable, causes listed above
- Epidemiology
  - Common

Gross Pathologic & Surgical Features

- Deformity of trunk visible on physical examination

Staging, Grading, or Classification Criteria

- Etiology
- Direction of curve
- Severity of curve

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Visible truncal deformity
  - Idiopathic scoliosis asymptomatic
  - Painful scoliosis indicates underlying abnormality

Demographics

- Age
  - Usually presents in childhood or adolescence
- Gender
  - Idiopathic M:F = 1:7

Natural History & Prognosis

- Most scoliosis is mild
- May progress rapidly, especially during growth spurts
- Degenerative disc disease common
  - Greatest along concave aspect of scoliosis
- Severe scoliosis
  - Respiratory compromise
  - Neurologic symptoms
  - Instability

Treatment

- Options, risks, complications
  - Observation for minor curves
  - Bracing for curves > 25°
  - Fusion for rapidly progressive curves, curves > 40°

DIAGNOSTIC CHECKLIST

Consider

- Short-curve scoliosis usually has underlying abnormalities

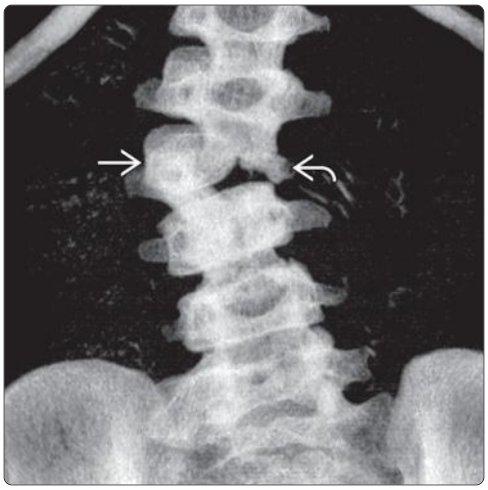
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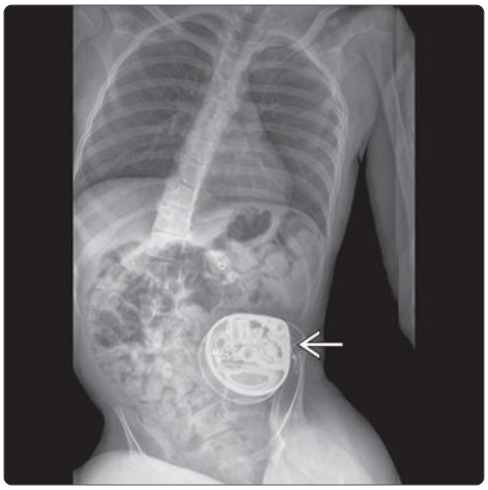
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(Left) *Coronal bone CT reveals multiple examples of vertebral segmentation failure. Several right-sided hemivertebra have failed to successfully segment from the adjacent vertebra, producing a jumble of malformed vertebra and multiple curve scoliosis.* **(Right)** *Frontal bone CT 3D surface rendering of congenital scoliosis, secondary to multiple vertebral anomalies (same patient), is helpful for fully characterizing contribution of the various anomalous vertebra to scoliosis and kyphosis to facilitate treatment planning.*





(Left) *Coronal 3D reformatted bone CT reveals a right L3 hemivertebra resulting in convex right congenital scoliosis. There are rudimentary left L3 pedicle and posterior elements.* **(Right)** *Anteroposterior radiograph of the thoracolumbar spine demonstrates C-shaped neuromuscular dextroscoliosis in a cerebral palsy patient. The intrathecal baclofen infusion pump and gastrostomy tube are clues to the diagnosis.*





(Left) *Anteroposterior radiograph depicts focal high cervical short curve scoliosis in a patient with neurofibromatosis type 1. Vascular clips in the right neck reflect the surgical site of prior neurofibroma resection.* **(Right)** *Anteroposterior radiograph demonstrates a large posterior paraspinal mass causing short segment, tumor-related scoliosis. Note osseous remodeling of the ipsilateral ribs.*

Kyphosis

KEY FACTS

TERMINOLOGY

- Spinal curvature secondary to vertebral anomalies
- Hemivertebra: Unilateral or anterior vertebral hypoplasia
- "Butterfly" vertebra: Central vertebral cleft due to failure of central vertebral body development
- "Fused" vertebrae: Embryological failure of segmentation rather than fusion
- Klippel-Feil syndrome: Multiple cervical segmentation anomalies

IMAGING

- Accentuated dorsal thoracic spine curvature on lateral image
- Look for vertebral anomalies in patient with scoliosis or kyphosis
- May have multiple kyphotic and scoliotic curves if multiple anomalies present

TOP DIFFERENTIAL DIAGNOSES

- Scheuermann kyphosis
- Idiopathic kyphosis
- Kyphosis or scoliosis due to syndromes
- Traumatic kyphosis
- Osteomyelitis, granulomatous

PATHOLOGY

- May be congenital or acquired
- Congenital abnormalities due either to failure of development &/or failure of segmentation

CLINICAL ISSUES

- May be isolated anomaly or associated with multisystem anomalies (VACTERL)

DIAGNOSTIC CHECKLIST

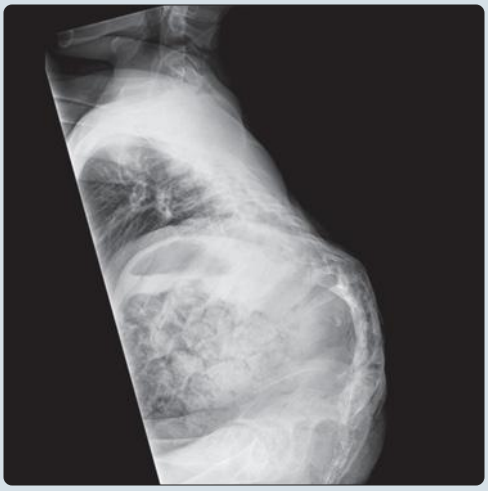
- Image entire spine (particularly in children) to exclude additional bone or cord abnormalities, Chiari 1 malformation

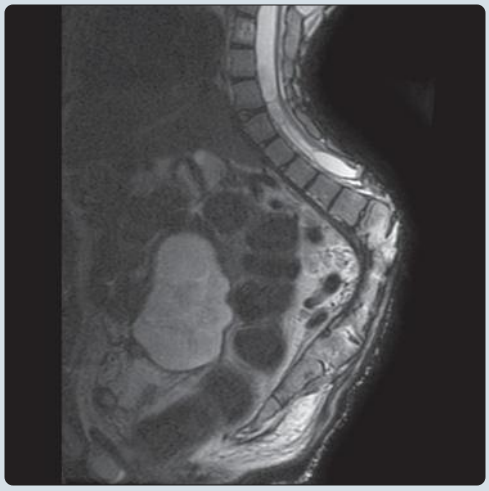
(Left) *Lateral radiograph demonstrates smoothly curved thoracic kyphosis, with premature upper thoracic degenerative disc disease, in this patient with degenerative kyphosis.* **(Right)** *Lateral chest radiograph (idiopathic kyphosis) reveals diffuse upper thoracic kyphosis with a round-back deformity. There is no underlying cause of kyphosis (e.g., Scheuermann disease, prior trauma, congenital anomaly, or infection).*





(Left) *Lateral radiograph (repaired high myelomeningocele, congenital vertebral segmentation failure with kyphosis) depicts severe focal lumbosacral kyphotic curvature. There is also posterior spinal dysraphism and segmentation failure of the lumbar and sacral vertebra associated with kyphosis.* **(Right)** *Sagittal T2WI MR (same patient) reveals distal spinal cord attenuation at the thoracolumbar junction myelomeningocele repair site. Note extensive congenital vertebral anomalies.*





TERMINOLOGY

Definitions

- Accentuated thoracic ± reduced normal cervical, lumbar lordotic spinal curvature
  - Spinal curvature secondary to vertebral anomalies, degenerative spine disease, or idiopathic causes

IMAGING

General Features

- Best diagnostic clue
  - Accentuated dorsal thoracic spine curvature on lateral image
  - Vertebral anomaly in patient with scoliosis or kyphosis
- Location
  - Most common in thoracic spine but can occur at any spinal level

Radiographic Findings

- Accentuated dorsal curvature of thoracic spine &/or reduced lordotic curvature of cervical, lumbar spine

CT Findings

- Improved visualization of anomalous vertebrae compared to radiographs
- Coronal and sagittal reformatted images essential
  - Helps confirm abnormal curvature and better demonstrates osseous abnormalities in osteopenic patients
  - 3D imaging helpful for surgical planning

MR Findings

- Similar to CT but better demonstrates spinal cord, soft tissues

Imaging Recommendations

- Best imaging tool
  - CT preferable for surgical planning in adults because of superior spatial resolution
  - Multiplanar MR best modality to evaluate full spine in children

DIFFERENTIAL DIAGNOSIS

Scheuermann Kyphosis

- Wedging of 3 or more vertebral bodies, undulation of endplates
- 15% have scoliosis as well as kyphosis

Idiopathic Kyphosis

- Vertebral anomalies absent

Kyphosis or Scoliosis Due to Syndromes

- Neurofibromatosis type 1
- Marfan syndrome
- Osteogenesis imperfecta
- Diastrophic dwarfism
- Ehlers-Danlos syndrome

Traumatic Kyphosis

- Short curve kyphosis, vertebral body deformity

Osteomyelitis, Granulomatous

- Paraspinous cold abscess, endplate destruction
- Kyphosis may be severe ("gibbus") deformity

PATHOLOGY

General Features

- Genetics
  - Sometimes associated with chromosomal abnormalities
- Associated abnormalities
  - Spinal cord abnormalities
    - Syringohydromyelia
    - Diastematomyelia
    - Tethered cord
  - Caudal regression
  - Component of VACTERL association

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Visible spinal axis deformity
- Clinical profile
  - May be isolated anomaly or associated with multisystem anomalies (VACTERL)

Demographics

- Age
  - Congenital kyphosis present at birth but may not be evident clinically until later in childhood or adolescence
  - Acquired kyphosis usually adolescence to adulthood
- Gender
  - M = F
- Epidemiology
  - Sporadic, relatively uncommon

Natural History & Prognosis

- Congenital kyphosis
  - Kyphosis tends to progress without treatment; fusion during childhood indicated
- Degenerative kyphosis frequently progressive
- Scheuermann kyphosis frequently progressive

Treatment

- Brace is of limited utility
- Fusion of congenital kyphosis to prevent paralysis

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Lateral radiography usually adequate to measure curvature
  - CT may be necessary in osteopenic patients
- Image entire spine to exclude additional bone or cord abnormalities, Chiari 1 malformation in children especially

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KEY FACTS

TERMINOLOGY

- Lateral curvature in spine due to degenerative disc and facet disease in older patients
- Deformity in a skeletally mature patient with Cobb angle of > 10° in coronal plane
- Predominance of lower lumbar curves

IMAGING

- Conventional standing full-length PA and lateral radiographs for monitoring curve progression
- Most common from L1 to L4
  - Lateral listhesis, vertebral rotation
  - Disc space loss, endplate sclerosis
  - Circumferential endplate spurring
  - Facet arthropathy
  - Spondylolisthesis, loss of lordosis

TOP DIFFERENTIAL DIAGNOSES

- Adult idiopathic scoliosis
- Neuromuscular
- Congenital scoliosis
- Post-traumatic, inflammatory, or neoplastic
- Dysplasias (NF1, Marfan)

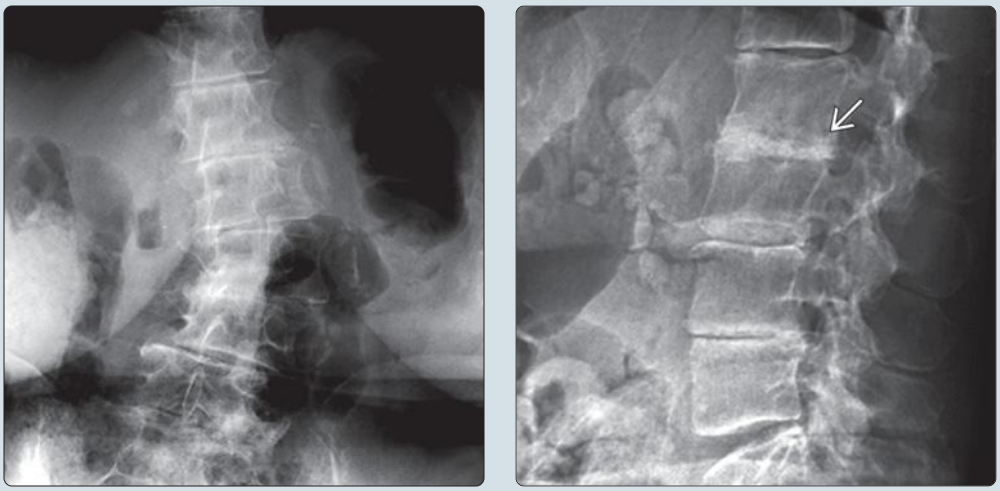
PATHOLOGY

- Asymmetric degenerative change at multiple levels
  - Asymmetric loading of spinal segments gives 3 dimensional deformity
  - Spondylolisthesis &/or rotatory listhesis

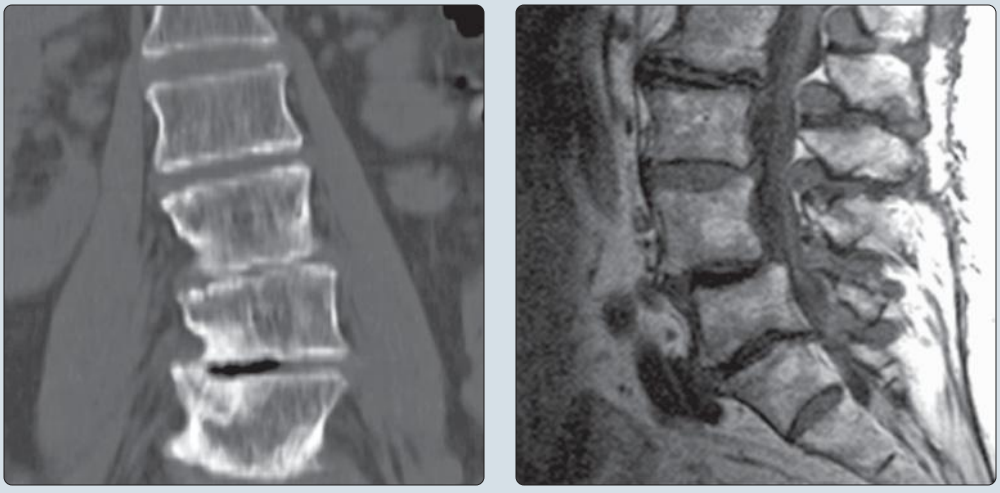
CLINICAL ISSUES

- Low back pain, radiculopathy
  - Pain worse with prolonged spinal extension
  - Radiculopathy not reliably relieved by flexion
- Risk factors for curve progression
  - Cobb angle > 30°
  - Lateral listhesis > 6 mm
  - More than 30° apical vertebral rotation

(Left) *Anteroposterior radiography shows lumbar rotatory levoscoliosis with associated multilevel marked disc height loss and endplate sclerosis. There is left lateral listhesis of L4 on L5.* **(Right)** *Lateral radiography shows loss of lumbar lordosis with multilevel disc space height loss and degenerative endplate bony eburnation, worst at L2-L3 ➡.*



(Left) *Coronal reformation of nonenhanced lumbar spine CT shows mild levoscoliosis at L4-L5 with vacuum disc phenomenon and right-sided endplate sclerosis. There is disc height loss at L3-L4 and L4-L5.* **(Right)** *Sagittal T1WI MR shows grade 1 anterolisthesis of L4 on L5 with severe disc degeneration and vacuum phenomenon. Degenerative disc and endplate disease is also present at L2-L3 and L5-S1. There is severe central stenosis at L4-L5 and L5-S1.*



TERMINOLOGY

Abbreviations

- "De novo" scoliosis

Definitions

- Lateral curvature in spine due to degenerative disc and facet disease in older patients
- Deformity in a skeletally mature patient with Cobb angle of > 10° in coronal plane
  - Advanced degenerative changes and predominance of lower lumbar curves

IMAGING

General Features

- Best diagnostic clue
  - Lateral curvature in spine with associated degenerative changes
- Location
  - T12 to L5
    - Most common from L1 to L4
  - Apex of curvature most common at L2-L3 interspace
- Size
  - Curvature ranging from 14-80°
    - Mean: 24-43°
  - Average curvature per segment < 10°
- Morphology
  - Moderate curvature over a short spinal segment
  - Levo or dextroscoliosis
    - Levoscoliosis (57-68%)

Radiographic Findings

- Radiography
  - Lateral listhesis
    - In > 3/4 of patients
    - Average translation: < 10 mm
  - Vertebral rotation
    - Most common grade 2
  - Disc space loss
    - Worse on concave side
    - Vacuum disc phenomenon
  - Endplate sclerosis
  - Circumferential endplate spurring
    - Most significant on concave side
  - Facet arthropathy
    - Concave > convex side
  - Spondylolisthesis
    - In > 50% of patients
    - L4-L5 most common
    - Usually not beyond grade 1

CT Findings

- Bone CT
  - Same findings as radiography
  - Better osseous detail
    - Sagittal and coronal reformation
    - Facet arthropathy well delineated
  - Similar accuracy compared to CT myelography and MR

MR Findings

- Same findings as CT myelography
- Underestimates nerve root compression in lateral recess compared to conventional myelography
  - 28-29% vs. 5-7%
- Disc herniation well visualized
  - Anterior, posterior, foraminal, far lateral
- Excellent characterization of central canal and neural foramina

Nonvascular Interventions

- Myelography
  - Post-myelographic CT improves sensitivity and specificity
    - Especially with multidetector row helical CT
  - Variable central stenosis
    - Disc herniation, endplate spurring, spondylolisthesis, ligamentum flavum hypertrophy
  - Subarticular recess stenosis
    - May be asymmetric
    - Concave side of curvature
    - Ligamentum flavum hypertrophy and facet arthropathy
  - Neural foraminal stenosis
    - Worse at concave side
    - Foraminal disc herniation, endplate spurring, or disc space loss

Nuclear Medicine Findings

- Bone scan
  - Spinal curvature
  - Asymmetric uptake of radiotracer at endplates and facets

Other Modality Findings

- Axial loading during CT myelography or MR may provide additional information
  - Worsening disc herniation
  - Worsening central canal, lateral recess, or neural foraminal narrowing

Imaging Recommendations

- Best imaging tool
  - Conventional standing full-length PA and lateral radiographs for monitoring curve progression
  - Noncontrast-enhanced MR
    - Similar sensitivity and specificity as CT myelography
    - Noninvasive
- Protocol advice
  - Include coronal T1 on lumbar MR
    - Characterization of scoliosis
    - Additional information on lateral endplate spurring, disc bulge, and far lateral nerve root

DIFFERENTIAL DIAGNOSIS

Adult Idiopathic Scoliosis

- Juvenile idiopathic scoliosis present in adults
- Most common
  - 85% of all scoliosis
- Strongly familial: 80%
  - Female predilection: 7-9:1
- Long, smooth S-shaped spinal curvature

- Thoracic and lumbar component

Neuromuscular

- Cerebral palsy, poliomyelitis, muscular dystrophy, syringohydromyelia, cord neoplasm
- Single long smooth curve

Congenital Scoliosis

- Failure of vertebral formation or segmentation
  - Wedge vertebra, hemivertebra, pedicle bar, block vertebra
- Acute curvature
- Associated with spinal dysraphic, genitourinary, or cardiac anomalies

Post-Traumatic, Inflammatory, or Neoplastic

- Juvenile rheumatoid arthritis
- Tuberculosis
- Radiation therapy
- Osteoid osteoma

Dysplasias

- Neurofibromatosis type 1 (NF1)
- Marfan syndrome
- Ehlers-Danlos syndrome
- Posterior vertebral scalloping
  - Dural ectasia
- High thoracic acute curvature in NF1

PATHOLOGY

General Features

- Etiology
  - Asymmetric degenerative change of discs and facets at multiple levels
    - Asymmetric loading of spinal segments ultimately gives a 3D deformity
      - Asymmetric loading with degeneration continues the cycle, which enhances the curvature
      - Spondylolisthesis &/or rotatory listhesis
      - Osteophyte formation give canal and lateral recess narrowing
- Associated abnormalities
  - Osteoarthritis
    - Hip and knee, may be unilateral
  - Spondylosis
    - Cervical and thoracic
  - No association with osteoporosis
- Degenerative disease of spine

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back pain
  - Other signs/symptoms
    - Radiculopathy
    - Neurogenic claudication
    - Deformity and waist asymmetry
    - Gait disturbance
- Clinical profile
  - Pain worse with prolonged spinal extension
  - Radiculopathy not reliably relieved by flexion
    - In contrast to spinal stenosis without scoliosis

Demographics

- Age
  - 6th decade
- Gender
  - M < F
- Ethnicity
  - More common in whites
- Epidemiology
  - Prevalence around 6%

Natural History & Prognosis

- Risk factors for curve progression
  - Cobb angle > 30°
  - Lateral listhesis > 6 mm
  - More than 30° apical vertebral rotation
  - Degenerative disease at lumbosacral junction
- May progress rapidly at 3° or more per year

Treatment

- Nonoperative
  - Physical therapy
  - NSAIDs
  - Epidural injection
  - Brace
- Surgery
  - Preserves neural function and prevents instability
  - Decompression, alone or with fusion
    - Posterior, combined anterior/posterior, or lateral approaches
  - Extreme lateral interbody fusion treating degenerative scoliosis results in lower complication rates and equivalent outcomes to more conventional approaches

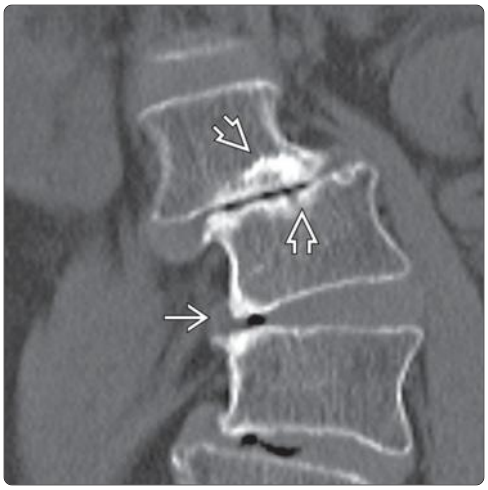
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Presence of extensive degenerative disc disease and facet arthropathy indicates degenerative scoliosis

SELECTED REFERENCES

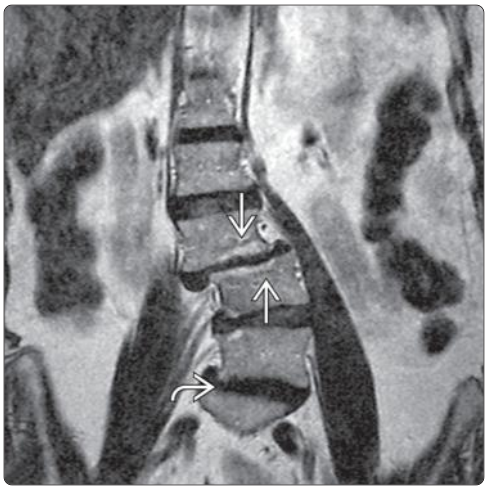
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(Left) *Coronal bone CT shows discogenic sclerosis on the left at L2-L3, and on the right at L3-L4. The asymmetric distribution is due to altered stresses in this patient with scoliosis. Vacuum phenomena are evident at multiple discs.* **(Right)** *Coronal NECT shows severe multilevel disc degeneration with loss of disc height, bony eburnation, and vacuum phenomenon at L5-S1, L3-L4, and L2-L3. Note the pronounced discogenic sclerosis at L5-S1 and convex left degenerative scoliosis.*





(Left) *Coronal T1WI MR shows moderate levoscoliosis centered at L3-L4. There is right lateral listhesis of L1 on L2, and L2 on L3. Disc height loss and degenerative endplate changes are present.* **(Right)** *Coronal T2WI MR shows fatty marrow adjacent to degenerated discs. Note the diffuse irregularity of the vertebral endplates and lateral listhesis due to the severe disc degeneration.*





(Left) *Coronal CT following extreme lateral interbody fusion in a patient with degenerative scoliosis shows the typical appearance of the interbody fusion graft with 3 separate radiopaque marker locations. Access to the disc is obtained through the psoas muscle.* **(Right)** *Coronal T2 MR shows normal imaging appearance of the postoperative change from an extreme lateral interbody fusion (XLIF). The interbody fusion graft is low signal.*

Flat Back Syndrome

KEY FACTS

TERMINOLOGY

- Loss of lumbar lordosis after lumbar or scoliosis distraction instrumentation, vertebral fracture, ankylosing spondylitis, degenerative disease
  - Sagittal imbalance with center of gravity of head anterior to sacrum

IMAGING

- Thoracolumbar or lumbar instrumentation with straightening of lumbar spine
- Full-length lateral radiograph obtained with patient's knees and hips fully extended
- Evaluate radiographs over time

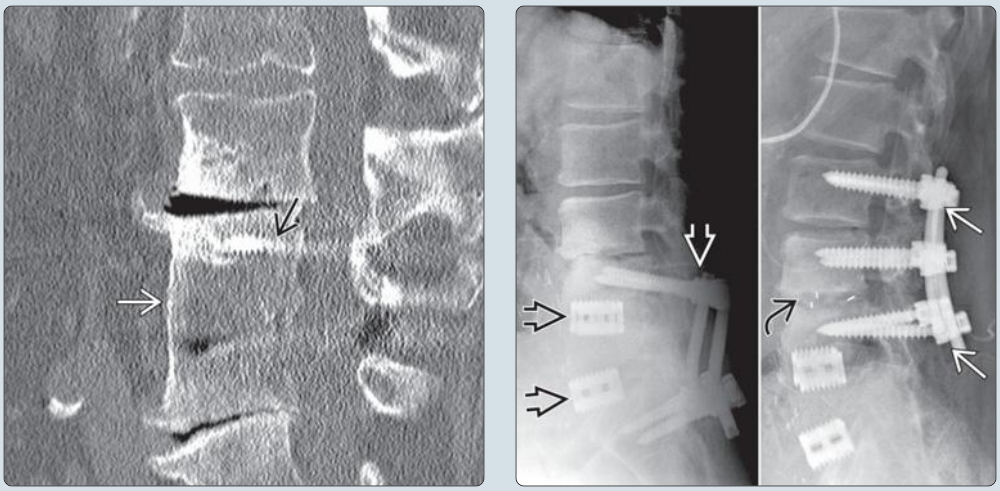
CLINICAL ISSUES

- Common at presentation
  - Inability to stand upright and forward inclination of trunk
  - Muscular pain in lower cervical area or upper back
  - Pain in lower extremities
- Single biggest risk factor is lower lumbar spine or sacrum distraction instrumentation
  - Magnitude of loss of lordosis increases the farther distally the instrumentation is placed
- Other causes: Pseudoarthrosis resulting in loss of sagittal plane correction, fixed thoracic hyperkyphosis, hip flexion contractures, preexisting thoracolumbar kyphosis
- Goal of surgery: Restore sagittal balance
  - Osteotomies
  - Anterior-posterior surgery
- Prevention by avoiding distraction instrumentation

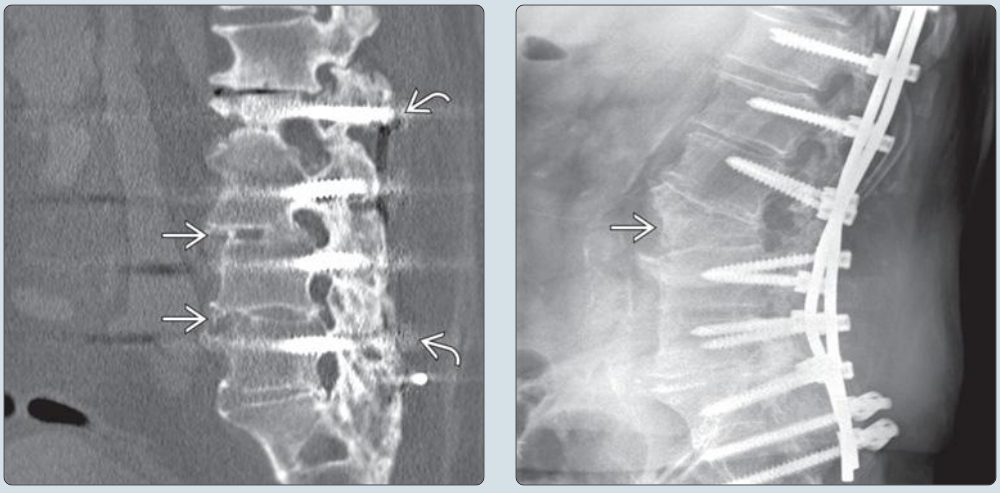
DIAGNOSTIC CHECKLIST

- Evaluate lumbar curvature
  - Normal lumbar lordosis: 40-70º
- Normal plumb line should be < 2 cm from anterior aspect of sacrum

(Left) *Sagittal CT reconstruction demonstrates anterior interbody ➡ and posterior ⇨ fusion at L4-L5. The resulting FBS produces increased biomechanical demands on surrounding muscles, ligaments, and intervertebral discs, leading to back pain and progressive degeneration.* **(Right)** *Lateral radiography reveals FBS due to anterior ⇨ and posterior ➡ fusion at L4-L5 and L5-S1. Note revision of lumbar instrumentation with interbody fusion at L3-L4 ⇨ and posterior transpedicular fusion from L2 to L4 ➡.*



(Left) *Sagittal CT reconstruction exhibits FBS due to anterior ➡ and posterior ➡ lumbar fusion. Physiologic sagittal alignment has a thoracic kyphosis of 30-40° and a lumbar lordosis of 40-70°.* **(Right)** *Lateral radiograph shows a pedicle subtraction osteotomy ➡ for the correction of sagittal imbalance. This involves removal of a wedge of bone with the base at the spinous process and the apex at the anterior margin of the vertebral body and the removal of the pedicles and transverse processes.*



TERMINOLOGY

Abbreviations

- Flat back syndrome (FBS)

Synonyms

- Fixed Sagittal Imbalance (FSI), iatrogenic flat back syndrome/deformity, abnormal spinal kyphosis

Definitions

- Loss of lumbar lordosis (LL) after scoliosis or lower lumbar distraction instrumentation, vertebral fracture, ankylosing spondylitis, or degenerative disease
  - Sagittal imbalance with center of gravity of head anterior to sacrum

IMAGING

General Features

- Best diagnostic clue
  - Thoracolumbar or lumbar instrumentation with straightening of lumbar spine
- Location
  - Thoracolumbar, lumbar spine
- Morphology
  - Loss of normal lumbar lordosis
    - Normal LL: 40-70º, increases with age

Radiographic Findings

- Abnormal plumb line (measure of sagittal balance)
  - Line from center of C7 and distance from this line to anterior aspect of sacrum > 2 cm
- Extension views: Flexibility of curve

CT Findings

- CT myelogram helpful to delineate areas of stenosis

MR Findings

- Delineate areas of stenosis

Imaging Recommendations

- Best imaging tool
  - Evaluate radiographs over time
  - Full-length lateral radiograph obtained with patient's knees and hips fully extended

DIFFERENTIAL DIAGNOSIS

Post-Polio Syndrome

- Weakness in trunk extensor musculature represents major contributing factor to FBS

Scoliosis Instrumentation

- Reducing lateral curvature in thoracic scoliosis can ↑ sagittal deformity and cause flattening of cervical, thoracic, ± lumbar spine

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Inability to stand upright and forward trunk leaning
  - Muscular pain in lower cervical area or upper back
- Patients compensate with hip extension and relative extension of thoracic spine
  - Knee flexion, cervical extension to maintain horizontal gaze

Natural History & Prognosis

- Single biggest risk factor is lower lumbar spine or sacrum distraction instrumentation
  - Magnitude of loss of lordosis ↑ the farther distally the instrumentation is placed
  - Hypolordosis in instrumented segments → increased loading of posterior column of adjacent segments → degenerative changes at junctional level
- Other causes: Pseudoarthrosis resulting in loss of sagittal plane correction
  - Fixed thoracic hyperkyphosis, preexisting thoracolumbar kyphosis
  - Hip flexion contractures
  - Patient positioning (e.g., knee-chest, kneeling) during transpedicular instrumentation
  - Segmental distraction for foraminal enlargement or for bone graft placement → focal kyphosis

Treatment

- Goal: Restore sagittal balance
- Surgical treatment can precipitate perioperative complications in > 60%
- FBS without thoracolumbar kyphosis → osteotomies below level of conus medullaris
  - With flexible thoracolumbar kyphosis → lumbar osteotomies with fusion to thoracic spine
  - With inflexible thoracolumbar kyphosis → osteotomy at thoracolumbar junction or multiple osteotomies
- Osteotomies: Extension (Smith-Petersen), polysegmental, closing wedge
  - Involves removing posterior elements, undercutting adjacent spinous processes
  - Extension osteotomy → risk of injury to vascular structures and neural elements
    - Closing of posterior elements and opening of anterior elements → hyperextension
    - Gap through disc space and biomechanics of axial compressive forces → pseudoarthrosis
  - Pedicle subtraction (PSO) → type of closing wedge, correction in sagittal and coronal planes
- Prevention with proper operative patient positioning
- Avoid distraction instrumentation

DIAGNOSTIC CHECKLIST

Reporting Tips

- Evaluate lumbar curvature

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Scoliosis Instrumentation

KEY FACTS

TERMINOLOGY

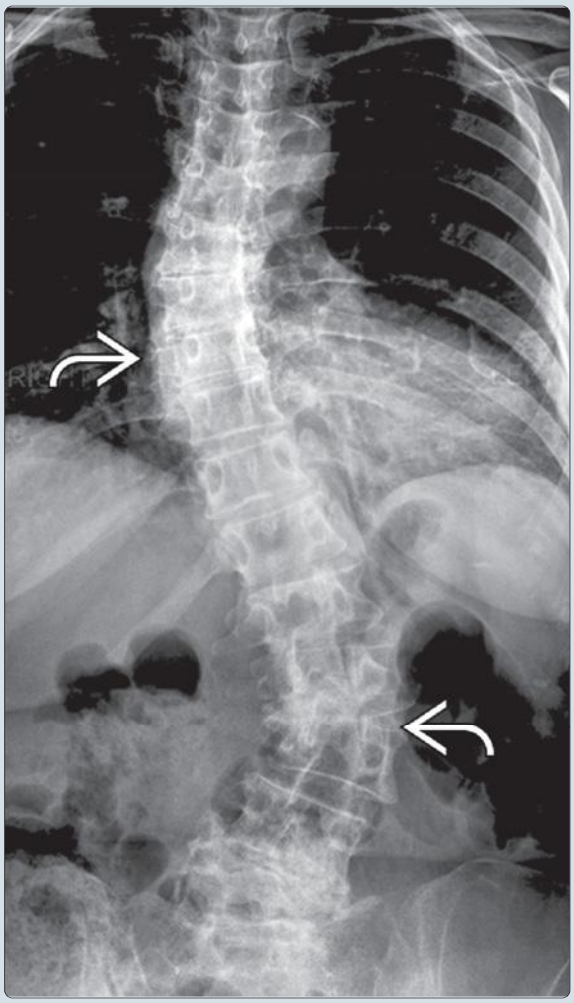
- Spinal fusion surgery recommended when curve magnitude > 40-45° for adolescent idiopathic scoliosis (AIS)
- Adult scoliosis presents with lumbar back, ± leg pain, L3–L4 rotatory subluxation, L4–L5 tilt, and L5–S1 disc degeneration on radiographs

IMAGING

- Radiographs
  - Main thoracic, thoracolumbar, and lumbar curves should be assessed for structural characteristics
  - 36" standing anteroposterior (AP) and lateral radiographs and supine side-bending radiographs
  - In adult scoliosis, assess for degenerated changes and rotatory ± lateral listhesis
- CT
  - Assess integrity of hardware
  - Look for osseous bridging at levels of interbody fusion and lucency along screw tracks
- MR
  - Preoperative planning to evaluate for central &/or foraminal stenosis and disc degeneration

CLINICAL ISSUES

- Adult bones tend to be weaker or osteoporotic, making instrumentation and fusion more difficult
- Degenerative disc changes, spinal stenosis, and facet arthropathy can be exacerbated and in turn exacerbate scoliosis, leading to more rigid spines
- Goals: Prevent progression, restore acceptability of clinical deformity, reduce curvature
  - Resolve pain ± make it more controllable with medications
  - Fuse spine in as normal anatomical position as possible



Anteroposterior radiograph shows sigmoid scoliosis of the thoracic and lumbar spines. Fusion is extended to L5 if there is fixed tilt or subluxation at L4–L5, or to the sacrum if L5-S1 central or foraminal decompression is needed.



Anteroposterior radiograph depicts posterior fusion from the thoracolumbar junction to the sacrum. Extension of the fusion to the sacrum increases the incidence of pseudarthrosis and reoperation.

TERMINOLOGY

Abbreviations

- Scoliosis instrumentation (SI)

Synonyms

- Scoliosis surgery, long spinal fusion surgery (more than 4 vertebral segments)

Definitions

- Spinal fusion surgery recommended when curve magnitude > 40-45° for adolescent idiopathic scoliosis
- Adult scoliosis patient presents with lumbar back, ± leg pain, L3–L4 rotatory subluxation, L4–L5 tilt, and L5–S1 disc degeneration on radiographs
  - Goals: Prevent progression, restore acceptability of clinical deformity, reduce curvature

IMAGING

General Features

- Location
  - Load-bearing axes of cervical and lumbar spinal segments reside in dorsal aspect of vertebral body (VB)
  - Load-bearing axis in thoracic region is located in ventral aspect of VB
    - Pedicle is site of most rigid fixation
    - Shorter constructs can be used with pedicle screws
      - Screw-bone interface has greater strength than hook-bone or wire-bone interfaces
      - Limits number of segments fused, thereby maintaining sagittal contour

Radiographic Findings

- Main thoracic, thoracolumbar, and lumbar curves should be assessed for structural characteristics
  - 36" standing AP and lateral radiographs and supine side-bending radiographs
  - ± 36" supine lateral or hyperextension lateral radiograph over bolster
    - Supine evaluation is helpful to determine curve flexibility
  - From Lenke classification system for adolescent idiopathic scoliosis (AIS), structural curve is defined by a Cobb angle 25° on side-bending radiographs
  - Overall flexibility is important to note → indicates expected curve correction
  - Coronal and sagittal balance, including center sacral vertical line and C7 plumbline
  - Shoulder height, apical vertebral translation of thoracic and lumbar curves, and relative curve magnitudes
  - In adult scoliosis, assess for degenerated changes and rotatory &/or lateral listhesis

CT Findings

- Look for osseous bridging at levels of interbody fusion and lucency along screw tracks
- Assess integrity of hardware
- May be helpful in delineating congenital osseous abnormalities (such as hemivertebrae or fused vertebrae)
- 3D reformations may be helpful for preoperative planning and postoperative evaluation

MR Findings

- Preoperative planning to evaluate for central &/or foraminal stenosis and disc degeneration
- Magnetic susceptibility artifact from hardware can limit evaluation of thecal sac contents and of neural foramina
- Helpful for postoperative evaluation of fluid collections

Imaging Recommendations

- Best imaging tool
  - Upright AP and lateral radiographs
    - Cobb angle measurements

DIFFERENTIAL DIAGNOSIS

Plates and Screws

- Instrumentation in fusion surgery used to stabilize bony elements

Flat Back Syndrome

- Loss of lumbar lordosis after lumbar or scoliosis distraction instrumentation, vertebral fracture, ankylosing spondylitis, degenerative disease
- Sagittal imbalance with center of gravity of head anterior to sacrum

CLINICAL ISSUES

Presentation

- Adult scoliosis curves are more rigid than AIS
  - Rotational deformity and multilevel sagittal vertebral slippages compound coronal malalignment
  - Often progresses over time and can lead to cosmetic and psychological problems, back pain, radiculopathy, spinal claudication, spinal instability disease, and cardiopulmonary compromise
  - Adult scoliosis curves have apices that are predominantly in lumbar spine vs. more common thoracic apices seen in adolescent scoliosis
    - Lumbar or thoracolumbar curves more commonly result in pain and dysfunction than thoracic curves

Natural History & Prognosis

- Adult bones tend to be qualitatively and quantitatively weaker or osteoporotic, making instrumentation and fusion more difficult
- Degenerative disc changes, spinal stenosis, and facet arthropathy can be exacerbated and in turn exacerbate scoliosis, leading to more rigid spines
- Complications
  - Surgical: Severe blood loss, UTI, pancreatitis, obstructive bowel dysfunction due to bowel immobilization, neurological damage
    - Retroperitoneal approach: Abdominal visceral, great vessel, and superior hypogastric nerve plexus injury
    - Transthoracic approach: Injury to great vessels, pulmonary complications, chylothorax, and postthoracotomy pain syndrome
  - Migration of bone graft
  - Breakage of implants
  - Penetration of implants into spinal canal or dorsally into cutaneous tissues
  - Compression of nerve roots by implant components

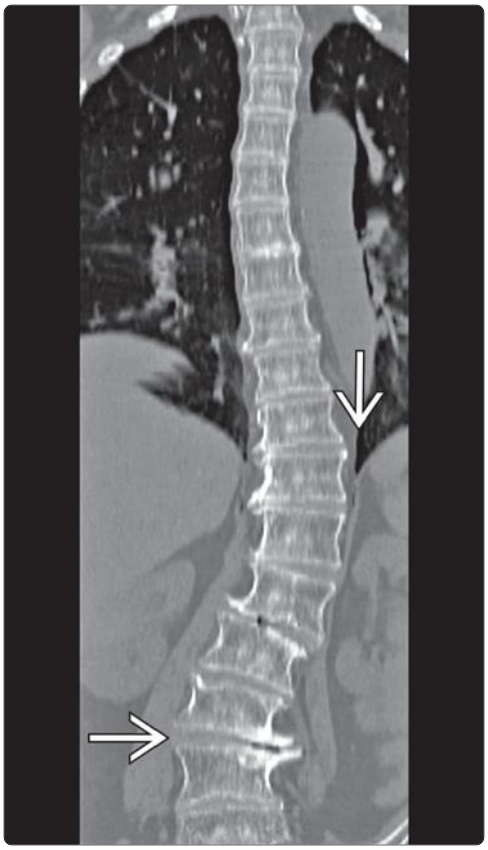
- Failure of spinal fusion requires reoperation to restore curvature correction
  - Pain is primary indication for reoperation
- Pseudoarthrosis
  - Can occur years after surgery
  - Most likely to occur at thoracolumbar junction
  - Reported in 15-27% of cases
    - Risk factors: Preoperative thoracolumbar kyphosis of > 20°
      - Age > 55 years
      - Arthrodesis to S1 compared with arthrodesis to L5 or cephalad level
      - Arthrodesis of more than 12 vertebrae
- Irreversible loss of normal active range of movement of spinal column, including nonfused segments
  - Side flexion is reduced by 20-60%
- Strain on unfused skeletal framework
  - Postsurgical degenerative changes within 2 years
  - Higher degree of correction results in higher rate of degenerative osteoarthritis
    - Rigid spine susceptible to serious injuries even with low impact
  - Reducing lateral curvature in thoracic scoliosis can exacerbate sagittal deformity and cause flattening of cervical, thoracic, ± lumbar spine
    - Loss of lumbar lordosis (flat back deformity)
    - Incidence of proximal junctional kyphosis is 26%
- Study of 34 patients with thoracic to L5 fusion showed subsequent L5–S1 disc degeneration in 66%
  - Cases with normal L5–S1 disc and fusion did not include L5–S1, initially normal L5–S1 disc (based on discography) degenerated within 3 years
    - 1 case degenerated to such a condition that L5–S1 spondylolisthesis was present
- Curvature progression
  - "Crank shaft" phenomenon described in children: Spinal growth causes rotation around fusion
- Pain at iliac graft site, rib resection site
- Infections reported in 5-10% patients at 11-45 months after surgery
  - More common in SI possibly due to larger instrumentation or increasing prevalence of multiresistant bacteria in hospital settings
- Inflammatory process
  - Particulate debris stimulates autoimmune response causing bone deterioration
- Increased torso deformity with rib hump
- Cardiopulmonary symptoms

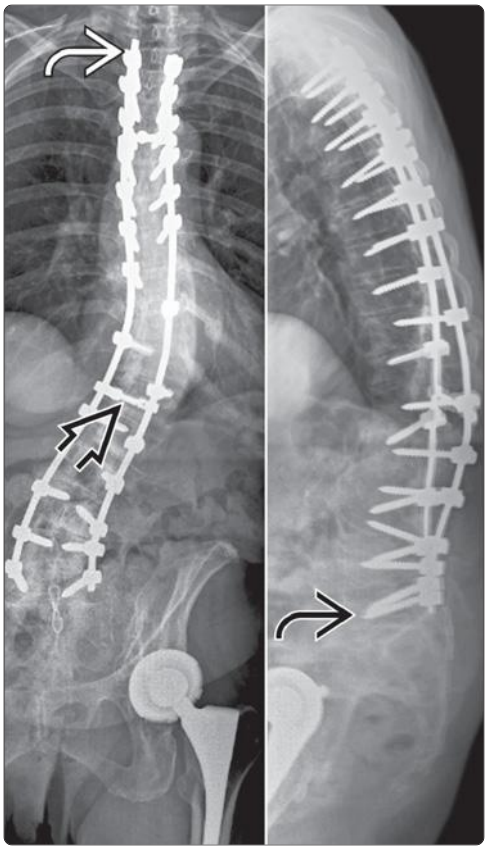
Treatment

- Goal of surgery in adult scoliosis
  - Resolve pain ± make it more controllable with medications
  - Fuse spine in as normal anatomical position as possible
  - Biggest operative decision is to determine proximal and distal extent of instrumentation and fusion
    - Operative levels include fusion of Cobb angle, typically L3 or L4, and extending to L5 when there is lateral or rotatory listhesis present
    - Proximal instrumented level should at least end at stable vertebra
      - If shoulders are level and main thoracic curve is minimal and well balanced, then stable end vertebra is appropriate level to end fusion
      - Generally, it is felt best to cross thoracolumbar junction transitional zone to reduce adjacent segment stresses and junctional kyphosis above instrumented fusion
    - Restoration of sagittal balance reported to be primary parameter associated with outcome
- Conservative management
  - Pain control
  - Physiotherapy
  - Scoliosis inpatient rehabilitation
  - Corrective bracing
  - Regarded as effective when curvature progression has been stopped below specific limits
- Scoliosis instrumentation recommended when magnitude of curvature exceeds 40-45° in AIS
  - Approaches: Posterior instrumentation and fusion alone, anterior instrumentation and fusion alone, and anterior release (to restore spinal flexibility) combined with posterior fusion, posterior only
    - Main principle: Spine is structural scaffold with parts cemented on via bone paste → overall straighter shape
    - Steel rods, screws, wires, etc. based on surgeon preference
      - Suggested extension of posterior instrumentation to upper thoracic spine when high risk for adjacent segment failure, such as cases of exaggerated thoracic kyphosis
  - Fusion often extended to sacrum if
    - L5–S1 spondylolisthesis
    - Previous L5–S1 laminectomy
    - Any form of L5–S1 stenosis, be it central, lateral recess, or foraminal
    - Oblique takeoff at L5–S1
    - Severe L5–S1 disc degeneration (but not necessarily if there is L5–S1 disc calcification)
  - If extension to S1 is undertaken, then iliac fixation with long fusions (greater than 3 levels)
  - Since degenerated lumbar curves present with loss of lordosis, surgical plan should specifically address this issue

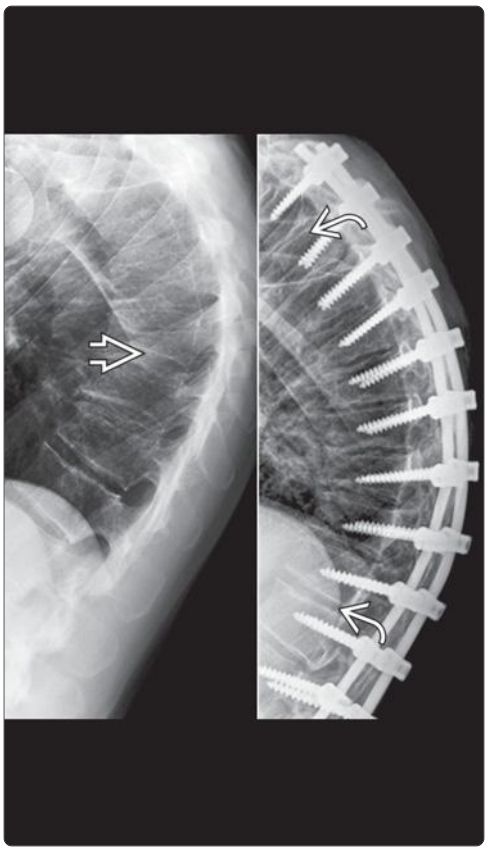
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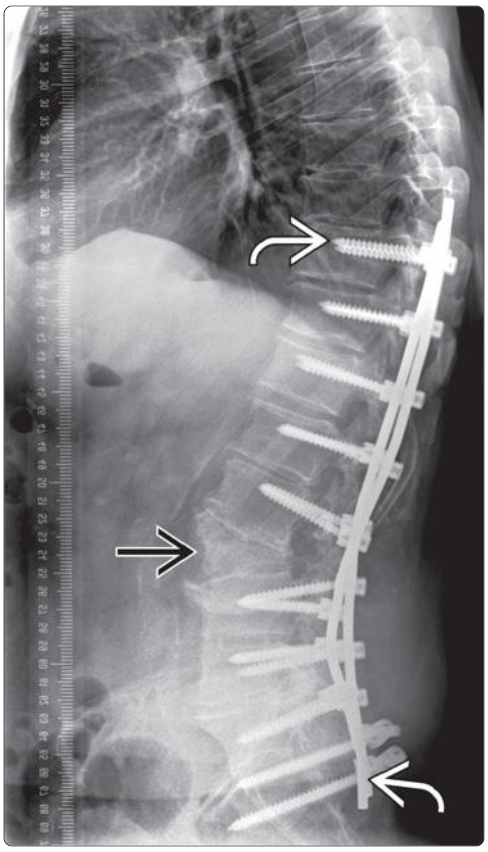
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(Left) *Coronal CT reconstruction displays levoscoliosis at the thoracolumbar junction and dextroscoliosis ➔ of the lumbar spine. CT with multiplanar reformations gives information about alignment and the effects of the listhesis on the vertebral canal and neural structures.* **(Right)** *AP and lateral radiographs illustrate extensive fusion from the thoracic spine ➔ to the L5 level ➔. In most cases, fusion is not terminated next to a severely degenerated segment, especially if there is fixed tilt or subluxation. The proximal extension of the fusion should not stop distal to a structural proximal thoracic curve. Stopping long fusions at L5 can lead to subsequent degeneration. Crosslinks ➔ are added to the construct primarily to improve overall construct torsional stiffness.*





(Left) *Lateral radiograph shows prominent thoracic kyphosis ➔. Studies show that severity of symptoms increases linearly with progressive sagittal imbalance. Kyphosis is more favorable in the upper thoracic region, but poorly tolerated in lumbar spine. After posterior pedicle instrumentation ➔, there is mildly improved thoracic kyphosis.* **(Right)** *Lateral radiograph shows thoracolumbar to sacrum posterior fusion ➔ and L3 pedicle subtraction osteotomy ➔ for restoration of lumbar lordosis. Because of the 3D nature of scoliosis and associated disc degeneration in adults, lumbar scoliosis frequently results in a significant loss of lumbar lordosis or actual kyphosis, resulting in positive sagittal balance position.*

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PART II
SECTION 1

Vertebral Column, Discs, and Paraspinal Muscle



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Craniocervical Junction

Occipital condyle fractures are classified into 3 types
- Type I = comminuted fractures due to axial loading; stable if contralateral side is intact
- Type II = occipital condyle fracture with skull base fractures; most of these are stable
- Type III = avulsion fracture due to tensile force on alar ligaments; may show occipitocervical instability

Recent data (Maserati 2009) suggests that initial evaluation should be primarily concerned with identification of craniocervical malalignment. Fusion or halo used in patients with initial scans show fracture and malalignment, with rigid cervical collar with delayed imaging follow-up for all others.

Atlantooccipital Disassociation (Dislocation)/C0-C1 Distraction Injury

Complete (disassociation) or partial (subluxation) ligamentous disruption between occiput and C1 which can occur in one of 3 directions: (1) Anterior superior displacement of cranium relative to spine most common; (2) Pure distraction injury with superior displacement of cranium; or (3) Posterior dislocation of cranium which is least common.

Numerous measurement techniques have been used to assess craniovertebral junction trauma, many of which were first defined in the plain film era. Many of these measurements have been superseded by the direct soft tissue visualization afforded by CT and MR. There is reasonable literature support for use of the following measurements.

Basion-dental interval (BDI) is abnormal if > 10 mm on sagittal CT.

Summed condylar displacement (sum of the bilateral distances between midpoint of occipital condyle and C1 condylar fossa) is abnormal if > 4.2 mm.

Single side condylar distance measurement of > 2 mm is also considered abnormal in adults. The 2 mm upper limit of C0-C1 spacing also applies to children up to 18 years of age.

Other measurements such as the Powers ratio and Lee lines do not have sufficient sensitivity and specificity to recommend their use. The Harris "rule of 12" for the BDI and basion-axial interval are for plain film use only, so are very limited given the use of CT for acute trauma evaluation.

C1 Fractures
- Anterior arch = vertical or transverse with avulsion from longus colli
- Anterior arch bilateral fractures with posterior atlantoaxial dislocation = plow fracture
- Lateral mass = stable if lateral ring intact; rare
- Posterior arch = common
- Jefferson = combined lateral mass displacement relative to C2 of 6.9 mm indicates disruption of transverse ligament and potential for instability

Atlantoaxial Instability
- Nonphysiologic motion between C1-C2
- Wide variety of causes
- → transverse ligament rupture (most common)
- → odontoid fracture
- → unstable Jefferson fracture
- → fracture of lateral mass of C1 or C2
- → unilateral alar ligament rupture
- → alar and tectorial membrane rupture

Classification of Atlantoaxial Rotatory Fixation (Fielding 1977)
- Type I = rotation about dens without anterior translation (no increase in ADI)
- Type II = rotation about 1 lateral mass with anterior translation of 3-5 mm (ADI) (transverse ligament injury)
- Type III = rotation about lateral mass with anterior translation > 5 mm (transverse and alar ligament injury)
- Type IV = posterior dislocation of C1 behind dens (rare, usually fatal)

Odontoid
- Type I = avulsion at tip of odontoid
- Type II = transverse fracture of dens above C2 body
- Type III = fracture involving superior portion of C2 body

C2 Ring Fractures (Effendi 1981)
- Type I = bilateral pars fractures with < 3 mm anterior subluxation (stable)
- Type II = displacement of pars fracture + anterior translation of C2 with discoligamentous injury
- Type III = pars fractures with C2-3 facet dislocations

C2 Body Fractures (Fujimura 1996)
- Type I = extension teardrop fracture of anterior inferior endplate of C2
- Type II = horizontal shear fracture through body (more caudal than type III odontoid fracture)
- Type III = C2 body burst fracture
- Type IV = unstable sagittal cleavage fractures

Cervical Fracture Classification

Hyperflexion
- Simple compression fracture
- Anterior subluxation: Posterior ligament disruption
- Bilateral interfacetal dislocation: Unstable
- Flexion teardrop fracture: Unstable
- Clay shoveler's fracture: Avulsion of spinous process of C7-T1

Hyperflexion and Rotation
- Unilateral facet dislocation (locked facet)
- May have associated facet fracture
- Radiograph shows forward displacement of vertebra < 1/2 AP diameter of cervical vertebral body

Hyperextension and Rotation
- Pillar fracture

Vertical Compression
- Jefferson fracture = fractures of both anterior and posterior rings with 2, 3, or 4 parts with radial displacement
- Burst fracture = middle column involvement with bony retropulsion

Hyperextension
- Hyperextension dislocation
- C1 anterior arch avulsion fracture = longus colli insertion around anterior tubercle of C1
- Extension teardrop fracture of C2
- C1 posterior arch fracture = compressed between occiput and C2 spinous process
- Lamina fracture = between articular mass and spinous process
- Hangman's fracture = bilateral pars fractures of C2
- Hyperextension fracture: Dislocation = bilateral facet fracture ± dislocation

Lateral Flexion
- Uncinate process fracture

Subaxial Cervical Spine Injury Classification (Vaccaro 2007)
- 3 major components including morphology of spinal column disruption, integrity of disco-ligamentous complex, and neurologic status
- Within each component, subgroups are graded from least to most severe (see table 1)

Thoracolumbar Fracture Classification

Holdsworth 2 Column Model (1963)
- Superseded by Denis classification
- Anterior column = ALL, vertebral body, disc, PLL
- Posterior column = skeletal and ligamentous structures posterior to PLL

Denis 3 Column Model (1983)
- Anterior: ALL, anulus, anterior vertebral body
- Middle: Posterior wall of vertebral body, anulus, PLL
- Posterior: Facets, posterior elements, posterior ligaments
- 3 column model also relevant to lower cervical injuries

Denis Subclassification of Burst Fracture (1984)
- **Denis type A**
- Axial load force; anterior and middle columns involved, unstable
- Upper and lower endplates involved
- **Denis type B and C**
- Flexion and axial load, anterior and middle columns, possibly unstable
- B upper endplate involved (most common)
- C lower endplate involved
- **Denis type D**
- Axial load and rotation, all columns, unstable
- Atlas modification of D injuries (1986)
- D1 burst lateral translation, D2 burst sagittal translation
- **Denis type E**
- Lateral compression, all columns, possibly unstable

Magerl AO Pathomorphologic System (1994)
- A, B, C types reflecting common injury patterns
- Each type has 3 groups, each with 3 subgroups (3-3-3 scheme)
- Type A vertebral compression fractures due to axial loading without soft tissue disruption in transverse plane (66%)
- Type B distraction of anterior and posterior elements with soft tissue disruption in axial plane (14.5%)
- Type C with axial torque forces giving anterior and posterior element disruption with rotation (19%)
- Severity progresses through type A to C, as well as within types, groups, and subdivisions
- Stable type A1 most common (wedge fracture)
- A3 corresponds to "burst fracture" of Denis classification
- Unstable: A3.2, A3.3, B, C types

McCormack "Load-Sharing" Classification (1994)
- Specifically designed to evaluate need for anterior column reconstruction following pedicle screw stabilization
- Also useful as a more generic guide to the magnitude of comminution and biomechanical instability
- Comminution graded
- → amount of vertebral of body damage
- → fragment of spread at fracture site
- → degree of corrected kyphosis

Thoracolumbar Injury Classification and Severity Score (TLICS) (Vaccaro 2006)
- 3 components give final numeric score that directs treatment
- Injury mechanism, integrity of posterior ligamentous complex, and neurologic status (see table 2)

Unstable Fractures

Cervical
- Atlantoaxial dissociation
- Atlantooccipital dislocation
- Occipital condyle fracture with malalignment
- Jefferson fracture where combined offset of C1 lateral masses > 7 mm or > 7 mm separation of fracture fragments
- Hangman II, III
- Odontoid types I, II
- Subaxial anterior subluxation of > 3.5 mm
- Hyperflexion fracture dislocation
- Hyperflexion teardrop
- Hyperextension fracture dislocation
- Burst

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Subaxial Cervical Spine Injury Classification

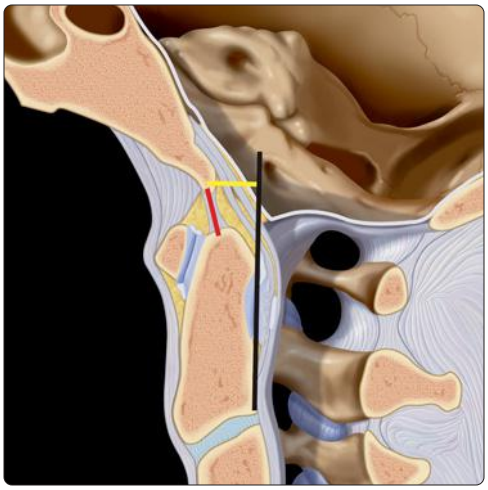
	Description	Points
Morphology		
	No abnormality	0
	Compression	1
	Burst	+ 1 = 2
	Distraction (perched facet, hyperextension)	3
	Rotation/translation (facet dislocation, unstable teardrop)	4
Disco-Ligamentous Complex		
	Intact	0
	Indeterminate (MR signal abnormality only, isolated interspinous widening)	1
	Disrupted	2
Neuro Status		
	Intact	0
	Root injury	1
	Complete cord injury	2
	Incomplete cord injury (most urgent situation so higher value than complete injury)	3
	Continuous cord compression in setting of neuro deficit (modifier)	+ 1

Surgical vs. nonsurgical determined by total score: 1-3 nonoperative treatment, ≥ 5 operative treatment recommended. (Vaccaro 2007.)

Thoracolumbar Injury Severity Score

	Description	Qualifier	Points
Injury Mechanism			
	Compression		
		Simple	1
		Lateral angulation > 15°	1
		Burst	1
	Translational/rotational		3
	Distraction		4
Posterior Ligamentous Complex			
	Intact		0
	Suspected/indeterminate for disruption		2
	Injured		3
Neuro Status			
	Nerve root involvement		2
	Cord, conus involvement (incomplete)		3
	Cauda equina involvement		3
	Cord, conus involvement (complete)		2

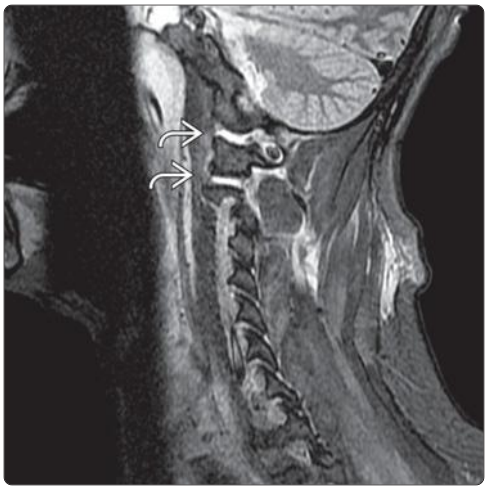
Score is a total of 3 components. Score ≤ 3 suggests nonoperative treatment, while score of 4 is indeterminate. Score ≥ 5 suggests operative treatment. For injury mechanism, the worst level is used, and the injury is additive. An example is distraction injury with burst without angulation is 1 (simple compression) + 1 (burst) + 4 (distraction) = 6 points. (Vaccaro 2006.)



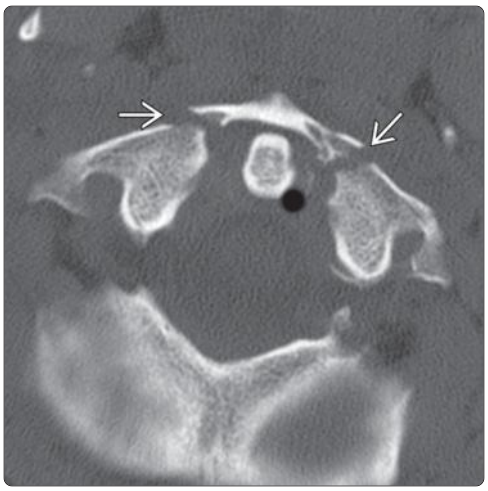


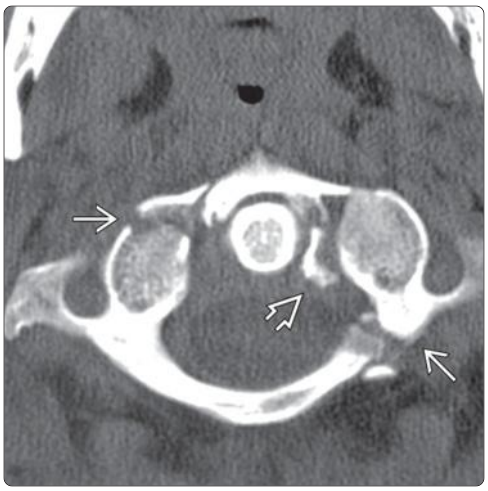
(Left) *Sagittal graphic shows normal basion-dental interval (BDI) (red line) and the basion-posterior axial line interval (BAI) (yellow line). BAI is the distance from basion to posterior axial line (black line). BDI is abnormal if > 10 mm on sagittal CT. BAI is abnormal if > 12 mm on plain films.* **(Right)** *Sagittal NECT of trauma patient with AOD shows abnormal distance between basion and dens (yellow), abnormal separation of basion from posterior axial line (orange). Posterior axial reference line is white.*





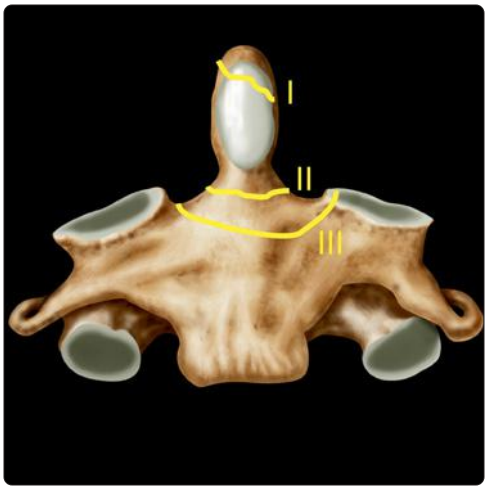
(Left) *Parasagittal NECT in a patient with AOD shows widening of both C0-C1 and C1-C2 articulations ➔. Summed condylar displacement (sum of the bilateral distances between midpoint of occipital condyle and C1 condylar fossa) is abnormal if > 4.2 mm.* **(Right)** *Sagittal STIR shows abnormally widened and hyperintense C0-C1 and C1-C2 articulations ➔. This patient underwent occiput to C3 posterior fusion for atlantooccipital and atlantoaxial dislocation.*





(Left) *Axial NECT shows multiple fracture sites involving the C1 ring ➔ without canal compromise.* **(Right)** *Axial NECT shows fractures involving both anterior and posterior rings of C1 ➔ and additional avulsion fracture off of mesial C1 ring at the level of attachment of transverse ligament ➔.*

(Left) *Coronal graphic of the C2 vertebra shows the schematic for location of types I, II, and III odontoid fractures.* **(Right)** *Coronal NECT shows an oblique type III fracture extending across the base of odontoid and the upper body of C2 with fragmentation of the left lateral mass of C2.*





(Left) *Sagittal NECT shows a C2 pars fracture without significant offset, angulation, or distraction.* **(Right)** *Axial bone CT shows a typical case of traumatic C2 pars fractures in classic hangman configuration. This patient demonstrates the mildest class of injury (type I) using the Levine and Edwards modification of the Effendi classification system and would be considered a stable fracture.*

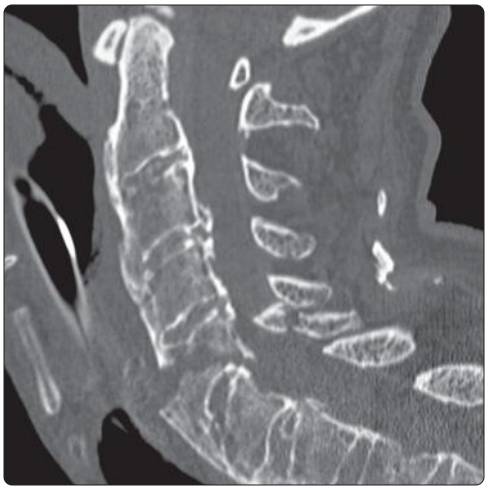


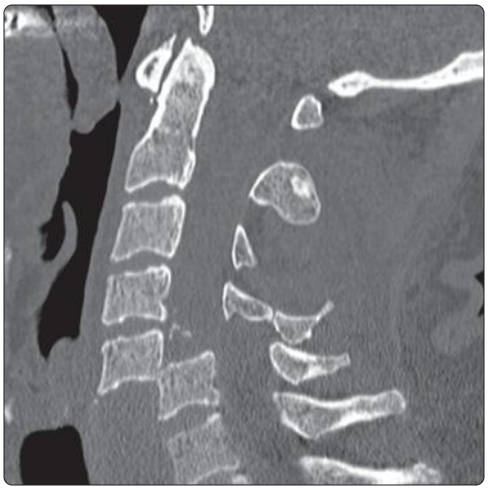


(Left) *Sagittal graphic shows an unstable cervical hyperflexion injury involving anterior and posterior longitudinal ligaments, disc, and interspinous ligaments with epidural hemorrhage and cord compression.* **(Right)** *Lateral radiograph shows C4-C5 flexion facet dislocation with bilateral "jumped" facets. There is disruption of all 3 columns by this injury.*



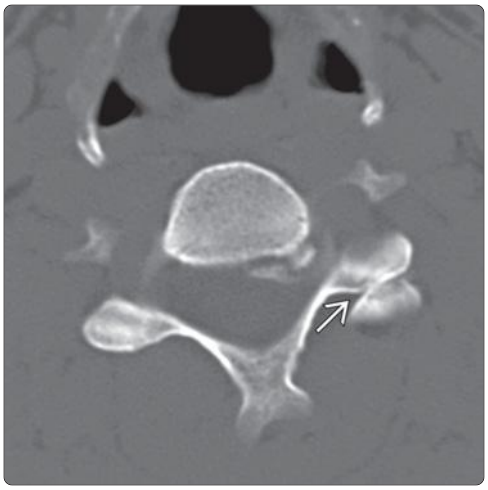




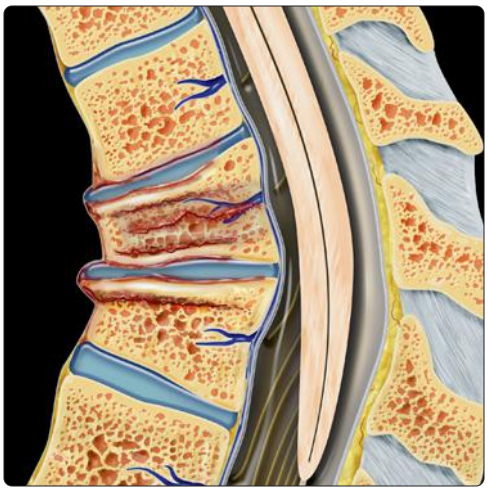


(Left) *Sagittal NECT of transverse extension fracture in a patient with ankylosing spondylitis shows horizontal fracture line extending through the C5 body and into the base of the spinous process with posterior displacement of the superior aspect of the fracture, indicating hyperextension mechanism. Note typical anterior ossification of AS.* **(Right)** *Sagittal NECT shows complete dislocation of the upper cervical spine at the C5-C6 level. There is over 100% listhesis of the C5 body relative to C6.*





(Left) *Lateral radiograph shows hyperflexion injury with bilateral locked facets, widened disc space, anterior subluxation of C6 on C7 of 50%, and widened spinous process distance.* **(Right)** *Axial bone CT shows unilateral left facet dislocation ➡. There is reversed relationship of the facets, with the inferior articular facet of C6 lying anterior to the superior articular facet of C7 (back-to-back apposition).*





(Left) *Sagittal graphic of the thoracolumbar junction shows compression (wedge) fractures involving primarily the anterior column with normal middle and posterior columns.* **(Right)** *Sagittal graphic shows a Chance ("seat belt") fracture of the thoracolumbar junction extending in the horizontal plane through body and posterior elements (3 column involvement).*

Atlantooccipital Dislocation

KEY FACTS

TERMINOLOGY

- Disruption of stabilizing ligaments between occiput and C1

IMAGING

- Widened prevertebral soft tissues (nonspecific)
- Condylar sum > 4.2 mm has 100% sensitivity, 69% specificity, and 76% accuracy
- Increased BDI distance > 8.5 mm (CT in adults)
- Widened unilateral AOI > 2 mm
  - Widened, fluid-filled facet joints between condyles and C1
- STIR/T2WI MR best show ligamentous injury

TOP DIFFERENTIAL DIAGNOSES

- Occipital condyle fracture
- C1 Jefferson fracture
- Odontoid fracture
- Rheumatoid arthritis, adult

PATHOLOGY

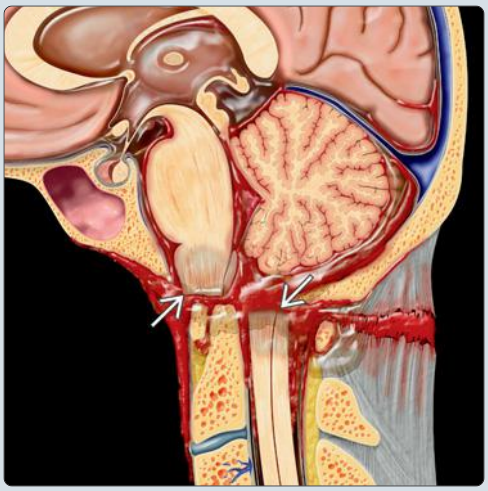
- High-speed motor vehicle accident
- Associated injuries
  - Brainstem and cranial nerve injury
  - Fractures of occipital condyles, C1 and C2

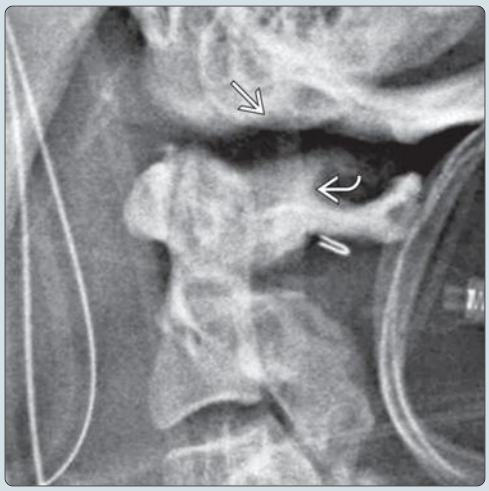
CLINICAL ISSUES

- < 1% of acute cervical spine injuries
- Often immediately fatal
- However, may have good outcome with recognition and fixation

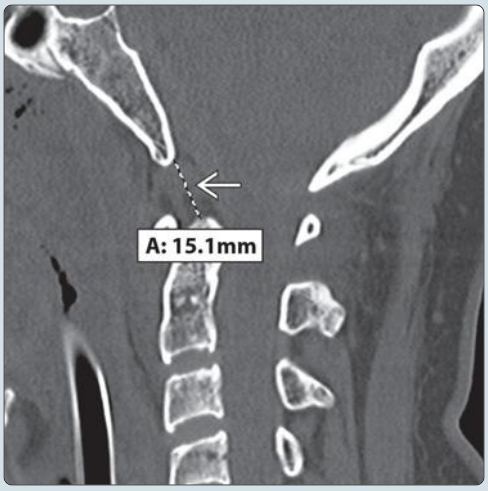
DIAGNOSTIC CHECKLIST

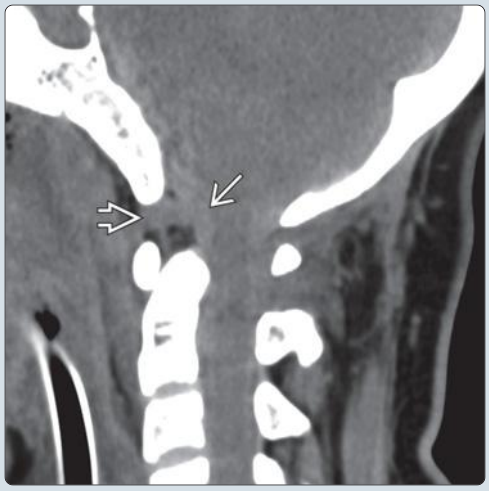
- No imaging sign is 100% sensitive
- Condylar sum > 4.2 mm most sensitive CT sign of AOD
- Powers ratio not recommended (has low sensitivity and specificity)
- Detection of subarachnoid hemorrhage at CCJ should direct search for atlantooccipital injuries





(Left) *Sagittal graphic depicts fatal atlantooccipital dissociation with cord transection ➡ at the craniocervical junction. Stretch injury to the spinal cord may occur, resulting in neurologic dysfunction.* **(Right)** *Lateral radiograph shows separation between occipital condyle ➡ and C1 ➡. The Powers ratio (> 1), BAI (> 12 mm), and BDI (> 12 mm) are classic plain film criteria used to determine craniocervical dissociation.*





(Left) *Sagittal CT reconstruction illustrates increase in the BDI ➡. Normal distance by CT is ≤ 8.5 mm in adults, or 12 mm on lateral radiographs.* **(Right)** *Sagittal CT in soft tissue algorithm exhibits hyperdense blood products in the ventral epidural space ➡. Abnormal soft tissue is also observed in the supraodontoid space ➡, compatible with hemorrhage along the ligaments. Cadaveric studies have shown that rupture of both the tectorial membrane and alar ligament are necessary for AOD to occur.*

TERMINOLOGY

Abbreviations

- Atlantooccipital dislocation (AOD)

Synonyms

- Craniocervical dissociation
- Occipitoatlantal dislocation

Definitions

- Disruption of stabilizing ligaments between occiput and C1 ± between C1 and C2

IMAGING

General Features

- Best diagnostic clue
  - Widening between occipital condyles and C1
  - Increased distance between basion and dens
- Location
  - Axial distraction across craniocervical junction (CCJ) can produce either atlantooccipital or atlantoaxial dislocation
    - Dimensions of superior and inferior extensions of cruciate ligaments, orientation of alar ligaments, and presence of prominent apical ligament shown in cadaveric studies to affect whether injury occurs at C0–C1 or C1–C2
    - One study reported > 1/3 of patients with craniocervical disassociation had both C0-C1 and C1–C2 distraction injuries

Radiographic Findings

- Widened prevertebral soft tissues (nonspecific)
- Increased distance from basion-dens interval (BDI) > 12 mm
- Powers ratio (not recommended)
  - BC = distance from tip of clivus (basion) to anterior cortex of C1 posterior arch
  - AO = distance from posterior cortex of C1 anterior arch to posterior margin foramen magnum (opisthion)
  - BC/AO > 1.0 indicates anterior AOD
  - Sensitivity is low, landmarks difficult to identify
- Wackenheim line
  - Along posterior margin of clivus
    - Normally extends through posterior tip of dens
  - In longitudinal or posterior AOD, line extends posterior to dens
  - In anterior AOD, line extends through center of dens or even further anterior
- Basion-axial interval (BAI)
  - Vertical line drawn through tip of basion
  - Vertical line drawn through posterior margin of C2 vertebral body
  - Normal in adults: +12 mm to - 4 mm
  - Normal in children: + 12 mm to 0 mm
- Atlantooccipital interval (AOI): Space between condyles and C1
  - > 4 mm considered diagnostic
  - Joint space normally bilaterally symmetric
  - AOD may widen space on just 1 side
- Radiographs technically limited: Inadequate exposure, patient motion, positioning difficulties, and poor visualization, particularly at CCJ

Fluoroscopic Findings

- Traction may be applied under fluoroscopy to assess widening of C0-C1

CT Findings

- Widened unilateral AOI > 2 mm
  - Normal range 0.5-1.8 mm in adults (wider in children)
    - 1.4 mm (max for 97.5% of population)
  - Condylar sum > 4.2 mm has 100% sensitivity, 69% specificity, and 76% accuracy
- Midline occiput to C1 spinolaminar line > 4.2 mm
  - May be artifactually shortened due to extension positioning in collar
- Basion-dens interval range 1.4-9.1 mm
  - Normal < 8.5 mm (max for 97.5% of population) in adults
- C1-C2 spinolaminar interval > 7.8 mm
  - C1-C2 facet joint distance > 3 mm
- Anterior or posterior position of C1 relative to basion
  - Incongruity of occiput-C1 facet joints well seen on sagittal images
- Avulsion fracture of occipital condyle or anterior arch of C1
- Subarachnoid hemorrhage at CCJ is associated with AOD & its detection should direct search for atlantooccipital injuries

MR Findings

- STIR, T2WI best shows ligamentous injury
  - Tectorial membrane disruption seen in 71% of 1 series of 16 pediatric patients
  - Nonvisualization of apical, alar, and anterior atlantoaxial ligaments
    - These ligaments are consistently seen in normal patients with high-resolution MR centered on craniocervical junction
  - Posterior atlantoaxial membrane often remains intact
  - No good data on patients showing only ligament injury with normal alignment on MR and CT
    - Cervical traction under fluoroscopy may be useful to determine stability
- Widened, fluid-filled facet joints between condyle and C1 > 2 mm
  - May be unilateral
- Anterior or posterior displacement of C1 relative to base of skull
  - Best seen on sagittal images
- Epidural &/or subdural hematoma in upper cervical spine
- Posterior fossa subarachnoid hemorrhage
- Vertebral artery injury
- Prevertebral hematoma

Imaging Recommendations

- Best imaging tool
  - Can be evaluated with CT or MR
  - CT useful for rapid triage
  - MR better shows extent of ligament injury
- Protocol advice
  - Advisable to have specific trauma protocol for full evaluation of craniocervical junction
    - Coronal and sagittal STIR, heavily T2-weighted sequences such as MR cisternography

- Axial images should extend superiorly through occipital condyles
- 3D volumetric T2 sequences can help delineate ligamentous structures at CCJ
- CTA/MRA for evaluation of vertebral dissection and carotid blunt trauma

DIFFERENTIAL DIAGNOSIS

Occipital Condyle Fracture

- Best seen on multiplanar CT reconstructions
- Condyle avulsions may be associated with AOD

Jefferson Fracture of C1

- Fractures of anterior and posterior ring of C1
- Often see lateral displacement of C1 lateral masses

Odontoid C2 Fracture

- Type 2 odontoid fracture often results in posterior displacement of dens
- Will disrupt Powers ratio

Atlantoaxial Rotatory Fixation

- Asymmetry of lateral masses of C1 relative to dens
- Occiput-C1 facet joints and atlantodental interval are normal

Down Syndrome

- Nontraumatic atlantooccipital instability
  - Hypermobility of C1-C2 joint due to laxity of alar ligament
  - Ligamentous laxity reported in 15–20% of Down syndrome patients

Rheumatoid Arthritis, Adult

- Nontraumatic atlantooccipital instability
  - Pannus destabilizes joints and ligaments
- Radiographs show erosions
- CT/MR shows pannus

PATHOLOGY

General Features

- Etiology
  - High-speed motor vehicle accident
    - Shear injury of head from spinal column **or**
    - Vertical distraction of head from spine
- Associated abnormalities
  - Intracranial hemorrhage
  - Brainstem and cranial nerve injury
  - Fractures of occipital condyles, C1 and C2
  - Ligamentous disruption, C1-2
  - Facial fractures
  - Chest trauma

Staging, Grading, & Classification

- Longitudinal AOD
  - Vertical displacement
  - Due to distracting force between head and spine
- Anterior AOD
  - Skull positioned anterior to C1
  - Due to anterior shear force on head
- Posterior AOD
  - Skull positioned posterior to C1
  - Due to posterior shear force on head

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Respiratory failure, cranial nerve and motor deficits
  - 20% have no neurological deficit, need high index of suspicion
- Other signs/symptoms
  - Delayed neurological deterioration may be due to post-traumatic pseudomeningocele
    - Seen in association with hydrocephalus

Demographics

- Age
  - More common in children
    - Due to relatively large head, horizontal orientation of condyles
- Epidemiology
  - < 1% of acute cervical spine injuries

Natural History & Prognosis

- Often immediately fatal
- However, with recognition and fixation may have good outcome
- Poor outcomes if basion-dens distance ≥ 16 mm
- Injury severity score and complete neurologic injury correlate with nonsurvivability

Treatment

- Occiput to C2 fusion required

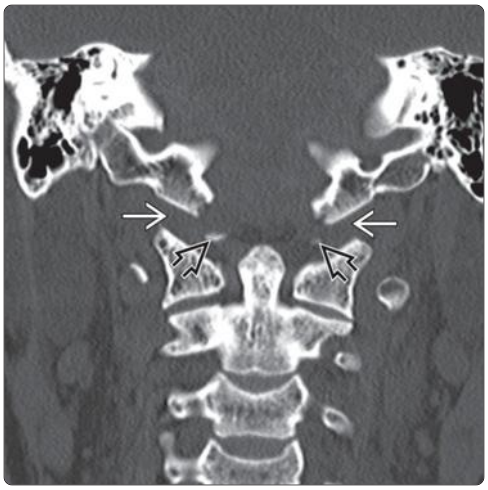
DIAGNOSTIC CHECKLIST

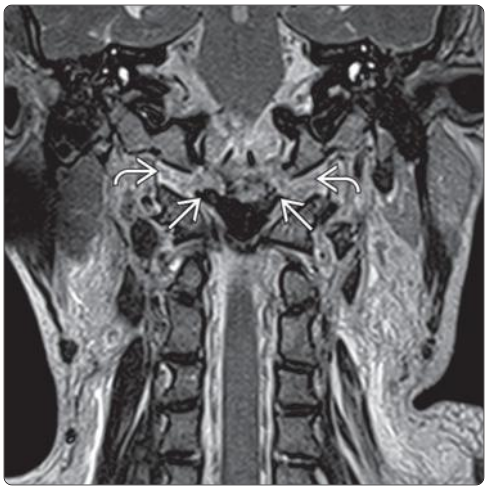
Consider

- No imaging sign is 100% sensitive
- Condylar sum > 4.2 mm is most sensitive CT sign of AOD
- Powers ratio not recommended
  - Basion difficult to see on most trauma radiographs
  - Will not detect posterior or vertical displacement
- All patients with upper cervical prevertebral soft tissue swelling should undergo CT scan

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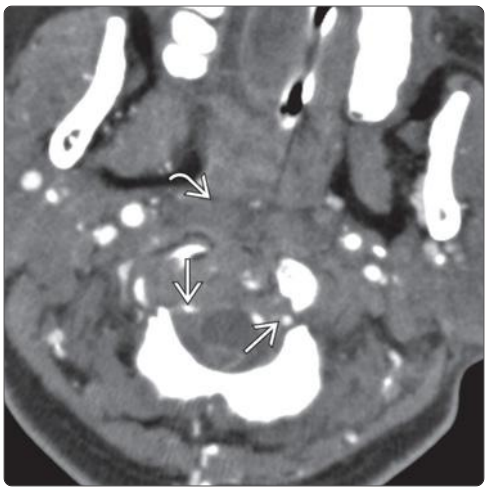


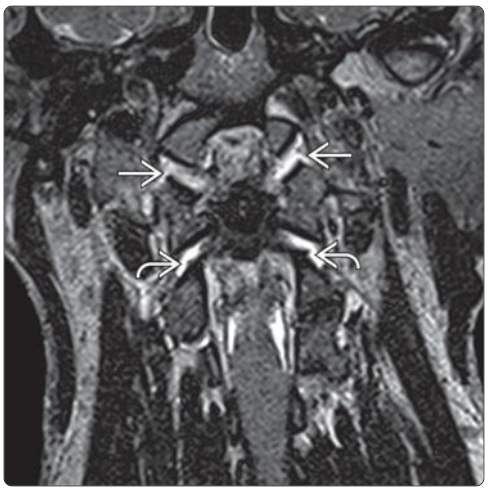
(Left) *Coronal CT reconstruction demonstrates widening of the atlantooccipital joints (AOJs) ➡. Small osseous fragments ⇨ avulsed from the inferomedial aspects of bilateral occipital condyles (OC) are noted. Fractures at the insertions of the alar ligaments are equivalent to intrinsic disruptions and contribute to instability of CCJ.* **(Right)** *Coronal T2WI MR reveals widening and hyperintensity within bilateral AOJs ➡. Discontinuity of bilateral alar ligaments ➡ is observed.*





(Left) *Sagittal CT illustrates anterior subluxation of the OC ➡ on C1 ➡. Small displaced OC fracture fragments ⇨ are seen.* **(Right)** *Sagittal T2WI MR exhibits extensive abnormality of the CCJ ligaments with thinning and irregularity of the tectorial membrane ⇨. The anterior atlantooccipital membrane is irregular and stretched anteriorly ➡. Abnormal hypointense soft tissue in the supraodontoid space ➡ suggests injury to the apical, alar, and Barkow ligaments.*





(Left) *Axial CTA reveals luminal irregularity and narrowing of bilateral vertebral arteries ➡. There is prevertebral soft tissue swelling and edema ➡. Vascular injuries range from vasospasm and dissection of the carotid and vertebral arteries to complete transection with extravasation.* **(Right)** *Coronal T2WI MR shows distraction of bilateral AOJs ➡ and atlantoaxial joints (AAJs) ➡. Mechanisms of hyperextension of the AOJ and hyperflexion of the AAJ may explain the combined injury pattern.*

Ligamentous Injury

KEY FACTS

IMAGING

- Best imaging method is T2W FS or STIR MR
  - Abnormal T2 hyperintensity or gross discontinuity of spinal ligamentous structure
- Flexion-extension films: Segmental instability
  - > 3.5 mm subluxation
- STIR and T2WI FS to evaluate spinal ligamentous injury
  - Normal ligament should be thin, contiguous, and low in signal intensity on both T1 and T2WI
  - Increased signal within ligamentous structure indicative of edema, hemorrhage, or inflammation (stretch injury, strain, partial tear)
  - Complete discontinuity of ligamentous structure on T2WI indicative of disruption

PATHOLOGY

- Forced motion of vertebral segment beyond limits of tissue elasticity
- Associated abnormalities
  - Spinal instability
  - Traumatic disc herniation
  - Vertebral fractures

CLINICAL ISSUES

- Spinal instability predisposing to progressive deformity or to impingement on spinal cord, nerve roots
  - Instantaneous (instability immediately following injury)
  - Delayed (> 20 days after injury)

DIAGNOSTIC CHECKLIST

- Ventral ligamentous injury of cervical spine without fracture is rare (< 0.7%)

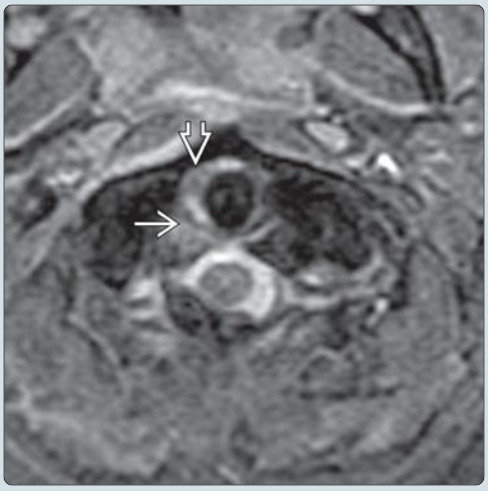
(Left) *Sagittal STIR MR of thoracic hyperextension injury shows disruption of the ALL and anterior disc space, contiguous with fracture extending through the superior endplate of the subjacent vertebral body.* **(Right)** *Sagittal T2 MR of cervical hyperflexion-distraction injury shows hyperintensity of the interspinous ligament and disruption of both ligamentum flavum and PLL. Thickening and increased signal are seen in prevertebral soft tissues.*





(Left) *Sagittal STIR MR of hyperflexion-distraction injury with bilateral facet dislocations shows anterolisthesis at C6-C7 and disruption of both ALL and PLL. There is edema in the dorsal cervical musculature.* **(Right)** *Axial T2* GRE MR shows increased signal to the right of midline in the transverse ligament due to disruption with mild asymmetric widening of the right dens-lateral mass interval.*





IMAGING

General Features

- Best diagnostic clue
  - Abnormal T2 hyperintensity or gross discontinuity of spinal ligamentous structure
- Location
  - Anatomy
    - Anterior longitudinal ligament (ALL)
      - Broad ligament along ventral aspect of spinal column
      - Extends from anterior arch of C1 through sacrum
    - Posterior longitudinal ligament (PLL)
      - Extends along dorsal aspect of vertebral bodies within spinal canal
      - From body of C2 through to sacrum
    - Ligamentum flavum
      - Paired ligaments connecting lamina of adjacent vertebra
    - Interspinous ligaments
      - Thin ligamentous structures connecting spinous processes of adjacent vertebra
    - Supraspinal ligament
      - Fibrous cord connecting apices of spinous processes from C7 through sacrum
    - Zygapophyseal capsuloligamentous complex
      - Joint capsule and associated connective tissue
      - Resists distraction/subluxation of zygapophyseal (facet) joints

Radiographic Findings

- Radiography
  - Conventional radiographs insensitive to ligamentous injury
  - Ligamentous disruption may be inferred by observing disruption of normal vertebral alignment
  - Flexion-extension films
    - Instability
      - > 3.5 mm subluxation
    - May be performed in neurologically intact patient with neck pain, normal cervical spine motion, and normal radiographs
      - Abnormal cervical spine radiographs, limited range of motion (< 60° angular movement between flexion and extension): Pain or limited movement may result in inadequate examination
      - Many investigators report limited or no utility of flexion-extension studies in patients with normal cervical spine radiographs

CT Findings

- NECT
  - CT insensitive to ligamentous injury
  - Ligamentous disruption may be inferred with varying degrees of success by observing disruption of normal vertebral alignment

MR Findings

- T1WI
  - Intact ligamentous structures should be continuous and low in signal intensity
  - Discontinuity of ligament on T1WI indicative of disruption
- T2WI
  - T2WI FS, STIR
    - Favored by many sources to evaluate spinal ligamentous injury
    - Increased signal within ligamentous structure indicative of edema, hemorrhage, or inflammation (stretch injury, strain, partial tear)
    - Complete discontinuity of ligamentous structure on T2WI indicative of disruption
  - Variable performance of MR for ligamentous injury in different studies
    - When MR compared to surgical exploration and direct evaluation of ligamentous structure, different investigators report good, moderate, or poor correlation
    - Variable performance of MR for ligamentous injury may relate to timing of examination(s)
      - Edema or hemorrhage may not have fully developed before 24 hours post injury
      - Soft tissue edema resolves 72 hours after injury

Imaging Recommendations

- Best imaging tool
  - MR best to evaluate soft tissue structures of cervical spine
- Protocol advice
  - Sagittal T1, T2, and STIR; axial T1 and T2*

DIFFERENTIAL DIAGNOSIS

Crystal Arthropathy

- Calcium pyrophosphate dihydrate deposition disease (CPPD), hydroxyapatite deposition
- Thickened ligaments, heterogeneous signal

Abscess, Paraspinal

- Associated with spondylodiscitis
- Phlegmon (especially due to spinal tuberculosis) may spread over several levels deep to ALL, PLL

Calcific Tendinitis, Longus Coli

- Soft tissue thickening and T2 hyperintensity in cervical prevertebral soft tissues
- Inflammation lies ventral to ALL

PATHOLOGY

General Features

- Etiology
  - Soft tissues injuries of spine occur with variety of injury mechanisms
  - Each pattern of cervical or thoracolumbar injury has pattern of osseous changes which, in most cases, is more characteristic of injury mechanism than soft tissue changes
  - Cervicooccipital junction (C0-C2)
    - Jefferson fracture
      - Transverse atlantal ligament disrupted **or** avulsion fracture of tubercle from 1 of lateral masses of C1
    - Hangman's fracture

        - Disruption of intervertebral disc and PLL with type II injury
        - Type II: > 3 mm translation and > 10° angulation
      - Occipital condyle fracture
        - Avulsion fracture of inferomedial occipital condyle (Anderson and Montesano type III fracture)
        - Disruption of tectorial membrane and contralateral alar ligament
    - Cervical spine
      - Hyperextension-distraction
        - ALL rupture
        - PLL failure with more severe injury associated; posterior displacement of supradjacent vertebral body
      - Hyperflexion-compression
        - Injury to interspinous and supraspinous ligaments
        - PLL failure with more severe injury associated; posterior displacement of supradjacent vertebral body
      - Hyperflexion-distraction
        - Disruption of interspinous and supraspinous ligaments
        - Disruption of facet capsuloligamentous complex
        - PLL avulsion and varying degree of damage to ALL with facet dislocation(s)
      - Lateral hyperflexion
        - Failure of facet capsuloligamentous complex in tension, on side ipsilateral to applied force
      - Whiplash
        - Stretch or disruption of ligamentum flavum ALL and zygapophyseal capsuloligamentous complex found in cadaveric studies
        - Sprain or disruption of alar ligaments
        - Ligamentous injuries due to whiplash often occult on MR
    - Thoracolumbar spine
      - Burst fracture
        - Variable disruption of ALL, PLL, &/or interspinous and supraspinous ligaments
      - Thoracolumbar flexion-distraction fracture (Chance fracture variant)
        - Disruption of interspinous ligaments ± zygapophyseal capsuloligamentous complex
      - Fracture-dislocation
        - 3-column injury: Disruption of ALL, PLL, zygapophyseal capsuloligamentous complex, &/or interspinous and supraspinous ligaments
      - Hyperextension
        - Disruption of ALL
- Associated abnormalities
  - Traumatic disc herniation
  - Vertebral fractures

Gross Pathologic & Surgical Features

- Partial or complete disruption of ligamentous structure
- Edema, hemorrhage

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability after trauma
  - Spinal instability predisposing to progressive deformity or to impingement on spinal cord, nerve roots
    - Instantaneous (instability immediately following injury)
    - Delayed (> 20 days after injury)
  - Fractures, spinal malalignment on trauma radiographs
  - Risk of spinal cord injury varies with mechanism, severity of injury
- Other signs/symptoms
  - Closed head injury, polytrauma

Natural History & Prognosis

- Varies with degree of neurologic injury

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Ventral ligamentous injury of cervical spine without fracture is rare (< 0.7%)

SELECTED REFERENCES

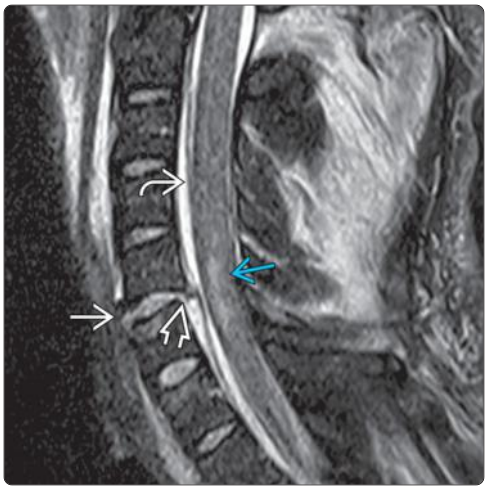
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(Left) *Sagittal STIR MR of a flexion-distraction injury shows edema due to disruption of the interspinous ligament injury at L2-L3. Note mild anterolisthesis of L2 on L3, anterior compression fracture of L3. Tiny epidural hematoma is present.* **(Right)** *Sagittal STIR MR of flexion-distraction injury shows edema/hemorrhage due to disruption of the interspinous ligament. The PLL is stripped from the T4 and T5 vertebral bodies with fracture of T6 with buckling of ALL.*





(Left) *Sagittal STIR MR shows hyperintensity between the neural arches of C1-C2 due to interspinous ligament disruption and thickening and hyperintensity dorsal to the dens, reflecting injury of the transverse ligament.* **(Right)** *Sagittal STIR MR shows disruption of the ALL and PLL, and posterior displacement of the ventral dural margin due to epidural hemorrhage. Cord contusion is also present as increased cord signal.*





(Left) *Sagittal T2* GRE MR of hyperflexion injury with bilateral jumped facets shows disruption of the PLL and a small fracture of C7 anteriorly. There is also contusion and hematoma of the cervical cord extending from C5 through C7.* **(Right)** *Coronal T2 MR of a child with a right occipital condyle fracture (not shown) shows partial avulsion of the right alar ligament from the occipital condyle.*

Occipital Condyle Fracture

KEY FACTS

TERMINOLOGY

- Bony disruption of occipital condyle

IMAGING

- Linear, comminuted, or avulsion-type fracture
  - Radiography insensitive due to overlapping structures
  - Lucency through occipital condyle on CT ± displaced bone fragment
  - Marrow edema on T1, STIR MR
  - Prevertebral or nuchal ligament edema on MR

TOP DIFFERENTIAL DIAGNOSES

- Accessory ossification center(s)
- Osteomyelitis
- Primary or secondary skull base neoplasm
- Rheumatoid arthritis

PATHOLOGY

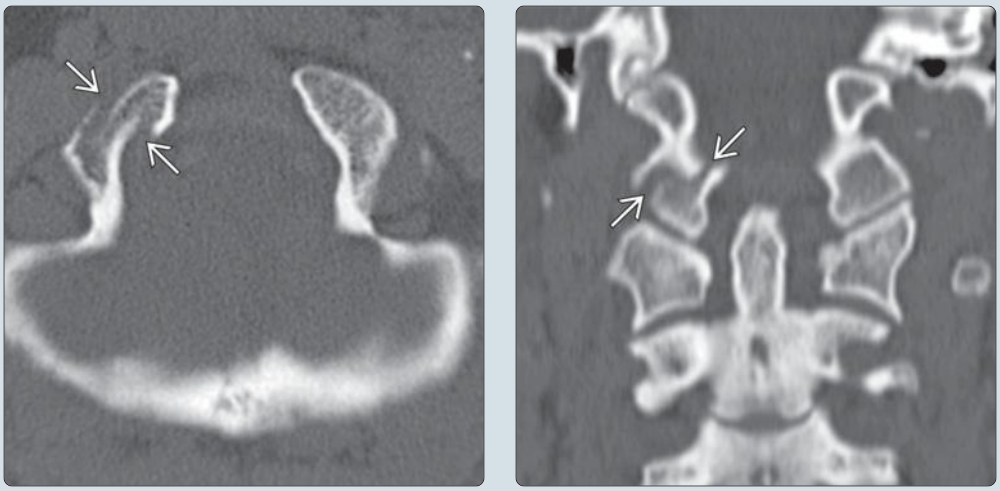
- High-energy blunt trauma, most often MVA
- Air bag deployment associated with craniocervical junction injury

CLINICAL ISSUES

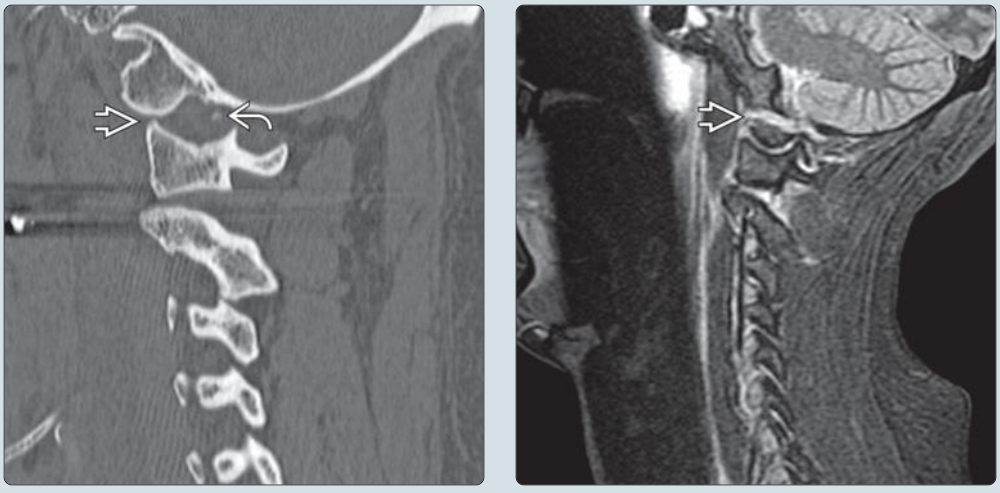
- Presenting symptoms usually related to severity of head injury
  - Most patients have mild/moderate ↓ GCS due to intracranial injury
  - Head injury is main determinant of outcomes
- Cranial nerve (CN) deficit(s) (up to 30%)
- Spasmodic torticollis from concomitant atlantoaxial rotatory fixation

DIAGNOSTIC CHECKLIST

- Easy fracture to overlook in severely injured trauma patients



(Left) *Axial bone CT shows a mildly displaced fracture of the right occipital condyle with cortical break ➡ and medial movement of the fracture fragment.* **(Right)** *Coronal reformatted bone CT in the same patient shows a lucent fracture line ➡ with minimal displacement. Articulation with C1 is preserved.*



(Left) *Sagittal reformatted NECT shows gross anterior displacement of the occipital condyle relative to the articulation with C1 ➡. A tiny portion of an occipital condyle fracture ➡ is also seen.* **(Right)** *Sagittal STIR MR shows hyperintense signal through a horizontal fracture ➡ of the occipital condyle. Signal in the adjacent marrow spaces is normal.*

TERMINOLOGY

Definitions

- Bony disruption of occipital condyle

IMAGING

General Features

- Best diagnostic clue
  - Lucency through occipital condyle on CT ± displaced bone fragment
- Location
  - Uni- or bilateral occipital condyle(s)
- Morphology
  - Linear, comminuted, or avulsion-type fracture
- Occipital condyle anatomy
  - Oriented obliquely forward and inward
  - Narrowest at midportion, slopes caudally from lateral to medial
  - Occipitoatloid articulations are cup-shaped joints = convex occipital condylar surfaces + concave superior surfaces of atlas articular facets
    - In coronal plane, joint slopes downward medially
  - Within base of each occipital condyles lie hypoglossal (anterior condyloid) canals containing CN12
  - Lateral to condyle & hypoglossal canal, posterior to carotid canal, is jugular foramen
    - Pars nervosa contains CN9, inferior petrosal sinus
    - Pars vascularis contains CN10, CN11, jugular bulb
  - Joint has fibrous capsule that blends into antero- & postero- atlantooccipital membranes
  - Alar ligaments extend laterally from superolateral dens to medial condyle
    - Very strong; bone often fails before ligaments
    - Checks lateral flexion & rotation, limiting cranial rotation with respect to atlas
  - Tectorial membrane attached to dorsal surfaces of C3 & C2 vertebral bodies, dens body, & anterior border of foramen magnum
    - Cephalad continuation of PLL
    - Checks extension, flexion, & vertical translation
  - Relationship of multiple neurovascular structures explains symptomatologies from occipital condyle fracture
    - Nerve root compression or stretching
    - Direct brain stem injury or vascular compromise

Radiographic Findings

- Radiography
  - Very insensitive (0/51 in 1 study) due to
    - AP → superimposition of maxilla and occiput
    - Lateral → superimposition of mastoid processes
    - Open mouth → difficult to obtain in unconscious, intubated, severely injured patients
  - May see prevertebral soft tissue swelling

Fluoroscopic Findings

- Flexion/extension fluoroscopy to assess for atlantooccipital ligamentous injury, instability
  - Following initial evaluation with CT to avoid unintentional displacement of undiagnosed fracture during flexion/extension

CT Findings

- Bone CT
  - Direct visualization of condylar fracture(s)
  - ± associated fractures of skull base, atlas, &/or axis
  - ± occipitoatlantal subluxation
- CTA
  - Evaluate vertebrobasilar arterial system for patency/injury

MR Findings

- STIR
  - Occipitoatloid joint subluxation, alar ligament disruption, joint effusion
  - Marrow edema
    - May be minimal in hyperacute period
  - Prevertebral or nuchal ligament edema
  - ± cord edema or hemorrhage
  - ± foramen magnum extradural, subdural hemorrhage
- MRA
  - Evaluate vertebrobasilar arterial system for patency/injury

Angiographic Findings

- Catheter angiography if CTA/MRA equivocal or unresolved concern for vertebral artery injury

Imaging Recommendations

- Best imaging tool
  - Unenhanced multidetector bone CT scan
  - MR for soft tissues
- Protocol advice
  - Thin slice (≤ 1 mm) axial CT in bone reconstruction algorithm
    - Sagittal and coronal reformatted images very useful
  - MR protocol: Include STIR in sagittal and coronal planes

DIFFERENTIAL DIAGNOSIS

Accessory Ossification Center(s)

- Anterior to occipital condyle
- Well corticated

Marrow Space Abnormality

- Infectious: Osteomyelitis
- Neoplastic
  - Metastatic neoplasms
  - Primary neoplasms
- Inflammatory: Rheumatoid arthritis

PATHOLOGY

General Features

- Etiology
  - High-energy blunt trauma, most often MVA
  - Air bag deployment associated with craniocervical junction injury
- Associated abnormalities
  - Intracranial hemorrhage, brain contusion
  - Cervical spine fracture (usually C1 or C2)
  - Thoracic, abdominal, & extremity injuries

Staging, Grading, & Classification

- Anderson & Montesano classification (1988)
  - Type I: Comminuted
    - Axial load injury with component of ipsilateral flexion; an impaction fracture
      - Comminuted condylar fracture without displacement
    - Least common
  - Type II: Extension of linear basilar skull fracture
    - Basilar skull fracture extending into occipital condyle(s), usually from direct blow to skull
    - Intact tectorial membrane & alar ligaments preserve stability
  - Type III: Avulsion of fragment
    - Avulsion of inferomedial condyle, usually result of severe contralateral flexion and rotation
    - May show medially displaced fragment, partial or complete disruption of tectorial membrane and contralateral alar ligament
    - Inferior clivus may be involved
    - Most common (75%)
- Tulli classification (1997)
  - Type 1 (most common): Nondisplaced; stable
  - Type 2A: Displaced fracture with intact ligaments; stable
  - Type 2B: Displaced fracture with any 1 or more criteria of radiographic instability
    - > 8° of axial occipitoatloid rotation to 1 side
    - > 1 mm of occipitoatloid translation
    - > 7 mm of overhang of C1 on C2
    - > 45° of axial rotation of C1-C2 to 1 side
    - > 4 mm of C1-C2 translation
    - < 13 mm distance between posterior body of C2 to posterior ring of C1
    - Avulsed transverse ligament with MR evidence of ligamentous disruption
- Hanson classification (2001)
  - Uses Anderson & Montesano classification as a base
  - Subdivides type III into stable & unstable

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Presenting symptoms usually related to head injury severity
  - Cranial nerve (CN) deficit(s) (up to 30%)
    - 1 or more CN9-C12 (most often 12)
    - 40% manifest days to months after trauma, possibly fragment migration or callus formation
    - Collet-Sicard syndrome = full CN9-C12 deficits
  - High cervical pain, impaired skull mobility
- Other signs/symptoms
  - Dysphagia from retropharyngeal hematoma
  - Spasmodic torticollis from concomitant atlantoaxial rotatory fixation
  - Hemi- or quadriparesis
  - Vertebrobasilar ischemia
- Clinical profile
  - No specific predictors for occipital condyle fracture
  - Most patients have mild/moderate ↓ GCS due to intracranial injury

Demographics

- Gender
  - M:F = 3:1
- Epidemiology
  - May be as high as 19% of patients with high-energy blunt trauma
    - True incidence, prevalence remain unknown
  - Increasingly reported, likely related to widespread use of CT & MR imaging

Natural History & Prognosis

- Functional recovery after unilateral CN deficit(s) is generally good
- Head injury is main determinant of outcome
- Limited & painful mobility of craniocervical junction can persist despite good osseous consolidation, especially with type III
  - Children generally have good osseous consolidation with normal & painless craniocervical mobility

Treatment

- By Anderson & Montesano classification
  - Type I & II are considered stable (tectorial membrane & contralateral alar ligament intact)
    - Treated conservatively → semirigid or rigid collar
  - Type III is potentially unstable injury if ligamentous injury present
    - Assess for instability → flexion/extension
    - Treated with rigid collar, halo traction vest, or surgical fixation
- By Tulli classification
  - Type 1: Require no specific treatment
  - Type 2A: May be treated with a rigid collar
  - Type 2B: Surgical instrumentation or halo traction
- Surgical therapy is controversial
  - Possible indications → neurovascular decompression &/or stabilization
  - Usually posterior fusion (occipitoatlantoaxial arthrodesis)

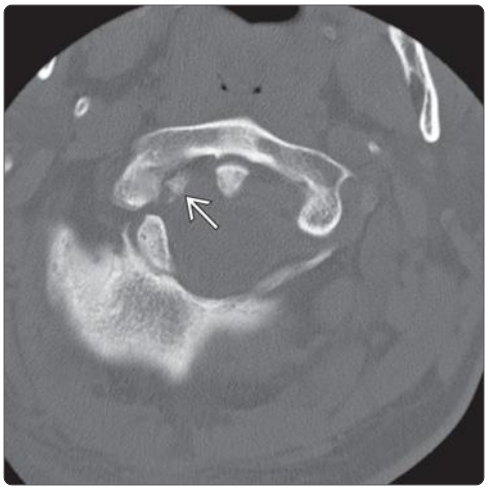
DIAGNOSTIC CHECKLIST

Consider

- Easy fracture to overlook in severely injured trauma patients

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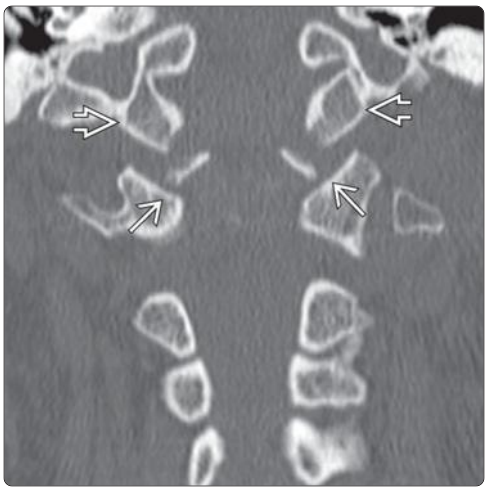


(Left) *Axial NECT shows a small avulsion fracture ⇨ off of the inferior right occipital condyle, medially displaced.* **(Right)** *Coronal reformatted NECT in the same patient again shows the small avulsion fracture ⇨ off of the inferior right occipital condyle. There is also an oblique fracture through the right lateral mass of C3 ⇨.*





(Left) *Axial NECT shows avulsion of the right occipital condyle ⇨, which is rotated and displaced inferomedially, residing supradjacent to the odontoid process.* **(Right)** *Coronal reformatted NECT in the same patient shows the right occipital condyle avulsion ⇨, rotated and displaced inferomedially, residing supradjacent to the tip of the dens.*





(Left) *Coronal NECT (bone window) shows symmetrical bilateral avulsion fractures ⇨ of the occipital condyles with fragments inferior and posterior to the condyles themselves ⇨. There is widening of the C0-C1 joint with atlantooccipital dislocation.* **(Right)** *Coronal reformatted bone CT shows a portion of a comminuted occipital fracture ⇨ extending into the margin of the left occipital condyle and through hypoglossal canal.*

Jefferson C1 Fracture

KEY FACTS

TERMINOLOGY

- Fracture(s) of C1 ring

IMAGING

- Multiple fractures of C1 arch (2, 3, and 4 part fractures)
- Combined offset of lateral masses of C1 relative to lateral margins of C2 ≥ 7 mm suggests interruption of transverse ligament
- Avulsion fragment off inner C1 pillar at insertion of transverse ligament indicates unstable fracture
- Widening of atlantoaxial interval
  - ≥ 4 mm concerning for interruption of transverse ligament
  - ≥ 7 mm presumed interruption of transverse ligament
- Associated C2 fracture (hangman's fracture, odontoid fracture)
- Fractures at lower levels not uncommon
- May see T2 hyperintense edema if cord contusion present

TOP DIFFERENTIAL DIAGNOSES

- Congenital variants, clefts, malformations of atlas
- Rotational malalignment of atlas, axis pillars
- Pseudospread of atlas in children

PATHOLOGY

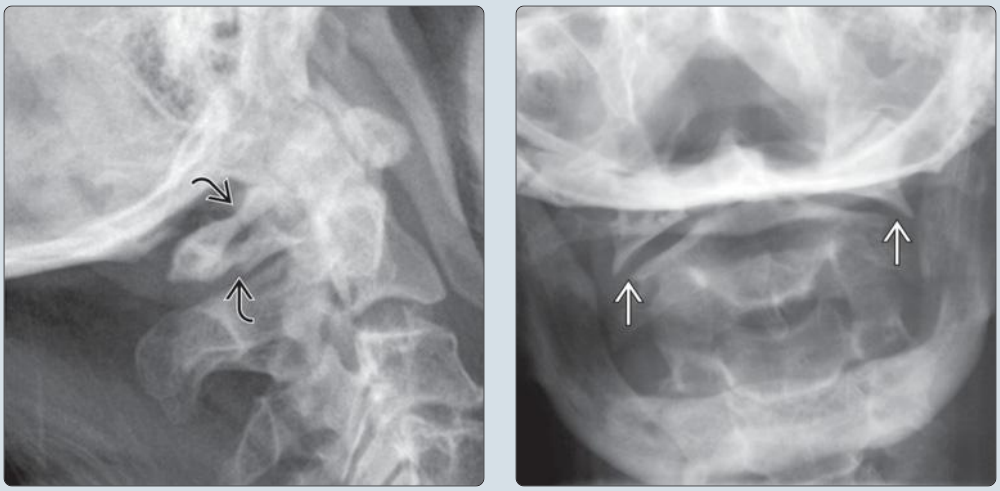
- Force transmitted down through occipital condyles onto sloped C1 pillars with head and neck rigidly erect
- Transverse ligament often intact
- If transverse ligament is interrupted, stability of fracture depends on integrity of alar ligaments

CLINICAL ISSUES

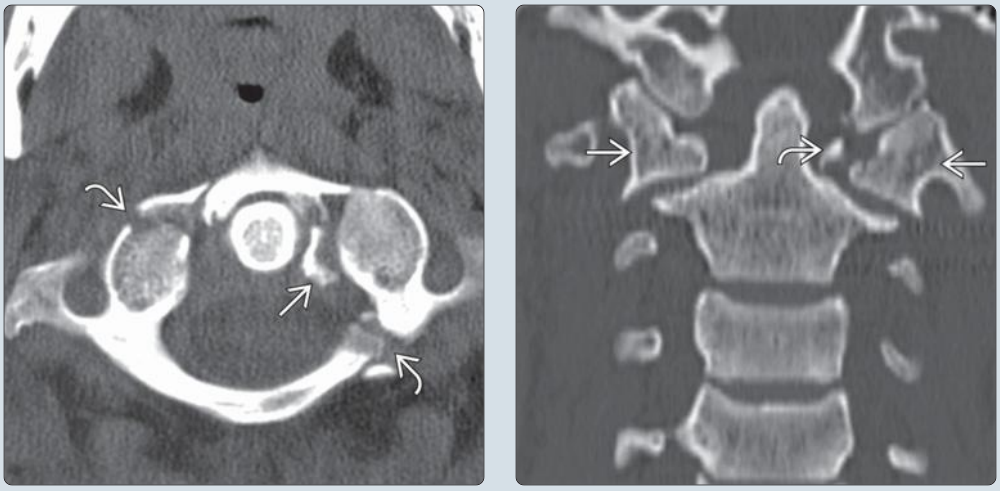
- Neurologic signs uncommon unless unstable fracture, injury at another level, or vascular injury

DIAGNOSTIC CHECKLIST

- Important to evaluate lower levels for additional fractures



(Left) *Lateral radiograph of the craniovertebral junction shows faint lucencies in the posterior arch of C1.* **(Right)** *Coronal radiograph, in which the interval between the dens and the lateral masses of C1 is obscured by the occipital bone, shows lateral displacement of the outer margins relative to C2.*



(Left) *Axial NECT shows fractures extending through the anterior and posterior arches of C1. A small bony fragment demonstrates the site of an avulsion fracture at the attachment of the transverse ligament.* **(Right)** *Coronal reformatted CT shows lateral displacement of both C1 lateral masses relative to the occipital condyles and C2 lateral masses. Also seen is a bony fragment due to transverse ligament tubercle avulsion.*

TERMINOLOGY

Synonyms

- Atlas burst fracture

Definitions

- Fracture(s) of C1 ring

IMAGING

General Features

- Best diagnostic clue
  - Lateral displacement of both articular masses of C1 relative to margins of C2 on open mouth radiograph

Radiographic Findings

- Radiography
  - Bony defects of C1
  - Widening of distance between odontoid and C1 lateral masses on open mouth view
    - Normal rotation may produce apparent offset of pilar, simulating fracture
    - Combined offset of lateral masses of C1 relative to lateral margins of C2 ≥ 7 mm suggests interruption of transverse ligament
      - Potentially unstable fracture
  - Widening of atlantoaxial interval
    - ≥ 4 mm concerning for interruption of transverse ligament
    - ≥ 7 mm presumed interruption of transverse ligament
      - Potentially unstable fracture
  - Upper prevertebral soft tissue swelling
  - Associated C2 fracture (hangman's fracture, odontoid fracture)
  - Fractures at lower levels not uncommon

Fluoroscopic Findings

- Subluxation if unstable

CT Findings

- NECT
  - Axial CT defines components of fracture to best advantage
    - Multiple fractures of C1 arch (2, 3, and 4 part fractures)
    - Avulsion fragment off inner C1 pillar at insertion of transverse ligament indicates unstable fracture
  - May demonstrate various patterns of arch disruption
  - May demonstrate hyperdensity in epidural space if bleeding occurs
- CTA
  - Loss of vertebral artery integrity if vertebrobasilar vascular syndrome present

MR Findings

- T1WI
  - Prevertebral soft tissue swelling anterior to C1
  - Disruption of cortical margins of C1
- T2WI
  - Edema in prevertebral soft tissues
  - May see hyperintense cord edema if contusion is present
    - Low signal within cord edema concerning for parenchymal hemorrhage
- MRA
  - Vertebral artery injury, if present, with dissection or occlusion

Angiographic Findings

- Useful if CTA/MRA equivocal or for persistent concern for vertebral artery injury
- Endovascular intervention

Imaging Recommendations

- Best imaging tool
  - Unenhanced multidetector CT scan
- Protocol advice
  - Any lateral spread of C1 pillars on open mouth x-ray view requires CT
  - Thin-slice (≤ 1 mm) axial CT in bone reconstruction algorithm
    - Fastest possible scan times to minimize motion
    - Sagittal and coronal reformatted images often helpful
      - Coronal especially for assessing lateral subluxation of lateral masses
  - Evaluate entire cervical spine as well as upper thoracic spine
    - Associated fractures occur in 24-48% of cases
  - Axial and sagittal T1WI and T2WI, sagittal STIR to evaluate fracture morphology, displacement, ligamentous injury, soft tissue edema

DIFFERENTIAL DIAGNOSIS

Congenital Variants, Clefts, Malformations of Atlas

- May show 1-2 mm offset of C1 pillars from those of C2
- Clefts found in 4% of posterior arches, 0.1% of anterior arches
- 97% of posterior clefts are midline, 3% through sulcus of vertebral artery
- Various deficiencies of arch development can be seen
- Most are partial hemiaplasias of posterior arch
- Clefts, congenital defects show smooth or well-corticated edges

Rotational Malalignment of Atlas, Axis Pillars

- Generally seen unilaterally, with rotation and abduction of head

Pseudospread of Atlas in Children

- Common finding in children 3 months to 4 years of age evaluated for minor trauma
- Seen in 90% or more of 2 year olds
- Caused by disparity in growth rates of atlas and axis
- Jefferson fracture rare in young children, greater plasticity, synchondroses of C1 arch serve as "buffer"

PATHOLOGY

General Features

- Etiology
  - Axial compressive force applied to skull vertex
  - Force transmitted down through occipital condyles onto sloped C1 pillars with head and neck rigidly erect
    - Wedge effect

- Transverse ligament often intact
- If transverse ligament is interrupted, stability of fracture depends on integrity of alar ligaments

- Associated abnormalities
  - Fractures at other levels
  - Vertebral artery injury: Dissection or occlusion
  - Cord injury (rare)

Staging, Grading, & Classification

- Type I: Bilateral single arch fractures
- Type II: Concurrent anterior and posterior arch fractures
  - Includes classic 4-point Jefferson fracture
- Type III: Lateral mass fracture

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Upper neck pain after compression trauma (e.g., diving)
  - Cervical muscle spasm
  - Limited range of motion
  - Head tilt
- Other signs/symptoms
  - Neurologic signs uncommon unless unstable fracture, injury at another level, or vertebral artery injured
    - Upper cord injury
      - Potentially catastrophic, including apnea
    - PICA injury, especially Wallenberg syndrome
- Clinical profile
  - Trauma victim
  - Upper neck pain

Demographics

- Epidemiology
  - C1 fractures represent 6% of all vertebral injuries
  - 1/3 of C1 fractures conform to classic burst Jefferson fracture
  - Rare in infants, young children

Natural History & Prognosis

- Stable fracture
  - Healing with conservative therapy in majority of cases

Treatment

- Nondisplaced isolated anterior or posterior atlas arch fractures and fractures of atlas lateral mass
  - External cervical immobilization
  - Rigid collars, suboccipital mandibular immobilizer braces, and halo ring-vest orthoses
  - 8-12 weeks
  - 96% rate of healing
- Combined anterior and posterior arch fractures of atlas with intact transverse ligament
  - Rigid collar, suboccipital mandibular immobilizer brace, or halo orthosis
  - 10-12 weeks
- Combined anterior and posterior arch fractures of the atlas with transverse ligament disruption
  - Halo orthosis for 12 weeks or surgical stabilization and fusion

DIAGNOSTIC CHECKLIST

Consider

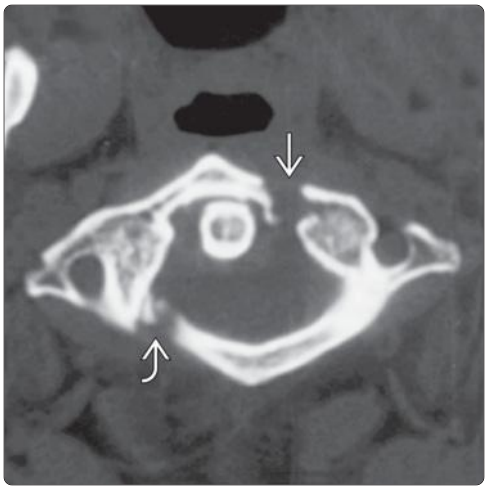
- Routine CT of cervical spine in trauma victims with severe neck pain
- Evaluate for extension of fracture into foramina transversarium
- Very important to evaluate lower levels for additional fractures

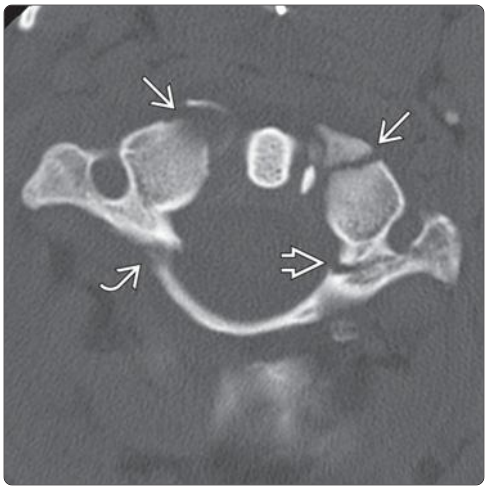
Image Interpretation Pearls

- Well-corticated edges of midline C1 arch defects are likely congenital clefts
- 1-2 mm offset of C1 lateral masses vs. C2 on open mouth view in infants may be normal variant

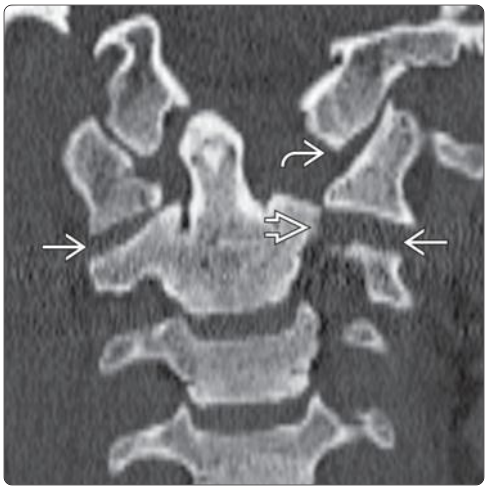
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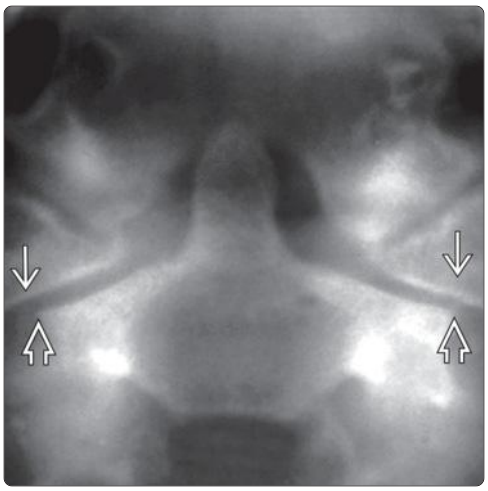


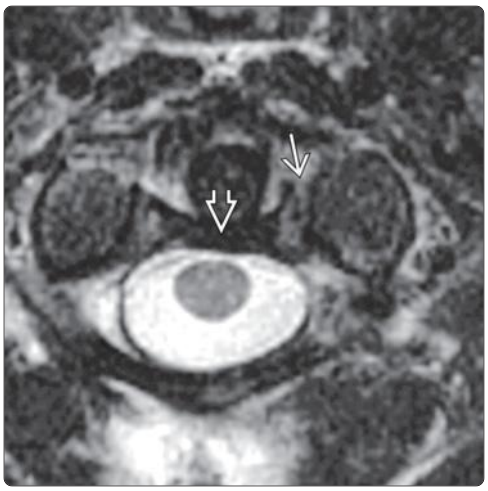
(Left) *Axial NECT shows a C1 burst fracture with displaced fractures of the anterior and posterior arches of C1 with lateral displacement of the fragments.* **(Right)** *Axial NECT shows symmetric fractures of the anterior arch, of the right lateral aspect of the posterior arch, and through the left lateral mass extending into the margin of the foramen transversarium.*





(Left) *Coronal reformatted CT shows displaced lateral masses of C1. The tip of the odontoid is at the margin of the foramen magnum due to loss of height at C1. Note disruption of the occiput/C1 articulation and C1-C2 facet joints. Depressed fracture of the left C2 articular pillar is also present.* **(Right)** *Sagittal reformatted bone CT shows a component of a C1 burst fracture through the right lateral mass, highlighting the role axial loading from occipital condyles plays in this injury mechanism.*





(Left) *Frontal tomographic image through the depth of the odontoid shows lateral displacement of the C1 lateral masses relative to the C2 lateral masses.* **(Right)** *Axial T2WI MR in a patient with a burst fracture of the C1 shows an intact transverse ligament. Note the left-sided bony avulsion fracture that does not involve the substance of the ligament.*

Atlantoaxial Rotatory Fixation

KEY FACTS

IMAGING

- Abnormal rotatory motion of C1 with respect to C2 defined by 3-position CT scan

TOP DIFFERENTIAL DIAGNOSES

- **Etiologies of AARF**
  - Trauma
  - Nasopharyngeal infection (Grisel syndrome)
  - Prior head and neck surgery

PATHOLOGY

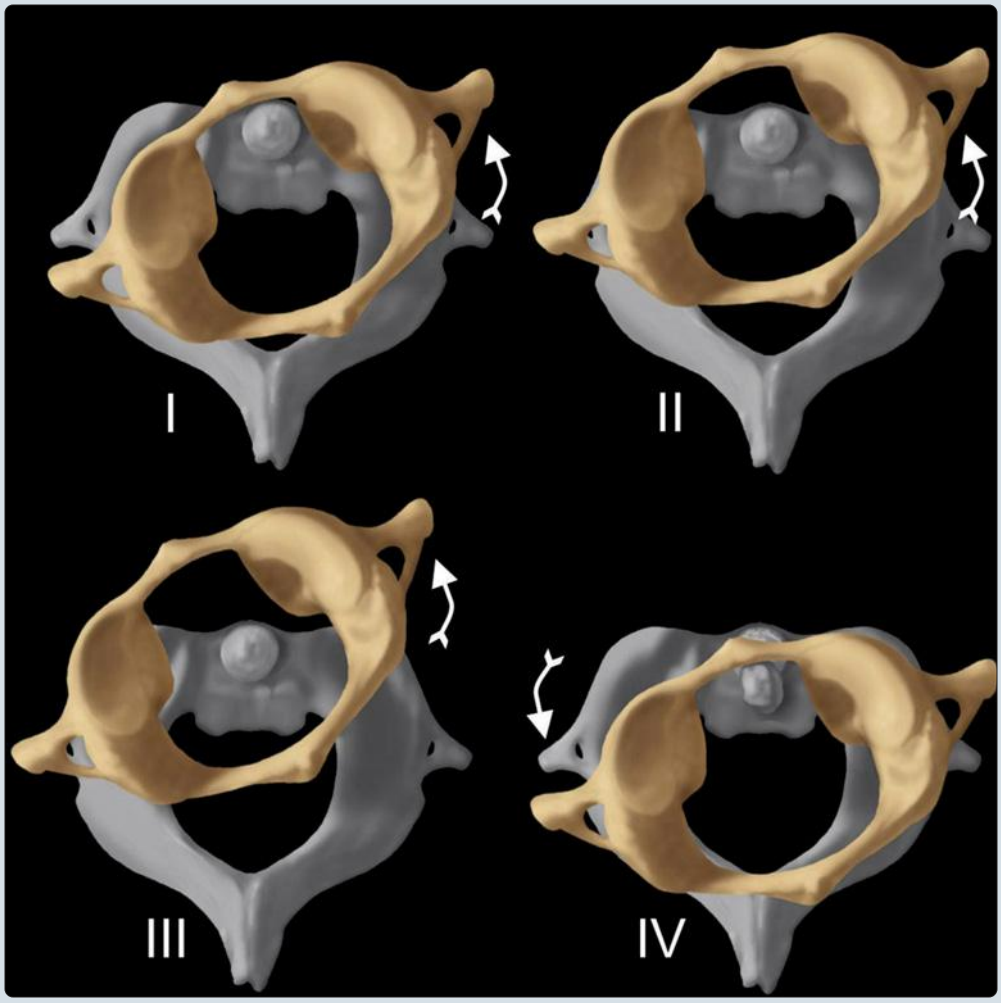
- **Pang type I AARF**: Unaltered or locked C1-C2 coupled configuration regardless of corrective counter-rotation
- **Pang type II AARF**: Reduced C1-C2 separation angle with forced correction, but C1 does not cross C2
- **Pang type III AARF**: Show C1-C2 crossover, but only with head turned far to opposite side

CLINICAL ISSUES

- Persistently rotated head that is painful during attempts at correction (painful torticollis)
- Head typically laterally flexed on side opposite to pointing chin ("cock-robin" appearance)

DIAGNOSTIC CHECKLIST

- **Fielding-Hawkins** type III, IV involve traumatic rupture of transverse atlantal and other stabilizing ligaments
  - Emergent and serious injuries requiring immediate surgical intervention to protect cord
  - These more serious injuries are separate from pure rotatory fixations, which are never acutely unstable (as in Pang type I-III AARF)



Graphic shows Fielding and Hawkins classification (1977) of atlantoaxial rotatory fixation: Type I shows rotatory displacement of C1 without ligamentous abnormality; type II demonstrates 3–5 mm anterior displacement of C1 and is associated with transverse ligament abnormality; type III shows > 5 mm of anterior displacement of C1 on C2 and is associated with deficiency of transverse and alar ligaments; type IV is the least common and shows C1 displacement posteriorly. Arrows show direction of movement.

TERMINOLOGY

Abbreviations

- Atlantoaxial rotatory fixation (AARF)

Synonyms

- Atlantoaxial rotatory subluxation, rotary subluxation

Definitions

- Rotatory fixation or subluxation: Persistent rotational deformity of C1-C2 complex with resultant torticollis and head in "cock-robin" position; normal atlantodental interval (ADI)
- Rotatory dislocation: More severe injury with anterior displacement of lateral mass of C1 relative to C2 with widening of ADI

IMAGING

General Features

- Best diagnostic clue
  - Abnormal rotatory motion of C1 with respect to C2 defined by 3-position CT scan
- Location
  - C1-C2 articulation

Radiographic Findings

- Radiography
  - Difficult to diagnose
  - Lateral mass of C1 rotated anteriorly, appears wider and closer to midline (medial offset)
  - Opposite lateral mass is narrower and away from midline (lateral offset)

CT Findings

- NECT
  - Abnormal motion curve of C1 and C2 given 3-position CT protocol

Imaging Recommendations

- Protocol advice
  - 3 separate scans covering occiput to C2
    - Head in presenting position, undisturbed (designated as P position)
    - Head turned to zero position by examiner (designated as P0 position)
    - Head turned to side opposite to presenting position as far as tolerable (designated as P_)
      - By convention, side toward chin was initially pointing is assigned + sign
      - All C1 angles at P position are positive; all angles on opposite side are negative
  - **Motion curve**
    - Angles to right of 0 on CT (chin turned to left) are designated (+); angles on opposite side are (-)
      - Sagittal axis of C1 is line between anterior and posterior tubercles
      - Sagittal axis of C2 is line joining midline body prominence and cleft of bifid spine
      - Angles made between sagittal axes of C1 and C2 and vertical 0 degrees
    - Separation angle then calculated for each of 3 positions: C1 degrees minus C2 degrees

DIFFERENTIAL DIAGNOSIS

Etiologies of AARF

- **Trauma**
  - More severe forms caused by violent trauma
  - Frequent coexistence of fractured clavicle on opposite side of chin
- **Nasopharyngeal infection (Grisel syndrome)**
  - Many purported mechanisms, including spread of infection to CV joints; synovial inflammation
- **Prior head and neck surgery**
  - Heads rotated or hyperextended with otopharyngeal inflammation; general anesthesia and muscle relaxants
- **Mimics**
  - Muscular torticollis related to overactivity or contracture of sternomastoid muscle on side opposite to chin
  - CV junction segmentation anomalies with instability
  - Os odontoideum
  - Developmental instabilities (Down syndrome, mucopolysaccharidoses, neurofibromatosis)

PATHOLOGY

Staging, Grading, & Classification

- **Fielding and Hawkins (1977)**
  - **Type I**: Rotatory fixation without anterior displacement of atlas (displacement of < 3 mm)
    - Most common type
  - **Type II**: Rotatory fixation with anterior displacement of atlas from 3-5 mm
    - Associated with abnormality of transverse ligament
  - **Type III**: Rotatory fixation with displacement > 5 mm
    - Associated with deficiency of transverse and alar ligaments
  - **Type IV**: Rotatory fixation with posterior displacement
    - Rare
- **Pang (2005)**
  - **5 distinct groups** based on motion curves obtained from 3-position CT
    - Does not include cases of concomitant acute translational instabilities of C1 on C2 as in Fielding-Hawkins types III, IV
  - **Type I AARF**: Unaltered or locked C1-C2 coupled configuration regardless of corrective counter-rotation
    - Motion curve is horizontal in upper 2 quadrants of C1C2 motion template
  - **Type II AARF**: Reduced C1-C2 separation angle with forced correction, but C1 does not cross C2
    - Motion curve slopes downward from right to left in upper quadrants, but does not traverse x-axis
  - **Type III AARF**: Show C1-C2 crossover but only with head turned far to opposite side
    - Motion curve traverses x-axis left of C1 = -20°
  - **Group IV**: Normal dynamics in muscular torticollis without injury to C1-C2 joints
  - **Group V**: Diagnostic gray zone
    - Motion curve shows features between normal and type III AARF

Pang Classification Summary

Group	C1-C2 Motion	Interpretation	Designation
I	Horizontal motion curve; reduction of C1-C2° < 20%	Bony interlock; most severe form of C1-C2 "stickiness"	Type I AARF
II	Reduction of C1-C2° > 20%; motion curve slopes right to left but does not cross x-axis; C1 does not cross C2	Not locked, but unyielding motion "stickiness"	Type II AARF
III	C1 crosses C2 left of C1 = -20° (far from normal null point of 0°)	Moderate abnormal "stickiness"	Type III AARF
IV	Motion curve normal	Normal rotational dynamics	Muscular torticollis
V	Curve slightly left of normal curve; C1 crosses C2 between -8° and -20°	Spasm and mild joint "stickiness"	Diagnostic gray zone; conservative tx

Adapted from Pang D et al: Atlantoaxial rotary fixation: parts 1-3. Neurosurgery. 55(3):614-25, 2004; 57(5):941-72, 2005.

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Persistently rotated head that is painful during attempts at correction (painful torticollis)
  - Head typically laterally flexed on side opposite to pointing chin ("cock-robin" appearance)
    - Strong resistance if patient's head manually counter-rotated to opposite side of chin
    - Little pain or resistance when head rotated further toward side of chin
      - Unique finding in all other body part joint subluxations where displacement along direction of dislocation always causes pain
- Other signs/symptoms
  - Palpable spasm in sternocleidomastoid muscle on side of chin
  - Occipital neuralgia

Demographics

- Age
  - Childhood (18 months to 18 years)
- Gender
  - M > F

Natural History & Prognosis

- Treatment in acute phase gives best outcome (Pang type I-III)
- With short treatment duration of < 4 months, in one study no acute patients required halo fixation or surgical fusion
- Subacute patients have worse outcome with longer treatment course; many of these patients require halo fixation
- Chronic patients have worst prognosis with Pang type I, II AARF with high percentage of persistent abnormal motion requiring fusion
- Best overall outcome with Pang acute type III AARF, and worst outcome chronic type I AARF

Treatment

- Acute and subacute patients immobilized with Guilford brace for 3 months with 1st successful reduction
- Recurrence may be treated with traction and Guilford bracing for additional 3 months
- 2nd recurrence treated with halo for 3 months
- 3rd recurrence or recurrence after halo immobilization treated with posterior C1-C2 fusion

DIAGNOSTIC CHECKLIST

Consider

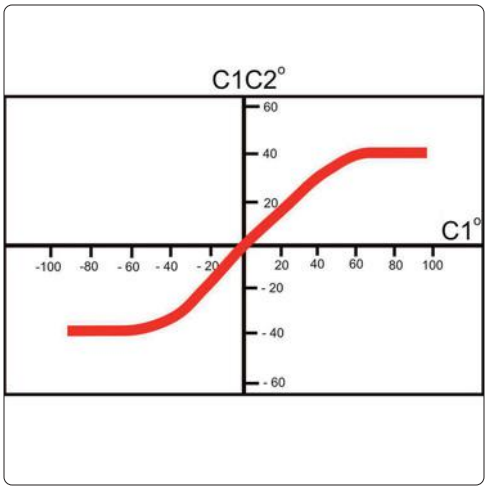
- Fielding-Hawkins type III, IV involve traumatic rupture of transverse atlantal and other stabilizing ligaments
  - Emergent and serious injuries requiring immediate surgical intervention to protect cord
  - These more serious injuries are separate from pure rotatory fixations, which are never acutely unstable

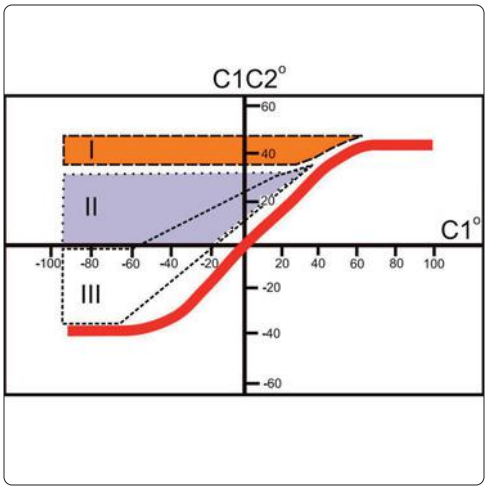
Image Interpretation Pearls

- Children with painful torticollis must receive 3-position CT
  - Secure diagnosis of AARF
  - Grade severity of condition via dynamic motion curve

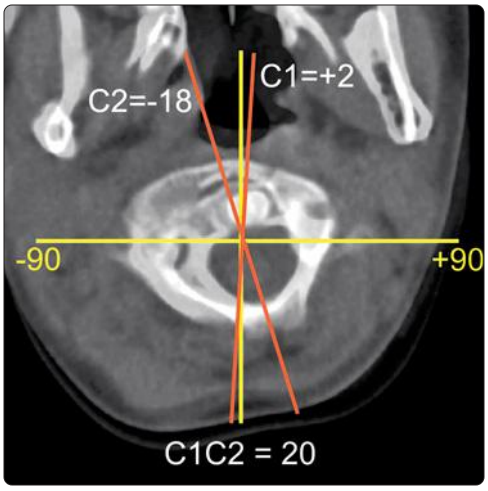
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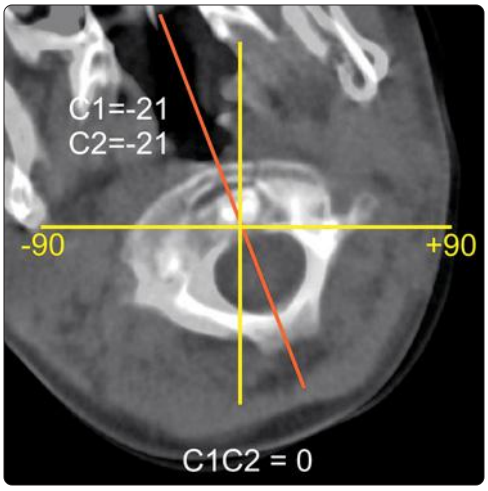
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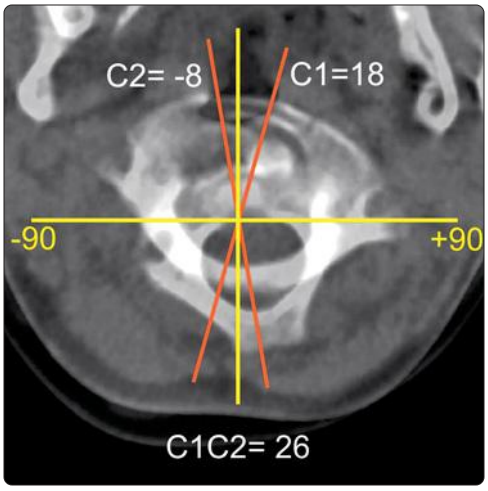


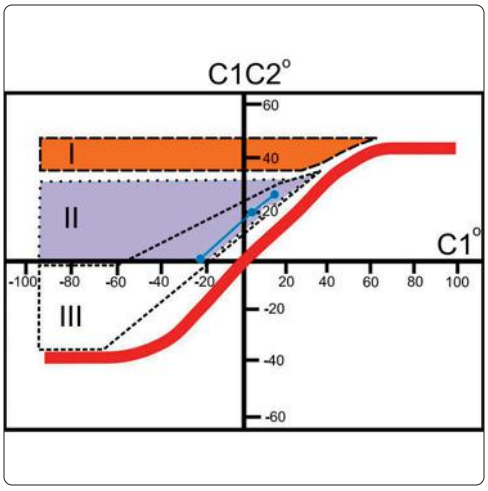
(Left) *Normal motion curve shows relationship between C1 and C2 during axial rotation within the full range of motion from right to left. X-axis is C1 angle (head position). Y-axis is C1-C2 angle, which is the separation angle between C1 and C2 (after Pang 2004).* **(Right)** *Schematic shows general regions for pathologic motion curves of the 3 types of Pang AARF (I-III) derived from 3-position CT scans. Classification depends upon clinical findings since overlap is present in regions II, III (after Pang 2005).*





(Left) *Axial NECT (neutral position) shows a 3-position study in a patient with Pang type II AARF. Slices through C1 and C2 have been overlaid. C1 angle is +2; C2 angle is -18 with a C1C2 difference of +20.* **(Right)** *Axial NECT (right head turn) shows a 3-position study in a patient with Pang type II AARF. C1 angle is -21; C2 angle is also -21 with a C1C2 difference of 0.*





(Left) *Axial NECT (left head turn) shows a 3-position study in a patient with Pang type II AARF. Slices through C1 and C2 have been overlaid. C1 angle is +18; C2 angle is -8 with a C1C2 difference of +26.* **(Right)** *Plot of 3 positions from head CT study (blue points) shows motion curve falls within general region of Pang type II AARF.*

(Left) *Fielding type I rotatory fixation: AP scout from a NECT shows the head rotated severely to the right with the right side of C1 closely applied to the odontoid process* ➡. **(Right)** *Fielding type I rotatory fixation: Axial CT with the head rotated to the right shows severe rotation of C1 with respect to C2, but a normal atlantodental interval and no fracture.*



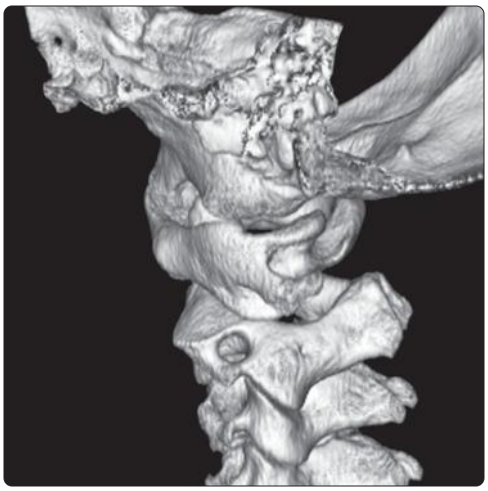


(Left) *Fielding type I rotatory fixation: Oblique CT due to the patient's rotation shows the rotated C1 arch with respect to the normal morphology of the odontoid process. Normal atlantodental interval is again seen.* **(Right)** *Fielding type I rotatory fixation: Anterior view from a volume CT reconstruction shows the morphology of the rotated C1 and morphologically normal odontoid. Axial of rotation is about the anterior atlantodental joint, consistent with intact ligamentous complex.*

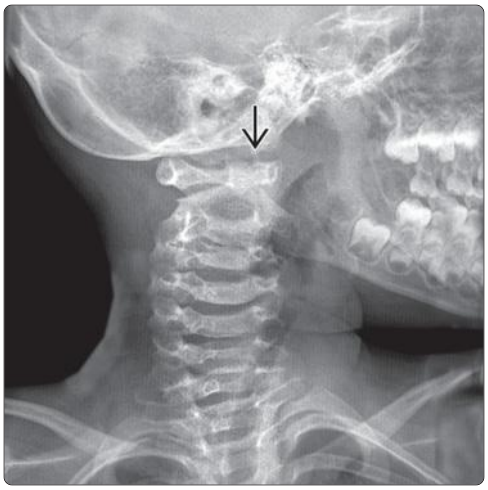




(Left) *Fielding type I rotatory fixation: Lateral view from a volume CT reconstruction shows the morphology of the rotated C1 and morphologically normal odontoid. Axial of rotation is about the anterior atlantodental joint, consistent with intact ligamentous complex.* **(Right)** *Fielding type I rotatory fixation: Coronal CT following reduction of the fixation under anesthesia shows normal relationship of C1 to C2.*

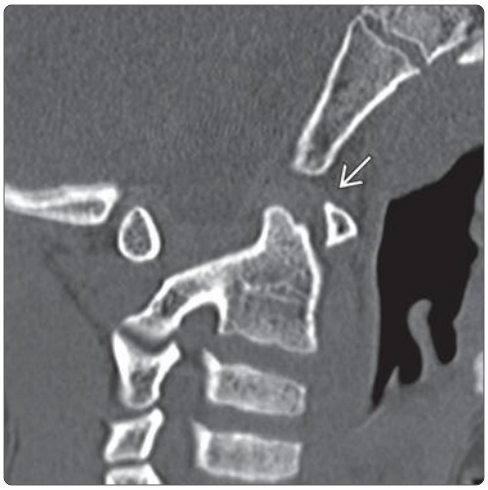


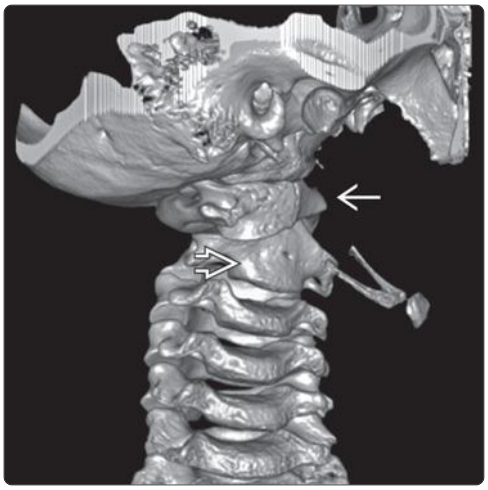






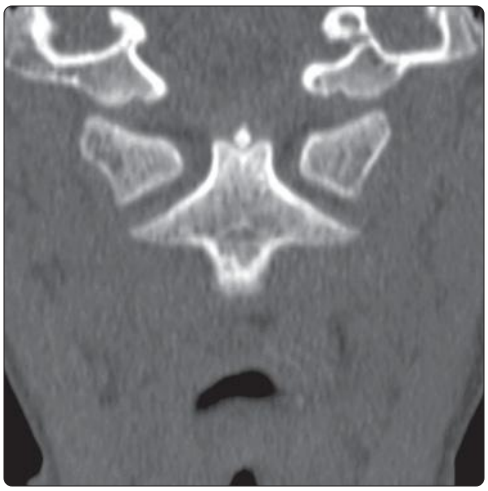
(Left) *Fielding-Hawkins type I atlantoaxial rotatory fixation: Initial plain radiograph shows rotation of the head to the left without fracture. Note the typical appearance of the rotated C1 being closely applied to the left side of C1 ⇨, with widening of the right C1-C2 spacing.* **(Right)** *Fielding-Hawkins type I atlantoaxial rotatory fixation: Lateral radiograph shows the head turned severely to the left, nearly 90°.*





(Left) *Fielding-Hawkins type I atlantoaxial rotatory fixation: Sagittal CT reconstruction is difficult to interpret due to the rotation, but no fracture is seen and the anterior atlantodental interval is normal ➡.* **(Right)** *Fielding-Hawkins type I atlantoaxial rotatory fixation: 3D CT reconstruction nicely visualizes the markedly left rotated C1 ➡ with respect to the mildly rotated C2 ➡.*





(Left) *Typical pattern of a Fielding-Hawkins type I atlantoaxial rotatory fixation: Following reduction under anesthesia, the lateral scout view from the CT study is normal.* **(Right)** *Typical pattern of a Fielding-Hawkins type I atlantoaxial rotatory fixation: Coronal CT following reduction shows the normal relationship of C1 lateral masses to the C2 body.*

Odontoid C2 Fracture

KEY FACTS

TERMINOLOGY

- Type I: Avulsion fracture from tip of odontoid at insertion of alar ligament
  - Usually stable injury
  - Usually seen in conjunction with more extensive craniocervical injury
- Type II: Transverse fracture through base of odontoid
  - Most likely to progress to nonunion without surgical fusion
- Type III: Oblique fracture extending from base of odontoid into body of C2

IMAGING

- Direct visualization of fracture line on radiography
  - Soft tissue swelling anterior to C2 in acute cases
  - Displacement of dens, C1 on lateral film
- CT protocol: Thin slice (1 mm or less) multidetector CT, fast scan time to minimize motion
  - Sagittal and coronal reformatted images mandatory
- MR
  - Effacement of thecal sac on MR due to displaced fracture
  - Cord injury, if present, hyperintense on T2WI
  - Fractures without compression &/or fractures with distraction do not reliably generate marrow edema and can lead to false-negative MR imaging

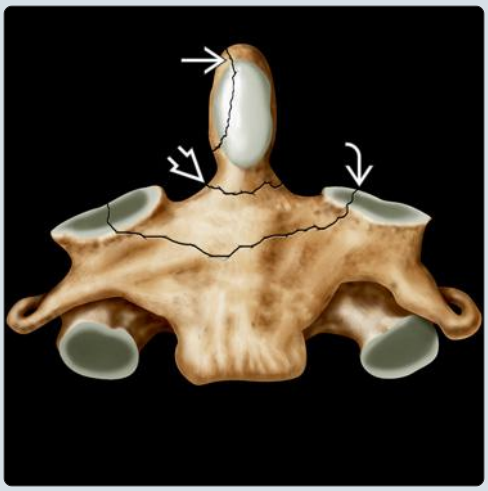
TOP DIFFERENTIAL DIAGNOSES

- Os odontoideum
- Congenital variation: 3rd occipital condyle (condylus tertius)
- Rheumatoid arthritis: C1/C2 subluxation
- Pathologic C2 fracture
- Ossiculum terminale persistens

PATHOLOGY

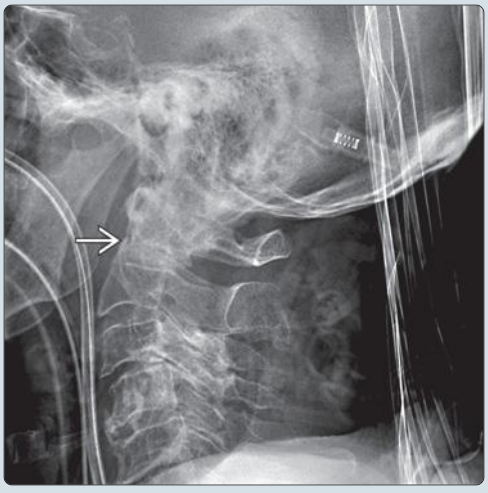
- Osteoporosis in elderly predisposes to type II fracture and nonunion

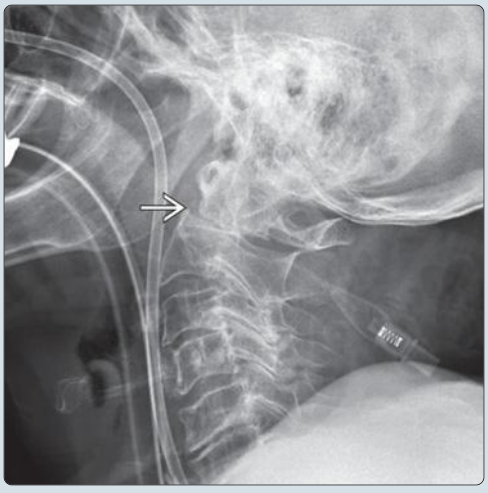
(Left) *Anterior graphic shows an avulsion fracture through the tip of the odontoid ➡ (type I), transverse fracture at the base of the odontoid ➡ (type II), and an odontoid fracture extending through the body of C2 ➡ (type III).* **(Right)** *Sagittal reformatted bone CT shows a nondisplaced fracture ➡ through the tip of the odontoid (type I odontoid fracture).*





(Left) *Lateral radiograph shows a subtle type II odontoid fracture through the base of the dens ➡, which displaced posteriorly with slight extension reflecting an unstable injury.* **(Right)** *Lateral projection radiograph in slight extension shows a fracture through the base of the odontoid ➡ (type II odontoid fracture) with displacement of the distal fracture fragment.*





TERMINOLOGY

Synonyms

- Dens fracture

Definitions

- Type I: Avulsion fracture from tip of odontoid at insertion of alar ligament
- Type II: Transverse fracture through base of odontoid
- Type III: Oblique fracture extending from base of odontoid into body of C2

IMAGING

General Features

- Best diagnostic clue
  - Lateral radiograph: Anterior or posterior displacement of C1 arch vs. C2, with prevertebral soft tissue swelling
  - Fracture visible on open mouth (dens) view

Radiographic Findings

- Radiography
  - Type I: Oblique fracture fragment at rostral aspect of odontoid on frontal view
  - Type II
    - Lucent fracture line through base of odontoid
    - Posterior displacement of C1 arch and odontoid relative to C2 body
  - Type III
    - Lucent fracture line through base of odontoid extending into C2 body
    - Fracture may extend into superior articular surfaces of C2
    - Interruption of C2 rings on lateral film
  - All: Swelling of prevertebral soft tissues

Fluoroscopic Findings

- Evaluate stability of fusion postoperatively

CT Findings

- NECT
  - Soft tissue swelling anterior to C2 in acute cases
  - Lucent fracture line through tip of odontoid (type I) or base of odontoid (type II) ± extent into C2 body (type III)
- CTA
  - Readily available screening for cerebrovascular injury in appropriate cases

MR Findings

- T1WI
  - Abnormal low T1 marrow signal due to osseous edema
  - Cortical disruption
  - May directly appreciate cortical defect at fracture site
  - Thickened prevertebral soft tissues
- T2WI
  - Variable and inconsistent hyperintense signal in C2 marrow due to osseous edema
    - Fractures without compression &/or fractures with distraction do not reliably generate marrow edema and can lead to false-negative MR imaging
  - Hyperintense soft tissue edema
  - Effacement of thecal sac due to displaced fracture
  - Cord injury, if present, hyperintense on T2WI
  - Epidural hematoma transitioning from hyperintense to hypointense during hyperacute → acute period

Nuclear Medicine Findings

- Bone scan
  - Healing fracture tracer-avid for some time after injury
  - May also be used to assess for pseudoarthrosis following surgical repair
  - Patients > 75 years old or debilitated may not show activity for days or up to 2 weeks after injury

Imaging Recommendations

- Best imaging tool
  - Unenhanced multidetector CT scan
- Protocol advice
  - Plain radiographs (especially lateral, open mouth views) initially suggest need for CT
  - Thin slice (≤ 1 mm) axial CT in bone reconstruction algorithm
    - Fastest possible scan times to minimize motion
    - Sagittal and coronal reformatted images mandatory
  - Coronal and sagittal T1WI, sagittal T2WI, sagittal STIR, axial T1WI, and axial T2 or T2* to evaluate fracture morphology, displacement, ligamentous injury, soft tissue edema

DIFFERENTIAL DIAGNOSIS

Os Odontoideum

- Odontoid replaced by ossicle with no continuity to C2 body
- Corticated margins on radiography, CT
- No soft tissue swelling
- No recent history of trauma, no pain

Pathologic C2 Fracture

- Can produce pathologic odontoid fracture
- Metastases, infection, other inflammatory arthritidis

Rheumatoid Arthritis: C1/C2 Subluxation

- Synovial proliferation erodes dens
- Laxity, subluxation

Ossiculum Terminale Persistens

- Nonfusion of ossiculum terminal (apical odontoid epiphysis) to body of dens beyond 12 years of age

Congenital Variation: 3rd Occipital Condyle (Condylus Tertius)

- One of several anomalous bony structures that may form at foramen magnum as remnant of 1st sclerotome
- Midline bony peg off anterior lip of foramen magnum may articulate to dens, simulate odontoid type I fracture

PATHOLOGY

General Features

- Etiology
  - Sudden forward or backward movement of head, neck rigidly erect, and articulations locked
  - Osteoporosis in elderly predisposes to type II fracture and nonunion
- Associated abnormalities

- Type I odontoid fracture rarely occurs as isolated abnormality, usually as part of more extensive pattern of craniocervical injury
- Embryology: Anatomy
  - 5 primary ossification centers and 1 secondary ossification center form C2
  - Os odontoideum represents unfused dentrocentral synchondrosis vs. unrecognized fracture through synchondrosis prior to fusion at 5-7 years of age
  - Type II fracture follows embryologic line of union between dens and body of C2
  - Ossiculum terminale represents secondary center at tip of odontoid, fuses by 12 years of age

Staging, Grading, & Classification

- Anderson and D'Alonzo (1974)
  - Type I fracture: Oblique fracture through upper portion of odontoid process
  - Type II fracture: Fracture across base of odontoid process near junction with axis body
  - Type III fracture: Fracture that includes odontoid and extends into axis body
- Hadley modification (1988)
  - Type IIA: Comminuted fracture of base of odontoid with associated free fracture fragments
    - Highly unstable
- Grauer modification (2005)
  - Type IIA: Minimally or nondisplaced fracture with no comminution; treated with external immobilization
  - Type IIB: Displaced odontoid fracture that extends from anterior-superior to posterior-inferior, or transverse; treated with anterior screw fixation if reducible
  - Type IIC: Fracture extending from anterior-inferior to posterior-superior or with significant comminution; considered for posterior internal fixation and fusion

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck pain
- Other signs/symptoms
  - Myelopathy
- Clinical profile
  - Frequently elderly, osteoporotic patients
  - Fluctuating long tract signs and spasticity may be only presentation in older patients with minor initial trauma; may be missed clinically

Demographics

- Epidemiology
  - 7-17% of all cervical spine fractures

Natural History & Prognosis

- Chronic nonunion or fibrous union in elderly
- Nonunion common in elderly without primary fusion
  - May stabilize by fibrous union with prolonged immobilization
- Fusion produces stability

Treatment

- Fracture pattern dictates management
- Type I fracture
  - Usually stable injury
    - Cases of gross instability with severe distraction mechanism
  - Treated with simple immobilization
- Type II fracture
  - Most likely to progress to nonunion
  - Surgical fusion may be necessary to achieve stability
- Type III fracture
  - Nonunion uncommon after treatment with traction followed by bracing

DIAGNOSTIC CHECKLIST

Consider

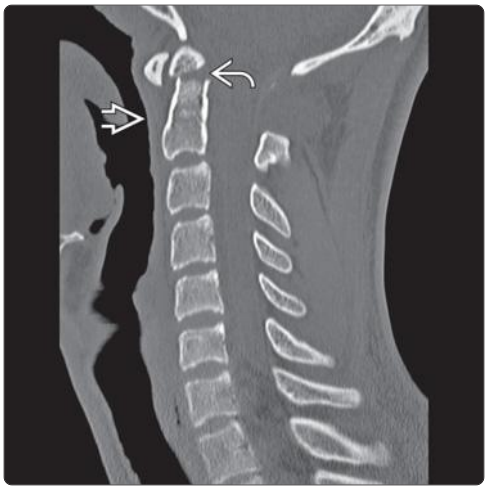
- STIR to show soft tissue edema in prevertebral space (missing in chronic nonunion)
  - Marrow edema an unreliable sign for presence of fracture in distraction injuries
- T1W sagittal and coronal images to depict disruption of bone contiguity and degree of displacement
  - Marrow shows signal loss from edema in acute cases
  - Normal signal indicates chronic nonunion
- Flexion/extension films or fluoroscopy for evaluating stability

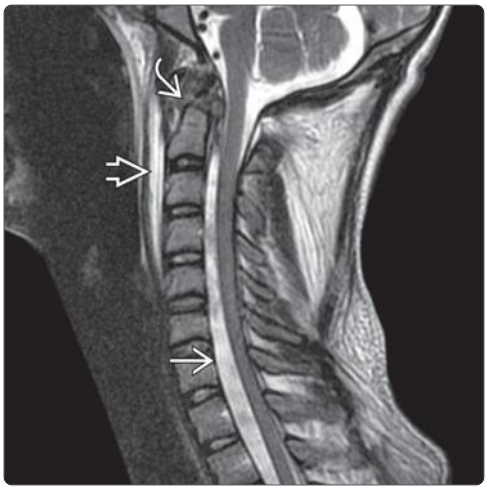
Image Interpretation Pearls

- Sclerotic margins of ununited dens indicate chronic nonunion of remote fracture
- High resolution T2W axial imaging for evaluation of transverse ligament integrity

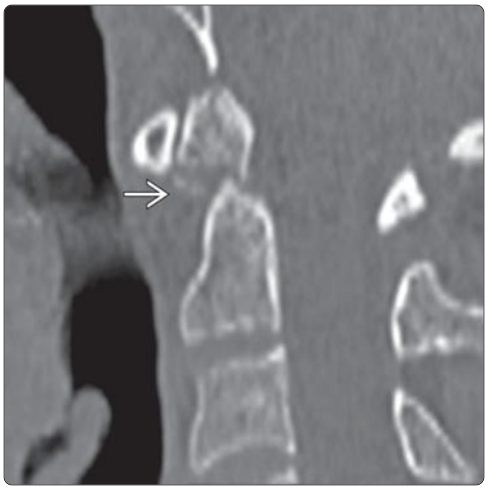
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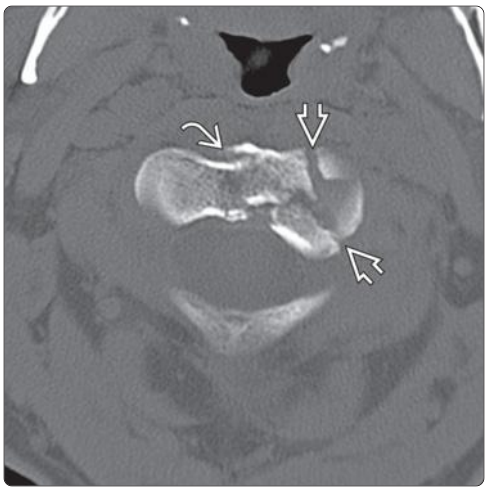


(Left) *Sagittal reformatted CT shows a type I odontoid fracture ➡ horizontally through the upper portion of the odontoid. The prevertebral soft tissues ➡ are thickened.* **(Right)** *Sagittal T2WI MR in the same patient shows an osseous defect ➡ of the upper dens but no marrow edema. Prevertebral soft tissues are thickened and edematous ➡. Extensive ventral epidural hematoma and pseudomeningocele ➡ are hyperintense on T2WI, with minimal heterogeneity.*





(Left) *Sagittal reconstructed CT shows a fracture ➡ extending through the base of the dens (type II). The dens is moderately displaced anteriorly.* **(Right)** *Coronal reformatted CT in the same patient shows the irregular fracture ➡ through the base of the dens. The dens is displaced slightly to the right.*





(Left) *Axial NECT shows a severely comminuted fracture at the base of the odontoid ➡ extending into the body and left lateral mass ➡ of C2 (type III odontoid fracture).* **(Right)** *Coronal reformatted CT in the same patient shows a type III fracture extending from the right of the base of the odontoid ➡ through the C2 body and into the left lateral mass ➡.*

Burst C2 Fracture

KEY FACTS

TERMINOLOGY

- Comminuted fracture through C2 vertebral body
- Displacement of fragments in AP direction

IMAGING

- AP displacement of fracture fragments
- "Fat" C2 sign
- Often see associated hangman's fracture, unstable injury
- Displaced fracture fragments may encroach on spinal canal
- CTA or MRA for screening vertebral artery injury
  - Especially if fracture extends to foramen transversarium

TOP DIFFERENTIAL DIAGNOSES

- Flexion teardrop fracture of C2
- Other nonodontoid, non-hangman's C2 body fracture
- Pathologic fracture

PATHOLOGY

- High-energy mechanisms cause most C2 fractures
- Fujimara (1996) classification of C2 body fractures (non-hangman's, nonodontoid C2 fractures)
  - Avulsion fracture
  - Transverse fracture
  - Burst fracture
  - Sagittal fracture

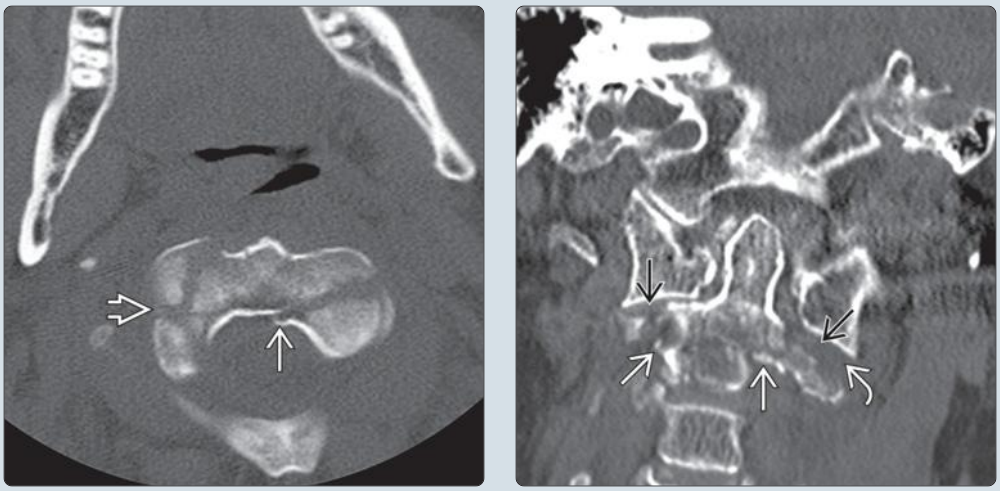
CLINICAL ISSUES

- High-energy, high-velocity trauma
- May have concomitant head injury
- Often managed with conservative/nonoperative treatment
- Surgical management for inability to reduce fragments, associated hangman's fracture, malalignment of atlantoaxial articulation

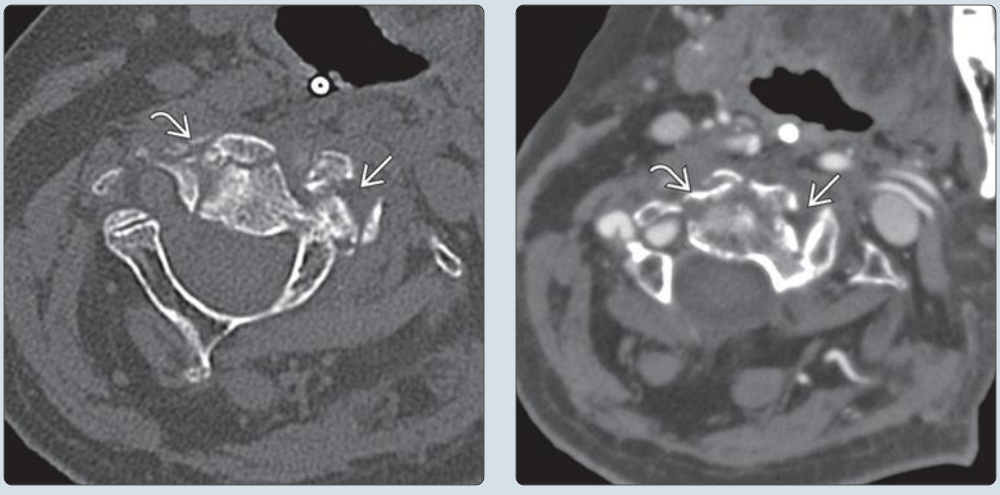
DIAGNOSTIC CHECKLIST

- C2 burst fractures can involve the body, pedicle, lateral mass, and transverse foramina

(Left) *Axial NECT shows a comminuted C2 body fracture with fragmentation of the right lateral mass. There is slight posterior displacement of fragments, flattening the thecal sac. C2 burst injuries may occur as a result of isolated or combined hyperflexion or hyperextension forces.* **(Right)** *Coronal CT illustrates a highly comminuted C2 burst fracture. The fracture extends into the superior articular facets with widening of the C1-C2 facet joint on the left.*



(Left) *Axial bone CT demonstrates a comminuted burst fracture of C2, involving the left transverse foramen. Primary hyperflexion mechanism predominantly disrupts the anterior margin, while primary hyperextension is suggested if the posterior margin is disrupted. Combined hyperextension-hyperflexion injuries may result in the interruption of both margins.* **(Right)** *Axial CTA exhibits occlusion of the left vertebral artery due to C2 burst fracture involving the left transverse foramen.*



TERMINOLOGY

Synonyms

- Nonodontoid, non-hangman's, miscellaneous C2 fracture

Definitions

- Comminuted fracture through C2 vertebral body, pedicle, superior articulating process, transverse foramen
- Displacement of fragments in AP direction

IMAGING

General Features

- Best diagnostic clue
  - Comminuted fracture of C2 body
  - Displaced fragment, widening of C2 body on lateral view
- Location
  - C2 body
- Morphology
  - Comminuted fracture disrupting C2 body

Radiographic Findings

- Radiography
  - Lateral radiograph
    - May not appreciate fracture line(s) directly
    - AP displacement of fracture fragments
    - "Fat" C2 sign
    - Prevertebral soft tissue swelling
    - Often see associated hangman's fracture
  - Frontal radiograph
    - Comminuted fracture lines within C2 body
    - Degree of displacement suggests magnitude of trauma
    - May extend into lateral masses

CT Findings

- NECT
  - Comminuted fracture through C2 vertebral body
  - Involves posterior cortex of body
  - Displaced fracture fragments
    - May encroach on spinal canal
  - Upper prevertebral soft tissue swelling
  - May demonstrate hyperdensity in epidural space if hemorrhage is present
- CTA
  - Screening for vertebral artery dissection or occlusion
    - Especially if fracture extends to foramen transversarium

MR Findings

- T1WI
  - Low signal in C2 body due to edema/hemorrhage
  - Disruption of cortical margins of C2
  - Prevertebral soft tissue edema
- T2WI
  - High signal edema in thickened prevertebral soft tissue
  - Effacement of thecal sac by displaced fracture fragments
  - May see hyperintense intramedullary signal if contusion is present
    - Intramedullary low signal within area of edema concerning for hematomyelia
- STIR
  - High signal in C2 vertebral body, adjacent soft tissues due to edema and hemorrhage
- T2* GRE
  - Accentuates susceptibility associated with blood products in canal or cord (parenchymal hematoma)
- MRA
  - Irregular contour of vertebral artery (dissection) or occlusion due to associated vascular injury
    - Axial T1WI MR with fat saturation increases conspicuity of mural hemorrhage

Angiographic Findings

- Useful if CTA/MRA equivocal or for persistent concern for vertebral artery injury

Nuclear Medicine Findings

- Bone scan
  - Patients > 75 years old or debilitated may not show activity for several days to as much as 2 weeks

Imaging Recommendations

- Best imaging tool
  - Unenhanced multidetector CT scan
- Protocol advice
  - Plain radiographs initially suggest need for CT
  - Thin-slice (≤ 1 mm) axial CT in bone reconstruction algorithm
    - Fastest possible scan times to minimize motion
    - Sagittal and coronal reformatted images mandatory
  - Coronal and sagittal T1WI, sagittal T2WI and STIR, axial T1 and T2WI to evaluate fracture morphology, displacement, ligamentous injury, soft tissue edema
    - Volumetric T2 and T1WI MR allow multiplanar reformations
  - T2* GRE imaging to detect hemorrhage if cord injury is present

DIFFERENTIAL DIAGNOSIS

Type III Odontoid Fracture

- Fracture through base of dens extending into C2 body (Anderson and D'Alonzo classification)
- Fracture often extends into C2 lateral masses, superior articular facets
- No or minimal retropulsion of fracture elements
- No vertical fracture line through C2 body

Flexion Teardrop Fracture of C2

- Anteroinferior body "teardrop" fragment, flexed alignment
- Distraction of posterior column
  - Facet subluxation/distraction
  - Spinous process splaying on lateral film
- Clinically important to distinguish from burst fracture
  - Most severe and unstable C-spine type of fracture
  - Difference in stability, management
  - May be difficult distinction on conventional radiographs

Other Nonodontoid, Non-Hangman's C2 Body Fracture

- Hyperextension "teardrop," transverse, and vertical C2 body fractures

Pathologic Fracture

- Destruction of cancellous and cortical bone
- Soft tissue mass
- Bowing of posterior cortex into epidural space

PATHOLOGY

General Features

- Etiology
  - High-energy mechanisms (MVA, falls) cause most C2 fractures
  - Mechanism of burst
    - Compression forces cause axial loading
    - Sudden "explosive" pressure ↑ causes vertebral body comminution
    - Fragments undergo centrifugal dispersion
    - Posteriorly retropulsed fragments enter canal and may impact, compress cord
  - Associated hangman's fracture implies unstable fracture
- Associated abnormalities
  - Hangman's fracture frequently associated with C2 burst fracture
  - Cord contusion ± hematoma
    - Infrequent, due to capacious canal diameter at C2
  - Vertebral artery injury, delayed neurologic sequelae
  - Closed head injury

Staging, Grading, & Classification

- Fujimara (1996) classification of C2 body fractures
  - Avulsion fracture: Triangular fragment at anteroinferior margin of C2 (hyperextension teardrop fracture)
  - Transverse fracture: Cross-sectional fracture of C2 body
    - Away from base of odontoid (which would be type III dens fracture)
  - Burst fracture: Comminuted fracture of axis body, displaced in AP dimension
  - Sagittal fracture: Vertical fracture, from adjacent to base of dens through inferior C2 endplate
- Benzel (1994) classification of C2 body fractures
  - Type I: Vertical, coronally oriented fracture plane
    - Incorrectly described as variant of hangman's fracture in some literature
  - Type II: Vertical, sagittal oriented
  - Type III: Transverse, axially oriented
    - Includes type III dens fracture

Gross Pathologic & Surgical Features

- Comminuted fracture
- Extruded blood, cellular debris, fragments of tissue envelope fracture site in hematoma
- Inflammation begins within hours and continues for several days until replaced by developing soft callus

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Symptoms are variable, from no deficit to quadriplegia
    - May have transient symptoms
  - With cord injury
    - Spinal shock: Flaccidity, areflexia, loss of anal sphincter tone, fecal incontinence, priapism, loss of bulbocavernosus reflex
    - Neurogenic shock: Hypotension, paradoxical bradycardia, flushed/dry/warm peripheral skin
    - Autonomic dysfunction: Ileus, urinary retention, poikilothermia
    - Tetraparesis
- Clinical profile
  - High-energy, high-velocity trauma
    - High-risk activities: Diving, equestrian activities, football, gymnastics, skiing, hang gliding
  - Axial loading injury, may have concomitant head injury

Demographics

- Age
  - C-spine injury: 80% of patients 18-25 years old
- Epidemiology
  - 25% of C2 fractures cannot be classified as odontoid or hangman's fractures
    - C2 burst is uncommon fracture pattern

Natural History & Prognosis

- Considered unstable injury given high frequency of association with hangman's fractures

Treatment

- C2 body fractures often managed with conservative/nonoperative treatment
  - Traction
  - Immobilization
- Inability to reduce fracture fragments, hangman's fracture associated with burst fracture, malalignment of atlantoaxial articulation
  - Operative management may be considered
  - No standard surgical therapy
- Cord injury
  - Steroids in acute cord injury controversial
  - Standard of management includes support of arterial oxygenation and spinal cord perfusion pressure

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- These fractures can involve the body, pedicle, lateral mass, and transverse foramina

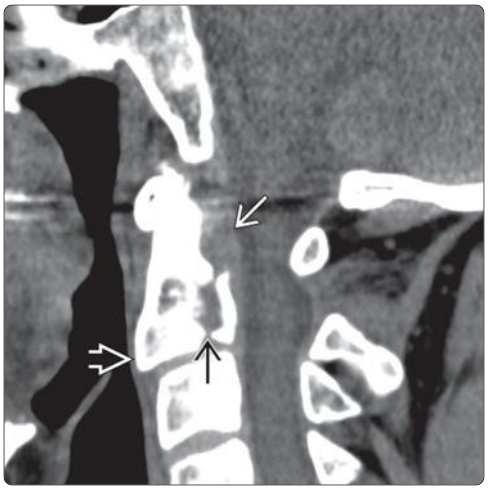
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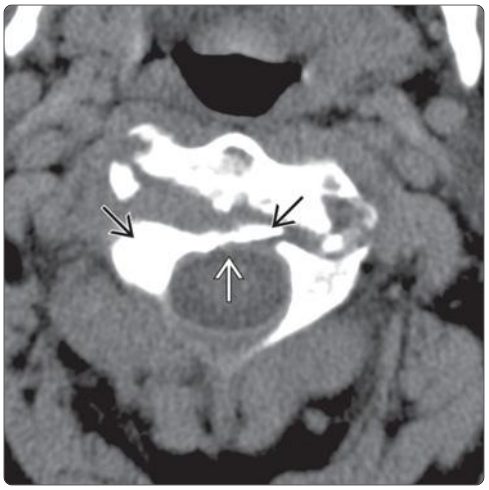
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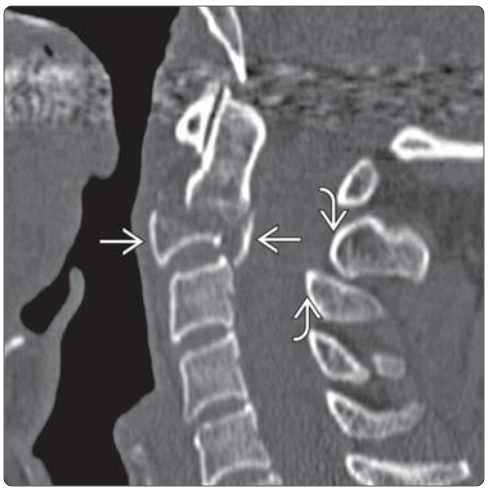


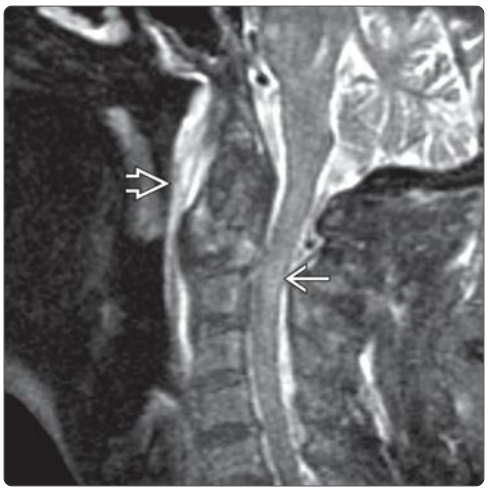
(Left) *Lateral radiograph reveals a vertically oriented C2 body fracture. The "fat" C2 sign refers to the increase in the AP distance between the margins of the C2 vertebra when compared to those of the C3 vertebra. There is also a displaced fracture of the C4 spinous process. Fracture/dislocation is observed at the C5-C6 level.* **(Right)** *Coronal CT demonstrates a transverse, horizontally oriented fracture of the C2 body, extending into the C2-C3 disc space.*





(Left) *Sagittal CT reconstruction shows a vertical, coronally oriented fracture of the C2 body. A small epidural hematoma is noted in the ventral spinal canal. C2 is slightly anteriorly subluxed with respect to C3.* **(Right)** *Axial NECT exhibits a coronally oriented fracture of the C2 body with retropulsion of the osseous fragment; this effaces the ventral CSF collar. The use of external immobilization is advocated as the initial treatment with reported 1.6% nonunion rate.*





(Left) *Sagittal bone CT reformat reveals centrifugal displacement of C2 burst fracture fragments. The dorsal fragment encroaches on the ventral spinal canal. Posteriorly, there is disruption of the spinolaminar line at C2-C3.* **(Right)** *Sagittal STIR MR of a comminuted C2 fracture shows posterior displacement of osseous fragments, which efface the thecal sac. Contusion of the spinal cord manifests with faint T2 hyperintensity. There is prevertebral soft tissue edema.*

Hangman's C2 Fracture

KEY FACTS

TERMINOLOGY

- Traumatic spondylolisthesis of axis (TSA)
- Bilateral C2 pars interarticularis fractures

IMAGING

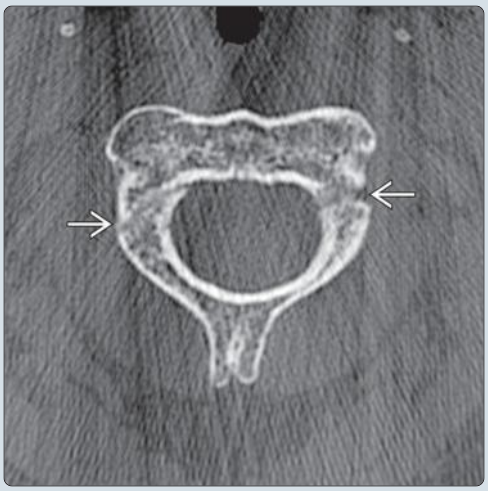
- Fracture through pars interarticularis of C2
  - Fracture may extend into C2 vertebral body
- Anterior displacement of C2 body relative to C3
- C1 arch and skull ride forward with C2 vertebral body
- Posterior elements and spinolaminar line of C2 and C3 remain aligned
- Flexion exaggerates C2-C3 subluxation
- Involvement of vertebral artery foramen would be concerning for vertebral artery injury
  - Consider CTA
- Additional fracture levels seen in 33% of cases, C1 most commonly
- Thin-section (1 mm) helical CT with sagittal and coronal reformations
  - Any anterior subluxation of C2 vs. C3 on lateral x-ray warrants CT
- High T2 signal (edema) in thickened prevertebral soft tissue
  - Possible spinal cord injury: Evaluate signal in cord on T2, STIR

PATHOLOGY

- TSA results from hyperextension with axial loading or forced hyperflexion with axial loading
- True hangman's fracture, seen in judicial hanging, is caused by hyperextension combined with sudden, violent distraction

CLINICAL ISSUES

- Type I: Stable lesion, no permanent deficits
- Types II, III: Higher rates of sequelae, disability





(Left) *Axial NECT shows nondisplaced fractures of bilateral C2 pars interarticularis in this adolescent with hyperextension injury.* **(Right)** *Sagittal CT reconstruction from the same patient shows nondisplaced fracture extending obliquely through the left C2 pars interarticularis.*





(Left) *Lateral radiograph shows fractures of bilateral C2 pars interarticularis with distraction and anterolisthesis of C2 on C3. C2-C3 spinolaminar line is preserved, with interruption at C2.* **(Right)** *Sagittal STIR MR shows disruption of the posterior longitudinal and interspinous ligaments with C2-C3 anterolisthesis. Prevertebral soft tissue edema is noted, and there is a ventral epidural hematoma causing canal stenosis and cord compression.*

TERMINOLOGY

Synonyms

- Traumatic spondylolisthesis of axis (TSA)

Definitions

- Fractures through bilateral C2 pars interarticularis
- Avulsion of C2 body from facets, neural arch

IMAGING

General Features

- Best diagnostic clue
  - Classic imaging appearance
    - Fractures through C2 pars interarticularis
    - Anterior displacement of C2 vertebral body relative to C3
    - C1 arch and skull ride forward with C2 vertebral body
      - May not see malalignment despite pars interarticularis disruption
    - Posterior elements and spinolaminar line of C2 and C3 remain aligned
    - Flexion exaggerates C2-C3 subluxation
  - CT best defines morphology of fracture
  - Soft tissue swelling in upper prevertebral tissues common
- Location
  - Both pars interarticularis of C2

Radiographic Findings

- Radiography
  - C2 vertebral body anteriorly subluxed relative to C3
  - Radiolucent gap in pars interarticularis of C2
  - Prevertebral soft tissue swelling
  - C1 arch and skull ride forward with C2 vertebral body
  - Posterior elements and spinolaminar line of C2 and C3 remain aligned

Fluoroscopic Findings

- Flexion exaggerates C2-C3 subluxation
- Useful for verifying stability of therapeutic fusion

CT Findings

- NECT
  - Bone window CT
    - Bilateral fractures of C2 pars interarticularis
    - Variation of fracture patterns common
      - Extension of fracture into C2 body
      - Unilateral pars with fracture or contralateral pedicle or lamina
    - Dens typically spared
    - Additional fracture levels seen in 33% of cases, C1 most commonly
    - Canal at C2 enlarged in AP dimension
  - Soft tissue windows
    - Prevertebral soft tissue swelling
    - Epidural hemorrhage
- CTA
  - May see evidence of concomitant vertebral artery injury (dissection, occlusion)

MR Findings

- T1WI
  - Low signal in neural arch of C2 due to edema
- T1WI FS
  - Hyperintense perivascular signal if associated vascular injury present
- T2WI
  - High-signal edema in thickened prevertebral soft tissue
  - Swelling and hyperintensity (edema) in cord indicates spinal cord injury
    - May see focal or irregular intramedullary hypointensity if hematoma is present
- STIR
  - High signal in neural arch of C2, adjacent soft tissues due to edema and hemorrhage
- T2* GRE
  - Exaggerates susceptibility (hypointensity) associated with blood products in canal or cord (hematoma)
- MRA
  - Irregular contour of vertebral artery (dissection) or occlusion due to associated vascular injury

Imaging Recommendations

- Protocol advice
  - Any anterior subluxation of C2 vs. C3 on lateral x-ray warrants CT
  - Evaluate entire cervical spine (even upper thoracic): Associated fractures occur in 33% of cases
  - Thin-section (1 mm) helical CT with sagittal and coronal reformations
    - Mandatory to assess fractures, degree of subluxation, canal status
  - MR of cervical spine if neurologic symptoms are present
  - MRA or CTA if fracture line involves transverse foramen
  - Catheter angiography for endovascular intervention

DIFFERENTIAL DIAGNOSIS

Pseudosubluxation

- Physiologic anterolisthesis in upper cervical spine, occurs in young children (< 8 years old)
- Reflects laxity of juvenile cervical ligaments
- C2-C3, and less commonly C3-C4
- Occurs on lateral x-ray view when mildly flexed
- No associated soft tissue swelling
- Spinolaminar alignment preserved

Rotatory Subluxation of C2-C3

- Unilateral fracture(s) of C2 arch, pedicle

Spondylolysis, Primary Cervical

- Rare congenital anomaly
- Persistence of unossified embryonic synchondrosis

PATHOLOGY

General Features

- Etiology
  - TSA results from hyperextension with axial loading or from forced hyperflexion with axial loading

  - True hangman's fracture, seen in judicial hanging, is caused by hyperextension combined with sudden, violent distraction
    - Classical hanging with knot in submental position produces complete disruption of C2, C3 disc and ligaments
    - Not seen in victims of suicidal hanging
  - Traumatic TSA has different mechanism (e.g., chin vs. dashboard), similar results in bony spine
- Associated abnormalities
  - Fractures at other levels, though not always contiguous
    - Associated C1 fracture is most common of other levels involved

Staging, Grading, & Classification

- Effendi
  - Type I: Hairline fracture, nondisplaced; no disruption of C2-C3 disc
  - Type II: Fracture with ≥ 3 mm anterior translation of C2 on C3, abnormal C2-C3 disc
  - Type IIA: Minimum degree of C2-C3 displacement but severe angulation associated with flexion distraction
    - Levine and Edwards modification to Effendi classification
  - Type III (rare): Displaced anterior fracture fragment with dislocated or subluxed unilateral or bilateral facet joints
- Effendi classification can be applied to other fractures of C2 pedicles and lamina
- Atypical hangman's fracture: Fracture extends into posterior cortex of vertebral body
  - Bony spike ventral to cord, associated with higher incidence of paralysis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute neck pain
- Other signs/symptoms
  - Neurological deficits
  - Cerebellar findings suggesting stroke due to vertebral artery dissection or occlusion
- Clinical profile
  - Upper neck pain after trauma
  - Neurological sequelae occur in minority of traumatic cases (25%)
    - Canal wide here, further decompressed by fracture
  - Vertebral artery injury may cause delayed neurological signs
    - Posterior circulation infarcts

Demographics

- Epidemiology
  - Almost all modern cases represent sequelae of accidents rather than hanging
    - Only a minority of judicial hanging victims show C2 arch fracture
  - TSA represents 4-7% of all cervical fractures &/or dislocations
  - Isolated TSA represented 7% of all craniovertebral fractures in 1 series

Natural History & Prognosis

- Depends on presence of neurological damage
- Type I: Stable lesion, should never result in permanent neurological deficit
- Type II: ~ 20% transient neurologic deficits, 5% permanent neurologic deficits
- Type III: Higher risk of permanent, disabling sequelae
- Delayed stroke is possibility if vertebral artery damaged
- Accelerated degenerative changes

Treatment

- Conservative treatment
  - Immobilization
- Fusion for unstable injuries
  - Severe angulation of C2 on C3, disruption of the C2-C3 disc, &/or inability to maintain fracture alignment with external immobilization

DIAGNOSTIC CHECKLIST

Consider

- Check vertebral body alignment, soft tissue thickness (4 mm or less) on lateral x-ray; get CT if abnormal
- Evaluate transverse foramen for integrity; get MRA/CTA to exclude vertebral artery injury

Image Interpretation Pearls

- C2-C3 vertebral body anterior subluxation with spinolaminar line normally aligned requires CT even without fracture shown on x-ray

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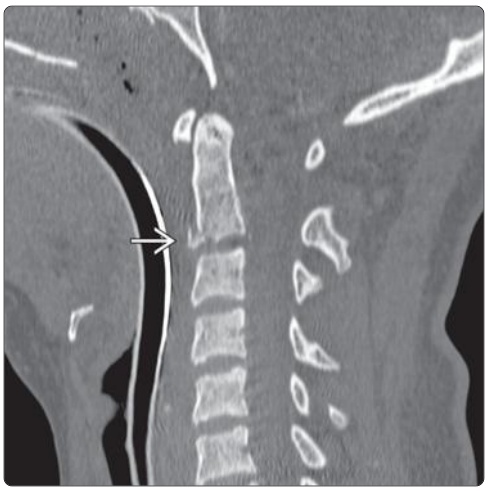
(Left) *Lateral cervical radiograph shows linear lucency in pars interarticularis of C2. There is subtle anterolisthesis and more conspicuous malalignment of the spinolaminar line at C1-C2.* **(Right)** *Axial NECT shows atypical-type traumatic spondylolisthesis of axis (TSA), in which left pars interarticularis fracture extends into inferior margin of posterior C2 vertebral body. If displaced, this sort of atypical fracture could potentially "scissor" the contents of the spinal canal.*





(Left) *Sagittal STIR MR shows cortical disruption at the site of a C2 pars interarticularis fracture. Minimal soft tissue edema is present. No marrow edema is present in this acute injury.* **(Right)** *Sagittal STIR MR in another patient with C2 pars interarticularis fractures shows only mild adjacent soft tissue edema. The fracture itself is not visible.*





(Left) *Sagittal CT reconstruction shows minimally distracted C2 pars interarticularis fracture.* **(Right)** *Sagittal reconstruction through the midline in the same patient shows a tiny flexion teardrop fracture at the anterior base of C2.*

Apophyseal Ring Fracture

KEY FACTS

TERMINOLOGY

- Fracture or avulsion of vertebral ring apophysis (RA) following injury in immature skeleton

IMAGING

- Concentric bone fragment displaced from vertebral endplate margin in skeletally immature patient
- Inferior or superior endplate may be involved; limbus vertebra (LV) usually superior
- Fractured apophyseal fragment usually midline

TOP DIFFERENTIAL DIAGNOSES

- Flexion teardrop fracture of anterior endplate corner
- Schmorl node
- Disc calcification or ossification
- Calcified disc fragment; posterior osteophyte
- Disc herniation

PATHOLOGY

- LV: Herniation of nucleus pulposus (NP) between ring apophysis and vertebral body
- PAR-Fx: 2 possible mechanisms described
  - Same as LV
  - Herniating NP spares Sharpey fibers, avulses ring apophysis

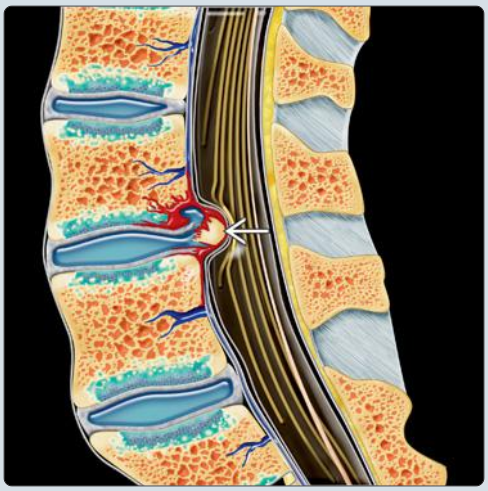
CLINICAL ISSUES

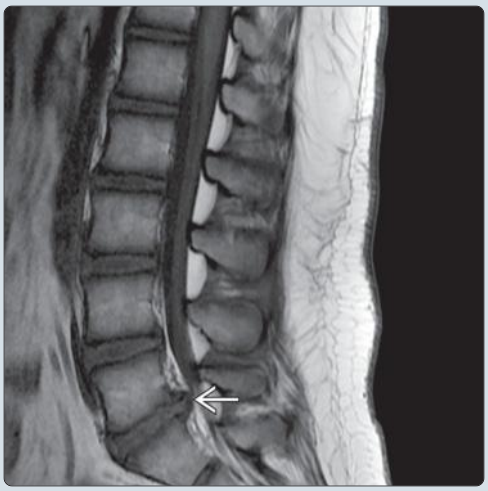
- Back pain in acute (adolescent) cases
- Adolescent athlete with acute low back pain
- Majority of patients with PAR-Fx/LV report engagement in sporting activities

DIAGNOSTIC CHECKLIST

- MR more sensitive than radiographs or CT in young children (RA not ossified)
- T2WI FS/STIR MR essential to assess for associated ligamentous injury

(Left) *Sagittal graphic demonstrates an acute lumbar apophyseal ring fracture ➡ involving the posterior inferior vertebral body corner with displacement and associated hemorrhage. There is compression of the adjacent thecal sac.* **(Right)** *Sagittal T1WI MR (posterior ring apophysis fracture with disc herniation) shows herniation of the L5-S1 disc and fracture with displacement of the L5 ring apophysis ➡. Note marrow edema and endplate irregularity.*





(Left) *Sagittal STIR MR (same patient) nicely confirms marrow edema ➡ and delineates the fracture line. Note also the superimposed congenital spinal stenosis with reduced AP canal diameter related to congenital short pedicles, which exacerbates clinical symptoms.* **(Right)** *Axial T2WI MR (same patient) shows posterior displacement of the fractured apophysis ➡, narrowing the central spinal canal and both lateral recesses.*





TERMINOLOGY

Synonyms

- Vertebral apophyseal fracture (Fx), limbus vertebra (LV), endplate avulsion Fx, corner Fx, slipped vertebral apophysis, lumbar posterior marginal node

Definitions

- Fx or avulsion of vertebral ring apophysis (RA) following injury in immature skeleton
  - Fx of anterior ring termed limbus vertebra
  - Fx of posterior ring termed posterior apophyseal ring Fx (PAR-Fx)

IMAGING

General Features

- Best diagnostic clue
  - Concentric bone fragment displaced from vertebral endplate margin in skeletally immature patient
- Location
  - Lumbosacral spine most common; rare in thoracic, cervical
    - L3-S1
    - PAR-Fx commonly L4, S1
  - Inferior or superior endplate may be involved; LV usually superior
  - Fractured apophyseal fragment usually midline
- Size
  - Fragment size variable
- Morphology
  - Rim-like morphology typical, but fragment(s) may be amorphous

Radiographic Findings

- Radiography
  - Bone fragment displaced from corner defect of endplate
  - Bone fragment not identified > 50% (cartilaginous or resorbed)

CT Findings

- Bone CT
  - Acute
    - PAR-Fx: Arc-shaped or rectangular bone fragment posterior to dorsal endplate margin
    - Vertebral donor site usually identified
    - LV: Same as PAR-Fx except located anteriorly, ± mild kyphosis
  - Subacute or chronic
    - Sclerosis of fragment and donor site margins
    - Donor defect often enlarges (particularly LV)

MR Findings

- T1WI
  - Corner defect in marrow of parent vertebral body (VB) with disc extending into defect
  - Hypointense bone fragment merges with adjacent ligaments
  - PAR-Fx fragment plus Sharpey fibers = "Y or 7" shape on sagittal MR
- T2WI
  - Defect in endplate; hyperintense disc between fragment and VB
  - Disc height loss common ± disc desiccation
    - Both ↑ over time
  - High signal in subjacent marrow in acute cases
- STIR
  - Same as T2WI, edema accentuated
- T2* GRE
  - Accentuation of sclerotic bone margins in chronic lesions
- T1WI C+
  - Enhancement of donor site marrow if acute fracture

Nonvascular Interventions

- Myelography
  - Epidural defect on contrast column in PAR-Fx

Nuclear Medicine Findings

- Bone scan
  - Increased radionuclide uptake in acute lesions

Other Modality Findings

- Discography (LV): Contrast diffuses into space between fragment and vertebral body

Imaging Recommendations

- Best imaging tool
  - Multiplanar CT in bone and soft tissue algorithm
- Protocol advice
  - Sagittal and axial reformatted images key for making diagnosis

DIFFERENTIAL DIAGNOSIS

Flexion Fracture of Anterior Endplate Corner

- Seen in older children, adults after apophysis fused
- Spinal cord injury more common than in apophyseal ring fracture

Schmorl Node

- Endplate defect is within interspace, not at endplate corner
- Associated edema and contrast enhancement when acute

Disc Calcification or Ossification

- Painful childhood nucleus pulposus (NP) calcification of unknown etiology
  - Multilevel common, cervical > thoracic, occasionally asymptomatic
  - Spontaneous resolution of calcifications and symptoms

Calcified Disc Fragment; Posterior Osteophyte

- Sequelae of longstanding degenerative disc disease, uncommon in children
- Fragment at level of disc, not above or below
- May see marrow if ossified

Disc Herniation

- Uncommon in adolescents, rare (traumatic) in 1st decade

PATHOLOGY

General Features

- Etiology
  - LV: Herniation of nucleus pulposus between ring apophysis and vertebral body

- PAR-Fx: 2 possible mechanisms described
  - Same as LV
  - Herniating NP spares Sharpey fibers, avulses ring apophysis
- Genetics
  - Often seen with Scheuermann disease
- Associated abnormalities
  - Disc herniation
  - Kyphosis
- Embryology-anatomy
  - Superior and inferior surface of developing vertebral body covered by thin, peripherally thickened cartilage plate → cartilaginous marginal ridge
  - Endochondral ossification of marginal ridge begins at 7-9 years → RA
  - RA seen as small bony triangles at corners of vertebral body (XR, sagittal/coronal CT reconstruction)
  - RA separated from vertebral body by thin cartilage layer until apophysis fuses to vertebral body at 18-20 years → relative weak point in disc/vertebral body complex until fusion occurs
  - Outermost fibers of annulus fibrosus (Sharpey fibers) embedded in RA, attaching disc to spine

Staging, Grading, & Classification

- Modified Takata classification
  - Type I: Avulsion posterior cortical vertebral rim (most common)
  - Type II: Central cortical and cancellous bone Fx
  - Type III: Lateralized chip Fx
  - Type IV: Spans entire posterior vertebral margin

Gross Pathologic & Surgical Features

- Displaced bony/cartilaginous rim fragment ± disc material
- Sharpey fibers and posterior longitudinal ligament usually intact

Microscopic Features

- Cancellous bone, hyaline cartilage, and acellular hyaline tissue (disc)
- Basophilic degeneration and foci hemorrhage in hyaline cartilage common

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain in acute (adolescent) cases
- Clinical profile
  - Adolescent athlete with acute low back pain
  - Majority of patients with PAR-Fx/LV report engagement in sporting activities
- PAR-Fx: Principal presentation
  - Acute > > prolonged history of central low back pain ± sciatica
    - 66% had minor trauma or lifting event (weightlifting, gymnastics)
  - Physical exam (PE) findings: Similar to herniated nucleus pulposus
- LV: No inciting event; chronic history of back pain; occasionally incidental finding
  - PE: ↓ ROM, ± kyphosis and pain with palpation of spinous processes

Demographics

- Age
  - Late childhood through adolescence
- Gender
  - M > F (up to 85% of males in PAR-Fx group)
- Epidemiology
  - 20% prevalence of PAR-Fx teen lumbar disc surgeries (33% in 14-17 years)

Natural History & Prognosis

- LV: Symptoms resolve in months (usual) to years (rare)
  - Reports document evolution of LV into anteriorly located Schmorl node
- PAR-Fx: Good to excellent with surgery; occasional mild, short-term deficits

Treatment

- LV: Conservative: Analgesics and limitation of physical activity in acute symptomatic setting
- PAR-Fx: Surgical therapy is mainstay; conservative approach rarely successful
  - Uni- or bilateral laminotomy with removal bone fragment, ± disc

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- MR more sensitive than radiographs or CT in young children (RA not ossified)
- T2WI FS/STIR MR essential to assess for associated ligamentous injury

SELECTED REFERENCES

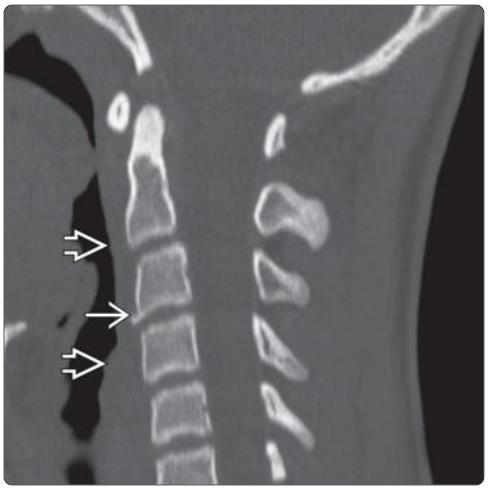
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(Left) *Sagittal T2WI MR (chronic lumbar ring apophysis fracture) shows displacement and remodeling of the anteroinferior L1 ring apophysis ➡ compared to normal ring apophyses at adjacent levels. Note interposition of disc material ➡ between the displaced apophysis and L1 vertebra.* **(Right)** *Axial bone CT (same patient) characterizes displaced ring apophysis ➡ more specifically as a ring apophysis fracture rather than an isolated anterior disc herniation.*





(Left) *Sagittal STIR MR (acute cervical ring apophysis fracture) shows marrow edema and avulsion of the anterior inferior corner of the C3 vertebra ➡. Note also prevertebral edema ➡ related to acute injury. The anterior longitudinal ligament (ALL) is intact.* **(Right)** *Sagittal bone CT (same patient) clearly demonstrates avulsion of the anterior inferior C3 ring apophysis ➡. The prevertebral soft tissues are widened ➡ with anterior displacement of the airway, confirming acute injury.*





(Left) *Sagittal bone CT (cervical posterior apophyseal ring fracture) demonstrates a small posteroinferior C6 vertebral fracture fragment ➡ with associated mild C6-7 anterolisthesis. The small osseous fragment is noted to represent the displaced posteroinferior C6 ring apophysis (empty donor site visible on comparison to other levels).* **(Right)** *Axial bone CT (same patient) confirms that the C6 osseous fragment ➡ represents the displaced C6 ring apophysis.*

Cervical Hyperflexion Injury

KEY FACTS

TERMINOLOGY

- Injury from cervical hyperflexion with compression or hyperflexion with distraction/shearing

IMAGING

- Hyperflexion with compression
  - Forced flexion of cervical spine with axial loading vector slightly anterior to vertebral column
  - Primary injury to anterior and (later) middle columns
- Hyperflexion with distraction
  - Forced flexion of cervical spine with rostrally oriented force vector nearly perpendicular to trunk
  - Primary failure of posterior and (later) middle columns
  - Focal kyphosis, ↑ space between spinous processes; distracted, perched, or jumped facets
- Mid or lower cervical spine more common
- Best imaging modality
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations
  - MR (especially STIR and GRE) to evaluate soft tissue structures, spinal cord

TOP DIFFERENTIAL DIAGNOSES

- Burst fracture
- Flexion-rotation injury
- Whiplash fracture

PATHOLOGY

- Spinal instability
- Spinal cord injury, radiculopathy
- Associated with closed head injury, polytrauma

CLINICAL ISSUES

- Immobilization, axial traction, surgical fusion
- Surgical decompression for spinal cord injury
- Methylprednisolone for treatment of acute spinal cord injury **not** recommended

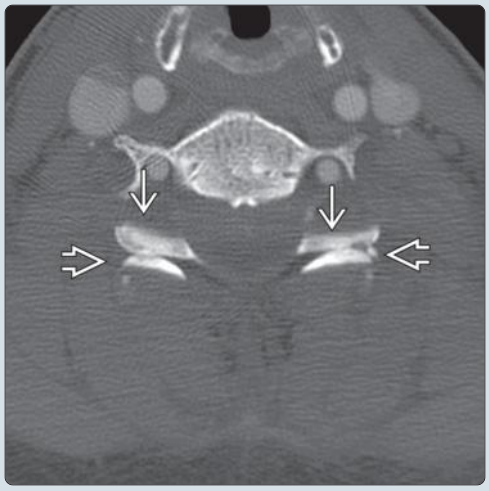
(Left) *Sagittal graphic shows flexion injury at C4-C5 with subluxation, disc disruption, disc herniation, ligamentous disruption, cord compression, and epidural hemorrhage.* **(Right)** *Sagittal T2WI MR shows severe flexion injury with subluxation of C5 on C6, flexion deformity and severe cord compression, and cord edema. Note the posterior ligamentous injury.*





(Left) *Sagittal CECT shows an inferior C6 facet anterior to the superior facet of C7 (known as "jumped facet") in this patient with flexion injury.* **(Right)** *Axial CECT in a flexion injury with "jumped facets" shows inferior facets of the C6 anterior to superior facets of C7 (known as "naked facets" sign or "hamburger" sign).*





TERMINOLOGY

Definitions

- Injury from cervical hyperflexion with compression or hyperflexion with distraction/shearing

IMAGING

General Features

- Best diagnostic clue
  - Focal kyphosis, anterior column compression deformity or "teardrop" fracture
  - Focal kyphosis, increased space between spinous processes; distracted, perched, or jumped facets
- Location
  - Mid or lower cervical spine more common
- Hyperflexion injury with axial compression
  - Hyperkyphosis of spine at level of injury
  - May show increased distance between adjacent spinous processes ("fanning")
  - Blunting of anterior margin of superior endplate extending to anterior vertebral wedging (mild injury)
  - Oblique coronal fracture through anteroinferior vertebral body with retrolisthesis on lower vertebral body (flexion "teardrop")
    - Severe, unstable injury
  - ± distraction of facet joints
- Hyperflexion injury with distraction or shear
  - Hyperkyphosis of spine at level of injury
  - Increased distance between adjacent spinous processes ("fanning")
  - ± blunting of anterior margin of superior vertebral endplate
  - Diastatic (> 2 mm), subluxed (articular apposition < 50%), perched, or jumped facet joints with anterolisthesis
    - 100% anterior displacement = "floating" vertebra

MR Findings

- T2WI
  - Need complete anterior to posterior assessment
    - Prevertebral soft tissues (edema or hemorrhage)
    - Anterior longitudinal ligament (ALL) may be intact
    - Possible disc distraction or abnormality increased signal
    - Posterior longitudinal ligament may be intact
    - Epidural space may be normal or hemorrhagic
    - Disc herniation may also be present
    - Possible cord signal abnormality (edema or hemorrhage)
    - Interspinous and supraspinous ligaments may be intact or abnormal with increased signal
- STIR
  - Prevertebral, ligamentous edema
  - Marrow edema due to vertebral compression or "teardrop" fracture
  - Increased signal in cord, if cord injury present
  - Epidural hemorrhage with thecal sac compression
- T2* GRE
  - Low signal intensity from cord hemorrhage

Imaging Recommendations

- Best imaging tool
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations
- Protocol advice
  - CT best to evaluate for fractures and facet widening/subluxation
  - MR (especially STIR and GRE) to evaluate ligaments, spinal cord

DIFFERENTIAL DIAGNOSIS

Burst Fracture

- Comminuted, centrifugally displaced vertebral body fracture fragments

Flexion-Rotation Injury

- Unilateral facet subluxation or facet fracture typical

Whiplash Fracture

- Predominantly ligamentous injury

PATHOLOGY

General Features

- Etiology
  - Hyperflexion with compression
    - Forced flexion of cervical spine with axial loading vector slightly anterior to vertebral column
    - Primary injury to anterior and (later) middle columns
      - Compression deformity, "teardrop" fracture
      - Retrolisthesis at level of injury, narrowing canal and compressing spinal cord
  - Hyperflexion with distraction or shearing
    - Forced flexion of cervical spine with rostrally oriented force vector nearly perpendicular to trunk
    - Primary failure of posterior and (later) middle columns
      - Widening of interspinous distance with disruption of interspinous ligaments
      - Distraction of facet joints, leading to subluxation or perching
      - With more severe force, jumped facets with anterolisthesis
      - PLL avulsion
- Associated abnormalities
  - Spinal instability
  - Spinal cord injury, radiculopathy
  - Closed head injury, polytrauma

Staging, Grading, & Classification

- Many different classification schemes of cervical spine injuries
- Holdsworth (1970)
  - Unified cervical, thoracic, and lumbar scheme including role of ligaments
  - Flexion, flexion-rotation, extension, vertical compression, direct shear
    - Stable and unstable
  - Loss of height > 50% seen as sign of ligamentous disruption and is unstable
- Marar (1974)
  - Correlated neurologic injury to mechanism

- No guidelines for treatment
  - Group 1: Complete cord injury (bilateral facet and burst fractures)
  - Group 2: Central cord injury (extension injury)
  - Group 3: Anterior cord syndrome (vertical compression and "teardrop")
  - Group 4: Partial motor loss with intact sensory (unilateral facet dislocation)
  - Group 5: Brown-Séquard syndrome
- Allen (1982)
  - 6 major mechanistic categories based on 4 cardinal force vectors (flexion, extension, compression, distraction)
  - Risk of neurologic injury increases with increasing stage for each type of injury
  - Good predictor of neurologic outcome
    - Compressive flexion (CF)
    - Vertical compression (VC)
    - Distractive flexion
    - Compressive extension
    - Distractive extension
    - Lateral flexion
- Moore (2006)
  - Morphological classification based on 4 pillars of cervical spine (anterior column, posterior column, right lateral column, left lateral column)
  - Each column given a score accounting for bony and ligamentous injuries
    - Score 0-20, 20 worst score
- Vaccaro (2007)
  - Subaxial cervical injury classification system
  - 3 major characteristics (injury morphology, status of discoligamentous complex (DLC), neurologic status)
  - Higher score (5 or more) = more severe injury, needs surgical treatment
  - Morphology
    - No abnormality = 0
    - Compression = 1
    - Burst modifier of compression = +1 = 2
    - Distraction = 3
      - Facet perch, hyperextension
    - Rotation/translation = 4
      - Facet dislocation, unstable "teardrop," advanced flexion compression)
  - DLC
    - Intact = 0
    - Indeterminate = 1
      - Isolated interspinous widening, MR imaging signal change only
    - Disrupted = 2
      - Widening of disc space, facet perch, dislocation
  - Neurologic status
    - Intact = 0
    - Root injury = 1
    - Complete cord injury = 2
    - Incomplete cord injury = 3
    - Continuous cord compression in setting of neurodeficit = +1
- Stability
  - Instability defined as "loss of ability of spine under physiological loads to maintain relationships between vertebrae in such a way that there is neither damage nor subsequent irritation to the spinal cord or nerve roots and, in addition no development of deformity with excessive pain"
  - White (1976)
  - 5 or more = unstable
    - Anterior elements destroyed or unable to function = 2
    - Posterior elements destroyed or unable to function = 2
    - Relative sagittal plane translation > 3.5 mm = 2
    - Relative sagittal plane rotation > 11° = 2
    - Positive stretch test = 2
    - Cord damage = 2
    - Root damage = 1
    - Abnormal disc narrowing = 1
    - Dangerous load anticipated = 1

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma ± radiculopathy, myelopathy

Natural History & Prognosis

- Varies with degree of neurologic injury
- Accelerated degenerative disease may be seen

Treatment

- Immobilization, axial traction, surgical fusion
- Surgical decompression for spinal cord injury
- Methylprednisolone for treatment of acute spinal cord injury **not** recommended

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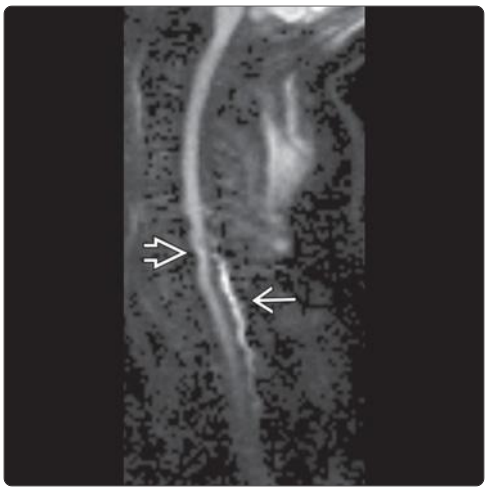
(Left) *Lateral radiograph shows anteriorly narrowed C6-C7 disc space with widened posterior interspinous distance and distracted facet joints (< 50% apposition of the articular surfaces).* **(Right)** *Sagittal bone CT through the lateral column of the cervical spine shows diastasis and widening of the C6-C7 facet joint (> 2 mm), indicating ligamentous injury.*





(Left) *Sagittal CT angiogram in this trauma patient shows a "teardrop-type" fracture of the anterior inferior aspect of C6. There is a fracture through the posterior elements as well.* **(Right)** *Sagittal T1WI MR shows flexion injury at C6 with fracture of the anterior inferior corner and disruption of the ALL. There is posterior epidural hemorrhage.*

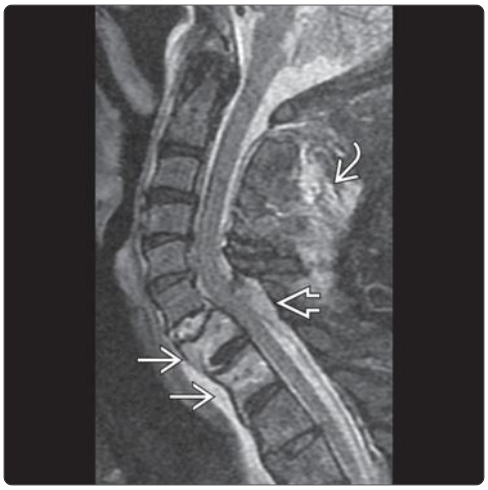




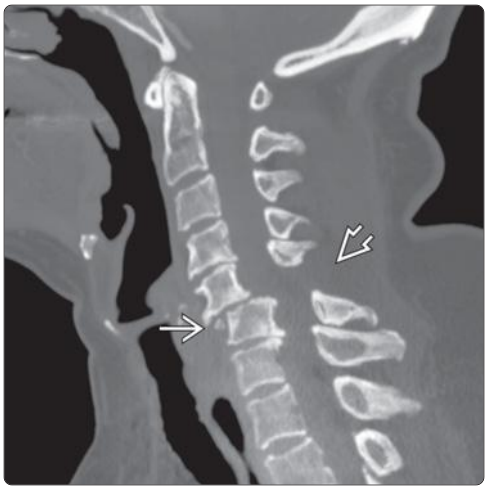
(Left) *Sagittal T2 TSE image in this patient with a flexion teardrop injury at C6 shows abnormal increased signal and ventral cortical disruption. A large epidural hemorrhage severely compresses the cord. Note the ventral and dorsal edema.* **(Right)** *Sagittal DWI MR shows high signal from a large posterior epidural hemorrhage and relatively normal diffusion signal from the cervical cord.*

(Left) *Sagittal bone NECT shows an anterior wedging burst fracture of C7 with anterior subluxation of C6 on C7 and anterior widening of the disc space, with overriding of the lamina.* **(Right)** *Sagittal STIR MR shows the marrow edema from fractures at C7 and T1 with extensive prevertebral edema. There is posterior soft tissue and ligamentous edema. There is a posterior epidural hematoma moderately compressing the cord. No cord hemorrhage is present.*





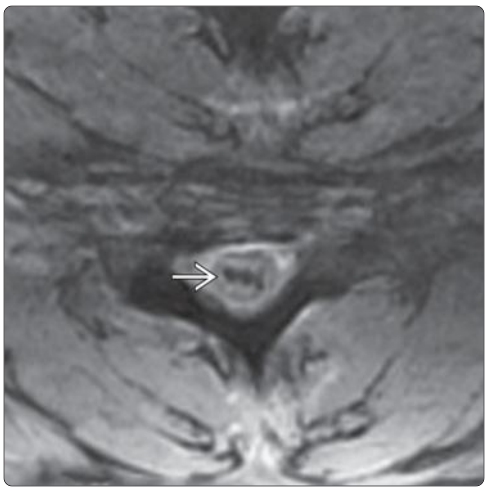
(Left) *CT angiogram shows the typical CT and MR appearance of severe hyperflexion injury with vertebral C5-C6 subluxation with C6 corner fracture and severe widening of the spinous processes, reflecting interspinous ligament disruption.* **(Right)** *Sagittal CT angiogram shows C5-C6 "jumped facets" with a fracture off of the superior articular process of C6. The vertebral artery shows normal luminal enhancement.*

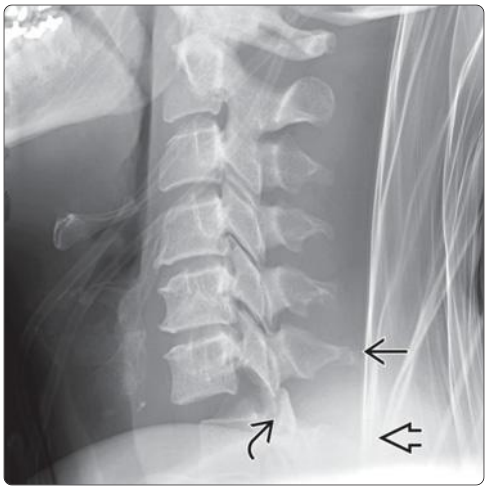


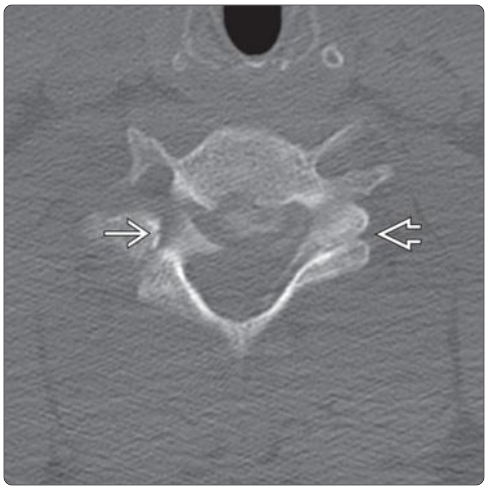


(Left) *Sagittal T2 TSE MR shows disruption of the low signal cortical bone from fractures at C6 and C7. There is extensive cord hemorrhage and edema, and post-traumatic disc herniation. There is extensive posterior soft tissue edema.* **(Right)** *Axial GRE MR in this patient with flexion injury shows extensive cord hemorrhage that is seen as central low signal intensity. This is a poor prognostic sign for functional recovery.*









(Left) *Lateral radiograph shows widening of space between the spinous process of C6 and C7. The inferior facet of C6 jumped the anterior superior facet of C7.* **(Right)** *Axial bone NECT in a severe flexion injury with facet subluxation shows the vertebral body and posterior element comminution, with facet displacement.*





(Left) *Sagittal T2WI MR shows anterolisthesis of C7 on T1 with fanning of spinous processes, edema of intraspinous ligament, and spinal cord edema.* **(Right)** *Sagittal bone CT shows perching of the inferior facet joint of C7 on superior facet joint of T1.*





(Left) *Sagittal bone NECT in a patient with a flexion and axial loading fracture of C7 (burst) with small posterior superior fracture retropulsion. There is no widening of the posterior elements to suggest ligamentous disruption.* **(Right)** *Sagittal STIR MR shows marrow edema at C7 from a burst fracture, with slight bony retropulsion and mild effacement of the ventral thecal sac. The cord signal is normal. Posterior edema relates to additional C2 fracture (not shown).*

Cervical Hyperextension Injury

KEY FACTS

TERMINOLOGY

- Injury from cervical hyperextension with compression or hyperextension with distraction/shearing

IMAGING

- Hyperextension injury with axial compression
  - Unilateral or bilateral fracture of posterior elements
  - Traumatic anterolisthesis with more severe injury; ligamentous disruption ± superior endplate impaction fracture of subjacent vertebral body
- Hyperextension injury with distraction or shear
  - May have minimal radiographic findings even with significant injury
  - ALL rupture, widening of anterior disc space
  - Minimally displaced fracture of anterior margin of inferior vertebral body ("extension teardrop") without retrolisthesis
- Best imaging modality
  - Thin-slice helical CT with sagittal and coronal reformations
  - MR (especially STIR) to evaluate ligaments, cord
    - STIR MR
      - Disruption of ALL, widened and ↑ signal in disc space anteriorly
      - Marrow edema if bone contusion, fracture
      - ↑ signal in cord, if cord injury present

TOP DIFFERENTIAL DIAGNOSES

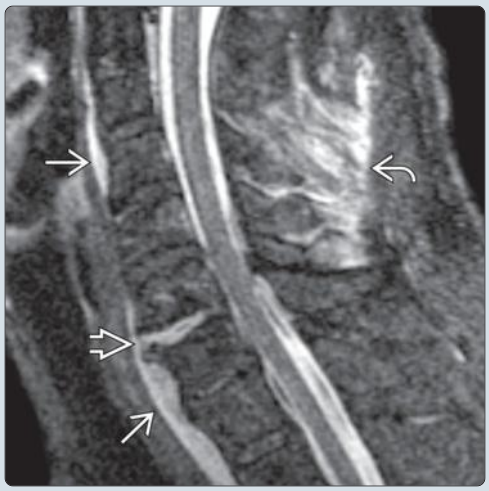
- Flexion "teardrop"
- "Clay shoveler" fracture
- Whiplash injury

PATHOLOGY

- Spinal cord injury
  - Especially central cord syndrome
  - Potential for injury increased with congenital spinal stenosis

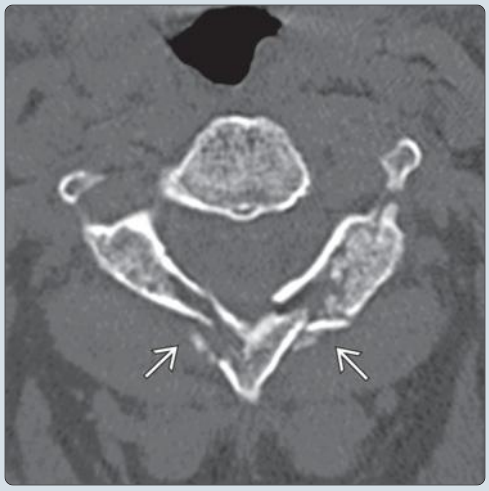
(Left) *Lateral radiograph shows mild prevertebral soft tissue swelling. Mild changes of preexisting cervical spondylosis are also noted.* **(Right)** *Sagittal STIR MR in the same patient shows prevertebral edema and disruption of the ALL. A nonphysiologic hyperintense signal within the C6-C7 disc space reflects disruption of the discovertebral unit by hyperextension-distraction injury at this level. Posterior soft tissue edema is also present.*





(Left) *Axial NECT shows mildly displaced fractures of bilateral C7 articular pillars due to a hyperextension-compression type of injury.* **(Right)** *Axial NECT shows fragmented, mildly displaced fractures of the bilateral lamina due to a hyperextension-compression injury.*





TERMINOLOGY

Definitions

- Injury from cervical hyperextension with compression or hyperextension with distraction/shearing

IMAGING

General Features

- Best diagnostic clue
  - Fracture(s) of neural arch of cervical vertebra
  - Widening of anterior portion of disc space with prevertebral soft tissue swelling
- Location
  - Typically mid or lower cervical spine
- Hyperextension injury with axial compression
  - Unilateral or bilateral fracture of posterior elements
    - Lamina fractures (> 1 adjacent level may be seen)
    - Fracture or compression of articular facet(s), pillar(s)
  - Traumatic anterolisthesis with more severe injury; ligamentous disruption ± superior endplate impaction fracture of subjacent vertebral body
- Hyperextension injury with distraction or shear
  - May have minimal radiographic findings even with significant injury
  - Anterior longitudinal ligament (ALL) rupture, widening of anterior disc space
  - Prevertebral edema, hematoma
  - Posterior displacement of supra-adjacent vertebral body, posterior longitudinal ligament (PLL) failure
  - Minimally displaced fracture of anterior margin of inferior vertebral body ("extension teardrop") without retrolisthesis
    - C2 most common level for extension teardrop

MR Findings

- STIR
  - Prevertebral soft tissue edema, hemorrhage
  - Disruption of ALL, widened and ↑ signal in disc space anteriorly
  - Marrow edema if bone contusion, fracture
  - ↑ signal in cord, if cord injury present

Imaging Recommendations

- Best imaging tool
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations
- Protocol advice
  - CT best to evaluate fracture
  - MR (especially STIR) to evaluate ligaments, spinal cord

DIFFERENTIAL DIAGNOSIS

Flexion Teardrop

- Anteroinferior body "teardrop" fragment, larger than with "extension teardrop" fracture
- Flexed alignment, distraction of posterior column
  - Facet subluxation/distraction
  - Spinous process splaying on lateral film
- Posterior displacement of supra-adjacent vertebral body, canal narrowing

Clay Shoveler Fracture

- Spinous process avulsion from sudden severe force placed on ligamentum nuchae
- Laminae typically spared, spinolaminar line preserved on lateral radiograph

Whiplash Injury

- Predominantly ligamentous injury, neurologic injury rare

PATHOLOGY

General Features

- Etiology
  - Hyperextension with compression
    - Forced extension of cervical spine with axial loading vector posterior to vertebral column
    - Primary injury to posterior column structures (lamina, articular facets) and, later, middle columns
  - Hyperextension with distraction or shearing
    - Forced extension of cervical spine with dorsally oriented force vector
    - Primary failure of anterior column and, later, middle column
      - ALL rupture, widening of anterior disc space
      - Posterior displacement of supra-adjacent vertebral body
      - Displacement spontaneously reduces, imaging underestimates maximal canal occlusion during time of injury
- Associated abnormalities
  - Spinal cord injury
    - Especially central cord syndrome
    - Potential for injury increased with congenital spinal stenosis
  - Radiculopathy
  - Vertebral artery dissection, occlusion
  - Spinal instability
  - Closed head injury, polytrauma

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma ± radiculopathy, myelopathy

Natural History & Prognosis

- Varies with degree of neurologic injury

Treatment

- Immobilization, axial traction, surgical fusion
- Surgical decompression for spinal cord injury
- Edematous cord: High-dose steroids (< 1 day)

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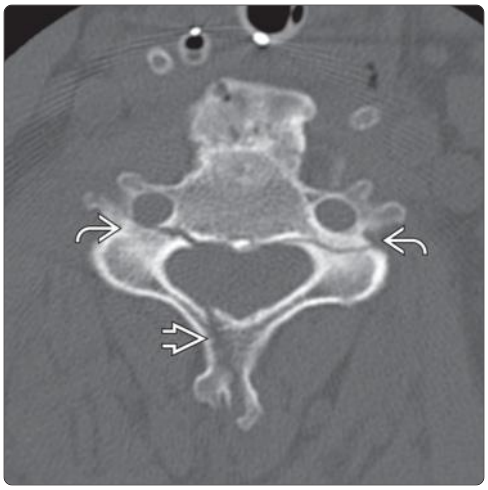
(Left) *Sagittal NECT (bone window) shows the importance of MR imaging to further define subtle or questionable CT abnormalities in patients with trauma. There is mild widening of the anterior disc at C6-C7 and prevertebral swelling.* **(Right)** *Sagittal STIR MR shows an obvious hyperextension injury at C6-C7 with disruption of the ALL, abnormal increased signal in the disc, and extensive prevertebral edema/hemorrhage.*





(Left) *Sagittal NECT (bone window) shows a hyperextension injury with widening of the C4-C5 disc space and fracture through the lamina and spinous process of C4.* **(Right)** *Axial NECT (bone window) in a patient with hyperextension injury shows fractures through the lamina and spinous process of C4 and a vertically oriented fracture through the C4 posterior body at junction with pedicles.*





(Left) *Lateral radiograph in a neutral position shows the normal appearance of disc spaces. C5-C6 and C6-C7 disc spaces become abnormal with extension in a hyperextension injury.* **(Right)** *Lateral radiograph of a trauma patient in extension shows widening of the C5-C6 and C6-C7 anterior disc spaces, consistent with ALL disruption in a hyperextension injury.*







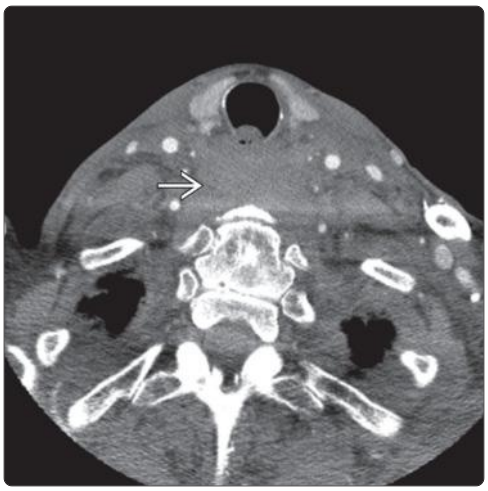


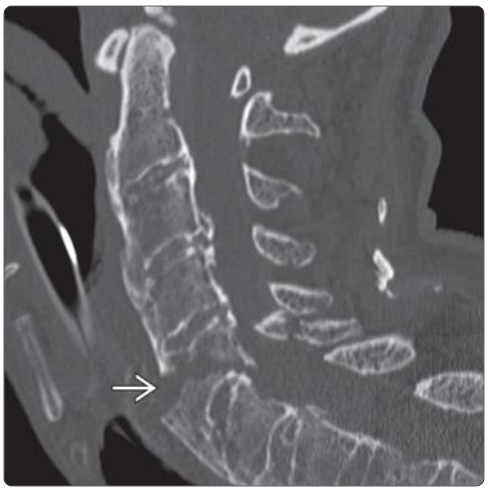
(Left) *Sagittal T1W MR in a hyperextension injury shows abnormal intermediate signal acute epidural hemorrhage ⇨ surrounding the thecal sac from the C3 inferiorly with diffuse cord compression.* **(Right)** *Sagittal T2WI MR shows abnormal increased signal from the disrupted ALL ⇨ at the C5-C6 level in a hyperextension injury. There is extensive acute epidural hemorrhage ⇨ surrounding the thecal sac, in addition to the diffuse cervical spondylosis ⇨.*





(Left) *Sagittal NECT (bone window) shows no definite abnormality in this trauma patient with a diminished level of consciousness and severe neck pain. Clearing the cervical spine in this patient would require MR imaging.* **(Right)** *Sagittal STIR MR in this patient with a normal cervical spine CT shows an abnormal increased signal from the disrupted ALL ⇨ at the C4-5 level and disc. Note the prevertebral edema ⇨.*





(Left) *Axial CT angiogram in a severe trauma patient shows a large prevertebral hematoma ⇨ with a cervical spine extension injury.* **(Right)** *Sagittal NECT (bone window) of the cervical spine shows horizontal fracture dislocation ⇨ through the C5 body and into the base of the spinous process in this patient with ankylosing spondylitis.*

Cervical Hyperextension-Rotation Injury

KEY FACTS

TERMINOLOGY

- Hyperextension injury to cervical spine with off-center force vector causing rotation and asymmetric injury
  - Pedicolaminar fracture-separation

IMAGING

- Asymmetric hyperextension pattern of injury to posterior column
- Below C2; most common at C6 and C7
- Fracture of articular facet
  - Associated with radiculopathy
- Fracture of articular pillar
- Simultaneous fractures of pedicle and ipsilateral lamina = traumatic isolation of cervical articular pillar
- May see degree of rotational deformity at level of injury
- Best imaging modality
  - Thin slice (≤ 1 mm) helical CT with sagittal and coronal reformations
  - MR (especially STIR) to evaluate ligaments, spinal cord
- Radiographs should include oblique views to evaluate facet alignment, articular pillars

TOP DIFFERENTIAL DIAGNOSES

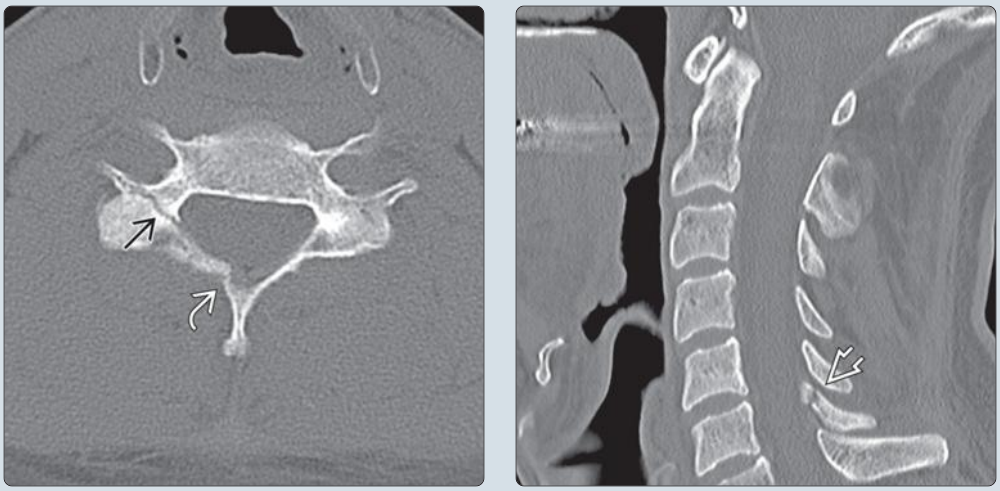
- Hyperflexion injury, cervical
- Hyperflexion-rotation injury, cervical
- Lateral flexion injury, cervical

PATHOLOGY

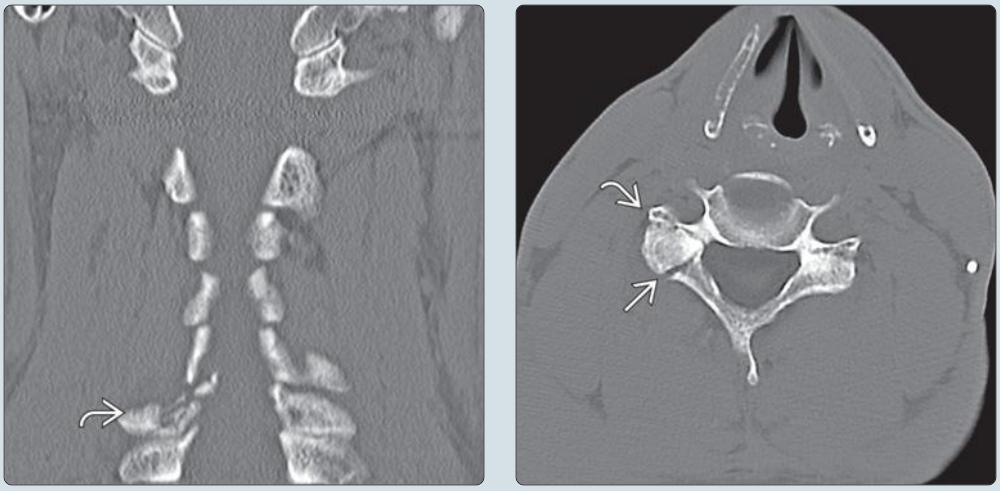
- Rotational instability
- Radiculopathy

CLINICAL ISSUES

- Surgical fusion to achieve rotational stability



(Left) *Axial NECT shows unilateral fractures of the right C6 articular pillar extending into the lamina.* **(Right)** *Sagittal reformatted CT in the same patient shows extension of the articular pillar fracture into the ipsilateral lamina.*



(Left) *Coronal reconstructed CT of the hyperextension-rotation injury shows a unilateral comminuted, mildly displaced fracture involving the right C7 articular pillar.* **(Right)** *Axial NECT shows fractures of the right C6 articular pillar and lamina, resulting in traumatic isolation of the articular pillar.*

TERMINOLOGY

Synonyms

- Pedicolaminar fracture-separation

Definitions

- Hyperextension injury to cervical spine with off-center force vector causing rotation and asymmetric injury

IMAGING

General Features

- Best diagnostic clue
  - Asymmetric hyperextension pattern of injury to posterior column
- Location
  - Below C2; most common at C6 and C7
- Fracture of articular facet
  - Simple fracture or impacted/comminuted
  - Fracture of superior articular facet more commonly associated with radiculopathy
- Fracture of articular pillar
  - Simple fracture or impacted/comminuted
  - May extend into lamina
- Simultaneous fractures of pedicle and ipsilateral lamina = traumatic isolation of cervical articular pillar
  - Unstable injury
- May see degree of rotational deformity at level of injury

MR Findings

- T1WI
  - Demonstrates malalignment but insensitive to fracture
- STIR
  - Soft tissue edema
  - Marrow edema due to bone contusion, fracture

Imaging Recommendations

- Best imaging tool
  - Thin slice (≤ 1 mm) helical CT with sagittal and coronal reformations
- Protocol advice
  - Radiographs should include oblique views to evaluate facet alignment, pillars
  - CT best to evaluate fracture
  - MR (especially STIR) to evaluate ligaments, spinal cord

DIFFERENTIAL DIAGNOSIS

Hyperflexion Injury, Cervical

- Posterior column injury tends to be symmetric
- Cord injury more frequent
- Rotational malalignment absent

Hyperflexion-Rotation Injury, Cervical

- Distraction injury rather than loading: Facets subluxed, perched, or jumped
- Rotational malalignment more noticeable

Lateral Flexion Injury, Cervical

- Vertebral body fracture present

PATHOLOGY

General Features

- Etiology
  - Forced extension of cervical spine
  - Off-center force vector results in asymmetric loading or shear injury to components of facet joint, articular pillars, neural arch
    - Fracture or comminuted, crush injury of facets, pillar
    - Fracture of lamina
  - Rotational listhesis may be present with more severe injury
- Associated abnormalities
  - Rotational instability
  - Radiculopathy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma
  - Radiculopathy
  - Spinal cord injury uncommon

Natural History & Prognosis

- Varies with degree of neurologic injury
- Accelerated degenerative disease may be seen

Treatment

- Options, risks, complications
  - Immobilization and axial traction
    - Failure of 0-50% with conservative therapy
  - Surgical fusion, ± decompression

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Cervical Burst Fracture

KEY FACTS

TERMINOLOGY

- Comminuted fracture of cervical vertebral body due to axial loading

IMAGING

- Loss of vertebral body height
- Vertically oriented fracture planes
  - Extending to endplates and posterior cortex
- Centrifugal displacement of fragments
  - Compromise of spinal canal
  - Cord injury on MR
- Typically mid or lower cervical spine

TOP DIFFERENTIAL DIAGNOSES

- Flexion teardrop fracture
- Hyperextension teardrop fracture
- Benign compression fracture
- Pathologic compression fracture

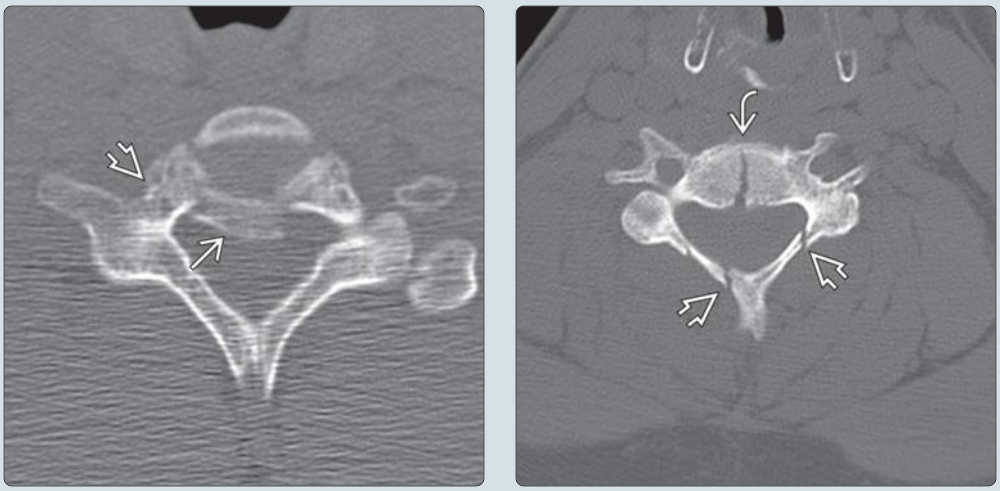
PATHOLOGY

- Axial loading with neck in neutral position
- Centrifugal displacement of comminuted fracture fragments
- Displacement into spinal canal → cord injury
  - Spinal cord injury common

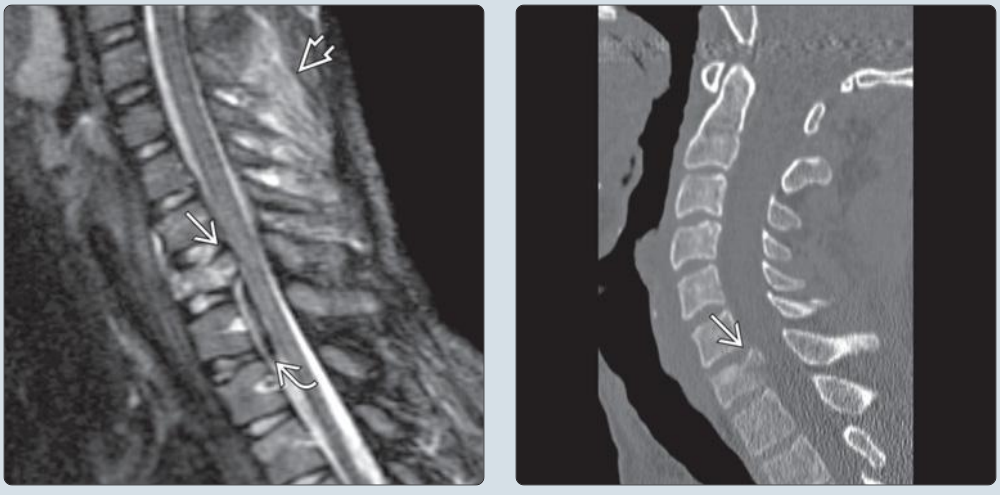
CLINICAL ISSUES

- Variable neurologic symptoms, from no deficit to tetraparesis
- Treatment
  - May be able to be managed conservatively
    - Traction, immobilization
  - Unstable injury may need surgical stabilization
    - ± surgical decompression of canal if cord injury present

(Left) *Axial bone CT shows a vertebral body fragment displaced into the spinal canal. Fracture of the right pedicle is also seen. This is the classic CT appearance of burst fracture, which causes loss of vertebral body height and posterior displacement of bone fragment into the spinal canal.* **(Right)** *Axial NECT shows a burst fracture in the sagittal fracture plane. There is widening of the interpedicular width and multiple fractures of the neural arch.*



(Left) *Sagittal STIR MR shows a C7 burst fracture with posterior displacement of a fracture fragment into the spinal canal, contacting the spinal cord. There are no signal changes in the cord. A small amount of ventral epidural hemorrhage is seen. Edema of the dorsal soft tissue is also present.* **(Right)** *Sagittal reconstructed CT shows a C7 burst fracture with loss of vertebral body height and posterior displacement of bone fragment into the spinal canal.*



TERMINOLOGY

Definitions

- Comminuted fracture of cervical vertebral body
- Vertical compression fracture

IMAGING

General Features

- Best diagnostic clue
  - Loss of vertebral body height, vertical fracture planes, centrifugal displacement of fragments
- Location
  - Typically mid or lower cervical spine

Radiographic Findings

- Radiography
  - Compression deformity of vertebral body
  - Disruption of posterior cortex
    - Fragments displaced into cervical canal
  - Prevertebral soft tissue swelling

CT Findings

- NECT
  - Comminuted vertebral body fracture, loss of height
    - Vertically oriented fracture planes, extending to endplates & posterior cortex
    - Centrifugal displacement of fragments
    - ± interpedicular widening, neural arch fracture
- CTA
  - Screening for vertebral artery dissection or occlusion if fracture extends to foramen transversarium

MR Findings

- T1WI
  - Wedging of vertebral body, cortical disruption
  - Low marrow signal due to edema
  - Prevertebral soft tissue edema
- T2WI
  - Effacement of thecal sac by displaced fracture fragments
  - Hyperintense cord edema if contusion is present
- STIR
  - Best for soft tissue and cord signal abnormality
- T2* GRE
  - Most sensitive for susceptibility artifact associated with blood products in canal or cord
- MRA
  - Irregular contour of vertebral artery (dissection) or occlusion due to associated vascular injury

Angiographic Findings

- Useful if CTA/MRA equivocal, or for persistent concern for vertebral artery injury

Imaging Recommendations

- Best imaging tool
  - Unenhanced multidetector CT scan
- Protocol advice
  - Thin-slice unenhanced axial CT with reformats
  - MR to evaluate cord injury and hemorrhage, include sagittal STIR and axial T2* GRE

DIFFERENTIAL DIAGNOSIS

Flexion Teardrop Fracture (Flexion-Compression)

- Anteroinferior body teardrop fragment, flexed alignment
- Distraction of posterior column
  - Facet subluxation/distraction
  - Spinous process splaying on lateral film

Hyperextension Teardrop Fracture

- Triangular avulsion fragment at anteroinferior margin of C2

Benign Compression Fracture

- No endplate-to-endplate comminution or retropulsion
- Osteoporotic type very rare in cervical spine; think pathologic if present

Pathologic Compression Fracture

- Destruction of cancellous and cortical bone, soft tissue mass
- Bowing of posterior cortex into epidural space

PATHOLOGY

General Features

- Etiology
  - Axial loading with neck in neutral position
  - Centrifugal displacement of comminuted fracture fragments
    - Displacement into spinal canal → cord injury
  - Spreading of vertebral body in lateral plane → fracture through neural arch
    - May be unstable injury
- Associated abnormalities
  - Spinal cord injury common with retropulsion

Gross Pathologic & Surgical Features

- Comminuted fracture of vertebral body, hematoma

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Variable neurologic symptoms, from no deficit to tetraparesis
- Clinical profile
  - Severe neck pain, disability after trauma with high-energy/high-velocity axial loading

Treatment

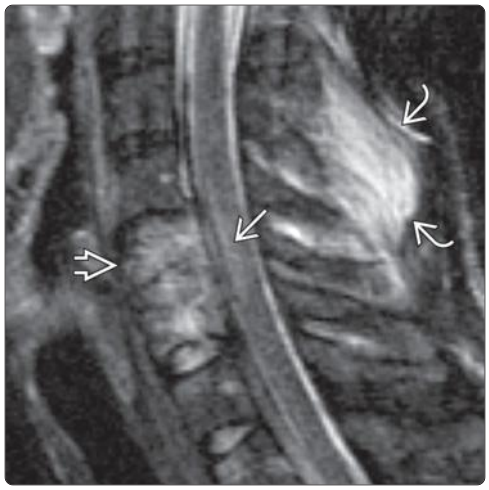
- Unstable injury may need surgical stabilization, others can be managed conservatively
- Traction, immobilization
- ± surgical decompression of canal if cord injury present

SELECTED REFERENCES

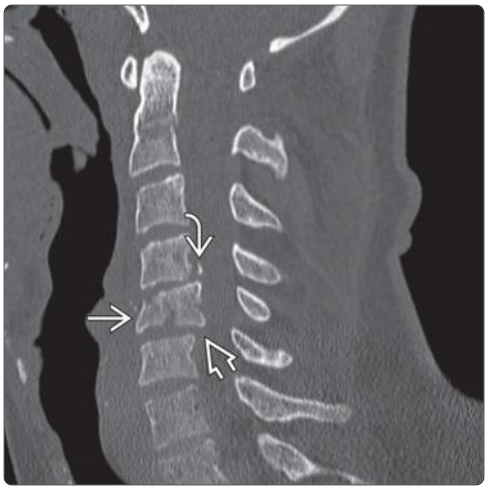
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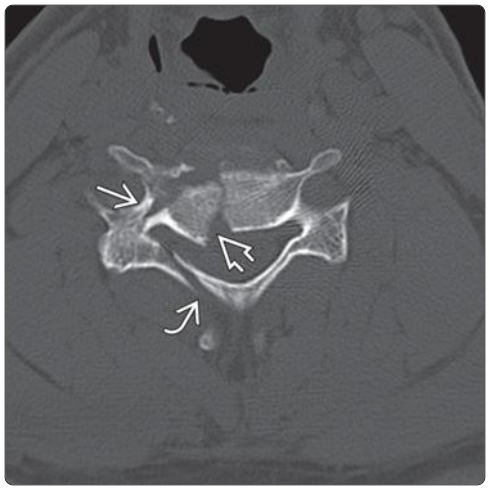
(Left) *Axial bone CT through the C5 vertebra shows a complex fracture pattern with a sagittally oriented vertebral body fracture ➡ from a burst-type injury and right pedicle ➡ lamina ➡ fracture separation.* **(Right)** *Sagittal STIR MR shows marrow edema ➡ from compressive fractures without malalignment. There is cord contusion ➡. Posterior ligamentous and muscular injuries are also evident ➡.*



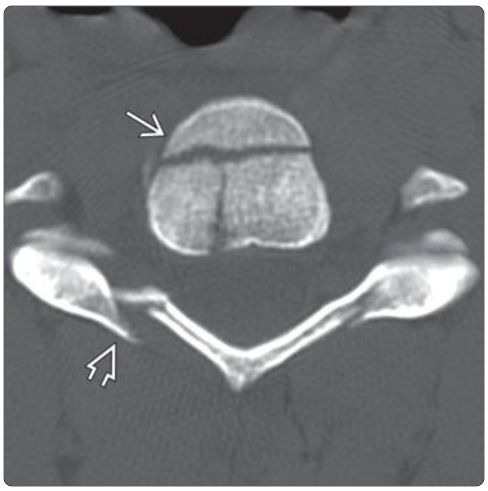


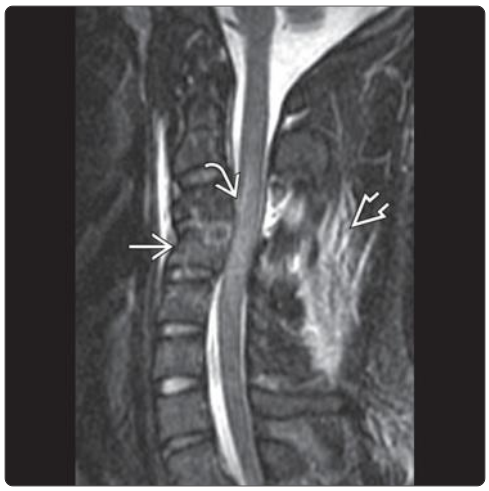
(Left) *Sagittal NECT (bone window) shows a comminuted burst fracture of C5 ➡ with vertical disruption and loss of height. There is posterior displacement of the fracture fragment ➡. An additional fracture is present in the posterior aspect of C4 ➡.* **(Right)** *Axial NECT (bone window) shows a comminuted burst fracture of C5 with vertical disruption, posterior displacement, and rotation of the fracture fragment ➡. Fractures extend through the right pedicle ➡ and right lamina ➡.*





(Left) *Axial NECT (bone window) shows the typical pattern of a burst fracture ➡ through the cervical vertebral body, with both transverse and sagittal components. There is also a fracture of the right facet and lamina, consistent with a rotatory component to the injury ➡.* **(Right)** *Sagittal STIR MR shows a compression fracture of C4 ➡, a flexion component to the injury mechanism with slight C3-C4 deformity, and posterior ligamentous edema ➡. There is also extensive cord contusion ➡.*







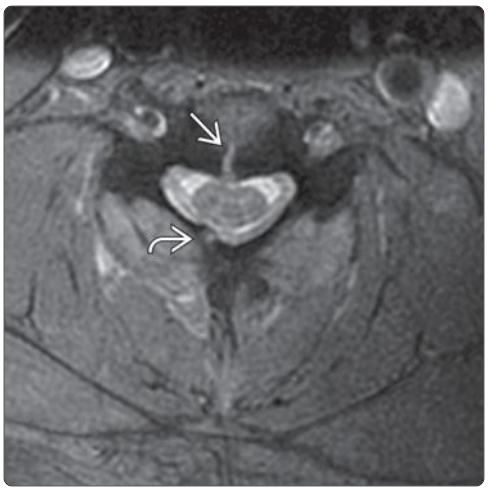


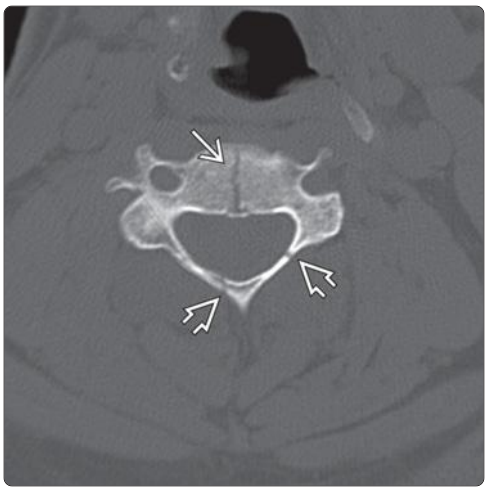
(Left) *Coronal CT reformat shows a vertical fracture through the C5 vertebral body in this patient with a C5 burst fracture. An additional fracture is seen involving the superior aspect of the C6 body.* **(Right)** *Sagittal T2* GRE MR shows extensive edema and low signal hemorrhage within the cervical cord in this patient with a C5 burst fracture. There is moderate retropulsion of the posterior aspect of the C5 body.*





(Left) *Sagittal NECT (bone window) shows a C5 burst fracture with mild height loss, as well as a 2nd mild burst fracture of the C4 and posterior element fracture at C6.* **(Right)** *Coronal NECT (bone window) shows C4, C5, and C6 burst fractures with predominately sagittal fractures through the vertebral bodies.*





(Left) *Axial GRE MR in this patient with a burst fracture shows the sagittal fracture line and disruption of the right lamina.* **(Right)** *Axial NECT (bone window) shows a sagittal fracture site in this patient with an axial compression injury. Additional fractures are present in the right and left lamina.*

Cervical Hyperflexion-Rotation Injury

KEY FACTS

TERMINOLOGY

- Hyperextension injury to cervical spine
  - Off-center force vector causes rotation, asymmetric injury

IMAGING

- Asymmetric hyperextension pattern of injury to posterior column
- Below C2
  - Most common sites = C6 and C7
- Fracture of articular facet
  - Associated with radiculopathy
- Fracture of articular pillar
- Simultaneous fractures of pedicle and ipsilateral lamina = traumatic isolation of cervical articular pillar
- May see degree of rotational deformity at level of injury
- Best imaging modality
  - Thin-slice (≤ 1 mm) helical CT
  - Sagittal and coronal reformations
  - MR (especially STIR) to evaluate ligaments, spinal cord
- Radiographs should include oblique views to evaluate facet alignment, articular pillars

TOP DIFFERENTIAL DIAGNOSES

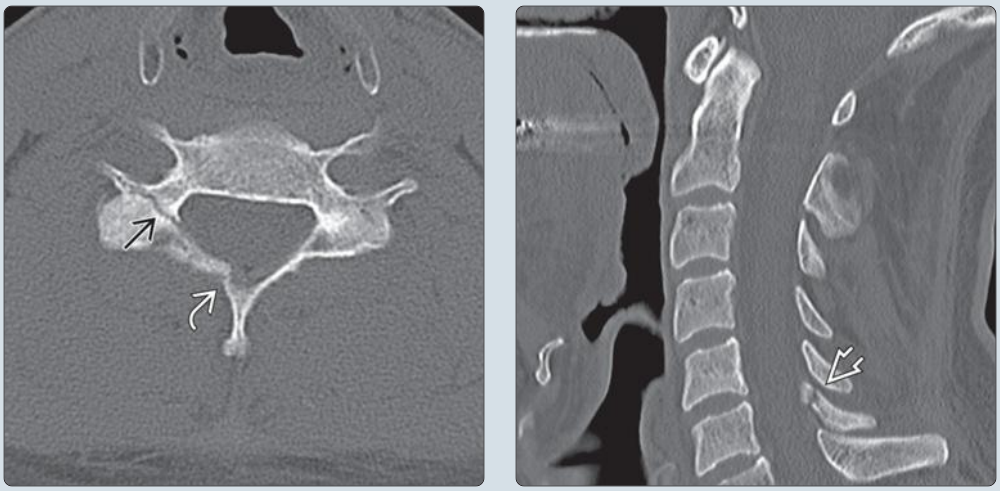
- Hyperflexion injury, cervical
- Hyperflexion-rotation injury, cervical
- Lateral flexion injury, cervical

PATHOLOGY

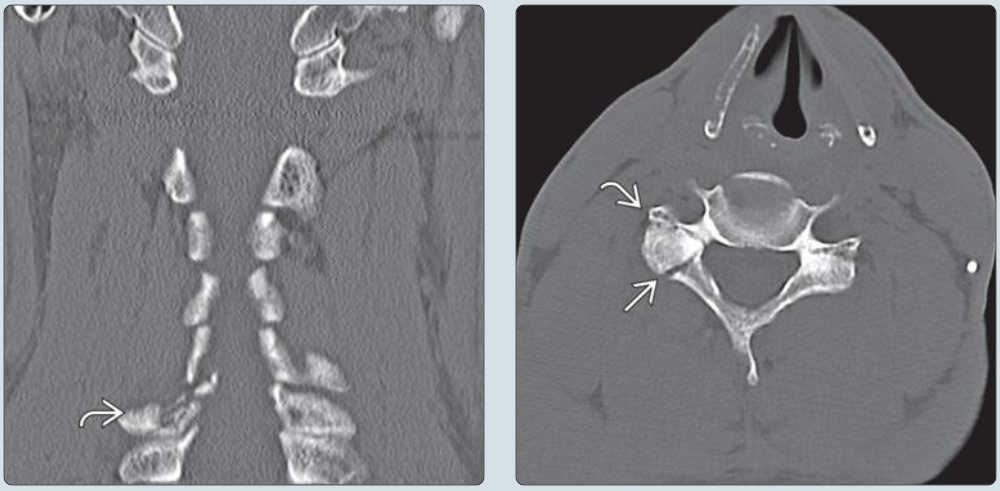
- Rotational instability
- Radiculopathy

CLINICAL ISSUES

- Surgical fusion to achieve rotational stability



(Left) *Axial NECT shows unilateral fractures of the right C6 articular pillar extending into the lamina.* **(Right)** *Sagittal reformatted CT in the same patient shows extension of the articular pillar fracture into the ipsilateral lamina.*



(Left) *Coronal reconstructed CT of a hyperextension-rotation injury shows a unilateral comminuted, mildly displaced fracture involving the right C7 articular pillar.* **(Right)** *Axial NECT (bone window) shows fractures of the right C6 articular pillar and lamina, resulting in traumatic isolation of the articular pillar.*

TERMINOLOGY

Definitions

- Hyperextension injury to cervical spine with off-center force vector causing rotation and asymmetric injury

IMAGING

General Features

- Best diagnostic clue
  - Asymmetric hyperextension pattern of injury to posterior column
- Location
  - Below C2; most common at C6 and C7
- Fracture of articular facet
  - Simple fracture or impacted/comminuted
  - Fracture of superior articular facet more commonly associated with radiculopathy
- Fracture of articular pillar
  - Simple fracture or impacted/comminuted
  - May extend into lamina
- Simultaneous fractures of pedicle and ipsilateral lamina = traumatic isolation of cervical articular pillar
  - Unstable injury
- May see degree of rotational deformity at level of injury

MR Findings

- T1WI
  - Demonstrates malalignment but insensitive to fracture
- STIR
  - Soft tissue edema
  - Marrow edema due to bone contusion, fracture

Imaging Recommendations

- Best imaging tool
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations
- Protocol advice
  - Radiographs should include oblique views to evaluate facet alignment, pillars
  - CT best to evaluate fracture
  - MR (especially STIR) to evaluate ligaments, spinal cord

DIFFERENTIAL DIAGNOSIS

Hyperflexion Injury, Cervical

- Posterior column injury tends to be symmetric
- Cord injury more frequent
- Rotational malalignment absent

Hyperflexion-Rotation Injury, Cervical

- Distraction injury rather than loading: Facets subluxed, perched, or jumped
- Rotational malalignment more noticeable

Lateral Flexion Injury, Cervical

- Vertebral body fracture present

PATHOLOGY

General Features

- Etiology
  - Forced extension of cervical spine
  - Off-center force vector results in asymmetric loading or shear injury to components of facet joint, articular pillars, neural arch
    - Fracture or comminuted, crush injury of facets, pillar
    - Fracture of lamina
  - Rotational listhesis may be present with more severe injury
- Associated abnormalities
  - Rotational instability
  - Radiculopathy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma
  - Radiculopathy
  - Spinal cord injury uncommon

Natural History & Prognosis

- Varies with degree of neurologic injury
- Accelerated degenerative disease may be seen

Treatment

- Options, risks, complications
  - Immobilization and axial traction
    - Failure of 0-50% with conservative therapy
  - Surgical fusion, ± decompression

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Cervical Lateral Flexion Injury

KEY FACTS

TERMINOLOGY

- Lateral bending cervical spine injury due to laterally applied force vector to side of head or upper neck

IMAGING

- Mid to lower cervical spine more common
  - Fracture of transverse process or uncinate process
  - Fracture of uncinate process
  - Lateral vertebral body compression fracture
  - Crushing fracture of articular pillar &/or facet
  - Ligamentous failure on side of impact, focal angulation on frontal views
- Best imaging modality
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations

TOP DIFFERENTIAL DIAGNOSES

- Hyperextension-rotation injury, cervical
- Hyperflexion-rotation injury, cervical

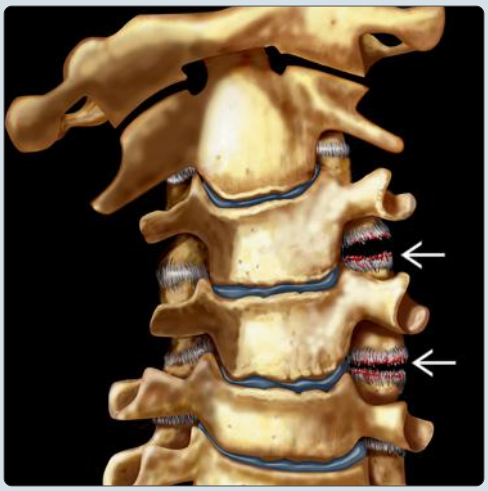
PATHOLOGY

- Associated injuries
  - Asymmetric fracture(s) of C0-C2
  - Occipital condyle
  - Superior articular process of C2
  - Brachial plexus or cord injury
- Allen mechanistic classification (1982)
  - Lateral flexion is major compressive injury causing vertebral arch fracture on one side and minor distractive injury on opposite side
- Harris classification (1986)
  - Uncinate process fracture
- Subaxial cervical injury classification system (SLIC) (2007)
  - Rotation/translation injury giving 4 points

CLINICAL ISSUES

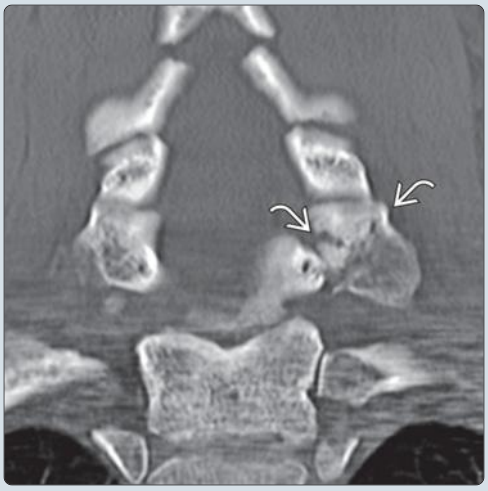
- Mild lateral vertebral body compression may be considered stable injury

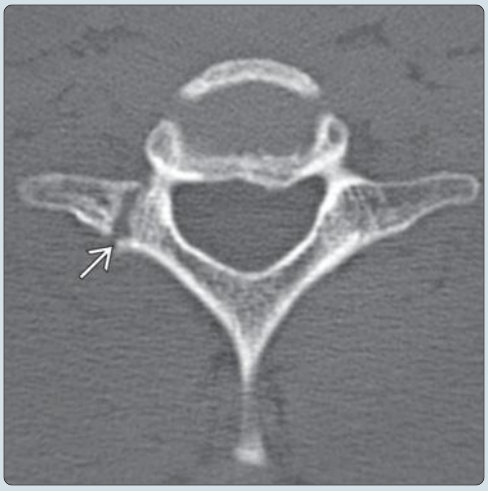
(Left) *Anterior graphic shows failure of left C3-C4 and C4-C5 facet joint capsular ligaments due to tension from lateral flexion injury.* **(Right)** *Sagittal bone CT shows a severely comminuted fracture of lateral mass of facet and pedicle. Healing of these unilateral, nondisplaced fractures is unpredictable under physiological loads and may be treated by conservatively or by surgical fusion.*





(Left) *Coronal CT reconstruction shows comminuted fracture of the left C7 articular pillar due to compression associated with forced lateral flexion of the head and neck.* **(Right)** *Axial NECT shows nondisplaced fracture of the right transverse process of C7.*





TERMINOLOGY

Definitions

- Cervical spine injury caused by laterally applied force vector to side of head or upper neck
  - Specific types of fractures included in this category vary widely depending upon the specific classification system utilized (mechanistic vs. morphological)

IMAGING

General Features

- Best diagnostic clue
  - Compression fracture of lateral vertebral body &/or articular pillar
- Mid to lower cervical spine
  - Fracture of transverse process, or uncinate process
  - Lateral vertebral body compression fracture
    - Lateral wedging of vertebral body
    - May encounter sagittally oriented vertebral body fracture
    - Focal lateral angulation deformity of cervical spine
  - Crushing fracture of articular pillar &/or facet
    - May be difficult to appreciate with radiography
    - Ipsilateral neural arch fracture
  - Ligamentous failure on side of impact, focal angulation on frontal views
    - Diastasis of facet joint

MR Findings

- T1WI
  - Hypointense marrow due to edema
    - ± lateral vertebral wedging deformity, best seen with coronal imaging plane
- STIR
  - Marrow edema due to fracture
    - Lateral vertebral body
    - Articular pillar
  - Paravertebral soft tissue edema
  - Hyperintensity on side of injury if ligamentous tension injury present

Imaging Recommendations

- Best imaging tool
  - Thin-slice (≤ 1 mm) helical CT with sagittal and coronal reformations
- Protocol advice
  - Radiographic evaluation must include AP and oblique views; will be missed on lateral film

DIFFERENTIAL DIAGNOSIS

Hyperextension-Rotation Injury, Cervical

- Asymmetric posterior column injury
  - Loss of articular pillar height not seen
- No lateral wedging of vertebral body

Hyperflexion-Rotation Injury, Cervical

- Asymmetric posterior column injury
  - Facet diastasis, subluxation, fracture
  - Articular pillar, neural arch fracture
- No lateral wedging of vertebral body

PATHOLOGY

General Features

- Etiology
  - Forced lateral flexion of cervical spine
  - Compression injury of lateral vertebral body, articular pillar, transverse process contralateral to force vector
  - With more severe injury, may see distractive ligamentous injury ipsilateral to force vector
- Associated abnormalities
  - Asymmetric fracture(s) of C0-C2
    - Occipital condyle
    - Lateral mass of C1
    - Superior articular process of C2
  - Brachial plexus or cord injury
  - Fractures at other levels of cervical spine

Staging, Grading, & Classification

- Allen mechanistic classification (1982)
  - Lateral flexion is a major compressive injury causing vertebral arch fracture on one side, and minor distractive injury on opposite side
  - More severe injuries show vertebral body displacement
- Harris classification (1986)
  - Uncinate process fracture
- Subaxial cervical injury classification system (SLIC) (2007)
  - Rotation/translation injury giving 4 points

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma
- Other signs/symptoms
  - Brachial plexus lesion, radiculopathy

Natural History & Prognosis

- Widely variable depending on neurological injury

Treatment

- Options, risks, complications
  - Mild lateral vertebral body compression may be considered stable injury
  - More extensive deformity, involvement of articular pillar/neural arch, significant neural lesion: May require surgical fusion

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Cervical Posterior Column Injury

KEY FACTS

IMAGING

- Fractures of
  - Superior and inferior articular facets
  - Articular pillars
  - Pedicles, lamina
- Disruption of
  - Interspinous ligament
  - Ligamentum flavum
  - Facet joint capsules
- Mechanisms of cervical posterior column injury
  - Hyperextension with axial compression
  - Hyperflexion with axial compression
  - Hyperflexion with distraction
  - Lateral flexion injury
  - Fracture-dislocation, hyperextension or hyperflexion injuries with rotational component
- Bony elements of posterior column best evaluated with CT
- Ligamentous structures best evaluated with MR

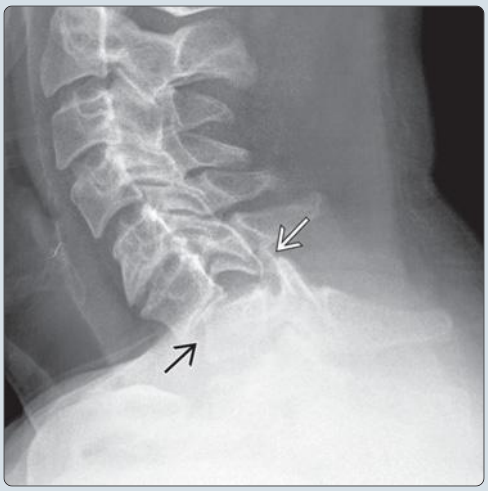
PATHOLOGY

- Subaxial Cervical Spine Injury Classification Scale (SLIC)
  - Numerical value given to 1 of each 3 components; sum of which is added to produce total score
  - Components
    - Morphology: Compression 1, burst 2, distraction 3, translation rotation 4
    - Discoligamentous complex: Intact 0, indeterminate 1, disrupted 2
    - Neurological status: Normal 0, root injury 1, complete cord injury 2, incomplete cord injury 3
- Scores higher than or equal to 5, surgery recommended

CLINICAL ISSUES

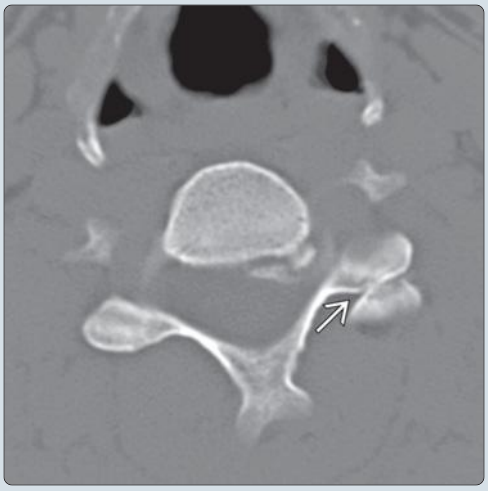
- Prognosis primarily depends on degree of neurologic injury
- Treatment decisions may depend on identifying mechanism of injury and potential for instability

(Left) *Lateral cervical spine radiograph shows bilateral jumped facets at C6-C7 due to a hyperflexion-distraction injury. Also noted is traumatic anterolisthesis of C6 on C7.* **(Right)** *Sagittal reformatted CT shows one of the bilateral C7 articular pillar fractures secondary to a hyperextension-compression injury.*





(Left) *Axial NECT shows the classic appearance of a unilateral jumped facet secondary to a hyperflexion-rotation pattern of cervical spine injury.* **(Right)** *Axial NECT shows a fracture through the right C7 articular pillar and lamina from a hyperextension-rotation pattern of injury.*





TERMINOLOGY

Definitions

- Posterior column of spine defined as structures dorsal to posterior longitudinal ligament
  - Superior and inferior articular facets, articular pillars, lamina and spinous processes
  - Facet joint capsules
  - Ligamentum flavum
  - Interspinous and supraspinous ligaments

IMAGING

General Features

- Posterior column injury seen as component of many mechanisms of cervical spine injury
- Posterior column failure in compression
  - Hyperextension with axial compression
    - Lamina fractures
    - Fracture, compression of articular facet(s), pillar(s)
  - Hyperflexion with axial compression
    - Disruption of interspinous ligament: Widening of interspinous distance
    - Distraction of facet joints
  - Lateral flexion injury
    - Crushing fracture of articular pillar &/or facet on direction of flexion
- Posterior column failure in tension
  - Hyperflexion with distraction
    - Disruption of interspinous ligament: Widening of interspinous distance
    - Diastatic (> 2 mm), subluxed (articular apposition < 50%), perched, or jumped facet joints
  - Lateral flexion injury
    - Facet diastasis on side opposite direction of flexion
- Posterior column failure in shear
  - Fracture-dislocation, hyperextension or hyperflexion injuries with rotational component
    - Horizontal fracture or fracture-dislocation of facet joints
    - Fractures of articular pillar, pedicle, lamina

MR Findings

- T2WI/STIR
  - Hyperintense T2 signal associated with disruption of soft tissue structures
    - Interspinous ligament
      - Weakest of the spinal ligamentous complex
      - Abnormal signal within only interspinous ligament may still be a stable fracture
    - Ligamentum flavum
    - Facet joint capsules
      - Strongest of spinal ligamentous complex
      - Disruption suggests instability
  - Hyperintense marrow signal due to fracture of posterior elements

Imaging Recommendations

- Best imaging tool
  - Thin-slice helical CT
    - Coronal and sagittal reformations to evaluate fracture

PATHOLOGY

Staging, Grading, & Classification

- Subaxial Cervical Spine Injury Classification Scale (SLIC)
  - Numerical value given to 1 of each 3 components; sum of which is added to produce total score
    - Scores higher than or equal to 5, surgery recommended
  - Components
    - Morphology: Compression 1, burst 2, distraction 3, translation rotation 4
    - Discoligamentous complex: Intact 0, indeterminate 1, disrupted 2
    - Neurological status: Normal 0, root injury 1, complete cord injury 2, incomplete cord injury 3

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma
  - Radiculopathy

Natural History & Prognosis

- Depends on degree of neurologic injury
- Accelerated degenerative change
- Spinal instability

Treatment

- Options, risks, complications
  - Treatment may depend on identifying mechanism of injury and potential for instability
  - Treatment modalities include
    - Immobilization
    - Traction
    - Surgical fusion

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Correlating with type of injury to anterior and middle columns necessary to determine mechanism of injury
- Characterizing mechanism of injury has utility to degree that doing so changes patient management
  - High-energy trauma may lead to injuries that are not clearly assignable to single mechanism

SELECTED REFERENCES

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Traumatic Disc Herniation

KEY FACTS

TERMINOLOGY

- Traumatic disruption of annularis fibrosis with associated displacement of nucleus pulposus

IMAGING

- Location: Cervical > thoracic > > lumbar
- Radiographs insensitive for disc pathology
  - Cause of spinal cord injury without radiographic abnormality (SCIWORA)
- Associated with spinal fracture, facet subluxation
- MR is modality of choice to evaluate intervertebral discs, soft tissue contents of spinal canal
  - Disc material effacing anterior CSF ± cord or nerve root compression
  - ± cord edema if cord compressed

TOP DIFFERENTIAL DIAGNOSES

- Nontraumatic intervertebral disc herniation
- Epidural abscess, phlegmon
- Epidural tumor

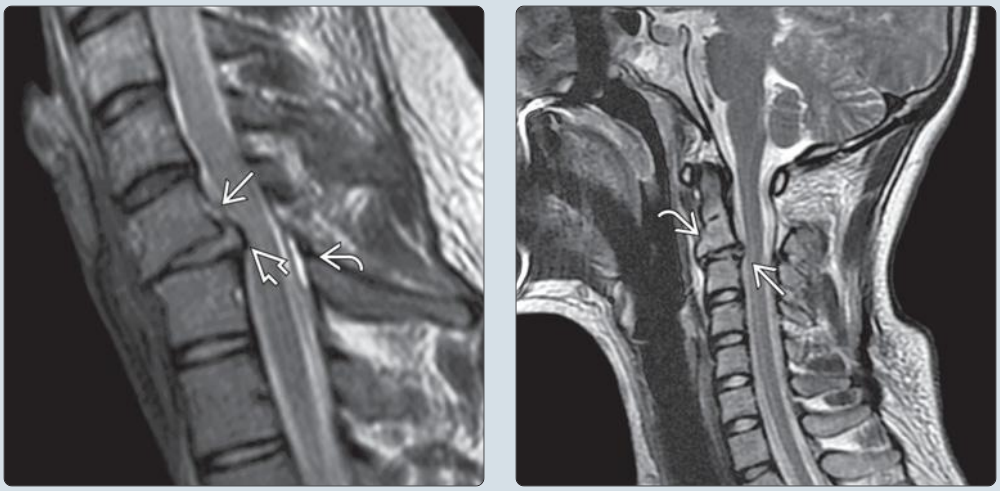
CLINICAL ISSUES

- Anterior cord syndrome highly associated with traumatic disc herniation
- Cauda equina syndrome with lumbar herniation: Pain, incontinence 2° to compression of lumbosacral spinal nerve roots
- 5-54% incidence of herniation with cervical spine trauma
- Cord compression from herniation is reported complication of reduction of cervical facet dislocation without discectomy

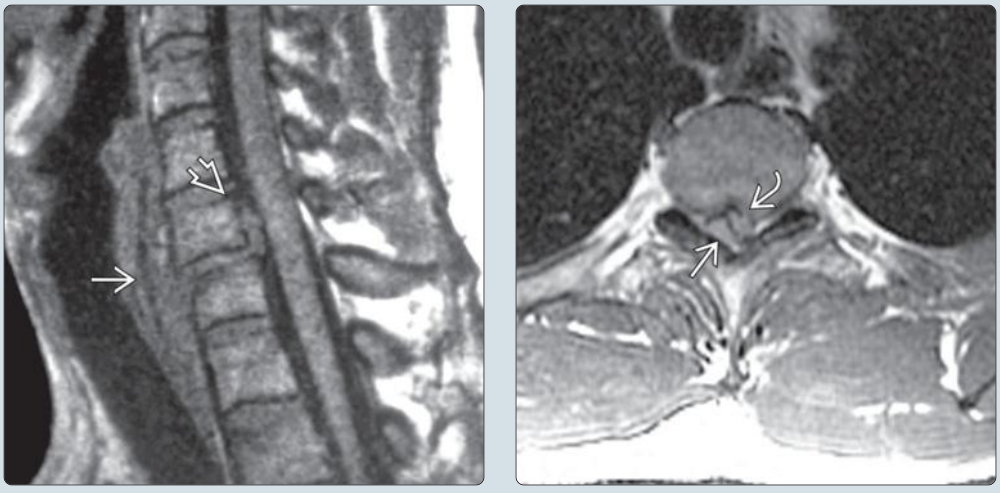
DIAGNOSTIC CHECKLIST

- Consider MR prior to reduction of facet dislocation
- MR to exclude anterior cord compression from herniated disc or hematoma prior to aggressive attempts at closed reduction

(Left) *Sagittal T2WI MR shows hyperflexion-compression injury, with focal kyphosis of C6-C7, disruption of the posterior longitudinal ligament, narrowing of the anterior disc space, and a posterior disc protrusion causing canal stenosis. There is splaying of the spinous processes and a small amount of dorsal epidural hematoma.* **(Right)** *Sagittal T2WI MR shows flexion teardrop fracture of C2, increased kyphosis at C2-C3, and a traumatic disc protrusion effacing the thecal sac at C2-C3.*



(Left) *Sagittal T1WI MR shows C5-C6 flexion injury with subluxation, prevertebral hemorrhage, and disc extrusion touching ventral cord.* **(Right)** *Axial T1WI MR shows traumatic disc extrusion in the thoracic canal, associated with canal stenosis and compression of the thoracic cord.*



TERMINOLOGY

Definitions

- Traumatic disruption of annularis fibrosis with associated displacement of nucleus pulposus

IMAGING

General Features

- Best diagnostic clue
  - Disc herniation on MR or CT myelography, with evidence of spinal trauma on imaging or history
- Location
  - Cervical > thoracic > > lumbar
  - At or adjacent to level of facet dislocation
- Size
  - Any
- Morphology
  - Same appearance as degenerative disc herniation

Radiographic Findings

- Radiography
  - Insensitive for disc pathology
    - Cause of spinal cord injury without radiographic abnormality (SCIWORA)
  - May see disc space narrowing
  - May see other signs of spinal injury
    - Focal kyphosis, traumatic spondylolisthesis, dislocated facet joints
    - Fracture, compression deformity

CT Findings

- NECT
  - Soft tissue density in anterior or anterolateral epidural space
    - Difficult to differentiate from epidural hematoma
  - ± narrowing of disc space
  - Spinal fracture, facet subluxation
- CT myelography
  - Ventral extradural mass effacing anterior thecal sac ± cord or nerve compression

MR Findings

- T2WI
  - Disc material effacing anterior CSF ± cord or nerve root compression
  - ± cord edema if cord compressed
  - ± disruption of posterior longitudinal ligament

Imaging Recommendations

- Best imaging tool
  - MR is modality of choice to evaluate intervertebral discs, soft tissue contents of spinal canal

DIFFERENTIAL DIAGNOSIS

Nontraumatic Intervertebral Disc Herniation

- Appears similar to traumatic herniation on imaging; other signs of spinal trauma absent

Epidural Abscess, Phlegmon

- Usually associated with spondylodiscitis, instrumentation
- May extend > 1 spinal segment; will enhance

Epidural Tumor

- Epidural extension of vertebral metastases, aggressive hemangioma; lymphoma

PATHOLOGY

General Features

- Etiology
  - Annularis fibrosis constrains nucleus pulposus within disc space under load
  - Traumatic disruption of annulus → herniated nucleus compressing spinal cord, nerve roots
- Associated abnormalities
  - Spinal cord injury, especially anterior cord syndrome
  - Facet joint dislocation, posterior longitudinal ligament (PLL) tear

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, disability following cervical trauma ± radiculopathy, myelopathy
  - Anterior cord syndrome highly associated with traumatic disc herniation
    - Pain & temperature lost, touch & vibration intact
  - Cauda equina syndrome: Pain, incontinence 2° to compression of lumbosacral spinal nerve roots

Demographics

- Epidemiology
  - 5-54% incidence with cervical spine trauma

Natural History & Prognosis

- Varies with degree of neurologic injury

Treatment

- Cord compression from traumatic herniation is reported complication of reduction of cervical facet dislocation without discectomy
- Controversial, but prereduction MR to rule out anterior compressive pathology is commonly utilized
  - MR to exclude anterior cord compression from herniated disc or hematoma prior to aggressive attempts at closed reduction

DIAGNOSTIC CHECKLIST

Consider

- MR of spine prior to reduction of facet dislocation

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Thoracic and Lumbar Burst Fracture

KEY FACTS

TERMINOLOGY

- Vertebral body fracture due to axial load, involving anterior and middle, ± posterior columns

IMAGING

- Usually at thoracolumbar junction
- Loss of vertebral body height
- Fracture involves posterior vertebral body cortex
  - ± retropulsion of posterior cortex
  - ± vertically oriented posterior element fractures
  - ± widening interpediculate distance on AP radiograph
- Cord contusion best seen on axial T2WI MR

TOP DIFFERENTIAL DIAGNOSES

- Compression fracture
- Chance fracture
- Pathologic fracture due to tumor
- Fracture-dislocation

PATHOLOGY

- Associated with other spinal fractures, pelvic and lower extremity fractures

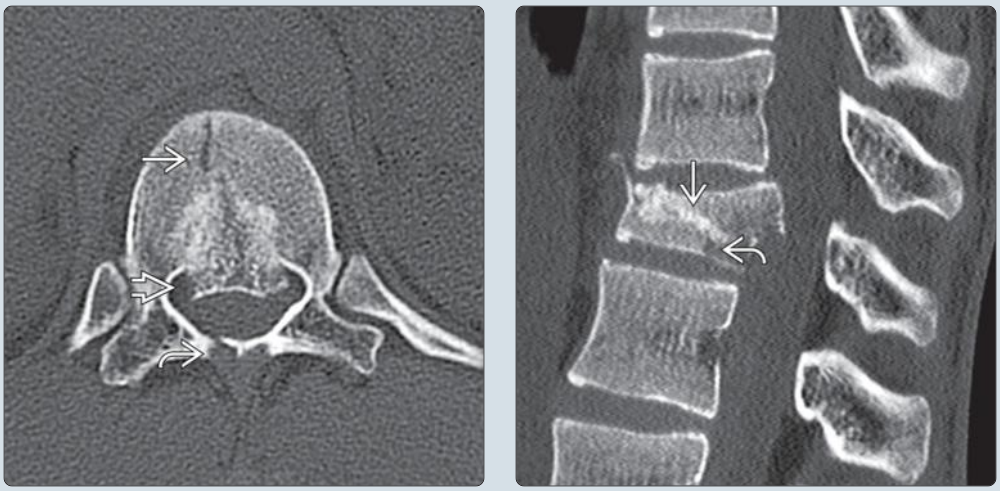
CLINICAL ISSUES

- Canal narrowing decreases significantly in size within 1 year following injury

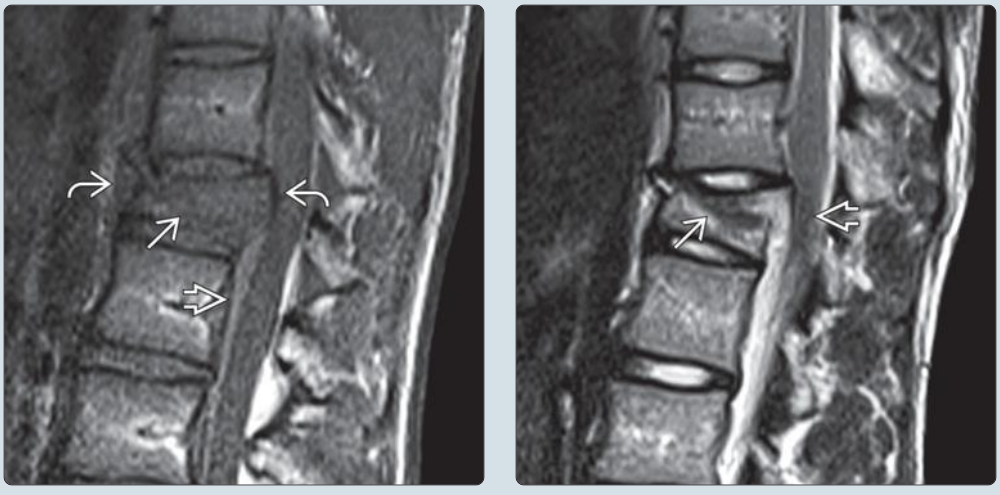
DIAGNOSTIC CHECKLIST

- Orientation of posterior element fractures useful to distinguish between Chance and burst fractures
  - Burst: Vertically oriented posterior element fractures reflect axial load force
  - Chance: Horizontally oriented posterior element fractures reflect distraction force

(Left) *Axial bone CT shows sagittal fracture line through vertebral body. The axial load force has continued through the right lamina. The posterior vertebral body cortex is displaced posteriorly.* **(Right)** *Sagittal bone CT in the same patient shows a sclerotic line reflecting trabecular impaction. There is also a coronal fracture line extending to the inferior vertebral body cortex.*



(Left) *Sagittal T1WI MR in the same patient shows the body deformity and epidural hematoma. However, the fracture line is difficult to see because of the surrounding red marrow.* **(Right)** *Sagittal T2WI MR in the same patient better shows the low signal intensity fracture line outlined by marrow hematoma. The tip of the conus is displaced posteriorly.*



TERMINOLOGY

Definitions

- Vertebral body fracture due to axial load, involving anterior and middle, ± posterior columns
- Anterior column: Anterior longitudinal ligament, anterior 1/2 vertebral body, & anterior anulus fibrosis
- Middle column: Posterior longitudinal ligament, posterior 1/2 vertebral body, & posterior anulus fibrosis
- Posterior column: Neural arch, facet joint capsular ligaments, ligamentum flavum, inter- & supraspinous ligaments

IMAGING

General Features

- Location
  - Usually at thoracolumbar junction
  - 60% of thoracolumbar spine injuries of all types occur between T11 and L1
    - Region is known as transition zone
  - Thoracic spine relatively stabilized by ribs/sternum
- Morphology
  - Loss of vertebral body height
  - Fracture extends through posterior vertebral body cortex
    - ± retropulsion of posterior cortex

Radiographic Findings

- Compression of vertebral body on anteroposterior, lateral radiography
  - Usually anterior portion of body compressed more than posterior
- Fracture involves posterior vertebral body cortex
  - Do not mistake vascular groove in center of posterior cortex for fracture line
- Posterior displacement of posterior vertebral body cortex may occur
  - Usually involves superior portion of body
- Neural arch fractures may occur
  - Vertically oriented
  - Widening of interpediculate distance on anteroposterior radiography
    - Present in more severe burst fractures
    - Axial load fractures pedicles, displaces them laterally
    - Normally, interpediculate distance widens in caudad direction
      - e.g., T12 interpediculate distance < L1 interpediculate distance < L2
  - Laminar fractures seen in more severe injuries

CT Findings

- Fracture of vertebral body extending to posterior cortex
  - Often have sagittally oriented fracture line
  - Often severely comminuted
- ± retropulsed bone fragments in spinal canal
  - Percentage of spinal canal narrowing should be reported
- ± vertically oriented posterior element fractures
- ± widening interpediculate distance
- Can be diagnosed on routine CT scan of abdomen in multitrauma patients

MR Findings

- Fracture line may be difficult to see
  - Low signal intensity on all sequences
- Band-like marrow edema surrounds fracture line
  - Low signal intensity on T1WI
    - May blend with erythropoietic marrow
  - High signal intensity on T2WI, STIR
- Retropulsion of posterior cortex best seen on T2WI
  - Axial T2WI to evaluate for compression of neural elements
- Cord contusion best seen on axial T2WI
  - High signal intensity in cord
  - May be mimicked by artifact on sagittal images

Imaging Recommendations

- Best imaging tool
  - CT for surgical planning
  - MR for evaluation of cord injury/epidural hematoma
- Protocol advice
  - Coronal, sagittal reformations essential

DIFFERENTIAL DIAGNOSIS

Compression Fracture

- Posterior vertebral body cortex intact
- Posterior element fractures absent
- < 40-50% loss of vertebral body height in patients with normal bone density

Chance Fracture

- Anterior vertebral body compression
- Horizontal fractures posterior elements **or**
- Separation of facet joints + spinous processes
- Retropulsion of posterior cortex may also be present ("burst-Chance")

Pathologic Fracture Due to Tumor

- More commonly compression fracture, but sometimes burst fracture
- Mass may be obscured by fracture and hematoma
- MR often shows rounded area of abnormal signal intensity
  - Distinguished from band-like configuration of traumatic fracture
- CT may show trabecular, cortical destruction
- Identification of other tumor sites is helpful

Fracture-Dislocation

- Due to shear injury or distraction
- Spondylolisthesis of vertebra
- ALL, PLL injury usually evident on STIR MR

PATHOLOGY

General Features

- Etiology
  - Most commonly due to falls from height
  - Insufficiency fracture: Minimal trauma
- Associated abnormalities
  - Dural laceration
  - Other spine fractures: Contiguous or noncontiguous
  - Pelvic fractures

- Lower extremity fractures: Calcaneus, tibial plateau, tibial plafond
- Adolescents may have greenstick fracture of posterior elements
- Degree of neurologic injury with lumbar burst highly dependent on position of conus
- Lower lumbar burst fractures may have minimal or no neurologic signs

Staging, Grading, & Classification

- **Denis classification**
  - Type A: Fracture of both superior and inferior endplates
    - Primarily seen in lower lumbar region
  - Type B: Fracture of superior endplate
    - Most common type
    - Primarily seen at thoracolumbar junction
  - Type C: Fracture of inferior endplate
  - Type D: Burst plus malrotation due to rotary force as well as axial load
  - Type E: Burst plus lateral compression due to lateral flexion as well as axial load
  - Spine mechanical instability: Involvement of 2 of 3 columns is necessary but not sufficient
  - Spine neurologic instability: Neurologic injury, especially to spinal cord
- **Thoracolumbar Injury Classification and Severity Score (TLICS)**
  - 3 injury categories: (1) injury morphology, (2) integrity of the posterior ligamentous complex (PLC), and (3) neurologic status of the patient
  - Point values from the 3 categories are totaled to provide a severity score
  - Identify injury features that are important in predicting biomechanical and neurologic spinal stability, facilitating appropriate treatment

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Traumatic back pain, ± lower extremity neurologic deficits

Demographics

- Epidemiology
  - Construction workers
  - Mountain climbers
  - Sky divers
  - Skiers
  - Sometimes osteoporotic insufficiency fracture
  - Occasionally pathologic fracture due to tumor

Natural History & Prognosis

- Fragments in spinal canal remodel over time
  - Canal narrowing decreases significantly in 1st year after injury
  - Best seen on CT scan but can be evaluated on lateral radiography

Treatment

- Options, risks, complications
  - Systemic steroids generally given at time of injury
  - Conservative management for neurologically intact patients, mild fracture, lower lumbar fracture
    - Long-term outcomes similar to fusion
  - Laminectomy and posterior fusion used if there is
    - Neurologic dysfunction
    - > 20-30° kyphosis
    - Subluxation of facet joints
    - > 50% loss of vertebral body height

DIAGNOSTIC CHECKLIST

Consider

- Position of vertebral fragments in canal on CT not true measure of displacement at time of injury
  - Fragments tend to move forward after initial posterior excursion
- High association with lower extremity & pelvis fractures

Image Interpretation Pearls

- Since spinal column is curved, axial load may not compress all 3 columns evenly
  - Often see mild widening of interspinous distance in burst fracture
  - Should not lead to classification as Chance fracture unless
    - Horizontally oriented posterior element fracture present **or**
    - Separation of facet joints present
- Orientation of posterior element fractures useful to distinguish between Chance and burst fractures
  - Burst: Vertically oriented posterior element fractures reflect axial load
  - Chance: Horizontally oriented posterior element fractures reflect distraction force

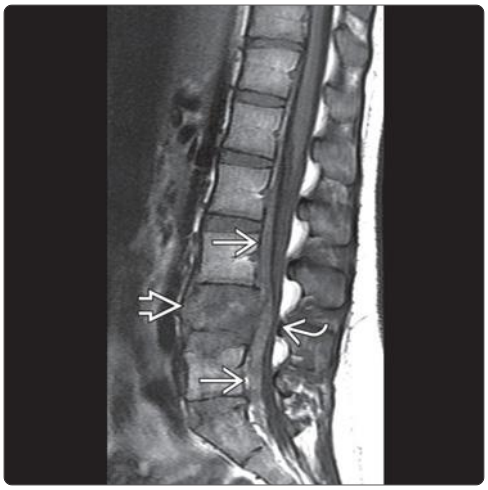
Reporting Tips

- Assessment of spine stability or instability is based on imaging + clinical
- Best to report injury extent without pronouncing spine stable or unstable based on imaging alone

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(Left) *CT study shows extensive comminuted burst fracture involving the L4 body, with anterior displacement of a fracture fragment, as well as severe posterior retropulsion.* **(Right)** *T1WI MR shows low signal from the fractured L4 body. The retropulsed fragments compress the ventral thecal sac, and there is a very large epidural hemorrhage. There is severe thecal sac compression due to the combination of retropulsion and epidural hemorrhage.*





(Left) *Sagittal CT shows L1 burst fracture with retropulsed posterior cortex. The fracture line extends into the posterior vascular groove. The normal vascular groove is distinguished from fracture by its smooth contour and characteristic location.* **(Right)** *Coronal bone CT in the same patient shows a vertical pars interarticularis fracture. This is characteristic of axial load injury, contrasted to the horizontally oriented posterior element fracture seen in flexion-distraction injury.*





(Left) *Sagittal bone CT shows severe burst fracture of L5 with a large fragment in the spinal canal. Despite the canal narrowing, the patient was neurologically intact.* **(Right)** *Sagittal bone CT shows burst fractures of T5 and T3. Since the thorax is stabilized by the ribs and sternum, burst fracture of the upper thoracic spine indicates a severe injury force.*

Facet-Lamina Thoracolumbar Fracture

KEY FACTS

IMAGING

- Fractures of articular facets and lamina typically seen as components of more extensive patterns of thoracic and lumbar spine injury
  - Burst fracture
    - Fracture of lamina with widening of interpediculate distance
  - Chance fracture
    - Distracted horizontal fracture through neural arch
  - Fracture-dislocation
    - Shearing fracture through articular facets
  - Hyperextension
    - Fractures of spinal process, lamina
- Distinctive involvement of anterior and middle columns of spine with each injury pattern
- Elements of posterior column spared with compression fracture
- 60% of thoracolumbar fractures occur between T12 and L2
  - Rib cage and sternum provide structural stability to majority of thoracic spine
- Imaging recommendations
  - Thin-slice helical CT with multiplanar reformations
  - MR for ligaments, intervertebral disc, spinal cord

PATHOLOGY

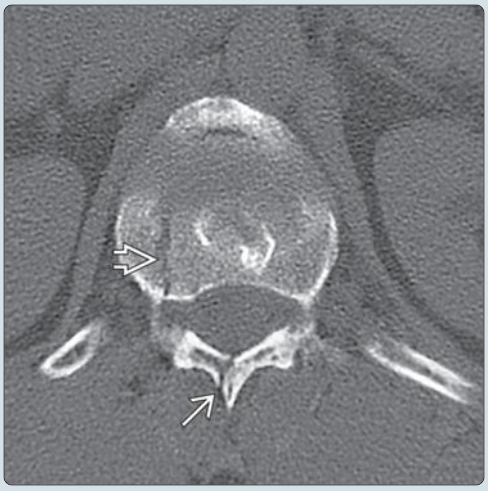
- High-energy mechanism with most spinal injuries resulting in fractures of facets and lamina
  - Motor vehicle accidents
  - Falls
  - Sport-related injuries
  - Penetrating injuries
- Associated abnormalities
  - Radiculopathy
  - Spinal cord injury
  - Dural tears

(Left) *Sagittal reconstructed bone CT of a patient with thoracic fracture-dislocation shows fractures of the lamina and posterior vertebral body, along with anterior wedging of the vertebral body above the level of injury and a mild traumatic anterolisthesis.* **(Right)** *Coronal reconstructed bone CT in a patient with a bony Chance fracture shows a horizontal fracture extending through the lamina and pars interarticularis of L4.*





(Left) *Axial NECT shows a portion of the burst fracture through T12. There is also a thin fracture through the lamina just to the right of the spinous process.* **(Right)** *Sagittal reconstructed bone CT of a patient with a low thoracic flexion-distraction injury shows fracture through the right lamina and pars interarticularis, and distraction of the facet joint. There is a tiny avulsion of the posterior vertebral body from attachment of the PLL.*





TERMINOLOGY

Synonyms

- Neural (vertebral) arch fracture
- Posterior column fracture

Definitions

- Posterior column of spine defined (by Denis) as structures dorsal to posterior longitudinal ligament
  - Superior and inferior articular facets, articular pillars, lamina and spinous processes
  - Facet joint capsules
  - Ligamentum flavum
  - Interspinous and supraspinous ligaments

IMAGING

General Features

- Best diagnostic clue
  - Fractures of articular facets and lamina typically seen as components of more extensive patterns of thoracic and lumbar spine injury
- Location
  - 60% of thoracolumbar fractures located between T12 and L2
    - Rib cage and sternum provide structural stability to thoracic spine
- **Burst fracture**
  - Axial loading injury, comminuted fracture of anterior and middle columns
  - Centrifugal displacement of fragments can cause lamina fracture(s)
    - Widening of interpediculate distances on radiography
- **Hyperflexion-distraction and Chance fractures**
  - Forced flexion of spine around anterior fulcrum
    - Flexion-distraction: Diastatic (> 2 mm), subluxed (articular apposition < 50%), perched, or jumped facet joints
      - "Naked facet" sign on CT
    - Chance fracture: Horizontal fracture(s) through pedicles, articular pillar, lamina
- **Fracture-dislocation**
  - Shear injury to spine causing displacement in AP or lateral directions
  - Fractures of neural arch
  - Horizontal fractures of articular facets with anterior fracture-dislocation
- **Hyperextension**
  - Forced hyperextension of spine
  - Fractures of lamina, spinous processes
    - Especially with mechanism of posterior impact
    - May be present at several adjacent levels

Radiographic Findings

- Radiography
  - Malalignment of vertebral column
    - Traumatic listhesis with fracture-dislocation
      - Fractures through articular facets
    - Focal kyphosis with hyperflexion-distraction or hyperextension injuries
      - Horizontal fractures through pedicles, articular pillars, lamina with bony Chance fracture
  - Fracture of lamina with widening of interpediculate distance with unstable burst fracture
  - Fractures of lamina, spinous processes from direct blow to back
  - Widening of paraspinal line due to hematoma

CT Findings

- CT myelography
  - May see extravasation of intrathecal contrast with dural laceration
    - If dural laceration, look for nerve root entrapment within fracture

MR Findings

- T2WI, STIR
  - Soft tissue edema associated with ligamentous disruption
  - Marrow edema and fluid associated with fractures of posterior elements
  - Epidural fluid associated with dural tears
  - Increased signal in spinal cord indicates contusion

Imaging Recommendations

- Best imaging tool
  - Thin-slice helical CT with multiplanar reformations
  - MR for ligaments, intervertebral disc, spinal cord

PATHOLOGY

General Features

- Etiology
  - Motor vehicle accidents
  - Falls
  - Sport-related injuries
  - Penetrating injuries
- Associated abnormalities
  - Radiculopathy
  - Spinal cord injury
  - Dural tears

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Fracture Dislocation

KEY FACTS

TERMINOLOGY

- Transversely applied force vector, resulting in shearing injury ± flexion &/or rotation

IMAGING

- Traumatic spondylolisthesis with fracture(s) of posterior elements &/or vertebral body
  - Vertebral column discontinuity/listhesis at level of injury, involving 3 columns
  - Complete disruption of disc or horizontal fracture through vertebral body with displacement &/or rotation
  - Soft tissue edema associated with ligamentous disruption
- Spinal cord injury common

TOP DIFFERENTIAL DIAGNOSES

- Chance fracture
  - Distracted, horizontal fracture through pedicles, no listhesis
- Burst fracture
  - Comminuted fracture of vertebral body extending to posterior cortex

PATHOLOGY

- Force vector perpendicular to vertebral column, ± rotation &/or flexion
- Shear injury, failure of all 3 columns
- High-energy mechanisms

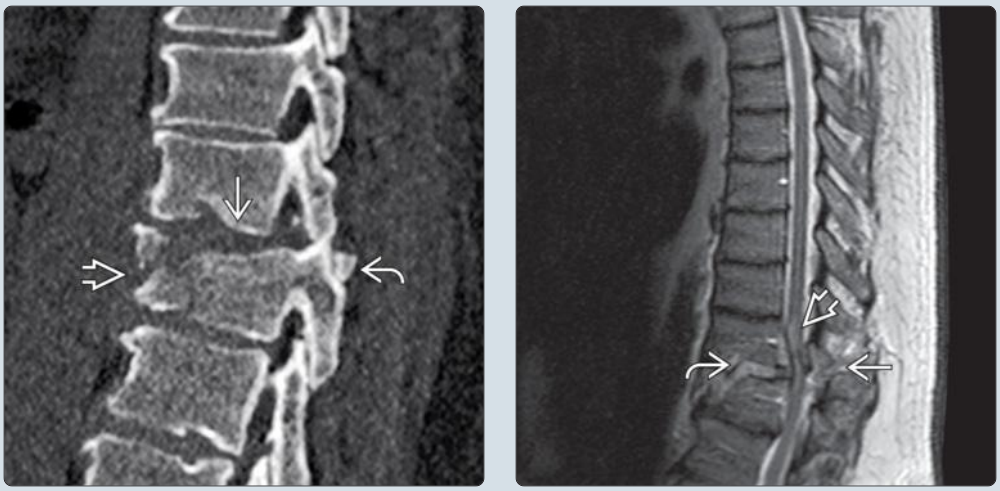
CLINICAL ISSUES

- Severely injured trauma patient, high-energy mechanism
- Lower extremity paralysis, sensory deficit
- Spinal shock with hypotension

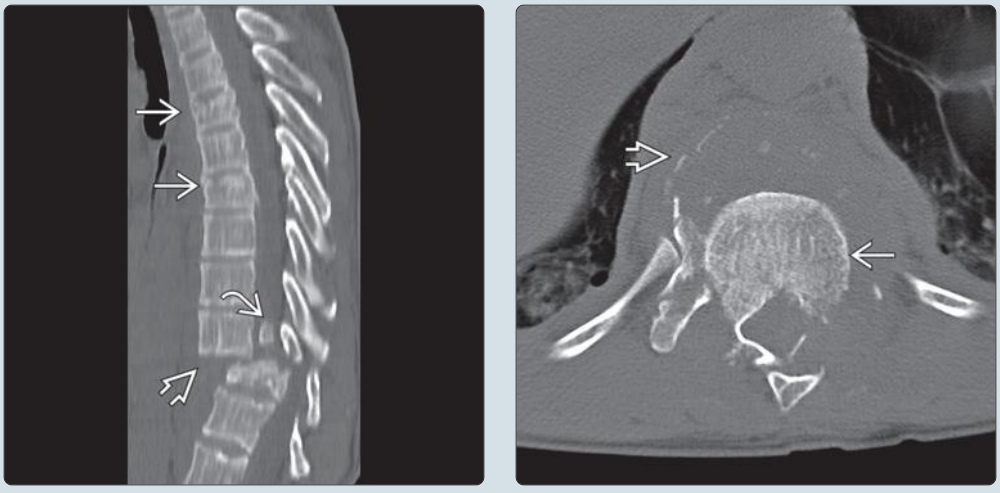
DIAGNOSTIC CHECKLIST

- Fracture-dislocation injuries have high rate of paraplegia, instability

(Left) *Sagittal CT reconstruction shows a widened T10-T11 disc space with mild traumatic anterolisthesis. The inferior T10 facet is fractured and perched. There is a fragmented fracture of the anterior T11 vertebral body.* **(Right)** *Sagittal T2WI MR in the same patient shows abnormal signal and widening of the T10-T11 disc space and fracture of the T10 spinous process and lamina. Traumatic anterolisthesis causes spinal canal stenosis and spinal cord contusion.*



(Left) *Sagittal reconstructed bone CT shows anterior fracture-dislocation of T10-T11 with comminuted fracture of the T11 vertebral body. There is severe canal stenosis due to 75% anterior translation and bone fragment displaced into the spinal canal. Mild compression fractures are noted at T5 and T7.* **(Right)** *Axial NECT (bone window) in the same patient shows the margin of T10 anteriorly displaced relative to T11. Thickening of the paravertebral soft tissues is due to hematoma.*



TERMINOLOGY

Definitions

- Transversely applied force vector, resulting in shearing injury ± flexion &/or rotation

IMAGING

General Features

- Best diagnostic clue
  - Traumatic spondylolisthesis with fracture(s) of posterior elements &/or vertebral body

Radiographic Findings

- Radiography
  - Displacement of spinal column at level of trauma in AP or lateral planes ± rotation
  - Widening of disc space or horizontal fracture through vertebral body
  - Vertebral body height tends to be preserved, compression minimal or absent

CT Findings

- Bone CT
  - Vertebral column discontinuity and listhesis at level of injury, involving all 3 columns
  - Complete disruption of intervertebral disc, or horizontal fracture through vertebral body with displacement &/or rotation
  - Horizontal fracture through facets with anterolisthesis
  - Fracture through neural arch: Pedicles, lamina

MR Findings

- T2WI, STIR
  - Marrow edema and fluid associated with fractures of vertebral body, posterior elements
  - Abnormally ↑ signal in widened disc space
  - Ligamentous injury
    - Soft tissue edema associated with ligamentous disruption
    - Disruption of ALL, PLL with anterior listhesis, stripping of ALL with posterior listhesis
  - Spinal cord injury common
    - Increased signal in spinal cord = contusion
    - Discontinuity of cord = transection
- T2* GRE
  - Hypointensity within cord indicates hemorrhage, heralds poorer prognosis

Imaging Recommendations

- Best imaging tool
  - Thin-slice helical bone CT with multiplanar reformations
  - MR for ligaments, intervertebral disc, spinal cord

DIFFERENTIAL DIAGNOSIS

Chance Fracture

- Injury due to forced flexion around anterior fulcrum
- Distracted, horizontal fracture through pedicles, no listhesis

Burst Fracture

- Comminuted fracture of vertebral body extending to posterior cortex
- Loss of vertebral body height
- Spondylolisthesis absent

PATHOLOGY

General Features

- Etiology
  - Force vector perpendicular to vertebral column, ± rotation &/or flexion
  - Shear injury, failure of all 3 columns
  - High-energy traumatic mechanisms
- Associated abnormalities
  - Closed head injury
  - Life-threatening intrathoracic and abdominal injuries

Gross Pathologic & Surgical Features

- Spinal cord contusion, hemorrhage, transection
  - Cord tends to be spared in variant injury pattern of fracture-dislocation with bilateral pedicle fractures and separation of neural arch from vertebral body at injury level
- Spinal instability nearly universal

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Severely injured trauma patient, high-energy mechanism
  - Lower extremity paralysis, sensory deficit, absent sphincter tone
- Other signs/symptoms
  - Spinal shock with hypotension

Natural History & Prognosis

- High rate of spinal cord, nerve root injury
  - High incidence of paraplegia (> 80%)
- Complications of paraplegia
  - Deep venous thrombosis
  - Decubitus ulcers
- Often high comorbidity associated with other injuries

Treatment

- Surgical fusion ± decompression
- Intensive rehabilitation

DIAGNOSTIC CHECKLIST

Consider

- Fracture-dislocation injuries have high rate of paraplegia, instability

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Chance Fracture

KEY FACTS

TERMINOLOGY

- Flexion-distraction injury, seat belt fracture
- Compression injury of anterior column with distraction of middle and posterior columns

IMAGING

- Usually occurs at T11-L3, occasionally midthoracic
- Wedging of anterior vertebral body
- Focal kyphosis ± fracture
- Bony or ligamentous injury to posterior column
  - Transversely oriented posterior element fracture &/or
    - Separation of facet joints
    - Increased interspinous distance
    - Ligament disruption on MR
- No subluxation of vertebral body

TOP DIFFERENTIAL DIAGNOSES

- Shear injury
- Distraction injury
- Burst fracture
- Traumatic compression fracture
- Pathologic vertebral fracture

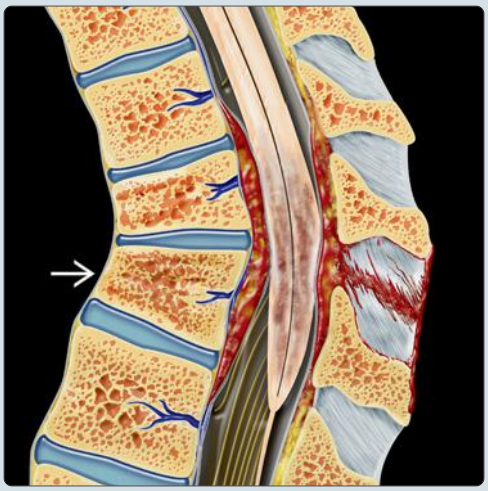
PATHOLOGY

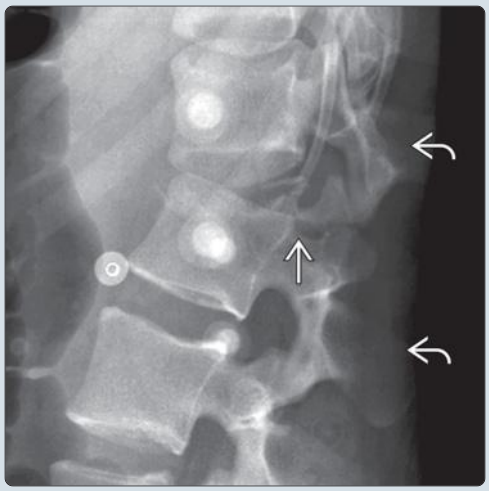
- Anterior compression, posterior distraction around fulcrum
- 15-80% have significant abdominal injuries (bowel and mesentery most common)

DIAGNOSTIC CHECKLIST

- MR evidence of hematoma between spinous processes is not sufficient to diagnose ligament disruption
  - Hematoma may occur due to compression force
  - Look for discontinuity of ligament on MR
- Chance fracture may have retropulsion of posterior vertebral body cortex mimicking burst fracture

(Left) *Sagittal graphic shows osteoligamentous Chance injury. There is a compression fracture of the vertebral body, but posterior distraction injury is ligamentous.* **(Right)** *Lateral radiograph shows a different variant in the osteoligamentous pattern. Wide separation of the spinous processes indicates rupture of supraspinous and interspinous ligaments. A fracture extends through the pedicle and superior articular facet into the vertebral body.*





(Left) *Sagittal T2WI MR in the same patient shows rupture of the interspinous and supraspinous ligaments. The ALL and PLL appear intact.* **(Right)** *Sagittal T2WI MR in the same patient shows vertebral body fracture and facet joint disruption. The combination of fracture and ligamentous injury is variable in Chance injury, but all types show evidence of anterior compression and posterior distraction without an anterior to posterior vector force.*





TERMINOLOGY

Synonyms

- Flexion-distraction injury, seat belt fracture

Definitions

- Compression injury of anterior column with distraction of middle and posterior columns
- Anterior column: Anterior longitudinal ligament (ALL), anterior 1/2 of vertebral body, anterior annulus fibrosis
- Middle column: Posterior longitudinal ligament (PLL), posterior 1/2 of vertebral body, posterior annulus fibrosis
- Posterior column: Posterior neural arch, facet joint capsular ligaments, ligamentum flavum, inter- and supraspinous ligaments
- Transitional zone: T11-L1 vertebrae, highly susceptible to fracture

IMAGING

General Features

- Best diagnostic clue
  - Wedging anterior vertebral body and posterior element fracture &/or increased intraspinous distance (ligamentous injury)
- Location
  - Usually occurs from T11-L3 levels
    - 78% occur between T12 and L2 levels
  - Occasionally at midthoracic spine
  - May have anterior injury at one level, posterior injury at adjacent one

Radiographic Findings

- Radiography
  - Wedging of anterior vertebral body
    - Sclerotic transverse fracture line due to trabecular impaction
    - Often > 40-50% loss of vertebral body height
    - Wedging often greater than seen in compression fracture in patients with normal bone density
  - Focal kyphosis ± fracture
  - Bony or ligament injury to posterior column
    - Separation of facet joints
    - Increased interspinous distance
    - Transversely oriented posterior element fracture
      - Fracture fragments usually mildly separated
  - No subluxation of vertebral body
  - "Empty body" sign on AP radiograph
    - Increased radiolucency secondary to displaced spinous processes

CT Findings

- Vertebral body fracture, often comminuted
  - Transversely oriented posterior element fracture
  - Posterior ligamentous injury
    - Separation of facet joints: "Naked facet" sign
    - Increased interspinous distance
    - Focal kyphosis
  - Gradual loss of pedicle definition on axial images: "Dissolving pedicle" sign
  - May have mild buckling or retropulsion of posterior cortex

MR Findings

- Fracture line low signal intensity on all sequences
- Band of edema may surround compressive aspect of fracture
  - Anterior loss of vertebral body height
  - Low signal intensity on T1WI, high signal intensity on T2WI, STIR
- ALL is not discontinuous
  - May be partially stripped from vertebral attachment
- Fractures of posterior elements are often difficult to see
  - There may be no marrow edema associated with distractive fracture in acute phase
- Discontinuity of interspinous, supraspinous ligaments
  - Seen in ligamentous and osteoligamentous patterns
- Look for spinal cord contusion and hemorrhage

Imaging Recommendations

- Best imaging tool
  - CT scan for acute evaluation, MR for more complete evaluation of discoligamentous complex
- Protocol advice
  - Coronal/sagittal reformations essential

DIFFERENTIAL DIAGNOSIS

Shear Injury

- Cause: Transverse shearing force
- All 3 columns disrupted
  - Anterior, posterior, or lateral listhesis of vertebra
- Vertebral displacement and ALL disruption distinguish from Chance fracture

Distraction Injury

- Cause: Vertical distraction or hyperextension
- All 3 columns disrupted
- Distraction &/or listhesis of vertebra
- Vertebral displacement and ALL disruption distinguish from Chance fracture

Burst Fracture

- Cause: Axial load force
- Also involves posterior vertebral body cortex
  - ± retropulsion of posterior cortex
- Vertically oriented fractures of posterior elements
  - Posterior element fractures are transverse in Chance fracture
- Normal interspinous distance
- May have posterior hematoma on MR, but interspinous ligaments are intact

Traumatic Compression Fracture

- Cause: Axial load force ± flexion
- < 40% loss of vertebral body height
- No posterior element fracture
- Normal interspinous distance

Osteoporotic Compression Fracture

- Cause: Normal weight-bearing stress or minor trauma
- May have complete loss of vertebral body height
- Pedicles may be involved
- Often at multiple levels
- Osteoporosis usually apparent on radiographs

Pathologic Vertebral Fracture

- Cause: Normal weight-bearing stress or minor trauma
- May have complete loss of vertebral body height
- Trabecular destruction on CT
- Tumor may be difficult to see at fracture site
- Often see marrow replacement at other levels on CT or MR

PATHOLOGY

General Features

- Etiology
  - Motor vehicle accident or fall
    - Anterior compression, posterior distraction around fulcrum
    - Injury patterns depend on position of fulcrum
  - Classic pattern: Lap seat belt without shoulder strap
    - Seat belt fulcrum anterior to vertebral column
    - Compression of anterior column, distraction of posterior column
  - Midthoracic Chance fractures have fulcrum in middle column
    - Posterior column tension failure
    - Anterior column compression failure
- Associated abnormalities
  - 15-80% have significant abdominal injuries (bowel and mesentery most common)
  - Retropulsion of posterior cortex has higher incidence of cord injury

Staging, Grading, & Classification

- Osseous Chance fracture
  - Vertebral body fracture
  - Posterior element fractures: Pedicles, transverse processes, laminae, spinous process
- Ligamentous Chance injury (uncommon)
  - Intervertebral disc
  - Facet dislocation
  - Ruptured interspinous ligaments
- Osteoligamentous Chance injury
  - Variable combination of fracture and ligament injury

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Traumatic back pain ± neurologic injury

Demographics

- Epidemiology
  - 5-11% of thoracolumbar fractures
  - Decreased prevalence after institution of automobile shoulder belts

Natural History & Prognosis

- Patients may develop kyphotic deformity

Treatment

- Options, risks, complications
  - Osseous injury
    - Acutely unstable
    - Often treated conservatively with bracing
    - May require fixation if reduction cannot be maintained
  - Osteoligamentous, ligamentous injury
    - Poor prognosis for healing unless fusion performed

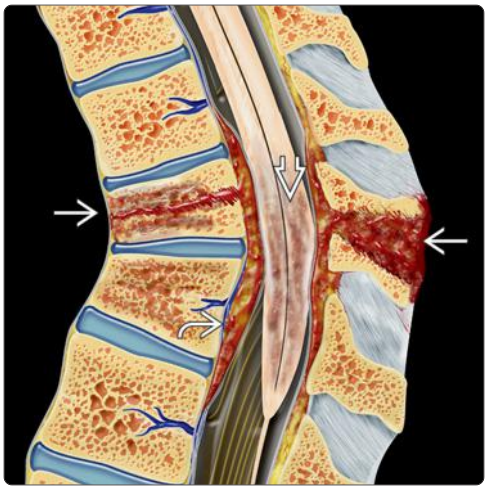
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- MR evidence of hematoma between spinous processes is not sufficient to diagnose ligament disruption
  - May also occur due to compression injury
- Chance fracture may have retropulsion of posterior vertebral body cortex mimicking burst fracture
  - Posterior element fracture is horizontal in Chance fracture, vertical in burst fracture
- Severe anterior wedging usually indicates Chance fracture in patient with normal bone density

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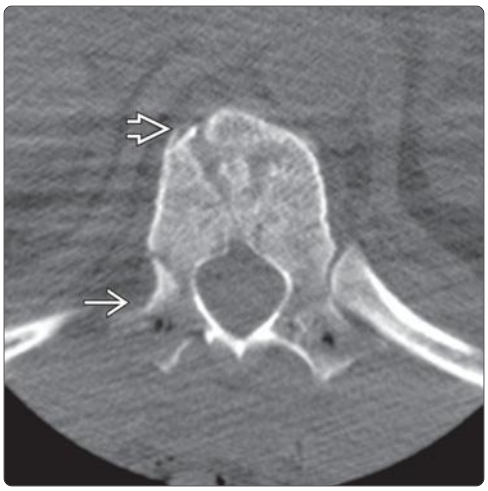


(Left) *Sagittal graphic shows bony Chance injury through vertebral body, pedicles, and spinous process. There is associated epidural hematoma and cord contusion. Bony Chance injury tends to heal more readily than the ligamentous Chance.* **(Right)** *Sagittal bone CT shows bony Chance injury with anterior compression-type fracture and transverse fracture of the posterior process.*





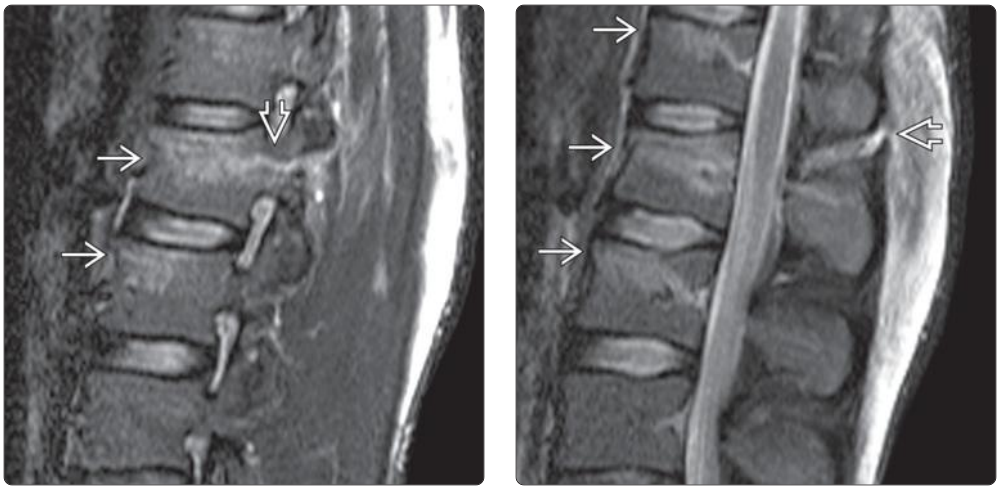
(Left) *Lateral radiograph of bony Chance injury at T11 shows anterior compression of T11 and widely distracted fracture of the lamina.* **(Right)** *Anteroposterior radiograph in the same patient shows loss of vertebral body height and horizontal fractures through right pedicle and left pars interarticularis. The T11 spinous process is displaced inferiorly.*





(Left) *Axial bone CT shows vertebral body fracture, which is usually not conspicuous on axial imaging. Right facet and pedicle are indistinct ("vanishing pedicle" sign), indicating posterior element fracture.* **(Right)** *Coronal bone CT shows horizontal fracture of posterior elements. Some patients show a bony fracture on one side and ligamentous disruption on the other.*

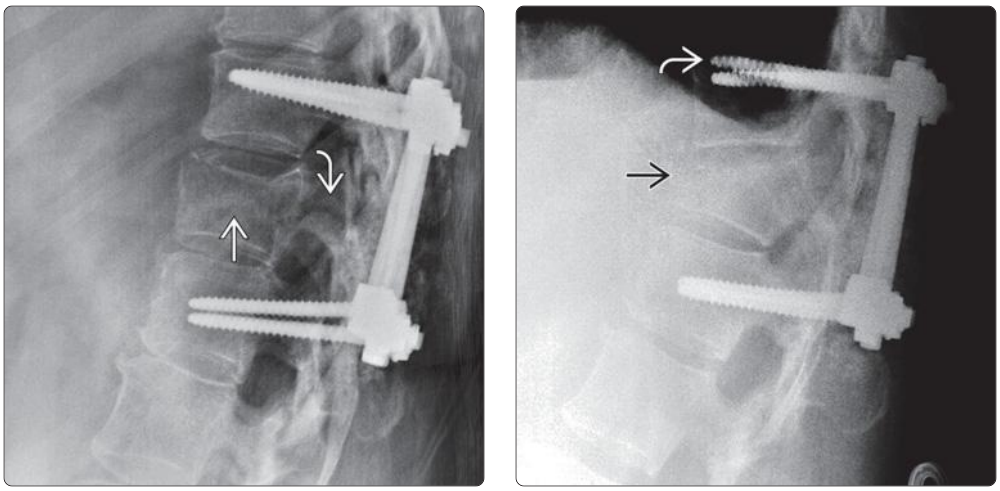
(Left) *Sagittal T2WI MR shows bone marrow edema in the anterior portions of T12 and L1 consistent with compression injury. The slightly distracted horizontal fracture through the T12 pedicle indicates a distraction force was present posteriorly.* **(Right)** *Sagittal STIR MR shows anterior compression at 3 levels. Posterior distraction is limited to the central level involved.*



(Left) *Sagittal bone CT shows Chance injury in a patient with underlying Scheuermann disease. The anterior wedging and focal kyphosis are beyond the range seen in Scheuermann disease, and the splaying of the spinous processes is another helpful sign of Chance injury.* **(Right)** *Sagittal STIR MR in the same patient confirms posterior ligament injury. Bone marrow edema is consistent with acute injury. Adjacent vertebrae show bone bruise and compression fracture.*



(Left) *Lateral radiograph shows a characteristic bony Chance injury with sclerotic vertebral body fracture line and lucent, slightly separated posterior element fracture. There is only minimal kyphosis on the 1st postoperative radiograph after ORIF.* **(Right)** *Lateral radiograph in the same patient 2 months later shows that the T12 vertebral body has lost its reduction and developed severe anterior compression. The T11 fixation screws have displaced superiorly.*





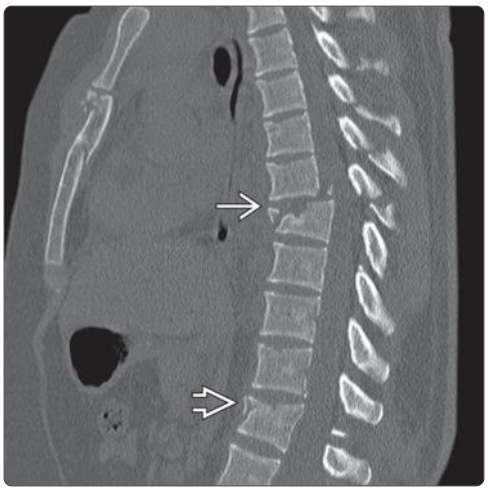


(Left) *Sagittal CT in a child shows the typical horizontal pedicle fracture of the Chance morphology. There is distraction of the posterior elements, with a compressive component anteriorly.* **(Right)** *Sagittal T2WI MR shows a horizontally directed focus of low signal within the L2 pedicle due to a Chance fracture in this child. Note there is no significant increased marrow signal in this acute fracture. Marrow edema cannot be relied upon for detection of fractures on MR.*





(Left) *Sagittal bone CT shows an upper thoracic Chance injury with anterior compression and posterior spinous process distraction. The 3 levels below this are also fractured, and posterior ligament injury should be suspected.* **(Right)** *Sagittal STIR MR in the same patient shows disruption of interspinous and supraspinous ligaments at 2 levels. ALL is partly stripped from the involved vertebral bodies but remains continuous.*





(Left) *Sagittal bone CT shows a shear injury at T7-T8 associated with a sternal fracture. In addition, there is a Chance fracture at T12. Spinal injuries often involve multiple levels, raising the danger of satisfaction of search.* **(Right)** *Sagittal STIR MR in the same patient is useful for contrasting the 2 fracture types. The shear injury shows a tear of the ALL, while the Chance fracture does not. Both injuries show disruption of the posterior ligaments.*

Thoracic and Lumbar Hyperextension Injury

KEY FACTS

IMAGING

- Widening of anterior disc space, fracture(s) of posterior elements
- Lower thoracic spine > upper thoracic or lumbar spine
- "Pure" hyperextension injury
  - Failure of anterior ± middle columns in tension
  - Widening of anterior disc space
  - ± mild retrolisthesis
  - Disruption of ALL and disc annulus on MR
  - Fracture(s) of spinous process, lamina
  - May have injury of facet joints
  - Deformity tends to spontaneously reduce, may be minimal indication of injury on plain films/CT
- Hyperextension fracture-dislocation
  - Subtype of thoracolumbar fracture-dislocation
  - Mechanism involves hyperextension with significant shear component
  - Anteriorly directed force vector dislocates vertebra anteriorly relative to supradjacent vertebra
  - Disruption of all 3 columns of spine, unstable injury
  - High rate of complete cord injury with paraplegia

TOP DIFFERENTIAL DIAGNOSES

- Thoracic and lumbar burst fracture
- Distraction fracture, low thoracic
- Fracture dislocation

CLINICAL ISSUES

- Hyperextension is rare mechanism of thoracolumbar injury

DIAGNOSTIC CHECKLIST

- Ligamentous hyperextension injury may have no signs of injury on radiographs or CT





(Left) *Sagittal NECT (bone window) shows a fracture extending through the inferior aspect of the vertebral body along the margin with the superior endplate. There is anterior widening of the fracture that is consistent with hyperextension injury.* **(Right)** *Sagittal STIR MR shows an abnormal increased signal involving the anterior longitudinal ligament, inferior vertebral body. There is no definite abnormal signal seen in the posterior longitudinal ligament or posterior elements.*





(Left) *Sagittal CT of a patient with ankylosing spondylitis shows a hyperextension fracture-dislocation injury. The spine below the injury is displaced anteriorly, resulting in severe canal compromise. A remote compression fracture is seen caudally.* **(Right)** *Sagittal CT shows a mildly distracted, oblique fracture extending through the midthoracic vertebral body, reflecting hyperextension fracture-dislocation pattern of injury. Vertical fractures are seen in the 2 lower vertebrae.*

TERMINOLOGY

Synonyms

- Extension-distraction injury
- Hyperextension-dislocation injury
- Lumbarjack paraplegia

IMAGING

General Features

- Best diagnostic clue
  - Widening of anterior disc space, fracture(s) of posterior elements
- Location
  - Lower thoracic spine > upper thoracic or lumbar spine
- "Pure" hyperextension injury
  - Failure of anterior ± middle columns in tension
    - Widening of anterior disc space
    - ± mild retrolisthesis
    - Disruption of ALL and disc annulus on MR
    - Rare reported complication: Entrapment of bowel or esophagus within intervertebral disc space
  - Fracture(s) of spinous process, lamina
    - Especially with mechanism involving posterior impact
    - May be present at multiple adjacent levels
  - May have injury of facet joints
    - Fracture
    - Widening of joint spaces
  - Deformity tends to spontaneously reduce, may be minimal indication of injury on plain films/CT
    - High index of suspicion necessary to identify potentially unstable spinal injury
- Hyperextension fracture-dislocation
  - Subtype of thoracolumbar fracture-dislocation
  - Mechanism involves hyperextension with significant shear component
  - Anteriorly directed force vector dislocates vertebra anteriorly relative to supradjacent vertebra
    - Complete disruption of discovertebral unit &/or fracture through vertebral body
    - Fracture(s) of neural arch(s)
      - May be present at multiple adjacent levels
  - Disruption of all 3 columns of spine, unstable injury
  - High rate of complete cord injury with paraplegia
    - Lumberjack paraplegia due to high prevalence of timber harvesters (50%) in 1 reported series of thoracolumbar hyperextension injuries

MR Findings

- T2WI/STIR
  - Edema with soft tissue/ligamentous injury
  - ↑ signal, disc space widening indicates disruption of intervertebral disc
  - Disruption of ALL, ± PLL
  - ↑ signal in spinal cord indicates cord contusion
    - Discontinuity of spinal cord indicates cord transection

Imaging Recommendations

- Best imaging tool
  - Thin-slice helical CT with sagittal and coronal reformatted images best for fracture
  - MR to evaluate cord injury with neurologic deficit or to evaluate radiographically occult ligamentous injury
- Protocol advice
  - MR should include sagittal STIR/T2WI, axial T2* GRE or T2WI, sagittal and axial T1WI

DIFFERENTIAL DIAGNOSIS

Thoracic and Lumbar Burst Fracture

- Structural failure due to axial loading
- Comminuted fracture of vertebral body involving posterior cortex

Distraction Fracture, Low Thoracic

- Mechanism involves forced flexion around anterior fulcrum, rather than hyperextension
- Widening of interspinous distance, anterior column relatively spared

Fracture Dislocation

- Disruption of vertebral column due to shearing force
- Hyperextension is infrequent mechanism of thoracolumbar fracture-dislocation

PATHOLOGY

Staging, Grading, & Classification

- Infrequent type of injury, omitted from many spinal classification systems
- Hyperextension fracture-dislocation is Magerl type B3

CLINICAL ISSUES

Demographics

- Epidemiology
  - Hyperextension is rare mechanism of thoracolumbar injury

Natural History & Prognosis

- High rate of paraplegia with fracture-dislocation

Treatment

- Immobilization, bracing
- Surgical fusion, especially for purely ligamentous injury or for paraplegia

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Ligamentous hyperextension injury may show no signs of injury on radiographs and CT

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Anterior Compression Fracture

KEY FACTS

TERMINOLOGY

- Vertebral body fracture compressing anterior cortex, sparing middle/posterior columns

IMAGING

- Vertebral body shorter anteriorly than posteriorly
  - < 40-50% loss of height in patients with normal bone density
- ± vertebral body endplate abnormality
- ± anterior cortical irregularity
- Normal middle and posterior vertebral columns
- Most common in middle and lower thoracic spine

TOP DIFFERENTIAL DIAGNOSES

- Compression-distraction injury (Chance fracture)
- Burst fracture
- Pathologic fracture due to tumor
- Schmorl node
- Scheuermann kyphosis
- Physiologic vertebral wedging
- Limbus vertebra

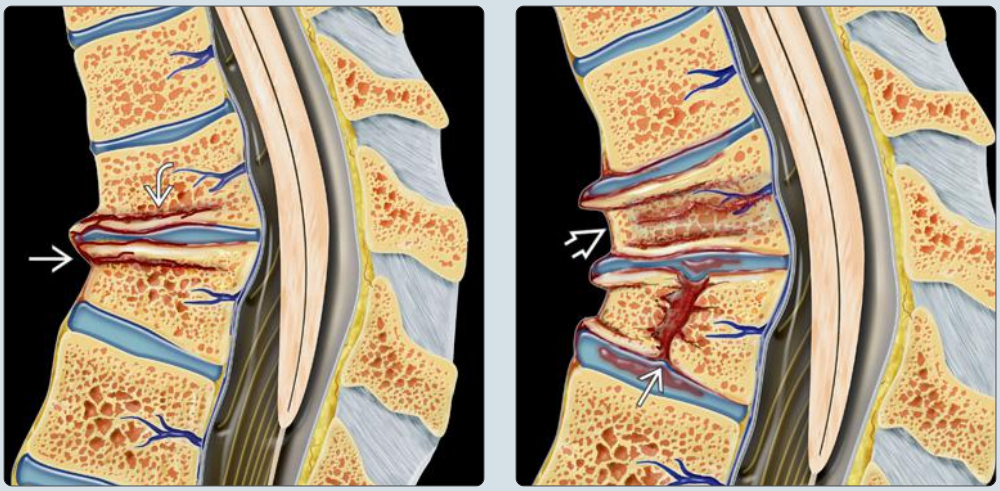
CLINICAL ISSUES

- Most common type of thoracic spine fracture due to blunt trauma
  - Young patients: Due to significant fall
  - Osteoporotic patients: Insufficiency fracture
- American Academy of Orthopaedic Surgeons (AAOS) practice guidelines (2011)
  - Against vertebroplasty for osteoporotic spinal compression fracture in patients who are neurologically intact
- AAOS guidelines recommend calcitonin for 4 weeks
  - Ibandronate and strontium ranelate are options to prevent additional symptomatic fractures

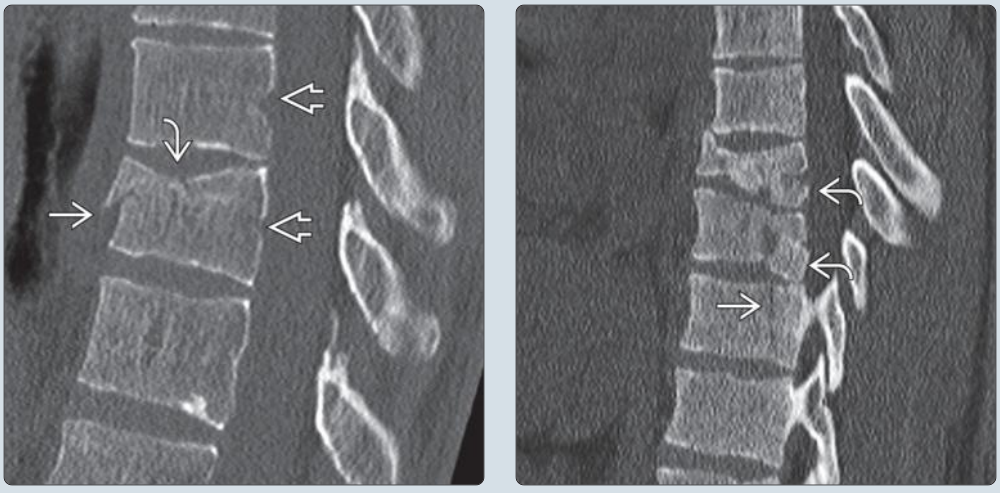
DIAGNOSTIC CHECKLIST

- Patients often have additional compression, burst, Chance, or shear fractures at other spinal levels

(Left) *Sagittal graphic shows 2 types of compression fractures. The most common type is compression of the superior endplate. Isolated compression of the inferior endplate is rare. Note that in these cases, there is angular deformity of the anterior cortex without focal endplate angulation.* **(Right)** *Sagittal graphic shows 2 additional types of compression fractures. The fracture involving both endplates is common, while the coronally oriented fracture through the vertebral body with wedge deformity is rare.*



(Left) *Sagittal bone CT shows a compression fracture causing angular deformity of the vertebral endplate and cortical step-off anteriorly. The normal vascular groove posteriorly should not be confused with a fracture.* **(Right)** *Sagittal bone CT shows multilevel injuries. The T12 compression fracture has a coronal split. T10 and T11 fractures are not compression fractures, since there is retropulsion of the posterior vertebral body cortex.*



TERMINOLOGY

Synonyms

- Wedge compression

Definitions

- Vertebral body fracture compressing anterior cortex, sparing middle/posterior columns

IMAGING

General Features

- Best diagnostic clue
  - Wedge-shaped vertebral body
- Location
  - May occur at multiple levels, contiguous or noncontiguous
  - Most common in middle and lower thoracic spine

Radiographic Findings

- Paraspinous hematoma apparent on AP radiograph
- Wedge-shaped vertebral body
  - Vertebral body shorter anteriorly than posteriorly
  - < 40-50% loss of height in patients with normal bone density
    - If greater loss of height, probably Chance fracture
- Vertebral body endplate abnormality
  - Most commonly superior endplate alone
  - May involve both endplates
  - < 5% involve inferior endplate only
- Variable contours of affected vertebral endplate
  - Focal, angular deformity
  - Diagonal orientation
  - Rounded concavity (usually with osteoporotic compression fracture)
- Rarely, coronal fracture through vertebral body
- Anterior vertebral body cortex shows angular deformity or step-off
- Posterior vertebral body cortex intact
- Normal middle and posterior vertebral columns
- In osteoporotic patients, may develop vertebra plana
- Upper thoracic vertebrae less commonly involved
  - Difficult to see on radiographs
  - Swimmer's view can be helpful

CT Findings

- Fracture best seen on coronal and sagittal reformatted images
- Horizontal, sclerotic fracture line
  - Due to trabecular impaction
  - Does not extend to posterior cortex of vertebral body
  - Often comminuted
- Deformity of endplate &/or anterior vertebral body cortex
  - Angulation or displacement (cortical step-off)
- Fractures of posterior elements absent
- Alignment of posterior elements normal on reformatted images
- Many trauma centers now go routinely to CT scan without radiographs

MR Findings

- Fracture line has low signal intensity on all sequences
  - May be obscured by marrow edema on STIR
  - Morphology as described for CT appearance
- Band-like edema seen surrounding fracture line
  - Low to intermediate signal on T1WI
  - High signal on T2WI, STIR
- Paraspinous hematoma can mimic tumor
- Fracture, bone marrow edema, and paraspinous hematoma enhances with gadolinium contrast

Nuclear Medicine Findings

- Bone scan
  - Positive in all 3 phases if acute
  - Indistinguishable from infection, tumor, Charcot arthropathy, degenerative instability

Imaging Recommendations

- Best imaging tool
  - CT best to differentiate from Chance and burst fractures
- Protocol advice
  - Multidetector CT with thin, overlapping helical sections
  - Sagittal/coronal reformations essential to detect signs of ligament injuries

DIFFERENTIAL DIAGNOSIS

Compression-Distraction Injury (Chance Fracture)

- Anterior compression + middle and posterior column distraction
- Horizontally oriented posterior element fractures **or**
- Disruption of facet joints and interspinous ligaments
- MR useful to confirm posterior column ligamentous injury

Burst Fracture

- Same injury mechanism (axial load) as compression fracture
- Involves posterior vertebral body cortex
- ± retropulsion of posterior vertebral cortex
- Less common in thoracic spine because of stabilizing effect of ribs

Pathologic Fracture Due to Tumor

- Cortical destruction
- Trabecular destruction, rounded mass on CT
- Rounded or diffuse bone marrow edema on MR
- Metastases often involve posterior elements as well as vertebral body
- Paraspinous mass may be due to hematoma (benign compression fracture) or tumor extension
- Often see additional foci of tumor at levels away from fracture
- Diffusion, in- and out-of-phase imaging of questionable usefulness in differential diagnosis

Schmorl Node

- Rounded depression of vertebral endplate
- Usually smoothly marginated
- Fracture may develop through Schmorl node
- MR may show bone marrow edema due to fracture or discogenic edema

Scheuermann Kyphosis

- 3 contiguous levels or vertebral body wedging > 5°
- Schmorl nodes
- Undulation of vertebral endplates

Physiologic Vertebral Wedging

- Involves T11, T12, &/or L1
- Loss of height is mild
- Usually involves both superior, inferior endplates
- No angular contour abnormality

Limbus Vertebra

- Failure of fusion of ring apophysis
- Creates a small, triangular ossicle at anterior corner of vertebral body
- Corticated margins distinguish from acute fracture

Old, Healed Fracture

- Absence of paraspinous hematoma, MR bone marrow edema useful to distinguish acute/subacute fracture from chronic deformity

PATHOLOGY

General Features

- Etiology
  - Axial load ± flexion component
  - Because of normal thoracic kyphosis, axial load affects anterior portion of vertebral body more than posterior
- Associated abnormalities
  - Other spine fractures, contiguous or not contiguous
  - Fractures of pelvis + lower extremities

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute trauma with focal back pain
  - Sudden onset back pain with no or minimal trauma in elderly patients
  - Other signs/symptoms
    - Radiculopathy
    - Kyphotic deformity

Demographics

- Epidemiology
  - Most common type of thoracic spine fracture due to blunt trauma
  - 2 distinct populations: Major trauma vs. insufficiency fracture
  - Young patients: Due to significant fall
  - Osteoporotic patients: Insufficiency fracture

Natural History & Prognosis

- Patients with normal bone mineral density heal well with conservative management
- Increased risk of premature degenerative disc disease in young patients
- Osteoporotic patients may have progressive fracture causing chronic pain
- Kyphotic deformity often progressive in osteoporotic patients
- Patients with 1 osteoporotic compression fracture at increased risk for other fractures
- May have delayed onset of neurologic symptoms

Treatment

- Conservative management usually successful
- Vertebroplasty and kyphoplasty used for chronic pain, kyphotic deformity
  - Impact on outcomes is controversial
  - American Academy of Orthopaedic Surgeons (AAOS) practice guidelines (2011)
    - Against vertebroplasty for osteoporotic spinal compression fracture on imaging in patients who are neurologically intact
      - Strength of recommendation is strong
- Bisphosphonates, calcitonin decrease pain and risk of further osteoporotic fracture
  - AAOS guidelines recommend calcitonin for 4 weeks
    - Ibandronate and strontium ranelate are options to prevent additional symptomatic fractures
  - Long-term bisphosphonate use is associated with femoral fractures

DIAGNOSTIC CHECKLIST

Consider

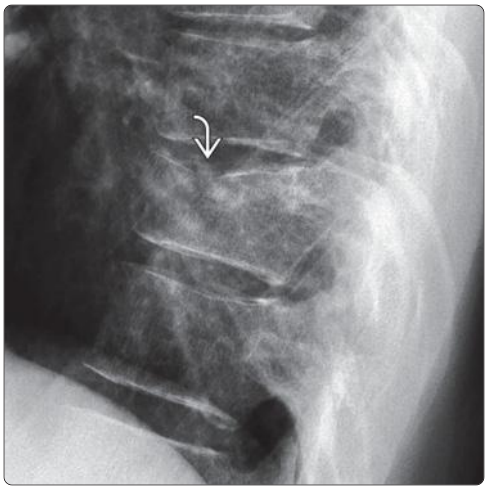
- Patients often have additional compression, burst, Chance, or shear fractures at other spinal levels

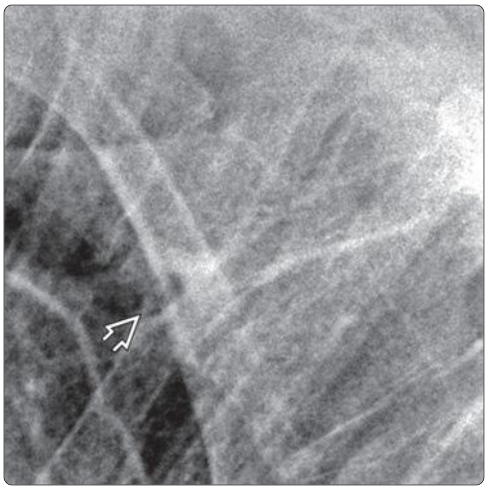
Image Interpretation Pearls

- Compression fractures involving inferior endplate with a normal superior endplate are more likely due to pathologic fracture
- Paraspinous soft tissue widening on trauma AP chest radiograph is often initial imaging sign of thoracic fracture
- Increasing thoracic kyphosis on lateral chest radiograph is often initial imaging sign of osteoporotic compression fracture
- If middle or posterior column abnormal, it is not compression fracture

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(Left) *Lateral radiograph shows angular contour deformity in the superior endplate of T10. There is only mild loss of vertebral body height anteriorly.* **(Right)** *Lateral radiograph shows a T3 compression fracture. Fractures at this level are difficult to see on radiographs but should be suspected when there is increased upper thoracic kyphosis.*





(Left) *Axial bone CT shows a compression fracture traversing the anterior vertebral body. Paraspinous hematoma is a useful secondary sign of injury. Sagittal reformatted images are very useful to distinguish between compression and burst fractures.* **(Right)** *Sagittal bone CT shows a compression fracture with minimal loss of vertebral height. The sclerotic band represents trabecular impaction.*





(Left) *Sagittal T1WI MR shows cup-shaped depression of the superior vertebral cortex in an osteoporotic patient. The low signal intensity fracture line is immediately beneath the cortex. Osteoporotic patients often describe minimal or no trauma but acute onset of back pain.* **(Right)** *Sagittal STIR MR shows a T12 burst fracture compared to T10, T11 compressions. The posterior cortex is not involved with the compression fractures.*

Lateral Compression Fracture

KEY FACTS

IMAGING

- Most common at thoracolumbar junction
  - Also seen in midthoracic spine
- Loss of lateral vertebral body height
  - Involves 1 side of vertebra more than other side
  - Posterior vertebral body cortex not involved
- Focal, short-curve scoliosis
- Focal kyphosis
- Fracture fragments centered on lateral vertebral body cortex

TOP DIFFERENTIAL DIAGNOSES

- Anterior compression fracture
- Burst fracture
- Pathologic fracture
- Hemivertebra
- Schmorl node
- Scheuermann disease

PATHOLOGY

- Typically result of flexion, axial forces, or both
- Shoulder-lap seat belt injury
- Typically result of flexion, axial forces, or both
- Associated with transverse process fracture

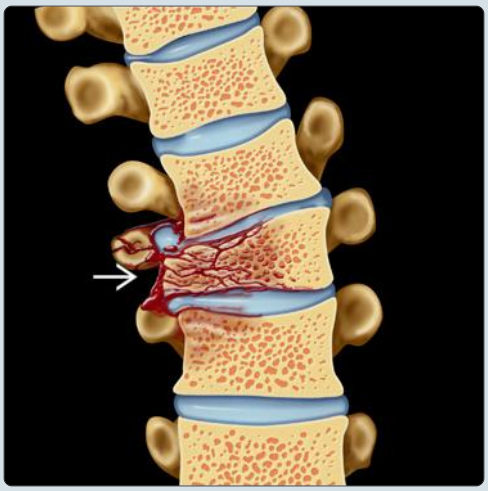
CLINICAL ISSUES

- 2 distinct populations: Major trauma vs. insufficiency fracture

DIAGNOSTIC CHECKLIST

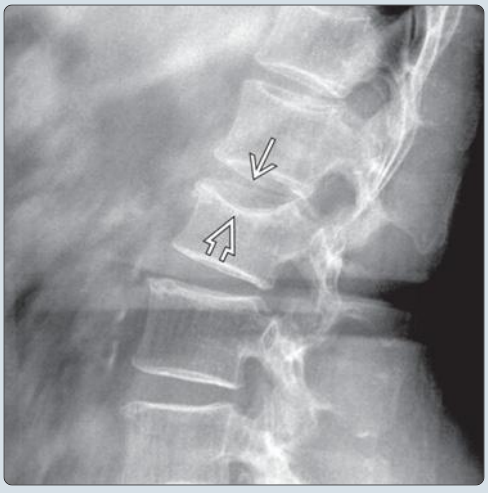
- Cause of painful, short-curve scoliosis
- Up to 20% of vertebral fractures are multiple
- May be clinically unsuspected in multitrauma patients, detected on screening CT scan

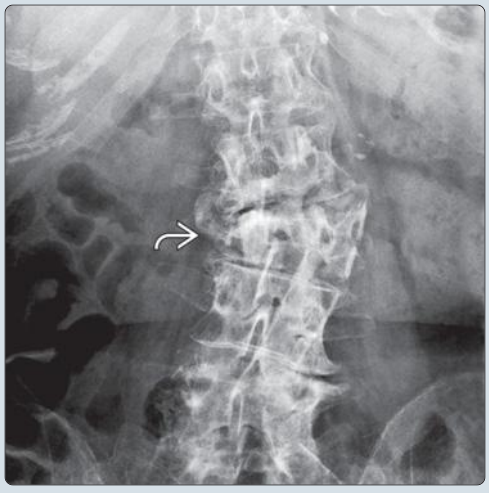
(Left) *Coronal graphic illustrates a right lateral thoracic compression fracture and transverse process fracture. There are also minimal fractures of the endplates of the adjacent vertebra.* **(Right)** *Anteroposterior radiograph demonstrates a left lateral compression fracture of L2.*





(Left) *Lateral radiograph shows a L2 lateral compression fracture. The lateral nature is evident by the appearance of the normal superior endplate and the abnormal superior endplate.* **(Right)** *Anteroposterior radiograph shows levocurvature of the lumbar spine associated with laterally wedged osteoporotic compression fracture of L3.*





IMAGING

General Features

- Best diagnostic clue
  - Asymmetric loss of vertebral body height
- Location
  - Most common at thoracolumbar junction
  - Also seen in midthoracic spine

Radiographic Findings

- Loss of lateral vertebral body height
  - Involves 1 side of vertebra more than other side
  - Posterior vertebral body cortex not involved
- Focal, short-curve scoliosis
  - Usually angular in morphology
- Focal kyphosis

CT Findings

- Fracture fragments centered on lateral vertebral body cortex
  - Often multiple small fragments
  - Intact posterior vertebral body cortex, posterior elements
- Angular deformity of lateral vertebral cortex
- Sclerotic band due to trabecular impaction
- Paraspinous hematoma

MR Findings

- Fracture lines have low signal intensity on all sequences
  - Intact posterior vertebral body cortex, posterior elements
- Band-like T2 marrow edema centered on fracture
- Angular deformity of lateral vertebral cortex
- Paraspinous hematoma
- May have soft tissue hematoma posteriorly, but posterior ligaments are intact

Imaging Recommendations

- Best imaging tool
  - CT more sensitive than radiographs
    - Coronal, sagittal reformations essential

DIFFERENTIAL DIAGNOSIS

Compression Fracture

- Anterior without lateral component

Burst Fracture

- Fracture extends into posterior vertebral body cortex
- ± fragment retropulsion into spinal canal

Pathologic Fracture

- FDG PET may be positive
- Often see rounded morphology of marrow abnormality
  - In contrast to band-like morphology of traumatic fracture

Hemivertebra

- Vertebra may be hypoplastic on 1 side
- Smooth contour
- Normal MR signal intensity

Schmorl Node

- Disc material invaginates through vertebral endplate
- May be asymmetric

Scheuermann Disease

- 3 contiguous levels of vertebral body wedging > 5°
- Undulation of vertebral endplates, Schmorl nodes
- 15% have scoliosis

PATHOLOGY

General Features

- Etiology
  - Typically result of flexion, axial forces, or both
  - Shoulder-lap seat belt injury
    - Lateral compression occurs on side opposite of restrained shoulder
    - Flexion and rotation about axis of shoulder strap
    - Results in anterolateral wedge compression of thoracolumbar vertebra

CLINICAL ISSUES

Demographics

- Age
  - Young patients: Motor vehicle accident or fall
  - Elderly patients: Minor injury in setting of osteoporosis
- Epidemiology
  - 11% of vertebral compression fractures are lateral
  - Most commonly due to osteoporosis

Treatment

- Conservative therapy
  - Consists of analgesic use, rest, and external bracing
- Vertebroplasty or kyphoplasty
- Surgery when neurologic dysfunction &/or instability occurs as result of VCF

DIAGNOSTIC CHECKLIST

Consider

- Cause of painful, short-curve scoliosis
- May be clinically unsuspected in multitrauma patients, detected on screening CT scan

Image Interpretation Pearls

- Up to 20% of vertebral fractures are multiple

SELECTED REFERENCES

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Lumbar Facet-Posterior Fracture

KEY FACTS

TERMINOLOGY

- Fracture of lumbar facet or neural arch (excluding pedicle and pars interarticularis)

IMAGING

- Trauma
  - Facet fracture &/or dislocation as component of fracture-dislocation of lumbar spine
  - Superior S1 facet fracture associated with Denis zone II sacral fracture (20%)
  - Transverse process fracture(s) due to direct blow to lumbar region
  - Burst fracture may be associated with fracture of lamina
- Stress (fatigue) fracture of articular facet, usually in competitive athletes, gymnasts
- Complication of lumbar surgery: Inferior articular facet fracture is one cause of failed back surgery syndrome
- Facet fracture usually occult on radiographs, unless oblique films obtained
- Thin-slice helical CT with sagittal and coronal reformatted images best tool for imaging posterior elements of lumbar spine

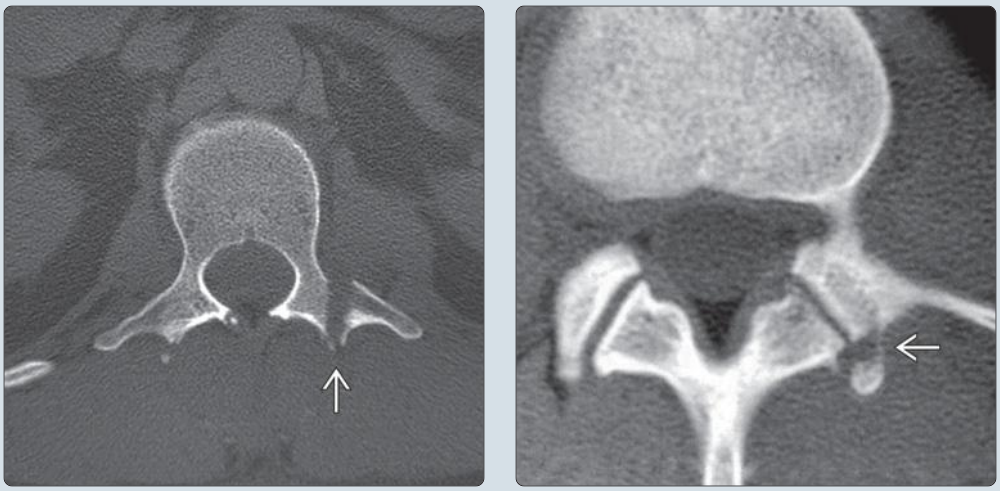
TOP DIFFERENTIAL DIAGNOSES

- Pars interarticularis fracture
- Hypoplastic or accessory ribs at T12, L1

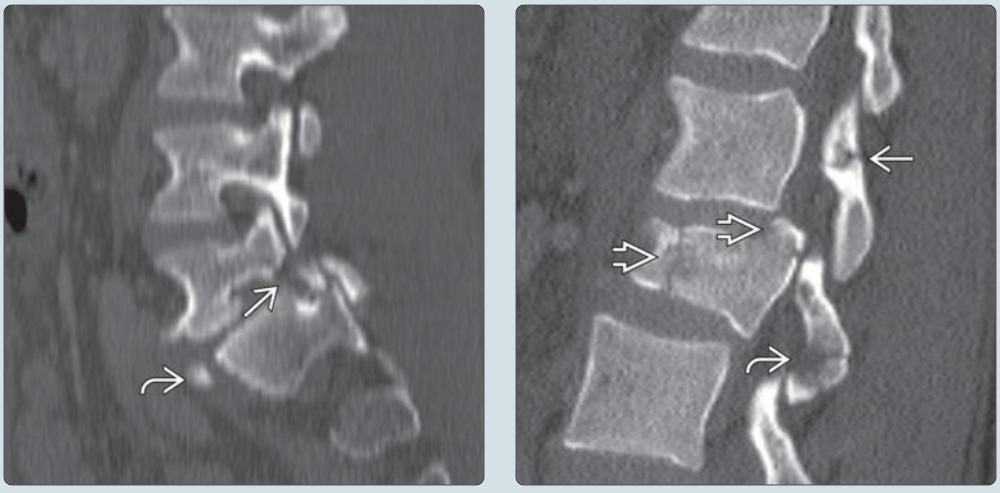
CLINICAL ISSUES

- Treatment
  - Conservative therapy → bracing and pain control
  - Fusion for instability, persistent pain

(Left) *Axial NECT (bone window) shows a linear fracture through the left L1 transverse process, with slight displacement.* **(Right)** *Axial NECT shows an L3 superior facet fracture in a young hockey player who experienced sudden onset of low back pain while playing. (Courtesy W.O. Shaffer, MD.)*



(Left) *Sagittal NECT (bone window) shows a traumatic pars fracture. There is an avulsion of the anterior longitudinal ligament indicating acute anterior column disruption.* **(Right)** *Sagittal reconstructed CT of the lumbar spine shows a L2 burst fracture with anterior wedging, a nondisplaced fracture of the inferior L2 articular process, and a nondisplaced fracture of the L1 lamina.*



TERMINOLOGY

Definitions

- Fracture of lumbar facet or neural arch (excluding pedicle and pars interarticularis)

IMAGING

General Features

- Best diagnostic clue
  - Fracture line through lumbar articular process, transverse process, lamina or spinous process
- Morphology
  - Trauma
    - Facet fracture &/or dislocation as component of fracture-dislocation of lumbar spine
      - L5-S1 most common, with superior S1 facet fracture
    - Superior S1 facet fracture associated with Denis zone II sacral fracture (20%)
    - Transverse process fracture(s) due to direct blow to lumbar region
    - Burst fracture may be associated with fracture of lamina
  - Stress fracture of articular facet, usually in competitive athletes, gymnasts
  - Complication of surgery: Inferior articular facet fracture is one cause of failed back surgery syndrome
    - Following laminectomy or placement of prosthetic intervertebral disc

Radiographic Findings

- Radiography
  - Facet fracture usually occult on radiographs, unless oblique films obtained

CT Findings

- Linear defect through facet, transverse process, lamina, or spinous process
- ± bony sclerosis with fatigue fracture of facet

MR Findings

- STIR
  - Marrow edema; surrounding soft tissue edema, hemorrhage
  - Facet fracture
    - Increased fluid within and around facet joint
    - Disruption of zygapophyseal capsuloligamentous complex

Nuclear Medicine Findings

- Bone scan
  - Increased uptake at fracture site
    - With traumatic fracture, tracer avidity may not appear for several days until healing begins
  - Facet fracture can be difficult to see due to small size
    - When seen, can be difficult to differentiate from spondylolysis (more common); SPECT helpful

Imaging Recommendations

- Best imaging tool
  - Thin-slice helical CT with sagittal and coronal reformatted images

DIFFERENTIAL DIAGNOSIS

Pars Interarticularis Fracture (Spondylolysis)

- More common cause of low back pain in young athletes

Hypoplastic or Accessory Ribs

- Small ribs at T12 or L1 articulating with vertebral body; normal variant, not fracture

PATHOLOGY

General Features

- Etiology
  - Trauma
    - High-energy mechanisms, such as MVA, falls
      - 35% incidence of facet fracture in one series of MVA fatalities
    - Lumbosacral dislocation: Shear force ± distraction
    - Fractures of transverse processes, spinous process: Direct blow to lumbar area
    - Burst fracture: Direct axial loading of vertebral body without flexion
  - Sports injury
    - Stress (fatigue) fracture in competitive athletes
    - Acute fracture: Rotated waist causing loading of facet joint, then posterior/posterolateral blow to body (e.g., "checking" in hockey)
  - Laminectomy defect encroaching onto medial margin of inferior articular process; structural weakness predisposing to fracture
- Associated abnormalities
  - Retroperitoneal injury associated with transverse process fracture(s)

CLINICAL ISSUES

Presentation

- Clinical profile
  - Athlete: Acute or acute-on-chronic pain: Exacerbated by movement, relieved with recumbency
  - Trauma victim with low back pain, radiculopathy, &/or cauda equina syndrome

Natural History & Prognosis

- Nonunion of isolated lumbar facet fracture in 5/5 patients in one series, despite resolution of symptoms

Treatment

- Conservative therapy → bracing and pain control
- Fusion for instability, persistent pain

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Sacral Traumatic Fracture

KEY FACTS

IMAGING

- Difficult to see on routine radiography
  - Up to 60% reportedly missed prior to CT
  - Disruption of arcuate line of neural foramina indicates zone II fracture
  - Lateral compression fracture causes decreased width of sacrum on affected side
  - Transverse fracture line usually causes angular deformity on lateral radiograph
- CT shows displacement not apparent on radiography
  - CT defines position of fracture relative to neural foramina, spinal canal
- Fracture difficult to see on MR
  - MR useful to evaluate for nerve root injury
  - Oblique coronal T1WI, STIR along plane of sacrum useful

TOP DIFFERENTIAL DIAGNOSES

- Stress fracture
- Tumor
- Accessory centers of ossification
- Normal variant coccyx

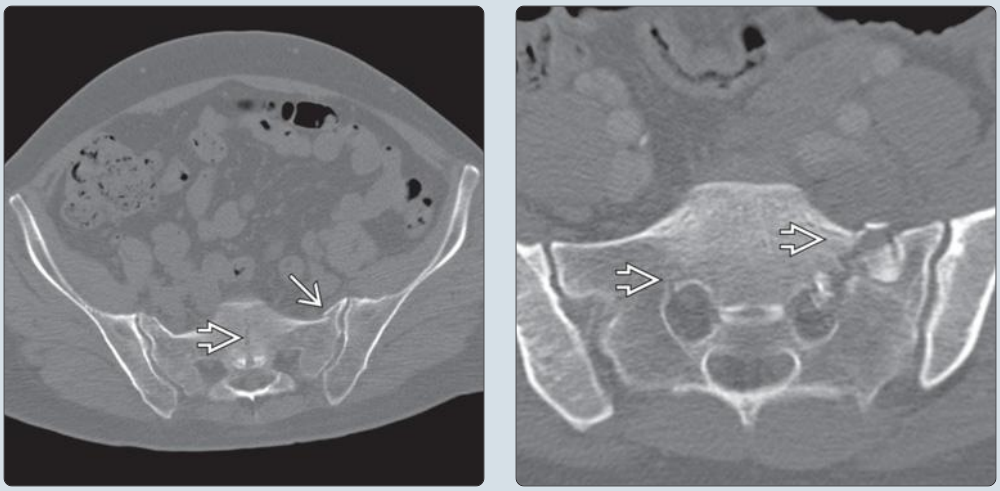
PATHOLOGY

- Associated with lumbar spine and pelvic fractures and injuries to nerves, vessels, and urinary tract
- **Denis classification (by zone)**
  - Zone I: Lateral to neural foramina
  - Zone II: Through neural foramina
  - Zone III: Through spinal canal

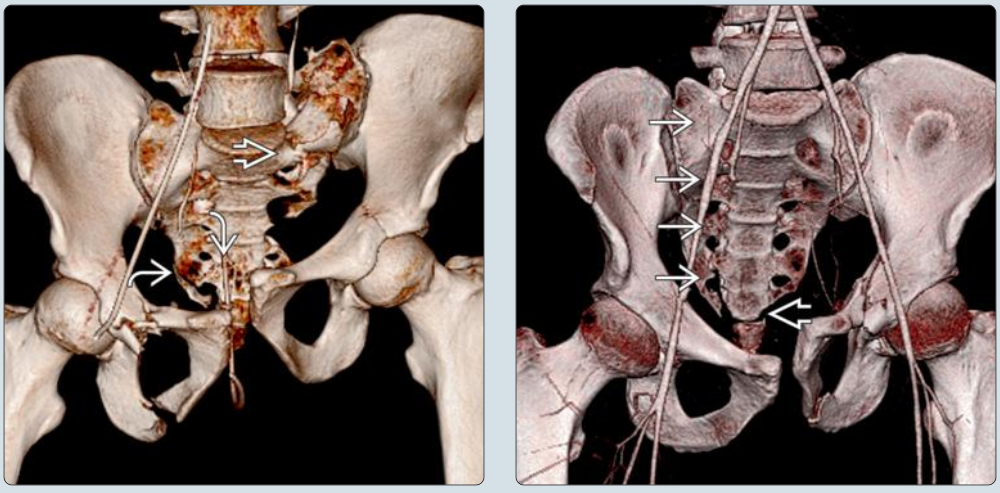
DIAGNOSTIC CHECKLIST

- Look for associated pelvic ring disruption in high-velocity injuries

(Left) *Axial bone CT shows a left sacral alar fracture ⇨ lateral to the neural foramen (Denis zone II) and a central vertical fracture extending into the spinal canal ⇨ (Denis zone III). A zone III fracture may be oriented vertically, as in this case, or horizontally across all 3 zones.* **(Right)** *Axial bone CT shows bilateral sacral fractures ⇨ extending into the neural foramina (zone II) at the S1 level.*



(Left) *Frontal 3D reformation CT shows a vertical shear injury. There is a vertical zone II fracture ⇨ on the left and a transverse zone III fracture ⇨ at the S4 level. Note associated anterior pelvic ring fractures and vertical displacement of the pubic symphysis.* **(Right)** *Frontal 3D reformation CT shows a right-sided zone II fracture ⇨. The normal sacrococcygeal junction ⇨ should not be mistaken for fracture. Anterior pelvic fractures and pubic symphysis disruption are also present.*



TERMINOLOGY

Definitions

- Pelvic ring: Pelvic bones form obliquely horizontal, ring-shaped structure around pelvic organs
- Pelvic ring disruption: Fractures &/or ligament injuries through both anterior and posterior portions of pelvis
  - Posterior ring fracture: Behind ischial spine; usually sacrum or sacroiliac joint
  - Anterior ring fracture: Anterior to ischial spine, usually pubic rami

IMAGING

General Features

- Best diagnostic clue
  - Disruption of neural foramen contour (arcuate line)
- Morphology
  - 95% vertical or oblique
  - 5% horizontal
  - U-shaped or H-shaped fractures: Horizontal + transverse components

Radiographic Findings

- Difficult to see on routine radiography
  - Up to 60% reportedly missed prior to institution of CT scanning
- Disruption of arcuate line of neural foramina indicates zone II fractures
  - Arcuate line normally forms smooth curve above neural foramen
  - Angular contour of arcuate line indicates fracture
- Lateral compression causes decreased width of sacrum on affected side
- Displacement of fracture line uncommonly visible
- Transverse fracture line is best seen on lateral radiograph
  - Angular deformity of sacrum
- Additional views improve visibility
  - Inlet view of pelvis: 25° caudad angulation
  - Outlet view of pelvis: 25° cephalad angulation
  - Ferguson view sacrum: 15° cephalad angulation
- Children may have "greenstick" fracture of sacrum

CT Findings

- Defines position of fracture relative to neural foramina, spinal canal
- Shows displacement not apparent on radiography
- Most accurate method to assess fracture displacement
  - Axial images: AP displacement, horizontal displacement, canal occlusion
  - Coronal reconstructions: Vertical translation
  - Sagittal reconstructions: AP translation, angulation

MR Findings

- Because sacrum is thin and obliquely positioned, fracture may be missed or mistaken for tumor
  - Oblique coronal T1WI, STIR along plane of sacrum useful
- Evaluate for nerve root injury
  - Traumatic avulsion may be masked acutely by surrounding hematoma
  - Evaluate injury of sacral plexus on STIR sequences

Imaging Recommendations

- Best imaging tool
  - CT scan
- Protocol advice
  - Often useful to angle reformatted images along plane of sacrum

DIFFERENTIAL DIAGNOSIS

Sacroiliac Joint Disruption/Dislocation

- May require CT to differentiate from fracture

Stress Fracture

- Back pain of insidious onset and disabling severity
- No history of trauma
- Insufficiency fracture due to osteoporosis
- Fatigue fracture in athletes
- Vertically oriented fracture through sacral ala
  - Unilateral or bilateral
  - Horizontal component connecting 2 vertical fractures in < 50%
    - Known as "Honda" sign or "H" sign
- Vacuum phenomenon often seen in anterior portion of fracture

Tumor

- Loss of arcuate lines on radiography
- Round or oval region of trabecular destruction on CT
- Round or oval region of high T2WI signal on MR
- Hematoma from traumatic fracture may mimic soft tissue mass

Accessory Centers of Ossification

- Seen at lateral margins of sacrum in children and adolescents
- Rounded contour, corticated
- Usually bilaterally symmetric

Normal Variant Coccyx

- Sacrococcygeal junction may have angular contour

Traumatic Lumbosacral Dislocation

- Due to high-speed injury, motor vehicle accident, or fall
- Complete disruption of lumbosacral ligaments
- Extremely rare
- See lateral, anterior, or posterior displacement of L5 relative to sacrum
- Diagnosed with lateral radiograph or CT

PATHOLOGY

General Features

- Etiology
  - Sacral fractures without pelvic ring disruption are usually due to falls
    - Transverse fracture may occur if patient lands in seated or supine position
    - Fractures at sacrococcygeal junction due to minor falls
  - 3 major mechanisms of pelvic ring disruption
    - Anteroposterior compression ("open book")
      - Vertically oriented sacral fracture, often slightly diastatic
    - Lateral compression ("T-bone injury")

- Vertically oriented fracture
- Impaction may cause sclerotic line &/or decreased mediolateral dimension of sacral ala

– Vertical shear (fall from height)
- Vertical displacement may be present, may have spontaneously reduced
- ± fractures of L5 transverse processes

- Associated abnormalities
  - 95% occur in association with pelvic ring fractures
  - Lumbosacral junction injury
    – Seen in up to 1/3 of sacral fractures with pelvic ring involvement
    – Facet joint injury or L5-S1 disc injury
  - Fracture of L5 transverse process
    – Due to avulsion of lumbosacral ligament
    – Highly suggestive of unstable fracture
  - Lumbar spine fractures
    – Spine fracture with neurologic deficit may cause sacral fracture to be overlooked clinically
  - Neurologic injuries
    – 40% of displaced fractures
    – Nerve roots or sacral plexus
    – Transection or stretch injury
    – More commonly seen with longitudinal sacral displacement
  - Vascular injuries
    – Therapeutic embolization may be needed
    – Active bleeding often seen on contrast-enhanced CT at time of trauma
  - Lower extremity fractures
  - Degloving injury of posterior soft tissues
- Neurologic injury may be masked by concurrent injuries at higher levels

Staging, Grading, & Classification

- **Denis classification (by zone)**
  - Zone I: Lateral to neural foramina
    – 50% of sacral fractures
    – Vertical or oblique orientation
    – Neurologic deficit in 6-24%
    – L5 nerve root may be entrapped in fracture
  - Zone II: Through neural foramina
    – 34% of sacral fractures
    – Vertical or oblique
    – Neurologic deficit in 28%
    – Traumatic "far-out syndrome"
      - L5 root caught between sacral alar fragment and transverse process
  - Zone III: Through spinal canal
    – 16% of sacral fractures
    – May be transverse or vertical
    – Transverse zone III fracture
      - Also extends through zones I and II
      - 35% have associated avulsion of nerve roots
      - "Sacral burst fracture": Transverse fracture with retropulsion of bone into spinal canal
      - Fracture-dislocation may occur through rudimentary disc at S1-S2
      - Neurologic deficit in 57-60%
    – Vertical zone III fracture
      - Majority have neurologic deficit
      - Bowel, bladder, sexual dysfunction
    – Gibbons grading system for neurologic deficits
      - Grade 1: No neurologic deficit
      - Grade 2: Sensory changes only
      - Grade 3: Motor weakness
      - Grade 4: Loss of bowel/bladder control

CLINICAL ISSUES

Natural History & Prognosis

- Unstable injuries require open reduction internal fixation (ORIF)
- Neurologic injury
  - S1, S2 deficits tend to improve within 1 year regardless of treatment
    – Unless nerve root avulsion present
  - Dural laceration may lead to cerebrospinal fluid leak

Treatment

- Options, risks, complications
  - Stable injury: Bed rest, slow progression to weight bearing
  - Unstable injury: Stabilization with transalar screws

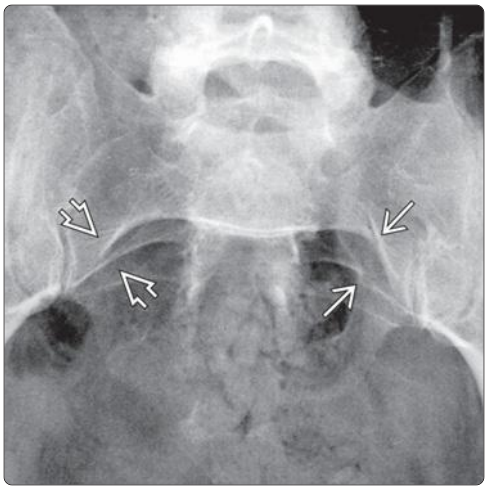
DIAGNOSTIC CHECKLIST

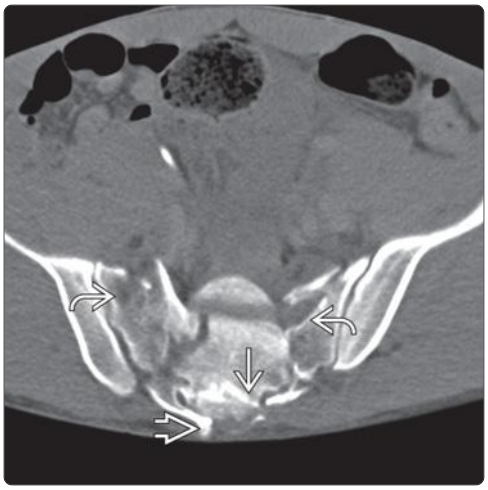
Image Interpretation Pearls

- Look for associated pelvic ring disruption in high-velocity injuries
- If L5 transverse process fracture is seen, suspect sacral fracture

SELECTED REFERENCES

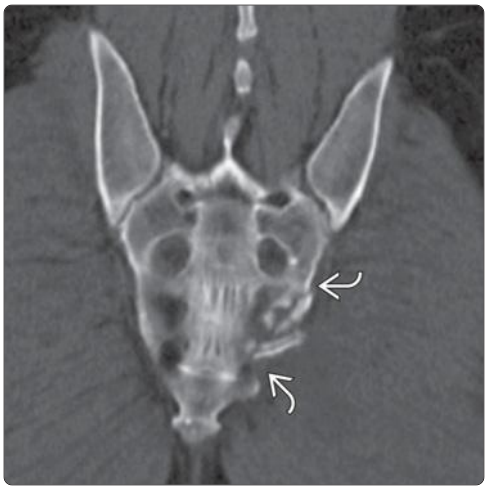
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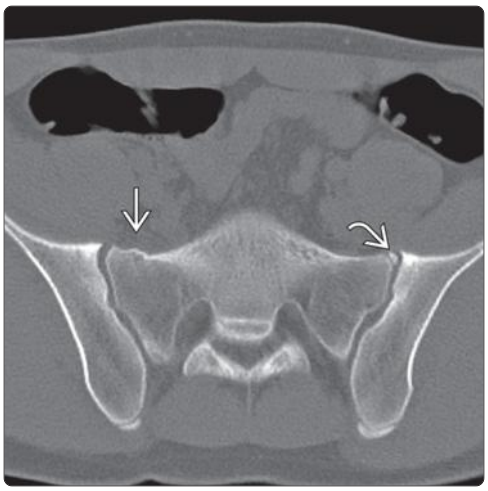


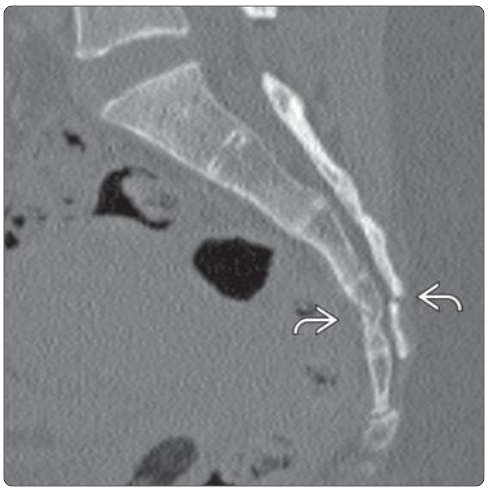
(Left) *Anteroposterior radiograph shows a left-sided zone II fracture due to lateral compression. Note angular deformity of arcuate lines on the left compared to the normal smooth contour of the uninjured arcuate lines on the right.* **(Right)** *Axial bone CT shows a very severely comminuted sacral fracture due to a parasailing accident. This includes a transverse fracture and vertical fractures through the neural foramina bilaterally and vertical fracture through the spinal canal.*





(Left) *Anteroposterior radiograph shows a zone II fracture. Fractures in this region are easily missed due to overlying soft tissue shadows. The most important clue in this case is disruption of the lateral sacral contour. Compare the normal right side.* **(Right)** *Coronal oblique bone CT in the same patient, angled with the plane of the sacrum, more readily shows the fracture.*





(Left) *Axial bone CT in a child shows a buckle fracture of the right sacrum. On the left, a normal apophysis is seen, recognizable by its rounded, sclerotic margins.* **(Right)** *Sagittal bone CT shows an isolated transverse sacral fracture due to a fall. Since these fractures are in the axial plane, they can be missed unless CT reformations or lateral radiograph of the sacrum are performed.*

Pedicle Stress Fracture

KEY FACTS

TERMINOLOGY

- Insufficiency fracture: Fracture due to physiological stress on abnormally weak bone
- Fatigue fracture: Fracture due to increased stress on normal bone

IMAGING

- CT, radiography: Vertically oriented, lucent fracture line through pedicle
  - Associated bony sclerosis reflects callus/healing &/or preexisting stress changes
  - ± contralateral posterior element defect (spondylolysis, pedicle fracture)
- MR: T1 hypointensity/T2 hyperintensity through pedicle marrow with acute or otherwise biomechanically unstable fracture
  - Marrow normalizes or converts to fatty marrow with healing
- Bone scan: ↑ radiotracer uptake in stress fracture

TOP DIFFERENTIAL DIAGNOSES

- Facet arthropathy, lumbar
- Spondylolysis
- Metastatic disease
- Primary bone tumor

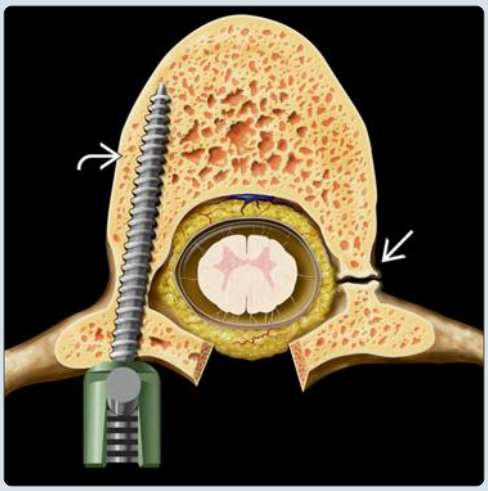
PATHOLOGY

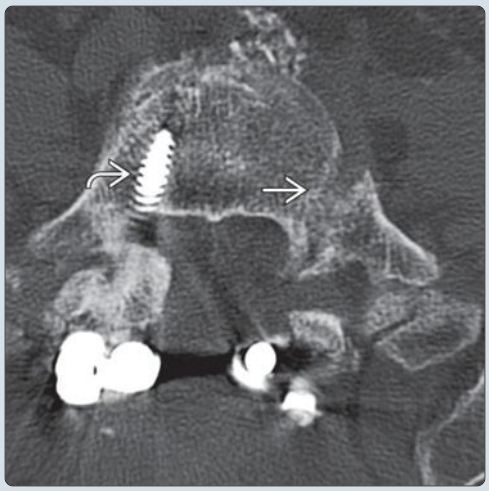
- Fatigue fracture: Increased &/or repetitive stress on normal bone, exceeding reparative capability
- Insufficiency fracture: Fracture from physiological levels of mechanical stress on weakened bone

DIAGNOSTIC CHECKLIST

- Pedicle stress fracture is one cause of abnormal STIR hyperintensity in pedicle marrow
- Pedicle marrow signal intensity changes seen in 1.7% of patients who underwent MR of the lumbar spine for back pain

(Left) *Axial graphic of lumbar vertebral body following unilateral fixation and midline laminectomy shows right pedicle screw placement, which altered spinal biomechanics to produce left pedicle stress fracture.* **(Right)** *There is a chronic stress fracture through the left L5 pedicle secondary to altered biomechanics with unilateral pedicle screw placement in this patient with multilevel laminectomies.*





(Left) *Sagittal T2WI MR demonstrates pedicle stress fracture as low signal with adjacent hyperintense edema. Pedicle screw tract from prior surgery also shows hyperintense signal in this patient with prior laminectomy and pedicle screw fixation.* **(Right)** *Sagittal STIR MR reveals hyperintense signal in the L5 pedicle extending into the superior articular facet and pars interarticularis due to stress fracture. Fine, linear hypointensity corresponds to the fracture line.*





TERMINOLOGY

Synonyms

- Pediculolysis

Definitions

- Insufficiency fracture: Fracture due to physiological stress on abnormally weak bone
- Fatigue fracture: Fracture due to increased stress on normal bone

IMAGING

General Features

- Best diagnostic clue
  - Lucent fracture line through pedicle on radiography, bone CT
- Location
  - Lumbar pedicle > cervical > thoracic
- Morphology
  - Vertically oriented fracture through pedicle
    - May be incomplete
  - May show bony remodeling from chronic mechanical stress
    - Cortical thickening, medullary sclerosis

Radiographic Findings

- Radiography
  - Linear fracture through pedicle
  - ± adjacent bony sclerosis
    - Vertebral anisocoria: Enlarged, sclerotic pedicle on AP radiograph

CT Findings

- Bone CT
  - Coronal oriented fracture through pedicle
    - Associated bony sclerosis reflects callus/healing &/or preexisting stress changes
    - Chronic pediculolysis may progress to hypertrophied pseudoarthrosis
  - Look for contralateral posterior element abnormality
    - Pedicle fracture
    - Spondylolysis
    - Laminectomy defect
  - ± degenerative disease of intervertebral disc and facet joints

MR Findings

- T1WI
  - T1 hypointensity through pedicle marrow with acute or otherwise biomechanically unstable fracture
    - Reflects edema or fibrovascular marrow replacement
  - Fracture line may be visible as a fine, linear T1 hypointensity
    - May be obscured by surrounding T1 marrow changes
  - With resolution of biomechanical instability, marrow changes resolve to normal or convert to fatty marrow signal (hyperintense on T1WI)
- T2WI, STIR
  - T2 hyperintensity through pedicle marrow with acute or otherwise biomechanically unstable fracture
    - Reflects edema or fibrovascular marrow replacement
  - Fracture line &/or cortical discontinuity may be visible as linear T2 hyperintensity
  - Marrow changes much more confidently seen on STIR than on T2WI
    - Obscured by fatty marrow signal on T2 FSE
  - With resolution of biomechanical instability, marrow changes resolve to normal or convert to fatty marrow signal (moderately hyperintense on T2WI, hypointense on STIR)
- T1WI C+
  - Areas of T1 marrow hypointensity (fibrovascular marrow replacement) enhance following intravenous contrast

Nuclear Medicine Findings

- Bone scan
  - Increased radionuclide uptake in pedicle with stress fracture due to reparative response
    - SPECT useful to reliably differentiate from spondylolysis
  - Increased uptake from associated abnormalities, such as contralateral spondylolysis

Imaging Recommendations

- Best imaging tool
  - MR (STIR or fat-suppressed T2WI) most sensitive for marrow changes in pedicles
    - Nonspecific for pediculolysis, unless fracture can be directly visualized
  - Limited bone CT is useful adjunct to assess for fracture, detect reparative sclerosis

DIFFERENTIAL DIAGNOSIS

Facet Arthropathy, Lumbar

- May see hypointense T1/hyperintense T2 marrow signal change in pedicle just above or below level of facet arthropathy
- Etiology unclear
  - Inflammation from facet arthropathy
  - Bony response to biomechanical instability/stress changes
  - Microfracture

Spondylolysis

- May see hypointense T1/hyperintense T2 marrow signal change in pedicle ipsilateral to acute spondylolysis
  - Tends to resolve with healing, especially in younger patients

Metastatic Disease

- Multiple lesions
- T1 hypointense, T2 hyperintense
- Destruction of cortex & medullary bone on CT

Primary Bone Tumor

- Osteoblastoma
- Osteoid osteoma

Paget Disease

- Bone expansion with thickened cortex and coarse trabecula
- May predispose to pathologic fracture

PATHOLOGY

General Features

- Etiology
  - Fatigue fracture
    - Increased &/or repetitive stress on normal bone, exceeding reparative capability → mechanical failure
    - Athletes
      - Typically sports requiring rotary movement of spine, such as baseball or cricket
    - Preexisting defect of neural arch
      - Congenital defect of neural arch
      - Contralateral spondylolysis
      - Laminectomy
    - Late middle-aged to elderly patients
      - Degenerative changes in intervertebral disc transfer axial loading to posterior elements
      - Degenerative changes in facet joints diminish ability of segment to maintain stability in AP direction
      - Increased mechanical stress on posterior elements, including pedicles → mechanical failure
    - Complication of spinal fusion
      - Increased stress transferred to vertebral components at margins of fusion → mechanical failure
  - Insufficiency fracture
    - Implies physiological levels of mechanical stress on weakened bone
      - Osteoporosis, radiation, Paget disease, etc.

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute-on-chronic back pain
- Other signs/symptoms
  - ± radiculopathy
    - May reflect dural inflammation or nerve root compression by coexisting spondylotic changes
- Clinical profile
  - Patient with new or suddenly worsening back pain

Demographics

- Age
  - Physically active adolescents & young adults, competitive athletes
  - Late middle-aged and elderly patients, associated with degenerative spondylosis
- Gender
  - No gender preference
- Epidemiology
  - Relatively uncommon, compared with fractures of pars interarticularis (spondylolysis)

Natural History & Prognosis

- Pain usually resolves with injury healing

Treatment

- Conservative treatment usually attempted 1st
  - Reduction in activity
  - Immobilization, bracing
- Surgical treatment (fusion or primary repair of pedicle with screw) reserved for failure of conservative treatment

DIAGNOSTIC CHECKLIST

Consider

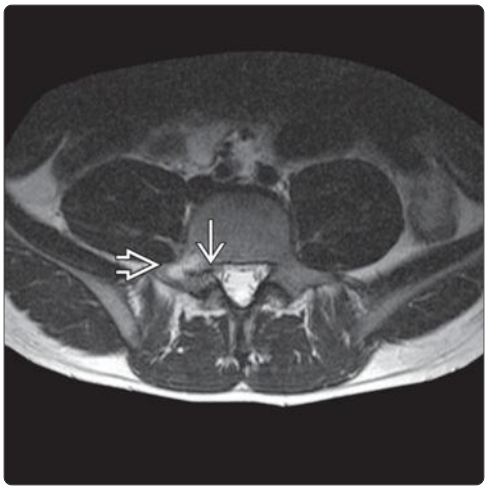
- Pedicle marrow signal intensity changes seen in 1.7% of patients who underwent MR of lumbar spine for back pain
  - Most commonly associated with 1 of 3 abnormalities: Degenerative facet disease, fractures of pars, or fracture of pedicle

Image Interpretation Pearls

- Pedicle stress fracture is one cause of abnormal STIR hyperintensity in pedicle marrow

SELECTED REFERENCES

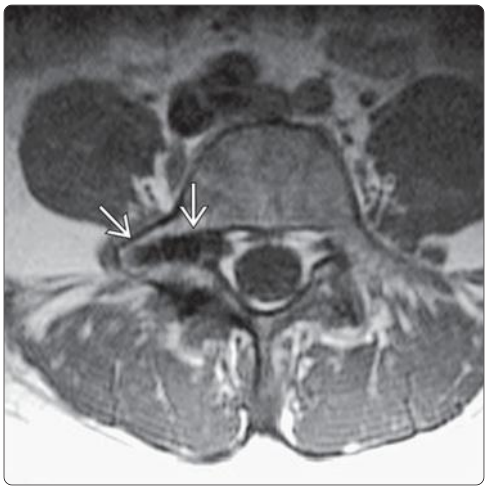
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(Left) *Axial T2WI MR in an adolescent with suddenly worsening back pain shows hyperintensity in the right L5 pedicle. The pedicle stress fracture is visualized as linear hypointense signal within the right pedicle. The left L5 pedicle is normal.* **(Right)** *Sagittal CT reconstruction in an adolescent patient with sudden onset of acute back pain reveals an acute, nondisplaced lumbar pedicular stress fracture of the right L5 pedicle.*





(Left) *Sagittal oblique bone CT shows a lucent cervical pedicle stress fracture line through C5-C6 left pedicle with sclerotic margins. Patient was post C5-C6 discectomy & failed interbody fusion.* **(Right)** *Axial T1WI MR demonstrates chronic right lumbar pedicle fracture as linear low signal with a large amount of bony hyperplasia and hyperintense fatty marrow replacement in the right pedicle and transverse process.*





(Left) *Axial T1WI MR demonstrates linear hypointense signal intensity in the bilateral L4 pedicles with normal surrounding marrow, reflecting healed pedicle stress fractures.* **(Right)** *Axial bone CT shows congenital left posterior element hypoplasia, and left pedicle aplasia. There is bony sclerosis of the compensatorily enlarged right pedicle, which contains a central horizontal lucency reflecting a fatigue stress fracture.*

Sacral Insufficiency Fracture

KEY FACTS

TERMINOLOGY

- Sacral fracture resulting from normal physiological stress on weakened (e.g., osteoporotic) bone

IMAGING

- Vertical fracture(s) through sacral alae
  - Either unilateral or bilateral
  - Located lateral to sacral foramina (zone I), roughly parallel to sacroiliac joint
- ± transverse fracture of sacral body
- Subtle ventral cortical disruption of sacral alae
- Sclerotic bands or irregular zones in sacral alae during healing phase
- Sacral marrow edema on MR
  - Hypointense on T1, hyperintense on T2
  - Greatest conspicuity on STIR or T2WI FS
  - May be overlooked on MR of lumbar spine obtained for generalized back pain
- Variable presence of classic H-shaped pattern of radiotracer uptake on bone scan: 19-62%
- Frequency of extrasacral tracer uptake 70% in 1 series
  - Multiple sites of tracer uptake may falsely raise concern for metastatic disease

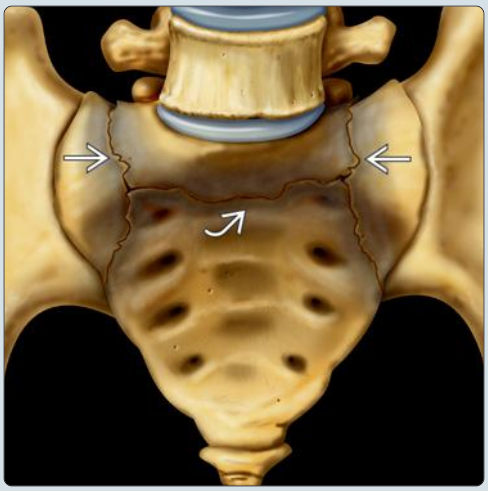
PATHOLOGY

- Associated abnormalities
  - Vertebral compression fractures
  - Other pelvic insufficiency fractures (pubic rami, iliac wing)
  - Intertrochanteric femur fracture

CLINICAL ISSUES

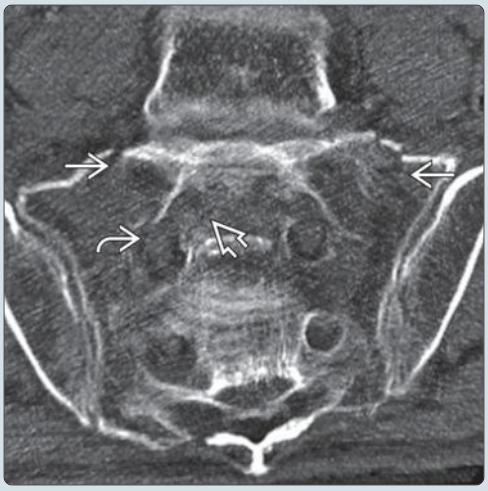
- In 1 series, only 43% with unilateral fractures and 0% with bilateral fractures regained pre-injury levels of mobility
- 1-year mortality: 14.3%

(Left) *Sagittal graphic depicts bilateral vertical sacral alar fractures, located lateral to the sacral foramina, and a transverse fracture through the sacral body, resulting in the classic H-shaped insufficiency fracture.* **(Right)** *Posterior bone scan shows increased sacral uptake in the classic "H" distribution due to insufficiency fracture. There is also uptake in a T12 compression fracture and in a pair of lower left rib fractures.*





(Left) *Coronal reformatted CT shows diffuse osteopenia and bilateral sacral alae cortical defects due to sacral insufficiency fracture. The right-sided fracture extends into the right S1 neural foramen. There is also a transverse fracture of the sacral body.* **(Right)** *Sagittal T1WI MR shows a small focus of low marrow signal involving the S3 body in the midline in this patient with insufficiency fracture. This location is important to evaluate in every lumbar spine MR.*





TERMINOLOGY

Definitions

- Sacral fracture resulting from normal physiological stress on weakened (e.g., osteoporotic) bone

IMAGING

General Features

- Best diagnostic clue
  - Abnormal sacral marrow signal (hypointense T1, hyperintense T2 or STIR)
- Location
  - Vertical fracture(s) through sacral alae
    - Either unilateral or bilateral
    - Located lateral to sacral foramina (zone I), roughly parallel to sacroiliac joint
  - ± transverse fracture of sacral body
- Morphology
  - Unilateral, bilateral, H-shaped

Radiographic Findings

- Radiography
  - Approximately 1/2 will be normal at time of presentation
  - Osteopenia
  - Vertical sclerotic bands in sacral ala
    - Healing fractures, seen weeks to months after onset of symptoms
  - Cortical disruption of sacral alae
  - Visualization of lucent fracture lines rare
  - Concomitant fractures of pubic rami, lumbar spine compression fractures, iliac wing fractures, intertrochanteric hip fractures

CT Findings

- Bone CT
  - Subtle cortical disruption
    - Ventral margins of sacral alae, just medial to sacroiliac joint(s)
    - Fracture typically lateral to sacral foramina
  - Direct visualization of fracture line, sacral deformity
    - Review sagittal reformatted images to identify sacral body fractures
  - Sclerotic bands or irregular zones in sacral alae
    - Healing phase
  - Vacuum phenomenon: Gas within fracture or sacroiliac joints
    - Seen with insufficiency fracture, not with pathologic fracture

MR Findings

- T1WI, T2WI, T2WI FS/STIR
  - Sacral marrow edema
    - Ill-defined and irregular
    - Hypointense on T1, hyperintense on T2
      - Greatest conspicuity on STIR or T2WI FS
      - Fatty marrow can obscure fluid signal on FSE T2WI
    - Unilateral, bilateral, or H-shaped (on coronal scans)
    - Adjacent to sacroiliac joints
      - Not involving or crossing sacroiliac joints
    - Seen within 3 weeks after symptom onset
    - Resolves after 3 months
    - May be overlooked on MR of lumbar spine obtained for generalized back pain
      - Best chance for diagnosis on sagittal STIR sequences
  - Presence of discrete fracture lines
    - Usually visualized between 3 weeks and 3 months
    - Lateral to sacral foramina, paralleling sacroiliac joints
- T1WI C+
  - Can enhance visualization of fracture line
  - Fracture appears as area of linear low signal surrounded by enhancing marrow edema

Nuclear Medicine Findings

- Bone scan
  - Bilateral or unilateral sacral radiotracer uptake ± horizontal component
  - Variable presence of classic H-shaped pattern of radiotracer uptake: 19-62%
    - "Honda" sign
  - Sensitive but not specific
    - Frequency of extrasacral tracer uptake 70% in 1 series
      - Due to fractures of pubic bone (50%) and spine (46%)
      - Multiple sites of tracer uptake may falsely raise concern for metastatic disease

Other Modality Findings

- Osteoporosis on bone densitometry

Imaging Recommendations

- Best imaging tool
  - MR most sensitive for marrow signal changes associated with subtle fracture
  - CT useful adjuvant to MR imaging
- Protocol advice
  - Sagittal, axial, and oblique coronal (plane of sacral body); axial and oblique coronal STIR or T2WI FS; post-contrast imaging may be considered but often not necessary

DIFFERENTIAL DIAGNOSIS

Sacral Traumatic Fracture

- 95% occur with other fractures of pelvis

Sacral Metastases

- Discrete marrow abnormality, random distribution
- Osteolysis on CT, rather than linear disruption of trabecula & cortex
- Extraosseous soft tissue mass
- Extrasacral involvement

Chordoma

- Solitary mass
- Cortical destruction, extraosseous soft tissue

Sacral Osteomyelitis

- Ill-defined, asymmetric marrow edema
  - May involve both sides of sacroiliac joint
- Osteolysis
- Soft tissue edema, abscess, sinus tract
- No fracture lines

Sacroiliitis

- Erosions, sclerosis, marrow edema adjacent to sacroiliac joints
- SI joint ankylosis

Osteoarthritis

- Sclerosis on iliac side of sacroiliac joints
- Anterior osteophytes

PATHOLOGY

General Features

- Etiology
  - Predisposing factors
    - Osteoporosis
    - Radiation therapy to pelvis
    - Endogenous or exogenous corticosteroid excess
    - Renal osteodystrophy
    - Paget disease
    - Other causes of osteopenia
  - History of minor trauma may be elicited
  - Finite-element modeling suggests stresses placed on sacrum by ambulation causes majority of insufficiency fractures in osteoporotic patients
  - Shearing causes vertical fracture through lateral sacral ala
    - Horizontal component of fracture seems to be secondary development
- Associated abnormalities
  - Vertebral compression fractures
  - Other pelvic insufficiency fractures
    - Pubic rami
    - Acetabular roof
    - Iliac wing
  - Intertrochanteric femur fracture
  - Neurologic symptoms rare
    - Sphincter disturbance, lower extremity paresthesias

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific buttock or lumbar spine pain
    - Exacerbated by movement
    - Radiating to leg or groin
  - Sacral tenderness
- Clinical profile
  - Pain exacerbated by activity
  - Relieved by rest

Demographics

- Age
  - 6th through 8th decades of life
- Gender
  - Vast majority occur in elderly females
- Ethnicity
  - No racial predilection
- Epidemiology
  - Incidence: 0.14-1.8%

Natural History & Prognosis

- In 1 series, only 43% with unilateral fractures and 0% with bilateral fractures regained pre-injury levels of mobility
- 1-year mortality: 14.3%

Treatment

- Conservative
  - Bed rest
  - Protected weight-bearing
  - Analgesics
  - Physical therapy
  - Treatment of underlying osteoporosis
- Sacroplasty
  - Injection of polymethylmethacrylate

DIAGNOSTIC CHECKLIST

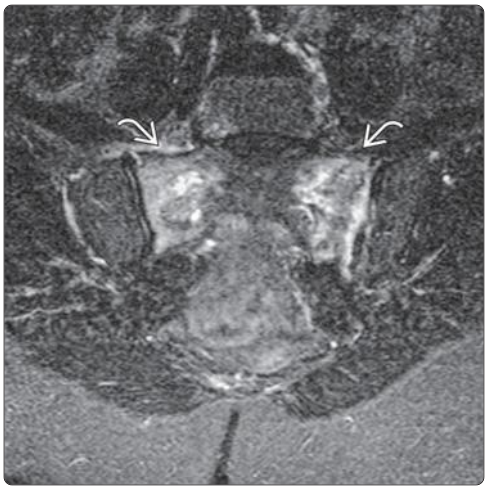
Image Interpretation Pearls

- Make sure to review visualized sacrum on lumbar spine MR (especially sagittal STIR)

SELECTED REFERENCES

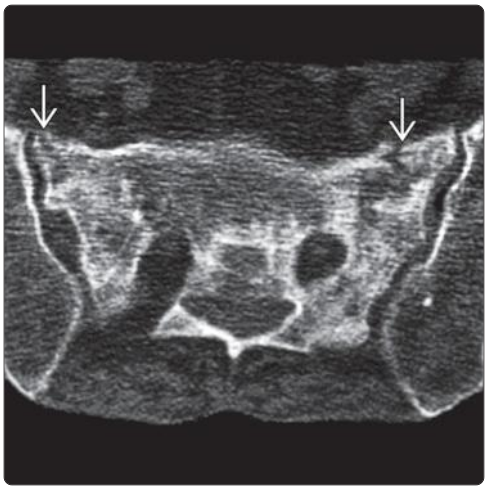
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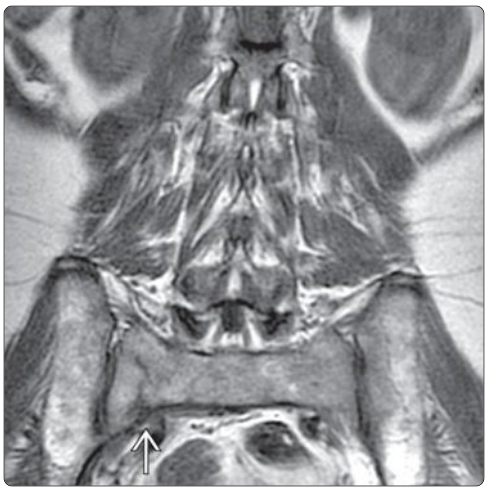


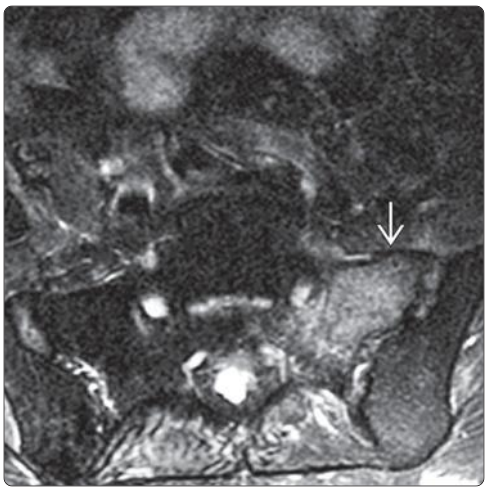
(Left) *Coronal T1WI MR shows abnormal hypointense marrow signal in both sacral alae due to bilateral sacral insufficiency fractures.* **(Right)** *Coronal T2WI FS MR in the same patient shows markedly hyperintense marrow in both sacral alae due to insufficiency fracture.*





(Left) *Axial NECT shows focal disruption of the ventral cortices of bilateral sacral alae due to insufficiency fractures. Note the severe osteopenia.* **(Right)** *Axial NECT shows linear fractures of bilateral sacral alae. Patchy sclerosis reflects healing of a subacute injury.*





(Left) *Coronal T1WI MR shows unilateral (right) sacral insufficiency fracture. The fracture line is visible as a fine, irregular hypointensity.* **(Right)** *Axial T2WI FS MR shows diffusely hyperintense signal in the left sacral ala due to unilateral insufficiency fracture. Focal cortical irregularity identifies the actual site of the fracture.*

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PART II
SECTION 2

Cord, Dura, and Vessels



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SCIWORA

KEY FACTS

TERMINOLOGY

- Spinal cord injury without radiological abnormality (SCIWORA)

IMAGING

- Plain radiographs, CT classically normal or negative for acute injury
- MR frequently abnormal
  - Spinal ligament disruption, intervertebral disc herniation, paraspinal muscle injury, &/or shearing of vertebral endplate subepiphyseal growth zone

TOP DIFFERENTIAL DIAGNOSES

- Penetrating spinal trauma
- Electrical shock
- Obstetric complications
- Congenital spine anomalies

PATHOLOGY

- Hyperextension/flexion or longitudinal distraction most common injury mechanisms
- Primary spinal cord injury &/or spinal cord infarction

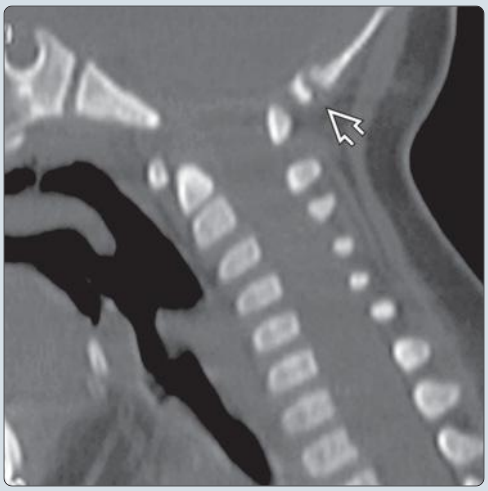
CLINICAL ISSUES

- Pediatric, adult, and thoracic SCIWORA subtypes
  - Children > adults
- Spinal cord abnormality demonstrated on MR is highly predictive of outcome
  - SCIWORA prognosis better than SCI with radiologic evidence of injury
- Traumatic brain injury commonly associated

DIAGNOSTIC CHECKLIST

- Plain radiographs, CT negative (by definition in SCIWORA) although subtle abnormalities may be visible on careful review

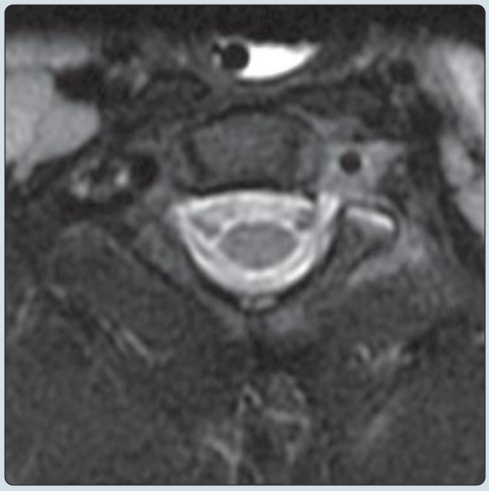
(Left) *Sagittal bone CT (closed head injury, extremity weakness) shows normal craniovertebral junction alignment and no cervical spinal fracture. There is a minimally displaced occipital bone fracture ⇨ with overlying scalp edema.* **(Right)** *Sagittal STIR MR (extremity weakness) reveals unexpected mild abnormal T2 prolongation within the upper cervical spinal cord ⇨. No spinal ligamentous injury was detected, although scalp edema from occipital bone ⇨ fracture is conspicuous.*





(Left) *Sagittal STIR MR (abusive head trauma, normal cervical CT) shows abnormal enlargement and T2 hyperintensity ⇨ within the cervical spinal cord in conjunction with interspinous/supraspinous ligament edema ⇨.* **(Right)** *Axial T2 FS MR (abusive head trauma, right hemiparesis) confirms abnormal T2 hyperintensity within the cervical cord involving both gray and white matter.*





TERMINOLOGY

Abbreviations

- Spinal cord injury without radiological abnormality (SCIWORA), spinal cord injury (SCI)

Definitions

- Pre-MR term (Pang and Wilberger, 1982)
  - Objective traumatic SCI without ligamentous injury or fracture detectable on radiographs
  - Original definition excludes penetrating trauma, electrical shock, obstetric complications, and congenital spine anomalies
- Term SCIWORA is antiquated in MR era
  - Intervertebral disc herniation, spinal epidural hematoma, spinal cord contusion, and hematomyelia may be occult on CT and radiographs but obvious on MR

IMAGING

General Features

- Best diagnostic clue
  - Clinical neurological deficit following injury localizes to spinal cord in absence of explanatory neuroimaging abnormality
- Location
  - Cervical > > thoracic

Radiographic Findings

- Radiography
  - May be normal or show chronic degenerative changes
  - No fracture or subluxation

CT Findings

- Bone CT
  - May be normal or show chronic degenerative changes
  - No fracture or subluxation
  - No facet joint or disc space widening in classic cases
    - Subtle abnormalities may be detectable on high quality multiplanar CT imaging in some cases

MR Findings

- T1WI
  - Ligamentous disruption, intervertebral disc herniation, paraspinal muscle injury, shearing of vertebral endplate subepiphyseal growth zone
- T2WI
  - Same as T1WI; edema more conspicuous than on T1WI but may be difficult to distinguish from bright fat without fat saturation
  - ± spinal cord injury (ranges from edema → spinal cord transection)
- STIR
  - Most sensitive for spinal cord injury, ligamentous edema, soft tissue injury
- T2* GRE
  - ± spinal cord or epidural/subdural hemorrhage

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging
- Protocol advice
  - Fat-saturated T2WI or STIR MR, T2* GRE necessary

DIFFERENTIAL DIAGNOSIS

Penetrating Spinal Trauma

- Osseus injury is frequently conspicuous but may be occult with penetrating blade injuries
- Search for bullet fragments in gunshot trauma

Electrical Shock

- Bones normal
- Look for soft tissue or spinal cord injury

Obstetric Complications

- Large infant, small maternal pelvic biometry, shoulder dystocia
- Distraction injuries may show intervertebral disc space widening at injury level
- MR most sensitive modality for plexus, spinal cord, soft tissue injuries
  - Obstetrical brachial plexus and spinal cord injuries usually occult on plain radiographs, CT

Congenital Spine Anomalies

- Vertebral anomalies usually obvious on plain radiographs
- Look for associated congenital spinal stenosis, kyphosis, diastematomyelia spur

PATHOLOGY

General Features

- Etiology
  - Injury mechanisms include hyperextension/flexion, longitudinal distraction, and ischemic spinal cord injury
    - Flexion and extension injuries most common mechanism
    - May also be lateral bending, longitudinal distraction, rotation, axial loading, or combination of mechanisms
  - Factors predisposing young children to SCIWORA include
    - Ligamentous laxity, hypermobility of pediatric osseous cervical, thoracic spine
      - Vertebral column > spinal cord elasticity
      - Permits nonphysiologic deformation of vertebral column → direct spinal cord contusion or secondary stretch injury
    - Spinal cord blood supply more tenuous than adults
  - Primary spinal cord injury
    - Contusion, transection, or stretch injury
    - Penetrating injuries
      - Most commonly displaced bone fragments penetrate spinal cord &/or segmental spinal nerves
      - Bullets or bladed weapons less common mechanism
  - Traumatic spinal cord infarction
    - SCIWORA subset
    - Traumatic occlusion of thoracic aortic arterial branches to thoracic spinal cord
    - Usually presents with delayed cord injury
    - High incidence of associated blunt thoracic or retroperitoneal trauma, arterial hypotension
- Associated abnormalities
  - Traumatic brain injury (TBI) most common associated injury (37%)
  - Other axial, appendicular skeletal injuries

- Intraabdominal injuries

Gross Pathologic & Surgical Features

- Usually single injury level

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pediatric SCIWORA
    - Predilection for upper cervical spine
    - More likely to have severe neurological injuries, delayed neurological deficits
  - Adult SCIWORA
    - Often concurrent cervical spondylosis
    - Vascular compromise from smoking or vascular disease may accentuate injury severity
  - Thoracic SCIWORA
    - High-speed direct impact, distraction from lap belts, and crush injury by slow-moving vehicles

Demographics

- Age
  - More common in children < 8 years of age than in adults
- Gender
  - Follows trauma demographics
- Epidemiology
  - Reported incidence of pediatric SCIWORA ranges from 3.3-36% (includes pre- and post-MR era studies)
  - Most common mechanisms include sports injuries, nonaccidental trauma, motor vehicle trauma

Natural History & Prognosis

- Type of spinal cord abnormality demonstrated on MR is highly predictive of outcome
  - More severe imaging abnormalities → worse prognosis
- Prognosis better in SCIWORA than SCI with radiologically evident traumatic injury

Treatment

- Many SCIWORA injuries are stable
  - Treatment based on which structures injured and their importance to spinal column stability
- Options range from conservative bracing for stable injuries to surgical fusion for unstable injuries

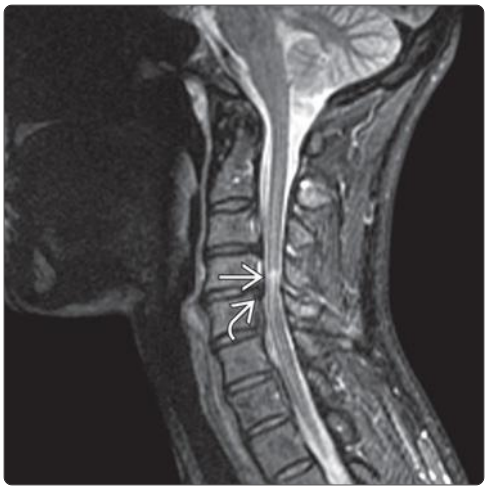
DIAGNOSTIC CHECKLIST

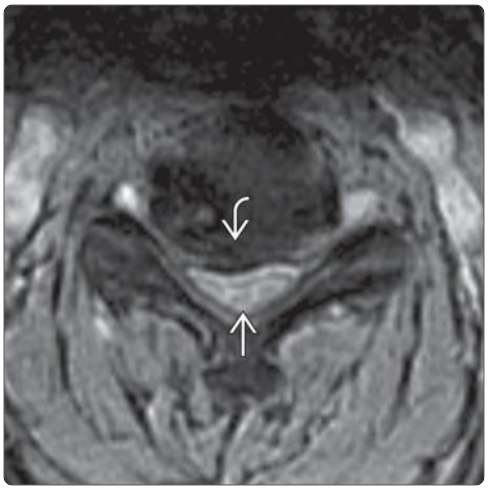
Consider

- Plain radiographs and CT negative by definition in SCIWORA, although subtle abnormalities may be visible on careful review
- MR findings include ligamentous or disc injury, spinal cord transection, spinal cord hemorrhage, or normal
- Term SCIWORA is antiquated in MR era because it does not take into account MR imaging detection of soft tissue or spinal cord injuries that are occult on CT or radiographs
  - Better term would be "spinal cord injury without neuroimaging abnormality" (SCIWONA)
  - "SCIWONA" acronym not currently in widespread use

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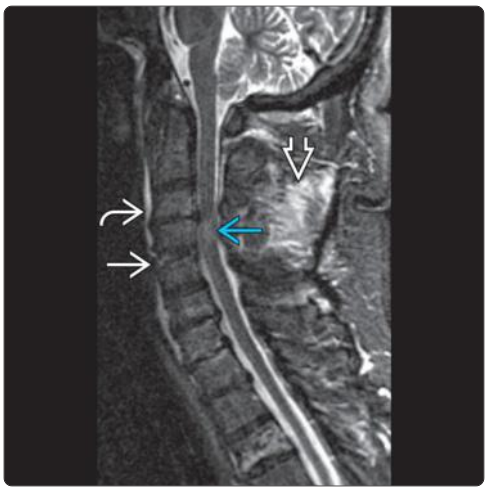
(Left) *Sagittal STIR MR (vehicular collision, clinical central cord syndrome), obtained following negative cervical radiographs and CT for acute injury, reveals an acute disc herniation and cord contusion superimposed on preexisting spinal stenosis. No significant acute ligamentous injury is demonstrated.* **(Right)** *Axial T2* GRE MR (vehicular collision, clinical central cord syndrome) demonstrates a right paracentral disc herniation and spinal cord contusion.*





(Left) *Sagittal bone CT (auto-pedestrian collision, closed head injury) shows subtle widening of the C2-C3 disc space and mild anterior displacement of C2 on C3. No fracture is demonstrated.* **(Right)** *Sagittal STIR MR shows prevertebral fluid, epidural hematoma, and abnormal C2-C3 disc space fluid signal. Extensive posterior soft tissue injuries include disruption of the posterior longitudinal ligament and supraspinal and interspinous ligaments.*





(Left) *Sagittal bone CT (helmeted bicyclist hit by car with acute upper extremity weakness) demonstrates moderately severe multilevel degenerative disc changes but no acute fracture.* **(Right)** *Sagittal STIR MR (upper extremity weakness and clinical central cord syndrome) reveals prevertebral fluid, C4-C5 disc injury, C3-C4 disc protrusion producing spinal cord contusion, and severe posterior muscular injury and interspinous ligament injury.*

Post-Traumatic Syrinx

KEY FACTS

TERMINOLOGY

- Post-traumatic syrinx (PTS)

IMAGING

- Fusiform intramedullary hyperintensity tracking CSF signal
  - Myelomalacia precedes overt syrinx formation = "presyrinx state"
- Cystic expansile cord lesion
  - May appear to be "expansile" lesion, relative finding in presence of cord atrophy
- Consider cine PC CSF flow study if suspected obstruction to CSF flow (e.g., arachnoid adhesions)

TOP DIFFERENTIAL DIAGNOSES

- Gibbs artifact
- Nontraumatic syrinx
- Myelitis
- Myelomalacia

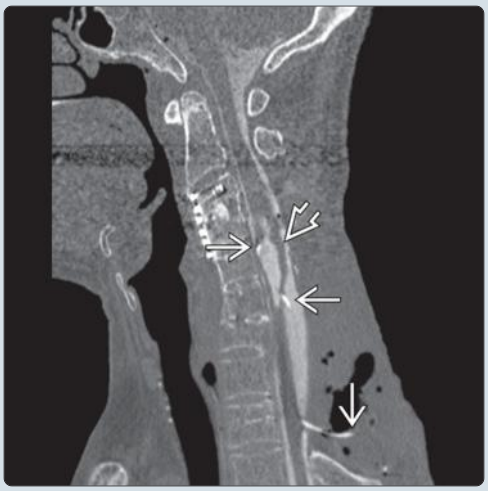
PATHOLOGY

- Current treatment assumes syrinx is related to post-traumatic arachnoid scarring and CSF flow obstruction at trauma level

CLINICAL ISSUES

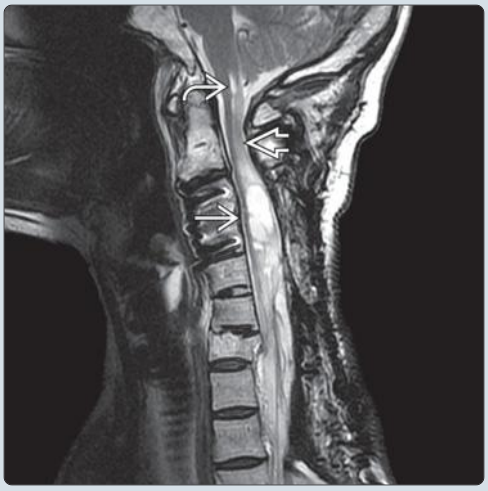
- Symptoms include spasticity, hyperhidrosis, pain, sensory loss, automotive hyperreflexia
- Classic presentation: Severe pain unrelieved by analgesics; ascending disassociated sensory loss
- Surgery reserved from patients with progressive neurological symptoms
  - First-line treatment has moved away from shunting of syrinx to restoring normal CSF flow patterns at traumatic site
    - Untethering of cord
    - Duraplasty
    - Spine realignment or fusion may be added if angulation or stability is problematic





(Left) *Sagittal images showing extension of syrinx over time into regions of "presyrinx" edema are presented. Initial T2 MR after C3-C4 fusion and prior flexion injury at C5-C6 shows the well-defined syrinx cavity at the C5-C6 level with cord expansion. T2 hyperintense signal within the cord extending cephalad from the syrinx to the C2 level due to cord edema.* **(Right)** *Sagittal CT myelogram following placement of a syringoperitoneal shunt shows the shunt catheter within the syrinx cavity, which is decreased in size.*





(Left) *Follow-up sagittal T2 MR 3 months later shows extension of the syrinx to the C3 level with slight edema cephalad to this site. Note the presence of a shunt catheter, which has not stopped the syrinx progression.* **(Right)** *Sagittal T2 MR 6 months later shows expansion of the syrinx at the C3 level and marked, increased "presyrinx" edema extending cephalad to the medulla with a small focus of syringobulbia.*

TERMINOLOGY

Abbreviations

- Post-traumatic syrinx (PTS)

Synonyms

- Syringomyelia, syringohydromyelia

Definitions

- Cystic cord cavity that may (hydromyelia) or may not (syringomyelia) communicate with central canal
  - Artificial distinction → syringomyelia is commonly used term
- PTS implies syrinx formation is related to prior trauma
  - Syrinx may occur in post-trauma patients with complete recovery from trauma, and patients without direct cord injury
- Cephalad extension to brainstem medulla is termed syringobulbia

IMAGING

General Features

- Best diagnostic clue
  - Cystic expansile cord lesion with CSF signal intensity on MR
- Location
  - Rostral to injury site in 81%, caudal in 4%, both directions in 15%
- Size
  - Average length: 6 cm; range: 5 mm to entire cord
- Morphology
  - Longitudinal spinal cord cavity with CSF signal/attenuation
  - Frequently has fusiform "beaded" appearance

Radiographic Findings

- Radiography
  - Signs of chronic spine injury

CT Findings

- NECT
  - Often difficult to appreciate CSF attenuation central cavity on unenhanced imaging

MR Findings

- T1WI
  - Fusiform intramedullary hypointensity relative to cord parenchyma
- T2WI
  - Fusiform intramedullary hyperintensity tracking CSF signal
    - Myelomalacia precedes overt syrinx formation = "presyrinx state"
    - May begin as small cysts, which do not precisely track CSF signal, and then coalesce into larger cysts demonstrating bulk water signal
  - Adjacent cord may be normal or atrophied
- CSF flow
  - Sagittal 2D cine phase-contrast technique for defining presence, direction of flow
  - May show abnormal CSF dynamics

Ultrasonographic Findings

- Intraoperative US: Delineates location, extent

Nonvascular Interventions

- Myelography
  - Focal changes in cord size
  - Delayed CT myelography
    - Syrinx fills in with delayed scanning
    - May change with position, characteristically ↓ in size when moving from prone to supine
  - Look for arachnoiditis, adhesions, or myelographic block at level of injury

Imaging Recommendations

- Best imaging tool
  - T2WI, T1 C+ MR
- Protocol advice
  - Axial images confirm location and clarify relationship to adjacent anatomical structures
  - Sagittal sequence most useful for defining craniocaudal extent
  - Initial contrast examination essential to exclude neoplasm; typically unnecessary for continued surveillance
  - Consider cine PC CSF flow study if suspected obstruction to CSF flow (e.g., arachnoid adhesions)

DIFFERENTIAL DIAGNOSIS

Gibbs Artifact

- Bright or dark lines parallel and adjacent to borders of abrupt intensity change
- Commonly seen when going from bright CSF to dark spinal cord on T2WI

Nontraumatic Syrinx

- Chiari 1
- Spinal cord tumor
  - Ependymoma, astrocytoma
  - Hemangioblastoma
  - Cord expansion; typically enhance
- Idiopathic
  - Nontraumatic hydromyelia
  - Ventriculitis terminalis (distal cord)

Myelitis

- Demyelinating disease (e.g., MS, NMO)

Myelomalacia

- Cord volume loss, gliosis
- Do not see CSF signal central cavitation on T1WI

PATHOLOGY

General Features

- Etiology
  - Current treatment assumes syrinx is related to post-traumatic arachnoid scarring and CSF flow obstruction at trauma level
    - May also develop from contusion, myelomalacia, or hemorrhage within cord
  - "One-way valve" theory: Phenomenon allowing CSF into, but not out of, cyst cavity

- "Slosh-and-suck" theory: ↑ epidural venous flow during activity (e.g., straining, sneezing, violent coughing), ↑ pressure around cord, which cannot be dissipated because of disruptions in CSF flow forcing CSF into cyst

Gross Pathologic & Surgical Features

- Cavitation of gray matter within spinal cord centrally or eccentrically

Microscopic Features

- Forms in partially damaged cord adjacent to area of complete injury
- May begin as small areas of cavitation that coalesce into larger cysts
- Tubular spinal cord cavitation surrounded by dense glial fibril wall (Rosenthal fibers)
  - Predominantly affecting decussating pain and temperature fibers
- Adhesions often present tethering cord with local scarring
- Enlargement secondary to cavity fluid motion in response to pressure changes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Spasticity, hyperhidrosis, pain, sensory loss, automotive hyperreflexia
  - May result in neurologic deterioration years after injury
    - Delays from 6 months to 35 years reported
    - Mean interval ranges 9-13 years
  - Cervical: Rarely CN deficit(s), 25% have bulbar signs
  - Rarely new onset urinary dysfunction, neuropathic joint
- Clinical profile
  - Most common symptom: Pain, which may increase with coughing, sneezing, or straining
    - Axial > radicular pain
  - Classic presentation: Severe pain unrelieved by analgesics, ascending disassociated sensory loss
    - Loss of pain and temperature, damage to spinothalamic tracts
  - Uncommon in childhood
    - Most childhood syrinx is simple hydromyelia or Chiari-associated

Demographics

- Age
  - May occur at any age; average age at time of injury ~ 28 years (range: 16-67 years)
- Gender
  - M > F
- Epidemiology
  - Trauma most common cause of secondary syrinx
  - PTS incidence difficult to determine
    - Spinal cord injury patients have 2% per year risk of deterioration from multiple causes
      - Instability, cord tethering, cord compression, syringomyelia

Natural History & Prognosis

- Variable; dependent on underlying etiology
- PTS onset earlier with ↑ age, cervical and thoracic levels, dislocated fracture, spinal instrumentation without decompression, complete SCI
- As cavity enlarges
  - Involves previously unaffected neural tissue
  - Results in progressive myelopathy
  - Heralded by changing or new symptoms
- Muscle wasting ± spasticity
- Neuropathic (Charcot) joints may occur long-term

Treatment

- Surgery reserved from patients with progressive neurological symptoms
- First-line treatment has moved away from shunting of syrinx to restoring normal CSF flow patterns at traumatic site
  - Untethering of cord
  - Duraplasty
  - Spine realignment or fusion may be added if angulation or stability is problematic
- Other surgical considerations
  - Spinal cord transection (cordectomy) is effective form of treatment for syrinx in patients with complete cord lesion
- Shunting is associated with high rate of long-term complications with frequent revisions
  - Only treats symptoms related to syrinx, and not symptoms associated with tethering or adhesions

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Main imaging goal is to define location and extent of syrinx, and to exclude other lesions

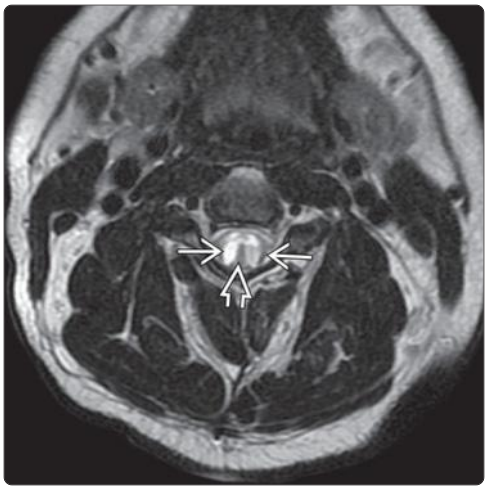
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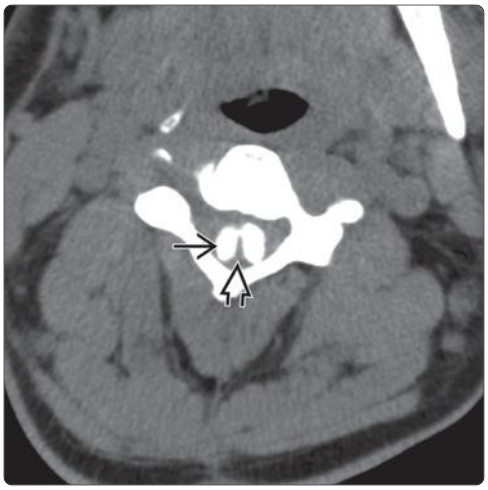
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(Left) *Sagittal T2 MR depicts fracture-dislocation at the C6-C7 level with associated cord contusion and T2 hypointensity on the cord surface due to blood products. There is disruption of the ligamentum flavum and the interspinous ligaments.* **(Right)** *Sagittal T2 MR in the same patient shows interval increase in the syrinx above the level of the cord injury. There is cord expansion and edema, which extends cephalad to the medulla.*





(Left) *Axial T2 MR reveals intramedullary hyperintensity related to syrinx within the right and left lateral aspects of the cord. There is a preserved cleft of tissue separating the syrinx.* **(Right)** *Axial CT myelogram demonstrates filling of the syrinx cavity with contrast. A cleft of tissue separates the syrinx cavity.*





(Left) *Sagittal T2 MR demonstrates a focal, post-traumatic kyphosis of the thoracic spine, and diffuse expansion and abnormal hyperintense signal of the thoracic cord. A 2-level laminectomy has been performed to decompress the thoracic canal.* **(Right)** *Axial CT myelogram (delayed acquisition following intrathecal contrast injection) shows a syrinx cavity in the midcervical cord, which has opacified with contrast, similar in density to the subarachnoid CSF.*

Presyrinx Edema

KEY FACTS

TERMINOLOGY

- Potentially reversible state of spinal cord edema caused by obstruction or alteration of normal cerebrospinal fluid (CSF) flow pathways

IMAGING

- Increased T2 signal within central aspect of cord, with slightly decreased and ill-defined T1 signal and cord expansion in setting of pathology that alters CSF flow dynamics

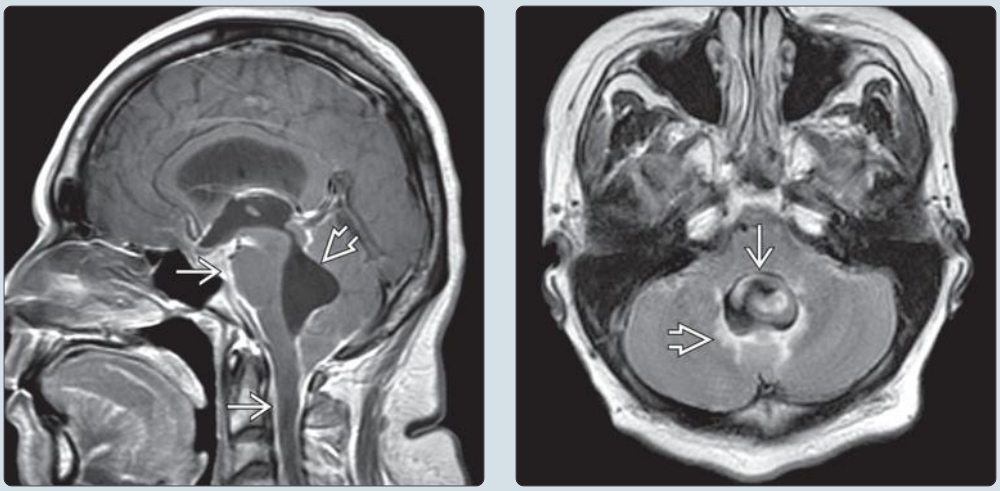
TOP DIFFERENTIAL DIAGNOSES

- Post-traumatic syringomyelia
- Nontraumatic syringomyelia
  - Chiari 1 malformation
  - Tumor associated (ependymoma, astrocytoma, hemangioblastoma)
  - Idiopathic
- Myelitis
  - Demyelinating disease
  - Viral infection
  - Vasculitis (SLE)
- Infarction
- Myelomalacia
- Type I dural arteriovenous fistula
- Posterior fossa dural fistula with intraspinal drainage
- Radiation myelopathy/necrosis

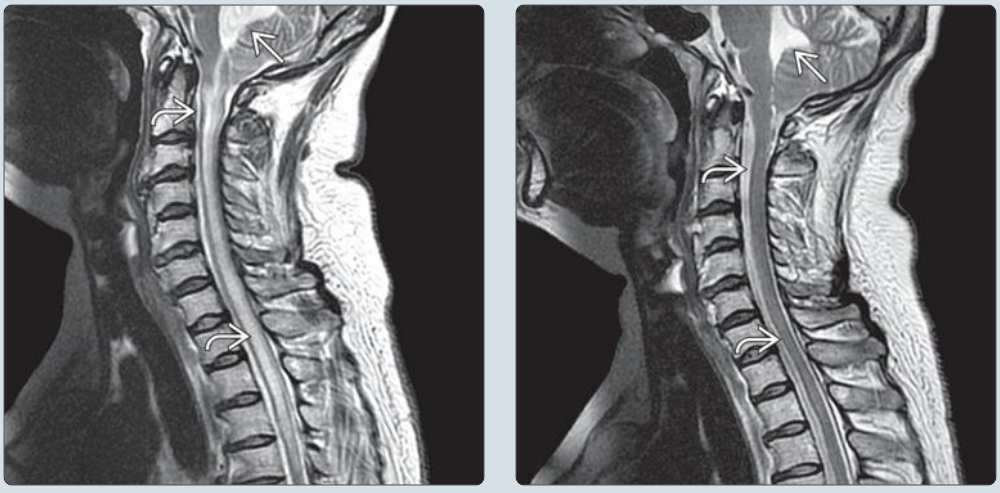
DIAGNOSTIC CHECKLIST

- Presyrinx edema likely represents a point on continuum to development of syringomyelia
- May be misinterpreted as syringomyelia on MR studies
- May be related to progressive post-traumatic myelomalacic myelopathy

(Left) *Sagittal T1WI C+ MR of brain shows severe hydrocephalus ⇨ from diffuse coccidioidomycosis basilar meningitis with diffuse leptomeningeal enhancement ⇨.* **(Right)** *Axial FLAIR MR through the posterior fossa in this case of coccidioidomycosis meningitis shows transependymal edema ⇨ surrounding the markedly dilated 4th ventricle ⇨.*



(Left) *Sagittal T2 MR shows extensive cervical cord presyrinx edema ⇨ that is due to coccidioidomycosis meningitis causing marked hydrocephalus ⇨. The cervical subarachnoid space and 4th ventricular outflow obstruction causes abnormal fluid shift into the cord.* **(Right)** *Extensive cervical cord edema (presyrinx) resolution following ventricular shunting: Follow-up MR study 7 days after lateral ventricular shunt placement shows resolved cervical cord edema ⇨ and diminished 4th ventricular size ⇨.*



TERMINOLOGY

Synonyms

- Presyrinx state, interstitial cord edema

Definitions

- Potentially reversible state of spinal cord edema caused by obstruction or alteration of normal cerebrospinal fluid (CSF) flow pathways
- ± presence of syrinx

IMAGING

General Features

- Best diagnostic clue
  - Increased T2 signal within central aspect of cord, with slightly decreased and ill-defined T1 signal and cord expansion in setting of pathology that alters CSF flow dynamics
- Location
  - Variable, but cervical cord most common
- Size
  - Varies from < 1 vertebral body length to longitudinally extensive

CT Findings

- CECT
  - Myelography shows CSF flow obstruction related to adhesive arachnoiditis (block or partial block)

MR Findings

- T1WI
  - Slightly decreased signal within central aspect of cord, with ill-defined margins
  - Does not tract CSF signal
- T2WI
  - Increased signal within central cord, with ill-defined margins (unlike syrinx with sharp margins and loculations)
- T2* GRE
  - Constructive interference in steady state (CISS)-type imaging is good for definition of CSF loculations with arachnoiditis, or subtle subarachnoid adhesions
- T1WI C+
  - No enhancement of cord unless secondary pathology is present
- MR cine
  - Shows abnormal flow, reflected in diminished or absent flow, or increased jet effect with stenosis/partial obstruction

Imaging Recommendations

- Best imaging tool
  - MR with contrast to exclude other intrinsic inflammatory conditions and neoplasms of spinal cord
  - Exclude leptomeningeal enhancement with meningitis
- Protocol advice
  - CISS imaging for best evaluation of membranes or adhesions

DIFFERENTIAL DIAGNOSIS

Post-Traumatic Syringomyelia

- Cystic expansile cord lesion, which tracts CSF signal on all sequences
- May show CSF dephasing with flow
- Loculations

Nontraumatic Syringomyelia

- Chiari 1 malformation
- Tumor associated
  - Ependymoma
  - Astrocytoma
  - Hemangioblastoma
- Idiopathic

Myelitis

- Demyelinating disease
  - Multiple sclerosis
  - Neuromyelitis optica
  - Acute disseminated encephalomyelitis
- Viral infection
- Vasculitis (SLE)

Infarction

- Acute symptom onset
- Ventral 2/3 of cord involved for anterior spinal artery disease

Myelomalacia

- Cord volume loss

Type I Dural Arteriovenous Fistula

- Long-segment cord edema with well-defined vascular flow voids in subarachnoid space
- Uncommon in cervical spine

Posterior Fossa Dural Fistula With Intraspinal Drainage

- Cord edema with prominent vascular flow voids in subarachnoid space mimics intraspinal dural fistula

Radiation Myelopathy/Necrosis

- Irregular enhancement
- 1 month to years following fractionated radiotherapy

PATHOLOGY

General Features

- Etiology
  - Pathophysiology of syrinx and presyrinx controversial, with multiple theories proposed
  - Whatever the specific cause, CSF flow at craniocervical junction or cervical spine is altered or restricted → mismatch between rate of CSF entering cervical cord and central canal vs. rate of CSF elimination from cord
    - Increased pulse pressure in subarachnoid space causing CSF to move through perivascular/interstitial spaces of spinal cord toward central canal, resulting in edema of spinal cord

        - Development of syrinx that does not communicate with 4th ventricle and is remote from site of CSF obstruction has been explained by variations in competence of central canal and degree and location of canal stenosis
        - Presyrinx may occur if central canal is not patent and CSF entering perivascular/interstitial spaces of spinal cord parenchyma is unable to enter central canal to form discrete cyst
      - Syrinx may be filled with extracellular fluid derived from microcirculation of spinal cord rather than directly from CSF
        - Studies have shown evidence that CSF is produced not only in choroid plexus but also in brain parenchyma and spinal cord
        - Increases in extracellular fluid caused by disturbed absorption from intramedullary venous system may be important mechanism of syringomyelia associated with spinal adhesive arachnoiditis
    - **Multiple underlying causes of presyrinx edema**
      - **Chiari 1 malformation** with obstruction of CSF flow at foramen magnum
      - **Foramen magnus mass**
        - Meningioma
        - Periodontal pseudotumor
        - Posterior fossa arachnoid cyst
      - **Adhesive arachnoiditis**
        - Post trauma
        - Post subarachnoid hemorrhage
        - Post infection/meningitis
      - **Chronic meningitis**
        - Tuberculosis or coccidioidomycosis
      - **Recurrent syrinx**
        - Edema seen at advancing syrinx margin
      - **Cervical spondylosis** with central canal stenosis (rare)
      - **Postoperative complication**
        - Following posterior fossa decompression with posterior fossa hygroma (rare)
- Associated abnormalities
  - Progression from presyrinx to syrinx has been documented by serial imaging
  - Look for causes of CSF obstruction due to adhesions
    - CSF loculations and arachnoid cyst formation with adhesive arachnoiditis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be associated with nonspecific myelopathy, weakness
    - Paraparesis, numbness
  - May be asymptomatic until frank cavitation of lesion and syrinx formation

Treatment

- Surgical removal of site of CSF flow alteration or obstruction will quickly eliminate cord edema
- With more focal areas of arachnoid scarring, satisfactory long-term results seen with arachnoid dissection and decompression of subarachnoid space by fascia lata graft
- Extensive arachnoid scarring gives poor prognosis with high incidence of recurrence
  - Syringopleural or syringoperitoneal shunting most effective

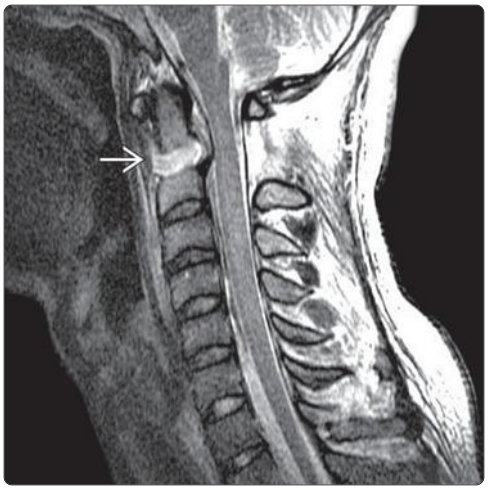
DIAGNOSTIC CHECKLIST

Consider

- Presyrinx is a cause of reversible spinal cord enlargement with T1 and T2 prolongation
  - Occurs in setting of CSF flow obstruction
  - May be misinterpreted as syringomyelia on MR studies
- Presyrinx edema likely represents a point on continuum to development of syringomyelia
  - May be pathophysiologically related to variations in patency of central canal
  - May be related to progressive post-traumatic myelomalacic myelopathy

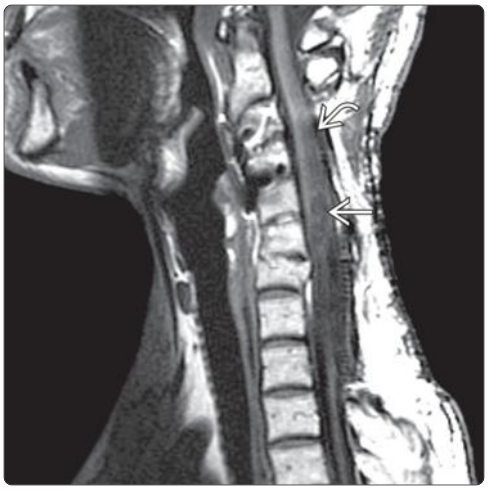
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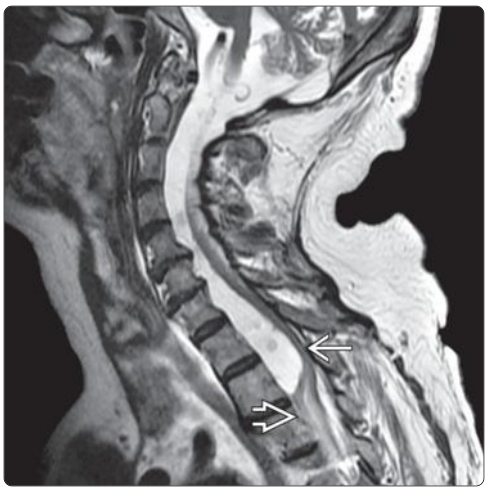
(Left) *Presyrinx cord edema developed due to post-traumatic arachnoiditis/adhesions. Initial MR study shows nonunion of type II odontoid fracture with wide separation of the fracture fragment. No cord signal abnormality is present.* **(Right)** *Presyrinx edema developed due to post-traumatic adhesions. Follow-up sagittal T2WI 2 years later, after occiput-C2 fusion, shows widening of the cervical cord with diffuse T2 hyperintensity. Focal area of ventral cord tethering is seen.*





(Left) *Sagittal T1WI MR status post flexion injury at C5-C6 shows the well-defined syrinx cavity at the C5-C6 level with cord expansion. Ill-defined low signal is seen at C3 and C4 level due to presyrinx edema.* **(Right)** *Sagittal T2WI shows increased signal within the cord extending cephalad from the syrinx to the C2 level due to presyrinx cord edema. This is the expected path of growth of the syrinx. Cephalad growth may be devastating if tenuous neurologic function is compromised.*





(Left) *Sagittal T2WI MR of Chiari 1 malformation with presyrinx edema shows marked cerebellar tonsillar ectopia with normal tectal plate and 4th ventricular position characteristic of Chiari 1 malformation. There is central intramedullary presyrinx edema in the cervical spinal cord without frank syringomyelia.* **(Right)** *Sagittal T2WI shows extensive arachnoiditis and adhesions following subarachnoid hemorrhage. Abnormal CSF flow dynamics and tethering has produced presyrinx edema.*

Spinal Cord Contusion-Hematoma

KEY FACTS

TERMINOLOGY

- Spinal cord injury (SCI)
- Traumatic axonal injury, cord edema, &/or hemorrhage

IMAGING

- Abnormal cord signal on MR in setting of trauma
- Most common level of adult SCI is C4-C6
- Commonly associated fracture or subluxation in younger adults (16-45 years)
- Underlying degenerative change (canal stenosis) predisposes to cord injury in older population
- SCI without radiographic abnormality (SCIWORA) is common in pediatric population (< 8 years)
- Cord injury typically occult on CT

PATHOLOGY

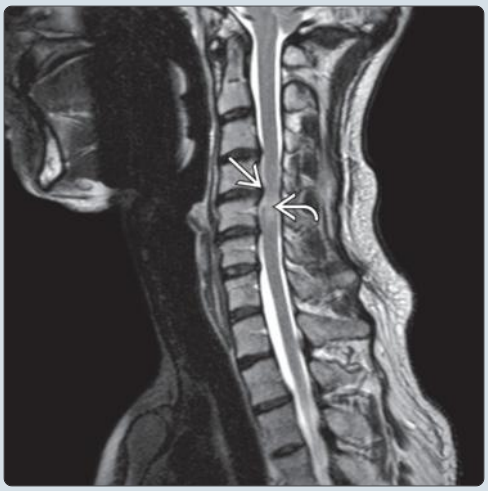
- Overall incidence of SCI in trauma estimated at 3.7%
- High-velocity mechanisms more common in youth and young adults
- If > 45 years, more likely due to fall; short falls (< 1 m) may result in significant injury in elderly

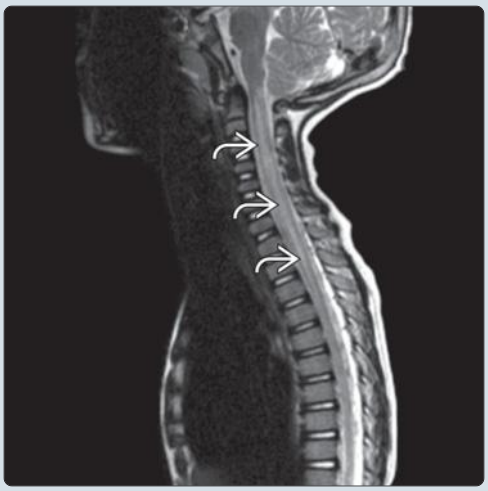
CLINICAL ISSUES

- Edema without hemorrhage: Good prognosis for recovery
- Hematoma: Poor prognosis, often without recovery
  - Extent of intramedullary hemorrhage and cord swelling are key predictors of neurologic recovery after traumatic cervical cord injury
- 30-60 new cases per million per year in USA

DIAGNOSTIC CHECKLIST

- Sagittal STIR is key sequence
  - Sensitive to cord edema
  - Ligamentous/muscular injury
  - Marrow edema
- Sagittal and axial gradient-echo images for cord hemorrhage





(Left) *Sagittal T2WI MR of a patient presenting days after a MVA with upper > lower extremity weakness shows congenital narrowing of the canal with multiple disc bulges and levels of canal stenosis, worst at C4-C5. Caudally, mild, patchy T2 hyperintensity shows nonhemorrhagic cord contusion.* **(Right)** *Sagittal T2WI MR in a child shows diffuse contusion in the cord from C1 through the upper thoracic cord. The patient presented with 4 extremity neurologic deficits after fall. Plain films would be normal (SCIWORA).*





(Left) *Sagittal T2 MR in this trauma patient shows low signal intensity cord hemorrhage at C4 with surrounding edema in the setting of severe cervical canal stenosis. Prevertebral edema is present.* **(Right)** *Sagittal T2WI shows a midthoracic burst fracture and disruption of ligamentum flavum. There is complete transection of the spinal cord, demonstrated by a CSF-filled cleft, with adjacent edema.*

TERMINOLOGY

Abbreviations

- Spinal cord injury (SCI)

Definitions

- Traumatic axonal injury, cord edema, &/or hemorrhage

IMAGING

General Features

- Best diagnostic clue
  - Abnormal cord signal on MR in setting of trauma
- Location
  - Most common level of adult SCI is C4-C6
    - Injury at higher levels in children due to larger heads, weaker musculature, and ligamentous laxity

Radiographic Findings

- Radiography
  - Commonly associated fracture or subluxation in younger adults (16-45 years)
  - Underlying degenerative change (canal stenosis) predisposes to cord injury in older population
  - SCI without radiographic abnormality (SCIWORA) is common in pediatric population (< 8 years)

CT Findings

- NECT
  - Predisposing factors: Spondylosis, spinal stenosis
  - Associated fracture, subluxation
  - Subdural/epidural hematoma
  - Cord injury typically occult on CT
    - Cord hematoma may be visible (hyperdensity)
- CTA
  - Quick screening for associated vascular injury (cervical spine)

MR Findings

- T1WI
  - Acute contusion
    - Iso- to hypointense
    - Cord swelling
  - Hematoma: Hyperintense after 3 days (methemoglobin)
  - Transection: Sagittal T1WI best to show cord discontinuity
  - Chronic contusion: Atrophy; ± cyst, syringomyelia
- T2WI
  - Acute contusion (edema): Hyperintense, distribution will increase over several days after injury, then plateau
  - Acute hematoma is hypointense
    - Look for traumatic disc herniation
  - Chronic hematoma: Hypointense due to hemosiderin deposition
    - Chronic contusion: Atrophy, hyperintense gliosis; ± cyst, syringomyelia
- T2* GRE
  - Hematoma: Hypointense (best for hemorrhage)
- MRA
  - Quick screening for associated vascular injury (cervical spine)

Nonvascular Interventions

- Myelography
  - Contusion: Focal cord enlargement effacing contrast
  - Hematoma: May mimic syrinx containing contrast
  - Transection: No cord visible in affected axial slice

Imaging Recommendations

- Best imaging tool
  - MR with gradient-echo images
- Protocol advice
  - Add STIR and GRE

DIFFERENTIAL DIAGNOSIS

Disc Herniation With Cord Compression

- Nontraumatic compressive myelopathy
- Severe spinal stenosis 2° to disc bulge/herniation, ligamentous hypertrophy, OPLL, etc.

Myelitis

- Demyelinating disease (e.g., MS), ADEM, transverse myelitis
- Patchy, hyperintense signal on T2WI, infrequently with fusiform enlargement
- All may enhance

Cavernous Malformation, Spinal Cord

- Best seen with GRE
- Hypointense due to susceptibility, may be multiple
- May be asymptomatic

Dural or Spinal Arteriovenous Fistula

- Flow voids posterior to cord: Dilated veins

Infarction, Spinal Cord

- Aortic dissection → conus infarction
- Vertebral dissection → cervical cord infarction (rare)

Syringomyelia

- Fusiform fluid within spinal cord

PATHOLOGY

General Features

- Etiology
  - Overall incidence of SCI in trauma estimated at 3.7%
  - MVA (45%), falls (22%), acts of violence (16%), sports injuries (13%)
    - High-velocity mechanisms more common in youth and young adults
    - If > 45 years, more likely due to fall; short falls (< 1 m) may result in significant injury in elderly
  - Spondylosis and canal stenosis both are significant risk factors for SCI

Staging, Grading, & Classification

- American Spinal Injury Association (ASIA)
  - Grade A: No motor/sensory function in sacral segments S4-S5
  - Grade B: No motor function below injury level
  - Grade C: > 50% key muscles below level < 3/5 strength
  - Grade D: > 50% key muscles below level > 3/5 strength
  - Grade E: Normal

- Neurologic level of injury = most caudal cord segment with normal sensory and motor function on both sides of body

Gross Pathologic & Surgical Features

- Ranges in severity
  - Cord edema without hemorrhage
  - Cord petechial hemorrhage
  - Cord pulverization and bleeding
  - Complete transection

Microscopic Features

- Cord microcirculation alterations maximize in 24-48 hours
- Intramedullary edema ± hemorrhage involving central gray matter
  - Extends outward with severity of trauma
  - Distribution of edema peaks in 3-6 days, then fades
  - Hemorrhage tends to be stable
- Hemorrhagic necrosis, liquefaction may occur
- Chronic: Mixed glial and fibrocytic scar, cavitation
  - Axonal degeneration both rostral and caudal to level of injury

CLINICAL ISSUES

Presentation

- Anterior cord syndrome
  - Injury to anterior 2/3 of cord
  - Motor, pain, and temperature affected
  - Posterior function spared (proprioception, light touch, vibration)
  - Highly associated with traumatic disc herniation; also seen with anterior spinal artery infarction
- Central cord syndrome
  - Motor deficit, predominantly affecting upper extremities
  - Bladder dysfunction
  - Varying degrees of sensory loss
  - Historically associated with central hematoma, predominantly a white matter injury
- Posterior cord syndrome: Uncommon; loss of dorsal column function
- Brown-Séquard syndrome
  - Injury to 1/2 of cord
  - Ipsilateral loss of touch and motor function
  - Contralateral loss of pain and temperature
  - Often superimposed on central cord syndrome
- Autonomic hyperreflexia
  - Arise in SCI above T6
  - Paroxysmal hypertension with agonizing headache
  - May result in intracerebral hemorrhage, seizure, cardiac arrhythmias, or death
- Contusion may be present in asymptomatic patients

Demographics

- Age
  - Mean age: 30.5 years; median age: 26 years
- Gender
  - > 80% male
- Epidemiology
  - 30-60 new cases per million per year in USA
  - Annual cost of SCI in USA estimated to be $9.73 billion in 1996

Natural History & Prognosis

- Contusion, no hemorrhage
  - Regresses over 1-2 weeks
  - Better prognosis for neurologic recovery
- Hematoma: Poor prognosis, often without recovery
  - Extent of intramedullary hemorrhage and cord swelling are key predictors of neurologic recovery after traumatic cervical cord injury
  - Poor prognostic factors: C4, C5, C6 level injuries, age > 50 years
- Anterior cord syndrome: 70% regain ambulation
- Central cord syndrome: < 50 years 70% ambulate, > 50 years 40%
- Posterior column syndrome: Ambulation usually possible unless proprioception involved, which then makes it difficult
- Brown-Séquard syndrome: 90% will ambulate
- Post-traumatic spinal cord syndrome (a.k.a. post-traumatic progressive myelopathy)
  - ≤ 2% post-traumatic SCI
  - Spinothalamic symptoms (i.e., pain, sensory loss)
  - Cysts may occur above or below original cord lesion

Treatment

- Surgical → stabilization, decompression
- Medical
  - Respiratory support
  - Spinal shock
    - Loss of sympathetic tone below injury level
    - Unopposed vagal tone → asystole → atropine
  - Neurogenic bladder: Intermittent catheter drainage to minimize potential complications
  - ↑ DVT risk due to immobilization
- Chronic care best approached from rehabilitation team approach

DIAGNOSTIC CHECKLIST

Consider

- Screening MR for all symptomatic SCI

Image Interpretation Pearls

- Sagittal STIR is key sequence

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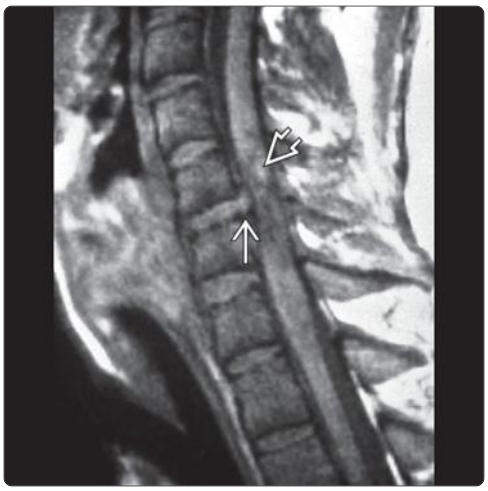


(Left) *Sagittal T2WI MR shows flexion injury with subluxation of C5 over C6 body and splaying of the neural arches. There is compromise of the canal diameter with compression and contusion of the cervical cord. Faint hypointensity marks the site of focal cord hemorrhage.* **(Right)** *Sagittal STIR MR shows congenital canal stenosis and a superimposed disc protrusion at C3-C4 compressing the cervical cord. The patient presented with upper extremity weakness after trauma. Note the site of focal cord contusion.*





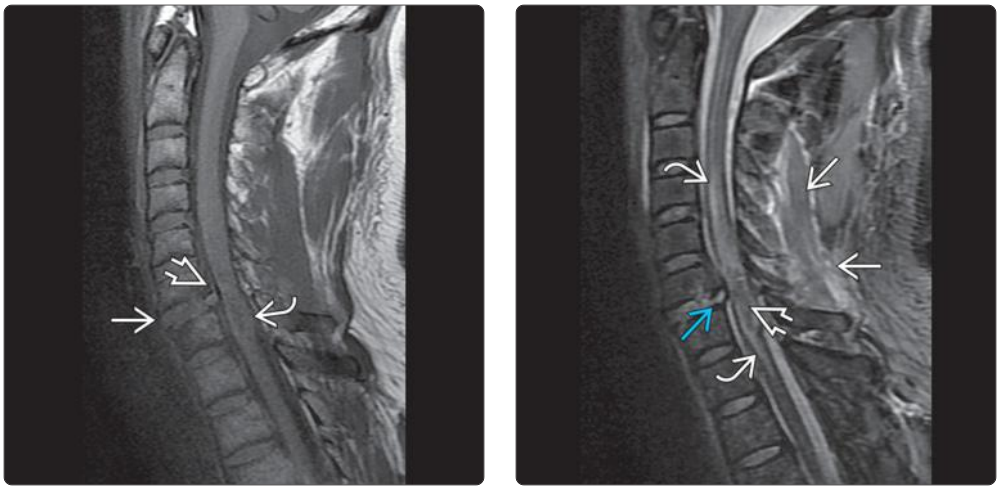
(Left) *Sagittal STIR MR shows extensive prevertebral edema in this patient with a known type II odontoid fracture. There is focal hemorrhage in the cord, seen as a faint area of hypointensity, surrounded by central cord edema rostrally and caudally.* **(Right)** *Sagittal T2* GRE MR performed on patient with C5 burst fracture is notable for extensive hypointensity within cord at C3-C4 through mid C6 level due to susceptibility from hematoma.*



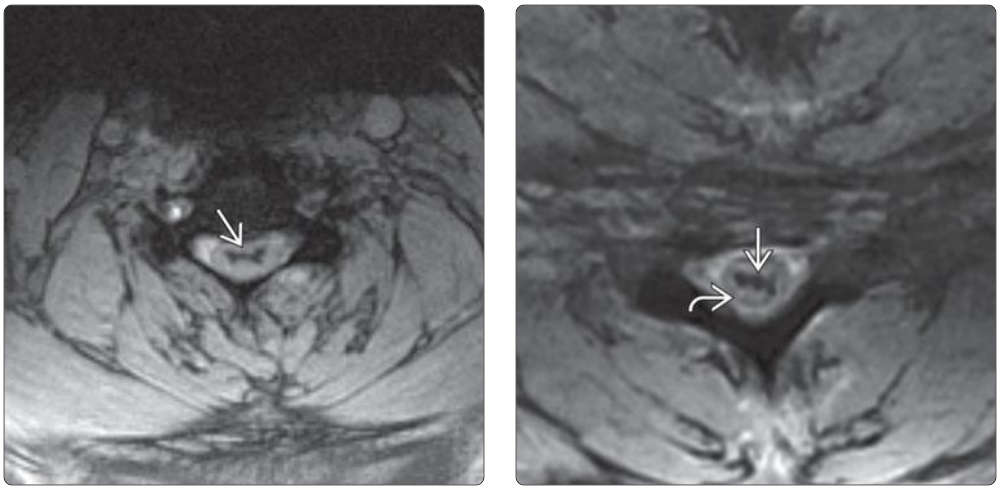


(Left) *Sagittal T1WI MR shows mild enlargement and a heterogeneously hypointense signal of the midcervical cord at the site of a traumatic disc herniation following hyperflexion injury. The area of hyperintensity relative to the normal cord demonstrates subacute cord hemorrhage.* **(Right)** *Sagittal T2WI MR shows transection of the cord at C6-C7 with sharply defined margins. The patient was tetraplegic. Note changes of spinal fixation with anterior plate and screws visible on this image.*

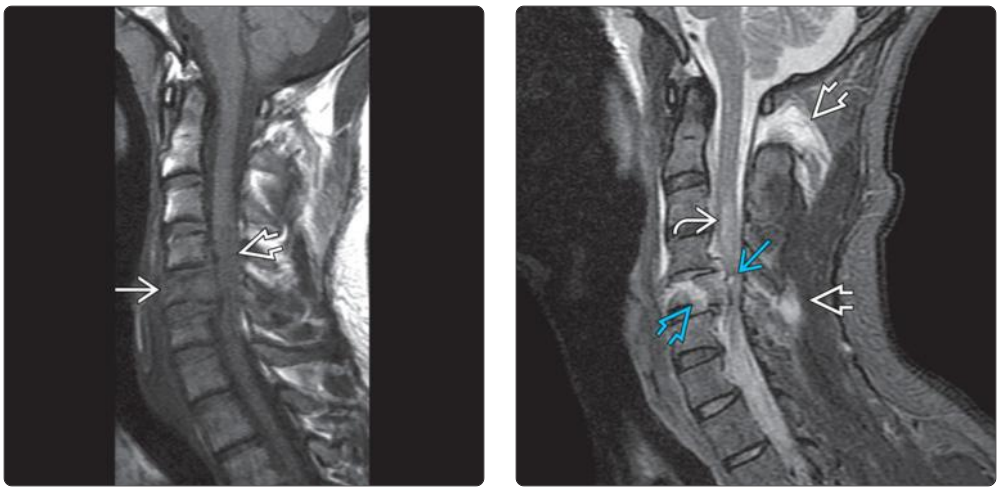
(Left) *Sagittal T1 MR shows marrow edema and subluxation at the C6-C7 fracture dislocation site, with post-traumatic disc herniation. The cord is expanded and low in signal.* **(Right)** *Sagittal STIR MR shows low signal cord hemorrhage with adjacent high signal cord edema. There is extensive posterior soft tissue edema. Traumatic disc herniation is also present.*

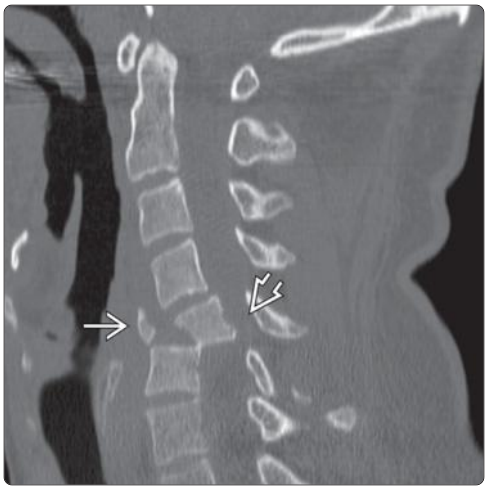


(Left) *Axial T2* GRE MR shows central very low signal within the cervical spinal cord in this trauma patient, reflecting cord hemorrhage. This gives a poor prognosis for functional recovery.* **(Right)** *Axial T2* GRE MR shows extensive bilateral cord hemorrhage as low signal with surrounding abnormal increased signal from edema.*



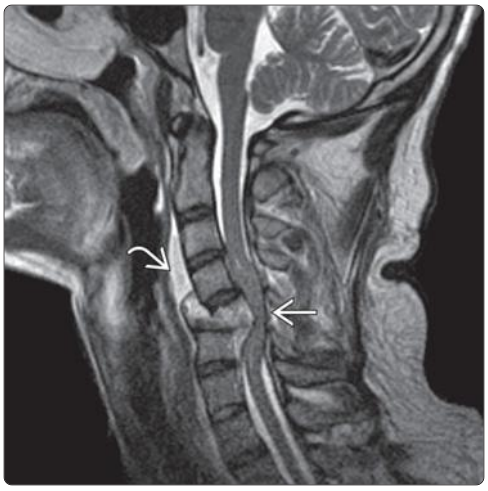
(Left) *Sagittal T1 MR in this patient with C5 burst fracture shows marrow edema from the poorly defined fracture and fusiform expansion of the cord with extensive edema.* **(Right)** *Sagittal STIR MR in this patient with C5 burst fracture shows fusiform expansion of the cord with extensive edema and low signal hemorrhage. The fracture site is well defined due to the comminution and displacement. Posterior ligamentous injury is also present.*







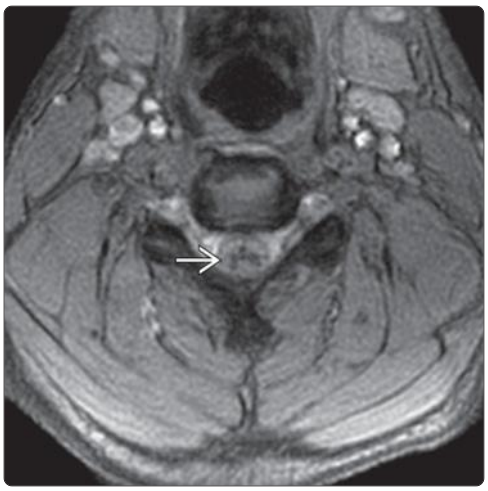
(Left) *Initial sagittal NECT (bone window) shows severe burst flexion fracture at the C5 level with dislocation ⇨. There is severe canal compromise ⇨.* **(Right)** *Axial CT (bone window) defines the severity of the vertebral body and posterior element comminution, with facet displacement ⇨. Note the marked narrowing of the spinal canal from the fragments and dislocation ⇨.*





(Left) *Sagittal T2 MR shows the burst fracture with severe cord compression ⇨ with very slight cord edema. Extensive high signal prevertebral edema is seen ⇨. The patient had a complete injury at presentation, and the T2 MR does not show the extensive nature of the cord injury.* **(Right)** *Postoperative sagittal T2 MR with canal decompression (C5 corpectomy) better shows the true nature of the cord injury. Expansion of the canal has allowed blooming of cord edema & hemorrhage & shows the focal cord disruption ⇨.*





(Left) *Postoperative sagittal T2* GRE MR shows a metal artifact from the anterior corpectomy with plate and screw fixation ⇨ and low signal hemorrhage in the cord ⇨.* **(Right)** *Axial T2* GRE MR shows low signal intensity hemorrhage within the cervical cord ⇨ in this patient with a prior C5 burst fracture dislocation.*

Idiopathic Spinal Cord Herniation

KEY FACTS

TERMINOLOGY

- Ventral cord herniation
- Herniation of spinal cord through defect in dura of ventral canal

IMAGING

- Best diagnostic clue: Focal anterior displacement of cord with expansion of dorsal subarachnoid space
- Location: Typically in midthoracic spine
  - T2-T8 level
- Focal cord deformity
- Cord displaced anteriorly against posterior edge of vertebral body
- Increased dorsal subarachnoid sac
- May see secondary collection of contrast in extradural sac
- Best imaging tool: CT postmyelography

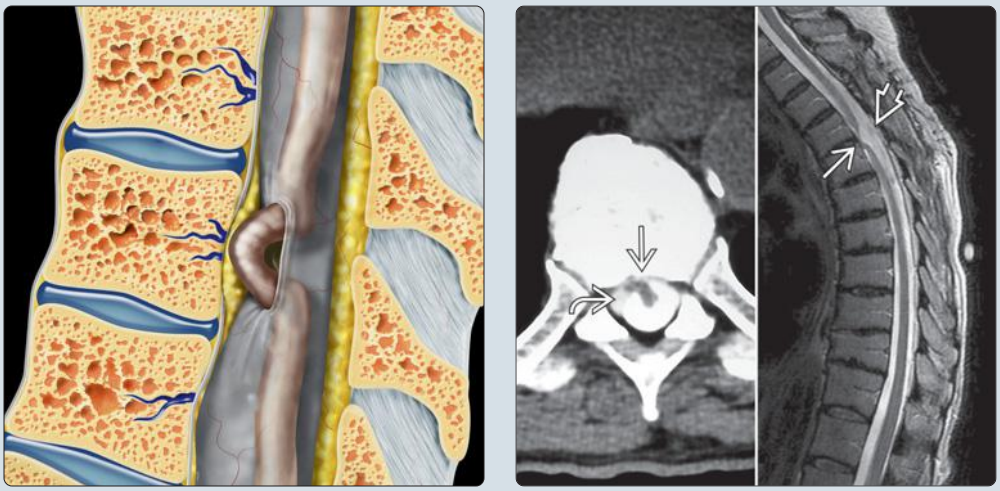
TOP DIFFERENTIAL DIAGNOSES

- Arachnoid cyst
- Epidermoid cyst
  - Restricted diffusion
- Adhesions
- Cystic schwannoma
  - Peripheral or nodular enhancement
- Epidural hematoma
- Epidural empyema

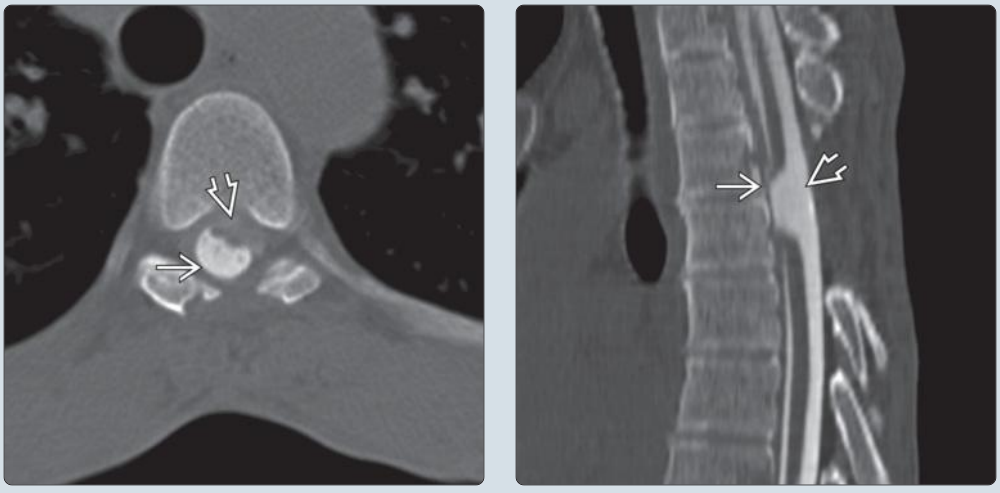
PATHOLOGY

- Defect or diverticulum in ventral dural sheet into which cord herniates
- Several proposed mechanisms
  - Congenital weakening of ventral dural fibers
  - Damage to ventral dura by disc herniation or other mechanism
  - Abnormal adhesion of cord to anterior dural sleeve progressively wears down dura leading to herniation

(Left) *Sagittal graphic shows focal dural defect in the thoracic spine allowing cord herniation. Note the distinctive and focal cord kink.* **(Right)** *Axial CECT myelogram (L) and sagittal T2WI MR (R) show ventral cord distortion and anterior displacement into the extradural cavity ➡ with focal enlargement of dorsal CSF ➡. Note the extradural CSF collection with slightly less density from delayed leakage of contrast ➡.*



(Left) *Axial CECT (postmyelography) shows an expanded dorsal subarachnoid space ➡ with a distorted cord displaced anteriorly against the posterior aspect of vertebral body ➡.* **(Right)** *Sagittal CECT (postmyelography) shows a focal kink of the thoracic cord ➡ and anterior adhesion to the back of the vertebral body. The expanded dorsal thecal sac mimics an arachnoid cyst ➡.*



TERMINOLOGY

Synonyms

- Ventral cord herniation

Definitions

- Herniation of spinal cord through defect in dura of ventral canal

IMAGING

General Features

- Best diagnostic clue
  - Focal anterior displacement of cord with expansion of dorsal subarachnoid space
- Location
  - Typically in midthoracic spine
    - T2-T8 level
    - Ventral dura involved
  - Typically at or near intervertebral disc level
- Morphology
  - S-shaped or kinked, thinned cord

MR Findings

- T1WI
  - Focal cord deformity
  - Cord displaced anteriorly against posterior edge of vertebral body
  - Increased dorsal subarachnoid sac
- T2WI
  - Focal increase in dorsal CSF space at level of herniation
  - Focal anterior cord displacement
  - Kink or S-shaped cord
- T2* GRE
  - Same as T2WI
- T1WI C+
  - Excludes extramedullary mass

Nonvascular Interventions

- Myelography
  - Focal deformity of cord and subarachnoid space
  - May be associated with adjacent extradural fluid collection

Other Modality Findings

- CT with intrathecal contrast (postmyelography)
  - Anterior displacement of cord
    - Axial images show cord ventrally displaced into dural defect
    - Portion of distorted cord may extend outside of dural margin
  - Focal cord deformity at level of displacement
  - Expanded dorsal subarachnoid space
  - May see secondary collection of contrast in extradural sac
    - Delayed scan to see filling of extradural sac
  - MR thin-section to define cord deformity and exclude other masses

Imaging Recommendations

- Best imaging tool
  - CT postmyelography or MR
- Protocol advice
  - CT thin-section slices
    - Sagittal reformations

DIFFERENTIAL DIAGNOSIS

Arachnoid Cyst

- Can produce identical findings if dorsal
- Usually lacks abrupt cord deformity
- Can accompany herniation (due to adhesions)

Epidermoid Cyst

- Variable T1 and T2 signal due to cyst protein content
- Dorsal nonenhancing mass with ventral cord displacement
- Restricted diffusion

Adhesions

- Simulates appearance of arachnoid cyst
- Cord adhesions usually present with herniation

Epidural Hematoma

- Can produce focal cord displacement
- If subacute, will show high signal on T1WI
- Usually more diffuse

Epidural Empyema

- Usually more extensive and not localized to disc level
- T1WI shows enhancement
- Associated disc space infection

Cystic Schwannoma

- Dorsal mass with ventral cord displacement
- Peripheral or nodular enhancement
- No restricted diffusion

PATHOLOGY

General Features

- Etiology
  - Several proposed mechanisms
    - Congenital weakening of ventral dural fibers
    - Damage to ventral dura by disc herniation or other mechanism
      - Herniated thoracic disc or osteophyte pierces, lacerates, and progressively erodes anterior dura
    - Abnormal adhesion of cord to anterior dural sleeve progressively wears down dura leading to herniation
    - CSF pulsations weaken dura over time
- Epidemiology
  - Rare
  - Literature consists of many case reports and small series (< 10 patient)
- General path comments
  - Defect or diverticulum in ventral dural sheet into which cord herniates

Gross Pathologic & Surgical Features

- Ventral dural defect or diverticulum
- Cord herniated into defect
- Adhesions
- May see arachnoid cyst

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic leg pain
  - Leg weakness
  - Other signs/symptoms
    - Brown-Séquard syndrome
      - Ipsilateral upper motor neuron paralysis with loss of proprioception and contralateral loss of pain and temperature sensation
    - Spasticity
    - Sphincter dysfunction
- Clinical profile
  - Unexplained chronic, progressive leg pain with onset of myelopathy

Demographics

- Age
  - Most reported cases in middle-aged individuals
  - Wide age range of 20-80, with mean of 50
- Gender
  - F > M
- Ethnicity
  - All

Natural History & Prognosis

- Progressive neurologic symptoms and signs until diagnosis made

Treatment

- Options, risks, complications
  - Surgery
    - Repair of dural defect
    - Repositioning of cord and reduction of herniated segment
    - Excision of arachnoid cyst if present
    - Enlargement of dural defect with surgery, freeing cord also successful
- Symptoms improve or resolve after successful surgery
  - 88% improve
  - 24% stable
  - 9% worsened

DIAGNOSTIC CHECKLIST

Consider

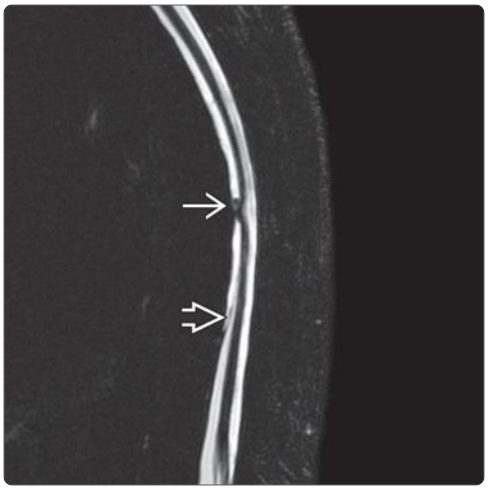
- Any focal cord displacement or subarachnoid space deformity as possible herniation
- Rare, but important to recognize size represents curable cause of myelopathy

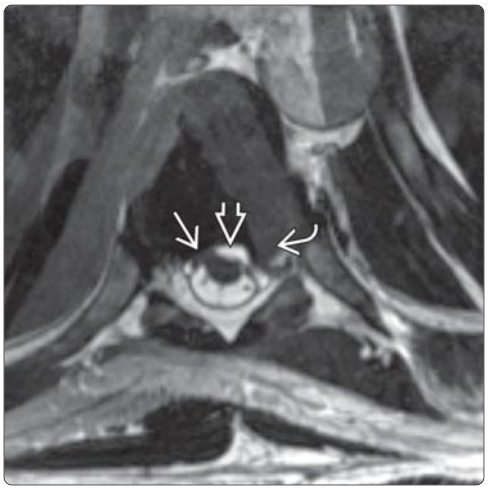
Image Interpretation Pearls

- Delayed filling of extradural fluid collection on CT after myelography with focal filling defect contiguous with cord = herniation

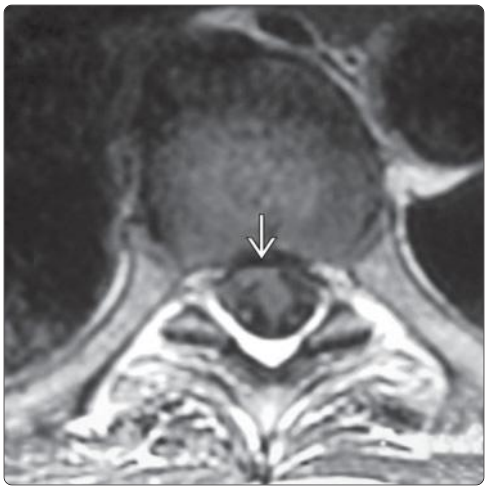
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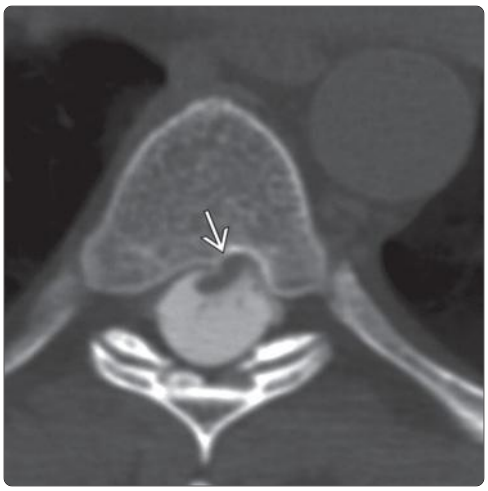
(Left) *Sagittal heavily T2-weighted MR myelography shows focal distortion and kinking of the thoracic cord ⇨. Note the displaced dural margin indicating associated extradural fluid collection related to the dural defect ⇨.* **(Right)** *Axial heavily T2-weighted CISS MR shows the site of cord herniation through the dura ⇨, as well as adjacent extradural fluid collection along the ventral thecal sac ⇨. Note the associated disc herniation ⇝.*





(Left) *Axial T1WI MR shows a distorted, anteriorly displaced cord ⇨. Surgery confirmed the dural defect in this patient with slowly progressive leg weakness.* **(Right)** *Sagittal T2 MR shows ventral focal distortion and displacement of the cord abutting the posterior vertebral body margin ⇨. Differential considerations include posterior arachnoid cyst cord herniation.*





(Left) *Sagittal T1 MR shows the nearly pathognomonic appearance of idiopathic transdural cord herniation affecting the upper thoracic spine with focal angulation of the cord ⇨ ventrally into the dural defect.* **(Right)** *Axial CT myelogram defines the ventral and left displacement of the cord adjacent to the posterior margin of the vertebral body ⇨ in this typical example of cord herniation.*

Central Spinal Cord Syndrome

KEY FACTS

TERMINOLOGY

- Central cord syndrome (CCS), acute traumatic central cord syndrome (ATCCS)
- Most common partial cord syndrome
- Traumatic cord injury, with clinical syndrome of
  - Bilateral weakness, arms > legs
  - Bladder dysfunction, variable sensory loss
- Injury to central gray matter, corticospinal and spinothalamic tracts

IMAGING

- Radiographs may show spondylosis or congenitally diminished canal diameter
- MR shows hyperintense signal in central spinal cord on T2WI
- Significant percentage show fracture/dislocations

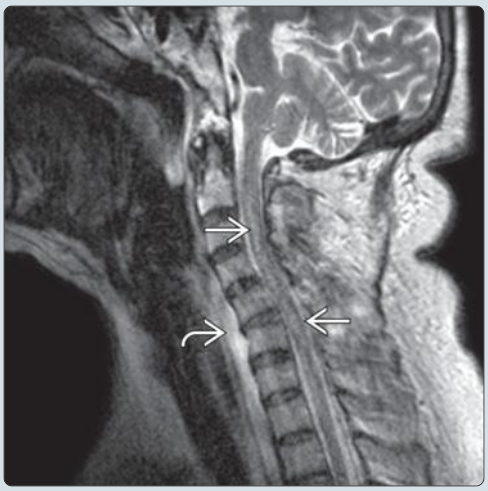
PATHOLOGY

- Often with preexisting canal stenosis
  - Congenital canal stenosis
  - Disc herniation, osteophyte
  - Facet/ligamentous hypertrophy

CLINICAL ISSUES

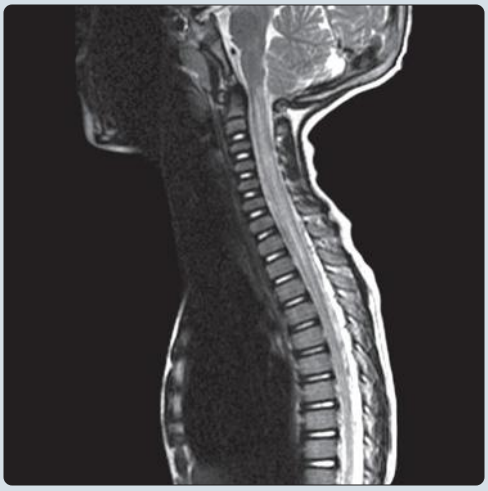
- **4 groups**
  - 10% of ATCCS patients have MR evidence of cord signal change with no other radiographic abnormality
    - Medical management
  - 20% of ATCCS patients present with acute disc herniation
    - Surgical intervention recommended
  - 30% of ATCCS patients have cervical spine fracture subluxation injuries
    - Early realignment of spinal column with spinal cord decompression is recommended
  - 40% of ATCCS patients have spinal stenosis without evidence of bony or ligamentous injury
    - Management remains controversial





(Left) *Sagittal reformatted spine CT shows focal flexion deformity at C5-C6 with splaying of the posterior elements, demonstrating a hyperflexion injury in this trauma patient with marked upper extremity weakness.* **(Right)** *Sagittal T2WI of the same patient again shows the focal flexion deformity at C5-C6. There is extensive moderately hyperintense signal in the cord from C2-C3 through C6-C7 demonstrating contusion within the spinal cord. Also note edema within prevertebral soft tissues.*





(Left) *Lateral radiograph in this injured child presenting with central cord syndrome shows borderline thickening of the prevertebral soft tissues but is otherwise unremarkable.* **(Right)** *Sagittal T2WI MR in the same child shows a normal vertebral column and prevertebral soft tissues (seen better on other sequences). There is extensive hyperintense signal within the cord from the cervicomedullary junction through T4-T5, demonstrating spinal cord contusion.*

TERMINOLOGY

Abbreviations

- Central cord syndrome (CCS), acute traumatic central cord syndrome (ATCCS)

Synonyms

- Partial cord syndrome (subset of)

Definitions

- Most common partial cord syndrome
- Traumatic cord injury, with clinical syndrome of
  - Bilateral weakness, arms > legs
  - Bladder dysfunction
  - Variable sensory loss
- Injury to central gray matter, corticospinal and spinothalamic tracts

IMAGING

General Features

- Best diagnostic clue
  - Hyperintense signal in central spinal cord on T2WI
- Location
  - Predominates at C3-C4 through C5-C6 levels

Radiographic Findings

- Radiography
  - Often negative for acute injury
  - May show spondylosis or congenitally diminished canal diameter

CT Findings

- NECT
  - May show
    - Disc herniation
    - Ligamentous hypertrophy
    - Congenitally small spinal canal

MR Findings

- T1WI
  - May show enlargement of cord due to edema
  - Canal stenosis
- T2WI
  - Hyperintense signal at level of injury
  - May show enlargement of cord due to edema
  - Canal stenosis
- T2* GRE
  - High intraparenchymal cord signal
  - Will exaggerate canal stenosis
  - Loss of CSF signal (narrow canal &/or swollen cord)
  - Rare: Low signal indicates hemorrhage and predicts permanent injury
    - If hemorrhage present, usually **not** a central cord syndrome

Nonvascular Interventions

- Myelography
  - Done only if no MR available
  - Mild cord enlargement
  - Predisposing canal stenosis

Imaging Recommendations

- MR in any case of post-traumatic cord signs is mandatory

DIFFERENTIAL DIAGNOSIS

Myelitis

- Demyelinating disease (e.g., MS), ADEM, transverse myelitis
- Subacute evolution rather than immediate traumatic onset

Cavernous Malformation, Spinal Cord

- Best seen with GRE
- May be multiple
- May be incidental finding in trauma that simulates cord injury

Syringomyelia

- Fusiform fluid within spinal cord
- May also have upper extremity weakness but is chronic in presentation

Infarction, Spinal Cord

- Secondary to aortic surgery, nontraumatic aortic dissection, atherosclerotic disease, hypotension/hypoperfusion, vasculitis
- Subset of infarctions are traumatic, due to
  - Traumatic aortic dissection → conus infarction
  - Vertebral artery dissection → cervical cord infarction (rare)

PATHOLOGY

General Features

- Etiology
  - Often with preexisting canal stenosis
    - Congenital canal stenosis
    - Disc herniation, osteophyte
    - Facet/ligamentous hypertrophy
  - Hyperextension
    - Buckling of redundant ligamentum flavum causes compression of cord and damages central gray matter
  - Axonal stretching

Staging, Grading, & Classification

- American Spinal Injury Association (ASIA)
  - Grade A: No motor/sensory function in sacral segments S4-S5
  - Grade B: No motor function below injury level
  - Grade C: > 50% key muscles below level < 3/5 strength
  - Grade D: > 50% key muscles below level > 3/5 strength
  - Grade E: Normal
  - Neurologic level of injury = most caudal cord segment with normal sensory & motor function on both sides of body

Gross Pathologic & Surgical Features

- Cord compression, swelling

Microscopic Features

- Primary injury to lateral corticospinal tract with loss of axons; predominantly white matter injury
- Cord hemorrhage does not have to be present
- Central gray matter intact in acute and reversible cases
- Wallerian degeneration distal to epicenter of injury

- Minimal degeneration within ventral corticospinal tract
- Neuronal loss in regions supplying hand musculature in cases of chronic injury

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Heterogeneous phenomenon
    - Minority of cases show classic hyperextension injury superimposed on spinal stenosis
    - 60% of ATCCS patients have fracture subluxations, acute disc herniation and rarely SCIWORA
    - Minimal form may result in symptoms such as "burning hands," but with normal neurological examination (contusion of spinothalamic tracts)
    - Potential for spontaneous recovery of function irrespective of the treatment provided
  - Bilateral weakness, arms > legs
    - Bladder dysfunction
    - Variable sensory loss
  - Upper extremity American Spinal Injury Association (ASIA) motor score should be at least 10 points less than lower extremities ASIA Motor Score
  - Exclude other causes of post-traumatic upper extremity weakness
    - Bilateral brachial plexus injury
    - Bell cruciate paralysis
- Clinical profile
  - Post-traumatic cord signs in patient with underlying congenital or acquired spinal stenosis
    - Bilateral weakness, arms > legs
    - High-energy mechanisms of injury more common for younger patients
    - Older patients have high incidence of injury sustained from fall

Demographics

- Ethnicity
  - Asians have higher incidence of ossified posterior longitudinal ligament as predisposing factor
- Epidemiology
  - 1 study estimated that 1.3 of every 10,000 USA football players sustained central cord syndrome
  - Up to 25% of traumatic paralysis associated with restricted canal

Natural History & Prognosis

- Considerable spontaneous recovery, from complete resolution to slight residual weakness and spasticity
- Generally complete or near-complete reversal of acute paralysis
- Prognosis is age related
  - Most under age of 50 recover completely
  - Those over age of 70 usually have significant residual deficits
- Severe cord compression, extensive edema, hemorrhage on MR indicate worse prognosis, poor recovery

Treatment

- In general, conservative therapy produces equal outcomes to surgical intervention
  - Intensive care unit management of patients recommended (level III evidence)
  - Cardiac, hemodynamic, respiratory monitoring, and maintenance of mean arterial blood pressure at 85-90 mm Hg for the 1st week after injury to improve spinal cord perfusion recommended (level III evidence)
  - Early reduction of fracture-dislocation injuries is recommended
- **4 groups**
  - 10% of ATCCS patients have MR evidence of cord signal change with no other radiographic abnormality
    - Medical management
  - 20% of ATCCS patients present with acute disc herniation
    - Surgical intervention recommended
  - 30% of ATCCS patients have cervical spine fracture subluxation injuries
    - Early realignment of the spinal column with spinal cord decompression is recommended
  - 40% of ATCCS patients have spinal stenosis without evidence of bony or ligamentous injury
    - Management remains controversial

DIAGNOSTIC CHECKLIST

Consider

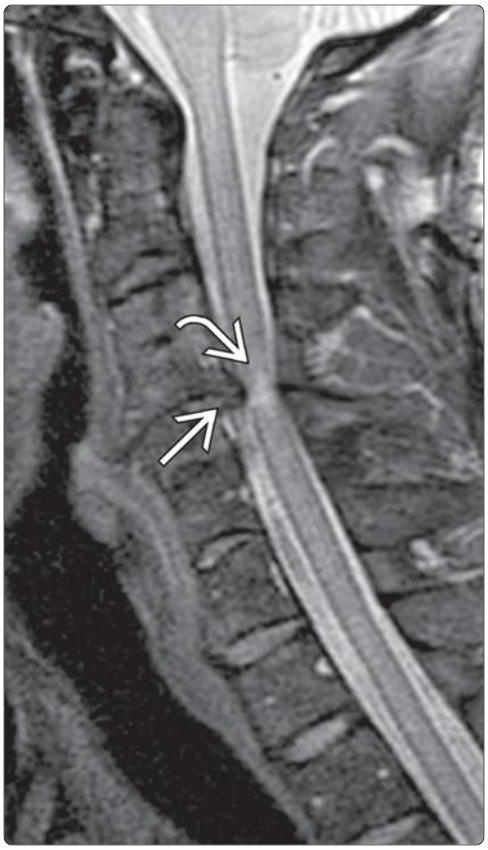
- Any cord signs after trauma are indication for MR

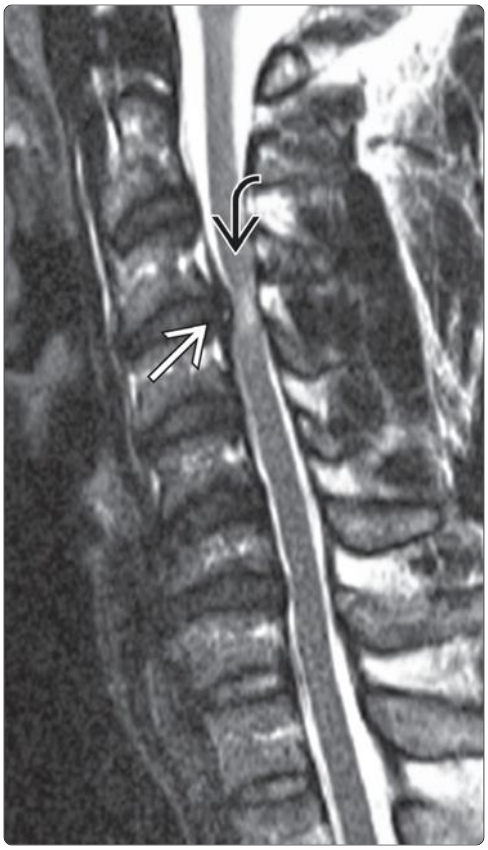
Image Interpretation Pearls

- While initially described with minimal imaging findings, current knowledge shows that significant percentage of ATCCS patients show fractures/dislocations

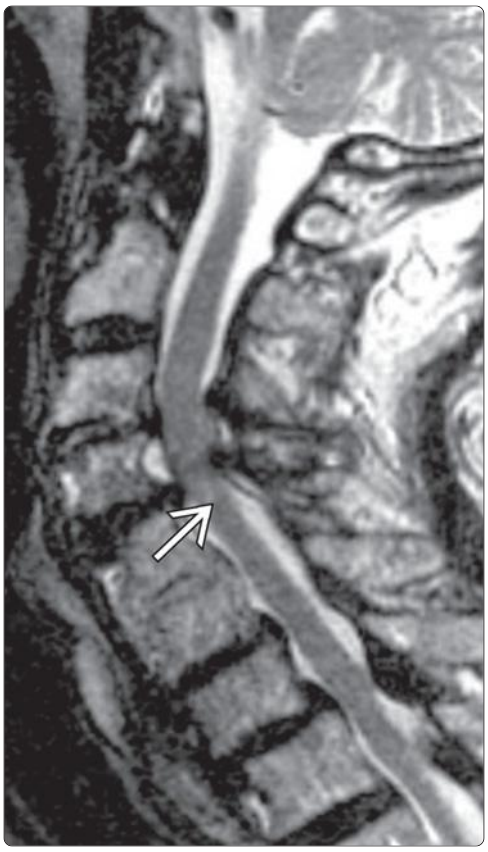
SELECTED REFERENCES

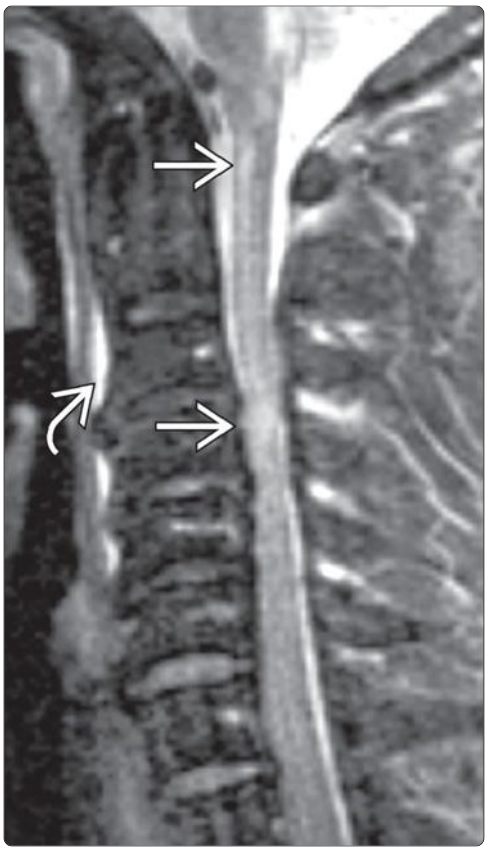
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(Left) *Sagittal T2WI shows focal hyperintense signal in the cord at C4-C5, demonstrating a contusion in this trauma patient with central cord syndrome. Note the preexisting canal stenosis due to the large endplate osteophyte at this level.* **(Right)** *Sagittal T2WI MR shows hyperintense signal in the cord at C3-C4. In addition to congenital canal stenosis, there is a superimposed C3-C4 disc protrusion, which leads to significant narrowing of the cervical canal.*





(Left) *Sagittal T2WI MR shows multilevel degenerative change in the cervical spine. At both C3-C4 and C4-C5 there is significant canal stenosis due to ventral disc bulge-endplate osteophyte complexes and dorsal ligamentous hypertrophy. This trauma patient presented clinically with central cord syndrome, and imaging shows a small focus of hyperintense signal at C4-C5 at the site of cord injury.* **(Right)** *Sagittal STIR MR of this trauma patient shows congenital canal stenosis with minimal superimposed degenerative change. Abnormally hyperintense T2 signal is present in the cord at the cervicomedullary junction and at C4, demonstrating sites of cord injury. Note hyperintense signal in anterior tissues, showing soft tissue signal changes from hyperextension injury.*

Traumatic Dural Tear

KEY FACTS

TERMINOLOGY

- Acquired dural defect, with CSF leak ± pseudomeningocele

IMAGING

- Most leaks not identified preoperatively, but rather identified at time of surgery with CSF leaking around injured segments
  - May see epidural or paravertebral fluid collection
  - Fluid can infiltrate surrounding tissues or become encapsulated, forming a circumscribed collection (pseudomeningocele)
    - Longstanding pseudomeningocele can be associated with osseous remodeling
- Look for associated severe spine trauma (e.g., burst fracture)
- CT myelography, radionuclide myelography
  - Useful to identify site of tear to guide surgical exploration or blood patch placement
- Imaging of brain may show signs of intracranial hypotension in chronic cases
  - "Sagging" midbrain
  - Pachymeningeal thickening, enhancement

PATHOLOGY

- Causative injuries
  - Burst fracture with laceration of dura
  - Nerve root avulsion with dural tear
  - Penetrating injury
- Dural defect results in collection of CSF outside of meninges

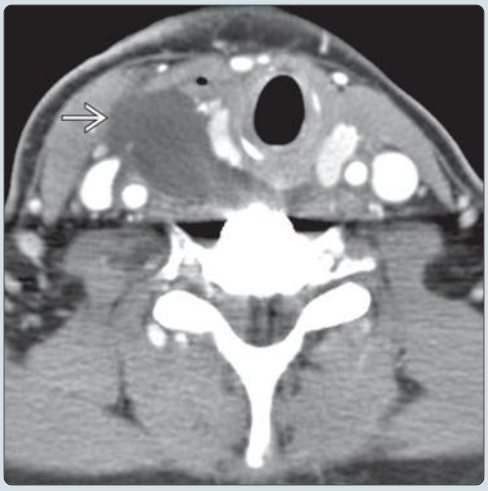
CLINICAL ISSUES

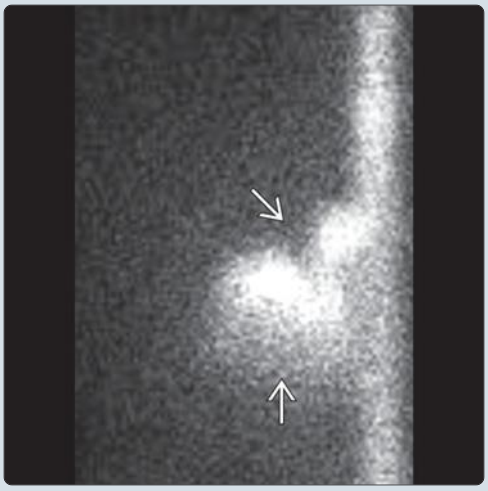
- Back pain
- Localized soft tissue swelling
- Syndrome of intracranial hypotension
  - Persistent, postural headache
  - Infrequent: Cranial neuropathies, visual disturbance





(Left) *Sagittal STIR shows comminuted burst fracture of L5 with large posterior pseudomeningocele secondary to dural tear and CSF leak. Traumatic spondylolisthesis also associated with severe effacement of the thecal sac at the injured level.* **(Right)** *Sagittal T2WI MR shows type I dens fracture and large ventral epidural fluid collection resulting in extensive effacement of the thecal sac with dorsal displacement and mild compression of the cervical cord.*





(Left) *Axial CECT of the neck in this patient recently status post ACDF shows a large pseudomeningocele extending from the discectomy site into the prevertebral space and then medial to the carotid and jugular vein.* **(Right)** *Nuclear myelography of this patient with symptoms of intracranial hypotension shows a large amount of radionuclide spilling from the thoracic thecal sac into the right chest. The patient was recently status post thoracotomy for resection of a pulmonary malignancy.*

TERMINOLOGY

Synonyms

- Traumatic meningocele
- Pseudomeningocele

Definitions

- Traumatically acquired dural defect, with CSF leak ± pseudomeningocele

IMAGING

General Features

- Location
  - Most leaks not identified preoperatively, but rather identified at time of surgery with CSF leaking around injured segments
    - May see epidural or paravertebral fluid collection
    - If paravertebral, communicates through neural foramen or through vertebral defect
- Morphology
  - Fluid can infiltrate surrounding tissues or become encapsulated, forming circumscribed collection

Radiographic Findings

- Radiography
  - Evidence of associated spinal trauma (e.g., burst fracture)

CT Findings

- Bone CT
  - Epidural or paravertebral fluid collection
  - May see vertebral fracture
  - Chronically, may result in osseous remodeling, foraminal widening
  - CT myelography
    - May see extravasation of contrast at site of dural defect
    - Useful to identify site of tear to guide surgical repair or blood patch placement
    - In nerve root avulsion
      - Rounded paraforaminal collection of contrast
      - Absent nerve root

MR Findings

- T1WI
  - Fluid hypointense on T1
  - Paravertebral and epidural fat provides natural contrast to define extent of collection
  - Acute setting: May see marrow edema associated with vertebral fracture, hypointense on T1WI
- T2WI
  - Fluid collection hyperintense on T2WI
  - Signs of intracranial hypotension on brain MR
    - Pachymeningeal thickening
    - "Sagging" midbrain
    - Cerebellar tonsillar herniation
- STIR
  - Fluid hyperintense on STIR
  - Acute setting: May see marrow edema associated with vertebral fracture, hyperintense on STIR
- T1WI C+
  - Essentially no enhancement associated with pseudomeningocele
  - Signs of intracranial hypotension on brain MR
    - Smoothly thickening, enhancing dura
    - Enlarged enhancing pituitary
      - Due to venous engorgement
  - MR myelography
    - Additional technique for identifying CSF leaks

Imaging Recommendations

- Protocol advice
  - CT most useful to define anatomy in setting of vertebral fracture
    - Thin-section (1-2 mm) cuts
    - Sagittal and coronal reformatted images
  - Low threshold for performing CT myelography
    - Intrathecal contrast useful to demonstrate exact site of dural tear
  - MR preferred to show fluid collection and to delineate neural structures

Nuclear Medicine Findings

- Radionuclide myelography
  - Useful to identify site of tear to guide blood patch placement

DIFFERENTIAL DIAGNOSIS

CSF Leakage Syndrome

- Includes all etiologies of decreased CSF volume/pressure
- Clinical syndrome of
  - Severe, persistent postural headaches
  - Low CSF pressure on lumbar puncture

Hematoma, Epidural, Traumatic

- Epidural collection of blood
- Contents may be similar to CSF on some pulse sequences at certain points in time
- Contents will not follow CSF on all sequences

Abscess, Epidural, Paravertebral

- Seen with spondylodiscitis, direct inoculation of epidural space
- Marked enhancement typical
  - Peripheral = abscess
  - Diffuse = phlegmon

PATHOLOGY

General Features

- Etiology
  - **Lumbar spine**
    - Burst fracture
      - Axial loading injury, vertically oriented fracture plane, centrifugal displacement of fragments
      - Laceration of ventral dura by osseous fragments
    - Fracture dislocation
  - **Thoracic spine**
    - Burst fractures and fracture dislocations are most common cause
  - **Cervical spine**

  - Most common underlying trauma is atlantooccipital dislocation (AOD), flexion compression or flexion distraction mechanisms
  - Nerve root avulsion
    - Traction on nerve root (due to lateral flexion of cervical spine, for example)
    - Nerve root avulsion and tear of nerve root sleeve
- **Other**
  - Penetrating injury
    - Knife
    - Projectile
    - Surgery
- Associated abnormalities
  - Vertebral fracture
  - Nerve root avulsion
  - Evidence of penetrating injury
  - History/signs of recent surgery
  - Nerve root or spinal cord herniating through dural defect
  - Nerve root entrapped in fracture
    - Particularly with burst fracture & disruption of neural arch

Gross Pathologic & Surgical Features

- Rent in dura

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Localized soft tissue swelling
  - Back pain
  - Persistent, postural headache in chronic cases
- Other signs/symptoms
  - Infrequent: Cranial neuropathies, visual disturbance

Demographics

- Epidemiology
  - In 1 series of operatively treated unstable spine fractures
    - Cervical tears in 9%
    - Thoracic tears in 10%
    - Lumbar tears in 18%
  - High rate of neurologic injury in the setting of traumatic dural tears
    - Complete neurologic injury (ASIA A) was present in ~ 50%

Natural History & Prognosis

- Presence of traumatic dural tear associated with high incidence of neurological injury
  - Prognosis depends on primary injury type
- Traumatic dural tear overall negative effect on the patient's postoperative course is minor
  - Similar to iatrogenically created dural tears in elective spine surgery
  - May lead to persistent CSF leak or pseudomeningocele formation

Treatment

- Blood patch
- Temporary subarachnoid drain
- Primary surgical repair
  - Surgical fixation of any unstable vertebral column injury
- There is no consistent algorithm regarding the management of dural tears
  - Wide variety of repair techniques and agents utilized

DIAGNOSTIC CHECKLIST

Consider

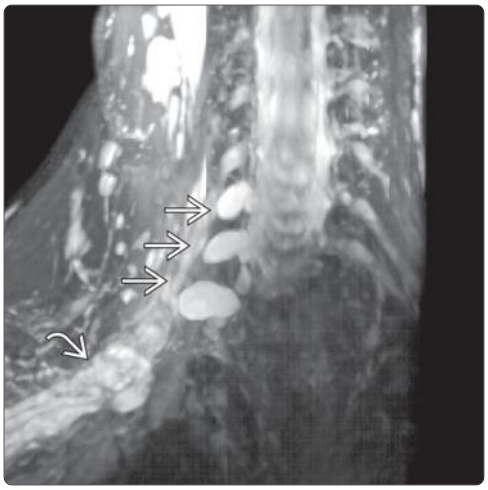
- CT myelography for evaluating any patient with neurological signs and burst fracture prior to surgery

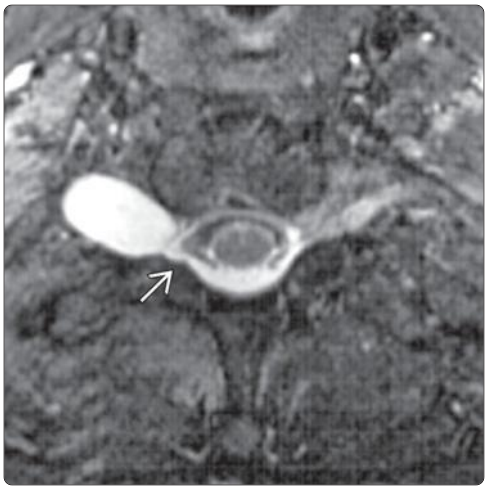
Image Interpretation Pearls

- Posterior vertebral body fragment displaced into canal may be cause of anterior dural tear

SELECTED REFERENCES

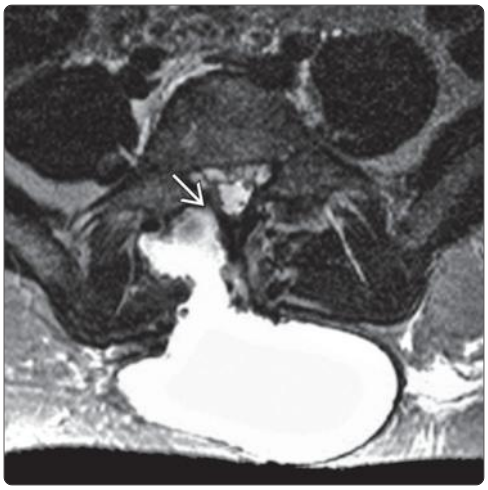
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(Left) *Coronal T2WI FS MR shows right C5, C6, and C7 pseudomeningoceles. Nerve roots are absent, due to chronic avulsion and distal retraction. Components of the brachial plexus are thickened and flaccid.* **(Right)** *Axial T2WI FS MR in the same patient shows pseudomeningocele extending through and widening the right C7-T1 neural foramen. C8 nerve root is chronically avulsed and not seen on this image.*





(Left) *Sagittal T2WI MR shows extensive dorsal epidural fluid collection due to CSF leak in this young female with severe headaches following lumbar puncture.* **(Right)** *Axial T2WI MR in a different patient shows a large CSF signal fluid collection in the dorsal lumbar soft tissues, extending from the right hemilaminectomy site to dorsal subcutaneous tissues. There is no displacement or mass effect upon the thecal sac.*





(Left) *Anteroposterior myelography shows rounded, extraforaminal collection of contrast on the left at C6-C7. The patient had avulsed the left C7 nerve root in a remote automobile accident.* **(Right)** *Sagittal T2WI MR shows a large dorsal fluid collection. Patient is status post multilevel laminectomy and release of tethered cord due to lipomyelomeningocele. Note the fatty mass and myeloschisis in a low-lying cord.*

Traumatic Epidural Hematoma

KEY FACTS

TERMINOLOGY

- Spinal epidural hematoma (SEH)
- Post-traumatic accumulation of blood into spinal epidural space

IMAGING

- May be located at any spinal level
  - Typically extends over multiple levels
  - Fusiform, oval, or tubular
- Extends more freely in dorsal epidural space; ventral spread limited by dural attachment to PLL, anulus
- "Capping" of hematoma by epidural fat on sagittal imaging
  - Confirms epidural (rather than subdural) location
- MR signal intensity depends on age of hematoma

TOP DIFFERENTIAL DIAGNOSES

- Epidural abscess (or phlegmon)
- Epidural lipomatosis
- Epidural tumor
- Extramedullary hematopoiesis
- Ossification of posterior longitudinal ligament (OPLL)
- Sequestered disc fragment

PATHOLOGY

- Clot characteristics depend on compartment, age of collection
- Venous source more common than arterial

CLINICAL ISSUES

- May be associated with significant compression of spinal cord or cauda equina
- Surgical evacuation/decompression may be necessary to alleviate compression of spinal cord, cauda equina

DIAGNOSTIC CHECKLIST

- Contrast injection and fat saturation for complete characterization
- CT may help identify hemorrhage when MR confusing





(Left) *Sagittal STIR MR shows disruption of the anterior longitudinal ligament (ALL) at C5-C6 and a small ventral epidural hematoma. There is a contusion of the cervical cord. Injury to the paraspinous musculature is shown by hyperintense signal.* **(Right)** *Sagittal STIR MR shows a small dorsal epidural hemorrhage in this patient with a L2 Chance fracture. This image shows a portion of the fracture extending through the posterior vertebral body and disruption of the interspinous ligaments at L1-L2.*





(Left) *Sagittal STIR MR shows compression deformity of T1 and dorsal epidural hematoma from C7 through T2-T3 effacing the thecal sac and compressing the cervicothoracic cord. Note how the STIR shows the complex signal of the hematoma distinct from the homogeneous CSF and the fat-suppressed epidural fat.* **(Right)** *Sagittal T2WI shows disruption of the ALL and post-traumatic herniation at C6-C7. There is a large dorsal epidural hematoma, with canal stenosis and cord compression.*

TERMINOLOGY

Abbreviations

- Spinal epidural hematoma (SEH)

Definitions

- Post-traumatic accumulation of blood into spinal epidural space

IMAGING

General Features

- Best diagnostic clue
  - Fusiform, extradural mass-like process with MR signal characteristics of hemorrhage
- Location
  - May be located at any spinal level
  - Dorsal to thecal sac > ventral
    - Extends more freely in dorsal epidural space
    - Ventral spread limited by dural attachments to posterior longitudinal ligament (PLL), anulus
- Size
  - Typically extends over multiple levels
    - Rarely focal
- Morphology
  - Typically fusiform, oval, or tubular
  - May see "capping" of ends of hematoma by epidural fat in thoracic spine on sagittal imaging
    - Confirms epidural (rather than subdural) location

CT Findings

- NECT
  - Hyperdense, extradural collection
- CECT
  - No hematoma enhancement

MR Findings

- T1WI
  - Epidural fluid collection, signal depends on age of hematoma
    - Acute hemorrhage can be similar in signal to cord or CSF on T1WI
    - Subacute (methemoglobin) is hyperintense to CSF and cord
- T2WI
  - Epidural fluid collection, signal depends on age of hematoma
    - Variable signal
      - Acute hematoma can be isointense or slightly hyperintense to CSF on T2WI
      - Diminished T2 signal in late acute to subacute hematoma
      - Isointense/mildly hyperintense relative to CSF (chronic)
- STIR
  - Hyperintense relative to cord and epidural fat
  - Variable signal intensity compared with CSF
- T2* GRE
  - Accentuates susceptibility artifact from blood products
- T1WI C+
  - Mild peripheral enhancement may be seen with subacute hematoma

Angiographic Findings

- No routine role for diagnostic catheter angiography
- Usually negative when searching for bleeding source

Nonvascular Interventions

- Myelography
  - Effaced or constricted subarachnoid space

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Fat-saturated sequences (FS T2WI, STIR)
    - Excludes epidural lipomatosis

DIFFERENTIAL DIAGNOSIS

Abscess, Epidural (or Phlegmon)

- Avid enhancement
  - Peripheral (abscess) or diffuse (phlegmon)
- Typically more focal than SEH, 1 or 2 segments
- Vertebral marrow and disc space changes if associated with spondylodiscitis

Epidural Lipomatosis

- Diffuse multisegmental epidural, almost always dorsal, mass-like process
- Signal intensity matches fat on all sequences
  - Fat saturation nulls signal
- Clinical picture of obesity or metabolic disturbance
- Indolent onset of symptoms or asymptomatic

Epidural Tumor

- Typically quite focal
- Adjacent bone often involved
- Lymphoma may simulate SEH
- Subdural (leptomeningeal) spread is isointense, enhances avidly

Extramedullary Hematopoiesis

- Multifocal lobulated paravertebral and epidural masses
- Most often thoracic spine
- Diffuse vertebral marrow signal abnormality with red marrow conversion

Ossification of Posterior Longitudinal Ligament

- Cervical location most common
- Ventral epidural space
- Longitudinal hypointense signal
  - With extensive OPLL, marrow space can develop
    - T1 hyperintensity can simulate methemoglobin from subacute hematoma
- Osseous nature easily shown with CT

Sequestered Disc Fragment

- Epidural mass, moderate T1/hyperintense T2 signal
- Usually anterior or anterolateral to thecal sac

PATHOLOGY

General Features

- Etiology
  - Venous source more common than arterial

- Spontaneous (1/3 of patients)
  - Reported association with disc herniations
- Spinal trauma
  - Vertebral fractures
  - Missile or other penetrating injury
  - Traumatic disc herniation, annular disruption
  - Blunt trauma
    - Mechanism may relate to suddenly increased pressure within spinal veins leading to vessel rupture
- Therapeutic anticoagulation
  - Coumadin
  - antiplatelet agents
- Instrumentation
  - Spinal hardware
  - Epidural anesthetic
  - Nerve block
  - Facet joint injection
  - Lumbar puncture
- Vascular malformation, vascular tumor

- Genetics
  - Inherited bleeding diatheses are predisposing factors

Gross Pathologic & Surgical Features

- Clot in various stages
- Clot characteristics depend on compartment, age of collection

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Severe, localized back pain
  - Radiculopathy
  - Motor, sensory disturbance
  - Sphincter disturbance
- Clinical profile
  - Spinal trauma
  - Recent instrumentation/penetrating injury
  - Bleeding diathesis, anticoagulation therapy

Demographics

- Epidemiology
  - Infrequent
    - SEH affects 1 per 1,000,000 in USA annually (all causes)
  - Nontraumatic and iatrogenic etiologies are more common
    - Coagulopathy
    - Instrumentation

Natural History & Prognosis

- May be associated with significant compression of spinal cord or cauda equina

Treatment

- Surgical evacuation/decompression may be necessary to alleviate compression of spinal cord or cauda equina
- Without significant neurologic compromise, surgeon may choose to manage SEH conservatively
- Vitamin K injection or fresh frozen plasma to reverse coagulopathy, if present

DIAGNOSTIC CHECKLIST

Consider

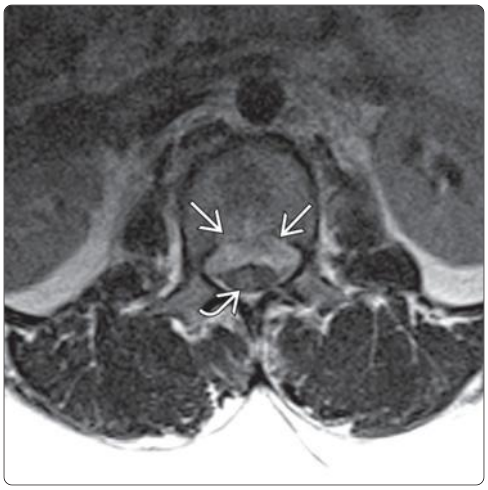
- Contrast injection and fat saturation for complete characterization
  - Active hemorrhage may enhance vividly

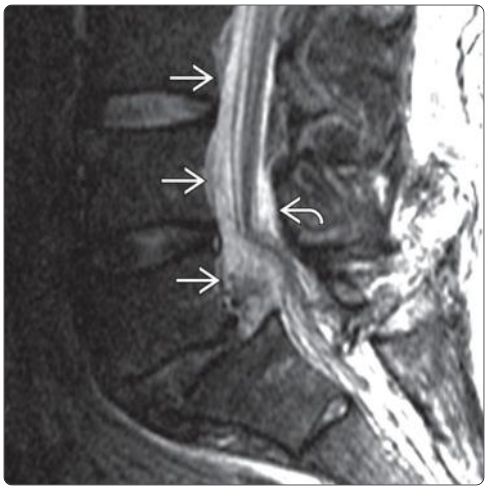
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- CT may help identify hemorrhage when MR confusing

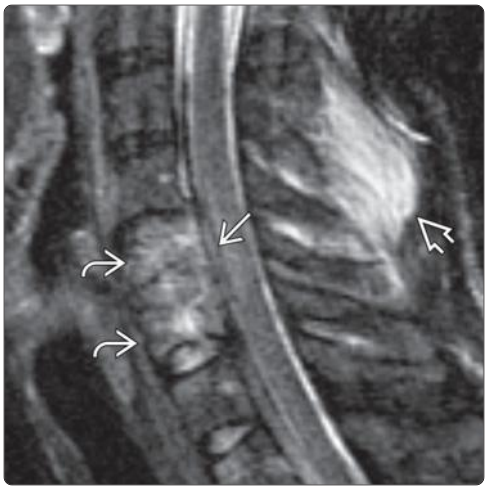
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(Left) *Axial T1WI MR of the lumbar spine shows a large ventral epidural hemorrhage compressing the conus medullaris in this patient with a L1 burst fracture. The hematoma approaches the signal of epidural fat on T1WI but is faintly heterogeneous.* **(Right)** *Sagittal STIR MR shows a large ventral and smaller dorsal epidural hematoma in this patient with a bilateral acute L5 pars fractures and traumatic spondylolisthesis. SEH compresses the thecal sac and cauda equina.*





(Left) *Sagittal STIR MR shows burst fractures of C5 and C6 associated with small ventral SEH. There is edema/hemorrhage in the paraspinous musculature. A contusion of the cord is also seen, with faint hyperintensity in the ventral cord.* **(Right)** *Sagittal STIR MR shows a L1 burst fracture with retropulsion causing severe thecal sac compression. Ventral SEH extends caudally to the L4-L5 level. A small amount of dorsal epidural hemorrhage is also seen at the T12 level.*





(Left) *Sagittal T1WI MR of the thoracic spine in this patient with a L1 burst fracture shows a thin, extensive dorsal epidural hemorrhage sandwiched between the brighter epidural fat and the darker CSF.* **(Right)** *Sagittal STIR MR of the same patient shows the extensive dorsal epidural hematoma, hyperintense and quite distinct from the suppressed epidural fat signal.*

Traumatic Subdural Hematoma

KEY FACTS

TERMINOLOGY

- Traumatic spinal subdural hematoma (tSSDH)
- Accumulation of blood between dura and arachnoid layers of spine

IMAGING

- Lobulated subdural mass with smooth margins
  - May be located at any level of spine
  - Tends to have biconvex or lentiform appearance on both sagittal and axial imaging
- MR signal depends on age of hematoma
  - MR most useful to assess size/extent and to evaluate impact on neurologic structures
- Variable degree of compression of spinal cord or nerve roots

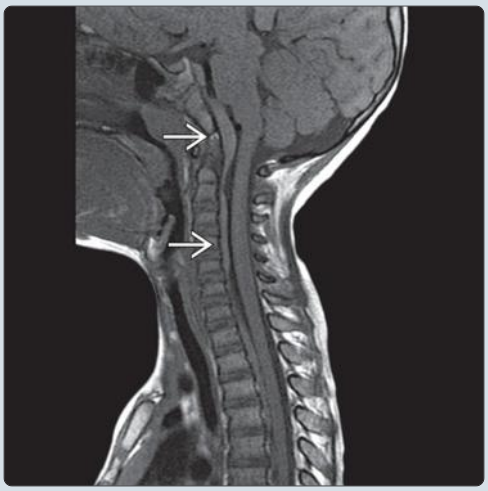
TOP DIFFERENTIAL DIAGNOSES

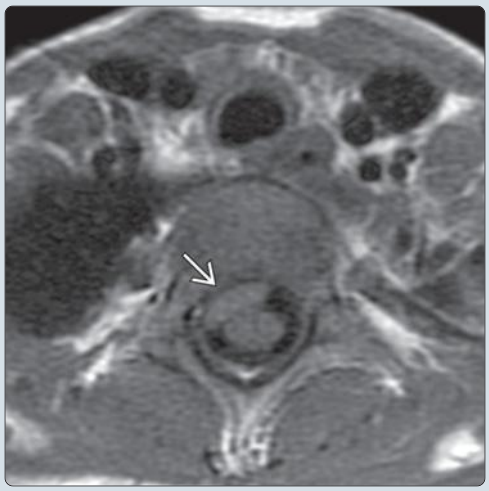
- Hematoma, epidural, traumatic
- Abscess, subdural
- Arachnoid cyst
- Meningioma

CLINICAL ISSUES

- Overall incidence of SSDH is rare, as subdural space is relatively avascular
  - tSSDH is particularly rare
- May be associated with significant compression of spinal cord or cauda equina
- Surgical decompression or percutaneous drainage may be performed to alleviate compression of spinal cord or cauda equina
- Conservative management may be considered
  - Usually resorbs within weeks or several months, depending on size
- Retroclival subdural hematoma requires evaluation for possible atlantooccipital dislocation

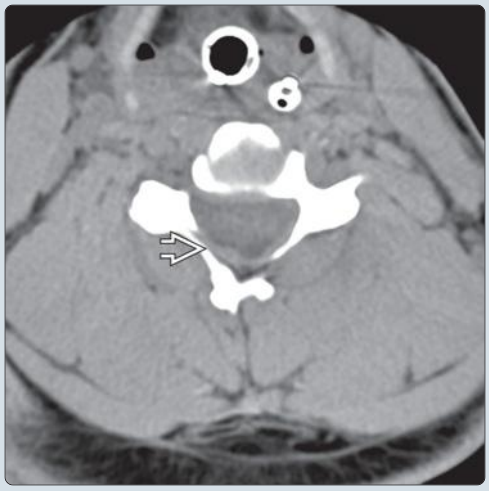
(Left) *Sagittal T1WI MR shows a typical case of early subacute clival and subdural hematoma, demonstrating typical lobulated margin of the arachnoid, effacing the ventral thecal sac.* **(Right)** *Axial T1WI MR through the lower cervical spine shows early subacute subdural hematoma, demonstrating typical lobulated margin of the arachnoid, effacing the ventral thecal sac, and impinging on the anterolateral cervical cord.*





(Left) *Sagittal CECT myelogram shows extensive post-traumatic spinal subdural hematoma with contiguous intracranial extension. Note the larger ventral and smaller dorsal components of the hematoma.* **(Right)** *Axial NECT shows post-traumatic spinal subdural hematoma, with a small, circumferential subdural hematoma at the level of the mid cervical spine.*





TERMINOLOGY

Abbreviations

- Traumatic spinal subdural hematoma (tSSDH)

Definitions

- Accumulation of blood between dura and arachnoid related to trauma

IMAGING

General Features

- Best diagnostic clue
  - Lobulated subdural mass with smooth margins
- Location
  - May be located at any level of spine and extend into or from posterior fossa
  - Cervical is least frequent
- Size
  - Focal or extensive
  - Variable degree of compression of spinal cord or nerve roots
- Morphology
  - Biconvex or lentiform appearance on both sagittal and axial imaging

Radiographic Findings

- May see associated vertebral fracture

CT Findings

- Hyperdense, subdural collection
- May be difficult to differentiate from other epidural or subdural/extramedullary processes on myelography

MR Findings

- T1WI
  - Subdural fluid collection, signal depends on age of hematoma
    - Subacute (methemoglobin) is hyperintense to CSF and cord
- T2WI
  - Subdural fluid collection, signal depends on age of hematoma
    - Varies from iso-/hyperintense (acute) to hypointense (subacute) relative to CSF
- STIR
  - Hyperintense relative to cord and epidural fat
  - Variable signal intensity compared with CSF
- T2* GRE
  - Usually hypointense due to susceptibility associated with blood products

Imaging Recommendations

- Best imaging tool
  - MR most useful to assess size/extent and to evaluate impact on neurologic structures
- Protocol advice
  - Contrast helpful to differentiate from meningioma or epidural abscess

DIFFERENTIAL DIAGNOSIS

Hematoma, Epidural, Traumatic

- "Capping" of hematoma by epidural fat on sagittal imaging
- Mostly dorsal epidural space
- Variable signal due to age of blood products

Abscess, Subdural

- Isointense on T1WI, hyperintense on T2WI
- Peripheral enhancement; vertebral changes if associated with spondylodiscitis

Arachnoid Cyst

- Follows CSF on all sequences, no enhancement
- May appear to be in subdural or epidural space

Meningioma

- Typically enhances or is densely calcified

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often presents with signs of neurologic compromise
  - May mimic presentation of herniated disc
  - Retroclival subdural hematoma requires evaluation for possible atlantooccipital dislocation
- Other signs/symptoms
  - Can be seen with concomitant subdural hematoma in head

Demographics

- Epidemiology
  - Overall incidence of SSDH is rare, as subdural space is relatively avascular
    - Nontraumatic etiologies are more common
      - Coagulopathy, AVM, or vascular tumor

Natural History & Prognosis

- May be associated with significant compression of spinal cord or cauda equina
- Prognosis is generally good if pressure can be relieved prior to irreversible injury
- Usually resorbs within weeks or several months

Treatment

- Conservative management may be considered
- Surgical decompression or percutaneous drainage may be performed to alleviate compression of spinal cord or cauda equina

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Vascular Injury, Cervical

KEY FACTS

TERMINOLOGY

- Traumatic injury to either carotid or vertebral arteries

IMAGING

- Intimal flap
- Long, tapered vascular narrowing or occlusion (stenoocclusive pattern)
- Pseudoaneurysm
  - Dissecting aneurysm pattern
- Traumatic vertebral dissection usually involves V2 or V3 segments
- Crescentic intramural hematoma

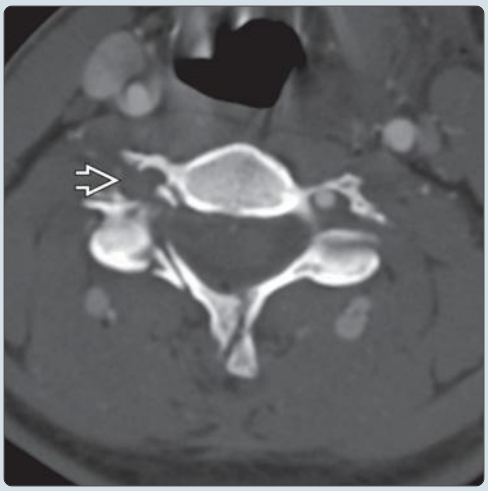
PATHOLOGY

- Direct blunt trauma to neck
- Abrupt rotation or hyperextension of neck
- Direct puncture or laceration
- Dissection
  - Intimal tear, allows jet of arterial blood into vessel wall, developing false lumen

CLINICAL ISSUES

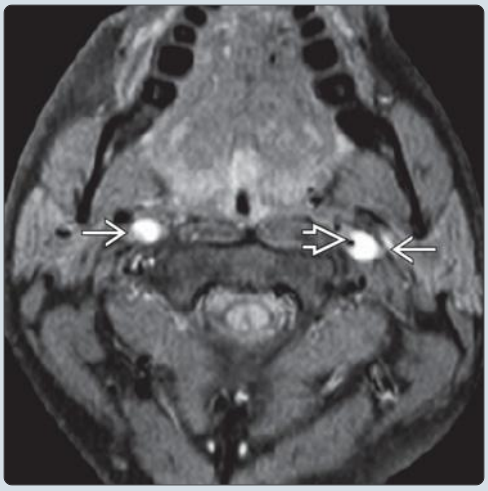
- Presenting symptoms
  - Ipsilateral pain, bruit
  - CNS ischemia, stroke
    - Neurologic symptoms may be delayed
  - Horner syndrome
- Blunt cerebrovascular injury seen in 0.1-1.1% of all trauma admissions
- Systemic anticoagulation to prevent ischemic sequela
- Larger dissections may require fenestration &/or endovascular stenting
- Treatment for pseudoaneurysms includes covered stent placement or sacrifice of host vessel

(Left) *Axial CTA shows occlusion of the right vertebral artery in this patient with comminuted fracture of the C5 vertebral body, which extends into the right foramen transversarium.* **(Right)** *Angiography performed from the right common carotid artery shows right ICA occlusion with a proximal stump in this patient with an occlusion due to carotid dissection.*





(Left) *Axial T1WI FS MR through the neck shows round hyperintensity from bilateral carotid dissections, due to methemoglobin within the vessel walls. Right ICA is occluded; flow void on the left demonstrates tiny residual vessel lumen.* **(Right)** *Parasagittal MRA MIP image in the same patient shows signal from methemoglobin along the distal internal carotid courses. Methemoglobin obscures the flow-related enhancement of the MRA technique, which could be falsely interpreted as patent flow.*





TERMINOLOGY

Synonyms

- Blunt cerebrovascular injury (BCVI)
- Carotid artery injury (CAI)
- Vertebral artery injury (VAI)

Definitions

- Traumatic injury to either carotid or vertebral arteries
  - Dissection
  - Thrombosis
  - Pseudoaneurysm
  - Transection

IMAGING

General Features

- Best diagnostic clue
  - Dissection
    - Intimal flap
    - Long, tapered vascular narrowing or occlusion (steno-occlusive pattern)
      - Occluded carotid dissection often leaves tapered, flame-shaped ICA stump
    - Pseudoaneurysm (dissecting aneurysm pattern)
  - Pseudoaneurysm
    - Eccentric vascular protrusion
- Location
  - Cervical ICA
    - Dissection most commonly begins at or near carotid bifurcation
    - Infrequently extends into intracranial ICA
  - Vertebral artery
    - Traumatic vertebral dissection usually involves V2 or V3 segments
- Morphology
  - Dissection
    - Smoothly tapered narrowing of vascular lumen
    - Intimal flap is pathognomonic
  - Occlusion
    - Segmental or extensive obliteration of vascular lumen
  - Pseudoaneurysm
    - Eccentric vascular enlargement or protrusion

CT Findings

- NECT
  - Evidence of cervical spine or skull base trauma
  - Subarachnoid hemorrhage associated with dissection of intracranial vertebral artery
- CTA
  - Dissection
    - Smooth, tapered narrowing
    - Crescentic intramural hematoma
    - Intimal flap
  - Vascular occlusion
  - Pseudoaneurysm
    - Focal, eccentric vascular protrusion
    - Long segment of vascular enlargement, typically irregular
    - ± perivascular hematoma

MR Findings

- T1WI FS
  - Hyperintense crescentic signal due to thrombosed blood in dissection flap
- T2WI
  - Loss of flow void within vessel due to thrombosis/occlusion or stenosis
- DWI
  - Brain infarct due to vessel occlusion or distal embolus of thrombus
- MRA
  - As per CTA
  - Thrombosed blood may be hyperintense on TOF source images; do not mistake for flow-related enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Luminal narrowing or occlusion
  - Echogenic thrombus
  - Visualization of intimal flap in carotid dissection
  - Widened vessel contour in pseudoaneurysm
- M-mode
  - Evidence of abnormal hemodynamics
    - Elevated velocities
    - High resistance waveforms
    - To-and-fro flow in pseudoaneurysm

Imaging Recommendations

- Best imaging tool
  - **CT angiography**
    - Simple, fast, noninvasive screening examination
    - Recommended as screening tool in patients after blunt cervical trauma who meet modified **Denver screening criteria** for suspected vertebral artery injury
  - MRA supplemented by axial FS T1WI also useful
  - Some centers consider angiography mandatory for BCVI screening

Angiographic Findings

- While the "gold standard" for evaluating vascular injury, it is often replaced by CTA as noninvasive, more widely available screening examination
- Dissection
  - Intimal flap is pathognomonic
  - Long, smoothly tapered stenosis
- Pseudoaneurysm
  - Irregular or eccentric enlargement of vessel lumen

DIFFERENTIAL DIAGNOSIS

Atherosclerotic Stenosis

- Stenosis typically short
- Often densely calcified plaques are seen
- Diffuse narrowing distal to critical stenosis (≥ 95%) due to adaptive narrowing

Spontaneous Dissection

- Absent history of trauma
- Underlying arteriopathy often present, such as fibromuscular dysplasia or Marfan

Catheter-Related Vasospasm

- Focal, reversible narrowing of vessel lumen seen on angiography

Takayasu Arteritis

- Inflammatory vasculitis affecting medium and large vessels
- Smooth, symmetric narrowing of arch vessels

PATHOLOGY

General Features

- Etiology
  - Causes
    - Direct blunt trauma to neck
    - Abrupt rotation or hyperextension of neck
    - Direct puncture or laceration
  - Dissection
    - Intimal tear, allows jet of arterial blood into vessel wall, developing false lumen
    - Intimal flap, dividing true lumen and false lumen
      - Pathognomonic of arterial dissection
    - False lumen may thrombose with complete vessel occlusion
      - Intramural hematoma results in diffuse, smoothly tapered stenosis
    - Subadventitial dissection may result in pseudoaneurysm formation

Staging, Grading, & Classification

- **Modified Denver screening criteria for blunt cerebrovascular injuries**
  - Lateralizing neurologic deficit not explained by CT
    - Infarct on CT
    - Cervical hematoma (nonexpanding)
    - Massive epistaxis
    - Anisocoria or Horner syndrome
    - GCS < 8
    - Cervical spine or skull base fracture
    - Severe facial fracture (Le Forte II or III)
    - Seatbelt sign above clavicle
    - Cervical bruit

Gross Pathologic & Surgical Features

- Dissection
  - Intimal defect, delamination of wall, intramural hematoma
- Pseudoaneurysm
  - Torn/attenuated or ruptured vessel wall with cavitated clot within adventitial tissues

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Ipsilateral pain, bruit, pulsatile tinnitus
  - CNS ischemia, stroke (neurologic symptoms may be delayed)
  - Cranial neuropathies, Horner syndrome
  - Expanding hematoma

Demographics

- Epidemiology
  - Blunt cerebrovascular injury seen in 0.1-1.1% of all trauma admissions
  - Extracranial vascular dissection (all etiologies) causes 1% of all ischemic strokes, 5% of ischemic strokes in young adults

Natural History & Prognosis

- Dynamic process
- Majority (57%) of small intimal irregularities heal on follow-up imaging
- Dissections with larger intimal flaps infrequently heal, and up to 43% develop pseudoaneurysms
- Pseudoaneurysm may heal (25%), decrease in size (15%), be stable (60%)
- Traumatic cervical vessel occlusions rarely recanalize
- Mortality of post-traumatic extracranial carotid dissection ranges from 20-40%
- Mortality of post-traumatic extracranial vertebral dissection ranges from 4-8%

Treatment

- Systemic anticoagulation to prevent ischemic sequela
- Larger dissections may require fenestration &/or endovascular stenting
- Treatment for pseudoaneurysms includes covered stent placement or sacrifice of host vessel

DIAGNOSTIC CHECKLIST

Consider

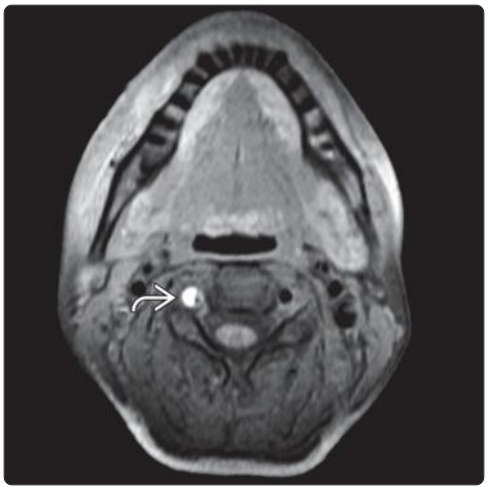
- CT angiographic screening in trauma patients with
  - Cervical spine injuries, especially fractures through foramen transversarium and facet subluxations
  - Evidence of severe head and facial trauma
  - Horner syndrome
  - Neck soft tissue injuries, especially expanding hematoma
  - Cervical bruit in younger (< 50 year old) patient
  - Neurologic abnormalities unexplained by known intracranial injuries

Image Interpretation Pearls

- If injury to 1 carotid or vertebral artery is seen, look carefully at others for additional injury

SELECTED REFERENCES

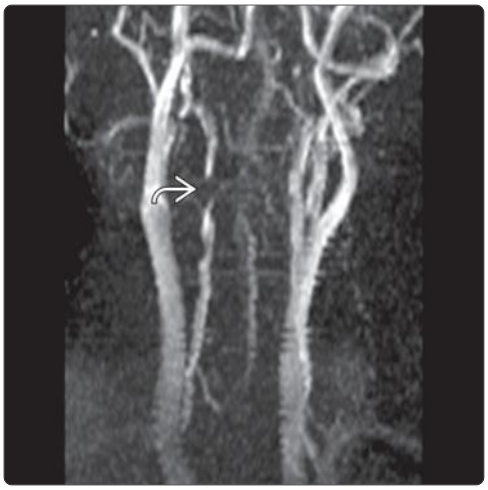
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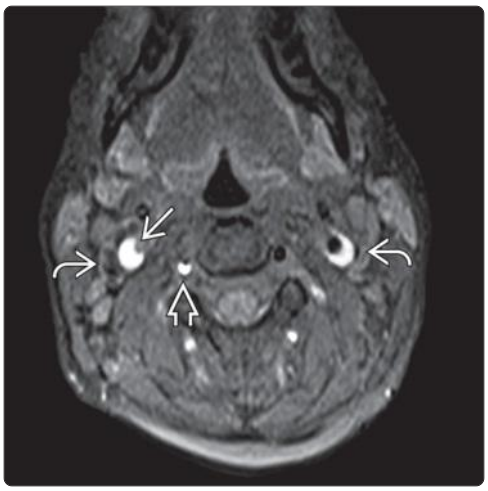


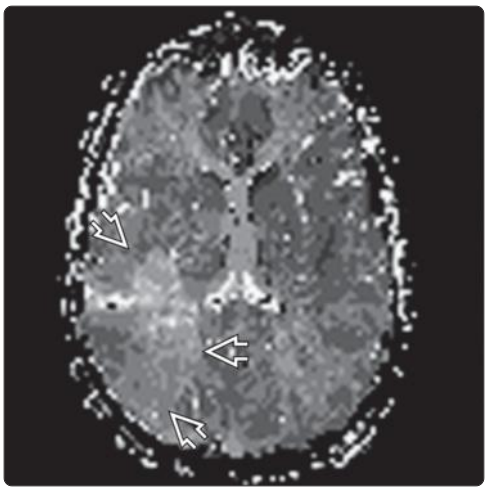
(Left) *Axial T1WI FS MR shows crescentic hyperintense signal in the right vertebral artery due to a dissection. The outer diameter of the vessel is enlarged. Note the patent flow void medially.* **(Right)** *Frontal projection DSA of the right vertebral artery in the same patient shows a short segment of fusiform narrowing of the distal V2 segment due to a focal vertebral artery dissection. The vessel is normal distally.*





(Left) *Lateral catheter angiography performed with injection of the left common carotid artery shows occlusion of the left internal carotid artery. Note the flame-shaped stump of the ICA.* **(Right)** *Coronal MIP projection of 2D TOF MRA in a patient with evidence of traumatic ligamentous injury on MR shows focal loss of flow-related enhancement within the right vertebral artery at site of vascular dissection.*

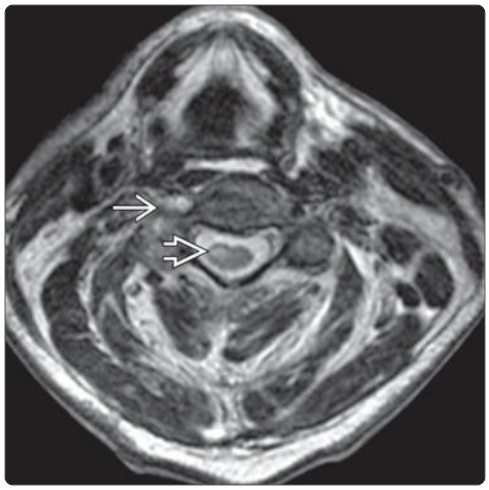




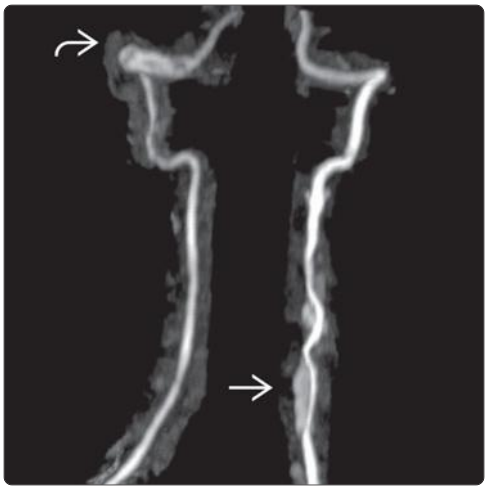
(Left) *Axial T1WI FS MR shows crescentic hyperintensity due to dissections of bilateral internal carotid arteries and the right vertebral artery. There is loss of flow void in the right ICA dissection due to occlusion of the residual lumen.* **(Right)** *Axial MR perfusion study shows prolonged time-to-peak in the dorsal right MCA distribution. Due to internal injuries, anticoagulation was contraindicated, and the patient completed infarct of this territory on follow-up.*

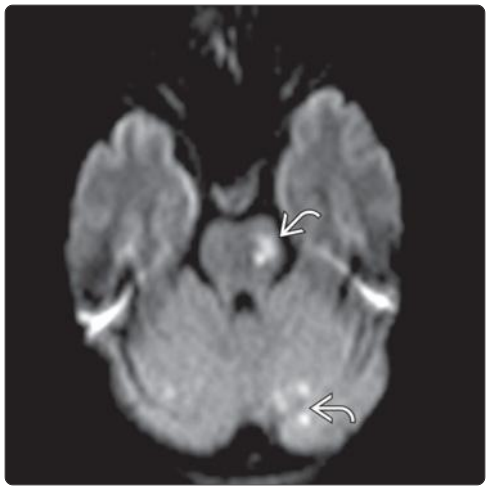
(Left) *Coronal MIP of cervical 2D time-of-flight MRA in this patient with C6-C7 fracture shows loss of flow-related enhancement, demonstrating occlusion of the right vertebral artery.* **(Right)** *Axial T2WI shows loss of flow void in the right vertebral artery due to occlusion. There is also hyperintensity in the right hemicord due to ischemia. One could hypothesize that hemicord infarct in this case could be due to variant anatomy where anterior spinal artery is paired at this level.*



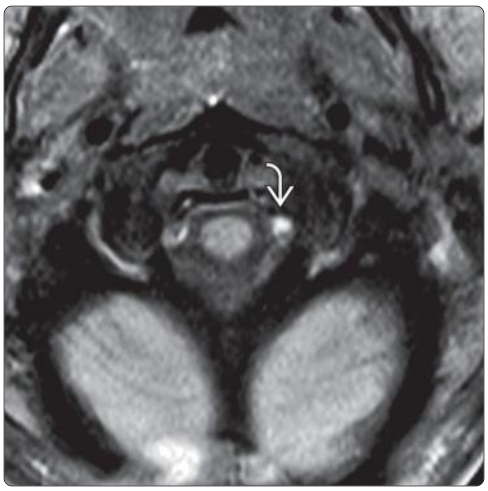


(Left) *Dissection of the V3 segment of the right vertebral artery appears enlarged on TOF MRA, though this was mostly due to T1 shortening from thrombus. There is also a dissection of the proximal left vertebral artery.* **(Right)** *Axial DWI MR of the same patient shows small infarcts in the cerebellum and left pons. This patient had presented with right facial numbness and arm weakness several days after chiropractic manipulation.*



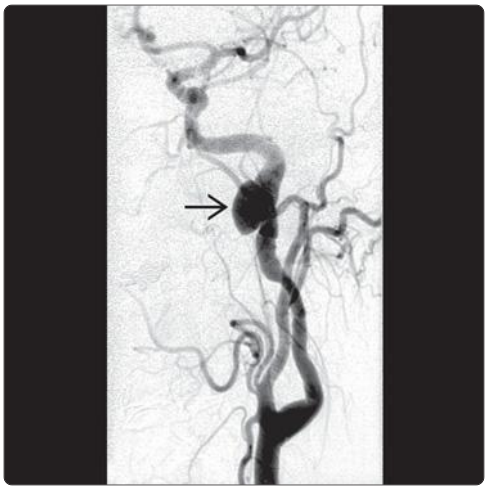


(Left) *Axial T1WI FS MR shows a thin rim of hyperintensity from dissection of the left vertebral artery as it enters the dura. The patient presented with basilar cistern subarachnoid hemorrhage.* **(Right)** *Coronal MIP image of MRA data shows occluded left internal carotid artery due to acute carotid dissection. Note the flame-shaped stump.*









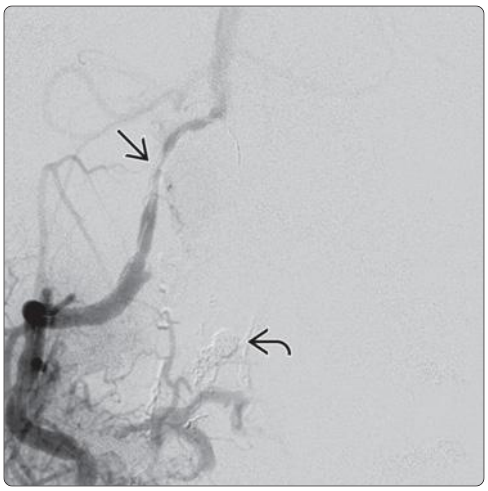
(Left) *AP projection carotid angiography shows large carotid artery aneurysm of the right ICA just below the level of the skull base. Early post-injection, slow-moving contrast flows around aneurysm margin, with incomplete opacification of the lumen. ICA is medially deviated due to mass effect. Note focal narrowing with post-stenotic dilatation.* **(Right)** *Lateral projection of left carotid angiography shows pseudoaneurysm. Beaded lumen proximally is due to fibromuscular dysplasia.*





(Left) *Lateral DSA of the left ICA shows an irregular saccular protrusion arising from the vessel just below the skull base, demonstrating the appearance of a pseudoaneurysm. DSA typically underestimates the size of a pseudoaneurysm, missing the surrounding lamellated thrombus typically present.* **(Right)** *Lateral projection left carotid angiogram shows tapered occlusion of right internal carotid artery, reflecting underlying dissection. Note absence of intracranial branches of the ICA.*





(Left) *Right vertebral angiogram shows a short segment of smooth, eccentric narrowing of the distal right vertebral artery due to vascular dissection.* **(Right)** *Frontal projection right vertebral angiography shows irregular narrowing of the intradural segment of the right vertebral artery, due to vertebral artery dissection and spasm associated with endovascular embolization of an upper cervical cord AVM. Note embolic material.*

Traumatic Arteriovenous Fistula

KEY FACTS

TERMINOLOGY

- Traumatic dural AV fistula (tDAVF)
- Trauma-induced arteriovenous fistula
- Vertebrovertebral arteriovenous fistula

IMAGING

- DSA is "gold standard" for diagnosing and defining tDAVF
  - DSA demonstration of AVF nidus with enlarged draining veins
  - Unlike spontaneous DAVF (sDAVF), often no cord enlargement/hyperintensity from congestive myelopathy
- Angiographic protocol for evaluation of suspected cervical tDAVF includes selective injections of both external carotid + vertebral arteries, thyrocervical trunks, and paired segmental arteries
  - Angiographic appearance is similar to sDAVF, but nidus may be extraforaminal/paravertebral instead of along nerve root

PATHOLOGY

- Any segment of spine, most frequently cervical
- Usually caused by direct penetrating wounds
- May also occur with blunt trauma and vertebral fracture

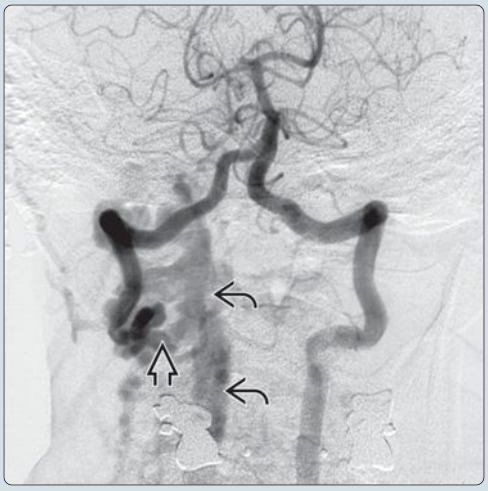
CLINICAL ISSUES

- Symptoms may relate to
  - Venous hypertensive myelopathy
  - Compression by enlarged draining veins
  - Steal phenomenon
- Endovascular treatment with embolization or surgical ligation





(Left) *Off-midline sagittal T1WI MR shows an enlarged ventral epidural venous plexus with a prominent flow void in this patient with a C2 fracture.* **(Right)** *Axial T2* GRE MR shows dilation of the epidural venous plexus with prominent flow voids causing compression of the nerve root in the lateral recess of the cervical spine.*





(Left) *Anteroposterior DSA from the left vertebral artery shows high-flow arteriovenous communication near the V2/V3 junction of the right vertebral artery, with reversed flow in the distal right vertebral artery supplying dilated epidural plexus and dilated deep cervical veins.* **(Right)** *Coronal CT myelography of the lumbar spine shows dilated, tortuous veins in the caudal thecal sac due to a post-traumatic (iatrogenic) arteriovenous fistula.*

TERMINOLOGY

Abbreviations

- Arteriovenous fistula (AVF)
- Traumatic dural AV fistula (tDAVF)

Synonyms

- Vertebrovertebral arteriovenous fistula

Definitions

- Trauma-induced arteriovenous fistula

IMAGING

General Features

- Best diagnostic clue
  - DSA demonstration of AVF nidus with enlarged draining veins
- Location
  - Dural or extradural nidus
  - Any segment of spine, most frequently cervical
- Size
  - Small nidus
  - May have extensive draining venous engorgement
- Morphology
  - Small, tight AVF nidus
  - Serpiginous veins

CT Findings

- CECT
  - Dilated enhancing perimedullary or epidural veins
- CT myelography
  - Serpiginous, dilated (intradural) perimedullary veins or epidural defects due to enlarged epidural venous plexus

MR Findings

- T1WI
  - Perimedullary or epidural flow voids from enlarged draining veins
- T2WI
  - Unlike spontaneous DAVF, often no cord enlargement/hyperintensity from congestive myelopathy
  - Perimedullary or epidural flow voids from enlarged draining veins
    - May compress cord or nerve roots
- T1WI C+
  - May ↑ conspicuity of enlarged draining veins
- MRA
  - Both TOF and contrast techniques have been described

Angiographic Findings

- Conventional
  - DSA is "gold standard" for diagnosing/defining tDAVF
  - Angiographic protocol for evaluation of suspected cervical tDAVF includes selective injections of both external carotid + vertebral arteries, thyrocervical trunks, and paired segmental arteries
  - Demonstrates solitary AVF nidus and perimedullary or epidural enlarged draining veins
    - Angiographic appearance is similar to sDAVF, but nidus may be extraforaminal/paravertebral instead of along nerve root

Imaging Recommendations

- Best imaging tool
  - Digital subtraction angiography

DIFFERENTIAL DIAGNOSIS

Spontaneous Dural AVF

- Typically older men who present with chronic progressive myelopathy
- Hyperintense cord expansion from congestive myelopathy

Spinal AVM

- Intradural; nidus on or within cord parenchyma
- Congenital vascular lesions

PATHOLOGY

General Features

- Etiology
  - Usually caused by direct penetrating wounds
    - Stabbing, gunshot wound
  - Also with blunt trauma and vertebral fracture

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Bruit, pain
  - Weakness, paresis, radiculopathy
  - Symptoms related to
    - Venous hypertensive myelopathy
    - Compression by enlarged draining veins
    - Steal phenomenon
- Other signs/symptoms
  - Expanding hematoma
  - Subarachnoid hemorrhage

Demographics

- Epidemiology
  - Rare lesions; much rarer than spontaneous DAVF

Natural History & Prognosis

- Rapidly progressive without treatment

Treatment

- MR with MRA provides improved screening for DAVF, guiding subsequent DSA studies by helping target fistula level
- Endovascular treatment with embolization or surgical ligation
  - Embolization preferred given decreased invasiveness

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Wallerian Degeneration

KEY FACTS

TERMINOLOGY

- Orthograde degeneration, secondary degeneration
- Disintegration of axon and myelin sheath after axonal connection to neuronal cell body is disrupted

IMAGING

- Increased signal from dorsal columns above site of cord injury
- Increased signal from corticospinal tracts below site of cord injury
- Most apparent in cervical cord
- Less apparent in lower thoracic cord and conus, since fewer axons in distal cord
- Usually seen > 7 weeks following cord injury

TOP DIFFERENTIAL DIAGNOSES

- Multiple sclerosis
- Viral myelitis
- Subacute combined degeneration
- Syrinx, post traumatic
- Infarction, spinal cord

PATHOLOGY

- Stage 1 (1st 4 weeks): Physical degradation of axon with no change in myelin
- Stage 2 (4-14 weeks): Myelin protein breakdown
- Stage 3 (> 14 weeks): Myelin lipid breakdown and gliosis
- Stage 4 (late): Volume loss from atrophy

CLINICAL ISSUES

- Typical history of severe prior trauma with paraparesis or paraplegia

DIAGNOSTIC CHECKLIST

- Important to recognize as sequelae of prior cord injury and not intrinsic new cord pathology

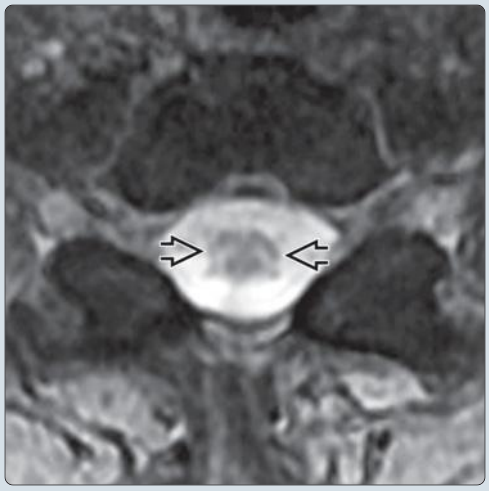
(Left) *Prior C7 trauma with extensive anterior cervical fusion and laminectomy. Sagittal T2WI shows instrumented fusion at C4-C5 and fusion at C6-T1 level on a congenital basis. There has been prior laminectomy with large focus of focal cord myelomalacia at C7 seen as cord hyperintensity.* **(Right)** *Axial T2* GRE MR shows the extensive cord myelomalacia at C7 with diffuse increased signal from the cord substance ⇨, with essentially no normal cord signal.*





(Left) *Above the cord lesion, the axial T2* GRE MR shows well-defined high signal intensity wallerian degeneration within the dorsal columns ⇨.* **(Right)** *Below the cord lesion, the axial T2* GRE MR shows high signal intensity wallerian degeneration within the lateral corticospinal tracts ⇨.*





TERMINOLOGY

Synonyms

- Orthograde degeneration, secondary degeneration

Definitions

- Disintegration of axon and myelin sheath after axonal connection to neuronal cell body is disrupted

IMAGING

General Features

- Best diagnostic clue
  - Increased T2 signal from dorsal columns above cord lesion; increased signal from lateral corticospinal tracts below cord lesion
- Location
  - Cervical spinal cord > thoracic
- Size
  - Variable longitudinal length, generally several vertebral bodies
- Morphology
  - Linear, following long axis of spinal cord

MR Findings

- T2WI
  - Increased signal from dorsal columns above site of cord injury
  - Increased signal from corticospinal tracts below site of cord injury
    - Most apparent in cervical cord
    - Less apparent in lower thoracic cord and conus since fewer axons in distal cord
    - Usually seen > 7 weeks following cord injury

Imaging Recommendations

- Best imaging tool
  - MR with axial T2W or GRE images

DIFFERENTIAL DIAGNOSIS

Multiple Sclerosis

- Multiple lesions with intervening normal cord

Viral Myelitis

- Long segment diffuse cord involvement

Subacute Combined Degeneration

- Dorsal column involvement only

Syrinx, Post Traumatic

- CSF signal focus within cord, with expansion

Infarction, Spinal Cord

- Ventral cord involved with anterior spinal artery territory

PATHOLOGY

General Features

- Etiology
  - Sensory tracts are posterior columns (gracilis, cuneatus) of spinal cord and carry touch, conscious proprioception, and vibration from periphery by neurons afferent to brain
  - Major lateral corticospinal tracts located in lateral aspect of spinal cord
    - Smaller, ventral corticospinal tract along ventral midline not usually visualized

Staging, Grading, & Classification

- Stage 1 (1st 4 weeks): Physical degradation of axon with no change in myelin
- Stage 2 (4-14 weeks): Myelin protein breakdown
- Stage 3 (> 14 weeks): Myelin lipid breakdown and gliosis
- Stage 4 (late): Volume loss from atrophy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Typical history of severe prior trauma with paraparesis or plegia

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Important to recognize as sequelae of prior cord injury and not intrinsic new cord pathology

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PART III
SECTION 1

Degenerative Diseases



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Disc Degeneration

Overview. Describing morphology alterations seen in degenerative disc disease requires common terminology to standardize communication. A series of pathoanatomic terms and definitions proposed by an interdisciplinary committee and endorsed by multiple societies is summarized here. These general terms are descriptive and independent of imaging modality.

It cannot be overemphasized that the specific terms described below do not imply knowledge of etiology, symptoms, prognosis, or need for treatment.

MR signal intensity. Any or all of the following can occur with **disc degeneration**: Real or apparent desiccation, fibrosis, narrowed intervertebral disc space, bulging, fissuring/mucinous degeneration of the anulus, osteophytes of the vertebral apophyses, and endplate/adjacent marrow changes.

Severely degenerated discs with markedly decreased signal intensity may demonstrate linear areas of high signal intensity on T2-weighted images that may represent free fluid within cracks or fissures of the degenerated complex. Signal intensity changes in the intervertebral disc on T1-weighted images, while much less common than the loss of signal noted on T2-weighted images, can also be seen with degeneration.

Regions of decreased or absent signal within **heavily calcified discs** may occur. Signal loss has been attributed to a low mobile proton density as well as, in the case of gradient echo imaging, its sensitivity to the heterogeneous magnetic susceptibility found in calcified tissue.

Focal or diffuse areas of hyperintensity on T1-weighted spin-echo sequences may also be encountered in densely calcified intervertebral discs. These changes are related to T1 relaxation times secondary to a surface relaxation mechanism. These regions of high signal intensity on T1 are unaffected by fat suppression, suggesting that it is a T1 shortening effect rather than the presence of lipid. Hyperintensities within severely degenerated intervertebral discs that suppress on fat-saturation techniques presumably relate to areas of fatty marrow.

Separation between, or avulsion of, anular fibers from their vertebral body insertions, or breaks through fibers that extend transversely in a radial fashion, or concentrically through layers of the anular lamellae, are referred to as **anular fissures**. On MR, these changes are seen on T2-weighted images as high signal intensity within the outer anulus/PLL complex (the so-called "high intensity zone"). These regions will also sometimes enhance following the administration of paramagnetic contrast, an effect thought to be secondary to reactive reparative tissue. The term "anular tear" should be avoided.

The role of anular disruption as the principle causal agent of disc degeneration has not been proven. In light of the continuing controversy surrounding the concept of "internal disc disruption," it is probably unwarranted to imply that radial tears are more than a manifestation of advanced degeneration. While no data clearly supports an unequivocal causal relationship between these degeneration changes and symptoms, anular disruption is important to consider because of a controversial concept of "discogenic pain" and its implications concerning the usefulness of discography for diagnosis. Back pain is thought to occur in some patients without morphological abnormalities, such as herniation or stenosis, which are thought to be related to leakage of nuclear material through the disrupted anulus into the epidural space.

Facet and ligament degenerative changes can occur with or without accompanying disc degeneration and can be readily identified on imaging studies. These changes are best described level by level, along with the presence of foraminal and canal narrowing, facet fluid, cysts, and other abnormalities.

Bulge vs. Herniation

The term **bulge** is used to describe a generalized extension greater than 50% of the circumference of the disc tissues, extending a short distance (< 3 mm) beyond the edges of the adjacent apophyses. A bulge is not a herniation, although 1 portion of the disc may be bulging and another portion of the disc may herniate.

A bulge is often a normal variant, particularly in children in whom all normal discs appear to extend slightly beyond the vertebral body margin. Bulge may also be associated with disc degeneration or may occur as a response to axial loading or angular motion with ligamentous laxity. Occasionally, a bulge in 1 plane is really a central subligamentous disc herniation in another plane. Asymmetric bulging of disc tissue greater than 25% of the disc circumference may be seen as an adaptation to adjacent deformity, and is not considered a form of herniation.

Herniations are a localized displacement of disc material beyond the limits of the intervertebral disc space in any direction. The disc space is defined in the cranio-caudad direction by the vertebral body endplates and peripherally by the outer edges of the vertebral ring apophyses, exclusive of any osteophytes. If the herniation is less than 25% of the disc circumference, it is referred to as **focal or localized**. If it is between 25% and 50%, it has been referred to as **broad based**. This term has been eliminated in the latest version of the nomenclature.

Protrusions are herniations in which the greatest distance in any plane between the edges of disc material beyond the disc space is less than the distance between the edges of the base in the same plane. In practical terms, this looks like a triangle on sagittal images, with the base of the triangle at the disc margin and the apex of the triangle within the epidural space.

Extrusions are herniations in which, in at least 1 plane, any 1 distance between the edges of the disc material beyond the disc space is greater than the distance between the edges of the base in the same plane, or when no continuity exists between the disc material beyond the disc space and that material within the disc space. In practical terms, this looks like the "toothpaste" sign on the sagittal image, with the larger component of the herniation within the epidural space and a smaller pedicle connecting to the intervertebral disc.

If extruded disc material is not contiguous to the disc space, it is referred to as a **sequestration** (or **"free fragment"**). The extrusion is referred to as "migrated" if it has been displaced from the site of extrusion, regardless of whether or not it is sequestrated. The signal intensity of the extruded portion may be increased or decreased on T2-weighted images. All disc herniations, small or large, can be associated with enhancement, and this enhancement may constitute a large

portion of the extradural mass. Acute disc herniations may also cause focal epidural hemorrhage.

Sequestrated fragments can lie anterior to the posterior longitudinal ligament, especially if they have migrated behind the vertebral bodies where the posterior longitudinal ligament is not in direct opposition. Fragments may also lie posterior to the ligament. Sequestrated fragments within the lateral recess and the neural foramen have been shown to produce eroded cortical bone and expansion of those spaces and thus, should be considered in the differential diagnosis of a mass arising and expanding the neural foramen and lateral recess.

In rare instances, disc herniation may extend through the dura and arachnoid. The mechanism of **intradural disc herniation** is thought to be the development of chronic inflammation leading to adhesions between the dura and the posterior longitudinal ligament. Nevertheless, there is almost invariably penetration through the posterior longitudinal ligament posteriorly, where it is fused with the anulus, or superiorly or inferiorly, where it fuses with the vertebral body margin. Intradural disc herniations may enhance and mimic neoplasm.

The characterization of a disc herniation is not always clear cut, and it may appear as a protrusion in 1 plane and an extrusion in another. If there is displacement away from the disc space in any plane, it should be referred to as an extrusion. Containment refers to the integrity of the outer anulus covering the disc herniation.

One may view the continuum of herniated disc disease as starting with anular disruption, proceeding on to small focal herniation (which is not broken completely through the anulus-ligamentous complex) to frank herniation (extrusion). The extrusion has dissected through the anulus and posterior ligamentous complex completely. These extrusions may show variable degrees of containment, and a line of decreased signal intensity has been reported around sequestrated fragments and large extruded discs where there has clearly been disruption of the anulus and ligament. This is thought to be secondary to anular and ligamentous fibers that are carried away with the disc herniation.

The anulus fibrosus and posterior longitudinal ligament are so intertwined at the level of the disc that a distinction between the 2 structures may be impossible or, for that matter, irrelevant. Technical limitations of CT and MR usually preclude the distinction of a contained from an uncontained disc herniation. In the transverse plane, the disc abnormality is usually described as **central**, **right** or **left central**, **subarticular**, **foraminal**, or **extraforaminal** (far lateral). In the sagittal plane, **discal**, **infrapedicular**, **suprapedicular**, and **pedicular** are most commonly employed.

Several nonstandard terms should be avoided. These include "disc material beyond the interspace" (DEBIT), which has been superseded by protrusion and extrusion categorization. "Herniated nucleus pulposus" is an old favorite but is not accurate since much of what herniates is not nucleus pulposus (cartilage, fibrous tissue). "Ruptured" disc has no role in a report since this implies a specific knowledge of a traumatic etiology, which is nearly always lacking. Finally, "prolapse" is synonymous with the standard term "protrusion."

Degenerative Endplate Changes

The relationship among the vertebral body, endplate, and disc has been studied by using both degenerated and chymopapain-treated discs as models. A common observation in MR imaging is signal intensity changes in the vertebral body marrow adjacent to endplates of a degenerative disc. These changes appear to take 3 main forms.

Type I changes demonstrate a decreased signal intensity on T1-weighted images and an increased signal intensity on T2-weighted images, and these changes have been identified in approximately 4% of patients scanned for lumbar disease. Type I changes are also seen in approximately 30% of chymopapain-treated discs, which may be viewed as a model of acute disc degeneration.

Type II changes are represented by increased signal intensity on T1-weighted images and an isointense or slightly hyperintense signal on T2-weighted images. These changes are seen in approximately 16% of cases. In both types I and II, there is evidence of associated degenerative disc disease at the level of involvement. Mild enhancement of type I vertebral body marrow changes is seen with Gd-DTPA, which at times can extend to involve the disc itself. This enhancement is presumably related to vascularized fibrous tissue within adjacent marrow.

Histopathological sections of a disc with type I changes demonstrate disruption and fissuring of the endplate and vascularized fibrous tissues within the adjacent marrow, producing prolongation of T1 and T2. Discs with type II changes also show evidence of endplate disruption with yellow marrow replacement in the adjacent vertebral body, resulting in a shorter T1. There appears to be a relationship between the different endplate types, as type I changes have been observed to convert to type II with time, and type II changes seem to remain stable.

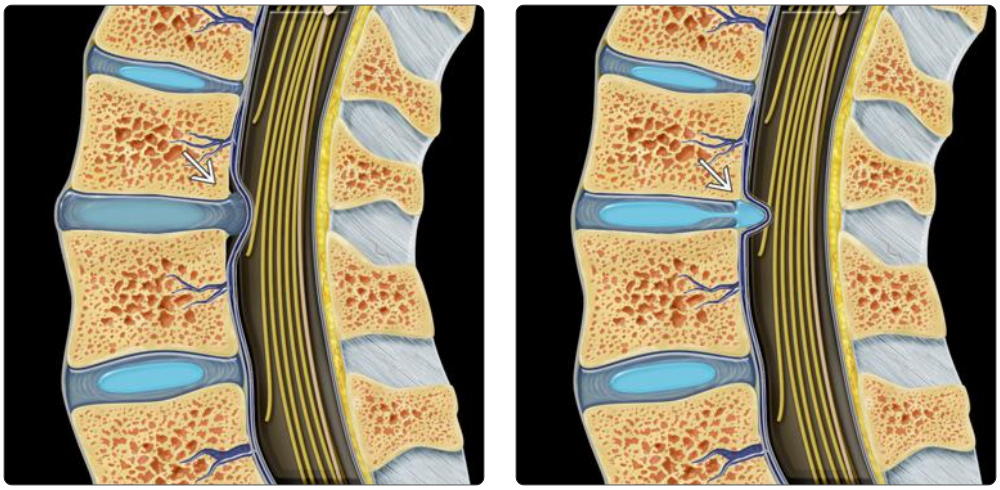
Type III changes are represented by decreased signal intensity on both T1- and T2-weighted images. These findings seem to correlate with the presence of extensive bony sclerosis.

While the signal intensity changes of type I may be similar to those seen in vertebral osteomyelitis, the distinguishing factor (at least in the adult population) is the involvement of the intervertebral disc, which shows an abnormal high signal intensity and abnormal configuration on T2-weighted images with infection. Increased signal from the disc may suggest an active inflammatory process. Disc narrowing, sclerosis, and vertebral endplate irregularity suggestive of osteomyelitis has also been demonstrated in long-term hemodialysis and calcium pyrophosphate disease. Classically, in the patient with hemodialysis spondyloarthropathy, the intervertebral disc maintains low signal intensity on both T1- and T2-weighted sequences. Crystal disorders should show increased signal intensity on long TE/TR sequences.

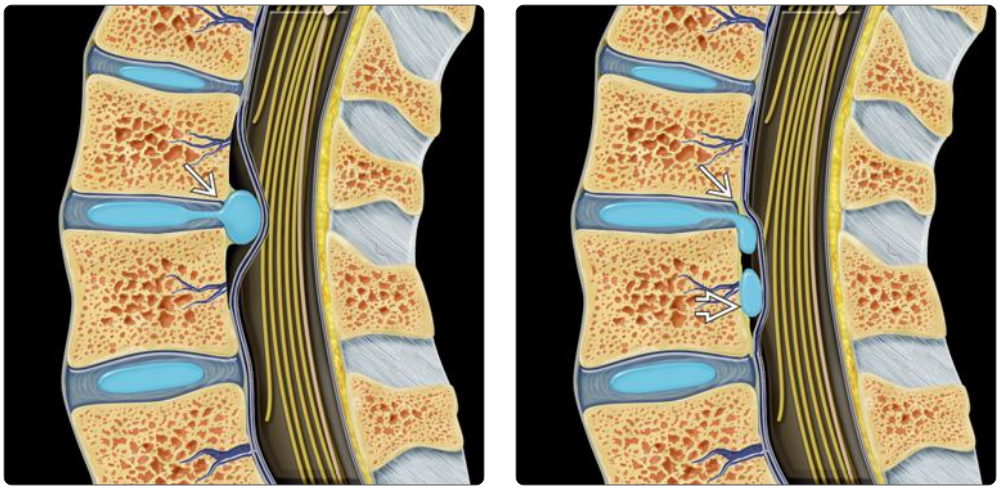
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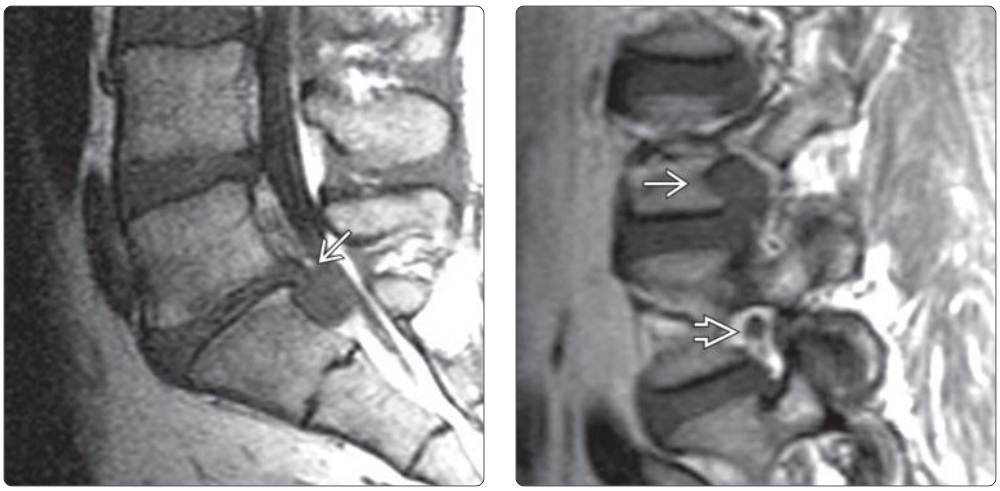
(Left) *Sagittal graphic of a disc bulge shows generalized extension of the disc margin beyond the vertebral body ⇨. An axial view must show generalized extension > 90° of the disc margin to be a bulge; less than 90° would be a protrusion.* **(Right)** *Sagittal graphic of a disc protrusion shows extension of disc material beyond the interspace margin where the base of the herniation is wider than the portion in the epidural space ⇨.*



(Left) *Sagittal graphic of a disc extrusion shows extension of disc material beyond the interspace where the base of the herniated material is smaller than the component in the epidural space ⇨.* **(Right)** *Sagittal graphic shows a disc extrusion with free fragment. Extruded disc demonstrates extension of disc material beyond the interspace with the base narrower than the portion in the epidural space ⇨. A 2nd component ⇨ has separated from the parent disc and is a free fragment or sequestered disc.*



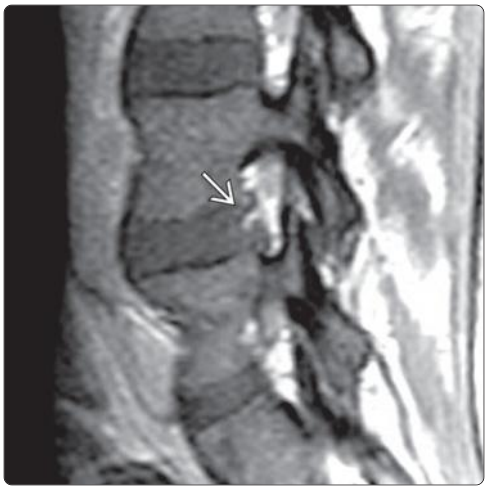
(Left) *Sagittal T1WI MR shows severe disc degeneration at L5-S1 with disc extrusion where the base of herniation is narrower than the portion extending into epidural space ⇨. There is inferior migration of herniation consistent with free fragment.* **(Right)** *Sagittal T1WI MR shows a large L3-L4 far lateral disc extrusion, which is located within the neural foramen (foraminal herniation) ⇨ and obscures the exiting L3 nerve root. Note the normal exiting root at the level below for comparison ⇨.*

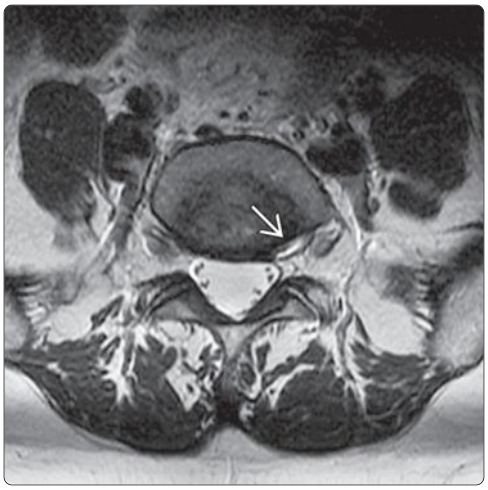




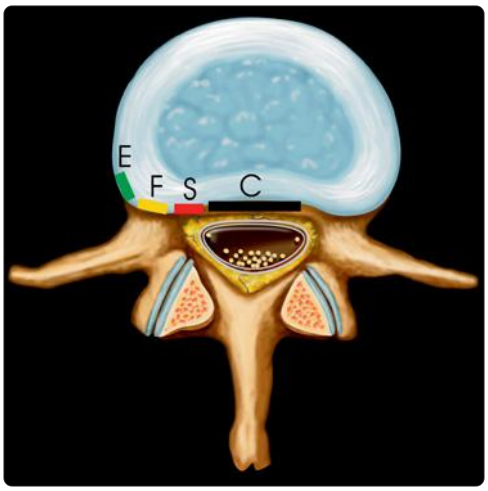


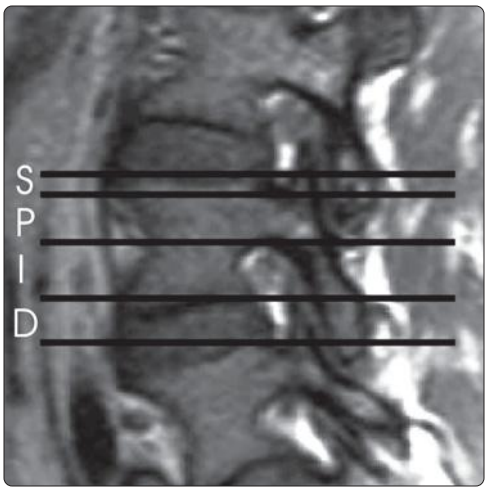
(Left) *Sagittal T2WI MR shows C5-C6 disc extrusion where the base of the herniation ➡ is smaller than the rest of the epidural component ➡.* **(Right)** *Axial T2* GRE MR shows a large left central disc extrusion with moderate compression of the thecal sac and cord ➡.*





(Left) *Sagittal T1 C+ MR shows an anular fissure as focal enhancement within the posterolateral anulus fibrosus ➡. There is no disc herniation and the foramen is normal in size. The term anular tear is to be avoided.* **(Right)** *Axial T2WI shows an anular fissure along the posterior lateral margin of the disc defined as linear hyperintensity without focal contour abnormality ➡.*





(Left) *Axial graphic of a lumbar intervertebral disc shows right-left location nomenclature classification for disc disease: C (central), S (subarticular), F (foraminal), and E (extraforaminal or far-lateral).* **(Right)** *Sagittal T1W1 MR through the neural foramen of the lumbar spine shows the level classification and nomenclature: S (suprapedicular), P (pedicular), I (infrapedicular), and D (disc).*

KEY FACTS

TERMINOLOGY

- Generalized and multifactorial process affecting discovertebral unit leading to biomechanical/morphologic alterations
- Asymptomatic or associated with back/neck pain ± radiculopathy

IMAGING

- Loss of disc space height, vacuum phenomenon seen as low signal within disc
- Degenerative endplate changes I → III
- T2 shows loss of signal from nucleus, loss of horizontal nuclear cleft
- Disc may show linear enhancement with DDD, enhancement within Schmorl nodes

TOP DIFFERENTIAL DIAGNOSES

- Disc space infection
- Hemodialysis spondyloarthropathy
- Seronegative spondyloarthropathy

PATHOLOGY

- Etiology of disc degeneration multifactorial
- Individuals involved in manual materials handling, with repeated heavy lifting, at increased risk
- Some studies show strong familial predisposition to discogenic back pain

CLINICAL ISSUES

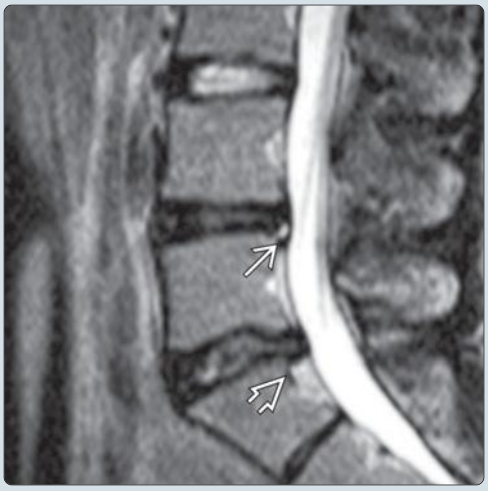
- Lifetime incidence of back pain in United States 50-80%
- Prevalence among adults ranges from 15-30%
- Back pain most common cause of disability in persons younger than 45 years
- LBP has benign natural history
  - LBP symptoms self-limited and resolve spontaneously (< 2 weeks)

(Left) *Sagittal graphic shows degeneration of L4-5 and L5-S1 intervertebral discs with loss of disc height, associated type II fatty endplate change, and osteophyte formation.* **(Right)** *Sagittal NECT reformat shows multilevel severe loss of disc height and vacuum phenomenon. Endplate eburnation is present at L3-4 and L4-5. Osteophyte is present at several levels.*





(Left) *Sagittal T2WI MR shows degenerated disc as low signal at L4-5, L5-S1 compared to normal disc signal intensity at L3-4. Degenerative disc disease is also seen as loss of disc height at L4-5, disc herniation at L5-S1 ➪, and anular tear at L4-5 ➪.* **(Right)** *Axial T2WI MR shows protrusion ➪ compressing the exiting root. Degenerated disc shows low signal centrally with disrupted internal disc contents pointing toward the herniation ➪.*





TERMINOLOGY

Synonyms

- Degenerative disc disease (DDD), disc degeneration, spondylosis

Definitions

- Generalized and multifactorial process affecting discovertebral unit leading to biomechanical/morphologic alterations
- Asymptomatic or associated with back/neck pain ± radiculopathy

IMAGING

General Features

- Best diagnostic clue
  - Decreased signal of intervertebral disc on T2WI
- Location
  - Intervertebral disc, adjacent endplates
- Size
  - Normal disc height with mild signal loss to marked loss of disc height
- Morphology
  - Nucleus/anulus alteration, with adjacent facet/foramina osteoarthritic degenerative change

Radiographic Findings

- Radiography
  - Late disease shows loss of disc space height, osteophyte formation, bony endplate eburnation "discogenic sclerosis," vacuum phenomenon

CT Findings

- NECT
  - Useful for assessment of bulge, focal disc herniation, osteophyte formation with facet arthropathy, central stenosis

MR Findings

- T1WI
  - Loss of disc space height, vacuum phenomenon seen as low signal within disc
  - Degenerative endplate changes I → III
- T2WI
  - T2 shows loss of signal from nucleus, loss of horizontal nuclear cleft
  - Degenerative endplate changes I → III
- T1WI C+
  - Disc may show linear enhancement with DDD, enhancement within Schmorl nodes

Nonvascular Interventions

- Myelography
  - Nonspecific extradural defects associated with central stenosis, osteophyte formation, herniation

Nuclear Medicine Findings

- Bone scan
  - Diffuse increased uptake with DDD, osteophyte formation, and facet degenerative arthropathy

Other Modality Findings

- Discography role limited in patients with DDD
  - Used in confirmatory role and to evaluate adjacent level disc degeneration
  - Provocative discography used for determination of levels included in fusion surgery

Imaging Recommendations

- Best imaging tool
  - MR shows intrinsic disc signal changes, anular tears, bulge, herniation, and stenosis
- Protocol advice
  - Standard T1WI and T2WI in sagittal/axial planes (4 mm maximum slice thickness)

DIFFERENTIAL DIAGNOSIS

Disc Space Infection

- Endplate destruction

Hemodialysis Spondyloarthropathy

- Low signal on T1WI, may be indistinguishable from pyogenic infection

Seronegative Spondyloarthropathy

- Inflammatory endplate changes may mimic type I degenerative endplate changes

PATHOLOGY

General Features

- Etiology
  - Etiology of disc degeneration multifactorial
  - Individuals involved in manual materials handling, with repeated heavy lifting, at increased risk
  - More controversial associations of back pain with psychosocial factors
    - Depression, mental stress, job dissatisfaction, high work pace, lack of job support
  - Cigarette smoking and obesity linked to back pain in some studies
  - Evidence of dose response relationship between physical workload and long duration low back pain (LBP)
- Genetics
  - 2 collagen IX alleles associated with sciatica and disc herniation
  - Disc degeneration associated with aggrecan gene polymorphism, matrix metalloproteinase 3 and 7 gene alleles
  - Some studies show strong familial predisposition to discogenic back pain
- Associated abnormalities
  - Bulging anulus, disc herniation, osteophyte formation, central and foraminal stenosis
  - Degenerative spondylolisthesis
    - M < F, occurs after age 40 years, 3x more common in African American women than Caucasian women, L4-L5 level
- Etiology of symptomatic DDD not fully known
- Multiple abnormalities can be present in symptomatic patients, such as disc space narrowing, endplate sclerosis, osteophytes, vacuum phenomenon

Staging, Grading, & Classification

- Classification systems
  - MR disc grading scheme I → V
  - Histologic classification system of age-related changes

Gross Pathologic & Surgical Features

- Disc abnormalities
  - Disc desiccation with decreased proteoglycan water binding
  - Collagen content increases
  - Loss of negatively charged proteoglycan side chains
  - Loss of hydrostatic properties of disc with loss of disc space height
  - Bulging of anulus with anular tears/herniation
  - Loss of boundary between anulus and nucleus
  - Nucleus becomes progressively disorganized and fibrous with fibrocartilage replacing distinct nucleus/anulus
  - Decrease in anular cells, with shift to more type II collagen
- Endplate abnormalities
  - Endplates show progressive thinning and hyalinization
  - Loss of nutritional function of endplate with decreased diffusion ability
  - Endplate marrow conversion to type I-III, end-stage of endplate bony eburnation
  - Osteophyte formation
- Facet joints
  - Loss of disc space height → rostrocaudal subluxation of facets with capsule laxity, ligamentous buckling/thickening
  - Shift of axial load pattern leading to increased facet degenerative arthropathy, osteophyte formation

Microscopic Features

- Nucleus becomes progressively disorganized and fibrous, cracks and fissures throughout nucleus
- Endplate fragmentation and disruption with new bone formation, granulation tissue
- Marrow conversion from hematopoietic to fat or fibrous (type II, type I endplate change, respectively)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Commonly asymptomatic with normal neurologic exam
  - Low back or neck pain
    - ± radiculopathy
  - Other signs/symptoms
    - Range of motion restricted
    - Extension may exacerbate pain

Demographics

- Age
  - Back pain peak 45-65 years
- Gender
  - M = F
- Epidemiology
  - Overall incidence of back pain ~ 45 per 1,000 person-years
  - Lifetime incidence of back pain in United States 50-80%
  - Prevalence among adults ranges from 15-30%
  - 2-5% of population receives medical care every year for back pain
  - 15 million physician visits in 1990 for back pain
  - 1% of population disabled because of back pain
  - Back pain most common cause of disability in persons younger than 45 years
  - Highest prevalence of back pain 40-60 years of age

Natural History & Prognosis

- LBP has benign natural history
- LBP symptoms self-limited and resolve spontaneously (< 2 weeks)
- 7% of patients with acute symptoms develop chronic pain
- Of patients complaining of chronic pain (> 3 months), 1/3 have disabling symptoms
- Over 5-year period, ~ 70% show clinical improvement

Treatment

- Nonoperative treatment with bedrest, exercise, drug therapy, manipulation, epidural steroid injection
  - Manipulation effective in treatment of LBP of short duration
- Operative treatment
  - Surgical treatment for axial pain from DDD most commonly involves fusion

DIAGNOSTIC CHECKLIST

Consider

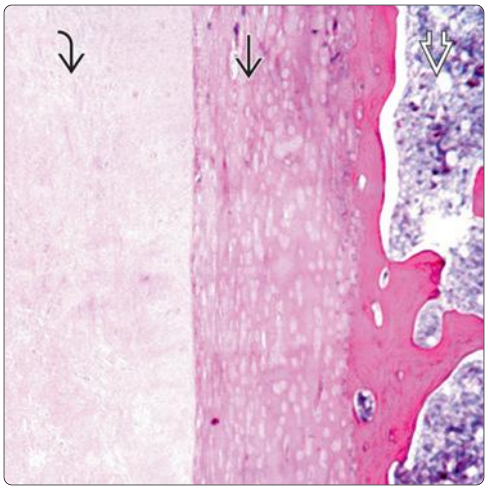
- 30% of normal individuals have abnormal signal in lumbar discs

Image Interpretation Pearls

- Discography should consist of 4 data points
  - Morphology of disc injected
  - Disc pressure/volume of fluid accepted
  - Patient subjective pain response
  - Lack of pain response at adjacent levels

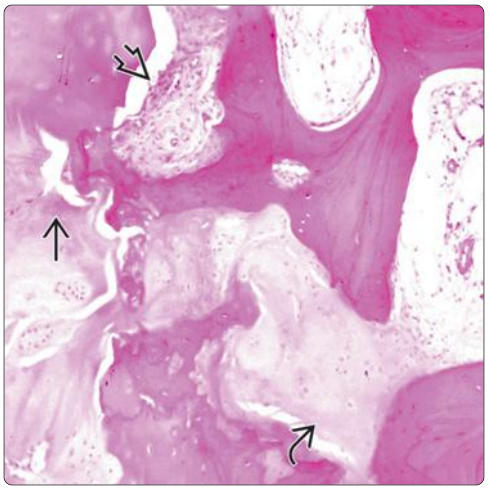
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(Left) *H&E stained section of normal discovertebral junction shows a normal endplate with nucleus, smooth cartilage junction, thin bony trabeculae, and normal hematopoietic marrow.* **(Right)** *Sagittal T2WI MR shows multiple aspects of DDD, including Schmorl node involving inferior endplates of L3 and T12, diffuse disc degeneration with loss of height at multiple levels, severe multilevel central canal stenosis with bulging discs effacing the thecal sac, and multilevel ligamentous hypertrophy.*





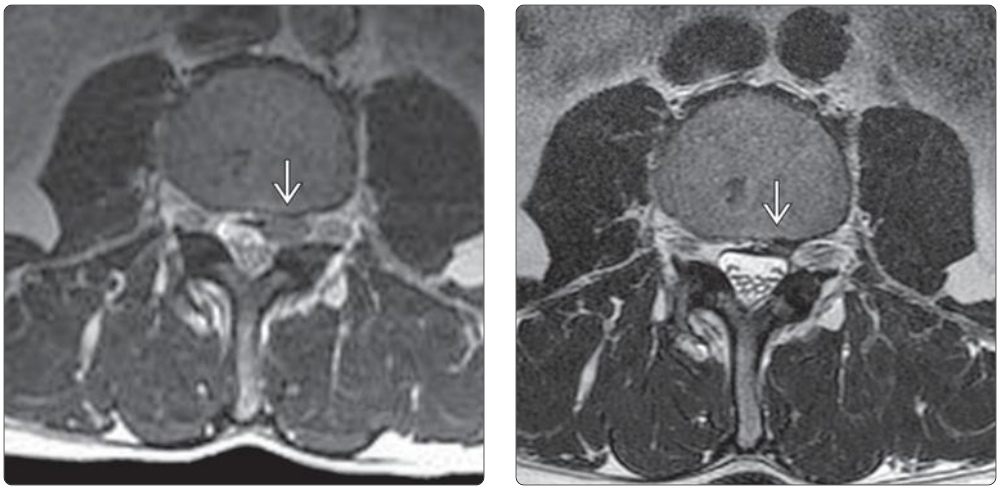
(Left) *Micropathology, low-power H&E of severely degenerated discovertebral junction shows a degenerated endplate with disruption of cartilage, fissures in nucleus, granulation tissue, and cartilage endplate herniation.* **(Right)** *Axial discogram CT shows contrast extension from the central nucleus pulposus into a midline anular tear extending to the epidural space.*



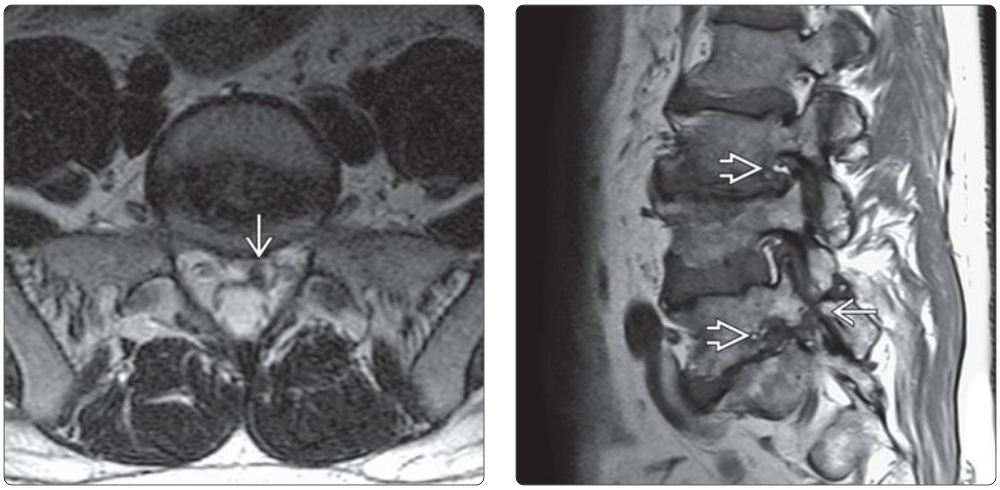


(Left) *Sagittal T1WI MR shows prominent type II endplate change at L4-L5 (fatty marrow) with marked loss of disc space height. The adjacent level L3-L4 shows type I change as low signal marrow with slight spondylolisthesis.* **(Right)** *Sagittal STIR MR level shows L3-L4 pronounced type I degenerative endplate change. There is a slight anterolisthesis of L3 on L4 and multilevel disc degeneration.*

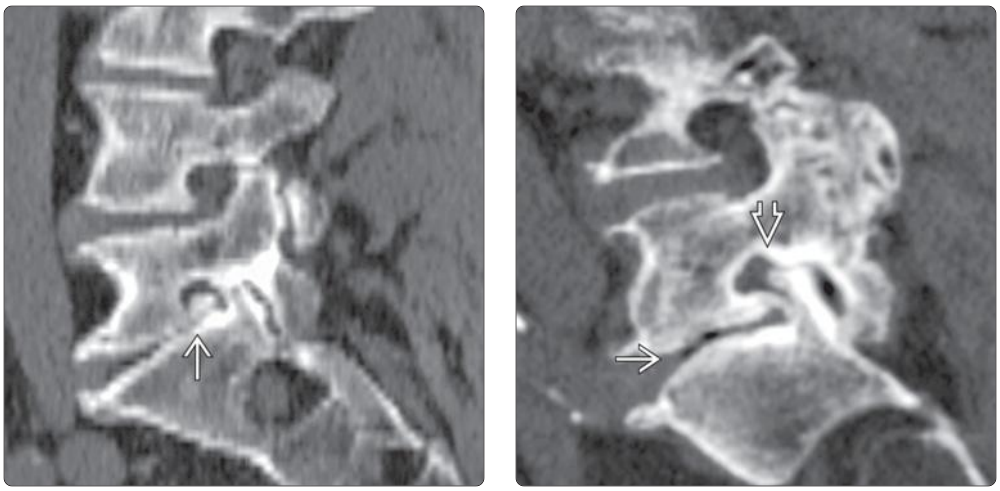
(Left) *Axial T2WI MR at presentation shows a large left-sided disc extrusion with thecal sac compression in this case of natural involution of large disc extrusion.* **(Right)** *Follow-up axial T2WI MR after conservative therapy shows a markedly decreased size of the previously large disc extrusion seen on the previous image.*



(Left) *Axial T2WI MR study 7 months after the initial diagnosis of typical disc extrusion shows well-defined fluid signal intensity that has replaced the disc herniation, consistent with a "discal cyst". This may reflect involution of a prior hemorrhage with the disc herniation.* **(Right)** *Sagittal T1WI MR shows chronic L5 spondylolysis with severe multilevel foraminal stenosis with extension of disc bulge and osteophyte into the inferior neural foramina.*



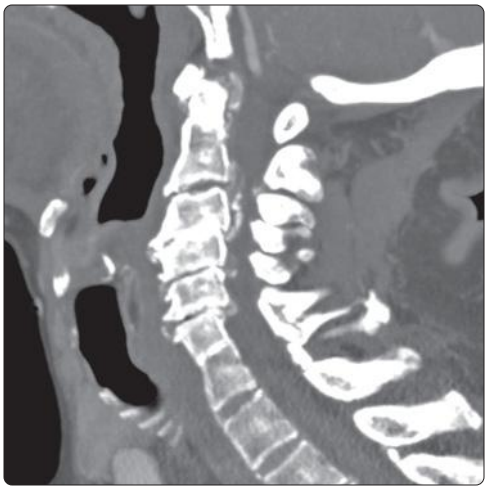
(Left) *Sagittal NECT shows loss of disc height at L5-S1 with resultant rostrocaudal subluxation of facets and foraminal stenosis. Osteophyte causes further stenosis extending into the foramen.* **(Right)** *Sagittal NECT reformat shows loss of disc space height at L5-S1 with vacuum phenomenon. Marked facet hypertrophic degenerative arthropathy gives severe foraminal stenosis.*







(Left) *Sagittal T1WI MR shows severe multilevel degenerative scoliosis with foraminal stenosis and L5-S1 spondylolysis. There is disc degeneration at every level with height loss and retrolisthesis.* **(Right)** *Sagittal T2WI MR in a patient with severe multilevel degenerative scoliosis shows multiple T2 hyperintense intervertebral discs as a manifestation of disc degeneration, not infection.*





(Left) *Sagittal CECT shows a classic pattern of multilevel cervical spondylosis with severe central stenosis with marked loss of disc height/bony eburnation from C2-C3 to C5-C6. There is associated disc and posterior longitudinal ligament calcification.* **(Right)** *Sagittal STIR MR shows severe cervical spondylosis with multilevel subarachnoid space narrowing with the abnormal cord T2 signal at C4-C5. An incidental finding is the congenital block vertebra at C6-C7.*





(Left) *Sagittal STIR MR shows low signal degenerative pseudopannus dorsal to the odontoid, with compression of the anterior thecal sac and ventral cord. There is diffuse subaxial degenerative disc disease with fusion of C5-C6.* **(Right)** *Sagittal T2WI MR shows multilevel cervical spondylosis, worst at the C5-C6 and C6-C7 levels. Both levels show central canal stenosis with disc osteophyte complexes, which efface the ventral cord. There is myelomalacia of the cord at C6-C7.*

Degenerative Endplate Changes

KEY FACTS

TERMINOLOGY

- Vertebral endplate change, Modic changes
  - Type I, II, III endplate change
- MR signal abnormalities involving vertebral body endplates related to degenerative disc disease
  - Type I: Hypointense on T1, hyperintense on T2
  - Type II: Hyperintense on T1, isointense on T2
  - Type III: Hypointense on T1 and T2

TOP DIFFERENTIAL DIAGNOSES

- Disc space infection
- Pseudoarthrosis/nonunion
- Seronegative spondyloarthropathy
- Metastatic disease

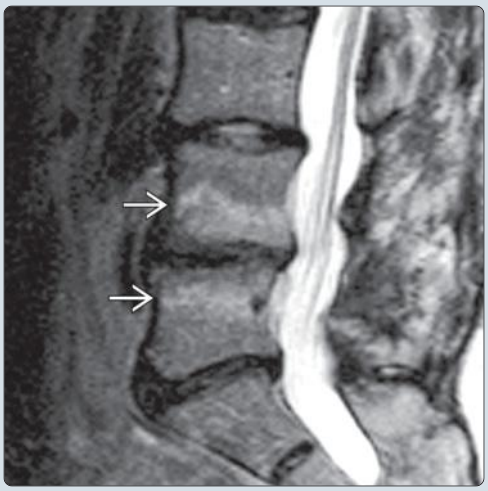
PATHOLOGY

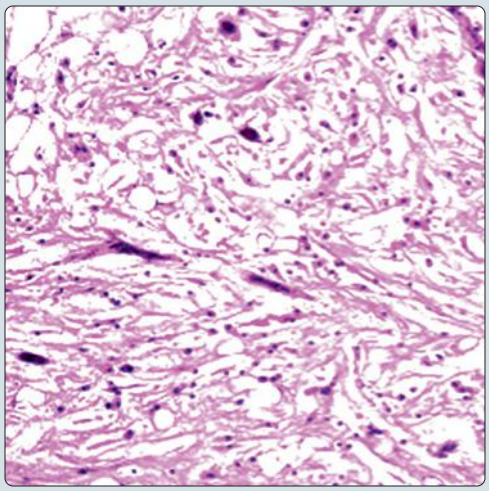
- Type I: Replacement with fibrovascular marrow
- Type II: Replacement by fatty marrow
- Type III: Replacement by bony sclerosis with little residual marrow

CLINICAL ISSUES

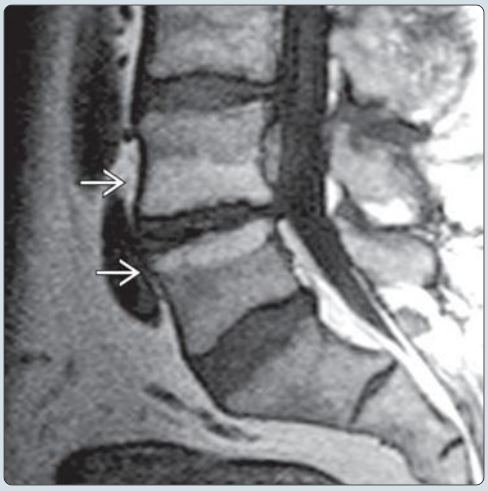
- Type I: Present in 4% of patients undergoing MR for disc disease
- Type II: Present in 16% of patients undergoing MR for disc disease
  - Commonly present after successful segmental fusion
- Type III: Least common (~ 1%)
- Relationship of type I endplate changes to low back pain (discogenic pain) controversial
  - Some studies suggest type I, II change have high specificity (> 90%) but low sensitivity (20-30%) for painful lumbar disc
  - Type I change shows high positive predictive value with discography

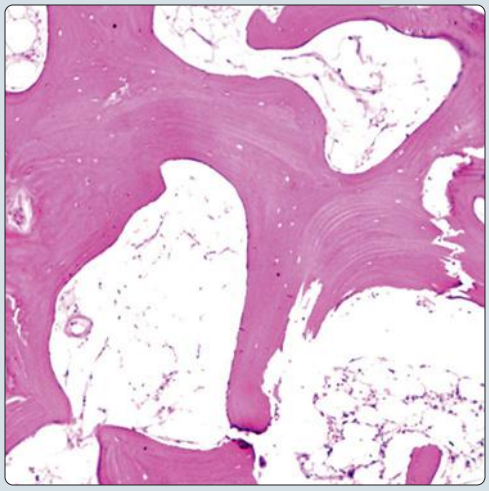
(Left) *Sagittal T2WI MR shows increased signal intensity from L4-L5 endplates ⇒. The disc at L4-L5 is low signal, reflecting disc degeneration with a slight retrolisthesis. No prevertebral or epidural soft tissue is present to suggest infection.* **(Right)** *Micropathology, low-power H&E shows type I change with fibrovascular replacement of hematopoietic marrow. There are prominent interstitial spaces and scattered capillaries among spindle-shaped cells.*





(Left) *Sagittal T1WI MR shows classic type II endplate changes ⇒ due to fatty marrow replacement at L4-L5 disc level. There is degeneration of L4-L5 with a loss of disc height and vacuum phenomenon.* **(Right)** *Micropathology, low-power H&E shows type II changes with fatty replacement of normal hematopoietic marrow. Note the thickened trabeculae with new woven bone formation.*





TERMINOLOGY

Synonyms

- Vertebral endplate change, Modic changes
- Type I, II, III endplate change

Definitions

- MR signal abnormalities involving vertebral body endplates related to degenerative disc disease
  - Type I: Hypointense on T1WI, hyperintense on T2WI
  - Type II: Hyperintense on T1WI, isointense on T2WI
  - Type III: Hypointense on T1WI and T2WI

IMAGING

General Features

- Best diagnostic clue
  - Parallel signal alteration of vertebral endplates, associated with evidence of disc degeneration
- Location
  - Most common in lumbar spine, may occur in any vertebral body
  - L4-L5, L5-S1 most common
- Size
  - Variable: Small linear bands to near-complete vertebral body involvement
- Morphology
  - Linear, parallel to intervertebral disc

Radiographic Findings

- Radiography
  - Insensitive to early changes, type III change seen as "discogenic" sclerosis of endplates

CT Findings

- NECT
  - Insensitive to early changes, type III seen as endplate sclerosis

MR Findings

- T1WI
  - Type I: Hypointense horizontal bands involving endplates
  - Type II: Hyperintense bands
  - Type III: Hypointense bands
- T2WI
  - Type I: Hyperintense horizontal bands involving endplates
  - Type II: Isointense to slightly hyperintense bands
    - Signal tracks fat on all pulse sequences
  - Type III: Hypointense bands involving endplates
- T1WI C+
  - Type I: May show prominent enhancement
  - Commonly associated with linear intervertebral disc enhancement

Nuclear Medicine Findings

- Bone scan
  - Nonspecific increased uptake with all degenerative disc disease
- PET
  - Hypometabolic with degenerative endplate changes
  - Hypermetabolic with disc space infection or vertebral osteomyelitis
  - PET accurate and specific to differentiate degenerative endplate change from infection
- Ga-67 scintigraphy
  - No increased uptake with degenerative endplate changes

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal T1WI and T2WI defines lesions and classification

DIFFERENTIAL DIAGNOSIS

Disc Space Infection

- Low signal involving endplates similar to type I change
- Endplate destruction
- Hyperintense intervertebral disc on T2WI
- Paravertebral or epidural soft tissue phlegmon/abscess

Pseudoarthrosis/Nonunion

- Low signal involving endplates similar to type I change
- Irregular endplates

Hemodialysis Spondyloarthropathy

- Low signal on T1WI involving endplates similar to pyogenic disc space infection
- May be indistinguishable from pyogenic disc space infection

Seronegative Spondyloarthropathy

- Inflammatory endplate changes (Anderson lesions) may mimic type I degenerative endplates
- Typical squaring of vertebral bodies
- Late changes with fusion of bodies and posterior elements

Metastatic Disease

- Focal low signal on T1WI with bone destruction ± epidural extension
- Low signal does not parallel endplates as in type I change

PATHOLOGY

General Features

- Etiology
  - Unknown
    - Type I: Probably reflects sequelae of acute disc degeneration
      - Present in 30% of chymopapain-treated discs (good model for acute disc degeneration)
    - Type II: Probably reflects sequelae of chronic disc degeneration
    - Type III: Probably reflects sequelae of chronic disc degeneration
    - Controversial potential cause is low-grade infection (Propionibacterium)
- Genetics
  - Interleukin 1 gene locus polymorphisms reported
- Disc degeneration
  - Loss of disc space height
  - Loss of signal from disc on T2WI
- Replacement of normal hematopoietic marrow

- Type I: Replacement with fibrovascular marrow
- Type II: Replacement by fatty marrow
- Type III: Replacement by bony sclerosis with little residual marrow

Staging, Grading, & Classification

- Type I, II, III classification is reliable and reproducible
  - Excellent interobserver agreement

Gross Pathologic & Surgical Features

- Disc degeneration with desiccation of disc, bony endplate eburnation

Microscopic Features

- Type I: Spindle cells, capillaries (vascularized fibrous tissue) with prominent interstitial space, thickened trabeculae with new bone production
- Type II: Adipose cells, prominent trabeculae with new bone production (woven bone)
- Type III: Dense woven bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Role of endplate changes in back pain is controversial
  - Nonspecific neck or back pain
  - Relationship of type I endplate changes to low back pain (discogenic pain) controversial
    - No significant correlation between concordant pain at discography and presence of endplate changes in some studies
    - Some studies suggest type I, type II change have high specificity (> 90%) but low sensitivity (20-30%) for painful lumbar disc
    - Some studies show type I correlation with pain (> 70%) but low with type II (10%)
    - Type I change shows high positive predictive value with discography
  - Type II may imply more stable discovertebral unit
    - Commonly develops after successful segmental surgical fusion
  - Chronic antibiotic treatment (placebo-controlled trial) significantly more effective for chronic low back pain associated with Modic I than placebo
- Clinical profile
  - Low back pain without major radicular component

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - Adult with disc degeneration
  - Type I: Present in 4% of patients undergoing MR for disc disease
  - Type II: Present in 16% of patients undergoing MR for disc disease
  - Type III: Least common (~ 1%)
    - Mixed lesions are possible (I/II and II/III)

Natural History & Prognosis

- Natural history variable
  - Type I change may convert to type II over course of months to years
  - Type II more stable change
    - Up to 14% of type II may convert over 3-year follow-up

Treatment

- Options, risks, complications
  - Degenerative endplate changes may be asymptomatic
  - Controversial as to role of endplate changes regarding spinal fusion
  - Endplate changes proposed as 1 criterion for spinal fusion for discogenic pain

DIAGNOSTIC CHECKLIST

Consider

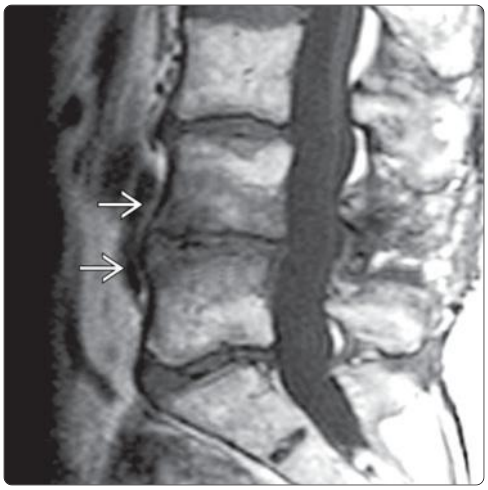
- Disc space infection if high signal within disc on T2WI
- Disc degeneration may also show increased signal within disc on T2WI due to cracks, fissures

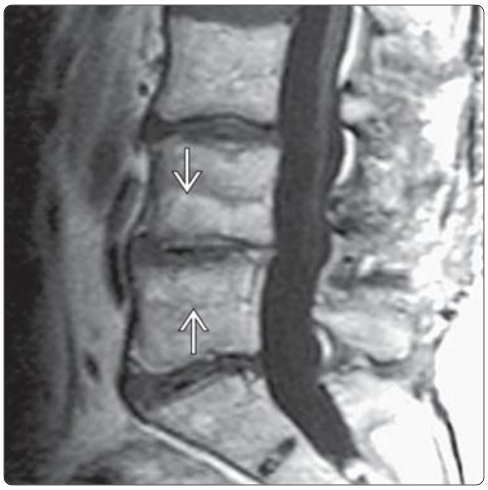
Image Interpretation Pearls

- Discrimination of early disc space infection and severe type I endplate changes may be difficult or impossible
- Degenerative endplate changes do not show paravertebral enhancing soft tissue, which is often present with disc space infection
- Use axial post-contrast fat-suppressed T1WI to better define paravertebral soft tissues

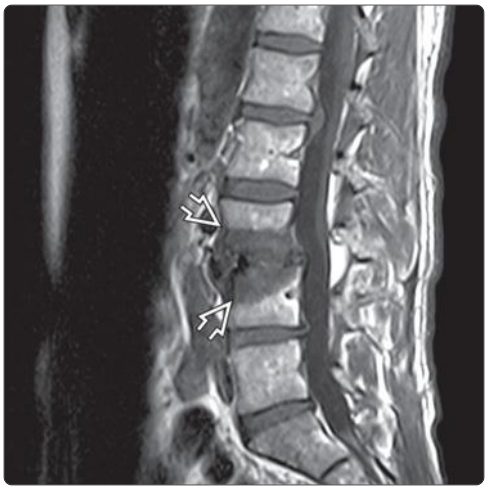
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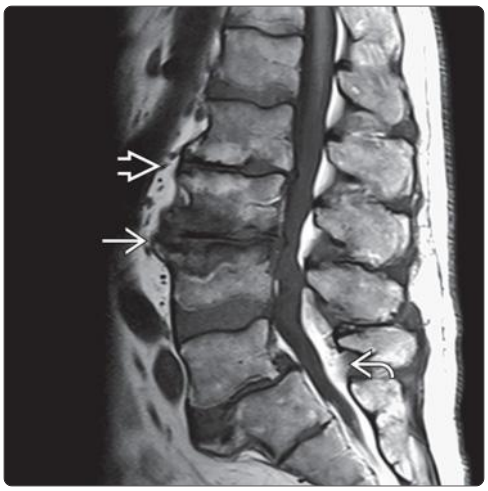


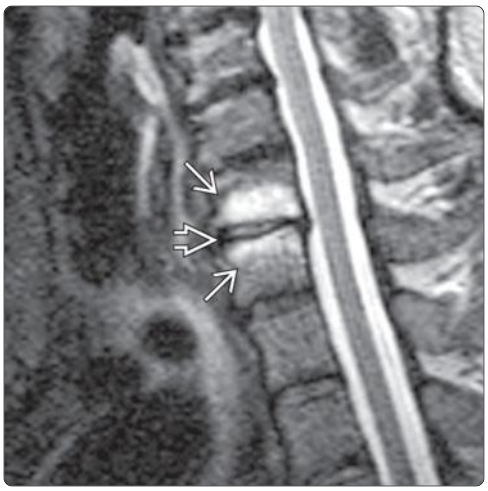
(Left) *Sagittal T1WI MR shows low signal involving L4-L5 endplates with vacuum phenomenon within several disc spaces due to degenerative change. Low signal endplate margins appear intact.* **(Right)** *Sagittal T1WI C+ MR shows prominent enhancement of the type I change at L4-L5. Mild linear enhancement due to disc degeneration is present within the disc itself, extending posteriorly to the anulus.*





(Left) *Sagittal T1WI MR shows aggressive type I endplate change at L3-L4 with grade I anterolisthesis, narrowed disc, irregular endplates, and hypointensity in adjacent bodies. Note the lack of bone destruction and paraspinous mass that would suggest infection.* **(Right)** *Sagittal T2WI MR shows endplate irregularity, thinned disc, and hyperintensity in the vertebral bodies. Severe endplate changes may mimic infection. Clinical correlation or biopsy may be required to make the distinction.*





(Left) *Sagittal T1WI MR shows type III endplate changes at L3-L4 and type II changes at L2-L3. There is spondylolisthesis and spondylolysis at L5-S1.* **(Right)** *Sagittal T2WI MR shows type I endplate changes at C5-6 level with increased signal. Slight increased signal is present within the disc itself due to disc degeneration with fissuring. No prevertebral soft tissue is present.*

KEY FACTS

TERMINOLOGY

- Joint-based abnormality due to cartilage damage

IMAGING

- Lateral atlantoaxial joints > atlantoodontoid joint > occipitocervical joint
  - Lateral atlantoaxial involvement typically unilateral
- Joint space narrowing, subchondral sclerosis, osteophytes
- Varying degrees of lateral mass collapse
- Ligament laxity + ligamentous hypertrophy
- Osteocartilaginous bodies (ossicles)
- Degenerative pannus
  - Variable subchondral signal changes on MR
- Imaging
  - Open mouth odontoid view is best screening
  - CT optimal for preoperative evaluation
  - Flexion/extension lateral films or CT to assess stability

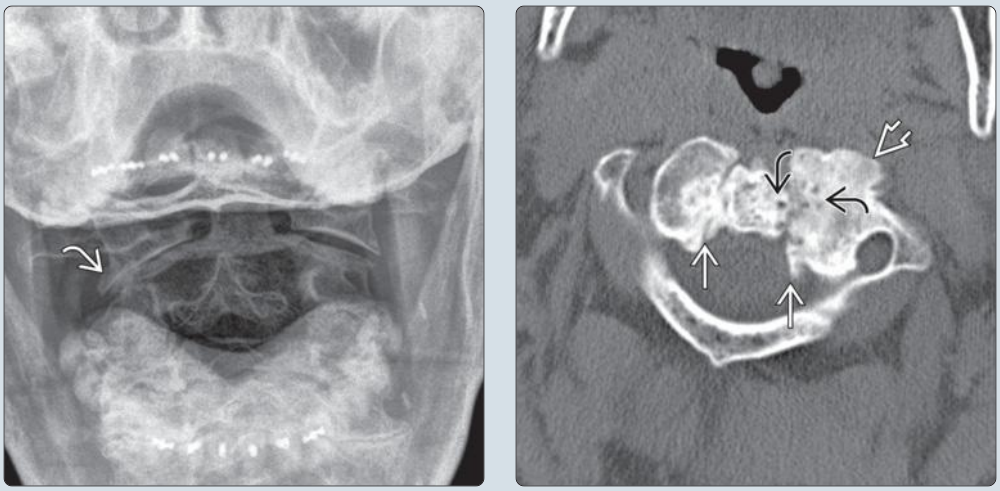
TOP DIFFERENTIAL DIAGNOSES

- Rheumatoid arthritis
  - Bilateral symmetric disease
  - Osteopenia, erosions
  - No subchondral sclerosis, osteophytes
- Calcium pyrophosphate deposition disease
  - Atlantoodontoid joint
  - Extensive mineralization

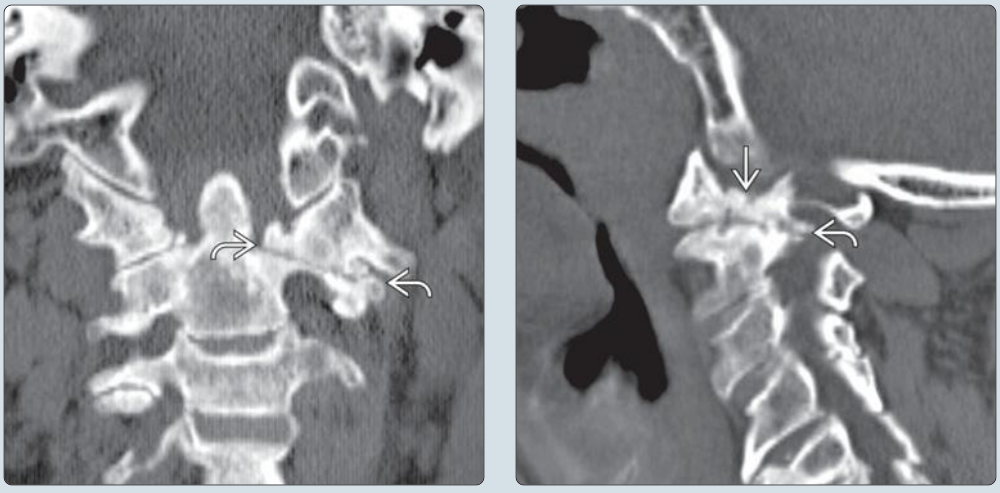
CLINICAL ISSUES

- F > M
- Increasing incidence with increasing age
- Pain: Occipital, occipitocervical, unilateral
  - Headache
  - Associated point tenderness ± crepitus
- Limited neck range of motion, especially rotation
- Myelopathy: Associated with subluxation; mass effect from ligament and synovial hypertrophy

(Left) *AP open mouth odontoid radiograph shows osteoarthrosis (OA) of the right atlantoaxial joint, which is significantly narrowed with a large osteophyte. Note that within the joint, narrowing is not uniform; it is more severe laterally. Compare to the normal left articulation.* **(Right)** *Axial bone CT shows arthritic changes in the atlantodental articulation with joint space narrowing, small subchondral cysts, sclerosis, and osteophytes. Hypertrophic changes from atlantoaxial OA are also evident.*



(Left) *Coronal reformatted bone CT shows advanced atlantoaxial OA. Both atlantoaxial articulations are involved, and the disease is asymmetric (left > right). Extensive osteophyte formation (on left) involves both medial and lateral joint margins.* **(Right)** *Sagittal reformatted bone CT shows severely osteoarthritic lateral atlantoaxial articulation. The joint is markedly narrowed with extensive sclerosis, articular surface irregularity, and osteophyte formation. Note height loss within the lateral mass.*



TERMINOLOGY

Abbreviations

- Degenerative joint disease (DJD)

Synonyms

- Osteoarthrosis (OA)
- Osteoarthritis
  - Out of favor; OA is not an inflammatory process

Definitions

- Joint-based abnormality secondary to cartilage damage

IMAGING

General Features

- Best diagnostic clue
  - Joint space narrowing, subchondral sclerosis, osteophytes
- Location
  - Midline atlantoodontoid joint is common
    - May be isolated process or associated with lateral atlantoaxial OA
  - Lateral atlantoaxial joint disease is uncommon
    - Lateral atlantoaxial involvement is typically unilateral
      - Asymmetric bilateral OA less common
      - Bilateral disease may be secondary OA, often from trauma or its sequelae
  - Occipitocervical joint disease is least common

Radiographic Findings

- Joint space narrowing
- Subchondral sclerosis
- Osteophytes
- Advanced cartilage loss and wear of subchondral bone may produce lateral mass collapse
  - Loss of lateral mass height leads to cranial settling
- Subchondral cysts can be present and of variable size
  - Large odontoid cysts increase fracture risk
- Associated subaxial degenerative changes are common

CT Findings

- Mirror radiographic findings and provide better detail
- Osteocartilaginous bodies (ossicles) are better seen
  - Variable sizes
  - Found in recesses at joint periphery
- Transverse ligament calcification is best seen with CT
  - Common feature of OA
  - Nonspecific

MR Findings

- Most useful to exclude other etiologies and evaluate spinal cord status
- Subchondral signal changes are variable
  - Edema-like
    - Decreased signal on T1WI
    - Increased signal on fluid-sensitive images
  - Sclerosis/fibrosis
    - Decreased signal on T1WI and fluid-sensitive images
  - Subchondral signal may have mixed appearance
- Capsular and ligamentous hypertrophy
- Ligament laxity secondary to lateral mass collapse
- Osteocartilaginous bodies (ossicles)
- Degenerative pannus (hypertrophied synovium)
  - Tumor-like appearance
  - Isointense signal on T1WI
  - Iso- to hypointense on T2WI
  - Cystic foci hypointense on T1WI and hyperintense on T2WI
  - Mass effect leads to spinal cord compression

Nuclear Medicine Findings

- Bone scan
  - Focal intense uptake seen unilaterally
    - Even with SPECT, may need CT to differentiate occipitocervical and atlantoaxial disease

C1-C2 Joint Injection

- Diagnostic and therapeutic
- Fluoroscopic or CT guided
- Posterolateral approach to avoid vertebral arteries
- Contralateral joint may opacify

Imaging Recommendations

- Best imaging tool
  - Open mouth odontoid view is best screening
  - CT is optimal for preoperative evaluation
    - Coronal and sagittal CT reconstructions provide best visualization
  - Flexion/extension lateral films and CT can be used to assess stability

DIFFERENTIAL DIAGNOSIS

Rheumatoid Arthritis (RA), Adult

- Distribution is bilateral and symmetric
- Osteopenia is key feature
  - Periarticular &/or generalized
- Erosions common
- No subchondral sclerosis, osteophytes, ossicles
  - Degenerative changes may be seen if RA is inactive and secondary OA develops
- Expect other joint involvement
  - Hands and feet are commonly affected
  - Hint: Check for temporomandibular joint involvement on cervical spine films

Calcium Pyrophosphate Deposition Disease (CPPD)

- Atlantoodontoid joint is more commonly involved than atlantoaxial joints
- Extensive mineralization within and around joint
  - Speckled, amorphous mineralization within joint
  - Calcification within transverse ligament and joint capsule
- Pseudotumor is common feature, may mimic pannus
- Subcortical cysts, especially on odontoid, are more numerous than OA and can be quite large
- Limited osteophyte formation

Pseudotumor

- MR appearance mimics pannus
- CT helps differentiate
  - No joint space narrowing, subchondral sclerosis, osteophytes
- May represent isolated manifestation of CPPD

PATHOLOGY

General Features

- Decreased proteoglycans and collagen within cartilage leads to decreased shock-absorbing capability
- Continued wear results in cartilage damage including generalized thinning, fissures, and partial and full thickness defects
  - Changes typically not directly detectable on imaging studies
    - Joint space narrowing is associated imaging finding
    - Cartilage in these joints is too thin to directly assess
  - Initially, cartilage swelling and fibrillation; may see attempts at cartilage repair
  - Fibrocartilage often fills areas of articular cartilage loss
- Loss of cartilage results in decreased shock-absorbing function and increased stress on subchondral bone
- Subchondral bone responds to stress with hypervascularity, microtrabecular fracture, and new bone formation
  - Hypervascularity is associated with edema-like changes on MR
  - New bone formation appears as subchondral sclerosis
  - Microtrabecular fractures contribute to bone collapse
  - Foci of avascular bone will be present
- At joint margin, hypervascularity contributes to cartilage overgrowth, which progresses to new bone formation creating osteophytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain
    - Unilateral on same side as disease
    - Radiating to occiput or occipitocervical region
    - Point tenderness
    - Crepitus
    - May also present as headache, especially in postauricular location
  - Intraosseous degenerative dens cysts and retro-dens synovitis (pseudopannus) are risk factors for dens fractures
    - Elderly patients with dens fractures 8x more likely to have intraosseous dens cyst and 5x more likely to have synovitis compared with those without fracture
- Other signs/symptoms
  - Limited neck range of motion, especially rotation
    - Diminished lateral flexion results from associated subaxial disease
  - Myelopathy
    - Results from joint subluxation or from mass effect from atlantoodontoid pannus

Demographics

- Age
  - Increasing incidence with age
    - 6th decade: 5.4%
    - 9th decade: 18.2%
- Gender
  - F > M

Natural History & Prognosis

- Associated with OA in other sites
  - Common other sites include subaxial cervical spine, lumbar spine, hip, knee

Treatment

- Usually conservative
  - Nonsteroidal anti-inflammatory agents
  - Soft collar
- Steroid injection: Diagnostic and therapeutic
  - Diagnostic information to confirm joint as pain source
  - Therapeutic benefit provides weeks to months of relief
- Surgery (fusion) when subluxation and neurologic symptoms are present
- Surgery (decompression and fusion) when myelopathic symptoms from pannus are present
  - Over time, fusion alone may produce regression of pannus
  - Decompression is typically transoral; fusion is posterior

DIAGNOSTIC CHECKLIST

Consider

- Consider OA in older patients with unexplained headache, especially with postauricular, occipital, or occipitocervical pain

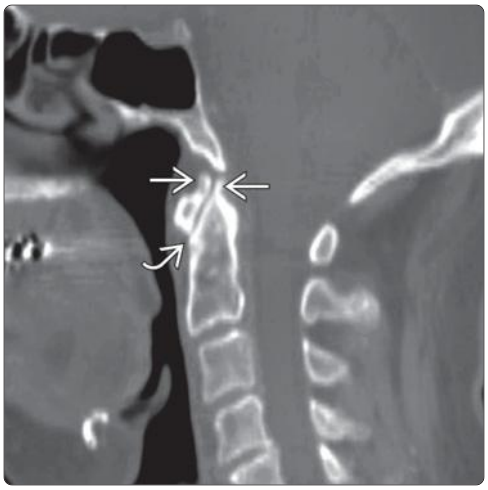
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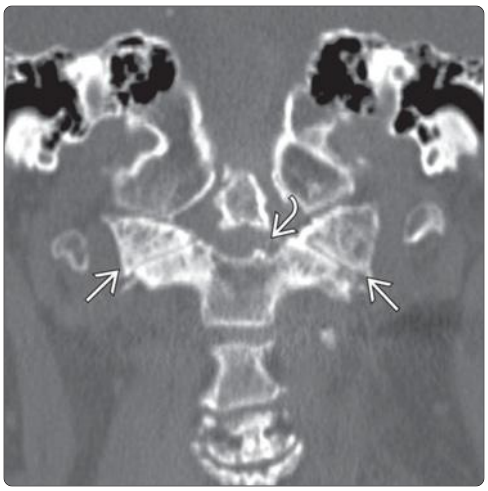
(Left) *Lateral atlantoaxial OA is easily appreciated on this coronal T1WI MR. The joint is narrowing and appears elongated secondary to osteophytes. Extensive subchondral low signal is present along both sides of the joint.* **(Right)** *Sagittal T2WI MR demonstrates advanced OA of the lateral atlantoaxial joint. The joint space is narrowed. Osteophytes are present at the joint margins, and an extensive edematous-like signal is present mixed with subchondral foci of low signal.*





(Left) *Sagittal reformatted bone CT shows that narrowing of the atlantodental interval is present, accompanied by osteophyte formation superiorly. The lack of mineralization and the presence of osteophytes help exclude CPPD as the etiology.* **(Right)** *Sagittal T1WI MR in the midline shows a large osseous structure superior to the body of C1. The structure is a large osteophyte at the margin of the midline articulation. The extensive low signal within the dens is marrow edema.*





(Left) *On this sagittal T2WI MR, the body of C1 and the osteophyte are difficult to appreciate. What can be appreciated is the thickening of the transverse ligament and synovial hypertrophy, creating slight mass effect.* **(Right)** *Coronal reformatted bone CT shows bilateral, symmetric OA of the lateral joints, which is uncommon. In this case, it is likely due to excessive motion from the nonunited dens fracture. The occipitocervical articulations also have mild osteoarthritic changes.*

Disc Bulge

KEY FACTS

TERMINOLOGY

- Anular bulge
- Generalized extension of disc beyond edges of vertebral ring apophyses

IMAGING

- Circumferential disc "expansion" beyond confines of vertebral endplates
  - Short radius of extension: ≤ 3 mm
  - > 25% of disc circumference
  - If morphologic abnormality is < 25% of disc circumference, then it is a herniation and not a bulge
- Smooth ventral extradural defect in contrast column, with indentation on anterior thecal sac
  - Central canal and subarticular recesses usually not compromised unless associated with ligamentous hypertrophy
- T1WI and T2WI MR with sagittal and axial planes
- Discography may help identify symptomatic disc

TOP DIFFERENTIAL DIAGNOSES

- Disc protrusion (< 25% of disc circumference)
- Ossification of posterior longitudinal ligament
  - Most common in cervical spine (70%)
- Vertebral endplate bony spur
  - Continuous with vertebral endplate

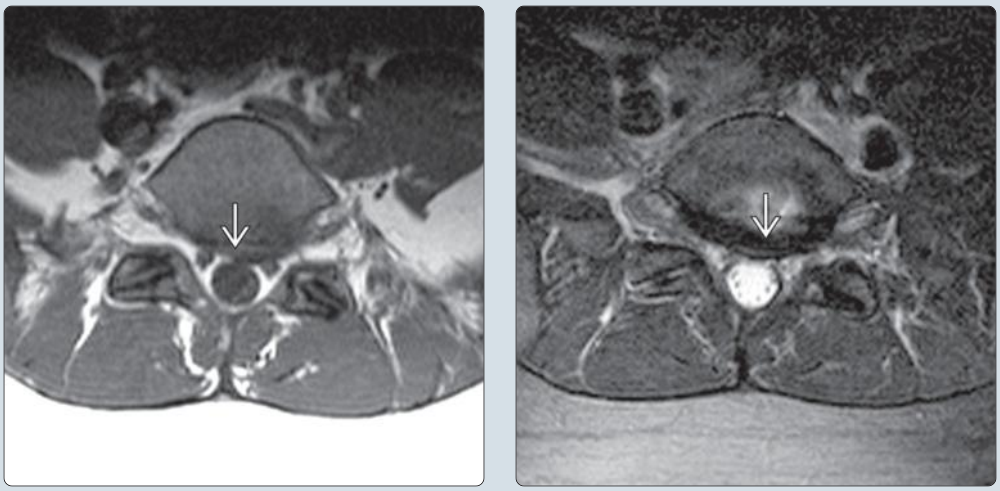
PATHOLOGY

- Bulge less important as separate entity, but associated with disc degeneration and anular fissures → "discogenic" pain

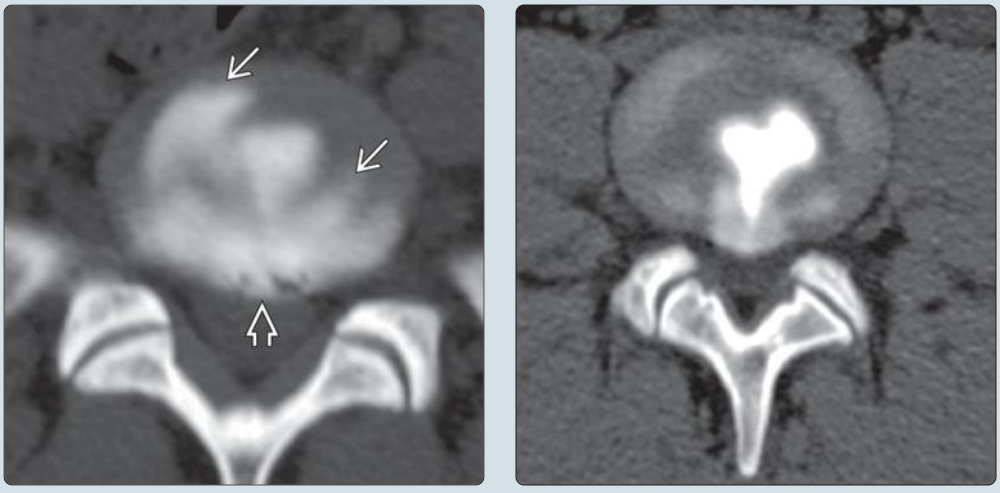
CLINICAL ISSUES

- Low back pain
- Up to 39% of asymptomatic adults have bulging discs
- > 80-90% success rate with conservative treatment

(Left) *Axial T1WI MR shows the typical pattern of a lumbar broad-based vertebral disc bulge at L5-S1. The disc extension beyond the vertebral body margin is diffuse, with no focal protrusion.* **(Right)** *Axial T2WI FS MR demonstrates the broad-based appearance of the bulging disc with predominately low signal intensity. Associated anular fissure would show focal high signal.*



(Left) *Axial NECT after discography shows multiple circumferential annular tears. These coalesce into a large dorsal annular defect, allowing contrast extending beyond annulus. Note the generalized extension of the posterior disc margin into the epidural space.* **(Right)** *Axial NECT after discography shows a radial tear extending into middle 1/3 of dorsal annulus with associated focal annular bulge into the ventral epidural space.*



TERMINOLOGY

Synonyms

- Anular bulge, annular bulge, bulging disc

Definitions

- Generalized extension of disc beyond edges of vertebral ring apophyses
  - Greater than 25% of disc circumference
  - Typically extends less than 3 mm beyond edge of apophyses

IMAGING

General Features

- Best diagnostic clue
  - Circumferential disc "expansion" beyond confines of vertebral endplates
- Location
  - Cervical: C5-C6 and C6-C7 most common
  - Lumbar: L4-L5 and L5-S1 most common
- Size
  - Short radius of extension: ≤ 3 mm
- Morphology
  - Broad based
    - > 25% of disc circumference
    - If morphologic abnormality is < 25% of disc circumference, then it is a herniation and not a bulge

Radiographic Findings

- Radiography
  - Not visible, but associated with degenerative changes
    - Disc height loss, endplate sclerosis
    - Facet arthropathy
    - Degenerative spondylolisthesis

CT Findings

- NECT
  - Smooth broad-based radial disc bulge
  - Vacuum disc phenomenon
- Bone CT
  - Better osseous detail with sagittal and coronal reformation

MR Findings

- T1WI
  - Hypointense bulging disc beyond vertebral body margin
  - Disc height loss
- T2WI
  - Hypointense bulging disc
    - Desiccation and fibrosis
    - Initially seen as linear hypointensity
    - Mild hypointensity likely related to normal aging
- T1WI C+
  - Degenerative change with enhancing anular tears
  - Enhancing fibrovascular marrow in adjacent vertebral endplates

Nonvascular Interventions

- Myelography
  - Smooth ventral extradural defect in contrast column, with indentation on anterior thecal sac
    - Central canal and subarticular recesses usually not compromised
  - Bulging disc accentuated by upright myelography
    - Especially at L4-L5

Nuclear Medicine Findings

- Bone scan
  - Uptake of radiotracer in endplates and posterior elements from degenerative changes
  - No direct imaging of anular disease

Other Modality Findings

- **Discography**
  - Indications
    - Failed conservative therapy
    - Normal or equivocal MR findings
    - To correlate disc pathology with symptoms
    - In absence of disc herniation
  - Pain responses
    - No pain, dissimilar pain, similar pain, exact pain
    - Concordant response: Absent painful response in control disc
    - Exact pain in symptomatic disc
  - Extension of contrast into anulus
    - Concentric or circumferential tear
    - Radial tear
  - Dallas discogram description
    - Grade 0: Contrast confined within nucleus pulposus
    - Grade 1: Inner 1/3 of anulus
    - Grade 2: Middle 1/3
    - Grade 3: Outer 1/3
      - Focal or < 30° of disc circumference
    - Grade 4 (April and Bogduk): Outer 1/3 of anulus
      - > 30° of disc circumference
    - Grade 5 (Schellhas)
      - Extension of contrast beyond anulus
      - Full thickness tear
      - Focal or diffuse

Imaging Recommendations

- Best imaging tool
  - T1WI and T2WI MR with sagittal and axial planes
  - Discography may help identify symptomatic disc

DIFFERENTIAL DIAGNOSIS

Disc Protrusion

- Focal contour abnormality, < 25% of disc circumference
- May exceed 3 mm beyond the edges of the ring apophyses

Ossification of Posterior Longitudinal Ligament

- Most common in cervical spine (70%)
- Continuous, segmental, mixed, or retrodiscal
- Hypointense on T1WI and T2WI
- Vertical extension
- Radiolucent line between ossified ligament and posterior margin of vertebral body on CT

Vertebral Endplate Spur

- Often associated with bulging disc
- Continuous with vertebral endplate
- Focal or diffuse

- Marrow signal may be present

PATHOLOGY

General Features

- Etiology
  - Bulge less important as separate entity, but associated with disc degeneration and anular fissures → "discogenic" pain
  - Environmental factors
    - Heavy lifting
    - Twisting
    - Flexion and extension
    - Acute trauma
  - Increasing age
  - Poor posture
  - Repetitive microtrauma
    - Endplate injury
    - Rim lesion: Horizontal tear in outer anulus at its attachment to ring apophysis
  - Interruption of nutritional supply to disc
  - Decreasing water content in nucleus pulposus
    - Proteoglycans replaced by fibrocartilage and fibrosis
    - Increasing disc stiffness
  - Ineffective cushioning
    - Decreasing intradisc pressure
  - Unequal force distribution to anulus
    - Increasing pressure on anulus posteriorly and posterolaterally
    - Concentric tear ± radial tear → disc bulge and herniation
- Genetics
  - Genetic predisposition to disc degeneration
- Associated abnormalities
  - Scoliosis
    - Asymmetric lateral bulge
  - Spondylolisthesis
    - Posterior bulge

Microscopic Features

- Myxomatous degeneration of anulus fibrosus
- Combination of anulus, nucleus, fibrocartilage, or apophyseal bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back pain
  - Other signs/symptoms
    - Radiculopathy
    - Neurogenic claudication
- Clinical profile
  - Worsening pain with flexion and increased intraabdominal pressure
    - Increasing intradisc pressure
  - Pain relieved by lying flat with flexed hips and knees

Demographics

- Age
  - 30-70 years
- Epidemiology
  - Up to 39% of asymptomatic adults have bulging discs

Natural History & Prognosis

- 82% without progression after 1 year
- > 80-90% success rate with conservative treatment

Treatment

- Conservative
  - NSAIDs
  - Physical therapy
  - Epidural injection
    - Cortisone ± lidocaine
- Surgical
  - Discectomy with stabilization if intractable pain
    - About 75% success rate

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Axial and sagittal CT or MR differentiates disc bulge from disc herniation

SELECTED REFERENCES

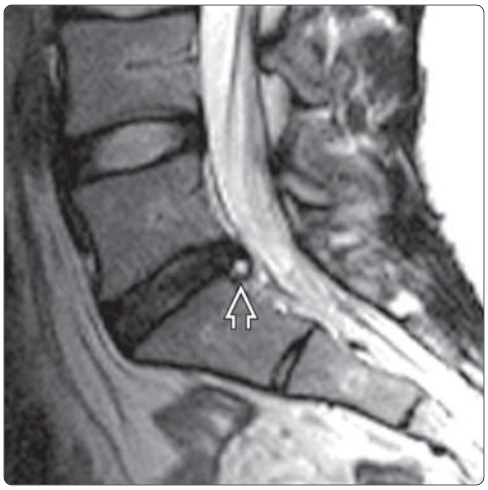
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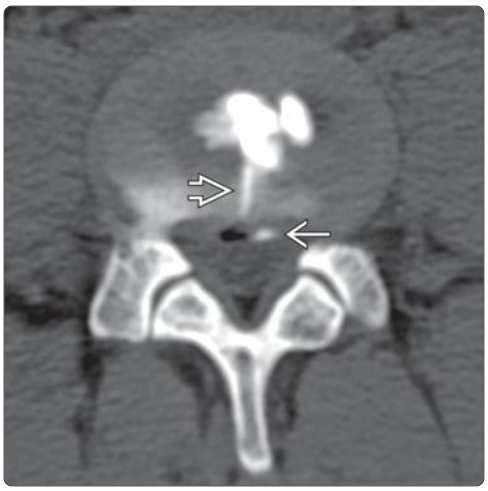
(Left) *Sagittal T2WI MR through the lumbar spine shows L4-L5 disc degeneration with loss of signal and a bulging disc touching the ventral thecal sac without significant deformity. A associated hyperintense annular fissure is present.* **(Right)** *Axial T2WI FS MR shows a bulging disc as broad-based extension of the annulus and without focal contour abnormality.*





(Left) *Sagittal T2WI MR shows bulging discs at L4-L5 and L5-S1 mildly effacing the ventral thecal sac. Both discs are degenerated with signal loss. Minimal height loss is present at L4-L5.* **(Right)** *Sagittal T2WI MR shows a disc bulge at L5-S1 with associated annular fissure. Disc degeneration is also present at L5-S1, manifested by signal loss.*





(Left) *Sagittal T2WI MR shows grade II spondylolisthesis with disc degeneration at L5-S1 with type I degenerative endplate changes. There is extensive "uncovering" of L4 and L5 discs which should not be confused with annular bulge. Note the annular fissures at L2-L3 and L3-L4.* **(Right)** *Axial NECT after discography shows a full thickness radial tear through the annulus. There is a broad-based annular bulge with a small amount of extravasated contrast.*

Anular Fissure, Intervertebral Disc

KEY FACTS

TERMINOLOGY

- Anular defect; high intensity zone (HIZ) in posterior anulus
- Disruption of concentric collagenous fibers comprising anulus fibrosus
- Term "tear" is nonstandard and may inaccurately imply acute traumatic etiology

IMAGING

- Focal HIZ in anulus on T2WI with low signal of parent disc
- Contrast enhancement on T1WI
- Focally enhancing nidus in posterior disc margin
- Discography is more provocative test (symptom simulation), rather than diagnostic imaging modality

TOP DIFFERENTIAL DIAGNOSES

- Disc space infection
- Focal disc protrusion
- Focal fat or ossification of disc with fatty marrow

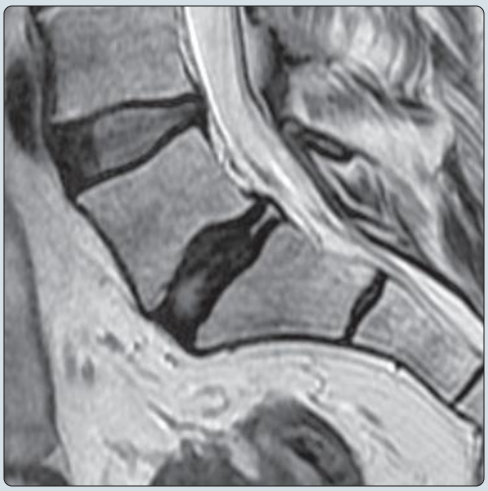
CLINICAL ISSUES

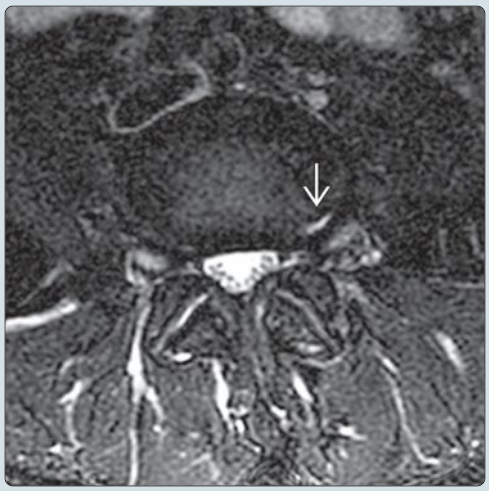
- Most anular tears are asymptomatic
  - Autopsy demonstrates high prevalence of fissures
  - MR shows anular fissures in majority of asymptomatic individuals
- Utility of discography remains controversial
- Direct association with disc degeneration
  - Recurrent meningeal nerve and ventral ramus of somatic spinal nerve are sources of innervation
  - Anular disruption may allow inflammatory substances to leak from nucleus

DIAGNOSTIC CHECKLIST

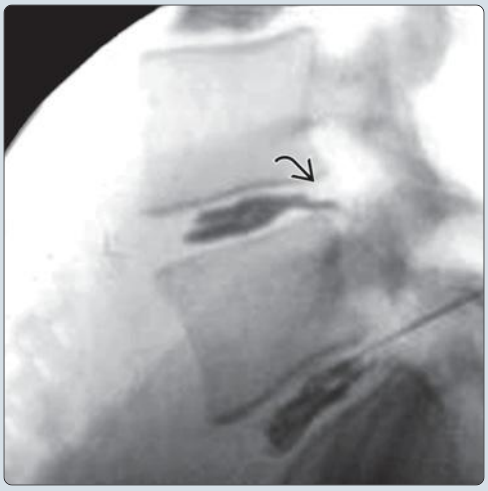
- Consider other sources of low back pain or disc pathology (e.g., disc infection)
- Incidental HIZ of otherwise normal disc is essentially normal finding

(Left) *Sagittal PD FSE MR demonstrates linear hyperintensity within the posterior aspect of the anulus fibrosus at L5-S1, reflecting the anular disruption of an anular fissure. There is mild bulging of the anulus at this level as well.* **(Right)** *Axial T2WI MR shows crescentic, focal high signal in L3-L4 disc ⇒, within the foraminal zone. Sagittal T2WI (not shown) revealed an identical focus cross-referenced to this site.*





(Left) *Lateral radiograph during discography shows leakage of intradiscal contrast at L4-L5 ⇨ and fissured and degenerated L4-L5 and L5-S1 discs. Symptoms were reproduced with injection of L5-S1 disc, not at L4-L5.* **(Right)** *Axial bone CT in the same patient following the discogram shows the leakage of contrast into the ventral epidural space at L4-L5, verifying the presence of the anular defect.*





TERMINOLOGY

Synonyms

- Anular defect; high-intensity zone (HIZ) in posterior anulus
  - Term "tear" is nonstandard and may inaccurately imply acute traumatic etiology rather than typical cause of repetitive stress

Definitions

- Disruption of concentric collagenous fibers comprising anulus fibrosus

IMAGING

General Features

- Best diagnostic clue
  - Abnormal signal focus (HIZ) at posterior disc margin on MR
- Location
  - Posterior margin of lumbar, thoracic, or cervical disc
- Size
  - Typically 1-2 mm focus
- Morphology
  - Well circumscribed
  - Typically seen with degenerating disc
  - Classic imaging appearance
    - Focal HIZ in anulus on T2WI with low signal of parent disc
    - Contrast enhancement on T1WI

CT Findings

- CECT
  - CECT can show enhancement of disc margin
  - CT post discography shows intradiscal contrast at posterior margin of anulus

MR Findings

- T1WI
  - No abnormality apparent on unenhanced image
- T2WI
  - High-signal zone in disc margin
- T1WI C+
  - Focally enhancing linear nidus in posterior disc margin

Other Modality Findings

- Discography demonstrates contrast leak from central site of injection through anulus
  - Utility is controversial
  - Discography is more provocative test (symptom simulation), rather than diagnostic imaging modality
    - Separates symptomatic fissures (internal disc disruption syndrome [IDD]) from incidental ones
    - If typical pain reproduced
    - No double-blind prospective studies exist to verify IDD theory
    - Several studies show poor correlation between presence of fissure and concordant symptoms at discography

Imaging Recommendations

- Sagittal heavily T2WI with thin sections
- Contrast-enhanced T1WI

DIFFERENTIAL DIAGNOSIS

Disc Space Infection

- Diffuse signal alteration of disc
- Enhancement throughout disc &/or paralleling endplate(s)
- Abnormal marrow adjacent to interspace
- Epidural abscess may be present

Focal Disc Protrusion

- High-signal zone represents both anular defect and disc material extending beyond normal margin of interspace
- Often accompanies anular defect when complete anular radial tear occurs

Focal Fat or Ossification of Disc With Fatty Marrow

- HIZ on T1WI in disc margin
- HIZ fades on T2WI or with fat-saturation pulse
- CT (or plain radiograph) shows ossification of posterior disc

PATHOLOGY

General Features

- Etiology
  - With age, anulus shows focal lamellar thickening
  - Concentric fissure
    - Repetitive stress on spinal motion element leads to lamellar separation; defect parallels lamellar orientation
  - Transverse fissure
    - Microtrauma produces defects in anular attachment at endplate rim
  - Radial fissure
    - Various factors, including loss of nutrient vessels, can lead to total lamellar disruption through outer margin
- Genetics
  - Genetic predisposition to "weak" collagen has been demonstrated in electron microscopic studies
  - Anular defects associated with certain familial conditions, such as Scheuermann syndrome
- Associated abnormalities
  - Disc protrusion or extrusion (herniation)
- Anulus composed of dense, concentric, crescentically ordered collagen layers
  - Vertically oriented
  - Attach to hyaline cartilage endplate at rim (Sharpey fibers)
  - Contains small blood vessels and nociceptive nerve fibers in outer lateral edge
  - Blood vessels and nerve fibers diminish with age
  - With age, inner anulus expands at expense of nucleus
- Anulus in cervical discs of adults differs from lumbar discs
  - Absence or attrition of thick posterior anulus
  - Only thin layer of vertical collagen fibers exists
  - Posterior longitudinal ligament contains disc within interspace

Staging, Grading, & Classification

- Concentric fissures are aging variant
- Rim fissures and radial tears may have clinical consequences in some cases
- Dallas discogram description
  - Grade 0: Contrast confined within nucleus pulposus

- Grade 1: Inner 1/3 of anulus
- Grade 2: Middle 1/3
- Grade 3: Outer 1/3
  - Focal or < 30° of disc circumference
- Grade 4 (April and Bogduk): Outer 1/3 of anulus
  - > 30° of disc circumference
- Extension of contrast beyond anulus
- Grade 5 (Schellhas)
- Full-thickness fissure
- Focal or diffuse

Gross Pathologic & Surgical Features

- Separation of lamellar structure in concentric tears
- Transverse disruption of fibers in rim, or radial fissures

Microscopic Features

- Granulation tissue with microvascularity invades fissures
  - Source of enhancement

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back or radicular pain
    - Recurrent meningeal nerve and ventral ramus of somatic spinal nerve are sources of innervation
    - Pain may be radicular in distribution
    - Anular disruption may allow inflammatory substances to leak from nucleus
    - Most anular fissures are asymptomatic
  - Other signs/symptoms
    - Reproducibility of pain with discography
- Clinical profile
  - Chronic back pain or sciatica in absence of mechanical nerve root compromise
  - Controversial role of anular fissure
    - Most anular fissures are incidental findings

Demographics

- Age
  - Up to 25% of normal individuals younger than 20 show anular defects
  - Majority of adult individuals harbor anular defects
- Gender
  - No gender differences
- Epidemiology
  - Autopsy demonstrates high prevalence of anular fissures
    - Increase with age
  - Direct association with disc degeneration
  - Discography shows presence of anular fissures in up to 80% of degenerated discs
  - MR shows anular fissures in majority of asymptomatic individuals
    - Some fissures are seen only on CE T1WI
    - 96% of all fissures enhance
    - Most anular fissures persist for months on follow-up MR

Natural History & Prognosis

- Most are asymptomatic or have self-limited symptoms
  - Scar endpoint of inflammation
- Some felt to be cause of chronic back pain and sciatica
- Up to 1/3 of chronic pain sufferers have recurrent symptoms despite any therapy

Treatment

- Symptomatic pain relief with NSAIDs
- Chronic pain may prompt patients to undergo interbody fusion as last resort

DIAGNOSTIC CHECKLIST

Consider

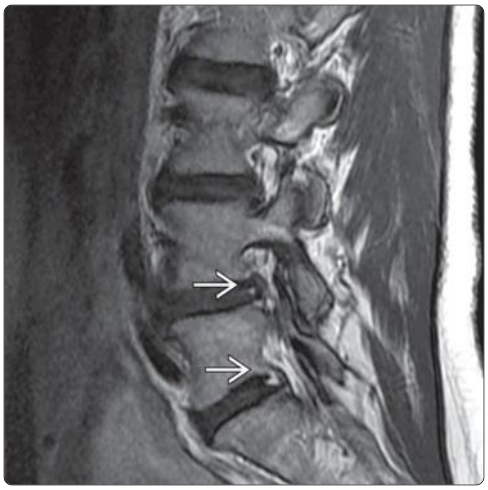
- Other sources of low back pain or disc pathology (e.g., disc infection)

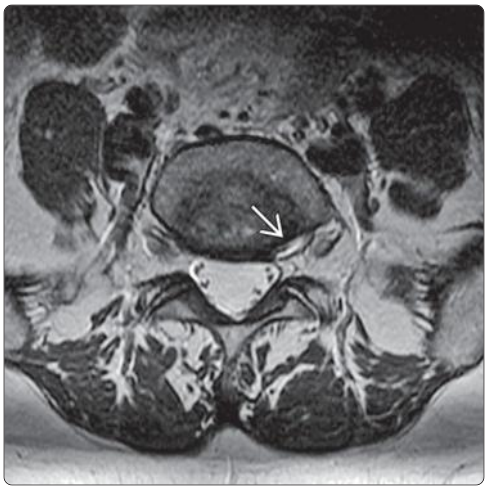
Image Interpretation Pearls

- Incidentally seen focal high intensity zone in anulus of otherwise normal disc is essentially normal finding

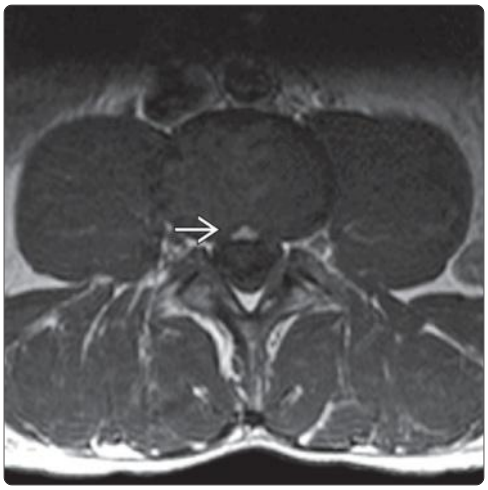
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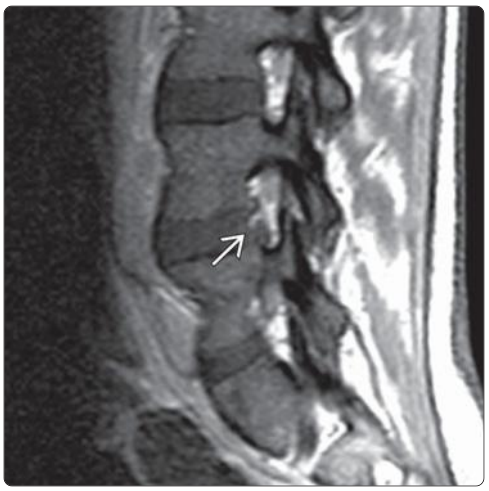
(Left) *Sagittal T2WI MR shows focal areas of T2 hyperintensity within the posterior anulus fibrosus at both L4-L5 and L5-S1, reflecting anular fissures. Both disc levels show anular bulging.* **(Right)** *Axial T2WI MR shows an anular fissure along the posterior lateral margin of the L5-S1 disc as linear hyperintensity without focal contour abnormality.*





(Left) *Axial T1WI C+ MR shows triangle-shaped enhancement within the anulus at the L4-L5 level without disc contour abnormality due to enhancement of an anular fissure.* **(Right)** *Sagittal T1WI C+ MR shows focal enhancement within posterior anulus. This is a typical pattern for focal enhancement within anular fissure. There is also enhancement within the posterior anulus at L2-L3 and severe degenerative disc disease at L5-S1.*





(Left) *Sagittal T2WI MR shows severe disc degeneration at L5-S1 with marked type I endplate changes and spondylolisthesis. Note the ventral anular fissures at L2-L3 and L3-L4 seen as linear hyperintensity.* **(Right)** *Sagittal T1WI C+ MR shows an enhancing high-signal focus in the foraminal margin of the L4-L5 disc due to anular fissure.*

Cervical Intervertebral Disc Herniation

KEY FACTS

TERMINOLOGY

- Protruded disc, extruded disc, free fragment, sequestered disc
- Localized (< 50% of disc circumference) displacement of disc material beyond edges of vertebral ring apophyses

IMAGING

- Small mass in ventral spinal canal, contiguous with intervertebral disc
- Protrusion is herniated disc with broad base at parent disc
- Extrusion is herniated disc with narrow or no base at parent disc
- Sequestered or free fragment: Extruded disc without contiguity to parent disc
- Migrated: Disc material displaced away from site of herniation, regardless of continuity to parent disc

TOP DIFFERENTIAL DIAGNOSES

- OPLL
- Osteophyte
- Tumor
- Hemorrhage
- Abscess

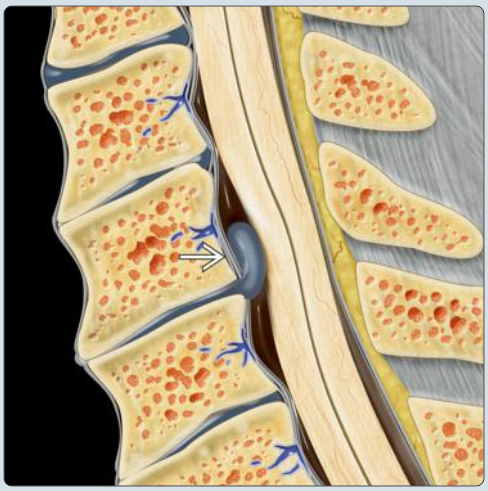
CLINICAL ISSUES

- 10% of people under age 40 have cervical herniation
- Acute radiculopathy usually self-limited disorder with full recovery expected
  - Neck pain (90%)
  - Radicular pain (65%)
  - Paresthesia (89%)
- Treatment
  - Conservative treatment
  - Multiple surgical approaches without clear consensus

DIAGNOSTIC CHECKLIST

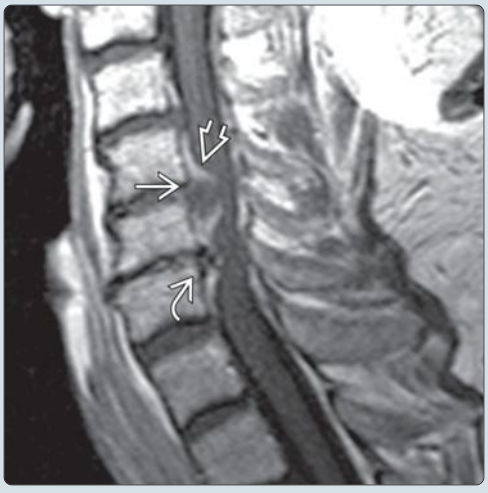
- Use fast STIR for cord disease
- Axial GRE, T2* GRE for disc definition

(Left) *Sagittal graphic shows disc extrusion, with the base of the herniation smaller than the epidural component, effacing the thecal sac and causing cord compression.* **(Right)** *Sagittal T2WI MR shows C5-C6 disc extrusion with the base of herniation smaller than the component extending into the epidural space.*





(Left) *Sagittal T1WI C+ MR shows C4-5 extrusion impinging upon the cord. Smaller C5-C6 protrusion also compresses the ventral cord. Peripheral enhancement is related to epidural plexus and granulation tissue.* **(Right)** *Axial T2* GRE MR shows left-sided extrusion effacing the left side of the thecal sac and compressing the cord, extending toward the left neural foramen.*





TERMINOLOGY

Abbreviations

- Cervical disc herniation (CDH)

Synonyms

- Protruded disc, extruded disc, free fragment, sequestered disc
- Nonstandard: Prolapse, herniated nucleus pulposus (HNP), rupture

Definitions

- Localized (< 50% of disc circumference) displacement of disc material beyond edges of vertebral ring apophyses

IMAGING

General Features

- Best diagnostic clue
  - Small mass in spinal canal, contiguous with intervertebral disc
- Location
  - Ventral epidural
  - C6-C7 level most common, with compression of C7 root
  - C5-C6 2nd most common location, compressing C6 root
- Size
  - Variable
- Morphology
  - Protrusion
    - Herniated disc with broad base at parent disc
    - Greatest diameter of protrusion in any plane greater than distance between edges of base in same plane
    - Focal: < 25% of disc circumference
    - Broad based: > 25% but < 50% of disc circumference (> 50% is defined as bulge)
  - Extrusion
    - Extrusion is herniated disc with narrow or no base at parent disc
    - Greatest diameter of extrusion in any plane greater than distance between edges of base in same plane
    - Sequestered or free fragment: Extruded disc without contiguity to parent disc
    - Migrated: Disc material displaced away from site of herniation, regardless of continuity to parent disc
  - Intravertebral herniation (Schmorl node)

CT Findings

- NECT
  - Ventral epidural soft tissue density extending dorsally into spinal canal contiguous with disc
  - Displaces exiting nerve roots and distorts thecal sac

MR Findings

- T1WI
  - Isointense to parent disc
  - May show low signal if calcified or associated with vacuum phenomenon
- T2WI
  - Isointense to hyperintense
- T2* GRE
  - Variable in signal reflecting hydration of disc material (hyperintense) or calcification (hypointense)
- T1WI C+
  - No enhancement of disc material
  - May enhance peripherally after intravenous contrast material due to granulation tissue or dilated epidural plexus
  - May rarely diffusely enhance if associated with granulation tissue or post-contrast imaging is delayed due to diffusion
- Variable amount of exiting root and thecal sac impingement
- Variable amount of degenerative changes at same or other disc levels

Nonvascular Interventions

- Myelography
  - Extradural defect at intervertebral disc level
  - Effacement or cutoff of root sleeves

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal, axial T1WI and T2WI; intravenous contrast if question of tumor, infection

DIFFERENTIAL DIAGNOSIS

OPLL

- Flowing low signal epidural space on all sequences
- May show increased signal on T1WI due to fatty marrow content

Osteophyte

- Sharp margins not arising directly from intervertebral disc level
- May show increased signal on T1WI due to fatty marrow content

Tumor

- Homogeneous enhancement, irregular and infiltrative
- Not arising out of intervertebral disc

Hemorrhage

- Elongated within epidural space
- Tends to be posterior
- Isointense in hyperacute phase, increased signal in subacute stage on T1WI

Abscess

- May mimic large herniation with peripheral enhancement
- Increased signal on T2WI, endplate destruction

PATHOLOGY

General Features

- Etiology
  - Degenerative breach in anulus
  - May be post-traumatic
  - Disc material extends through defect
  - Protrusions may be contained by thin annuloligamentous complex
- Genetics

- Congenitally weak collagen predisposes to degenerative disc disease
- Increased incidence of spondylosis in Marfan syndrome
- Associated abnormalities
  - 20% of people over age 40 have cervical foraminal stenosis
- Composed of combination of nucleus pulposus, fragmented anulus, cartilage, and fragmented apophyseal bone

Gross Pathologic & Surgical Features

- Nucleus pulposus
  - Gelatinous material with high water content and few collagen fibers
  - Water content decreases with age
- Anulus fibrosus
  - Fibrocartilage, with collagen fibers in concentric lamellae

Microscopic Features

- Degeneration shows cracks, fissures within nucleus, endplate fragmentation, granulation tissue, thickening endplate trabeculae

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Clinical symptoms affected by level, location, and size of herniation
  - Neck pain and stiffness, with pain radiating to shoulder and upper extremity
  - May include paresthesia, hyperesthesia, and weakness
    - Neck pain (90%)
    - Paresthesia (89%)
    - Radicular pain (65%)
    - Weakness (15%)
  - Central herniations produce cord compression; paracentral compress ipsilateral hemicord
  - Lateral herniations compress anterior cervical root giving weakness without pain
  - Lateral herniations compress root and dorsal root ganglion causing pain
- Clinical profile
  - 4th through 5th decades
  - Risk factors include increasing age, heavy physical work, work dissatisfaction, depression, history of headache

Demographics

- Age
  - Adult population
- Gender
  - M > F
- Epidemiology
  - 10% of people under age 40 have cervical herniation

Natural History & Prognosis

- Acute radiculopathy usually self-limited disorder with full recovery expected
- Chronic nonspecific pain much more variable
  - 90% back to work by 3 months
  - Continuous or recurrent pain affects 30-40%

Treatment

- Options, risks, complications
  - Conservative treatment
    - Exercise and physical conditioning
    - Nonsteroidal anti-inflammatory drugs
    - Transcutaneous electrical stimulation
    - Epidural steroid injection
    - Opioids
    - Atypical analgesics such as antidepressants, antiepileptic drugs, and neuroleptics
  - Multiple surgical approaches without clear consensus
    - Anterior discectomy with/without fusion
    - Hemilaminectomy and foraminotomy
    - Cervical artificial disc placement
    - Compressive cervical myelopathy with spondylosis treated by corpectomy/strut graft or multilevel laminectomy

DIAGNOSTIC CHECKLIST

Consider

- OPLL if extradural disease low in signal on all sequences

Image Interpretation Pearls

- Define herniation by level, size, location (central, paracentral, lateral), effect on thecal sac and cord, and migration away from disc level
- Axial T1WI + T2* GRE better define disc contour due to variable disc signal
- FLAIR insensitive for intrinsic cord signal abnormality such as contusion, myelomalacia
  - Use fast STIR for cord disease combined with standard fast spin-echo T2WI

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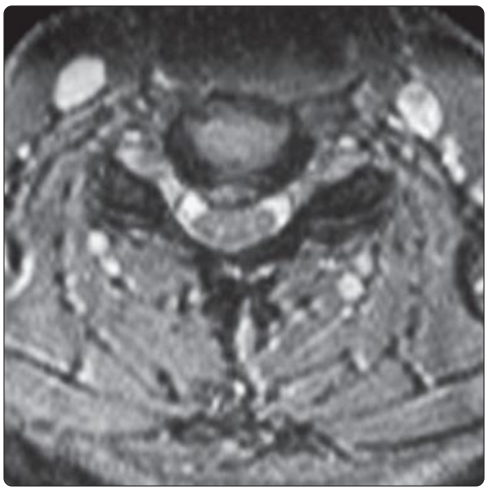
(Left) *Sagittal T2WI MR shows severe cervical spondylosis (worst at C4-C5) with cord compression related to anterior disc osteophyte complex and ligamentous hypertrophy. T2 hyperintensity is present in the cord due to contusion or myelomalacia.* **(Right)** *Sagittal T1WI C+ MR shows C4-C5 stenosis where there is cord compression related to anterior disc osteophyte complex and posterior ligamentous hypertrophy. Focal cord enhancement present at C4-C5 is due to myelomalacia.*





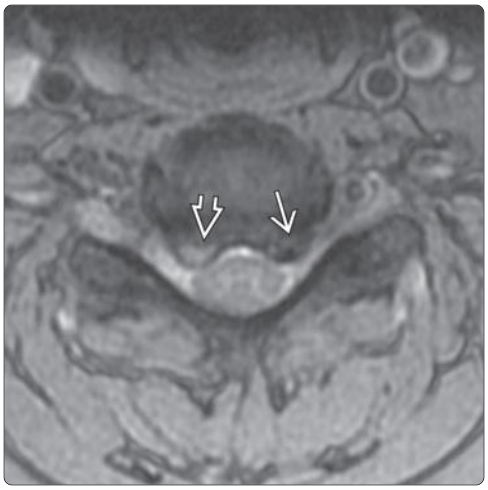
(Left) *Sagittal T2WI MR shows severe multilevel subarachnoid space narrowing reflecting a combination of disc herniation, osteophyte, and OPLL. Increased signal is present in the cord from myelomalacia.* **(Right)** *Sagittal T2WI MR demonstrates a metal artifact at C6-C7 from prior cervical fusion. There is a large extrusion at C5-C6 severely compressing the ventral cord.*





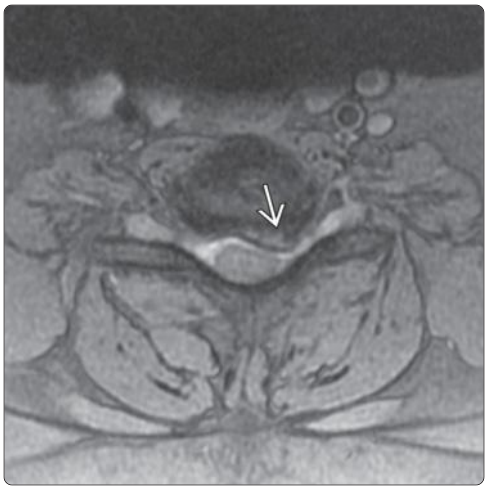
(Left) *Sagittal T2WI MR shows a small central protrusion as low signal effacing the ventral thecal sac but not distorting the cord. Type I degenerative endplate changes are present as high signal at C5-C6.* **(Right)** *Axial T2* GRE MR shows small focal protrusion touching the ventral cord. This focal herniation is less than 25% of the disc margin.*

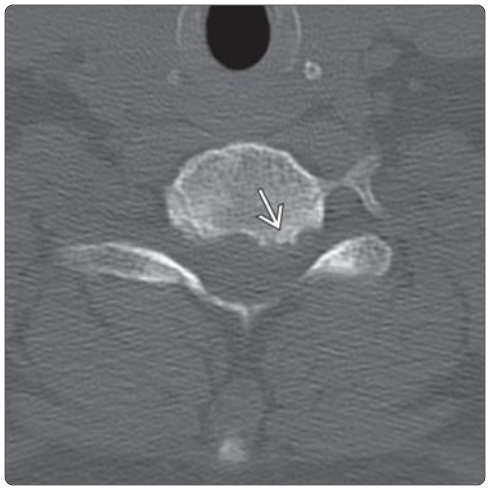
(Left) *Axial GRE MR shows a right-sided protrusion ➔ abutting the right ventral cord, which shows no calcification on the accompanying CT study. Low signal left-sided protrusion ➔ contains calcification on the CT.* **(Right)** *Axial NECT (bone window) shows a small focus of calcification that resides within the left-sided disc protrusion ➔, which is much better identified on the accompanying MR. There is a right-sided protrusion on the MR at this level that does not have calcium; therefore it cannot be identified ➔.*





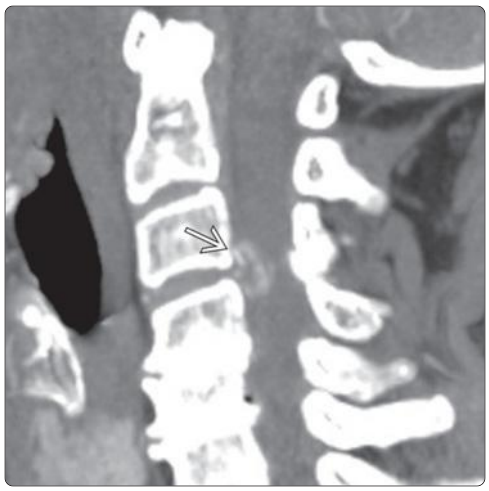
(Left) *Axial GRE MR shows a large left-sided protrusion with a relatively increased signal ➔, suggesting a "soft disc" or one with minimal calcification. There is mild mass effect upon the cord.* **(Right)** *Axial CT shows minimal osteophyte formation ➔ at the base of the left-sided disc extrusion, which is present on the MR study.*





(Left) *Sagittal T2 MR shows a large extrusion at the C3-4 level ➔, severely compressing the cervical cord. There is cord T2 hyperintensity at C3-4 that is consistent with myelomalacia.* **(Right)** *Sagittal NECT (bone window) shows extensive calcification within the large disc extrusion ➔ at the C3-4 level, with marked narrowing of the thecal sac space.*

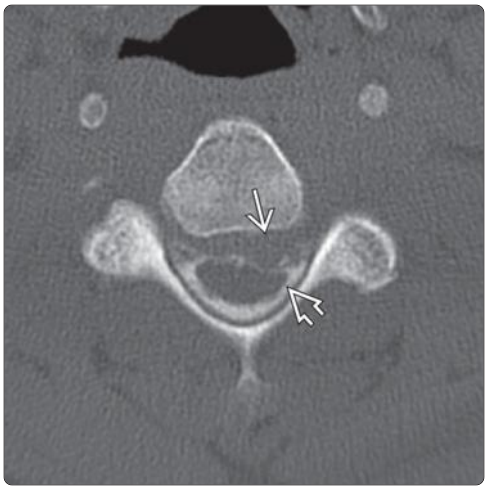


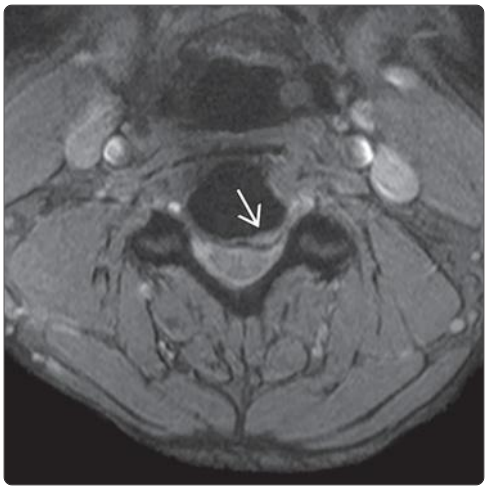






(Left) *Lateral view of a cervical myelogram shows a large ventral extradural defect ➡ in the contrast column from a disc herniation. There is severe narrowing of the subarachnoid space at that level. This patient has had prior ACDF ➡.* **(Right)** *Sagittal CT myelogram shows a large central protrusion of the intervertebral disc ➡ with mild cord compression. This patient has a solid fusion from prior ACDF at C5-6.*





(Left) *Axial CT myelogram shows a broad-based left-sided protrusion ➡ of the intervertebral disc with effacement of the thecal sac and mild cord compression ➡.* **(Right)** *Axial GRE MR shows the left-sided disc protrusion ➡ as a broad-based focus of increased signal, suggesting minimal associated calcification or osteophyte.*





(Left) *Axial T1WI MR show a large disc extrusion ➡ with compression of the right anterolateral cord ➡ and extension to the right neural foramen.* **(Right)** *Sagittal T2WI MR show a large disc extrusion at C6-7 ➡ with migration cephalad from the parent disc, suggesting a free disc fragment.*

Thoracic Intervertebral Disc Herniation

KEY FACTS

IMAGING

- Small mass in spinal canal contiguous with intervertebral disc
  - Ventral epidural
  - Rare in upper thoracic spine (T1-T3)
- May enhance peripherally after intravenous contrast material due to granulation tissue or dilated epidural plexus
  - Peripheral enhancement may give "lifted band" or "tent" configuration

TOP DIFFERENTIAL DIAGNOSES

- Osteophyte
  - Sharp margins not arising directly from intervertebral disc level
- Tumor
  - Homogeneous enhancement
- Hemorrhage
  - Elongated within epidural space, tends to be posterior
- Abscess
  - May mimic large herniation with peripheral enhancement

CLINICAL ISSUES

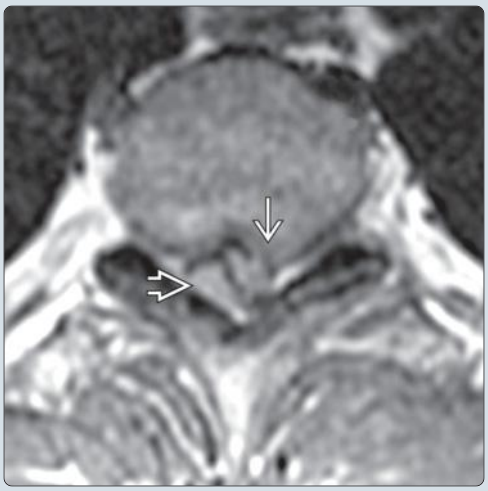
- 5th decade
- Uncommon entity
- Thoracic disc surgery uncommon, represents 1-2% of all disc surgery
- History of Scheuermann

DIAGNOSTIC CHECKLIST

- Calcification (65%)
- Multiple herniation (14%)
- T2WI critical since herniation may not be visible on T1WI due to calcification
- **Check and recheck herniation level, counting from C2 and L5 levels**

(Left) *Sagittal T2WI MR shows a large low signal disc extrusion at T2-T3 effacing the thecal sac and tenting the ventral dura. A smaller disc protrusion is present at T7-T8 with no neural compromise.* **(Right)** *Axial T1WI MR demonstrates the large left-sided extrusion ➡ severely compressing the left side of the cord ➡.*





(Left) *Sagittal T2WI MR shows multiple large, lower thoracic disc herniations, which severely compress the cord at multiple levels. Thoracic herniations are most common from T7-T12.* **(Right)** *Sagittal T1WI C+ MR shows pronounced peripheral enhancement surrounding the herniations related to tented and distended epidural plexus and herniation-associated granulation tissue.*





TERMINOLOGY

Abbreviations

- Thoracic disc herniation (TDH)

Synonyms

- Protruded disc, extruded disc, free fragment, sequestered disc
- Nonstandard: Prolapse, herniated nucleus pulposus (HNP), rupture
- Dorsal spinal disc herniation

Definitions

- Localized (< 50% of disc circumference) displacement of disc material beyond edges of vertebral ring apophyses

IMAGING

General Features

- Best diagnostic clue
  - Small mass in spinal canal contiguous with intervertebral disc
- Location
  - Ventral epidural
  - T6 → T11 most common
  - Rare in upper thoracic spine (T1-T3)
- Size
  - Variable
- Morphology
  - Protrusion
    - Herniated disc with broad base at parent disc
    - Greatest diameter of herniated disc in any plane greater than distance between edges of base in same plane
    - Focal: < 25% of disc circumference
  - Extrusion
    - Herniated disc with narrow or no base at parent disc
    - Greatest diameter of herniated disc in any plane greater than distance between edges of base in same plane
    - Sequestered or free fragment: Extruded disc without contiguity to parent disc
    - Migrated: Disc material displaced away from site of herniation, regardless of continuity to parent disc
  - Intravertebral herniation (Schmorl node)

CT Findings

- NECT
  - Ventral epidural soft tissue density extending dorsally into spinal canal contiguous with disc
  - Displaces cord and thecal sac
  - May be completely or partially calcified

MR Findings

- T1WI
  - Isointense to hypointense to parent disc
  - May show low signal if calcified or associated with vacuum phenomenon
  - Herniation may only be inferred by mass effect upon cord if lesion very low in signal
- T2WI
  - Variable signal reflecting hydration state and presence or absence of calcification
- STIR
  - Variable signal reflecting hydration state and presence or absence of calcification
- T2* GRE
  - Variable signal reflecting hydration of disc material (hyperintense) or calcification (hypointense)
- T1WI C+
  - No enhancement of disc material
  - May enhance peripherally after intravenous contrast material due to granulation tissue or dilated epidural plexus
  - Peripheral enhancement may give "lifted band" or "tent" configuration

Nonvascular Interventions

- Myelography
  - Extradural defect at intervertebral disc level
  - Effacement or cutoff of root sleeves
  - Displacement or compression of cord

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal, axial T1WI, and T2WI; intravenous contrast if question of tumor, infection

DIFFERENTIAL DIAGNOSIS

Osteophyte

- Sharp margins not arising directly from intervertebral disc level
- May show increased signal on T1WI due to fatty marrow content

Tumor

- Homogeneous enhancement
- Not arising out of intervertebral disc
- Irregular and infiltrative

Hemorrhage

- Elongated within epidural space, tends to be posterior
- Isointense in hyperacute phase; increased signal in subacute stage on T1WI

Abscess

- May mimic large herniation with peripheral enhancement
- Increased signal on T2WI
- Associated with endplate destruction of disc space infection and adjacent osteomyelitis

PATHOLOGY

General Features

- Etiology
  - Degenerative breach in anulus
  - Prior history of trauma (37%)
  - Disc material extends through defect
  - Protrusions may be contained by thin annuloligamentous complex
- Genetics

- Congenitally weak collagen predisposes to degenerative disc disease
- Associated abnormalities
  - Signs of disc degeneration elsewhere in spine
    - Loss of disc space height
    - Loss of disc signal on T2WI
    - Vacuum disc
    - Scheuermann endplate irregularity
    - Linear remodeling of endplate directed to herniation
  - Disc bulge or tear at other thoracic levels
- Composed of combination of nucleus pulposus, fragmented anulus, cartilage, and fragmented apophyseal bone

Gross Pathologic & Surgical Features

- Nucleus pulposus
  - Gelatinous material with high water content and few collagen fibers
  - Water content decreases with age
- Anulus fibrosus
  - Fibrocartilage, with collagen fibers in concentric lamellae

Microscopic Features

- Degeneration shows cracks, fissures within nucleus, endplate fragmentation, granulation tissue, thickening endplate trabeculae

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Protean symptomatology
  - Pain: Axial, localized, or radicular (76%)
  - Myelopathy
    - Motor impairment (60%)
    - Hyperreflexia, spasticity (58%)
    - Sensory impairment (61%)
    - Bowel or bladder dysfunction (24%)
  - Other signs/symptoms
    - May rarely present as abdominal pain
- Clinical profile
  - 5th decade
  - Prior trauma history
  - History of Scheuermann

Demographics

- Age
  - Adult population, mean age ~ 50 years
- Gender
  - M = F
- Epidemiology
  - Uncommon entity
  - Estimated 2-3 cases per 1,000 patients with disc protrusions
  - 1 patient per million population per annum

Natural History & Prognosis

- Asymptomatic population
  - Thoracic disc herniation (37%)
- Thoracic disc herniations
  - As in cervical and lumbar spine, high percentage of herniations remain without major neurologic compromise
  - 20-30% may require surgery
  - Small herniations (0-10% canal compromise) tend to remain stable in size and remain asymptomatic
  - Large herniations (> 20% canal compromise) tend to decrease in size
  - May be multilevel

Treatment

- Options, risks, complications
  - Thoracic disc surgery uncommon, represents 1-2% of all disc surgery
  - Surgery for intractable pain, neurologic dysfunction
  - Variety of surgical approaches
    - Transthoracic
    - Transfacet pedicle sparing
    - Lateral extracavitary/costotransversectomy
    - Transpedicular
  - Causes of surgical failure for thoracic herniations
    - Misidentified level
    - Cord injury/vascular injury
    - Migrated disc fragment missed
    - Intradural disc extension missed
    - Lack of adequate visualization during surgery

DIAGNOSTIC CHECKLIST

Consider

- Calcification (65%)
- Multiple herniations (14%)
- Intradural herniations (7%)

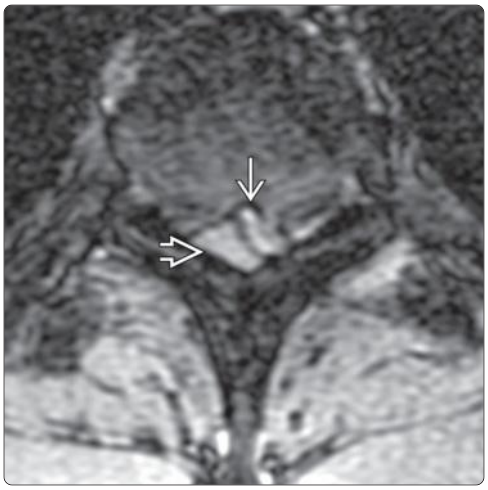
Image Interpretation Pearls

- T2WI critical since herniation may not be visible on T1WI due to calcification
- Check and recheck herniation level, counting from C2 and L5 levels
- Combination of axial T1WI and gradient-echo T2* better define disc contour due to variable disc signal

SELECTED REFERENCES

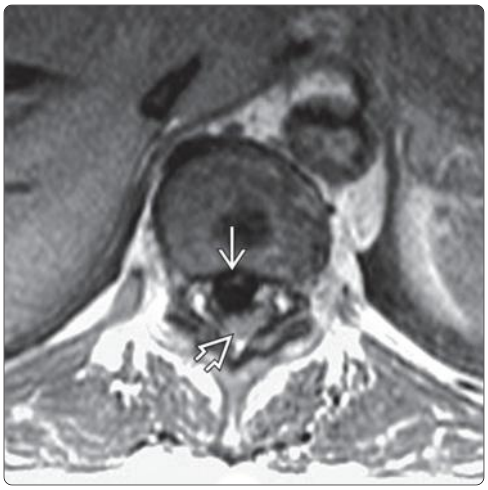
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(Left) *Sagittal T2WI MR shows a large thoracic disc extrusion compressing the anterior cord. There are sloping margins to the lesion related to displaced posterior longitudinal ligament and veins.* **(Right)** *Axial T2* GRE MR through the upper thoracic spine shows a large left-sided extrusion compressing the left lateral margin of the thoracic cord.*





(Left) *Sagittal T1WI MR with contrast shows a large hypointense mass contiguous with disc space reflecting calcified disc extrusion. Prominent distended epidural veins "tent" around herniation.* **(Right)** *Axial T1WI C+ MR shows a large thoracic calcified extrusion as a low signal ventral epidural mass effacing the cord.*





(Left) *Sagittal T1WI MR shows a large thoracic disc extrusion at T10-T11 migrating superiorly behind the T10 vertebral body. There is severe disc degeneration at T10-T11 and T11-T12 with vacuum disc.* **(Right)** *Sagittal T2WI MR shows the large disc extrusion as intermediate signal intensity, migrating superiorly behind the T10 body reflecting free fragment.*

Lumbar Intervertebral Disc Herniation

KEY FACTS

TERMINOLOGY

- Localized (< 25% of disc circumference) displacement of disc material beyond confines of disc space
- Protrusion
  - Herniated disc with broad base at parent disc
- Extrusion
  - Herniated disc with narrow or no base at parent disc
- Sequestered: Free fragment

IMAGING

- Anterior extradural mass contiguous with disc space extending into spinal canal
- Axial: Central, subarticular (lateral recess), foraminal, or extraforaminal (far lateral)
- Sagittal: Disc level, infrapedicle, pedicle, suprapedicle

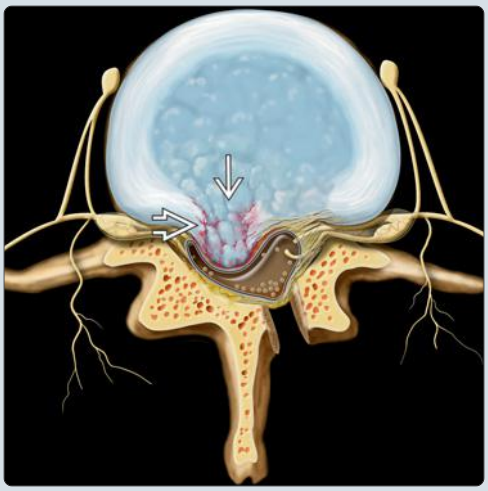
TOP DIFFERENTIAL DIAGNOSES

- Peridural fibrosis
- Epidural abscess
- Epidural metastasis
- Nerve sheath tumor

CLINICAL ISSUES

- Low back pain
- Radiculopathy: Posterolateral radiating pain down lower extremity
  - Positive straight-leg raising test (Lasègue's sign)
  - Cauda equina syndrome
- Back pain ± radiculopathy resolves within 6-8 weeks
- Treatments
  - Conservative
  - Minimally invasive techniques (many variations)
  - Standard open techniques, such as laminotomy/discectomy
    - ≥ 90% success rate

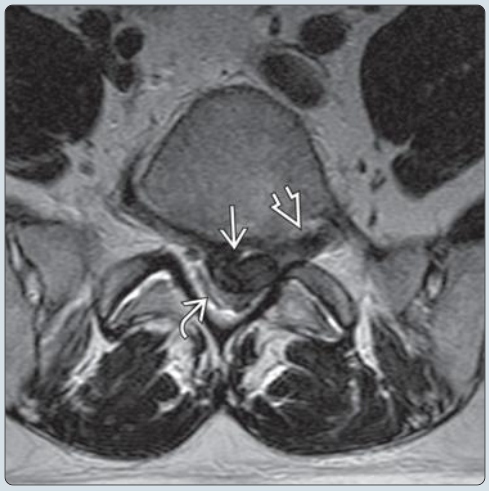
(Left) *Graphic of large recurrent disc herniation shows displacement of the nuclear material ➡ through a large defect in the posterior annular fibers ➡, with effacement of the ventral thecal sac and displacement of the intrathecal nerve roots.* **(Right)** *Sagittal T1 MR shows a very large disc extrusion at L5-S1 level ➡ with superior migration of a large component to L4-L5 level ➡. The thecal sac is nearly completely effaced by the large herniation.*





(Left) *Sagittal T2 TSE shows a large extrusion of the intervertebral disc at the L5-S1 level with a sequestered component (free fragment) ➡ that is migrated superiorly.* **(Right)** *Axial T2WI MR shows a large central and left extrusion of the intervertebral disc ➡ with severe effacement of the thecal sac ➡. The herniation extends into the left neural foramen ➡.*





TERMINOLOGY

Synonyms

- Nonstandard: Herniated nucleus pulposus
- Nonstandard: "Slipped" or prolapsed disc

Definitions

- Localized (< 25% of disc circumference) displacement of disc material beyond confines of disc space
- Protrusion
  - Herniated disc with broad base at parent disc
    - Greatest dimension of disc herniation in any plane ≤ distance between edges of base in same plane
- Extrusion
  - Herniated disc with narrow base or no base at parent disc
    - Greatest dimension of disc herniation in any plane > distance between edges of base in same plane
- Sequestered: Free fragment
  - Extruded disc without continuity to parent disc
- Migrated
  - Disc material displaced away from site of herniation
  - Regardless of continuity
- Intravertebral herniation: Schmorl node (endplate herniation)

IMAGING

General Features

- Best diagnostic clue
  - Anterior extradural mass contiguous with disc space extending into spinal canal
- Location
  - Most common: L4-L5 or L5-S1
    - 90% of lumbar disc herniation
  - Axial plane
    - Central, subarticular (lateral recess), foraminal, or extraforaminal (far lateral)
  - Sagittal plane
    - Disc level, infrapedicle, pedicle, or suprapedicle
- Size
  - Variable
- Morphology
  - Focal: < 25% of disc circumference
  - Nonstandard: Broad based is > 25%, but < 50% of disc circumference (overlaps with asymmetric bulge nomenclature)

CT Findings

- NECT
  - Anterior extradural soft tissue attenuation mass
    - Displacing nerve root and distorting thecal sac

MR Findings

- T1WI
  - Isointense to parent disc
- T2WI
  - Iso- to hyperintense
    - Depending on degree of disc hydration
- T1WI C+
  - Peripheral enhancement
    - Diffuse enhancement if imaged > 30 minutes after injection
- Variable extent of nerve impingement and central stenosis
  - Enhancing nerve root likely due to venous congestion ± inflammation
- Disc hypointensity and height loss

Nonvascular Interventions

- Myelography
  - Extradural mass indenting thecal sac and effacing or displacing nerve root sleeves

Other Modality Findings

- Discography
  - Pain responses
    - No pain, dissimilar pain, similar pain, or exact pain
    - Positive response if exact pain reproduced by pressurization (symptomatic disc)
  - Extension of contrast beyond anulus fibrosus
    - Grade 5 (Schellhas)
    - Full thickness tear

Imaging Recommendations

- Best imaging tool
  - MR: T2WI and T1WI in sagittal and axial planes
- Protocol advice
  - Intravenous gadolinium in postoperative patients
    - Distinguishes scar tissue from recurrent disc herniation

DIFFERENTIAL DIAGNOSIS

Peridural Fibrosis

- Early and homogeneous enhancement
- Surrounding thecal sac and nerve root

Epidural Abscess

- Look for discitis and osteomyelitis
- Peripheral enhancement

Epidural Metastasis

- Osseous involvement
- Epidural component elongated in cranialcaudal dimension
- Paravertebral extension

Nerve Sheath Tumor

- Avid enhancement
- 15% with "dumbbell" configuration

PATHOLOGY

General Features

- Etiology
  - Environmental factors
    - Heavy lifting
    - Twisting
    - Flexion and extension
    - Acute trauma
  - Repetitive microtrauma
    - Rim lesion: Horizontal tear in outer anulus at its attachment to ring apophysis
    - Endplate separation → interrupted nutritional supply to disc

- Decreased water content in nucleus pulposus
- Unequal force distribution to anulus
  - Coalescing concentric tears → radial tear → disc herniation
- Genetics
  - Gene variations associated with lumbar disc degeneration
    - Collagen IX and XI genes
    - Vitamin D receptor gene
    - Aggrecan gene
- Associated abnormalities
  - Transitional vertebra below level of lumbar herniation
  - Adjacent level degeneration/herniation cephalad to interbody fusion
  - Scoliosis
- Anatomy
  - Central nucleus pulposus
    - Shock absorbing
    - Proteoglycans and glycosaminoglycans
    - High water content
  - Transition zone
  - Peripheral anulus fibrosus
    - Tensile strength
    - Outer zone: Dense collagen fibers in concentric lamellae
    - Inner zone: Fibrocartilage

Microscopic Features

- Combination of nucleus pulposus, anulus, cartilage, or fragmented apophyseal bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Radiculopathy: Posterolateral radiating pain down lower extremity
  - Other signs/symptoms
    - Low back pain
    - Positive straight-leg raising test (Lasègue's sign)
    - Cauda equina syndrome
- Clinical profile
  - Worsening symptoms during lumbar flexion
    - Sitting, bending
    - Increased pressure on nucleus pulposus
  - Worsening pain with increased intraabdominal pressure
    - Coughing, sneezing
  - Pain relieved by lying flat

Demographics

- Age
  - 30-60
    - Mean: 40s
  - Increasing age correlates with more cephalad level of herniation
- Gender
  - Slight male predominance
- Epidemiology
  - Up to 1/3 of asymptomatic adults have disc herniation

Natural History & Prognosis

- Back pain ± radiculopathy resolves within 6-8 weeks
  - Conservative treatment alone
  - 90% of patients
    - 70% within 1st 4 weeks
- 5% recurrent disc herniation

Treatment

- Conservative
  - NSAIDs
  - Physical therapy
  - Epidural injection
    - Cortisone ± lidocaine
- Chymopapain chemonucleolysis
  - Fallen out of use due to complication rate
  - 3.7% complication rate (discitis)
- Indications of surgery
  - Intractable pain
  - Cauda equina syndrome
  - Progressive neurologic deficits
- Surgical
  - Minimally invasive techniques (many variations)
    - Dekompressor, automated percutaneous discectomy, laser discectomy
    - Percutaneous endoscopic lumbar discectomy
  - Standard open techniques, such as laminotomy/discectomy
    - ≥ 90% success rate
    - 5% "failed back" syndrome

DIAGNOSTIC CHECKLIST

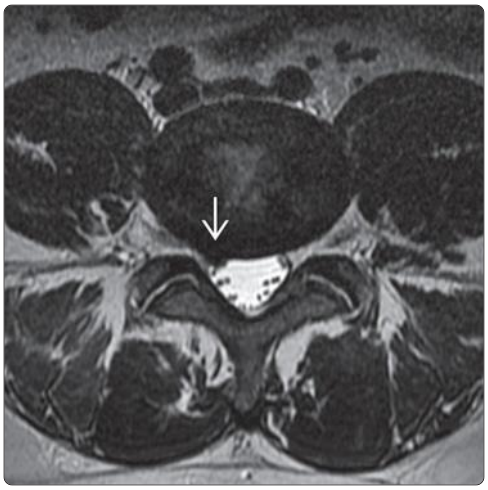
Image Interpretation Pearls

- Sagittal imaging best at discriminating extrusion from protrusion
  - "Mushroom" appearance of extrusion due to focal expansion

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(Left) *Sagittal T2WI MR shows a tongue-like disc protrusion at L4-L5 ➡. The greatest cranialcaudal dimension of the protruded disc is the same as that of its base, making protrusion the correct term.* **(Right)** *Axial T2WI MR shows small focal protrusion of the intervertebral disc at L4-L5 level on the right, extending into the region of the exiting right L5 root ➡.*





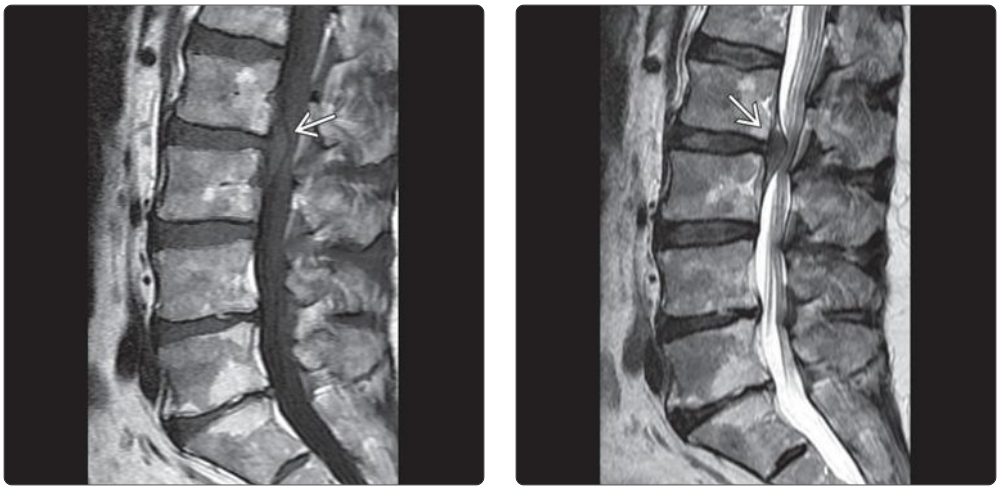
(Left) *Sagittal T1WI MR shows a large herniation at L3-L4 level with inferior migration of the fragment at L4 level ➡, consistent with sequestered disc (free fragment).* **(Right)** *Sagittal T2WI MR shows a large disc extrusion with relatively low signal intensity that is migrated inferiorly. There is severe effacement of the ventral thecal sac by the herniation.*



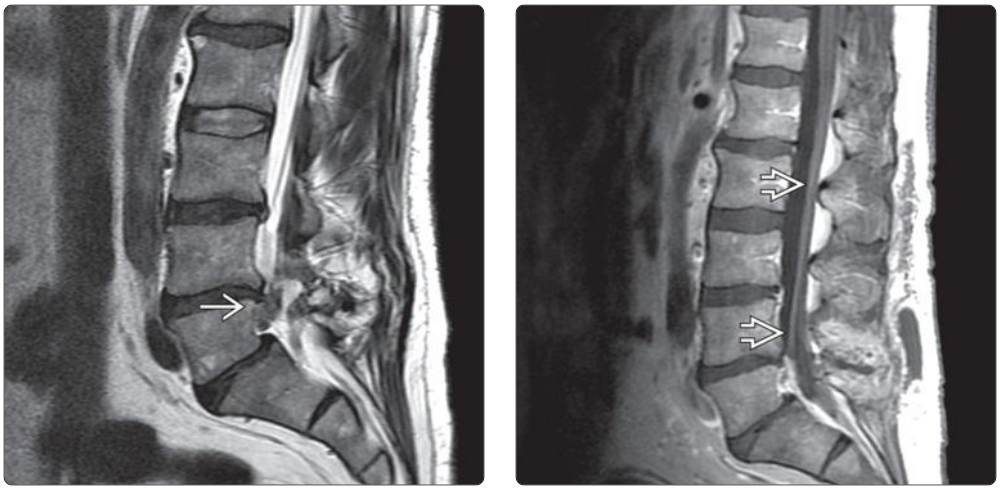


(Left) *Sagittal T2WI MR shows an extremely large herniation at L4-L5 level which completely effaces the thecal sac and severely compresses the cauda equina at that level. Smaller herniations are also present at L3-L4 and L5-S1.* **(Right)** *Sagittal T1WI MR shows a foraminal disc extrusion ➡ at L4-L5 obliterating perineural fat. The exiting L4 nerve root is impinged.*

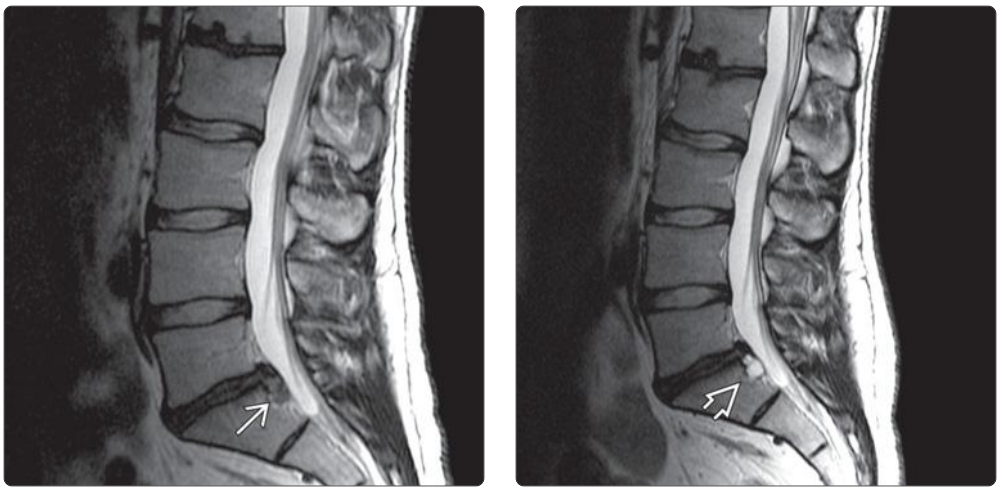
(Left) *Sagittal T1WI MR shows a large disc extrusion at L2-L3 level with severe effacement of the ventral thecal sac. There is extension of disc material both above and below the disc level.* **(Right)** *Sagittal T2WI MR shows a large disc extrusion at L2-L3. Note the "mushroom" or "toothpaste" appearance of extrusion with the base smaller than the portion extending into the epidural space. Fatty degenerative endplate changes are seen at L5-S1.*



(Left) *Sagittal T2WI MR shows a large left-sided extrusion with inferior migration from L4-L5 level.* **(Right)** *Sagittal postoperative enhanced MR study shows intense enhancement of a single root's dorsal and ventral components extending from L4-L5 to the conus. Single root enhancement is unusual but is typically associated with a disc herniation and root compression due to radiculitis or engorgement of the radicular vein.*



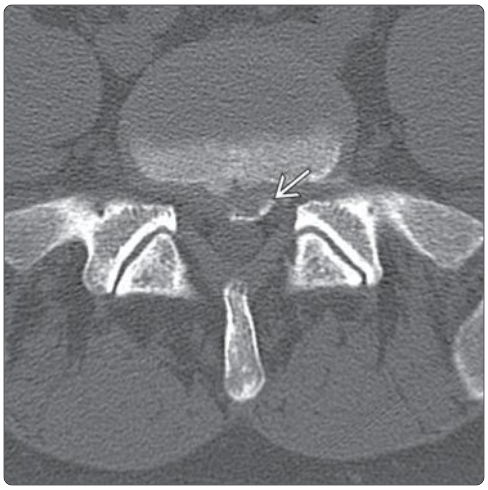
(Left) *Sagittal T2WI MR shows evolution of discal cyst development. Initial MR study shows a large disc extrusion at L5-S1 with inferior migration behind the superior S1 body.* **(Right)** *Sagittal T2WI MR shows evolution of discal cyst development with a well-defined fluid signal intensity that has replaced the initial disc herniation signal. This change may reflect involution of a prior hemorrhage associated with the disc herniation.*







(Left) *Sagittal T1 C+ MR with fat suppression shows intravertebral disc herniation ⇨ (Schmorl node) at the superior endplate of L4 with surrounding marrow enhancement.* **(Right)** *Axial nonenhanced CT with soft tissue window shows a large ventral soft tissue density arising from a large disc extrusion ⇨. Note the severe facet degenerative change bilaterally.*





(Left) *Axial NECT through the L5-S1 level shows partial calcification ⇨ of the moderate-sized central disc herniation.* **(Right)** *Plain film myelogram shows an extradural defect in the contrast column on the left at L4-L5 ⇨ with cutoff of the exiting left L5 root sleeve, which should be seen underneath the L5 pedicle ⇨. Note the normal S1 root ⇨.*





(Left) *Sagittal CECT following myelogram shows the large disc extrusion effacing the ventral thecal sac at the L4-L5 level, seen as a nonspecific extradural defect in the contrast column.* **(Right)** *Axial CECT following myelogram shows the large disc extrusion effacing the ventral thecal sac and extending eccentrically to the left. A small focus of gas is present within the disc herniation ⇨ from the degenerative vacuum phenomenon of the intervertebral disc.*

Intervertebral Disc Extrusion, Foraminal

KEY FACTS

TERMINOLOGY

- Extruded disc material within neural foramen
- Far lateral is disc material lateral to neural foramen

IMAGING

- Obliterated perineural fat in neural foramen on sagittal images
- Soft tissue mass contiguous with parent disc
- T1WI isointense to parent disc
- T2WI iso-, hypo-, or hyperintense to parent disc
- May enhance peripherally
- Often missed on myelography

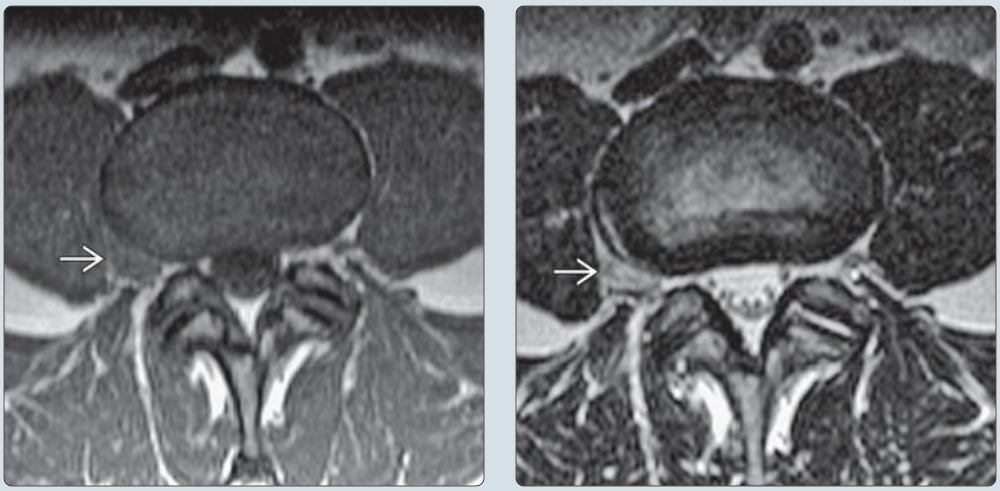
TOP DIFFERENTIAL DIAGNOSES

- Schwannoma
- Spinal nerve root diverticulum
- Large facet osteophyte

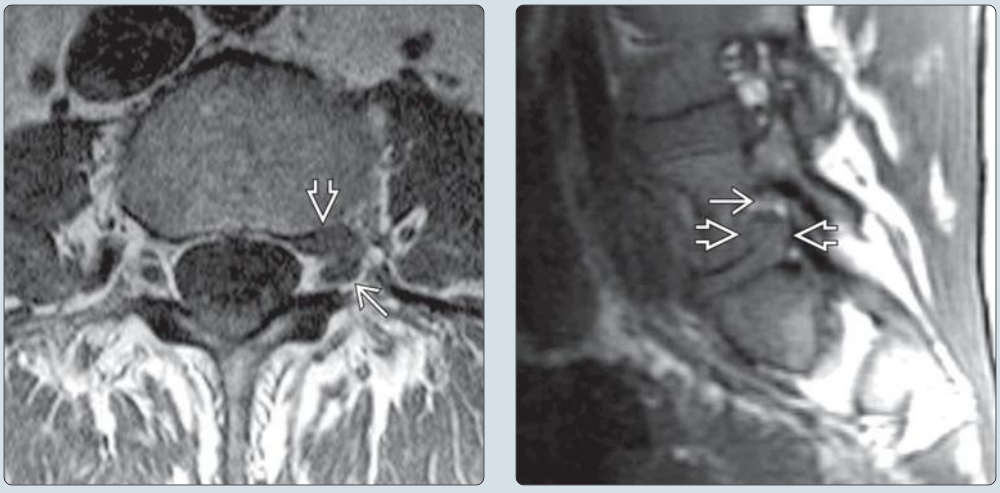
CLINICAL ISSUES

- May stabilize or resolve spontaneously
- Severe radicular pain
- Mass effect on exiting nerve root in narrow confines of neural foramen
- 5-10% of all disc herniations
- Surgery
  - Failed conservative therapy after 6-8 weeks; progressive deficits
  - Interlaminal approach with partial medial facetectomy
  - Endoscopic lateral approaches more commonly used, but no change in outcome relative to open procedures

(Left) *Axial T1WI MR shows large right lateral and intraforaminal disc herniation at L4-L5. The disc herniation involves the lateral aspect of the neural foramen and extends to the root ganglion level.* **(Right)** *Axial T2WI MR shows the large right lateral and intraforaminal disc herniation at L4-L5 as near isointense signal to adjacent paravertebral soft tissue. There is slight lateral displacement of the psoas muscle.*



(Left) *Axial T1WI MR shows a left L4-L5 foraminal disc extrusion, contacting the exiting left L4 nerve root at the level of the nerve root ganglion.* **(Right)** *Sagittal T1WI MR shows L5-S1 foraminal disc extrusion displacing the exiting L5 nerve root superiorly within the foramen.*



TERMINOLOGY

Synonyms

- Lateral/far lateral herniation
- Nonstandard: Lateral or foraminal herniated nucleus pulposus (HNP)

Definitions

- Extruded disc material within neural foramen
- Far lateral is disc material lateral to neural foramen

IMAGING

General Features

- Best diagnostic clue
  - Obscured perineural fat in neural foramen on sagittal images
    - Soft tissue mass contiguous with parent intervertebral disc
- Location
  - Lumbar: L3-L4 and L4-L5 most common
  - Cervical: C5-C6 and C6-C7 most common
  - Thoracic: Rare
    - Herniation: 3 in 1,000 incidence
- Size
  - Restricted by neural foramen
- Morphology
  - Typical "mushroom" appearance of central or subarticular disc extrusion absent
    - Conforms to neural foramen on sagittal images

Radiographic Findings

- Radiography
  - Nonspecific degenerative changes
    - Disc height loss, endplate sclerosis, facet arthropathy
    - Disc material not directly visualized

CT Findings

- NECT
  - Foraminal soft tissue mass
    - Same density as disc
  - Neural foramen not enlarged
- CECT
  - May show peripheral enhancement
- Bone CT
  - Same as radiography; nonspecific degenerative changes

MR Findings

- T1WI
  - Isointense to parent disc
- T2WI
  - Iso-, hypo-, or hyperintense to parent disc
    - Depending on hydration status
- T1WI C+
  - May enhance peripherally
  - Variable extent of impingement on exiting nerve root
    - Contacted, displaced, flattened
    - Nerve root may show post-gadolinium enhancement
      - Venous congestion ± inflammation
  - Extraforaminal component best seen on axial imaging
    - May involve adjacent nerve root
  - Disc height loss
  - Degenerative endplate changes

Nonvascular Interventions

- Myelography
  - Often missed on myelography
  - Sensitivity < 13%
  - Post-myelography CT improves sensitivity

Nuclear Medicine Findings

- Bone scan
  - Limited role
    - Radiotracer uptake in endplates and facet joints

Other Modality Findings

- Discography
  - Contrast extravasation into neural foramen and extraforaminal space
    - Invasive

Imaging Recommendations

- Best imaging tool
  - T1WI and T2WI MR in sagittal and axial planes

DIFFERENTIAL DIAGNOSIS

Schwannoma

- Enlarged neural foramen due to chronic remodeling
- "Dumbbell" appearance on axial imaging
  - 15% of schwannomas
- Diffuse post-contrast enhancement
  - Unless necrosis present

Spinal Nerve Root Diverticulum

- Cerebral spinal fluid intensity on all sequences
- No enhancement
- May opacify with contrast on myelography
- Enlarged neural foramen

Large Facet Osteophyte

- Hypointense on T1WI and T2WI
- Contiguous with facet joint
- Cephalad and posterior portion of neural foramen
- Osseous density on CT

Endplate Osteophyte

- Often associated with herniation
- Contiguous with endplate
- Hypointense on T1WI and T2WI
- Marrow intensity may be present

PATHOLOGY

General Features

- Etiology
  - Environmental factors
    - Heavy lifting
    - Twisting
    - Flexion and extension
    - Lateral bending
    - Acute trauma
  - Age
  - Poor posture

- Repetitive microtrauma
  - Endplate injury
  - Rim lesion: Horizontal tear in outer anulus at its attachment to ring apophysis
- Interruption of nutritional supply to disc
- Decreasing water content in nucleus pulposus
  - Proteoglycans replaced by fibrocartilage and fibrosis
  - Increasing stiffness
  - Decreasing intradisc pressure
- Ineffective cushioning
- Unequal force distribution to anulus
  - Concentric tear ± radial tear → disc herniation
- Genetics
  - Genetic predisposition to herniation
    - If positive family history of herniation
- Associated abnormalities
  - Spondylolisthesis
  - Scoliosis
  - Anatomy
    - Central nucleus pulposus
      - Proteoglycans and aminoglycans
      - High water content
      - Water content diminishes with age
      - Avascular
    - Peripheral anulus fibrosus
      - Tensile strength
      - Outer collagen fibers in concentric lamellae
      - Inner fibrocartilage

Microscopic Features

- Myxomatous degeneration of anulus fibrosus
- Composed of combination of nucleus pulposus, fragmented anulus, cartilage, and fragmented apophyseal bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Severe radicular pain
    - More symptomatic compared to other disc herniations
    - Mass effect on exiting nerve root in narrow confines of neural foramen
    - Lumbar: 25% sciatic and 75% femoral distribution
  - Other signs/symptoms
    - Muscle weakness
    - Positive femoral stretch test
- Clinical profile
  - Lateral bending, sitting, and increased intraabdominal pressure worsen pain
  - Pain relieved by rest with hip and knee in flexion

Demographics

- Age
  - 50s to 70s
    - Older than those with posterolateral herniation
- Gender
  - M = F
- Ethnicity
  - No racial predilection
- Epidemiology
  - Uncommon
    - 5-10% of all disc herniations

Natural History & Prognosis

- May stabilize or resolve spontaneously
- Favorable outcome with conservative measures

Treatment

- Conservative
  - Indicated if no or minor neurologic impairment
  - NSAIDs
  - Physical therapy
  - Selective nerve root block
    - Cortisone ± lidocaine
    - 60-80% asymptomatic after 1 year
- Surgery
  - Indications
    - Failed conservative therapy after 6-8 weeks; progressive deficits
  - Lumbar
    - Interlaminal approach with partial medial facetectomy
    - Pars interarticularis fenestration preserves facet integrity
    - Endoscopic lateral approaches more commonly used, but no change in outcome relative to open procedures
  - Cervical
    - Anterior or posterior approach
    - Laminotomy-foraminotomy
    - Discectomy; stabilization

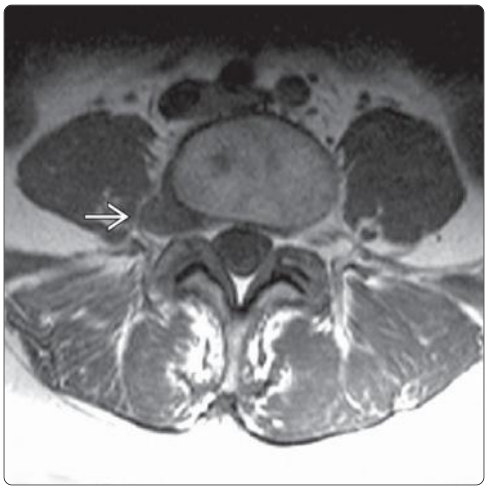
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Look for foraminal mass contiguous with disc on sagittal plane

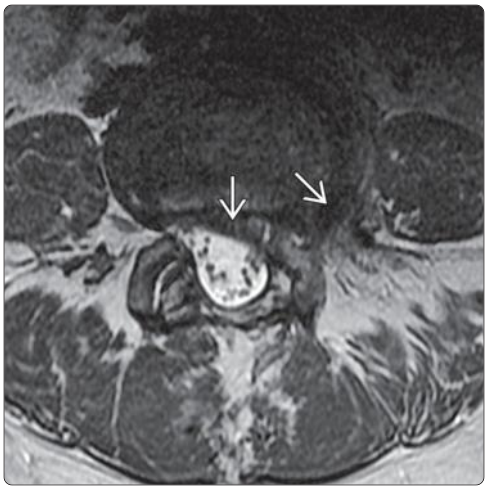
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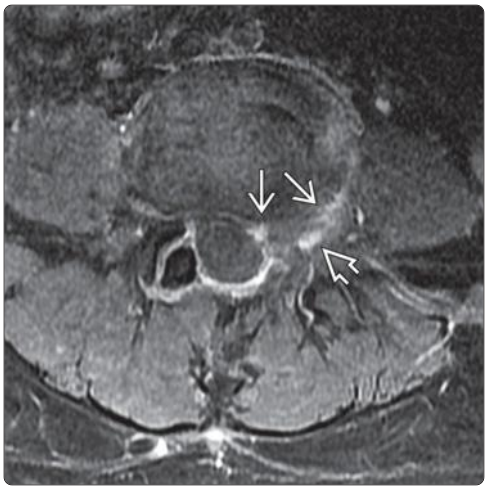
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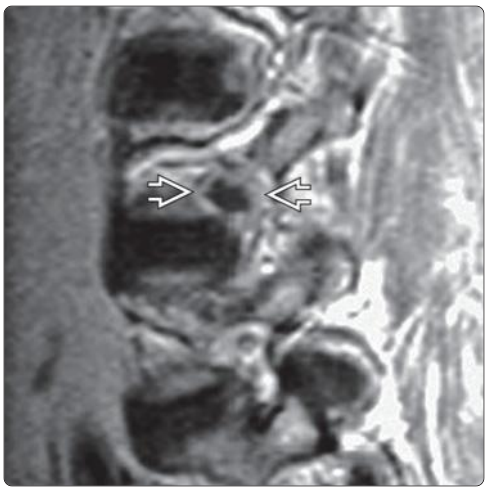
(Left) *Axial T1WI MR shows a large soft tissue lesion arising contiguous with the L4-L5 disc margin extending into the right foramen and the right lateral extraforaminal soft tissue* ⇨. **(Right)** *Sagittal T1WI shows the large lateral and foraminal disc herniation* ⇨ *obscuring the foraminal fat and displacing the exiting L4 nerve root superiorly* ⇨.





(Left) *Axial T2WI MR shows broad-based disc extrusion* ⇨ *extending from the left paracentral location into the left neural foramen in this postoperative patient.* **(Right)** *Axial T1WI C+ MR in a case of recurrent foraminal herniation shows slight peripheral enhancement* ⇨ *surrounding the large left lateral and foraminal herniation* ⇨.





(Left) *Sagittal T1WI MR shows a large L4-L5 disc extrusion* ⇨ *completely filling the neural foramen and obliterating perineural fat. The exiting L4 nerve root cannot be identified as a separate entity.* **(Right)** *Sagittal T1WI C+ MR shows L4-L5 foraminal disc extrusion with pronounced peripheral enhancement* ⇨.

Cervical Facet Arthropathy

KEY FACTS

TERMINOLOGY

- Facet arthrosis, degenerative facet disease, degenerative joint disease

IMAGING

- Osseous facet overgrowth impinging on neural foramina in conjunction with articular joint space narrowing
- Facet joint osteophytes producing foraminal narrowing
- "Mushroom cap" facet appearance
- Joint space narrowing with sclerosis and bone eburnation
- Intraarticular gas ("vacuum phenomenon")
- Enhancing inflammatory soft tissue changes surrounding facet joint are common
- Joint space narrowing, thinning of articular cartilage
- Facet effusions as linear hyperintensity

TOP DIFFERENTIAL DIAGNOSES

- Septic facet
- Healing facet fracture
- Inflammatory arthritides
- Paget disease
- Myositis ossificans
- Metastasis

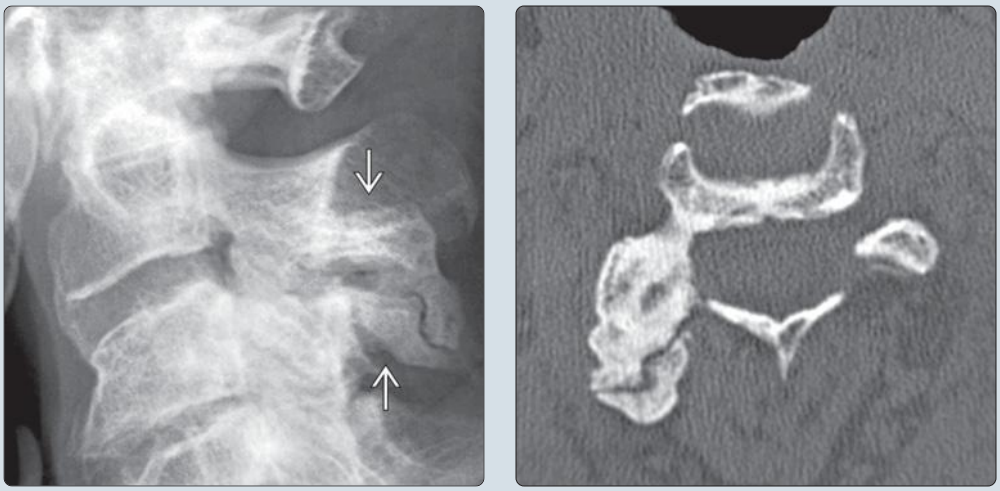
CLINICAL ISSUES

- Clinical pain aggravated by rest and alleviated by movement
- Poor correlation between duration/severity of pain and extent of facet degeneration

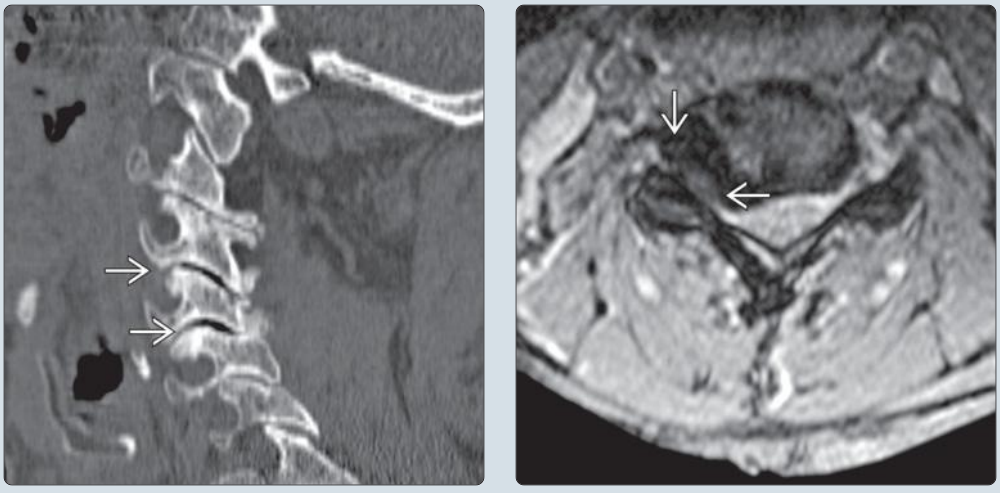
DIAGNOSTIC CHECKLIST

- Best detail with thin-section CT
- T2* images overemphasize degree of foraminal or central narrowing

(Left) *Lateral radiograph shows severe facet hypertrophic degenerative arthropathy at C2-C3 ➔ with marked hypertrophy of the facets.* **(Right)** *Axial NECT shows severe, exuberant right facet degenerative arthropathy.*



(Left) *Sagittal NECT shows multilevel hypertrophic facet degenerative arthropathy with "vacuum phenomenon" ➔, bony eburnation, and osteophyte formation.* **(Right)** *Axial T2* GRE MR shows severe right uncovertebral hypertrophic degenerative arthropathy with severe foraminal stenosis ➔ with facet degenerative change.*



TERMINOLOGY

Synonyms

- Facet arthrosis, degenerative facet disease, degenerative joint disease

Definitions

- Osteoarthritis of synovially lined apophyseal joints

IMAGING

General Features

- Best diagnostic clue
  - Osseous facet overgrowth impinging on neural foramina in conjunction with articular joint space narrowing
- Location
  - Normal anatomy
    - C2 → C7 levels have 2 sets of paired joints
    - Facet or zygapophyseal joints posteriorly, with oblique orientation
    - Uncovertebral joints (joints of Luschka, neurocentral joints) formed by curved edges of vertebral bodies at lateral margins
  - Arthropathy most common in mid/lower cervical spine
- Size
  - Facets may show minimal abnormality or large osteophytes approximating size of facet themselves
- Morphology
  - Osseous facet overgrowth and cartilage erosion with joint space narrowing

Radiographic Findings

- Radiography
  - Poor for demonstrating early facet degeneration
  - Later changes well demonstrated
  - Plain films demonstrate facet arthrosis well, not soft tissues
  - Oblique views demonstrate facet joints best
    - Facet joint osteophytes producing foraminal narrowing
    - Joint space narrowing with sclerosis and bone eburnation
    - Intraarticular gas ("vacuum phenomenon")

Fluoroscopic Findings

- Same as plain radiography; can observe motion abnormalities on flexion-extension

CT Findings

- NECT
  - CT more sensitive than plain films for detecting presence and degree of arthrosis
  - Facet joint osteophytes producing foraminal narrowing
  - "Mushroom cap" facet appearance
  - Joint space narrowing with sclerosis and bone eburnation
  - Intraarticular gas ("vacuum phenomenon")
  - CT myelography shows facet relationship to adjacent contrast-opacified thecal sac and nerve root sleeves
- CECT
  - Inflammatory soft tissue changes surrounding facet joint are common
    - May appear aggressive even in absence of infection

MR Findings

- T1WI
  - Enhancing inflammatory soft tissue changes surrounding facet joint are common
    - May appear aggressive even in absence of infection
- T2WI
  - Osteophyte proliferation limiting neural foramina
  - Joint space narrowing, thinning of articular cartilage
  - Facet effusions as linear hyperintensity
- T2* GRE
  - Shows osseous changes well
  - Tends to overemphasize degree of foraminal or central canal narrowing
- T1WI C+
  - Enhancement of synovium, epidural, and foraminal venous plexus

Nonvascular Interventions

- Myelography
  - CT myelography portrays facet relationship to adjacent contrast-opacified thecal sac and nerve root sleeves

Nuclear Medicine Findings

- Bone scan
  - Increased uptake with degenerative disc disease, facet arthropathy

Imaging Recommendations

- Plain films useful to demonstrate presence and severity of facet degenerative changes
- Sagittal and axial T1WI and T2WI best demonstrate degenerative facet compression of adjacent thecal sac
  - 3D gradient echo T2* for foraminal detail

DIFFERENTIAL DIAGNOSIS

Septic Facet

- T2 hyperintensity extending into adjacent soft tissues

Healing Facet Fracture

- Trauma history, search for fracture line

Inflammatory Arthritides

- Look for ankylosis or erosions, cranial settling, atlantoaxial subluxation (rheumatoid arthritis)

Paget Disease

- Thickened cortex, trabeculae, bony expansion

Myositis Ossificans

- Trauma history, circumscribed calcification

Tumor

- Metastasis
- Lymphoma
- Sarcoma

PATHOLOGY

General Features

- Etiology
  - Frequently seen in elderly population

  - Probably related to minor repetitive trauma coupled with abnormal biomechanics
  - May see earlier presentation after trauma, with kyphosis/scoliosis, or following surgical fusion at adjacent level
- Genetics
  - Genetic predisposition pondered but not proven
- Associated abnormalities
  - Frequently seen in conjunction with spondylosis
- Degenerative (hypertrophic) inflammatory changes in synovial joints
  - Progression similar to that in other joints except for early focal full thickness cartilage necrosis
- Normal bone mineralization (in contrast to rheumatoid arthritis)
- Joint traction during subluxation may produce gas in joint ("vacuum phenomenon")
- Associated with synovial cysts, degenerative disc disease

Staging, Grading, & Classification

- Grading scale (Pathria scale): Based on imaging studies
  - Grade 0: Normal
  - Grade I: Mild narrowing and joint irregularity
  - Grade II: Moderate narrowing and joint irregularity, sclerosis, and osteophyte formation
  - Grade III: Severe narrowing and almost total loss of joint space, sclerosis, and osteophyte formation

Gross Pathologic & Surgical Features

- Most common locations for facet arthropathy
  - Mid/lower cervical spine, lower lumbar spine
  - Thoracic spine uncommon due to rigidity from ribs/sternum
- Joint space narrowing, capsular laxity may permit subluxation of superior facet on inferior facet (degenerative spondylolisthesis)

Microscopic Features

- Findings are similar to those seen in synovial joints at other locations
  - Osseous proliferation
  - Fibrillation and erosion of articular joint cartilage
  - Preservation of bone density

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Mechanical neck pain (facet arthrosis syndrome)
    - Related to irritation of innervation of joint from medial branch(es) of dorsal primary ramus
    - Capsular distension, inflammatory synovitis, entrapment of synovial villi between 2 articular processes, or nerve impingement by osteophytes
    - Aggravated by rest, worse in morning, and relieved by repeated gentle motion
  - Radiculopathy
  - Other signs/symptoms
    - Facet disease may be asymptomatic incidental finding on imaging
    - Myelopathy (rarely)
- Clinical profile
  - Clinical pain syndrome produces symptoms that frequently are aggravated by rest and alleviated by movement
  - Poor correlation between duration and severity of pain, as well as extent of facet degeneration

Demographics

- Age
  - Universal after age 60
- Gender
  - No gender preference
- Epidemiology
  - Facet joint degeneration begins in 1st 2 decades of life

Natural History & Prognosis

- Can show progressive symptoms/signs
- Variable (depending on severity)

Treatment

- Mechanical pain → conservative medical therapy
- Foraminal narrowing with radiculopathy → foraminotomy
- Subluxation
  - Cervical → transarticular fusion with lateral mass screws or anterior cervical discectomy and fusion

DIAGNOSTIC CHECKLIST

Consider

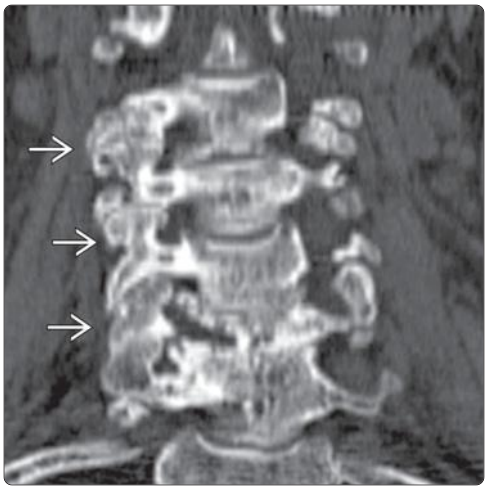
- Best detail of foraminal bony narrowing with thin-section CT with oblique reformats

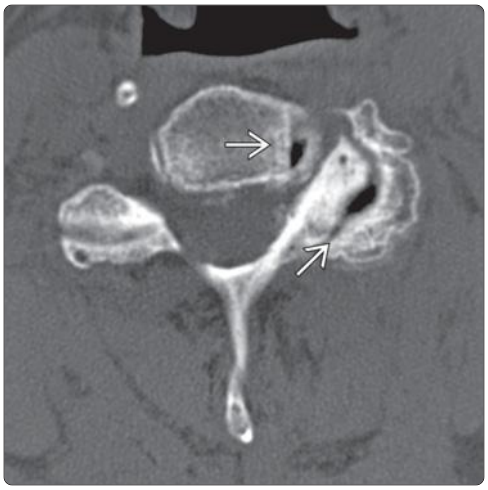
Image Interpretation Pearls

- 3D gradient echo T2* images overemphasize degree of foraminal or central canal narrowing

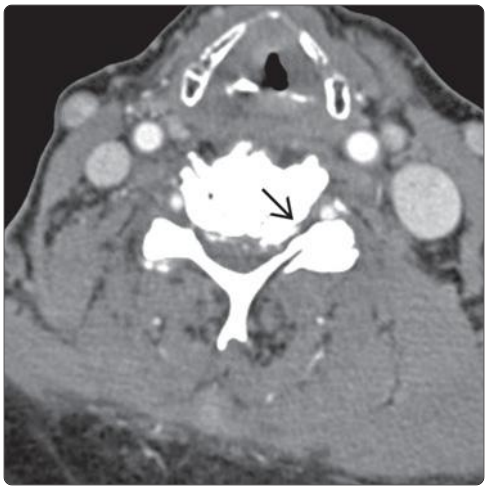
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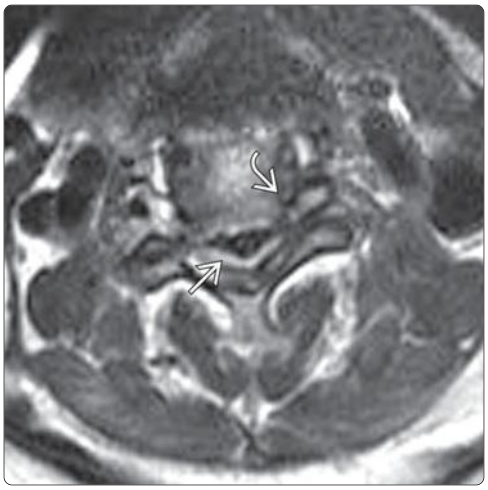
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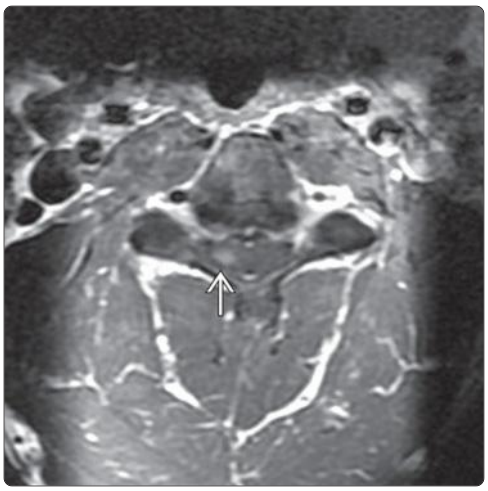
(Left) *Coronal NECT shows multilevel severe facet hypertrophic degenerative arthropathy with foraminal stenosis.* **(Right)** *Axial NECT shows severe left facet degenerative arthropathy and foraminal stenosis. "Vacuum phenomenon" is present in facet joint and uncovertebral joint.*





(Left) *Axial CECT shows severe canal stenosis due to diffuse disc bulge and diffuse broad-based herniations anteriorly with associated disc calcification. There is severe bilateral foraminal stenosis with uncovertebral joint hypertrophy and facet degeneration.* **(Right)** *Axial T2WI MR shows markedly thinned cord as relatively high signal outlined ventrally by disc osteophyte complex and posteriorly by thickened low signal ligamentum flavum. There is severe bilateral foraminal stenosis.*





(Left) *Axial T2* GRE MR shows moderate foraminal stenosis from mild facet degenerative change and uncovertebral joint hypertrophy. High signal is present within the cord from chronic compression and myelomalacia.* **(Right)** *Axial T1WI C+ MR shows foraminal stenosis from uncovertebral hypertrophy and facet degeneration with enhancing cord myelomalacia.*

Lumbar Facet Arthropathy

KEY FACTS

TERMINOLOGY

- Facet arthrosis, degenerative facet disease, degenerative joint disease, facet hypertrophy
- Osteoarthritis of synovially-lined lumbar apophyseal joints

IMAGING

- Osseous overgrowth impinging on foramina in conjunction with articular joint space narrowing
  - "Mushroom cap" facet appearance
  - Joint space narrowing with sclerosis/bone eburnation, ligamentum flavum hypertrophy
  - Intraarticular gas, effusion
  - Spondylolisthesis not uncommon
- CT more sensitive than plain films for detecting presence and degree of arthrosis
  - Facet hypertrophic degenerative arthropathy, particularly superior articular facet extending into lumbar foramen
- MR best demonstrates degenerative facet compression of thecal sac and fat-filled neural foramina
  - Enhancing inflammatory soft tissue changes surrounding facet joint not uncommon
- Consider CT myelography if MR contraindications or MR does not demonstrate facet relationship to neural foramina

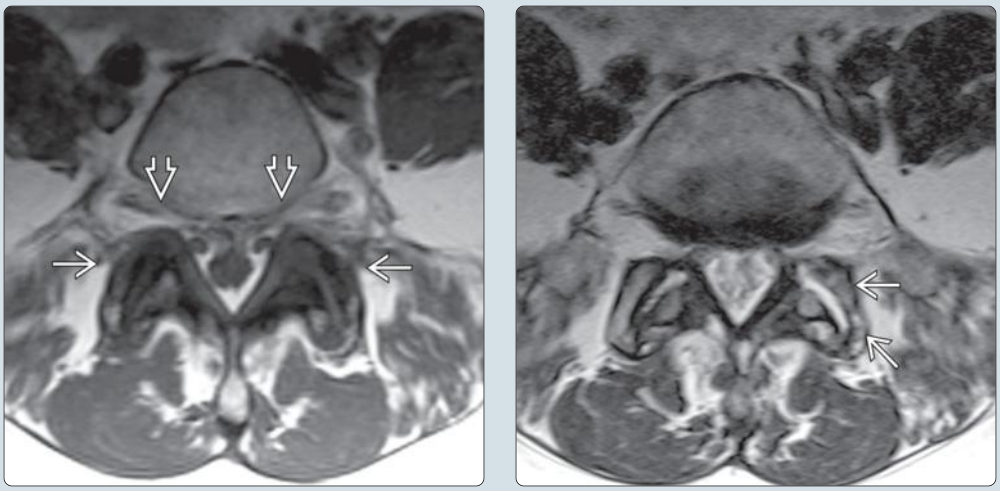
TOP DIFFERENTIAL DIAGNOSES

- Septic facet
- Inflammatory arthritides
- Paget disease
- Tumor
  - Metastasis
  - Lymphoma

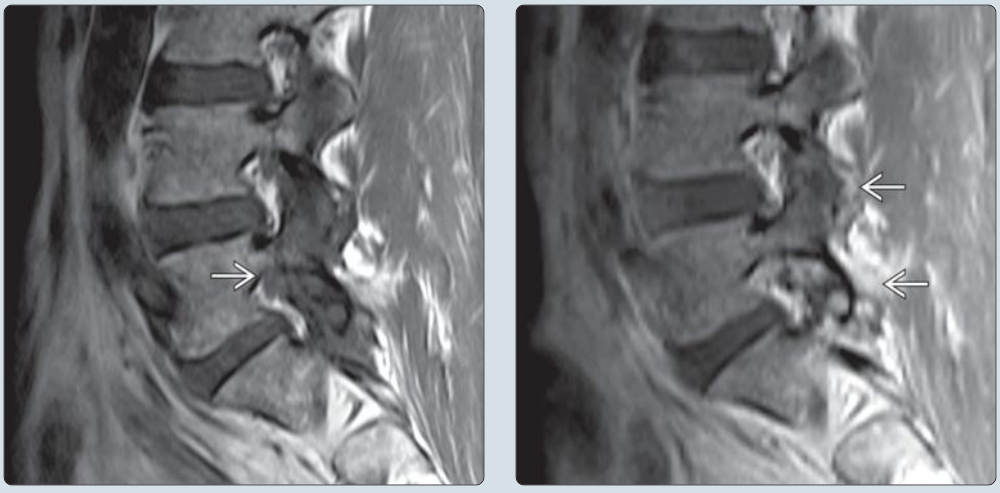
DIAGNOSTIC CHECKLIST

- Look for associated **synovial cysts**

(Left) *Axial T1WI MR shows marked facet hypertrophic degenerative arthropathy with enlarged facets, producing moderately severe foraminal stenosis.* **(Right)** *Axial T2WI MR shows bilateral facet hypertrophy and small bilateral facet effusions. There is mild deformity of the posterior margin of the thecal sac, but no significant central stenosis.*



(Left) *Sagittal T1WI MR shows L4-L5, L5-S1 facet degenerative arthropathy with bony hypertrophy and low signal involving the facets from bony sclerosis. There is narrowing of the L5-S1 foramen primarily by the hypertrophied superior articular facet of S1.* **(Right)** *Sagittal T1WI C+ MR shows enhancement of the L4-L5, L5-S1 facets due to degenerative arthropathy.*



TERMINOLOGY

Synonyms

- Facet arthrosis, degenerative facet disease, degenerative joint disease, facet hypertrophy

Definitions

- Osteoarthritis of synovially lined lumbar apophyseal joints

IMAGING

General Features

- Best diagnostic clue
  - Osseous facet overgrowth impinging on neural foramina in conjunction with articular joint space narrowing
- Location
  - Facet joints of lumbar spine
- Size
  - Minimal → large osteophytes approximating size of facet themselves
- Morphology
  - Osseous facet overgrowth and cartilage erosion with joint space narrowing

Radiographic Findings

- Radiography
  - Poor for mild facet degeneration, severe well demonstrated
  - Oblique views demonstrate facet joints best
    - "Mushroom cap" facet appearance
    - Joint space narrowing with sclerosis/bone eburnation
    - Intraarticular gas ("vacuum phenomenon")
    - Spondylolisthesis not uncommon

Fluoroscopic Findings

- Same as plain radiography; can observe motion abnormalities on flexion/extension

CT Findings

- NECT
  - CT more sensitive than plain films for detecting presence and degree of arthrosis
  - Facet joint osteophytes producing foraminal narrowing
  - Joint space narrowing with sclerosis and bone eburnation
  - CT myelography shows facet relationship to adjacent contrast-opacified thecal sac and nerve root sleeves
- CECT
  - Inflammatory soft tissue changes surrounding facet joint are common
    - May appear aggressive even in absence of infection

MR Findings

- T1WI
  - Joint space narrowing, thinning of articular cartilage
    - Some patients show variable synovial thickening
    - Irritation of synovium can produce synovial hyperplasia with paradoxical joint space widening
- T2WI
  - Osteophyte proliferation narrowing neural foramina
  - Foraminal compromise often combination of disease
    - Facet hypertrophic degenerative arthropathy, particularly superior articular facet extending into lumbar foramen
    - Ligamentous buckling/thickening
    - Disc bulge/osteophyte extending into inferior portion of lumbar foramen
    - Joint space effusions
- STIR
  - May show marrow hyperintensity due to edema, type I endplate-like changes
- T2* GRE
  - Shows osseous changes well
  - Overemphasize degree of foraminal or central canal narrowing
- T1WI C+
  - Enhancing inflammatory soft tissue changes surrounding facet joint not uncommon
    - May appear aggressive even in absence of infection

Nonvascular Interventions

- Myelography
  - CT myelography shows facet relationship to adjacent contrast-opacified thecal sac and nerve root sleeves

Nuclear Medicine Findings

- Bone scan
  - Increased uptake with degenerative disc disease, facet arthrosis

Imaging Recommendations

- Plain films to demonstrate presence and severity of facet changes
- MR best demonstrates degenerative facet compression of thecal sac and fat-filled neural foramina
- Consider CT myelography if MR contraindications or MR does not demonstrate facet relationship to neural foramina

DIFFERENTIAL DIAGNOSIS

Septic Facet

- T2 hyperintensity extending into soft tissues, epidural abscess/phlegmon

Inflammatory Arthritides

- Look for ankylosis/erosions involving facet and SI joints

Paget Disease

- Thickened cortex, trabeculae, bony expansion

Tumor

- Metastasis
- Lymphoma
- Soft tissue enhancing mass, bone destruction

Tumoral Calcinosis

- Large, lobulated calcific mass, no enhancing soft tissue component (rare)

PATHOLOGY

General Features

- Etiology
  - Elderly population

- Probably related to minor repetitive trauma coupled with abnormal biomechanics
- May see earlier presentation after trauma, with kyphosis/scoliosis, or following surgical fusion at adjacent level

- Genetics
  - No genetic basis proven
- Associated abnormalities
  - Foraminal stenosis, central canal stenosis
  - Synovial cyst, extending dorsal or ventral to facet joint
- Normal bone mineralization (in contrast to rheumatoid arthritis)
- Degenerative (hypertrophic) inflammatory changes in synovial joints
  - Progression is similar to that seen in other joints except for early focal full thickness cartilage necrosis
- Joint traction during subluxation may produce gas in joint ("vacuum phenomenon")
- Associated with synovial cysts, degenerative disc disease, canal stenosis
- Biomechanical stress is exacerbated in facets that are more horizontal in sagittal plane and more angled in axial plane

Staging, Grading, & Classification

- Grading scale (Pathria et al); based on imaging studies
  - Grade 0: Normal
  - Grade I: Mild narrowing and joint irregularity
  - Grade II: Moderate narrowing and joint irregularity, sclerosis, and osteophyte formation
  - Grade III: Severe narrowing and almost total loss of joint space, sclerosis, and osteophyte formation

Gross Pathologic & Surgical Features

- Joint space narrowing, capsular laxity may permit subluxation of superior facet on inferior facet (degenerative spondylolisthesis)

Microscopic Features

- Findings similar to those seen in synovial joints at other locations
  - Osseous proliferation
  - Fibrillation and erosion of articular joint cartilage
  - Preservation of bone density

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Mechanical back pain (facet arthrosis syndrome) most common symptom
    - Low back stiffness
    - Pain related to irritation of innervation of joint from medial branch dorsal primary ramus, capsular distension, inflammatory synovitis, entrapment of synovial villi between 2 articular processes, or impingement by osteophytes
    - Aggravated by rest, worse in morning, and relieved by repeated gentle motion
    - Pain is centered in hips, buttocks, or thighs; does not extend below knees, has no radicular pattern, and is aggravated by hyperextension
    - Straight leg raise usually negative
  - Other signs/symptoms
    - Facet disease may be asymptomatic incidental finding on imaging
- Clinical profile
  - Symptoms frequently aggravated by rest and alleviated by movement
  - Poor correlation between duration and severity of pain and extent of degeneration

Demographics

- Age
  - Virtually universal after age 60 years
  - Identifiable to varying degrees in majority of adults
- Gender
  - No gender preference
- Epidemiology
  - Facet joint degeneration begins in 1st 2 decades of life

Natural History & Prognosis

- Can show progressive symptoms/signs
- Variable (depending on severity)

Treatment

- Mechanical pain: Conservative medical therapy
- Foraminal narrowing with radiculopathy: Nerve root blocks, foraminotomy
- Subluxation
  - Lumbar: Posterior fusion with pedicle screws and rods (rarely in conjunction with anterior interbody fusion)

DIAGNOSTIC CHECKLIST

Consider

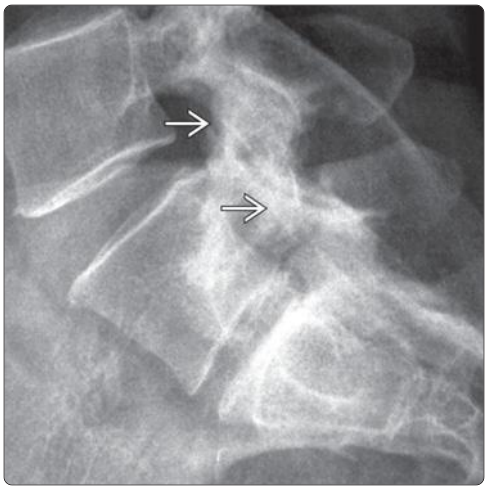
- Best bony detail with thin-section CT with reformats

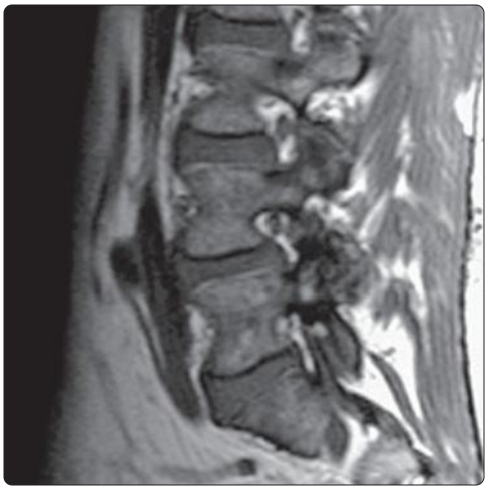
Image Interpretation Pearls

- Multifactorial nature of foraminal narrowing: Bony facet, ligament, disc, osteophyte
- Look for associated **synovial cysts**

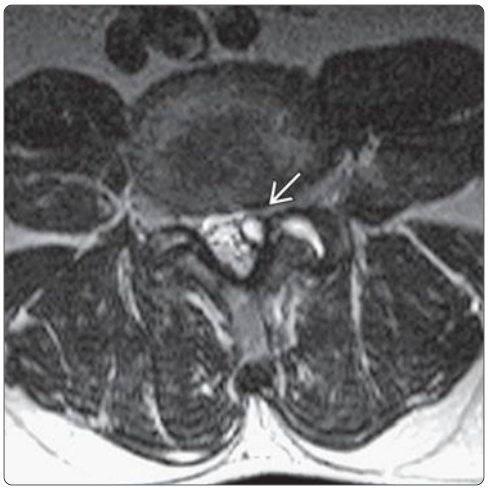
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(Left) *Lateral radiograph shows facet hypertrophy and bony eburnation of the L4-L5 and L5-S1 facets* ➡. **(Right)** *Sagittal T1WI MR shows degenerated facets spanning L3-L4 through L5-S1 levels, narrowing the neural foramina from the dorsal aspect, with superior articular facet hypertrophy causing the impingement.*





(Left) *Axial T2WI MR shows degenerated left facet with effusion and synovial cyst compressing the left dorsal aspect of thecal sac* ➡. **(Right)** *Axial T2WI MR shows lumbar juxtaarticular cyst as focal lesion of low signal with central T2 hyperintensity involving the left L4 ligamentum flavum* ➡. *The lesion compresses the left posterior lateral aspect of the thecal sac. Note the severe facet degenerative change with facet hypertrophy and bilateral facet effusions.*





(Left) *Axial T2WI MR shows cystic interspinous ligament degeneration (Baastrup disease)* ➡ *and facet degenerative change with small facet effusions* ➡. **(Right)** *Axial T2WI MR shows degenerative facet hypertrophy with bilateral effusions* ➡, *thickened ligamentum flavum* ➡, *and posteriorly directed juxtaarticular (synovial) cysts* ➡.

Facet Joint Synovial Cyst

KEY FACTS

TERMINOLOGY

- Juxtaarticular cyst
- Facet joint ganglion cyst
- Synovial cyst formed from degenerative facet joint

IMAGING

- Posterolateral extradural cystic mass communicating with facet joint
- Posterolateral to thecal sac
- Adjacent to facet joint
- Lumbar spine: 90%

TOP DIFFERENTIAL DIAGNOSES

- Extruded disc fragment
- Nerve sheath tumor
- Septic facet arthritis/epidural abscess
- Asymmetric ligamentum flavum hypertrophy

PATHOLOGY

- Facet osteoarthropathy
- Facet instability and hypermobility
- Greatest mobility at L4-L5
- Degenerative spondylolisthesis

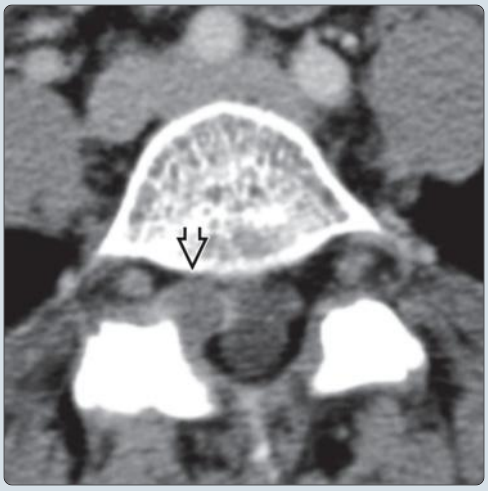
CLINICAL ISSUES

- Chronic low back pain, radicular symptoms
- May spontaneously regress
- High postsurgical success rate in symptomatic patients
- Myelopathy if cervical or thoracic location
- Treatment
  - Conservative
  - Laminectomy with cyst excision
  - Percutaneous cyst aspiration/steroid injection





(Left) *Axial graphic depicts a right facet joint synovial cyst. Fluid expands the right facet joint, extending into subarticular recess in a loculated collection. There is mass effect on the thecal sac.* **(Right)** *Axial T2WI MR shows bilateral subarticular hyperintense masses ⇨ at L4-L5, consistent with bilateral facet synovial cysts. There is bilateral facet degeneration with facet effusions.*





(Left) *Axial CECT shows a right-sided circumscribed juxtaarticular mass ⇨, slightly hyperdense than CSF with a thin rim of mild enhancement, causing subarticular stenosis.* **(Right)** *Axial bone CT after myelography with coronal reformation shows an extradural mass abutting the right L4-L5 facet joint, severely distorting the thecal sac.*

TERMINOLOGY

Synonyms

- Juxtaarticular cyst
- Facet joint ganglion cyst

Definitions

- Synovial cyst formed from degenerative facet joint

IMAGING

General Features

- Best diagnostic clue
  - Posterolateral extradural cystic mass communicating with facet joint
- Location
  - Posterolateral to thecal sac
  - Adjacent to facet joint
  - Lumbar spine: 90%
    - 70-80% at L4-L5
    - L3-L4 and L5-S1: Less common
  - Uncommon
    - Cervical and thoracic spine
    - Foraminal
    - Far lateral extraforaminal
    - Bilateral
- Size
  - 1-2 cm
- Morphology
  - Round
  - Lobulated
  - Sharply marginated

Radiographic Findings

- Radiography
  - Nonspecific degenerative changes
    - Worst at level of synovial cyst
      - Disc space loss
      - Vacuum disc phenomenon
      - Endplate sclerosis and spurring
      - Facet arthropathy
      - Spondylolisthesis without spondylolysis
      - Lateral listhesis
      - Scoliosis

CT Findings

- NECT
  - Difficult to detect because of fluid density
  - Increased density from hemorrhage
  - Mural calcifications
  - Intracystic or facet joint gas
- CECT
  - Mild rim enhancement
- Bone CT
  - Same as radiographic findings
  - Better osseous detail with sagittal and coronal reformation
  - Erosion of vertebral arch uncommon

MR Findings

- T1WI
  - Hypointense
    - Similar to CSF
  - Hyperintense
    - Hemorrhagic or proteinaceous content
- T2WI
  - Hyperintense
    - If direct communication with facet joint present
    - Best seen on sagittal T2WI
  - Hypointense
    - Hemorrhage
  - Low intensity rim
- STIR
  - Hyperintense
- T1WI C+
  - Enhancing wall
  - Well circumscribed
  - Associated ligamentum flavum hypertrophy and facet arthropathy
    - Variable central canal, subarticular, or lateral recess narrowing
      - Impingement of traversing nerve root
  - MR has high sensitivity and specificity

Nonvascular Interventions

- Myelography
  - Circumscribed posterolateral extradural mass
  - Central canal and subarticular narrowing
  - High sensitivity
  - Low specificity

Nuclear Medicine Findings

- Bone scan
  - Radiotracer uptake in vertebral endplates and posterior elements
    - Degenerative changes

Other Modality Findings

- Facet joint arthrogram
  - Contrast → facet joint into synovial cyst
    - Not a consistent finding

Imaging Recommendations

- Best imaging tool
  - MR of lumbar spine
    - Axial and sagittal T2WI/STIR

DIFFERENTIAL DIAGNOSIS

Extruded Disc Fragment

- Not contiguous with facet joint
- Usually anterior epidural
- Posterolateral uncommon
- Not as hyperintense on T2WI

Nerve Sheath Tumor

- Intradural extramedullary most common
- "Dumbbell" lesion less common
- Intense post-contrast enhancement

Septic Facet Arthritis/Epidural Abscess

- Abscess extending out from facet joint
  - Simulating synovial cyst

- Surrounding soft tissue edema and enhancement
- Adjacent marrow edema

Asymmetric Ligamentum Flavum Hypertrophy

- Hypointense on T2WI
- More broad-based contour
- Diffuse ligamentum flavum thickening

Ganglion Cyst

- Likely from ligamentum flavum
- Difficult to distinguish by imaging
- Different pathologically
- Contains myxoid material
- Lined by fibrous connective tissue capsule

PATHOLOGY

General Features

- Etiology
  - Stress loading on lumbar spine
  - Facet osteoarthropathy
  - Joint fluid accumulation
  - Facet instability and hypermobility
    - Greatest mobility at L4-L5
  - Synovial proliferation
  - Cyst formation
- Associated abnormalities
  - Degenerative spondylolisthesis
  - Rheumatoid arthritis
  - Calcium pyrophosphate deposition disease
- Thickened connective tissue and synovium
- Invariably associated with degenerative disc and facet disease

Gross Pathologic & Surgical Features

- Encapsulated
- Serous, mucinous, ± hemorrhagic fluid
- White myxoid material

Microscopic Features

- Fibrous connective tissue
- Inflammatory cell infiltration
- Calcific deposits
- Lined with hypervascular synovium

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic low back pain
  - Other signs/symptoms
    - Acute pain from hemorrhage
    - Radicular symptoms
    - Neurogenic claudication
    - Cauda equina syndrome
    - Myelopathy if cervical or thoracic

Demographics

- Age
  - ≥ 60
- Gender
  - More common in females
- Ethnicity
  - No racial predilection

Natural History & Prognosis

- May spontaneously regress
- High postsurgical success rate in symptomatic patients

Treatment

- Conservative
  - Bed rest
  - Analgesics
  - Epidural and facet injection
  - Bracing
- Laminectomy with cyst excision
  - In setting of intractable pain
  - To relieve nerve compression and central stenosis
  - Hemilaminectomy and flavectomy also performed
- Less permanent
  - Percutaneous cyst aspiration
    - Under CT guidance
  - Steroid injection into cyst
    - 1/2 to 2/3 of patients have excellent symptomatic relief after 6 months

DIAGNOSTIC CHECKLIST

Consider

- Thin-section sagittal T2WI may show communication between facet joint and adjacent synovial cyst

Image Interpretation Pearls

- Well-circumscribed T2 hyperintense mass
- Thin hypointense rim
- Posterolateral to thecal sac
- Abutting facet joint

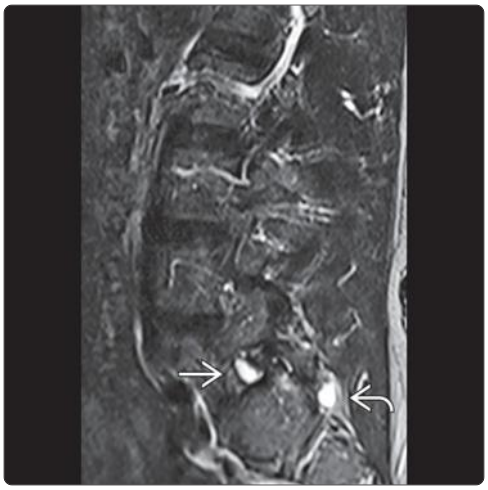
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(Left) *Axial T2WI MR shows a very large, well-circumscribed hyperintense mass with a hypointense rim abutting the left L4-L5 facet joint, severely narrowing the central canal.* **(Right)** *Axial T1WI MR with fat suppression shows a hyperintense extradural mass within the right subarticular/lateral recess, compatible with a hemorrhagic or proteinaceous synovial cyst.*





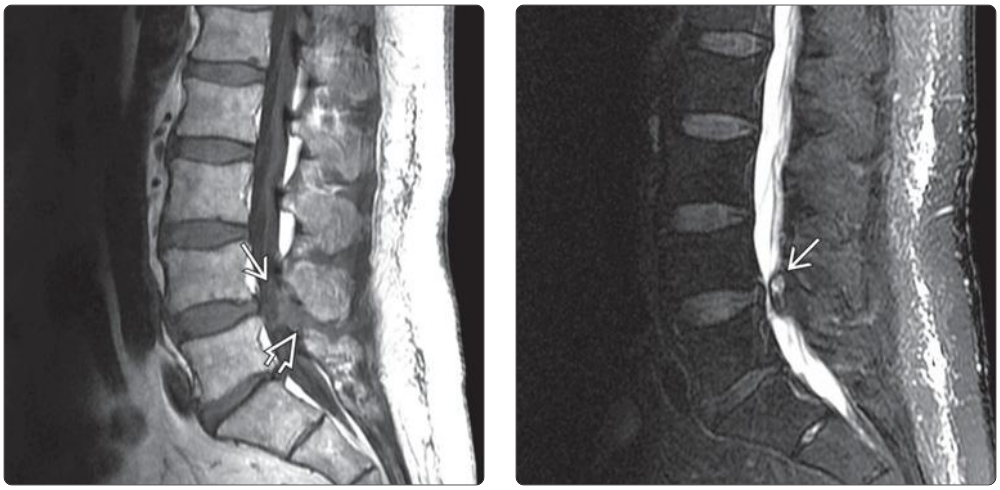
(Left) *Sagittal STIR MR shows a severely degenerated facet joint with synovial cysts that extend into the neural foramen ➡ and dorsally into posterior soft tissues ➡. Foraminal location is unusual but may occur due to far lateral extension of facet joint, as well as lateral extension of the ligamentum flavum into superior and posterior margin of foramen along superior articular facet.* **(Right)** *Axial T2WI MR shows a well-defined juxtaarticular cyst, which extends into the posterior soft tissues ➡.*



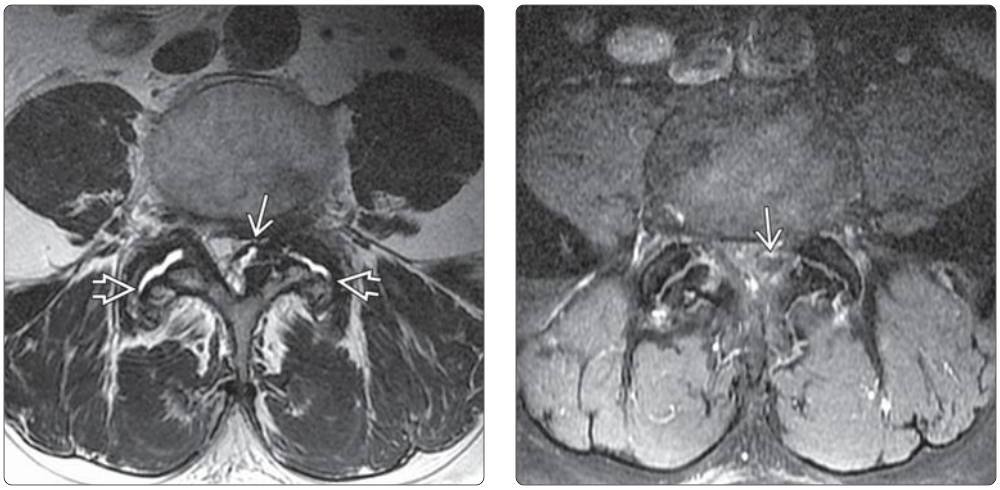


(Left) *Sagittal NECT shows a well-defined mass ➡ with central near fluid attenuation, which is difficult to further characterize without axial images. This cyst arises from the left facet joint.* **(Right)** *Axial NECT shows a well-defined mass with central fluid attenuation ➡ arising from the left facet joint and severely compressing the left lateral margin of the thecal sac. Note the facet hypertrophy and degenerative change.*

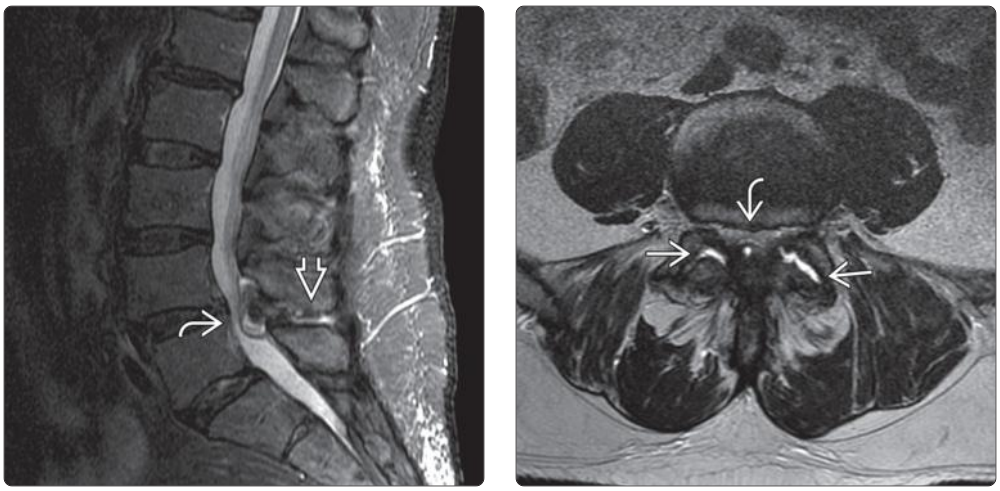
(Left) *Sagittal T1WI MR shows an ill-defined lesion merging with the intermediate signal of the ligamentum flavum and obscuring the dorsal margin of the thecal sac and normal epidural fat at the L4-L5 disc level.* **(Right)** *Sagittal STIR MR shows a focal lesion of low peripheral signal with central T2 hyperintensity involving the left L4 ligamentum flavum. There is severe thecal sac compression.*



(Left) *Axial T2 TSE MR shows a focal lesion of low peripheral signal with central T2 hyperintensity arising from the left L4 ligamentum flavum. Note the severe facet degenerative change with facet hypertrophy and bilateral facet effusions.* **(Right)** *Axial T1 C+ FS MR shows no significant enhancement within the lesion adjacent to the left facet and ligamentum flavum, consistent with a synovial cyst. Peripheral enhancement may occur with these lesions.*



(Left) *Sagittal STIR MR shows degeneration of the interspinous ligament with T2 hyperintensity and a large juxtaarticular cyst with central hyperintensity compressing the dorsal thecal sac.* **(Right)** *Axial T2 TSE MR shows severe canal stenosis with facet degenerative change, facet effusions, and midline juxtaarticular (facet) cysts within the ligamentum flavum.*







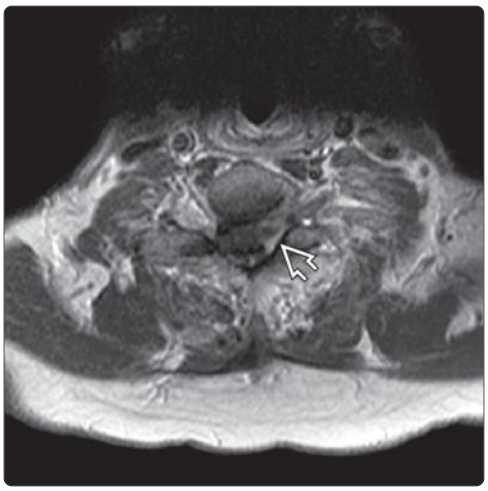
(Left) *Sagittal T2WI MR shows peripheral low signal and central heterogeneous hyperintensity ⇨ within a dorsal extradural mass at C7-T1 due to a juxtaarticular cyst.* **(Right)** *Sagittal T1 C+ MR shows a peripherally enhancing extradural lesion ⇨ based along the posterior elements at C7-T1, consistent with synovial cyst.*





(Left) *Sagittal T1 C+ MR shows a peripherally enhancing mass with central low signal at the C7 level due to a synovial cyst ⇨ in this patient with multilevel severe facet degenerative change. A cystic schwannoma could also present with this appearance.* **(Right)** *Sagittal STIR MR shows a mass with peripheral low signal and central hyperintensity ⇨ within the spinal canal at the C7 level, due to a facet synovial cyst.*





(Left) *Axial T1 C+ MR defines the relationship of the right synovial cyst ⇨ with the facet joint, displacing the cord to the left ⇨.* **(Right)** *Axial T1 C+ MR shows an extradural mass on the left side of the thecal sac ⇨ in this patient with a cervical synovial cyst. There is peripheral enhancement and the cyst is contiguous with the ligamentum flavum.*

Baastrup Disease

KEY FACTS

TERMINOLOGY

- Close approximation and contact of adjacent spinous processes with enlargement, flattening, and sclerosis of apposing interspinous surfaces
- Putative source of back pain exacerbated by extension, relieved with flexion

IMAGING

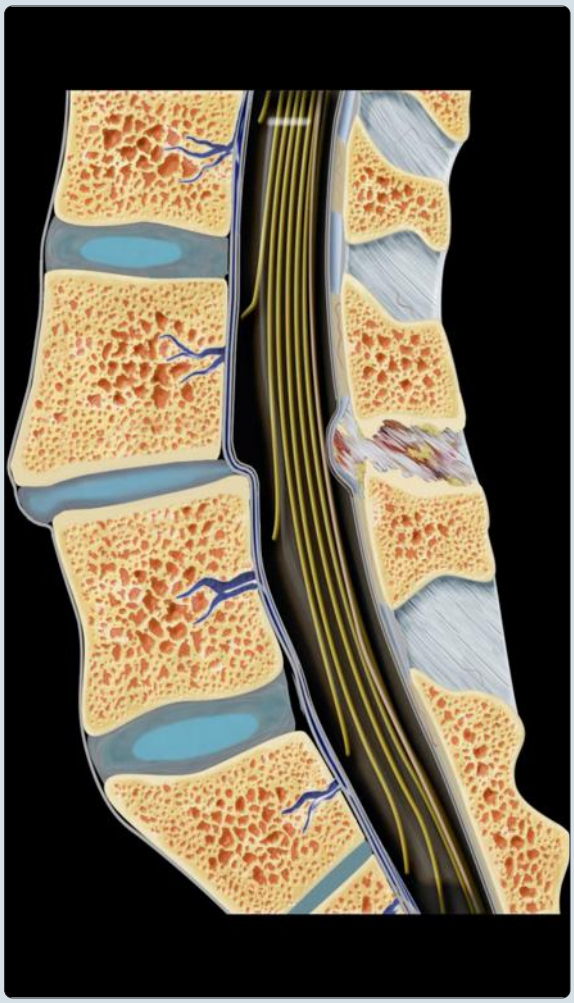
- Plain films/CT: Contact of adjacent spinous processes with sclerosis ("kissing" spinous processes)
- T1WI: Apposing surfaces of spinous processes may show low signal with bony sclerosis or marrow edema
- T2WI: Increased signal from interspinous ligament cystic degeneration and bursa formation

TOP DIFFERENTIAL DIAGNOSES

- Ligamentum flavum hypertrophy
- Osteomyelitis, pyogenic
- Metastases, lytic osseous
- Juxtaarticular cyst
- Tumoral calcinosis

CLINICAL ISSUES

- Interspinous bursitis appearance seen in 8% of subjects undergoing MR of lumbar spine for back or leg pain
- Associations with Baastrup
  - Increasing age
  - Central canal stenosis
  - Anterolisthesis
- Treatment
  - Direct steroid injection into bursa
  - X-Stop or other interspinous spacing device for treatment of canal stenosis
  - Resection of offending spinous processes



Sagittal graphic shows degeneration of the L4-L5 interspinous ligament with spinous process flattening sclerosis of apposing interspinous surfaces. There is associated ligamentum flavum hypertrophy and spondylolisthesis.



Sagittal STIR MR shows irregularity of the apposed margins of L4-L5 spinous processes, with ligamentum flavum and interspinous process hypertrophy. Cystic degeneration is shown as focal high signal.

TERMINOLOGY

Synonyms

- Spinous process impingement syndrome
- "Kissing" spinous process disease
- Interspinous ligament bursitis (diarthrosis interspinosae)
- Baastrup sign

Definitions

- Defined by Baastrup in 1933
- Close approximation and contact of adjacent spinous processes with enlargement, flattening, and sclerosis of apposing interspinous surfaces
- Putative source of back pain exacerbated by extension, relieved with flexion

IMAGING

General Features

- Best diagnostic clue
  - Contact of adjacent spinous processes with sclerosis ("kissing" spinous processes)
- Location
  - Lumbar spinous processes and interspinous ligaments
- Size
  - Ligament abnormality 1-3 cm long
    - Shallow transverse diameter
    - Vertical height a few mm
- Morphology
  - Variable, with involvement of adjacent spinous processes, interspinous ligament, and adjacent soft tissues

Radiographic Findings

- Flexion and extension radiographs
  - Contact of apposing spinous processes, with sclerosis of superior and inferior aspects of adjacent processes
- Spinous processes may be enlarged

CT Findings

- Similar to plain film findings with bony sclerosis of adjacent spinous processes
- May detect associated juxtaarticular cysts
- Stress-related fractures through spinous processes have been reported

MR Findings

- T1WI
  - Low or normal striated appearance of interspinous ligament
  - Diminished height of interspinous ligament with close apposition of adjacent processes
  - Apposing surfaces of spinous processes may show low signal with bony sclerosis or marrow edema
  - Look for associated ligamentum flavum hypertrophy
- T2WI
  - Increased interspinous ligament soft tissue
    - Low signal from ligament hypertrophy
    - Increased signal from interspinous ligament cystic degeneration and bursa formation
  - Look for associated dorsal and ventral juxtaarticular cysts
- T1WI C+
  - Nonspecific enhancement of interspinous ligament
  - Look for peripheral enhancement of degenerated facet and associated synovial cysts

Nuclear Medicine Findings

- Bone scan
  - Nonspecific increased uptake from spinous processes
  - Increased uptake from interspinous ligaments reported with FDG PET

Other Modality Findings

- Facet arthrography
  - Facet injection may communicate to adventitious bursa within interspinous ligament
  - Opacification of bilateral facets and midline interspinous ligament bursa gives "butterfly" contrast pattern

Imaging Recommendations

- Best imaging tool
  - MR to show T2 hyperintensity of interspinous ligament, associated degenerative changes of facets (effusions), ligamentum flavum hypertrophy, juxtaarticular cysts

DIFFERENTIAL DIAGNOSIS

Normal Aging/Degenerative Process

- Common in elderly (> 80 years of age) as incidental finding

Ligamentum Flavum Hypertrophy

- Low T2 signal enlargement of ligamentum flavum with variable canal stenosis

Osteomyelitis, Pyogenic

- Bone destruction with ill-defined margins
- Extension of inflammation into adjacent soft tissues

Metastases, Lytic Osseous

- Bone destruction with focal mass

Juxtaarticular Cyst

- Low T2 signal rim with central hyperintensity centered within ligamentum flavum or posterior epidural space

Tumoral Calcinosis

- Mass-like lobulated lesions with low T2 signal, centered on posterior elements

PATHOLOGY

Staging, Grading, & Classification

- Bursal index (plain radiographs)
  - Percentage figure found by dividing summed heights of individual interspinous spaces L1-L2 through L4-L5 by total height of spine from top of L1 spinous process to bottom of L5 spinous process
    - 6% in most "crowded" spines to 22% for most "spread-out" spines
    - The larger the number, lower the number of interspinous bursa found at autopsy

Gross Pathologic & Surgical Features

- Insertion tendinopathy
- Bony sclerosis of adjacent spinous process surfaces
- Interspinous region degenerates with formation of adventitious bursa with creation of synovial articulation

- Propagation of bursa to dorsal epidural space intraspinal juxtaarticular cysts
  - May cause central stenosis and claudication

Microscopic Features

- Interspinous cavities bordered by highly vascular eburnated bone with marked sclerosis
- Bursa shows areas of cellular lining
  - Acellular regions with fibrin deposition
- Villous processes with blood vessels
- Bursal cavity covered with partially calcified fibrocartilage
  - Occasional conversion to hyaline cartilage
- More severe cases show bone cysts, which contain fibrocartilage, adjacent new bone formation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - "Kissing" spinous processes a very common finding in elderly, often incidental
    - Increases with increasing age; found in 80% of subjects > 80 years of age
  - Controversial as to whether real source of back pain
    - Associated with many other aspects of spinal degenerative disc disease, confounding analysis
  - Classic pattern is back pain when standing erect with relief by flexion

Natural History & Prognosis

- Interspinous bursitis appearance seen in 8% of subjects undergoing MR of lumbar spine for back or leg pain
- Associations with Baastrup
  - Increasing age
  - Central canal stenosis
  - Ligamentum flavum hypertrophy
  - Disc degeneration with disc bulge
  - Anterolisthesis
  - Facet degenerative change with facet effusions

Treatment

- Conservative therapy with NSAIDs
- Direct steroid injection into bursa
- Resection of offending spinous processes
  - Not universally successful in relieving back pain
- X-Stop or other interspinous spacing device for treatment of canal stenosis

DIAGNOSTIC CHECKLIST

Consider

- Flexion extension plain films to define apposition of spinous processes with extension and to define segmental instability

Image Interpretation Pearls

- Lumbar interspinous ligament T2 hyperintensity reported as **Baastrup sign** since clinical source of back pain unknown

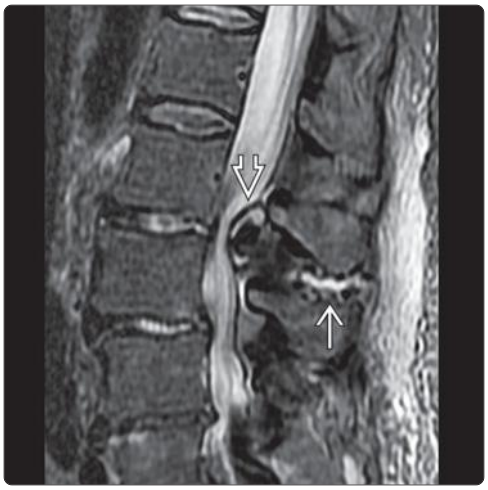
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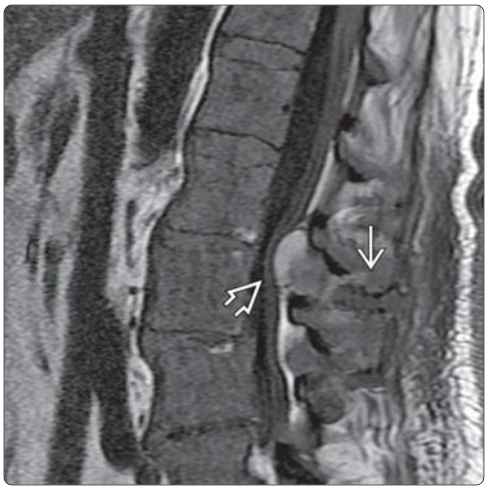
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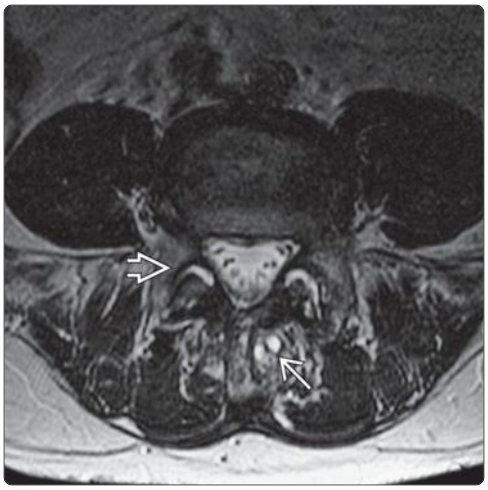


(Left) *Sagittal T1WI MR shows irregularity of the apposed margins of the L4 and L5 spinous processes, with ligamentum flavum and interspinous process hypertrophy, in a classic pattern.* **(Right)** *Sagittal T2WI MR shows cystic degenerative nature of Baastrup as focal high signal along spinous processes in a classic pattern. There is marked ligamentum flavum hypertrophy. Severe degenerative facet disease is also present in this patient.*





(Left) *Sagittal STIR MR shows Baastrup disease as irregular hyperintensity at the L2-L3 interspinous ligament level. There is an associated protein containing a synovial (juxtaarticular) cyst with central low signal within the dorsal epidural space, causing severe canal stenosis.* **(Right)** *Sagittal T1WI MR shows Baastrup disease as ill-defined prominence of the interspinous ligament and adjacent spinous process cortical irregularity. Note the associated protein-containing epidural cyst.*





(Left) *Axial T2WI MR shows the cystic degenerative nature of Baastrup as focal high signal along spinous processes and within the adjacent soft tissues. Severe degenerative facet disease is also present with facet effusions bilaterally.* **(Right)** *Sagittal STIR MR shows bursa formation of the interspinous ligament at L4-L5 level seen as linear hyperintensity. No adjacent marrow signal abnormality is seen. Note the grade II spondylolisthesis at L4-L5.*

Bertolotti Syndrome

KEY FACTS

TERMINOLOGY

- Back pain related to unilateral or bilateral enlargement of transverse process of most caudal lumbar vertebra, which may articulate or fuse with sacrum or ilium

IMAGING

- Partial sacralization of caudal segment of lumbar body on plain films, CT, or MR

TOP DIFFERENTIAL DIAGNOSES

- Normal variant
- Osteochondroma
- Post-traumatic ossification
- SI joint degenerative change
- Facet joint arthropathy

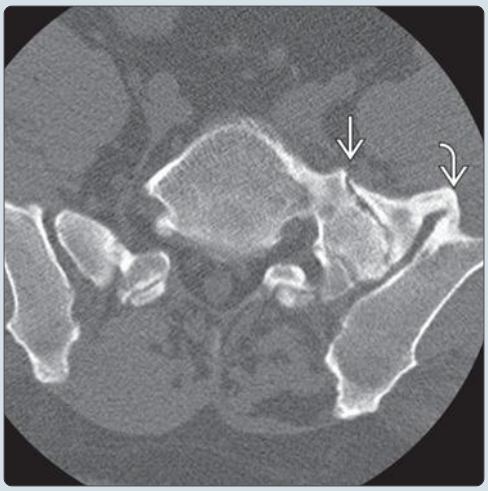
PATHOLOGY

- 4-6% of normal patients show transitional segment
  - Hypothesized mechanism is reduced and asymmetrical motion at lumbosacral junction
- Type I: Unilateral (Ia) or bilateral (Ib) dysplastic TP measuring at least 19 mm craniocaudal
- Type II: Unilateral (IIa) or bilateral (IIb) lumbarization/sacralization with enlarged TP with a diarthrodial joint with sacrum
- Type III: Unilateral (IIIa) or bilateral (IIIb) lumbarization/sacralization with complete osseous fusion of TP to sacrum
- Type IV: Unilateral type II transition with type III on contralateral side

CLINICAL ISSUES

- Causal relationship to low back pain controversial
- Conservative treatment with NSAIDs, physical therapy
- Steroid and local anesthetic infiltration into anomalous lumbosacral articulation
- Resection of accessory joint
- Posterolateral fusion of transitional segment

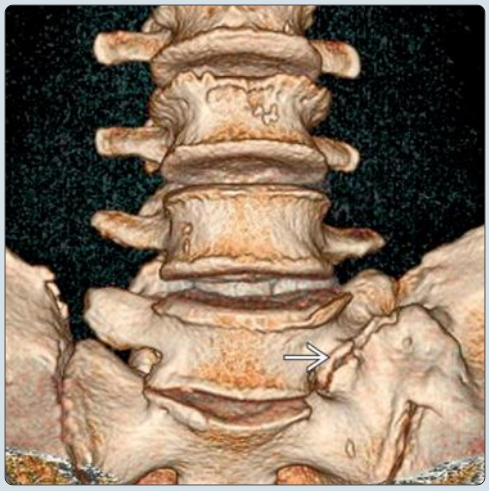
(Left) *Axial NECT shows large bridging pseudoarticulation ➡ of the caudal lumbar segment with the sacrum and ilium ➡ on the left, contrasted with the normal right-sided anatomy.* **(Right)** *Coronal NECT shows the large neoarticulation of the caudal lumbar body with the sacrum, with articular irregularity and vacuum phenomenon ➡.*





(Left) *Axial T2WI MR shows the transitional anatomy at the caudal lumbar segment with partial sacralization on the left ➡.* **(Right)** *Coronal CT reconstruction shows the anomalous pseudoarticulation ➡ at the caudal lumbar level on the left between the body, sacrum, and ilium.*





TERMINOLOGY

Definitions

- Back pain related to unilateral or bilateral enlargement of transverse process (TP) of most caudal lumbar vertebra, which may articulate or fuse with sacrum or ilium

IMAGING

General Features

- Best diagnostic clue
  - Partial sacralization of caudal segment of lumbar body on plain films, CT, or MR
- Location
  - Caudal segment of lumbar spine
- Size
  - Variable
- Morphology
  - Fusion of caudal segment to sacrum or ilium, or neoarticulation

CT Findings

- NECT
  - Transitional segment with either fusion or anomalous articulation of caudal segment with sacrum/ilium

MR Findings

- T1WI
  - Transitional caudal lumbar segment anatomy
- T2WI
  - T2 hyperintensity in joint and adjacent marrow not a prominent feature in reported cases

Nuclear Medicine Findings

- Bone scan
  - Syndrome has been described with both positive and negative bone scans at anomalous junction

DIFFERENTIAL DIAGNOSIS

Normal Variant

- Lumbar transitional anatomy common

Osteochondroma

- Cauliflower-like excrescence arising from cortex with cartilage cap

Post-Traumatic Ossification

- Irregular calcification within soft tissues

SI Joint Degenerative Change

- Degenerative osteoarthritis with bony eburnation and osteophyte formation

Facet Joint Arthropathy

- Degenerative disease with facet hypertrophy, eburnation, joint effusion

PATHOLOGY

General Features

- Bertolotti 1st described mechanical low back pain with transitional lumbar body in 1917
  - 7% of patients with low back pain show transitional segment
  - 4-6% of normal patients show transitional segment
- Hypothesized mechanism is reduced and asymmetrical motion at lumbosacral junction
  - Early degenerative change in neoarticulation
  - Early degenerative change in normal contralateral facet
  - Accelerated degenerative change directly superior to transitional segment
    - Disc at transitional level typically small and fibrotic and rarely source of disc herniation

Staging, Grading, & Classification

- Lumbosacral transitional vertebrae defined by Castellvi (1984)
  - Type I: Unilateral (Ia) or bilateral (Ib) dysplastic TP measuring at least 19 mm craniocaudal
  - Type II: Unilateral (IIa) or bilateral (IIb) lumbarization/sacralization with enlarged TP with a diarthrodial joint with sacrum
  - Type III: Unilateral (IIIa) or bilateral (IIIb) lumbarization/sacralization with complete osseous fusion of TP to sacrum
  - Type IV: Unilateral type II transition with type III on contralateral side

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Causal relationship to low back pain is controversial
    - Mechanical low back pain typical
    - Clinically may manifest as SI joint pain or facet pain

Treatment

- Nonsurgical
  - Conservative treatment with NSAIDs, physical therapy
  - Steroid and local anesthetic infiltration into anomalous lumbosacral articulation
- Surgical
  - Resection of accessory joint, posterolateral fusion

DIAGNOSTIC CHECKLIST

Consider

- Clinical diagnosis of exclusion, given prevalence of transitional anatomy in normal population

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Schmorl Node

KEY FACTS

TERMINOLOGY

- Definition: Node within vertebral body due to vertical disc extension (intravertebral disc herniation) through vertebral endplate

IMAGING

- Focal invagination of endplate by disc material surrounded by sclerotic (old) or edematous (acute) bone
- Plain films
  - Contour defect within endplate, extending from disc space into vertebral body spongiosa with well-corticated margins
- CT
  - Island of low density surrounded by condensed bone on axial slice through vertebral body
- MR
  - Focal defect in endplate filled by disc ± adjacent marrow edema, fatty marrow conversion

TOP DIFFERENTIAL DIAGNOSES

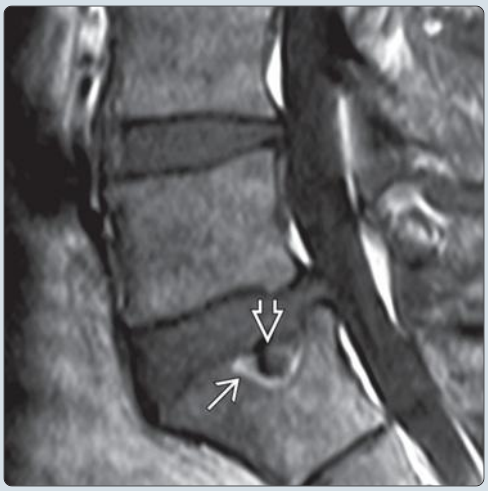
- Acute compression fracture
- Degenerative endplate change
- Discitis
  - Both endplates show defect
- Limbus vertebrae
  - Seen at vertebral body corners
- Bone island
  - Sclerotic nodule
- Focal metastasis
  - No contiguity with parent disc

CLINICAL ISSUES

- Seen in up to 75% of all normal spines
- Conservative management

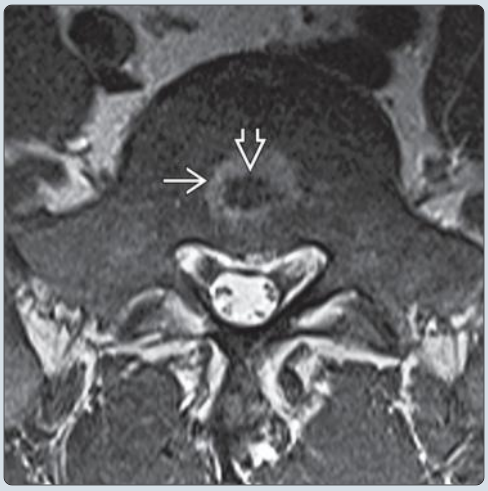
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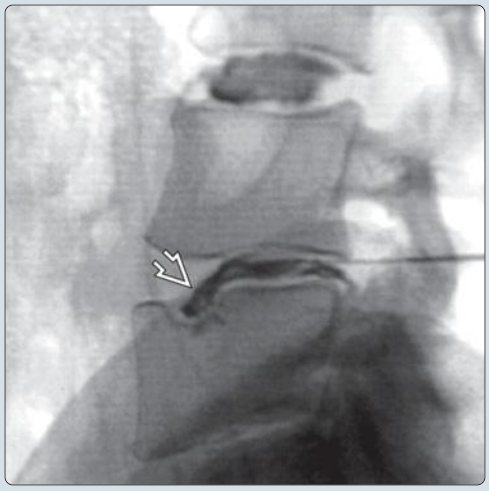
- Schmorl node (SN) is always contiguous with parent disc





(Left) *Sagittal T1WI MR shows a well-defined focus of low signal ⇨ involving the superior endplate of S1, due to endplate herniation. There is a thin rind of fatty marrow conversion adjacent to the Schmorl node (SN) ⇨.* **(Right)** *Sagittal T2WI MR (same patient) demonstrates the typical pattern of fatty marrow ⇨ outlining chronic endplate herniation (SN) ⇨.*





(Left) *Axial T2WI MR (same patient) shows a well-defined focus of low signal ⇨ in the superior S1 endplate representing a SN. There is a thin concentric rind of fatty marrow conversion ⇨ surrounding the SN.* **(Right)** *Lateral fluoroscopy image (different patient) during discography reveals a superior endplate SN ⇨. Contrast injected into the contiguous intervertebral disc fills the herniated disc within the vertebral SN defect.*

TERMINOLOGY

Abbreviations

- Schmorl node (SN)

Synonyms

- Intravertebral disc herniation

Definitions

- Node within vertebral body due to vertical disc extension through weakened vertebral body endplate

IMAGING

General Features

- Best diagnostic clue
  - Focal invagination of endplate by disc material surrounded by either sclerotic (old) or edematous (acute) bone
- Location
  - T-8 to L1 region most common
- Size
  - Varies from several millimeters to "giant"
- Morphology
  - Typically upwardly round or cone-shaped defect contiguous with disc space

Radiographic Findings

- Radiography
  - Contour defect within endplate, extending from disc space into vertebral body spongiosa with well-corticated margins

CT Findings

- NECT
  - Island of low density surrounded by condensed bone on axial slice through vertebral body
  - Sagittal reformation shows endplate defect contiguous with disc space, capped by sclerotic bone
- Bone CT
  - Same as NECT
  - SN may calcify

MR Findings

- T1WI
  - Focal defect in endplate filled by disc
  - Low signal in adjacent marrow if acute
- T2WI
  - Defect in endplate filled by disc
  - Normal marrow signal if chronic
  - High signal in adjacent marrow if acute
- STIR
  - Edema in adjacent vertebral body if acute
- T1WI C+
  - Localized marginal contrast enhancement in subacute stage
  - Diffuse marrow enhancement in acute stage
  - May show central enhancement of cartilaginous component associated with disc degeneration

Nuclear Medicine Findings

- Bone scan
  - Increased uptake in acute cases

Imaging Recommendations

- Best imaging tool
  - MR with contrast will show endplate lesion and exclude concurrent pathology
- Analyze contiguity with parent disc on all sequences

DIFFERENTIAL DIAGNOSIS

Acute Compression Fracture

- Simulates diffuse edema of acute SN; may in fact predispose to its ultimate formation
- Lacks imploded disc nodule within abnormal marrow

Degenerative Endplate Change

- Seen as reactive change to disc degeneration; typically both adjacent vertebral bodies affected
- Represent granulation tissue and edema incited by degenerating disc
- Edema replaced by fat on follow-up studies
- No focal endplate defect

Discitis

- Both endplates show defect
- Disc signal diffusely abnormal and enhances after contrast

Limbus Vertebrae

- Seen only at vertebral body corners
- Truncated anterior margin of vertebral body
- Bone fragments anterior to defect

Bone Island

- Sclerotic nodule
- No endplate defect

Focal Fatty Marrow

- Hyperintense on T1WI

Focal Metastasis

- Does not show contiguity with parent disc or its signal intensity

PATHOLOGY

General Features

- Etiology
  - Developmental, degenerative, traumatic, and disease influences
  - Typically repetitive stress of gravity on immature endplate
  - Acute axial traumatic load can lead to SN formation with focal back pain
  - Osteoporosis, neoplasm, and infection can weaken endplate
- Genetics
  - Twin studies show SN common in middle-aged women (30%) and strongly genetically determined
    - Associated with lumbar degenerative change
    - Not an independent risk factor for back pain
    - Heritability is > 70%
- Cartilaginous disc tissue with degenerative or inflammatory changes
  - Pathologic staging mirrors that of focal endplate fracture

- Typical SN is healed focal endplate fracture
- Embryology/anatomy
  - Annulus actually biomechanically more resistant to mechanical failure than endplate in young individuals
  - Focal weakness of endplate predisposes to SN formation
  - Associated with endplate weakening of Scheuermann disease

Staging, Grading, & Classification

- Edema of vertebral body next to endplate with acute trauma, pain, no endplate defect on initial MR
- Subsequent formation of chronic and asymptomatic SN on follow-up MR

Gross Pathologic & Surgical Features

- Identical to endplate fracture

Microscopic Features

- Fibrocartilaginous tissue surrounded by marrow with sclerotic cancellous bone or inflammatory changes
- Osteonecrosis may also be present beneath cartilaginous endplate
  - Increased reactive woven bone with thickened trabeculae and prominent osteoclasts and osteoblasts

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sudden onset, localized, nonradiating pain and tenderness in acute cases
  - Most cases found incidentally as chronic, "burned out" lesions
- Clinical profile
  - Teenager involved in axial-loading sports
  - Acute onset of localizing pain

Demographics

- Age
  - Adolescents and young adults
- Gender
  - M > F, up to 9:1 ratio
- Epidemiology
  - Seen in up to 75% of all normal spines
  - Most acute cases occur in individuals 11-30 years old
  - Can occur with single traumatic episode

Natural History & Prognosis

- Self-limited
- Good prognosis, unless systemic osteoporosis leads to recurrent compression fractures

Treatment

- Observational, with pain management in symptomatic cases

DIAGNOSTIC CHECKLIST

Consider

- Follow-up MR in cases of unexplained vertebral body edema and localized pain

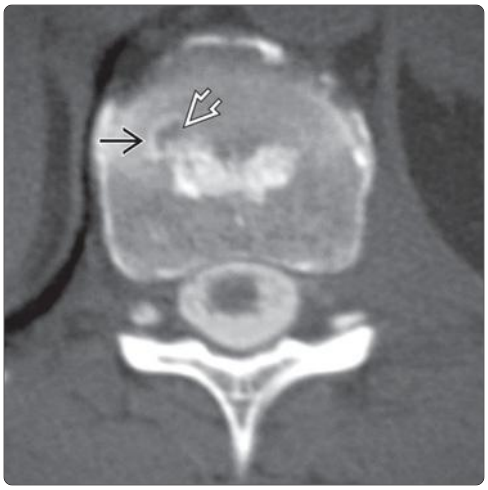
Image Interpretation Pearls

- Always contiguous with parent disc

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(Left) *Sagittal bone CT shows multiple SN of varying sizes. There is marginal sclerosis adjacent to each node, representing reparative bone changes. This patient does not meet criteria for Scheuermann disease because there are not 3 adjacent levels with at least 5° of anterior wedging.* **(Right)** *Axial CT myelogram reveals sclerotic rim of bone surrounding low-density material (SN) within the vertebral body, just above the interspace.*





(Left) *Sagittal T1WI MR shows a small inferior endplate SN. There is no marrow edema or fatty conversion surrounding this nonacute SN.* **(Right)** *Sagittal STIR MR confirms absence of marrow edema surrounding an asymptomatic SN incidentally discovered on MR imaging performed for other reasons. The remaining intervertebral disc spaces are normal.*





(Left) *Sagittal T1WI MR demonstrates a focal undersurface endplate defect filled by disc material representing a SN. There is prominent low signal intensity within adjacent marrow representing a recent injury in this patient with focal back pain.* **(Right)** *Sagittal T1WI C+ MR with fat saturation (same patient) depicts diffuse marrow enhancement surrounding the inferior endplate SN suggesting a subacute time point.*

Scheuermann Disease

KEY FACTS

TERMINOLOGY

- Juvenile kyphosis, Scheuermann kyphosis
- Kyphosis secondary to multiple Schmorl nodes → vertebral body wedging

IMAGING

- Wedged-shaped thoracic vertebrae with irregular endplates
  - ≥ 3 contiguous vertebrae, each showing ≥ 5° of kyphosis
  - Undulation of endplates secondary to extensive disc invaginations
  - Disc spaces narrowed, with greatest narrowing anteriorly
  - Well-defined Schmorl nodes

TOP DIFFERENTIAL DIAGNOSES

- Postural kyphosis
- Wedge compression fractures
- Congenital kyphosis
- Tuberculosis
- Osteogenesis imperfecta
- Neuromuscular disease

PATHOLOGY

- Disc extrusions through weakened regions of vertebral endplates
- Weightlifting, gymnastics, and other spine-loading sports may contribute

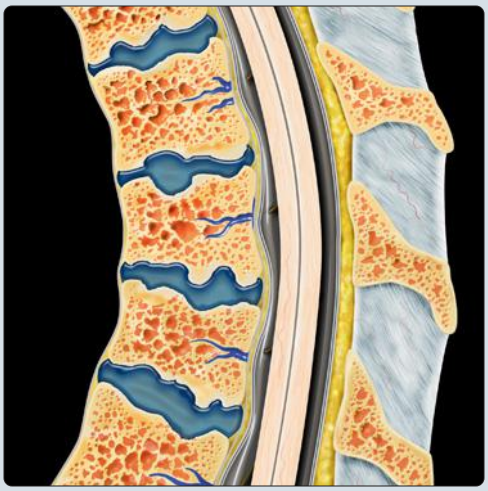
CLINICAL ISSUES

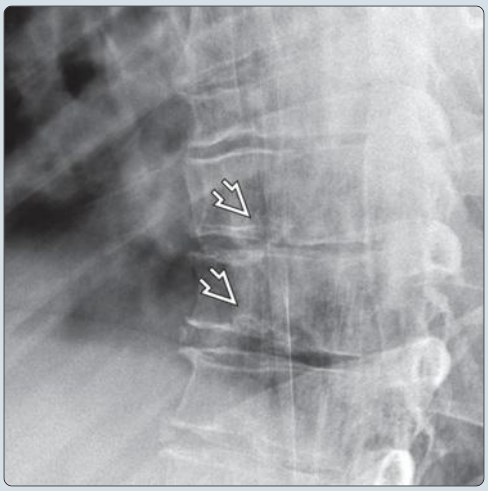
- Thoracic spine pain worsened by activity
- Kyphosis develops in adolescence, may present later in life
- Peak incidence: 13-17 years
- Initial treatment includes observation, bracing
- Surgical treatment for > 75° kyphosis in skeletally immature person

DIAGNOSTIC CHECKLIST

- Schmorl nodes without anterior wedging are not indicative of Scheuermann disease

(Left) *Sagittal graphic demonstrates anterior vertebral wedging and herniation of disc material through the vertebral endplates, creating focal subcortical bone defects in conjunction with thoracic kyphotic deformity. Undulation of endplates reflects sequelae of the osseous reparative process.* **(Right)** *Lateral radiography reveals anterior wedging deformity and undulation of vertebral endplates with multiple Schmorl nodes at each level ⇨.*





(Left) *Sagittal bone CT shows loss of anterior vertebral height and endplate irregularity involving multiple thoracic discs. There are multiple (> 3) contiguous levels of endplate irregularities (Schmorl node) and increased thoracic kyphosis.* **(Right)** *Sagittal T2WI MR (same patient) confirms loss of disc height and signal intensity at multiple thoracic levels. The thoracic spinal cord is normal, and there are no associated thoracic disc herniations into the spinal canal.*





TERMINOLOGY

Synonyms

- Juvenile kyphosis, Scheuermann kyphosis

Definitions

- Kyphosis secondary to multiple Schmorl nodes → vertebral body wedging
- Schmorl node: Invagination of disc material through vertebral body endplate

IMAGING

General Features

- Best diagnostic clue
  - ≥ 3 wedged thoracic vertebrae with irregular endplates
- Location
  - 75% thoracic
  - 20-25% thoracolumbar
  - < 5% lumbar only
  - Rarely cervical
- Size
  - Normal kyphosis of thoracic spine increases with age
  - Kyphosis > 40° considered abnormal

Radiographic Findings

- Radiography
  - ≥ 3 contiguous vertebrae, each showing ≥ 5° of kyphosis
  - Undulation of endplates secondary to extensive disc invaginations
  - Disc spaces narrowed, with greatest narrowing anteriorly
  - Well-defined Schmorl nodes
  - Thoracic spine: Measure kyphosis from T3 to T12
  - Thoracolumbar or lumbar involvement
    - Measure sagittal plane deformity from 1 vertebra above affected vertebrae to 1 vertebra below
    - This method of measuring gives a better estimate of functional kyphosis
    - Loss of normal lumbar lordosis functionally significant
  - ± limbus vertebrae
  - 15% have scoliosis as well as kyphosis
    - Hyperextension lateral radiograph to assess flexibility of kyphosis

CT Findings

- Bone CT
  - Endplate abnormalities more apparent than on radiography

MR Findings

- T1WI
  - Schmorl nodes, disc herniation with low signal intensity
  - ± discogenic sclerosis
- T2WI
  - Disc degeneration seen in 50% of involved discs
  - Disc herniations
  - Schmorl nodes may be low or high signal intensity
    - ± bone marrow edema adjacent to Schmorl nodes

Nuclear Medicine Findings

- Bone scan
  - May be normal or show increased activity

Other Modality Findings

- Bone mineral density normal (DEXA, CT bone densitometry)

Imaging Recommendations

- Plain film for diagnosis
- MR to exclude disc herniations

DIFFERENTIAL DIAGNOSIS

Postural Kyphosis

- Vertebral endplates normal
- Deformity usually corrects in hyperextension unless longstanding

Wedge Compression Fractures

- May involve contiguous levels
- Anterior vertebral cortex often shows angular deformity
- MR shows fracture lines, bone marrow edema

Congenital Kyphosis

- Vertebral anomalies present

Tuberculosis

- Kyphosis often severe
- Endplate destruction ± vertebral fusion

Osteogenesis Imperfecta

- Platyspondyly
- Severe osteopenia

Neuromuscular Disease

- Persistence of infantile thoracolumbar kyphosis in nonambulatory patients
- Kyphoscoliosis

Ankylosing Spondylitis (AS)

- Fusion of vertebral bodies
- Abnormalities of sacroiliac joints

Spondyloepiphyseal Dysplasia Tarda (SED)

- Platyspondyly throughout spine
- Epiphyses abnormal

PATHOLOGY

General Features

- Etiology
  - Unknown
    - Weightlifting, gymnastics, and other spine-loading sports may contribute
  - Disc extrusions through weakened regions of vertebral endplates
    - Disc space loss
    - Limbus vertebrae
    - Schmorl node
  - Delayed growth in anterior portion of vertebrae causes wedging
- Genetics
  - Familial tendency
  - Danish twin registry showed major genetic contribution
- Associated abnormalities
  - Lordosis of cervical and lumbar segments

Gross Pathologic & Surgical Features

- Schmorl nodes invaginate into vertebral bodies through fissures in weakened growth plates
- Limbus vertebrae occur when disc material protrudes through growth plate of ring apophysis

Microscopic Features

- Vertebral body growth plates
  - Abnormal chondrocytes
  - Loose cartilage matrix
  - Diminished number or thickness of collagen fibers
  - Increased proteoglycan content
- Osteonecrosis, osteochondrosis not seen

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Kyphosis
  - Other signs/symptoms
    - Thoracic spine pain and tenderness worsened by activity
    - Neurologic symptoms from kyphosis or disc herniations
    - Fatigue

Demographics

- Age
  - Develops in adolescence, may present later in life
- Gender
  - Slight male predominance
- Epidemiology
  - Prevalence: 0.4-8%
  - Peak incidence: 13-17 years

Natural History & Prognosis

- Increases in magnitude during adolescent growth spurt
- Mild progression after growth is complete
- Severe deformity uncommon
- Kyphosis > 70° has poor functional result
- Premature disc degeneration
- Disc herniations related to degeneration, mechanical stress from spine deformity

Treatment

- Observation
  - Indications
    - Growth still remains
    - Kyphotic deformity < 50°
  - Elimination of specific strenuous activity
  - Analgesics
  - Spine exercises
  - Follow-up until growth plate fuses (~ age 25)
- Brace treatment
  - Indications
    - At least 1 year of growth remains
    - < 70° kyphosis
    - At least partial correction of kyphosis on hyperextension
    - Efficacious in skeletally immature patients
- Surgical treatment
  - Uncommonly felt to be necessary
  - Indications
    - > 75° kyphosis in skeletally immature person
    - > 60° kyphosis in mature person
    - Excessive pain
    - Neurologic deficit
  - Posterior instrumentation and fusion
  - Anterior and posterior fusion for more severe kyphosis

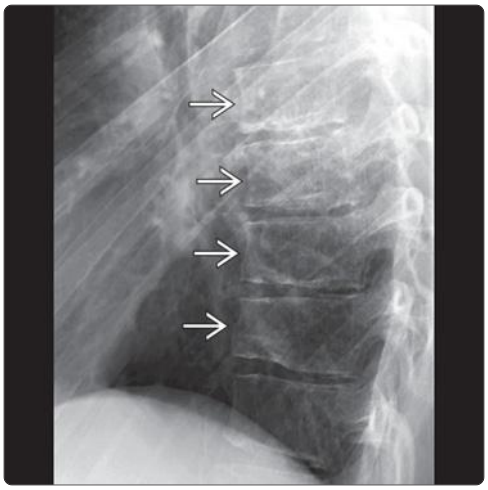
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Schmorl nodes without anterior wedging are not indicative of Scheuermann disease
- Radiography may show undulating endplates rather than discrete Schmorl nodes

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(Left) *Lateral radiograph reveals endplate irregularities and vertebral wedging. The vertebral endplate depressions have an undulating contour, unlike the angular depressions that occur in fractures.* **(Right)** *Sagittal bone CT shows multilevel endplate irregularities and vertebral wedging. Diagnostic criteria for Scheuermann disease include involvement of 3 contiguous levels with Schmorl nodes and at least 5° of wedging at each level, which were met in this case.*





(Left) *Sagittal T1WI MR (teen rodeo rider with back pain) demonstrates multilevel thoracic vertebral anterior wedging with endplate undulations, Schmorl nodes, and kyphosis characteristic of fairly severe Scheuermann disease.* **(Right)** *Sagittal STIR MR (same patient) confirms multilevel thoracic vertebral anterior wedging, endplate undulations, Schmorl nodes, and fairly severe kyphosis.*





(Left) *Sagittal T2WI MR illustrates lumbar variant of Scheuermann disease with diffuse loss of intervertebral disc signal, wedging of vertebral bodies, undulation of endplates, and discrete Schmorl nodes. Note the large disc extrusion.* **(Right)** *Sagittal T2WI MR depicts multiple level endplate irregularity with Schmorl nodes and disc degeneration throughout visualized spine. Multiple contiguous levels are identified. Type I degenerative endplate changes are present as well.*

Acquired Lumbar Central Stenosis

KEY FACTS

TERMINOLOGY

- Spinal canal narrowing in lumbar spine
  - Secondary to multifactorial degenerative changes, being a progressive, and dynamic, process

IMAGING

- Trefoil appearance of lumbar spinal canal on axial imaging
- Sagittal diameter of bony lumbar canal < 12 mm relative stenosis
- Sagittal diameter of bony lumbar canal < 10 mm absolute stenosis

TOP DIFFERENTIAL DIAGNOSES

- Disc herniation
- Metastatic disease
- Paget disease
- Epidural hemorrhage

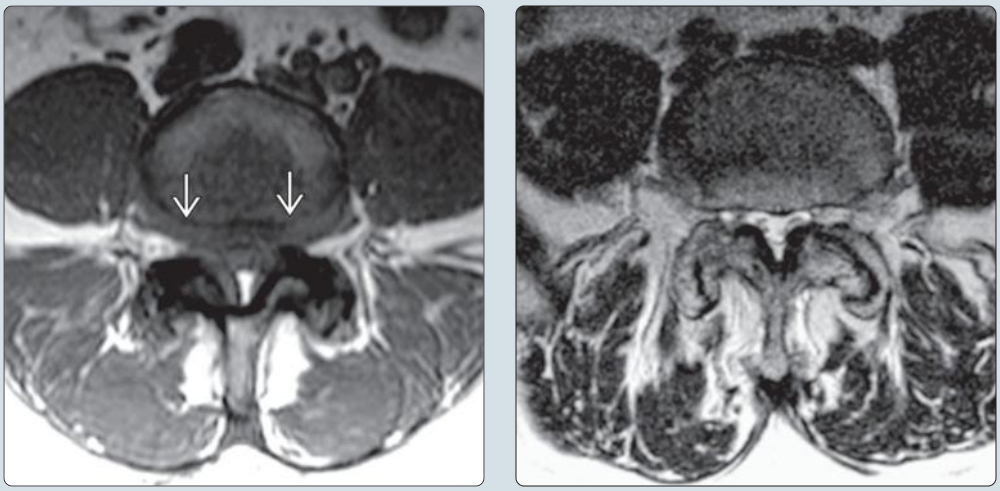
CLINICAL ISSUES

- Chronic low back pain
- Neurogenic claudication
  - Leg pain (80%)
  - Bilateral lower extremity pain, paresthesia, and weakness
  - Relief of pain by squatting or sitting (flexion) in 80%
- Bladder dysfunction and sexual difficulty (10%)
- Radicular pain (10%)
- Operative treatment includes surgical decompression or X-Stop interspinous implant-like devices
- Natural history: Majority of symptomatic patients stable over months to years (40-70%)
  - 1/3 improve with nonoperative treatment
  - 1/3 deteriorate

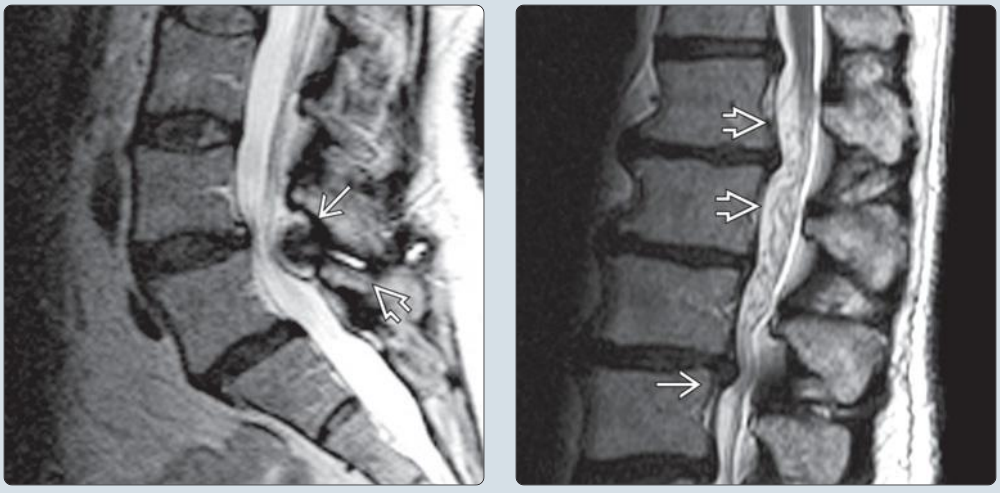
DIAGNOSTIC CHECKLIST

- Axial T2WI mandatory for stenosis identification

(Left) *Axial T1WI MR shows severe central canal stenosis due to a diffusely bulging anulus ⇨ anteriorly, and posterior facet hypertrophic degenerative arthropathy, which combine to compress the thecal sac.* **(Right)** *Axial T2WI MR demonstrates severe central canal stenosis with marked facet degenerative arthropathy. The thecal sac has assumed a trefoil appearance.*



(Left) *Sagittal T2WI MR shows severe central stenosis at L4-L5 due to marked posterior ligamentum flavum thickening ⇨. Hyperintensity within the interspinous ligament denotes degeneration ⇨, which is also called Baastrup disease or interspinous bursitis.* **(Right)** *Sagittal T2WI MR shows canal stenosis at L3-L4 with a bulging disc and ligamentous thickening ⇨. Note serpentine areas of low signal within the cephalad thecal sac due to redundant nerve roots ⇨.*



TERMINOLOGY

Synonyms

- Spondylosis, central canal stenosis, lumbar canal stenosis

Definitions

- Spinal canal narrowing in lumbar spine
- Secondary to multifactorial degenerative changes, being a progressive, and dynamic, process

IMAGING

General Features

- Best diagnostic clue
  - Trefoil appearance of lumbar spinal canal on axial imaging
- Location
  - Most common in lower lumbar spine where there is the most mobility (L4-L5)
- Size
  - Sagittal diameter of lumbar canal < 12 mm relative stenosis
  - Sagittal diameter of lumbar canal < 10 mm absolute stenosis
- Morphology
  - Obliterated perineural fat in lumbar neural foramina on sagittal imaging
  - Narrowed lumbar lateral recess on axial imaging

Radiographic Findings

- Radiography
  - Disc space narrowing, osteophytes
  - Facet osteoarthritis, spondylosis, spondylolisthesis
  - Interpedicular distance narrowing if combined with congenital stenosis

CT Findings

- NECT
  - Direct visualization of compressing disc disease (bulge), facet arthropathy, and ligamentous thickening
  - Trefoil pattern of canal, with compression of thecal sac
  - Reformats allow visualization of foraminal stenosis
    - Rostrocaudal subluxation of facets associated with loss of disc space height
    - Osteophyte impinging upon neural foramen

MR Findings

- T1WI
  - MR equivalent to CT myelogram in diagnosing lumbar spinal stenosis but provides additional information about spinal cord
  - Degenerative disc disease with variable degree of herniation
  - Trefoil appearance of spinal canal on axial imaging
  - Thickened ligamentum flavum
  - Vertebral endplate osteophytes
  - Facet joint hypertrophy
  - Obliterated perineural fat in neural foramina on sagittal imaging
  - Short pedicles
- T2WI
  - Trefoil appearance of spinal canal on axial imaging
  - Hourglass appearance of central canal on sagittal T2WI
  - Elongated and redundant nerve roots above and below level of stenosis
  - Narrowed lateral recess on axial imaging
- T1WI C+
  - Enhancing and crowded nerve roots

Nonvascular Interventions

- Myelography
  - 93% accurate for spinal stenosis
  - Hourglass constriction at single or multiple levels
  - Patient positioning in extension accentuates stenosis
    - Dynamic changes: If block at stenotic level, forward flexion often allows contrast past stenotic level

Imaging Recommendations

- Best imaging tool
  - MR
  - Conventional T1W and T2W sagittal and axial images with 4-5 mm slice thickness

DIFFERENTIAL DIAGNOSIS

Infection

- Disc space and vertebral body hyperintensity on T2WI, with endplate irregularity
- Enhancement of epidural phlegmon, peripheral enhancement of abscess

Disc Herniation

- Focal lesion arising from intervertebral disc

Metastatic Disease

- Diffuse or focal hypointensity on T1WI, ventral epidural soft tissue from contiguous extension

Paget Disease

- Expansion of vertebral body
- Patchy hyperintensity of vertebral body on T1WI

Epidural Hemorrhage

- Variable signal intensity depending on evolving hemoglobin
- Acute onset of symptoms

PATHOLOGY

General Features

- Etiology
  - Degenerative changes involving discs, vertebral endplates, facet joints, and ligamentum flavum
  - Congenitally short pedicles often contribute to acquired spinal stenosis
- Genetics
  - None for acquired stenosis
- Associated abnormalities
  - Congenitally short pedicles often present
- Age-related degenerative disease involving disc and facets

Staging, Grading, & Classification

- Normal lumbar canal
  - > 12 mm AP diameter
  - Cross section area > 70 mm square
- 50% reduction in cross section area of bony canal motor and sensory deficits occur (animal model)

Gross Pathologic & Surgical Features

- Disc degeneration
  - Loss of nuclear definition, fibrosis, and fissuring
  - Osteophyte formation
  - Anular disruption
  - Loss of facet joint space and cartilage, subchondral sclerosis, osteophytosis

Microscopic Features

- Fissuring of disc, loss of cartilaginous endplate, granulation tissue, bony sclerosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Symptoms include
    - Chronic low back pain
    - Bilateral lower extremity pain, paresthesia, and weakness
    - Exacerbation by prolonged standing and walking
    - Relief of pain by squatting or sitting (flexion) in 80%
  - Neurogenic claudication
    - Leg pain (80%)
    - Numbness, tingling (60%)
    - Cramping, weakness (50%)
  - Other signs/symptoms
    - Bladder dysfunction and sexual difficulty (10%)
    - Radicular pain (10%)
    - Negative straight leg raise
  - Degree of spinal stenosis on imaging may not correlate with symptomatology
- Clinical profile
  - Insidious onset of low back pain, paresthesias, difficulty walking

Demographics

- Age
  - > 50 years
- Gender
  - M = F
  - Associated degenerative spondylolisthesis 4x more common in women
- Epidemiology
  - 5th decade and later

Natural History & Prognosis

- Majority of symptomatic patients stable over months to years (40-70%)
- 1/3 improve with nonoperative treatment
- 1/3 deteriorate
- Patients with history of back pain < 5 years and leg pain < 6 months have success with nonoperative treatment

Treatment

- Nonoperative
  - Analgesic medications: Salicylates, NSAIDs
  - Antidepressants (manage chronic pain and associated depression)
  - Exercise
  - Injection therapy (epidural steroids)
  - Other: TENS, thermal therapy
- Operative
  - Operative treatment primarily surgical decompression with laminectomies, fusion
    - > 70% complete relief of preoperative symptoms
  - Other surgery: X-Stop interspinous implant device, expansive laminoplasty, or fenestration procedure
    - X-Stop patients showed improvement at 12 months in 54%
  - Postoperative complications
    - Mortality 0.09%; increases to 0.6% in patients more than 75 years old
    - Radicular deficit (5%)
    - CSF fistula/pseudomeningocele (5%)
    - Infection (0.5-8%)

DIAGNOSTIC CHECKLIST

Consider

- MR myelography using bright CSF echo planar sequence or thin-section fast T2-weighted images

Image Interpretation Pearls

- Border of thecal sac and ligamentum flavum indistinct on axial T1WI
- Axial T2WI mandatory for stenosis identification

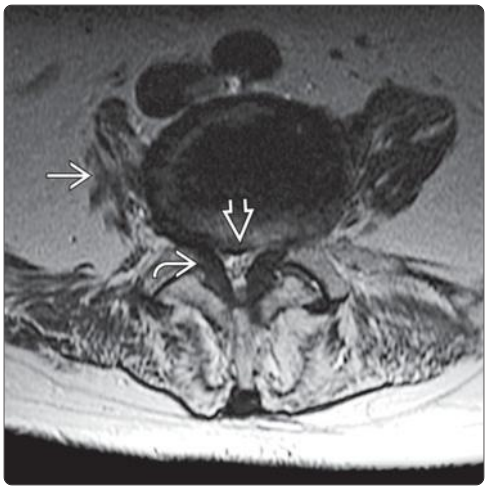
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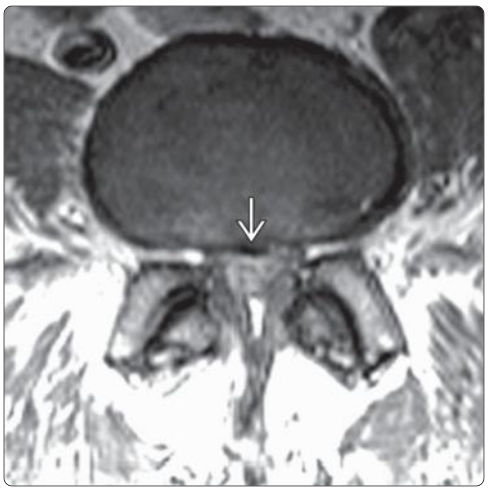
(Left) *Sagittal T1WI MR shows severe disc degeneration at L1-L2 through L4-L5 with marked loss of disc space height and vacuum discs. There is fusion of the L5-S1 disc level. Note severe central stenosis at each disc level due to ventral disc bulge/osteophyte and posterior ligamentous hypertrophy.* **(Right)** *Sagittal T2WI MR shows severe central stenosis at each disc level due to ventral disc bulge/osteophyte and posterior ligamentous hypertrophy seen as low signal extradural disease.*





(Left) *Axial T2WI MR shows severe central stenosis due to ventral disc bulge and marked facet and ligamentous hypertrophic degenerative arthropathy. Note right psoas muscle atrophy.* **(Right)** *Sagittal T2WI MR shows severe disc degeneration at L2-L3 through L5-S1 with loss of disc height and severe canal stenosis at multiple levels. There is intradural nerve root redundancy from the multilevel central stenosis.*





(Left) *Sagittal T1WI C+ MR shows focal enhancement of the roots at the stenotic L4-L5 level. This pattern of focal enhancement at the stenotic level should not be confused with tumor seeding.* **(Right)** *Axial T1WI C+ MR shows central canal stenosis with facet arthropathy and ligamentous thickening. There is diffuse enhancement of the intrathecal roots.*

Congenital Spinal Stenosis

KEY FACTS

TERMINOLOGY

- Reduced AP canal diameter secondary to short, squat pedicles and laterally directed laminae

IMAGING

- Central canal diameter is smaller than normal
  - Cervical spine: Absolute AP diameter < 14 mm
  - Spinal stenosis present if AP canal diameter at L1 < 20 mm, L2 < 19 mm, L3 < 19 mm, L4 < 17 mm, L5 < 16 mm, S1 < 16 mm
    - Lumbar spine: Critical stenosis at L4 < 14 mm, L5 < 14 mm, S1 < 12 mm
- Short, thick pedicles
- Trefoil-shaped lateral recesses
- Laterally directed laminae

TOP DIFFERENTIAL DIAGNOSES

- Acquired spinal stenosis
- Inherited spinal stenosis
  - Achondroplasia
  - Mucopolysaccharidoses

PATHOLOGY

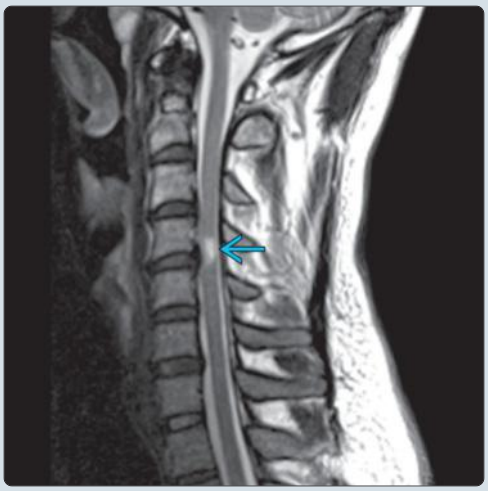
- Torg ratio (AP canal diameter/AP vertebral body diameter) < 0.8
- Idiopathic

CLINICAL ISSUES

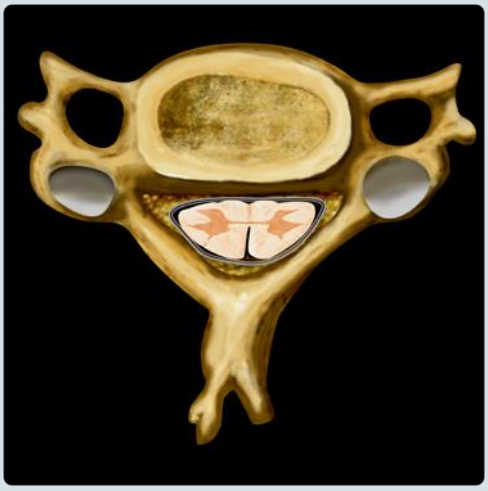
- Symptomatic cervical or lumbar stenosis symptoms at a younger age than typical of degenerative stenosis
  - These patients typically lack complicating medical problems (diabetes or vascular insufficiency)
- Athletes present with temporary neurological deficit following physical contact that subsequently resolves
- Lumbar: Decompressive laminectomy, posterior foraminotomy at involved levels
- Cervical: Posterior cervical laminectomy or laminoplasty

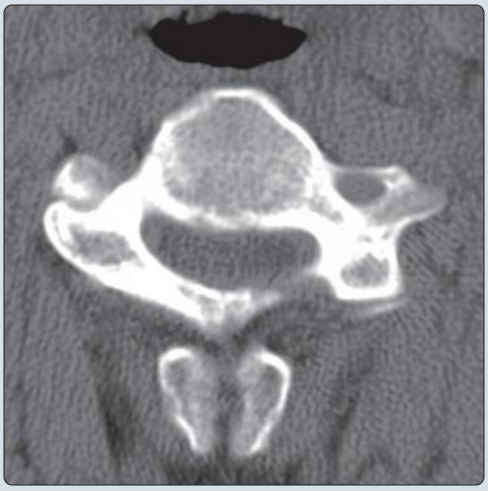
(Left) *Sagittal graphic shows marked congenital anteroposterior narrowing of the central spinal canal.* **(Right)** *Sagittal T2WI MR reveals moderate congenital AP canal narrowing exacerbated by C4-C5 disc herniation. The protrusion produces spinal cord T2 hyperintensity ⇨ corresponding to clinical myelopathy.*





(Left) *Axial graphic depicts congenital cervical AP spinal narrowing with generalized narrowing of the subarachnoid space. The pedicles are thick and laterally directed with resultant flattening of laminae.* **(Right)** *Axial bone CT demonstrates marked AP canal reduction attributable to developmentally short pedicles and laterally oriented laminae.*





TERMINOLOGY

Synonyms

- Short pedicle syndrome, congenital short pedicles, developmental spinal stenosis

Definitions

- Reduced AP canal diameter secondary to short, squat pedicles and laterally directed laminae

IMAGING

General Features

- Best diagnostic clue
  - Short, thick pedicles producing narrowed anteroposterior spinal canal diameter
- Location
  - Lumbar > cervical > thoracic spine
- Size
  - Central bony canal diameter is smaller than normal
    - Cervical spine: Absolute AP diameter < 14 mm
    - Lumbar spine: Critical stenosis at L4 < 14 mm, L5 < 14 mm, S1 < 12 mm
- Morphology
  - Short, thick pedicles
  - Trefoil-shaped lateral recesses
  - Laterally directed laminae

Radiographic Findings

- Radiography
  - Shortened AP distance between posterior vertebral body and spinolaminar line
    - ± superimposed degenerative disc, facet disease
  - Lateral radiograph normally shows articular pillar ending before spinolaminar line
    - Imaging pearl: If articular pillar takes up entire AP canal dimension on lateral radiograph, central canal stenosis is present

CT Findings

- Bone CT
  - Short, thick pedicles, trefoil-shaped lateral recesses, and laterally directed laminae
    - ± acquired disc, facet degenerative changes
  - Sagittal plane useful to survey extent of congenital narrowing
  - Axial plane best demonstrates reduced AP canal diameter, short, thick pedicles

MR Findings

- T1WI
  - Short AP diameter ± superimposed acquired facet, disc degenerative changes
  - ± hypointense facet, vertebral body marrow changes indicative of superimposed degenerative changes
- T2WI
  - Similar osseous findings to T1WI MR
  - ± cord compression
- T2* GRE
  - Best depicts osseous structures
  - Caveat: Overestimates true degree of canal narrowing secondary to susceptibility "blooming" artifact

Nonvascular Interventions

- Myelography
  - Confirms shortened AP dimension, clarifies severity of neural compression

Imaging Recommendations

- CT with sagittal and coronal reformats to evaluate osseous structures
  - Axial plane best for degree of narrowing and neural compromise
- MR to assess degree/presence of spinal cord, dural sac compression
  - Also demonstrates osseous anatomy well; permits complete imaging assessment with single imaging study
  - Sagittal MR best demonstrates AP canal narrowing, assesses for cord/cauda equina compression
  - Axial MR confirms pedicle configuration, assesses severity of canal narrowing

DIFFERENTIAL DIAGNOSIS

Acquired Spinal Stenosis

- Normal pedicle length
- ± subluxation
- ± spondylolysis
- ± disc degenerative changes
- ± facet degenerative changes

Inherited Spinal Stenosis

- Genetic predisposition; most common
  - Achondroplasia
  - Mucopolysaccharidoses
    - Frequently associated with characteristic brain, visceral, &/or extremity abnormalities enabling specific diagnosis

PATHOLOGY

General Features

- Etiology
  - Idiopathic
- Associated abnormalities
  - Superimposed acquired (degenerative) stenosis
- Short, thickened pedicles

Staging, Grading, & Classification

- Cervical spine
  - Torg ratio (AP canal diameter/AP vertebral body diameter) < 0.8
  - Absolute diameter < 14 mm
- Lumbar spine
  - Spinal stenosis present if AP canal diameter at L1 < 20 mm, L2 < 19 mm, L3 < 19 mm, L4 < 17 mm, L5 < 16 mm, S1 < 16 mm
  - Lumbar spine: Critical stenosis where surgery required at L4 < 14 mm, L5 < 14 mm, S1 < 12 mm

Gross Pathologic & Surgical Features

- Sagittal canal narrowed secondary to short pedicles, thick laminae, and protrusion of inferior facet articular processes
- Lateral recess often narrowed secondary to coexistent hypertrophic superior facet articular processes

Microscopic Features

- Histologically normal bone

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Lumbar
    - Low back pain, radiating leg pain (unilateral or bilateral), ± bladder and bowel dysfunction
    - Neurogenic claudication: Radiating leg pain with walking, relieved by rest, bending forward
    - Cauda equina syndrome (rare): Bilateral leg weakness, urinary retention due to atonic bladder
  - Cervical
    - Radiating arm pain or numbness
    - Progressive myelopathy (spinal cord dysfunction) or reversible acute neurologic deficits ("stingers")
- Clinical profile
  - Symptomatic cervical or lumbar stenosis symptoms at younger age than typical of degenerative stenosis
    - These patients typically lack complicating medical problems (diabetes or vascular insufficiency)
  - Athletes present with temporary neurological deficit following physical contact that subsequently resolves

Demographics

- Age
  - Symptoms may arise in teens; more commonly in 4th to 5th decade
- Gender
  - M ≥ F
- Ethnicity
  - No ethnic or racial predilection
- Epidemiology
  - Prevalence in general population difficult to establish; up to 30% of surgically proven lumbar stenosis in 1 study
    - Not uncommonly seen in clinical practice during routine cervical or lumbar spine evaluation
    - Congenital cervical stenosis reported in 7.6% of 262 high school and college football players
  - Symptomatic at earlier age than expected

Natural History & Prognosis

- May be incidental finding in younger patients
  - Borderline cases are usually asymptomatic until superimposed acquired spinal degenerative disease occurs
- Many patients eventually develop symptomatic spinal stenosis
  - Minor superimposed acquired abnormalities (bulge, herniation, osteophyte) cause severe neurologic symptoms
- Early surgical treatment important for best outcome
  - Surgical decompression usually relieves symptoms effectively
  - Long-term pain relief following surgery common

Treatment

- Lumbar: Decompressive laminectomy, posterior foraminotomy at involved levels
  - Risk of complications increases with more levels of decompression, diabetes, and long-term steroid use
- Cervical: Posterior cervical laminectomy or laminoplasty
  - Same risk factors for complication as lumbar decompression

DIAGNOSTIC CHECKLIST

Consider

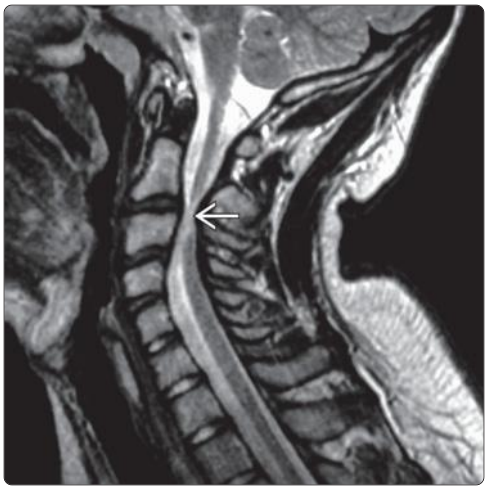
- Patients present at younger age than typical for degenerative spine disease
- Congenital spinal stenosis may not produce symptoms until superimposed acquired degenerative changes accumulate

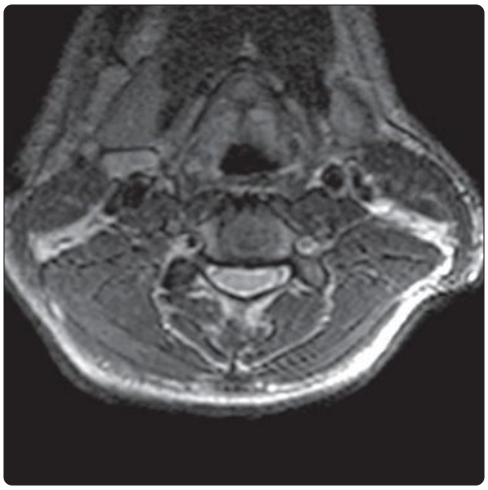
Image Interpretation Pearls

- Recognition of short, thick pedicles, AP canal diameter reduction key to diagnosis
- Look for superimposed degenerative changes

SELECTED REFERENCES

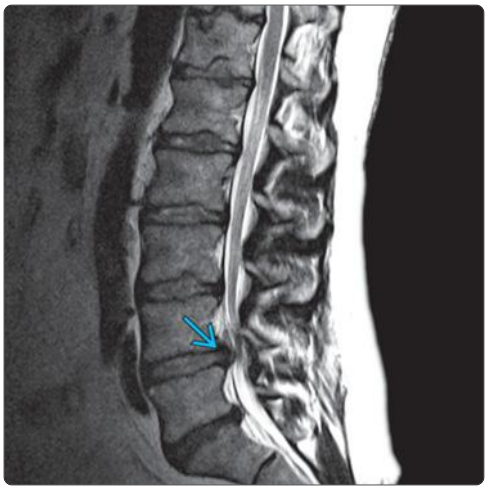
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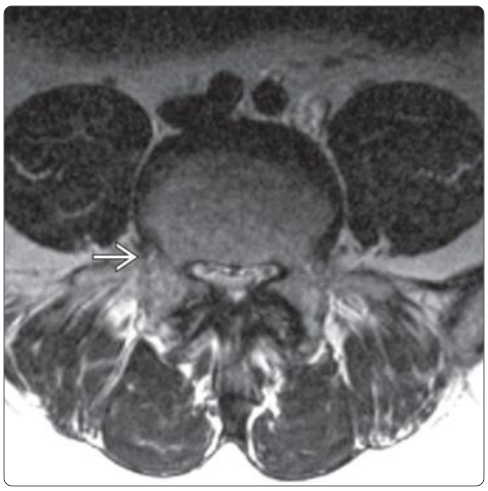


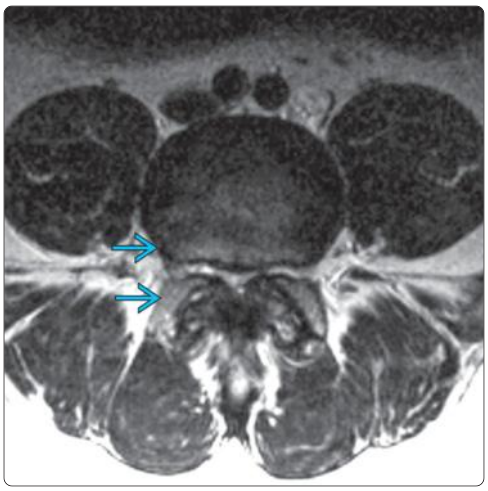
(Left) *Sagittal T2WI MR shows marked congenital stenosis at C3 due to abnormally short pedicles. Cord T2 hyperintensity ➡ corresponds to clinical myelopathy.* **(Right)** *Axial T2WI MR demonstrates moderate narrowing of the cervical AP canal diameter. No disc herniation or osseous degenerative changes are noted. Typical cervical findings include reduced AP canal diameter and short, laterally oriented pedicles.*





(Left) *Axial T1W MR shows congenital central stenosis with short and squat pedicles and narrowing of the AP diameter of the bony canal.* **(Right)** *Sagittal T2WI MR reveals congenital lumbar AP canal diameter reduction complicated by superimposed degenerative disc disease. A L4-L5 disc herniation ⇨ severely narrows central canal.*

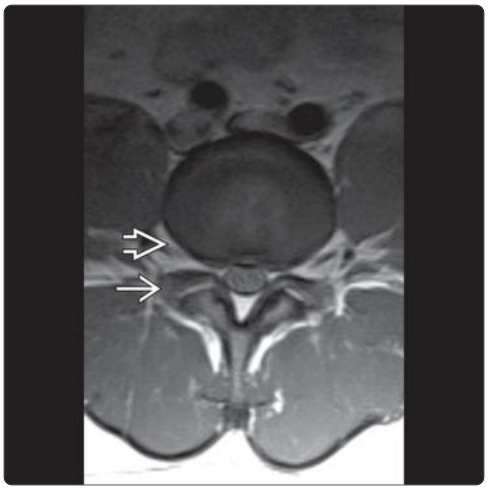




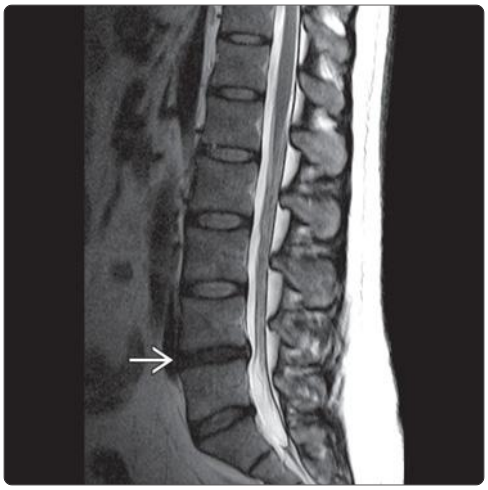
(Left) *Axial T2WI MR shows the typical pattern of L4-L5 central canal stenosis with a combination of acquired facet hypertrophy and congenitally shortened pedicles. Congenitally short pedicles ➡ with narrowed lateral recesses are also demonstrated.* **(Right)** *Axial T2WI shows spinal stenosis secondary to disc bulge, ligamentum flavum laxity, hypertrophied facets, and component of congenital stenosis with close approximation of facets to the vertebral body ⇨.*

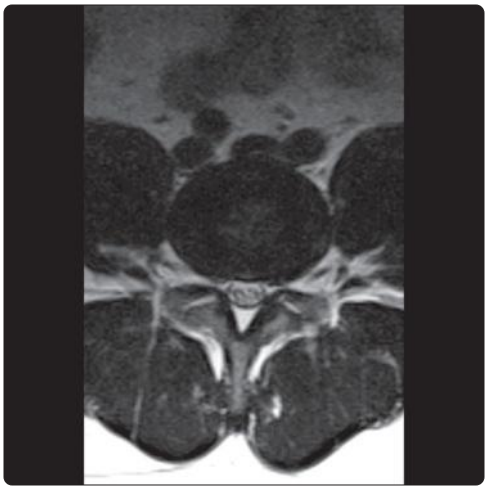
(Left) *Sagittal T1WI MR demonstrates moderate congenital AP canal diameter reduction spanning the L3 through the sacrum. In a normal spine imaging study, there should be an increase rather than reduction of AP diameter within the lower lumbar spine.* **(Right)** *Axial T2WI MR shows congenital spinal canal stenosis with a small AP diameter of the bony canal and facets ⇨ that approach the posterior margin of the intervertebral disc ⇨, reflecting the short pedicles that are present.*



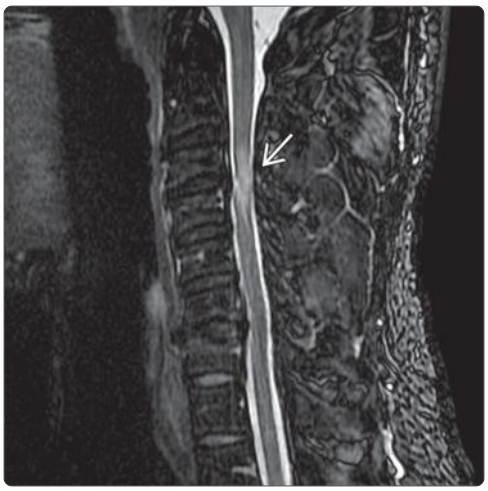


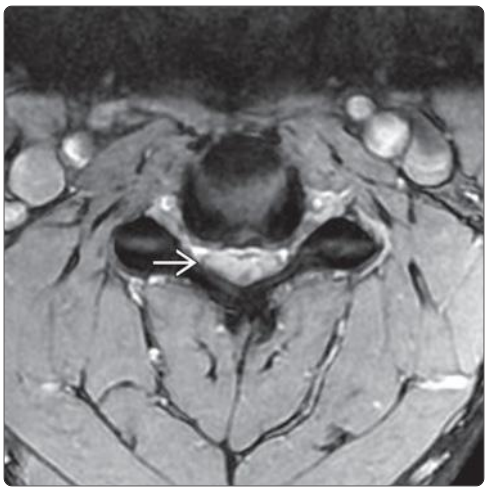
(Left) *Sagittal T2WI MR shows diffuse narrowing of the AP diameter of the subarachnoid space in this patient with diffuse congenital spinal canal stenosis. Note the mild acquired stenosis at L4-L5 ⇨ with bulging of the anulus and disc degeneration with signal loss.* **(Right)** *Axial T2 TSE shows the typical appearance of congenital spinal stenosis with facets closely opposed to the intervertebral disc due to short pedicles.*



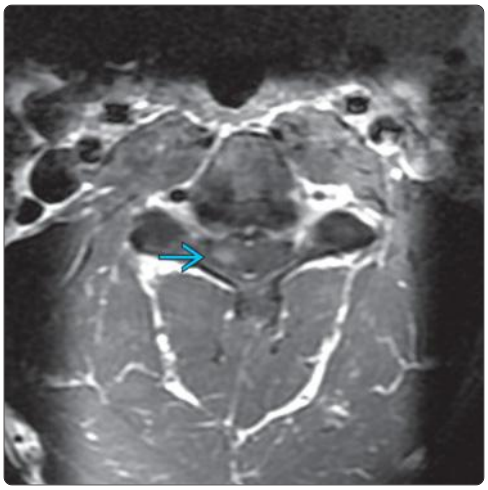


(Left) *Sagittal STIR MR shows focal cord T2 hyperintensity at the C3-C4 disc level ⇨ from myelomalacia due to minor disc disease coupled with congenital canal stenosis.* **(Right)** *Axial GRE study at C3-C4 shows diffuse high signal throughout the substance of the cord, which is a typical pattern for chronic traumatic myelomalacia related to disc disease. Note the small AP diameter of the bony canal due to congenital stenosis ⇨.*



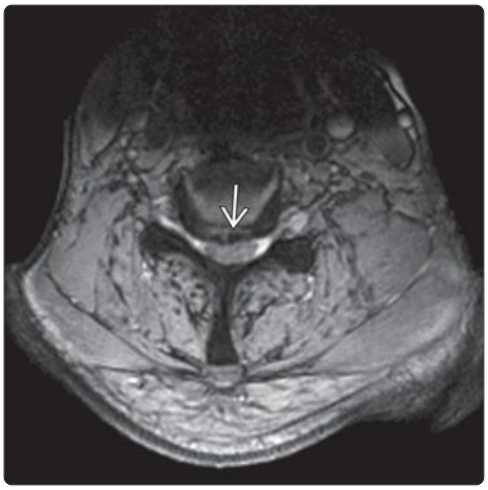






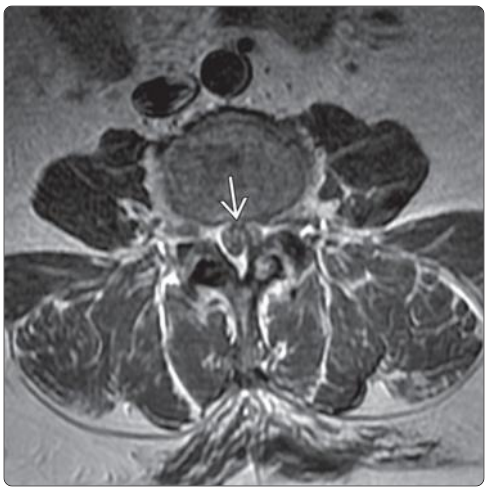
(Left) *Sagittal T1 C+ MR shows diffuse narrowing of the subarachnoid space, reflecting congenital canal stenosis. There is focal enhancement at C3-C4 within the right side of the cord due to myelomalacia from chronic cord trauma from the stenosis.* **(Right)** *Axial T1 C+ MR shows cord enhancement related to myelomalacia from minor disc disease coupled with congenital canal stenosis, causing repetitive cord trauma.*





(Left) *Sagittal T2WI MR shows diffuse congenital canal stenosis with a very small AP diameter of the subarachnoid space. There is cord myelomalacia at each level from repetitive trauma due to the tight canal.* **(Right)** *Axial GRE MR shows severe congenital central canal stenosis, the severity of which is compounded by acquired disc degeneration with a broad-based protrusion.*





(Left) *Sagittal T1 C+ MR shows diffuse congenital central canal stenosis, with superimposed acquired stenosis at L4-L5 and L5-S1 levels with disc bulges. Note the root enhancement at L3-L4 secondary to the stenosis.* **(Right)** *Axial T1 C+ MR shows congenital plus acquired central canal stenosis causing focal root enhancement with disruption of the blood-root barrier.*

Cervical Spondylosis

KEY FACTS

TERMINOLOGY

- Spinal canal and neural foraminal narrowing in cervical spine 2° to multifactorial degenerative changes

IMAGING

- Cervical spine, ventral epidural, centered at disc levels
- Sagittal diameter of cervical canal < 13 mm
- Disc osteophyte complex protruding into canal and compressing thecal sac and cord
- Completely effaced subarachnoid space at disc levels in cervical spine → "washboard spine"
- Variable degrees of cord compression

TOP DIFFERENTIAL DIAGNOSES

- Ossification of posterior longitudinal ligament
- DDx of abnormal cord signal
  - Cord tumor
  - Syrinx
  - Multiple sclerosis
  - Motor neuron disease

PATHOLOGY

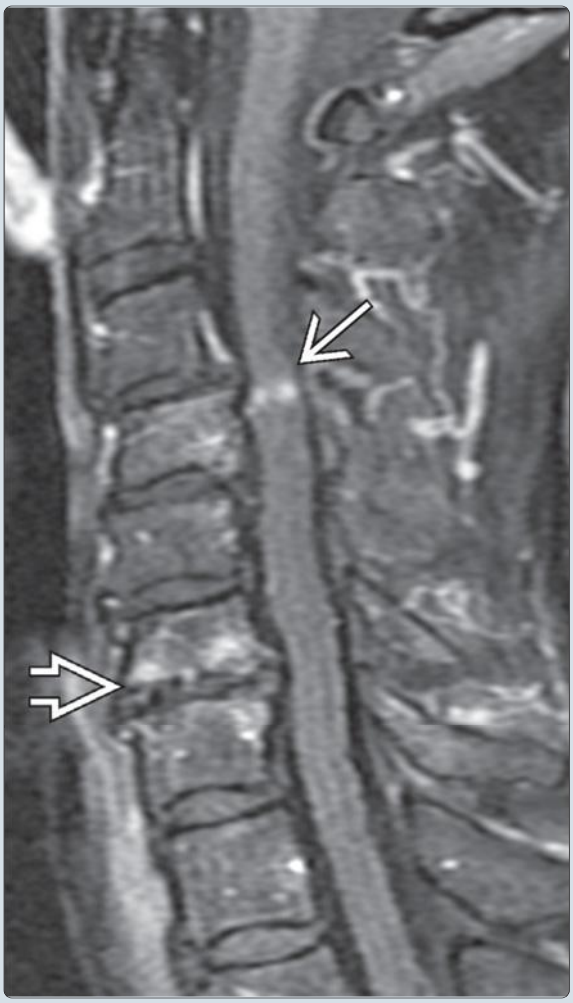
- Multilevel disc degeneration with decreased disc hydration and fissures
- Disc herniation, bulge with associated broad osteophytes
- Uncovertebral joint osteoarthritic change with foraminal stenosis

CLINICAL ISSUES

- ASSC has no pathognomic symptoms or signs
- Spastic paraparesis commonly seen
- Upper extremity numbness, weakness ("myelopathic hand")
- Natural history of insidious onset, periods of static disability, and episodic worsening



Sagittal T2WI MR shows multilevel degenerative disc disease and disc osteophyte complexes with canal stenosis spanning C3-C4 through C6-C7. T2 hyperintensity present within the cord centered at C3-C4 reflects myelomalacia.



Sagittal T1WI C+ FS MR shows horizontal linear enhancement within the cervical cord ⇨ at C3-C4 due to chronic myelomalacia. There is also enhancement of type I degenerative endplate changes at C6-C7 ⇨.

TERMINOLOGY

Abbreviations

- Acquired spinal stenosis, cervical (ASSC)

Synonyms

- Cervical spondylosis, cervical spondylotic myelopathy (CSM)

Definitions

- Spinal canal and neural foraminal narrowing in cervical spine 2° to multifactorial degenerative changes

IMAGING

General Features

- Best diagnostic clue
  - Completely effaced cerebral spinal fluid in cervical spine at disc levels
- Location
  - Cervical spine, ventral epidural, centered at disc levels
- Size
  - Sagittal diameter of cervical canal < 13 mm
- Morphology
  - Extradural degenerative change compressing thecal sac and cord
  - Congenitally short pedicles often contribute to acquired spinal stenosis

Radiographic Findings

- Radiography
  - Disc space height loss, osteophyte extending into canal, multilevel degenerative change

CT Findings

- NECT
  - Disc osteophyte complex protruding into canal and compressing thecal sac and cord
  - Uncovertebral and facet joint hypertrophy
  - Narrowing of neural foramina

MR Findings

- T1WI
  - Completely effaced subarachnoid space at disc levels in cervical spine → "washboard spine"
  - Variable degree of cord compression
  - Disc disease: Disc herniations, osteophytes, disc bulges
- T2WI
  - Narrowing of subarachnoid space due to ventral and dorsal epidural disease
    - Ventral low T2 signal due to disc osteophyte complex
    - Dorsal low T2 signal due to ligamentous thickening
  - Intramedullary T2 hyperintensity represents myelomalacia, demyelination, or edema
- T2* GRE
  - Uncovertebral and facet joint hypertrophy
  - Narrowing of neural foramina
- T1WI C+
  - Cord myelomalacia, contusion may enhance after gadolinium
  - Typical horizontal linear pattern involving central and peripheral cord

Nonvascular Interventions

- Myelography
  - Obliterated subarachnoid space at disc levels
  - Multiple horizontal "bars" or ridges on AP view due to subarachnoid space narrowing
  - Cord widened on AP view due to compression
  - Multiple levels of root cutoff

Imaging Recommendations

- Protocol advice
  - Sagittal and axial T1WI, sagittal T2WI, axial T2* GRE imaging (2-3 mm thickness, 3D preferred)

DIFFERENTIAL DIAGNOSIS

Ossification of Posterior Longitudinal Ligament

- A cause of cervical spinal stenosis
  - Thick band of hypointensity along posterior vertebral margin
  - Central hyperintensity within band represents fatty marrow

DDx of Abnormal Cord Signal

- Cord tumor
  - Intramedullary mass with enhancement
- Syrinx
  - Linear hyperintensity on T2WI, ± cord expansion, multiple septations
- Multiple sclerosis
  - Focal cord hyperintensity on T2WI
  - Brain periventricular lesions

DDx of Clinical Symptoms

- Motor neuron disease
  - Distinguished on clinical grounds, diagnosis of exclusion

PATHOLOGY

General Features

- Etiology
  - Nontraumatic, progressive, chronic compression of cervical spinal cord by static and dynamic factors
    - Static: Degenerative changes involving discs, vertebral endplates, uncovertebral joints, facet joints, and ligamentum flavum
    - Dynamic: Repetitive injury to compressed cord from flexion and extension movements
  - Congenitally short pedicles often present
- Associated abnormalities
  - Age-related degenerative disease
  - Lumbar canal stenosis (> 5%)

Staging, Grading, & Classification

- Sagittal diameter of spinal canal < 13 mm stenotic
- Torg ratio
  - Spinal canal: Vertebral body ratio < 0.8 on plain radiographs = cervical stenosis
  - Poor correlation of Torg ratio and sagittal diameter of spinal canal on CT
  - Not absolute, multiple factors involved in producing neurapraxia with trauma

- AP compression ratio (AP diameter cord/transverse diameter)
  - < 0.40 have worse outcome after surgical decompression
- Cord T2 hyperintensity
  - More clinically symptomatic
  - Less postoperative improvement

Gross Pathologic & Surgical Features

- Multilevel disc degeneration with decreased disc hydration and fissures
- Disc herniation, bulge with associated broad osteophytes
- Uncovertebral joint osteoarthritic change with foraminal stenosis

Microscopic Features

- Cord destruction of gray and white matter, with demyelination above and below compression levels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - ASSC has no pathognomic symptoms or signs
    - Spastic paraparesis commonly seen
    - Upper extremity numbness, weakness ("myelopathic hand")
    - Gait disturbance, loss of position and vibration sense
    - Chronic neck pain radiating to occiput and upper extremities
  - Cord syndromes
    - Transverse: Severe spasticity, sphincter involvement, Lhermitte
    - Motor system: Spasticity but minimal sensory disturbance
    - Central cord: Severe motor and sensory disturbance, worse for upper extremities
    - Brown-Séquard: Contralateral sensory and ipsilateral motor deficits
    - Brachialgia cord syndrome: Lower motor neuron → upper extremity involvement, upper motor neuron → lower extremity involvement, radicular pain
- Clinical profile
  - Neck pain, "heaviness" in arms, ataxic gait, hyperreflexia, extensor plantar reflex

Demographics

- Age
  - > 50
- Gender
  - M > F
- Epidemiology
  - 5th decade and later

Natural History & Prognosis

- Most common cause of spinal cord dysfunction worldwide
- Natural history of insidious onset, periods of static disability, and episodic worsening
  - New signs and symptoms (75%)
  - Slow steady progression of disability (20%)
  - Rapid onset with long period of stability (5%)
- Continued pain, disability, and poor outcomes (30-50%)
- More favorable outcome if treated early
- Severity and duration of preoperative neurologic deficits predictive of degree of neurologic recovery

Treatment

- Cervical spine
  - Medications
    - Anti-inflammatories, narcotics, antidepressants, muscle relaxants
  - Soft collar immobilization and traction
  - Physical therapy
    - Isometrics
    - Aerobic conditioning
    - Flexibility exercises
  - Depending on site of compression
    - Anterior corpectomy or interbody arthrodesis
    - Posterior decompression with laminectomies
    - Open-door laminaplasties ± foraminotomies
- Prognostic factors for good surgical outcome
  - Young age
  - < 1 year of symptoms
  - Unilateral motor deficit
  - Presence of Lhermitte sign
- Postoperative complications
  - Neurologic deterioration (5%)
  - Vertebral artery injury
  - Graft extrusion or fracture
  - Instrumentation failure or migration
  - Airway obstruction
  - Adjacent level degeneration
  - Reduced mobility

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- MR of cervical spine may overestimate neural foraminal stenosis
  - Short TE on axial T2* images decreases this artifact

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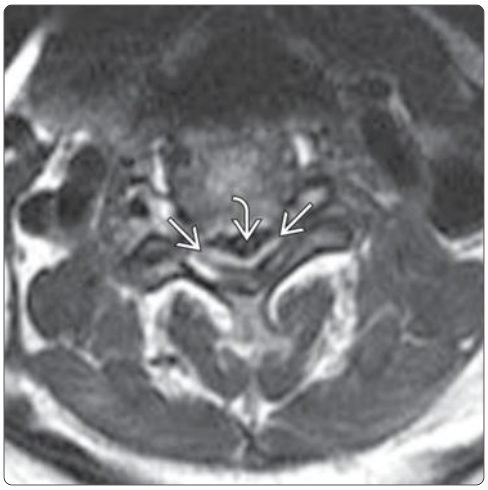
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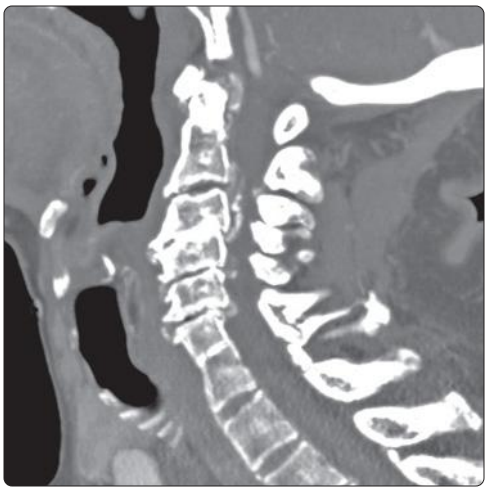


(Left) *Sagittal graphic shows disc degeneration with osteophyte formation causing central stenosis and cord compression at multiple levels (cervical spondylosis).* **(Right)** *Sagittal STIR MR shows severe multilevel stenosis, which is worst at C2-C3, C3-C4. Cord hyperintensity reflecting myelomalacia is present at both levels. Congenital fusion of C4 and C5 bodies accelerates the degenerative process.*





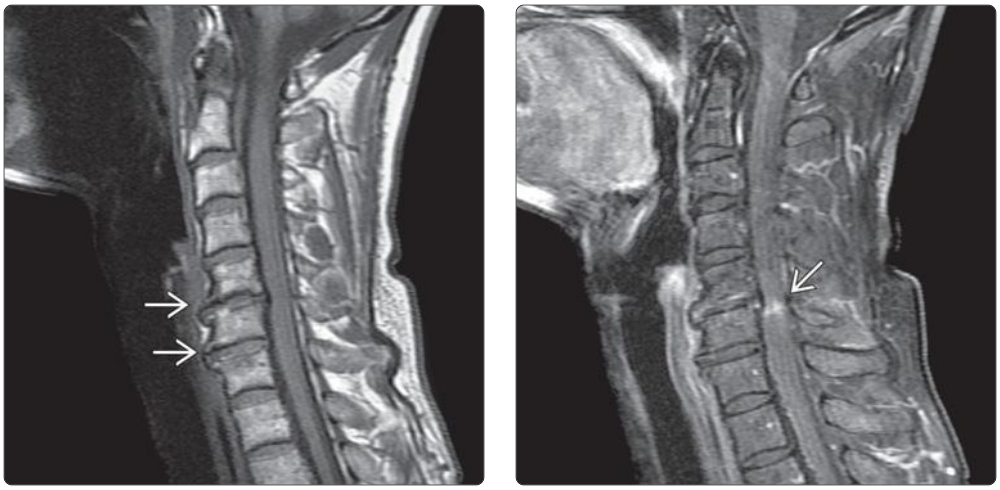
(Left) *Sagittal T2WI MR shows diffuse disc degeneration with severe multilevel subarachnoid space narrowing from disc osteophyte complexes. There is abnormal cord T2 signal at C4-C5 due to chronic trauma and myelomalacia.* **(Right)** *Axial T2WI MR shows a markedly thinned cord as relatively high signal that is outlined ventrally by low signal disc osteophyte complex and posteriorly by thickened low signal ligamentum flavum.*



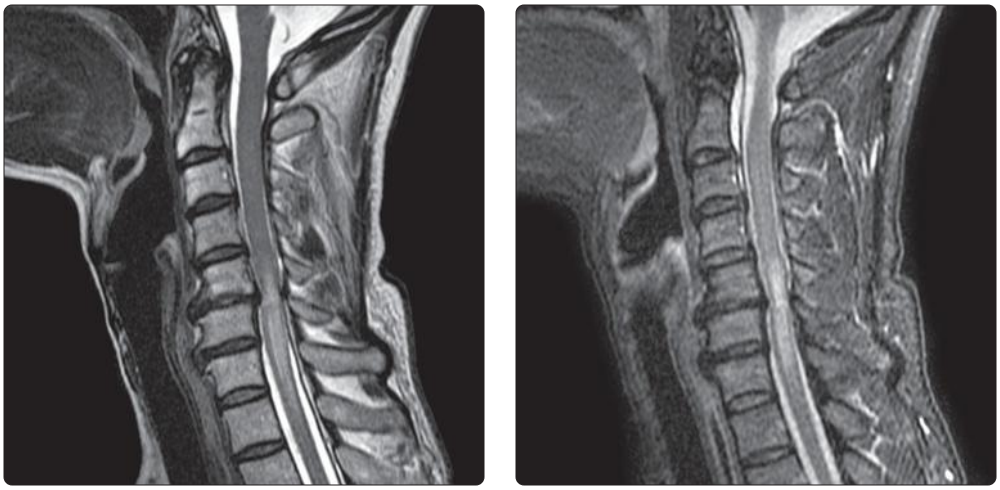


(Left) *Sagittal CECT shows very severe multilevel central canal stenosis from C2-C3 through C5-C6 due to diffuse disc bulge and diffuse broad-based herniations anteriorly with associated disc and PLL calcification. Note congenital fusion at C6-C7.* **(Right)** *Axial CECT shows central canal stenosis due to broad-based herniation anteriorly with associated disc and PLL calcification. Posterior ligamentous hypertrophy adds to severe central stenosis. There is bilateral foraminal stenosis with uncovertebral joint hypertrophy.*

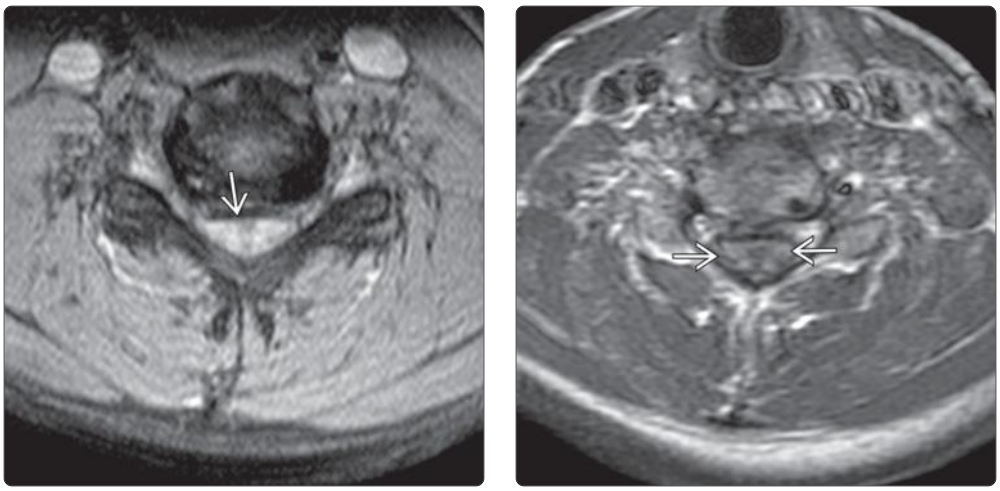
(Left) *Sagittal T1WI MR shows disc degeneration at C5-C6 and C6-C7. There is severe central canal stenosis at C5-C6 with effacement of the thecal sac and moderate cord compression.* **(Right)** *Sagittal T1 C+ FS MR shows focal linear enhancement extending transversely through the cord at the level of most severe central stenosis. Enhancement is due to blood-cord barrier disruption from chronic repetitive compression.*



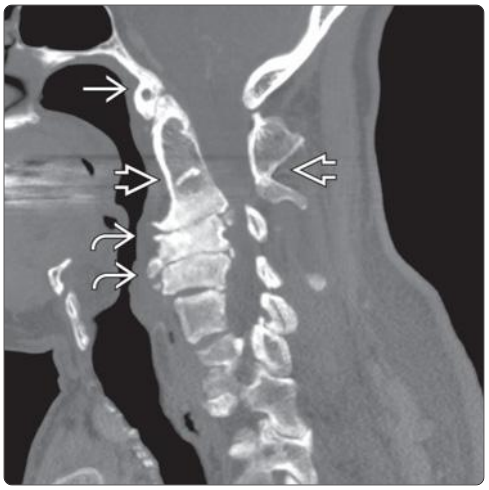
(Left) *Sagittal T2WI MR shows the typical appearance of cord myelomalacia from cervical spondylosis. Disc degeneration as manifested by loss of T2 signal and loss of disc space height at C5-C6 and C6-C7. There is severe central canal stenosis at C5-C6 with effacement of the thecal sac.* **(Right)** *Sagittal STIR MR shows a cord increased signal (myelomalacia) that has occurred from the disc-osteophyte complex at the C5-C6 level.*



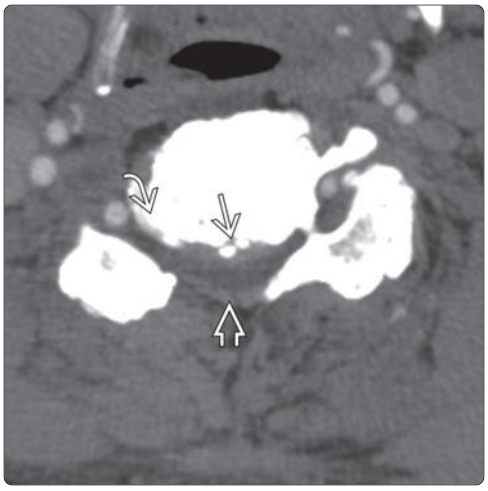
(Left) *Axial GRE MR shows the severity of the central canal stenosis with flattening of the ventral cord and loss of all normal surrounding CSF signal. There is diffuse cord hyperintensity, with a slightly more prominent signal from the central gray matter.* **(Right)** *Axial T1 C+ FS MR shows diffuse patchy enhancement extending transversely through the cord at the level of most severe central stenosis, due to enhancing myelomalacia.*

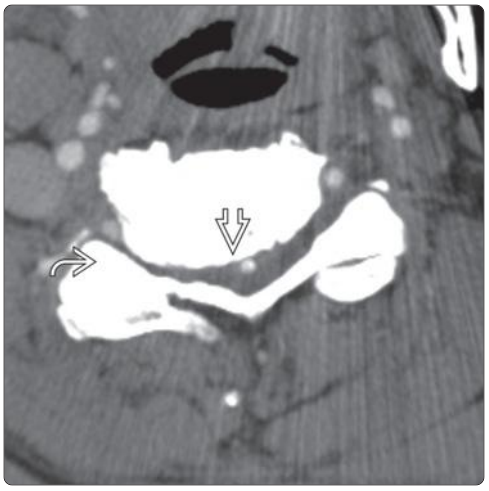






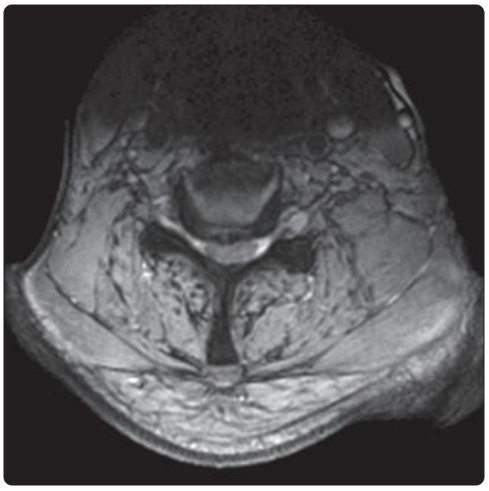
(Left) *Coronal bone NECT shows severe accelerated degenerative disc disease with foraminal stenosis complicating Klippel-Feil segmentation anomalies and assimilation of C1 into the occiput.* **(Right)** *Sagittal bone NECT shows assimilation of C1 to occiput with a fused anterior arch with the basion. The C2 is congenitally fused to C3. Severe disc degeneration is present at multiple levels, with marked loss of disc height and extensive bony sclerosis with osteophytes.*





(Left) *Axial CT angiogram shows severe central canal stenosis from ventral disc-osteophyte complex producing cord compression. There is severe bony foraminal stenosis with facet hypertrophy and uncovertebral degeneration. Thickening of the ligamentum flavum is also present.* **(Right)** *Axial CT angiogram shows severe central canal stenosis from ventral disc-osteophyte complex with marked cord compression. There is severe right foraminal stenosis with uncovertebral joint hypertrophy.*





(Left) *Sagittal T2WI MR shows diffuse congenital central stenosis with acquired mild disc osteophyte complex at multiple levels. This stenosis results in multilevel cord myelomalacia.* **(Right)** *Axial GRE MR shows severe central canal stenosis from ventral disc-osteophyte complex with underlying congenital central stenosis.*

DISH

KEY FACTS

TERMINOLOGY

- Diffuse idiopathic skeletal hyperostosis (DISH)
- Forestier disease, senile ankylosing hyperostosis, asymmetrical skeletal hyperostosis

IMAGING

- Flowing anterior vertebral ossification with minimal degenerative disc disease, facet arthropathy, absent facet ankylosis
- Thoracic spine (100%) > cervical (65-80%), lumbar spine (68-90%); R > L
- Lateral radiography inexpensive, reliable
- Reserve MR to evaluate for coexistent OPLL or spondylosis-related cord compression

TOP DIFFERENTIAL DIAGNOSES

- Spondylosis
- Ankylosing spondylitis
- Psoriatic or reactive (Reiter) arthritis

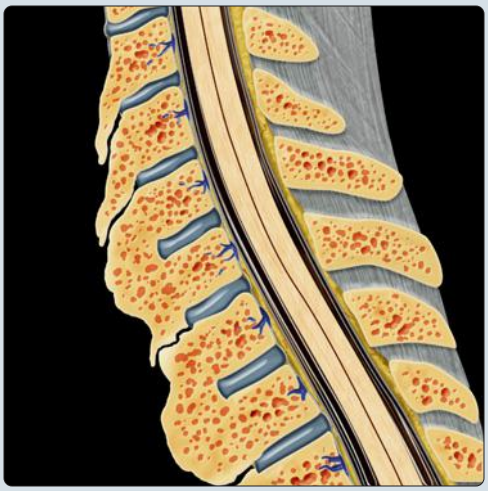
PATHOLOGY

- Exact cause for exaggerated new bone formation stimuli unknown
  - Exuberant enthesial reaction at tendon, ligament, and joint capsule insertions
  - Associated with OPLL
  - Dysphagia related to DISH multifactorial
- Primary diagnostic criteria for DISH
  - Flowing anterior ossification extending over at least 4 contiguous vertebral bodies
  - No apophyseal or SI joint ankylosis
  - Mild degenerative disc changes, no facet ankylosis

CLINICAL ISSUES

- Majority of cases incidental
- Osteophyte resection if severe symptoms
- Increased risk of extension-type fractures with high morbidity

(Left) *Sagittal graphic shows bulky flowing ossification of the anterior longitudinal ligament extending over more than 4 contiguous vertebra. Disc spaces are relatively preserved.* **(Right)** *Lateral radiograph shows a large anterior ossified mass that is discontinuous at several disc spaces, a variation implying some degree of continued cervical spine mobility.*





(Left) *Sagittal T1WI MR shows bulky anterior flowing ossification ➡ consistent with DISH. Fatty marrow is responsible for high T1 signal intensity within the anterior ossification.* **(Right)** *Sagittal T1WI MR right parasagittal slice shows bulky flowing ALL ossification ➡ spanning more than 4 vertebral levels, but minimal disc abnormality that is typical of DISH.*





TERMINOLOGY

Abbreviations

- Diffuse idiopathic skeletal hyperostosis (DISH)

Synonyms

- Forestier disease, senile ankylosing hyperostosis, asymmetrical skeletal hyperostosis
- Spondylosis hyperostotica, spondylitis ossificans ligamentosa

Definitions

- Flowing coarse osteophytes bridging 4 vertebral adjacent bodies

IMAGING

General Features

- Best diagnostic clue
  - Flowing anterior vertebral ossification with minimal degenerative disc disease, facet arthropathy, and absent facet ankylosis
- Location
  - Thoracic spine (100%) > cervical (65-80%), lumbar spine (68-90%); R > L
- Size
  - Range from small, focal to large, and extensive
- Morphology
  - Bulky flowing multilevel ossification anterior to vertebral bodies
  - Predominantly right side of thoracic spine

Radiographic Findings

- Radiography
  - Flowing anterior vertebral ossification, variably distinguishable from anterior vertebral body cortex
    - Earliest sign is new bone formation adjacent to midvertebral body
    - May coexist with spondylosis
  - Relative preservation of disc spaces and facets

Fluoroscopic Findings

- Lateral flexion-extension imaging assesses dynamic spinal mobility

CT Findings

- Bone CT
  - Thick ALL ossification, R > L
  - Relatively minimal degenerative disc, facet disease
  - Absent facet, sacroiliac joint erosion, or ankylosis
  - DISH changes detectable between disc levels

MR Findings

- T1WI
  - Flowing ALL ossification along anterior vertebral bodies; may be difficult to distinguish from anterior vertebral body
    - ALL marrow is not contiguous with vertebral body marrow
    - Hypointense if predominantly calcified
    - Isointense or hyperintense if marrow fat present
- T2WI
  - Similar findings to T1WI; ALL ossification may be hypointense unless substantial fatty marrow content
- T2* GRE
  - Hypointense flowing "mass" within ALL
- T1WI C+
  - May show minimal enhancement (similar to vertebral marrow)

Nonvascular Interventions

- Myelography
  - Similar osseous findings to plain radiography
  - No canal stenosis unless concurrent ossification of posterior longitudinal ligament (OPLL)

Imaging Recommendations

- Best imaging tool
  - Lateral radiography inexpensive, reliable for detecting DISH
- Protocol advice
  - AP and lateral plain radiographs
  - Axial bone algorithm CT with sagittal and coronal reformats to confirm plain film diagnosis
  - MR unnecessary for DISH diagnosis
    - MR to evaluate for coexistent OPLL or if cord compression or spondylosis are being considered within differential diagnosis
    - MR for evaluation of CT/plain film occult fractures in setting of trauma

DIFFERENTIAL DIAGNOSIS

Spondylosis

- Rarely contiguous across 4 or more vertebral levels; usually confined to disc interspace vicinity
- More substantial facet, disc degenerative changes than DISH
- No predisposition to right or left side

Ankylosing Spondylitis

- Thin intervertebral syndesmophytes instead of bulky flowing DISH ossification
- Sacroiliac (SI) joint, facet erosion/ankylosis
- Appropriate clinical history; HLA B27 ≥ 90%

Psoriatic or Reactive (Reiter) Arthritis

- Large bulky osteophytes; often lateral with skip areas
- Erosive or ankylosing changes of SI joints, appropriate clinical history

PATHOLOGY

General Features

- Etiology
  - Exact cause for exaggerated response to new bone formation stimuli unknown
  - Possible associations with diabetes mellitus, dyslipidemia, hyperuricemia, alcohol intake, and poor dietary habits postulated but not definitively proven
  - Multiple possible genetic influences including COL6A1 (type VI collagen), IGF-1, Dickkopf-related protein 1 (*DKK-1*), bone morphogenic protein 2 (BMP2)
- Associated abnormalities
  - OPLL

- Exuberant enthesial reaction at tendon, ligament, and joint capsule insertions
  - Iliac crest (66%), ischial tuberosities (53%) producing "pelvic whiskering"
  - Lesser (42%) and greater (36%) femoral trochanters, patellar quadriceps insertion (29%)
  - Osseous bridging of fibula to tibia (10%)
  - Distal metacarpal, phalangeal periarticular capsular hyperostosis (13%)
- Decreased spine mobility due to ALL ossification; relative protection from degenerative disc disease and facet arthropathy
- May have coexistent exuberant ossification of tendon, ligament, or joint capsule insertions ("enthesitis")
- Dysphagia due to DISH probably multifactorial
  - Combination of direct mechanical compression + inflammation/fibrosis of esophageal wall, esophageal denervation
- R > L predilection; attributed to effect of repetitive aortic pulsations retarding proliferative ossification

Staging, Grading, & Classification

- Several classification schemes related to radiographic abnormalities, Resnick (1976) most commonly utilized
- Primary diagnostic criteria for DISH
  - Flowing anterior ossification extending over at least 4 contiguous vertebral bodies
  - No apophyseal or SI joint ankylosis
  - Relatively minimal degenerative disc changes, no facet ankylosis

Gross Pathologic & Surgical Features

- Bulky anterior vertebral ossification with normal cortical bone, marrow appearance

Microscopic Features

- Normal-appearing Haversian bone, marrow within ossified ligament

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Frequently incidental finding; patient may complain of intermittent spinal stiffness, restricted mobility
    - Often worse in morning, after prolonged sitting, or during cold winter weather
    - May be relieved by mild activity
  - Pain related to associated enthesitis, tendinitis
  - Predisposition to extension-type fractures of spine
  - Other signs/symptoms
    - Anterior osteophytes may compress esophagus resulting in dysphagia
    - Stridor requiring tracheostomy (rare)
- Clinical profile
  - Usually incidental observation in asymptomatic Caucasian patient imaged for unrelated reasons

Demographics

- Age
  - Middle-aged to older adults; uncommon before age 50, common in elderly
- Gender
  - M:F = 2:1
- Ethnicity
  - Caucasian > > African American, Native American, or Asian populations
  - Decreased incidence in osteoporotic Caucasian populations
- Epidemiology
  - Reported incidence varies widely
    - Conservative estimate: 6-12% of adults > 40 years of age manifest some findings of DISH; actual incidence may be substantially higher
    - More common in elderly men

Natural History & Prognosis

- Nearly always incidental finding without additive morbidity or mortality
  - Severe cases may rarely produce dysphagia
  - Increased predisposition to fracture due to decreased fusion with increased lever arm
  - Increased risk of heterotopic ossification after total hip arthroplasty

Treatment

- Vast majority of cases incidental and merit conservative (observational) management
- Consider osteophyte resection if severe symptoms directly referable to DISH

DIAGNOSTIC CHECKLIST

Consider

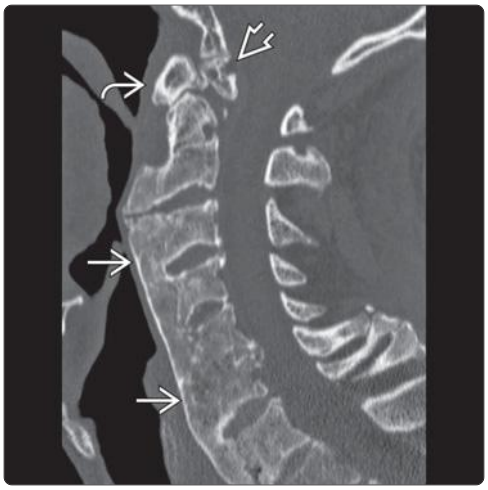
- DISH is largely asymptomatic and incidentally detected
  - Look carefully for signs of associated OPLL producing myelopathy due to central canal stenosis
- Ankylosed spine (whether due to ankylosing spondylitis or DISH) is at risk for hyperextension-type spine injuries
  - High morbidity

Image Interpretation Pearls

- Flowing ossification classically R > L
- Discs/facet joints relatively preserved, no SI joint erosion or ankylosis
- May coexist with degenerative spondylosis

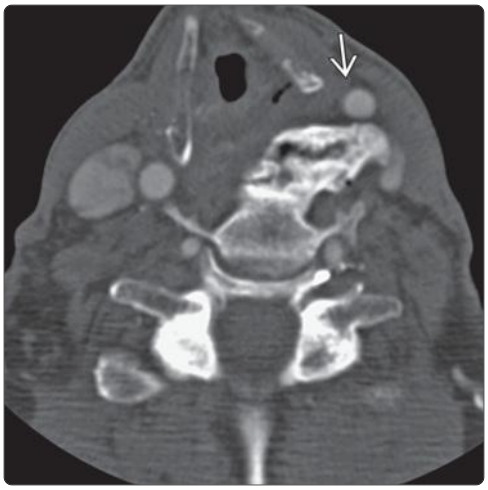
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(Left) *Sagittal NECT shows large flowing ventral osteophytes from C2 through the upper thoracic spine. There is no bony fusion at C2-C3, allowing limited motion at this segment. Note the blunted odontoid with a hypertrophic anterior C1 arch and os odontoideum articulating with the basion.* **(Right)** *Sagittal NECT shows exuberant anterior osteophyte formation throughout the cervical and upper thoracic spine, displacing the hypopharynx anteriorly.*





(Left) *Axial NECT shows a huge osteophyte displacing the left carotid artery anteriorly and effacing the left hypopharynx and larynx.* **(Right)** *Sagittal NECT shows bone formation at multiple sites in this patient with DISH. Typical anterior vertebral ossification is present, as well as OPLL and disc calcification. There is also ossification of the ligamentum flavum.*





(Left) *Sagittal NECT shows a C6-C7 level hyperextension injury with widening of the anterior disc space in this patient with DISH and diffuse spinal ankylosis.* **(Right)** *Sagittal thick slice MIP projection of the cervical spine shows severe fracture dislocation at the C7-T1 level. There is extensive bony ankylosis from DISH throughout the cervical spine with coarse vertically oriented osteophytes.*

OPLL

KEY FACTS

TERMINOLOGY

- Ossification of posterior longitudinal ligament

IMAGING

- Flowing multilevel ossification posterior to vertebral bodies, with relatively minimal degenerative disc disease, absent facet ankylosis
  - Midcervical (C3-C5) > midthoracic (T4-T7)
  - PLL ossification narrows AP spinal canal dimension → spinal stenosis, cord compression
- Characteristic "upside-down T" or "bowtie" PLL configuration on axial images
  - CT with sagittal reformats to confirm MR diagnosis, clarify extent of ossification for surgical planning
  - Sagittal T1WI, T2WI to evaluate spinal cord compression, extent of ligamentous ossification

TOP DIFFERENTIAL DIAGNOSES

- Spondylosis
- Calcified herniated disc
- Meningioma

PATHOLOGY

- Continuous → ossified mass over several vertebral segments
- Segmental → fragmented ossified lesions behind each vertebral body
- Mixed → combination of continuous and segmental
- Other → ossification confined to disc level

CLINICAL ISSUES

- Spastic paresis → paralysis (17-22%)
- ↑ risk for developing progressive myelopathy if more than 60% canal stenosis, ↑ cervical range of motion

(Left) *Sagittal graphic shows flowing multilevel ossification within the posterior longitudinal ligament (PLL) producing canal narrowing and cord compression.* **(Right)** *Sagittal STIR MR shows the typical pattern of continuous type OPLL seen as low signal ventral to the cord ⇨ with severe cord compression. Note the associated DISH with multilevel fusion ⇨.*





(Left) *Axial GRE MR shows a rectangular focus of markedly low signal ventral to the cord, consistent with OPLL ⇨. There is severe cord compression ⇨.* **(Right)** *Sagittal T1 C+ MR shows a slightly heterogeneous mass ventral to the cervical cord, with severe cord compression. OPLL may show low or heterogeneous increased signal on T1W images due to marrow content with fat elements.*





TERMINOLOGY

Abbreviations

- Ossification of posterior longitudinal ligament (OPLL)

Definitions

- Ossification within spinal posterior longitudinal ligament (PLL)

IMAGING

General Features

- Best diagnostic clue
  - Flowing multilevel ossification posterior to vertebral bodies, with relatively minimal degenerative disc disease, absent facet ankylosis
- Location
  - Midcervical (C3-C5) > midthoracic (T4-T7)
- Size
  - Mild focal thickening → extensive flowing ossification (≈ 2-5 mm thickness)
- Morphology
  - PLL ossification narrows AP spinal canal dimension → spinal stenosis, cord compression

Radiographic Findings

- Radiography
  - Flowing posterior ossification behind vertebral bodies
    - Often superimposed over facet complex on lateral projection
    - Requires high index of suspicion to diagnose; subtle finding, easy to overlook

Fluoroscopic Findings

- Flex-extend imaging determines dynamic range

CT Findings

- NECT
  - Characteristic "upside-down T" or "bowtie" PLL configuration on axial images
- Bone CT
  - PLL ossification appearance similar to NECT findings
    - Cortical bone surrounding central marrow space
    - May appear contiguous with adjacent vertebral cortex or distinct from posterior vertebral body
    - PLL, vertebral marrow spaces discrete

MR Findings

- T1WI
  - Posterior flowing ossification extending over multiple levels on sagittal images
  - Characteristic "upside-down T" or "bowtie" configuration on axial images
  - Usually low signal intensity on all pulse sequences
    - May show high signal intensity if significant marrow fat content
- T2WI
  - PLL findings similar to T1WI
  - ± cord hyperintensity 2° to myelomalacia, edema
- T2* GRE
  - Hypointense signal intensity within PLL ossification
  - Caveat: Exaggerates severity of stenosis 2° to magnetic susceptibility effects

Nonvascular Interventions

- Myelography
  - Flowing PLL ossification narrows ventral spinal canal
    - Spinal stenosis → cord compression
    - ± spinal block (severe cases)

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Sagittal T1WI, T2WI to evaluate spinal cord compression, extent of ligamentous ossification
  - Axial T2WI to determine stenosis severity, confirm cord signal abnormality
  - CT with sagittal reformats to confirm MR diagnosis, clarify extent of ossification for surgical planning

DIFFERENTIAL DIAGNOSIS

Spondylosis

- Usually centered at disc interspace; rarely contiguous across 4 or more vertebral levels
- More substantial facet, disc degenerative changes than OPLL
- Absence of characteristic T-shaped PLL ossification

Calcified Herniated Disc

- Focal calcified "mass" centered at a single disc space level
- Lacks characteristic T-shaped PLL ossification

Meningioma

- Avidly enhancing dural-based mass + dural "tail," smooth margins
- Often T2 hypointense 2° to calcification
- Lacks characteristic T-shaped PLL ossification

Cervical Peridural Calcification of Hemodialysis

- Curvilinear dural calcification; may be circumferential
- Hemodialysis patient ± myelopathy, sensory abnormalities
- Requires dural resection in addition to laminectomy/laminoplasty

PATHOLOGY

General Features

- Etiology
  - Not conclusively determined
  - Postulated etiologies include infectious agents, autoimmune disorders, or trauma
- Genetics
  - Chromosome 6 gene (*COL11A2*) polymorphisms → abnormal α chain of type XI collagen → OPLL susceptibility
  - Polymorphisms of collagen 6A1 gene (*COL6A1*) → abnormal α chain of type VI collagen → OPLL susceptibility
- Associated abnormalities
  - Hypertrophy of posterior longitudinal ligament (HPLL) seen as precursor to OPLL

- Diffuse idiopathic skeletal hyperostosis (DISH) seen in 25% of patients with OPLL
- OPLL present in 16% of ankylosing spondylitis cases in Mexico and 23% in Canada
- OPLL present in 16-20% of cases of ossification of ligamentum flavum (OLF)

- Idiopathic PLL ossification predominately described in Japanese > other Asian populations, less common in Caucasians
- More recently described early stage "OPLL in evolution" noted in both Asian, Caucasian populations
  - Hypertrophied PLL with punctate calcifications rather than smooth, flowing ossific mass
  - Difficult to distinguish from more common spondylosis at this early stage
- OPLL patients have higher bone mineral density (BMD) than age-matched controls
  - Possible predisposition for excessive bone deposition

Staging, Grading, & Classification

- Continuous → ossified mass over several vertebral segments
- Segmental → fragmented ossified lesions behind each vertebral body
- Mixed → combination of continuous and segmental
- Other → ossification confined to disc level

Gross Pathologic & Surgical Features

- Bulky posterior ligamentous ossification with normal cortical bone, marrow
- OPLL may transgress dura

Microscopic Features

- Histologically normal Haversian bone and marrow within ossified ligament

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Incidental observation in asymptomatic patient
  - Symptomatic patients present with myelopathy referable to stenosis level
    - Symptomatic myelopathy almost universal if canal diameter < 6 mm; rare if > 14 mm
    - Canal diameter 6-14 mm variable presentation; greater cervical mobility → more likely to be symptomatic
- Clinical profile
  - Classic presentation is Japanese patient with progressive quadriparesis or paraparesis

Demographics

- Age
  - Usually age > 50 years; rare < 30 years
- Gender
  - M:F = 2:1
- Ethnicity
  - ↑ prevalence in Japanese population
- Epidemiology
  - 2-4% prevalence in Japan; much less common elsewhere

Natural History & Prognosis

- Mild cases asymptomatic → incidental discovery
- Spastic paresis → paralysis (17-22%)
  - Patients with mild OPLL at diagnosis rarely develop severe canal stenosis on follow-up
  - Patients presenting with myelopathy very likely to clinically progress
  - Mild trauma may exacerbate myelopathy
  - Progression more marked in young patients with continuous type disease
- ↑ risk for developing progressive myelopathy if > 60% canal stenosis, ↑ cervical range of motion
  - ↓ spinal mobility appears to protect spinal cord from injury

Treatment

- Asymptomatic patients: Careful observation, nonoperative management
- Symptomatic or high-grade stenosis patients: Anterior (corpectomy) or posterior (laminectomy or laminoplasty) decompression

DIAGNOSTIC CHECKLIST

Consider

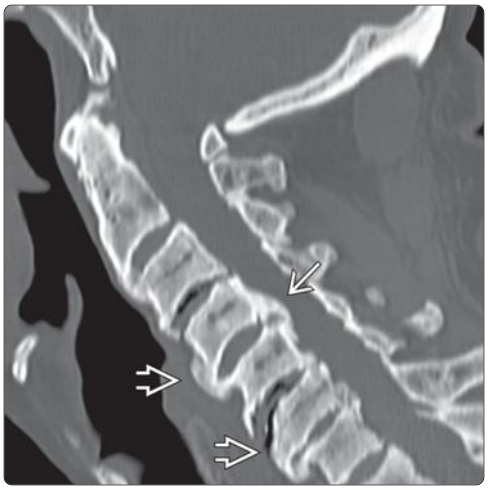
- Look carefully for OPLL on lateral plain films; confirm with multiplanar CT or MR

Image Interpretation Pearls

- Search for flowing multilevel ossification behind vertebral bodies
- Overly "thick" hypointense PLL on MR strongly suggestive of OPLL
- Requires high index of suspicion, careful observation to diagnose on plain radiographs

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(Left) *Sagittal NECT demonstrates a focal type of OPLL occurring primarily at the disc space level and associated with DISH.* **(Right)** *Axial NECT shows the large focal area of ossification from OPLL projecting into the spinal canal with severe canal compromise.*





(Left) *Sagittal bone CT shows extensive flowing PLL ossification that severely narrows the central canal. Also noted is concurrent ALL ossification from severe ankylosing spondylitis.* **(Right)** *Axial bone CT reveals bulky "upside-down T" PLL ossification that severely narrows the central canal at C2. There is also mild concurrent ALL ossification that is typical of ankylosing spondylitis.*





(Left) *Sagittal T2WI MR demonstrates large, flowing OPLL severely compressing the thoracic cord. Note the associated ossification of ligamentum flavum.* **(Right)** *Sagittal T1WI MR in continuous type OPLL of the thoracic spine shows flowing low signal mass ventral to the severely compressed cord. The patient was severely myelopathic. Note ossification of ligamentum flavum.*

Ossification Ligamentum Flavum

KEY FACTS

TERMINOLOGY

- Ossification of ligamentum flavum (OLF), ossification of vertebral arch ligaments (OVAL), ligamentum flavum "pseudogout"
- Calcification/ossification of ligamentum flavum

IMAGING

- Linear thickening of ligamentum flavum with imaging characteristics similar to adjacent vertebral marrow ossification
- Curvilinear hyperdense thickening of ligamentum flavum
- Hypointense linear "mass" within ligamentum flavum ± cord myelomalacia, ↑ signal intensity 2° to compression
- CT imaging best modality for primary diagnosis, "lesion conspicuity"

TOP DIFFERENTIAL DIAGNOSES

- Facet arthrosis
- Meningioma

PATHOLOGY

- Ectopic bone formation within ligamentum flavum
- In majority of patients (idiopathic presentation), mechanism is unclear
  - Some cases clearly associated with metabolic or endocrine disease

CLINICAL ISSUES

- Incidental observation on imaging study ordered for other reasons
- Chronic thoracic myelopathy
- 4th → 6th decade
- Symptomatic presentation more common in Japanese, North African descent > > Caucasian > African American

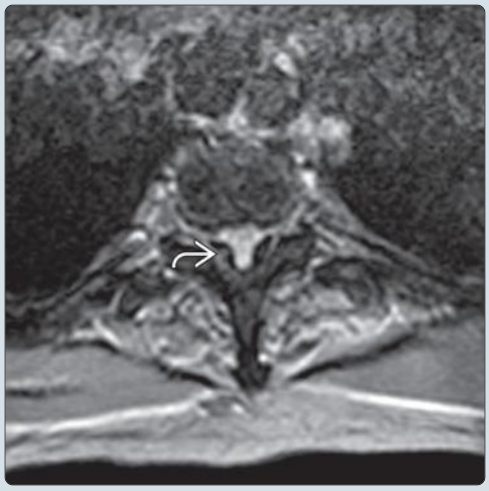
(Left) *Sagittal NECT shows that, at essentially every thoracic level, there are various patterns of ligamentum flavum ossification. These areas of ossification merge with the facets along their ventral margins.* **(Right)** *Axial NECT shows typical thoracic ligamentum flavum ossification as clumped and linear calcifications ventral and medial to the facets. They may have variable mass effect upon the thecal sac but can be a source of thoracic myelopathy.*





(Left) *Sagittal T2WI MR shows ossified ligamentum flava as low signal at every thoracic level, merging with the low signal of the cortex of the facet joints.* **(Right)** *Axial T2* GRE MR demonstrates the ossified ligaments as low signal with compression of the dorsal thecal sac producing central canal stenosis. This should be distinguished from congenital stenosis with shortened pedicles.*





TERMINOLOGY

Synonyms

- Ossification of ligamentum flavum (OLF), ossification of vertebral arch ligaments (OVAL), ligamentum flavum "pseudogout"

Definitions

- Calcification &/or ossification of ligamentum flavum

IMAGING

General Features

- Best diagnostic clue
  - Linear thickening of ligamentum flavum with imaging characteristics similar to adjacent vertebral marrow ossification
- Location
  - Ligamentum flavum (ventromedial to facets and lamina, dorsal to thecal sac)
  - Lower thoracic > > cervical, upper thoracic, lumbar
- Size
  - Multi- or single level, small or large
- Morphology
  - Linear ossification localized within ligamentum flavum
  - Symmetric, bilateral > unilateral

Radiographic Findings

- Radiography
  - Thin curvilinear "calcification" ventral to lamina; difficult to appreciate on plain radiography

CT Findings

- NECT
  - Curvilinear hyperdense thickening of ligamentum flavum
  - CT best shows ossification, but poor for determining cord status
- Bone CT
  - Hyperdense nodular or linear ossification within ligamentum flavum
  - "V" shape on axial images

MR Findings

- T1WI
  - Hypo- to hyperintense linear "mass" within ligamentum flavum ± cord compression
    - "V" shape on axial images
    - Thinner lesions usually hypointense 2° to minimal marrow relative to cortical bone
    - Thicker lesions more likely to mimic marrow signal intensity (isointense → hyperintense) of adjacent vertebral body
- T2WI
  - Hypointense linear "mass" within ligamentum flavum ± cord myelomalacia, ↑ signal intensity 2° to compression
  - Large lesions may mimic marrow signal intensity of adjacent vertebra but usually hypointense
- T2* GRE
  - Hypointense ligamentum flavum thickening
  - GRE always overemphasizes degree of canal narrowing 2° to susceptibility artifact

Nonvascular Interventions

- Myelography
  - Thin curvilinear "calcification" ventral to lamina
  - ± dorsal thecal sac impression
    - Usually minimal; rare cases show significant compression → myelographic block

Imaging Recommendations

- CT imaging best modality for primary diagnosis, "lesion conspicuity"
  - Sagittal reformats excellent for determining longitudinal extent
- Multiplanar MR imaging to determine relationship to and effect on regional soft tissues
  - Sagittal T1WI, T2WI evaluate longitudinal extent of ligamentous ossification, degree of cord compression
  - Axial T1WI, T2WI evaluate canal caliber

DIFFERENTIAL DIAGNOSIS

Facet Arthrosis

- Substantially more common than OLF
- Facet joint space shows characteristic degenerative changes
  - Centered in joint space rather than ligamentum flavum
- Commonly coexists with OLF as unrelated finding

Meningioma

- "En plaque" calcified meningioma mimics OLF when small
  - Arises from dural surface; centered in spinal canal, not ligamentum flavum
  - Almost always enhance; look for dural "tail"

PATHOLOGY

General Features

- Etiology
  - In majority of patients (idiopathic presentation), mechanism is unclear
    - Some cases clearly associated with metabolic or endocrine disease
  - Pathogenesis probably related to hydroxyapatite (HAD) or calcium pyrophosphate deposition (CPPD) in ligament → calcification, ossification + genetic predilection
- Genetics
  - Collagen 6A1 gene single nucleotide polymorphisms shown in Han Chinese population with OLF
- Associated abnormalities
  - Variably coexists with DISH, OPLL
  - Rare association with Bartter syndrome, hypomagnesemia
- Calcium pyrophosphate dihydrate crystal deposition disease (CPPD/CDD) noted in variety of vertebral structures other than ligamentum flavum
  - Intervertebral disc, anterior longitudinal ligament (ALL), posterior longitudinal ligament (PLL), interspinous/supraspinous ligaments, apophyseal and SI joints
- Debated whether ossified ligamentum flavum is different from "calcification" of ligamentum flavum 2° to crystal deposition

Gross Pathologic & Surgical Features

- Ectopic bone formation within ligamentum flavum
- Infiltration of fibroblast mesenchymal cells into ligamentum flavum
- Calcification may involve adjacent dura

Microscopic Features

- Ligamentous tissue hyperplasia, cell proliferation → endochondral ossification of ligamentum flavum
- Polarized-light microscopy: Characteristic rod-shaped, birefringent crystals

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Incidental observation on imaging study ordered for other reasons
  - Chronic thoracic myelopathy
    - Ambulation difficulty, weakness, back pain, and lower extremity paresthesias
    - Posterior column findings usually 1st followed by progressive spastic paraparesis
  - Back pain ± radicular symptoms
- Clinical profile
  - Symptomatic patients present similarly to those with other cause of myelopathy

Demographics

- Age
  - 4th-6th decade
- Gender
  - M > F
- Ethnicity
  - Symptomatic presentation more common in Japanese, North African descent > > Caucasian > African American
- Epidemiology
  - Probably more common than currently appreciated
    - Most cases incidental, asymptomatic, and unrecognized on imaging

Natural History & Prognosis

- Most patients remain asymptomatic
- Minority of patients manifest progressive myelopathy
  - Symptoms may improve or stabilize following surgery
  - Persistent postoperative spasticity suggests irreversible cord injury
- Duration of preoperative symptoms = most important predictor of long-term postoperative outcome

Treatment

- Conservative observation in asymptomatic, mild cases
- Posterior decompression (laminectomy or laminoplasty), ligamentum flavum resection in symptomatic cases

DIAGNOSTIC CHECKLIST

Consider

- Consider OLF in patient presenting with progressive myelopathy, particularly if Japanese or North African descent
- Important to exclude coexistent OPLL, DISH, other myelopathic lesions

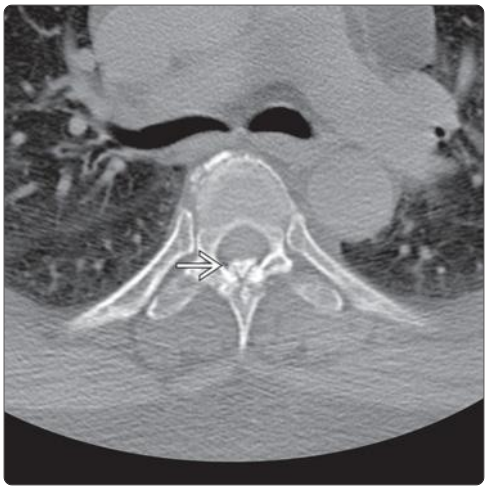
Image Interpretation Pearls

- Distinguish from facet arthropathy by location in ligamentum flavum, observation above or below facet joint level

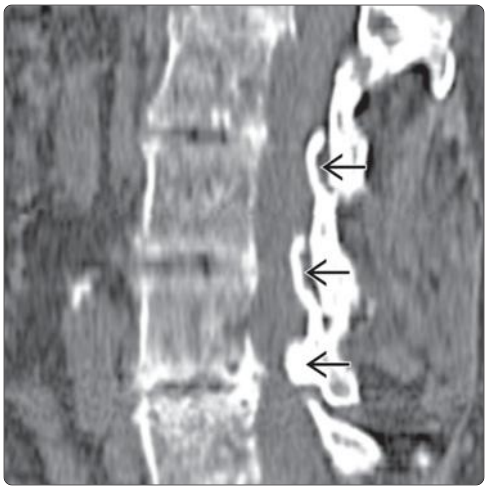
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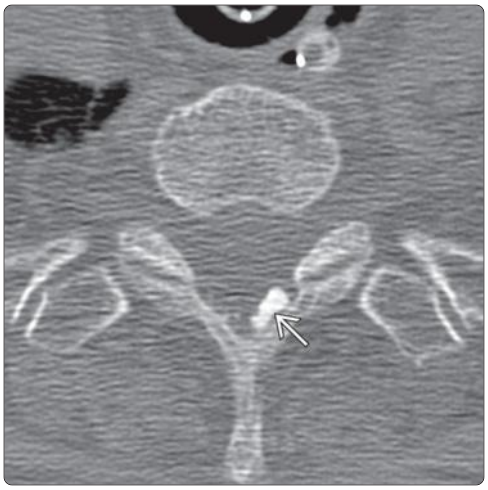
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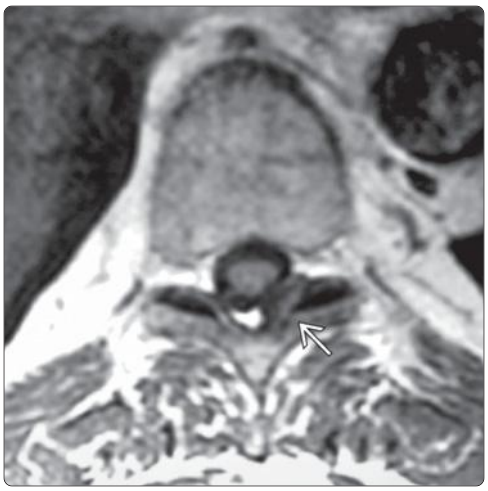


(Left) *Sagittal NECT shows focal calcification of the ligamentum flavum involving 2 levels of the midthoracic spine. There is no significant stenosis from these deposits.* **(Right)** *Axial NECT shows focal midline calcification of the ligamentum flavum involving the midthoracic spine. This is an incidental finding as there is no mass effect on the thecal sac and no metabolic abnormality (such as CPPD).*





(Left) *Sagittal NECT shows dorsal curvilinear ossification within ligamentum flavum producing mild central canal narrowing. Thoracic ossification of ligamentum flavum is much more common.* **(Right)** *Axial bone CT obtained for trauma shows focal nodular ossification in the ligamentum flavum, an incidental asymptomatic finding.*





(Left) *Axial T1WI MR reveals focal ossification within the ligamentum flavum as a low signal intensity mass contiguous with the left facet.* **(Right)** *Axial T2WI MR depicts unilateral hypointense ossification within the left ligamentum flavum producing mild narrowing of the lateral spinal canal and neural foramen.*

Periodontoid Pseudotumor

KEY FACTS

TERMINOLOGY

- Calcium pyrophosphate dihydrate deposition disease (CPPD); articular chondrocalcinosis
- Crowned dens syndrome: Fever with cervicooccipital junction pain, stiffness with associated calcification of transverse ligament and nonenhancing retroodontoid soft tissue; associated with CPPD or hydroxyapatite deposition
- Pseudogout: Arthritis caused by CPPD crystal inflammation with symptoms like gout

IMAGING

- Calcification of transverse ligament on CT
- T1WI: Iso to low signal mass posterior to odontoid
- T2WI: Low signal mass posterior to odontoid
- T1WI C+: May show peripheral enhancement

TOP DIFFERENTIAL DIAGNOSES

- Rheumatoid arthritis
- Osteoarthritis
- OPLL
- Osteomyelitis
- Meningioma
- Metastatic disease

PATHOLOGY

- Nodular deposits of birefringent, rhomboid crystals

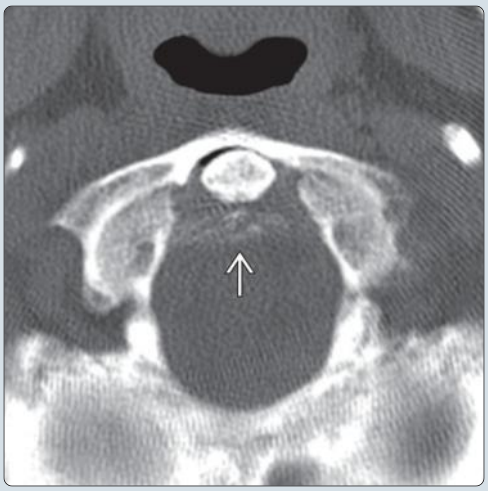
CLINICAL ISSUES

- Age 60 or greater, prevalence of CPPD deposition is 34%
- Crowned dens: Inflammatory indicators + (fever, CRP, elevated WBC count)

DIAGNOSTIC CHECKLIST

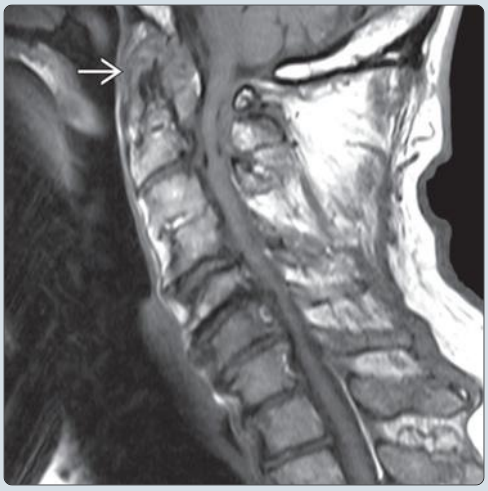
- Calcification of transverse ligament common in elderly, but symptomatic CPPD very uncommon

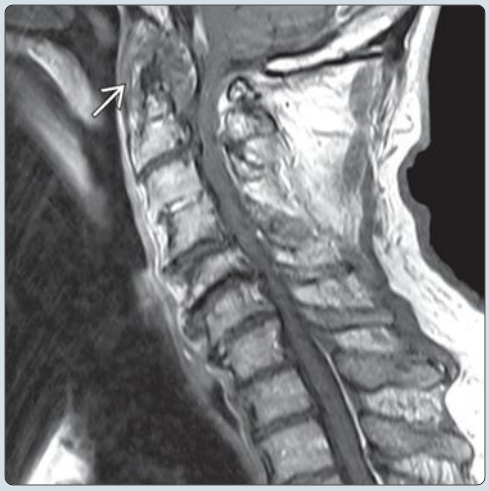
(Left) *Axial NECT shows amorphous calcification within the transverse ligament at the C1-C2 junction. There is also osteoarthritic degenerative change at the anterior C1-C2 joint with vacuum phenomenon.* **(Right)** *Sagittal T2WI MR shows low signal pseudotumor posterior to odontoid process, with moderate mass effect upon the cervical cord. Low signal mass extends over superior margin of odontoid. Note the associated calcification of ligamentum flavum.*





(Left) *Sagittal T1WI MR shows prominent soft tissue surrounding the odontoid process, with mass effect upon ventral thecal sac and cord at C1 level. There is severe multilevel disc degeneration and canal stenosis.* **(Right)** *Sagittal T1WI C+ MR shows mild peripheral enhancement of the CPPD pseudotumor involving the periodontal soft tissues. Low T2 signal and peripheral enhancement are typical of this soft tissue.*





TERMINOLOGY

Synonyms

- Calcium pyrophosphate dihydrate deposition disease (CPPD); articular chondrocalcinosis
- Pseudogout; crystal-associated arthropathy
- Crowned dens syndrome
- Calcification of transverse ligament
- Crystal deposition at craniovertebral junction

Definitions

- Crowned dens syndrome: Fever with cervicooccipital junction pain, stiffness with associated calcification of transverse ligament and nonenhancing retroodontoid soft tissue; associated with CPPD or hydroxyapatite deposition
- Pseudogout: Arthritis caused by CPPD crystal inflammation with symptoms like gout
- Tophaceous pseudogout: Large accumulation of material giving pattern of soft tissue tumor with mass effect/cord compression

IMAGING

General Features

- Best diagnostic clue
  - Calcification of transverse ligament on CT
- Location
  - Retroodontoid soft tissues and transverse ligament
- Size
  - Few cms
- Morphology
  - Rounded soft tissue posterior to odontoid with calcification

CT Findings

- NECT
  - Curvilinear, stippled, or mixed pattern of calcification posterior to odontoid
  - Variable soft tissue mass with effacement of ventral thecal sac
  - Variable cystic or erosive changes within adjacent bone

MR Findings

- T1WI
  - Iso to low signal mass posterior to odontoid
- T2WI
  - Low signal mass posterior to odontoid
- T1WI C+
  - May show peripheral enhancement
  - Generally does not show homogeneous soft tissue enhancement

Imaging Recommendations

- Best imaging tool
  - CT for calcification of transverse ligament
  - MR for soft tissue component and mass effect upon the cervical cord
- Protocol advice
  - T1WI C+ should not show homogeneous enhancement of the retroodontoid soft tissue in pseudogout

DIFFERENTIAL DIAGNOSIS

Rheumatoid Arthritis

- Erosion of odontoid
- Enhancing soft tissue pannus dorsal to irregular odontoid
- Cranial settling seen with RA and not CPPD

Osteoarthritis

- Bony sclerosis and joint space loss
- Affects anterior C1-C2 articulation and lateral C1-C2 joints

OPLL

- Focal low signal on all pulse sequences, flowing or segmented
- CT to show calcification and bow tie pattern of ossification

Osteomyelitis

- Destruction of odontoid with enhancing soft tissue from phlegmon
- Focal abscess may show peripheral enhancement
- Adjacent soft tissue T2 hyperintensity from inflammatory response

Meningioma

- Intradural extramedullary lesion
- Diffuse enhancement with dural base

Metastatic Disease

- Soft tissue mass with bone destruction
- Calcification in mass very uncommon

Synovial Cyst

- T2 hyperintense round mass arising from posterior atlantodental joint
- Peripheral enhancement

Calcific Retropharyngeal Tendinitis

- Calcific mass ventral to odontoid/C2 body; no retrodental component

Os Odontoideum

- Well-corticated bony density with truncated odontoid peg

Clival Chordoma

- Enhancing mass secondarily involving upper cervical spine
- No matrix, purely lytic lesion

Osteoblastoma

- Expansile mass involving posterior elements
- C2 body involvement rare

PATHOLOGY

Gross Pathologic & Surgical Features

- **CPPD**: Firm, yellowish brown tissue at surgery involving transverse ligament
  - Nodular deposits of birefringent, rhomboid crystals in soft tissue on H&E sections
    - Decalcification process for bone samples dissolves CPPD crystals
  - 90% of calcification at CVJ occurs posterior to odontoid
    - Calcification can develop anywhere around odontoid, including synovial membrane, articular capsule, and alar ligament

- **Hydroxyapatite deposition disease** (HDD): Also affects cervical spine
  - Calcific tendinitis of longus colli
    - 30-60 years of age
    - Inflammation at anterior C2 body level
  - Also can give crowned dens syndrome

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Heterogeneous clinical presentations ranging from incidental finding to mass with myelopathy; neck pain
  - Crowned dens
    - Suboccipital to posterior neck pain
    - Markedly restricted neck motion, with no rotation
    - Positive inflammatory indicators, such as fever, C-reactive protein, slightly elevated WBC count
  - Myelopathy with pseudotumor presentation from mass effect on cervical cord

Demographics

- Age
  - Age 60 or greater, prevalence of CPPD deposition is 34%
  - Age 80 or greater, prevalence of CPPD deposition is 49%
- Gender
  - Crowned dens syndrome: F > M, > 60 years

Natural History & Prognosis

- Most cases asymptomatic, finding is incidental
  - 7% of adults by age 70
  - 1 case per 1,000 persons
- CPPD associated with many other metabolic and inflammatory diseases
  - Hemochromatosis
  - Wilson disease
  - Gout
  - Diabetes
  - Rheumatoid arthritis
  - Hyperparathyroidism
  - Hypophosphatemia
- Joints most commonly involved include knee, hand, hip, pubic symphysis
- Less commonly involved regions include shoulder, elbow, spine, TMJ

Treatment

- Conservative treatment for inflammatory symptoms of crowned dens, including NSAIDs, steroids, or both; physical therapy
- Pain or cord compression from nonenhancing mass may necessitate surgery
  - Transoral-transpharyngeal approach with resection of CPPD mass
    - Variable need dorsal fixation (occipital-C2)

DIAGNOSTIC CHECKLIST

Consider

- Associated symptomatic involvement of ligamentum flavum with CPPD may present with cord compression and myelopathy
- Neuropathologist should be informed of CPPD diagnosis before specimen obtained since typical decalcification processing of tissue will dissolve CPPD crystals

Image Interpretation Pearls

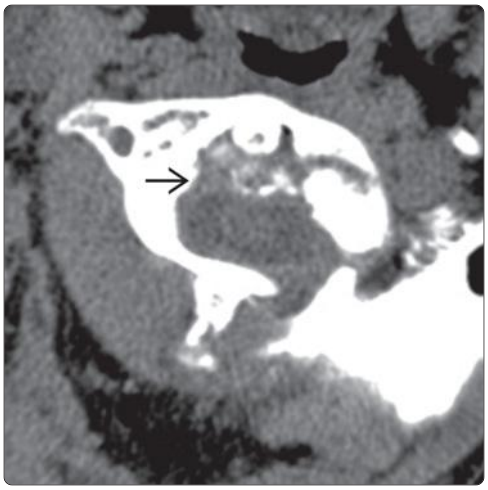
- T2 low signal retrodental mass is key imaging finding

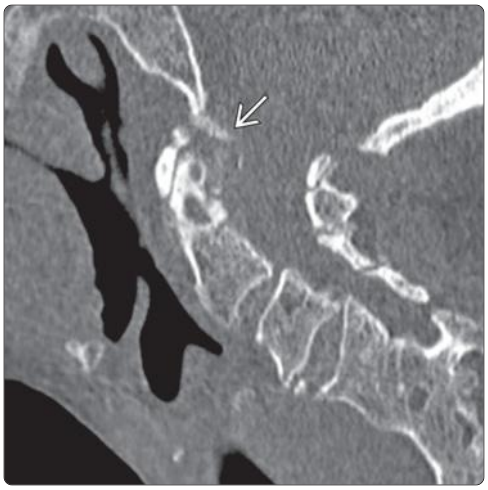
Reporting Tips

- Calcification of transverse ligament is common in elderly, but symptomatic CPPD is very uncommon

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(Left) *Axial NECT shows extensive amorphous calcification involving the retrodental soft tissue ⇨ with mild mass effect upon the ventral thecal sac. This pattern may be asymptomatic but may also be seen with inflammatory or mass-like symptoms.* **(Right)** *Sagittal NECT shows ill-defined calcification posterior to the odontoid process ➡ in this case of CPPD. Note the cystic degenerative change of the odontoid process and osteoarthritic narrowing of the C1-C2 articulation.*





(Left) *Sagittal T1WI MR shows low signal pseudotumor ➡ dorsal to the odontoid, with compression of the anterior thecal sac and ventral cord. There is also dorsal compression from an anteriorly displaced posterior arch of C1.* **(Right)** *Sagittal STIR MR shows CPPD arthropathy at the C1-C2 junction with pseudopannus compressing the cervical cord with myelomalacia ⇨ associated with anterior subluxation of C1. Note calcification within ligamentum flavum as associated finding ➡.*





(Left) *In this differential diagnosis example, sagittal T2WI MR shows rheumatoid cranial settling with upward translocation of odontoid into foramen magnum due to ligamentous laxity.* **(Right)** *In this differential diagnosis, sagittal T2WI MR in C1-C2 osteomyelitis demonstrates phlegmon at C1-C2 articulation ➡ with T2 hyperintensity. This is distinctly different from the low signal seen with CPPD.*

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PART III
SECTION 2

Spondylolisthesis and Spondylolysis



Spondylolisthesis	**508**
Spondylolysis	**512**
Instability	**516**

Spondylolisthesis

KEY FACTS

TERMINOLOGY

- Anterolisthesis: Anterior displacement of vertebral body relative to one below
- Retrolisthesis: Posterior displacement of vertebral body relative to one below

IMAGING

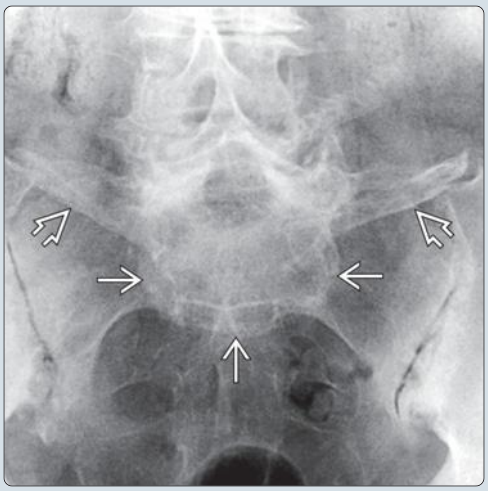
- Lateral flexion and extension to evaluate for instability
  - "Napoleon's hat" sign on AP plain film
  - Instability uncommon in degenerative listhesis
    - 90% of normal volunteers show 1-3 mm translation on flexion-extension radiographs
- Spondylolysis may be difficult to identify on MR
  - T1-weighted sagittal images critical
  - CT for definitive diagnosis of subtle fracture

PATHOLOGY

- Degenerative (DS)
  - Degenerative retrolisthesis associated with disc degeneration
  - Sagittal oriented facets more likely to have DS
- Spondylolysis (isthmic)
  - Bilateral in 80%
- Postsurgical: Loss of posterior element stability
- Dysplastic: Small L5 body leading to pars lysis
- Trauma: Severe to produce vertebral body displacement
- Pathologic: Underlying tumor with instability

CLINICAL ISSUES

- **9.2% overall complication rate for treatment of spondylolisthesis**
  - Complications related to higher grade spondylolisthesis, DS > isthmic, older age (> 65)
- DS + stenosis treated surgically show greater improvement in pain and function over 4 years compared to nonsurgical treatment





(Left) *Anteroposterior radiograph shows "Napoleon's hat" sign. The hat is inverted, with the crown ➡ representing the anterior cortex of vertebral body, and the brim ➡ being the transverse processes.* **(Right)** *Sagittal T1WI MR shows postoperative spondylolisthesis following lumbar laminectomy ➡. There is advanced degenerative changes of the L4-L5 intervertebral disc space with anterior subluxation of L4 on L5. Note the acute superior endplate compression fracture of L1 ➡.*





(Left) *Sagittal T2WI MR shows grade I spondylolisthesis of L4 on L5 ➡, with intervertebral disc degeneration and associated fatty endplate change. There is also degeneration of the interspinous ligament ➡.* **(Right)** *Sagittal T2WI MR shows a grade I spondylolisthesis of L4 on L5 with resultant severe foraminal stenosis ➡.*

TERMINOLOGY

Synonyms

- Degenerative spondylolisthesis (DS)

Definitions

- Displacement of vertebral body, described relative to inferior vertebra
  - Anterolisthesis: Anterior displacement of vertebral body relative to one below
  - Retrolisthesis: Posterior displacement of vertebral body relative to one below
  - Spondyloptosis: Vertebral body displaced completely anteriorly, with inferior displacement to level of one below
- Degenerative spondylolisthesis describes anterior slippage of vertebral body on another in presence of intact neural arch

IMAGING

General Features

- Best diagnostic clue
  - Displacement of posterior cortex of vertebral body on lateral radiograph
- Location
  - Most common in lower lumbar spine

Radiographic Findings

- Radiography
  - Evaluate percentage or grade of listhesis on neutral, flexion, and extension laterally
  - Lateral flexion and extension views to evaluate for instability
    - Instability uncommon in degenerative listhesis
    - Instability may be present with spondylolysis
    - Instability almost always seen with traumatic spondylolisthesis
  - "Napoleon's hat" sign
    - Severe spondylolisthesis results in focal kyphosis
    - Kyphotic, subluxed vertebra resembles upside-down curved hat on AP radiographs
  - Evaluate for fracture, spondylolysis
  - Degenerative type shows degenerative disc disease and facet osteoarthritis

CT Findings

- Bone CT
  - Evaluate for pars interarticularis defects, fractures, spinal stenosis

MR Findings

- Spondylolysis difficult to see on MR
- Obtain stack axial images as well as axial images angled through intervertebral disc
- Sagittal, axial T1WI, and T2WI to evaluate spinal stenosis and neural foraminal encroachment
- Look for hematoma as sign of acute traumatic spondylolysis

Nuclear Medicine Findings

- Bone scan
  - Increased uptake at acute or subacute pars defects
  - Chronic pars defects photopenic
  - Increased uptake at degenerated facet joints and discs

Imaging Recommendations

- Best imaging tool
  - CT scan
- Protocol advice
  - Thin-slice CT with sagittal reformations

DIFFERENTIAL DIAGNOSIS

Physiologic Motion (Pseudo-Subluxation)

- Slight subluxations normal in young patients

Displaced Vertebral Body Fracture

- Fracture is often best seen on CT

PATHOLOGY

General Features

- Etiology
  - Degenerative
    - Anterolisthesis may be related to alignment of facet joints
    - More sagittally oriented L4-L5 facets are more likely to have degenerative listhesis
    - Etiology-based classification describes degenerative spondylolisthesis as either developmental or acquired
      - Degenerative is subtype of acquired
      - Primary is typical of middle-aged women with spinal stenosis
      - Secondary relates to predisposing factors, such as prior surgical fusion
    - Degenerative retrolisthesis associated with disc degeneration
      - Decreased disc height with telescoping of adjacent articular processes
      - Most frequent in upper lumbar spine
  - Postsurgical
    - Destabilization of spine at wide laminectomy
    - Late complication above or below fusion with adjacent segment accelerated degeneration
  - Dysplastic
    - Small L5 body leading to altered biomechanics and pars defects
  - Spondylolysis (isthmic)
    - Acquired abnormality becoming symptomatic in adulthood as disc degeneration progresses
    - Progression of adult isthmic listhesis is uncommon
    - Bilateral in 80%
  - Trauma
    - At site other than pars interarticularis
    - Shear injury
    - Fractures of posterior elements
  - Pathologic
    - Tumor
      - Bone destruction with lysis of posterior elements
    - Infection
      - Bone destruction involving endplates and less likely posterior elements
      - Late instability due to altered biomechanics with body collapse
    - Neuropathic arthropathy

- Associated abnormalities
  - Scoliosis may develop

Staging, Grading, & Classification

- Meyerding classification
  - Grade I: < 25% displacement of vertebral body
  - Grade II: 25-50% displacement of vertebral body
  - Grade III: 50-75% displacement of vertebral body
  - Grade IV: 75-100% displacement of vertebral body
  - Grade V: Spondyloptosis
- Progression of < 10% is difficult to define with this classification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, radiculopathy
  - Degenerative listhesis presents as lumbar stenosis
- Other signs/symptoms
  - Neurogenic claudication (leg pain with walking)
    - Pain predominant symptom
    - Numbness and weakness less common

Demographics

- Age
  - Degenerative spondylolisthesis > 40 years old
  - Isthmic spondylolisthesis 4-6% of general population
- Gender
  - Degenerative more common in women (4:1)
    - Thought to be related to ligamentous laxity
  - Isthmic listhesis: M > F
- Epidemiology
  - > 60% prevalence of isthmic listhesis in certain Eskimo communities

Natural History & Prognosis

- Degenerative
  - Not well characterized
  - 10% of degenerative listhesis show clinical deterioration
  - Progressive slip shows no correlation with clinical deterioration
- Increased pelvic incidence angle shows increase of slippage
  - Increased shear forces across affected segment with resultant hyperlordosis

Treatment

- **9.2% overall complication rate for treatment of spondylolisthesis**
  - Complications related to higher grade spondylolisthesis, DS > isthmic, older age (> 65)
- DS + stenosis treated surgically show greater improvement in pain and function over 4 years compared to nonsurgical treatment
  - Spinal stenosis + degenerative listhesis has better clinical outcome when decompression combined with arthrodesis
    - Role of augmented fusion with pedicle screws is controversial
  - Poor surgical outcomes in middle-aged men who smoke + compensation or litigation claims at time of surgical treatment
  - Increased fusion with instrumentation and anterior grafting does not improve clinical outcome compared to instrumented posterolateral grafting

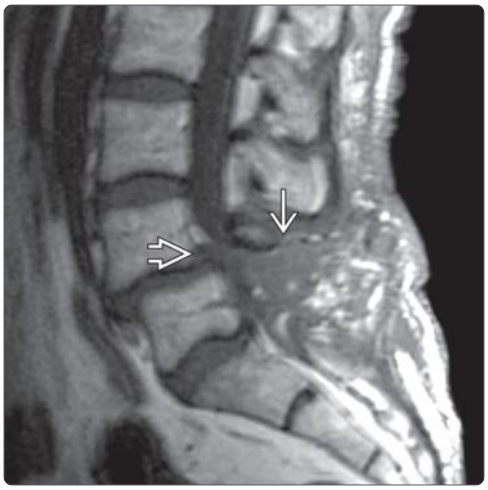
DIAGNOSTIC CHECKLIST

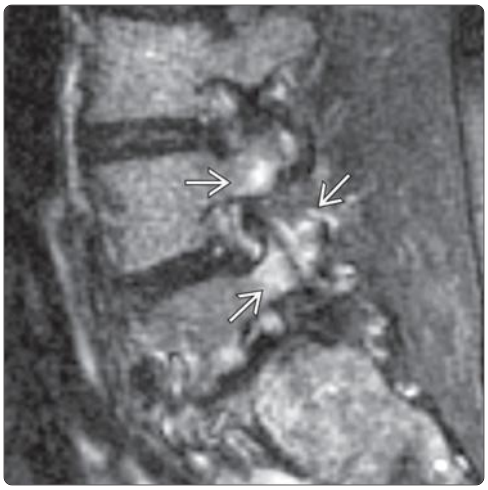
Image Interpretation Pearls

- 90% of normal volunteers show 1-3 mm translation on flexion-extension lateral radiographs
  - > 4 mm translation thus abnormal
- Discordance of imaging and scintigraphic findings not uncommon, with acute pars defects difficult to detect on plain radiographs, but metabolically active

SELECTED REFERENCES

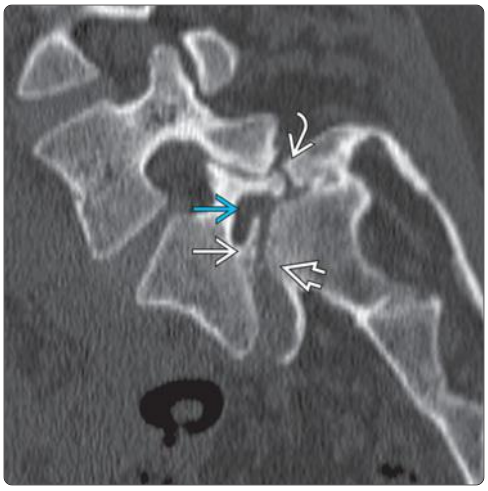
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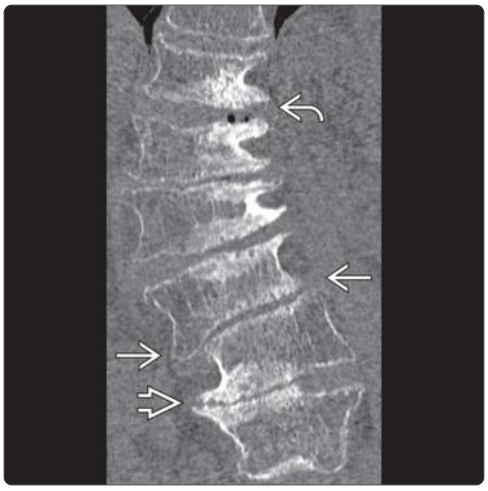


(Left) *Sagittal T1WI MR of mild spondylolisthesis following lower lumbar laminectomy shows mild anterolisthesis of L4 on L5 with evidence of prior L4 and L5 laminectomy.* **(Right)** *Sagittal STIR MR shows hyperintense marrow edema adjacent to L4-L5 facet, L4 and L5 pedicles. Type I pedicle marrow signal can be seen with degenerative facet arthropathy and may reflect altered biomechanical loading of the facet joints and neural arch in the setting of disc space narrowing and spondylolisthesis.*





(Left) *Sagittal T2WI MR shows severe multilevel cervical spondylosis with anterolisthesis of C3 on C4 with canal stenosis, cord compression. Cord myelomalacia due to repetitive trauma from the stenosis is seen as T2 cord hyperintensity.* **(Right)** *Sagittal NECT (bone window) shows that the posterior margin of the L5 vertebral body is almost at same position as the anterior margin of S1. Pars defect is readily apparent, as is the severe neural foraminal stenosis.*





(Left) *Coronal NECT shows severe multilevel disc degeneration and lateral listhesis. Note the increased bone production along the stress lines of the right and left side components.* **(Right)** *Sagittal T1WI MR shows L5/S1 spondylolisthesis in a patient run over by a truck. Mixed low and high signal hematoma is present behind L5 vertebral body and between posterior elements L5 and S1.*

Spondylolysis

KEY FACTS

TERMINOLOGY

- Defects in pars interarticularis (PI) thought to result from repetitive stress injury
  - Pars is junction of pedicle, lamina, and facet

IMAGING

- Most common at L5: 80-90%
- Discontinuity in neck of "Scotty dog" on oblique plain films
- Elongation of spinal canal at level of pars defects on axial imaging
- "Incomplete ring" sign on axial imaging

TOP DIFFERENTIAL DIAGNOSES

- Acute traumatic fracture of posterior elements
- Facet arthropathy with marrow edema
- Septic facet with marrow edema
- Bone tumor with marrow edema
- Pedicle stress fracture
- Congenital defect with pars cleft

PATHOLOGY

- Repeated microfractures of PI lead to fatigue fracture

CLINICAL ISSUES

- 6-8% in general population
- 10-20 years old is typical age range
- Symptoms of chronic low back pain
- Conservative measures in grade 1-2 spondylolisthesis
- Wide variety of practice patterns for treatment
  - 50% of surgeons agreed on surgical treatment
  - No consensus on optimum surgical strategy

DIAGNOSTIC CHECKLIST

- Look for integrity of PI on sagittal MR and pars/pedicle marrow edema

(Left) *Lateral graphic shows chronic L5 spondylolisthesis and spondylolysis with defect in the pars interarticularis (PI) and resultant foraminal stenosis.* **(Right)** *Coronal oblique radiograph shows a thin lucency in the right L5 PI with adjacent bony sclerosis.*





(Left) *Lateral radiography shows a defect and mild angulation in L5 PI. There is disc height loss at L5-S1 with associated sclerotic endplate changes.* **(Right)** *Sagittal bone CT reformation shows L5 pars defect without anterolisthesis.*





TERMINOLOGY

Synonyms

- Isthmic spondylolysis (IS)

Definitions

- Unilateral or bilateral defects in pars interarticularis (PI) thought to result from repetitive stress injury
  - Pars is junction of pedicle, lamina, and facet

IMAGING

General Features

- Best diagnostic clue
  - Bony defect or cortical discontinuity in PI
- Location
  - Most common at L5: 80-90%
  - L4 2nd most common
  - 10-15% unilateral defects
    - Unilateral healing or union of fractures that were initially bilateral
- Morphology
  - Horizontal orientation on axial imaging

Radiographic Findings

- Radiography
  - Radiolucent band in PI on oblique views of lumbar spine
    - Discontinuity in neck of "Scotty dog"
  - Oblique lucency at base of laminae on lateral view
  - Contralateral pedicle and lamina hypertrophy and sclerosis
    - Unilateral spondylolysis
    - Best seen on frontal lumbar spine
  - Variable degree of spondylolisthesis
  - Disc height loss at level below spondylolysis
  - Secondary degenerative sclerotic endplate changes and spurring

CT Findings

- Bone CT
  - "Incomplete ring" sign on axial imaging
    - Disruption of ring formed by vertebral body and arch
  - May simulate facet joints
    - "Extra facet" sign
    - Horizontal vs. oblique orientation
    - Irregular vs. smooth bony cortex
  - Pars defects well seen on oblique reformation
  - Variable spondylolisthesis and foraminal narrowing on sagittal reformation
    - Disc space loss
    - Degenerative endplate changes
  - Canal may be widened even without spondylolisthesis

MR Findings

- T1WI
  - Focally decreased signal in PI on sagittal and axial imaging
  - Similar appearance on T2WI
- T2WI
  - Hyperintensity may be present within pars defects and adjacent marrow of pedicle
- STIR
  - Marrow edema adjacent to pars defects and pedicle
- Elongation of spinal canal
- Horizontal configuration of affected neural foramina on sagittal imaging
  - Anterolisthesis
  - Disc height loss
  - Loss of fat surrounding exiting nerve roots
- MR sensitivity: 57-86%
- MR specificity: 81-82%

Nonvascular Interventions

- Myelography
  - Neural foraminal narrowing with anterolisthesis and disc height loss

Nuclear Medicine Findings

- Bone scan
  - Foci of increased radiotracer uptake in posterior elements
    - Suggests bone healing
    - SPECT is better, but still relatively high false-positives and false-negatives

Imaging Recommendations

- Best imaging tool
  - Axial thin-section CT with bone algorithm
    - Sagittal and oblique reformation

DIFFERENTIAL DIAGNOSIS

Acute Traumatic Fracture of Posterior Elements

- History of acute trauma
- Sharp noncorticated margins

Facet Arthropathy With Marrow Edema

- Hypertrophied facet and ligamentum flavum
- Edema centered on facet and dorsal facet soft tissues

Septic Facet With Marrow Edema

- Clinical findings of fever, elevated WBC, and sedimentation rate
- Extensive adjacent soft-tissue inflammation

Bone Tumor With Marrow Edema

- Osteoid osteoma typical
  - Edema may be extensive with small sclerotic tumor nidus
- Metastatic disease
  - Multiple lesions
  - Focal and round on T1W images

Pedicle Stress Fracture

- Stress reaction may show adjacent marrow edema
- CT to define exact location of stress reaction or fracture

Congenital Defect With Pars Cleft

- Hypoplasia of spinal accessory process
- Rotation of spinous process contralateral to spondylolysis
- Prominent ipsilateral lamina

PATHOLOGY

General Features

- Etiology

- Participation in gymnastics, weightlifting, wrestling, football, etc.
  - Starting at young age
  - Training > 15 hours per week
  - Repetitive exposure to rotation, flexion-extension, and hyperextension
- Repeated microfractures of PI lead to fatigue fracture
  - Usually occurs during growth spurt
- Genetics
- Predisposing familial conditions to spondylolysis
  - Marfan syndrome
  - Osteogenesis imperfecta
  - Osteopetrosis
- Associated abnormalities
- Scoliosis
- 50% with spondylolisthesis
- Spina bifida occulta
- Scheuermann disease
- Thought to result from combination of "weak" bone and repetitive trauma during growth spurt

Microscopic Features

- New bone and cartilaginous matrix if healing present
- Fibrocartilaginous tissue within pars defects if not healed

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
- Commonly asymptomatic
- Chronic low back pain, particularly in children and adolescents
  - Usually presents during adolescent growth spurt
- Other symptoms
  - Back spasm
  - Hamstring tightness
  - Radiculopathy and cauda equina syndrome
  - Gait disturbance
- Clinical profile
- Pain exacerbated by rigorous activities
- 80% without symptoms

Demographics

- Age
- 10-20 years old
- Gender
- M:F = 2-3:1
- Ethnicity
- > 30-40% of Eskimos
- Higher incidence in Caucasian males: 6.4% vs. African American females: 1.1%
- Epidemiology
- 6-8% in general population
  - 22-44% in competitive athletes in diving, weightlifting, wrestling
- Prevalence of 4% at age 6
- Rare under age 5

Natural History & Prognosis

- Progressive spondylolisthesis occurs during skeletal immaturity
- More common in females
- Risk of progression: 3-28%
- Little slippage with horizontal sacrum
- Lumbosacral angle ≥ 100°
- Progressive spondylolisthesis with vertical sacrum
- Lumbosacral angle < 100°
- Progression from grade 1 spondylolisthesis: Superior vertebral body subluxed by 1/4 of vertebral body
- To grade 2: Subluxation by 1/2 of vertebral body
- To grade 3: Subluxation by 3/4 of vertebral body
- To grade 4: Subluxation by whole width of vertebral body

Treatment

- Conservative measures in patients with grade 1 and 2 spondylolisthesis
- Anti-inflammatory medications
- Epidural steroid injection
- Brace/physical therapy
- Surgical intervention
- Fail conservative treatment
- Have worsening slippage
- Preserve neural function and prevent instability
  - Posterior or posterolateral fusion
  - Stabilization with fusion or cast immobilization
  - Decompression
- Wide variety of practice patterns for treatment
- 50% of surgeons agreed on surgical treatment in 1 study
- No consensus on optimum surgical strategy
- Laminectomy/fusion
- TLIF/PLIF
- Anterior lumbar interbody fusion (ALIF) with posterior instrumentation

DIAGNOSTIC CHECKLIST

Consider

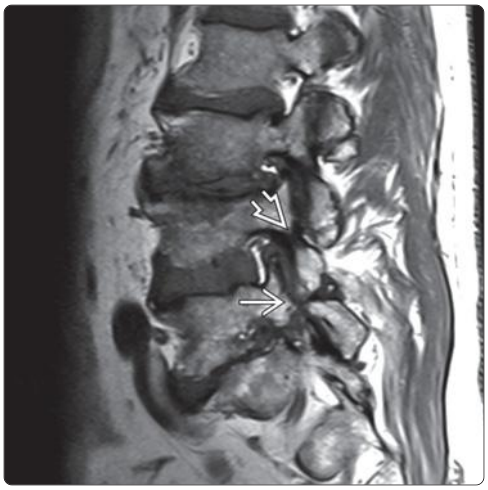
- L5-S1 IS stand-alone ALIF failure has been reported
- Unique axial load and shear stresses
- Supplementary posterior fixation recommended

Image Interpretation Pearls

- Look for integrity of PI on sagittal MR

SELECTED REFERENCES

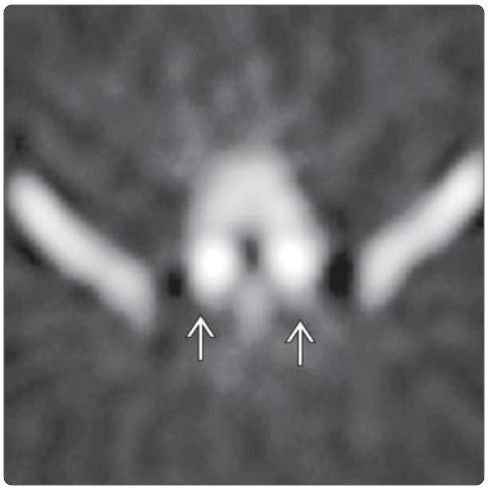
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(Left) *Sagittal T1WI MR shows chronic pars defects ➡ that maintain normal fatty marrow signal. There is severe foraminal stenosis at L5-S1. Note the normal low cortical bone signal at L4 from the intact pars ➡, which can mimic a pars defect.* **(Right)** *Sagittal T1WI MR shows a defect in L4 PI ➡. Fractures at this level and above are much less common than those at L5.*





(Left) *Stress reaction of the pars gives increased marrow signal on this sagittal T2WI MR within the pedicle ➡, superior articular process, and pars. This lesion went on to become a complete fracture.* **(Right)** *Axial SPECT bone scan in a patient with bilateral spondylolysis shows increased radiotracer uptake in bilateral L5 PI ➡.*





(Left) *Sagittal bone NECT in this patient with a congenital cleft involving pars shows a well-corticated large defect in the region of the L5 pars ➡, with hypertrophy and dysmorphism of the inferior articular facet of L4 ➡.* **(Right)** *Axial bone NECT in this patient with a congenital cleft in the right pars shows a large hypertrophied right lamina ➡ and rotated spinous process ➡.*

KEY FACTS

TERMINOLOGY

- Loss of spine motion segment stiffness, when applied force produces greater displacement than normal, with pain/deformity

IMAGING

- Deformity, which increases with motion and over time
- Various parameters used for degenerative instability by plain films
  - Dynamic slip > 3 mm in flexion/extension
  - Static slip of ≥ 4.5 mm
  - Angulation > 10-15° suggests need for surgical intervention
- Flexion/extension plain films best for definition of motion

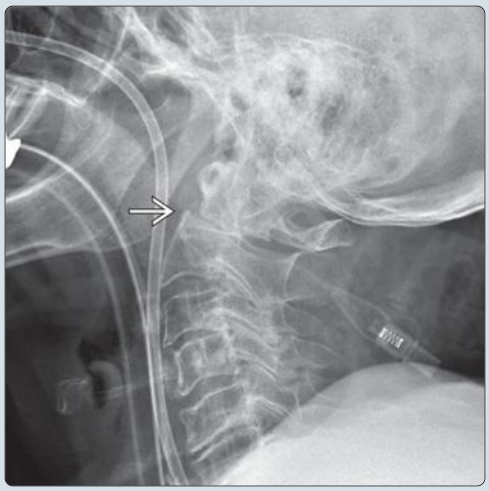
TOP DIFFERENTIAL DIAGNOSES

- Pseudoarthrosis
- Infection
  - Endplate destruction, disc T2 hyperintensity
- Tumor
  - Enhancing soft tissue mass
- Postoperative
  - Following multilevel laminectomy or facetectomy

PATHOLOGY

- Degenerative instabilities
  - Axial rotational
  - Translational; plain films show spondylolisthesis, traction spurs, vacuum phenomenon
  - Retrolisthesis; plain films show increased retrolisthesis with extension
  - Degenerative scoliosis
  - Post laminectomy; resection of 50% of bilateral facets alters segmental stiffness
  - Post fusion; altered biomechanics





(Left) *Lateral radiograph shows fracture through the base of the odontoid, which shows satisfactory alignment in neutral position.* **(Right)** *Lateral radiograph shows fracture through the base of the odontoid, which displaces posteriorly with slight extension. This patient underwent occiput to C3 fusion.*





(Left) *Sagittal T1WI MR shows a well-defined fracture line through the base of the odontoid in this type II fracture. Fracture was unstable on extension plain film examination.* **(Right)** *Sagittal STIR MR shows extensive prevertebral edema in this patient with type II odontoid fracture. There is cord edema and focal cord hemorrhage seen as slight hypointensity.*

TERMINOLOGY

Synonyms

- Spine instability (SI), segmental instability, abnormal spinal motion, degenerative instability

Definitions

- Loss of spine motion segment stiffness, when applied force produces greater displacement than normal, with pain/deformity
- Neoplastic instability: Loss of spinal integrity from neoplastic process associated with movement-related pain, symptomatic or progressive deformity, &/or neural compromise under physiologic loads

IMAGING

General Features

- Best diagnostic clue
  - Deformity that increases with motion and over time
- Location
  - Any spinal motion segment (comprised of 2 adjacent vertebrae, discs, and connecting spinal ligaments)
- Size
  - Displacement may vary from few mm to width of vertebral body
- Displacement of vertebral body with respect to adjacent body
- Stabilizing anatomic structures
  - Ligaments
    - Anterior longitudinal ligament
      - Resists hyperextension
    - Posterior longitudinal ligament
    - Intertransverse ligaments
      - Connect neighboring transverse processes
    - Interspinous ligaments
      - Resists hyperflexion
    - Facet capsule (strong)
    - Ligamentum flavum
  - Intervertebral disc
    - Main stabilizer of lumbar and thoracic spine
  - Muscular attachments
    - Both global (rectus and abdominal muscles) and local paraspinal muscle groups

Radiographic Findings

- Radiography
  - Various parameters used for degenerative instability by plain films
    - Dynamic slip > 3 mm in flexion/extension
    - Static slip ≥ 4.5 mm
    - Angulation > 10-15° suggests need for surgical intervention
  - Traction spurs
  - Vacuum phenomenon

Fluoroscopic Findings

- Increased motion with flexion/extension or translation

CT Findings

- NECT
  - Nonspecific findings of degenerative disc disease ± spondylolisthesis

MR Findings

- T1WI
  - Anterolisthesis, retrolisthesis, lateral translation
  - Nonspecific changes of degenerative disc disease
  - Controversial as to role in defining instability: Type I degenerative endplate changes
- T2WI
  - Loss of disc signal ± disc space height
- STIR
  - Type I endplate changes may be more evident on this sequence
- T1WI C+
  - Nonspecific enhancement of disc due to degenerative disc disease
  - Enhancement of type I degenerative endplate changes

Imaging Recommendations

- Best imaging tool
  - Flexion/extension plain films
- Protocol advice
  - MR findings useful as secondary tool for degeneration, endplate changes, stenosis, and herniation

DIFFERENTIAL DIAGNOSIS

Pseudoarthrosis

- Abnormal low T1 signal extending through disc, posterior elements, and ligaments

Infection

- Endplate destruction, disc T2 hyperintensity

Tumor

- Enhancing soft tissue mass

Postoperative

- Following multilevel laminectomy or facetectomy

PATHOLOGY

General Features

- Etiology
  - Multifactorial
- Clinical and imaging relationship of disc degeneration to instability controversial
- Many causes of spinal instability
  - Fractures
  - Infection (especially anterior column involvement)
  - Primary bone and metastatic tumors (vertebral body destruction, neural compression, post resection)
  - Isthmic spondylolisthesis (L5-S1 progressive deformity in children)
  - Scoliosis
  - Degenerative instabilities
    - Axial rotational (recurrent pain worse with twisting)
      - Plain films show malaligned spinous processes and pedicle rotation
    - Translational (degenerative spondylolisthesis)
      - Plain films show spondylolisthesis, traction spurs, vacuum phenomenon

- Retrolisthesis (posterior displacement of cephalad body, loss of posterior disc height, facet subluxation)
  - Plain films show increased retrolisthesis with extension
- Degenerative scoliosis (low back pain ± claudication, radiculopathy)
  - Imaging shows central and foraminal stenosis
- Post laminectomy (resection of 50% of bilateral facets alters segmental stiffness)
  - Plain films show new or worsening segmental motion
- Post fusion (altered biomechanics from fusion, but little correlation between radiographic findings and clinical symptoms)

Staging, Grading, & Classification

- Spine Instability Neoplastic Score (SINS)
- 6 components, with numerical scores given for each component
  - Location, mechanical pain, bone lesion quality, spine alignment, vertebral body collapse, involvement of posterior elements
  - SINS is generated by tallying each score from the 6 components
    - 0-6 denotes stability
    - 7-12 denotes indeterminate
    - 13-18 denotes instability

Gross Pathologic & Surgical Features

- Acutely is related to trauma
- In chronic state, mainly findings of degenerative disc disease
- Wide variety of primary and metastatic neoplasms may cause spinal instability

Microscopic Features

- Same as degenerative disc disease, or specific neoplastic process

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific symptoms
  - Low back pain with occasional radiation
  - Physical signs suggesting instability include palpable step off deformity ± spinous process lateral deviation
  - Paraspinal muscle spasms
  - Nonspecific clinical findings lead to use of radiographic surrogate findings
- Clinical profile
  - Elderly woman with nonspecific back pain with evidence of L4-L5 spondylolisthesis by plain films

Demographics

- Age
  - > 50 years
- Gender
  - M < F for degenerative anterior slip, especially at L4-L5
- Epidemiology
  - Translational instability occurs 5-6x more often in women, 3x more common in African American women than Caucasian women
  - Facet joint angulation has been suggested as cause

Natural History & Prognosis

- 20% of patients have instability resolve over 10-year time period
- Increased flexion on plain films does not adversely affect long-term clinical outcome
- L5-S1 isthmic spondylolisthesis rarely unstable in adults

Treatment

- Conservative medical treatment of pain, including exercise, physical therapy, NSAIDs
- Surgical treatment
  - Posterolateral fusion (± pedicle screw fixation)
    - Disadvantages include variable satisfactory outcomes (16-95%), pseudoarthrosis (14-70%), reoperation rates (25%)
  - Interbody fusions via anterior, posterior, or circumferential approaches
    - Posterior lumbar interbody fusion (PLIF)
    - Anterior lumbar interbody fusion (ALIF)
    - Transforaminal lumbar interbody fusion (TLIF)
- Risks: PLIF (arachnoiditis, epidural hemorrhage, cage displacement), ALIF (retrograde ejaculation, cage displacement, iatrogenic herniation)

DIAGNOSTIC CHECKLIST

Consider

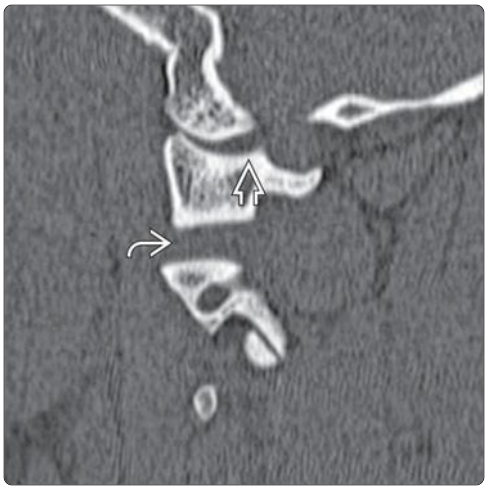
- Findings of "mechanical back pain" present with degenerative instability
- Specificity of plain films limited by high prevalence of lumbar spondylosis in general population

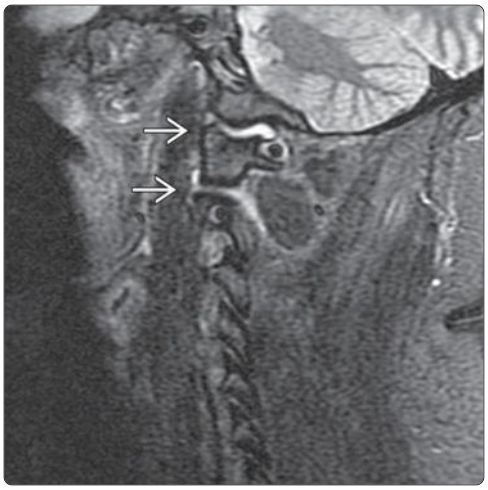
Image Interpretation Pearls

- Radiographic assessment begins with standing AP and lateral views of lumbar spine with flexion/extension views

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(Left) *Sagittal NECT in this trauma patient shows posteriorly widened atlantooccipital joint and widened C1-C2 articulation in this combined C0-C1 and C1-C2 disassociation.* **(Right)** *Sagittal STIR MR in this trauma patient shows effusions at C0-C1 and C1-C2 joints. Fluoroscopy at the time of surgery showed instability with marked widening of the C0-C1 joint. Patient underwent successful occiput to C3 fusion.*





(Left) *Sagittal T1WI MR in an adolescent shows severe spondylolisthesis L5 on S1, with disc degeneration and extensive type I endplate changes.* **(Right)** *Sagittal T1WI C+ MR shows progressive deformity and retrolisthesis at the operative site following extensive posterior debridement for infection, with marked thecal sac compression from the deformity and residual phlegmon, and extensive dorsal pseudomeningocele.*



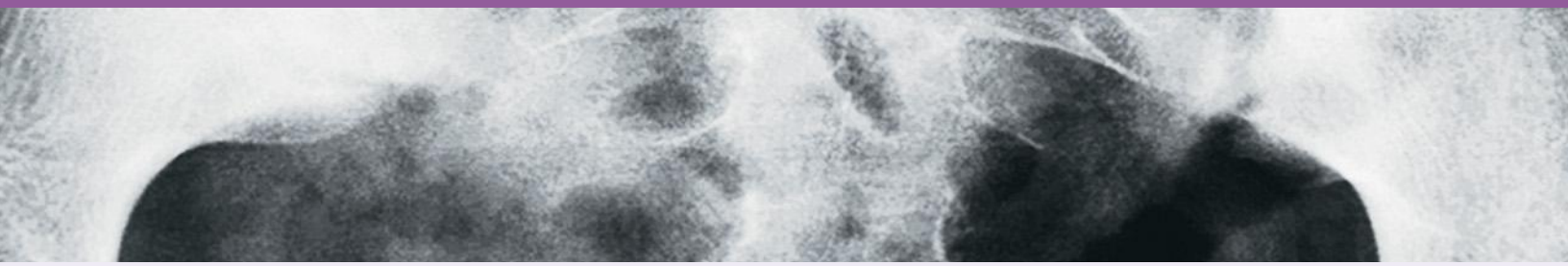


(Left) *Sagittal T1WI MR shows retrolisthesis L2 on L3 with type II fatty degenerative endplate changes. Anterolisthesis was present at L4-L5 with vacuum phenomenon.* **(Right)** *Sagittal T1WI MR shows progressive retrolisthesis at L2-L3, with loss of previous type II fatty endplate changes reflecting ongoing instability at that level.*

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PART III
SECTION 3

Inflammatory, Crystalline, and Miscellaneous Arthritides



Adult Rheumatoid Arthritis	**522**
Juvenile Idiopathic Arthritis	**528**
Spondyloarthropathy	**534**
Neurogenic (Charcot) Arthropathy	**540**
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Ankylosing Spondylitis	**546**
CPPD	**550**
Gout	**556**
Longus Colli Calcific Tendinitis	**558**

KEY FACTS

IMAGING

- Erosions of dens, uncovertebral joints, facet joints
- Atlantoaxial instability in 20-86% patients with RA
- Cranial settling occurs in 5-8% of RA patients
- Lower cervical spine: Facet and uncovertebral joint erosions, instability
- Neutral, flexion, and extension lateral radiographs performed for evaluation of instability
  - Normal: 2 mm between inner margin anterior ring of C1 and dens
  - High correlation to neurologic symptoms with distance ≥ 9 mm
- Pannus is mass-like and surrounds and erodes dens, facet joints, uncovertebral joints
  - Low signal on T1WI
  - Heterogeneous signal on T2WI, STIR
  - Enhances avidly with gadolinium

TOP DIFFERENTIAL DIAGNOSES

- Seronegative spondyloarthropathy
- Calcium pyrophosphate dihydrate deposition (CPPD) disease
- Juvenile chronic arthritis
- Osteoarthritis

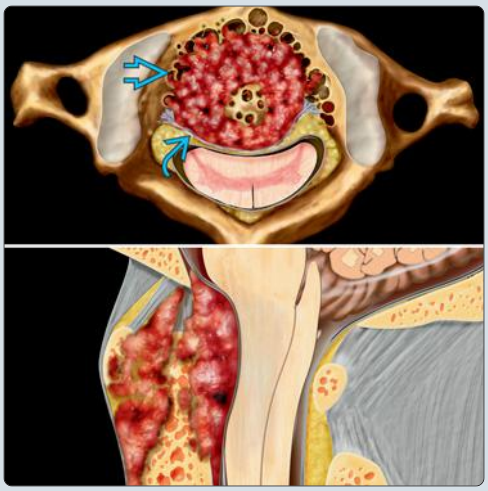
CLINICAL ISSUES

- 50-60% of RA patients have involvement of cervical spine
- Never involves spine before hands &/or feet
- May develop radiculopathy, myelopathy
- Instability → significant morbidity, mortality

DIAGNOSTIC CHECKLIST

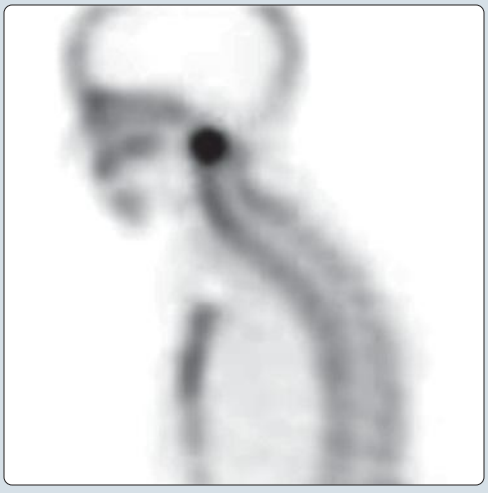
- Calcifying mass with odontoid erosions is **not** RA
  - Indicates crystalline arthropathy, usually CPPD

(Left) *Axial and sagittal graphics show erosion of dens by hypertrophied synovial tissue (pannus). Pannus has eroded the transverse ligament of the dens, resulting in instability. The spinal cord is compressed.* **(Right)** *Coronal CT reconstruction illustrates erosive changes at the right C1-2 joint and lateral subluxation of C1 with respect to C2. Inflammatory synovial proliferation and destruction of surrounding bone also affect the uncovertebral joints in the subaxial spine.*





(Left) *Lateral view from a bone scan shows focal marked uptake at the C1-2 junction in this patient with RA.* **(Right)** *Sagittal NECT scan shows upward translocation of the odontoid and widened atlantodental interval. The skull and C1 have remained together, with the ligamentous laxity and disruption at C1-2 (coupled with the C1 lateral mass collapse), allowing C2 to migrate cephalad into the foramen magnum with brainstem compression (not shown).*





TERMINOLOGY

Abbreviations

- Rheumatoid arthritis (RA)

IMAGING

General Features

- Location
  - Cervical spine: Approximately 50-60% of RA patients
    - C1-odontoid articulations, occipital condyles
    - Facet and uncovertebral joints
  - Severity of craniovertebral junction (CVJ) involvement is factor of disease duration and severity

Radiographic Findings

- Radiography
  - Erosions of dens, uncovertebral joints, and facet joints
    - Dens may be completely eroded
  - Neutral, flexion, and extension lateral radiographs to evaluate for instability
  - C1-C2 instability
    - Normal: < 2 mm between inner margin anterior ring of C1 and dens (anterior atlantodental interval [AADI])
      - Neurologic symptoms usually present if distance ≥ 9 mm
    - Spinal canal < 14 mm at C2 (posterior atlantodental interval [PADI]) also correlates with neurologic symptoms
    - Atlantoaxial instability (AAI) in 20-86% of patients with RA
    - Extension view is useful to determine if pannus precludes full reduction
  - Acquired basilar invagination (BI): Decreased distance occiput to C2
    - Used synonymously with cranial settling
    - Bony erosions of occipital condyles and C1 lateral masses where cranium moves downward (settles) relative to dens
      - Dens may protrude through foramen magnum
      - C1 maintains normal relationship with clivus
      - Alar and apical ligaments involved
      - Occurs in 5-8% of RA patients
  - Instability may also be present at lower levels of cervical spine
    - Often multilevel ("stepladder") subluxations
  - Ankylosis of spine not seen in adult RA
  - Disc and adjacent vertebral body destruction (extremely rare)

CT Findings

- Bone CT
  - Increased conspicuity of odontoid erosions compared to radiographs
  - Pannus around dens may be seen; **never** calcifies
    - Pannus is mass-like, surrounds and erodes dens, facet joints, uncovertebral joints
  - Erosions of uncovertebral and facet joints

MR Findings

- Pannus
  - Low signal intensity (SI) on T1WI
  - Heterogeneous SI on fluid-sensitive sequences
  - Contrast-enhanced T1WI MR may be able to discriminate between joint effusion and various forms of pannus
    - Hypervascular pannus is distinguished from hypovascular and fibrous pannus on T2WI MR
    - Joint effusion and pannus are detected in patients with negative radiographic findings
    - No concordance between MR findings and clinical symptoms
- Subluxations may lead to spinal stenosis
  - Myelopathic changes are common

Imaging Recommendations

- Best imaging tool
  - Cervical spine radiographs in flexion/extension to assess for instability
    - CT of CVJ in flexion/extension
  - Plain radiographs of hands &/or feet to confirm diagnosis
  - Thin-section bone algorithm CT with sagittal and coronal reformats for surgical planning
  - MR imaging in patients with cord symptoms

DIFFERENTIAL DIAGNOSIS

Seronegative Spondyloarthropathy

- Corner erosions of anterior cortex vertebral bodies
- Bony ankylosis of vertebral bodies, facet joints

Calcium Pyrophosphate Dihydrate Deposition (CPPD) Disease

- Crystal deposition causes inflammatory mass around odontoid
- Calcification is typically present
  - Seen in ligaments, discs, facet joints, around dens

Juvenile Idiopathic Arthritis

- Fusion of vertebral bodies, facet joints, erosions
- Growth disturbance is common

Osteoarthritis

- Involves synovial joints of spine
- Osteophytes and joint narrowing
- May cause "pseudopannus" at dens

Gout

- Rare in spine
  - Usually centered on disc
- Involvement of cervical vertebrae may produce bone erosion and ligament softening
  - Potential subluxation can produce spinal cord compression

PATHOLOGY

General Features

- Etiology
  - Unknown, strong hereditary component
  - Autoimmune
    - T cells produce IL-17, cytokine promoting synovitis
    - B cells present antigens, produce autoantibodies and cytokines
- Laboratory findings
  - ESR, C-reactive protein (CRP) elevated

- Autoantibodies, such as rheumatoid factor (RF) and anticitrullinated protein antibody (ACPA)
  - Can precede clinical manifestation of RA by many years
  - RF may be negative early in disease course
    - Eventually positive in up to 5% of RA patients
    - False-positive RF is seen in elderly patients, systemic lupus erythematosus, sarcoidosis, cryoglobulinemia, cirrhosis, endocarditis

Staging, Grading, & Classification

- 2010 American College of Rheumatology/European League Against Rheumatism criteria
  - Target population: Patients with at least 1 joint with clinical synovitis not explained by other disease
  - Joint involvement
    - Large ± small joints
  - Serology
    - Presence of RF, ACPA
  - Acute-phase reactants
    - CRP, ESR
  - Duration of symptoms
    - Acute and chronic

Gross Pathologic & Surgical Features

- Inflammatory proliferation of synovium = pannus
  - Osteoclast-rich portion of synovial membrane destroys bone
- Enzymes secreted by synoviocytes and chondrocytes degrade cartilage

Microscopic Features

- Thickened synovium containing plasma cells, multinucleated giant cells, polymorphonuclear leukocytes and lymphocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Morning pain and stiffness
    - Atlantoaxial subluxation may cause suboccipital pain secondary to compression of C2 nerve
  - Radiculopathy
- Other signs/symptoms
  - Cervical myelopathy
  - Autonomic centers can be compromised, leading to labile blood pressures, arrhythmias, respiratory depression, or sudden death
  - Anterior spinal arteries may also be involved, leading to neurologic deficits, vertebrobasilar insufficiency, or transient ischemic attacks
  - Pathologic fracture

Demographics

- Age
  - Occurs at all ages, most common in middle age
- Gender
  - 3x more common in women
    - Men represent only 25% of population yet have greater risk of advanced cervical involvement
- Epidemiology
  - 0.3-1.5% of population has RA
  - 33% of all RA patients have spine involvement
    - Involvement of cervical spine in 17-86%
      - CVJ most often affected

Natural History & Prognosis

- Increased morbidity and mortality with CVJ instability
  - RA may start with AAI and progress to BI
- Neurologic symptoms poor prognostic sign as mortality rate increases dramatically
- After stabilizing surgery, periodontoid pannus has been shown to decrease in size

Treatment

- Medical treatment
  - Corticosteroids
  - Disease-modifying agents slow disease progression
    - Tumor necrosis factor inhibitors, methotrexate (MTX), cyclosporine
    - Biological treatment, such as infliximab, for failure of initial MTX treatment
- Surgical indications for AAI
  - Pain, myelopathy, cord compression, symptomatic vertebral artery compromise
  - AADI > 8-10 mm
  - Early surgery for AAI may prevent BI/cranial settling
  - C1-C2 transarticular surgical fusion for atlantoaxial subluxation
- Surgical indications for BI
  - Lower cranial neuropathy (C9-C12)
  - Myelopathy, pain, symptomatic vertebral artery compromise, deformity
  - Severe compression of cervicomedullary junction
    - Cervicomedullary angle < 135° represents significant disease
- Transoral odontoidectomy for cord compression

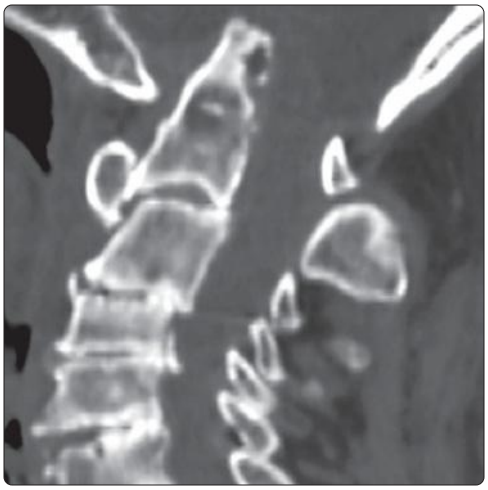
DIAGNOSTIC CHECKLIST

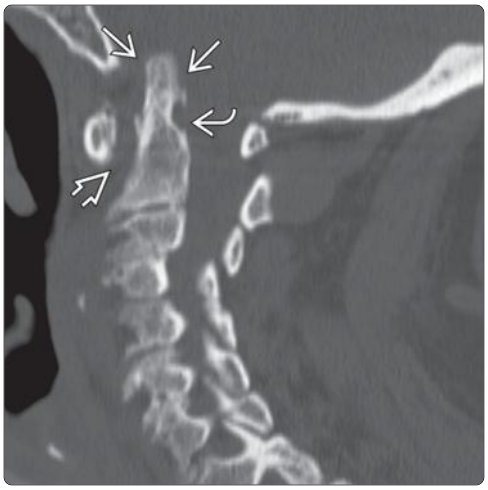
Image Interpretation Pearls

- Flexion/extension views often technically inadequate due to lack of patient motion
  - Do not misinterpret as stability
- Calcifying epidural mass with odontoid erosions is not feature of RA
  - Indicates crystal deposition, usually CPPD

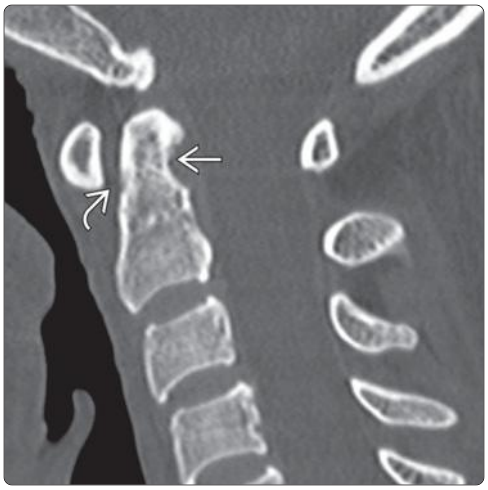
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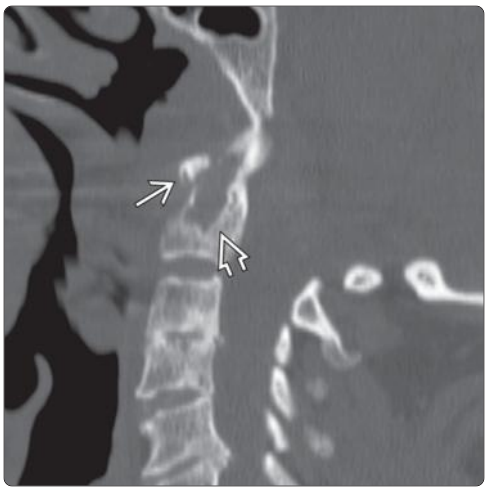


(Left) *Sagittal CT demonstrates superior migration of the dens. Cranial settling may lead to compression of the brainstem by the dens with compression of the autonomic centers with labile blood pressures, arrhythmias, or sudden death.* **(Right)** *Sagittal NECT shows cranial settling with upward translocation of the odontoid process relative to foramen magnum. There are also odontoid erosions and increased atlantodental interval.*





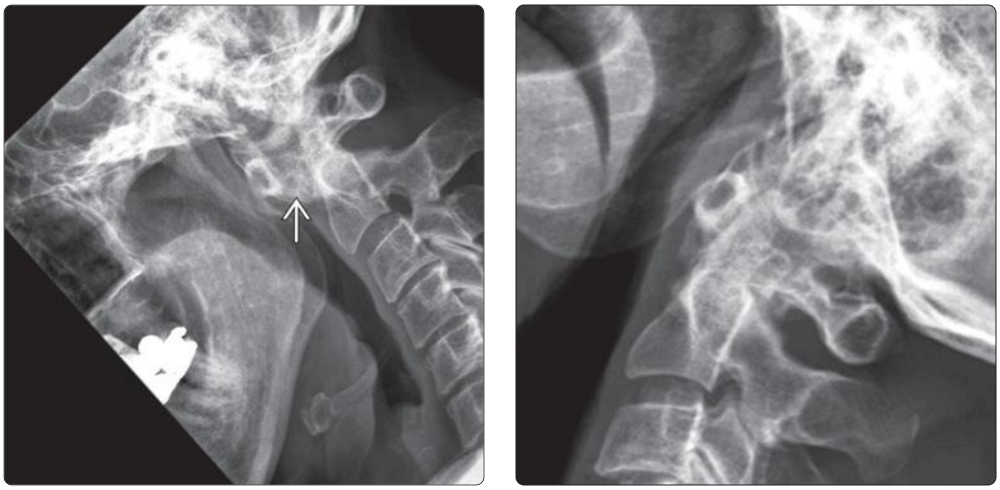
(Left) *Sagittal CT reconstruction exhibits erosions along the posterior margin of the dens and widening of the AADI.* **(Right)** *Sagittal T1WI MR depicts widening of the AADI related to inflammatory pannus and erosive changes of the posterior dens, consistent with known RA.*



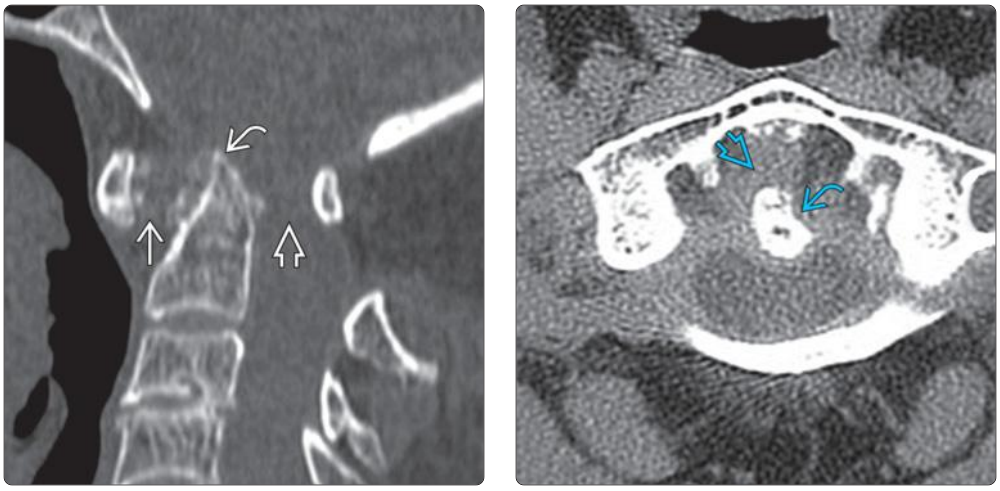


(Left) *Sagittal NECT (bone window) shows extensive erosive changes of the anterior arch of C1 and odontoid process. The odontoid is malpositioned and touching the clivus.* **(Right)** *Sagittal T1WI MR shows a large amount of pannus both ventral and dorsal to the odontoid with effacement of the thecal sac and posterior displacement of cord.*

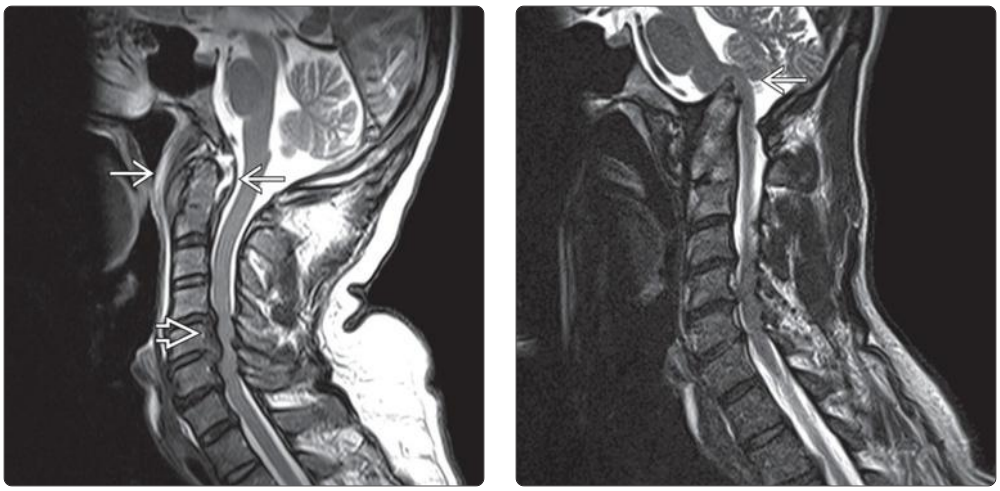
(Left) *Sagittal radiograph in the flexed position illustrates marked widening of the AADI. Inflammatory destruction via synovitis of the atlantal transverse ligament leads to anteroposterior instability of the atlantoaxial joint.* **(Right)** *Sagittal radiograph in the same patient shows a decrease in the AADI with the neck extended. However, PADI has been shown to be a more reliable predictor of whether neurologic symptoms will develop. MR is recommended when PADI is ≤ 14 mm.*



(Left) *Sagittal CT demonstrates marked erosive change of the dens. Erosion of the odontoid process frequently coincides with the development of anterior atlantoaxial subluxation. Widening of the AADI is also noted. There is narrowing of the PADI, and the space for the cord is markedly narrowed.* **(Right)** *Axial NECT displays abnormal soft tissue widening the predental space. There are erosive changes along the margin of the dens. Pannus is the main cause of bone destruction.*



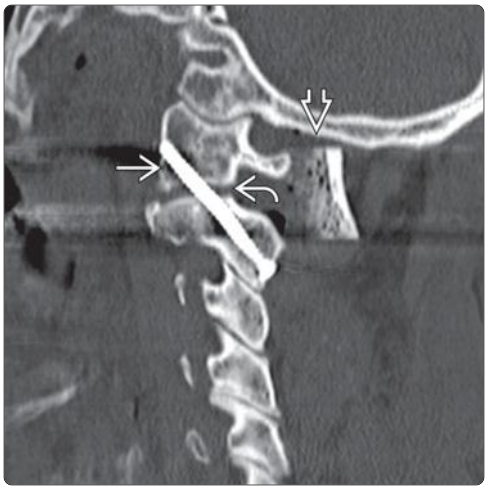
(Left) *Initial MR shows rapid development cranial settling & severe inflammatory response surrounding C1-2 articulation with a ventral and dorsal soft tissue mass, but a relatively preserved relationship of C2 with the foramen magnum. Large disc extrusion is seen at C5.* **(Right)** *Follow-up MR 6 months later (a further example of rapid development cranial settling) shows rapid progression of cranial settling with upward translocation of the odontoid process through the foramen magnum with severe cervicomedullary cord compression.*





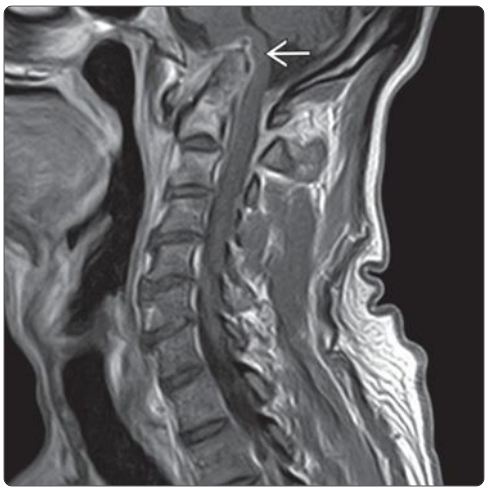


(Left) *Sagittal CT reconstruction displays marked erosive changes at the dens, likely due to pannus formation. There is also severe cervical degeneration with ankylosis at multiple levels and kyphosis.* **(Right)** *Coronal bone CT shows subluxation of the C1 lateral masses of C1 relative to both C2 and the occipital condyles. The dens is absent. Erosions involve multiple uncovertebral and facet joints as well as the intervertebral disc.*





(Left) *Sagittal bone CT in the same patient shows screw placement from the C2 articular pillar into the C1 lateral mass and posterior strut graft. In this early postoperative case, CT shows that the facet joint is still open.* **(Right)** *Coronal bone CT shows resection of the dens and posterolateral C1-C2 fusion in this patient with unstable RA. There is progressive erosion at the C3-C4 uncovertebral joints. Fixation screws traverse the C1-C2 facet joints.*





(Left) *Sagittal T1 C+ MR (an example of severe cord compression treated with odontoid resection) shows upward translocation of C2 with cranial settling and severe cervicomedullary cord compression.* **(Right)** *Postoperative MR (a further example of severe cord compression treated with odontoid resection) shows transoral resection of the odontoid process, which has decreased the mass effect upon the cervicomedullary junction. Patient has also undergone an occipital/upper cervical fusion.*

KEY FACTS

TERMINOLOGY

- Spectrum of idiopathic inflammatory arthropathies occurring in childhood

IMAGING

- Cervical spine subluxations, fusions, and growth disturbance
- Bony overgrowth due to hyperemia may also be seen
- Cranial settling and basilar invagination are common
- Involves discs, facets, and uncovertebral, costovertebral and costotransverse joints
- Progresses to ankylosis
  - Fused vertebrae are small in anteroposterior dimension if fused early in childhood
- Pannus on MR: Rounded, periarticular mass
  - ↓ signal on T1WI; intermediate, heterogeneous signal on T2WI, STIR
- Synovitis: Effusions in facets and sacroiliac joints

TOP DIFFERENTIAL DIAGNOSES

- Congenital spinal fusion
- Physiologic cervical spine subluxations
- Down syndrome
- Osteogenesis imperfecta
- C1-C2 osteomyelitis

PATHOLOGY

- Rheumatoid factor (RF) is negative

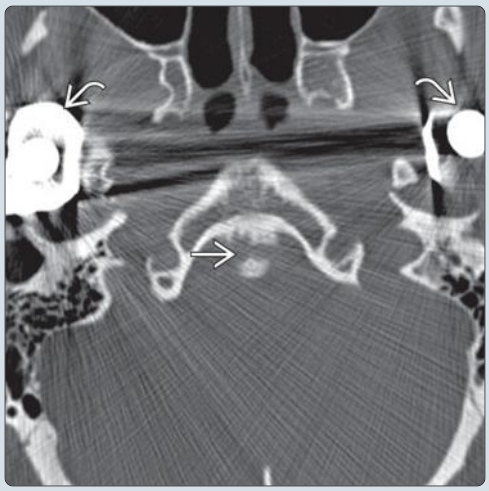
CLINICAL ISSUES

- Vague, pauciarticular pain
- Limited neck movement

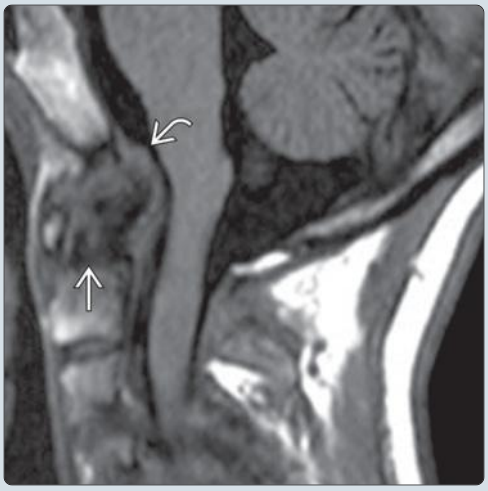
DIAGNOSTIC CHECKLIST

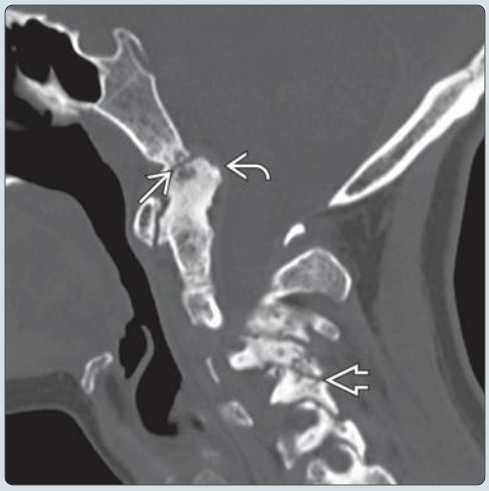
- Diagnosis is often delayed due to subtle imaging findings and nonspecific symptoms





(Left) *Coronal CT shows erosions along the dens, C1-C2 facet joint, and C3-C4 uncovertebral joint. Joint space narrowing and growth disturbances are key radiologic manifestations of juvenile idiopathic arthritis (JIA).* **(Right)** *Axial bone CT reveals bilateral temporomandibular joint (TMJ) prostheses. Erosions of the dens are also seen. Destruction of the joint and the mandibular condyle may result in growth retardation of the TMJ with consequent micrognathia, hypoplasia, and asymmetry.*





(Left) *Sagittal T1WI MR depicts a hypointense retroodontoid soft tissue pannus, which effaces the ventral CSF collar at the cervicomedullary junction. The dens marrow hypointensity likely reflects hyperemia/edema.* **(Right)** *Sagittal CT demonstrates superior migration of the dens into the foramen magnum. Erosive changes of the dens, clivus, and subaxial facet joints are seen. Erosions in children develop later in the course of JIA as compared with those observed in adult RA.*

TERMINOLOGY

Abbreviations

- Juvenile idiopathic arthritis (JIA)

Synonyms

- Juvenile chronic arthritis (JCA), juvenile rheumatoid arthritis (JRA)

Definitions

- Heterogeneous group of arthritides that begin before patient is 16 years of age, persist for more than 6 weeks, and are of unknown origin

IMAGING

General Features

- Best diagnostic clue
  - Cervical spine subluxations and growth disturbance

Radiographic Findings

- Cervical vertebral fusions
  - Involve discs &/or facet joints
  - May involve multiple levels
  - Accelerated degenerative disc disease at unfused levels
  - Anterior atlantoaxial subluxation (33%)
  - C1-C2 arthritis (17%)
  - Apophyseal joint arthritis (16%)
- Growth disturbances
  - Fused vertebrae are small in anteroposterior dimension if fused early in childhood
    - More differences in sizes of individual vertebrae, representing growth disturbances
  - Bony overgrowth due to hyperemia may also be seen
  - Cranial settling and basilar invagination are common
- Erosions
  - Odontoid process is characteristically involved
  - Facet, uncovertebral, costovertebral, costotransverse joints
  - May progress to ankylosis
  - Because cartilage is thicker in children, bone erosions are less commonly seen than in adults
- Multilevel vertebral subluxations
  - Craniocervical or subaxial
  - May be rotary, as well as anteroposterior
- In 1 study, radiographs showed cervical lesions in 65% of polyarticular JIA (pJIA)
  - Presence of cervical lesions correlates with more severe disease
  - Patients with severe hand JIA usually also have cervical spine involvement

CT Findings

- Can be used for early diagnosis of erosions

MR Findings

- Erosions
  - Cartilage erosion is seen before bone erosion
  - Usually surrounded by edema with ↓ signal on T1WI and ↑ signal on T2WI or STIR
- Pannus
  - Rounded, periarticular mass
  - ↓ signal on T1WI, intermediate and heterogeneous signal on T2WI or STIR
  - Moderate enhancement with gadolinium
- Synovitis
  - Effusions in facet joints
  - Nodular thickening of synovium
  - Synovium excretes contrast
    - Apparent synovial thickening may represent contrast diffusing into joint

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan
  - Oligoarticular disease easily misdiagnosed as infection on bone scan
- Gallium and indium scans
  - High concentration of WBC in joints affected by JIA

Imaging Recommendations

- Best imaging tool
  - Earliest diagnosis by MR
- Protocol advice
  - Sagittal T1WI, STIR, axial T2WI FSE
  - Gadolinium increases sensitivity in early diagnosis, but is usually not needed

DIFFERENTIAL DIAGNOSIS

Congenital Spinal Fusion

- Isolated anomaly or in association with syndromes, such as VACTERL
- No systemic symptoms

Physiologic Cervical Spine Subluxations

- Normal vertebral morphology, < 2 mm

Down Syndrome

- Normal vertebral morphology, ± subluxations

Osteogenesis Imperfecta

- Diffuse osteopenia, no systemic symptoms
- Vertebra plana and fractures

C1-C2 Osteomyelitis

- Pain, fever
- Disc destruction > JIA
- C1-C2 instability (Grisel syndrome)

PATHOLOGY

General Features

- Etiology
  - Heterogeneous group of diseases
  - Oligoarticular and rheumatoid factor (RF)-positive polyarticular JIA: Autoimmune disease of adaptive immune system
    - Antinuclear antibody (ANA) and RF are common; associated with *HLA* genes
    - Environmental trigger may cause uncontrolled innate and adaptive immune response toward self-antigen, resulting in inflammation and disease
  - Systemic JIA: Autoinflammatory disease, primarily involving innate immune system

- Uncontrolled activation of phagocytes by unknown triggers, leading to increase of inflammatory cytokines
- Associated abnormalities
  - Young children may have fever, anemia, hepatosplenomegaly
  - Children with JIA are at risk of osteopenia and osteoporosis, leading to increased risk of fracture

Staging, Grading, & Classification

- International League of Association for Rheumatology criteria
  - Oligoarticular JIA
    - Involves ≤ 4 joints
      - Large joints (e.g., knee, hip, shoulder, elbow)
    - Uveitis in 30%
    - RF(-)
  - Seropositive pJIA
    - Involves ≥ 5 joints
      - Arthritis is often erosive and symmetric, involving wrists and small joints of hands and feet
    - Adolescent
  - Seronegative pJIA
    - Involves ≥ 5 joints
    - 25% ANA(+)
    - ± cervical spine and temporomandibular joint (TMJ)
  - Systemic onset JIA
    - Arthritis and systemic features (e.g., fever or rash)
    - Lymphadenopathy and hepatosplenomegaly
    - Cardiac disease and pericardial effusions in ~ 10%
    - Leukocytosis, thrombocytosis, anemia, ↑ transaminase levels, ↑ inflammatory markers
    - ANA(+) in 5-10%; RF is rare
  - Enthesitis-related arthritis
    - Involves lower limb, especially hip and intertarsal joints
    - Sacroiliac joint involved later in course of disease
      - Some develop ankylosing spondylitis within 10-15 years of disease onset
  - Psoriatic JIA
    - Arthritis, psoriasis
    - Dactylitis, nail changes
  - Undifferentiated JIA

Gross Pathologic & Surgical Features

- Growth disturbance is often most outstanding feature
- Fusion of joints common in appendicular skeleton, as well as spine

Microscopic Features

- Thickened synovium containing plasma cells, multinucleated giant cells, PMNs, lymphocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Vague, pauciarticular pain
  - Limited neck movement
  - Peripheral tenosynovitis may be presenting complaint
- Other signs/symptoms
  - Rash, iridocyclitis, pericarditis, tenosynovitis, intermittent fever, or morning stiffness
  - MR evidence of TMJ arthritis was detected in 43% of patients in 1 study

Demographics

- Age
  - Younger children often have systemic disease
  - Older children/adolescents present with joint pain
- Gender
  - F > M
- Epidemiology
  - Prevalence rate: 0.07-4.01 per 1,000 children
  - Annual incidence: 0.008-0.226 per 1,000 children
  - European descent may be important predisposing factor for oligoarticular JIA and psoriatic JIA

Natural History & Prognosis

- Growth disturbances lead to premature osteoarthritis
- Early onset of neck involvement correlates to worse prognosis
- Often "burns out" as children approach adulthood
  - Duration of symptoms in childhood correlated to risk of continued disease in adulthood
  - At least 1/3 will have ongoing active disease in their adult years

Treatment

- NSAIDs, steroids, methotrexate, sulfasalazine, leflunomide, antitumor necrosis factor medications
- Interleukin inhibition: Tocilizumab (anti-interleukin-6)
- T-cell costimulatory modulator
- B cell depletion: Rituximab

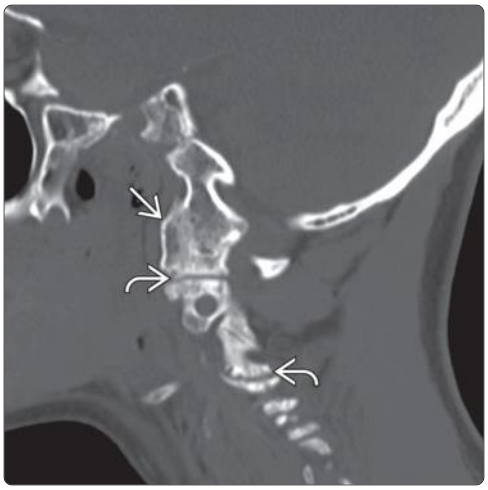
DIAGNOSTIC CHECKLIST

Consider

- Diagnosis is often delayed due to subtle imaging findings and nonspecific symptoms
- Consider JIA in child with vague, chronic articular, neck pain, or chronic systemic illness

SELECTED REFERENCES

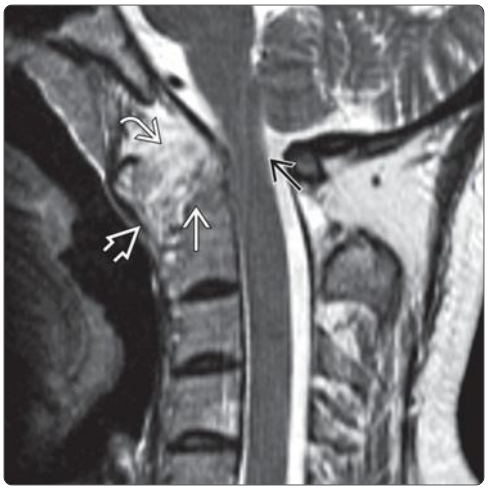
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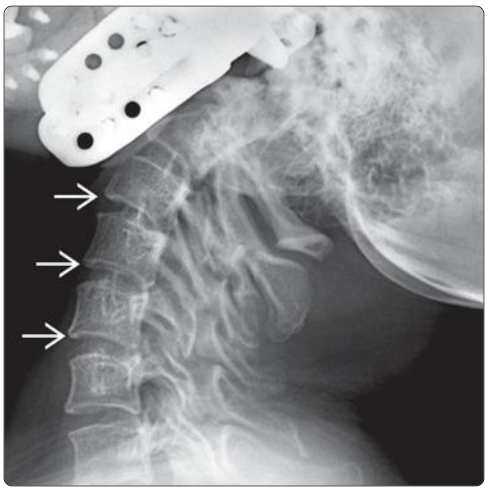
(Left) *Sagittal CT demonstrates ankylosis of the atlantooccipital joint. There are erosive changes of the facet joints. Joint ankylosis occurs more promptly in JIA than in adult RA, particularly in the carpal and tarsal joints and in the cervical spine.* **(Right)** *Sagittal CT exhibits erosive changes at the C1-C2 facet joint. There is mild anterior subluxation of C4 on C5, and C5 on C6. Radiographic narrowing of a joint space is the reflection of cartilage loss.*





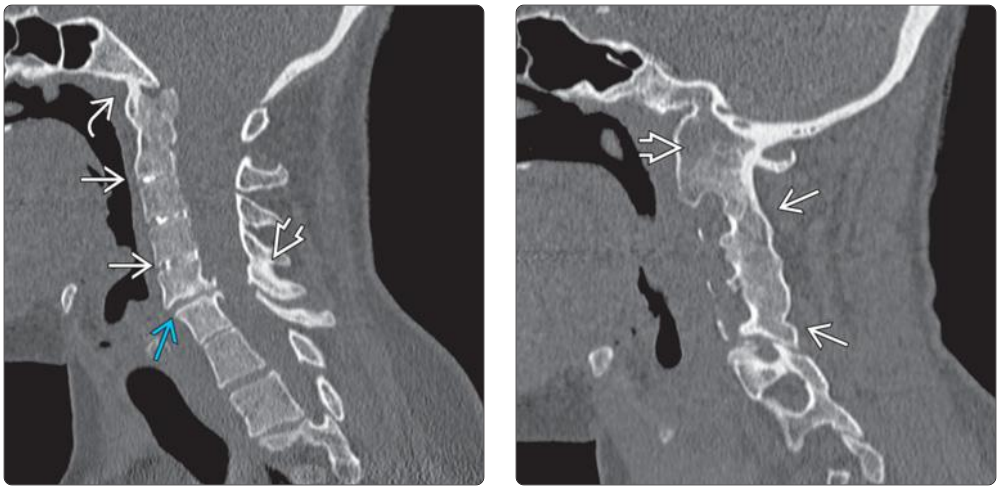
(Left) *Sagittal bone CT in an adult patient with a history of JIA shows a triangular dens from healed erosions and ossification. C1 is fused to the occiput posteriorly. Although many children experience spontaneous remission, at least 1/3 will have ongoing active disease in their adult years.* **(Right)** *Sagittal T2WI MR depicts an eroded dens with pannus widening the atlantodental space. The anterior longitudinal ligament is displaced anteriorly. There is marked narrowing of the foramen magnum.*



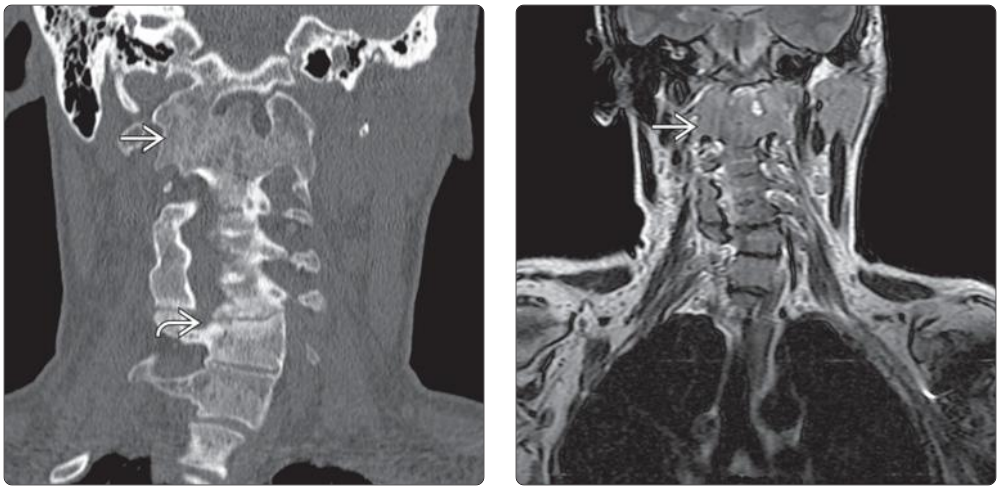


(Left) *Lateral radiograph in a flexed position illustrates anterior subluxation of multiple subaxial cervical vertebral levels. Metallic TMJ prosthesis are seen.* **(Right)** *Lateral radiograph of the same patient in extension shows that the subluxations get smaller. JIA tends to ankylose the apophyseal joints and destroy the atlantoaxial joints, resulting in anterior atlantoaxial subluxation or impaction. This inflammatory process can restrict rotatory and bending movements.*

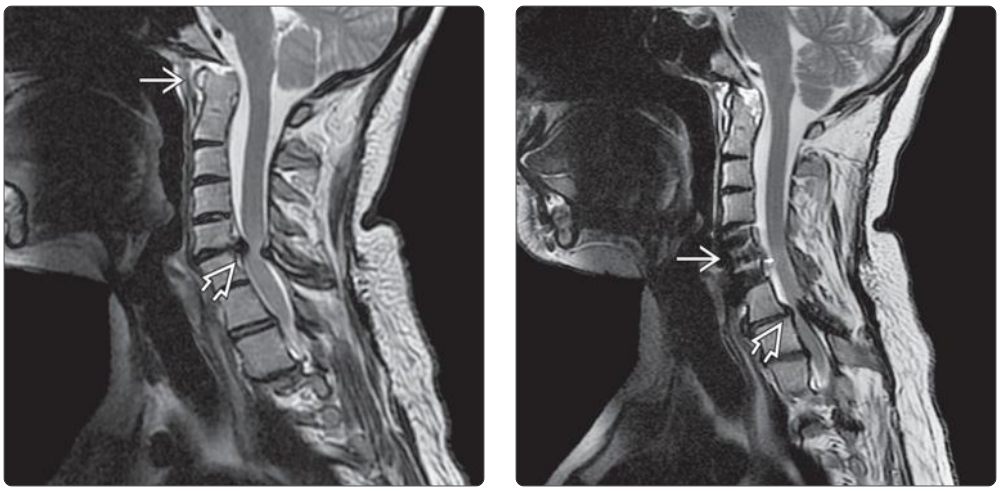
(Left) *Sagittal NECT (bone window) of the cervical spine shows multiple fused vertebral bodies, also involving the posterior elements. The fusion extends to the craniocervical junction. There is severe disc degeneration below the fusion site.* **(Right)** *Sagittal NECT (bone window) through the lateral masses shows extension fusion of the facets with extension to the skull base.*



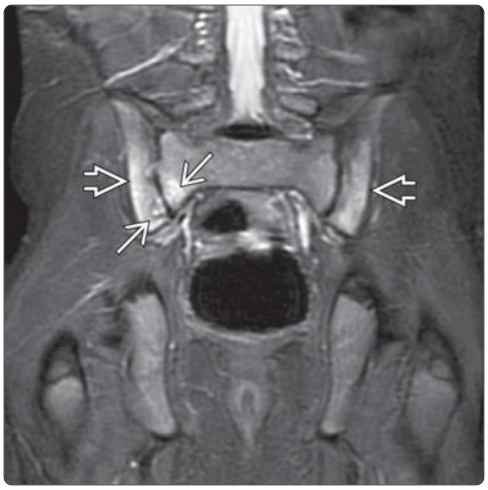
(Left) *Coronal NECT through the cervical spine shows extensive fusion of the vertebral bodies to the craniocervical junction. Note the disc degeneration with bony sclerosis at the inferior margin of the fused segment, so-called accelerated degenerative change.* **(Right)** *Coronal T2 MR in the same patient shows the appearance of the bony fusion mass in this patient with long standing JIA.*



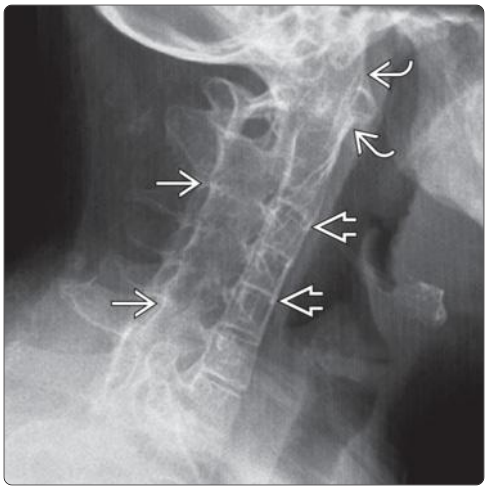
(Left) *Sagittal T2 MR shows fusion most pronounced at the C1-C2 level. The patient has vertebral fusions from C1 to C5, which caused accelerated degeneration at C5-C6 with large disc osteophyte complex compressing the cord.* **(Right)** *Follow-up sagittal T2 MR in the same patient 1 year later shows metal artifact after successful anterior discectomy and fusion for the severe central canal stenosis. Patient is now developing inferior disc degeneration with stenosis.*

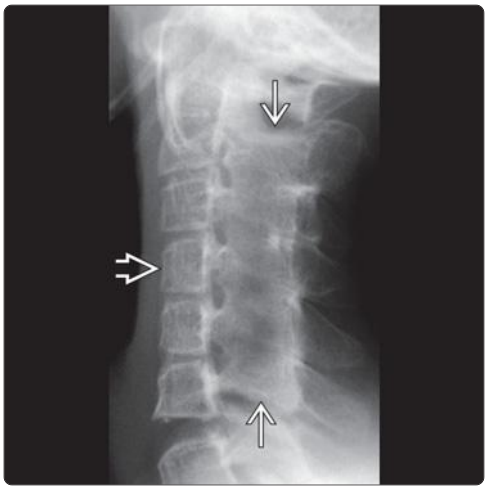






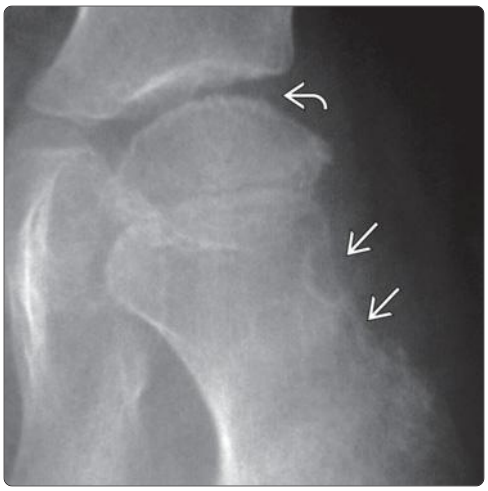
(Left) *AP pelvis radiograph shows right side erosion ⇨. Subtle subcortical indistinctness is present on the iliac side of both SI joints.* **(Right)** *Coronal STIR MR shows intense bone marrow edema ⇨ adjacent to both SI joints. The mild asymmetry is more common in early disease, as in this case. Also note that the bone marrow edema is more extensive and intense on the iliac side of the SI joints, also a common pattern in active sacroiliitis. Also note the right SI joint erosions ⇨.*





(Left) *Lateral radiograph shows extensive osteoporosis. Extensive fusion of the facet joints is present ⇨. Fusion has developed along several disc spaces ⇨ and has extended cephalad to involve the occipitocervical junction ⇨.* **(Right)** *Lateral radiograph shows fusion of the posterior elements and vertebral bodies from C2-C6 ⇨. Since fusion occurred at a young age, the vertebral bodies across the fused section ceased to grow. Note the small body size in the fused portion ⇨ compared with a normal vertebral body in a nonfused portion.*





(Left) *Coronal T1 MR illustrates severe synovitis and innumerable erosions of the carpal bones ⇨ and metacarpal bases ⇨.* **(Right)** *AP radiograph shows severe erosive disease along the femoral neck ⇨, crenelation of the femoral capital epiphysis, and joint space narrowing ⇨. This patient's disease progressed rapidly over 1 year.*

Spondyloarthropathy

KEY FACTS

TERMINOLOGY

- Rheumatoid factor (RF) negative inflammatory arthritis and enthesopathy affecting spine and sacroiliac joints
- Syndesmophyte: Paraspinous ligamentous or disc ossification bridging 2 adjacent vertebral bodies

IMAGING

- Sacroiliac joint (SIJ) erosive arthritis
  - Best seen in lower 1/3 of joint
- Corner erosions of vertebral bodies
- Ossification of paraspinous ligaments and anulus fibrosis
- Inflammatory arthritis of synovial joints
- Fractures: Through syndesmophytes &/or vertebral bodies
- MR primary method to diagnosis early sacroiliitis associated with axial spondyloarthropathy

TOP DIFFERENTIAL DIAGNOSES

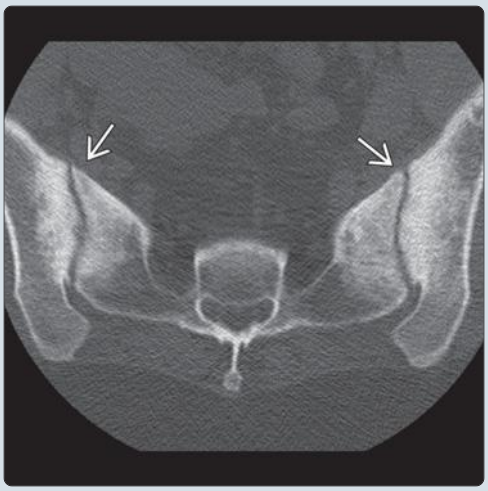
- Rheumatoid arthritis
- Juvenile idiopathic arthritis
- Infectious sacroiliitis
- Diffuse idiopathic skeletal hyperostosis (DISH)
- Retinoid therapy

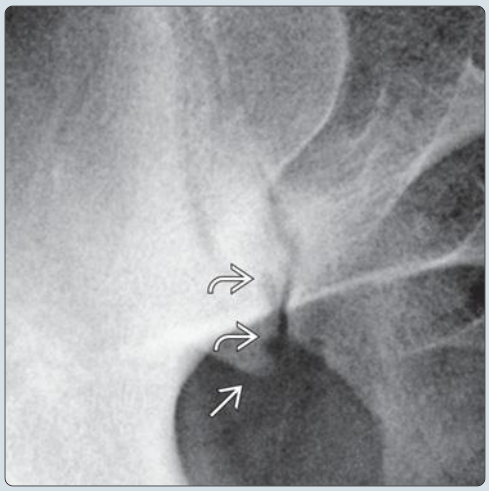
PATHOLOGY

- Classified as
  - Ankylosing spondylitis
  - Reactive spondyloarthropathy (formerly Reiter syndrome)
  - Psoriatic spondyloarthropathy
  - Spondyloarthropathy associated with inflammatory bowel disease
  - Undifferentiated spondyloarthropathy (uSpA)
- Strong association with HLA-B27 haplotype
  - 95% AS, 80% Reiter, 50% psoriatic patients are positive

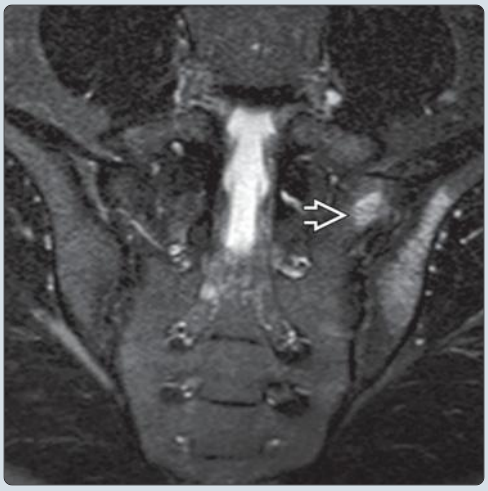
CLINICAL ISSUES

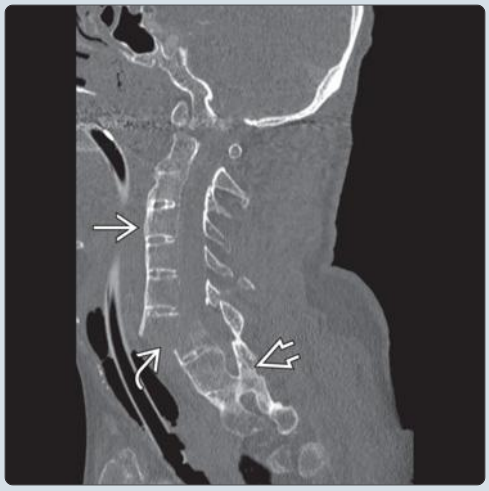
- TNF-α inhibitors effective and have rapid onset





(Left) *Axial bone NECT through the SI joints shows very symmetrical sclerosis surrounding the joints, involving both the sacral ala and iliac wing. There is irregularity of the joint margin, with widening that is slightly worse on the right.* **(Right)** *Anteroposterior radiograph in a patient with reactive arthropathy shows small, serrated erosions. Periosteal reaction has developed at the inferior margin of the joint.*





(Left) *Coronal T2WI FS MR in a patient with early ankylosing spondylitis is angled along the plane of the sacrum. Bone marrow edema is a critical finding and helps to corroborate an active inflammatory process essential for early diagnosis and prompt treatment.* **(Right)** *Sagittal bone NECT in a trauma patient with AS shows fusion extending inferiorly from the C2 involving ALL, discs, and posterior elements. There is fracture dislocation at the C7-T1 level with anterior displacement of the superior spine.*

TERMINOLOGY

Synonyms

- Formerly called seronegative spondyloarthropathy

Definitions

- Rheumatoid factor negative inflammatory arthritis and enthesopathy affecting spine and sacroiliac joints (SIJ)
  - Archetype is ankylosing spondylitis (AS)
- Enthesopathy: Inflammation at attachments of ligaments and tendons (entheses)
- Syndesmophyte: Paraspinous ligamentous or disc ossification bridging 2 adjacent vertebral bodies

IMAGING

General Features

- Best diagnostic clue
  - SI joint erosion or ankylosis
- Location
  - Initially involves sacroiliac joints
  - May involve entire spine, discs, synovial joints
  - Also involves joints of axial and appendicular skeleton
    - Psoriatic arthritis: Involves hands > feet
    - Reactive arthritis: Involves feet > hands
  - Involves tendon and ligament attachments (entheses)

Radiographic Findings

- Sacroiliac joint erosive arthritis
  - Primarily visible in lower 1/3 of joint
  - Erosions seen 1st on iliac side of joint
  - Earliest sign: Loss of definition of subchondral bone plate
  - Erosions small, multiple, "serrated steak knife"
  - Fusion a late phenomenon
  - Bilaterally symmetric: Ankylosing spondylitis, arthritis of inflammatory bowel disease
  - Bilateral but asymmetric: Reactive arthropathy, psoriatic arthritis
- Corner erosions vertebral bodies
  - "Squaring" of anterior vertebral margin
  - Progresses to corner erosions
  - Sclerotic repair leads to "shiny corner" sign (Romanus lesion)
- Ossification of paraspinous ligaments
  - May be bulky; more common in psoriatic and reactive arthropathy
  - Distinguished from osteophytes because originates from midvertebral body rather than endplate
- Ossification of outer fibers of anulus fibrosis
  - When multilevel, results in "bamboo spine" appearance; more common in AS
  - Susceptible to fracture
- Inflammatory arthritis of synovial joints
  - In spine and periphery
  - Erosions often progress to fusion
  - Erosions + periosteal reaction at tendon, ligament attachments
  - May see atlantoaxial subluxation
- Kyphotic deformity
- Fractures
  - Through syndesmophytes &/or vertebral bodies
  - Often difficult to see because of osteopenia, spinal deformity
  - Unstable; typically involve all spinal columns
- Accelerated degenerative disc disease
  - Seen in unfused regions due to increased stress
  - May mimic discitis or neuropathic disease
- Osteopenia

CT Findings

- Bone CT
  - Highly sensitive to early sacroiliac erosions
  - Useful to evaluate fractures

MR Findings

- T1WI
  - Low signal intensity bone marrow at corner and SIJ erosions
  - Joint erosions and ankylosis
- T2WI
  - High signal intensity bone marrow at early corner and SIJ erosions
  - High signal from endplates when inflammatory-type Andersson lesions present
- STIR
  - High signal intensity bone marrow at early corner and SIJ erosions
- T1WI C+
  - Avid enhancement in active disease
- Marrow in disc spaces (later stages), preservation of central canal

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan in active disease

Imaging Recommendations

- MR primary method to diagnosis early sacroiliitis associated with axial spondyloarthropathy
- MR/CT in combination to evaluate bone and cord status following trauma

DIFFERENTIAL DIAGNOSIS

Rheumatoid Arthritis

- Uncommonly involves SIJs
- Primarily involves cervical spine
- Bony ankylosis not a feature

Juvenile Idiopathic Arthritis

- Onset before age 18
- Growth disturbances of vertebrae with fusions
- Juvenile onset ankylosing spondylitis usually limited to SIJs, peripheral enthesopathy

Infectious Sacroiliitis

- Unilateral

Diffuse Idiopathic Skeletal Hyperostosis (DISH)

- Flowing ossification of paraspinous ligaments
- Sacroiliac joints normal

Retinoid Therapy

- Flowing ossification of paraspinous ligaments

- Sacroiliac joints normal

PATHOLOGY

General Features

- Etiology
  - Psoriatic arthritis: Associated with psoriasis but may precede psoriatic rash
  - Reactive arthritis: Follows prior bacterial infection, e.g., venereal or dysentery
  - Also associated with ulcerative colitis and Crohn disease
- Genetics
  - Strong association with HLA-B27 haplotype
    - 95% AS, 80% Reiter, 50% psoriatic patients are positive
    - 6-8% of normal population is HLA-B27 positive
    - Psoriatic arthritis in HLA-B27 negative patients spares spine
    - 1-2% of people with HLA-B27 develop spondyloarthropathy
- Associated abnormalities
  - Uveitis, iritis, conjunctivitis seen with all forms
  - AS: Inflammatory bowel disease, aortitis, upper lobe pulmonary fibrosis, rash
  - Reactive: Urethritis/cervicitis, balanitis, heel pain
  - Peripheral arthritis
    - AS, reactive: Predilection for lower extremity, especially hips, knees, plantar aponeurosis
    - Psoriatic: Predilection for hands
  - Secondary amyloid leading to nephropathy
- RF negative, elevated ESR

Staging, Grading, & Classification

- Divided into 5 types
  - Ankylosing spondylitis
  - Reactive spondyloarthropathy (formerly Reiter syndrome)
  - Spondyloarthropathy associated with inflammatory bowel disease
  - Psoriatic spondyloarthropathy
  - Undifferentiated spondyloarthropathy (uSpA)

Gross Pathologic & Surgical Features

- AS: Extensive ankylosis creates high risk for spinal fractures

Microscopic Features

- Enchondral ossification discs
- Inflammation synovium, subchondral bone, less severe than RA

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sacroiliitis usually 1st manifestation
    - Low back pain, greatest in morning
  - Pathologic fracture
    - Increased fracture risk with multilevel spine ankylosis
    - Often due to minor injury
    - Unstable, high morbidity in elderly
    - Most common at cervicothoracic or thoracolumbar junction
    - May be chronic with instability and pseudoarthrosis (noninflammatory-type Andersson lesion)
  - Other signs/symptoms
    - Fatigue, low-grade fever
    - Rarely, cauda equina syndrome (due to synovial cysts)
  - Cauda equina syndrome as late manifestation of adhesive arachnoiditis associated with AS

Demographics

- Age
  - Early to mid adulthood
- Gender
  - All types more common in men than women
- Epidemiology
  - Approximately 0.1-1% of population
    - Lowest incidence in sub-Saharan Africa
  - Often mild, undiagnosed

Natural History & Prognosis

- Most cases mild, limited to SIJs
- Some progress to extensive spine ankylosis over period of years
- Often see exacerbations and remissions

Treatment

- TNF-α inhibitors effective and have rapid onset
- NSAIDs also commonly utilized

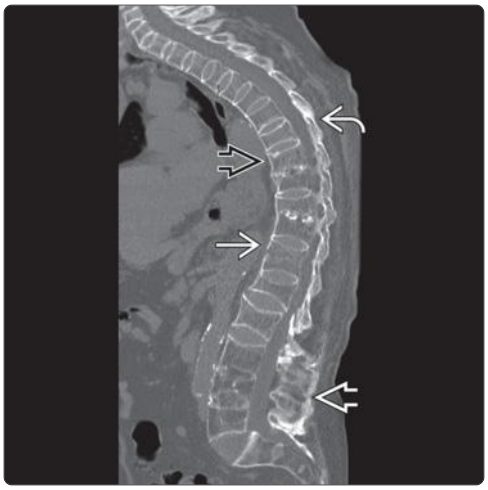
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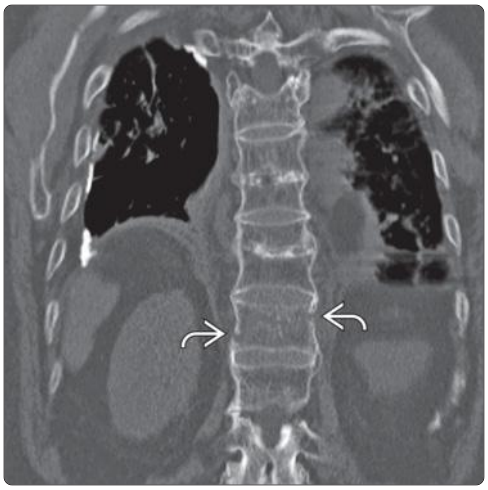
Image Interpretation Pearls

- Look for loss of definition of subchondral bone plate at SIJs as earliest sign
- If SIJs normal, cannot make imaging diagnosis of seronegative spondyloarthropathy

SELECTED REFERENCES

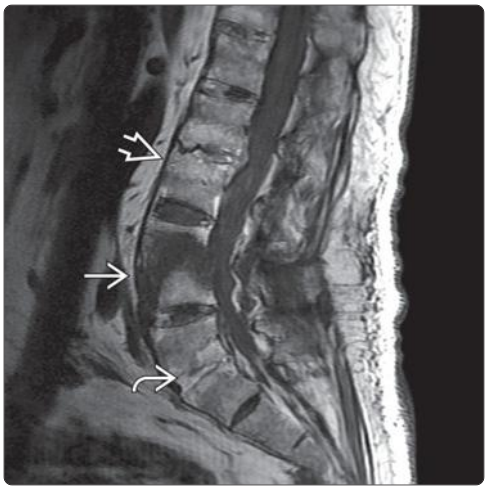
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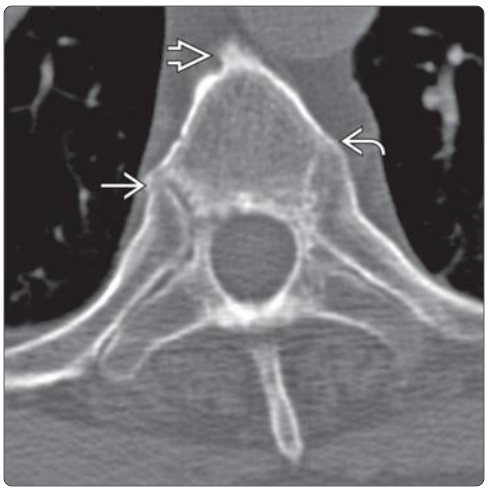


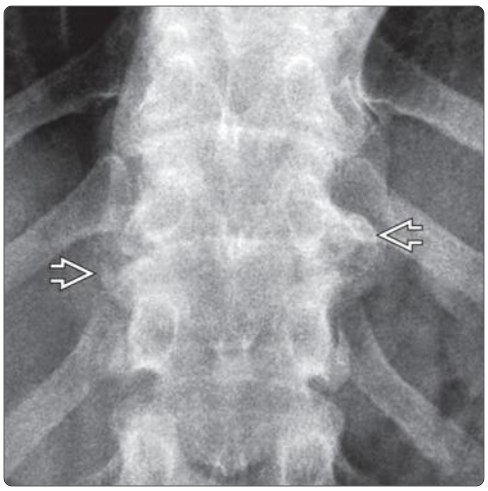
(Left) *Sagittal bone CT shows T12 acute fracture in a patient with nearly complete spine ankylosis. Note the facet joint fusion as well as thin syndesmophytes anteriorly and spinous process fusion. There is mild wedging of L1 and L2, which are age indeterminate.* **(Right)** *Coronal bone CT in the same patient shows a vertebral body fracture. Note pulmonary interstitial fibrosis. Bone is severely osteopenic.*





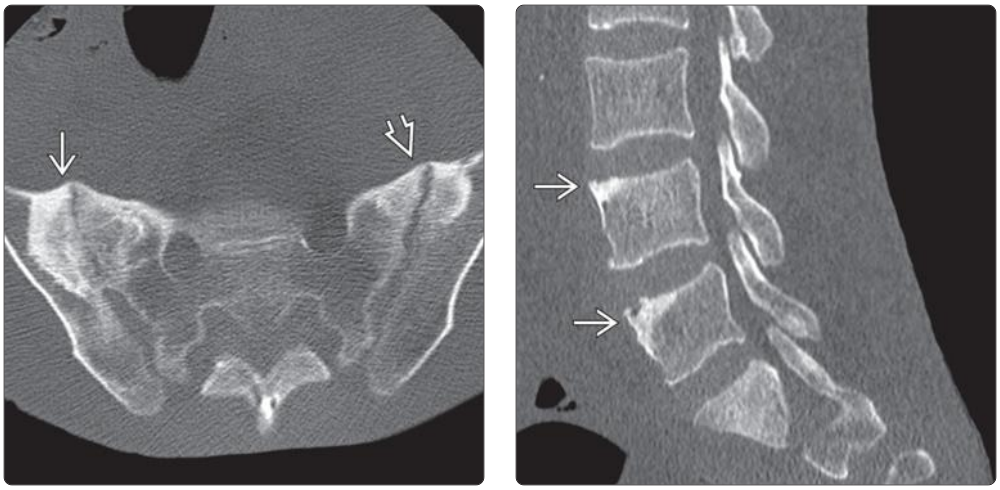
(Left) *Sagittal lumbar bone NECT shows a 3-column distraction/Chance-type fracture through the L4-5 disc extending obliquely through the L4 body and posterior elements.* **(Right)** *Sagittal lumbar T1W MR in this AS patient shows a 3-column injury with distraction of the L4 body components. Note the anterior fusion seen as linear low signal. Discs show increased signal due to fusion with fatty marrow.*



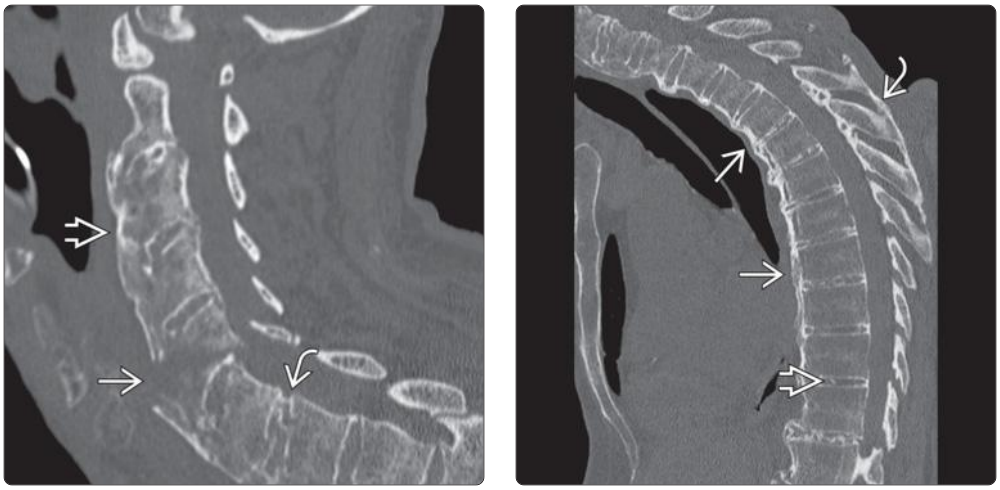


(Left) *Axial bone CT in a patient with reactive arthropathy shows fusion of left costovertebral joint and erosions at right costotransverse joint. Ossification is present in the anterior longitudinal ligament.* **(Right)** *Anteroposterior radiography shows bulky lateral paraspinous ligamentous ossifications in a patient with psoriatic arthritis. DISH has a similar appearance but spares the sacroiliac joints.*

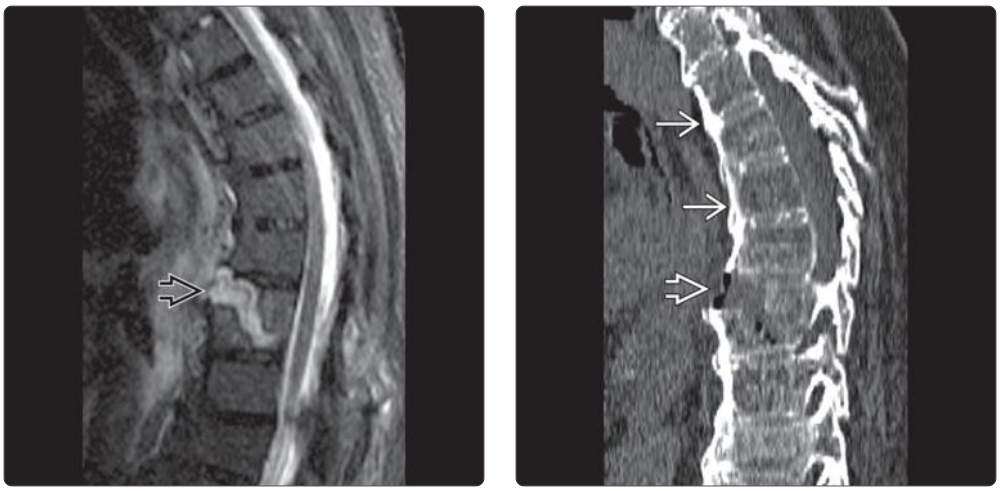
(Left) *Axial bone NECT through the SI joints shows marked bilateral sclerosis, right-sided fusion ⇨ of the SI joint, and left-sided erosive changes ⇨ in this patient with AS.* **(Right)** *Sagittal bone NECT in a patient with axial spondyloarthropathy shows sclerosis of the inferior endplates and vertebral body corners at L4 and L5 ⇨, reflecting the anterior inflammatory lesions of ankylosing spondylitis (Romanus lesions).*



(Left) *Sagittal bone NECT of the cervical spine shows a fracture extending through the C5 body ⇨. There is posterior displacement of the superior aspect of the fracture indicating a hyperextension mechanism. Note the typical anterior ossification of AS ⇨ and the extensive intervertebral disc calcification ⇨.* **(Right)** *Sagittal bone NECT of the thoracic spine in a patient with AS shows vertically oriented syndesmophytes ⇨, disc calcification ⇨, and extensive supraspinous ligament ossification ⇨.*



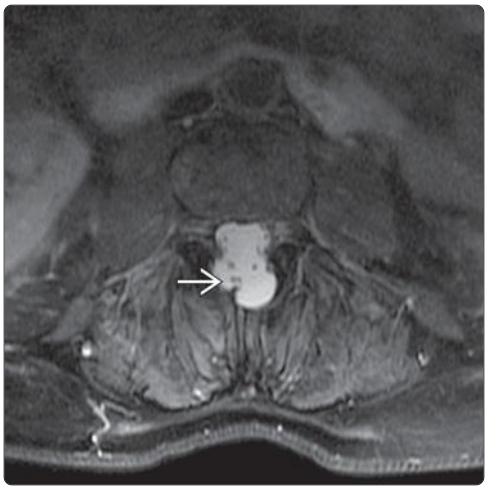
(Left) *Sagittal T2 MR shows the typical appearance of a chronic pseudoarthrosis in a patient with longstanding AS. There is an oblique increased signal across the disc and a vertebral body with sharp margins ⇨.* **(Right)** *Sagittal bone NECT shows longstanding ankylosing spondylitis with anterior syndesmophytes ⇨.There is a pronounced oblique fracture involving the lower thoracic spine with angulation ⇨ with a small amount of gas.*

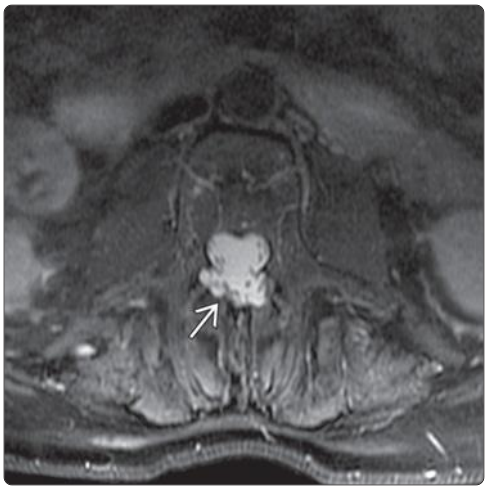






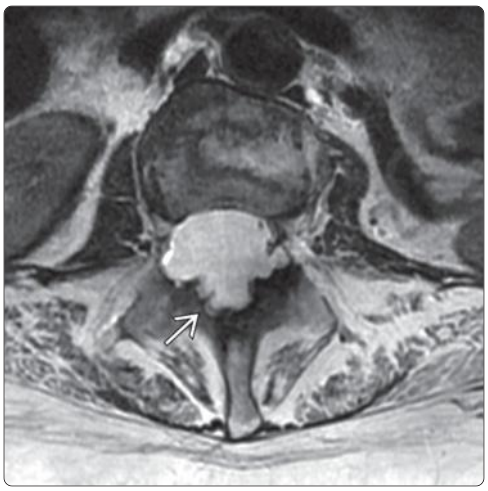
(Left) *Sagittal T1WI MR shows a lobulated contour to the dorsal dural margin with small outpouchings (dorsal dural diverticula) ⇒. There is squaring of the vertebral bodies and disc calcification reflecting chronic AS. Lumbar thecal sac distortion, as seen in this case, may be seen with cauda equina syndrome.* **(Right)** *Sagittal T2WI MR shows lobulation of the dorsal dural margin ⇒ in this patient with chronic AS. No normal nerve roots are seen in the distal thecal sac due to chronic arachnoiditis related to AS inflammation.*





(Left) *Axial T2 TSE FS MR shows the multiple dorsal dural diverticula and remodeling of the posterior elements with dorsal displacement of multiple nerve roots ⇒.* **(Right)** *Axial T2 TSE FS MR shows the multiple nerve roots, which are posteriorly displaced and adhesed ⇒ onto the dorsal dural diverticula in this patient with chronic AS and cauda equina syndrome.*





(Left) *Sagittal T2 TSE FS shows squaring of the vertebral bodies and diffuse disc calcification from chronic AS. The roots of the cauda equina are tethered posteriorly to the dural margin ⇒. The underlying mechanism is poorly understood, but may reflect the inflammation of the posterior elements during the more active phase of the disease.* **(Right)** *Axial T2W MR shows nerve roots tethered into the dorsal irregularly-shaped dural margin. Dorsal dural diverticula ⇒ extend into the remodeled lamina.*

Neurogenic (Charcot) Arthropathy

KEY FACTS

TERMINOLOGY

- Destructive arthropathy occurring when pain and proprioception are impaired while joint mobility is maintained

IMAGING

- Almost always in lumbar spine
- Vertebral endplate destruction
- Facet joint destructive arthropathy
- Preserved bone density
- Nonunited fractures
- Bony debris around vertebrae
- Vertebral subluxations
- Soft tissue mass, which may be large
- Heterogeneous enhancement
  - Both of affected vertebrae and soft tissue mass

TOP DIFFERENTIAL DIAGNOSES

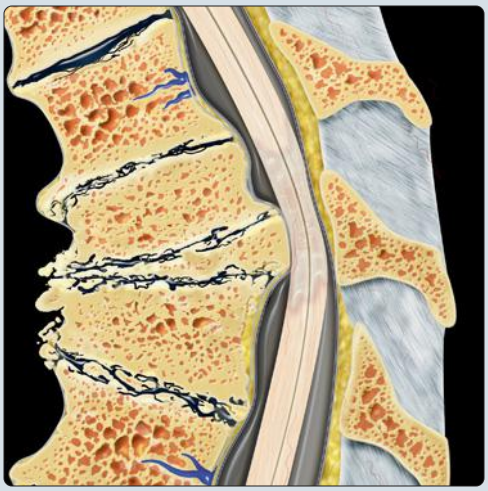
- Pyogenic infection
- Atypical infection: Tuberculosis, fungi
- Soft tissue sarcoma
- Bone tumor
- Degenerative disc disease (DDD) with instability

CLINICAL ISSUES

- Instability of spine
- Spine deformity
- Treated with rigid internal fixation and fusion of affected spinal levels

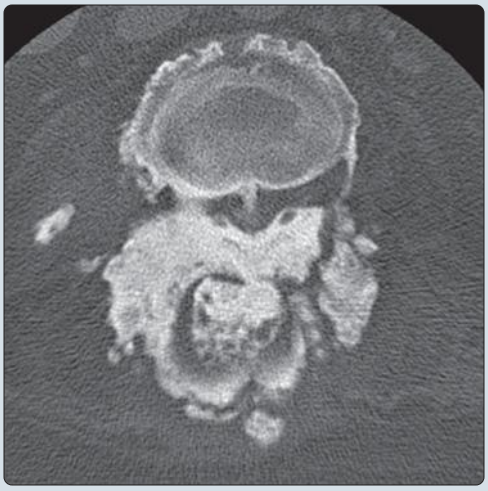
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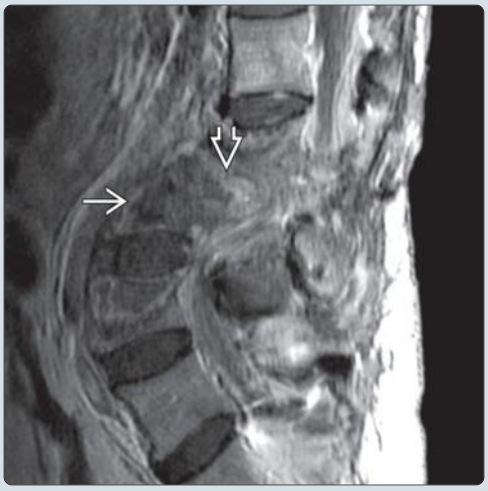
- Distinction between neuropathic joint and infection is one of the most difficult problems radiographically
- Only 2 processes can destroy joint in 1 month: Infection and neurogenic arthropathy





(Left) *Sagittal graphic shows arthropathy centered at 3 adjacent discs with endplate erosions, bony debris, and deformity.* **(Right)** *Sagittal NECT shows Charcot joint with pseudoarthrosis at L2-L3 and remodeling of adjacent vertebral bodies with extensive hypertrophic bone production about the posterior elements. Note the posterior retrolisthesis of the spinal canal at the L2-L3 level. Note the linear calcification involving the dura in the lower thoracic spine from arachnoiditis.*





(Left) *Axial NECT shows the typical massive bone production associated with neuropathic pseudoarthrosis.* **(Right)** *Sagittal T1 C+ MR in a paraplegic with cord transection shows heterogeneous enhancement at destructive arthropathy at L2-L3 with posterior subluxation, with central fluid filled areas surrounded by enhancing reparative tissues. L3-L4 is affected to a lesser extent. The striking soft tissue abnormalities seen on MR of Charcot arthropathy often raise concern for infection or sarcoma.*

TERMINOLOGY

Abbreviations

- Neuropathic spine, neuropathic arthropathy

Definitions

- Destructive arthropathy occurring when pain and proprioception are impaired while joint mobility is maintained

IMAGING

General Features

- Best diagnostic clue
  - Florid destruction of discs and facet joints with preserved bone density, involving 1-2 spinal levels
- Location
  - Almost always in lumbar spine
  - Sometimes occurs in lower thoracic spine

Radiographic Findings

- Radiography
  - Early stages mimic severe degenerative disc disease
    - Disc space narrowing
    - Discogenic sclerosis
    - Large osteophytes, often much larger than in degenerative disc disease
    - Vacuum phenomenon
  - Vertebral endplate destruction
  - Facet joint destructive arthropathy
  - Preserved bone density
  - Nonunited fractures
  - Bony debris around vertebrae
    - Small, nonunited fracture fragments
  - Vertebral subluxations
  - Often rapidly progressive
  - Disorganization

CT Findings

- Bone CT
  - Destruction of vertebral endplates, facet joints
    - Erosions and bone cysts common
    - Endplate involvement usually less uniform than seen in infection
  - Bony debris and nonunited fractures
  - Soft tissue mass, which may be large
    - Enhances with contrast administration

MR Findings

- Abnormalities centered at disc
  - Destruction of endplates
  - Bone marrow edema (↓ on T1WI, ↑ on T2WI, STIR)
  - Enhancement with Gd in disc and vertebral bodies
- Abnormalities centered at facet joints
  - Destruction of subchondral bone plate
  - Joint effusion
  - Bone marrow edema
  - Enhancement with Gd around facet joints
- Soft tissue mass
  - Variably present
  - Heterogeneous signal intensity
    - Predominantly ↓ on T1WI, ↔ ↑ on T2WI, STIR
  - Bone debris within mass often difficult to see
- Fluid collections
  - Variably present
  - Rim enhancing
  - Mimic abscess

Nuclear Medicine Findings

- Bone scan
  - Positive on all 3 phases
- PET
  - Uptake with FDG
- Gallium scan
  - Positive
- WBC scan
  - Positive in active neuropathic joint

Imaging Recommendations

- Best imaging tool
  - CT: Preserved bone density, bony debris best seen on CT, helps distinguish from infection
- Protocol advice
  - Thin-section CT with reformatted images in coronal and sagittal planes

DIFFERENTIAL DIAGNOSIS

Pyogenic Infection

- Bones usually osteopenic but occasionally see sclerosing variant
- Destruction of adjacent endplates, intervening disc
- Rarely see cloaca, sequestrum in bone
- Paraspinous soft tissue abscess shows rim enhancement
- Uncommon to destroy both discs and facet joints except in patients with failed fusion
- Fever, elevated ESR and C-reactive protein useful
- Cultures negative in 50%

Atypical Infection: Tuberculosis, Fungi

- Bone density preserved
- Paraspinous soft tissue mass
- Amorphous calcifications in intraosseous and soft tissue cold abscesses
- Cultures often negative
- Pulmonary tuberculosis may be absent
- Rely on histology rather than culture for diagnosis

Soft Tissue Sarcoma

- Malignant fibrous histiocytoma
- Osteosarcoma
- Reactive spindle cells in soft tissue mass of Charcot joint may be misdiagnosed as sarcoma by unwary pathologists
- Centered in soft tissues
- Soft tissue sarcomas rarely destroy intervertebral discs or facet joints

Bone Tumor

- Rare to involve 2 adjacent vertebrae
- Tends to spare disc space

Degenerative Disc Disease (DDD) With Instability

- Severe DDD may show endplate erosion, especially if instability present

- Erosion similar to very early neuropathic joint; however, other signs of neuropathic joint absent
- DDD slowly progressive

PATHOLOGY

General Features

- Etiology
  - Occurs when there is preserved mobility with impaired proprioception and pain
    - Because proprioception is impaired, joint is stressed beyond normal range of motion
    - Because pain is diminished or absent, patient does not self-protect following minor injury
    - Abnormal joint motion results in
      - Disruption of joint capsule and stabilizing ligaments
      - Microfractures &/or major fractures
    - Fractures are not immobilized, leading to
      - Exuberant, nonbridging peripheral callus formation
      - Joint subluxation &/or dislocation
      - Soft tissue reparative response
      - Fracture nonunion
      - Bone debris
      - Preserved bone density (i.e., no disuse osteopenia)
- Associated abnormalities
  - Most common cause: Neuropathy due to diabetes mellitus
  - Neurosyphilis is cause originally described by Charcot
    - More commonly involves hips, knees than spine
  - Sequela of traumatic paraplegia
    - Paraplegic patients who are physically active put stress on insensate spine
    - Arthropathy occurs > 5 years following traumatic paraplegia
    - Below level of sensation loss
  - Congenital insensitivity to pain
  - Other causes or neuropathic joint seen in peripheral joints but not in spine

Gross Pathologic & Surgical Features

- Bone sclerosis due to
  - Affected level continues to be stressed (i.e., no disuse osteopenia)
  - Callus formation
  - Bone debris

Microscopic Features

- Bone and cartilage fragments
- Histiocytes and lymphoplasmacytic infiltrate
- Fibrosis, granulation tissue without acute inflammation
- Mimics chronic infection or sometimes neoplasm

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Instability of spine, sometimes with audible "clicking"
  - New onset spine deformity
  - Common misconception is that pain must be absent to make diagnosis of neuropathic joint
    - Pain is, however, less severe than expected for severity of joint destruction
- Clinical profile
  - Diabetic patient with nonspecific back pain
  - Paraplegic patient complaining of instability

Natural History & Prognosis

- Most cases do well following fusion
- May become superinfected

Treatment

- Options, risks, complications
  - Rigid internal fixation and fusion of affected spinal levels
  - Can also be treated with bracing but success limited

DIAGNOSTIC CHECKLIST

Consider

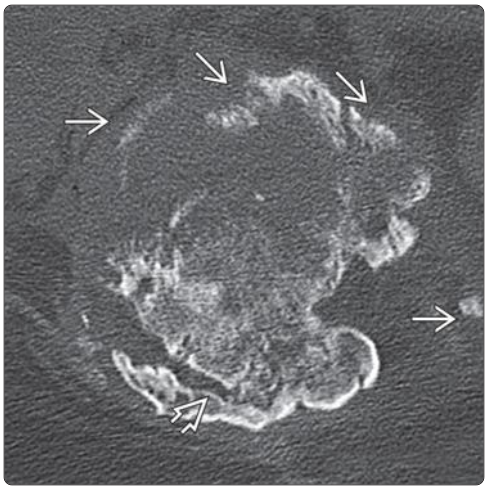
- Distinction between neuropathic joint and infection is one of most difficult problems radiographically
- Important differential diagnostic consideration for infection when patient has
  - Predisposing cause for neuropathic joint
  - No systemic signs of infection
  - Negative cultures (percutaneous or open)
  - Preserved bone density, disorganization, and bone debris

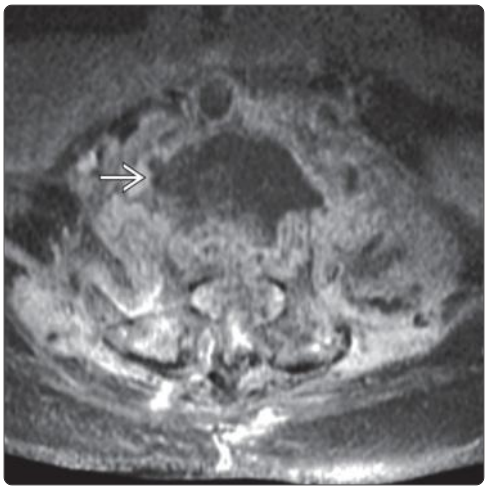
Image Interpretation Pearls

- Only 2 processes can destroy joint in 1 month: Infection and neurogenic arthropathy
- Nuclear medicine studies unreliable in differentiation of neuropathic joint from infection

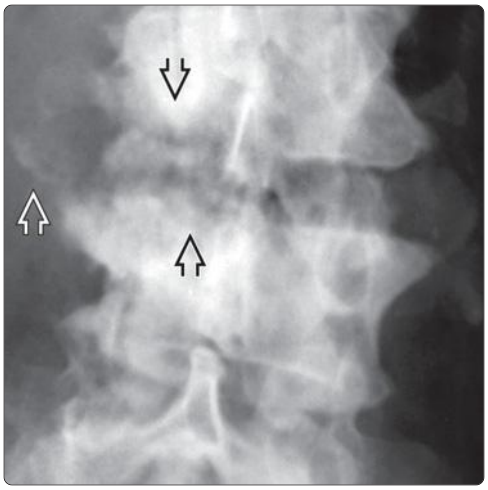
SELECTED REFERENCES

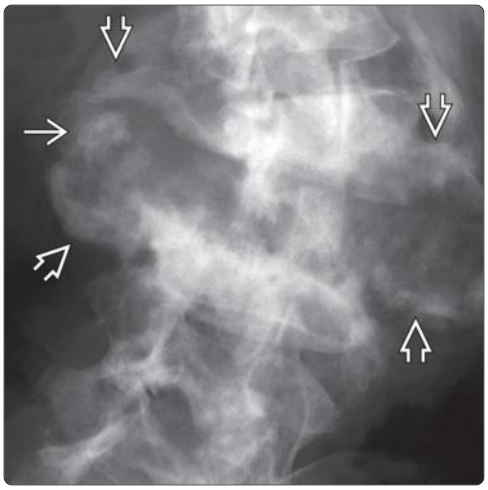
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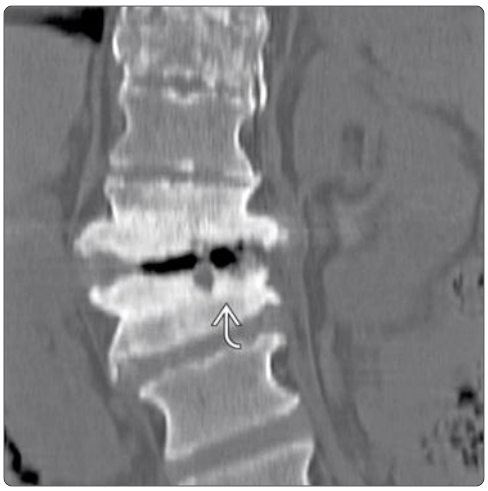
(Left) *Axial bone CT shows bone almost obliterating spinal canal ➡ in neuropathic joint due to traumatic paraplegia. Multiple nonunited bone fragments ➡ create bony debris. Bone density is preserved.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows fluid collection ➡ surrounded by heterogeneously enhancing soft tissue. There is also heterogeneous enhancement in the bone. Infection was excluded by open cultures, laboratory findings, and follow-up examinations.*





(Left) *Anteroposterior radiography shows early neuropathic joint. Clues to diagnosis are focal endplate destruction ➡ and bony debris ➡. Infection usually involves the endplate more uniformly and tends to cause osteopenia.* **(Right)** *Anteroposterior radiography in the same patient 2 months later shows rapid progression of neuropathic joint 1. Note scoliosis and subluxation, florid osteophytes ➡, bone debris ➡, and bony destruction centered on disc.*





(Left) *Sagittal bone CT shows a less severe neuropathic joint. Clues that this is more than degenerative disc disease are the history of paraplegia, the involvement of both disc and facet joints, and the bony remodeling ➡ due to instability. Clues that this is not infection include the extensive gas in the disc and the preserved bone density.* **(Right)** *Coronal bone CT in the same patient shows large erosions ➡ involve only a portion of the endplate. Infection usually involves the entire endplate.*

Hemodialysis Spondyloarthropathy

KEY FACTS

TERMINOLOGY

- Destructive spondyloarthropathy (DSA)
- Discocentric destructive arthritis in patient on long-term hemodialysis

IMAGING

- Vertebral and endplate destruction in patient on long-term hemodialysis, ± vertebral collapse
  - Cervical, thoracic, or lumbar spine
  - Often involves multiple levels
- Endplate destruction with sharply marginated erosions
- Amorphous material in disc, spinal canal, &/or prevertebral soft tissues
  - Usually lower signal intensity than infection on T2WI, STIR

TOP DIFFERENTIAL DIAGNOSES

- Infection
- Neuropathic joint
- Gout
- Calcium pyrophosphate deposition disease

PATHOLOGY

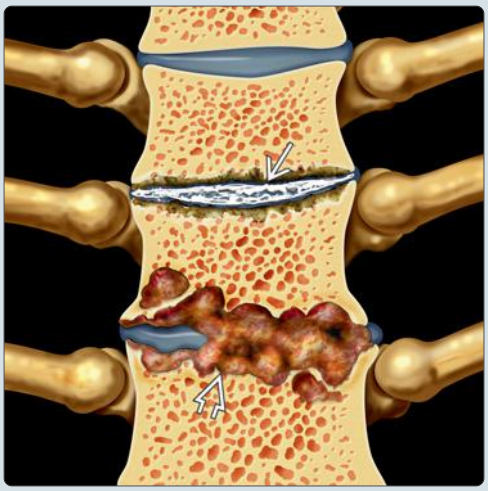
- 2 types of arthropathy associated with hemodialysis
  - Amyloid deposition: β-2 microglobulin
  - Crystal deposition: Hydroxyapatite

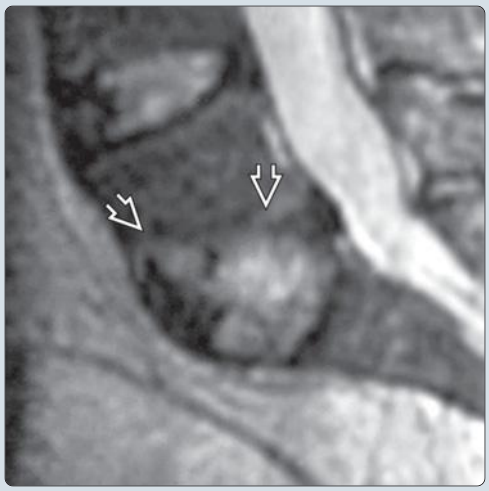
CLINICAL ISSUES

- Indolent
- May result in spinal instability
- Back pain, radiculopathy, cord compression

DIAGNOSTIC CHECKLIST

- Specimen must be sent for crystal analysis in saline or ethanol; formalin dissolves crystals
- History is key to making diagnosis





(Left) *Coronal graphic shows 2 types of hemodialysis arthropathy, crystal deposition and amyloid deposition, both of which may cause endplate erosions.* **(Right)** *Sagittal T2WI MR shows amyloid deposit at L5-S1 causing extensive erosion of the adjacent vertebral bodies, which may be confused for pyogenic infection or even metastatic disease when asymmetric endplate involvement occurs.*





(Left) *Sagittal T1WI MR shows severe bone destruction centered at the C4-C5 disc, with smaller, "punched out" erosions at C5-C6 and C6-C7. Patient was on hemodialysis and had chronic, worsening neck pain and findings consistent with cord compression. Soft tissue posteriorly could represent discs or crystal deposition.* **(Right)** *Sagittal STIR MR of the same patient shows severe cord compression. Of note, discs and masses remain very low signal intensity, unlike pyogenic infection.*

TERMINOLOGY

Synonyms

- Destructive spondyloarthropathy (DSA)

Definitions

- Discocentric destructive arthritis in patient on long-term hemodialysis

IMAGING

General Features

- Best diagnostic clue
  - Vertebral and endplate destruction in patient on long-term hemodialysis, ± vertebral collapse
- Location
  - Cervical, thoracic, or lumbar spine
  - Often involves multiple levels

Radiographic Findings

- Radiography
  - Endplate destruction
  - Soft tissue mass
    - May contain amyloid
    - May contain crystals with visible calcification

CT Findings

- Sharply marginated endplate erosions
- Soft tissue mass
  - ± calcifications
  - Vertebral body collapse

MR Findings

- Vertebral endplate destruction
- Abnormal signal intensity in vertebral bodies
  - High signal intensity on T2WI, STIR mimics infection
- Amorphous material in disc, spinal canal, &/or prevertebral soft tissues
  - Usually lower signal intensity than infection on T2WI, STIR

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan

Imaging Recommendations

- Best imaging tool
  - MR
  - Differentiation from active infection may require biopsy

DIFFERENTIAL DIAGNOSIS

Infection

- Endplate destruction, adjacent soft tissue abscess
- High signal intensity in disc on T2WI, STIR
- Calcifications absent

Neuropathic Joint

- Lumbar spine
- Bone debris, subluxations prominent

Gout

- Imaging appearance may be identical

Calcium Pyrophosphate Deposition Disease

- Disc, ligamentum flavum linear calcifications
- Endplate destruction, erosions

PATHOLOGY

General Features

- Etiology
  - 2 types of arthropathy associated with hemodialysis
    - Amyloid deposition: β-2 microglobulin
      - Less common today due to advances in hemodialysis
    - Crystal deposition: Hydroxyapatite
- Associated abnormalities
  - Adynamic bone disease
    - Absolute or partial parathyroid hormone deficit associated with low or absent bone remodeling
    - Patients often present with bone fractures and collapse, myopathy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic unless severe
  - Other signs/symptoms
    - Back pain, radiculopathy, cord compression

Demographics

- Epidemiology
  - Uncommon, incidence increases with length of time on dialysis

Natural History & Prognosis

- Indolent, may result in instability

Treatment

- Options, risks, complications
  - Fusion for stabilization

DIAGNOSTIC CHECKLIST

Consider

- Specimen must be sent for crystal analysis in saline or ethanol; formalin dissolves crystals

Image Interpretation Pearls

- History is key to making diagnosis

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Ankylosing Spondylitis

KEY FACTS

IMAGING

- Location
  - Discs and synovial joints of spine
  - Joints of axial skeleton, less commonly peripheral joints
  - Tendon and ligament attachments (entheses)
- Sacroiliac joints: Bilaterally symmetric erosive arthropathy → fusion
- Facet, uncovertebral joints: Erosions → fusion
- Squaring of vertebral bodies → corner erosions → "shiny corner" (corner sclerosis) → ankylosis
- Erosions, new bone formation at tendon and ligament attachments
- Trauma
  - Often hyperextension injury, involving all 3 columns of spine
  - Fracture may occur through fused regions
  - MR should be considered to exclude occult fracture, as these may be subtle secondary to osteopenia

TOP DIFFERENTIAL DIAGNOSES

- DISH
- Psoriatic arthritis and reactive arthropathy
- Rheumatoid arthritis
- Osteitis condensans ilii

PATHOLOGY

- Associated findings
  - Pulmonary interstitial fibrosis, decreased chest excursion, uveitis, aortitis, aortic valve insufficiency
- 90% of patients HLA-B27 positive

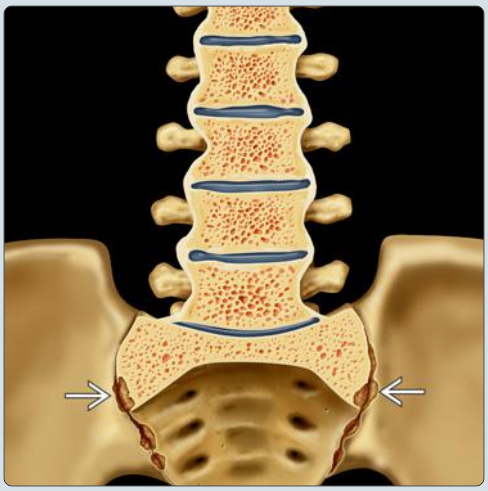
CLINICAL ISSUES

- Complications: Kyphotic deformity, fractures of fused spine
- Pain centered over SIJs, back stiffness

DIAGNOSTIC CHECKLIST

- Normal SIJs preclude imaging diagnosis of AS

(Left) *Graphic shows sacroiliac joint (SIJ) erosions. Erosions are best seen in the inferior 1/3 of the joint, which is primarily synovial. More superiorly, the articulation is primarily ligamentous, with only a small synovial portion, and erosions are difficult to see.* **(Right)** *Axial bone CT shows classic appearance of SIJ inflammatory arthropathy: Small erosions, blurring of the subchondral bone plate, and adjacent sclerosis. Findings are more severe on the iliac side of joint.*





(Left) *Anteroposterior radiograph shows the "bamboo spine" typical of advanced Ankylosing Spondylitis (AS), where ossification of the outer fibers of the annulus fibrosus mimics the joints of the bamboo plant. Fusion of the costotransverse joints and between the spinous processes is also visible.* **(Right)** *Lateral radiograph shows the shiny corner sign. Bone resorption beneath the anterior longitudinal ligament creates a squared vertebral body contour. A single, thin syndesmophyte is visible.*





TERMINOLOGY

Definitions

- Syndesmophyte: Paraspinous ligamentous or disc ossification bridging 2 adjacent vertebral bodies

IMAGING

General Features

- Location
  - Intervertebral discs
  - Synovial joints of spine: Facet, uncovertebral, costotransverse, costovertebral
  - Joints of axial skeleton
  - Tendon and ligament attachments (entheses)
    - Plantar fascia origin erosions common

Radiographic Findings

- **Sacroiliac joints: Sacroiliitis**
  - Bilaterally symmetric erosive arthropathy
  - Findings seen on iliac side of joint before sacral side
  - Early: Loss of definition of subchondral bone plate
  - Later: Erosions, widened joint space
    - Erosions often small, indistinct
  - Later: Sclerosis surrounding joint
  - Endstage: Fusion of joint
- **Vertebral bodies**
  - Earliest: Squaring of vertebral bodies → corner erosions → "shiny corner" (corner sclerosis)
  - Later: Thin syndesmophytes bridge vertebrae
  - End stage: Widespread ankylosis ("bamboo spine")
- **Facet, uncovertebral joints**
  - Erosions → fusion
- **Enthesopathy**
  - Erosions, periosteal new bone formation at tendon and ligament attachments
- **Osteopenia**

CT Findings

- Same findings as radiograph, but ↑ sensitivity

MR Findings

- Bone marrow edema: Low signal T1, high signal T2 and STIR
  - Sacroiliac joints
  - Corners of vertebral bodies, facet joints
- Joint effusions
- Erosions: Loss of subchondral bone plate, irregular margins of joint
- Squaring of vertebral bodies
- Ankylosis not a striking MR finding
  - Syndesmophytes are thin, low signal intensity on all sequences

Imaging Recommendations

- Best imaging tool
  - MR and CT have good sensitivity for sacroiliitis

Imaging of Trauma in AS

- C2, cervicothoracic junction, thoracolumbar junction
  - Often a hyperextension injury, involving all 3 columns of spine
  - Fracture may be through vertebral body or fused disc space
  - MR should be considered to exclude occult fracture, as these may be subtle secondary to osteopenia

DIFFERENTIAL DIAGNOSIS

DISH

- Bulky ossification of paraspinous ligaments
- SIJs usually normal, rarely fused by bulky osteophytes

Rheumatoid Arthritis

- Ankylosis not a feature
- Involves synovial joints, spares discs
- Primarily in cervical spine

Psoriatic Arthritis and Reactive Arthropathy

- Bulky ossification of paraspinous ligaments
- SIJ involvement often asymmetric

Osteitis Condensans Ilii

- Sclerosis adjacent to SIJ
- Subchondral bone plate preserved, no erosions
- Joint space normal or narrow

Infection

- Unilateral sacroiliitis

PATHOLOGY

General Features

- Associated abnormalities
  - Pulmonary interstitial fibrosis, decreased chest excursion, uveitis, aortitis, aortic regurgitation
  - 90% of patients HLA-B27 positive

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain centered over SIJs, back stiffness
- Other signs/symptoms
  - Kyphotic deformity, fracture

Treatment

- Corticosteroids
- Disease modifying agents (DMARDs): Tumor necrosis factor inhibitors, sulfasalazine, methotrexate
  - Early initiation of treatment helps prevent ankylosis

DIAGNOSTIC CHECKLIST

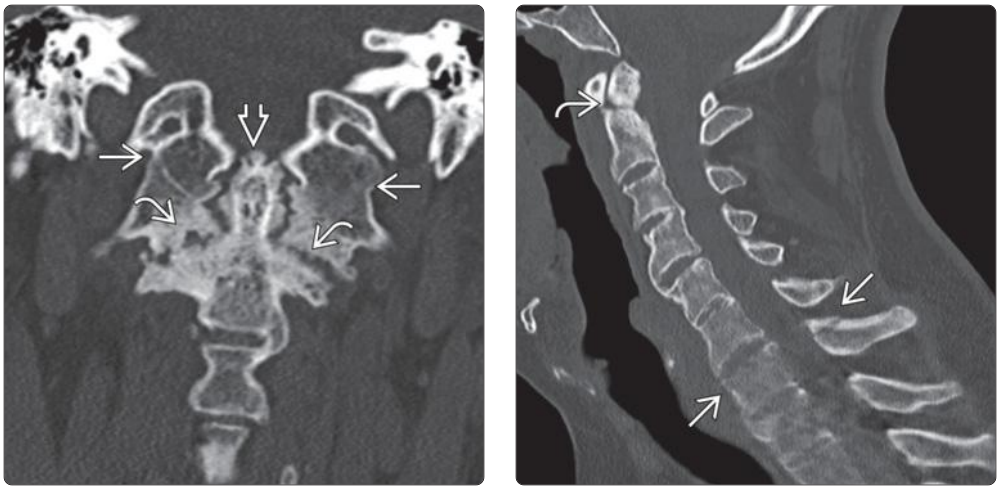
Image Interpretation Pearls

- Normal SIJs preclude imaging diagnosis of AS

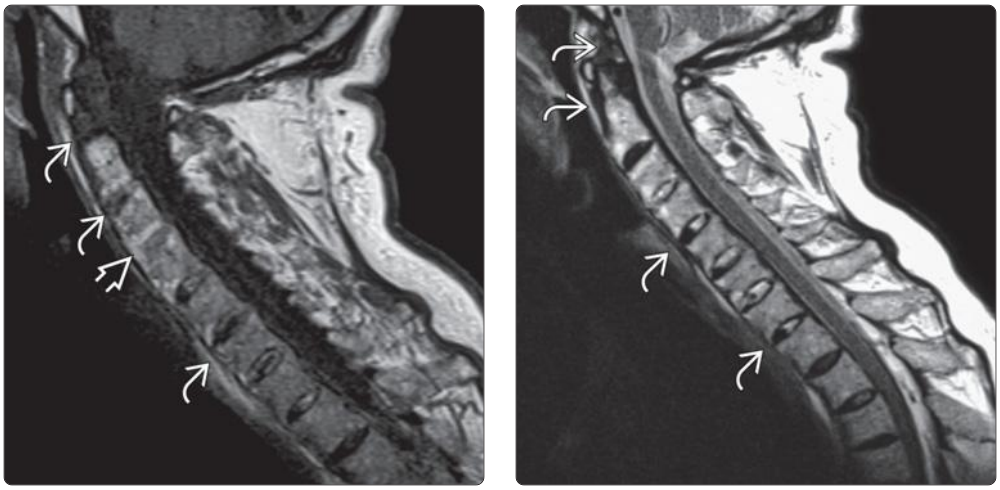
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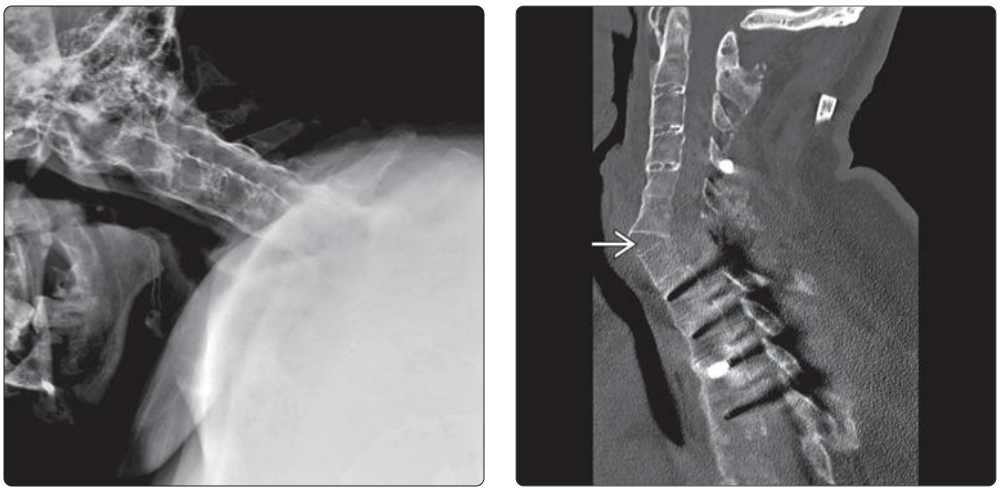
(Left) *Coronal bone CT shows erosions and sclerosis at the C1-2 facet joints. There is extensive periosteal reaction (enthesopathy) surrounding the dens. Occiput-C1 joints are fused.* **(Right)** *Sagittal bone CT shows fractures at C2 and C7 in a patient with AS. Fractures in this population are often multilevel and difficult to see on CT because of osteopenia. Fracture of the C7 spinous process alerts the radiologist to look for a fracture of the anterior column.*

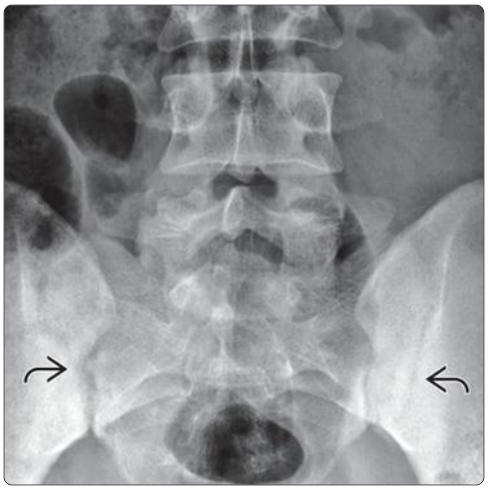


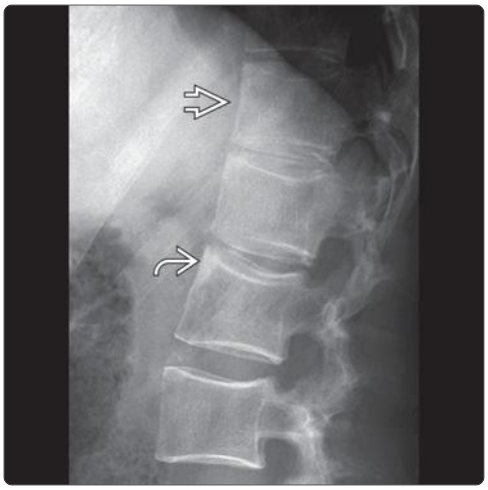
(Left) *Sagittal T1WI MR shows the characteristically unimpressive MR appearance of syndesmophytes. At C3-4 the entire disc has ossified.* **(Right)** *Sagittal T2WI MR in a different patient shows ligamentous ossification extending from the clivus to the thoracic spine. Note the upper thoracic kyphosis. Spinal deformity in patients with AS may preclude use of a cervical MR coil, and flexible surface coils are often a better option.*



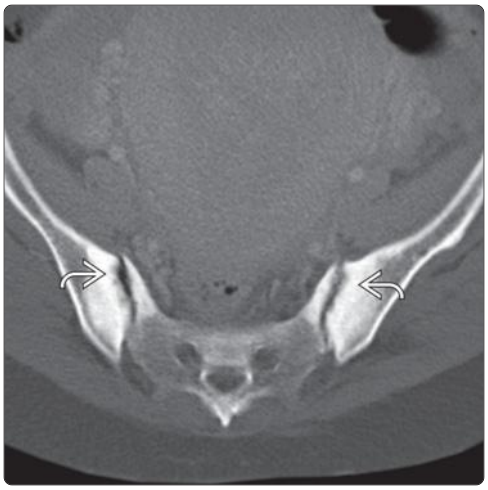
(Left) *Lateral radiograph in a 46-year-old woman shows fixed deformity from AS. Vertebral bodies and facet joints are fused, with loss of cervical lordosis. Thoracic kyphosis is severe, and the patient's chin was on her chest. Spinal deformity of AS can be debilitating. Patient also had limited chest expansion due to ankylosis of rib articulations.* **(Right)** *Sagittal bone CT shows the same patient after extension osteotomy and posterior fusion. A focal lordosis has been created at C6-7.*





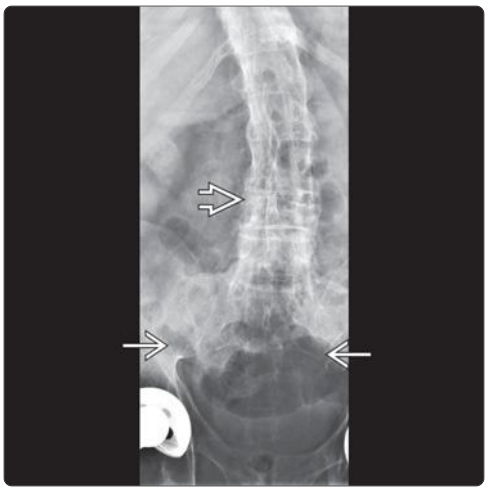


(Left) *Anteroposterior radiograph shows SIJ sclerosis. The subchondral bone plate is poorly defined, but erosions are subtle. Sacroiliac joints are difficult to evaluate on radiograph. CT or MR may be performed to improve early diagnosis and allow prompt DMARD treatment.* **(Right)** *Lateral radiograph shows "shiny corner" sign at L1 and L2. There is squaring of the vertebral body contour of T12-L3. AS initially involves the SIJs, then typically skips to the thoracolumbar junction.*





(Left) *Axial bone CT shows bilaterally symmetric sacroiliitis. Small erosions are seen with widening of the joint and reactive sclerosis. Osteitis condensans ilii, in contrast, shows bone sclerosis without erosions or joint space widening.* **(Right)** *Axial T2WI MR shows bilateral sacroiliac joint erosions. Bone marrow edema is slightly more prominent on the left. As is typical, the erosions are more severe on the iliac side of the joint.*





(Left) *Anteroposterior radiograph shows fused SIJs and bamboo spine. Fracture of L4 is difficult to see because of severe osteopenia. Hip arthroplasties were performed for advanced AS.* **(Right)** *Lateral radiograph in the same patient shows hyperextension-type fracture of L4. The rigidity of the ankylosed spine renders it vulnerable to fracture. Fractures may occur through syndesmophytes or, as in this case, through the vertebral body or ossified disc space.*

CPPD

KEY FACTS

TERMINOLOGY

- Crowned dens syndrome: Pain due to CPPD deposition around dens

IMAGING

- Soft tissue calcifications
  - Usually linear, occasionally globular
  - Seen in ligaments, discs, facet joint capsules, hyaline cartilage
  - Horseshoe-shaped calcification around dens
- Erosions of odontoid process, vertebral endplates
  - Usually sharply demarcated, often corticated
- MR findings nonspecific
  - Calcium usually not visible, low signal intensity on all sequences
  - Soft tissue mass surrounding dens
  - Erosions of dens, vertebral endplates

TOP DIFFERENTIAL DIAGNOSES

- Degenerative disc disease
- Rheumatoid arthritis, adult
- Osteomyelitis, pyogenic
- Seronegative spondyloarthropathy
- Hemodialysis arthropathy
- Hyperparathyroidism
- Gout

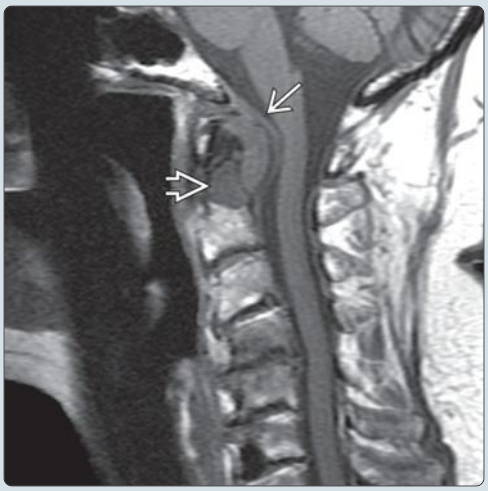
CLINICAL ISSUES

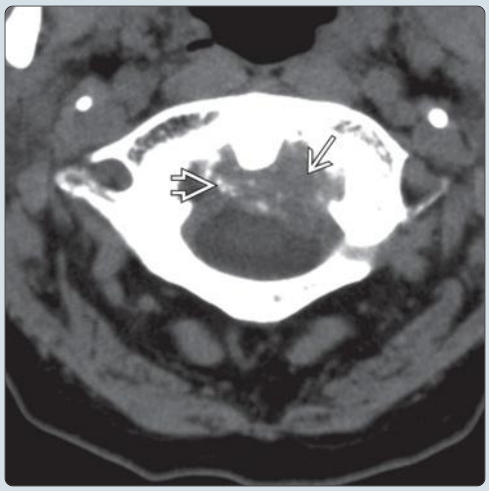
- Chronic back pain
- Acute attack of pain ± fever ± radiculopathy
- Myelopathy due to cord compression

DIAGNOSTIC CHECKLIST

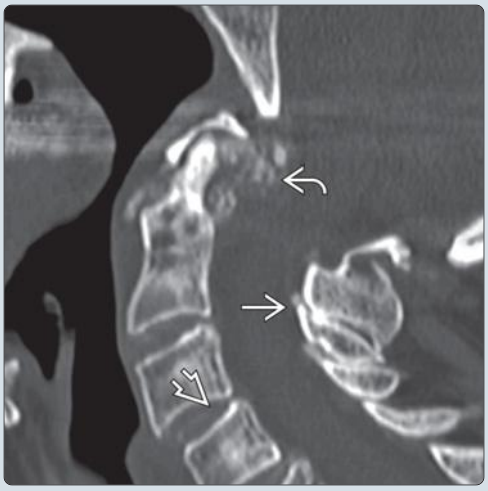
- CT useful in distinguishing from rheumatoid arthritis, which does not calcify

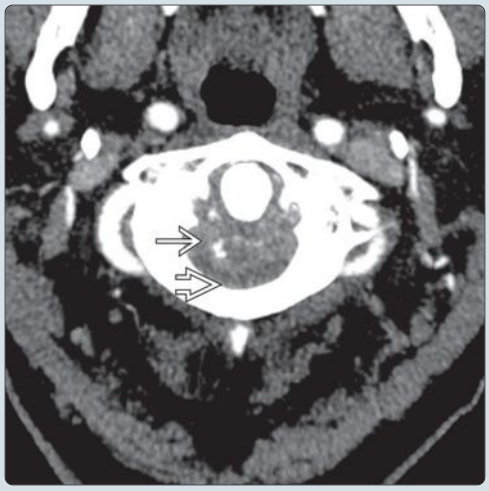
(Left) *Sagittal T1WI MR shows a large pseudopannus posterior to the odontoid process ⇨ with moderately severe effacement of the ventral thecal sac and mild mass effect upon the cervicomedullary junction. A large degenerative cyst is present in the odontoid process ⇨.* **(Right)** *Axial bone CT shows marked thickening of the retroodontoid soft tissues ⇨ with scattered calcifications ⇨ that are consistent with a diagnosis of CPPD.*





(Left) *Sagittal bone CT shows severe CPPD at the craniocervical junction. There is a large calcified mass posterior to the dens ⇨ and severe odontoid erosion. Calcification is seen in the ligamentum flavum ⇨ and intervertebral discs ⇨.* **(Right)** *Axial CTA study shows multiple punctate foci of calcification ⇨ within a large soft tissue mass posterior to the odontoid process, which causes cord compression ⇨.*





TERMINOLOGY

Abbreviations

- Calcium pyrophosphate dihydrate deposition (CPPD) disease

Definitions

- Pseudogout: Acute painful episode due to CPPD
  - Terms with "pseudo" should generally be abandoned in favor of CPPD
- Crowned dens syndrome: Pain due to calcium pyrophosphate crystal deposition around dens

IMAGING

General Features

- Best diagnostic clue
  - Linear calcific deposits in disc, ligament, hyaline cartilage, or joint capsule
- Location
  - Cervical, thoracic, or lumbar spine
    - Calcification in ligaments, discs, facet joint capsules, hyaline cartilage
  - Sacroiliac joints, pubic symphysis
  - Peripheral skeleton: Most common in knee and triangular fibrocartilage of wrist
  - Spine involvement often seen in absence of peripheral skeleton involvement

Radiographic Findings

- Radiography
  - Linear disc calcifications
  - Majority of cases also show calcification in pubic symphysis &/or triangular fibrocartilage of wrist

CT Findings

- Soft tissue calcifications
  - Usually linear, occasionally globular and mass-like
- Ligament calcification
  - Most commonly in PLL, ligamentum flavum
  - May result in spinal stenosis
- Facet joint effusions
- Hyaline cartilage calcification in facet and uncovertebral joints
- Erosions of vertebral endplates
  - Sharply demarcated, often corticated
- Crowned dens syndrome
  - Horseshoe-shaped calcification around dens
  - Erosion of dens, often severe
  - C1-C2 instability with flexion/extension

MR Findings

- Calcium low signal intensity on all sequences
  - May see punctate foci
  - Often not visible because isointense to discs, ligaments
- Intervertebral discs narrow and low signal intensity on all sequences
  - Indistinguishable from degenerative disc disease
- Ligament thickening
  - Most commonly PLL, ligamentum flavum
  - May result in spinal stenosis
- Erosions of dens, vertebral endplates
- Abnormal signal intensity in vertebrae
  - Usually mild
  - Centered on disc
  - T1 findings often more prominent than on T2, STIR
  - Minimal enhancement with gadolinium
- Soft tissue mass adjacent to vertebra
  - Low to intermediate signal intensity on all sequences
  - Minimal enhancement with gadolinium
- Facet joint effusions
- Variable enhancement with gadolinium administration

Nuclear Medicine Findings

- Bone scan
  - Positive on delayed images
  - Positive 3-phase scan in acute pseudogout attack

Imaging Recommendations

- Best imaging tool
  - CT scan
- Protocol advice
  - Thin-section CT with sagittal, coronal reformats

DIFFERENTIAL DIAGNOSIS

Degenerative Disc Disease

- Disc bulges and herniations may calcify when chronic
- Mild irregularity of vertebral endplate often occurs
  - Generally less severe than erosions of CPPD

Pseudopannus

- Thickening of tectorial membrane due to degenerative changes
  - Causes soft tissue fullness around dens
  - Retroodontoid soft tissue thickness in patients with CPPD crystal deposition greater than in patients without CPPD crystal deposition
- May be seen with abnormal motion secondary to more inferior cervical fusion

Rheumatoid Arthritis, Adult

- Odontoid erosions, soft tissue mass may appear identical to CPPD on MR
  - Absence of calcifications distinguishes from CPPD on CT, radiographs
- Spares intervertebral discs
- Look for subaxial erosive changes

Seronegative Spondyloarthropathy

- Ligament ossifications
- May appear identical to CPPD on MR

Osteomyelitis, Pyogenic

- Usually affects only 1 level
- High signal intensity in disc on T2WI, STIR
- Phlegmon or abscess often present in spinal canal or prevertebral soft tissues
- Infected areas show enhancement with gadolinium

Hemodialysis Arthropathy

- Endplate erosions
- Calcifications due to hydroxyapatite deposition

Hyperparathyroidism

- Osteopenia, endplate erosions, soft tissue calcifications
- Laboratory values most useful to distinguish from CPPD

Ochronosis

- Diffuse disc calcifications and degeneration

Gout

- Much less common in spine than CPPD
- Endplate erosions, soft tissue masses
- Calcification usually minimal
- Focal "punched-out" lesions typical

Hydroxyapatite Deposition Disease

- Rounded calcific deposits in soft tissues
- Uncommon in spine

PATHOLOGY

General Features

- Etiology
  - Crystal deposition of unknown etiology
- Associated abnormalities
  - Multiple crystals may be present in same patient
    - CPPD, gout, hydroxyapatite
  - Elderly patients may have both rheumatoid arthritis and CPPD
- Specimens must be sent to pathology in alcohol or saline
  - Crystals dissolve in formalin

Gross Pathologic & Surgical Features

- Chalky white, linear deposits of crystals
- Occasionally tumoral deposits of calcification develop
  - Less common, usually due to hydroxyapatite crystals in addition to CPPD

Microscopic Features

- Polarizing microscope shows crystals with weakly positive birefringence

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be asymptomatic
  - Chronic back pain
  - Acute attack of pain ± fever ± radiculopathy
  - Myelopathy due to cord compression
  - Acute neck pain

Demographics

- Age
  - Usually > 50 years old
- Epidemiology
  - Atlantoaxial CPPD seen in 12.5% of population
  - Prevalence increases with age
    - > 60 years: 34%
    - > 80 years: 50%
  - ANKH (human homologue of progressive ankylosis) mutations in familial CPPD disease

Treatment

- Options, risks, complications
  - Nonsteroidal anti-inflammatory agents
  - Some medications used to treat gout are also effective against CPPD
  - Surgical decompression, stabilization

DIAGNOSTIC CHECKLIST

Consider

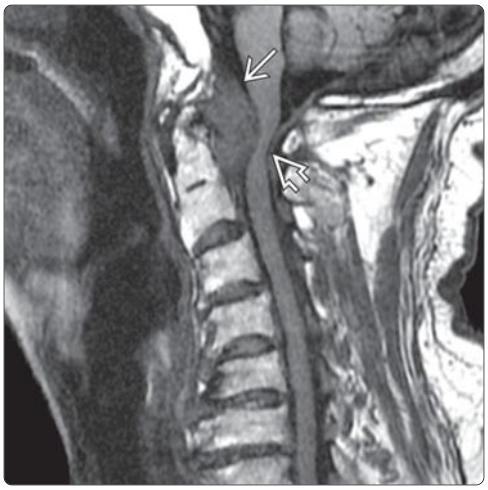
- CPPD may be present in axial skeleton without peripheral involvement
- Diagnosis may be missed at surgery unless surgeon is alerted to possibility of CPPD from imaging
  - Surgical specimens usually sent to pathology in formalin
  - Formalin dissolves crystals

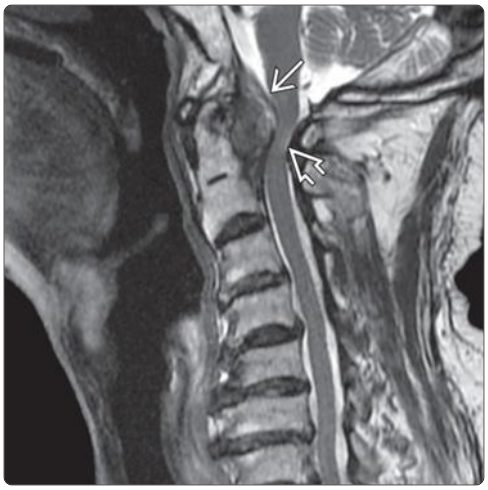
Image Interpretation Pearls

- CT useful in distinguishing from rheumatoid arthritis, which does not calcify

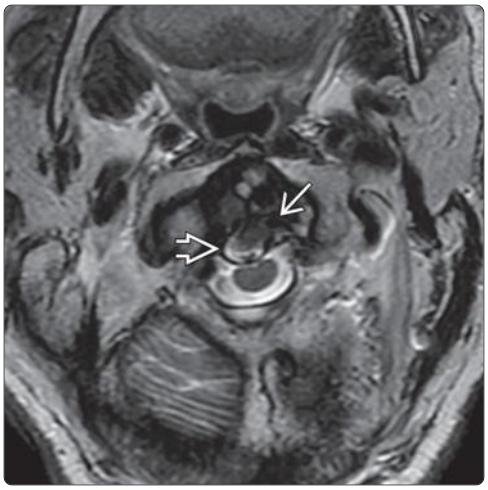
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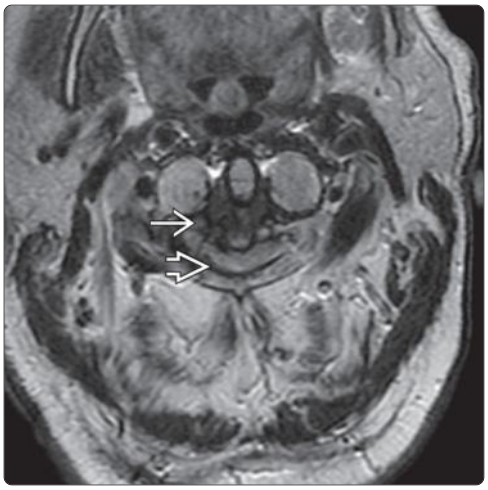
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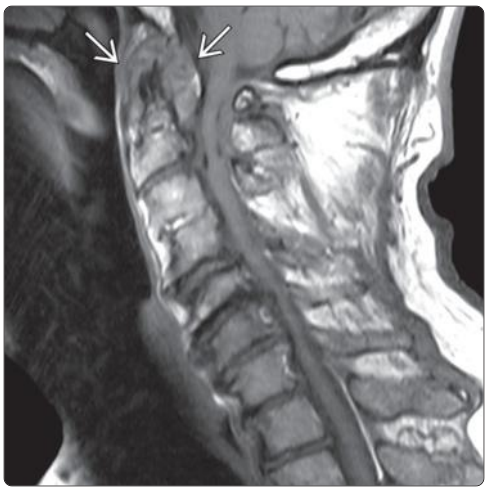


(Left) *Sagittal T1WI MR shows the typical pattern of CPPD involving C1-C2 articulation with a large mass ⇨ posterior to the odontoid process producing severe cord compression ⇨. The odontoid process appears otherwise intact, and without gross erosions.* **(Right)** *Sagittal T2 TSE image shows the large retroodontoid mass ⇨ to be relatively low in signal intensity. Note the marked effacement of the thecal sac and severe cord compression ⇨.*





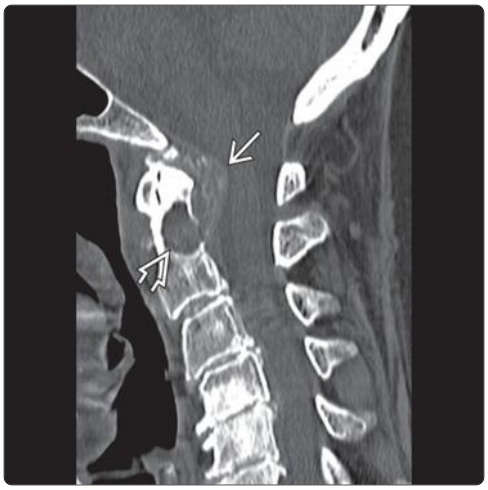
(Left) *Axial T2 TSE image shows a large mass with low T2 signal posterior to the odontoid process with severe cord compression ⇨. A small cystic focus is present within the dorsal aspect of the pseudotumorous mass due to an associated juxtaarticular cyst ⇨.* **(Right)** *Axial T2WI MR demonstrates a large low T2 signal mass engulfing the retroodontoid space ⇨ and causing severe cord compression ⇨. Note the intact cortex of the odontoid process.*





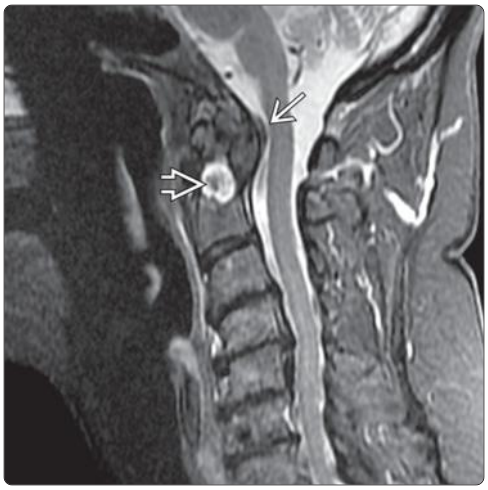
(Left) *Sagittal T1WI MR shows a mass surrounding the superior and posterior aspect of the odontoid process ⇨ producing cord compression. The anterior aspect of C1 is poorly identified and is also involved in the pseudomass. Multilevel degenerative disc disease is also present.* **(Right)** *Sagittal T2 TSE T2WI MR shows the periodontoid mass to be of low signal ⇨. Note the severe effacement of the thecal sac and cord compression. Cord integrity is compromised by multilevel subaxial spondylosis ⇨.*

(Left) *Sagittal bone NECT shows a large retroodontoid pseudomass effacing the ventral thecal sac ➡ that contains scattered calcifications. This pattern is pathognomonic of CPPD involvement of the craniovertebral junction. Note the large degenerative cyst involving the odontoid ➡.* **(Right)** *Axial bone NECT shows a large retroodontoid mass that contains scattered calcifications. The odontoid process is intact. The presence of calcium in the soft tissue distinguishes this CPPD from RA, which does not calcify.*





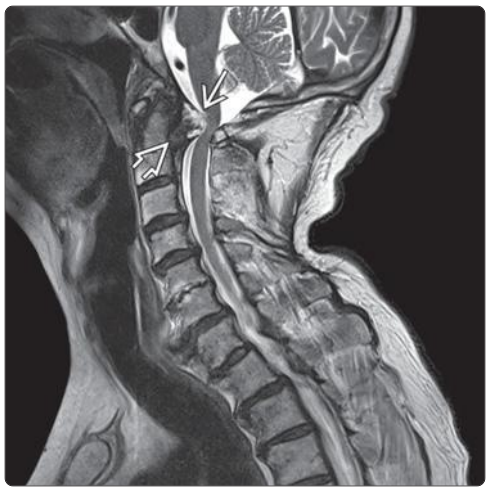
(Left) *Sagittal STIR MR shows a low signal intensity mass ➡ posterior to the odontoid process with moderate effacement of the ventral thecal sac. The high signal is due to associated degenerative cyst ➡ involving an odontoid process.* **(Right)** *Sagittal T1W C+ MR shows no enhancement ➡ of the low T2 signal retroodontoid tissue, which is typical for CPPD. The degenerative cyst within the odontoid process also does not enhance ➡.*

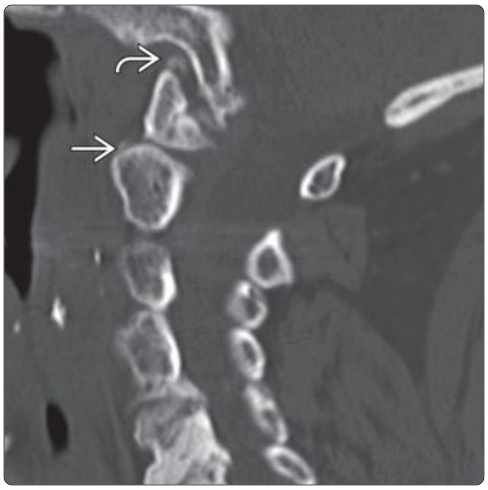


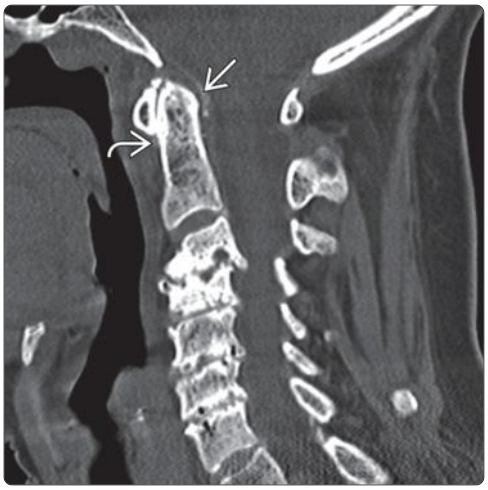


(Left) *Sagittal T1 C+ MR shows a juxtaarticular cyst ➡ associated with a CPPD-induced periodontal pseudotumor. There is peripheral enhancement of the cyst and no significant enhancement from the increased periodontal pseudotumor tissue ➡.* **(Right)** *Sagittal T2WI MR demonstrates an increased signal from the juxtaarticular cyst ➡ associated with the large low signal intensity pseudomass ➡ from CPPD deposition. There is severe cord compression.*

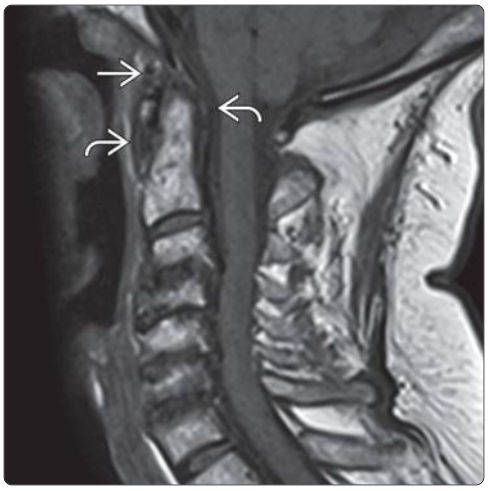








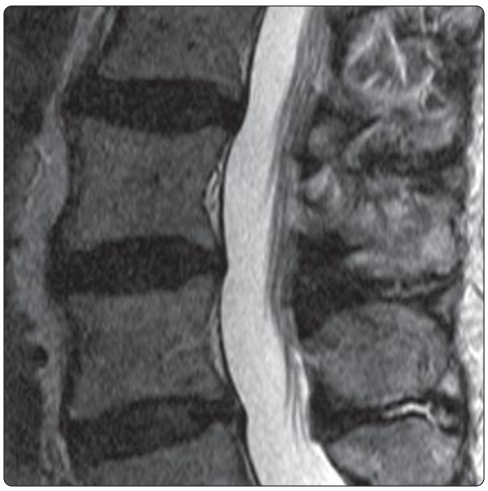
(Left) *Sagittal bone CT shows calcifications in the facet joints at C0-1 and C1-2 . CPPD involves synovial joints as well as discs and ligaments.* **(Right)** *Sagittal bone CT shows multiple sites of calcification in CPPD. This includes the tectorial membrane , the anterior longitudinal ligament , and multiple intervertebral discs. Endplate erosions are sharply demarcated and multiple levels are involved, helping to distinguish this case from infection.*





(Left) *Sagittal T1WI MR in the same patient shows soft tissue fullness around the dens. Punctate calcifications are difficult to see. Multilevel endplate erosions are evident, with adjacent decreased marrow signal intensity.* **(Right)** *Sagittal STIR MR in the same patient shows minimal abnormal signal intensity in the vertebral bodies despite extensive CT abnormalities. This is characteristic of CPPD. The discs remain low signal intensity, helping to distinguish CPPD from infection.*





(Left) *Lateral radiograph shows linear disc calcification , suggesting CPPD. In contrast to these linear calcifications, the calcified disc bulges or herniations due to disc degeneration are very common and cannot be used as a sign of CPPD.* **(Right)** *Sagittal STIR MR in the same patient is indistinguishable from degenerative disc disease. As a general rule, CT is preferable to MR in the differential diagnosis of all forms of spine arthritis.*

Gout

KEY FACTS

TERMINOLOGY

- Arthropathy secondary to urate crystal deposition

IMAGING

- Generally involves only 1-2 levels
- Disc or facet joints, sacroiliac joint
- Most common in lumbar spine
- Typically have peripheral joint disease
- Endplate, facet erosions
  - Usually sharply demarcated ("punched out")
- Preserved bone density
- Tophi
  - Mass in disc space, facet joint, paravertebral, spinal canal
  - May compress cord or nerve roots
  - May contain faint, amorphous calcifications on CT
  - Low signal intensity on T1WI
  - Low to intermediate signal intensity on T2WI, STIR
  - Variable enhancement with Gd
  - ± bone marrow edema adjacent to tophi

TOP DIFFERENTIAL DIAGNOSES

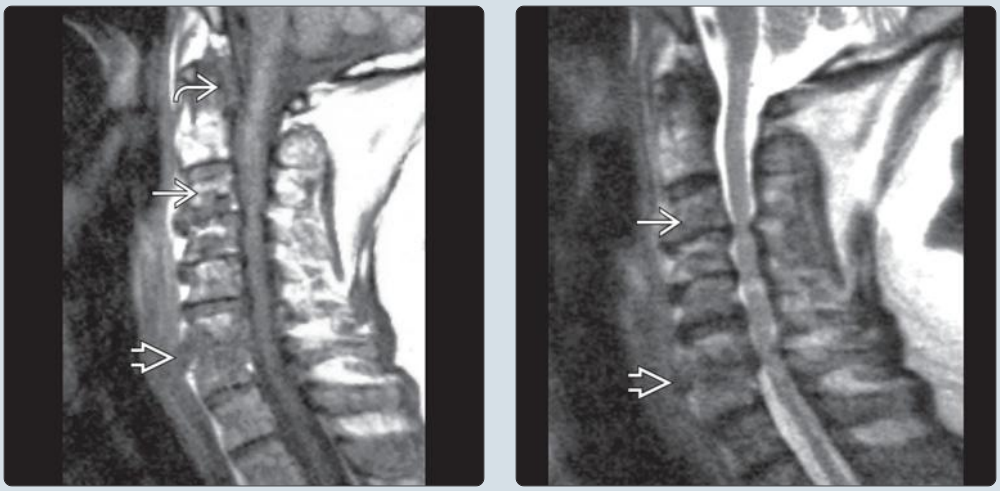
- Osteomyelitis
- Hemodialysis arthropathy
- Calcium pyrophosphate deposition
- Neuropathic arthropathy
- Seronegative spondyloarthropathy
- Rheumatoid arthritis

PATHOLOGY

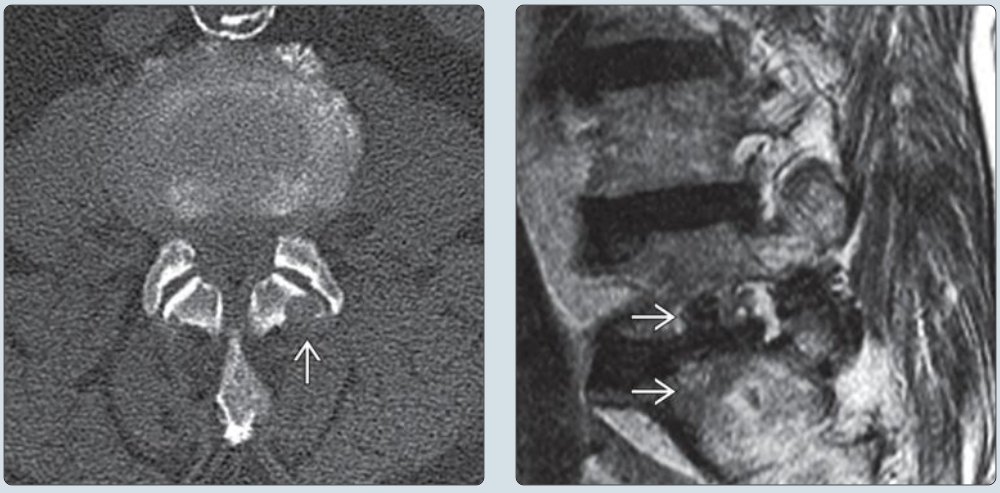
- Uric acid crystal deposition in soft tissues
- May be secondary to chronic diseases

CLINICAL ISSUES

- Treatment
  - Colchicine
  - Uric acid production inhibitors (e.g., allopurinol)
  - Uricosuric medications (e.g., probenecid)



(Left) *Sagittal PD/intermediate MR shows "punched out" vertebral body erosions at multiple levels. There is nearly complete endplate destruction at C6-C7. Tophus is present posterior to C2, and there are multiple intraosseous tophi.* **(Right)** *Sagittal T2 MR in the same patient shows endplate destruction at C6-C7. Intraosseous tophus is low signal intensity and almost indistinguishable from the surrounding marrow.*



(Left) *Axial NECT shows focal "punched out" lesion of soft tissue density in left facet, not directly contiguous with the joint space. Metastatic disease was primary consideration, but gout was diagnosed with biopsy. (Courtesy N. Farhataziz, MD.)* **(Right)** *Sagittal T2 MR shows focal erosions of L5 and S1 in a patient with uncontrolled peripheral gout and flare of back pain. There was minimal enhancement with gadolinium.*

TERMINOLOGY

Definitions

- Arthropathy secondary to urate crystal deposition

IMAGING

General Features

- Location
  - Most common in lumbar spine
  - Usually peripheral disease as well

Radiographic Findings

- Radiography
  - Endplate, facet erosions
  - Prevertebral soft tissue mass

CT Findings

- Endplate, facet erosions
  - Usually sharply demarcated ("punched out")
- Preserved bone density
- Soft tissue masses
  - May contain faint, amorphous calcifications

MR Findings

- Vertebral endplate destruction
  - Rounded masses may extend from disc into vertebrae
  - Discs low to intermediate signal intensity on T2WI, STIR
- Facet joint erosions and tophi
- Tophi
  - Mass in disc space, facet joint, paravertebral, spinal canal
  - May compress cord or nerve roots
  - Low signal intensity on T1WI
  - Low to intermediate signal intensity on T2WI, STIR
  - Variable enhancement with Gd
- ± bone marrow edema adjacent to tophi

Imaging Recommendations

- Best imaging tool
  - MR to show tophi, impingement on cord and nerve roots

DIFFERENTIAL DIAGNOSIS

Osteomyelitis

- Focal osteopenia
- Endplate erosions
- High signal intensity in discs on T2WI, STIR

Hemodialysis Arthropathy

- Patient history is key, otherwise same appearance

Calcium Pyrophosphate Deposition

- Calcifications tend to be more extensive, linear

Neuropathic Arthropathy

- Extensive bone destruction
- Bony debris, malalignment

Seronegative Spondyloarthropathy

- Vertebral body corner erosions
- Ligament ossification, vertebral fusion

Rheumatoid Arthritis

- Soft tissue calcifications absent

PATHOLOGY

General Features

- Etiology
  - Uric acid crystal deposition in soft tissues
  - May be secondary to chronic diseases
    - Renal disease, myeloproliferative disorders
- Specimens must be sent to pathology in alcohol; formalin dissolves crystals

Gross Pathologic & Surgical Features

- Chalky white tophi

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, often acute and severe
  - Other signs/symptoms
    - Fever, neurologic symptoms
- Clinical profile
  - Associated with obesity, rich diet
  - Serum uric acid may be normal at time of presentation

Demographics

- Gender
  - M:F = 20:1
    - Female patients almost always postmenopausal
- Epidemiology
  - Spine involvement rare

Natural History & Prognosis

- Most cases respond to medical therapy

Treatment

- Medical
  - Nonsteroidal anti-inflammatory medications
  - Colchicine
  - Uric acid production inhibitors (e.g., allopurinol)
  - Uricosuric medications (e.g., probenecid)
- Surgical decompression for neurologic symptoms

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Longus Colli Calcific Tendinitis

KEY FACTS

TERMINOLOGY

- Deposition of calcium hydroxyapatite crystals in superior oblique tendon fibers of longus colli muscle

IMAGING

- Focal calcification and soft tissue swelling in prevertebral space
- Almost always C1-C3
- CT
  - Retropharyngeal soft tissue swelling and calcification
  - Fluffy, amorphous calcification of longus colli
  - Elongated fluid collection extending along fascial planes
  - Decreased attenuation in longus colli muscle
- MR
  - Calcifications are globular and low signal intensity on all sequences
  - Prevertebral soft tissue swelling seen, ↓ signal intensity on T1WI, ↑ on T2WI and STIR
  - Nonenhancing fluid collection with adjacent diffuse soft tissue enhancement

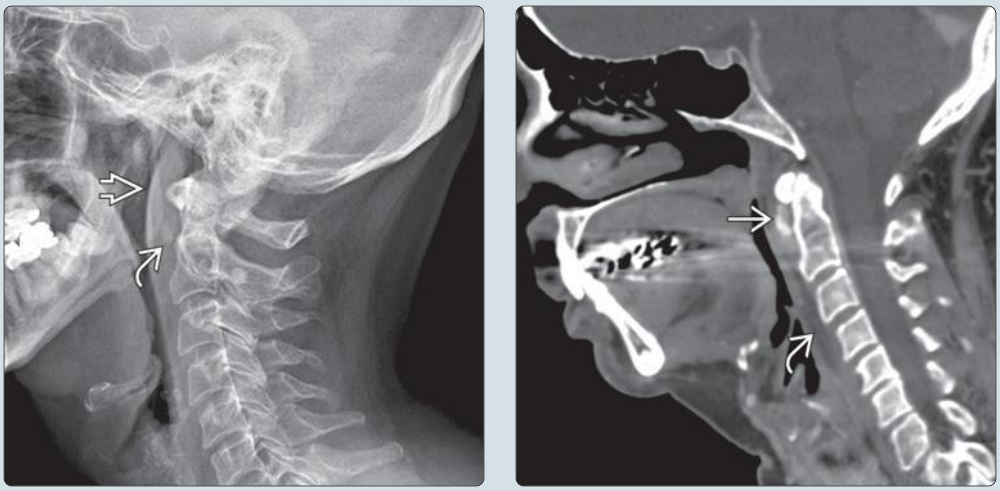
TOP DIFFERENTIAL DIAGNOSES

- Retropharyngeal abscess
- Cervical osteomyelitis
- Diffuse idiopathic skeletal hyperostosis (DISH)
- Calcium pyrophosphate dihydrate deposition (CPPD) disease
- Gout
- Tumoral calcinosis

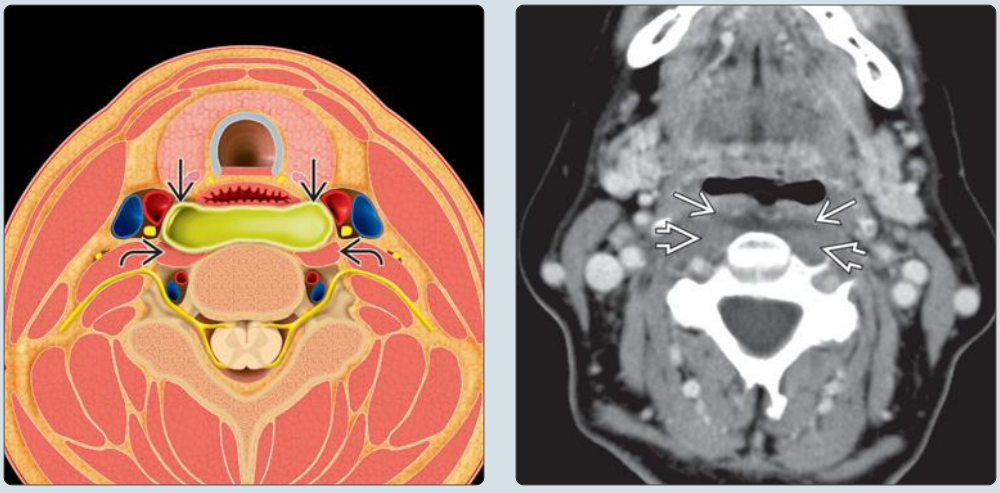
CLINICAL ISSUES

- Neck pain and stiffness, odynophagia
- Mild leukocytosis and fever
- Middle-aged and older patients
- Resolves in 1-2 weeks
- Nonsteroidal anti-inflammatory medications

(Left) *Lateral radiograph demonstrates a small density inferior to the C1 arch in the prevertebral space. There is marked prevertebral soft tissue swelling.* **(Right)** *Sagittal CT reconstruction better illustrates the ossification inferior to the C1 arch. There is fluid in the retropharyngeal space. It is important to differentiate longus colli tendinitis from infection, which needs antibiotic treatment.*



(Left) *Axial graphic expansion shows the retropharyngeal space with fluid. The longus colli muscle is a deep flexor of the neck, located anterior to the anterior longitudinal ligament. The longus colli is bounded by prevertebral fascia anteriorly and vertebral column posteriorly, and is composed of superior oblique, inferior oblique, and vertical portions.* **(Right)** *Axial CECT shows hypodense fluid in the retropharyngeal space, anterior to the paired longus colli muscles.*



TERMINOLOGY

Synonyms

- Calcific prevertebral/retropharyngeal tendinitis

Definitions

- Deposition of calcium hydroxyapatite crystals in superior oblique tendon fibers of longus colli muscle

IMAGING

General Features

- Best diagnostic clue
  - Focal calcification and soft tissue swelling in prevertebral space
- Location
  - Almost always C1-C3
    - Has been reported in lower cervical spine: C4-C5 and C5-C6 levels
  - Longus colli muscle is paired accessory neck flexor located in prevertebral space
    - Bounded by prevertebral fascia anteriorly and vertebral column posteriorly

Radiographic Findings

- Radiography
  - Retropharyngeal soft tissue swelling and calcification

CT Findings

- Fluffy, amorphous calcification of longus colli
- Decreased attenuation in longus colli muscle
- Elongated fluid collection extending along fascial planes
- No enhancement of fluid with contrast administration

MR Findings

- Calcifications are globular and low signal intensity on all sequences
- Prevertebral soft tissue swelling seen, ↓ signal intensity on T1WI, ↑ on T2WI and STIR
- Nonenhancing fluid collection with adjacent diffuse soft tissue enhancement

Imaging Recommendations

- Best imaging tool
  - CT is best to detect calcification

DIFFERENTIAL DIAGNOSIS

Retropharyngeal Abscess

- Rounded fluid collection on CT and MR
  - Compared to elongated fluid collection seen in longus colli tendinitis (LCT)
- Rim enhancement of fluid

Cervical Osteomyelitis

- Endplate erosions
- Peripherally enhancing collection in perivertebral space, epidural space
- Tuberculosis spondylitis can be associated with soft tissue calcification

Diffuse Idiopathic Skeletal Hyperostosis (DISH)

- Bone formation in anterior longitudinal ligament
  - Compared to amorphous calcification seen in LCT

Calcium Pyrophosphate Dihydrate Deposition (CPPD)

- Calcification around dens, in discs and ligaments
- May present similarly, with acute neck pain

Gout

- Peridiscal erosions
- Soft tissue mass, faint calcifications

Tumoral Calcinosis

- Calcific mass centered in large synovial joints
- Complication of renal dialysis, may be familial or result of degenerated or inflamed tissue

PATHOLOGY

General Features

- Etiology
  - Hydroxyapatite crystals deposition in longus colli muscle
    - May be due to local or systemic metabolic derangements, such as following ischemia, necrosis, or trauma

Gross Pathologic & Surgical Features

- Chalky, semiliquid calcific deposits

Microscopic Features

- Crystals amorphous on light microscopy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck pain and stiffness, limited range of motion
  - Odynophagia, dysphagia
  - Mild leukocytosis, ↑ inflammatory markers and fever

Demographics

- Age
  - 30-60 years
- Gender
  - M = F

Natural History & Prognosis

- Calcifications of LCT undergo spontaneous resolution
  - Margins become less well defined as calcifications are resorbed; complete resorption in ~ 2 weeks

Treatment

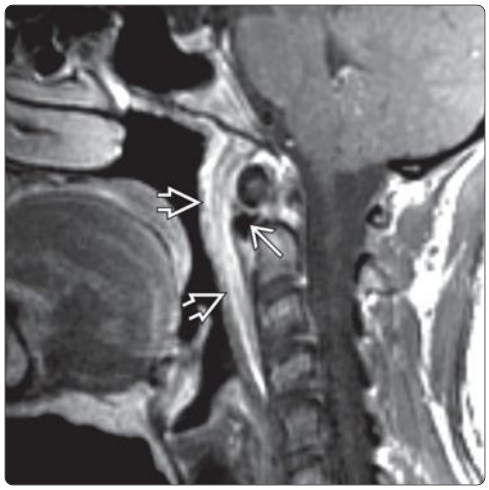
- Short course of nonsteroidal anti-inflammatory drugs and neck stabilization may help to alleviate symptoms

SELECTED REFERENCES

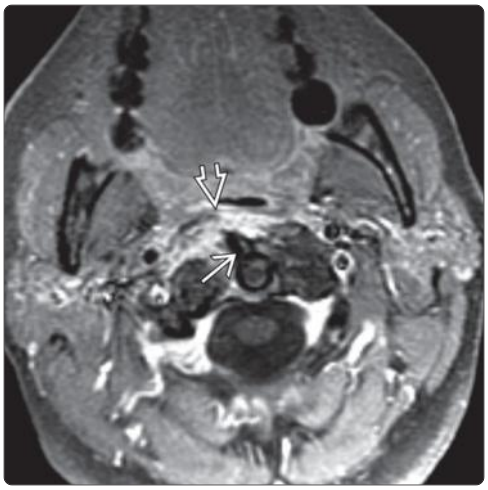
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(Left) *Axial CECT exhibits calcification of the longus colli tendon at the C1 level. There is edema in the adjacent soft tissues.* **(Right)** *Sagittal T1WI C+ MR displays enhancement of the thickened prevertebral soft tissue, indicating that underlying inflammatory soft tissue is present. The area of calcification is seen as low signal. The inflammatory changes seen on MR resolved completely in the course of 1 week without definitive therapy or intervention.*



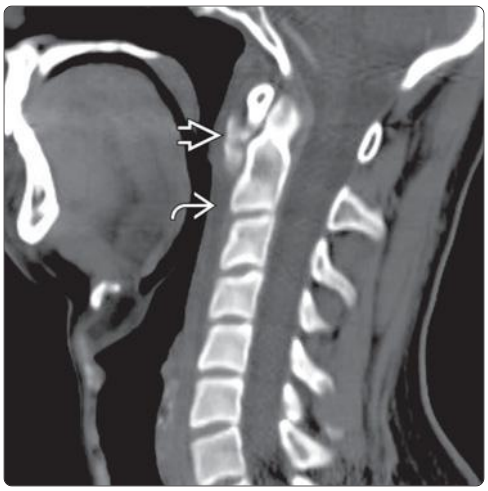


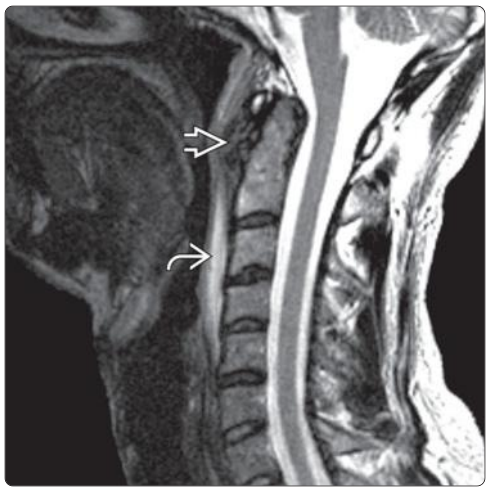
(Left) *Axial T1WI C+ FS MR shows enhancement of the prevertebral soft tissues, consistent with inflammation. The area of calcification is seen as low signal.* **(Right)** *Axial T1WI C+ FS MR demonstrates thickening and enhancement of the prevertebral muscles, particularly on the right. The classic clinical profile is a middle-aged patient with sudden onset of neck pain and stiffness. Acute LCT is considered relatively benign, typically resolving on its own within several weeks.*

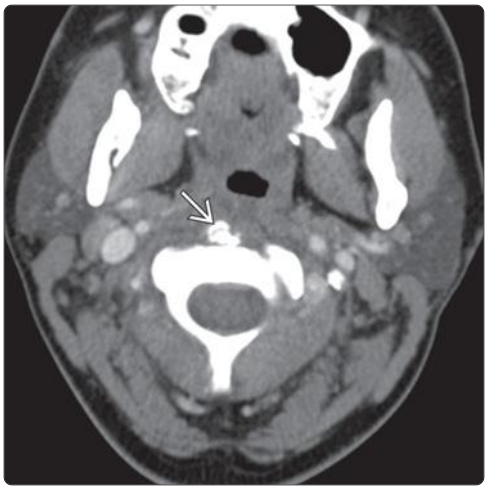




(Left) *Sagittal CT reconstruction illustrates smooth dense calcification in the superior insertion of the left longus colli muscle. Slight distension of the retropharyngeal space with fluid is also noted. The amorphous calcification lies anterior and inferior to the C1-C2 joint.* **(Right)** *Sagittal T2 MR in the same patient reveals the low signal of the calcific mass below the anterior atlantodental joint and marked hyperintensity of the swollen prevertebral tissues.*

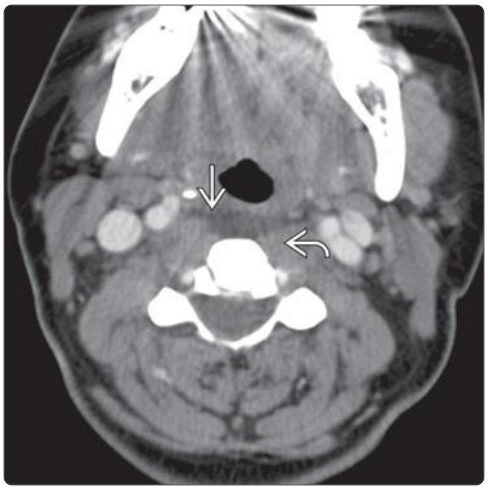


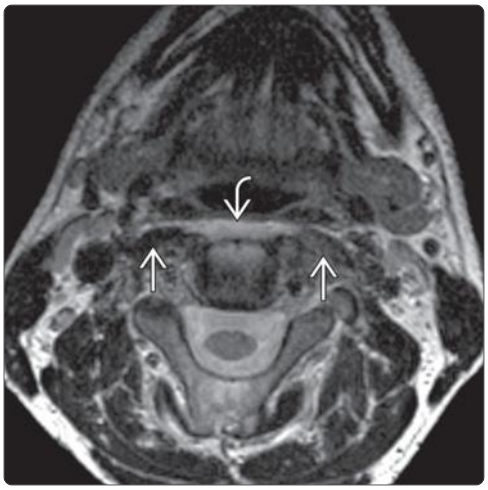




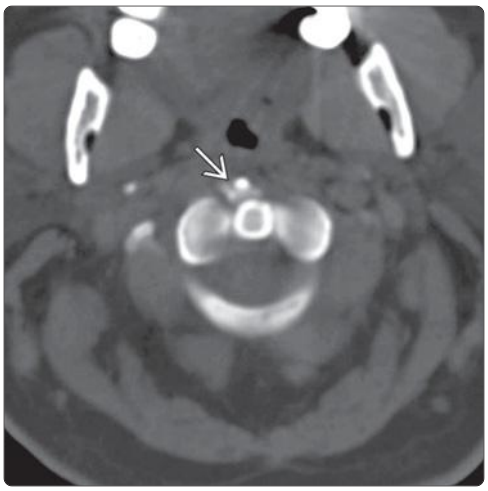


(Left) *Axial CECT illustrates calcification in the prevertebral tissues. LCT typically manifests on CT as paramidline calcium hydroxyapatite crystal deposits anterior to the C1 and C2 vertebral bodies. However, recent studies have brought attention to LCT existing at the C4-C5 and C5-C6 levels.* **(Right)** *Axial CECT displays thickening of the prevertebral muscles. The differential diagnosis for LCT includes retropharyngeal abscess, infectious spondylitis, trauma, or foreign body aspiration.*





(Left) *Axial CECT shows fluid in the retropharyngeal space, posteriorly displacing the longus colli muscles. The fluid collection in LCT is nonenhancing and elongated. This differs from a peripherally rounded fluid collection seen with retropharyngeal abscess.* **(Right)** *Axial T2 MR demonstrates retropharyngeal edema. The longus colli muscles are posteriorly displaced. Mild hyperintensity is observed in the left longus colli muscles more than in the right.*





(Left) *Axial bone CT exhibits calcification in the prevertebral soft tissues anterior to the C1-C2 joint. The patient presented with acute-onset neck pain with odynophagia, neck rigidity, and headache. A concurrent low-grade fever and elevated erythrocyte sedimentation rate and C-reactive protein level were present.* **(Right)** *The patient was treated with antibiotics for 8 days but without relief of symptoms. Axial NECT reveals increased calcification.*

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PART IV
SECTION 1

Infections



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Anatomy-Based Imaging Issues

Spread of infection may occur along one of many different tracts, including **direct extension**, **lymphatic spread**, **hematogenous spread**, and along the **cerebrospinal fluid pathways**. **Direct extension**, as its name implies, occurs when bone or soft tissue comes into contact with a directly adjacent infection leading to a soft tissue abscess or osteomyelitis. For the spine, this route is typically seen adjacent to a decubitus ulcer where there is adjacent osteomyelitis. An infection of the disc space can extend into the adjacent paravertebral soft tissues and produce psoas muscle abscesses. Direct extension is also the mechanism for epidural abscess involvement cranial or caudal to the site of disc space infection. This route can also be seen for an intramedullary spinal cord abscess where the infection occurs through congenital dysraphism or a dermal sinus tract. Lymphatic spread is of limited importance in the spine relative to the much more commonly seen direct extension and hematogenous spread. **Lymphatic spread** may be seen in cases of retroperitoneal node enlargement from pelvic or abdominal primary neoplasms.

Hematogenous Spread

Hematogenous spread is the major pathway of infection spread to the axial skeleton. Which route is more important (arterial or venous) is controversial. The arterial route is classically more important for spread of spinal infection. Vertebral bodies have areas that function physiologically in a similar manner to long bone metaphyses. The metaphyseal equivalent bone occurs near the anterior longitudinal ligament and has an end-arteriole network making it susceptible to bacterial seeding. These areas have distal nonanastomosing vessels that have slow flow, and occlusion of these vessels will lead to avascular necrosis. In the vertebral bodies, segmental arteries usually supply two adjacent vertebral bodies and the intervening disc, giving the typical disc space infection patterns. The venous route is classically through Batson plexus, which is a longitudinal network of valveless veins running parallel to the spinal column. These veins lie outside of the thoracoabdominal cavity. These veins communicate with multiple aspects of the venous system, including the vena cava, portal venous system, azygos system, intercostal veins, and pulmonary and renal veins. Flow direction within the plexus is variable due to the variable intrathoracic and intraabdominal pressures. The pharyngovertebral plexus serves the same physiologic purpose. The contiguity of the cerebrospinal fluid spaces between the intracranial vault and the thecal sac allows for direct communication of neoplasm and infection. Intracranial neoplasms may seed throughout the cervical, thoracic, or lumbar thecal sac. Likewise, even distal neoplasms involving the caudal thecal sac may propagate cephalad and extend into the intracranial cerebrospinal fluid space.

Pathologic Issues

Types of spinal infection can be divided into disc space infection/vertebral osteomyelitis, subdural empyema, meningitis, intramedullary cord abscess, and septic arthritis/facet joint involvement. Disc space infection shows the typical pattern of low signal intensity on T1-weighted images involving the disc space proper and extending to the adjacent endplates. Endplate irregularity is a typical feature. T2 hyperintensity is generally present within the intervertebral disc in a nonanatomic pattern, with adjacent T2 hyperintensity extending to the vertebral bodies. Contrast enhancement tends to be irregular when it involves the intervertebral disc, with diffuse enhancement extending to the involved vertebral bodies. Extension into the paravertebral soft tissues is an important aspect of disc space infections and should be evaluated via either fat-suppressed, post-contrast T1-weighted images looking for enhancement of the paravertebral and psoas musculature, or on T2-weighted images looking for T2 hyperintensity. It is important to comment not only on the level of involvement, but also on any instability or malalignment that may be present and whether there is extension into the paravertebral regions, epidural space, and psoas musculature.

Epidural Abscess and Meningitis

Isolated epidural abscesses can occur without concomitant disc space infections, but they can be associated with indwelling spinal catheters or prior spinal instrumentation. Uncommonly, these may occur as a result of hematogenous spread. Meningitis typically manifests on post-contrast T1-weighted images as linear enhancement along the pial surface of the cord or the roots of the cauda equina. With fungal infection, a more nodular enhancement pattern can be seen, which mimics the appearance of neoplastic spread. Spinal subdural empyemas are an uncommon manifestation of infection but may be seen in the setting of a severe disc space infection with adjacent extension into the epidural space. Presumably this is the result of direct extension through the dura and infection of the subdural space.

Intramedullary spinal cord abscesses are uncommon but can occur via both the hematogenous route and by direct extension. In adults, direct extension is the more typical mechanism. In children, the typical mechanism is direct extension through a dermal sinus. Septic arthritis/facet joint involvement may occur via hematogenous extension or by direct extension. Early infection may only be identified by slight T2 hyperintensity involving the bone of the facets, associated with facet effusion.

Adult vs. Pediatric

The routes of pyogenic infection will differ between adults and children due to developmental differences. In adults, the vertebral endplates become infected first, spreading to adjacent disc space and subsequently to the adjacent vertebral body, paravertebral tissues, and epidural space. In children, vascular channels are present across the growth plate, allowing primary infection of the intervertebral disc with subsequent secondary infection of the vertebral body. Disc space infections occur most commonly in the lumbar spine, followed by thoracic and cervical regions. Risk factors are many but include age over 50 years, diabetes, rheumatoid arthritis, AIDS, steroid administration, urinary tract instrumentation, prior spinal fracture, and paraplegia. *Staphylococcus aureus* is the most common organism. Pseudomonas may occur in the setting of drug abuse. *Salmonella* is the classic infection seen in sickle cell patients; however, *S. aureus* is still the most common overall in this population.

Classification

The Cierny and Mader classification of bone infection divides the pathology into four anatomic disease types and three host categories, yielding twelve clinical stages. The four anatomic disease types are: (1) Early hematogenous or medullary osteomyelitis, (2) superficial osteomyelitis (contiguous spread), (3) localized or full thickness sequestration, and (4) diffuse osteomyelitis. The three host classifications are: (A)

Normal physiologic response, (B) locally or systemically compromised response, and (C) treatment of the osteomyelitis would be worse than infection itself.

The spinal tuberculosis classification of Mehta (2001) divides the disease into four groups: (1) Stable anterior lesions without kyphotic deformity are treated with anterior debridement and strut grafting, (2) global lesions with kyphosis and instability are treated with posterior instrumentation and anterior strut grafting, (3) patients who are at high risk if treated by transthoracic surgery are treated with posterior decompression and instrumentation, and (4) isolated posterior lesions can be treated with posterior decompression.

Clinical Implications

Spinal involvement with infection represents 2-5% of all osteomyelitis sites. Axial spine pain is the most common presentation. This is progressive, although it can have a fairly insidious onset, producing pain without relief from rest. Fever is variable, and may be present in < 50% of cases. High-grade fever is present in < 5%, and motor and sensory deficits occur in 10-15% of patients. Rarely, intramedullary abscess can present with motor or sensory neurological deficits. Delay in diagnosing spinal infection is common. Intramedullary abscesses are fatal in 8%, with persistent neurological deficits in over 70%. Erythrocyte sedimentation rate is positive in more than 90%. C-reactive protein is also elevated. Blood cultures are positive with spinal osteomyelitis in 25-60% of cases.

Operative debridement with fusion may be necessary for a variety of reasons, including necessity to obtain a specific microorganism, abscess drainage, persistent neurological deficit, presence of spine instability and deformity, and failure of medical treatment. Long-term intravenous antibiotics remain the first line of therapy if there is no acute or evolving neurological deficit. A 6-week course of intravenous antibiotics is typical, which may also include an additional oral antibiotic regimen at the completion of the intravenous phase. External spine immobilization and bracing may be used. Recurrent bacteremia, paravertebral abscesses, and chronically draining sinuses are associated with relapse. Chronic auto-fusion of the infected level with successful nonoperative treatment is a common outcome.

Differential Diagnosis

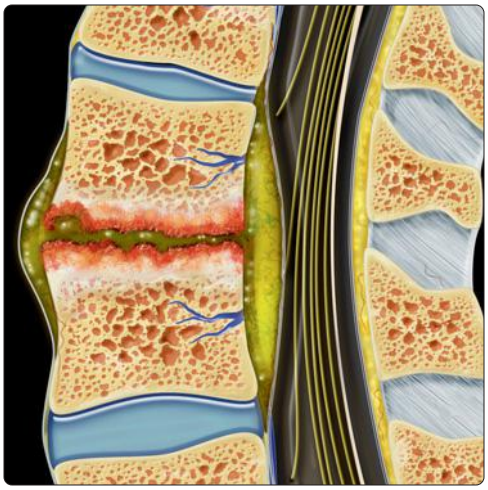
The primary diagnostic modality in the evaluation of epidural abscess is MR, which is as sensitive as CT myelography for epidural infection but also allows the exclusion of other diagnostic choices, such as herniation, syrinx, tumor, and cord infarction. MR imaging of epidural abscess demonstrates a soft tissue mass in the epidural space with tapered edges and an associated mass effect on the thecal sac and cord. The epidural masses are usually isointense to the cord on T1-weighted images and of increased signal on T2-weighted images. Contrast-enhanced MR is necessary for full elucidation of the abscess. The patterns of MR contrast enhancement of epidural abscess include: (1) Diffuse and homogeneous, (2) heterogeneous, and (3) thin peripheral. Enhancement is a very useful adjunct for identifying the extent of a lesion when the plain MR scan is equivocal, demonstrating activity of an infection, and directing needle biopsy and follow-up treatment. Successful therapy should cause a progressive decrease in enhancement of the paraspinal soft tissues, disc, and vertebral bodies. In the initial stages of vertebral osteomyelitis, when the disc space is not yet involved, it may be difficult to exclude neoplastic disease, type I degenerative endplate changes, or compression fracture from the differential diagnosis using only MR. Follow-up studies are usually necessary to further define the nature of the lesion.

Boden et al. suggested that in the postoperative spine, the triad of intervertebral disc space enhancement, annular enhancement, and vertebral body enhancement leads to the diagnosis of disc space infection, with the appropriate laboratory findings, such as an elevated sedimentation rate. However, there is a group of normal postoperative patients with annulus enhancement (at the surgical curette site), intervertebral disc enhancement, and vertebral endplate enhancement without evidence of disc space infection. In postoperative normal enhancement, the intervertebral disc enhancement is typically seen as thin bands paralleling the adjacent endplates, and the vertebral body enhancement is enhancement associated with type I degenerative endplate changes. This pattern should be distinguished from the amorphous enhancement seen within the intervertebral disc with disc space infection.

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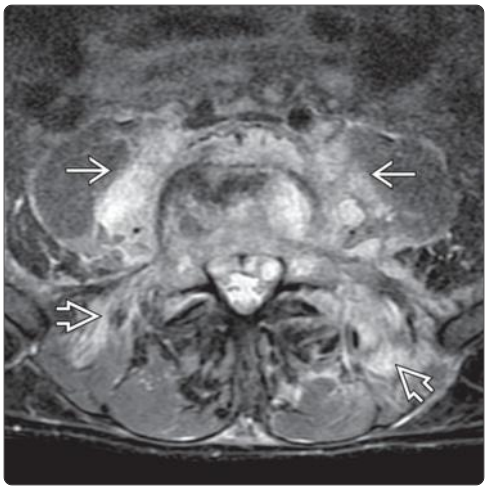
(Left) *Sagittal graphic shows lumbar disc space infection with vertebral body osteomyelitis with endplate destruction and marrow edema. There are ventral and dorsal abscess collections.* **(Right)** *Sagittal T1WI C+ FS MR in this case of disc space infection shows enhancement of L5 and S1 bodies and intervertebral disc, with prevertebral and epidural phlegmon extension.*



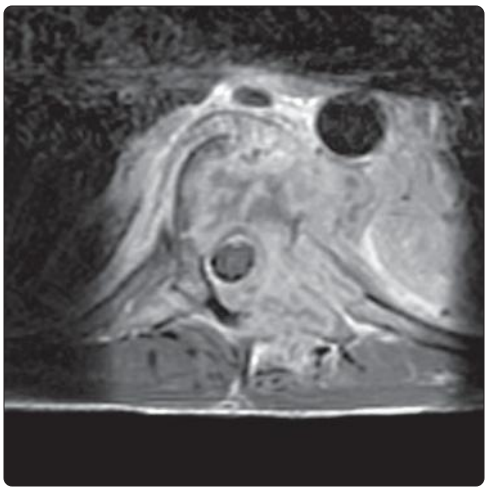


(Left) *Axial T1WI C+ MR of a disc space infection shows inflammatory extension into the prevertebral space, psoas muscles, and dorsal spinal muscles. Phlegmon extends into the ventral epidural space with thecal sac compression.* **(Right)** *Axial T2WI FS MR shows inflammatory extension into the prevertebral space, psoas muscles, and dorsal spinal muscles.*

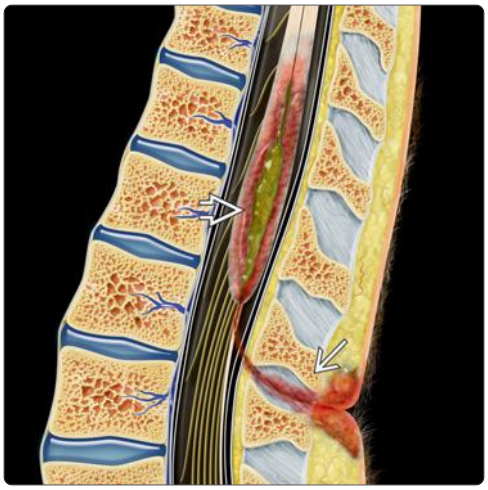


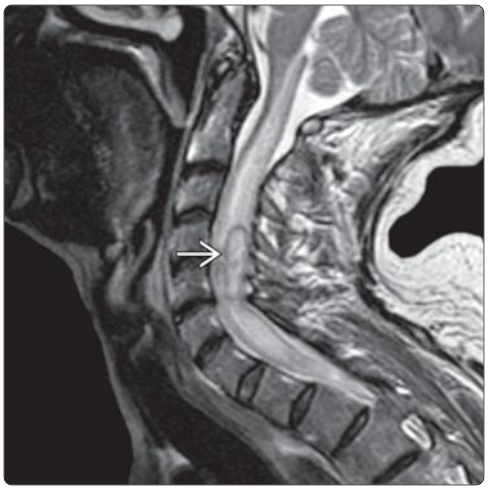


(Left) *Axial T1WI C+ MR shows disseminated coccidioidomycosis with diffuse bone and soft tissue involvement and adjacent paraspinal extension and extension into lung.* **(Right)** *Axial T2WI MR in coccidioidomycosis shows huge paraspinal abscesses. There is effacement of the normal thecal sac within the spinal canal due to disc space infection and osteomyelitis.*



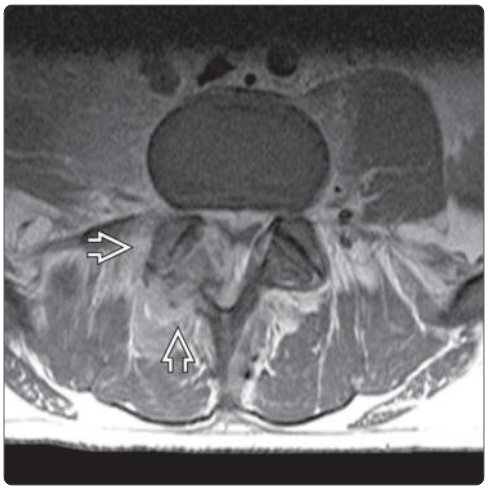




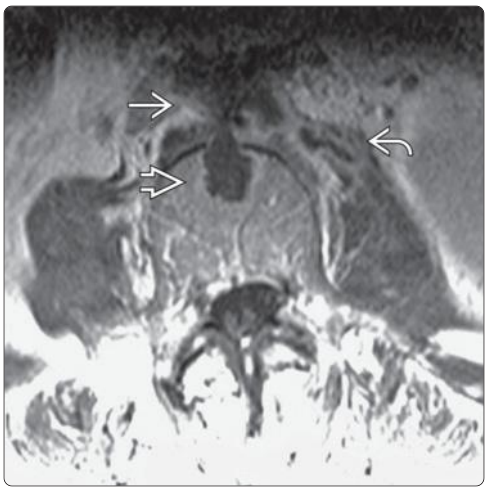


(Left) *Sagittal graphic shows dermal sinus ⇨ extending from skin surface to conus, with conus abscess ⇨ and extensive cord edema.* **(Right)** *Sagittal T2WI MR in a patient with a cervical cord abscess and streptococcal endocarditis shows diffuse cord expansion, with a ring-shaped area of low T2 signal (abscess capsule) within the cord from C4 to C5-C6 ⇨.*





(Left) *Sagittal T1WI C+ MR with fat suppression shows extensive subdural empyema with peripheral enhancement ⇨ throughout the cervical spine and extending along clivus ⇨.* **(Right)** *A septic facet joint is shown. Axial T1WI C+ MR at L4-L5 shows extension of the infection to the right facet joint with diffuse facet bone enhancement and juxta facet soft tissue involvement ⇨.*





(Left) *Axial T1WI C+ MR shows direct extension of infection from mycotic aortic aneurysm ⇨ into the ventral vertebral body, producing bone destruction and osteomyelitis ⇨. There is also direct extension of infection into psoas muscle ⇨.* **(Right)** *Axial CECT shows direct extension of infection from mycotic aortic aneurysm ⇨ into vertebral body ⇨ and left psoas muscle ⇨.*

Spinal Meningitis

KEY FACTS

IMAGING

- MR
  - Diffuse, extensive subarachnoid enhancement
  - Smooth or irregular meningeal enhancement

TOP DIFFERENTIAL DIAGNOSES

- Carcinomatous meningitis
  - Focal or diffuse, sheet-like or nodular enhancement along cord or nerve roots
- Sarcoidosis
  - Leptomeningeal + nerve root enhancement mimics spinal meningitis
- Lumbar arachnoiditis
  - "Empty sac" sign with nerve roots adherent to periphery of thecal sac
- Guillain-Barré syndrome
  - Inflammatory demyelination typically following recent viral illness

PATHOLOGY

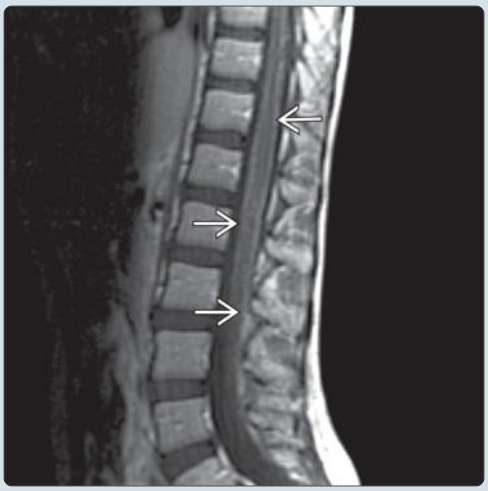
- Infection of CSF and meningeal coverings surrounding spinal cord
- Associated findings
  - Spondylodiscitis
  - Spinal epidural abscess
  - Blocked CSF flow → increased pressure within cord → syringomyelia

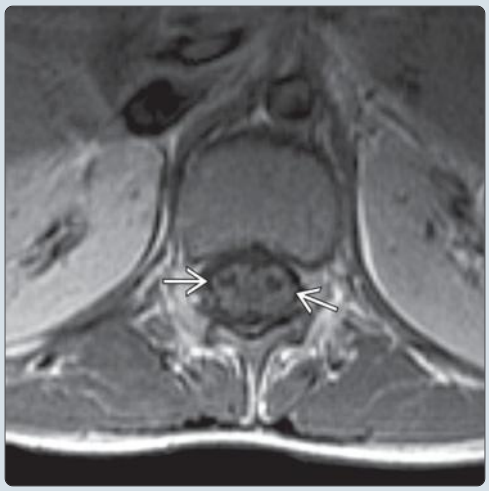
CLINICAL ISSUES

- Acute onset of fever, chills, headache, and altered level of consciousness

DIAGNOSTIC CHECKLIST

- Imaging often negative in early spinal meningitis
  - Positive in advanced bacterial meningitis or granulomatous infection
- Intravenous gadolinium increases sensitivity in detecting meningeal disease





(Left) *Sagittal T1WI C+ MR shows diffuse mildly irregular leptomeningeal enhancement. No extradural or vertebral inflammatory changes are apparent. Abnormal contrast agent enhancement is noted in only 55-70% of patients with proven infectious meningitis on contrast-enhanced MR images. Contrast-enhanced MR is particularly insensitive to viral meningitis.* **(Right)** *Axial T1WI C+ MR image reveals diffuse nerve root enhancement.*





(Left) *Sagittal T1 C+ MR exhibits diffuse leptomeningeal enhancement extending into the posterior fossa.* **(Right)** *Sagittal T2 MR reveals diffuse intramedullary hyperintensity consistent with spinal cord ischemia complicating meningitis. Inflammatory vasculitis acutely produces vascular compromise. Vascular insufficiency secondary to arachnoiditis gives delayed complications, such as paraparesis, sensory loss, and urinary incontinence.*

TERMINOLOGY

Synonyms

- Infectious arachnoiditis

Definitions

- Infection of spinal cord leptomeninges and subarachnoid space

IMAGING

General Features

- Best diagnostic clue
  - Diffuse, extensive subarachnoid enhancement
- Location
  - All spinal segments involved
- Size
  - Diffuse or focal
- Morphology
  - Smooth or irregular meningeal enhancement
  - Diffuse cerebral spinal fluid (CSF) enhancement

CT Findings

- CECT
  - Enhancing CSF ± meninges

MR Findings

- T1WI
  - Increased CSF intensity
  - Indistinct cord-CSF interface
  - Irregular cord outline
  - Clumped nerve roots
- T2WI
  - Obliterated subarachnoid space
  - Nodular or band-like filling defects in subarachnoid space
  - Complication of cord involvement: Hyperintense cord signal intensity (SI)
    - ± focal or diffuse cord swelling
- T1WI C+
  - Smooth or nodular leptomeningeal enhancement
    - May show homogeneously enhancing CSF
  - Smooth or nodular nerve root enhancement
    - ± segmental or focal intramedullary enhancement

Imaging Recommendations

- Best imaging tool
  - Axial and sagittal C+ T1WI
    - Positive in advanced bacterial meningitis or granulomatous infection
- Protocol advice
  - MR contrast mandatory for detecting meningeal disease

DIFFERENTIAL DIAGNOSIS

Carcinomatous Meningitis

- Primary CNS neoplasm
- Metastatic lung, breast carcinoma, melanoma, lymphoma
- Thickened, blurred nerve roots on T1WI and T2WI
  - Obliterated CSF
- Focal or diffuse, sheet-like or nodular enhancement along cord or nerve roots

Sarcoidosis

- Noncaseating granulomatous inflammation of spinal cord and its coverings
- Protean imaging findings
  - Leptomeningeal + nerve root enhancement mimics spinal meningitis
  - Cord edema with focal intramedullary enhancement simulates myelitis
- Concurrent systemic manifestations and elevated angiotensin-converting enzyme level help make diagnosis
  - Clinical CNS involvement in 5% of patients with sarcoidosis

Lumbar Arachnoiditis

- Commonly associated with prior surgery
- Cauda equina typically involved
- Clumped nerve roots forming central mass or multiple cords
  - ± mild nerve root enhancement
- "Empty sac" sign with nerve roots adherent to periphery of thecal sac

Guillain-Barré Syndrome

- Inflammatory autoimmune demyelination typically following recent viral illness
- Ascending paralysis
- Diffuse enhancement of conus and cauda equina
  - ± nerve root thickening (uncommon)

Intracranial Hypotension

- From prior spinal trauma, diagnostic or interventional procedure, or spontaneous
  - Low opening pressure on lumbar puncture
  - Increased dural venous engorgement
  - Diffuse smooth meningeal thickening & enhancement
- May see cerebellar tonsillar descent + effaced prepontine space in posterior fossa

PATHOLOGY

General Features

- Etiology
  - Acute meningitis: Onset of symptoms < 24 hr
    - Almost always bacterial
    - Newborn: Group B *Streptococcus*, gram-negative bacilli, *Listeria* monocytogenes
    - 2 months to 12 years: *Haemophilus influenzae, Streptococcus pneumoniae,* and *Neisseria meningitides*
    - Adults: Above, plus streptococci + staphylococci
  - Subacute meningitis: Symptoms develop in 1-7 days
    - Mostly viral (e.g., HIV-related CMV radiculomyelitis), some bacterial (e.g., Lyme disease)
  - Chronic meningitis: Fluctuating symptoms for > 7 days
    - Tuberculosis
    - Syphilis
    - Fungal: Coccidioidomycosis, cryptococcosis, and aspergillosis
  - Mechanism of inoculation
    - Hematogenous dissemination from extraspinal focus of infection

  - Contiguous spread from adjacent spondylodiscitis, spinal epidural abscess
  - Direct inoculation through trauma or interventional procedures
  - Unexplained source of infection: Probably bacteria (meningococci) colonized in nasopharynx
- Pathophysiology of bacterial meningitis
  - Initial acute inflammatory exudate in subarachnoid space
  - Toxic mediators potentiate inflammatory response
  - Increased permeability of blood-cord barrier
  - Influx of inflammatory cells
  - Spinal cord swelling and edema likely due to ischemia from vasculitis, venous congestion, &/or direct infection
- Associated abnormalities
  - Spondylodiscitis
  - Spinal epidural abscess
  - Subdural empyema
  - Subarachnoid cysts
    - Fibrin deposition results in loculation of subarachnoid space
  - Myelitis
  - Cord abscess
  - Syringomyelia
    - Blocked CSF flow results in increased pressure within cord and subsequent central canal expansion

Microscopic Features

- Cellular debris, inflammatory cells, and microorganisms
- Tuberculous meningitis
  - Small tubercles consist of epithelioid cells, Langhans giant cells, and foci of caseation
- Loss of integrity of brain-CSF barriers, oxidative stress and S-100B (mediator of astrocytes activation/injury)
  - → contribute to severity and neurological complications of bacterial meningitis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute onset of fever, chills, headache, and altered level of consciousness
  - Other signs/symptoms
    - Generalized convulsions
    - Neck stiffness
    - Paraparesis
    - Paresthesia
    - Gait disturbance
    - Urinary bladder dysfunction
- Clinical profile
  - Milder symptoms with protracted course in tuberculous or fungal meningitis

Demographics

- Age
  - Newborns
  - Infants: Peak age 3-8 months
  - Adults: 20s; 60s
- Gender
  - No gender preference among adults
  - M:F = 3:1 in neonates
- Epidemiology
  - Incidence of bacterial meningitis: 2-3 per 100,000

Natural History & Prognosis

- Prognosis depends on severity of disease, causative pathogen, patient age and comorbidities
  - Mortality of bacterial meningitis (20-90%)
    - Depending on initial neurologic impairment and rate of progression
    - Chronic disabilities include paralysis, seizures, deafness, etc.
  - Viral meningitis generally less severe
    - Full recovery within 2 weeks in most cases

Treatment

- Bacterial meningitis presenting with normal CSF is uncommon
  - Incidence (0.5-12%)
  - "Normal" CSF in meningitis does not correlate with age of child or subsequently demonstrated organism
    - Related to duration of illness; incidence is higher when lumbar puncture (LP) is performed within 1st 24 hours of illness (before inflammatory response has developed)
      - Repeat LP after 24-48 hours in clinically suspected cases
      - Repeat LP in blood culture-positive cases if initial CSF is clear
- CSF lactate level → distinguish bacterial infection where lactate is ↑ from nonbacterial (viral) meningitis
  - High sensitivity, specificity, and predictive values
  - Level on 1st LP has no prognostic value, but decrease of CSF lactate during treatment → good prognosis
- Supportive care with hydration and pain management
- Intravenous dexamethasone to decrease inflammatory reaction and brain/cord edema
- Empiric intravenous antibiotics based on suspected organisms in each age group
- Organism-specific intravenous antibiotics
- Preventive oral antibiotics for close contacts of patients with *Neisseria meningitides*

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Imaging often negative in early spinal meningitis
- Increased CSF SI on T1WI with diffuse post-gadolinium enhancement suggestive of spinal meningitis

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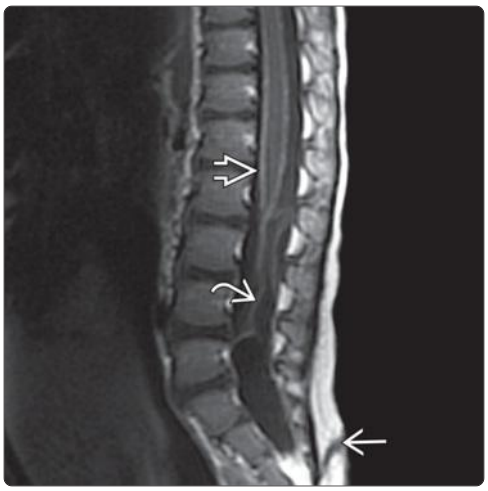


(Left) *Sagittal T1WI C+ MR shows diffuse abnormal enhancement of the cauda equina and distal cord surface with central clumping of the nerves within the thecal sac. The leptomeningeal enhancement is due to histoplasmosis (successfully treated with antifungal medication).* **(Right)** *Sagittal T1WI C+ MR shows extensive thickening of the dura with enhancement and diffuse epidural phlegmon. There is massive infection involving soft tissues, epidural space, and subarachnoid space.*





(Left) *Sagittal T1 C+ MR reveals extensive leptomeningeal enhancement surrounding the spinal cord. There is enlargement of the 4th ventricle due to severe coccidioidomycosis meningitis.* **(Right)** *Diffuse T2 hyperintensity due to severe coccidioidomycosis meningitis extends inferiorly from the obex to the T4 level. There is relative sparing of the cord periphery. Extensive cervical cord edema (presyrinx) is related to marked hydrocephalus and 4th ventricular outflow obstruction.*





(Left) *Diffuse pial and cauda equina enhancement is due to meningitis. Minimal enhancement outlines an epidermoid. There is a dorsal dermal sinus tract in the low sacral region.* **(Right)** *Thickening and enhancement of the cauda equina is due to adhesive arachnoiditis. A fibrinous exudate with minimal cellular infiltrate adheres to the arachnoid membranes and nerve roots. Fibroblasts infiltrate the exudate and produce collagen bands. Meningeal scarring can alter CSF flow dynamics.*

Pyogenic Osteomyelitis

KEY FACTS

TERMINOLOGY

- Bacterial suppurative infection of vertebrae and intervertebral disc

IMAGING

- Ill-defined hypointense T1 vertebral marrow with loss of endplate definition on both sides of disc
- Loss of disc height and abnormal disc signal
- Destruction of vertebral endplate cortex
- Vertebral collapse
- Paraspinal ± epidural infiltrative soft tissue ± loculated fluid collection
- Follow-up MR
  - Should focus on soft tissue findings
  - No single MR imaging parameter is associated with clinical status

TOP DIFFERENTIAL DIAGNOSES

- Degenerative endplate changes
- Tuberculous vertebral osteomyelitis
- Spinal neuropathic arthropathy

PATHOLOGY

- Predisposing factors
  - Intravenous drug use
  - Immunocompromised state
  - Chronic medical illnesses (renal failure, cirrhosis, cancer, diabetes)
- *Staphylococcus aureus* is most common pathogen

CLINICAL ISSUES

- Acute or chronic back pain
- Focal spinal tenderness
- Fever
- ↑ ESR, ↑ CRP, ↑ WBC





(Left) *Sagittal T1WI MR in a patient with a history of lumbar surgery shows findings of disc space infection at L4-L5, with hypointense marrow, vertebral collapse, endplate erosion, disc space loss, and epidural phlegmon.* **(Right)** *Sagittal T1WI C+ MR demonstrates enhancing vertebral bodies and intervening disc. There is an epidural abscess extending from L4-L5 to S1, consistent with pyogenic vertebral osteomyelitis. Severe central canal narrowing is present at L4-L5.*





(Left) *Sagittal STIR MR shows increased fluid in the retropharyngeal/prevertebral space. Marrow edema in C6 and C7 vertebral bodies is seen. There is fluid signal within the disc space with irregularity along the endplate cortical margins.* **(Right)** *Sagittal T1WI C+ FS MR shows homogeneously enhancing epidural phlegmon at C6-C7 level, causing mass effect on the cord. C6 & C7 vertebral bodies exhibit avid homogeneous enhancement. Note the prevertebral, enhancing soft tissues, representing phlegmon.*

TERMINOLOGY

Synonyms

- Pyogenic spondylodiscitis, disc space infection

Definitions

- Bacterial suppurative infection of vertebrae and intervertebral disc

IMAGING

General Features

- Best diagnostic clue
  - Ill-defined hypointense vertebral marrow on T1WI with loss of endplate definition on both sides of disc
- Location
  - All spinal segments involved
    - Lumbar (48%) > thoracic (35%) > cervical spine (6.5%)
- Morphology
  - Loss of disc height
  - Abnormal disc signal
  - Destruction of vertebral endplate cortex
  - Ill-defined marrow signal alteration
  - Vertebral collapse
  - Paraspinal ± epidural infiltrative soft tissue ± loculated fluid collection
    - Present in 75% of pyogenic vertebral osteomyelitis

Radiographic Findings

- Radiography
  - Negative up to 2-8 weeks after onset of symptoms
  - Initial endplate and vertebral osteolysis followed by increased bone density
  - Paraspinal soft tissue density and loss of fat planes
  - Fusion across disc space late in disease course

CT Findings

- NECT
  - Endplate osteolytic/osteosclerotic changes
  - Spinal deformity best seen on coronal and sagittal reformation
  - Increase in paraspinal soft tissue
- CECT
  - Enhancing disc, marrow, and paravertebral soft tissue

MR Findings

- Disc space
  - Hypointense on T1WI
  - Variable, typically hyperintense on T2WI
  - Diffuse or rim enhancement with gadolinium
  - Loss of height
- Vertebral marrow signal abnormality abutting disc
  - Hypointense on T1WI
  - Hyperintense on fat-saturated T2WI or STIR
  - Avid enhancement with gadolinium
- Paraspinal and epidural phlegmon or abscess
  - Isointense to muscle on T1WI
  - Hyperintense on T2WI
  - Diffuse or rim enhancement
- Cord compression
- DWI
  - Acute: Vertebral bodies, endplates, and discs hyperintense
  - Chronic: Hypointense
- Follow-up MR
  - No single MR imaging finding was associated with clinical status
  - Less paraspinal inflammation and less epidural enhancement compared with baseline
  - Vertebral body, disc space enhancement, and bone marrow edema equivocal or worse compared with baseline

Nuclear Medicine Findings

- Bone scan
  - 3-phase technetium-99m diphosphonate scan shows increased activity in all phases
- Gallium scan
  - Increased uptake of gallium citrate (Ga-67)
    - Increased sensitivity with SPECT
- WBC scan
  - Often false-negative in patients with chronic vertebral osteomyelitis

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI and T1WI MR
    - Sensitivity (96%), specificity (92%), accuracy (94%)
  - SPECT Ga-67 scan good alternative
    - Sensitivity and specificity in low 90%
- Protocol advice
  - STIR or FSE T2 with fat suppression most sensitive for marrow edema, epidural involvement
  - Post-gadolinium T1WI with fat suppression also improves MR sensitivity
    - Improves evaluation of epidural and soft tissues

DIFFERENTIAL DIAGNOSIS

Degenerative Endplate Changes

- Most common mimic
- Disc desiccation
  - Hypointense on T1WI and T2WI
  - Minimal or absent enhancement with gadolinium
- Vertebral endplates preserved

Tuberculous Vertebral Osteomyelitis

- Midthoracic or thoracolumbar > lumbar or cervical
- Vertebral collapse, gibbus deformity
- ± endplate destructive changes
- Large dissecting paraspinal abscesses out of proportion to vertebral involvement

Spinal Neuropathic Arthropathy

- Sequela of spinal cord injury
- Disc space loss/T2 hyperintensity, endplate erosion/sclerosis, osteophytosis, soft tissue mass
  - Present in both spondylodiscitis and neuropathic spine
- Vacuum disc/rim enhancement, facet involvement, spondylolisthesis, debris, disorganization

Chronic Hemodialysis Spondyloarthropathy

- Cervical spine most common

- Disc space loss, endplate erosion, vertebral destruction
- Vertebral marrow hypointense on both T1WI and T2WI
- Low to intermediate disc signal intensity on T2WI

Spinal Metastases

- Discrete or ill-defined vertebral lesions
  - Hypointense on T1WI
  - Hyperintense on T2WI
  - Post-gadolinium enhancement
- Posterior elements commonly affected
- Disc space preserved

PATHOLOGY

General Features

- Etiology
  - Predisposing factors
    - Intravenous drug use
    - Immunocompromised state
    - Chronic medical illnesses (renal failure, cirrhosis, cancer, diabetes)
  - *Staphylococcus aureus* is most common pathogen
    - *Escherichia coli* most common within gram-negative bacilli
    - *Salmonella* more common in patients with sickle cell disease
  - Bacteremia from extraspinal primary source
    - Most common route of infection
    - GU or GI tract, pulmonary, cardiac, mucous/cutaneous sources
    - Vascularized subchondral bone adjacent to endplate seeded primarily
    - Secondary infection of intervertebral disc, adjacent vertebra
    - Intervertebral disc 1st site of infection in children due to presence of vascularity
  - Direct inoculation from penetrating trauma, surgical intervention, or diagnostic procedures
  - Extension from adjacent infection
    - Diverticulitis, appendicitis, inflammatory bowl disease
    - Pyelonephritis
- Associated abnormalities
  - Spinal meningitis
  - Vascular compromise of cord and myelitis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute or chronic back pain
  - Focal spinal tenderness
  - Fever
- Other signs/symptoms
  - Myelopathy if cord compromised
  - Elevated erythrocyte sedimentation rate, C-reactive protein, white cell count
- Clinical profile
  - Average duration of symptoms for 7 weeks before diagnosis

Demographics

- Age
  - Bimodal distribution
    - Pediatric patients
    - 6th-7th decade
- Epidemiology
  - 2-7% of all osteomyelitis occurs in spine

Natural History & Prognosis

- Mortality rate: 2-12%
- Favorable outcome; resolution of symptoms if prompt diagnosis & treatment
  - Residual functional deficits may be present in 15% of patients
- Recurrence due to incomplete treatment (2-8%)
- Irreversible neurological deficits
  - Delay in diagnosis & neurologic impairment at diagnosis significant predictors of neurologic deficit at follow-up
  - Previous spinal surgery associated with adverse outcome: Readmission within 1 year of discharge following 1st spinal infection
  - Independent predictors of long-term disability
    - Neurological impairment at time of diagnosis, time to diagnosis ≥ 8 weeks, & debilitating disease
- Improvement in imaging findings may lag behind clinical improvement

Treatment

- CT-guided or open biopsy yields causative organism more often than blood cultures (77% vs. 58%)
  - Previous antibiotic treatment significantly lowers yield (23% vs. 60%)
- Early empiric antibiotics, broad spectrum coverage until causative pathogen isolated
- Organism-specific parenteral antibiotics for 6-8 weeks
- Spinal immobilization with bracing for 6-12 weeks
- Surgical treatment
  - Laminectomy, debridement, ± stabilization
  - Especially if epidural abscess, instability present

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Diffusely enhancing disc, adjacent vertebral marrow, soft tissue with endplate erosion highly suggestive of vertebral osteomyelitis

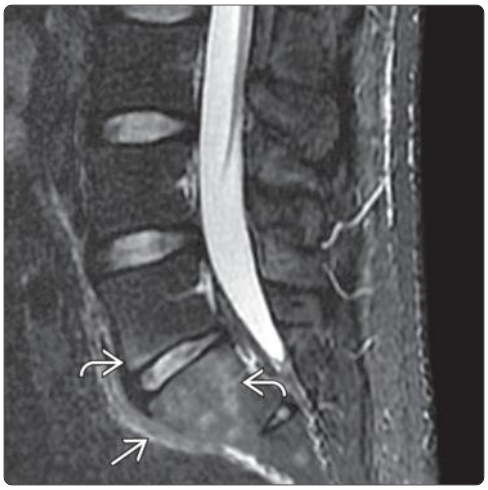
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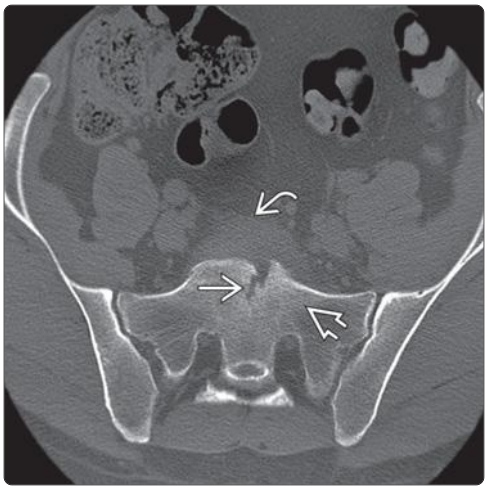
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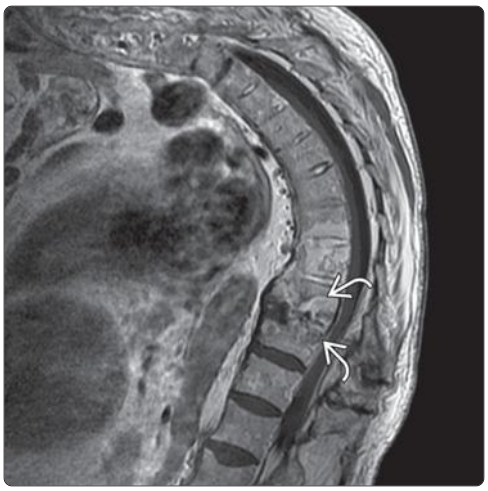
(Left) *Sagittal STIR MR illustrates marrow edema related to discitis-osteomyelitis at the T12-L1 level. Intervertebral fluid extends into the paraspinal soft tissues. Cortical irregularity along the adjacent endplate margins is observed.* **(Right)** *Sagittal T1WI C+ MR shows peripheral enhancement of the T12/L1 disc and paraspinal enhancing phlegmon. Follow-up MRs often depict less paraspinal inflammation and less epidural enhancement compared with baseline.*





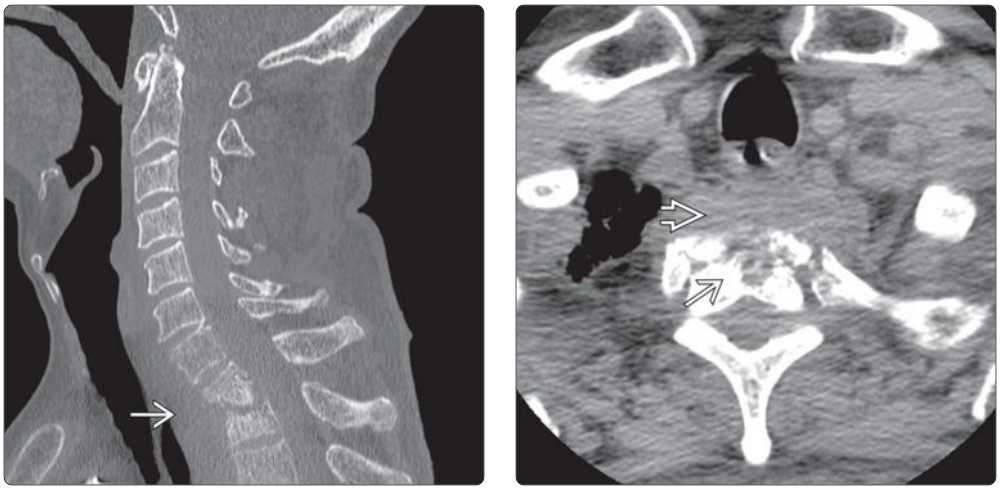
(Left) *Sagittal STIR MR depicts marrow edema in L5 and S1 vertebral bodies. Presacral fluid-intensity collection is observed. The anterior cortical margin is obscured. On follow-up MR, vertebral body, disc space enhancement, and bone marrow edema may be equivocal or appear worse compared with baseline.* **(Right)** *Axial CT reconstruction exhibits an irregular cortical break along the anterior superior margin of the S1 body, subtle sclerosis, and a presacral soft tissue component.*



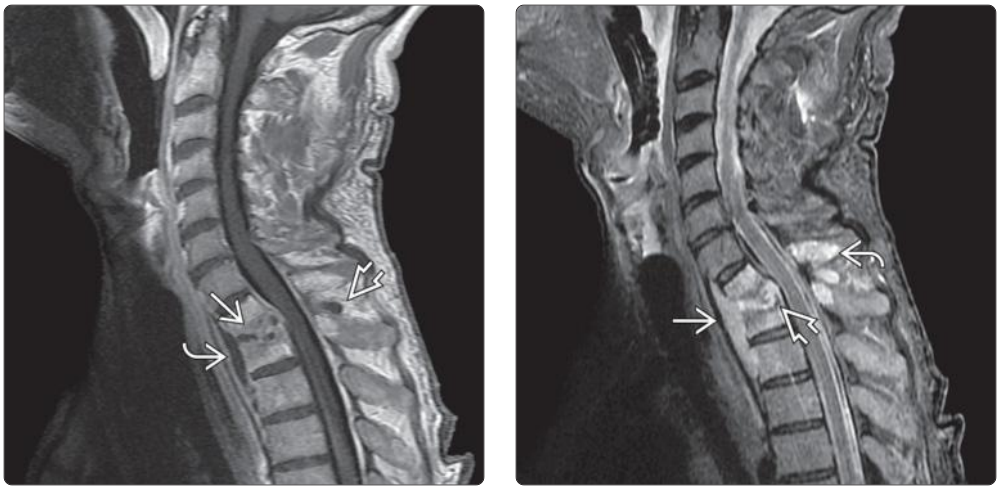


(Left) *Abnormal T1 hypointensity is seen in the marrow of 2 adjacent midthoracic vertebral bodies. Thin syndesmophytes are compatible with ankylosing spondylitis (AS).* **(Right)** *T1 C+ MR shows enhancement of adjacent irregular endplates. Aseptic spondylodiscitis can complicate AS. Proliferative epidural tissue without inflammatory infiltrates and new bone reaction, suggesting the contribution of mechanical factors, may cause neurological complications.*

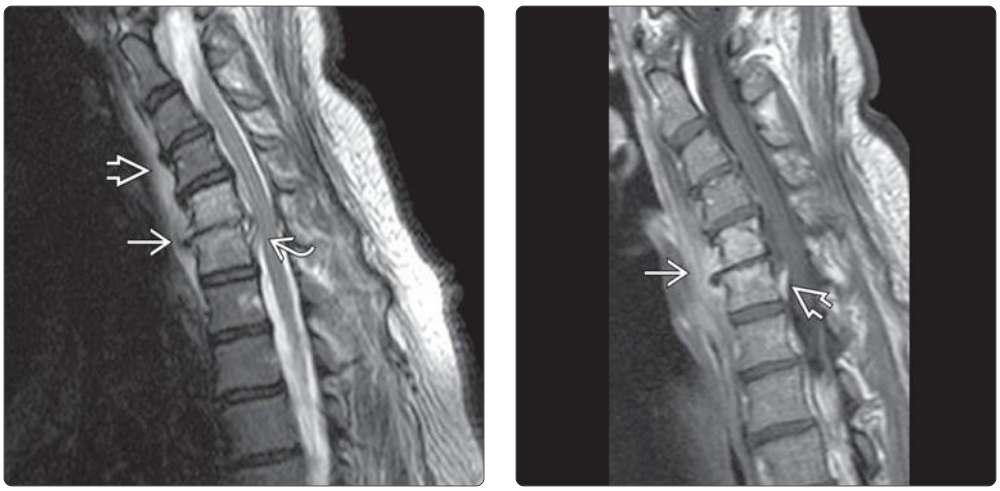
(Left) *Sagittal NECT(bone window) of upper thoracic disc space infection shows destruction of the T1 and T2 bodies centered about the collapsed disc space with marked endplate irregularity and kyphotic deformity.* **(Right)** *Axial NECT shows destruction of the T1 and T2 bodies with marked endplate irregularity and a large ventral paravertebral soft tissue mass (abscess).*



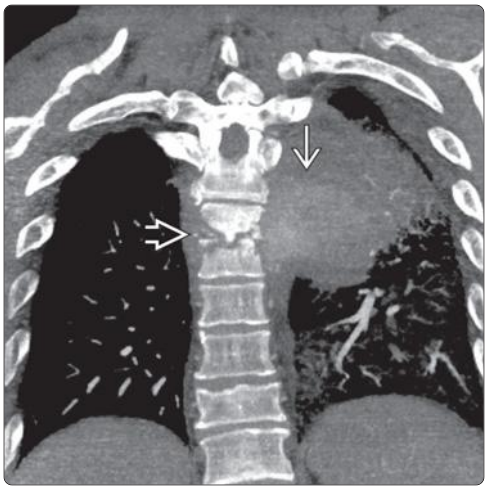
(Left) *Sagittal C+ MR shows multiple compartments involved by infection with destruction and collapse of the adjacent vertebral endplates, a large ventral abscess, and extension to involve the posterior elements.* **(Right)** *Sagittal STIR MR shows T1 and T2 collapse with endplate destruction and a large ventral abscess that displaces the anterior longitudinal ligament. There is extension to involve the posterior elements.*



(Left) *Sagittal T2 MR shows C5-C6 disc space infection with increased signal from the contiguous bodies and extensive prevertebral edema. There is a small epidural abscess that mildly effaces the cord.* **(Right)** *Sagittal T1 C+ MR shows enhancement of C5 and C6 bodies with mild disc irregularity and diffuse prevertebra soft tissue enhancement. Small ventral epidural abscess is also present.*

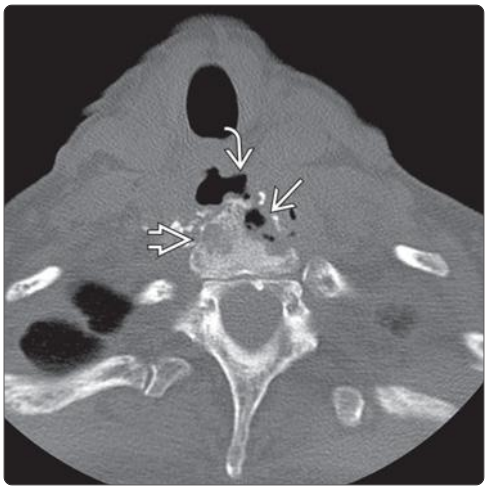






(Left) *Axial CTA shows aortic mycotic aneurysm from extension from the adjacent thoracic osteomyelitis. There is a large enhancing lesion consistent with pseudoaneurysm within mediastinum that displaces the aorta anteriorly. There is vertebral body destruction from the osteomyelitis.* **(Right)** *Coronal CTA in an aortic mycotic aneurysm from adjacent osteomyelitis shows the pseudoaneurysm and surrounding inflammatory mass, as well as bony destruction centered on the disc space.*





(Left) *Sagittal bone NECT shows a complicated case of esophageal perforation (prior neck irradiation and esophageal stricture) with fistula formation into the intervertebral disc (with adjacent vertebral osteomyelitis).* **(Right)** *Axial NECT (bone window) shows lytic destruction of the C7 vertebral body with gas extending from the posterior margin of the esophageal lumen into the intervertebral disc.*





(Left) *Sagittal STIR MR in a patient with esophageal fistula to disc shows abnormal increased signal extending from the thickened prevertebral soft tissues into the C7-T1 disc. Abnormal increased signal also presents within the cord due to either radiation effects or meningitis with myelitis.* **(Right)** *Sagittal T1 C+ MR shows diffuse enhancement of the prevertebral soft tissues adjacent to the fistula at C7 with evidence of diffuse meningitis with leptomeningeal enhancement.*

Tuberculous Osteomyelitis

KEY FACTS

TERMINOLOGY

- Tuberculous spondylitis (TS)
- Granulomatous infection of spine and adjacent soft tissue secondary to tuberculosis

IMAGING

- Gibbus vertebrae with relatively intact intervertebral discs, large paraspinal abscesses
- Midthoracic or thoracolumbar > lumbar, cervical
- Isolated posterior element involvement possible
- Sagittal STIR or FSE T2 with fat saturation most sensitive for bone marrow edema, epidural involvement
  - MR best modality to evaluate extent of disease, assess response to treatment

TOP DIFFERENTIAL DIAGNOSES

- Pyogenic spondylitis
  - Initial infection in subchondral bone
  - Intervertebral discs typically affected
- Fungal spondylitis
- Spinal metastases
  - Extraosseous epidural or paraspinal extension
  - Disc space preserved
- Brucellar spondylitis

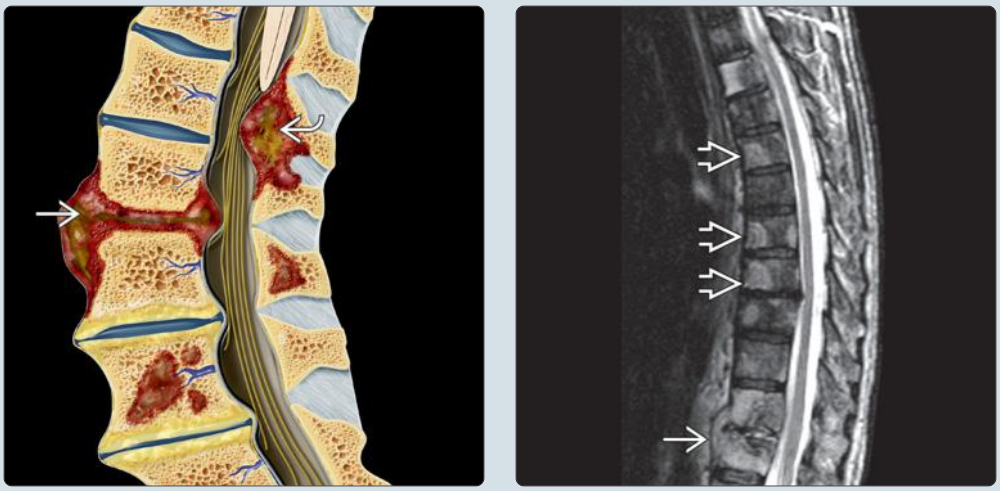
PATHOLOGY

- Hematogenous or lymphatic spread
- Initial inoculum in anterior vertebral body
- Spread to noncontiguous vertebral bodies beneath longitudinal ligaments

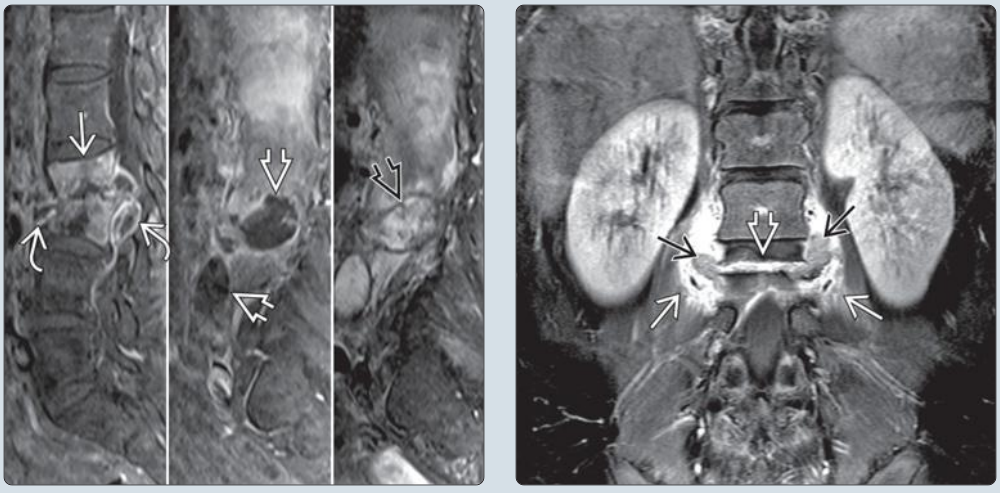
CLINICAL ISSUES

- Chronic back pain, focal tenderness, fever
- Neurologic deficits more common with TS than other granulomatous infections

(Left) *Sagittal graphic through lumbar spine depicts multifocal granulomatous osteomyelitis. Frank abscesses are present at L3-L4 disc space and between spinous process of L2 and L3.* **(Right)** *Sagittal STIR MR in a patient with TB infection shows involvement of contiguous vertebral bodies, with subligamentous abscess spread and partial disc involvement. Multiple focal bone lesions are present without adjacent disc involvement.*



(Left) *Sagittal T1WI C+ FS MR demonstrates focal kyphosis at L2-L3, collapse of the disc space, avid vertebral body enhancement, and ventral and dorsal paravertebral abscesses. There are peripherally enhancing abscesses in the paraspinal soft tissues, which exhibit hypointense rims.* **(Right)** *Coronal T1WI C+ FS MR shows TB osteomyelitis with L2 vertebra plana. Psoas involvement with swelling and marked enhancement is present. Inflammatory soft tissue surrounds the disc.*



TERMINOLOGY

Abbreviations

- Tuberculous spondylitis (TS)

Definitions

- Granulomatous infection of spine and adjacent soft tissue secondary to tuberculosis (TB)

IMAGING

General Features

- Best diagnostic clue
  - Gibbus vertebrae with relatively intact intervertebral discs, large paraspinal abscesses
- Location
  - Midthoracic or thoracolumbar > lumbar, cervical
  - Anterior vertebral body
  - Isolated posterior element involvement possible
  - Laminae > pedicles > spinous process > transverse process
- Size
  - Multiple (non)contiguous vertebrae
- Morphology
  - Vertebral collapse, gibbus deformity
  - ± destruction of intervertebral discs
  - Epidural soft tissue mass
  - Large dissecting paraspinal abscesses over considerable distance
  - TS has higher rate of psoas abscess and involvement of posterior elements compared to brucellar spondylitis

Radiographic Findings

- Radiography
  - Endplate irregularity, osteolysis
  - Diffuse vertebral sclerosis
  - Fusion across disc space in late TS
  - Findings may not be present until weeks after onset of infection

CT Findings

- NECT
  - Endplate destruction with vertebral body fragmentation
  - Calcified chronic paravertebral abscesses: TS > brucellar spondylitis (BS)
- CECT
  - Diffuse or peripherally enhancing epidural and paraspinal soft tissue

MR Findings

- T1WI
  - Hypointense marrow in contiguous vertebrae
  - Hypointense intraosseous, extradural, paraspinal abscesses
- T2WI
  - Hyperintense marrow, disc, phlegmon/abscess
- STIR
  - Hyperintense marrow, disc, phlegmon/abscess
- DWI
  - Vertebral bodies, endplates, and discs: Acute → hyperintense; chronic → hypointense
- T1WI C+
  - Marrow, subligamentous, discal, dural enhancement
  - Diffusely enhancing soft tissue (phlegmon)
  - Peripherally enhancing soft tissue (abscess)
- Epidural abscess → cord displacement or compression
- May have extradural infection without bone destruction
- Atypical findings in TS
  - Isolated vertebral body or posterior element involvement
    - Posterior element tuberculosis: 3-5% of TS
      - Potential for coronal decompensation
  - Sacral involvement

Nuclear Medicine Findings

- Bone scan
  - Increased spinal radionuclide uptake
  - Sensitive but not specific
- Gallium scan: Increased radionuclide uptake in spine and paraspinal soft tissue
  - Highly sensitive and specific for vertebral osteomyelitis

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI, T2WI, and T1 C+ MR
    - Evaluate extent of disease, assess response to treatment
- Protocol advice
  - Sagittal STIR or FSE T2 with fat saturation most sensitive for bone marrow edema, epidural involvement

DIFFERENTIAL DIAGNOSIS

Pyogenic Osteomyelitis

- Peak incidence in older patients
- Predilection for lower lumbar spine
- Initial infection in subchondral bone adjacent to endplate
  - Intervertebral discs typically affected
- Posterior element involvement less common
- Soft tissue calcifications and spinal deformity infrequent

Fungal Osteomyelitis

- May be indistinguishable from TS

Brucellar Spondylitis

- Anterosuperior epiphysitis at L4 with associated sacroiliitis
- May be indistinguishable from TS

Lytic and Blastic Osseous Metastases

- Hypointense T1, hyperintense T2 signal
  - Post-gadolinium enhancement
  - Posterior elements typically involved
- Extraosseous epidural or paraspinal extension
- Pathologic compression fractures
- Disc space preserved
- May be difficult to distinguish from isolated tuberculous, fungal, or BS

Degenerative Disc Disease

- Modic type 1 changes may mimic infection
  - Hypointense T1, hyperintense T2 signal
  - Inflammatory marrow change

PATHOLOGY

General Features

- Etiology
  - Hematogenous spread or through lymphatics from pulmonary origin
  - Initial inoculum in anterior vertebral body
  - Spread to noncontiguous vertebral bodies beneath longitudinal ligaments
  - Sparing of intervertebral disc secondary to lack of proteolytic enzymes
  - Paraspinal, subarachnoid dissemination of disease
  - Other pathogens causing granulomatous osteomyelitis (*Streptomyces*, *Madurella*) uncommon
- Associated abnormalities
  - Intramedullary abscess
  - Arachnoiditis
- Granulomatous destruction of spinal column with adjacent soft tissue infection

Microscopic Features

- Caseating granulomas, nonspecific inflammatory reaction
- Acid-fast bacilli isolated < 50% of time

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic back pain (~ 95%), focal tenderness, fever
- Other signs/symptoms
  - Paraparesis, kyphosis, sensory disturbance
  - Bladder and bowel dysfunction
  - Osseo-ligamentous destruction at cranio-vertebral junction → atlantoaxial instability and compression of cervico-medullary junction
    - → quadriparesis, bulbar dysfunction, and respiratory insufficiency
- Clinical profile
  - Gradual, insidious onset of symptoms results in diagnostic delay
  - Fever relatively infrequent in TS
  - Neurologic deficits more common with TS than other granulomatous infections, such as BS
  - TS has significant high rate of accompanying chronic renal failure, constitutional symptoms, history of tuberculosis, ↑ ESR, ± surgical treatment
  - QuantiFERON assay: Interferon-γ release assay
    - Sensitivity (84%), specificity (95%)
    - Combination of radiological criteria, bone scan, ELISA, QuantiFERON assay is 90% predictive of TS

Demographics

- Age
  - Most prevalent in 5th decade
- Gender
  - M = F in TS
- Epidemiology
  - Spinal TB accounts for 2% of all TB cases
    - Rising incidence of tuberculosis in past 2 decades
    - Trend toward increased incidence in parallel with growing number of immunocompromised patients
  - Concomitant pulmonary tuberculosis in ~ 10% of patients
  - Reactivation of latent tuberculosis with anti-TNF therapies
  - Tuberculosis-associated immune reconstitution inflammatory syndrome (TB-IRIS) → complication in HIV type I infected TB patients on antiretroviral treatment
    - Neurological manifestations occur in more than 10% of TB-IRIS cases
  - Evidence for increased risk of serious local infections after kyphoplasty in patients with history of systemic infection
  - TS more aggressive in children
    - Kyphosis, cord compression more common

Natural History & Prognosis

- Prognosis depends on early diagnosis and institution of appropriate therapy
- Proper treatment leads to
  - Favorable outcome with resolution of symptoms
    - Particularly favorable if early presentation and lack of neurologic deficits or spinal deformity
- No treatment leads to
  - Progressive vertebral collapse
  - Irreversible neurologic deficits
  - Death

Treatment

- Long-term antituberculous medication for at least 1 year
- Surgical decompression in setting of neurologic deficits ± spinal deformity
  - Indicated in 10-25% of TS
  - Laminectomy and debridement in absence of vertebral destruction
  - Debridement and fusion if spinal deformity present

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Thoracic spondylitis with posterior element involvement, large paraspinal abscesses suggests TS

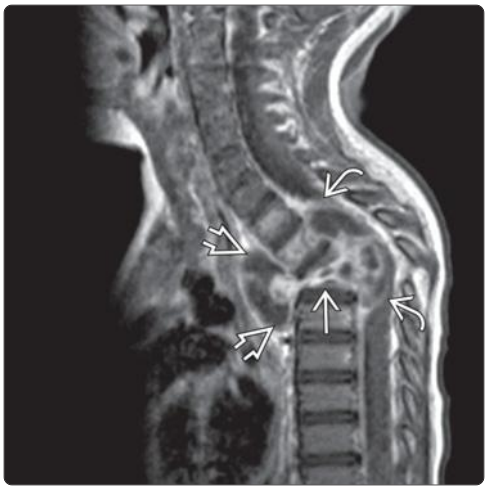
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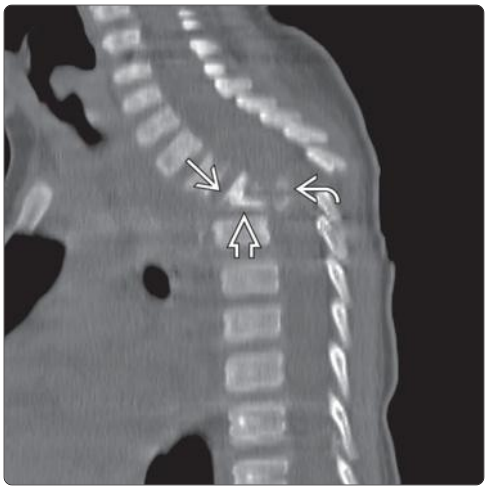
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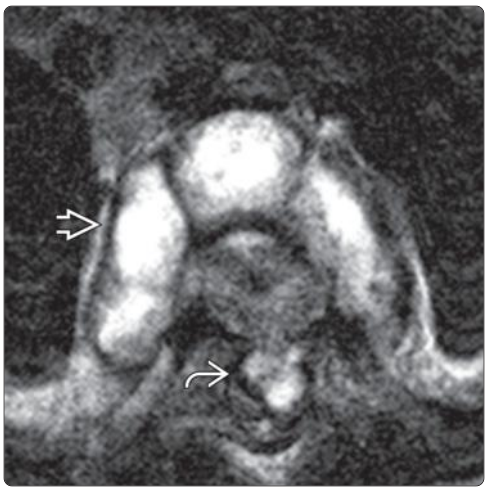


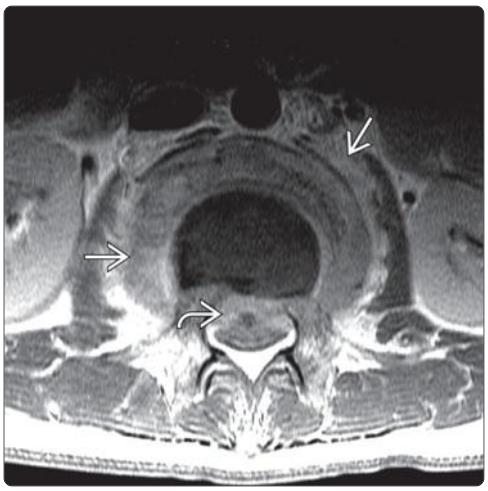
(Left) *Sagittal T1WI C+ MR in TB shows contiguous vertebral body extension with subligamentous abscess spread and partial disc involvement. Note multiple other focal bone lesions with mild epidural extension, which can mimic metastases.* **(Right)** *Sagittal T2WI FS MR demonstrates collapse of the L2 vertebral body. The adjacent intervertebral discs are now contiguous to each other. There is a focal abscess collection protruding posteriorly with compression of thecal sac contents.*





(Left) *Sagittal T1WI C+ MR illustrates thoracic spinal TB with abnormally enhancing vertebral bodies and kyphotic deformity. Large epidural abscess causes severe cord compression. The paraspinal abscess shows typical peripheral enhancement.* **(Right)** *Sagittal CT exhibits destruction of the T4-T6 bodies with kyphotic deformity and extension of calcific abscess/bone into the paravertebral region and spinal canal. Only residual T4 and T6 endplates remain after T5 destruction.*

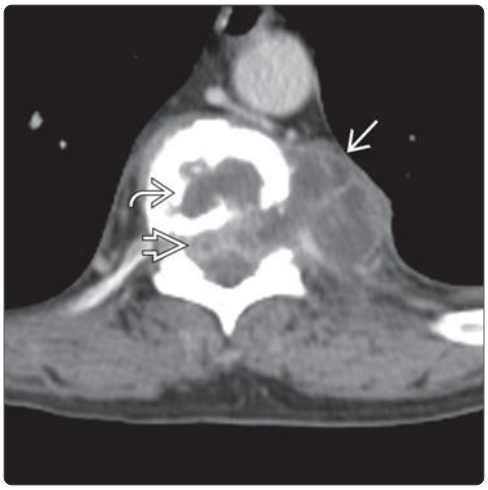




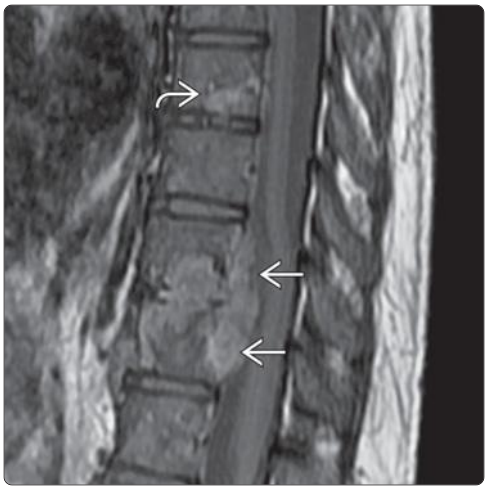
(Left) *Axial T2WI MR of a patient with thoracic TB demonstrates a large prevertebral abscess with extension into the spinal canal, which causes mild mass effect on the thecal sac.* **(Right)** *Axial T1WI C+ MR illustrates a focal abscess collection protruding posteriorly with compression of thecal sac contents. Inflammatory soft tissue surrounds the disc and extends into the adjacent psoas muscles. Pott disease, named after Percivall Pott, is extrapulmonary TB affecting the spine.*

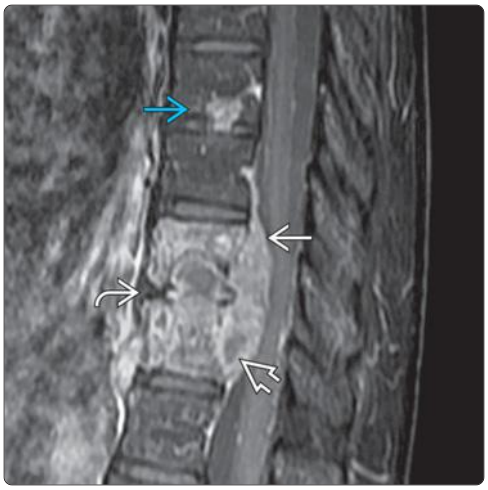
(Left) *Axial T1 C+ MR shows bilateral large psoas abscesses, right much larger than left in this patient with tuberculous spondylitis (TS).* **(Right)** *Contrast-enhanced CT shows a rim-enhancing abscess with central low attenuation. Enhancing material is present in the epidural space with cord compression. Large erosions are present in the vertebral body.*



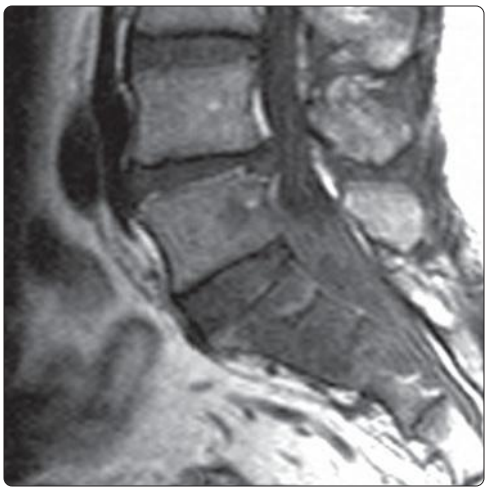


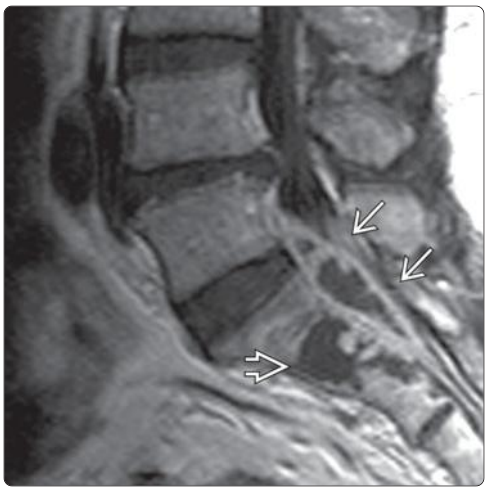
(Left) *Sagittal T1 MR without contrast shows typical pattern of disc space infection with large ventral epidural phlegmon in this patient with TS. Note the 2nd area of vertebral body involvement.* **(Right)** *Sagittal T1 C+ FS MR shows diffuse enhancement of the large ventral epidural phlegmon, with a small focus of nonenhancing abscess. There is disc space and adjacent vertebral body involvement. Note the 2nd vertebral body focus.*

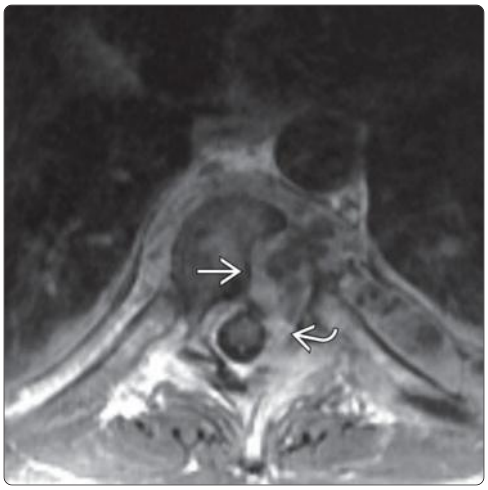


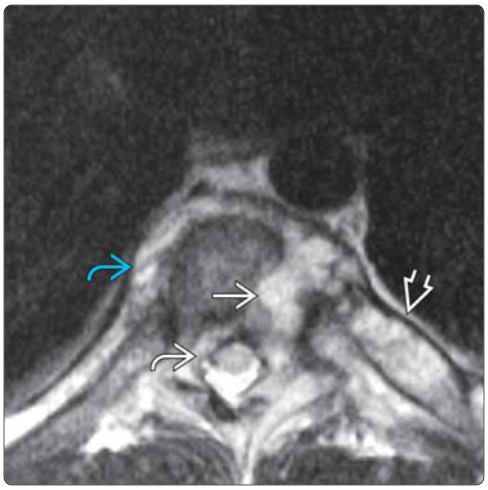


(Left) *Sagittal T1WI shows abnormal low signal from posterior L5, S1 and S2 bodies, and obscuration of thecal sac below L5 disc level. There is sparing of L5 disc. Patient was diagnosed with TS.* **(Right)** *Sagittal T1 C+ MR shows a large, nonenhancing area of bone abscess/necrosis with extension of large abscess into ventral epidural space compressing distal sac.*

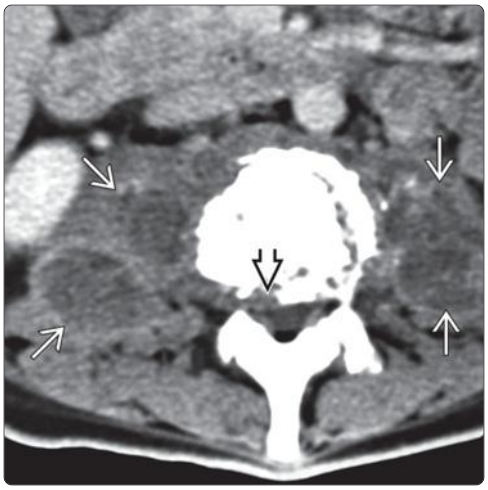








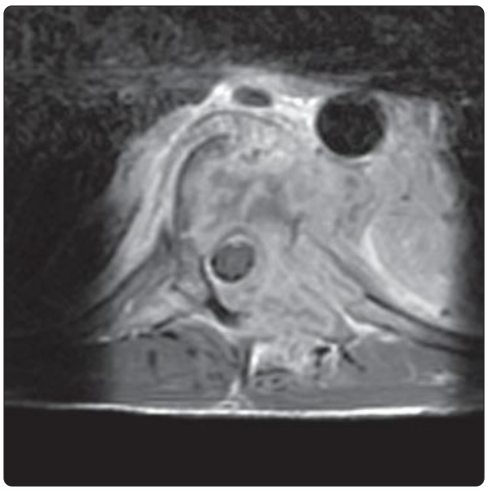
(Left) *Axial T1 C+ MR in this patient with brucellar osteomyelitis shows bone destruction with diffuse enhancing phlegmon within the epidural space and paravertebral region.* **(Right)** *Axial T2WI MR in this patient with brucellar osteomyelitis shows abnormal increased signal with bone destruction and expansion. There is involvement of epidural space and paravertebral region.*





(Left) *Axial CECT reveals a case of brucellosis granulomatous discitis-osteomyelitis. There is vertebral destruction and bilateral psoas abscesses present. There is also epidural extension narrowing the central spinal canal.* **(Right)** *CT study shows multiple focal areas of bone destruction involving L2, L3, and L5 with preservation of the disc spaces and minimal prevertebral soft tissue in this patient with coccidioidomycosis.*





(Left) *Sagittal STIR MR shows diffuse abnormal T2 signal from bone destruction and soft tissue phlegmon involving multiple thoracic bodies and posterior elements, conspicuously sparing the intervertebral discs. Pathology was coccidioidomycosis.* **(Right)** *Axial T1 C+ MR shows soft tissue enhancement and involvement of both vertebral body and posterior elements with epidural extension. Note the paraspinal and adjacent lung extension. Patient had disseminated coccidioidomycosis.*

KEY FACTS

TERMINOLOGY

- Noncaseating, acid-fast negative or fungal infections primarily occurring as opportunistic infection in immunocompromised patient
- Involvement of spine and adjacent soft tissue typically secondary to fungal pathogen

IMAGING

- Osseous destruction ± disc, epidural, or paraspinal involvement
  - Mixed lytic and sclerotic foci within vertebral bodies
  - Marrow edema
  - ± paravertebral mass, epidural phlegmon
- Diffuse > focal, lobulated contours
- May produce spinal deformity

TOP DIFFERENTIAL DIAGNOSES

- Pyogenic osteomyelitis
- Granulomatous osteomyelitis
  - Tuberculosis
  - Brucellosis

PATHOLOGY

- Hematogenous spread
- Direct extension from adjacent tissues
  - Direct implantation from trauma, hematogenous, local extension, iatrogenic after lumbar puncture, nucleoplasty

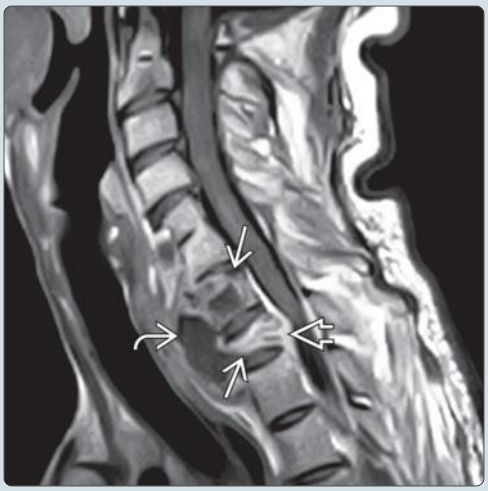
CLINICAL ISSUES

- Neck pain or back pain
- Clinical signs of systemic illness
- Risk factors: Immunosuppression, diabetes, hemodialysis, corticosteroid use, chemotherapy, or malnutrition

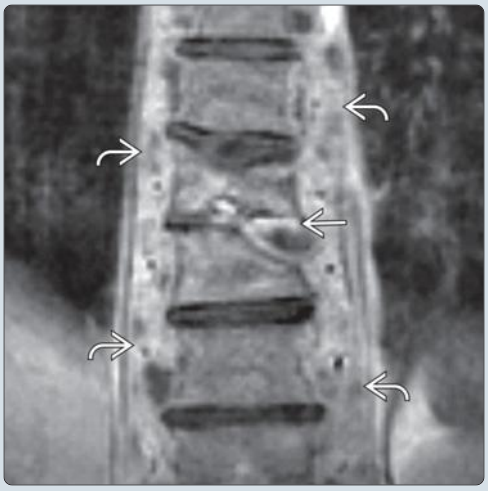
DIAGNOSTIC CHECKLIST

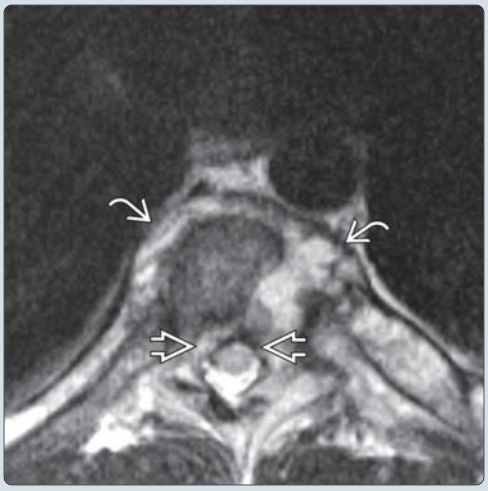
- Also consider fungal entities when tuberculosis is in imaging differential diagnosis list





(Left) *Diffuse abnormal low T1 signal (coccidioidomycosis) reflects diffuse anemia and marrow hyperplasia ± marrow infiltration. Note bone destruction with soft tissue mass involving multiple midthoracic bodies and posterior elements, sparing intervertebral discs.* **(Right)** *Sagittal T1 C+ MR (coccidioidomycosis) shows a rim-enhancing prevertebral abscess due to chronic infection. There is destruction of C7 and T1 vertebral bodies, relative sparing of the intervertebral discs, and ventral epidural phlegmon.*





(Left) *Coronal T1WI C+ MR (blastomycosis) demonstrates narrowing and destruction of a thoracic disc space with vertebral marrow signal abnormality and compression fracture. There is extensive paraspinal phlegmon and heterogeneous enhancement of the abnormal disc space.* **(Right)** *Axial T2WI MR (blastomycosis) illustrates hyperintense paraspinal phlegmon. A ventral epidural mass mildly displaces the cord without cord signal abnormality.*

TERMINOLOGY

Definitions

- Noncaseating, acid-fast negative or fungal infections primarily occurring as opportunistic infections in immunocompromised patients
  - Involvement of spine and adjacent soft tissue typically secondary to fungal pathogen

IMAGING

General Features

- Best diagnostic clue
  - Osseous destruction ± disc, epidural, or paraspinal involvement
- Morphology
  - Diffuse > focal, lobulated contours
  - Intervertebral disc sparing, simultaneous thoracic infiltrates, intraspinal disease, and intramuscular abscesses suggest actinomycosis

Radiographic Findings

- Mixed lytic and sclerotic foci within vertebral bodies

CT Findings

- NECT
  - Mixed lytic and sclerotic lesions within vertebral bodies
  - Osseous destruction → spinal deformity

MR Findings

- ± paravertebral mass, epidural phlegmon
- ± disc involvement
  - Actinomycosis commonly spares disc space

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR with contrast of entire spine to assess full disease extent

DIFFERENTIAL DIAGNOSIS

Pyogenic Osteomyelitis

- Ill-defined hypointense T1 vertebral marrow, loss of endplate definition on both sides of disc

Granulomatous Osteomyelitis

- Tuberculosis: Gibbus vertebrae with relatively intact intervertebral discs, large paraspinal abscesses
- Brucellosis: Anterosuperior epiphysitis at L4 with sacroiliitis, disc involvement, large paraspinal abscesses

PATHOLOGY

General Features

- Etiology
  - Hematogenous spread or direct extension from adjacent tissues
  - Coccidiomycosis, blastomycosis, invasive aspergillosis, candidiasis, cryptococcosis
    - Fungal infection ± coexistent with bacterial infection
  - Uncommon compared to bacterial pathogens

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck pain/back pain
  - Blastomycosis, actinomycosis: Pneumonia is primary infection
    - Bone and joint lesions account for 1/4 to 1/2 of all extrapulmonary dissemination
    - Bone is infected by hematogenous dissemination or direct invasion from adjacent tissues
- Other signs/symptoms
  - Glucatell serum β-D-glucan detection assay
  - Diagnosis → potassium hydroxide (KOH) preparation, polymerase chain reaction (PCR)

Demographics

- Epidemiology
  - Risk factors: Immunosuppression, diabetes, hemodialysis, corticosteroid use, chemotherapy for tumor, or malnutrition
  - Direct implantation from trauma, hematogenous, local extension, iatrogenic after lumbar puncture, nucleoplasty

Natural History & Prognosis

- Prolonged delay in initiation of antifungal treatment is associated with poorer outcome
  - Majority of patients with > 60-day delay in treatment → motor deficits and incomplete recovery
- Clinical course and prognosis of postoperative fungal spondylodiscitis equals that reported for postoperative pyogenic spondylodiscitis

Treatment

- Aggressive antifungal therapy
  - Preferred treatment for blastomycosis: Itraconazole, amphotericin B
  - Successfully treated patients → resolution of pain, may fuse spontaneously
- Abscess drainage, bone fusion, posterior instrumentation, surgical reconstruction of deformity
  - Spinal decompression ± stabilization → those not responding to pharmacologic treatment
    - → those with progressive or severe neurologic deficits, spinal deformity, or instability

DIAGNOSTIC CHECKLIST

Consider

- Consider fungal entities when tuberculosis is in imaging differential diagnosis list
- Mixed lytic and sclerotic foci within vertebral bodies

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Osteomyelitis, C1-C2

KEY FACTS

TERMINOLOGY

- Infection of C1-C2 articulation (pyogenic or tuberculous)

IMAGING

- Soft tissue mass within prevertebral space centered at C1-C2
  - Prevertebral increased soft tissue/edema
  - Variable extension into epidural space, dural sac, or cord compression
  - Low T1 signal, increased T2/STIR signal from vertebral bodies, soft tissue mass
- ± bone destruction involving anterior arch of C1, odontoid, and body of C2
- Diffuse enhancement of vertebral bodies, soft tissue mass within prevertebral region/epidural space
  - May show nonenhancing abscess focus
- MRA/CTA: Evaluation of skull base, C1-C2 instability with vertebral artery compromise

TOP DIFFERENTIAL DIAGNOSES

- Rheumatoid arthritis
- Odontoid fracture
- C1-C2 osteoarthritis
- Primary bone tumor/metastases

PATHOLOGY

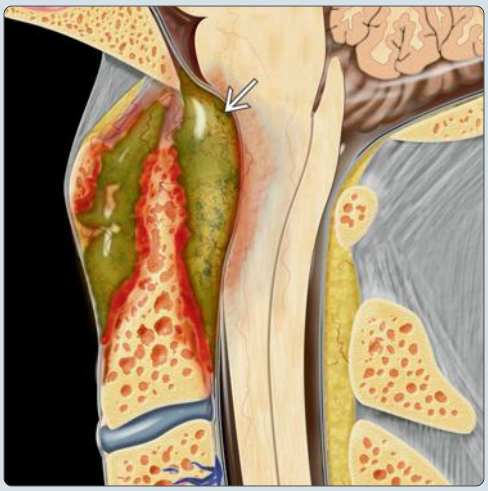
- Hematogenous seeding to capillary ends/end arterioles in subchondral regions

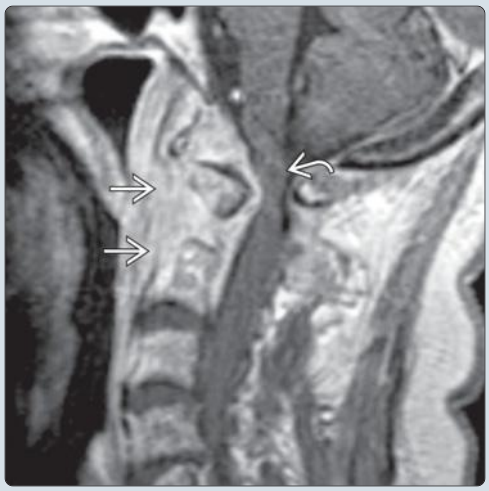
CLINICAL ISSUES

- Neck pain, limited range of motion, dysphagia
- C1-C2 subluxation, medulla compression, and motor/sensory deficit
- Vertebral artery compression with posterior circulation infarction

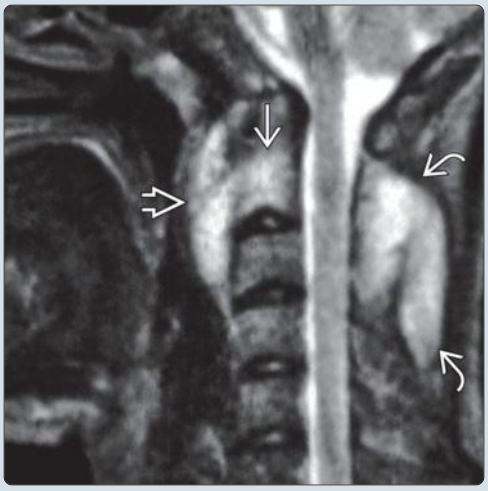
DIAGNOSTIC CHECKLIST

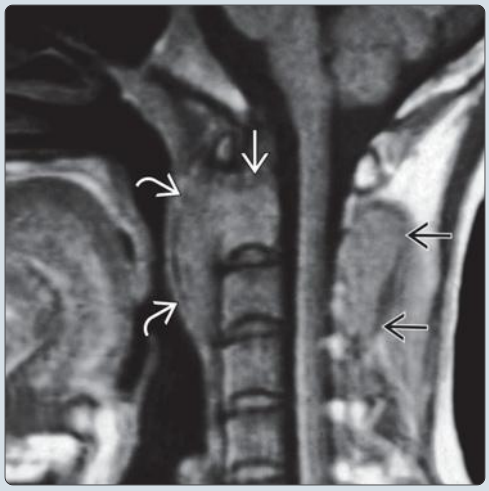
- Severe C1-C2 subluxation ± epidural phlegmon/abscess





(Left) *Sagittal graphic illustrates osteomyelitis involving the odontoid with bone destruction, extension to the anterior arch of C1, and formation of epidural abscess ⇒.* **(Right)** *Sagittal T1WI C+ MR demonstrates C1-C2 osteomyelitis with extensive enhancement of the C2 body and odontoid, and considerable prevertebral phlegmon ⇒. There is atlantoaxial subluxation and cord compression ⇒. The cervical spine is a rare site of bacterial epidural abscess, accounting for only 10-15% of such abscesses.*





(Left) *Sagittal T2WI MR shows tuberculous (TB) osteomyelitis involving the C2 body ⇒ and C2 and C3 posterior elements ⇒. There is extension into the prevertebral space ⇒.* **(Right)** *Sagittal T1 MR shows TB osteomyelitis that involves C2 body ⇒ with a prevertebral phlegmon ⇒, spares the disc spaces, and preferentially involves the posterior elements ⇒. Presence of a multilocular, calcified abscess with a thick enhancing irregular rim in the presence of vertebral body fragmentation is pathognomonic of tuberculosis.*

TERMINOLOGY

Synonyms

- Atlantoaxial osteomyelitis

Definitions

- Infection of C1-C2 articulation (pyogenic or tuberculous)

IMAGING

General Features

- Best diagnostic clue
  - Soft tissue mass and bone destruction at C1-C2 level
- Location
  - C1 anterior arch, atlantoaxial joint, dens, C2 body

Radiographic Findings

- Radiography
  - Gross bone destruction manifested late in course
    - Sensitivity (80%), specificity (50-60%)
  - Early changes with indistinct anterior margin of C1, prevertebral soft tissue swelling

Fluoroscopic Findings

- Useful in assessing C1-C2 subluxation

CT Findings

- NECT
  - Variable bone destruction involving anterior arch of C1, odontoid, and body of C2
  - Soft tissue mass adjacent to bone lesions within prevertebral space, variable extension into epidural space
- CECT
  - Enhancement of soft tissue component; may show nonenhancing abscess focus
- CTA
  - Useful if question of skull base, C1-C2 instability with vertebral artery compromise

MR Findings

- T1WI
  - Low signal mass centered at C1-C2 with variable involvement of odontoid and lateral masses
  - May show enlarged atlantodental interval
  - Prevertebral increased soft tissue/edema
  - Epidural mass with thecal sac/cord compression
- T2WI
  - Diffuse increased signal from vertebral bodies, soft tissue mass
- STIR
  - Diffuse increased signal from vertebral bodies, soft tissue mass
- DWI
  - May show restricted diffusion of abscess component
  - ± brain DWI abnormality in posterior circulation if vertebral artery compromise
- T1WI C+
  - Diffuse enhancement of vertebral bodies, soft tissue mass within prevertebral region/epidural space
    - Phlegmon shows diffuse enhancement
    - Abscess shows peripheral enhancement, central nonenhancing pus
- MRA
  - Useful if question of skull base, C1-C2 instability with vertebral artery compromise

Nonvascular Interventions

- Myelography
  - May show nonspecific extradural defect in contrast column at C1-C2 related to epidural extension of infection

Nuclear Medicine Findings

- Bone scan
  - Tc-99m methylene diphosphonate SPECT
    - Arterial hyperemia with progressive increased uptake in osteomyelitis
  - High sensitivity (90%), low specificity (75%)
    - 3-phase bone scanning overall accuracy (90%)
    - Specificity increased by combining indium-labeled WBC scan or gallium-67 scan
    - Unreliable for diagnosis of active TB (scans cold in 35-40%)
- PET
  - FDG PET shows nonspecific increased uptake with infection, tumor
    - No uptake with degenerative endplate changes
- Gallium scan
  - Increased uptake, nonspecific
- WBC scan
  - Increased uptake in acute phase
  - High specificity, low sensitivity

Imaging Recommendations

- Best imaging tool
  - MR with contrast ± MRA; shows bone involvement, prevertebral soft tissues, epidural space, vertebral arteries
- Protocol advice
  - T1WI, T2WI axial, sagittal images; post-contrast axial, sagittal with fat suppression; 3D TOF MRA

DIFFERENTIAL DIAGNOSIS

C1-C2 Osteoarthritis

- May show pseudopannus from osteoarthritic degenerative change
- Bone irregularity, low signal, and soft tissue mass dorsal to odontoid

Rheumatoid Arthritis

- Erosion of odontoid, soft tissue pannus, C1-C2 subluxation, subaxial subluxations

Chordoma

- Soft tissue mass, increased T2WI signal

Metastatic Disease

- Focal low signal on T1WI, with bone destruction, epidural extension
- Multiple lesions

Extension of Skull Base Osteomyelitis

- Sphenoid, petrous apex inflammatory change with secondary inferior extension

Odontoid C2 Fracture

- Fracture line defined by GRE, T1WI, STIR shows marrow/prevertebral edema

Hemodialysis Spondyloarthropathy

- Low signal on T1WI involving endplates similar to pyogenic disc space infection
- May be indistinguishable from pyogenic disc space infection

Gout

- Rarely vertebral involvement with low signal involving endplates, similar to disc space infection

PATHOLOGY

General Features

- Etiology
  - Hematogenous seeding to capillary ends/end arterioles in subchondral regions
  - Many vascular anastomoses
    - Pharyngovertebral plexus drains posterosuperior nasopharynx inferiorly to basiocciput penetrating atlantooccipital membrane
    - Pharyngovertebral plexus ↔ lymphatics
    - Periodontoidal venous plexus ↔ pharyngovertebral veins
    - Batson plexus allows seeding from viscera
  - *Staphylococcus aureus* most common organism in USA
  - *Mycobacterium tuberculosis* most common worldwide
  - *Brucella*, *Pseudomonas*, *Serratia*, and *Candida* organisms common in IV drug addicts, immunocompromised patients
  - Infection starts as septic arthritis of C1-C2
- Associated abnormalities
  - C1-C2 subluxation, cervicomedullary junction compression; vertebral artery compression

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute and chronic neck pain, stiffness, limited range of motion, dysphagia
  - Other signs/symptoms
    - Triad of fever, pain, and neurologic deficit in minority of cases
      - 4 stages: Spinal ache, root pain, weakness, and paralysis
    - Severe neck spasm
    - C1-C2 subluxation, medulla compression, and motor/sensory deficit
    - Vertebral artery compression with posterior circulation infarction
- Other signs/symptoms
  - Reports of Collet-Sicard syndrome resulting from cranial-based infection → multiple cranial nerve palsies and extremity symptoms
  - ↑ ESR, ↑ CRP, peripheral leukocytosis
  - ≥ 50% associated with bacteremia, particularly *S. aureus*

Demographics

- Age
  - Adult > child
- Gender
  - No gender preference
- Epidemiology
  - Uncommon; most cervical infections occur in C5 and C6 region
  - Risk factors include advanced age, immunosuppression, alcoholism, diabetes, end-stage renal disease, drug abuse, endocarditis
  - Reports after steroid injection into C1-C2 facet joint

Natural History & Prognosis

- Increasing age and degree of thecal sac compression
  - Independent association with poor outcome

Treatment

- Open surgical decompression/stabilization with subsequent long-term IV antibiotics favored treatment
  - Variable approaches; anterior debridement and posterior cervical-occipital arthrodesis
- Successful medical management reported
  - Initial course of IV antibiotics if patient neurologically stable
    - IV antibiotics, followed by prolonged oral antibiotic administration
  - Identify offending organism via blood cultures ± biopsy if patient unstable
  - Surgical intervention for patients with significant or progressive neurological deficits

DIAGNOSTIC CHECKLIST

Consider

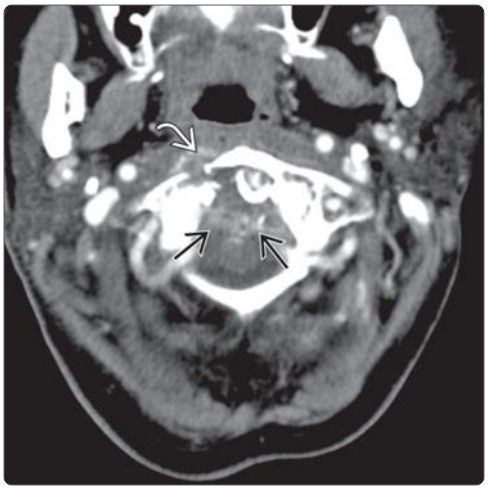
- Grisel syndrome: Inflammatory, nontraumatic subluxation of C1-C2 following peripharyngeal infection

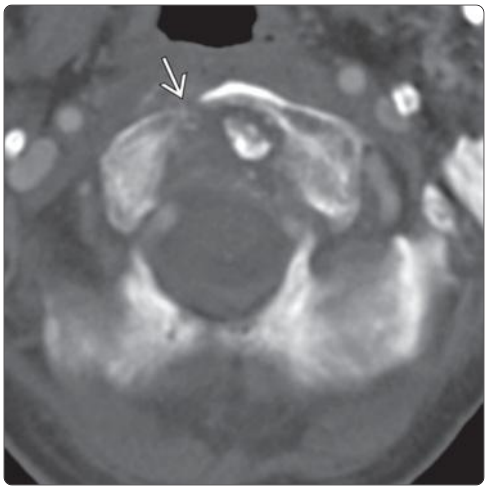
Image Interpretation Pearls

- Severe C1-C2 subluxation ± epidural phlegmon/abscess
  - ± posterior circulation infarcts due to vertebral compression
  - ± medullary compression

SELECTED REFERENCES

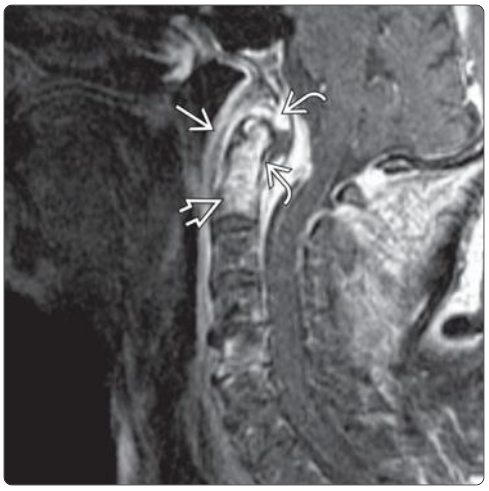
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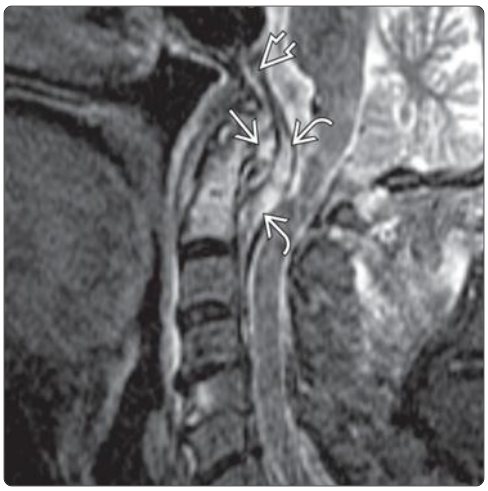


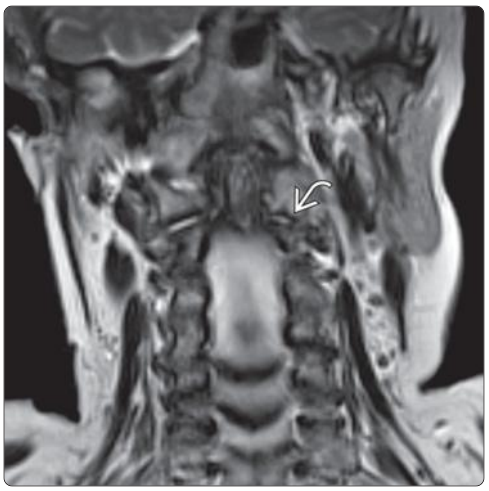
(Left) *Axial CECT shows a heterogeneously ventral epidural collection at the atlantoaxial articulation. There is osseous destruction and extension into the prevertebral space. There is mild mass effect on the ventral thecal sac.* **(Right)** *Axial NECT exhibits the osseous destruction of the C1 anterior arch due to Staphylococcus aureus osteomyelitis. Destruction of the dens is typically caused by rheumatoid arthritis; however, destruction due to purulent infection is quite common.*





(Left) *Sagittal T1WI MR illustrates abnormal hypointensity in the C2 body and dens. There is an associated retrodental soft tissue lesion displacing the cervicomedullary junction posteriorly.* **(Right)** *Sagittal T1WI C+ FS MR demonstrates avid enhancement of the C2 vertebral body. There is abnormal prominent soft tissue in the prevertebral space. In the retrodental region, there is a heterogeneously enhancing fluid collection consistent with an epidural abscess.*





(Left) *Sagittal STIR MR shows not only an epidural abscess, but also a subdural fluid collection. The epidural space is anatomically closed at the foramen magnum where the spinal dura attaches with the endosteal dura of the cranium. The subdural space is a potential space extending along the retroclival region.* **(Right)** *Coronal T2WI MR depicts left C1-C2 facet osteoarthritis. There is irregularity of the endplates and decrease in the joint space. The dark cortical line is preserved.*

Brucellar Spondylitis

KEY FACTS

TERMINOLOGY

- Brucellar spondylitis (BS)
- Granulomatous infection of spine and adjacent soft tissue due to brucellosis

IMAGING

- Lower lumbar spine (L4) > cervical = thoracic
  - Anterosuperior epiphysitis at L4
  - Associated sacroiliitis
- May have extradural infection without bone destruction
  - Epidural abscess → cord displacement or compression
- 2 presentations
  - Focal BS: Anterior endplate at discovertebral junction involved
  - Diffuse BS: Entire vertebral body affected

TOP DIFFERENTIAL DIAGNOSES

- Tuberculosis osteomyelitis
- Pyogenic osteomyelitis
- Fungal osteomyelitis
- Neurogenic (Charcot) arthropathy

PATHOLOGY

- Hematogenous dissemination to spine
  - Direct extension to adjacent discs and vertebrae

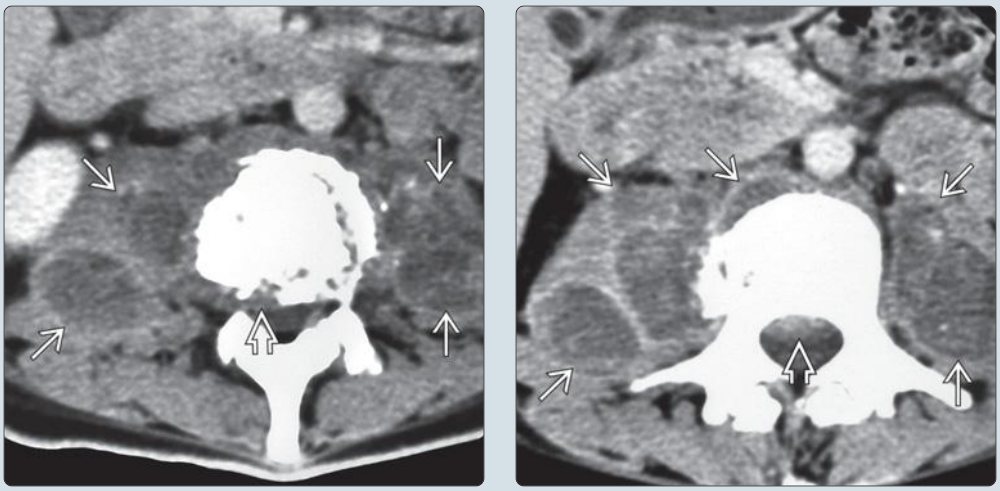
CLINICAL ISSUES

- Chronic back pain, focal tenderness, fever
- Prognosis depends on early diagnosis and institution of appropriate therapy
  - Antibrucellar medications highly effective
  - Surgical debridement rarely indicated

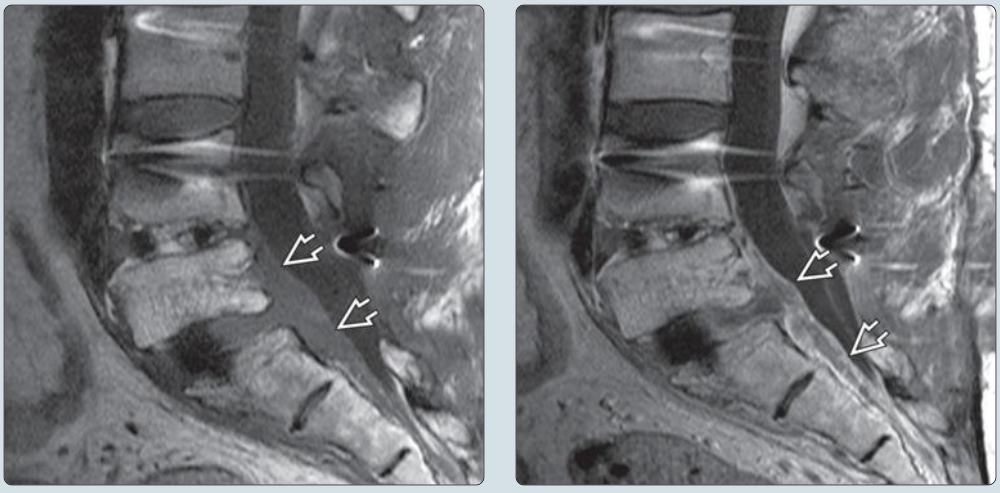
DIAGNOSTIC CHECKLIST

- BS should be differential diagnosis of L4 lumbar spondylitis with associated bilateral sacroiliitis
- MR best modality to evaluate extent of disease, assess response to treatment

(Left) *Axial CECT demonstrates vertebral destruction and bilateral psoas muscle abscesses ➪. There is also epidural extension narrowing the central spinal canal ➪.* **(Right)** *Axial CECT depicts vertebral destruction and bilateral psoas abscesses ➪. There is also epidural extension narrowing the central spinal canal ➪. In the acute stage on the DWI, vertebral bodies, endplates, and discs are all hyperintense but hypointense in the chronic stage.*



(Left) *Sagittal T1WI MR shows hypointense epidural soft tissue ➪ lifting the posterior longitudinal ligament and spanning from L4-L5 to S2. Vertebral bodies, endplates, and disc spaces are T1 hypointense/T2 hyperintense in the acute stage and T1 hypointense/T2 heterogeneous in the subacute and chronic stages.* **(Right)** *Sagittal T1WI C+ MR reveals heterogeneously enhancing soft tissue ➪ in the ventral epidural space reflecting an epidural abscess related to brucellosis infection.*



TERMINOLOGY

Abbreviations

- Brucellar spondylitis (BS)

Definitions

- Granulomatous infection of spine and adjacent soft tissue secondary to brucellosis

IMAGING

General Features

- Best diagnostic clue
  - Anterosuperior epiphysitis at L4 ± sacroiliitis
- Location
  - Lower lumbar spine (L4) > cervical = thoracic
    - Posterior elements not affected
  - Sacroiliac joints
  - Focal: Anterior endplate at discovertebral junction
  - Diffuse: Entire vertebral body
- Size
  - Multiple (non)contiguous vertebrae
- Morphology
  - Vertebrae morphologically intact despite osteomyelitis
    - Spinal deformity rare
  - Destruction of intervertebral discs
  - Epidural soft tissue mass
  - Paraspinal soft tissues rarely affected

Radiographic Findings

- Endplate irregularity, osteolysis
- Focal vertebral sclerosis
- Fusion across disc space in late BS
- Findings may not be present until weeks after onset of infection

CT Findings

- Endplate destruction with fragmentation of vertebral bodies
- Calcified chronic paravertebral abscesses: BS < TB osteomyelitis
- Diffuse or peripherally enhancing epidural, paraspinal soft tissue

MR Findings

- T1WI
  - Hypointense marrow in contiguous vertebrae
  - Hypointense intraosseous, extradural, paraspinal abscesses
- T2WI
  - Hyperintense marrow, disc, phlegmon/abscess
- DWI
  - Vertebral bodies, endplates, and discs
  - Acute → hyperintense; chronic → hypointense
- T1WI C+
  - Marrow, subligamentous, discal, dural enhancement
  - Diffusely enhancing soft tissue (phlegmon)
  - Peripherally enhancing soft tissue (abscess)
    - Epidural abscess → cord displacement or compression
- May see extradural infection without bone destruction

DIFFERENTIAL DIAGNOSIS

Tuberculosis Osteomyelitis

- Gibbus vertebrae with relatively intact intervertebral discs, large paraspinal abscesses

Pyogenic Osteomyelitis

- Predilection for lower lumbar spine
- Initial infection in subchondral bone adjacent to endplate
  - Intervertebral discs typically affected

Fungal Osteomyelitis

- May be indistinguishable from BS

Neurogenic (Charcot) Arthropathy

- Destructive lumbar arthropathy in setting of pain, proprioception impairment, and joint mobility preservation

PATHOLOGY

General Features

- Etiology
  - Hematogenous dissemination to spine
    - Direct extension to adjacent discs and vertebrae
  - Intramedullary abscess

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic back pain (~ 95%), focal tenderness, fever (up to 92%)
- Other signs/symptoms
  - Paraparesis, kyphosis, sensory disturbance
  - Bladder and bowel dysfunction
  - Neurologic deficits less common with BS than TB

Demographics

- Epidemiology
  - Uncommon in USA: 100-200 cases per year
    - Prevalent in Mediterranean, South and Central America, and Middle East (6-58% incidence)

Natural History & Prognosis

- Prognosis depends on early diagnosis and institution of appropriate therapy

Treatment

- Antibrucellar medications highly effective
- Surgical debridement rarely indicated

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- BS should be differential diagnosis of L4 lumbar spondylitis with associated bilateral sacroiliitis

SELECTED REFERENCES

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3. Sanaei Dashti A et al: Skeletal Involvement of Brucella melitensis in Children: A Systematic Review. Iran J Med Sci. 38(4):286-92, 2013

Septic Facet Joint Arthritis

KEY FACTS

TERMINOLOGY

- Suppurative bacterial infection of facet joint, adjacent soft tissue

IMAGING

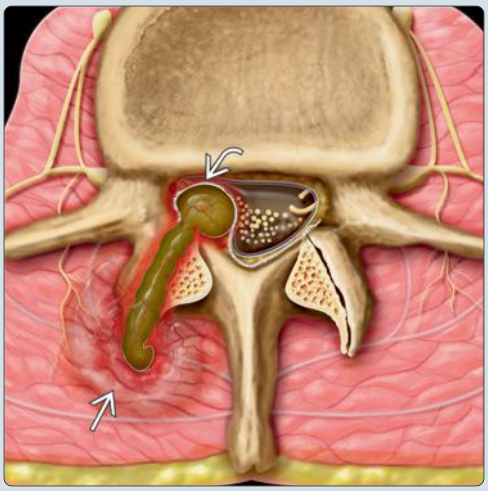
- Abnormal enhancement within facet joint with associated facet marrow, adjacent soft tissue edema
  - Typically single level, unilateral involvement
  - Facet joint widening
  - Ill-defined facet marrow signal alteration
  - Eroded facet cortex
- Lumbar spine most common
- Protocol advice
  - Sagittal STIR or FSE T2 with fat saturation most sensitive for bone marrow edema and epidural involvement
  - Post-gadolinium T1WI with fat saturation better delineates extent of facet, epidural, and paraspinal involvement

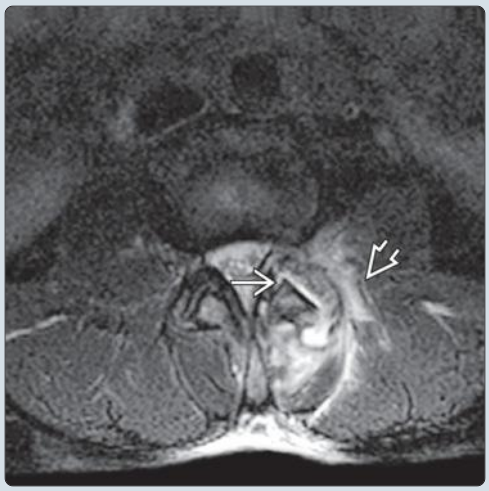
TOP DIFFERENTIAL DIAGNOSES

- Facet joint osteoarthritis
  - Bilateral joint space narrowing with vacuum phenomenon
- Facet synovial cyst
  - Juxtaarticular thin-walled, well-defined mass
- Rheumatoid arthritis
  - Widened facets with enhancing synovium
  - Facet joint erosion

PATHOLOGY

- Most common cause: Hematogenous contamination
- Associated findings
  - Spondylodiscitis
  - Epidural paravertebral abscess





(Left) *Axial graphic at a lumbar vertebral level demonstrates a septic right facet joint with adjacent osteomyelitis and abscess extending into subarticular recess and posterior paraspinal muscle.* **(Right)** *Axial T2WI MR with fat suppression through a lumbar vertebra shows fluid in the left facet joint with posterior extension and loculation. Edema is present in surrounding soft tissue.*





(Left) *Sagittal STIR MR shows multilevel discitis-osteomyelitis with interdiscal fluid and adjacent endplate erosive changes. There is also infection of the facet joints of 2 adjacent levels. Fluid is seen within the facet joints. There is marrow edema of the articular pillars as well as the surrounding soft tissues.* **(Right)** *Sagittal T1WI C+ FS MR depicts the peri facet inflammation associated with septic facet arthritis. Epidural phlegmon extends into the neural foramen.*

TERMINOLOGY

Synonyms

- Pyogenic facet joint infection

Definitions

- Suppurative bacterial infection of facet joint, adjacent soft tissue

IMAGING

General Features

- Best diagnostic clue
  - Abnormal enhancement within facet joint with associated facet marrow, adjacent soft tissue edema
- Location
  - Lumbar spine most common (86-97%)
- Size
  - Typically single-level involvement
    - Unilateral
- Morphology
  - Facet joint widening
  - Eroded facet cortex
  - Ill-defined facet marrow signal alteration

Radiographic Findings

- Radiography
  - May be negative up to 2-8 weeks after onset of infection
  - Osteolytic/sclerotic facet joint
  - ± instability on flexion/extension views

CT Findings

- NECT
  - Low density within expanded facet joint
  - ± juxtaarticular, epidural, and paraspinal phlegmon or abscess
- CECT
  - Diffuse or rim enhancement within facet joint
    - ± extraarticular extension
  - Diffuse or peripherally enhancing epidural and paraspinal soft tissue

MR Findings

- T1WI
  - Hypointensity within facet joint
  - Ill-defined hypointense facet marrow
- T2WI
  - Hyperintensity within facet joint
  - Periarticular soft tissue edema
- STIR
  - Hyperintense facet marrow
- T1WI C+
  - Diffuse or rim enhancement within joint
  - Enhancing marrow abutting facet joint
  - Enhancing soft tissue or peripherally enhancing fluid collection
    - Contiguous with facet joint
    - Epidural or paraspinal extension

Nuclear Medicine Findings

- Bone scan
  - Uptake of Tc-99m diphosphonate in posterior elements
    - More laterally located and vertically oriented
    - Improved lesion localization with SPECT
- Gallium scan
  - Increased uptake of gallium citrate (Ga-67)
    - Increased sensitivity with SPECT

Imaging Recommendations

- Best imaging tool
  - MR, particularly fat-suppressed post-contrast T1W images
- Protocol advice
  - Sagittal STIR or FSE T2 with fat saturation most sensitive for bone marrow edema and epidural involvement
  - Post-gadolinium T1WI with fat saturation better delineates extent of facet, epidural, and paraspinal involvement

DIFFERENTIAL DIAGNOSIS

Facet Joint Osteoarthritis

- Joint space narrowing, vacuum phenomenon
- Cortical eburnation with subcortical cysts
- Osteophytosis
- Marrow T2 signal reflects stress reaction
- ± fluid within facet joint
- Tends toward bilateral symmetry
- Associated ligamentum flavum thickening
- ± spondylolisthesis
- No soft tissue edema or abscess
- Normal erythrocyte sedimentation rate, C-reactive protein

Facet Synovial Cyst

- Juxtaarticular thin-walled, well-defined mass
  - Hypointense on T1WI
  - Hyperintense on T2WI
  - ± mild peripheral enhancement
- No marrow signal abnormality or cortical erosion
- No soft tissue edema or enhancement
- Associated facet arthropathy and ligamentum flavum thickening

Rheumatoid Arthritis

- Cervical spine most commonly affected
- Atlantoaxial subluxation & dens erosion most common findings
- Facet joint erosion with enhancing synovium
- Enhancing soft tissue pannus

Metastases

- Multifocal discrete or ill-defined lesion in posterior elements
  - Hypointense on T1WI
  - Hyperintense on T2WI
  - Post-gadolinium enhancement
  - ± surrounding marrow edema
- Osseous destruction
- Facet joint may be preserved

PATHOLOGY

General Features

- Etiology

- Most common cause hematogenous contamination
  - *Staphylococcus aureus* in 86%; *Streptococcus* in 9%
  - GU or GI tract, lungs, or cutaneous source
    - 36% of cases, 1 or more concomitant infectious processes due to same microorganism: Arthritis, skin & soft tissue infections, endocarditis, & urinary tract infections
  - Predisposing factors
    - Intravenous drug abuse
    - Immunocompromised state
    - Diabetes, cirrhosis, renal failure, cancer, other chronic medical illnesses
- Direct inoculation from penetrating trauma, surgical intervention, or diagnostic procedures
  - Facet joint injection
- Extension from adjacent infection in paraspinal soft tissues
  - Diverticulitis, appendicitis, inflammatory bowl disease
  - Renal abscess

- Associated abnormalities
  - Spondylodiscitis
  - Epidural abscess (25%)
    - Cord compression
    - Foraminal narrowing
  - Paraspinal abscess
    - Presence of paraspinal ± epidural extension does not indicate worse prognosis
  - Spinal meningitis

Gross Pathologic & Surgical Features

- Necrotic bone
- Suppurative soft tissue

Microscopic Features

- Bone/synovial fragments
- Leukocytes, microorganisms, cellular debris
- Granulation tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute or chronic back pain
  - Focal tenderness
  - Fever (up 83%)
  - Other signs/symptoms
    - Neurological impairment in 38% of cases
    - Radiculopathy
    - Paraparesis
    - Sensory disturbance
    - Sphincter dysfunction
    - Elevated erythrocyte sedimentation rate, C-reactive protein, white count
- Clinical profile
  - Acute back pain mimics disc herniation
  - Difficult to distinguish from spondylodiscitis clinically

Demographics

- Age
  - 6th-7th decade
- Gender
  - Slight male predominance
- Epidemiology
  - Uncommon entity
    - Literature is primarily case reports (~50)
    - 4-5% of infectious spondylitis in 1 series
      - Incidence to as much as 20% of spontaneous spinal infections

Natural History & Prognosis

- Osseous destruction
- Spinal instability
- Progressive neurologic deterioration
- Sepsis and death
  - Mortality rate (2%)
- Favorable outcome with intravenous antibiotics alone (71%)
- Success rate improved if combined with percutaneous drainage (85%)

Treatment

- Intravenous antibiotics
- Percutaneous drainage
- Decompressive laminectomy indicated when epidural abscess and neurological deficits present

DIAGNOSTIC CHECKLIST

Consider

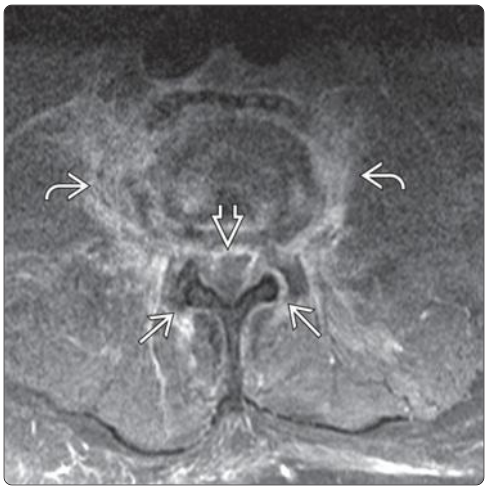
- Image-guided facet joint aspiration if blood culture negative
- High sensitivity of scintigraphy can help rule out other remote active septic foci

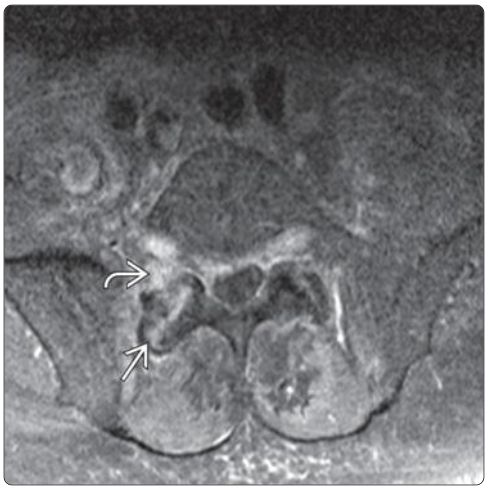
Image Interpretation Pearls

- Enhancing facet joint with juxtaarticular, paraspinal, or epidural phlegmon or abscess characteristic of septic facet joint arthritis
- Delay in MR findings with respect to clinical resolution
  - Consider scintigraphy with marked leukocytes as post-treatment control, confirms disappearance of inflammatory activity at that level

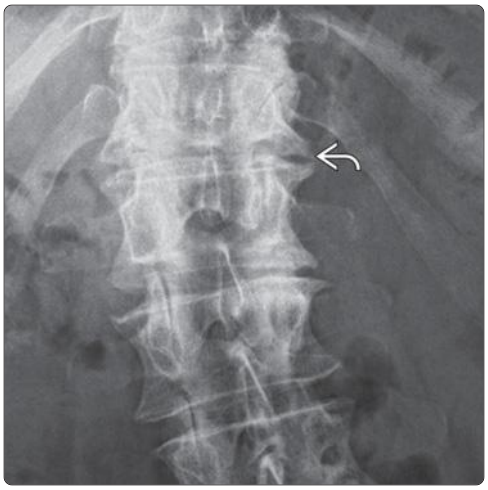
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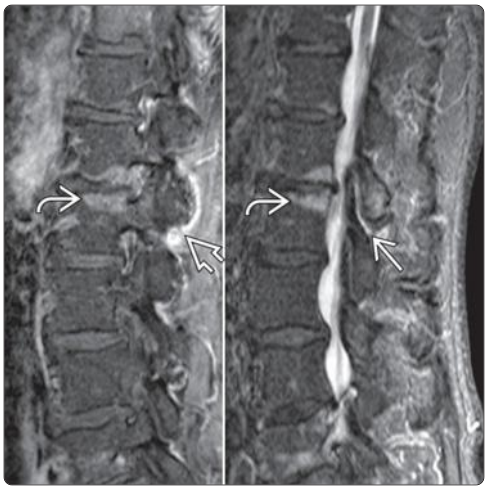
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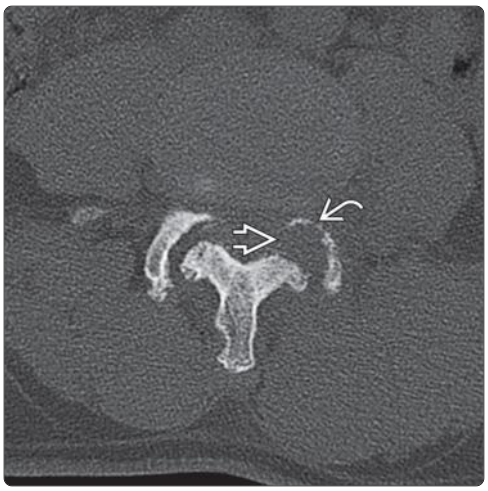
(Left) *Axial T1WI C+ FS MR depicts paraspinal enhancing phlegmon. There is epidural phlegmon circumferentially narrowing the thecal sac. There is bilateral septic facet arthritis with associated discitis-osteomyelitis. The hematogenous source was from a lower extremity cellulitis.* **(Right)** *Axial T1WI C+ FS MR displays right L5-S1 septic facet arthritis. Peri facet inflammation extends into the right neural foramen. Ventral epidural phlegmon effaces the right S1 lateral recess.*





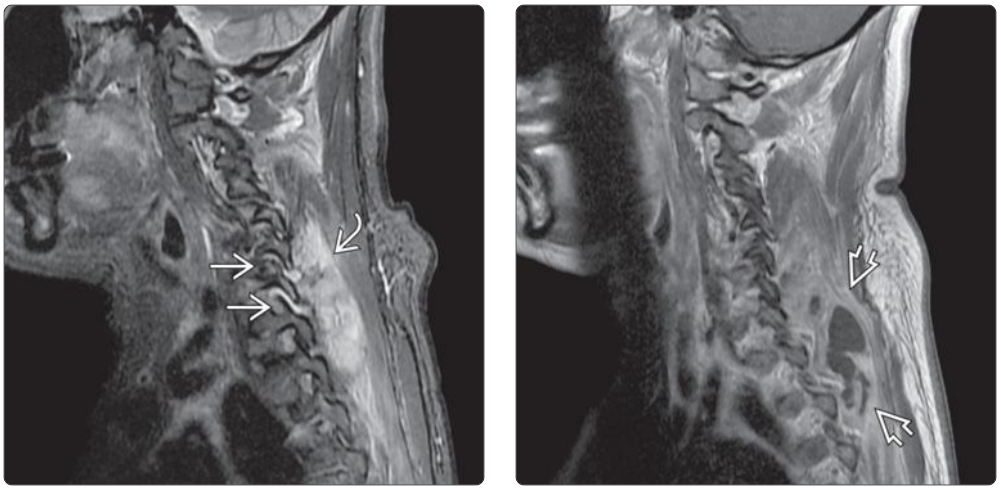
(Left) *Anteroposterior radiograph exhibits focal dextroscoliosis at the thoracolumbar junction. There is loss of disc height and endplate spurring along concavity.* **(Right)** *Sagittal T1WI C+ FS and STIR MR in the same patient illustrate minimal fluid in the left L1-L2 facet joint and enhancing peri facet soft tissue. On the ipsilateral side, there are endplate discogenic changes due to stress reaction (not infection) from the scoliosis.*



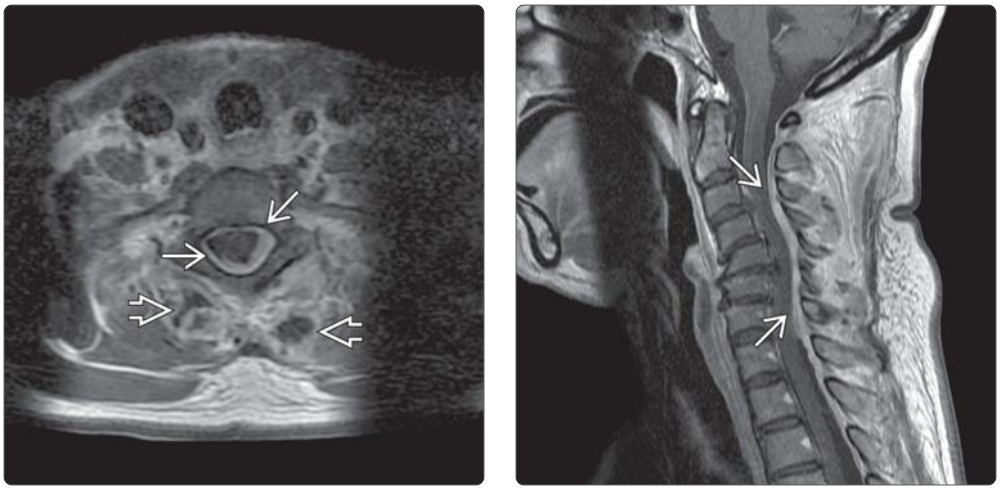


(Left) *Sagittal CT reconstruction shows widening of the left L4-L5 facet joint with erosive changes. Inflammatory spondyloarthopathies and metabolic disorders, such as gout, affect the facet joints. There may be spondylolisthesis due to bony erosion of the pars interarticularis and facet joint.* **(Right)** *Axial CT demonstrates widening of the left facet joint. There is smooth erosion and thinning of the cortex. On MR, the marrow of adjacent vertebral bodies is normal.*

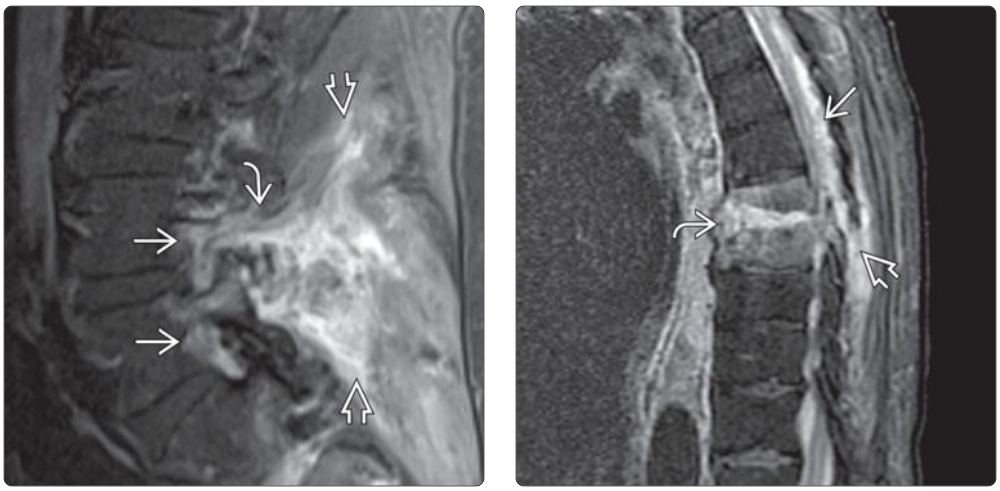
(Left) *Sagittal STIR MR in this patient with epidural abscess and facet involvement shows extensive posterior soft tissue edema and abnormal fluid in the C6-7 and C7-T1 joints.* **(Right)** *Sagittal T1 C+ MR in this patient with soft tissue infection (staph aureus) with posterior epidural phlegmon shows soft tissue abscesses and enhancing phlegmon extending to the posterior elements.*



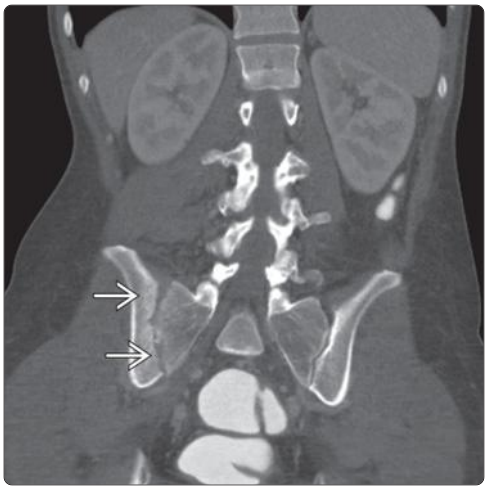
(Left) *Axial post-contrast T1WI MR shows the large, posterior diffusely enhancing epidural phlegmon and soft tissue abscesses.* **(Right)** *Sagittal T1 C+ in this patient with multicompartment infection shows the posterior epidural phlegmon with diffuse dural thickening and enhancement.*



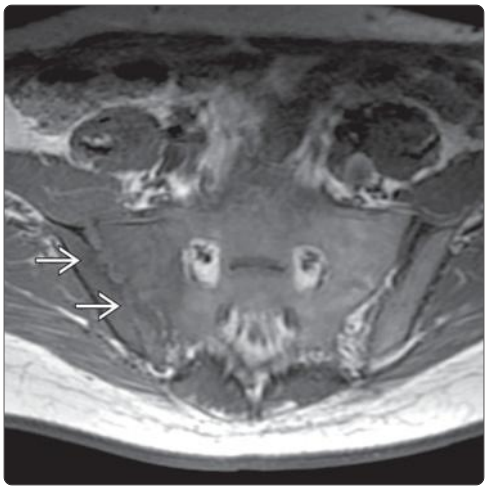
(Left) *Sagittal T1 C+ FS MR shows diffuse irregular enhancement of the dorsal soft tissues extending to involve the posterior elements and neural foramen in this patient with multicompartment infection. There is destruction of the facet joint.* **(Right)** *Sagittal STIR MR shows the typical findings of disc space infection with loss of disc margin, T2 hyperintense intervertebral disc, and hyperintense vertebral bodies. There is extension of abnormal signal into the facets and dorsal epidural abscess.*

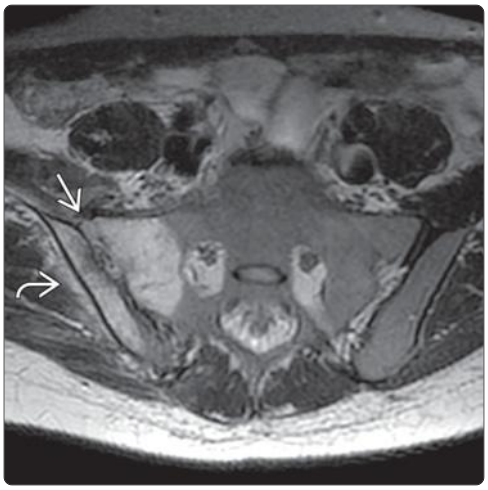




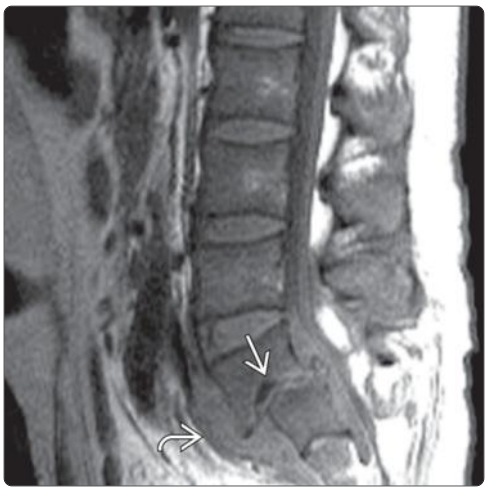


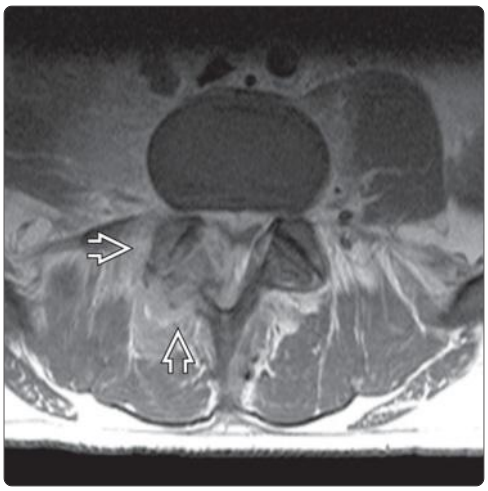
(Left) *Initial axial CECT shows a large right iliopsoas multiloculated abscess with mild narrowing of the right SI joint.* **(Right)** *Follow-up CECT study after catheter drainage of the right psoas abscess shows sclerosis and bony irregularity of the right SI joint margins, reflecting SI joint septic arthritis and adjacent osteomyelitis.*





(Left) *Axial T1W MR shows low T1 signal from the right sacral ala and iliac wing with diffuse irregularity of the infected SI joint, contrasted to the normal-appearing left joint.* **(Right)** *Axial T2W MR shows abnormal increased signal from the right SI joint, and adjacent iliac wing and sacral ala in this patient with right SI joint infection. Note the adjacent muscle edema.*





(Left) *Sagittal T1W MR in this patient with multicompartment infection shows typical disc space infection at L5-S1 with abnormal disc morphology, endplate irregularity, and presacral phlegmon.* **(Right)** *Axial T1 C+ MR at L4-5 shows extension of the infection to the right facet joint with diffuse facet bone enhancement and juxtafacet soft tissue involvement.*

Paraspinal Abscess

KEY FACTS

TERMINOLOGY

- Paraspinal phlegmon surrounding peripherally enhancing fluid collections

IMAGING

- **Pre**vertebral/**para**vertebral space
- Multiple or multiloculated collections along muscle plane
- NECT
  - Amorphous low-density intramuscular collection
  - Calcified psoas abscesses characteristic of tuberculous paraspinal abscess
  - Endplate destruction
- MR
  - Iso- to hypointense on T1WI
  - Hyperintense fluid collection and surrounding muscle on T2WI and STIR
  - Diffuse enhancement: Phlegmon
  - Peripherally enhancing fluid collection(s): Abscess

TOP DIFFERENTIAL DIAGNOSES

- Neoplasm, primary or metastatic
- Retroperitoneal hematoma
- Extramedullary hematopoiesis

PATHOLOGY

- Most common pathogens
  - *Staphylococcus aureus*
  - *Mycobacterium tuberculosis*
  - *Escherichia coli*

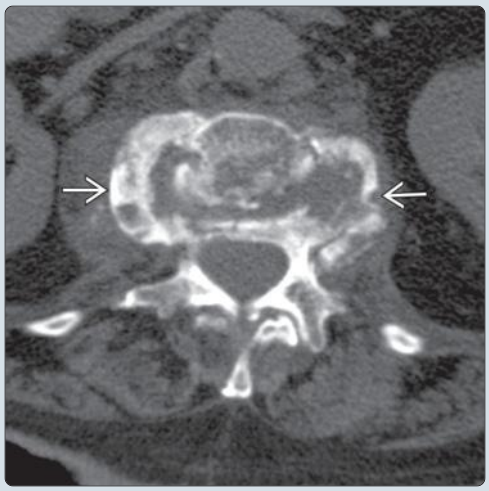
CLINICAL ISSUES

- Fever (50%) at presentation
- Back pain and tenderness
- ↑ ESR, ↑ WBC

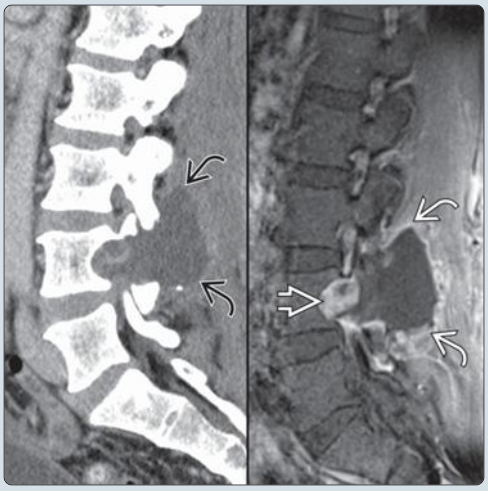
DIAGNOSTIC CHECKLIST

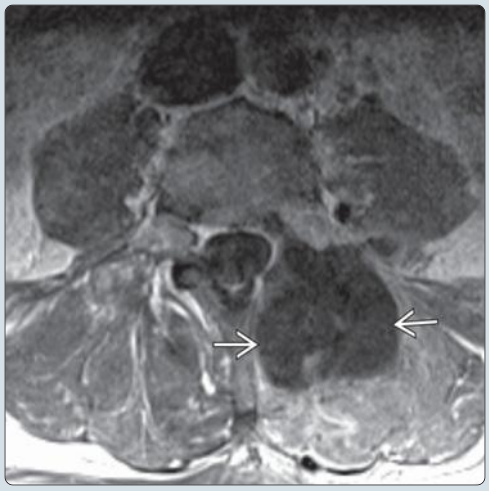
- Peripherally enhancing collection in paravertebral soft tissue with associated spondylitis characteristic of PA





(Left) *Axial graphic through a lumbar disc space demonstrates an extensive abscess infiltrating bilateral psoas muscles and epidural space. Abnormal retroperitoneal lymph nodes are also present.* **(Right)** *Axial NECT depicts bilateral calcified paraspinal masses in a patient with tuberculosis spondylodiscitis. Anterior aspect of the vertebral body adjacent to the subchondral plate is affected with spread to adjacent discs. Abscesses may descend down the sheath of the psoas and are typically calcified.*





(Left) *Sagittal CT (left) illustrates a low-density collection in the operative bed status post schwannoma resection. The collection has faint peripheral enhancement. Sagittal T1WI C+ MR (right) in the same patient better delineates irregular peripheral enhancement of the postoperative collection and enhancement in the neural foramen. The patient presented with fever and leukocytosis.* **(Right)** *Axial T1WI C+ MR shows a peripherally enhancing collection in the left neural foramen operative site.*

TERMINOLOGY

Abbreviations

- Paraspinal abscess (PA)

Definitions

- Infection of soft tissues surrounding spine

IMAGING

General Features

- Best diagnostic clue
  - Paravertebral enhancing phlegmon or peripherally enhancing liquefied collection
- Location
  - Prevertebral space
  - Paravertebral soft tissue
    - Psoas
    - Iliacus
    - Posterior paraspinous muscles (erector spinae)
- Size
  - Variable
    - Multiple or multiloculated
  - Craniocaudal extension
    - Subligamentous
    - Along muscle plane
- Morphology
  - Ill-defined infiltrative paraspinal soft tissue
  - Focal or diffuse muscle enlargement
    - Intramuscular fluid collection
    - Thick or thin, smooth or irregular wall
  - Obliterated soft tissue fascial planes

Radiographic Findings

- Radiography
  - Paraspinal soft tissue density
  - Enlarged psoas shadow
  - Osseous abnormalities after 3-4 weeks
    - Endplate irregularity and osteolysis
    - Disc space narrowing
    - Vertebral collapse

CT Findings

- NECT
  - Amorphous soft tissue density
  - Low-density intramuscular collection
    - ± intraabscess gas
  - Calcified psoas abscesses characteristic of tuberculous paraspinal abscess
- CECT
  - Diffuse or peripheral enhancement
  - Enhancing disc space
- Bone CT
  - Endplate destruction
  - Bony sequestra
  - Spinal deformity

MR Findings

- T1WI
  - Iso- to hypointense
    - May be difficult to distinguish from normal musculature
    - Obscuration of musculofascial planes
- T2WI
  - Hyperintense fluid collection and surrounding muscle
  - Hypointense rim
- PD/intermediate
  - Hyperintense fluid collection and surrounding muscle
- STIR
  - Hyperintense fluid collection and surrounding muscle
- T2* GRE
  - Iso- to hyperintense
  - Hypointense rim
- DWI
  - Can increase conspicuity of epidural, vertebral, and paraspinal abscesses
- T1WI C+
  - Phlegmon: Diffuse enhancement
  - Abscess: Peripherally enhancing fluid collection(s)
- Features of spondylitis
  - Hyperintense disc on T2WI
  - Endplate erosion
  - Marrow edema
  - Enhancing disc and marrow
- Intraspinal epidural extension with cord compression
  - Involvement of lateral epidural space with neural foraminal narrowing

Ultrasonographic Findings

- Grayscale ultrasound
  - Anechoic or hypoechoic intramuscular collection
- Power Doppler
  - Hyperemia and increased vascularity surrounding abscess

Nuclear Medicine Findings

- Gallium scan
  - Increased radionuclide uptake
- WBC scan
  - Increased uptake on In-111 WBC scan

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI C+ and T2WI MR

DIFFERENTIAL DIAGNOSIS

Neoplasm, Primary or Metastatic

- Discrete or infiltrative soft tissue mass
  - Isointense to muscle on T1WI
  - Hyperintense on T2WI
  - Post-gadolinium enhancement
    - Necrotic component mimics abscess
- Variable vertebral involvement
  - Intervertebral discs typically spared

Retroperitoneal Hematoma

- Infiltrative soft tissue
  - Hyperdense on CT
  - Iso- to hypointense on T1WI and T2WI in acute stage
  - Hyperintense on T1WI and T2WI in later stage

- ± mild enhancement
- Diffuse muscular enlargement
  - Fluid-fluid level present if anticoagulated

Extramedullary Hematopoiesis

- Paravertebral mass along thoracic and lumbar spine
  - Homogeneous and well defined
  - Iso- to hypointense on T1WI
  - Iso- to hyperintense on T2WI
  - ± mild enhancement
- Diffuse vertebral marrow hypointensity
  - Relatively hyperintense intervertebral discs spared

PATHOLOGY

General Features

- Etiology
  - Most common pathogens
    - *Staphylococcus aureus*
    - *Mycobacterium tuberculosis*
    - *Escherichia coli*
  - Fungal infection rare
    - More common in immunocompromised host
  - Predisposing factors
    - Intravenous drug abuse
    - Immunocompromised state
    - Diabetes mellitus, alcoholism, cirrhosis, chronic renal failure, and other chronic medical illnesses
  - Direct extension from adjacent infection
    - Spondylodiscitis
    - Septic facet arthritis
    - Appendicitis
    - Diverticulitis
    - Inflammatory bowel disease
    - Perinephric abscess
  - Transcutaneous infection of deep tissue
    - Trauma
    - Epidural injection or catheter placement
    - Facet joint injection
    - Spine or GI surgery
  - Hematogenous spread from distant sites
    - Pyomyositis
      - Commonly caused by gram-positive organisms
  - Primary PA uncommon
- Associated abnormalities
  - Epidural abscess

Gross Pathologic & Surgical Features

- Necrotic soft tissue with thick green-brown fluid

Microscopic Features

- Leukocytes, microorganisms, cellular debris
- Granulation tissue, increase vascularity

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever (50%) at presentation
  - Back pain and tenderness
  - ↑ ESR, ↑ WBC
  - Other signs/symptoms
    - Lower extremity pain
    - Paraspinal muscle spasm
  - If epidural component present
    - Weakness, paresthesia, sphincter dysfunction
- Clinical profile
  - Diagnosis may be delayed due to chronic and insidious symptomatology

Demographics

- Age
  - Bimodal pattern
    - Adults (6th to 7th decades)
    - Children, especially 10-19 years
- Gender
  - M:F = 3:1
- Epidemiology
  - > 90% of tuberculous spondylitis cases associated with intra- or paraspinal abscess

Natural History & Prognosis

- Depends on host immune response
  - May be contained with early treatment
  - Overwhelming sepsis leading to death in debilitated host
- Progressive neurological impairment if spondylitis ± epidural abscess present
- Dependent on
  - Comorbidities
  - Extent of spinal involvement
  - Degree of neurologic compromise

Treatment

- Long-term intravenous antibiotics
- Analgesic medications
- Percutaneous catheter drainage
- Surgical debridement
  - Spinal stabilization may be required if instability present

DIAGNOSTIC CHECKLIST

Consider

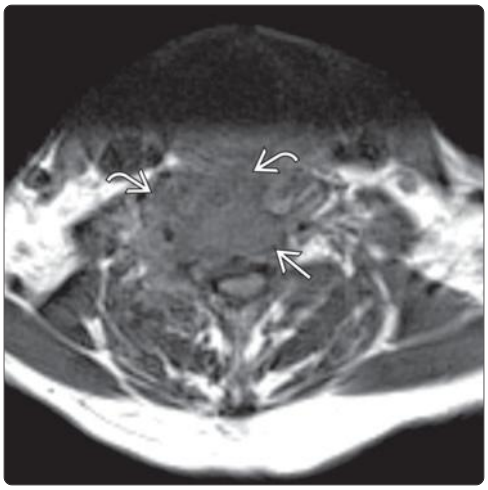
- Evaluate adjacent vertebral bodies and intervertebral disc spaces for spondylodiscitis
- Assess epidural soft tissues, particularly in setting of neurologic deficits

Image Interpretation Pearls

- Peripherally enhancing collection in paravertebral soft tissue with associated spondylitis characteristic of PA

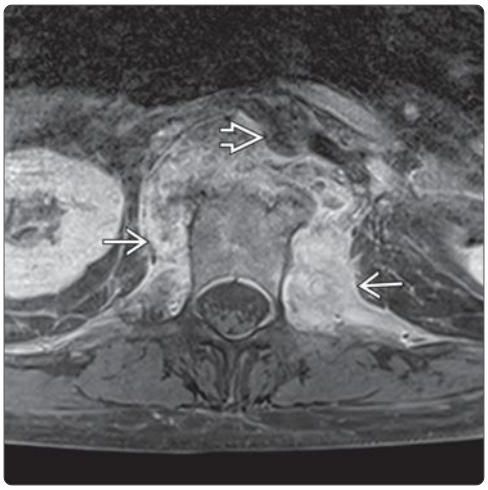
SELECTED REFERENCES

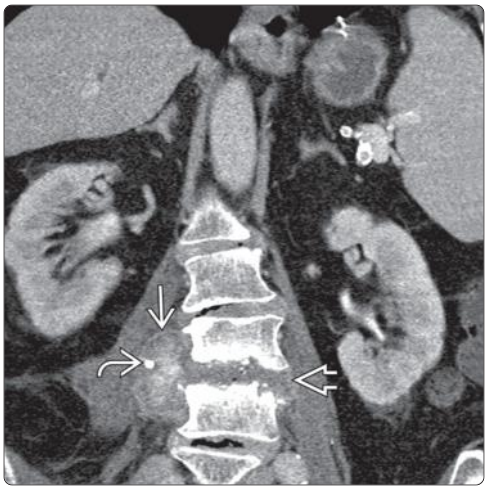
1. Siddiq DM et al: Spinal and paraspinal pneumococcal infections-a review. Eur J Clin Microbiol Infect Dis. 33(4):517-27, 2014
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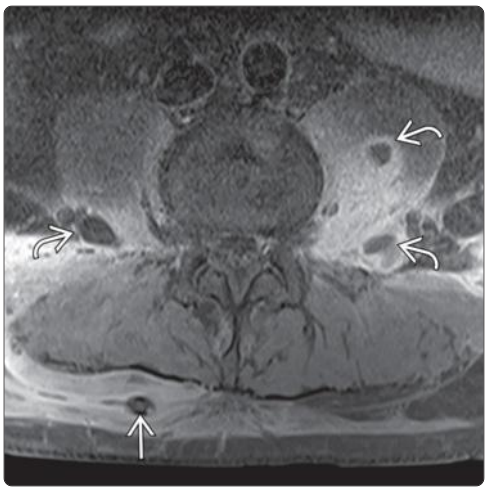
(Left) *Axial T1WI MR depicts amorphous low signal of the cervical vertebral body and ventral longus colli musculature. The paraspinal phlegmon is an extension of discitis/osteomyelitis.* **(Right)** *Axial T2WI MR demonstrates a heterogeneous collection in the right psoas muscle. It has a thick, irregular hypointense rim and central hyperintensity. The right psoas shows diffuse hyperintensity due to edema.*





(Left) *Axial T1WI C+ FS MR exhibits an avidly heterogeneously enhancing paraspinal collection, which abuts the anterior and lateral margins of the lumbar vertebral body. The enhancing material comes in close proximity to the aorta.* **(Right)** *Coronal NECT illustrates a right paraspinal abscess related to lumbar discitis/osteomyelitis. A drainage catheter is seen within the collection. Internal hyperdensity may be a combination of blood products and purulent material.*





(Left) *Coronal STIR MR shows multiple heterogeneously hyperintense collections in the edematous left psoas muscle. These paraspinal abscesses were likely hematogenous in etiology given the history of IV drug abuse in this patient. STIR improves detection of early paraspinal inflammation.* **(Right)** *Axial T1WI C+ FS MR displays multiple small, peripherally enhancing, hypointense collections in bilateral psoas muscles. A posterior subcutaneous drain was placed for superficial abscesses.*

Epidural Abscess

KEY FACTS

TERMINOLOGY

- Spinal epidural abscess (SEA)
- Extradural spinal infection with abscess formation

IMAGING

- Lower thoracic and lumbar > upper thoracic and cervical
- CT
  - Enhancing epidural mass narrowing central canal
- MR
  - T1WI: Iso- to hypointense to cord
  - T2WI/STIR: Hyperintense
  - T1WI C+: Homogeneously or heterogeneously enhancing **phlegmon**
  - Peripherally enhancing necrotic **abscess**
- Fat saturation: STIR, T2WI FS, T1WI C+ FS
  - Increases lesion conspicuity by suppressing signal from epidural fat and vertebral marrow
- Signal alteration in spinal cord secondary to compression, ischemia, or direct infection
- Persistent epidural enhancement without mass effect on follow-up MR imaging
  - Probable sterile granulation tissue or fibrosis
  - Correlate with ESR and CRP for disease activity

TOP DIFFERENTIAL DIAGNOSES

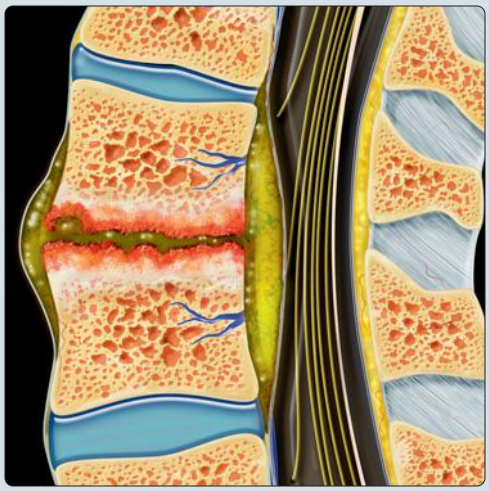
- Extradural metastasis
  - Often contiguous with vertebral lesion
- Epidural hematoma
  - ± mild peripheral enhancement

PATHOLOGY

- *Staphylococcus aureus* most common cause; *Mycobacterium tuberculosis* next most frequent

CLINICAL ISSUES

- Fever, acute or subacute spinal pain and tenderness





(Left) *Sagittal graphic through the lumbar spine demonstrates vertebral osteomyelitis with an intervertebral abscess extending ventrally and dorsally and narrowing the central canal.* **(Right)** *Sagittal T1WI C+ MR in an intravenous drug abuser shows a large ventral epidural abscess collection ⇨ with peripheral enhancement causing a severe mass effect upon the cervical cord. There is relative preservation of the C5-C6 endplates ⇨ with no disc enhancement.*





(Left) *Sagittal T1WI MR without contrast shows low signal from the C5 and C6 vertebral bodies and an intermediate signal ventral epidural mass causing cord compression in a patient with cervical abscess.* **(Right)** *Sagittal STIR MR shows a large ventral epidural abscess ⇨ as T2 hyperintensity with severe mass effect upon the cord. Involvement of the posterior C5-C6 disc is seen as linear hyperintensity ⇨. There is cord hyperintensity in this patient, who was quadriparetic.*

TERMINOLOGY

Abbreviations

- Spinal epidural abscess (SEA)

Synonyms

- Spinal epidural empyema

Definitions

- Extradural spinal infection with abscess formation

IMAGING

General Features

- Best diagnostic clue
  - Findings of spondylodiscitis with adjacent enhancing epidural phlegmon ± peripherally enhancing fluid collection
- Location
  - Posterior (80%) or anterior (20%) epidural space; circumferential (caudal to S2)
  - Lower thoracic and lumbar > upper thoracic and cervical
- Size
  - May extend over many segments
- Morphology
  - Focal or diffuse elongated epidural soft tissue

Radiographic Findings

- Radiography
  - Not directly visualized
  - Associated findings of spondylodiscitis: Endplate irregularity/destruction; disc height loss

CT Findings

- CECT
  - Enhancing epidural mass narrowing central canal
- Bone CT
  - Increased epidural soft tissue
  - May be difficult to distinguish from disc

MR Findings

- T1WI
  - Iso- to hypointense to cord
- T2WI
  - Hyperintense
- STIR
  - Hyperintense
- T2* GRE
  - Iso- to hyperintense
- DWI
  - Hyperintense, ADC hypointense
- T1WI C+
  - Homogeneously or heterogeneously enhancing **phlegmon**
  - Peripherally enhancing necrotic **abscess**
  - Diffuse dural enhancement in extensive SEA
  - Enhancing prominent anterior epidural veins or basivertebral venous plexus above/below abscess
- Various degrees of encroachment on central canal and neural foramina
- Signal alteration in spinal cord secondary to compression, ischemia, or direct infection
- Persistent epidural enhancement without mass effect on follow-up MR imaging
  - Probable sterile granulation tissue or fibrosis
  - Correlate with ESR and CRP for disease activity

Nonvascular Interventions

- Myelography
  - Epidural mass impeding cerebral spinal fluid flow

Nuclear Medicine Findings

- Gallium scan
  - Increased uptake in spinal or epidural area

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI and T1WI MR with gadolinium
- Protocol advice
  - Fat saturation: STIR, T2WI FS, T1WI C+ FS
    - Increases lesion conspicuity by suppressing signal from epidural fat and vertebral marrow

DIFFERENTIAL DIAGNOSIS

Extradural Metastasis

- Well-circumscribed, extraosseous soft tissue mass
  - Hypointense on T1WI, hyperintense on T2WI
  - Diffuse enhancement
  - Often contiguous with vertebral lesion
    - Destruction of posterior vertebra/pedicle
  - Intervertebral discs unaffected
- Pathologic compression fractures
- Sparing of spinal column in some cases
  - Spinal epidural lymphoma

Epidural Hematoma

- Heterogeneously hyperintense on T2WI
- Acute hemorrhage isointense on T1WI
  - Hyperintense subacute and chronic hemorrhage
- ± mild peripheral enhancement
- Intact vertebrae in absence of trauma

Extruded/Migrated Disc

- Associated parent disc height loss, protrusion, degeneration
  - Iso- to hypointense on T2WI
- More focal appearance
- ± mild peripheral enhancement
- Vertebral endplates intact

Epidural Lipomatosis

- Excessive epidural fat in thoracic and lumbar canal
- Homogeneous and hyperintense on T1WI and T2WI
- Signal loss with fat suppression on STIR
- Mass effect on spinal cord and nerve roots

PATHOLOGY

General Features

- Etiology
  - Up to 1/3 may not have identifiable source
    - *Staphylococcus aureus* is most common pathogen (57-73% of reported cases)

- *Mycobacterium tuberculosis* is next most frequent cause (25% of reported cases)
- Fungal infection is less common cause
- Predisposing factors
  - Intravenous drug abuse
  - Immunocompromised state
  - Diabetes mellitus, chronic renal failure, alcoholism, cancer, other chronic illnesses
- Anterior SEA arises from adjacent discitis and vertebral osteomyelitis
- Posterior SEA arises from GU or GI tract, lungs, cardiac, and mucous/cutaneous sources through hematogenous dissemination
- Direct inoculation from penetrating trauma, surgical intervention, or diagnostic procedures
  - Risk of SEA in epidural anesthesia (5.5%)
    - Especially in presence of indwelling catheters
  - Extradural hematoma may become infected in blunt trauma
- Extension from adjacent infection in paraspinal soft tissues
  - Diverticulitis, appendicitis, pyelonephritis
- Cord symptoms are likely from combination of compressive and ischemic effects
  - Cord ischemia from compromised epidural venous plexuses
- Epidural space
  - Dura is adherent to bone above foramen magnum → subaxially true epidural space is posterior and lateral to spinal cord
    - Epidural space is very small in cervical region, larger in lumbosacral region
    - Anteriorly, dura is loosely adherent to vertebral bodies from foramen magnum to L1 → potential epidural space
    - Therefore, majority of SEAs are located posteriorly; anterior SEAs are usually below L1

- Associated abnormalities
  - Discitis, osteomyelitis, paraspinal abscess, septic facet arthritis
- Suppuration of epidural space from adjacent infection or bacteremia

Microscopic Features

- Granulation tissue, leukocytes, cellular debris

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, acute or subacute spinal pain and tenderness
- Other signs/symptoms
  - Radiculopathy, paraparesis/paralysis, paresthesia, loss of bladder and bowel control
- Clinical profile
  - In patients with septicemia or chronic illness, neurologic symptoms may be obscured by systemic complaints
  - Abscess from hematogenous spread progresses rapidly
  - Abscess from osteomyelitis/discitis may smolder

Demographics

- Age
  - All ages reported
  - Peak incidence in 6th and 7th decades
- Gender
  - M:F = 1:0.56
- Epidemiology
  - 0.2-2.8 cases per 10,000

Natural History & Prognosis

- Irreversible neurologic deficit and death if untreated or if treatment is delayed
- Factors influencing prognosis (mortality rate of 12-30%)
  - Age: Better prognosis in children than adults
  - Severity of initial neurologic deficits
    - Thecal compression > 50% is associated with poor prognosis
  - Location of SEA: Poor outcome with SEA at higher levels
  - Duration between onset of neurologic deficits and surgical intervention
    - Early diagnosis and institution of treatment improve prognosis
  - Comorbidities such as diabetes

Treatment

- Some studies advocate emergent surgical decompression with abscess drainage, even in absence of initial neurologic compromise
  - Rapid progression of neurologic deficits can occur despite appropriate medical therapy
- Others report no increased risk of poor outcome if not treated with early surgery
  - Anatomy of SEA (ventral or dorsal) should play role in determining treatment plan
    - Dorsal > ventral: Paraplegia or quadriplegia
- Early empiric antibiotics with broad-spectrum coverage until causative pathogen is isolated
- Organism-specific intravenous antibiotics followed by long-term parenteral antibiotics for 6-8 weeks

DIAGNOSTIC CHECKLIST

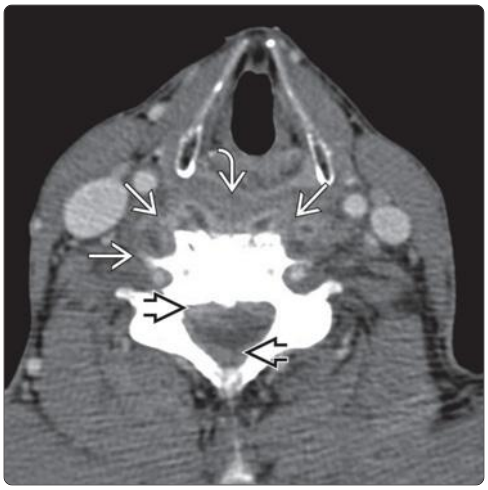
Image Interpretation Pearls

- Epidural soft tissue with diffuse or peripheral enhancement and adjacent spondylodiscitis is characteristic

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(Left) *Sagittal T1WI C+ MR illustrates ventral epidural phlegmon from C5 to C7 with a focal hypointense abscess at C6-C7. Note the extensive prevertebral phlegmon.* **(Right)** *Axial CECT demonstrates an inflammatory process in the perivertebral space due to osteomyelitis/discitis. Heterogeneously enhancing enlarged prevertebral muscles, a small amount of retropharyngeal fluid, and extradural epidural phlegmon compressing the cervical cord are identified.*





(Left) *Sagittal T1WI C+ MR shows osteomyelitis/discitis at L3-L4 with enhancing irregular endplates and peripherally enhancing ventral and dorsal SEA. Involvement of the subarachnoid space is defined with thick, avid enhancement of the cauda equina nerve roots.* **(Right)** *Sagittal T1WI C+ FS MR shows homogeneously enhancing epidural phlegmon in the ventral epidural space adjacent to the osteomyelitis/discitis. Dorsal epidural phlegmon contributes to the mass effect on the cord.*





(Left) *Sagittal T1WI C+ MR depicts peripheral enhancement around a central nonenhancing abscess in the ventral epidural space. The abscess results in significant compression of the thecal sac.* **(Right)** *Sagittal T1WI C+ MR exhibits a complex, peripherally enhancing dorsal epidural collection with enhancing internal septations at the cervicothoracic junction. Extensive dorsal spinal cord compression explains the patient's myelopathy. Plegia is associated with dorsal SEA > ventral SEA.*

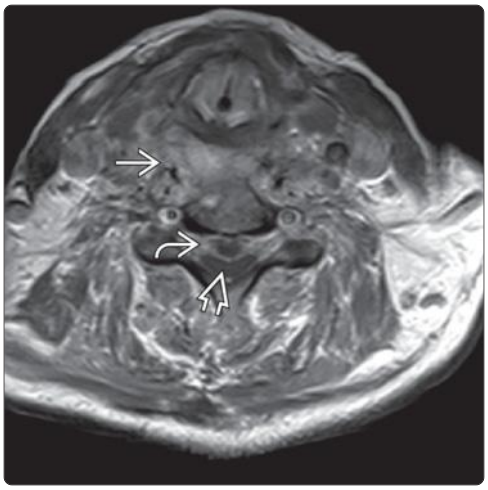
(Left) *Sagittal T1 MR shows loss of signal at C4 and C5 with indistinct anterior vertebral body margins. There is also loss of the ventral margin of the cord at the C4-C5 level due to epidural abscess.* **(Right)** *Sagittal STIR MR shows slight hyperintensity from the C4 and C5 bodies and from the ventral intervertebral disc. There is high signal from the prevertebral soft tissues. There is extensive hyperintensity along the ventral margin of the cord from C3-C4 to C5-C6 due to abscess.*





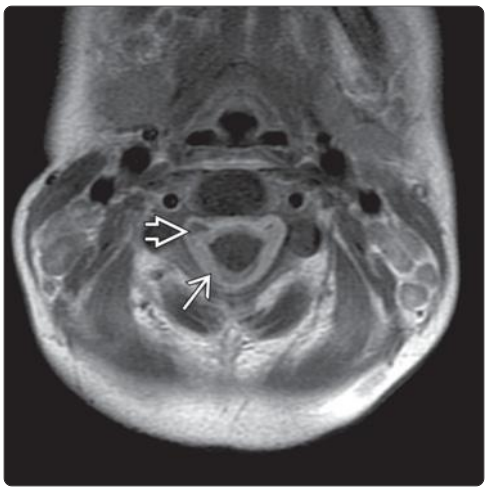
(Left) *Sagittal T1 C+ MR shows extensive enhancement of the prevertebral phlegmon and peripheral enhancement of the well-defined ventral epidural abscess. Note the mass effect upon the cervical cord.* **(Right)** *Axial T1 C+ MR defines the enhancing prevertebral phlegmon, peripheral enhancing ventral epidural abscess, and severe degree of cord compression.*





(Left) *Sagittal T1 C+ MR shows abnormal circumferential epidural thickening and enhancement in this patient with meningitis complicated by epidural phlegmon and abscess.* **(Right)** *Axial T1 C+ MR shows extensive epidural thickening due to phlegmon as well as small pockets of fluid signal epidural abscess.*

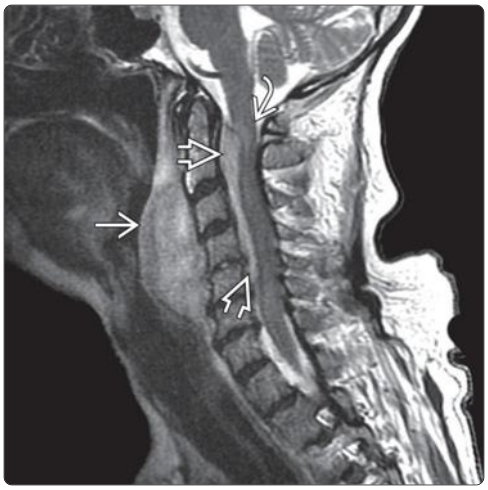








(Left) *Sagittal CECT performed for evaluation of neck pain shows a large prevertebral abscess and a large ventral epidural abscess with cord compression.* **(Right)** *Axial CECT shows a large prevertebral abscess with foci of gas. There is extension of the abscess into the ventral epidural space with posterior displacement of the cervical cord.*





(Left) *Sagittal T2WI MR shows a large prevertebral abscess as increased signal. Note the extensive ventral epidural abscess with posterior displacement of the cord. A small amount of cord edema is present.* **(Right)** *Sagittal T1 C+ MR shows peripheral enhancement of both the large ventral paravertebral abscess and epidural involvement with cord compression.*





(Left) *Sagittal T1 C+ MR after laminectomy shows a large infected fluid collection extending from the laminectomy site. Additional focal fluid is present in the posterior epidural space with mass effect on the cauda equina (which is enhancing due to meningitis).* **(Right)** *Axial T1 C+ MR shows a large fluid collection extending from the operative site with severe mass effect on the cauda equina. The patient was clinically infected.*

Subdural Abscess

KEY FACTS

TERMINOLOGY

- Spinal subdural abscess (SSA)
- Intradural extramedullary ring-enhancing fluid collection of pus in "potential" space between dura and arachnoid

IMAGING

- Thoracolumbar region most common
- Presence suggested by mass effect on cord and obliterated subarachnoid space
- Cord displacement: ± impingement and edema
- Increased subdural density on NECT
- MR
  - T2 hyperintense/T1 hypointense
  - Heterogeneous, diffuse subdural enhancement
  - Rim-enhancing fluid collection
- Best imaging modality
  - MR with contrast → ↑ sensitivity in detecting SSA
  - Axial sequences confirm subdural location (inside dural margin)

TOP DIFFERENTIAL DIAGNOSES

- Abscess: Epidural, paravertebral
  - Typically associated with spondylodiscitis
- Subdural hematoma
  - Predominantly hypointense on T2WI or gradient-echo

PATHOLOGY

- Predisposing factors
  - Intravenous drug abuse, immunocompromised state, diabetes, cirrhosis, renal failure

CLINICAL ISSUES

- Fever
- Neck or back pain and tenderness

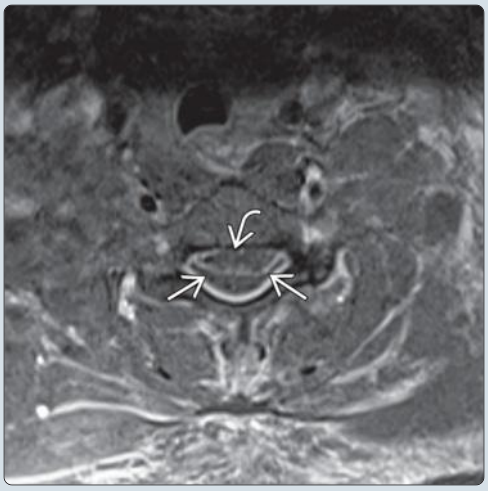
DIAGNOSTIC CHECKLIST

- Ring-enhancing intradural extramedullary collection outlined by epidural fat and CSF on axial imaging





(Left) *Sagittal T2WI MR shows a heterogeneously hyperintense collection in the dorsal intradural extramedullary space. It has a hypointense rim and causes marked mass effect upon the cervical cord.* **(Right)** *Sagittal CT demonstrates a hyperdense SSA in the dorsal spinal canal. Osseous destruction of the L4 and L5 endplates and intradiscal hypodense fluid at this level are observed. This extensive SSA began as a lumbar spinal epidural abscess caused by Serratia marcescens.*





(Left) *Axial T1WI C+ MR depicts a rim-enhancing fluid collection in the dorsal spinal canal, causing mass effect upon and anteriorly displacing the cervical cord.* **(Right)** *Sagittal T1 C+ MR exhibits a low intensity collection with thick peripheral enhancement spanning the cervical and thoracic dorsal spinal canal. The SSA causes severe impingement of the cord. Initially, CT-guided drainage was attempted but with limited success. Subsequently, surgical decompression was performed.*

TERMINOLOGY

Synonyms

- Spinal subdural empyema
- Spinal subdural abscess (SSA)

Definitions

- Collection of pus in "potential" space between dura and arachnoid

IMAGING

General Features

- Best diagnostic clue
  - Intradural extramedullary ring-enhancing fluid collection on axial imaging
- Location
  - Thoracolumbar region most common
- Size
  - Focal abscess
  - Multifocal phlegmon or abscesses extending over multiple spinal levels
- Morphology
  - Thin, elongated fluid collection
  - Diffuse subdural thickening

Radiographic Findings

- Radiography
  - Typically noncontributory unless spondylodiscitis also present

CT Findings

- NECT
  - Increased subdural density
  - ± intradural extramedullary gas
- CECT
  - Homogeneous subdural enhancement
  - Peripherally enhancing fluid collection

MR Findings

- T1WI
  - Intermediate signal intensity (SI)
  - Effaced CSF
- T2WI
  - Hyperintense
    - Presence suggested by mass effect on cord
  - Obliterated subarachnoid space
- PD/intermediate
  - Hyperintense
- STIR
  - Hyperintense
- T2* GRE
  - Hyperintense
- T1WI C+
  - Heterogeneous, diffuse subdural enhancement
  - Rim-enhancing fluid collection
  - Intraspinal contents from outer to inner on axial imaging
    - Epidural fat, enhancing dura and subdural abscess, CSF, cord
- Cord displacement
  - ± impingement
  - ± edema

Nonvascular Interventions

- Myelography
  - Block of CSF
  - Intradural irregularities
  - ± cord swelling

Nuclear Medicine Findings

- Gallium scan
  - Increased intraspinal uptake of gallium citrate (Ga-67)
    - Improved sensitivity with SPECT

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI and T2WI MR
- Protocol advice
  - Intravenous gadolinium increases sensitivity in detecting SSA
    - Axial sequences confirm subdural location

DIFFERENTIAL DIAGNOSIS

Epidural Abscess

- Lower thoracic and lumbar spine
- Typically associated with spondylodiscitis
- Intermediate SI on T1WI, hyperintense on T2WI
  - Diffuse or rim enhancement
- Variable mass effect on spinal cord

Subdural Hematoma

- Other than trauma, lumbar puncture in patients with coagulopathy is most common cause
- More common in lumbar or thoracolumbar region
- Predominantly hypointense on T2WI or gradient-echo
  - Isointense on T1WI
- ± mild peripheral enhancement

Spinal Meningitis

- Hematogenous dissemination of bacteria from extraspinal source
- Increased CSF intensity on T1WI
- Diffuse CSF enhancement
- Smooth or nodular leptomeningeal or nerve root enhancement
- ± cord edema

CSF Leakage Syndrome

- From prior spinal trauma, diagnostic or interventional procedure, or spontaneous
- Low opening pressure on lumbar puncture
- Diffuse smooth dural thickening and enhancement
  - Likely from dural venous engorgement
- May see cerebellar tonsillar descent and effaced prepontine space in posterior fossa

Idiopathic Hypertrophic Spinal Pachymeningitis

- Fibrous inflammatory thickening of dura mater
- Thickened dura iso- to hypointense on T1WI and T2WI
  - Marked post-contrast enhancement

PATHOLOGY

General Features

- Etiology
  - Predisposing factors
    - Intravenous drug abuse
    - Immunocompromised state
      - Meds, underlying infection, or systemic disease, such as malignant neoplasm
    - Obesity and alcoholism reported important predictive factors for spinal sepsis
    - Diabetes, cirrhosis, renal failure, or AIDS, recurrent UTI
  - Mechanism of inoculation
    - Hematogenous dissemination from extraspinal focus of infection
    - Mucous/cutaneous source most common
    - Contiguous spread from adjacent abscess
    - Direct inoculation through trauma or interventional procedures
      - Lumbar puncture, placement of catheters, cervical/lumbar discogram
    - Dermal sinus tract
      - Ends in intradural space in ~ 60%
    - Unexplained source of infection
  - Common pathogens
    - *Staphylococcus aureus* most common
    - Gram-negative bacilli
  - Cord symptoms likely from combination of compressive and ischemic effects
- Associated abnormalities
  - Spinal meningitis
  - Spinal epidural abscess
  - Usually **not** associated with vertebral osteomyelitis
- Suppuration of spinal subdural space
- Anatomy
  - Subdural space
    - "Potential" space between dura mater and arachnoid membrane

Gross Pathologic & Surgical Features

- Turbid CSF
- Frank pus

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever
  - Neck or back pain
  - Tenderness
  - Other signs/symptoms
    - Paraparesis, quadriparesis
    - Paresthesia, sensory level
    - Gait disturbance
    - Meningismus
    - Hyporeflexia
    - Bladder or bowl dysfunction
    - Papilledema 2° to ↑ CSF protein or disrupted CSF flow in thecal sac → ↑ intracranial pressure
- Other signs/symptoms
  - Staging system for abscesses
    - Stage 1: Fever ± spinal or nerve root pain
    - Stage 2: Mild neurological deficits are added to clinical picture
    - Stage 3: Paralysis and complete sensory loss occur below level of lesion
- Clinical profile
  - Triad of fever, neck or back pain, and cord compression
    - 38% of reported cases
  - Source of infections, such as cellulitis, furuncles, dental abscess may be present
  - Difficult to distinguish from spinal epidural abscess clinically

Demographics

- Age
  - All ages reported
    - Most frequently between 60-70 years
- Gender
  - M:F = 1:2
- Epidemiology
  - Rare; about 50 reported cases of SSA in the literature

Natural History & Prognosis

- If left untreated, progressive neurologic deterioration
  - Sepsis, death
- 20% survival rate if intravenous antibiotics only
- 82% survival rate if surgical treatment

Treatment

- Supportive care with hydration and pain management
- Intravenous dexamethasone to decrease inflammatory reaction and brain/cord edema
- Percutaneous image-guided catheter drainage
- Surgery
  - Laminectomy and durotomy for abscess drainage and irrigation
  - Recommended in all surgical candidates
- Empiric intravenous antibiotics with gram-positive cocci coverage until positive culture results

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Ring-enhancing intradural extramedullary collection outlined by epidural fat and CSF on axial imaging

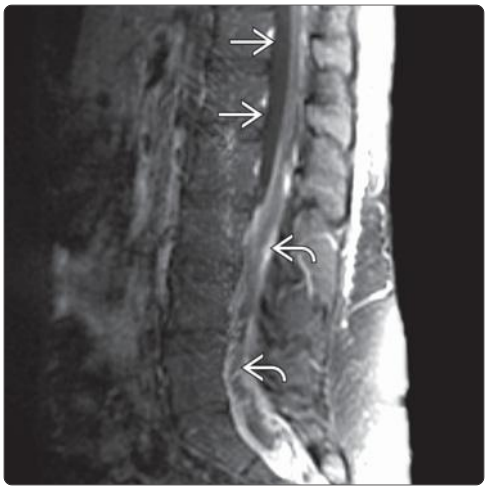
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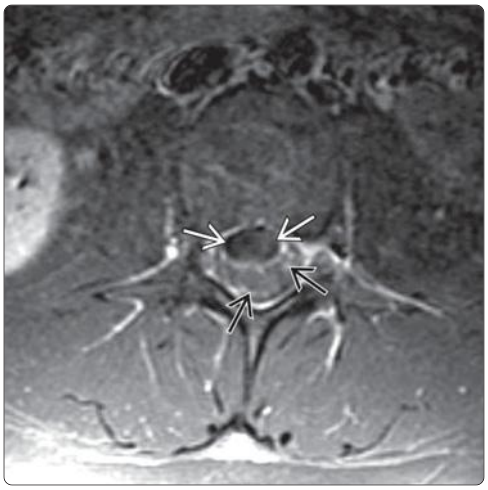
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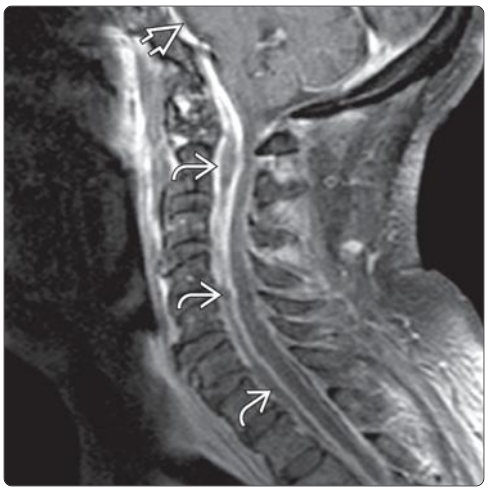


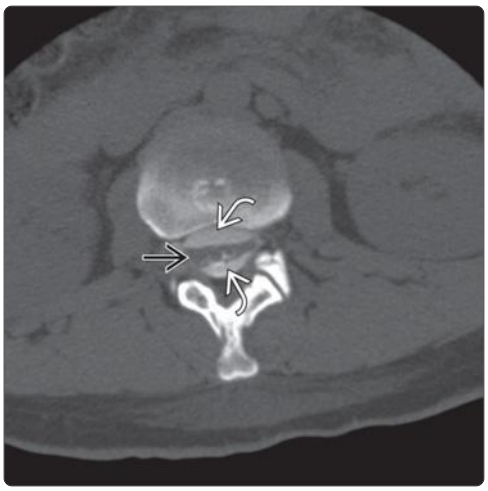
(Left) *Sagittal T1WI C+ MR exhibits low signal intensity, loculated collections along the course of the cauda equina and extending cephalad to the T12 level. There is associated enhancement of the cauda equina due to meningitis.* **(Right)** *Sagittal T2WI MR shows a subdural hygroma secondary to intracranial hypotension. This may mimic a subdural abscess; however, there will be no clinical signs of infection. Note the preservation of the dorsal epidural fat.*





(Left) *Sagittal T1WI C+ FS MR demonstrates a hypointense collection in the ventral intradural extramedullary space that displaces the cord posteriorly. This patient has Pseudomonas meningitis with clumping and intense enhancement of the cauda equina in distal thecal sac.* **(Right)** *Axial T1WI C+ FS MR reveals a peripherally enhancing subdural collection that posteriorly displaces the cord. There is linear enhancement along the pial surface of the cord and along the nerve roots.*





(Left) *Sagittal T1WI C+ MR with fat suppression through the cervical spine demonstrates diffuse ventral subdural abscess with intracranial extension. SSA is most commonly caused by Staphylococcus aureus and arises mostly from adjacent infections.* **(Right)** *Axial CT myelogram illustrates the separate spinal spaces due to a split injection of contrast. There is circumferential narrowing of the cauda equina due to contrast dissecting along the subdural space.*

Abscess, Spinal Cord

KEY FACTS

TERMINOLOGY

- Infection of spinal cord with necrosis

IMAGING

- Ring-enhancing mass within cord, with appropriate clinical history of inflammation/infection
- Increased T2WI signal from abscess core and surrounding edema; cord expansion
- Capsule may show low signal as in brain abscess
- May show positive diffusion (reduced reduced apparent diffusion coefficient [ADC]) similar to brain abscess
- Irregular ring-enhancing lesion with cord expansion

TOP DIFFERENTIAL DIAGNOSES

- Acute transverse/viral myelitis
- Hypervascular cord neoplasms
- Multiple sclerosis
- Cavernous malformation

PATHOLOGY

- 2 major etiologies
  - Adult intramedullary spinal cord abscess (ISCA) either hematogenous seeding from cardiopulmonary source or idiopathic
  - Children often have spinal dysraphism allowing direct extension of infection (40%)
  - Dermal sinus tract or dysraphism commonly present in children presenting with cord abscess

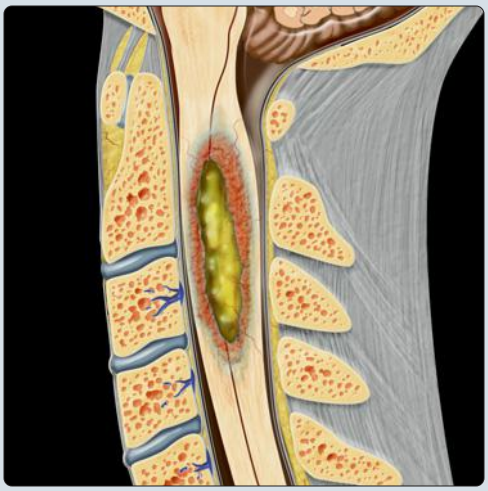
CLINICAL ISSUES

- Often present with signs/symptoms of structural cord lesion, rather than infection
- Mortality of 8% in antibiotic era (1977-1997)
- Surgically treated mortality approximately 14%

DIAGNOSTIC CHECKLIST

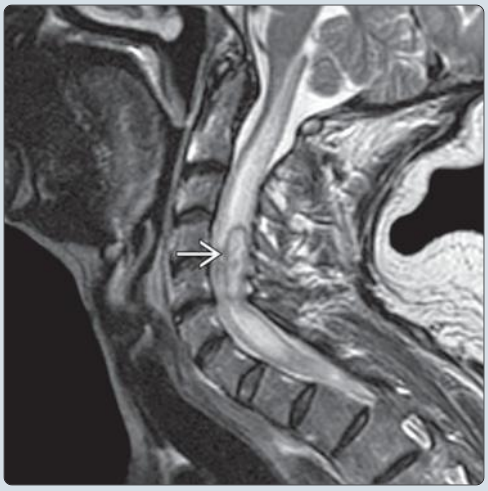
- MR brain to exclude concomitant brain abscess

(Left) *Sagittal graphic shows irregular abscess cavity in cervical cord, with cord expansion and edema.* **(Right)** *Sagittal T2WI MR shows high signal intensity centrally with a dark rim, often seen in inflammatory lesions and extensive edema, tracking rostrally into the medulla and caudally into the thoracic spinal cord. This appearance is not specific for abscess, and primary or secondary cord neoplasm would have to be considered as well.*





(Left) *Sagittal T2WI MR in a patient with streptococcal endocarditis shows diffuse cord expansion with a ring-shaped area of low T2 signal (abscess capsule) within the cord from C4 to C5-6* ➪. **(Right)** *Sagittal T1WI C+ MR in a patient with streptococcal endocarditis demonstrates the abscess capsule region showing ring enhancement* ➪.





TERMINOLOGY

Abbreviations

- Intramedullary spinal cord abscess (ISCA)

Definitions

- Infection of spinal cord with necrosis
  - Pyogenic infection most common

IMAGING

General Features

- Best diagnostic clue
  - Ring-enhancing mass within cord, with appropriate clinical history of inflammation/infection
- Location
  - Intramedullary
- Size
  - Variable; usually < 2 cm
- Morphology
  - Round or oblong irregular area of ring enhancement, with associated edema

CT Findings

- NECT
  - Normal or ill-defined cord irregularity and expansion

MR Findings

- T1WI
  - Ill-defined low signal from expanded cord
  - Abscess cord may show focal low signal
- T2WI
  - Increased T2WI signal from abscess core and surrounding edema; cord expansion
  - Capsule may show low signal as in brain abscess
- PD/intermediate
  - Increased signal from abscess and surrounding edema
- STIR
  - Good definition of cord edema
- DWI
  - May show positive diffusion (reduced apparent diffusion coefficient [ADC]) similar to brain abscess
    - Lack of diffusion restriction does not exclude abscess
- T1WI C+
  - Irregular ring-enhancing lesion with cord expansion

Nonvascular Interventions

- Myelography
  - Nonspecific findings of symmetrical cord enlargement, or myelographic block from intramedullary lesion

Imaging Recommendations

- Best imaging tool
  - MR with contrast defines cord abnormality, edema, enhancing abscess
- Protocol advice
  - Sagittal and axial T1W and T2W images, post-contrast sagittal and axial T1W images

DIFFERENTIAL DIAGNOSIS

Acute Transverse/Viral Myelitis

- Rapid onset of paraplegia ± sensory symptoms, bladder dysfunction
- Multiple viral agents implicated, either immune-mediated or direct viral invasion of cord
  - Echovirus, Coxsackie, cytomegalovirus, varicella-zoster, herpes simplex, Epstein-Barr, mumps, hepatitis, rubella, measles
- 1/3 improve, 1/3 partial improvement, 1/3 fail to improve
- Nonspecific T2 hyperintensity in cord, with variable enhancement (no ring enhancement)

Hypervascular Cord Neoplasms

- Ependymoma (mass centrally located; no syrinx)
- Hemangioblastoma (focal pial-based mass ± cyst)
- Astrocytoma (usually not hypervascular; peritumoral edema common)
- Vascular metastasis (known primary, e.g., renal cell carcinoma)

Multiple Sclerosis

- Peripheral in location
- > 2 vertebral segments in length
- 90% incidence of associated intracranial lesions
- May show ring enhancement

Type I Dural Fistula

- May rarely show only cord expansion and edema on T2WI, without prominent vessels
- No cord ring enhancement

Cavernous Malformation

- Mottled or speckled pattern of prior hemorrhage, hemosiderin rim

Cord Infarction

- Abrupt onset of weakness and loss of sensation
- Focal hyperintensity on T2WI in slightly expanded cord
- May show patchy, ill-defined enhancement in subacute phase

Intradural Extramedullary Tumors

- Homogeneous enhancement with demarcation between lesion and cord
  - Schwannoma
  - Meningioma

PATHOLOGY

General Features

- Etiology
  - 2 major etiologies
    - Adult ISCA either hematogenous seeding from cardiopulmonary source or idiopathic
    - Children often have spinal dysraphism allowing direct extension of infection (40%)
- Associated abnormalities
  - Dermal sinus tract or dysraphism commonly present in children presenting with cord abscess
- Rare: < 100 cases described

- Routes of infection: Majority of cases idiopathic/cryptogenic
  - Hematogenous spread (10%)
  - Contiguous spread from adjacent infection (25% of these cases from dermal sinus tract)
  - Direct implantation, such as trauma or surgery
- Microbiology
  - Multiple organisms involved
    - *Staphylococcus aureus*
    - *Staphylococcus epidermidis*
    - *Bacteroides*
    - *Haemophilus* species
    - *Listeria* monocytogenes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often present with signs/symptoms of structural cord lesion, rather than infection
  - Mixed neurologic deficits, including fever, pain, motor deficit, sensory disturbance, and sphincter dysfunction
  - Other signs/symptoms
    - Incontinence, meningismus
    - Triad of fever, pain, neurologic deficit in 25%
  - Delay in diagnosis common (time from symptom onset to diagnosis > 30 days)
    - Fever (< 50%)
  - Children
    - Paralysis (60%)
    - Fever (> 50%)
    - Urinary retention (30%)
    - Back pain (28%)
    - Nausea/vomiting (10%)
    - 20% recover without neurologic sequelae

Demographics

- Age
  - Adults and children (mean age: 34 years)
- Gender
  - M:F = 3:1
- Ethnicity
  - None known
- Epidemiology
  - Intramedullary infection rare
  - 0.2-2.2 cases per 10,000 tertiary care hospital admissions

Natural History & Prognosis

- Associated with high mortality and morbidity
- Mortality of 24% reported from years 1944-1977
  - Mortality of 8% in antibiotic era (1977-1997)
- Quick surgical drainage and aggressive intravenous antibiotics required
- Persistent neurologic deficit with successful treatment (> 70%)

Treatment

- Early surgical drainage followed by antibiotic therapy
  - Laminectomy with myelotomy and drainage
  - IV antibiotic coverage 4-6 weeks, with 2-3 additional months oral antibiotics
- Surgically treated mortality ~ 14%

DIAGNOSTIC CHECKLIST

Consider

- Ampicillin empiric coverage in idiopathic cases for *Listeria* monocytogenes
- Serial WBC count, ESR, enhanced MR important for postoperative follow-up, detection of recurrence

Image Interpretation Pearls

- MR brain to exclude concomitant brain abscess

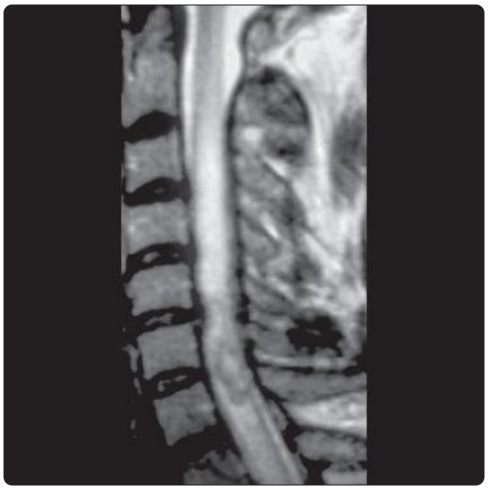
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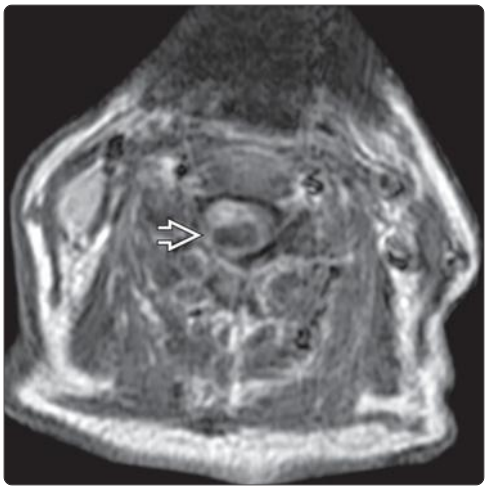
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(Left) *Sagittal T2WI MR in tuberculous myelitis shows a hypointense intramedullary mass surrounded by hyperintense edema at C7. The T2 hypointensity is typical for this infection but can be seen with tumor as well (such as leukemia).* **(Right)** *Sagittal T1 C+ FS MR in tuberculous myelitis shows ring-enhancing "target" lesion in the cord. Tuberculosis (TB) is much less common in the cord compared to the vertebral bodies and paraspinal soft tissues. Tumor is also in the differential.*





(Left) *Sagittal T2WI MR shows cord abscess as irregular low signal within the cord at C6-C7. There is extensive T2 hyperintense cord edema extending to the C2 level. (Courtesy J. Illes, MD.)* **(Right)** *Axial T1WI C+ MR in a patient with streptococcal endocarditis shows the intramedullary nature of the ring-enhancing mass. Brain examination (not shown) showed a typical pattern of abscess with positive diffusion study.*





(Left) *Sagittal T2WI MR in dorsal dermal sinus, with infection extending intradurally into the distal cord, shows a complex intradural mass engulfing the distal cord and cauda equina with extensive cord edema.* **(Right)** *Sagittal T1WI C+ MR in a case of dorsal dermal sinus, with infection extending into the cord, shows enhancement along the sinus tract and irregular enhancement surrounding the intradural cord abscess.*

Viral Myelitis

KEY FACTS

TERMINOLOGY

- Acute inflammatory insult of spinal cord due to direct viral infection or postviral immunologic attack

IMAGING

- Swollen, edematous spinal cord with segmental contiguous involvement
  - From 1 segment to extensive cord involvement
- Fusiform expansion of spinal cord
- Diffuse increase in T2 signal intensity through involved segment
- Variable, patchy enhancement of involved cord segment

TOP DIFFERENTIAL DIAGNOSES

- "Idiopathic" transverse myelitis
- Multiple sclerosis
- Acute cord infarct

PATHOLOGY

- Since near complete eradication of polio, other enteroviruses most common etiology
- Edematous, boggy cord ± necrosis

CLINICAL ISSUES

- Acute onset of weakness following febrile illness or upper respiratory tract infection
- CSF shows elevated mononuclear counts and protein level
- Principal treatment supportive with antiviral drugs ± steroids, variable efficacy

DIAGNOSTIC CHECKLIST

- Strong diagnostic clue in patients with acute onset of myelopathy: Long, segmental cord enlargement and edema without focal lesions

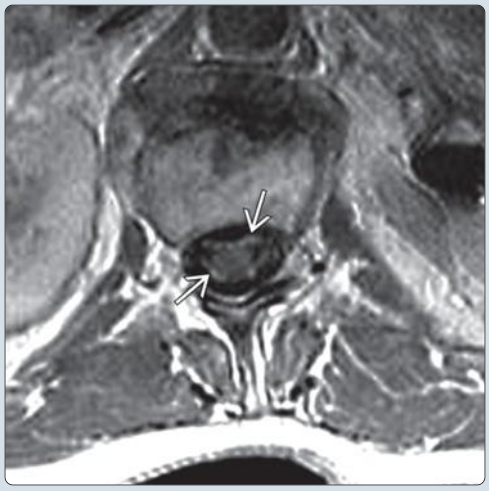
(Left) *Sagittal T2WI MR shows hyperintensity in the distal thoracic cord and conus ⇨ in this patient with West Nile virus (WNV), confirmed with positive IgM serology. WNV can cause meningitis, encephalitis, ± an irreversible acute flaccid paralysis (AFP). WNV shows a predilection for the deep gray nuclei in the brain and the spinal cord.* **(Right)** *Axial T2WI MR in the same patient with WNV demonstrates hyperintensity in the distal thoracic cord central gray matter ⇨.*





(Left) *Sagittal T1WI C+ FS MR displays enhancement along the distal cord and cauda equina ⇨ due to WNV. This patient had a poliomyelitis-like syndrome.* **(Right)** *Axial T1WI C+ MR of the same patient illustrates linear enhancement of the cauda equina ⇨. Most WNV-associated acute flaccid paralysis is due to spinal motor neuron involvement producing an anterior myelitis.*





TERMINOLOGY

Abbreviations

- Acute transverse myelitis (ATM)

Definitions

- Acute inflammatory insult of spinal cord due to direct viral infection or postviral immunologic attack

IMAGING

General Features

- Best diagnostic clue
  - Swollen, edematous spinal cord with segmental contiguous involvement
- Location
  - Cervical, thoracic segments; isolated conus involvement rare
- Size
  - From 1 segment to extensive cord involvement
- Morphology
  - Fusiform expansion of spinal cord

CT Findings

- CECT
  - ± vague patchy enhancement

MR Findings

- T1WI
  - Expanded cord fills canal
  - ± central low signal simulating syrinx, but intensity higher than CSF
- T2WI
  - Diffuse increase in signal intensity through involved segment
- T1WI C+
  - Variable, patchy enhancement of involved cord segment

Nonvascular Interventions

- Myelography
  - Focal or diffuse spinal cord enlargement
  - CT myelogram outlines circumferentially swollen spinal cord

Imaging Recommendations

- Best imaging tool
  - T2WI and STIR MR
- Protocol advice
  - T2WI with thin (3 mm) sagittal sections; T1 C+ to exclude focal lesion as cause of spinal cord edema

DIFFERENTIAL DIAGNOSIS

"Idiopathic" Transverse Myelitis

- Identical clinical picture
- No etiology found
- Up to 40% of cases preceded by upper respiratory tract infection
- Presence of CSF lymphocytes and neutrophils indicative of some type of inflammation in most cases
- Typically long segment of cord demonstrates swelling, edema, vague diffuse enhancement
- "Idiopathic" likely because of incomplete evaluation &/or follow-up
- May represent collateral damage of immune attack following infection already abated by host's defenses

Multiple Sclerosis (MS)

- Up to 33% may have isolated cord lesions (no brain lesions)
- Most lesions are focal (1-2 segments), may be multiple
- 20% demonstrate monosegmental involvement
- Acute lesions exhibit focal enhancement with short segment edema
- No peripheral nervous system involvement
- 90% of cases show oligoclonal bands

Acute Disseminated Encephalomyelitis (ADEM)

- Mimic of multiple sclerosis
- Related to vaccination or immune insult
- Monophasic illness

Neuromyelitis Optica

- Myelitis and optic neuritis
- Associated with several systemic diseases, including collagen vascular diseases, autoantibody syndromes, infections, and toxic exposures

Spinal Arteriovenous Malformation (AVM)

- High-flow AVM may cause acute myelopathy due to bleed or venous thrombosis
- Subacute onset of myelopathy, including edema; cord swelling may be due to "steal" and resulting ischemia
- Dural arteriovenous fistula (AVF) may cause stuttering symptoms or acute myelopathy (due to venous thrombosis → Foix-Alajouanine syndrome)

Arteritis

- Small vessel vasculitis may have no imaging abnormality

Infarction, Spinal Cord

- Acute stroke-like presentation
- Motor signs predominate
- Etiologies
  - Frequent: Atherosclerotic occlusion of anterior spinal artery
    - Aortic aneurysm repair, aortic dissection (vertebral artery dissection if cervical), arteriovenous malformation
    - Cryptogenic, transient ischemic attack, and cardiac arrest
- Conus involvement common (due to single ventral spinal artery)

Mycoplasma Myelitis

- Coincident with mycoplasma pneumonitis

PATHOLOGY

General Features

- Etiology
  - Since near complete eradication of polio, other enteroviruses most common etiology
    - Coxsackie, echovirus, hepatitis, rubella, measles, mumps
  - Herpes

  - Ebstein-Barr, varicella-zoster, cytomegalovirus, herpes simplex, herpesvirus 6, herpes B (monkey virus)
  - Retrovirus
    - Immunocompromised hosts
    - Human T-lymphotrophic virus I and II, HIV
  - Other
    - Influenza, rabies, West Nile virus (WNV)
- Associated abnormalities
  - Peripheral neuropathy
  - Encephalitis
  - Swollen, soft spinal cord
- Cytolytic infection of anterior horn cells → acute flaccid paralysis
  - Polioviruses-1, -2, and -3; Coxsackie viruses A and B; enterovirus-71; and flaviviruses, including WNV
    - WNV infection → local immune activation within spinal cord → failure of astrocyte glutamate reuptake despite ↑ number of astrocytes → rising extracellular glutamate levels → excitotoxic injury of infected and uninfected anterior horn cells
- Involvement of mainly white matter → virus-specific and autoimmune host cellular immune responses contribute to spinal cord injury
  - May be associated with infection by many different viruses

Gross Pathologic & Surgical Features

- Edematous, boggy cord
- ± necrosis

Microscopic Features

- Lymphocytic infiltrate
- Vascular thickening
- Demyelination

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Limb weakness
  - Areflexia
  - Other signs/symptoms
    - Sensory dysesthesia
    - Pain
- Clinical profile
  - Acute onset of weakness following febrile illness or upper respiratory tract infection
  - Reaches nadir at 1 week
  - CSF shows elevated mononuclear counts, protein level
  - Specific diagnosis through viral titers or polymerase chain reaction on CSF specimens
  - Acute viral myelitis
    - Can present as acute flaccid paralysis (poliomyelitis) or neurologic dysfunction due to involvement of WM
      - Transverse expanse of spinal cord affected → asymmetric motor and sensory symptoms
      - Both halves of spinal cord affected → **acute transverse myelitis**, symmetric weakness, sensory loss, and urinary bladder involvement
  - Chronic viral myelitis
    - May be due to either direct infection of spinal cord with HTLV-1 or metabolic disturbance due to HIV-1 infection in AIDS patients
      - No other human virus is known to chronically infect spinal cord without involvement of brain

Demographics

- Age
  - Typically younger patients
- Epidemiology
  - 1/100,000 per year
  - More common in immunocompromised

Natural History & Prognosis

- Most cases leave lasting residual effects; significant minority resolve
- Disease course: 1-12 weeks

Treatment

- Principal treatment: Antiviral drugs immediately upon virus isolation/identification by PCR
  - Variable efficacy
  - When indicated, high doses of methylprednisolone
- Supportive

DIAGNOSTIC CHECKLIST

Consider

- Brain MR to exclude multiple sclerosis, ADEM

Image Interpretation Pearls

- Strong diagnostic clue in patients with acute onset of myelopathy: Long, segmental cord enlargement and edema without focal lesions

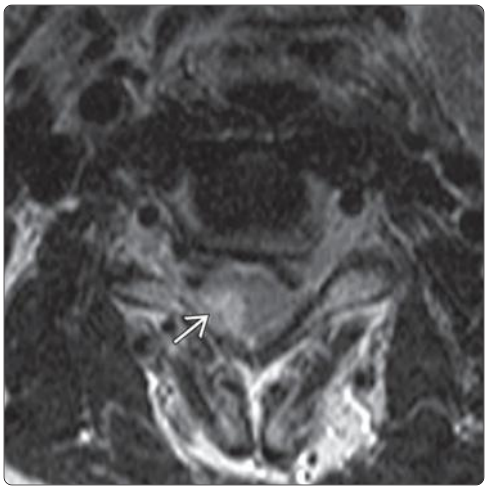
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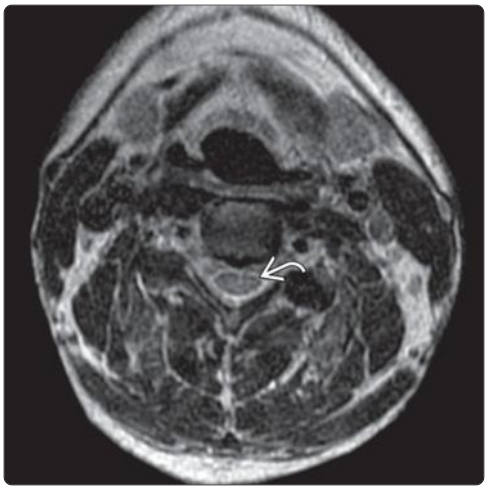
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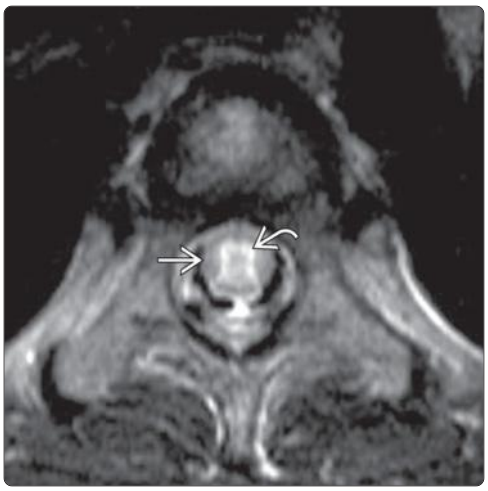
(Left) *Sagittal T1WI C+ MR demonstrates subtle clumping and linear enhancement of the cauda equina.* **(Right)** *Axial T1WI C+ MR shows predominant enhancement of the anterior nerve roots. Enhancement of the ventral surface of the conus and the anterior nerve roots of the ventral cauda equina has been described with various entities, including enterovirus 71, tick-borne encephalitis virus, WNV, and Guillain-Barré.*





(Left) *Axial T2WI MR of a patient with shingles depicts right cervical hemicord hyperintensity. A subset of patients with varicella-zoster reactivation may develop longitudinally extensive transverse myelitis.* **(Right)** *Axial T2WI MR shows atrophy of the cervical spinal cord with slight left-sided asymmetry in this patient with a history of polio.*





(Left) *Sagittal T2WI MR displays a swollen cervical Spinal cord and extensive hyperintensity in this patient with rabies.* **(Right)** *Axial T2WI MR of rabies radiculomyelitis exhibits marked involvement of the central cord gray matter and sparing of the peripheral white matter, a pattern suggestive of the diagnosis.*

HIV Myelitis

KEY FACTS

TERMINOLOGY

- Myelopathy resulting from primary HIV infection

IMAGING

- Most common: Atrophy
- Spinal cord T2 hyperintensity ± patchy enhancement
  - Thoracic > cervical with ↑ rostral involvement as disease progresses

TOP DIFFERENTIAL DIAGNOSES

- B12 deficiency
- Varicella-zoster virus
- Multiple sclerosis
- Transverse myelitis
- Human T-cell leukemia/lymphoma virus

PATHOLOGY

- DNA lentivirus/retrovirus attacks spine monocytes and macrophages
  - Disease of cord and brain often occurs separately, suggesting different pathogenetic mechanisms
- Opportunistic CNS and PNS infections and malignancies

CLINICAL ISSUES

- Insidious progressive spastic paraparesis with ataxia, urinary problems, and sensory loss
  - May have acute myelitic syndrome shortly after seroconversion
- Immune reconstitution inflammatory syndrome → myelopathy
- Diagnosis of exclusion based on clinical, laboratory, and radiologic findings
  - MR excludes other extrinsic or intrinsic processes

DIAGNOSTIC CHECKLIST

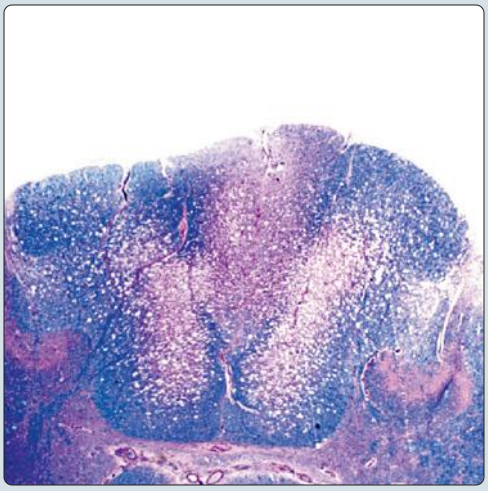
- Important to exclude other treatable causes of myelopathy
- Spinal cord atrophy is most common MR abnormality, typically involving thoracic SC ± cervical cord involvement

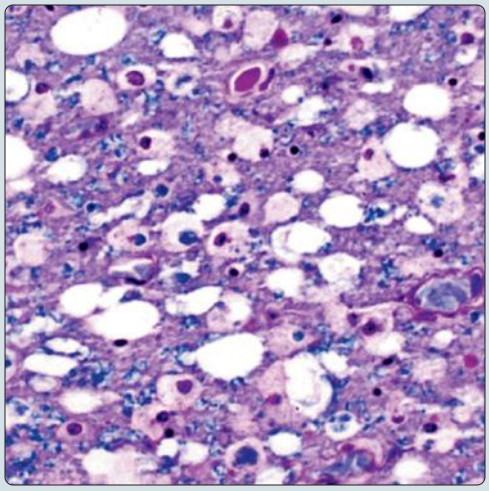
(Left) *High signal intensity abnormalities are seen in the cervical spinal cord on sagittal FLAIR MR. The cord is not swollen. The findings are consistent with HIV-related changes in the spinal cord.* **(Right)** *Axial T2WI MR demonstrates hyperintense signal in the right hemicord. The cord is not swollen. No enhancement was observed on post-contrast T1WI (not shown). The findings are consistent with HIV-related changes in the spinal cord.*





(Left) *Micropathology, low power with Luxol fast blue stain, shows extensive spongiform changes within white matter from vacuolar myelopathy. Disease of cord and brain often occur separately, suggesting different pathogenetic mechanisms. (Courtesy R. Schmidt, MD.)* **(Right)** *Micropathology, high power with Luxol fast blue stain, shows marked vacuolation within white matter from HIV myelopathy. (Courtesy R. Schmidt, MD.)*





TERMINOLOGY

Definitions

- Myelopathy resulting from primary HIV infection

IMAGING

General Features

- Best diagnostic clue
  - Spinal cord (SC) T2 hyperintensity ± patchy enhancement
- Location
  - Thoracic > cervical; mid to low thoracic cord with ↑ rostral involvement as disease progresses
- Morphology
  - Most common: Atrophy (72%)
  - Common
    - Diffuse nonspecific T2 hyperintensity of SC without definite pattern (29%)
    - Atrophy + diffuse abnormality (14%)
  - Classic: Symmetric T2 hyperintensity involving white matter (WM) tracts laterally

MR Findings

- T1WI
  - May be normal
  - SC atrophy
- T2WI
  - May be normal
  - Diffuse hyperintensity or symmetric hyperintensity involving posterior columns
  - SC atrophy
- STIR
  - Foci of hyperintensity
- T1WI C+
  - ± enhancement of visible lesions

Imaging Recommendations

- Best imaging tool
  - T2WI and T1WI C+ MR

DIFFERENTIAL DIAGNOSIS

B12 Deficiency

- May appear identical to HIV myelopathy
- Negative HIV test

Varicella-Zoster Virus

- Myelopathy
- CSF PCR-positive for virus

CMV Myelitis

- Cause of HIV-related polyradiculopathy
- ± nerve root and conus leptomeningeal thickening, enhancement on MR

Multiple Sclerosis

- Similar appearance with posterior and posterolateral location of demyelinating plaques
- Waxing and waning symptoms
- Negative HIV test

Transverse Myelitis

- May be indistinguishable by imaging
- Inflammation across width of enlarged spinal cord
- Uncertain etiology
- Will have normal CD4 count and negative HIV test
  - Some reports now of transverse myelitis developing after retroviral therapy in HIV(+) patients

Human T-Cell Leukemia/Lymphoma Virus (HTLV1)

- Myelopathy
  - Chronic, progressive spastic paresis
- Cerebral WM lesions (52%), acute phase: Diffuse T2 SC hyperintensity, chronic phase: SC atrophy (74%)
- Myelin and axon loss in lateral and anterior columns
- ± respond to corticosteroids or interferon

Anticardiolipin Antibody

- Relapsing-remitting myelopathy
- Enlarged SC with elongated multisegmental lesions in thoracic spine

PATHOLOGY

General Features

- Etiology
  - Primary HIV infection = DNA lentivirus/retrovirus that attacks spine monocytes and macrophages
- Genetics
  - Neurotropism is specific to certain HIV strains and may be related to monocyte tropism
    - May explain why HIV expression occurs in CNS of only certain AIDS patients
  - May relate to polymorphism of genes encoding for enzymes of transmethyl cycle
    - Methylation is essential for myelin formation, stabilization, and repair → interference with transmethyl pathway can cause WM vacuolization in SC
    - Usually mild or asymptomatic in normal individuals
    - ↑ demand for methyl donors or inhibition of methylation (as in HIV infection) may induce metabolic stress and hypomethylation
    - Result is genetic predisposition to cord disease in these susceptible and HIV-infected individuals
- Associated abnormalities
  - Opportunistic CNS and PNS infections (e.g., cytomegalovirus, toxoplasmosis, progressive multifocal leukoencephalopathy)
  - Opportunistic malignancies (e.g., lymphoma)

Staging, Grading, & Classification

- Criteria for diagnosis of AIDS-associated myelopathy
  - Male or female > 18 years with HIV-1 infection
  - AIDS-associated myelopathy, ± neuropathy and dementia
    - At least 2 of following symptoms
      - Paresthesia ± numbness in lower extremities or all 4 limbs
      - Weakness of limbs
      - Unsteady, stiff, or uncoordinated gait
      - Sensation of electric shock through back and legs on neck flexion
      - Urinary frequency, urgency, incontinence, or retention

        - Fecal incontinence or retention
        - Sexual dysfunction with male erectile impairment
      - At least 2 of following signs
        - Reduction in vibratory ± position sensation
        - Brisk deep tendon reflexes
        - Abnormal plantar response
        - Lhermitte sign
        - Spastic, ataxic, or ataxospastic gait
    - Signs and symptoms of AIDS-associated myelopathy for at least 6 weeks
    - Abnormal somatosensory evoked potentials
    - No other determinable cause for SC disease by serologic and CSF studies

Microscopic Features

- Vacuolar myelopathy (VM)
  - Direct injury of neurons by HIV
  - Intramyelin and periaxonal vacuolation of WM with posterior and lateral column demyelination
- Electron microscopy
  - Intramyelinic or periaxonal vacuoles, rarely disrupted axons
  - Myelin within macrophages
  - HIV-1 budding from macrophages
- RNA analysis demonstrates
  - HIV-1 expression in mononuclear and multinucleated macrophages in areas of myelopathy
  - HIV RNA detected in 100% of SC with VM
  - Level of RNA expression correlates directly with extent of SC pathology and clinical findings
- In children, SC WM is also affected
  - Diffuse demyelination with axonal loss and more prominent inflammatory infiltrate
  - Typical VM is rarely observed
  - Difference may be from different myelin structure or alternative pathologic mechanisms in children

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Insidious onset of progressive spastic paraparesis with ataxia, urinary problems, and sensory loss
    - Difficulty in obtaining or maintaining erection
    - Stiffness in legs develops, often accompanied by cramps and spasms → walking becomes difficult
    - In final stages, there may be severe paralysis of legs, loss of bladder, and anal sphincter control
    - Painless initially, in later stages cramps and spasms may cause serious discomfort or pain
    - Arms and hands generally not affected
- Other signs/symptoms
  - Immune reconstitution inflammatory syndrome
    - Myriad of manifestations: Disseminated tuberculosis, myelopathy, encephalopathy, deep venous thrombosis, cryptococcal meningoradiculitis
    - May cause progression of vacuolar myelopathy to symptomatic disease after initiation of antiretroviral therapy
- Clinical profile
  - Very low CD4(+) lymphocyte counts
  - Often have vitamin B12 deficiency
    - May be related to polymorphism of transmethyl cycle enzyme genes
  - CSF: Normal in HIV-associated VM
    - Used to exclude other viral pathogens

Demographics

- Epidemiology
  - AIDS-associated myelopathy prevalence of 20-55%
  - Up to 20% in clinical and 55% in histologic studies

Treatment

- MR is essential in exclusion of other extrinsic or intrinsic processes
  - However, normal-appearing SC often seen in mild and severe myelopathy
  - Cord atrophy and signal abnormality are found in both mildly and severely myelopathic patients
- Treatment studies using highly active antiretroviral therapy (HAART) → less common SC pathology
  - Estimated < 10% of patients with AIDS now develop SC pathology
- Improvement with L-methionine has been reported
- Symptomatic treatment includes antispasticity agents, management of sphincter dysfunction, and physical therapy

DIAGNOSTIC CHECKLIST

Consider

- Need to exclude other treatable causes of myelopathy

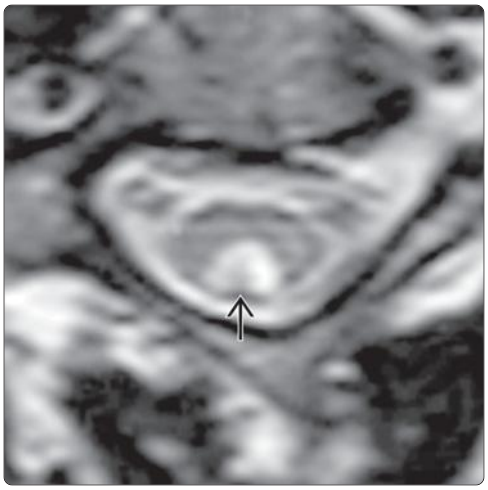
Image Interpretation Pearls

- MR is often noncontributory, but may reveal unsuspected coexisting conditions or other explanation for patient symptoms
- SC atrophy is most common MR abnormality, typically involving thoracic SC ± cervical cord involvement

SELECTED REFERENCES

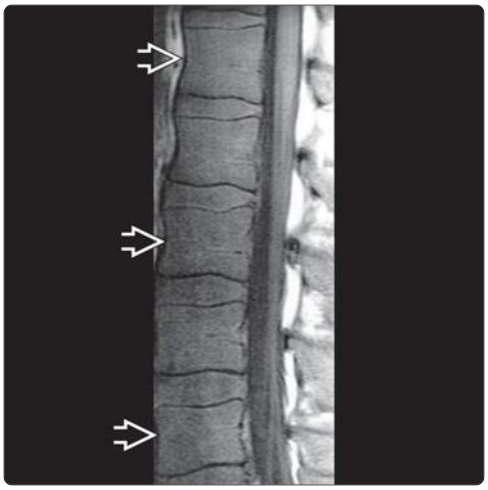
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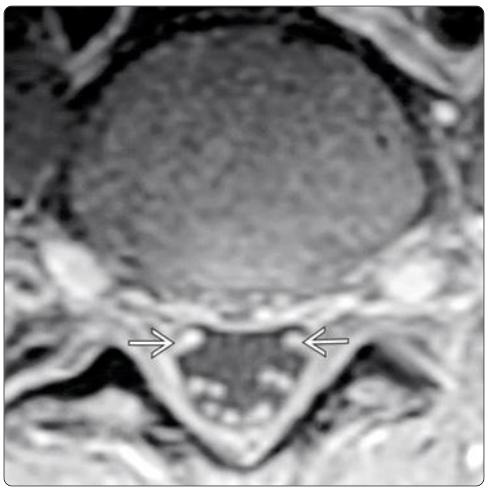
(Left) *Sagittal T2WI MR displays intramedullary hyperintensity within the posterior cervical cord due to HIV myelopathy. Infection by HIV-1 may stimulate macrophages to phagocytize myelin. These macrophages may secrete factors damaging neural tissue. HIV infection can interfere with transmethyl pathway and cause cord white matter (WM) vacuolization.* **(Right)** *Axial T2WI MR shows hyperintensity within posterior cervical cord in a patient with HIV myelopathy. (Courtesy F. Thomas, MD, PhD.)*





(Left) *Sagittal T2WI MR demonstrates diffuse, central T2 hyperintensity from primary HIV myelitis. There is mild diffuse cord atrophy. (Courtesy American Journal of Neuroradiology [AJNR].)* **(Right)** *Sagittal T1WI MR depicts diffuse hypointensity of the vertebral marrow. This abnormality may be the result of markedly increased amounts of storage iron within the bone marrow, probably secondary to the anemia of chronic disease.*





(Left) *Sagittal T1WI C+ FS MR exhibits diffuse enhancement of the cauda equina extending to the conus. No focal masses are present. There is an enhancing annular tear at the L4-L5 level. Without the history, other differentials include primarily CSF spread of tumor and bacterial meningitis.* **(Right)** *Axial T1WI C+ FS MR demonstrates diffuse cauda equina enhancement due to AIDS polyradiculopathy. This is the typical pattern for CMV radiculitis.*

Syphilitic Myelitis

KEY FACTS

TERMINOLOGY

- Synonyms: Neurosyphilis, syphilitic meningomyelitis
- CNS infection by spirochete *Treponema pallidum*

IMAGING

- Variable nodular and linear enhancement with diffuse cord T2 hyperintensity

TOP DIFFERENTIAL DIAGNOSES

- Spinal vascular malformation
- Postinfectious demyelination
- Viral myelitis
- Ischemia
- Sarcoidosis

PATHOLOGY

- Neurosyphilis divided into early and late forms and may occur during primary or secondary infection
- **Early** affects CSF, vessels, and meninges more often than brain or cord
- **Late** affects meninges and brain or cord parenchyma
  - Very rare in antibiotic era
  - General paresis, dementia, tabes dorsalis (sensory ataxia with bowel and bladder dysfunction)

CLINICAL ISSUES

- Lower extremity weakness, pyramidal signs, bowl and bladder dysfunction, polyradiculopathy
- Nontreponemal serology (RPR, VDRL) 78-86% sensitive in 1° syphilis, 100% sensitive in 2°
- Varying incidence rates by geography, race, and sexual orientation
- Negative CSF fluorescent treponemal antibodies (FTA-abs) excludes neurosyphilis





(Left) *Sagittal T2WI MR shows diffuse cord edema as T2 hyperintensity with diffuse cord expansion obscuring any CSF surrounding cervical cord. (Courtesy E. Gasparetto, MD.)* **(Right)** *Sagittal T1WI C+ FS MR shows multiple nodular foci of enhancement primarily located on the cord pial surface extending from medulla throughout the cervical cord. More severe pial involvement is referred to as "candle guttering" pattern with linear and nodular foci. (Courtesy E. Gasparetto, MD.)*





(Left) *Sagittal STIR MR shows diffuse T2 hyperintensity throughout the thoracic cord in this patient with subacute syphilitic myelitis. Slight irregularity or T2 hyperintensity ➡ corresponds to foci of enhancement. (Courtesy E. Gasparetto, MD.)* **(Right)** *Sagittal T1WI C+ FS MR shows multiple nodular foci of enhancement throughout the thoracic cord, with a strong predominance toward the cord pial surface. (Courtesy E. Gasparetto, MD.)*

TERMINOLOGY

Synonyms

- Neurosyphilis, syphilitic meningomyelitis

Definitions

- CNS infection by spirochete *Treponema pallidum*

IMAGING

General Features

- Best diagnostic clue
  - Neurosyphilis diagnosis requires 1 of the following CSF pathologies
    - Increased cell count or protein
    - Reactive VDRL or RPR
    - Positive iTPA index
    - Positive PCR for *T. pallidum*
- Location
  - Pial surface of cord &/or intramedullary
- Size
  - Variable nodular and linear enhancement

Imaging Recommendations

- Best imaging tool
  - Enhanced MR to define cord edema and enhancing leptomeningeal or cord parenchymal enhancement

DIFFERENTIAL DIAGNOSIS

Spinal Vascular Malformation

- Flow voids on cord surface (type 1) or within cord (type 2, 3)

Postinfectious Demyelination

- Diffuse cord hyperintensity with variable enhancement

Viral Myelitis

- Nonspecific cord T2 hyperintensity
- Diffuse, rather than nodular, enhancement

Ischemia

- Abrupt onset, with minimal enhancement

Sarcoidosis

- May mimic syphilis appearance with cord edema, leptomeningeal, and cord nodular enhancement

PATHOLOGY

Staging, Grading, & Classification

- Syphilis infection occurs in series of overlapping stages
  - Primary infection by contact with infectious lesion or body fluids
    - Skin lesion classic (chancre)
  - Secondary syphilis occurs 4-10 weeks after chancre
    - Rash is presenting complaint in 70%
  - Tertiary syphilis occurs in 1/3 of untreated patients
    - Cardiovascular and gummatous forms
- Neurosyphilis divided into early and late forms and may occur during primary or secondary infection
  - **Early** affects CSF, vessels, and meninges more often than brain or cord
    - Symptomatic forms include meningitis, meningovascular disease, or stroke
  - **Late** affects meninges and brain or cord parenchyma
    - Very rare in antibiotic era
    - General paresis, dementia, tabes dorsalis (sensory ataxia with bowel and bladder dysfunction)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Lower extremity weakness, pyramidal signs, bowl and bladder dysfunction, polyradiculopathy
- Other signs/symptoms
  - Nontreponemal serology (RPR, VDRL) 78-86% sensitive in primary syphilis, 100% sensitive in secondary
  - Treponemal specific tests (TPPA) and fluorescent treponemal antibodies (FTA-abs) are sensitive but nonspecific
    - Negative CSF FTA-abs excludes neurosyphilis

Demographics

- Epidemiology
  - Varying incidence rates by geography, race, and sexual orientation
    - Heterosexual rates highest in southern USA
    - Rate in black women 42x higher than in white women
    - Increased incidence in HIV-positive men

Natural History & Prognosis

- 1/3 of patients with early syphilis have CNS invasion
- Symptomatic neurosyphilis is rare

Treatment

- Neurosyphilis: 18-24 million units of penicillin IV per day for 10-14 days
  - Steroids added to control cord edema

DIAGNOSTIC CHECKLIST

Consider

- Patients with neurosyphilis should have repeat CSF exam every 3-6 months after therapy until CSF normal

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KEY FACTS

TERMINOLOGY

- Infection in setting of immunocompromised state, chronic/systemic illness, steroid therapy, cancer ± chemotherapy

IMAGING

- Discitis/osteomyelitis → loss of disc height, bony endplate erosive changes
  - Enhancement of irregular adjacent endplates, blurring of dark cortical margin of endplates
- Epidural abscess/phlegmon → soft tissue in ventral/dorsal extradural space
  - Homogeneous enhancement of phlegmon
  - Rim enhancement of abscess
- Paraspinal abscess/phlegmon
- Infectious arachnoiditis → thickening, clumping of nerve roots
- Intramedullary abscess → peripherally enhancing intramedullary lesion

TOP DIFFERENTIAL DIAGNOSES

- Osteomyelitis, pyogenic
- Meningitis, spinal
- ADEM, spinal cord
- Acute transverse myelitis, idiopathic

PATHOLOGY

- CMV → intranuclear inclusions

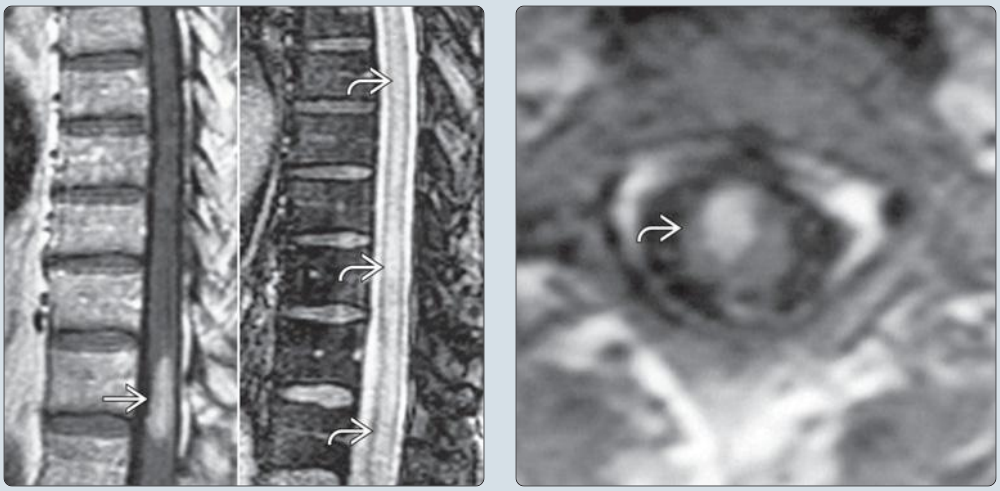
CLINICAL ISSUES

- Fever
- Back/neck pain
- Paraparesis
- Treatment targeted to pathogen

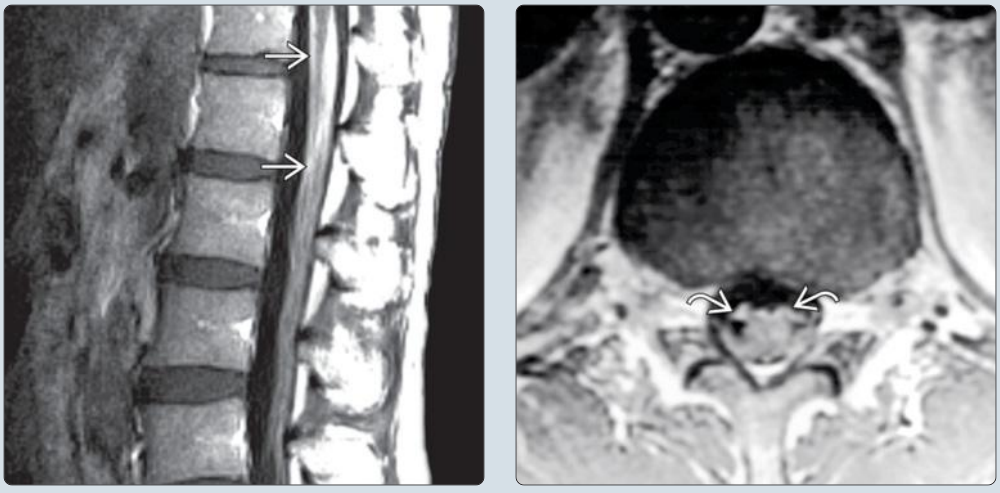
DIAGNOSTIC CHECKLIST

- Neuraxis imaging in immunocompromised patients with neurologic symptoms

(Left) *Sagittal T1WI C+ MR (left) shows an intramedullary enhancement in the lower thoracic cord representing toxoplasma lesion. On the sagittal T2WI MR (right), abnormal hyperintense signal extends across multiple segments.* **(Right)** *Axial T1WI C+ MR demonstrates a nodular enhancing intramedullary lesion, which represents toxoplasma abscess of the spinal cord. The cord is slightly swollen. Spinal toxoplasmosis has only been described in HIV patients with simultaneously present cerebral toxoplasmosis.*



(Left) *Sagittal T1WI C+ MR in this case of CMV polyradiculopathy illustrates mild thickening of the proximal cauda equina and avid pial and cauda equina enhancement.* **(Right)** *Axial T1WI C+ MR depicts diffuse cauda enhancement due to CMV radiculitis. The multiple root involvement with relatively smooth enhancement suggests CMV infection. CSF spread of tumor, lymphoma, and pyogenic meningitis can give a similar pattern, although tends to be more nodular.*



TERMINOLOGY

Definitions

- Infection in setting of immunocompromised state, chronic illness, systemic infection, corticosteroid therapy, cancer ± chemotherapy

IMAGING

General Features

- Best diagnostic clue
  - Intramedullary, intradural extramedullary, or extradural involvement
  - Additional systemic manifestations, such as pulmonary disease

CT Findings

- Discitis/osteomyelitis → loss of disc height, bony endplate erosive changes
  - Enhancement of irregular adjacent endplates
  - ± enhancement of intervertebral disc
- Epidural abscess/phlegmon → hyperdense collection in ventral/dorsal extradural space
  - Homogeneous enhancement of phlegmon
  - Rim enhancement of abscess
- Paraspinal abscess → hyperdense collection involving paravertebral soft tissues
  - Homogeneous enhancement of phlegmon
  - Rim enhancement of abscess
- Infectious arachnoiditis → thickening, clumping of nerve roots
  - Avid enhancement of nerve roots
- Intramedullary abscess → peripherally enhancing intramedullary low-density lesion
  - Surrounding edema
  - Cord expansion

MR Findings

- Discitis/osteomyelitis → loss of disc height, ± T2 hyperintensity in disc space
  - Enhancement of irregular adjacent endplates, blurring of dark cortical margin of endplates
  - ± enhancement of intervertebral disc
- Epidural abscess → soft tissue in ventral/dorsal extradural space
  - Homogeneous enhancement of phlegmon
  - Rim enhancement of abscess
- Paraspinal abscess → soft tissue infiltrating paravertebral soft tissues
  - Homogeneous enhancement of phlegmon
  - Rim enhancement of abscess
- Infectious arachnoiditis → enhancing thick, clumped nerve roots
  - CMV myelitis → nerve root and conus leptomeningeal thickening and enhancement
- Intramedullary abscess → peripherally enhancing intramedullary low T1 lesion with surrounding edema
  - Cord expansion

Imaging Recommendations

- Best imaging tool
  - CT → define osseous involvement
  - MR → evaluate extent of intramedullary, intradural extramedullary, and extradural involvement

Nuclear Medicine Findings

- Labeled leukocyte scintigraphy
  - Cells labeled with Tc-99m exametazime or In-111 oxine
    - Less useful for illnesses in which predominant cellular response is other than neutrophilic, such as most opportunistic infections and spinal osteomyelitis
- ^{123}I-labeled chitinase shown to accumulate in tissue infected with *Candida albicans* and *Aspergillus fumigatus*
  - → tracer for fungal infection

DIFFERENTIAL DIAGNOSIS

Osteomyelitis, Granulomatous

- Infection of spine and adjacent soft tissues caused by TB or brucellosis
- TB: Gibbus vertebrae with relative sparing of intervertebral discs and large paraspinal abscess
  - Posterior element involvement
  - Spread to (non)adjacent vertebral bodies beneath longitudinal ligaments
- Brucellosis: Lower lumbar spondylitis with bilateral sacroiliitis
  - Anterior endplate at discovertebral junction

Osteomyelitis, Pyogenic

- Bacterial suppurative infection of vertebrae and intervertebral disc
- Ill-defined hypointense T1/hyperintense T2 vertebral marrow with obscuration of endplate definition
  - Initial infection of subchondral bone adjacent to endplate
  - Intervening disc space narrowing ± T2 hyperintensity ± enhancement
  - Paraspinal ± epidural infiltrative soft tissue ± fluid collection
  - Cord compression ± intramedullary T2 hyperintensity

Fungal Osteomyelitis and Miscellaneous

- Noncaseating, acid-fast-negative infections primarily occurring as opportunistic infections in immunocompromised patients
- Osseous destructive process ± disc, epidural, or paraspinal involvement

Meningitis, Spinal

- Infection of spinal cord leptomeninges and subarachnoid space
- Clumped nerve roots → smooth or nodular meningeal enhancement

HIV Myelitis, Spine

- Spinal cord T2 hyperintensity ± patchy enhancement
- Cord atrophy
- Thoracic > cervical with rostral involvement as disease progresses

Viral Myelitis

- Acute inflammatory injury of spinal cord due to direct viral infection or post-viral immunologic attack

- Long, segmental cord enlargement and edema without focal lesions
- Variable, nonfocal enhancement of involved segment

ADEM, Spinal Cord

- Immune-mediated demyelinating disorder
- Multifocal flame-shaped white matter lesions with relatively little mass effect or vasogenic edema
- Variable enhancement, related to stage of disease
- Brain almost always involved

Acute Transverse Myelitis, Idiopathic

- Inflammatory disorder involving both halves of spinal cord
  - → bilateral motor, sensory, and autonomic dysfunction
- Peripheral enhancement in centrally located T2 hyperintensity

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, back/neck pain
  - Paraparesis
  - CMV lumbosacral polyradiculopathy → acute ascending flaccid paralysis of lower limbs with areflexia, paresthesia, urinary, and bowel symptoms
- Other signs/symptoms
  - Pulmonary symptoms

Demographics

- Epidemiology
  - Conditions causing immunocompromised state
    - Chronic steroid treatment
    - Neutropenia, reduced neutrophil activity, reduced hematopoietic stem cells
    - HIV infection
      - CD4 count can be used as predictor of clinical course, as follows
      - Mild to moderate decrease (≥ 200 cells/mm³) → discitis/osteomyelitis responds to antibiotics
      - Severe decrease (50-200 cells/mm³) → spinal TB
      - Lowest CD4 counts → epidural abscess
      - Occurrence of spondylodiscitis in HIV-positive patients is associated with low CD4 T-cell count
      - Probability of mixed infections rises with CD4 T-cell count < 100/microL
    - Cancer ± cytotoxic chemotherapy
      - *Blastoschizomyces capitatus* → emergent fungal pathogen causing spondylodiscitis in cancer patients
    - Organ transplant and immunosuppressive therapy
      - Recipients of solid organ transplantation (because of immunosuppressive therapy) → spinal TB
  - Immune reconstitution inflammatory syndrome (IRIS)
    - Patients with acquired immunodeficiency syndrome (AIDS) are at increased risk for developing CNS-IRIS upon initiation of highly active antiretroviral therapy
      - Typically associated with mycobacterial infections (such as *Mycobacterium avium*), herpes virus infections (such as herpes zoster and Cytomegalovirus [CMV]), and mycoses
      - Varicella-Zoster virus vasculopathy
      - Recurrent myelopathy due to TB
  - CMV lumbosacral polyradiculopathy → complication of HIV patients with CD4 count < 40/microL
  - *Toxoplasma gondii* can produce acute focal or diffuse meningitis and encephalitis, with destruction of cells and signs of inflammation
    - Cord abscess, adhesive arachnoiditis
  - Fungal spondylitis
  - Sarcoid → cryptococcal spondylitis with spinal cord involvement
  - *Nocardia* → respiratory tract disease is main syndrome
    - 50% with hematogenous dissemination
    - Primary CNS infection (5-7%)
  - Systemic lupus erythematosus → increased risk of developing osteoarticular TB
  - Invasive *Aspergillus* disease → 0-12% of those infected with HIV, highly lethal in advanced AIDS
    - *Aspergillus* infection → prolonged immunosuppression and higher numbers of opportunistic infections
    - AIDS patients develop acute invasive pulmonary aspergillosis → rapid evolution and hematogenous spread

Natural History & Prognosis

- Occurrence of spondylodiscitis in HIV-positive patients is accompanied by high mortality

Treatment

- Treatment targeted to pathogen
- Conservative or surgical treatment for spondylodiscitis
  - No increase in surgical complication rate in operative therapy of spondylodiscitis in HIV-positive patients

DIAGNOSTIC CHECKLIST

Consider

- Neuraxis imaging in immunocompromised patients with neurologic symptoms

Image Interpretation Pearls

- Fungal osteomyelitis should be considered in patients with evidence of destructive spinal process, particularly when tuberculosis is in differential

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(Left) *Sagittal T2WI MR exhibits expansion of cord with intrinsic heterogeneous hyperintensity. Abnormal signal extends from C2 through upper thoracic cord. Patient was taking steroid chronically for arthritis and developed spinal cord abscess from Listeria with associated Listeria meningitis.* **(Right)** *Sagittal T1WI C+ MR reveals hypointense cord expansion and heterogeneous intramedullary and leptomeningeal enhancement due to Listeria cord abscess and meningitis.*





(Left) *Sagittal T2WI MR shows cord expansion and a hypointense intramedullary mass surrounded by hyperintense edema at C7. T2 hypointensity is characteristic for TB.* **(Right)** *Axial CECT demonstrates an irregular mass surrounding an infected abdominal aortic graft and adjacent psoas muscle, with the abscess extending into the anterior aspect of the L3 vertebral body. This mycotic infection causes osseous destruction in the anterior vertebral body.*





(Left) *Sagittal T2WI MR shows extensive hyperintensity from the cervical cord, with diffuse cord expansion down to the thoracic level due to herpes infection. The long segment of involvement with enhancement favors direct viral infection or post-viral demyelinating disease.* **(Right)** *Sagittal T1WI C+ MR demonstrates extensive, patchy, and nonfocal enhancement of the cord over the C4 to the C7-T1 level. The extensive cord edema and enhancement is due to herpes infection.*

Echinococcosis

KEY FACTS

TERMINOLOGY

- Synonyms: Hydatidosis, hydatid disease, hydatid cyst disease
- Disease caused by cyst stage of infestation by tapeworm *Echinococcus* spp.
- Liver, lung involvement most common

IMAGING

- CT delineated bone morphology, degree of bone destruction
  - Minimally enhancing, multiloculated, osteolytic lesions in vertebral body, posterior elements
- MR shows bone, epidural, paravertebral extension and degree of neural compromise
  - Multiloculated, multiseptated, T2-hyperintense vertebral body/posterior element mass without significant enhancement

PATHOLOGY

- Bone involvement rare: Incidence ranges from 0.5-4% of all cases of echinococcosis
  - Vertebrae most common bone involved
  - Typical involvement of vertebral bodies, contiguous ribs, and paravertebral tissue
  - Disc space generally spared
  - No pericyst formation in bone involvement

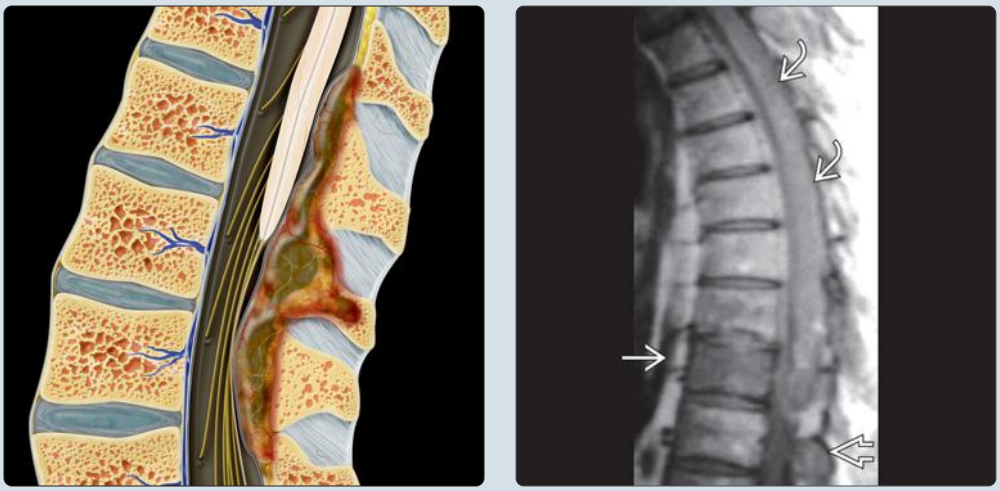
CLINICAL ISSUES

- Back pain, myelopathy, radiculopathy depending upon anatomic location of infestation
- Progressive weakness, numbness of extremities
- Indolent, progressive course without treatment

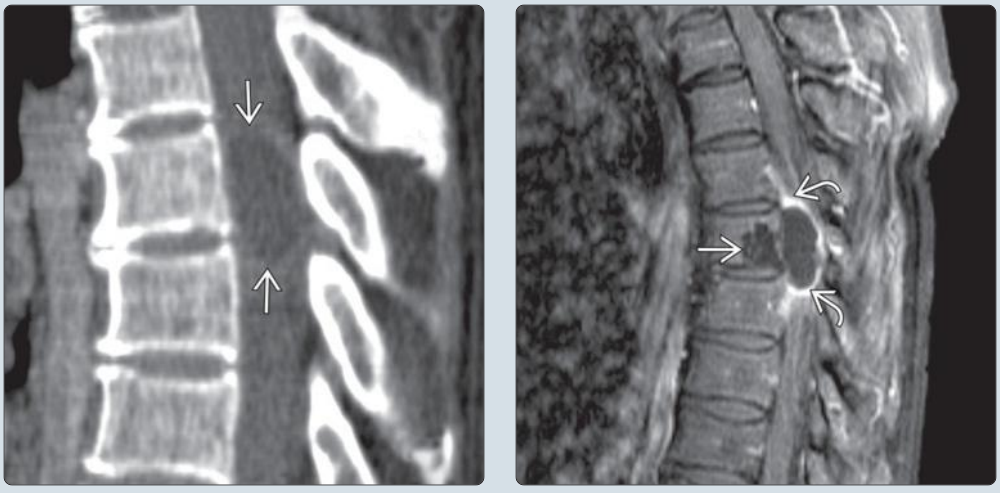
DIAGNOSTIC CHECKLIST

- Lack of significant enhancement is critical differential feature of echinococcal cysts

(Left) *Sagittal graphic of the lumbar spine shows cystic mass involving posterior elements with extension into dorsal epidural space, thecal sac, and cauda equina compression.* **(Right)** *Sagittal T1WI MR shows complex echinococcal mass involving vertebral body, posterior elements, and diffuse epidural extension. Cortical vertebral weakening due to compression or avascular necrosis may eventually lead to fractures resulting in spinal cord or nerve root compression. (Courtesy J. Beltran, MD.)*



(Left) *Sagittal CT illustrates a low-density rounded lesion in the thoracic spinal canal.* **(Right)** *Sagittal T1WI C+ FS MR exhibits a peripherally enhancing intraspinal lesion likely extending from the vertebral body. The primary site of the infestation and the precise extension of the spinal hydatid disease can be difficult to verify. There are 5 types: Primary intramedullary, intradural extramedullary, extradural intraspinal, vertebral hydatid disease, & paravertebral hydatid disease.*



TERMINOLOGY

Synonyms

- Hydatidosis, hydatid disease, hydatid cyst disease

Definitions

- Disease caused by cyst stage of infestation by tapeworm *Echinococcus* spp.
- Liver, lung involvement most common
  - Bone (0.5-4%) → spine involved in 50% of cases
- Multiple echinococcal species produce disease; complex life cycle
  - *Echinococcus granulosus*: Causative agent of hydatid disease in man/other mammals
    - Dog is definite host; parasitized by adult tapeworm
      - Gravid proglottids shed by tapeworm, disintegrate in dog intestine
      - Eggs passed in feces
    - Mammals act as intermediate host after ingestion of eggs (typically sheep, cattle)
      - Eggs hatch in intestine, penetrate gut wall, travel via lymphatic/blood vessels throughout body
      - Cyst develops into infective protoscolices in 1-2 years
    - Dogs then ingest protoscolices on death of intermediate host (predation/scavenging)
      - Cyst wall digested in gut, allowing development of adult worms
  - *Echinococcus multilocularis*: Causative agent of alveolar hydatid disease in man/other mammals
    - Fox is most important definitive host
    - Cyst grows invasively by external budding, diffuse growth through infected organ
      - Presents as ramifying, porous, necrotic mass
  - *Echinococcus vogeli*: Polycystic form in Central and South America
    - Bush dog is definitive host

IMAGING

General Features

- Best diagnostic clue
  - Multiloculated, multiseptated, T2-hyperintense vertebral body/posterior element mass without significant enhancement in endemic area for *Echinococcus*
- Location
  - Thoracic 50%, cervical 10%, lumbar 20%, sacrum 20%
    - Extradural
    - Intradural extramedullary is rare, intramedullary is **extremely** rare
- Size
  - Variable, up to several centimeters
- Morphology
  - Round, multiseptated, multiloculated cysts; bone expansion

Radiographic Findings

- Radiography
  - Multiloculated osteolytic lesions have "cluster of grapes" morphology
  - Nonspecific bone destruction

CT Findings

- NECT
  - Multiloculated osteolytic lesions in vertebral body, posterior elements
  - Paraspinal mass associated with vertebral body lesions
- CECT
  - Minimal/no enhancement of paraspinal/vertebral body cysts

MR Findings

- T1WI
  - CSF signal multiseptated/multicystic lesions
  - Degenerated cysts may be isointense to muscle
- T2WI
  - CSF signal multiseptated/multicystic lesions
  - Epidural extension of soft tissue with cord compression
  - Degenerated cysts may show low signal relative to CSF
- STIR
  - CSF signal multiseptated/multicystic lesions
- DWI
  - Can help differentiate complicated infected hydatidosis from abscesses
- T1WI C+
  - Minimal or no enhancement of large cystic lesions

Nonvascular Interventions

- Myelography
  - May show block with significant epidural extension of cysts

Imaging Recommendations

- Best imaging tool
  - MR shows bone, epidural, paravertebral extension and degree of neural compromise
  - CT shows bone morphology, degree of bone destruction
- Protocol advice
  - Sagittal, axial T1WI + T2WI, post-contrast sagittal, axial T1WI

DIFFERENTIAL DIAGNOSIS

Primary Bone Neoplasm

- Osteosarcoma
- Chondrosarcoma
- Aneurysmal bone cyst
- Giant cell
- Chordoma

Metastatic Disease

- Cystic metastases, such as renal cell, thyroid

Granulomatous Osteomyelitis

- Vertebral body, posterior element, and epidural space involvement
- Diffuse + peripheral enhancement of phlegmon, cold abscesses

Other Parasitic Diseases

- Cord inflammatory myelopathy with schistosomiasis
- Complex cystic intramedullary enhancing mass with cysticercosis

Arachnoid Cyst

- Extradural or intradural extramedullary
- CSF signal intensities on all sequences, no enhancement

PATHOLOGY

General Features

- Etiology
  - Usual cause is accidental ingestion of dog feces containing echinococcal eggs
  - Cysts slowly grow in vertebral body/posterior elements with subsequent extension into pre-/paravertebral region, ribs, epidural space with cord compression
- Bone involvement rare: Incidence ranges from 0.5-4% of all cases of echinococcosis
  - Vertebrae most common bone involved
  - Vertebrae > long bone epiphyses > ilium > skull > ribs
- No pericyst formation in bone involvement
  - Parasite expands by exogenous proliferation with irregular pattern
  - Bone resorption due to pressure erosion and necrosis
- Typical involvement of vertebral bodies, contiguous ribs, and paravertebral tissue
  - Disc space generally spared

Staging, Grading, & Classification

- 5 categories of spine involvement
  - Intramedullary cyst
  - Intradural extramedullary
  - Extradural intraspinal
  - Vertebral disease
  - Paravertebral

Gross Pathologic & Surgical Features

- Spherical, fluid-filled, hollow cyst containing multiple protoscolices, brood capsules, and daughter cysts
- Pearly white

Microscopic Features

- Outer nonnucleated hyaline-laminated wall
- Inner nucleated germinal layer, studded with developing brood capsules
  - Protoscolices formed within brood capsules

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, myelopathy, radiculopathy depending upon anatomic location of infestation
  - Other signs/symptoms
    - Progressive weakness, numbness of extremities
    - Nerve root compression
- Other signs/symptoms
  - Eg1 antigen in ELISA system used in diagnosis of cerebrospinal hydatidosis

Demographics

- Age
  - Any age
- Gender
  - No gender predilection
- Ethnicity
  - Related to endemic geographic locations
- Epidemiology
  - *Echinococcus* common in southern South America, Mediterranean, Middle East, Central Asia, and Africa
  - USA: California, Arizona, New Mexico, and Utah

Natural History & Prognosis

- Indolent, progressive course without treatment
- Alveolar form has 100% mortality rate
- Secondary infection of cyst often causes death of parasite

Treatment

- Oral treatment with albendazole or mebendazole
- Surgical resection for neural compression
  - Anterior decompression ± laminectomy
- Recurrence rate of spinal hydatid cysts after surgery is 18%

DIAGNOSTIC CHECKLIST

Consider

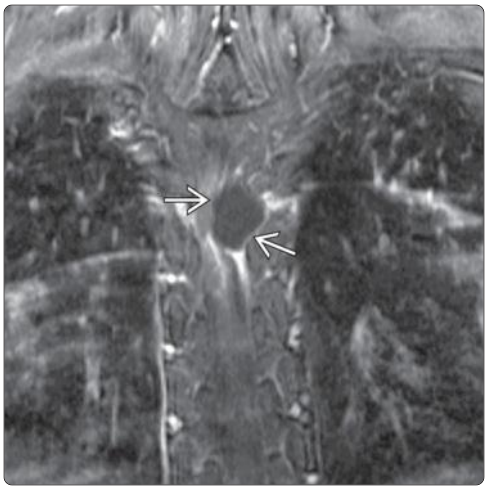
- Hydatid cyst fluid highly allergenic; cyst rupture may cause anaphylaxis
- Disease is indolent with latent period of multiple years before cysts reach symptomatic size

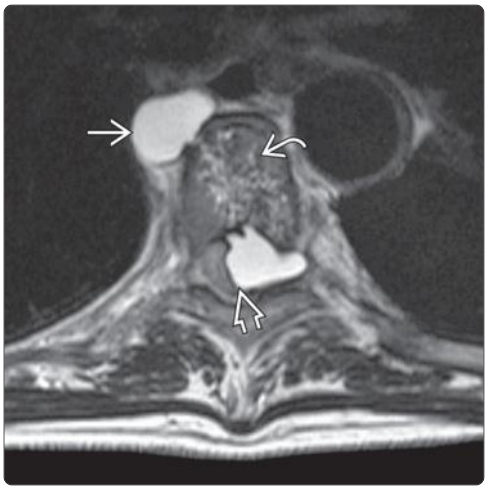
Image Interpretation Pearls

- Lack of significant enhancement is critical differential feature of echinococcal cysts

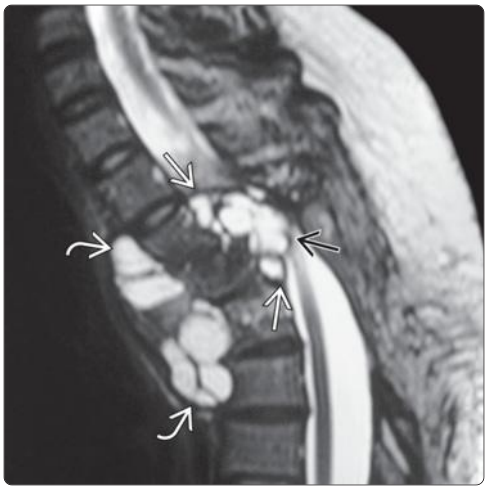
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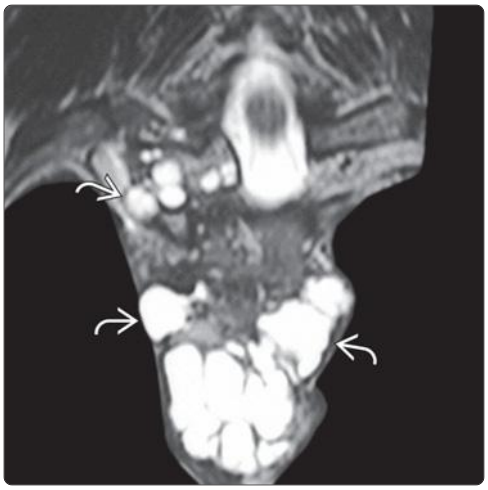
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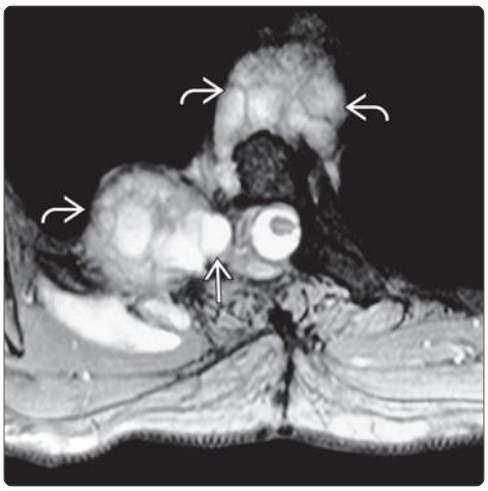


(Left) *Coronal T1WI C+ FS MR shows a peripherally enhancing spinal lesion. Recurrence is a predictor of poor prognosis, with rates of 30-100%. Up to 40% of recurrences occurred within 2 years of the initial operation* **(Right)** *Axial T2WI MR depicts echinococcal involvement of a midthoracic vertebral body. Anterior extension of the cyst results in a spherical cyst. A posterior cortical break into the epidural space causes marked compression of the cord. (Courtesy P. Rodriguez, MD.)*





(Left) *Sagittal T2WI MR exhibits a multicystic mass encircling the paravertebral thoracic spine. A cortical breach allows extension into the ventral epidural space with mass effect on the cord. The dura typically remains intact.* **(Right)** *Coronal T2WI MR illustrates an echinococcal lesion. Small loculi separate from the main cyst and form new cysts resulting in a mass of cysts. The disease starts in the vertebral body and may spread to the neural arch and adjacent rib.*





(Left) *Axial T2* GRE MR demonstrates a multiloculated extradural cystic mass. The mass extends into the epidural space through the right neural foramen.* **(Right)** *Sagittal T1WI MR shows a complex echinococcal mass involving multiple vertebral bodies and posterior elements, but sparing the disc spaces. Minimal host reaction in the bone results in a thin cyst wall. The cyst permeates within the bone and destroys the medullary cancellous bone. (Courtesy J. Beltran, MD.)*

Schistosomiasis

KEY FACTS

TERMINOLOGY

- CNS infection from parasitic trematodes (blood flukes) of genus *Schistosoma*

IMAGING

- Myelopathy, cord enhancement, edema in patient from endemic area
- Enlarged thoracic cord, conus medullaris
  - Diffuse increased signal within cord over several segments
  - Heterogeneous cord enhancement

TOP DIFFERENTIAL DIAGNOSES

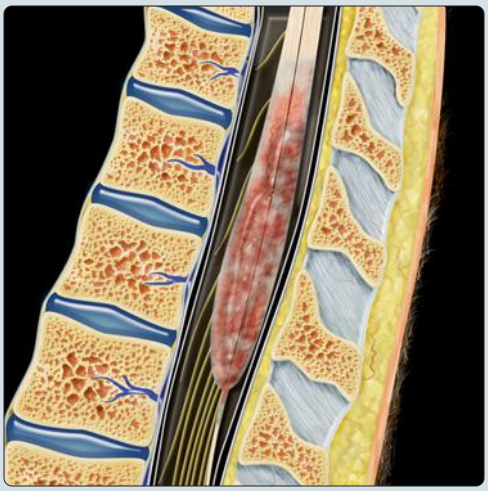
- Multiple sclerosis
- ADEM/viral myelitis
- Tuberculosis
- Intramedullary neoplasm
- Idiopathic acute transverse myelitis

PATHOLOGY

- Human schistosomiasis secondary to *Schistosoma haematobium* (*S. haematobium*), *Schistosoma mansoni* (*S. mansoni*), *Schistosoma japonicum* (*S. japonicum*)
- Early stage of disease more often produces CNS symptoms
- Chronic hepatosplenic, cardiopulmonary, urinary schistosomiasis usually asymptomatic

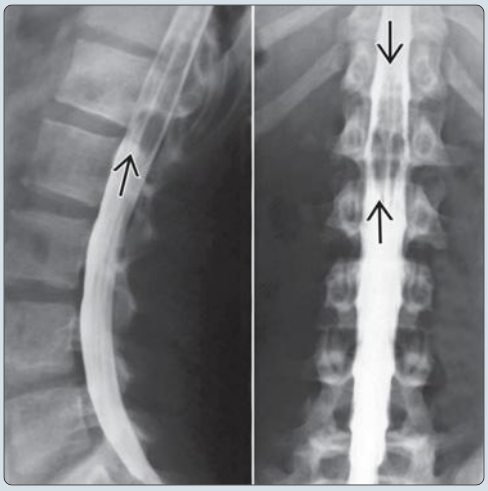
CLINICAL ISSUES

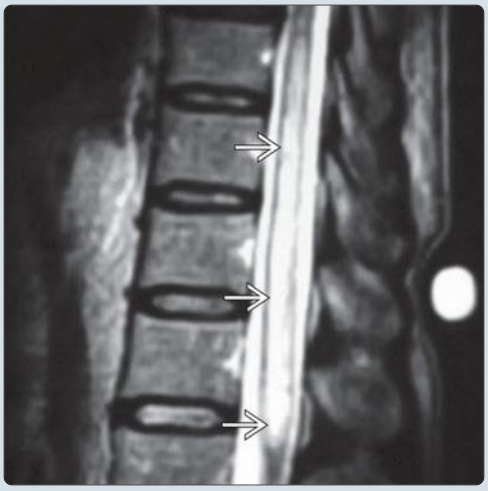
- Presence and clinical significance of neurological lesions depend on combination of presence of eggs and host immune response
- Schistosomal myelopathy tends to occur early after infection and is more likely to be symptomatic than cerebral schistosomiasis
- Conus medullaris and cauda equina are most common sites of involvement
  - Complete flaccid paraplegia with areflexia, sphincter dysfunction, and sensory disturbances





(Left) *Sagittal graphic shows schistosomiasis involving conus medullaris with granulomatous inflammatory response. Myelopathy, cord enhancement, and edema are diagnostic clues for NS in a patient from endemic area.* **(Right)** *Sagittal T1WI C+ MR shows enhancement of distal thoracic cord due to schistosomiasis. CSF examination shows inflammatory pattern with increased protein and mononuclear cells in 90% of cases ± IgG against schistosomal antigens. (Courtesy R. Mendonca, MD.)*





(Left) *Myelography reveals a filling defect in the thecal sac due to NS of the conus. (Courtesy Ahmed El-Razek, MD.)* **(Right)** *Sagittal STIR MR depicts diffuse hyperintensity in the distal cord and conus, which are mildly enlarged. Schistosomal myelopathy tends to occur early after infection and is more likely to be symptomatic (symptoms of flaccid paraplegia with areflexia, sphincter dysfunction, and sensory disturbances) compared to cerebral schistosomiasis. (Courtesy A. El-Razek, MD.)*

TERMINOLOGY

Abbreviations

- Neuroschistosomiasis (NS)

Synonyms

- Bilharziasis
- Katayama fever = acute schistosomiasis

Definitions

- CNS infection from parasitic trematodes (blood flukes) of genus *Schistosoma*
- Complex life cycle
  - Infect particular species of fresh water snails within endemic areas
  - Infected snails release free swimming larvae (cercariae), which attach to host mammal
  - Cercariae migrate through skin to dermal veins → pulmonary vasculature
  - Cercariae metamorphose to "schistosomula" → losing forked tails, developing lipid teguments, incorporating host proteins (major histocompatibility complexes and blood group antigens)
  - Migrate through lung capillaries → portal veins where they mature
  - Male and female pair off and migrate together, produce eggs
    - *Schistosoma mansoni*, *Schistosoma japonicum* → mesenteric veins
    - *Schistosoma haematobium* → vesicular (bladder) veins
  - Eggs are antigenic and induce inflammatory response
    - Migrate through bowel or bladder wall and shed in feces, urine
  - Shed eggs mature into free swimming "miracidia" → infect freshwater snail
    - 2 generations of "sporocysts" multiply in snail intermediate host

IMAGING

General Features

- Best diagnostic clue
  - Myelopathy, cord enhancement, edema in patient from endemic area
- Location
  - Thoracic cord, conus

CT Findings

- CECT
  - Diffuse cord enlargement and patchy enhancement

MR Findings

- T1WI
  - Enlarged cord/conus medullaris
- T2WI
  - Diffuse increased signal within cord over several segments
- T2* GRE
  - No gross hemorrhage
- T1WI C+
  - Heterogeneous cord enhancement
  - May extend over several segments

Imaging Recommendations

- Protocol advice
  - Sagittal, axial T1WI + T2WI, sagittal + axial post contrast

DIFFERENTIAL DIAGNOSIS

Multiple Sclerosis

- Central enhancement of peripheral cord lesions
- May show ring enhancement
- Presence of typical intracranial periventricular, subcallosal, brainstem, cerebellar white matter lesions

ADEM

- Diffuse cord enlargement, patchy multisegmental enhancement

Intramedullary Neoplasm

- No leptomeningeal enhancement
- Peritumoral edema
- Cystic and hemorrhagic components

Tuberculosis

- Vertebral body, epidural, leptomeningeal abnormalities

Pyogenic Abscess

- Focal, irregular focus of ring enhancement with edema

Viral Myelitis

- Long segmental cord enlargement and edema without focal lesions

Idiopathic Acute Transverse Myelitis

- Usually single lesion
- May be mono- or multisegmental

Sarcoidosis

- Cord and leptomeningeal enhancement
- Intracranial leptomeningeal enhancement, particularly midline about optic chiasm

PATHOLOGY

General Features

- Etiology
  - Early stage of disease more often produces CNS symptoms
    - 2 main pathways by which schistosomes cause NS
      - Egg embolism and worm migration, via either artery or vein system, especially valveless perivertebral Batson plexus
      - Anomalous migration of adult worms causes CNS infestation, with subsequent in situ egg deposition
  - Chronic hepatosplenic, cardiopulmonary, urinary schistosomiasis usually asymptomatic
    - Random, occasional embolization of eggs from portal mesenteric system to brain and cord
  - Vasculitis of immune etiology → may cause ischemic lesions, neurological symptoms
- Associated abnormalities
  - Egg retention and granuloma formation in bowel causes bloody diarrhea, cramping, inflammatory colon polyps
  - Urinary tract involvement leads to dysuria, hematuria, bladder polyps, ulcers, obstructive uropathy, increased risk of bladder squamous carcinoma

- Unshed eggs → portal circulation → hepatic perisinusoidal obstruction, portal hypertension, splenomegaly, ascites
- Presence of eggs in CNS induces cell-mediated periovular granulomatous reaction
- Spinal cord symptoms secondary to *S. mansoni* or *S. haematobium*
- Cerebral invasion secondary to *S. japonicum*
- Vascular obstruction secondary to granuloma formation with intense inflammatory reaction, ischemic necrosis

Staging, Grading, & Classification

- CSF bilharzia enzyme-linked immunosorbent assay (ELISA) test
  - Antibody testing cannot distinguish active infection from inactive infection
- Vestibular-evoked myogenic potential assesses vestibulospinal pathway
  - Abnormal in schistosomal myeloradiculopathy, alterations may precede image abnormalities
- Urine, stool analysis
- Colon biopsy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - CNS involvement frequently asymptomatic
    - 20-30% of patients with schistosomiasis have some CNS lesion
    - Presence and clinical significance of neurological lesions depend on combination of presence of eggs and host immune response
      - Majority → delayed immune response (type IV granulomatous), higher in beginning of schistosomiasis
      - Chronic forms → localized and lighter (immunological modulation phenomenon)
    - Myelopathy (acute transverse myelitis and subacute myeloradiculopathy) is most common neurological complication of *S. mansoni* infection
      - Schistosomal myelopathy tends to occur early after infection and is more likely to be symptomatic than cerebral schistosomiasis
      - Conus medullaris and cauda equina are most common sites of involvement
      - Severe schistosomal myelopathy → complete flaccid paraplegia with areflexia, sphincter dysfunction, and sensory disturbances
  - Fever, malaise, arthralgias, diarrhea
  - 3 clinical patterns of cord involvement
    - Myelitic
    - Granulomatous/pseudotumoral
    - Radicular/myeloradicular
  - Spine involvement generally as transverse myelitis pattern or spinal cord infarction
    - Transverse myelitis
    - Tetraparesis, paraparesis
    - Anterior spinal artery occlusion
    - Radiculopathy
  - Other signs/symptoms
    - Cerebral and cerebellar tumor-like neuroschistosomiasis → increased intracranial pressure, headache, nausea and vomiting, and seizures
    - Bowel, bladder dysfunction
    - Portal hypertension
- Clinical profile
  - CSF pleocytosis, elevated protein content, oligoclonal IgG bands
  - Stool smear examination with quantitative Kato-Katz oogram if suspected schistosomiasis

Demographics

- Age
  - Any age (peak in 2nd decade)
- Gender
  - M > F
- Ethnicity
  - Infection related to location of endemic schistosomiasis
- Epidemiology
  - Infection affects 200 million individuals in 74 countries of Latin America, Africa, and Asia
    - 2nd to malaria in worldwide socioeconomic impact
  - Man, other mammals as definitive hosts; aquatic and amphibious snails as intermediate hosts
  - Prevalence of CNS involvement varies from 1-30% of infected individuals
  - Neuroschistosomiasis 3rd most common cause of myelopathy after trauma and tumor involvement

Natural History & Prognosis

- Most patients improve with appropriate antihelmintic drug treatment
- Acute schistosomiasis: Up to 25% mortality
- End-stage hepatosplenic disease, varices, pulmonary hypertension, CNS disease associated with high mortality

Treatment

- Surgical biopsy may be required
- Oral treatment with antihelmintics: Praziquantel and oxamniquine
- Steroids to decrease inflammatory response
- High cervical electrical stimulation may improve chronic pain arising from spinal cord injury due to NS
- Rehabilitation and multidisciplinary team care

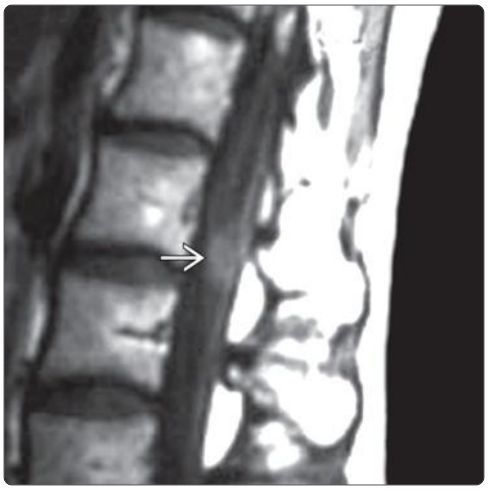
DIAGNOSTIC CHECKLIST

Consider

- Patients with spinal involvement may have no other clinical evidence of schistosomiasis

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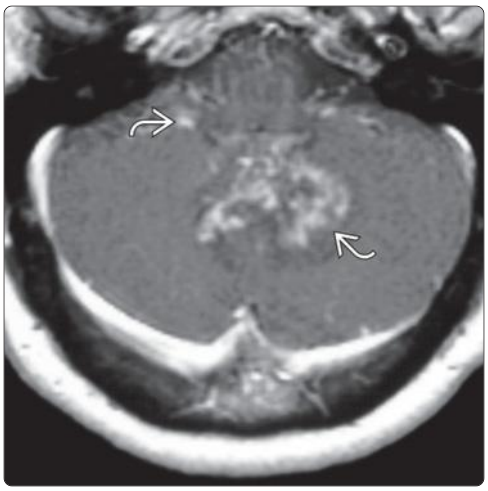


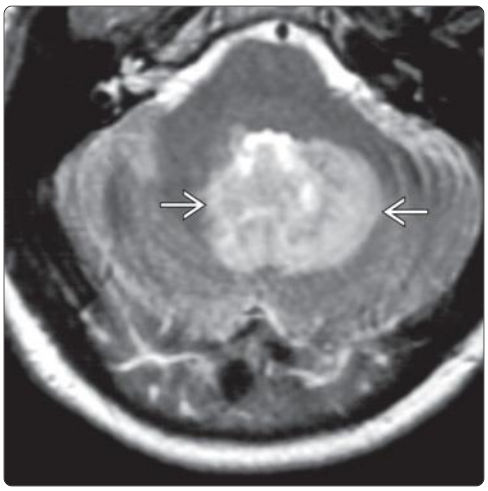
(Left) *Sagittal T1WI C+ MR shows ovoid cauda equina lesion due to schistosomiasis granulomatous inflammatory mass. NS presents as a low cord syndrome of acute/subacute progression. Lower limb pain, weakness, sensory disturbance, autonomic dysfunctions, and impotence in men are often present.* **(Right)** *Sagittal T1WI C+ MR illustrates patchy conus enhancement due to schistosomiasis infection. (Courtesy A. Aul, MD.)*





(Left) *Sagittal T1WI MR reveals mild diffuse cord/conus enlargement with schistosomiasis infection. (Courtesy A. Aul, MD.)* **(Right)** *Sagittal T2WI MR illustrates diffuse patchy T2 hyperintensity throughout distal thoracic cord and conus in a patient with schistosomiasis. Vascular obstruction secondary to granuloma formation results in an intense inflammatory reaction and ischemic necrosis.*





(Left) *Axial T1WI C+ MR demonstrates irregular punctate enhancement due to schistosomiasis. Cerebral NS presents as a slowly expanding intracranial tumor-like lesion. Its clinical manifestations are variable, depending on the site of the lesion and increased intracranial pressure.* **(Right)** *Axial T2WI MR shows hyperintense regions in the medial cerebellar hemispheres related to edema from cerebellar tumor-like involvement with schistosomiasis.*

KEY FACTS

TERMINOLOGY

- CNS parasitic infection caused by pork tapeworm, *Taenia solium*
- Spinal extraparenchymal form uncommon (5% of cases)
  - Often accompanied by intracranial disease
  - Divided in intramedullary, subarachnoid, or extradural in location
  - Subarachnoid form most common spinal type
  - Spinal thoracic involvement in 60-75%

IMAGING

- Intradural cyst with evidence of similar lesions in brain (cyst with "dot" appearance)
- CT: Focal cystic low attenuation, subarachnoid or intramedullary, cord enlargement ± enhancement
- MR
  - Subarachnoid: CSF signal cystic lesions, variable mass effect on adjacent cord/cauda equina
  - Intramedullary: Focal cystic lesion(s), ± syrinx cavitation, cord edema extending over several segments
  - ± peripheral cyst enhancement

PATHOLOGY

- Caused by larval form of pig tapeworm, *T. solium*
- Man intermediate host in tapeworm life cycle

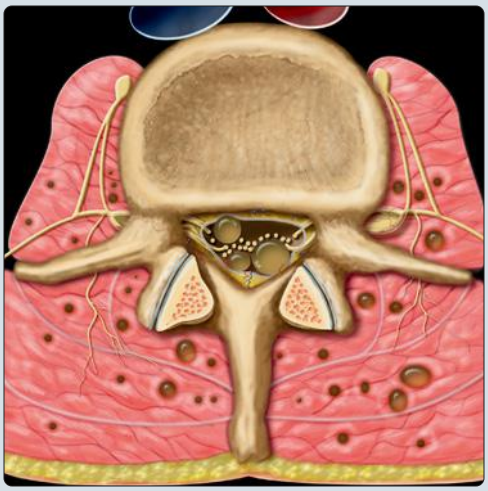
CLINICAL ISSUES

- Symptoms due to mechanical compression, inflammatory changes and cord edema, gliosis due to vasculopathy, pachymeningitis, and syrinx formation
- Medical therapy in selected patients in whom symptoms are stable and nonprogressive
- Surgical intervention when severe or progressive deficits occur to prevent permanent injury

DIAGNOSTIC CHECKLIST

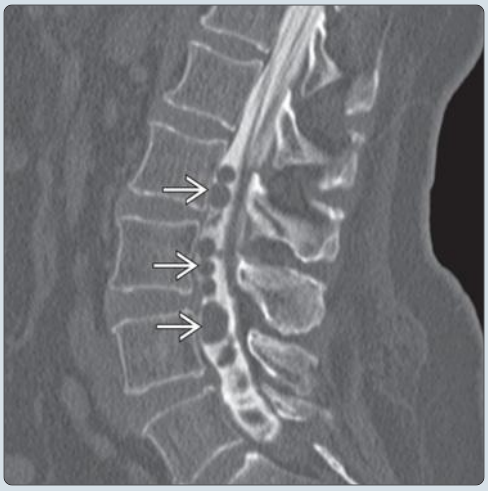
- MR is investigative and follow-up modality of choice

(Left) *Axial graphic shows multiple subarachnoid cysts (cysticerci) distorting nerve roots, with multiple additional lesions within muscles.* **(Right)** *Axial CT myelogram illustrates multiple cysts ➡ of various sizes within the lumbar subarachnoid space in this patient with known neurocysticercosis. Most subarachnoid SCCs are associated with intracranial lesions, spreading by the ventriculo-ependymal route. Rare reports exist of primary infection through hemopoietic spread or direct larval migration.*





(Left) *Sagittal CT myelogram demonstrates multiple subarachnoid cysts ➡ from cysticercosis. These subarachnoid cysts are hypodense on NECT. SCC occurs in the subarachnoid space where mass effect can cause spinal cord compression, although obstruction of cerebrospinal fluid pathways due to scarring of the subarachnoid space can also cause symptoms.* **(Right)** *Sagittal T2WI MR exhibits an intradural extramedullary cyst in the ventral thecal sac ➡, causing mass effect on the cervicothoracic cord ➡.*





TERMINOLOGY

Abbreviations

- Neurocysticercosis (NCC), spinal cysticercosis (SCC)

Definitions

- CNS parasitic infection caused by pork tapeworm
- 2 forms of NCC: Parenchymal and extraparenchymal
  - SCC: Uncommon form of extraparenchymal NCC (5% of cases of NCC)
    - Divided in intramedullary, subarachnoid, or extradural in location
    - Subarachnoid form most common spinal type

IMAGING

General Features

- Best diagnostic clue
  - Intradural cyst with evidence of similar lesions in brain (cyst with "dot" appearance)
- Location
  - Parenchymal, leptomeningeal, intraventricular, spinal
    - SCC rare
- Size
  - Cyst size variable (5-20 mm)
    - Cysts contain scolex, which is 1-4 mm
- Morphology
  - Round or ovoid cyst

Radiographic Findings

- Radiography
  - May show soft tissue calcifications

CT Findings

- CECT
  - Vesicular (antigenically nonviable): Small and rounded low-density areas
    - No perilesional edema and enhancement
    - Eccentric hyperdense nodule = scolex
  - Colloidal (acute encephalitic phase): Ill-defined lesions surrounded by edema due to release of metabolic products, ring pattern of enhancement
  - Granular nodular: Lesion starts to mineralize, nodular hyperdense lesions surrounded by edema
  - Final involution: Calcified cysticerci → small hyperdense nodules without perilesional edema or abnormal enhancement

MR Findings

- T1WI
  - Subarachnoid: CSF signal cystic lesions, variable mass effect on adjacent cord/cauda equina
  - Intramedullary: Focal cystic lesion(s), ± syrinx cavitation
- T2WI
  - Subarachnoid: CSF signal cystic lesions
  - Intramedullary: Focal cystic lesion showing ↑ T2 signal, cord edema extending over several segments
- DWI
  - Scolex hyperintense within dark cyst probably caused by water restriction
  - High signal intensity on DWI in colloid stage of NCC, resulting from water-restricted products of parasite degradation and host immune response
- T1WI C+
  - Subarachnoid: Peripheral cyst enhancement
  - Intramedullary: Peripheral enhancement of cyst
    - May show only nonspecific sheet-like enhancement of subarachnoid space, cord pial surface
- Vesicular (antigenically nonviable): CSF signal intensity on T1WI and T2WI MR
  - Scolex is high intensity nodule within cyst: "Hole-with-dot" pattern
- Colloidal: Thick, hypointense cyst walls, varied perilesional edema
- Granular nodular: Areas of signal void on both T1WI and T2WI MR surrounded by edema or gliosis
- Final involution: Calcified cysticerci → small hypointense areas

Nonvascular Interventions

- Myelography
  - Variety of appearances depending upon region involved
    - Intradural extramedullary lesions with subarachnoid cysts
    - Enlarged cord ± myelographic block with intramedullary disease

Imaging Recommendations

- Best imaging tool
  - MR imaging with contrast defines intradural extramedullary, intramedullary cysts, associated edema
- Protocol advice
  - Sagittal, axial T1WI, T2WI, T1 C+; include brain imaging in addition to spine imaging

DIFFERENTIAL DIAGNOSIS

Pyogenic Abscess

- Irregular ring enhancement with cord edema, expansion

Osteomyelitis, Granulomatous

- Tuberculosis, sarcoidosis
- Generally leptomeningeal involvement

Neoplasm

- Primary
- Metastatic
- Irregular solid or ring enhancement
- Associated cysts with astrocytoma, ependymoma, hemangioblastoma

Arachnoid Cyst

- CSF signal/attenuation, no enhancement

Other Parasitic Infections

- May have cystic component (echinococcosis)
- May show myelitis pattern without focal cyst (schistosomiasis)

Syrinx

- Nonenhancing central cord linear CSF signal intensity associated with Chiari 1

Schwannoma

- Intradural extramedullary mass
- May show cystic degeneration with thin rim enhancement

PATHOLOGY

General Features

- Etiology
  - Caused by larval form of pig tapeworm, *Taenia solium*
  - Man intermediate host in tapeworm life cycle
    - Ova or pregnant proglottids in contaminated water, food: Fecal-oral route
    - Ova are digested in GI tract → release oncospheres that penetrate intestinal wall → bloodstream
    - Oncospheres develop into cysticerci in any organ → cysticerci lodge permanently in tissue
  - Downward migration of larvae from cerebral to spinal subarachnoid space → leptomeningeal form
  - Hematogenous dissemination, spread through ventriculo-ependymal pathways → intramedullary
  - Subarachnoid cysticercosis caused by 2 different larval types
    - *Cysticercus cellulosae* → larval worm with simple bladder and scolex
    - *Cysticercus racemosus* → grape-like cluster of larvae (without scolex) → chronic granulomatous meningitis
- Most frequently affected site is CNS (70-80%)
  - Parasites may lodge in brain parenchyma, subarachnoid space, and ventricular system, or less commonly spine (1-6% of cases)
  - Incidence of spinal ≤ brain, as blood flow to brain is ~ 100x greater than to spine
- SCC: Extraspinal or intraspinal
  - Intraspinal: Epidural, subarachnoid (leptomeningeal), and intramedullary
    - Intramedullary lesions ≤ 20% of SCC cases
      - Spinal distribution: Cervical (34%), thoracic (44.5%), lumbar (15.5%), sacral region (6%)

Gross Pathologic & Surgical Features

- Thickened, opaque arachnoid, pachymeningitis
- Subarachnoid or intramedullary cysts with cleavage plane from normal tissue; surrounding soft yellowish granulation tissue

Microscopic Features

- Cyst wall has 3 layers: Outer (cuticular layer), middle cellular (pseudoepithelial) layer, inner reticular (fibrillary) layer
- Inflammatory reaction with edema, adjacent gliosis
- Adjacent acute, chronic inflammatory cell infiltrate causing "cysticercal abscess," arachnoiditis
- Cyst wall degeneration with calcification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Commonly asymptomatic
  - Spine symptoms
    - Chronic myelopathy, sensory level, back pain
    - Spastic para-/quadriparesis, flaccid paralysis
    - Bladder dysfunction
- Clinical profile
  - Diagnosis confirmed by ELISA of serum or CSF
    - > 70% sensitivity, highly specific for cysticercal antigens; sensitivity ↑ if assay performed on CSF
    - Increased sensitivity as number of lesion increase if assay performed on CSF
    - 2/3 of spinal NCC reported with negative ELISA
    - Enzyme-linked immunoelectrotransfer blot (EITB) reportedly > 90% sensitivity
  - Peripheral blood eosinophilia (25%)
  - CSF: Nonspecific protein elevation, lymphocytosis
  - Stool specimens occasionally may be positive for *T. solium* ova or proglottids

Demographics

- Ethnicity
  - In USA, Hispanic and Asian patients common → endemic area immigration
- Epidemiology
  - Cysticercosis most common worldwide parasitic infection
    - In endemic regions, incidence of NCC approaches 4% of general population
    - Endemic in Mexico, Central and South America, Eastern Europe, Asia, India, Africa

Natural History & Prognosis

- Parenchymal gliosis, pachyleptomeningitis may cause cord vascular compromise

Treatment

- Options, risks, complications
  - Surgical intervention when severe or progressive deficits occur to prevent permanent injury
    - Recovery may be limited as result of inflammatory injury to spinal cord or arachnoidal adhesions
  - Medical therapy in selected patients in whom symptoms are stable and nonprogressive
    - Steroids to decrease inflammation
    - Oral treatment with praziquantel or albendazole
  - Postoperative course of anticysticercal necessary since generalized disease with focal spinal manifestation

DIAGNOSTIC CHECKLIST

Consider

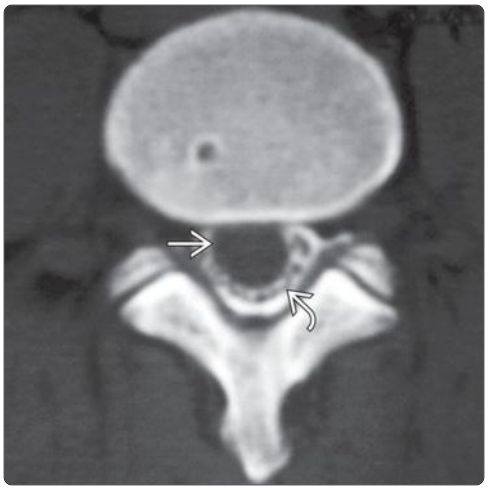
- SCC often accompanied by intracranial disease

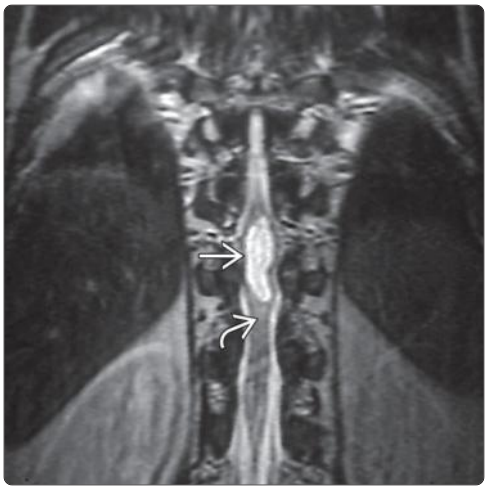
Image Interpretation Pearls

- Only spinal findings may be chronic inflammatory arachnoiditis → altered CSF flow dynamics → syringomyelia

SELECTED REFERENCES

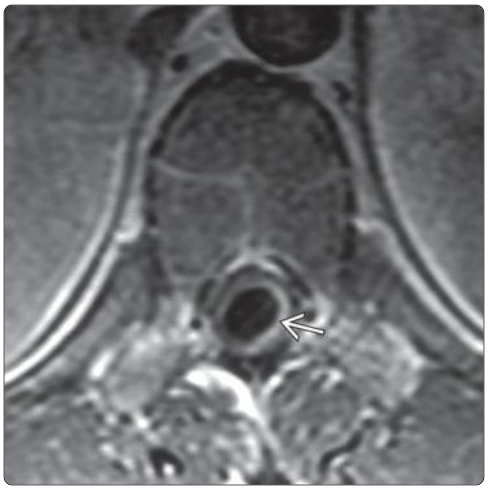
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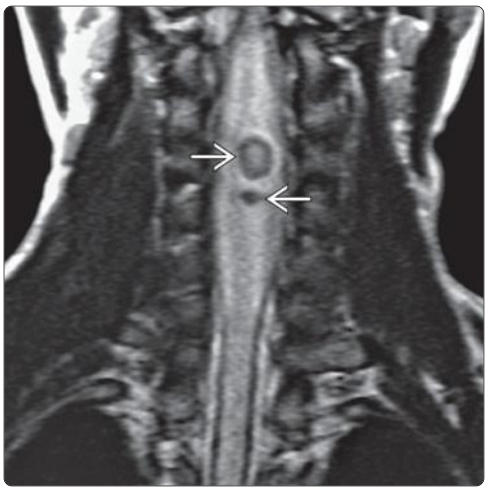
(Left) *Axial CT myelogram shows a low-density intrathecal filling defect due to a cysticercosis cyst. It displaces the cauda equina nerve roots. (Courtesy P. Rodriguez, MD.)* **(Right)** *Coronal T2WI MR illustrates a focal cystic mass involving distal thoracic cord, with fusiform cord expansion. The cord cyst is hyperintense with more diffuse edema within the cord proximal and distal to the cyst. The differential of the cord lesion would be primarily intramedullary tumor.*





(Left) *Sagittal T1WI C+ MR demonstrates focal ring-enhancing cystic mass in distal thoracic cord, with fusiform cord expansion. Intramedullary lesions comprise up to 20% of SCC cases. Hematogenous dissemination is most probable route of intramedullary SCC.* **(Right)** *Axial T1WI C+ MR depicts an intramedullary peripherally enhancing cysticercosis cyst in the distal thoracic cord. It causes fusiform cord expansion.*





(Left) *Sagittal T2WI MR exhibits diffuse cord expansion and hyperintensity of the cervical cord. There are 2 intramedullary foci of decreased signal. Extensive edema is seen in the colloidal and granular nodular stages due to release of metabolic products with thickening and hypointensity of the cyst wall.* **(Right)** *Coronal T2WI MR shows intramedullary cysticercosis involvement with marked edema. Fusiform cord expansion at the site of the lesions is observed.*

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PART IV
SECTION 2

Inflammatory and Autoimmune Disorders



Acute Transverse Myelopathy	**644**
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Acute Transverse Myelopathy

KEY FACTS

TERMINOLOGY

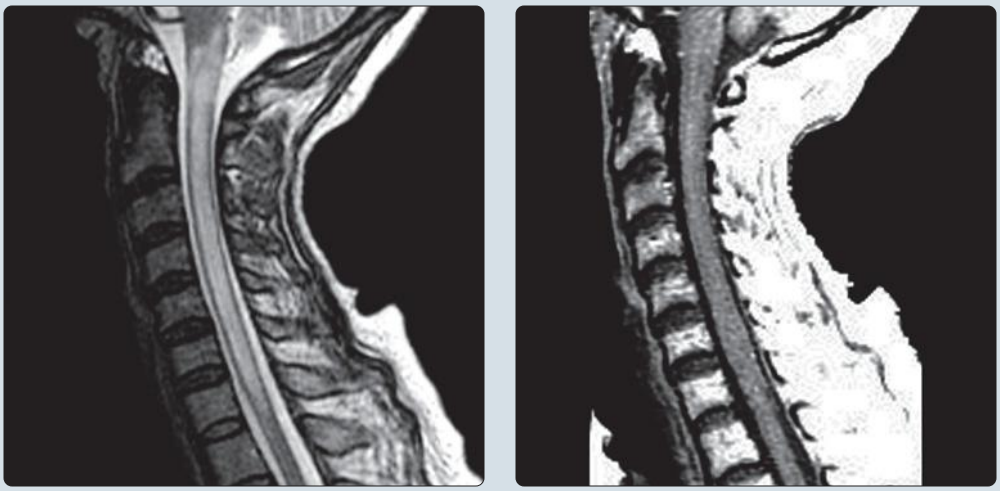
- Noncompressive myelopathy
- Acute transverse myelitis (ATM) is a broad generic term encompassing heterogeneous group of disorders causing cord dysfunction
  - Etiologies include inflammatory causes (ATM) + vascular disease + radiation + paraneoplastic + idiopathic (unknown)
  - ATM is a more specific term for cord dysfunction that is inflammatory in etiology
    - Inflammation defined by CSF findings and MR enhancement

IMAGING

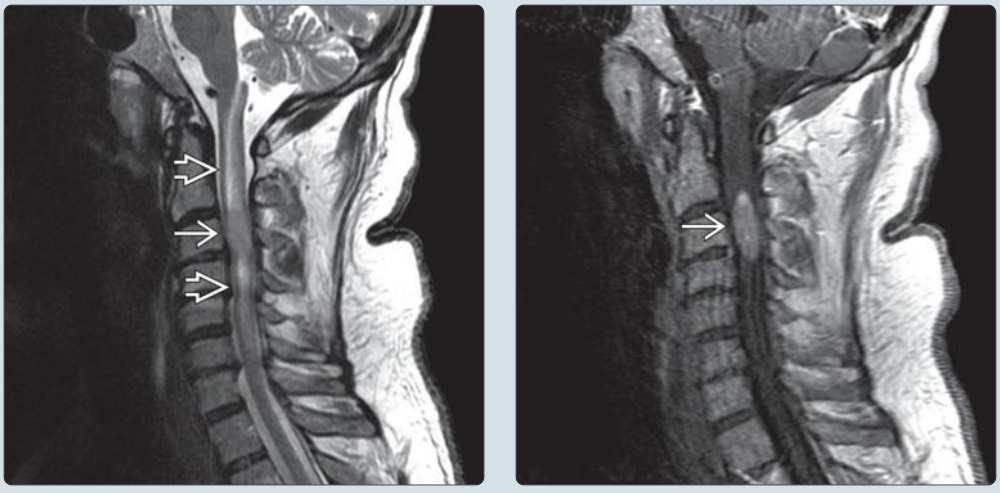
- Hyperintense lesion on T2WI with mild cord expansion ± enhancement
- Longitudinally extensive = NMO, ADEM, viral infection

TOP DIFFERENTIAL DIAGNOSES

- Inflammatory Etiologies
  - Acquired demyelinating
    - Multiple sclerosis (MS)
    - Neuromyelitis optica (NMO)
    - Acute disseminated encephalomyelitis (ADEM)
  - Parainfectious
  - Systemic autoimmune
  - Paraneoplastic (antineuronal immune disorder associated with cancer)
- Noninflammatory mimics
  - Radiation
  - Metabolic
  - Tumor and cysts
  - Vascular
    - Cord infarction
    - Dural fistula (type I AVF)
- Idiopathic ATM



(Left) *Sagittal T2WI MR shows longitudinally extensive T2 hyperintensity throughout the cervical cord and lower medulla. Differential diagnoses include NMO, ADEM, parainfectious and systemic autoimmune disorders.* **(Right)** *Sagittal T1 C+ MR in the same patient does not demonstrate significant post-gadolinium enhancement. The presence of longitudinally extensive abnormality in this patient suggests MS is not the underlying abnormality.*



(Left) *Sagittal T2 TSE MR in this patient with lung cancer shows a focal mass at the C3 level ⇨ with adjacent cord edema ⇨ from a focal cord parenchymal metastasis.* **(Right)** *Sagittal T1 C+ MR shows enhancement of focal cord metastasis ⇨ in this patient with lung carcinoma and worsening myelopathy.*

TERMINOLOGY

Abbreviations

- Acute transverse myelitis (ATM)
- Multiple sclerosis (MS)
- Neuromyelitis optica (NMO)

Synonyms

- Acute noncompressive myelopathies

Definitions

- Acute transverse myelopathy is a broad generic term encompassing heterogeneous group of disorders causing cord dysfunction
  - Etiologies include inflammatory causes (ATM) + vascular disease + radiation + paraneoplastic + idiopathic (unknown)
  - ATM more specific term for cord dysfunction that is inflammatory in etiology
    - Inflammation defined by CSF findings and MR enhancement

IMAGING

General Features

- Best diagnostic clue
  - Hyperintense lesion on T2WI with mild cord expansion
  - Enhancement suggests inflammatory etiology; thus it is transverse myelitis
- Location
  - Any cord level
- Size
  - Segmental, multisegmental, or holocord
  - Less than 2 vertebral segments suggests MS
  - Longitudinally extensive signal change associated with NMO
- Morphology
  - Fairly well-defined signal abnormality

MR Findings

- T1WI
  - Normal
  - Focal or diffuse hypointensity
  - Mild hyperintensity due to petechial hemorrhage (rare)
- T2WI
  - Focal or diffuse hyperintensity
  - Swollen cord
- STIR
  - Hyperintense
- T1WI C+
  - Variable enhancement
    - None to focal, diffuse, or heterogeneous
    - May be mass-like, or peripheral/meningeal
- Cord swelling and enhancement typically resolve if related to demyelinating disease
  - Corresponding to clinical improvement
    - Cord atrophy may be present in chronic stage

Nonvascular Interventions

- Myelography
  - Nonspecific cord swelling

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI and T2WI MR with contrast
    - 40% of clinical ATM with normal MR findings

DIFFERENTIAL DIAGNOSIS

Inflammatory Etiologies

- Acquired demyelinating
  - MS
    - Difficult to distinguish from idiopathic ATM by cord imaging alone
      - Lesion peripheral in location
      - Less than 2 vertebral segments in length
      - Less than half cross sectional area of cord
      - Homogenous or ring enhancement in acute or subacute phase
    - MS following ATM: 2-8%
    - 90% with associated intracranial lesions
    - Relapsing remitting clinical course
  - NMO
    - Longitudinally extensive intramedullary hyperintensity on T2WI
    - Cord swelling
    - Bilateral retrobulbar optic neuritis preceding ATM
      - ATM as initial presentation in 20% of cases
    - Little clinical involvement beyond spinal cord and optic nerves
    - Positive serologic test for NMO-IgG
  - Acute disseminated encephalomyelitis (ADEM)
    - May be indistinguishable from cord MS
    - Thalamic involvement more common intracranially
    - Younger age at presentation
    - Constitutional symptoms
    - Monophasic clinical course
- Parainfectious
  - Viral, bacterial, fungal, or parasitic
- Systemic autoimmune
  - Sjögren syndrome
  - SLE
  - Antiphospholipid syndrome
  - Behcet disease
  - Mixed connective tissue disease
- Paraneoplastic (antineuronal immune disorder associated with cancer)
  - Multiple antibodies documented: Often small cell lung carcinoma
    - Antibodies present in approximately 60% of patients with paraneoplastic disease of central nervous system (CNS)
      - Anti-Hu (ANNA 1), anti-Ri (ANNA 2), anti-Yo (PCA1), Amphiphysin-IgG
      - CRMP-5-IgG, anti-GAD6, NMDAR
- Toxic
  - Cassava toxicity
  - Lathyrism
  - Hepatic myelopathy

Noninflammatory Mimics

- Radiation

- Metabolic
  - Vitamin B12 deficiency
    - Mild cord enlargement
    - Signal abnormality in dorsal ± lateral columns
      - Hypointense on T1WI and hyperintense on T2WI
    - No enhancement
    - Macrocytic anemia with decreased plasma B12 level
- Tumor and cysts
  - Syringohydromyelia
    - Central cystic lesion
    - Cerebral spinal fluid intensity on all pulse sequences
    - No post-gadolinium enhancement
  - Cord tumor
    - Diffuse or nodular contrast enhancement
    - Extensive peritumoral edema
    - Cystic ± hemorrhagic components
- Vascular
  - Cord infarction
    - Immediate onset: Minutes, rather than hours or days
    - Ventral cord location with little initial mass effect
    - Motor sign predominate
    - No enhancement in acute phase
  - Dural fistula (type I AVF)
    - Chronic course in elderly patients
    - Prominent vascular flow voids on MR along cord surface
    - Most are distal thoracic or conus lesions
    - Dynamic enhanced spinal MRA to diagnose

Idiopathic ATM

- Diagnosis of exclusion
  - Lesion centrally located
  - Variable enhancement
- No associated intracranial lesions

PATHOLOGY

General Features

- Etiology
  - Collagen vascular disease: ATM may be the initial manifestation
    - Autoimmune vasculitis
      - SLE
      - Antiphospholipid syndrome
      - Sjögren disease, scleroderma
  - Infectious: Typically viral
    - Direct cord involvement ± autoimmune phenomenon
    - Epstein-Barr virus, cytomegalovirus, herpes simplex virus, varicella, or enteroviruses
    - Nonviral pathogens: Syphilis, Lyme disease, mycoplasma pneumoniae, or schistosomiasis
  - Postvaccination
    - Autoimmune response with small vessel vasculopathy
    - Polio, rubella, rabies, smallpox, influenza, or hepatitis B vaccinations
  - Postirradiation
    - Dose of 45-50 Gy in daily 1.8-2 Gy fractions considered safe
    - Symptoms develop 6-30 months after therapeutic irradiation
  - Spinal dural fistula (AVF)
    - Course more indolent compared to other etiologies
  - Paraneoplastic syndrome
    - May be initial presentation of underlying neoplasm
    - Lung, breast, hepatocellular carcinoma, or lymphoma
- Genetics
  - No genetic predilection
- Epidemiology
  - 4.6 new cases of transverse myelitis per million people per year in US

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sensory deficit
    - Loss of pain and temperature sensation
    - Well-defined upper level
    - Ascending paresthesia in bilateral upper/lower extremities
  - Other signs/symptoms
    - Back ± radicular pain
    - Paraplegia/quadriplegia
    - Bladder and bowel urgency, frequency, and retention
- Clinical profile
  - Progression to severe neurologic deficits within days

Demographics

- Age
  - All ages can be affected
- Gender
  - Female predominance in collagen vascular disease

Natural History & Prognosis

- 30-50% with complete recovery after treatment

Treatment

- Collagen vascular disease
  - Corticosteroids
  - Immunosuppressive therapy
  - Plasmapheresis, intravenous immunoglobulins (IVIg)
- Infectious etiologies
  - Steroids, antiviral, or antibacterial medications
- Postirradiation myelitis
  - Hyperbaric oxygen treatment

DIAGNOSTIC CHECKLIST

Consider

- Brain MR imaging to exclude intracranial lesions associated with MS, ADEM, and NMO

SELECTED REFERENCES

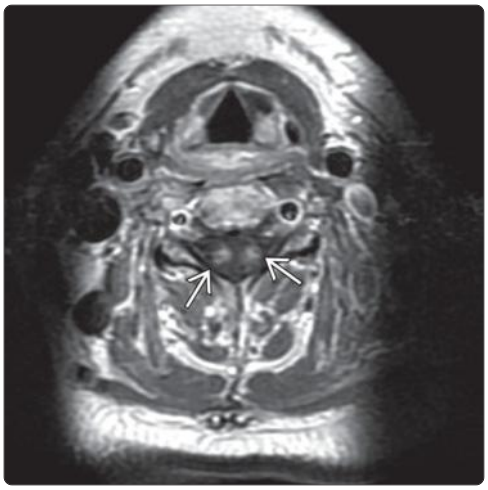
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(Left) *Sagittal T2WI MR with fat suppression in this young patient with rapid onset myelopathy after recent viral infection shows patchy hyperintensities in ventral lower thoracic cord, consistent with ATM.* **(Right)** *Sagittal STIR MR in this 25 year old shows increased signal within the cord spanning the C6 and C7 levels, with an additional lesion in the upper thoracic cord. Findings are suggestive of MS, given the short segment of involvement and multiplicity of lesions.*





(Left) *Sagittal T2 FS MR in this patient with multiple bone metastases and prior radiation therapy shows diffuse increased signal in the cord. Primary considerations would be radiation myelopathy vs. paraneoplastic myelopathy.* **(Right)** *Axial T1 C+ MR shows focal enhancement of the central gray matter of the spinal cord in this patient with extensive bony metastatic disease. This pattern is suggestive of paraneoplastic myelopathy.*





(Left) *Sagittal STIR MR in a patient with chronic cervical myelopathy shows fusiform hyperintense cord expansion from C4-C6 with multiple superficial flow voids, compatible with arteriovenous malformation.* **(Right)** *Sagittal T2WI MR in a patient with a history of head and neck cancer treated with radiotherapy demonstrates postirradiation myelopathy with diffuse hyperintensity and cord swelling from C2-C6.*

Idiopathic Acute Transverse Myelitis

KEY FACTS

TERMINOLOGY

- Inflammatory disorder involving both halves of spinal cord, resulting in bilateral motor, sensory, and autonomic dysfunction

IMAGING

- Central cord expansile lesion > 2 vertebral segments in length ± variable eccentric enhancement

TOP DIFFERENTIAL DIAGNOSES

- Multiple sclerosis
- Neuromyelitis optica
- Spinal cord neoplasm
- Spinal cord infarction

PATHOLOGY

- Autoimmune phenomenon with formation of antigen-antibody complexes
- Small vessel vasculopathy → cord ischemia
- Associated demyelinating process

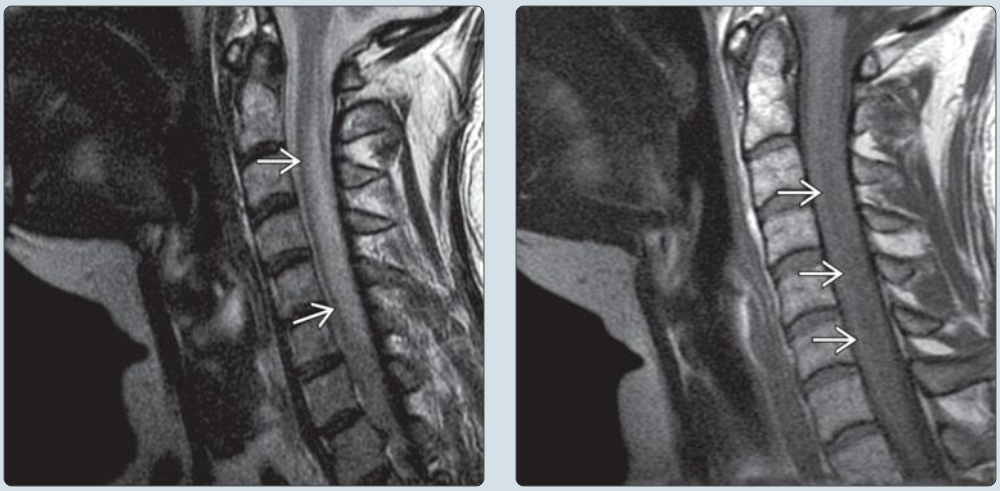
CLINICAL ISSUES

- Inclusion criteria
  - Sensory, motor, or autonomic dysfunction attributable to spinal cord
  - Bilateral signs/symptoms with well-defined sensory level
  - Compressive etiology excluded by neuroimaging
  - Cord inflammation confirmed by CSF pleocytosis, elevated IgG index, or gadolinium enhancement
- Exclusion criteria
  - History of spinal radiation within past 10 years
  - Cord ischemia/infarction, arteriovenous malformation
  - Connective tissue diseases
  - CNS infections, sarcoidosis, SLE, or Sjögren syndrome

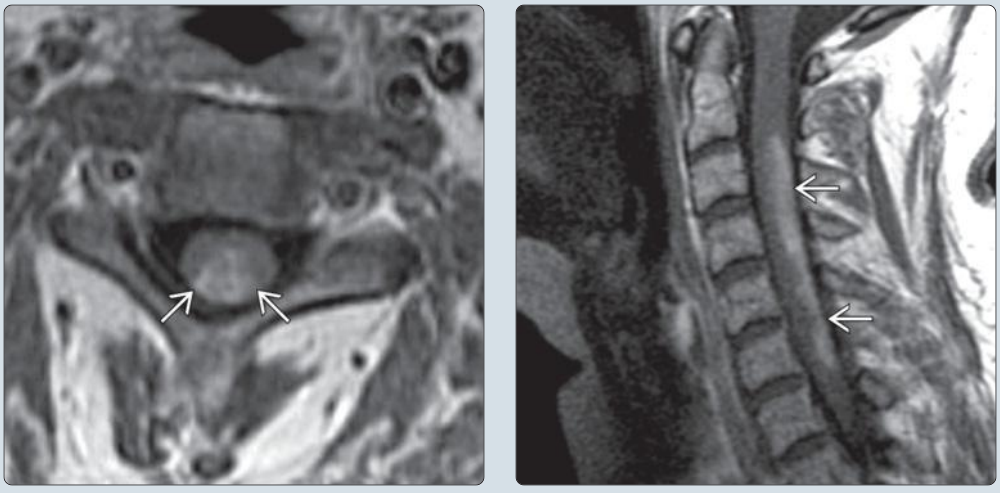
DIAGNOSTIC CHECKLIST

- Central cord lesion > 2 vertebra in length, eccentric enhancement → consider ATM

(Left) *Sagittal T2WI MR demonstrates mild, multilevel cervical cord expansion with confluent intramedullary T2 hyperintensity ➡ extending over 5 levels.* **(Right)** *Sagittal T1WI MR (same patient) reveals mild cervical cord expansion with subtle central intramedullary hypointensity ➡. Lesion extent on T1 MR is usually less conspicuous than on T2 MR.*



(Left) *Axial T1WI C+ MR (same patient) illustrates patchy, bilateral, central heterogeneous cervical spinal cord enhancement ➡ involving both gray and white matter.* **(Right)** *Sagittal T1WI C+ MR (same patient) confirms extensive parenchymal enhancement within the abnormal dorsal cervical spinal cord ➡.*



TERMINOLOGY

Abbreviations

- Idiopathic acute transverse myelitis (IATM)

Synonyms

- Idiopathic transverse myelopathy

Definitions

- Inflammatory disorder involving both halves of spinal cord, resulting in bilateral motor, sensory, and autonomic dysfunction

IMAGING

General Features

- Best diagnostic clue
  - Central cord lesion > 2 vertebral segments in length with eccentric enhancement
- Location
  - Thoracic most common
  - 10% in cervical cord
  - Central cord location on axial imaging
- Size
  - > 2 vertebral segments in length
    - Commonly 3-4 segments
  - > 2/3 of cross-sectional area of cord on axial imaging
- Morphology
  - Well circumscribed

MR Findings

- T1WI
  - Smooth cord expansion in acute phase
    - Less extensive than T2 signal abnormality
  - Iso- to hypointense
  - Cord atrophy in late stage
- T2WI
  - Hyperintense cord expansion
  - Central "dot" sign
    - Central gray matter surrounded by edema
- STIR
  - Hyperintense cord expansion
- DWI
  - Significantly ↓ fractional anisotropy (FA) in lesions and distal (normal-appearing) spinal cord
  - FA ↓ in distal normal-appearing spinal cord might be related to clinical outcome
- T1WI C+
  - Variable appearance post-gadolinium enhancement
    - No enhancement in up to 40-50% cases
    - Enhancement more common when cord enlargement present
      - More frequent in subacute than acute or chronic stages
        - Resolves over time
      - Enhancing area less extensive than T2 hyperintensity
    - Solitary or multifocal lesions
      - Focal, nodular, subtle, or patchy enhancement
      - Peripheral lesion enhancement
    - ± meningeal enhancement
  - Enhancement not predictive of clinical course

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI, T1WI C+ MR

DIFFERENTIAL DIAGNOSIS

Multiple Sclerosis

- < 2 vertebral segments in length, peripheral location
- < 1/2 of cord cross-sectional area
- 90% have associated intracranial lesions
- Relapsing and remitting clinical course

Neuromyelitis Optica

- Autoimmune inflammatory disorder involving neuronal myelin of optic nerves and spinal cord
- Extensive (> 3 vertebral segments) cord T2 hyperintensity + optic nerve enhancement
- Limited brain involvement

Spinal Cord Neoplasm

- Cord expansion and extensive peritumoral edema invariably present
- Cystic ± hemorrhage components
- Diffuse or nodular contrast enhancement
- Slower clinical progression

Spinal Cord Infarction

- Ventral or central cord location, less mass effect initially
- Patients present more often with motor signs than sensory signs
- Immediate onset (minutes, rather than hours, days)

Spinal Dural Fistula

- Increased T2 cord signal
- Prominent flow voids (dilated veins) on cord surface

PATHOLOGY

General Features

- Etiology
  - Autoimmune phenomenon with formation of antigen-antibody complexes
  - Small vessel vasculopathy results in cord ischemia
  - Associated demyelinating process
  - Neoplastic, paraneoplastic, collagen vascular, and iatrogenic etiologies
    - Not considered "idiopathic" in those cases
  - Association with previous viral infection or vaccination postulated in some cases
- Associated abnormalities
  - Depression
    - Lifetime risk (> 50%), correlates with severity of sensory symptoms
- Perivascular inflammation
- Demyelination

Microscopic Features

- Gray and white matter necrosis
- Destruction of neurons, axons, and myelin
- Astrocytic gliosis
- Perivascular lymphocytic infiltrate

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sensory deficit
    - Loss of pain and temperature sensation
    - Clearly defined upper level
    - Band-like dysesthesia at lesion level
    - Ascending paresthesia in bilateral lower extremities
  - Other signs/symptoms
    - Paraplegia or quadriplegia
    - Back ± radicular pain
    - Bladder and bowel dysfunction
    - Hypotonia and hyporeflexia initially
    - Spasticity and hyperreflexia over time
- Clinical profile
  - Prodrome of generalized body aches
  - Preceding viral-like illness
  - Rapid progression to maximal neurologic deficits within days
- Transverse Myelitis Consortium Working Group
  - Inclusion criteria
    - Sensory, motor, or autonomic dysfunction attributable to spinal cord
    - Bilateral signs and symptoms
    - Well-defined sensory level
    - Cord inflammation confirmed by CSF pleocytosis, elevated IgG index, or gadolinium enhancement
    - Progression to nadir between 4 hours and 21 days from onset
    - Symptoms must progress if present upon awakening
    - Compressive etiology excluded by neuroimaging
    - May repeat MR ± lumbar puncture between 2-7 days if signs of inflammation absent initially
  - Exclusion criteria
    - History of spinal radiation within past 10 years
    - Cord ischemia and infarction due to anterior spinal artery thrombosis
    - Cord arteriovenous malformation
    - Connective tissue disorder (sarcoidosis, SLE, or Sjögren syndrome)
    - CNS infections (syphilis, Lyme disease, mycoplasma, HIV, HTLV-1, or other viral infections)
    - Brain MR suggestive of multiple sclerosis
    - Clinical optic neuritis
  - 2005 modifications
    - Acute partial transverse myelitis (TM)
      - Small cord lesions, associated with multiple sclerosis (MS)
    - Acute complete TM
      - Associated with longitudinally extensive TM
      - Within spectrum of neuromyelitis optica (NMO) as antiaquaporin 4 antibody is present in 1/2 of recurrent cases of IATM

Demographics

- Age
  - Any, but peaks at 10-19 and 30-39 years
- Epidemiology
  - Incidence 2 per 500,000 population
    - 4.6 new cases of IATM per million people per year in USA
      - 1,200 new cases per year in USA
  - Majority of cases occurred in late winter through spring in 1 series

Natural History & Prognosis

- Typically monophasic
  - Recurrence rate reported between 24-40%
  - If recurrent, must consider
    - MS: Progression to multiple sclerosis in 2-8% of cases of transverse myelitis
    - SLE, antiphospholipid syndrome
    - Vascular malformation
- 1/3 of patients experience good to complete recovery
  - Symptomatic improvement 2-12 weeks after onset
  - Children have slightly better prognosis than adults
- 1/3 of patients experience fair recovery
  - Residual spasticity and urinary dysfunction
- 1/3 of patients experience poor recovery
  - Persistent complete deficits
  - Assistance required in activities of daily living
- Factors portending poor prognosis
  - Back pain
  - Rapid clinical deterioration
  - Spinal shock: Loss of motor, sensation, sphincter control, and areflexia
  - MR signal alteration > 10 spinal segments
  - Significant denervation on electromyogram or abnormal somatosensory evoked potential
  - Presence of antiaquaporin 4 antibody

Treatment

- High-dose intravenous steroid pulse therapy
- Physical therapy

DIAGNOSTIC CHECKLIST

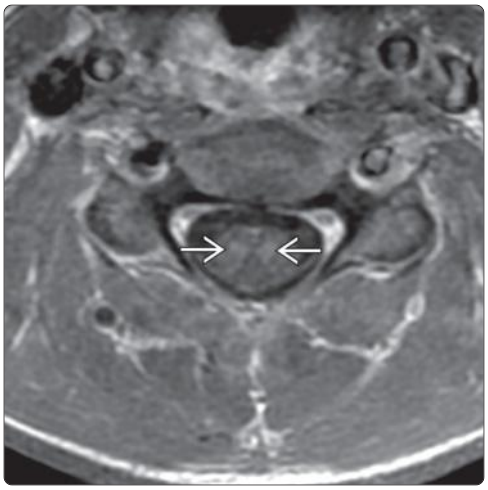
Image Interpretation Pearls

- IATM is diagnosis of exclusion with multiple differential diagnostic possibilities
- Central enhancement in peripherally located T2 hyperintensity more characteristic of multiple sclerosis

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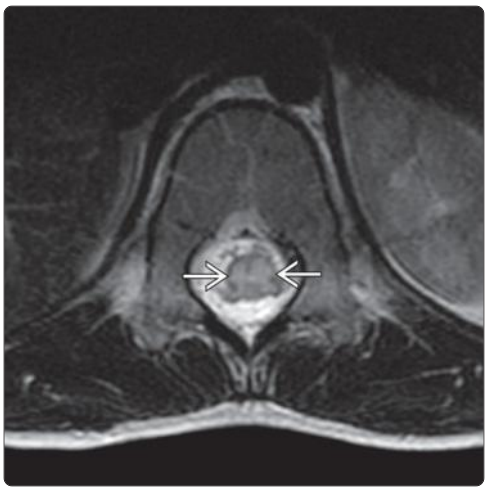
(Left) *Sagittal STIR MR demonstrates abnormal enlargement and edema within the midcervical spinal cord spanning 3 vertebral levels. Low signal intensity within the CSF ventral to the cord represents CSF pulsation artifact.* **(Right)** *Axial T1 C+ MR (same patient) confirms abnormal, bilateral, central intramedullary enhancement within the abnormally enlarged cervical spinal cord.*





(Left) *Sagittal T2WI MR demonstrates long segment T2 hyperintensity within the thoracic cord spanning 6 levels. This is the typical central edema pattern for ATM, with relative sparing of the cord periphery. CSF dephasing, a common artifact, mimics an intradural extramedullary lesion.* **(Right)** *Sagittal T1WI C+ FS MR (same patient) reveals a single focus of enhancement within the abnormal thoracic spinal cord segment that is much smaller than the extent of T2 signal abnormality.*





(Left) *Sagittal T2WI MR of the thoracolumbar spine demonstrates mild conus enlargement and abnormal T2 hyperintensity representing edema spanning nearly 2 vertebral levels.* **(Right)** *Axial T2WI MR (same patient) confirms abnormal conus T2 signal intensity, mainly within central gray mater. There is conus enlargement secondary to edema but no central fluid signal to indicate syringohydromyelia.*

KEY FACTS

TERMINOLOGY

- Primary demyelinating disease of central nervous system with multiple lesions disseminated over time and space
  - Concomitant intracranial lesions in periventricular, subcallosal, brain stem, or cerebellar white matter

IMAGING

- Isolated spinal cord disease (10-20%)
- Cervical segment is most commonly affected
  - Dorsolateral aspect of cord
  - < 1/2 of cross-sectional area of spinal cord
  - < 2 vertebral segments in length
- Sagittal and axial T1WI & T2WI sequences with gadolinium
  - Lesions typically oval, peripheral, and asymmetric
  - Discrete vs. vague hyperintense lesions
  - Enhancement lasts 1-2 months but does not reflect disease progression

TOP DIFFERENTIAL DIAGNOSES

- Intramedullary neoplasm
- Idiopathic transverse myelitis
- Neuromyelitis optica

PATHOLOGY

- Autoimmune, cell-mediated inflammatory process focused on CNS myelin

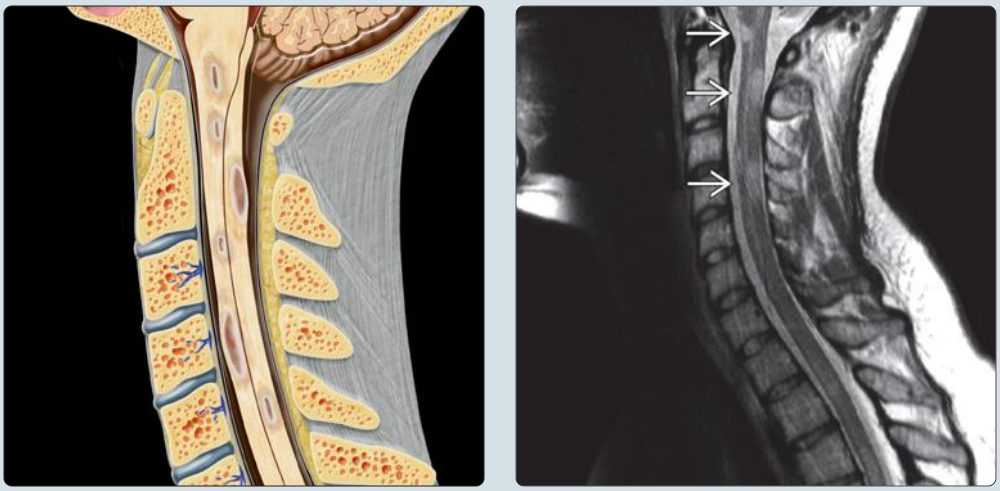
CLINICAL ISSUES

- Peak onset: 20-40 years
- Relapsing remitting (RR)
- Secondary progressive (SP)
- Primary progressive (PP)
- Progressive relapsing (PR)

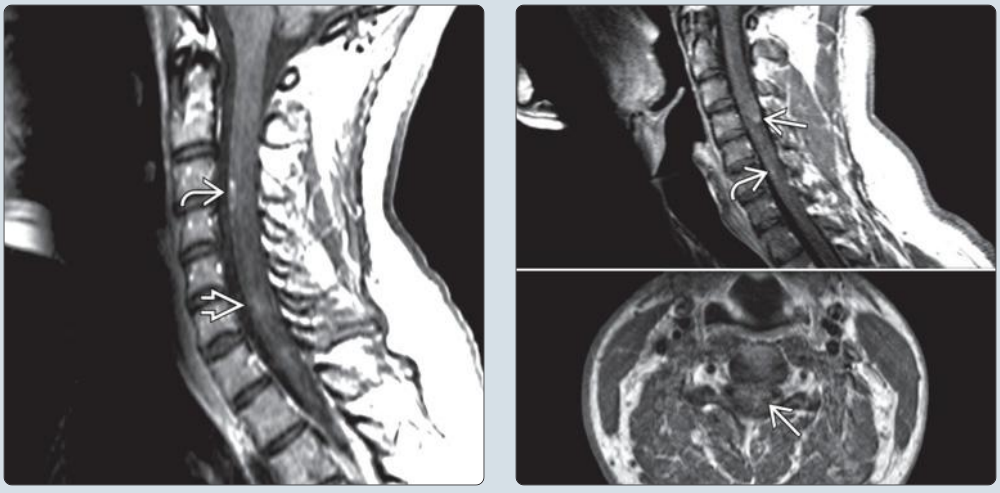
DIAGNOSTIC CHECKLIST

- Imaging findings must be correlated with clinical and laboratory features to confirm diagnosis

(Left) *Sagittal graphic depicts multiple demyelinating plaques within the cervical spinal cord.* **(Right)** *Sagittal T2WI MR of the cervical spinal cord demonstrates multiple T2 hyperintense foci, some well defined and others ill defined. The multiplicity of lesions & lack of edema or significant cord expansion is typical for demyelinating disease.*



(Left) *Sagittal T1WI C+ MR shows multiple enhancing demyelinating lesions within the cervical spinal cord. Enhancement varies from focal to ill defined. The enhancement pattern changes with evolution of inflammation.* **(Right)** *T1WI C+ MR (sagittal on top, axial on bottom) illustrates an incomplete rim-enhancing lesion in the dorsal cervical cord at the C3-C4 level. A 2nd small enhancing focus is noted in the ventral cord at the C6 level.*



TERMINOLOGY

Abbreviations

- Spinal cord multiple sclerosis (MS)

Definitions

- Primary demyelinating disease of central nervous system with multiple lesions disseminated over time and space

IMAGING

General Features

- Best diagnostic clue
  - Concomitant intracranial lesions in periventricular, subcallosal, brain stem, or cerebellar white matter
- Location
  - Isolated spinal cord disease (10-20%)
  - Cervical segment is most commonly affected
    - 2/3 of cord lesions
  - Dorsolateral aspect of cord
  - Does not respect gray-white boundary
- Size
  - < 1/2 of cross-sectional area of spinal cord
  - < 2 vertebral segments in length
- Morphology
  - Wedge-shaped on axial MR
    - Apex directed centrally

MR Findings

- T1WI
  - Iso- to hypointense lesions
    - In cord (unlike brain), rarely visible as hypointense
- T2WI
  - Discrete or ill-defined hyperintense lesions
    - May be related to extent of demyelination
    - Lesions ↑ in size due to edema associated with inflammatory infiltrates → reaches max size at 4 weeks
    - Slow decrease in size over 6-8 weeks as edema resolves ± remyelination
  - Lesions typically oval, peripheral, and asymmetric
- STIR
  - Improved lesion detection
- FLAIR
  - Lower sensitivity compared to STIR in cord imaging
- DWI
  - ↑ mean diffusivity, ↓ fractional anisotropy in plaques & areas without T2 abnormality
- T1WI C+
  - Variable
    - Homogeneous, nodular, or ring enhancement during acute or subacute phase
      - Enhancement lasts 1-2 months
      - Does not reflect disease progression
    - No enhancement during chronic phase
- Solitary or multifocal lesions
  - Larger lesions formed by coalescence of smaller plaques
- Normal or mild focal cord expansion
  - Cord edema
    - Resolves after 6-8 weeks
- Cord atrophy
  - Usually in late stage
  - May be seen in early disease course
  - Useful for monitoring disease progression and therapeutic efficacy
  - Correlates with clinical disability
- Functional MR (fMR)
  - Tactile-associated cervical cord fMR activity increased in relapse-onset MS patients
    - Overactivation more prominent in patients with more severe locomotor disability

DIFFERENTIAL DIAGNOSIS

Intramedullary Neoplasm

- Cord expansion
- Entire cross section of spinal cord
- Peritumoral edema
- Diffuse or partial enhancement
- Cystic ± hemorrhagic components

Idiopathic Transverse Myelitis

- Lesion centrally located
- 3-4 segments in length
- Involving > 2/3 of cord cross-sectional area
- Variable enhancement
- No associated intracranial lesions
- Diagnosis of exclusion

Neuromyelitis Optica

- Autoimmune inflammatory disorder involving myelin of neurons of optic nerves & spinal cord
- Limited brain involvement
- Longitudinally extensive (> 3 vertebral segments) T2 hyperintensity within cord + enhancement of optic nerves
- T2 abnormality involves entire cross section of cord

PATHOLOGY

General Features

- Etiology
  - Autoimmune, cell-mediated inflammatory process focused on CNS myelin
    - Infectious agents may play 1° or 2° role
      - Humoral mechanism: Cross-reactivity between infectious and self-epitopes
  - May be association between MS & altered venous return due to multiple extracranial venous strictures
    - Hampered cerebrospinal venous drainage in patients with MS determines complex hemodynamic picture → chronic cerebrospinal venous insufficiency (CCSVI)
      - Multiple substitute circles with very high incidence of reflux in both intracranial and extracranial venous segments
      - Loss of postural regulation of cerebral venous outflow
    - ↑ cerebral blood flow and volume & ↓ mean transit time (compared with baseline values before relapse) precede development of plaques
    - Previously, these findings interpreted as sign of local flow disturbances mediated by inflammatory & neurodegenerative processes

- However, may be attributable to recent findings of significant stenoses in extracranial veins draining brain & spinal cord
- Genetics
  - MS may be inherited as complex multifactorial disorder resulting from interaction of genetic and environmental factors
    - Estimated risk to siblings of proband ~ 3.0-5.0%, ↑ to 29.5% if 1 or both parents have MS
    - Risk to offspring of person with MS is 2.0-3.0% and higher if both parents have MS
- Associated abnormalities
  - 90% incidence of associated intracranial lesions
  - Neurofibromatosis type 1

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Cord lesions may be asymptomatic
  - Paresthesia
- Other signs/symptoms
  - Muscle weakness
  - Hyperreflexia
  - Gait disturbance
  - Bladder & bowel dysfunction
- Surveillance includes periodic neurological examination to track disease progression & periodic brain & spinal cord MRs to monitor disease activity

Demographics

- Age
  - Peak onset: 20-40 years
  - Onset: < 18 years of age in 3-5% of MS cases
- Gender
  - Adult females more susceptible than males (1.7:1)
    - Men more likely to have PR & SP MS
    - Women more likely to have RR MS
  - Both sexes are equally affected in PP MS
- Ethnicity
  - Western Europeans have higher risk
- Epidemiology
  - Increasing prevalence further north from equator
    - 30-80 per 100,000 in northern USA & Europe
    - 6-14 per 100,000 in southern USA & Europe
    - 1 per 100,000 in equatorial regions

Natural History & Prognosis

- Benign (20%)
  - Complete recovery after 1-2 attacks
  - Some may experience progressive MS after 10-15 years
- Relapsing remitting (RR) (25%)
  - Distinct periods of new or worsening symptoms alternating with complete or partial recovery
  - 90% will evolve into progressive MS after 25 years
- Secondary progressive (SP) (40%)
  - From relapsing-remitting MS
  - Worsening deficits & disabilities
  - Incomplete & infrequent remission
- Primary progressive (PP) (12%)
  - Steady progression of symptoms
    - Motor dysfunction common
    - Primary cord involvement
    - No distinct attacks
- Progressive relapsing (PR) (3%)
  - Similar to primary progressive MS
  - Includes distinct periods of exacerbation but without recovery
  - High mortality rate

Treatment

- Agents decrease clinical relapse rate & accompanying inflammation within CNS
  - IV & oral prednisone
  - Plasmapheresis
    - If no response to steroid therapy
  - Interferon β (interferon β-1b, interferon β-1a)
    - Inhibition of immune cells
  - Glatiramer acetate
    - Synthetic protein similar to myelin protein
    - Serves as substrate for T cells
  - Mitoxantrone
    - Suppression of T lymphocytes & B lymphocytes
- Supportive therapy for symptomatic treatment of pain, muscle spasms, fatigue, depression, sexual dysfunction, & bladder & bowel dysfunction
  - Anticholinergics
  - Smooth muscle relaxants

DIAGNOSTIC CHECKLIST

Consider

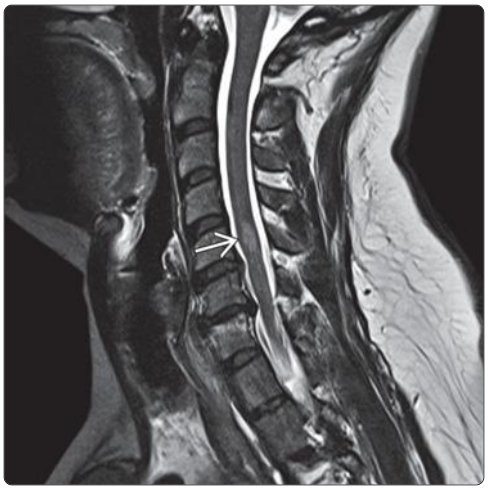
- Brain MR, including high-resolution fast spin-echo T2 through corpus callosum
  - Periventricular, subcallosal, brain stem, or cerebellar white matter lesions suggest MS
- Gray matter atrophy correlates with disability

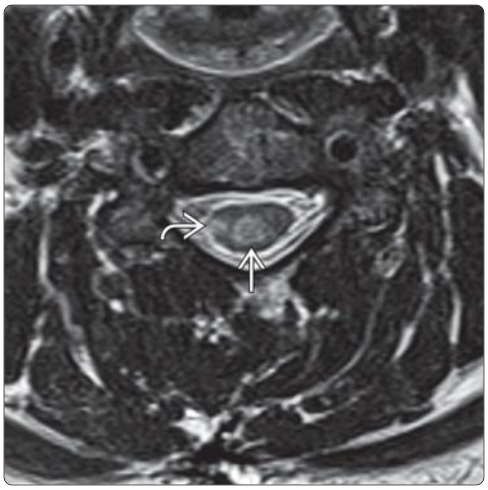
Image Interpretation Pearls

- Imaging findings must be correlated with clinical & laboratory features to confirm diagnosis
- Acute MS can mimic cord neoplasm

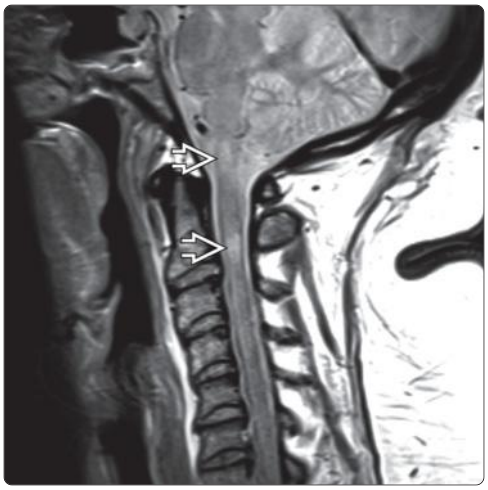
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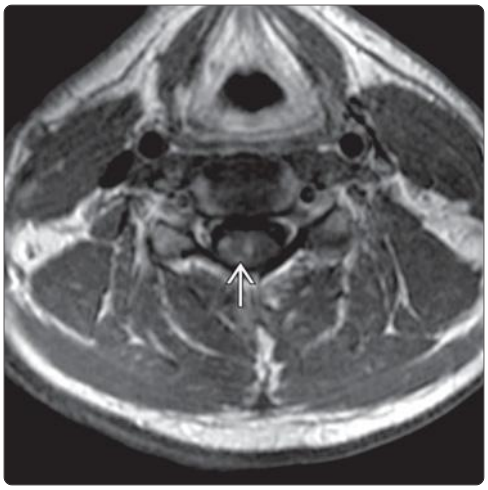
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(Left) *Sagittal T2 MR shows focal T2 hyperintensity within the cervical spinal cord is seen at C5 level. There is minimal cord expansion produced by the fairly large lesion, characteristic of demyelinating lesions.* **(Right)** *Axial T2WI MR shows focal T2 hyperintense demyelinating lesions with both central and peripheral involvement. T2 hyperintense lesions are not specific for plaque age, degree of myelin & axon loss, or amount of edema & inflammation.*





(Left) *Sagittal PD FSE MR of the cervical spinal cord demonstrates characteristic ovoid hyperintense intramedullary demyelinating lesions without significant cord expansion.* **(Right)** *Axial T1 C+ MR of the cervical spinal cord depicts focal ring enhancement within an active multiple sclerosis demyelinating lesion.*





(Left) *Sagittal STIR MR shows a focal hyperintense demyelinating plaque within the thoracic cord without significant cord expansion. STIR MR is more sensitive for lesion depiction than T2WI MR at the price of more artifacts.* **(Right)** *Sagittal T2WI MR of the thoracic spinal cord illustrates numerous hyperintense intramedullary foci.*

Neuromyelitis Optica

KEY FACTS

TERMINOLOGY

- Synonyms: Devic disease, optic-spinal multiple sclerosis
- Autoimmune inflammatory disorder involving myelin of neurons of optic nerves and spinal cord, with limited brain parenchymal involvement

IMAGING

- LTE (> 3 vertebral segments) T2 hyperintensity within cord + enhancement of optic nerves (85% of cases)
- T2 abnormality tends to involve entire cross section of cord, unlike more focal involvement of MS

TOP DIFFERENTIAL DIAGNOSES

- Inflammatory etiologies
  - Multiple sclerosis
  - Acute disseminated encephalomyelitis (ADEM)
  - Parainfectious myelitis (viral, bacterial)
- Systemic autoimmune disease
- Noninflammatory mimics
  - Dural fistula
  - Tumor
- Idiopathic acute transverse myelitis

PATHOLOGY

- Autoimmune disease targeting water channel proteins (aquaporin-4)

CLINICAL ISSUES

- **Revised diagnostic criteria (2006)**
  - Optic neuritis, myelitis, and at least 2 of 3 supportive criteria
    - Contiguous cord lesion 3 or more segments in length, initial brain MR nondiagnostic for MS, or NMO-IgG seropositivity

DIAGNOSTIC CHECKLIST

- Simultaneous ON and myelitis → NMO cannot be distinguished from ADEM at 1st attack





(Left) *Sagittal T2WI MR shows a long segment of cord enlargement with T2 hyperintensity.* **(Right)** *Sagittal T1WI C+ MR shows cord enlargement and ill-defined enhancement. Neuromyelitis optica (NMO) is an autoimmune disease possibly targeting the aquaporin-4 transmembrane water channel. The vasculocentric distribution of antigen of NMO-IgG correlates with the sites of immunoglobulin and complement deposition seen in spinal cord lesions of patients with NMO.*





(Left) *Axial T1WI C+ FS MR reveals intense enhancement and mild enlargement of the intracanalicular and prechiasmatic segments of the right optic nerve.* **(Right)** *Coronal T1WI C+ FS MR reveals intense enhancement and mild enlargement of the intracanalicular and prechiasmatic segments of the right optic nerve. This is the classic appearance of Devic disease involving the optic nerves and spinal cord, with no brain parenchymal abnormalities.*

TERMINOLOGY

Abbreviations

- Neuromyelitis optica (NMO)
- Longitudinally extensive (LTE)

Synonyms

- Devic disease, Devic syndrome, Devic neuromyelitis optica, optic-spinal multiple sclerosis (MS)

Definitions

- Autoimmune inflammatory disorder involving myelin of neurons of optic nerves and spinal cord, with limited brain parenchymal involvement

IMAGING

General Features

- Best diagnostic clue
  - Longitudinally extensive (> 3 vertebral segments) T2 hyperintensity within cord + enhancement of optic nerves (85% of cases)
- Location
  - Spinal cord + optic nerves/chiasm
  - Studies have reported MS-like lesions in 10-30% of NMO patients; brain MR is usually normal
- Size
  - > 3 segments of T2 abnormality more likely in NMO or acute transverse myelitis than in classic MS
    - Most specific nonserological feature to differentiate NMO from MS
- Morphology
  - T2 abnormality tends to involve entire cross section of cord, unlike more focal involvement of MS

Imaging Recommendations

- Best imaging tool
  - STIR, T2WI MR, C+ MR of cord/brain
- Protocol advice
  - Proton density or STIR imaging of cord; sole use of heavily T2WI will lower sensitivity to cord lesions

MR Findings

- T2WI
  - Lesions more linear than MS patients
- DWI
  - Mean diffusivity and fractional anisotropy in patients with relapsing NMO are significantly different from those in patients with relapsing MS
- MRS
  - NAA:Cr, Cho:Cr ratios, myoinositol values normal in NMO vs. abnormal in MS

DIFFERENTIAL DIAGNOSIS

Inflammatory Etiologies

- Multiple sclerosis
  - < 1 vertebral segment length of cord T2 signal abnormality
  - Tends to involve cord periphery
  - Multiple cord lesions
  - Associated typical periventricular white matter lesions
- Acute disseminated encephalomyelitis (ADEM)
  - Encephalopathy
  - May be indistinguishable from MS on imaging
  - Thalamic involvement
  - Monophasic
- Parainfectious myelitis (viral, bacterial)
  - Long segment of T2 signal abnormality
  - Usually prominent irregular enhancement (not ring-shaped)
- Systemic autoimmune disease
  - Sjögren, SLE, antiphospholipid syndrome

Noninflammatory Mimics

- Dural fistula
  - Elderly men most common
  - Central cord T2 hyperintensity
  - Diffuse, mild enhancement
  - Associated intradural vessel flow voids
- Tumor
  - Irregular enhancement
  - Cord expansion, may be asymmetric

Idiopathic Acute Transverse Myelitis

- Diagnosis of exclusion
- Cord lesion may be indistinguishable from NMO
- Monophasic

PATHOLOGY

General Features

- Etiology
  - Autoimmune disease → possibly targeting of aquaporin-4 (AQP4)
    - Aquaporins = family of transmembrane channel proteins highly selective for water transport
    - AQP4 predominant water channel expressed in brain
    - Localized to astrocytic foot processes surrounding cerebral capillary endothelium
    - NMO-IgG binds to abluminal face of microvessels, pia, and Virchow-Robin sheath
  - Possible parainfectious NMO syndromes
    - Reported association with viral and bacterial pathogens (Varicella-Zoster virus and *Mycobacterium pneumonia*)
    - Acute febrile illness preceding or in close relation with neurological symptoms
    - Monophasic course in majority with disability sustained
  - Longitudinally extensive transverse myelitis (LETM) may confer high risk of conversion into NMO
    - Autoimmune disorder caused by pathogenic antibodies to aquaporin-4
    - Cases of infectious or parainfectious NMO/LETM (associated with herpes zoster) → etiological diversity in NMO/LETM
    - May be pathogenic role of AQP4-Ab in LETM
- Associated abnormalities
  - Brain white matter lesions
    - Recurrent optic neuritis, myelitis with normal brain MR, or when few brain WM lesions fail MS criteria → NMO very likely
    - WM lesions common in patients with otherwise typical NMO (60%)

- Asymptomatic brain lesions are common in NMO, and symptomatic brain lesions do not exclude diagnosis of NMO
- Atypical brain lesions can occur with involvement of thalamus, hypothalamus, brain stem, cerebellum
  - Diencephalic, brainstem, or cerebral hemispheric lesions observed in pediatric patients
  - Children with NMO are more susceptible to atypical brain lesions
- Other concomitant or associated diseases
  - Prior viral infection
  - ANA positive (10%)
  - Autoimmune thyroiditis (9%)
  - Myasthenia gravis (2%)
  - Sjögren syndrome

Gross Pathologic & Surgical Features

- Optic nerves show nonspecific segments of demyelination + atrophy
- Spinal cord shows perivascular inflammatory demyelination with necrosis
- May show ↑ PMNs + eosinophils (not seen in MS)

Microscopic Features

- Prominent vasculocentric IgG and C9 neoantigen deposits
- Eosinophilic and neutrophilic inflammatory infiltrates, necrosis, vascular hyalinization, and extensive vasculocentric immunoglobulin and complement deposition

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Recurrent optic neuritis, recurrent transverse myelitis, simultaneous attacks of ON + myelitis
  - Bilateral simultaneous ON or sequential ON more suggestive of NMO than MS
  - Respiratory failure due to extensive cervical myelitis in up to 1/3 of cases (very uncommon in MS)
  - Radicular pain in 35% (uncommon in MS)
  - L'hermitte symptom common in MS (40%) and NMO (35%)
  - CSF in NMO may show prominent pleocytosis (especially with myelitis) compared to MS
    - Oligoclonal bands in 20-30% (vs. 85% with MS)
- Other signs/symptoms
  - **Revised diagnostic neurology criteria**, Wingerchuk **(2006)**
    - Optic neuritis, myelitis, and at least 2 of 3 supportive criteria
      - → MR evidence of contiguous spinal cord lesion 3 or more segments in length, onset brain MR nondiagnostic for multiple sclerosis, or NMO-IgG seropositivity
      - CNS involvement beyond optic nerves and spinal cord is compatible with NMO
    - NMO-IgG seropositivity 76% sensitive and 94% specific for NMO

Demographics

- Age
  - Mean age range: 30-40 years
- Gender
  - F > M (4:1)
- Ethnicity
  - NMO greater proportion of CNS demyelinating disease in people of non-Caucasian background

Natural History & Prognosis

- Outcome worse than in MS
- Frequent and early relapses (annual relapse rate of 1.3%)
  - Increased risk of relapse during pregnancy in NMO
- Compared with MS, NMO patients have older age at onset (depending on ethnicity), females more frequently affected, course more severe
  - Vision/ambulation impaired within 5 years of onset in 50%
  - 20% succumb to respiratory failure
- Some reports state predictor of poor evolution is high number of MR brain lesions at diagnosis → predictive of residual visual acuity

Treatment

- Intravenous steroids used as first-line treatment of ON/myelitis
- Immunosuppressive medications (e.g., azathioprine, corticosteroids, and rituximab)
- Plasmapheresis as rescue therapy
- Optimal prevention therapy unknown
  - Standard MS immunomodulatory therapies
  - Azathioprine + oral prednisone

DIAGNOSTIC CHECKLIST

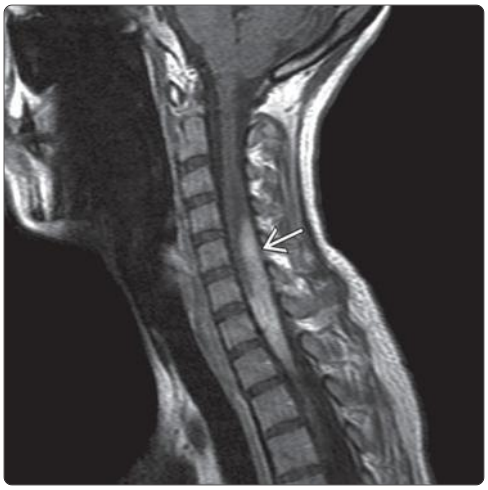
Consider

- Simultaneous ON and myelitis → NMO cannot be distinguished from ADEM at 1st attack
- Brain MR should be normal or near normal (≤ 2 lesions)
- Longitudinally extensive cord T2 hyperintensity in 85%
- Best diagnostic combination (99% sensitive and 90% specific)
  - At least 2 of 3 elements: LTE cord lesion, onset brain MR nondiagnostic for MS, or NMO-IgG seropositivity

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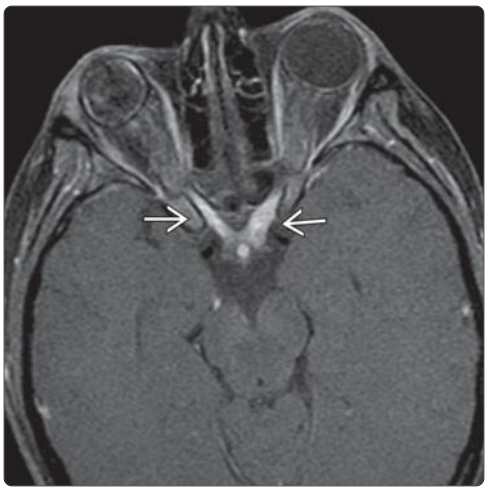


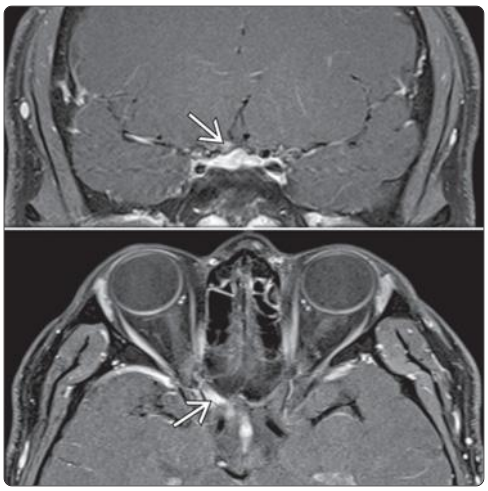
(Left) *Sagittal T2WI MR shows fusiform expansion of the cervical cord with longitudinally extensive T2 signal abnormality > 3 vertebral segments in this patient with NMO.* **(Right)** *Sagittal T1 C+ MR shows extensive diffuse enhancement of the cervical cord with fusiform cord expansion.*





(Left) *Sagittal T1WI C+ MR exhibits a long segment of diffuse enhancement. This initial pattern is nonspecific, and the differential would include other acute transverse myelopathies, such as the noninflammatory group (dural fistula, cord infarct, radiation change) and the inflammatory group (MS, NMO, ADEM, SLE, and other connective tissue disorders).* **(Right)** *Sagittal T2WI MR depicts a longitudinally extensive focus of hyperintensity within the thoracic cord, with slight fusiform expansion.*





(Left) *Axial T1 C+ FS MR of the optic nerves in this patient with NMO demonstrates marked enhancement of both optic nerves and the optic chiasm. There is phthisis bulbi on the right.* **(Right)** *T1W C+ FS MR images demonstrate mild enlargement & enhancement of the intracanalicular right optic nerve. Systemic autoimmune disorders & autoantibodies may coexist in NMO (e.g., thyroid disease, myasthenia gravis, celiac disease, systemic lupus erythematosus, & Sjögren syndrome).*

ADEM

KEY FACTS

TERMINOLOGY

- Para-/postinfectious immune-mediated inflammatory disorder of white matter
  - Antibodies to pathogens exhibit cross-immunoreactivity with myelin basic protein

IMAGING

- Multifocal white matter lesions with relatively little mass effect or vasogenic edema
  - Flame-shaped lesions with slight cord swelling
  - Dorsal white matter more voluminous
  - May see gray matter involvement
  - Variable enhancement, depending on stage of disease
  - May see nerve enhancement
- Brain almost always involved

TOP DIFFERENTIAL DIAGNOSES

- Inflammatory etiologies
  - Acquired demyelinating
    - Multiple sclerosis
    - Neuromyelitis optica
  - Parainfectious myelitis
  - Systemic autoimmune (vasculitis)
- Noninflammatory mimics
  - Vascular malformation
  - Infarction, spinal cord
  - Tumor and cysts
- Idiopathic acute transverse myelitis

CLINICAL ISSUES

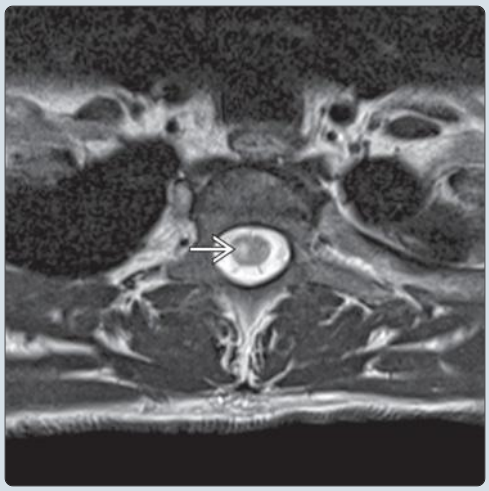
- Typically monophasic illness lasting from 2-4 weeks
  - Encephalopathy, paresis
- Typically childhood or young adult

DIAGNOSTIC CHECKLIST

- Rescanning if initially negative
  - Typically, delay between clinical onset and appearance of imaging findings

(Left) *Sagittal T1WI C+ MR shows linear enhancement along the dorsal surface of the midcervical cord. Acutely, there is perivenous edema, demyelination, and infiltration with macrophages and lymphocytes, with relative axonal sparing. The late course of the disease is characterized by perivascular gliosis.* **(Right)** *Axial T2WI MR depicts a hyperintense focus in the right hemicord with minimal cord enlargement. ADEM is a postinfectious disease mediated by autoreactive cells or molecules.*





(Left) *Sagittal T2WI MR demonstrates a vague intramedullary focus of hyperintensity. There is mild cord expansion.* **(Right)** *Sagittal T1WI C+ MR illustrates an ovoid intramedullary focus of enhancement in the upper thoracic cord. Any 2 of the following criteria are proposed to distinguish MS from ADEM: 1) absence of a diffuse bilateral lesion pattern 2) presence of black holes 3) presence of 2 or more periventricular lesions (81% sensitivity and 95% specificity).*





TERMINOLOGY

Abbreviations

- Acute disseminated encephalomyelitis (ADEM)

Definitions

- Para-/postinfectious immune-mediated inflammatory disorder of white matter

IMAGING

General Features

- Best diagnostic clue
  - Multifocal white matter lesions with relatively little mass effect or vasogenic edema
- Location
  - Anywhere in spinal cord white matter
  - Brain almost always involved
- Size
  - Punctate to segmental
- Morphology
  - Plump lesions with feathery edges

CT Findings

- CECT
  - May see (multi)focal intramedullary enhancement

MR Findings

- T1WI
  - Focal low signal and slight cord swelling
- T2WI
  - Multifocal flame-shaped white matter lesions with slight cord swelling
  - Dorsal white matter more voluminous
  - May see gray matter involvement
- PD/intermediate
  - Sometimes depict lesions more sensitively
- T2* GRE
  - Same as T2WI; not as sensitive
- DWI
  - Hyperintensity
    - Acute lesions reported to have restricted diffusion and free diffusion in subacute stage
- T1WI C+
  - Variable enhancement depending on stage of disease
  - Punctate, ring-shaped, or fluffy enhancement
  - May see nerve enhancement
- MRS
  - NAA/Cho, Cho/Cr ratios significantly different between lesions in acute and subacute stages
    - Progressive decrease in NAA/Cho ratios
- Brain findings
  - Lesions in deep cerebral hemispheric and subcortical white matter, basal ganglia, gray-white junction, diencephalon, brainstem, cerebellum
- Delayed development of MR lesions in deep gray matter and brainstem may herald prolonged clinical course and lack of response to glucocorticoid therapy
  - Possible that lesions in these areas take longer to evolve and become visible with standard resolution MR
  - Plasmapheresis might be effective therapeutic intervention in these patients

Imaging Recommendations

- Best imaging tool
  - T2WI and T1WI pre- and post-contrast MR
- Protocol advice
  - Include brain MR whenever suspicious cord lesions found
    - Sagittal FLAIR to assess for callosal lesions

DIFFERENTIAL DIAGNOSIS

Inflammatory Etiologies

- Acquired demyelinating
  - Multiple sclerosis
    - Indistinguishable on single study
    - Brain MR may help differentiate (cranial nerve involvement indicates ADEM)
    - Clinical course (relapsing-remitting) indicates MS
  - Neuromyelitis optica
    - Longitudinally extensive intramedullary hyperintensity on T2WI
    - Little clinical involvement beyond optic nerves and spinal cord
- Parainfectious myelitis
  - Acute inflammatory insult of spinal cord due to direct viral infection or post-viral immunologic attack
- Systemic autoimmune (vasculitis)
  - Systemic lupus erythematosus

Noninflammatory Mimics

- Vascular malformation
  - High-flow AVM demonstrates obvious serpentine structures with flow void at periphery of abnormal cord
  - Dural AVF often shows distal cord signal increase only, not multifocal
    - Stuttering course of paresis in adult patient
- Infarction, spinal cord
  - Stroke-like, acute presentation
  - Focal, segmental lesion in gray matter
  - Typically conus involvement
- Tumor and cysts
  - Cord tumor
    - Diffuse or nodular contrast enhancement
    - Peritumoral edema
    - Cystic and hemorrhagic components

Idiopathic Acute Transverse Myelitis

- Diagnosis of exclusion
- Usually single lesion

PATHOLOGY

General Features

- Etiology
  - Autoimmune process producing inflammatory reaction
    - Antibodies to pathogens may exhibit cross-immunoreactivity with myelin basic protein in white matter
  - Predisposing infection
    - Mumps, measles, chickenpox, influenza, Epstein-Barr
    - *Campylobacter* spp, *Streptococcus* spp, *Leptospira* spp
    - Mycoplasma
  - Postvaccinial association in some reported cases

- Polio, rabies, rubella vaccine, DPT
- Following intravenous immunoglobulin administration

- Genetics
  - HLA-DR link
- Tumefactive swelling in some larger acute lesions
  - Most lesions show no gross features
- Increasingly recognized
  - More sensitive imaging tools

Gross Pathologic & Surgical Features

- Slight swelling to tumefactive necrosis

Microscopic Features

- Acute myelin loss
  - Perivenous demyelination → pathological hallmark of ADEM; confluent demyelination → hallmark of acute multiple sclerosis
- Lymphocytic infiltrate
- Axonal preservation
- Astroglial reaction, proliferation
- Distinct pattern of cortical microglial activation without cortical demyelination → pathological correlate of depressed level of consciousness in ADEM

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Encephalopathy
    - Altered consciousness (lethargy, coma)
    - Behavioral changes (confusion, irritability)
  - Other signs/symptoms
    - Paresis
    - Cranial nerve palsies
    - Convulsions
    - Lower urinary tract dysfunction
      - Up to 33%, associated with occurrence of paraparesis or tetraparesis
      - Especially with lower limb pyramidal involvement
      - Voiding dysfunction, urinary retention
      - Incontinence more often in patients with frontoparietal white matter changes
- Clinical profile
  - Usual prodromal phase
    - Fever, malaise, myalgia
  - Abnormal CSF
    - Increased protein
    - Usually no oligoclonal bands
    - Leukocytosis

Demographics

- Age
  - Typically childhood or young adult
- Gender
  - Some reports suggest male predominance

Natural History & Prognosis

- Typically monophasic illness lasting from 2-4 weeks
  - Relapses have been reported: Biphasic disseminated encephalomyelitis or multiphasic disseminated encephalomyelitis
  - Current diagnostic criteria → presence of new lesions on MR repeated 3 months after attack is predictive of MS
    - Most children with MS show new lesions on MR repeated within 2 years even when they remain asymptomatic
- 50-60% recover completely
- Neurologic sequelae in 30-40%
  - Patients with extensive lesions in brainstem or cervical cord reported to have more deficits compared with those whose lesions are mainly in cerebellum or subcortical white matter
- Positive myelin oligodendrocyte glycoprotein (MOG) antibodies
  - Associated with increased frequency of longitudinal extensive transverse myelitis and better outcome
- Mortality (10%)

Treatment

- Options, risks, complications
  - Immunomodulation, supportive
    - Steroid treatment with intravenous methylprednisolone is treatment of choice, immunoglobulin therapy
  - Plasmapheresis in fulminant case
    - Potential complications: Central line sepsis, hypotension, anemia, electrolyte abnormalities, and heparin-induced thrombocytopenia

DIAGNOSTIC CHECKLIST

Consider

- Rescanning if initially negative
  - Typically, delay between clinical onset and appearance of imaging findings

Image Interpretation Pearls

- Check thin-section intracranial slices on T1 C+ for cranial nerve enhancement
- ADEM can mimic cord neoplasm!

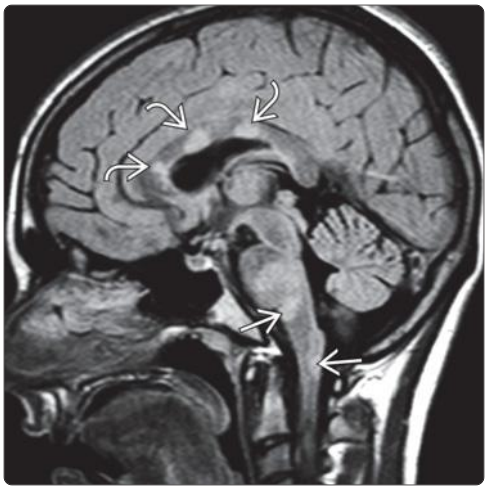
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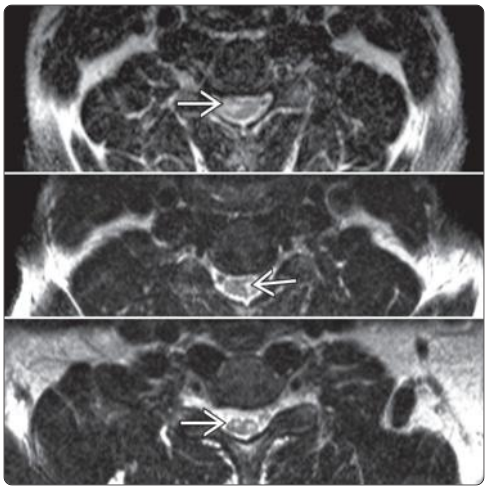
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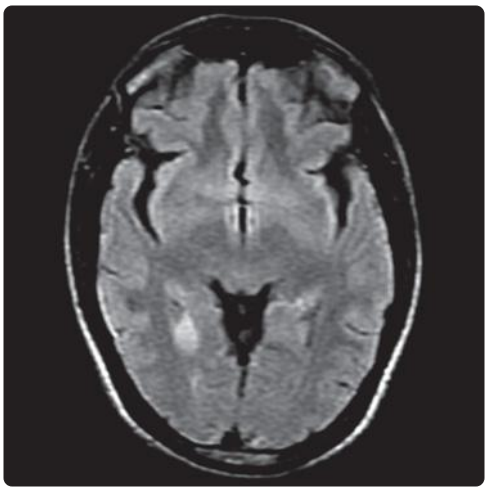


(Left) *Sagittal T1 C+ MR reveals multiple intramedullary foci of enhancement reflecting active demyelination and disruption of the blood cord barrier.* **(Right)** *Sagittal T2WI MR demonstrates multiple foci of hyperintensity scattered throughout the cervical cord with focal cord expansion. ADEM and fulminant multiple sclerosis may show similar findings, the difference being a monophasic illness with ADEM and multiple lesions over time and location in multiple sclerosis.*





(Left) *Sagittal FLAIR MR of the brain shows multiple white matter lesions in this monophasic demyelinating disease. There are lesions in the corpus callosum and brainstem. The new Callen criteria for multiple sclerosis (MS)/acute disseminated encephalomyelitis (ADEM) are the most useful for differentiating a 1st attack of MS from monophasic ADEM: Sensitivity 75%, specificity 95%.* **(Right)** *Axial T2WI MR illustrates abnormal intramedullary hyperintense foci.*





(Left) *Axial FLAIR image shows focal demyelination involving right posterior temporal white matter in this patient with post-vaccination demyelination.* **(Right)** *Sagittal T2W image in this patient with post-vaccination ADEM shows diffuse lower cervical and thoracic cord expansion and T2 hyperintensity.*

Guillain-Barré Syndrome

KEY FACTS

TERMINOLOGY

- Acute inflammatory demyelinating polyradiculoneuropathy (AIDP)
- Autoimmune postinfectious or postvaccinial acute inflammatory demyelination of peripheral nerves, nerve roots, cranial nerves

IMAGING

- Smooth pial enhancement of cauda equina and conus medullaris
  - Nerve roots may be slightly enlarged
  - Conus not enlarged
- Axial T1WI C+ MR shows preferential contrast accentuation of ventral roots in cauda

TOP DIFFERENTIAL DIAGNOSES

- Miller-Fischer syndrome (MFS)
- Chronic polyneuropathies
  - Subacute inflammatory demyelinating polyradiculoneuropathy
  - Chronic inflammatory demyelinating polyradiculoneuropathy
  - Hereditary polyneuropathies
- Carcinomatous or lymphomatous meningitis
- Anterior lumbar radiculopathy

PATHOLOGY

- Antecedent event or "trigger" in 70% of GBS cases
  - Usually follows recent viral illness
- *Campylobacter jejuni* infection can be trigger

CLINICAL ISSUES

- Classically presents with "ascending paralysis"
  - Ascent up to brainstem may involve cranial nerves
  - Respiratory paralysis requiring ventilator in severe cases
- Sensory loss common but less severe



Sagittal T2WI MR in a GBS patient reveals thickened cauda equina ➡. The conus has a normal morphology and exhibits normal signal intensities. Elevated antiganglioside antibody titer confirmed Guillain-Barré syndrome.



Sagittal T1WI C+ FS MR demonstrates smooth linear cauda equina thickening and enhancement ➡. This patient presented with typical acute onset of weakness and areflexia of the distal extremities.

TERMINOLOGY

Abbreviations

- Guillain-Barré syndrome (GBS)

Synonyms

- Acute inflammatory demyelinating polyradiculoneuropathy (AIDP)
- "Ascending paralysis"

Definitions

- Autoimmune postinfectious or postvaccinial acute inflammatory demyelination of peripheral nerves, nerve roots, cranial nerves

IMAGING

General Features

- Best diagnostic clue
  - Smooth pial enhancement of cauda equina and conus medullaris
- Location
  - On imaging, typically cauda equina, especially ventral roots
- Size
  - Nerve roots may be slightly enlarged
- Morphology
  - Symmetric, smooth appearance of roots

CT Findings

- CECT
  - May see symmetric enhancement of lumbar roots

MR Findings

- T2WI
  - Should see normal conus
  - May see slight prominence of root size
- T1WI C+
  - Avid enhancement of cauda equina
    - Roots may be slightly thickened, not nodular
  - Axial images show preferential contrast accentuation of ventral roots in cauda
  - Pial surface of distal cord and conus enhances variably

Nonvascular Interventions

- **Lumbar puncture**

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI ± gadolinium contrast

DIFFERENTIAL DIAGNOSIS

Miller-Fischer Syndrome (MFS)

- GBS variant: Ataxia, areflexia, ophthalmoplegia
- Anti-GQ1b antibody present in serum of > 85% of patients with MFS, but not specific
- May have similar trigger

Chronic Polyneuropathies

- Subacute inflammatory demyelinating polyradiculoneuropathy (SIDP)
  - Patients who fall between 4 weeks and 2 months (SIDP)
- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
  - Slower onset, more protracted course
  - Minimum duration of symptoms until patients reach nadir: 2 months
- Hereditary polyneuropathies
  - Charcot-Marie-Tooth
  - Dejerine-Sottas

Vasculitic Neuropathy

- Polyarteritis nodosa, Churg-Strauss most common
- Cranial and respiratory nerves frequently spared

Acute Transverse Myelitis

- Cranial nerves always spared

Bacterial or Granulomatous Meningitis

- Acute onset
- Fever, headache, + LP

Carcinomatous or Lymphomatous Meningitis

- Enhancement is frequently more nodular than GBS
- Conus deposits frequently cause T2 signal abnormality

Chemical or Postsurgical Arachnoiditis

- Hemorrhage-induced arachnoidal inflammation
- Systemic chemotherapy → vincristine neuropathy

Anterior Lumbar Radiculopathy

- Transient or permanent paraplegia uncommon complication following intrathecal chemotherapy
- Smooth linear enhancement of ventral cauda equina and conus pia

Postradiation Radiculopathy

- History of prior radiation therapy
- Smooth enhancement of both ventral and dorsal cauda equina nerve roots

Physiological Nerve Root Enhancement

- Much more subtle enhancement of normal roots
- Absent clinical syndrome
- May see postoperative up to 6 weeks

PATHOLOGY

General Features

- Etiology
  - Inflammatory (postulated autoimmune or viral) demyelination
    - Antecedent event or "trigger" in 70% of GBS cases
    - Usually follows recent viral illness
    - *Campylobacter jejuni* infection can be trigger
    - 1/3 have antibodies against nerve gangliosides, which cross react with *C. jejuni* lipopolysaccharide constituents
      - *C. jejuni* infection associated with serum antibodies to gangliosides GM1 and GD1a, axonal neuropathy, and greater disability
    - Vaccination (influenza, others) can be trigger in rare cases
  - More tenuous association with prior recent surgery or systemic illness
- Genetics

- May be 1st presentation of genetic or hereditary disorders
  - *17p12* mutation
  - Reported association with HLA typing and Guillain-Barré subtypes (possible HLA DR2)
- Lesions are scattered throughout peripheral nerves, nerve roots, cranial nerves
- Both cell-mediated and humoral mechanisms involved in pathogenesis

Staging, Grading, & Classification

- Spectrum of disease extends to subacute inflammatory demyelinating polyradiculoneuropathy (SIDP)
  - SIDP: Bridge to chronic inflammatory demyelinating polyradiculopathy (CIDP)
    - Both entities exhibit slower onset, protracted course, exacerbations, lesser cranial nerve involvement

Gross Pathologic & Surgical Features

- Thickened nerve roots

Microscopic Features

- Focal segmental demyelination
- Perivascular and endoneural lymphocytic/monocytic (macrophages) infiltrates
- Axonal degeneration in conjunction with segmental demyelination in severe cases
- Antiganglioside antibodies play pathogenic role
  - Nerve injury 2° to complement activation or dysfunction of molecules, such as voltage-gated sodium and calcium channels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Classically presents with "ascending paralysis"
  - Ascent up to brainstem may involve cranial nerves
    - Respiratory paralysis requiring ventilator in severe cases (~ 25%)
  - Other signs/symptoms
    - Sensory loss common but less severe
    - Numbness, tingling
- Distal paresthesias rapidly followed by "ascending paralysis"
  - Frequently bilateral and symmetric
- Autonomic disturbances
- Cranial nerve involvement common
  - Facial nerve involved in up to 50% of cases
  - Ophthalmoparesis in 10-20% of cases
- Association with posterior reversible encephalopathy syndrome (PRES) attributed to autoregulatory loss

Demographics

- Age
  - Typically children, young adults
  - Clinical factors associated with poor outcome
    - Increasing age, presence of preceding diarrhea, severity of disability in early course of disease
- Epidemiology
  - Incidence: 0.6-2.73 cases per 100,000 population per year
  - Affects all ages, races, socioeconomic statuses
  - Most common cause of paralysis in Western countries

Natural History & Prognosis

- Acute flaccid paralysis or distal paraesthesia followed by rapid ascending paralysis
- Cranial nerve involvement common
  - Facial nerve in 50%
  - Ophthalmoparesis in < 10%
- Clinical nadir at 4 weeks
- Most patients somewhat better by 2-3 months
  - 30-50% have persistent symptoms at 1 year
  - Permanent deficits in 5-10%
  - ~ 20% are unable to walk unaided after 6 months
- Relapse (2-10%)
  - Early relapse occurring < 9 weeks after onset more common in patients with GBS
  - 6% develop chronic course resembling CIDP
  - Earliest stages of SIDP, CIDP may be indistinguishable clinically from Guillain-Barré
- Mortality (up to 8%)

Treatment

- Medical management with plasma exchange or intravenous immunoglobulin (IVIG)
  - Severe GBS may benefit from plasma exchange after IVIG in refractory cases
  - Plasma exchange should be considered early in cases with axonal involvement and in recurrent or familial GBS forms
- Corticosteroids given alone do not significantly hasten recovery from GBS or affect long-term outcome
- Intensive care management in severe cases

DIAGNOSTIC CHECKLIST

Consider

- Clinical setting of nerve root enhancement in cauda critical in differential diagnosis

Image Interpretation Pearls

- Anterior cauda nerve root and pial conus enhancement without significant enlargement or nodularity is strong clue

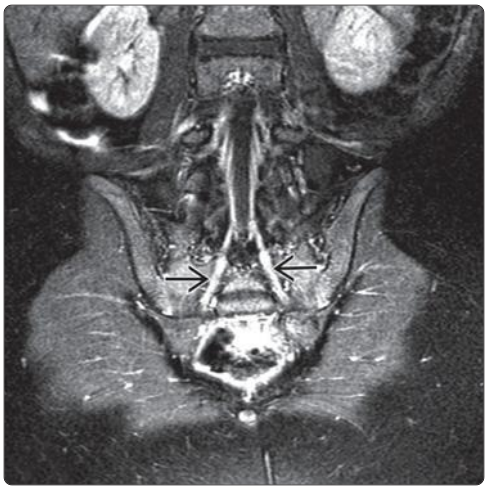
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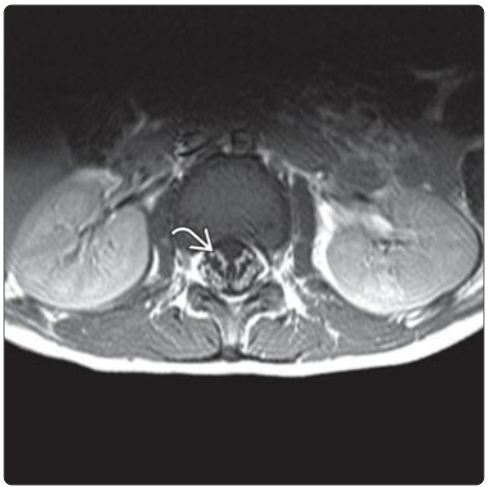
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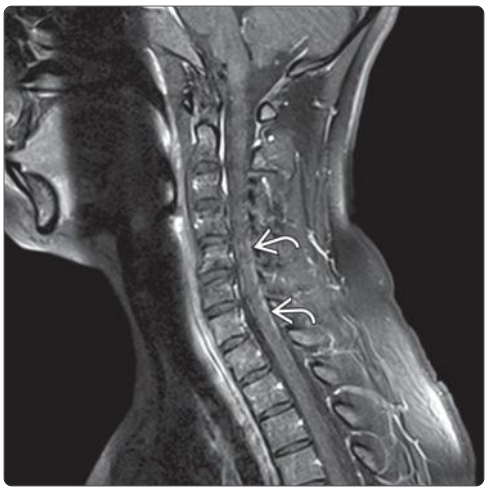


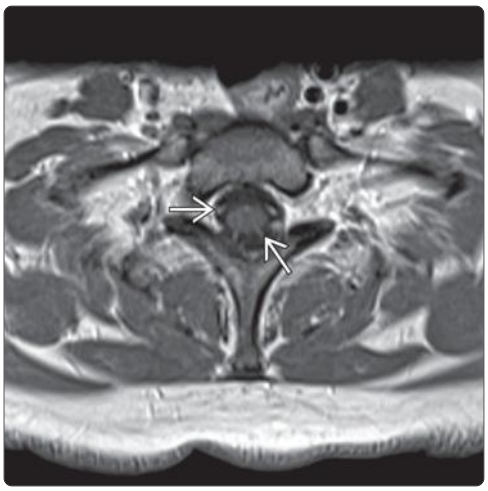
(Left) *Sagittal T1WI C+ MR shows avid enhancement of the ventral conus pia and cauda equina, sparing the dorsal sensory roots. Involvement of the autonomic nervous system, facial weakness, a preceding infectious illness, and the need for mechanical ventilation are more common in GBS as compared to CIDP.* **(Right)** *Axial T1WI C+ MR reveals intense enhancement of the ventral cauda equina. CSF analysis in early stages is helpful to assess chemistry, protein levels, and infectious process.*





(Left) *Coronal T1WI C+ FS MR demonstrates avid enhancement of spinal nerves and lumbar plexus roots in a child with GBS. (Courtesy S. Blaser, MD.)* **(Right)** *Axial T1WI C+ MR reveals extensively enhancing ventral and dorsal nerve roots. Intrathecal and extraforaminal enhancement can be seen in the acute and chronic polyneuropathies. GBS may actually be the 1st presentation of a chronic or hereditary disorder.*





(Left) *Sagittal T1WI C+ FS MR depicts thin pial enhancement along the pial surface of the midcervical cord. This patient had < 4 weeks of weakness, muscle pain, and areflexia involving her distal > proximal extremities. She had no abnormal sensation changes. CSF analysis revealed ↑ protein with 2 white blood cells.* **(Right)** *Axial T1WI C+ MR exhibits avid enhancement and slight thickening of intradural nerves at the cervicothoracic junction.*

CIDP

KEY FACTS

TERMINOLOGY

- Clinically heterogeneous, grossly symmetric, sensory and motor neuropathy evolving as monophasic, relapsing, or progressive disorder
  - Develops over > 8 weeks

IMAGING

- Enlargement and abnormal T2 hyperintensity of nerve roots, plexi, or peripheral nerves
- Spinal nerve roots and peripheral nerves (extraforaminal > intradural)
- Lumbar > cervical, brachial plexus, thoracic/intercostal > cranial nerve

TOP DIFFERENTIAL DIAGNOSES

- Guillain-Barré (AIDP)
- Inherited demyelinating neuropathy
- Neurofibromatosis type 1

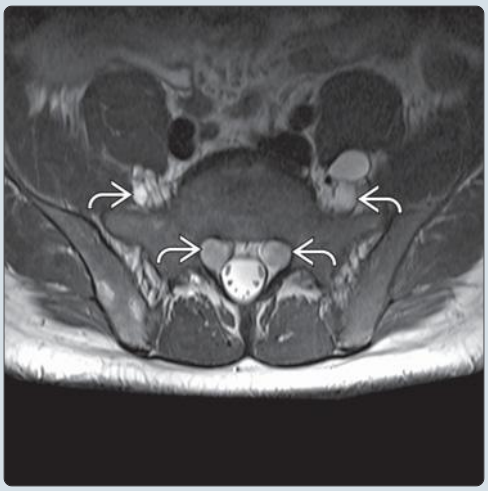
PATHOLOGY

- Autoimmune disease involving cellular and humoral immunity
- Hallmarks of CIDP: Enlarged nerves with "onion bulb" formations, demyelination

CLINICAL ISSUES

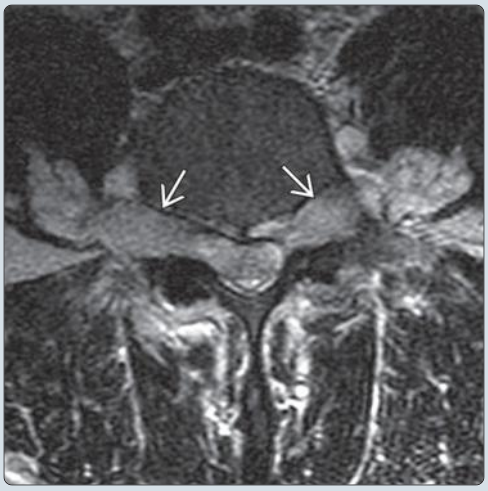
- CIDP is usually a **clinical** diagnosis based on presence of progressive weakness/sensory loss and response to corticosteroid therapy
  - Typical form: Symmetric proximal and distal weakness, sensory loss
- Abnormal EMG/NCV: Key electrophysiologic features → nerve conduction block, slowed conduction velocities suggestive of demyelination
- Diagnosis relies primarily on clinical, electrophysiologic examination supplemented by nerve biopsy





(Left) *Axial T1WI C+ FS MR demonstrates symmetric enlargement and mild enhancement of the lumbar nerve roots. Classically, CIDP presents with sensory and motor symptoms in the distal and proximal segments of the 4 limbs with areflexia, evolving over > 8 weeks. Raised protein concentrations in CSF and heterogeneous slowing of nerve conduction are typical of the condition.* **(Right)** *Axial T2WI MR illustrates mild enlargement and iso- to hyperintensity of bilateral lumbar nerves.*





(Left) *Sagittal T2WI MR reveals enlargement and abnormal T2 hyperintensity of the exiting extradural lumbosacral nerves. Diffuse nerve root hypertrophy and enhancement can be extensive in CIDP.* **(Right)** *Axial T2WI MR exhibits enlargement and abnormal T2 hyperintensity of the exiting extradural lumbosacral nerves. Typical cases associate progressive or relapsing-remitting motor and sensory deficit with increased CSF protein content and electrophysiological features of demyelination.*

TERMINOLOGY

Abbreviations

- Chronic inflammatory demyelinating polyneuropathy (CIDP)

Definitions

- Chronic acquired, immune-mediated demyelinating neuropathy characterized by relapsing or progressive muscle weakness ± sensory loss

IMAGING

General Features

- Best diagnostic clue
  - Enlargement and abnormal T2 hyperintensity of nerve roots, plexi, or peripheral nerves
- Location
  - Spinal nerve roots and peripheral nerves (extraforaminal > intradural)
  - Lumbar > cervical, brachial plexus, thoracic/intercostal > cranial nerve
- Size
  - Nerve size varies; small → very large
- Morphology
  - Focal or diffuse fusiform enlargement of cauda equina, nerve roots/plexi, and peripheral nerves

CT Findings

- NECT
  - Isodense nerve enlargement

MR Findings

- T1WI
  - Isointense enlargement of cauda equina and proximal peripheral nerves
- T2WI
  - Enlargement, abnormal hyperintensity of intradural and extradural spinal nerves/branches
- FLAIR
  - Sagittal FLAIR may reveal hyperintense brain lesions similar to multiple sclerosis
- DWI
  - Diffusion neurography: FLAIR-based diffusion tensor scan that can demonstrate spinal cord and nerve root abnormalities
- T1WI C+
  - Mild to moderate nerve enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic, hypertrophic nerves

Imaging Recommendations

- Best imaging tool
  - T2WI, enhanced coronal and axial T1WI sequences with fat suppression best delineate nerve lesions
  - Brain MR to detect subclinical CNS demyelination

DIFFERENTIAL DIAGNOSIS

Guillain-Barré (AIDP)

- Pial, nerve root enhancement similar to CIDP
- Differs from CIDP in onset duration, clinical course
  - Acute onset of ascending paralysis with relative sensory preservation

Inherited Demyelinating Neuropathy

- Charcot-Marie-Tooth, Dejerine-Sottas neuropathies
- Genetic testing, clinical phenotype help distinguish from CIDP

Neurofibromatosis Type 1

- Diffuse nerve root enlargement, enhancement
- Genetic testing, distinctive clinical stigmata distinguish from CIDP

Lateral Meningocele

- CSF density/signal intensity (not solid) ± foraminal enlargement, dural ectasia
- Usually coexisting NF1 or connective tissue disorder (Marfan syndrome)

PATHOLOGY

General Features

- Etiology
  - Exact pathogenesis of CIDP unclear; involves both cellular and humoral immune factors
    - Chemokines released into CSF from inflamed spinal nerve roots → attract T cells, monocytes, and macrophages to PNS and precipitates release of proinflammatory cytokines
      - ↑ systemic tumor necrosis factor and interleukin → T cell activation
      - ↑ adhesion molecules, matrix metalloproteinases, and chemokines in CSF and nerves for interaction between activated T cells and adhesion molecules on endothelial cells
    - Activated CD4 T cells, antimyelin antibodies/complement cross-react with myelin and PNS antigens
  - Idiopathic or secondary (CIDP with concurrent disease)
    - Connective tissue disease, Sjögren syndrome, inflammatory bowel disease, IgG or IgA monoclonal gammopathy, hepatitis C, human immunodeficiency virus, and lymphoma
    - ± diabetes mellitus
    - Rare cases of CIDP associated with melanoma in which tumor cells share carbohydrate epitopes with Schwann cells
    - Reported in patients with Charcot-Marie-Tooth disease → superimposed inflammatory mechanisms might occur in hereditary neuropathies → contribute to disability
    - Although precise relationship of secondary diseases to CIDP is unclear, neuropathy frequently responds to therapy-like idiopathic CIDP
- Genetics
  - Homozygous genotype for low repeat number of tandem GA in *SH2D2A* gene coding for T-cell-specific adapter protein
    - Could result in defective control and elimination of autoreactive T cells

- Acquired, immune-mediated multifocal demyelinating neuropathy affecting primarily spinal nerves, plexi, and proximal nerve trunks
  - Putative pathogenic mechanisms broadly resemble MS; considered by some to be peripheral counterpart of MS

Gross Pathologic & Surgical Features

- Extensive fusiform nerve enlargement ± gross "onion bulb" formations

Microscopic Features

- Hallmarks of CIDP: Enlarged nerves with "onion bulb" formations, demyelination
  - Macrophage, T-cell infiltration → perivascular inflammatory infiltrates, nerve demyelination and remyelination
  - "Onion bulb" formation 2° to excessive Schwann cell process proliferation → segmental repetitive demyelination/remyelination

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Mixed sensorimotor neuropathy
  - Typical form: Symmetric proximal and distal weakness, sensory loss
    - Rarer atypical form (Lewis-Sumner syndrome)
      - Predominantly uni- or multifocal as well as distal involvement
  - Other signs/symptoms
    - Occasional pure motor neuropathy
    - Symptomatic lumbar stenosis secondary to nerve root hypertrophy
- Other signs/symptoms
  - Monophasic: Progress to nadir then fully recover with treatment and usually do not relapse (7–50%)
  - Relapsing–remitting: Complete remissions between relapses (20–35%)
  - Chronic progressive: Progressively deteriorate until treatment is given
- Clinical profile
  - CIDP is usually a **clinical** diagnosis based on presence of progressive weakness/sensory loss and response to corticosteroid therapy
    - ± concurrent CNS demyelination (frequently subclinical)
  - Electrophysiologic and laboratory abnormalities
    - Abnormal EMG/NCV: Key electrophysiologic features → nerve conduction block, slowed conduction velocities suggestive of demyelination
      - F-wave conduction velocities decrease in parallel with increase in diameters of some spinal nerve roots
    - ↑ CSF protein; however, protein content can be normal in at least 10% of patients
      - Total protein in CSF reported to show significantly positive correlation with diameters of spinal nerve roots
  - Diagnosis relies primarily on clinical, electrophysiologic examination supplemented by nerve biopsy
  - ≥ 50% of CIDP patients have atypical features; asymmetry, distal weakness, sensory loss only
    - Heterogeneous distribution of symptoms, disease course, and disability → underdiagnosis

Demographics

- Age
  - Adult > childhood
- Gender
  - M = F
- Epidemiology
  - Incidence (1.6/100,000/year) and prevalence (8.9/100,000)

Natural History & Prognosis

- Clinical course: Chronic progressive, step-wise progressive, or recurrent behavior
  - Prolonged remission/cure is exception, not rule
  - Atypical and secondary CIDP respond similarly to classic CIDP
- Average disease duration: 7.5 years

Treatment

- Immunomodulation or immunosuppression therapy
  - Prednisolone therapy, plasmapheresis, or intravenous immunoglobulin (IVIg)
  - Immunosuppressive therapies like methotrexate, azathioprine, cyclophosphamide, cyclosporine A, mycophenolate mofetil have been used anecdotally
  - Immunomodulatory agents, such as β interferon, reported beneficial

DIAGNOSTIC CHECKLIST

Consider

- Consider CIDP in differential of nerve root/peripheral nerve enlargement

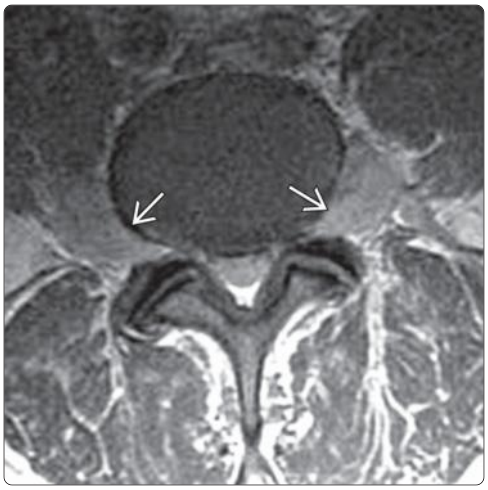
Image Interpretation Pearls

- MR findings imperfectly correlate with clinical disease activity/severity, laboratory findings

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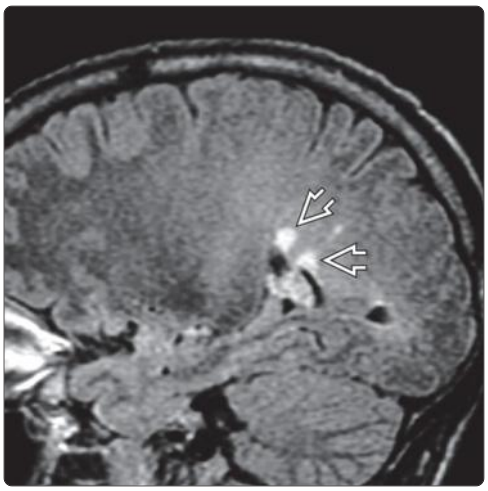
(Left) *Sagittal T1WI C+ MR reveals massive cauda equina enlargement and prominent abnormal enhancement. Axon loss associated with demyelination is the most important factor of disability, resistance to treatment.* **(Right)** *Axial T1WI C+ MR depicts enlargement and abnormal enhancement of exiting extradural lumbosacral nerves. Blood–nerve barrier breakdown at the root level can cause contrast enhancement. Root hypertrophy also may cause stenosis symptoms.*





(Left) *Sagittal T1 C+ MR of the cervical spine demonstrates marked hypertrophy and enhancement of all exiting cervical nerve roots. In the cervical region, 5.0 mm is considered an adequate cut-off value of the spinal nerve root diameter, discriminating CIDP patients from controls.* **(Right)** *Axial T2WI MR shows symmetric hyperintensity and enlargement of bilateral cervical nerve roots. Demyelination and remyelination are pathological hallmarks of CIDP.*





(Left) *Coronal T2WI MR demonstrates marked abnormal fusiform enlargement and T2 hyperintensity of the lumbar extradural nerve roots. High signal shown on MR includes dura mater and cerebrospinal fluid, and their diameters are usually larger than the actual size of spinal nerve roots.* **(Right)** *Sagittal FLAIR MR exhibits periventricular ovoid hyperintensities in a patient with a typical case of marked fusiform CIDP nerve enlargement with brain demyelination.*

Sarcoidosis

KEY FACTS

TERMINOLOGY

- Neurosarcoidosis
- Noncaseating granulomatous disease of spine and spinal cord

IMAGING

- Combination of leptomeningeal and peripheral intramedullary (IM) mass-like enhancement suggestive of spinal sarcoidosis
- Intramedullary
  - Fusiform cord enlargement
  - Enhancing intramedullary masses
  - Focal or diffuse T2-hyperintense lesions
- Intradural extramedullary
  - Leptomeningeal enhancement
  - Dural masses
- Extradural
  - CT/radiographs: Sclerotic or mixed lytic and sclerotic lesions

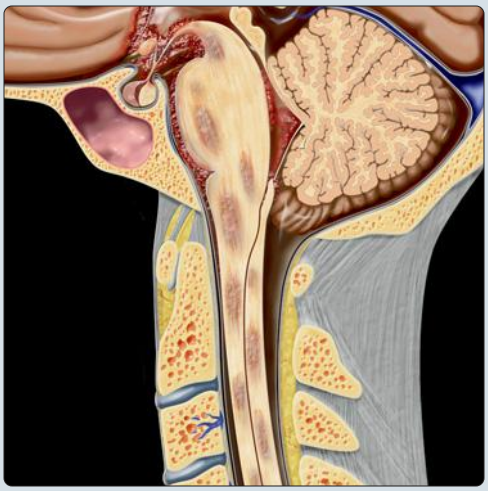
TOP DIFFERENTIAL DIAGNOSES

- Intramedullary neoplasm
- Idiopathic transverse myelitis
- Lymphoma
- Intradural metastases
- Multiple sclerosis

CLINICAL ISSUES

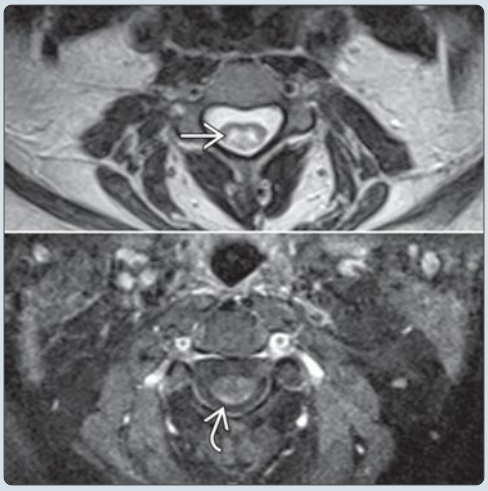
- Clinical CNS involvement in patients with sarcoidosis (5%)
  - Spinal intramedullary sarcoidosis (< 1%)
- Lower extremity weakness, paresthesia
- Bladder and bowel dysfunction
- Treatment: Intravenous ± oral corticosteroids
  - Immunosuppressive therapy

(Left) *Graphic depicts multiple intramedullary sarcoid granulomas in brainstem & upper cervical cord. The intramedullary, intradural extramedullary, & extradural regions can be involved. Rare findings, such as calcifications & cyst formation, have also been described.* **(Right)** *Sagittal T1WI C+ MR demonstrates diffuse linear leptomeningeal enhancement with areas of nodularity. There is intracranial involvement with sarcoidosis. Bulky leptomeningeal disease is unusual with neurosarcoidosis.*





(Left) *Axial T2WI MR (top) shows intramedullary hyperintensity. Axial T2WI MR (bottom) reveals irregular leptomeningeal enhancement along the cord.* **(Right)** *Sagittal T1WI C+ MR exhibits multiple enhancing subarachnoid nodules interspersed among the cauda equina. Intradural extramedullary lesions are usually represented by leptomeningeal sarcoidosis infiltration, present in up to 60% of spinal cord lesions.*





TERMINOLOGY

Definitions

- Noncaseating granulomatous disease of spine and spinal cord

IMAGING

General Features

- Best diagnostic clue
  - Combination of leptomeningeal and peripheral intramedullary (IM) mass-like enhancement suggestive of spinal sarcoidosis
- Location
  - Intramedullary
    - Cervical, upper thoracic
  - Intradural extramedullary
    - Cauda equina
    - Dural involvement without segmental predilection
  - Extradural
    - Vertebral
      - Lower thoracic and upper lumbar spine
      - ~ 4% patients with sarcoidosis have bone lesions: Small bones of hands & feet involved most often
      - Spine involvement is **rare** but usually symptomatic → back pain associated with activity

Radiographic Findings

- Lytic, compact, or diffuse lesions
  - Lytic lesions may develop sclerotic rim with healing
  - Osteosclerotic lesions are rare
- Radiographic osseous abnormalities (1-20%)

CT Findings

- Bone CT
  - Sclerotic or mixed lytic and sclerotic lesions

MR Findings

- T1WI
  - Fusiform cord enlargement
  - Cord atrophy in late stages
  - Iso- to hypointense lesions
- T2WI
  - Focal or diffuse hyperintense lesions
- STIR
  - Lesions hyperintense
- T1WI C+
  - Enhancing dural masses
    - Variable cord and nerve root compression
  - Leptomeningeal enhancement
    - Smooth or nodular
  - Peripheral IM enhancement
    - Broad-based contact with cord surface
  - Mass-like IM enhancement
    - Focal or multifocal
  - Diffuse or patchy IM enhancement
  - Vertebral lesions typically enhance
- Regression of findings during steroid therapy
  - Decreased enhancement
  - Poor correlation with clinical response
- Rare: IM calcifications or cysts

Nuclear Medicine Findings

- Bone scan
  - Multifocal uptake of radiotracer in vertebral sarcoid lesions
- PET
  - Increased metabolic activity in cord lesions
    - 18F FDG PET detects active granulomatous foci with higher sensitivity (87%) than Ga-67 scintigraphy (67%)
- Gallium scan
  - Increased central nervous system uptake
    - < 5% of patients

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI and T2WI MR with gadolinium

DIFFERENTIAL DIAGNOSIS

Intramedullary Neoplasm

- Lack of leptomeningeal enhancement
- Typically holocord enhancement on axial imaging
- Peritumoral edema
- Cystic ± hemorrhagic components

Multiple Sclerosis

- No leptomeningeal enhancement
- Central enhancement in peripheral lesions
- < 1/2 of cross-sectional area of spinal cord
- Periventricular brain lesions

Idiopathic Transverse Myelitis

- Peripheral enhancement in central lesions
- 3-4 segments in length
- No leptomeningeal or dura enhancement
- Diagnosis of exclusion

Lymphoma

- Protean manifestations, may be indistinguishable from sarcoid
- Nodular leptomeningeal enhancement
- Look for vertebral & epidural masses

Intradural Metastases

- Smooth enhancement along cord or roots
- Multifocal discrete nodules, focal masses
- Thickening of cauda equina

Infarction, Spinal Cord

- Immediate onset, ventral location

PATHOLOGY

General Features

- Etiology
  - Exaggerated T-cell-mediated immune response suspected
    - Self-antigen or infectious agents
  - Faulty suppression mechanism
- Genetics
  - Major histocompatibility complex or complement receptor gene on macrophages may be involved
    - Exaggerated response to chemotactic factors
- Associated abnormalities

- Intracranial sarcoidosis
- Systemic sarcoidosis

- Syringomyelia may result from blood-spinal cord barrier breakdown, impairment of extracellular fluid drainage, & leakage of subarachnoidal CSF into cord

Staging, Grading, & Classification

- 4 stages of intraspinal sarcoidosis correlating with histologic stages
  - 1: Early inflammation → linear leptomeningeal enhancement
  - 2: Centripetal spread of leptomeningeal inflammation through perivascular spaces → faint parenchymal enhancement & swelling
  - 3: Less prominent swelling with focal or multiple enhancement
  - 4: Resolution of inflammatory process with normal size or atrophy of cord & no enhancement

Gross Pathologic & Surgical Features

- Granuloma resembles neoplasm
- Intraoperative frozen sections may be misinterpreted for gliomas

Microscopic Features

- Epithelioid cell noncaseating granulomas
  - Surrounded by lymphocytes and mononuclear phagocytes
  - Negative stains for infectious agents
  - Granulomas smaller compared to those in systemic disease
    - Fewer multinucleated giant cells
- Areas of neural tissue infarction
- Perivascular lymphocytic infiltrate
  - Also involving adventitia of vessels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Lower extremity weakness
  - Paresthesia
  - Radiculopathy
  - Bladder and bowel dysfunction
- Other signs/symptoms
  - > 10-20% of patients with sarcoidosis have no clinical and radiographic findings of sarcoidosis
  - ↑ angiotensin-converting enzyme (ACE) levels
    - Can be used to monitor disease progress
  - ↑ serum calcium levels & urinary calcium excretion in 20% of patients secondary to ↑ calcitriol production by sarcoid granuloma
- Clinical profile
  - Acute to subacute onset

Demographics

- Age
  - 20-40 years old
- Gender
  - Male > female in spinal sarcoidosis
  - Slight female predominance in systemic sarcoidosis
- Ethnicity
  - Sarcoidosis relatively more common in Northern Europeans and African Americans
  - Rare in Asians
- Epidemiology
  - Incidence of sarcoidosis
    - 10-40 per 100,000 in USA and Europe
  - Clinical CNS involvement in patients with sarcoidosis (5%)
    - Spinal intramedullary sarcoidosis (< 1%)
      - Intramedullary lesions are rarely 1st manifestation of sarcoidosis, especially when there is no systemic involvement
  - Imaging evidence of CNS involvement (10%)
  - CNS involvement at autopsy (15-25%)
  - Isolated neurosarcoidosis (1.5%)

Natural History & Prognosis

- Favorable response to corticosteroid therapy
  - 2/3 of neurosarcoidosis
- 1/3 with remitting relapsing course

Treatment

- Intravenous ± oral corticosteroids
  - Chronic steroid therapy may be required
- Immunosuppressive therapy
  - Treatment correlates with improvement in CSF T-lymphocyte subpopulation ratios
- Low-dose radiation (rarely used)
  - 1-3,000 rads over days to weeks
- Operative intervention may be indicated in any case of spinal sarcoidosis that either requires tissue biopsy for diagnosis or is associated with progressive neurologic symptoms

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Protean imaging manifestations
  - Mimicking multiple spinal pathologies
  - Typically, systemic disease present; rarely, intramedullary sarcoid may be initial presentation

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5. Varron L et al: Spinal cord sarcoidosis: report of seven cases. Eur J Neurol. 16(3):289-96, 2009
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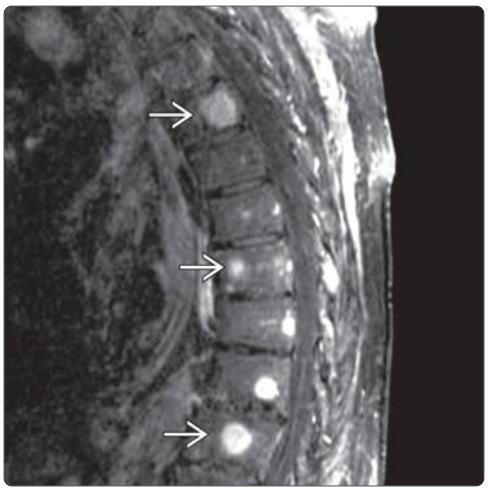


(Left) *Sagittal T2WI MR shows diffuse leptomeningeal nodular hypointensity and thickening of the cauda equina* ⇒. **(Right)** *Sagittal T1WI C+ MR demonstrates marked nodular leptomeningeal enhancement along the cauda equina* ⇒. *Axial enhanced images can be used to confirm the intradural extramedullary nature of the distal cord and cauda involvement. Correlation between symptom resolution and resolution of imaging findings is poor, especially with spinal cord lesions.*





(Left) *Sagittal T2WI MR reveals intramedullary hyperintensity in the distal cord* ⇒, *which is mildly expanded. Note subtle patchy intramedullary enhancement* ⇒. *At clinical presentation, there may be faint parenchymal enhancement and diffuse swelling or less prominent swelling and possible normal-sized spinal cord with focal or multiple areas of enhancement.* **(Right)** *Axial T2WI MR shows intramedullary T2 hyperintensity and cord expansion* ⇒.





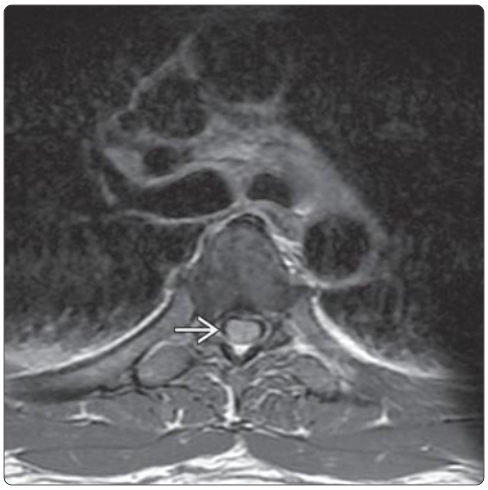
(Left) *Sagittal T1WI C+ MR with fat suppression illustrates "multiple vertebral sarcoid" lesions with homogeneous enhancement* ⇒. *Osseous involvement is reported in 1–13% of patients, but the actual frequency may be higher as most of the osseous lesions are asymptomatic.* **(Right)** *Sagittal T1WI MR shows discrete, hypointense* ⇒, *minimally enhancing* ⇒ *lesions in the vertebral bodies. These treated sclerotic lesions with sclerotic margins may extend into the pedicles and paraspinal region.*

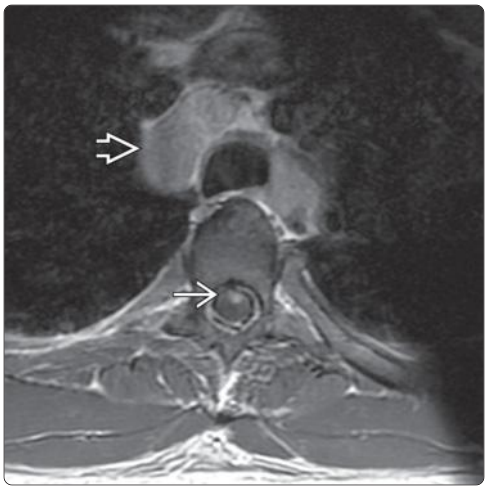
(Left) *Sagittal T2WI MR shows diffuse enlargement of the upper and midthoracic cord with increased T2 signal. This longitudinally extensive increased signal is nonspecific. Transbronchial biopsy of the lymph nodes revealed noncaseating granulomata consistent with sarcoidosis.* **(Right)** *Sagittal T1 C+ MR shows multiple intramedullary enhancing lesions centered in the thoracic cord.*



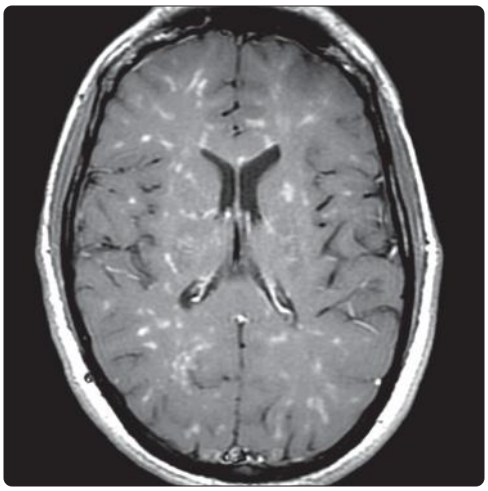


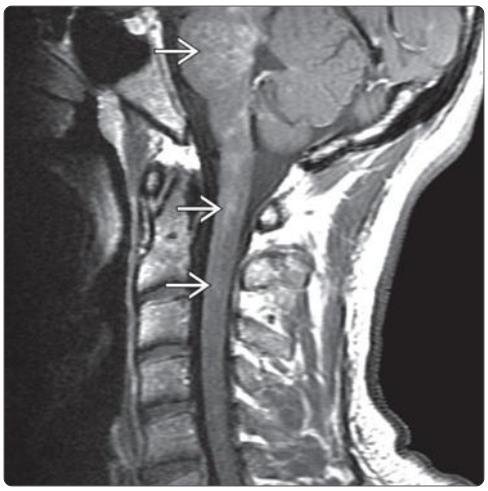
(Left) *Axial T1 C+ MR shows focal homogeneous enhancement of 1 of multiple lesions within the thoracic cord in this patient with sarcoidosis. Differential considerations are many, including metastatic disease and lymphoma.* **(Right)** *Axial T1 C+ MR show a small focus of enhancement within the ventral thoracic cord due to sarcoidosis. Note the enlarged mediastinal lymph nodes.*



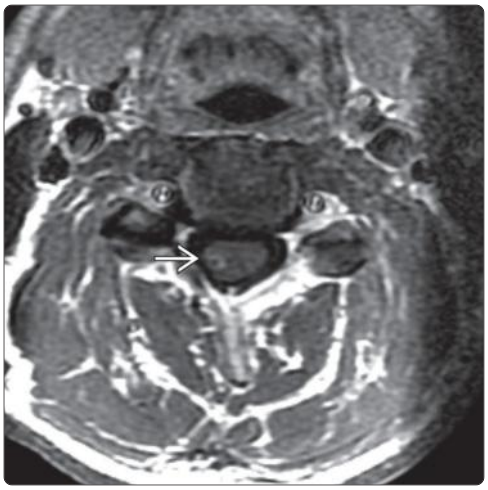


(Left) *Axial T1 C+ MR shows a miliary pattern of enhancement throughout the brain parenchyma with no mass effect or edema in this case of neurosarcoidosis. Lymphoma and TB may also give such a pattern.* **(Right)** *Sagittal T1 C+ MR shows punctate and linear enhancement within the brainstem and extending into the upper cervical spinal cord.*







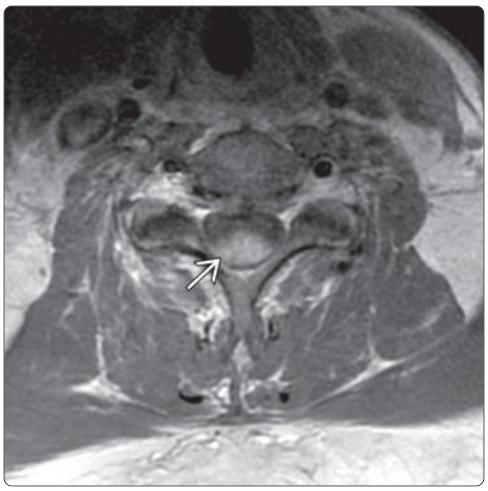


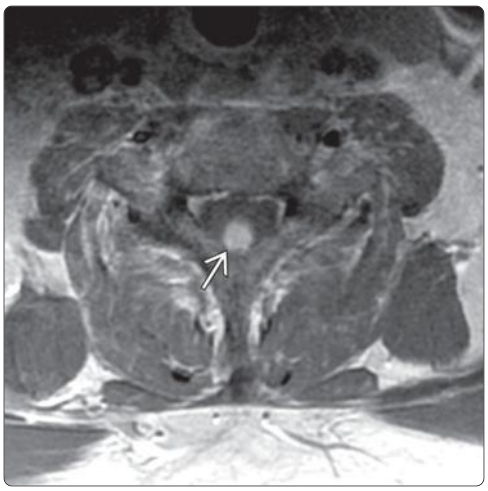
(Left) *Sagittal STIR MR shows mild patchy increased signal from the upper cervical cord in this patient with path proven neurosarcoidosis.* **(Right)** *Axial T1 C+ MR shows a small focus of enhancement involving the right lateral cervical cord.*





(Left) *Sagittal STIR MR shows diffuse cord edema extending from C2 inferiorly to the upper thoracic spinal cord. There is slight cord expansion.* **(Right)** *Sagittal T1 C+ MR shows a large area of diffuse enhancement of the cervical cord. Patient has unrelated prior anterior fusion at C6-C7.*





(Left) *Axial T1 C+ MR shows a large area of diffuse enhancement involving the dorsal cervical cord. Patient had spinal cord biopsy and path-proven neurosarcoidosis.* **(Right)** *Axial T1 C+ MR shows a nodular area of enhancement involving the posterior aspect of the lower cervical cord due to neurosarcoidosis. Finding is nonspecific, and lymphoma is a primary consideration.*

Subacute Combined Degeneration

KEY FACTS

TERMINOLOGY

- Vitamin B_{12} deficiency produces selective degeneration of dorsal ± lateral spinal cord columns

IMAGING

- Longitudinal dorsal cord T2 signal abnormality
- Axial image → "inverted V" or "inverted rabbit ears" within dorsal spinal cord
- ± mild dorsal column enhancement

TOP DIFFERENTIAL DIAGNOSES

- Infectious myelitis
  - HIV vacuolar myelopathy
  - Varicella-Zoster/herpes
- Inflammatory demyelination
  - Multiple sclerosis
  - Acute disseminated encephalomyelitis
- Acute transverse myelitis
  - Acute presentation → diffuse multisegmental cord hyperintensity, swelling
- Infarction, spinal cord
  - Hyperacute presentation

PATHOLOGY

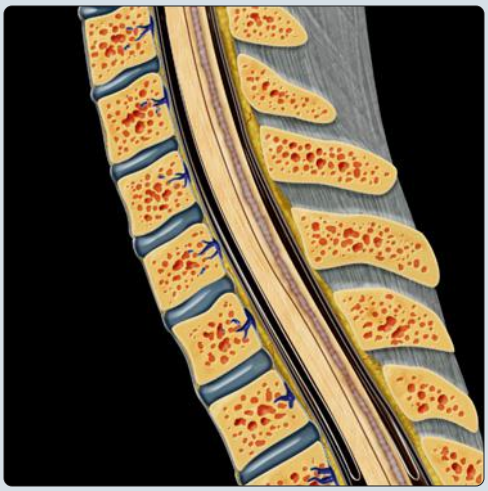
- Malabsorption (most common)
  - Metabolic conditions: Copper deficiency, nitrous oxide anesthesia susceptibility, transcobalamin II deficiency

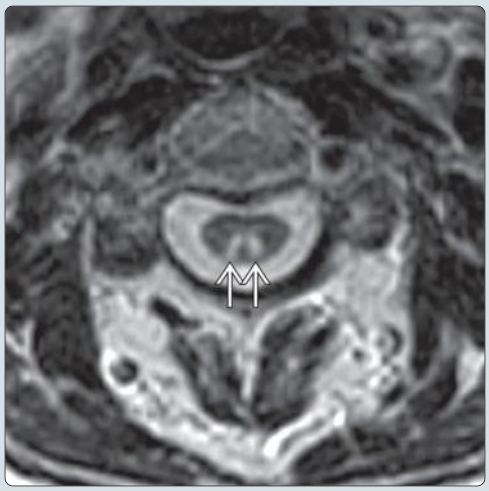
CLINICAL ISSUES

- Paraesthesia, stiffness, mild sensory ataxia, loss of position and vibration sense, spasticity, hyperreflexia
- Degree of resolution of clinical symptoms is inversely proportional to their duration and severity

DIAGNOSTIC CHECKLIST

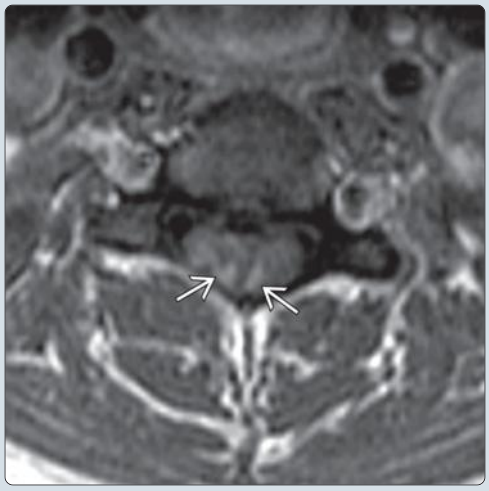
- Neurologic findings may precede anemia and may not completely resolve following treatment





(Left) *Graphic shows dorsal column demyelination 2° to vitamin B_{12} deficiency. Major clinical findings of dysesthesia, gait disturbance and impairment of position, and vibration sense in the lower ± upper limbs are 2° to dorsal column, lateral corticospinal tract, and sometimes lateral spinothalamic tract dysfunction.* **(Right)** *Axial T2WI MR reveals T2 hyperintensity confined to the dorsal columns in an "inverted V" configuration, characteristic of B_{12} deficiency.*





(Left) *Sagittal T1WI C+ MR reveals vague dorsal intramedullary enhancement. Lack of cobalamin (required as a cofactor for conversion of methylmalonyl-CoA to succinyl-CoA) leads to accumulation of methylmalonyl-CoA, causing ↓ in normal myelin synthesis and incorporation of abnormal fatty acids into neuronal lipids.* **(Right)** *Axial MR reveals patchy enhancement in bilateral dorsal columns, which reflects breakdown of the blood-cord barrier caused by perivascular demyelination and inflammation.*

TERMINOLOGY

Abbreviations

- Subacute combined degeneration (SCD)

Definitions

- Vitamin B_{12} deficiency produces selective degeneration of dorsal ± lateral spinal cord columns

IMAGING

General Features

- Best diagnostic clue
  - Mild spinal cord enlargement, abnormal T2 hyperintensity within dorsal columns
- Location
  - Dorsal ± lateral columns of thoracic ± cervical cord
- Size
  - Imaging abnormality confined to dorsal spinal tracts
- Morphology
  - Mild spinal cord enlargement, abnormal signal intensity within dorsal ± lateral columns only

MR Findings

- T1WI
  - Mild spinal cord enlargement, dorsal spinal cord hypointensity
- T2WI
  - Hyperintensity confined to dorsal ± lateral columns
    - Longitudinal dorsal cord T2 signal abnormality
    - Axial image → "inverted V" or "inverted rabbit ears" within dorsal spinal cord
- T1WI C+
  - ± mild dorsal column enhancement

Other Modality Findings

- Brain MR imaging
  - ± white matter T2 hyperintensities
  - May see severe brain volume loss that improves following parenteral B_{12} therapy in infants

Imaging Recommendations

- Best imaging tool
  - Multiplanar T2WI to confirm localization in dorsal columns, exclude cord infarct or spondylosis

DIFFERENTIAL DIAGNOSIS

Infarction, Spinal Cord

- Hyperacute presentation, motor > sensory symptoms
- Predominantly ventral cord or central gray matter signal changes

Contusion-Hematoma, Spinal Cord

- Cord swelling, T2 hyperintensity ± hemorrhage
- Associated fracture, soft tissue injury
- History, clinical findings determine diagnosis

Inflammatory Demyelination

- Multiple sclerosis
- Acute disseminated encephalomyelitis
- Lesions more focal, patchy than B_{12} deficiency, do not show specificity for lateral or dorsal columns
- Characteristic clinical presentation

Infectious Myelitis

- HIV vacuolar myelopathy, Varicella-Zoster, Lyme disease
- Imaging findings may be identical to B_{12} deficiency
- Clinical, laboratory findings help distinguish

Acute Transverse Myelitis

- Acute (nontraumatic) presentation → diffuse multisegmental cord hyperintensity, swelling
- Idiopathic or disease-associated; clinical and laboratory findings may help distinguish cause

PATHOLOGY

General Features

- Etiology
  - Malabsorption (most common)
    - Pernicious anemia (antibody mediated intrinsic factor disorder) most common cause of vitamin B_{12} malabsorption in USA
      - Immune-mediated process → destruction of gastric parietal cells →
      - Achlorhydria, atrophic gastritis, and decreased availability of intrinsic factor
      - Mean age 60 years; often with other autoimmune disorders, such as Graves disease
    - Other causes include post-gastrectomy or ileal resection, fish tapeworm (*D. latum*) infestation, Crohn disease, celiac disease, bacterial overgrowth in intestinal blind loops
  - Other metabolic conditions (nitrous oxide anesthesia susceptibility, transcobalamin II deficiency)
    - Nitrous oxide (N_2O) inactivates B_{12} → accumulation of methylmalonic acid → myelin toxicity
      - Irreversible oxidization of active reduced B_{12} form to inactive form → excreted
      - May bring about manifestations of B_{12} deficiency
  - Inadequate B_{12} intake (rare)
    - Infants of strict vegetarian mothers, vegans
  - Copper deficiency myelopathy (CDM)
    - Risk factors include previous upper gastrointestinal surgery, zinc overload, and malabsorption syndromes → impaired copper absorption in upper gastrointestinal tract
    - Spinal MR reported abnormal in 47% → high T2 signal in posterior cervical and thoracic cord
- Genetics
  - Serum transcobalamin II (TC-II) transporter deficiency in some patients
- Associated abnormalities
  - Megaloblastic anemia
    - > 1/4 of patients with neurologic syndromes have normal complete blood count
  - Extramedullary hematopoiesis (severe anemia)
  - Brain derangements; dementia, psychiatric disorders, optic neuropathy
  - Other (rare) sites of demyelination: Peripheral nerves, optic nerves, pyramidal and spinocerebellar tracts, cerebrum/cerebellum
- Vitamin B_{12} (cobalamin) mostly found in meat, lacking in most vegetables

- B_{12} storage mainly in liver; effects of deficiency may not be appreciated until stores depleted
- B_{12} is coenzyme for 2 important biochemical enzymatic reactions
  - Homocysteine methylation → methionine
  - Methylmalonyl coenzyme A + succinate → succinyl coenzyme A + methylmalonic acid
- Methylmalonic acid accumulation believed to cause myelin toxicity → dorsolateral cord demyelination (myelin sheaths + axons)
  - → focal myelin swelling progresses to vacuolization → axonal loss, Wallerian degeneration of posterior and lateral columns

Gross Pathologic & Surgical Features

- Gray discoloration of posterior ± lateral columns
- ± patchy demyelination in cerebral white matter

Microscopic Features

- Early neuropathological findings: Myelin sheath swelling of cervical posterior columns (funiculus gracilis and spinocerebellar tracts), lateral columns
  - Separation of myelin lamellae and formation of intramyelinic vacuoles, leading eventually to complete destruction of myelin sheaths
- Late sequelae: Myelin sheath + axonal degeneration/loss → long tract wallerian degeneration
  - Marked gliosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Paraesthesia, stiffness, numbness or tingling of limbs (earliest neurological manifestation)
  - Mild sensory ataxia, loss of position and vibration sense, spasticity, hyperreflexia and positive Babinski sign
  - Often symmetric, distal → proximal
  - CDM: Progressive sensory ataxia and weakness in lower and upper limbs
- Other signs/symptoms
  - Atypical clinical features: Motor symptoms, bladder dysfunction, bronchospasm
  - Bladder symptoms occur in advanced cases of SCD
    - Detrusor areflexia and neurogenic detrusor overactivity with high-pressure voiding occur and respond to B_{12} therapy
- Clinical profile
  - Insidious subacute course of clinical symptom onset; functional disturbances increase fairly rapidly → severe disability within weeks to months
    - Cord symptoms appear 1st with motor (spastic paraparesis, gait unsteadiness) and sensory (paraesthesia, absent reflexes, loss of joint position and vibration sense) findings
    - Mental status decline may follow → progressive psychomotor regression (confusion, depression, delusions, mental slowness)
- Laboratory abnormalities
  - Macrocytic anemia (MCV > 100)
  - ↓ plasma B_{12}
  - Abnormal electroneurography, electromyography, and evoked potentials
  - CDM: Low serum copper and ceruloplasmin levels, low urinary copper levels

Demographics

- Age
  - Diagnosis in 5th-8th decades; uncommonly identified in infants, young adults
  - CDM: Frequently present in 5th and 6th decades
- Gender
  - M ≥ F
  - CDM: More common in women (F:M = 3.6:1)
- Ethnicity
  - Pernicious anemia more common in Scandinavian and "English-speaking" populations, but found within all racial groups

Natural History & Prognosis

- Spontaneous improvement without treatment uncommon
- Variable clinical improvement following B_{12} therapy
  - Degree of resolution of clinical symptoms is inversely proportional to their duration and severity
- Treatment arrests degenerative process but does not restore destroyed neural fibers

Treatment

- Address treatable causes
- Lifelong parenteral B_{12} administration
- Avoiding N_2O anesthesia in vulnerable patients
- CDM: Copper supplementation and modification of any risk factors → hematological normalization and neurological improvement or stabilization

DIAGNOSTIC CHECKLIST

Consider

- Neurologic findings may precede anemia
- Imaging changes may not completely resolve following treatment

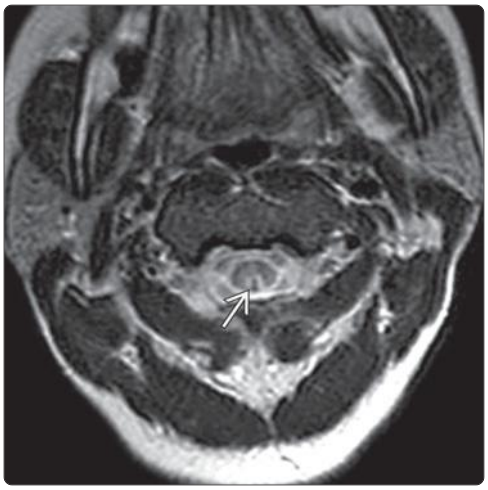
Image Interpretation Pearls

- T2 hyperintensity confined to dorsal columns highly suggestive of diagnosis; confirm with laboratory studies

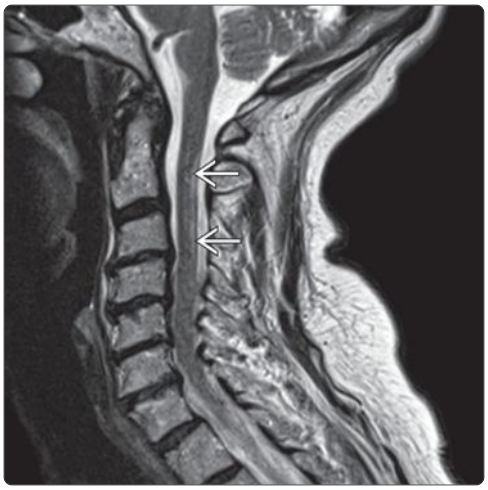
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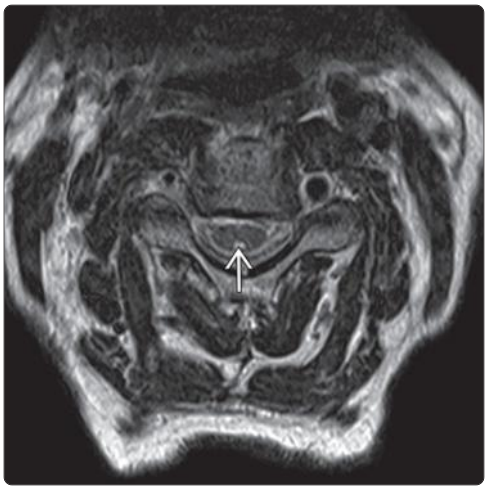
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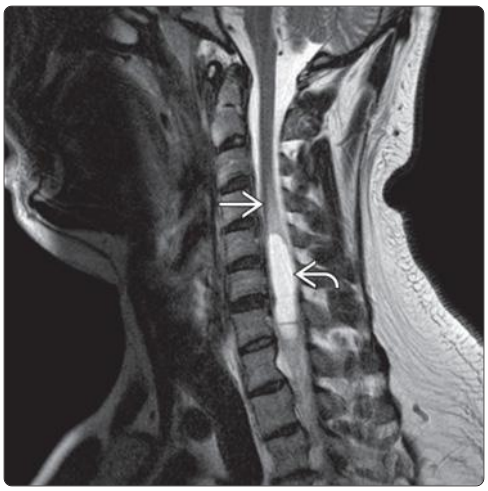


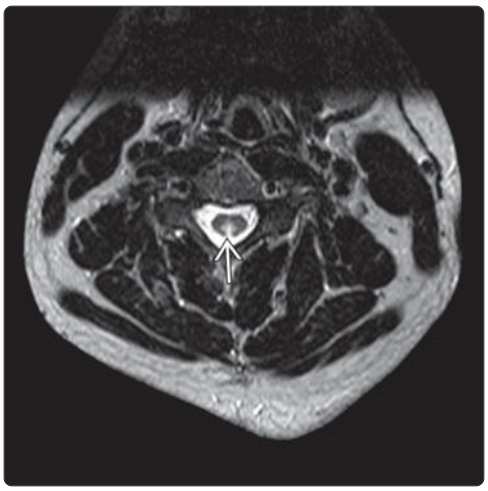
(Left) *Sagittal T2WI MR demonstrates ill-defined hyperintensity in a mildly enlarged mid cervical cord. The abnormal signal intensity in the dorsal columns is usually contiguous in length over several vertebral bodies.* **(Right)** *Axial T2WI MR exhibits the characteristic "inverted V-shaped" T2 hyperintensity along the dorsal columns, which is highly suggestive of SCD. Immediate treatment with parenteral B_{12} may improve symptoms but imaging abnormalities may not completely resolve.*





(Left) *Sagittal T2WI MR shows linear hyperintensity in the dorsal cervical cord due to SCD in this patient with hypocupremia. Copper deficiency myelopathy is a treatable cause of noncompressive myelopathy, which closely mimics SCD due to B_{12} deficiency.* **(Right)** *Axial T2WI MR illustrates focal hyperintensity in the dorsal columns due to SCD in this patient with hypocupremia.*





(Left) *Sagittal T2WI MR shows a post-traumatic syrinx with hyperintensity cephalad to the lesion in keeping with wallerian degeneration (WD). WD is seen in the dorsal columns above the lesion (which can mimic SCD) and in corticospinal tracts below the lesion.* **(Right)** *Axial T2WI MR shows hyperintensity in the dorsal columns above the level of the lesion. The WD above the injury indicates complete or partial interruption of the ascending axons in the posterior columns.*

Chronic Recurrent Multifocal Osteomyelitis

KEY FACTS

TERMINOLOGY

- Autoinflammatory disorder of bone with fever, multifocal painful nonpyogenic bone lesions, relapsing and remitting course in children and adolescents
- Adult disease is SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis)

IMAGING

- Spine involvement uncommon (3% of lesions)
- Most common site complicated by pathologic fracture

TOP DIFFERENTIAL DIAGNOSES

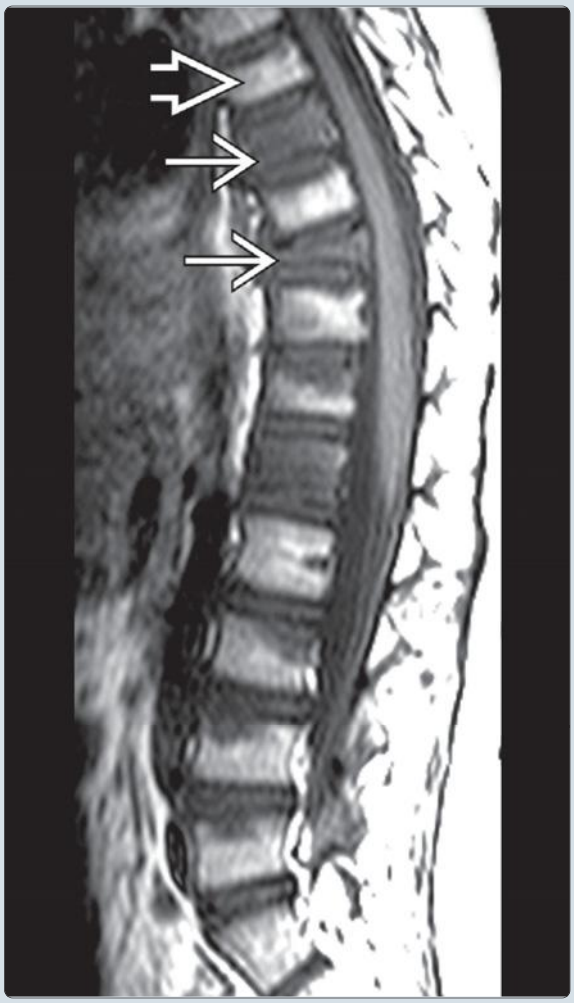
- Septic multifocal osteomyelitis
- Langerhans cell histiocytosis
- Ewing sarcoma
- Juvenile idiopathic arthritis

PATHOLOGY

- CRMO part of heterogeneous group of disorders of innate immune system (autoinflammatory disease)
- Mutations in either pyrin or tumor necrosis factor receptor superfamily
- Major criteria: Imaging-proven osteolytic/sclerotic bone lesion, multiple bone lesions, pustulosis palmoplantaris or psoriasis, sterile bone biopsy
- Minor criteria: Normal blood cell count, ESR mild elevation, course > 6 months, hyperostosis, other autoimmune disease

CLINICAL ISSUES

- NSAID, steroids
- Infliximab and bisphosphonates have also shown clinical improvement



Sagittal T1WI MR shows multiple areas of ↓ signal in multiple vertebral bodies without paraspinal or epidural soft tissue. Compression fractures are present at multiple levels ⇨, some with normal marrow due to old lesions ⇨.



Sagittal STIR MR shows increased signal lesions within multiple vertebral bodies. Multiple pathologic compression fractures are present. Intervening disc signal is normal, and there is no paraspinal abnormality.

TERMINOLOGY

Abbreviations

- Chronic recurrent multifocal osteomyelitis (CRMO)

Synonyms

- Chronic sclerosing osteomyelitis, condensing osteitis, primary chronic osteomyelitis, pustulotic arthrosteitis, lymphoplasmacellular osteomyelitis
- Autosomal recessive syndromic form of disease is Majeed syndrome (OMIM 609628)

Definitions

- Autoinflammatory disorder of bone with fever, multifocal painful nonpyogenic bone lesions, relapsing and remitting course in children and adolescents
- Adult disease is SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis)

IMAGING

General Features

- Best diagnostic clue
  - Involvement of multiple noncontiguous vertebral bodies with sparing of intervertebral disc
- Location
  - Spine involvement uncommon (3% of lesions)
    - Most common site complicated by pathologic fracture
  - Thoracic most common spine site > lumbar > cervical

Radiographic Findings

- Radiography
  - Loss of vertebral body height
  - Lytic lesions with surrounding sclerosis or purely sclerotic

MR Findings

- T1WI
  - Low signal focal lesions with loss of vertebral body height, normal intervertebral disc
- T2WI
  - Focal increased signal lesions with spared intervertebral disc
- T1WI C+
  - Lesions enhance without significant associated paraspinal or epidural soft tissue

DIFFERENTIAL DIAGNOSIS

Septic Multifocal Osteomyelitis

- Bone destruction with soft tissue component

Langerhans Cell Histiocytosis

- Single level spine involvement (typical)

Ewing Sarcoma

- Aggressive bone destruction

Juvenile Idiopathic Arthritis

- Fusion as late complication

PATHOLOGY

General Features

- Etiology
  - CRMO part of heterogeneous group of disorders of innate immune system (autoinflammatory disease)
  - Archetypal disease of this class is familial Mediterranean fever
  - Mutations in either pyrin or tumor necrosis factor receptor superfamily
    - Homozygous mutation in *LPIN2* gene in Majeed syndrome

Staging, Grading, & Classification

- Major criteria: Imaging-proven osteolytic/sclerotic bone lesion, multiple bone lesions, pustulosis palmoplantaris or psoriasis, sterile bone biopsy
- Minor criteria: Normal blood cell count, ESR mild elevation, course > 6 months, hyperostosis, other autoimmune disease

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, scoliosis, kyphosis, rarely cord compression

Demographics

- Age
  - Infant to 55 years old
  - Median age of 10 years old

Natural History & Prognosis

- Waxing and waning symptoms over multiple years
- Self-limited course with limited long-term skeletal abnormalities

Treatment

- NSAID, steroids
- Infliximab and bisphosphonates have also shown clinical improvement

DIAGNOSTIC CHECKLIST

Consider

- CRMO may cause vertebra plana, but restitution of vertebral height with treatment has not been described (unlike Langerhans cell histiocytosis)

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Grisel Syndrome

KEY FACTS

TERMINOLOGY

- Atlantoaxial rotary subluxation accompanying respiratory infection or otolaryngolic surgery

IMAGING

- Fixed, rotated head occurring in child soon after upper respiratory infection or otolaryngologic surgery
- Difficult to obtain adequate films 2° to head position
- Open-mouth odontoid: Asymmetric C1 lateral masses
  - Anteriorly rotated side wider and closer to odontoid
  - Opposite side smaller and farther from odontoid
- Lateral view: ± widened atlantodental interval
- Dynamic CT: Should include occiput C1-C2
  - Initial scan: Maintain position of comfort
  - 2nd scan: Rotate head to contralateral side
  - Interpretation: C1-C2 relationship fixed or C1 unable to rotate past neutral to contralateral side
- MR may replace CT to eliminate radiation dose

TOP DIFFERENTIAL DIAGNOSES

- Torticollis
  - Resolves within a few days to weeks
- Septic C1-C2 joint

PATHOLOGY

- Multiple theories: Common theme tracking of inflammatory mediators from upper airway

CLINICAL ISSUES

- Pain, head tilt, restricted neck movement
- Cock-robin position: Head rotated and tilted to 1 side with chin lifted to opposite side
- Associated with otitis media, pharyngitis, retropharyngeal abscess, upper respiratory infection, otolaryngologic surgery
- 68% < 12 years old; 90% < 21 years old
- Leads to permanently fixed deformity if left untreated
- Reduction and immobilization are key

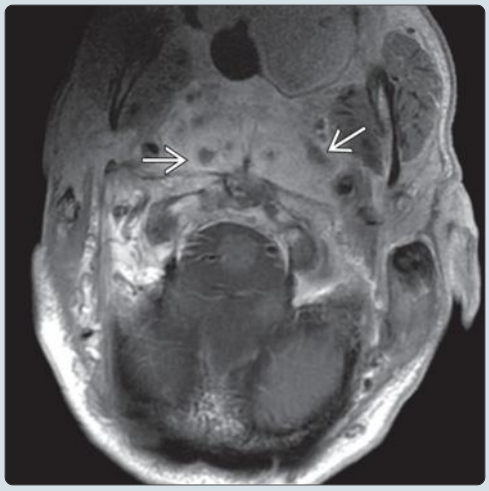
(Left) *Lateral radiograph shows typical retropharyngeal effusion due to strep pharyngitis. Grisel syndrome may occur with any stage of infection, from effusion to extensive abscess.* **(Right)** *Axial CECT shows retropharyngeal space abscess due to left palatine tonsillitis. Although the C1-C2 articulation is not affected, one can appreciate how easily inflammatory changes can track to that articulation.*





(Left) *Sagittal STIR MR in a florid case of craniovertebral junction bacterial infection shows marked increased signal involving prevertebral space, odontoid, and loss of anterior C1 arch margin. This is to be contrasted with the less aggressive appearance of Grisel syndrome.* **(Right)** *Axial T1 C+ MR shows infection involving the craniovertebral junction with multiple small abscesses within the posterior nasopharyngeal tissue. Note the close anatomic relationship which would allow for C1-C2 joint involvement.*





TERMINOLOGY

Synonyms

- Nontraumatic atlantoaxial subluxation

Definitions

- Atlantoaxial rotary subluxation accompanying respiratory infection or otolaryngolic surgery

IMAGING

General Features

- Best diagnostic clue
  - Child with fixed, rotated head soon after upper respiratory infection or otolaryngologic surgery

Radiographic Findings

- Radiography
  - Difficult to obtain adequate films due to head position
  - Open-mouth odontoid: Asymmetric C1 lateral masses
    - Anteriorly rotated side wider and closer to odontoid
    - Opposite side smaller and farther from odontoid

CT Findings

- NECT
  - Dynamic CT: Should include occiput C1-C2
    - Initial scan: Maintain position of comfort
    - 2nd scan: Rotate head to contralateral side
    - Interpretation: C1-C2 relationship fixed or C1 unable to rotate past neutral to contralateral side

MR Findings

- MR may replace CT to eliminate radiation dose

Imaging Recommendations

- Protocol advice
  - Dynamic CT or MR
    - Do contralateral rotation 2nd to minimize overall discomfort
    - Consider doing only rotation to contralateral side to eliminate radiation dose

DIFFERENTIAL DIAGNOSIS

Torticollis

- Resolves within a few days to weeks
- Normal motion at C1-C2

Septic C1-C2 Joint

- Extensive edema surrounding odontoid process
- C1-C2 joint effusion
- Bone erosion/destruction

PATHOLOGY

General Features

- Etiology
  - Multiple theories: Common theme tracking of inflammatory mediators from upper airway
    - Nasopharyngeal veins drain into periodontoid plexus without intervening lymph nodes
    - Laxity of transverse ligament or trapped hypertrophied synovium
    - Relative horizontal orientation of C1-C2 joints and generalized ligamentous laxity in children are contributing factors

Staging, Grading, & Classification

- Type 1: Rotation of axis around odontoid without anterior displacement (within normal range of motion)
- Type 2: Rotation of axis relative to 1 lateral facet with 3-5 mm anterior displacement
- Type 3: Rotation of axis with > 5 mm anterior displacement
- Type 4: Rotation of axis with posterior dislocation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, head tilt, restricted neck movement
    - Acute onset
    - Cock-robin position: Head rotated and tilted to 1 side with chin lifted to opposite side
  - Sudek sign: Palpable spinous process of C2 on same side of midline as chin
- Other signs/symptoms
  - Associated with otitis media, pharyngitis, retropharyngeal abscess, upper respiratory infection, otolaryngologic surgery
  - Neurologic complications in 15% of cases

Demographics

- Age
  - 68% < 12 years old; 90% < 21 years old
    - In an adult, think septic C1-C2 joint, not Grisel syndrome

Natural History & Prognosis

- Leads to permanently fixed deformity if left untreated
- Long-term untreated condition leads to facial flattening on ipsilateral side

Treatment

- Reduction and immobilization are key
  - Early presentation is treated with soft collar
  - Later presentations require more aggressive intervention, including head halter or halo traction

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Need to confirm limited/fixed motion to diagnose

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Paraneoplastic Myelopathy

KEY FACTS

TERMINOLOGY

- Paraneoplastic neurologic disorder (PND)
- Antineuronal immune mechanism targeting CNS, occurring in conjunction with cancer

IMAGING

- Longitudinally extensive, symmetric, tract-specific signal changes within spinal cord that often enhance after gadolinium administration in patient with cancer
- Signal abnormality in ~ 70% of cases
- Lateral, dorsal columns
- Central gray matter

TOP DIFFERENTIAL DIAGNOSES

- Cord infarction
- Neuromyelitis optica
- B12 deficiency (subacute combined degeneration)
- Metastatic disease
- Neurosarcoidosis

PATHOLOGY

- Paraneoplastic disease usually
  - Antibodies against intracellular antigens
  - Older individuals, resistant to treatment
  - Mediated by T-cell mechanisms
- Most common with **lung** and **breast** cancer
- Most common antibodies causing myelopathy
  - Amphiphysin-IgG
  - Collapsin response-mediator protein-5 (CRMP-5)

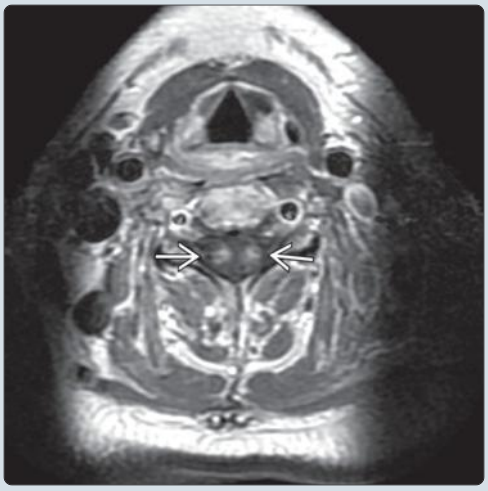
CLINICAL ISSUES

- Insidiously progressive myelopathy
- Often manifesting before detection of underlying cancer (2/3 of patients)
  - Median of 12 months from myelopathy onset to cancer detection
- Poor prognosis, often becoming wheelchair dependent





(Left) *Sagittal T1 MR shows multiple metastatic lesions ➡ involving the cervical and upper thoracic vertebral bodies, with marrow conversion from prior radiation therapy ➡. The cord is mildly expanded.* **(Right)** *Sagittal T2 MR shows longitudinally extensive high signal within the cervical cord, with mild cord expansion ➡. This edema pattern is nonspecific, and cord metastases and radiation change should also be considered.*





(Left) *Axial T1 C+ MR shows the typical pattern of paraneoplastic myelopathy with focal central tract enhancement ➡ in a symmetrical fashion. This symmetrical tract involvement would not be seen with metastases or radiation change.* **(Right)** *Sagittal T1 C+ MR in a patient with a focal cord metastasis ➡ shows the very localized nature of the tumor to be contrasted, with the longitudinally extensive abnormality with paraneoplastic disease of the cord.*

TERMINOLOGY

Abbreviations

- Paraneoplastic neurologic disorder (PND)

Definitions

- Antineuronal immune mechanism targeting CNS, occurring in conjunction with cancer

IMAGING

General Features

- Best diagnostic clue
  - Longitudinally extensive, symmetric, tract-specific signal changes within spinal cord that often enhance after gadolinium administration in patient with cancer
- Location
  - Cervical or thoracic cord
- Size
  - > 3 vertebral segments in length
- Morphology
  - Linear signal abnormality following spinal cord tract

MR Findings

- T1WI
  - Minimal signal change, may be normal
- T2WI
  - Signal abnormality in ~ 70% of cases
  - Longitudinally extensive
  - Look for symmetrical bilateral tract involvement
    - Lateral, dorsal columns
    - Central gray matter
- T1WI C+
  - Enhancement variable, but may be linear and involve long segments of cord tracts

Nuclear Medicine Findings

- PET
  - No spinal cord hypermetabolic uptake

DIFFERENTIAL DIAGNOSIS

Cord Infarction

- Acute onset
- May show "owl eye" or "snake eye" central pattern of gray matter involvement

Neuromyelitis Optica

- Longitudinally extensive T2 signal abnormality in cord
- Predilection for gray matter
- Usually patchy contrast enhancement

B12 Deficiency (Subacute Combined Degeneration)

- Dorsal column signal abnormality, without enhancement
- Laboratory findings make diagnosis

Metastatic Disease

- Focal enhancing mass within cord, with edema
- May show rim or flame signs

Neurosarcoidosis

- Leptomeningeal enhancement typical
- May show direct cord involvement, but usually nodular

PATHOLOGY

General Features

- Etiology
  - Antineuronal immune mechanism occurring in conjunction with cancer
  - Paraneoplastic disease usually
    - Antibodies against intracellular antigens
    - Affects older individuals
    - Mediated by T-cell mechanisms
    - Largely resistant to treatment
  - Contrast with autoimmune encephalitis
    - Antibodies to synaptic or cell surface antigens
    - Children and younger adults
    - May or may not be associated with cancer
    - Responsive to immunomodulatory therapy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Insidiously progressive myelopathy
    - Often manifesting before detection of underlying cancer
  - May develop quickly and be severe
- Other signs/symptoms
  - Multifocal neurologic symptoms
    - Encephalopathy
    - Cranial neuropathies
    - Cerebellar ataxia

Natural History & Prognosis

- Poor prognosis
- Minority of patients improve with current treatment

Treatment

- Immunotherapy or treatment of underlying cancer may give mild improvement
- Steroids, intravenous immunoglobulin, cyclophosphamide have been tried

DIAGNOSTIC CHECKLIST

Consider

- 1/3 of patients have normal spinal MR study

Image Interpretation Pearls

- Longitudinally extensive bilateral tract involvement is characteristic

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PART V
SECTION 1

Neoplasms



Introduction and Overview

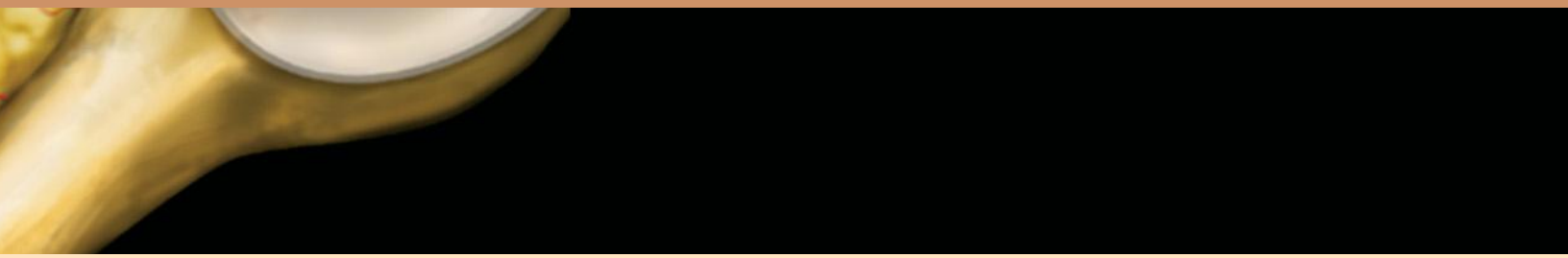
Spread of Neoplasms 690

Extradural

Blastic Osseous Metastases 694
Lytic Osseous Metastases 698
Hemangioma 702
Osteoid Osteoma 706
Osteoblastoma 712
Aneurysmal Bone Cyst 716
Giant Cell Tumor 720
Osteochondroma 724
Chondrosarcoma 728
Osteosarcoma 732
Chordoma 736
Ewing Sarcoma 742
Lymphoma 746
Leukemia 752
Plasmacytoma 756
Multiple Myeloma 760
Neuroblastic Tumor 764
Langerhans Cell Histiocytosis 768
Angiolipoma 772

Intradural Extramedullary

Schwannoma 776
Meningioma 780
Solitary Fibrous Tumor/Hemangiopericytoma 786
Neurofibroma 790
Malignant Nerve Sheath Tumors 794
Metastases, CSF Disseminated 798
Paraganglioma 802



Intramedullary

Astrocytoma	**806**
Cellular Ependymoma	**810**
Myxopapillary Ependymoma	**814**
Hemangioblastoma	**818**
Spinal Cord Metastases	**824**
Primary Melanocytic Neoplasms/Melanocytoma	**828**
Ganglioglioma	**830**

Anatomy-Based Imaging Issues

Neoplasms affecting the spine may spread by direct extension via the lymphatic system, the hematogenous route, or along the cerebrospinal fluid pathways. There can also be combinations of these pathways involved with the most typical being hematogenous metastatic dissemination to vertebral bodies with subsequent direct extension into the epidural space.

Primary tumors located in soft tissues may extend into the vertebral column by direct extension. An example would include lung carcinoma extending into the chest wall and subsequently the paravertebral region and into the spinal column and epidural space. Prostate, bladder, or bowel carcinoma may extend into the presacral space and subsequently into the vertebral column and epidural space. Nasopharyngeal carcinoma extends into the clivus and skull base and may track along the cranial nerves. In rare cases, there may be direct extension of a CNS tumor along the biopsy or surgical tract. There are also rare cases of CNS tumors extending through access via indwelling shunt tubing to give systemic metastases. Findings of direct extension of neoplasm in the spine consists of a soft tissue mass with adjacent bone destruction and variable neural compromise. Direct extension of neoplasm into the epidural space is more likely from vertebral body through the posterior longitudinal ligament. The anterior longitudinal ligament and disc are relatively resistant to tumor invasion. The anterior longitudinal ligament is stronger than the posterior longitudinal ligament and has fewer perforating vessels. Once the tumor has access into the epidural space, it comes against the tough dura, which is an effective barrier to tumor penetration. These barriers result in the distinguishing features between a disc space infection with adjacent vertebral osteomyelitis (with the epicenter at the level of the disc space) and neoplastic involvement (with the epicenter involving the vertebral bodies with sparing of the disc space). Direct extension of tumor may also be seen with a primary cord tumor within the cervical spine, with extension into the infratentorial space. Rarely, brainstem or cerebellar neoplasms may extend into the upper cervical cord.

Lymphatic spread is of limited importance in spine imaging relative to the more ubiquitous hematogenous spread. Local spread of pelvic tumors within the lumbar spine without pulmonary metastases would suggest venous or lymphatic route of extension.

Hematogenous spread is the major pathway of extension of malignant tumors to the axial skeleton. Batson plexus is a longitudinal network of valveless veins running parallel to the spinal column. These veins lie outside of the thoracoabdominal cavity and communicate with multiple aspects of the venous system, including vena cava, spinal, portal, azygos, intercostal, pulmonary, and renal veins. Flow direction in Batson plexus is variable due to the variable intrathoracic and intraabdominal pressures. Tumors in multiple anatomic sites could cause metastatic lesions along the course of the venous plexus without lung or liver involvement. Prostate carcinoma cells could seed vertebral bodies via Batson plexus and not necessarily extend into the vena cava. Breast carcinoma might also seed the vertebral bodies via the azygos system into Batson plexus. There is only 5-10% of portal blood flow that might shunt to Batson plexus, explaining the relatively low frequency of spinal metastatic lesions with GI and GU primaries. For the vast majority of spinal metastases, there may be no clear answer to the precise route to the end target. Homing properties of the tumor cells and receptive properties of the implantation site may be more important than any particular vascular route.

Spread along the CSF pathways is an important route for primary intracranial tumors. Tumor emboli gain access to the CSF via fragmentation, as well as being shed during surgical manipulation. CNS tumor types showing subarachnoid spread include medulloblastoma, ependymoma, pineal tumors, astrocytoma, lymphoma and leukemia, choroid plexus carcinoma, and retinoblastoma (poor prognosis with *MYCN* gene amplification). Spread along the CSF pathways can also occur following initial hematogenous dissemination of tumor. For example, this pattern of spread would be present in cord and leptomeningeal metastatic disease following the initial hematogenous dissemination in lung and breast metastatic disease.

Pathologic Issues

Forty percent of patients with cancer will develop visceral or bony metastases during their illness. The spinal column is the most common site of osseous metastases. Men are more frequently affected with vertebral metastases, with a male to female ratio of 3:2. Prostate, lung, and breast carcinomas account for the vast majority of spinal metastases. Locations are primarily thoracic (70%) > lumbar (20%) > cervical.

Primary tumors are generally composed of a variety of biologically different cells in regard to ultimate metastatic potential. Cells continually shed from the primary tumor and gain access to the circulatory system. Fewer than 0.01-0.1% of tumor cells survive to reach a distant site. Successful tumor spread requires completion of a complex pathway, including tumor separation from the primary source, access to blood, CSF or lymphatic system, survival within the transport process, attachment to the endothelium of a distant vessel as well as exiting that vessel into the interstitial space, and finally developing the vascular supply at a distant site. The distant host environment site is a complex milieu. Multiple anatomic pathways may be involved with varying flow patterns within the veins and arteries.

Mechanisms of Tumorigenesis

Tumorigenesis in humans appears to be multistep process. These steps reflect genetic alterations that drive progressive transformation of normal human cells into highly malignant versions. Cancers in humans have been shown to have an age-dependent incidence which implicates 4-7 rate limiting, stochastic events. Tumor development proceeds via a succession of genetic changes, each conferring one or another type of growth advantage, which leads to the progressive conversion of normal human cells into cancer cells. By definition, cancer cells will have defects in regulatory circuits that govern cell proliferation and homeostasis. These defects have been categorized into 6 types of alterations in cell physiology: 1) Self-sufficiency in growth signals, 2) insensitivity to growth inhibitory signals, 3) evasion of programmed cell death (apoptosis), 4) limitless replicative potential, 5) sustained angiogenesis, and 6) tissue invasion and metastasis. Each of these 6 physiologic changes reflects the successful breaching of an anticancer defense mechanism that is hardwired into cells and tissues.

Normal cells require growth signals before they can move into an active proliferative state. The signals are transmitted to the cell via transmembrane receptors that bind a variety of

signaling molecules, such as diffusible growth factors, extracellular matrix components, and cell-to-cell adhesion molecules (CAMs). Many cancer cells acquire the ability to synthesize growth factors to which they are responsive, which creates an abnormal positive feedback loop. Examples would be the production of platelet-derived growth factor (PDGF) by glioblastoma and tumor growth factor alpha (TGF) by sarcomas. Epidermal growth factor receptor (EGF-R) is upregulated in brain tumors.

Normal tissues have a variety of mechanisms which limit proliferation. The signals include both growth inhibitors and inhibitors within the extracellular matrix on the surface of adjacent cells. Many antiproliferative signals are channeled through the retinoblastoma tumor suppressor protein (pRb) and its variations. Disruption of the retinoblastoma protein pathway will allow cell proliferation by rendering the cells insensitive to antigrowth factors by repressing the E2F transcription factors.

Evasion of programmed cell death (apoptosis) commonly occurs through mutations involving the *p53* tumor suppressor gene. The inactivation of this *p53* protein is seen in over 50% of human cancers.

Limitless replicative potential of cancer tumors is seen in cultures of tumor cells that have become immortalized. Normal human cell types in culture have a capacity for no more than 60-70 doublings. The ends of the chromosomes appear to be involved in this process, which are called the telomeres. The erosion of the telomeres occurs through successive cycles of normal cell replication causing them to lose their ability to protect the ends of the chromosome DNA. This leads to eventual cell crisis and cell death. In contrast, telomere maintenance is evident in all types of malignant cells. In cancer, telomeres are maintained at a length above some critical threshold, which allows unlimited cell multiplication.

Abnormal angiogenesis is perhaps one of the more recognized aspects of abnormal cell physiology in cancer. Angiogenesis promoting signals are classically seen with vascular endothelial growth factor (VEGF) and fibroblast growth factors (FGF). More than 2 dozen angiogenic-inducing factors are known, with a similar number of inhibiting-type proteins.

Metastatic Disease and Cell Motion

Tumor metastases are the cause of 90% of human cancer deaths. Tumor invasion and tumor metastases are very complex processes, with multiple genetic and biochemical factors. These can be described as tumor invasion, tumor cell dissemination through the blood stream or lymphatic system, colonization of distant organs, and outgrowth of the metastasis. Several classes of proteins are involved in tethering cells to their surroundings and are altered in invasion and metastases. CAMs and integrins are involved in this cell-to-cell regulatory signaling. The basement membrane is the first barrier tumor cells must breech. Receptors on the surface of cells recognize the glycoprotein of the basement membrane to which they attach. Attachment is followed by proteolysis of type IV collagen of the basement membrane by tumor-specific collagenase. Locomotion follows basement membrane lysis, with cells crossing that defective basement membrane with access to the interstitial space, lymphatics, and blood vessels. Chemoattractants can activate cell migration and invasion through activation of specific receptors and downstream intracellular signaling pathways. This appears to ultimately reorganize the actin cytoskeleton. Rho/Rac GTPases are well-known regulators of cytoskeletal organization. Rac activation induces membrane ruffles, adhesion complexes, and lamellipodia formation.

Tumor dissemination and cell motility occurs along three different pathways including epithelial-to-mesenchymal transition (EMT), amoeboid transcription, and collective migration. In EMT, the cell elongates and degrades the local matrix by enzymes and migrates with pseudopodia-like projections. In amoeboid transcription the cells become spherical and pass through gaps in the extracellular matrix. In collective migration, sheets or clusters of tumor cells migrate. EMT includes the downregulation of epithelial markers in tight junctions and cytokeratin filament network and upregulation of mesenchymal markers, like N-cadherin, vimentin, integrins, tenascin C, and fibronectin, fibroblast- specific protein 1. N-cadherin seems to be the most important of this group of markers. EMT can be controlled by intrinsic oncogenic activation like K-Ras mutation or Her2 overexpression. In some cancers, such as lung, the central role of EMT regulator belongs to TGFβ. TGFβ ultimately promotes EMT by regulating genes that control cell proliferation, apoptosis, differentiation, motility and migration.

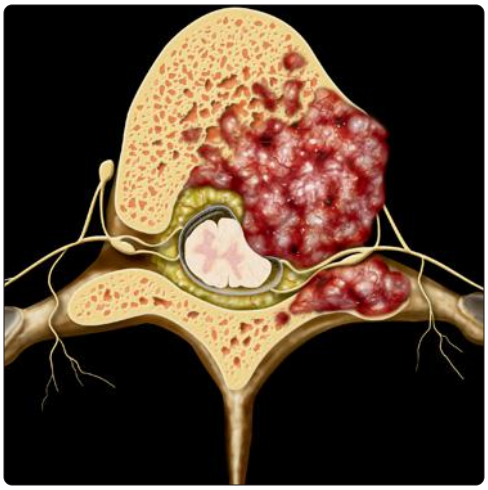
Clinical Implications

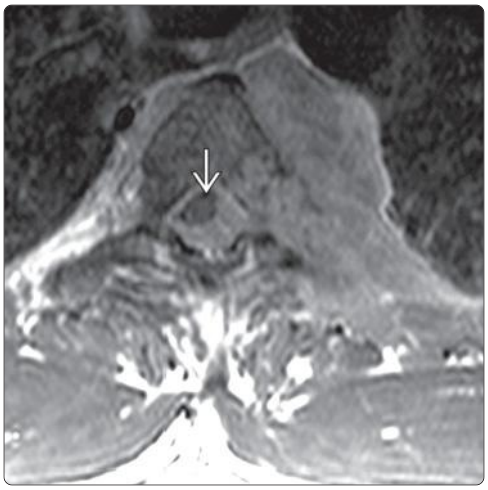
Spine metastatic involvement presents with unrelenting back pain. Objective signs are uncommon, or occur late in the disease (such as palpable mass and deformity). Back pain and weakness are a sign of epidural tumor extension. Due to spinothalamic tract crossing pattern, sensory levels may be 1-2 segments below the site of compression. Sensory abnormalities are an uncommon presenting sign of metastatic disease in the spine.

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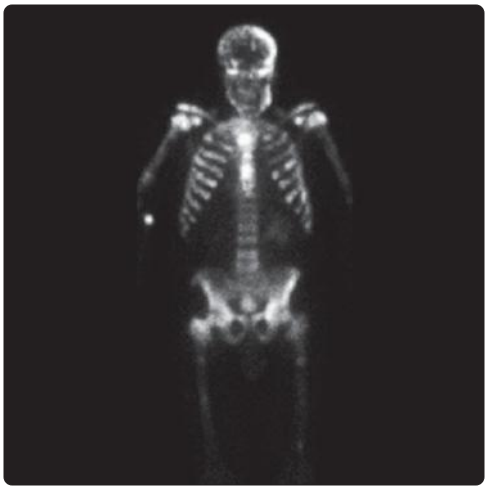
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(Left) *Axial graphic shows hematogenously disseminated lytic metastatic lesion to thoracic vertebral body and pedicle, with subsequent direct epidural tumor extension and cord compression.* **(Right)** *Axial T1WI C+ MR shows large hematogenously disseminated metastatic paravertebral mass lesion involving thoracic body, with direct tumor extension to posterior elements, chest wall, and epidural space with cord compression ⇒.*





(Left) *Bone scan in a case of extensive blastic prostate carcinoma shows multiple foci of increased radiotracer uptake consistent with diffuse bone metastasis.* **(Right)** *Sagittal T1WI C+ MR in a patient with extensive blastic prostate carcinoma shows diffuse abnormal decreased signal from all vertebral bodies and posterior elements with multiple foci of ventral epidural tumor extension ⇒, causing mild cord compression.*

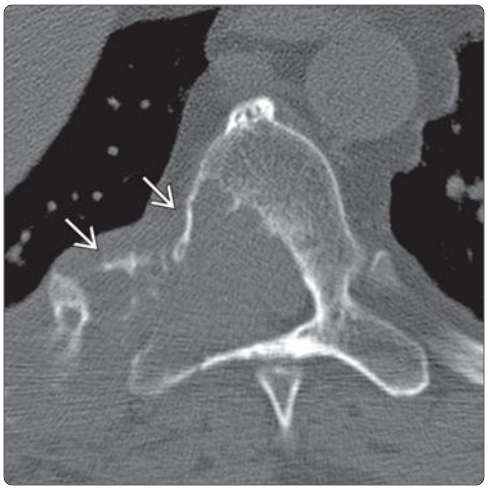


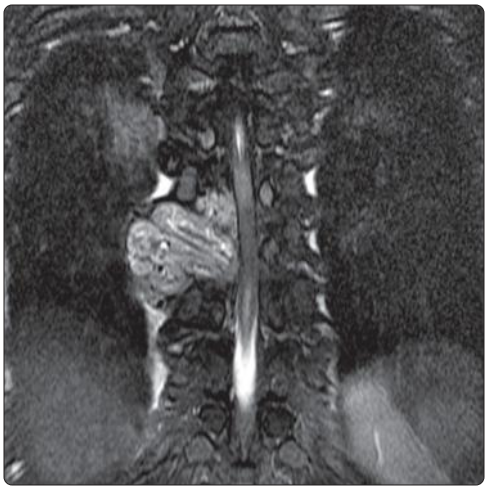


(Left) *Sagittal T2WI MR in a patient with multiple myeloma shows 2 levels of bony tumor involvement and slight spinous process expansion. There is severe cord compression at both levels.* **(Right)** *Sagittal T1 C+ FS MR of multiple myeloma shows 2 levels of focal tumor involvement with enhancement and slight spinous process expansion. Cord compression is present at both levels.*









(Left) *Axial NECT in a patient with renal cell metastasis shows mass destroying the right side of T7-T8 bodies, extending into posterior elements and crossing to involve right ribs and costovertebral joint. There is bony expansion with a thin rim "soap bubble" pattern ⇨.* **(Right)** *Coronal T2WI FS MR in a patient with renal cell metastasis shows mass involving the ribs and right side of T7-T8 bodies, with epidural extension and cord compression. Multiple flow voids are present within the mass.*





(Left) *Unenhanced T2WI MR in a patient with a large cervical cord metastatic lesion from lung carcinoma shows diffuse cord edema from cervicomedullary junction inferiorly to C5. More focal signal abnormality is present at the C2-C3 level.* **(Right)** *Sagittal T1WI C+ MR in a patient with large cervical cord metastatic lesion from lung carcinoma via hematogenous dissemination shows enhancement of large intramedullary metastatic lesion at the C2-C3 level.*





(Left) *Sagittal T1 C+ MR in a patient with extensive nodular leptomeningeal metastatic disease from high-grade brain oligodendroglioma shows massive nodular-enhancing leptomeningeal tumor following contrast administration ⇨. This child had rapid onset of flaccid paraplegia suggesting cord infarction from tumor compression.* **(Right)** *Sagittal T1 C+ MR shows focal epidural mass with cord compression from metastatic melanoma. Route of dissemination is presumed hematogenous.*

Blastic Osseous Metastases

KEY FACTS

TERMINOLOGY

- Extension of primary tumor to spine where bone production exceeds bone destruction

IMAGING

- Multiple osteoblastic lesions in spine
  - May coexist with areas of osteolytic tumor, soft tissue mass
- Lesion distribution proportional to red marrow (lumbar > thoracic > cervical)
- MR signal typically diminished on T1, T2WI in areas of osteoblastic metastases
- Sclerotic metastases usually tracer avid on bone scan

TOP DIFFERENTIAL DIAGNOSES

- Treated metastases
- Discogenic sclerosis
- Hemangioma
- Paget disease
- Osteosarcoma

PATHOLOGY

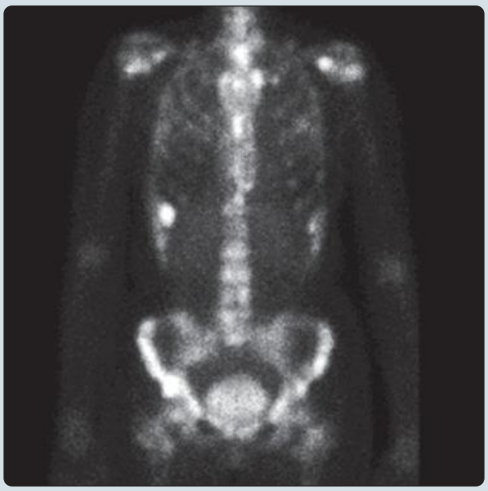
- Marrow infiltration, tumor stimulates osteoblastic response
  - New bone deposition on trabeculae, within intertrabecular spaces
- Primary tumor, adults: Prostate, breast, carcinoid, lung, GI, bladder, nasopharynx, pancreas
- Primary tumor, children: Medulloblastoma, neuroblastoma, Ewing sarcoma

CLINICAL ISSUES

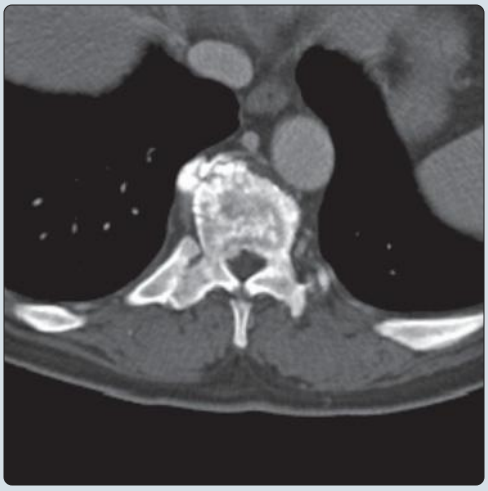
- Pain: Progressive axial, referred, or radicular
- Epidural tumor, if present, may cause neurologic dysfunction
- 90% of prostate metastases involve spine, with lumbar 3x more often than cervical

(Left) *Anteroposterior plain film shows diffuse extensive bone sclerosis of the spine and pelvis due to diffuse osseous metastases from breast carcinoma.* **(Right)** *Anteroposterior bone scan shows diffuse, patchy tracer uptake in the thoracolumbar spine, pelvis, sternum, and multiple ribs due to diffuse osseous metastases from breast carcinoma.*





(Left) *Axial NECT through the lower thoracic spine shows patchy sclerosis of the vertebral body due to blastic metastases from prostate carcinoma.* **(Right)** *Sagittal T1-weighted MR image shows low signal involving the L3 body and sacrum, reflecting diffuse metastatic disease. Extraosseous extension into the epidural space is seen at L3. There is no specific signal change on the MR to define the blastic nature of these metastatic lesions.*





TERMINOLOGY

Synonyms

- Sclerotic metastases, osteosclerotic metastases, osteoblastic metastases

Definitions

- Spread of primary tumor to spine where bone production exceeds bone destruction

IMAGING

General Features

- Best diagnostic clue
  - Multiple blastic lesions in spine
- Location
  - Vertebral body and posterior elements
  - Lesion distribution proportional to red marrow (lumbar > thoracic > cervical)
- Size
  - Any size from a few millimeters to entire vertebral body (ivory vertebra)
- Morphology
  - Round focus of sclerosis or mixed lytic/sclerotic

Radiographic Findings

- Radiography
  - Discrete or mottled areas of sclerosis, typically multiple

CT Findings

- NECT
  - Multiple sclerotic lesions, possibly coexisting with areas of osteolysis
  - May see paravertebral and epidural soft tissue mass
- CECT
  - Enhancement typically not detectable due to sclerosis
  - May see enhancement in adjacent areas of osteolysis or in extraosseous extension of tumor
- CT myelography
  - Multiple sclerotic lesions as per NECT
  - Extradural compression by epidural tumor
    - Better assessment of cord compression than with NECT or CECT
    - Reserve for when patient is unable to undergo MR

MR Findings

- T1WI
  - Low or no signal in areas of osteoblastic metastases
  - ± compression fracture
  - ± paravertebral and epidural soft tissue
  - Intervertebral discs generally spared
- T2WI
  - Variable, may be hyperintense or hypointense
- STIR
  - Variable, may be hyperintense or hypointense
- DWI
  - Controversial efficacy of DWI for spinal metastases
  - Several reports of false-negatives for sclerotic metastases with DWI
- T1WI C+
  - Variable enhancement depending upon degree of sclerosis

Nuclear Medicine Findings

- Bone scan
  - Sclerotic metastases typically show increased uptake
  - Diffusely increased uptake ("Superscan") most common with prostate carcinoma
  - Diffusely increased uptake excluding calvarium ("headless bone scan") uncommon pattern of metastatic prostate carcinoma
- PET
  - Reports of FDG negative blastic metastases despite radiographic lesion progression
    - Treated osteolytic metastases also FDG negative, yet tracer avid on bone scan

Imaging Recommendations

- Protocol advice
  - MR scan the entire spine to evaluate for osseous metastasis, epidural tumor, cord compression
    - Include STIR or fat-suppressed T2WI and contrast-enhanced (± fat-saturated) T1WI
  - Bone scan useful for screening of entire skeleton

DIFFERENTIAL DIAGNOSIS

Treated Metastases

- Osteolytic metastases "heal" with treatment, may become sclerotic

Discogenic Sclerosis

- Sclerosis limited to endplate marrow, adjacent to degenerated intervertebral disc

Hematopoietic Malignancy

- Plasmacytoma
  - Typically osteolytic; sclerosis uncommon (3%)
- Multiple myeloma (MM)
  - Typically osteolytic; sclerotic multiple myeloma uncommon (1%)
  - POEMS: Variant with sclerotic lesions
- Lymphoma
  - "Ivory" vertebra a rare presentation of osseous lymphoma

Hemangioma

- "Corduroy" appearance of thickened, vertical trabeculae within geographic area of lucency

Osteosarcoma

- Extraosseous extension, periosteal new bone, soft tissue mass

Bone Cement (Vertebroplasty/Kyphoplasty)

- Very dense material in midline or paramidline location of vertebral body with morphologic changes of compression fracture

Paget Disease

- Expansion of body with thickened trabeculae ("picture frame" vertebrae)

Benign (Osteoporotic) Compression Fracture

- May see linear (horizontal) sclerosis due to trabecular impaction or due to healing
- Loss of vertebral body height

Myelofibrosis

- Often normal radiographs; may see diffuse vertebral sclerosis, peripheral skeleton spared
- Diffusely diminished T1, T2 marrow signal

Osteopetrosis

- Thick sclerosis in endplate marrow ("rugger jersey spine")
- No extraosseous extension

Renal Osteodystrophy

- "Rugger jersey spine" appearance on radiographs
- Vertebral body low T1 signal, no paravertebral or epidural soft tissue component

PATHOLOGY

General Features

- Etiology
  - Hematogenous dissemination (arterial or venous via Batson plexus) > perineural, lymphatic, CSF spread
  - Marrow infiltration, tumor stimulates osteoblastic response
  - Primary tumor, adults: Prostate, breast, carcinoid, lung, GI, bladder, nasopharynx, ovaries, pancreas
  - Primary tumor, children: Medulloblastoma, neuroblastoma, Ewing sarcoma
  - Tumors promote pro-osteoblastic factors, such as RANKL, parathyroid hormone-related protein (PTHrP), BMPs, TGF-β, IGFs, FGFs, interleukins, and VEGF
    - Leads to unregulated osteoblastic function through activation of proliferation and differentiation signaling pathways like Wnt

Staging, Grading, & Classification

- Surgical stratification
  - Grade of malignancy (slow growth, 1 point → rapid growth, 4 points)
  - Visceral metastases (no metastases, 0 points → untreatable, 4 points)
  - Bone metastases (solitary, 1 point → multiple, 2 points)
  - Total 2-3 points suggest wide excision for long-term control
  - Total 8-10 points indicate nonoperative supportive care

Gross Pathologic & Surgical Features

- Deposition of new bone on existing trabecula and in intertrabecular spaces
- May coexist with osseous destruction ± adjacent soft tissue mass

Microscopic Features

- Varies with histology of primary, osteoclastic/blastic response

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Progressive axial, referred, or radicular pain
  - Epidural tumor extension may cause neurologic dysfunction
  - Compression fracture
- Other signs/symptoms
  - Hypocalcemia or hypercalcemia
- Clinical profile
  - Elderly male with prostate carcinoma, new onset leg weakness, back pain

Demographics

- Age
  - Children and adults
- Gender
  - Predilection varies with specific tumor
- Epidemiology
  - Spine most frequent site of skeletal metastases
  - 90% of prostate metastases involve spine, with lumbar 3x more often than cervical
  - Epidural spinal cord compression in 5% of adults with systemic cancers (70% solitary, 30% multiple sites)

Natural History & Prognosis

- Relentless, progressive; pathologic fracture, spinal cord compression
- Prognosis varies with histology of primary lesion

Treatment

- Radiation therapy
- Surgical decompression, stabilization
- Vertebroplasty, embolization

DIAGNOSTIC CHECKLIST

Consider

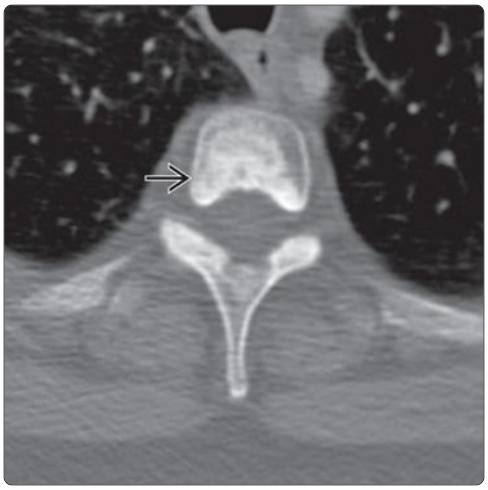
- Defining response to therapy of sclerotic lesion difficult since osteolytic conversion (tumor progression) and fading (good response) may look identical

Image Interpretation Pearls

- Metastatic "ivory vertebral body" seen with prostate, lymphoma, myeloma, chordoma

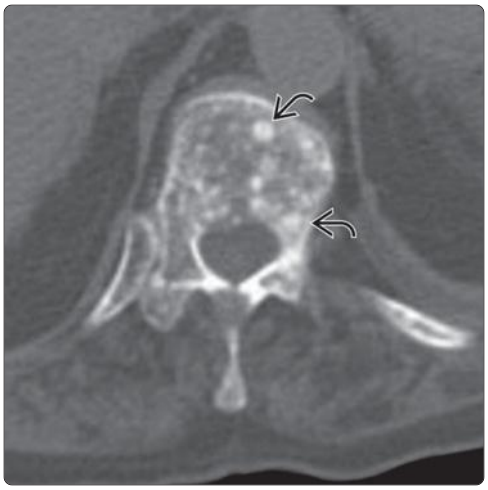
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(Left) *Axial NECT shows diffusely sclerotic metastasis from breast carcinoma involving most of the vertebral body.* **(Right)** *Sagittal T2WI MR in the same patient shows sclerotic metastases from breast carcinoma involving 2 upper thoracic vertebral bodies. There is loss of signal, similar to that of cortical bone, due to the dense mineralization.*





(Left) *Axial NECT shows multiple small, rounded sclerotic lesions in the lower thoracic vertebra due to blastic metastases from breast carcinoma.* **(Right)** *Sagittal T1WI MR in the same patient shows the sclerotic metastases as rounded foci of markedly low signal involving multiple thoracic and lumbar vertebral bodies.*





(Left) *Axial NECT shows diffuse sclerosis involving the left half of the vertebral body due to blastic metastasis in this patient with metastatic prostate carcinoma.* **(Right)** *Sagittal T1WI MR shows diffuse marrow replacement throughout the visualized spine and multiple compression fractures. Diffuse sclerotic and lytic metastases from Ewing sarcoma were present on radiography. Low signal in the T6 and T10 vertebral bodies corresponded to 2 of the more sclerotic lesions.*

Lytic Osseous Metastases

KEY FACTS

TERMINOLOGY

- Spread of primary tumor to spine, where bone destruction exceeds bone production

IMAGING

- Multiple osteolytic lesions in spine
- Compression fracture with bowing of posterior cortex, osteolysis extending into neural arch, extraosseous soft tissue
- Lesion distribution proportional to red marrow (lumbar > thoracic > cervical)
- Radiography requires 50-70% bone destruction and tumor size > 1 cm for detection
- Bone scan can give false-negatives with aggressively lytic tumor or with very small lesions

TOP DIFFERENTIAL DIAGNOSES

- Hematopoietic malignancy
- Benign (osteoporotic) compression fracture
- Schmorl node
- Normal heterogeneous marrow
- Spondylodiscitis

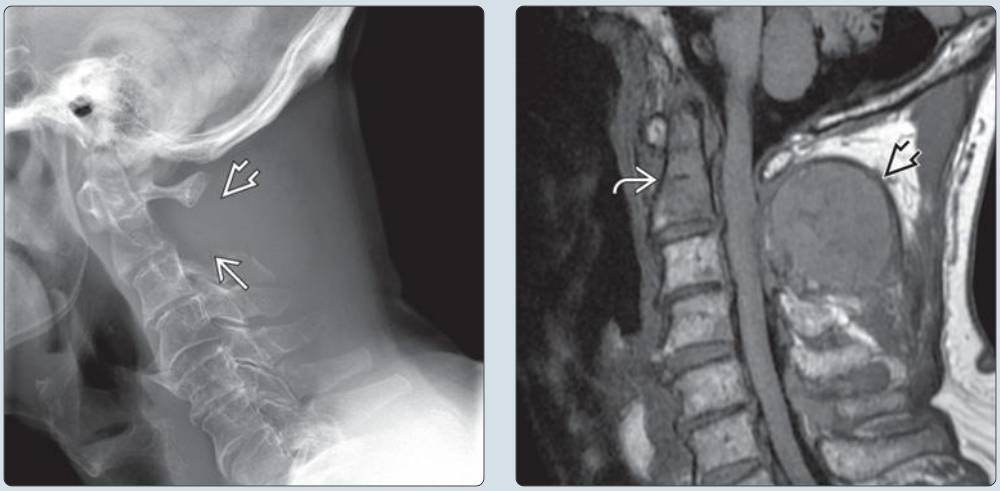
PATHOLOGY

- Spine is most common site of osseous metastases
- Common primaries causing osteolytic metastases
  - Renal, lung, breast, thyroid, GI tract, urothelial, ovarian, melanoma, chordoma, paraganglioma

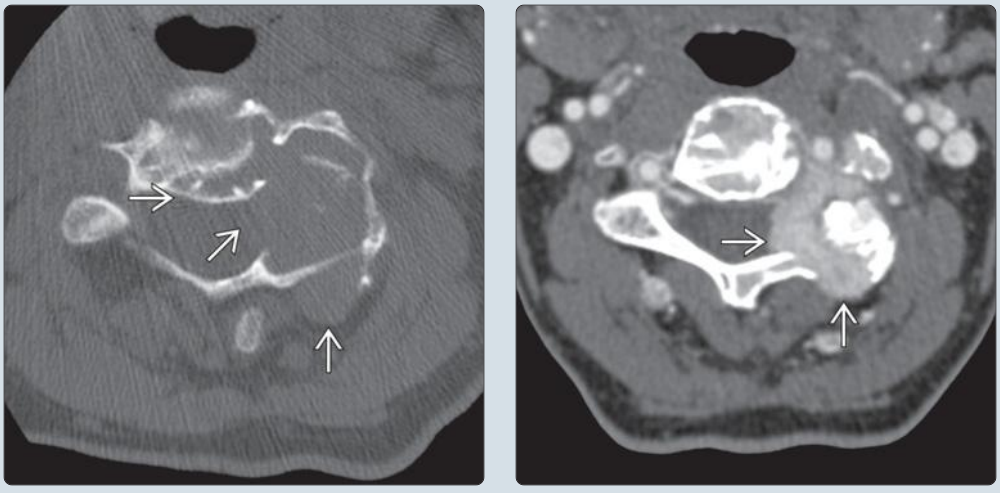
CLINICAL ISSUES

- Pain: Progressive axial, referred, or radicular
- Epidural tumor extension may cause neurologic dysfunction
- Compression fracture
- Cord compression in 5% of adults with systemic cancers (70% solitary, 30% multiple sites)
- Spine metastases found in 5-10% of cancer patients

(Left) *Lateral radiograph shows osteolysis of the C2 neural arch and superior margin of the C3 arch.* **(Right)** *Sagittal T1WI MR in the same patient shows a large soft tissue mass replacing the C2 arch. There is also replacement of marrow in the C2 body and dens. Subsequent work-up disclosed non-small cell lung carcinoma in the right upper lobe.*



(Left) *Axial NECT shows large thyroid carcinoma metastasis to the vertebral body and left facet/lamina of C3. There is a thin rim of expanded bone partially surrounding the lesion. The extraosseous soft tissue mass is not clearly seen.* **(Right)** *Axial contrast-enhanced CT shows enhancement of the mass, typical of hypervascular tumors like thyroid carcinoma. The medial margin of the mass is in the spinal canal, effacing the thecal sac and likely contacting the cervical cord.*



TERMINOLOGY

Synonyms

- Osteolytic metastases

Definitions

- Spread of primary tumor to spine, where bone destruction exceeds bone production

IMAGING

General Features

- Best diagnostic clue
  - Multiple lytic lesions in spine
  - Compression fracture with bowing of posterior cortex, osteolysis extending into neural arch, extraosseous soft tissue
- Location
  - Vertebral body and posterior elements
  - Lesion distribution proportional to red marrow (lumbar > thoracic > cervical)
- Size
  - Any size
- Morphology
  - Typically round (due to centrifugal growth) destruction of bone

Radiographic Findings

- Radiography
  - Requires 50-70% bone destruction and tumor size > 1 cm for detection
    - AP: Absent ("missing") pedicle, ± paraspinous soft tissue mass
    - Lateral: Destroyed posterior cortical line
  - Plain films detect level of neural compression < 25%

CT Findings

- NECT
  - Destruction of cortex and cancellous bone
    - ± compression fracture
  - Paravertebral and epidural soft tissue
    - Rim of hematoma/edema can be seen with benign compression fracture, typically < 5 mm
  - Margins usually indistinct, permeative
- CECT
  - Variable enhancement

MR Findings

- T1WI
  - Hypointense, replacing normal T1 hyperintense marrow
    - Diffuse osseous involvement causes generalized low marrow signal (discs "brighter" than bone)
    - ± compression fracture
  - Paravertebral and epidural soft tissue
    - "Curtain" sign
  - Intervertebral discs generally spared
- T2WI
  - Hypointense → hyperintense to normal marrow
- STIR
  - Greater conspicuity of vertebral metastasis than on fast spin-echo T2WI due to fat suppression
- DWI
  - Hyperintense
  - Controversial utility
    - DWI technically difficult in spine
    - Many reports of elevated DWI signal seem to be actually due to T2 shine-through
- T1WI C+
  - Diffuse enhancement typical
  - Without fat saturation, contrast may decrease conspicuity of metastasis relative to normal fatty marrow

Nonvascular Interventions

- Myelography
  - Osseous destruction as per NECT
  - Extrinsic compression of thecal sac by epidural tumor
    - Better assessment of cord compression than with NECT or CECT
    - Reserve for when patient is unable to undergo MR

Nuclear Medicine Findings

- Bone scan
  - Tracer avidity in regions of bone production at margins of tumor, not directly by lytic tumor
  - False-negatives with aggressively lytic tumor, very small lesions
- PET
  - FDG PET has better specificity but lower sensitivity for detecting malignant bone metastases when compared with bone scan
  - FDG PET/CT has better specificity for detection of malignant involvement of spine than does FDG PET alone

Imaging Recommendations

- Protocol advice
  - MR imaging of entire spinal axis to evaluate for osseous metastasis, epidural tumor, cord compression
    - Include STIR or fat-suppressed T2WI and contrast-enhanced (± fat-saturated) T1WI
  - Bone scan useful for screening of entire skeleton

DIFFERENTIAL DIAGNOSIS

Hematopoietic Malignancy

- Plasmacytoma
- Multiple myeloma (MM)
  - Bone scan negative/equivocal in 25% of MM
- Lymphoma
- Diffuse marrow involvement more common than in metastasis

Benign (Osteoporotic) Compression Fracture

- May be difficult to distinguish acute osteoporotic compression fracture (DWI may be helpful)
  - 1/3 of fractures in patients with known primary tumor are benign
  - 1/4 of fractures in apparently osteopenic patients are from malignant disease
- Marrow signal with late subacute/chronic benign fractures similar to normal marrow (suppresses on STIR)

Schmorl Node

- Circumscribed hypointense T1/hyperintense T2 signal in vertebral body adjacent to disc space

- May have faint rim of enhancement

Normal Heterogeneous Marrow

- Patchy fatty marrow signal in older patients
- Intact pedicle, posterior vertebral cortex, and no epidural soft tissue

Spondylodiscitis

- Destruction of disc space with ↑ fluid signal, enhancement, and abnormal signal in marrow on either side of disc space

PATHOLOGY

General Features

- Etiology
  - Hematogenous dissemination (arterial or venous via Batson plexus) > perineural, lymphatic, CSF spread
  - Marrow infiltration, tumor stimulates osteoclastic response
    - Posterior vertebral body usually 1st, then pedicle
  - Primary tumor, adults: Kidney, lung, breast, thyroid, GI tract, GU tract, ovaries, pancreas, melanoma, chordoma, paraganglioma
    - Unknown primary in 15-25%
  - Primary tumor, children: Ewing sarcoma, other sarcoma, neuroblastoma, hematologic malignancies

Staging, Grading, & Classification

- Surgical stratification (Tokuhashi et al)
  - 6 parameters scored
    - General condition, number of extraspinal bone metastases, number of metastases in vertebral body, metastases to major internal organs, primary site of cancer, severity of spinal cord palsy
  - Excisional operation > 9 points
  - Palliative operation < 5 points
- Surgical stratification (Tomita et al)
  - Grade of malignancy (slow growth, 1 point → rapid growth, 4 points)
  - Visceral metastases (no metastasis, 0 points → untreatable, 4 points)
  - Bone metastases (solitary, 1 point; multiple, 2 points)
    - Total 2-3 points suggests wide excision for long-term local control
    - Total 8-10 points indicates nonoperative supportive care

Gross Pathologic & Surgical Features

- Softened, eroded bone ± adjacent soft tissue mass

Microscopic Features

- Varies with histology of primary, osteoclastic/blastic response

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain: Progressive axial, referred, or radicular
  - Epidural tumor extension may cause neurologic dysfunction
  - Compression fracture
- Other signs/symptoms
  - Hypercalcemia, focal tenderness, soft tissue mass
- Clinical profile
  - Back pain, motor or sensory disturbance in patient with known primary tumor

Demographics

- Age
  - Middle-aged to elderly most common
- Epidemiology
  - Spine most frequent site of skeletal metastases
  - Vertebral metastases
    - 10-40% of patients with systemic cancer
    - Account for 40% of all bone metastases
  - Cord compression in 5% of adults with systemic cancers (70% solitary, 30% multiple sites)
  - Cord compression in 5% of children with malignant solid tumors

Natural History & Prognosis

- Spine metastases found in 5-10% of cancer patients
- Relentless, progressive; pathologic fracture, spinal cord compression
- Prognosis varies with histology of primary lesion

Treatment

- Radiation therapy
- Surgical decompression, stabilization
- Vertebroplasty, embolization

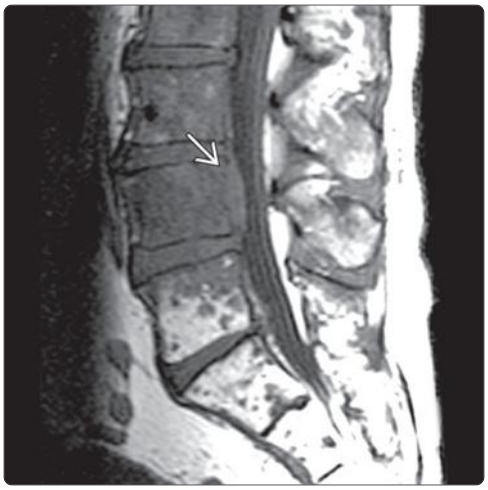
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Persistence of marrow low T1 signal after therapy may be either residual active tumor or fibrosis

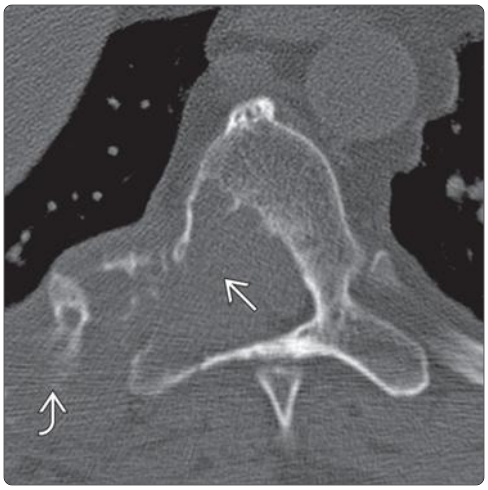
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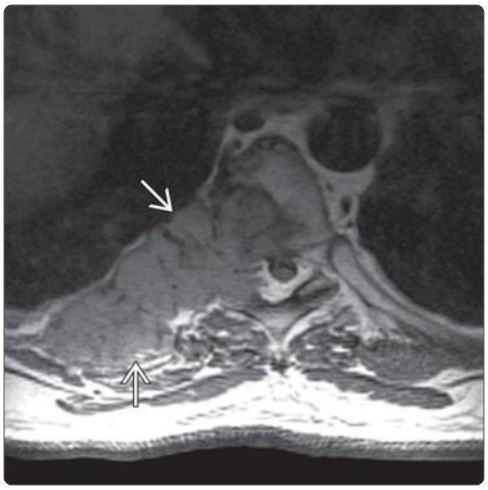
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(Left) *Sagittal T1WI MR shows recurrent disease in this patient with lung carcinoma and prior radiation therapy to spine. Diffuse low signal metastatic disease is present in L2-L5, with epidural extension at L4.* **(Right)** *Sagittal T1WI C+ MR shows multiple low signal metastatic lesions, which are less conspicuous following contrast administration due to diffuse tumor enhancement matching high signal of fatty marrow.*





(Left) *Axial NECT shows a large osteolytic metastasis from renal cell carcinoma destroying the posterior right T7 vertebral body extending into the posterior elements and right T7 rib.* **(Right)** *Axial T1WI MR in the same patient shows the full extent of the soft tissue mass involving the ribs and right side of T7 vertebral body. Minimal intraspinal involvement is seen on this image.*





(Left) *Axial NECT in this patient with metastatic prostate carcinoma shows permeative, lytic mixed with densely blastic metastases. Extraosseous extension is not identified on this slice, and the bony cortex appears preserved.* **(Right)** *Sagittal T1WI MR shows collapse of L3 with diffuse low signal from metastatic disease. There is convex posterior margin to L3, with severe cauda equina compression and anterior paravertebral soft tissue reflecting tumor infiltration.*

Hemangioma

KEY FACTS

TERMINOLOGY

- Common benign venous malformation within vertebrae
- Usually intraosseous, may have epidural component
- Typically, incidental lesion identified on imaging performed for unrelated reasons

IMAGING

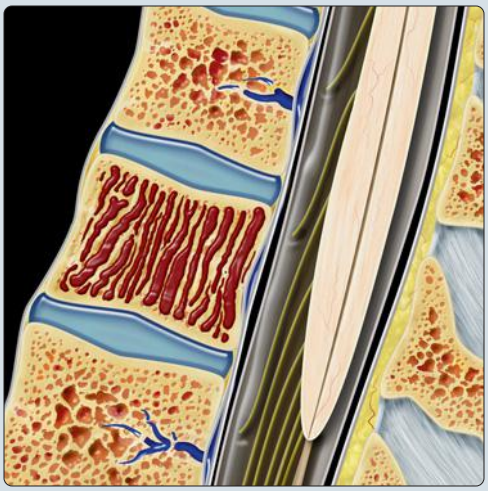
- CT: Well-circumscribed, hypodense lesion with coarse vertical trabeculae ("white polka dots" on axial CT)
- MR: Circumscribed lesion, hyperintense on both T1 and T2WI, with hypointense vertical striations
  - Atypical hemangiomas may have reduced T1 signal due to paucity of fat
- Often multiple (20-30%)
- Bone CT may supplement MR evaluation of suspected hemangioma with atypical imaging features concerning for neoplasm

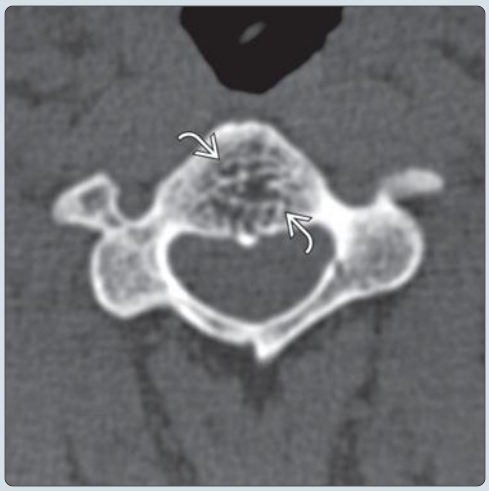
PATHOLOGY

- Complications in < 1%
  - Pathologic compression fracture
  - Epidural hemangioma component with cord compression
- Histology
  - Thin-walled sinusoidal channels lined by vascular endothelium
  - Interspersed bony trabeculae
  - Fatty tissue

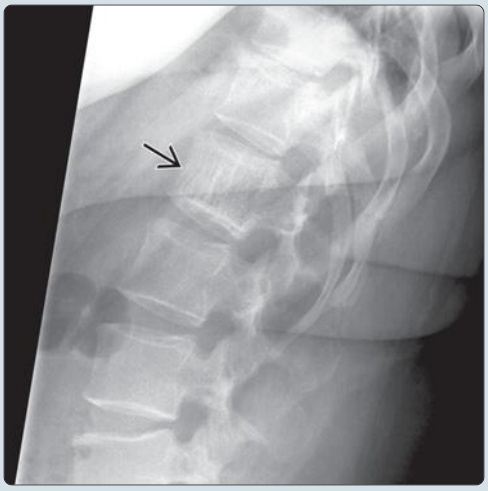
CLINICAL ISSUES

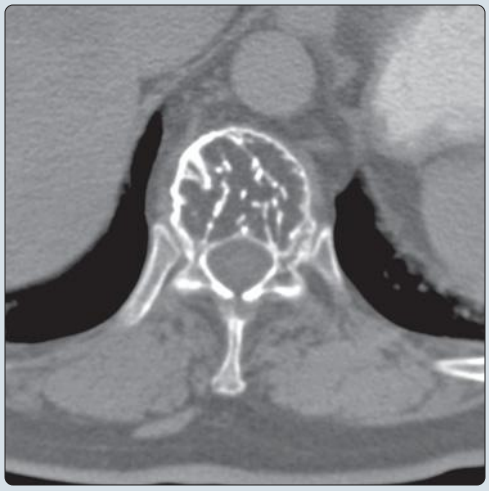
- No follow-up typically necessary with pathognomonic imaging with small lesions and no extraosseous extension





(Left) *Sagittal graphic of the thoracolumbar junction shows the typical striated pattern of hemangioma, with thickened bony trabeculae. There is neither extraosseous extension nor thecal sac compromise.* **(Right)** *Axial NECT shows lucency with interposed thickened bony trabecula, demonstrating characteristic CT findings of a vertebral hemangioma.*





(Left) *Lateral radiograph shows vertical striations within the L1 vertebral body, the so-called "corduroy vertebra," due to vertebral hemangioma.* **(Right)** *Axial CECT shows fatty attenuation within the lower thoracic vertebral body with multiple punctate thickened trabeculae.*

TERMINOLOGY

Definitions

- Common benign venous malformation within vertebrae
  - Usually intraosseous, may have epidural component
  - Rare: Purely epidural spinal hemangioma
- Typically, incidental lesion identified on imaging performed for unrelated reasons

IMAGING

General Features

- Best diagnostic clue
  - CT: Well-circumscribed, hypodense lesion with coarse vertical trabeculae ("white polka dots" on axial CT)
  - MR: Circumscribed lesion, hyperintense on both T1 and T2WI, with hypointense vertical striations
- Location
  - Vertebral body
    - Infrequently extends into posterior elements
  - Often multiple (20-30%)
- Size
  - Variable, may encompass whole vertebral body
- Morphology
  - Well-circumscribed margins, thickened trabeculae
  - Infrequently, associated epidural soft tissue

Radiographic Findings

- Radiography
  - Vertebral body lesion with coarse vertical trabeculae resembling corduroy or honeycomb

CT Findings

- NECT
  - Hypodense vertebral lesion
  - Thickened, vertically aligned trabecula
    - "Polka dot" appearance on axial images

MR Findings

- T1WI
  - Typical (fatty stroma): Hyperintense
  - Atypical (mostly vascular soft tissue): Isointense or hypointense
    - Atypical hemangioma may simulate vertebral metastasis
- T2WI
  - Usually hyperintense, due to vascular elements
- T1WI C+
  - Variable enhancement
    - Milder enhancement typically seen in lesions with larger fatty component
- "Aggressive" hemangioma features
  - T1 isointense to hypointense, with avid contrast enhancement
  - Extraosseous soft tissue extension
  - Pathologic fracture

Angiographic Findings

- Variable enhancement
  - Angiography unnecessary unless embolization is being considered

Nuclear Medicine Findings

- Bone scan
  - Studies usually normal, lesions not visualized

Imaging Recommendations

- Best imaging tool
  - Pathognomonic findings usually present on either CT and MR
- Protocol advice
  - Bone CT may supplement MR evaluation of hemangioma with atypical imaging features concerning for neoplasm

DIFFERENTIAL DIAGNOSIS

Focal Fatty Marrow

- Incidental rounded focus of marrow fat conspicuous on MR imaging
- Fat-suppressed sequences (e.g., STIR) will show marked lesion hypointensity; hemangiomas typically retain some high signal due to vascular components

Vertebral Metastasis

- Hypointense on T1WI, hypointense to hyperintense (to marrow) on T2WI
- May be difficult to distinguish from "aggressive" hemangioma; consider CT
- Typically multiple

Paget Disease

- Expanded body with thickened low signal cortex, heterogeneous signal containing fat
- No regular pattern of vertically oriented trabeculae
- No epidural soft tissue extension (unless degenerated into sarcoma)

Degenerative Endplates, Type II

- Abuts intervertebral disc
- Adjacent degenerate disc with endplate spondylosis

Spinal Radiation Treatment

- Fatty marrow replacement throughout vertebral body
- Conforms to radiation port
- No coarse, vertical trabeculations as in hemangioma

PATHOLOGY

General Features

- Etiology
  - Developmental, hamartomatous lesions
- Genetics
  - Rare: Krev interaction-trapped 1 (*KRIT1*) gene in families with cerebral cavernous malformations, autosomal dominant
- Associated abnormalities
  - Complications in < 1%
    - Pathologic compression fracture
    - Epidural hemangioma component with cord compression

Gross Pathologic & Surgical Features

- Majority confined to vertebral body proper
  - May be small or occupy entire vertebral body

- Uncommonly involve posterior elements/pedicles (10-15%)
- Thoracic lesions are more often aggressive than those at other locations

Microscopic Features

- Thin-walled sinusoidal channels lined by vascular endothelium
- Interspersed bony trabeculae
- Fatty tissue
  - Atypical lesions contain less fat and more vascular stroma
- Slow venous flow
- Similar to venous malformations

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Incidental finding; present in 11% of general population
  - Aggressive lesions can present with pathologic compression fracture or spinal cord compression
    - Rare: Enlargement of hemangioma during pregnancy resulting in cord compression
- Clinical profile
  - Incidental finding on spine imaging ordered for unrelated reason

Demographics

- Age
  - Peak incidence 4th to 6th decades
- Gender
  - Similar incidence, or up to 2x as frequent in females
- Epidemiology
  - Most common spinal tumor
  - 11% of adult population
  - 25-30% multiple, particularly in thoracic spine

Natural History & Prognosis

- Benign (fatty) hemangiomas: Incidental lesions, no clinical sequela
- Aggressive vascular hemangiomas: Variable depending on size of lesion, degree of epidural extension, and presence/absence of cord compression

Treatment

- No follow-up typically necessary with pathognomonic imaging
- Annual neurological and radiological examinations for patients with hemangiomas associated with pain may be appropriate
- Aggressive hemangiomas
  - Direct intralesional injection of ethanol (post ethanol treatment body collapse reported complication)
  - Embolization with Onyx recently reported
  - Vertebroplasty in conjunction with embolization may be considered if concern for pathologic fracture
  - Surgical resection (corpectomy) or radiation therapy have also been utilized

DIAGNOSTIC CHECKLIST

Consider

- Atypical (fat-poor) hemangioma in low-risk patient with solitary, indeterminate vertebral lesion on MR

Image Interpretation Pearls

- Focused CT of suspected atypical (fat-poor) hemangioma to identify characteristic osseous findings

SELECTED REFERENCES

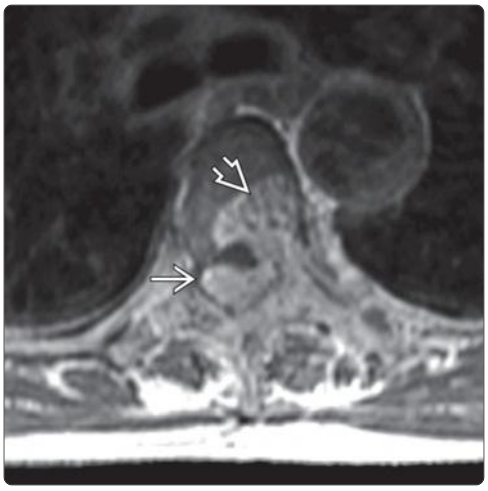
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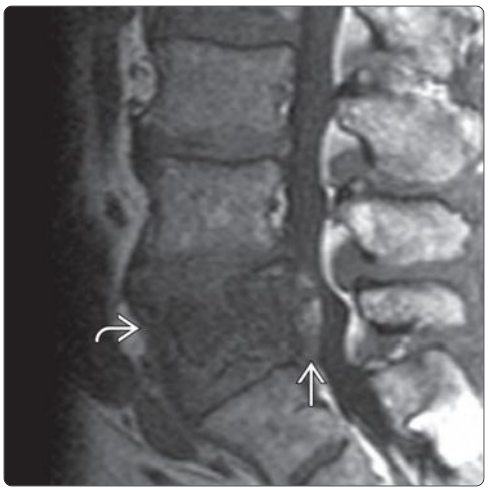
(Left) *Sagittal T1WI MR of the cervical spine shows T1 hyperintensity involving majority of the C6 vertebral body, due to a large vertebral hemangioma.* **(Right)** *Sagittal T2WI MR in the same patient shows the hemangioma to have a diffusely hyperintense signal. Fine vertical striations correspond to the thickened trabecula that can be seen on CT with these lesions.*





(Left) *Sagittal T1 C+ MR shows prominent vertical striations within the vertebral body consistent with hemangioma. There is extraosseous extension of the tumor into the ventral epidural space.* **(Right)** *Axial T1WI C+ MR shows enhancing mass involving posterior thoracic body and posterior elements, with large amount of epidural extension and cord compression. Thickened low signal trabeculae give a clue to the diagnosis of aggressive hemangioma with bone and soft tissue involvement.*





(Left) *Axial T2WI MR through a lumbar vertebral body shows a circumscribed hyperintense lesion. Finely speckled hypointense foci correspond to the thickened trabecula that can be seen on CT.* **(Right)** *Sagittal T1 MR shows lipid-poor hemangioma of the L5 as it manifests with diffuse T1 hypointensity throughout the vertebral body. There is contiguous ventral epidural soft tissue effacing the thecal sac, with mixed fat and soft tissue signal, representing extraosseous extension of the hemangioma.*

Osteoid Osteoma

KEY FACTS

TERMINOLOGY

- Benign osteoid-producing tumor < 1.5 cm in size
- Tumor often called "nidus" to distinguish it from surrounding reactive zone due to host response

IMAGING

- 10% of OO occur in spine, in neural arch
- Focal scoliosis, concave on side of tumor
- Central nidus
  - Variable amount of ossification
- Reactive zone
  - Dense sclerosis, edema around nidus
  - Involves much larger area than tumor
  - Periosteal reaction variably present
  - Soft tissue mass or pleural thickening/effusion
  - Low signal on T1WI, high signal on T2WI, STIR
  - Enhances with gadolinium, iodinated contrast

TOP DIFFERENTIAL DIAGNOSES

- Osteoblastoma
- Stress fracture of pedicle or lamina
- Unilateral spondylolysis
- Unilateral absent pedicle or pars interarticularis
- Sclerotic metastasis
- Lymphoma
- Osteomyelitis
- Ewing sarcoma

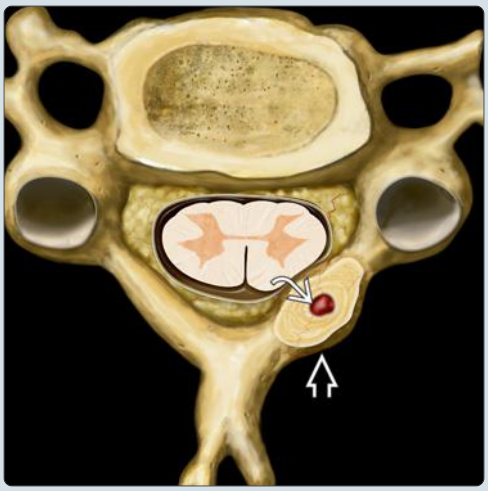
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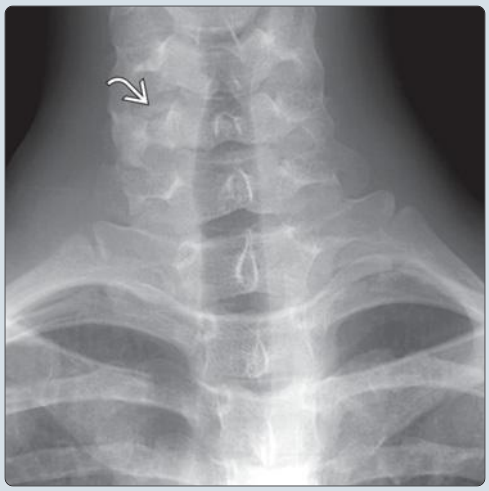
- Night pain relieved by aspirin, NSAIDs
- 70% have scoliosis related to muscle spasm, concave on side of tumor

DIAGNOSTIC CHECKLIST

- Thin section CT most accurate in visualizing nidus
- Edema on MR mimics infection, malignancy

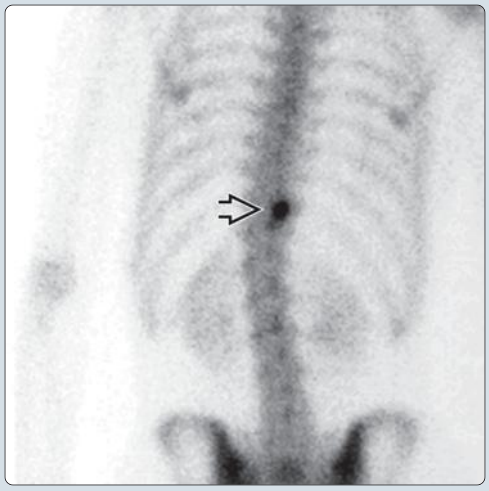
(Left) *Axial graphic shows a small, highly vascular tumor nidus of osteoid osteoma in the left lamina, surrounded by dense reactive bone.* **(Right)** *Anteroposterior radiograph shows focal, short-curve levoscoliosis in a young man with neck pain and no trauma history. Painful scoliosis raises concern for a tumor or infection. Sclerosis at C6 on the right suggested a possible diagnosis of OO.*





(Left) *Axial bone CT in the same patient shows a sclerotic C6 OO nidus. Nidus may range from purely radiolucent to completely sclerotic. The lesion is sharply demarcated and surrounded by reactive sclerosis. The lesion was missed on a prior routine cervical spine MR.* **(Right)** *Posteroanterior bone scan (3-hour image) shows scoliosis and intense, focal uptake on the right at a T11 osteoid osteoma. Scoliosis due to osteoid osteoma is always concave on the side of the tumor.*





TERMINOLOGY

Abbreviations

- Osteoid osteoma (OO)

Definitions

- Benign osteoid-producing tumor < 1.5 cm in size
- Tumor often called "nidus" to distinguish it from surrounding reactive zone due to host response

IMAGING

General Features

- Best diagnostic clue
  - Small radiolucent tumor nidus with surrounding sclerosis
- Location
  - 10% of OO occur in spine
  - Almost all involve neural arch
  - 59% lumbar, 27% cervical, 12% thoracic, 2% sacrum
- Size
  - < 1.5 cm nidus; larger lesions called osteoblastoma
- Morphology
  - Nidus round or oval

Radiographic Findings

- Radiography
  - Central, lucent tumor nidus often obscured by reactive sclerosis
  - Focal scoliosis concave on side of tumor

CT Findings

- CECT
  - Variable enhancement of nidus and reactive zone
    - Contrast administration may obscure bony matrix
- Bone CT
  - Central nidus
    - Variable amount of ossification in nidus
    - Nidus usually predominantly lucent
    - Occasionally nidus is sclerotic
  - Reactive zone
    - Dense sclerosis around tumor
    - Periosteal reaction variably present, usually unilaminar
    - Soft tissue mass or pleural thickening/effusion often seen
    - Bones adjacent to OO may show sclerosis, periosteal reaction
      - Ribs, adjacent vertebrae affected
    - Ossification of ligamentum flavum has been reported

MR Findings

- Central nidus
  - Low signal intensity on T1WI
  - Variable low to high signal intensity on T2WI, STIR
  - Avid, rapid enhancement with gadolinium
- Reactive zone
  - Edema surrounding nidus, due to prostaglandin release
    - Low signal on T1WI, high signal on T2WI, STIR
    - Enhances with gadolinium
      - Dynamic scan may show delayed enhancement relative to nidus
  - Involves much larger area than tumor
  - May lead to misdiagnosis of malignancy or infection
  - Periosteal reaction and cortical thickening common
  - May extend to adjacent vertebrae, ribs, and paraspinous soft tissues
    - Pleural thickening, enhancement, and effusion common with thoracic OO
- Literature reports high rate of missed MR diagnosis of OO
  - Lesion may not be seen due to small size of nidus
    - Thin shape, oblique orientation of posterior elements
    - Partial volume averaging with adjacent structures
    - Improve visualization with thin slices, 3-3.5 mm
    - Axial, coronal, and sagittal planes
  - Lesion misidentified based on appearance of reactive zone rather than nidus
    - Misdiagnosis of infection or malignancy common

Nuclear Medicine Findings

- Bone scan
  - Positive on all 3 phases of Tc-99m MDP bone scan

Imaging Recommendations

- Best imaging tool
  - CT scan

DIFFERENTIAL DIAGNOSIS

Osteoblastoma

- Larger (> 1.5 cm)
- Expansile lesion of neural arch/pedicle

Stress Fracture of Pedicle or Lamina

- Sclerosis around fracture mimics reactive sclerosis around osteoid osteoma
- CT with reformatted images or high-resolution MR will show fracture line
- May occur in patients with preexisting scoliosis, due to altered stresses
- Pain related to activity, improves at night

Unilateral Spondylolysis

- Linear defect pars interarticularis
- Contralateral side is sclerotic
- Painful
- Presents in young patients
- Scoliosis may develop
- May be diagnosed on oblique radiographs, CT

Unilateral Absent Pedicle or Pars Interarticularis

- Contralateral side is sclerotic
- Congenital absence of pedicle or pars interarticularis confirmed on CT

Sclerotic Metastasis

- Older patients

Lymphoma

- Older patients
- Often involves pedicle, destroys posterior body cortex
- Poorly defined, wide zone of transition
- Associated soft tissue mass common
- Often presents with night pain relieved by aspirin, NSAIDs

Osteomyelitis

- Sequestrum or focal abscess can mimic nidus of osteoid osteoma
  - Tends to have irregular shape, unlike round OO nidus
- Usually involves vertebral bodies rather than neural arch
- CT usually shows endplate destruction or destructive arthritis of facet joints
- Often presents with night pain relieved by aspirin, NSAIDs

Ewing Sarcoma

- Marrow replacement looks similar on MR
  - Centered in vertebral body
  - Diffuse edema may involve adjacent vertebrae and ribs

PATHOLOGY

General Features

- Etiology
  - Benign tumor of osteoblastic origin

Gross Pathologic & Surgical Features

- Sharply demarcated, round, pink-red mass (nidus)
- Nidus can be shelled out from surrounding sclerotic, reactive bone that does not contain tumor

Microscopic Features

- Web of osteoid trabeculae showing variable amounts of mineralization
- Vascular, fibrous connective tissue
- Similar histologically to osteoblastoma
- No malignant potential
- Reactive zone may contain lymphocytes and plasmocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Night pain relieved by aspirin, NSAIDs
- Clinical profile
  - 70% have scoliosis related to muscle spasm, concave on side of tumor
  - Gait disturbance, muscle atrophy, torticollis sometimes seen

Demographics

- Age
  - Majority occur in 2nd decade of life
  - Reported as late as 7th decade, but very rare
- Gender
  - M:F = 2-3:1
- Epidemiology
  - 12% of all benign skeletal neoplasms

Natural History & Prognosis

- Resection is curative in most cases
  - Entire nidus must be removed or recurrence is probable
  - Radionuclide labeling can be used to localize intraoperatively
- Spontaneous healing has been reported

Treatment

- Open excision
  - Proximity to vertebral artery should be noted in cervical OO
- CT-guided percutaneous excision
- Thermo-/photocoagulation
- Conservative observation (patients with well-controlled symptoms)

DIAGNOSTIC CHECKLIST

Consider

- Important cause of painful scoliosis in child or young adult

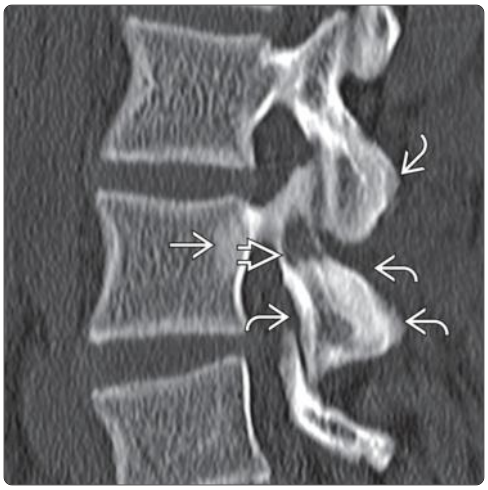
Image Interpretation Pearls

- Thin-section CT most accurate in visualizing nidus
- Edema on MR mimics infection, malignancy

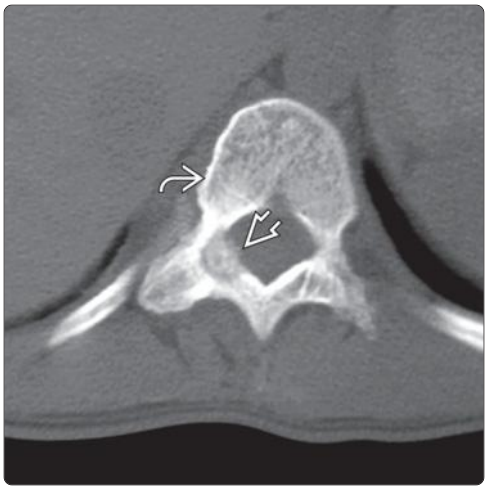
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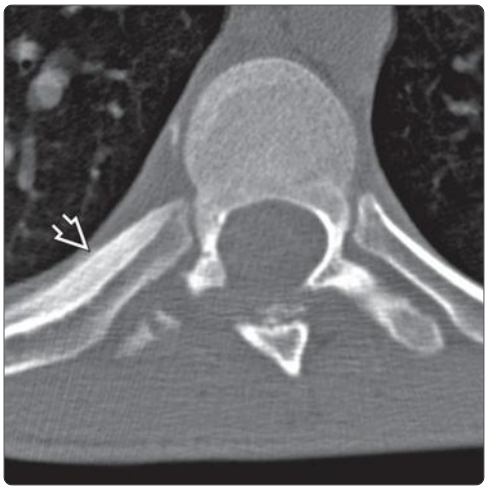
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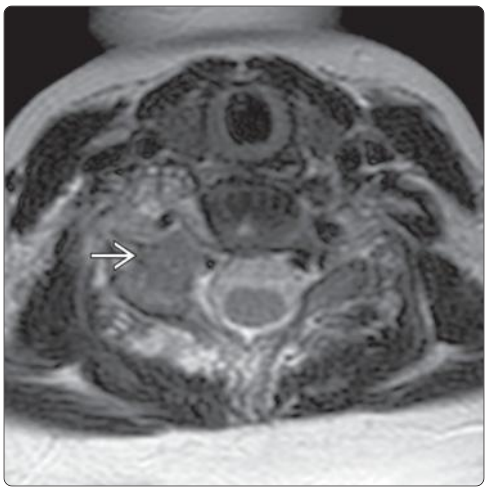


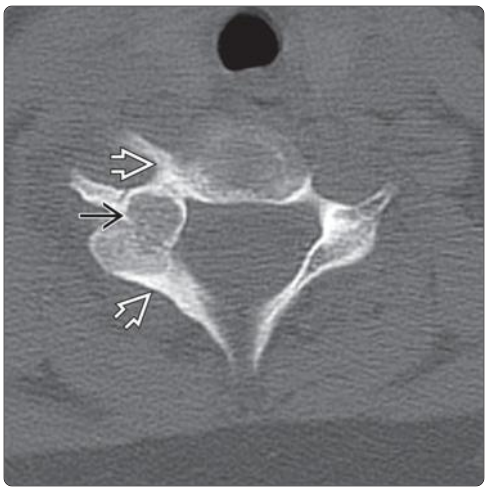
(Left) *Axial bone CT shows OO that is purely lytic and appears inconspicuous relative to an enlarged, sclerotic pedicle and transverse process. Sclerosis extends into the vertebral body.* **(Right)** *Sagittal bone CT in the same patient shows reactive vertebral body sclerosis, which might easily be mistaken for the lesion of concern. Nidus is centered in pars interarticularis. There is extensive, thick periosteal reaction that also involves the adjacent vertebral level.*





(Left) *Axial bone CT shows a sclerotic OO nidus centered in the lamina. Reactive sclerosis extends into the vertebral body.* **(Right)** *Axial bone CT in the same patient shows a thick, uniform reactive periosteal reaction of the rib at the level above the vertebra with OO. Mild pleural thickening is also visible adjacent to the rib.*





(Left) *Axial T2WI MR shows an ill-defined intermediate signal intensity mass in the right C6 neural foramen. The mass appears poorly marginated, mimicking malignancy. Because of the patient's age, osteoid osteoma was considered and a CT scan was performed.* **(Right)** *Axial bone CT in the same patient shows a lytic OO nidus with surrounding host sclerosis accounting for edema on MR.*

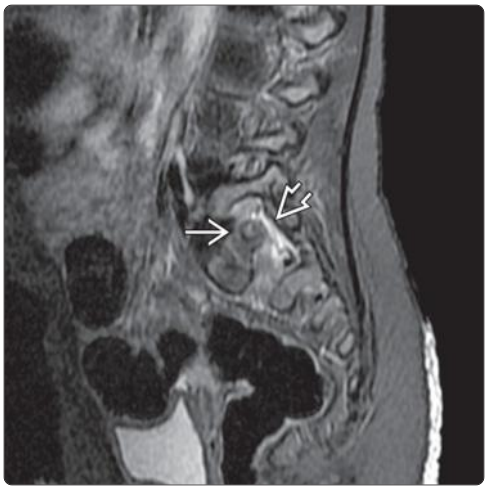
(Left) *Axial T1W MR shows an ill-defined lesion within the posterior S1 body just anterior to the superior articular facet in this child with back pain. There is adjacent inflammatory change in the epidural space.* **(Right)** *Anteroposterior bone scan shows intense focal uptake in the left side of the S1 body.*



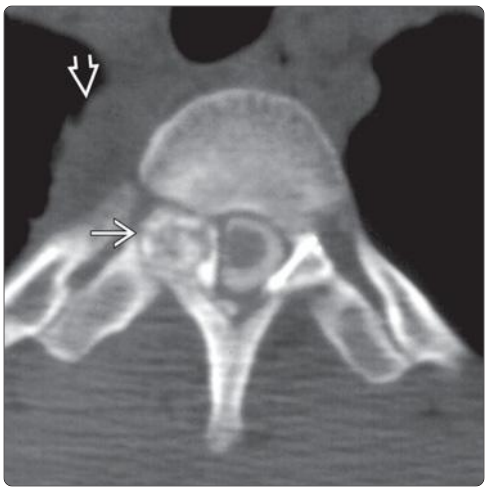


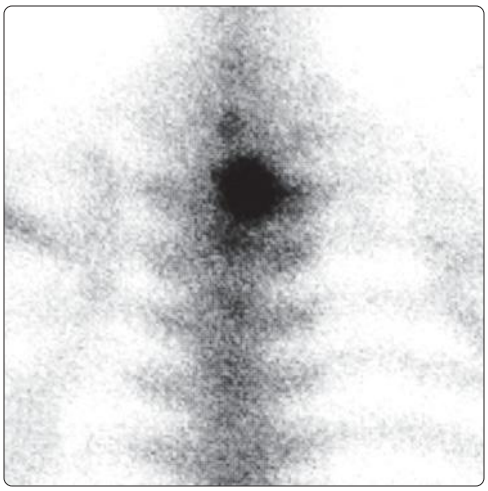
(Left) *Sagittal T1WI MR shows a target-like nidus within the posterior S1 body just anterior to the superior articular facet.* **(Right)** *Sagittal T2WI MR shows a target-like nidus of OO with an adjacent marrow increased signal from an inflammatory reaction.*



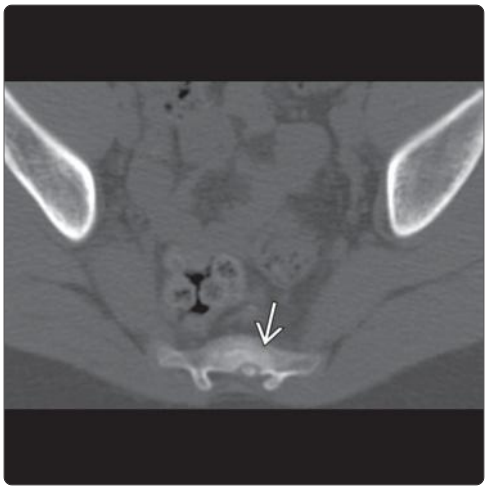


(Left) *Axial CT myelogram shows the characteristic, sharply demarcated tumor nidus of OO. Local pleural reaction is commonly seen in thoracic osteoid osteomas.* **(Right)** *Posterior bone scan shows not only marked uptake by the tumor itself but a "corona" of increased uptake involving the adjacent bones. This reflects the local response to prostaglandin secreted by the tumor.*

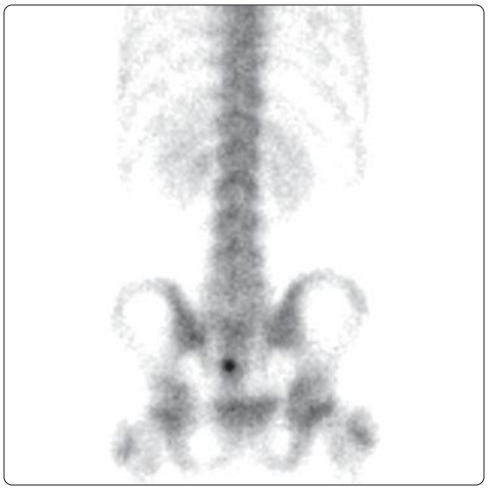


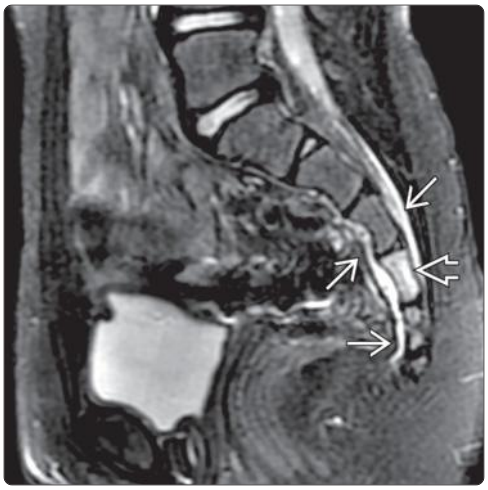




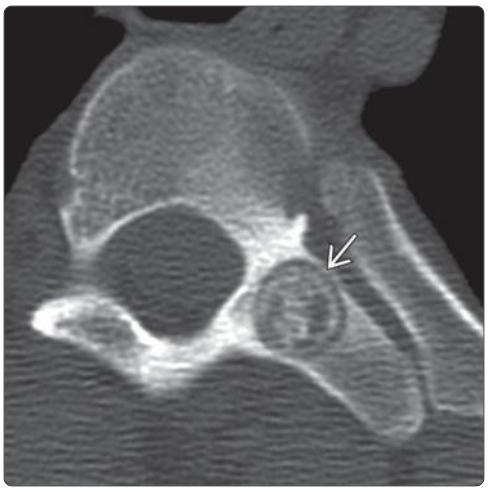


(Left) *Coronal NECT (bone window) of the sacrum in this child shows the "bull's eye" sclerotic nidus ➡ that is typical for OO surrounded by lucency and sclerosis.* **(Right)** *Axial NECT (bone window) shows a well-defined sclerotic nidus with a target sign of surrounding lucency and adjacent sclerosis, typical for OO ➡.*





(Left) *Posterior bone scan shows a single small area of intense uptake in the sacrum.* **(Right)** *Sagittal STIR MR shows an increased signal of S4 ➡ and adjacent parasacral soft tissues ➡ from inflammatory reaction of OO.*





(Left) *Axial NECT (bone window) shows a partially calcified well-defined nidus at the base of the left transverse process of T3 ➡. Note the adjacent sclerosis.* **(Right)** *Axial NECT (bone window) shows an ill-defined extradural soft tissue lesion arising from the dorsal vertebral body ➡ with surrounding sclerotic change. OOs may be subtle, and are easiest to detect on bone CTs and fat-suppressed T2 MR or STIR MR techniques.*

Osteoblastoma

KEY FACTS

TERMINOLOGY

- Benign tumor that forms osteoid
- Differentiated grossly from osteoid osteoma by larger size (> 1.5 cm)

IMAGING

- 40% of osteoblastomas (OB) occur in spine
- Well-circumscribed, expansile lesion of neural arch
  - Frequent extension into vertebral body
  - Narrow zone of transition, sclerotic rim
- Periosteal inflammatory response of adjacent ribs, pleural thickening, effusion
- Peritumoral edema (flare phenomenon)
  - Edema enhances, obscures tumor margins, mimics malignancy (MR)

TOP DIFFERENTIAL DIAGNOSES

- Osteoid osteoma (OO)
- Aneurysmal bone cyst (ABC)
- Metastasis
- Osteogenic sarcoma (OGS)
- Chordoma
- Infection

PATHOLOGY

- Tumor prostaglandins release causes extensive peritumoral edema

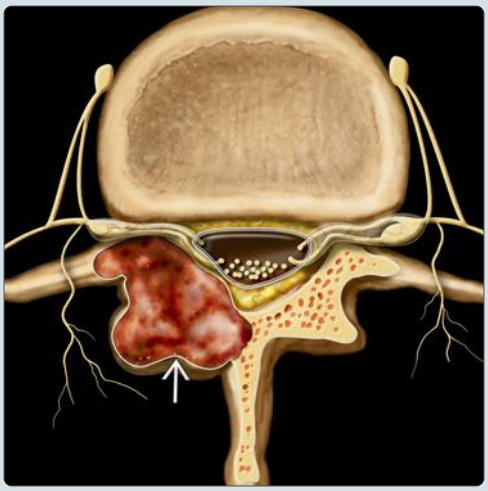
CLINICAL ISSUES

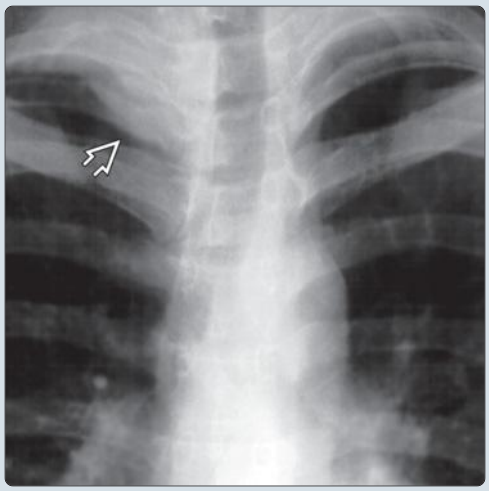
- 90% in 2nd-3rd decades of life
- Dull, localized pain
- Painful scoliosis
- Neurologic symptoms due to compression of cord, nerve roots

DIAGNOSTIC CHECKLIST

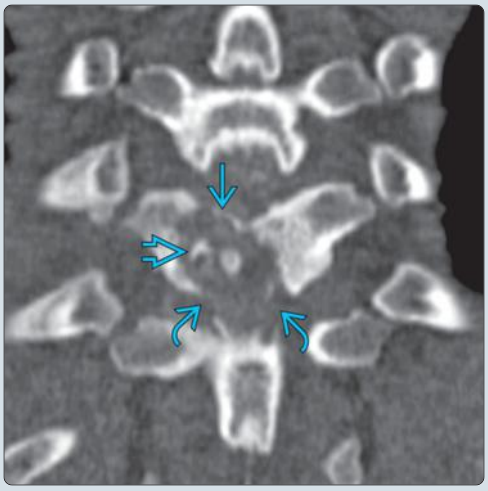
- May be occult on radiographs; consider MR in young patients with painful scoliosis

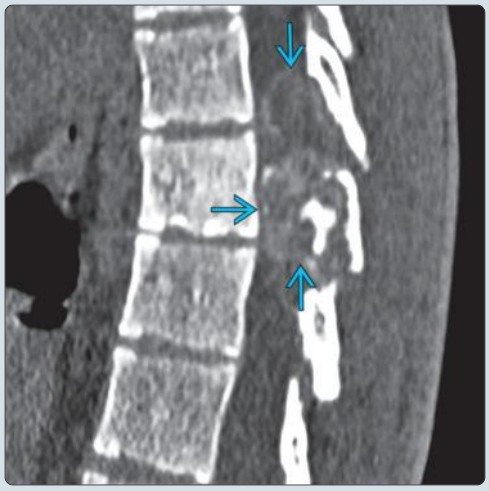
(Left) *Axial graphic shows expansile, highly vascular osteoblastoma (OB) arising in the right lamina and impinging on the exiting nerve root.* **(Right)** *Anteroposterior radiograph shows focal levoscoliosis secondary to upper thoracic OB. The tumor is not visible. Note focal pleural thickening, a secondary sign of OB. The patient also had upper thoracic pain. This combination of symptoms and radiographic findings should prompt CT evaluation.*





(Left) *Coronal bone CT shows expansile OB of the right lamina. The matrix mimics "rings and arcs" of chondroid matrix. Cortical breakthrough is evident at the inferior tumor margin.* **(Right)** *Sagittal bone CT shows expansile lower thoracic OB extending from the lamina into spinal canal. The lesion is sharply circumscribed and bilobed, involving 2 adjacent levels.*





TERMINOLOGY

Definitions

- Benign tumor forming osteoid
- Differentiated grossly from osteoid osteoma by larger size (> 1.5 cm)

IMAGING

General Features

- Best diagnostic clue
  - Expansile mass occurring in posterior elements
- Location
  - 40% of osteoblastomas (OB) occur in spine
    - 40% cervical, 25% lumbar, 20% thoracic, 15-20% sacrum
  - Originate in neural arch
  - May be centered in pedicle, lamina, transverse or spinous process, articular pillar, or pars interarticularis
  - Often extend into vertebral body
- Size
  - > 1.5 cm

Radiographic Findings

- Radiography
  - Geographic, expansile lesion
  - Matrix often not visible on spine radiographs, better seen on CT
  - Usually radiolucent, occasionally sclerotic
  - Subtle imaging findings
    - Compare posterior elements to contralateral side and to levels above and below area of concern
  - AP radiograph
    - Lucent or sclerotic pedicle
    - Expansion of transverse process
    - Scoliosis in 50-60% of cases
    - Scoliosis concave on side of tumor
  - Lateral radiograph
    - Expanded pedicle, lamina, or spinous process
    - Sharply demarcated lucency in posterior portion of vertebral body

CT Findings

- CECT
  - Heterogeneous enhancement obscures bone matrix
- Bone CT
  - Most common appearance
    - Well-circumscribed, expansile lesion of neural arch
    - Often extends into vertebral body
    - Narrow zone of transition, sclerotic rim
  - Aggressive OB
    - Cortical breakthrough, wide zone of transition
    - Difficult differential diagnosis with osteosarcoma
  - Matrix mineralization
    - Variable amount of mineralization
    - May see small, irregular trabeculae
    - Can be difficult to distinguish from calcified cartilage of chondrosarcoma, enchondroma
  - Inflammatory response may spread far beyond lesion
    - Widespread, ill-defined sclerotic bone around lesion
    - Periosteal reaction of adjacent ribs
    - Pleural thickening &/or effusion
    - Ossification of ligamentum flavum

MR Findings

- Low/intermediate signal intensity on T1WI
- Low to high signal intensity on T2WI, STIR
  - Depending on extent of ossification in tumor
- Variable enhancement with gadolinium
- Peritumoral edema is prominent (flare phenomenon)
  - Due to prostaglandin secretion
  - Edema enhances, obscures tumor margins
  - May mimic malignancy
  - May involve adjacent bones and soft tissues
    - Pleural effusion is common
- Periosteal reaction common in involved or adjacent bone
  - Due to prostaglandin excretion
- Bone matrix is often not visible
  - May see small, irregularly shaped areas of low signal intensity on all sequences
- Tumor margins often obscured by surrounding bone marrow edema
  - Better seen on CT scan
- Often associated with aneurysmal bone cyst
  - Fluid-fluid levels
- Epidural hematoma with minor trauma has been reported

Angiographic Findings

- Intense tumor blush

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan

Imaging Recommendations

- CT with sagittal, coronal reformations

DIFFERENTIAL DIAGNOSIS

Osteoid Osteoma (OO)

- Smaller (< 1.5 cm)
- Round nidus with surrounding sclerotic bone
- Same age group
- Pain is usually more intense
- Scoliosis is common

Aneurysmal Bone Cyst (ABC)

- Expansile lesion of posterior elements
- ABC component present in 10-15% of OB
- ABC may also be isolated or associated with other tumors
- Multiple blood-filled cavities with fluid-fluid levels
- Matrix absent in ABC without OB

Metastasis

- Older patients
- Usually destroys cortex rather than expanding
- May be expansile, especially in renal cell carcinoma
- Involves posterior elements &/or vertebral body

Osteogenic Sarcoma (OGS)

- Sarcoma containing bone matrix
- Rare in spine
- More aggressive appearance on radiographs, CT
- Wider zone of transition

- Cortical breakthrough rather than cortical expansion
- Involves neural arch &/or vertebral body

Chordoma

- Involves vertebral body rather than posterior elements
- Common in sacrum, rare in vertebrae
- No matrix; purely lytic tumor

Infection

- MR appearance of OB may mimic infection because of inflammatory change in adjacent bones
- Distinction between OB and infection easily made on CT

Fibrous Dysplasia

- Rare in spine
- Expansile lesion of posterior elements
- May be lytic or contain ground-glass matrix or tiny trabeculae of bone

Chondrosarcoma

- Rare in spine
- Involves vertebral body &/or posterior elements
- May have fairly unaggressive features on imaging studies
- Cartilage calcification in arcs and rings

PATHOLOGY

General Features

- Prostaglandins released by tumor cause extensive peritumoral edema
- Pain usually less intense than with OO

Staging, Grading, & Classification

- Classic OB
- Aggressive OB
  - Borderline lesion with osteosarcoma
  - Locally aggressive but does not metastasize

Gross Pathologic & Surgical Features

- Friable, highly vascular tumor
- Red color due to vascularity
- Well demarcated from surrounding host bone

Microscopic Features

- Prominent osteoblasts
- Rims of osteoblasts along trabeculae
- Vascular fibrous stroma
- Woven bone matrix of variable quantity
- ABC component found in 10-15%
- "Aggressive" OB
  - Same features as above, plus
    - More nuclear pleomorphism
    - Epithelioid osteoblasts

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Dull, localized pain
  - Neurologic symptoms due to compression of cord, nerve roots
- Painful scoliosis

Demographics

- Age
  - 90% in 2nd-3rd decades of life
  - Have been diagnosed in patients up to 7th decade of life
- Gender
  - M:F = 2-2.5:1

Natural History & Prognosis

- Grow slowly
- 10-15% recurrence for typical OB
- 50% recurrence for aggressive OB

Treatment

- Curettage with bone graft or methylmethacrylate placement
- Preoperative embolization may be useful

DIAGNOSTIC CHECKLIST

Consider

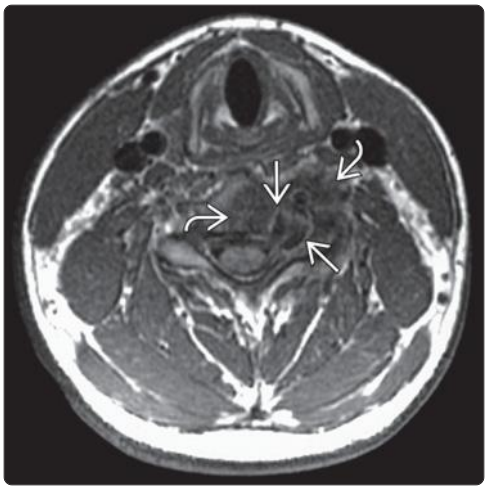
- May be occult on radiographs; consider MR in young patients with painful scoliosis

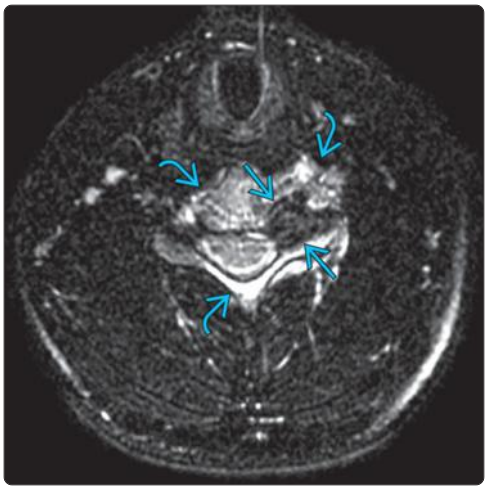
Image Interpretation Pearls

- Aggressive OB is difficult on imaging and histology to distinguish from osteogenic sarcoma
  - Wide zone of transition suspicious for osteogenic sarcoma
  - Cortical breakthrough is common in OB
    - Without wide transition zone, should not be considered an indication of malignancy

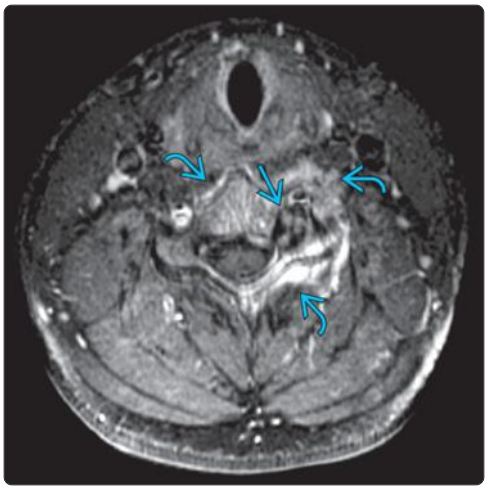
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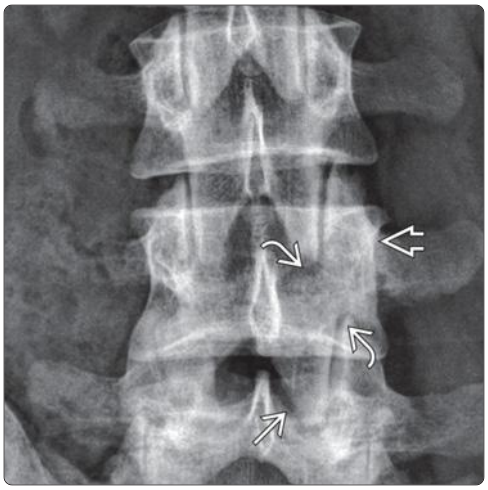
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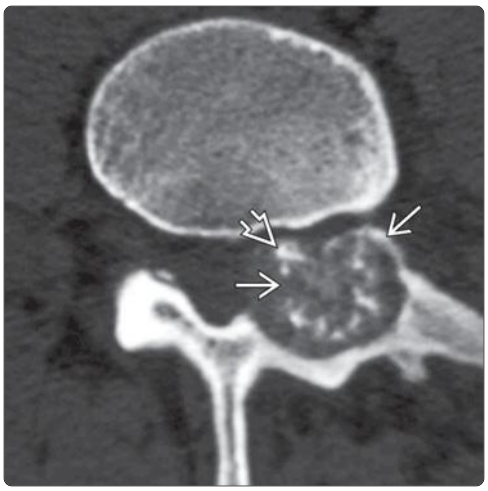


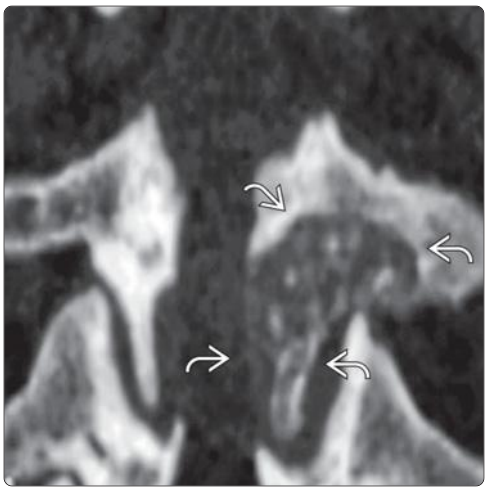
(Left) *Axial T1WI MR shows a low signal intensity, densely ossified mass expanding the left pedicle. There is extensive reactive edema in the adjacent posterior elements, vertebral body, and soft tissues that mimic a more aggressive tumor.* **(Right)** *Axial T2WI FS MR shows that the central, densely ossified tumor remains low signal intensity, while the surrounding edema is high signal intensity.*





(Left) *Axial T1WI C+ FS MR in the same patient shows enhancement of nonossified portions of tumor as well as the peritumoral edema.* **(Right)** *Anteroposterior radiograph shows a lytic lesion involving the left L4 pars, pedicle, and inferior articular facet. OB presents with vague pain, and radiographs are often the modality initially requested for evaluation. Evaluation of radiographs should always include comparing density of posterior element structures to adjacent levels.*





(Left) *Axial bone CT in the same patient shows an expansile mass containing thin, irregular bone trabeculae characteristic of OB. Although cortical breakthrough is present, the zone of transition to adjacent bone is narrow and sclerotic.* **(Right)** *Coronal bone CT in the same patient is useful to confirm the tumor extent from the transverse process to the inferior articular facet. Compare this image to the AP radiograph to better understand the radiographic findings.*

Aneurysmal Bone Cyst

KEY FACTS

IMAGING

- 10-30% of aneurysmal bone cysts (ABCs) occur in spine and sacrum
- Centered in neural arch, extends into vertebral body
- Balloon-like expansile remodeling of bone
  - Thinned, "eggshell" cortex
  - Focal cortical destruction common
- Absent pedicle sign: Expansion of pedicle results in loss of pedicle contour on AP radiographs
- Contains multiple round cysts with fluid-fluid levels
  - Caused by hemorrhage, blood product sedimentation
- Blood-filled cysts separated by septa of varying thickness
  - Periphery and septa enhance
  - Solid ABC variant enhances diffusely
- Calcified tumor matrix absent
- Narrow, nonsclerotic zone of transition with adjacent bone
- CT best to differentiate from telangiectatic osteogenic sarcoma
  - Narrow zone of transition in ABC
  - Absence of infiltration into surrounding soft tissues
- MR shows epidural extent, cord compromise

TOP DIFFERENTIAL DIAGNOSES

- Osteoblastoma
- Telangiectatic osteogenic sarcoma
- Metastases
- Giant cell tumor
- Tarlov cyst

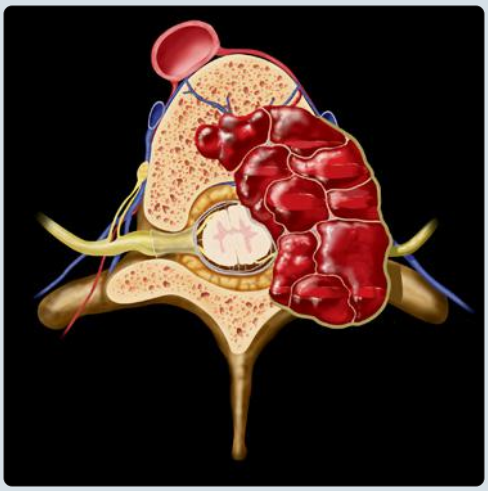
PATHOLOGY

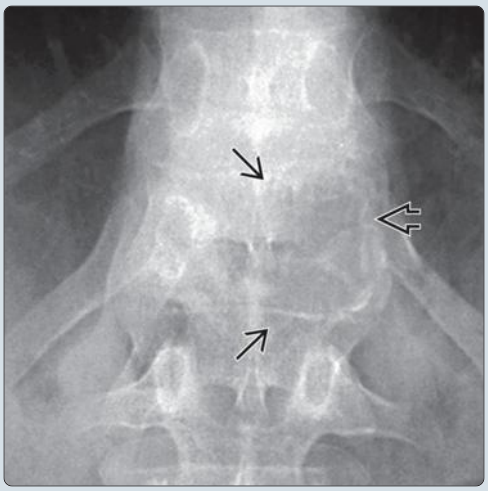
- Now considered true neoplasm since cytogenetic abnormalities found in > 50%
- Translocation between chromosomes 16 and 17

CLINICAL ISSUES

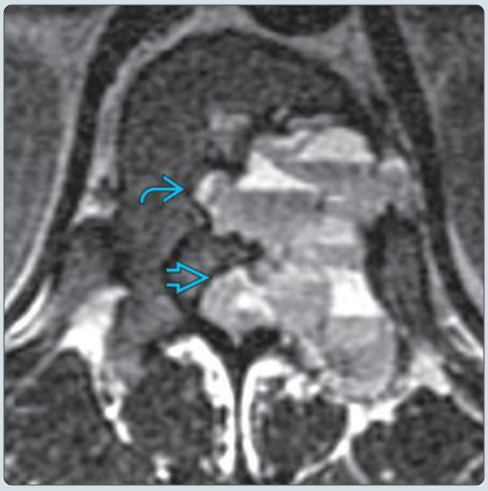
- Young patient with back pain of insidious onset

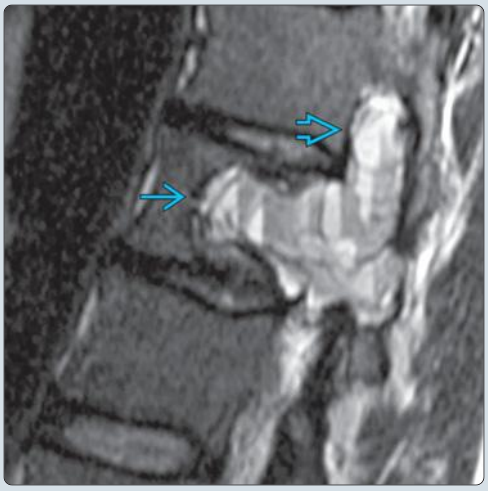
(Left) *Axial graphic shows aneurysmal bone cyst with an expansile, multicystic mass in the posterior vertebral body and pedicle extending into epidural space. Fluid-fluid levels are characteristic.* **(Right)** *Anteroposterior radiography shows aneurysmal bone cyst of T12 with an absent pedicle sign. The superior and inferior extent of tumor is shown by bone destruction, which involved the vertebral body as well as the neural arch.*





(Left) *Axial T2WI MR shows aneurysmal bone cyst of T12 with multiple fluid-fluid levels due to layering blood products. The zone of transition to normal bone is narrow and sclerotic. There is cortical breakthrough, as well as extension into the spinal canal, compressing the spinal cord.* **(Right)** *Sagittal T2WI MR in the same patient shows extension into the vertebral body and spinal canal. Determining the center of origin of a bone tumor is critical in differential diagnosis.*





TERMINOLOGY

Definitions

- Expansile benign neoplasm containing thin-walled, blood-filled cavities

IMAGING

General Features

- Best diagnostic clue
  - Multiple fluid-fluid levels in expansile mass
- Location
  - 10-30% of aneurysmal bone cysts (ABCs) occur in spine/sacrum
    - Arise in neural arch
    - 75-90% extend into vertebral body
- Morphology
  - Bone cannot truly expand
  - Expansile appearance reflects containment of tumor by appositional periosteal new bone

Radiographic Findings

- Radiography
  - Balloon-like expansile remodeling of bone
    - Centered in neural arch, extends into vertebral body
  - Cortical thinning
  - Focal cortical destruction common
  - Absent pedicle sign: Expansion of pedicle results in loss of pedicle contour on AP radiographs
  - Rare: Vertebral body collapse (vertebra plana)
  - Rare: Extends to > 1 vertebral level
  - Rare: Involves adjacent ribs

CT Findings

- Balloon-like expansile mass centered on posterior elements
  - Commonly extends into epidural space, may severely narrow spinal canal
- Contains rounded cysts that show fluid-fluid levels
  - Caused by hemorrhage, blood product sedimentation
- Calcified tumor matrix absent
  - Thin, bony septa may be present
- Narrow, nonsclerotic zone of transition with adjacent bone
- Thinned, "eggshell" cortex
- Focal cortical destruction common
- Periphery of tumor, septa between cysts enhance
- Solid ABC variant enhances diffusely

MR Findings

- Lobulated neural arch mass ± extension into vertebral body
- Well-defined rim of low signal intensity around mass
  - Reflects periosteum &/or pseudocapsule
- Peritumoral edema
  - High signal intensity on T2WI, STIR extends beyond tumor margins
  - Enhances with gadolinium (Gd)
- Cystic spaces of varying sizes within mass
  - Contain fluid-fluid levels due to blood products
  - Signal intensity variable
  - Cysts separated by septa of varying thickness
  - Gd enhancement at periphery, septa between cysts
- Part or all of mass may be solid (solid ABC)
  - Diffuse Gd enhancement of solid portions

Angiographic Findings

- Conventional
  - Hypervascular
  - Vessels more prominent at periphery, drape around lesion

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan
    - May have rim of activity around photopenic region (donut sign)

Imaging Recommendations

- CT best for diagnosis based on specific imaging features
- CT best to differentiate from telangiectatic osteogenic sarcoma
  - Narrow zone of transition in ABC
  - Absence of infiltration into surrounding soft tissues
- MR shows epidural extent, cord compromise

DIFFERENTIAL DIAGNOSIS

Osteoblastoma

- Same age range
- Expansile lesion of neural arch
- Bone matrix visible on plain films or CT
- May be associated with ABC

Telangiectatic Osteogenic Sarcoma (OGS)

- Same age range or older
- Involves vertebral body &/or neural arch
- Also shows fluid-fluid levels
- Has more permeative bone destruction
- Wider zone of transition
- Infiltrates into surrounding soft tissues

Metastases

- Older patients
- Involves vertebral body ± neural arch
- Destructive lesion with associated soft tissue mass
- Rare: Vascular metastasis can have fluid-fluid levels
- Usually destroys bone cortex rather than expanding it
- Renal cell carcinoma can have "soap bubble" expansile appearance

Giant Cell Tumor (GCT)

- Slightly older patients
- Originates in vertebral body rather than neural arch
- Expansile lytic lesion ± soft tissue mass
- May be associated with ABC

Plasmacytoma

- Older patients, usually over 40 years old
- Involves vertebral body, usually spares neural arch
- May expand vertebral body

Tarlov Cyst

- Perineural cyst occurring in sacrum
- Arises in neural foramina or spinal canal
- Causes bone remodeling
- No enhancement

- Simple fluid on all pulse sequences

Simple Bone Cyst (SBC)

- In peripheral bones, SBC is another cystic lesion
- May have fluid-fluid levels
- Not seen in spine

PATHOLOGY

General Features

- Etiology
  - Now considered true neoplasm since cytogenetic abnormalities found in > 50%
    - Previously thought to result from trauma or local circulatory disturbance
  - Divided into primary and secondary types
    - Primary ABC is isolated neoplasm of bone
    - Secondary ABC is associated with another tumor
      - Most commonly giant cell tumor or osteoblastoma

Gross Pathologic & Surgical Features

- Spongy, red mass
- Multiple blood-filled spaces

Microscopic Features

- Typical
  - Cystic component predominates
    - Cavernous blood-filled cysts of variable sizes
    - Lined by fibroblasts, giant cells, histiocytes, hemosiderin
  - Solid components
    - Septations interposed between blood-filled spaces
    - Contain bland stroma with fibrous tissue, reactive bone, giant cells
- Solid ABC is rare variant
  - 5-8% of all ABCs
  - Solid component predominates
  - Propensity for spine

Genetics

- Translocation t(16;17)(q22;p13)
- Results in fusion of promoter region of osteoblast cadherin 11 gene (CDH11) on chromosome 16q22 to the ubiquitin protease USP6 gene on chromosome 17p13
  - Promoter swapping mechanism
  - Up-regulates transcription of USP6
  - Immature cells of osteoblast lineage likely involved in overexpression

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, most severe at night
  - Other signs/symptoms
    - Scoliosis
    - Neurologic signs and symptoms from root &/or cord compression
    - Pathologic fracture
- Clinical profile
  - Young patient with back pain of insidious onset

Demographics

- Age
  - 80% under 20 years old
- Gender
  - Slightly more common in females
- Epidemiology
  - 1-2% of primary bone tumors are ABC
    - Familial incidence has been reported
  - 0.15 per million individuals in general population

Natural History & Prognosis

- Long-term history of untreated ABC variable
  - Grows initially, then usually stabilizes
  - No malignant degeneration
- Recurrence rate 20-30% (increased if incomplete excision)

Treatment

- Embolization
  - Curative as sole therapy in some cases
  - Can also be used preoperatively
- Surgical excision or curettage
- May require instrumentation for stabilization of spine
- Radiation therapy may predispose to radiation-induced sarcoma

DIAGNOSTIC CHECKLIST

Consider

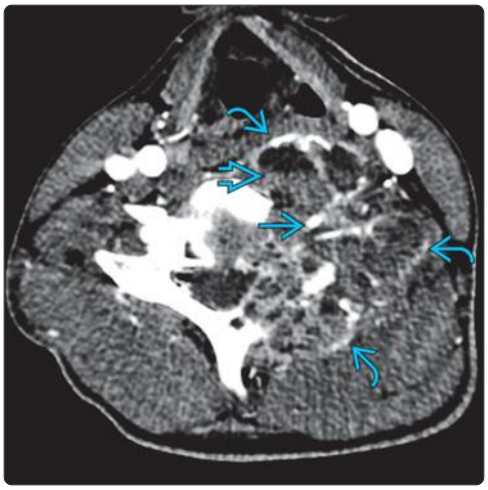
- Absent pedicle sign caused by ABC, osteoblastoma, lytic osteogenic sarcoma, metastasis, trauma, congenital absence of pedicle

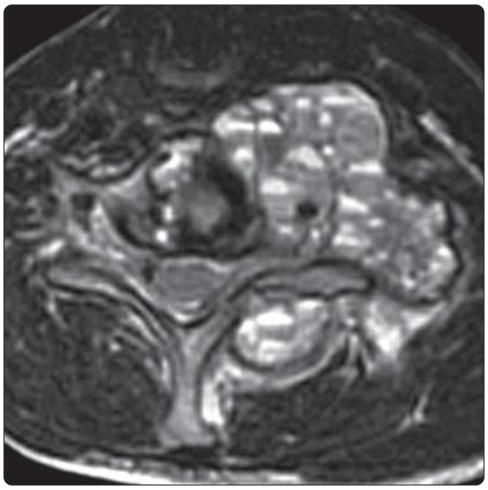
Image Interpretation Pearls

- Evaluation of pedicles on AP radiograph should be part of routine on every patient

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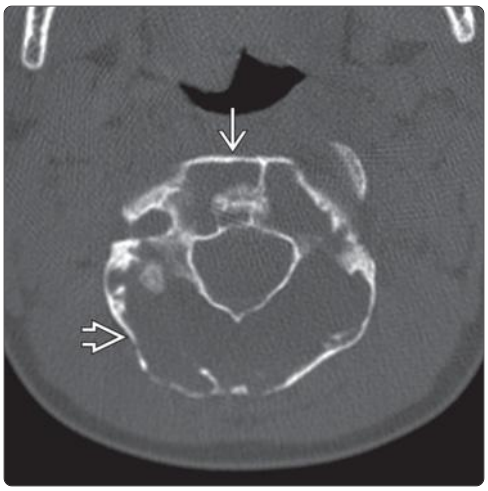


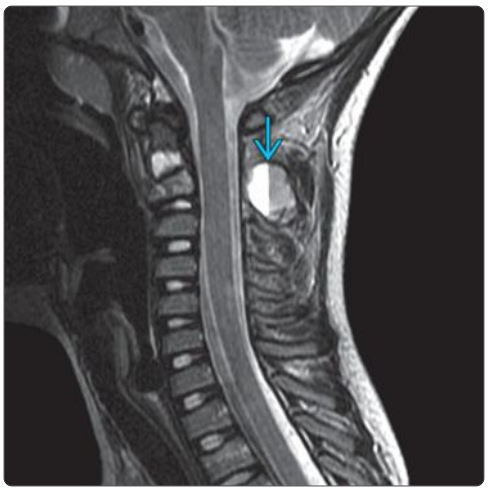
(Left) *Axial CECT shows a markedly expansile mass arising from the C5 articular pillar and containing multiple fluid-fluid levels. Contrast enhancement around the periphery of the lesion is difficult to distinguish from reactive bone. Note the displaced but patent vertebral artery.* **(Right)** *Axial T2WI MR in the same patient shows the classic appearance of multiple fluid-fluid levels in small cysts, which are separated by thin septa.*





(Left) *Sagittal T1WI MR shows aneurysmal bone cyst filling neural foramen. Fluid-fluid levels are present but less striking than on T2WI.* **(Right)** *Sagittal T2WI MR in the same patient shows well-defined fluid-fluid levels. The mass is clearly centered posterior to the vertebral body, a useful diagnostic clue. A small amount of peritumoral edema is present. Peritumoral edema is a common finding in benign tumors and should not be mistaken for a sign of malignancy.*





(Left) *Axial NECT (bone window) shows an extensive aneurysmal bone cyst involving the entire C2 body with both body and posterior element expansion. The bony expansion is the typical "soap bubble" type with intact cortical margins.* **(Right)** *Sagittal T2 MR shows a heterogeneous signal mass involving the C2 body and posterior elements without epidural extension and no cord compression. The mass characteristically shows fluid-fluid levels.*

Giant Cell Tumor

KEY FACTS

TERMINOLOGY

- Locally aggressive neoplasm composed of osteoclast-like giant cells

IMAGING

- 3% of all GCT occur in spine
- 4% of all GCT occur in sacrum
- Lytic, expansile lesion of vertebral body or sacrum
- Narrow zone of transition, margin not sclerotic
- Matrix absent, but may have residual bone trabeculae
- May have cortical breakthrough
- Heterogeneous contrast enhancement
- Nonenhancing areas of necrosis often present
- May be associated with aneurysmal bone cyst component

TOP DIFFERENTIAL DIAGNOSES

- Metastasis
- Myeloma
- Chordoma
- Osteogenic sarcoma

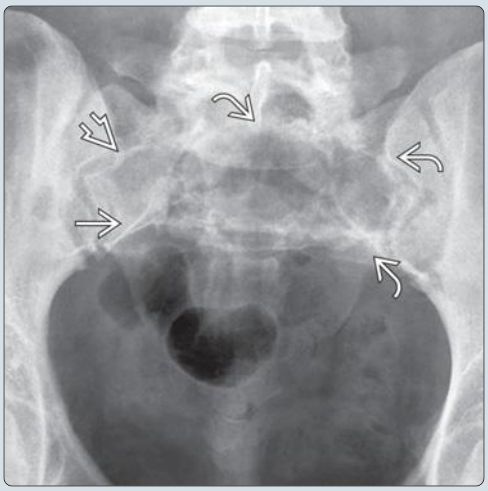
CLINICAL ISSUES

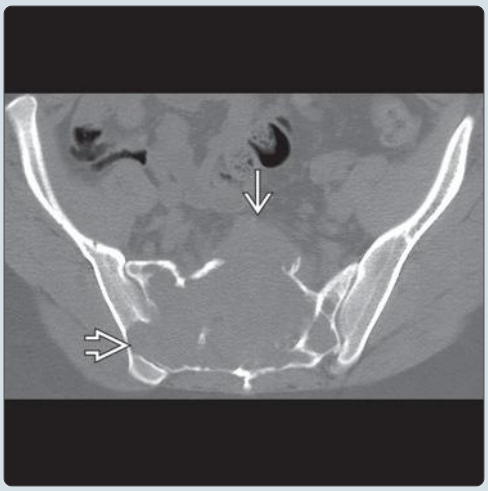
- Back pain of insidious onset, greatest at night
- Locally aggressive: 12-50% recurrence rate
- Curettage alone associated with high rate of recurrence
  - Curettage defect filled with methylmethacrylate or bone graft
  - Denosumab: Human monoclonal antibody to RANKL
    - In one study, 86% of patients had tumor response

DIAGNOSTIC CHECKLIST

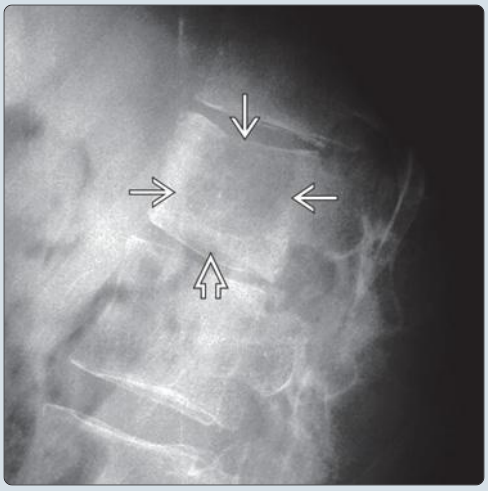
- Most likely causes of solitary sacral mass in adult patient are GCT, chordoma, and plasmacytoma
- Malignant GCT rare, difficult to distinguish from typical GCT
  - Zone of transition usually less well defined

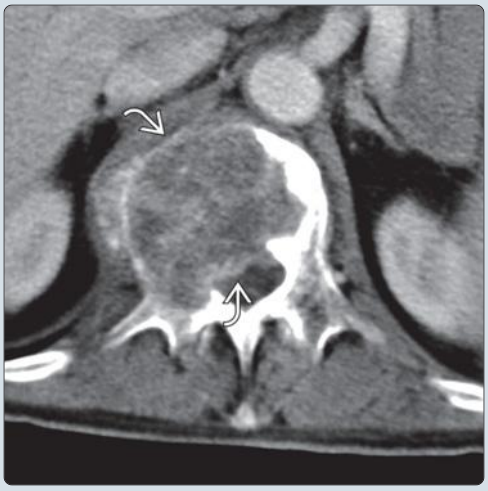
(Left) *AP radiograph shows a giant cell tumor ⇨ of the sacrum. Lytic lesions in this region are difficult to see on radiographs and may be mistaken for bowel gas. Arcuate lines are a useful landmark. In this case, the S1 ⇨ and S2 ⇨ arcuate lines are visible on the normal right side, but destroyed on the left.* **(Right)** *Axial NECT shows nearly complete cortical rim around a large sacral mass. There is a small region of cortical breakthrough ⇨ and extension of the tumor from sacrum into the right iliac wing, across the SI joint ⇨.*





(Left) *Lateral radiograph shows a lytic lesion ⇨ of L1. The lesion is sharply demarcated with a nonsclerotic margin. There is probable cortical breakthrough inferiorly ⇨. This constellation of findings is consistent with GCT and other intermediate aggressivity lesions, such as plasmacytoma, chordoma (rare at this site), or some metastases.* **(Right)** *Axial CECT in the same patient shows a heterogeneously enhancing mass with cortical breakthrough ⇨.*





TERMINOLOGY

Abbreviations

- Giant cell tumor (GCT)

Definitions

- Locally aggressive neoplasm composed of osteoclast-like giant cells

IMAGING

General Features

- Best diagnostic clue
  - Lytic lesion in vertebral body or sacrum
- Location
  - 3% of all GCT occur in spine
    - Centered in vertebral body
  - 4% of all GCT occur in sacrum
  - Rarely involves multiple sites

Radiographic Findings

- Radiography
  - Lytic, expansile lesion of vertebral body or sacrum
  - Narrow zone of transition
  - Margin usually not sclerotic
  - Matrix absent, but may have residual bone trabeculae
  - May have cortical breakthrough

CT Findings

- Features described for radiography are more clearly seen
- May contain fluid attenuation regions due to necrosis
- Contrast enhancement may mimic bone matrix
- Fluid-fluid levels suggest associated ABC

MR Findings

- Expansile mass
- Soft tissue extension common
- Sharply demarcated, narrow zone of transition
- Low to intermediate signal intensity on T1WI
- Intermediate to high signal intensity on T2WI, STIR
- Fluid-fluid levels suggest associated ABC
- Thin, curvilinear bands of low signal intensity on all sequences
  - Correspond to residual bone trabeculae or fibrous septa
- ± low signal intensity hemosiderin
- Heterogeneous contrast enhancement
- Nonenhancing areas of necrosis often present

Nuclear Medicine Findings

- Bone scan
  - Positive on all 3 phases

Imaging Recommendations

- Best imaging tool
  - CT scan for diagnosis
  - MR for evaluation of spinal canal, nerve roots
- Protocol advice
  - Thin-section CT with sagittal, coronal reformations

DIFFERENTIAL DIAGNOSIS

Metastasis

- Usually appears more aggressive, but can be expansile
- Involves vertebral body &/or neural arch
- Often multiple
- Usually in older patients

Myeloma

- Lytic lesion, often expansile
- Vertebral body or sacrum
- Can have sharp zone of transition, geographic appearance
- Usually in older patients
- Look for lesions elsewhere

Chordoma

- Lytic, expansile lesion
- Most common in sacrum but can involve vertebral body
- Arises in midline
- No matrix
- Can have large soft tissue component

Osteogenic Sarcoma (OGS)

- Wide zone of transition, permeative appearance
- Vertebral body or neural arch
- Soft tissue mass
- Osseous matrix visible in 80%
- May contain large numbers of giant cells

Aneurysmal Bone Cyst (ABC)

- Markedly expansile
- Arises in neural arch, can extend into vertebral body
- Can coexist with GCT

Osteoblastoma

- Lytic, expansile lesion neural arch
- May extend into vertebral body
- Bone matrix visible on CT scan

Brown Tumor of Hyperparathyroidism

- Radiologically and histologically identical to GCT
- Due to osteoclast stimulation in hyperparathyroidism
- Regresses with successful treatment of hyperparathyroidism

PATHOLOGY

Staging, Grading, & Classification

- Typical GCT
  - Grading systems have been proposed, but not predictive of behavior
- Malignant GCT
  - High risk of local recurrence and metastasis to bone, lung, and liver

Gross Pathologic & Surgical Features

- Soft, tan tumor
- 10-15% have ABC component
- Surrounding bone is "expanded" with thinning of cortex
  - Actually represents containment of tumor by deposition of periosteal new bone
- Periphery shows thin layer of fibrous and reactive bone tissue
- Apparent soft tissue extension usually covered by periosteum

Microscopic Features

- Multinucleated osteoclastic giant cells
  - Giant cells have average of 15 nuclei
  - Mitoses are absent
  - Nuclei round or oval, not pleomorphic
  - Abundant cytoplasm
  - No bone or cartilage matrix production by giant cells
- Spindle cell stroma
  - Each cell has single nucleus similar in appearance to giant cells
- Reactive osteoid may be present
  - Periosteal reaction, not made by tumor cells
- Hemorrhage, necrosis, hemosiderin may be present
- Malignant giant cell tumor
  - Rare tumor, difficult histologic diagnosis
  - Anaplasia and pleomorphism of giant cells
  - Distinguished from giant cell-rich OGS by lack of osteoid formation by tumor cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain of insidious onset, greatest at night
- Clinical profile
  - Pathologic fracture in 30%
  - Limited range of motion of joint adjacent to affected area

Demographics

- Age
  - 80% in 3rd to 5th decade of life
  - In spine, peak incidence in 2nd and 3rd decades
  - Rare before skeletal maturity
- Gender
  - F:M = 2.5:1 in spine
    - More marked female preponderance than in appendicular skeleton
- Epidemiology
  - 5% of all primary bone tumors
  - 6th most common primary bone tumor

Natural History & Prognosis

- Locally aggressive: 12-50% recurrence rate
- Can undergo sarcomatous transformation
  - Either spontaneously or in response to radiation therapy
- Pulmonary implants: 1-2%
  - Self-limited growth potential
  - May regress spontaneously
  - Resection is curative
  - Occur within 3 years after removal of primary GCT
  - Histologically identical to primary GCT
- Primary malignant GCT (rare)
  - Poor prognosis
  - Metastasis to lung, liver, and bone

Treatment

- Curettage alone associated with high rate of recurrence
  - Cryotherapy or thermocoagulation used to reduce recurrence
  - Curettage defect filled with methylmethacrylate or bone graft
- Chemotherapy used as adjunct if tumor cannot be completely resected
- Morbidity of complete resection of sacral tumor is high
- Denosumab: Human monoclonal antibody to RANKL
  - In one study, 86% of patients had tumor response
    - Histologic elimination of 90% of giant cells or no radiologic progression of the target lesion to week 25
- Radiation only for cases of unresectable tumor
  - Risk of sarcoma years after treatment
- Arterial embolization
  - Used preoperatively
  - Used for therapy for unresectable tumor
- Surgical resection of pulmonary implants
  - Good prognosis following resection
- Malignant GCT treated with wide resection, chemotherapy, radiation

DIAGNOSTIC CHECKLIST

Consider

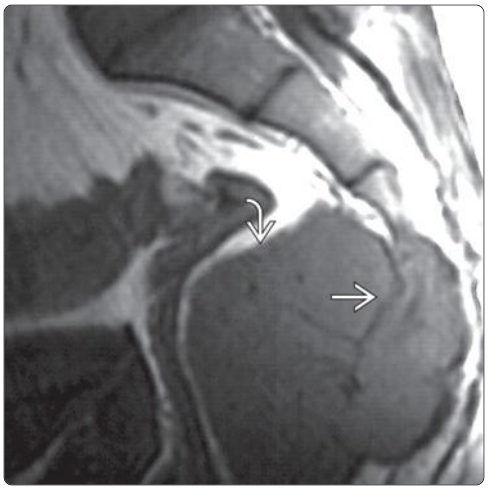
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- Malignant GCT are rare, difficult to distinguish from typical GCT
  - Zone of transition usually less well defined

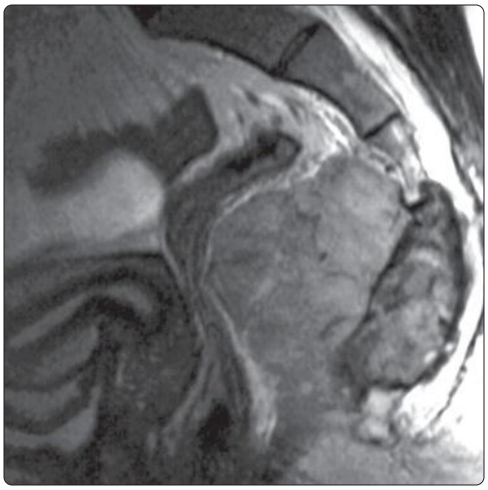
Image Interpretation Pearls

- Sacral GCT may be occult on radiographs; have low threshold for CT in patients with atypical sacral pain
- Patients treated with radiation must be followed with MR because of risk of recurrence, sarcomatous degeneration

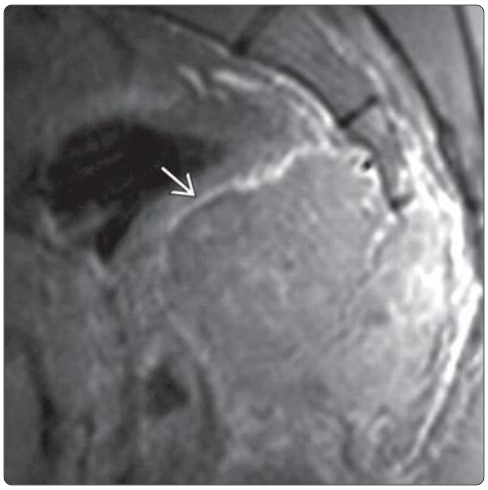
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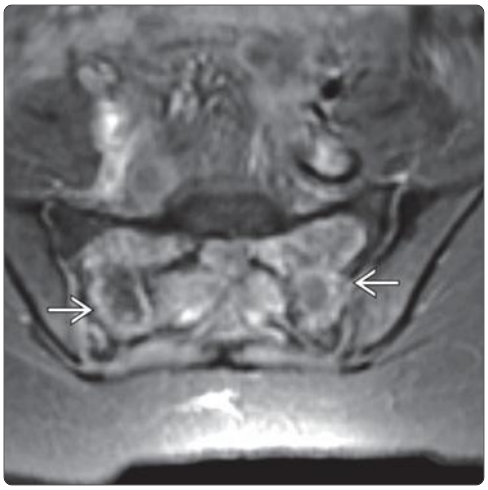
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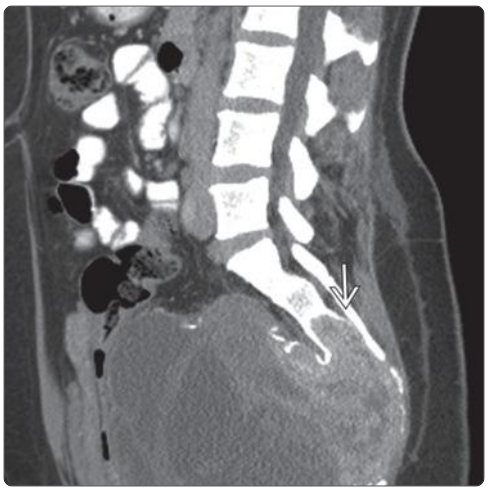


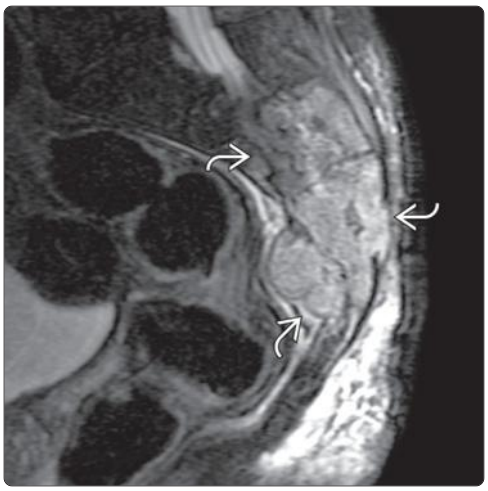
(Left) *Sagittal T1WI FS MR shows a large sacral GCT deviating the rectum anteriorly. Note thin, curvilinear bands of low signal intensity within the tumor, corresponding to fibrous or bony septa.* **(Right)** *Sagittal T2WI MR in the same patient shows that the signal intensity of the lesion is heterogeneous, but remains fairly low. The signal intensity of GCT is variable and of limited utility in differential diagnosis.*





(Left) *Sagittal T1WI C+ FS MR in the same patient shows the large solid and heterogenously enhancing tumor arising from the sacrum. GCT is often markedly heterogeneous in enhancement.* **(Right)** *Axial T1WI C+ FS MR shows a treated GCT, with rim enhancement around fluid-filled regions. Because of morbidity associated with complete surgical resection, sacral tumors may be treated with chemotherapy, and serial MR is used to evaluate for recurrence.*





(Left) *Sagittal NECT shows a massive pelvic mass with involvement of the inferior sacrum. The differential is primarily one of sacral chordoma vs. giant cell tumor. Metastasis is unlikely since it appears to be an indolent lesion.* **(Right)** *Sagittal STIR MR shows malignant sacral GCT. Clues suggesting the tumor is more aggressive than most GCT include a larger ST mass and more extensive cortical destruction.*

Osteochondroma

KEY FACTS

TERMINOLOGY

- Osteochondroma (OC)
- Osteocartilaginous exostosis, exostosis
- Cartilage-capped osseous growth contiguous with parent bone

IMAGING

- Sessile or pedunculated osseous "cauliflower" lesion
- Continuity of bony cortex and medullary space between lesion and and underlying bone
- May see chondroid calcifications in cartilage cap
- Center follows normal marrow signal on MR
- Cartilage cap of similar intensity to cord on T1WI, hyperintense on T2WI

TOP DIFFERENTIAL DIAGNOSES

- Chondrosarcoma
- Osteoblastoma
- Aneurysmal bone cyst (ABC)
- Tumoral calcinosis
- Enthesopathy

PATHOLOGY

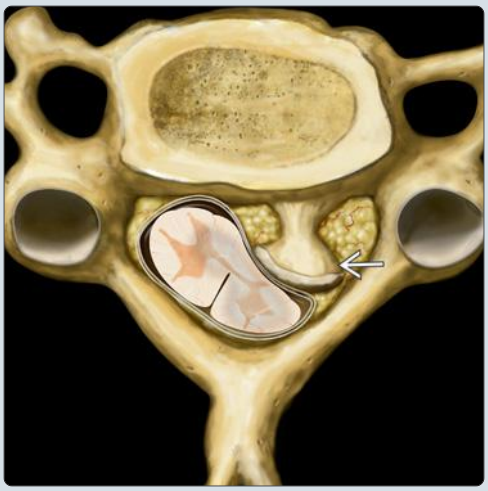
- Idiopathic, trauma, perichondrial ring deficiency
- Radiation-induced OC
- Syndromic: Hereditary multiple exostoses
- Vertebral OC rare; 1-5% of sporadic OC, 1–9% OC in HME

CLINICAL ISSUES

- Often asymptomatic; incidental diagnosis on radiography
- Palpable mass
- Mechanical impingement of joint, muscle
- Cord compression, radiculopathy unusual
- Peak age = 10-30 years

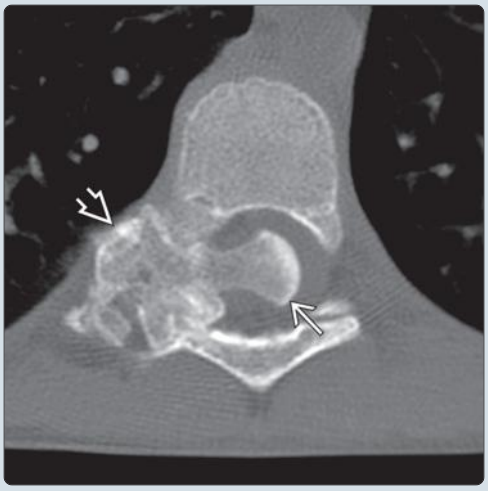
DIAGNOSTIC CHECKLIST

- Multiplicity → consider HME
- Cartilage cap > 1.5 cm in adults raises concern for malignant transformation (chondrosarcoma)





(Left) *Axial graphic of the cervical spine demonstrates a typical osteochondroma (exostosis) protruding into the spinal canal, causing canal stenosis and cord compression.* **(Right)** *Axial NECT shows a large osseous excrescence projecting ventrally from the right lateral mass of C2. There is ossification of the cartilaginous cap.*





(Left) *Axial bone CT shows pedunculated osseous lesion extending into canal from the right pedicle, with cortical and medullary continuity. There is resulting canal stenosis. The right pedicle and superior facet are irregularly enlarged due to sessile component of the osteochondroma.* **(Right)** *Axial T2WI FS MR in the same patient shows the medullary continuity of the osseous mass with the right pedicle and a hyperintense cartilaginous cap. Spinal cord is displaced and compressed.*

TERMINOLOGY

Abbreviations

- Osteochondroma (OC)
- Hereditary multiple exostoses (HME)

Synonyms

- Osteocartilaginous exostosis, exostosis
- HME, a.k.a. diaphyseal aclasis, multiple cartilaginous exostoses

Definitions

- Cartilage-capped osseous excrescence contiguous with parent bone

IMAGING

General Features

- Best diagnostic clue
  - Sessile or pedunculated osseous "cauliflower" lesion with corticomedullary continuity with parent bone
- Location
  - Bones forming through endochondral ossification
    - Metaphysis of long tubular bones (85%) common, particularly knee
  - < 5% occur in spine
    - Cervical (50%, C2 predilection) > thoracic (T8 > T4 > other levels) > lumbar > > sacrum
    - Spinous/transverse processes > vertebral body
- Size
  - Varies dramatically at time of presentation; 1-10 cm

Radiographic Findings

- Radiography
  - Sessile/pedunculated osseous protuberance with flaring of parent bone cortex at OC attachment
  - Cartilage cap visible only if extensively mineralized

CT Findings

- NECT
  - Sessile or pedunculated osseous lesion
  - Cortex and medullary cavity continuous with attached bone
  - May see chondroid calcifications in cartilage cap
- CECT
  - May see peripheral, septal enhancement in cartilage cap

MR Findings

- T1WI
  - Central hyperintensity (marrow) surrounded by hypointense cortex
  - Hypo-/isointense hyaline cartilage cap
- T2WI
  - Central iso- to hyperintense signal (marrow) surrounded by hypointense cortex
  - Hyperintense hyaline cartilage cap
- T1WI C+
  - May see peripheral, septal enhancement in cartilage cap

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic nonmineralized cartilage cap easily distinguished from adjacent fat, muscle
  - Cartilage cap mineralization and osseous stalk → posterior acoustic shadowing

Nuclear Medicine Findings

- Bone scan
  - Variable; direct correlation with enchondral bone formation
    - ↑ radionuclide uptake = metabolically active osteochondroma
    - No ↑ radionuclide uptake = quiescent osteochondroma

Imaging Recommendations

- Best imaging tool
  - MR imaging
- Protocol advice
  - MR to measure cartilage cap, determine status of regional neural and musculoskeletal tissue
  - Bone CT to assess mineralization, confirm continuity with vertebral marrow space

DIFFERENTIAL DIAGNOSIS

Chondrosarcoma

- Isolated or 2° to malignant OC degeneration
- Lytic destructive lesion with sclerotic margins, soft tissue mass
- Chondroid matrix ("rings and arcs," 50%)

Osteoblastoma

- Expansile lesion, typically involving posterior elements
- Lytic lesion with osseous matrix, narrow zone of transition

Aneurysmal Bone Cyst (ABC)

- Expansile, lytic lesion
- Multicystic, fluid-fluid levels

Tumoral Calcinosis

- Calcific periarticular mass
- Spinal involvement uncommon

Enthesopathy

- DISH, ankylosing spondylitis, psoriatic arthropathy, disorders of calcium metabolism

PATHOLOGY

General Features

- Etiology
  - Idiopathic, trauma, perichondrial ring deficiency
    - Peripheral portion of epiphyseal cartilage herniates out of growth plate
    - Metaplastic cartilage is stimulated; enchondral bone formation → bony stalk
  - Radiation-induced OC
    - Most common benign radiation-induced tumor
      - Prevalence 6-24%
    - Radiation dose 1,500–5,500 cGy; occur at treatment field periphery
      - Patients generally < 2 years old at time of XRT
      - Latent period 3-17 years
    - Pathologically and radiographically identical to other exostoses

- Genetics
  - Hereditary multiple exostoses
    - Autosomal dominant transmission
    - Exostoses (multiple) 1 gene (a.k.a. *EXT1*) on long arm of chromosome 8; produces exostosin 1
    - Exostoses (multiple) 2gene (a.k.a. *EXT2*) on short arm of chromosome 11; produces exostosin 2
    - Exostoses (multiple) 3 gene (a.k.a. *EXT3*) on short arm of chromosome 19
    - Inactivation of one EXT gene → exostosis; subsequent inactivation of 2nd EXT gene → malignant transformation
- Benign cartilaginous bone tumor
  - 9% of all bone tumors, most common benign bone tumor (30-45%)
  - Rapid growth, new pain, and continued growth of cartilage cap > 1.5 cm thickness after skeletal maturity imply malignant transformation to chondrosarcoma
  - OC complications include deformity, fracture, vascular compromise, neurologic sequelae, overlying bursa formation, and malignant transformation
- Vertebral OC rare; 1-5% of sporadic OC, 1–9% OC in HME
  - Narrow stalk (pedunculated) or broad attachment base (sessile) contiguous with vertebral cortex, medullary space
  - Hyaline cartilage cap thickness proportional to patient age
- Many spinal OC are asymptomatic
  - May cause mechanical impingement
  - May compress spinal cord or nerve root

Gross Pathologic & Surgical Features

- Osseous excrescence with corticomedullary contiguity with parent bone
- Cartilage cap ranges from thick bosselated glistening blue-gray surface (young patients) to several millimeters thick or entirely absent (adults)

Microscopic Features

- Mature cartilaginous, cancellous, and cortical bone
- Cartilage cap histology reflects classic growth plate zones

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often asymptomatic; incidental diagnosis on radiography
  - Palpable mass
  - Mechanical impingement of joint, muscle
- Other signs/symptoms
  - Myelopathy or radiculopathy, more common with HME
  - Canal stenosis may predispose to cord injury with mild trauma
  - Cranial nerve deficits
  - Pharyngeal mass
  - Scoliosis

Demographics

- Age
  - Peak age = 10-30 years
  - Most HME patients diagnosed by age 5, virtually all by 12 years
- Gender
  - M:F = 3:1
- Epidemiology
  - Prevalence of solitary OC unknown since many asymptomatic
  - Prevalence of HME 1:50,000-100,000 in Western populations; up to 1:1,000 in ethnic Chamorros

Natural History & Prognosis

- Postsurgical local recurrence rate < 2%
- Benign lesions; no propensity for metastasis
- Malignant transformation < 1% solitary lesions; 3-5% HME
  - Growth or new pain after skeletal maturity, ↑ cartilage cap thickness (> 1.5 cm in adults)

Treatment

- Conservative management in asymptomatic patients
- Surgical excision, deformity correction in symptomatic patients

DIAGNOSTIC CHECKLIST

Consider

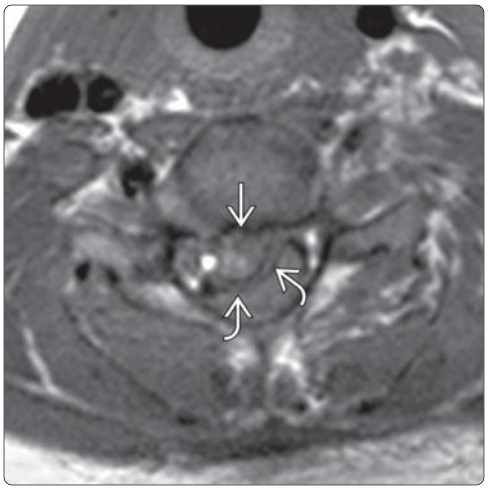
- Multiplicity → consider HME
- Interpret cartilage thickness in context of patient age

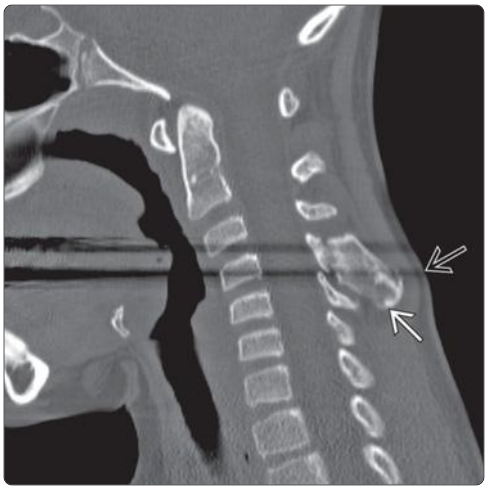
Image Interpretation Pearls

- Cartilage cap > 1.5 cm in adults raises concern for malignant transformation (chondrosarcoma)
- Radiologic features pathognomonic, reflect pathologic appearance

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(Left) *Axial T1WI MR shows the marrow space of a cervical osteochondroma within the cervical canal, with the predominately hypointense cartilaginous cap causing stenosis of the cervical canal and compressing the spinal cord.* **(Right)** *Sagittal reformatted NECT shows pedunculated bony mass arising from the spinous process of C4 in this young child. The mass causes a small bulge of the overlying skin.*





(Left) *Sagittal T1WI MR shows cauliflower-like, heterogeneous signal mass arising from the left L5-S1 facet joint. T1 hyperintensity reflects marrow elements.* **(Right)** *Coronal CT reconstruction shows an osteochondroma arising from the left posterior elements of a thoracic vertebral body. There is corticomedullary continuity with the underlying bone. The adjacent ribs are splayed. (Courtesy J. Lichtenberger, III, MD.)*





(Left) *Axial T1WI MR shows a large osteochondroma from the L4 spinous process, with hyperintense marrow centrally and peripheral isointense cartilage cap. Osteochondroma with more classical cauliflower morphology noted at the right iliac crest.* **(Right)** *Axial NECT shows a tiny bony excrescence arising from the left pedicle demonstrating corticomedullary continuity with the underlying bone in this patient with MHE.*

Chondrosarcoma

KEY FACTS

TERMINOLOGY

- Malignant tumor of connective tissue, characterized by formation of cartilage matrix by tumor cells

IMAGING

- Lytic mass with chondroid matrix
  - Higher grade lesions tend to have larger areas without calcifications
- Lobular morphology often seen
- Enhancing periphery and internal septa
- May penetrate cortex with epidural or paravertebral extension
- Thoracic most commonly involved spine segment
  - Both posterior elements and body (45%)
  - Posterior elements (40%)
- Variable size, typically large at presentation

TOP DIFFERENTIAL DIAGNOSES

- Metastases
- Plasmacytoma
- Lymphoma
- Osteosarcoma
- Malignant fibrous histiocytoma

PATHOLOGY

- Lobulated tumor composed of translucent hyaline nodules (resemble normal cartilage)
- May be primary or secondary

CLINICAL ISSUES

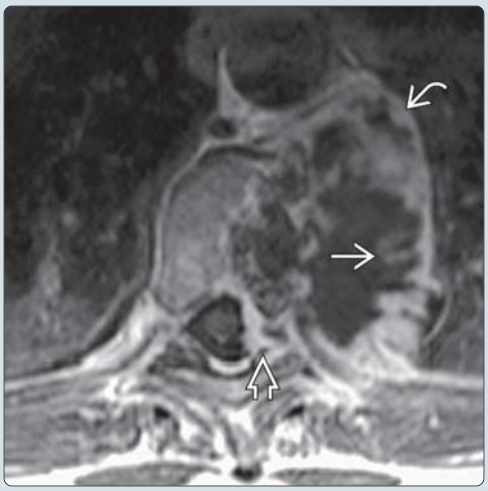
- Palpable mass
- Localized pain, typically long duration of symptoms
- Neurologic symptoms in 45% of vertebral chondrosarcoma
  - Weakness, paresthesias, paralysis
- Age at presentation: 20-90 years; peak: 40-60 years

DIAGNOSTIC CHECKLIST

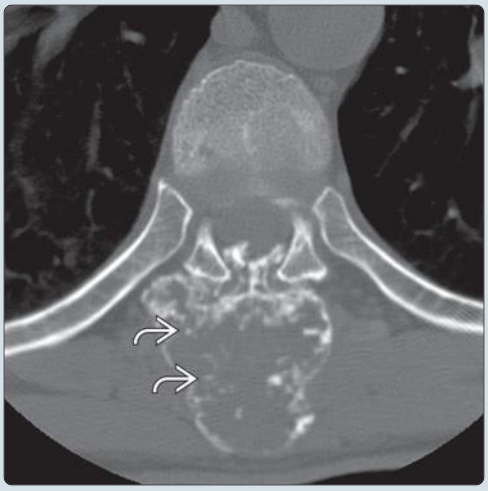
- Enlarging or painful enchondroma or osteochondroma suspicious for sarcomatous transformation

(Left) *Sagittal graphic of the lumbosacral junction shows a large soft tissue mass centered within the sacrum. This tumor produces bone destruction with presacral and epidural extension.* **(Right)** *Axial T1WI C+ MR shows a large vertebral mass with paraspinal and epidural extension. There is irregular peripheral enhancement with some faintly enhancing central septations.*





(Left) *Axial unenhanced bone CT shows a large mass arising from the neural arch, containing ringed and stippled calcifications, a typical appearance chondroid tumor matrix.* **(Right)** *Left paravertebral sagittal T2WI in the same patient shows a large, lobulated mass, and predominately hyperintense signal with thin, hypointense signal around the periphery of the lobules due to calcification. This is the typical MR appearance of a chondroid neoplasm.*





TERMINOLOGY

Abbreviations

- Chondrosarcoma (CHS)

Definitions

- Malignant tumor of connective tissue, characterized by formation of cartilage matrix by tumor cells

IMAGING

General Features

- Best diagnostic clue
  - Lytic mass with chondroid matrix
- Location
  - Most commonly involved segment of spine is thoracic
  - Posterior elements (40%)
  - Vertebral body (15%)
  - Both posterior elements and body (45%)
  - Rarely seen in sacrum
- Size
  - Variable, typically large at presentation
- Morphology
  - Lytic lesion with or without chondroid matrix ("ring and arc" calcifications)
  - Lobular morphology often seen
  - May penetrate cortex with epidural or paravertebral extension

Radiographic Findings

- Radiography
  - Destructive mass
  - Chondroid matrix may be seen

CT Findings

- NECT
  - Lytic bone lesion
  - Chondroid matrix mineralization
  - Nonmineralized portions of tumor hypodense to muscle
  - Variable degree of extraosseous soft tissue mass

MR Findings

- T1WI
  - Destructive bony lesion
  - Low to intermediate signal intensity
  - Variable degree of extraosseous soft tissue mass
- T2WI
  - Lobular high signal due to high water content of hyaline cartilage
  - Areas of low signal intensity due to calcification
- T1WI C+
  - Enhancing periphery and internal septa
  - Nonenhancing areas represent hyaline cartilage, cystic mucoid tissue, necrosis

Nuclear Medicine Findings

- Bone scan
  - Increased uptake of radiotracer
  - Overestimates extent of lesion due to surrounding hyperemia and edema

Imaging Recommendations

- Best imaging tool
  - CT shows bone destruction, calcification, extent
- Protocol advice
  - CT to evaluate for chondroid matrix, cortical destruction, intra- and extraosseous extension
  - MR to evaluate extent of tumor within bone and degree of extension into adjacent soft tissues

DIFFERENTIAL DIAGNOSIS

Metastases

- Osteolytic tumor, irregular margins
- Typically multiple
- Paravertebral, epidural extension
- Compression fractures
- Some primaries may produce osteoblastic metastases

Plasmacytoma

- Osteolytic tumor, irregular margins
- Typically solitary
- Paravertebral, epidural extension
- Compression fracture

Lymphoma

- Soft tissue (e.g., epidural disease) predominates spine involvement
- Destructive, permeative tumor, when osseous structures involved
- Does not calcify

Osteosarcoma

- Osteoid matrix, not chondroid
- Usually younger patients

Malignant Fibrous Histiocytoma (MFH)

- Often purely lytic
- Can have areas of sclerosis, no chondroid matrix
- No significant periosteal reaction

PATHOLOGY

General Features

- Etiology
  - Primary CHS
    - Can arise as sequel of radiation therapy
  - Secondary CHS, malignant degeneration of osteochondroma or enchondroma
- Genetics
  - Grade 1 CHS: Diploid
  - Grade 2 and 3 CHS: Aneuploid, correlate with aggressive behavior
  - High-grade lesions: Complex chromosomal aberration with nonreciprocal translocations and deletions
    - Accumulation of p53 protein, may be indicator of poor prognosis
- Malignant tumor in which neoplastic cells form hyaline cartilage

Staging, Grading, & Classification

- Grade 1 CHS: Slowly growing, locally aggressive, indolent course
  - Recurrent growth potential, no metastatic spread

- Diagnosis requires supportive evidence from clinical and radiographic data
- Grade 2 CHS: Locally aggressive tumor with great potential for local recurrence
  - Less chondroid matrix
  - Metastases in 10-15%
- Grade 3 CHS: Highly aggressive, rapidly growing
  - Metastases > 50%
  - Sparse or absent chondroid matrix
  - Increased myxoid material

Gross Pathologic & Surgical Features

- Lobulated tumor composed of translucent hyaline nodules (resemble normal cartilage)
  - Mineralization at periphery of nodules
- Focal ivory-like areas represent extensive enchondral calcification
- Hemorrhagic necrosis, especially in high-grade tumors
- Central lesions erode and destroy cortex with extension into surrounding soft tissue
  - Cortical disruption earlier in flat bones due to narrow medullary cavity
- Extraosseous extension with soft tissue mass adjacent to cortical penetration

Microscopic Features

- Irregular-shaped lobules of cartilage
- May be separated by narrow, fibrous bands
- Chondrocytes arranged in clusters, can be mononuclear or multinucleated
- Matrix: Mature hyaline cartilage or myxoid stroma
- Immunohistochemistry: Positive for S100 protein and vimentin

Histologic Subtypes

- Conventional intramedullary (80-90%)
  - Typically large at diagnosis
  - Lobulated architecture, "ring and arc" calcification
  - Less chondroid matrix in higher grade lesions
- Clear cell
  - Rare, low-grade, slow-growing variant
  - Resembles chondroblastoma
- Juxtacortical
  - Arises on surface of bone
  - Cortical erosion seen, intramedullary extension rare
  - Fibrous pseudocapsule contiguous with periosteum
- Myxoid
  - Rare, intermediate-grade tumor
  - High water content due to myxoid stroma
- Mesenchymal
  - Rare, high-grade variant
  - Low-grade CHS with nests of small, round undifferentiated cells
- Dedifferentiated
  - "Collision" of low-grade CHS admixed with higher grade sarcoma (MFH, osteosarcoma, fibrosarcoma)
  - 5-year survival of < 10%

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Localized pain, typically long duration of symptoms
  - Neurologic symptoms in 45% of vertebral CHS
    - Weakness, paresthesias, paralysis
  - Palpable mass

Demographics

- Age
  - 20-90 years, peak: 40-60 years
- Gender
  - M:F = 2:1
- Epidemiology
  - 3rd most common primary malignancy of bone
  - 2nd most common primary nonlymphoproliferative malignancy of vertebral column
  - 10-25% of all primary bone sarcomas
  - Extraskeletal CHS: 2% of all soft tissue sarcomas

Natural History & Prognosis

- Prognosis depends on location, size, and histologic grade; completeness of surgical resection
  - Overall 5-year survival: 48-60%
  - 5-year survival 90% for grade 1, 81% for grade 2, 29% for grade 3 tumors
- Recurrence 5-10 years after surgery
  - Recurrence can be associated with increased histologic grade and more aggressive behavior
- Metastases (lung, lymph nodes, liver, kidneys, brain): 66% (grade 3 CHS)
- Degeneration into fibrosarcoma, malignant fibrous histiocytoma, osteosarcoma in 10%
- Pelvic CHS may invade bladder or colon
- Pathologic fractures rare

Treatment

- En bloc surgical excision
  - Radical excision impossible in spine
- Evidence for mild decrease in recurrence with adjuvant radiotherapy

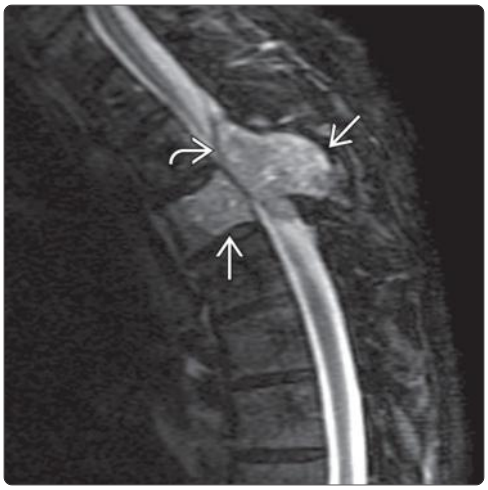
DIAGNOSTIC CHECKLIST

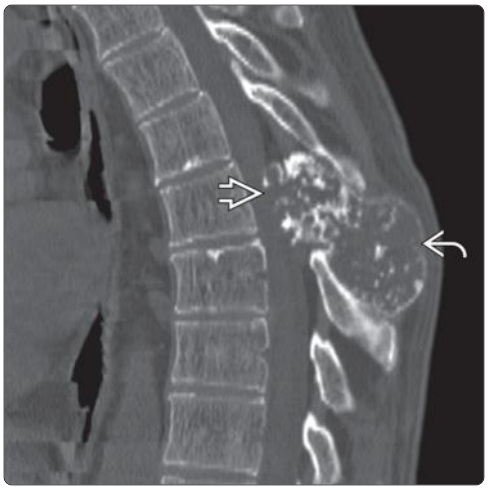
Consider

- Enlarging or painful enchondroma or osteochondroma suspicious for sarcomatous transformation
- Clear cell CHS may be indistinguishable from benign chondroid lesions (chondroblastoma, enchondroma)
- Substantial risk of tumor seeding and ↑ recurrence with biopsy or with piecemeal resection

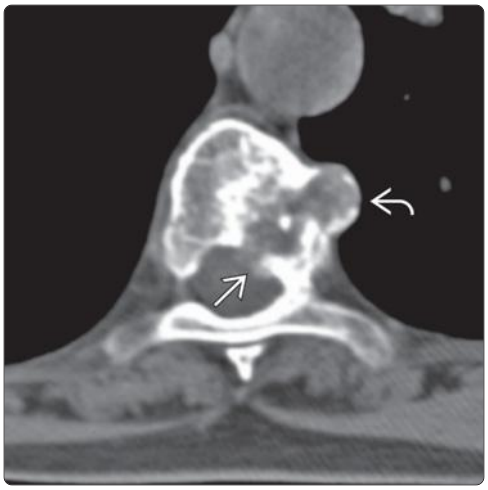
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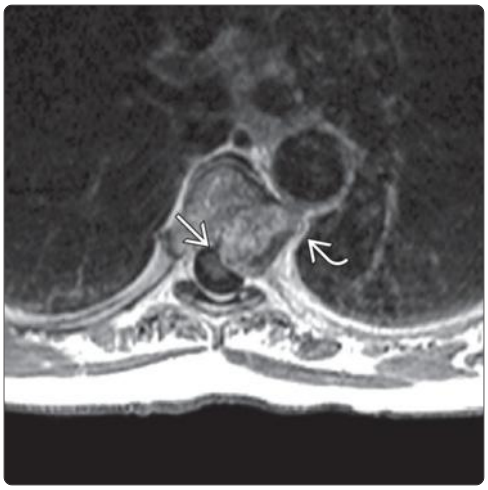
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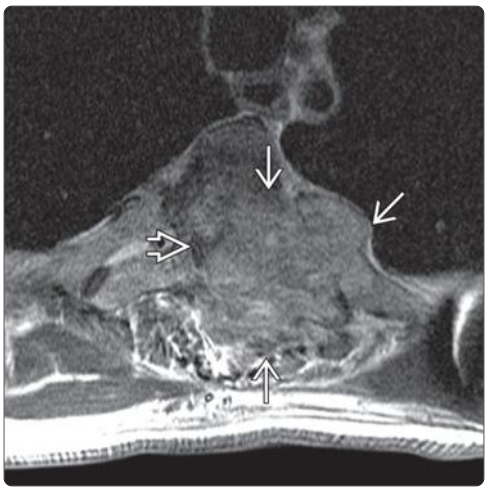


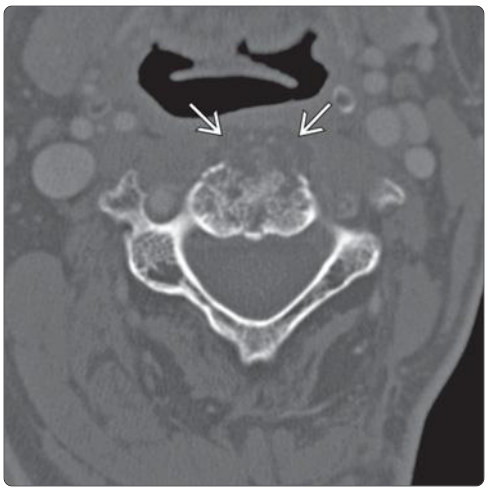
(Left) *Sagittal STIR MR shows a mass involving spinous process & body of upper thoracic vertebra. Epidural extension effaces the thecal sac with cord compression. MR does not visualize mineralization well, and the tumor appearance is nonspecific.* **(Right)** *Sagittal reconstructed bone CT (same patient) again shows expansile mass replacing the spinous process of the involved vertebra. Expansion of the neural arch dorsally effaces the spinal canal, resulting in significant stenosis.*





(Left) *Axial NECT reveals an expansile, lytic mass containing stippled calcifications. There is extension into the epidural and left paravertebral spaces.* **(Right)** *Axial enhanced T1WI in the same patient better defines epidural extension effacing the thecal sac and flattening the spinal cord. There is moderate, heterogeneous enhancement within the mass. Extension into the left paravertebral tissues is also seen.*





(Left) *Axial T1 C+ MR shows a large, destructive, heterogeneously enhancing mass. In addition to extension into the paravertebral and paraspinous soft tissues, there is epidural extension with severe canal stenosis and compression of the spinal cord.* **(Right)** *Axial bone CT of a cervical vertebra shows a destructive vertebral neoplasm, with a permeative growth pattern, extending into the prevertebral soft tissues. Fine, stippled calcifications are present.*

Osteosarcoma

KEY FACTS

TERMINOLOGY

- Sarcoma containing osteoid matrix produced directly by malignant cells

IMAGING

- Permeative or moth-eaten appearance
- Cortical breakthrough
- Bone sclerosis due to production of immature bone
- Wide zone of transition
- Discontinuous periosteal reaction, usually multilaminar
- Soft tissue mass
  - Fluid-fluid levels seen in telangiectatic OGS

TOP DIFFERENTIAL DIAGNOSES

- Sclerotic metastasis
- Osteoblastoma (OB)
- Aneurysmal bone cyst (ABC)
- Chordoma
- Osteomyelitis
- Ewing sarcoma
- Chondrosarcoma
- Lymphoma
- Malignant giant cell tumor

PATHOLOGY

- 4% of all primary OGS occur in spine and sacrum
- Majority of OGS of unknown etiology = primary OGS
- Association with retinoblastoma (*Rb* gene mutation)

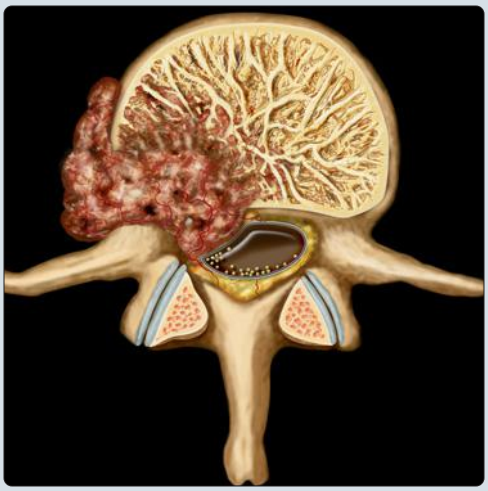
CLINICAL ISSUES

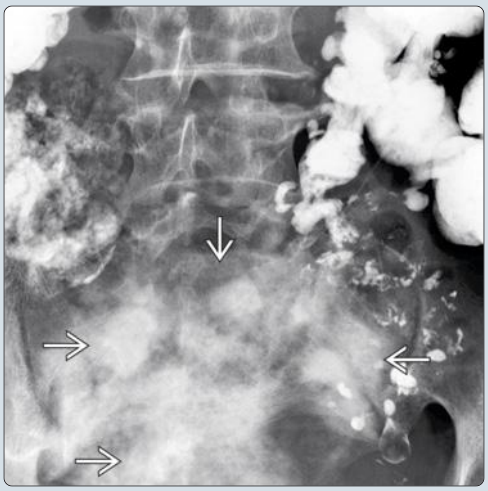
- Insidious onset of back pain, greatest at night
- Neurologic symptoms including radicular pain, weakness
- 3% of 10-year survivors of all OGS develop 2nd malignancy

DIAGNOSTIC CHECKLIST

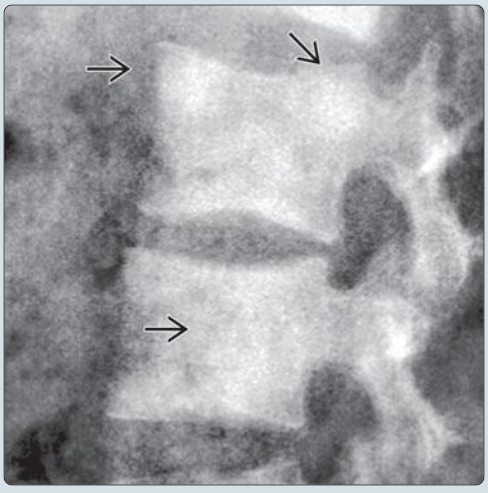
- CT scan best method for evaluation of tumor matrix, zone of transition
- All telangiectatic OGS are lytic on radiographs and CT, but not all lytic OGS are telangiectatic

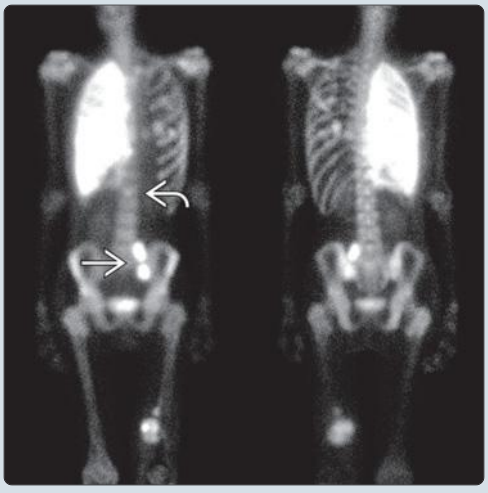
(Left) *Axial graphic shows secondary osteosarcoma arising in Pagetic vertebral body, destroying cortex and invading adjacent soft tissues. The soft tissue mass in osteosarcoma usually contains ossification.* **(Right)** *Anteroposterior radiograph shows multiple, poorly defined areas of sclerosis ➡ occupying the majority of the sacrum. This is a case of secondary osteosarcoma arising in Paget disease. The differential diagnosis includes sclerotic metastases.*





(Left) *Lateral radiograph shows metastatic OGS to lumbar spine. Multiple deposits of amorphous bone matrix ⇨ are visible in vertebral bodies, obliterating normal bony architecture.* **(Right)** *Nuclear medicine bone scan (same patient) shows metastatic OGS to spine ➡, ribs, pleural space, and iliac lymph nodes ➡ from femoral primary.*





TERMINOLOGY

Abbreviations

- Osteogenic sarcoma (OGS)

Definitions

- Sarcoma containing osteoid produced directly by malignant cells

IMAGING

General Features

- Best diagnostic clue
  - Aggressive-appearing lesion forming immature bone
- Location
  - 4% of all primary OGS occur in spine and sacrum
  - 79% arise in posterior elements
  - 17% involve 2 adjacent spinal levels
  - 84% invade spinal canal

Radiographic Findings

- Permeative or moth-eaten appearance
  - Multiple small areas of bone destruction
  - Best visualized at tumor periphery
- Wide zone of transition
  - Difficult to demarcate where tumor ends and normal bone begins
- Bone sclerosis due to production of immature bone
  - 80%: Bone matrix seen on radiographs or CT scan
  - 20%: Lytic appearance without visible bone matrix
- Cortical breakthrough
  - Usually extensive
- Discontinuous periosteal reaction, usually multilaminar
- Soft tissue mass
  - Often contains immature bone
- May cross sacroiliac joint or disc space

CT Findings

- Same as on radiographs, but better detail
- Best method to evaluate tumor matrix
  - OGS matrix is cloud-like, amorphous
- Contrast administration tends to obscure bone matrix and is not recommended

MR Findings

- Aggressive, heterogeneous mass similar to other malignancies
- Matrix difficult to see, CT more specific
- Densely ossified portions of tumor ↓ signal intensity on all sequences
- Nonossified tumor ↓ signal on T1WI, ↑ on T2WI, STIR
- Fluid-fluid levels seen in telangiectatic OGS

Nuclear Medicine Findings

- Bone scan
  - Increased uptake on all 3 phases
  - Used for staging, detection of skip lesions, metastases

Imaging Recommendations

- Best imaging tool
  - CT scan
- Protocol advice
  - Avoid IV contrast on bone CT to allow visualization of tumor matrix
  - MR useful to evaluate for cord, nerve root impingement
  - Staging should also include bone scan and chest CT scan

DIFFERENTIAL DIAGNOSIS

Sclerotic Metastasis

- Most commonly prostate, breast, gastrointestinal
- Sclerosis rarely extends beyond borders of bone
- Often multiple

Osteoblastoma (OB)

- Expansile, bone forming-lesion in posterior elements
- Can extend into vertebral body
- Cortical breakthrough often seen
- Narrow zone of transition
- Aggressive osteoblastoma mimics OGS on imaging studies

Aneurysmal Bone Cyst (ABC)

- Fluid-fluid levels similar to telangiectatic OGS
- Expansile lesion centered in posterior elements
- Cortical breakthrough often seen
- Narrow zone of transition

Chordoma

- Most common in sacrum
- Arises in midline
- Lytic, aggressive mass, no sclerosis

Osteomyelitis

- Occasionally sclerotic
- Usually involves 2 contiguous vertebrae and intervening disc space

Ewing Sarcoma

- Sometimes sclerotic
- Tends to permeate through cortex rather than cause large visible area of destruction

Chondrosarcoma

- Ring and arc calcifications

Lymphoma

- Moth-eaten lytic bone destruction
- Can rarely be sclerotic

Malignant Giant Cell Tumor

- Rare entity, most common in sacrum
- Lytic, aggressive mass without bone matrix formation
- Histologically similar to giant cell-rich OGS

Giant Bone Island

- Sclerotic focus in medullary bone
- Mature cortical bone
- Shows "brush border": Trabeculae merge with adjacent bone

PATHOLOGY

General Features

- Etiology
  - Majority of OGS of unknown etiology = primary OGS
  - Associated with retinoblastoma (*Rb* gene mutation)

- Secondary osteosarcoma associated with
  - Paget disease
  - Bone infarct
  - Radiation

Staging, Grading, & Classification

- Classified by predominant cell type histologically
  - Osteoblastic (conventional), chondroblastic, fibroblastic, telangiectatic, small cell, giant cell, epithelioid
- AJCC staging, based on size, grade, and metastases

Gross Pathologic & Surgical Features

- Heterogeneous mass with ossified and nonossified components
- Ossified areas: Yellow-white, firm, may be as hard as cortical bone
- Less ossified areas: Soft, tan, with foci of hemorrhage and necrosis
- Necrosis common
- Cortical breakthrough
- Often large extraosseous tumor mass

Microscopic Features

- Pluripotential neoplasm
- Malignant cells produce some osteoid in all subtypes, but it may be difficult to find
  - Osteoid is immature
- Classic: High-degree anaplasia, high mitotic rate
  - Tumor cells may be spindled or round
  - Size of cells ranges from small to giant
- Telangiectatic OGS
  - Dilated vascular channels lined by multinucleated giant cells
  - Stroma forms osteoid, which may not be prominent feature
- Other malignant tumors (e.g., chondrosarcoma) may contain reactive bone but osteoid is not produced by malignant cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Insidious onset of back pain, greatest at night
  - Neurologic symptoms including radicular pain, weakness
- Clinical profile
  - Pathologic fracture
  - Most common sites of metastases are liver, lung, bone, and lymph nodes
    - Metastases often calcify
  - Pulmonary metastases may result in pneumothorax
  - Increased serum alkaline phosphatase

Demographics

- Age
  - Spine OGS has peak incidence in 4th decade
    - Later than for appendicular OGS
  - Range: 8-80 years in 1 large series
- Gender
  - M = F
- Epidemiology
  - 2nd most common primary bone malignancy (after multiple myeloma)

Natural History & Prognosis

- Median survival: 23 months in recent series
- Survival rate lower than for peripheral OGS due to difficulty of surgical resection
- 3% of 10-year survivors of all OGS develop 2nd malignancy
- Patients should have close CT (or PET/CT) surveillance

Treatment

- Neoadjuvant chemotherapy ± radiation therapy
- Surgical resection with wide margins
- Biopsies must be planned with future tumor excision in mind

DIAGNOSTIC CHECKLIST

Consider

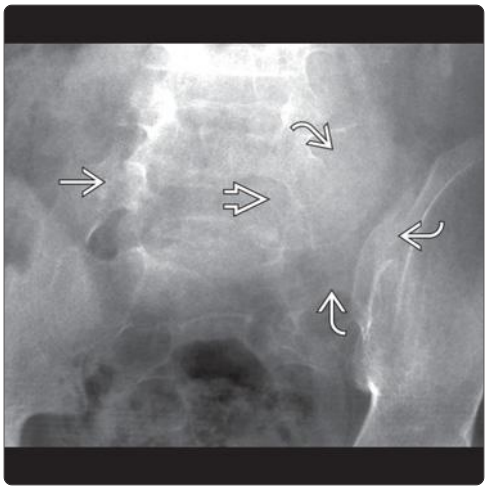
- CT scan best method for evaluation of tumor matrix, zone of transition
- Patients may present with multiple synchronous tumors (osteosarcomatosis)

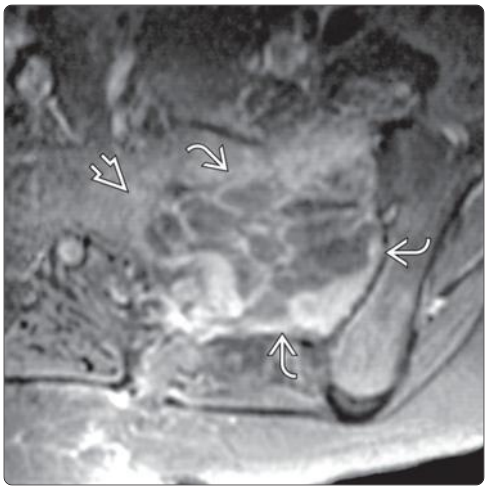
Image Interpretation Pearls

- All telangiectatic OGS are lytic on radiographs and CT, but not all lytic OGS are telangiectatic

SELECTED REFERENCES

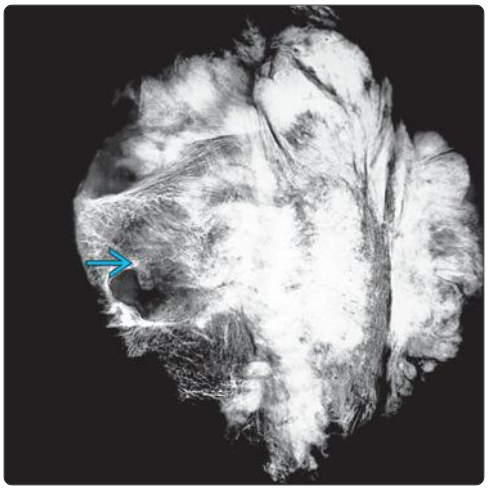
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(Left) *Anteroposterior radiograph shows lytic OGS at left transitional vertebra. The left L5 pedicle is destroyed (compare to the normal right pedicle). Loss of symmetry is a useful clue to spine neoplasms.* **(Right)** *Axial T1WI C+ MR in the same patient shows telangiectatic OGS of sacrum. Fluid-filled nonenhancing regions are present, as well as enhancing tumor nodules. Zone of transition medially is broad and difficult to demarcate.*





(Left) *Axial bone CT shows an aggressive-appearing, densely ossified mass characteristic of osteosarcoma. Mass involves both the sacrum and the ilium, crossing the sacroiliac joint. Immature ossification extends into the soft tissues.* **(Right)** *Anteroposterior specimen radiograph in the same patient shows resected tumor, which extends to the S1 neural foramen. Although other neoplasms may develop reactive bone, it is rarely as extensive as in osteosarcoma.*





(Left) *Sagittal T1WI MR shows a permeative, destruction lesion of L3 with epidural extension. There is loss of vertebral body height. This appearance is more commonly seen with Ewing sarcoma, and it must be remembered that osteosarcoma may mimic Ewing sarcoma (and vice versa).* **(Right)** *Sagittal STIR MR in the same patient shows that the mass is heterogeneously increased in signal intensity. Bone matrix is poorly seen on MR. However, MR is useful to assess epidural extension.*

Chordoma

KEY FACTS

TERMINOLOGY

- Malignant tumor arising from notochord remnants

IMAGING

- Sacrococcygeal > sphenooccipital > mobile spine
- Osseous destruction with disproportionately large soft tissue mass
- Circumscribed, scalloped, or sclerotic bony margins
- Amorphous intratumoral calcifications
- Hyperintense to disc on T2WI, with multiple septa
- May extend into disc space, involve 2 or more adjacent vertebrae

TOP DIFFERENTIAL DIAGNOSES

- Chondrosarcoma
- Giant cell tumor
- Metastases
- Plasmacytoma

PATHOLOGY

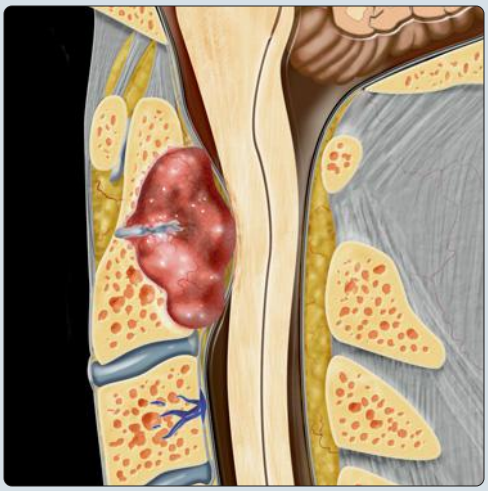
- Lobulated, soft, grayish gelatinous (myxoid) mass
- Areas of calcification and hemorrhage

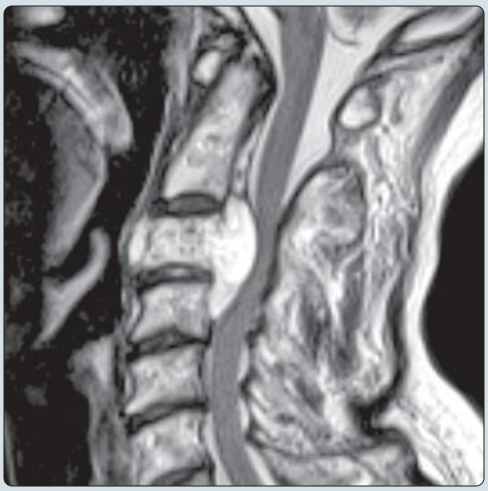
CLINICAL ISSUES

- Skull base
  - Diplopia (CNV2 palsy, most common)
  - Headaches
  - Facial pain
- Mobile spine: Cord compression, radiculopathy (50%)
- Sacral: Altered sacrogluteal sensation
- Symptoms tend to be longstanding (4-24 months)
- Recurrence common
- 5-year survival: 50-68%; 10-year survival: 28-40%

DIAGNOSTIC CHECKLIST

- High signal intensity mass on T2WI with septations, little enhancement is chordoma or chondrosarcoma





(Left) *Sagittal graphic of the cervical spine shows extradural soft tissue mass with the epicenter in the posterior aspect of the C2 body, causing bone destruction and epidural extension with cord compression.* **(Right)** *Sagittal T2WI MR shows hyperintense mass involving the C3 body with extensive epidural extension and cord compression.*





(Left) *Sagittal T2WI show a large, well-defined soft tissue mass engulfing the distal sacrum and coccyx, with extension into the presacral space and dorsal soft tissues. The mass is hyperintense with linear hypointense septations.* **(Right)** *Sagittal T2WI shows a large heterogeneously hyperintense mass arising from the C2-C3 level with extension into the cervical epidural space and anterolateral prevertebral tissues.*

TERMINOLOGY

Definitions

- Malignant tumor arising from notochord remnants

IMAGING

General Features

- Best diagnostic clue
  - Destructive process with disproportionately large soft tissue mass
  - Located in clivus or sacrum
  - Mass is hyperintense to disc on T2WI, with multiple septa
- Location
  - Sacrococcygeal > sphenooccipital > mobile spine
- Size
  - Variable
  - Sacrococcygeal chordomas can be huge at presentation
- Morphology
  - Midline lobular soft tissue mass with osseous destruction

Radiographic Findings

- Radiography
  - Heterogeneous destructive mass of sacrum or vertebral body
  - May extend into disc space, involve 2 or more adjacent vertebrae
  - May enlarge neural foramen
  - Osseous sclerosis in up to 60% of cases
    - Solitary sclerotic "ivory" vertebra (rare)

CT Findings

- NECT
  - Destructive, lytic lesion
  - Soft tissue mass usually present
    - Often disproportionately large relative to bony destruction
  - Amorphous intratumoral calcifications
    - Sacrum: > 70%
    - Vertebra: 30%
- CECT
  - Mild to moderate enhancement
  - ± inhomogeneous areas (cystic necrosis, hemorrhage)

MR Findings

- T1WI
  - Heterogeneous hypo- to isointense (compared to marrow)
- T2WI
  - Hyperintense to CSF, intervertebral discs
  - May have low signal septations (fibrous)
- T1WI C+
  - Variable enhancement: Blush → intense enhancement

Nonvascular Interventions

- Myelography
  - Large extradural mass

Nuclear Medicine Findings

- Bone scan
  - Normal or decreased uptake

Imaging Recommendations

- Best imaging tool
  - MR for soft tissue (STIR/fat-saturated T2WI, contrast-enhanced T1WI)
  - NECT for bone detail

DIFFERENTIAL DIAGNOSIS

Chondrosarcoma

- Neural arch > vertebral body
- Chondroid matrix (rings and arcs)
- Mild peripheral and internal septal enhancement
- Similar MR characteristics

Giant Cell Tumor

- Heterogeneous, intermediate to hyperintense T2 signal
- May infiltrate disc space, with spread to adjacent vertebral body

Metastases

- Hypointense T1, variably hyperintense T2WI lesion(s)
- Often multiple, involving vertebral bodies and posterior elements

Plasmacytoma

- Destructive vertebral body lesion, similar appearance to lytic metastases

Lymphoma

- Multifocal disease; heterogeneous T2 signal

Sacrococcygeal Teratoma

- Heterogeneous MR signal
- Areas of macroscopic fat hyperintense on T1WI
- Pediatric patients

Ecchordosis Physaliphora (Rare)

- Benign, nonneoplastic ectopic notochordal remnant(s)
- Usually at skull base/C2 but can occur anywhere (including intradural)

PATHOLOGY

General Features

- Etiology
  - Arises from notochord remnants
- Genetics
  - Losses on chromosomal arms 3p (50%) and 1p (44%)
  - Gains on 7q (69%), 20 (50%), 5q (38%), and 12q (38%)
- Location
  - 2-4% of primary malignant bone tumors
  - Sacrococcygeal (50%), sphenooccipital (35%), mobile spine (15%)
  - Mobile spine: Cervical (20-50%) > lumbar > thoracic
- Embryology-anatomy
  - Tumor arises from notochordal remnants
    - Notochord (column of cells ventral to neural tube) arises 3rd gestational week, disappears by 7th week
    - Rest of notochord cells occur in axial skeleton from coccyx to dorsum sellae

Staging, Grading, & Classification

- Enneking system for staging musculoskeletal sarcomas

- Grade of biologic aggressiveness
- Anatomic setting
- Presence of metastasis

Gross Pathologic & Surgical Features

- Lobulated, soft, grayish gelatinous (myxoid) mass
- Areas of calcification and hemorrhage

Microscopic Features

- 3 types described
  - Typical: Lobules, sheets, and cords of clear cells with intracytoplasmic vacuoles (physaliphorous cells); abundant mucin
  - Chondroid: Hyaline cartilage (usually sphenooccipital region)
  - Dedifferentiated: Sarcomatous elements (rare, highly malignant)
- Immunohistochemistry: Positive for S100, keratin (AE1/AE3; CK8, CK19, and often CK5), HBME-1, epithelial membrane antigen; usually negative for CK7, CK20

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Location dependent
    - Clival
      - Diplopia (CNV2 palsy, most common)
      - Headaches
      - Facial pain
    - Sacral
      - Weakness
      - Sacral mass
      - Altered sacrogluteal sensation
      - Bowel/bladder/sexual dysfunction
    - Spinal
      - Cord compression, radiculopathy (50%)
      - Cervical spine chordomas may present with airway obstruction, dysphagia
  - Symptoms tend to be longstanding (4-24 months)
- Clinical profile
  - Middle-aged man presenting with pain, neurologic dysfunction

Demographics

- Age
  - Peak incidence 5th-6th decades (rare in children)
- Gender
  - M:F = 2:1
    - More equal sex distribution for clival chordoma
- Ethnicity
  - Rare in African Americans
- Epidemiology
  - 2-4% of primary malignant bone neoplasms
  - Most common primary nonlymphoproliferative malignancy of spine
  - Incidence rate of 0.08 per 100,000
  - Rare among patients < 40 years old

Natural History & Prognosis

- Slow growing
- Distant metastases 5-40% (lung, liver, lymph nodes, bone)
- Poor prognostic factors
  - Large size
  - Subtotal resection, local recurrence
  - Microscopic necrosis
  - Ki-67 index > 5%
- 5-year survival up to 67-84%
- 10-year survival: 40%

Treatment

- Surgical resection with adjuvant XRT
  - En bloc resection yields best outcome
- Recurrence common
  - Local (90%)
  - Regional lymph nodes (5%)
  - Distant metastases (5%); lung, bone
  - 5-year survival: 50-68%; 10-year survival: 28-40%

DIAGNOSTIC CHECKLIST

Consider

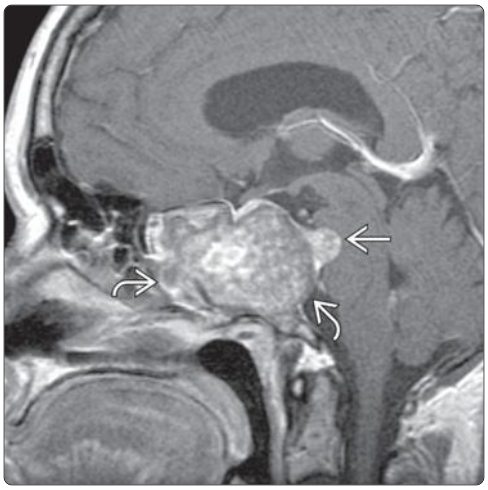
- High signal intensity mass on T2WI with septations, little enhancement is chordoma or chondrosarcoma

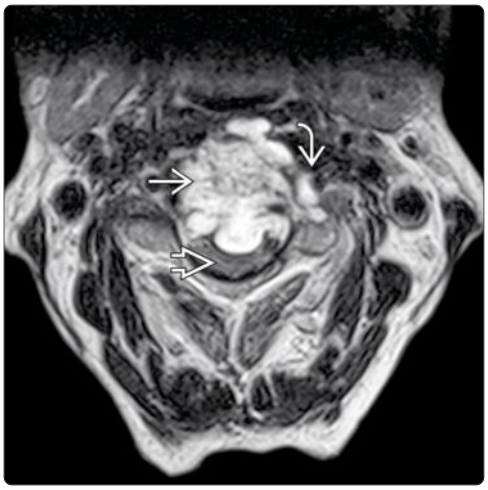
Image Interpretation Pearls

- Recurrence (seeding) along operative tract is not uncommon, modify field of view to include operative approach

SELECTED REFERENCES

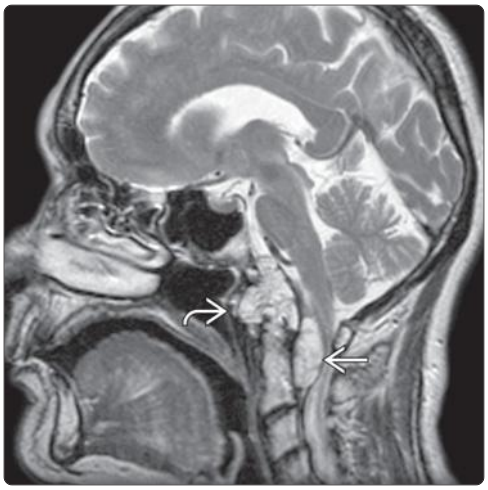
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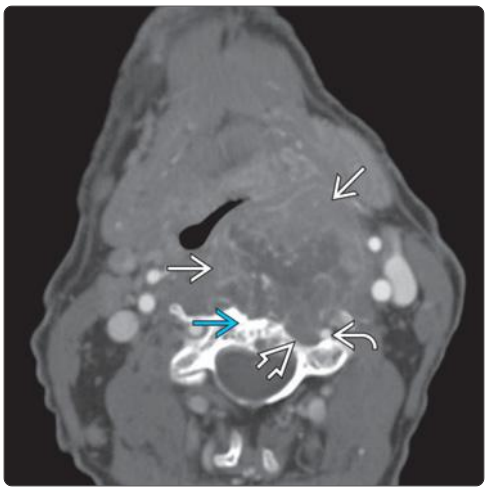
(Left) *Sagittal T1WI C+ MR shows an expansile, heterogeneously enhancing mass arising from the clivus and basisphenoid. Prepontine cistern is effaced, and there is focal nodular excrescence deforming the surface of the pons.* **(Right)** *Axial T2WI MR shows chordoma replacing the C3 body, extending into epidural space and compressing the cord. Tumor extends into left foramen transversarium and along the vertebral artery. Typical T2 hyperintensity and internal septations are seen.*





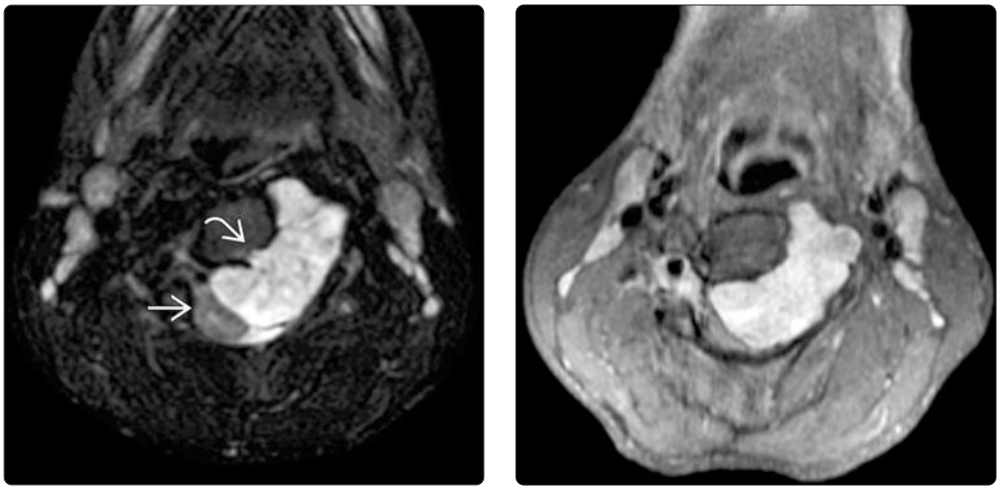
(Left) *Axial T2WI through the skull base shows an expansile mass replacing the clivus. Hyperintense T2 signal, lobulated morphology, and fine hypointense internal septations are characteristic of chordoma.* **(Right)** *Sagittal T2WI MR shows an expansile, destructive mass arising from an epicenter in the basion. Epidural extension caudally along the dorsal aspect of the dens effaces the thecal sac and compresses the cervicomedullary junction.*



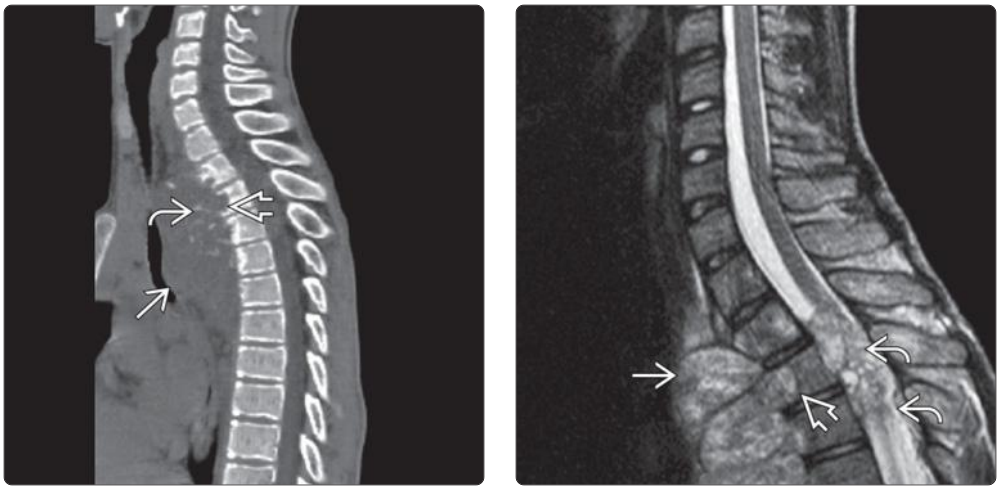


(Left) *Sagittal T2WI MR shows heterogeneous, moderately hyperintense prevertebral mass extending from mid C2 through the C4-C5 level. There is contiguous involvement of the anterior C3 vertebral body with a faint sclerotic margin.* **(Right)** *Axial CTA in the same patient shows an exophytic mass in the prevertebral tissues with smoothly scalloped margins of vertebral involvement and areas of osseous sclerosis. There is no matrix. The vertebral artery is displaced, but patent.*

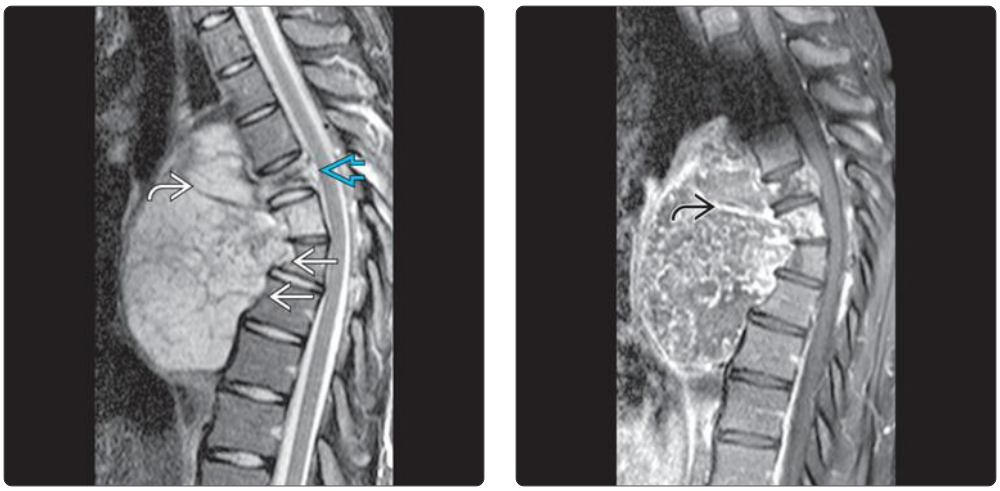
(Left) *Axial T2WI FS MR shows a high signal extradural mass compressing the cervical cord and extending into the left prevertebral region. Fine internal septations are hypointense. There is focal loss of the C3 cortical bone. Osseous involvement is minimal relative to the soft tissue mass. The mass enlarges the left neural foramen.* **(Right)** *Axial T1WI FS C+ MR in the same patient shows diffuse, mildly heterogeneous enhancement of the mass.*



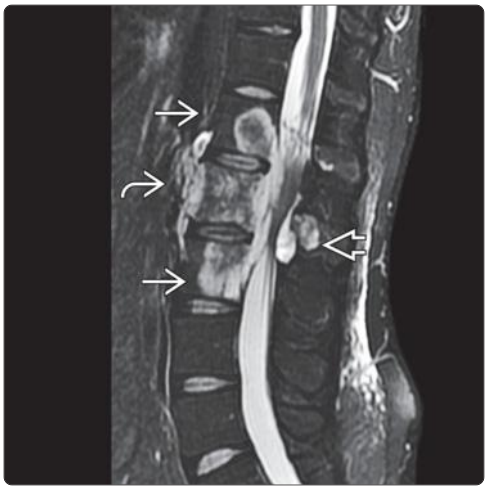
(Left) *Sagittal NECT shows a small amount of bony destruction with a disproportionately large paravertebral/posterior mediastinal component. Irregularly calcific matrix is present.* **(Right)** *Sagittal T2WI MR in the same patient shows osseous destruction and a large paravertebral soft tissue component. A large volume of tumor within the thoracic canal was occult on NECT. Soft tissue components are moderately and heterogeneously hyperintense with lobulated morphology.*



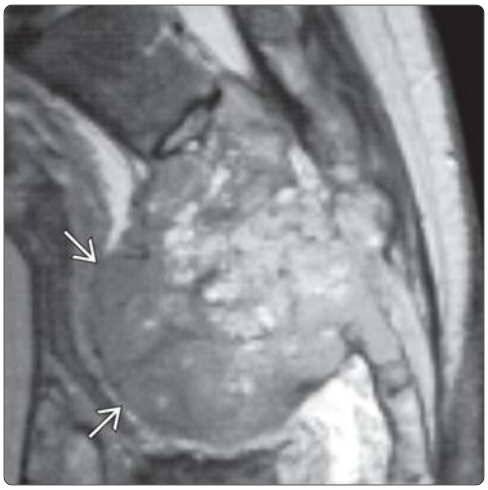
(Left) *Sagittal T2WI FS MR shows a large posterior mediastinal mass with involvement of 4 adjacent thoracic vertebral bodies. The mass is hyperintense with fine septations. There is a pathologic compression fracture of the top vertebra, with ventral epidural tumor. Inferiorly, there are scalloped anterior margins.* **(Right)** *Sagittal T1WI C+ FS MR in the same patient shows a posterior mediastinal mass. Markedly heterogeneous enhancement mirrors the pattern of T2 hypointense septations.*

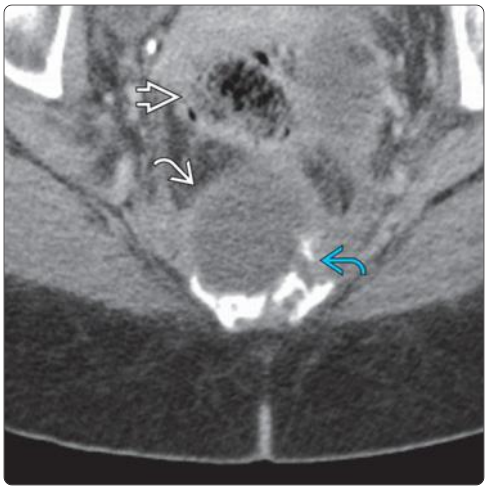




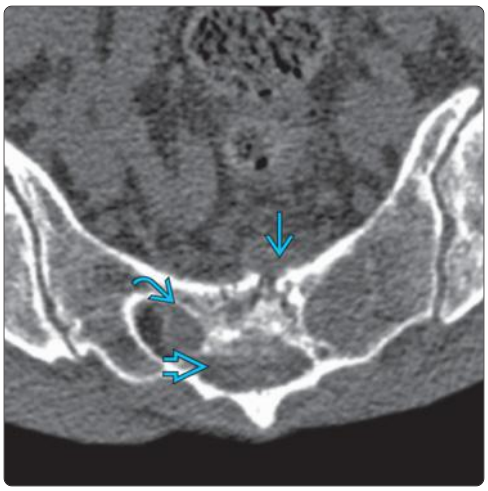


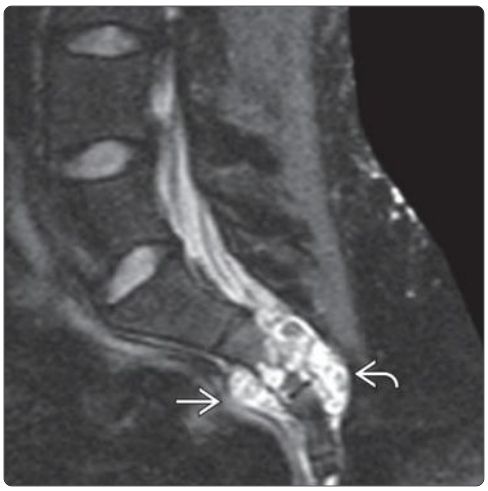
(Left) *Sagittal NECT shows predominately osteosclerotic involvement of the L1-L3 bodies in this large lumbar chordoma.* **(Right)** *Sagittal STIR MR shows a bony and soft tissue mass involving multiple vertebral bodies without involvement of disc spaces, with extension to the posterior elements. There is ventral paraspinal soft tissue involvement and epidural extension with severe compression of the thecal sac at L2.*





(Left) *Sagittal T2WI shows a destructive sacral mass with a large extraosseous component in the presacral space. The signal is heterogeneous and only mildly to moderately hyperintense relative to the skeletal muscle.* **(Right)** *Axial CECT shows destructive changes in the sacrococcygeal junction and fragment of calcification. There is a cystic mass, an unusual variant appearance of sacrococcygeal chordoma, extending ventrally from this region, displacing the rectum.*





(Left) *Axial NECT (bone window) shows a heterogeneous, destructive mass of the sacrum with infiltrative margins. There is focal penetration of the anterior cortex. Appreciation of extension into the sacral canal and right S2 neural foramen is limited by lack of contrast.* **(Right)** *Sagittal STIR MR in the same patient shows an expansile, destructive mass in the S2 segment with extension into the presacral tissues & sacral canal. The mass is heterogeneously hyperintense, with low signal septa.*

Ewing Sarcoma

KEY FACTS

TERMINOLOGY

- Ewing sarcoma family of tumors (ESFT)
- Aggressive childhood cancer which includes Ewing sarcoma, Askin tumor, and peripheral primitive neuroectodermal tumor (PNET)

IMAGING

- Spine: 5% of all Ewing tumors
  - Sacrum most common spinal site
  - Spreads along peripheral nerves
- May originate in epidural or paraspinous soft tissues
- Permeative/"moth-eaten" bone destruction
  - 5% sclerotic (represents host reaction, not tumor matrix)
  - Areas of central necrosis common
  - "Percolates" through tiny perforations in cortex
- Lower signal intensity than disc or muscle on T1WI
  - May be isointense to red marrow on T2WI
  - Intermediate to high signal intensity on STIR
- MR best shows involvement of adjacent bones and soft tissues, which can be underestimated on CT scan
  - Heterogeneous enhancement with gadolinium (Gd)
- CT useful to confirm absence of tumor matrix, distinguish from osteogenic sarcoma (OGS)

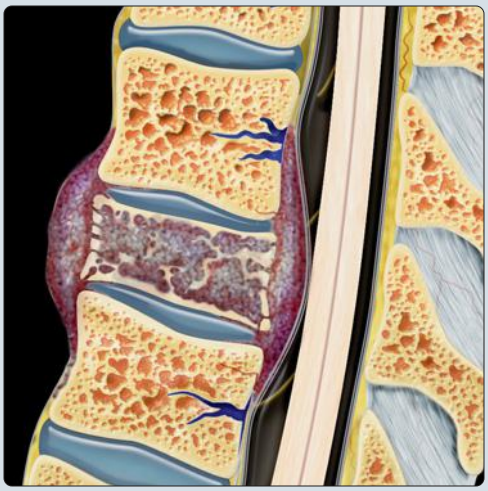
TOP DIFFERENTIAL DIAGNOSES

- Primitive neuroectodermal tumor (PNET)
- Langerhans cell histiocytosis
- Osteosarcoma
- Metastatic neuroblastoma
- Osteomyelitis

CLINICAL ISSUES

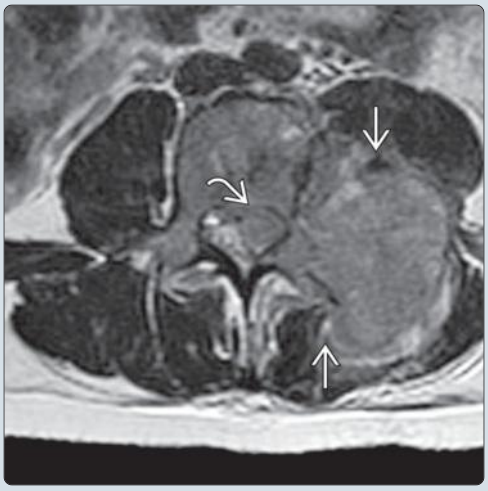
- 90% of all Ewing sarcoma patients present before 20 years
- Fever, leukocytosis, elevated ESR (simulating osteomyelitis)
- Spine and sacral lesions often in older patients than in peripheral Ewing sarcoma

(Left) *Sagittal graphic shows vertebral body replaced by Ewing sarcoma, resulting in mild collapse. Tumor extends into adjacent soft tissues through small perforations in bone cortex.* **(Right)** *Axial CECT shows large paraspinal soft tissue mass of Ewing sarcoma engulfing the left psoas muscle with extension into the dorsal musculature, destruction of left side of lumbar body, and epidural extension.*





(Left) *Axial T2WI MR shows Ewing sarcoma involving the paraspinal muscles and extending into the epidural space, compressing and displacing the thecal sac to the right. Infiltration of vertebral body is poorly defined on this sequence.* **(Right)** *Axial T1WI C+ MR shows diffuse heterogeneous enhancement of the tumor involving the left side of lumbar body, epidural space, and paravertebral and dorsal muscles.*





TERMINOLOGY

Synonyms

- Ewing sarcoma family of tumors (ESFT)
- Round cell sarcoma of bone

Definitions

- Aggressive childhood cancer which includes Ewing sarcoma, Askin tumor, and peripheral primitive neuroectodermal tumor (PNET)

IMAGING

General Features

- Best diagnostic clue
  - Permeative lytic lesion of vertebral body or sacrum
- Location
  - Spine: 5% of all Ewing tumors
  - Involves vertebral body before neural arch
  - Sacrum most common spinal site
  - May involve adjacent bones: Vertebrae, ribs, or ilium
  - Spreads along peripheral nerves
  - May originate in epidural or paraspinous soft tissues

Radiographic Findings

- Radiography
  - Centered in vertebral body or sacrum
  - Permeative/"moth-eaten" bone destruction
  - Wide zone of transition
    - Difficult to identify tumor margin
  - Cortical destruction difficult to see
    - Tiny perforations of cortex better seen on CT
  - 50% have extraosseous, noncalcified soft tissue mass
  - 5% sclerotic (represents host reaction, not tumor matrix)
  - May cause vertebra plana
  - May involve 2 or more adjacent bones
  - If adjacent vertebrae involved, do not see disc height loss and endplate erosions as in osteomyelitis

CT Findings

- Permeative intramedullary mass ± soft tissue mass
- Areas of central necrosis common
- "Percolates" through tiny perforations in cortex
  - Usually do not see large areas of cortical breakthrough
- Heterogeneous contrast enhancement
- Rarely sclerotic, due to reactive new bone formation
  - Mimics osteogenic sarcoma (OGS)
  - Ossification is not present in soft tissue component
- Soft tissue Ewing is nonspecific mass

MR Findings

- Poorly defined lesion originating in bone or soft tissue
  - Lower signal intensity than disc or muscle on T1WI
  - May be isointense to red marrow on T2WI
  - Intermediate to high signal intensity on STIR
- Central necrosis often present
  - Heterogeneous enhancement with Gd
    - Cannot reliably distinguish between tumor and peritumoral edema
      - Peritumoral edema enhances with Gd
- Bone cortex often visible despite soft tissue extension of bone mass
  - Has "smudged" appearance due to perforation of tumor through cortex

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan
- PET
  - Increased FDG uptake of tumor, metastases

Imaging Recommendations

- Best imaging tool
  - MR best shows involvement of adjacent bones and soft tissues, which can be underestimated on CT scan
    - May overestimate tumor size due to peritumoral edema
- Protocol advice
  - MR to determine extent of tumor
    - Sagittal T1WI
    - Axial and sagittal STIR, post-contrast T1WI
  - CT useful to confirm absence of tumor matrix, distinguish from OGS

DIFFERENTIAL DIAGNOSIS

PNET

- Clinically/radiologically identical to Ewing sarcoma
- May occur in bone or soft tissue
- Greater neuroectodermal differentiation of tumor cells

Langerhans Cell Histiocytosis

- May have identical radiographic appearance to Ewing sarcoma
- May form discrete geographic lytic lesion

Lymphoma and Leukemia

- Same radiographic appearance as Ewing sarcoma
- Ill-defined lytic lesion showing permeative pattern
- Involves vertebral body more than neural arch
- Often involves multiple vertebrae

Metastatic Neuroblastoma

- Adrenal or extraadrenal primary
- Occurs in children
- Identical appearance in bone

Osteosarcoma

- Ill-defined lytic lesion showing permeative pattern
- 80% show bone matrix on radiographs or CT
- Involves vertebral body or neural arch
- May involve adjacent vertebral body
- Tends to have cortical destruction rather than permeation through cortex

Osteomyelitis

- Ill-defined lytic lesion showing permeative pattern
- May be more geographic (intraosseous abscess with peripheral enhancement)
- Involves vertebral body more than neural arch
- Discocentric: Extends from 1 vertebral body to adjacent vertebra across disc

- Disc height loss, enhancement of disc on MR, endplate erosions
- Soft tissue mass common

Multiple Myeloma

- Occurs in older patients
- Usually multiple vertebrae

Chordoma

- Most common in sacrum
- Originates in midline
- Bone expansion and destruction

Giant Cell Tumor

- Originates in vertebral body
- Narrow zone of transition
- Focal cortical destruction

Chondrosarcoma

- Usually show areas of chondroid matrix
  - Rings and arcs of calcification

PATHOLOGY

General Features

- Etiology
  - Mesenchymal cells with slight differentiation toward neuroectodermal cells
- Genetics
  - Reciprocal translocation between *EWS* gene on chromosome 22 and *ETS*-like genes on chromosome 11 - t(11;22)

Staging, Grading, & Classification

- No internationally recognized risk classification scheme for Ewing sarcoma patients

Gross Pathologic & Surgical Features

- Grayish-white tumor
- Poorly demarcated
- Areas of hemorrhage, cyst formation, necrosis

Microscopic Features

- Small, round cells (2-3x larger than lymphocytes), meager cytoplasm
- Cell outlines indistinct
- Round nuclei, frequent indentations, high mitotic rate
- Solid sheets of cells divided into irregular masses by fibrous strands

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Localized pain
  - Fever, leukocytosis, elevated ESR (simulating osteomyelitis)
  - Neurologic symptoms ranging from radiculopathy to paralysis
  - Vertebra plana

Demographics

- Age
  - 90% of all Ewing sarcoma patients present before 20 years
  - 2nd (smaller) peak at 50 years
  - Spine and sacral lesions often in older patients than in peripheral Ewing sarcoma
- Gender
  - M:F = 2:1
- Epidemiology
  - Annual incidence of Ewing sarcoma (all locations): 3/1,000,000 Caucasian children < 15 years old
  - 6th most common malignant bone tumor

Natural History & Prognosis

- Metastases to lung, regional lymph nodes, and other bones in 30% at presentation
- Prognosis worse in spinal than peripheral Ewing sarcoma due to difficulty of surgical resection
- Significant risk of 2nd malignancy
- Current treatments yield long-term survival in > 50% of patients with localized disease at presentation
- Complications common > 5 years after treatment
  - Local recurrence
  - Metastases
  - 2nd malignancies

Treatment

- Surgery or radiotherapy; without chemotherapy universally fatal
- Neoadjuvant chemotherapy given prior to surgery
- Surgical resection with wide margins
- Radiation therapy for surgically inaccessible lesions, stage III disease, poor response to chemotherapy

DIAGNOSTIC CHECKLIST

Consider

- Often metastatic at time of presentation

Image Interpretation Pearls

- Always consider osteomyelitis and other small round cell tumors in imaging differential diagnosis

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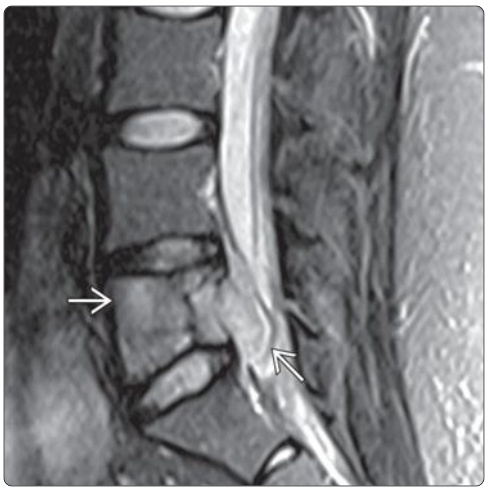
(Left) *Anteroposterior radiograph shows a Ewing tumor of L2. The bone destruction is difficult to see, but the left pedicle is destroyed and the vertebral body appears mottled. There is mild loss of vertebral body height.* **(Right)** *Axial bone CT in the same patient shows a lytic lesion in the vertebral body extending into the left pedicle. There is cortical destruction posteriorly, but the lateral vertebral body cortex is slightly thinned, with a small amount of periosteal new bone.*





(Left) *Sagittal T1WI MR in a 30-year-old man shows Ewing sarcoma involving nearly the complete L5 vertebral body, with epidural extension. The low signal intensity line is due to reactive sclerosis and is not the tumor margin. Tumor is slightly lower in signal intensity than red marrow and muscle on T1WI.* **(Right)** *Axial bone CT in the same patient better shows the sclerotic reactive bone within the tumor. There is soft tissue extension laterally.*





(Left) *Sagittal T2WI MR in the same patient shows the tumor is primarily isointense to red marrow.* **(Right)** *Sagittal STIR MR in the same patient shows the tumor is hyperintense to red marrow. Ewing sarcoma tends to be most conspicuous on STIR sequences.*

Lymphoma

KEY FACTS

TERMINOLOGY

- Lymphoreticular neoplasms with a myriad of specific diseases and cellular differentiation

IMAGING

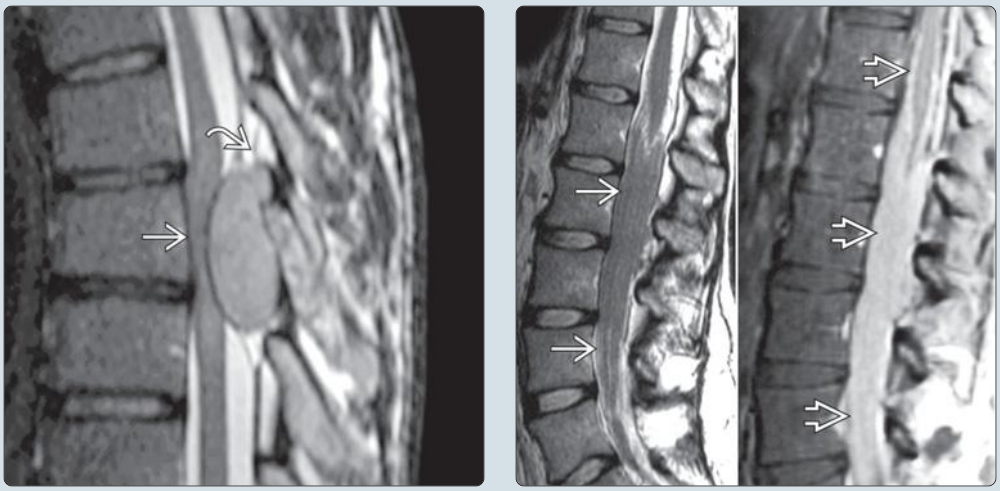
- Multiple types with variable imaging manifestations
- Epidural lymphoma: Thoracic > lumbar > cervical
  - Enhancing epidural mass ± vertebral involvement
- Osseous lymphoma: Long bones > spine
  - Bone destruction ("ivory" vertebra, rare), vertebra plana
- Lymphomatous leptomeningitis
  - Smooth/nodular pial enhancement
- Intramedullary lymphoma: Cervical > thoracic > lumbar
  - Poorly defined, enhancing mass
- Secondary > primary involvement
- Extradural > intradural > intramedullary
- FDG PET useful for staging, monitoring treatment response, predicting treatment outcomes, and risk stratifying lymphoma patients

PATHOLOGY

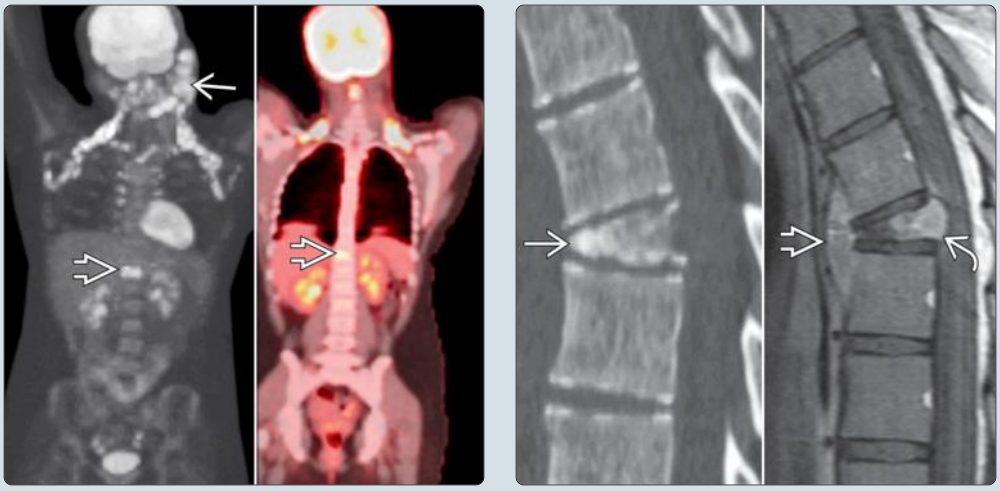
- NHL > > Hodgkin disease (HD); 80-90% are B cell
  - CNS lymphoma > 85% NHL (B cell > > > T cell)
- CNS lymphoma may be primary or secondary (hematogenous or direct geographic extension)

CLINICAL ISSUES

- Most common presenting symptom = back pain
- Intramedullary = myelopathy (weakness, numbness)
- Cord compression occurs in up to 5-10% of systemic lymphomas
- Generally poor prognosis for CNS lymphoma
- Markedly sensitive to chemotherapy/XRT
- Depressed humoral and cell-mediated immunity leads to opportunistic infections



(Left) *Sagittal T2WI MR demonstrates a discrete hypointense mass in the posterior epidural space with a cap of epidural fat. The spinal cord is displaced anteriorly. Lymphoma is the most common malignancy of the epidural space.* **(Right)** *Sagittal T2WI MR (left) shows amorphous hypointense tumor mass insinuating along the cauda equina. Sagittal T1WI C+ MR (right) reveals enhancing lymphoma within the leptomeninges surrounding the distal conus and involving the nerve roots diffusely.*



(Left) *Coronal PET (left) shows FDG uptake in the left cervical lymph node mass that was clinically evident. PET/CT fused image (right) shows abnormal activity at T11. This is an example of osseous metastases from cervical Hodgkin lymphoma primary tumor.* **(Right)** *Sagittal CT (left) demonstrates a pathologic compression fracture of a midthoracic vertebral body. Sagittal T1WI C+ MR (right) confirms abnormal marrow enhancement and a ventral paraspinal mass.*

TERMINOLOGY

Definitions

- Lymphoreticular neoplasms with a myriad of specific diseases and cellular differentiation

IMAGING

General Features

- Location
  - Multiple types with variable imaging manifestations
    - Epidural lymphoma: Thoracic > lumbar > cervical
      - Epidural extension from adjacent vertebral/paraspinous disease
    - Osseous lymphoma: Long bones > spine
    - Lymphomatous leptomeningitis
    - Intramedullary lymphoma: Cervical > thoracic > lumbar
  - Secondary > primary involvement
    - 30% of systemic lymphomas have skeletal involvement
    - Primary osseous lymphoma = 3-4% of all malignant bone tumors
  - Extradural > intradural > intramedullary
- Best imaging clues
  - Epidural: Enhancing epidural mass ± vertebral involvement
  - Osseous: Bone destruction ("ivory" vertebra rare)
  - Leptomeningitic: Smooth/nodular pial enhancement
  - Intramedullary: Poorly defined enhancing mass

Radiographic Findings

- Radiography
  - Epidural: May see bony erosion
  - Osseous
    - Bone destruction (30-40%)
    - Rare "ivory" vertebral body, vertebra plana

CT Findings

- NECT
  - Epidural: Homogeneous, slightly hyperdense mass, ± bone involvement
- CECT
  - Epidural: Homogeneous enhancement
- Bone CT
  - Osseous: Lytic, permeative bone destruction
    - Multilevel involvement, crossing disc spaces

MR Findings

- T1WI
  - Epidural: Isointense homogeneous epidural mass (often multisegmental ± extend through foramina)
  - Osseous: Hypointense to normal marrow (± epidural extension)
  - Leptomeningitic: Thick nerve roots ± focal nodules (isointense with cord)
  - Intramedullary: Mass isointense to spinal cord
- T2WI
  - Epidural: Iso-/hyperintense to cord
  - Osseous: Variable; iso-/hyperintense
  - Leptomeningitic: Thick nerve roots ± focal nodules (isointense with cord)
  - Intramedullary: Hyperintense + surrounding edema
- STIR
  - Osseous: Increased conspicuity of T2 characteristics
- DWI
  - ADC measurements inversely correlate with histopathologic assessment of tumor cellular density
- T1WI C+
  - Epidural: Enhances intensely, uniformly
  - Osseous: Diffuse uniform enhancement
  - Leptomeningitic: Nerve roots enhance
  - Intramedullary: Variable; patchy/confluent, infiltrating/discrete
- Dynamic, contrast–enhanced MR
  - Demonstrates ↑ bone marrow (BM) enhancement in patients with lymphoproliferative diseases with diagnostic accuracy of 99%
  - ↓ BM enhancement after treatment in all patients who respond to treatment, but not in 2/3 patients who do not respond to treatment

Nuclear Medicine Findings

- Bone scan
  - Increased uptake
- PET
  - FDG-18 PET has proven useful for staging, monitoring treatment response, predicting treatment outcomes, and risk stratifying lymphoma patients
    - High negative predictive value in diffuse large B-cell lymphoma and Hodgkin lymphoma (HL)
    - HL: Positive scan should be interpreted with other staging investigations
    - Non-Hodgkin lymphoma (NHL): Few false-positive results but negative FDG PET scans must be viewed with caution
    - Metabolic response assessment at least 3 weeks after chemo (immuno) therapy and 8–12 weeks after (chemo)radiation
- Gallium scan
  - 67Ga scintigraphy has high sensitivity and specificity for diagnosis of bone lymphoma
  - Discriminates between patients who achieve complete response and those who show no or only partial response to induction therapy

Imaging Recommendations

- Best imaging tool
  - MR + C
- Protocol advice
  - Fat-saturated T1WI
  - STIR may be helpful

DIFFERENTIAL DIAGNOSIS

DDx for Epidural Lymphoma

- Hematoma: Heterogeneous > homogeneous signal
- Abscess: Rim > solid enhancement, central low signal
- Metastasis: Epidural met without bone involvement rare

DDx for Osseous Lymphoma

- Metastasis: Destructive, ± soft tissue mass
- Eosinophilic granuloma: Vertebra plana; younger patients

DDx for Lymphomatous Meningitis

- Other neoplastic/granulomatous or infectious meningitides

DDx for Intramedullary Lymphoma

- Ependymoma: Hemorrhage, cysts common
- Astrocytoma: Multisegmental; cysts common
- Metastasis: Usually round, more sharply delineated

PATHOLOGY

General Features

- Etiology
  - Exact causes are unknown
  - CNS lymphoma may be primary or secondary (hematogenous or direct geographic extension)
  - Risk factors
    - Exposure to chemicals, such as pesticides, fertilizers, or solvents
    - Epstein-Barr virus plays role in immunocompromised
    - Infection with human T-lymphotropic virus I (HTLV-I)
    - AIDS/transplant immunocompromised patients
    - Family history of NHL, although no hereditary pattern has been established
- Genetics
  - Inherited immunodeficiency
  - Previous concomitant malignant neoplasm (e.g., leukemia) in ~ 8% of immunocompetent primary CNS lymphoma patients
- Associated abnormalities
  - Multisystem/multiorgan involvement

Staging, Grading, & Classification

- NHL: Ann Arbor staging system
  - B cell (80-85%)
    - AL: Rare systemic disease in which there is partial or complete occlusion of blood vessels by neoplastic cells of B-cell lineage
  - T cell (15-20%)
  - Leukemia-like picture with peripheral lymphocytosis and bone marrow involvement in up to 50% of children and ~ 20% of adults
    - Generally, patients with more extensive nodal involvement (especially mediastinal), fewer circulating abnormal cells, and fewer blast forms in the marrow (< 25%) = lymphoma
  - Aggressive
    - Rapidly progressive but responsive to therapy and often curable
  - Indolent
    - Slowly progressive and responsive to therapy but not curable with standard approaches
- HL: Cotswold system (modified Ann Arbor staging system)
  - Extranodal involvement infrequent

Microscopic Features

- Neoplastic lymphocytes in marrow, meninges
- Pack perivascular spaces
- AL: Intravascular neoplastic cell proliferation typically found within lumina of capillaries, venules, arterioles, and small arteries
- HL: Clonal transformation of cells of B-cell origin → pathognomic binucleated Reed-Sternberg cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most common presenting symptom = back pain
  - Intramedullary = myelopathy (weakness, numbness)
- Other signs/symptoms
  - Depressed humoral and cell-mediated immunity contributes to opportunistic infections

Demographics

- Age
  - Adults; peak = 4th-7th decade
- Gender
  - Slight male predominance
- Epidemiology
  - NHL > > Hodgkin disease (HD); 80-90% are B cell
    - CNS lymphoma > 85% NHL (B cell > > > T cell)
  - Primary epidural = 1-7% of NHL; 10-30% of epidural malignancies
  - Secondary epidural in 5% of patients with systemic lymphoma
  - Primary osseous = 3-4% of malignant bone tumors
  - Bone marrow involvement in 25-50% of NHL patients, 5-15% HD
  - Intramedullary = 1-3% of CNS lymphoma
  - Leptomeningitic nearly always occurs as spread from intracranial lymphoma

Natural History & Prognosis

- Cord compression occurs in up to 5-10% of systemic lymphomas
- Generally poor prognosis for CNS lymphoma
- Best prognosis with primary osseous lymphoma
  - 5-/10-year survival rates 91% and 87%, respectively

Treatment

- XRT ± chemotherapy
  - Markedly sensitive to chemotherapy/XRT
- ± surgery

DIAGNOSTIC CHECKLIST

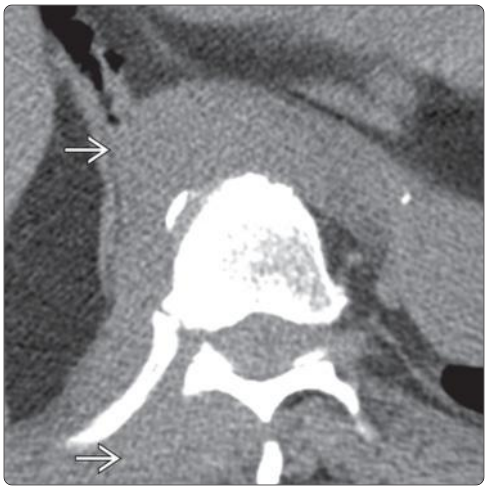
Consider

- Lymphoma has protean manifestations

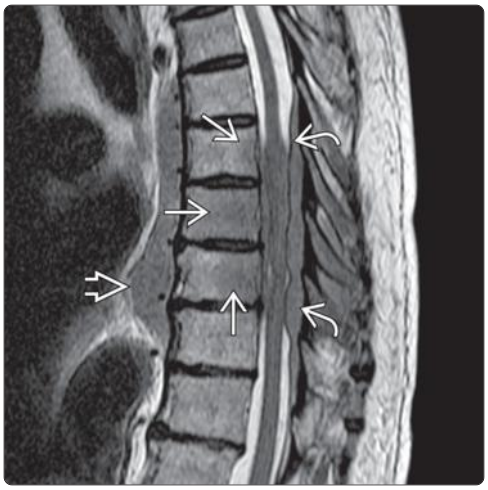
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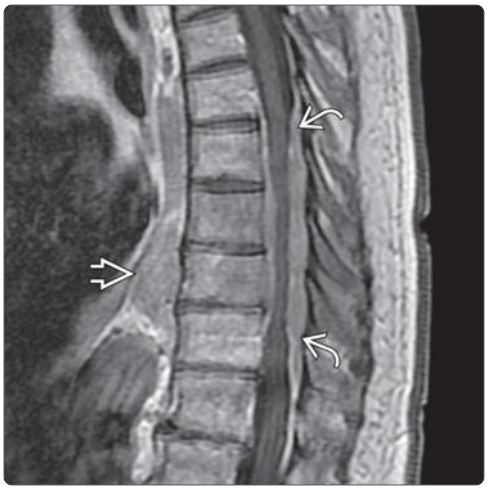
1. Mechtler LL et al: Spinal cord tumors: new views and future directions. Neurol Clin. 31(1):241-68, 2013
2. Hanrahan CJ et al: MRI of spinal bone marrow: part 2, T1-weighted imaging-based differential diagnosis. AJR Am J Roentgenol. 197(6):1309-21, 2011
3. Barajas RF Jr et al: Diffusion-weighted MR imaging derived apparent diffusion coefficient is predictive of clinical outcome in primary central nervous system lymphoma. AJNR Am J Neuroradiol. 31(1):60-6, 2010
4. Geus-Oei LF et al: Predictive and prognostic value of FDG-PET. Cancer Imaging. 8:70-80, 2008





(Left) *Sagittal NECT illustrates a single sclerotic vertebral body in this patient with lymphoma. Lymphoma represents an increasingly important cause of morbidity and mortality in older patients.* **(Right)** *Axial NECT shows an infiltrative soft tissue mass in the prevertebral space with extension along the right paravertebral region and into the right dorsal paraspinal musculature. Despite this extensive paravertebral and epidural infiltration, there is bone destruction.*





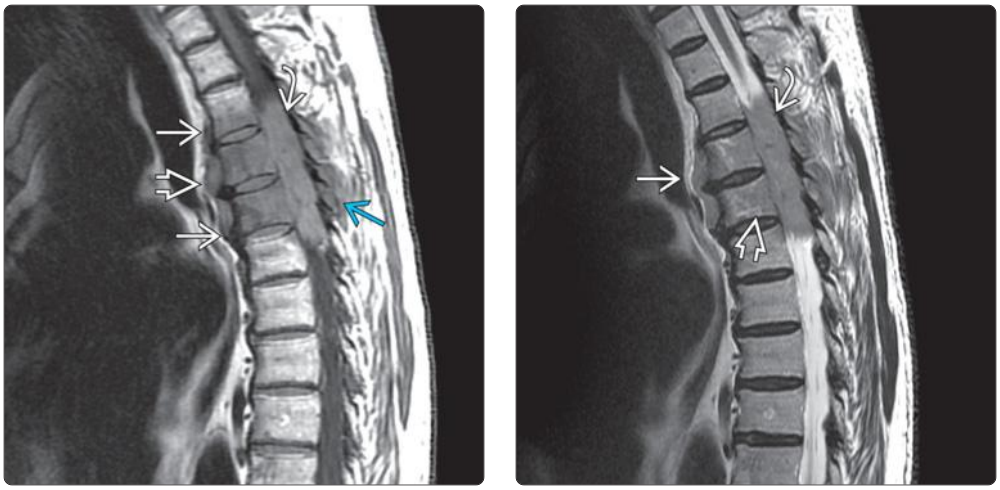
(Left) *Sagittal T2WI MR demonstrates multiple thoracic bodies with abnormally low signal, a large paravertebral, and epidural mass. The mass is typical for lymphoma given the infiltrative nature of the body involvement without bone destruction and the low T2 signal.* **(Right)** *Sagittal T1WI C+ MR reveals homogeneous enhancement of the paravertebral and epidural mass. The latter causes circumferential cord compression.*



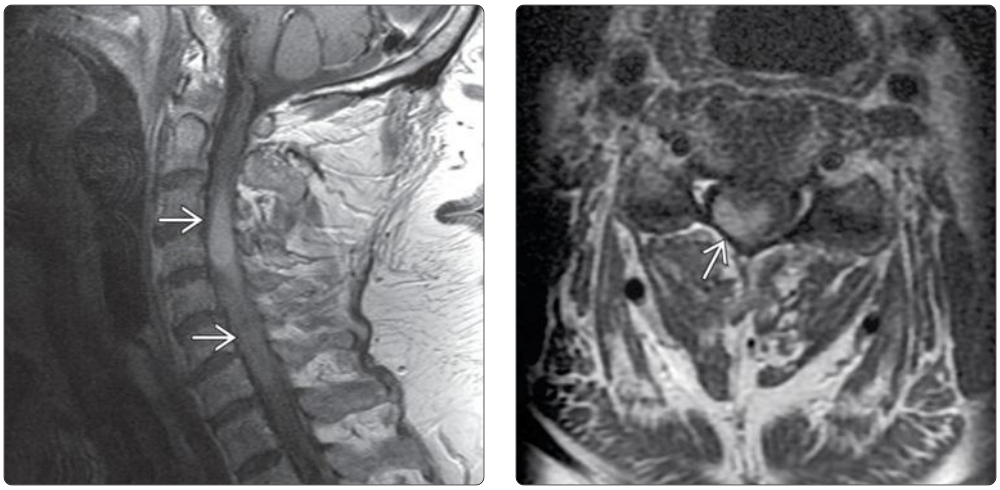


(Left) *Sagittal STIR (left) displays a hypointense posterior epidural mass, causing significant mass effect on the thecal sac and cauda equina. Sagittal T1WI C+ MR (right) shows diffuse, intense enhancement of this epidural mass. No definite bone involvement is present. Low signal T2 mass with diffuse enhancement is consistent with high cellularity tumor, such as lymphoma.* **(Right)** *Sagittal T1WI C+ MR exhibits patchy enhancement in the conus due to angiotropic large cell lymphoma.*

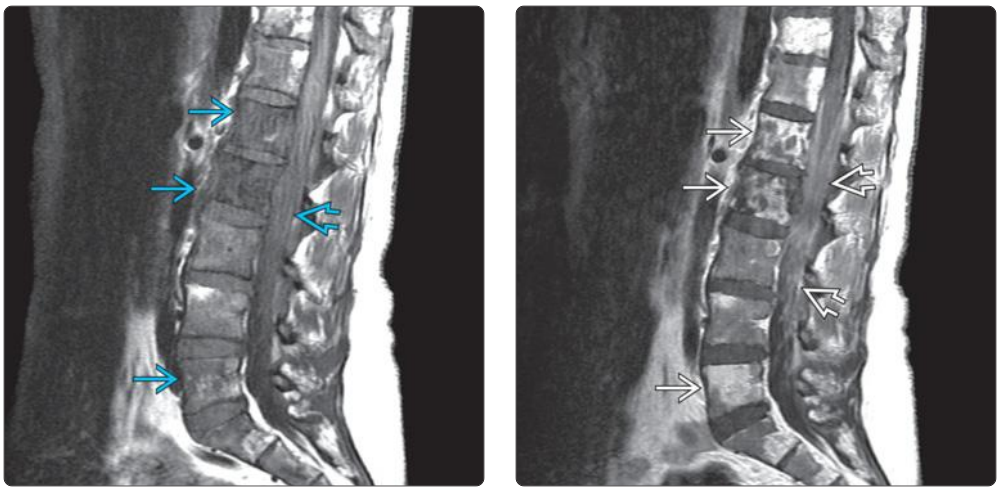
(Left) *Sagittal T1WI MR shows abnormal low signal from marrow infiltration at multiple thoracic levels, as well as prevertebral extension and a large epidural mass. The marrow filtration involves the posterior elements.* **(Right)** *Sagittal T2WI MR shows the typical low signal intensity of lymphoma involving the prevertebral space, vertebral bodies, and epidural space.*

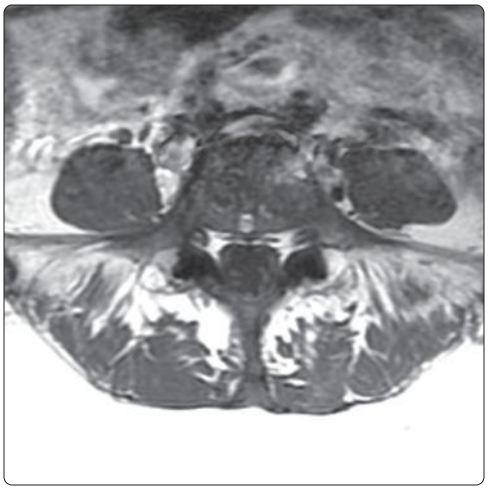


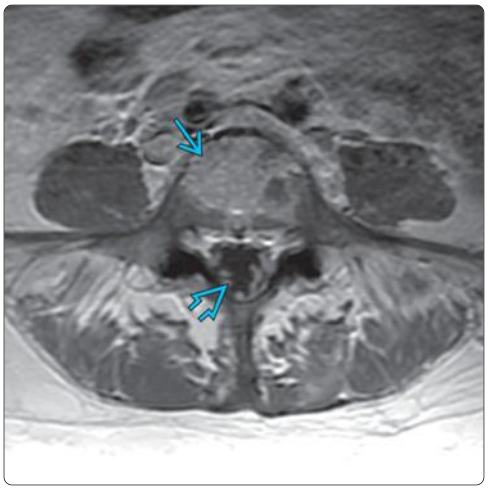
(Left) *Sagittal T1 C+ MR shows extensive and homogeneous enhancement of the cervical cord in this patient with lymphoma. Note that the cord involvement occurs without obvious leptomeningeal enhancement.* **(Right)** *Axial T1 C+ MR shows a large region of homogeneous enhancement within the cervical cord due to lymphomatous involvement.*



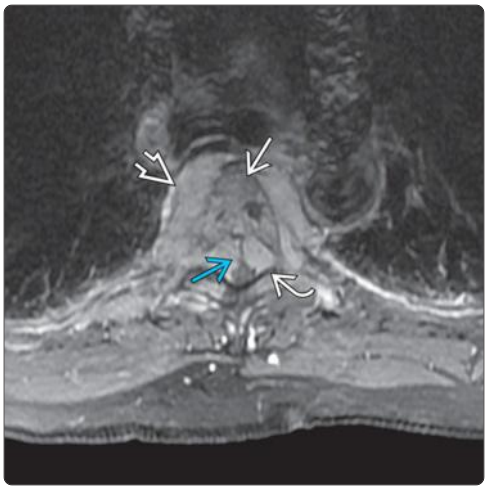
(Left) *Sagittal T1WI MR shows extensive marrow abnormality with low signal within multiple vertebral bodies from lymphoma. The cauda equina is indistinct.* **(Right)** *Sagittal T1 C+ MR shows extensive enhancement of the lymphoma involving essentially every vertebral body, as well as extensive leptomeningeal enhancement.*





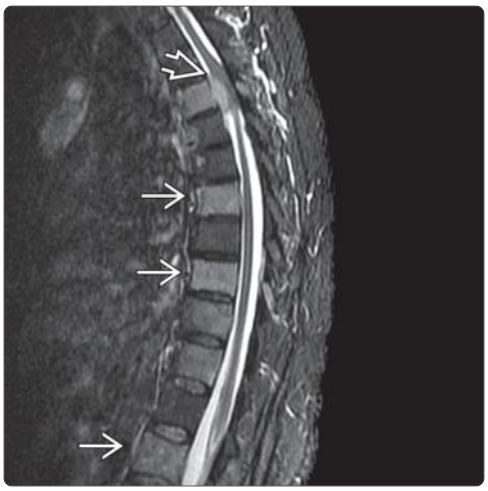


(Left) *Axial T1WI MR shows diffuse abnormal low signal intensity marrow in the lumbar vertebral body and posterior elements in this patient with lymphoma. The roots of the cauda equina are slightly prominent due to extensive leptomeningeal disease.* **(Right)** *Axial T1 C+ MR shows enhancement of the marrow involvement within the vertebral body, as well as extensive enhancement of the leptomeningeal disease involving the cauda equina.*





(Left) *Axial T1 C+ FS MR shows extensive lymphomatous involvement of the thoracic vertebral body, paravertebral soft tissues, and epidural space. Linear low signal in the midline between the compressed cord and PLL is the Hoffman ligament.* **(Right)** *Axial T1 MR shows an extensive soft tissue mass replacing the normal epidural fat signal at the sacral level in this patient with lymphoma.*





(Left) *Sagittal STIR MR shows extensive lymphoma metastatic disease involving multiple vertebral bodies, with epidural extension in the upper thoracic spine causing cord compression.* **(Right)** *Sagittal T1 C+ FS MR show heterogeneous enhancement of the metastatic involvement of the thoracic vertebral bodies. The epidural extension with cord compression shows homogeneous enhancement.*

Leukemia

KEY FACTS

TERMINOLOGY

- Acute or chronic myeloid or lymphoid white blood cell neoplasia with spinal involvement as component of systemic disease

IMAGING

- Radiographs/CT
  - Diffuse osteopenia with multiple vertebral fractures ± lytic spine lesions
  - Variable enhancing isodense soft tissue mass with adjacent bone destruction
- MR
  - T1WI: Hypointense marrow and focal tumor masses
  - T2WI: Hyperintense marrow ± focal vertebral mass, cord signal abnormality
  - T1WI C+: Abnormal enhancement of marrow, focal lesion, or leptomeninges

TOP DIFFERENTIAL DIAGNOSES

- Metastases
- Lymphoma
- Ewing sarcoma
- Langerhans cell histiocytosis

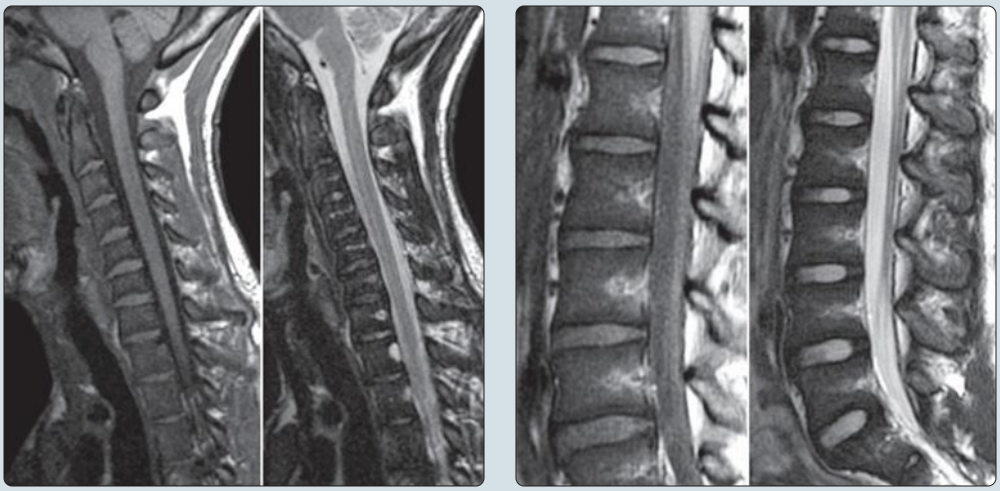
CLINICAL ISSUES

- Localized or diffuse bone pain
- Symptomatic patients present with fever, ↑ ESR, hepatosplenomegaly, lymphadenopathy, joint effusions, petechial and retinal hemorrhage, anemia, frequent infections

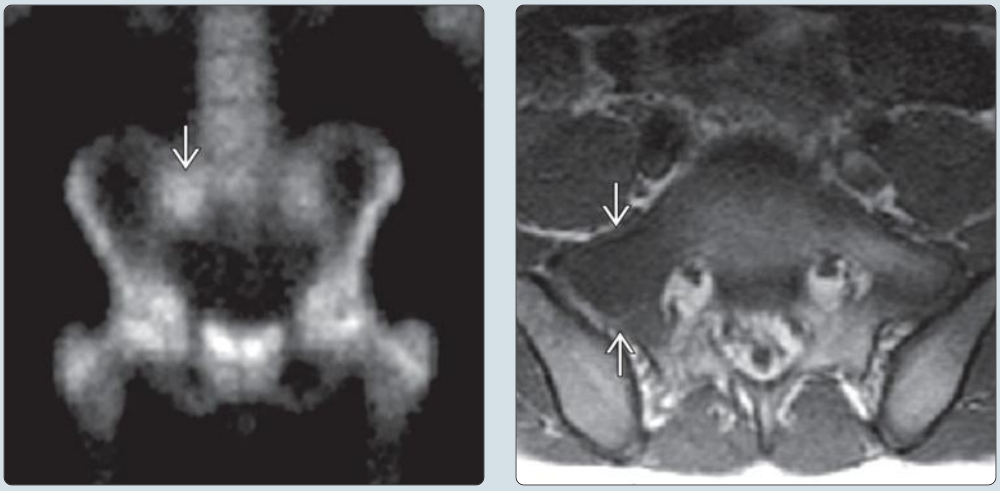
DIAGNOSTIC CHECKLIST

- Marrow infiltration in child with osteoporosis raises suspicion for leukemia
- Consider leukemia in patient with unexplained compression fractures

(Left) *Sagittal T1WI MR (left) and T2WI MR (right) of the cervical spine demonstrate diffuse abnormal hypointense signal intensity within the vertebra. Signal intensity is lower than the adjacent disc spaces.* **(Right)** *Sagittal T1WI MR (left) and T2WI MR (right) of the lumbar spine exhibit diffuse abnormal hypointense signal intensity within the vertebra as compared to the adjacent disc spaces. The signal abnormality may be a combination of tumor marrow infiltration and hyperplastic marrow secondary to anemia.*



(Left) *Coronal planar bone scan of the pelvis reveals abnormal increased uptake within the right sacral ala ➔ in this case of focal leukemic metastasis to the axial skeleton. Bone scan may underestimate disease extent, especially in absence of significant cortical destruction.* **(Right)** *Axial T1WI MR in the same patient clarifies the right sacral alar metastatic lesion ➔. There is no definite cortical breakthrough.*



TERMINOLOGY

Synonyms

- Acute lymphocytic leukemia (ALL), chronic lymphocytic leukemia (CLL), acute myelogenous leukemia (AML), chronic myelogenous leukemia (CML), granulocytic sarcoma (GS), chloroma, myeloid sarcoma (MS)

Definitions

- Acute or chronic myeloid or lymphoid white blood cell neoplasia with spinal involvement as component of systemic disease

IMAGING

General Features

- Best diagnostic clue
  - Diffuse osteopenia with multiple vertebral fractures ± lytic spine lesions
- Location
  - Children: Multiple long bones and spine (14%)
  - Adults: Predominately axial skeleton
    - Spinal GS: Multiple extramedullary masses with diffuse leukemic bone marrow infiltration
- Morphology
  - Osteopenia ± moth-eaten bone destruction of multiple vertebral bodies, leptomeningeal enhancement, focal mass ("chloroma")

Radiographic Findings

- Radiography
  - Diffuse vertebral, long bone osteopenia
    - Coarse cancellous trabeculation ± pathologic vertebral compression fractures
    - Radiographs may look normal, even if extensive disease
  - ± "leukemic lines" (horizontal vertebral bands)
  - GS: Focal lytic mass

CT Findings

- NECT
  - Isodense soft tissue mass with adjacent bone destruction
  - Leptomeningeal disease: ↑ density of subarachnoid with lumbar nerve root enlargement
  - GS: Isodense to muscle
- Bone CT
  - Permeative bone destruction ± focal lytic lesions, pathologic vertebral fractures

MR Findings

- T1WI
  - Leukemic marrow, focal tumor masses relatively hypointense
  - GS: Isointense
- T2WI
  - Increased leukemic marrow signal intensity ± focal vertebral mass, cord signal abnormality
  - GS: Hyperintense (mild to moderate)
- STIR
  - Hyperintense leukemic marrow
- T1WI C+
  - Abnormal enhancement of marrow, focal lesion, or leptomeninges
  - GS: Variable enhancement; peripheral rim enhancement may mimic abscess
  - Dynamic, contrast-enhanced MR (DCE-MR) based on time-intensity curves can be used to distinguish between highly cellular hematopoietic marrow and highly cellular neoplastic infiltration

Nuclear Medicine Findings

- Bone scan
  - ± ↑ radiotracer uptake; often underestimates disease extent, especially in absence of significant cortical destruction
- PET/CT
  - Reports of increased FDG uptake in extramedullary AML

Imaging Recommendations

- Protocol advice
  - Multiplanar T1WI, T2WI (+ fat saturation) or STIR, T1 C+ MR
  - Whole body STIR MR proposed for staging, assessing lesion burden
  - Bone CT with multiplanar reformats to delineate osseous lesions, quantitate compression fractures

DIFFERENTIAL DIAGNOSIS

Metastases

- Metastatic neuroblastoma or rhabdomyosarcoma in children, carcinomas in adults
- Multifocal bone involvement similar to leukemia

Langerhans Cell Histiocytosis (LCH)

- Lytic lesion with periosteal reaction, endosteal scalloping, soft tissue mass
- May have systemic symptoms similar to leukemia

Osteomyelitis, Pyogenic

- Bacterial suppurative infection of vertebrae and intervertebral disc
- Soft tissue extension
- May have systemic symptoms similar to leukemia

Osteomyelitis, Granulomatous

- Gibbus vertebrae with relatively intact intervertebral discs, large paraspinal abscesses

Lymphoma

- Older patient with large soft tissue mass; predilection for paraspinal, epidural locations
- Systemic lymphomatous metastasis or primary vertebral lesion

Ewing Sarcoma

- Marked periosteal reaction + soft tissue mass
- No metaphyseal lucent lines
- May have systemic symptoms similar to leukemia

PATHOLOGY

General Features

- Etiology

- External factors: Alkylating drugs, ionizing radiation, chemicals (benzene)
- Internal factors: Chromosomal abnormalities
- Predisposing hematological disorders: Aplastic anemia, chronic myeloproliferative disorders
- GS: Bone involvement may be via transhaversian canal migration of leukemia cells from bone marrow to periosteum and dura
  - CNS involvement may occur via perivascular or perineural routes from direct dural extension or through capillary migration

- Genetics
  - Strong association with chromosomal abnormalities
    - ALL: Trisomy 21, chromosomal translocations
    - CLL: Trisomy 12
    - CML: 90% have Philadelphia chromosome t(9;22)
- Associated abnormalities
  - Diagnosis of acute leukemia established by bone marrow aspirate showing at least 30% blast cells
  - Long bone periostitis (12-25%), "leukemic" metaphyseal lines
  - Focal destruction of flat/tubular bones, pathologic fractures
- Neuropathologic features include
  - Spinal manifestations of primary disease: Fractures, marrow or meningeal infiltration
  - Treatment effects (radiation therapy, chemotherapy, BMT): Secondary neoplasms (usually aggressive CNS tumors), hemorrhage, anterior lumbosacral radiculopathy (intrathecal methotrexate toxicity)
  - Immunosuppression complications: Fungal or other opportunistic infection
- GS/MS (chloroma): Extramedullary neoplasm of immature granulocytic cells → focal lytic mass
  - Most common in AML (concurrent presentation ≤ 9.1% AML cases)
  - Multiple soft tissue masses in CNS, subcutaneous tissues, and genitourinary system

Microscopic Features

- Diffuse bone marrow infiltration by poorly differentiated hematologic cells
  - ALL: Infiltrates of small blue cells
  - AML: Auer rods (condensed lysosomal, cytoplasmic, rod-shaped structures) are diagnostic
  - CML: Leukocytosis with increase in basophils, eosinophils, neutrophils; Philadelphia chromosome t(9;22)
  - CLL: Mature lymphocytes, < 55% atypical cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Localized or diffuse bone pain
  - Recurrent paraarticular arthralgias (75%)
- Other signs/symptoms
  - CSF of patients with neoplastic meningitis shows ↑ molecules involved in CNS penetration, tumor cell tropism, and angiogenesis
  - GS: Mass effect from soft tissue lesions may produce symptoms such as cauda equina syndrome
- Clinical profile
  - Chronic leukemia may be asymptomatic
  - Symptomatic patients present with fever, ↑ ESR, hepatosplenomegaly, lymphadenopathy, joint effusions, petechial and retinal hemorrhage, anemia, frequent infections

Demographics

- Age
  - ALL: Peak 2-10 years
  - AML: Peak > 65 years
  - CML: Rare in childhood (< 5%), peak > 40 years
  - CLL: Peak 50-70 years
- Gender
  - M:F = 2:1
- Epidemiology
  - Most common malignancy of childhood (ALL: 75%, AML: 15-20%, CML: 5%)
  - 20th most common cause of cancer death (all age groups)

Natural History & Prognosis

- 5-year survival (all leukemias): 25-30%
  - Children with ALL: 90% complete remission, 80% 5-year disease-free survival
  - Adults with ALL: Remission in 60-80%, 20-30% 5-year disease-free survival
  - AML: 45% 5-year survival
  - CLL: 6-year median survival
  - CML: 5-year median survival

Treatment

- Chemotherapy: Induction phase, consolidation phase, maintenance therapy phase
  - Systemic chemotherapy with cytarabine and methotrexate
  - Intrathecal chemotherapy for CNS involvement
  - ↓ bone marrow density with chemotherapy, ↑ fracture risk
- Radiation therapy
- Bone marrow transplant

DIAGNOSTIC CHECKLIST

Consider

- Marrow infiltration in child with osteoporosis raises suspicion for leukemia
- Laboratory CSF evaluation > > MR sensitivity for leptomeningeal metastases

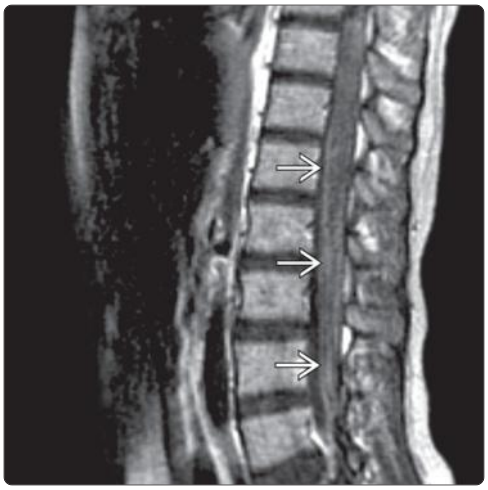
Image Interpretation Pearls

- Consider leukemia in patient with unexplained compression fractures

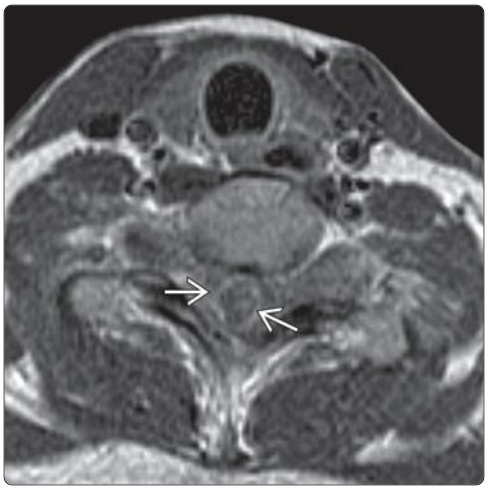
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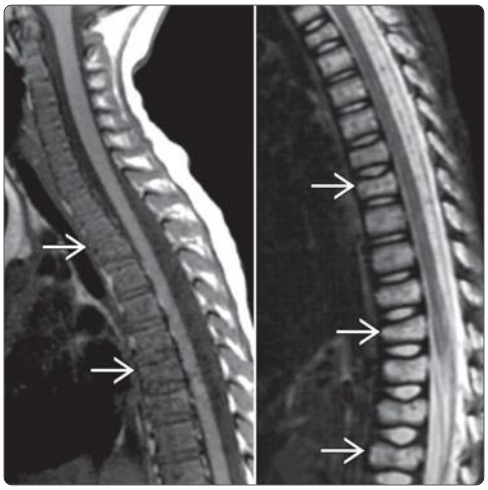
1. Chamberlain MC: Comprehensive neuraxis imaging in leptomeningeal metastasis: a retrospective case series. CNS Oncol. 2(2):121-8, 2013
2. Mäkitie O et al: Long-term skeletal consequences of childhood acute lymphoblastic leukemia in adult males: a cohort study. Eur J Endocrinol. 168(2):281-8, 2013



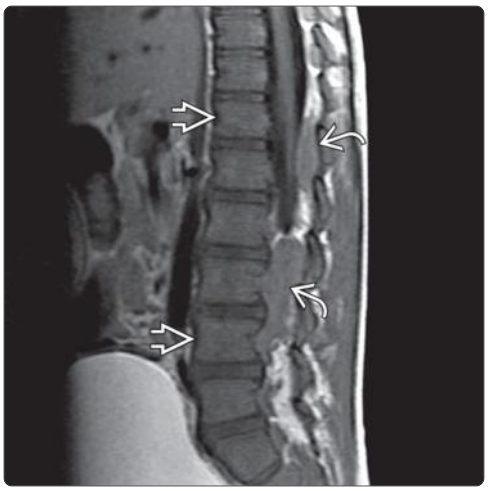


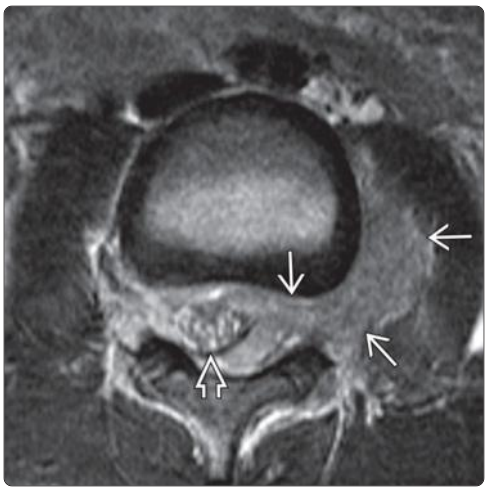
(Left) *Sagittal T2WI MR demonstrates abnormal soft tissue surrounding the cervicothoracic spinal cord. Sagittal T1WI C+ MR confirms diffuse leptomeningeal enhancement from disseminated intrathecal leukemic metastasis encircling the spinal cord.* **(Right)** *Sagittal T1WI C+ MR displays diffuse leptomeningeal enhancement from disseminated intrathecal leukemic metastasis encircling the spinal cord and infiltrating the cauda equina. The cauda equina nerve roots are also abnormally thickened.*





(Left) *Axial T1WI C+ MR reveals diffuse leptomeningeal enhancement from CSF disseminated leukemic metastasis encircling the spinal cord.* **(Right)** *Sagittal T1WI MR (left) and STIR MR (right) illustrate abnormal hypointense and hyperintense marrow signal, respectively. Numerous compression fractures are present. This represents a case of leukemic marrow infiltration precipitating initial leukemia presentation with compression fractures.*





(Left) *Sagittal T1WI MR shows diffuse abnormal hypointensity in the vertebral body marrow. There is leukemic involvement of the extradural space as exemplified by the hypointense epidural masses.* **(Right)** *Axial T2WI MR demonstrates a soft tissue mass, slightly hyperintense to muscle, infiltrating into the epidural space from the left paraspinal region. The mass causes mild mass effect on the thecal sac, which is displaced medially.*

Plasmacytoma

KEY FACTS

TERMINOLOGY

- Solitary monoclonal plasma cell tumor of bone or soft tissue
- Diagnosis of solitary bone plasmacytoma (SBP) requires
  - Solitary lesion, biopsy showing plasma cells
  - Negative skeletal survey, negative MR spine, pelvis, proximal femora/humeri
  - Negative clonal cells in marrow aspirate
  - No anemia, hypercalcemia, or renal involvement suggesting systemic myeloma

IMAGING

- Axial skeleton > extremities
  - Thoracic vertebral body most common site
- Radiographs/CT
  - Lytic, multicystic-appearing lesion ± vertical dense striations
  - Pathologic compression fracture common
- T1 hypointense, T2/STIR hyperintense marrow with low signal, curvilinear areas
  - Posterior elements involved in most cases
  - ± associated soft tissue mass (paraspinous or epidural with "draped curtain" sign)

PATHOLOGY

- SBP may reflect early (stage I) multiple myeloma (MM)
- SBPs considered clinical stage I Durie-Salmon lesions

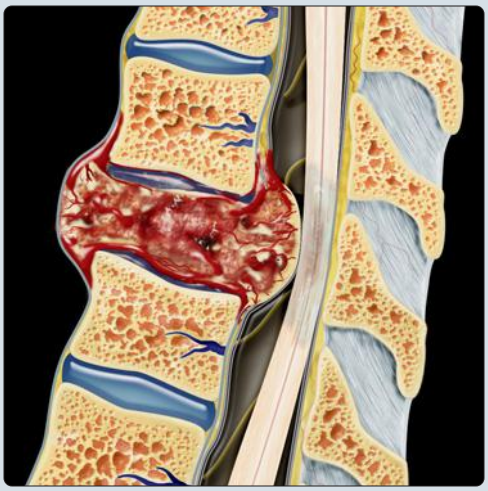
CLINICAL ISSUES

- Most common symptom = pain due to bone destruction
- Epidural extension may cause compression of cord or nerve root
- Mean age = 55 years (younger than age of patients with multiple myeloma)

DIAGNOSTIC CHECKLIST

- Must exclude 2nd unanticipated lesion (33% of cases)

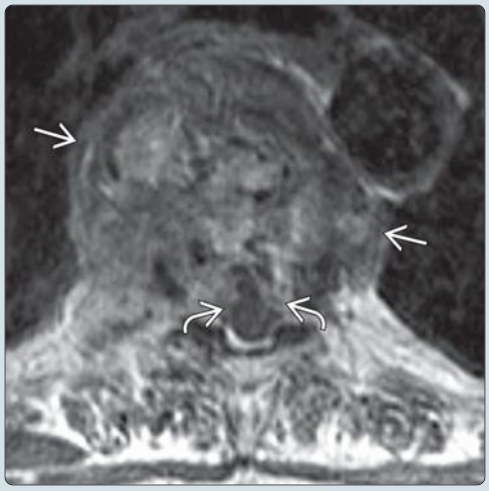
(Left) *Sagittal graphic shows collapse of a thoracic vertebral body due to tumor infiltration. There is retropulsion of the anterior and posterior margins. Posterior retropulsion and tumor mass may result in cord compression.* **(Right)** *Sagittal T2WI MR demonstrates a heterogeneously hyperintense thoracic vertebral body lesion with hypointense ventral epidural ➔ and paravertebral ➔ soft tissue components producing cord compression.*





(Left) *Sagittal bone CT exhibits the characteristic appearance of thickened cortical struts ➔, a result of stress phenomenon from the lytic process forcing remaining bone to increase thickness as a compensatory response to weakening bone.* **(Right)** *Axial T1WI C+ MR depicts tumor infiltration of a thoracic vertebral body, with extension into the paravertebral ➔ and epidural ➔ spaces. The latter significantly narrows the canal.*





TERMINOLOGY

Synonyms

- Solitary bone plasmacytoma (SBP), solitary myeloma, solitary plasma cell tumor

Definitions

- Solitary monoclonal plasma cell tumor of bone or soft tissue, without evidence of multiple myeloma (MM)

IMAGING

General Features

- Best diagnostic clue
  - T1 hypointense marrow with curvilinear hypointensities
- Location
  - Axial skeleton most common site of involvement (25-60%), followed by extremities
    - Vertebral body = most common site of SBP
    - Thoracic > lumbar > cervical

Radiographic Findings

- Radiography
  - Can be normal early
  - Lytic, multicystic-appearing lesion ± vertical dense striations
  - Pathologic compression fracture common
  - No periosteal reaction

CT Findings

- NECT
  - Common
    - Lytic, destructive vertebral body lesion
    - Compression fracture ± associated soft tissue mass
  - Uncommon: Osteosclerosis (3%)
  - Rare: Involvement of intervertebral disc, adjacent vertebrae (if present, helpful differentiating feature from metastasis)
  - Collapse of vertebral body

MR Findings

- T1WI
  - Solitary vertebral body lesion
    - Marrow iso-/hypointense (compared to muscle)
    - Contains curvilinear low signal areas ± cortical irregularities ("infoldings" caused by endplate fractures)
    - "Mini brain" appearance due to thick cortical struts in expanded vertebral body
    - Variable degrees of compression
    - Posterior elements involved in most cases
    - ± associated soft tissue mass (paraspinous or epidural with "draped curtain" sign)
  - Scanning entire spine reveals 2nd lesion in 1/3 of cases
- T2WI
  - Heterogeneous signal
    - Focal hyperintensities (compared to fat)
    - Curvilinear areas of signal void
- STIR
  - STIR: Hyperintense (corresponds to lytic lesions on NECT)
- T1WI C+
  - Common: Mild/moderate diffuse enhancement
  - Uncommon: Peripheral (rim) enhancement

Nuclear Medicine Findings

- Bone scan
  - Intense uptake but may be normal early in disease
  - Unreliable for staging and monitoring
- PET
  - FDG PET reliably detects active MM
  - Negative FDG PET supports diagnosis of monoclonal gammopathy of undetermined significance (MGUS)
- Tc-99m-MIBI
  - Increased uptake in MM, with extent and intensity correlated with disease activity

Imaging Recommendations

- Protocol advice
  - Standard MR + STIR, scan entire spine
  - CT-guided biopsy/fine needle aspiration

DIFFERENTIAL DIAGNOSIS

Multiple Myeloma

- 2nd lesion found in 33% of cases with presumed spine SBP

Metastasis

- May be indistinguishable from SBP
- Posterior element involvement not useful in differentiating from SBP
- Does not involve disc or adjacent vertebrae

Benign (Osteoporotic) Compression Fracture

- Common in older patients, including those with SBP and MM
- 50-60% of compression fractures in MM appear benign on MR
- Signal intensity (subacute/chronic fractures) like normal marrow

Vertebral Hemangioma (VH)

- Aggressive VHs may mimic SBP, metastases are rare
- Marrow signal of aggressive VH may resemble SBP
  - Most benign VHs are T1/T2 hyperintense
- Intense enhancement

Paget Disease

- Vertebral body expansion with thickened trabeculae

PATHOLOGY

General Features

- Etiology
  - SBP may reflect early (stage I) MM
  - Some patients stable for decades, suggesting relationship to MGUS
- Genetics
  - Unknown for SBP
  - In situ hybridization studies show cytogenetic abnormalities in 80-90% of MM patients
    - Chromosome 13 deletion most common (13q)
    - Other: Chromosome 14q32 deletions/translocations, 11q, miscellaneous translocations

- Monosomy 13 correlated with poor prognosis, especially combined with elevated β2 microglobulin, concomitant chromosome 11 aberration

Staging, Grading, & Classification

- SBPs considered clinical stage I Durie-Salmon lesions
  - Durie-Salmon staging based on
    - Urine and blood monoclonal immunoglobulin
    - Blood calcium levels
    - Bone destruction on x-rays
    - Blood hemoglobin
- Diagnostic criteria for SBP
  - Single area of bone damage due to clonal plasma cell hyperplasia
  - Histologically normal marrow aspirate and trephine (plasma cell < 5%)
  - Normal results on skeletal survey, including radiology of long bones
  - No anemia, hypercalcemia, or renal impairment due to plasma cell dyscrasia
  - Absent or low serum or urinary level of monoclonal immunoglobulin (> 20 g/L suspicious of MM)
  - No additional lesions on MR scan of spine
- Some variations in definitions, such as 2 bone lesions or < 10% bone marrow plasmacytosis

Gross Pathologic & Surgical Features

- Compressed vertebra with gray-purple fatty marrow replacement

Microscopic Features

- Monotonous collection of neoplastic plasma cells
- Neoplastic cells express more plasma cell antigens, including CD138, CD38, and PC

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Clinical manifestations related to anatomic sites
    - Most common symptom = pain due to bone destruction
      - Painful phase with mean of 6 months
    - Can be asymptomatic
    - Epidural extension may cause compression of cord or nerve root
    - Pathologic fracture may cause cord compression
  - Other signs/symptoms
    - Low levels of serum/urine monoclonal proteins (25-75%)
    - Uncommon presentation of demyelinating polyneuropathy
      - Consider POEMS (**p**olyneuropathy, **o**rganomegaly, **e**ndocrinopathy, **M** protein, **s**kin changes) syndrome

Demographics

- Age
  - Mean age = 55 years (younger than patients with MM)
- Gender
  - M > F (2:1)
- Epidemiology
  - Solitary bone plasmacytic lesions represent 3-5% of monoclonal gammopathies
  - Spine is most common site
  - 20% have solitary lesions in rib, sternum, clavicle, scapula

Natural History & Prognosis

- SBPs typically have indolent course (median overall survival period of 7.5-12 years)
  - 5-year survival rate ~ 70%
- Common progression patterns include new bone lesions, increased myeloma protein levels, marrow plasmacytosis
- When MM evolves from SBP, tend to have low tumor mass disease with high chemotherapy response rate
- Prognostic factors associated with ↑ relapse rates
  - Age > 60
  - Extraaxial skeletal lesions location
  - Tumors > 5 cm
  - Presence of M protein at time of diagnosis not shown to have prognostic value

Treatment

- Definitive local radiotherapy (40-60 Gy)
  - ± supported by autologous stem cell transplantation
- Surgical intervention is preferably indicated for patients with neurologic involvement and tumor-induced spinal instability
- Studies show gross total tumor resection or total spondylectomy with adjuvant radiotherapy can ↓ local recurrences and ↓ possibility of progressing to MM
- Some asymptomatic patients with stage I MM not treated until more aggressive disease demonstrated at clinical follow-up
- Bisphosphonate treatment ~ 2 years can effectively prevent osteolysis or bone loss

DIAGNOSTIC CHECKLIST

Consider

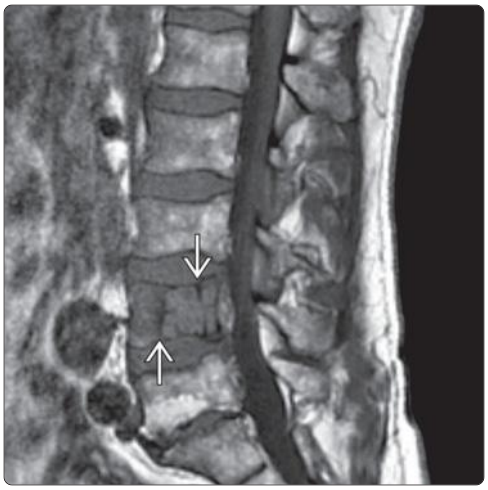
- Must exclude 2nd unanticipated lesion (33% of cases)
- Diagnosis of SBP can only be established after exclusion of extramedullary invasion by MM

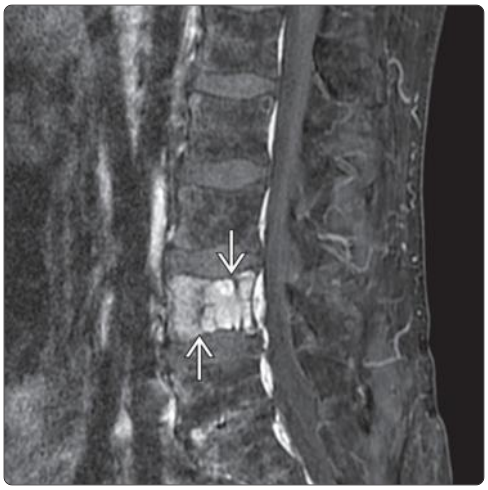
Image Interpretation Pearls

- Classic imaging appearance = hypointense vertebra (T1WI) with cortical "infoldings," curvilinear low signal areas

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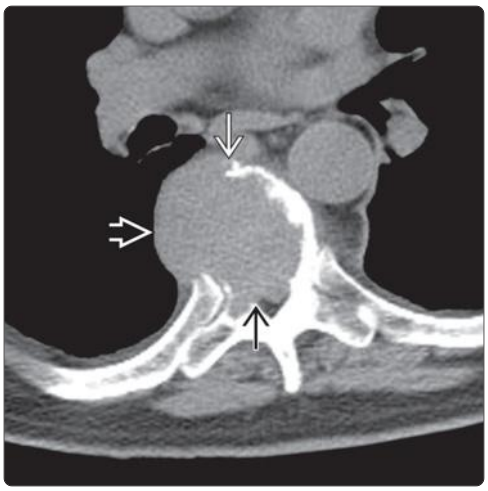


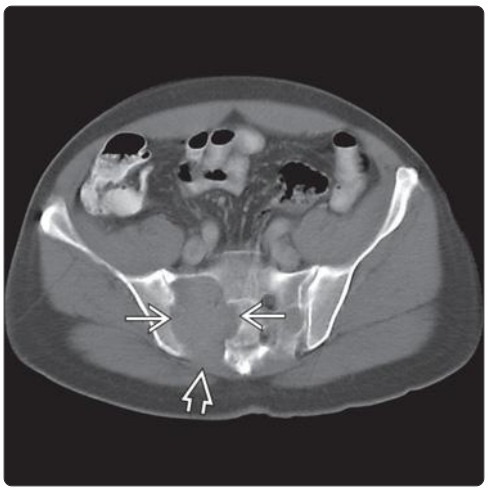
(Left) *Sagittal T1WI MR illustrates the "mini brain" appearance of solitary bone plasmacytoma (SBP). Curvilinear low signal intensity structures extend partially through the vertebral body and resemble sulci seen in the brain. These are the thick cortical struts seen in SBP with bone CT.* **(Right)** *Sagittal STIR MR of the lumbar spine reveals diffuse hyperintensity of the infiltrated L4 vertebral body with internal thick cortical bone struts.*





(Left) *Axial T1WI MR demonstrates a lobulated tumor mass in a lumbar vertebral body with posterior extension into the ventral epidural space. This mildly narrows the spinal canal and left lateral recess.* **(Right)** *Coronal STIR MR of the thoracic spine demonstrates hyperintense lesions involving 3 contiguous vertebral bodies with an associated soft tissue mass extending in the right paravertebral region.*





(Left) *Axial NECT depicts a lytic neoplastic lesion involving a midthoracic vertebral body. The lesion also extends into the right paraspinal region. Epidural extension displaces the thecal sac posterolaterally with significant mass effect.* **(Right)** *Axial bone CT shows a lytic, expansile lesion involving the right aspect of the sacrum. The lesion is sharply demarcated with a narrow zone of transition. However, there is posterior cortical breakthrough, which indicates intermediate lesion aggressivity.*

KEY FACTS

TERMINOLOGY

- Multifocal malignant proliferation of monoclonal plasma cells within bone marrow

IMAGING

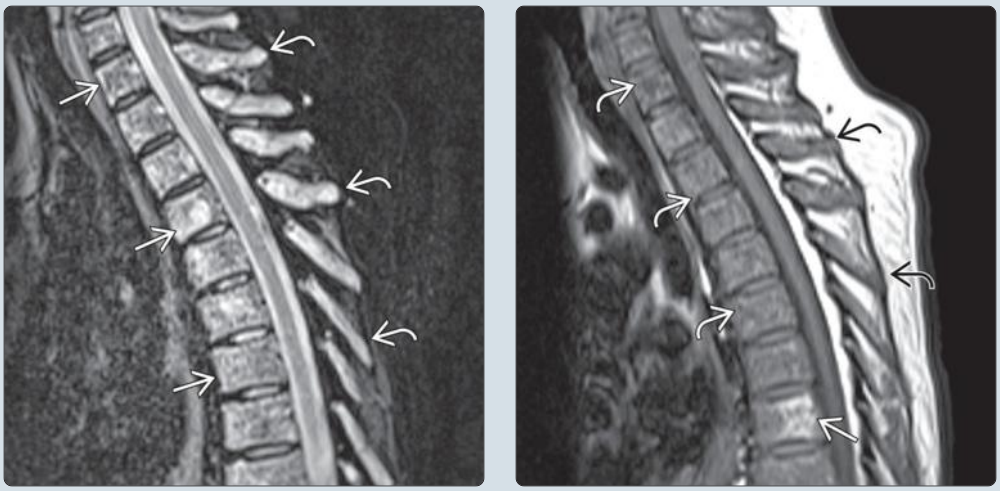
- Skeletal survey is initial diagnostic imaging evaluation
  - Diffuse osteopenia and multiple lytic lesions
- NECT (bone algorithm)
  - Multifocal lytic lesions
  - Vertebral destruction and fractures
- MR patterns
  - Normal
  - Focal marrow involvement
  - Diffuse marrow involvement
  - Variegated pattern (micronodular, "salt and pepper")
- Compression fractures with variable central canal narrowing
- FDG PET
  - Identifies active MM; useful in monitoring treatment response
- FSE T2 with fat saturation, STIR, or T1WI C+ with fat suppression increase lesion conspicuity

TOP DIFFERENTIAL DIAGNOSES

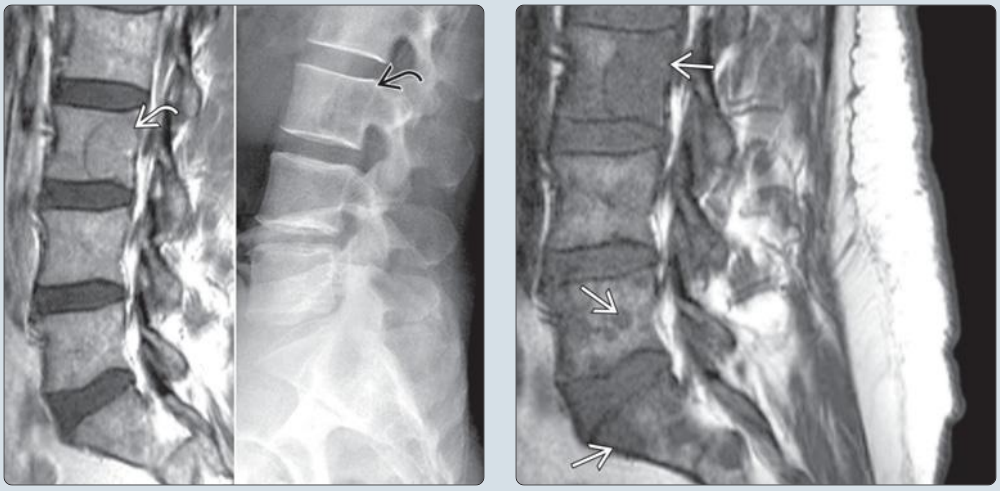
- Metastases
- Leukemia/lymphoma
- Osteoporosis

CLINICAL ISSUES

- Bone pain: 75%
- Marrow failure: Anemia, infection
- Renal insufficiency/failure
- M protein (monoclonal immunoglobulin): Blood ± urine
- Hypercalcemia



(Left) *Sagittal STIR MR illustrates innumerable hyperintense foci in the cervical and thoracic vertebral bodies and spinous processes. Advanced imaging is recommended in those with normal radiographs and monoclonal gammopathy or a solitary plasmacytoma.* **(Right)** *Sagittal T1WI MR demonstrates heterogeneous marrow signal due to countless hypointense lesions in the cervical and thoracic vertebral bodies and the spinous processes. There is a benign vertebral hemangioma at T8.*



(Left) *Sagittal T2WI MR (left) shows marrow heterogeneity with a hypointense L3 vertebral body lesion, barely detectable on lateral MR (right). Radiographs allow identification of only those lesions with advanced destruction affecting at minimum 30% of trabecular bone.* **(Right)** *Sagittal T1WI MR depicts fatty marrow replacement by several hypointense vertebral body lesions of varying sizes. Post-treatment T1WI MR shows gradual replacement of infiltrates by red marrow, then by fat.*

TERMINOLOGY

Abbreviations

- Multiple myeloma (MM)

Definitions

- Heterogeneous group of plasma cell neoplasms involving primarily bone marrow ± soft tissues

IMAGING

General Features

- Best diagnostic clue
  - Multifocal, diffuse, or heterogeneous T1 hypointensity
- Location
  - Axial skeleton (red marrow) > long bones
    - Spine, skull (mandible), ribs, pelvis
  - 87% vertebral fractures between T6 and L4

Radiographic Findings

- Radiography
  - Skeletal survey (SS) is initial diagnostic imaging evaluation
    - Identifies those lesions with at least 30% destruction
  - Diffuse osteopenia: 85%
  - Multiple lytic lesions: 80%
    - Approximately 1% of lesions sclerotic
  - Endosteal scalloping
  - Soft tissue mass adjacent to bone destruction
  - Vertebral compression fractures

CT Findings

- NECT (bone window)
  - Multifocal lytic lesions
  - Vertebral destruction and fractures
  - CT with sagittal and coronal reformation
    - Cortical disruption, extraosseous soft tissue component, potential instability better evaluated

MR Findings

- Normal
- Focal marrow involvement
  - Low to intermediate signal intensity (SI) compared to marrow on T1WI
  - Hyperintense on T2WI or STIR
  - Post-gadolinium enhancement
- Diffuse marrow involvement
  - Fatty marrow replaced by low SI on T1WI
    - Iso- or hypointense to intervertebral discs
  - Diffuse marrow enhancement
- Variegated pattern (micronodular, "salt and pepper")
  - Patchy, heterogeneous, or mottled T1 SI
  - Heterogeneous post-gadolinium enhancement
- Compression fractures → variable central canal narrowing
  - 67% of vertebral fractures appear benign
  - Cortical breakthrough → epidural extension, possible cord compression
- Patterns of response
  - T1WI: Gradual replacement of infiltrates by red marrow, then by fat
  - STIR: Uniform ↑ SI or rim of ↑ SI with healing
  - T1WI C+: Diffuse enhancement → active MM
    - Peripheral enhancement → ± viable tumor cells
  - Focal MM lesions may persist up to 58 months

Nuclear Medicine Findings

- Bone scan
  - Typically negative → MM inhibits osteoblastic activity
    - Bone scintigraphy detects 10% of lesions
  - Photopenic areas
- PET
  - MM lesions metabolically active
    - Sensitivity: 84-92%
    - Specificity: 83-100%
  - Identifies active MM; useful in monitoring treatment response
    - More accurate than MR for detecting extramedullary disease
  - Concordant PET and whole-body MR findings have specificity and positive predictive value of 100%

Imaging Recommendations

- Best imaging tool
  - MR and FDG PET/CT more sensitive than plain films or bone scintigraphy
    - Provide complimentary information for staging and post-treatment monitoring

DIFFERENTIAL DIAGNOSIS

Metastases

- Pedicles often involved earlier in metastases
- Increased radiotracer uptake on bone scan

Leukemia/Lymphoma

- Diffuse marrow signal abnormality and enhancement

Osteoporosis

- No discrete lesions
- Difficult to distinguish from diffuse MM marrow involvement on MR

Hyperplastic Marrow

- Diffuse or patchy T1 hypointensity
- No discrete lesions

PATHOLOGY

General Features

- Etiology
  - Largely unknown
  - Risk factors
    - Ionizing radiation
    - Exposure to pesticide, herbicide, dioxin
    - HIV, human herpesvirus 8
  - Cell of origin: Memory B lymphocyte
- Genetics
  - Favorable prognosis
    - Upregulation of cell cycle proteins, hyperdiploidy
  - Poor prognosis
    - Certain translocations, hypodiploidy, loss of *P53* tumor suppressor gene, ↑ plasma cell proliferation, high serum β_2-microglobulin

Staging, Grading, & Classification

- Durie-Salmon staging system: Based on Hgb and serum calcium levels, M (monoclonal) protein burden, and bone involvement on radiographs
  - Stage I: Low tumor mass
    - Normal and variegated marrow on MR
  - Stage II: Intermediate tumor mass
    - Focal and diffuse pattern on MR
  - Stage III: High tumor mass
    - Focal and diffuse pattern on MR
  - Each stage divided into A (serum creatinine < 2 mg/dl) or B (serum creatinine > 2 mg/dl)
  - Adapted to incorporate x-sectional imaging: "Durie and Salmon PLUS"
    - Number of focal lesions 5 mm or more
    - Extent of diffuse bone marrow disease on MR or FDG PET/CT

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Bone pain: 75%
  - Pathologic fractures
  - Marrow failure: Anemia, infection
  - Renal insufficiency/failure
  - M protein (monoclonal immunoglobulin): Blood ± urine
  - Amyloidosis (10%)
  - Hypercalcemia
- Clinical profile
  - 20% asymptomatic at presentation
  - Excessive production of nonfunctional M protein: 60% IgG, 20% IgA, 20% free light chain
  - Lab abnormalities indicate residual disease even without imaging findings

Demographics

- Age
  - Peak incidence from 40s-80s
- Gender
  - M:F = 3:2
- Ethnicity
  - 2x more common in African Americans than Caucasians
- Epidemiology
  - Most common primary tumor of bone
    - 10% of hematologic malignancies

Natural History & Prognosis

- Median survival rate of 8.5 years with high-dose chemotherapy and peripheral stem cell transplantation
  - 5% complete remission
- Serum level of M protein and percentage of bone marrow plasma cells stratify risk of progression
  - Monoclonal gammopathy of undetermined significance (MGUS)
    - 1% per year risk of progression to MM
  - Monoclonal gammopathy of borderline significance (MGBS)
    - 10-30% proportion of bone marrow plasma cells (as compared to ≤ 10% for MGUS)
  - Smoldering MM (SMM): Absence of disease-related end-organ damage
    - ≥ 3g/dL
  - Symptomatic MM : Requires therapy
  - Other
    - Plasma cell leukemia
      - More aggressive form, ≥ 20% proportion of circulating plasma cells
    - Nonsecretory MM
      - Meets bone marrow criteria for MM but **no** elevation of M protein
    - POEMS syndrome: **P**olyneuropathy, **O**rganomegaly, **E**ndocrine disorders, **M**onoclonal gammopathy, **S**kin changes
    - Plasmacytoma
      - Better prognosis → some cases cured with radiation therapy
      - Many cases progress to MM
- Factors portending poor prognosis
  - Low hemoglobin
  - Hypercalcemia
  - Extensive lytic lesions
  - High immunoglobulin production rate
  - Impaired renal function

Treatment

- Supportive care
- Local radiation
- Chemotherapy
- Transplants
  - Autologous hematopoietic stem cell: Patients usually < 65 years old with newly diagnosed MM
  - Allogeneic bone marrow: Treatment-related mortality up to 50%

DIAGNOSTIC CHECKLIST

Consider

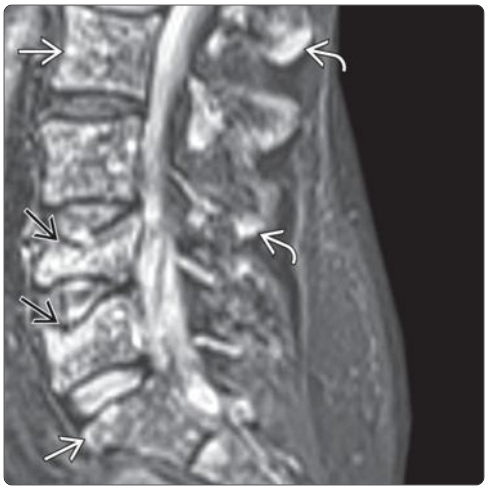
- MM when multifocal, diffuse, or heterogeneous T1 marrow hypointensity present
- Whole-body MR has higher sensitivity and specificity than radiographic SS
  - Recommended for solitary plasmacytoma or monoclonal gammopathy and normal radiographs or < 5 lesions
- PET identifies active lesions

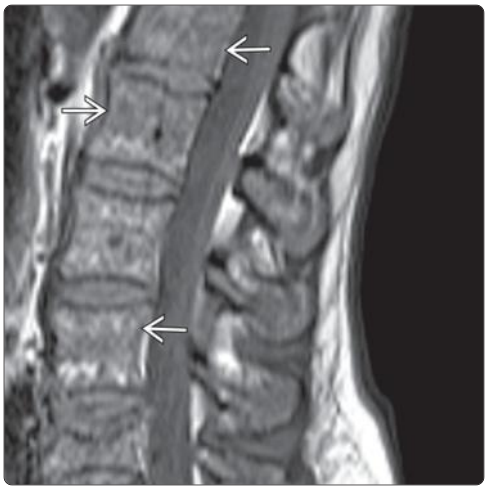
Image Interpretation Pearls

- Lytic lesions on CT almost always persist despite successful treatment

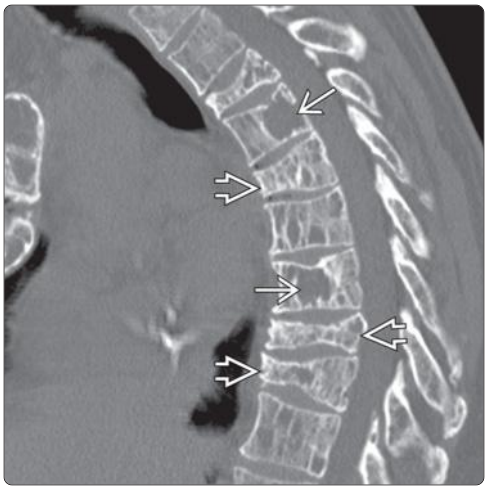
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(Left) *Sagittal STIR MR shows marrow heterogeneity that is due to numerous lesions in the anterior ⇨ and posterior ⇨ elements of the lumbosacral spine. Most compression fractures ⇨ in MM have a benign appearance. There may be abnormal patterns of marrow infiltration including focal, diffuse, and micronodular.* **(Right)** *The multitude of lesions are difficult to appreciate individually on this sagittal T1WI MR. The "salt and pepper" appearance ⇨ of the marrow is suspicious for an infiltrative process.*





(Left) *Sagittal CT shows diffuse osteopenia with multiple vertebral body lytic lesions ⇨. Pathologic compression fractures ⇨ are seen in multiple midthoracic vertebral bodies. Compression fractures can cause variable central canal narrowing.* **(Right)** *Sagittal PET illustrates mild heterogeneity of the spine bone marrow ⇨. The spine has a background of mottled diffuse activity and a large hypermetabolic area at L3 ⇨. False-negative findings may occur in lesions smaller than 10 mm.*





(Left) *Sagittal T1WI MR demonstrates diffuse thoracic spine heterogeneity due to marrow infiltration. The large focal lesions in T2 ⇨ and the posterior T6 body ⇨ cause slight bowing of the posterior convexity. Cortical breakthrough with epidural extension may result in cord compression.* **(Right)** *Sagittal STIR MR increases the conspicuity of the variegated marrow. Hyperintense foci are noted in spinous processes ⇨. Larger lesions are seen in the T2 and T6 vertebral bodies ⇨.*

Neuroblastic Tumor

KEY FACTS

TERMINOLOGY

- Neuroblastoma (NB), ganglioneuroblastoma (GNB), and ganglioneuroma (GN) are tumors of varying maturity derived from primordial neural crest cells that form sympathetic nervous system

IMAGING

- Abdominal (adrenal, paraspinal ganglia) > thoracic > pelvic > cervical
- Radiographs
  - Widened paraspinal soft tissues ± scoliosis
  - ± stippled abdominal or mediastinal calcifications
- CT
  - Widened neural foramina & intercostal spaces, pedicle erosion, adjacent rib splaying (GN, GNB) or destruction (NB)
- MR for diagnosis, presurgical planning
  - T1WI: Hypo-/isointense paraspinal mass
  - T2WI: Hypo-/hyperintense paraspinal mass
  - ± epidural extension through neural foramina
  - Variable enhancement ± internal hemorrhage, necrosis
- MIBG for NB staging, post-treatment surveillance

TOP DIFFERENTIAL DIAGNOSES

- Ewing sarcoma
- Vertebral metastasis
- Lymphoma

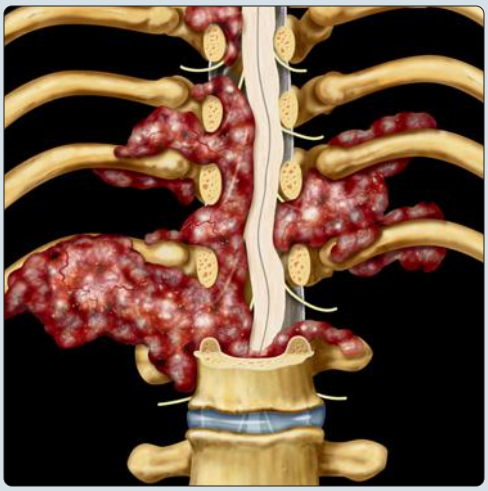
CLINICAL ISSUES

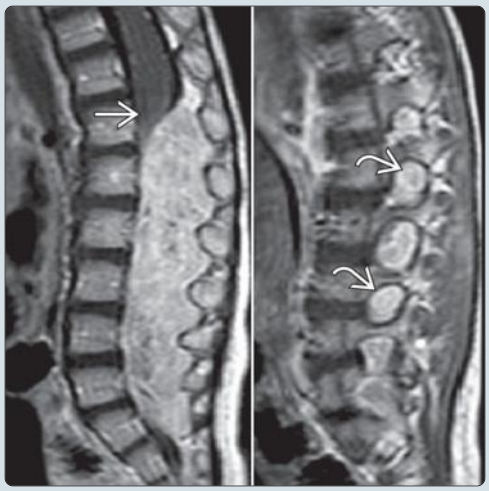
- Abdominal mass/pain, bone pain, fatigue, weight loss, blanching subcutaneous nodules
- Paraparesis/paraplegia (cord compression)

DIAGNOSTIC CHECKLIST

- Critical to determine whether tumor extends into spinal canal or neural foramina

(Left) *Coronal graphic depicts a vascular paraspinal neuroblastoma (NB) originating on the right, with spread through contiguous neural foramina across midline to the left (stage 3).* **(Right)** *Sagittal T1 C+ MR illustrate a large mass involving the entire lumbar epidural space with marked compression of the thecal sac and compression of the conus medullaris. This NB mass enhances avidly and homogeneously. The mass extends from the retroperitoneum through multiple lumbar foramen.*





(Left) *Axial STIR MR displays a paraspinal NB transgressing across the neural foramen into the epidural space and displacing the thecal sac medially. Report whether the tumor extends into the spinal canal or neural foramina, as epidural extension complicates the surgical management.* **(Right)** *Sagittal T1 C+ FS MR shows an avidly enhancing posterosuperior mediastinal mass. This GNB directly contacts adjacent vertebral bodies and ribs without abnormal marrow enhancement to imply invasion.*





TERMINOLOGY

Definitions

- Neuroblastic tumors = ganglioneuroma (GN), ganglioneuroblastoma (GNB), and neuroblastoma (NB)
  - Embryonal tumors derived from neural crest cells

IMAGING

General Features

- Best diagnostic clue
  - Abdominal or thoracic paraspinal mass ± intraspinal extension, calcification
- Location
  - Adrenal (40%) > paraspinal ganglia (25%) > thoracic (15%) > pelvic (5%) > cervical (3%); miscellaneous (12%)
- Size
  - Variable: 1-10 cm diameter
- Morphology
  - Marrow replacement, "dumbbell" paraspinal-intraspinal tumor

Radiographic Findings

- Radiography
  - Widened paraspinal soft tissues ± scoliosis
    - Rigid, progressive curve & atypical left curve in skeletally immature adolescent should undergo further investigation
  - ± stippled abdominal or mediastinal calcifications

CT Findings

- CECT
  - Enhancing paraspinal mass ± epidural extension, finely stippled calcifications
- Bone CT
  - Widened neural foramina & intercostal spaces, pedicle erosion, adjacent rib splaying (GN, GNB) or destruction (NB)

MR Findings

- T1WI
  - Hypo-/isointense paraspinal mass ± epidural extension through neural foramina
  - ± hypointense marrow replacement
- T2WI
  - Hypo-/hyperintense paraspinal mass ± epidural extension, spinal cord compression
- DWI
  - Significant difference of ADC of NB compared to ADC of GN/GNB
    - No GN/GNB with ADC < 1.1 x 10^{-3} mm^2/s
- T1WI C+
  - Variable enhancement ± internal hemorrhage, necrosis

Nuclear Medicine Findings

- Bone scan
  - Tc-99m MDP uptake in osseous metastatic lesions
- PET
  - Avid FDG uptake
- MIBG (metaiodobenzylguanidine)
  - Uptake by sympathetic catecholaminergic cells
    - I-123 MIBG for NB staging, post-therapy surveillance
    - I-123 MIBG shows early therapeutic promise for NB treatment

Imaging Recommendations

- Best imaging tool
  - MR imaging for diagnosis, presurgical planning
  - MIBG for staging, post-treatment surveillance
- Protocol advice
  - Multiplanar enhanced MR for tumor evaluation
  - Bone CT with multiplanar reformats to evaluate bone disease, detect calcifications

DIFFERENTIAL DIAGNOSIS

Ewing Sarcoma

- "Small round blue cell tumor"; relatively T2 hypointense
- Arises from adjacent flat bones (rib, chest wall, pelvis) → 2° to vertebral invasion

Vertebral Metastasis

- Variable signal intensity; imaging characteristics follow primary tumor
- Multifocal disease common

Nerve Sheath Tumor

- Contiguous with neural foramen ± "dumbbell" configuration

Lymphoma

- Systemic metastasis or primary vertebral lesion; predilection for paraspinal, epidural locations

Wilms Tumor

- Majority arise from renal parenchyma; distinctive histopathology, slightly older patient age facilitate distinction from NB

PATHOLOGY

General Features

- Etiology
  - No specific environmental exposure or risk factors definitively identified
- Genetics
  - 1p chromosomal deletion in 70-80% of NB patients
  - Myc-N (proto-oncogene chromosome 2p) amplification → rapid tumor progression, poor outcome
  - Gain on chromosome arm 17q linked with advanced stage tumors
  - HER2/neu oncogene overexpression is unfavorable prognostic indicator
  - ↑ CD44 (glycoprotein on surface of NB cells) & expression of TRK-A (nerve growth factor) correlate with better prognosis
- Associated abnormalities
  - Orbit/skull/mandible osseous/dural metastases, "hair on end" periostitis, primary brain neuroblastoma (PNET)
- Arise from primordial neural crest cell derivatives along sympathetic chain
  - Ganglioneuroma = most benign, differentiated stromal and cellular components, mature ganglion cells (100%)

- Ganglioneuroblastoma = intermediate malignant potential, varying proportions of neuroblastoma and mature ganglion cells (> 50%)
- Neuroblastoma = malignant "small round blue cell tumor," < 50% differentiated elements

Staging, Grading, & Classification

- Evans anatomic staging for NB (overall % survival)
  - Locoregional (combines stages 1-3)
    - Stage 1: Confined to organ of interest (90%)
    - Stage 2: Extension beyond organ but not crossing midline (75%)
    - Stage 3: Extension crossing midline (include vertebral column) (30%)
  - Stage 4: Systemic, widespread distal metastases (10%)
  - Stage 4S: < 1 year at diagnosis; metastatic disease confined to skin, liver, and bone marrow; may spontaneously regress (nearly 100% survival)
- International Neuroblastoma Staging System
  - Considers surgical resectability, radiologic findings, and lymph node & bone marrow involvement

Gross Pathologic & Surgical Features

- GN, GNB: Firm, gray-white nodules
- NB: Soft, gray-tan nodules ± hemorrhage, necrosis, calcification

Microscopic Features

- GN: Mature ganglion cells, Schwann cells, neuritic processes
- GNB: Internal spectrum ranging from GN → NB
- NB: Undifferentiated neuroblasts, ganglion cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Abdominal mass/pain, bone pain, fatigue, weight loss, blanching subcutaneous nodules
  - Paraparesis/paraplegia (cord compression)
- Other signs/symptoms
  - Diarrhea due to vasoactive intestinal peptide (VIP)
  - Proptosis, periorbital/conjunctival ecchymoses ("raccoon eyes")
  - Opsoclonus-myoclonus-ataxia (OMA) paraneoplastic syndrome (2-3%)
  - Horner syndrome (cervical NB)
  - Cauda equina syndrome
  - Children with malignancies have higher proportion of abnormal ribs (18%)
    - Homeobox genes, involved in vertebral and rib sequencing, are abnormally expressed in many different malignancies
  - Laboratory findings
    - > 90% ↑ urine homovanillic acid (HVA) ± vanillylmandelic acid (VMA)
      - HVA: Metabolite of dopamine, elevated in more mature NBs and GNBs
      - VMA: Less mature metabolite of epinephrine and norepinephrine
      - VMA to HVA ratio: Indicator of maturity; ratio < 1 favorable; ratio > 1 poor prognosis
- Clinical profile
  - Kerner-Morrison syndrome: Intractable secretory diarrhea 2° to VIP secretion (GN, GNB > NB)
  - Pepper syndrome: Infant with overwhelming liver metastatic NB → respiratory compromise
  - "Blueberry muffin" baby: Infant with subcutaneous metastatic NB
  - Hutchinson syndrome: Widespread bone metastasis → bone pain, limping, pathologic fractures
  - Nonaccidental trauma mimic: Metastatic retrobulbar neuroblastoma → rapidly progressive painless proptosis, periorbital ecchymosis

Demographics

- Age
  - 40% < 1 year, 35% 1-2 years, and 25% > 2 years at diagnosis; rare after age 10 years
- Gender
  - M > F (1.3:1)
- Epidemiology
  - 7-10 new NB cases/1,000,000 children (USA); true incidence of GNB, GN unknown because many asymptomatic

Natural History & Prognosis

- GN prognosis excellent after surgical resection
- GNB prognosis dependent on proportion of GN, NB
- NB 5-year survival ~ 83% for infants, 55% for children 1-5 years, and 40% for children > 5 years
  - Favorable: Locoregional, stage 4S, ↓ n-myc amplification, hyperdiploid DNA
  - Unfavorable: Stage 4 disease, ocular & CNS involvement, ↑ serum levels of neuron-specific enolase & ferritin, ↑ urine VMA/HVA

Treatment

- Chemotherapy, surgery, radiation, steroids
- No differences in neurologic improvement in patients with spinal cord compression treated with radiotherapy, laminectomy, or chemotherapy alone

DIAGNOSTIC CHECKLIST

Consider

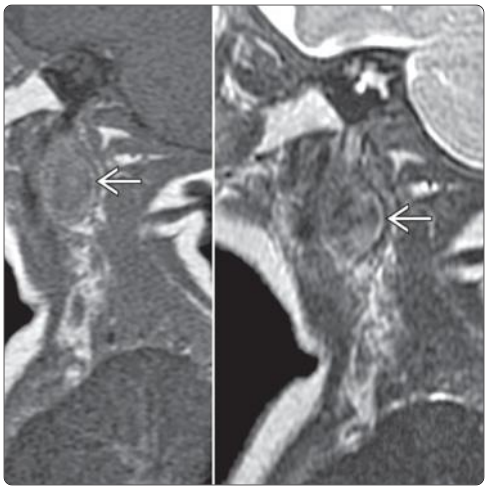
- NB presentation depends on patient age, primary site, metastatic burden, metabolically active products
- Infants more often present with thoracic, cervical tumors; older children with abdominal tumors

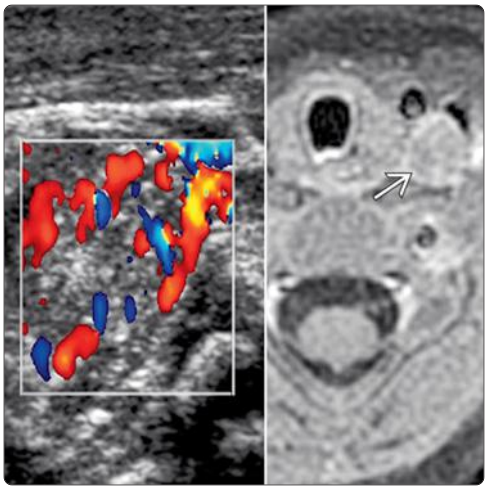
Reporting Tips

- Critical to determine preoperatively whether tumor extends into spinal canal or neural foramina, as epidural extension complicates surgical management

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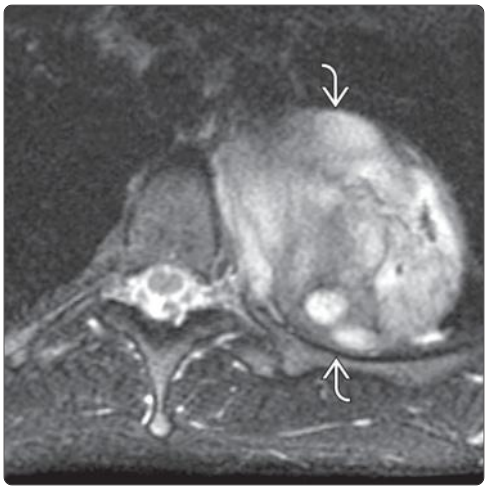
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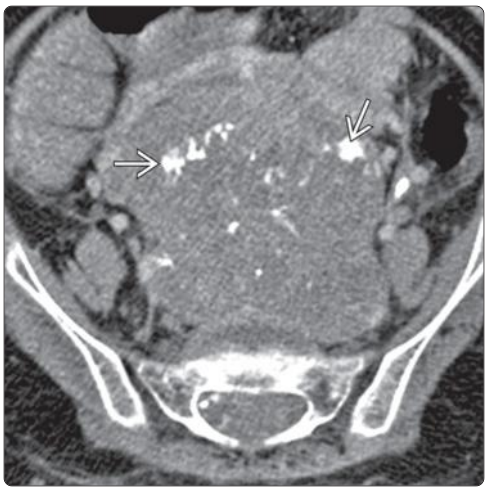


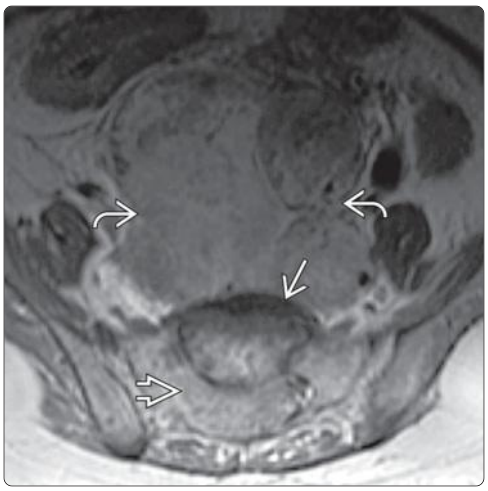
(Left) *Sagittal T1WI C+ (left) & T2WI (right) show a round, mildly enhancing suprahyoid carotid space mass. The patient presented with Horner syndrome. This cervical NB is T2 hypointense, suggesting hypercellularity.* **(Right)** *Color flow duplex US (left) shows a vascular hyperechoic mass in the left carotid space. This NB shows mild enhancement on T1WI C+ MR (right).*





(Left) *Sagittal T1WI C+ MR reveals heterogeneous vertebral body marrow replacement compatible with stage 4 NB. Pathologic compression fractures and dorsal extradural heterogeneous enhancing mass are also present.* **(Right)** *Axial STIR MR shows a posterior mediastinal GN affixed to adjacent ribs and vertebra without marrow signal abnormality to imply direct invasion. Note the absence of neural foraminal involvement.*





(Left) *Axial CECT demonstrates calcifications in a large pelvic mass. Calcifications are seen in > 80% of NBs at CT. Small tumors are usually homogeneous. Larger tumors show heterogeneous attenuation because of the presence of hemorrhage and necrosis, but they do not contain fat.* **(Right)** *Axial T1WI C+ MR in the same patient illustrates a large presacral mass with sacral bone involvement and epidural extension.*

Langerhans Cell Histiocytosis

KEY FACTS

TERMINOLOGY

- Abnormal histiocyte proliferation producing granulomatous skeletal lesions

IMAGING

- Calvarium > mandible > long bones > vertebrae
- Spinal involvement greater in children
  - Thoracic > lumbar > cervical
- Destructive lytic lesion ± pathologic fracture, paraspinal/epidural mass, spinal canal extension
- ± vertebra plana (adjacent discs, posterior elements rarely involved)

TOP DIFFERENTIAL DIAGNOSES

- Ewing sarcoma
- Metastases
- Neuroblastoma
- Hemopoietic malignancies

PATHOLOGY

- Restricted LCH
  - Only skin lesions
  - Osseous lesions ± diabetes insipidus (DI), adjacent lymph node involvement, or rash
- Extensive LCH
  - Visceral organ involvement ± bone lesions, DI, adjacent lymph node involvement, ± rash, ± hemopoietic organ system dysfunction

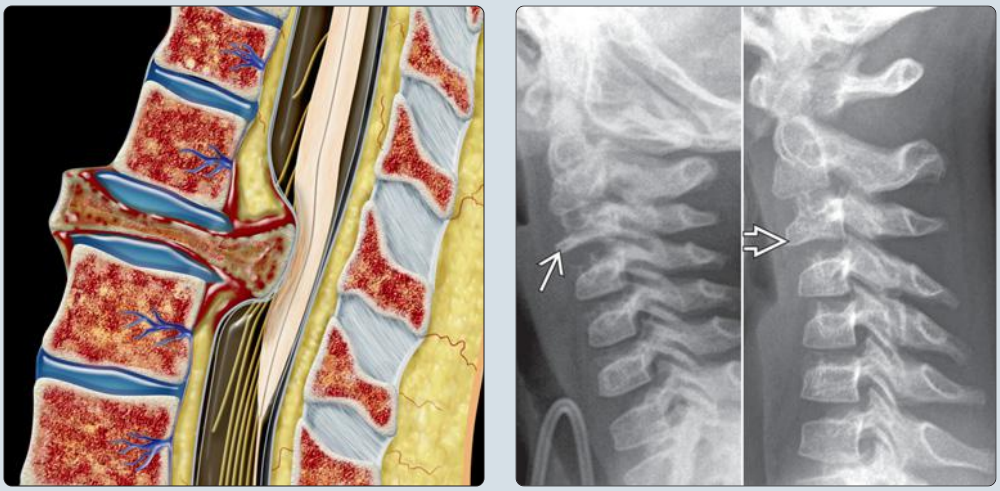
CLINICAL ISSUES

- Asymptomatic or localized pain due to medullary expansion or pathologic fracture
- ± fever, leukocytosis

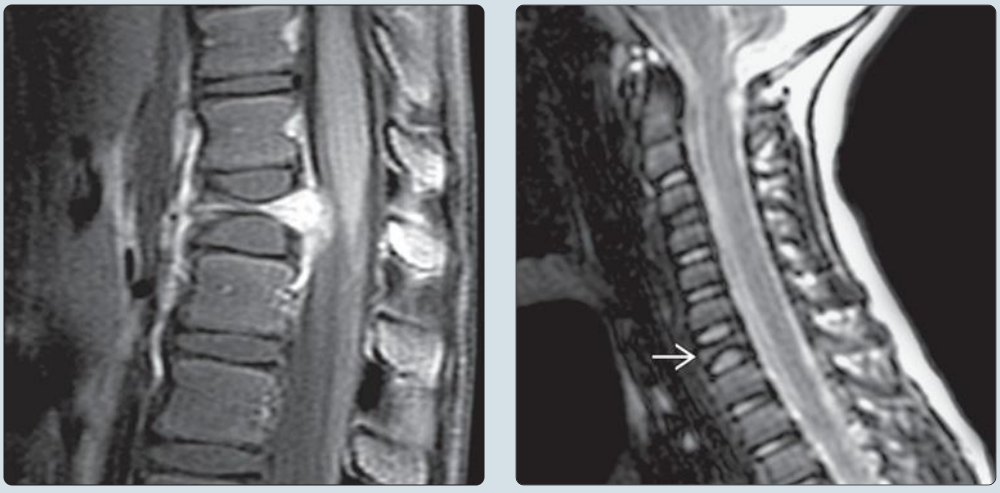
DIAGNOSTIC CHECKLIST

- Vertebra plana with relative absence of epidural mass, disc abnormalities classic for LCH

(Left) *Sagittal graphic demonstrates diffuse vertebral marrow infiltration with pathologic fracture producing vertebra plana sparing the disc spaces. Epidural extension results in ventral cord compression.* **(Right)** *Lateral radiographs show acute C3 vertebra plana ⇨ (left). Follow-up (right), 3 years later, exhibits substantial height reconstitution ⇨. Reconstitution of vertebral height results from endochondral ossification at the apophyseal plate.*



(Left) *Sagittal T1WI C+ MR shows pathological vertebral collapse with enhancing epidural mass. Vertebral endplates and adjacent discs are intact. (Courtesy S. Blaser, MD., & A. Illner, MD.)* **(Right)** *Sagittal T2WI MR (chronic LCH) demonstrate substantial C7 vertebral height loss ⇨. Marrow signal is normal, and no prevertebral or epidural mass is present.*



TERMINOLOGY

Synonyms

- Langerhans cell histiocytosis (LCH), Langerhans cell granulomatosis, eosinophilic granuloma (EG), histiocytosis X

Definitions

- Abnormal histiocyte proliferation producing granulomatous skeletal lesions

IMAGING

General Features

- Best diagnostic clue
  - ± vertebra plana sparing disc space
- Location
  - Calvarium > mandible > long bones > ribs > pelvis > vertebrae
  - Spinal involvement
    - Thoracic (54%) > lumbar (35%) > cervical (11%)
    - Children > adults
- Morphology
  - Destructive lytic lesion ± pathologic fracture, soft tissue mass, spinal canal extension

Radiographic Findings

- Radiography
  - Lytic nonsclerotic destructive vertebral lesion
    - ± vertebra plana (adjacent discs, posterior elements rarely involved)
    - ± scoliosis; kyphosis uncommon

CT Findings

- CECT
  - Lytic (nonsclerotic) destructive vertebral lesion with enhancing soft tissue mass ± paraspinal, epidural extension
- Bone CT
  - Lytic vertebral lesion ± collapsed vertebral body (vertebra plana)

MR Findings

- T1WI
  - Hypointense vertebral soft tissue mass ± pathologic fracture
- T2WI
  - Heterogeneously hyperintense soft tissue mass ± pathologic fracture
  - Disc spaces generally spared
- T1WI C+
  - Homogeneous enhancement

Nuclear Medicine Findings

- Bone scan
  - Variable uptake; bone lesions may be hot, cold, or mixed ("ring") activity
  - False-negatives common (35%)

Imaging Recommendations

- Protocol advice
  - Multiplanar enhanced MR to evaluate soft tissues, determine epidural extension
  - Bone algorithm CT with multiplanar reformats to define osseous destruction, &/or vertebral height loss

DIFFERENTIAL DIAGNOSIS

Ewing Sarcoma

- Permeative bone destruction ± pathological fracture
- More likely than LCH to have large soft tissue mass, adjacent bone destruction

Metastases

- Extension of primary tumor to spine
  - Bone production > destruction (blastic)
  - Bone destruction > production (lytic)

Neuroblastic Tumors

- Abdominal or thoracic paraspinal mass ± intraspinal extension

Hemopoietic Malignancies

- Multifocal disease; may be indistinguishable from LCH
- Widespread abnormal marrow signal, enhancement with extensive disease

Pyogenic Osteomyelitis

- Abnormal marrow signal ± vertebral collapse
- Hyperintense narrowed disc ± fluid signal

Giant Cell Tumor

- Expansile, lytic vertebral lesion + soft tissue mass
- Usually older patient (> 30 years)

PATHOLOGY

General Features

- Etiology
  - Dysregulated growth, activity, and trafficking of Langerhans cells
  - Unknown cause; infectious agents (especially viruses), immune system dysfunction, neoplastic mechanisms, genetic factors, cellular adhesion molecules proposed
- Associated abnormalities
  - Pituitary-hypothalamic axis → diabetes insipidus
  - Purulent otitis media → deafness
  - Orbital involvement → proptosis
  - Cutaneous LCH (≤ 50%)
  - Pulmonary involvement (20-40%); male predominance, older age (20-40 years), smoking association
  - Gastrointestinal (GI) bleeding, liver/spleen abnormalities
  - Lymph node enlargement ± suppuration, chronic drainage (30%)
- < 1% of biopsy-proven primary bone lesions; vertebral involvement (6%)
  - Most common cause of vertebra plana in children
  - Variable vertebral body restoration occurs over months to years
  - Pedicles often involved; posterior elements, adjacent disc spaces usually spared
- PCR-based assay demonstrates all LCH forms are clonal → LCH probably clonal neoplastic rather than reactive disorder as previously believed

Staging, Grading, & Classification

- Classic categorization of LCH based on age, severity, and extent of involvement
  - Eosinophilic granuloma
  - Hand-Schüller-Christian disease
  - Letterer-Siwe disease
- Revised LCH classification according to disease extent
  - Restricted LCH
    - Skin lesions without other site of involvement
    - Monostotic lesion ± diabetes insipidus (DI), adjacent lymph node involvement, or rash
    - Polyostotic lesions involving several bones or ≥ 2 lesions in 1 bone ± DI, adjacent lymph node involvement, or rash
  - Extensive LCH
    - Visceral organ involvement ± bone lesions, DI, adjacent lymph node involvement, &/or rash
      - ± lung, liver, or hemopoietic organ system dysfunction

Microscopic Features

- Light microscopy → abnormal granulomatous histiocytes (Langerhans cells) with folded nuclei, cytoplasmic S100 reactivity, areas of bone necrosis
- EM: Birbeck granule cytoplasmic inclusions

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Localized pain due to medullary expansion or pathologic fracture, reduced mobility, swelling, fever, or leukocytosis
  - Myelopathy or radiculopathy
- Clinical profile
  - EG (70%)
    - Localized form (bone lesions only), older children (5-15 years), good prognosis
    - Back pain, stiffness, scoliosis, neurologic complications, fever, leukocytosis
  - Hand-Schüller-Christian disease (20%)
    - Chronic disseminated osseous and visceral lesions, younger children (1-5 years), intermediate prognosis (fatal 10-30%)
    - Triad of diabetes insipidus, exophthalmos, osteolytic lesions
  - Letterer-Siwe disease (10%)
    - Acute form, rapid visceral organ dissemination, infants (< 3 years), poor prognosis
    - Fever, cachexia, anemia, hepatosplenomegaly, lymphadenopathy, rash, gum hyperplasia
    - Most patients die ≤ 1-2 years
  - Multisystem disease represents > 2/3 (68.6%) of total cases among adult patients with LCH
    - Skin and pulmonary involvement representing 51% and 62% of cases, respectively

Demographics

- Age
  - Predominantly affects children, adolescents, or young adults, but may occur at any age
    - Most severe manifestations peak at younger ages
- Gender
  - M:F = 2:1

Natural History & Prognosis

- Variable depending on presentation age, extent of systemic disease
  - Single-system disease 5-year event-free survival: 100%
  - Isolated pulmonary manifestation: 87.8% survival
  - Multisystem disease: 91.7% survival
- Spontaneous remission of lesions common, but relapse or reactivation possible
  - Lesions usually begin to regress after ~ 3 months, but may take up to 2 years to resolve
  - Lower rate of recurrence in skeletally immature
- Vertebra plana prognosis favorable for symptomatic improvement, vertebral height restoration with conservative orthopedic treatment
  - Vertebral height restoration varies 18.2% → 63.8% before, 72.2% → 97% after skeletal maturity

Treatment

- Conservative management initially with observation ± bracing
  - Combined chemotherapy options for multisystem disease
- Limited disease has excellent prognosis, usually without need for systemic therapy
  - Multifocal skeletal involvement, refractory cutaneous lesions, and disseminated or recurrent organ disease benefit from systemic treatment
- Advanced-stage disease: Local treatment, radiation therapy, chemotherapy, immunomodulation, and liver, lung, and stem cell transplantation
- ± surgical intervention (partial intralesional curettage, fusion), external beam radiotherapy, chemotherapy, steroids in patients with neurological deficits, conservative treatment failure

DIAGNOSTIC CHECKLIST

Consider

- Vertebra plana much more common in children than adults
- Unexplained vertebral compression fractures in children or adolescents merit evaluation to exclude LCH or leukemia

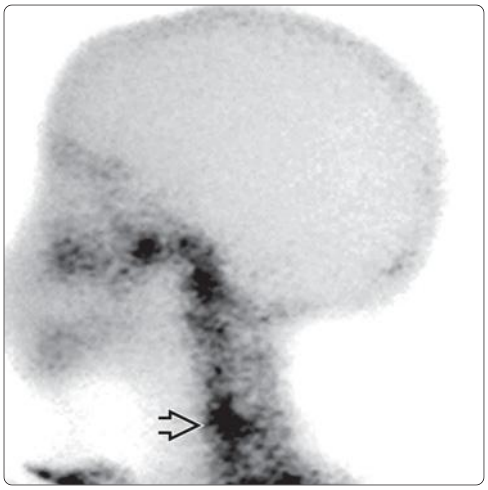
Image Interpretation Pearls

- Vertebra plana with relative absence of epidural mass, disc abnormalities classic for LCH

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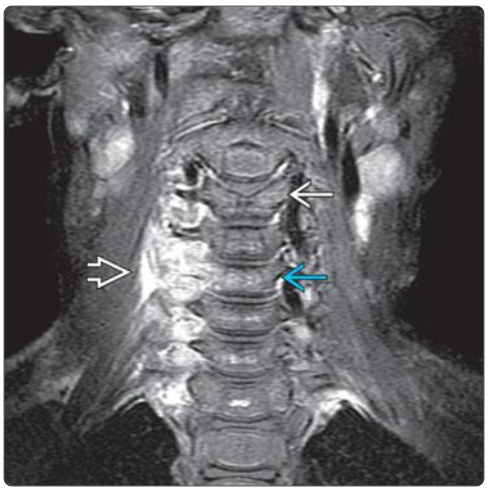


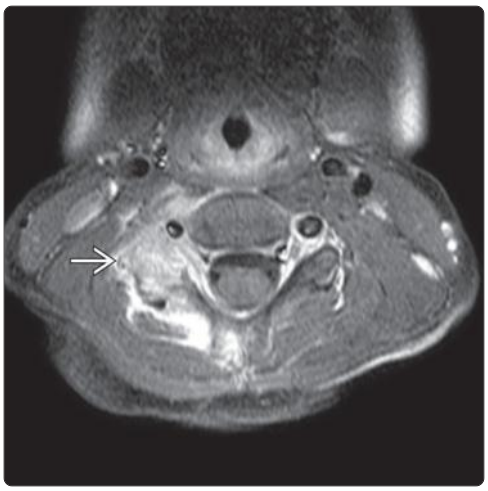
(Left) *Lateral radiograph of the cervical spine reveals complete flattening of the C6 vertebra ➡ in a typical "vertebra plana" deformity. Note that the posterior elements remain normally aligned.* **(Right)** *Lateral planar spot image from a Tc-99m MDP bone scan confirms abnormal increased radioisotope uptake ⇨ in this active LCH lesion.*





(Left) *Sagittal T1WI C+ FS MR of the lumbar spine demonstrates intense homogeneous enhancement of a pathologic L3 fracture ➡ resulting in vertebra plana and an epidural mass. Note that the intervertebral disc spaces are preserved.* **(Right)** *Axial T1 C+ FS MR reveals relatively mild abnormal enhancement of the flattened L3 vertebra (volume averaging with disc space) with posterior extension into the epidural space.*





(Left) *Coronal T1 C+ FS MR shows central collapse plana of C3 ➡ and mild height loss at C5 ⇨. There is minimal enhancement of older lesion at C3 and moderate enhancement of the more active C5 lesion. Note asymmetric right-sided soft tissue enhancement and lymphadenopathy ➡.* **(Right)** *Axial T1WI C+ FS MR confirms asymmetric right perifacet enhancement extending into the adjacent soft tissues ➡. Involvement of posterior elements is unusual.*

Angiolipoma

KEY FACTS

TERMINOLOGY

- Benign tumor of adipose and vascular elements

IMAGING

- Extradural mass with fat attenuation (-20 to -60 HU) with scattered soft tissue reticulation on CT
- Thoracic predominance; usually extends over 1-4 vertebral body segments
- Hyperintense on unenhanced T1WI, + enhancement on fat-suppressed T1WI
  - Inhomogeneous signal (attributed to vascular channels)
- Spine involvement uncommon: Extradural
  - Degree of central hypointensity on T1WI MR is predictive of degree of vascularity
- Mildly increased signal relative to CSF on T2WI/STIR

TOP DIFFERENTIAL DIAGNOSES

- Lipoma
- Liposarcoma
- Hematoma
- Abscess, epidural, paravertebral
- Epidural lipomatosis

PATHOLOGY

- Infiltrating form more common in anterior epidural space, may destroy adjacent bone
- Focal form more common in posterior thoracic epidural space, no bone destruction

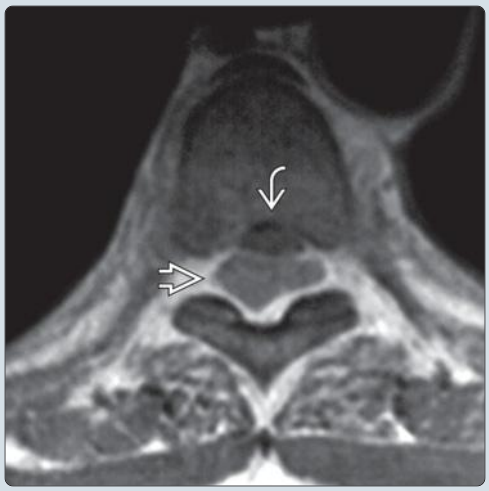
CLINICAL ISSUES

- Slowly progressive paraparesis with back pain

DIAGNOSTIC CHECKLIST

- Fat-suppressed T1WI most useful for lesion definition, showing heterogeneous enhancement
- Not to be confused with angiomyolipoma, which is found in kidney of tuberous sclerosis patients





(Left) *Sagittal T1WI MR shows a large, predominately T1 hyperintense mass involving a long segment of the thoracic dorsal epidural space, with moderate spinal cord compression. Intermixed with the fatty component are islands of intermediate T1 signal (angiomatous tissue).* **(Right)** *Axial T1WI C+ MR of a large thoracic dorsal epidural angiolipoma demonstrates cord compression. Within the fatty component is the angiomatous component of the lesion, which exhibits hypointense signal.*





(Left) *Sagittal STIR MR depicts a lobulated, heterogeneous dorsal epidural mass. The fatty component suppresses on STIR; the vascular component remains hyperintense.* **(Right)** *Sagittal T1WI MR (left) reveals a heterogeneous lesion within the dorsal epidural space at the L3-L4 level. Sagittal T1 C+ MR (right) confirms mild heterogeneous enhancement of the angiomatous portions.*

TERMINOLOGY

Abbreviations

- Spinal angiolipoma (SAL)

Synonyms

- Vascular lipoma, hemangiolipoma, fibromyolipoma

Definitions

- Benign tumor of adipose and vascular elements

IMAGING

General Features

- Best diagnostic clue
  - Hyperintense mass on unenhanced T1WI, showing enhancement on fat-suppressed T1WI
- Location
  - Uncommon tumors of extremities, trunk, neck
  - Spine involvement uncommon: Extradural
    - Thoracic predominance may be result of regional variation in blood supply
      - Midthoracic spine is least perfused
    - Intramedullary SAL is **extremely rare**
  - SAL can infiltrate from paravertebral areas into spinal canal
  - Rare reports of mediastinal angiolipoma with spinal canal extension
- Size
  - Extend over 1-4 vertebral body segments
  - Average extent > 2 vertebral bodies in length
- Morphology
  - Focal or infiltrating mass showing heterogeneous fat signal

Radiographic Findings

- Often negative
- Some cases show erosions of pedicle and widening of spinal canal
- Infiltrating tumors cause trabeculation of affected vertebral body

CT Findings

- NECT
  - Extradural mass of fat attenuation (-20 to -60 HU), with scattered soft tissue reticulation
  - Slight enhancement
  - Rare calcification
  - Rare bony remodeling

MR Findings

- T1WI
  - Hyperintense on T1WI, with inhomogeneous signal
  - Heterogeneity thought to be related to vascular channels
  - Prominence of vascular component may give more isointense T1 signal
- T2WI
  - Mild increased signal relative to CSF
- PD/intermediate
  - Increased signal relative to CSF, but decreased signal relative to fat
- T1WI C+
  - Fat-suppressed T1WI most useful for lesion definition, showing heterogeneous enhancement

Nonvascular Interventions

- Myelography
  - Nonspecific extradural mass
  - May show block with aggressive lesion

Imaging Recommendations

- Best imaging tool
  - Fat-suppressed contrast-enhanced T1WI
- Protocol advice
  - Axial, sagittal T1, STIR, T2WI MR precontrast
  - Axial, sagittal post-contrast fat-suppressed T1WI MR

Nuclear Medicine Findings

- PET
  - SAL can be detected by using F18 FDG PET

DIFFERENTIAL DIAGNOSIS

Lipoma

- Little or no enhancement, minimal septations
- May be intradural, extradural, or subcutaneous
- Distinguishable microscopically, with angiolipoma showing vessels branching pattern and fibrous scarring

Liposarcoma

- May have irregular thickened septa, with areas of T2 hyperintensity
- Liposarcoma show cellular pleomorphism and mitotic activity
- May show considerable T1 isointense tissue

Hematoma

- No suppression with fat sat
- No enhancement, unless more chronic
- May show low signal on T2* images

Abscess, Epidural, Paravertebral

- May show heterogeneous T1 signal within epidural fat, with focal areas of low signal
- Prominent peripheral enhancement
- May show heterogeneous signal and enhancement like angiolipoma

Epidural Lipomatosis

- Homogeneous fat signal, without enhancement
- ± history of steroid use
- Typical dorsal thoracic epidural location

PATHOLOGY

General Features

- Etiology
  - Unknown, with several theories
    - May originate from primitive pluripotential mesenchymal cells
    - Congenital malformation
    - Hamartomatous lesion, enlarging in response to injury/inflammation
- Genetics

- Subcutaneous angiolipomas show predominately normal karyotype
  - vs. characteristic chromosomal aberrations in other lipomatous lesions, such as lipoma, lipoblastoma, hibernomas
- Rare familial occurrence of angiolipomas
- Benign tumor usually found in extremities, trunk, or neck
- Spine angiolipomas rare
- May occur as focal or infiltrating forms
  - Infiltrating form more common in anterior epidural space, may destroy adjacent bone
  - Focal form more common in posterior thoracic epidural space, no bone destruction
    - May occur at multiple sites in subcutaneous tissues

Gross Pathologic & Surgical Features

- Gross features of fatty tissue; although, may show port wine or dark brown coloration

Microscopic Features

- Presence of fat and vascular tissues
  - Vascular proliferation with microthrombi
  - Consistent marker of angiolipomas is presence of fibrin microthrombi in vascular channels associated with disrupted endothelial cells
- SAL is distinct from cutaneous angiolipoma in that caliber of its predominant vascular component is far larger than that of fat cells
- Vessels variously described as sinusoids, thin walled, thick walled with smooth muscle proliferation
- Mitoses and pleomorphism not present
- Features vary
  - Predominately lipomatous with few small angiomatous regions
  - Predominately vascular with small lipomatous component

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific back pain
  - Other signs/symptoms: Cord compression
    - Progressive paraparesis
    - Symptoms may progress with pregnancy
      - May be associated with weight gain
      - Increase in tumor volume resulting in impaired spinal venous drainage
      - Hormonal changes resulting in increased extravascular fluid volume
    - Obesity may lead to symptoms induced by greater fatty component
    - Prolonged neurological course can suddenly deteriorate
      - Vascular engorgement
      - Enlarging or degenerated blood vessels
      - Vascular steal phenomenon
      - Venous stasis with thrombosis
      - Hemorrhage into lesion
- Clinical profile
  - Slowly progressive paraparesis with back pain

Demographics

- Age
  - 4th-5th decade; although, may occur in children
  - Mean age 42 years
- Gender
  - F > M
- Epidemiology
  - Rare, ~ 0.14-1.2% of all spinal tumors
  - 2-3% of extradural tumors
  - Focal form more common in spine
  - Typically midthoracic level, but can occur anywhere in spine
  - CNS involvement rare, but 90% occur in spine

Natural History & Prognosis

- Slowly progressive
- Bone infiltration associated with more aggressive behavior and worse prognosis

Treatment

- Surgical excision for painful, symptomatic lesions
  - Focal form: Encapsulated, low rate of recurrence following excision
  - Infiltrative form: 50% recurrence rate
    - Wider resection followed by radiotherapy should be considered
- Partial resection may give good symptomatic relief in infiltrative form
- Degree of hypointensity on T1WI predictive of degree of vascularity encountered at surgery

DIAGNOSTIC CHECKLIST

Consider

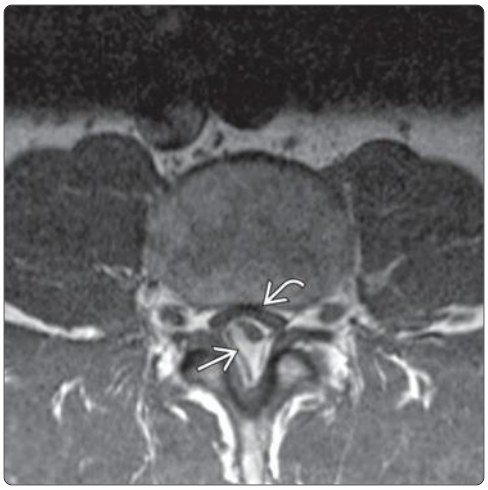
- Not to be confused with angiomyolipoma, which is seen in tuberous sclerosis and occurs in kidney

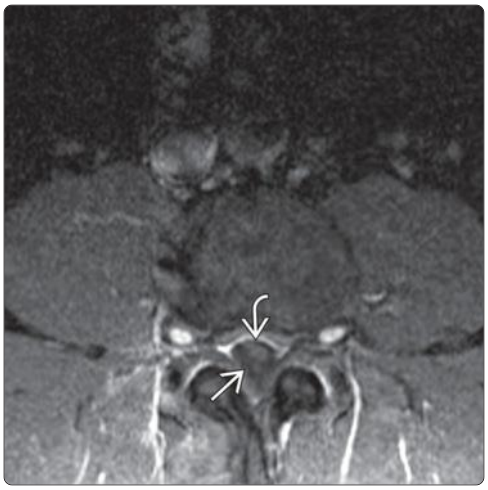
Image Interpretation Pearls

- Angiolipomas do not show vascular flow voids

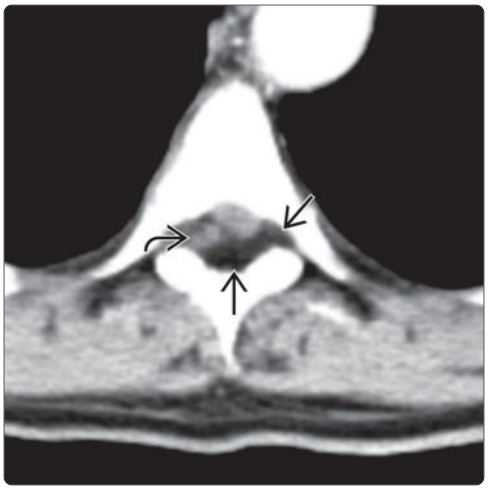
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(Left) *Axial T1WI MR illustrates thecal sac compression by a heterogeneous extradural SAL . The degree of central hypointensity on T1WI MR is predictive of the degree of vascularity likely to be encountered at surgery.* **(Right)** *Axial T1WI C+ FS MR reveals minimal peripheral enhancement of the posterior angiolipoma. The fatty lesion elements appear dark due to fat suppression.*





(Left) *Axial CECT through the thoracic spine demonstrates a fat attenuation dorsal epidural mass with a soft tissue component filling much of the spinal canal.* **(Right)** *Sagittal T1WI MR demonstrates a large, heterogeneous, predominately T1 hyperintense dorsal epidural mass producing regional spinal cord compression.*





(Left) *Sagittal T1WI C+ FS MR reveals a slightly heterogeneous, intensely enhancing epidural mass involving dorsal upper thoracic spine with cord compression. This is a nonspecific MR appearance of a dorsal epidural mass, and preoperative diagnosis of SAL can be difficult.* **(Right)** *Sagittal STIR MR depicts a lobulated hyperintense dorsal epidural mass involving upper thoracic spine with cord compression.*

Schwannoma

KEY FACTS

TERMINOLOGY

- Neurinoma, neurilemmoma (outdated terms)
- Neoplasm of nerve sheath (Schwann cells)

IMAGING

- 70-75% intradural extramedullary
  - Most common intradural extramedullary mass
- 15% completely extradural
- 15% transforaminal, "dumbbell" masses
- Bone remodeling due to large intraspinal or intraforaminal tumor common
- Cystic change common
- Calcifications, hemorrhage are rare
- Uniform, heterogeneous, or peripheral enhancement patterns

TOP DIFFERENTIAL DIAGNOSES

- Neurofibroma (NF)
- Perineural root sleeve cyst
- Myxopapillary ependymoma
- Meningioma
- Leptomeningeal carcinomatosis
- Neuroblastic tumor

PATHOLOGY

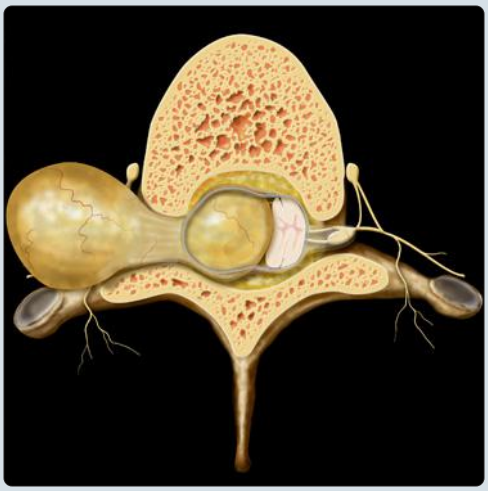
- WHO grade I
- NF2: Loss of tumor suppressor (merlin) on chromosome 22
- Sporadic schwannoma more common than NF2
  - Inactivating mutations of merlin gene in ~ 60%

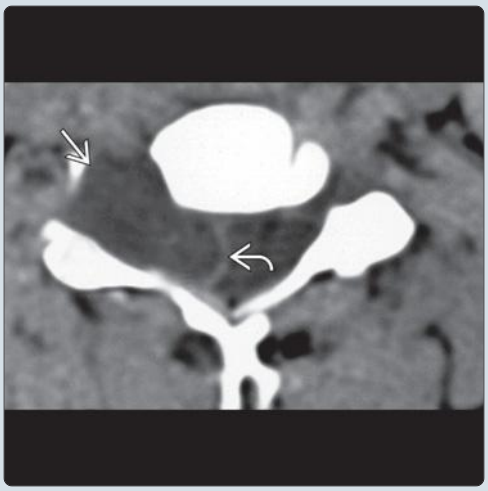
CLINICAL ISSUES

- Pain, weakness, paresthesias most common clinical findings
- Typically solitary unless part of inherited tumor syndrome

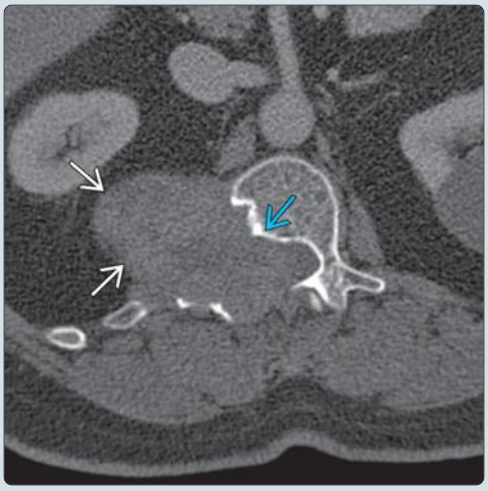
DIAGNOSTIC CHECKLIST

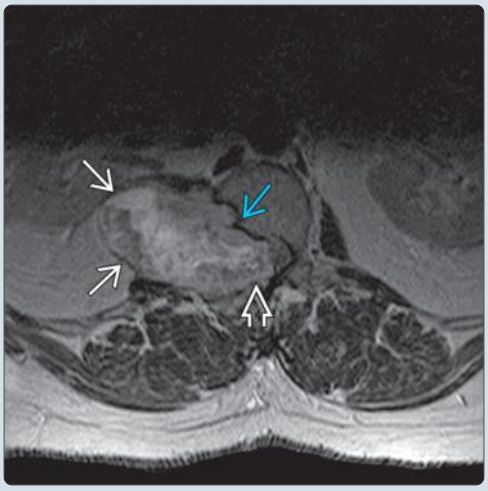
- Schwannoma most likely when solitary enhancing "dumbbell-shaped" spinal lesion present





(Left) *Axial graphic portrays a right-sided dumbbell-shaped spinal nerve root schwannoma, enlarging the neural foramen and compressing the spinal cord. Both intra- and extradural components ("dumbbell") are present.* **(Right)** *Axial CECT shows a hypodense transforaminal mass enlarging the right neural foramen. The intraspinal component effaces the thecal sac and causes canal stenosis.*





(Left) *Axial NECT demonstrates a large soft tissue mass enlarging the right L1-2 neural foramen. There is conspicuous scalloping of the vertebral body. The full intraspinal component is not seen.* **(Right)** *Axial T2WI MR in the same patient reveals heterogeneous mass signal intensity. Foraminal enlargement and vertebral body scalloping are again seen. The extradural component within the spinal canal is causing significant canal stenosis.*

TERMINOLOGY

Abbreviations

- Neurofibromatosis type 2 (NF2)
  - Multiple inherited schwannomas, meningiomas, and ependymomas (MISME)

Synonyms

- Neurinoma, neurilemmoma (outdated terms)
- Nerve sheath tumor (NST)

Definitions

- Neoplasm of nerve sheath (Schwann cells)

IMAGING

General Features

- Best diagnostic clue
  - Well-circumscribed, "dumbbell-shaped' transforaminal mass
  - Solitary or multiple enhancing cauda equina nodules
- Location
  - 70-75% intradural extramedullary
    - Most common intradural extramedullary mass
  - 15% completely extradural
  - 15% "dumbbell"
    - Both intra- and extradural
  - Rare intramedullary
  - Thoracic > cervical = lumbar
- Size
  - Most are small: Few millimeters in diameter
  - Giant schwannoma
    - Extend ≥ 2 vertebral segments
    - Paraspinal extension > 2.5 cm
    - Posterolateral extension into myofascial planes: **Giant invasive schwannoma**
    - Lumbosacral spine most common
- Morphology
  - Round
  - Lobulated

Radiographic Findings

- Radiography
  - Bony remodeling due to large intraspinal or transforaminal mass
    - Thinned pedicle, enlarged neural foramen, vertebral body scalloping, widened interpedicular distance
  - May see paravertebral mass in thoracic spine

CT Findings

- NECT
  - Well-marginated mass
    - Intraforaminal, paravertebral, or intraspinal (more difficult to see on NECT)
  - Isodense to cord, nerve roots
    - Cystic change common
    - Calcifications rare
- CECT
  - Moderate solid or peripheral enhancement
- Bone CT
  - Bone remodeling adjacent to tumor

MR Findings

- T1WI
  - Iso-/hypointense relative to cord, roots
  - Melanotic schwannoma hyperintense
- T2WI
  - 75% hyperintense
  - 40% cystic change
  - 10% hemorrhage
  - Occasional: "Target" sign
    - High signal rim, low-intensity center
    - More typical of neurofibroma
  - ± myelomalacia or edema due to spinal cord compression
- T1WI C+
  - Uniform, heterogeneous, or peripheral enhancement patterns

Angiographic Findings

- Variable vascularity

Nonvascular Interventions

- Myelography
  - Intradural mass or extradural compression of thecal sac
  - May cause myelographic block if intraspinal

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR
- Protocol advice
  - Include post-gadolinium imaging with fat suppression
  - Scan entire spine in asymptomatic patients with suspected neurofibromatosis type 2 (NF2)

DIFFERENTIAL DIAGNOSIS

Neurofibroma (NF)

- Difficult or impossible to distinguish solitary neurofibroma from schwannoma by imaging
- NF more often fusiform
- "Target" sign more common with NF than schwannoma
- Hemorrhage, cystic or fatty degeneration more common with schwannoma

Perineural Root Sleeve Cyst

- Cystic nonenhancing mass
- CSF density/signal intensity

Myxopapillary Ependymoma

- Large vascular intraspinal mass
- Hemorrhage common
- Small ependymoma may be indistinguishable from schwannoma

Meningioma

- Dural-based enhancing mass
- Calcification common

Leptomeningeal Carcinomatosis

- Smooth or nodular enhancement along pial surface of spinal cord, cauda equina

Neuroblastic Tumor

- Paraspinal mass, usually in pediatric patient

- May see intraspinal extension through neural foramen ("dumbbell" tumor)

Extruded Disc Fragments

- Simulate appearance of intraspinal (extradural) mass
- Do not enhance

PATHOLOGY

General Features

- Genetics
  - NF2 (MISME syndrome)
    - Hereditary, with autosomal dominant transmission
    - Incidence: 1:30,000-40,000
    - Mutation on long arm of chromosome 22
      - Tumor suppressor: Merlin (a.k.a. schwannomin)
      - Relative stable phenotype for a particular *NF2* gene mutation
    - Bilateral acoustic schwannomas diagnostic
    - Multiple schwannomas, meningiomas, ependymomas
  - Sporadic: Most common
    - Inactivating mutations of *NF2* gene in ~ 60%
  - Schwannomatosis
    - Multiple peripheral schwannomas
    - Other NF2 features absent
  - Carney complex
    - Hereditary, with autosomal dominant transmission
    - Gene loss or inactivation on long arm of chromosome 17
      - Tumor suppressor
    - Melanotic schwannoma, cutaneous myxomas, potentially life-threatening cardiac myxomas, pigmented adrenal tumors

Staging, Grading, & Classification

- WHO grade I

Gross Pathologic & Surgical Features

- Circumscribed, well encapsulated
- Light tan/yellow, round/ovoid
- Cysts may occur
- Gross hemorrhage, frank necrosis uncommon

Microscopic Features

- 3-layered capsule: Fibrous layer, nerve tissue, transitional layer
- Cell of origin: Schwann cells
- Classic "biphasic" pattern
  - Antoni A: Compact, elongated cells with occasional palisading
  - Antoni B: Less cellular, loosely textured, often lipidized
- Hyalinized blood vessels
- Fatty degeneration, cystic change, hemorrhage
- May contain melanin: 50% of melanotic schwannomas occur with Carney complex
- Immunohistochemistry: S100(+) and anti-leu-7(+); EMA(+)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain
  - Weakness
  - Paresthesias
- Clinical profile
  - Pain associated with movement

Demographics

- Age
  - 30-60 years
- Gender
  - M = F
- Ethnicity
  - No racial predilection
- Epidemiology
  - 30% of primary spine tumors
  - Typically solitary unless part of inherited tumor syndrome

Natural History & Prognosis

- Slow growing
- Malignant degeneration rare
- No recurrence for typical schwannoma
  - NF2 and schwannomatosis may develop new lesions
  - Melanotic schwannoma: 15% recurrence, 26% metastasis

Treatment

- Total surgical resection
- Highly conformal radiosurgery may have role for symptom relief while preserving function

DIAGNOSTIC CHECKLIST

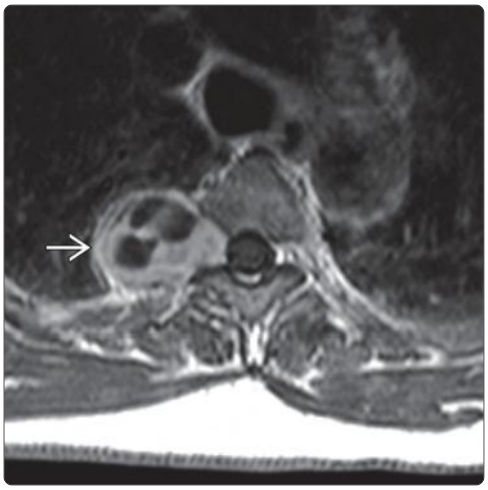
Image Interpretation Pearls

- Schwannoma most likely when solitary enhancing "dumbbell-shaped" spinal lesion present

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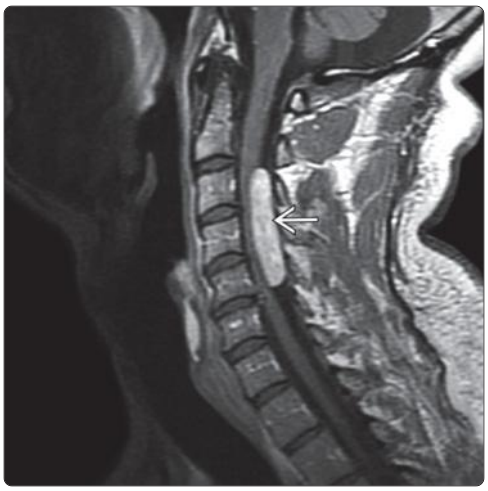


(Left) *Oblique radiograph of the cervical spine demonstrates marked pedicle thinning, foraminal enlargement, and vertebral body scalloping due to remodeling by a large intraspinal schwannoma.* **(Right)** *Axial T1WI C+ MR depicts a well-defined enhancing mass extending from the right neural foramen. Intratumoral cysts are typical of schwannoma.*





(Left) *Sagittal T1WI C+ FS MR reveals a well-defined intradural mass at the L3 level with vivid enhancement.* **(Right)** *Sagittal T1WI C+ MR shows an extensive intraspinal lumbar schwannoma with markedly heterogeneous enhancement extending from the lower thoracic spine to the L4 level. Chronic bony remodeling results in scalloping of multiple vertebral bodies.*





(Left) *Sagittal T1WI C+ MR shows an oblong intradural extramedullary mass dorsal to the cervical cord with cord compression. Homogeneous enhancement of the intradural mass is noted on post-contrast imaging. Meningioma would be an alternative diagnostic consideration, but the mass does not appear to be dural based and there is no dural tail.* **(Right)** *Sagittal T2WI FS MR demonstrates innumerable schwannomas of different sizes within the cauda equina in this patient with neurofibromatosis type 2.*

Meningioma

KEY FACTS

IMAGING

- Intradural extramedullary mass
- Thoracic (80%) > cervical (16%) > lumbar (4%)
- Typically round or ovoid (en globe)
  - en plaque variety pancaked or flat along dura
- Broad dural attachment (more often ventral or ventrolateral)
- Strong homogeneous enhancement
- Calcification in 1-5%
- No bony remodeling or hyperostosis in spine

TOP DIFFERENTIAL DIAGNOSES

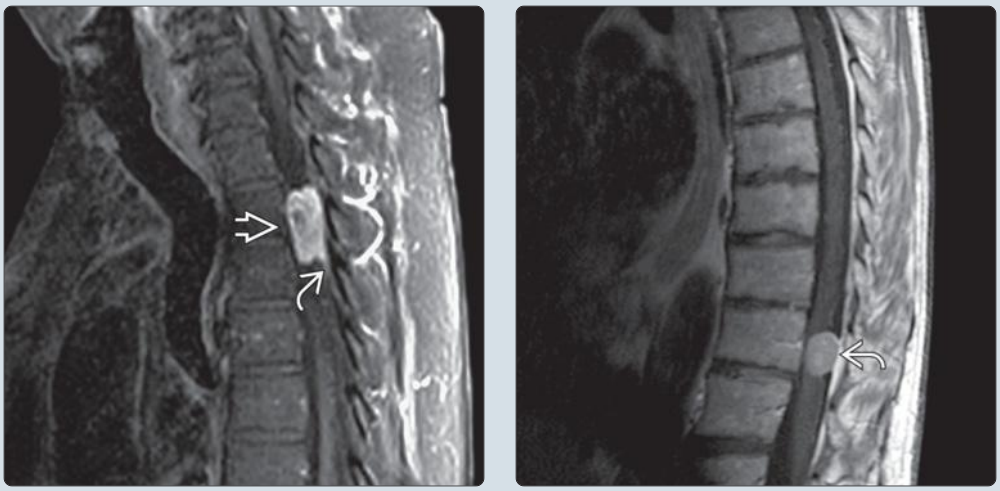
- Schwannoma
- Ependymoma, myxopapillary
- Lymphoma
- Intradural metastases

PATHOLOGY

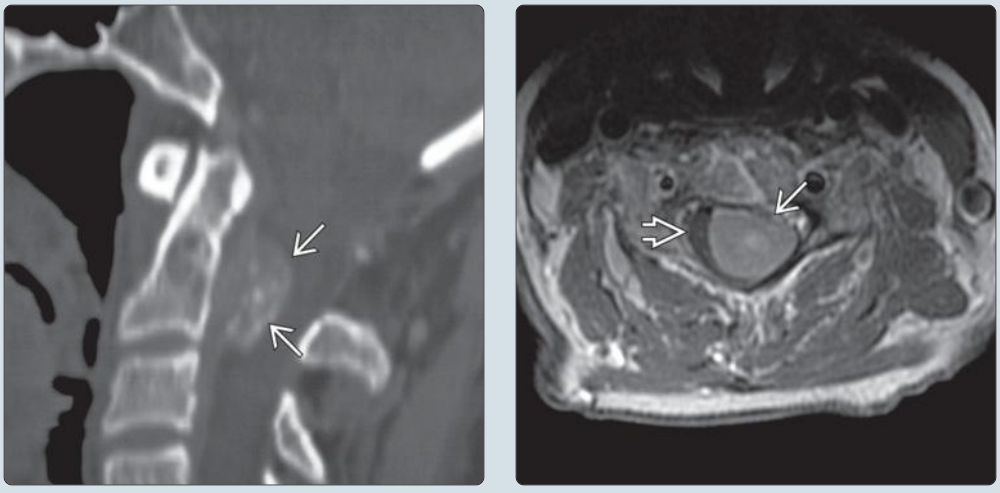
- Arise from arachnoid cap cells
- > 95% WHO grade I
- Most solitary, sporadic
  - Almost all have 22q12 abnormalities
- Syndromes
  - Multiple lesions with NF2
    - Intradural extramedullary meningiomas and schwannomas
    - Intramedullary ependymomas
  - Familial clear cell meningioma syndrome
  - Multiple meningiomatosis

CLINICAL ISSUES

- Peak incidence: 5th-6th decade
- > 80% female
- 2nd most common intradural extramedullary tumor
- Sensory and motor deficits (84%)
- Gait disturbances (83%)
- Local pain (47%)



(Left) *Sagittal T1WI C+ FS MR of the upper thoracic spine shows an intensely enhancing intradural mass with a broad dorsal dural attachment. A small enhancing dural tail is seen, a finding suggestive of but not exclusive to meningioma.* **(Right)** *Sagittal T1WI C+ MR shows a rounded, circumscribed, intradural extramedullary mass with homogeneous, intense enhancement positioned dorsal to the thoracic cord. A dural tail is not evident in this case.*



(Left) *Sagittal reconstructed CT image shows a sizable, partially calcified meningioma arising on the ventral margin of the canal at the level of C2. The mass results in significant canal stenosis and cord compression.* **(Right)** *Axial T1 C+ image shows a large cervical meningioma with a ventral, dural-based attachment at the C5-6 level, severely compressing and displacing the cervical cord posteriorly and to the right.*

TERMINOLOGY

Definitions

- Slow-growing, benign tumor originating from dura mater

IMAGING

General Features

- Best diagnostic clue
  - Intradural extramedullary mass that enhances avidly with contrast
- Location
  - Intradural extramedullary > 95%
    - Can rarely be intraosseous, extradural, or even paraspinous
  - Thoracic (80%) > cervical (16%) > lumbar (4%)
    - Site frequency is roughly proportional to surface area of dura in spine
- Size
  - Variable
- Morphology
  - Typically round or ovoid (en globe)
    - en plaque variety is pancaked or flat along dura
  - Broad dural attachment (more often ventral or ventrolateral)
  - May see dural tail (58% in 1 series of spinal meningiomas)

Radiographic Findings

- Radiography
  - Focal intraspinal opacity (if calcified)

CT Findings

- NECT
  - Most often of similar attenuation to spinal cord, may be occult on NECT
  - Calcification in 1-5%
  - Hyperostosis not a feature of spinal meningiomas
  - Rare bony remodeling (e.g., foraminal enlargement)
- CECT
  - Strong, usually homogeneous enhancement

MR Findings

- T1WI
  - Isointense to spinal cord
- T2WI
  - Majority isointense to cord
    - Hypointense if calcified
    - Cystic degeneration in 2-4%, hyperintense on T2WI
  - Very vascular meningioma may have prominent flow voids
- T1WI C+
  - Prominent, diffuse enhancement
  - Dural tails

Angiographic Findings

- Conventional
  - Early capillary blush
  - Vascular stain well into venous phase

Nonvascular Interventions

- Myelography
  - Rounded intradural extramedullary mass
  - Sharp meniscus of contrast caps lesion (classic for intradural extramedullary mass)
  - Ipsilateral subarachnoid space widened (cord, roots displaced away from mass)
  - May result in myelographic block

Imaging Recommendations

- MR ± contrast
- CT (if densely calcified)

DIFFERENTIAL DIAGNOSIS

Schwannoma

- Variable enhancement, may be heterogeneous
- Not dural based, but may broadly abut dura when large and difficult to differentiate from meningioma
- Higher signal on T2WI with more pronounced cystic change
- Less commonly posterior to cord

Myxopapillary Ependymoma

- Highly enhancing mass of lumbar canal
- Not dural based, but may broadly abut dura; may be difficult to differentiate from meningioma
- Hemorrhage or hemosiderin cap

Other Intradural Extramedullary Masses

- Paraganglioma
  - Looks like myxopapillary ependymoma
  - Rare
- Epidermoid cysts
  - Intradural mass
  - Low T1 signal, similar or mildly increased signal relative to CSF
  - No central enhancement
- Arachnoid cyst
  - Cystic intradural or epidural mass, identical to CSF on CT, MR
- Intradural metastasis
  - Often multiple
  - Advanced disease

Lymphoma

- Solitary intradural mass uncommon

Extradural Masses

- Can simulate rare extradural meningioma if large or difficult to compartmentalize
  - Hemangioma, epidural
  - Lymphoma
  - Chordoma

PATHOLOGY

General Features

- Etiology
  - Arise from arachnoid cap cells
- Genetics
  - Most lesions are solitary and sporadic
    - Almost all have 22q12 abnormalities
  - Multiple tumors with NF2
    - Meningiomas, schwannomas, ependymomas
    - Meningioma in a child, think NF2
  - Multiple meningiomatosis

- Rare familial clear cell meningioma syndrome
  - Loss of function mutation in SMARCE1

Staging, Grading, & Classification

- > 95% WHO grade I

Gross Pathologic & Surgical Features

- Firm, well-demarcated, lobulated/rounded mass with dural attachment
- Expands centripetally within dural sac
- May rarely have dumbbell morphology simulating schwannoma
- Occasionally cystic

Microscopic Features

- Most are typical (low grade) meningiomas
  - Common histologic subtypes
    - Most common: Psammomatous type with Ca++ concretions
    - Meningothelial
    - Fibrous
    - Transitional
  - Less common
    - Angiomatous
    - Microcystic
    - Clear cell
    - Choroid
    - Lipomatous
    - Hemorrhagic
- Rare
  - Atypical (increased mitoses, cellularity, etc.)
  - Anaplastic (malignant)
    - Angioblastic, difficult to distinguish from hemangiopericytoma
    - Can metastasize or be multicentric

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sensory and motor deficits (84%)
  - Gait disturbances (83%)
  - Local pain (47%)
- Clinical profile
  - Female with onset of myelopathy

Demographics

- Age
  - Peak incidence: 5th-6th decade
  - Under 50, more commonly genetic, worse prognosis
- Gender
  - > 80% female
- Epidemiology
  - 2nd most common intradural extramedullary tumor
    - 25% of primary spinal tumors
  - F:M = 4:1
  - Intracranial: Spine meningiomas = 8:1

Natural History & Prognosis

- Slow growing, compresses but does not invade adjacent structures
- Nonspecific symptoms, typically does not come to attention until significant neurologic compromise
- 0-13% overall recurrence rate; higher with incomplete excision or with infiltrating or en plaque tumors

Treatment

- Surgical resection (best outcome with Simpson I resection)
- Radiosurgical treatment shown to have at least short-term benefit in select patients

DIAGNOSTIC CHECKLIST

Consider

- Angiogram to distinguish between meningioma and neurofibroma

Image Interpretation Pearls

- Tumor dorsal to cord likely meningioma, not schwannoma (nerve roots are anterolaterally located)
- Thoracic tumor in female patient is more likely meningioma
- MR features suggesting schwannoma instead of meningioma: Foraminal enlargement, located within lumbar spine, fluid signal on T2WI, rim enhancement

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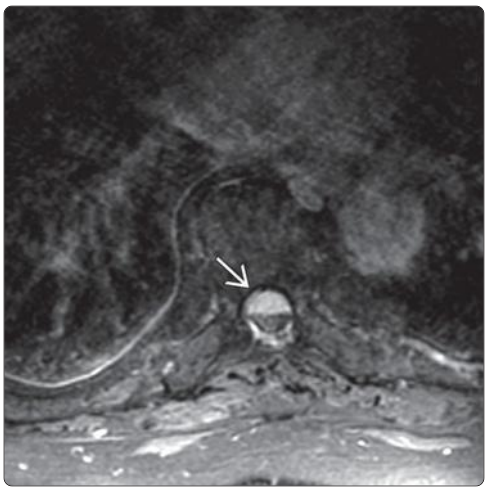
(Left) *Sagittal T2 MR shows a round, ventral, dural-based mass at C5-6 level, compressing and displacing the cervical cord posteriorly. Intradural extramedullary mass displaces the cord with concomitant widening of the CSF spaces adjacent to the mass. This T2 image shows CSF flow dephasing with signal loss adjacent to the mass.* **(Right)** *Large intradural extramedullary meningioma shows prominent enhancement. Note the broad ventral dural attachment, and severe cord compression.*





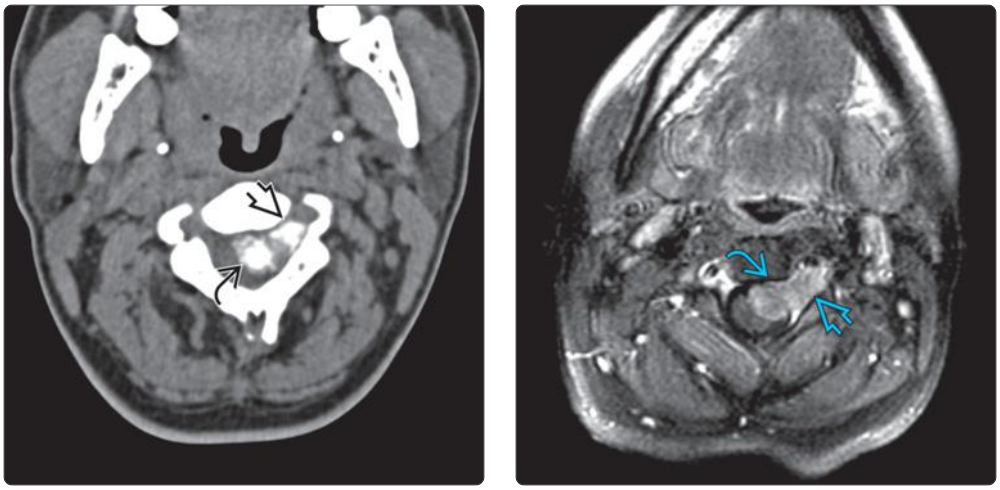
(Left) *Sagittal T2WI MR demonstrates an intradural extramedullary mass with a broad dural attachment along the dorsal aspect of the thoracic canal. The mass is mildly heterogeneous and iso- to hypointense relative to the cord.* **(Right)** *Sagittal T1 C+ FS MR in the same patient demonstrates relatively homogeneous enhancement throughout the mass.*



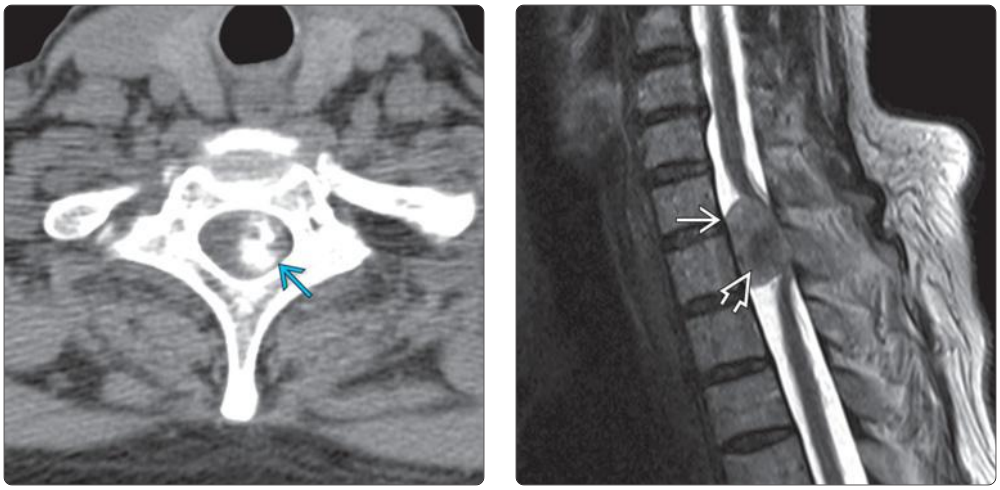


(Left) *Sagittal T1 C+ FS MR shows the en plaque type of morphology of a thoracic meningioma with a long segment of involvement of the ventral dura.* **(Right)** *Axial T1 C+ FS MR shows a homogeneously enhancing mass ventral to the spinal cord with a broad ventral dural base.*

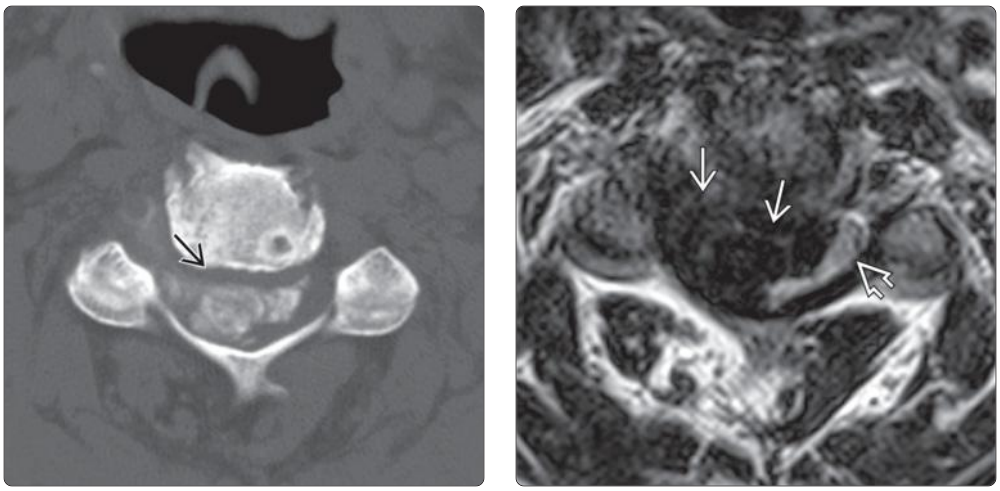
(Left) *Axial NECT shows a densely calcified meningioma within the spinal canal causing severe canal stenosis. Extension into the left neural foramen is an unusual finding for meningioma.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows diffuse enhancement throughout the meningioma. There is both an intraspinal and an intraforaminal component; therefore, this would be considered a dumbbell tumor, for which schwannoma would be a more likely diagnosis.*

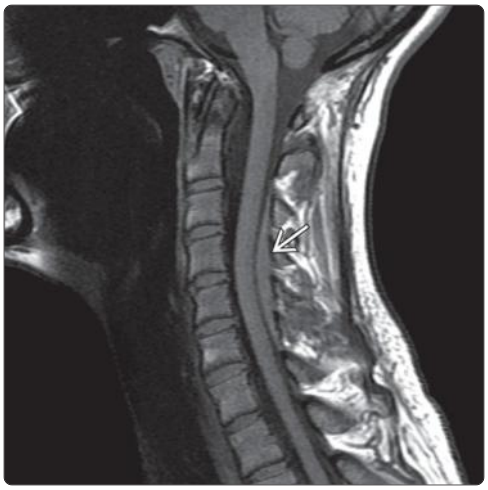


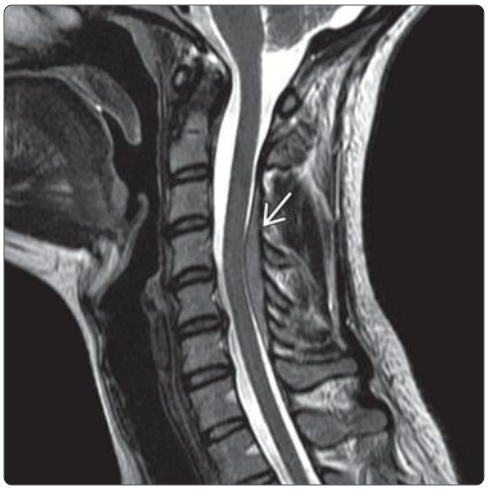
(Left) *Axial CT study shows extensive calcification within a lesion at the cervicothoracic junction consistent with meningioma.* **(Right)** *Sagittal T2 MR shows a large intradural extramedullary mass at the cervicothoracic junction, with marked mass effect upon the cord, which is displaced posteriorly. Low signal in the lesion reflects calcification.*



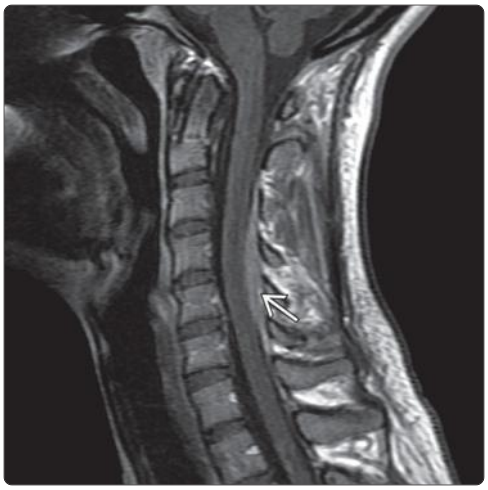
(Left) *Axial bone CT (a variant case of a heavily calcified cervical meningioma) shows dense meningioma calcification with a soft tissue separation from the dorsal aspect of the vertebral body (making this very unlikely to be OPLL).* **(Right)** *Axial T2WI MR (another case of a heavily calcified cervical meningioma) shows a broad ventral dural base of the very low signal meningioma producing severe spinal cord compression.*

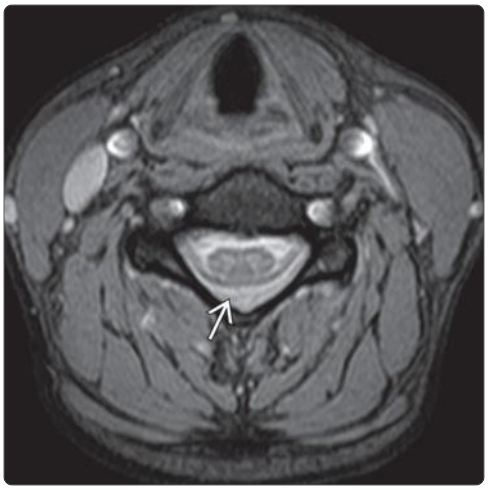




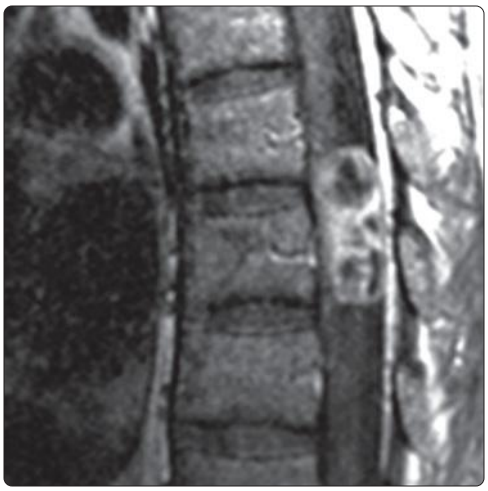


(Left) *Sagittal T1 MR (an uncommon case of extradural meningioma) shows a nonspecific-appearing broad-based mass ⇨ dorsal to the low signal dural margin, with ventral displacement of the cord (path-proven meningioma).* **(Right)** *Sagittal T2WI MR shows a low signal mass ⇨ dorsal to the dural margin in the midcervical spine, favored to be lymphoma prior to surgery. Meningothelial meningioma was found at surgery.*





(Left) *Sagittal T1 C+ MR (a variant case of extradural meningioma) shows a long segment of abnormal homogeneous enhancement ⇨ within the dorsal epidural space.* **(Right)** *Axial T2 GRE MR (a variant case of extradural spinal meningioma) shows a nonspecific appearance to the dorsal epidural mass ⇨, which displaces the cord ventrally.*





(Left) *Sagittal T1WI C+ MR shows an enhancing multicystic mass in a patient with a family history of NF2. The cysts are more consistent with schwannoma than meningioma. The dorsal extraaxial location favors meningioma. Meningioma was found at surgery.* **(Right)** *Sagittal T2WI MR in a patient with a family history of NF2 shows an extramedullary intradural multicystic mass ⇨. Despite similarity to neurinoma, dorsal location favors meningioma, which was proven at surgery.*

Solitary Fibrous Tumor/Hemangiopericytoma

KEY FACTS

TERMINOLOGY

- Hemangiopericytoma (HPC)
- Solitary fibrous tumor (SFT)

IMAGING

- HPC may occur in any compartment of spine except intramedullary
- Circumscribed, multilobulated morphology
- Vivid enhancement characteristically seen

TOP DIFFERENTIAL DIAGNOSES

- Meningioma
- Schwannoma
- Chordoma and other primary bone malignancies
- Aggressive hemangioma
- Vascular metastases
- Angiosarcoma

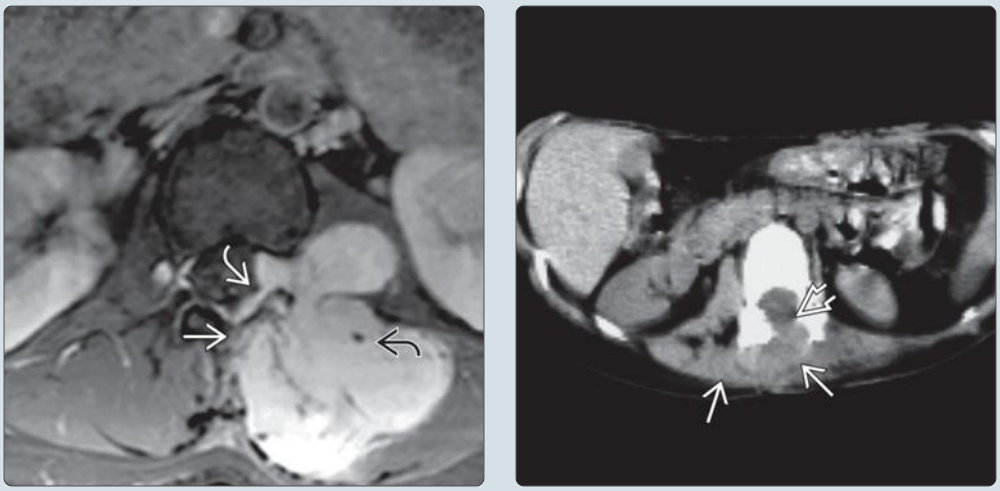
PATHOLOGY

- Hypervascular neoplasm; currently considered mesenchymal tumor of unknown etiology
- Hemangiopericytoma considered part of SFT spectrum ("cellular" or "malignant" SFT)
- Majority of HPC likely classified within cellular end of SFT spectrum

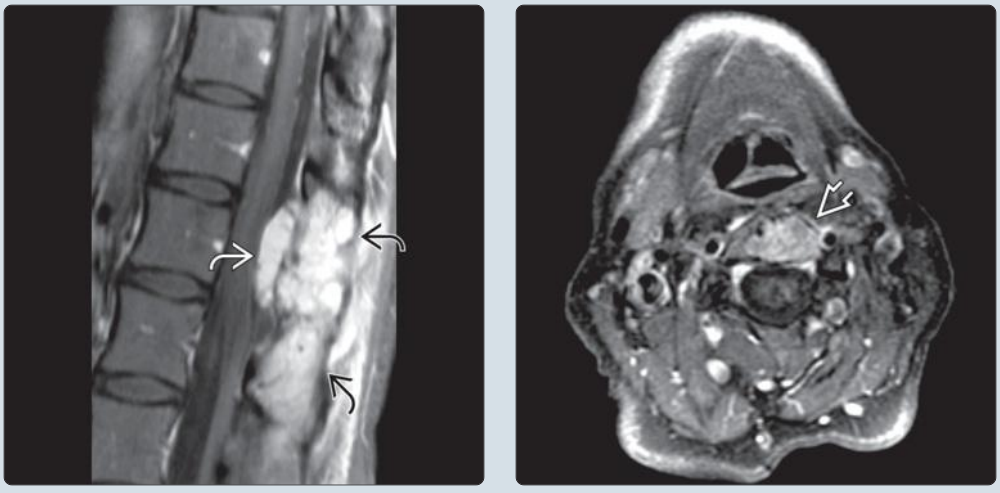
CLINICAL ISSUES

- Natural history
  - Progressive growth
  - Local recurrence
  - Metastases
- Primary treatment is surgical resection
- Radiation therapy as adjuvant to surgery, primary treatment for unresectable tumor
  - Limited benefit of chemotherapy
- Continued radiographic surveillance for late recurrence

(Left) *Axial T1WI C+ MR shows lobulated, avidly enhancing paraspinous mass with internal flow voids due to hypervascularity. There is transforaminal extension with an intradural component. Bony invasion of the lamina is also seen.* **(Right)** *Axial NECT shows dorsal soft tissue mass destroying lamina and extending into dorsal epidural space.*



(Left) *Sagittal T1WI C+ MR shows avidly enhancing mass invading the dorsal elements and adjacent soft tissues at 3 levels of the thoracic spine. An intraspinal component is not clearly in the epidural or intradural extramedullary compartments.* **(Right)** *Axial T1WI C+ MR shows an avidly enhancing osseous metastasis from an intracranial hemangiopericytoma to the C3 vertebral body.*



TERMINOLOGY

Abbreviations

- Hemangiopericytoma (HPC)
- Solitary fibrous tumor (SFT)

Definitions

- Hypervascular neoplasm; currently considered mesenchymal tumor of unknown etiology
- Hemangiopericytoma considered part of SFT spectrum ("cellular" or "malignant" SFT)

IMAGING

General Features

- Best diagnostic clue
  - Vividly enhancing lesion expanding/eroding adjacent osseous structures, with large soft tissue component
- Location
  - In spine, most often dural-based, epidural, or paraspinal
    - Intradural/pial and intramedullary locations also described
  - May see osseous metastases from extraspinal HPC
- Size
  - Variable
- Morphology
  - Circumscribed, multilobulated

Radiographic Findings

- Radiography
  - Erosive soft tissue mass in spinal column or appendages

CT Findings

- NECT
  - Osseous erosion, destruction
  - Expansion of canal by isodense mass with circumscribed borders
  - No phleboliths or calcific components
- CECT
  - Vivid enhancement

MR Findings

- T1WI
  - Hypointense multilobular mass expanding/eroding bone and canal
- T2WI
  - Mild to moderate hyperintensity
- STIR
  - Osseous invasion hyperintense, normal marrow suppressed
- T1WI C+
  - Vivid, homogeneous enhancement

Angiographic Findings

- Conventional
  - Marked hypervascularity in arterial phase with abnormal irregular vessels

Nonvascular Interventions

- Myelography
  - Intradural or extradural mass, effacing contrast column
  - Destructive osseous changes

Nuclear Medicine Findings

- Bone scan
  - "Hot" focal uptake

Imaging Recommendations

- Best imaging tool
  - T1 C+ with fat saturation
- Protocol advice
  - Scan entire neural axis to look for primary, as metastases common, especially in previously treated cases

DIFFERENTIAL DIAGNOSIS

Meningioma

- Usually well circumscribed, nonerosive
- Calcification helpful clue
- Common in women, thoracic spine

Schwannoma

- Avidly enhancing mass in spinal canal, neural foramen
- Hyperintense on T2WI
- Bony remodeling, foraminal enlargement, or vertebral scalloping

Chordoma and Other Primary Bone Malignancies

- Destructive process centered in bone
- Chordoma most commonly seen in sacrum, sinus
- Extraosseous/epidural soft tissue mass

Aggressive Hemangioma

- Thickened vertical trabecula within area of circumscribed osteolysis
- Avid enhancement
- Associated epidural or paravertebral soft tissue mass

Vascular Metastases

- Renal cell, thyroid carcinoma
- Usually centered in cancellous bone, posterior vertebral body commonly involved
- If dural-based, quite focal

Angiosarcoma

- Very rarely in spine
- More haphazard boundaries

PATHOLOGY

General Features

- Etiology
  - Latest WHO classification (2002) abandons traditional pericyte progenitor, favors fibroblastic cell of origin
    - Majority of HPC likely classified within cellular end of SFT spectrum
    - Few remaining tumors with likely pericyte lineage: Some sinonasal HPC, infantile HPC/myofibromatosis, myopericytoma
  - Wide variety of tumors show areas of HPC-like growth
- Immunohistochemistry
  - CD34 (80-90%)
    - Less common in cellular (HPC-like) forms of SFT
  - CD99 (70%)
  - Bcl-2 (30%)
  - SMA (20%)

- S100 usually negative

Gross Pathologic & Surgical Features

- Discrete, vascular mass of dural, epidural space, &/or perivertebral soft tissues

Microscopic Features

- Well-developed, branching/"stag horn," thick-walled vessels surrounded by connective tissue sheath
- Moderate to high cellularity
- Monotonous appearance under light microscopy
- Round to oval, monomorphic nuclei
- Little intervening fibrosis
- Myxoid change, foci of chronic inflammation, interstitial mast cells
- HPC-like histology found in up to 15% of all soft tissue sarcomas

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain
  - Other signs/symptoms
    - Neuropathy
    - Myelopathy

Demographics

- Age
  - 20-70 years of age, median age in the 40s
- Gender
  - No preference
- Epidemiology
  - Rare

Natural History & Prognosis

- Progressive growth
- Local recurrence
- Metastases
  - Lung
  - Liver
  - Bone

Treatment

- Options, risks, complications
  - Surgical resection is mainstay of therapy
  - Radiation therapy as adjuvant to surgery, primary treatment for unresectable tumor
  - Limited benefit of chemotherapy, may be a role for antiangiogenic therapies
  - Continued radiographic surveillance for late recurrence

DIAGNOSTIC CHECKLIST

Consider

- Angiography and embolization for operative planning

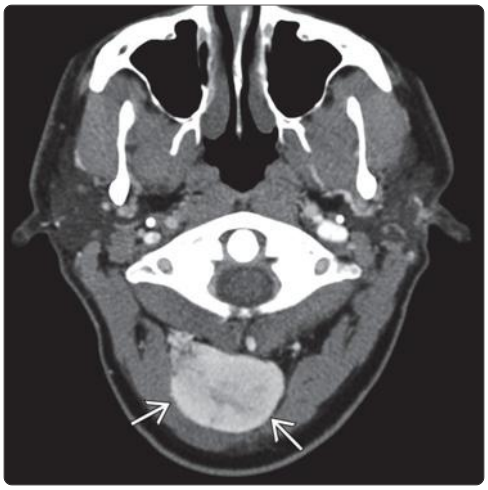
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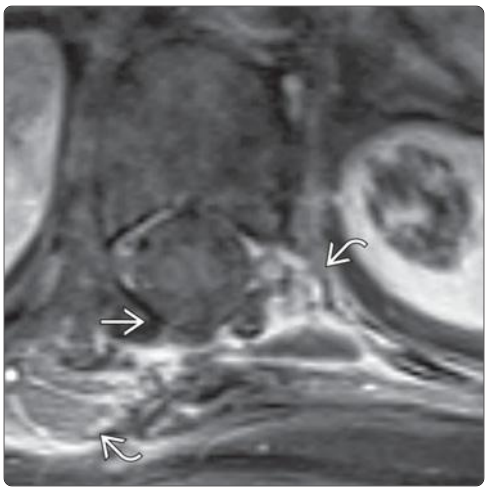


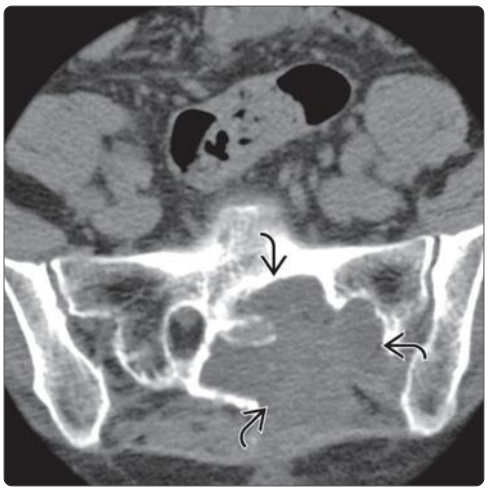
(Left) *Sagittal CT reconstruction shows a destructive process extending into the sacral body from the canal.* **(Right)** *Sagittal T1WI C+ MR in the same patient shows heterogeneously and avidly enhancing HPC-like tumor filling the lower lumbosacral canal, associated with erosion into the sacral body.*





(Left) *Axial CECT shows vividly enhancing, circumscribed soft tissue mass in the dorsal soft tissues of the upper neck.* **(Right)** *Lateral projection DSA in the same patient shows marked hypervascularity of the hemangiopericytoma.*





(Left) *Axial T1WI C+ FS MR shows infiltrative, enhancing soft tissue mass with destruction of the lamina due to local recurrence of HPC of the thoracic spine.* **(Right)** *Axial NECT shows large soft tissue attenuation lesion eroding the sacrum and neural foramen due to osseous metastasis from previously treated intracranial hemangiopericytoma.*

Neurofibroma

KEY FACTS

TERMINOLOGY

- Neoplasm containing Schwann cells, fibroblasts, myxoid material, and peripheral nerve fibers

IMAGING

- Locations
  - Extradural/paraspinal
  - Intradural extramedullary
- Variable involvement of spinal root, neural plexus, peripheral nerve, or end organs
- Size varies from small circumscribed mass to large plexiform NF involving multiple body compartments
- Plexiform neurofibroma pathognomonic for NF1
- Target sign on T2WI suggestive but not pathognomonic for NF
- FDG avidity suggests malignant degeneration

TOP DIFFERENTIAL DIAGNOSES

- Schwannoma
- Spinal meningioma
- Perineural root sleeve cyst
- Chronic interstitial demyelinating polyneuropathy
- Malignant nerve sheath tumors

PATHOLOGY

- Neoplastic Schwann cells + fibroblasts
- Tumor, nerve fascicles intermixed
  - Presence of axons characteristic of NF
- 90% of neurofibromas are sporadic

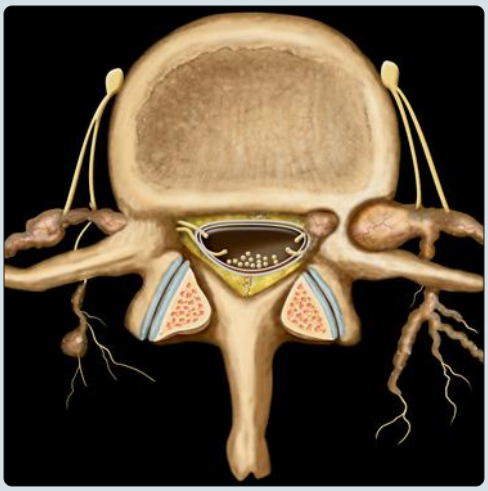
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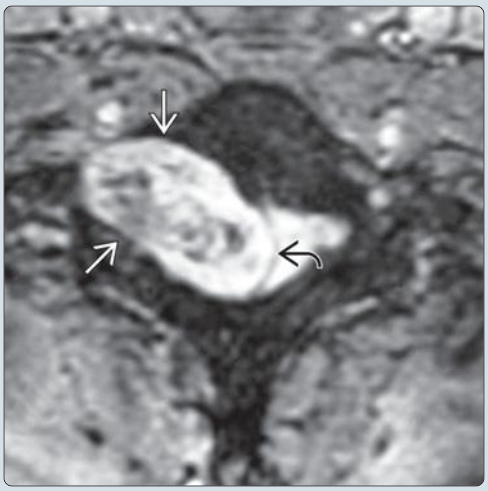
- Malignant transformation of plexiform NF to MPNST in 10% of NF1 patients

DIAGNOSTIC CHECKLIST

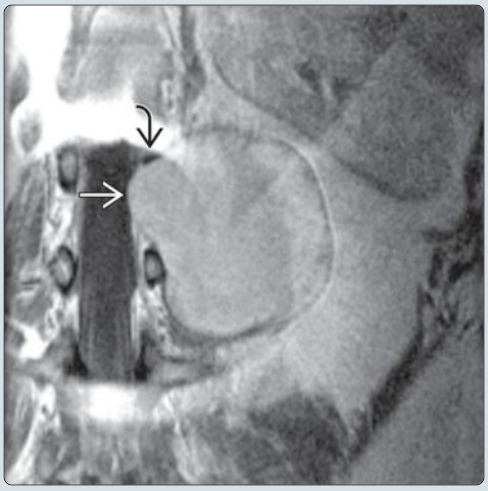
- Rapidly growing NF or atypical pain concerning for malignant transformation to MPNST
- Solitary spinal lesion more likely schwannoma than NF

(Left) *Axial graphic portrays bilateral lobulated plexiform neurofibromas in NF1. There is erosion of the left pedicle by the tumor.* **(Right)** *Axial T2* MR shows a mass centered within and widening the right neural foramen ➡ and the posterolateral margin of the cervical vertebral body. The intraspinal extradural component of the mass ⇨ causes severe canal stenosis. The mass is predominantly hyperintense with some whorled hypointensity centrally.*





(Left) *Coronal T1WI C+ MR shows an enhancing paraspinal mass on the left at L1-2 extending into the left neural foramen, with minimal impact on the thecal sac ➡. Remodeling causes enlargement of the left L1-2 neural foramen with thinning of the L1 pedicle ⇨.* **(Right)** *Axial T2WI in the same patient shows central areas of T2 hypointense signal ➡ with peripheral rim of hyperintensity. This target sign is not specific, but is seen more commonly with neurofibroma than with schwannoma.*





TERMINOLOGY

Abbreviations

- Neurofibroma (NF)

Synonyms

- Nerve sheath tumor (NST)
  - Category of NST also includes schwannomas

Definitions

- Neoplasm containing Schwann cells, fibroblasts, myxoid material, and peripheral nerve fibers

IMAGING

General Features

- Best diagnostic clue
  - Bulky multilevel spinal nerve root tumors in patient with stigmata of neurofibromatosis type 1 (NF1)
- Location
  - Intradural extramedullary
  - Extradural/paraspinal
  - Variable involvement of spinal root, neural plexus, peripheral nerve, or end organs
  - NF1
    - Cervical, lumbosacral
    - Uni- or bilateral
- Size
  - Spectrum: Small, circumscribed mass to large, plexiform NF involving multiple body compartments
- Morphology
  - Localized NF
    - Solitary nodular or fusiform nerve enlargement
    - Typically < 5 cm
    - Sporadic, not associated with NF1
    - 90% of all NFs
  - Diffuse
    - Infiltrative tumor
    - Children and young adults
    - Usually affects subcutaneous tissues of head, neck
    - Spinal nerve involvement rare
    - 90% sporadic, isolated, not associated with NF1
  - Plexiform
    - Long, bulky, multinodular nerve enlargement
    - Interdigitating with surrounding soft tissue
    - Bilateral, multilevel
    - Often affects lumbosacral or brachial plexus
    - Virtually pathognomonic for NF1

Radiographic Findings

- Radiography
  - Bone remodeling due to tumor
    - Neuroforaminal widening
    - Thinned pedicles
    - Vertebral body scalloping

CT Findings

- NECT
  - Soft tissue masses isodense to spinal cord
  - Calcifications rare
- CECT
  - Mild to moderate enhancement
- Bone CT
  - May show enlarged neural foramina, bone remodeling

MR Findings

- T1WI
  - Similar intensity to cord and nerve roots
- T2WI
  - Iso- or hyperintense
  - Target sign suggestive but not pathognomonic for NF
    - Peripherally hyperintense
    - Central hypo- to isointense
  - Hypointense septations throughout plexiform NF
- STIR
  - Hyperintense
- T1WI C+
  - Variable
    - Mild to moderate
    - Relatively homogeneous

Nuclear Medicine Findings

- PET
  - Malignant peripheral nerve sheath tumor (MPNST) metabolically active, elevated FDG uptake compared to benign NF
    - Above 1.8 SUV 1 hour post FDG injection
      - 100% sensitive for MPNST, 83% specific

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR ± contrast
- Protocol advice
  - T2WI with fat suppression or STIR improves lesion detection

DIFFERENTIAL DIAGNOSIS

Schwannoma

- Indistinguishable from solitary NF by imaging
- Target is sign more common with NF than schwannoma
- Hemorrhage, cystic, or fatty degeneration more common with schwannoma

Spinal Meningioma

- 90% intradural
- May see enhancing dural "tail"
- T2 hypointensity due to calcification (< 5%)

Perineural Root Sleeve Cyst

- Cystic nonenhancing mass
- CSF density/signal intensity
- Often opacifies with intrathecal contrast on myelography

Chronic Interstitial Demyelinating Polyneuropathy (CIDP)

- Caused by repeated episodes of demyelination, remyelination
- "Onion skin" layered enlargement of spinal, peripheral nerves
- Mimics plexiform NF on imaging studies
- Features of NF1 absent

Malignant Nerve Sheath Tumors

- Usually arise from malignant transformation of NF
- Heterogeneous T2 signal and enhancement helps distinguish from NF
  - Due to necrosis and hemorrhage

Other Causes of Multiple, Enlarged, Enhancing Spinal Nerves

- Inflammatory neuritis
  - CMV radiculopathy in HIV(+)
  - Mechanical/chemical nerve root irritation
    - Disc herniation
    - Post surgery
  - Guillain-Barré syndrome
- Leptomeningeal carcinomatosis
- Charcot-Marie-Tooth syndrome

PATHOLOGY

General Features

- Genetics
  - Sporadic NF
    - 30-50% are spontaneous
    - Probably due to NF1 gene alteration
  - NF1 (von Recklinghausen disease)
    - Hereditary, with autosomal dominant transmission
    - Essentially 100% penetrance, wide variation in phenotype
    - Incidence: 1:3,000
    - Gene loss or inactivation on long arm of chromosome 17
      - Tumor suppressor, NF1, controls cell growth/proliferation by down-regulating p21-ras

Staging, Grading, & Classification

- NFs: WHO grade I
- MPNSTs: WHO grade II-IV

Microscopic Features

- Neoplastic Schwann cells + fibroblasts
- Tumor, nerve fascicles intermixed
  - Presence of axons characteristic of NF
- Mitotic figures rare in NF
  - High mitotic rate in MPNST
- Immunohistochemistry
  - S100(+) and anti-leu-7(+)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain
  - Weakness
  - Sensory deficit
- Other signs/symptoms
  - Myelopathy
  - NF1 patients: Café au lait spots, skin fold freckling, cutaneous NFs, Lisch nodules

Demographics

- Age
  - Peak presentation: 20-30 years
- Gender
  - No gender predilection
- Ethnicity
  - No racial predilection
- Epidemiology
  - 5% of all benign soft tissue tumors
  - 90% of NFs occur as sporadic, solitary tumors
  - 13-65% of NF1 patients have spinal NFs
    - 57% single lesions
  - About 10% of NF1 patients develop MPNST
  - 50-60% of MPNSTs are associated with NF1

Natural History & Prognosis

- Slow growing
- Malignant transformation of plexiform NF to MPNST in 10% of NF1 patients
  - Rapid growth of NF
  - Pain
  - Rare with sporadic NF

Treatment

- Resection of sporadic/solitary NF
  - > 80% cure rate
- Plexiform NFs difficult to resect
  - High recurrence rate

DIAGNOSTIC CHECKLIST

Consider

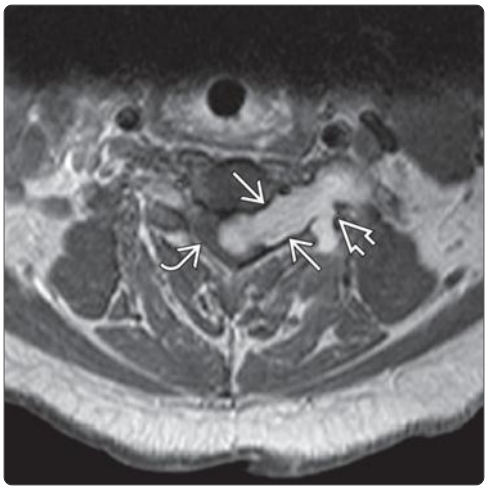
- Rapidly growing NF or atypical pain concerning for malignant transformation to MPNST

Image Interpretation Pearls

- All spinal nerve root tumors are NFs in NF1
  - Schwannomas or mixed tumors in NF2
- Solitary spinal lesion most likely schwannoma

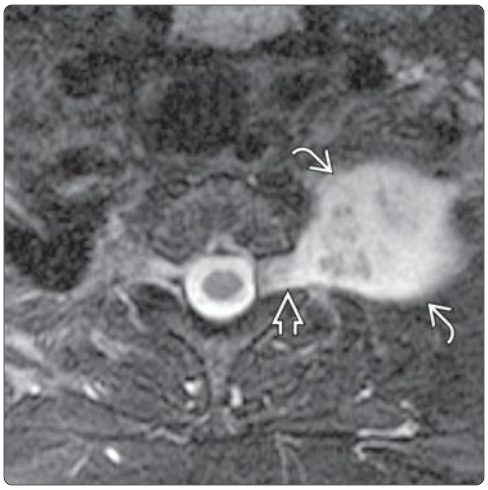
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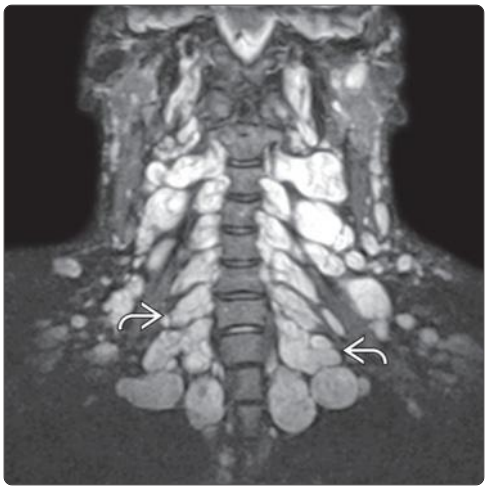
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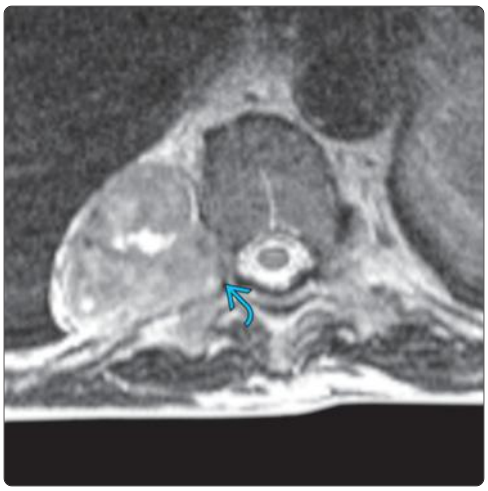


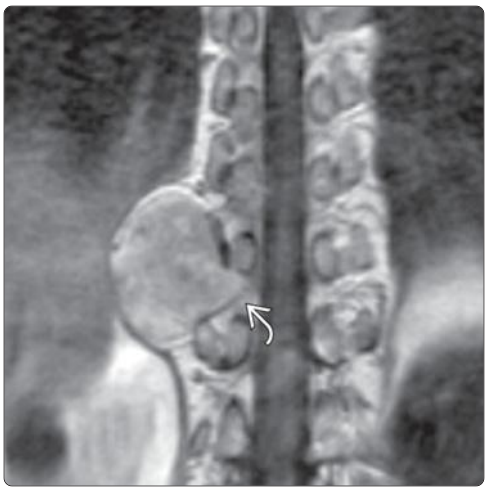
(Left) *Axial T1WI C+ MR shows an oblong enhancing mass extending through the left C4-5 neural foramen. Longstanding remodeling has enlarged the neural foramen. The intraspinal component of the mass causes canal stenosis and cord compression.* **(Right)** *Sagittal T1WI C+ MR of the same patient shows the intraspinal component of the mass. An enhancement pattern is notable for a concentric or target appearance, analogous to the classic target sign seen with neurofibromas on T2WI.*





(Left) *Axial T2WI FS MR shows a hyperintense, paravertebral mass continuous with the nerve root. Signal is mildly heterogeneous.* **(Right)** *Coronal STIR shows extensive, bulky plexiform neurofibromas involving bilateral cervical nerve roots. This appearance is virtually pathognomonic for neurofibromatosis type I.*





(Left) *Axial T2WI MR shows a heterogeneous, circumscribed mass adjacent to the lower right thoracic spine abutting the margin of the neural foramen.* **(Right)** *Coronal T1WI C+ MR in the same patient shows slightly heterogeneous enhancement throughout the mass. The mass extends along right L2 nerve root into the neural foramen, but there is no intraspinal component.*

KEY FACTS

TERMINOLOGY

- Malignant peripheral nerve sheath tumor (MPNST)
- Soft tissue sarcoma that arises from or differentiates toward cells of peripheral nerve sheath

IMAGING

- CT and MR: Large infiltrative, often hemorrhagic, soft tissue mass
  - Arise spontaneously or from malignant degeneration within preexisting neurofibroma
  - Heterogeneous areas correspond to hemorrhage, calcifications, and necrosis
  - Bony erosion, destruction
  - Marked enhancement
  - Indistinct margins
- FDG PET diagnoses neurofibromatosis type 1-associated MPNST with a sensitivity of 89% and a specificity of 95%
  - CT significantly less accurate than FDG PET for characterizing tumors as malignant or benign

TOP DIFFERENTIAL DIAGNOSES

- Benign peripheral nerve sheath tumor
- Other soft tissue sarcoma
- Hematoma

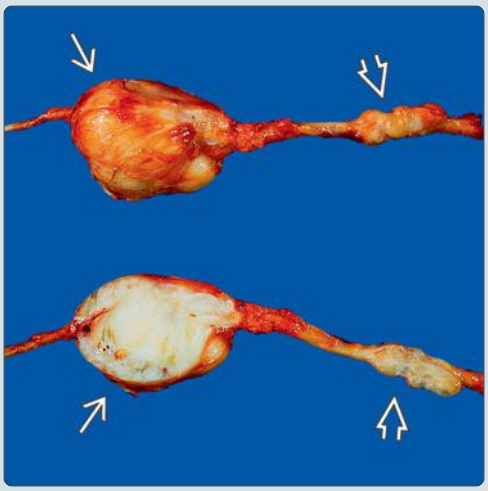
PATHOLOGY

- Incidence in general population: 0.001%
- 50-60% associated with neurofibromatosis type 1 (NF1)

CLINICAL ISSUES

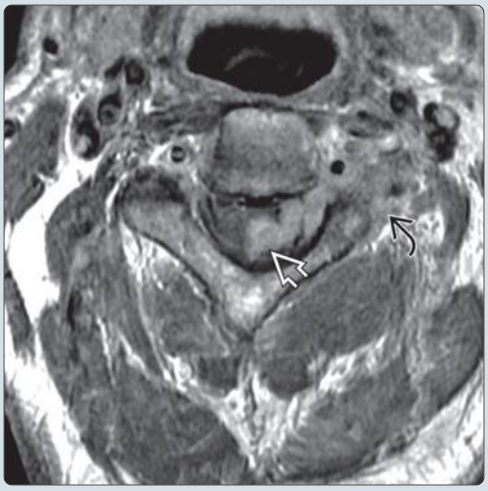
- Enlarging soft tissue mass
- Local or radicular pain, sensory disturbance
- Paraparesis
- Sporadic MPNST presents in 4th decade (mean age: 39.7 years)
- If associated with NF1: 26-42 years (mean age: 28.7)
- Local recurrence: 26-65%; metastases: 20-65%
- Worse prognosis with NF1

(Left) *Malignant peripheral nerve sheath tumors (MPNSTs) often arise from a major nerve trunk, such as this sciatic nerve tumor forming a fusiform, lobulated, intraneural mass. These tumors can extend along a nerve to form satellite nodules.* **(Right)** *Coronal T1 C+ FS MR shows a large heterogeneously enhancing mass in the right pelvis extending through a sacral foramen into the spinal canal and through the sciatic notch into the buttocks.*





(Left) *Axial T1 C+ MR shows enhancing dumbbell-shaped mass involving the cervical spine. There is infiltration of the paravertebral soft tissues, and within the spinal canal, tumor invades the cord parenchyma.* **(Right)** *Axial T1 C+ MR in a patient with NF1 shows neurofibromas (NF) in the left neural foramen and within the spinal canal. Enhancing tumor in the right sacrospinalis muscle has slightly indistinct margins and was pathologically proven to be a MPNST.*





TERMINOLOGY

Abbreviations

- Malignant peripheral nerve sheath tumor (MPNST)
- Neurofibroma (NF)

Synonyms

- Neurofibrosarcoma
- Malignant schwannoma
- Malignant neurilemmoma
- Neurogenic sarcoma

Definitions

- Soft tissue sarcoma that arises from or differentiates toward cells of peripheral nerve sheath

IMAGING

General Features

- Best diagnostic clue
  - Large infiltrative, often hemorrhagic, soft tissue mass arising from plexiform neurofibroma
- Location
  - Paravertebral, rare intraspinal
    - Posterior mediastinum
    - Retroperitoneum
  - Proximal portion of extremities along neurovascular bundle
    - Especially brachial plexus and sciatic nerve
- Size
  - > 5 cm
- Morphology
  - Circumscribed or infiltrative soft tissue mass

Radiographic Findings

- Radiography
  - Deep or superficial soft tissue mass
  - Intervertebral foraminal widening

CT Findings

- NECT
  - Soft tissue mass iso- or hypodense to muscle
  - Heterogeneous areas correspond to hemorrhage, calcifications, and necrosis
  - Bony erosion, destruction
- CECT
  - Marked enhancement

MR Findings

- T1WI
  - Soft tissue mass isointense to muscle
  - Indistinct margins
- T2WI
  - Heterogeneously hyperintense
    - Reflecting hemorrhage, cystic degeneration, calcification
  - Loss of homogeneity, loss of "target" sign within plexiform neurofibroma
- STIR
  - Hyperintense
- T1WI C+
  - Marked enhancement, infiltration of adjacent tissue planes

Angiographic Findings

- Hypervascular soft tissue mass

Nuclear Medicine Findings

- Bone scan
  - Increased radiotracer uptake on blood pool and delayed images
- PET
  - PET can differentiate benign PNSTs from MPNSTs in neurofibromatosis type 1 patients
  - MPNST metabolically active, elevated FDG uptake distinguishes from NF
    - SUVmax threshold of 6.1 g/mL differentiates malignant from benign PNST with a sensitivity and specificity of 94% and 91%, respectively
  - Schwannomas are less reliably discriminated from MPNST by FDG PET imaging
    - Schwannoma may exhibit SUVs > 5 g/mL

Imaging Recommendations

- Best imaging tool
  - FDG PET/CT scans every 6 months for first 2 years and annually thereafter
- Protocol advice
  - T1WI, T2WI , and fat-saturated T1WI C+ MR in multiple planes for soft tissue definition

DIFFERENTIAL DIAGNOSIS

Benign Peripheral Nerve Sheath Tumor

- No invasion of surrounding structures
- Well circumscribed
- Variable enhancement
- Sudden, painful enlargement of preexisting neurofibroma in neurofibromatosis type 1 (NF1) suspicious for MPNST

Other Soft Tissue Sarcoma

- Not necessarily related to neurovascular bundle
- Hemorrhagic and necrotic components
- Intense enhancement

Hematoma

- No enhancing solid components
- T2 hypointensity invariably present
- Surrounding edema

PATHOLOGY

General Features

- Etiology
  - Cells that differentiate toward Schwann cells, perineurial cells, fibrocytes
  - Arise spontaneously or from malignant degeneration within preexisting neurofibroma
    - 50-60% associated with NF1
    - MPNST in patients without NF1 also associated with neurofibroma
  - 11% radiation-induced
    - Latency: 10-20 years
- Genetics

- NF1: von Recklinghausen disease
  - Deletion on long arm of chromosome 17
    - Longer deletion associated with higher incidence of MPNST
    - Autosomal dominant transmission
    - Tumor suppressor: Neurofibromin
  - 50% of cases from new mutations
- Associated abnormalities
  - NF1
    - Café au lait spots
    - Kyphoscoliosis, other vertebral anomalies
    - Sphenoid dysplasia, ribbon-shaped ribs, long bone pseudoarthroses
    - Meningocele, dural ectasia
    - Optic nerve glioma, astrocytoma
    - Lisch nodules

Staging, Grading, & Classification

- Surgical staging
  - Stage I: Low grade
  - Stage II: High grade
  - Stage III: Low or high grade, metastases
    - Within each stage: A = intracompartmental; B = extracompartmental
- Low grade: Perineurial cell differentiation
- High grade: High mitotic rate, ± necrosis, sarcomatous divergent differentiation

Gross Pathologic & Surgical Features

- Large globoid or fusiform mass associated with major nerve
- No true capsule
- Tan-gray, fleshy
- Areas of hemorrhage and necrosis
- Infiltrates surrounding soft tissue
- Spreads along perineurium and epineurium

Microscopic Features

- Fasciculated hyperchromatic mitotically active spindle cells
  - Uncommon variant composed primarily of epithelioid cells
  - Rare divergent differentiation: Striated muscle (Triton tumor), glandular tissue
- Areas of myxoid stroma, mineralization
- May see residual neurofibroma, transition from neurofibroma to sarcoma
- Immunohistochemistry stains for S100 protein, LEU-7, vimentin, and neuron-specific enolase

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Local or radicular pain, sensory disturbance
  - Paraparesis
  - Enlarging soft tissue mass
- Clinical profile
  - Involves major nerve trunks
    - Sciatic nerve, brachial or lumbosacral plexus
  - Sudden enlargement of preexisting neurofibroma in NF1
  - Usually presents as stage IIb lesion (85%)

Demographics

- Age
  - Presents in 4th decade (mean age: 39.7 years)
  - If associated with NF1: 26-42 years (mean age: 28.7 years)
- Gender
  - M = F
  - In NF1: M > F
- Epidemiology
  - Incidence in general population: 0.001%
    - 10% of NF1 patients develop MPNST
  - 50-60% of MPNST patients have NF1
  - 5-10% of all malignant soft tissue tumors

Natural History & Prognosis

- Local recurrence: 26-65%
- Metastases: 20-65%
  - Lung, bone, pleura, liver
- 5-year survival: 10-50%
- NF1 patients have worse prognosis
  - Higher recurrence rate
  - Shorter survival
- Better prognosis associated with
  - Diameter < 5 cm
  - Gross total resection
  - Younger age

Treatment

- Surgical resection
  - Improved resectability with distal lesions
    - 20% in paraspinal MPNST vs. 95% in extremity lesions
  - Resection with wide margins (> 3 cm)
- Pre- or postoperative radiation therapy
- Chemotherapy for metastatic disease effectiveness unproven
- Radiotherapy
  - Local control, little impact on survival

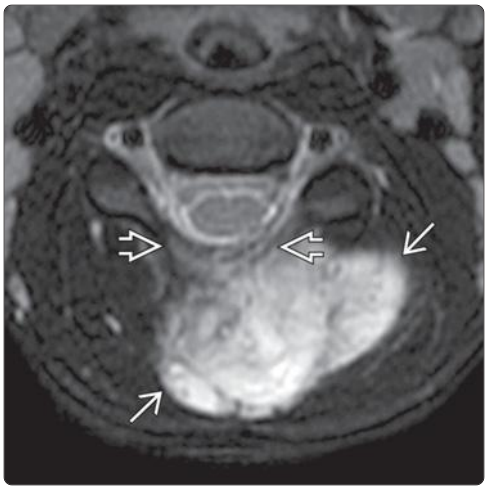
DIAGNOSTIC CHECKLIST

Consider

- Malignant transformation when preexisting plexiform neurofibroma enlarges
- CT significantly less accurate than FDG PET for characterizing tumors as malignant or benign

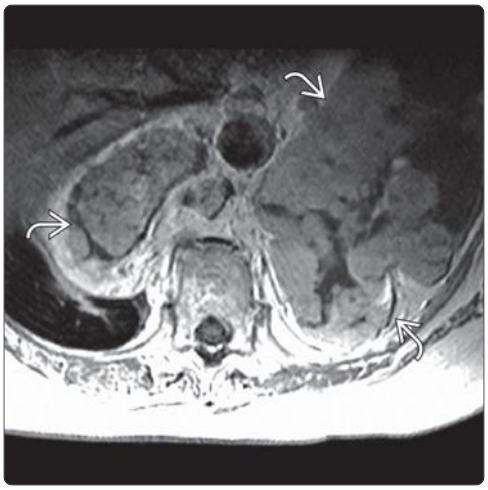
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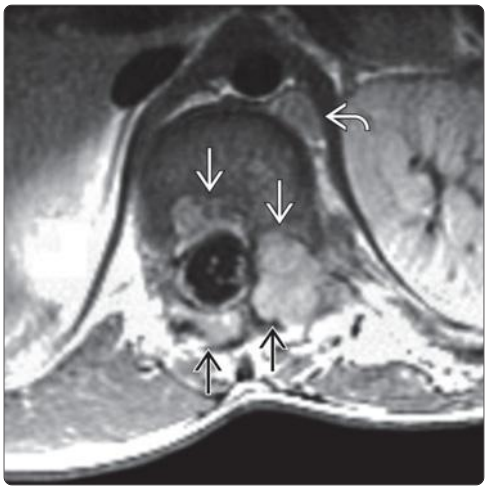
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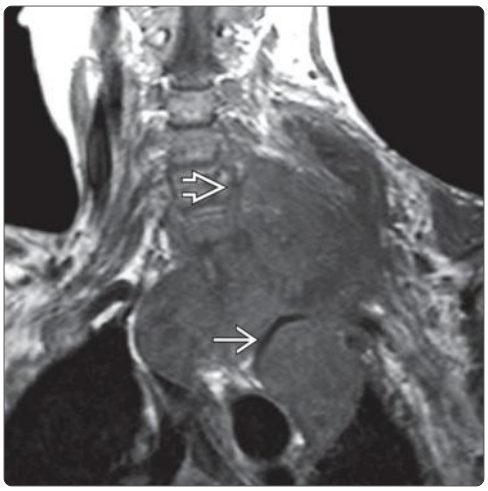


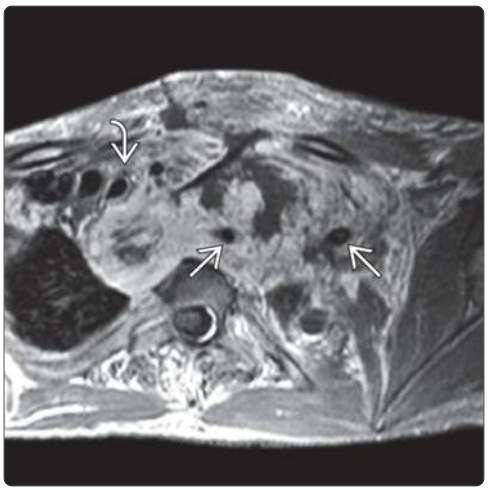
(Left) *Axial T2 FS MR of a MPNST arising within the posterior cervical soft tissues, presumably from a branch of the dorsal primary ramus, shows a large heterogeneously hyperintense mass with infiltrative, destructive changes in the adjacent neural arch.* **(Right)** *Sagittal T1 C+ MR shows a large diffusely enhancing intraspinal MPNST extending from the L1-2 level through the spinal canal. A small circumscribed neurofibroma is seen at the T12 level.*





(Left) *Axial T1 C+ MR shows a large irregularly lobulated MPNST in the retroperitoneum with heterogeneous enhancement.* **(Right)** *Axial T1 C+ MR through the lower thoracic spine shows an enhancing mass centered in left neural foramen, eroding left vertebral body and posterior element. A left retrocrural neurofibroma also enhances.*





(Left) *Coronal T1 MR of a patient with NF1 shows a large MPNST in the upper mediastinum and lower neck, encasing the left subclavian artery and infiltrating the left brachial plexus.* **(Right)** *Axial T1 C+ MR in the same patient shows a large heterogeneously enhancing MPNST in the upper mediastinum, displacing the trachea, encasing the left subclavian artery, and extending into the left supraclavicular fossa. Central irregular area without enhancement was due to necrosis.*

Metastases, CSF Disseminated

KEY FACTS

TERMINOLOGY

- Spread of malignant tumor through subarachnoid spaces of brain and spinal cord

IMAGING

- Smooth or nodular enhancement along cord, cauda equina
- Located at any point along CSF pathway
- 4 basic patterns
  - Solitary focal mass at bottom of thecal sac or along cord surface
  - Diffuse, thin, sheet-like coating of cord/roots ("carcinomatous meningitis")
  - Rope-like thickening of cauda equina
  - Multifocal discrete nodules along cord/roots

TOP DIFFERENTIAL DIAGNOSES

- Multifocal primary tumor
- Pyogenic meningitis
- Granulomatous meningitis
- Chemical meningitis
- Recent lumbar puncture
- Congenital hypertrophic polyradiculoneuropathies
- Thick nerve roots/cauda equina

PATHOLOGY

- Hematogenous dissemination from solid tumors (1-5%)
- "Drop" metastases from patients with primary CNS tumor (1-2%)
- Leukemia, lymphoma (5-15%)
- Falsely negative CSF cytology in up to 40%

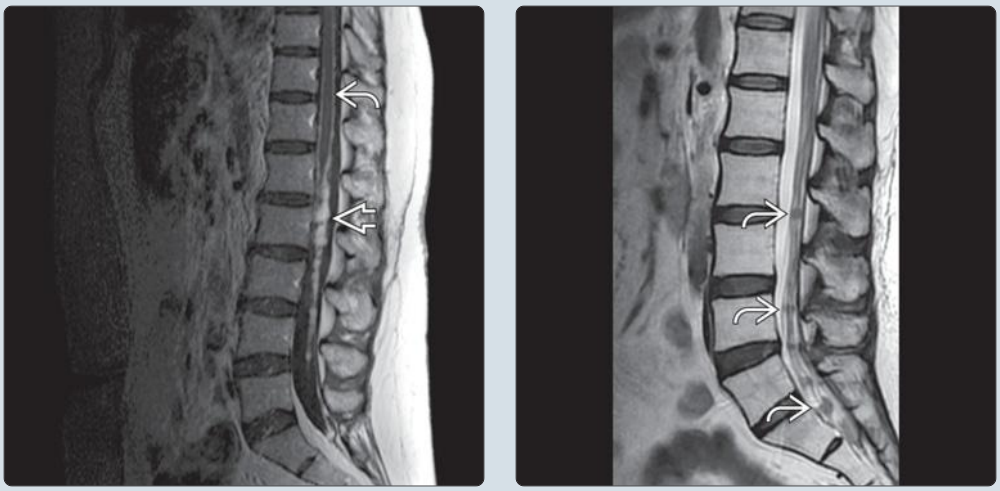
CLINICAL ISSUES

- Typically seen in advanced cancer cases
- Prevalence increasing as cancer patients living longer
- Median patient survival of 3-6 months with treatment, 4-6 weeks without

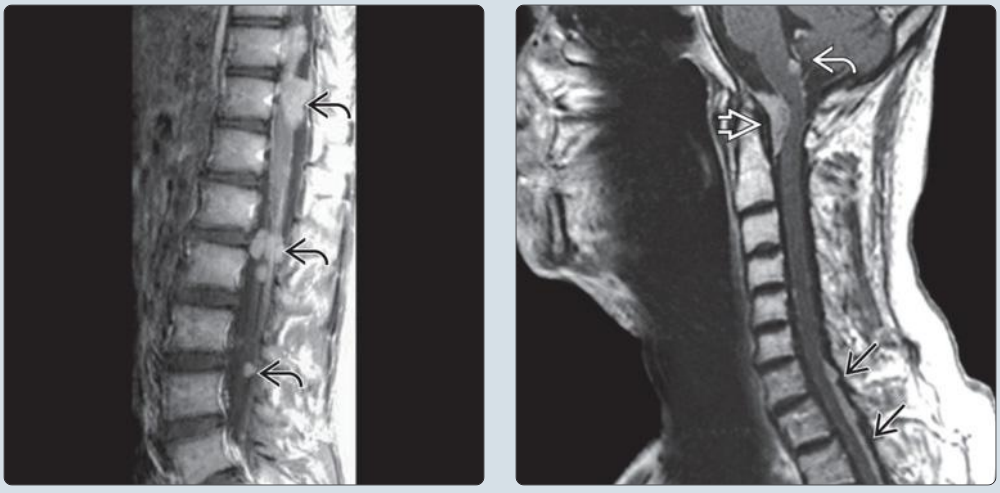
DIAGNOSTIC CHECKLIST

- MR more sensitive than CSF cytology

(Left) *Sagittal T1WI C+ MR shows a mixed nodular and smooth leptomeningeal tumor along the distal cord and conus medullaris due to drop metastases from glioblastoma multiforme.* **(Right)** *Sagittal T2WI shows multiple nodular low signal intensity rounded drop metastases involving roots of the cauda equina, spreading from an intracranial glioblastoma multiforme.*



(Left) *Sagittal T1WI C+ MR shows bulky nodular drop metastases from a pineal germinoma along the conus and cauda equina.* **(Right)** *Sagittal T1WI C+ MR shows CSF metastasis from small cell lung carcinoma with a bulky enhancing dural mass compressing the cervicomedullary junction and linear enhancing metastases along the dorsal pial surface of the cord from C7 caudally. An additional enhancing focus is seen in the region of the gracile tubercle.*



TERMINOLOGY

Synonyms

- Leptomeningeal carcinomatosis
- Carcinomatous meningitis
- Neoplastic meningitis
- Drop metastases

Definitions

- Spread of malignant tumor through subarachnoid spaces of brain and spinal cord

IMAGING

General Features

- Best diagnostic clue
  - Smooth or nodular enhancement in basilar cisterns, along cord (especially dorsally) and cauda equina
- Location
  - Any point along CSF pathways
- Size
  - Variable
- Morphology
  - 4 basic patterns
    - Solitary focal mass at bottom of thecal sac or along cord surface
    - Diffuse, thin, sheet-like coating of cord/roots (carcinomatous meningitis)
    - Rope-like thickening of cauda equina
    - Multifocal discrete nodules along cord/roots
  - Intramedullary nodule(s)
    - Rarely due to CSF spread, impossible to distinguish from hematogenous metastatic disease

CT Findings

- NECT
  - Often normal
  - Metastases may be seen in vertebral column, regional soft tissues
- CECT
  - May be normal
  - May be able to appreciate enhancement of cauda equina

MR Findings

- T1WI
  - Metastases usually isointense with cord, roots
  - Tumor may fill thecal sac, elevating normally low CSF signal
    - Nerve roots, dura appear blurred, "smudged"
    - CSF has ground-glass appearance
- T2WI
  - Thickened nerve roots
  - Tumor isointense or hyperintense to cord, nerve roots
  - May see vasogenic edema in cord
- T1WI C+
  - Variable enhancement
    - Single or multiple enhancing, nodular masses
    - "Sugar coating" of cord, roots
    - Rounded intramedullary mass

Nonvascular Interventions

- Myelography
  - Expanded cord, thickened nerve roots
  - Nodular or mass-like filling defects in contrast column
  - Myelographic block

Imaging Recommendations

- Protocol advice
  - With high-risk CNS tumors, image entire neuraxis for metastatic disease prior to craniotomy
    - High-resolution T2WI and STIR
    - Contrast-enhanced T1WI ± fat suppression

Nuclear Medicine Findings

- Radionuclide cisternography
  - Communicating hydrocephalus
  - CSF flow obstruction in spinal canal, skull base from subarachnoid tumor

DIFFERENTIAL DIAGNOSIS

Multifocal Primary Tumor

- Hemangioblastoma
- Astrocytoma (rare)
- Myxopapillary ependymoma

Pyogenic Meningitis

- Typically chronic bacterial or fungal meningitis
- Subdural empyema
- Clinical/laboratory findings helpful

Granulomatous Meningitis

- Tubercular meningitis
- Fungal meningitis
- Sarcoidosis

Chemical Meningitis

- Postoperative change
- Subarachnoid blood, adhesions can mimic leptomeningeal mets

Recent Lumbar Puncture

- Leptomeningeal, dural enhancement

Congenital Hypertrophic Polyradiculoneuropathies

- Charcot-Marie-Tooth
- Dejerine-Sottas

Thick Nerve Roots/Cauda Equina

- Guillain-Barré syndrome
  - Post-viral or vaccinial ascending paralysis
  - Predominantly motor signs
- AIDS-associated polyneuropathy (e.g., CMV)
- Chemotherapy-associated polyneuropathy
- Chronic interstitial demyelinating polyneuropathy (CIDP)

PATHOLOGY

General Features

- Etiology
  - Hematogenous spread (Batson plexus, arterial) from solid tumors (1-5%)
    - Most common: Adenocarcinomas (lung, breast), melanoma, small cell lung carcinoma
    - Unknown primary in 1-7%

- "Drop" metastases from patients with primary CNS tumor (1-2%)
  - Adults: Anaplastic astrocytoma, glioblastoma multiforme (GBM) (0.5-1% of cases), ependymoma
  - Children: Most common - PNET (medulloblastoma)
    - Others: Germinoma, ependymoma, choroid plexus papilloma/carcinoma
- Leukemia, lymphoma (5-15%)
- Associated abnormalities
  - Elevated CSF protein, leukocytes; decreased glucose

Microscopic Features

- Varies with histology of primary neoplasm
- CSF cytology negative in up to 40% of patients with autopsy-proven leptomeningeal carcinomatosis
  - Yield increases with
    - 2 samples
    - Adequate CSF volumes (> 10.5 cc)
    - Sampling from point on neuraxis near to clinical/radiographic disease

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Headache
  - Confusion, cognitive impairment, seizures
  - Cranial nerve dysfunction
    - Diplopia, especially CN VI
    - Trigeminal neuralgia, anesthesia
    - Optic neuropathy
    - Deafness
  - Weakness, legs more frequently than arms
  - Dermatomal or segmental sensory loss
  - Radicular pain
  - Nuchal rigidity (15%)
  - Disturbance of bowel, bladder function
- Other signs/symptoms
  - Abnormal CSF flow dynamics in 30-70%, with obstruction at level of
    - Spine
    - Skull base
    - Arachnoid granulations (communicating hydrocephalus)
- Clinical profile
  - Typically seen in advanced cancer cases

Demographics

- Epidemiology
  - 5% of all spinal metastases
  - Prevalence increasing as cancer patients living longer

Natural History & Prognosis

- Relentless progression typical
- Median patient survival of 3-6 months with treatment, 4-6 weeks without

Treatment

- Radiation therapy for focal tumor
- Intrathecal chemotherapy

DIAGNOSTIC CHECKLIST

Consider

- MR more sensitive than CSF cytology; especially fat-saturated T1 C+

Image Interpretation Pearls

- Postoperative enhancement of meninges can mimic leptomeningeal tumor spread

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(Left) *Sagittal T1WI MR shows diffusely abnormal increased CSF signal in the visualized thecal sac. Further imaging demonstrated leptomeningeal carcinomatosis due to spread from an intracranial glioma.* **(Right)** *Sagittal T2WI MR shows drop metastasis from a 4th ventricle ependymoma, presenting as a slightly inhomogeneous mass in the caudal thecal sac.*





(Left) *Sagittal T1 C+ MR shows an intradural extramedullary lesion at the C5-6 level in this patient with a path-proven metastatic lesion from intracranial GBM. Note the lack of diffuse leptomeningeal enhancement.* **(Right)** *Sagittal T1WI C+ MR of the spine, performed during follow-up after resection of a nonenhancing GBM of the right insula, shows multiple nonenhancing drop metastases along the surface of the cord.*





(Left) *Sagittal T1WI C+ MR of the cervical spine shows an enhancing high-grade oligodendroglioma in the midbrain. Enhancing, nodular, and confluent drop metastases are seen compressing the cord in the cervical spine.* **(Right)** *Sagittal T1WI C+ MR of the lumbar spine shows bulky nodular drop metastases in the lumbar canal, extending throughout the fibers of the cauda equina.*

Paraganglioma

KEY FACTS

TERMINOLOGY

- Synonyms: Glomus tumor, extraadrenal pheochromocytoma, chemodectoma

IMAGING

- Hypervascular, intensely enhancing intradural extramedullary mass
  - Prominent flow voids due to enlarged draining veins
  - ± cystic areas, hemosiderin from prior hemorrhage
- Large tumors may show osseous remodeling
- Malignant paraganglioma may → osteolytic spinal metastases
- Usually positive on I-123 or I-131 MIBG scan

TOP DIFFERENTIAL DIAGNOSES

- Myxopapillary ependymoma
  - Usually indistinguishable from paraganglioma on imaging studies
- Schwannoma
  - Rarely associated with dilated vessels
- Meningioma
- Metastasis
- Hemangioblastoma of filum terminale

PATHOLOGY

- WHO grade I
- Slow-growing, generally benign behavior

CLINICAL ISSUES

- Presents with back pain/radiculopathy (often chronic)
- Prognosis varies with tumor location but generally excellent

DIAGNOSTIC CHECKLIST

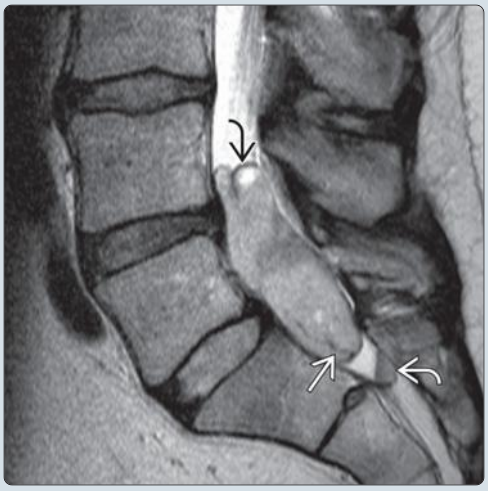
- MR imaging features nonspecific
- Ependymoma and schwannoma much more common than paraganglioma

(Left) *Sagittal T1WI C+ MR of the lumbar spine demonstrates a large, homogeneously enhancing intradural mass filling the lumbar canal from T12 through L4.* **(Right)** *Axial T1WI C+ MR of the same patient shows posterior vertebral body scalloping, neuroforaminal enlargement, and thinning of the left pedicle secondary to longstanding osseous remodeling. Recognition of these osseous changes confirms the slow-growing nature of the tumor.*





(Left) *Sagittal T2WI MR reveals a heterogeneous mass within the caudal thecal sac, demonstrating a cystic area superiorly and a prominent flow void indicating hypervascularity. A small amount of hemorrhage layers dependently in the caudal tip of the thecal sac.* **(Right)** *Sagittal T1WI C+ MR in the same patient confirms diffuse, mildly heterogeneous enhancement throughout the tumor. Mild scalloping of the posterior L5 vertebral body is also noted, indicating chronic remodeling.*





TERMINOLOGY

Synonyms

- Glomus tumor
- Extraadrenal pheochromocytoma
- Chemodectoma

Definitions

- Paraganglioma presenting as intradural extramedullary mass within cauda equina

IMAGING

General Features

- Best diagnostic clue
  - Hypervascular, enhancing intradural mass involving cauda equina mass
- Location
  - Spine is rare extraadrenal site of paraganglioma
  - Primary spinal paraganglioma most often occurs within filum terminale
  - Other spinal paragangliomas
    - Malignant paraganglioma may result in osteolytic spinal metastases
    - There are scattered reports of primary intraosseous and extradural paragangliomas
- Size
  - 10 mm to > 50 mm
- Morphology
  - Well-defined intradural extramedullary mass

CT Findings

- NECT
  - Large intradural tumors may show bony remodeling
    - Vertebral scalloping
    - Thinning of pedicles
    - Foraminal enlargement
- CECT
  - Enhancing intradural mass with large draining veins

MR Findings

- T1WI
  - Extramedullary mass in distal thecal sac
  - Iso- to hypointense relative to cord
- T2WI
  - Heterogeneously hyperintense relative to cord
  - May see cystic areas
  - May see hemosiderin from prior hemorrhage
  - Prominent flow voids due to enlarged draining veins
- T1WI C+
  - Intense enhancement
  - Tends to be mildly inhomogeneous

Angiographic Findings

- Hypervascular mass with large draining veins

Nonvascular Interventions

- Myelography
  - Smoothly lobulated intradural extramedullary mass
  - ± serpentine filling defects due to enlarged intrathecal vessels

Nuclear Medicine Findings

- > 90% paraganglioma positive on radioiodine MIBG scan
- May also show octreotide uptake due to somatostatin receptors

Imaging Recommendations

- Contrast-enhanced MR (image entire spine to evaluate for "uphill" metastases)

DIFFERENTIAL DIAGNOSIS

Myxopapillary Ependymoma

- Usually indistinguishable from paraganglioma on imaging studies
- Much more common tumor in cauda equina

Schwannoma

- Rarely hemorrhage
- Rarely associated with dilated vessels
- Much more common tumor in cauda equina

Meningioma

- Dural-based mass
- Unusual in lumbar spine
- Not associated with dilated vessels

Metastasis

- Consider drop metastases within differential for enhancing intradural mass

Hemangioblastoma of Filum Terminale

- Rare location for hemangioblastoma
- Hypervascular, enlarged vessels

PATHOLOGY

General Features

- Etiology
  - Arise from neural ectoderm
  - Typical distribution of extraadrenal pheochromocytoma parallels autonomic nervous system
  - Histogenesis of spinal paraganglioma debated
    - Paraganglionic tissue not normally found in cauda equina
    - Peripheral neuroblasts in filum may undergo paraganglionic differentiation
- Genetics
  - No reports of familial spinal paraganglioma, unlike nonspinal extraadrenal paraganglioma

Staging, Grading, & Classification

- WHO grade I
- Histological evidence of malignancy in 6.5% of paraganglioma

Gross Pathologic & Surgical Features

- Encapsulated, soft, dark red-brown tumor
- Richly vascular

Microscopic Features

- Well-differentiated tumor (resembles normal paraganglia)
  - Chief (type I) cells arranged in compact nests ("Zellballen")

- Surrounded by inconspicuous single layer of sustentacular (type II) cells
- Round/oval nuclei with finely stippled chromatin, indistinct nucleoli
- Sinusoidal blood vessels (occasionally thick-walled, hyalinized)
- Immunohistochemical staining
  - Chromogranin and synaptophysin most important markers for paraganglioma
  - Helpful to differentiate unusual paraganglioma variants from ependymoma, metastatic carcinoma
- Electron microscopy shows dense core neurosecretory granules

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Presents with back pain/radiculopathy (tends to be chronic)
  - Approximately 1-3% of paraganglioma have secretory activity (norepinephrine, epinephrine)
    - Headaches, facial flushing
    - Hypertension, palpitations, and diaphoresis
    - Urinary catecholamine levels elevated
- Other signs/symptoms
  - Sensory/motor loss, bowel/bladder dysfunction
  - Few reports of increased intracranial pressure
- Clinical profile
  - Middle-aged patient with myelopathy, intradural extramedullary enhancing mass

Demographics

- Age
  - Range: 13-70 years
  - Average: 45-50 years
- Gender
  - M = F
- Epidemiology
  - Most neural crest tumors occur in adrenal medulla (pheochromocytoma)
  - 80-90% of extraadrenal paragangliomas occur in/near carotid body, jugular bulb
  - Spine is uncommon site

Natural History & Prognosis

- Slow-growing, generally benign behavior
- Varies with tumor location (generally excellent for spinal paraganglioma)
- Recurrence < 5% after gross total removal

Treatment

- May require preoperative embolization to minimize intraoperative hemorrhage
- Surgical excision usually curative

DIAGNOSTIC CHECKLIST

Consider

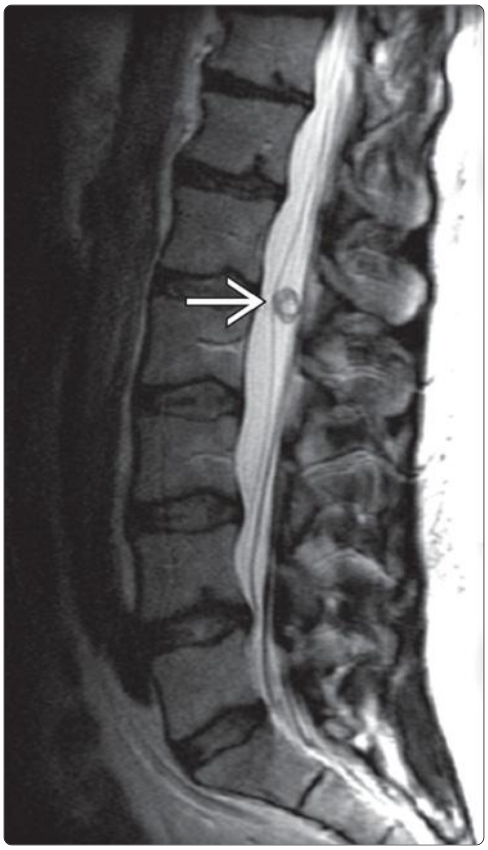
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Image Interpretation Pearls

- Ependymoma and schwannoma much more common

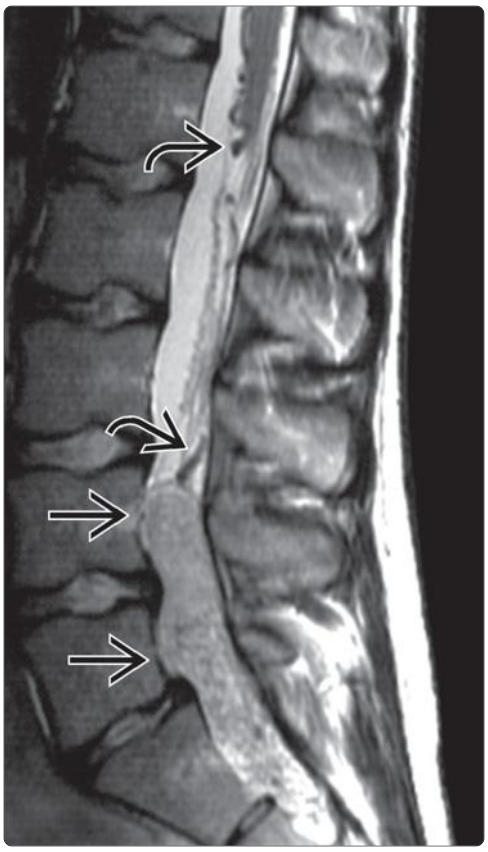
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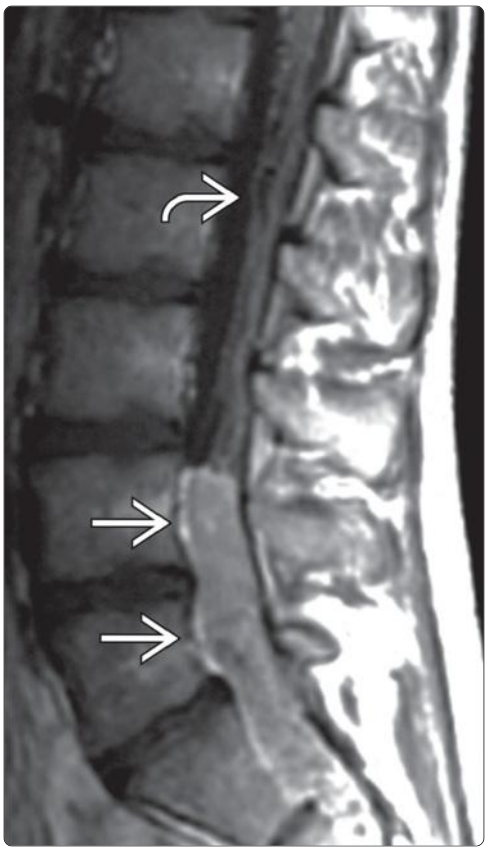
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(Left) *Sagittal T2WI MR demonstrates a small, solitary nodular intradural mass ➡ interposed between the cauda equina nerves within the lumbar canal at the L1-L2 level. The mass is heterogeneously hyperintense.* **(Right)** *Sagittal T1WI C+ FS MR of the same patient reveals diffuse enhancement of the intradural mass ➡. No additional pathological enhancement to indicate tumor dissemination is demonstrated.*





(Left) *Sagittal T2WI MR depicts a hyperintense intradural extramedullary mass within the caudal osseous spinal canal. Mild scalloping of the dorsal L4 and L5 ⇨ vertebral bodies due to chronic osseous remodeling is noted. Enlarged vessels ⇨ are seen in the thecal sac, along the surface of the cauda equina and conus medullaris, confirming a hypervascular tumor.* **(Right)** *Sagittal T1WI C+ MR in the same patient demonstrates diffuse enhancement throughout the intradural mass. The posterior vertebral scalloping ➡ indicates slow tumor growth giving chronic osseus remodeling. Multiple enlarged vessels ➡ confirm the hypervascular nature of the intradural mass.*

Astrocytoma

KEY FACTS

TERMINOLOGY

- Primary neoplasm of astrocytic origin within spinal cord

IMAGING

- Fusiform expansion of cord with enhancing component of variable morphology
  - Almost always enhances
- Cervical > thoracic
- Usually ≤ 4 segments
  - Occasionally multisegmental, even holocord (more common with pilocytic astrocytoma)
- ± cyst/syrinx (fluid slightly T1 hyperintense to CSF)
- Hyperintense on proton density and T2WI MR
- Myelopathy should be evaluated with contrast-enhanced MR

TOP DIFFERENTIAL DIAGNOSES

- Ependymoma
- Hemangioblastoma
- Syringohydromyelia
- Autoimmune or inflammatory myelitis

PATHOLOGY

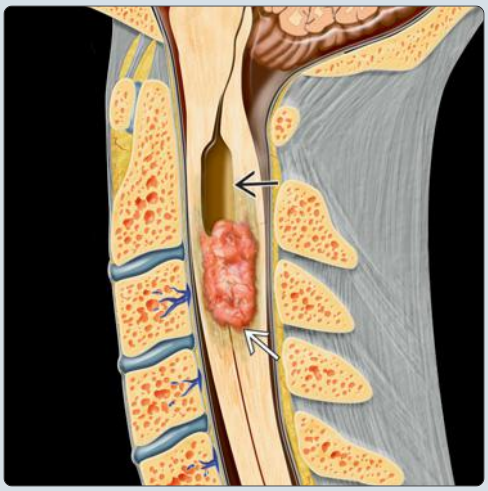
- 80-90% low grade
- 10-15% high grade

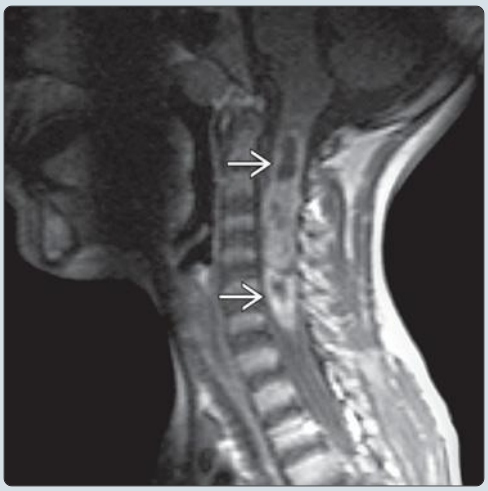
CLINICAL ISSUES

- Slow onset of myelopathy
- May cause painful scoliosis
- Most are slow growing
  - Malignant tumors may cause rapid neurologic deterioration
- Most common intramedullary tumor in children and young adults
- Overall, ependymomas > astrocytomas (2:1)

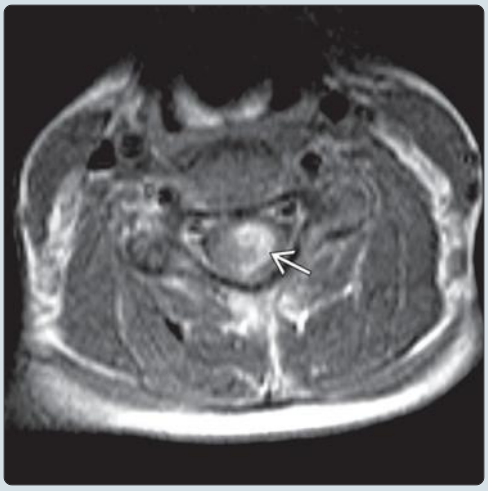
DIAGNOSTIC CHECKLIST

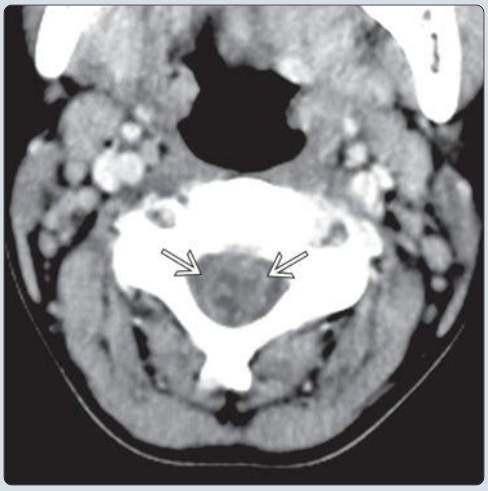
- Myelopathy should be evaluated with MR





(Left) *Sagittal graphic of cervical spine astrocytoma shows a fusiform solid mass ➡ with a rostral cystic cord component ⇨. (Right) Sagittal T1WI C+ MR reveals a heterogeneously enhancing cervical cord mass ➡. The tumor subtype and histological grade are the most important prognostic factors. Malignant transformation, while common in recurrent adult low-grade gliomas, is unusual in pediatric low-grade intramedullary spinal cord tumors.*





(Left) *Axial T1WI C+ MR depicts an intramedullary enhancing tumor in the cervical cord ➡. Astrocytomas tend to have an eccentric growth pattern and are T2 hyperintense. Up to 1/3 are reportedly nonenhancing but have mass effect and cord expansion. (Right) Axial CECT illustrates a heterogeneously enhancing mass ➡ within the central spinal canal in this case of cervical spinal cord astrocytoma with holocord edema (not shown).*

TERMINOLOGY

Definitions

- Primary neoplasm of astrocytic origin within spinal cord (SC)

IMAGING

General Features

- Best diagnostic clue
  - Enhancing infiltrating mass expanding SC
- Location
  - Cervical > thoracic
- Size
  - Usually ≤ 4 segments
  - May be extensive, especially with pilocytic histology
- Morphology
  - Fusiform spinal expansion, with enhancing component of variable morphology
  - Occasionally asymmetric, even exophytic
    - Eccentric > central growth pattern

Radiographic Findings

- Radiography
  - ± scoliosis
  - ± expansion of osseous canal

CT Findings

- NECT
  - Enlarged SC
  - ± expansion, remodeling of osseous canal
- CECT
  - Mild/moderate enhancement

MR Findings

- T1WI
  - Cord expansion
    - Usually < 4 segments
    - Occasionally multisegmental, even holocord (more common with pilocytic astrocytoma [PA])
  - ± cyst/syrinx (fluid slightly hyperintense to cerebrospinal fluid [CSF])
  - Solid portion hypo-/isointense
    - Minority of cases may have areas of hyperintensity (methemoglobin)
- T2WI
  - Hyperintense
- T2* GRE
  - Hyperintense
  - Minority of cases may have hypointense areas if hemorrhagic products
- DWI
  - ↓ fractional anisotropy, ↑ ADC
- T1WI C+
  - Usually enhance (partial > total)
  - Mild/moderate > intense enhancement
  - Heterogeneous/infiltrating > homogeneous/sharply delineated

Nuclear Medicine Findings

- Anaplastic astrocytomas demonstrate high uptake of FDG-18 and 11C-methionine

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced multiplanar MR
- Protocol advice
  - Sagittal and axial T2WI and T1WI C+ MR

DIFFERENTIAL DIAGNOSIS

Ependymoma

- Intense, sharply delineated enhancement
- Central > eccentric growth pattern
- More often seen in low thoracic cord
- Cystic or necrotic component, hemorrhage common
- Typically older patients than those with astrocytoma

Other Neoplasms

- **Ganglioglioma**
  - Mixed T1 signal intensity due to solid, cystic components
  - Homogeneous > heterogeneous T2 hyperintensity
- **Lymphoma**
  - Intramedullary form presents as poorly defined, enhancing lesion
- **Metastasis**
  - Intramedullary nidus of enhancement much more focal with extensive edema
  - Pial metastasis can simulate hemangioblastoma
  - Usually older patients
- **Hemangioblastoma**
  - Focal, enhancing pial/subpial nodule
  - Associated syrinx simulates astrocytoma

Syringohydromyelia

- Cyst fluid similar to CSF; no enhancement

Autoimmune or Inflammatory Myelitis

- Demyelinating disease (± patchy, ill-defined enhancement if acute)
  - Multiple sclerosis
    - Cord may show mild focal swelling, flame-shaped
    - Typically multifocal
  - Transverse myelitis
    - Long cord segment ± patchy enhancement
  - Infectious myelitis
    - Viral myelitis, granulomatous osteomyelitis, bacterial meningitis
    - Rapid onset, constitutional signs
- Spinal cord infarction
  - Abrupt onset
  - Risk factors: Atherosclerosis, hypertension, diabetes
  - Clinical setting: Aortic dissection, abdominal aortic aneurysm, or surgery
- Dural vascular malformation
  - Typically, cord shows mild edema and enlargement
  - Prominent pial vessels
  - Slow onset of upper leg weakness

PATHOLOGY

Staging, Grading, & Classification

- 80-90% low grade
  - Pilocytic astrocytoma = WHO grade I

  - Focal or diffuse cord-enlarging, irregularly-shaped masses with cystic elements and hydromyelia
  - Propensity for thoracic and cervical regions
  - Different degrees of enhancement, high water diffusivity
- Fibrillary astrocytoma = WHO grade II
- Pilomyxoid astrocytoma (similar to PA but with distinct histopathological characteristics)
  - Rare in SC
  - More aggressive behavior than PA
  - ± CSF dissemination at presentation
- Pleomorphic xanthoastrocytoma
  - Rare in SC
  - Favorable prognosis; however, 15-20% undergo progressive anaplastic transformation
  - Superficially located and partly infiltrates leptomeninges
- Ganglioglioma, mixed gliomas

- 10-15% high grade
  - Most are anaplastic astrocytomas (WHO grade III)
  - Glioblastoma (WHO grade IV) uncommon
  - Higher grades (WHO grades III-IV) exhibit high rate of local and distant recurrence throughout neuraxis

Microscopic Features

- Fibrillary astrocytoma
  - Hypercellularity, variable atypia/mitoses
  - Parenchymal infiltration
- Pilocytic astrocytoma
  - Rosenthal fibers, glomeruloid/hyalinized vessels
  - Low prevalence of nuclear atypia/mitoses
- Anaplastic astrocytoma
  - Hypercellularity
  - Nuclear atypia and mitotic activity
  - Absent microvascular proliferation and necrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Slow onset of myelopathy
- Other signs/symptoms
  - Painful scoliosis
  - Radiculopathy
  - Sensory or motor deficits
  - Incontinence

Demographics

- Age
  - Most common intramedullary tumor in children/young adults
  - 60% of intramedullary SC tumors (IMSCTs) in children are astrocytomas, 30% are ependymomas
- Gender
  - M:F = 1.3:1
- Epidemiology
  - 2nd most common SC neoplasm overall
  - IMSCTs = 5-10% of all CNS tumors
    - 20% of intraspinal neoplasms in adults
    - 30-35% of intraspinal neoplasms in children
  - 90-95% of IMSCTs are gliomas
    - Overall, ependymomas > astrocytomas (2:1)
    - Compared to PAs in other sites, cord PAs occur in older patients and comprise large proportion (58%) of spinal astrocytic tumors
  - M:F = 1.3:1
  - Association with NF2

Natural History & Prognosis

- Most slow growing
- Malignant tumors may cause rapid neurologic deterioration
- Survival varies with tumor histology/grade and gross total resection
  - 5-year survival: 80% for low grade; 30% for high grade
- Postoperative neurologic function determined largely by degree of preoperative deficit

Treatment

- Serial monitoring for asymptomatic individuals
- Resection reserved for progressive neurologic decline
- Microsurgical resection (low-grade tumors)
- Adjuvant therapy
  - Option for WHO grades III-IV astrocytomas
  - No evidence that radiation therapy and chemotherapy improve long-term outcome

DIAGNOSTIC CHECKLIST

Consider

- Myelopathy should be evaluated with MR

Image Interpretation Pearls

- Axial and sagittal T1WI enhanced fat-saturated MR to exclude dural or pial lesion inciting syringomyelia

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(Left) *Sagittal T2WI MR demonstrates an expansile hyperintense intramedullary mass in the upper cervical cord.* **(Right)** *Sagittal T1WI C+ MR in the same patient shows minimal enhancement within this expansile intramedullary astrocytoma. Glial fibrillary acidic protein-δ immunoreactivity is reported to significantly correlate with the grade of spinal cord astrocytoma compared to that in normal control tissues.*





(Left) *Sagittal T1WI C+ MR depicts the typical pattern of fusiform enlargement of the cervical cord. There is irregular patchy enhancement within the cord at the C3 and C4 levels.* **(Right)** *Sagittal T2WI MR (left) illustrates an anaplastic astrocytoma of the thoracic cord with a long segment of mild cord expansion and hyperintensity. Sagittal T1WI C+ FS MR (right) reveals minimal enhancement of the lesion.*





(Left) *Sagittal T1WI C+ MR exhibits a lobulated intramedullary, ventrally exophytic mass with slight displacement of the cord posteriorly. The lesion displays patchy enhancement.* **(Right)** *Sagittal T1WI C+ MR of a pilomyxoid astrocytoma shows a predominantly solid and well-circumscribed homogeneously enhancing mass. Such tumors have shorter, progression-free and lower overall survival probabilities than classical low-grade astrocytomas, and have a high rate of CSF spread.*

Cellular Ependymoma

KEY FACTS

TERMINOLOGY

- Neoplasm arising from ependyma lining spinal cord central canal

IMAGING

- Circumscribed, enhancing hemorrhagic cord mass with surrounding edema
- Associated cysts common
- Cervical > thoracic > conus
- T1WI: Isointense or slightly hypointense to spinal cord
- T2WI: Hyperintense relative to spinal cord
- Cap sign: Hemosiderin at cranial or caudal margin
- Most tumors enhance

TOP DIFFERENTIAL DIAGNOSES

- Astrocytoma
- Hemangioblastoma
- Demyelinating disease
- Idiopathic transverse myelitis

PATHOLOGY

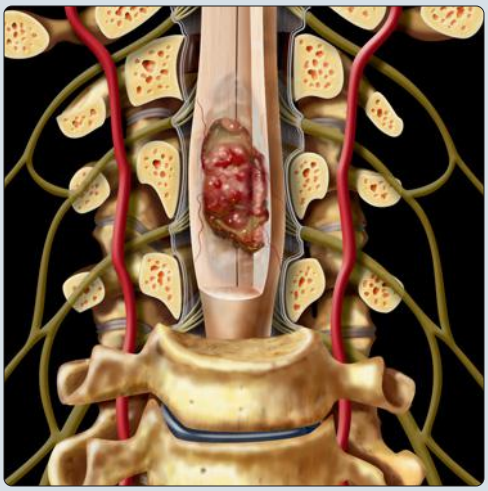
- Arises from ependymal cells of central cord canal
  - Most intramedullary
  - Rarely can be intradural extramedullary
- Most WHO grade II
- Rarely WHO grade III
  - Anaplastic ependymoma
- Association with NF2

CLINICAL ISSUES

- Most common presentation
  - Neck or back pain
- Other presentations
  - Progressive paraparesis, paresthesias

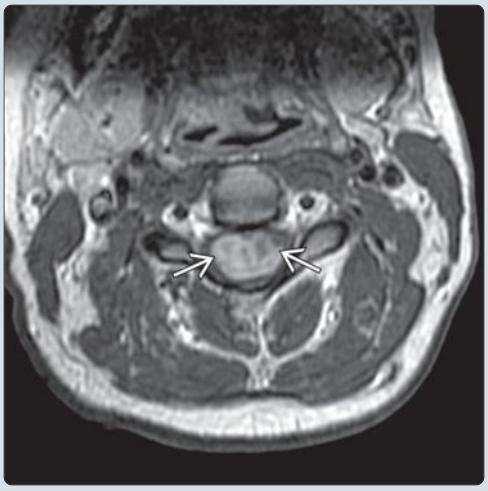
DIAGNOSTIC CHECKLIST

- Spinal cord neoplasm with associated peripheral hemorrhage suggestive of cellular ependymoma





(Left) *Coronal graphic depicts an intramedullary ependymoma mildly expanding the cervical cord. Cranial and rostral cysts, as well as hemorrhagic products, are associated with this mass.* **(Right)** *Sagittal T1WI C+ MR demonstrates a cervical intramedullary mass with solid and cystic components causing fusiform cord expansion. There are 2 areas of enhancing solid components and rostral cyst extending cephalad into brainstem. The adjacent solid portion shows little enhancement.*





(Left) *Axial T1WI C+ MR reveals an expansile intramedullary ependymoma with robust enhancement pattern.* **(Right)** *Sagittal T2WI MR illustrates the mixed cystic and solid appearance of cervical ependymomas in a patient with neurofibromatosis type 2. In addition, there is a dural-based intradural extramedullary meningioma.*

TERMINOLOGY

Synonyms

- Spinal ependymoma

Definitions

- Neoplasm arising from ependyma lining spinal cord central canal

IMAGING

General Features

- Best diagnostic clue
  - Circumscribed, enhancing hemorrhagic cord mass
- Location
  - Cervical > thoracic > conus
- Size
  - Multisegmental: Typically 3-4 segments
- Morphology
  - Symmetric cord expansion
  - Well circumscribed
  - ± exophytic component

Radiographic Findings

- Radiography
  - Osseous spinal canal widening: 20%
  - Erosion of medial pedicles, posterior vertebral scalloping
    - More common in lower spinal canal and filum ependymomas than cervical
  - ± scoliosis

CT Findings

- Bone CT
  - Osseous spinal canal widening
    - Thinned pedicles
    - Widened interpediculate distance
    - Posterior vertebral scalloping

MR Findings

- T1WI
  - Isointense or slightly hypointense to spinal cord
  - Hemorrhage hyperintense
  - Cord atrophy may be present
    - Correlates with surgical morbidity
- T2WI
  - Hyperintense relative to spinal cord
  - Focal hypointensity: Hemosiderin
    - Cap sign: Hemosiderin at cranial or caudal margin
    - 20-64% of cord ependymomas
  - Surrounding cord edema
  - 3 types of cysts (50-90%): Tumoral cyst; rostral or caudal cyst; central canal reactive dilatation (syrinx)
    - Tumoral cyst: Arises from degeneration, necrosis, and liquefaction within neoplasm
      - Contains mixture of protein, old hemorrhage, and necrotic tumoral tissue
      - Inhomogeneous composition → variable signal
      - Lined by abnormal glial cells
    - Rostral/caudal cyst
      - Location at pole(s) of neoplastic mass
    - Syrinx: Likely related to partial obstruction of central canal by tumor mass
      - Central location within spinal cord
      - CSF-equivalent signal
      - Location beyond tumor margins (above &/or below)
- T2* GRE
  - Hypointensity (hemorrhage)
    - May be 2° to highly vascular connective tissue stroma
    - May be 2° to lack of intervening neural tissue; therefore, vulnerable interface between tumor and normal cord substance
- T1WI C+
  - Tumor enhancement patterns
    - Intense, well-delineated homogeneous enhancement (50%)
    - Nodular, peripheral, or heterogeneous enhancement
    - Minimal or no enhancement (rare)
  - Peripheral enhancement of tumoral cysts
  - Polar cysts and syrinx do not demonstrate contrast enhancement around borders

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging
- Protocol advice
  - Sagittal, axial T2WI and T1WI MR ± gadolinium

DIFFERENTIAL DIAGNOSIS

Astrocytoma

- May be indistinguishable from ependymoma
  - Often longer, may be holocord
  - More often eccentric, infiltrative with indistinct margins
- Hemorrhage uncommon
- Tumor cysts, syrinx less common
- Most common primary cord neoplasm in children

Hemangioblastoma

- Cyst with enhancing highly vascular nodule
  - Flow voids often prominent
- Extensive surrounding edema relative to tumor size
- Thoracic > cervical
- Older patients
- 1/3 with von Hippel-Lindau disease

Demyelinating Disease

- Multiple sclerosis
- ADEM
- Lesions more often peripheral, posterolateral
- Ill-defined
- Typically < 2 vertebral segments in length
- Faint nodular or patchy enhancement
- Frequently multifocal
  - 90% have concurrent brain lesions

Idiopathic Transverse Myelitis

- Cord expansion less pronounced than ependymoma
- Centrally located, 3-4 vertebral segments in length
- Variable enhancement
- Thoracic > cervical
- Diagnosis of exclusion

PATHOLOGY

General Features

- Etiology
  - Arises from ependymal cells of central cord canal
- Genetics
  - Cord ependymomas are genetically different from intracranial lesions
    - Chromosome copy number aberrations using comparative genomic hybridization: Gain on chromosomes 2, 7, 12, etc.
    - Structural abnormalities on chromosomes 1, 6, 17, etc.
  - Ependymoma are associated with NF2
    - Deletions, translocations of chromosome 22
- Associated abnormalities
  - Subarachnoid hemorrhage (especially papillary type)
  - Superficial siderosis
  - NF2
    - Schwannomas
    - Meningiomas
- 4 subtypes: Cellular, papillary, clear-cell, tanycytic
  - Cellular most common intramedullary tumor subtype
  - Tanycytes: Precursors of astrocytes and ependymal cells

Staging, Grading, & Classification

- Most are WHO grade II
- Rarely WHO grade III
  - Anaplastic ependymoma

Gross Pathologic & Surgical Features

- Soft red or grayish-purple mass
  - Small blood vessels on tumor surface
- Well-circumscribed
  - May be encapsulated
- Cystic change common
- Hemorrhage at tumor periphery

Microscopic Features

- Immunohistochemistry: Positive for GFAP, S100, vimentin
- Cellular: Cuboidal or low columnar cells arranged in diffuse fashion
  - Perivascular pseudorosettes
  - True ependymal rosettes less common (some classify as epithelial-type)
  - Moderate cellularity with low mitotic activity
- Papillary: Glial stroma within papillary core
  - Single layer of cuboidal tumor cells form finger-like projection
- Clear-cell: Preferentially located in supratentorial compartment of young patients
- Tanycytic: Spindly, bipolar elements
  - Absent ependymal rosettes
  - Most commonly in spinal cord

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck or back pain
- Other signs/symptoms
  - Progressive paraparesis
  - Paresthesia
- Clinical profile
  - Delay in diagnosis due to slow tumor growth
    - Average duration of symptoms before diagnosis: 2.5 years

Demographics

- Age
  - 35-45 years
- Epidemiology
  - Ependymomas: 4% of all primary central nervous system neoplasms in adults
    - 30% of ependymomas are spinal
  - Most common primary spinal cord tumor in adults
    - 60% of primary spinal cord neoplasms
  - 2nd most common primary spinal cord tumor in children

Natural History & Prognosis

- Less preoperative neurologic deficit at presentation, better postoperative outcome
- Thoracic tumors have worse surgical outcome
- Metastasis rare
  - Lung, skin, kidney, lymph nodes
- 5-year survival: 85%

Treatment

- Grade II ependymomas may be observed carefully after imaging confirms complete resection
- Grade III tumors require adjuvant radiation treatment
- Surgical resection
  - Gross total resection in > 85% of cases
  - Very low risk of local and intracranial recurrence
- Radiotherapy for subtotal resection or recurrent disease
  - Typical doses of radiation therapy range from 4,000-5,400 cGy
  - No dose-response relationship

DIAGNOSTIC CHECKLIST

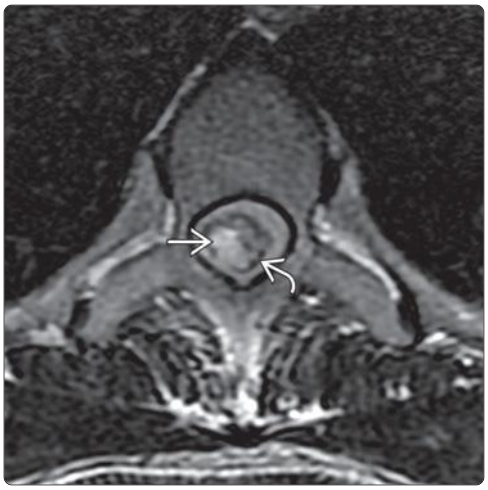
Image Interpretation Pearls

- Associated peripheral hemorrhage suggestive of cord ependymoma

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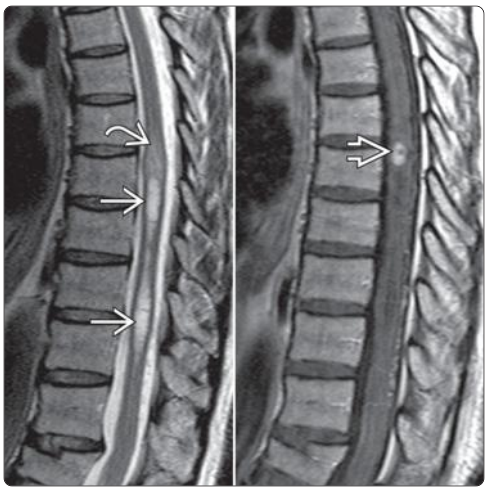


(Left) *Sagittal T2WI MR shows a large, heterogeneous, expansile mass involving the cervical and thoracic spinal cord with cyst formation and areas of low signal reflecting prior hemorrhage. Hemosiderin is seen capping the margins of the lesion. Note extensive cord edema, seen at the superior margin as a flame-shaped termination.* **(Right)** *Axial T2WI MR reveals tumoral cyst formation, edema, and hemosiderin deposition reflecting evidence of prior hemorrhage.*





(Left) *Sagittal T2WI MR illustrates the classic pattern of cord ependymoma with cyst formation and evidence of prior hemorrhage. This large heterogeneous mass expands the thoracic cord. The areas of low signal reflect hemosiderin deposition from prior hemorrhage. There is extensive caudal cord edema.* **(Right)** *Sagittal T1WI C+ FS MR reveals a heterogeneous thoracic spinal cord mass with cord expansion and cyst formation. A large central area of tumor enhancement is observed.*





(Left) *Sagittal T2WI MR demonstrates a heterogeneous lesion expanding the midthoracic spine with cystic areas and edema. The contrast-enhanced MR shows a nodular enhancing focus within the superior aspect of the mass.* **(Right)** *Sagittal T2WI MR depicts a round, hypointense, avidly enhancing intramedullary lesion at the C4-C5 level. The cord is mildly expanded due to this tanycytic ependymoma. T2 hyperintensity reflecting edema extends cranially and caudally.*

Myxopapillary Ependymoma

KEY FACTS

TERMINOLOGY

- Slow-growing glioma arising from ependymal cells of conus, filum terminale, cauda equina

IMAGING

- Usually spans 2-4 vertebral segments
  - May fill entire lumbosacral thecal sac
- Ovoid, lobular, sausage-shaped
- CT/radiographs
  - ± osseous canal expansion, thinned pedicles, vertebral scalloping
  - May enlarge, extend through neural foramina
- T1WI: Isointense→ hyperintense to cord
- T2WI: Almost always hyperintense to cord
  - Hypointensity at tumor margin = hemosiderin
- T1WI C+: Intense enhancement

TOP DIFFERENTIAL DIAGNOSES

- Nerve sheath tumor (NST)
- Intradural metastases
- Meningioma
- Paraganglioma

PATHOLOGY

- WHO grade I
- May have local seeding or subarachnoid dissemination
- Subarachnoid hemorrhage

CLINICAL ISSUES

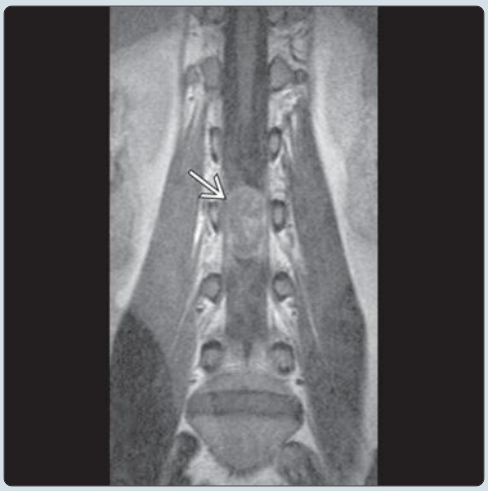
- Symptoms mimic disc herniation
- Back pain most common
- Other issues include paraparesis, radiculopathy, or bladder and bowel dysfunction

DIAGNOSTIC CHECKLIST

- Slow tumor growth may delay diagnosis
- Always image the conus in patients presenting with back pain

(Left) *Sagittal graphic shows a multilevel cauda equina myxopapillary ependymoma. Mass is vascular, with old intratumoral hemorrhage & acute subarachnoid hemorrhage along the dorsal conus. Indolent tumor growth has enlarged spinal canal & remodeled posterior vertebral cortex.* **(Right)** *Coronal T1WI MR shows a well-delineated, intradural extramedullary mass in lumbar spine. Lesion is predominantly isointense with cord & nerves. Almost 70% of filum terminale masses are ependymomas, mostly myxopapillary type.*





(Left) *Sagittal T2WI MR reveals an extensive intradural tumor distorting the conus and cauda equina. ME are unique in their intracellular and perivascular accumulation of proteinaceous mucin, which appears T1/T2 hyperintense.* **(Right)** *Sagittal T1WI C+ MR exhibits avid homogeneous tumor enhancement. Note diffuse intradural tumor seeding. Smaller tumors tend to displace the cauda equina nerve roots, whereas large tumors often compress or encase them.*





TERMINOLOGY

Abbreviations

- Myxopapillary ependymoma (ME)

Definitions

- Slow-growing glioma arising from ependymal cells of filum terminale

IMAGING

General Features

- Best diagnostic clue
  - Enhancing cauda equina mass with hemorrhage
- Location
  - Almost exclusively in conus, filum terminale, or cauda equina
  - Ependymomas outside of CNS are rare
    - Metastases or direct extension of primary CNS lesion after surgery
    - Direct extension to sacrococcygeal area from cord ependymoma or ME
    - Primary presacral, pelvic, or abdominal lesion
    - Primary ME of skin or subcutaneous tissue in sacrococcygeal region
      - Originates from ectopic ependymal remnants
- Size
  - Usually spans 2-4 vertebral segments
    - May fill entire lumbosacral thecal sac
- Morphology
  - Well circumscribed
  - Ovoid, lobular, sausage-shaped

Radiographic Findings

- Radiography
  - Vertebral remodeling changes
    - Widened interpediculate distance
    - Eroded pedicles
    - Posterior vertebral scalloping
    - Intervertebral foraminal widening

CT Findings

- NECT
  - Isodense intradural mass
- CECT
  - Homogeneous, avid enhancement
- Bone CT
  - ± osseous canal expansion
    - Thinned pedicles
    - Widened interpediculate distance
    - Scalloped vertebral bodies
    - May enlarge and extend through neural foramina

MR Findings

- T1WI
  - Usually isointense with cord
  - Hyperintensity attributed to mucin accumulation
- T2WI
  - Nearly always hyperintense to cord
  - Hypointensity at tumor margin = hemosiderin
    - 70% of intradural spine tumors with hemorrhage are ependymomas
  - May see flow voids
- STIR
  - Hyperintense
- T1WI C+
  - Intense enhancement
    - Reports of heterogeneous enhancement reflect unencapsulated tumors
      - Unencapsulated tumors may have extensive tumor necrosis, intratumoral hemorrhage, and progressive perivascular hyalinization → variable signal characteristics

Nonvascular Interventions

- Myelography
  - Well-demarcated lobular mass extending from conus
    - Nerve roots drape around mass
  - Meniscus of contrast delineates intradural extramedullary mass

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR with contrast
- Protocol advice
  - Always image conus in patients presenting with back pain

DIFFERENTIAL DIAGNOSIS

Nerve Sheath Tumor (NST)

- Small tumor associated with nerve root rather than filum terminale
- Large, multilevel NST may be indistinguishable
- Often extends through neural foramina
- Hemorrhage less common

Intradural Metastases

- Smooth or nodular enhancement along conus, nerve roots
- Enhancing mass(es), frequently multiple

Acquired Epidermoid Cyst

- Hypointense on T1WI
- Hyperintense on T2WI: Similar to CSF
- No enhancement
- Typically small

Meningioma

- Usually isointense with cord on T1 and T2WI
- More common in thoracic, cervical spine
  - Conus/filum location unusual
- Hemorrhage uncommon
- Osseous remodeling rare

Paraganglioma

- Rare tumor of cauda equina
- May be highly vascular
  - Indistinguishable from ME, although usually smaller

PATHOLOGY

General Features

- Etiology
  - Originates from ependymal cells of filum terminale

- Subcutaneous sacrococcygeal ME arises from coccygeal medullary vestige
  - Ependyma-lined cavity of caudal neural tube at postanal pit
- Genetics
  - Spinal ependymomas genetically different from intracranial ependymomas
- Associated abnormalities
  - Acute nonaneurysmal subarachnoid hemorrhage
  - Superficial siderosis

Staging, Grading, & Classification

- WHO grade I
- May have local seeding, subarachnoid dissemination
- No malignant degeneration

Gross Pathologic & Surgical Features

- Soft, lobulated, ovoid
- Grayish-white surface
- Noninfiltrating, often encapsulated
- May be highly vascular

Microscopic Features

- Spindle, columnar, cuboidal tumor cells radially arranged in papillary manner around vascularized stromal cores
- Fibrous, mucoid matrix
- Cysts, hemorrhage, calcifications common
- Absent, low mitotic activity
  - MIB (marker of cell proliferation) 0.4-1.6%
- Immunohistochemistry: Glial fibrillary acidic protein, S100, vimentin positive
  - Cytokeratin negative

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain
- Other signs/symptoms
  - Paraparesis
  - Radiculopathy
  - Bladder and bowel dysfunction: 20-25%
- Clinical profile
  - Symptoms mimic disc herniation
  - Slow tumor growth may delay diagnosis
    - ~ 2 year duration of symptoms prior to diagnosis

Demographics

- Age
  - Broad age range: Reported at all ages
    - Peak: 30-40 years
- Gender
  - M:F = 2:1
- Epidemiology
  - Ependymoma: 4% of all primary central nervous system neoplasms in adults
    - 30% of ependymomas are spinal
  - Cord ependymoma: Most common primary spinal cord tumor in adults
    - 60% of primary spinal cord neoplasms
  - ME: 27-30% of all ependymomas
    - Most common tumor of conus, filum terminale, cauda equina
    - 90% of filum terminale tumors

Natural History & Prognosis

- Excellent prognosis with complete resection
  - Late recurrence, distant metastases uncommon after complete resection
- Risk of local recurrence if incomplete resection
  - CSF dissemination if tumor capsule is violated
- Gross characteristics appear to be of greater prognostic significance than histologic features
  - Encapsulated tumors amenable to total surgical removal have lower recurrence rate (~ 10%) than those removed either piecemeal or subtotally (~ 19%)
  - Overall survival is related to residual disease
    - Total removal of tumor results in longer survival than subtotal resection
- Pediatric patients more likely to develop metastases and display more aggressive disease
  - Higher rate of local recurrence and tumor dissemination within neural axis

Treatment

- Resection
  - Gross total resection in > 85% of cases
- Radiotherapy for recurrence or subtotal resection
  - MEs are radiosensitive
- Adjuvant therapy for multifocal lesions

DIAGNOSTIC CHECKLIST

Consider

- Scanning up to at least midthoracic spine if conus lesion is found
- Slow tumor growth may delay clinical presentation, diagnosis

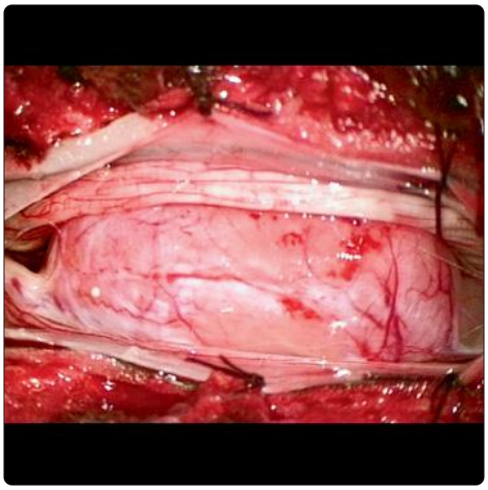
Image Interpretation Pearls

- T1/T2 hyperintense, enhancing, hemorrhagic mass associated with filum terminale is highly suggestive of ME

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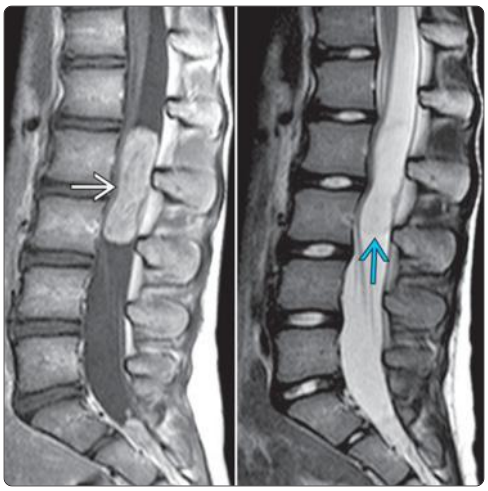


(Left) *Sagittal T1WI C+ MR illustrates a mildly enhancing, well-delineated, intradural extramedullary mass in the lumbar spine. ME usually appears as a round mass in the filum, sometimes involving the conus. A connective tissue capsule without firm attachment to, or incorporation of, surrounding spinal nerve root makes MEs highly amenable to gross total removal.* **(Right)** *Intraoperative photo shows dural retraction with exposure of an intradural ME. Note the mildly ↑ vasculature on the surface of the mass.*





(Left) *Sagittal T1WI C+ MR exhibits multiple nodular enhancing lesions along the cauda equina, representing CSF spread of ME.* **(Right)** *Sagittal T2WI MR displays a massive tumor distending the thecal sac from T12 inferiorly into the lumbar spine. Additionally, there is extensive posterior scalloping of the vertebra. Foci of low signal within the tumor reflect calcification &/or hemorrhage.*





(Left) *Sagittal T1W C+ (left) and T2WI (right) images show a well-circumscribed, enhancing T2 hyperintense mass arising from the filum terminale.* **(Right)** *Sagittal T2WI MR demonstrates a variant case of a rare subcutaneous ME in a premature newborn. There is fluid signal in a tumor cyst adjacent to solid tissue component. Sacral truncation is also visible. At surgery, the mass was connected by a thin fibrous tract into the thecal sac.*

Hemangioblastoma

KEY FACTS

TERMINOLOGY

- Low-grade, capillary-rich neoplasms of cerebellum and spinal cord that occur sporadically or in setting of von Hippel-Lindau (VHL) syndrome

IMAGING

- Thoracic > cervical > lumbar, sacral
- Subpial and posterior aspect of spinal cord
- Usually shows uniform contrast enhancement
- Large lesions have vessel flow voids
- Often associated with intraspinal cyst
- Extensive syrinx suggests hemangioblastoma (HB)
- Spinal cord thickening remote from tumor and without syrinx

TOP DIFFERENTIAL DIAGNOSES

- Arteriovenous malformation (AVM)
- Cavernous malformation
- Hypervascular cord neoplasms
  - Ependymoma
  - Astrocytoma
  - Vascular metastasis
- Intradural extramedullary tumors

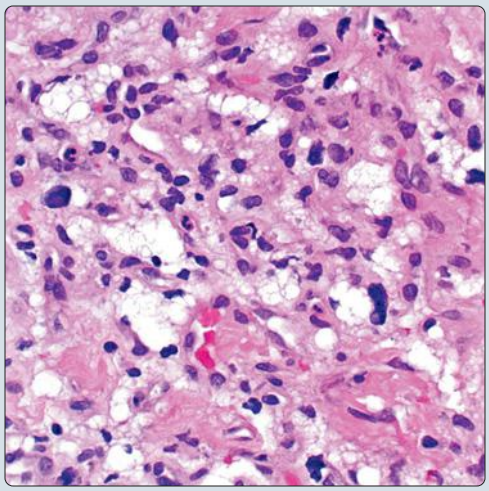
PATHOLOGY

- WHO grade I
- No malignant degeneration

CLINICAL ISSUES

- 68% of spinal HBs are sporadic (32% VHL-associated)
  - Multiple HBs indicate VHL syndrome
- Patients with suspected HB/VHL should undergo contrast-enhanced MR imaging of entire neural axis to exclude multiple lesions
- Symptoms: Sensory > motor > pain





(Left) *Sagittal graphic depicts a focal intramedullary mass in the cervical cord with prominent feeding vessels, adjacent cysts, and cord expansion with edema.* **(Right)** *Low-power H&E shows typical "stromal cells" (neoplastic component of the tumor) with pale vacuolated cytoplasm (clear cells) containing lipid droplets.*





(Left) *Sagittal T1WI C+ MR shows a focal enhancing mass in the dorsal cervical cord at the C4 level. Distinct appearances of spinal hemangioblastoma (HB) have been described, including associated cyst/syrinx, diffuse cord enlargement, exophytic morphology with minimal spinal cord reaction, and extramedullary location.* **(Right)** *T2WI MR displays a peripheral cystic component and focal mass with edema enlarging the cervical cord. The ring of hypointensity suggests susceptibility due to prior hemorrhage.*

TERMINOLOGY

Synonyms

- Capillary hemangioblastoma (HB)

Definitions

- Low-grade, capillary-rich neoplasms of cerebellum and spinal cord that occur sporadically or in setting of von Hippel-Lindau syndrome (VHL)

IMAGING

General Features

- Best diagnostic clue
  - Intramedullary mass with serpentine vascular flow voids
- Location
  - Subpial
  - Posterior aspect of spinal cord (SC), often associated with intraspinal cyst
  - Thoracic > cervical > lumbar, sacral
- Size
  - Few mm to several cm
  - Multiple tumors (often small) in VHL
- Morphology
  - Round, well-defined margins

CT Findings

- CTA
  - May show enlarged feeding vessels, enhancing mass

MR Findings

- T1WI
  - Depends on lesion size, presence of syrinx
  - Small
    - Isointense with cord (may be invisible unless hemorrhage has occurred)
    - Delineated syrinx (hypointense) present in > 50%
  - Large
    - Mixed hypo-/isointense
    - Lesions ≥ 2.5 cm almost always show "flow voids" (enlarged feeding arteries ± draining veins)
- T2WI
  - Small lesions usually uniformly hyperintense
    - ± peritumoral edema
  - Syrinx fluid often slightly hyperintense to CSF
  - Larger lesions ± mixed hyperintensity (flow voids, hemorrhage common)
  - May show extensive, long segment cord edema without syrinx
- T1WI C+
  - Small
    - Subpial nodule (often on surface of dorsal cord)
    - Well-demarcated, intense, homogeneous enhancement
  - Large
    - Heterogeneous enhancement
    - If syrinx present, wall does not enhance
- MRA
  - Noninvasive modality can provide preoperative detail of architecture of feeding vessels

Angiographic Findings

- Conventional
  - Intense, prolonged vascular stain
  - ± arteriovenous shunting
  - Enlarged spinal arteries (anterior > posterior) supply mass

Imaging Recommendations

- Best imaging tool
  - Sagittal, axial T2WI and T1 MR with contrast
- Protocol advice
  - Patients with suspected HB/VHL should undergo contrast-enhanced MR of entire neural axis to exclude multiple lesions
  - DSA for large lesions/preoperative embolization

DIFFERENTIAL DIAGNOSIS

Arteriovenous Malformation (AVM)

- Cord often normal/small, gliotic
- Syrinx, focal enhancing nodule absent

Cavernous Malformation

- Mottled or speckled pattern of prior hemorrhage, hemosiderin rim
- Minimal enhancement

Hypervascular Cord Neoplasms

- Ependymoma
  - Centrally located mass
  - Intense enhancement with well-defined margins and occasional central cavitation
  - ± syrinx
- Astrocytoma
  - Heterogeneous enhancement
  - Usually not hypervascular
  - Peritumoral edema common
- Vascular metastasis (known primary, e.g., renal cell carcinoma)

Intradural Extramedullary Tumors

- Meningioma
- Schwannoma
  - Rarely associated with syrinx, uncommon flow voids
- Paraganglioma
  - Filum much more common than spinal cord

PATHOLOGY

General Features

- Etiology
  - Suppressor gene product (VHL protein) causes neoplastic transformation
- Genetics
  - Familial HB (VHL)
    - Autosomal dominant
    - Chromosome 3p, other gene mutations common
    - VEGF highly expressed
    - Erythropoietin often upregulated
    - Tumors arise after loss or inactivation of wild-type allele in cell

  - In 20% of VHL families, no deletion or mutation can be detected
- Sporadic HB (unknown origin)
- Associated abnormalities
  - VHL
    - Cerebellar HBs, retinal angiomas, pheochromocytoma, renal cell carcinoma, angiomatous or cystic lesions of kidney/pancreas/epididymis, endolymphatic sac tumor

Staging, Grading, & Classification

- HBs are WHO grade I
- Exophytic HB
  - HB originating at or near surface of cord (66%)
  - Incites little or no reaction in adjacent cord parenchyma
  - May mimic nerve sheath tumors, meningioma, or ependymoma
- HB associated with syrinx (40-55%)
  - Syrinx may be small and cyst-like or may extend length of cord
- HB associated with cord enlargement (23%)
  - Cord enlargement beyond margins of enhancing tumor and distinct from syrinx
  - May be caused by arteriovenous shunting, venous congestion, or edema-promoting factor produced by tumor
    - Cord swelling will usually improve after enhancing nodule resected
- Extramedullary HB
  - 8% of HBs are intradural extramedullary
- Intramedullary HB with no spinal cord reaction (23%)
  - Small intramedullary HB may not elicit significant cord enlargement or syrinx
  - 25% of HBs are completely intramedullary

Gross Pathologic & Surgical Features

- Red to orange, well-circumscribed vascular mass on dorsal surface
- Rare extensive involvement of leptomeninges = leptomeningeal hemangioblastomatosis
- Prominent arteries, veins

Microscopic Features

- Large vacuolated stromal cells + rich capillary network
- If present, cyst wall is usually compressed cord and not part of tumor

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific clinical symptoms
    - Sensory (39%) > motor deficit or pain
  - VHL patients usually have 1 dominant symptomatic lesion; may have other smaller, asymptomatic lesions
    - 95% symptom-producing spinal HBs associated with syringomyelia
- Other signs/symptoms
  - May cause secondary polycythemia (erythropoietin upregulated)
  - Long clinical course
    - Mean duration of symptoms: 38 months
  - Rarely, may be source of subarachnoid hemorrhage or hematomyelia

Demographics

- Age
  - Mean at presentation: 30 years
    - Patients with VHL may present earlier
- Gender
  - M = F
- Epidemiology
  - 1.6-5.8% of all spinal tumors
  - 68% of spinal HBs are sporadic (32% associated with VHL)
  - Often multiple (VHL usually has 1 large ± many small HBs)

Natural History & Prognosis

- Grows slowly
- Does not undergo malignant degeneration
- Life expectancy of VHL patients: 50 years
  - Cause of death: CNS hemangioblastomas, followed by renal cell carcinoma

Treatment

- Preoperative arterial embolization
- Microsurgical resection of symptomatic tumors
  - Ventral tumors or completely intramedullary tumors associated with ↑ risk of postoperative worsening
- Stereotactic radiosurgical ablation (Gamma Knife therapy)
  - Particularly useful in VHL patients, who tend to present with multiple HBs that would otherwise require multiple surgical procedures

DIAGNOSTIC CHECKLIST

Consider

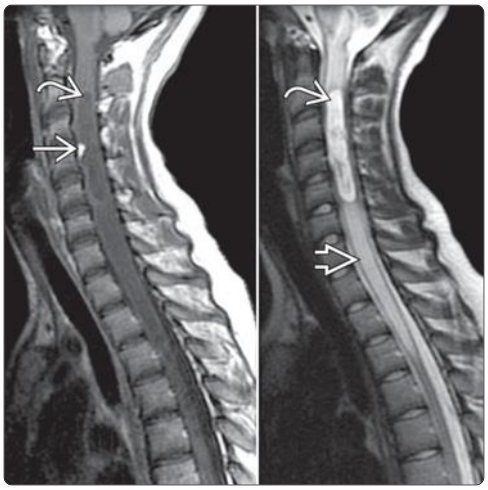
- VHL: Annual physical and ophthalmologic examinations should begin in infancy
  - Imaging of abdominal organs, CNS (brain and spine) added in teenagers and adults

Image Interpretation Pearls

- Typical pattern of intensely enhancing small masses on dorsal pial surface of cord

SELECTED REFERENCES

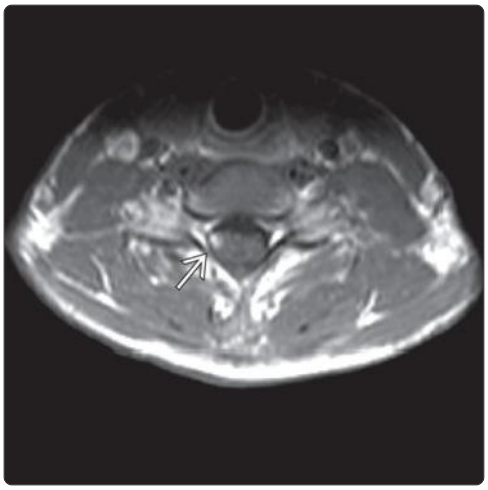
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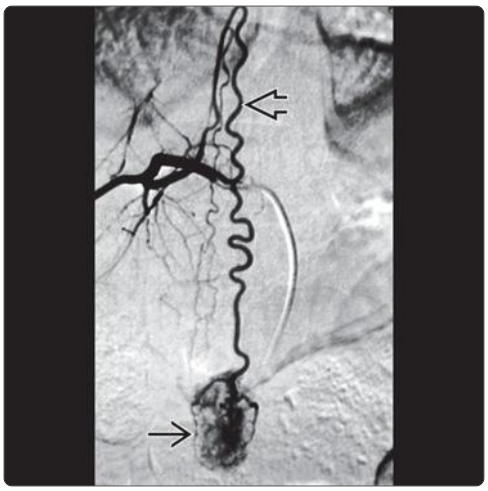
(Left) *Sagittal T1WI C+ MR (left) demonstrates an enhancing nodule in the ventral cord with an associated cyst. Sagittal T2WI MR (right) reveals the heterogeneous cyst and extensive surrounding edema.* **(Right)** *Sagittal T1W (left), T1W C+ (middle), and T2W (right) images exhibit an intramedullary enhancing mass with a small cystic component superiorly. There is a prominent serpentine vascular structure along the dorsal surface of the cord.*





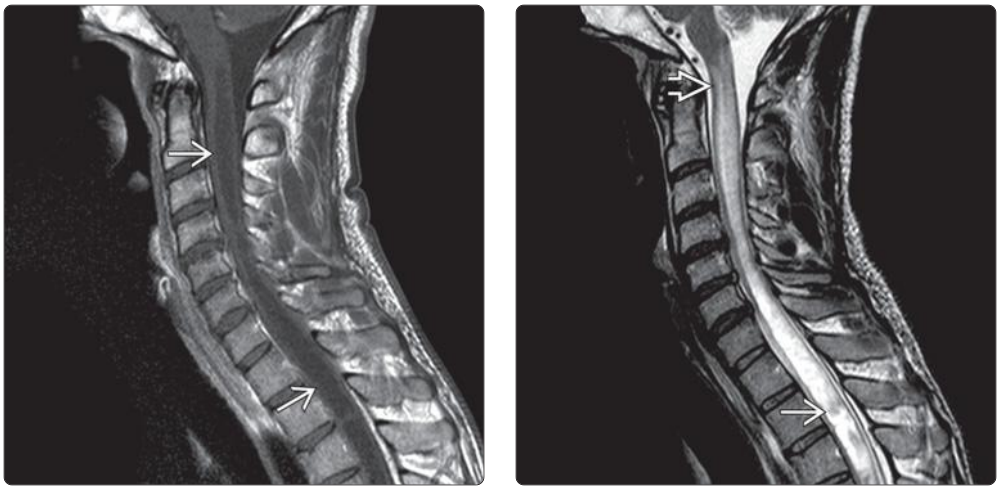
(Left) *Sagittal T1WI C+ MR shows multiple focal enhancing HBs along the cord surface in this patient with von Hippel Lindau (VHL). Note the HB at the level of the obex of 4th ventricle.* **(Right)** *Axial post-contrast image through the cervical spine shows the peripheral location of a small cervical nodule of HB.*





(Left) *Sagittal T1WI C+ FS MR displays a small enhancing lesion with an associated cyst and prominent vessel. HB is a highly vascular tumor, and MRIs of large HBs show flow voids resulting from prominent vessels. Rarely, spinal HBs may hemorrhage in either the cord parenchyma or the subarachnoid space.* **(Right)** *Anteroposterior angiography shows an enlarged anterior spinal artery feeding a hypervascular mass at the thoracic lumbar junction.*

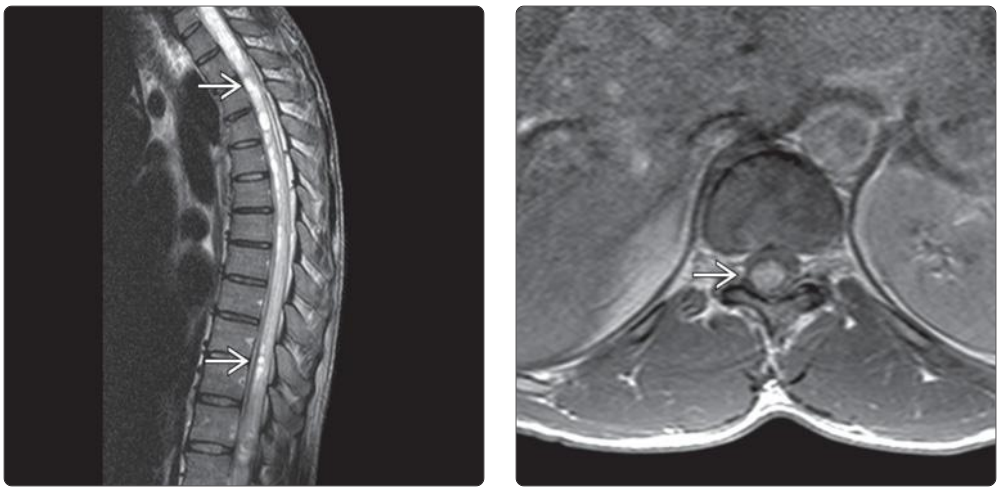
(Left) *Sagittal MR shows extensive syrinx formation ➡ as low signal on T1 images throughout the spinal cord and extending to the CVJ in this patient with a conus hemangioblastoma.* **(Right)** *Sagittal T2WI MR shows syrinx formation seen as increased signal throughout the spinal cord ➡ and extending cephalad. Low-signal flow dephasing is seen within the syrinx ➡. The tumor was at the level of the conus medullaris.*



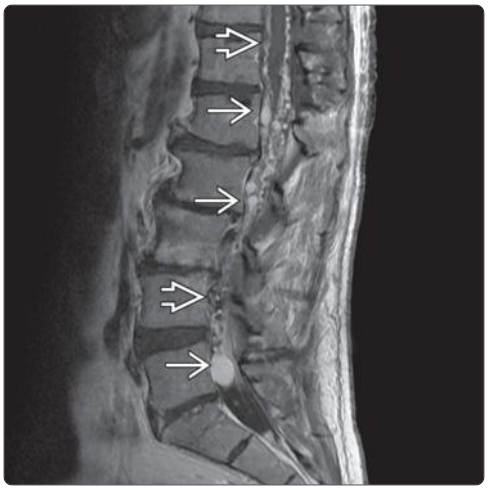
(Left) *Sagittal T1 C+ MR thoracolumbar image shows a solitary focus of intense enhancement ➡ in the conus. Syrinx formation from the tumor extended to the CVJ.* **(Right)** *Lumbar T2W image shows a rounded focus of more intermediate signal ➡, suggesting an underlying mass lesion within the conus medullaris.*



(Left) *Sagittal T2WI MR through the thoracic spine shows extensive loculated syrinx formation as increased signal throughout the spinal cord ➡ in this patient with a conus level hemangioblastoma.* **(Right)** *Axial T1 C+ MR shows a solitary focus of intense enhancement ➡ in the conus in this patient with pathology-proven HB.*

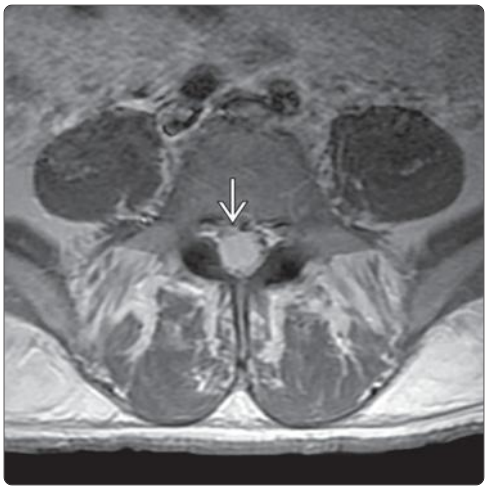






(Left) *Sagittal T1 C+ MR, in a rare case of hemangioblastomatosis in a patient without VHL, shows enhancement of the multiple enlarged intrathecal vessels and multiple intradural, homogeneously enhancing masses.* **(Right)** *Sagittal T1WI C+ MR shows enhancement of the multiple, enlarged intrathecal vessels supplying multiple intradural, homogeneously enhancing masses. Hemangioblastomatosis may occur with prior posterior fossa surgery with tumor seeding into the CSF.*





(Left) *Sagittal T2WI MR shows extensive serpentine flow voids within the thecal sac, which might be mistaken for a vascular malformation except that there are also round soft tissue masses within the thecal sac that do not exhibit flow voids.* **(Right)** *Axial T1 C+ MR shows the solid enhancing mass within the lumbar thecal sac in this patient with hemangioblastomatosis (without VHL).*





(Left) *Sagittal T2WI MR shows findings of an intradural extramedullary HB posterior to the distal cord. There is severe cord compression from the lesion, with adjacent cord edema. Note the multiple flow voids associated with feeding vessels.* **(Right)** *Sagittal T1 C+ MR in a patient with intradural extramedullary HB shows the dorsal, pial-based mass with marked enhancement and multiple adjacent enhancing, enlarged feeding vessels.*

Spinal Cord Metastases

KEY FACTS

TERMINOLOGY

- Metastatic lesion from primary carcinoma in another organ (including brain)

IMAGING

- Focal, enhancing cord lesion(s) with extensive edema
- Typically small (< 1.5 cm)
- T1WI: Enlarged cord
- T2WI/PD/STIR: Focal high signal represents diffuse edema
  - Rarely syrinx
- T1WI C+: Focal enhancement
- T2* GRE: Hypointensity due to hemorrhagic components

TOP DIFFERENTIAL DIAGNOSES

- Demyelinating disease
  - Multiple sclerosis
  - Acute disseminated encephalomyelitis
- Primary cord tumor
- Inflammatory granuloma
- Tuberculosis
- Sarcoidosis
- Inflammatory myelitis

CLINICAL ISSUES

- Brown-Séquard syndrome
- Rapidly progressive flaccid paraparesis
- Intramedullary spinal cord metastatic (ISCM) heralds poor prognosis

DIAGNOSTIC CHECKLIST

- Full craniospinal imaging when focal cord lesion found
- Edema out of proportion to focal small cord lesion suggests metastasis, even if solitary





(Left) *Graphic depicts metastases to the skeleton, epidural space and cord. The hemorrhagic intramedullary metastasis expands the cord. Rapid onset of symptoms is characteristic for ISCM. Asymmetric dysfunction of the spinal cord mimicking Brown-Séquard has been reported in 30-40% in ISCM but is exceptional in patients with epidural spinal cord compression.* **(Right)** *Sagittal T1WI C+ MR shows an avidly enhancing non-small cell metastasis to the conus. Note the slight enhancement of cauda equina roots.*





(Left) *Sagittal T2WI MR demonstrates a well-defined intramedullary lesion in the thoracic spinal cord due to angiosarcoma metastases. The lesion has a hyperintense center and hypointense rim. Also note spinal cord edema extending above and below the lesion.* **(Right)** *Sagittal T1WI C+ MR reveals subtle enhancement of the angiosarcoma metastasis. Nonneoplastic lesions, such as radiation myelopathy, demyelinating plaques, and paraneoplastic necrotizing myelopathy, can be difficult to differentiate from ISCM.*

TERMINOLOGY

Synonyms

- Intramedullary spinal cord metastatic (ISCM) disease

Definitions

- Metastatic lesion from primary carcinoma in another organ (including brain)

IMAGING

General Features

- Best diagnostic clue
  - Focal, enhancing cord lesion(s) with extensive edema
- Location
  - Cervical > thoracic > lumbar, conus
- Size
  - Typically small (< 1.5 cm)
- Morphology
  - Well circumscribed
  - Spherical or oval

CT Findings

- CECT
  - May rarely depict hypervascular mets as enhancing intraspinal foci (e.g., hypernephroma)

MR Findings

- T1WI
  - Enlarged cord
  - Rarely, syrinx cavity
  - Hyperintense T1 signal of melanoma metastasis may be 2° to degradation products of intralesional hemorrhage or 2° to paramagnetic properties of stable free radicals within melanin
- T2WI
  - Focal high signal due to diffuse edema
  - Rarely syrinx
  - Rarely, low signal due to hemorrhagic metastasis (e.g., from thyroid, melanoma)
- PD/intermediate
  - Focal high signal: Edema
- T2* GRE
  - Hypointensity due to hemorrhagic components
- T1WI C+
  - Focal enhancement
    - Rim sign: Intense, thin rim of peripheral enhancement around enhancing lesion
    - Flame sign: Ill-defined, flame-shaped region of enhancement at superior/inferior margins

Ultrasonographic Findings

- Grayscale ultrasound
  - Helps in surgical removal
  - Focal hyperechoic lesion

Angiographic Findings

- Conventional
  - Helps differentiate arteriovenous malformation (AVM) or hemangioblastoma from metastasis

Nonvascular Interventions

- Myelography
  - May miss lesion in substantial minority of cases
    - False-negative in 40-50%
  - When positive, depicts focal cord swelling
  - CT with intrathecal contrast (post myelography)
    - As myelography, can miss lesion
    - Symmetric fusiform cord expansion over multiple levels

Nuclear Medicine Findings

- PET
  - Has been anecdotally reported to detect metastases

Imaging Recommendations

- Best imaging tool
  - MR
    - T2WI and T1WI C+
    - Fat saturation helps conspicuity
- Protocol advice
  - Delayed imaging (10-20 min) after contrast to increase lesion detection in clinically suspected cases

DIFFERENTIAL DIAGNOSIS

Demyelinating Disease

- Multiple sclerosis (MS)
  - Usually in dorsal cord, flame-shaped on T2WI
  - Less cord swelling and edema compared to metastasis
  - Multiple lesions
  - Lesion(s) may not enhance
  - Most have brain involvement
- Acute disseminated encephalomyelitis (ADEM)
  - Multiple lesions
  - Simulates MS

Primary Cord Tumor

- Astrocytoma
- Ependymoma
- Hemangioblastoma
- Neurofibroma

Cord Arteriovenous Malformation

- Vascular flow voids leading to and from high-flow lesions
- Angiographically occult AVMs have hemosiderin stain, no edema

Inflammatory Granuloma

- Tuberculosis (TB)
- Sarcoidosis
  - Look for pretracheal nodes

Inflammatory Myelitis

- Transverse myelitis
  - Usually very long cord segment involved
  - Haphazard, if any, enhancement
- Viral myelitis
  - Preexisting illness
  - CSF viral titers may prove diagnosis

Radiation Myelopathy

- Do not present with pain
- Insidious progression of neurological deficits

PATHOLOGY

General Features

- Etiology
  - Hematogenous dissemination (most common metastatic route)
    - Either through arterial supply or retrograde spread from venous system
  - Carcinomatous meningitis, contiguous spread
  - Spread may occur through central canal via CSF pathways
  - Lung most common primary
    - Small cell > non-small cell
  - Breast 2nd most common
  - Virtually any primary can be source, including brain, thyroid, and urinary bladder
- Genetics
  - Predisposing conditions, such as von Hippel-Lindau syndrome for renal cell carcinoma (RCC)
- Associated abnormalities
  - Hemorrhage
  - Syrinx
    - Fluid in cavity typically higher in signal compared to developmental syringohydromyelia
  - Brain mets in approximately 20-59%
  - ~ 25% have leptomeningeal carcinomatosis
- Seen in < 1% of general autopsies
- Only 5% diagnosed antemortem
- CSF: Negative or increase in protein and mild pleocytosis

Gross Pathologic & Surgical Features

- Well-circumscribed fleshy lesion

Microscopic Features

- Typically reflect primary source

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Rapidly progressive flaccid paraparesis
- Other signs/symptoms
  - Sphincter disturbance
    - Urinary and bowel dysfunction with conus involvement predominate
  - Brown-Séquard syndrome
  - Pain
    - Radicular pain in 25-33%
  - Hypesthesia
  - Constitutional symptoms, such as weight loss or decreased appetite or energy
  - Increased hematocrit specific clinical feature of RCC because it may produce erythropoietin
- Clinical profile
  - Most cases have known primary CA and develop sudden myelopathy
  - In 20% of ISCM, this is 1st presentation of cancer

Demographics

- Age
  - Any; typically over 50
- Gender
  - No preference
- Epidemiology
  - ISCM represents only 4-8.5% of all CNS mets
    - May reflect different percentage shared by cardiac output (20% to brain vs. 5% to spinal cord) and consequent degree of tissue perfusion

Natural History & Prognosis

- ISCM heralds poor prognosis
  - Minority survive > 1 year
  - Survival varies from 2 weeks to 9 months after onset of myelopathy
- Preoperative neurological status, nature of primary cancer, presence of systemic &/or CNS metastases may influence survival

Treatment

- Options, risks, complications
  - Steroids, chemotherapy, radiation therapy
  - Surgery an option in solitary cases
    - May improve survival rate and neurologic function
  - Variable response to radiotherapy
    - Positive response in patients in whom early diagnosis was made or who had radiosensitive tumors
  - Chemotherapy
    - Most studies show no effect on survival
  - Stereotactic spine radiosurgery may be improve symptoms, including pain relief
- Goal of therapy is stabilization or reversal of myelopathy, and preservation of ambulation

DIAGNOSTIC CHECKLIST

Consider

- Full craniospinal imaging when focal cord lesion found

Image Interpretation Pearls

- Edema out of proportion to focal small cord lesion suggests metastasis, even if solitary

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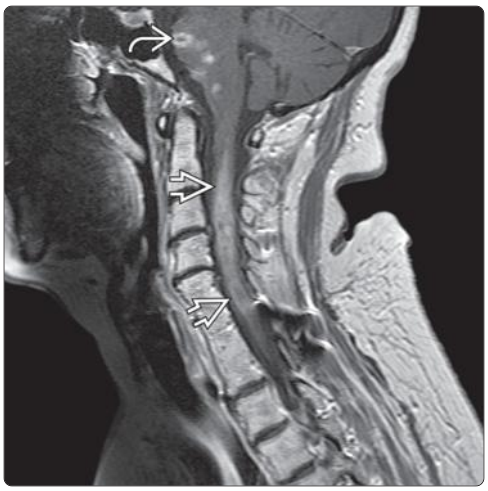


(Left) *Sagittal T2WI MR displays diffuse cord edema from cervicomedullary junction inferiorly to C5. A focal hypointense signal abnormality at C2-C3 is due to a cord metastatic lesion from lung carcinoma.* **(Right)** *Sagittal T1WI C+ FS MR depicts an intramedullary enhancing focus at the C3 level, which is a metastasis from renal cell carcinoma. The space-occupying effect of the tumor together with the edema leads to fusiform widening of the cord.*





(Left) *Sagittal STIR MR shows high signal intensity edema within the distal thoracic cord in this patient with small cell lung carcinoma metastases.* **(Right)** *Sagittal T1 C+ MR shows focal area of enhancement within the distal thoracic spinal cord from metastatic small cell lung carcinoma.*





(Left) *Sagittal T1WI C+ MR reveals direct extension of glioma from brainstem inferiorly into cervical cord. There are multiple foci of abnormal enhancement within the brainstem. Spinal metastases of GBM are quite rare and dissemination usually occurs late in the course of the disease.* **(Right)** *Sagittal MR exhibits a T1-hyperintense intramedullary lesion. This ill-defined enhancing lesion on T1WI C+ FS MR was found to be a melanoma metastasis. There is extensive intramedullary edema.*

Primary Melanocytic Neoplasms/Melanocytoma

KEY FACTS

TERMINOLOGY

- Uncommon neoplasms derived from normal leptomeningeal melanocytes
- Morphologic spectrum from low-grade melanocytoma to melanocytic tumor of intermediate differentiation to malignant melanoma
  - Distinct from other melanotic lesions, such as meningioma, schwannoma, melanoma

IMAGING

- Intradural mass, either intradural extramedullary or rarely intramedullary
- MR
  - T1WI: Isointense to hyperintense intradural lesion
  - T2WI: Isointense to hypointense to normal cord
  - T2* GRE: May show blooming of low signal related to melanin susceptibility effect
  - T1WI C+: Heterogeneous enhancement

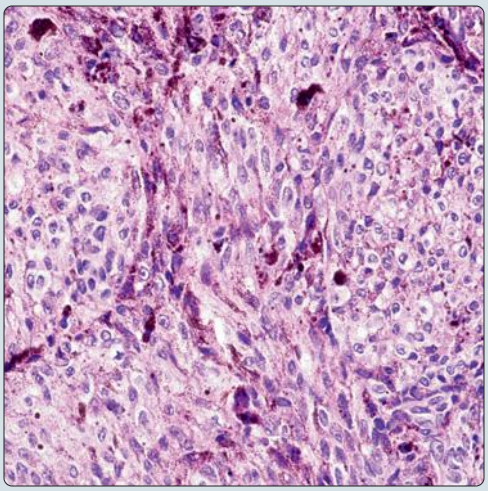
TOP DIFFERENTIAL DIAGNOSES

- Hemorrhagic cord neoplasm
- Cavernous malformation
- Arteriovenous malformation
- Pigmented intradural extramedullary tumors
  - Malignant melanoma, meningioma, schwannoma

CLINICAL ISSUES

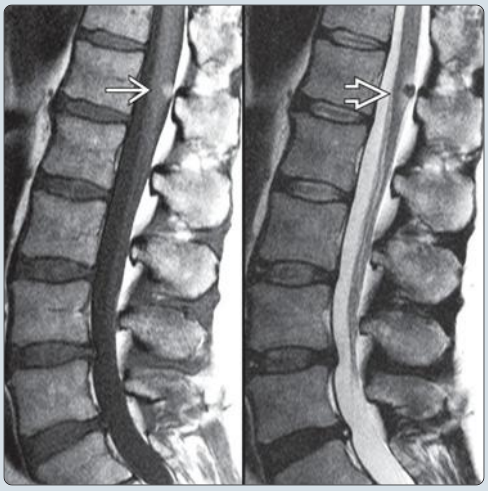
- Progressive back pain, extremity numbness, weakness
- Progressive, low-grade lesions curable by resection
- Can be locally invasive
- Treatment
  - Total tumor resection is optimal to prevent recurrence
  - Adjuvant radiotherapy after incomplete resection

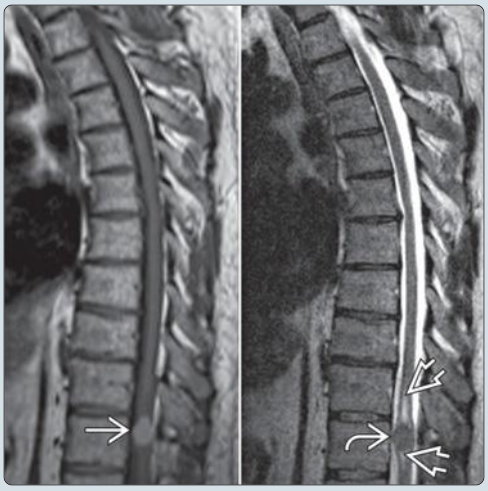
(Left) *Tight nests of bland, slightly spindled cells are key features of melanocytoma. Sparsely pigmented or amelanotic tumors may be misinterpreted as meningioma.* **(Right)** *Sagittal T1 C+ MR shows that there is homogeneous, marked enhancement of a T1 hyperintense intramedullary lesion ➡ in the conus medullaris.*





(Left) *Sagittal nonenhanced T1 MR (left) shows a primary intramedullary melanocytoma of the cord with intrinsic T1 shortening ➡ that is characteristic of melanin. Sagittal T2 MR (right) shows focal low signal ➡. (Courtesy P. Hildenbrand, MD.)* **(Right)** *Sagittal T1 MR shows a well-delineated intramedullary mass in the lower thoracic cord that demonstrates high T1 ➡ (left) and low T2 signal ➡ (right). There is surrounding cord edema ➡.*





TERMINOLOGY

Definitions

- Uncommon neoplasms derived from normal leptomeningeal melanocytes
  - Morphologic spectrum from low-grade melanocytoma to melanocytic tumor of intermediate differentiation to malignant melanoma

IMAGING

General Features

- Best diagnostic clue
  - Intradural enhancing mass showing T1 hyperintensity, not fat suppressing
- Location
  - Intradural mass, either intradural extramedullary or rarely intramedullary
  - Highest concentration of melanocytes in upper cervical leptomeninges
    - Can occur in thoracolumbar region
- Size
  - Variable

MR Findings

- T1WI
  - Isointense to hyperintense intradural lesion
- T2WI
  - Isointense to hypointense to normal cord
- T2* GRE
  - May show blooming of low signal related to melanin susceptibility effect
- T1WI C+
  - Heterogeneous, nonspecific enhancement

Imaging Recommendations

- Best imaging tool
  - MR C+ of entire spine and brain to exclude additional lesions and possibility of metastatic melanoma
- Protocol advice
  - T1, T2WI, and post-contrast T1WI

DIFFERENTIAL DIAGNOSIS

Hemorrhagic Cord Neoplasm

- Ependymoma may show hemorrhage, hemosiderin
- Vascular metastasis, such as renal cell carcinoma

Cavernous Malformation

- Mottled or speckled pattern of prior hemorrhage, hemosiderin rim

Arteriovenous Malformation

- Multiple flow voids

Pigmented Intradural Extramedullary Tumors

- Malignant melanoma
- Meningiomas, schwannomas may show melanin

PATHOLOGY

General Features

- Etiology
  - Unknown, may arise from neoplastic transformation of meningeal melanocytes

Staging, Grading, & Classification

- No established cutoffs defining grades, with much overlap
- Melanotic lesions probably constitute spectrum from well-differentiated melanocytoma → overtly malignant melanoma
  - Melanocytoma MIB-1 staining low (< 1-2%)
  - Intermediate-grade lesions have occasional mitoses, MIB-1 staining 1-4%
  - Melanoma MIB-1 labeling indices (mean: 8.1%) higher than melanocytomas

Gross Pathologic & Surgical Features

- Dark pigmentation and encapsulation, superficial siderosis

Microscopic Features

- Monomorphic spindle cells with prominent nucleoli and melanin in cytoplasm
  - Minimal to absent mitotic activity
- Immunoreactive for antimelanoma antibody HMB-45 and S100 protein, vimentin antibodies, and negative for epithelial membrane antigen

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Progressive back pain, extremity numbness, weakness (≥ 1 year)

Demographics

- Age
  - Wide age range (13-73 years); mean age: 45 years

Natural History & Prognosis

- Progressive, low-grade lesions curable by resection
- Generally lack metastatic potential

Treatment

- Total tumor resection is optimal to prevent recurrence
  - Adjuvant radiotherapy after incomplete resection

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Signal relates to paramagnetic free radicals in melanin, with shortening of T1, T2 by proton-proton dipole-dipole interaction

SELECTED REFERENCES

1. Foit NA et al: Bifocal extra- and intradural melanocytoma of the spine: case report and literature review. Eur Spine J. 22 Suppl 3:S521-5, 2013
2. El-Khashab M et al: Intermediate grade meningeal melanocytoma of cervical spine. Childs Nerv Syst. 25(4):407-10, 2009
3. Horn EM et al: Surgical treatment for intramedullary spinal cord melanocytomas. J Neurosurg Spine. 9(1):48-54, 2008

Ganglioglioma

KEY FACTS

TERMINOLOGY

- Intramedullary lesion containing both neoplastic astrocytes and neural cells

IMAGING

- Cervical > thoracic
  - Holospinal involvement
- CT/radiographs
  - Scoliosis and bone remodeling
- MR
  - T1WI: Mixed signal intensity
  - T2WI: Homogeneous > heterogeneous
  - Cysts or syrinx
  - Absence of edema
  - Patchy enhancement, no enhancement within tumor center
  - Hemosiderin or calcification

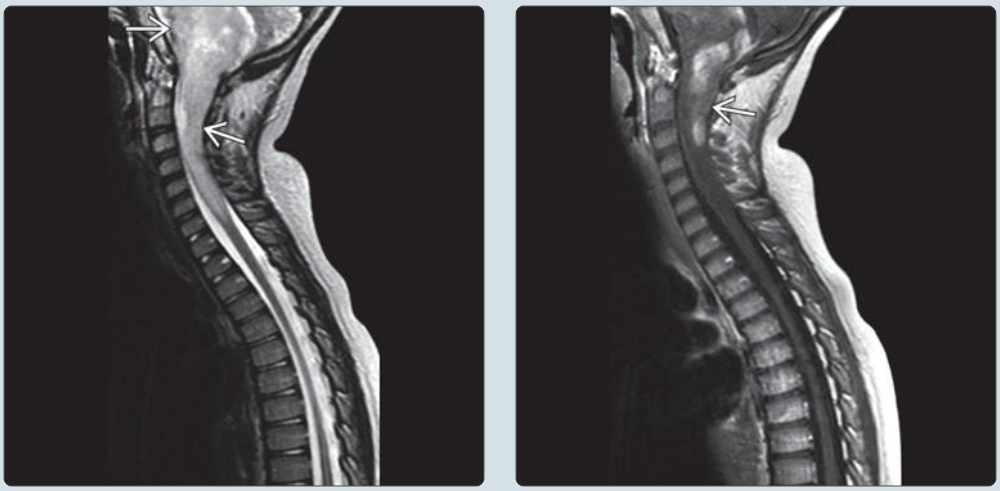
TOP DIFFERENTIAL DIAGNOSES

- Spinal cord astrocytoma
- Spinal cord cellular ependymoma

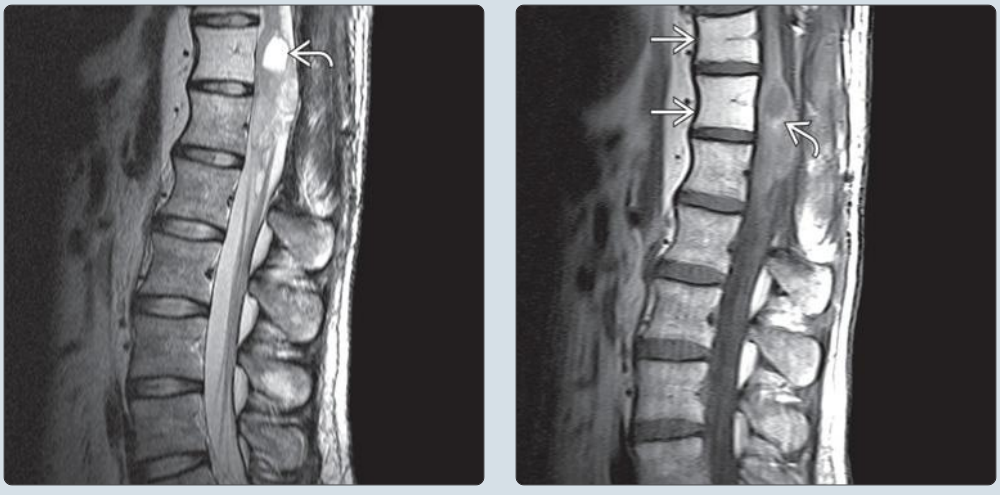
CLINICAL ISSUES

- Young age
  - Highest incidence in first 3 decades
- Back pain, limb weakness
- Progressive myelopathy
- 1.1% of all spinal cord tumors
- Higher relative risk of recurrence (3.5x) than cerebral gangliogliomas
- Rare reports of malignant degeneration of glial > gangliglionic elements (reported high as 10%)
- Extent of surgical resection is main prognostic factor

(Left) *Sagittal T2WI MR depicts an infiltrating brainstem and cervical cord mass. Infratentorial GG are unusual and tend to involve the brainstem. These patients have a better prognosis than patients with other brainstem gliomas.* **(Right)** *Sagittal T1WI C+ MR illustrates an infiltrating brainstem and cervical cord mass with patchy, irregular enhancement. The proportion of neural and glial cells varies in GG. There is frequent local involvement of the subarachnoid space; however, leptomeningeal spread is rare.*



(Left) *Sagittal T2WI MR exhibits a heterogeneous expansile mass with cystic and solid components. Most GG grow slowly, and symptoms result from tumor expansion with neurological findings reflecting the site of tumor.* **(Right)** *Sagittal T1WI MR shows a heterogeneous lesion expanding the distal cord. There is a curvilinear hyperintensity, which may be due to blood products ± dystrophic mineralization. Fatty marrow replacement of lower thoracic vertebral bodies from prior radiation treatment is noted.*



TERMINOLOGY

Abbreviations

- Ganglioglioma (GG)

Synonyms

- Gangliocytoma, ganglioneuroma, neuroastrocytoma

Definitions

- Well-differentiated, slowly growing neuroepithelial tumor, composed of neoplastic, mature ganglion cells in combination with neoplastic glial cells

IMAGING

General Features

- Location
  - Cervical (67% of cases) > thoracic
- Size
  - Long tumor length
- Morphology
  - Eccentrically located within spinal cord

Imaging Recommendations

- Protocol advice
  - MR delineates full extent of disease

MR Findings

- T2WI
  - Homogeneous SI in 60%, heterogeneous SI in 40%
- T1WI C+
  - Patchy enhancement and cord surface enhancement
  - Tumoral cysts more often in GG compared to astrocytoma and ependymoma
  - Reactive cysts do not show mural enhancement

DIFFERENTIAL DIAGNOSIS

Astrocytoma, Spinal Cord

- Enhancing infiltrating mass expanding cord
- Usually < 4 segments
- Almost always enhances
- ± cyst/syrinx

Ependymoma, Cellular, Spinal Cord

- Circumscribed, enhancing cord mass with hemorrhage
- Sharply delineated enhancement
- Polar or intratumoral cysts (50-90%)

Ependymoma, Myxopapillary, Spinal Cord

- Slow-growing glioma arising from ependymal cells of filum terminale
- Well circumscribed

PATHOLOGY

General Features

- Etiology
  - Benign tumors of mixed neuronal and glial components

Staging, Grading, & Classification

- Presence of neoplastic astrocytic component: Ganglioglioma
- Substantial population of nonneoplastic stroma and small neoplastic mature neurons: Ganglioglioneurocytoma
- Tumor consists solely of large relatively mature neoplastic neurons or ganglion cells and nonneoplastic stroma: Gangliocytoma
- Most GG are WHO grade 1

Microscopic Features

- GG suspected when otherwise astrocytic neoplasm contains component of large cells potentially representing neurons
- Mature ganglionic cells and nonganglionic glial elements

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Back pain, limb weakness
  - Progressive myelopathy
  - Scoliosis
- Other signs/symptoms
  - Paraparesis, paraplegia
  - Incontinence
  - Gait or sensory disturbance
  - Floating-Harbor syndrome: Expressive language delay, developmental delay, short stature, facial anomalies

Demographics

- Age
  - Young age (mean: 23 years)
- Gender
  - No gender predominance for spinal GG
- Epidemiology
  - 0.4-6.25% of all primary CNS neoplasms
    - 2nd most common intramedullary tumor in children
  - 1.1% of all spinal cord tumors
    - GG most often in cerebral hemispheres, particularly temporal lobes and 3rd ventricle
  - Report of GG in neurofibromatosis 2 (NF2)
    - Possible association because of predilection of NF2 individuals to form intramedullary tumors of glial lineage

Natural History & Prognosis

- Slow growing
  - Low malignant potential
  - Symptoms result from tumor expansion
- Extent of surgical resection is main prognostic factor

Treatment

- Gross total resection is treatment of choice for grade I and II gangliogliomas
- Radiation therapy reserved for anaplastic gangliogliomas and for recurrent tumors

SELECTED REFERENCES

1. Yang C et al: Intramedullary gangliogliomas: clinical features, surgical outcomes, and neuropathic scoliosis. J Neurooncol. 116(1):135-43, 2014
2. Lotfinia I et al: 'Intramedullary cervical spinal cord ganglioglioma, review of the literature and therapeutic controversies'. Spinal Cord. 47(1):87-90, 2009

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PART V
SECTION 2

Nonneoplastic Cysts and Tumor Mimics



Cysts

CSF Flow Artifact	**834**
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Perineural Root Sleeve Cyst	**842**
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Nonneoplastic Masses and Tumor Mimics

Epidural Lipomatosis	**850**
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Fibrous Dysplasia	**854**
Kümmell Disease	**856**
Hirayama Disease	**858**
IgG4-Related Disease/Hypertrophic Pachymeningitis	**860**

CSF Flow Artifact

KEY FACTS

TERMINOLOGY

- CSF flow-related phenomenon due to time of flight (TOF) effects and turbulent flow

IMAGING

- Location: Intrathecal, subarachnoid space
  - Most prominent in cervical and thoracic spine
- Low or high signal intensity
- Ill-defined margins

TOP DIFFERENTIAL DIAGNOSES

- Vascular malformation, type I dural arteriovenous fistula
- Vascular malformation, type IV arteriovenous fistula
- CSF drop metastases

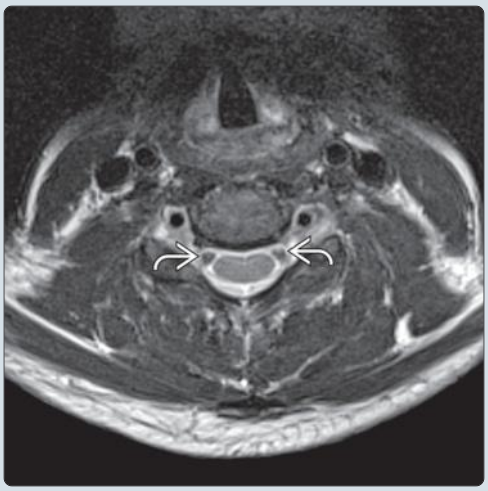
CLINICAL ISSUES

- Time of flight effects
  - Dark CSF signal
  - Positive relationship between CSF velocity and TOF losses
  - Typically occurs in spin-echo or fast spin-echo imaging
- Flow-related enhancement produces bright CSF signal
- Turbulent flow
  - Abnormally dark signal intensity
- Ghosting artifact motion
  - Periodic motion, such as CSF pulsation, cardiac motion, and respiratory motion

DIAGNOSTIC CHECKLIST

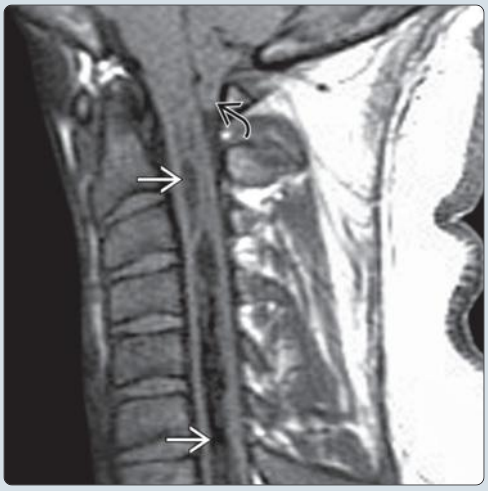
- Flow artifacts frequently have bizarre nonanatomic appearance
- Cross-reference other imaging planes and compare with faster imaging sequences (GRE, true FISP) that reduce superimposed flow phenomenon
- Ghosting artifacts in phase-encoding direction often accompany TOF- and FRE-related signal changes, help point out true nature of those changes





(Left) *Sagittal STIR MR shows an ill-defined signal void in the dorsal subarachnoid space of the thoracic spine. This artifact is due to a combination of respiratory and cardiac-related pulsatile CSF flow superimposed on cranially directed bulk CSF flow. There is also contribution from turbulent CSF flow moving from the ventral to dorsal subarachnoid space. This complex CSF motion results in phase incoherence leading to signal loss.* **(Right)** *Axial T2WI MR confirms the CSF flow artifact.*





(Left) *Axial T1WI MR depicts isointense CSF flow artifact dorsal to the thoracic cord in the subarachnoid space.* **(Right)** *Sagittal T1WI MR exhibits syringomyelia in the cervical cord. CSF flow dynamics are altered because of the cerebellar tonsillar ectopia. The turbulent flow results in a broader spectrum of proton velocities, which result in more rapid dephasing and signal loss. There is also a positive relationship between CSF velocity and time of flight (TOF) losses.*

TERMINOLOGY

Definitions

- CSF flow-related phenomenon due to time-of-flight (TOF) effects, turbulent flow

IMAGING

General Features

- Location
  - Intrathecal, subarachnoid space
  - More prominent in cervical and thoracic spine

Imaging Recommendations

- Protocol advice
  - Turbulence dephasing ↓ by shorter TE sequences and smaller voxel volume (decreased slice thickness or higher matrix)
  - Flow-related enhancement (FRE) ↓ by applying saturation bands or by using ultrafast techniques, such as true FISP
  - BLADE shown to be superior to turbo spin-echo sequences
  - ↓ ghosting artifact by ↑ number of excitations
  - T1 FLAIR at 3T and 1.5T shows superior CSF nulling, better conspicuity of normal anatomic structures and degenerative and metastatic lesions, and improved image contrast

MR Findings

- T1WI
  - Hyperintense relative to CSF
- T2WI
  - Hypointense relative to CSF
  - Ill-defined margins
- FLAIR
  - CSF flow artifacts at 3T > 1.5T
    - Crosstalk between each inversion pulse
- T2* GRE
  - Resistant to TOF loss because of short TE
- **T1 FLAIR**
  - Relatively short TI/TR pair to produce effective nulling of CSF signal, to achieve heavy T1 weighting, and to optimize contrast between abnormal tissues and cord/bone marrow

DIFFERENTIAL DIAGNOSIS

Vascular Malformation, Type I Dural Arteriovenous Fistula (DAVF)

- Dilated pial veins cover abnormally enlarged, hyperintense distal cord

Vascular Malformation, Type IV AVF

- Intradural extra-/perimedullary AVF with multiple well-defined serpentine flow voids on ventral or dorsal surface of cord

CSF Drop Metastases

- Administer intravenous contrast, image in multiple planes if necessary

PATHOLOGY

General Features

- Etiology
  - Pulsatile to-and-fro movement of CSF due to expansion and contraction of brain produced by expansion and contraction of intracranial vessels associated with cardiac cycle
    - Results in significant motion-related effects that can alter visualized signal of CSF

CLINICAL ISSUES

Demographics

- Age
  - More prominent in pediatric patients

Natural History & Prognosis

- TOF effects
  - Dark CSF signal
    - More pronounced with faster proton velocity, thinner slices, longer TE, and imaging plane perpendicular to flow
    - Typically occurs in spin-echo or fast spin-echo imaging
  - Single-shot fast spin-echo (SS FSE): TOF loss during systole, without TOF loss in diastole
  - Relationship between CSF velocity and TOF losses
  - Laminar flow: Peripherally located protons move at slower velocity → reduction in TOF losses
- FRE produces bright CSF signal
  - Bright signal from unsaturated protons flowing into a slice replace outflowing, partially saturated protons during TR
  - Seen on more slices with single slice acquisition techniques in which every slice is an entry slice
- Turbulent flow
  - Abnormal dark signal intensity
  - Varied flow velocities and directions → more rapid dephasing and signal loss, "intravoxel dephasing"
- Ghosting artifact motion
  - Periodic motion, such as CSF pulsation, cardiac motion, and respiratory motion
  - Phase encoding direction
  - Bright if in phase or dark if out of phase with background signal

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Flow artifacts frequently have bizarre nonanatomic appearance

SELECTED REFERENCES

1. Lavdas E et al: Reduction of motion, truncation and flow artifacts using BLADE sequences in cervical spine MR imaging. Magn Reson Imaging. 33(2):194-200, 2015
2. Kwon JW et al: Three-dimensional isotropic T2-weighted cervical MRI at 3T: comparison with two-dimensional T2-weighted sequences. Clin Radiol. 67(2):106-13, 2012
3. Lisanti C et al: Normal MRI appearance and motion-related phenomena of CSF. AJR Am J Roentgenol. 188(3):716-25, 2007

Meningeal Cyst

KEY FACTS

TERMINOLOGY

- Intraspinal extramedullary loculated CSF collection

IMAGING

- Nonenhancing extramedullary loculated CSF intensity collection displacing cord or nerve roots
  - Solitary, multiple, or multiloculated
- Extradural or intradural extramedullary location
  - Extradural meningeal cyst (MC) may extend through enlarged neural foramina
  - Presence of intradural MC suggested by mass effect on spinal cord
- CSF intensity on T1WI, T2WI, STIR
- Cyst wall may be imperceptible
  - No enhancement
- "Cap" sign: Extradural MC outlined by rostral and caudal epidural fat

TOP DIFFERENTIAL DIAGNOSES

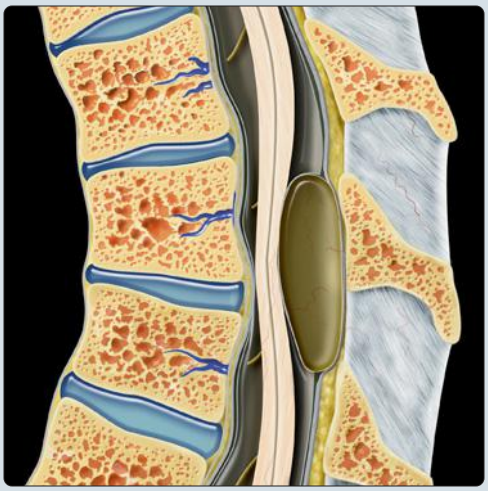
- Idiopathic spinal cord herniation
  - Focal cord atrophy and ventral deviation
- Dural ectasia
  - Spinal cord not distorted
- Spinal nerve root avulsion
  - Contiguous with subarachnoid space

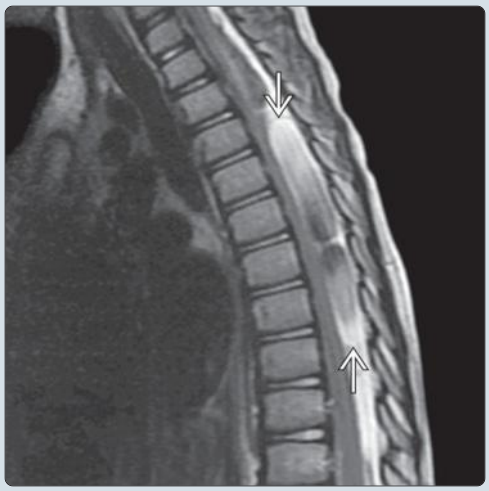
PATHOLOGY

- Nabors classification of spinal MC
  - Type I: Extradural MC without spinal nerve root fibers
  - Type II: Extradural MC with spinal nerve root fibers
  - Type III: Intradural MC

CLINICAL ISSUES

- Most patients asymptomatic
- Other signs/symptoms
  - Pain, paraparesis, paresthesia
- Worsening neurologic deficits with enlarging cyst





(Left) *Sagittal graphic demonstrates a type III intradural meningeal cyst in the mild dorsal thoracic canal, with moderate mass effect on the spinal cord.* **(Right)** *Sagittal T2WI MR depicts an intradural dorsal meningeal cyst causing mild spinal cord compression. The cyst margins can be visualized ➡ adjacent to areas of turbulent CSF flow. This is a typical case of an intradural meningeal cyst presenting with myelopathy. The cyst was treated with surgical fenestration.*





(Left) *Sagittal STIR MR shows a compression fracture of the L4 vertebral body ➡. A post-traumatic dural tear with arachnoid herniation results in an intradural arachnoid cyst ➡.* **(Right)** *Sagittal T2WI MR shows a loculated arachnoid cyst dorsal to the cord at the C3-4 level ➡. There is cord edema ➡ related to coccidiomycosis meningitis and cord compression from the arachnoid cyst. Note the hydrocephalus and ballooned 4th ventricle ➡ due to the cranial meningeal involvement.*

TERMINOLOGY

Abbreviations

- Spinal meningeal cyst (MC), arachnoid cyst (AC)

Synonyms

- Dural dissection cyst, intradural dissecting meningocele

Definitions

- Intraspinal extramedullary loculated CSF collection

IMAGING

General Features

- Best diagnostic clue
  - Nonenhancing extramedullary loculated CSF intensity collection displacing cord or nerve roots
- Location
  - Extradural or intradural extramedullary
  - Primary MC
    - Extradural: Posterior or posterolateral lower thoracic spine
    - Intradural: Dorsal midthoracic spine
    - Anterior location uncommon
  - Secondary to trauma, instrumentation, subarachnoid hemorrhage
    - No specific location
- Size
  - Variable
    - Average between 2-4 vertebral segments
- Morphology
  - Well circumscribed
  - Oval, elongated
  - "Dumbbell" lesion: Extension through neural foramen
  - Cyst wall may be imperceptible
    - Presence of intradural MC suggested by mass effect on spinal cord
  - Solitary, multiple, or multiloculated
  - "Cap" sign: Extradural MC outlined by rostral and caudal epidural fat

Radiographic Findings

- Radiography
  - ± bony canal expansion
    - Posterior vertebral scalloping
    - Widened interpedicular distance

CT Findings

- NECT
  - Axial postmyelography CT may delineate communication between extradural MC and subarachnoid space
  - Contrast-filled intradural MC may be difficult to visualize
    - Suggested by mass effect on cord and nerve roots
    - Delayed imaging to show late filling of cyst
- Bone CT

MR Findings

- T1WI
  - CSF intensity
- T2WI
  - Typically CSF intensity
  - May be more hyperintense than CSF
    - Slow or lack of CSF flow
  - Hypointensity related to flow-related signal loss
- PD/intermediate
  - CSF intensity
- STIR
  - CSF intensity
  - CSF pulsation may be more pronounced
- T1WI C+
  - No enhancement
  - Variable mass effect on spinal cord
    - Myelomalacia
  - Syringohydromyelia at or away from site of MC
    - May result from partial obstruction of CSF by MC
- MR cine
  - 2D-cine phase-contrast CSF-flow study may show abrupt change in flow at cyst margin
  - May demonstrate compression at rostral side of cyst during diastole
- MR myelography
  - Heavily T2-weighted sequences may be helpful to detect cyst wall
    - Noninvasive
    - Some techniques are 3D acquisitions → multiplanar reformations
  - Intrathecal gadolinium sensitive for detecting cysts communicating with CSF spaces

Nonvascular Interventions

- Myelography
  - Intradural MC: Intrathecal filling defect
  - Extradural MC: Effaced subarachnoid space
  - Spinal cord compression
  - Variable myelographic block
    - May have delayed filling of cyst

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI and T1WI MR with gadolinium
- Protocol advice
  - Delayed imaging on postmyelography CT to allow filling of cyst

DIFFERENTIAL DIAGNOSIS

Idiopathic Spinal Cord Herniation

- Upper to midthoracic level most common
- Focal cord atrophy and ventral deviation
  - No dorsal intradural cyst on myelography
  - Herniation through ventral dural defect

Facet Synovial Cyst

- Typically lumbosacral spine
  - L4-L5 most common
- Posterolateral aspect of spinal canal
  - Adjacent to facet joint
- Associated facet arthropathy

Dural Ectasia

- Associated with Marfan syndrome, NF-1
- Diffuse dilatation of thecal sac
- Spinal cord not distorted
- No block on myelography

Spinal Nerve Root Avulsion

- History of trauma
  - Contiguous with subarachnoid space
  - No discrete intraspinal lesion

Pseudomeningocele

- Postoperative fluid collection contiguous to operative site

PATHOLOGY

General Features

- Etiology
  - Primary **intradural** MC thought to arise from diverticulum of arachnoid
    - Arachnoid membrane splits to form cyst wall
  - Possible etiologies of primary **extradural** MC
    - Congenital diverticulum of dura
    - Arachnoid protrusion through dural defect
  - Both ± communication with arachnoid space through a neck
    - Enlargement possibly due to "ball valve" mechanism
  - Secondary MC results from prior trauma, iatrogenic (LP, spinal infection, surgery), infection, or hemorrhage
    - Post-traumatic dural tear with arachnoid herniation
    - Postinflammatory or surgical adhesions compartmentalize subarachnoid space
- Genetics
  - Multiple MCs may be associated with adult polycystic kidney disease
- Primary MC: Diverticulum of dura, arachnoid, or nerve root sheath

Staging, Grading, & Classification

- Nabors classification of spinal MC
  - Type I: Extradural MC without spinal nerve root fibers
    - A: Extradural MC/AC
    - B: Sacral meningocele
  - Type II: Extradural MC with spinal nerve root fibers
    - Tarlov perineural cyst
    - Spinal nerve root diverticulum
  - Type III: Intradural MC
    - Intradural MC
    - Acquired intradural MC also known as subarachnoid cyst

Gross Pathologic & Surgical Features

- Smooth-walled cystic mass
- Clear, colorless fluid

Microscopic Features

- Primary MC ± arachnoid lining inner cyst wall
  - Sometimes duplicated
  - Variable connective tissue also present
  - Typically avascular
  - No significant inflammatory changes
- Secondary cyst formed by fibrous connective tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most patients asymptomatic
    - Especially type II MC
  - Other signs/symptoms
    - Pain, paraparesis, paresthesia
    - Hyperreflexia
    - Bladder and bowel incontinence
- Clinical profile
  - Pain may worsen with Valsalva maneuver
    - Increased intracystic pressure

Demographics

- Age
  - May present at any age
    - Type I thoracic MC more common in adolescents
    - Others more common in adults
- Epidemiology
  - Primary extradural MC relatively rare

Natural History & Prognosis

- Worsening neurologic deficits with enlarging cyst
- Excellent prognosis
  - Complete relief of symptoms > 80%
- Degree of cord atrophy predictive of neurological outcome

Treatment

- Laminectomy with complete cyst wall resection
- If complete resection not possible
  - Wide marsupialization of cyst, shunting, fenestration

DIAGNOSTIC CHECKLIST

Consider

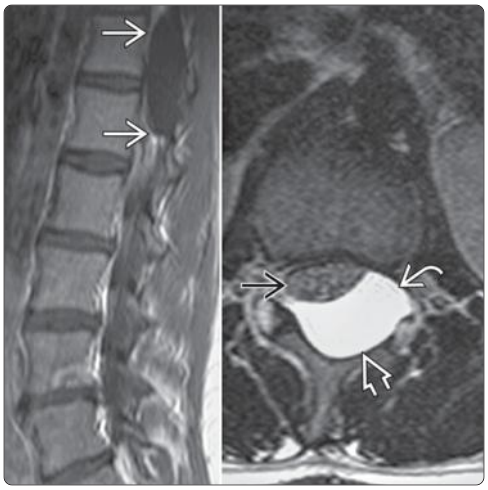
- Thin-section axial CT myelography to define communication between extradural MC and subarachnoid space
- Heavily T2-weighted MR myelography may be helpful to detect cyst wall

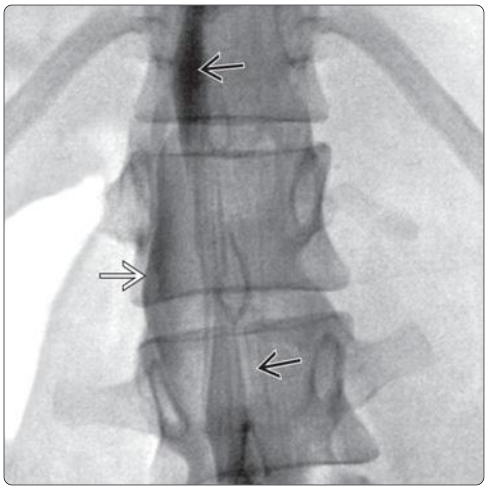
Image Interpretation Pearls

- Nonenhancing thoracic extramedullary CSF intensity collection with mass effect on spinal cord highly suggestive of spinal MC

SELECTED REFERENCES

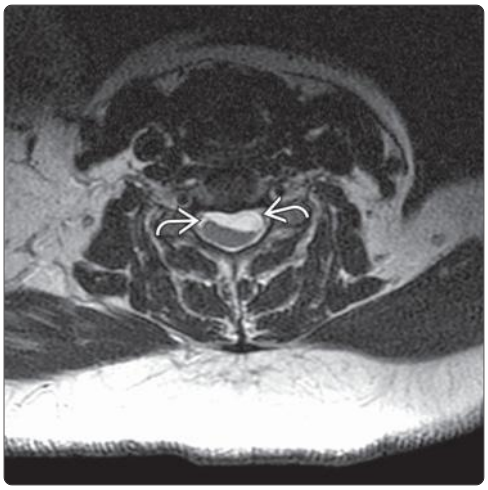
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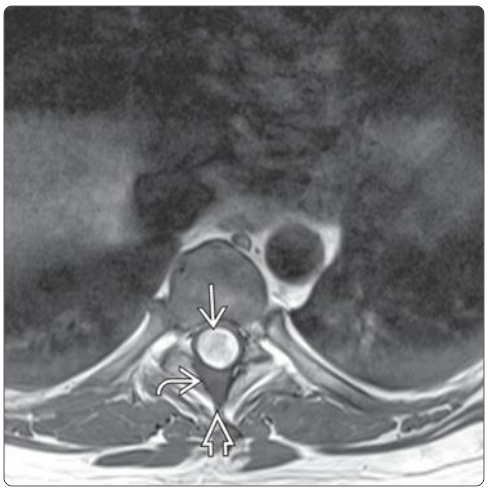
(Left) *Sagittal T1WI C+ MR (left) illustrates a nonenhancing intradural MC ➡ at the thoracolumbar junction. Axial T2 MR (right) shows internal hyperintense signal within the MC ➡. There is smooth scalloping of the lamina ➡. The thecal sac and distal cord are displaced anteriorly ⇨.* **(Right)** *Anteroposterior myelography demonstrates intrathecal opacification with contrast ⇨. There is pooling of contrast separate from the intrathecal subarachnoid component within the MC ➡.*





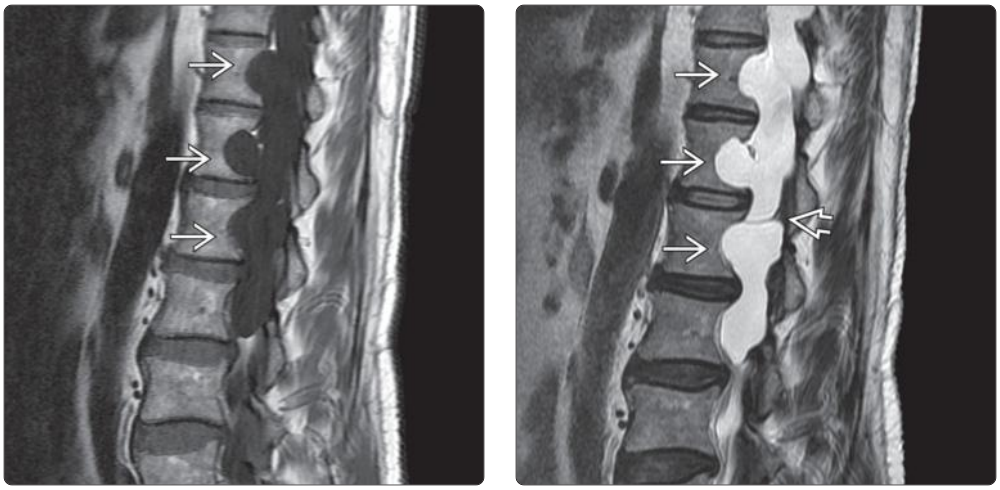
(Left) *Axial CT myelogram after conventional myelogram displays the intradural extramedullary MC ➡ opacified with contrast but distinctly separate from the thecal sac, which is displaced anteriorly ⇨. The smooth scalloping of the posterior elements is observed ⇨.* **(Right)** *Axial T2WI MR shows a hyperintense extramedullary lesion ➡ causing mild mass effect on the cervical cord. This is a subdural hematoma mimicking an arachnoid cyst. The blood products were better exhibited on the T2* GRE MR.*



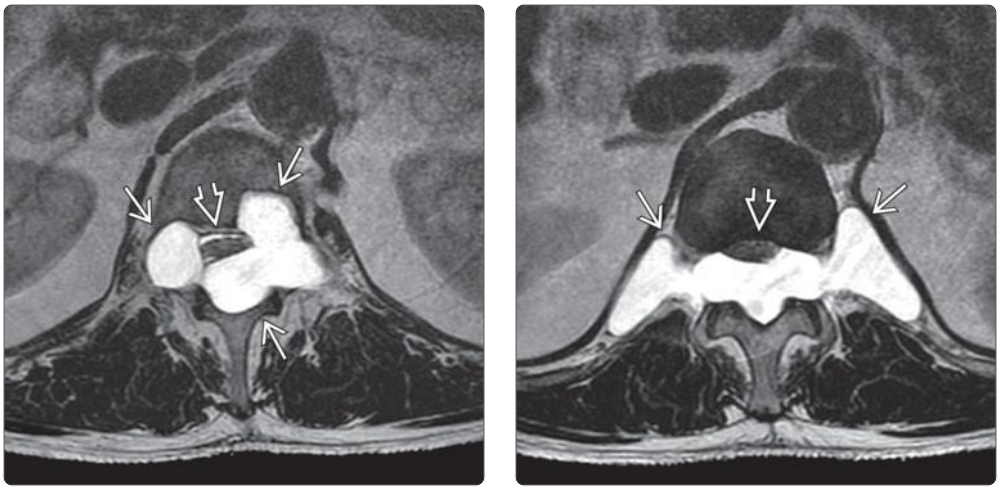


(Left) *Sagittal T2WI MR depicts an extradural septated MC ➡ in the midthoracic spine. It is associated with an intradural extramedullary dermoid cyst ➡, which causes severe compression of the cord. MCs can occur anywhere along the canal; middle and lower thoracic regions are most frequently involved.* **(Right)** *Axial T1WI MR illustrates the intradural dermoid cyst ➡, which shows intrinsic T1 hyperintensity. The dorsal extradural MC is T1 hypointense ➡. Spinal dysraphism is noted ➡.*

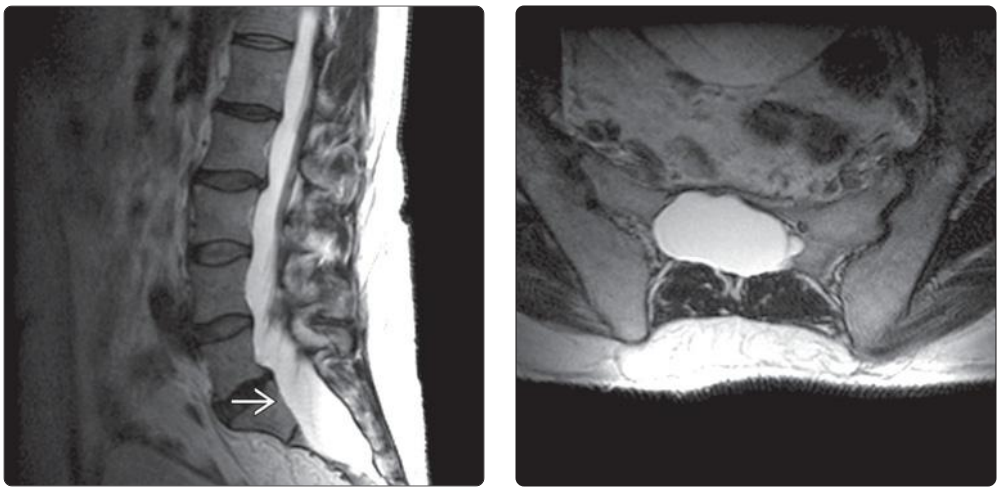
(Left) *Sagittal T1WI MR shows a giant extradural meningeal cyst extending over multiple segments with extensive remodeling and scalloping ⇨ of the vertebral bodies.* **(Right)** *Sagittal T2 TSE MR shows the giant extradural meningeal cyst ⇨ with high signal, extensive bony remodeling, and intracystic septation ⇨.*



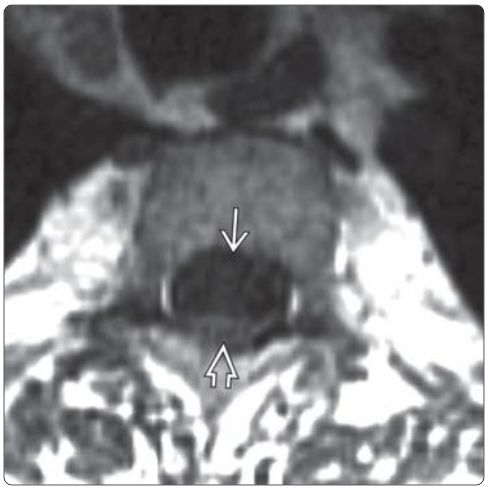
(Left) *Axial T2WI MR shows bilateral extent of a giant extradural meningeal cyst with vertebral body and posterior element remodeling ⇨, and ventral displacement and compression of the cord ⇨.* **(Right)** *Axial T2WI MR shows bilateral extension of a giant extradural meningeal cyst out along both neural foramina ⇨, with bony expansion and remodeling. There is ventral cord displacement and compression ⇨.*



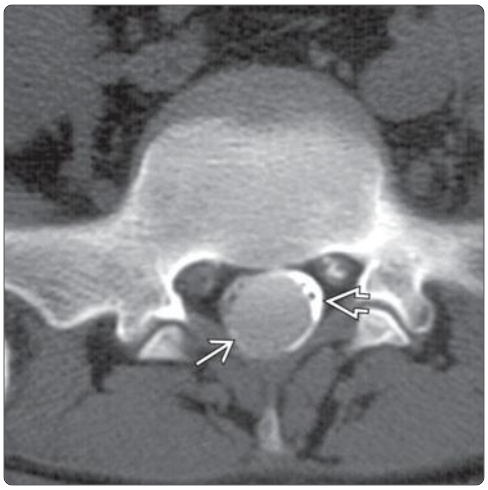
(Left) *Sagittal T2WI MR shows a large "occult" intrasacral meningocele that expands and remodels the sacrum ⇨.* **(Right)** *Axial T2W MR shows high signal from the large meningeal cyst within the sacrum with adjacent sacral bony expansion and remodeling.*

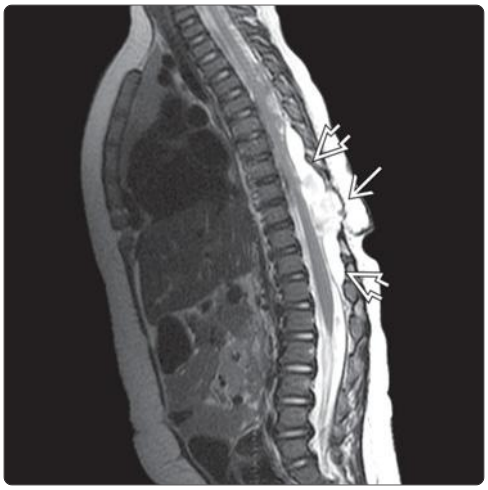




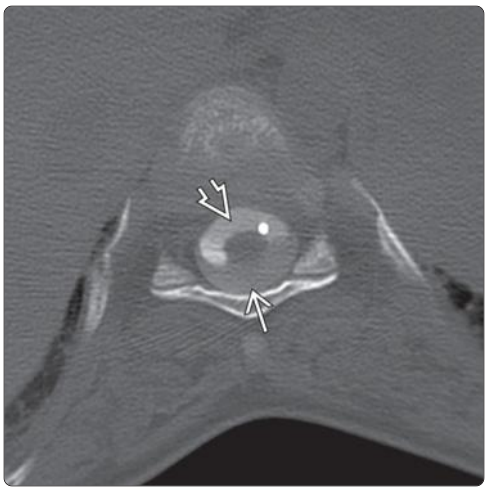


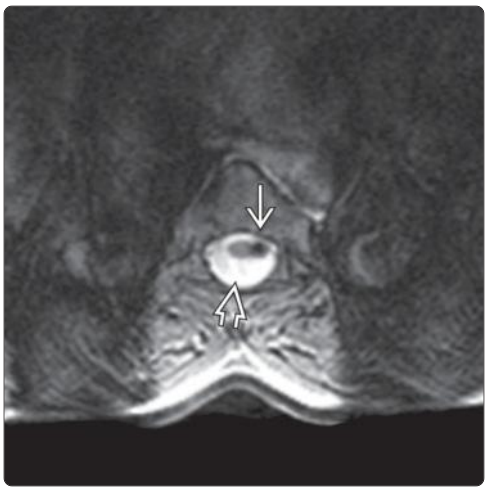
(Left) *Sagittal T2WI MR demonstrates a hyperintense CSF signal intensity intradural mass ⇨ that compresses and displaces the ventral spinal cord ⇨. Arachnoid cysts follow CSF signal intensity on all sequences unless complicated by hemorrhage.* **(Right)** *Axial T1W MR confirms a CSF signal intensity mass ⇨ that appears within the dural sac, with posterior displacement and flattening of the adjacent spinal cord ⇨.*





(Left) *Axial CT following myelography shows a central lesion ⇨ well-defined margins and decreased contrast density due to an intradural meningeal cyst. Higher attenuation of the normal subarachnoid space due to contrast is seen peripheral to the cyst ⇨.* **(Right)** *Sagittal T2W MR shows a CSF signal collection in the dorsal skin connecting to the thoracic thecal sac ⇨ in this patient with dorsal dysraphism. There is expansion of the bony canal ⇨ due to the associated meningeal cyst.*





(Left) *CT myelography shows differential amounts of contrast within the thecal sac due to partial filling of a posterior intradural meningeal cyst ⇨. The ventral CSF, with a Baclofen delivery catheter, shows a normal contrast amount ⇨.* **(Right)** *Axial GRE MR of the lower thoracic spine in this patient with a dorsal intradural meningeal cyst ⇨ shows the presence of a Baclofen delivery catheter ⇨ but cannot visualize the cyst margins.*

Perineural Root Sleeve Cyst

KEY FACTS

TERMINOLOGY

- Dilatation of arachnoid and dura of spinal posterior nerve root sheath, containing nerve fibers

IMAGING

- Occur anywhere along spine
  - Most common in lower lumbar spine and sacrum
  - S2 and S3 nerve roots most commonly involved
- Thin-walled cyst mass
  - Contents follow CSF density/signal intensity
  - No enhancement
- ± neural foraminal widening (bone remodeling)

TOP DIFFERENTIAL DIAGNOSES

- Facet synovial cyst
- Nerve sheath tumor (NST)
- Spinal nerve root avulsion
- Metastases
- Meningocele

PATHOLOGY

- Nabors classification of spinal meningeal cyst (MC)
  - Type I: Extradural MC without spinal nerve root fibers
    - IA: Extradural MC
    - IB: Occult sacral meningocele (outdated term)
  - Type II: Extradural MC with spinal nerve root fibers
  - Type III: Intradural MC

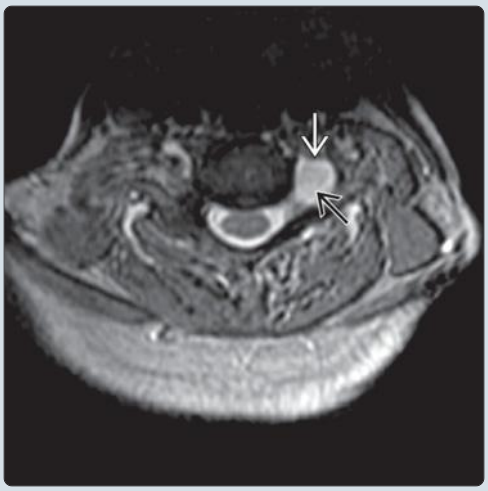
CLINICAL ISSUES

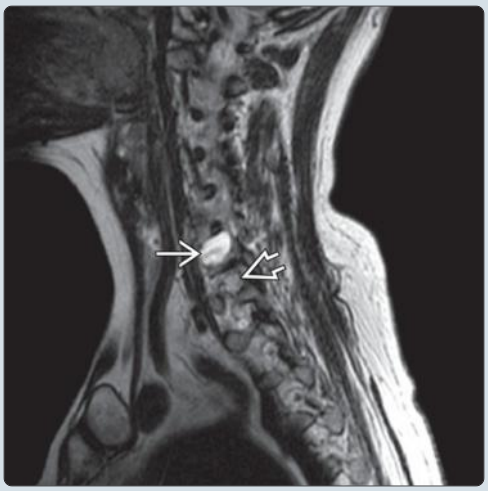
- Majority asymptomatic: > 80%
- Symptoms may worsen with postural changes, Valsalva maneuvers
- Cyst rupture → spontaneous intracranial hypotension
- Symptoms simulate disc herniation and spinal stenosis

DIAGNOSTIC CHECKLIST

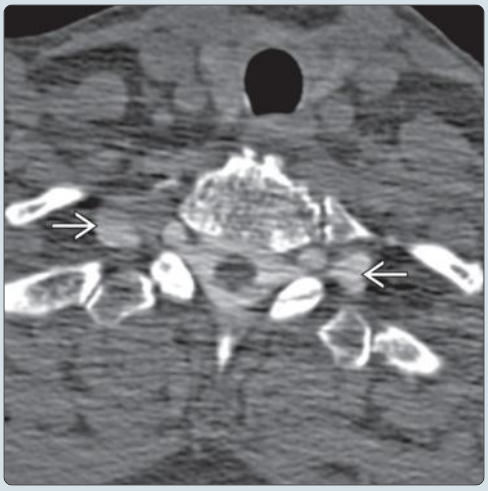
- CSF intensity mass enlarging neural foramen or sacral canal characteristic of perineural root sleeve cyst

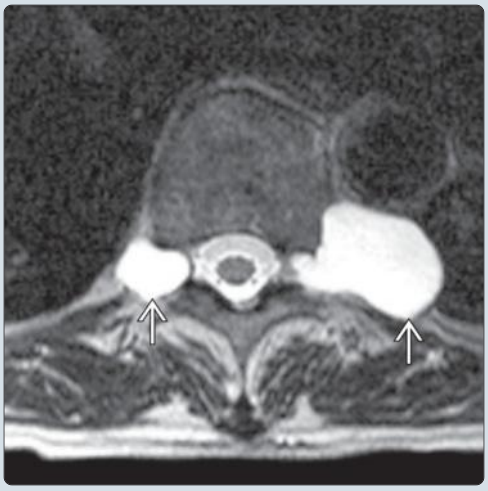
(Left) *Axial T2WI MR demonstrates a focal, well-circumscribed, hyperintense perineural cyst ➡ within the left C6-7 foramen. Subtle linear low signal ⇨ within the cyst represents the exiting nerve within it.* **(Right)** *Sagittal T2WI MR confirms focal expansion of the left C6-7 foramen by a large perineural root sleeve cyst ➡. Note hyperintense cyst signal compared to the normal dorsal root ganglia ➡ within the adjacent level neural foramina.*





(Left) *Axial CT myelography shows multiloculated bilateral T1 nerve root contrast-filled cysts ➡ confirming contiguity with the contrast-filled dural sac. Only part of the right cyst contains contrast.* **(Right)** *Axial T2WI MR of the thoracic spine demonstrates bilateral, well-circumscribed CSF intensity perineural cysts ➡ extending through the neural foramina. There is enlargement of the right neural foramen by the cyst.*





TERMINOLOGY

Synonyms

- Spinal nerve root diverticulum
- Tarlov cyst
- Perineural cyst

Definitions

- Dilatation of arachnoid and dura of spinal posterior nerve root sheath, containing nerve fibers

IMAGING

General Features

- Best diagnostic clue
  - Thin-walled cerebral spinal fluid (CSF) density/intensity masses extending into/through neural foramina
- Location
  - Anywhere along spinal axis
    - Most common in lower lumbar spine and sacrum
    - S2 and S3 nerve roots most commonly involved
- Size
  - Variable
- Morphology
  - Well circumscribed
  - Thin-walled cystic mass
  - Round, lobular, multiloculated
  - Often multiple and bilateral

Radiographic Findings

- Radiography
  - Limited diagnostic role
  - Enlarged neural foramen
  - Thinned pedicle

CT Findings

- NECT
  - Discrete CSF density mass
  - May see expanded neural foramen, thinned pedicle
    - Intact, remodeled cortex
- CECT
  - No enhancement
- Bone CT
  - Neural foraminal widening, pedicle thinning reflecting bone remodeling

MR Findings

- T1WI
  - Circumscribed intra-/transforaminal mass, isointense to CSF
- T2WI
  - Circumscribed intra-/transforaminal mass, isointense to CSF
- T1WI C+
  - No enhancement

Nonvascular Interventions

- CT myelography
  - Opacification of cyst by intrathecal contrast
  - Delayed imaging sometimes needed to show cyst filling

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI and T1WI MR
- Protocol advice
  - Gadolinium contrast to exclude solid mass if signal intensity not definitely cystic

DIFFERENTIAL DIAGNOSIS

Facet Synovial Cyst

- Posterolateral lumbosacral spine
- Abuts degenerative facet joint

Nerve Sheath Tumor (NST)

- Circumscribed, rounded, or fusiform mass
- Variable signal on MR
- + enhancement

Spinal Nerve Root Avulsion

- More common in lower cervical and upper thoracic spine
- Usually unilateral, may be multiple at contiguous levels
- History of trauma
- Immediate filling of intrathecal contrast on myelography

Metastases

- Soft tissue intensity mass usually enhances
- Intradural extramedullary
  - Foraminal enlargement due to bony destruction

Meningocele

- Anterior sacral meningocele
  - Solitary, CSF intensity lesion in pelvis
    - Protrusion of meninges through anterior sacral defect
  - ± sacral anomaly (scimitar) and anorectal malformation
    - Currarino triad
- Distal or occult intrasacral meningocele
  - Type IB meningeal cyst
    - No neural elements
  - CSF intensity mass filling sacral canal
  - Connected to caudal tip of thecal sac by a pedicle
- Lateral thoracic meningocele
  - Sharp angle thoracic scoliosis
  - Neurofibromatosis type I in 75-85% of cases

PATHOLOGY

General Features

- Etiology
  - Controversial origin, congenital vs. acquired lesion
- Genetics
  - No genetic predisposition
- Pronounced remodeling/thinning of adjacent bones may result in insufficiency fracture
- Cyst rupture may cause spontaneous intracranial hypotension

Staging, Grading, & Classification

- Nabors classification of spinal meningeal cyst (MC)
- Type I: Extradural MC without spinal nerve root fibers
  - IA: Extradural MC
    - a.k.a. extradural arachnoid cyst
    - Relatively rare

  - Dorsal or dorsal-lateral lower thoracic spine
  - May extend into neural foramina
- IB: Occult sacral meningocele (outdated term)
  - Actually sacral extradural arachnoid cyst
- Type II: Extradural MC with spinal nerve root fibers
  - Tarlov perineural cyst
  - Spinal nerve root diverticulum
- Type III: Intradural MC
  - a.k.a. intradural arachnoid cyst
  - Posterior aspect of midthoracic spine

Gross Pathologic & Surgical Features

- Origin at junction of dorsal nerve root and its ganglion
- Spinal nerve root traverses through cyst or within cyst wall
- ± communication with subarachnoid space

Microscopic Features

- Outer wall with epineurium lined by arachnoid
- Inner wall lined with pia mater
- Fibrocollagenous tissue present
- Old hemorrhage
- Nerve fibers (75%) or ganglionic cells (25%) within cyst or cyst wall

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Majority asymptomatic: > 80%
- Other signs/symptoms
  - Low back or sacral pain
  - Radicular or perineal pain
  - Paresthesia
  - Leg weakness
  - Bladder or bowel dysfunction
- Clinical profile
  - Symptoms simulate disc herniation and spinal stenosis
  - Symptoms may worsen with postural changes and Valsalva maneuvers

Demographics

- Age
  - 30-40 years old
- Gender
  - M = F
- Ethnicity
  - No racial predilection
- Epidemiology
  - Common, incidental, usually asymptomatic
  - 4.6-9% of adults

Natural History & Prognosis

- Progressive cyst enlargement
- Cyst may recur following aspiration or partial resection
- Symptomatic relief after complete resection: > 70%
  - Better outcome associated with
    - Cysts < 1.5 cm
    - Radicular pain
    - Bladder/bowel dysfunction
    - Worsening symptoms with Valsalva maneuver

Treatment

- Conservative medical
  - Anti-inflammatory medication
  - Physical therapy
- Interventional
  - Temporary relief with cyst aspiration
    - Usually several months symptom relief
- Surgical
  - Sacral laminectomies with microsurgical cyst fenestration and imbrication
  - Complete cyst/nerve root resection

DIAGNOSTIC CHECKLIST

Consider

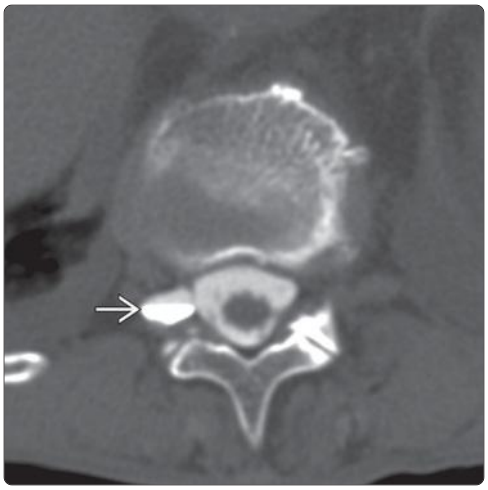
- No reliable imaging method to differentiate symptomatic from asymptomatic lesions

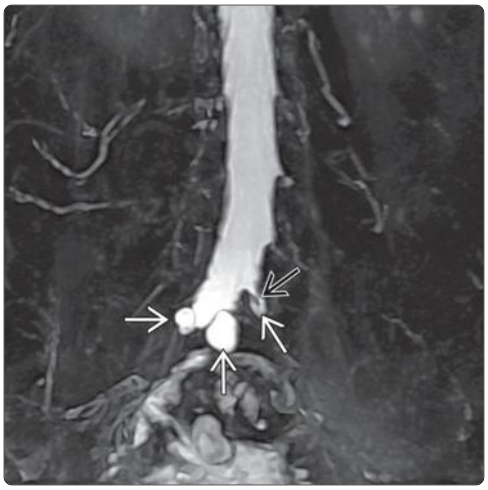
Image Interpretation Pearls

- Sacral CSF intensity mass enlarging neural foramen or sacral canal characteristic of perineural root sleeve cyst
- Delayed CT (30-60 minutes) after intrathecal contrast injection to visualize filling of perineural cyst

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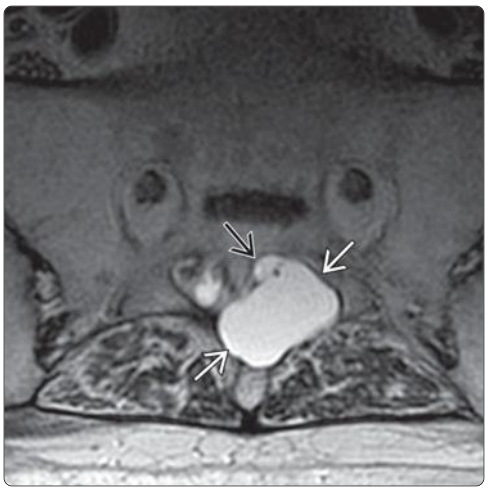
(Left) *Axial CT myelography shows a right-sided perineural cyst ➡ in the lower thoracic spine that opacifies with intrathecal contrast. Note the dependent contrast-CSF level in the cyst representing incomplete mixing.* **(Right)** *MR myelography demonstrates several sacral nerve root sleeve (Tarlov) cysts ➡. One of the left cysts contains a nerve root ⇨.*





(Left) *Axial T2WI MR at the L5 level reveals an extradural CSF intensity mass ➡ within the left lateral recess.* **(Right)** *Axial T1WI C+ MR in the same patient confirms absence of intrinsic or wall contrast enhancement, compatible with a typical perineural root sleeve cyst ➡.*





(Left) *Axial CT myelogram in the same patient demonstrates contrast filling within the perineural cyst ➡, confirming cyst contiguity with the thecal sac.* **(Right)** *Axial T2WI MR through the sacrum in a different patient reveals a large, eccentric cyst ➡ of the 2nd left sacral nerve root sleeve. The large cyst produces ventral displacement of the S2 nerve dorsal root ganglion ⇨.*

Syringomyelia

KEY FACTS

TERMINOLOGY

- Hydromyelia = cystic central canal dilatation
- Syringomyelia = cystic spinal cord cavity not contiguous with central cord canal
- Syringobulbia = brainstem syrinx extension

IMAGING

- Expanded spinal cord due to dilated, beaded, or sacculated cystic cavity
- Normal or canal enlargement, vertebral scalloping (severe, longstanding syrinx)

TOP DIFFERENTIAL DIAGNOSES

- Ventriculus terminalis
- Cystic spinal cord tumor
- Myelomalacia

PATHOLOGY

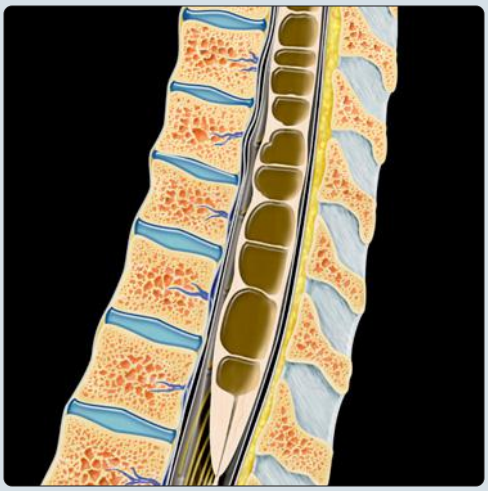
- Hydrocephalus, Chiari 1 or 2 malformation, myelomeningocele or other spinal dysraphism, tethered cord, congenital scoliosis, spinal cord injury

CLINICAL ISSUES

- Cloak-like pain and temperature/sensory loss with preservation of position sense, proprioception, light touch
- Distal upper extremity weakness, gait instability
- Cranial neuropathy (2° to syringobulbia)

DIAGNOSTIC CHECKLIST

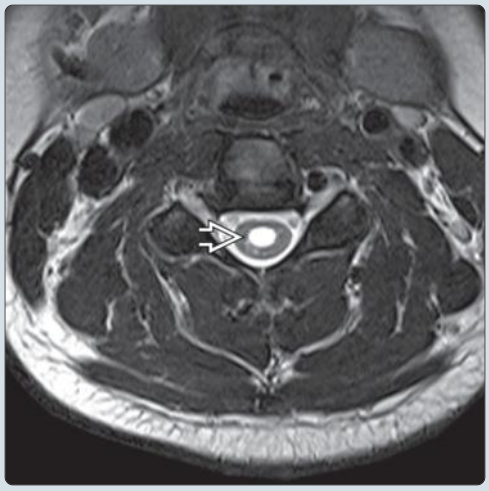
- Despite septated appearance, large syrinx cavities usually contiguous
- Contrast administration essential to exclude tumor in complicated cavitary lesions





(Left) *Sagittal graphic demonstrates a large, sacculated, "beaded" spinal cord syrinx extending to the conus. Despite the loculated appearance of large syringes, the individual fluid spaces are contiguous and drainable using a single shunt catheter.* **(Right)** *Sagittal T1WI MR (Chiari 2 malformation, not shown) depicts a large sacculated spinal cord syrinx that extends the entire length of the spinal cord into the low-lying terminal spinal cord that inserts into the dural closure at L4.*





(Left) *Sagittal T2WI FS MR (Chiari 1 malformation) shows inferior displacement of pointed ectopic cerebellar tonsils ⇨ below the foramen magnum. Note associated cervical syringohydromyelia ⇨.* **(Right)** *Axial T2WI MR (Chiari 1 malformation, not shown) reveals typical cervical spinal cord syringohydromyelia ⇨ characterized by smooth dilation of the central spinal cord canal and absence of nodularity, eccentric cavitation, or myelomalacia.*

TERMINOLOGY

Definitions

- Hydromyelia = cystic central canal dilatation
- Syringomyelia = cystic spinal cord cavity not contiguous with central cord canal
  - Syringobulbia = brainstem syrinx extension
  - Syringocephaly = brain/cerebral peduncle syrinx extension
- Syringohydromyelia = features of both syringomyelia and hydromyelia
- "Presyrinx state" = reversible spinal cord edema produced by alterations in CSF flow dynamics

IMAGING

General Features

- Best diagnostic clue
  - Expanded spinal cord due to dilated, beaded, or sacculated cystic cavity
- Location
  - Intramedullary spinal cord
- Size
  - Small → markedly dilated
- Morphology
  - Cystic intramedullary lesion; small syringes are tubular, while large are beaded or loculated with septations

Radiographic Findings

- Radiography
  - Normal or wide osseous canal
  - ± atrophic neuroarthropathy (cervical syrinx)

CT Findings

- CECT
  - Cord expansion, nonenhancing CSF-density spinal cord cavitation
- Bone CT
  - Normal or canal enlargement, vertebral scalloping (severe, longstanding syrinx)

MR Findings

- T1WI
  - Hypointense spinal cord cleft
    - Sagittal images demonstrate longitudinal extent
    - Axial images confirm syrinx topology, clarify relationship to adjacent structures
- T2WI
  - Hyperintense intramedullary cavity ± adjacent gliosis, myelomalacia
    - ± arachnoidal adhesions, cord tethering, or edema
- T1WI C+
  - Nonenhancing cavity; enhancement suggests inflammatory or neoplastic lesion

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic intramedullary cavitation or central canal dilatation

Nonvascular Interventions

- Myelography
  - Expanded spinal cord ± spinal canal widening
  - Delayed syrinx contrast uptake

Other Modality Findings

- 2D MR cine phase contrast (PC) CSF flow study
  - ± abnormal CSF dynamics across tonsils (Chiari 1)
  - Normal conus motion argues against tethered cord
  - Larger syrinx commonly shows internal CSF flow artifact

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal and axial T1WI, T2WI, T1 C+ MR
  - Cine PC CSF flow MR

DIFFERENTIAL DIAGNOSIS

Ventriculus Terminalis

- Asymptomatic (normal) dilatation of terminal cord central canal only

Cystic Spinal Cord Tumor

- Cord expansion, cystic cavity surrounded by abnormal T2 signal, nodular enhancement

Myelomalacia

- Cord volume loss, gliosis
- No CSF signal cavitation on T1WI

PATHOLOGY

General Features

- Etiology
  - Actively debated; 2 most popular theories not mutually exclusive
    - Abnormal subarachnoid space drives CSF into cord through perivascular spaces → formation, enlargement of syrinx
      - Arachnoid adhesions, mass, Chiari malformation, spinal dysraphisms, diastematomyelia
    - Cord destruction 2° to primary disease process (trauma, infectious or inflammatory myelitis, spondylosis) → cord cavitation
- Associated abnormalities
  - Hydrocephalus, Chiari 1 or 2 malformation, myelomeningocele or other spinal dysraphism, tethered cord, congenital scoliosis
- Longitudinally oriented CSF-filled cavity ± surrounding gliosis, myelomalacia
- Associated with scoliosis (↑ with left curve, high or low apex/end vertebra, male sex, Chiari 1 or 2 malformation)
- Hydromyelia: Dilated ependymal-lined central canal
  - "Spinal cord hydrocephalus"
  - Central canal patency determines syrinx location, extent
  - Slit-like central canal remnant detected in small percentage of asymptomatic adults
- Syringomyelia: Paracentral spinal cord cavitation lined by gliotic parenchyma independent of central canal
  - ± eccentric extension into anterior commissure, posterior horn

- ↑ intramedullary pressure → neurological dysfunction 2° to compression of long tracts, neurons, and microcirculation
- "Presyrinx state": Reversible cervical spinal cord edema caused by CSF flow dynamic alteration that does not produce frank cord cavitation
  - Usually described with Chiari 1 or 2 malformation obstruction to CSF flow at foramen magnum

Gross Pathologic & Surgical Features

- Dilated cavity ± contiguous with central canal
- ± arachnoid adhesions, mass lesion, tethered cord, Chiari malformation, blood products/trauma detritus

Microscopic Features

- Central canal dilatation communicates directly with 4th ventricle
  - Cavity lined by ependyma; length influenced by age-related central canal stenosis
- Noncommunicating (isolated) central canal dilation below syrinx-free cord segment
  - Variable distance below 4th ventricle; cavity margins defined by adjacent central canal stenosis
  - ± paracentral dissection into parenchyma, gliosis
- Extracanalicular (parenchymal) syrinx noncontiguous with central canal
  - Cord watershed area ± myelomalacia, paracentral dissection lined by glial/fibroglial tissue, central chromatolysis, neuronophagia, wallerian degeneration

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Cloak-like pain and temperature/sensory loss with preservation of position sense, proprioception, light touch
  - Distal upper extremity weakness, gait instability
  - Other signs/symptoms
    - Mechanical spinal pain, radicular pain, spastic paraparesis, scoliosis
    - Cranial neuropathy (2° to syringobulbia)
- Clinical profile
  - Symmetrically enlarged central cavity asymptomatic or presents with nonspecific neurological signs (spasticity, weakness, segmental pain)
  - Paracentral cavitation associated with combination of long tract and segmental signs referable to level, side, and quadrant of spinal cord cavitation

Demographics

- Age
  - Usual presentation in late adolescence or early adulthood; uncommon in childhood
- Gender
  - M = F
- Epidemiology
  - Primary syrinx usually in young patients
    - ↑ prevalence with basilar invagination, Chiari 1 or 2 malformation
  - Secondary syrinx at any age; timing of appearance determined by primary disease behavior
    - 25% of spinal cord injury (SCI) patients develop syrinx

Natural History & Prognosis

- Variable; dependent on underlying etiology
  - Chronic, slow progression usual; occasional acute course
  - Spontaneous resolution rare
- ± reversibility after Chiari decompression, mass removal, adhesion lysis, or cord untethering
  - Long-term improvement in < 50%
  - Sensory disturbances, dysesthesias, pain more likely to improve than motor weakness or gait disturbance
- CSF diastolic flow determines cyst size, biological behavior
  - Diastolic flow velocity ↓ after successful surgery; persistent ↑ implies poor outcome

Treatment

- Primarily address underlying causative etiology
  - Correct osseous deformities, decompress/untether spinal cord, lyse adhesions
  - Consider syrinx drainage with indwelling catheter only if not possible to restore normal cord CSF dynamics

DIAGNOSTIC CHECKLIST

Consider

- Syrinx etiology influences treatment approach
- Despite septated appearance, large syrinx cavities usually contiguous

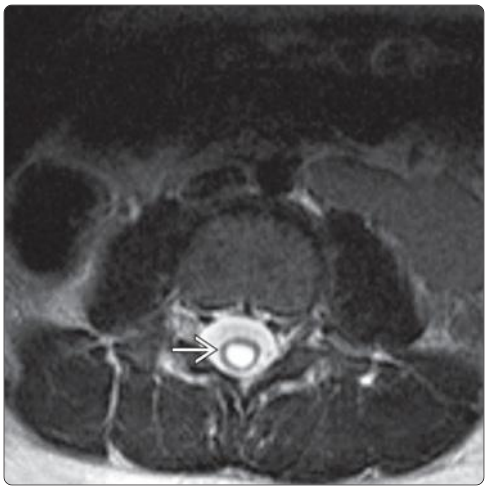
Image Interpretation Pearls

- Simple syringomyelia rarely enhances or produces diagnostic dilemma
- Contrast administration essential to exclude tumor in complicated cavitary lesions

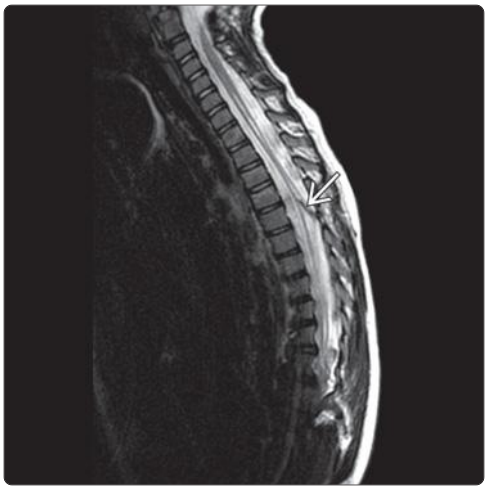
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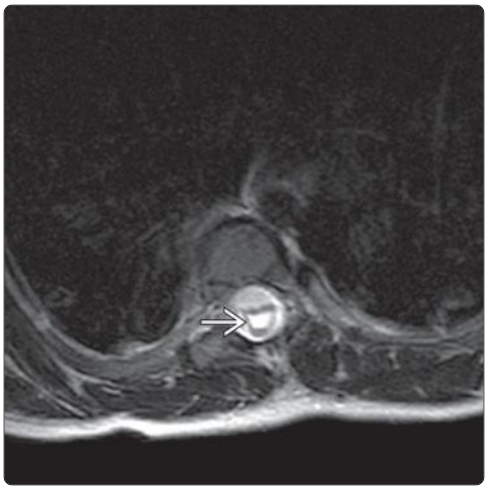
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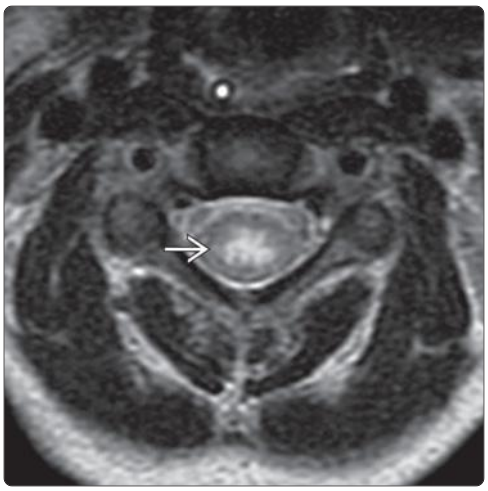
(Left) *Sagittal T1WI MR (clinical tethered spinal cord) demonstrates a low-lying, hydromyelic ⇨ tethered spinal cord inserting into a sacral lipomyelomeningocele ⇨.* **(Right)** *Axial T2WI MR (sacral lipomyelomeningocele with clinical tethered spinal cord) reveals CSF signal intensity dilation of the central spinal cord canal that is characteristic of hydrosyringomyelia ⇨.*





(Left) *Sagittal T2WI MR (Chiari 2 malformation, myelomeningocele) of the thoracic spine reveals a small syrinx cavity within the atrophic spinal cord. A hypointense syringopleural shunt catheter is visible ⇨, extending into the syrinx cavity.* **(Right)** *Axial T2WI MR (Chiari 2 malformation, surgically closed myelomeningocele at birth with syringopleural shunt placed later for syringomyelia) depicts an irregularly shaped syringomyelia cavity ⇨ within thoracic spinal cord.*





(Left) *Sagittal T2WI MR (Chiari 1 malformation) shows marked cerebellar tonsillar ectopia ⇨ with a normal tectal plate and 4th ventricular position, characteristic of Chiari 1 malformation. The clivus ⇨ is also mildly foreshortened. Note the intramedullary edema without frank syringomyelia ⇨, representing a "presyrinx state."* **(Right)** *Axial T2WI MR (Chiari 1 malformation) shows central intramedullary edema in the cervical spinal cord without frank syringomyelia ⇨ ("presyrinx state").*

Epidural Lipomatosis

KEY FACTS

TERMINOLOGY

- Excessive accumulation of intraspinal fat causing cord compression and neurologic deficits

IMAGING

- Thoracic spine: 58-61%
  - T6-T8, dorsal to spinal cord
  - Epidural fat ≥ 7 mm thick
- Lumbar spine: 39-42%
  - L4-L5, surrounding thecal sac
- Y-shaped configuration to lumbar thecal sac on axial imaging
- Mass effect on thecal sac and nerve roots
- Follows fat signal intensity on all sequences
  - Fat suppression to confirm adipose tissue, exclude blood products

TOP DIFFERENTIAL DIAGNOSES

- Subacute epidural hematoma
- Spinal angiolipoma
- Epidural metastasis
- Epidural abscess

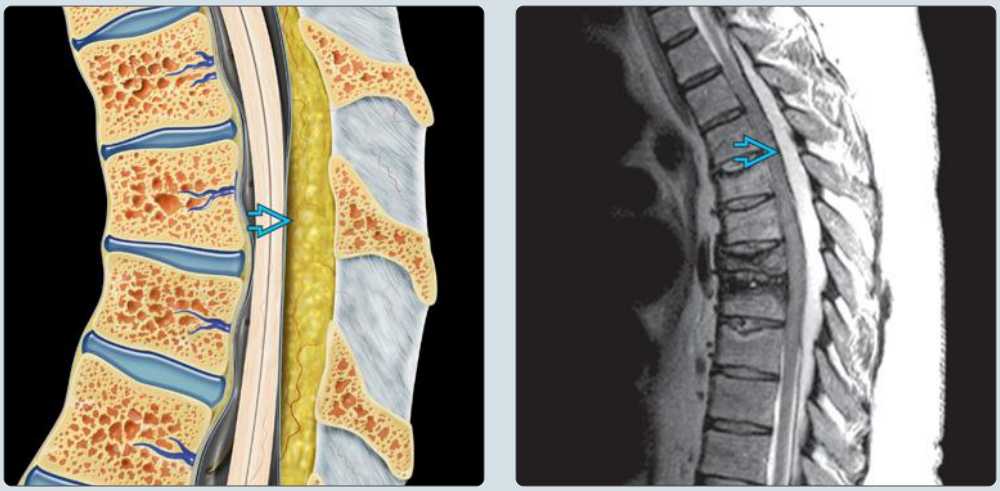
PATHOLOGY

- Exogenous or endogenous steroids
- General obesity
- Idiopathic

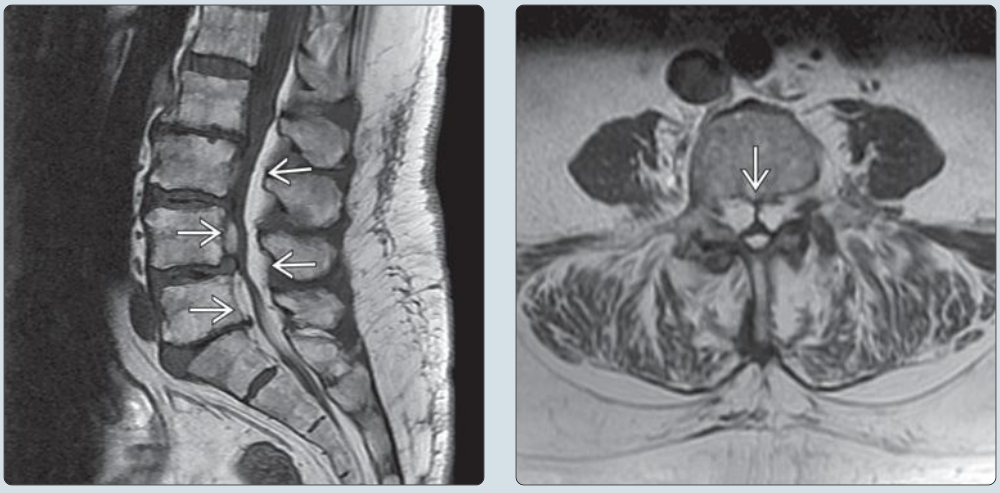
CLINICAL ISSUES

- Gradual progression of symptoms
- Weakness: > 85%
- Back pain, sensory loss, polyradiculopathy, altered reflexes, incontinence, ataxia

(Left) *Sagittal graphic illustrates confluent abundant epidural fat in dorsal thoracic canal, with effacement of the dorsal thecal sac and mild mass effect on spinal cord, which is displaced ventrally.* **(Right)** *Sagittal T2WI MR of thoracic spine shows smooth and homogeneous dorsal epidural tissue, isointense to subcutaneous fat. Vertebral compression fractures are due to steroid-induced osteoporosis.*



(Left) *Sagittal T1 MR shows abundant epidural fat both dorsal and ventral to the thecal sac. The thecal sac is small in AP diameter due to compression by the fat.* **(Right)** *Axial T1 MR shows marked prominence of the epidural fat in the lumbar canal, which causes the thecal sac to assume a trefoil pattern. The abundant fat in the epidural space allows for good visualization of the normal anatomy of the Hoffman ligament, which connects the posterior longitudinal ligament to the ventral dura.*



TERMINOLOGY

Definitions

- Excessive accumulation of intraspinal fat causing cord compression and neurologic deficits

IMAGING

General Features

- Best diagnostic clue
  - Abundant epidural fat in midthoracic and distal lumbar spinal canal compressing thecal sac
- Location
  - Thoracic spine: 58-61%
    - T6-T8, dorsal to spinal cord
  - Lumbar spine: 39-42%
    - L4-L5, surrounding thecal sac
- Size
  - Epidural fat ≥ 7 mm thick in thoracic spine
  - Over multiple vertebral segments
- Morphology
  - Y-shaped configuration to lumbar thecal sac on axial imaging

CT Findings

- Increased fat in spinal canal
- Compression of cord or thecal sac
- No abnormal enhancement
- No bony erosions

MR Findings

- Mass effect on thecal sac and nerve roots
  - Obliterated CSF, cord compression, crowded cauda equina
- Follows fat signal intensity on all sequences
- No abnormal enhancement

Nonvascular Interventions

- Myelography
  - Normal or effacement of CSF

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Fat suppression to confirm adipose tissue, exclude blood products

DIFFERENTIAL DIAGNOSIS

Subacute Epidural Hematoma

- Acute onset of symptoms
- High signal intensity on T1WI persists on fat-suppressed sequences

Spinal Angiolipoma

- Benign neoplasm of adipose and vascular elements
- Focal, rounded mass
- Infiltrative type invades surrounding bone
- Usually ↓ relative to fat on T1WI, ↑ on STIR
  - Due to vascular component
- Enhancement with gadolinium (Gd)

Epidural Metastasis

- Low signal intensity on T1WI, intermediate to high signal intensity on T2WI, STIR
- Enhancement with Gd

Epidural Abscess

- Hypointense on T1WI, hyperintense on T2WI
- Diffuse or peripheral enhancement with Gd

PATHOLOGY

General Features

- Etiology
  - Long-term exogenous steroid administration
  - Excessive endogenous steroid production
  - General obesity
  - Idiopathic

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Weakness: > 85%
- Other signs/symptoms
  - Back pain, sensory loss, polyradiculopathy, altered reflexes, incontinence, ataxia
- Clinical profile
  - Gradual progression of symptoms

Demographics

- Uncommon; seen in middle-aged patients (M > F)

Natural History & Prognosis

- Presurgical low steroid dose and idiopathic cases have better prognosis
- > 80% with postsurgical symptomatic relief

Treatment

- Correction of underlying endocrinopathies
- Weight reduction in case of general obesity
- Multilevel laminectomy and fusion
  - Indicated when cord compression and radiculopathy present

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Normal Fatty Marrow Variants

KEY FACTS

TERMINOLOGY

- Red marrow: Hematopoietic marrow
- Yellow marrow: Fatty marrow

IMAGING

- Yellow marrow often distributed around central perforating vein
- Yellow marrow adjacent to endplates in degenerative disc disease
- After bone marrow transplantation: Fat centrally located in vertebral body, red marrow at periphery
  - Yellow marrow high T1W signal intensity
  - Red marrow low to intermediate T1W signal intensity
- Signal intensity equal or slightly higher than muscle

TOP DIFFERENTIAL DIAGNOSES

- Leukemia
- Multiple myeloma
- Metastasis
- Myelofibrosis
- Hemangioma

PATHOLOGY

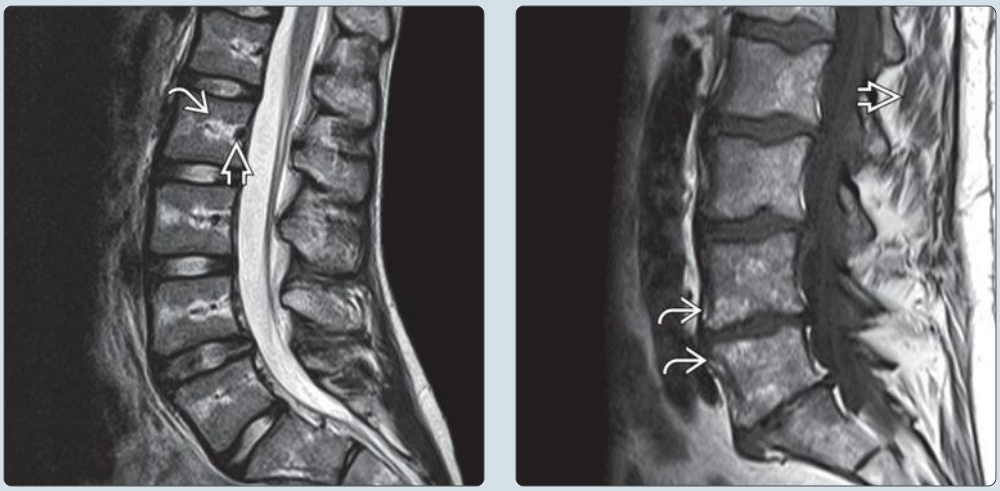
- Normal marrow pattern changes with age
- Primarily erythropoietic marrow in childhood
- Gradually converts to fatty marrow during adulthood
- Fatty marrow 1st seen around central draining vein
- Fatty, erythropoietic marrow often intermixed in marbled pattern
- Converts to predominantly fatty marrow in later adulthood
- Red marrow reconversion occurs with anemia, obesity, high athletic activity, bone marrow transplantation, and erythropoietic stimulating agents

CLINICAL ISSUES

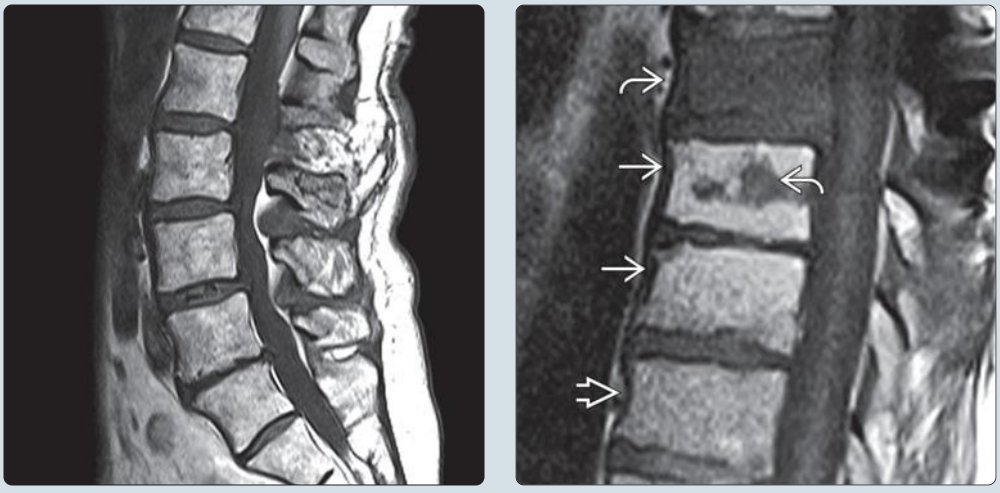
- Asymptomatic

DIAGNOSTIC CHECKLIST

- Marrow fat patterns vary with age, health



(Left) *Sagittal T2WI MR in a 15-year-old patient shows fat centered on the draining vein at each vertebral level. The remainder of the vertebral body contains red marrow, the usual pattern in young patients.* **(Right)** *Sagittal T1WI MR in a 50-year-old patient shows a polka dot appearance of yellow and red marrow. Fat tends to be centered around the draining veins and adjacent to degenerated discs. Red marrow is of equal or slightly higher signal intensity compared to muscle.*



(Left) *Sagittal T1WI MR in an 85-year-old patient shows marrow is predominantly fatty, but small foci of erythropoietic marrow are still present interspersed throughout. Although red marrow involutes with aging, there is always some red marrow in the spine, unless a patient has been treated with radiation therapy.* **(Right)** *Sagittal T1WI MR shows a difference in signal intensity between normal fatty marrow, irradiated marrow, and tumor infiltration.*

TERMINOLOGY

Definitions

- Red marrow: Hematopoietic marrow
- Yellow marrow: Fatty marrow

IMAGING

General Features

- Location
  - Vertebral bodies
  - Yellow marrow often distributed around central perforating vein
  - Yellow marrow adjacent to endplates in degenerative disc disease
  - After bone marrow transplantation: Fat centrally located in vertebral body, red marrow at periphery
  - Loss of red marrow in regions of radiation therapy

CT Findings

- Normal trabecular pattern preserved

MR Findings

- T1WI
  - Yellow marrow high signal intensity
  - Red marrow low to intermediate signal intensity
    - Signal intensity equal or slightly higher than muscle
- T2WI
  - Yellow marrow high signal intensity unless fat suppression used
  - Red marrow intermediate signal intensity, similar to muscle
- STIR
  - Yellow marrow low signal intensity
  - Red marrow low to intermediate signal intensity
- DWI
  - Not shown to be of use in distinguishing red marrow from tumor
- T1WI C+
  - Erythropoietic marrow enhances slightly
- In-phase and out-phase gradient-echo imaging
  - Contribution of fat and water to signal cycle in- and out-phase with increasing TE
  - Erythropoietic marrow contains fat and water: Low signal on out-phase images
  - Neoplasm usually does not contain fat: High signal on out-phase images
  - No large series to validate this concept
    - Early myeloma has same appearance as red marrow

Imaging Recommendations

- Best imaging tool
  - CT in questionable cases to exclude lytic lesions

DIFFERENTIAL DIAGNOSIS

Leukemia

- Early: May appear identical to red marrow on MR
- Late: Lower signal on T1WI than intervertebral disc
- CT scan shows focal trabecular destruction
- Out-phase MR: High signal (usually)

Multiple Myeloma

- Same appearance as leukemia

Metastasis

- Lower signal on T1WI than intervertebral disc
- Usually high signal on STIR, but intermediate in sclerotic metastasis
- Usually better defined, larger focus of marrow involvement than with red marrow
- Out-phase MR: High signal

Myelofibrosis

- Low signal intensity on all MR sequences

Hemangioma

- Round mass usually containing fat
- Corduroy appearance of thickened residual trabeculae traversing lesion

PATHOLOGY

General Features

- Etiology
  - Increased yellow marrow
    - Normal aging phenomenon
      - Marbled appearance of vertebrae
    - Radiation therapy
      - Corresponds to radiation ports
      - Erythropoietic elements more susceptible to radiation necrosis
    - Degenerative disc disease
      - Primarily adjacent to vertebral endplates
  - Increased red marrow
    - Reconversion from fatty to erythropoietic marrow
    - Anemia, obesity, high athletic activity, bone marrow transplantation, erythropoietic stimulating agents

Staging, Grading, & Classification

- Normal marrow pattern changes with age
  - Primarily erythropoietic marrow in childhood
  - Gradually converts to fatty marrow during adulthood
  - Fatty marrow 1st seen around central draining vein
  - Fatty, erythropoietic marrow often intermixed in marbled pattern
  - Converts to primarily fatty marrow in later adulthood
  - Anemia, obesity cause red marrow reconversion

DIAGNOSTIC CHECKLIST

Consider

- Marrow fat patterns vary with age, health

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Fibrous Dysplasia

KEY FACTS

TERMINOLOGY

- McCune-Albright syndrome: Polyostotic fibrous dysplasia (FD), precocious puberty, café au lait skin lesions

IMAGING

- Most spine lesions occur with polyostotic disease
- Often causes scoliosis
- Neural arch > vertebral body
- Fusiform expansion of bone
- Cortical thinning
- Commonly "ground-glass" matrix
  - However, matrix may range from purely lytic to purely sclerotic lesion
- Narrow zone of transition ± sclerotic margin
- Low to intermediate signal intensity on T1WI, heterogeneous on T2WI, STIR, variable enhancement
- Mild to marked increase in radionuclide uptake

TOP DIFFERENTIAL DIAGNOSES

- Aneurysmal bone cyst (ABC)
- Paget disease
- Osteoblastoma
- Osteosarcoma
- Tuberous sclerosis

PATHOLOGY

- Sporadic mutation in *GNAS* gene

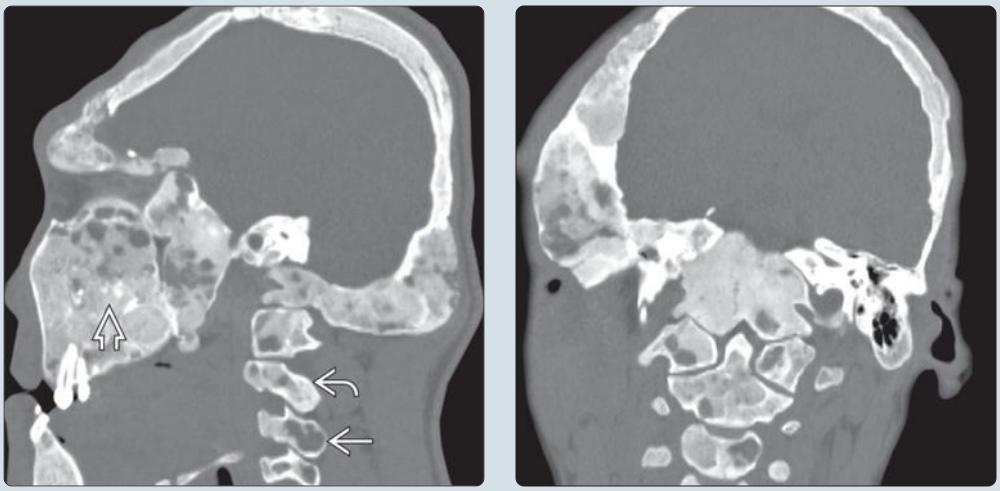
CLINICAL ISSUES

- Growth disturbance, pathologic fracture
- Rarely undergoes sarcomatous transformation

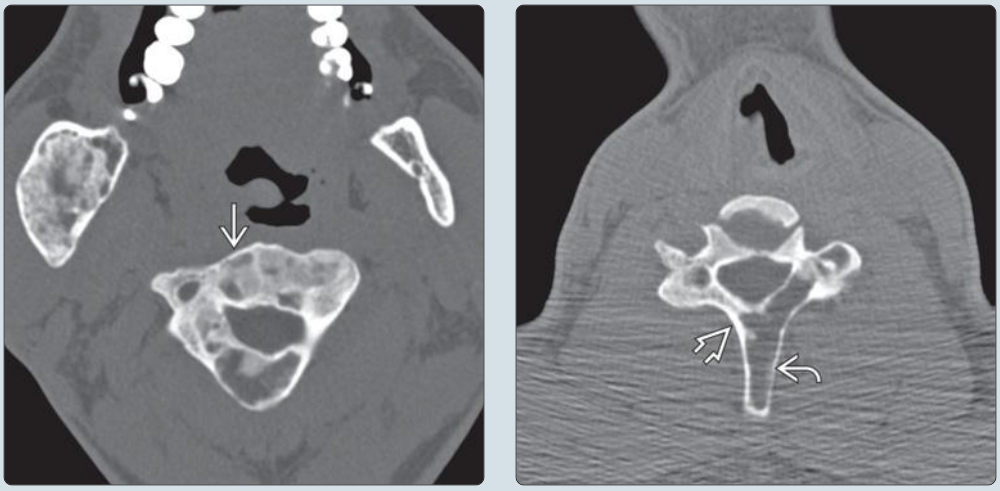
DIAGNOSTIC CHECKLIST

- Do not confuse with Paget disease on imaging
  - Paget disease thickens cortex and trabeculae
  - FD thins cortex and replaces trabeculae

(Left) *Sagittal bone CT shows severe polyostotic fibrous dysplasia (FD) involving skull, facial bones, and cervical spine. Some areas are "ground-glass", others are purely lytic, and there are a few foci of calcified cartilage.* **(Right)** *Coronal bone CT in the same patient shows loss of normal trabeculae in the majority of the included bones, replaced by fibrous dysplasia matrix. Thinning of tables of skull is a characteristic finding.*



(Left) *Axial bone CT shows variation in density in a single vertebra with nearly complete marrow replacement. This variability is common and should not raise suspicion for malignant degeneration.* **(Right)** *Axial bone CT shows lytic FD in the posterior elements. The narrow zone of transition (sometimes sclerotic) helps to distinguish FD from more aggressive processes. Vertebral bodies tend to be less severely involved than the posterior elements.*



TERMINOLOGY

Abbreviations

- Fibrous dysplasia (FD)

Definitions

- Monostotic: Single lesion
- Polyostotic: Multiple lesions, often associated with growth disturbances
- McCune-Albright syndrome: Polyostotic FD, precocious puberty, café au lait skin lesions

IMAGING

General Features

- Location
  - Rarely involves spine
    - Most spine lesions occur in patients with polyostotic disease
    - Neural arch > vertebral body

Radiographic Findings

- Radiography
  - Fusiform expansion of bone
  - Cortical thinning
  - Commonly ground-glass matrix
    - However, matrix may range from purely lytic to purely sclerotic lesion
    - May contain islands of cartilage
  - Narrow zone of transition ± sclerotic margin
  - Scoliosis

CT Findings

- Matrix, transition zone better seen than on radiographs
- Cortical breakthrough not uncommon

MR Findings

- T1WI
  - Low to intermediate signal intensity
- T2WI FS
  - Heterogeneous intermediate to high signal intensity
- T1WI C+
  - Variable enhancement

Nuclear Medicine Findings

- Bone scan
  - Mild to marked increase in radionuclide uptake
  - Radionuclide uptake not correlated to symptoms

Imaging Recommendations

- Best imaging tool
  - CT scan: 1-3 mm sections

DIFFERENTIAL DIAGNOSIS

Aneurysmal Bone Cyst (ABC)

- Balloon-like expansion of neural arch
- Fluid-fluid levels on CT or MR
- Can be associated with fibrous dysplasia

Paget Disease

- Lytic phase may appear similar
- Reparative phase distinctive: Cortical thickening, coarsened trabeculae, bone enlargement

Osteoblastoma

- Usually shows more discrete bone matrix
- May mimic "ground-glass" matrix

Tuberous Sclerosis

- Sclerotic bone lesions
- Characteristic brain abnormalities

Osteosarcoma

- ± amorphous, immature bone matrix
- Wide zone of transition
- Cortical breakthrough, soft tissue mass
- Periosteal reaction

PATHOLOGY

General Features

- Genetics
  - Sporadic mutation in *GNAS* gene
- Associated abnormalities
  - Endocrine abnormalities: Hyperparathyroidism, acromegaly, premature puberty, phosphaturia
  - May have associated ABC

Gross Pathologic & Surgical Features

- White or tan lesion with gritty consistency

Microscopic Features

- Stroma of bland fibrous tissue
- Small spicules of woven bone ("alphabet soup")
- Occasionally has foci of cartilage

CLINICAL ISSUES

Natural History & Prognosis

- Growth disturbance, pathologic fracture common in polyostotic form
- Rarely undergoes sarcomatous transformation to osteosarcoma or malignant fibrous histiocytoma

DIAGNOSTIC CHECKLIST

Consider

- Do not confuse with Paget disease on imaging
  - Paget disease thickens cortex and trabeculae
  - FD thins cortex and replaces trabeculae

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Kümmell Disease

KEY FACTS

TERMINOLOGY

- Post-traumatic avascular necrosis of vertebral body

IMAGING

- Radiographs, CT
  - Loss of height, sclerosis of vertebral body
  - Narrow, horizontally oriented band of gas in vertebral body
  - Filling cleft in vertebral body due to fracture nonunion
- MR
  - Vertebral body collapse
  - Gas-filled cleft is low signal intensity on all sequences (unless fluid-filled)
  - Fracture line may or may not be visible
  - Less well seen than on CT scan

TOP DIFFERENTIAL DIAGNOSES

- Infection
- Nontraumatic bone infarction
- Gas within degenerated intervertebral discs
- Calcium pyrophosphate dihydrate deposition

PATHOLOGY

- Radiographically occult vertebral body clefts common in patients with fracture
- Nonunited vertebral body fracture cleft undergoes 2° necrosis, collapse
- Nitrogen accumulates in fracture cleft

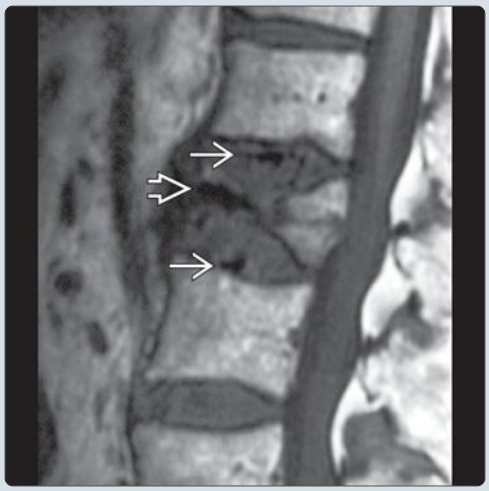
CLINICAL ISSUES

- Presents with pain, kyphosis
- Usually occurs in elderly, osteoporotic patients
- Progressive vertebral body collapse if untreated

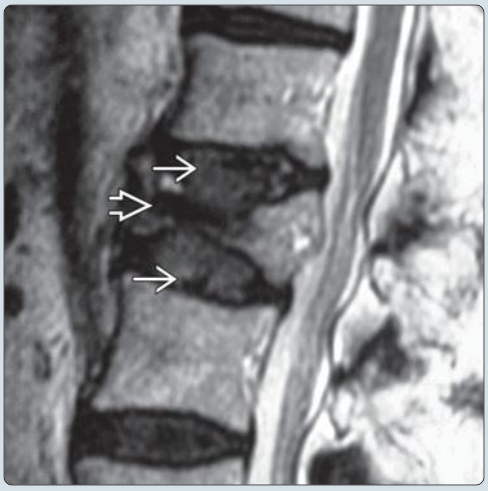
DIAGNOSTIC CHECKLIST

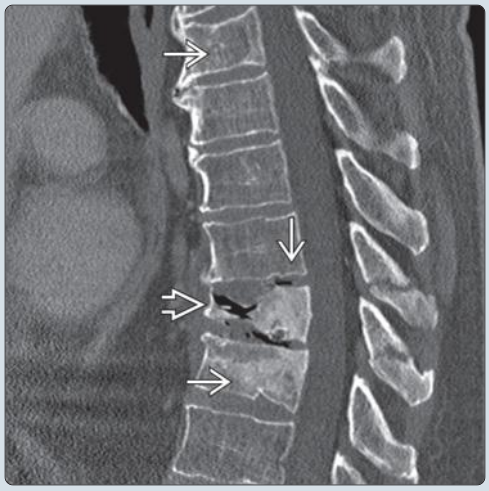
- Kümmell disease may be rarely associated with pathologic fracture





(Left) *Coronal bone CT shows gas within a collapsed, sclerotic vertebral body. Gas is also present in the adjacent disc spaces. The gas seen in Kümmell disease probably migrates into the vertebral body from a degenerated disc.* **(Right)** *Sagittal T1WI MR shows gas in an osteoporotic burst fracture and adjacent discs. Gas could easily be mistaken for calcification on the MR.*





(Left) *Sagittal T2WI MR reveals band-like gas in the vertebral body and gas in the intervertebral discs. The findings are less conspicuous and less specific than demonstrated with bone CT.* **(Right)** *Sagittal bone CT shows gas in a collapsed vertebral body. This unusual case has occurred in a patient with metastatic disease. Kümmell disease should not be presumed to always indicate a benign compression fracture. This patient had blastic metastases from prostate carcinoma.*

TERMINOLOGY

Definitions

- Post-traumatic avascular necrosis of vertebral body

IMAGING

General Features

- Best diagnostic clue
  - Gas-filled cleft in flattened vertebral body
- Location
  - Thoracic or lumbar vertebral body

Radiographic Findings

- Radiography
  - Loss of height, sclerosis of vertebral body
  - Narrow, horizontally oriented band of gas in vertebral body
  - Filling cleft in vertebral body due to fracture nonunion

CT Findings

- Bone CT
  - Horizontal band of gas-filling cleft in vertebral body
  - Gas often in adjacent disc space

MR Findings

- Vertebral body collapse
- Fracture line may or may not be visible
  - Horizontal low signal intensity line on T1WI
  - Band-like high signal intensity on T2WI, STIR
- Gas-filled cleft
  - Low signal intensity on all sequences
    - May occasionally be fluid-filled, follow fluid signal intensity
  - Less well seen than on CT scan

Nuclear Medicine Findings

- Bone scan
  - Positive 3-phase bone scan

Imaging Recommendations

- Best imaging tool
  - CT scan
- Protocol advice
  - MDCT with sagittal, coronal reformations

DIFFERENTIAL DIAGNOSIS

Infection

- Small bubbles of gas sometimes present, but not a cleft
- Endplate destruction
- Heterogeneous enhancement of disc and vertebral body
- Fluid collections in paraspinous soft tissues

Nontraumatic Bone Infarction

- Serpentine contour of infarction with peripheral enhancement
- Associated with steroids, sickle cell disease, pancreatitis, vasculitis, emboli, and caisson disease

Gas Within Degenerated Intervertebral Discs

- Gas forms in degenerated discs, may enter Schmorl nodes

Calcium Pyrophosphate Dihydrate Deposition (CPPD)

- Calcifications appear similar to gas on MR

PATHOLOGY

General Features

- Etiology
  - Kyphoplasty data suggests radiographically occult vertebral body clefts are common in patients with fracture
  - Nonunited vertebral body fracture undergoes secondary necrosis and collapse
  - Nitrogen accumulates in fracture cleft

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, kyphosis

Demographics

- Age
  - Usually elderly, osteoporotic patients
- Epidemiology
  - Uncommon

Natural History & Prognosis

- Progressive vertebral body collapse if untreated

Treatment

- Options, risks, complications
  - Vertebroplasty or kyphoplasty relieves pain

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Kümmell disease rarely can occur in pathologic fracture due to tumor
- Gas may be seen in infection
  - Usually small bubbles, not linear cleft

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Hirayama Disease

KEY FACTS

TERMINOLOGY

- Cervical myelopathy related to anterior displacement of posterior cervical dura with flexion

IMAGING

- Asymmetric cord atrophy
- Flexion study shows increased posterior epidural space with ventral dural displacement, cord compression
- T1WI C+ shows enhancing enlarged posterior epidural space with flexion

TOP DIFFERENTIAL DIAGNOSES

- Motor neuron disease
- Chronic radiculopathy

PATHOLOGY

- Tight dural canal during flexion related to disproportionate length between vertebral column and dural canal
- Generally sporadic, but familial cases have been reported
- Anterior horn cells of spinal cord levels C5-T1 show shrinkage, degeneration, and necrosis with mild gliosis

CLINICAL ISSUES

- Nonprogressive muscular atrophy confined to hand and forearm
  - 2nd to 3rd decades
- Avoidance of neck flexion can stop progression
- Selected patients treated with posterior decompression with duraplasty

DIAGNOSTIC CHECKLIST

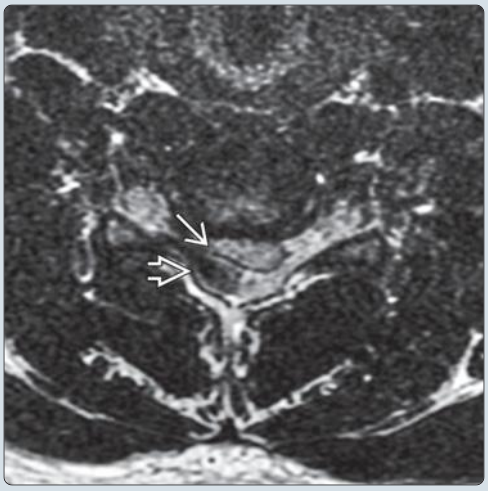
- Asymmetric atrophy of lower cervical cord on routine MR in patient with distal upper limb weakness is highly suspicious for Hirayama
  - Flexion cervical MR recommended





(Left) *Sagittal T2WI MR shows the classic appearance of Hirayama disease. The neutral position of the MR shows mild cord atrophy at the C5-C6 level, but is otherwise normal.* **(Right)** *Flexion T2WI MR shows marked ventral displacement of the posterior dural margin with cord compression. A hypointense T2 signal filling the dorsal epidural space is a distended venous plexus, which will homogeneously enhance with contrast (not shown).*





(Left) *Flexion T1WI MR shows marked ventral displacement of the posterior dural margin with a long segment of cord compression. A slightly heterogeneous signal filling the dorsal epidural space reflects a distended venous plexus.* **(Right)** *Axial T2WI MR in flexion shows a markedly distended posterior epidural plexus. There is ventral displacement of the posterior dura with asymmetrical cord compression, worse on the right.*

TERMINOLOGY

Synonyms

- Hirayama flexion myelopathy, juvenile spinal muscular atrophy, monomelic amyotrophy, juvenile asymmetric segmental spinal muscular atrophy

Definitions

- Cervical myelopathy related to anterior displacement of posterior cervical dura with flexion

IMAGING

General Features

- Best diagnostic clue
  - Anterior displacement of posterior cervical dura on MR
- Location
  - Cervical
- Size
  - Variable
- Morphology
  - Anterior displacement of linear posterior dura

CT Findings

- Myelography shows unilateral cord atrophy
- Flexion studies difficult due to contrast movement

MR Findings

- T1WI
  - Asymmetric cord atrophy
  - Flexion study shows increased posterior epidural space with ventral dural displacement, cord compression
- T2WI
  - Asymmetric cord atrophy, cord hyperintensity at atrophic area
  - Flexion study shows increased posterior epidural space with ventral dural displacement, cord compression
- T1WI C+
  - Enhancing enlarged posterior epidural space with flexion

Imaging Recommendations

- Best imaging tool
  - Flexion MR

DIFFERENTIAL DIAGNOSIS

Motor Neuron Disease

- Diagnosis of exclusion without spinal imaging findings

Chronic Radiculopathy

- Disc disease

PATHOLOGY

General Features

- Etiology
  - Controversial
  - Dynamic spinal cord compression with neck flexion
  - Tight dural canal during flexion related to disproportionate length between vertebral column and dural canal
    - Length of cervical canal increases with flexion
    - Dural sac compensates for this lengthening in normal subjects
    - Imbalance of growth of vertebrae and dura in Hirayama causes tight dural canal with flexion with anterior shift
- Genetics
  - Generally sporadic, but familial cases have been reported

Gross Pathologic & Surgical Features

- Anterior horn cells of spinal cord levels C5-T1 showed shrinkage, degeneration, and necrosis with mild gliosis
  - Suggested circulatory insufficiency in these areas related to chronic trauma of cord compression
- Abnormal posterior dura with few elastic fibers

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Muscular atrophy confined to hand and forearm
  - Usually unilateral, but can be bilateral
  - Disease onset insidious
  - No sensory or pyramidal tract involvement

Demographics

- Age
  - 2nd to 3rd decades
- Gender
  - M > F

Natural History & Prognosis

- Clinical course characterized by steady progression with eventual stabilization

Treatment

- Avoidance of neck flexion can stop progression
  - Cervical collar for 3-4 years
- Selected patients treated with posterior decompression with duraplasty, rarely anterior fusion

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Asymmetric atrophy of lower cervical cord on routine MR in patient with distal upper limb weakness suspicious for Hirayama
  - Flexion MR recommended

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IgG4-Related Disease/Hypertrophic Pachymeningitis

KEY FACTS

TERMINOLOGY

- Idiopathic hypertrophic pachymeningitis (IHP)
- IgG4-related hypertrophic pachymeningitis (IgG4-RHP)
- Multiorgan immune mediated condition, which mimics multiple malignant, infectious, and inflammatory disorders
  - Chronic progressive diffuse inflammatory fibrosis of dura of brain or spine

IMAGING

- Linear low signal mass involving dura with variable mass effect upon cord
- Low signal linear mass effacing CSF and cord
- Peripheral enhancement related to peripheral zone of active inflammation vs. chronic central fibrosis

TOP DIFFERENTIAL DIAGNOSES

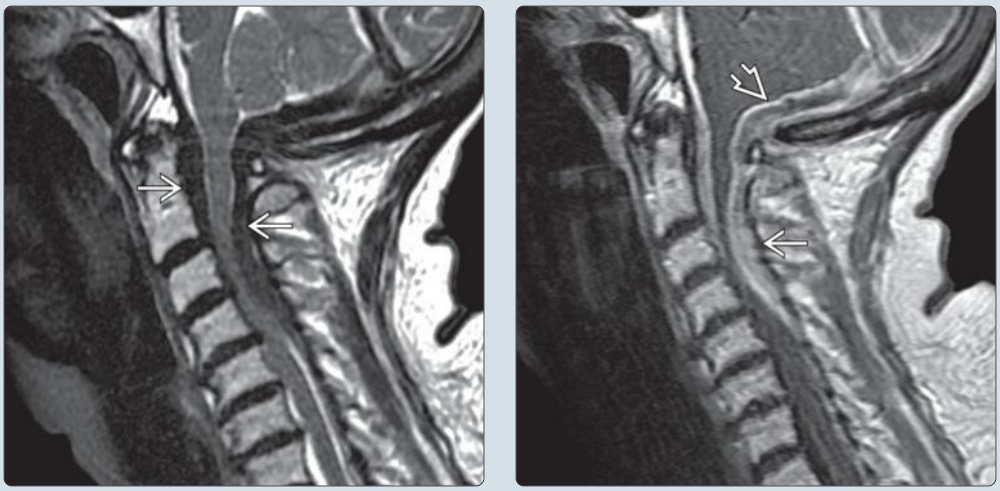
- Meningioma
- Lymphoma
- Tuberculosis
- Sarcoidosis
- Dural metastases
- Venous engorgement (CSF leak)

PATHOLOGY

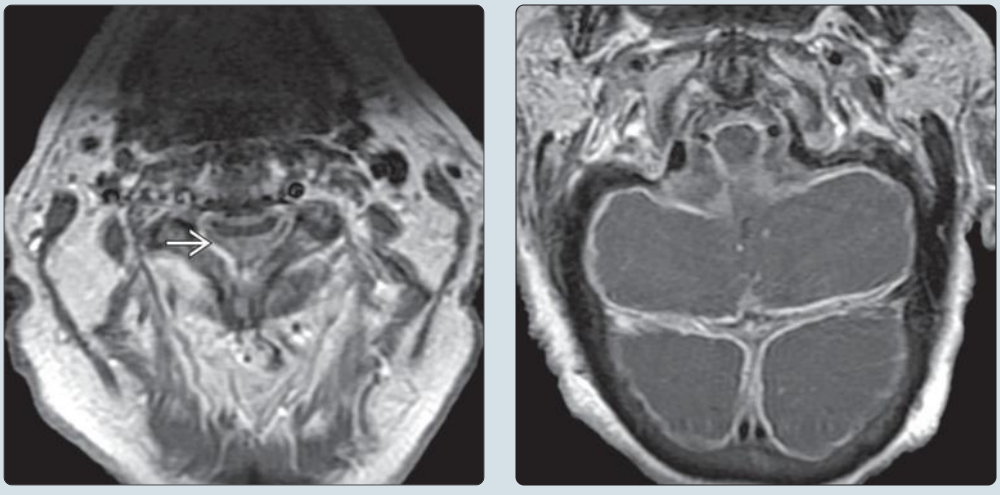
- Autoreactive IgG4 antibodies observed in IgG4-related disease
  - No evidence they are directly pathogenic
- **Central pathology features**
  - Lymphoplasmacytic infiltration
  - Obliterative phlebitis
  - Storiform fibrosis
  - Mild to moderate tissue eosinophilia

CLINICAL ISSUES

- Spinal presentation as mass with myelopathy or radiculopathy
- Histopathology remains cornerstone of diagnosis



(Left) *Sagittal T2WI MR shows extensive low signal intensity thickening of the dura involving posterior fossa and extending to involve upper cervical dura to the C4 level. There is severe cord compression with loss of normal CSF signal from the C1-C3 level.* **(Right)** *Sagittal T1WI C+ MR shows diffuse enhancement of dural thickening of posterior fossa and upper cervical dura. Note the variable enhancement with areas that homogeneously enhance and other areas with more peripheral enhancement.*



(Left) *Axial T1WI C+ MR shows diffuse enhancement of the markedly thickened dura circumferentially surrounding the cord.* **(Right)** *Axial T1WI C+ MR shows the extension of the diffuse dural enhancement and thickening to involve the posterior fossa and tentorium. Note the lack of direct brain parenchymal involvement.*

TERMINOLOGY

Synonyms

- Idiopathic hypertrophic pachymeningitis (IHP)
- IgG4-related disease
- IgG4-related hypertrophic pachymeningitis (IgG4-RHP)

Definitions

- Multiorgan immune mediated condition, which mimics multiple malignant, infectious, and inflammatory disorders
  - 1st defined for pancreatic involvement in 2003
  - May involve pachymeninges of CNS
    - Chronic progressive diffuse inflammatory fibrosis of dura of brain or spine

IMAGING

General Features

- Best diagnostic clue
  - Diffuse smooth dural thickening with low T2 signal
  - Does not involve brain parenchyma or spinal cord directly
- Location
  - Any meningeal location, including skull base and spine
- Morphology
  - Linear thickening of dura

MR Findings

- T1WI
  - Linear low signal mass involving dura with variable mass effect upon cord
- T2WI
  - Low signal linear mass effacing CSF and cord
- T1WI C+
  - Homogeneous or marked peripheral enhancement of thickened dural mass
  - Peripheral enhancement related to peripheral zone of active inflammation vs. chronic central fibrosis

Imaging Recommendations

- Protocol advice
  - T1WI C+ best shows dural abnormality, T2WI shows low signal mass

DIFFERENTIAL DIAGNOSIS

Meningioma

- Focal dura lesion with homogeneous enhancement

Lymphoma

- Homogeneous enhancement, look for leptomeningeal disease
- Tends to show iso to low signal on T2W imaging, and increased diffusion signal due to high cell density

Tuberculosis

- Multifocal disease
- Look for intracranial parenchymal lesions
- Parenchymal involvement suggests diagnosis other than IgG4-related disease

Sarcoidosis

- Multifocal disease
- Intracranial lesions, leptomeningeal enhancement

Dural Metastases

- Not low T2 signal
- Does not show peripheral enhancement that IHP may

Venous Engorgement

- Typically from CSF leak
- Look for intracranial hypotension signs

PATHOLOGY

General Features

- Etiology
  - Unknown; autoimmune phenomenon
  - Autoreactive IgG4 antibodies observed in IgG4-related disease
    - No evidence they are directly pathogenic
    - Exact role IgG4 plays in disease process has yet to be determined
  - Inflammatory and fibrotic process appears driven by Th2 cells, or combination of Th2 cells and regulatory T cells (Treg cells)
- Genetics
  - Limited genetic studies available
    - Major histocompatibility (MHC) class II bias in IgG4-related disease in patients of Asian ancestry
    - Potential genetic links to HLA genes and FcRL3, CTLA4, KCNA3 loci
- Associated abnormalities
  - Riedel thyroiditis
    - Fibrosing Hashimoto thyroiditis may also be in spectrum of IgG4-related disease
  - Retroperitoneal fibrosis (Ormond disease)
  - Periaortitis and periarteritis
    - Chronic periaortitis three components
      - IgG4-related retroperitoneal fibrosis
      - IgG4-related abdominal aortitis
      - IgG4-related perianeurysmal fibrosis
  - Inflammatory pseudotumor
  - Mikulicz disease
    - Salivary and lacrimal glands
    - Unilateral or bilateral diffuse or focal swelling
  - Küttner tumor
    - Submandibular gland
    - Firm swelling difficult to differentiate from a neoplasm
  - Inflammatory aortic aneurysm
  - Multifocal fibrosclerosis
    - Orbits, thyroid, retroperitoneum, mediastinum, other organs
  - Fibrosing mediastinitis
  - Sclerosing mesenteritis
  - Kidney
    - Tubulointerstitial nephritis (most common renal manifestation)
    - Membranous glomerulonephritis
  - Autoimmune (sclerosing) pancreatitis

Staging, Grading, & Classification

- Absolute number of IgG4-positive plasma cells in tissue samples must be interpreted according to specific tissue

- Sialadenitis cutoff value should be at least 100 cells per high-power field
- Pancreas more than 50 cells per high-power field is compatible with diagnosis of autoimmune pancreatitis
- Ratio of IgG4 to IgG-positive plasma cells must be at least 40%
- IgG4-related disease cannot be diagnosed on basis of infiltration by IgG4-positive cells alone

Microscopic Features

- **Central pathology features**
  - Lymphoplasmacytic infiltration
  - Obliterative phlebitis
  - Storiform fibrosis
  - Mild to moderate tissue eosinophilia
- High numbers of IgG4-positive plasma cells
  - Lymphocytes and plasma cells polyclonal
- Necrosis, discrete granulomata, and xanthogranulomatous changes atypical
- Storiform fibrosis characterized by radially arranged collagen fibers that weave through tissue
  - Unique pattern associated with IgG4-related disease
- Phlebitis is partial or complete obliteration of medium-sized veins
  - Lumina small and medium-sized veins are partially or completely occluded by inflammatory infiltrate
  - Infiltrate composed of lymphocytes and plasma cells
- Infiltrate shows abundance of CD4(+) T cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - **Most common forms**
    - Autoimmune pancreatitis
    - Sialadenitis (particularly of the submandibular gland)
    - Dacryoadenitis
    - Retroperitoneal fibrosis
  - Brain involvement
    - Headache
    - Cranial nerve palsies
    - Ataxia
    - Seizures
    - Increased intracranial pressure
  - Spinal presentation
    - Radiculopathy
    - Myelopathy
  - IgG4 serum concentrations elevated, but range varies widely
    - Increased serum IgG4 concentrations neither sufficiently sensitive nor specific for diagnosis
      - Serum IgG4 concentrations useful for screening but unreliable as single diagnostic marker
  - Tissue biopsy is gold standard for diagnosis

Demographics

- Age
  - Peak in 6th decade
  - M > F, 3.5:1
    - Involvement of H&N M:F closer to 1:1
    - Male predilection in striking contrast to classic autoimmune diseases, which have marked female preponderance

Treatment

- Steroids
  - Most clinical manifestations of IgG4-related disease respond to glucocorticoids
- Immune modulators/suppressants in steroid resistant cases
  - Azathioprine
  - Mycophenolate mofetil
  - Methotrexate
  - Rituximab is promising therapeutic approach
    - Works by B-cell depletion
- Surgery for decompression

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Diagnosis of exclusion
- **Tissue biopsy is gold standard for diagnosis**

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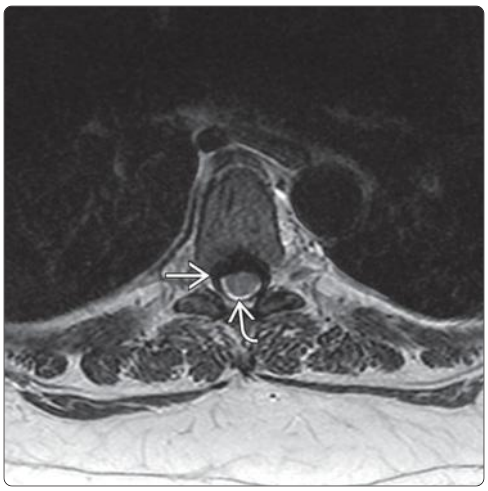


(Left) *Sagittal T2W MR shows large linear areas of very low signal involving the dura of the spine diffusely, with marked attenuation of CSF signal. Cord compression with slight cord edema is present.* **(Right)** *Sagittal T1W C+ MR shows smooth linear enhancement outlining the margins of the thickened fibrotic low signal dura throughout the spine.*





(Left) *Sagittal T2W MR shows extensive linear low signal involving the dura throughout the lumbar spine, both the ventral and dorsal aspects. Patient has undergone dural biopsy.* **(Right)** *Sagittal T1W C+ MR demonstrates linear enhancement along the dural/CSF interface of the diffusely thickened fibrotic dura throughout the lumbar spine, sparing only the most inferior L5 level and sacrum.*



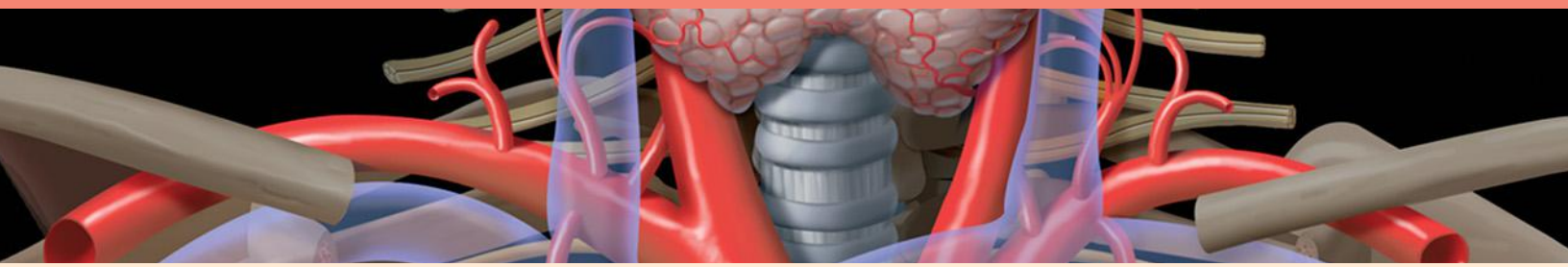


(Left) *Axial T2W MR through the thoracic spine shows the circumferential nature of the dural thickening, showing very low signal intensity. There is marked narrowing of the subarachnoid space with very little visible CSF signal.* **(Right)** *Axial T2W MR shows a thick rind of low signal intensity thickened dura surrounding the nerve roots, with little intervening CSF signal.*

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PART V
SECTION 3

Vascular and Systemic Disorders



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Terminology

Abbreviations: Anterior spinal artery (ASA), posterior spinal artery (PSA), artery of Adamkiewicz (AA)

Synonyms: Great anterior radicular artery, arteria radicularis magna = artery of Adamkiewicz radicularis magna = artery of Adamkiewicz

Imaging Anatomy

Vertebral Artery

The vertebra artery is divided into 4 segments. The **first segment** (V1) of the vertebral artery extends from its origin to the point of entrance into the foramen of the cervical transverse process, which is usually the sixth body. The vertebral artery is usually the most proximal and largest branch off of the subclavian artery. Multiple variations in the anatomic course and origins of the vertebral arteries have been described. The most common variation in vertebral artery origin is in the origination from the proximal subclavian artery. The origin of the left vertebral artery from the aortic arch between the left common carotid artery and left subclavian artery has been described in 2.4-5.8% of cases. When there is an origin of the vertebral artery from the arch, the vertebral artery usually enters the foramen of the transverse process of the fifth cervical vertebrae. With a normal origin of the left vertebral artery from the subclavian artery, the vertebral artery enters the transverse foramen of the sixth cervical vertebrae in nearly 88% of cases. The site of entrance at the level of C4 is seen in 0.5%, C5 in 6.6%, and C7 in 5.4%. Rare examples of origins of the left vertebral artery from the left common carotid artery, or external carotid artery, have been described. Also rare are variations in the origin of the right vertebral artery (less than 1%) from the aorta, carotid arteries, or brachiocephalic arteries.

The **second segment** (V2) of the vertebral artery extends superiorly through the foramen of the transverse processes in a vertical course until it reaches the transverse process of C2. The **third segment** (V3) of the vertebral artery extends from the exit of C2 to its entrance into the spinal canal. After leaving the transverse foramen of C2, it courses laterally and posteriorly to pass through the transverse foramen of C1. The vertebral artery then extends posterior and medially in a horizontal groove on the upper surface of the posterior arch of C1. The vertebral artery turns abruptly as it nears the midline and pierces the posterior atlantooccipital membrane and enters into the vertebral canal. Anomalous connections in this region are uncommon but include the proatlantal intersegmental artery, which can communicate between the internal or external carotid artery and the vertebral artery at this level. Local duplication or fenestration of the V3 segment can occur. The occipital artery also can arise from the V3 segment. A persistent first intersegmental artery can occur where the vertebral artery courses below the C1 arch after exiting the transverse foramen of C2 and enters the spinal canal without passing through the C1 transverse foramen (3-4%). The origin of the posterior inferior cerebellar artery

The **fourth segment** (V4) segment pierces the dura and extends through the foramen magnum where it lies anterior to the medulla and eventually joins the contralateral vertebral artery to form the basilar artery. Major branches arising off the vertebral artery include multiple muscular branches from the extracranial segments to supply the deep muscles of the neck and meningeal branches. The posterior meningeal branch arises from the vertebral artery above the level of C1, and below the foramen magnum, and supplies the falx cerebelli and the medial portion of the dura of the occipital posterior fossa. Just before joining to form the basilar artery, each of the vertebral arteries give off a branch that will become the anterior spinal artery, which extend downward and medially to join in the midline with a corresponding branch from the other vertebral artery. The posterior spinal arteries can originate from the posterior inferior cerebellar arteries or from the intracranial portion of the vertebral arteries.

Spinal Arteries

The spinal cord circulation is derived from segmental branches off of the vertebral arteries, as well as multiple radicular arteries arising from segmental vessels. These segmental vessels include the descending cervical, deep cervical, intercostal, lumbar, and sacral arteries. The anterior spinal artery arises at the junction of the intradural segment of the vertebral arteries, caudal to the basilar artery. The anterior spinal artery descends in the midline without interruption from the foramen magnum to the filum terminale. The anterior spinal artery supply is reinforced by multiple segmental feeders. The segmental feeders give rise to sulcal, or central branches, which supply the anterior 2/3 of the cord. The anterior 2/3 includes the anterior horns, spinothalamic tracts, and corticospinal tracts. The anterior spinal artery lies in the midline on a ventral aspect of the cord, in the groove of the anterior median fissure. The posterior spinal arteries arise from the posterior rami of the vertebral artery or from the posterior inferior cerebellar artery.

The posterior spinal arteries are a paired longitudinal system of vessels on the dorsal cord medial to the dorsal roots. These form a plexiform and variable network between the two dorsal arteries. The posterior spinal artery supplies the posterior 1/3 of the cord, which includes supply to the posterior columns, and a variable supply to the corticospinal tracts. Continuation of both the anterior and posterior spinal artery supply is derived from segmental anastomoses. These segmental vessels arise as dorsal rami from vertebral, subclavian, thoracic intercostal, and lumbar intercostal arteries. The dorsal rami enter the canal through the neural foramen and then penetrate the dura and divide into two main branches: The dural artery, which supplies the nerve root sleeve and the dura, and the radiculomedullary branch. The radiculomedullary branch then divides into a radicular artery, which penetrates the subarachnoid space to supply the anterior and posterior roots. There is also a variable medullary artery branch, which joins the anterior spinal artery and posterior spinal artery. The radicular arteries arise from the division of the radiculomedullary arteries along the anterior and posterior nerve roots. The anterior radicular artery extends along the anterior surface of the spinal cord, while the posterior radicular artery likewise extends along the posterior cord surface.

The multiple fetal segmental vessels will regress with adulthood, leaving 2-14 (average 6) anterior radicular arteries persisting in the adult, and 11-16 posterior radicular arteries in the adult. The major cervical radicular feeders to the spinal cord occur between the C5-C7 levels. There are 2-3 anterior cervical cord feeders that measure 400-600 microns in size. There are also 3-4 posterior cervical cord feeders, which are smaller in size, on the order of 150-400 microns. The V3 segment never gives rise to radiculomedullary branches. In the thoracic spine, there are 2-3 anterior thoracic cord feeding segmental vessels. These are usually left-sided and are on the

order of 550-1200 microns in size. Small ventral feeding vessels may also be present on the order of 200 microns. There is an inverse relationship between the number and caliber of ventral radicular vessels. There can occur "pauci-segmental" anatomy with fewer vessels (less than 5) with larger caliber or "pluri-segmental" anatomy with more vessels with smaller caliber. The dominant thoracic anterior radicular artery is also called the artery of **Adamkiewicz**. The artery of Adamkiewicz tends to have a left-sided origin (73%) and arises from T9-T12 (62%), with less common origins in the lumbar region (26%) and from T6-T8 (12%). A major segmental feeder may also occur in the upper thoracic spine, often the T5 level. This has been termed the artery of **von Haller**. The number of posterior thoracic cord feeding vessels varies from 9-12, with an average of 8. The posterior thoracic feeding vessels have no right to left lateralization. These vessels are on the order of 150-400 microns in size. The lumbosacral and pelvic regions have from 0-1 major feeding vessels to the spinal cord. The anterior spinal artery ends at the conus with communicating branches (rami cruciantes) to the posterior spinal artery. The posterior division of the iliac artery gives rise to the inferior and superior lateral sacral branches, which give rise to the spinal arteries via the anterior sacral foramen. The anterior division of the iliac artery gives rise to the inferior gluteal artery, which supplies the sciatic nerve. The posterior division of the internal iliac artery gives rise to the iliolumbar artery, which supplies the femoral nerve at the iliac wing level.

Spinal Veins

The veins of the spinal cord parallel the spinal arterial pattern. There is a very symmetrical pattern of venous drainage (compared with the highly asymmetrical arterial supply) with minimal anterior to posterior, right to left segmental variation. There are two sets of intrinsic radial draining veins, which drain into the anastomoses on the cord surface. The central group of veins provides for return of the anterior horns and surrounding white matter and drain into the central veins in the anterior median fissure. This forms the anterior median vein. Peripheral dorsal and lateral cord drainage is via small valveless radial vein plexus, which extends to the coronal venous plexus on the cord surface, and then drains to the epidural venous plexus of Batson. The epidural plexus consists of anterior and posterior internal vertebral plexus components and connects with the superior and inferior vena cava, azygos and hemiazygos systems, and the intracranial dural sinuses. There are from 30-70 medullary radicular veins. The anterior median vein continues caudally along the filum terminale to the end of the dural sac. The coronal and median veins drain to the medullary veins, which leave the intradural space at the root sleeve and extend into the epidural plexus. Medullary veins have a functional valve-like mechanism at the dural margin, which prevents epidural reflux into the intradural space. There are no intradural valves present.

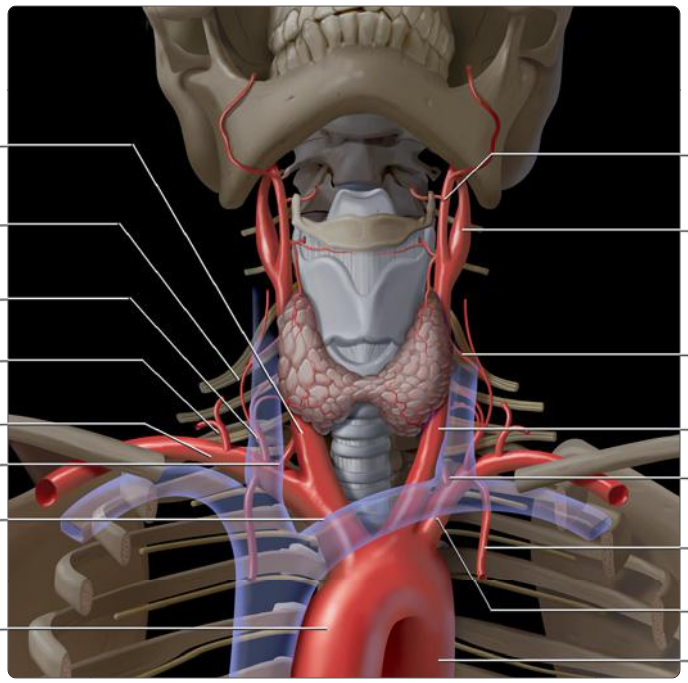
Embryology

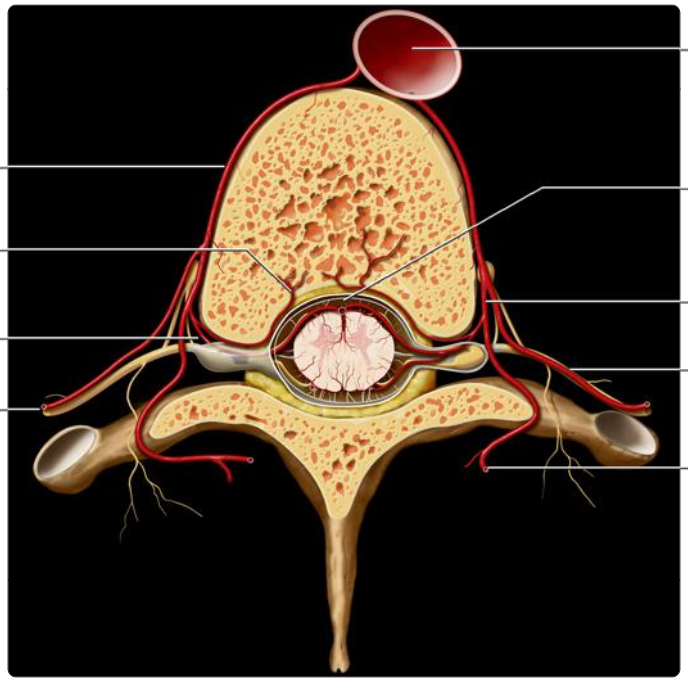
The embryogenesis of the vertebral artery begins at approximately day 32 and is completed by day 40. The vertebral artery is formed from fusion of the longitudinal anastomosis that link cervical intersegmental arteries, which branch off primitive paired dorsal aorta. The intersegmental arteries regress, except for the seventh vessel, which will come to form the proximal portion of the subclavian artery, including the origin of the vertebral artery. As the connections to the primitive dorsal aorta disappear, the vertebral artery takes shape and initially has a more beaded anastomotic appearance and a tortuous course. The basilar artery is formed by fusion of the two primitive vertebral arteries.

Spinal cord vessels originate from a capillary network on the ventral lateral surface of the cord connected with segmental aortic branches. Two primitive longitudinal systems are formed. By the end of the second month, the ventrolateral systems transform into longitudinal solitary anterior median anterior spinal artery. The plexus-like pattern remains more prominent on the dorsal surface of the cord. The anterior spinal artery formation is followed by a variable regression of segmental feeding vessels (initially 31) and is completed by the fourth month of gestation. The reduction is most pronounced in the thoracic and lumbar areas. Segmental arteries persist as intercostal and lumbar arteries. In the cervical spine, dorsal intersegmental anastomoses persist as components of the vertebral arteries. The ventral anastomoses persist as the thyrocervical trunk.

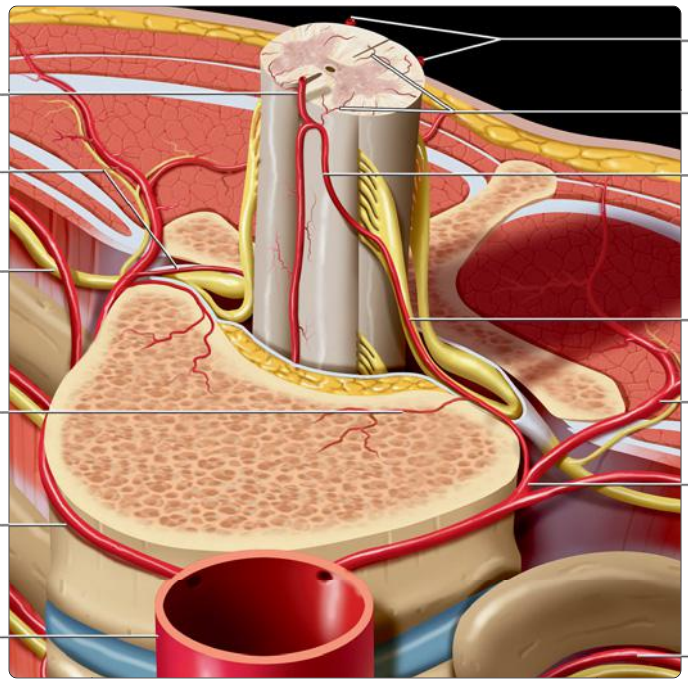
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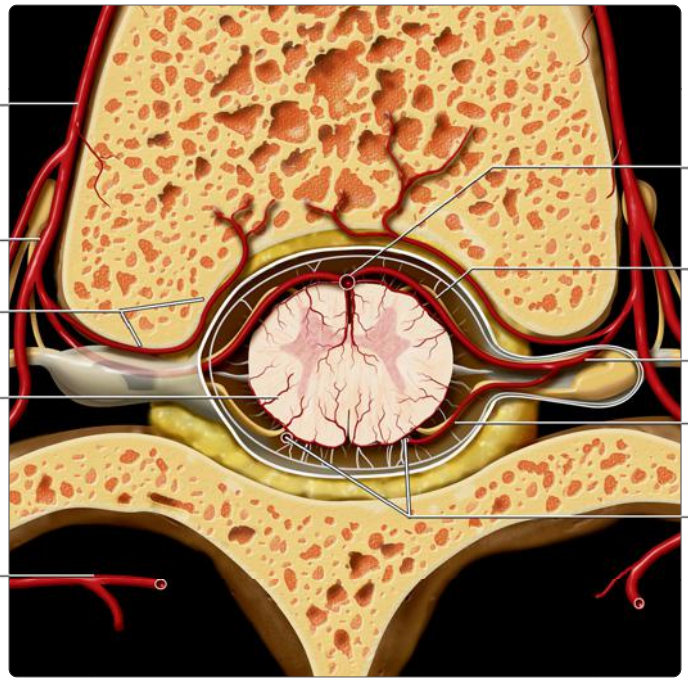
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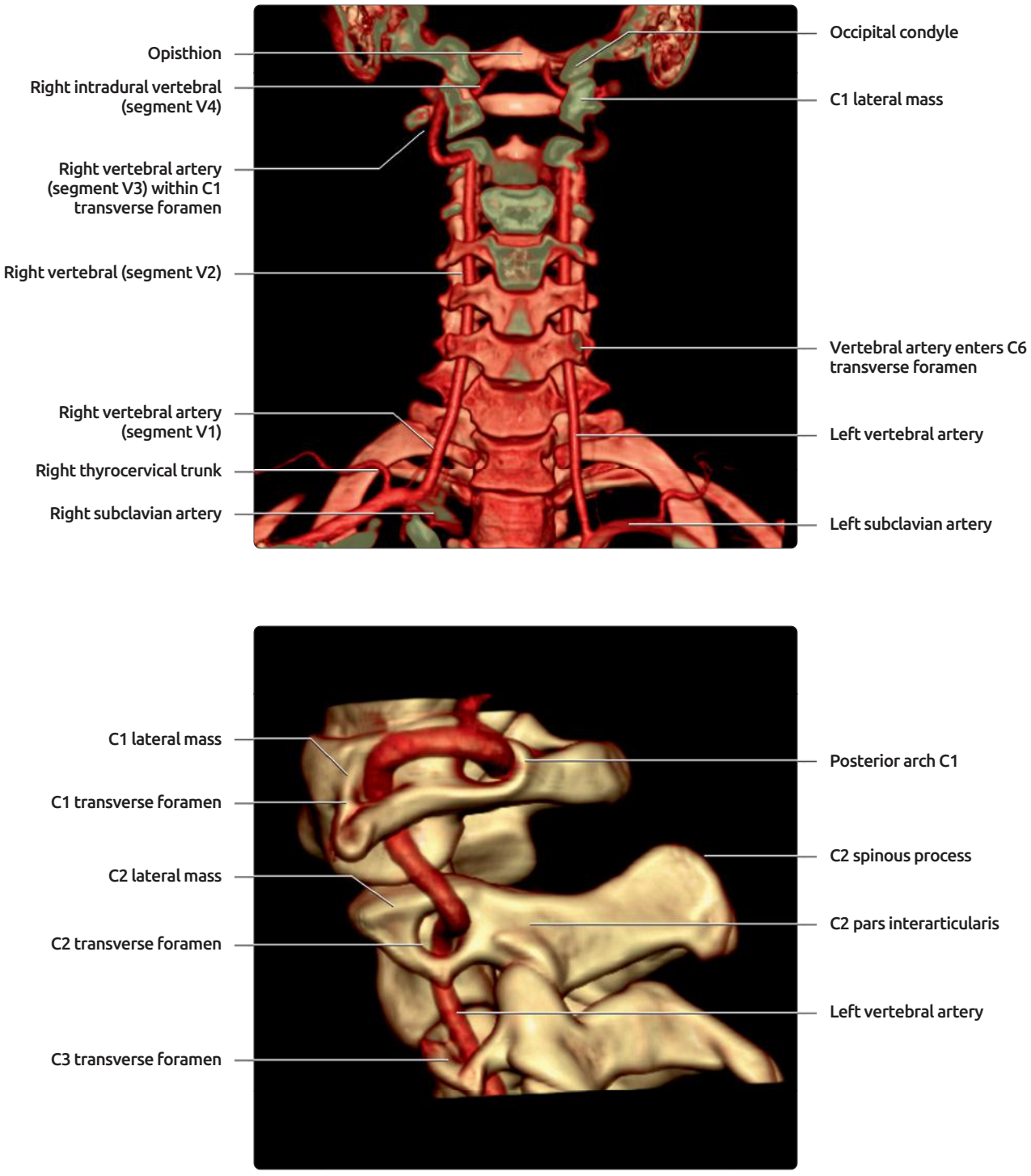


(Top) *AP graphic shows the aortic arch and arterial great vessels in red. The vertebral arteries give rise to the anterior and posterior spinal arteries. The ascending cervical arteries, branches of the thyrocervical trunks, give off anterior and posterior segmental medullary arteries that anastomose with the ASA and PSA on the cord surface. Complete spinal angiography includes evaluation of all these vessels.* **(Bottom)** *Axial graphic shows overview of the arterial supply to the vertebral column and its contents, depicted here for the lower thoracic spine. A series of paired segmental arteries (cervical region arise from the vertebral and thyrocervical arteries, thoracic region are intercostal arteries, and lumbar region are lumbar arteries) divide into anterior and posterior branches. The posterior branch gives rise to a muscular branch, a branch to the vertebral body, and the radiculomedullary artery. The radiculomedullary artery enters the vertebral canal via neural foramen.*

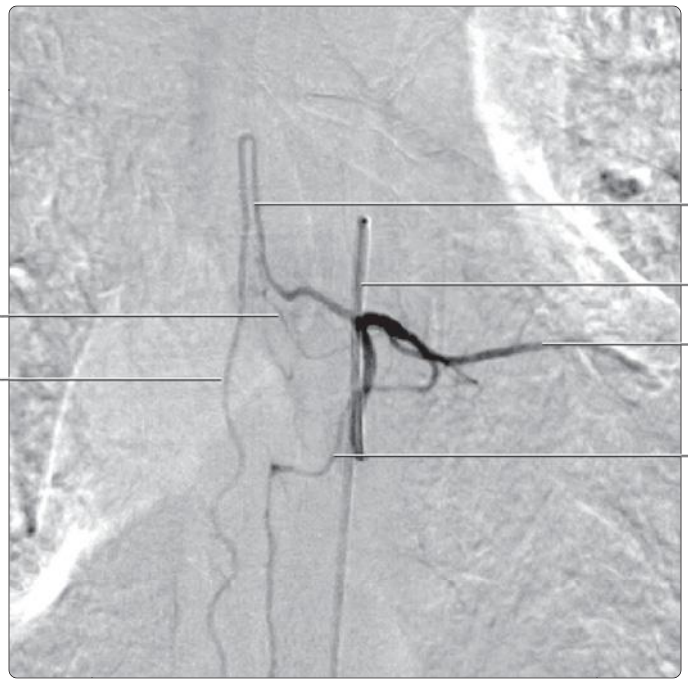


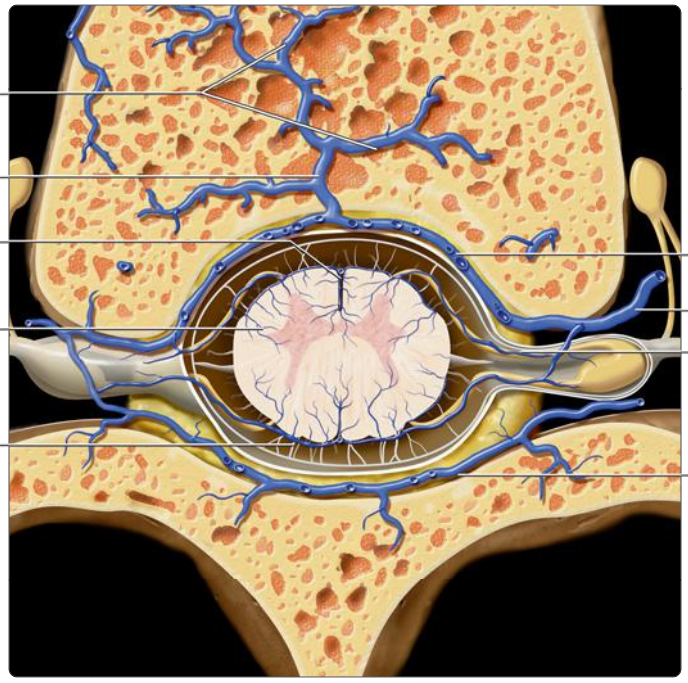


(Top) *Oblique axial graphic rendering of T10 depicts segmental intercostal arteries arising from the lower thoracic aorta. The artery of Adamkiewicz is the dominant segmental feeding vessel to the thoracic cord, supplying the anterior aspect of the cord via the anterior spinal artery. Note its characteristic "hairpin" turn on the cord surface as it first courses superiorly, then turns inferiorly.* **(Bottom)** *Anterior and posterior radiculomedullary arteries anastomose with the anterior and posterior spinal arteries. Penetrating medullary arteries in the cord are largely end-arteries with few collaterals. The cord "watershed" zone is at the central gray matter.*



(Top) *AP volume rendered image of CTA shows the course of the vertebral arteries, entering the transverse foramen and ascending to the foramen magnum. Both vertebral arteries in this patient enter the C6 level, but this can show wide normal variation. Left vertebral arteries arising from the arch enter more cephalad, at C5.* **(Bottom)** *Lateral volume rendered CTA shows the course of the distal left vertebral artery passing through C1 and horizontally oriented C2 transverse foramen.*





(Top) *AP view of a left T8 intercostal injection gives rise to the characteristic hairpin turn of the major segmental feeding vessel to the thoracic cord, the artery of Adamkiewicz. Extending inferiorly from the top of the hairpin turn is the anterior spinal artery, which supplies the anterior 2/3 of the cord.* **(Bottom)** *Magnified graphic of the internal vertebral venous plexus. The radicular veins course along the dorsal and ventral rami, eventually draining into components of the anterior or posterior internal plexus, and subsequently the segmental veins, which will drain into the superior or inferior vena cava.*

Persistent First Intersegmental Artery

KEY FACTS

TERMINOLOGY

- Anatomic variation of V3 segment of vertebral artery (VA) coursing into spinal canal below C1 arch level, at C1-C2
- Unilateral in up to 4% of individuals

IMAGING

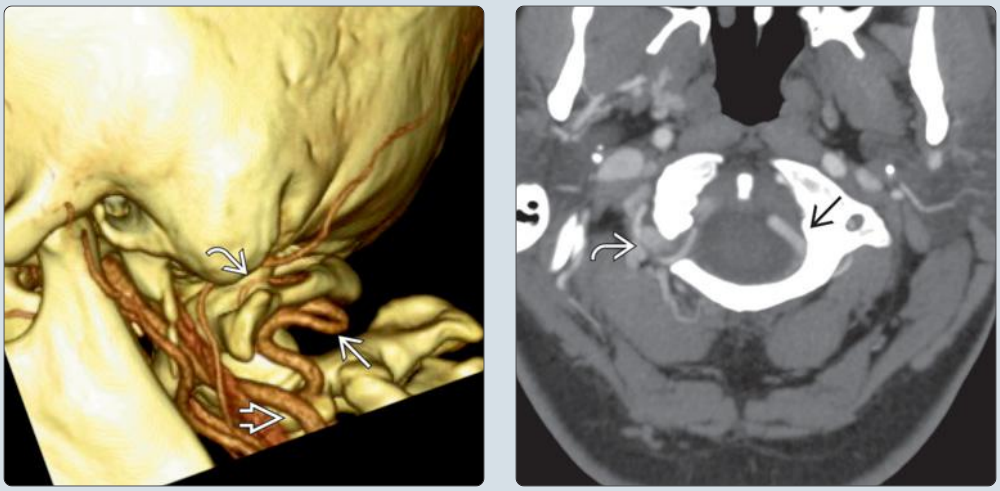
- Rounded vascular structure medial to C1 arch
  - May show associated bony groove in medial aspect of posterior arch of C1 at vessel location
- Flow void below C1 arch on parasagittal images
- Flow void medial to C1 arch on axial images

TOP DIFFERENTIAL DIAGNOSES

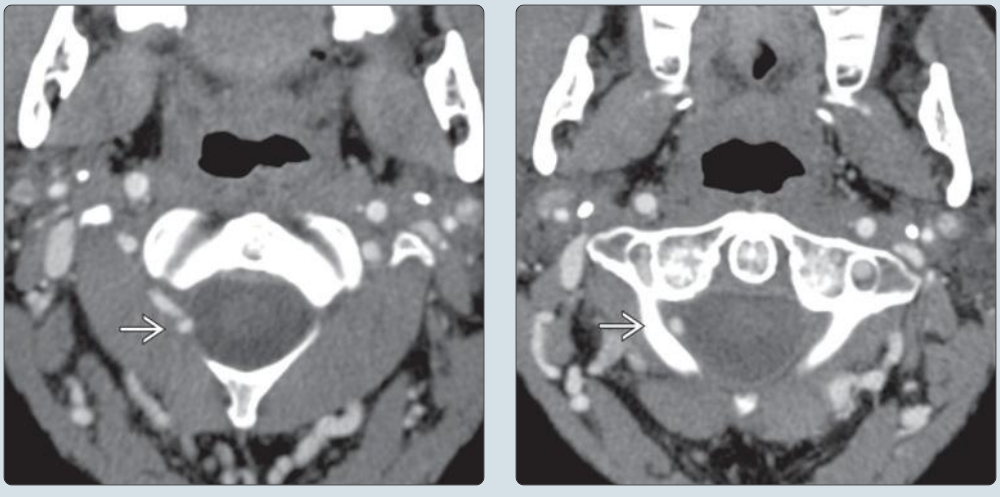
- Proatlantal intersegmental artery
  - Persistent carotid-basilar anastomoses
- Fenestrated vertebral artery
  - Normal V3 course superior to C1, but 2nd segment enters canal at C1-C2
- Low posterior inferior cerebellar artery (PICA) origin
  - Normal V3 course of vertebral superior to C1, but PICA has low origin and enters canal at C1-C2
- Cervicomuscular collateral to vertebral artery

CLINICAL ISSUES

- Usually asymptomatic
- Can present problem for interventional or surgical approaches
  - C1 lateral mass screws may be dangerous with this variant with risk of vascular injury, and C1-C2 transarticular screws are preferred
  - Increased risk of vascular injury with C1-C2 lateral puncture for myelography



(Left) *Left lateral 3D reformat of a CTA shows the course of the left vertebral artery extending below the arch of C1 as a persistent left 1st intersegmental artery. There is a hypoplastic normal course of the left vertebral artery through the C1 foramen and over the top of the C1 arch.* **(Right)** *Axial CTA shows the position of the left persistent 1st intersegmental artery traveling cephalad from below the C1 arch. The right vertebral artery has a normal position superior to the C1 arch.*



(Left) *Axial CTA study shows a persistent 1st intersegmental artery variant of the right vertebral artery entering the spinal canal below the right side of the C1 arch in the C1-C2 interspace.* **(Right)** *Axial CTA study shows the typical appearance of a 1st intersegmental artery variant of the right vertebral artery extending into the spinal canal below the C1 arch level and positioned medial to the C1 arch as it extends cephalad.*

TERMINOLOGY

Definitions

- Anatomic variation of V3 segment of vertebral artery (VA) coursing into spinal canal below C1 arch level, at C1-C2
- Unilateral in up to 4% of individuals
- Bilateral persistence is uncommon (< 1%)

IMAGING

General Features

- Best diagnostic clue
  - VA travels between atlas and axis in space normally occupied only by C2 nerve root
  - Seen on axial images as vessel medial to lateral aspect of C1 arch
- Location
  - C1-C2 level, inferior to C1 arch
- Size
  - Variable, like normal VA

CT Findings

- CTA
  - Rounded vascular structure medial to C1 arch
  - May show associated bony groove in medial aspect of posterior arch of C1 at vessel location
  - Look for marked asymmetry of transverse foramen size as general indicator of VA variation

MR Findings

- T2WI
  - Flow void is below C1 arch on parasagittal images
  - Flow void is medial to C1 arch on axial images

DIFFERENTIAL DIAGNOSIS

Proatlantal Intersegmental Artery

- Persistent carotid-basilar anastomoses
- Type 2 persistent proatlantal intersegmental artery originates from external carotid artery and joins the normal course of vertebral artery at C1-C2 interspace

Fenestrated Vertebral Artery

- Normal V3 course superior to C1, but 2nd segment enters canal at C1-C2

Low Posterior Inferior Cerebellar Artery (PICA) Origin

- Normal V3 course of vertebral superior to C1, but PICA has low origin and enters canal at C1-C2

Cervicomuscular Collateral to Vertebral Artery

- Collateral flow to distal vertebral if inferior vertebral artery is occluded
- At C1-C2 level, likely originating from external carotid (occipital artery)

PATHOLOGY

General Features

- Etiology
  - Persistence of cervical 1st intersegmental artery during embryogenesis
  - Failure of resegmentation of embryonic sclerotome

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
  - Can present problem for interventional or surgical approaches
    - C1 lateral mass screws may be dangerous with this variant with risk of vascular injury, and C1-C2 transarticular screws are preferred
    - Injuries include
      - Formation of arteriovenous fistula
      - Occlusion
      - Dissection of VA
      - Massive bleeding
    - Increased risk of vascular injury with C1-C2 lateral puncture for myelography
  - Rare cases of cord compression reported due to bilateral persistent 1st intersegmental arteries
  - Rare reports related to such vascular anomaly of
    - Cervical pain
    - Occipital neuralgia
    - Accessory nerve palsy

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Persistent Hypoglossal Artery

KEY FACTS

TERMINOLOGY

- Persistent fetal carotid-basilar anastomosis with course from internal carotid artery through hypoglossal canal to basilar artery

TOP DIFFERENTIAL DIAGNOSES

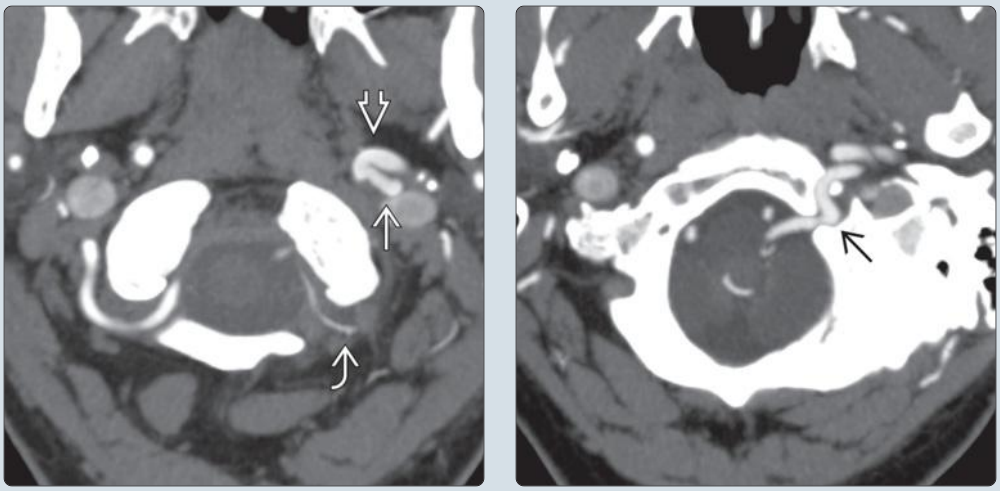
- Proatlantal intersegmental artery
  - Type 1 artery arises from caudal internal carotid artery and courses between arch of C1 and occiput to join distal ipsilateral vertebral artery
  - Type 2 artery arises from external carotid artery and travels within C1–C2 interspace to join ipsilateral vertebral artery

PATHOLOGY

- 4 potential carotid-basilar anastomoses have been described
- **Trigeminal** (0.1-0.5%)
- **Otic** (very rare, ≤ 0.001%)
- **Hypoglossal** (0.1%)
  - Primitive hypoglossal artery originates from cervical part of internal carotid artery at C1-C3 level
  - Proximal vertebral arteries usually hypoplastic
- **Proatlantal intersegmental** (0.02%)

CLINICAL ISSUES

- Typically asymptomatic
- Scattered case reports of mixed types of pathology related to this anomaly
  - Aneurysm (probably not greater rate than general population)
  - Atherosclerotic stenosis of persistent hypoglossal
  - Dysfunction of hypoglossal nerve and glossopharyngeal neuralgia



(Left) *Axial CTA shows the left internal carotid artery giving rise to the anomalous hypoglossal artery. Note the associated small distal left vertebral artery.* **(Right)** *Axial CTA at the level of the left hypoglossal canal shows the persistent hypoglossal artery passing into the posterior fossa through the enlarged left hypoglossal canal and reconstituting the vertebrobasilar junction.*



(Left) *A series of axial images from an MRA show a large artery arising from the left internal carotid artery above the carotid bifurcation and passing dorsally through the hypoglossal canal to form the basilar artery.* **(Right)** *3D MRA reconstruction shows the aberrant course of the vessel as it forms the basilar artery.*

TERMINOLOGY

Synonyms

- Primitive hypoglossal artery, fetal hypoglossal artery

Definitions

- Persistent fetal carotid-basilar anastomosis with course from internal carotid artery through hypoglossal canal to basilar artery

IMAGING

General Features

- Best diagnostic clue
  - Vessel within hypoglossal canal originating from distal cervical internal carotid artery and supplying basilar artery
- Location
  - Hypoglossal canal
- Size
  - Variable; may approximate size of distal internal carotid artery

CT Findings

- CTA
  - Tubular vascular structure extending through hypoglossal canal and forming basilar artery

MR Findings

- T1WI
  - Tubular flow void extending through enlarged hypoglossal canal

Imaging Recommendations

- Best imaging tool
  - CTA/MRA

DIFFERENTIAL DIAGNOSIS

Proatlantal Intersegmental Artery

- Type 1 artery arises from caudal internal carotid artery and courses between arch of C1 and occiput to join distal ipsilateral vertebral artery
- Type 2 artery arises from external carotid artery and travels within C1–C2 interspace to join ipsilateral vertebral artery

PATHOLOGY

General Features

- Etiology
  - 4 potential carotid-basilar anastomoses have been described
    - **Trigeminal** (0.1-0.5%)
    - **Otic** (very rare, ≤ 0.001%)
    - **Hypoglossal** (0.1%)
      - 1st described by Batujeff in 1889
      - Primitive hypoglossal artery originates from cervical part of internal carotid artery at C1-C3 level
      - After tortuous course, it penetrates enlarged hypoglossal canal and joins lower portion of basilar artery
      - Proximal vertebral arteries usually hypoplastic
      - Ipsilateral vertebral artery may be absent
      - Rare variant where hypoglossal arises from external carotid
    - **Proatlantal intersegmental** (0.02%)
  - These anastomoses remain functional for 7-10 days during early stage of fetal development
    - Obliterate at rate posterior communicating arteries develop
    - Failure to obliterate results in persistence of embryonic artery

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Typically asymptomatic
  - Scattered case reports of mixed types of pathology related to this anomaly
  - Aneurysm (probably not greater rate than general population)
  - Carotid atherosclerosis with embolization to posterior circulation
  - Atherosclerotic stenosis of persistent hypoglossal
  - Dysfunction of hypoglossal nerve and glossopharyngeal neuralgia

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Persistent Proatlantal Artery

KEY FACTS

TERMINOLOGY

- Persistent fetal carotid-basilar anastomosis with course from internal carotid artery (ICA) or external carotid artery (ECA) to basilar artery at C1 level

IMAGING

- Anomalous vessel connecting ECA or ICA to basilar system through foramen magnum, with horizontal course below occiput

TOP DIFFERENTIAL DIAGNOSES

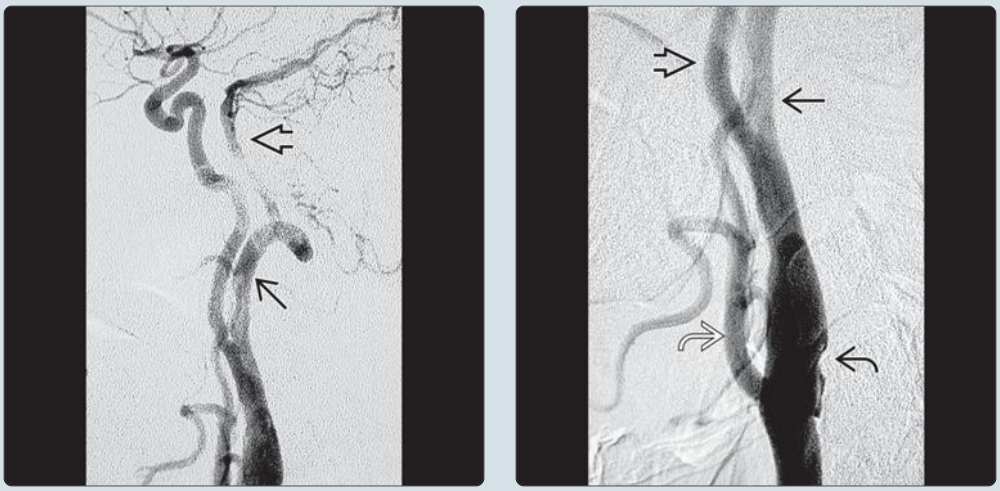
- Persistent hypoglossal artery
  - Fetal carotid-basilar anastomosis with course from ICA through hypoglossal canal to basilar artery
- Persistent 1st intersegmental artery
  - Vertebral artery travels between atlas and axis in space normally occupied only by C2 nerve root
- Fenestrated vertebral artery
  - Normal course of V3 superior to C1, with 2nd segment entering canal at C1-C2

PATHOLOGY

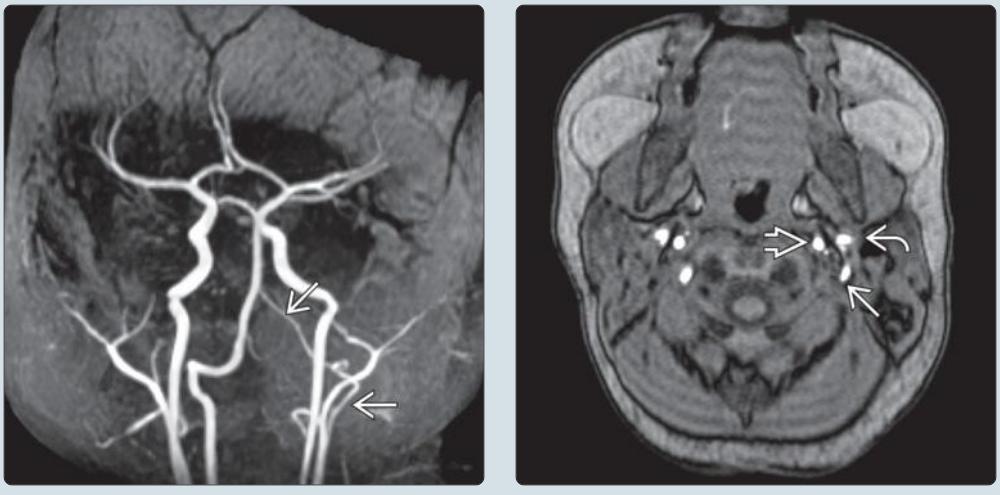
- Type 1 proatlantal artery arises from ICA and ascends to C0-C1 space without passing through transverse foramen (~ 50%)
- Type 2 proatlantal artery (C1 intersegmental variety) arises from ECA

CLINICAL ISSUES

- Usually asymptomatic
- Can present problem for interventional or surgical approaches
- Rare reports of associated embolic infarcts to posterior circulation from anterior circulation (carotid bifurcation) atherosclerosis in setting of proatlantal intersegmental artery



(Left) *Lateral catheter angiography shows a type 1 proatlantal intersegmental artery ⇨ arising off the ICA and supplying the basilar artery ⇨ via the foramen magnum.* **(Right)** *Lateral catheter angiography of the carotid bifurcation shows an anomalous proatlantal intersegmental artery ⇨ arising off the proximal ICA. Note the course of the normal ICA ⇨ and ECA ⇨. There is atheromatous irregularity of the carotid bulb ⇨, which caused an embolic infarct (not shown) in this individual.*



(Left) *Oblique MRA shows an anomalous type 2 proatlantal intersegmental artery ⇨ arising off the left ECA and supplying the basilar artery. (Courtesy A. Illner, MD.)* **(Right)** *Axial MRA source image shows the positions of the left ECA ⇨, ICA ⇨, and type 2 proatlantal intersegmental ⇨ arteries in the neck. Note the lack of normal left vertebral artery signal within the left transverse foramen. (Courtesy A. Illner, MD.)*

TERMINOLOGY

Synonyms

- Persistent primitive proatlantal intersegmental artery
- Persistent proatlantal artery

Definitions

- Persistent fetal carotid-basilar anastomosis with a course from internal carotid artery (ICA) or external carotid artery (ECA) to basilar artery at C1 level

IMAGING

General Features

- Best diagnostic clue
  - Anomalous vessel connecting ECA or ICA to basilar system through foramen magnum, with horizontal course below occiput
- Location
  - Suboccipital region through foramen magnum
- Size
  - Variable, may approximate size of distal ICAs

CT Findings

- CTA
  - Tubular vascular structure extending from distal cervical ICA or ECA to basilar artery via foramen magnum

MR Findings

- T1WI
  - Tubular flow void extending from anterior circulation to basilar artery

Imaging Recommendations

- Best imaging tool
  - CTA/MRA

DIFFERENTIAL DIAGNOSIS

Persistent Hypoglossal Artery

- Fetal carotid-basilar anastomosis with course from ICA through hypoglossal canal to basilar artery

Persistent 1st Intersegmental Artery

- Vertebral artery travels between atlas and axis in space normally occupied only by C2 nerve root
- No connection to ECA or ICA

Fenestrated Vertebral Artery

- Normal course of V3 superior to C1, with 2nd segment entering canal at C1-C2

Cervicomuscular Collateral to Vertebral Artery

- Collateral flow to distal vertebral if inferior vertebral is occluded

PATHOLOGY

General Features

- Associated abnormalities
  - Aplasia or hypoplasia of 1 or both vertebral arteries proximal to anastomotic vessel in 50% of cases
  - Rare reports of intracranial aneurysms with proatlantal arteries but may reflect incidence equivalent to general population

Staging, Grading, & Classification

- Type 1 proatlantal artery arises from ICA and ascends to C0-C1 space without passing through transverse foramen (~ 50%)
  - Artery takes course dorsal to transverse process of C1 and extends cephalad to enter foramen magnum
- Type 2 proatlantal artery (C1 intersegmental variety) arises from ECA
  - More lateral in position than type 1 variety
  - Joins horizontal segment of vertebral artery before entering foramen magnum

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
  - Can present problem for interventional or surgical approaches
    - C1 lateral mass screws may be dangerous with this variant
  - Rare reports of associated embolic infarcts to posterior circulation from anterior circulation (carotid bifurcation) atherosclerosis in setting of proatlantal intersegmental artery

Demographics

- Epidemiology
  - Rare: < 50 cases reported

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Type 1 Vascular Malformation (dAVF)

KEY FACTS

TERMINOLOGY

- Synonyms: Type 1 spinal AVF, dural AVF, dural fistula
- Definition: Spinal arteriovenous (AV) fistula, present within dura, with intradural distended draining veins

IMAGING

- Cord enlarged, T2WI hyperintense, vessel flow voids on cord surface
- Multiple enhancing serpentine veins on cord surface
- Dynamic contrast-enhanced MRA capable of defining dilated intradural veins; very useful to guide catheter angiography
- Spinal catheter arteriography is gold standard for confirming diagnosis/treatment

TOP DIFFERENTIAL DIAGNOSES

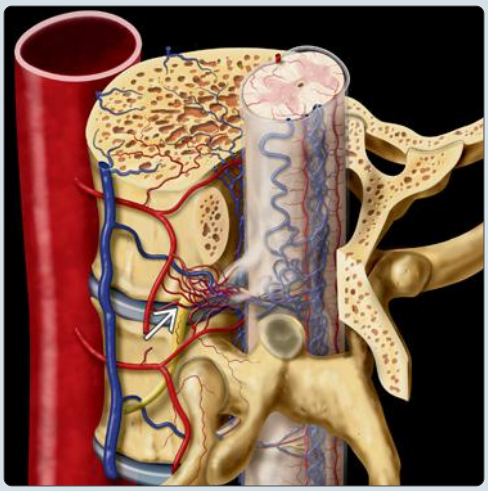
- CSF pulsatile flow artifact
- Spinal cord tumor
- Spinal cord arteriovenous malformation
- Tortuous "redundant" roots from spinal stenosis

PATHOLOGY

- Venous hypertension from engorgement reduces intramedullary AV pressure gradient, causing reduced tissue perfusion and cord ischemia

CLINICAL ISSUES

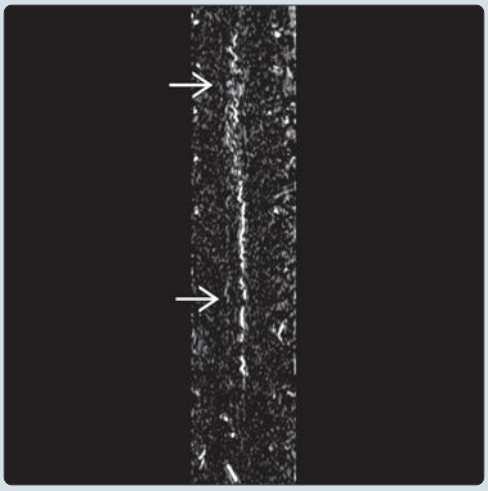
- Most common presentation is progressive lower extremity weakness with both upper + lower motor neuron involvement
- Very rarely presents with subarachnoid hemorrhage
- Middle-aged male with progressive lower extremity weakness exacerbated by exercise
- Persistent edema and enhancement of cord can occur even with successful clinical treatment of fistula





(Left) *Sagittal oblique graphic of the thoracic cord shows site of a type 1 dural fistula at the dural root sleeve level ⇒, with secondary dilatation of intradural venous plexus due to AV shunt.* **(Right)** *Sagittal T2WI MR shows central cord T2 hyperintensity (edema) related to venous hypertension from the peripheral fistula shunting, which typically spares the cord periphery. There are multiple serpentine intradural flow voids from the arterialized and distended venous plexus ⇒.*





(Left) *Sagittal T1WI C+ FS MR shows ill-defined diffuse enhancement of the distal thoracic cord ⇒ in this case of type 1 dural fistula. Faint to moderate cord parenchymal enhancement is common with this lesion.* **(Right)** *Coronal reformat of double dose dynamic enhanced spinal MRA shows the dilated coronal venous plexus throughout the thoracic spine ⇒.*

TERMINOLOGY

Synonyms

- Type 1 spinal AVF, dural AVF (dAVF)
- Dural fistula; type 1 spinal vascular malformation; radiculomeningeal fistula

Definitions

- Spinal arteriovenous (AV) fistula, present within dura, with intradural distended draining veins

IMAGING

General Features

- Best diagnostic clue
  - Abnormally enlarged, T2WI hyperintense distal cord covered with dilated pial vein flow voids
- Location
  - Intradural extramedullary flow voids from distended draining veins
  - Variable, from upper thoracic to sacrum
    - Most commonly occur at level of conus
- Size
  - Variable
- Morphology
  - Serpentine vessels on cord surface

CT Findings

- CECT
  - Slightly enlarged distal spinal cord
  - Enhancing pial veins on cord surface
  - Much more difficult to diagnose on routine CT than MR imaging
- CTA
  - Nidus in neural foramen shows focal enhancement, with prominent intradural draining dilated veins
  - Post-processing with multiplanar reformats mandatory

MR Findings

- T1WI
  - Cord enlarged, hypointense
    - Abnormal small vessel flow voids on cord pial surface
- T2WI
  - Cord enlarged, hyperintense
    - Multiple small abnormal vessel flow voids (dilated pial veins) on cord pial surface
    - Edema spares cord periphery
    - Edema has flame-shaped margins superiorly and inferiorly
    - Low signal cord periphery on T2WI consistent with venous hypertensive myelopathy
- STIR
  - Cord enlarged, hyperintense signal from cord edema
- T1WI C+
  - Multiple enhancing serpentine veins on cord surface
  - May show patchy, ill-defined enhancement within cord
    - Enhancement can be pronounced
- MRA
  - Dynamic contrast-enhanced MRA capable of defining dilated intradural veins; may guide catheter angiography
  - Rarely MR imaging may be normal or demonstrate only cord hyperintensity with no flow voids in cases with catheter confirmed fistula

Angiographic Findings

- Spinal catheter arteriography is gold standard for confirming diagnosis
  - Permits identification of exact level of arteriovenous shunt
  - Provides precise localization of anterior spinal artery
  - Provides access for interventional therapy/embolization
  - Fistula may arise anywhere from vertebral artery to internal iliac superiorly

Nonvascular Interventions

- Myelography
  - Multiple curvilinear filling defects due to dilated veins

Imaging Recommendations

- 1st perform focused MR imaging with small field of view, thin slices in both sagittal (3 mm/0 mm gap) and axial planes (4 mm/0 mm gap)
  - T1WI, T2WI, T1 C+ sequences in both planes
- Dynamic MRA capable of resolution of peripheral nidus location within foramen and defining draining veins
  - CTA may also be used, but dynamic MRA is first-line vascular imaging choice
  - Myelography no longer primary or secondary diagnostic tool with availability of MRA, CTA
- Use selective spinal arteriography to confirm diagnosis and direct treatment planning

DIFFERENTIAL DIAGNOSIS

CSF Flow Artifact

- Typically dorsal to cord on T2WI, ill-defined margins

Spinal Cord Tumor

- Distinguishable by imaging with intramedullary mass showing focal enhancement
- May be associated with prominent feeding intradural vessels

Spinal Cord Arteriovenous Malformation (SCAVM)

- Usually acute presentation (compared to insidious presentation of dAVF)
- Intramedullary or subarachnoid hemorrhage relatively common

Tortuous "Redundant" Roots From Spinal Stenosis

- Distinguished by presence of severe central stenosis

Collateral Venous Flow

- IVC occlusion may cause prominent epidural/intradural veins
- Check for small or ill-defined IVC, or metal artifact in IVC region due to filter

PATHOLOGY

General Features

- Etiology
  - Postulated to be acquired lesions, possibly from thrombosis of extradural venous system

- Venous drainage from dAVF results in increased pial vein pressure that is transmitted to intrinsic cord veins
- Venous hypertension from engorgement reduces intramedullary AV pressure gradient, causing reduced tissue perfusion and cord ischemia

- Genetics
  - No association with other CNS vascular malformations
- Lesions are extramedullary AVFs, not true AVMs
  - No intervening small vessel network
  - Fistula drains directly into venous outflow tract
  - Usually intradural
- Supplied by small tortuous arteries originating from dura mater

Staging, Grading, & Classification

- Anson-Spetzler classification: Type 1 spinal vascular malformation
- Spetzler 2002 classification: Intradural dorsal AVF

Gross Pathologic & Surgical Features

- Most commonly occurs at thoracolumbar level (T5-L3)
- Usually located either adjacent to intervertebral foramen or within dural root sleeve
- Arterial supply arises from dural branch of radicular artery
- Intradural vein drains directly into cord pial veins
- Frequently poor correlation between location of AV shunt and clinical level of spinal dysfunction
  - Subacute necrotizing myelopathy as a cause of acute deterioration ("Foix-Alajouanine syndrome") currently not considered a real entity

Microscopic Features

- True arteriovenous fistula, no intervening small vessel network

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most common presentation is progressive lower extremity weakness with both upper + lower motor neuron involvement
  - Other signs/symptoms
    - Additional symptoms include back pain, bowel/bladder dysfunction, impotence
    - Thoracolumbar fistula location spares upper extremities
    - Very rarely presents with subarachnoid hemorrhage
- Clinical profile
  - Middle-aged male with progressive lower extremity weakness exacerbated by exercise
  - Time from symptom onset to diagnosis often delayed

Demographics

- Age
  - Usually presents in 5th or 6th decade
- Gender
  - M > F
- Epidemiology
  - 80% of all spinal vascular malformations
  - 80% of patients are male
  - Usually presents in 5th or 6th decade (range: 20-80)

Natural History & Prognosis

- Slowly progressive clinical course over several years leading to paraplegia
- Cord ischemia/venous congestive myelopathy is reversible if treated early, but may become irreversible when untreated
- Bowel/bladder dysfunction and impotence rarely improve, even after successful obliteration of fistula

Treatment

- Endovascular fistula occlusion with permanent embolic agents
- Surgical fistula obliteration
  - 40-60% improve following obliteration of fistula, 40-50% stabilize myelopathic symptoms
  - T2WI cord edema decreases over 1-4 months following successful embolization
  - Improved cord appearance on MR following treatment does not necessarily correlate with improved symptoms
  - Persistent edema and enhancement of cord can occur even with successful clinical treatment of fistula

DIAGNOSTIC CHECKLIST

Consider

- Dynamic enhanced MRA is first-line vascular imaging choice
- Consider other types of spinal vascular malformation if evidence of intramedullary flow voids
- Rarely, cervical type 1 dAVF may drain intracranially with high flow, varix formation, and SAH
- Posterior fossa dural fistula may mimic cervical spinal type 1 fistula when intracranial fistula has caudal intraspinal venous drainage

Image Interpretation Pearls

- Imaging and clinical findings are frequently subtle or nonspecific; early diagnosis requires high level of suspicion

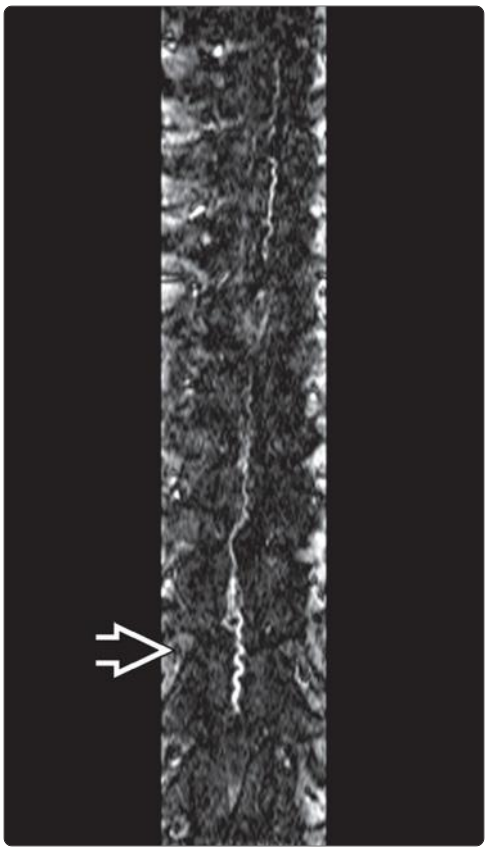
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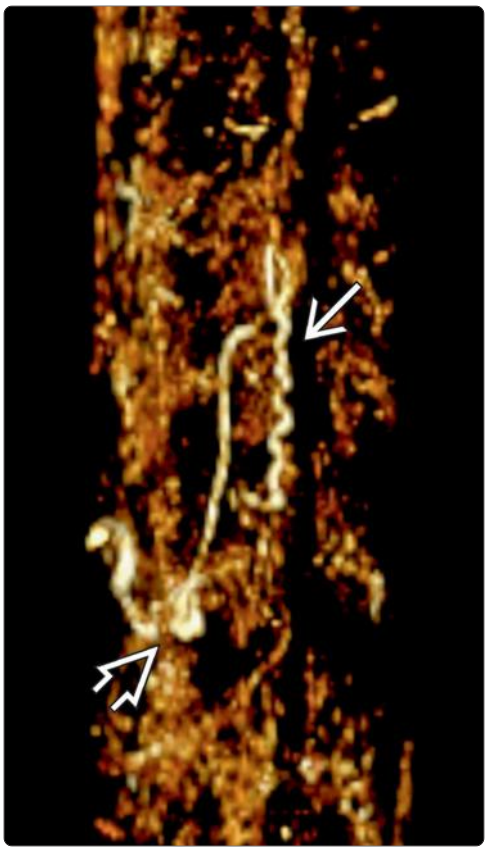
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(Left) *Sagittal T2WI MR in this case of type 1 fistula shows diffuse T2 hyperintensity throughout the thoracic cord with relative sparing of the cord periphery. There are no conspicuous flow voids on the dorsal cord periphery.* **(Right)** *Sagittal T1WI C+ MR in this case of type 1 fistula shows extensive confluent enhancement of the distal thoracic cord. The differential is extensive, including infectious and inflammatory etiologies, such as demyelinating disease.*





(Left) *Coronal reformat of dynamic enhanced spinal MRA images shows abnormal vessels on the dorsal aspect of the distal thoracic cord ⇨ reflecting the arterialized veins.* **(Right)** *Coronal 3D volume render of the dynamic MRA shows the site of the peripheral fistula at the right L1 level ⇨ with shunting of flow to the coronal venous plexus around the cord ⇨.*

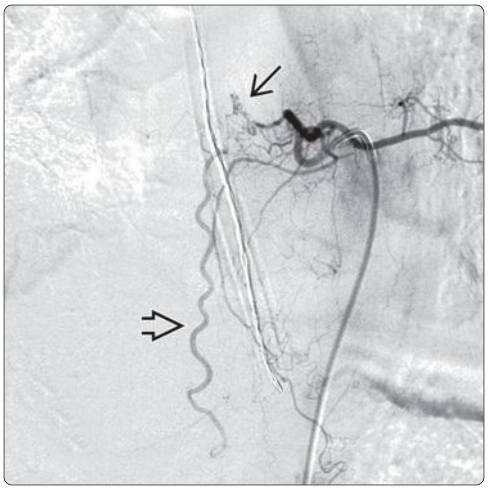
(Left) *Sagittal T2WI MR shows the typical cord edema pattern of a spinal dural fistula, with central cord T2 hyperintensity and multiple flow voids on the cord surface.* **(Right)** *Sagittal T1WI C+ FS MR shows the classic pattern of cord enhancement, which is ill-defined, diffuse, and without sharp margination.*





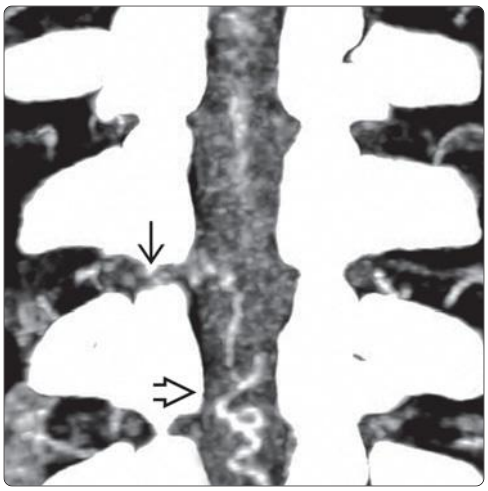
(Left) *Coronal reformat of dynamic enhanced MR angiography shows the abnormal cord surface vessels, and more precisely defines the site of the fistula on the left at T9 as a more compact focus of vessels within the neural foramen.* **(Right)** *Catheter angiography with injection of the left T9 intercostal artery shows type 1 fistula site at the neural foraminal level and the flow into the dilated coronal venous plexus.*

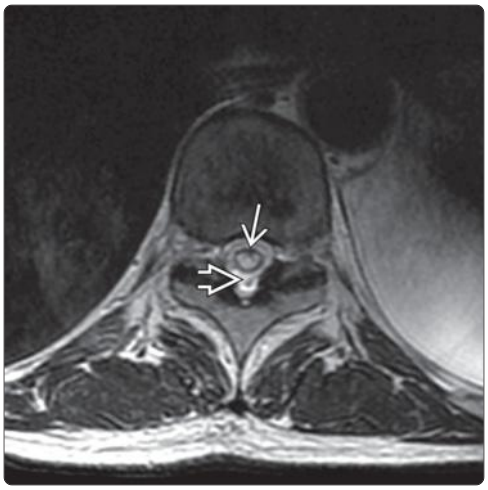




(Left) *Sagittal T2WI MR shows enlarged draining veins as multiple flow voids surrounding the cord. High T2 signal in the cord tapers at both ends and tends to spare periphery, typical of venous hypertensive edema.* **(Right)** *Coronal reformat from a CTA shows type 1 fistula in the right thoracic neural foramen, with enlarged inferior draining veins.*

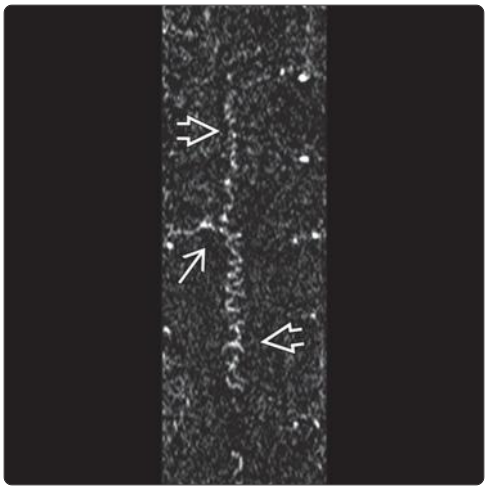








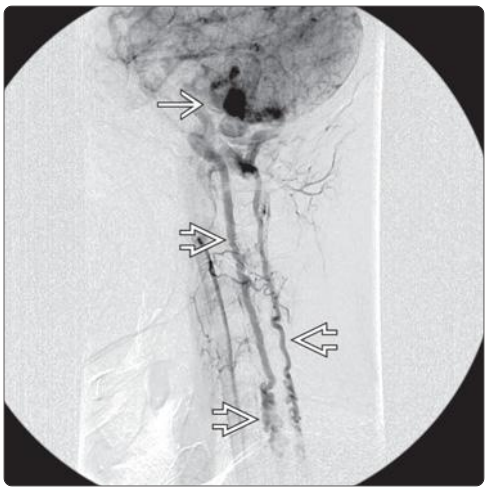
(Left) *Axial T2WI MR shows T2 cord signal change without vascular flow voids and central cord hyperintensity* ⇨ *with sparing of the cord periphery in this patient with type 1 dAVF. CSF flow voids are present surrounding the cord* ⇨*, but no definite vessel flow voids are seen.* **(Right)** *Sagittal T1 C+ FS MR shows faint serration to the dorsal margin of the cord* ⇨ *from mild vascular enhancement in this type 1 fistula. No definite cord enhancement is present. Lack of enhancement does not exclude a fistula.*





(Left) *Coronal reconstruction from a dynamic contrast-enhanced MRA data set shows the location of the fistula within the neural foramen* ⇨ *and shunting to the dilated intradural spinal veins* ⇨*.* **(Right)** *Catheter angiogram shows shunting of contrast into the venous plexus surrounding the cord* ⇨ *from the right-sided type 1 fistula* ⇨*.*





(Left) *Sagittal T1WI C+ MR demonstrates diffuse enhancement of cord from cervicomedullary junction to C5. The pattern mimics the look of spinal type 1 dural fistula, but myelopathy was caused by the posterior fossa fistula* ⇨ *with caudal intraspinal venous drainage* ⇨*.* **(Right)** *Venous phase of the left vertebral injection shows a posterior fossa dural fistula* ⇨ *(Cognard type 5) supplied by meningeal branches with extensive intraspinal draining veins dorsal and ventral to the cord* ⇨*; a type 1 fistula mimic.*

Type 2 Arteriovenous Malformation (AVM)

KEY FACTS

TERMINOLOGY

- Synonyms
  - Glomus-type AVM
  - Plexiform AVM
- Direct arterial/venous communications forming compact nidus within cord

IMAGING

- Multiple, well-defined serpentine flow voids within substance of cord
- Large cord, heterogeneous signal (blood products), flow voids
- Dynamic enhanced MRA capable of defining enlarged feeding arteries, nidus, and enlarged draining veins
- Supplied by anterior spinal artery &/or posterior spinal artery
  - Nidus drains to coronal venous plexus (on cord surface), which drains anterograde to extradural space

TOP DIFFERENTIAL DIAGNOSES

- Intramedullary neoplasm
- Cavernous hemangioma
- Paraganglioma with large feeding vessels (rare)

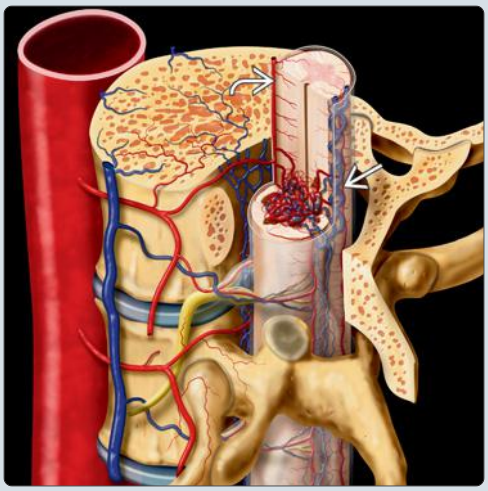
PATHOLOGY

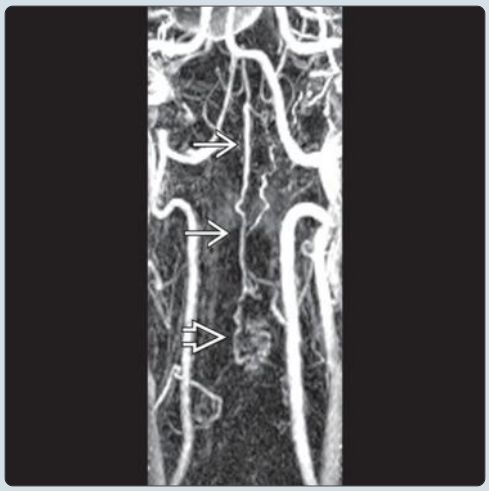
- Associated with cutaneous angiomas, Klippel-Trenaunay-Weber, Rendu-Osler-Weber syndromes
- Neurologic deterioration with subarachnoid hemorrhage (SAH), ischemia from vascular steal, cord compression, venous hypertension

CLINICAL ISSUES

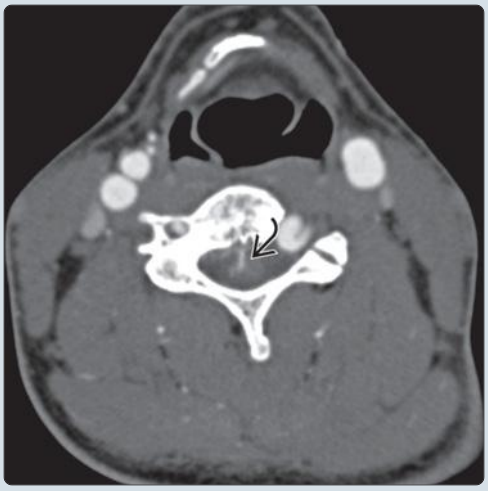
- SAH most common symptom; pain, myelopathy
- Surgical resection + preop embolization (aneurysms, nidus)

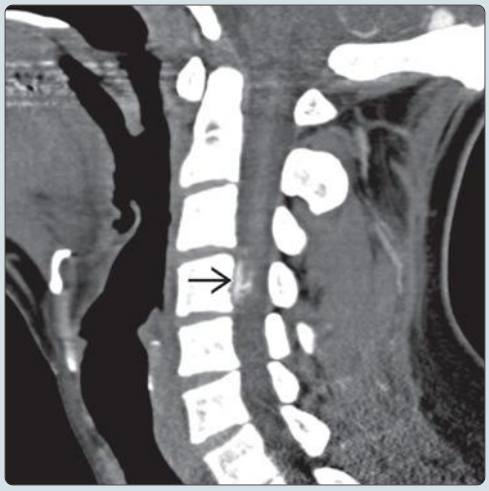
(Left) *Sagittal oblique graphic of the thoracic cord shows focal, compact intramedullary nidus of type 2 arteriovenous malformation (AVM) fed by branches from the anterior spinal artery.* **(Right)** *Dynamic enhanced coronal MRA of the cervical spine allows high-resolution MRA within 30-60 seconds and illustrates the feeding anterior spinal artery extending inferiorly to the AVM nidus.*





(Left) *Axial CTA in a 29-year-old man with bilateral extremity weakness demonstrates a prominent intramedullary nidus.* **(Right)** *Sagittal CTA in the same patient depicts a vascular nidus in the ventral cord at the C3 level. Spinal AVMs may lead to spinal cord dysfunction by various pathophysiological mechanisms, including venous hypertension with secondary congestive myelopathy, venous congestion, hemorrhage, mass effect, and vascular steal leading to ischemia.*





TERMINOLOGY

Synonyms

- Glomus-type AVM, plexiform AVM

Definitions

- Direct arterial/venous communications forming compact nidus within cord
- General spine vascular malformation classification
  - **Type 1**: Dural arteriovenous fistula (DAVF)
    - Most common type (up to 80%)
  - **Type 2**: Intramedullary glomus-type AVM (similar to brain AVM)
  - **Type 3**: Juvenile-type AVM (intramedullary extramedullary)
  - **Type 4**: Intradural extra-/perimedullary AVF

IMAGING

General Features

- Best diagnostic clue
  - Type 1: Flow voids on cord surface with cord T2 hyperintensity
  - Type 2: Intramedullary nidus (may extend to dorsal subpial surface)
  - Type 3: Nidus may have extramedullary and extraspinal extensions
  - Type 4: Ventral fistula on cord surface (venous varices displace or distort cord)
- Location
  - **Type 2**: Intramedullary nidus within cervical or thoracic cord
- Size
  - Variable
- Morphology
  - Multiple, well-defined serpentine flow voids within substance of cord

CT Findings

- NECT
  - Usually normal
- CECT
  - May show enlarged cord with enhancing nidus
- CTA
  - Shows enhancing nidus position with respect to cord
  - Delineates multiple feeding vessels and draining veins

MR Findings

- T1WI
  - Large cord, heterogeneous signal (blood products), flow voids
- T2WI
  - Cord hyperintense (edema, gliosis, ischemia) or mixed (blood)
- STIR
  - Shows flow voids, edema within cord
- T2* GRE
  - More sensitive to blood by-products of AVM hemorrhage
- T1WI C+
  - Variable enhancement of nidus, cord, and vessels
- MRA
  - Dynamic enhanced MRA capable of defining enlarged feeding arteries, nidus, and enlarged draining veins
    - Different MRA sequences improve temporal resolution, such as TWIST and TRICKS
- Compare: Types 1 and 4 fistulas
  - T1WI: Ventral type 4 fistula, large flow voids distort ± displace cord
  - T2WI: Type 1 fistula with cord hyperintensity + flow voids
  - Contrast: Enhancing pial vessels, epidural plexus, ± patchy enhancement distal cord

Angiographic Findings

- Conventional catheter
  - Type 2: Supplied by anterior spinal artery (ASA) or posterior spinal artery (PSA); nidus drains to coronal venous plexus (on cord surface), which in turn drains anterograde to extradural space
- Rotational angiography: Post-processed images used to characterize lesion in relation to surrounding bone and to enhance ability to precisely localize and obliterate abnormality

Nonvascular Interventions

- Myelography
  - Serpentine filling defects in contrast column from draining veins, enlarged feeding vessels
  - May see slight cord expansion at nidus level

Imaging Recommendations

- Contrast-enhanced MR; consider spinal angiography ± embolization

DIFFERENTIAL DIAGNOSIS

Intramedullary Neoplasm

- Ependymoma: Heterogeneous (cysts, blood products)
- Astrocytoma: Multisegmental enhancing mass, no enlarged vessels
- Hemorrhagic metastatic disease (melanoma)

Intradural Extramedullary Tumor With Prominent Vascular Supply

- Paraganglioma: Markedly enhancing intradural extramedullary mass
- Hemangioblastoma: Larger lesions may show mixed hyperintensity with pial flow voids, hemorrhage

Cavernous Hemangioma

- Well-defined lesion showing speckled pattern on T1WI and T2WI (similar to brain)
- No prominent vessels

PATHOLOGY

General Features

- Etiology
  - Type 2: Compact nidus, high flow, aneurysms common (20-44%)
- Genetics
  - Sporadic or syndromic
    - Type 2: Associated with cutaneous angiomas, Klippel-Trenaunay-Weber, Rendu-Osler-Weber syndromes
- Associated abnormalities

- Aneurysms of feeding vessels in type 2 AVMs (40%)
- Causes of neurologic deterioration include subarachnoid hemorrhage (SAH), ischemia from vascular steal, cord compression, and venous hypertension
- Location
  - Type 2 (glomus): Cervical/upper thoracic (may occur anywhere)
- Embryology
  - Persistence of primitive direct communications between arterial and venous channels, without intervening capillary bed

Staging, Grading, & Classification

- Newest classification of AVMs as extradural, extradural intradural, or intradural
  - Intradural: Dorsal or ventral AVF, intramedullary (compact, diffuse, conus)
- Angiographic classification based on involvement of axial artery in supply of AVF/AVM, involvement of axial vein(s) in drainage of AVF/AVM, and presence or absence of nidus
  - Axial arteries consist of 1 anterior and 2 posterior spinal arteries
  - Axial veins are perimedullary veins that consist of up to 3 anterior and 3 posterior spinal veins

Gross Pathologic & Surgical Features

- Type 2: Compact intramedullary nidus lacks normal capillary bed
  - **No parenchyma within nidus**; nidus may have pial extension

Microscopic Features

- Type 2: Abnormal vessels with variable wall thickness, internal elastic lamina
- Reactive change in surrounding tissue: Gliosis, cytoid bodies, Rosenthal fibers; hemosiderin deposition common; ± Ca++

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Type 2 AVM: SAH (most common symptom), pain, myelopathy
- Clinical profile
  - Type 2: Adult with acute SAH, myelopathy
- Pregnancy associated with worsening symptoms
  - 1 study showed ↑ risk of hemorrhage from AVMs during pregnancy: Hemorrhage rate of 8.1% per pregnancy or 10.8% per year

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - Intramedullary (types 2, 3): 15-20% of spinal AVMs
  - AVMs account for 10% of spinal masses

Natural History & Prognosis

- Good outcome in type 2 (glomus) and type 4 (perimedullary)
- Pooled analysis shows annual hemorrhage rate prior to treatment of 4%, increasing to 10% for AVMs with prior hemorrhage
  - Overall rates of complete obliteration 78% for surgery, 33% for endovascular treatment
  - Clinical worsening occurred in 12% of patients after surgical treatment, 13% after endovascular treatment
  - No hemorrhage after complete obliteration, annual hemorrhage rate of 3% after partial surgical treatment
    - No hemorrhages reported over 196 patient-years after partial endovascular treatment
  - Postoperative reduction in symptoms is frequent despite small remnants
    - Risk of neurological deficits seems relatively low even in residual AVMs

Treatment

- Type 2: Surgical resection + preop embolization (aneurysms, nidus)
  - Reports of endovascular therapy with Onyx

DIAGNOSTIC CHECKLIST

Consider

- CTA or dynamic MRA as localization tool prior to catheter angiography
  - Can define malformation type, location of ASA

Image Interpretation Pearls

- Definition of intramedullary involvement is critical for classification, prognosis, treatment options

SELECTED REFERENCES

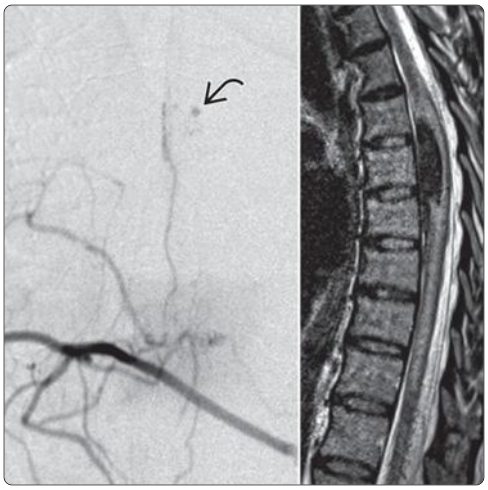
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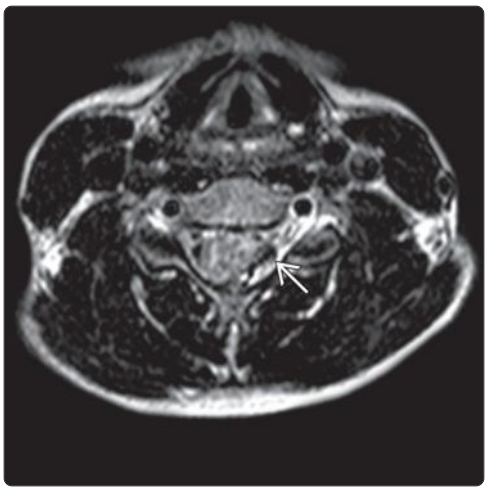


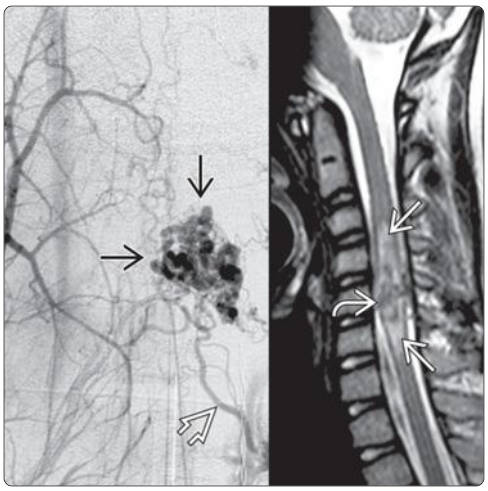
(Left) *Sagittal T2WI MR in a patient with a type 2 AVM exhibits mild cord expansion and heterogeneous signal due to hyperacute/acute blood products. Type 2 AVMs with prior hemorrhage, particularly within 10 years, demonstrate a greater risk of hemorrhage. Complete obliteration and even partial endovascular treatment have been shown to significantly decrease the hemorrhage rate.* **(Right)** *Sagittal T1WI C+ MR in the same patient shows irregular enhancement of the intramedullary nidus.*





(Left) *Lateral DSA reveals a glomerular AVM arising from a radicular branch of the right vertebral artery at the C4-C5 level. There is opacification of the anterior spinal artery, the origin of which is not distinct from the radicular branch supplying the AVM.* **(Right)** *DSA (left) reveals a thoracic type 2 AVM with an ill-defined nidus and an intranidal aneurysm. Sagittal T2WI MR (right) displays intramedullary hemorrhage and cord expansion, presumably due to rupture of the intranidal aneurysm.*





(Left) *Axial T2WI MR reveals fusiform cord expansion with multiple serpentine flow voids around and within the cord. Abnormal intramedullary hyperintensity is likely due to edema.* **(Right)** *Anteroposterior DSA (left) demonstrates the compact AVM nidus involving the cervical cord. The draining vein is seen extending inferiorly to the cervical epidural plexus. Sagittal T2WI MR (right) shows mild cord expansion with heterogeneous intramedullary T2 signal due to blood products and edema.*

Type 3 Arteriovenous Malformation (AVM)

KEY FACTS

TERMINOLOGY

- Synonyms: Juvenile AVM; intramedullary extramedullary AVM
- Juvenile-type AVM (large, complex, intramedullary extramedullary AVM)
- Rarest of spinal AVMs (7%)

IMAGING

- Large complex nidus, multiple feeding vessels; may be intramedullary and extramedullary, and even extraspinal

TOP DIFFERENTIAL DIAGNOSES

- Intramedullary neoplasm
- Cavernous angioma
- Type 4 perimedullary fistula
- Paraganglioma: Enhancing intradural extramedullary mass (rare)

PATHOLOGY

- Associated with Cobb syndrome (metameric vascular malformation with triad of spinal cord, skin, bone)
- Variety of vascular lesions may be present, including paravertebral and epidural hemangioma

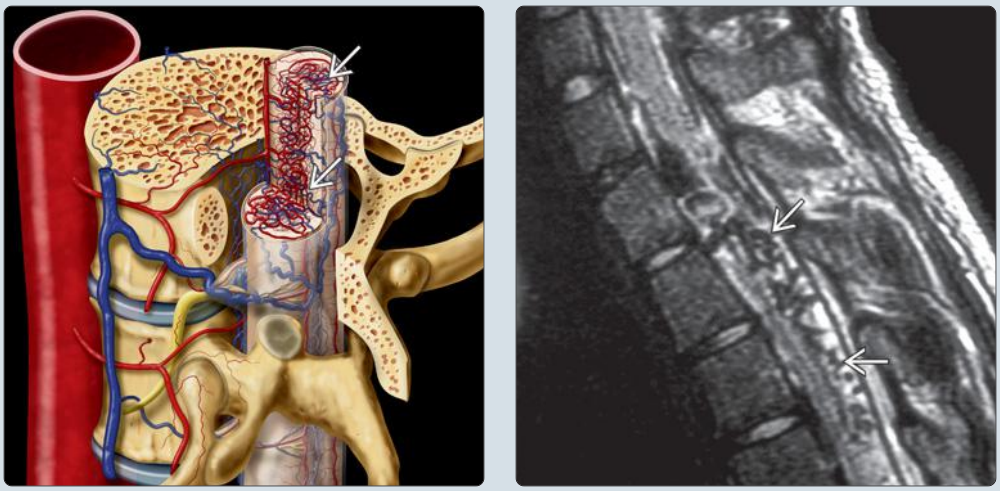
CLINICAL ISSUES

- Progressive neurologic decline (weakness), subarachnoid hemorrhage
  - Pain in lower extremities or back; motor deficit
  - Poor prognosis for type 3 (juvenile) AVM

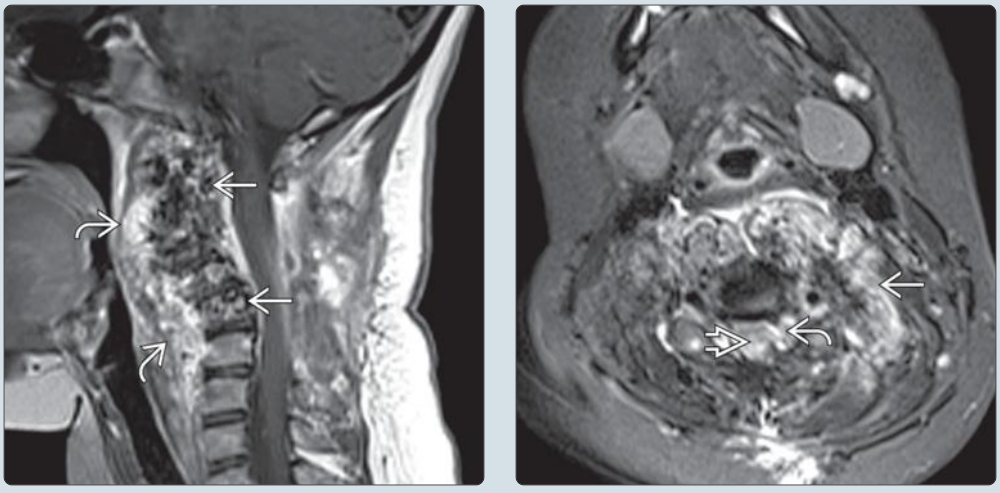
DIAGNOSTIC CHECKLIST

- Definition of intramedullary involvement is critical for classification, prognosis, treatment options
- CTA or dynamic MRA serves as localization tool prior to catheter angiography

(Left) *Sagittal oblique graphic of the thoracic cord shows a large complex intramedullary nidus of a type 3 arteriovenous malformation (AVM) with multiple feeding vessels.* **(Right)** *Sagittal T2WI MR exhibits extensive flow voids, involving a long segment of the cervical cord, seen throughout the subarachnoid space. These lesions are fed by multiple arteries at multiple vertebral levels. An apoplectic presentation from SAH or intramedullary hemorrhage is strongly associated with intramedullary AVMs.*



(Left) *Sagittal T1WI C+ MR displays a vascular lesion composed of multiple flows in the cervical spine, involving the vertebral bodies and prevertebral muscles.* **(Right)** *Axial T1WI C+ FS MR in the same patient illustrates the intramedullary, extramedullary, and extradural components of this type 3 AVM. MR has the advantage of demonstrating the intraspinal vascular component as well as the extraspinal portion insinuating into the perivertebral space.*



TERMINOLOGY

Synonyms

- Juvenile arteriovenous malformation (AVM); intramedullary extramedullary AVM

Definitions

- Direct arterial/venous communications without capillary bed involving cord
- General spine vascular malformation classification
  - **Type 1**: Dural arteriovenous fistula (DAVF)
    - Most common type (up to 80%)
  - **Type 2**: Intramedullary glomus-type AVM (similar to brain AVM)
  - **Type 3**: Juvenile-type AVM (large, complex, intramedullary extramedullary AVM)
    - Rarest of spinal AVMs (7%)
  - **Type 4**: Intradural extra-/perimedullary AVF

IMAGING

General Features

- Best diagnostic clue
  - Type 1: Flow voids on cord surface with cord hyperintensity
  - Type 2: Intramedullary nidus (may extend to dorsal subpial surface)
  - Type 3: Nidus may have extramedullary and extraspinal extension
  - Type 4: Pial fistula (venous varices displace ± distort cord)
- Location
  - Type 3: Intramedullary extramedullary
- Size
  - Type 3 AVMs tend to be large, complex
- Morphology
  - Multiple serpentine flow voids within substance of cord + subarachnoid and extradural locations

CT Findings

- NECT
  - Usually normal (rarely, widened interpedicular distance ± posterior vertebral scalloping)
- CECT
  - Enlarged cord with enhancing nidus and pial vessels
- CTA
  - Shows enhancing nidus position with respect to cord
  - Delineates multiple feeding arteries and draining veins

MR Findings

- T1WI
  - Large cord, heterogeneous signal (blood products), flow voids
- T2WI
  - Cord is hyperintense (edema, gliosis, ischemia) or mixed (blood)
- STIR
  - Shows flow voids, edema within cord
- T2* GRE
  - More sensitive to blood by-products of AVM hemorrhage
- T1WI C+
  - Variable enhancement of nidus, cord, vessels
- MRA
  - Dynamic, enhanced MRA capable of defining enlarged feeding arteries, nidus, and enlarged draining veins
    - Newer dynamic, enhanced MRA techniques have improved spatial and temporal resolution
      - Strongly improved vessel-to-background contrast by using fast acquisition combined with contrast agent bolus injection
      - Able to visualize vessels supplying or draining spinal cord and to differentiate spinal cord arteries from veins
      - Guide-selective catheterization of all arteries potentially supplying spinal cord and meninges

Angiographic Findings

- Conventional
  - Type 1 DAVF: Peripheral dural fistula may arise anywhere from vertebral artery superiorly to internal iliac artery inferiorly
  - Type 2 AVM: Nidus supplied by anterior spinal artery (ASA) ± posterior spinal artery (PSA)
    - Nidus drains to coronal venous plexus (on cord surface), which in turn drains anterograde to extradural space
  - Type 3 AVM: Large complex nidus, multiple feeding vessels; may be intramedullary and extramedullary, or even extraspinal
  - Type 4 AVF: Feeding vessel from ASA or PSA connects directly with spinal vein (no nidus)
- Regarded as only technique sufficiently reliable for pretreatment detection and characterization of spinal vascular malformations
  - Drawbacks: Invasive, exposure to ionizing radiation, small risk of neurologic complications

Nonvascular Interventions

- Myelography
  - Serpentine filling defects from draining veins, enlarged feeding vessels
  - Cord expansion due to edema, may result in myelographic block

Imaging Recommendations

- Enhanced MR and dynamic MRA; CTA
- Consider spinal DSA ± embolization

DIFFERENTIAL DIAGNOSIS

Intramedullary Neoplasm

- Ependymoma: Heterogeneous (cysts, blood products)
- Astrocytoma: Multisegmental enhancing mass, no enlarged vessels
- Uncommon: Metastasis with hemorrhage (melanoma, renal cell)

Intradural Extramedullary Tumor With Prominent Vascular Supply

- Paraganglioma: Markedly enhancing intradural extramedullary mass (rare)
- Hemangioblastoma: Enhancing nodule with cyst, often in subpial location with associated prominent vascular flow voids

Cavernous Hemangioma

- Well-defined lesion showing speckled pattern on T1 and T2 images without prominent vessels

Type 4 Perimedullary Fistula

- Prominent high-flow feeders with varices may compress cord and thus simulate intramedullary lesion

PATHOLOGY

General Features

- Etiology
  - Type 1: Hypothesized to be acquired, possibly from thrombosis of extradural venous system
  - Type 2: Aneurysms common (20-44%), congenital, compact nidus, high flow
  - Type 3: Large diffuse nidus, cord ischemia, venous hypertension, congenital
  - Type 4: Congenital; may be acquired after trauma; (A) venous hypertension; (B, C) arterial steal; (C) cord compression
- Genetics
  - Sporadic or syndromic
    - Type 3: Associated with Cobb syndrome (metameric vascular malformation with triad of spinal cord, skin, bone involvement)
- Associated abnormalities
  - Type 3 AVM is associated with Cobb syndrome
    - Also called "cutaneomeningospinal angiomatosis"
    - Variety of vascular lesions may be present, including paravertebral and epidural hemangiomas
    - Pathogenesis of neurological deficit in Cobb syndrome is thought to be ischemia secondary to diversion of blood into malformation or spinal cord compression
- Location
  - Type 1 DAVF: Lower thoracic/conus common
  - Type 2 AVM (glomus): Cervical/upper thoracic (may occur anywhere)
  - Type 3 AVM (juvenile): Cervical/upper thoracic (may occur anywhere)
  - Type 4 AVF: Conus medullaris (types A, B), thoracic (type C)
- Embryology
  - Persistence of primitive direct communications between arterial/venous channels, without intervening capillary bed

Gross Pathologic & Surgical Features

- Type 3: Large, complex intramedullary lesion; **normal neural parenchyma inside nidus** (may involve extramedullary ± extradural spaces)

Microscopic Features

- Abnormal vessels with variable wall thickness, internal elastic lamina
- Reactive change in surrounding tissue: Gliosis, cytoid bodies, Rosenthal fibers; hemosiderin deposition common; ± Ca++

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - **Type 3**: Progressive neurologic decline (weakness), SAH
    - Pain in lower extremities or back, motor deficit
    - Cobb syndrome shows port-wine skin lesions with dermatomal distribution
- Other signs/symptoms
  - Lumbar puncture: SAH, ↑ protein and cell counts
- Clinical profile
  - **Type 3**: Young adult with acute (subarachnoid hemorrhage) SAH, myelopathy

Demographics

- Age
  - 1st-3rd decades
- Gender
  - Slight male predominance for Cobb syndrome
- Epidemiology
  - Intramedullary (types 2, 3):15-20% of spinal AVM; type 4: 10-20%
  - AVMs account for < 10% of spinal masses

Natural History & Prognosis

- Poor prognosis for type 3 (juvenile) AVM

Treatment

- Type 3: Complete resection generally not possible; palliative therapy
  - Treatment algorithm for massive AVMs must be individualized
  - Combination of staged embolization and subsequent surgery

DIAGNOSTIC CHECKLIST

Consider

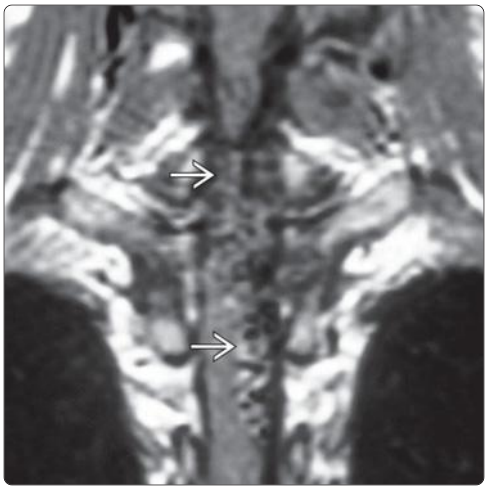
- CTA or dynamic MRA as localization tool prior to catheter angiography
  - Can define malformation type, location of ASA

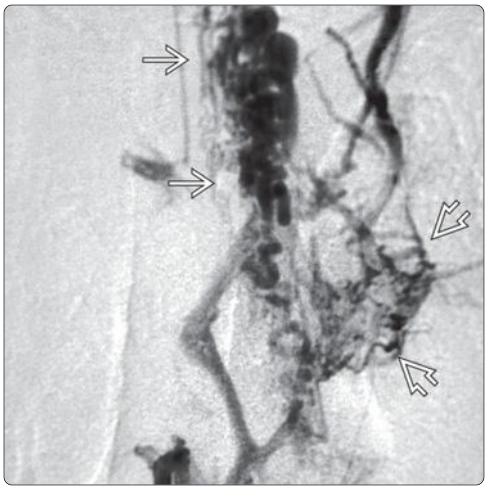
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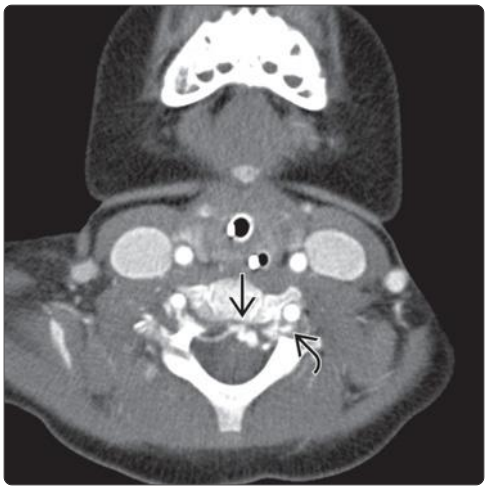
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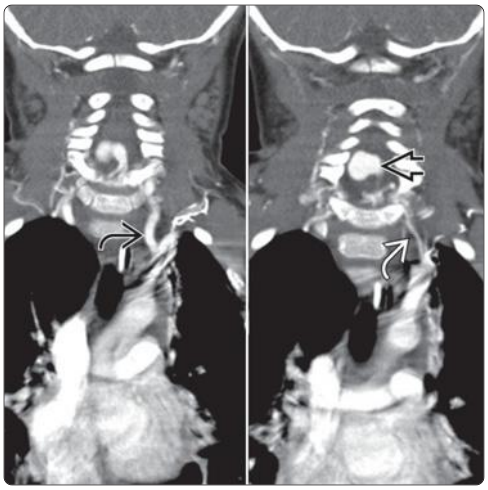
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(Left) *Coronal T1WI MR shows a long segment of parenchymal flow voids extending into the adjacent subarachnoid space to the left of the cord in this extensive intra- and extramedullary AVM.* **(Right)** *AP vertebral angiogram in a patient with Cobb syndrome shows both extradural draining veins and an intradural component in the subarachnoid space. Catheter angiography is the ultimate imaging technique for diagnosing, localizing, and classifying spinal vascular lesions.*





(Left) *Axial CTA demonstrates a convergence of arterial vessels at the ventral cord surface related to a type 3 AVM. This appears to be supplied by the anterior spinal artery and numerous tortuous arterial feeders that enter bilateral neural foramina.* **(Right)** *Coronal CTA in the same patient reveals an intramedullary aneurysm. Some of the arterial feeders to this AVM appear to arise from thyrocervical branches and from muscular branches of the proximal vertebral arteries.*





(Left) *Sagittal CTA in the same patient shows a 2nd plaque of abnormal vasculature within the thoracic cord at the level of T5-T8, which is predominantly intramedullary in location. The cervical intramedullary aneurysm is also seen at the C5-C6 level.* **(Right)** *Sagittal MRA in the same patient illustrates multiple abnormal vessels entering the vertebral canal in the cervical region that communicate with the aneurysm, which is approximately 1 cm in dimension.*

Type 4 Vascular Malformation (AVF)

KEY FACTS

TERMINOLOGY

- Synonyms: Perimedullary fistula; type 4 spinal vascular malformation, pial arteriovenous fistula (AVF)
- Direct intradural extramedullary arterial/venous communication from ASA, or PSA to draining vein without capillary bed

IMAGING

- Hyperintense cord + flow voids
- Intradural location for fistula, adjacent to cord
- Draining veins may be pronounced on dorsal or ventral surface of cord
- Feeding vessel from ASA or PSA connects directly with spinal vein (no nidus)

TOP DIFFERENTIAL DIAGNOSES

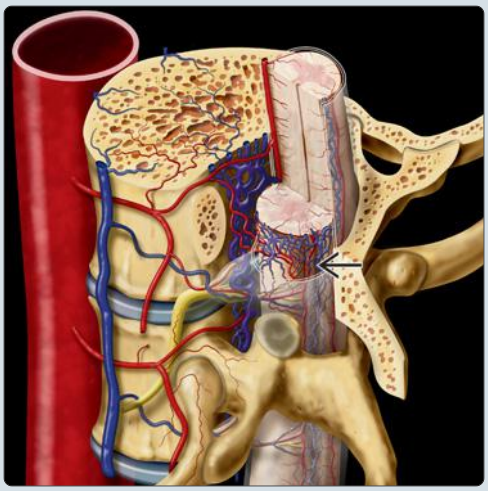
- Normal CSF flow artifact
- Type 1 dural fistula
- Lumbar canal stenosis with tortuous intradural roots
- Intramedullary neoplasm

PATHOLOGY

- 4-A: Small AVF with slow flow, mild venous enlargement
- 4-B: Intermediate AVF, dilated feeding arteries; high flow rate
- 4-C: Large AVF, dilated feeding arteries; dilated, tortuous veins
- Associations
  - Hereditary hemorrhagic telangiectasia
  - Cobb syndrome
  - Klippel-Trénaunay-Weber syndrome

CLINICAL ISSUES

- > 90% of patients present with neurological deficits
  - Hemorrhage at presentation in 36%
- Embolization or surgical resection based upon anatomy and size: Surgical resection, surgical resection or embolization, embolization





(Left) *Coronal oblique graphic of the thoracic cord shows the intradural site of arteriovenous fistula (AVF, type 4) on the dorsal cord surface with diffuse venous engorgement. A fistula is more typically along the ventral cord surface.* **(Right)** *Sagittal T2 MR shows multiple serpentine intradural extramedullary flow voids dorsal to the thoracic cord, plus focal cord abnormality due to a high-flow aneurysm.*





(Left) *Sagittal T1 C+ MR shows multiple enhancing intradural vessels surrounding the distal cord.* **(Right)** *Sagittal T2 MR shows high signal within distal cord due to edema, and more focal low signal reflecting compressing perimedullary vessels.*

TERMINOLOGY

Synonyms

- Perimedullary arteriovenous fistula (AVF), type 4 spinal vascular malformation, pial AVF, ventral AVF

Definitions

- **General spine vascular malformation classification**
  - **Type 1**: Dural arteriovenous fistula (DAVF)
    - Most common = type 1 (up to 80%)
  - **Type 2**: Intramedullary glomus-type AVM (similar to brain AVM)
  - **Type 3**: Juvenile-type AVM (intramedullary, extramedullary)
    - 2nd most common = intramedullary types 2 and 3 (15-20%)
  - **Type 4**: Intradural extra-/perimedullary AVF (types A, B, C)
- Type 4: Direct intradural extramedullary arterial/venous communication from anterior spinal artery (ASA), or posterior spinal artery (PSA) to draining vein without capillary bed
  - Heterogenous group of lesions ranging from slow flow fistula (IVa) to large high flow (IVc)

IMAGING

General Features

- Best diagnostic clue
  - Type 1: Flow voids with cord hyperintensity
  - Type 2: Intramedullary nidus (may extend to dorsal subpial surface)
  - Type 3: Nidus may have extramedullary and extraspinal extension
  - **Type 4**: Ventral fistula (venous varices displace, distort cord)
- Location
  - Type 4: Intradural location for fistula, adjacent to cord
  - Draining veins may be pronounced on dorsal or ventral surface of cord
  - Tends to be along ventral pial surface
- Size
  - Variable depending on number of feeding vessels, size of draining veins, presence of aneurysms
- Morphology
  - Multiple well-defined serpentine flow voids

CT Findings

- CECT
  - May show enlarged cord with enhancing nidus, pial vessels (rare)
- CTA
  - Intradural draining veins identifiable; precise fistula site more difficult
  - Postprocessing mandatory for best anatomic display

MR Findings

- T1WI
  - Type 4 (perimedullary/pial)
    - Ventral fistula, large flow voids distort/displace cord
    - May be difficult to distinguish from type 1 AVF on imaging
- T2WI
  - Hyperintense cord + flow voids
- T1WI C+
  - Enhancing pial vessels, epidural plexus, ± patchy enhancement distal cord
- MRA
  - Dynamic enhanced MRA capable of defining distending draining veins
  - Spatial resolution generally inadequate for arterial feeders

Angiographic Findings

- Conventional
  - Type 1: Peripheral dural fistula may arise anywhere from vertebral artery inferiorly to internal iliac supply
  - Type 2: Supplied by anterior spinal artery or posterior spinal artery; nidus drains to coronal venous plexus (on cord surface), which in turn drains anterograde to extradural space
  - Type 3: Large complex nidus, multiple feeding vessels; may be intramedullary and extramedullary and even extraspinal
  - **Type 4**: Feeding vessel from ASA or PSA connects directly with spinal vein (no nidus) on pial surface

Nonvascular Interventions

- Myelography
  - Serpentine filling defects along cord

Imaging Recommendations

- Contrast-enhanced MR; CTA; consider spinal angiography ± embolization

DIFFERENTIAL DIAGNOSIS

Normal CSF Flow Artifact

- Most prominent dorsal to thoracic cord
- Ill-defined margins, appearance varies with different sequences

Type 1 Dural Fistula

- May be indistinguishable from type 4 by MR, CTA

Lumbar Canal Stenosis With Tortuous Intradural Roots

- Distinguished by presence of severe central stenosis
- Redundant serpentine roots stop at conus

Intramedullary Neoplasm

- Ependymoma: Heterogeneous (cysts, blood products)
- Astrocytoma: Multisegmental enhancing mass, no enlarged vessels

Intradural Extramedullary Tumor With Prominent Vascular Supply

- Paraganglioma: Markedly enhancing intradural extramedullary mass

PATHOLOGY

General Features

- Etiology
  - Type 4: Unknown, but several associations
- Genetics

- Sporadic or syndromic
  - Type 1: None
  - Type 2: Associated with cutaneous angiomas, Klippel-Trénaunay-Weber, Rendu-Osler-Weber syndromes
  - Type 3: Associated with Cobb syndrome (metameric vascular malformation involving triad of spinal cord, skin, bone)
  - **Type 4**
    - Hereditary hemorrhagic telangiectasia
    - Cobb syndrome
    - Klippel-Trénaunay-Weber syndrome
- Associated abnormalities
  - Location
    - Type 1: Lower thoracic/conus common
    - Type 2 (glomus): Cervical/upper thoracic (may occur anywhere)
    - Type 3 (juvenile): Cervical/upper thoracic (may occur anywhere)
    - Type 4: Conus medullaris (type A, B), thoracic (type C)
  - Embryology
    - Persistence of primitive direct communications between arterial and venous channels, without intervening capillary bed

Staging, Grading, & Classification

- 4-A: Small AVF with slow flow, mild venous enlargement
- 4-B: Intermediate AVF, dilated feeding arteries; high flow rate
- 4-C: Large AVF, dilated feeding arteries; dilated, tortuous veins

Gross Pathologic & Surgical Features

- Type 4: Direct fistula between ASA/PSA and draining vein, no nidus

Microscopic Features

- Abnormal vessels with variable wall thickness, internal elastic lamina
- Reactive change in surrounding tissue: Gliosis, cytoid bodies, Rosenthal fibers; hemosiderin deposition common; ± Ca++

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - **Type 4:** Progressive conus/cauda equina syndrome, subarachnoid hemorrhage (SAH)
    - Type 4-A with slow flow, venous hypertension primary cause of symptoms
    - Types 4-B, C show arterial steal and cord compression from venous varices
  - > 90% of patients present with neurological deficits
    - Paraparesis 78%
    - Sensory deficit 66%
    - Bowel or bladder symptoms 62%
  - Hemorrhage at presentation in 36%
    - Subarachnoid 75%
    - Hematomyelia 32%
    - No definite variation across type 4 subgroups with respect to incidence of hemorrhage
    - 2.5% annual hemorrhage rate if left untreated
    - Hemorrhagic lesions have greater annual rate of subsequent hemorrhage (5.6%)
  - Rarely, patients may present with respiratory symptoms with cervical pial AVF

Demographics

- Age
  - Child and adult
  - Mean age: 32 years
- Gender
  - ~ 60% male
- Epidemiology
  - Intramedullary (type 2, 3): 15-20% of spinal AVM
  - Type 4: 10-20% of spinal AVMs
  - AVMs account for < 10% of spinal masses

Natural History & Prognosis

- Type 4a: Generally treated with surgery with good outcome (> 90% obliteration rate)
- Type 4c: Poorer prognosis with lower treatment obliteration rate (~ 70%)

Treatment

- Type 4: Embolization or surgical resection based upon anatomy and size: Surgical resection, surgical resection or embolization, embolization
  - Type 4-A tends to surgical resection since normal caliber ASA is inaccessible to embolization
  - Type 4-B have dilated feeding vessels allowing embolization, or surgery
  - Type 4-C have high flow state, are most amenable to embolization

DIAGNOSTIC CHECKLIST

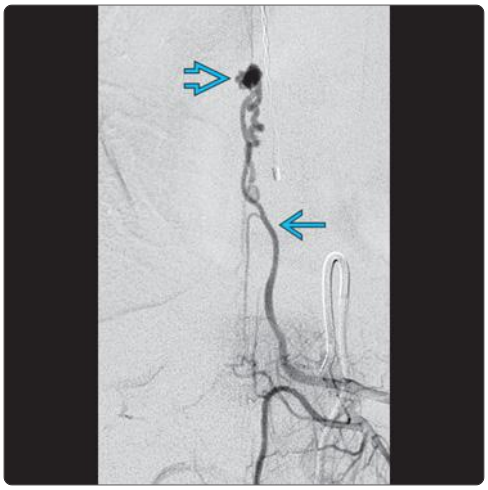
Consider

- CTA or dynamic enhanced MRA as localization tool prior to catheter angiography
  - Can define malformation type, location of ASA

SELECTED REFERENCES

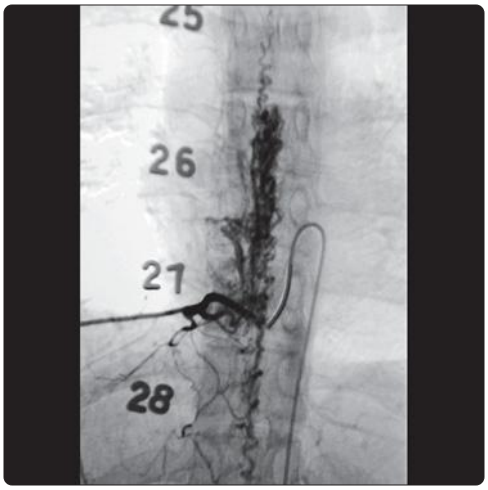
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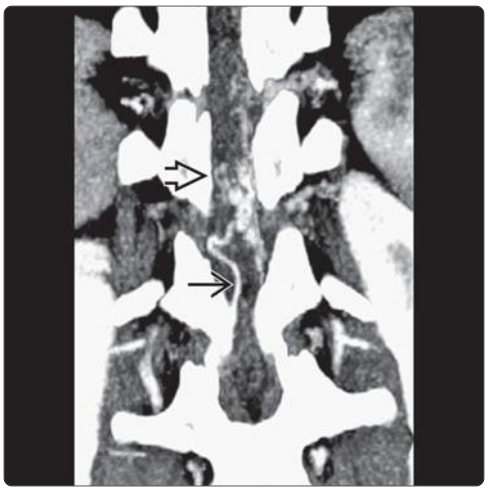


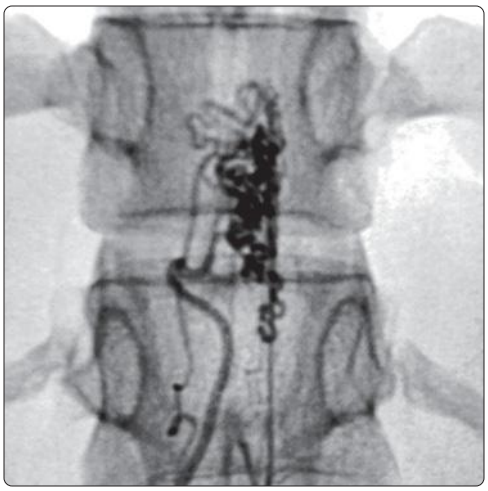
(Left) *Sagittal STIR MR shows a long segment of cord hyperintensity with ill-defined flow voids along the ventral cord surface. There is a more focal area of low signal, which at angiography was a high-flow aneurysm* ⇨. **(Right)** *AP view of a catheter angiogram (arterial phase) shows a prominent segmental feeder* ⇨ *to the cord, which shunts into the coronal venous plexus, with no intervening nidus. There is high-flow aneurysm at the fistula site* ⇨.





(Left) *Sagittal T1 MR shows multiple serpentine flow voids dorsal to the thoracic cord from the intradural type 4 fistula.* **(Right)** *Anteroposterior angiogram shows the type 4 fistula fed by right radicular artery, with multiple dilated perimedullary veins.*





(Left) *Coronal CTA shows a large tangle of vessels along the ventral cord surface fed by the right radicular artery* ⇨ *with dilated veins on the cord* ⇨. **(Right)** *Anteroposterior angiography shows a vascular malformation supplied by the radicular artery on the pial surface of cord. Shunting is at the cord level (type 4), not at the dural level (type 1).*

Conus Arteriovenous Malformation

KEY FACTS

TERMINOLOGY

- Spinal vascular lesions with angioarchitectural hallmarks
  - Dual arterial supply from anterior spinal arteries (ASA) and posterior spinal arteries (PSA)
  - Multiple glomus-type nidi
  - Complex venous drainage at level of conus medullaris

IMAGING

- Multiple flow voids surrounding distal thoracic cord on MR
- May be indistinguishable on MR from type I or IV spinal fistulas
- Multiple enlarged feeding vessels with relatively fast shunting, including ASA
- Multiple fistulous points, which may show aneurysmal dilatation
- Multiple glomus-type nidi that may be intramedullary or extramedullary
- Catheter angiography for definitive diagnosis

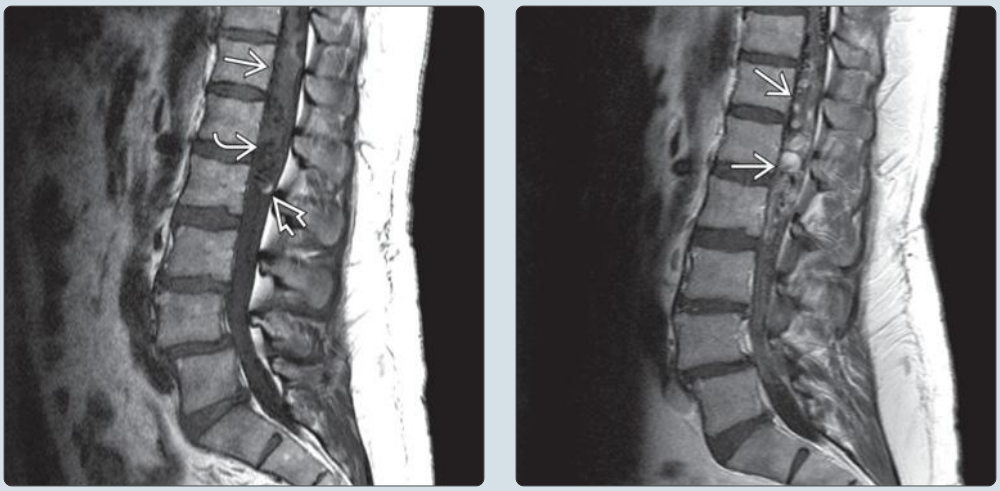
TOP DIFFERENTIAL DIAGNOSES

- Normal CSF flow artifact
- Type 1 dural fistula
- Type 4 pial fistula
- Lumbar canal stenosis with tortuous intradural roots
- Intradural extramedullary tumor with prominent vascular supply

CLINICAL ISSUES

- Progressive myelopathy, radiculopathy, or myeloradiculopathy
  - Subarachnoid hemorrhage (30% at presentation)
- Embolization followed by microsurgical resection
- Microsurgical resection only

(Left) *Sagittal T1 MR shows loss of normal morphology surrounding the conus with ill-defined flow voids. A small focus of high signal could reflect thrombus within a varix or be flow-related enhancement.* **(Right)** *Sagittal T1 C+ MR shows marked enhancement of multiple dilated and serpentine vessels filling the thecal sac and obscuring the underlying conus and cauda equina.*



(Left) *Sagittal STIR MR shows flow voids within multiple enlarged intradural vessels surrounding the distal spinal cord. These vessels are a combination of enlarged feeding arteries and dilated draining veins.* **(Right)** *Sagittal T1 C+ FS MR shows marked enhancement of extensive draining veins from the conus arteriovenous malformation extending superiorly to the midthoracic level.*



TERMINOLOGY

Definitions

- Spinal vascular lesions with angioarchitectural hallmarks of (1) dual arterial supply from anterior and posterior spinal arteries, (2) multiple glomus-type nidi, and (3) complex venous drainage at level of conus medullaris

IMAGING

General Features

- Best diagnostic clue
  - Multiple flow voids surrounding distal thoracic cord on MR
  - May be indistinguishable on MR from type I or IV spinal fistulas
- Location
  - Conus
- Size
  - Variable, depending upon number and size of feeding vessels
- Morphology
  - Multiple serpentine enlarged vessels surrounding conus related to multiple nidi/fistulas

MR Findings

- T1WI
  - Multiple serpentine intradural flow voids
  - Little or no cord parenchymal flow voids
  - CSF may show subarachnoid hemorrhage
- T2WI
  - Intradural extramedullary flow voids
  - Cord edema
- T1WI C+
  - May show patchy enhancement of cord due to venous ischemia
- MRA
  - Dynamic enhanced MRA will confirm vascular nature of flow voids
  - Multiple feeding vessels may be difficult to define by MRA

Angiographic Findings

- Multiple enlarged feeding vessels with relatively fast shunting, including ASA
- Multiple fistulous points, which may show aneurysmal dilatation
- Multiple glomus-type nidi that may be intramedullary or extramedullary
- Complex venous drainage

Imaging Recommendations

- Best imaging tool
  - Screening with MR to show abnormal flow voids and point to spinal vascular malformation
  - Catheter angiography for definitive diagnosis

DIFFERENTIAL DIAGNOSIS

Normal CSF Flow Artifact

- Most prominent dorsal to thoracic cord

Type 1 Dural Fistula

- Fistula at level of neural foramen via radicular artery

Type 4 Pial Fistula

- May be indistinguishable on MR, MRA

Lumbar Canal Stenosis With Tortuous Intradural Roots

- Look for central canal stenosis with redundant roots cephalad

Intradural Extramedullary Tumor With Prominent Vascular Supply

- Paraganglioma: Markedly enhancing intradural extramedullary mass

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Progressive myelopathy, radiculopathy, or myeloradiculopathy
    - Caused by venous hypertension, ischemia, or local mass effect
- Other signs/symptoms
  - Subarachnoid hemorrhage (30% at presentation)
  - Bowel or bladder dysfunction (50%)

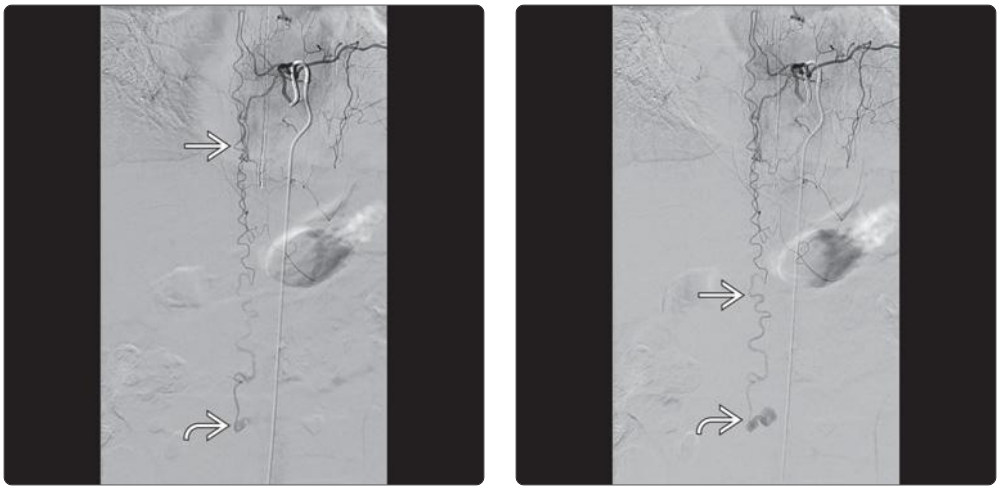
Treatment

- Embolization followed by microsurgical resection
- Microsurgical resection only
  - Obliteration of vascular malformation in ~ 80%

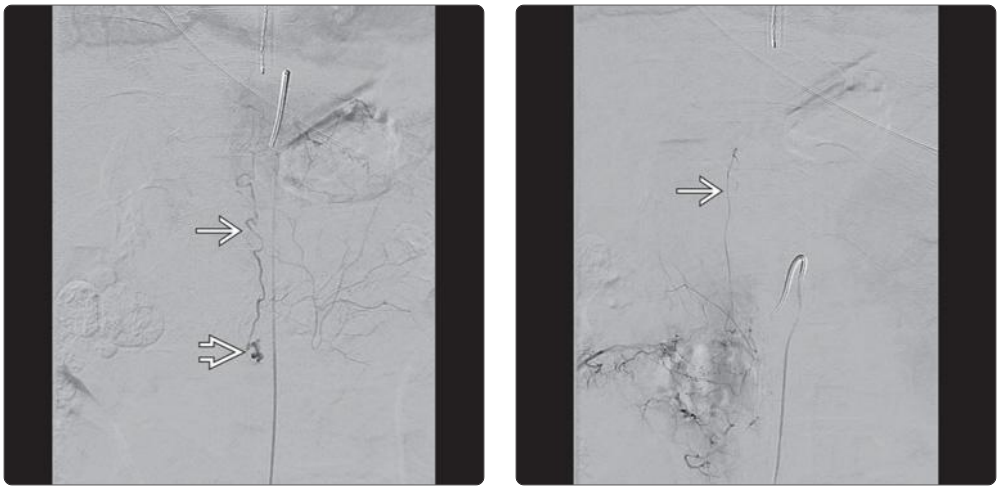
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(Left) *AP spinal angiogram view showing 1 of 4 feeding vessels to a large conus arteriovenous malformation (AVM). A catheter is injecting the right T7 intercostal with an enlarged ASA ➡ showing a fistula with a dilated vein ➡.* **(Right)** *AP angiogram showing 1 of 4 feeding vessels to a large conus AVM. A slightly later phase shows the catheter at the right T7 intercostal with an enlarged ASA ➡ and fistula with dilated draining vein ➡.*



(Left) *AP angiogram showing 2 of 4 feeding vessels to a large conus AVM. A catheter is injecting the right T10 intercostal with an enlarged spinal artery ➡ showing a small vascular nidus ➡.* **(Right)** *AP angiogram showing 3 of 4 feeding vessels to a large conus AVM. A catheter is injecting the right L1 lumbar artery with a small feeding artery ➡ ascending towards the conus AVM.*



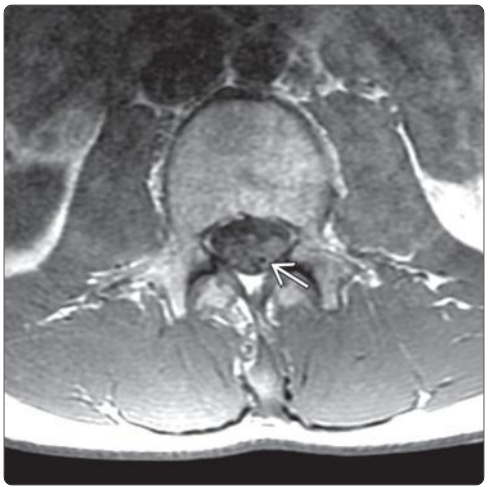
(Left) *AP angiogram showing 4 of 4 feeding vessels to a large conus AVM. A catheter is injecting the left L2 lumbar artery with an enlarged ASA ➡ showing a fistula ➡ with a dilated draining vein and varix.* **(Right)** *AP angiogram showing 4 of 4 feeding vessels to a large conus AVM. A catheter is injecting the left L2 lumbar artery. This venous phase shows the markedly dilated draining veins ➡ with varix formation ➡.*

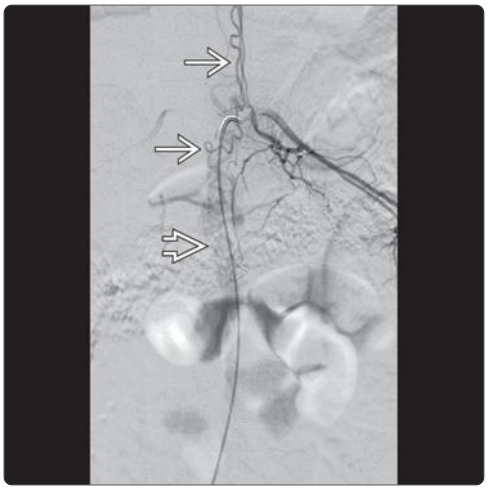




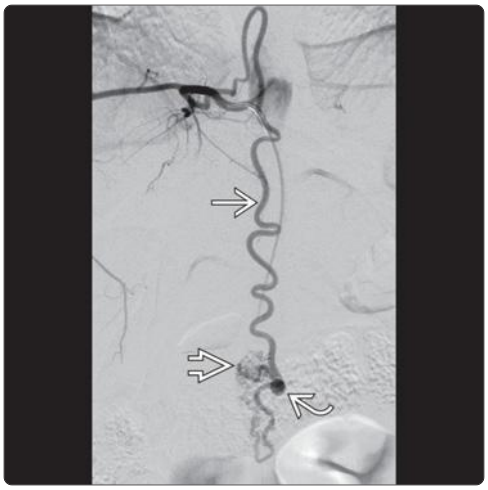


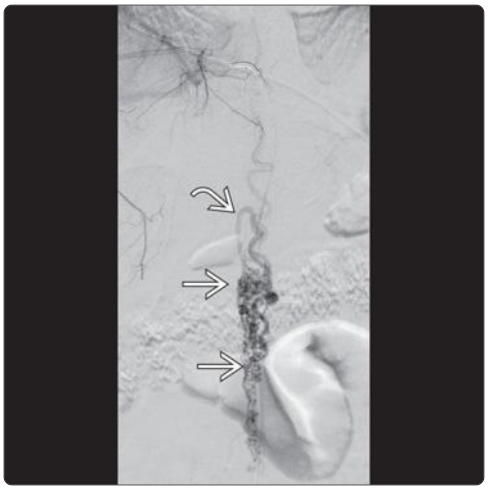
(Left) *Sagittal T1 MR shows apparent thickening or clumping of the cauda equina due to the presence of subarachnoid hemorrhage and intrathecal clot. Scattered abnormal flow voids are also present.* **(Right)** *Sagittal T2 MR shows extensive and prominent flow voids within the thecal sac in this patient with a large conus malformation. The vessels identified are primarily venous, with dilated and arterialized draining vessels.*





(Left) *Axial T1 MR shows an abnormal flow void within the cauda equina, which nerves appear thickened due to adjacent subarachnoid hemorrhage.* **(Right)** *AP spinal angiogram view showing 1 of 2 feeding vessels to a large conus AVM. A catheter is injecting the left T12 intercostal, which shows an enlarged spinal artery and early venous opacification due to AVM shunting.*





(Left) *AP spinal angiogram view showing 2 of 2 feeding vessels to a large conus AVM. A catheter is injecting the right T10 intercostal with filling of an enlarged ASA and vascular nidus. An intranidal aneurysm is present.* **(Right)** *AP spinal angiogram view showing 2 of 2 feeding vessels to a large conus AVM. A catheter is injecting the right T10 intercostal. This later phase shows the extent of the vascular nidus and a portion of the enlarged draining veins.*

Posterior Fossa Dural Fistula With Intraspinal Drainage

KEY FACTS

TERMINOLOGY

- Dural arteriovenous fistula (DAVF): Anomalous connection between dural arteries and venous sinuses &/or cortical veins without presence of normal intervening capillary bed
- Rare DAVF variant has preferential venous drainage inferiorly into spinal veins

IMAGING

- Increased T2 signal in upper cervical cord/medulla with abnormal perimedullary flow voids
- Fistula located in posterior fossa, with symptoms related to site of venous drainage (myelopathy)
- Catheter angiography remains gold standard

TOP DIFFERENTIAL DIAGNOSES

- Spinal DAVF
  - Cord signal change and flow voids may look identical to posterior fossa DAVF with intraspinal drainage
- Demyelinating disease
  - Cord enhancement and increased T2 signal but no abnormal perimedullary flow voids

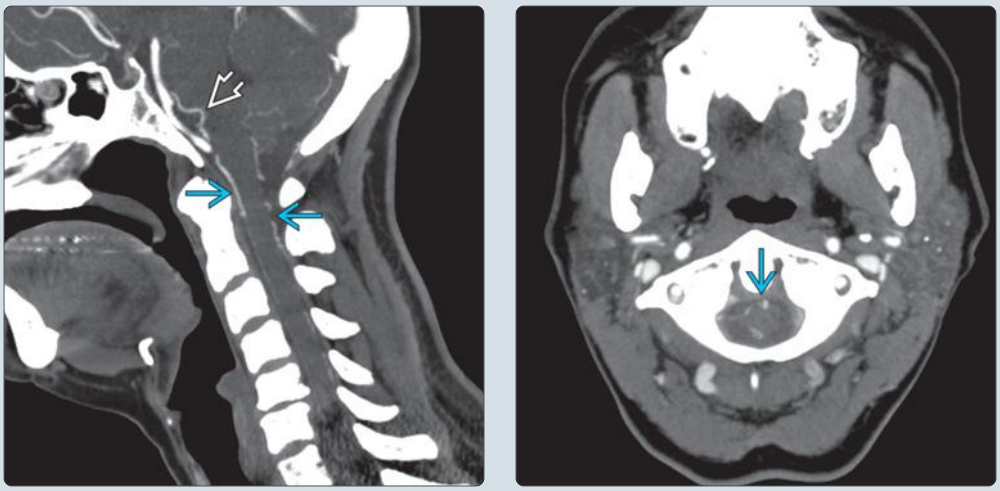
CLINICAL ISSUES

- Symptoms tend to be chronic and progressive
- Good prognosis for improvement of symptoms or complete recovery

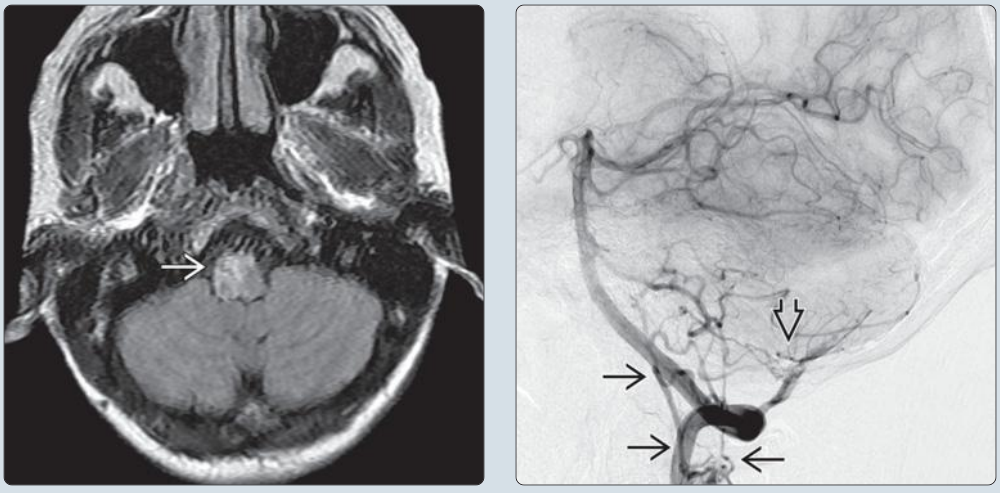
DIAGNOSTIC CHECKLIST

- Catheter angiography still required for exclusion of DAVF even if negative MRA
- DAVF clinical manifestations related to anatomical distribution of draining veins, not the fistula site
- Cognard type V fistula presenting symptoms relate to dysfunction of spinal cord rather than brain

(Left) *CTA shows a variant AV fistula off the right PICA with drainage into spinal veins that produces venous hypertension and subsequent T2 hyperintensity within the medulla. Prominent veins on both the dorsal and ventral surface of the upper cervical cord, as well as a prominent median medullary vein, are well seen.* **(Right)** *CTA shows prominent veins on both the dorsal and ventral surface of the upper cervical cord. These vessels could be normal if no other imaging abnormality was found.*



(Left) *Axial FLAIR shows abnormal T2 hyperintensity within the right side of the medulla. Diffusion study was normal (not shown). The abnormality within the medulla, coupled with the prominent veins, prompted the recommendation for a catheter angiogram to exclude a dural fistula.* **(Right)** *Catheter angiogram shows a pial fistula arising off the distal right PICA with shunting into the medullary and spinal veins.*



TERMINOLOGY

Synonyms

- Intracranial dural arteriovenous fistula (DAVF) with spinal perimedullary venous drainage, Cognard type V fistula

Definitions

- DAVF: Anomalous connection between dural arteries and venous sinuses &/or cortical veins without presence of normal intervening capillary bed
  - Rare DAVF variant has preferential venous drainage inferiorly into spinal veins

IMAGING

General Features

- Best diagnostic clue
  - Increased T2 signal in upper cervical cord/medulla with abnormal perimedullary flow voids
- Location
  - Fistula located in posterior fossa, with symptoms related to site of venous drainage (myelopathy)
- Size
  - Variable, from small focus of increased T2 signal in medulla to extensive cervical cord edema

CT Findings

- CTA
  - Increased number and size of leptomeningeal vessels within posterior fossa and surrounding cord
  - Venous ectasias/aneurysms

MR Findings

- T2WI
  - Increased signal in medulla &/or upper cervical cord
  - Perimedullary flow voids (variable)
- T1WI C+
  - Patchy diffuse enhancement of medulla or upper cervical cord

Imaging Recommendations

- Best imaging tool
  - Catheter angiography remains gold standard

DIFFERENTIAL DIAGNOSIS

Spinal DAVF

- Cervical location for spinal dural fistula uncommon
- Cord signal change and flow voids may look identical to posterior fossa DAVF with intraspinal drainage

Demyelinating Disease

- Cord enhancement and increased T2 signal but no abnormal perimedullary flow voids

PATHOLOGY

General Features

- Etiology
  - DAVF in cases with intraspinal drainage is usually located in dura of posterior fossa

Staging, Grading, & Classification

- Presence of cortical venous drainage (Cognard IIb–V) indicates more aggressive lesion
  - Associated with annual mortality rate of up to 10.4%
  - Hemorrhage rate of up to 8.1%
  - Risk of nonhemorrhagic neurological deficit of up to 6.9%

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Myelopathy related to venous hypertension secondary to AV shunting and insufficient venous drainage of spinal cord
  - Symptoms tend to be chronic and progressive
    - 25% have acute onset of neurological disorder

Treatment

- Occlusion/resection of fistula by endovascular means or open surgery
- Good prognosis for improvement of symptoms or complete recovery

DIAGNOSTIC CHECKLIST

Consider

- DAVF clinical manifestations related to anatomical distribution of draining veins, not fistula site
  - Cognard type V fistula presenting symptoms relate to dysfunction of spinal cord rather than brain
- Catheter angiography still required for exclusion of DAVF even if negative MRA

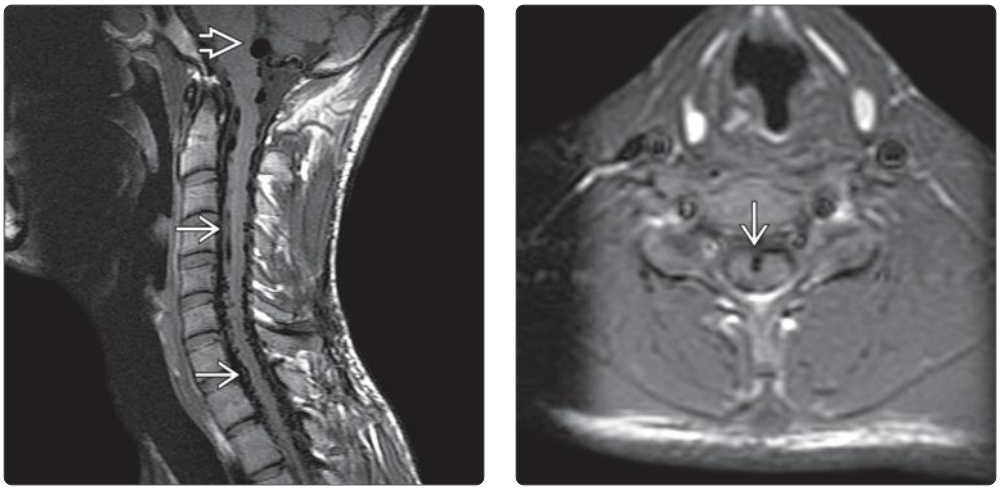
Image Interpretation Pearls

- Look for abnormal flow voids in posterior fossa when edema present in upper cervical cord

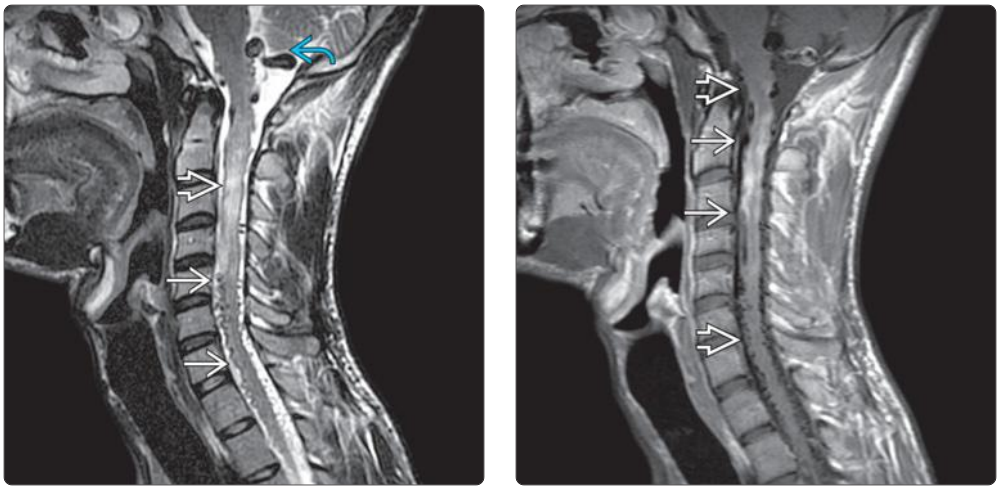
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(Left) *Sagittal T1WI shows multiple serpentine flow voids adjacent to the cervical cord. Prominent flow voids also present within the posterior fossa with venous ectasia.* **(Right)** *Axial T1WI shows an abnormally large midline ventral flow void adjacent to the cervical cord.*



(Left) *Sagittal T2WI shows extensive cord hyperintensity and mild fusiform cord expansion from C1 to C4. There are multiple abnormal pronounced perimedullary flow voids. Dilated draining veins are present in the posterior fossa.* **(Right)** *Following contrast administration, sagittal T1WI shows diffuse enhancement of the upper cervical cord from the cervicomedullary junction to C5. Multiple intradural extramedullary flow voids are present.*



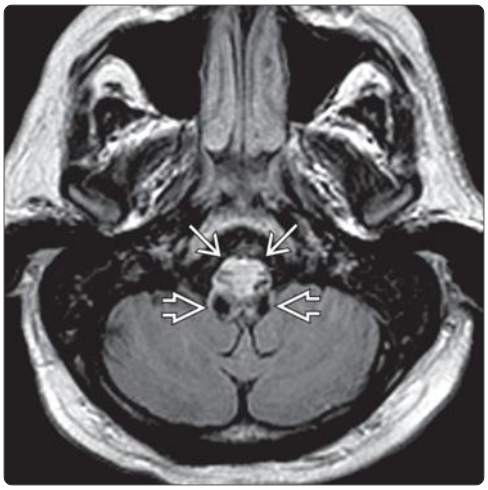
(Left) *Arterial phase of a left vertebral catheter angiogram shows a dural fistula shunt supplied by posterior meningeal branches.* **(Right)** *The venous phase of a left vertebral injection shows the posterior fossa dural fistula site with extensive intraspinal draining veins both dorsal and ventral to the cord.*

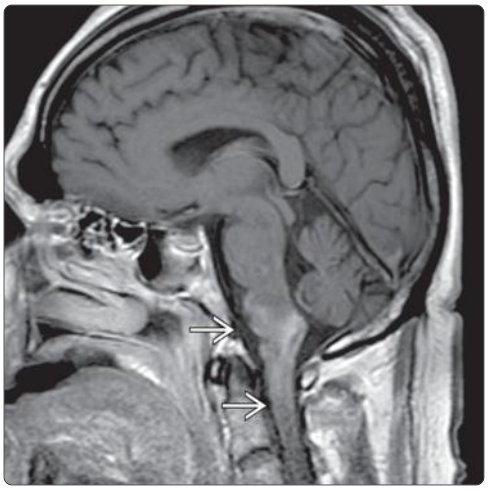




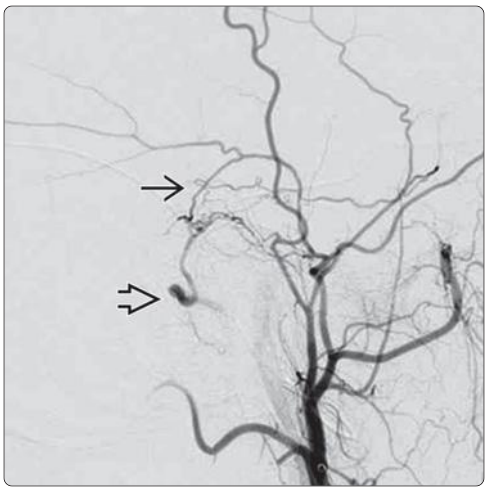


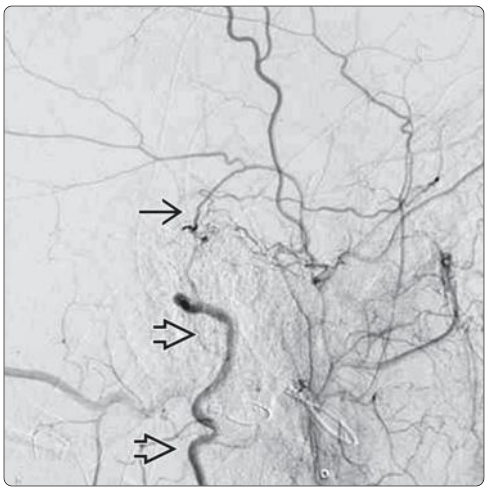
(Left) *Sagittal T1WI shows a posterior fossa tentorial dural fistula with intraspinal venous drainage and cord venous hypertension. Abnormal serpentine flow voids along the cord surface ⇒ are seen as the only clue that vascular lesion is present.* **(Right)** *Sagittal T2WI shows diffuse medullary and upper cervical cord T2 hyperintensity ⇒.*





(Left) *Axial FLAIR brain MR confirms the brainstem abnormality ⇒, with hemosiderin staining of the lower medullary nuclei ⇒, presumably from old hemorrhage.* **(Right)** *Sagittal T1WI C+ shows diffuse medullary and upper cervical cord contrast enhancement ⇒. This diffuse type of enhancement pattern with ill-defined margins over a long segment is unusual for a neoplasm.*





(Left) *Catheter angiogram injection of the external carotid artery shows the fistula feed in part by the middle meningeal artery ⇨, with venous shunting ⇨. Cervical cord myelopathy can occur due to venous hypertension from an intracranial fistula.* **(Right)** *Late arterial phase from an external carotid catheter angiogram shows the fistula feed in part by the middle meningeal artery ⇨, with venous shunting ⇨ that drains into the spinal canal.*

Cavernous Malformation

KEY FACTS

TERMINOLOGY

- Vascular lesion with lobulated, thin, sinusoidal vascular channels and no interspersed neural tissue

IMAGING

- Locules of blood with fluid-fluid levels surrounded by very T2-hypointense rim
- Spinal cord uncommon site: 3-5% of all cavernous malformations (CMs)
- Round heterogeneous signal abnormality, well-defined margins
- Brain MR to identify supratentorial lesions

TOP DIFFERENTIAL DIAGNOSES

- Intramedullary neoplasm
- Arteriovenous malformation (AVM)
- Multiple sclerosis

PATHOLOGY

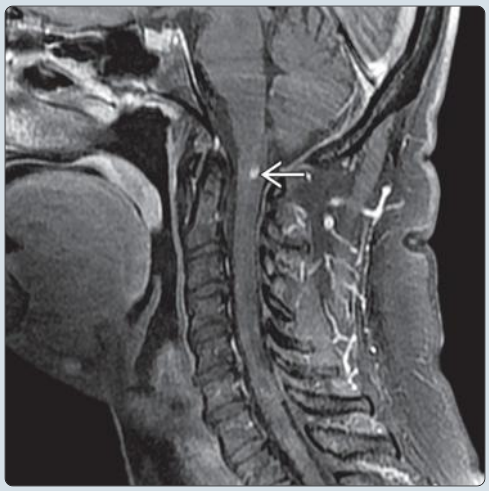
- Cervical (40%), thoracic (50%)
  - Discrete, lobulated blue-reddish brown (mulberry-like) nodule
  - Vascular spaces with single layer of endothelial cells
  - No intervening neural tissue between vascular spaces
- Multiple (familial) CM syndrome (20%)
  - Familial CMs are at high risk for hemorrhage and formation of new lesions
  - Mutations in 3 genes (*CCM1*, *CCM2*, *CCM3*) implicated in familial forms

CLINICAL ISSUES

- Typically occurs in 3rd-6th decades
- M:F = 1:2
- Broad range of dynamic behavior (may progress, enlarge, or regress)
- Surgical resection is mainstay of symptomatic CM
- Variable in outcomes literature: 50-66% improve, 28% stabilize, and 6% deteriorate postoperatively

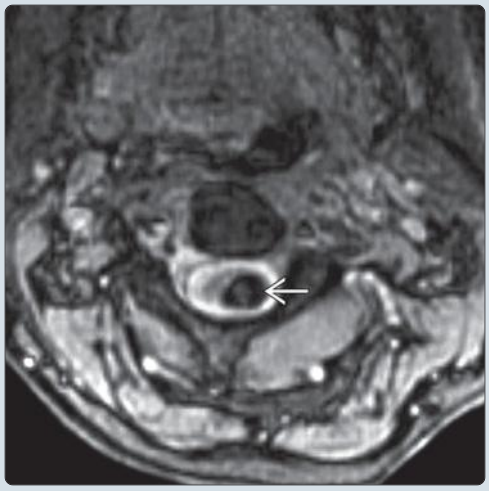
(Left) *Sagittal T2WI MR shows a small heterogeneous intramedullary lesion ➡ at the C1 level. There is slight cord expansion but no adjacent edema. Mass effect and intrinsic cord edema generally occur with more acute hemorrhage.* **(Right)** *Sagittal T1 C+ FS MR in the same patient demonstrates minimal enhancement ➡ of the CM. Acute neurological decline occurs secondary to hemorrhage. Chronic progressive myelopathy occurs due to microhemorrhages and resulting gliotic reaction to blood products.*





(Left) *Sagittal T2WI MR depicts a classic mixed hyperintense ("popcorn" or "salt and pepper") lesion surrounded by a hypointense rim ➡ from hemosiderin. Cavernous malformation was found at surgery.* **(Right)** *Axial T2* GRE MR exhibits a cavernous malformation in the cervical cord as a large area of low signal from susceptibility artifact ➡ from the blood by-products. There should be blooming of this type of lesion with longer echo times.*





TERMINOLOGY

Abbreviations

- Cavernous malformation (CM)

Synonyms

- Cavernous angioma, cavernous hemangioma, "cryptic" vascular malformation

Definitions

- Vascular lesion with lobulated, thin sinusoidal vascular channels and no interspersed neural tissue

IMAGING

General Features

- Best diagnostic clue
  - Locules of blood with fluid-fluid levels surrounded by T2-hypointense rim
- Location
  - Spinal cord uncommon site: 3-5% of all cavernous malformations
- Size
  - Few millimeters (punctate to > 1 cm)
- Morphology
  - Round heterogeneous signal abnormality, well-defined margins

CT Findings

- NECT
  - Often normal; cord may appear widened

MR Findings

- T1WI
  - Heterogeneity due to blood products of varying ages
  - Typical speckled "popcorn" heterogeneous signal
  - Small lesion may not show obvious heterogeneous signal
- T2WI
  - Heterogeneous, hypointense hemosiderin rim
  - Small lesion may not show obvious heterogeneous signal, may show only focal low signal
  - Typically no edema unless recent hemorrhage
- T2* GRE
  - Prominent "blooming" due to susceptibility effects
- T1WI C+
  - Enhancement absent/minimal

Angiographic Findings

- Conventional
  - Typically negative (one of "angiographically occult" vascular malformations)
  - Flat-panel angiotomography during spinal angiogram capable of defining associated developmental venous anomalies

Imaging Recommendations

- MR spine (use gradient-echo, contrast sequences to exclude other etiologies)
- MR brain to identify supratentorial lesions

DIFFERENTIAL DIAGNOSIS

Intramedullary Neoplasm

- Ependymoma: Enhancing mass with cysts, blood products; flow voids are rare
- Astrocytoma: Multisegmental enhancing mass (hemorrhage uncommon)
- Hemangioblastoma: Vascular nodule; "flow voids" are common
- Metastasis: Enhancing mass with edema, hemorrhage

Arteriovenous Malformation (AVM)

- Enlarged flow voids with vascular nidus
- DSA reveals enlarged feeding arteries, nidus, early draining veins

Multiple Sclerosis

- Clinical course of spinal cavernous malformations resembles multiple sclerosis with chronicity, exacerbations
- No blood by-products/hemosiderin in multiple sclerosis

PATHOLOGY

General Features

- Etiology
  - Angiogenically immature lesions with endothelial proliferation, increased neoangiogenesis
    - VEGF, β-FGF, and TGF-α are expressed
- Genetics
  - Multiple (familial) CM syndrome (20%)
    - Autosomal dominant, variable penetrance
    - Mutation in chromosomes 3, 7q (Hispanic Americans)
    - Mutations in 3 genes (*CCM1*, *CCM2*, *CCM3*) implicated in familial forms
      - *CCM1* = *KRIT1*, *CCM3* =*PDCD10*
      - Loss of *CCM1*: Insufficient angiogenesis due to incomplete cell adhesion can lead to development of CM
  - Familial CMs at high risk for hemorrhage and formation of new lesions
    - Familial characterized by higher frequency multiple CMs, infratentorial location, lower age at clinical presentation
- Identical to intracranial cavernous malformations
- Location
  - Thoracic (50%), cervical (40%), conus (10%)
  - Rare reports of intradural extramedullary or extradural CM
- 9-35% of CMs occur in brainstem
- Multiple spinal cord CMs are extremely rare
  - 1 case report involving *CCM3* (*PDCD10* gene) mutation
- Few case reports of multiple CMs in patients with Klippel-Trenaunay-Weber syndrome (KTWS)
  - *RASA1* mutation in KTWS
  - *KRIT1* and *RASA1* interact with Rap-1A protein, which is guanosine triphosphatase signaling cellular adhesion molecule

Staging, Grading, & Classification

- Type 1: Subacute hemorrhage
- Type 2: Mixed subacute and chronic; classic "popcorn" lesion

- Type 3: Chronic
- Type 4: Small punctate hypointensity on GRE images

Gross Pathologic & Surgical Features

- Discrete, lobulated, blue-reddish brown (mulberry-like) nodule
- Pseudocapsule (gliotic, hemosiderin-stained cord)

Microscopic Features

- Vascular spaces with single layer of endothelial cells with gaps between them, abnormal basal lamina
- Vascular spaces separated by collagenous walls without smooth muscle or elastin, no ensheathing pericytes or astrocytic processes
- Histologic features of arteries, capillaries, and veins generally lacking
- No intervening neural tissue between vascular spaces
  - Ca++ are rare (common in brain cavernous malformations)
  - Blood products in different stages of evolution
  - Areas of partial and complete thrombosis
- No true surrounding capsule
  - Surrounding neural tissue shows gliosis; atrophy; hemosiderin, calcium, and iron deposits
- Some authors speculate that CM and capillary telangiectasias may represent different manifestations of same pathophysiology

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sensorimotor deficits, progressive paraparesis
  - 4 clinical patterns
    - Multiple episodes of neurological deterioration, intermittent recovery
    - Slowly progressive neurological decline
    - Sudden symptom onset with rapid decline (hours to days)
    - Mild symptoms with acute onset, gradual decline (weeks to months)
  - Extremely rare: Subarachnoid hemorrhage
  - In 1 study, infratentorial, basal ganglia, or spinal location and preexisting neurologic deficit found to be independent risk factors for unfavorable outcome
- Clinical profile
  - Young adult with sudden paraplegia

Demographics

- Age
  - Typically, 3rd-6th decades
    - Range = 12-88 years; peak = 4th decade
- Gender
  - M:F = 1:2
- Epidemiology
  - Prevalence of cerebral CM in general population is 0.5%
  - 10-30% multiple, familial
  - 70% of spinal CMs are in females
  - Cranial irradiation is suspected risk factor for cerebral CMs
    - Case report of de novo development of spinal CM following spinal axis radiation
  - Simultaneous occurrence, affecting both brain and spinal cord, is observed with varying prevalence (37-47%)

Natural History & Prognosis

- Broad range of dynamic behavior (may progress, enlarge, regress)
- De novo lesions may develop (especially in familial CM syndrome)
- Clinical course varies from slow progression to acute quadriplegia
- Bleeding rate: ~ 1.4-4.5% per lesion per year

Treatment

- Conservative management if asymptomatic
  - Follow with annual MR to assess growth
- Surgical resection for symptomatic lesions
  - Long pretreatment duration (~ 3 years)
  - Postoperative outcome related to preoperative neurologic status
  - Variable in outcomes literature: 50-66% improve, 28% stabilize, and 6% deteriorate postoperatively
  - Intraoperative electrophysiological monitoring helps to reduce risk of perioperative injury of myelon and intradural fibers
  - CO_2 laser useful for treating lesions in brainstem, thalamus, and spinal cord

DIAGNOSTIC CHECKLIST

Consider

- Other hemorrhagic lesions if no classic speckled pattern or hemosiderin margins

Image Interpretation Pearls

- Heterogeneous mass (locules of blood with "popcorn" appearance) surrounded by dark rim (hemosiderin)
- Scan brain for additional lesions!
- Screen with GRE

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(Left) *Sagittal T1WI MR shows the typical appearance of a CM of the spinal cord. A heterogeneous, slightly expansile intramedullary lesion is seen at the C2-C3 level. Faint "salt and pepper" pattern is present from repeated hemorrhages and hemosiderin deposition.* **(Right)** *Sagittal T1WI C+ MR in the same patient illustrates the faint speckled pattern of enhancement in the cord CM at C2-C3. Faint mild enhancement is typical although larger lesions may show significant enhancement.*





(Left) *Sagittal T2WI MR shows hemorrhage into the conus from a cavernous malformation. Lesion shows heterogeneous signal from prior hemorrhage, with no mass effect or adjacent cord edema.* **(Right)** *Sagittal T2* GRE MR shows focal low signal within conus, which is more evident on this sequence.*





(Left) *Sagittal T2WI MR reveals extensive acute intramedullary hemorrhage from cavernous malformation. There is extensive cord edema extending the cephalad and caudad to the intramedullary hemorrhage.* **(Right)** *Sagittal T1WI C+ MR in the same patient shows more focal "speckled" enhancement from the cavernous malformation as the underlying cause of the massive cord hemorrhage. Extensive cord hemorrhage is poorly defined on this sequence, but diffuse cord expansion is evident.*

KEY FACTS

TERMINOLOGY

- Fusiform or saccular dilatation of artery supplying spinal cord, particularly anterior spinal artery (ASA) but including radiculomedullary branches

IMAGING

- Catheter spinal angiography required for diagnosis

TOP DIFFERENTIAL DIAGNOSES

- Hemorrhage from intramedullary tumor
- Hemorrhage from spinal vascular malformation

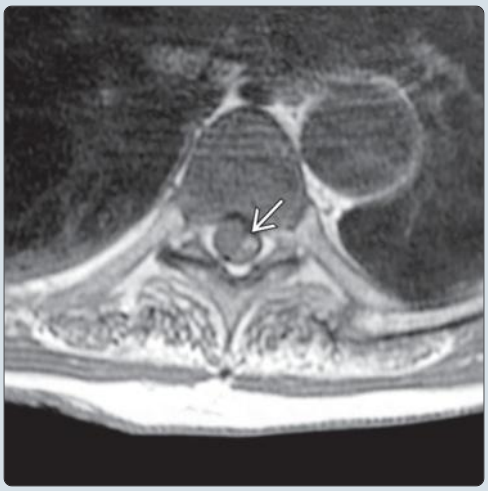
PATHOLOGY

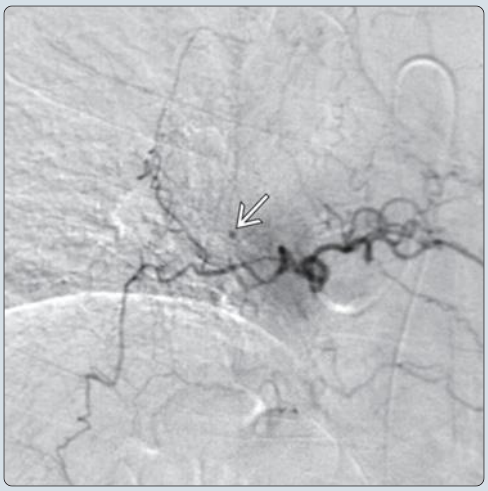
- Most commonly associated with spinal vascular malformations
- Coarctation of aorta
- Arteritis (including inflammatory etiologies, such as syphilis or mycotic)
- Fibromuscular dysplasia
- Pseudoxanthoma elasticum
- Klippel-Trenaunay-Weber syndrome
- Idiopathic
  - Tend to be fusiform in shape, without a defined neck (as seen commonly with intracranial aneurysms)
  - Often unrelated to arterial branching sites

CLINICAL ISSUES

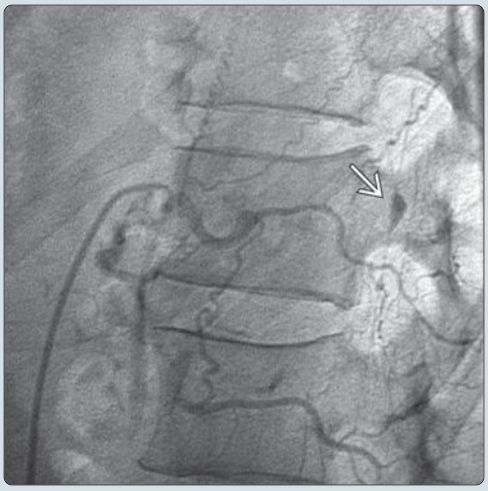
- Presenting symptoms include back pain, headache, vomiting, weakness, paraparesis, and paralysis
- Treatment may include clipping if neck is present or trapping with occlusion of parent vessel or wrapping with muslin
- Isolated reports of coil embolization of spinal aneurysms
- Spontaneous regression reported in aneurysms with inflammatory etiology

(Left) *Axial T1WI MR shows high signal intensity blood throughout the subarachnoid space ⇨ in this patient with spinal aneurysm.* **(Right)** *Anteroposterior catheter angiography with injection of left-sided intercostal artery shows filling of a small anterior spinal aneurysm ⇨.*





(Left) *Lateral catheter angiography with injection of the thoracolumbar intercostal artery shows filling of a fusiform anterior spinal artery aneurysm ⇨.* **(Right)** *Anteroposterior catheter angiography 3D spin study shows small outpouching of contrast off of the ventral medial aspect of the left vertebral artery, due to an aneurysm at the origin of the anterior spinal artery ⇨. There is a small amount of reflux extending down the right vertebral artery ⇨.*





TERMINOLOGY

Definitions

- Fusiform or saccular dilatation of artery supplying spinal cord, particularly anterior spinal artery (ASA) but including radiculomedullary branches

IMAGING

General Features

- Best diagnostic clue
  - Catheter angiographic finding of aneurysm in setting of spinal subarachnoid hemorrhage
- Location
  - Intradural extramedullary location, primarily along ventral cord surface
- Size
  - Variable, but generally small (3 mm)
- Morphology
  - Fusiform or saccular

MR Findings

- T1WI
  - Increased T1 signal from CSF due to subarachnoid hemorrhage (may be near isointense if acute)
- T2WI
  - Variable signal abnormality in CSF due to subarachnoid hemorrhage

Imaging Recommendations

- Best imaging tool
  - Catheter spinal angiography

DIFFERENTIAL DIAGNOSIS

Hemorrhage From Intramedullary Tumor

- Ependymoma

Hemorrhage From Spinal Vascular Malformation

- Types II-IV

PATHOLOGY

Staging, Grading, & Classification

- Heterogeneous group of lesions with variable anatomic and pathophysiologic characteristics
  - Most commonly associated with spinal vascular malformations
  - Coarctation of aorta
  - Arteritis (including inflammatory etiologies, such as syphilis or mycotic)
  - Fibromuscular dysplasia
  - Pseudoxanthoma elasticum
  - Klippel-Trenaunay-Weber syndrome
  - Idiopathic

Gross Pathologic & Surgical Features

- Tend to be fusiform in shape, without defined neck (as seen commonly with intracranial aneurysms)
- No definite predilection for specific site or arterial involvement
- Often unrelated to arterial branching sites
- Most frequent vessel involved is ASA
  - Aneurysms of posterior spinal, lateral spinal, and segmental arteries may also occur

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Subarachnoid hemorrhage (SAH) of spinal origin is rare event (< 1% of all SAHs)
  - Presenting symptoms include back pain, headache, vomiting, weakness, paraparesis, and paralysis
  - Symptoms may relate either to subarachnoid hemorrhage or cord compression
- Other signs/symptoms
  - Abrupt onset of back pain near universal finding with rupture

Treatment

- Variable due to ventral intraspinal position, blood supply to cord, and often fusiform shape complicating treatment
  - Clipping if neck is present
  - Trapping with occlusion of parent vessel
  - Wrapping with muslin
  - Isolated reports of coil embolization of spinal aneurysms
  - Conservative management since spontaneous regression reported in aneurysms with inflammatory etiology

DIAGNOSTIC CHECKLIST

Consider

- "Wait and see" strategy may be employed since lesions can spontaneously occlude
- Unlike intracranial aneurysms, risk of surgical treatment of ruptured aneurysm may exceed expected benefit

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Spinal Cord Infarction

KEY FACTS

TERMINOLOGY

- Spinal cord ischemia
- Cord infarction 2° to vessel occlusion (radicular artery)

IMAGING

- MR with contrast, + diffusion
- Hyperintensity on T2WI within cord; central "owl's eye" pattern
  - Slight cord expansion in acute phase
- Hyperintense central gray matter or entire cross-sectional area

TOP DIFFERENTIAL DIAGNOSES

- Multiple sclerosis
- Transverse myelitis
- ADEM/viral myelitis
- Neuromyelitis optica
- Type I dural fistula
- Spinal cord neoplasm
- Radiation myelopathy

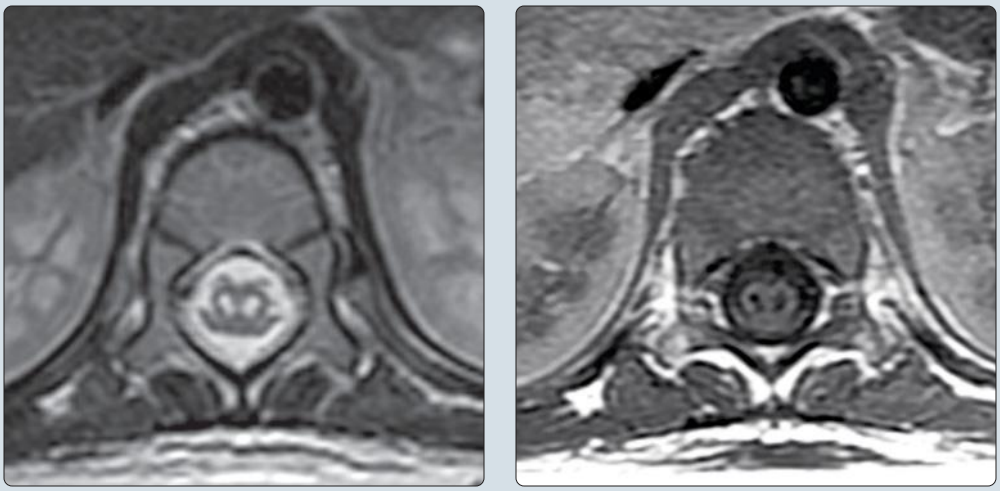
PATHOLOGY

- Up to 50% of cases have no known etiology
- Majority of known causes relate to aortic pathology
- Septicemia; systemic hypotension
- Blunt trauma with dissection
- Fibrocartilaginous embolism
- Iatrogenic; transforaminal steroid injection, selective root block

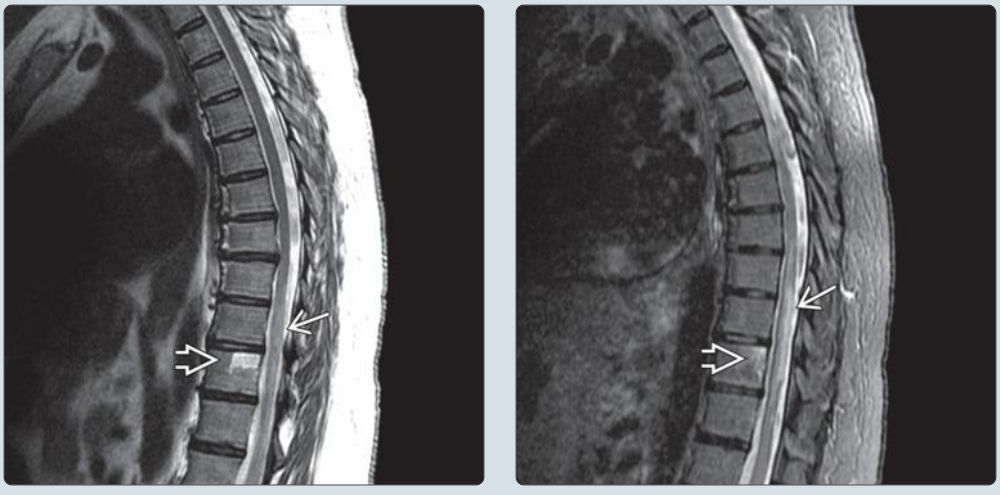
CLINICAL ISSUES

- Abrupt onset weakness, loss of sensation
- Rapid progression; maximum deficit within hours
- Poor prognosis, with permanent disabling sequelae
- Pain is frequent and disabling feature of cord infarct

(Left) *Axial T2WI MR in this case of hypoxic-ischemic encephalopathy (HIE) shows typical "owl's eye" abnormal hyperintensity within the central gray matter from chronic spinal cord infarction.* **(Right)** *Axial T1WI MR in case of severe HIE shows focal low signal within the central cord due to old infarction with myelomalacia.*



(Left) *Sagittal T2 MR shows slight hyperintensity in the distal thoracic cord in this patient with acute onset of leg weakness. There is a geographic region of increased signal in the superior aspect of the thoracic vertebral body due to concomitant cord and vertebral body infarcts.* **(Right)** *Sagittal STIR MR shows the cord hyperintensity involving the mid and dorsal aspect of the thoracic cord, along with the vertebral body infarction. The classic anterior 2/3 of the cord involved is not always present.*



TERMINOLOGY

Abbreviations

- Spinal cord infarction (SCI)
- Anterior spinal artery (ASA)

Synonyms

- Spinal cord ischemia

Definitions

- Cord infarction 2° to vessel occlusion (radicular artery)

IMAGING

General Features

- Best diagnostic clue
  - Hyperintensity on T2WI within cord; central "owl's eye" pattern
- Location
  - Distal 1/2 of thoracic cord → arterial border zone
- Size
  - Usually > 1 vertebral body segment
- Morphology
  - Central hyperintensity on T2WI, which involves central gray matter, more variable involvement of cord periphery

CT Findings

- NECT
  - Noncontributory
- CTA
  - Useful to define underlying aortic disease causing infarct, or to discover other etiologies mimicking cord infarct (dural fistula)
  - Not directly useful for identifying ASA since lack of visualization of ASA does not prove diagnosis

MR Findings

- T1WI
  - Slight cord expansion in acute phase
  - Cord atrophy as chronic finding
  - Hemorrhage conversion → hyperintense (rare)
- T2WI
  - Hyperintense central gray matter or entire cross-sectional area
- DWI
  - Hyperintense, as in brain infarcts
- MRA
  - Dynamic contrast enhanced
    - Assess other etiologies that may mimic infarct, such as dural arteriovenous fistula
    - Not directly useful for identifying ASA since lack of visualization of ASA does not prove diagnosis

Imaging Recommendations

- Best imaging tool
  - MR with contrast, + diffusion
- Protocol advice
  - T2WI sagittal & axial, DWI

DIFFERENTIAL DIAGNOSIS

Multiple Sclerosis (MS)

- Peripheral location, < 2 segments

Transverse Myelitis

- Secondary to known cause (MS, ADEM, or idiopathic [15%])
- Central, usually > 2/3 cross-sectional area of cord
- 3-4 segments, occupies > 2/3 of cord diameter
- CSF pleocytosis
- Progression of symptoms within 4 hours to 21 days

ADEM/Viral Myelitis

- Child/young adult
- Brain affected more than cord

Neuromyelitis Optica

- Longitudinally extensive (> 3 vertebral body segments) T2 hyperintensity in cord

Type I Dural Fistula

- Cord expansion and edema on T2WI
- Enlarged serpentine pial veins on cord surface

Spinal Cord Neoplasm

- Expansion, enhancement, edema, cyst

Radiation Myelopathy

- Typically with dose > 50 Gray

Parasitic or Bacterial Infection

- Focal enhancement with cord edema and enlargement

PATHOLOGY

General Features

- Etiology
  - Up to 50% of cases have no known etiology
  - Majority of known causes relate to aortic pathology
    - Atherosclerosis → dissection or emboli
    - Thoracoabdominal aneurysm
    - Aortic surgery (rates of vascular complications vary from 0.4% in coarctation repair to 12% in type IIIb dissecting aneurysms)
  - Septicemia; systemic hypotension
  - Fibrocartilaginous embolism
  - Blunt trauma with dissection
  - Penetrating trauma
  - Vasculitis; decompression sickness
  - Iatrogenic; transforaminal steroid injection, selective root block
    - Cervical radicular arteries classically travel in ventral aspect of neural foramen
    - Ascending and deep cervical branches are present in posterior foramen, which occasionally supply segmental vessels to cervical cord
- Genetics
  - None
- Associated abnormalities
  - Look for associated vertebral body infarction at general level of cord infarct
    - Thoracic level most common location for vertebral body infarcts

  - Correlated with aortic pathology
- Embryology/anatomy
  - Radicular arteries → 1 anterior, 2 posterior spinal arteries
  - Spinal cord circulation from
    - Vertebral arteries
    - Segmental vessels at multiple levels
      - Ascending cervical
      - Deep cervical
      - Intercostal
      - Lumbar
      - Sacral
  - Anterior spinal artery branches supply GM, adjacent WM mantle
    - Arises at junction of intradural segment of vertebral arteries and lies in midline of cord on ventral surface
    - Courses inferiorly to filum terminale
  - Posterior branches supply 1/3 cord periphery
    - Arise from PICA or vertebral arteries
    - Paired longitudinal channels on dorsal cord
    - Numerous anastomoses
  - Thoracolumbar region derives supply from major radiculomedullary artery (great radicular artery or artery of Adamkiewicz)
    - Usually arises on left, from T9-T12 in 75%
    - Has characteristic "hairpin" turn on MRA, DSA

Gross Pathologic & Surgical Features

- Soft, pale, swollen tissue

Microscopic Features

- Acute: Ischemic neurons with cytotoxic + vasogenic edema, swelling of endothelial cells + astrocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Anterior spinal syndrome: Paralysis, pain and temperature sensation loss, bladder and bowel dysfunction
  - Posterior spinal cord infarction: Loss of vibration sense, proprioception
  - Sudden onset
- Clinical profile
  - Abrupt onset weakness, loss of sensation
  - Rapid progression; maximum deficit within hours

Demographics

- Age
  - > 50
- Gender
  - M = F

Natural History & Prognosis

- Poor prognosis, with permanent disabling sequelae
- Initial severity of neurological deficit is best predictor of outcome
- Worse outcome when proprioceptive and walking impairment present at onset
  - 1/2 of ASA territory infarcts have some element of proprioceptive impairment
- Pain is frequent and disabling feature of cord infarct
  - Central pain more common with interruption of spinothalamic tracts
- Complications of infarction
  - Ventilator dependence
  - Decreased bowel or bladder function
  - Respiratory and urinary tract infections

Treatment

- Anticoagulation with heparin and aspirin
- Steroids
- Supportive care and rehabilitative physical therapy

DIAGNOSTIC CHECKLIST

Consider

- Look for associated vertebral body infarctions as confirmatory evidence of cord ischemia in aortic disease

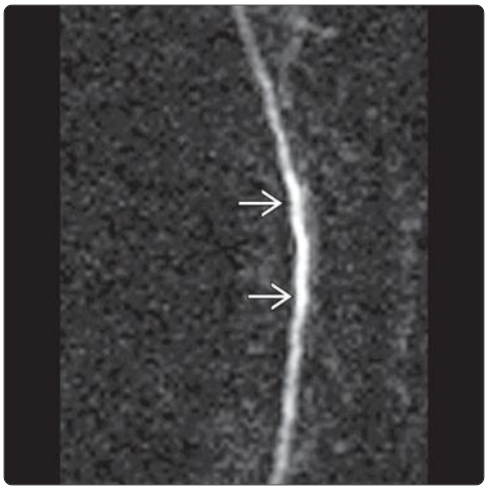
Image Interpretation Pearls

- Classic imaging appearance: T2 hyperintensity involving anterior horn cells

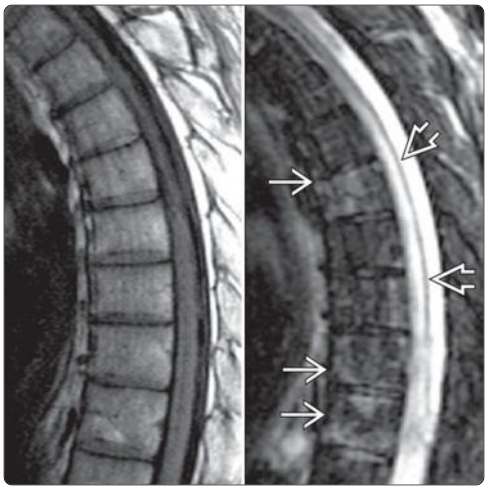
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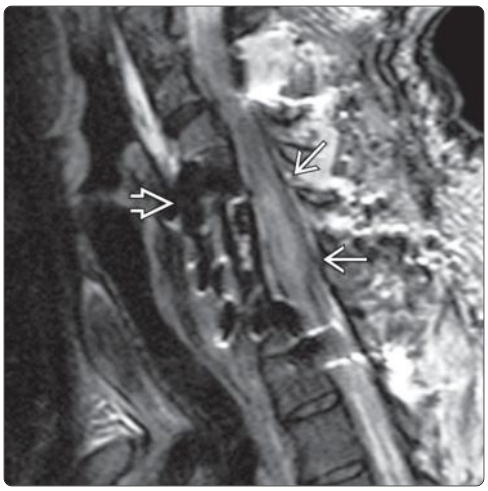


(Left) *Sagittal T2WI MR shows hyperintensity in thoracic spinal cord in this patient following vascular surgery.* **(Right)** *Sagittal DWI MR shows restricted diffusion as hyperintensity in cord due to acute infarction.*





(Left) *Sagittal T1WI MR (left side) is unremarkable. STIR MR (right side) shows hyperintense foci in thoracic bodies and cord due to bone with parenchymal infarcts.* **(Right)** *Sagittal T2WI MR shows cord infarction as complication of embolization of extensive spinal cord AVM. The medullary and cord hyperintensity is nonspecific, but the superior margin has a horizontal, well-defined transition not typical for cord edema and is much more suggestive of a vascular territory. DWI showed cord infarction.*





(Left) *Sagittal T2WI MR following multilevel corpectomy for spondylosis shows diffuse hyperintensity with enlargement of cervical cord due to infarct.* **(Right)** *Axial T2WI MR shows infarction as complication of thoracic discectomy. Cord hyperintensity present in anterior 2/3 of the cord, typical for infarction within the ASA distribution. Patient is status post postero-lateral approach for disc herniation, with unilateral pedicle screw. Presumed mechanism is occlusion of Adamkiewicz.*

Subarachnoid Hemorrhage

KEY FACTS

TERMINOLOGY

- Hemorrhage into spinal subarachnoid space from variety of etiologies
  - Trauma (> 50%)
  - Aneurysmal subarachnoid hemorrhage (SAH) with spinal extension
  - Spinal arteriovenous malformations
    - Mainly types II, III, IVc, conus malformations
  - Tumor
  - Anticoagulant therapy
  - Infection (pneumococcal meningitis, herpes)
  - Systemic disease
  - Spinal artery aneurysm (rare)

IMAGING

- Fluid-fluid level within thecal sac
- Variable depending on stage of blood breakdown and by-products
- Dynamic enhanced MRA useful as screen for spinal vascular malformation

TOP DIFFERENTIAL DIAGNOSES

- Epidural hemorrhage
- Subdural hemorrhage
- Intramedullary hemorrhage

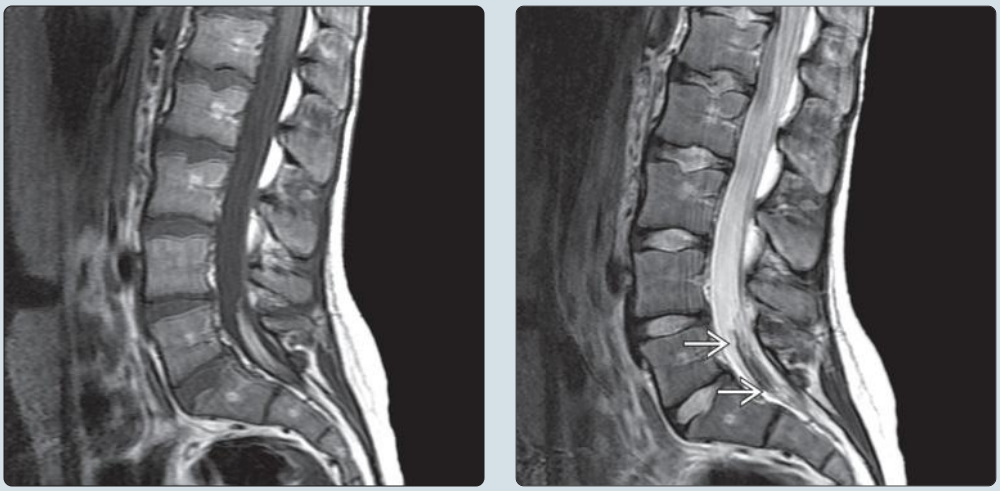
PATHOLOGY

- Rare reports of cervical/thoracic arachnoiditis developing following spinal SAH

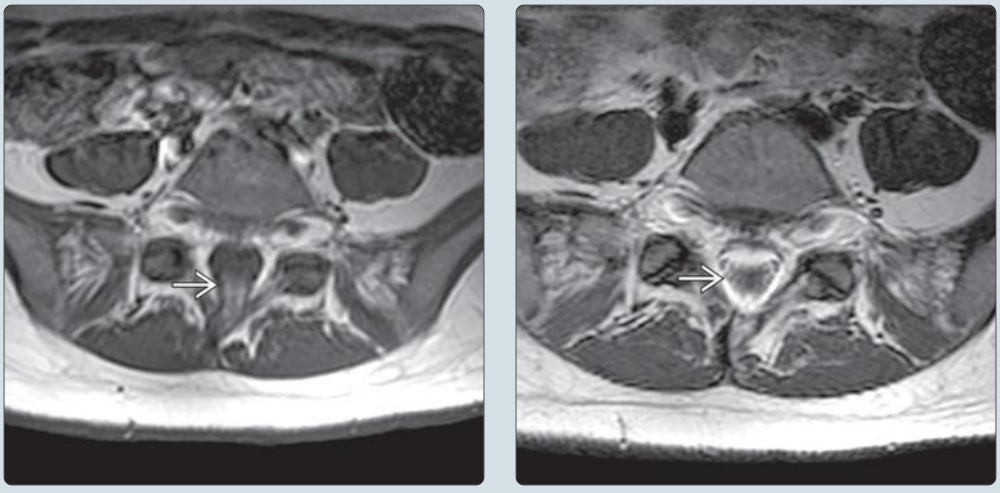
CLINICAL ISSUES

- Acute back or radicular pain ± signs of cord compression (numbness, weakness)
- Massive spinal SAH may give acute cord compression, paraplegia, fecal and urinary incontinence

(Left) *Sagittal T1WI MR of subarachnoid blood following epidural blood patch, presumably related to incorrect needle position, shows oblong focus of T1 hyperintensity within distal thecal sac at the L5-S1 level due to methemoglobin. No epidural lesion is present.* **(Right)** *Sagittal T2WI MR of subarachnoid blood following epidural blood patch, presumably related to incorrect needle position, shows blood to be of low signal. Epidural space is normal.*



(Left) *Axial T1WI MR of subarachnoid blood following epidural blood patch shows focus of T1 hyperintensity within distal thecal sac at the L5-S1 level.* **(Right)** *Axial T2WI MR shows diffuse subarachnoid blood in the distal thecal sac as hypointensity.*



TERMINOLOGY

Abbreviations

- Subarachnoid hemorrhage (SAH)

Definitions

- Hemorrhage into spinal subarachnoid space from variety of etiologies
  - Trauma (> 50%)
    - MVA, occupational, etc.
    - Postoperative
    - Lumbar puncture, epidural or intradural catheter placement
  - Related to brain pathology
    - Aneurysmal SAH with spinal extension
  - Spinal arteriovenous malformations
    - Mainly types II, III, IVc, conus malformations
    - Intradural extramedullary cavernous malformation (very rare)
  - Tumor
    - Spinal ependymoma
    - Spinal schwannoma (rare)
    - Hemangioblastoma (rare)
    - Astrocytoma (rare)
    - Endometriosis (rare)
  - Bleeding diatheses
  - Anticoagulant therapy
  - Infection (pneumococcal meningitis, herpes)
  - Systemic disease
    - Lupus erythematosus, polyarteritis nodosa
  - Spinal artery aneurysm (rare)
    - High flow varieties associated with AVM, or coarctation of aorta
    - Congenital type seen with collagen gene mutations; connective tissue disorders
  - Spontaneous idiopathic (rare)

IMAGING

General Features

- Best diagnostic clue
  - Fluid-fluid level within thecal sac
- Location
  - Intrathecal: Lumbar > thoracic > cervical
- Size
  - Variable
- Morphology
  - Variable depending on degree of clot formation; mass-like to fluid-fluid level

CT Findings

- NECT
  - High attenuation in subarachnoid space
- CECT
  - May show enhancement of underlying etiology (AVM, tumor)
- CTA
  - Useful in defining underlying spinal vascular malformation (nidus, enlarged draining veins)

MR Findings

- T1WI
  - Variable depending on stage of blood breakdown and by-products
    - Slight to pronounced increased signal within thecal sac
    - Fluid-fluid level or more focal globular clot within cauda equina
    - May show indistinct CSF/cord/root distinction within thecal sac due to diffuse SAH
- T2WI
  - Variable depending on stage of blood breakdown and by-products
- T2* GRE
  - Low signal from blood breakdown products
- T1WI C+
  - May show mild enhancement of cauda equina due to 2° inflammatory reaction
  - Look for enhancement of underlying pathology
- MRA
  - Dynamic enhanced MRA useful as screen for spinal vascular malformation (enlarged draining veins)

Angiographic Findings

- Conventional
  - Spinal angiography for elucidation of underlying pathology (spinal vascular malformation)

Imaging Recommendations

- Best imaging tool
  - MR for evaluation of underlying etiology (AVM, tumor)
- Protocol advice
  - Sagittal, axial T1WI, T2WI, gradient-echo with post-contrast T1WI
  - Scan whole spinal axis, + brain if unknown etiology

DIFFERENTIAL DIAGNOSIS

Epidural Hemorrhage

- Linear mass displacing low signal dural margin, cord
- Variable signal depending on age, may be isointense in acute phase

Subdural Hemorrhage

- Lobulated extramedullary signal abnormality, with well-preserved outer dural margin
- Axial may show lobulation as "Mercedes Benz" sign

Intramedullary Hemorrhage

- Focal increased signal within cord on T1WI
- Focal low signal on T2WI within cord
- Increase in size with gradient-echo imaging
- Look for enhancing mass separate from any T1 shortening related to methemoglobin

PATHOLOGY

General Features

- Etiology
  - Anticoagulant-induced hemorrhage
    - Any part of spine: Epidural, subdural, or subarachnoid spaces

  - May occur without antecedent trauma
  - May occur with PT, PTT; clotting times are within therapeutic range
  - Spinal presentation most commonly epidural
- Traumatic lumbar puncture (LP) vs. true SAH
  - Estimated 20% of LPs are traumatic
  - No consensus as to what constitutes traumatic tap
  - General guideline of < 1,000 cells/mm3
  - Opening pressure normal with traumatic tap, elevated opening pressure in 60% of true SAH cases
  - Traumatic tap initially bloody with clearing over 3 tubes, true SAH persistently bloody
  - Xanthochromia (RBC catabolism of hemoglobin → methemoglobin and bilirubin) with true SAH
  - Spectrophotometry positive in true SAH for hemoglobin breakdown products
  - Persistence of ↑ RBC count over multiple tubes with true SAH
  - WBC count proportional to peripheral blood with traumatic tap
  - CSF from true SAH should not clot (defibrination)
  - D-dimer positive with true SAH (problem with false-positive and negative)
- Associated abnormalities
  - Rare reports of cervical/thoracic arachnoiditis developing following spinal SAH
  - Incidence of SAH 6/100,000 people annually
  - 85% relate to aneurysm rupture
    - Nonaneurysmal perimesencephalic hemorrhage 10%
    - Primary spinal SAH 1%

Staging, Grading, & Classification

- Hunt and Hess scale for SAH
  - I: Asymptomatic
  - II: Severe headache or nuchal rigidity, no neurologic deficit
  - III: Drowsy, minimal neurologic deficit
  - IV: Stuporous, hemiparesis
  - V: Coma, decerebrate posturing

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute back or radicular pain ± signs of cord compression (numbness, weakness)
    - Spinal SAH characterized by sudden headache, acute sciatic pain, xanthochromic CSF, meningeal irritation, sensory deficit or paralysis
    - Massive spinal SAH may give acute cord compression, paraplegia, fecal and urinary incontinence

Demographics

- Age
  - Adult > children
- Gender
  - Predilection related to underlying etiology
- Ethnicity
  - Predilection related to underlying etiology
- Epidemiology
  - Trauma major cause of spinal hemorrhage
    - 10,000 new cases of spinal cord injury in USA per year

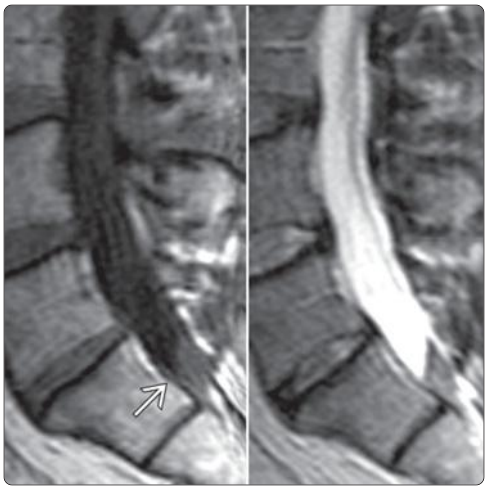
DIAGNOSTIC CHECKLIST

Consider

- CT brain > 95% sensitive to SAH during 1st 24 hours
  - Sensitivity at 1 week: 50%

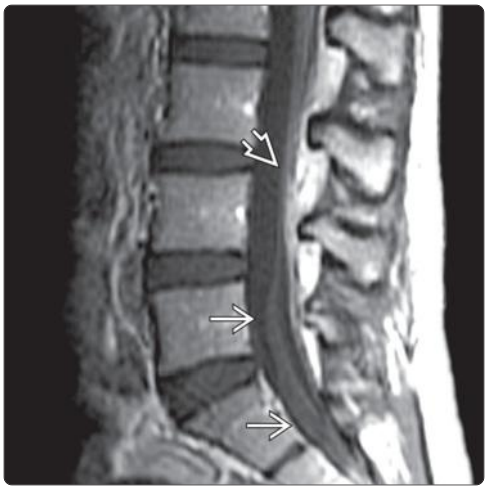
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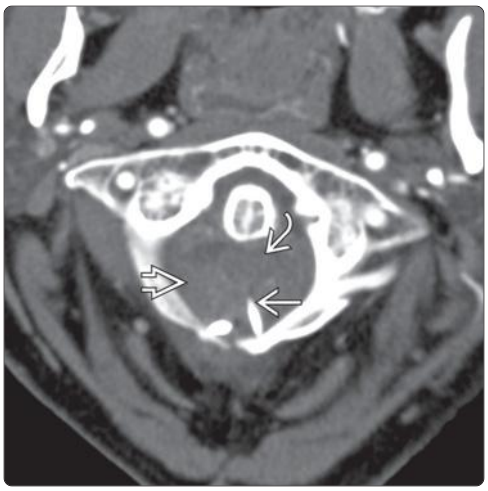


(Left) *Sagittal T1WI MR (left) and T2WI MR (right) shows CSF fluid level related to subarachnoid hemorrhage (intracranial aneurysm rupture).* **(Right)** *Sagittal T1WI C+ MR (left) shows poor discrimination of conus/cauda equina but no intradural enhancement. T2WI MR (right) shows subarachnoid hemorrhage as low signal (post lumbar catheter placement).*





(Left) *Sagittal T1WI C+ MR demonstrates variable appearance of spinal subarachnoid hemorrhage with nonenhancing clot in caudal thecal sac and mild inflammatory nerve root enhancement.* **(Right)** *Sagittal T2 MR shows the appearance of a large subarachnoid hemorrhage, which is layering within the thecal sac with a well-defined fluid-fluid level.*





(Left) *CT spine reconstructed from CTA shows posterior cerclage wires at C1-C2 with fracture of wire, which extends through dura and is adjacent to dorsal cord. There is large subarachnoid hemorrhage present with high attenuation of subarachnoid space.* **(Right)** *Axial NECT following posterior fossae tumor surgery shows extensive subarachnoid blood as intradural high attenuation. Note intrathecal postoperative gas.*

Spontaneous Epidural Hematoma

KEY FACTS

TERMINOLOGY

- Hemorrhage build-up between dura and spine not caused by significant trauma or iatrogenic procedures

IMAGING

- Extradural multisegmental T1 hyperintense fluid collection
- Thoracic, lumbar > cervical
  - Lentiform or biconvex
  - Outlined by dorsal epidural fat cranially and rostrally
- T1WI acute: < 48 hours
  - Isointense > hypo-/hyperintense
- T1WI subacute and chronic
  - Hyperintense > isointense
- T2WI heterogeneously hyperintense

TOP DIFFERENTIAL DIAGNOSES

- Epidural metastasis
- Lymphoma
- Disc extrusion, migration
- Epidural abscess
- Subdural hematoma
- Epidural fat lipomatosis

PATHOLOGY

- Idiopathic: 40-50%
- Minor trauma
- Anticoagulation
- Coagulopathy
- Disc herniation
- Vascular anomaly

CLINICAL ISSUES

- Acute onset of neck, back pain
- Progressive paraparesis
- Bladder or bowel dysfunction



Sagittal T1WI MR shows an indistinct margin of the cervical cord due to an isointense mass within the dorsal epidural space. The location of the mass is best identified at the inferior margin as extradural.



Sagittal T2WI MR shows a slightly heterogeneous, long segment hyperintense mass within the posterior epidural space spanning the C2-T4 level. The cord is severely compressed and displaced anteriorly.

TERMINOLOGY

Abbreviations

- Spontaneous spinal epidural hematoma (SSEDH)

Definitions

- Accumulation of hemorrhage between dura and spine not caused by significant trauma or iatrogenic procedures

IMAGING

General Features

- Best diagnostic clue
  - Extradural multisegmental T1 hyperintense fluid collection
- Location
  - Thoracic, lumbar > cervical
    - Cervicothoracic region more common in children
  - Dorsal > ventral > circumferential
- Size
  - Multilevel > single level
- Morphology
  - Broad-based
  - Lentiform or biconvex
  - Outlined by dorsal epidural fat cranially and rostrally

Radiographic Findings

- Radiography
  - Noncontributory

CT Findings

- NECT
  - Hyperdense epidural mass
    - Soft tissue window essential
  - Variable cord compression
  - Sagittal reformation better evaluates craniocaudal extent

MR Findings

- T1WI
  - Variable signal intensity with age of SSEDH
  - Acute: < 48 hours
    - Isointense > hypo- or hyperintense
  - Subacute and chronic
    - Hyperintense > isointense
  - Fat-suppressed T1WI to distinguish blood vs. fat
- T2WI
  - Heterogeneously hyperintense
  - Hypointense foci due to deoxyhemoglobin or fibrous septa in epidural fat
- PD/intermediate
  - Diffusely hyperintense
- T2* GRE
  - Hyperintense with prominent hypointense deoxyhemoglobin
- T1WI C+
  - Focal enhancement in acute SSEDH represents active extravasation of contrast-enhanced blood
  - Peripheral enhancement due to adjacent dural hyperemia
  - Linear enhancement may represent epidural septa or vessels
- Cord compression
  - ± cord edema

Nonvascular Interventions

- Myelography
  - Block of cerebral spinal fluid
    - Postmyelography CT confirms epidural location

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T1WI and T2WI MR
    - NECT may be helpful in confirming acute T1 isointense SSEDH with focal enhancement
    - Postmyelography CT in patients with instrumentation
  - Spinal angiography not indicated unless findings of arteriovenous malformation present

DIFFERENTIAL DIAGNOSIS

Epidural Metastasis

- Diffuse soft tissue enhancement

Lymphoma

- Well-circumscribed extraosseous soft tissue mass
  - Hypointense on T1WI
  - Hypo- to isointense on T2WI
  - Diffuse enhancement unless necrotic (unusual)
  - Often contiguous with vertebral lesion
    - Destruction of posterior vertebral body, pedicle
- Pathologic compression fractures
- Spinal column may be spared in some cases
  - Spinal epidural lymphoma

Disc Extrusion, Migration

- Associated parent disc height loss, desiccation, protrusion; anular tear
- More focal appearance
  - < 1/2 vertebral height in craniocaudal dimension
- Similar signal intensity as parent disc
  - Isointense on T1WI
  - Iso- to hypointense on T2WI
  - ± mild peripheral enhancement
- SSEDH may be associated with disc extrusion
  - Difficult to distinguish by imaging

Epidural Abscess

- More common in ventral epidural space
- Typically associated with infectious spondylodiscitis
- Iso- to hypointense on T1WI
- Hyperintense on T2WI
- Diffuse or peripheral enhancement

Subdural Hematoma

- Other than trauma, lumbar puncture in patients with coagulopathy most common cause
- More common in lumbar or thoracolumbar region
- Conforms to thecal sac
- Predominantly hypointense on T2WI or gradient-echo
  - Isointense on T1WI
- No post-contrast enhancement

Epidural Fat Lipomatosis

- Prominent epidural fat in thoracic spine normal

- Excessive epidural fat in spinal canal = lipomatosis
  - Thoracic spine most common
- Homogeneous and hyperintense on T1WI and T2WI
- Signal loss with fat suppression
- No post-contrast enhancement

PATHOLOGY

General Features

- Etiology
  - Minor trauma
    - Rupture of valveless epidural venous plexus
  - Anticoagulation
  - Coagulopathy
  - Transient venous hypertension
    - Coughing, sneezing, or other sudden Valsalva maneuvers
  - Disc herniation
    - 78% of lumbar SSEH in 1 series
    - Rupture of adjacent epidural veins
  - Vascular anomaly
  - Pregnancy
  - Paget disease
  - Idiopathic: 40-50%
- Genetics
  - No genetic predilection
- Associated abnormalities
  - Disc herniation
  - Arteriovenous malformation
- Epidural venous hemorrhage ± minimal trauma

Gross Pathologic & Surgical Features

- Isolated hematoma in epidural space

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute onset of neck, back pain
  - Radicular pain
  - Other signs/symptoms
    - Progressive paraparesis
    - Sensory deficit
    - Altered deep tendon reflexes
    - Bladder or bowel dysfunction
- Clinical profile
  - Rapid progression of neurologic impairment
  - May mimic acute disc herniation

Demographics

- Age
  - Bimodal distribution
    - Childhood
    - 50s and 60s
- Gender
  - M:F = 4:1
  - M = F in children
- Ethnicity
  - No racial predilection
- Epidemiology
  - Incidence of approximately 0.1 per 100,000 per year
  - < 1% of spinal space-occupying lesions
    - Leading cause of spinal epidural hematoma

Natural History & Prognosis

- May resolve spontaneously
- Favorable clinical and imaging outcome with conservative management
- Severity and duration of neurologic deficit predictive of postoperative neurologic recovery
  - Return to neurologic baseline after surgery
    - 89-95% of patients with incomplete neurologic deficit vs. 38-45% of patients with complete impairment
  - Improved outcome when surgery performed ≤ 36 hours in patients with complete sensorimotor loss
    - ≤ 48 hours in patients with incomplete deficit
  - Localization in lumbosacral spine or single level involvement also has favorable surgical prognosis
- Mortality rate: 6%

Treatment

- Decompressive laminectomy with hematoma evacuation
- Management
  - Correction of underlying coagulopathy with vitamin K, protamine sulfate, platelet transfusions, etc.
  - Intravenous dexamethasone to decrease inflammatory reaction and cord edema
  - In cases of mild symptoms at presentation
    - Regardless of degree of cord compression on MR
  - In cases of rapid and progressive neurologic recovery

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Biconvex epidural mass hyperintense on T1WI and T2WI with hypointense foci highly suggestive of spinal epidural hematoma

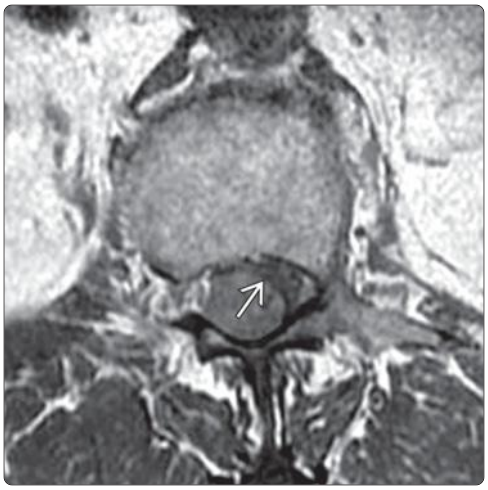
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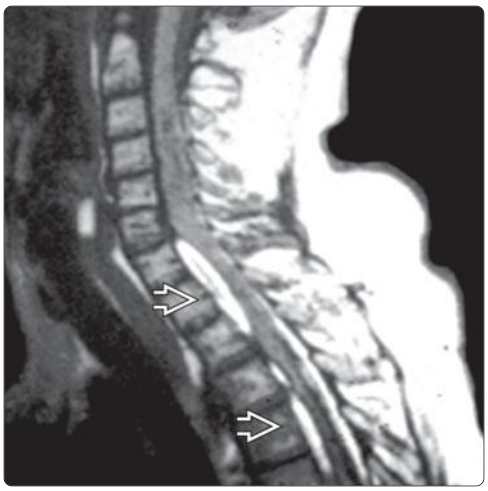


(Left) *Sagittal T1WI MR shows 2 extensive masses ⇨ that compress the distal thoracic cord and filum terminale in a patient with a history of anticoagulation. The inferior lesion is the most obviously extradural.* **(Right)** *Sagittal T2WI MR shows a larger cephalad extradural mass ➡, which is mixed iso- and hypointense, displacing ventrally the cord and filum. A smaller caudal epidural hemorrhage shows mild effacement of the thecal sac at the L3-L4 level ➡.*





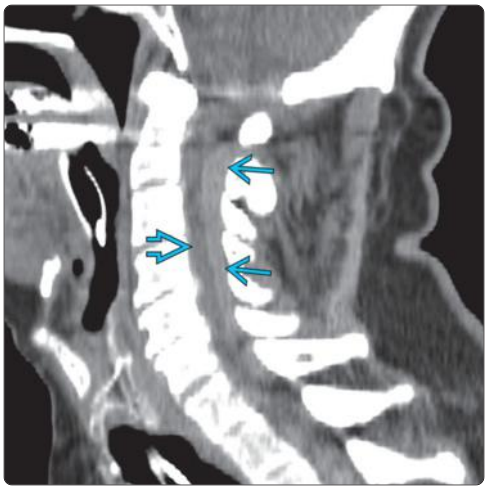
(Left) *Axial T1WI MR shows a mass that is isointense and appears to displace the dura ➡, indicating that the lesion is extradural. The mass did not enhance following contrast administration.* **(Right)** *Axial T2WI MR shows the mass ➡ is clearly extradural and severely compresses the thecal sac.*

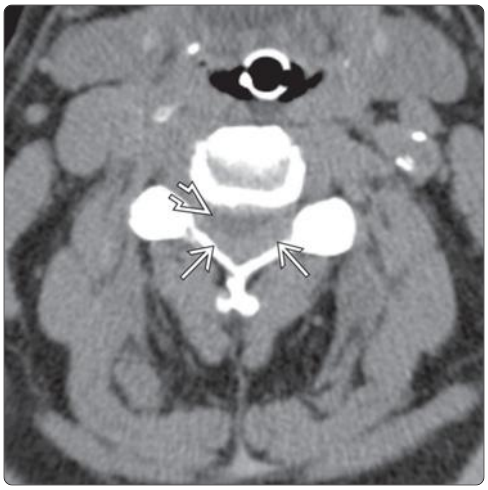




(Left) *Sagittal T1WI MR through the cervicothoracic spine shows 2 lentiform-shaped hyperintense ventral epidural hematomas ➡ with severe compression of the cord. Thoracic dorsal epidural fat is noted.* **(Right)** *Sagittal T1WI MR through the thoracic spine shows 2 subacute hyperintense SSEDHs, 1 ventral ⇨ and 1 dorsal ⇨.*

(Left) *Sagittal NECT with soft tissue window shows an extensive epidural hematoma as slightly increased attenuation in the posterior canal with displacement of the cord anteriorly.* **(Right)** *Axial NECT shows increased attenuation within the cervical epidural space from an epidural hematoma with circumferential mass effect upon the cord. Patient was on aspirin and Plavix.*





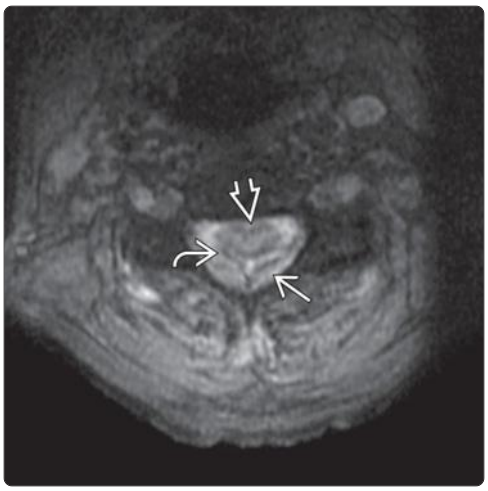
(Left) *Sagittal T1 MR shows an intermediate signal posterior epidural hematoma displacing the cord anteriorly against disc osteophyte complexes.* **(Right)** *Sagittal T2WI MR shows a heterogeneous posterior hematoma effacing the thecal sac and compressing the cord. Note the contusion at the level of the disc osteophyte complex.*



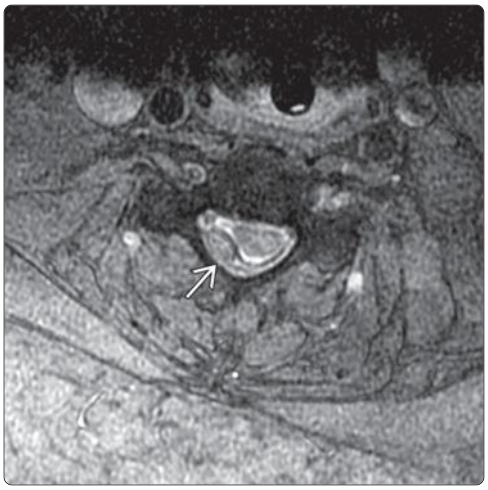


(Left) *Sagittal T1WI MR shows a long segment intermediate signal collection dorsal to the thecal sac which displaces the cord anteriorly.* **(Right)** *Axial T2* GRE MR shows a posterior epidural hematoma as central increased signal with linear low peripheral signal, in part due to displaced dura. The cord is mildly displaced anteriorly.*









(Left) *Sagittal T2WI MR shows a heterogeneous low signal collection within the dorsal epidural space of the cervical spine, extending inferiorly from C6 into the upper thoracic spine. There is cord compression from the hematoma.* **(Right)** *Axial T2* GRE MR shows a cervical epidural hematoma displacing the dorsal dural margin and the cord anteriorly. The hemorrhage is eccentric to the right.*





(Left) *Sagittal T1WI MR shows acute epidural hemorrhage as an intermediate signal mass dorsal to the cord. Note that in the acute phase, there will not be T1 hyperintensity from the blood products.* **(Right)** *Sagittal T1 C+ MR shows a thin rim of peripheral enhancement outlining the large dorsal cervical epidural hematoma.*





(Left) *Sagittal T1WI MR shows a focal extradural collection dorsal to thecal sac at the C4 and C5 level, severely compressing the cervical cord. The lesion shows classic T1 hyperintensity of methemoglobin.* **(Right)** *Axial T2* GRE MR shows heterogeneous low signal from the well-defined epidural hemorrhage. There is moderate mass effect upon the cervical cord.*

Subdural Hematoma

KEY FACTS

TERMINOLOGY

- Accumulation of blood between dura, arachnoid

IMAGING

- Intradural collection hyperintense on T1WI, predominantly hypointense on T2WI or gradient-echo imaging
- Thoracolumbar > lumbar or lumbosacral > cervical
- Clumped, loculated masses of hemorrhagic density/intensity

TOP DIFFERENTIAL DIAGNOSES

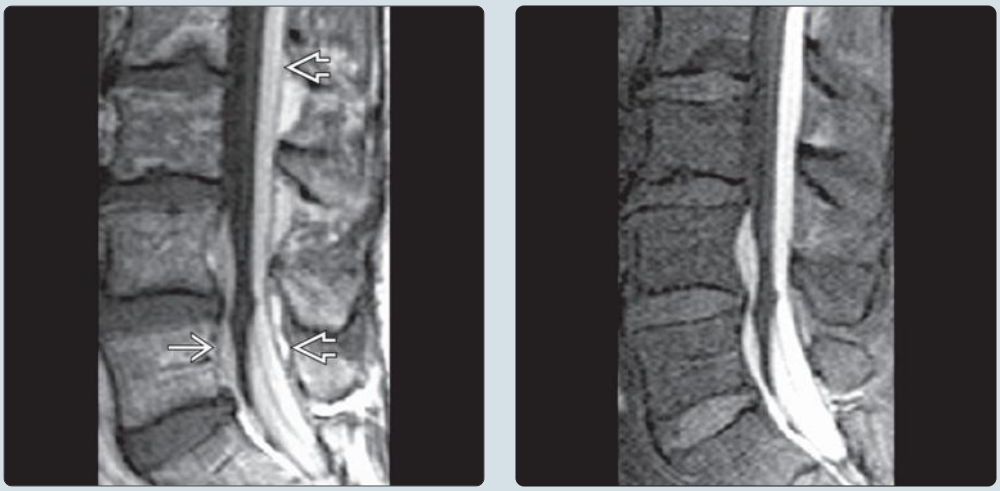
- Epidural hematoma
- Subdural abscess
- CSF leakage syndrome
- Spinal meningitis
- Idiopathic hypertrophic spinal pachymeningitis

PATHOLOGY

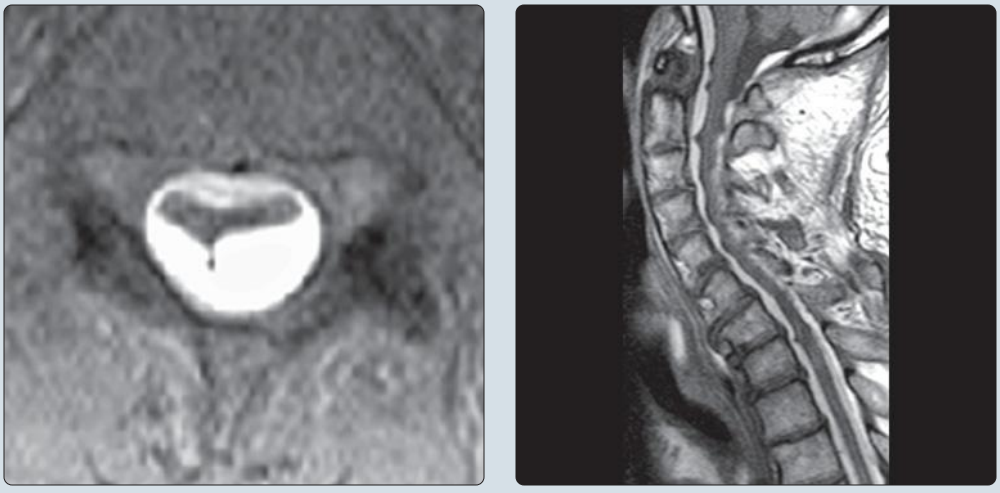
- Trauma
- Bleeding diathesis: 54% of reported cases
- Iatrogenic cause is factor in 2/3 of those with abnormal coagulation parameters
- Neoplasm
- Arteriovenous malformation
- Postoperative complication
- Spontaneous: 15%

CLINICAL ISSUES

- Acute onset of neck or back pain
- Radicular pain, bladder/bowel dysfunction
- Much less common than spinal epidural hematoma
- Treatment
  - May resolve spontaneously
  - Decompressive laminectomy with clot evacuation
    - Indicated with severe and progressive deterioration of neurologic symptoms



(Left) *Sagittal T1WI MR without fat saturation shows prominent hyperintense linear collections both ventral and dorsal within the bony spinal canal, with narrowing of the thecal CSF space.* **(Right)** *Sagittal T1WI MR with fat saturation shows hyperintense linear subdural blood collections both ventral and dorsal within the bony canal, with marked narrowing of thecal CSF space. Iatrogenic cause is a factor in 2/3 of those with abnormal coagulation parameters.*



(Left) *Axial T1WI FS MR shows the typical appearance of subdural hemorrhage, with well-defined outer margin bounded by the dura, and inner lobulated margin, giving a "Mercedes Benz" sign.* **(Right)** *Sagittal T1WI MR in a patient with chronic neck pain demonstrates diffuse ventral and dorsal intraspinal hyperintense collections, with intracranial extension.*

TERMINOLOGY

Abbreviations

- Spinal subdural hematoma (SSDH)

Definitions

- Accumulation of blood between dura, arachnoid

IMAGING

General Features

- Best diagnostic clue
  - Intradural collection hyperintense on T1WI, predominantly hypointense on T2WI or gradient-echo imaging
- Location
  - Thoracolumbar > lumbar or lumbosacral > cervical
  - Ventral, dorsal, lateral, or circumferential
- Size
  - Variable cranial-caudal extension
    - Typically over multiple levels
- Morphology
  - Clumped, loculated masses of hemorrhagic density/intensity
  - Conforms to dura
  - Distinct from epidural fat and adjacent osseous structures

Radiographic Findings

- Radiography
  - Noncontributory except in setting of trauma

CT Findings

- NECT
  - Hyperdense intradural collection
    - Outlined by epidural fat and relatively low-density cord
  - Sagittal reformation best depicts craniocaudal extent
  - Cord compression
- CECT
  - No significant enhancement
  - Bone CT
    - Vertebral fractures in trauma patients

MR Findings

- T1WI
  - Heterogeneous, variable signal intensity depending on duration of SSDH
    - Acute: Iso- to slightly hyperintense
    - Subacute to chronic: Hyperintense
- T2WI
  - Heterogeneously hyperintense
    - Significant hypointense portion
- STIR
  - Hyperintense
- T2* GRE
  - Significant hypointense portion due to deoxyhemoglobin
- T1WI C+
  - ± slight enhancement
  - Variable mass effect on spinal cord and cauda equina
    - Hyperintense cord edema on T2WI

Nonvascular Interventions

- Myelography
  - Effaced subarachnoid space over multiple levels
  - Cord compression

Imaging Recommendations

- Best imaging tool
  - Sagittal and axial T2WI and T1WI MR
  - Spinal angiography not indicated unless findings of arteriovenous malformation present

DIFFERENTIAL DIAGNOSIS

Epidural Hematoma

- Thoracolumbar spine most common
- "Capping" of hematoma by dorsal epidural fat on sagittal imaging
- Directly adjacent to osseous structures with effacement of epidural fat
  - Not limited by dura
- Iso- to hyperintense on T1WI acute, hyperintense subacute to chronic
- Hyperintense on T2WI with hypointense deoxyhemoglobin
- ± central or peripheral enhancement

Subdural Abscess

- Thoracolumbar spine most common
- Multi segmental dural/subdural thickening
- Obliterated CSF
- Isointense on T1WI, hyperintense on T2WI
- Diffuse vs. rim enhancement

CSF Leakage Syndrome

- Spontaneous or from prior spinal trauma, diagnostic or interventional procedure
- Dural venous engorgement and enhancement
- Diffuse smooth meningeal thickening, enhancement
- May see cerebellar tonsillar descent, effaced prepontine space in posterior fossa

Spinal Meningitis

- Normal or increased CSF intensity on T1WI
- Indistinct cord-CSF interface
- Diffusely enhancing CSF
- Smooth or nodular meningeal/nerve root enhancement
- ± cord edema/swelling

Idiopathic Hypertrophic Spinal Pachymeningitis

- Diagnosis of exclusion
- Fibrous inflammatory thickening of dura mater
- Thickened dura iso- to hypointense on T1WI and T2WI
  - Marked post-contrast enhancement

PATHOLOGY

General Features

- Etiology
  - Trauma
    - Mechanism
      - Look for vertebral fracture-dislocation
      - Post-traumatic disc herniation

- Hemorrhagic cord contusion: Hyperintense on T2WI with focal hypointensity
- Other proposed mechanisms of SSDH formation
- Rupture of valveless radiculomedullary veins that cross subdural and subarachnoid spaces
- Rupture of extraarachnoidal vessels on inner dural surface
- Bleeding diathesis: 54% of reported cases
- Anticoagulation
- Coagulopathy: Hemophilia, leukemia, thrombocytopenia, polycythemia
- Iatrogenic cause is factor in 2/3 of those with abnormal coagulation parameters
- Lumbar puncture
- Spinal anesthesia
- Neoplasm
- Arteriovenous malformation
- Postoperative complication
- Spontaneous: 15%
- Genetics
- No genetic predilection
- Embryology-anatomy
- Inner dura (dural border cell layer) structurally weaker than outer layer
- May tear during trauma
- Creating potential space within dural border cell layer
- SSDH forms between strong external dura and arachnoid
- No true spinal subdural space demonstrated in human autopsy studies

Gross Pathologic & Surgical Features

- Subdural (partially) organized blood clot under pressure
- Bluish tense dura
- ± subarachnoid/epidural hemorrhage

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
- Acute onset of neck or back pain
- Radicular pain
- Other signs/symptoms
- Headache
- Meningismus
- Paraplegia
- Paresthesia
- Bladder/bowel dysfunction
- Diminished deep tendon reflexes
- Clinical profile
- Neurologic impairment may be delayed
- Onset of neurologic deficits varies from hours to > 1 week
- Chronic SSDH may not have significant pain
- Mimics subarachnoid hemorrhage clinically

Demographics

- Age
- All ages affected
- Slightly increased incidence in those ≥ 60 years
- Gender
- M = F
- Ethnicity
- No racial predilection
- Epidemiology
- Much less common than spinal epidural hematoma

Natural History & Prognosis

- May resolve spontaneously
- Favorable clinical and imaging outcome in those suitable for conservative management
- > 40% of patients undergoing surgery have near complete neurologic recovery
- Concurrent subarachnoid hemorrhage portends poor outcome

Treatment

- Decompressive laminectomy with clot evacuation
- Indicated with severe and progressive deterioration of neurologic symptoms
- Nonoperative management
- In cases of mild symptoms at presentation
- In patients with rapid neurologic recovery
- Analgesics
- ± intravenous dexamethasone to decrease inflammatory reaction and cord edema

DIAGNOSTIC CHECKLIST

Consider

- NECT to confirm intradural location and hyperdense acute hematoma if MR findings equivocal

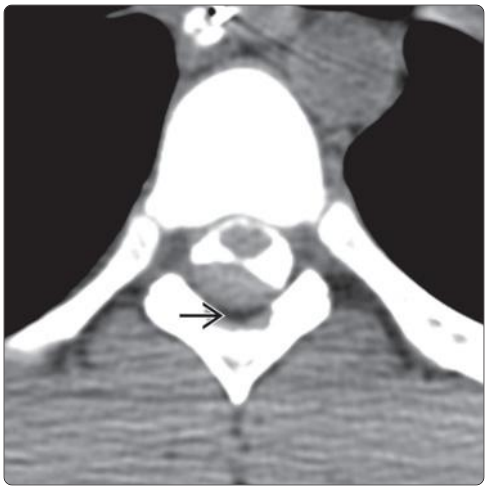
Image Interpretation Pearls

- Intradural extramedullary collection hyperintense on T1WI and predominantly hypointense on T2WI or gradient-echo imaging without significant enhancement characteristic of SSDH

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(Left) *Sagittal T1 MR shows a diffuse marrow signal abnormality due to metastatic disease with a mild pathologic compression fracture at L3 ⇨. Subdural hemorrhage ⇨, seen as slightly increased signal, surround the distal cord and lumbar CSF.* **(Right)** *Axial CECT following myelography shows lobulated contour deformity to the contrast column, which is clearly due to subdural collection in the thoracic spine with the maintained normal epidural space ⇨.*





(Left) *Sagittal T2WI MR shows a large ventral heterogeneous blood collection ⇨, with loculation and prominent hypointense areas. Considerable mass effect is present on the cauda equina, which is displaced posteriorly.* **(Right)** *Sagittal T1WI MR through the lumbosacral spine shows extensive ventral ⇨ and minimal dorsal ⇨ hyperintense subdural hematomas. Note the marked narrowing of the thecal sac.*





(Left) *Sagittal T1WI MR shows clival and cervical ventral subdural hematoma ⇨ following trauma, with a subacute blood product with slightly hyperintense lobulated contour and absence of ligamentous disruption.* **(Right)** *Sagittal T1 MR shows a diffuse marrow signal abnormality due to metastatic disease with a mild pathologic compression fracture at L3 ⇨. Subdural hemorrhage ⇨, seen as slightly increased signal, surround the distal cord and lumbar CSF.*

Superficial Siderosis

KEY FACTS

TERMINOLOGY

- Synonyms: Central nervous system siderosis, hemosiderosis
- Recurrent subarachnoid hemorrhage (multiple etiologies) causing hemosiderin deposition on cord and nerve surface

IMAGING

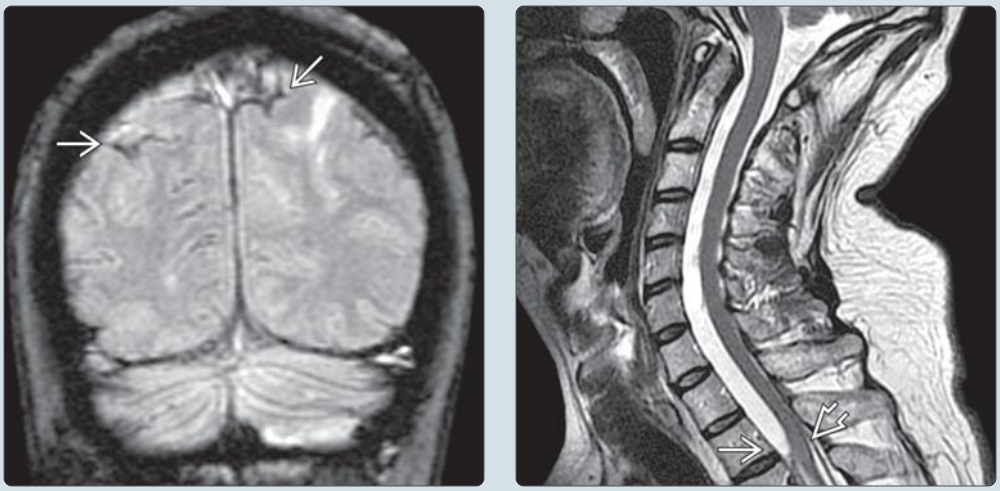
- Diffuse hypointensity of cord/brain surface on T2WI, gradient-echo images
- MR brain/spine
- Diagnosis of siderosis necessitates search for underlying cause of repetitive subarachnoid hemorrhage

TOP DIFFERENTIAL DIAGNOSES

- Artifact with low signal adjacent to cord related to CSF flow dephasing
- Subdural hemorrhage
- Cord peripheral T2 hypointensity in dural fistula
- Underlying etiology of subarachnoid hemorrhage
  - Dural source with spinal cystic collection, such as meningocele, pseudomeningocele, or diverticula
  - Ependymoma
  - Astrocytoma
  - Arteriovenous malformations
  - Post-traumatic or postoperative

CLINICAL ISSUES

- Bilateral sensorineural hearing loss (95%)
- Ataxia (88%)
- Pyramidal signs (76%)
- Preclinical phase often prolonged (> 10 years)
- Insidious, progressive abnormality



(Left) *Coronal T2* GRE MR shows low signal superficial siderosis following aneurysmal subarachnoid hemorrhage.* **(Right)** *Sagittal T2WI MR shows severe spinal arachnoiditis with ventral tethering of the cord and superficial siderosis seen as leptomeningeal low signal following aneurysmal subarachnoid hemorrhage. This is an unusual occurrence following subarachnoid hemorrhage, but should be kept in mind in a patient with new cord symptoms that occur late after subarachnoid hemorrhage (SAH).*



(Left) *Sagittal T2WI MR shows arachnoiditis and superficial siderosis following aneurysmal subarachnoid hemorrhage with abrupt ventral displacement of the thoracic cord at T3. Cord shows T2 hyperintensity below level of ventral tethering and arachnoiditis with low cord surface signal.* **(Right)** *Sagittal T2WI MR in a patient following thoracic ependymoma resection shows diffuse hemosiderin staining of thoracic cord with diffuse low signal, volume loss, and local tethering to laminectomy site.*

TERMINOLOGY

Abbreviations

- Superficial siderosis (SS), sensorineural hearing loss (SNHL)

Synonyms

- Central nervous system siderosis, hemosiderosis

Definitions

- Recurrent subarachnoid hemorrhage (multiple etiologies) causing hemosiderin deposition on cord and nerve surface

IMAGING

General Features

- Best diagnostic clue
  - Diffuse hypointensity of cord/brain surface on T2WI, gradient-echo images
- Location
  - Cerebellum, brainstem, cerebrum, cord, cranial nerves
- Size
  - Linear low signal < few mm in thickness
- Morphology
  - Curvilinear, following surface contour of involved CNS structure

CT Findings

- NECT
  - Cord, brain volume loss
- CECT
  - No enhancement

MR Findings

- T1WI
  - Usually normal, occasional low signal on cord surface
- T2WI
  - Linear low signal involving cord surface, nerve roots
  - Look for typical surface involvement of cerebellum, brainstem
- T2* GRE
  - Linear low signal involving cord surface, nerve roots
  - Findings more pronounced than with T2WI, T1WI
- T1WI C+
  - No enhancement of leptomeninges at site of hemosiderin deposition
  - May see pachymeningeal enhancement as with intracranial hypotension due to CSF leak if that is etiology of repetitive hemorrhage

Imaging Recommendations

- Best imaging tool
  - MR brain/spine
  - Diagnosis of siderosis necessitates search for underlying cause of repetitive subarachnoid hemorrhage
- Protocol advice
  - Brian MR: T1WI, T2WI, FLAIR, post-contrast T1WI, T2* GRE
  - Spine: Scan total spine with sagittal T2WI, T2* GRE, post-contrast T1WI

DIFFERENTIAL DIAGNOSIS

MR Artifact

- Low signal adjacent to cord, related to CSF flow dephasing
- Appearance varies with different sequences, plane of acquisition

Cord Central T2 Hyperintensity

- Variety of pathologies may give cord central T2 hyperintensity, causing periphery of cord to look abnormally low signal
  - Cord infarction: Acute symptoms onset
  - Look for cord expansion, enhancement with demyelinating disease, or tumor which does not occur with SS

Subdural Hemorrhage

- Lobulated low T2 signal adjacent to cord, not on cord surface

Cord Peripheral T2 Hypointensity in Dural Fistula

- True cord decreased T2 signal with type I dural fistula, thought related to venous hypertensive myelopathy

Underlying Etiology of Subarachnoid Hemorrhage

- Intramedullary neoplasm
  - Ependymoma
  - Astrocytoma
  - Cavernous malformation(s)
- Arteriovenous malformations
- Cranial pathology (postoperative)
- Dural source with spinal cystic collection, such as meningocele, pseudomeningocele, or diverticula

PATHOLOGY

General Features

- Etiology
  - Repeated subarachnoid hemorrhage → hemosiderin encrustation of surface of CNS
    - Hemosiderin is cytotoxic
  - Involvement of brain, cord surface is active process requiring glial tissues (microglia, Bergmann glia, subpial astrocytes)
    - Peripheral myelinated structures much less involved
    - Long length of central myelination of CN8 explains preferential involvement, along with CN1 and CN2 involvement
  - Hemosiderin deposition may extend into cord gray matter to affect neurons
    - Ferritin biosynthesis overwhelmed by large iron load → excess free iron → stimulation of lipid peroxidation → local tissue necrosis
- Associated abnormalities
  - Hydrocephalus
- Hemosiderin coating surface of cerebellum, brainstem, and spinal cord
- Extends to and involves pial, subpial, and subependymal surfaces
- Variable neuronal loss, reactive gliosis, demyelination
  - Cerebellum typically shows loss of Purkinje cells and Bergmann gliosis

Gross Pathologic & Surgical Features

- Dark brown/black staining of pial/subpial surface of cord, nerve roots

Microscopic Features

- Staining of cord surface by hemosiderin
- Leptomeninges fibrotic, thickened with hemosiderin-laden macrophages

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Bilateral sensorineural hearing loss (95%)
    - Ataxia (88%)
    - Pyramidal signs (76%)
    - Dementia (20%)
    - Nystagmus (19%)
    - Anosmia (17%)
  - Poor correlation of MR findings with clinical symptoms
  - Cause of recurrent subarachnoid hemorrhage found in 50%
    - Postoperative change with friable surgical cavity (hemispherectomy)
    - Dural source with fluid collection, such as pseudomeningocele or diverticulum
      - Traumatic nerve root avulsion
      - Congenital or postoperative peridural fluid collection
    - Neoplastic hemorrhage (35%); ependymoma most common tumoral cause
    - Arteriovenous malformation
    - Aneurysm
    - Cavernous malformation (0.5-0.9% of population affected)
  - Idiopathic (25-46%)
- Clinical profile
  - Classic description: Sensorineural hearing loss, cerebellar ataxia, dementia, myelopathy (uncommon to see all manifestations)
  - CSF analysis
    - Xanthochromia
    - Elevated RBC count
    - Elevated iron and ferritin levels
    - May be normal

Demographics

- Age
  - Adult and children (average: 50 years)
- Gender
  - M:F = 3:1
- Ethnicity
  - No predilection
- Epidemiology
  - Insidious, progressive abnormality
  - 0.15% patients undergoing MR imaging
    - 85% showed no symptoms

Natural History & Prognosis

- Preclinical phase often prolonged (> 10 years)
  - Reports of SS developing > 30 years following initial surgical procedure

Treatment

- Treat underlying abnormality by surgical or endovascular means
  - Arteriovenous malformation (AVM), aneurysm, tumor
- Cochlear implant for SNHL
- Iron/copper chelates unsuccessful for treatment

DIAGNOSTIC CHECKLIST

Consider

- Superficial siderosis is secondary to repetitive subarachnoid hemorrhage
  - Look for underlying etiology in brain/spine

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(Left) *Axial T2WI MR shows superficial siderosis with hypointense deposition along entire cord surface.* **(Right)** *Sagittal T2WI MR reveals hypointense signal intensity coating of the spinal cord surface from hemosiderin deposition.*





(Left) *Axial T2WI MR shows superficial siderosis with hypointense deposition along entire cord pial surface from subarachnoid blood product distribution.* **(Right)** *Sagittal T2WI MR reveals hypointense signal intensity coating of the spinal cord surface from hemosiderin deposition.*





(Left) *Sagittal T2WI MR of recurrent ependymoma shows superficial siderosis extending cephalad along cord.* **(Right)** *Sagittal T2WI MR shows large filum terminale ependymoma with drop metastases, superficial siderosis of cauda equina with linear low signal.*

Hematomyelia/Nontraumatic Cord Hemorrhage

KEY FACTS

TERMINOLOGY

- Synonyms
  - Intramedullary hemorrhage, spontaneous cord hemorrhage
- Definition: Hemorrhage within spinal cord from a variety of etiologies, primarily anticoagulant therapy, vascular, and neoplastic

IMAGING

- MR to show variable signal mass within cord, depending upon stage of blood product breakdown

TOP DIFFERENTIAL DIAGNOSES

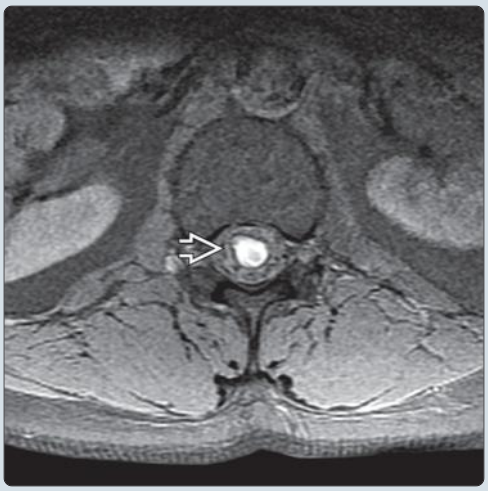
- Coagulopathy
- Vascular malformation
  - Cavernous malformation
  - Arteriovenous malformation
    - Most commonly seen with intramedullary nidus, as in type II and III
- Neoplasm
  - Ependymoma
  - Astrocytoma
  - Metastatic
  - Hemangioblastoma (very rare)
- Iatrogenic
  - Postsurgical hemorrhage into operative site
- Infarct
- Vasculitis, SLE (rare)
- Idiopathic

CLINICAL ISSUES

- Critical factors for recovery
  - Extent of preoperative neurological deficit
  - Operative interval
    - The sooner the operation, the better
- Complete recovery in 40% of spinal hemorrhage patients

(Left) *Sagittal T2WI MR of cord hemorrhage from conus level AVM shows hemorrhage varying from fluid-like cavity in the conus tip ⇨ to hemosiderin ⇨. There are multiple serpentine intradural flow voids about the dorsal distal cord.* **(Right)** *Axial T1WI FS MR shows hemorrhage from conus level AVM. Methemoglobin is present within the central aspect of the conus ⇨.*





(Left) *Sagittal T1WI MR in a typical case of intramedullary and bony metastases shows hypointense vertebral body ⇨ metastases and a rounded hyperintense intramedullary lesion in the conus ⇨. Conus lesion signal relates to hemorrhage.* **(Right)** *Sagittal T2WI MR of intramedullary hemorrhagic metastases shows rounded hypointense intramedullary lesion in the conus ⇨ with surrounding vasogenic edema. Vertebral metastases are difficult to identify ⇨.*





TERMINOLOGY

Synonyms

- Hematomyelia, intramedullary hemorrhage, spontaneous cord hemorrhage

Definitions

- Hemorrhage within spinal cord from a variety of etiologies, primarily anticoagulant therapy, vascular, and neoplastic

IMAGING

General Features

- Best diagnostic clue
  - T1 hyperintensity (methemoglobin) within cord
- Location
  - Intramedullary
- Size
  - Variable
- Morphology
  - Linear to lobulated

Imaging Recommendations

- Best imaging tool
  - MR to show variable signal mass within cord, depending upon stage of blood product breakdown
- Protocol advice
  - Include gradient-echo sagittal/axial

DIFFERENTIAL DIAGNOSIS

Coagulopathy With Cord Hemorrhage

- Nonspecific imaging findings with appropriate clinical history

Vascular Malformation

- Cavernous malformation
- Arteriovenous malformation (AVM): Most commonly seen with intramedullary nidus, as in type II and III
  - Type II and III may give subarachnoid hemorrhage
  - Type I fistulas do not present with hemorrhage
  - Perimedullary venous drainage of AVM increases risk of hemorrhage

Neoplasmic Hemorrhage

- Ependymoma
- Astrocytoma
- Metastatic (melanoma)
- Hemangioblastoma (very rare)
- Intradural extramedullary lesions give subarachnoid hemorrhage (SAH), not parenchymal
  - Schwannoma (rare)
  - Meningioma (rare)

Iatrogenic Hemorrhage

- Postsurgical hemorrhage into operative site
- Rare reported cases of direct injury to cord from percutaneous needle placement

Infarct

- Hemorrhagic transformation (rare)

Vasculitis, SLE (Rare)

- Nonspecific imaging findings given appropriate history

Idiopathic

- Diagnosis of exclusion

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May present with flaccid paraplegia due to spinal shock
  - Spastic paraplegia also possible
  - Brown Séquard seen with unilateral cord involvement

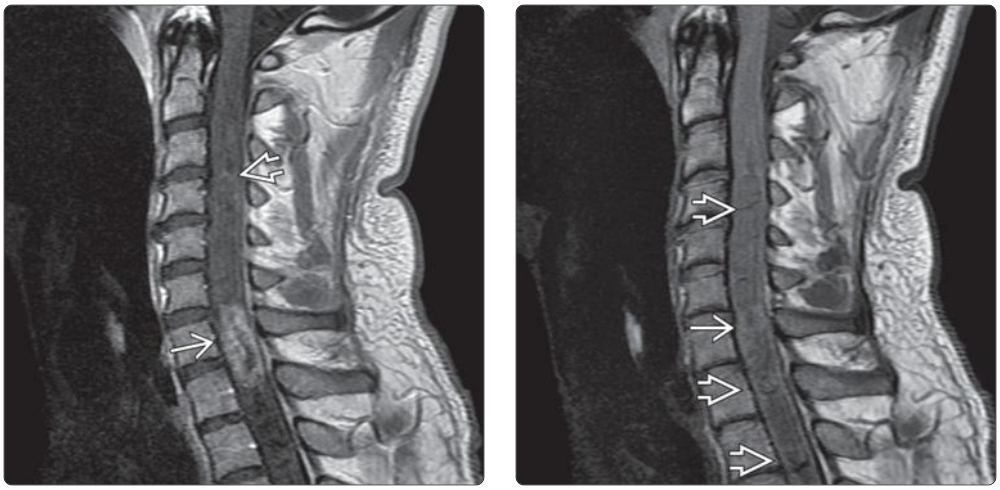
Natural History & Prognosis

- Critical factors for recovery
  - Extent of preoperative neurological deficit
  - Operative interval (sooner the operation, the better)
- Complete recovery in 40% of spinal hemorrhage patients

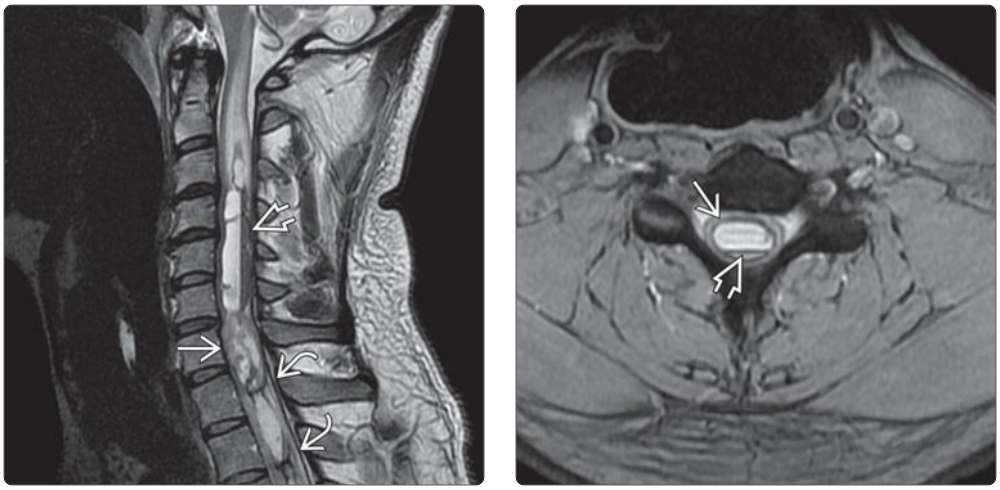
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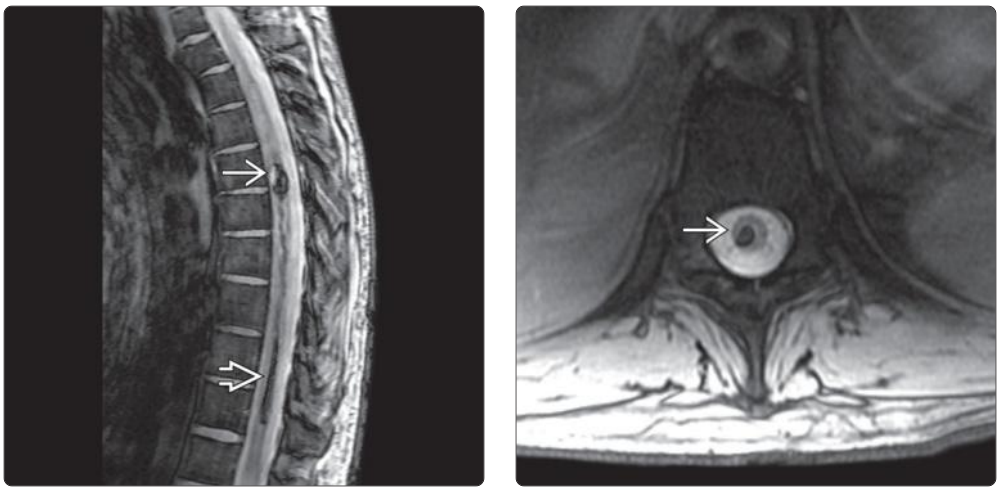
(Left) *Sagittal T1 C+ MR shows irregular enhancing intramedullary spinal cord ependymoma at C7. Faint tumor enhancement extends to C3 level.* **(Right)** *Unenhanced sagittal T1 MR in this patient with a cord hemorrhagic ependymoma shows mild heterogeneous signal from the mass and regions of hemorrhage and hemosiderin.*



(Left) *Sagittal T2 MR shows very heterogeneous signal from the large hemorrhagic cervical cord ependymoma. Hemorrhage and debris fluid-fluid levels are seen, along with more chronic hemosiderin capping of loculated cystic regions.* **(Right)** *Axial T2* GRE MR shows low signal hemosiderin lining the cyst wall in this patient with a hemorrhagic ependymoma. Note the low signal fluid-fluid level within the cyst due to more recent hemorrhage.*



(Left) *Sagittal T2* GRE MR shows large focus of low signal from a thoracic cord cavernous malformation, which has hemorrhaged into the more inferior cord.* **(Right)** *Axial T2* GRE MR shows focal low signal within the substance of the thoracic cord from a hemorrhagic cavernous malformation.*







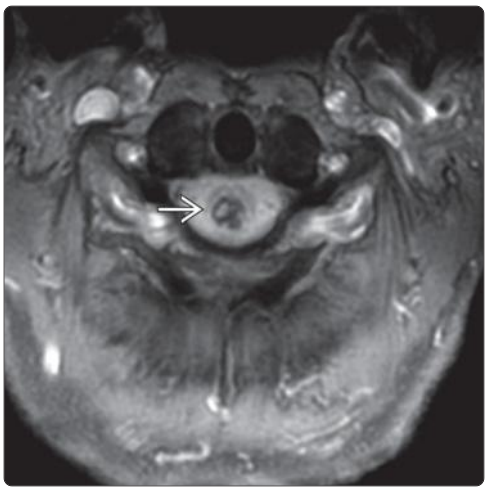
(Left) *Sagittal unenhanced T1 MR in this patient with cord hemorrhage from a cavernous malformation shows heterogeneous signal within the cord at C5-C7 levels, which is very nonspecific. This could reflect tumor or hemorrhage.* **(Right)** *Sagittal T2WI MR of intramedullary hemorrhage from cavernous malformation shows large oblong area of intramedullary low T2 signal. There is extensive cord edema extending cephalad and caudad to the intramedullary hemorrhage.*





(Left) *Sagittal unenhanced T1 MR in this patient with hematomyelia from a thoracic cavernous malformation shows heterogeneous signal involving a long segment of the cord from blood dissecting along the spinal cord tracts.* **(Right)** *Sagittal T1 C+ MR in this patient with von Hippel-Lindau disease shows hemangioblastoma involving the dorsal medulla with hemorrhage extending inferiorly into the cervical cord. Hemorrhage from hemangioblastomas is very rare.*





(Left) *Sagittal T2 MR in this patient with hemangioblastoma shows extensive low signal hemorrhage extending inferiorly from the tumor into the cervical cord. There is extensive cord edema.* **(Right)** *Axial T2* GRE MR shows low signal hemorrhage within the central aspect of the cervical cord in this patient with hematomyelia from a cord tumor.*

Bow Hunter Syndrome

KEY FACTS

TERMINOLOGY

- Bow hunter stroke, positional occlusion of vertebral artery, rotational occlusion of vertebral artery
- Vertebrobasilar insufficiency secondary to mechanical occlusion or stenosis of vertebral artery during head rotation

IMAGING

- Occlusion or stenosis of vessel with head positional dependence based on ultrasound, MRA, CTA, or catheter angiography

PATHOLOGY

- Vertebral artery at C1-C2 particularly prone to mechanical compression
- May occur anywhere along vertebral course related to spondylosis, atlantoaxial instability, hypertrophy of atlantooccipital membranes, paravertebral muscle fascial bands

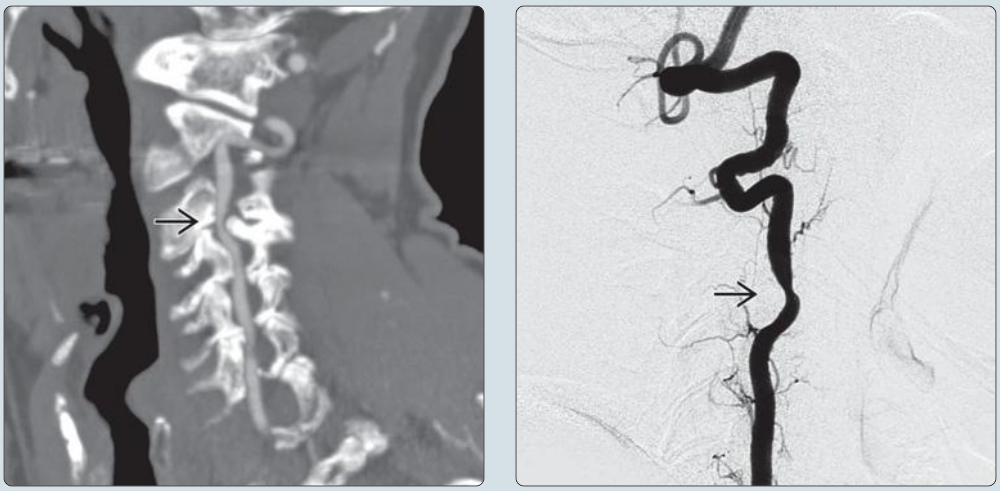
CLINICAL ISSUES

- Vertebrobasilar insufficiency
  - Motor or sensory deficits, ataxia, diplopia, dysarthria, dysmetria, vertigo, visual field deficit, cranial nerve dysfunction, syncope
- Treatment
  - Brace to restrict head motion, surgical fusion to prevent atlantoaxial rotation, vertebral artery decompression
  - Vertebral artery stenting

DIAGNOSTIC CHECKLIST

- Temporary positional occlusion of 1 vertebral artery during course of daily activities may be normal, if asymptomatic
- Hypoplasia or stenosis of contralateral vertebral artery predisposes patients to vertebrobasilar ischemic attacks during head rotation

(Left) *Coronal oblique CTA shows focal extrinsic narrowing of the left vertebral artery at the C3-C4 level due to facet degenerative hypertrophy and osteophyte formation* ⇨. **(Right)** *Lateral catheter angiography in a neutral position shows moderate focal stenosis at the C3-C4 level due to osteophytic compression* ⇨.



(Left) *Anteroposterior catheter angiography with the head turned to the right (asymptomatic head turn side) shows patent left vertebral with mild C3-C4 level narrowing* ⇨. **(Right)** *Anteroposterior catheter angiography with the head turned to the left (symptomatic head turn side) shows occlusion of the vertebral artery at the level of the osteophytic compression* ⇨.



TERMINOLOGY

Synonyms

- Bow hunter stroke, positional occlusion of vertebral artery, rotational occlusion of vertebral artery

Definitions

- Vertebrobasilar insufficiency secondary to mechanical occlusion or stenosis of vertebral artery during head rotation due to fibrous band or bony prominence

IMAGING

General Features

- Best diagnostic clue
  - Occlusion or stenosis of vessel with head positional dependence based on ultrasound, MRA, CTA, or catheter angiography
- Location
  - Along course of vertebral artery, typically at C1-C2 junction
- Morphology
  - Stenosis or occlusion of vertebral artery with head position change

CT Findings

- CTA
  - Stenosis or occlusion of vertebral artery with head turned to symptom-producing side

Ultrasonographic Findings

- Color Doppler
  - Reduction of forward flow velocity in involved vertebral artery with head turning to symptom-producing side

Imaging Recommendations

- Best imaging tool
  - Catheter angiography allows rapid assessment of dynamic component of stenosis/occlusion with head turning

DIFFERENTIAL DIAGNOSIS

Vertebral Occlusion From

- Atherosclerosis
- Traumatic dissection

PATHOLOGY

Gross Pathologic & Surgical Features

- Mechanical production of stenosis or occlusion along course of vertebral artery, which is positional
  - Hypoplasia or stenosis of contralateral vertebral artery predisposes patients to vertebrobasilar ischemic attacks during head rotation
  - May occur anywhere along vertebral course related to spondylosis, atlantoaxial instability, hypertrophy of atlantooccipital membranes, paravertebral muscle fascial bands
  - Vertebral artery at C1-C2 particularly prone to mechanical compression
    - For example, rotation of head to the right fixes right atlantoaxial joint and opens up left joint, which may stretch or compress left vertebral artery
  - Rare reports of positional vertebral occlusion related to congenital craniovertebral junction bony anomalies such as occipitalization of atlas

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Vertebrobasilar insufficiency
    - Motor or sensory deficits, ataxia, diplopia, dysarthria, dysmetria, vertigo, visual field deficit, cranial nerve dysfunction, syncope
    - May lead to stroke with permanent neurological deficit

Treatment

- Brace to restrict head motion, surgical fusion to prevent atlantoaxial rotation, vertebral artery decompression
- Vertebral artery stenting

DIAGNOSTIC CHECKLIST

Consider

- Temporary positional occlusion of 1 vertebral artery during course of daily activities may be normal, if asymptomatic

Image Interpretation Pearls

- Hypoplasia or stenosis of contralateral vertebral artery predisposes patients to vertebrobasilar ischemic attacks during head rotation

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Vertebral Artery Dissection

KEY FACTS

TERMINOLOGY

- Vertebral artery (VA) dissection
- Irregularity of VA contour from intimal tear or subadventitial hematoma

IMAGING

- **Stenoocclusive dissection**
  - Dissection to subintimal plane with vessel luminal narrowing or occlusion
- **Dissecting aneurysm**
  - Dissection into subadventitial plane with dilatation of outer wall
- Intramural hematoma is pathognomonic
  - Best seen as **bright crescent** on T1 FS MR
- CTA source images show contour changes of lumen
- Conventional angiography is gold standard

TOP DIFFERENTIAL DIAGNOSES

- Extracranial atherosclerosis
- Fibromuscular dysplasia
- Miscellaneous vasculitis
- Congenital VA hypoplasia

PATHOLOGY

- **Traumatic** VA dissection
  - Direct or indirect arterial injury
- **Spontaneous** VA dissection
  - Many associations and predisposing factors

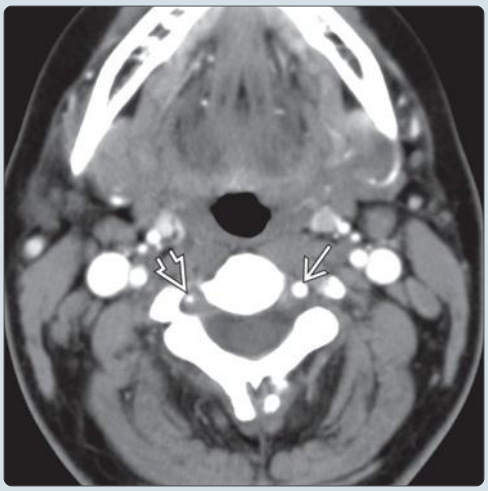
CLINICAL ISSUES

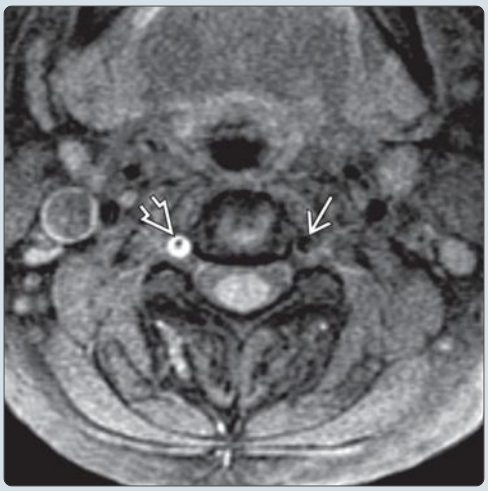
- Age: Adults < 45 years

DIAGNOSTIC CHECKLIST

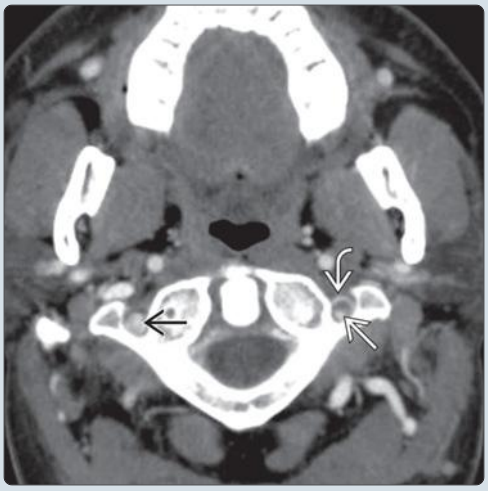
- Check other vessels carefully for 2nd dissection
- Beware of "suboccipital rind" sign
  - Intramural hematoma without narrow lumen
- Cerebellar infarction in young to middle-aged adults; need to exclude PICA infarction due to VA dissection

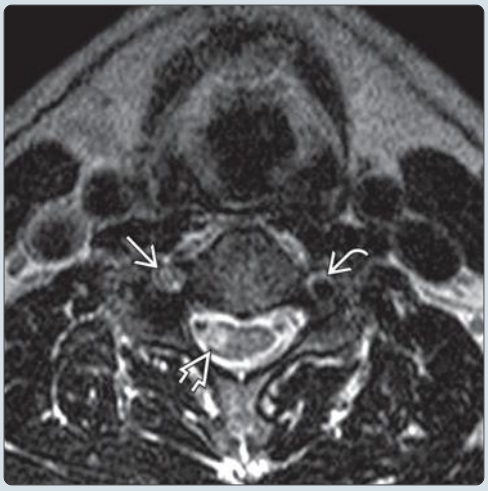
(Left) *Axial CECT in a patient with idiopathic vertebral artery dissection demonstrates normal caliber of left vertebral artery and narrowed, eccentric lumen of right vertebral artery. Note normal and symmetric foramina transversarium.* **(Right)** *Axial T1 FS MR in the same patient reveals a hyperintense crescent of mural hematoma with small hypointense patent lumen. Contralateral vertebral artery shows no methemoglobin hyperintensity. This is unilateral vertebral artery dissection.*





(Left) *Axial CTA shows rim enhancement of the left vertebral artery with low-density mural hematoma and contrast filling the narrowed lumen. Subtle linear lucency in the right vertebral artery proved not to be a dissection flap.* **(Right)** *Axial T2 MR in a patient with neck trauma and cervical fractures reveals traumatic vertebral artery dissection as loss of right vertebral flow void, as compared to the normal left side. Note right cervical hemicord hyperintensity from cord ischemia.*





TERMINOLOGY

Abbreviations

- Vertebral artery (VA) dissection

Definitions

- Irregularity of VA contour from intimal tear or subadventitial hematoma

IMAGING

General Features

- Best diagnostic clue
  - Important finding on axial T1 FS MR is **crescentic hyperintense** mural hematoma
- Location
  - VA is most mobile and, therefore, most vulnerable to mechanical injury at C1-C2 as it exits C1 transverse foramen and pierces dura
- Morphology
  - 2 distinct forms of VA dissection
  - **Stenoocclusive dissection**
    - Tapered stenosis or occlusion
    - Long segment stenosis → "string" sign
    - May involve intra- and extradural VA
  - **Dissecting aneurysm**
    - Focal or fusiform aneurysmal dilatation ± stenosis
    - Usually involves intradural VA
    - May present with subarachnoid hemorrhage (SAH)

CT Findings

- CTA
  - Axial source images show contour changes of lumen
  - **"Suboccipital rind" sign**
    - Normally opacified VA lumen but thickened wall from intramural hematoma
    - May be difficult to delineate on MRA or catheter angio

MR Findings

- T1WI
  - Mural hematoma signal varies with time
    - Hyperacute or acute blood
      - Oxy-/deoxyhemoglobin: Iso- → hyperintense
    - Subacute blood (~ 2-3 days)
      - Methemoglobin intrinsically bright on T1 MR
- T1WI FS
  - FS makes mural methemoglobin more conspicuous
- T2WI
  - Loss of normal flow void
- MRA
  - Reveals lumen caliber changes
    - Aneurysmal dilatation and stenosis/occlusion
  - Intimal flap and double lumen occasionally seen
- Magnetization-prepared radiofrequency pulses and rapid gradient-echo sequences (MP-RAGE)
  - Luminal crescentic T1 hyperintensity related to methemoglobin
- Fusion of time-of-flight MRA with true fast imaging with steady-state precession images may provide more information regarding arterial vessel wall

Angiographic Findings

- Shows vessel contour changes, including aneurysmal dilatation and stenosis/occlusion
- Intimal flap and double lumen are specific

Imaging Recommendations

- Best imaging tool
  - MR + MRA or CTA sensitive, relatively specific
  - **Intramural hematoma** is **pathognomonic**
    - Best seen with T1 FS MR and MP-RAGE
  - Conventional angiography is gold standard
    - May be difficult to detect if no change in lumen
- Protocol advice
  - NECT to exclude subarachnoid hemorrhage
  - Whole-brain imaging to evaluate for infarcts
    - Look for posterior circulation infarctions, particularly posterior inferior cerebellar artery territory
  - CTA or MR/MRA of neck and head for evaluation of full extent of VA
  - Conventional angiography if clinical suspicion is high, but noninvasive test negative
  - Where conventional angiography is reference standard, CTA reportedly more sensitive (100%) than either MRA (77%) or Doppler ultrasonography (71%)

DIFFERENTIAL DIAGNOSIS

Extracranial Atherosclerosis

- Multiple vessels with irregular stenoses
- Calcifications of intimal plaque

Fibromuscular Dysplasia

- Multiple, focal strictures ("string of beads") or long segment stenosis

Congenital Hypoplasia of VA

- Osseous transverse foramen of congenitally diminutive VA will be small as well

Miscellaneous Vasculitis

- Stenoses involving multiple, variably sized vessels

PATHOLOGY

General Features

- Etiology
  - **Traumatic VA dissection**
    - Direct arterial injury, such as stabbing
    - Indirect injury ± cervical fractures, probably due to stretch injury
      - Cerebrovascular injuries in 1% of blunt trauma
      - Screen patients with mandible fractures, complex skull fractures, traumatic brain injury with thoracic injuries, scalp degloving, and thoracic vascular injuries
      - Spinal manipulation, yoga, ceiling painting, coughing
      - Systematic review found no conclusive evidence for strong association between neck manipulation and stroke; however, evidence is also absent for no association
  - **Spontaneous VA dissection**
    - Many associations and predisposing factors

        - Hypertension
        - Vascular abnormality: Fibromuscular dysplasia, Marfan syndrome, collagen vascular disease, homocystinuria
        - Viral meningitis: Inflammation-associated injury of arterial wall, leading to weakness of vessel wall and subsequent dissection
    - Pathophysiology unclear
      - Intimal tear with hematoma dissecting into media
      - Hemorrhage from vasa vasorum within media
- Associated abnormalities
    - In 1 study, subarachnoid hemorrhage patients had significantly higher rate of smoking
      - Dissection of dominant VA, steep VA of dissecting side, posterior inferior cerebellar artery involvement, basilar artery bending < 3 mm, pearl-and-string sign

Gross Pathologic & Surgical Features

- Stenoocclusive dissection
    - Dissection to **subintimal** plane with vessel **luminal narrowing or occlusion**
- Dissecting aneurysm
    - Dissection extends into **subadventitial** plane with **dilatation of outer wall**

Microscopic Features

- Intradural VA has no external elastic lamina, thinner adventitia, fewer elastic fibers in media → more prone to aneurysmal dilatation and rupture

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
    - Systematic review showed dizziness/vertigo (58%), headache (51%), and neck pain (46%)
    - Stroke may be presenting feature or develop later
      - Ischemic stroke by thromboemboli at injury site or result of hemodynamic insufficiency due to severe stenosis/occlusion
      - Stroke reportedly common (63%), especially with extracranial dissections (66% vs. 32%)
      - Cerebellar, lateral medulla
      - Symptoms of vertebrobasilar ischemia: Vertigo, hoarseness, dysphagia, and cerebellar dysfunction
    - Transient ischemic attack (14%) and subarachnoid hemorrhage (10%)
      - Subarachnoid hemorrhage seen usually with intracranial dissections (57% vs. 0%)

Demographics

- Age
    - Adults < 45 years
- Epidemiology
    - Dissection is cause of 5-20% of strokes in young patients
    - Internal carotid artery (ICA) dissections 5x more common than VA dissections
      - In 1 study, annual incidence of spontaneous ICA dissection = 2.5-3 per 100,000
      - Annual incidence of spontaneous VA dissection = 1-1.5 per 100,000
    - ≤ 5% of spontaneous SAH due to VA dissecting aneurysm

Natural History & Prognosis

- Systematic review showed good outcome (modified Rankin scale 0-1) in 67% and poor (modified Rankin scale 5-6) in 10%
- Healing of arterial dissections occurs within 3-6 months, with resolution of stenosis seen in 90% and recanalization of occlusions in as many as 50%
- Dissecting aneurysms resolve on follow-up imaging in 5-40%, decrease in size in 15-30%, and remain unchanged in 50-65%
- 1 study showed that most ruptured intracranial VA dissections have dilatation without stenosis or pearl-and-string appearance
    - Primary shape of unruptured dissections: Stenosis-without-dilatation group and pearl-and-string group
- Resolution is more common in VA dissections than in ICA dissections

Treatment

- Stenoocclusive dissection → anticoagulation and antiplatelet agents
    - Current data from nonrandomized studies show no evidence for superiority of anticoagulation or antiplatelet therapy in prevention of stroke after ICA and VA dissection
    - Nonrandomized Cervical Artery Dissection in Stroke Study data shows lower rate of recurrent stroke than reported in some previously
- Dissecting aneurysm → ligation or coil embolization

DIAGNOSTIC CHECKLIST

Consider

- Check other vessels carefully (ICA, other VA) for 2nd or 3rd dissection
- Cerebellar infarction in young to middle-aged adults, need to exclude PICA infarction due to VA dissection

Image Interpretation Pearls

- Evaluate axial CTA carefully for suboccipital rind sign if normal lumen, but high clinical suspicion
- Look for eccentric mural T1 hyperintensity on axial T1 FS

SELECTED REFERENCES

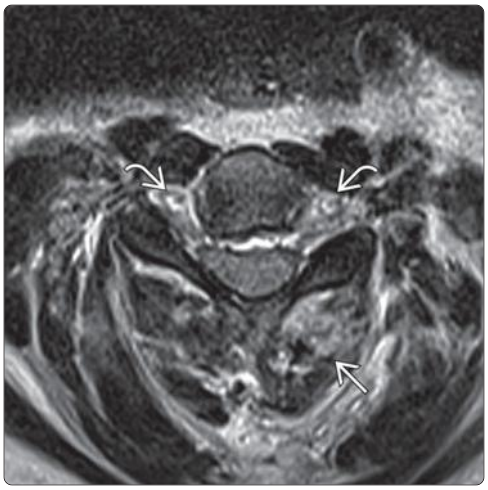
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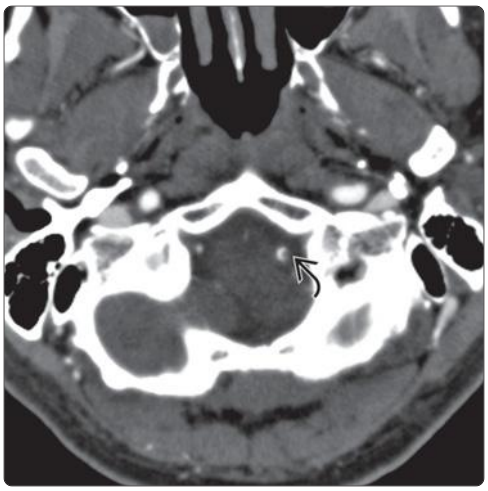


(Left) *Axial T1 MP-RAGE demonstrates T1 hyperintensity in the right V3 arterial segment due to thrombus. Left ICA dissection with mural thrombus is also seen. Spontaneous VA dissection most commonly occurs in the extradural VA.* **(Right)** *Axial CTA in the same patient shows irregularity and narrowing of the right vertebral artery. There is also a nearly complete occlusion of the left ICA due to dissection.*





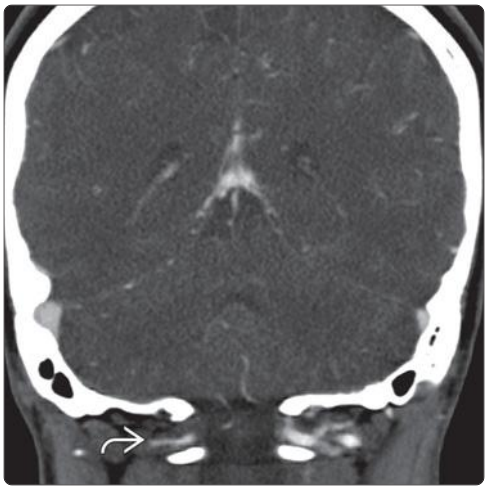
(Left) *Axial CTA reveals no luminal opacification of the vertebral arteries. Severe stenosis or occlusion of the extradural VA may result in brainstem or cerebellar ischemia if there is concomitant dissection of the contralateral VA, which limits collateral flow.* **(Right)** *Axial T2 MR in the same patient shows the lack of normal flow voids in bilateral vertebral arteries. Note the paraspinal muscle edema/hematoma due to blunt trauma.*





(Left) *Axial CTA demonstrates an intraluminal filling defect in the left vertebral artery related to a proximal dissection.* **(Right)** *Anteroposterior DSA in the same patient exhibits intraluminal filling defect related to thrombus. There is irregularity of the proximal V4 segment of the left vertebral artery. Irregularity and luminal narrowing of the V3 segment are related to dissection.*

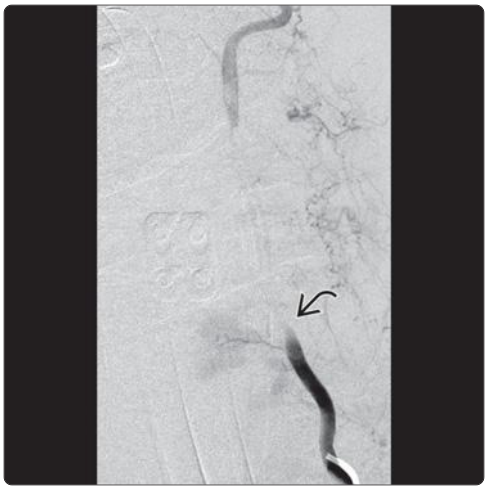
(Left) *Coronal CTA illustrates irregularity and narrowing of the extradural right VA.* **(Right)** *Axial CTA shows narrowing and irregularity of the V3/V4 segment of the left VA. There is subarachnoid hemorrhage (SAH) surrounding the cervicomedullary junction. Patients with intradural dissection usually present with severe headache and SAH, as well as symptoms of possible brainstem or cerebellar ischemia.*



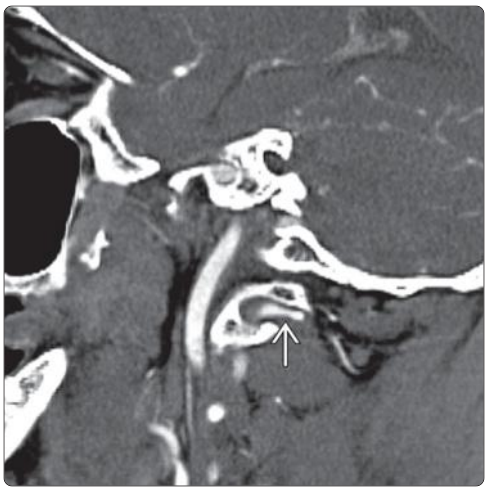


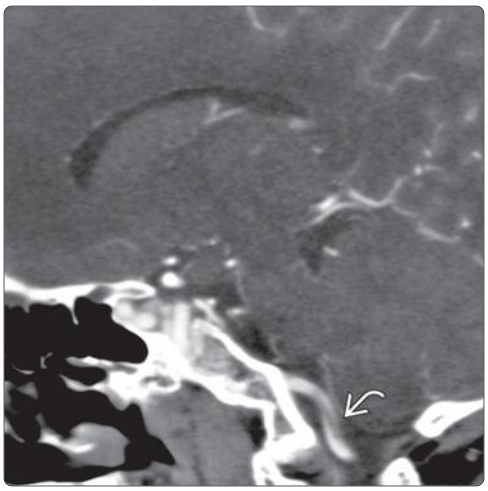
(Left) *Sagittal CTA reveals dissection of the V2 segment of the vertebral artery with a short segment of contrast filling the lumen. There is opacification of the V3 segment from retrograde flow from the basilar artery and possibly from muscular collaterals.* **(Right)** *Anteroposterior DSA with selective left VA injection in the same patient demonstrates tapered morphology of the VA. Conventional angiography is still considered the gold standard method to diagnosis of VA dissection.*

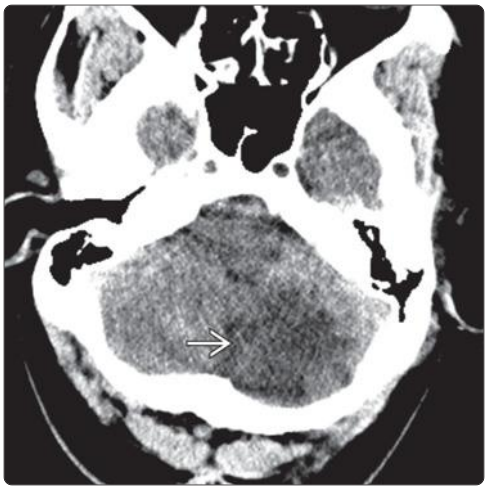


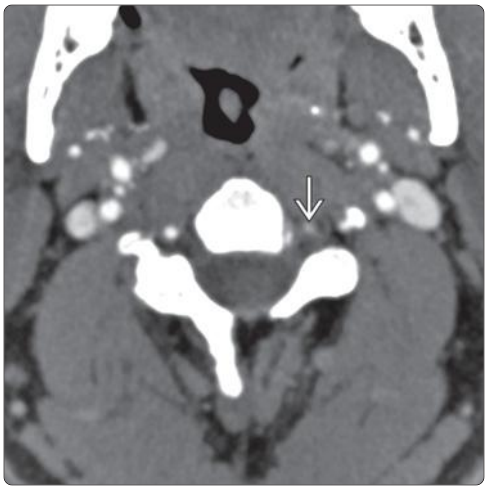


(Left) *Sagittal CTA displays irregularity and narrowing of the VA lumen due to dissection. However, in some cases, the lumen of the segment between the C2 foramen transversaria and foramen magnum may be normal in caliber with the only visible abnormality being dorsal thickening of the arterial wall against the adjacent fat, the "suboccipital rind" sign.* **(Right)** *Sagittal CTA in the same patient reveals extension of the dissection to the intradural V4 segment.*



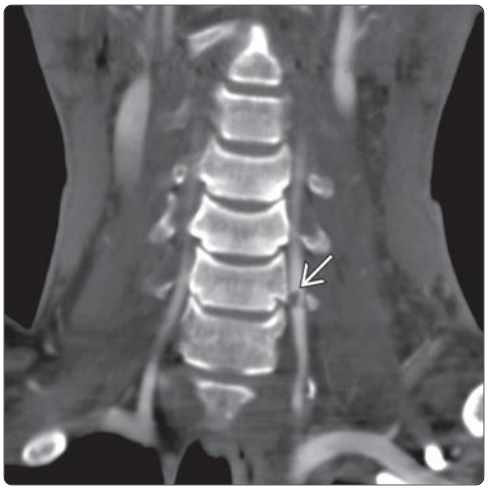




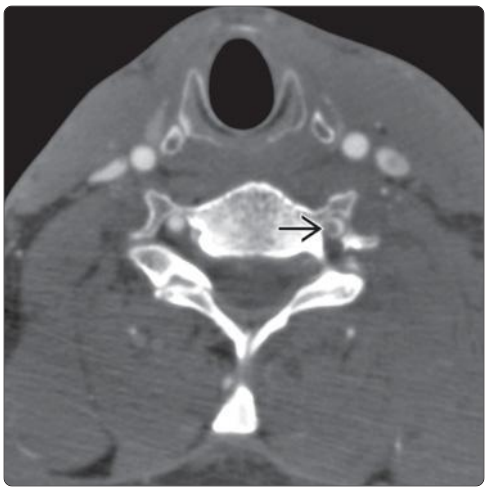


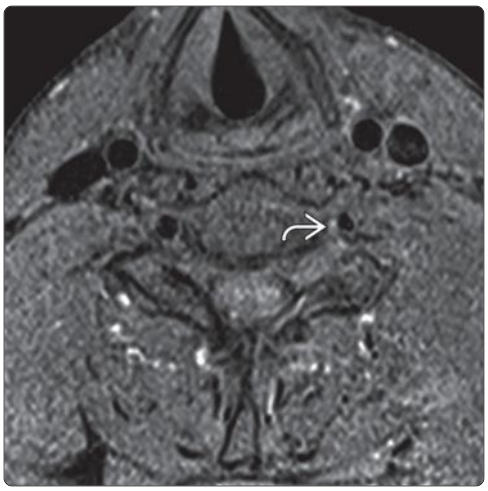
(Left) *Axial NECT shows a wedge-shaped area of hypodensity in the inferior left cerebellar hemisphere in the vascular distribution of the left PICA.* **(Right)** *Axial CTA in the same patient demonstrates luminal irregularity and poor opacification. There is a lateral eccentric hypodensity compatible with mural thrombus. An intraluminal clot can then break off, embolize, and occlude branches anywhere in the posterior circulation distal to the source.*





(Left) *Anteroposterior DSA of the posterior circulation displays tapered morphology of the intradural left VA just proximal to the vertebrobasilar junction. The intradural VA is more susceptible to rupture than the extradural VA, because it has a thicker internal elastic lamina, but no external elastic lamina, a thinner adventitia, and fewer elastic fibers in the media.* **(Right)** *Coronal CT reconstruction depicts intraluminal thrombus related to focal dissection of the left vertebral artery.*





(Left) *Axial CTA from the same patient shows > 50% luminal narrowing of the left vertebral artery due to a dissection-related mural thrombus.* **(Right)** *Axial MP-RAGE in the same patient illustrates irregularity of the left vertebral artery with minimal visible hyperintense mural hematoma. Acute vertebral dissections with only wall hematoma may be difficult to define on MR without the T1 high signal from more chronic blood by-products.*

Osteoporosis

KEY FACTS

TERMINOLOGY

- Skeletal disorder characterized by low bone strength and increased risk of fracture

IMAGING

- Bone scintigraphy and MR are most sensitive to detect osteoporotic spine and sacral insufficiency fractures
- Bone mineral density (BMD)
  - Standard measure for diagnosis of osteoporosis (OP) and assessment of fracture risk
  - WHO T-score cut-off value for OP: -2.5
- Thinning of cortex on radiographs and CT
- Normal or heterogeneous signal intensity (SI) on T1/T2WI MR
- MR features of **benign** osteoporotic fracture
  - Incomplete replacement of bone marrow, spared normal bone marrow SI of vertebral body
  - Multiple compression fractures
  - Retropulsion of posterior bone fragment
  - Low signal intensity band on T1/T2WI MR

TOP DIFFERENTIAL DIAGNOSES

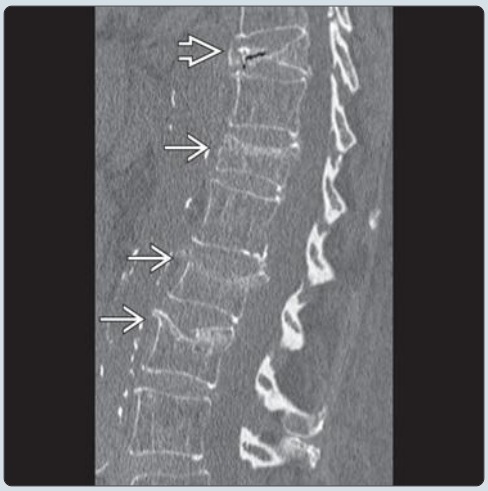
- Lytic or blastic osseous metastases
- Multiple myeloma
- Hyperparathyroidism

PATHOLOGY

- Risk factors
  - Age
  - Postmenopausal
  - Glucosteroids
- Underlying disease → secondary osteoporosis
- Fragility fracture and insufficiency fracture

CLINICAL ISSUES

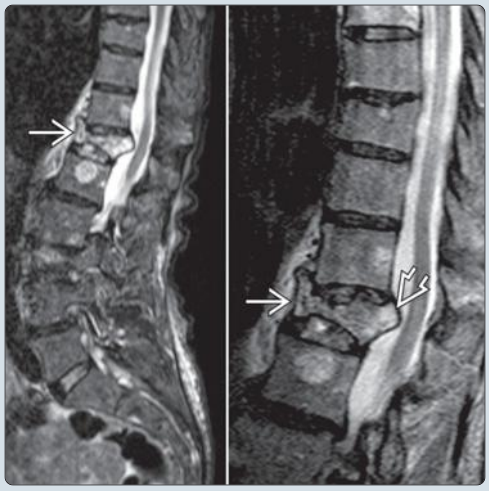
- Asymptomatic bone loss
- Affects up to 40% of postmenopausal women





(Left) *Sagittal NECT shows compression fractures involving multiple levels from L3 inferiorly ➡ to T10 superiorly. The T10 anterior wedging compression fracture shows gas within the body ➡, compatible with Kümmel disease (osteonecrosis). There is endplate sclerosis at all the fracture levels from T12-L3.* **(Right)** *Sagittal T1 MR demonstrates that the L3 fracture is chronic with fatty marrow signal ➡. The L2 and T12 fractures are acute with low signal superior endplates ➡.*





(Left) *Lateral radiograph reveals diffuse osteopenia with thinning of the cortices and accentuated appearance of the bony trabeculae ➡ due to background of demineralized bone.* **(Right)** *Sagittal STIR MR illustrates a benign osteoporotic fracture of the T12 vertebral body ➡ with retropulsion of the posterior cortex ➡. MR findings of metastatic fracture include, convex posterior vertebral body border, abnormal signal of pedicle or posterior elements, epidural or paraspinal mass, and other metastases.*

TERMINOLOGY

Abbreviations

- Osteoporosis (OP), sacral insufficiency fracture (SIF), bone marrow (BM)

Definitions

- Decreased bone mass/bone mineral density (BMD)
- Skeletal disorder characterized by low bone strength and increased risk of fracture

IMAGING

General Features

- Location
  - Vertebral body, sacrum

Imaging Recommendations

- Best imaging tool
  - Bone scan and MR are most sensitive to detect osteoporotic spine and SIFs

Radiographic Findings

- Thinning of cortex
- SIF: Vertical bands of sclerosis oriented parallel to sacroiliac joints (SIJ) in zone 1 of sacrum
  - Cortical disruption ± fracture lines
  - Insensitive for detection of SIF: Only 20–38% of SIFs and pelvic ring fractures identified

CT Findings

- Bone CT
  - Thinning of cortex
  - SIF: Sclerosis in sacral ala and lateral sacrum paralleling SIJ
    - Fracture lines ± bony callous in approximately 75% of patients
    - Useful to determine if fracture lines extend into neural foramina
    - Sensitivity 60-75%

MR Findings

- T1WI
  - Normal or heterogeneous
- T2WI
  - Normal or heterogeneous
- STIR
  - Sensitive to early edema associated with fracture
  - SIF: Hypointense fracture line is usually evident within edema, not seen in 7% of cases
- DWI
  - Hypo- or isointensity suggests acute benign collapse
  - Hyperintensity suggests tumorous nature
  - Apparent diffusion coefficients (ADCs) are lower in metastatic disease than in BM edema
- **MR features of benign osteoporotic fracture**
  - Inhomogeneous T1/T2/STIR signal intensity (SI)
  - Incomplete replacement of BM, spared normal BM SI of vertebral body
  - Multiple compression fractures
  - Retropulsion of posterior bone fragment
  - Low SI band on T1/T2WI MR
  - Fluid sign: OP and minor trauma → osteonecrosis vs. ischemia at site of fractured endplate
- **MR features of malignant fracture**
  - Paravertebral/epidural soft tissue
  - Infiltration of posterior elements
  - Homogeneous replacement of BM by tumor cells
  - Convex posterior margin
  - Other spinal metastases
- **BMD**
  - Standard measure for diagnosis of OP and assessment of fracture risk
  - Dual x-ray absorptiometry → grams of mineral/cm²
    - Related to reference value for young, normal adults of same sex using T-score
      - WHO T-score cut-off value for osteoporosis: -2.5

Nuclear Medicine Findings

- Bone scan
  - Tc-99m–labeled methylene diphosphonate is 1 of the most sensitive examinations for detection of SIFs
    - "Honda" sign or H-pattern in 20–40%
    - Follow-up is variable: Range of uptake pattern from resolution of abnormal activity to stable to worsening activity

DIFFERENTIAL DIAGNOSIS

Lytic Osseous Metastases

- Primary tumor extends to spine with bone destruction > bone production

Blastic Osseous Metastases

- Discrete nodular, mottled, or diffuse areas of sclerosis

Multiple Myeloma

- Multifocal, diffuse, or heterogeneous T1 hypointensity/STIR hyperintensity

Hyperparathyroidism

- Osteopenia, cortical thinning
- Subperiosteal bone resorption

Sacral Traumatic Fracture

- Fractures ± ligament injuries through anterior and posterior portions of pelvis
- Disruption of sacral arcuate lines
- Soft tissue edema and hematoma

PATHOLOGY

General Features

- Etiology
  - Bone remodeling is due to regulated resorption and formation
    - Osteoclasts resorb bone → reversal period, after which osteoblasts fill cavity with collagenous matrix → mineralization of matrix
    - Signals determining replication, differentiation, function, and death of cells of both lineages dictate degree of bone remodeling
  - Wnt/β-catenin signal pathway induces osteoblastogenesis and bone formation

- Bone morphogenic proteins (BMP) stimulate chondrocyte maturation and function
- IGF-1 mediates effects of growth hormone on longitudinal bone growth
  - Needed for skeletal development and maintenance of bone mass
- Postmenopausal OP
  - Estrogen deficiency → excessive bone resorption and bone loss
- Glucosteroids interfere with Wnt signaling, impair osteoblastogenesis, and inhibit osteoblastic function
  - Inhibit IGF-1 synthesis

- Genetics
  - Heritability of BMD estimated between 50-85% in twin studies, with strongest effects in axial skeleton
  - Phenotypes are determined by combined effects of several genes and environmental influences
- Associated abnormalities
  - Fragility fracture: Low-energy fracture from fall no greater than standing height
  - Insufficiency fractures are subtype of stress fracture resulting from normal stress on abnormal bone that has lost its elastic resistance
  - Sacral insufficiency fractures
    - Commonly affects elderly women with osteoporosis
    - Other risk factors: Pelvic radiation, steroid-induced osteopenia, renal osteodystrophy, multiple myeloma
    - Incidence 1-5% in at-risk patient populations
    - Most commonly present with diffuse low back pain ± radiating to buttock, hip, or groin
    - Neurologic symptoms are unusual (5–6%)
    - High incidence of concomitant pelvic insufficiency fractures
- Risk factors for osteoporosis and related fractures
  - History of fracture as adult
  - Low body weight
  - Current smoking
  - Oral corticosteroid therapy
  - Increased age

Staging, Grading, & Classification

- 2 forms of osteoporosis
  - Low turnover: Reduction of bone formative and bone resorptive activities
    - Elderly, following medications (chemotherapy, steroids, bisphosphonate)
  - High turnover: Increased activity of osteoclasts, normal or increased osteoblast activity
    - Most common
    - Postmenopausal females, hyperparathyroidism, transient OP in males
- Secondary OP due to underlying disease
  - Thyrotoxicosis or renal disease
- Disuse OP due to immobilization
  - Bone may have patchy or permeative appearance
- Transient OP
  - Women in 3rd trimester of pregnancy or middle-aged men
  - Self-limited spontaneous onset of pain, progressive over several weeks
  - Radiographic osteopenia involving 1 or several joints, commonly hip

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic bone loss
  - ~ 2/3 vertebral fractures are asymptomatic
- Other signs/symptoms
  - Degradation products in serum and urine
    - Bone formation and resorption marker analysis used to monitor clinical efficacy

Demographics

- Age
  - Typically > 55 years
- Gender
  - Female > male
- Epidemiology
  - Most common metabolic bone disease
  - Affects up to 40% of postmenopausal women

Natural History & Prognosis

- Fracture risk increases 2x for every T-score standard deviation below mean of reference young adult
- Fracture Risk Assessment Tool (FRAX)
  - Predict fractures with use of clinical risk factors ± femoral neck bone mineral density

Treatment

- Considered for patients with low bone mineral density (T-score between -1.0 and -2.5)
- Antiresorptive therapy → reduce bone resorption → stabilize bone architecture, ↓ incidence of fractures
- Anabolic agents → increase bone formation and bone mass
- Acute osteoporotic fracture
  - Strict bed rest and pain control
  - Vertebroplasty/kyphoplasty/sacroplasty

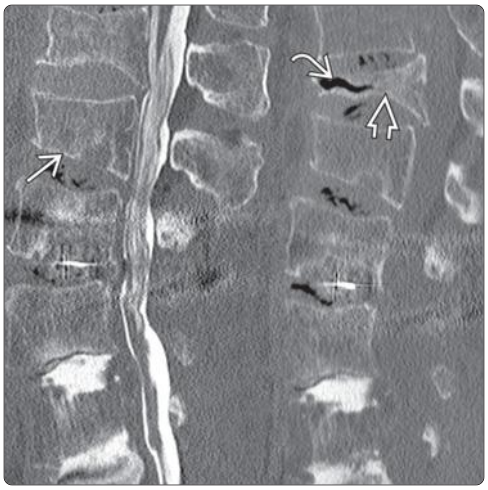
DIAGNOSTIC CHECKLIST

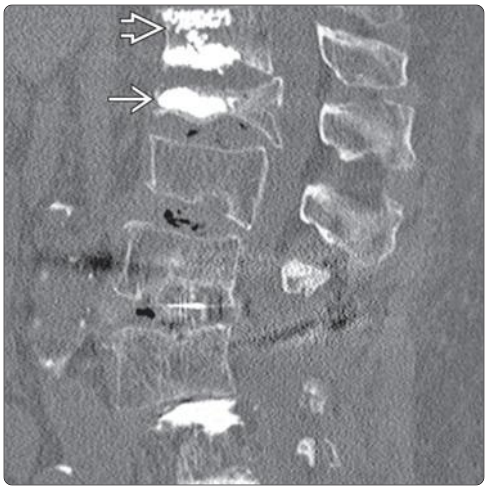
Image Interpretation Pearls

- Increased risk of fractures
  - MR most sensitive to edema related to acute fractures

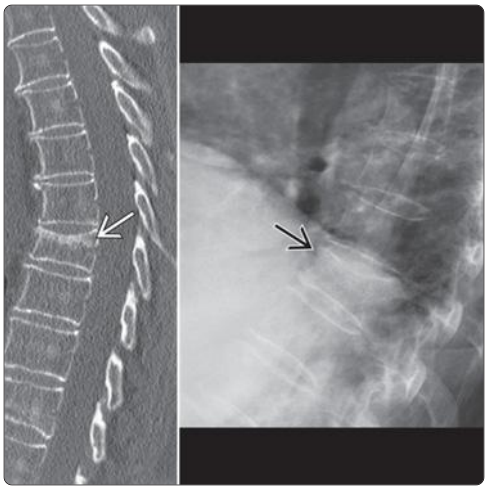
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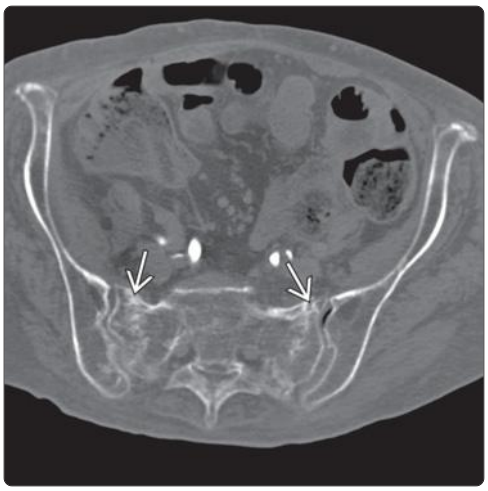
(Left) *Sagittal CT myelogram demonstrates diffuse osteopenia and mild anterior wedging of the L2 vertebral body. Within 1 year, the patient developed an osteoporotic compression fracture of L1. There is an inverse correlation between the presence of an intravertebral vacuum and bone mineral density.* **(Right)** *Sagittal CT reconstruction shows hyperdense methylmethacrylate within the L1 and T12 bodies after percutaneous vertebroplasty procedure.*





(Left) *Sagittal CT and correlative radiograph depict an osteoporotic fracture of a midthoracic vertebral body.* **(Right)** *Sagittal STIR MR reveals hyperintensity within a midthoracic vertebral body due to edema from an acute osteoporotic compression fracture. T1WI shows hypointensity and a low SI band related to the fracture line. Inhomogeneous T1/T2/STIR SI and incomplete marrow replacement are features of benign fractures.*





(Left) *Sagittal T1WI MR illustrates hypointensity parallel to the endplate. STIR MR shows hyperintensity in the corresponding region and a fluid sign. Fluid sign is more often seen in acute benign osteoporotic fractures. Osteoporosis and minor trauma result in osteonecrosis at the site of the fractured endplate. Fluid is pressed into the space of osteonecrosis and causes the MR "fluid" sign.* **(Right)** *Axial CT exhibits bilateral sacral insufficiency fractures. Minor or no antecedent trauma is reported in 2/3 of patients.*

Paget Disease

KEY FACTS

TERMINOLOGY

- Osteitis deformans
- Chronic metabolic disorder of abnormal bone remodeling in adult skeleton

IMAGING

- Enlarged vertebra with trabecular coarsening and cortical thickening
  - Both vertebral body, neural arch involved
- Lumbar spine most common
- "Picture frame" vertebra
  - Central osteopenia
  - Coarse and sclerotic peripheral trabecular pattern
- Diffusely dense "ivory" vertebra
- Fibrovascular marrow in active phase
- Fatty marrow in mixed phase
- Blastic inactive phase
  - Marrow space low T1/T2 signal representing sclerosis-fibrosis

TOP DIFFERENTIAL DIAGNOSES

- Osteoblastic metastases
- Vertebral hemangioma

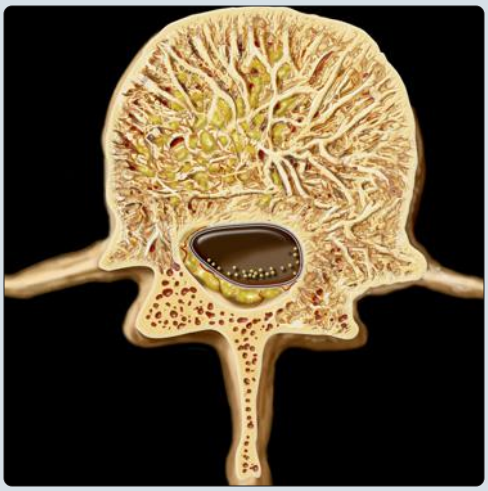
PATHOLOGY

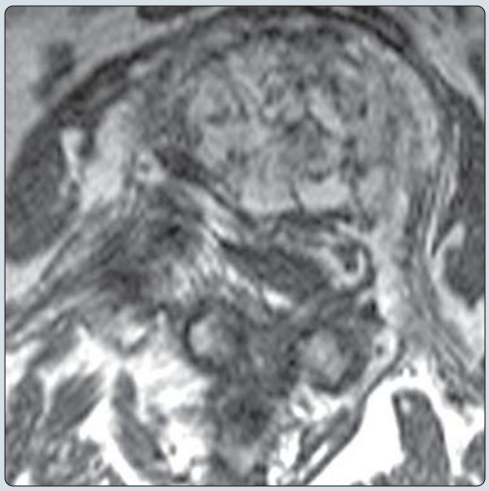
- Sites of osseous involvement
  - Pelvis = spine > femur > skull > tibia > clavicle > humerus > ribs
  - Polyostotic, asymmetric > monostotic
- Possible viral etiology
  - Measles virus of Paramyxovirus family found in osteoclasts

CLINICAL ISSUES

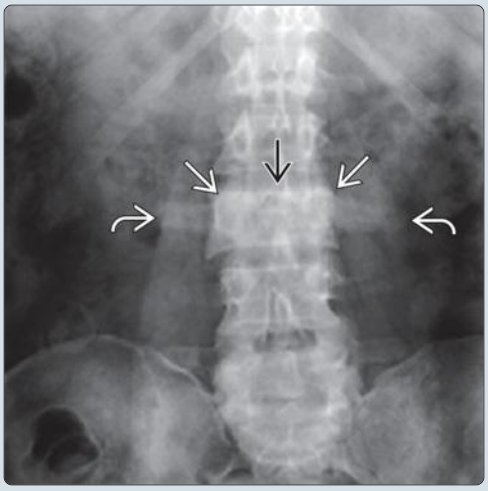
- 20% asymptomatic
- Deep, dull bone pain
- Sarcomatous transformation: < 1%
- Myelopathy, cauda equina syndrome from canal narrowing

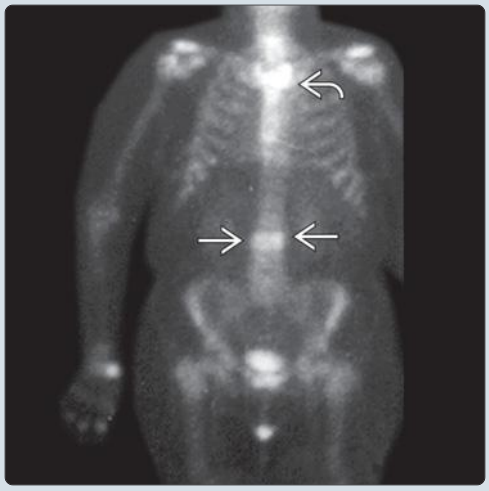
(Left) *Axial graphic of pagetic vertebra demonstrates an enlarged vertebral body, pedicles, and left facet with trabecular thickening and increased fatty marrow. PD is a chronic metabolic disorder of abnormal bone remodeling and may virtually affect every bone in the skeleton.* **(Right)** *Axial T1WI MR through a lumbar vertebra shows coarse and irregular trabeculae with mild vertebral expansion. The pagetic bone marrow contains fatty areas with a heterogeneous distribution.*





(Left) *Anteroposterior radiograph depicts an enlarged L3 vertebral body, pedicles, and transverse processes. Increased density gives the "ivory" vertebra appearance. The coarse and sclerotic peripheral trabecular pattern and central osteopenia gives the "picture frame" appearance.* **(Right)** *Anteroposterior bone scan reveals ↑ uptake in the L3 vertebra. Increased uptake in the sternum is suspicious for PD. Bone pain is common with lumbar spine.*





TERMINOLOGY

Abbreviations

- Paget disease (PD)

Synonyms

- Osteitis deformans

Definitions

- Chronic metabolic disorder of abnormal bone remodeling in adult skeleton

IMAGING

General Features

- Best diagnostic clue
  - Enlarged vertebra with trabecular coarsening and cortical thickening
- Location
  - Lumbar spine most common: L3, L4
- Size
  - Single or multiple levels
- Morphology
  - Vertebra expanded, "squared"
    - Loss of anterior concave margin
    - Thickened cortex
  - Coarse trabeculae in vertical orientation
  - Both vertebral body, neural arch involved

Radiographic Findings

- Radiography
  - "Picture frame" vertebra
    - Coarse and sclerotic peripheral trabecular pattern
    - Central osteopenia
  - Diffusely dense "ivory" vertebra
  - Rare solitary discrete lytic lesion

CT Findings

- Bone CT
  - Better characterization of sclerotic cortex and disorganized trabeculae
  - Evaluate risk for vertebral body collapse

MR Findings

- T1WI
  - Hypointense cortex + thickened trabeculae
- T2WI
  - Cortex hypointense
- STIR
  - Hypointense bone
- Variable degree of spinal stenosis + foraminal narrowing
- Marrow pattern
  - Fibrovascular marrow in active phase
    - Heterogeneous but predominantly hypointense on T1WI, hyperintense on T2WI
    - Foci of fatty marrow
    - Post-gadolinium enhancement
  - Fatty marrow in mixed phase
    - Hyperintense on T1WI and T2WI
    - Most often imaged
    - May be increased compared to normal vertebrae
  - Blastic inactive phase
    - Marrow space low T1/T2 signal representing sclerosis-fibrosis

Nuclear Medicine Findings

- Bone scan
  - Diffusely increased uptake in enlarged vertebra
  - May be normal in inactive phase
    - Radiographic abnormality persists
- PET
  - Most cases not hypermetabolic
    - Low- to high-grade uptake without sarcomatous transformation has been reported

Imaging Recommendations

- Best imaging tool
  - Radiography cost effective
  - CT: Extent of osseous involvement and degree of cancellous and cortical bone loss
  - MR: Evaluation of epidural space and neural structures
  - Bone scan: Determine disease extent, assess disease activity, monitor treatment response

DIFFERENTIAL DIAGNOSIS

Osteoblastic Metastases

- Multifocal, patchy, or diffuse
- Hyperintense on T2WI with fat suppression or STIR
- Post-gadolinium enhancement
- May be indistinguishable if "ivory" vertebra present
  - Lymphoma, metastases
- Epidural, paravertebral soft tissue mass common
- Pathologic compression fractures

Vertebral Hemangioma

- Typically hyperintense on T1WI and T2WI
  - Post-gadolinium enhancement
- Stippled or honeycomb appearance on axial CT
- No vertebral enlargement or cortical thickening

PATHOLOGY

General Features

- Etiology
  - Possible viral etiology
    - Measles virus of paramyxovirus family found in osteoclasts
    - Upregulation of interleukin-6 and its receptor gene reported
- Genetics
  - 40% with family history of disease
    - Majority of cases harbor germline mutations in the sequestosome 1 gene (*SQSTM1*)
    - *SQSTM1* mutations also in sporadic PD
  - Valousin-containing protein mutation
- Associated abnormalities
  - Other sites of osseous involvement
    - Pelvis = spine > femur > skull > tibia > clavicle > humerus > ribs
    - Polyostotic, asymmetric > monostotic
  - Pathologic compression fracture
  - Kyphoscoliosis
  - Spondylosis

- Basilar invagination
  - Narrowing at foramen magnum due to cervical spine prolapsing into pagetic calvarium
- Cord ischemia
  - Hyperemic bone shunts blood away from spinal cord → hypoxia
    - Pagetic bone with ↑ blood flow contains 6x more vessels per cross-sectional area than normal bone
  - Vascular compression
- Extramedullary hematopoiesis
- Sarcomatous transformation: < 1%
  - Increased risk with polyostotic disease
  - Hip, pelvis, shoulder most common
  - Uncommon in skull and spine relative to high prevalence of disease in axial skeleton
  - Osteosarcoma: 50-60%
    - Mostly in older persons with longstanding polyostotic disease
    - Pagetic osteosarcoma tends to spare spine, more common in pelvis, femur, humerus, and skull
  - Fibrosarcoma/MFH: 20-25%
  - Chondrosarcoma: 10%
  - Histologic distinction of secondary sarcoma not critical due to high degree of anaplasia
  - Osseous destruction, enhancing soft tissue mass with necrosis, metastases

Microscopic Features

- Active phase: Osteolytic phase
  - Aggressive bone resorption
  - Giant osteoclasts with multiple nuclei and inclusion bodies
  - Osteoclastic > osteoblastic activity
  - Replacement of hematopoietic bone marrow by fibrous connective tissue
  - Increased vascular channels
- Mixed blastic-lytic phase
  - ↑ osteoblastic activity eventually surpasses bone resorption
  - Disorganized trabecular pattern
    - "Mosaic" or "jigsaw": Multiple remodeling lines
  - "Pumice" bone
    - Weak bone lacking normal interconnections
- Inactive quiescent phase
  - ↓ bone turnover
    - Diminishing osteoblasts, osteoclasts
  - Loss of excessive vascularity
  - Fatty marrow

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Deep, dull bone pain
  - Other signs/symptoms
    - 20% asymptomatic
    - Myelopathy, cauda equina syndrome from canal narrowing
    - Radiculopathy from foraminal stenosis
    - ↑ serum alkaline phosphatase, urine hydroxyproline
    - Involvement of cervical spine can lead to cervical cord compression and cervical myelopathy
      - ± craniocervical instability 2° to pathologic C2 fracture
      - Basilar impression with brain stem compression
- Clinical profile
  - Sarcomatous transformation
    - Severe pain at site of previous asymptomatic disease with large lytic lesion + soft tissue mass

Demographics

- Age
  - 55-85 years
    - Rare < 40 years
- Gender
  - M:F = 3-4:2
- Ethnicity
  - Caucasians of Northern European descent
  - Rare in Asians and African Americans
- Epidemiology
  - 3-4% of adults over 40 years
  - 10-11% of adults over 80 years

Natural History & Prognosis

- Disease process may be halted with return of normal osseous architecture after medical treatment
- Poor prognosis with sarcomatous degeneration
  - 3-year mortality rate: > 90%

Treatment

- Analgesic medications
- Medications to inhibit osteoclastic bone resorption and relieve pain
  - Bisphosphates: Alendronate (Fosamax), pamidronate (Aredia), etc.
  - Calcitonin
  - Mithramycin
- Surgical decompression of spinal stenosis

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Expanded, square vertebra with cortical thickening and central lucency ("picture frame") highly suggestive of Paget disease

SELECTED REFERENCES

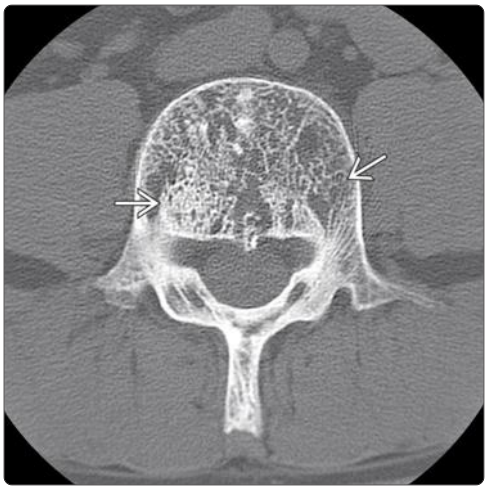
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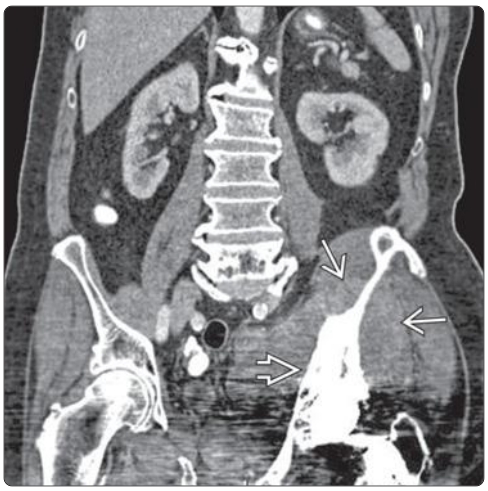
(Left) *Sagittal T1WI MR depicts the fibrovascular marrow phase of PD with heterogeneous but predominantly T1 hypointense signal. Overactive osteoclasts and osteoblasts result in abnormal and disordered bone remodeling. The newly formed bone is abnormally enlarged, deformed, and weak.* **(Right)** *Sagittal T2WI MR exhibits the fibrovascular marrow phase of PD with heterogeneous but predominantly T2 hyperintense signal.*





(Left) *Sagittal CT reveals cortical thickening of the L4 vertebral body and facet. There is also mild expansion of the posterior elements. The disease seldom appears before the age of 40, but its prevalence tends to double each decade from the age of 50 onwards, reaching about 10% after the 9th decade.* **(Right)** *Axial CT illustrates sclerotic cortex and disorganized trabeculae. A therapeutic approach is based on 2nd generation bisphosphonates, which are used when bone alkaline phosphatase is high.*





(Left) *Sagittal T1WI MR shows heterogeneous signal from the S1 and S2 bodies due to fat, thickened, and dark trabeculae. There is no soft tissue component. Complications depend on the skeletal sites involved and range from secondary osteoarthritis to malignant degeneration.* **(Right)** *Coronal CT demonstrates a soft tissue mass about an enlarged, thickened left iliac bone. Sarcomatous transformation of the lesion is rare (< 1% of cases).*

Hyperparathyroidism

KEY FACTS

TERMINOLOGY

- Increased levels of hyperparathyroid hormone

IMAGING

- Osteopenia
  - Resorption of secondary trabeculae
    - Interlinking, non-weight-bearing trabeculae
  - Accentuation of primary trabeculae
    - Weight-bearing trabeculae
  - Cortical thinning
- Subperiosteal bone resorption
- Sacroiliac joint erosions
- Rare: Vertebral endplate erosions

TOP DIFFERENTIAL DIAGNOSES

- Multiple myeloma
- Renal osteodystrophy
- Senile osteoporosis
- Seronegative spondyloarthropathy

PATHOLOGY

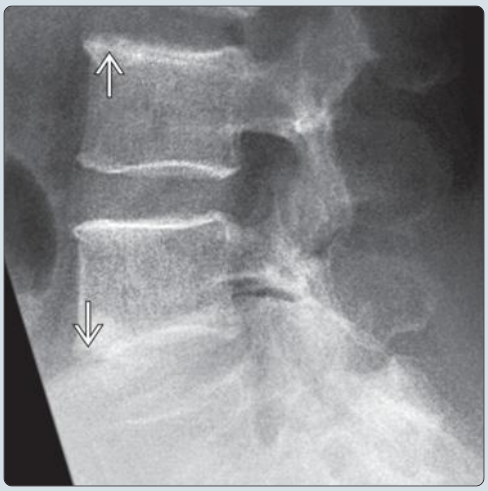
- Primary
  - Parathyroid adenoma, hyperplasia, or carcinoma
- Secondary
  - Due to renal failure, or rarely intestinal malabsorption
- Tertiary
  - Longstanding secondary hyperparathyroidism (HPT) leads to autonomous function of parathyroid glands

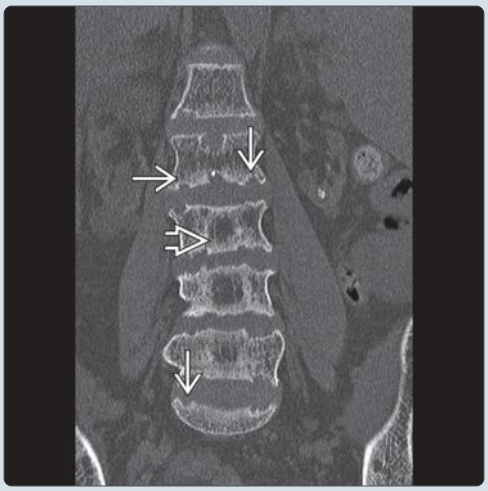
CLINICAL ISSUES

- Bone pain
- Renal colic
- Insufficiency fractures

DIAGNOSTIC CHECKLIST

- Hand radiographs useful to corroborate diagnosis





(Left) *Lateral radiograph shows diffuse, nonspecific osteopenia. Endplate erosions ⇨ are a clue to diagnosis but can also be seen in infection, CPPD, renal osteodystrophy, or due to disc degeneration. Involvement of 2 noncontiguous levels in this case makes infection unlikely.* **(Right)** *Coronal bone CT shows multiple endplate erosions due to hyperparathyroidism (HPT) ⇨. Paucity of trabeculae ⇨ in central portions of vertebral bodies can be distinguished from tumor because of their normal, fatty attenuation.*





(Left) *Sagittal bone CT shows severe hyperparathyroidism in a 50-year-old patient. Primary (vertical) trabeculae are prominent due to resorption of secondary trabeculae. There are prominent Schmorl nodes and an insufficiency fracture of T12 ⇨.* **(Right)** *Coronal bone CT in the same patient shows sacroiliac joint erosions ⇨. These are a common finding in HPT but may lead the unwary to an erroneous diagnosis of ankylosing spondylitis.*

TERMINOLOGY

Abbreviations

- Hyperparathyroidism (HPT)

Definitions

- Increased levels of hyperparathyroid hormone
- Osteitis fibrosa cystica: Florid bony changes due to longstanding HPT
- Brown tumor: Reactive giant cell lesion due to HPT
- HPT may be primary, secondary, or tertiary

IMAGING

General Features

- Best diagnostic clue
  - Osteopenia
- Location
  - Peripheral skeleton > spine

Radiographic Findings

- Radiography
  - Osteopenia
    - 30-50% bone loss necessary to detect osteopenia
  - Resorption of secondary trabeculae (interlinking, non-weight-bearing trabeculae)
  - Accentuation of primary trabeculae (weight-bearing trabeculae)
  - Cortical thinning
    - Sharp distinction between cortex, medullary cavity
  - Subperiosteal bone resorption
  - Sacroiliac joint erosions
  - Schmorl nodes
  - Rare: Vertebral endplate erosions
  - Rare: Brown tumor (osteoclastoma)
    - Lytic, expansile lesion without matrix production
    - Histologically identical to giant cell tumor
  - Soft tissue calcifications
    - Usually involve extremities

CT Findings

- Bone CT
  - More sensitive than radiographs to detect osteopenia, bone resorption

Nuclear Medicine Findings

- Bone scan
  - Increased bone turnover

Other Modality Findings

- Bone densitometry most sensitive in detection of osteopenia

DIFFERENTIAL DIAGNOSIS

Multiple Myeloma

- Most frequently presents as diffuse osteopenia of spine
- Marrow replacement visible on CT, MR

Renal Osteodystrophy

- Secondary HPT plus osteomalacia, bone sclerosis
- "Rugger jersey" spine, blurred trabeculae

Senile Osteoporosis

- Cortical thinning, resorption primary bone trabeculae

Seronegative Spondyloarthropathy

- Erosions of sacroiliac joints and vertebral corners
- Syndesmophytes

PATHOLOGY

General Features

- Etiology
  - Primary
    - Parathyroid adenoma, hyperplasia, or carcinoma
  - Secondary
    - Due to renal failure, or rarely intestinal malabsorption
    - Hyperplasia of parathyroid glands
  - Tertiary
    - Longstanding secondary HPT leads to autonomous function of parathyroid glands
- Associated abnormalities
  - Hypophosphatemia, renal calculi, peptic ulcer disease, pancreatitis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Bone pain, renal colic

Natural History & Prognosis

- Many cases remain mild, stable

Treatment

- Options, risks, complications
  - Major risk parathyroidectomy: Recurrent laryngeal nerve injury
- Mild cases can be followed conservatively
- Bone mineral density may not normalize after parathyroidectomy

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Hand radiographs useful to corroborate diagnosis

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Renal Osteodystrophy

KEY FACTS

TERMINOLOGY

- **Chronic kidney disease-mineral and bone disorder** (CKD-MBD): Constellation of abnormalities seen in progressive kidney disease, including metabolic abnormalities, disturbances in bone modeling and remodeling, and extraskeletal calcification in arteries and soft tissues
- **Renal osteodystrophy** (OD): Only defines the bone pathology associated with CKD

IMAGING

- Rugger jersey spine
- Retarded skeletal age
- Pathologic fracture
- Changes of osteomalacia/rickets
  - Blurred, coarse bone trabeculae
- Changes of hyperparathyroidism (HPT)
  - Prominent primary trabeculae
  - Resorption of secondary trabeculae
  - Cortical thinning
  - Erosions at entheses, endplates, sacroiliac (SI) joints
  - Brown tumors: Lytic lesion, no matrix, sharp zone transition

TOP DIFFERENTIAL DIAGNOSES

- Primary hyperparathyroidism
- Osteomalacia and rickets
- Dialysis arthropathy
- Osteopetrosis
- Paget disease
- Discogenic sclerosis

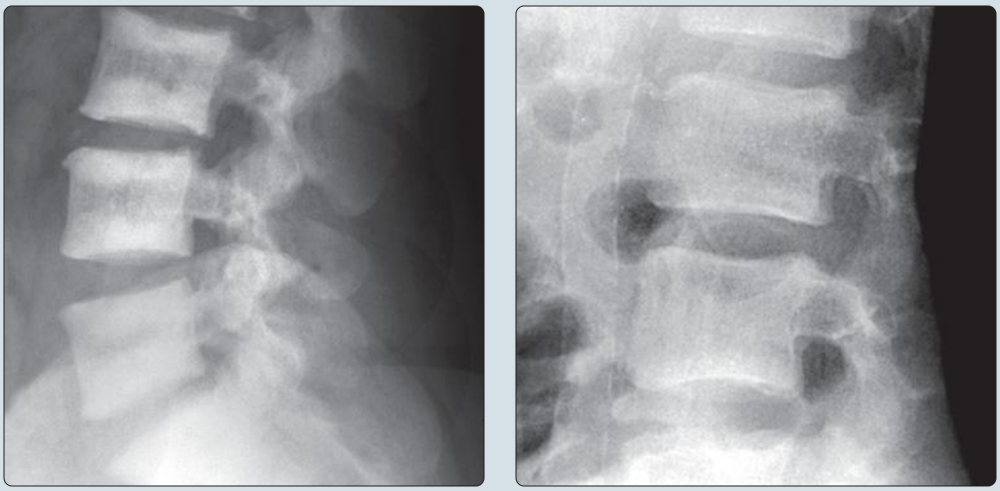
PATHOLOGY

- Secondary hyperparathyroidism
  - ↑ parathyroid hormone (PTH) → ↑ bone resorption
- Osteomalacia
- Soft tissue calcifications

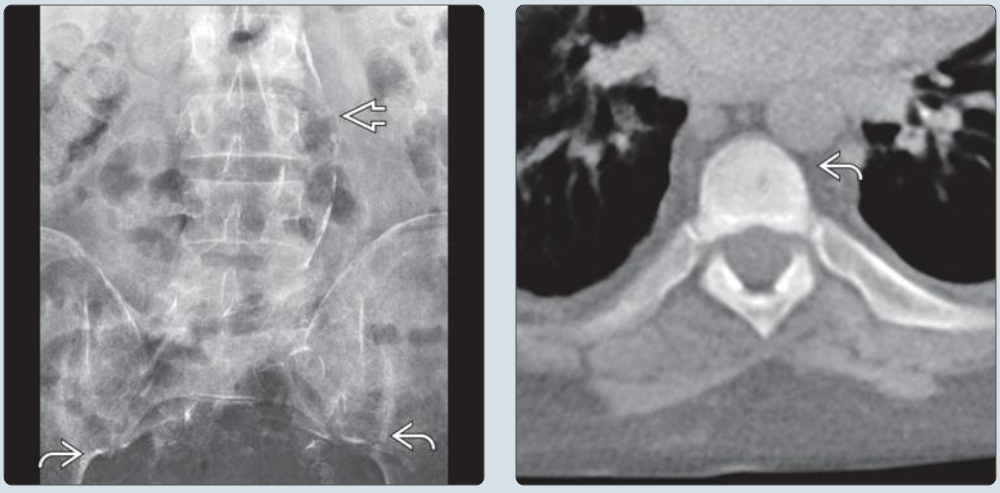
CLINICAL ISSUES

- Asymptomatic unless pathologic fracture occurs

(Left) *Lateral radiograph shows severe findings of renal osteodystrophy. There is marked sclerosis of the vertebral bodies adjacent to the endplate creating a striped appearance like a rugby jersey. The margins of the sclerosis are poorly defined, in distinction to the sharply demarcated sclerotic regions seen in osteopetrosis.* **(Right)** *Lateral radiograph shows fairly mild changes of renal osteodystrophy (OD). Ill-defined sclerosis is present adjacent to the endplates, and bony trabeculae are blurred.*



(Left) *Anteroposterior radiograph in the same patient shows sacroiliac erosions due to 2° hyperparathyroidism. Accelerated arterial calcifications are due to the abnormal calcium/phosphate levels.* **(Right)** *Axial bone CT through the thoracic spine shows sclerosis and blurred trabeculae due to renal osteodystrophy. The appearance is more uniform and diffuse than that of sclerotic metastases.*



TERMINOLOGY

Definitions

- **Chronic kidney disease-mineral and bone disorder** (CKD-MBD): Constellation of abnormalities seen in progressive kidney disease, including metabolic abnormalities, disturbances in bone modeling and remodeling, and extraskeletal calcification in arteries and soft tissues
- **Renal osteodystrophy** (OD): Only defines bone pathology associated with CKD

IMAGING

General Features

- Best diagnostic clue
  - Rugger jersey spine
- Location
  - Involves axial and appendicular skeleton

Radiographic Findings

- Radiography
  - Osteosclerosis &/or osteopenia
  - Rugger jersey spine
    - Bands of sclerosis paralleling endplates
    - Sclerotic bands have hazy, poorly defined margins
  - Changes of hyperparathyroidism (HPT)
    - Prominent primary trabeculae
    - Resorption of secondary trabeculae
    - Cortical thinning
    - Erosions at entheses, endplates, sacroiliac (SI) joints
    - Subperiosteal bone resorption
    - Brown tumors: Lytic lesion, no matrix, sharp zone transition
  - Changes of osteomalacia/rickets
    - Blurred, coarse bone trabeculae
    - Children: Fraying, cupping metaphyses
  - Retarded skeletal age
  - Pathologic fracture

MR Findings

- Bone marrow has heterogeneous, nonspecific appearance
- Brown tumor
  - T1WI: Low signal intensity focal mass
  - T2WI: High signal intensity focal mass

Nuclear Medicine Findings

- Bone scan may show diffusely increased uptake ("superscan")

Imaging Recommendations

- Best imaging tool
  - Radiography

DIFFERENTIAL DIAGNOSIS

Primary Hyperparathyroidism

- Osteopenia, resorption of secondary trabeculae
- Erosions of vertebral endplates, sacroiliac joints

Osteomalacia and Rickets

- Blurring of trabeculae
- Loss of distinction between cortex, medullary bone

Dialysis Arthropathy

- Arthritis due to deposition of crystals or amyloid
- Only in patients on dialysis, usually hemodialysis
- Endplate erosions, destruction
- Soft tissue masses due to amyloid or crystal deposits

Osteopetrosis

- Sharply demarcated sclerotic bands adjacent to endplates

Paget Disease

- Thickening of cortex around vertebra ("picture frame")
- Thickened, disorganized trabeculae

Discogenic Sclerosis

- Trabeculae have normal appearance
- Osteophytes, disc space narrowing also present

PATHOLOGY

General Features

- Etiology
  - Secondary HPT
    - Phosphate retention by kidneys → ↓ serum calcium → ↑ parathyroid hormone (PTH)
    - ↑ PTH → ↑ bone resorption
  - Osteomalacia
    - Failure to convert vitamin D to active form
    - Aluminum intoxication historically contributed to osteomalacia
      - Change in dialysate has eliminated this problem
- Associated abnormalities
  - Soft tissue calcifications

CLINICAL ISSUES

Presentation

- Asymptomatic unless pathologic fracture occurs

Demographics

- Epidemiology
  - Common in chronic renal failure

Natural History & Prognosis

- Increased fracture risk
- Hemodialysis may result in hemodialysis arthropathy

Treatment

- Vitamin D, dietary restriction, phosphate binders

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Hyperplastic Vertebral Marrow

KEY FACTS

TERMINOLOGY

- Physiologic process where fatty marrow is converted to red marrow in response to systemic stress

IMAGING

- Location: Calvarium, spine, ribs, pelvis initially involved
  - Followed by extremities
  - Reverse comparison to maturation
- T1WI: Iso- to hypointense signal intensity (SI) relative to disc
- T2WI: Intermediate to low SI
- STIR: Similar, but not hyperintense, compared to muscle
- Mild post-gadolinium enhancement

TOP DIFFERENTIAL DIAGNOSES

- Normal hematopoietic marrow
  - Children up to ~ 10 years
- Marrow infiltration/replacement
  - Leukemia/lymphoma
  - Multiple myeloma/metastases
- Marrow edema
  - Trauma
  - Osteomyelitis

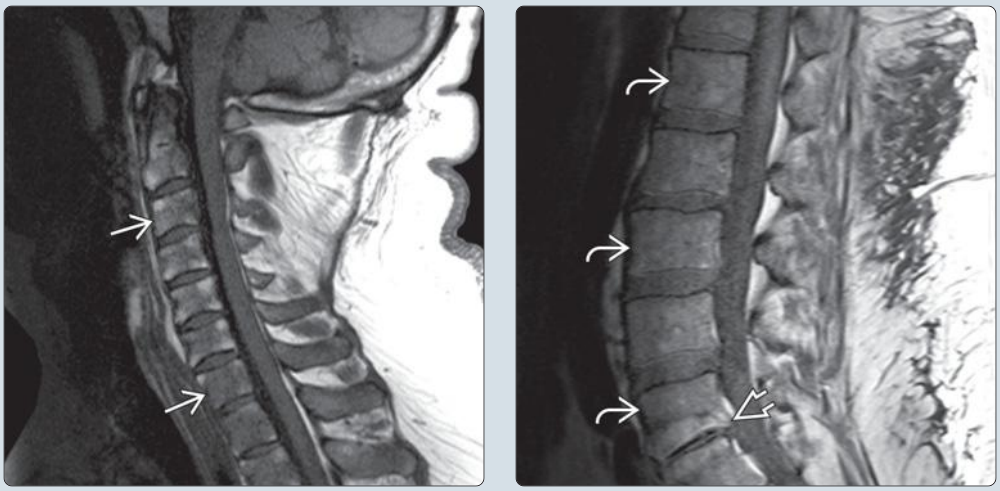
PATHOLOGY

- Iron deficiency anemia
- Anemia of chronic disease
- Hemolytic anemia
- Cyanotic heart disorder, heart failure
- Hematopoietic growth factors

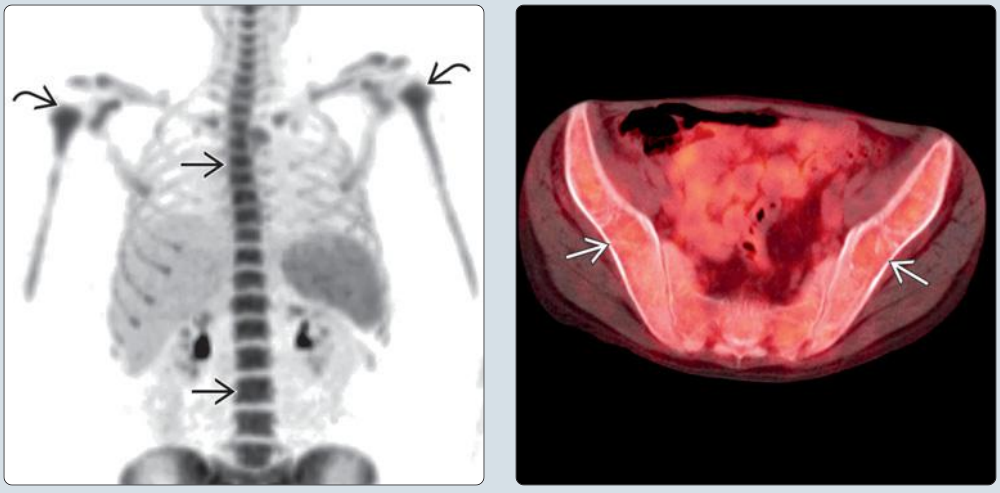
DIAGNOSTIC CHECKLIST

- Conversion of red to yellow marrow occurs in 1st 2 decades of life
  - Adult marrow pattern by 25 years of age
  - From peripheral and distal to central and proximal
- Diffuse marrow T1 hypointensity without marked post-gadolinium enhancement or cortical disruption suggests hyperplastic marrow

(Left) *Sagittal T1WI MR demonstrates heterogeneous SI in the cervical marrow with areas of hypointensity due to marrow reconversion. Focal alterations in SI may reflect local variation in the amount of normal expected vertebral components, including fat and hematopoietic cells, bone, and vessels.* **(Right)** *Sagittal T1WI MR reveals heterogeneous hypointensity 2° to chronic hypoxemia and anemia in this obese patient. Degenerative disc and fatty endplate changes are seen at L5-S1.*



(Left) *Coronal PET exhibits diffusely ↑ uptake in the axial and appendicular skeleton. In cancer patients, hematopoietic marrow hyperplasia can be encountered after granulocyte colony-stimulating factor administration, which is used minimize treatment-related myelosuppression.* **(Right)** *Axial PET/CT illustrates expansion of the medullary space of bilateral iliac bones with diffusely increased uptake in this patient with thalassemia major 1.*



TERMINOLOGY

Synonyms

- Marrow reconversion

IMAGING

General Features

- Best diagnostic clue
  - Vertebral marrow iso- to hypointense relative to intervertebral discs (hyperintense on T1WI)
- Size
  - Entire spinal column typically affected
- Morphology
  - Diffuse, homogeneous
  - Patchy, geographic

Radiographic Findings

- Radiography
  - Hemolytic anemias
    - Medullary expansion in ribs, calvarium, long bones
    - Cortical thinning
    - "Hair on end" striations in calvarium
    - Biconcave vertebrae in sickle cell anemia

CT Findings

- Bone CT
  - H-shaped vertebrae due to endplate infarction in sickle cell anemia
  - Expansion of medullary cavity in thalassemia

MR Findings

- T1WI
  - Iso- to hypointense signal intensity (SI) to disc
    - Similar, or slightly hyperintense, to muscle
    - Normal vertebral marrow SI typically higher than muscle
  - Marrow SI < skeletal muscle or disc on T1WI is **not** normal hematopoietic bone marrow at 1.5T
  - Highest diagnostic accuracy using SI of muscle at 3T as calibration standard
- T2WI
  - Intermediate to low SI
    - Similar to, but not higher than, muscle on fat-suppressed T2WI
- STIR
  - Similar, but not hyperintense, compared to muscle
  - Hematopoietic marrow SI > fatty marrow SI
- DWI
  - Hematopoietic marrow diffusion > fatty marrow diffusion
  - Diffusion decreases in skeleton with age
- T1WI C+
  - Mild post-gadolinium enhancement
    - Normal marrow SI should not ↑ by > 35% in adults > 35 years
  - Bone marrow in vertebral bodies displays variable behavior on chemical shift MR
    - ↓ in SI > 20% on out-phase compared with in-phase images: Threshold to differentiate between benign and malignant vertebral marrow abnormalities

Nuclear Medicine Findings

- Bone scan
  - Tc-99m diphosphonate or Tc-99m sulfur colloid
    - Sites of ↑ uptake due to marrow expansion
      - Distal appendicular skeleton, calvarium
- PET
  - Diffusely ↑ uptake of FDG seen with granulocyte colony-stimulating factor (GCSF) treatment
    - ↑ uptake and accumulation of FDG in normal, but stimulated cells due to upregulation of glucose transport and metabolism
  - Focal marrow hyperplasia may show slight increase in uptake in comparison to adjacent red marrow on FDG PET

Other Modality Findings

- Iron oxide-enhanced MR marrow imaging
  - Normal and hypercellular reconverted marrow accumulate supermagnetic iron oxides
    - Significant signal loss on STIR 45-60 minutes after iron oxide infusion
  - May be useful in distinguishing tumor infiltration from marrow reconversion

Imaging Recommendations

- Best imaging tool
  - MR modality of choice in marrow imaging
    - High sensitivity in detecting increased marrow cellularity on T1WI
- Protocol advice
  - Pre-, post-gadolinium T1WI with fat suppression also helpful
  - STIR or T2WI MR with fat saturation
    - To distinguish hematopoietic elements from marrow edema or neoplastic infiltration

DIFFERENTIAL DIAGNOSIS

Normal Hematopoietic Marrow

- Children up to ~ 10 years

Marrow Infiltration/Replacement

- Leukemia/lymphoma
- Multiple myeloma/metastases
  - Diffuse or focal T1 hypointensity
    - Multifocal disease more easily distinguished from hyperplastic normal marrow
    - Diffuse marrow infiltration may appear very similar
  - SI usually greater than muscle on STIR or fat-suppressed T2WI
  - Post-gadolinium enhancement
  - Increased metabolic activity on PET scan
  - Pathologic compression fractures
  - Epidural/paravertebral tumor extension

Myeloproliferative Disorder

- Polycythemia vera, chronic myelogenous leukemia, primary thrombocythemia, myelofibrosis with myeloid metaplasia
- MR may look like hyperplastic normal marrow

Marrow Edema

- Multiple factors, such as trauma or infection

- Hypointensity on T1WI
  - Typically less than muscle SI
- Focal or multisegmental hyperintensity on STIR
- Soft tissue and intraspinal hematoma in trauma

PATHOLOGY

General Features

- Etiology
  - Iron deficiency anemia
    - Most common cause of anemia
    - Chronic blood loss
    - Increased requirements (pregnancy, adolescence, etc.)
    - Inadequate diet
  - Anemia of chronic disease
    - 2nd most common cause of anemia
    - Infections (e.g., AIDS), inflammatory disease (e.g., rheumatoid arthritis), cancer (most common)
    - Interferons, interleukins from monocytes inhibit erythropoietin production, erythroid proliferation
    - Inadequate release of iron from reticulum cells to plasma
    - Increased destruction of red blood cells by reticuloendothelial system
  - Hemolytic anemia
    - Accelerated destruction of red blood cells
    - Sickle cell disease, thalassemia, etc.
      - Osteonecrosis in sickle cell anemia
  - Chronic hypoxemia
    - Cyanotic heart disorder, heart failure
    - Heavy smoking: > 1 pack per day
    - Others: High altitude, obesity, marathon athletes
  - Hematopoietic growth factors
    - Such as GCSF
      - Promotes growth and differentiation of hematopoietic stem cells
    - Antiviral HIV meds
- Embryology-anatomy
  - At birth, entire skeletal marrow space occupied with red marrow
  - Conversion of red to yellow marrow occurs in 1st 2 decades of life
    - 1st year: Conversion to fatty marrow begins
      - Starts centrally within vertebral body T1 signal compared to disc
      - < 1 year hypointense; 1-5 years isointense
      - > 5 years hyperintense marrow T1 signal compared to disc
    - Adult marrow pattern by 25 years of age
  - From peripheral and distal to central and proximal
  - With age, red marrow decreases from 58% of total marrow volume in 1st decade to 29% by 8th decade
  - Also to replace bone loss
- Process of reconversion starts in axial skeleton
  - Beginning with calvarium, then vertebrae, ribs, sternum, and pelvis
  - Followed by extremities, from proximal to distal
  - Reverse comparison to maturation
  - Epiphyses usually spared unless extreme needs
  - Extensive reconversion in appendicular skeleton suggests severe anemia or neoplastic involvement of axial skeleton
- Adults < 40 years have better marrow reconversion capacity than older adults

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May not have symptoms referable to spine
  - Other signs/symptoms related to anemia, hypoxemia
    - Skin and mucous membrane pallor
    - Weakness and fatigue
    - Dyspnea
    - Congestive heart failure

Demographics

- Ethnicity
  - No racial predilection, except certain anemias
    - Sickle cell disease: African Americans
    - Thalassemia: Races along eastern Mediterranean Sea (e.g., Greek, Italian, Persian)

Treatment

- Treatment of underlying disease
- Transfusions
- Recombinant erythropoietin

DIAGNOSTIC CHECKLIST

Consider

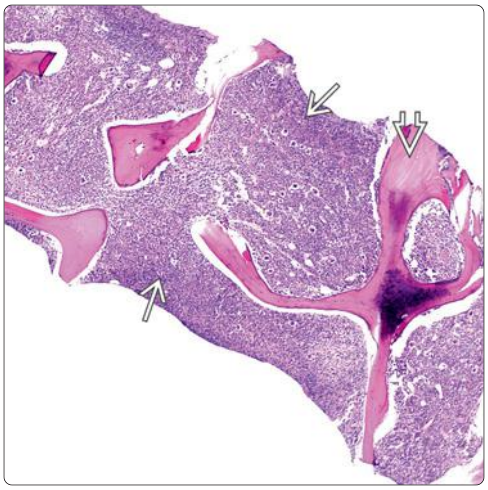
- Fat-suppressed T2WI (or STIR) and T1WI with gadolinium when hypointense marrow present on T1WI

Image Interpretation Pearls

- Diffuse marrow T1 hypointensity without marked post-gadolinium enhancement or cortical disruption suggests hyperplastic marrow
- Normal-appearing marrow on T1 may not exclude infiltration by plasmocytes or lymphocytes when water-fat balance is not sufficiently altered

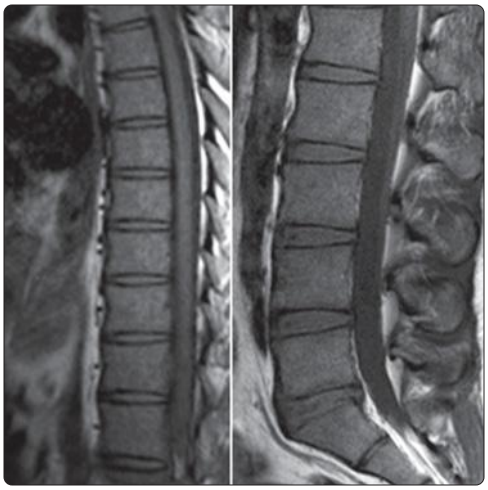
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(Left) *Micrograph of bone marrow in a patient with HIV infection shows markedly hypercellular marrow ➡ in between the bony trabeculae ➡. This type of marrow hypercellularity is due to trilineage hyperplasia.* **(Right)** *Sagittal MR images illustrate marrow SI isointense to discs on T1WI ➡. There is no abnormal hyperintensity on STIR ➡. Marrow hyperplasia demonstrates signal similar to that of red marrow.*





(Left) *Sagittal T1WI MR of the thoracic and lumbar spine shows marrow isointensity in this patient with HIV infection. Studies report ↓ marrow fat in the setting of ↓ bone density and have shown that marrow may be affected by specific protease inhibitors and lipodystrophy status.* **(Right)** *Coronal T1WI MR of the lumbar spine in a patient with anemia of chronic disease exhibits heterogeneously hypointense marrow, similar to muscle. Note multiple nodules of decreased SI ➡ due to focal red marrow hypertrophy.*





(Left) *Sagittal T1 MR reveals diffuse darker vertebral marrow hypointensity ➡ compared to intervertebral discs in this sickle cell patient with hyperplastic vertebral marrow. The dark vertebral marrow represents hematopoietic elements. Classic biconcave, or "fish mouth" ➡, and "H" vertebrae ➡ are present.* **(Right)** *Sagittal T1WI MR depicts diffusely hypointense marrow ➡ relative to discs. This patient with diffuse metastatic breast cancer had also received marrow-stimulating agents.*

Myelofibrosis

KEY FACTS

TERMINOLOGY

- Myelodysplastic syndrome characterized by marrow replacement with fibrous tissue

IMAGING

- Bone sclerosis
- Splenomegaly, hepatomegaly, paraspinous mass
- Marrow is low signal intensity on all sequences
  - Slightly higher on T2WI, STIR than on T1WI
  - Abnormalities may be diffuse or patchy
  - Signal intensity does not change between in- and out-phase imaging
  - Minimal or no enhancement with gadolinium
- Splenomegaly, hepatomegaly

TOP DIFFERENTIAL DIAGNOSES

- Leukemia and lymphoma
- Erythropoietic marrow
- Sclerotic metastases
- Osteopetrosis
- Regenerating marrow

PATHOLOGY

- Primary: Neoplasm of stem cells stimulates nonneoplastic proliferation of fibroblasts and collagen
- Secondary: Associated with leukemia, lymphoma, metastatic disease, toxins such as benzene

CLINICAL ISSUES

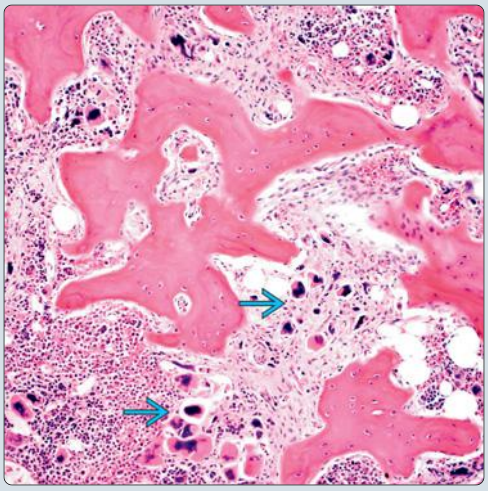
- Usually indolent process, may be asymptomatic
  - Anemia, thrombocytosis, or pancytopenia
  - Extramedullary hematopoiesis → splenomegaly, hepatomegaly
  - Acute myelofibrosis (rare): Abrupt onset, anemia, panmyeloid proliferation, rapidly fatal
- Ruxolitinib is an oral *JAK1* and *JAK2* inhibitor approved by FDA for treatment of intermediate or high-risk myelofibrosis

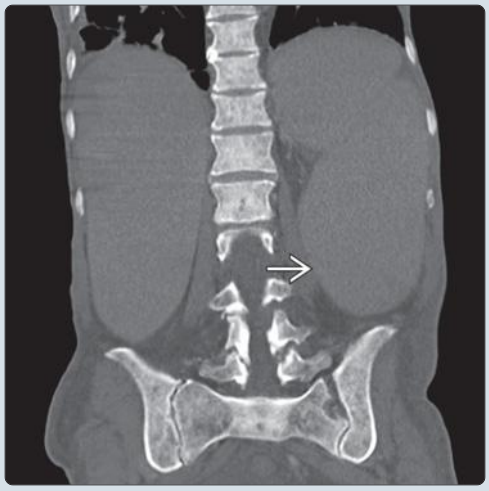
(Left) *Sagittal T1WI MR in a 70-year-old woman with a long history of thrombocytosis and subsequent development of pancytopenia shows extensive, mildly heterogeneous low signal intensity in bone marrow. Biopsy confirmed myelofibrosis.* **(Right)** *Sagittal T2WI MR in the same patient shows that bone marrow is low signal intensity. The patient was refractory to treatment, but she survived 8 years from diagnosis of myelofibrosis.*





(Left) *Marked osteosclerosis, myelofibrosis, and megakaryocytic pleomorphism are the hallmark histologic features of primary myelofibrosis. Megakaryocytes range from small to large and are hyperchromatic and clustered.* **(Right)** *Coronal bone CT in the same patient shows bone sclerosis and splenomegaly. Extramedullary hematopoiesis most commonly involves the spleen, but it may also be seen in paraspinous soft tissues.*





TERMINOLOGY

Synonyms

- Myelosclerosis
- Agnogenic myeloid metaplasia

Definitions

- Myelodysplastic syndrome characterized by marrow replacement with fibrous tissue

IMAGING

Radiographic Findings

- Radiography
  - Usually normal; bone sometimes diffusely sclerotic
  - Extramedullary hematopoiesis
    - Splenomegaly, hepatomegaly
    - Paraspinous mass

CT Findings

- Bone sclerosis
- Extramedullary hematopoiesis

MR Findings

- Marrow is low signal intensity on all sequences
  - Slightly higher on T2WI, STIR than on T1WI
  - Abnormalities may be diffuse or patchy
  - Signal intensity does not change between in- and out-phase imaging
  - Minimal or no enhancement with gadolinium

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Sagittal T1WI, T2WI, STIR

DIFFERENTIAL DIAGNOSIS

Leukemia and Lymphoma

- Low signal intensity on T1WI
- Intermediate signal intensity on T2WI, STIR

Erythropoietic Marrow

- Low signal intensity on T1WI but higher than disc or muscle
- Admixed with fat
- Intermediate signal intensity on T2WI, STIR

Sclerotic Metastases

- Very low signal intensity on T1WI and T2WI
- Usually patchy, nonuniform

Osteopetrosis

- Very low signal intensity on T1WI and T2WI
- "Bone-in-bone" appearance on radiographs

Regenerating Marrow

- After bone marrow transplant or marrow stimulating agent
- Usually admixed with fat, see ↓ signal on out-phase imaging

PATHOLOGY

General Features

- Etiology
  - Primary: Neoplasm of stem cells stimulates nonneoplastic proliferation of fibroblasts and collagen
    - → loss of hematopoietic elements
    - May be precursor to chronic myeloid leukemia
  - Related, in part, to mutations creating overactive signaling of Janus kinase (JAK)/signal transducer and activator of transcription (STAT) pathway
  - Secondary: Associated with leukemia, lymphoma (Hodgkin and non-Hodgkin), metastatic disease, toxins such as benzene

Microscopic Features

- Increased reticulin fibers in bone marrow; other findings variable
- Mutations in *JAK2* and *MPL* genes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually indolent process, may be asymptomatic
  - Anemia, thrombocytosis, or pancytopenia
  - Extramedullary hematopoiesis → splenomegaly, hepatomegaly
  - Acute myelofibrosis (rare): Abrupt onset, anemia, panmyeloid proliferation, rapidly fatal

Demographics

- Age
  - Primary: 50-70 years

Natural History & Prognosis

- Variable, depending on causative disease entity

Treatment

- Options, risks, complications
  - Androgens, prednisone, thalidomide, bone marrow transplants have some effectiveness
  - Ruxolitinib is oral *JAK1* and *JAK2* inhibitor approved by FDA for treatment of intermediate or high-risk myelofibrosis

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Cannot exclude concomitant tumor by MR; biopsy needed for diagnosis

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Bone Infarction

KEY FACTS

TERMINOLOGY

- Infarction of vertebral body cancellous bone and marrow secondary to systemic disease or aortic pathology
  - **Not** osteonecrosis (Kümmell disease)

IMAGING

- Well-defined, geographic signal abnormality, which tends to involve anterior 1/3 to 1/2 of vertebral body
- T2WI shows increased signal in multiple bodies, with abrupt transition to normal marrow signal
- T1WI C+ shows markedly diminished enhancement of affected vertebral body areas
  - No associated epidural or paravertebral soft tissue
  - Rare to involve posterior elements

TOP DIFFERENTIAL DIAGNOSES

- Infarction secondary to underlying systemic disease
  - Sickle cell
  - Acute leukemia (ALL or AML)
  - SLE
  - Lymphoma
  - Transplantation with graft vs. host disease
- Infarction secondary to aortic disease
  - Dissection
  - Abdominal aortic surgery
- Metastatic disease marrow infiltration
- Leukemia or lymphoma marrow tumor infiltration
- Granulomatous or fungal infection

DIAGNOSTIC CHECKLIST

- Infarction as sign of systemic illness or malignancy, such as leukemia
- Associated with spinal cord infarction; useful as a confirmatory sign that nonspecific T2 hyperintensity in cord reflects infarction

(Left) *Sagittal T1WI C+ MR in a patient with a new diagnosis of lymphocytic leukemia shows multiple well-defined, geographic foci of diminished enhancement ➡ in multiple vertebral bodies, with a rim of increased enhancement. There is no associated soft tissue mass and no disc involvement.* **(Right)** *Axial T1WI C+ MR in a patient with multiple vertebral infarcts and a new diagnosis of ALL shows sharply marginated lesions in both right and left sides of the vertebral body ➡ with mild peripheral enhancement.*





(Left) *Sagittal T1WI MR shows multiple infarcts in a patient with ALL status post chemotherapy. Note well-defined low signal present in the anterior 1/2 of multiple vertebral bodies. (Courtesy M. Pathria, MD.)* **(Right)** *Sagittal T2WI FS MR in a patient with ALL status post chemotherapy shows focal, well-defined hyperintensity in the anterior 1/2 of multiple vertebral bodies, which also involves the sacrum. (Courtesy M. Pathria, MD.)*





TERMINOLOGY

Synonyms

- Immature bone infarct, bone marrow infarction

Definitions

- Infarction of vertebral body cancellous bone and marrow secondary to systemic disease or aortic pathology
- **Not** osteonecrosis (Kümmell disease)

IMAGING

General Features

- Best diagnostic clue
  - Geographic T2 hyperintensity of multiple vertebral bodies with diminished enhancement in setting of systemic disease
- Location
  - Multiple vertebral bodies
- Size
  - Variable from entire vertebral body to small percentage
- Morphology
  - Well-defined, geographic signal abnormality, which tends to involve anterior 1/3 to 1/2 of vertebral body

Radiographic Findings

- Radiography
  - Usually negative for immature infarction, or subtle loss of density

CT Findings

- NECT
  - Acute and subacute infarction shows no abnormality, or mild decreased density
  - Chronic phase may involve sclerosis, or typical H pattern of infarction seen in sickle cell disease

MR Findings

- T1WI
  - Low signal in multiple bodies with well-defined margins; vertical orientation
- T2WI
  - Increased signal in multiple bodies, with abrupt transition to normal marrow signal
- T1WI C+
  - Central nonenhancing geographic regions of vertebral body, which may show peripheral enhancement
  - No associated epidural or paravertebral soft tissue
  - Rare to involve posterior elements

DIFFERENTIAL DIAGNOSIS

Secondary to Underlying Systemic Disease

- Sickle cell
- Acute leukemia (ALL or AML)
- SLE
- Lymphoma
- Transplantation with graft vs. host disease

Secondary to Aortic Disease

- Dissection
- Abdominal aortic surgery

Metastatic Disease

- Vertebral body and posterior elements involved
- Focal masses with epidural or paravertebral soft tissue extension

Leukemia or Lymphoma Marrow Tumor Infiltration

- More homogeneous marrow involvement without well-defined margins
- Will enhance homogeneously

Granulomatous or Fungal Infection

- Usually has some element of soft tissue extension
- Commonly involves disc space

PATHOLOGY

Gross Pathologic & Surgical Features

- Vascular stasis, thrombosis, platelet dysfunction
- GVHD-related microangiopathy major cause for bone marrow infarction in transplant population

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific back pain in setting of systemic illness
  - Myelopathy, paraparesis if associated with spinal cord infarction from aortic disease

DIAGNOSTIC CHECKLIST

Consider

- Infarction as sign of systemic illness or malignancy, such as leukemia
- Associated with spinal cord infarction; useful as a confirmatory sign that nonspecific T2 hyperintensity in cord reflects infarction

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KEY FACTS

TERMINOLOGY

- Epidural ± paravertebral proliferation of hematopoietic tissue in response to profound chronic anemia
- Minimally enhancing isointense thoracic intra- or paraspinal masses with associated diffuse marrow hypointensity

IMAGING

- Midthoracic > cervical, lumbar
- CT
  - Soft tissue density
  - Central canal narrowing
  - No bony erosion or calcifications
- MR
  - T1: Isointense to cord
  - T2: Iso- to mildly hyperintense to cord
  - Minimal, mild, or moderate enhancement

TOP DIFFERENTIAL DIAGNOSES

- Epidural/paraspinal metastasis
- Spinal epidural lymphoma
- Paravertebral phlegmon/abscess
- Peripheral nerve sheath tumor
- Epidural hematoma

PATHOLOGY

- Ectopic hematopoietic rests stimulated in response to chronic anemic states
  - Intermediate β-thalassemia: Most common
  - Sickle cell anemia
  - Polycythemia vera
  - Myelofibrosis with myeloid metaplasia

CLINICAL ISSUES

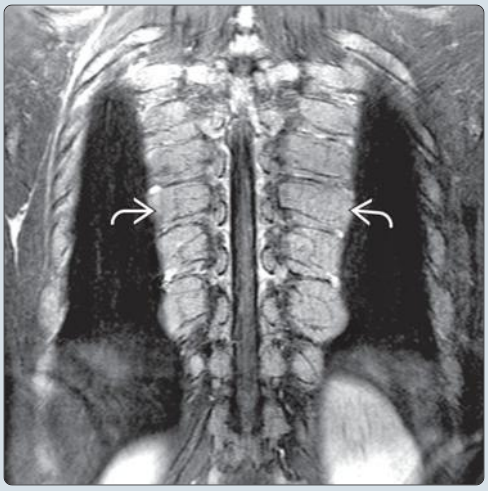
- Asymptomatic
- Back ± radicular pain

(Left) *Sagittal graphic of extramedullary hematopoiesis (EMH) depicts abundant hematopoietic marrow in lumbar vertebrae, extending into prevertebral ➡ and dorsal epidural space ⇨. EMH is a common compensatory phenomenon to chronic hemolytic anemias.* **(Right)** *Sagittal T1WI MR shows an ovoid, well-circumscribed, isointense epidural soft tissue mass ➡ compressing the distal thoracic cord. This is a typical MR case of extramedullary hematopoiesis presenting as epidural soft tissue mass.*





(Left) *Coronal T1WI C+ FS MR of the spine and paraspinal tissues demonstrates deposits of extramedullary hematopoiesis extending from the ribs ➡. Due to insufficient production of blood elements by the bone marrow, hematopoietic tissues at other sites proliferate to compensate for the circulatory demands for blood elements.* **(Right)** *Axial T1WI MR reveals foci of extramedullary hematopoiesis breaking through the vertebral bodies into the neural foramina ➡ from the ribs ➡.*





TERMINOLOGY

Abbreviations

- Extramedullary hematopoiesis (EMH)

Definitions

- Epidural ± paravertebral proliferation of hematopoietic tissue in response to profound chronic anemia

IMAGING

General Features

- Best diagnostic clue
  - Minimally enhancing isointense thoracic intra- or paraspinal masses with associated diffuse marrow hypointensity
- Location
  - Midthoracic > cervical, lumbar
    - Epidural, paravertebral
- Size
  - multisegmental
- Morphology
  - Well circumscribed
  - Homogeneous and lobular soft tissue mass

Radiographic Findings

- Radiography
  - Bilateral (a)symmetrically widened paraspinal stripe on frontal radiograph
  - ± rib, clavicle medullary expansion

CT Findings

- NECT
  - Soft tissue density
  - Central canal narrowing
  - Cord displacement and compression
  - ± rib, clavicle medullary expansion
- CECT
  - Mild enhancement
- Bone CT
  - No bony erosions or calcifications

MR Findings

- T1WI
  - Isointense to cord
- T2WI
  - Iso- to mildly hyperintense to cord
    - Hypointensity may represent increased iron content in hematopoietic tissue
- STIR
  - Iso- to hyperintense
- T1WI C+
  - Minimal, mild, or moderate
- Variable mass effect on spinal cord
  - Cord compression most commonly reported with β-thalassemia
  - Intramedullary T2 hyperintensity may be present
    - Edema or myelomalacia
- Nerve root compression
- Diffuse vertebral marrow T1 hypointensity on all sequences

Nonvascular Interventions

- Myelography
  - Nonspecific hypodense epidural mass effacing central canal
  - Vertebrae are enlarged and translucent ~ 86% in 1 report

Nuclear Medicine Findings

- Technetium sulfur colloid scan
  - Foci of epidural/paraspinal uptake correspond to EMH

Imaging Recommendations

- Best imaging tool
  - Sagittal, axial T2WI, T1WI, T1 C+ MR

DIFFERENTIAL DIAGNOSIS

Epidural/Paraspinal Metastasis

- Extension from adjacent vertebral lesions
  - Posterior cortex typically involved
    - High intensity on STIR
  - Compression fractures
- Isolated epidural/paraspinal disease sparing spinal column rare
- Moderate enhancement invariably present

Spinal Epidural Lymphoma

- Isointense to cord on T1WI
  - Iso- to hyperintense on T2WI
- Intense homogeneous post-gadolinium enhancement
- ± adjacent vertebral involvement
- ± diffuse marrow hypointensity

Paravertebral Phlegmon/Abscess

- Associated with infectious spondylitis
  - Destructive changes in adjacent vertebrae
- Hyperintense on STIR
  - Especially liquefied components
- Diffuse (phlegmon) or peripheral (abscess) enhancement

Peripheral Nerve Sheath Tumor

- Often at a single level
- Multiple neurofibromas in neurofibromatosis type 1
- Isointense to cord on T1WI, ↑ signal on T2WI
- Avid post-contrast enhancement
- Widened intervertebral foramina

Epidural Hematoma

- Often hyperintense on T1WI
- Iso- to hypointense on T2WI
- ± post-gadolinium enhancement
- Sudden onset of symptoms

PATHOLOGY

General Features

- Etiology
  - Ectopic hematopoietic rests stimulated in response to chronic anemic states
    - Intermediate β-thalassemia: Most common
      - Transfusion independent because of compensatory mechanisms of bone marrow
    - Sickle cell anemia

- Polycythemia vera
- Myelofibrosis with myeloid metaplasia
- Source of extravertebral hematopoiesis controversial
  - Stimulation of embryonic multipleuritant hematopoietic stem cells in epidural space
  - Direct extension of hematopoietic marrow from vertebrae into epidural space
  - Fetal hematopoietic capacity of dura
  - Embolic phenomenon
- Associated abnormalities
  - Marrow expansion in ribs
  - Small infarcted spleen in patients with sickle cell disease
    - Splenomegaly in thalassemia
  - Hemothorax with paraspinal EMH
  - Labs: Chronic microcytosis hemolytic anemia
- Compensatory mechanism when normal marrow insufficient to meet circulatory demands in chronic anemia
- Common sites of EMH: Liver, spleen, lymph nodes
  - Uncommon sites: Retroperitoneal tissue, kidneys, adrenal area, paravertebral region, thymus, breasts, sweat glands, pleura, prostate, broad ligaments
  - Unusual sites: Peripheral nerves and cranial dura matter
  - Sites of fetal hematopoiesis

Gross Pathologic & Surgical Features

- Discrete flesh-colored mass

Microscopic Features

- Resembles bone marrow on biopsy
  - Trilineage hematopoiesis
    - Erythroid and granulocytic precursors, megakaryocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Back ± radicular pain
  - Other signs/symptoms
    - Paraparesis
    - Sensory deficit
    - Gait disturbance
    - Bladder, bowel dysfunction
    - Diminished deep tendon reflexes
    - Anemia, pancytopenia
- Clinical profile
  - Cord compression from epidural EMH should be suspected in patients with chronic anemia who present with neurologic complaints

Demographics

- Age
  - More common in adults
    - 3rd-4th decade
- Ethnicity
  - Some hemoglobinopathies more common in certain ethnic groups
    - Sickle cell disease: African Americans
    - Thalassemia: Races along Eastern Mediterranean Sea (Greek, Italian, Persian, etc.)
  - Myeloproliferative disorders: No ethnic predilection
- Epidemiology
  - Spinal involvement most common after hepatosplenic EMH
    - Spinal EMH: 27% of nonhepatosplenic involvement
    - Intraspinal EMH occurs in 11-15% of patients with thalassemia

Natural History & Prognosis

- Excellent prognosis: Resolution of symptoms 3-7 days after radiotherapy
  - Overall, prognosis limited by underlying hemoglobinopathy or myeloproliferative disorder

Treatment

- Intravenous steroids
  - Decrease cord edema
- Radiation therapy
  - Ectopic hematopoietic tissue extremely radiosensitive
  - Alone or in conjunction with surgery
  - Risk of marrow suppression
    - May be stimulus for further EMH
- Decompressive laminectomy with surgical resection
  - Indicated when significant myelopathy present
- Transfusions
  - When radiation or surgery contraindicated
  - Hematopoietic tissues regress as stress of anemia relieved
  - Frequent recurrences
- Hydroxyurea
  - Increases production of hemoglobin-F
  - Role in myelosuppression
- Fetal hemoglobin-inducing agents that decrease hematopoietic drive

DIAGNOSTIC CHECKLIST

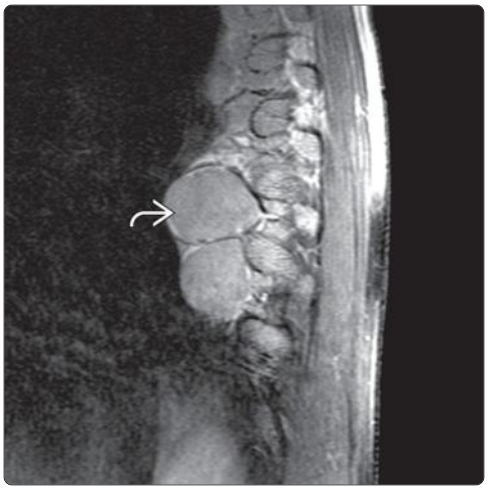
Image Interpretation Pearls

- Spinal EMH should be considered when thoracic epidural/paraspinal isointense masses are present in patients with hemoglobinopathies or myeloproliferative disorders

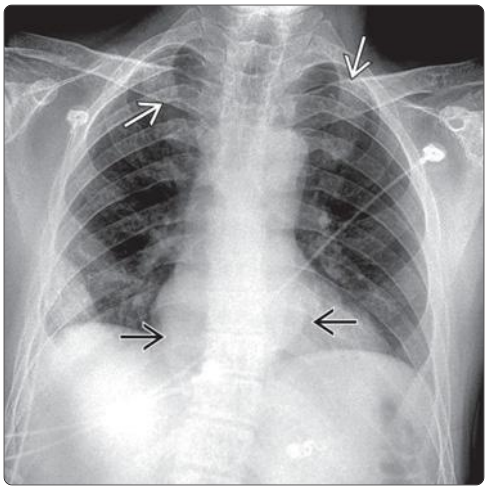
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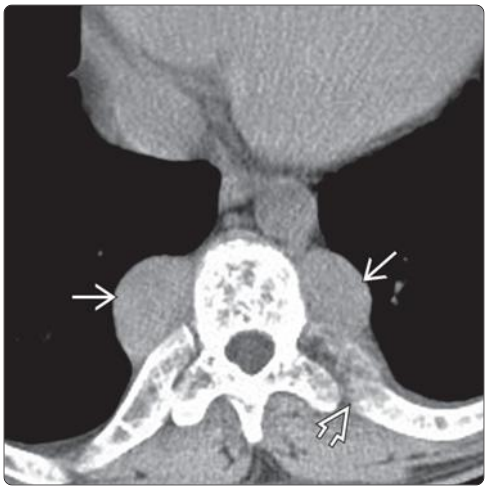
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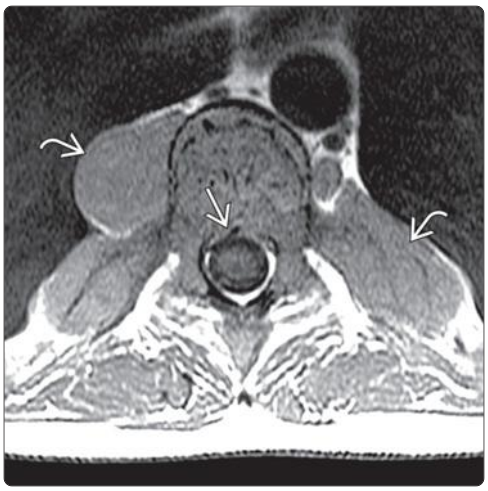


(Left) *Axial T2WI MR exhibits intermediate-intensity foci breaking through vertebral bodies into the epidural space and from the ribs. The pathogenesis includes the extrusion of proliferating bone marrow stem cells through the cortex of the vertebral bodies and ribs into the subperiosteal region, promoted by the negative pressure of the thoracic cavity.* **(Right)** *Sagittal T1WI C+ FS MR illustrates diffuse homogeneous enhancement of the paraspinal deposits of extramedullary hematopoiesis.*





(Left) *Anteroposterior chest radiograph shows medullary cavity expansion in bilateral clavicles and ribs. There is also widening of paraspinal soft tissue stripe.* **(Right)** *Axial NECT through lower thoracic spine confirms the presence of paraspinal soft tissue masses and medullary expansion of vertebra and ribs. In this case of extramedullary hematopoiesis, there is marrow expansion from severe anemia and paraspinal masses as extramedullary hematopoietic elements.*





(Left) *Axial T1WI MR demonstrates abnormal marrow signal and deposits of EMH breaking through the vertebral bodies into the epidural space and from the ribs. In older patients, differential considerations of an epidural mass on MR include lymphoma, myeloma, and metastatic disease.* **(Right)** *Sagittal T1WI MR reveals isointense soft tissue masses in the dorsal epidural space. The marrow SI of the vertebral bodies is diffusely hypointense due to hyperplastic hematopoietic elements.*

Tumoral Calcinosis

KEY FACTS

TERMINOLOGY

- Benign periarticular soft tissue hyperplasia, calcification

IMAGING

- Nonaggressive appearing calcific mass centered about large synovial joints
- Predilection for large joints
  - Hip
  - Shoulder
  - Elbow
  - Spinal involvement uncommon
- Radiographs/CT
  - Calcific mass with clustered calcific aggregates surrounding joint
- T1/T2/STIR
- Lobulated low signal masses centered on facet joint
- May extend into adjacent paraspinal soft tissue
- May extend beyond midline to involve dorsal elements, ligamentum flavum
- Minimal enhancement

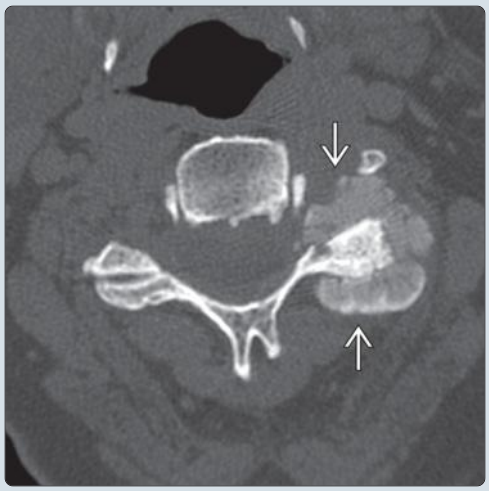
TOP DIFFERENTIAL DIAGNOSES

- Calcium pyrophosphate deposition disease
- Primary bone tumor
- Synovial chondromatosis
- Neuropathic joint
- Synovial cyst

PATHOLOGY

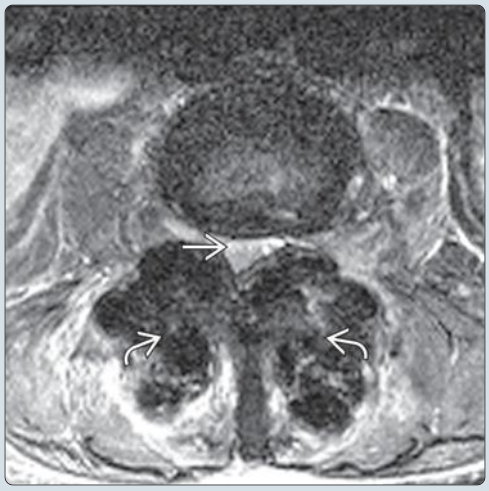
- Complication of chronic renal failure, typically on renal dialysis
- Familial tumoral calcinosis (FTC)
  - Normophosphatemic FTC
    - Likely related to mutations in the gene encoding for SAMD9 protein
  - Hyperphosphatemic FTC
    - Increased renal absorption of phosphate due to loss-of-function mutations





(Left) *Sagittal lumbar CT shows mass-like calcifications involving posterior elements. Calcified masses extend into bony canal causing central canal encroachment. Radiodense periarticular masses are usually calcium hydroxyapatite crystals surrounded by foreign body giant cell and histiocytic reaction.* **(Right)** *Axial NECT through cervical spine shows lobulated density surrounding the facet joint with well-defined margins and no soft tissue component. (Courtesy N. Stence, MD.)*





(Left) *Axial CT reveals central canal encroachment due to copious posterior element calcifications.* **(Right)** *Axial T2WI MR exhibits hypointensity of these posterior element calcified masses. There is moderate mass effect upon the nerve roots within the thecal sac. Tumoral calcinosis (TC) is usually associated with hereditary disorders of calcium metabolism or renal dialysis. It also occurs in degenerated tissues in the absence of systemic disorders.*

TERMINOLOGY

Abbreviations

- Tumoral calcinosis (TC)
- Familial tumoral calcinosis (FTC)

Synonyms

- Extraskeletal tumoral calcification, calcifying pseudoneoplasm

Definitions

- Periarticular soft tissue hyperplasia, calcification

IMAGING

General Features

- Best diagnostic clue
  - Nonaggressive appearing calcific mass centered about large synovial joints
    - Rare and commonly misdiagnosed
- Location
  - Predilection for large joints
    - Hip, shoulder, elbow
    - Spinal involvement uncommon
- Size
  - Variable
- Morphology
  - Well-defined calcific mass

Radiographic Findings

- Radiography
  - Calcific mass with clustered calcific aggregates surrounding joint
  - May show bone remodeling but no destruction
  - May show degenerative changes, including spondylolisthesis or spondylosis deformans

CT Findings

- NECT
  - Calcific mass with clustered calcific aggregates surrounding joint
  - Occasional bone remodeling without bone destruction
  - No significant noncalcified soft tissue mass

MR Findings

- T1WI
  - Lobulated low signal masses centered on facet joint
  - May extend into adjacent paraspinal soft tissue
  - May extend beyond midline to involve dorsal elements, ligamentum flavum
  - Diagnosis can be difficult on MR → CT confirmation
- T2WI
  - Lobulated, low signal masses
  - Mass effect on adjacent thecal sac
- STIR
  - Lobulated, low signal masses
- T2* GRE
  - Low signal from expansile dorsal element lobulated masses
- T1WI C+
  - Minimal enhancement
  - No associated enhancing soft tissue component

Nuclear Medicine Findings

- Bone scan
  - Markedly increased uptake

Imaging Recommendations

- Best imaging tool
  - CT shows calcific lesions, extent, presence or absence of bone destruction
- Protocol advice
  - Axial thin-section CT, multiplanar reformats

DIFFERENTIAL DIAGNOSIS

Calcium Pyrophosphate Deposition Disease

- Calcification involving ligamentum flavum
- May compress dorsal thecal sac
- No facet joint involvement

Primary Bone Tumor

- Ill-defined margins, infiltrative, bone destruction
- Enhancing soft tissue component separate from calcific mass

Synovial Chondromatosis

- Usually affects larger joints as does tumoral calcinosis
- Monoarticular involvement, unknown cause
- Progressive periarticular swelling and pain
- Multiple intraarticular densities on CT, plain film

Neuropathic Joint

- Endplate and facet joint destruction, bone debris
- Preserved bone density

Baastrup Disease

- Degeneration of interspinous ligament, ligamentum flavum
- Thickening, buckling of dorsal spinal ligaments
- Cystic degeneration of interspinous ligament with hyperintensity on T2WI

Infection/Septic Facet

- Abnormal low signal in facet, with irregular adjacent soft tissue extension
- Joint effusion, bone destruction
- Peripheral enhancement of epidural abscess

Synovial Cyst

- Well-defined mass arising from ventral margin of facet joint/ligamentum flavum
- Central high signal on T2WI, peripheral T1 enhancement
- May show peripheral calcification

PATHOLOGY

General Features

- Etiology
  - Complication of chronic renal failure, typically on renal dialysis
  - FTC
    - Normophosphatemic FTC
      - Calcium, phosphate, vitamin D3 metabolites, and parathyroid-hormone levels normal
      - Typically solitary calcifications
      - Present before 2nd decade of life

        - Likely related to mutations in the gene encoding for SAMD9 protein
      - Hyperphosphatemic FTC
        - Increased renal absorption of phosphate due to loss-of-function mutations
- Genetics
  - Regulation of phosphate is specific, involving phosphatonins, especially fibroblast growth factor 23 (*FGF23*)
  - Hyperphosphatemic FTC
    - OMIM #211900
    - Mutation in the *GALNT3* gene (601756), *FGF23* gene (605380), or *KL* gene (604824)
  - Normophosphatemic FTC
    - OMIM #610455
    - Mutation in the gene encoding the sterile alpha motif domain-containing-9 protein (SAMD9; 610456)
- Associated abnormalities
  - Hyperphosphatemic FTC associated with abnormal dentition
    - Short, abnormal roots and obliteration of the root canals and pulp chambers
  - Chronic renal failure
    - TC-like lesion associated with disorder of calcium and phosphate metabolism, a result of secondary hyperparathyroidism due to chronic renal failure
    - Estimated incidence 0.5-7% of hemodialysis patients
    - Secondary hyperparathyroidism
    - ↑ serum parathyroid (PTH) → osteoclast activation → phosphate retention, relative hypocalcemia, altered vitamin D metabolism, and altered feedback regulation of PTH
  - Need to exclude other causes of soft tissue calcification
    - Collagen vascular disease, hypervitaminosis D, chronic renal disease
    - CPPD

Gross Pathologic & Surgical Features

- Calcific mass, with solid and "milk of calcium" components
- Thickened dura, and chalky white paravertebral fluid

Microscopic Features

- Granulomatous reaction of histiocytes, osteoclastic-like multinucleated giant cells, fibroblasts forming thin septa around hydroxyapatite, scant inflammation
- Hydroxyapatite + calcium pyrophosphate dihydrate crystals identified

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Enlargement of joint, which may be initially painless
    - With enlargement, may cause discomfort, joint motion limitation
    - Large soft tissue masses may cause overlying skin ulceration
  - Other signs/symptoms
    - Usually lesions at other large synovial joints when spinal involvement present
    - Leg, back, or neck pain, myalgia
    - May cause neural compression → radiculopathy, paraparesis, or sensory deficits
    - Underlying medical conditions contribute to pathogenesis
- Clinical profile
  - Painless growth about large joint in adult

Demographics

- Age
  - Adult
- Gender
  - M = F
- Ethnicity
  - African American > Caucasian
- Epidemiology
  - 2nd to 3rd decades of life

Natural History & Prognosis

- Progressive
- Multiple locations may be involved

Treatment

- Options, risks, complications
  - Symptomatic treatment
  - Wide surgical excision
  - Recurrence common
  - Evaluate for underlying metabolic abnormality
  - Drug treatment with phosphorus deprivation by oral aluminum hydroxide, or acetazolamide

DIAGNOSTIC CHECKLIST

Consider

- Tumoral calcinosis is diagnosis of exclusion
- Look for lesions in other large joints by CT

Image Interpretation Pearls

- Recognition of this entity important
  - Benign nature
  - Potential for confusion with malignant lesions, such as sarcoma
- No enhancement

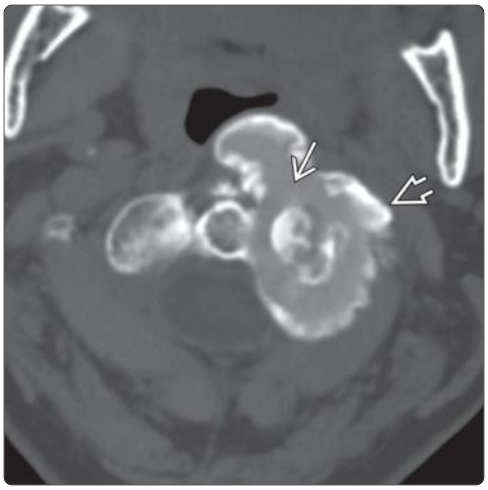
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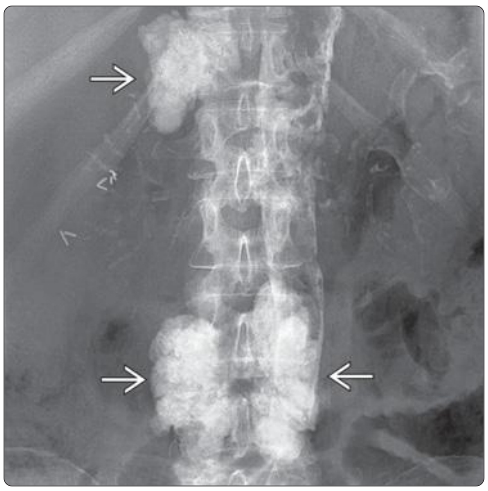


(Left) *Sagittal T2WI MR shows lobulated, hypointense masses arising from the facets, which are typical for benign tumoral calcinosis. The margins are sharp and without discrete soft tissue mass or adjacent edema.* **(Right)** *Axial T1WI C+ FS MR illustrates peripherally enhancing, hypointense masses arising from bilateral facet joints. No discrete soft tissue mass is seen.*





(Left) *Axial NECT shows a well-defined, calcified mass centered on the left C1-C2 articulation with large regions of homogeneous internal increased density, with peripheral higher attenuation.* **(Right)** *Sagittal T2WI MR shows low signal from the mass, which showed calcification on CT. A 2nd small focus of tumoral calcinosis is present at C7-T1. The MR findings are nonspecific, and CT is needed to make the diagnosis.*



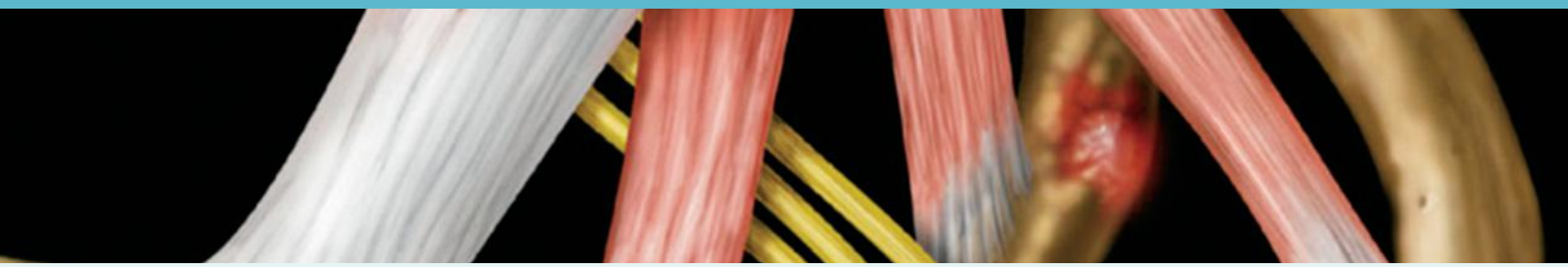


(Left) *Anteroposterior radiograph demonstrates abundant mass-like calcifications involving posterior elements of T12, L1, L3, and L4 vertebrae. TC may not be visible on plain radiographs. Note extensive vascular calcifications in this patient with renal disease.* **(Right)** *Axial bone CT exhibits the diffuse high attenuation of the lobulated masses. This is consistent with calcification or "milk of calcium." These should not be confused with more aggressive bone-producing neoplasm.*

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PART VI
SECTION 1

Plexus and Peripheral Nerve Lesions



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Tibial Neuropathy	**1030**

Terminology

Nerve rootlets: Individual neural filaments of dorsal and ventral roots directly exiting from spinal cord

Nerve roots: Composed of multiple individual dorsal or ventral nerve rootlets

- Dorsal sensory roots exit from dorsolateral cord and have cell bodies within dorsal root ganglion (DRG)
- Ventral motor roots arise from anterior cord gray matter and have cell bodies within the cord

Dorsal root ganglion (DRG): Dorsal nerve root sensory ganglion, resides in neural foramen

Spinal nerve (proper): Union of dorsal and ventral nerve roots

- 31 nerve pairs (8 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 1 coccygeal)
- Proper spinal nerve is short in length, bifurcates into ventral and dorsal rami

Ramus: 1st spinal nerve branch

- Larger ventral primary ramus (VPR) supplies ventral musculature and facets
- Smaller dorsal primary ramus (DPR) supplies paraspinal muscles and part of facet

Peripheral nerve: Combination of 1 or more rami into single neural conduit

Plexus: Neural network of anastomosing nerves

Imaging Anatomy

Cervical Plexus

The cervical plexus forms from the ventral rami of C1-C4 and variably a C5 minor branch. It has ascending superficial, descending superficial, and deep branches that supply nuchal muscles, diaphragm, and cutaneous head/neck tissues.

Brachial Plexus

The brachial plexus (BP) forms from the ventral rami of C5-T1 and variably from minor branches of C4 or T2. The brachial plexus contributes to several nerves originating above the BP proper, including dorsal scapular nerve, long thoracic nerve, nerves to scalene/longus colli muscles, and a branch to the phrenic nerve. The remaining minor and all major terminal nerve branches arise from the brachial plexus proper.

Anatomically, the BP is classically divided sequentially (proximal to distal) into five segments: Roots/rami, trunks, divisions, cords, and terminal branches. The roots/rami originate directly from the spinal cord levels C5 to T1. The three trunks include the superior or upper (C5-C6), middle (C7), and inferior or lower (C8, T1) trunks. Important minor nerves arising directly from the trunks include the suprascapular nerve and nerve to subclavius muscle. Two divisions are described: Anterior division innervates anterior (flexor) muscles, and posterior division innervates posterior (extensor) muscles. No named minor nerves arise directly from the anterior or posterior divisions. The three cords include the lateral cord (anterior divisions of superior, middle trunks) that innervates anterior (flexor) muscles, the medial cord (anterior division of inferior trunk) that innervates anterior (flexor) muscles, and the posterior cord (posterior divisions of all three trunks) that innervates posterior (extensor) muscles. The cords branch to form several important named terminal peripheral nerve branches.

Clinically, the BP is divided into three discrete segments based on anatomic relationship to the clavicle. These include the supraclavicular (roots, trunks), retroclavicular (divisions), and infraclavicular (cords, terminal branches) plexus.

Lumbar Plexus

The lumbar plexus (LP) derives from the L1-L4 ventral rami and a minor branch from T12. Important named minor nerve branches include the iliohypogastric, ilioinguinal, genitofemoral, lateral femoral cutaneous (L2-L3), and superior (L4-S1) and inferior (L5-S2) gluteal nerves. Important major nerve branches include the femoral (posterior division L2-L4) and obturator (anterior division L2-L4) nerves (table 2).

Lumbosacral Trunk

The lumbosacral trunk (LST) is derived from the ventral rami of L4 (minor branch) and L5, and it is easily followed on axial imaging as it transits along the ventral surface of the sacral ala to join the sacral plexus in the pelvis.

Sacral Plexus

The sacral plexus (SP) is composed of the LST, S1-S3 ventral rami, and a minor branch from S4. The sacral rami and LST converge into the upper sacral neural band (LST and S1-S3) that forms the sciatic nerve and the lower sacral neural band (S3-S4) that forms the pudendal nerve.

Coccygeal Plexus

The coccygeal plexus (CP) converges from the S5 ventral ramus, Cx1, and a minor branch of S4. The major named branch is the anococcygeal nerve.

Anatomy-Based Imaging Issues

Normal Nerve Findings

Surrounding perineural fat provides excellent visualization of nerves and allows them to be easily distinguished from adjacent soft tissues. The normal peripheral nerve is a round/ovoid shape with well-defined internal fascicular architecture. Normal nerve fascicles are uniform size and shape, and this characteristic fascicular pattern helps distinguish peripheral nerves from other lesions, such as schwannoma or ganglion cyst, which also demonstrate high intrinsic T2 signal intensity.

Intrafascicular signal intensity is determined predominately by endoneurial fluid and axoplasmic water, whereas the interfascicular signal is dominated by fibrofatty connective tissue that is amenable to fat suppression. Therefore, normal nerve fascicles are isointense to adjacent muscle tissue on T1WI and mildly hyperintense relative to muscle interspersed within hypointense fibrofatty connective tissue on fat-saturated T2WI or STIR MR. No abrupt change in nerve caliber or course should be observed in an anatomically normal nerve.

Abnormal Nerve Findings

The acutely abnormal nerve demonstrates one or more of the following findings: Segmental nerve enlargement, disruption of nerve anatomic continuity, T2 signal intensity approaching that of regional blood vessels on fat-saturated T2WI or STIR sequences, or disruption or distortion of normal fascicular architecture. Entrapped or scarred nerves may show abrupt change in caliber or course.

The abnormal nerve, therefore, remains isointense on T1WI, but becomes increasingly hyperintense to muscle on T2WI. In the setting of injury, it is postulated that increased endoneurial free water content alters the normal signal characteristics of peripheral nerves. The cause of abnormal high signal on T2WI and STIR sequences is not definitively known, but it has been speculated that edema from increased

endoneurial fluid due to disordered endoneurial fluid flow or local venous obstruction may explain abnormal T2 hyperintensity. Alterations in axoplasmic flow may also produce increased signal. Axoplasmic flow is impeded by nerve compression, and increased axoplasm proximal and distal to the injury site may produce T2 hyperintensity.

Imaging Pitfalls
It can sometimes be difficult to distinguish peripheral nerve from adjacent vascular structures, particularly if the abnormal nerve displays high T2 signal intensity. Vessels demonstrate internal flow voids, branch at large angles, and show intense contrast enhancement. On the other hand, nerves do not show flow voids, branch at relatively acute angles, enhance minimally, and display a discrete distinctive fascicular architecture on transverse imaging.

Clinical Implications

High-resolution MR technique readily identifies the large major nerves and major plexi and permits visualization of their internal neural anatomy. Conversely, smaller major and essentially all minor peripheral nerves are too small to directly visualize.

Successful peripheral nerve imaging requires a strong working knowledge of normal plexus and nerve anatomy. Additionally, peripheral nerve imaging is time consuming and necessitates constraining the imaging volume to clinically relevant regions rather than general "screening" surveys. It is critical to have all pertinent clinical and electrodiagnostic data available to appropriately constrain imaging volume and help detect subtle abnormalities.

Differential Diagnosis

Normal Nerve/Plexus
The normal nerve/plexus shows normal course, caliber, contour, and internal fascicular architecture. Consider myopathic or other nonneural etiology in symptomatic patients.

Nerve/Plexus Mass
Neural neoplasms are most commonly of neural sheath origin. Consider solitary or plexiform neurofibroma, schwannoma, or malignant peripheral nerve sheath tumor (MPNST). Less common considerations include neurolymphomatosis and peripheral nerve metastasis.

Trauma
Etiologies include traction (stretch or avulsion), laceration (projectile, fracture fragment, sharp object), or direct compression (hematoma, fracture).

MR imaging is sensitive for the detection and in some cases discrimination of all three described peripheral nerve injury levels. **Neuropraxia**, the least severe type of injury, is characterized by focal damage to the myelin sheath without axonal disruption and manifests as identifiable, but swollen and hyperintense nerve fascicles. **Axonotmesis** is an intermediate level of crush or traction injury that produces axonal disruption and subsequent wallerian degeneration, but leaves the Schwann cells and endoneurium intact. Axonotmesis will display a homogeneously increased signal intensity nerve with loss of fascicular architecture at the injury site. **Neurotmesis**, the most severe form of nerve injury, cannot always be distinguished from axonotmesis in the cases of functional rather than anatomical transaction, but may display axonal disruption with discontinuity of some or all of the surrounding connective tissues and subsequent wallerian degeneration in definitive cases.

Entrapment Syndrome
Neural compression occurs at characteristic locations. These injuries are often, but not always, related to poor ergonomics or overuse injuries.

Hereditary Motor and Sensory Neuropathy (HMSN)
Inherited peripheral nerve disorders are characterized by abnormally enlarged peripheral nerves (usually palpable if not deep) and variable clinical neuropathy presentations. Some demonstrate characteristic "onion bulb" appearance on micropathology following nerve biopsy, reflecting recurrent episodes of demyelination and remyelination. The most common HMSN disorder is Charcot-Marie-Tooth, which has characteristic clinical manifestations and may involve the cauda equina, peripheral nerves, or both.

Infection/Inflammation
Myriad pathological etiologies and clinical manifestations characterize this diverse group of disorders. Important causes include syphilis (tabes dorsalis), leprosy, infectious neuritis (usually viral), and sarcoidosis. Immune-mediated noninfectious disorders include post viral or vaccination (Guillain-Barré syndrome), chronic immune demyelinating polyneuropathy (CIDP), and idiopathic brachial plexitis (Parsonage-Turner syndrome).

Drug/Toxic Injury
Neural injuries have been linked to vinca alkaloids, therapeutic gold, amiodarone, dapsone, thalidomide, and lead or mercury intoxication.

Vascular Insult
Injury may result from either nerve ischemia related to peripheral vascular disease or vascular trauma or sequelae of vasculitis. The most common vasculitis etiologies are diabetic, Churg-Strauss, polyarteritis nodosa, and Wegener granulomatosis.

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Brachial Plexus Major Nerves

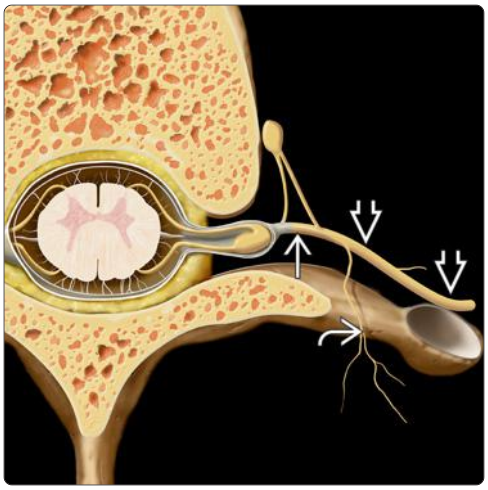
Nerve	Definition	Motor/Sensory Innervation	Important Branches
Radial nerve (RN)	Terminal branch of posterior brachial plexus cord	Innervates extensor muscles of arm and forearm (triceps, brachioradialis, extensor forearm muscles)	Most important branch is posterior interosseous nerve (PIN)
Median nerve (MN)	Terminal branch arises from both lateral and medial brachial plexus cords	Innervates flexor muscles of forearm and thumb as well as 1st and 2nd lumbricals	Most important branch is anterior interosseous nerve (AIN)
Ulnar nerve (UN)	Terminal branch of medial brachial plexus cord	Innervates flexor carpi ulnaris, 3rd and 4th lumbricals, and majority of intrinsic hand muscles	
Musculocutaneous nerve (MuscN)	Terminal branch of lateral brachial plexus cord	Innervates flexor muscles of arm (coracobrachialis, biceps, and brachialis)	
Axillary nerve (AxN)	Terminal branch of posterior brachial plexus cord	Innervates deltoid and teres minor muscles	

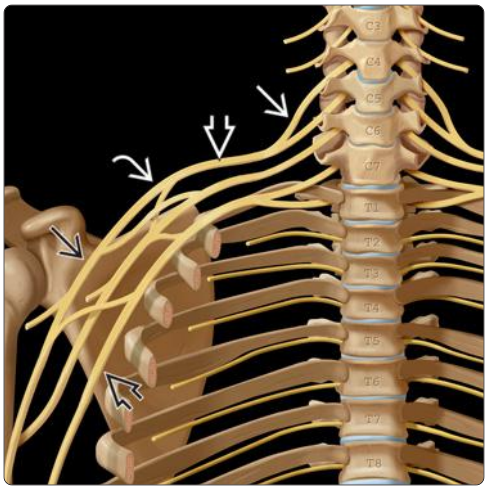
Lumbosacral Plexus Major Nerves

Nerve	Definition	Motor/Sensory Innervation
Obturator nerve (ObN)	Terminal branch of lumbar plexus (anterior division)	Innervates thigh adductor muscles
Femoral nerve (FN)	Terminal branch of lumbar plexus (posterior division)	Innervates iliacus, psoas, and quadriceps muscles
Sciatic nerve (SN)	Largest peripheral nerve branch of sacral plexus	Innervates posterior thigh (biceps femoris, semitendinosus, semimembranosus, adductor magnus) and all leg muscles (via tibial and common peroneal nerve)
Common peroneal nerve (CPN)	Major anterior terminal branch of sciatic nerve	Innervates anterior leg muscles; superficial peroneal nerve (SPN) innervates peroneus muscles, extensor digitorum brevis; deep peroneal nerve (DPN) innervates tibialis anterior, extensor digitorum longus, extensor hallucis longus muscles
Tibial nerve (TN)	Major posterior branch of sciatic nerve	Innervates posterior leg muscles (gastrocnemius, soleus, tibialis posterior, flexor digitorum longus, flexor hallicus longus)

High-Resolution MR Imaging Protocols

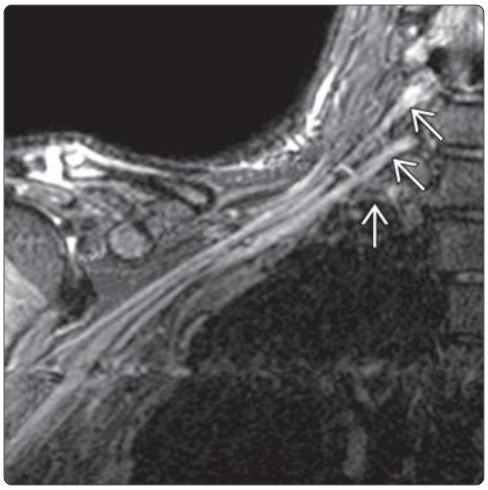
MR Pulse Sequence	Technical Parameters	Technical Comments
Coronal T1WI MR	3-4 mm slice thickness, 20-24 cm FOV, no interslice gap	Direct coronal plane, not oblique
Coronal fat-saturated T2WI or STIR MR	3-4 mm slice thickness, 20-24 cm FOV, no interslice gap	Direct coronal plane, not oblique
Direct axial or sagittal oblique T1WI MR	5-7 mm slice thickness, 16-20 cm FOV, no interslice gap	Sagittal oblique plane oriented perpendicular to plexus
Direct axial or sagittal oblique fat-saturated T2WI or STIR MR	5-7 mm slice thickness, 16-20 cm FOV, no interslice gap	Sagittal oblique plane oriented perpendicular to plexus
Coronal and axial fat-saturated T1WI C+ MR (optional)	Same planes as unenhanced imaging sequences	Use if suspected or confirmed mass or infection



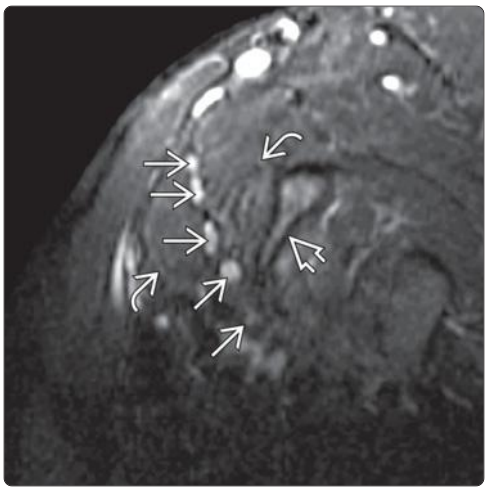


(Left) *Axial graphic of the thoracic spine shows formation of a typical spinal nerve from dorsal and ventral rootlets to form the spinal nerve proper. The short spinal nerve bifurcates into large ventral and small dorsal primary rami.* **(Right)** *Coronal graphic shows the classic anatomic classification of the 5 brachial plexus sections from proximal to distal: Roots (technically ventral primary rami) of C5-T1, trunks, divisions, cords, and terminal major branches.*





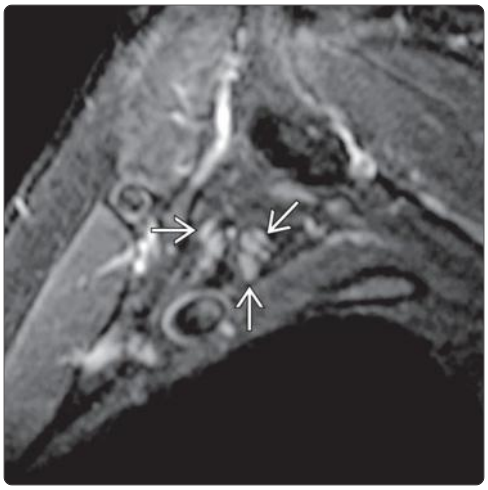
(Left) *Coronal STIR MR of the upper right brachial plexus depicts normal mildly hyperintense brachial plexus roots/rami of C5 to T1. C5 and C6 form the superior (upper) trunk, C7 the middle trunk, and C8 and T1 the inferior (lower) trunk.* **(Right)** *Coronal STIR MR of the lower right brachial plexus demonstrates normal C7, C8, and T1 roots/rami sequentially forming trunks, divisions, and cords. The normal brachial plexus courses retroclavicular into the axilla.*

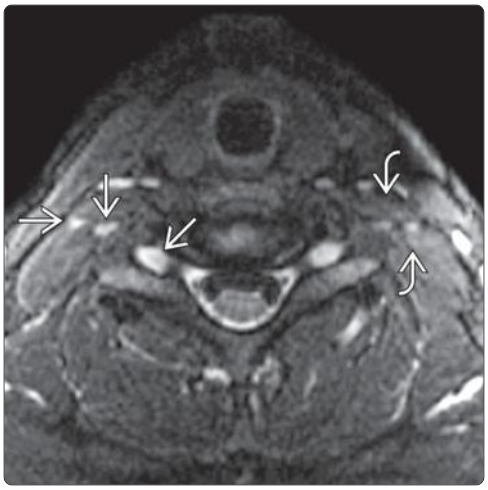




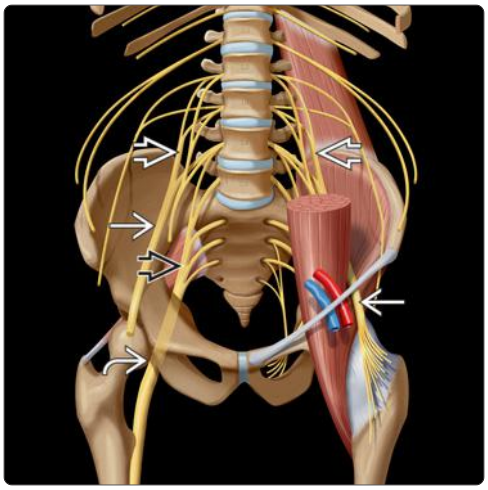
(Left) *Sagittal oblique STIR MR demonstrates the ventral primary rami of C5 through T1 proximal to the trunks. C8 exits above the 1st rib, while T1 exits below. The brachial plexus is "sandwiched" between the anterior and middle scalene muscles.* **(Right)** *Sagittal oblique STIR MR distal to the root level shows formation of the upper, middle, and lower trunks arranged in a vertical line between the scalene muscles.*

(Left) *Sagittal oblique STIR MR of the right brachial plexus distal to the trunks and divisions demonstrates formation of the 3 cords (lateral, medial, and posterior).* **(Right)** *Axial STIR MR depicts the C5, C6, and C7 ventral primary rami transiting between the anterior and middle scalene muscles in preparation to form the upper brachial plexus. This image demonstrates the conspicuity of the brachial plexus on routine cervical spine MR imaging studies.*



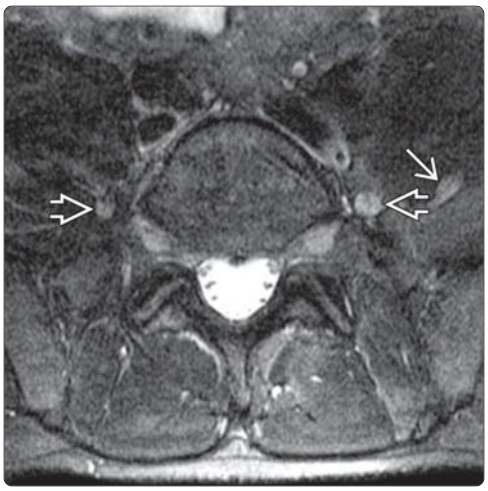


(Left) *Coronal graphic of the lumbosacral plexus demonstrates the lumbar plexus ventral to quadratus lumborum and iliacus muscles and medial to psoas muscle. The femoral nerve, the major terminal branch of the lumbar plexus, travels between iliacus and psoas muscles under inguinal ligament. The sciatic nerve is the major output of the lumbosacral plexus.* **(Right)** *Coronal T2WI FS MR shows the L4 and L5 nerve root forming the lumbosacral trunk.*

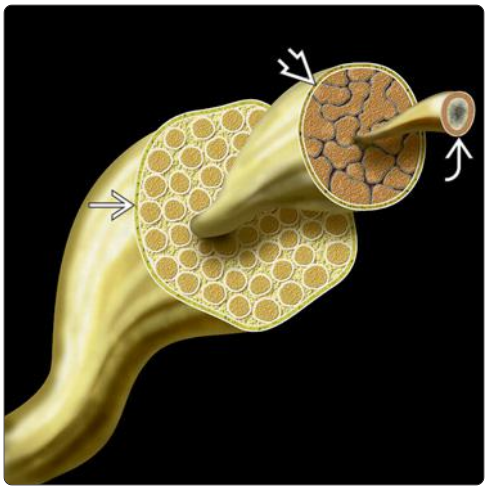


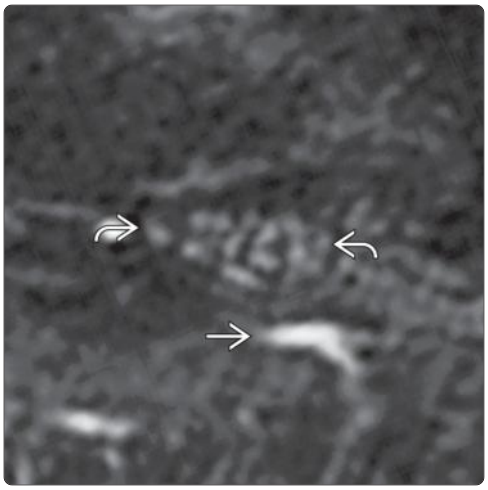


(Left) *Axial T2WI FS MR at the lumbosacral junction demonstrates the bilateral lumbar plexus after the L4 contribution in expected location medial to the psoas muscles. The left femoral nerve has branched off and is tracking within the iliopsoas groove in expected location.* **(Right)** *Coronal T1WI MR through the pelvis demonstrates the normal left S2 nerve transiting its named neural foramen to contribute to sacral plexus and left sciatic nerve as it passes through the sciatic notch.*

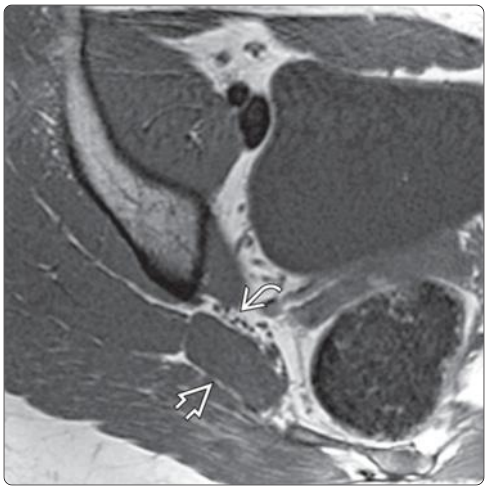


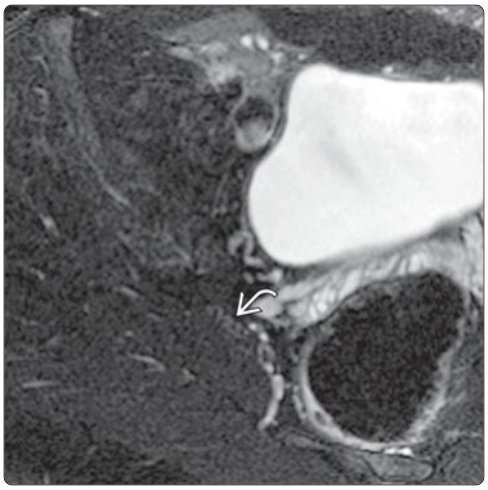




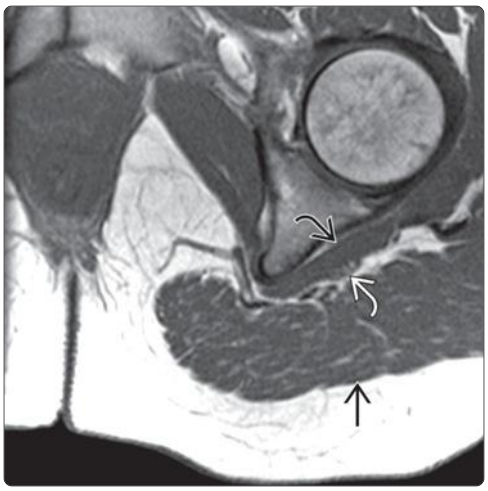


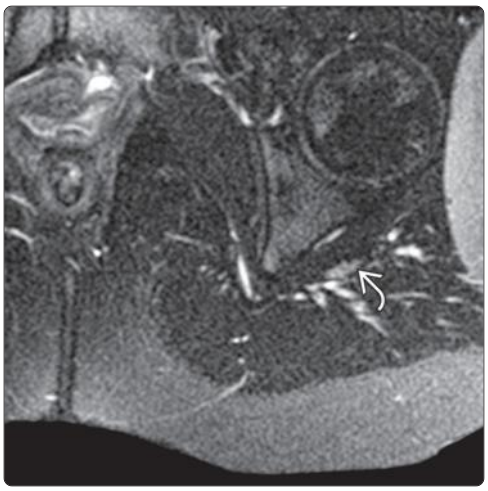
(Left) *3D graphic depicts normal peripheral nerve hierarchical anatomy. The peripheral nerve itself, composed of numerous fascicles, is surrounded by epineurium. Each fascicle is individually enveloped by perineurium. Each fascicle contains many endoneurium wrapped axons.* **(Right)** *Axial STIR MR of the normal sciatic nerve shows normal peripheral nerve imaging characteristics. The fascicles are mildly hyperintense to muscle, but not as bright as regional vessels.*





(Left) *Axial oblique T1WI MR shows normal sciatic nerve on the ventral piriformis muscle. Although the nerve (largest single nerve in the body) is enveloped by epineurium, the abundant fibrofatty epineurium gives the impression that individual fascicles are free in pelvic fat.* **(Right)** *Axial oblique T2WI FS MR demonstrates normal sciatic nerve with distinctive mildly hyperintense fascicles separated by interspersed dark (fat-suppressed) fibrofatty connective tissue.*





(Left) *Axial T1WI MR of normal sciatic nerve at the obturator internus level is readily identified between the obturator internus and gluteus maximus muscles. Normal sciatic nerve is smaller and flatter at this level than at piriformis level.* **(Right)** *Axial T2WI FS MR shows normal sciatic nerve at level of gluteus maximus and obturator internus. The normal neural fascicular architecture is distinctive and permits nerve discrimination from adjacent veins.*

Superior Sulcus Tumor

KEY FACTS

TERMINOLOGY

- Synonym: Pancoast tumor
- Benign or malignant neoplasm extending to superior thoracic inlet with severe shoulder/arm pain along C8, T1, T2 nerve trunks, Horner syndrome, weakness + atrophy of intrinsic hand muscles (Pancoast syndrome)

IMAGING

- Soft tissue apical lung mass with variable extension into chest wall, adjacent bone destruction, brachial plexus invasion

TOP DIFFERENTIAL DIAGNOSES

- Metastatic disease
- Other thoracic tumors (besides non-small cell lung carcinoma [NSCLC])
- Brachial plexus neural tumors
- Hematologic neoplasms
- Radiation fibrosis
- Vascular (venolymphatic) malformation
- Infection

PATHOLOGY

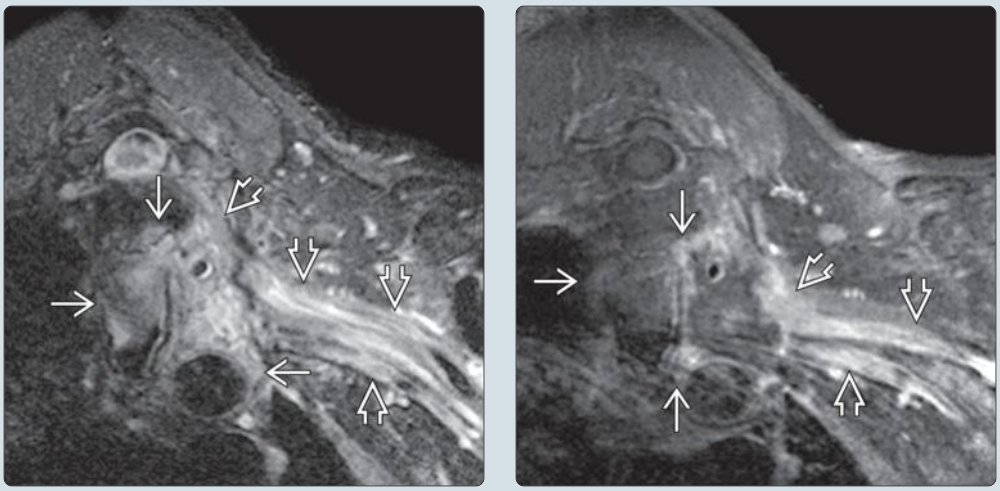
- Bronchogenic carcinomas may arise from either upper lobe
- Invades parietal pleura, endothoracic fascia, subclavian vessels, brachial plexus, vertebral bodies, and upper ribs
- NSCLC most frequent etiology

CLINICAL ISSUES

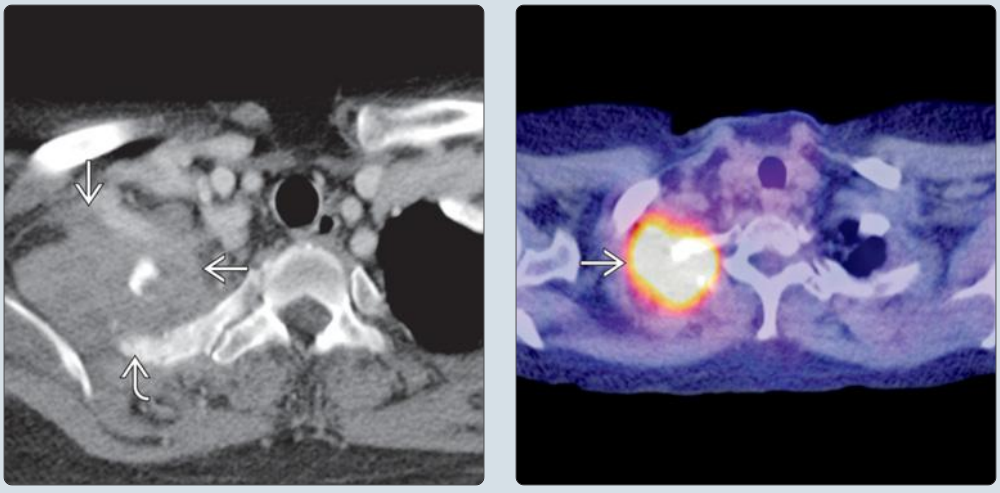
- Clinical findings determined by tumor location relative to scalene muscles
- Severe shoulder and arm pain
- Pulmonary symptoms uncommon early in disease course

DIAGNOSTIC CHECKLIST

- Apical lung mass with bone destruction = bronchogenic carcinoma until proven otherwise
- Rare benign tumors or infection may mimic lung carcinoma



(Left) *Coronal STIR MR (non-small cell lung carcinoma [NSCLC], arm pain and weakness) shows extensive architectural distortion of the lung apex by a peripheral lung mass with extension along the brachial plexus elements.* **(Right)** *Coronal T1 C+ FS MR (NSCLC, arm pain and weakness) demonstrates a heterogeneously enhancing apical lung neoplasm with tumor extension along the lower brachial plexus elements.*



(Left) *Axial CECT demonstrates a soft tissue mass involving the right upper lobe with extension beyond the chest wall producing destruction of right T1, T2 ribs.* **(Right)** *Axial PET (FDG) CT fusion image reveals marked hypermetabolic radiotracer uptake within the right upper lobe bronchogenic carcinoma.*

TERMINOLOGY

Synonyms

- Pancoast tumor

Definitions

- Benign or malignant neoplasm extending to superior thoracic inlet with severe shoulder/arm pain along C8, T1, T2 nerve trunks, Horner syndrome, weakness + atrophy of intrinsic hand muscles (Pancoast syndrome)
  - Non-small cell lung carcinoma (NSCLC) most frequent etiology

IMAGING

General Features

- Best diagnostic clue
  - Soft tissue mass involving lung apex with adjacent rib destruction
- Location
  - Lung apex and adjacent chest wall, brachial plexus, cervicothoracic junction vertebral bodies
- Size
  - Variable
- Morphology
  - Apical soft tissue mass with ill-defined margins, bone destruction

Radiographic Findings

- Radiography
  - Soft tissue density at lung apex, ± 1st or 2nd rib destruction

CT Findings

- CECT
  - Soft tissue attenuation mass involving lung apex, with variable extension into chest wall and adjacent bone destruction
    - 1st, 2nd rib bone destruction
    - May show vertebral body invasion with epidural/foraminal extension ± spinal cord compression
- CTA
  - May confirm encasement or narrowing of subclavian artery and branches

MR Findings

- T1WI
  - Intermediate signal mass involving lung apex, with chest wall/paravertebral extension
    - Demonstrates encasement of subclavian vessels, relationship of mass to brachial plexus elements
- T2WI
  - ± T2 hyperintense apical lung mass with chest wall/paravertebral extension
- T1WI C+
  - Variably enhancing mass
    - Fat suppression improves detection of epidural/foraminal extension
- MRA
  - 2D and 3D TOF techniques or TRICKS contrast dynamic imaging confirms aortic arch/proximal great vessel involvement

Angiographic Findings

- Conventional
  - Generally superseded by MRA/CTA for evaluation of great vessel encasement by tumor

Nonvascular Interventions

- Myelography
  - May show typical extradural defect within myelographic contrast column at cervicothoracic junction in advanced cases with epidural tumor extension

Nuclear Medicine Findings

- Bone scan
  - Positive with rib/vertebral body involvement ± diffuse bone metastases
- PET
  - Positive with primary cancer and nodal involvement
    - Mediastinoscopy performed regardless of PET positive nodes since N2, N3 node involvement is negative prognostic factor

Imaging Recommendations

- Best imaging tool
  - MR and CT complementary techniques
    - MR demonstrates apical soft tissue involvement and relationship to brachial plexus/subclavian vessels
    - CT shows mediastinal nodes and pulmonary, hepatic, or adrenal metastases
- Protocol advice
  - Multiplanar T1WI, T2WI, T1 C+ FS MR to define bone and epidural extension, relationship to subclavian vessels/scalene musculature

DIFFERENTIAL DIAGNOSIS

Metastatic Disease

- Usually nodal metastasis from breast or lung cancer

Other Thoracic Tumors (Besides NSCLC)

- Mesothelioma
- Hemangiopericytoma
- Sarcomas

Brachial Plexus Neural Tumors

- Schwannoma shows enhancing neural mass tracking between anterior + middle scalene muscles
- Neurofibroma infiltrates neural architecture of brachial plexus elements, enhancement often subtle

Hematologic Neoplasms

- Lymphoma
- Plasmacytoma

Radiation Fibrosis

- Hypointense on T2-weighted images in treatment field
- Variable contrast enhancement
- May also insinuate into brachial plexus

Vascular (Venolymphatic) Malformation

- Marked T2 hyperintensity (lymphatic malformation), variable enhancement, lobulated margins

Infections

- Staphylococcal pneumonia

- Aspergillosis
- Cryptococcosis
- Tuberculosis
- Hydatid cyst

PATHOLOGY

General Features

- Etiology
  - Bronchogenic carcinomas arising from either upper lobe
  - Cigarette smoking major cause of lung cancer
    - 80% of lung cancer attributed to tobacco exposure
    - 20x increase risk with smokers vs. nonsmokers
- Genetics
  - *EGFR*-overexpressed in 40-80% NSCLC
    - Response to tyrosine kinase inhibitor chemotherapy (which targets epidermal growth factor receptor) dependent upon specific mutation in *EGFR* gene
    - Mutations lead to increased growth factor signaling conferring susceptibility to inhibitor molecule
  - Variety of other genetic abnormalities in lung cancer
    - *TP53* tumor suppressor gene mutation most common, clustered around codon 157
    - *KRAS* mutations associated with smoking (ras oncogenes encode for GDP/GTP binding proteins)
    - *PTGS2*, *ERBB2*, *VEGFA* overexpression
- Carcinoma invades parietal pleura, endothoracic fascia, subclavian vessels, brachial plexus, vertebral bodies, and upper ribs

Staging, Grading, & Classification

- Staging/diagnosis by chest x-ray, CT chest, bronchoscopy, needle biopsy
- Tumor extent into brachial plexus assessed by examination, EMG, MR imaging
- Vascular involvement assessed by CTA, MRA, or catheter angiography (arterial and venous)

Gross Pathologic & Surgical Features

- Firm, gray-white polypoid mass infiltrating lung and chest wall

Microscopic Features

- Variety of bronchogenic carcinoma cell types: Squamous > adenocarcinoma > large cell

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain
    - Severe neuropathic pain in shoulder/arm in C8-T2 distribution
    - Pain in upper chest wall with 1st intercostal nerve involvement
  - Pulmonary symptoms uncommon early in disease course
    - Tumors peripheral in location, late appearance of pulmonary symptoms (cough, hemoptysis, dyspnea)
- Other signs/symptoms
  - Horner syndrome (ptosis, anhidrosis, miosis)
  - Atrophy of hand muscles

Demographics

- Age
  - Adult
- Gender
  - M > F
- Ethnicity
  - Marked variation in lung cancer incidence + mortality by ethnicity
- Epidemiology
  - Lung carcinoma leading cause of cancer deaths in USA in both men and women
  - Superior sulcus lesions < 5% of all bronchogenic carcinomas

Natural History & Prognosis

- Median survival with preoperative radiation and resection 22 months, 5-year survival 27%
- N2, N3 lymph node involvement major negative prognostic factor
- Horner syndrome associated with poor survival
- Involvement of vertebral bodies or subclavian vessels associated with poor survival after resection

Treatment

- Induction chemoradiation followed by surgical resection is standard treatment approach
- Other treatments include radiation alone, preoperative radiotherapy followed by resection, or chemotherapy without resection
- Pain palliation with radiation alone successful in 75%
  - Radiation alone long-term survival 5%
- Absolute surgical contraindications include extrathoracic metastatic disease or mediastinal node involvement
  - Brachial plexus invasion above T1 relative contraindication; new microsurgical techniques may permit preservation of hand function when C8 or T1 is involved

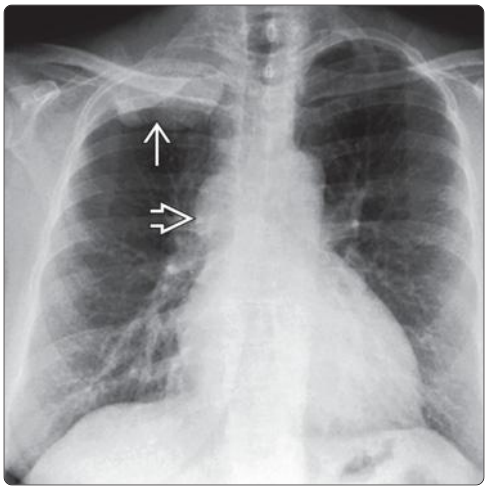
DIAGNOSTIC CHECKLIST

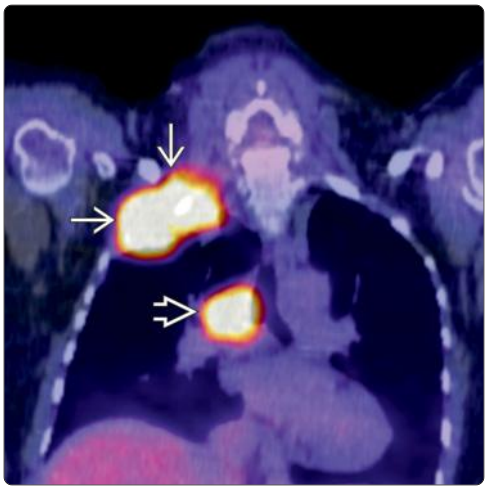
Consider

- Apical lung mass with bone destruction = bronchogenic carcinoma until proven otherwise
- Rare cases of benign tumors or infection may mimic lung carcinoma

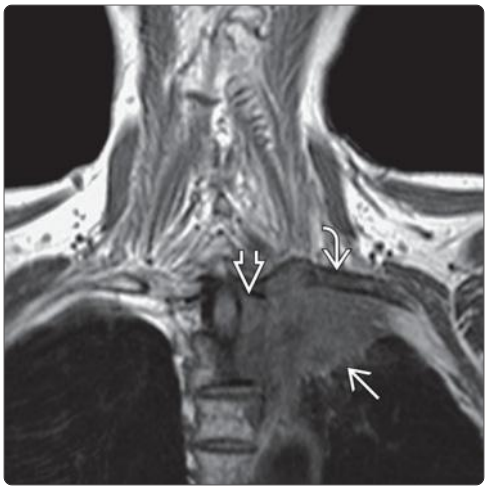
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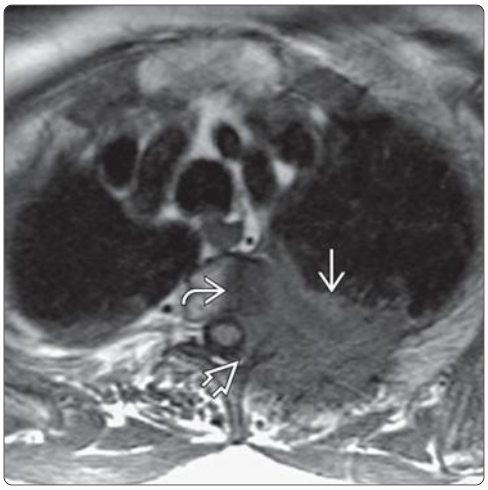
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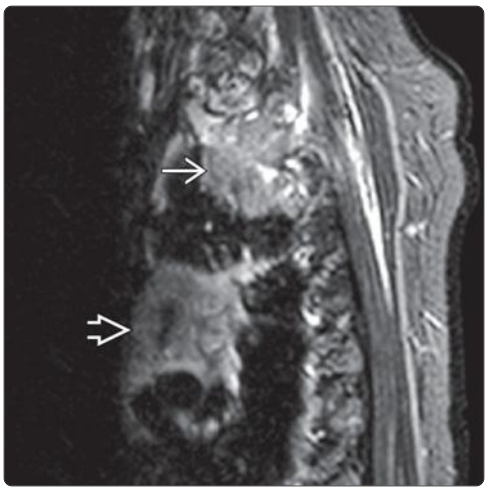


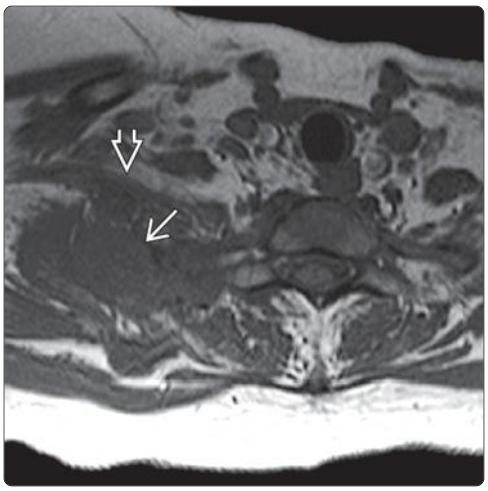
(Left) *Anteroposterior radiograph reveals a lobulated right apical primary lung neoplasm producing bone destruction of the right 2nd and 3rd ribs. Note also the ipsilateral mediastinal lymphadenopathy.* **(Right)** *Coronal PET/CT fusion image confirms intense hypermetabolism within a right apical lung mass. Note also the additional ipsilateral hypermetabolic tumor uptake within right mediastinal adenopathy.*





(Left) *Coronal T1WI MR demonstrates a large infiltrative lung mass involving left posterior chest wall and lung apex with local extension into vertebral body, ribs, paraspinal soft tissues, neural foramina, and epidural space.* **(Right)** *Axial T1WI MR confirms a large invasive lung neoplasm involving the left posterior chest wall, lung apex with extension into vertebral body, paraspinal soft tissues, and neural foramina into the epidural space.*





(Left) *Sagittal STIR MR demonstrates a large hyperintense apical lung mass extending from the posterior lung apex into the adjacent chest wall and soft tissues. Note also ipsilateral mediastinal lymphadenopathy.* **(Right)** *Axial T1WI MR depicts a large soft tissue mass representing local extension of a primary lung neoplasm producing rib destruction and soft tissue extension adjacent to the neurovascular bundle.*

Thoracic Outlet Syndrome

KEY FACTS

TERMINOLOGY

- Thoracic outlet syndrome (TOS)
- Neural, venous, &/or arterial compressive syndrome at thoracic outlet (TO)
- Diagnosis made by 1 of 3 methods (by event, affected structure, or compression cause)

IMAGING

- ± cervical rib, elongated C7 transverse process
- ± brachial plexus compression or distortion, scalene muscle inflammation or fibrosis, abnormal vascular flow voids within TO
- Positional occlusion or narrowing of SCA with arm hyperabduction, external rotation

TOP DIFFERENTIAL DIAGNOSES

- Primary and secondary plexus tumors
- Radiation plexopathy
- Trauma

PATHOLOGY

- Cervical ribs, abnormal transverse processes, fibrous bands, scalene compression of TO contents
- Neural injury with fibrosis, axonal loss

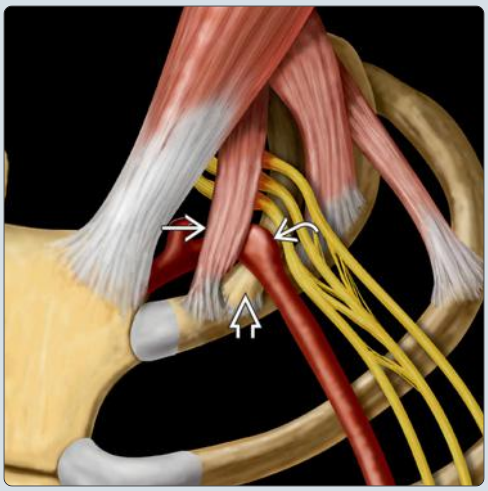
CLINICAL ISSUES

- "True" neurological TOS: Intermittent arm pain, numbness, and weakness with hyperabduction, external rotation
  - Pain in shoulder, proximal upper extremity → neck
  - Paresthesias, numbness in forearm/hand
- Vascular TOS: Paresthesias 2° to arterial or venous ischemia
  - Obliteration of brachial, radial pulses with arm hyperabduction and elevation

DIAGNOSTIC CHECKLIST

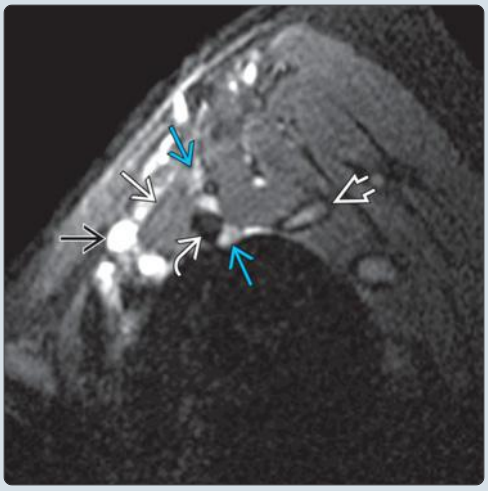
- SCA aneurysm, SCV thrombosis, brachial plexus compression with abnormal T2 hyperintensity at TO strongly suggest TOS

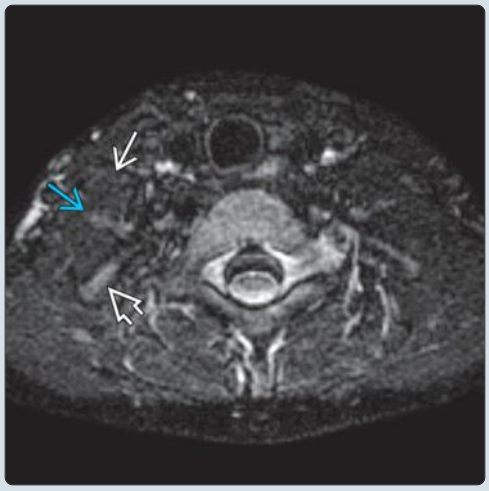
(Left) *Coronal graphic of the thoracic outlet (TO) demonstrates brachial plexus compression + a subclavian artery (SCA) aneurysm arising secondary to the C7 cervical rib and anterior scalene muscle compression.* **(Right)** *Anteroposterior radiograph demonstrates bilateral rudimentary C7 cervical ribs. The right cervical rib articulates with the 1st thoracic rib.*





(Left) *Sagittal oblique STIR MR shows normal anatomy of the interscalene triangle: Brachial plexus (BP) trunks, anterior and middle scalene muscles, SCA, subclavian vein (SCV), and 1st thoracic rib.* **(Right)** *Axial STIR MR demonstrates normal axial thoracic outlet anatomy. The brachial plexus passes between the anterior and middle/posterior scalene muscles and over the 1st thoracic rib.*





TERMINOLOGY

Abbreviations

- Thoracic outlet syndrome (TOS)

Definitions

- Neural, venous, &/or arterial compressive syndrome at thoracic outlet (TO)
- Diagnosis made by 1 of 3 methods (by event, affected structure, or compression cause)

IMAGING

General Features

- Best diagnostic clue
  - Radiography: Cervical rib in symptomatic patient
  - MR: Neural or vascular compression within interscalene or costocervical tunnels
- Location
  - Interscalene, costoclavicular space, retropectoralis minor space (subcoracoid tunnel)

Radiographic Findings

- Radiography
  - Cervical radiographs usually normal
    - ± cervical rib, elongated C7 transverse process

CT Findings

- CECT
  - Usually normal; ± abnormal soft tissue process, scalene thickening, vascular anomaly within TO
- Bone CT
  - ± cervical rib, elongated C7 transverse process
- CTA
  - ± vascular compression, subclavian artery (SCA) aneurysm or subclavian vein (SCV) thrombosis

MR Findings

- T1WI
  - Often normal; abnormal cases → brachial plexus (BP) compression or distortion, abnormal vascular flow void at TO
- T2WI
  - ± focal abnormal brachial plexus T2 hyperintensity in TO
  - ± brachial plexus compression or distortion, scalene muscle inflammation or fibrosis, abnormal vascular flow voids within TO
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± focal enhancement at TO
- MRA
  - ± SCA compression, aneurysm on MRA
  - Positional occlusion or narrowing of SCA with arm hyperabduction, external rotation
- MRV
  - ± subclavian vein compression or thrombosis, positional occlusion or narrowing of SCV

Ultrasonographic Findings

- Grayscale ultrasound
  - Muscular or osseous compression of brachial plexus, SCA, or SCV
- Color Doppler
  - ± SCA aneurysm, SCV thrombosis

Angiographic Findings

- Conventional
  - Positional occlusion/narrowing of SCV or SCA with arm hyperabduction ± mural thrombi, emboli, aneurysm

Imaging Recommendations

- Best imaging tool
  - MR imaging
  - Reported sensitivity (41%) and specificity (33%) for MR diagnosis of TOS relatively low
- Protocol advice
  - Multiplanar T1WI and STIR MR through TO
  - Plain radiography or CT → cervical rib, aberrant C7 transverse process
  - MRA/MRV in neutral, abducted arm position

DIFFERENTIAL DIAGNOSIS

Primary and Secondary Plexus Tumors

- Superior sulcus tumor
- Metastatic breast carcinoma
- Nerve sheath tumor

Radiation Plexopathy

- Diffuse plexus T2 hyperintensity ± enhancement
- Most common following radiation therapy for breast carcinoma, Hodgkin disease
- Clinical history makes diagnosis

Trauma

- Stretch injury or avulsion
- Trauma history, clinical findings make diagnosis

PATHOLOGY

General Features

- TO consists of interscalene triangle, costoclavicular space, and retropectoralis minor space (subcoracoid tunnel)
  - Compression of neural, arterial, or venous structures crossing these tunnels → TOS
  - Narrowing of costoclavicular distance may be most important abnormality in symptomatic patients
- Neuropathic TOS: Symptomatology 2° to brachial plexus compression (most symptomatic patients)
  - Up to 98% of symptomatic patients have plexus compression; minority 2° to arterial or venous impingement
- Vascular TOS: Compression of subclavian vessels
  - Repetitive arterial trauma → focal stenosis, aneurysm formation, microembolization, tissue loss
  - Venous compression → SCV thrombosis
- Interscalene triangle bordered by anterior scalene muscle anteriorly, middle and posterior scalene muscles posteriorly, and 1st rib inferiorly
  - SCA passes through lower portion of interscalene triangle, superior (C5-C6) and middle (C7) trunks of brachial plexus pass through upper portion, and lower (C8-T1) trunk crosses inferior portion behind SCA

- SCV courses between clavicle anteriorly and anterior scalene muscle posteriorly
- Congenital bony/fibromuscular anomalies, trauma, phenotype, and posture contribute to compression or elongation of neurovascular bundle as it passes through these compartments
  - Raising arms above head further compresses already narrowed TO
- Etiology
  - Neural > > vascular compression (symptomatic patients)
- Epidemiology
  - TOS incidence, prevalence disputed; may be ≤ 1/1,000,000
  - Cervical ribs: 6% of general population → 13% of TOS patients, up to 90% of patients receiving surgery for SCA aneurysm
- Associated abnormalities
  - Cervical ribs
  - Unusually large or elongated C7 transverse process
  - Fibrous band (1%)
  - Klippel-Feil syndrome

Gross Pathologic & Surgical Features

- Vascular compression, SCA aneurysm, SCV thrombosis may be noted at surgery
- Nerves may be swollen and edematous or atrophic and fibrotic
- Cervical ribs, abnormal transverse processes, fibrous bands, scalene compression of TO contents

Microscopic Features

- Neural injury with fibrosis, axonal loss

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Paresthesias, numbness in forearm/hand
  - Pain in shoulder, proximal upper extremity → neck
  - Obliteration of brachial, radial pulses with arm hyperabduction and elevation
- Clinical profile
  - "True" neurological TOS: Intermittent arm pain, numbness, and weakness with hyperabduction, external rotation
    - Lower trunk (C8, T1) ≥ upper, middle trunks
    - Nearly always 2° to fibrous band extending from cervical rib compressing lower trunk
    - Hand wasting, concordant EMG findings
  - "Disputed" neurological TOS: No consistent clinical or laboratory findings
  - Vascular TOS: Paresthesias 2° to ischemia
    - More diffuse than neurological TOS
    - ± obliteration of brachial, radial pulses

Demographics

- Age
  - Adult > > children; average age at diagnosis 26 years
- Gender
  - M < F

Natural History & Prognosis

- Symptomatic patients rarely recover spontaneously without treatment
- Postoperative prognosis of vascular TOS better than neurological TOS

Treatment

- Initial conservative treatment with physical therapy, analgesics
  - Role of anesthetic injections unclear; may be helpful for diagnosis
- TOS decompression ± 1st rib resection for conservative treatment failures

DIAGNOSTIC CHECKLIST

Consider

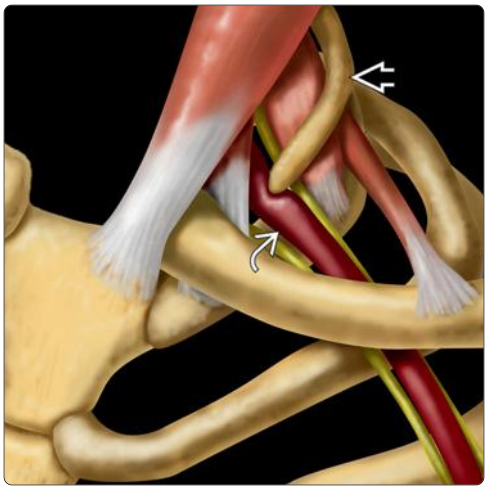
- TOS probably end result of diverse symptomatic etiologies rather than a single disease process

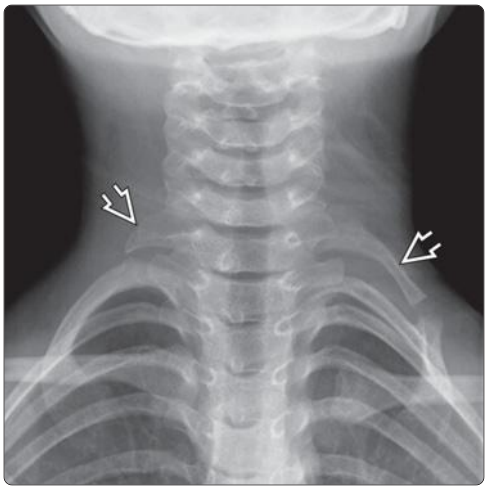
Image Interpretation Pearls

- SCA aneurysm, SCV thrombosis, brachial plexus compression with abnormal T2 hyperintensity at TO strongly suggest TOS

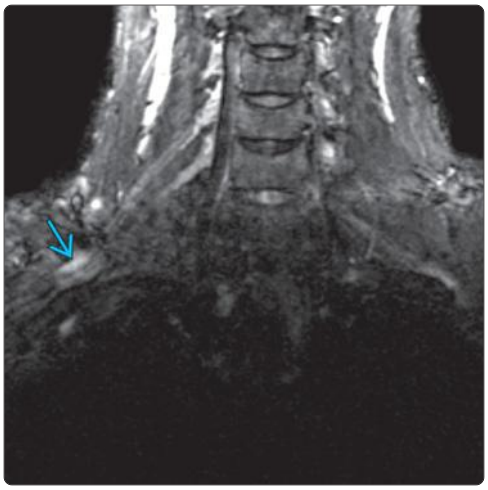
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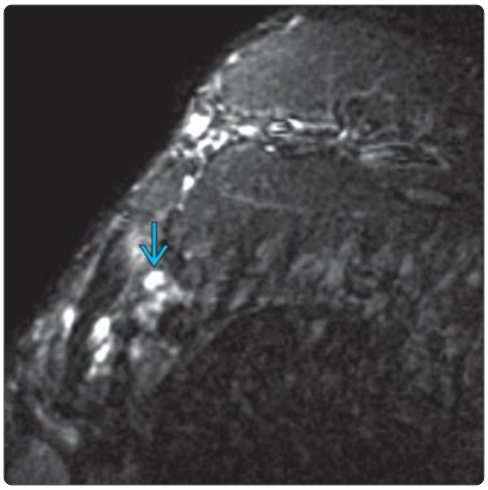
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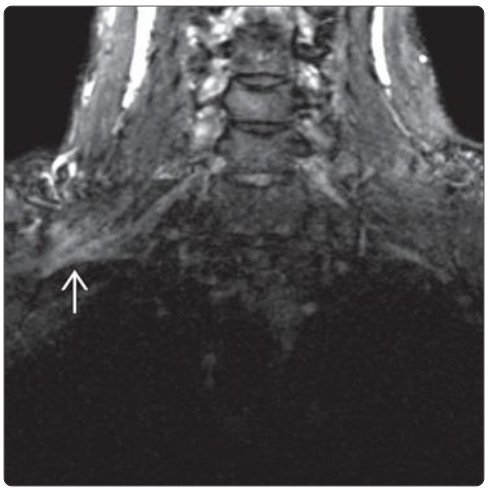


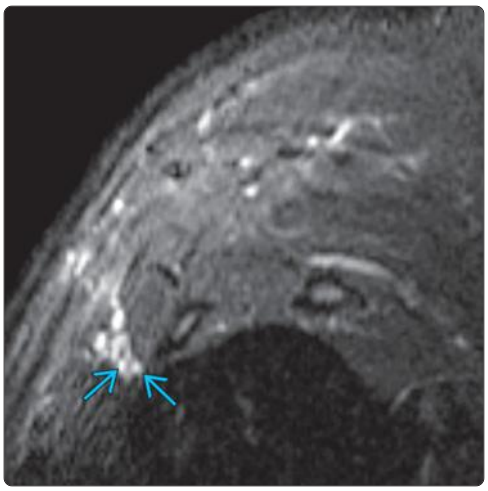
(Left) *Coronal graphic demonstrates a vascular thoracic outlet syndrome (TOS) variant with a predominately SCA compression produced by an elongated C7 cervical rib.* **(Right)** *Anteroposterior radiograph (palpable left supraclavicular mass, TOS) reveals bilateral C7 cervical ribs. The left C7 cervical rib is elongated and shows an aberrant articulation with the left 1st thoracic rib.*





(Left) *Coronal STIR MR (right neural TOS) demonstrates focal abnormal T2 hyperintensity of the brachial plexus upper trunk elements within the TO.* **(Right)** *Sagittal oblique STIR MR depicts focal T2 hyperintensity in the brachial plexus upper trunk corresponding to clinical right neural TOS symptoms.*





(Left) *Coronal STIR MR in a professional musician (drummer) with symptomatic right neural TOS shows abnormal brachial plexus T2 hyperintensity within the TO.* **(Right)** *Sagittal oblique STIR MR (in a professional drummer) through the TO confirms mild T2 hyperintensity of some fascicles within the lower trunk, correlating with the clinical presentation with lower brachial plexopathy.*

Muscle Denervation

KEY FACTS

TERMINOLOGY

- Secondary muscle injury resulting from denervation following nerve injury

IMAGING

- **Acute denervation**
  - Muscle appears normal on T1WI
  - Diffusely increased signal intensity on T2WI, STIR
  - Mild, homogeneous enhancement with gadolinium
- **Chronic denervation**
  - Fatty atrophy evident on T1WI
  - Muscle volume decreased
  - Denervation edema often persists for prolonged period

TOP DIFFERENTIAL DIAGNOSES

- Disuse atrophy
- Muscle trauma
- Muscle inflammation or infection
- Radiation myopathy

PATHOLOGY

- Nerve tumor, infection, autoimmune neuritis, peripheral neuropathy or injury

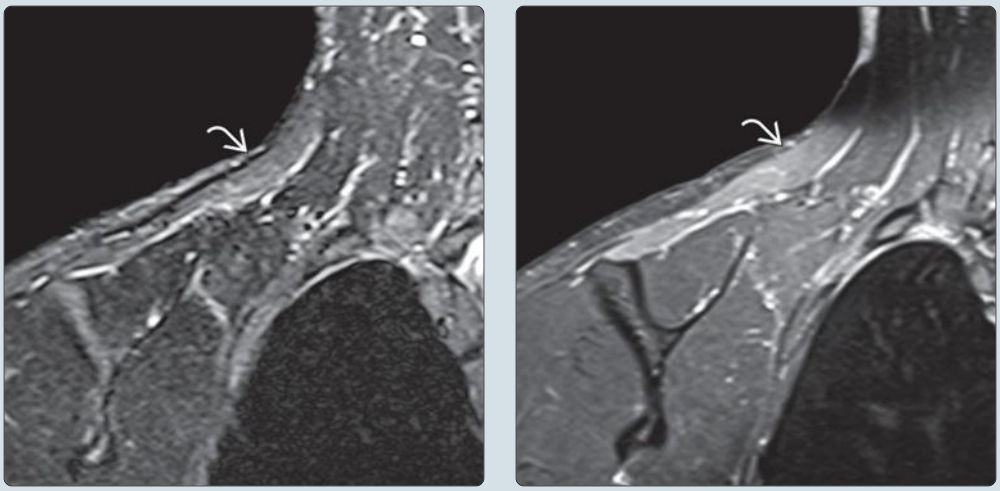
CLINICAL ISSUES

- Weakness, muscle volume loss in distribution of injured nerve
  - May be painful
- Acute denervation may partially or totally recover depending on nerve injury severity
- Chronic denervation changes permanent

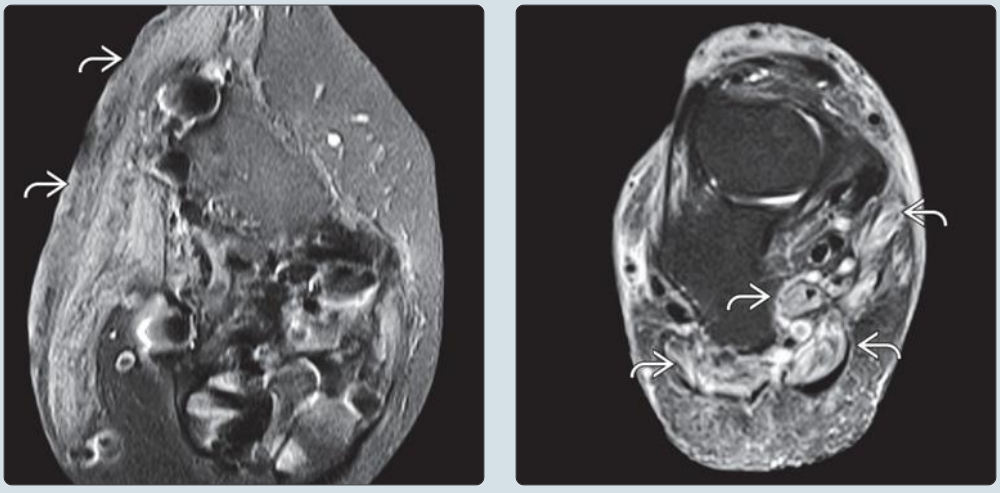
DIAGNOSTIC CHECKLIST

- Distribution of muscle signal abnormalities indicates location of nerve lesion
- Obtain fluid-sensitive sequence that shows muscle of concern in cross section

(Left) *Coronal STIR MR shows denervation edema of the trapezius, which is innervated by the spinal accessory nerve. This nerve can be traumatized by carrying heavy loads on the shoulder.* **(Right)** *Coronal T1WI C+ FS MR in the same patient shows uniform enhancement of the affected muscle. Muscle tears, in contrast, had a heterogeneous appearance.*



(Left) *Axial T2WI FS MR shows a free muscle flap to the leg. The muscle shows severe, diffuse edema with a few streaks of fat interspersed. This is an expected finding and should not be confused with infection.* **(Right)** *Coronal T2WI FS MR shows denervation edema throughout the intrinsic muscles of the foot. Denervation edema due to diabetes primarily involves the feet. It tends to be streakier and less homogeneous than denervation from other causes.*



TERMINOLOGY

Definitions

- Secondary muscle injury resulting from denervation following nerve injury

IMAGING

General Features

- Best diagnostic clue
  - Diffusely increased signal intensity on T2WI, STIR with intact muscle fibers
- Size
  - Acute: ± enlarged 2° muscle edema
  - Chronic: Atrophic muscle

CT Findings

- Muscle often appears normal in acute denervation
- ± mild muscle enlargement ± enhancement (acute)
- Volume loss, hypodense fatty replacement (chronic)

MR Findings

- **Acute denervation**
  - Muscle shows normal signal intensity on T1WI
  - Diffusely increased signal intensity on T2WI, STIR
    - "Denervation edema"
    - Muscle is homogeneously involved
      - Rarely, if distal muscular nerve branch is involved, only a segment of muscle is affected
    - Muscle fibers are intact
      - Helps to distinguish from muscle tear
    - Small amount of fluid often seen surrounding muscle
  - Mild, homogeneous enhancement with gadolinium
- **Chronic denervation**
  - Fatty atrophy evident on T1WI
  - Muscle volume decreased
  - Denervation edema often persists for prolonged period
  - ± enhancement with contrast

Other Modality Findings

- Needle electromyography (EMG)
  - Up to 3 weeks after injury before EMG abnormal

Imaging Recommendations

- Best imaging tool
  - MR imaging
- Protocol advice
  - Imaging planes chosen to show affected muscle in cross section

DIFFERENTIAL DIAGNOSIS

Disuse Atrophy

- Fatty atrophy of muscle
- Edema usually absent

Muscle Trauma

- Disruption/discontinuity of muscle fibers
- Hematoma usually appears heterogeneous on T2WI, STIR

Tendon Tear

- Large tendon tears lead to muscle atrophy

Muscle Inflammation or Infection

- Abnormal T2 signal and contrast enhancement are more patchy and heterogeneous
- Multiple different causes, including
  - Polymyositis/dermatomyositis
  - Infectious myositis
  - Diabetic myonecrosis
  - Rhabdomyolysis
  - Sickle cell crisis

Radiation Myopathy

- Vasculitis, tissue injury → uniform muscle edema
- Straight, sharp edema margins correspond to radiation port

PATHOLOGY

General Features

- Etiology
  - Nerve tumor, infection, autoimmune neuritis, peripheral neuropathy or injury

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Weakness, muscle volume loss in distribution of injured nerve
  - May be painful

Natural History & Prognosis

- Acute denervation may partially or totally recover depending on nerve injury severity
- Chronic denervation changes permanent

Treatment

- Address cause of nerve injury if applicable
- Physical therapy to strengthen residual muscle mass

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Distribution of muscle signal abnormalities indicates location of nerve lesion

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Brachial Plexus Traction Injury

KEY FACTS

TERMINOLOGY

- Synonyms: Brachial plexus stretch injury, brachial plexus avulsion, avulsion pseudomeningocele
- Stretch injury or avulsion of ≥ 1 cervical roots, brachial plexus

IMAGING

- Stretch injury: Enlargement, abnormal edema of neural elements
- Avulsion: Abnormal CSF signal intensity within empty thecal diverticulum

TOP DIFFERENTIAL DIAGNOSES

- Nerve sheath tumor
- Lateral meningocele
- Nerve root sleeve cyst
- CIDP

PATHOLOGY

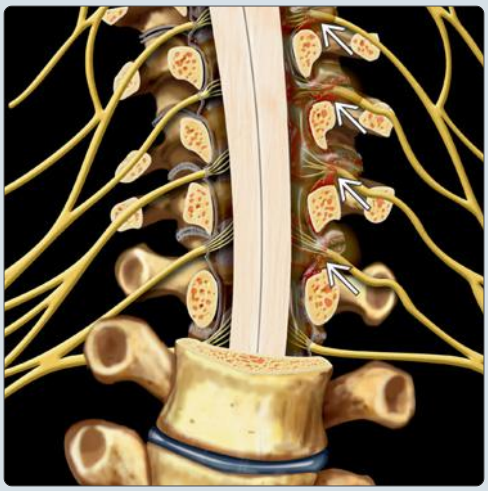
- Adults: Majority post-traumatic injuries 2° to high-energy force
- Infants: 2° to excessive traction on plexus during difficult delivery (breech or forceps)

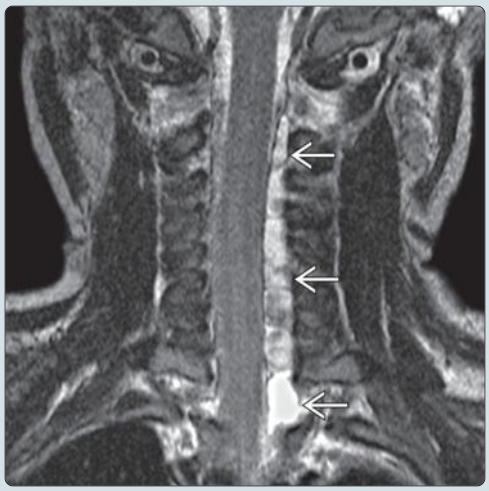
CLINICAL ISSUES

- Complete brachial plexus avulsion produces useless "flail arm"
- Clinical incomplete paralysis possible with complete root avulsion(s) because of redundant muscle innervation from multiple roots

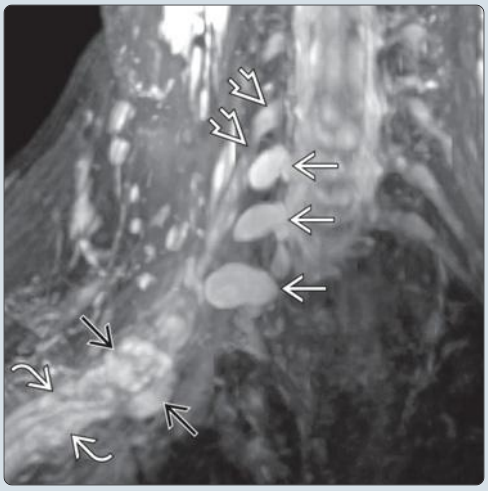
DIAGNOSTIC CHECKLIST

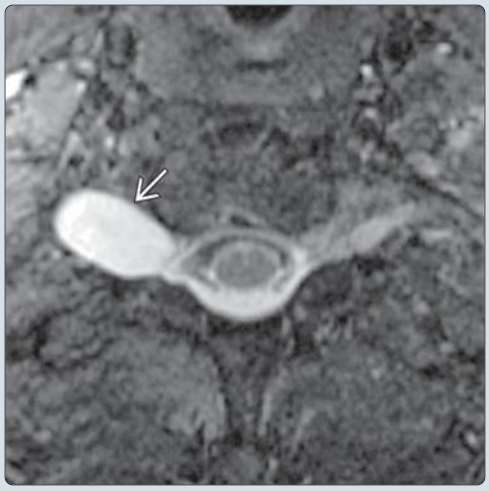
- Familiarity with normal brachial plexus anatomy essential for MR interpretation
- Muscle denervation pattern predicts abnormal nerves





(Left) *Coronal graphic demonstrates post-traumatic avulsion of left C5-C8 nerve roots producing local hemorrhage at the site of root injury and associated pseudomeningoceles.* **(Right)** *Coronal T2WI MR in a patient with post-traumatic paralyzed "flail arm" demonstrates an extensive extradural CSF signal intensity collection admixed with blood products, representing CSF leakage following avulsion of multiple nerve roots, displacing the spinal cord to the right.*





(Left) *Coronal STIR MR in a patient with severe brachial plexus traction injury shows avulsion pseudomeningoceles of right C6, C7, and C8 roots. The C5 root is attenuated with abnormally ↑ signal (incomplete stretch injury). The avulsed nerve roots have formed a "retraction ball", with abnormal enlargement and T2 hyperintensity of the distal (denervated) plexus.* **(Right)** *Axial STIR MR confirms the CSF signal and lack of demonstrable neural elements within the right C7 pseudomeningocele.*

TERMINOLOGY

Synonyms

- Brachial plexus stretch injury, brachial plexus avulsion, avulsion pseudomeningocele

Definitions

- Stretch injury or avulsion of ≥ 1 cervical roots, brachial plexus

IMAGING

General Features

- Best diagnostic clue
  - Lateral CSF-containing dural sac outpouching(s) devoid of neural elements
- Location
  - Brachial plexus, pre- or postganglionic nerve roots
- Morphology
  - Stretch injury: Enlargement or attenuation of stretched (but contiguous) plexus elements
  - Avulsion injury: Attenuated or disrupted proximal roots/rami within or immediately distal to diverticulum ± retracted distal nerve roots, nerve "retraction ball"
  - Obstetric brachial plexus palsy (OBPP): C5-6 roots (Erb-Duchenne palsy, 80%) > C8-T1 nerve roots (Klumpke palsy, 20%)

CT Findings

- NECT
  - Difficult CT diagnosis
  - ± pseudomeningocele(s), paraspinal hematoma
  - Most useful following myelography
- Bone CT
  - ± vertebral fracture(s)

MR Findings

- T1WI
  - Enlargement, hypo- to isointense signal of neural elements (stretch injury)
  - Dilated hypointense CSF signal intensity within empty thecal diverticulum (avulsion)
- T2WI
  - Enlargement, hyperintense signal of neural elements (stretch injury)
  - Hyperintense CSF signal intensity within empty thecal diverticulum (avulsion)
  - ± spinal cord edema, myelomalacia, syringomyelia
    - Central spinal cord edema in acute stage 2° to root avulsion
  - Denervation changes in posterior cervical paraspinal muscles (especially multifidus) → preganglionic injury
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - Enhancement absent in acute injury; chronic injury may show enhancing scar tissue
  - Enhancing muscle implies denervation

Nonvascular Interventions

- Myelography
  - ± dilated empty root sleeves, CSF/contrast leak, spinal cord "divot" at avulsion site

Imaging Recommendations

- Best imaging tool
  - High-resolution MR (MR neurography)
- Protocol advice
  - Coronal, sagittal oblique T1WI and STIR MR
  - CT myelography if MR inconclusive

DIFFERENTIAL DIAGNOSIS

Nerve Sheath Tumor

- Plexiform neurofibroma (NF1), solitary nerve sheath tumor mimics root avulsion(s)
  - + tumoral enhancement
- Look for clinical, genetic, or imaging stigmata of NF1

Lateral Meningocele

- Marfan syndrome, Ehlers-Danlos syndrome, NF1
- Lateral meningocele mimics pseudomeningocele, but contains neural elements
- Clinical history/stigmata ± dural dysplasia distinguish from trauma

Nerve Root Sleeve Cyst

- Usually incidental finding; may be large
- Intact nerve root peripherally displaced by cyst
- Spontaneous rupture → intracranial hypotension
- Clinical history helps distinguish from avulsion

CIDP

- Characteristic clinical, laboratory findings; often bilateral

PATHOLOGY

General Features

- Etiology
  - Majority of adult brachial plexus palsies are post-traumatic injuries 2° to high-energy force
    - Forced abduction or downward displacement of arm
    - Penetrating injury following gunshot wound or lacerations (knives, glass, automobile metal, chainsaw, or animal bite)
  - In infants, brachial plexus palsy 2° to excessive traction on plexus during difficult delivery (breech or forceps)
- Associated abnormalities
  - ± coexisting soft tissue or vascular injury, vertebral fractures
  - ± muscle denervation; pattern predicts abnormal nerve roots
  - Transdural spinal cord herniation (rare)
- Brachial plexus traction avulses 1 or more nerve roots from spinal cord
- Injury level critical to treatment planning, prognosis
  - Preganglionic injury: Lesion central to dorsal root ganglion (worse prognosis)
  - Postganglionic injury: Injury peripheral to ganglion (better prognosis)

Staging, Grading, & Classification

- Bonney classification (1998)

- Type A: Roots torn central to transition zone (true avulsion)
- Type B: Roots torn distal to transition zone
  - Type 1: Dura torn within spinal canal, DRG displaced into neck
  - Type 2: Dura torn at mouth of foramen, DRG more or less displaced
  - Type 3: Dura not torn, DRG not displaced
  - Type 4: Dura not torn, DRG not displaced, either ventral or dorsal roots intact

Gross Pathologic & Surgical Features

- Evidence for root avulsion: Spinal cord displacement/edema, hemorrhage or scarring in spinal canal, absence of roots in intervertebral foramina, pseudomeningocele

Microscopic Features

- Nerve discontinuity, axonal loss, demyelination, hemorrhage, scar

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, paralysis of ipsilateral limb ± phrenic nerve palsy
    - Complete brachial plexus avulsion produces useless "flail arm"
    - Incomplete paralysis is common, however, with complete root avulsion(s) because of redundant muscle innervation from multiple roots
  - Clinical examination cannot reliably distinguish between pre- and postganglionic injuries
- Clinical profile
  - Erb-Duchenne palsy
    - Upper plexus injury (C5, C6 roots, upper trunk) → proximal muscle weakness
    - Direct blow to shoulder, birth traction injury
  - Middle radicular syndrome
    - C7 root, middle trunk → radial nerve abnormalities
  - Klumpke palsy
    - Lower plexus injury (C8, T1 roots, lower trunk) → distal muscle weakness
    - Fall with overhead grasp slowing descent

Demographics

- Age
  - Young to middle-aged adult > infants, children
- Gender
  - M > F; follows trauma epidemiology
- Epidemiology
  - Strong association with off-road vehicle, motorcycle accidents
  - Incidence follows trauma epidemiological patterns

Natural History & Prognosis

- Variable; generally poor in avulsion, better with stretch injury + nerve contiguity
- Postganglionic injury has better prognosis than preganglionic injury
- Obstetrical injury has better prognosis than adult stretch injuries
- Integrity of nerve roots critical for surgical decision-making, prognostication
  - Stretched (but contiguous) nerves may recover some function, while avulsed nerve roots produce irreversible sensory and motor deficits
  - Preganglionic avulsions are usually not amenable to surgical repair; function of some denervated muscles is restored with nerve transfers from accessory nerves, cervical plexus, or intercostal nerves
  - Postganglionic avulsions are repaired with excision of damaged segment and nerve autograft between nerve ends

Treatment

- Conservative management
  - Physical therapy, rehabilitation
- Surgical management
  - Reanastomosis of avulsed roots to spinal cord is not generally advocated, but new microsurgical techniques are prompting rereview
  - Nerve bypass or graft, reconstruction with neurolysis, nerve graft, nerve transfer, functioning muscle/tendon transfer
  - Amputation of nonfunctional or painful "flail arm"
  - Dorsal root entry zone (DREZ) lesion palliation for intractable pain

DIAGNOSTIC CHECKLIST

Consider

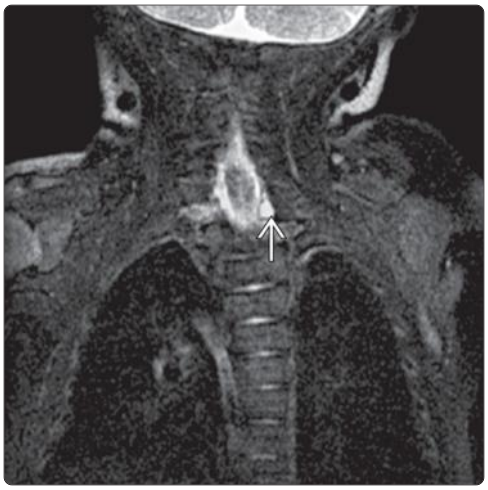
- Familiarity with normal brachial plexus anatomy is essential for MR interpretation
- MR may distinguish pre- from postganglionic injury for surgical planning, prognostication

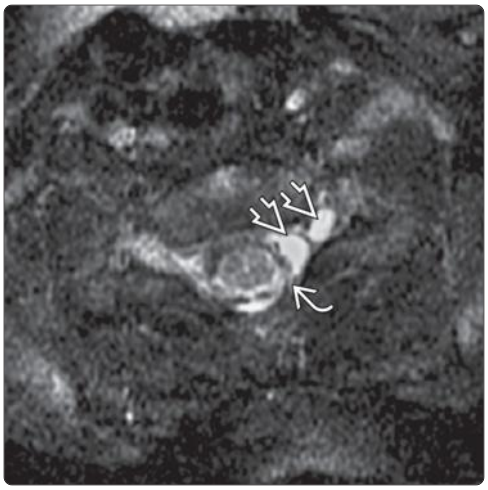
Image Interpretation Pearls

- Search for soft tissue trauma (edema, hemorrhage) or fracture to support diagnosis
- Muscle denervation pattern predicts abnormal nerves

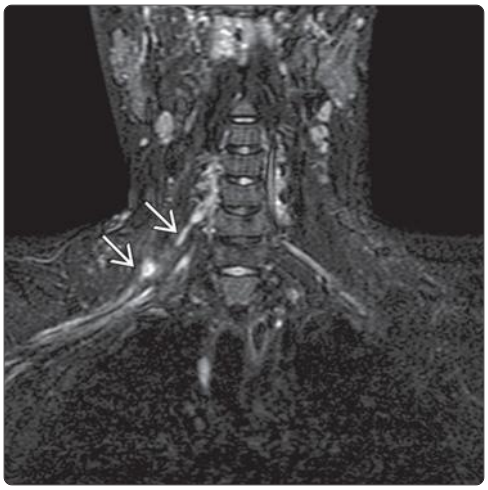
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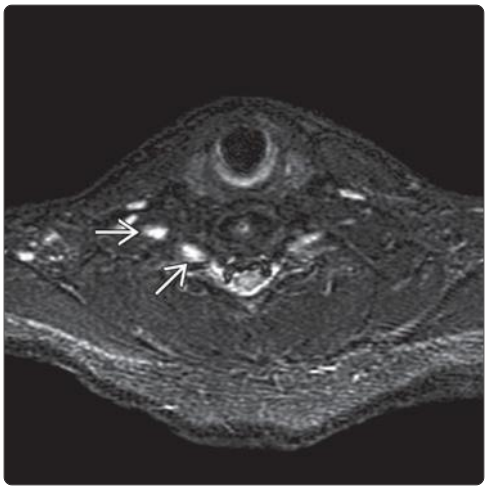
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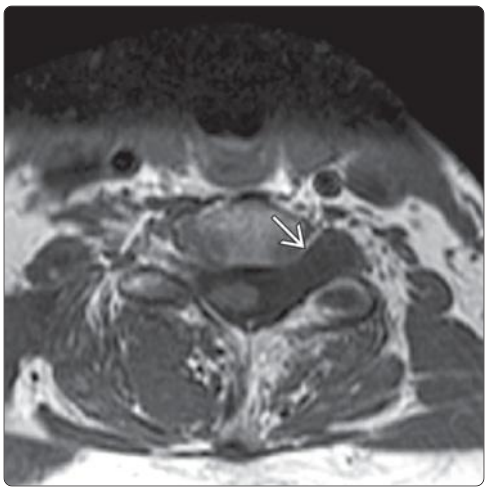


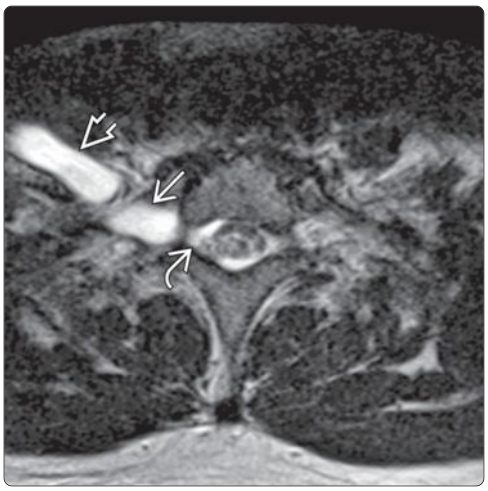
(Left) *Coronal T2WI FS MR in a patient with obstetrical left brachial plexus stretch injury and Klumpke palsy demonstrates a small left C8 CSF signal intensity avulsion pseudomeningocele.* **(Right)** *Axial T2WI FS MR in a patient with obstetrical left brachial plexus stretch injury and Klumpke palsy demonstrates extradural compression on the thecal sac from the adjacent empty nerve root sleeve sac avulsion pseudomeningocele.*





(Left) *Coronal STIR MR in a patient with combination of projectile- and traction-related partial transection of the right brachial plexus following a gunshot injury depicts complete transection of the C6 ramus. No avulsion pseudomeningocele is detected.* **(Right)** *Axial STIR MR in the same patient demonstrates abnormal enlargement and T2 hyperintensity in transected right brachial plexus elements.*





(Left) *Axial T1WI MR in a patient with traumatic avulsion injury depicts the classic MR appearance of a chronic nerve root avulsion with development of pseudomeningocele.* **(Right)** *Axial T2WI MR reveals an abnormal extradural fluid collection within the spinal canal, indicating an avulsion pseudomeningocele of T1. Note also marked enlargement of the C8 and T1 ventral primary rami following this severe brachial plexus stretch injury.*

Idiopathic Brachial Plexus Neuritis

KEY FACTS

TERMINOLOGY

- Parsonage-Turner syndrome

IMAGING

- Can affect any muscle innervated by brachial plexus
  - Most common: Rotator cuff, deltoid, biceps, triceps
- Denervation edema is earliest finding
  - Diffuse, homogeneous high signal on T2WI, STIR throughout affected muscle
- Fatty atrophy occurs in chronic denervation
  - Uncommonly seen
- Often muscles innervated by 2 or more different peripheral nerves are affected

TOP DIFFERENTIAL DIAGNOSES

- Cervical radiculopathy
- Suprascapular nerve entrapment
- Brachial plexus neoplasm
- Brachial plexus or cervical nerve root avulsion
- Radiation neuritis/myositis
- Quadrilateral space syndrome
- Pancoast tumor
- Muscle injury

PATHOLOGY

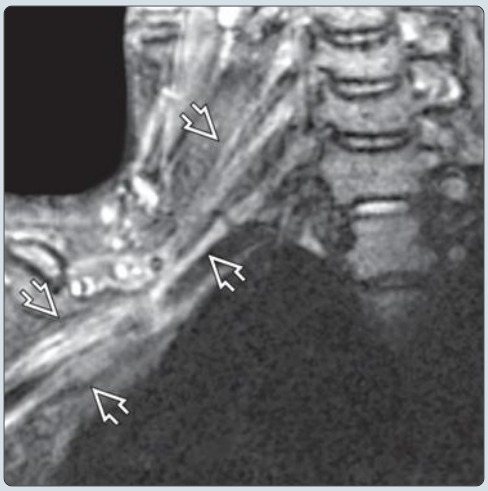
- Often associated with viral or bacterial infection
- Can also be post-traumatic or post surgery

CLINICAL ISSUES

- Sudden onset of pain, followed by weakness, paresthesias
- M > F
- Most cases resolve in 3 months to 2 years
- Physical therapy to preserve range of motion

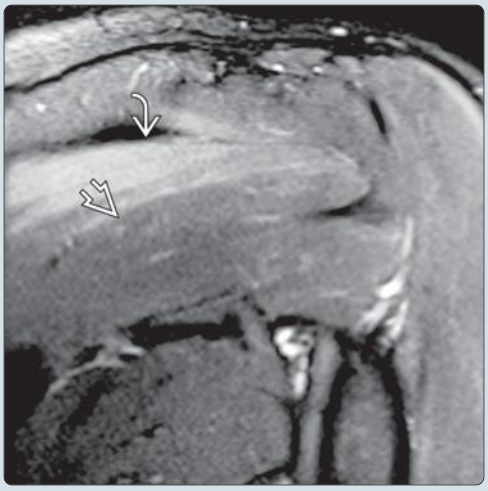
DIAGNOSTIC CHECKLIST

- Often an unexpected finding on shoulder MR performed to evaluate weakness, pain
- Abnormal muscle signal often involves more than 1 peripheral nerve distribution





(Left) *Coronal STIR MR shows diffusely increased signal intensity in brachial plexus due to idiopathic brachial neuritis.* **(Right)** *Coronal oblique T2WI MR shows denervation edema of the teres minor due to brachial neuritis. Edema is homogeneous and uniform, and there is no disruption of muscle fibers. Quadrilateral space shows no evidence of mass involving axillary nerve. Whenever denervation edema is seen, a search should be made for nerve mass or extrinsic compression.*





(Left) *Coronal oblique T2WI MR shows diffuse increase in signal intensity throughout superior fibers of infraspinatus muscle. Interestingly, the inferior portion of the muscle is spared. Unusual distributions of denervation edema are common in brachial neuritis.* **(Right)** *Sagittal T2WI MR shows diffusely abnormal signal intensity in the infraspinatus and supraspinatus muscles. Differential diagnosis, in this case, includes suprascapular nerve entrapment.*

TERMINOLOGY

Synonyms

- Parsonage-Turner syndrome
- Acute brachial neuritis
- Neuralgic amyotrophy

Definitions

- Immune-mediated neuropathy of brachial plexus

IMAGING

General Features

- Best diagnostic clue
  - Homogeneously increased signal on T2WI in 1 or more muscles of shoulder
- Location
  - Any muscle innervated by brachial plexus
    - Most common: Rotator cuff, deltoid, biceps, triceps
    - Uncommon: Brachialis, forearm muscles, diaphragm, serratus anterior (long thoracic nerve)
  - Sometimes bilateral
  - May cause pure sensory nerve deficit
- Size
  - Mild, uniform enlargement of affected muscles
  - Muscle atrophy seen in chronic cases
- Morphology
  - Diffuse involvement of muscle without focal mass

CT Findings

- Used to exclude mass involving brachial plexus or peripheral nerves
- Muscle atrophy, fatty infiltration evident in chronic cases

MR Findings

- **Brachial plexus**
  - Affected nerves high signal intensity on T2WI, STIR
  - Nerves show mild enhancement with gadolinium
  - No mass along course of nerves
- **Affected muscles**
  - Denervation edema is earliest finding
    - Muscle normal on T1WI
    - Diffuse, homogeneous high signal on T2WI, STIR throughout affected muscle
      - No disruption of muscle fibers
    - Diffuse, homogeneous enhancement with gadolinium
  - Fatty atrophy occurs in chronic denervation
    - Fatty streaking greater than in unaffected muscles
    - Decreased size of muscle
    - Uncommonly seen; most cases spontaneously resolve
  - Often muscles innervated by 2 or more different peripheral nerves are affected
    - e.g., infraspinatus (suprascapular nerve) + teres minor (axillary nerve)

Imaging Recommendations

- Best imaging tool
  - MR imaging
- Protocol advice
  - T1WI, T2WI, or STIR along long and short axes of symptomatic muscles
  - MR of shoulder should have 16-18 cm FOV on coronal images
    - Include spinoglenoid notch to rule out impingement on suprascapular nerve
    - Include quadrilateral space to rule out impingement on axillary nerve
  - Imaging of brachial plexus excludes brachial plexus mass
    - If patient gives characteristic history, not needed unless symptoms do not resolve

DIFFERENTIAL DIAGNOSIS

Cervical Radiculopathy

- Evaluate with cervical spine MR
- Most commonly see involvement of single nerve root
  - Brachial neuritis involves multiple nerve roots
- Severe cases may have denervation changes in affected muscles

Suprascapular Nerve Entrapment

- Causes denervation edema
- Mass in suprascapular notch: Affects supraspinatus + infraspinatus
- Mass in spinoglenoid notch: Affects infraspinatus

Brachial Plexus Neoplasm

- Presents with pain, muscle weakness
- Causes denervation edema
- Mass seen on MR of brachial plexus
- Usually metastasis; primary tumor uncommon

Brachial Plexus or Cervical Nerve Root Avulsion

- Severe, high velocity injury
- Loss of nerve continuity, fluid around nerve course seen on MR

Radiation Neuritis/Myositis

- Diffusely enlarged nerves and muscles showing increased T2 signal
- Geographic distribution
- History of radiation therapy

Quadrilateral Space Syndrome

- Mass between teres minor and teres major muscles
- Compresses axillary nerve, affects deltoid, teres minor muscles

Diabetic Neuropathy

- Usually distal, symmetric
- Sometimes involves 1 or more proximal muscles

Pancoast Tumor

- Bronchogenic carcinoma at lung apex ("superior sulcus")
- May directly extend to involve brachial plexus

Myositis

- Diffuse T2 signal abnormalities in muscle
- Edema in surrounding fat
- Usually bilaterally symmetric, multiple muscles

Muscle Injury

- High T2 signal intensity is heterogeneous, focal
- Disruption of muscle fibers usually visible
- Blood products often evident

- Surrounding soft tissues usually show edema/hematoma

Rotator Cuff Tear

- Chronic, large tears show muscle atrophy
- Fluid in tendon defect on MR

Burner/Stinger Syndrome

- Common athletic injury in contact sports
- Pain, paresthesias, muscle weakness
- Direct blow to brachial plexus in supraclavicular region
- Alternatively caused by traction to brachial plexus or cervical nerve root
- Usually transient, but may persist for weeks
- MR may be performed to exclude nerve avulsion

PATHOLOGY

General Features

- Etiology
  - Often associated with viral or bacterial infection
    - EBV, CMV, HIV among viral infections
  - Can occur post vaccination
  - Can also be post-traumatic or post surgery
    - Surgery may not be in same region
      - e.g., hip arthroplasty has been reported as cause

Gross Pathologic & Surgical Features

- Perineural edema, edema in affected muscles

Microscopic Features

- Perineural inflammatory infiltrate
- Thinning of myelin sheath
- Axonal degeneration

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Sudden onset of pain followed by weakness, paresthesias
  - May have sensory abnormalities only
  - Long thoracic nerve involvement: Winging of scapula
  - Phrenic nerve involvement: Shortness of breath

Demographics

- Age
  - May occur at any age
- Gender
  - M > F

Natural History & Prognosis

- Most cases resolve in 3 months to 2 years

Treatment

- Options, risks, complications
  - Physical therapy to preserve range of motion

DIAGNOSTIC CHECKLIST

Consider

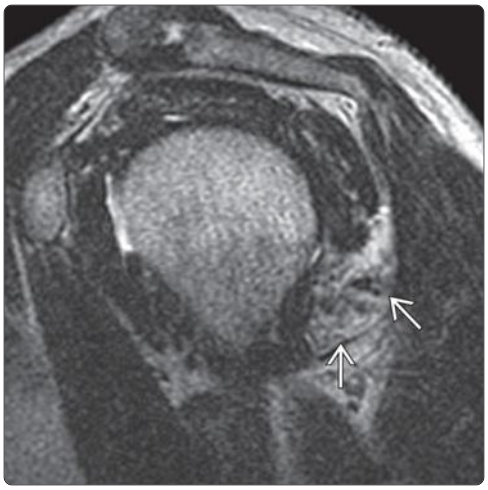
- Often an unexpected finding on shoulder MR performed to evaluate weakness, pain
- Brachial plexus MR performed to exclude other causes in atypical cases
  - Muscles outside of shoulder girdle
  - Respiratory compromise
  - Pure sensory loss

Image Interpretation Pearls

- Abnormal muscle signal often involves more than 1 peripheral nerve distribution
- Distribution of signal abnormalities helps to distinguish from nerve entrapment syndromes

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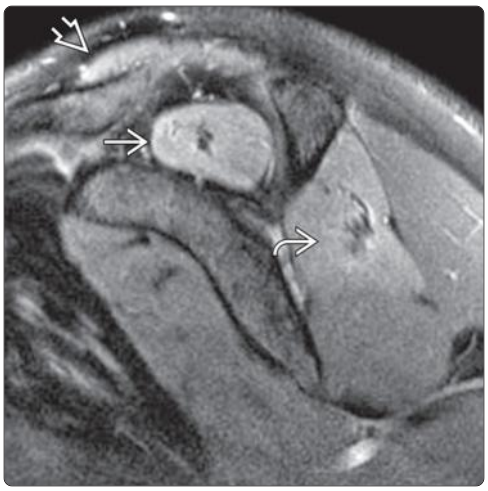


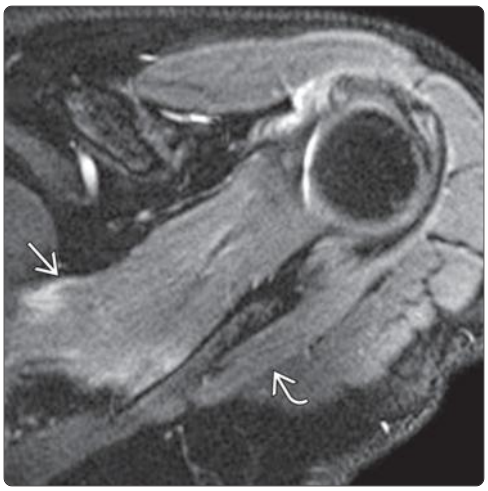
(Left) *Sagittal T2WI MR shows severe fatty atrophy of the teres minor muscle, manifested in volume loss and increased signal intensity. Most cases of idiopathic brachial plexus neuritis resolve without developing such severe atrophy.* **(Right)** *Coronal T1WI MR in the same patient again shows marked fatty atrophy limited to the teres minor. Quadrilateral space is normal, indicating that the atrophy is not due to compression of the axillary nerve.*





(Left) *Sagittal T2WI FS MR demonstrates denervation edema of the supraspinatus and infraspinatus muscles. This pattern may also occur from impingement of the suprascapular nerve at the suprascapular notch.* **(Right)** *Coronal T2WI FS MR in the same patient confirms that the edema spares a portion of the infraspinatus muscle. This pattern is not seen in nerve entrapment at the spinoglenoid or suprascapular notch, which affects all of the muscles innervated distal to the entrapment.*





(Left) *Sagittal T2WI FS MR demonstrates increased signal intensity representing a mild case of denervation edema involving the supraspinatus, infraspinatus, and trapezius muscles.* **(Right)** *Axial T2WI FS MR confirms hyperintense denervation edema of the supraspinatus and infraspinatus muscles.*

Traumatic Neuroma

KEY FACTS

TERMINOLOGY

- Amputation neuroma (stump neuroma)
  - Subtype of traumatic neuroma following traumatic or surgical amputation
- Morton neuroma: Traumatic neuroma between metatarsal heads

IMAGING

- Bulbous enlargement of nerve
- Enlargement of nerve fascicles
- Normal signal intensity on T1WI
- Intermediate to high signal intensity on T2WI, STIR
- Avid enhancement after gadolinium enhancement
- Any site of nerve injury or surgery

TOP DIFFERENTIAL DIAGNOSES

- Benign or malignant peripheral nerve tumor
- Perineural cyst
  - Eccentric to nerve, does not enhance with contrast
- Soft tissue metastasis or recurrent malignancy
- Neuritis

PATHOLOGY

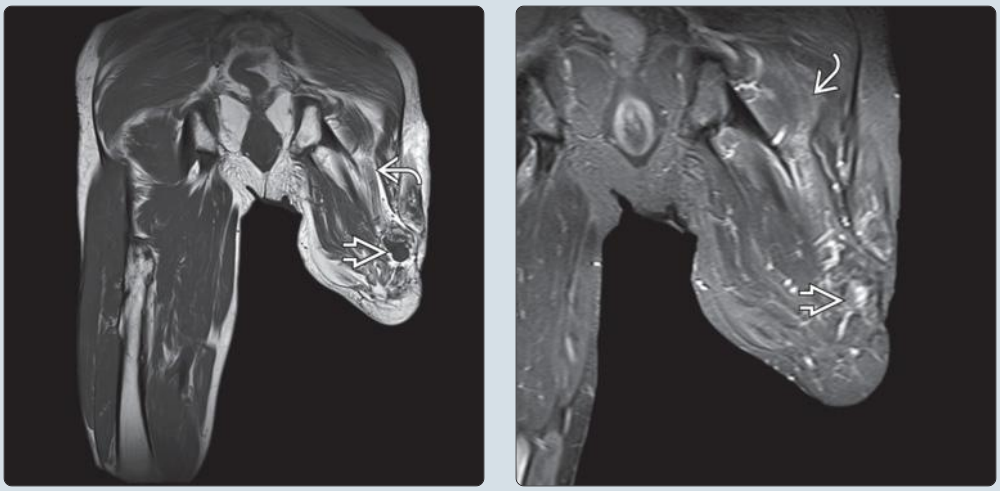
- Secondary to nerve injury: Amputation, crush injury, deep burns, minor trauma, nerve compression
- All elements of nerve present in neuroma in addition to scar tissue

CLINICAL ISSUES

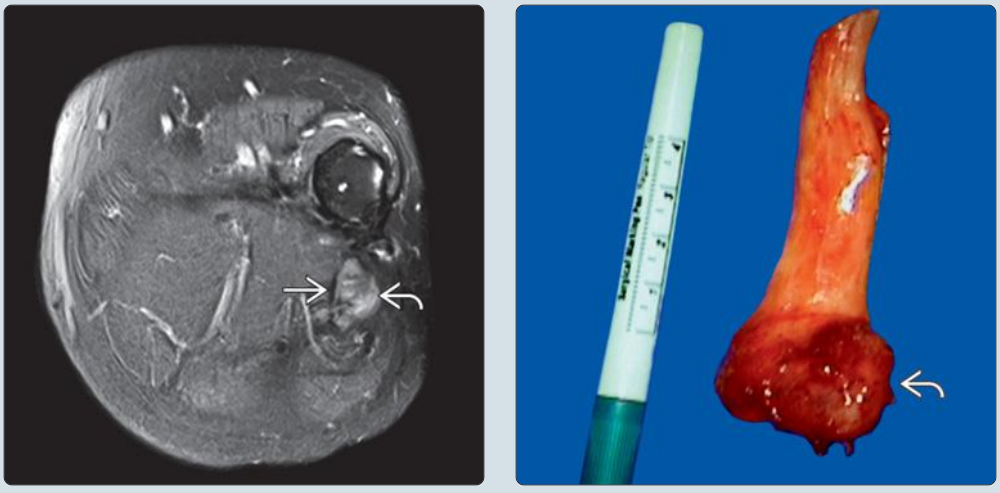
- Severe pain or paresthesias
- Arises 1-12 months after nerve injury
- As many as 1/4 of amputees develop stump neuroma

DIAGNOSTIC CHECKLIST

- May be mistaken for recurrent sarcoma



(Left) *Coronal T1WI MR in a patient status post above-knee amputation for trauma shows the sciatic nerve terminating in a large stump neuroma.* **(Right)** *Coronal STIR MR in the same patient nicely demonstrates the stump neuroma. The normal sciatic nerve is much less visible than on T1WI, as it blends with fat. T1WI are best for locating normal nerves, since the nerves' low signal intensity is conspicuous against the adjacent high signal intensity fat.*



(Left) *Axial T1WI C+ FS MR in the same patient shows marked enlargement of the abnormal sciatic nerve at the neuroma site. The separate tibial and peroneal nerves are distinctly seen.* **(Right)** *Surgical photograph of a resected stump neuroma illustrates bulbous enlargement of nerve stump tip, gradually tapering proximally into normal nerve.*

TERMINOLOGY

Synonyms

- Amputation neuroma (stump neuroma)
  - Subtype of traumatic neuroma following traumatic or surgical amputation
- Morton neuroma: Traumatic neuroma between metatarsal heads

Definitions

- Nonneoplastic nerve enlargement 2° to major or minor trauma

IMAGING

General Features

- Best diagnostic clue
  - Enlargement of transected nerve or nerve at site of soft tissue injury
- Location
  - Any site of nerve injury or surgery
    - Described in face, ear, neck, extremities, biliary tree, perineum
- Size
  - Usually 3-4x normal size

CT Findings

- Nerve enlargement
- Uniform enhancement with contrast

MR Findings

- **Transected nerve**
  - Bulbous enlargement of nerve most severe at amputation site
    - Nerve tapers proximally
  - Enlargement of nerve fascicles
    - Polka dot appearance on cross section
  - Normal signal intensity on T1WI
  - Intermediate to high signal intensity on T2WI, STIR
  - Avid enhancement after gadolinium enhancement
- **Lacerated nerve**
  - Irregular enlargement of nerve
  - Look for adjacent soft tissue injury
- **Compressed nerve**
  - Nerve enlargement mimics tumor
  - Process causing compression usually distinct from nerve

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - T1WI and T2WI along long and short axes of nerve
  - T1WI C+ in 2 planes to confirm nerve origin

DIFFERENTIAL DIAGNOSIS

Benign or Malignant Peripheral Nerve Tumor

- Signal characteristics, fusiform shape may be identical
- If no history of trauma, biopsy is needed
- If history of trauma, may monitor with serial exams

Perineural Cyst

- External to nerve
- No enhancement with gadolinium

Ganglion Cyst

- Associated with tendon
- No enhancement with gadolinium
- May compress nerve and cause neurologic symptoms

Soft Tissue Metastasis or Recurrent Malignancy

- Usually envelopes or displaces nerve
- Rarely contiguous with nerve

Mass Associated With Arthritis

- Gouty tophi or rheumatoid nodule mistaken for Morton neuroma
- May be presenting sign of arthritis

Neuritis

- Also shows enlargement of nerve fascicles
- Also enhances uniformly with gadolinium
- Nerve enlargement is more diffuse

PATHOLOGY

General Features

- Etiology
  - Secondary to nerve injury
    - Severed or crushed nerve
    - May occur with minor trauma
    - Morton neuroma: Tight shoes, high heels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Severe pain or paresthesias

Demographics

- Epidemiology
  - Arises 1-12 months after nerve injury
  - Affects as many as 1/4 of amputees

Treatment

- Steroid injection, physical therapy, resection

DIAGNOSTIC CHECKLIST

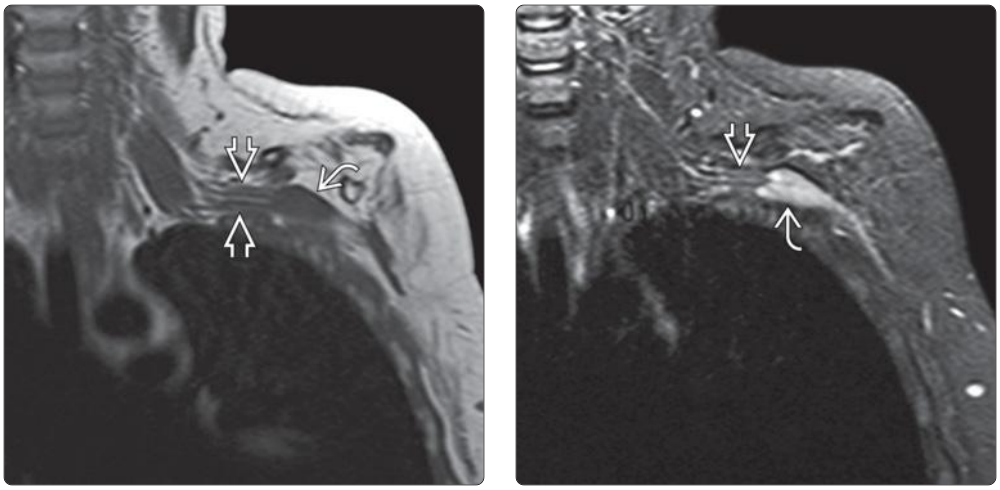
Consider

- May be mistaken for recurrent sarcoma

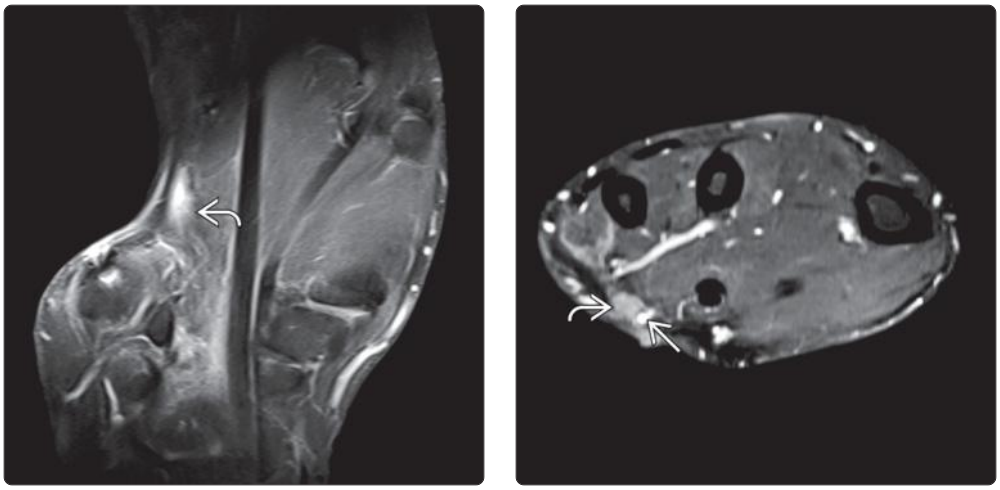
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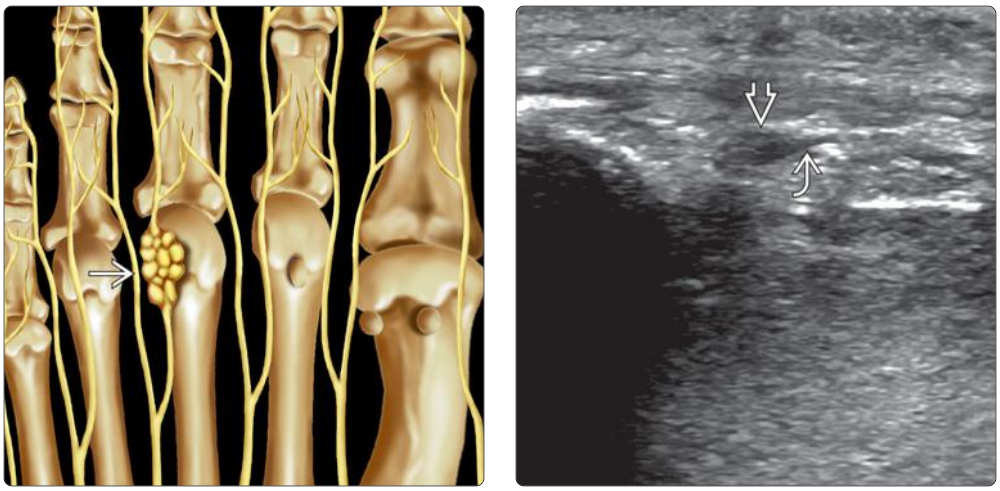
(Left) *Coronal T1WI MR in a patient status post forequarter amputation shows a bulbous mass contiguous with the brachial plexus. The shape and location are helpful in distinguishing traumatic neuroma from tumor recurrence.* **(Right)** *Coronal STIR MR in the same patient shows heterogeneously increased signal intensity of the traumatic neuroma compared to the proximal brachial plexus. Multiple small, rounded areas within the mass reflect enlarged nerve fascicles.*



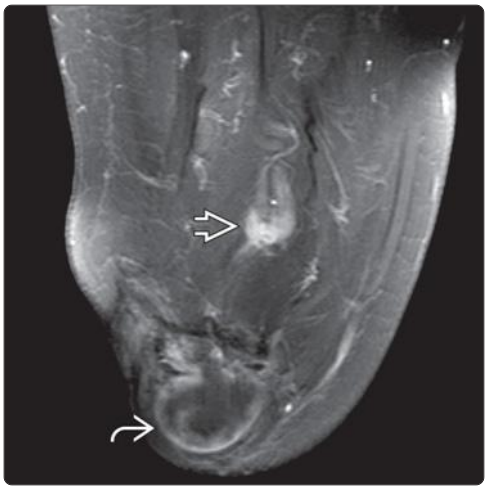
(Left) *Coronal T2WI FS MR shows a fusiform mass adjacent to amputation of the 4th and 5th digits for extremity sarcoma. The mass was felt to be suspicious for recurrence because it was not clearly associated with the nerve. Excision and pathologic evaluation confirmed an ulnar stump neuroma.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows avid neuroma enhancement. In retrospect, location adjacent to artery should have raised suspicion for a stump neuroma.*



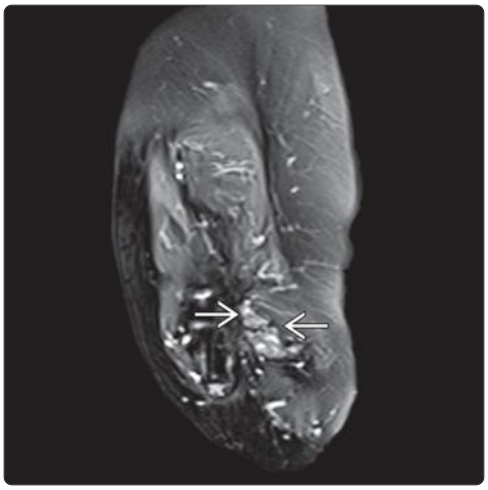
(Left) *Graphic shows a Morton neuroma along the plantar interdigital nerve. This traumatic neuroma is associated with tight shoes and high heels. Intermetatarsal bursitis and rheumatoid nodules are often diagnosed as Morton neuroma on MR and ultrasound.* **(Right)** *Longitudinal ultrasound shows a Morton neuroma located at the plantar aspect of the space between the metatarsal heads. The mass is hypoechoic and has a characteristic "tail" extending into the interdigital nerve.*

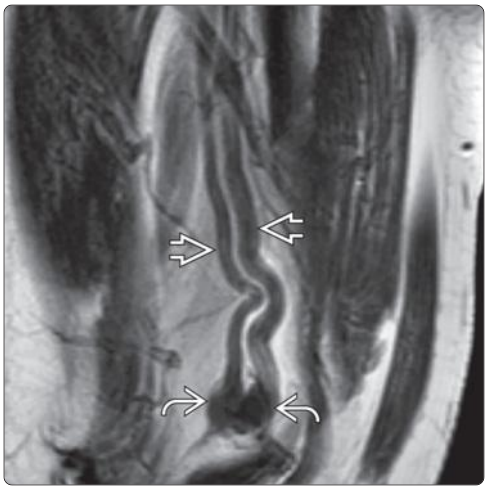






(Left) *Coronal T2WI FS MR through distal femur after above-knee amputation shows sciatic nerve enlargement and edema, with clear distinction of the peroneal and tibial components. A 2nd mass at the amputation site represents unrelated bursitis.* **(Right)** *Coronal T1WI C+ FS MR confirms avid neuroma enhancement. Bursitis, in comparison, demonstrates rim enhancement, and contrast is starting to diffuse into the center of the bursitis, an expected finding due to delay after contrast administration.*





(Left) *Sagittal STIR MR shows multiple round foci of high signal intensity near amputation site. This pattern reflects enlarged fascicles seen in cross section typical of nerve, and should prompt the radiologist to look on other planes to connect the lesion with the amputated nerve.* **(Right)** *Coronal T1WI MR of a stump is pathognomonic for stump neuroma, with bulbous mass clearly contiguous with a tortuous distorted sciatic nerve.*





(Left) *Coronal T1WI MR of an arm amputation demonstrates a neuroma continuous with the parent nerve. After arm amputation, neuromas are often seen at all 3 major nerves (median, ulnar, and radial).* **(Right)** *Coronal STIR MR in the same patient shows the mass is heterogeneous in signal intensity. Contiguity with the nerve enables the correct diagnosis. Heterogeneously high signal intensity mass adjacent to the neuroma is probably scar and can be monitored with sequential MR exams.*

Radiation Plexopathy

KEY FACTS

TERMINOLOGY

- Synonyms: Radiation-induced plexitis (RIP), radiation-induced fibrosis (RIF)

IMAGING

- Smooth, diffuse T2 hyperintensity ± enhancement of multiple plexus elements
- Upper brachial plexus (BP) (C5-C7) > lower BP (C8, T1) following radiation therapy (XRT) for breast and lung cancer, lymphoma
- Lumbosacral plexus following XRT for prostate, colorectal, and gynecologic tumors and lymphoma

TOP DIFFERENTIAL DIAGNOSES

- Malignant brachial plexus infiltration
- Plexiform neurofibroma
- Plexus traction injury

PATHOLOGY

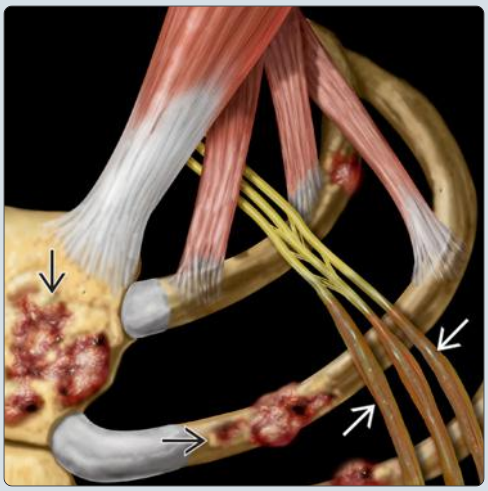
- Combination of direct cell damage 2° to ionizing radiation and progressive radiation-induced vascular damage to vaso nervosum
- Potentiated by chemotherapy

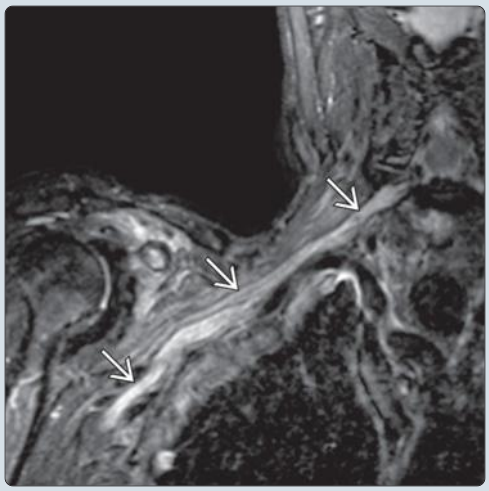
CLINICAL ISSUES

- Pain, paresthesia, motor deficits of ipsilateral extremity
- Prognosis variable; related to dose delivered to plexus, severity of nerve injury

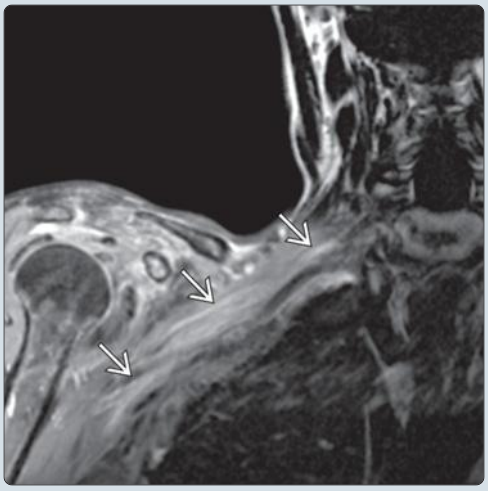
DIAGNOSTIC CHECKLIST

- Important to correctly diagnosis in order to avoid inappropriate additional radiation therapy
- Smooth T2 hyperintensity, linear enhancement of upper BP favors XRT plexitis > malignancy





(Left) *Coronal graphic of the left brachial plexus (BP) in breast cancer patient with skeletal metastases ⇨ following radiation therapy demonstrates diffuse swelling of irradiated brachial plexus elements ➡. Segmental involvement of all neural elements in radiation field is characteristic of radiation plexopathy.* **(Right)** *Coronal STIR MR (3 years post radiation for breast cancer with vague arm symptoms) shows diffuse smooth abnormal brachial plexus T2 hyperintensity ➡. No nodal mass was evident.*





(Left) *Coronal T1WI C+ FS MR (3 years post radiation for breast cancer with vague arm symptoms) reveals only mild patchy plexus enhancement ➡. No nodular mass lesion or enhancement was present, a finding that would imply neoplastic infiltration.* **(Right)** *Sagittal oblique STIR MR shows that abnormal T2 hyperintensity in upper plexus ventral primary rami ➡ preserves fascicular architecture. Plexus nodal tumor invasion more commonly involves the lower plexus because of axillary nodal location.*

TERMINOLOGY

Synonyms

- Radiation-induced plexitis (RIP), radiation-induced fibrosis (RIF)

Definitions

- Post-radiation brachial plexus (BP) or lumbosacral plexus (LSP) inflammation

IMAGING

General Features

- Best diagnostic clue
  - Smooth, diffuse T2 hyperintensity ± enhancement of multiple plexus elements
- Location
  - Upper BP (C5-7) > lower BP (C8, T1) following radiation therapy (XRT) for breast and lung cancer, lymphoma
  - Lumbosacral plexus following XRT for prostate, colorectal, and gynecologic tumors and lymphoma
- Size
  - Mild diffuse enlargement of abnormal nerves
- Morphology
  - Smooth, diffuse T2 hyperintensity of multiple trunks/divisions

CT Findings

- CECT
  - Difficult CT diagnosis
  - ± diffuse plexus enlargement, enhancement (acute)
  - ± distortion of tissue planes, ↑ density of axillary fat

MR Findings

- T1WI
  - Acute: Diffuse nerve enlargement, mild T1 hypointensity ± diffuse enhancement without focal nodularity, muscle denervation changes
  - Chronic: Nerves normal or architectural distortion ± surrounding tissue scarring, fibrosis, muscle denervation changes
- T2WI
  - Acute: Diffuse plexus enlargement, relatively homogeneous T2 hyperintensity, preservation of plexus internal architecture ± muscle denervation changes
  - Chronic: Nerves normal or architectural distortion ± surrounding tissue scarring, fibrosis, muscle denervation changes
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± mild homogeneous enhancement

Nuclear Medicine Findings

- PET
  - ± mild diffuse FDG uptake; avid focal uptake implies neoplastic metastasis

Other Modality Findings

- Electromyography: Subacute to chronic plexopathy with myokymic discharges in affected muscles

Imaging Recommendations

- Best imaging tool
  - High-resolution MR (MR neurography)
- Protocol advice
  - Coronal, sagittal oblique T1WI, STIR, fat-saturated T1WI C+ MR

DIFFERENTIAL DIAGNOSIS

Malignant Brachial Plexus Infiltration

- Direct extension from axillary adenopathy (breast cancer) or lung apex (Pancoast tumor) > systemic metastasis
- More focal, nodular than RIP
- Most commonly involves inferior BP (C8, T1), pain usually more severe than XRT plexopathy

Plexiform Neurofibroma

- More nodular, focal than RIP
- Clinical phenotype, genetic testing usually distinguish

Plexus Traction Injury

- Trauma history usually available
- Usually more focal than RIP, involves fewer plexus elements
- ± avulsion pseudomeningocele

PATHOLOGY

General Features

- Etiology
  - Combination of direct cell damage 2° to ionizing radiation → free radicals + progressive radiation-induced ischemia (vascular damage to vaso nervosum)
  - Uncommon < 60 Gy dose; dose-dependent, more common with large daily fractions or brachytherapy
  - Potentiated by chemotherapy
- Associated abnormalities
  - ± residual malignant tissue, fibrotic scar tissue
  - Unilateral left vocal cord paralysis (5%)
  - Radiation arteritis, accelerated atherosclerotic plaque formation
- Pathogenesis poorly understood
  - Connective tissue response spectrum from edema, acute inflammation → chronic inflammation, progressive fibrosis & neovascularization, and disruption of vascular supply to nerve
  - Acute edema may resolve if injury is minor & becomes subclinical, but frequently some level of nerve injury persists pathologically
  - Damaged neural tissue replaced by fibrosis in chronic plexopathies
- Incidence of complications ↑ with time following radiation

Staging, Grading, & Classification

- Modified LENT-SOMA scale (brachial plexopathy)
  - Grade 1: Mild sensory deficits, no pain
  - Grade 2: Moderate sensory deficits, tolerable pain, mild arm weakness
  - Grade 3: Continuous paresthesia with incomplete paresis
  - Grade 4: Complete paresis, excruciating pain, muscle atrophy

Gross Pathologic & Surgical Features

- Edema, demyelination, neural tissue atrophy, fibrous nerve sheath thickening, fibrous replacement of nerve fibers, connective tissue fibrosis

Microscopic Features

- Acute: Excessive extracellular matrix deposition, inflammatory infiltrate
- Chronic: Dense, noninflammatory fibrous matrix with fewer cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain, paresthesia, motor deficits of ipsilateral extremity
    - BP plexopathy usually referable to upper (C5-C7) > lower BP (C8-T1)
    - Lumbosacral plexopathy most commonly affects sciatic nerve distribution
    - Pain often relatively mild compared to malignant plexopathy but may be severe
  - Other signs/symptoms
    - Horner syndrome (rare; more common with malignant plexopathy)
- Clinical profile
  - Unilateral BP plexopathy following radiotherapy for breast cancer, Pancoast tumor, systemic metastasis
  - Bilateral brachial plexopathy after mantle radiotherapy for Hodgkin disease
  - Lumbosacral plexopathy following radiotherapy for prostate, cervical cancer

Demographics

- Age
  - Age at onset parallels demographics of initial cancer diagnosis + development time for radiation plexopathy
- Gender
  - M < F (reflects pertinent cancer demographics)
- Epidemiology
  - Rare (< 1% incidence)
  - Delayed onset 6 months to > 20 years post XRT
    - Peak onset at 10–20 months post XRT, median 1.5 years

Natural History & Prognosis

- Variable; related to dose delivered to plexus, severity of nerve injury
  - Some patients resolve spontaneously
  - Others slowly progress despite treatment
- Risk for brachial plexopathy may be lifelong
- All patients have potential to develop delayed symptoms, worsening of RIP grade

Treatment

- Avoid inappropriate additional radiation therapy
- Corticosteroids ± combined pentoxifylline and tocopherol (Vit E) therapy
- No traditional role for surgery; consider on case-by-case basis
  - Cervical dorsal root entry zone lesioning for treatment of neuropathic pain
  - Transferred myocutaneous flap + BP neurolysis may improve blood supply to fibrotic brachial plexus
- Palliative long-term narcotics, sympathectomy for pain relief

DIAGNOSTIC CHECKLIST

Consider

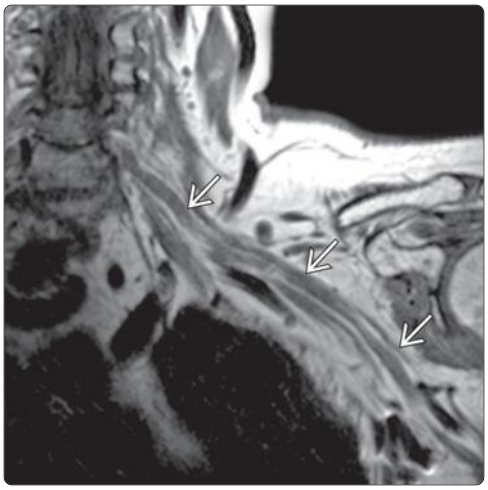
- Important to correctly diagnosis to avoid inappropriate additional radiation therapy

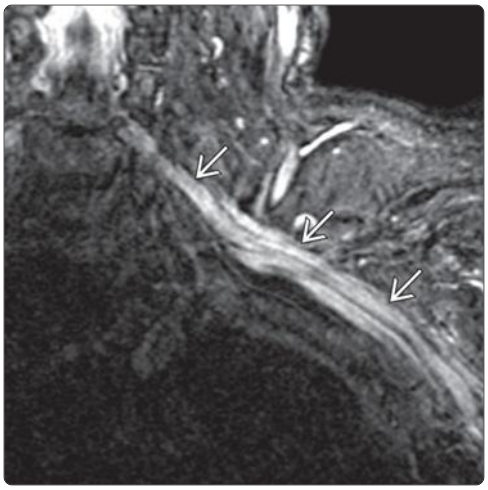
Image Interpretation Pearls

- Magnetic resonance plexus imaging limited by size of nerve
- May be difficult to distinguish XRT plexitis from recurrent tumor on imaging studies
- Smooth T2 hyperintensity, linear enhancement of upper BP favors XRT plexitis > malignancy

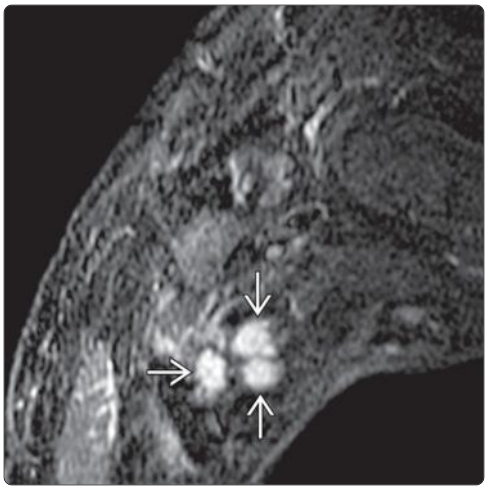
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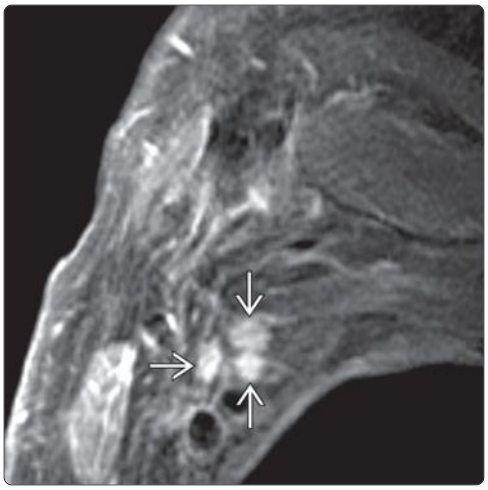
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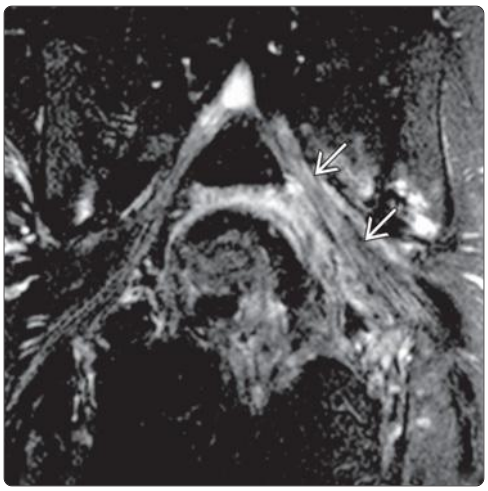


(Left) *Coronal T1WI MR (breast cancer post XRT with vague symptoms following radiation therapy) demonstrates mild diffuse enlargement of the upper brachial plexus. T1WI MR is the least sensitive sequence for detecting post-radiation plexus injury.* **(Right)** *Coronal STIR MR (breast cancer post XRT with vague symptoms following radiation therapy) shows mild diffuse brachial plexus T2 hyperintensity without focal nodular mass, suggesting local nodal tumor extension.*





(Left) *Sagittal oblique STIR MR (post-radiation therapy for breast cancer, mild symptoms) depicts diffuse enlargement, fascicular swelling, and abnormal T2 hyperintensity of all 3 BP cords. The characteristic fascicular pattern in short axis confirms neural origin.* **(Right)** *Sagittal oblique T1WI C+ MR (post-radiation therapy for breast cancer, mild symptoms) demonstrates diffuse enlargement and mild enhancement of the abnormal BP cords.*





(Left) *Coronal STIR MR (post-radiation therapy for cervical cancer, chronic symptoms) shows mild left lumbosacral plexus T2 hyperintensity surrounded by ill-defined hyperintense fibrovascular scar tissue. No dominant mass to suggest neural metastases is evident.* **(Right)** *Coronal T1 C+ MR (following radiation therapy for cervical cancer, chronic symptoms) shows mild enhancement within the fibrovascular scar tissue engulfing the irradiated lumbosacral plexus.*

Peripheral Nerve Sheath Tumor

KEY FACTS

TERMINOLOGY

- Grouping of benign and malignant primary tumors of peripheral nerves

IMAGING

- Mass associated with a peripheral nerve
  - Schwannoma: Round mass, eccentric to nerve
  - Neurofibroma: Fusiform, cord-like, or lobulated mass centered on nerve
  - Plexiform neurofibroma: Arborizing tangle of lobular masses
  - Malignant peripheral nerve sheath tumor (MPNST): Round, ovoid, fusiform, or lobulated
- Low signal intensity on T1WI, PDWI
- Intermediate to high signal intensity on T2WI, STIR
- Avid enhancement with gadolinium
- Muscles innervated by nerve often show denervation changes

TOP DIFFERENTIAL DIAGNOSES

- Traumatic neuroma
- Hemangioma
- Neuritis
- Other soft tissue sarcomas
- Metastases, soft tissue
- Lymphoma

PATHOLOGY

- Malignant degeneration seen in neurofibroma, but rare in schwannoma

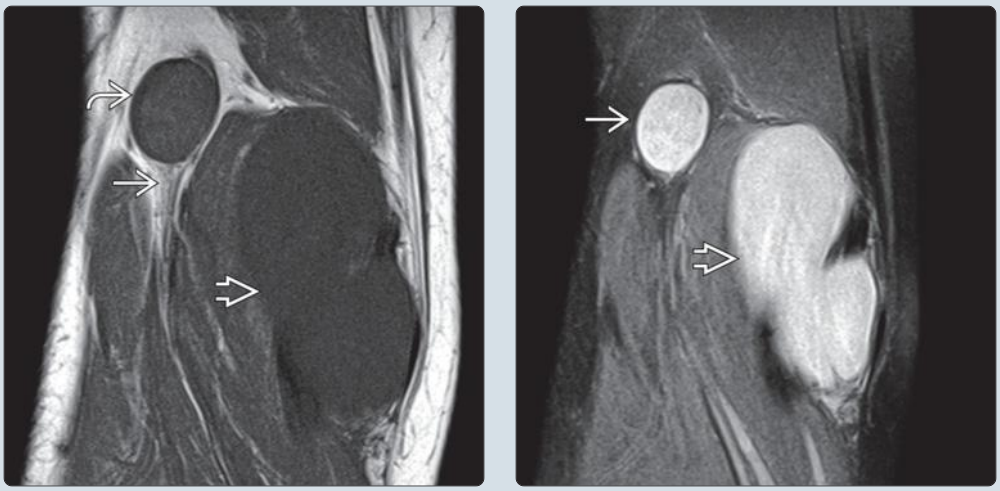
CLINICAL ISSUES

- Palpable mass, often painful
- May cause weakness in distribution of affected nerve

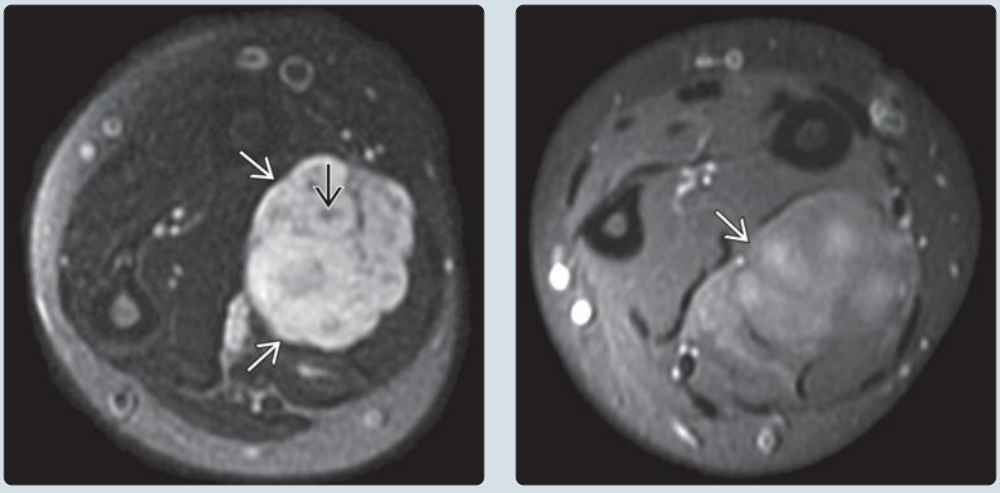
DIAGNOSTIC CHECKLIST

- Imaging cannot reliably distinguish between benign and malignant nerve sheath tumors

(Left) *Coronal T1WI MR demonstrates a large mass arising from the peroneal nerve. The mass has the same signal intensity as the adjacent popliteal cyst.* **(Right)** *Coronal PD FSE FS MR in the same patient confirms that both the tumor and fluid have a similar degree of minimal signal heterogeneity. It can be challenging to discern these tumors from cysts even with MR.*



(Left) *Axial T2 FS MR reveals marked T2 hyperintensity characteristic of neural sheath tumor in a large median nerve mass. Note the characteristic "target" appearance of plexiform neurofibroma.* **(Right)** *Axial T1 C+ FS MR confirms typical heterogeneous, relatively understated enhancement (compared to schwannoma) within the large median nerve plexiform neurofibroma.*



TERMINOLOGY

Definitions

- Grouping of benign and malignant primary tumors of peripheral nerves
- Malignant peripheral nerve sheath tumor (MPNST) replaces earlier terms (e.g., malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma)

IMAGING

General Features

- Best diagnostic clue
  - Mass along course of peripheral nerve
- Morphology
  - Schwannoma: Round mass, eccentric to nerve
  - Neurofibroma: Fusiform, cord-like, or lobulated mass centered on nerve
  - Plexiform neurofibroma: Arborizing tangle of lobular masses
  - MPNST: Round, ovoid, fusiform, or lobulated

CT Findings

- Fusiform, lobulated, or ovoid mass along course of nerve
- Benign nerve sheath tumors often erode adjacent bone
  - Erosions are corticated, reflecting slow enlargement of tumor
- Malignant nerve sheath tumors may erode or invade bone

MR Findings

- Mass associated with peripheral nerve
  - Usually sharply demarcated
  - Margins may be obscured by peritumoral edema
  - Cannot use margins to distinguish between benign and malignant
- Low signal intensity on T1WI, PDWI
  - These sequences best show relationship to adjacent nerve
  - Trace tumor into nerve to confirm origin from nerve
- Intermediate to high signal intensity on T2WI, STIR
  - May be homogeneous or heterogeneous
- Avid enhancement with gadolinium
  - Central necrosis common in MPNST
  - Cystic degeneration often seen in schwannoma
  - Pattern of enhancement is not a reliable sign of tumor aggressivity
- Muscles innervated by nerve often show denervation changes
  - Uniform high signal intensity on T2WI, STIR
  - Uniform enhancement with gadolinium

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - T1WI or PDWI best for showing relationship to adjacent nerve
  - Gadolinium enhancement often useful to distinguish from cyst
    - Not useful in distinguishing benign from malignant

Ultrasonographic Findings

- May see uniform echotexture or mixed solid/fluid tumor

DIFFERENTIAL DIAGNOSIS

Traumatic Neuroma

- History of trauma or amputation
- Bulbous enlargement of nerve at site of trauma
- Diffuse enhancement with gadolinium

Hemangioma

- Enlarged vessels may mimic multilobulated neurofibroma
- Draining veins visible

Neuritis

- Nerve mildly enlarged without focal mass
- Long segment of nerve enhances with gadolinium
- Increased T2 signal and enhancement in muscles innervated by affected nerve

Other Soft Tissue Sarcomas

- Mass not contiguous with nerve on MR, CT

Metastases, Soft Tissue

- Metastases may involve nerve or displace nerve

Lymphoma

- Neurolymphoma may primarily involve peripheral nerves
- Non-Hodgkin lymphoma

PATHOLOGY

General Features

- Associated abnormalities
  - 5-13% of patients with neurofibromatosis develop MPNST
  - Malignant degeneration seen in neurofibroma, but rare in schwannoma

Staging, Grading, & Classification

- Benign tumors
  - Schwannoma (neurilemoma)
  - Neurofibroma
  - Plexiform neurofibroma: Arborizing tumor of peripheral nerves occurring in neurofibromatosis
  - Nerve sheath myxoma (neurothekeoma)
  - Perineurioma
- Malignant tumors
  - Malignant peripheral nerve sheath tumor

Gross Pathologic & Surgical Features

- Schwannoma: Encapsulated; solid areas, edematous areas, cysts
- Neurofibroma: Not encapsulated, glistening, tan-white
- Nerve sheath myxoma: Gelatinous
- Perineurioma: Circumscribed white mass
- MPNST: Usually large, white-tan, hemorrhage, and necrosis

Microscopic Features

- Schwannoma: 2 types of cells, Antoni A (spindle cells) and Antoni B (tumor cells surrounded by fluid)
- Neurofibroma: All elements of peripheral nerve proliferate
- Nerve sheath myxoma: Myxoid material, cells arranged in bundles

- Perineurioma: Elongated cells in parallel bundles; storiform growth
- MPNST: Serpentine shape of tumor cells, arranged in whorls or palisades
- Variety of gene mutations have been described

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Palpable mass
    - Often painful
    - Positive Tinel sign
      - Pain along distal course of nerve with percussion of mass
  - Other signs/symptoms
    - May cause weakness in distribution of affected nerve

Demographics

- Age
  - Localized neurofibroma: Usually ages 20-30 years
  - Nerve sheath myxoma, perineuroma: Childhood to early adulthood
  - Schwannoma and MPNST: Ages 20-50 years

Natural History & Prognosis

- MPNST has poor prognosis
  - Adjacent nerve is at risk for local spread or skip metastases
    - Must distinguish from traumatic neuroma due to surgery
  - Frequent tumor surveillance is needed postoperatively
  - Lung metastases are common

Treatment

- Options, risks, complications
  - Most are amenable to surgical excision
    - "Jump graft" may be performed to bridge gap in nerve
  - MPNST treated with radiation and chemotherapy in addition to excision

DIAGNOSTIC CHECKLIST

Consider

- Percutaneous biopsy tends to be very painful
  - Use intravenous medication for pain management during biopsy

Image Interpretation Pearls

- Peripheral nerve lesions are often mistaken for cysts on MR
  - Gadolinium contrast should be administered for all soft tissue masses
  - Any mass not purely cystic should be biopsied
- Often can see tapered transition to normal contiguous nerve at margin of mass
  - Helps to distinguish nerve sheath tumor from mass compressing nerve

Reporting Tips

- Imaging cannot reliably distinguish between benign and malignant nerve sheath tumors
  - Report should use nonspecific terms, such as "consistent with tumor of nerve sheath origin"
- "Target" sign strongly suggestive of plexiform neurofibroma

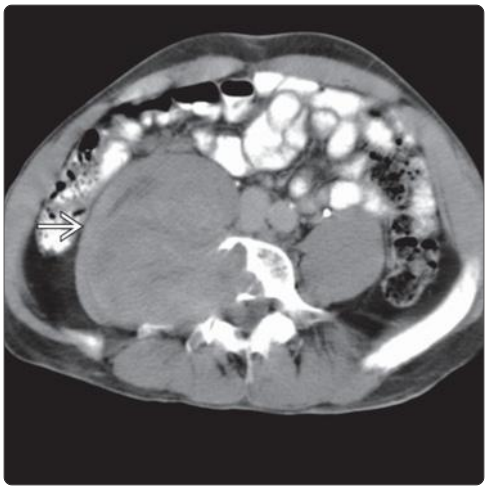
SELECTED REFERENCES

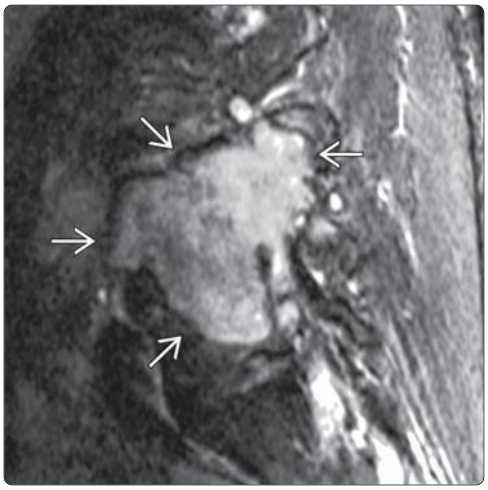
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(Left) *Coronal T1WI FS MR shows a heterogeneous mass ➡ arising in the left femoral nerve. Preoperative and frozen section diagnoses were schwannoma, but final histology showed malignant peripheral nerve sheath tumor.* **(Right)** *Coronal T1WI C+ FS MR in the same patient shows that the mass ➡ has a heterogeneous enhancement pattern, which can be seen in both benign and malignant lesions.*





(Left) *Axial CECT shows neurofibroma ➡ centered in right L4 neural foramen, extending into the abdomen, and eroding bone. Patient had vague pain for 14 years before undergoing radiography, which showed the mass.* **(Right)** *Sagittal T2WI MR (same patient) through lumbar vertebral body and pedicle shows heterogeneous signal intensity in the neurofibroma ➡. There is unusually extensive bone erosion of 2 adjacent spinal levels.*





(Left) *Coronal T1WI MR shows a schwannoma ➡ arising from the brachial plexus. Differential diagnosis for this isolated lesion should include metastasis as well as a primary neural tumor.* **(Right)** *Coronal STIR MR in the same patient shows that the lesion ➡ is markedly heterogeneous. Signal intensity and margins are not helpful in distinguishing benign from malignant nerve sheath tumors.*

KEY FACTS

TERMINOLOGY

- Synonyms: Peripheral neurolymphomatosis (NL), perineural lymphomatosis
- Perineural plexus or peripheral nerve lymphomatous infiltration

IMAGING

- Diffusely infiltrating plexus or peripheral nerve lesion in patient with lymphoma
  - Abnormal nerve T2 hyperintensity
  - Disruption/distortion of normal neural fascicular morphology
  - Variable enhancement

TOP DIFFERENTIAL DIAGNOSES

- Peripheral nerve metastasis
- CIDP
- Hypertrophic neuropathy
- Nerve/plexus stretch injury

PATHOLOGY

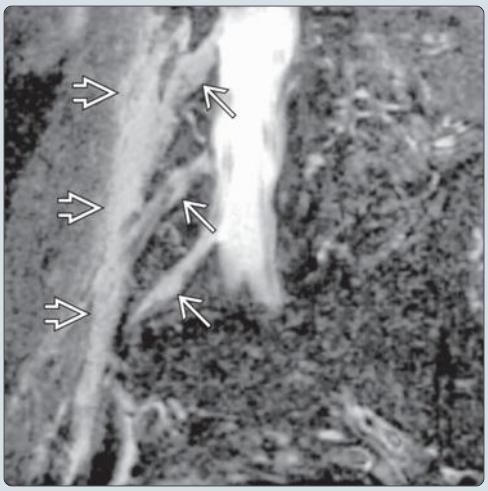
- Perineural lymphomatous nerve infiltration produces thickened neural elements

CLINICAL ISSUES

- Progressive sensorimotor neuropathy ± plexopathy
- Relentlessly progressive; variable response to chemotherapy, radiotherapy

DIAGNOSTIC CHECKLIST

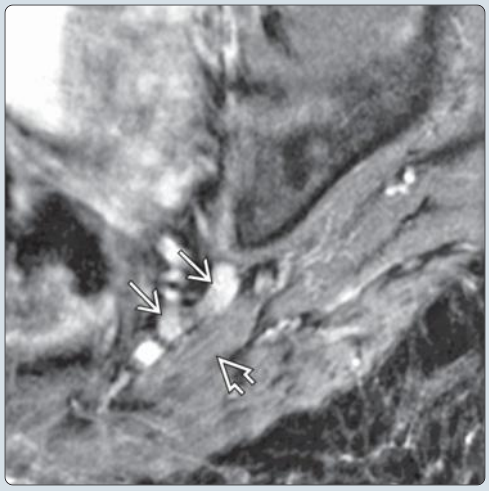
- Careful evaluation of NL patients often reveals evidence for subclinical systemic lymphoma
- Imaging findings are often subtle
  - Carefully scrutinize clinically abnormal nerves and plexi for thickening, abnormal T2 signal
  - High index of suspicion important for recognition





(Left) *Coronal STIR MR (stage IV thoracic lymphoma, progressive leg atrophy and pain) demonstrates abnormal L2-L4 nerve root and dorsal root ganglia ➡ hyperintensity and enlargement. Also present is concomitant abnormal thickening and T2 hyperintensity of the right lumbar plexus ➡.* **(Right)** *Axial T1WI C+ MR (stage IV thoracic lymphoma, progressive leg atrophy and pain) reveals abnormal thickening and avid enhancement of the infiltrated right lumbar plexus ➡.*





(Left) *Axial T1WI C+ FS MR (non-Hodgkin lymphoma, systemic metastases with progressive arm pain and weakness) demonstrates an ill-defined, enhancing infiltrative mass ➡ centered within the left radial nerve neurovascular bundle near the humeral spiral groove.* **(Right)** *Sagittal oblique T1WI C+ MR with fat saturation (known lymphoma, progressive left sciatic neuropathy) demonstrates abnormal left sciatic nerve enlargement and avid enhancement ➡ at the piriformis muscle ➡ level.*

TERMINOLOGY

Synonyms

- Peripheral neurolymphomatosis (NL), perineural lymphomatosis

Definitions

- Perineural plexus or peripheral nerve lymphomatous infiltration

IMAGING

General Features

- Best diagnostic clue
  - Diffusely infiltrating plexus or peripheral nerve lesion in patient with lymphoma
- Location
  - Plexus or peripheral nerve
- Morphology
  - Abnormal plexus/nerve thickening with disruption/distortion of normal fascicular morphology

CT Findings

- CECT
  - Often normal; abnormal cases show fusiform or nodular thickening + variable enhancement

MR Findings

- T1WI
  - Hypo- to isointense nerve/plexus thickening
- T2WI
  - Abnormal nerve T2 hyperintensity, disruption/distortion of normal fascicular morphology
- STIR
  - Abnormal nerve T2 hyperintensity, disruption/distortion of normal fascicular morphology
- T1WI C+
  - Variable enhancement

Imaging Recommendations

- Protocol advice
  - Multiplanar high-resolution MR (T1WI, fat-saturated T2WI or STIR, fat-saturated T1WI C+)

DIFFERENTIAL DIAGNOSIS

Peripheral Nerve Metastasis

- Usually more focal; signal intensity mimics primary nerve tumor

CIDP

- Fusiform nerve/plexus thickening
- Often bilateral, symmetric

Hypertrophic Neuropathy

- Fusiform nerve/plexus thickening
- Often bilateral, symmetric

Nerve/Plexus Stretch Injury

- Most commonly brachial plexus; often asymmetric

PATHOLOGY

General Features

- Etiology
  - Isolated disease entity or in association with systemic or primary CNS lymphoma
- Associated abnormalities
  - Careful evaluation of NL patients usually reveals subclinical systemic lymphoma

Gross Pathologic & Surgical Features

- Perineural lymphomatous nerve infiltration → thickened neural elements

Microscopic Features

- Perineural lymphomatosis infiltration ("small blue cell tumor")

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain ± motor weakness, sensory deficit
- Clinical profile
  - Progressive sensorimotor neuropathy ± plexopathy
  - Laboratory abnormalities
    - Electrodiagnostic studies: Plexopathy/neuropathy ± superimposed axonal peripheral neuropathy
    - CSF studies: ± elevated protein, malignant cells

Demographics

- Age
  - Adults
- Gender
  - M = F
- Epidemiology
  - Autopsy study showed NL ~ 40% of lymphoma patients

Natural History & Prognosis

- Relentlessly progressive; ± response to chemotherapy, radiotherapy

Treatment

- Biopsy to establish diagnosis (if findings of systemic lymphoma lacking)
- Chemotherapy, external beam radiation

DIAGNOSTIC CHECKLIST

Consider

- Careful evaluation of NL patients often reveals evidence for subclinical systemic lymphoma

Image Interpretation Pearls

- Often subtle; carefully scrutinize clinically abnormal nerves and plexi for thickening, abnormal T2 signal

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Hypertrophic Neuropathy

KEY FACTS

TERMINOLOGY

- Clinically and genetically heterogeneous group of inherited disorders characterized by focal or diffuse peripheral nerve enlargement

IMAGING

- Focal or diffuse fusiform peripheral nerve enlargement
  - Peripheral nerves ± intradural nerve roots
  - ± involvement of cauda equina nerve roots
- Acute &/or chronic muscle denervation changes
- Best imaging tool: High-resolution MR imaging (MR neurography)

TOP DIFFERENTIAL DIAGNOSES

- Guillain-Barré syndrome
- Chronic inflammatory demyelinating polyneuropathy (CIDP)
- Nerve sheath tumor
- Traumatic nerve stretch injury

PATHOLOGY

- Hypertrophic nerve roots, peripheral nerves
- Histologic features of demyelination, remyelination + "onion bulb" formations (HMSN I)

CLINICAL ISSUES

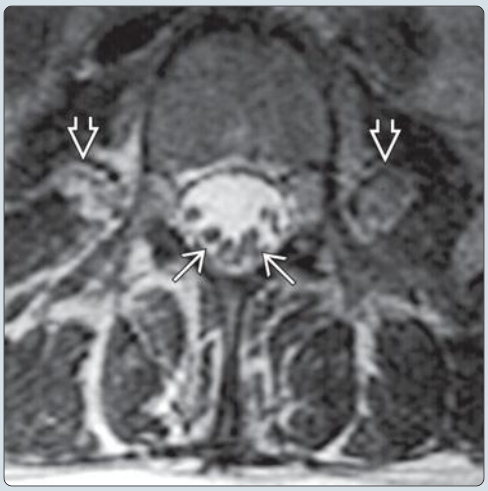
- Distal extremity muscle weakness/atrophy (motor > sensory), foot deformities
- Back/lower extremity radicular pain ± myelopathy
- Sensory loss, focal tenderness, dysesthesias
- ± palpable nerve mass

DIAGNOSTIC CHECKLIST

- Consider HMSN when abnormally enlarged peripheral nerves identified on MR imaging
- Genetic consultation recommended for patients with fusiform nerve enlargement on MR imaging

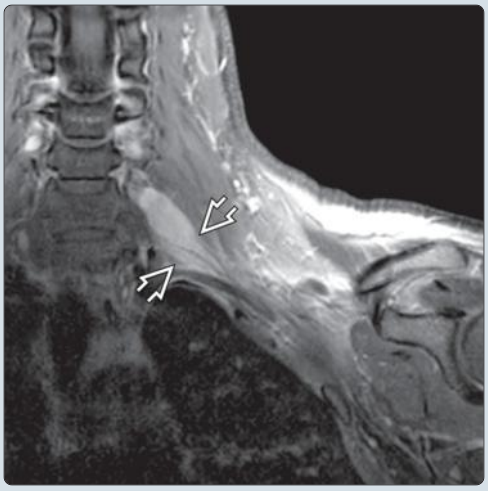
(Left) *Coronal T1WI MR (Charcot-Marie-Tooth, painful scoliosis) demonstrates convex left scoliosis and bilateral abnormal fusiform enlargement of the extradural spinal nerve roots and lumbar plexus.* **(Right)** *Axial T2WI MR (Charcot-Marie-Tooth, painful scoliosis) confirms abnormal enlargement and T2 hyperintensity of the bilateral extradural lumbar nerve roots as well as aberrant enlargement of the intradural cauda equina nerve roots.*





(Left) *Coronal STIR MR (unknown type hypertrophic neuropathy, left arm pain and weakness) demonstrates abnormal fusiform enlargement of the left proximal brachial plexus C7 and C8 roots/rami.* **(Right)** *Coronal T1WI C+ FS MR (unknown type hypertrophic neuropathy, left arm pain and weakness) of the left brachial plexus confirms fusiform enlargement and diffuse homogeneous nerve enhancement within the left proximal brachial plexus C7 and C8 roots/rami.*





TERMINOLOGY

Synonyms

- Hereditary motor-sensory neuropathy (HMSN), hypertrophic neuropathy (HN), hereditary sensory neuropathy (HSN), hereditary motor neuropathy (HMN), hereditary sensory and autonomic neuropathy (HSAN), Charcot-Marie-Tooth (CMT) syndrome

Definitions

- Clinically and genetically heterogeneous group of inherited disorders characterized by focal or diffuse peripheral nerve enlargement
- Subdivided into 3 main categories based on predominant involvement of motor &/or sensory nerves
  - Both motor and sensory nerves affected → HMSN
    - HMSN I (Charcot-Marie-Tooth syndrome type I, CMT 1)
    - HMSN II (neuronal-type peroneal muscular atrophy, CMT 2)
    - HMSN III (Dejerine-Sottas disease [DSD], hypertrophic neuropathy of infancy, congenital hypomyelinating neuropathy)
  - Motor nerve fibers primarily → distal HMN, spinal CMT, distal spinal muscular atrophy
  - Mainly peripheral sensory ± autonomic nerves → hereditary sensory neuropathy (HSN) or hereditary sensory and autonomic neuropathy (HSAN)

IMAGING

General Features

- Best diagnostic clue
  - Focal or diffuse peripheral nerve enlargement + distal extremity atrophy
- Location
  - Peripheral nerves ± intradural nerve roots

CT Findings

- CECT
  - Enlarged nerves ± enhancement

MR Findings

- T1WI
  - Fusiform hypointense mass(es) of peripheral nerves ± cauda equina nerve roots
  - ± abnormal muscle T1 hyperintensity, volume loss (chronic denervation → fatty atrophy)
- T2WI
  - Fusiform hyperintense mass(es) of peripheral nerves ± cauda equina nerve roots
  - Internal fascicular architecture disrupted over abnormal segment
  - ± abnormal muscle hyperintensity, swelling on fat-saturated T2WI (acute denervation)
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± abnormal enhancement of enlarged nerves (most commonly HMSN I, III)

Nonvascular Interventions

- Myelography
  - ± nerve root thickening, myelographic block 2° to spinal stenosis

Imaging Recommendations

- Best imaging tool
  - High-resolution MR imaging (MR neurography)
- Protocol advice
  - Axial T1WI, fat-saturated T2WI or STIR, fat-saturated T1WI C+ MR

DIFFERENTIAL DIAGNOSIS

Guillain-Barré Syndrome

- Acute onset of ascending paralysis with relative sensory preservation
- Symptom onset, progression characteristic
- Pial, nerve root enhancement similar to CIDP

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Repeated episodes of demyelination, remyelination → "onion skin" spinal, peripheral nerve enlargement
- Mimics HMSN, plexiform neurofibroma on MR
- Inflammatory infiltrate (histology) helps distinguish from HMSN

Nerve Sheath Tumor

- Schwannoma, solitary neurofibroma, plexiform neurofibroma
  - Diffuse nerve root enlargement, enhancement
  - ± cutaneous stigmata of NF1 on physical exam
  - Genetic testing, distinctive clinical stigmata distinguish from CIDP, HMSN

Traumatic Nerve Stretch Injury

- Nerve enlargement, architectural distortion
- History, trauma imaging findings make diagnosis

PATHOLOGY

General Features

- Etiology
  - HMSN I (CMT 1): Disorder of peripheral myelination
    - Mutation in peripheral myelin protein-22 (*PMP-22*) gene → unstable myelin that spontaneously demyelinates
    - Schwann cells proliferate and remyelinate axons
    - Repeated cycles of demyelination, remyelination produce concentric myelin layers around axons ("onion bulb" formations)
  - HMSN II (CMT 2): Primary axonal loss, wallerian degeneration without remyelination/demyelination cycles
  - HMSN III (Dejerine-Sottas disease): Severe segmental demyelination, myelin thinning around nerve
- Genetics
  - HMSN IA (CMT 1A): Autosomal dominant, short arm of chromosome 17
  - HMSN IB (CMT 1B): Long arm of chromosome 1
  - HMSN II: Chromosome 1
  - HMSN III: Autosomal recessive (controversial), chromosome 17

Staging, Grading, & Classification

- HMSN types IA, IB (CMT 1A, 1B)
- HMSN type II (neuronal-type peroneal muscular atrophy)
- HMSN type III (hypertrophic neuropathy of infancy, Dejerine-Sottas disease)
- HMSN type IV (hypertrophic neuropathy associated with phytanic acid excess)
- HMSN type V (associated with spastic paraplegia)
- HMSN type VI (associated with optic atrophy)
- HMSN type VII (associated with retinitis pigmentosa)

Gross Pathologic & Surgical Features

- Hypertrophic thickened nerve roots, peripheral nerves
- Roots may fill thecal sac at surgery

Microscopic Features

- Histologic features of demyelination, remyelination
  - HMSN I: Primary hypertrophic myelin pathology → demyelination/remyelination, S100(+) concentric "onion bulb" formations, secondary axonal changes
  - HMSN II: Primary axonal pathology, axon loss and wallerian degeneration
  - HMSN III: Severe hypomyelination/demyelination, axonal loss, myelin sheath thinning

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - HMSN I
    - Distal extremity muscle weakness/atrophy (motor > sensory), foot deformities (pes cavus, hammer toes), ↓ tendon reflexes, ± palpable extremity nerve masses
    - Relative lack of pain (in contrast to acquired neuropathies)
    - Similarly affected relatives common
  - HMSN II: Often minimally symptomatic
  - HMSN III: Clinical symptoms similar but more severe than HMSN I, with earlier presentation
    - Delayed motor development, progressive muscular weakness of legs and arms
- Other signs/symptoms
  - ± palpable nerve mass
  - Back/lower extremity radicular pain ± myelopathy
  - Sensory loss, focal tenderness, dysesthesias
  - Cold feet, hair loss, leg edema
- Clinical profile
  - HMSN type I: Distal extremity weakness, muscle atrophy (legs > arms)
    - Hyporeflexia or areflexia, orthopedic foot deformities, scoliosis (37-50%), tremor (≤ 25%)
  - HMSN type II: Peripheral nerves not clinically enlarged, lower extremity > hand weakness, sensory loss in distal extremities, foot deformities (< HMSN I)
  - HMSN type III: Infant with hyporeflexia, palpable enlarged peripheral nerves, sensory loss ± ataxia

Demographics

- Age
  - HMSN I: Onset in 1st decade typical, delayed in some patients until early or mid adulthood
  - HMSN II: Often diagnosed later in life; symptoms commonly begin during 2nd decade
  - HMSN III: Onset typically during infancy or early childhood
- Gender
  - M = F
- Epidemiology
  - HMSN I: Incidence 15/100,000
    - CMT IA incidence 10.5/100,000 (70% of HMSN I cases)
  - HMSN II: Incidence 7/100,000
  - HMSN III: Rare

Natural History & Prognosis

- Normal life expectancy
- Slowly progressive condition; disability severity highly variable

Treatment

- Conservative: Injury prevention (lifestyle adjustment, avoidance of occupational injury), treatment of acquired neuropathies, physical therapy, ankle-foot orthoses, anti-inflammatory medications
- Surgical: Correction of orthopedic deformities

DIAGNOSTIC CHECKLIST

Consider

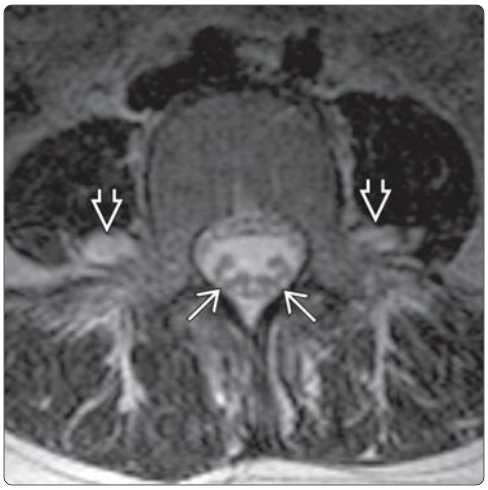
- Genetic consultation for patients with fusiform nerve enlargement identified on MR imaging

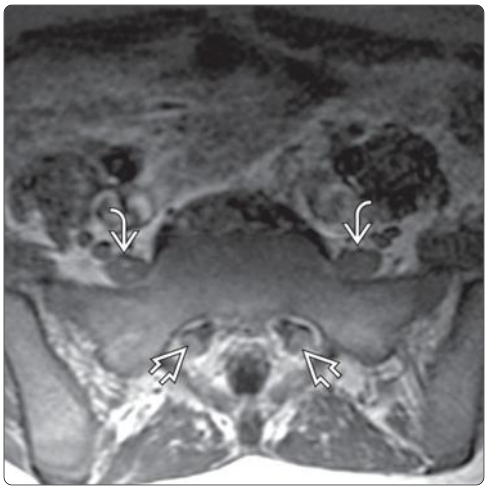
Image Interpretation Pearls

- Consider HMSN when abnormally enlarged peripheral nerves or nerve roots are identified on MR imaging

SELECTED REFERENCES

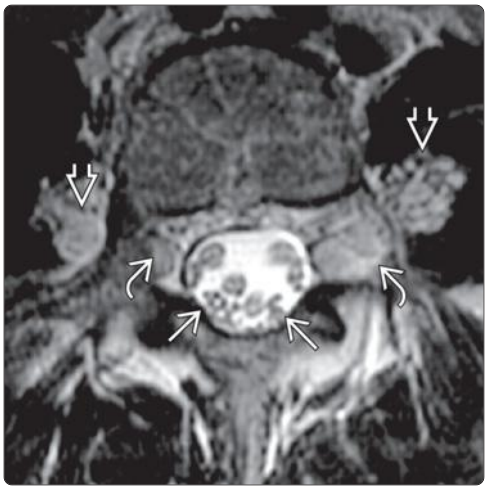
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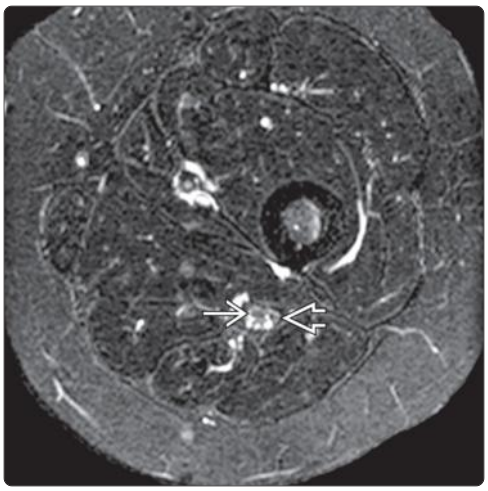


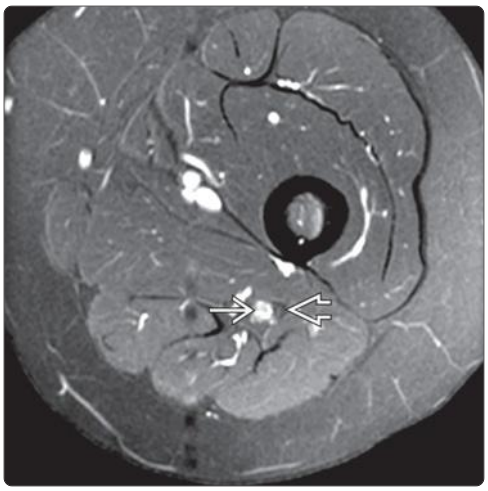
(Left) *Axial T2WI MR (Charcot-Marie-Tooth, CMT 1) demonstrates mild diffuse enlargement of the intradural cauda equina nerve roots as well as bilateral extradural nerve enlargement in the lumbar plexus medial to the psoas muscles.* **(Right)** *Axial T1WI C+ MR (Charcot-Marie-Tooth, CMT 1) at the sacral level confirms mild enhancement and abnormal enlargement of the bilateral extradural sacral nerves and lumbosacral trunk nerves.*





(Left) *Sagittal T1WI MR (Charcot-Marie-Tooth) through the lumbar neural foramina demonstrates abnormal enlargement of the extradural spinal nerves in the neural foramina.* **(Right)** *Axial T2WI MR (Charcot-Marie-Tooth) confirms abnormal enlargement of the cauda equina nerve roots as well as abnormal enlargement and T2 hyperintensity of the foraminal and paraspinal extradural nerves.*





(Left) *Axial STIR MR (unknown type HMSN, S1 radiculopathy) demonstrates a central hypointense tibial nerve mass displacing several mildly swollen, abnormally hyperintense fascicles to the periphery. The peroneal division is normal.* **(Right)** *Axial T1WI C+ FS MR (unknown type hypertrophic neuropathy, S1 radiculopathy) reveals modest abnormal enhancement of the tibial division neural mass. There is no abnormal enhancement of the adjacent normal peroneal division.*

Femoral Neuropathy

KEY FACTS

TERMINOLOGY

- Synonyms: Femoral neuropathy, femoral mononeuropathy, femoral nerve (FN) palsy
- FN entrapment or injury 2° to direct trauma, compression, stretch injury, or ischemia

IMAGING

- Nerve enlargement ± loss of internal fascicular architecture, ↑ T2 hyperintensity
- Injury most common in psoas muscle body, iliopsoas groove, or femoral canal

TOP DIFFERENTIAL DIAGNOSES

- Neoplastic femoral nerve infiltration
- Nerve sheath tumor
- Lumbosacral disc syndromes
- Lumbar plexopathy

PATHOLOGY

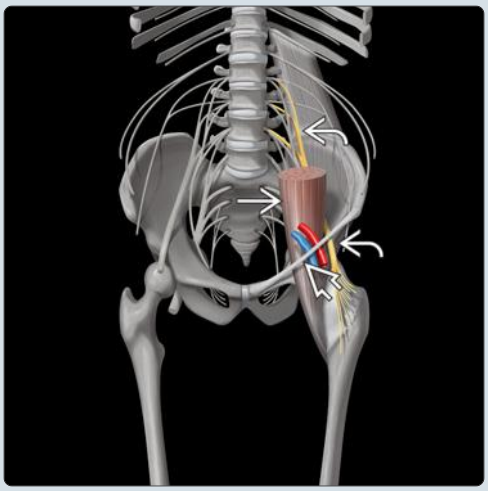
- Reported causes include self-retaining retractor, thigh tourniquet, traumatic injury, heparin anticoagulation (retroperitoneal hematoma), arterial catheterization complication, obstetrical complication, diabetic neuropathy

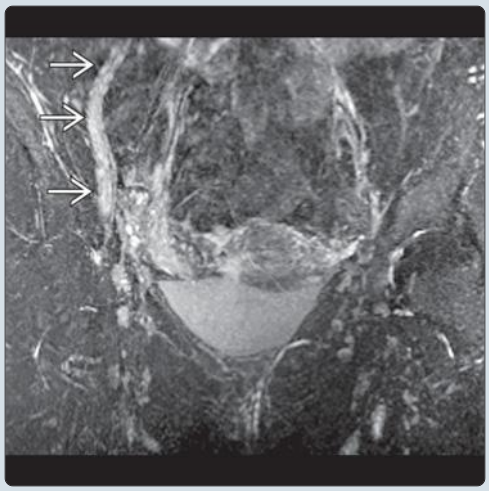
CLINICAL ISSUES

- Acute symptom onset, pain/weakness in FN distribution, diminished/absent knee jerk reflex, thigh muscle atrophy
- Severe back/groin pain (retroperitoneal hematoma)
- Recovery is rule over days → months

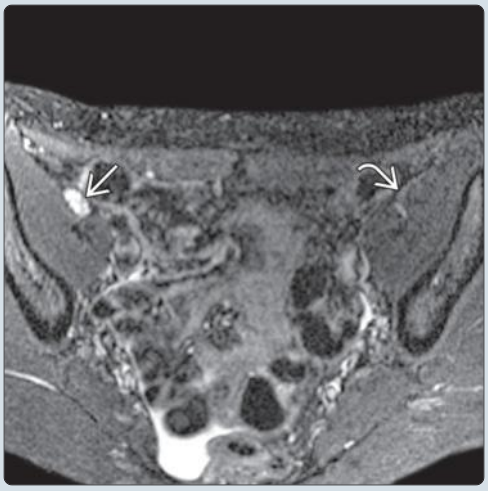
DIAGNOSTIC CHECKLIST

- Femoral neuropathy is uncommon
- Look carefully for lesion or hematoma in iliopsoas groove or femoral canal





(Left) *Coronal graphic shows the normal course of the femoral nerve relative to the psoas muscle and inguinal ligament. The femoral nerve produces multiple peripheral branches to the anterior thigh muscles.* **(Right)** *Coronal STIR MR (femoral neuropathy postsurgical herniorrhaphy) depicts marked enlargement, T2 hyperintensity of the right femoral nerve, with abrupt transition at the right groin. In this case, the femoral nerve was accidentally ligated during herniorrhaphy.*





(Left) *Axial STIR MR (femoral neuropathy postsurgical herniorrhaphy) through the pelvis confirms that the right femoral nerve in the iliopsoas groove is markedly enlarged with discrete T2 hyperintense fascicles (compared to the normal left femoral nerve).* **(Right)** *Axial CECT (severe hemophilia) depicts large left iliacus and psoas hematomas. Femoral neuropathy occurs from compression of the adjacent femoral nerve, which runs along the psoas muscle and iliopsoas groove.*

TERMINOLOGY

Synonyms

- Femoral mononeuropathy, femoral nerve (FN) palsy

Definitions

- FN entrapment or injury 2° to direct trauma, compression, stretch injury, or ischemia

IMAGING

General Features

- Best diagnostic clue
  - Femoral nerve enlargement, abnormal T2 hyperintensity
- Location
  - Injury most common at psoas muscle body, iliopsoas groove, or femoral canal

CT Findings

- CECT
  - ± retroperitoneal/psoas mass, hematoma, lymphadenopathy

MR Findings

- T1WI
  - Isointense → hypointense nerve enlargement ± retroperitoneal/psoas mass
  - ± hyperintense quadriceps muscle (fatty atrophy 2° to chronic denervation)
- T2WI
  - Hyperintense nerve (± fascicular) enlargement
  - ± abnormal quadriceps muscle T2 hyperintensity (acute/ongoing denervation)
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± enhancement (if neoplastic/inflammatory)

Other Modality Findings

- Electromyography (EMG): Denervation of anterior thigh muscles

Imaging Recommendations

- Best imaging tool
  - High-resolution MR imaging
- Protocol advice
  - Axial T1WI, fat-saturated T2WI or STIR MR, fat-saturated T1 C+ MR

DIFFERENTIAL DIAGNOSIS

Neoplastic Femoral Nerve Infiltration

- Neurolymphomatosis
- Systemic metastasis
- Local regional tumor extension

Nerve Sheath Tumor

- Solitary/plexiform neurofibroma (NF1), schwannoma
- Neural mass + variable tumoral enhancement

Lumbosacral Disc Syndromes

- Disc herniation compresses L2, L3, or L4 root
- EMG → radiculopathy, not mononeuropathy

Lumbar Plexopathy

- Diabetic, ischemic, traumatic, neoplastic etiologies
- EMG → plexopathy

PATHOLOGY

General Features

- Etiology
  - Reported causes include self-retaining retractor, thigh tourniquet, traumatic injury, heparin anticoagulation (retroperitoneal hematoma), arterial catheterization complication, obstetrical complication, diabetic neuropathy

Gross Pathologic & Surgical Features

- FN arises from L2–L4 ventral rami, descends in iliopsoas groove, exits pelvis below inguinal ligament
- Innervates iliacus, sartorius, and quadriceps muscles

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute symptom onset, pain/weakness in FN distribution, diminished/absent knee jerk reflex, thigh muscle atrophy
- Other signs/symptoms
  - Severe back/groin pain (retroperitoneal hematoma)

Demographics

- Age
  - All ages
- Gender
  - M = F
- Epidemiology
  - Uncommon

Natural History & Prognosis

- Recovery is rule over few days → months in most cases

Treatment

- Physical therapy, avoidance of excessive hip abduction/external rotation, knee bracing, treatment of causative mass lesions

DIAGNOSTIC CHECKLIST

Consider

- Femoral neuropathy is uncommon

Image Interpretation Pearls

- Look carefully for lesion or hematoma in iliopsoas groove or femoral canal

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Ulnar Neuropathy

KEY FACTS

TERMINOLOGY

- Synonyms: Ulnar nerve (UN) entrapment, cubital tunnel syndrome
- Partial fixation, compression, or distortion of UN

IMAGING

- UN enlargement ± abnormal T2 hyperintensity, architectural distortion
- Most commonly occurs within cubital tunnel (elbow); uncommon at Guyon tunnel (wrist) or brachial plexus

TOP DIFFERENTIAL DIAGNOSES

- Acute direct nerve trauma
- Idiopathic brachial plexitis (Parsonage-Turner syndrome)
- Nerve sheath tumor
- Enlarged perineural vein
- Medial epicondylitis

PATHOLOGY

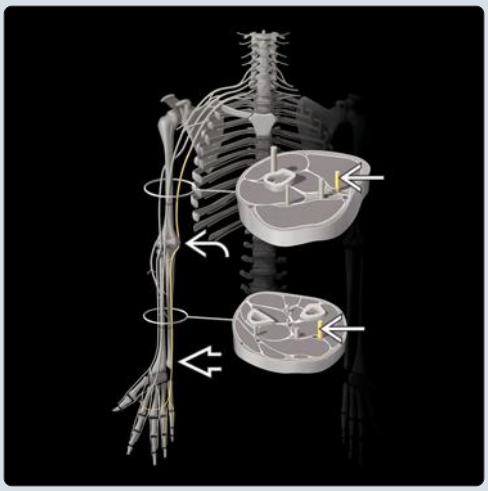
- Edematous/indurated UN ± thickened retinaculum, fibrous infiltration, fascicular atrophy/loss
- ± anconeus epitrochlearis, enlarged medial triceps head

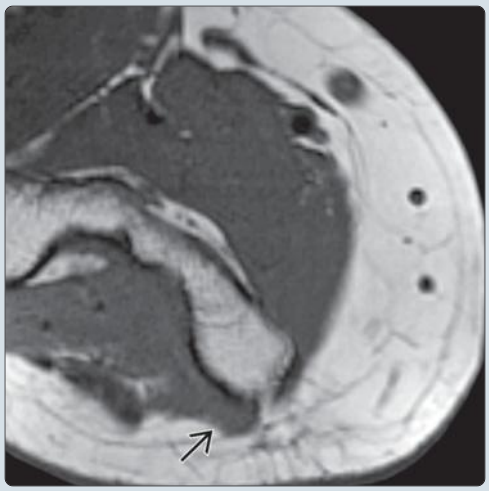
CLINICAL ISSUES

- Symptoms range from mild transient paresthesias of 4th and 5th digits → claw hand/digits, intrinsic muscle atrophy
- ± severe elbow/wrist pain radiating proximally or distally

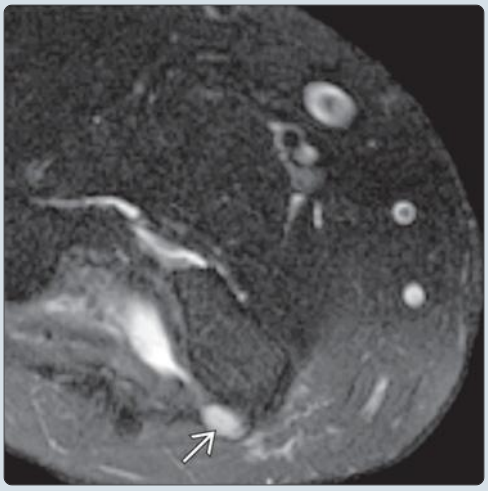
DIAGNOSTIC CHECKLIST

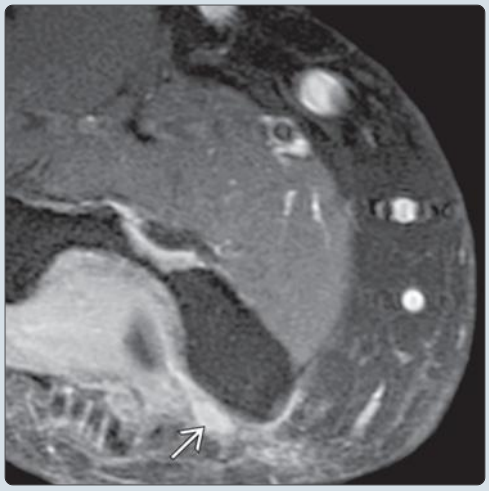
- Focal nerve enlargement, abnormal T2 hyperintensity distal to medial epicondyle
- Search for anatomic abnormalities (enlarged triceps medial head or anconeus muscle, osteophyte, thickened retinaculum)





(Left) *Coronal graphic of the right upper extremity demonstrates the normal course of the ulnar nerve (yellow). Vulnerable locations to injury include the cubital tunnel at the medial elbow and Guyon canal at the medial wrist.* **(Right)** *Axial T1WI MR of the right elbow demonstrates mild abnormal enlargement and rounded configuration of the right ulnar nerve within the cubital tunnel.*





(Left) *Axial T2WI FS MR of the right elbow demonstrates mild enlargement and abnormal T2 hyperintensity of the right ulnar nerve with complete loss of internal fascicular architecture, indicating an axonotmetic or neurotmetic injury rather than the less severe neuropraxic injury.* **(Right)** *Axial T1WI C+ FS MR of the right elbow confirms diffuse abnormal enhancement of the mildly enlarged right ulnar nerve. Note also mild edema and inflammation in the adjacent soft tissues.*

TERMINOLOGY

Synonyms

- Ulnar nerve entrapment, cubital tunnel syndrome

Definitions

- Partial fixation, compression, or distortion of ulnar nerve (UN)
  - Most commonly occurs within cubital tunnel at elbow; uncommon within Guyon tunnel at wrist, or 2° to brachial plexus inflammation

IMAGING

General Features

- Best diagnostic clue
  - UN enlargement ± T2 hyperintensity at cubital or Guyon tunnel in symptomatic patient
- Location
  - Most common at elbow near medial epicondyle (cubital tunnel); less common at ulnar wrist (Guyon tunnel)
- Size
  - Usually enlarged 1.5-2x, with change from mildly flattened configuration → rounded
- Morphology
  - Nerve swelling ± enlarged hyperintense fascicles (neuropraxic injury), axonal injury with loss of internal architecture (axonotmesis), or complete anatomic/functional transection (neurotmesis)

CT Findings

- NECT
  - Acute: ± soft tissue nerve mass, anconeus epitrochlearis
  - Chronic: Fatty atrophy of affected muscles (volume loss, hypodense fat infiltration)
- CECT
  - ± UN enhancement

MR Findings

- T1WI
  - UN thickening ± architectural distortion
  - Thickened cubital tunnel retinaculum
- T2WI
  - UN enlargement, abnormal T2 hyperintensity
  - Loss of internal UN architecture in more severe cases
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± UN enhancement (focal fascicles or diffusely)

Ultrasonographic Findings

- Grayscale ultrasound
  - Enlarged UN ± loss of reticular fascicular pattern (round hypoechoic areas surrounded by hyperechoic lines)

Other Modality Findings

- Electromyography: Helps localize most likely level of nerve injury

Imaging Recommendations

- Best imaging tool
  - High-resolution MR peripheral nerve imaging (MR neurography)
- Protocol advice
  - Axial T1WI, fat-saturated T2WI, and STIR imaging using surface coil

DIFFERENTIAL DIAGNOSIS

Acute Direct Nerve Trauma

- Appropriate history, traumatic soft tissue findings

Idiopathic Brachial Plexitis (Parsonage-Turner Syndrome)

- Brachial plexus inflammation, multiple nerve/muscle denervation patterns
- ± viral prodrome: Pain → weakness

Nerve Sheath Tumor

- Focal nerve enlargement, displacement/obliteration of internal fascicle architecture
- Tumoral enhancement, T2/STIR hyperintensity

Enlarged Perineural Vein

- Mimics enlarged UN
- Flow void, contrast enhancement similar to other veins

Medial Epicondylitis

- "Thrower's elbow," "golfer's elbow"
- Chronic valgus stress → ± medial collateral ligament (MCL) tear, hyperintensity within flexor pronatus muscle belly

PATHOLOGY

General Features

- Etiology
  - Intermuscular septum (arcade of Struthers) → compression 2° to musculofascial band 5-8 cm proximal to medial epicondyle
  - Thickened cubital tunnel retinaculum → entrapment with dynamic elbow flexion
  - Congenitally shallow groove, absent/dysfunctional retinaculum
  - Repeated valgus stress → medial epicondylitis (throwing athletes)
  - Malunion, callus around condylar fracture
  - Fascial compression at flexor carpi ulnaris (FCU) heads
- Associated abnormalities
  - Anconeus epitrochlearis muscle
  - Enlarged medial triceps head
  - ± medial elbow osteophyte, condylar osseous abnormality
- Important normal anatomy
  - UN lies in anterior proximal arm compartment along intermuscular septum
  - Enters posterior compartment at mid humerus, piercing arcade of Struthers
  - Passes distally through posterior/medial intermuscular septum → cubital tunnel
  - Distal to cubital tunnel, UN passes between 2 heads of FCU muscle (most frequent compression site)
- Cause of abnormal high signal on T2WI/STIR sequences not definitively known

- Speculated that axoplasmic flow alterations or edema from increased endoneurial fluid 2° to disordered endoneurial fluid flow, local venous obstruction → T2 hyperintensity

Staging, Grading, & Classification

- 3 severities of nerve injury
  - Neuropraxic injury (mildest)
    - Focal myelin sheath damage without axonal disruption
    - Contiguous swollen/T2 hyperintense nerve fascicles
  - Axonotmetic injury (intermediate)
    - Axon/myelin sheath disruption; subsequent wallerian degeneration leaves Schwann cells and endoneurium intact
    - Homogeneously increased T2 signal intensity, loss of fascicular architecture at injury site
  - Neurotmetic injury (most severe)
    - Functional ± anatomical transection
    - Axon/myelin sheath disruption with discontinuity of some/all of surrounding connective tissues → wallerian degeneration, regeneration failure
    - Cannot always distinguish from axonotmesis

Gross Pathologic & Surgical Features

- Edematous/indurated UN ± thickened retinaculum, fibrous stranding
- ± anconeus epitrochlearis, enlarged medial triceps head

Microscopic Features

- Nerve edema ± inflammatory cells, fibrous infiltration, fascicular atrophy/loss

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Range of symptoms: Mild transient paresthesias of 4th and 5th digits → claw hand/digits, intrinsic muscle atrophy
  - ± severe elbow/wrist pain radiating proximally or distally
- Clinical profile
  - Pain, paresthesias in ulnar nerve distribution
  - Throwing athletes with elbow pain of medial epicondylitis + paresthesias

Demographics

- Age
  - Adults > children
- Gender
  - M ≤ F
- Epidemiology
  - 2nd most common upper extremity entrapment syndrome (after carpal tunnel syndrome)
    - Up to 22% of USA population may have chronic compression during flexion 2° to thickened retinaculum
    - 10% static compression in association with anconeus epitrochlearis muscle

Natural History & Prognosis

- Rest, avoidance of provocative activities may produce remission in mild cases
- More severe cases show variable response to surgical decompression
  - Chronic pain, muscle weakness, atrophy portend worse prognosis

Treatment

- Conservative: Physical therapy, avoidance of provocative maneuvers
- Surgical: Osteophyte or anconeus resection, decompression ± nerve transposition
  - In situ decompression brings rapid relief, but transposition may bring longer symptom remission

DIAGNOSTIC CHECKLIST

Consider

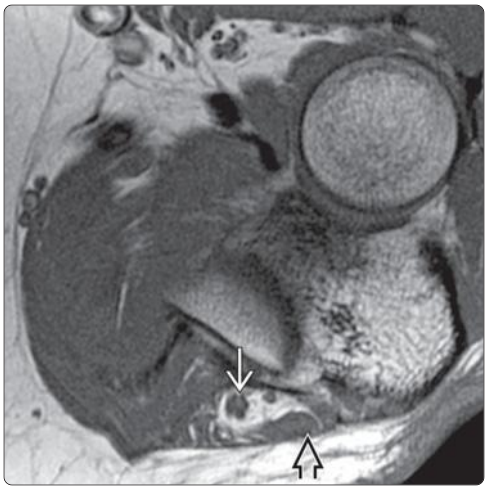
- MR imaging more sensitive for acute than chronic neuropraxia
- Search for anatomic abnormalities (enlarged medial head of triceps, anconeus muscle, osteophyte, thickened retinaculum)

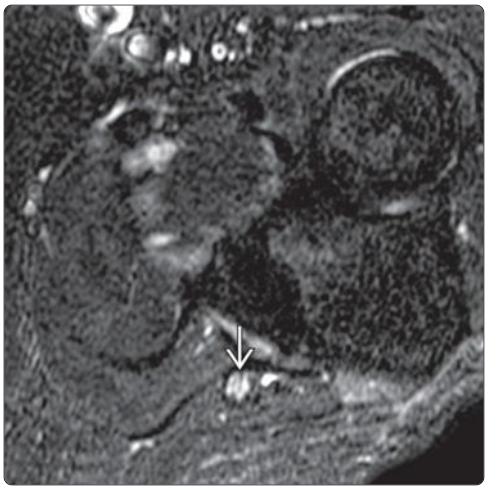
Image Interpretation Pearls

- Focal ulnar nerve enlargement, abnormal T2 hyperintensity at retinaculum just distal to medial epicondyle diagnostic for UN neuropathy

SELECTED REFERENCES

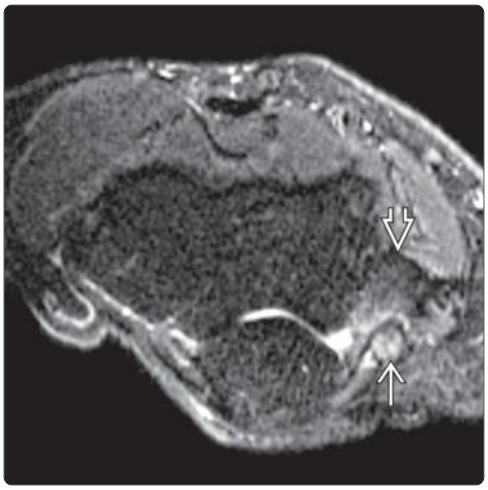
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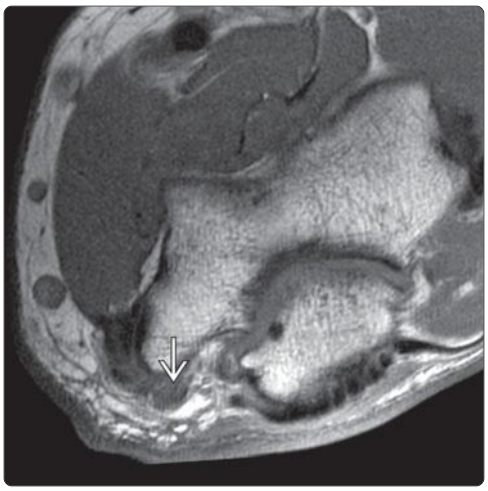


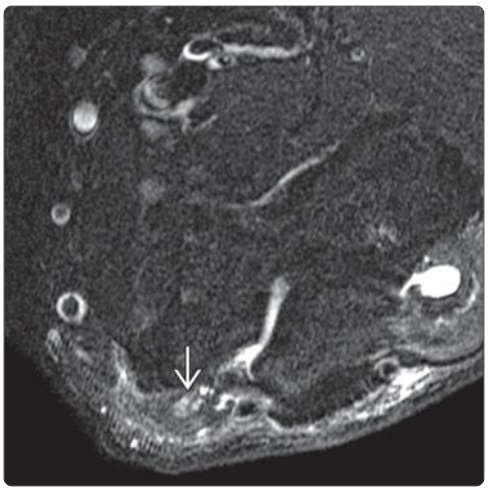
(Left) *Axial T1WI MR of acute ulnar neuropathy demonstrates rounding of the ulnar nerve ⇨ at the cubital tunnel. The fascicles show normal signal intensity but are mildly enlarged. Variant anconeus epitrochlearis muscle ⇨ forms the cubital tunnel roof.* **(Right)** *Axial STIR MR of acute ulnar neuropraxic injury demonstrates abnormal ulnar nerve ⇨ rounding within the cubital tunnel. The nerve fascicles demonstrate marked abnormal T2 hyperintensity approaching that of regional vessels.*





(Left) *Axial T1WI MR of neuropraxic ulnar neuropathy demonstrates abnormal ulnar nerve ⇨ rounding. There is mildly hypointense marrow edema within the medial epicondyle ⇨ suggesting a component of medial epicondylitis.* **(Right)** *Axial STIR MR confirms ulnar nerve ⇨ fascicular enlargement and abnormal T2 hyperintensity with preservation of underlying nerve architecture (neuropraxic injury). The medial epicondyle ⇨ reveals mildly T2 hyperintense marrow edema.*





(Left) *Axial T1WI MR of intermediate severity neuropathy reveals mild hypointensity and ulnar nerve enlargement ⇨ within the cubital tunnel. Regional perineural soft tissue architectural distortion is present.* **(Right)** *Axial T2WI FS MR shows ulnar nerve ⇨ enlargement. Note the internal architectural distortion with several abnormally enlarged hyperintense nerve fascicles with internal nerve architectural distortion; this is typical of axonotmetic injury.*

Suprascapular Neuropathy

KEY FACTS

TERMINOLOGY

- Suprascapular nerve impingement with muscle denervation

IMAGING

- Suprascapular nerve impingement may occur at either spinoglenoid or suprascapular notch
- Spinoglenoid notch: Between neck and blade of scapula, roofed by spinoglenoid ligament
  - Entrapment affects infraspinatus mm only
- Suprascapular notch: At superior border of scapula, roofed by superior transverse ligament
  - Entrapment affects both supraspinatus and infraspinatus muscles
- Denervation edema is 1st sign on MR
  - Uniformly increased signal intensity of muscle
- Fatty atrophy occurs later

TOP DIFFERENTIAL DIAGNOSES

- Cervical radiculopathy
- Parsonage-Turner syndrome (brachial neuritis)
- Rotator cuff tear
- Traction neuropathy
- Neural tumor

PATHOLOGY

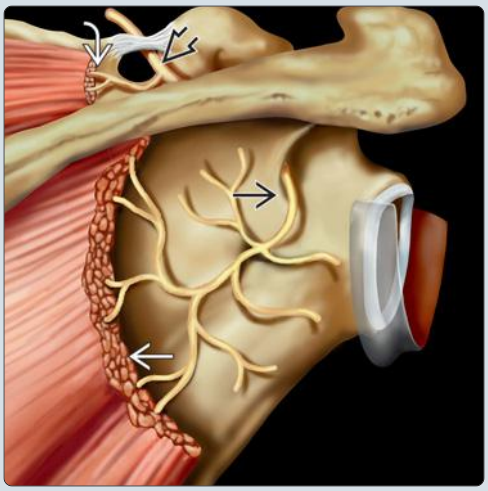
- Paralabral cyst due to tear of posterosuperior glenoid labrum
- Mass or venous varicosities compressing nerve
- Post-traumatic scar or heterotopic ossification

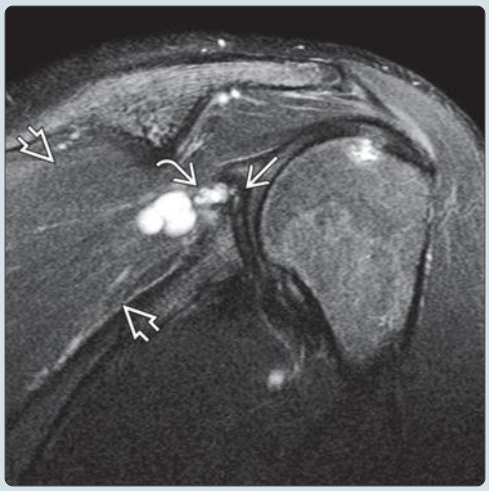
CLINICAL ISSUES

- Young or middle-aged patients, M > F
- Overhead athletes

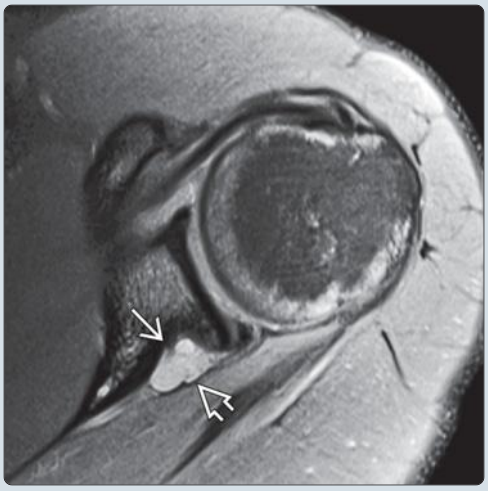
DIAGNOSTIC CHECKLIST

- Denervation edema best seen on sagittal fluid-sensitive sequences





(Left) *Coronal graphic shows suprascapular nerve coursing through suprascapular notch, after which it gives rise to motor branch to supraspinatus muscle. Nerve then passes through the spinoglenoid notch and innervates the infraspinatus muscle.* **(Right)** *Coronal T2WI FS MR shows an elongated, multilocular cyst extending from a posterosuperior glenoid labral tear through the spinoglenoid notch. Infraspinatus muscle shows a mild, homogeneous increase in signal intensity.*





(Left) *Axial PD FSE FS MR in the same patient shows the cyst at the level of the spinoglenoid notch. Denervation edema is less visible on this sequence than on T2 FS or STIR.* **(Right)** *Sagittal T2WI FS MR in the same patient shows the cyst at the spinoglenoid notch. The sagittal image most readily shows the abnormal signal intensity of the infraspinatus relative to the supraspinatus muscle.*

TERMINOLOGY

Definitions

- Suprascapular nerve impingement with muscle denervation

IMAGING

General Features

- Location
  - Suprascapular nerve: Motor nerve to supraspinatus and infraspinatus muscles (mm)
    - Arises from C5 + C6 nerve roots
  - Impingement may occur at either spinoglenoid or suprascapular notch
    - Spinoglenoid notch: Between neck and blade of scapula, roofed by spinoglenoid ligament
      - Entrapment affects infraspinatus mm only
    - Suprascapular notch: At superior border of scapula, roofed by superior transverse ligament
      - Entrapment affects both supraspinatus and infraspinatus mm

MR Findings

- **Rotator cuff abnormalities**
  - Denervation edema is 1st sign
    - Diffuse, homogeneous increase in signal intensity
    - Muscle fibers are intact
  - Fatty atrophy occurs later
    - Loss of muscle mass
    - Fatty streaking between muscle fibers
- **Causes of nerve entrapment**
  - Paralabral cyst
    - Often multilocular
    - Thin cyst neck extends to labrum
    - Rim enhancement with gadolinium
  - Venous varicosities
    - Serpentine, dilated vessels
    - Dilated draining vein
  - Scar
    - Poorly defined soft tissue fullness along course of nerve

Other Modality Findings

- Electrodiagnosis confirms diagnosis

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - Routine shoulder MR; field of view to include spinoglenoid notch

DIFFERENTIAL DIAGNOSIS

Cervical Radiculopathy

- Disc protrusion, osteophytes

Parsonage-Turner Syndrome (Brachial Neuritis)

- Muscles involved are variable
- Denervation changes without visible cause
- Diagnosis based on history and exclusion

Rotator Cuff Tear

- Fluid signal in tendon defect
- Muscle atrophy with chronic tear

Traction Neuropathy

- Large, retracted rotator cuff tear
- Overhead injury in sports

Nerve Sheath Tumor

- Fusiform, follows nerve course

Muscle Tear

- Discontinuity of muscle fibers
- Heterogeneously increased signal intensity on T2WI, STIR

PATHOLOGY

General Features

- Etiology
  - Paralabral cyst due to tear of posterosuperior glenoid labrum
  - Mass or venous varicosities compressing nerve
  - Post-traumatic scar or heterotopic ossification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Weakness, pain

Demographics

- Age
  - Young or middle-aged patients
- Gender
  - M > F
- Epidemiology
  - Overhead athletes

Natural History & Prognosis

- Atrophy of mm tends to progress without treatment

Treatment

- Arthroscopic debridement of labral tear, release of spinoglenoid notch
- Open surgery for other masses

DIAGNOSTIC CHECKLIST

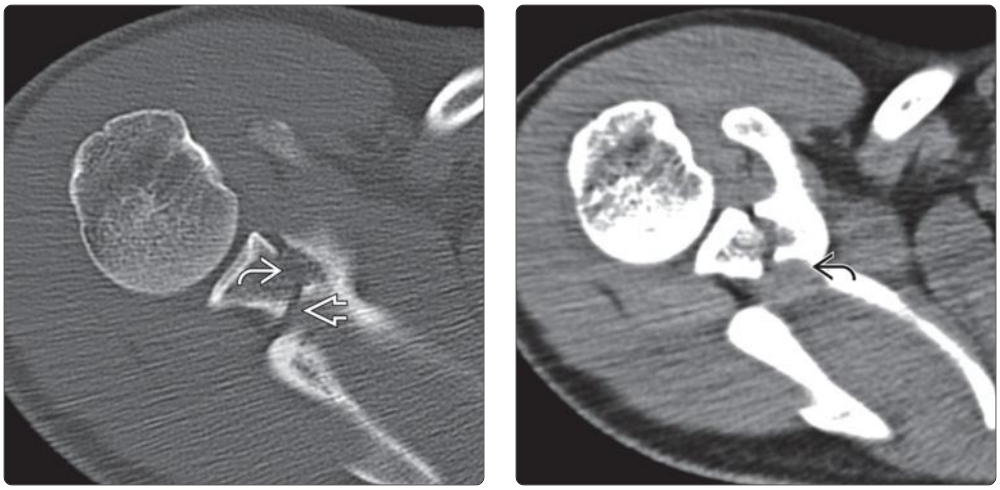
Image Interpretation Pearls

- Denervation edema best seen on sagittal fluid-sensitive sequences

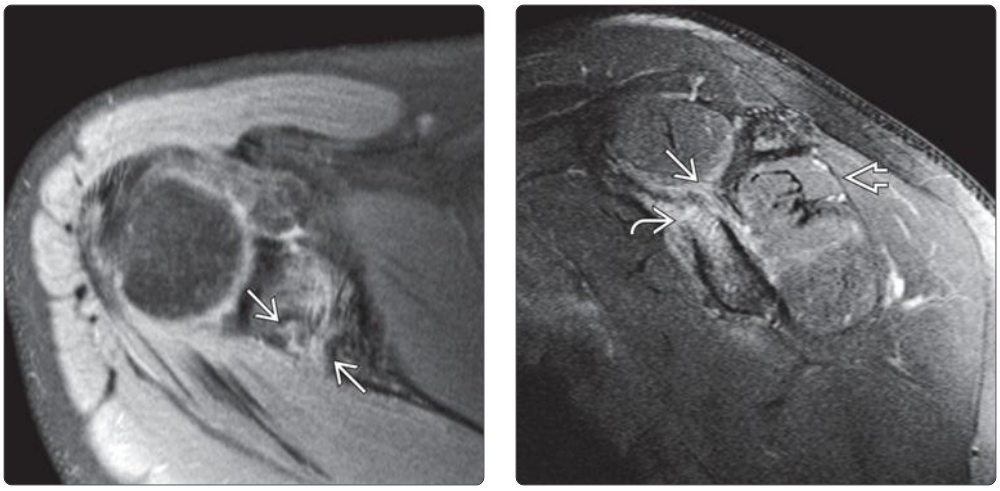
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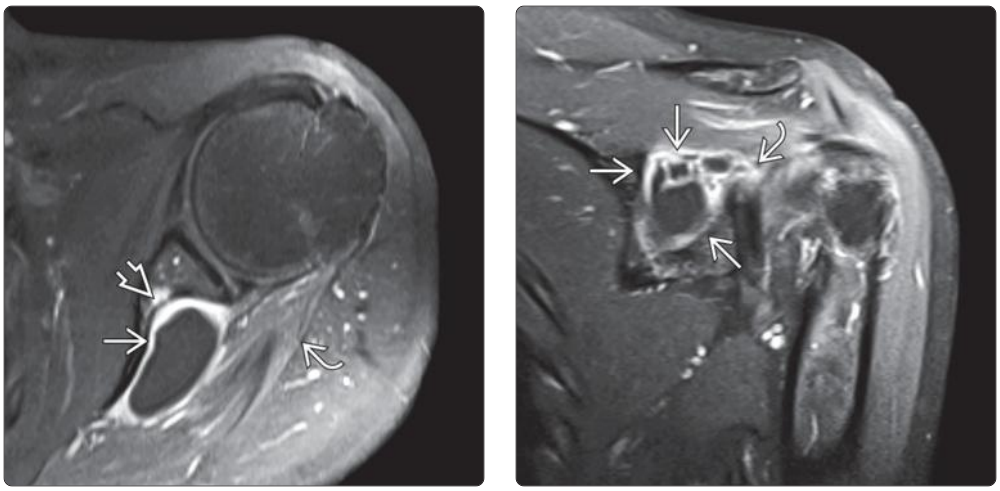
(Left) *Axial bone CT shows a fracture of the scapular neck involving the spinoglenoid notch due to a skiing accident. The patient had infraspinatus muscle weakness.* **(Right)** *Axial NECT shows hematoma filling the spinoglenoid notch. Any space-occupying process in the spinoglenoid notch may cause nerve impingement. Either traumatic nerve laceration or traction injury is more likely than mass effect to be responsible for this patient's symptoms.*



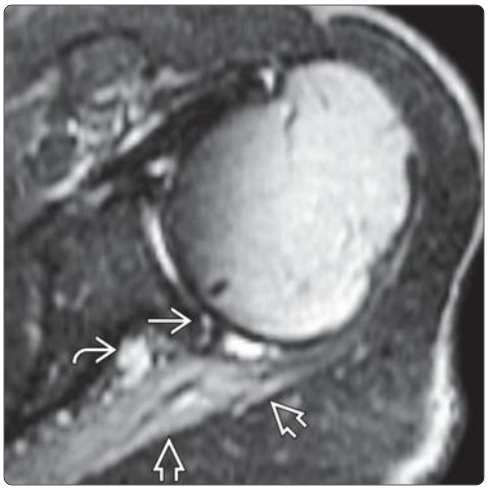
(Left) *Axial PD FSE FS MR in the same patient several weeks later shows soft tissue replacing fat in the spinoglenoid notch. MR was performed because of persistent infraspinatus weakness.* **(Right)** *Sagittal T2WI FS MR in the same patient shows soft tissue consistent with scar in the spinoglenoid notch. There is denervation edema of the infraspinatus muscle. Fracture of the scapula shows blurred margins consistent with early healing.*



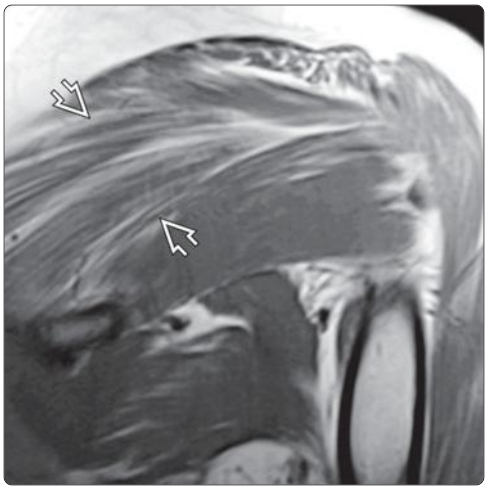
(Left) *Axial T1WI C+ FS MR shows a large cyst with rim enhancement. The cyst has resulted in remodeling of the spinoglenoid notch. Denervation is seen in the infraspinatus muscle.* **(Right)** *Coronal oblique T1WI C+ FS MR in the same patient shows that the cyst is multilocular. A narrow neck extends from the cyst to the posterior glenoid labrum. Cysts tend to recur after excision unless the labral tear is debrided or repaired.*

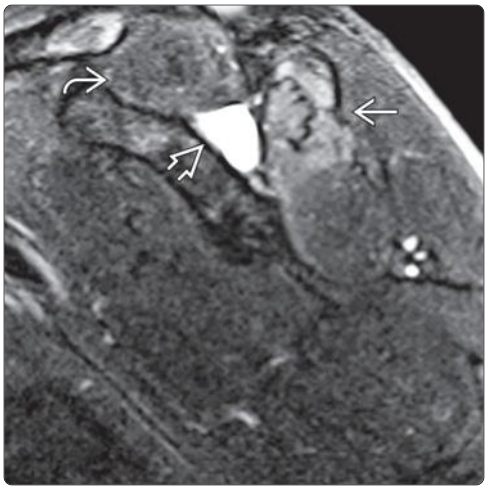




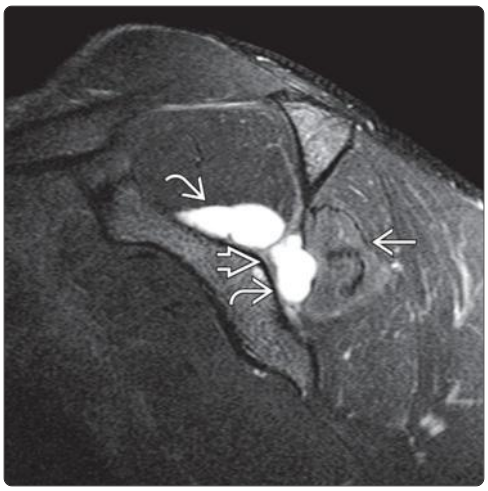


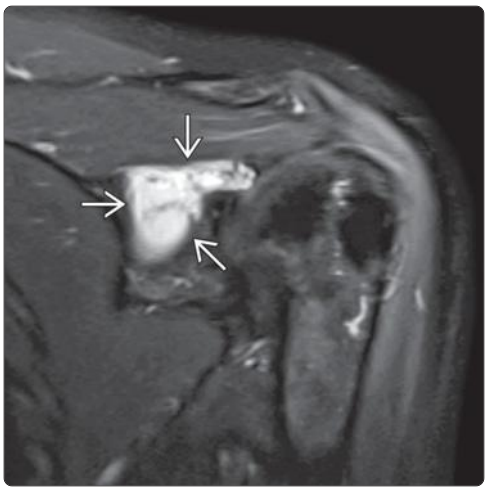
(Left) *Sagittal T2WI MR shows increased signal intensity in the infraspinatus muscle. The central low signal intensity structure is the tendon, which is relatively unaffected by the denervation.* **(Right)** *Axial T2WI MR shows a cyst in the spinoglenoid notch and a posterior labral tear. Despite the small size of the cyst, denervation is evident in the infraspinatus muscle.*





(Left) *Coronal T1WI MR shows infraspinatus muscle atrophy due to longstanding impingement. Atrophy of this severity is uncommonly seen with suprascapular nerve impingement and is more commonly associated with a rotator cuff tear.* **(Right)** *Sagittal T2WI MR in the same patient shows cyst in spinoglenoid notch and striking, homogeneously increased signal intensity in the infraspinatus muscle, characteristic of denervation. Supraspinatus muscle is normal in signal intensity.*





(Left) *Sagittal T2WI FS MR shows a cyst extending through the spinoglenoid notch. Homogeneously increased signal intensity in the infraspinatus muscle is mild but clearly different from surrounding muscles and consistent with denervation.* **(Right)** *Coronal T2WI FS MR shows a large, multiloculated cyst in the spinoglenoid notch. Cavernous hemangioma may have a similar appearance, and gadolinium administration should be performed to exclude hemangioma.*

Median Neuropathy

KEY FACTS

TERMINOLOGY

- Pronator syndrome: Nerve entrapment at pronator teres
- Carpal tunnel syndrome: Nerve entrapment at carpal tunnel

IMAGING

- Nerve enlargement distal to region of entrapment
  - Fascicles indistinct
- Perineural enhancement
- ± mass compressing nerve
- Denervation changes in muscles distal to entrapment

TOP DIFFERENTIAL DIAGNOSES

- Cervical radiculopathy
- Tenosynovitis
- Peripheral nerve sheath tumor
- Thoracic outlet syndrome

PATHOLOGY

- Overuse
- Arthritis
- Tumors
- Fractures
- Anatomic variants

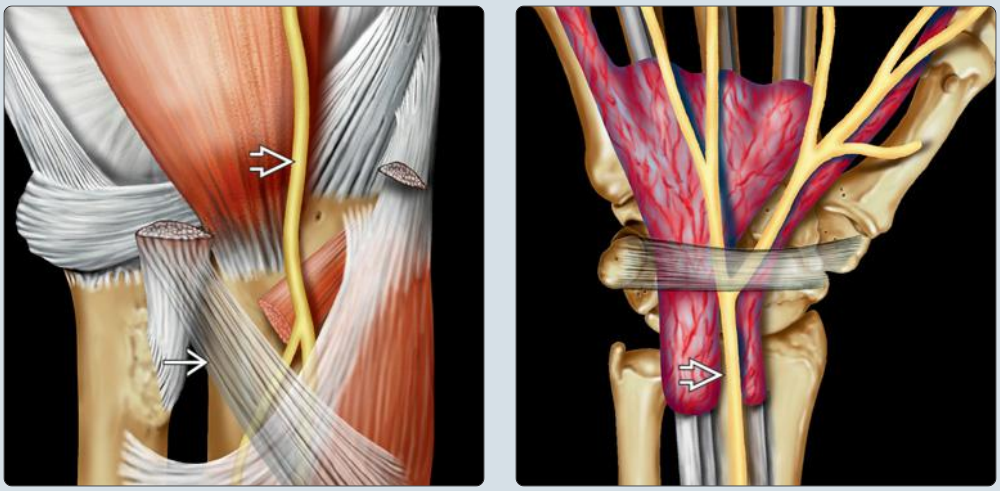
CLINICAL ISSUES

- Tinel sign: Tingling along course of nerve when nerve tapped at point of entrapment
- Electrodiagnosis standard for diagnosis

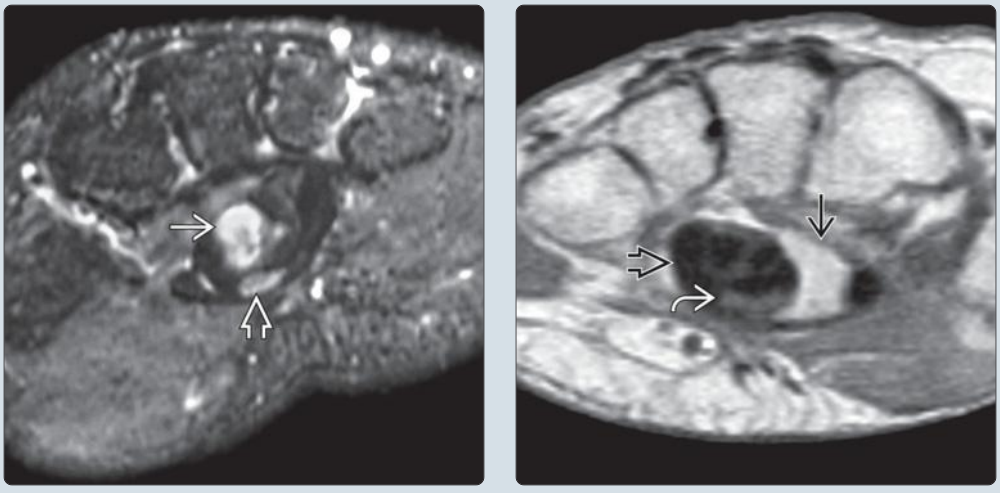
DIAGNOSTIC CHECKLIST

- Imaging diagnosis challenging
  - Nerve normally high signal intensity (in carpal tunnel) on FSE T2WI
  - Nerve normally flattened at level of hook of hamate
- Diagnosis usually made on EMG, not MR

(Left) *Graphic of anterior elbow shows median nerve passing anterior to brachialis muscle. It extends between the heads of the pronator teres and beneath the biceps aponeurosis. Entrapment of the nerve in this region is rare compared to carpal tunnel syndrome. It usually presents with numbness with repeated pronation/supination of the forearm.* **(Right)** *Coronal graphic of the carpal tunnel shows the median nerve at the ventral margin of the carpal tunnel, superficial to the flexor tendons.*



(Left) *Axial T2 FS MR performed at the wrist demonstrates a ganglion cyst in the carpal tunnel, exerting mass effect on the median nerve.* **(Right)** *Axial T1WI MR reveals a lipoma within the carpal tunnel. The flexor tendons are displaced ulnarly, and the median nerve is compressed.*



TERMINOLOGY

Synonyms

- Pronator syndrome: Nerve entrapment at pronator teres
- Carpal tunnel syndrome: Nerve entrapment at carpal tunnel

Definitions

- Carpal tunnel: Fibroosseous tunnel at volar aspect of wrist
  - Contains median nerve and flexor tendons

IMAGING

General Features

- Best diagnostic clue
  - Increased intrinsic nerve signal on STIR MR

MR Findings

- Nerve enlargement distal to region of entrapment
- Indistinct nerve fascicles on cross-sectional T2WI
- Perineural enhancement with gadolinium (Gd)
- Increased anterior bowing of flexor retinaculum
- Denervation changes in muscles distal to entrapment
- Mass compressing nerve in rare patients

Other Modality Findings

- Electrodiagnosis standard for diagnosis

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging
- Protocol advice
  - STIR sequences in long and short axis of nerve
  - T1WI or PDWI in axial plane for definition of anatomy

DIFFERENTIAL DIAGNOSIS

Cervical Radiculopathy

- Consider nerve impingement on cervical spine MR

Tenosynovitis

- Pain, muscle weakness
- May occur ± nerve entrapment

Peripheral Nerve Sheath Tumor

- Fusiform mass arising in or adjacent to nerve

Thoracic Outlet Syndrome

- Cervical rib or soft tissue compression

PATHOLOGY

General Features

- Etiology
  - Overuse
    - Carpal tunnel: Keyboarding, manual labor
    - Pronator: Athletes repeatedly pronating forearm
  - Arthritis
    - Rheumatoid arthritis, amyloid, gout, sarcoidosis
  - Tumors
    - Soft tissue tumor, ganglion cyst, osteochondroma
  - Fractures
    - Supracondylar humerus, distal radius
  - Anatomic variants
    - Supracondylar process and ligament of Struthers
    - Accessory flexor muscles

Gross Pathologic & Surgical Features

- Variable appearance of nerve: Normal, flattened, or enlarged

Microscopic Features

- Perineural inflammation, ± nerve ischemia

Relevant Anatomy

- Course of median nerve at elbow
  - Deep to bicipital aponeurosis (a.k.a. lacertus fibrosus)
  - Superficial to brachialis muscle
  - Passes between ulnar and humeral heads of pronator teres muscle
- Course of median nerve at wrist
  - Superficial and radial aspect of carpal tunnel
  - May divide within carpal tunnel
- Anatomic variants
  - Muscle anomalies, small carpal tunnel
  - Supracondylar process of humerus
    - Ligament of Struthers extends from shaft to medial epicondyle of humerus

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Tinel sign: Tingling along course of nerve when nerve tapped at point of entrapment
  - Tenderness at pronator region or carpal tunnel

Demographics

- Gender
  - Carpal tunnel: F:M = 3:1

Treatment

- Options, risks, complications
  - Conservative: Wrist splint in neutral position, steroid injection
  - Surgery: Open or endoscopic
  - Complications: Reflex sympathetic dystrophy, median nerve injury, incomplete release

DIAGNOSTIC CHECKLIST

Consider

- Diagnosis usually made on EMG, not MR

Image Interpretation Pearls

- Normal nerve appears flat at level of hook of hamate
- Nerve normally high signal intensity on FSE T2WI

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Common Peroneal Neuropathy

KEY FACTS

TERMINOLOGY

- Synonyms: Common peroneal nerve (CPN) palsy, CPN entrapment
- Common peroneal nerve entrapment at fibular head

IMAGING

- CPN swelling, abnormal T2 hyperintensity
  - Fascicular architecture preserved in milder cases (neuropraxic injury)
  - Loss of internal fascicular architecture in more severe cases (axonotmetic, neurotmetic)

TOP DIFFERENTIAL DIAGNOSES

- Ganglion cyst
- Viral neuritis
- Nerve sheath tumor
- Direct acute CPN trauma

PATHOLOGY

- Usual etiology is entrapment or sequelae of continued pressure on CPN at fibular head level
- Edematous/indurated CPN ± thickened "fibular tunnel" at surgery

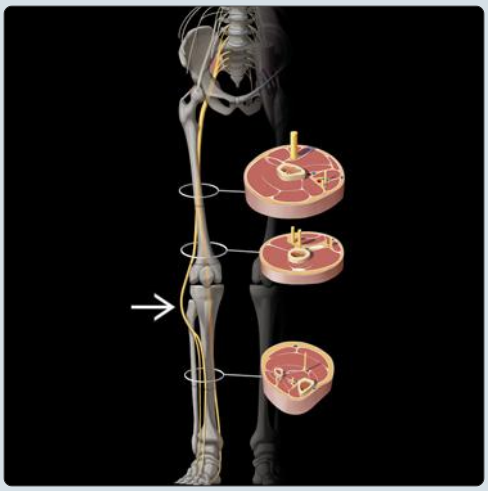
CLINICAL ISSUES

- Foot drop, sensory abnormality along anterolateral leg
- Prognosis variable; frequently good recovery following conservative management
- Surgical decompression reserved for recalcitrant cases

DIAGNOSTIC CHECKLIST

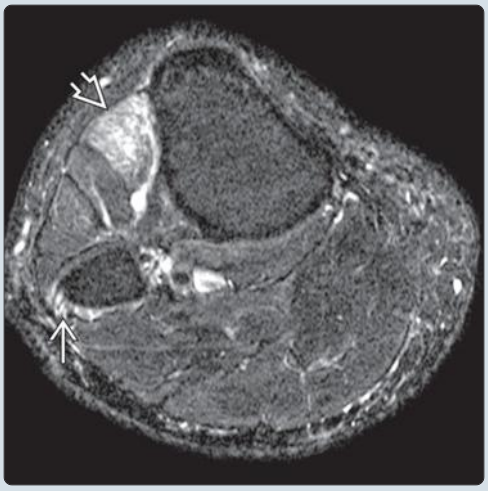
- Focal CPN enlargement, abnormal T2 hyperintensity at fibular head suggests CPN neuropathy
- MR imaging more sensitive for detecting acute than chronic nerve injuries

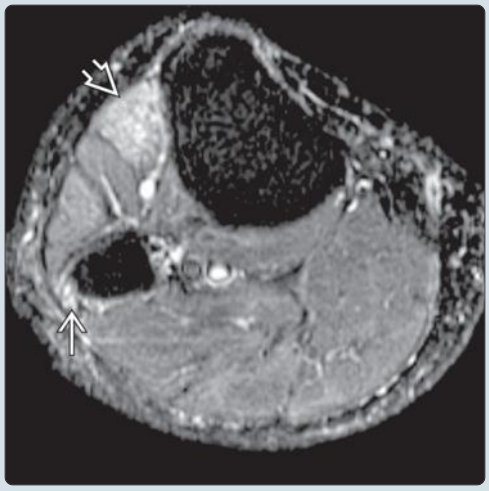
(Left) *Coronal graphic depicts the normal course of the common peroneal neuropathy (CPN) originating from the sciatic nerve, around the fibular head, into the leg. Note the vulnerable position of the CPN as it courses around the fibular head.* **(Right)** *Axial T1WI MR demonstrates atrophy and fatty infiltration of the right anterolateral compartment muscles innervated by the CPN. Mild fascicular enlargement within the right CPN is not as conspicuous on T1WI as it is on T2WI FS MR or STIR MR.*





(Left) *Axial T2WI FS MR reveals abnormal T2 hyperintensity in some of the anterolateral compartment muscles, confirming ongoing denervation superimposed on chronic denervation. The right CPN is mildly enlarged, with swollen, hyperintense fascicles.* **(Right)** *Axial STIR MR confirms abnormal T2 hyperintensity in some anterolateral compartment muscles. The CPN shows mild enlargement and abnormal T2 signal intensity with preservation of intrinsic fascicular architecture.*





TERMINOLOGY

Synonyms

- Common peroneal nerve (CPN) palsy, CPN entrapment

Definitions

- Common peroneal nerve entrapment at fibular head

IMAGING

General Features

- Best diagnostic clue
  - CPN enlargement ± T2 hyperintensity, architectural distortion at fibular head in symptomatic patient
- Location
  - Knee (fibular head level)

MR Findings

- T1WI
  - CPN enlargement ± architectural distortion
- T2WI
  - CPN enlargement, abnormal T2 hyperintensity
  - Loss of internal architecture (more severe cases)
- STIR
  - Similar to fat-saturated T2WI
- T1WI C+
  - ± mild enhancement of nerve, scar tissue

Other Modality Findings

- Nerve conduction velocity: Conduction block at fibular head
- EMG: Anterior compartment (tibial anterior, peroneus longus muscles) muscle denervation

Imaging Recommendations

- Best imaging tool
  - High-resolution MR imaging (MR neurography)

DIFFERENTIAL DIAGNOSIS

Ganglion Cyst

- Uncommon; variable location around knee, ± synovial lining

Viral Neuritis

- History, laboratory findings suggest diagnosis

Nerve Sheath Tumor

- Focal nerve enlargement, loss of fascicular architecture, tumoral enhancement
- Lacks soft tissue inflammatory changes seen in CPN entrapment

Direct Acute CPN Trauma

- Blunt or lacerating injury history

PATHOLOGY

General Features

- Etiology
  - Entrapment or sequelae of continued pressure on CPN at fibular head
  - Orthopedic procedural complication, direct nerve trauma, compression by traction splint, poor operative positioning, or following knee dislocation
- Associated abnormalities
  - Fibular osseous abnormalities ± intraneural or extrinsic ganglion cyst

Staging, Grading, & Classification

- Neuropraxic injury: Mild CPN swelling ± enlarged, hyperintense fascicles
- Axonotmesis: Axonal injury, loss of internal architecture
- Neurotmesis: Complete transection

Gross Pathologic & Surgical Features

- Edematous/indurated CPN ± thickened "fibular tunnel" (musculoaponeurotic arch derived from soleus, peroneus longus muscles)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Foot drop, anterolateral leg sensory abnormality
- Clinical profile
  - Foot drop ± history of acute or chronic lateral knee trauma, recent orthopedic procedure

Demographics

- Age
  - Adults > > children, infants
- Epidemiology
  - Uncommon

Natural History & Prognosis

- Variable; frequently good recovery with conservative management

Treatment

- Conservative: Physical therapy, rigid ankle-foot orthosis (AFO), avoidance of provocative maneuvers
- Surgical decompression if no improvement with conservative measures

DIAGNOSTIC CHECKLIST

Consider

- MR imaging more sensitive for acute than chronic nerve injuries

Image Interpretation Pearls

- Focal CPN enlargement, abnormal T2 hyperintensity at fibular head diagnostic for CPN neuropathy

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Tibial Neuropathy

KEY FACTS

TERMINOLOGY

- Tarsal tunnel syndrome

IMAGING

- Denervation of affected plantar intrinsic muscles
  - Uniform high signal intensity on fluid-sensitive sequences
- Mass in tarsal tunnel
  - Ganglion cyst: Follows fluid signal intensity, homogeneous, ± thin rim of enhancement with gadolinium
  - Nerve sheath tumor: Round or ovoid shape, often shows "tail" where it arises from nerve, diffuse enhancement
  - Venous varicosities: Serpentine, enlarged vessels in tarsal tunnel, can be followed beyond tunnel
- Scar: Amorphous material around tibial nerve
- Osseous impingement: Fracture malunion, subtalar coalition

TOP DIFFERENTIAL DIAGNOSES

- Radiculopathy
- Diabetic neuropathy
- Calcaneal stress fracture
- Isolated fatty atrophy of plantar muscles

PATHOLOGY

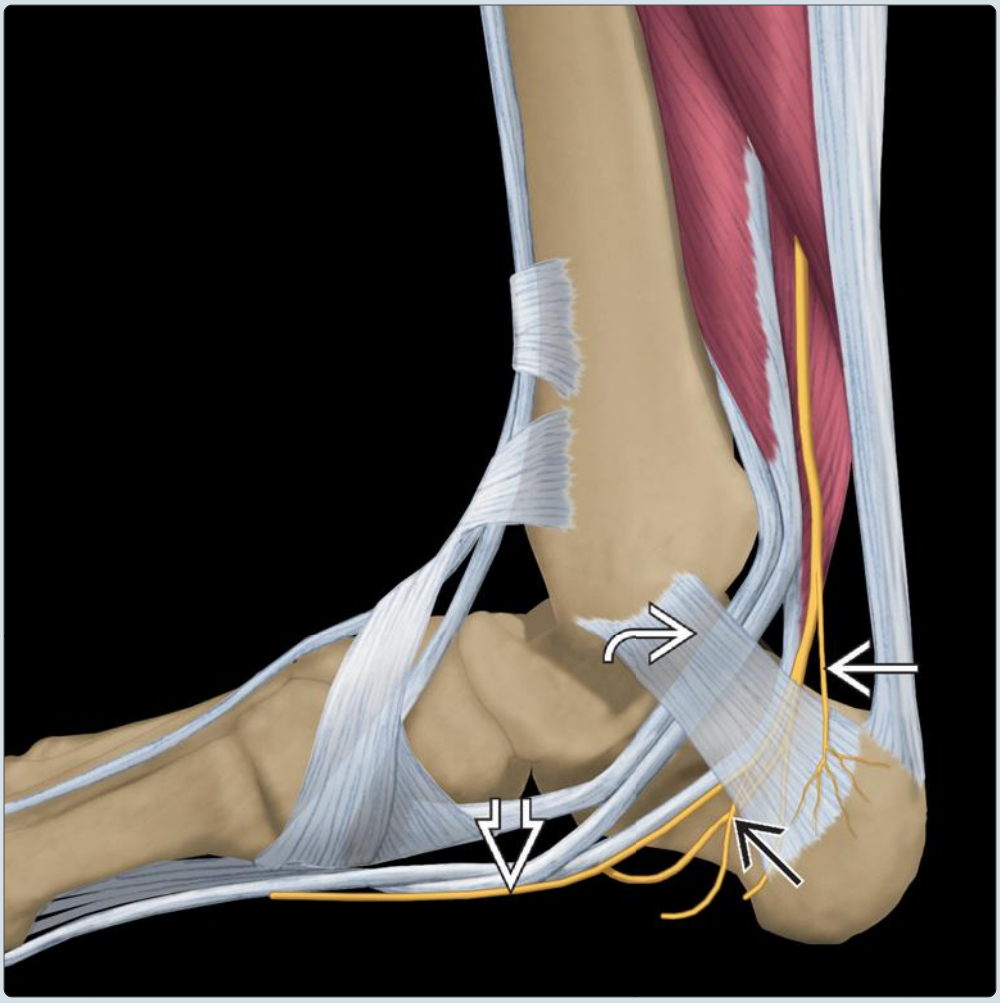
- Up to 50% of cases are idiopathic

CLINICAL ISSUES

- Burning, tingling pain, numbness at plantar aspect of foot

DIAGNOSTIC CHECKLIST

- Fatty atrophy of plantar intrinsic muscles can be incidental finding, increasing with age
- Intramuscular edema is most reliable sign of muscle denervation



Sagittal graphic of the medial side of the ankle shows the borders and contents of the tarsal tunnel. The inferior flexor retinaculum forms the superficial border of the tarsal tunnel, and the floor is composed of the adjacent talus and calcaneus. Posterior tibialis, flexor digitorum longus, and flexor hallucis longus tendons also occupy the tunnel. The tibial nerve is vulnerable to compression throughout its course from behind the medial malleolus to the midfoot. The lateral calcaneal nerve arises in the proximal tarsal tunnel. More distally, the tibial nerve divides into the medial and lateral plantar nerves.

TERMINOLOGY

Synonyms

- Tarsal tunnel syndrome

IMAGING

General Features

- Location
  - Fibroosseous tunnel deep to inferior flexor retinaculum

MR Findings

- Mass in tarsal tunnel
  - Ganglion cyst: Follows fluid signal intensity, homogeneous, ± thin rim of enhancement with gadolinium
  - Nerve sheath tumor: Round or ovoid shape, often shows "tail," where it arises from nerve
    - Uniform or mildly heterogeneous high signal intensity on T2WI, diffuse gadolinium enhancement
  - Venous varicosities: Serpentine, enlarged vessels in tarsal tunnel, can be followed beyond tunnel
  - Accessory muscle: Fusiform, elongated, follows muscle signal intensity on all sequences
- Scar: Amorphous material around tibial nerve, often shows gadolinium enhancement
- Altered osseous morphology of tarsal tunnel
  - Malunited fracture
  - Subtalar coalition with osseous overgrowth
- Enlargement of tibial nerve: Not a reliable finding
- Denervation of plantar intrinsic muscles
  - High signal intensity on fluid-sensitive sequences
    - Uniform appearance throughout affected muscle(s)
    - No disruption of muscle fibers
  - Fat atrophy of muscles is late finding
  - May involve all intrinsic foot muscles innervated by tibial nerve
  - May involve only muscles innervated by individual nerve branches
    - Depending on location where nerve impingement arises

Imaging Recommendations

- Best imaging tool
  - MR

DIFFERENTIAL DIAGNOSIS

Radiculopathy

- L5 or S1 radiculopathy may be clinical mimic

Diabetic Neuropathy

- Diffuse denervation of all plantar intrinsic muscles

Calcaneal Stress Fracture

- Incomplete or complete fracture line

Isolated Fatty Atrophy of Plantar Muscles

- Seen in asymptomatic patients; increases with age

PATHOLOGY

General Features

- Etiology
  - Up to 50% of cases are idiopathic
  - Osseous impingement on tunnel: Due to trauma or subtalar coalition
  - Space-occupying lesion: Ganglion cyst, nerve sheath tumor, venous varicosities, accessory muscle, lipoma, synovitis
  - Scar tissue: Post-traumatic or postsurgical
  - Biomechanical: Valgus heel + pronated forefoot
- Relevant anatomy: Tarsal tunnel
  - Roof: Inferior flexor retinaculum
  - Floor: Medial borders of talus and calcaneus
  - Contains extrinsic flexor tendons and neurovascular bundle
  - Posterior tibial nerve gives off lateral calcaneal branch, then divides into medial and lateral plantar nerves

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Burning, tingling pain, numbness at plantar aspect of foot
- Other signs/symptoms
  - Positive Tinel sign: Percussion of tibial nerve at tarsal tunnel reproduces symptoms

Natural History & Prognosis

- Masses usually require excision
- Idiopathic cases tend to recur

Treatment

- Conservative: NSAIDs, orthotics, physical therapy
- Surgical: Fasciotomy, excision of space-occupying lesion

DIAGNOSTIC CHECKLIST

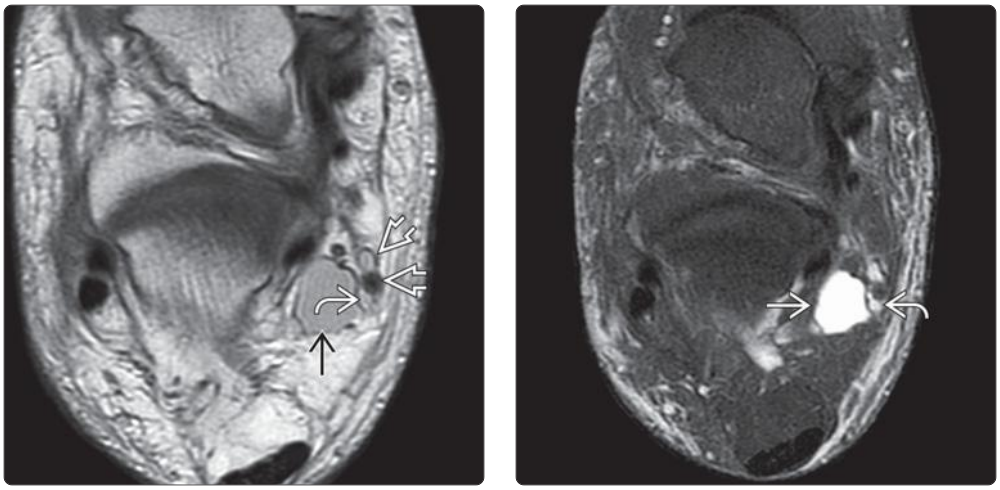
Consider

- Fatty atrophy of plantar intrinsic muscles can be incidental finding, increasing with age
- Intramuscular edema is most reliable sign of muscle denervation

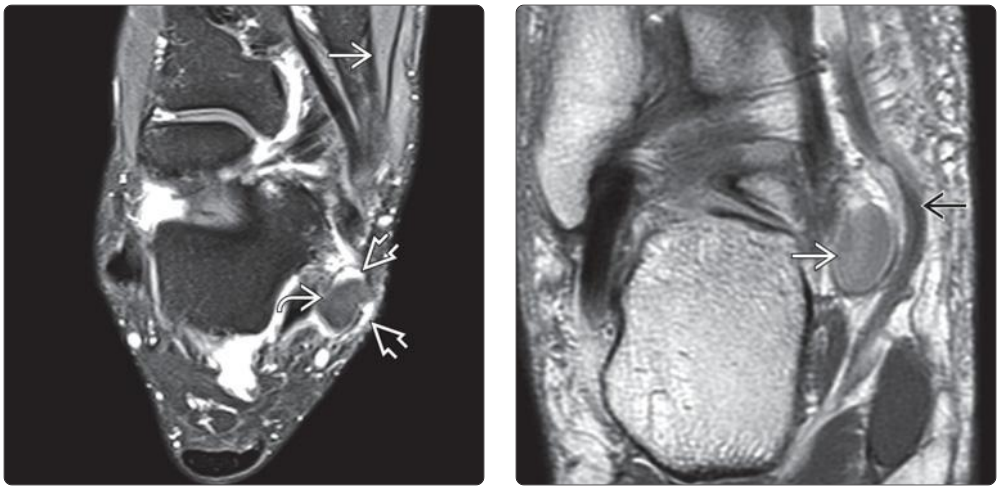
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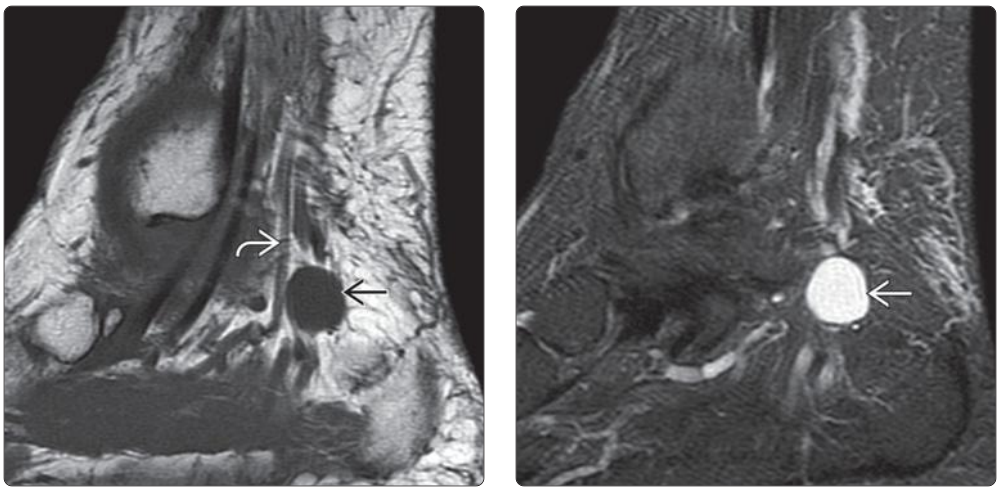
(Left) *Axial PDWI MR shows a ganglion displacing the tibial nerve and posterior tibial vessels medially.* **(Right)** *Axial T2WI FS MR from the same patient shows the ganglion cyst displacing the tibial nerve.*

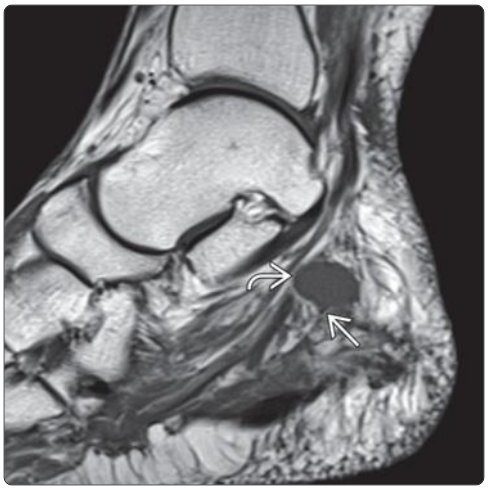


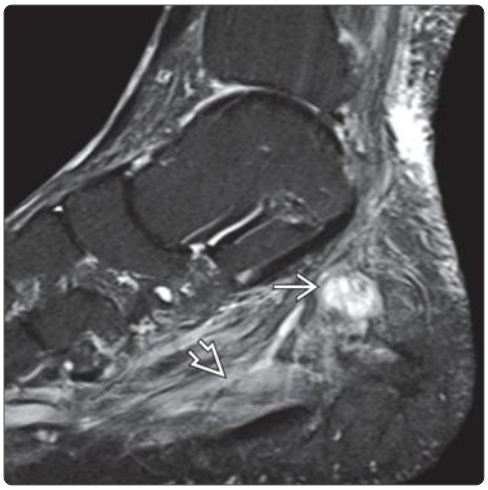
(Left) *Axial T1WI C+ FS MR from the same patient shows that the mass has homogeneously low signal intensity without any enhancement, confirming its cystic nature. Enhancing blood vessels are present adjacent to the mass. There is mild enhancement of the abductor hallucis muscle related to denervation.* **(Right)** *Coronal PDWI MR shows a ganglion displacing the posterior tibial artery medially.*



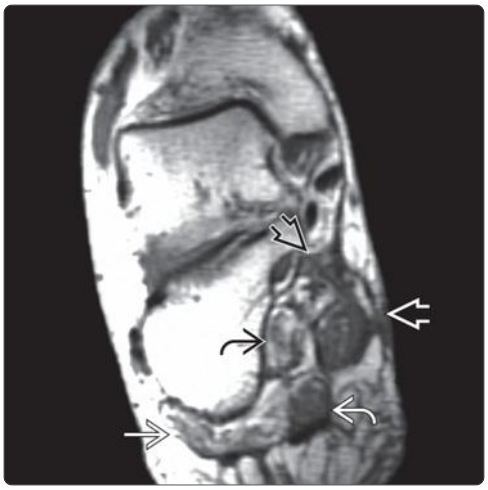
(Left) *Sagittal T1WI MR shows a lobular ganglion cyst in the tarsal tunnel adjacent to the posterior tibial vein. The patient presented with tibial neuropathy.* **(Right)** *Sagittal T2WI FS MR from the same patient shows the cyst is homogeneous in signal intensity, smoothly contoured, and slightly lobulated.*

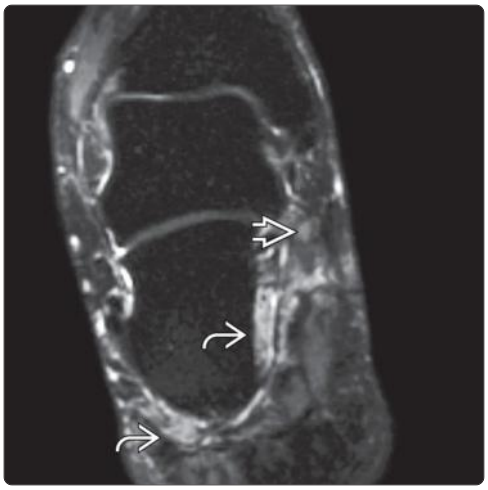






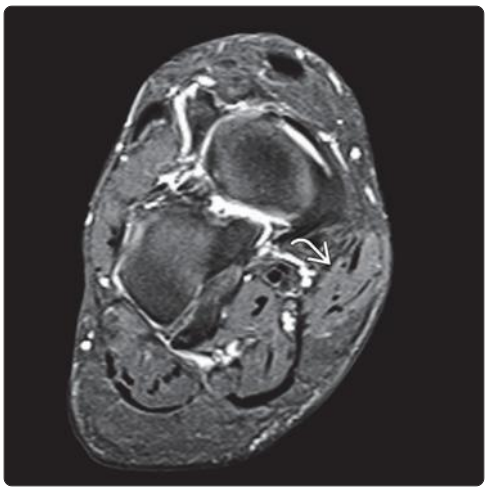
(Left) *Sagittal T1WI MR shows a low signal intensity mass in the tarsal tunnel. The mass has a small "tail", characteristic of nerve sheath tumors. At surgery, this was found to be a malignant peripheral nerve sheath tumor.* **(Right)** *Sagittal STIR MR in the same patient shows that the mass is heterogeneously high in signal intensity. The degree of heterogeneity is greater than expected for a synovial cyst. There is denervation edema in the abductor hallucis muscle.*





(Left) *Coronal PDWI MR in a patient with tarsal tunnel syndrome shows atrophy of the quadratus plantae and abductor digiti minimi muscles, innervated by the lateral plantar nerve. Muscles innervated by the medial plantar nerve (abductor hallucis and flexor digitorum brevis) are spared. The tibial nerve is surrounded by scar.* **(Right)** *Coronal T2WI FS MR from the same patient shows denervation edema of the affected muscles. The tibial nerve appears normal.*



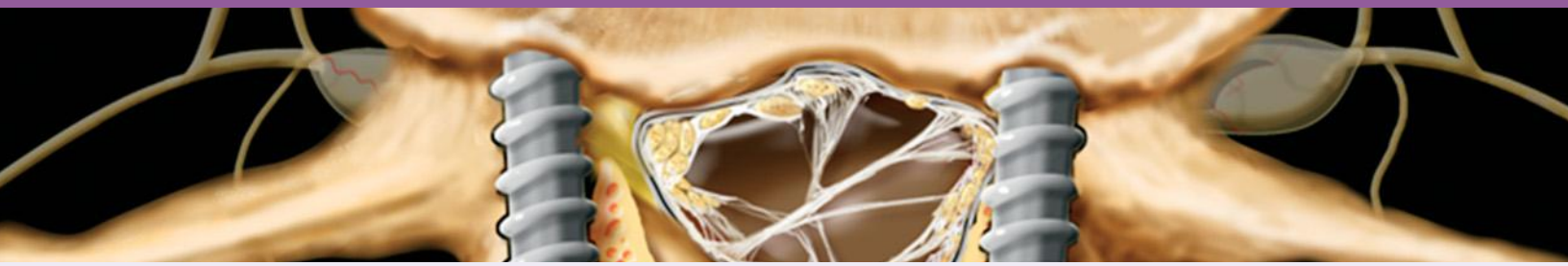


(Left) *Coronal T2WI FS MR shows post-traumatic deformity of the calcaneus due to subacute fracture. The tibial nerve is enlarged due to osseous impingement in this patient who presented with tarsal tunnel syndrome.* **(Right)** *Coronal T2WI FS MR shows isolated mild abductor hallucis muscle denervation due to distal tarsal tunnel syndrome involving the medial plantar nerve.*

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PART VII
SECTION 1

Postoperative Imaging and Complications



Surgical Approaches	**1036**
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Considerations

A particular surgical approach is dictated by multiple factors. Some of these factors include the experience and training of the surgeon, specific normal regional anatomy, symptomatology, the relationship of the pathology to the neural structures, length of pathology to be removed, need for fusion, the physical consistency of the pathology (calcified or not), patient body habitus, and patient comorbidities.

Upper Cervical Spine

Approaches to the anterior upper cervical spine can be generally defined as **transoral** and **retropharyngeal**.

The **transoral approach** is used to access the atlantoaxial complex. This approach can be extended inferiorly to the C3-C4 level via more complex lip splitting with a mandibulotomy. Cephalad extension of the transoral approach can be achieved by splitting the uvula and soft palate, which will allow visualization of the clivus. Complications of this approach include infection by the pharyngeal flora, problems with wound closure, venous hemorrhage, arterial disruption (particularly the vertebral artery), and airway compromise.

The **retropharyngeal approach** can be divided into the **anteromedial** and **anterolateral** techniques. The anteromedial approach goes medial to the carotid sheath, and the anterolateral approach goes lateral to the carotid sheath. Exposure to the upper cervical spine is by partial transection of the sternocleidomastoid muscle and by extending laterally to the carotid sheath. The advantage of this approach is that the major arteries, such as the external carotid artery, and nerves are not involved. The superior extent with this technique is limited to the level of the ring of C1. Complications of the retropharyngeal approach include airway obstruction, hemorrhage, and nerve injury. In particular, the laryngeal nerve and pharyngeal dysfunction are to be avoided. The anterolateral approach also runs the risk of damage to the spinal accessory nerve.

Lower Cervical Spine

Like the approach to the upper cervical spine, the approach to the lower cervical spine can be divided into **anteromedial** and **anterolateral**. The anteromedial approach uses the space between the sternocleidomastoid muscle and the tracheoesophageal complex medially. The cervical spine is often approached from the left, especially at the C6-C7 level, to minimize the chance of laryngeal nerve injury (since the laryngeal nerve leads the carotid sheath higher on the right). Surgical dissection should avoid the tracheoesophageal groove region since the recurrent laryngeal nerve lies within that space.

The anterolateral approach posterior to the carotid sheath avoids the vessels to the thyroid, the vagus nerve, and the superior laryngeal nerve. The surgeon carries the dissection down anterior to the anterior scalene muscle and anterior to the anterior tubercle of the transverse process to avoid the vertebral artery or exiting nerve root. The primary complication to be avoided is direct neurologic injury of the cord or root. Airway obstruction may occur due to edema and soft tissue hemorrhage. Hematoma is a postoperative complication in approximately 9% of cases where the anterior approach is followed. Esophageal perforation is also a possible complication. Hoarseness or involvement of the recurrent laryngeal nerve is encountered infrequently, on the order of 1%. Injury to the sympathetic plexus can give rise to Horner syndrome. The sympathetic chain lies on the anterior surface of the longus colli muscle, posterior to the carotid sheath.

Posterior Cervical Spine

The **posterior approach** is a common and generally safe approach to the cervical spine allowing for direct access to the posterior elements from the skull base to the thoracic spine. The laminectomy is used as a basic approach to deal with a variety of spine lesions, such as tumor, trauma, and congenital malformations. Subperiosteal dissection of muscles exposes the spinous processes, lamina, and lateral mass and facets. The facet capsule is a stabilizing structure and is kept intact except when a fusion is being performed. Posterior screw plating is a common method of fusion. Multiple different techniques are utilized for screw placement with the objective of solid placement, which avoids the cord, roots, and vertebral arteries. The original description suggested a screw entry point in the center of lateral mass with screws angled 10° laterally. Magerl recommended 25° laterally and 45° superiorly.

For hemilaminectomy with foraminotomy, a midline incision is made with dissection to the deep nuchal ligaments, with paraspinal muscles being elevated to expose the lamina and medial facets. The medial facet is removed, ligamentum flavum opened, and nerve root exposed. Complications from this approach include air embolus (if the sitting position is utilized), wound infection, and rarely, nerve root injury.

Thoracic Spine

Laminectomies were initially used to excise thoracic disc herniations starting in the 1950s but quickly fell out of favor due to the unacceptably high incidence of postoperative complications (including paraplegia) related to cord manipulation. A wide variety of approaches has been developed to avoid this complication. Approaches to the thoracic spine can be categorized as anterior (**transthoracic**, **transsternal**, and **thoracoscopic**) and **posterolateral.**

Anterior approaches include transthoracic with thoracotomy for exposure of T2-L2 levels and is used for a variety of thoracic pathologies. The patient is placed in a lateral decubitus position and generally approached from the left side since it is easier to handle aorta and segmental vessels from this approach. The appropriate rib is resected with care not to injure the intercostal vessels. The lung is retracted and rib separator used to give exposure to the spine. The parietal pleura is excised, allowing visualization of the disc. The left-sided approach is much more difficult above the T4 level due to presence of the mediastinum.

Posterior thoracic approaches are numerous but include **laminectomy**, **transpedicular**, **costotransversectomy**, and **lateral extracavitary**. Laminectomies are generally performed for a wide range of pathologies, including posterior decompression, thoracic discectomy, posterior fusions, tumor resection, intradural masses, and a variety of stabilization procedures. Laminectomy is not utilized for anterior disease in the thoracic spine. The thoracic cord is particularly susceptible to manipulation, which has caused the development of multiple approaches attempting to minimize this manipulation but still provide direct disc or vertebral body access. The transpedicular approach starts in the midline with reflection of the paraspinal muscles to the side. The dissection is carried laterally beyond the facet joint. The facet and pedicle are resected, and the disc space is exposed for removal. After

decompression of the cord is performed, a laminectomy may also be performed. A bilateral transpedicular approach can also be performed for more complete vertebral body removal. Costotransversectomy is a posterior lateral approach used for partial vertebral body resections, fusions, and decompressions. There is resection of the medial rib and disarticulation of the costotransverse and costovertebral junctions. The sympathetic trunk and parietal pleura may be elevated away from the vertebral body, allowing direct vertebral body access.

The lateral extracavitary approach allows for a combined anterior and posterior exposure through a single excision but is technically demanding and requires a longer operative time with increased blood loss. After the rib is resected and joint disarticulated, the intercostal nerves and vessels are isolated (if a vascular graft is used) or resected. Remaining outside the pleural cavity, the pedicle is removed, allowing visualization of the thecal sac and roots. One key advantage of this approach is allowing visualization of the lateral aspect of the dura before visualization of the pathology.

Lumbar Spine

The anterior approach to the lumbar spine is guided by the regional anatomy: The quadratus lumborum and psoas muscles; iliohypogastric and ilioinguinal nerves along the lateral margin of the psoas; splanchnic nerves, sympathetic tracks between the medial psoas and vertebral body. The aorta bifurcates at L4-L5, with the common iliac arteries lying across L5. The inferior vena cava forms from the confluence of the right and left common iliac veins at L5. The common iliac veins lie below and to the right of the arteries. The ureters extend over the iliac vessels laterally.

The **transperitoneal** approach opens the peritoneum, and the viscera are retracted to expose the posterior peritoneum and vessels. The **retroperitoneal** approach dissects the peritoneum off of the abdominal wall laterally. At the psoas level, the peritoneum and ureter are lifted off the psoas and retracted medially. Potential complications from the anterior approach of the lumbar spine are varied due to the presence of major arteries, veins, and sympathetic plexus. Injury may occur to the superior hypogastric plexus, which may lead to retrograde ejaculation. This complication appears much more commonly with the transperitoneal and retroperitoneal approaches. The overall incidence of vascular complications is 6% with a 1% mortality. Thrombotic occlusion of the left common iliac artery after anterior transperitoneal approach has been reported by multiple authors. Hemorrhage from disruption or tear of the inferior vena cava or lumbar vein may require suture, clip ligation, cautery, or Gelfoam. Visceral complications must also be considered, such as ileus, pancreatitis, and bowel perforation. The anterior approached to the lumbar spine shows a complication rate of 20%, with 12% being major complications.

A variety of spinal fusion techniques are available to the surgeon, including **posterior fusion** (PF), **posterior lateral fusion** (PLF), **anterior lumbar interbody fusion** (ALIF), **posterior lumbar interbody fusion** (PLIF), and **extreme lateral interbody fusion** (XLIF). PLIF is often used for treatment of segmental spinal instability from degenerative disc disease or spondylotic spondylolisthesis. This approach can also tackle all three spinal columns via a single posterior approach by removing the disc, correcting segmental instability, and providing segmental fixation (via pedicle screws or wiring). It also avoids the vascular and sympathetic neural injuries of anterior lumbar interbody fusion. A conventional PLIF exposure requires bilateral laminectomies and medial facetectomies with preservation of the posterior midline spinous processes and interspinous ligaments. This approach provides a small working window but maintains stability. A more extensive PLIF exposure requires total facetectomies and wider laminectomies. This approach gives a larger working window but requires internal fixation. Endoscopic or "minimally invasive" versions of this have been developed. Avoidance of neural injury requires adequate mobilization of neural elements, retraction of the thecal sac only to the midline, and intermittent release of traction, which does not produce compression of the neural elements for more than 30 minutes at a time. The minimum area of disc and endplate cartilage to be removed to provide a graft bed is controversial, but decorticated endplates on the order of 2.5 x 2.5 cm have been advocated. PLIF surgery with this degree of bed preparation shows a high degree of fusion. Multiple different materials may be used as graft material or fusion devices. Autograft from posterior iliac crest is common. Allograft may give an area too small to use by themselves without additional autograft or internal fixation to avoid graft subsidence.

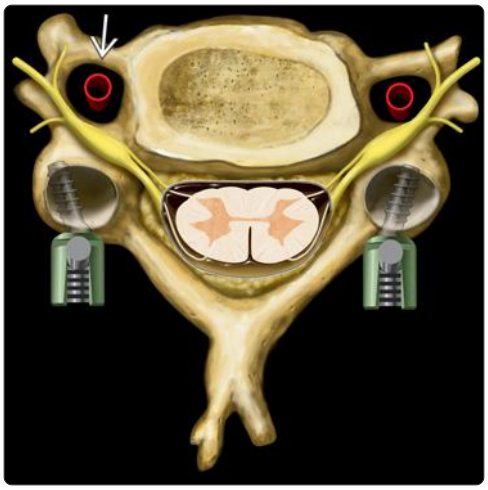
The **extreme lateral interbody fusion technique** (XLIF) involves entering the retroperitoneum through minimally invasive incisions, and then passing the specialized instruments transversely through the retroperitoneal fat and psoas muscle, (avoiding the lumbar plexus branches). This allows direct access to the lateral margin of the annulus and intervertebral disc. This far lateral approach does not involve peritoneal disruption and avoids the difficulty of manipulating the anterior major vessels and the associated potential complications. A discectomy can be performed while the posterior annulus remains intact. An implant/graft is then placed in the disc space. The XLIF implants have a typical long, rectangular shape which is easily recognized on axial CT or MR, and provides a large surface area for fusion to occur.

Selected References

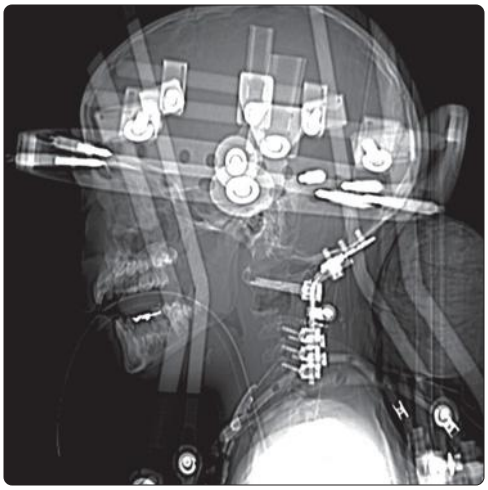
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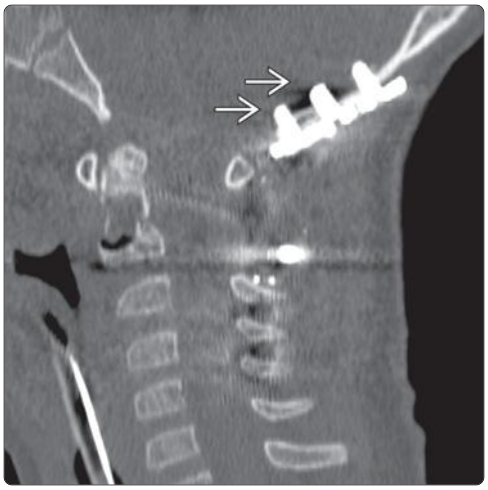
(Left) *Axial graphic shows the anteromedial approach (green line), which uses the space between the sternocleidomastoid muscle and the tracheoesophageal complex medially. The anterolateral approach (black line) posterior to the carotid sheath avoids the vessels to the thyroid, the vagus nerve, and the superior laryngeal nerve.* **(Right)** *Axial graphic shows posterior instrumented fusion with facet screws and longitudinal rod fixation. Exiting roots and vertebral arteries within the transverse foramen ➡ are to be avoided.*



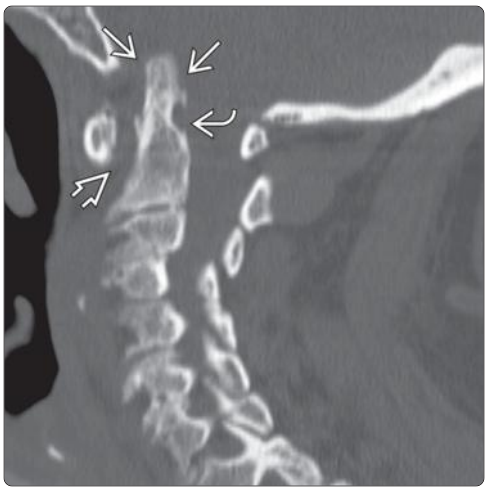


(Left) *Imaging was taken following resection of C2 aneurysmal bone cyst. Postoperative CT exam shows the normal appearance following C2 corpectomy and resection of the posterior elements with occiput to C3-C5 fusion with patient in halo frame.* **(Right)** *Sagittal NECT shows the occipital part of the hardware construct with 3 screws extending through the inner table ➡ following C2 corpectomy and resection of the posterior elements with occiput to C3-C5 fusion with patient in halo frame.*

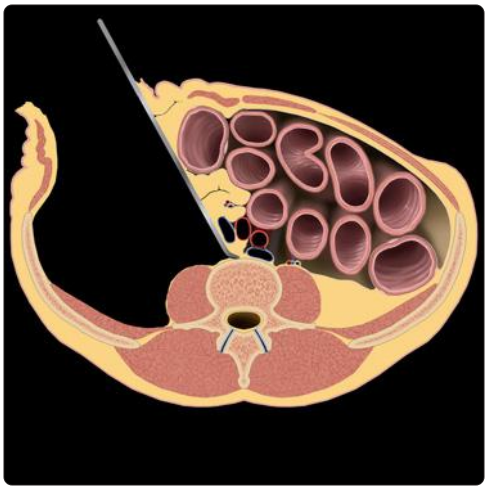


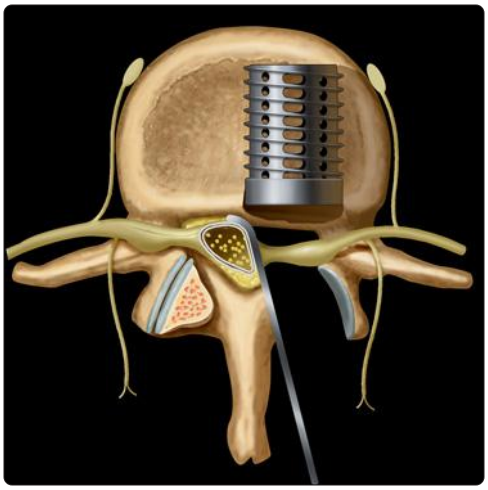


(Left) *CT image in this patient with RA shows cranial settling with upward translocation of the odontoid process relative to foramen magnum ➡. There are odontoid erosions ➡, and increased atlantodental interval ➡.* **(Right)** *Sagittal CT shows changes of 2 stage surgery, with the 1st being posterior occiput to C4 fusion ➡ and the 2nd stage the anterior resection of odontoid process ➡. Bone graft material has been laid down along the posterior elements ➡.*

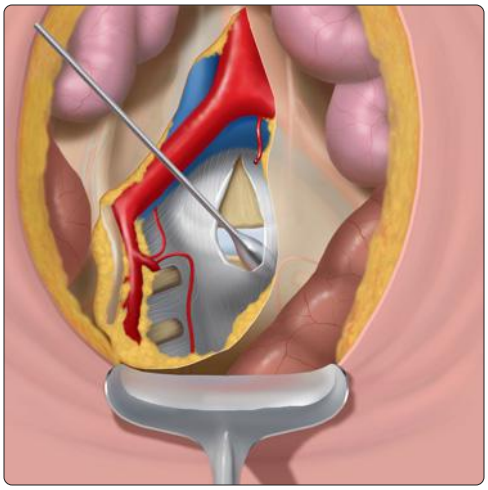






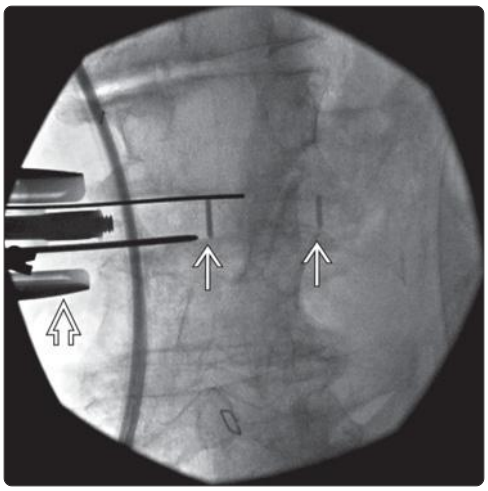


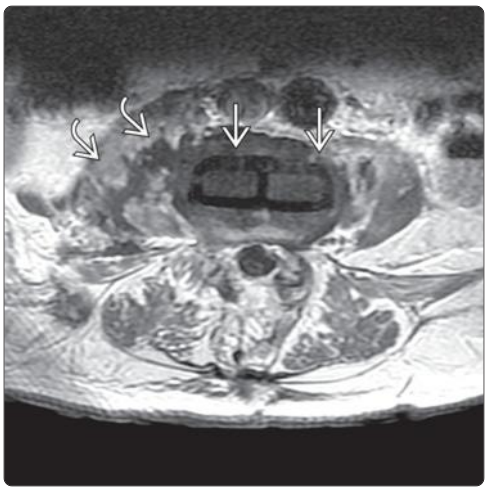
(Left) *Axial graphic of retroperitoneal approach shows dissection of the peritoneum off of abdominal wall laterally. At the psoas level, the peritoneum and ureter are lifted off the psoas and retracted medially.* **(Right)** *Axial graphic shows conventional posterior lumbar interbody fusion exposure with laminectomy and medial facetectomies, with preservation of the posterior midline spinous processes and interspinous ligaments. This approach provides a small working window but maintains stability.*





(Left) *Coronal graphic shows transperitoneal approach for lumbar anterior interbody fusion at L5-S1 level. The anterior approach to the lumbar spine shows complication rate of 20% (12% major).* **(Right)** *Sagittal T2WI MR shows the normal appearance of dorsal soft tissues immediately following successful multilevel laminectomy. Low signal ➪ reflects serosanguineous fluid in operative tract and Gelfoam. Moderate mass effect on thecal sac may occur without symptomatology.*



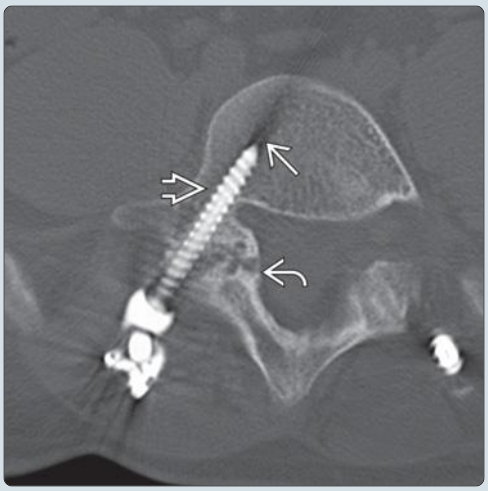


(Left) *Intraoperative image of extreme lateral interbody fusion (XLIF) approach shows specialized instruments ➪ allowing placement of an interbody fusion graft ➪ through the dilated and retracted tissue from a direct lateral approach through the psoas.* **(Right)** *Normal imaging appearance of the postoperative change from an XLIF that accesses the disc space through the psoas muscle. MR study shows the operative tract through the right psoas muscle ➪ and the interbody fusion graft ➪.*

KEY FACTS

TERMINOLOGY

- Substantial imaging overlap in routine early postoperative findings vs. surgical complications
  - Early postoperative imaging not routinely obtained
  - Edema, granulation tissue at operative site may simulate residual/recurrent disc herniation
  - Complex fluid in surgical bed may simulate infected fluid collection
  - Marrow changes and fluid associated with intervertebral fusion may simulate appearance of spondylodiscitis
  - Clumping, enhancement of nerve roots may simulate arachnoiditis
- Metallic hardware interferes with postoperative imaging
  - Beam-hardening artifact on CT
  - Susceptibility artifact, warping, poor fat saturation on MR
- Clear zones around pedicle screws
  - Not unusual
  - Not necessarily indicative of loosening
  - Tend to resolve over time
  - Enlarging lucencies worrisome for pseudarthrosis
- Pseudarthrosis
  - Pitfalls of interpretation on all imaging modalities
  - Sometimes difficult to identify geometrically complex pseudarthrosis on CT
  - Intact hardware may limit appreciation of movement on flexion/extension radiographs
  - Many false-negatives and false-positives on bone scan
  - Operative exploration remains gold standard
- Dorsal fluid collection may simulate early pseudomeningocele
  - Severity of compression by epidural fluid does not predict a patient's neurological status
  - Poor neurological exam after decompressive surgery may require immediate return to the operating room for exploration, since immediate postoperative imaging not sufficiently sensitive to pathology





(Left) *Axial NECT shows pedicle screw placement in L5 as part of posterior fusion for spondylolytic spondylolisthesis at L5-S1(note chronic L5 spondylolysis). There is no abnormal lucency at the bone-screw interface except for beam hardening artifact near the tip.* **(Right)** *Sagittal reformatted NECT shows intervertebral fusion of L5-S1 using fusion cages. Mature bone bridges the intervertebral space, demonstrating a good bony union.*





(Left) *Axial image from a CT myelogram shows changes of a left hemilaminotomy. The actual bony defect is difficult to appreciate on this image. What is apparent is the absence of the left ligamentum flavum on this image.* **(Right)** *Sagittal T2 MR shows early appearance of L5-S1 PLIF with low signal graft material within the disc space and surrounding hyperintense fluid.*

TERMINOLOGY

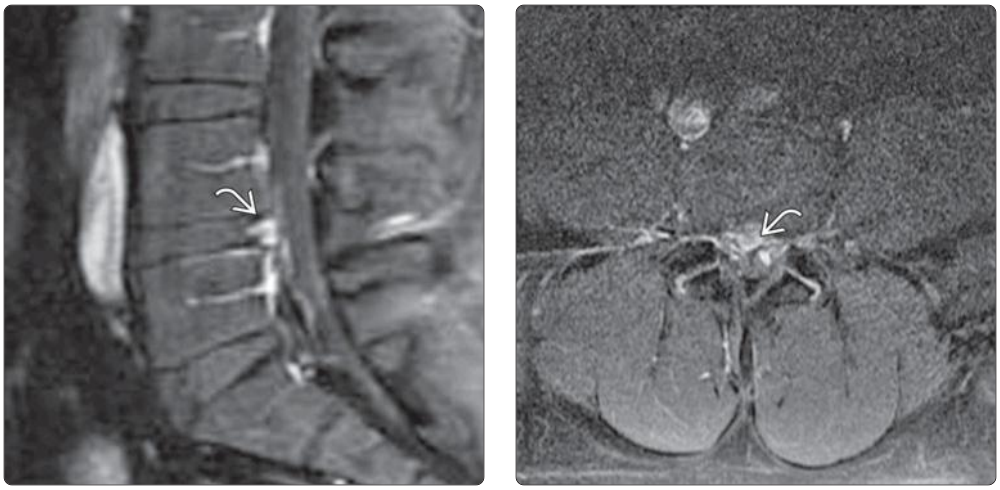
Definitions

- Discectomy or partial discectomy
  - Surgical removal of herniated portion of intervertebral disc
- Laminectomy
  - Removal of lamina to decompress spinal canal
    - Unilateral (hemilaminectomy) or bilateral
    - Partial removal of lamina and ligamentum flavum is technically a **laminotomy**, although terminology sometimes used interchangeably
- Interbody fusion
  - Disc removal by posterior (posterior lumbar intervertebral fusion [PLIF]) or anterior (anterior lumbar intervertebral fusion [ALIF]) approach
  - Insertion of bone graft &/or fusion hardware
  - Goal is arthrodesis (fusion) across disc space
- Posterior instrumentation
  - Including pedicle screws, paraspinous rods, transverse rods, laminar hooks
    - Translaminar or facet screws; can be inserted with minimally invasive techniques
- Posterolateral fusion
  - With severe loss of disc height, in lieu of interbody fusion
  - Lateral bone graft placement, fusion of transverse processes
  - Usually supplemented by posterior instrumentation
- Disc replacement
  - Variety of intervertebral hardware
  - Preserves motion across segment: Arthroplasty rather than arthrodesis
- Pseudoarthrosis
  - Failure to obtain bony union after fusion
  - Ideally, fused segment will show
    - Mature bridging bone on radiographs, CT
    - Cold on bone scan 6-12 months postop
    - Resolution of type I endplate marrow or conversion to type II
    - No motion on flexion/extension radiographs
  - However
    - Geometry of pseudoarthrosis may be complex and difficult to appreciate on radiographs, CT
    - Bone scan performs poorly, with significant false-negative and false-positive findings
      - Somewhat better performance at 12 months relative to 6 months postop
    - Stability provided by instrumentation may prevent motion on flexion/extension films, despite pseudoarthrosis
      - 2-3° motion may be present with fusion due to compliance of normal bone
    - Pedicle screw fracture can be seen with pseudoarthrosis
      - Instrumentation temporizing mechanism until bony union can occur
- Malpositioned pedicle screw
  - Pedicle screw should transverse pedicle and be securely positioned within vertebral body
  - Malpositioning includes
    - Perforation of anterior cortex of vertebral body
    - Perforation of cortex of pedicle
      - ± compromise of intervertebral neural foramen or spinal canal
- Lucency around pedicle screw
  - Clear zones (≥ 1 mm) around pedicle screws may be encountered on postoperative radiographs
    - Traditionally, concerning for loosening or infection
  - 1 longitudinal series describes majority (2/3) of clear zones as resolving over several years
    - Persistence 2 years postoperatively predictive of pseudoarthrosis
- Peridural fibrosis
  - Some degree of peridural fibrosis along margin of thecal sac is typical finding following discectomy
    - Edema and tissue disruption at discectomy site particularly conspicuous in 1st 6 weeks postoperatively
      - Can simulate disc residual/recurrent disc herniation
  - Scarring has been implicated with nerve root irritation in failed back surgery syndrome (FBSS)
- Postoperative fluid collection
  - Fluid collection in operative bed is common in immediate postoperative setting
    - May have complex signal, with fluid-fluid levels
    - Will demonstrate peripheral enhancement
  - Can be difficult to differentiate from postoperative hematoma, pseudomeningocele, infected collection
  - Severity of compression by epidural fluid does not predict a patient's neurological status
    - Poor neurological exam after decompressive surgery may require immediate return to the operating room for exploration, since immediate postoperative imaging not sufficiently sensitive to pathology
- Nerve root clumping, enhancement
  - Transient nerve root clumping can be identified in early postoperative period, often resolves spontaneously
  - Solitary nerve root enhancement may be seen perioperatively; inflammation related to compression, manipulation

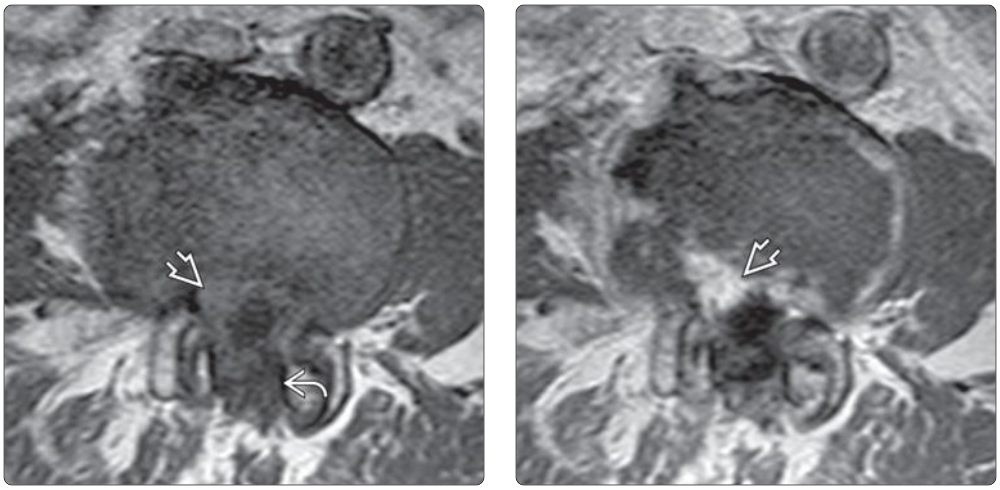
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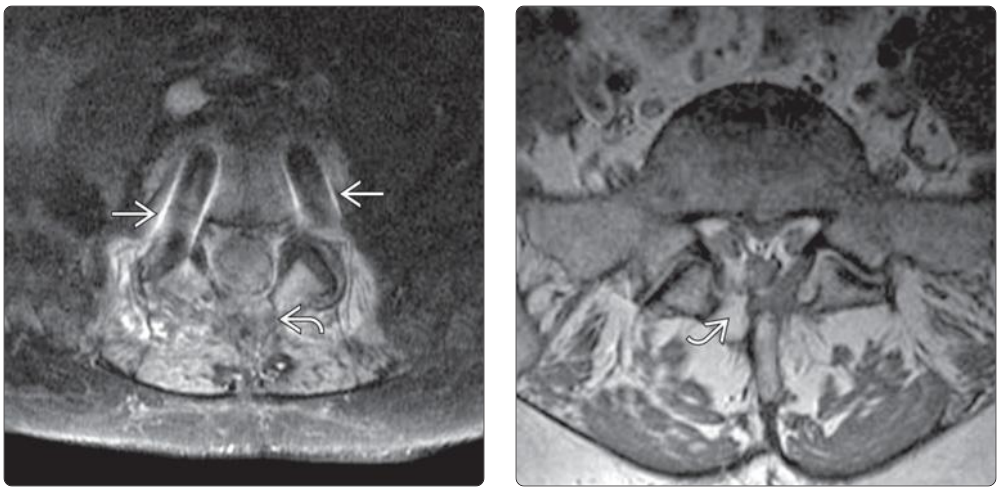
(Left) *Sagittal T1 C+ FS MR shows enhancement in the dorsal margin of the L4-L5 disc at the site of a partial discectomy.* **(Right)** *Axial T1 C+ FS MR shows a small amount of enhancing peridural fibrosis at the site of a partial discectomy. The enhancement differentiates fibrosis from a recurrent disc herniation.*

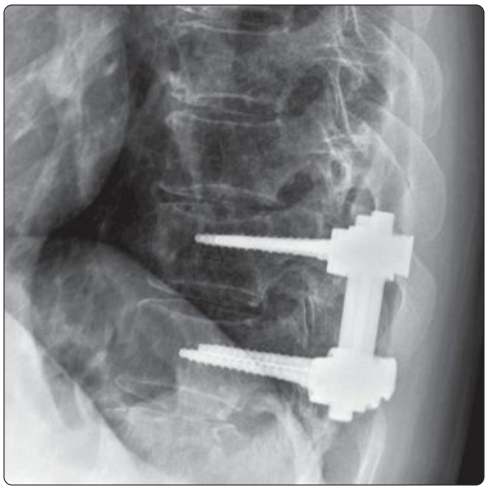


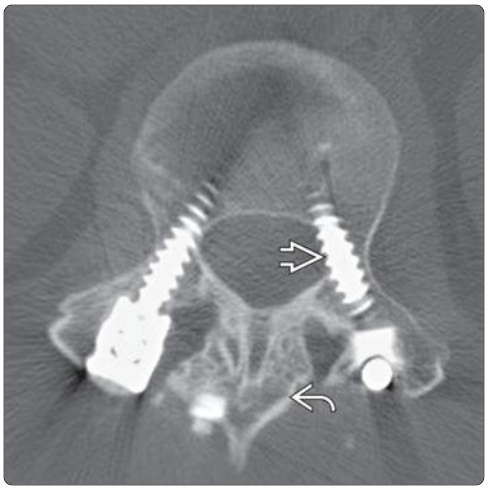
(Left) *Axial T1 MR through a level of prior laminectomy shows soft tissue flattening ventral margin of thecal sac, particularly prominent in the right anterolateral position where appearance is concerning for representing recurrent disc herniation.* **(Right)** *Axial T1 C+ MR in the same patient shows the epidural tissue to enhance; thus, representing peridural fibrosis rather than a herniated disc.*



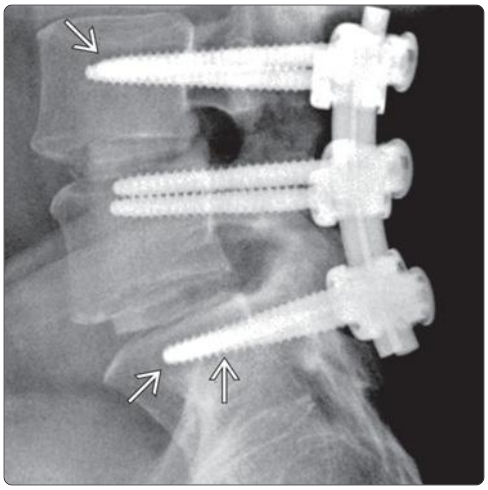
(Left) *Axial T1 C+ FS MR shows pedicle screws and a laminectomy defect. Diffusely increased signal probably reflects the effect of field inhomogeneity generated by the pedicle screws and resulting in poor fat saturation that obscures any potential enhancement due to underlying peridural fibrosis.* **(Right)** *Axial T1 MR shows a defect in the right lamina and ligamentum flavum at the site of a left L5 hemilaminectomy.*





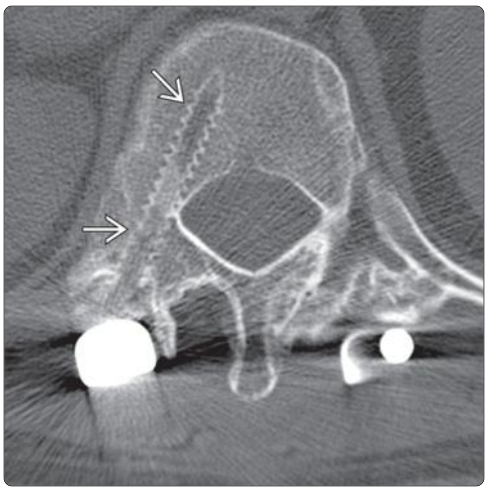


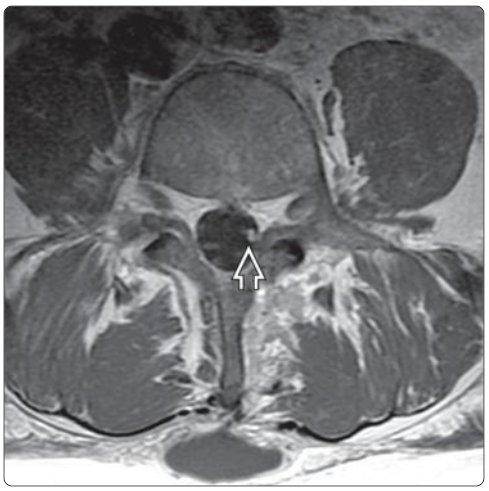
(Left) *Lateral radiograph shows 2-level posterior instrumentation in the lower thoracic spine. Hardware is intact and appears appropriately positioned, with no abnormal clear zones around the pedicle screws.* **(Right)** *Axial NECT in the midlumbar spine shows well-positioned screw on the right. On the left, however, the screw breaks through the medial cortex of the pedicle and likely impinges on the thecal sac. Note the incorporation of a bone graft into the neural arch.*





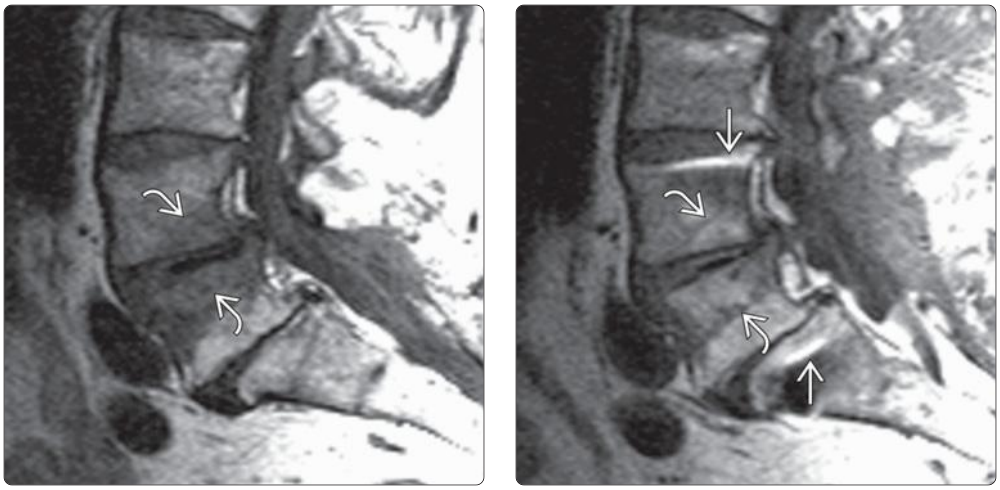
(Left) *Lateral radiograph of a 2-level intervertebral fusion with posterior instrumentation shows lucency around 1 of the L4 and 1 of the sacral screws.* **(Right)** *Axial NECT in the same patient shows the lucency around the left sacral screw. The degree of the lucency decreased on 1-year follow-up films.*



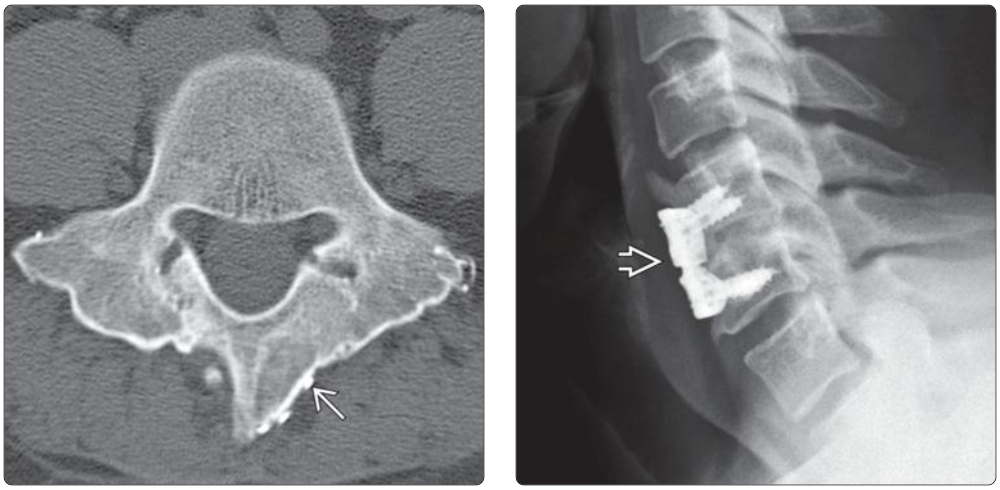


(Left) *Axial NECT shows a linear lucency through the right pedicle and body of L2 that reveals the tract of prior pedicle screw placement; the screw was removed during revision of lumbar spine fixation.* **(Right)** *Axial contrast-enhanced T1 C+ MR following removal of a large left-sided disc extrusion shows enhancement of a single nerve root due to inflammatory radiculitis from disc herniation and surgical manipulation.*

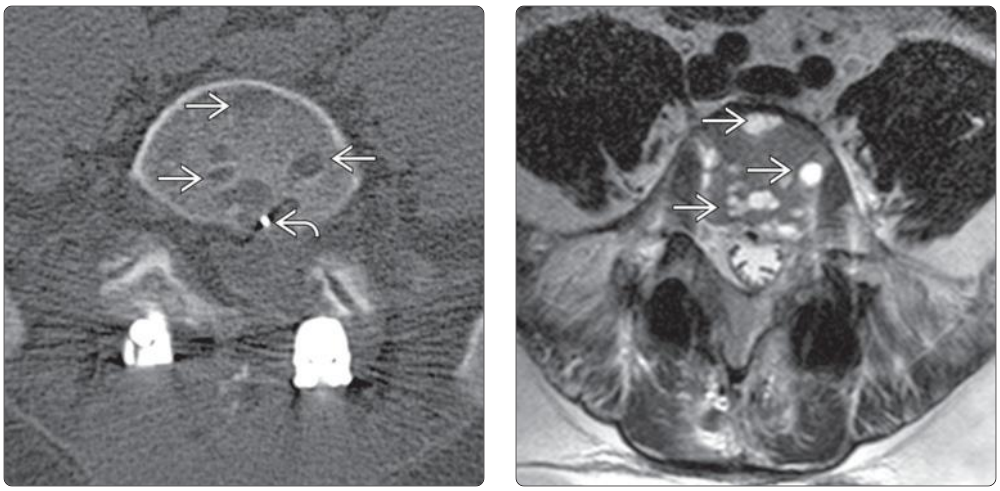
(Left) *Preoperative T1 MR shows type I marrow changes adjacent to degenerative spondylolisthesis of L4-L5.* **(Right)** *Postoperative sagittal T1 MR in the same patient demonstrates conversion of type I to type II marrow adjacent to the L4-L5 disc space. The patient has undergone posterior instrumentation and fusion of L4-S1. Note the metallic artifact from a pedicle screw placement. L4-L5 laminectomy is also seen. The L4-L5 anterolisthesis is stable from the preoperative study.*



(Left) *Axial NECT shows incorporation of bone graft into the posterior elements in this L5-S1 fusion. The metallic wire is from placement of a bone stimulator to promote bone growth.* **(Right)** *Lateral radiograph of the cervical spine shows C5-C6 discectomy and anterior fusion (ADCF) with metallic plate and screw placement. Hardware is intact and appears satisfactorily positioned without evidence of loosening, fracture, or subsidence.*



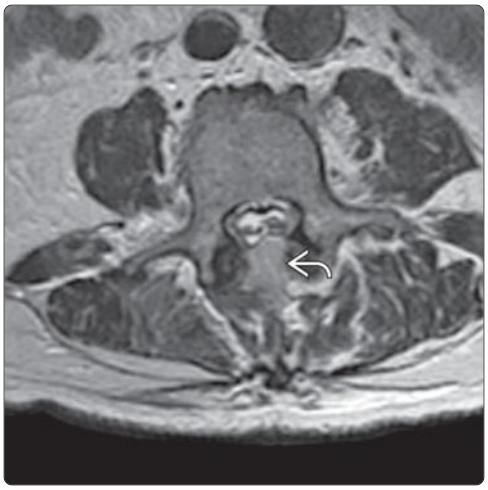
(Left) *Axial NECT following L5-S1 PLIF shows cystic lucencies in the adjacent L5 body due to bony remodeling/resorption from rh-BMP-2. The radiopaque marker denotes the margin of the intervertebral graft.* **(Right)** *Axial T2 MR in the same patient shows "cystic" lesions in the L5 endplates, reflecting osseous resorption/remodeling due to rh-BMP-2 placement for intervertebral fusion. Margins are smooth, and no edema is appreciated in surrounding normal vertebral marrow.*





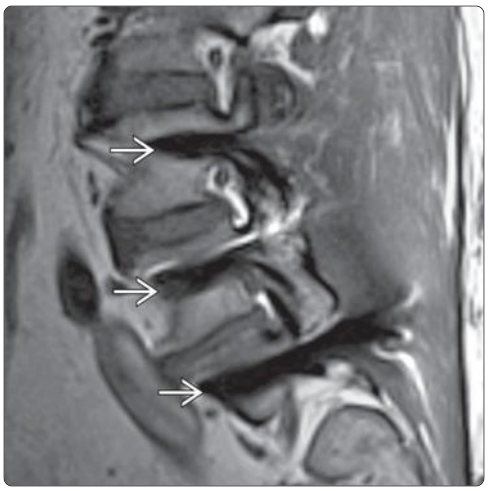


(Left) *Axial T2 FS MR following lumbar laminectomy shows a complex fluid collection in the laminectomy bed, as well as a small amount of subdural fluid. The patient did well postoperatively.* **(Right)** *Sagittal T2 FS MR shows fluid within the L4-L5 laminectomy bed effacing the dorsal thecal sac. A small amount of extraarachnoid fluid tracks cephalad up to the L1 level.*





(Left) *Axial T2 MR shows a fluid collection in the laminectomy bed, effacing the dorsal thecal sac.* **(Right)** *Sagittal T1 C+ MR shows L2-L5 laminectomy with postoperative fluid collection within the surgical bed, effacing the dorsal thecal sac. A small amount of enhancement is seen superiorly. There was no clinical suspicion of infection, and patient did well without further intervention.*





(Left) *Sagittal T1 MR shows screws in L4 and L5 pedicles and in the sacrum. Some geometric distortion is inevitable despite optimized MR technique. Degree of intraforaminal fat around nerve root suggests no more than mild L4-L5 stenosis and more pronounced narrowing with impingement of the nerve root sleeve at L5-S1.* **(Right)** *Sagittal T2* GRE MR shows changes of a C4-C5 discectomy. Ventral extradural defect actually represents metallic susceptibility artifact and was not present on T1WI.*

Postoperative Spinal Complications

KEY FACTS

TERMINOLOGY

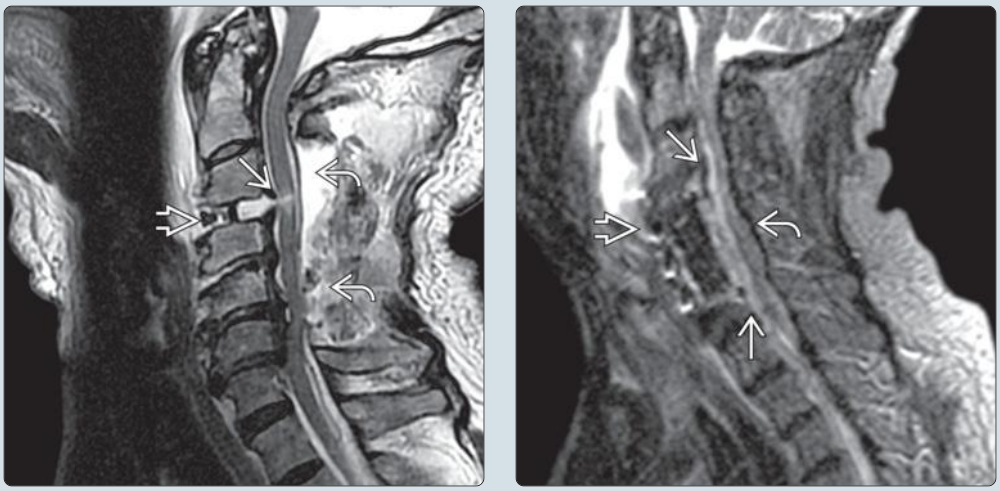
- Cord abnormality, neurologic deficit or hardware failure, or malposition arising as direct or indirect result of prior surgery (including altered biomechanics)
- **Acute**: Multiple diverse etiologies
  - Hemorrhage with cord compression
  - Cord infarction
  - Graft/hardware malposition
  - Spinal anesthesia related
  - Infection
- **Chronic**
  - Accelerated disc degeneration (junctional or transitional disease) with herniation/stenosis
  - Pseudomeningocele/CSF leak
  - Arachnoiditis
  - Peridural fibrosis
  - Recurrent herniation
  - Instability
- **Remote** from surgical site
  - Intracranial hemorrhage
  - Postoperative ischemic optic neuropathy

IMAGING

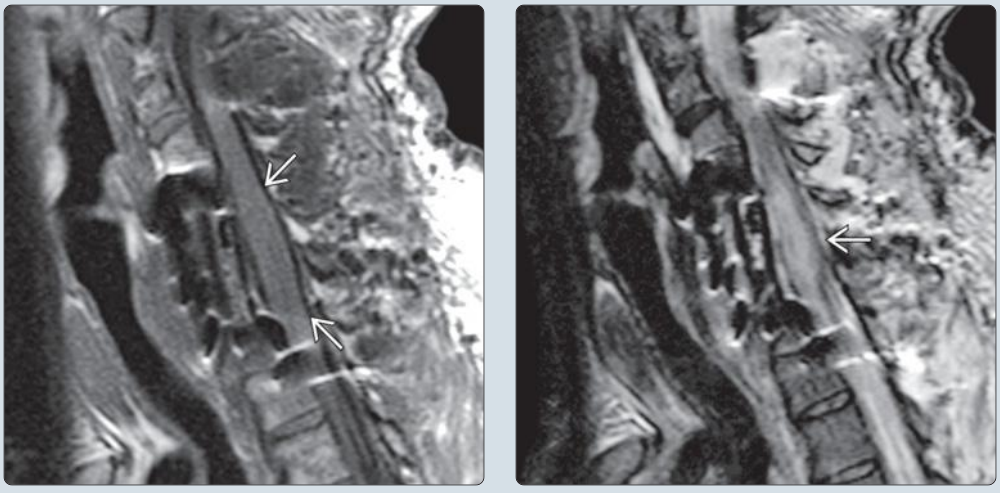
- New abnormality present postoperatively, outside usual scope of postoperative changes
- Infarct
  - Slight cord expansion, decreased signal
- Hemorrhage
  - Isointense to hypointense in acute setting

DIAGNOSTIC CHECKLIST

- MR + CT for definition of soft tissue abnormality (hemorrhage, ischemia) plus bone/graft detail
- Hyperacute hemorrhage may show nonspecific T1WI + T2WI "fluid" signal, defined only by mass effect on neural structures



(Left) *Sagittal T2WI MR shows focal high signal within cervical cord at C3-C4 level due to iatrogenic trauma, which was not present preoperatively. Changes of discectomy and graft placement are seen at C3-C4, and multilevel laminectomy.* **(Right)** *Sagittal T2WI MR shows C4-C7 strut graft placement + anterior plate for spondylosis. Cord compression results from acute epidural hematoma, which shows T2 hyperintensity in the acute phase. Note the posterior displacement of the cord.*



(Left) *Sagittal T1WI MR shows a multilevel corpectomy + strut graft placed for spondylosis. The cord shows fusiform enlargement with slight T1 hypointensity from acute infarction.* **(Right)** *Sagittal T2WI MR shows an anterior corpectomy + strut graft, a posterior multilevel laminectomy. The cord shows fusiform enlargement and T2 hyperintensity from acute infarction.*

TERMINOLOGY

Synonyms

- Iatrogenic complications

Definitions

- Cord abnormality, neurologic deficit or hardware failure, or malposition arising as direct or indirect result of prior surgery (including altered biomechanics)
- **Acute**: Multiple diverse etiologies
  - Hemorrhage with cord compression
    - Epidural
    - Subdural (may extend from posterior fossa surgery site)
    - Intramedullary (rarely due to surgery)
    - Hemosiderosis (chronic sequelae)
  - Cord infarction
    - Aortic aneurysms repair with anterior spinal artery occlusion or embolization
    - Spinal vascular malformation repair/embolization
    - Direct operative cord trauma
    - Secondary to compression from hematoma
    - Hypoperfusion/cardiac tamponade
    - Rare: Hyperextension injury to cord from operative malpositioning of neck
  - Graft/hardware malposition
    - Graft extrusion/displacement with cord compression
    - Hardware (pedicle screw) malposition with cord/thecal sac compression
  - Spinal anesthesia related
    - Catheter-induced hematoma
    - Celiac plexus block (chemical toxicity)
    - Epidural catheter malposition with intracord injection of anesthetic
  - Infection
    - Graft collapse with cord compression
    - Cord compression from abscess
    - Rare: Intramedullary abscess
- **Chronic**
  - Accelerated disc degeneration (junctional or transitional disease) with herniation/stenosis
  - Pseudomeningocele/CSF leak
  - Arachnoiditis
  - Peridural fibrosis
  - Recurrent herniation
  - Instability
    - Altered biomechanics with accelerated degeneration
- **Remote** from surgical site
  - General risks associated with surgery: Risk related to anesthesia, blood loss &/or hypovolemia, myocardial infarction, stroke, pulmonary embolism, deep vein thrombosis, and nosocomial pneumonia
  - Intracranial hemorrhage
    - While rare, when does occur is most commonly involving cerebellar hemispheres
  - Postoperative ischemic optic neuropathy
    - Not an imaging diagnosis

IMAGING

General Features

- Best diagnostic clue
  - New abnormality present postoperatively, outside usual scope of postoperative changes
- Location
  - Cervical, thoracic, or lumbar spine
- Size
  - Variable depending upon etiology
    - Infarction: May involve several cord segments
    - Direct trauma: More focal abnormality
  - Hemorrhage extends over multiple segments
- Morphology
  - Variable depending upon etiology
    - Infarction: Fusiform cord enlargement in acute setting
  - Hemorrhage forms craniocaudal linear signal abnormality

Radiographic Findings

- Radiography
  - Defines hardware position

CT Findings

- NECT
  - MDCT shows alignment, graft position, mass effect on thecal sac/cord

MR Findings

- T1WI
  - Infarct
    - Slight cord expansion, decreased signal
    - Hyperacute infarcts may show normal T1 signal
  - Hemorrhage
    - Isointense to hypointense in acute setting
    - Typical hyperintensity with subacute
- T2WI
  - Infarct
    - T2 hyperintensity in gray matter
    - "Snake eye" pattern of T2 central hyperintensity unfavorable prognostic factor for recovery of upper extremity motor weakness
  - Hemorrhage
    - Isointense to markedly hypointense on T2WI in acute setting
- T2* GRE
  - Infarct
    - May show susceptibility effect from hemorrhage
  - Hemorrhage
    - Acute hemorrhage hypo- to isointense
- T1WI C+
  - Infarct
    - Generally does not acutely enhance
  - Enhanced studies limited usefulness in acute postoperative setting, although may increase conspicuity of postoperative fluid collections
  - Chronic myelomalacia may enhance, linear → focal

Nonvascular Interventions

- Myelography

- Secondary technique to MR for postoperative complication imaging
  - Useful with MR contraindications (aneurysm clip, pacemakers)
  - May show nonspecific cord enlargement with infarct/extradural defect related to epidural hemorrhage

Imaging Recommendations

- Best imaging tool
  - MR
- Protocol advice
  - T1WI, T2WI, GRE show intrinsic cord signal abnormality, cord deformity, epidural collections

DIFFERENTIAL DIAGNOSIS

Epidural Hemorrhage

- Linear mass displacing low signal dura

Subdural Hemorrhage

- Lobulated extramedullary signal abnormality, with well-defined preserved dural peripheral margin

Disc Herniation

- Focal abnormality arising out of parent disc

Graft Extrusion/Malposition

- Plain film/CT quick and accurate for graft position

Abscess

- Subacute peripheral enhancing soft tissue mass, central nonenhancing fluid component

Subdural Effusion

- Benign subdural CSF collection may extend into cervical spine following posterior fossa surgery
- May occur following lumbar puncture/surgery

Cord Tethering

- Focal cord deformity with apposition of cord to dural surface at surgical site
- Cord displacement must be distinguished from mass effect from fluid collection

PATHOLOGY

General Features

- Etiology
  - Postoperative complication
- Infarction: Soft, pale, swollen cord tissue with increasingly distinct margin with more normal tissue over time

Microscopic Features

- Acute infarction: Ischemic neurons with cytotoxic, vasogenic edema, swelling of endothelial cells + astrocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Infarction
    - Anterior spinal syndrome presents with paralysis, loss of pain/temperature sensation, bladder/bowel dysfunction
    - Posterior spinal cord infarction characterized by loss of proprioception/vibration sense; paresis and sphincter dysfunction
  - Epidural postoperative hemorrhage acute symptoms within 24 hours of surgery
    - New focal neurologic deficit
    - Respiratory distress + flaccid quadriparesis/plegia
  - Rare cases of more subacute postoperative hemorrhage 3-5 days after surgery
    - Characterized by severe sharp pain with radiation to extremities
- Clinical profile
  - New neurologic deficit immediately following spinal surgery

Demographics

- Age
  - Child or adult
- Gender
  - M = F
- Epidemiology
  - Postoperative epidural hemorrhage associated with multilevel procedures + preoperative coagulopathy

Natural History & Prognosis

- Acute spinal cord ischemia syndrome has severe prognosis with permanent disabling sequelae
  - Substantial recovery < 20%
- Autonomic dysfunction, pain, paresthesia, and depression are common sequelae and impede recovery

Treatment

- Immediate reoperation for decompression of hematoma or graft malposition/compression
- Infarction without compression: Supportive care

DIAGNOSTIC CHECKLIST

Consider

- MR + CT for definition of soft tissue abnormality (hemorrhage, ischemia) plus bone/graft detail

Image Interpretation Pearls

- Hyperacute hemorrhage may show nonspecific T1WI + T2WI "fluid" signal, defined only by mass effect on neural structures

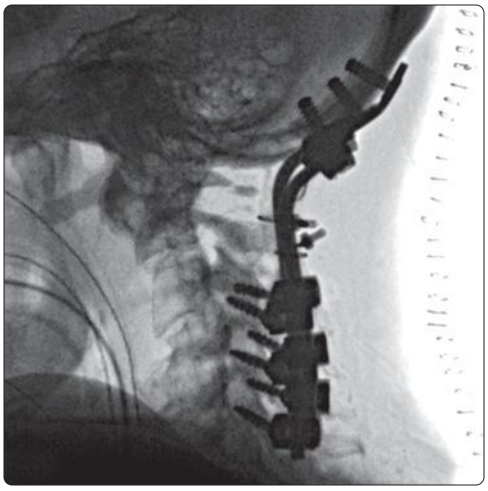
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5. Fayyazi AH et al: Accuracy of computed tomography in assessing thoracic pedicle screw malposition. J Spinal Disord Tech. 17(5):367-71, 2004
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(Left) *Sagittal T2WI MR after thoracic discectomy shows abnormal increased signal from the anterior cord ⇒. Patient is status post posterolateral approach for disc herniation, with unilateral pedicle screw fixation. Presumed mechanism is occlusion of the artery of Adamkiewicz.* **(Right)** *Axial T2WI shows abnormal T2 hyperintensity within the anterior 2/3 of the cord, typical for infarction within the anterior spinal distribution ⇒. Presumed mechanism is occlusion of the artery of Adamkiewicz.*





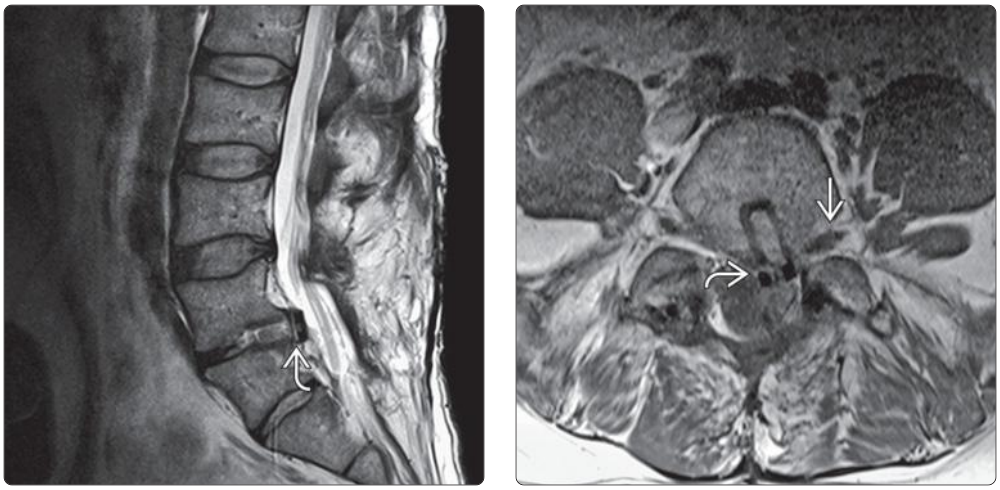
(Left) *Postoperative lateral radiograph after C1-C5 fusion for rheumatoid arthritis with collapse of C1 lateral masses and diffuse subaxial involvement shows satisfactory hardware position, in contrast to grossly abnormal MR.* **(Right)** *Sagittal T2WI shows the metal artifact from the occiput to C5 fusion with large fluid collection at the laminectomy site causing mass effect on the dorsal cord ⇒. Note the cord myelomalacia at the C4 level ⇒. There is relatively little metal artifact given the extensive posterior fusion.*



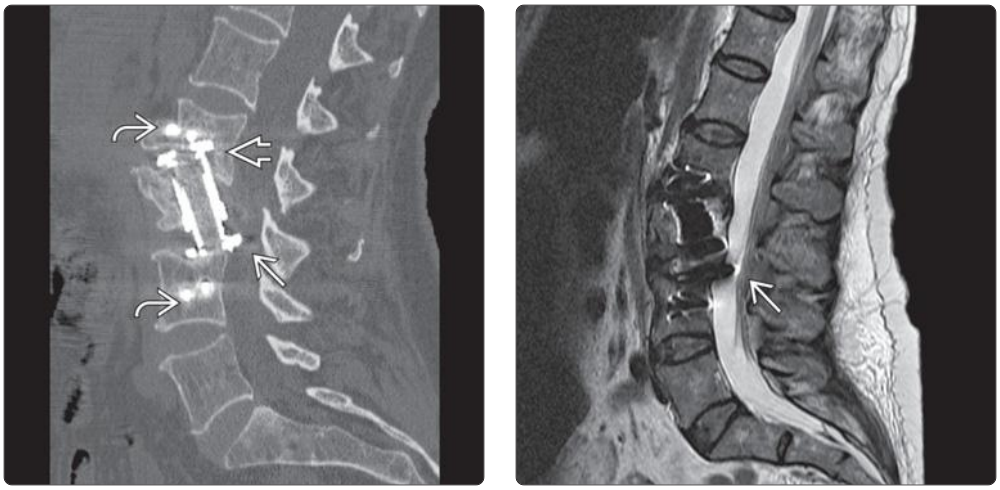


(Left) *Sagittal T1WI MR shows a multilevel laminectomy. Cord is compressed from an epidural hematoma ⇒, causing ventral cord deformity from disc/osteophytes. Note the drainage catheter ⇒.* **(Right)** *Sagittal T2WI MR shows a laminectomy for spondylosis. The dorsal epidural hematoma ⇒ shows T2 hyperintensity compressing the dorsal cord over multiple segments. Note the distinct dural margin ⇒.*

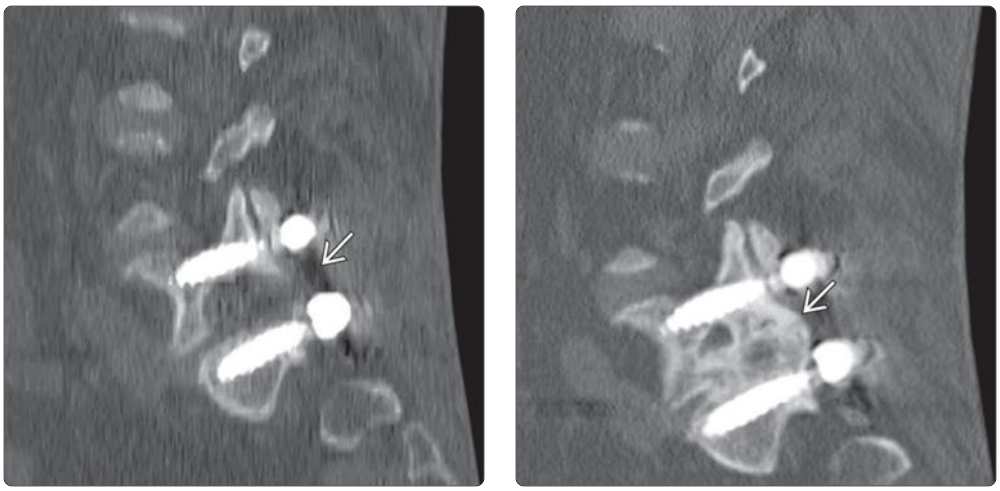
(Left) *Sagittal T2WI in a patient following L4 PLIF shows abnormal posterior positioning of the interbody graft at the L4-L5 level, compressing the ventral thecal sac.* **(Right)** *Axial T1WI in a patient following L4 PLIF shows the position of the posterior margin of the malpositioned graft with respect to the dural margin and the exiting left root.*

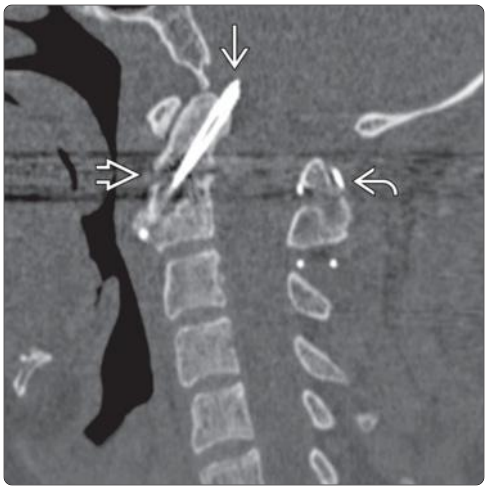


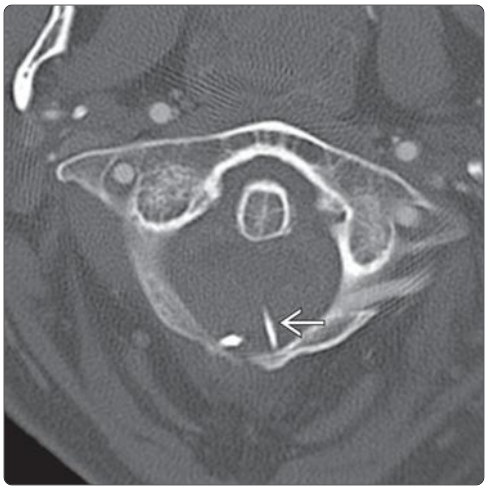
(Left) *Sagittal NECT shows rotation of the L2-L4 fusion cage with subsidence of the superior margin of the cage into the L2 body and posterior displacement of the cage into the ventral epidural space at L3-L4. Vertebral body screws are present from lateral sideplate fixation.* **(Right)** *Sagittal T2WI shows extensive signal loss related to the fusion cage, but suggests abnormal posterior position of the cage due to metal artifact extending into the spinal canal at the inferior L3 level.*



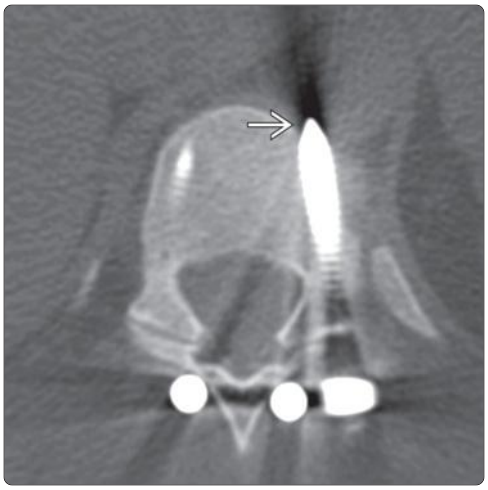
(Left) *Initial sagittal CECT study in this case of heterotopic bone formation shows a large facetectomy defect on the left at L5 with pedicle screw placement. Contrast this with the follow-up CT.* **(Right)** *Sagittal CECT performed 2 years later shows new heterotopic bone filling in the operative defect, which remarkably resembles the facet joint, resulting in moderate bony foraminal stenosis.*





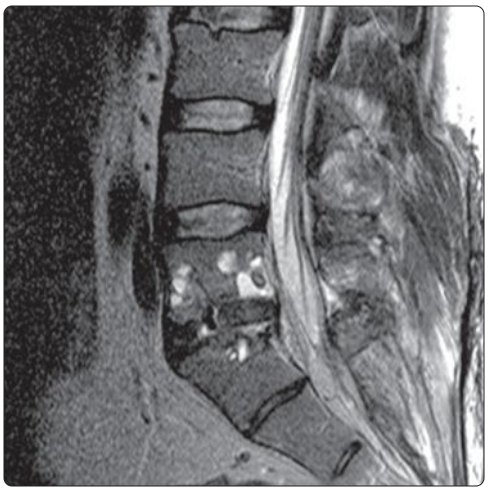


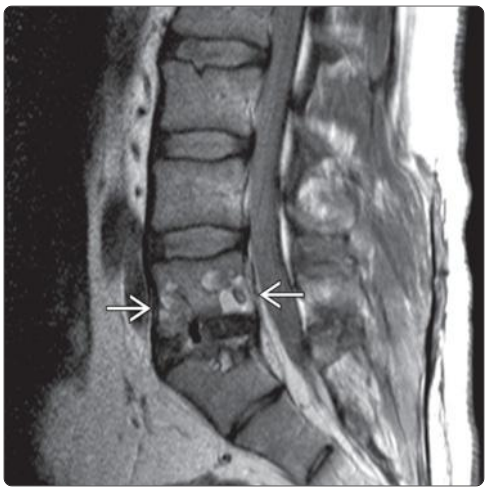
(Left) *Sagittal NECT in this case of a type II odontoid fracture shows extension of the screw tip beyond the confines of the distal odontoid cortex into the premedullary cistern. There has been posterior C1-C2 wiring.* **(Right)** *Axial CECT shows a posterior fixation wire fracture with intradural extension to the dorsal cervical cord. A large subarachnoid hemorrhage was present due to the wire fracture.*





(Left) *Axial NECT shows a left pedicle screw immediately adjacent to the aorta, extending beyond the anterior vertebral body cortical margin. The patient was taken back to surgery, and the screw backed out to appropriate depth.* **(Right)** *AP view of a lumbar myelogram shows an implanted drug delivery pump and an irregular, incomplete myelographic block to the caudal flow of contrast below the L4-L5 level, with an intradural filling defect from arachnoiditis.*





(Left) *Sagittal T2WI MR shows rh-BMP-2 osseous remodeling with vertebral body bone resorption, multiple small cystic-appearing lesions, and T2 hyperintensity involving the L5 and S1 bodies. Note the smooth margins with abrupt transition to more normal marrow signal and lack of adjacent marrow edema.* **(Right)** *Sagittal PD FSE MR in rh-BMP-2 osseous remodeling shows enhancement within several of the lesions, confirming their solid nature. This appearance does not imply failure of fusion.*

Myelography Complications

KEY FACTS

TERMINOLOGY

- Minor complications
  - Postdural puncture headache (most common)
  - Incorrect needle position (subdural, epidural)
  - Transient neurologic sequela
  - Contrast reaction (minor)
- Major complications
  - CSF leak
  - Symptomatic spinal subdural or epidural hemorrhage
  - Contrast reaction (major)
  - Cord injury
  - Intracranial hemorrhage
  - Seizure
  - Paralysis
- Delayed complications
  - Iatrogenic epidermoid (rare)

IMAGING

- Myelography technique
  - Monitor initial contrast injection fluoroscopically to confirm subarachnoid placement
  - Use smaller gauge, atraumatic needles
  - Fluoroscopic guidance for C1-C2 puncture, injection carefully monitored to avoid cord injection
  - Avoid hyperextension during cervical myelography: Rare cause of periprocedural cord injury
- Most common complications have no imaging findings

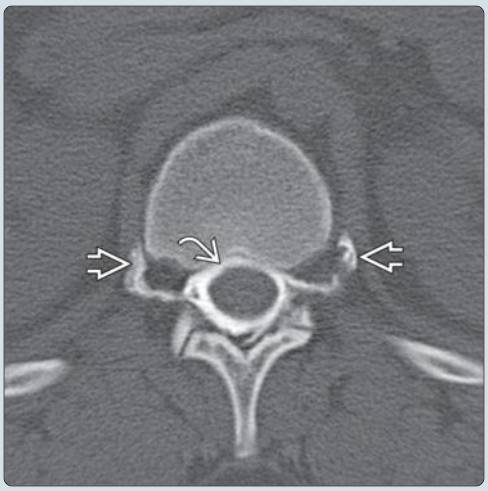
CLINICAL ISSUES

- Dural puncture headache usually begins by 3 days and lasts 3-5 days

DIAGNOSTIC CHECKLIST

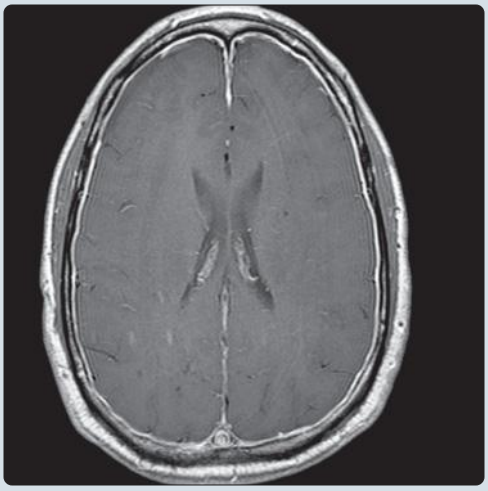
- Use cross-table fluoroscopy to watch initial intrathecal contrast fall away from needle tip into the subarachnoid space





(Left) *Lateral myelography shows inadvertent epidural injection of contrast material. The unopacified thecal sac presents as a tubular filling defect within the contrast column.* **(Right)** *Axial bone CT myelogram in the same patient shows opacification of the epidural space with myelographic contrast. Contrast extends along the extradural nerve roots into the paraspinal tissues. The unopacified thecal sac is seen centrally.*





(Left) *Axial CT myelogram demonstrates extensive subdural contrast material outlining the cord and adjacent suspensory ligaments. Note the outlining of the dentate ligaments extending to the lateral dura by contrast.* **(Right)** *Axial contrast-enhanced T1WI MR of the brain shows diffuse, smooth pachymeningeal thickening and enhancement in this patient with intracranial hypotension following lumbar puncture.*

TERMINOLOGY

Definitions

- Any unexpected sequela following myelography
  - Minor complications
    - Postdural puncture headache (PDPH)
    - Incorrect needle position (subdural, epidural)
    - Transient neurologic sequela
    - Contrast reaction (minor)
  - Major complications
    - CSF leak
    - Symptomatic spinal subdural, epidural, or subarachnoid hemorrhage
    - Contrast reaction (major)
    - Cord injury
    - Intracranial hemorrhage
    - Seizure
    - Paralysis
    - Death
  - Delayed complications
    - Iatrogenic epidermoid

IMAGING

General Features

- Best diagnostic clue
  - Most complications (e.g., headache) have no imaging findings
  - Incorrect injection: Epidural, subdural, or mixed compartment opacification
  - CSF leak: CT myelography, MR myelography, or radionuclide cisternography collection
    - Intracranial hypotension: "Sagging" midbrain, dural thickening, enhancement
  - Aseptic meningitis: ± meningeal enhancement
  - Arachnoiditis: Clumped nerve roots, mild pial enhancement
  - Cord injury: MR cord contusion, hemorrhage
  - Iatrogenic epidermoid: Circumscribed near CSF intensity/density mass

CT Findings

- CECT
  - Iatrogenic epidermoid: Nonenhancing hypodense lesion
- CT myelography
  - Incorrect injection: Most often epidural or subdural
  - CSF leak: ± extravasated contrast at site of leak

MR Findings

- T1WI
  - CSF leak may lead to intracranial hypotension (best seen in sagittal plane)
    - "Sagging" midbrain, loss of basilar cisterns, acquired tonsillar herniation
    - May result in subdural hematoma → tearing of bridging veins occurs after "sagging" brain motion
  - Cord injury: Acute blood isointense, becoming hyperintense with conversion to methemoglobin
  - Iatrogenic epidermoid: Well-defined intradural extramedullary lesion iso- or mildly hyperintense
- T2WI
  - Cord injury: Acutely hyperintense
  - Iatrogenic epidermoid: Well-defined intradural extramedullary lesion iso- or mildly hyperintense to CSF
- STIR
  - Cord injury: Acutely hyperintense
- T1WI C+
  - CSF leak and intracranial hypotension
    - Diffuse, smooth pachymeningeal enhancement
    - Enlarged, enhancing pituitary
    - May involve both brain and spine
  - Aseptic meningitis: May see enhancement
  - Arachnoiditis: Smooth, nodular, or diffuse pial/subarachnoid enhancement
  - Iatrogenic epidermoid: Faint peripheral enhancement possible

Nonvascular Interventions

- Myelography
  - Incorrect injection
    - Most often epidural or mixed subarachnoid/epidural injection
    - Contrast in epidural space may initially simulate contrast in thecal sac if not carefully monitored
  - Replacing needle stylet before withdrawing helps to prevent headache
  - Fluoroscopic guidance for C1-C2 puncture, initial injection carefully monitored to avoid cord injection
  - Avoid hyperextension during cervical myelography
    - Rare cause of periprocedural cord injury

Nuclear Medicine Findings

- Extravasation of intrathecal In-111 DTPA at site of CSF leak

Imaging Recommendations

- Best imaging tool
  - Incorrect injection: CT
  - CSF leak: Radionuclide cisternography, CT myelography, MR myelography
  - Intracranial hypotension, aseptic meningitis, arachnoiditis, iatrogenic epidermoid: MR C+
  - Cord injury: MR with STIR

DIFFERENTIAL DIAGNOSIS

Other Causes of Dural Thickening, Enhancement (Seen With Intracranial Hypotension)

- Septic meningitis
- Carcinomatosis
- Granulomatous disease

Other Causes of CSF Hypotension

- Spontaneous, postoperative, or traumatic CSF leak

Other Causes of Arachnoiditis

- Failed back surgery syndrome
- Lumbar arachnoiditis

Meningitis

- Infection
- Metastases, CSF disseminated

PATHOLOGY

General Features

- Etiology
  - PDPH: Dural puncture allows persistent CSF leak
  - Transient neurologic sequela: Neurotoxic effect of contrast, contaminants, idiosyncratic reaction
  - Cord injury
    - 2/3 due to cervical spine hyperextension and 1/3 after lateral C1-C2 puncture
    - Contributing factors are narrow canal diameter and severe cervical spondylosis
  - Seizure
    - May be potentiated by drugs that lower seizure threshold (antidepressants, psychoactive drugs)
    - Inadvertent intrathecal injection of ionic contrast → ascending tonic-clonic seizure
  - Iatrogenic epidermoid
    - Implantation of squamous cells during spinal puncture, associated with use of unstyletted needle
    - Majority are late complications of spinal puncture during early neonatal period
    - Have been discovered 2-23 years following apparent time of implantation
- Associated abnormalities
  - Subdural, subarachnoid hematoma: Rare
  - Nerve rootlet avulsion: Rare

Microscopic Features

- Iatrogenic epidermoid: Contains keratin, cholesterol crystals, desquamated epithelial cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most common complication: Headache
    - Usually begins within 3 days and lasts 3-5 days
  - Nausea, vomiting, hearing loss, tinnitus, vertigo, dizziness, paresthesias
  - Seizures: Dehydration, cervical myelography increase incidence
  - Visual disturbances, such as diplopia or cortical blindness, have been reported
  - Cranial nerve palsies are not uncommon
  - Transient neurologic sequela rare (e.g., expressive dysphasia)
  - Iatrogenic epidermoid: Progressive radicular and back pain, gait abnormalities, sphincter disturbances

Demographics

- Age
  - PDPH: More common in younger patients
- Epidemiology
  - PDPH
    - Occurs in 36-58% of myelograms performed with 22-gauge Quincke needles
    - Incidence of headache with 22-gauge atraumatic needle is 5% vs. 30% for 20-gauge beveled needles
    - Incidence with 24-gauge atraumatic needle is 1-2% but requires aspiration if CSF is needed
  - Aseptic meningitis: 0.1% of lumbar myelography with iopamidol (Isovue)
  - Iopamidol myelography of nonepileptic patient associated with seizure rate of 0-0.847%
  - Iatrogenic epidermoid
    - Epidermoid cysts are uncommon → 0.5-1% of all spinal tumors; acquired tumors account for ~ 40%
    - Incidence significantly ↓ after nonstyletted needles replaced by tight-fitting stylet needles

Natural History & Prognosis

- PDPH
  - 72% resolve within 7 days, 87% in 6 months
  - To decrease risk
    - Use smaller gauge, atraumatic (blunt) needles
    - Take paramedian approach
    - Maintain parallel orientation of needle bevel to dural fibers
    - Replace stylet before withdrawing
  - IV hydration, no removal of CSF, experience of myelographer, and inpatient status have been inconsistently associated with lower PDPH rates

Treatment

- Contrast reaction: Standard care
- PDPH: Treatment for headache is supportive
  - Nonnarcotic analgesics, hydration, and time
  - Epidural blood patch
    - Rarely necessary, reserved for PDPH persisting > 1 week
    - Very effective → produces remission in 89% of cases, and further 8% relieved by 2nd injection
- CSF leak
  - Blood patch (15-30 cc) is often successful
- Seizure: Supportive care, benzodiazepines, phenytoin
- Iatrogenic epidermoid (rare): Excision is curative

DIAGNOSTIC CHECKLIST

Consider

- Pre-myelography MR screening recommended for patients with canal stenosis, spondylosis, and myelopathy

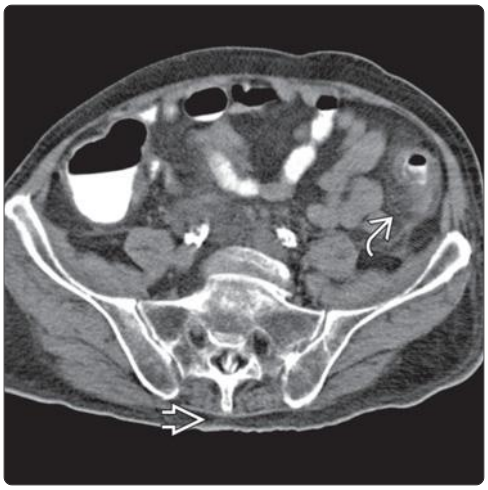
Image Interpretation Pearls

- Use cross-table fluoroscopy to watch initial intrathecal contrast fall away from needle tip into subarachnoid space
- **Always** visually verify type and expiration date of contrast that you are injecting into subarachnoid space

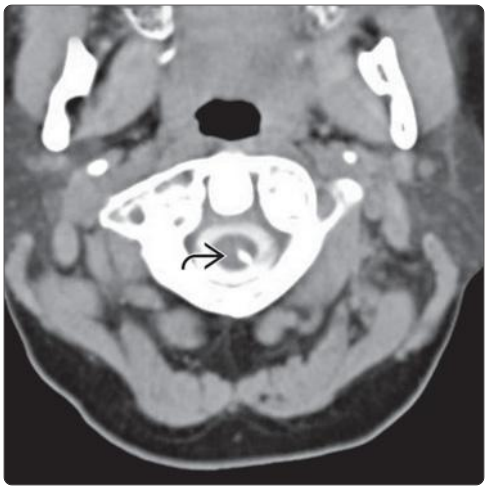
SELECTED REFERENCES

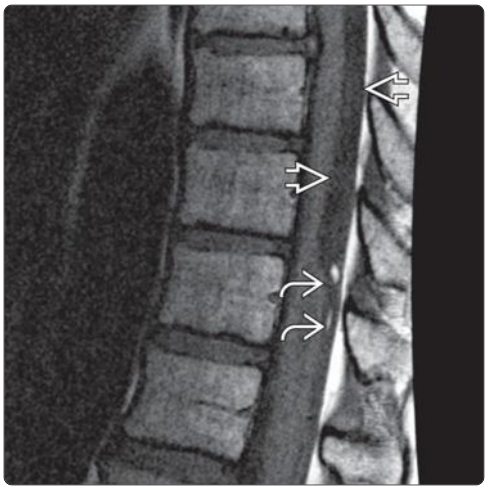
1. Willson MC et al: Postoperative spine complications. Neuroimaging Clin N Am. 24(2):305-26, 2014
2. Chin KR et al: Myelographic evaluation of cervical spondylosis: patient tolerance and complications. J Spinal Disord Tech. 21(5):334-7, 2008
3. Sandow BA et al: Myelography complications and current practice patterns. AJR Am J Roentgenol. 185(3):768-71, 2005





(Left) *Sagittal reformatted image from CT myelography demonstrates a mixed compartment injection; normal intradural contrast is present, as well as a lentiform-shaped subdural contrast collection.* **(Right)** *Axial abdominal NECT of this ICU patient with colitis also shows hyperdense material in the caudal thecal sac secondary to remote iodophenyl undecylic acid (Pantopaque) myelography.*





(Left) *Axial CT myelogram shows contrast in the cervical cord from an inadvertent cord puncture during a lateral C1-C2 injection. Initial placement was subarachnoid. The cord was accidentally punctured during intraprocedural adjustment of needle positioning.* **(Right)** *Sagittal T1WI MR shows arachnoid cyst formation dorsal to the thoracic cord, and secondary to arachnoiditis caused by Pantopaque myelography. Residual Pantopaque is visible as droplets of T1 hyperintensity.*





(Left) *Lumbar myelogram demonstrates a filling defect within the lumbar canal, spreading fibers of the cauda equina, which is due to a lumbar epidermoid cyst.* **(Right)** *Axial T2 MR shows a peripheral low signal involving the distal sac due to severe calcific arachnoiditis from remote Pantopaque myelography. Residual Pantopaque is visible as amorphous hyperintensity.*

Vertebroplasty Complications

KEY FACTS

IMAGING

- Extravasation of cement (into spinal canal, neural foramen, paravertebral spaces, epidural or paravertebral venous plexus) or pulmonary cement embolism
  - Improper needle placement
  - Inadequate intraprocedural monitoring of polymethylmethacrylate (PMMA) injection
  - Inadequate PMMA opacification
  - Low cement viscosity or too large of volume injected
  - Underlying neoplasm with deficient cortex
- New vertebral compression fracture (VCF) adjacent to previously treated level
- Fat embolism
  - Branching or globular hyperdense material in lung parenchyma on fluoroscopy, radiography, or CT
- Vertebral osteomyelitis
  - Osseous destruction adjacent to PMMA

PATHOLOGY

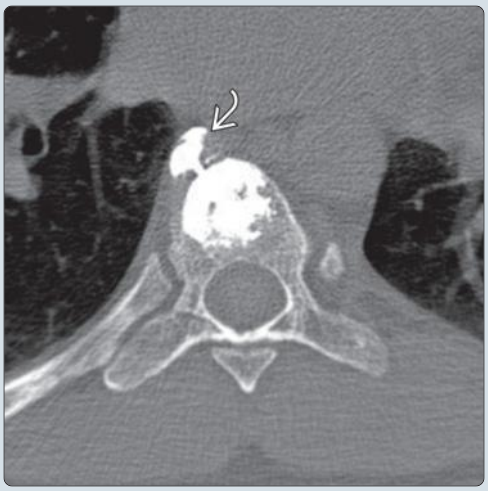
- Adjacent VCF
  - 10-15% of treated patients
  - Controversial relationship to percutaneous vertebroplasty (PVP)
- Fat embolism
  - Embolism of fatty marrow displaced by PMMA injection

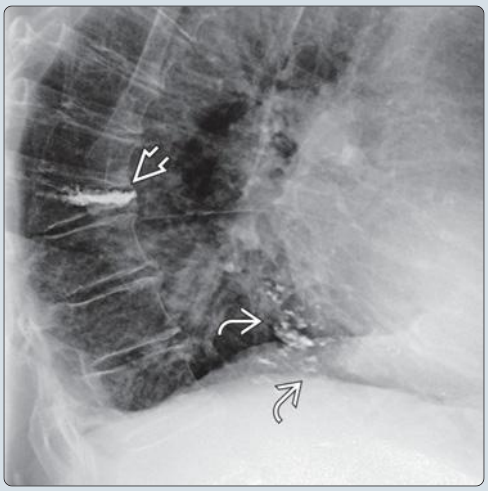
CLINICAL ISSUES

- Clinical presentation variable depending on complication type
- Symptomatic complications of vertebroplasty are rare
  - < 1% for osteoporotic compression fractures
  - 2-5% for treatment of osteolytic metastases

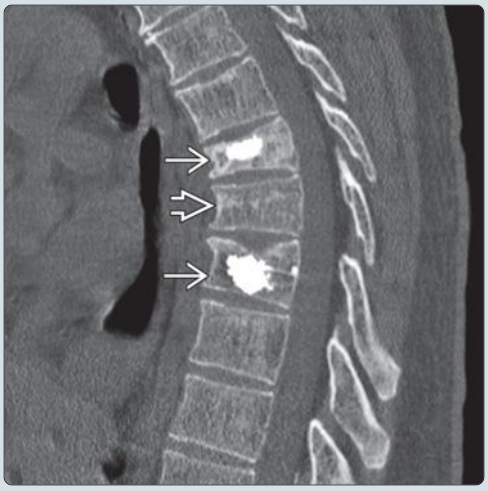
DIAGNOSTIC CHECKLIST

- Consider CT to investigate any unexpected peri- or postprocedural symptomatology





(Left) *Axial NECT shows bone cement extravasating from a lower thoracic vertebral body through a small cortical defect into the paravertebral soft tissues.* **(Right)** *Lateral radiograph shows a compression fracture of a midthoracic vertebral body treated with vertebroplasty. Embolization of cement to the right lower lobe is seen as striated opacities projecting in the retrocardiac space.*





(Left) *Sagittal NECT shows bone cement from prior 2-level vertebroplasty. Imaging for acute back pain shows mild, acute compression fracture of T7 with horizontal sclerosis due to trabecular impaction.* **(Right)** *Sagittal T2WI MR shows high signal surrounding cement several weeks after vertebroplasty in patient on chronic steroid therapy. Aspiration showed Staphylococcus aureus infection. (Courtesy S. Dunnagan, MD.)*

TERMINOLOGY

Definitions

- Vertebroplasty
  - Placement of polymethylmethacrylate (PMMA) into compressed vertebral body to stabilize fracture
- Kyphoplasty
  - Injection of PMMA preceded by balloon inflation within vertebral body
    - Restore vertebral body height
    - Reduce kyphotic deformity
    - Create cavity to facilitate injection of cement under low pressure

IMAGING

General Features

- Best diagnostic clue
  - Extravasation of cement
    - Into spinal canal, neural foramen, paravertebral spaces
      - Cement extending beyond border of vertebral body
    - Into epidural or paravertebral plexus
      - Serpiginous cement, slowly flowing toward central veins on fluoroscopy
  - Pulmonary embolization of cement
    - Branching or globular hyperdense material in lung parenchyma on fluoroscopy, radiography, or CT
  - Adjacent vertebral compression fracture (VCF)
    - New VCF adjacent to previously treated level
  - Vertebral osteomyelitis
    - Osteolysis of vertebral body on delayed radiographs, CT
    - Hyperintense T2, hypointense T1, enhancement signal surrounding cement

Imaging Recommendations

- Best imaging tool
  - CT is most sensitive for defining cement extension beyond vertebral body into canal or foramen, venous plexus, and pulmonary embolization
  - MR best for detecting osteomyelitis, early signs of adjacent VCF
- Protocol advice
  - Extravasation or embolization of cement: Use postprocedural fluoroscopy or plain films
    - Use CT if any question remains or if symptoms present despite negative radiograph
    - Bone or wide soft tissue windows especially useful
    - Use of intrathecal contrast can help prevent cement extravasation when visualization of vertebral body borders suboptimal
  - STIR and pre- and post-contrast T1WI used for detecting osteomyelitis, adjacent VCF

DIFFERENTIAL DIAGNOSIS

DDx of Extravasation of Cement Into Spinal Canal

- Retropulsion of disc/bone fragment into canal
- Ossification of posterior longitudinal ligament

DDx of Extravasation of Cement Into Foramen

- Osteophytic spur or bone fragment in foramen

DDx of Extravasation of Cement Into Perivertebral Venous Plexus

- Phlebolith

DDx of Pulmonary Embolization of Cement

- Calcified granuloma

DDx of Vertebral Osteomyelitis

- Osteonecrosis

PATHOLOGY

General Features

- Etiology
  - Extravasation or embolization of cement
    - Improper needle placement
    - Inadequate intraprocedural monitoring of PMMA injection
    - Low viscosity cement
    - Too large volume of cement injected
    - Underlying neoplasm with deficient vertebral cortex
  - Adjacent VCF
    - Seen in 10-15% of treated patients
    - Controversial relationship to percutaneous vertebroplasty (PVP)
    - May relate to
      - Altered compliance of vertebral body following PMMA injection
      - Increased transmission of forces to adjacent vertebral body
      - Sudden ambulation of previously bedridden patient
    - May merely reflect continued action of same forces that caused initial VCF
      - Osteoporosis
      - Mechanical loading over same segment of spine
  - Fat embolism
    - Embolism of fatty marrow displaced by PMMA injection
    - Likely occurs (to some degree) with all vertebroplasty/kyphoplasty patients
    - Hypotension, circulatory arrest
    - Increased chance of clinically significant fat embolism with multiple levels of treatment, preexisting pulmonary disease
  - Vertebral osteomyelitis
    - Suboptimal sterile technique
    - Omission of prophylactic antibiotic
    - Immunological compromise
    - Active urinary tract infection, other causes of bacteremia

Gross Pathologic & Surgical Features

- Extravasation, embolization of cement
  - Hardened cement beyond vertebral body
    - Variable compromise of spinal canal, neural foramina
  - Occlusion of vessel if embolization
- Adjacent VCF
  - Cortical defect, trabecular impaction
- Fat embolism

- Droplets of fat and spicules of trabecular bone in pulmonary arteries
- Osteomyelitis
  - Typical inflammatory changes, even liquefaction of bone surrounding cement

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Extravasation of cement into canal, neural foramen
    - Most often asymptomatic
    - Back pain, radiculopathy most common symptoms
    - Myelopathy in severe cases
  - Embolization of cement into vein or lungs
    - Most often asymptomatic
    - Shortness of breath, chest pain in minority of pulmonary emboli
    - Compromised pulmonary function rarely seen
    - Cardiopulmonary collapse and death reported
    - Patients with advanced pulmonary compromise not good candidates for aggressive vertebroplasty
  - Adjacent VCF
    - Delayed onset of new back pain
  - Fat embolism
    - Rare complication of vertebroplasty
    - Dyspnea, respiratory insufficiency, circulatory arrest, death
  - Osteomyelitis
    - Delayed onset of back pain
    - Elevated sedimentation rate, C-reactive protein, white cell count
    - Positive blood cultures
    - Positive aspirate or bone biopsy

Demographics

- Epidemiology
  - Symptomatic complications of vertebroplasty are rare
    - < 1% for osteoporotic compression fractures
    - 2-5% for treatment of osteolytic metastases
  - Extravasation of cement
    - Most common complication
    - Seen in up to 20% of cases if looked for
  - Pulmonary embolization
    - Clinically significant embolization rare
    - Probably more common than recognized
    - Fat, displaced by cement placed in marrow, likely embolizes subclinically more often than cement
  - Osteomyelitis
    - Very rare (< 10 reported as of 2009)

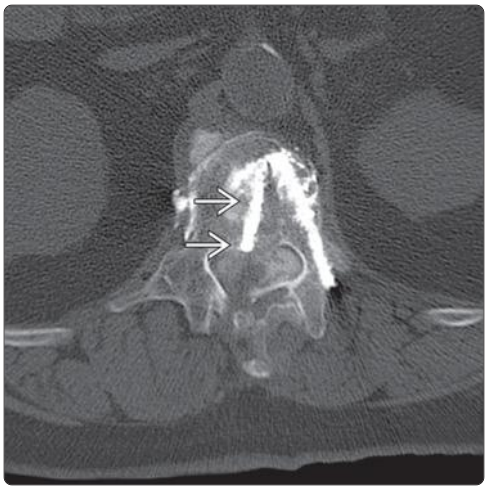
DIAGNOSTIC CHECKLIST

Consider

- CT in any case of unexplained symptomatology following procedure

SELECTED REFERENCES

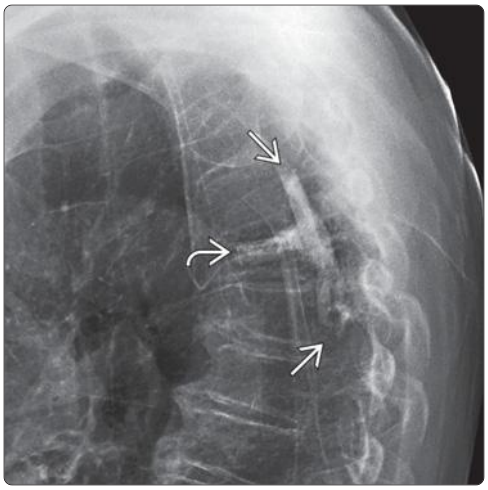
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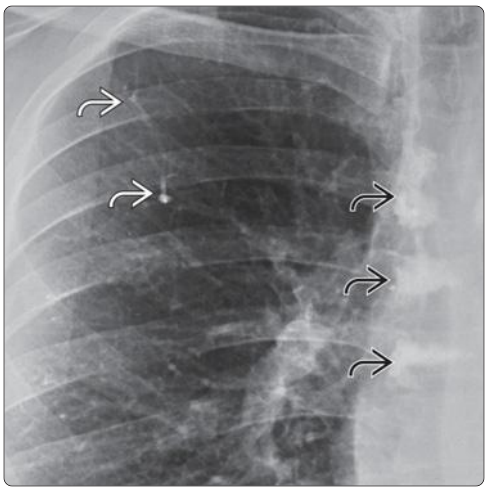
(Left) *CT myelogram shows abnormal trajectory of the vertebroplasty cement in the midportion of the vertebral body, which must have been placed by a malpositioned transdural needle placement very medial to the right pedicle location.* **(Right)** *Axial bone CT shows destruction of the right L2 body and pedicle by osteolytic metastasis. Vertebroplasty has been performed, with extravasation of the cement through a deficient dorsal cortex into the ventral epidural space.*





(Left) *Axial NECT shows extrusion of a large volume of polymethylmethacrylate (PMMA) into the epidural space from vertebroplasty of an upper thoracic compression fracture, resulting in spinal canal stenosis.* **(Right)** *Lateral radiograph in the same patient again shows the rather large volume of PMMA extruded into the thoracic canal from the upper thoracic vertebroplasty site.*





(Left) *Lateral radiograph shows extravasation of bone cement from a treated vertebral compression fracture into the intervertebral disc space.* **(Right)** *Anteroposterior chest x-ray demonstrates serpiginous hyperdense material in the right upper lobe following embolism of bone cement in this patient during multilevel vertebroplasty.*

Failed Back Surgery Syndrome

KEY FACTS

TERMINOLOGY

- Continued low back pain ± radicular pain after lumbar surgery

IMAGING

- Stenosis: "Trefoil" appearance of lumbar spinal canal on axial imaging
- Instability: Deformity increases with motion and increases over time
- Recurrent herniation: Nonenhancing, well-defined mass arising out of intervertebral disc
- Fibrosis: Infiltration of epidural/perineural fat by enhancing soft tissue density (intensity)
- Arachnoiditis: Clumping, adhesion of cauda equina nerve roots

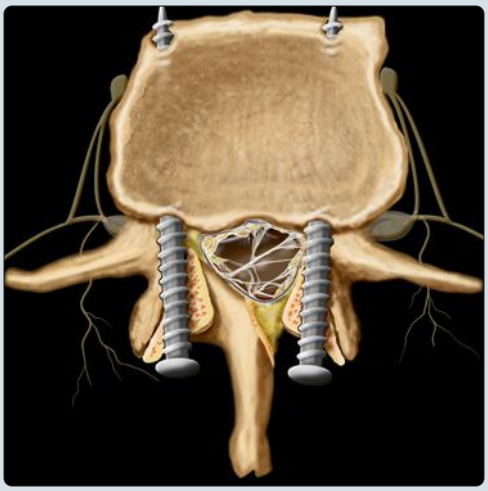
TOP DIFFERENTIAL DIAGNOSES

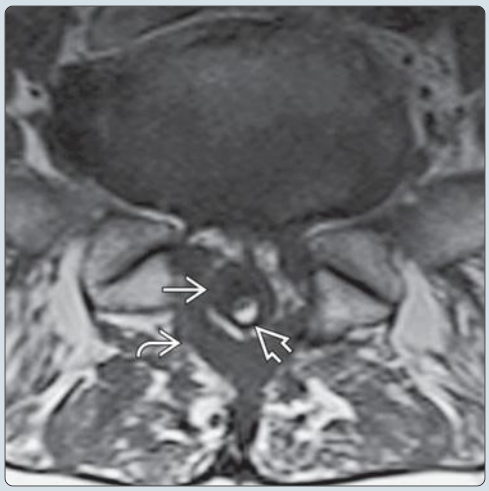
- Infection
  - Endplate destruction, disc T2 hyperintensity
- Tumor
  - Enhancing soft tissue mass
- Hemorrhage
  - Intermediate T1 signal if transition from acute to subacute
- Pseudoarthrosis
  - Abnormal low T1 signal extending through disc, posterior elements, and ligaments

PATHOLOGY

- Multiple underlying etiologies for late failure
  - Foraminal/central stenosis (20-60%)
  - Pseudoarthrosis/instability (14%)
  - Recurrent herniation (7-12%)
  - Epidural fibrosis (5-25%)
  - Arachnoiditis

(Left) *Axial graphic of the lumbar spine shows postoperative change with bilateral pedicle screws and left laminectomy defect. There is a clumping of nerve roots, reflecting arachnoiditis. Screws are too anterior and have breached the anterior vertebral body cortex.* **(Right)** *Axial T1WI MR shows a laminectomy at L5-S1. There is a peripheral clumping of roots in the thecal sac due to arachnoiditis. Note the droplet of T1-hyperintense Pantopaque. Posterior epidural fibrosis is present at the laminectomy site.*





(Left) *Sagittal T1WI C+ MR shows a central clumping of intrathecal roots into a central rope-like mass, simulating a low-lying cord. Note the multilevel laminectomy defect and degenerative disc space enhancement, particularly at L4-L5.* **(Right)** *Axial T2WI MR shows a paucity of nerve roots within the thecal sac at the L5 level, with central clumping consistent with arachnoiditis. Note that the patient has undergone prior laminectomy and posterolateral fusion.*





TERMINOLOGY

Abbreviations

- Failed back surgery syndrome (FBSS)

Definitions

- Continued low back pain ± radicular pain after lumbar surgery

IMAGING

General Features

- Best diagnostic clue
  - Stenosis: "Trefoil" appearance of lumbar spinal canal on axial imaging
  - Instability: Deformity increases with motion and increases over time
  - Recurrent herniation: Nonenhancing, well-defined mass arising out of intervertebral disc
  - Fibrosis: Infiltration of epidural/perineural fat by enhancing soft tissue density (intensity)
  - Arachnoiditis: Clumping, adhesion of cauda equina nerve roots
- Location
  - Lumbar spine
- Size
  - Variable
- Morphology
  - Variable depending upon specific pathology

Radiographic Findings

- Radiography
  - Instability
    - Dynamic slip > 3 mm in flexion/extension
    - Static slip of ≥ 4.5 mm
    - Angulation > 10-15° suggests need for surgical intervention

CT Findings

- NECT
  - Herniation: Ventral epidural soft tissue density extending dorsally into spinal canal contiguous with disc

MR Findings

- T1WI
  - Stenosis: "Trefoil" appearance of spinal canal, thick ligamentum flavum
  - Instability: Anterolisthesis, retrolisthesis, lateral translation
  - Herniation: Isointense to parent disc ventral epidural mass
  - Fibrosis: Intermediate signal on T1WI
  - Arachnoiditis: Peripheral or central clumping of cauda equina roots
- T2WI
  - Stenosis: "Trefoil" appearance of spinal canal, thick ligamentum flavum
  - Instability: Nonspecific loss of disc signal, disc space height
  - Herniation: Isointense to hyperintense ventral epidural lesion
  - Fibrosis: Typically slightly increased in signal relative to disc herniation
  - Arachnoiditis: Peripheral or central clumping of cauda equina
- T1WI C+
  - Stenosis: Crowded ± enhancing nerve roots
  - Instability: Nonspecific enhancement of disc due to degenerative disc disease
  - Herniation: Disc material shows no enhancement, scar will enhance
  - Fibrosis: Immediate, homogeneous post-contrast enhancement
  - Arachnoiditis: Little or no enhancement of roots

Nonvascular Interventions

- Myelography
  - Stenosis: "Hourglass" constriction(s) at single or multiple levels
  - Herniation: Extradural defect at intervertebral disc level

Nuclear Medicine Findings

- Bone scan
  - Nonspecific increased uptake with all degenerative disc disease
- PET
  - Hypometabolic with degenerative endplate changes
  - Hypermetabolic with disc space infection, vertebral osteomyelitis

Imaging Recommendations

- Best imaging tool
  - Flexion/extension plain films
  - MR defines degeneration, endplate changes, stenosis, fibrosis, herniation
- Protocol advice
  - Sagittal, axial T1WI and T2WI; C+ sagittal, axial T1WI

DIFFERENTIAL DIAGNOSIS

Infection

- Endplate destruction, disc T2 hyperintensity
- Abscess shows peripheral enhancement, as do recurrent herniations
- Typical clinical and laboratory findings of infection (elevated sedimentation rate, C-reactive protein)

Tumor

- Enhancing soft tissue mass
- Not arising out of intervertebral disc
- Irregular and infiltrative

Hemorrhage

- Intermediate T1 signal if transition from acute to subacute age

Pseudoarthrosis

- Abnormal low T1 signal extending through disc, posterior elements, and ligaments

PATHOLOGY

General Features

- Etiology

- Multiple underlying etiologies for late failure
  - Foraminal/central stenosis (20-60%)
  - Pseudoarthrosis/instability (14%)
  - Recurrent herniation (7-12%)
  - Epidural fibrosis (5-25%)
  - Arachnoiditis
- Early surgical failure etiologies
  - Postoperative hemorrhage with neural compression
  - Disc space infection, vertebral osteomyelitis, meningitis
  - Dural tear with pseudomeningocele
  - Hardware malposition
  - Wrong level surgery
- Extent of fibrosis possibly related to extent of surgical dissection
- Increased recurrent herniation with larger initial surgical anular defect
- Some degree of FBSS found in 15% of operative patients

- Postoperative scarring is part of normal reparative mechanism
- Herniation = combination of nucleus pulposus, fragmented anulus, cartilage, fragmented apophyseal bone

Staging, Grading, & Classification

- Best reoperative result in patients with herniation at new level away from operation site
- Intermediate operative success results in patients with new herniation at previously operated level
- Only fibrosis is contraindication to reoperation

Gross Pathologic & Surgical Features

- Multiple findings related to specific pathology, such as fibrosis, nonunion, herniation, bony facet arthropathy

Microscopic Features

- Fibrous tissue can extend to vertebral canal, adhere to dura and roots
- Disc material surrounded by granulation tissue, capillaries, macrophages

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Continued low back pain or radiculopathy
  - Weakness, numbness
  - Other signs/symptoms
    - Patients may also be incapacitated by psychiatric, psychologic, social/vocational factors, which relate indirectly to back complaints
- Clinical profile
  - Continued back pain following lumbar discectomy

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - Reintervention rates after lumbar discectomy range from 5-33%
  - Multiple revision: Rate of epidural fibrosis and instability increases to > 60%

Natural History & Prognosis

- Better prognosis
  - Young female
  - History of good results from previous operations
  - Absence of epidural scar
  - Employment before surgery
  - Predominance of radicular (as opposed to axial) pain

Treatment

- Conservative measures
  - Physiotherapy, back-strengthening exercises
  - Transcutaneous electrical nerve stimulation
  - Epidural steroids
- Spinal cord stimulators
- Surgery directed to specific pain generator: Recurrent herniation, foraminal stenosis, instability
  - 34% successful outcome (50% sustained relief of pain for 2 years and patient satisfaction with result)

DIAGNOSTIC CHECKLIST

Consider

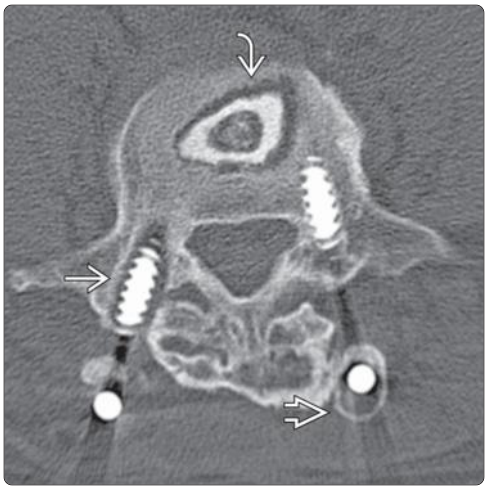
- 30-35% success rate for repeat surgery (range 12-100%)

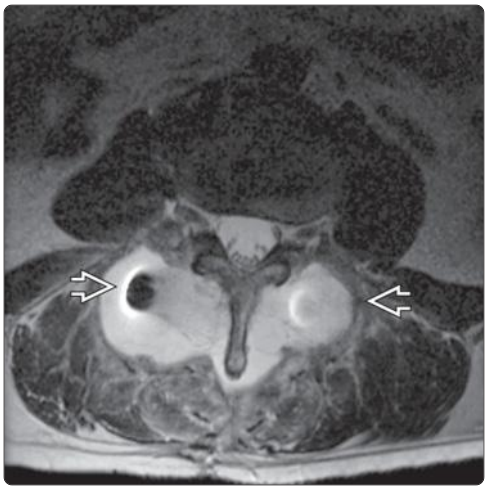
Image Interpretation Pearls

- Contrast-enhanced MR 96-100% accurate in detecting peridural fibrosis vs. recurrent herniation

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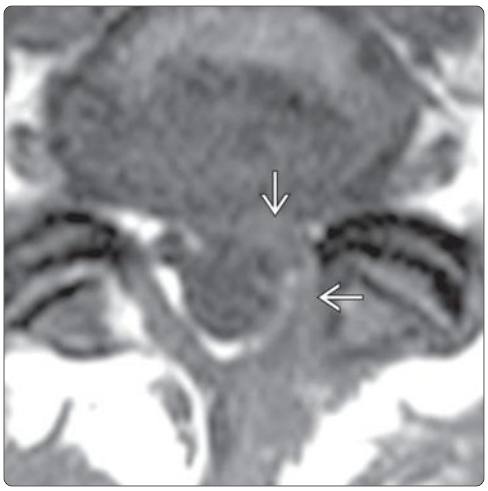
(Left) *Axial NECT shows bilateral loosened pedicle screws with prominent lucency surrounding the mid and distal aspects. There is also lucency and bony remodeling surrounding the junction with the longitudinal rod. Nonunion of the graft spanning L3-L5 is shown with perigraft lucency.* **(Right)** *Axial T2WI MR shows an L4/L5 PLIF with posterior laminectomy and abnormal fluid surrounding the posterior hardware and into the dorsal soft tissues in this postoperative pseudomeningocele.*





(Left) *Sagittal CECT after myelography shows large soft tissue filling defect that merges and engulfs the distal cauda equina, reflecting a chronic inflammatory pseudomass of arachnoiditis.* **(Right)** *Axial T2WI MR shows abnormal posterior positioning of the interbody graft at the L4-L5 level approaching the dural margin and the exiting left S1 root.*





(Left) *Sagittal T2WI MR shows a large recurrent extrusion at the L5-S1 level. There is disc degeneration at L5-S1 with loss of disc height and decreased T2 signal.* **(Right)** *Axial T1WI MR shows homogeneous soft tissue surrounding the ventral and left lateral aspect of the thecal sac with minimal sac deformity from peridural fibrosis.*

Recurrent Disc Herniation

KEY FACTS

TERMINOLOGY

- Recurrent protrusion/extrusion
- Failed back surgery syndrome (FBSS)
- Focal extension of disc material beyond endplate margins at previously operated intervertebral disc level

IMAGING

- Nonenhancing well-defined mass arising out of intervertebral disc
- Disc material shows no enhancement
- May enhance peripherally after intravenous contrast material due to granulation tissue or dilated epidural plexus
- Rare: Diffuse enhancement if associated with granulation tissue or if post-contrast imaging delayed
- Fat suppression of T1WI (pre-/post-gadolinium) may increase sensitivity in detecting peridural fibrosis, differentiating fibrosis from disc

TOP DIFFERENTIAL DIAGNOSES

- Peridural fibrosis
  - Immediate homogeneous post-contrast enhancement
- Hemorrhage
- Abscess
- Osteophyte
- Synovial cyst/ganglion cyst
- Vertebral body/epidural tumor
  - Homogeneous enhancement

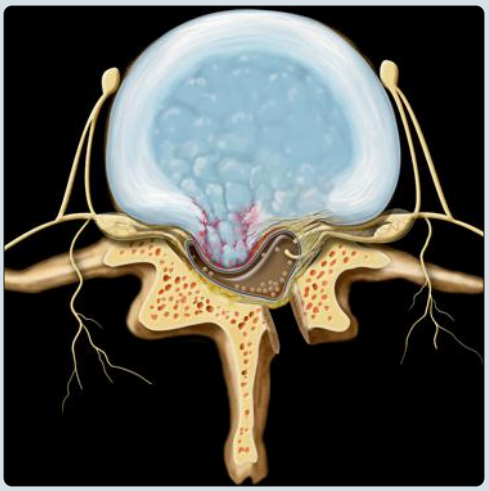
CLINICAL ISSUES

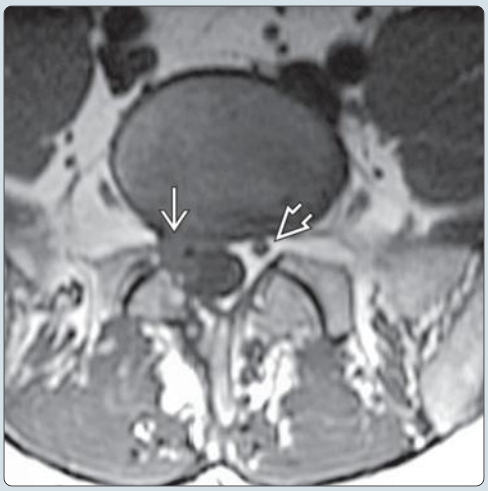
- Revision microdiscectomy with PLIF/TLIF versus revision microdiscectomy
  - 30-35% success rate for repeat surgery (range: 12-100%)

DIAGNOSTIC CHECKLIST

- Best reoperative result in patients with herniation at new level away from operation site

(Left) *Axial graphic of lumbar spine shows left laminotomy defect. Large right paracentral herniation is present, compressing thecal sac and roots adjacent to site of prior discectomy.* **(Right)** *Axial T1WI MR shows right-sided recurrent herniation as a focal epidural mass of intermediate signal with compression of the thecal sac ⇨. Exiting right S1 root is obscured by the herniation. Note the normal position of the left S1 root ⇨.*





(Left) *Axial T2WI MR shows right-sided recurrent extrusion ⇨ as a focal low signal mass displacing right S1 root and right lateral margin of the thecal sac. Note the normal left S1 root ⇨.* **(Right)** *Axial T1WI C+ MR shows typical central lack of enhancement of herniation ⇨, with peripheral enhancement of granulation tissue ⇨.*





TERMINOLOGY

Synonyms

- Failed back surgery syndrome (FBSS)
- Recurrent protrusion/extrusion

Definitions

- Focal extension of disc material beyond endplate margins at previously operated intervertebral disc level

IMAGING

General Features

- Best diagnostic clue
  - Nonenhancing well-defined mass arising out of intervertebral disc
- Location
  - Primarily lumbar disc levels
- Size
  - Varies from few mm to > 10 mm
- Morphology
  - Round, smooth-margined soft tissue
  - Protrusion
    - Protrusion is herniated disc with broad base at parent disc
    - Greatest diameter of protrusion in any plane < distance between edges of base in same plane
    - Focal: < 25% of disc circumference
  - Extrusion
    - Extrusion is herniated disc with narrow or no base at parent disc
    - Greatest diameter of extrusion in any plane > distance between edges of base in same plane
    - Sequestered or free fragment: Extruded disc without contiguity to parent disc
    - Migrated: Disc material displaced away from site of herniation, regardless of continuity to parent disc

Radiographic Findings

- Radiography
  - Plain film shows postoperative laminectomy site; not useful for disc herniation detection

CT Findings

- NECT
  - Ventral epidural soft tissue density extending dorsally into spinal canal contiguous with disc
    - Changes of prior discectomy/laminectomy
- CECT
  - Herniation seen as soft tissue attenuation mass at disc level, ± peripheral enhancement related to fibrosis or granulation tissue
  - Replaced by enhanced MR as primary diagnostic modality

MR Findings

- T1WI
  - Isointense to parent disc
  - May show low signal if calcified, or associated with vacuum phenomenon
- T2WI
  - Isointense to hyperintense
- T1WI C+
  - Disc material shows no enhancement
  - May enhance peripherally after intravenous contrast material due to granulation tissue or dilated epidural plexus
  - Rare: Diffuse enhancement if associated with granulation tissue or if post-contrast imaging delayed

Nonvascular Interventions

- Myelography
  - Shows nonspecific extradural defect in contrast column centered at disc level
    - Root sleeves effaced or appear "cut off"
    - May fail to show L5-S1 herniation due to large epidural space

Imaging Recommendations

- Best imaging tool
  - Fat suppression of T1WI (pre-/post-gadolinium) may increase sensitivity in detecting peridural fibrosis, differentiating fibrosis from disc
- Protocol advice
  - Sagittal, axial T1WI/T2WI; C+ sagittal, axial T1WI

DIFFERENTIAL DIAGNOSIS

Peridural Fibrosis

- Immediate homogeneous post-contrast enhancement
- Enhancement regardless of time elapsed since surgery
- Can be associated with ipsilateral enlargement of exiting root sleeve (cicatrization)

Hemorrhage

- Intermediate T1 signal if acute to subacute age
- Low T2 signal mass within epidural space at operative site

Abscess

- May show homogeneous enhancement within epidural space
- Abscess shows peripheral enhancement as do recurrent herniations
- Typical clinical/laboratory findings of infection (elevated sedimentation rate, C-reactive protein)

Osteophyte

- Sharp margins not arising directly from intervertebral disc level
- May show increased signal on T1WI due to fatty marrow content

Synovial Cyst/Ganglion Cyst

- Extension of synovial cyst lateral to thecal sac may mimic ventral epidural process
- Central T2 hyperintensity, with peripheral enhancement

Vertebral Body/Epidural Tumor

- Homogeneous enhancement
- Not arising out of intervertebral disc
- Irregular and infiltrative

PATHOLOGY

General Features

- Etiology

- Increased recurrent herniation with larger initial surgical anular defect
- Associated abnormalities
  - Evidence of prior surgical intervention
    - Fusion, peridural fibrosis, laminectomy defect, pseudomeningocele, arachnoiditis
- Composed of combination of nucleus pulposus, fragmented anulus, cartilage, fragmented apophyseal bone
- Disc material may be intermixed with granulation tissue/fibrosis

Staging, Grading, & Classification

- 3 categories of postoperative disc/fibrosis
  - Recurrent herniation ± peridural fibrosis
  - Herniation at new level
  - Peridural fibrosis only

Gross Pathologic & Surgical Features

- Nucleus pulposus
  - Gelatinous material with high water content, few collagen fibers
- Anulus fibrosus
  - Fibrocartilage, with collagen fibers in concentric lamellae

Microscopic Features

- Disc material surrounded by granulation tissue, capillaries, macrophages

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back pain or radiculopathy
  - Other signs/symptoms
    - Weakness, numbness
- Clinical profile
  - Adult with new sudden onset low back pain/radiculopathy following initially successful disc surgery

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - Estimated 1 million spinal operations worldwide each year
  - Recurrent disc herniation implicated in FBSS in 7-12%
  - Recurrent herniation in 12% of cases, median of 8 months after primary discectomy

Natural History & Prognosis

- FBSS complex, is multifactorial
- 7-year follow-up following microdiscectomy: 25% free of pain, 66% marked improvement, 9% had no improvement/worsening of pain
  - Of those that did not improve
    - 15% required changing profession following discectomy
    - 6% were incapacitated and unable to work
    - 14% were forced into early retirement

Treatment

- Conservative treatment
  - Exercise and physical conditioning
  - Nonsteroidal anti-inflammatory/opioid drugs
  - Epidural steroid injection
- Revision microdiscectomy with PLIF/TLIF versus revision microdiscectomy
  - 30-35% success rate for repeat surgery (range: 12-100%)

DIAGNOSTIC CHECKLIST

Consider

- Best reoperative result in patients with herniation at new level away from operation site
- Intermediate operative success results in patients with new herniation at previously operated level
  - Better results than surgery on scar only with no disc
  - Worse results than surgery on disc at new level
- Identification of only epidural fibrosis in FBSS patient is contraindication to reoperation, yields poor reoperative result

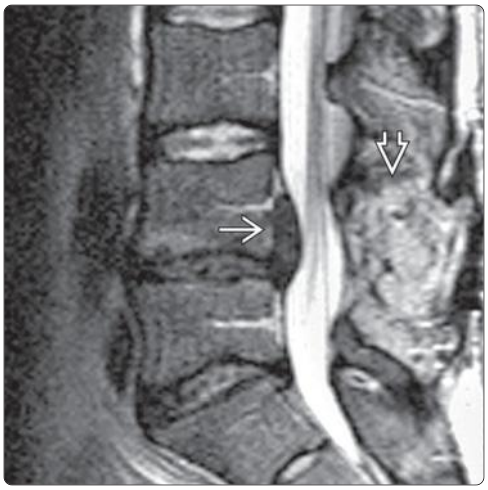
Image Interpretation Pearls

- Immediate post-contrast MR 96-100% accurate detecting peridural fibrosis vs. recurrent herniation

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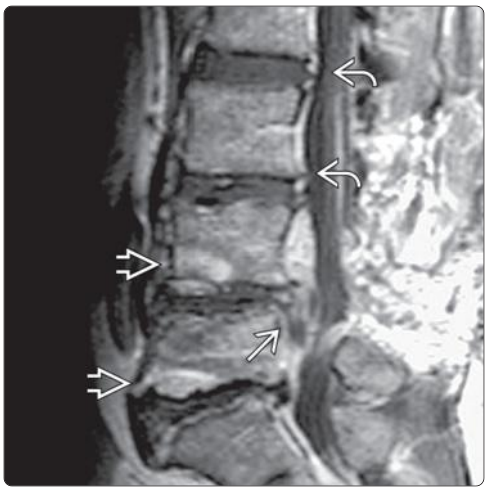
(Left) *Sagittal T1WI C+ MR shows recurrent disc extrusion as a centrally nonenhancing mass with peripheral enhancing granulation tissue, migrating superiorly from the L4-L5 level. Findings are suspicious for free fragment. Note fatty degenerative endplate changes at L4-L5.* **(Right)** *Sagittal T2WI MR shows large recurrent extrusion (free fragment) migrating cephalad from the L4-L5 parent disc level. Note the posterior laminectomy defect.*





(Left) *Sagittal STIR MR in this patient status post L5 discectomy shows a large recurrent herniation at L5-S1 displacing the exiting S1 root. Herniation shows well-defined margins and is slightly hyperintense.* **(Right)** *Sagittal T1WI C+ FS MR shows peripheral enhancement surrounding the large nonenhancing herniation due to granulation tissue and epidural plexus.*





(Left) *Sagittal T2WI MR shows a large recurrent L4-L5 extrusion effacing the thecal sac and migrating inferiorly behind the L5 body. There is extensive disc degeneration at L3-L4 through L5-S1.* **(Right)** *Sagittal T1WI C+ MR shows recurrent nonenhancing herniation mixed with enhancing fibrosis at L4-L5. Severe multilevel disc degeneration is present with type II degenerative endplates and multiple anular fissures.*

Peridural Fibrosis

KEY FACTS

TERMINOLOGY

- Scar formation within epidural space after lumbar surgery
- Part of failed back surgery syndrome (FBSS)

IMAGING

- Infiltration of epidural/perineural fat by enhancing soft tissue density (intensity)
- Variable signal intensity on T2WI
- Immediate homogeneous post-contrast enhancement

TOP DIFFERENTIAL DIAGNOSES

- Recurrent disc herniation
- Epidural abscess/phlegmon
- Pseudomeningocele
- Postoperative hemorrhage
- Arachnoiditis

PATHOLOGY

- Postoperative scarring is part of normal reparative mechanism
- May be asymptomatic; contribution to clinical symptoms controversial
- Up to 1/4 of all FBSS cases
- Most patients with some degree of fibrosis are asymptomatic

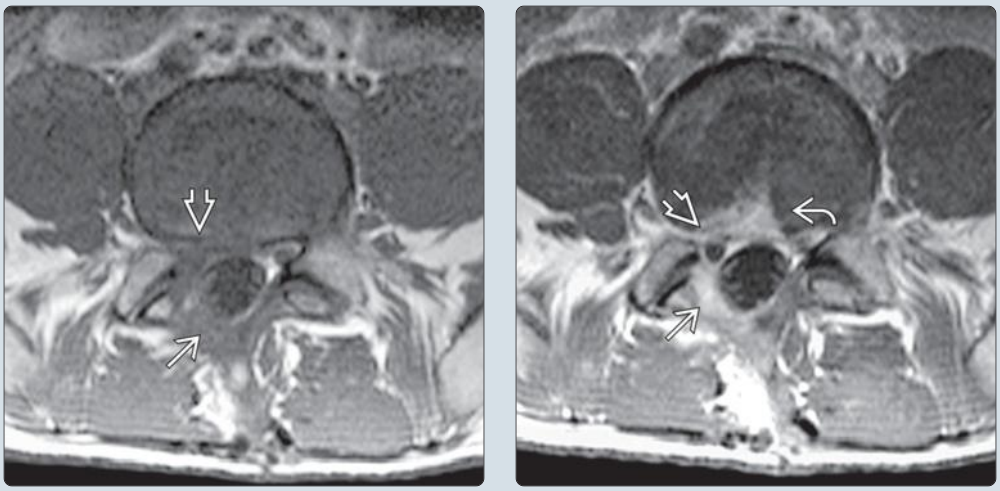
CLINICAL ISSUES

- Adult with gradual onset low back pain following initially successful disc surgery

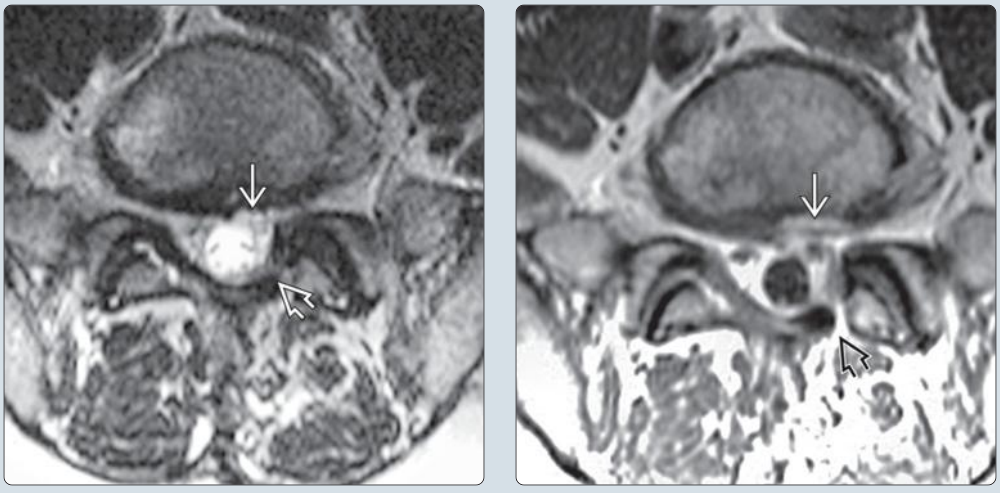
DIAGNOSTIC CHECKLIST

- Identification of only epidural fibrosis in FBSS patient is contraindication to reoperation, yields poor reoperative result

(Left) *Axial T1WI MR shows a right laminectomy defect, and extensive epidural fibrosis ➔ surrounding the right lateral and dorsal aspect of the thecal sac and the exiting root ➔.* **(Right)** *Axial T1WI C+ MR shows a right hemilaminectomy defect and diffuse enhancement of epidural fibrosis surrounding the right lateral aspect of the thecal sac ➔. Nerve root is seen as a nonenhancing structure within the fibrosis ➔. Enhancement continues into operative defect in the disc ➔.*



(Left) *Axial T2WI MR a shows hemilaminectomy defect and absence of ligamentum flavum ➔. Epidural fibrosis surrounding the left S1 root shows slight increased signal relative to adjacent disc material ➔.* **(Right)** *Axial T1WI C+ MR shows left hemilaminectomy defect ➔. A small amount of epidural fibrosis surrounds the exiting left S1 root, which diffusely enhances ➔, as does operative defect in posterior anulus.*



TERMINOLOGY

Synonyms

- Epidural fibrosis, epidural scar

Definitions

- Scar formation within epidural space after lumbar surgery
- Part of failed back surgery syndrome (FBSS)

IMAGING

General Features

- Best diagnostic clue
  - Infiltration of epidural/perineural fat by enhancing soft tissue density (intensity)
- Location
  - Epidural space at operative level
- Size
  - Few mm to 1-2 cm
- Morphology
  - Smooth marginated soft tissue, usually without mass effect

Radiographic Findings

- Radiography
  - Nonspecific postoperative changes, not directly visualized

CT Findings

- NECT
  - Nonspecific epidural soft tissue attenuation
- CECT
  - Epidural soft tissue density
    - Enhances after intravenous contrast

MR Findings

- T1WI
  - Peridural soft tissue intensity
    - Isointense
    - Often surrounds nerve root
    - Occasionally can be mass-like
  - Combination of disc + scar may be present
  - Postsurgical changes in posterior elements
  - Can be associated with ipsilateral enlargement of exiting root sleeve (cicatrization)
- T2WI
  - Variable signal intensity
  - Typically slightly increased in signal relative to disc herniation
- T1WI C+
  - Immediate homogeneous post-contrast enhancement
    - Enhancement may persist for years after
    - Pre- and post-gadolinium-enhanced MR approximately 96% accurate in differentiating peridural fibrosis from disc herniation
  - Involved nerve root may show post-gadolinium enhancement

Nonvascular Interventions

- Myelography
  - Nonspecific extradural defect on contrast column

Imaging Recommendations

- Fat suppression of T1WI (pre- and post-gadolinium) may increase sensitivity in detecting peridural fibrosis and differentiating fibrosis from disc

DIFFERENTIAL DIAGNOSIS

Recurrent Disc Herniation

- No central enhancement when imaged early after intravenous gadolinium
  - Peripheral enhancement common
- Delayed central enhancement if imaged after 30 minutes or later
  - Diffusion of contrast into disc

Epidural Abscess/Phlegmon

- May show homogeneous enhancement within epidural space
- Abscess shows peripheral enhancement
- Typical clinical and laboratory findings of infection (elevated sedimentation rate, C-reactive protein)

Pseudomeningocele

- CSF collection within dorsal soft tissues arising out of surgical bed

Postoperative Hemorrhage

- Intermediate T1 signal if acute to subacute age
- Low T2 signal mass within epidural space at operative site
- Mass effect upon thecal sac/roots

Arachnoiditis

- Peripheral intradural clumping of roots in type II arachnoiditis may extend contiguous to epidural fibrosis
- May mimic epidural disease

PATHOLOGY

General Features

- Etiology
  - Extent of fibrosis possibly related to extent of surgical dissection
  - Degree of host inflammatory response also plays role
  - Scar tissue compresses, irritates, puts abnormal traction on nerve roots
    - Blood supply compromised
    - Axoplasmic transport interrupted
- Genetics
  - None
- Associated abnormalities
  - Look for evidence of arachnoiditis
- Postoperative scarring is part of normal reparative mechanism
- May be asymptomatic; contribution to clinical symptoms controversial
- Peridural fibrosis
  - Up to 1/4 of all FBSS cases
  - Most patients with some degree of fibrosis are asymptomatic
  - Still controversial whether a source of recurrent pain

- 1 prospective study (1996) shows that patients with extensive peridural fibrosis are 3.2x more likely to have recurrent radicular pain than those with less extensive scarring; other studies show no association (2008)

Gross Pathologic & Surgical Features

- Scar tissue surrounding thecal sac and exiting roots at operative level

Microscopic Features

- Invasion of postoperative hematoma by dense fibrous tissue originating within deep surface of paravertebral musculature
- Fibrous tissue can extend to thecal sac, adhere to dura and roots

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back or radicular pain
  - Numbness
  - Weakness
  - 10% rate of recurrent back/radicular pain 6 months after discectomy
  - Causes of recurrent pain
    - Recurrent disc herniation
    - New herniation at another level
    - Peridural fibrosis
- Clinical profile
  - Adult with gradual onset low back pain following initially successful disc surgery

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - Estimated 1 million spinal operations worldwide each year
    - 3/4 involve decompression of lumbar roots or cauda equina
    - Peridural fibrosis implicated in 3-5% of cases as cause of FBSS

Natural History & Prognosis

- Chronic, indolent course of FBSS
- 30-35% success rate for repeat surgery (range: 12-100%)
- Worse reoperative success rates for only epidural fibrosis found at surgery, without disc herniation
- 50-70% success rate for spinal cord stimulation

Treatment

- Symptomatic treatment with physical therapy, wide variety of pain medication
- Periradicular injection of corticosteroids and local anesthetics
- Spinal cord stimulation by implanted electrodes
- Surgical lysis of scar tissue rarely performed

DIAGNOSTIC CHECKLIST

Consider

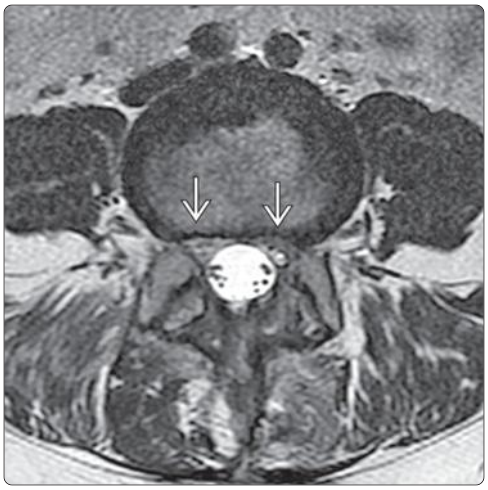
- Identification of only epidural fibrosis in FBSS patient is contraindication to reoperation, yields poor reoperative result
- Best reoperative result in patients with herniation at new level
- Intermediate operative success results in patients with new herniation at previously operated level
  - Results better than re-op on scar only, worse results than if re-op on disc at nonoperative level

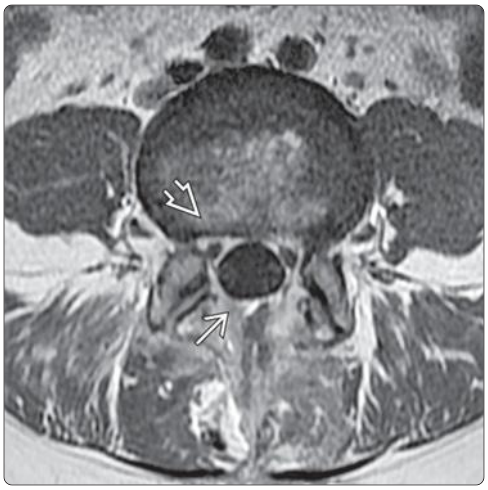
Image Interpretation Pearls

- Classic imaging appearance: Enhancing epidural soft tissue surrounding typically enlarged nerve root
- Primary function of imaging is to exclude recurrent disc herniation or other cause of FBSS (e.g., conus/filum tumor)

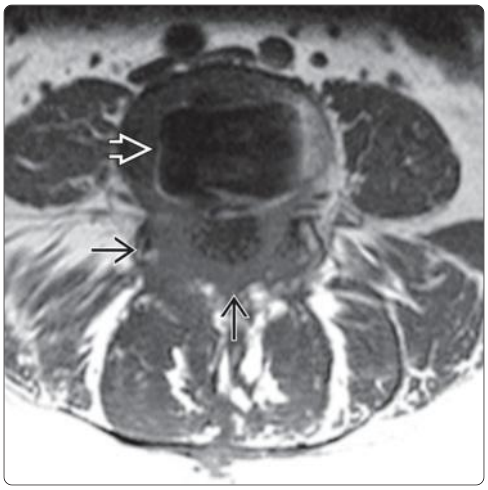
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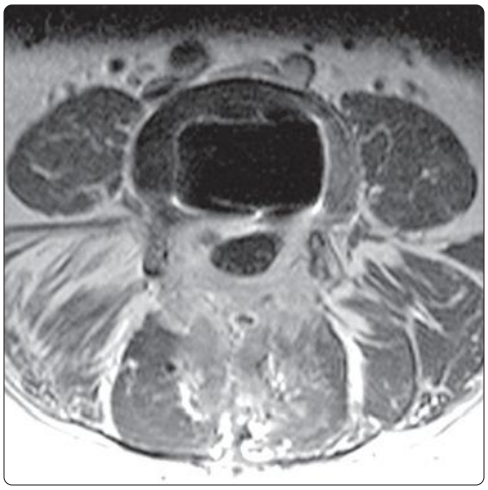
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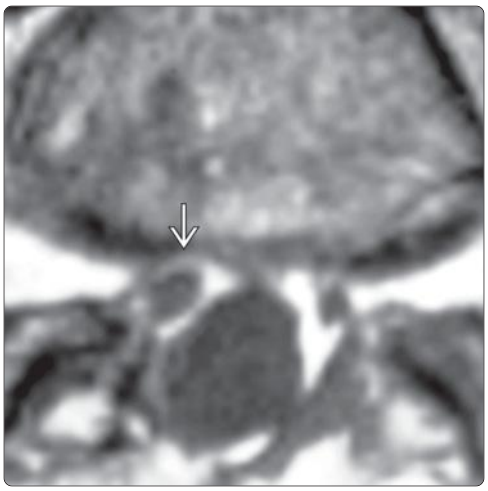


(Left) *Axial T2WI MR shows small bilateral laminectomy defects, with obscuration of the ventral epidural fat by isointense soft tissue ⇨. There is no mass effect on the thecal sac and no evidence of herniation.* **(Right)** *Axial T1WI C+ MR shows enhancement of the soft tissues at the laminectomy site ⇨ and diffuse mild enhancement of the ventral epidural space and surrounding the exiting roots ⇨.*





(Left) *Axial T1WI MR in a patient following posterior lumbar interbody fusion (PLIF) shows extensive low signal surrounding the thecal sac from peridural fibrosis ⇨. Artifact is present in disc space from fusion cages ⇨.* **(Right)** *Axial T1WI C+ MR in the same patient following posterior lumbar interbody fusion (PLIF) shows diffuse enhancement of extensive epidural fibrosis with minimal thecal sac deformity. Exiting roots are subsumed within the peridural fibrosis.*





(Left) *Axial T1 C+ MR shows a right hemilaminectomy defect and enlargement of the exiting root sleeve on right, with peripheral enhancement due to scar cicatrization ⇨.* **(Right)** *Axial T1WI MR shows a small left hemilaminectomy site ⇨. Soft tissue signal surrounds the exiting left S1 root, without mass effect upon the adjacent thecal sac ⇨.*

Arachnoiditis/Adhesions

KEY FACTS

TERMINOLOGY

- Postinflammatory changes in cauda equina

IMAGING

- Thickening and clumping of nerve roots in cauda equina
- Adhesion of nerve roots to peripheral dura ("empty sac" sign)
- Soft tissue mass (pseudomass)
- Minimal to mild pial, dural enhancement
- Nerve root calcification (rare) or calcific mass (arachnoiditis ossificans)

TOP DIFFERENTIAL DIAGNOSES

- Spinal stenosis
- Cauda equina neoplasms
- Carcinomatous meningitis
- Intradural metastases

PATHOLOGY

- Inflammatory, collagenous mass
- Historically related to trauma or spinal meningitis, now more commonly associated with prior lumbar surgery

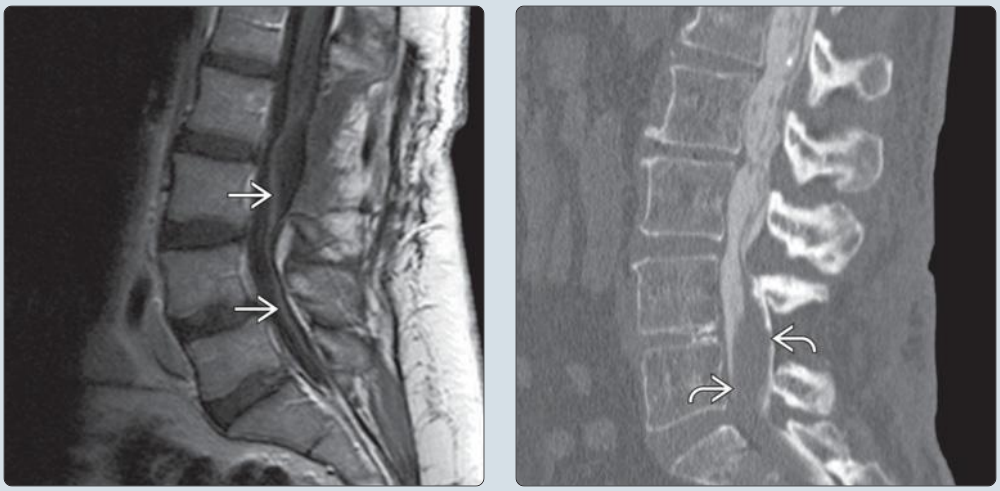
CLINICAL ISSUES

- Most common symptoms are chronic low back pain or leg pain (radicular or nonradicular)
  - Simulates spinal stenosis and polyneuropathy
- Less common are paraparesis, hypoesthesia, gait disorder, bowel/bladder dysfunction

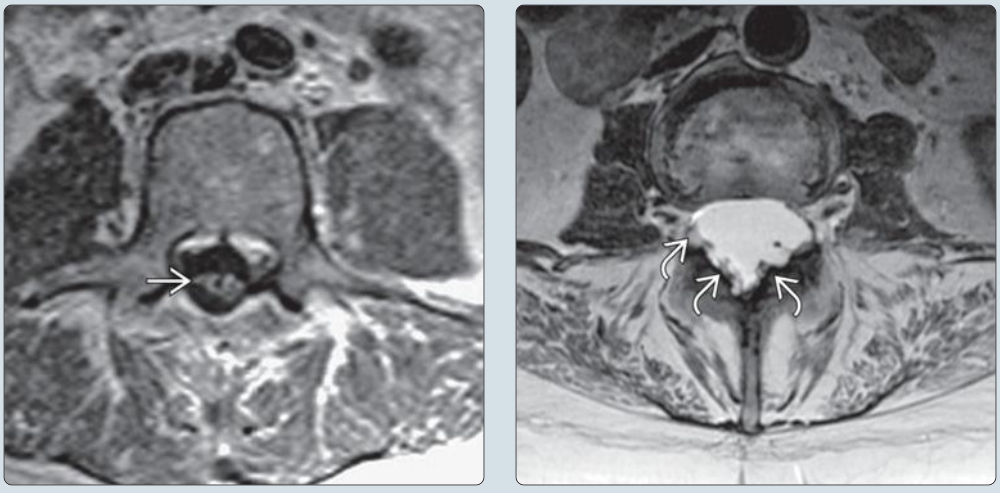
DIAGNOSTIC CHECKLIST

- Absent discrete nerve roots in thecal sac with clumping or "empty sac" sign highly suggestive of lumbar arachnoiditis
- Radiological findings may be present without clinical symptoms

(Left) *Sagittal T1 C+ MR shows clumping of the nerve roots of the cauda equina. There is faint enhancement. A L2-L3 laminectomy defect is noted.* **(Right)** *Sagittal CECT myelography shows a large, mass-like filling defect in the caudal thecal sac (pseudomass) engulfing the cauda equina due to chronic changes of arachnoiditis.*



(Left) *Axial T1WI C+ MR shows clumping of nerve roots in the thecal sac due to arachnoiditis with mild enhancement.* **(Right)** *Axial T2WI MR in this patient with ankylosing spondylitis shows peripheral adhesion of the nerve roots to the margins of the thecal sac, resulting in the "empty sac" sign of arachnoiditis. This pattern may cause cauda equina syndrome in patients with longstanding AS.*



TERMINOLOGY

Definitions

- Postinflammatory changes involving spinal cord, cauda equina, and thecal sac

IMAGING

General Features

- Best diagnostic clue
  - Thickening and clumping of nerve roots in cauda equina
  - Adhesion of nerve roots to peripheral dura ("empty sac" sign)
- Location
  - Lumbar spine
    - Cauda equina
- Morphology
  - Intrathecal clumping of nerve roots causing enlarged cords
  - "Empty sac" appearance with peripheral clumping of nerves to dura
  - Soft tissue mass (pseudomass) filling thecal sac
  - Intradural cysts

Radiographic Findings

- Radiography
  - Evidence of prior lumbar surgery

CT Findings

- CECT
  - Faintly enhancing nerve roots and dura
- Bone CT
  - Rare calcifications of nerve roots or calcific mass (arachnoiditis ossificans)
- CT myelography
  - "Empty sac" sign due to nerve roots adhering to periphery of sac
  - Absent nerve root sleeve filling
  - Thickening, clumping of nerve roots
  - Thickened, retracted dura
  - Distorted thecal sac

MR Findings

- T1WI
  - Indistinct cord due to increased CSF intensity
  - May encounter sequelae of iodophenylundecylic acid (Pantopaque) myelography (high signal droplets)
- T2WI
  - Clumping of nerve roots
    - Centrally = type 1
      - May involve only 2-3 nerve roots
    - Peripheral = type 2
    - Nerve roots adhering to walls of thecal sac
      - "Empty thecal sac" sign
      - Central CSF without nerve roots
      - Thickened thecal sac wall
  - Soft tissue mass filling most of thecal sac (type 3)
    - Clumped nerve roots
    - Decreased sac diameter
    - Obliterated subarachnoid space
  - Intradural cysts
    - Loculated CSF, may be septated
    - Formed by adhesions
- T1WI C+
  - Minimal to mild pial and dural enhancement
    - Smooth, linear
    - Nodular
    - Mass-like intradural enhancement
  - Enhancement also encountered in immediately postoperative patients without myelographic evidence of arachnoiditis
  - No correlation between degree of enhancement and severity of symptoms
- Findings extend over at least 2 lumbar vertebrae

Nonvascular Interventions

- Myelography
  - Type 1
    - Localized or diffuse intrathecal filling defects
    - Thickened roots present as large linear filling defect in contrast column
    - Loculated, clumped contrast: Must be distinguished from epidural or subdural injection
  - Type 2
    - "Empty sac" sign
    - "Candle-guttering" sign with thickening of peripheral dura with adherent roots
    - Homogeneous filling of thecal sac
    - Absent or blunted nerve root sleeve filling
  - Type 3
    - Conglomerate inflammatory mass
    - Enhances more poorly than typical intradural neoplasm
  - Types form continuum of findings

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR with T1WI and T2WI in sagittal and axial planes
  - Contrast administration necessary

DIFFERENTIAL DIAGNOSIS

Spinal Stenosis

- Clumped nerve roots at site of stenosis
- Tortuous, elongated roots cephalad to stenosis
- Degenerative changes prominent
  - Disc bulge, herniation, endplate osteophytosis
  - Facet arthropathy, degenerative hypertrophy of thickened ligamentum flavum

Cauda Equina Neoplasms

- Myxopapillary ependymoma
- Large nerve sheath tumor
- Paraganglioma
- All demonstrate avid post-gadolinium enhancement ± conus displacement

Carcinomatous Meningitis

- Smooth or nodular nerve root enhancement
- Clumping of nerve roots usually absent

Intradural Metastases

- Nodules or masses within cauda equina
- Significant post-gadolinium enhancement

PATHOLOGY

General Features

- Etiology
  - Lumbar surgery
    - Especially when multilevel or complicated
    - In combination with antecedent myelography
      - Especially Pantopaque
  - Infectious spinal meningitis
    - Tuberculosis
    - Syphilis
    - Fungal
    - Parasitic
  - Intrathecal hemorrhage
    - Shown to irritate meninges in animal models
    - Dorsal thoracic arachnoiditis more common in subarachnoid hemorrhage
  - Other causes
    - Spinal anesthesia, epidural blood patch
    - Spinal trauma
    - Degenerative disc disease
  - Pathophysiology
    - Infectious meningitis: Vascular congestion, inflammatory exudate
    - Perineural and leptomeningeal fibrosis
    - Collagen deposition by fibroblasts
    - Nerve roots adhere to each another, thecal sac
    - CSF loculation
  - Inflammation of all 3 meningeal layers and nerve roots
- Genetics
  - No familial predilection
- Associated abnormalities
  - Syringomyelia
    - Alteration in CSF circulation
    - Increased amount of CSF entering cord
    - Symptoms higher in cord

Gross Pathologic & Surgical Features

- Inflammatory, collagenous mass
- May be calcified
- Posterior nerve roots involved more commonly

Microscopic Features

- Chronic lymphocytic infiltration
- Collagen deposition
- Small foci of calcifications

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Chronic low back pain
  - Radicular or nonradicular leg pain
    - Simulates spinal stenosis and polyneuropathy
  - Other signs/symptoms
    - Paraparesis
    - Hypoesthesia
    - Gait disorder
    - Bladder and bowel dysfunction
- Clinical profile
  - No defining clinical symptomatology
  - Radiological findings may be present without clinical symptoms

Demographics

- Gender
  - No gender predilection
- Ethnicity
  - No racial predilection
- Epidemiology
  - Uncommon
  - Clinical syndrome more common
    - 6-16% of postoperative patients

Natural History & Prognosis

- Symptoms usually static
  - Fluctuate in severity
- Small percentage with progressive neurologic deficits
  - 1.8% in 1 series

Treatment

- Intrathecal steroid injection
- Spinal cord stimulation
  - Treatment of choice when pain is predominant symptom
  - Immediate success rate (pain relief): > 70%
  - Intermediate success rate: > 50%
  - Long-term success rate: > 30%
- Pain rehabilitation
- Laminectomy with microlysis of adhesions
  - Reserved for patients with progressive neurologic deficits
  - 50% initial success rate
  - Effectiveness decreases over time

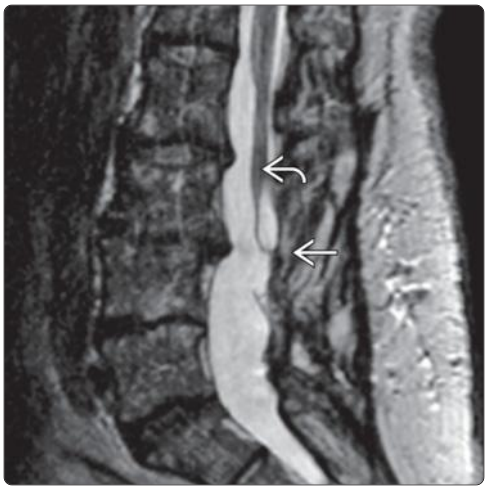
DIAGNOSTIC CHECKLIST

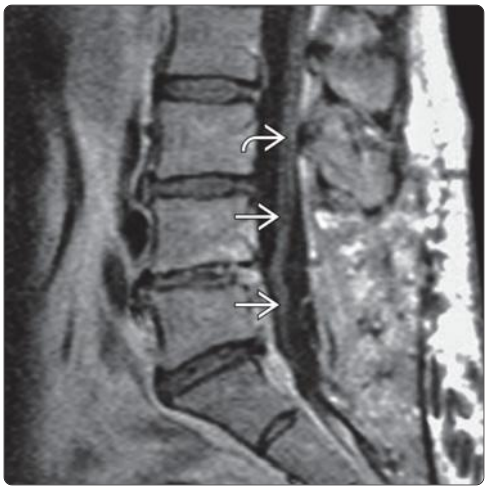
Image Interpretation Pearls

- Absent discrete nerve roots in thecal sac with clumping or "empty sac" sign highly suggestive of lumbar arachnoiditis

SELECTED REFERENCES

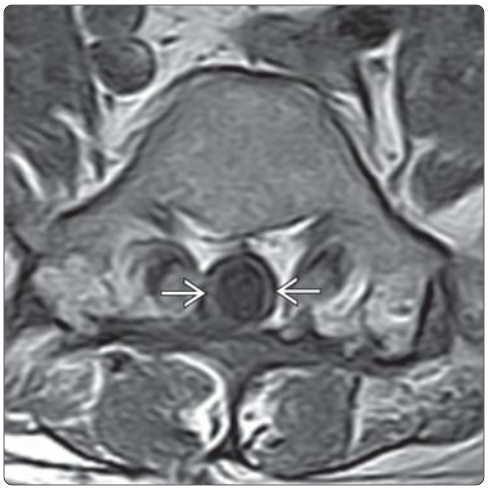
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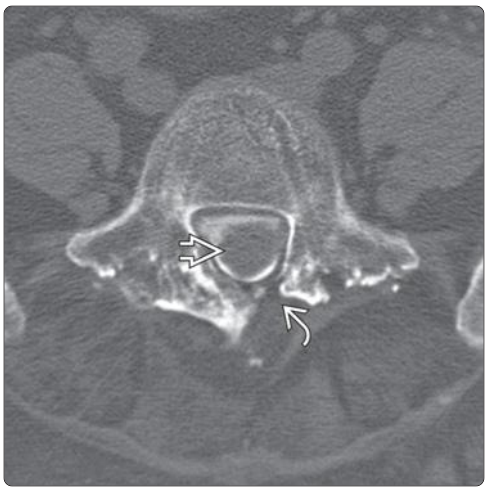
(Left) *Sagittal T2WI shows clumping of the cauda equina at the L2-L3 level due to type 1 arachnoiditis. Caudally, the nerve roots are tethered to the dorsal thecal sac. There is a laminectomy defect extending through L3-L5.* **(Right)** *Sagittal contrast-enhanced T1WI shows central clumping of the nerve roots of the cauda equina. Note the minimal pial enhancement. There has been a L4-L5 laminectomy.*





(Left) *Sagittal T2WI MR shows clumping of the cauda equina from arachnoiditis. A cystic mass is seen at the L2-L3 level, where there has been formation of an arachnoid cyst.* **(Right)** *Axial T1WI MR shows a thick rind of signal along the dural margin with an empty thecal sac, reflecting type 2 arachnoiditis. Note the prior laminectomy and dorsal epidural fibrosis.*





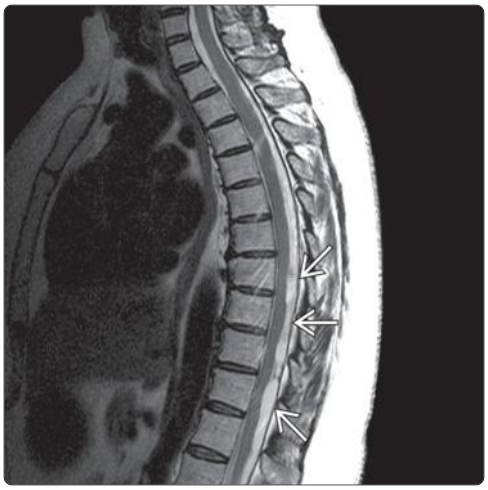
(Left) *Sagittal CECT myelography shows a soft tissue mass filling the distal thecal sac and blocking contrast flow at the L2-L3 level in this patient with type 3 arachnoiditis.* **(Right)** *Axial CT myelography shows a large, mass-like filling defect centrally in the thecal sac in this patient with type 3 arachnoiditis. The mass engulfs rather than displaces the nerve roots. Changes of the left hemilaminectomy are noted.*

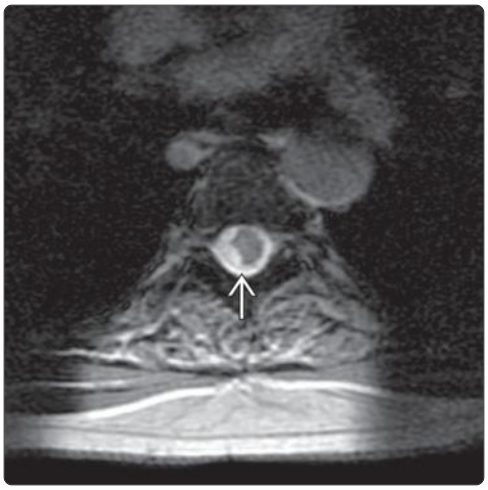
(Left) *AP myelogram shows abnormal contrast distribution with a poorly defined cauda equina and irregular caudal thecal sac in this patient with arachnoiditis. This pattern looks similar to a subdural injection.* **(Right)** *Axial CT myelogram shows abnormal clumping of the nerve roots both ventral and dorsal within the thecal sac. The focal outline of nerve roots would not be seen if this was a subdural injection, which can give a similar appearance.*



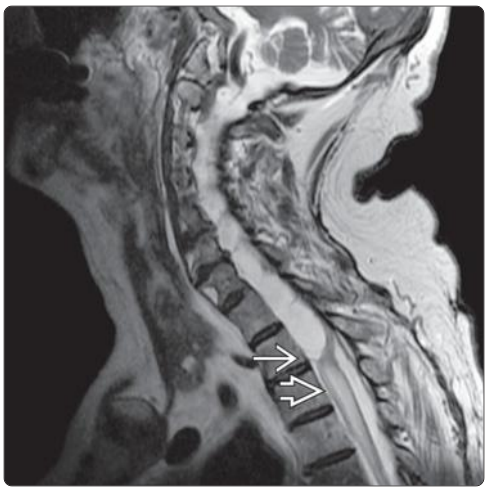


(Left) *Sagittal T2WI MR shows a lobulated posterior margin of the mid and lower thoracic spinal cord due to adhesive arachnoiditis that developed as a late complication after an intracranial subarachnoid hemorrhage.* **(Right)** *Axial GRE MR shows the extramedullary abnormality of adhesive arachnoiditis with ventral displacement of the cord and thin adhesions attached to the cord and dura.*

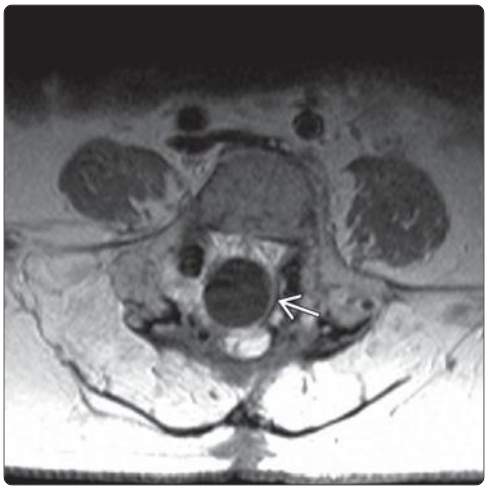


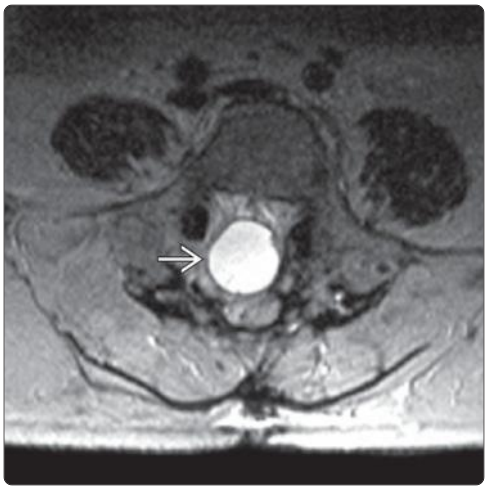


(Left) *Sagittal T2WI MR 1 year later after subarachnoid hemorrhage shows development of extensive adhesions/ arachnoiditis with tethering of the cord dorsally at the T3 level. The abnormal CSF flow dynamics and tethering has now produced cord edema.* **(Right)** *Sagittal T2WI MR shows complications of coccidioidomycosis meningitis with hydrocephalus, secondary arachnoiditis, and cervical arachnoid cyst formation. There is cord edema from altered CSF flow.*

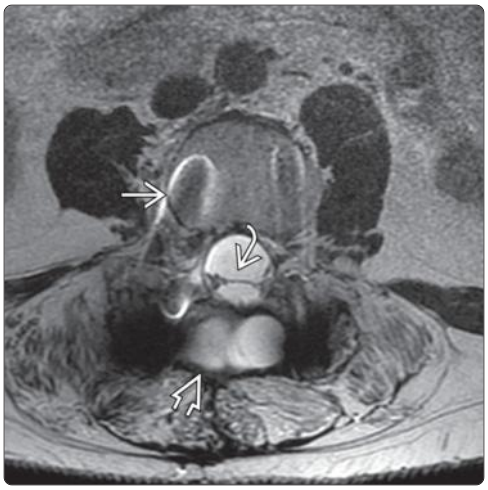






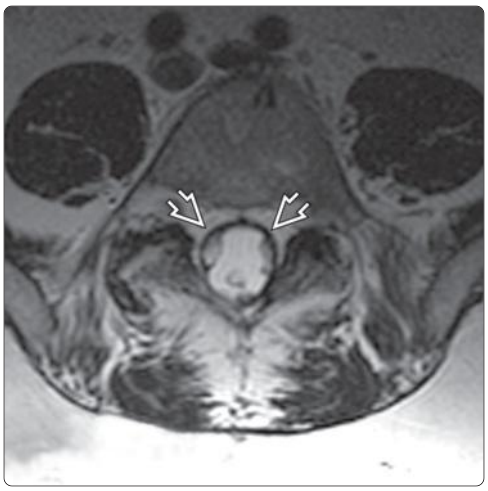


(Left) *Axial T1W C+ MR shows extensive postoperative changes with posterior laminectomy. There is slight peripheral dural enhancement, but no roots within the low central CSF signal.* **(Right)** *Axial T2WI MR shows the lack of nerve roots within the thecal sac below the L4 level ("empty thecal sac" sign), with extensive postoperative changes.*





(Left) *Axial T2WI MR shows extensive postoperative changes with a metal artifact from bilateral pedicle screws, laminectomy defect, and a small sterile fluid collection at the laminectomy site. There is abnormal clumping of the nerve roots from arachnoiditis.* **(Right)** *Axial T2WI MR shows thickening and peripheral clumping of the nerve roots into the dura with central cystic-like accumulation of CSF due to extensive adhesive postoperative arachnoiditis.*





(Left) *Axial T2WI MR shows diffuse thickening of the distal dural sac and clumping of the roots consistent with severe arachnoiditis. Note the thickening low signal dural margin suggestive of calcific arachnoiditis.* **(Right)** *Axial bone NECT shows irregular calcification involving the distal thecal sac at the sacral level. This finding is consistent with end-stage calcific arachnoiditis.*

Arachnoiditis Ossificans

KEY FACTS

TERMINOLOGY

- Intradural ossification associated with post-inflammation adhesion and clumping of lumbar nerve roots

IMAGING

- Calcification morphology
  - Thin, linear
  - Mass-like, globular
- Calcific density on CT
  - Contrast may obscure calcifications on CT myelography
- T1WI: Areas of ossification are variable, mixed signal
  - Hypointense, isointense, or hyperintense
- T2WI: Linear or globular hypointensity if calcifications are present
  - Larger areas of ossification can occasionally be hyperintense on T2WI
- May exert mass effect on conus and cauda equina

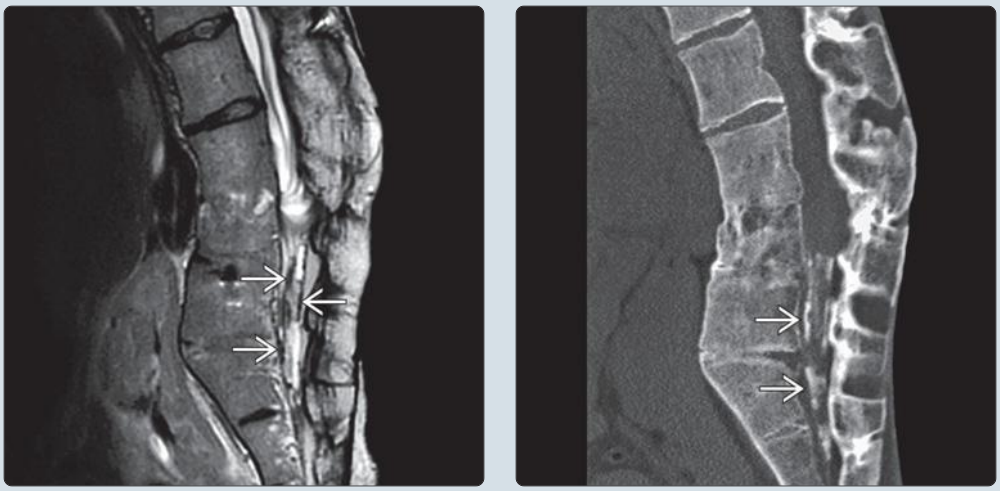
PATHOLOGY

- Etiologies
  - Prior trauma
  - Spinal surgery
  - Subarachnoid hemorrhage
  - Pantopaque myelography
  - Spinal anesthesia
- Gross pathology: Calcified, inflammatory, collagenous mass

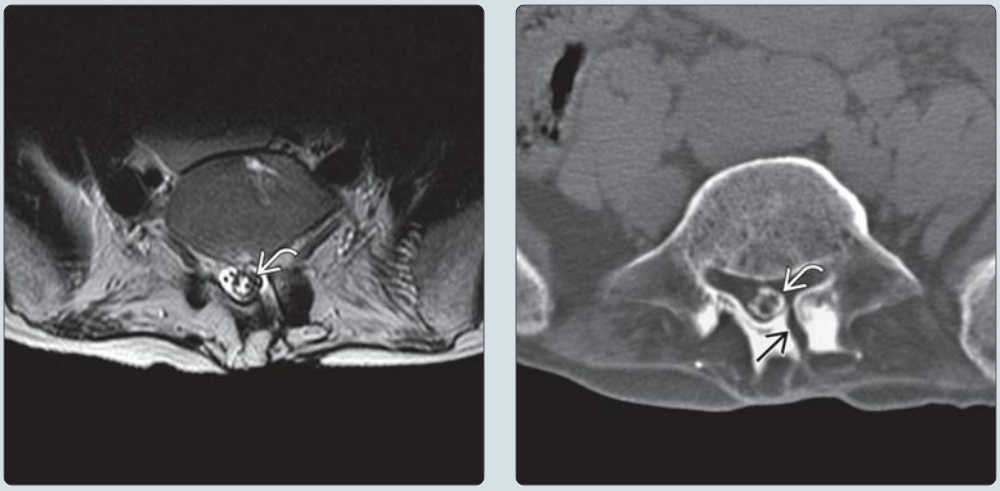
CLINICAL ISSUES

- Low back pain
- Radicular or nonradicular leg pain
- Paraparesis
- Bladder and bowel dysfunction

(Left) *Sagittal T2WI MR through the lumbar spine shows a nodular hypointense signal along the margins of the caudal thecal sac due to calcific arachnoiditis.* **(Right)** *Sagittal NECT through the lumbar spine more clearly shows the coarse calcifications of the thecal sac. CT also shows intervertebral fusion at L3-L4, L4-L5, and L5-S1.*



(Left) *Axial T2WI MR again shows irregular hypointensity about the margins of the thecal sac due to dystrophic calcifications.* **(Right)** *Axial NECT shows coarse calcifications of the caudal thecal sac. Also seen is a surgical defect of the left L5 lamina.*



TERMINOLOGY

Definitions

- Intradural ossification associated with post-inflammation nerve root clumping, arachnoid adhesions

IMAGING

General Features

- Best diagnostic clue
  - Calcific density on CT within lumbar canal
- Location
  - Lumbar spine
- Morphology
  - Thin, linear or mass-like, globular

Radiographic Findings

- Radiography
  - Intraspinal calcification

CT Findings

- Bone CT
  - Calcific densities within clumped nerve roots

MR Findings

- T1WI
  - Areas of ossification are of variable signal on T1WI
    - Hypointense, isointense, or hyperintense
- T2WI
  - Clumped nerve roots forming cords and mass(es)
  - Secondary adhesions and arachnoid cysts
  - Linear or globular hypointensity if calcifications are present
    - Larger areas of ossification can occasionally be hyperintense on T2WI
  - May exert mass effect on conus and cauda equina
- T1WI C+
  - Minimal or no nerve root enhancement

Nonvascular Interventions

- Myelography
  - Intrathecal contrast may obscure calcifications

Imaging Recommendations

- Best imaging tool
  - Axial thin section bone CT with sagittal reformation

DIFFERENTIAL DIAGNOSIS

Calcified Dural Plaques

- Thin dural calcifications
- Asymptomatic, incidental finding in elderly

Spinal Stenosis

- Clumped nerve roots
- Degenerative changes prominent
- No intrathecal calcification

Intrathecal Neoplasm

- Enhancing mass, noncalcified soft tissues
- Displacement of cauda equina fibers

Retained Pantopaque

- Hyperintense on T1WI
- Iso- to hypointense on T2WI
- Differentiated from calcification by CT

PATHOLOGY

General Features

- Etiology
  - Prior trauma, lumbar surgery, subarachnoid hemorrhage, myelography, spinal anesthesia
  - Pathophysiology of lumbar arachnoiditis
    - Fibrin deposition
    - Proliferating fibroblasts
    - Perineural and leptomeningeal fibrosis
  - Ossification results from
    - Intrathecal hematoma
    - Incorporated bone fragments
    - Osseous metaplasia from chronic inflammation and fibrosis
    - End-stage chronic arachnoiditis
- Associated abnormalities
  - Syringomyelia

Gross Pathologic & Surgical Features

- Calcified inflammatory, collagenous mass

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Low back pain
  - Other signs/symptoms
    - Radicular or nonradicular leg pain
    - Paraparesis, bladder and bowel dysfunction
- Clinical profile
  - No defining clinical symptomatology
    - Simulates spinal stenosis and polyneuropathy

Demographics

- Epidemiology
  - Rare

Natural History & Prognosis

- Possible progressive neurologic impairment

DIAGNOSTIC CHECKLIST

Consider

- Thin section bone CT when heterogeneous intrathecal signal present in setting of arachnoiditis

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Accelerated Degeneration

KEY FACTS

TERMINOLOGY

- Accelerated degeneration of disc space/facets at level(s) adjacent to surgical fusion

IMAGING

- Degenerative disc/facet changes directly above or below fusion
- Also occurs adjacent to congenital segmentation anomalies
- May show increased motion at degenerated level adjacent to fused segment
- Flexion/extension views for definition of instability
- Plain films most economical way to demonstrate presence of adjacent segment degenerative changes and to serially follow for progression
- MR best identifies soft tissue abnormalities that are occult on plain film

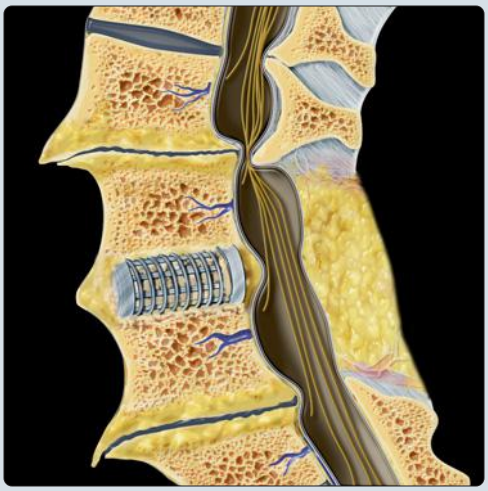
TOP DIFFERENTIAL DIAGNOSES

- Disc space infection
- Pseudoarthrosis
- Spondylolysis
- Spondylolisthesis
- Normal postoperative changes

PATHOLOGY

- Produced by aberrant biophysical stresses from altered normal spinal motion
- Solid fusion alters biomechanics at adjacent mobile levels
- More common with multilevel fusion but also seen following single level fusion
- Presence of osteopenia at time of cervical surgery increases risk of adjacent segment degeneration
- Concurrent lumbar spine degeneration at time of cervical surgery carries higher risk of adjacent segment degeneration

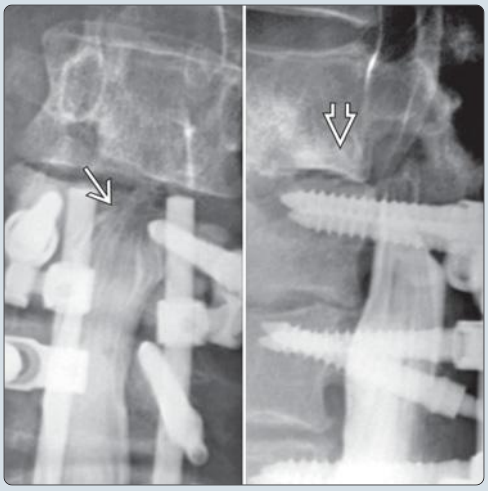
(Left) *Sagittal graphic shows solid L4-L5 interbody fusion and laminectomy. Note severe disc degeneration at L3-L4 with spondylolisthesis, loss of disc height, osteophytes and central stenosis, and severe degeneration at L5-S1 due to altered biomechanics.* **(Right)** *Sagittal T1WI MR shows a solid fusion at C5-C6 with normal fatty marrow signal. There is disc degeneration at C6-C7 with decreased disc height and large disc extrusion compressing cord ⇨.*





(Left) *Sagittal T2WI MR shows patient post C4-C7 fusion with anterior plating giving low signal artifact ⇨. There is a large synovial cyst showing central hyperintensity at the C3-C4 level ⇨.* **(Right)** *Anteroposterior myelography (left) shows block to contrast at L1-L2 ⇨ at the upper margin of posterior fusion. Lateral view (right) shows contrast block and severe L1-L2 degeneration ⇨.*





TERMINOLOGY

Synonyms

- Spinal "transitional" degenerative syndrome, accelerated segmental degeneration
- Adjacent segment degeneration

Definitions

- Accelerated degeneration of disc space/facets at level(s) adjacent to surgical fusion

IMAGING

General Features

- Best diagnostic clue
  - Degenerative disc/facet changes directly above or below fusion
  - Also occurs adjacent to congenital segmentation anomalies
- Location
  - Degenerative disc/facet changes directly adjacent to fused vertebra
- Size
  - Varies from mild to severe disc degenerative change with deformity
- Morphology
  - Disc degenerative change related to disc, endplates, facets

Radiographic Findings

- Radiography
  - Plain films show surgical change with adjacent segment degenerative changes
  - Flexion/extension views for definition of instability

Fluoroscopic Findings

- May show increased motion at degenerated level adjacent to fused segment

CT Findings

- NECT
  - Findings of degenerative disc disease at transitional level
    - Loss of disc space height
    - Disc vacuum phenomenon
    - Bony endplate eburnation
    - Osteophyte
    - Facet degenerative arthropathy with foraminal stenosis
    - Anterolisthesis or retrolisthesis
    - Disc bulge, protrusion, extrusion
    - Central canal stenosis

MR Findings

- T1WI
  - Typical findings seen in degenerative disc and facet disease
    - Loss of disc space height
    - Low signal foci within disc: Vacuum phenomena &/or calcification
    - May show degenerative type I, II endplate changes
    - Disc bulge, herniation
    - Central &/or foraminal stenosis
    - Facet degenerative arthropathy
    - Malalignment
- T2WI
  - Typical findings seen in degenerative disc and facet disease
    - Loss of signal within intervertebral disc
    - Degenerative type I, II endplate changes
- STIR
  - High signal marrow edema with type I endplate changes at transitional level
- T1WI C+
  - Adjacent vertebral body endplates may show marked enhancement with type I endplate changes
  - Linear enhancement within intervertebral disc due to degeneration
    - No endplate destruction

Nonvascular Interventions

- Myelography
  - Nonspecific extradural disease (stenosis, herniation) at transitional level
  - May show myelographic block to contrast material with severe disease

Nuclear Medicine Findings

- Bone scan
  - Increased uptake related to disc degeneration
- PET
  - Hypometabolic as with degenerative disc disease
  - Hypermetabolic with disc space infection/osteomyelitis
  - PET accurate and specific to distinguish infection from degenerative endplate changes

Imaging Recommendations

- Plain films most economical way to demonstrate presence of adjacent segment degenerative changes and to serially follow for progression
- MR best identifies soft tissue abnormalities that are occult on plain film
  - Disc herniation, ligamentum flavum laxity, synovial proliferation

DIFFERENTIAL DIAGNOSIS

Normal Postoperative Changes

- Low signal from interbody fusion cages with typical spatial mismapping of signal

Disc Space Infection

- Low signal involving adjacent endplates similar to type I degenerative change
- Endplate destruction
- ± paravertebral or epidural soft tissue phlegmon/abscess

Pseudoarthrosis

- Accelerated degenerative change may progress to pseudoarthrosis with abnormal motion, involvement of posterior elements/ligaments
- Abnormal horizontal signal extends through disc to posterior elements or interspinous ligament

Spondylolysis

- Pars defect as cause of instability

Spondylolisthesis

- Diagnosis rests with presence or absence of prior fusion, instrumentation, and level of listhesis
- Instability and abnormal biomechanics of fusion site may lead to stress fractures at transitional level

PATHOLOGY

General Features

- Etiology
  - Produced by aberrant biophysical stresses from altered normal spinal motion
    - Wolff law: Living tissue responds to chronic changes in stresses and strains
  - Solid fusion alters biomechanics at adjacent mobile levels
  - Increased mobility in these remaining mobile segments is hypothesized to cause accelerated degenerative pathologic changes
    - Segmental motion and intradiscal pressure above or below fusion increases
  - More common with multilevel fusion, but also seen following single level fusion
    - Most commonly affects proximal levels in lumbar spine
  - Not all patients at risk demonstrate accelerated degeneration
  - Pathological findings are typical of degenerative disc and facet disease
  - Presence of osteopenia at time of cervical surgery increases risk of adjacent segment degeneration
  - Concurrent lumbar spine degeneration at time of cervical surgery carries higher risk of adjacent segment degeneration
  - Preexisting facet degeneration is associated with high risk of adjacent segment problems following lumbar fusion procedures

Staging, Grading, & Classification

- Spondylolisthesis may be graded I-IV

Gross Pathologic & Surgical Features

- Disc space height loss, disc bulge or herniation, central stenosis, facet osteoarthritic change

Microscopic Features

- Disc dehydration, fissuring, anular disruption, disc protrusion or extrusion

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be asymptomatic
  - Mechanical pain
  - Radicular or myelopathic symptoms referable to level(s) above or below surgical fusion
  - Herniation above site of fusion < 2%
- Clinical profile
  - Prior surgical fusion with new and progressing pain in different distribution than preoperative pain

Demographics

- Age
  - Adult
- Gender
  - M = F
- Epidemiology
  - More common after surgical fusion than following decompression only
  - Bertolotti syndrome refers to association of back pain with lumbosacral transitional vertebrae
    - Disc bulge/herniation 9x more common at interspace immediately above transitional vertebra than at any other level

Natural History & Prognosis

- Difficult to distinguish natural history of disc degeneration from accelerated degeneration following fusion
- Progressively worsens
  - Highly variable course
- Decreased signal within adjacent discs on T2WI may occur as early as 12 months following fusion
- Below scoliosis fusion chance of herniation up to 34%, chance of disc signal abnormality 50%

Treatment

- Surgical decompression and fusion at adjacent symptomatic levels

DIAGNOSTIC CHECKLIST

Consider

- Pseudoarthrosis if extensive signal abnormality involving anterior and posterior elements in horizontal fashion

Image Interpretation Pearls

- Presence of prior surgical fusion, instrumentation
- Look for transitional lumbosacral vertebral body

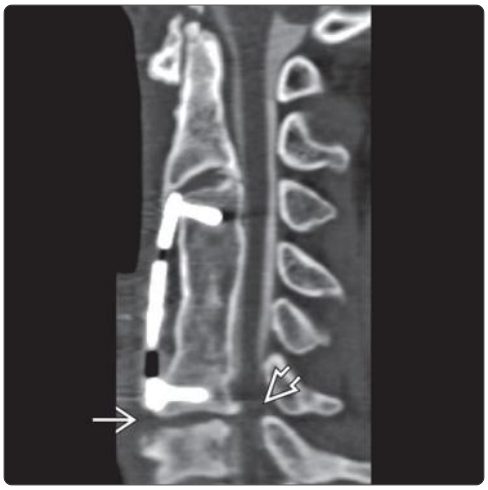
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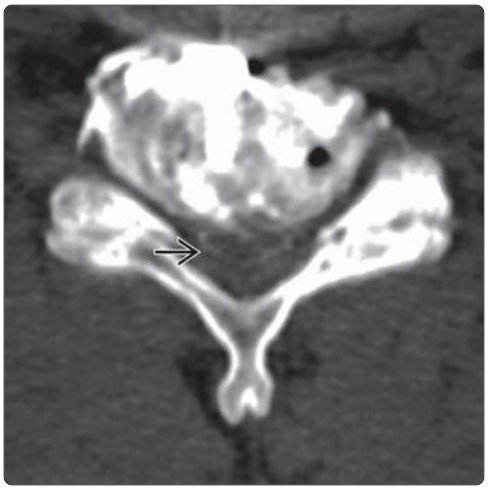
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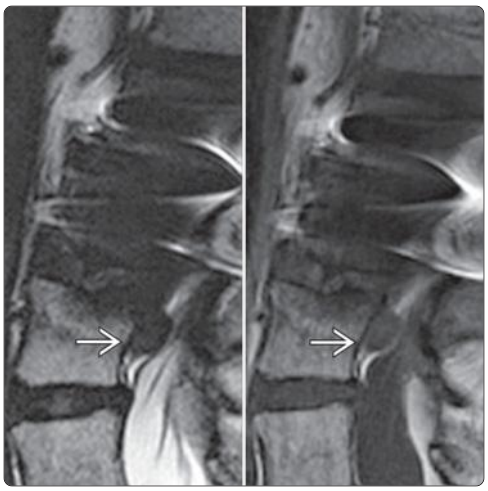


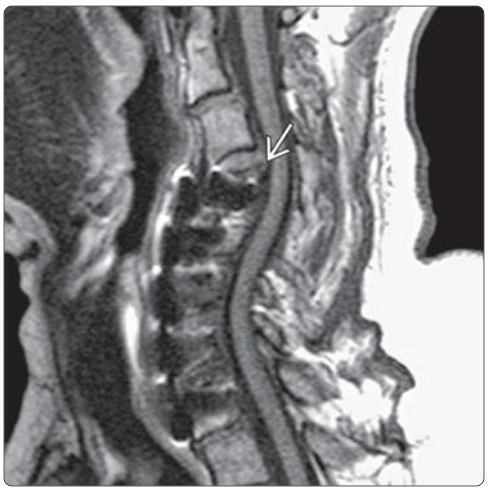
(Left) *Sagittal T1WI MR shows patient post C3-C5 fusion with a graft. There is degeneration of C5-C6 with central stenosis ⇨. Congenital fusion of C6-C7 further increases biomechanical stress on the C5-C6 disc level.* **(Right)** *Sagittal T2WI MR shows a solid fusion of C3-C5. Degeneration is present at C5-C6 with central stenosis and narrowed subarachnoid space. Hyperintensity at C5-C6 within the cervical cord is due to myelomalacia ➡.*





(Left) *Sagittal CT reformat following myelography shows a C3-C6 solid fusion with anterior plate and screws. The sagittal reformat shows long segment fusion and marked disc space height loss at C6-C7 ➡. There is severe central stenosis with loss of the contrast column ➡.* **(Right)** *Axial CECT at C6-C7 shows severe central stenosis at the level below the cervical fusion ⇨, as well as severe bilateral bony foraminal stenosis with facet and uncovertebral hypertrophy.*





(Left) *Sagittal T2WI MR (left) and T1WI C+ MR (right) with lumbar pedicle screws giving artifact demonstrate peripheral enhancing disc extrusion ➡ below the fusion site with thecal sac compression.* **(Right)** *Sagittal T1WI MR shows that the patient is post extensive C4-T1 fusion with anterior plate/screws. There is herniation above the fusion site at C3-C4 ➡ with impingement upon the cord as well as increased kyphosis.*

KEY FACTS

TERMINOLOGY

- Infectious sequelae following operative procedures

IMAGING

- Unexpected abnormal MR enhancement following spinal surgery
  - Begins at manipulated disc level as discitis
  - Spreads to adjacent local structures → "osteomyelitis-discitis"
  - Phlegmon/abscess may be paravertebral &/or epidural
- Radiographs insensitive early but later show demineralization with endplate lysis/erosion
- CT demonstrates adjacent endplate ± facet erosions
- MR bone marrow edema is T1 hypointense and T2 hyperintense
  - STIR best images marrow edema
  - Abscess appears T2 hyperintense/T1 hypointense with peripheral enhancement
  - Discitis appears as amorphous enhancement
  - Fat-suppressed techniques ↑ infection conspicuity
- FDG PET reveals increased ↑ glucose utilization 2° to activated neutrophils and macrophages in inflammatory reactions
  - Negative predictive value, sensitivity, specificity, and accuracy for diagnosing postoperative spine infection are 100%, 100%, 81%, and 86%, respectively
- MR C+; consider PET in presence of MR hardware susceptibility artifact

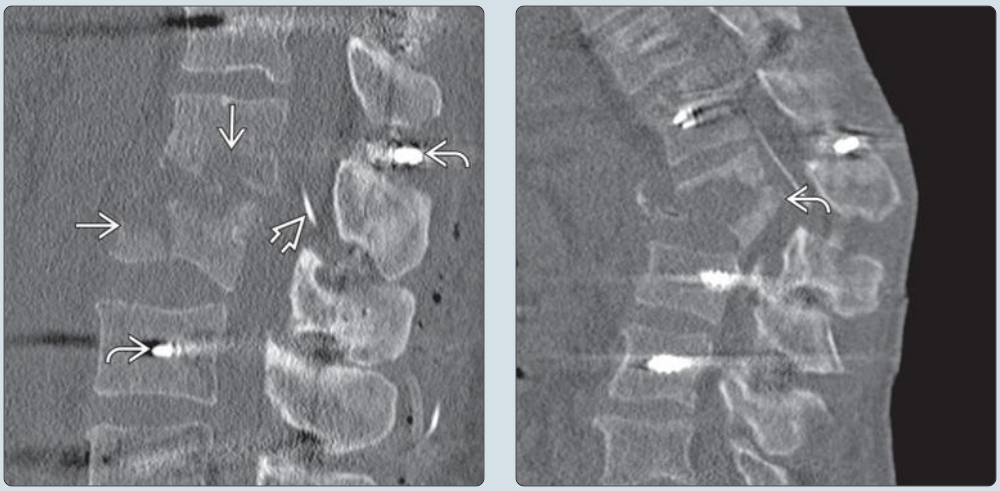
TOP DIFFERENTIAL DIAGNOSES

- Osteomyelitis, pyogenic
- Osteomyelitis, granulomatous
- Neoplasm

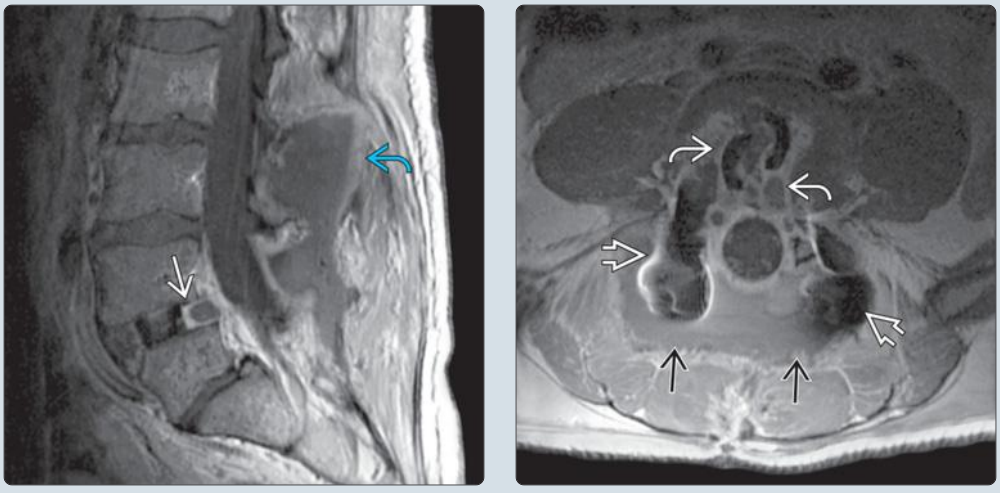
CLINICAL ISSUES

- Biopsy to establish pathogen
- Effective treatment may include removal of implants, irrigation, and debridement

(Left) *Sagittal CT reconstruction of the midthoracic spine shows endplate destructive changes due to postoperative infection. The bones are osteopenic, and there is bone destruction extending across the disc space. An intrathecal catheter and pedicle screws are partially seen.* **(Right)** *Follow-up CT reveals further endplate osteolytic changes, vertebral body collapse, and focal kyphosis.*



(Left) *Sagittal T1WI C+ MR demonstrates L4/L5 PLIF (posterolateral intervertebral fusion) with posterior laminectomy. Abnormal fluid signal intensity is seen in the L4/L5 disc interspace, and rim-enhancing fluid collection is noted within the dorsal soft tissues. The posterior soft tissue abscess collection has a fluid-debris interface.* **(Right)** *Abscess collection surrounds the posterior spinal fusion hardware (metallic susceptibility). Enhancing fluid envelopes the interbody spacers.*



TERMINOLOGY

Definitions

- Infectious sequelae following operative procedures

IMAGING

General Features

- Best diagnostic clue
  - Unexpected postoperative fluid collection with bone destruction
- Location
  - Begins at manipulated disc level as discitis
    - Less commonly at facets as facetitis
  - Local spread → "osteomyelitis-discitis"
  - Phlegmon/abscess may be paravertebral ± epidural
- Size
  - Focal to widespread process
- Morphology
  - Epidural "curtain" sign = bilobed posterior bulging of abscess (or other process) within epidural space on both sides of meningovertebral ligament

Radiographic Findings

- Radiography
  - Early: Insensitive
  - Later: Demineralization with endplate lysis/erosion

CT Findings

- CECT
  - Phlegmon: Enhancing soft tissue collection without low-density collection
  - Abscess: Hypodense collections within phlegmon
- Bone CT
  - Discitis: Adjacent endplate erosion
  - Facetitis: Osseous demineralization ± erosion

MR Findings

- T1WI
  - Edema: Hypointense marrow
  - Discitis: Disc hypointensity with loss of disc height; may be complete obliteration of disc space
    - Degenerative endplate changes may also show low signal on T1WI and may show enhancement
  - Facetitis: Hypointense marrow
  - Phlegmon: Paravertebral soft tissue mass (intermediate signal intensity)
  - Abscess: Hypointense collections within phlegmon
- T2WI
  - Edema: Hyperintense marrow
  - Discitis: Disc hyperintensity or fluid intensity with loss of disc height
  - Facetitis: Hyperintense facet fluid collection
  - Phlegmon: Hyperintense paravertebral soft tissue
  - Abscess: Hyperintense collections within phlegmon
  - Arachnoiditis: Nerve root clumping, adherence to thecal sac ("empty sac" sign)
- STIR
  - Best to visualize hyperintense marrow and soft tissue edema
- T1WI C+
  - Discitis: Disc enhancement
    - Normal postop disc: Linear bands of enhancement paralleling endplate margins in 20% at 3 months
  - Facetitis: Facet enhancement with ill-defined margin
  - Phlegmon: Enhancing soft tissue
  - Abscess: Fluid within enhancing phlegmon
  - Osteomyelitis: Marrow enhancement
  - Arachnoiditis: Nerve root clumping and enhancement
- Chronic MR findings
  - Anterior wedging of affected vertebrae with fusion across narrowed/obliterated disc space
  - Sclerosis: T1/T2 hypointense

Nonvascular Interventions

- Myelography
  - Nonspecific, showing distortion of contrast-filled thecal sac from extradural disease

Nuclear Medicine Findings

- Bone scan
  - Increased uptake, but limited value given uptake secondary to procedure itself
- PET
  - 18F-fluorodeoxyglucose (FDG) PET may become standard imaging technique
    - ↑ glucose utilization of activated neutrophils and macrophages in inflammatory reactions
    - Not hindered by presence of metallic implants
    - Provides rapid results (2 hours) and high-resolution images (4–5 mm)
  - Negative predictive value, sensitivity, specificity, and accuracy for diagnosing postoperative spine infection are 100%, 100%, 81%, and 86%, respectively
- WBC scan
  - Of limited use 2° to high uptake of labeled leukocytes in normal hematopoietically active bone marrow

Imaging Recommendations

- Best imaging tool
  - C+ MR; consider PET when hardware susceptibility limits MR imaging quality
- Protocol advice
  - STIR best images for marrow edema
  - Fat-suppressed techniques ↑ infection conspicuity

DIFFERENTIAL DIAGNOSIS

Osteomyelitis, Pyogenic

- Most often bacterial (usually *Staphylococcus aureus*)
- IV drug abusers at higher risk

Osteomyelitis, Granulomatous

- Usually TB (immunocompromised at ↑ risk)
- Often 2 or more contiguous levels, may spare intervening disc

Neoplasm (Metastatic)

- Adjacent vertebral bodies typically sparing disc
- May have extensive epidural, paravertebral extension

PATHOLOGY

General Features

- Etiology
  - Most common etiologic agent is *S. aureus*
  - Risk factors: Increasing age, diabetes, postoperative incontinence, posterior approach, surgery for tumor resection, morbid obesity, increased blood loss, longer operative times, number of operated levels, smoking
    - Elevated pre- and postoperative serum glucose associated with increased risk
  - Epidural catheter use → epidural abscess, osteomyelitis, psoas abscess
  - Scoliosis surgery
    - Postop closure of thoracolumbar wounds difficult due to tautness and lack of usable tissue
    - Resulting dead space containing metallic fixation devices is predisposed to infection
    - Presence of ventriculoperitoneal shunt increases infection risk for neuromuscular scoliosis (5%)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Variety of clinical symptoms; spontaneous drainage is most common
  - Symptoms and clinical findings are often nonspecific
  - High index of suspicion in any patient presenting with ↑ back pain after invasive spinal procedure
- Clinical profile
  - Abnormal erythrocyte sedimentation rate, C-reactive protein, and blood cultures
  - C-reactive protein is most sensitive clinical laboratory marker to assess presence of infection and effectiveness of treatment response
  - Many patients have fluctuant mass, localized drainage, or abscess
  - Abscesses or drainage material is typically contiguous with instrumentation and fusion mass
  - Clinical setting and presentation are key in diagnosis
    - 1-4 weeks following surgery, patient develops back pain, which may be severe
      - May radiate into abdomen, perineum, testes, legs

Demographics

- Age
  - Risk increases with age
- Epidemiology
  - Overall uncommon; 0.2-3% spinal surgeries
  - Postprocedural discitis: Incidence ~ 0.2%
  - Scoliosis surgery: Postoperative infection due to lack of vascularized tissue and presence of metallic hardware near the wound is common

Natural History & Prognosis

- Majority (90%) have good long-term outcomes
- Majority occur in early postop (< 3 months)

Treatment

- Biopsy to establish pathogen
- Effective treatment may include
  - Removal of implants, irrigation, and debridement
    - Post-discectomy/laminectomy infections confined to disc space can be treated with percutaneous transpedicle drainage
    - Epidural/paraspinal abscess → open debridement
    - Instrumentation can be retained safely in patients with infections that manifest during 1st month after implantation
      - Single surgical debridement in selected cases
      - Multiple debridements → granulation tissue in wound, ↓ CRP → effective
  - Intravenous organism-specific antibiotic treatment and spinal immobilization
- Prophylaxis
  - 1 preoperative administration of cefotiam has been shown to be effective for ↓ rate of postoperative wound infections in lumbar disc surgery
    - Rate of infection 0.2% after antibiotic administration vs. 2.8% without prophylaxis
  - Intraoperative antibiotic prophylaxis has been advocated by some
- Scoliosis surgery
  - Lower thoracic and thoracolumbar: Modified extended latissimus dorsi myocutaneous flap to close and supply blood to wounds
  - Allows for wound closure and drastically ↓ risk of postoperative infection
  - Effective for closure and increased blood supply to already infected wounds

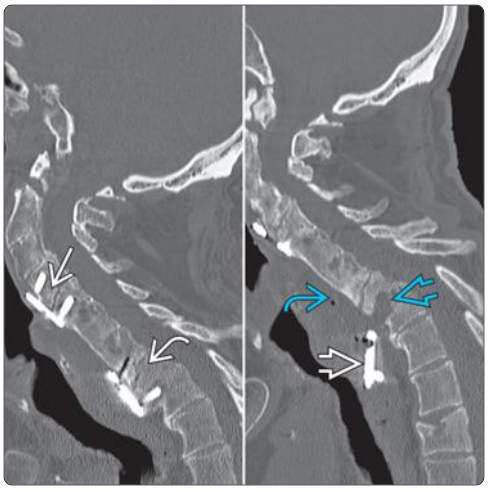
DIAGNOSTIC CHECKLIST

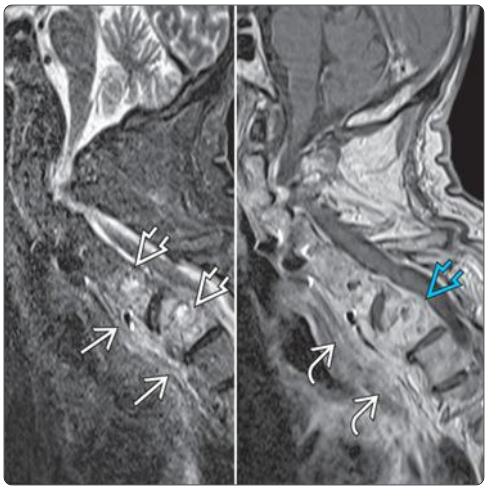
Image Interpretation Pearls

- 2 bands of enhancement in intervertebral disc paralleling endplates at surgical level ± type I endplate changes suggest **normal** postoperative changes
  - Pattern is different from amorphous enhancement seen with disc space infection

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(Left) *Initial CT shows C4-C5 and cervicothoracic ACDF. Later CT performed when the patient presented with dysphagia shows migrated lower cervical plate, soft tissue swelling, and gas. Note erosions in the fused vertebral bodies.* **(Right)** *Sagittal STIR MR from the same patient depicts prevertebral and retropharyngeal soft tissue edema as well as vertebral body marrow edema. T1WI C+ MR shows corresponding enhancement, compatible with phlegmon and osteomyelitis.*





(Left) *Sagittal CT demonstrates collapse of midthoracic vertebral bodies with focal kyphosis. Note the methylmethacrylate from vertebroplasty. Adjacent endplates show irregularity and erosive changes.* **(Right)** *Postprocedural epidural abscess is noted in the midthoracic spine. Collapse of midthoracic vertebral bodies with focal kyphosis. Methylmethacrylate from vertebroplasty appears as a signal void. Osteomyelitis is seen as diffuse avid enhancement of adjacent vertebral body.*





(Left) *Sagittal T1WI C+ FS MR shows rim-enhancing fluid collection in dorsal soft tissues extending from lower lumbar laminectomy site for tethered cord release. Moderate enhancement of neural placode posteriorly adherent to the thecal sac. Low-lying conus medullaris is noted.* **(Right)** *In this differential diagnosis, CT reconstruction of neurogenic arthropathy shows destruction of endplates, bony debris, and heterotopic bone. Involved vertebral bodies show increased sclerosis.*

Pseudomeningocele

KEY FACTS

TERMINOLOGY

- Dural dehiscence, pseudocyst, CSF leak
- Spinal fluid collection contiguous with thecal sac, not lined with meninges

IMAGING

- CSF-filled spinal axis fluid collection with supportive postoperative or post-traumatic ancillary findings
- Most common location is dorsal to thecal sac at lumbar laminectomy site

TOP DIFFERENTIAL DIAGNOSES

- Paraspinous abscess
- Postoperative hematoma
  - Does not follow CSF signal on all pulse sequences
- Extradural spinal meningeal cyst
  - Nabors type 1
- True meningocele
  - Disease or syndrome related such as Marfan or NF-1
- Plexiform neurofibroma

PATHOLOGY

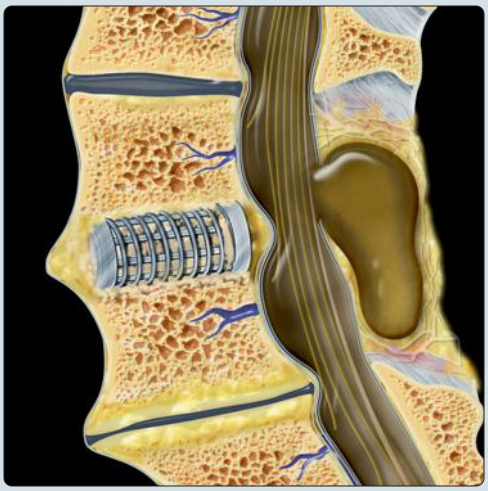
- Post-traumatic
  - Most commonly following cervical root avulsion
  - May also see with posterior element fractures + dural laceration
- Postsurgical

CLINICAL ISSUES

- Commonly asymptomatic
- Nonspecific back pain
- Headache if associated with CSF hypotension

DIAGNOSTIC CHECKLIST

- Classic imaging appearance: CSF signal/density collection contiguous with thecal sac in correct clinical context





(Left) *Sagittal graphic shows changes of L4-L5 interbody fusion with a cage. A large well-defined CSF collection extends into the dorsal soft tissue from the thecal sac from the site of operative dural tear.* **(Right)** *Sagittal T1WI MR shows a large CSF cyst at the surgical site extending dorsally from the dural sac into subcutaneous tissues. The CSF collection compresses the thecal sac at L4 and L5 levels.*





(Left) *Axial T2* MR shows CSF signal pseudomeningocele extending out the neural foramina, with septation separating the collections from the thecal sac, reflecting old trauma and nerve root avulsion.* **(Right)** *Sagittal T2WI MR in a patient with C2 fracture and traumatic anterior cervical pseudomeningocele shows CSF signal collection anterior to the cord and with cord displacement. The surgery was pseudomeningocele decompression with discectomy at C5-C6, followed by C0-C2 fusion.*

TERMINOLOGY

Synonyms

- Dural dehiscence, pseudocyst, CSF leak

Definitions

- Spinal fluid collection contiguous with thecal sac, not lined with meninges

IMAGING

General Features

- Best diagnostic clue
  - CSF-filled spinal axis fluid collection with supportive postoperative or post-traumatic ancillary findings
- Location
  - Most common location is dorsal to thecal sac at lumbar laminectomy site
  - Rare reports of intraosseous location (lamina/spinous process)
- Size
  - 1 to > 10 cm
- Morphology
  - Round, lobulated CSF attenuation/signal collection

Radiographic Findings

- Radiography
  - Nonspecific postoperative changes, such as laminectomy defect

CT Findings

- NECT
  - CSF-density cyst adjacent to thecal sac
  - Postoperative pseudomeningocele cyst usually oriented along operative approach
    - Dural connection is difficult to demonstrate without intrathecal contrast
  - Cervical root avulsions are anterolaterally oriented, contiguous with neural foramen, devoid of neural elements

MR Findings

- T1WI
  - CSF signal intensity cystic fluid collection
    - Note relationship to posterior fusion hardware
      - Fluid may surround posterior hardware
  - May be difficult to precisely define with presence of posterior fusion hardware/metal artifact
- T2WI
  - In postoperative cases, can frequently identify thecal sac communication using axial and sagittal T2-weighted imaging
- DWI
  - CSF signal intensity lesion, with no restricted diffusion
- T1WI C+
  - Does not enhance unless inflamed or infected; may see thin peripheral enhancement within 1 year of surgery
  - Spinal and intracranial dural thickening and enhancement in patients with symptomatic CSF hypotension

Ultrasonographic Findings

- Hypoechoic cyst

Nonvascular Interventions

- Myelography
  - Typical postoperative collection seen as round/lobulated contrast collection dorsal to thecal sac within subcutaneous tissues
    - ± ability to define precise point of communication with thecal sac
    - Note relationship to posterior fusion hardware
    - ± incorporated into CSF collection
    - Note extension of contrast to skin surface
  - Post-traumatic/postsurgical pseudomeningoceles usually devoid of neural elements
    - May see cord or nerve herniation into pseudomeningocele with severe dural laceration

Other Modality Findings

- While radionuclide cisternography can show activity within pseudomeningoceles, generally CT/MR preferred for direct visualization

Imaging Recommendations

- Fat-saturated T2WI best sequence to demonstrate pseudomeningocele and to localize dural communication
- Sagittal plane useful for diagnosis and to tailor thin-section axial imaging for definitive localization

DIFFERENTIAL DIAGNOSIS

Paraspinous Abscess

- Clinical history of critical importance, with supportive lab values (sedimentation rate, C-reactive protein)
- May be indistinguishable from postoperative pseudomeningocele

Postoperative Hematoma

- Does not follow CSF signal on all pulse sequences
- T2* images show susceptibility artifact ("blooming")

Extradural Spinal Meningeal Cyst

- Nabors type 1
- Dorsal thoracolumbar region typical
- Well defined, epidural location
- May show adjacent bone remodeling

Soft Tissue Tumor

- Cystic components of
  - Giant cell
  - Aneurysmal bone cyst
  - Osteosarcoma
- Easily distinguished from pseudomeningocele due to bone destruction, soft tissue component, enhancement

True Meningocele

- Lined with arachnoid
- Disease or syndrome related
  - Neurofibromatosis type 1
  - Marfan syndrome
  - Homocystinuria
  - Ehlers-Danlos syndrome
- No pertinent trauma or surgical history
- Often coexists with dural dysplasia

Plexiform Neurofibroma

- T2 hyperintense; T1 low signal, which may be as low as CSF
  - Neurofibroma not as bright as CSF
- Variable contrast enhancement
- "Target" sign common

PATHOLOGY

General Features

- Etiology
  - Post-traumatic
    - Most commonly following cervical root avulsion
    - May also see with posterior element fractures + dural laceration
  - Postsurgical
    - Iatrogenic dural laceration with CSF leak
    - May also see after dural graft following spinal tumor resection
- Genetics
  - None
- Associated abnormalities
  - May rarely show peripheral calcification/ossification
- CSF-containing cyst in contiguity with thecal sac
- Not lined by meninges
- Usually devoid of neural elements
- In some cases neural elements may herniate into defect

Gross Pathologic & Surgical Features

- CSF-containing collection

Microscopic Features

- No true meningeal covering
- Reactive fibrous tissue forms cyst capsule

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Commonly asymptomatic
  - Nonspecific back pain
  - Headache if associated with CSF hypotension
  - Occasionally extends superficially, palpable within subcutaneous tissues
  - Meningitis/abscess if extends to/communicates with skin surface
- Clinical profile
  - Adult patient with palpable subcutaneous mass overlying lumbar laminectomy site

Demographics

- Age
  - Adult or children
- Gender
  - M = F
- Epidemiology
  - 0.19-2% of patients undergoing lumbar laminectomies

Natural History & Prognosis

- May be asymptomatic
- Postoperative defects may close spontaneously if small but can require treatment
- Cervical root avulsion with pseudomeningocele usually leaves permanent neurological deficits

Treatment

- Closure of underlying dural defect when possible
  - Surgical repair for large defects
  - Subarachnoid catheter drainage occasionally successful
  - Blood patch may be effective for small defects

DIAGNOSTIC CHECKLIST

Consider

- Relationship of postoperative pseudomeningocele to patient pain is difficult
- Causes of pain related to pseudomeningocele include
  - Tension of cyst against nerve roots
  - Entrapment of nerve roots
  - Periradicular fibrosis

Image Interpretation Pearls

- Classic imaging appearance: CSF signal/density collection contiguous with thecal sac in correct clinical context
- May need to widen window on MR studies to visualize subcutaneous component due to surface coil signal inhomogeneity

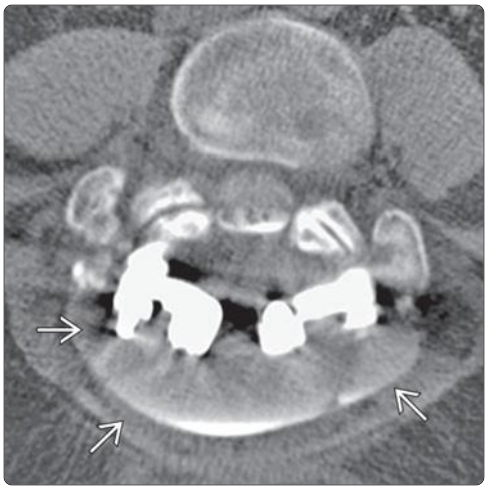
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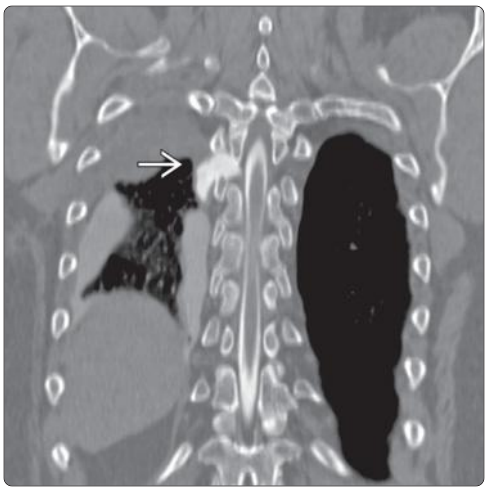
(Left) *Sagittal T2WI MR shows hyperintense fluid collection at L4-L5 levels. Focal low signal within the collection is flow dephasing from CSF movement into the collection at dural tear.* **(Right)** *Axial T2WI MR shows hyperintense fluid collection within dorsal soft tissues at the laminectomy site. There are scattered pieces of bone graft material within the fluid collection.*





(Left) *Axial CT postmyelography shows contrast collection at the laminectomy/fusion site in posterior soft tissues. There is incorporation of hardware into the collection.* **(Right)** *Axial T1WI MR shows ovoid CSF signal collection within dorsal soft tissues at the prior laminectomy site. There is a left pedicle screw causing artifact. Presence of infection cannot be determined by MR signal intensity of the fluid.*

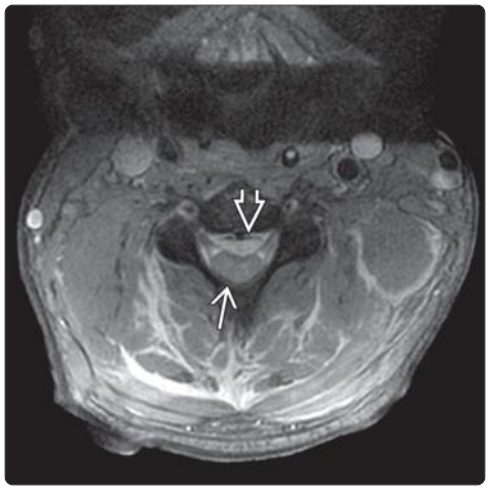




(Left) *Sagittal T2WI MR shows extensive hyperintense dorsal pseudomeningocele in this patient following multilevel lumbar laminectomy. There are multiple loculations and extension into subcutaneous tissues.* **(Right)** *Coronal postmyelography CT shows leakage of contrast from the thoracic thecal sac through the right T4-T5 neural foramen into the right pleural space. The patient required right laminectomy at T4 to repair the leak.*

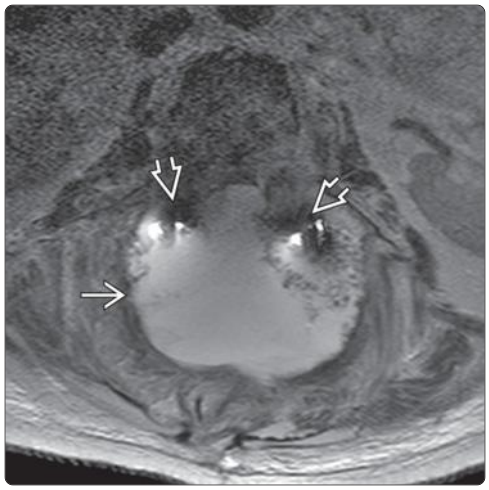
(Left) *Sagittal STIR MR shows disruption of the C2-C3 disc with a small traumatic protrusion. Inferior to the C2-C3 disc is increased fluid that posteriorly displaces the cord. This represents a traumatic dural tear with a ventral epidural CSF leak.* **(Right)** *Axial T2W MR shows fluid signal ventral to the cord that is posteriorly displaced in this trauma patient with C2 disruption. This finding reflects traumatic dural tear with epidural CSF leak mixed with hemorrhage.*



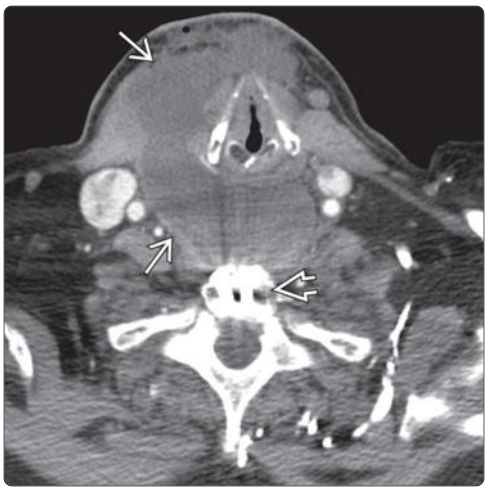


(Left) *Sagittal T2WI MR in this patient following pedicle subtraction osteotomy shows metal artifact at multiple levels from the pedicle screws. There is a large posterior operative site fluid collection (pseudomeningocele) surrounding the posterior hardware.* **(Right)** *Axial T2WI MR in this patient following multilevel fusion with pedicle screws shows a large postoperative soft tissue fluid collection contiguous to the hardware.*



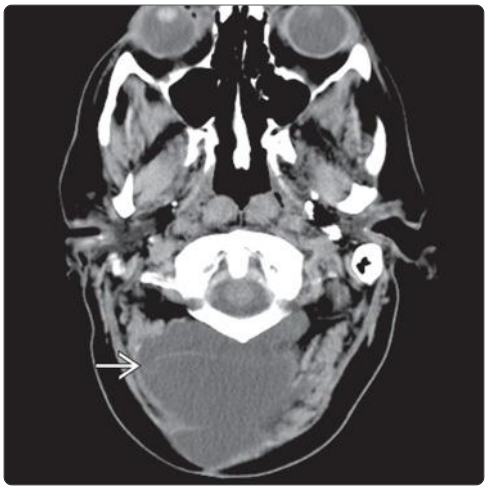


(Left) *Axial NECT shows a very large fluid collection within the right neck extending from the subcutaneous tissues to the anterior vertebral body between the carotid space and the trachea in this patient following multilevel ACDF.* **(Right)** *Sagittal T1WI MR 1 year following severe trauma shows sequelae of multiple lumbar root avulsions with the development of multiple CSF signal intensity pseudomeningoceles.*



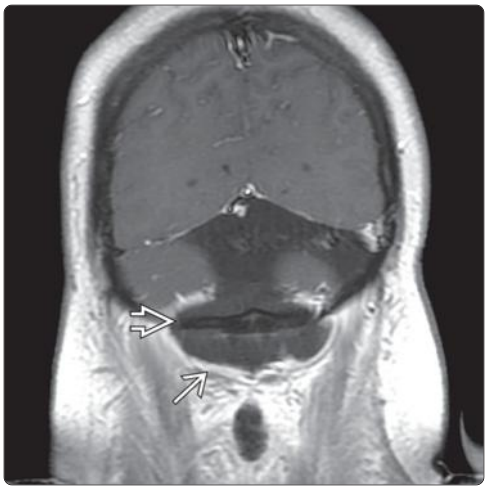




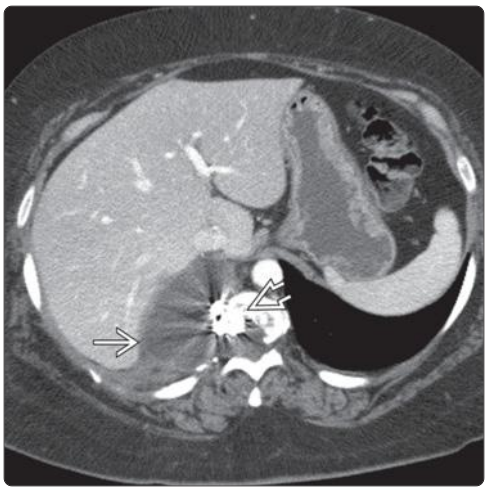


(Left) *Sagittal T2WI MR following suboccipital craniectomy for resection of cavernous malformation shows a large occipital and upper cervical pseudomeningocele.* **(Right)** *Axial CT shows the typical appearance of a pseudomeningocele within the upper dorsal cervical soft tissues following posterior fossa craniectomy.*





(Left) *Sagittal T1 C+ MR shows an irregular pseudomeningocele within the dorsal upper cervical soft tissues in this patient following occipital craniotomy for fenestration of a posterior fossa arachnoid cyst.* **(Right)** *Coronal T1 C+ MR shows CSF signal intensity on both sides of the operative site in this patient with a postoperative pseudomeningocele following occipital craniotomy.*





(Left) *Axial CT of the abdomen following thoracic corpectomy and discectomy shows a large CSF-attenuation pseudomeningocele in the right hemithorax which is contiguous to the operative site. Note the metal artifact from the cage.* **(Right)** *CT myelogram shows increased density of the mass consistent with a CSF collection communicating with the subarachnoid space. Metal artifact is present from the thoracic corpectomy and cage placement.*

CSF Leakage Syndrome

KEY FACTS

TERMINOLOGY

- Symptomatic leakage of CSF
- Postoperative, iatrogenic, or post-traumatic CSF leak, ± pseudomeningocele

IMAGING

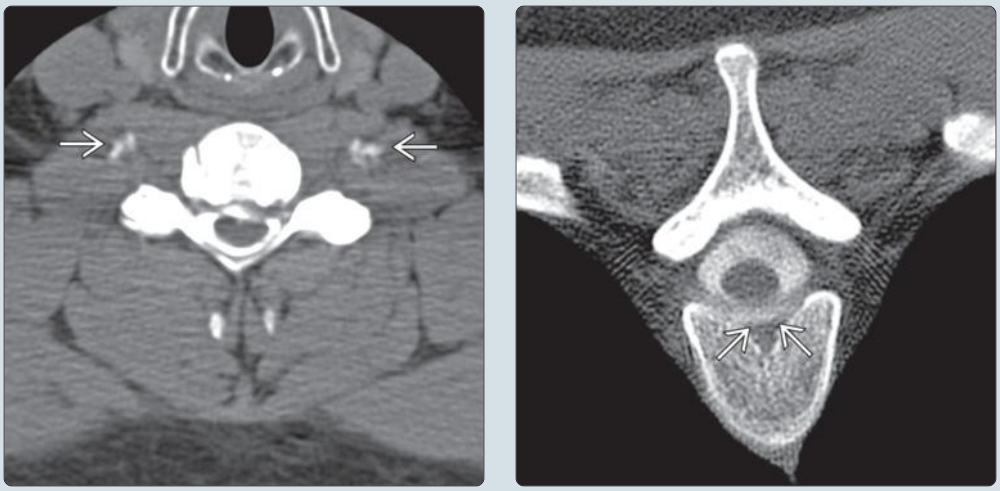
- Intracranial findings
  - Smooth intracranial dural thickening
  - "Sagging" midbrain
  - Subdural hygromas
  - Subdural hematomas
  - Descent of cerebellar tonsils
- Spinal findings
  - Spinal hygromas
  - Dilatation of anterior epidural venous plexus, dural thickening/enhancement
  - Paraspinal fluid
  - Symmetric anterolateral epidural masses (dilated epidural veins)
  - Nerve root sleeve cysts/diverticula
- Radionuclide cisternography (RNC)
  - Paucity or absence of radiotracer over cerebral convexities at 24-48 hours
  - Early washout
  - Early appearance of radionuclide in kidneys, bladder common
- Myelography/CT myelography
  - Demonstrate anatomic abnormalities (arachnoid diverticula)
  - Localize site of CSF leak
  - Variable performance relative to RNC

PATHOLOGY

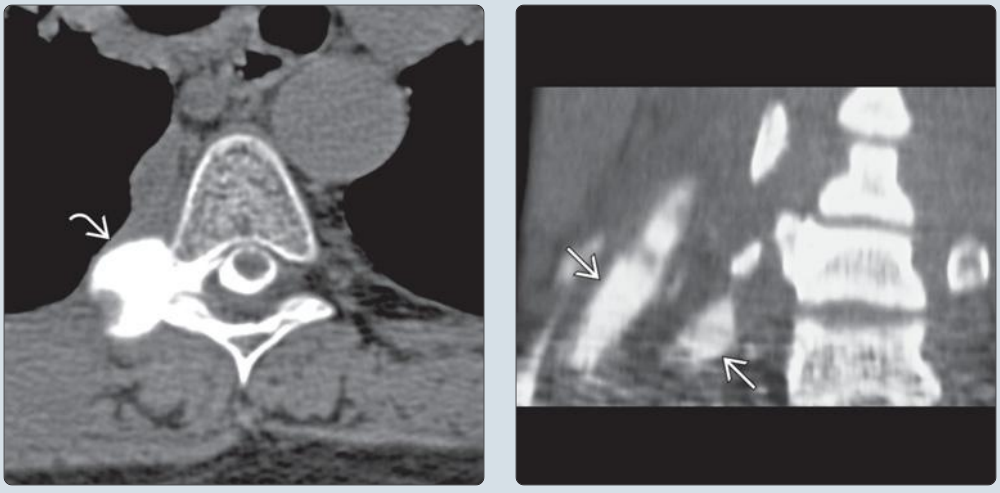
- Low CSF volume secondary to leakage or overdrainage

CLINICAL ISSUES

- Autologous blood patch is mainstay
  - 15-30 mL, lumbar route



(Left) *Axial CT myelography of the cervical spine shows leakage of contrast from the bilateral C5 nerve roots in this patient with spontaneous intracranial hypotension (SIH).* **(Right)** *Delayed CT myelography of CSF leakage syndrome shows leak of myelographic contrast into a ventral extraarachnoid fluid collection. The contrast was not present in this location on the initial scan.*



(Left) *CT myelography shows leakage of contrast along the right T4 nerve root into extraforaminal soft tissues.* **(Right)** *Coronal reformatted image from a bone CT of the cervical spine shows extravasation of myelographic contrast at 2 levels, extending through the perineural soft tissues of the brachial plexus.*

TERMINOLOGY

Definitions

- Symptomatic leakage of CSF
- Postoperative, iatrogenic, or post-traumatic CSF leak, ± pseudomeningocele

IMAGING

General Features

- Best diagnostic clue
  - Intracranial findings
    - Smooth intracranial dural thickening
    - Sagging midbrain
    - Subdural hygromas
    - Subdural hematomas
    - Descent of cerebellar tonsils
  - Spinal findings
    - Spinal hygromas
      - Extraarachnoid (subdural or epidural) CSF collections
      - Ventral or dorsal in canal
      - Close to or remote from site of leakage
    - Dilatation of anterior epidural venous plexus, dural thickening/enhancement
    - Paraspinal fluid

CT Findings

- NECT
  - Symmetric anterolateral epidural masses (dilated epidural veins)
  - Spinal hygromas
  - Nerve root sleeve cysts/diverticula
- CECT
  - May see "draped curtain" sign due to intensely enhancing, engorged epidural veins

MR Findings

- T2WI
  - Spinal hygromas
  - Nerve root sleeve cysts/diverticula
  - Dilated epidural venous plexus, increased incidence of flow voids
- T1WI C+
  - Enlarged, enhancing ventral epidural venous plexus

Other Modality Findings

- Radionuclide cisternography (RNC)
  - Paucity or absence of radiotracer over cerebral convexities at 24-48 hours
  - Early washout
  - Early appearance of radionuclide in kidneys, bladder common
  - Up to 60% demonstrate site of CSF leak
- Myelography/CT myelography
  - Demonstrate anatomic abnormalities (arachnoid diverticula)
  - Localize site of CSF leak
  - Variable performance relative to RNC
- MR myelography
  - Low dose intrathecal gadolinium chelate (0.5 cc, diluted in saline)
    - Off-label use
  - Allows for dynamic, multiplanar imaging of CSF leakage

Imaging Recommendations

- Scan brain 1st (look for intracranial findings of spontaneous intracranial hypotension [SIH])
  - Dural thickening, enhancement
  - "Sagging" midbrain
  - Tonsillar descent/herniation
  - Subdural hygroma
- Scan spine, search for actual leakage site only if
  - 2 technically adequate blood patches fail
  - Post-traumatic leak suspected
- Use RNC and CT myelography to search for sites of occult leakage
  - Or to differentiate CSF leak from other causes of postoperative fluid collections

DIFFERENTIAL DIAGNOSIS

DDX of Postoperative Fluid Collections

- Hematoma
- Empyema, abscess, phlegmon
- Pseudomeningocele

DDX of Enlarged Spinal Venous Plexi

- AVM
- Jugular vein thrombosis (collateral drainage)
- IVC thrombosis (collateral drainage)
- Venous engorgement above high-grade spinal stenosis

DDX of Dural Thickening, Enhancement

- Infection
  - Epidural abscess
    - Rarely occurs in absence of bone, disc involvement
- Neoplasm (posterior cortex, pedicle often destroyed/infiltrated)
- Miscellaneous (e.g., sarcoid)

PATHOLOGY

General Features

- Etiology
  - Low CSF volume secondary to leakage or overdrainage
    - Surgery or trauma (including trivial fall)
    - Vigorous exercise, violent coughing
    - After lumbar puncture (postdural puncture headache [PDPH])
    - Abnormal dura (e.g., Marfan syndrome)
    - Ruptured arachnoid diverticulum
    - Severe dehydration
- Genetics
  - 20% of patients with spontaneous CSF leaks have minor skeletal features of Marfan syndrome
- Associated abnormalities
  - Intracranial subdural hematoma
  - Marfan syndrome
    - Defect in dural sac
    - May be ruptured nerve sleeve or its diverticulum

Gross Pathologic & Surgical Features

- Fluid collection associated with dural defect

Microscopic Features

- Thick dura with fibrosis, numerous dilated thin-walled vessels
  - Occasionally show nonspecific inflammation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Postural headaches, unrelenting
  - Lumbar puncture shows low CSF pressure ± pleocytosis, increased protein
  - Other signs/symptoms
    - Unilateral or bilateral CN6 palsy, other visual changes
    - Posterior neck and interscapular pain, radicular pain
    - Nausea, vomiting
    - Rare: Obtundation, death
- Clinical profile
  - Onset of postural headache after lumbar puncture or spinal surgery

Demographics

- Epidemiology
  - M:F = 1:2 (spontaneous)
  - Peak age = 30-40 years

Natural History & Prognosis

- 75% resolve spontaneously within 3 months (dural thickening, venous engorgement disappear; fluid collections resorbed)
- 20-25% persistent leak, chronic headaches
- Generally excellent
- Rare: Coma, death from intracranial herniation

Treatment

- Conservative treatment to restore CSF volume (bed rest, hydration, caffeine/theophylline)
- Other
  - Autologous blood patch is mainstay
    - 15-30 mL, lumbar route
      - Repeat with higher volume if initial failure
    - Targeted blood patch at documented leak site shows higher success rate
    - Targeted fibrin glue placement may also be used for failed blood patch cases
  - Epidural saline infusion (uncommon)
  - Surgery if large dural tear, ruptured diverticulum, or Tarlov cyst
- Postoperative
  - Treatment includes repeated LPs or CSF diversion (lumbar drain) to decrease size

DIAGNOSTIC CHECKLIST

Consider

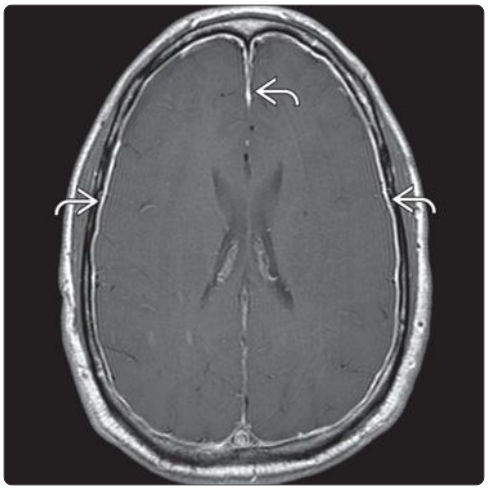
- Intracranial cisternography to look for congenital defects in middle fossa or temporal bone
- Rare reported cases of rebound intracranial **hypertension** following epidural blood patch placement

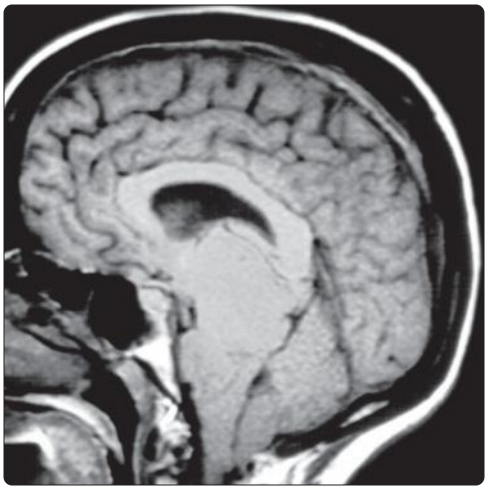
Image Interpretation Pearls

- "Sagging" brain, posterior fossa with diffuse dural thickening in patient evaluated for headache should suggest CSF leak

SELECTED REFERENCES

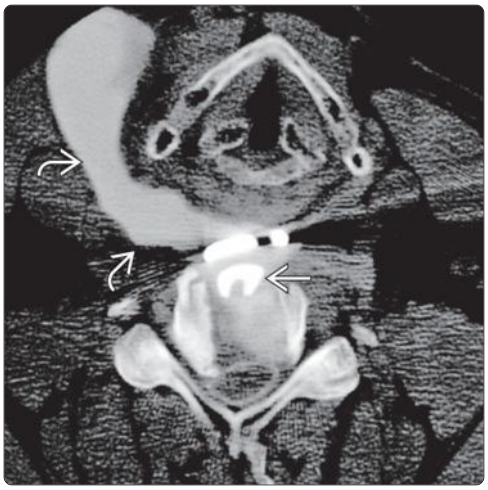
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(Left) *Axial T1 C+ MR of the brain shows diffuse, smooth dural thickening and enhancement in this patient with intracranial hypotension.* **(Right)** *Sagittal T1 MR shows "settling" of the midbrain, with flattened pons, loss of basilar cisterns, and herniation of the cerebellar tonsils through the foramen magnum. There is elongation of the 4th ventricle.*





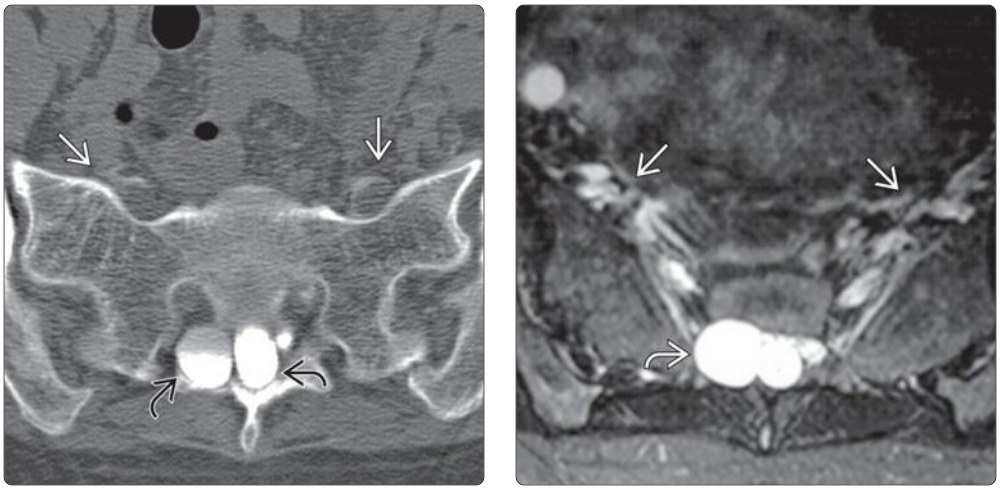
(Left) *Axial enhanced T1 C+ MR through the posterior fossa again shows diffuse dural thickening and enhancement, extending even into the internal auditory canals. Settling of the brainstem effaces the prepontine and CP angle cisterns.* **(Right)** *Axial CT myelogram shows changes of anterior cervical fusion, with a strut graft and metallic plate. There is a large volume of extravasated myelographic contrast through a dural rent into the deep tissue of the right neck.*



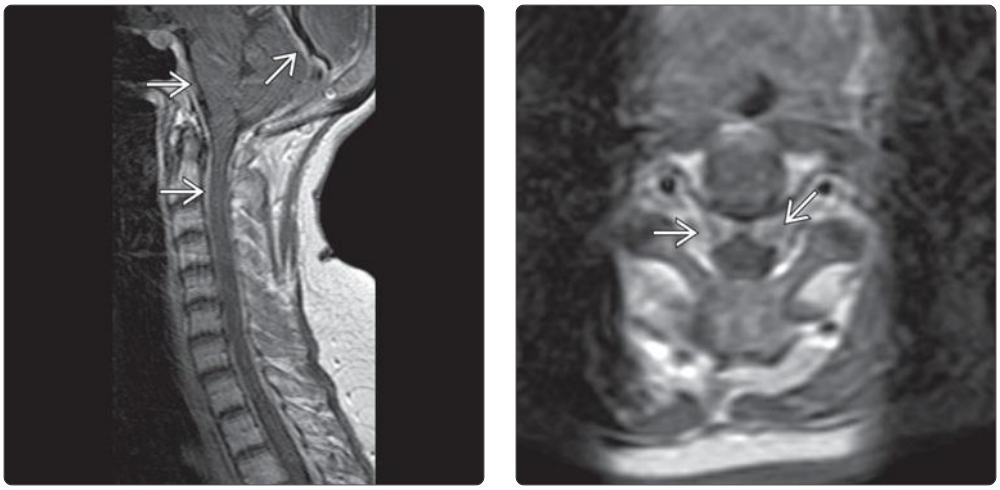


(Left) *Sagittal post-contrast T1WI of the cervical spine shows dilated, enhancing ventral epidural venous plexus and a small retrospinal fluid collection at the C1-C2 level.* **(Right)** *Lateral myelography of the cervical spine in the same patient shows extravasation of the myelographic contrast into the C1-C2 retrospinal fluid collection, demonstrating the site of an active CSF leak.*

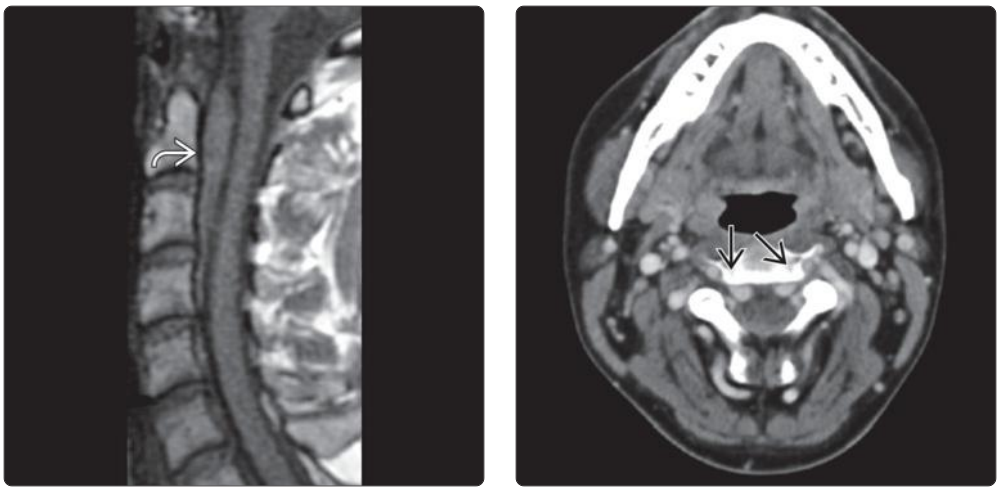
(Left) *Axial CT myelography through the level of the lower sacral canal shows multiple sacral nerve root cysts filling with contrast. There is extravasation of contrast along the sacral nerve roots into the pelvis.* **(Right)** *Axial fat-saturated T2WI MR in the same patient shows the sacral nerve root cysts, as well as the fluid leaking along the sacral nerve roots into the pelvis.*

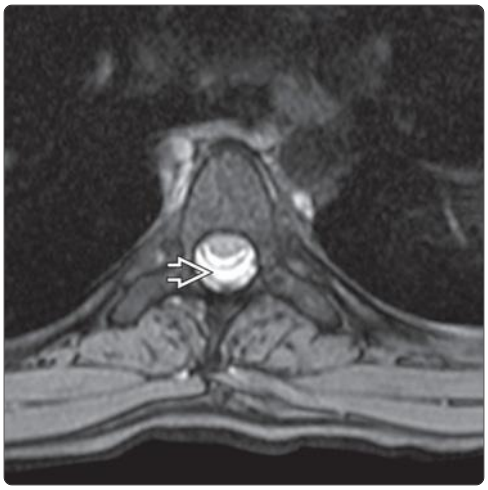


(Left) *Sagittal contrast-enhanced T1WI in a patient with a caudal CSF leak shows diffuse dural thickening and enhancement in the cervical spine, tentorium, and along the clivus.* **(Right)** *Axial contrast-enhanced T1WI in the same patient shows enhancing anterolateral epidural "masses", representing dilation of the cervical venous plexus due to CSF leakage syndrome.*



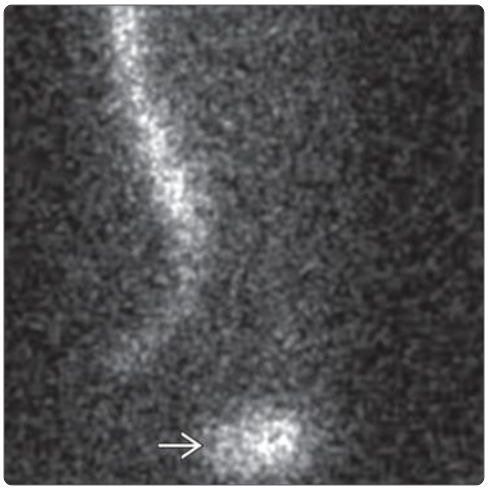
(Left) *Sagittal T1 MR of the cervical spine shows marked dilation of the ventral epidural venous plexus of the cervical spine due to intracranial hypotension.* **(Right)** *Axial CECT of the upper neck shows dilated, enhancing epidural venous plexus in this patient with intracranial hypotension due to CSF leak.*

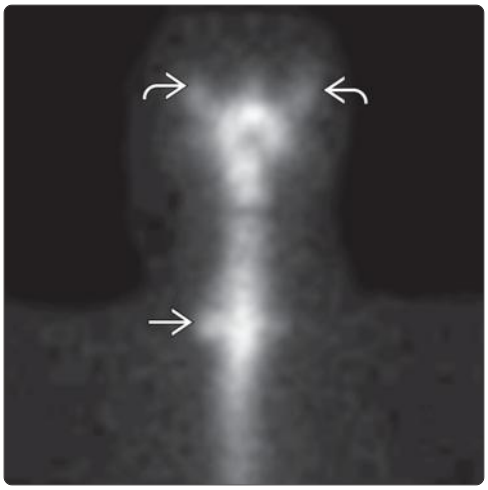




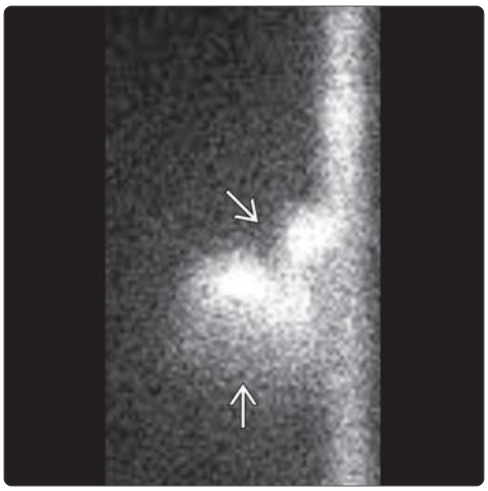


(Left) *Axial T2* GRE MR of the thoracic spine shows a dorsal fluid collection in the spinal canal, which appears to be in the subdural space, flattening the thecal sac. This patient had a disabling headache that persisted nearly 1 week following lumbar puncture.* **(Right)** *Sagittal T2WI MR in the same patient shows subdural hygroma extending from the cervicothoracic junction to the upper lumbar spine. Slightly increased protein concentration may account for mild hyperintensity to CSF.*





(Left) *Patient with SIH imaged after intrathecal injection of In-111 DTPA. Lateral projection 4 hours after injection shows early appearance of activity in the bladder, an indirect sign of CSF leak.* **(Right)** *Image of the same patient, 24 hours after injection, shows amorphous accumulation of tracer around the right T1-T2 level at the site of CSF leak from a dural diverticulum. Smaller diverticulum is seen on the left. Poor ascent of radiotracer over the convexities is an indirect sign of CSF leak.*





(Left) *Radionuclide cisternography shows a large amount of tracer extravasated from the thoracic spine. This patient had severe postural headaches and was discovered to have suffered a dural injury during recent thoracotomy.* **(Right)** *Delayed axial CT myelogram in the same patient shows extravasation of myelographic contrast into a right pleural effusion corresponding to the large collection of radiotracer. The patient eventually required open surgical repair of the dural defect.*

Postsurgical Deformity

KEY FACTS

TERMINOLOGY

- Deformity: Abnormality of alignment, angulation, or shape of vertebral column following surgery
  - May be associated with clinically significant findings such as instability

TOP DIFFERENTIAL DIAGNOSES

- Infection
- Tumor
- Degenerative instability
- Isthmic spondylolisthesis

PATHOLOGY

- Risk factors for deformity
  - Age (pediatric at increased risk), number of levels
  - Location of laminectomy
  - Intramedullary disease
  - Facet joint involvement
- Preexisting degenerative disease risk factor for cervical deformity
  - 30% incidence of kyphosis in patients with straight spine
- Thoracolumbar junction
  - Particularly susceptible to deformity
- Spinal deformity reoperation complications as high as 33%
  - Implant failure
  - Adjacent segment degeneration (10%)
  - Pseudoarthrosis

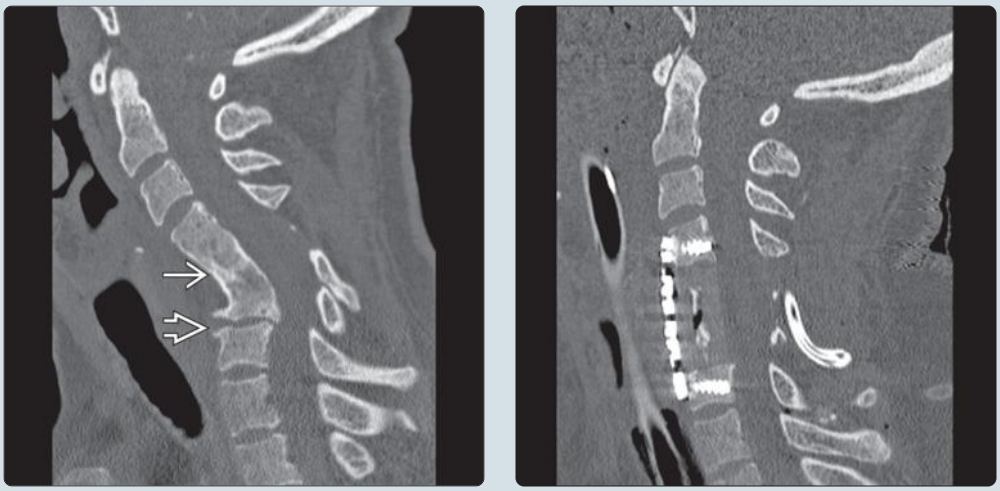
CLINICAL ISSUES

- Surgery for progressive neurological decline, intractable pain, cosmetic appearance

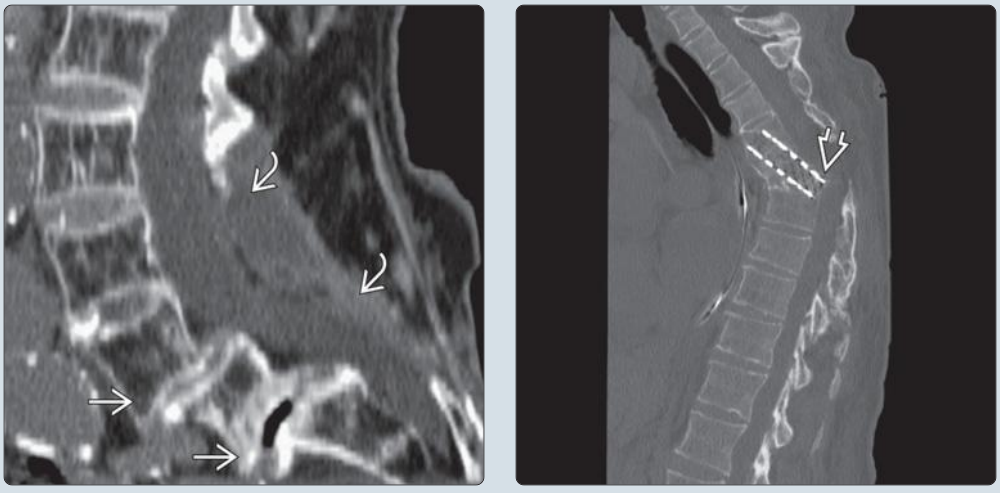
DIAGNOSTIC CHECKLIST

- Thresholds for optimal correction of adults deformity include sagittal vertical axis (SVA) < 50 mm, pelvic tilt (PT) < 25°, and pelvic incidence-lumbar lordosis (PI-LL) < 10°

(Left) *Sagittal NECT shows severe kyphotic deformity at site of anterior fusion ➡ at C5-C6 and adjacent segment degeneration ➡ at C6-C7. There had been prior laminectomy and posterior fusion.* **(Right)** *Sagittal NECT shows near normal alignment after 2 stages of surgery. The 1st stage was posterior osteotomies; the 2nd stage was anterior vertebrectomies with strut graft with plate/screws. The 3rd stage was posterior instrumented fusion (not shown).*



(Left) *Sagittal NECT shows severe multilevel spondylolisthesis ➡ with multilevel laminectomy defect ➡.* **(Right)** *Sagittal CT shows increased kyphotic angulation in this patient following posterior laminectomy and corpectomy. Note the angulation of the cage with the displacement of the posterior superior margin into the canal ➡.*



TERMINOLOGY

Synonyms

- Iatrogenic deformity, iatrogenic instability, postsurgical instability

Definitions

- Deformity: Abnormality of alignment, angulation, or shape of vertebral column following surgery
  - May be associated with clinically significant findings such as instability
- Instability: Postoperative loss of spine motion segment stiffness, in which applied force produces greater displacement than normal, with pain/deformity

IMAGING

General Features

- Best diagnostic clue
  - New postoperative deformity that increases with motion and increases over time
- Location
  - Any spinal motion segment
  - Deformity most commonly occurs involving cervical spine in children, operated upon for intramedullary cord disease
- Size
  - Displacement may vary from few mm to width of vertebral body
- Morphology
  - Displacement or angulation of vertebral body with respect to adjacent body

Radiographic Findings

- Radiography
  - Various parameters used for degenerative **instability** by plain films
    - Dynamic slip > 3 mm in flexion/extension
    - Static slip of ≥ 4.5 mm
    - Angulation > 10-15° suggests need for surgical intervention

Fluoroscopic Findings

- Increased motion with flexion/extension or translation

CT Findings

- NECT
  - Nonspecific postoperative changes with malalignment

MR Findings

- T1WI
  - Anterolisthesis, retrolisthesis, lateral translation
- T2WI
  - Loss of disc signal, disc space height in adult with disc degeneration
- T1WI C+
  - In adult, nonspecific enhancement of disc due to degenerative disc disease

Imaging Recommendations

- Best imaging tool
  - Flexion/extension plain films

DIFFERENTIAL DIAGNOSIS

Infection

- Endplate destruction, disc T2 hyperintensity

Tumor

- Enhancing soft tissue mass destroying vertebral body/posterior elements

Degenerative Instability

- Marked degenerative disc disease with intact endplates, no soft tissue mass

Isthmic Spondylolisthesis

- Lysis involving pars interarticularis
- Progressive L5-S1 deformity in child (rarely unstable in adults)

PATHOLOGY

General Features

- Etiology
  - Postoperative alteration in spinal biomechanics
  - Post-laminectomy cervical kyphosis > 40% in children
  - Adult change in alignment > 20% but < 40% (but may not be clinically significant)
    - Dependent upon age, spinal levels treated, and surgery
  - Risk factors for deformity
    - Age (pediatric at increased risk)
    - Number of levels treated
    - Location of laminectomy
      - Decompression at higher levels at increased risk
    - Intramedullary disease
    - Prior radiation therapy
    - Facet joint involvement
  - Flat back syndrome
    - Symptomatic forward fixation of trunk after scoliosis surgery
      - Loss of lumbar lordosis with anterior translation of vertical axis and center of gravity
      - Rigid imbalance compensated by hyperextension of thoracolumbar segments not in fusion
      - Extension of cervical spine and flexion at hips
      - Incidence as high as 49%, 25% requiring reoperation
  - Preexisting degenerative disease risk factor for cervical deformity
    - 30% incidence of kyphosis in patients with straight spine
    - Laminoplasty does not appear to improve rate of deformity for cervical spine
  - Thoracolumbar junction
    - Particularly susceptible to deformity
    - Transition from costal support of thoracic vertebrae to unsupported lumbar spine
- Resection of ≥ 50% of both facets alters segmental stiffness
- Plain films show new or worsening segmental motion

Staging, Grading, & Classification

- Spinal deformity reoperation complications as high as 33%

- Early complications include
  - Neurologic deterioration (2%)
  - Wound infection (1-8%)
  - CSF leak
  - Thromboembolic events
- Late complications include
  - Adjacent segment degeneration (10%)
  - Implant failure
  - Junctional kyphosis cephalad to instrumented fusion
  - Pseudoarthrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neck, back pain with later appearance of new neurologic symptoms
    - Pain may be radicular or related to facet arthropathy
  - In adolescents with scoliosis, treatment of deformity occurs without symptomatology
  - In adults with deformity, pain and disability determine treatment modalities more than deformity itself
  - Adults
    - Evaluation of sagittal alignment is critical when planning surgery
    - Regional measurements
      - Thoracic kyphosis (TK), lumbar lordosis (LL), and cervical lordosis
      - Thoracic kyphosis is angle between upper endplate of T4 and inferior endplate of T12
      - Lumbar lordosis is angle between upper endplate of L1 and superior endplate of S1
    - Global measurements
      - Sagittal vertical axis (SVA)
      - SVA is linear offset from C7 plumbline to posterosuperior corner of S1
    - Pelvic measures such as pelvic incidence (PI), pelvic tilt (PT), and sacral slope (SS)
      - PT is the angle between vertical and the line connecting the midpoint of superior sacral endplate to center of femoral heads
    - SVA and PT were the most correlated with health related quality of life scores
- Clinical profile
  - Early good operative result with progressive axial spine pain, new neurologic symptoms

Demographics

- Age
  - Children and adults
- Gender
  - No gender predilection
- Epidemiology
  - Predisposing factors for postoperative instability
    - Children > adults
    - No preoperative lordosis in cervical spine
    - Surgical disruption of facets
    - C2 laminectomy ± increased risk
    - More laminectomy levels ± increased risk

Natural History & Prognosis

- Generally progressive

Treatment

- Mild deformity treated conservatively
- Medical management includes hip extension, back extension exercises; bracing; NSAIDs
- Surgery for progressive neurological decline, intractable pain, cosmetic appearance
  - Anterior correction via distraction with discectomy or vertebrectomy
  - Subsequent posterior instrumentation to strengthen construct and reduce graft complications
  - Posterior osteotomies allows kyphotic deformity correction
- Prophylactic fusion considered for patients at risk of postoperative kyphosis
  - Prevention of deformity easier than treatment
  - Facet fusion and lateral mass fixation of cervical spine

DIAGNOSTIC CHECKLIST

Consider

- Wide range of normal curvature
  - Normal cervical spine has lordotic curve of 14-35°
    - Any cervical kyphosis is abnormal
  - Thoracic kyphosis
    - 37-42° (superior T3-T4 to inferior T12)
  - Lumbar lordosis
    - 50-75° (superior L1 to superior S1)
- Spinal constructs should not end at apex of a curve
  - Shorter fusions ending near apex lead to junction kyphosis
  - Should not end at thoracolumbar junction (T12, T11)
- Thresholds for optimal correction of adults deformity include SVA < 50 mm, PT < 25°, and pelvic incidence-lumbar lordosis (PI-LL) < 10°

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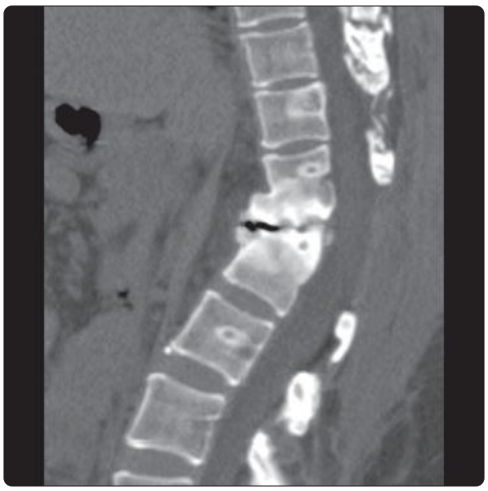


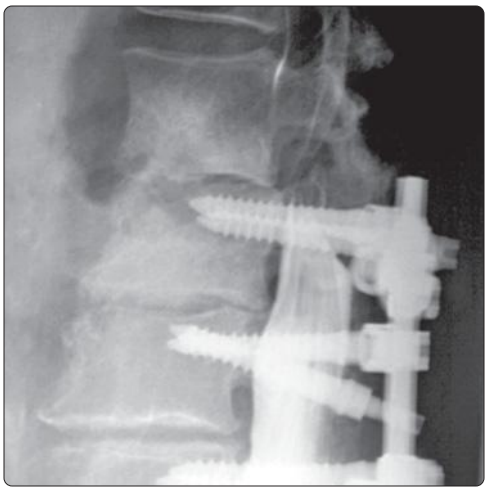
(Left) *Lateral extension radiograph in this patient with wide laminectomy, which destabilized the spine at L3-L4, shows grade 1 anterolisthesis ⇨ that worsened with flexion.* **(Right)** *Flexion lateral radiograph shows worsening grade 2 anterolisthesis at L3-L4 ➡ in this patient with wide laminectomy that destabilized the lumbar spine.*





(Left) *Sagittal T1WI MR shows postoperative changes of the posterior spinal elements at L4 and L5 following laminectomy. There are advanced degenerative changes of the L4-L5 intervertebral disc space with anterior subluxation of L4 on L5 ➡. Note also endplate compression fracture of L1 ➡.* **(Right)** *Sagittal T2WI MR shows postoperative spondylolisthesis following laminectomy at L4-L5 with severe disc degeneration and fatty endplate change. Note unrelated compression deformity of L1 ➡.*



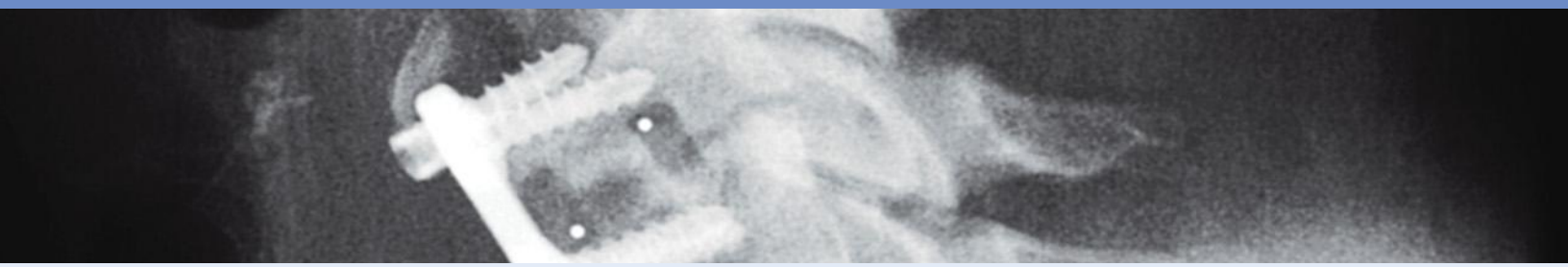


(Left) *Sagittal NECT reformat shows increased kyphotic deformity and pseudoarthrosis at the thoracolumbar junction following multilevel laminectomy for intramedullary ependymoma.* **(Right)** *Sagittal myelography shows anterolisthesis of the vertebral body at the upper margin of lumbar fusion with accelerated disc degeneration and severe central canal stenosis.*

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PART VII
SECTION 2

Hardware



Metal Artifact	**1106**
Occipitocervical Fixation	**1110**
Plates and Screws	**1112**
Cages	**1116**
Interbody Fusion Devices	**1118**
Interspinous Spacing Devices	**1122**
Cervical Artificial Disc	**1126**
Lumbar Artificial Disc	**1130**
Hardware Failure	**1134**
Bone Graft Complications	**1140**
rhBMP-2 Complications	**1144**
Heterotopic Bone Formation	**1148**

Metal Artifact

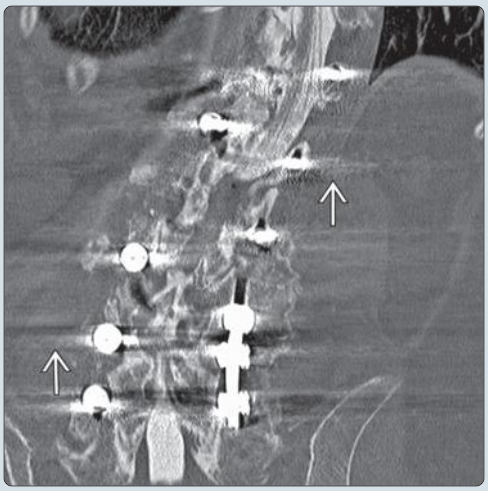
KEY FACTS

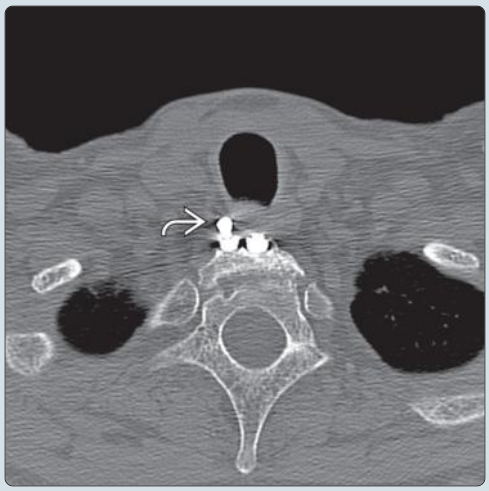
TERMINOLOGY

- CT: Beam-hardening artifact and blooming artifact
- MR: Magnetic susceptibility artifact
- Image degradation related to metal prostheses/implants

IMAGING

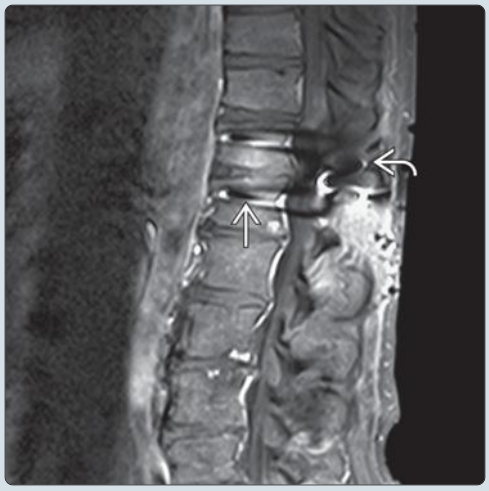
- CT: Artifact from metal hardware related to image reconstruction algorithm, tube current, x-ray kilovolt peak, pitch, hardware composition, shape, and location
  - Beam-hardening artifact is dark banding between dense objects, such as bone
  - Bloom artifacts result of partial volume effects or areas of photon starvation propagated by high-density structures, such as metal, within scanned object
  - Materials with lower x-ray attenuation coefficients produce less artifactual distortions
  - Reduce metal-related artifacts
    - Thick sections, lower kernel values (similar to standard reconstruction algorithm), and extended CT scale
- MR: Susceptibility artifact due to geometric distortion + signal loss secondary to dephasing
  - General MR methods to minimize metal artifact
    - Fast spin-echo better than conventional spin-echo, which is better than gradient-echo
    - Larger field of view better
    - Smaller voxel size better
    - Increase transmit and receive bandwidth
    - Frequency encoding direction along long axis of hardware
    - Lower magnetic field strength
    - Frequency selective fat saturation yields poor image quality with metal implants
      - STIR sequences are alternative method of fat suppression and less dependent on homogeneity of main magnetic field





(Left) *Coronal CT reconstruction illustrates bloom hardening artifact. Beam artifacts are a result of partial volume effects, or underranging, caused by areas of photon starvation propagated by high-density structures, such as metal, within the scanned object. When subtle, these artifacts appear as shading and, when severe, as high intensity streaks and areas of photon starvation.* **(Right)** *Axial NECT shows star artifact along the margins of the right corporal screw, which has migrated out.*





(Left) *Differences in magnetic susceptibilities of adjacent tissues and implants create local magnetic field inhomogeneities. Spins are mapped to an erroneous location within image giving distortion of the shape of the metallic object along the frequency encoding and section selection axes, and loss of signal.* **(Right)** *Magnetic susceptibility artifact around pedicle screws is seen. Mismapping of a disproportionate number of spins results in hyperintense rim around metallic object.*

TERMINOLOGY

Synonyms

- CT: Beam-hardening artifact and blooming artifact
- MR: Magnetic susceptibility artifact

Definitions

- Image degradation related to metal prostheses/implants
- Magnetic susceptibility
  - Partial magnetization of material in presence of applied external magnetic field
  - Nonferromagnetic metals may produce local electrical currents induced by changing scanner magnetic field
  - Tissues with greatly different magnetic susceptibilities in uniform magnetic field leads to
    - Difference of susceptibilities causes distortion in magnetic field → distortion on MR image
    - Magnetic susceptibility artifact consists of 2 additive components
      - Geometric distortion + signal loss secondary to dephasing

IMAGING

General Features

- CT: Artifact from metal hardware related to image reconstruction algorithm (filter)
  - Tube current (in mA)
  - X-ray kilovolt peak and pitch
  - Hardware composition, shape, and location
  - Polychromatic nature of x-ray beam leaving x-ray tube housing combined with eliminating lower energy photons from beam produce beam-hardening artifacts
    - Presents as dark banding between dense objects, such as bone
  - Partial volume effects or underranging due to areas of photon starvation propagated by high-density structures, such as metal → bloom artifacts
    - Subtle → shading; severe → high intensity streaks and areas of photon starvation
    - Emanate from attenuation at sites of orthopedic implants, surgical staples and clips, and calcium deposits
  - Metal causes severe x-ray attenuation with missing data in selected planes
  - Missing data or hollow projections cause classic "starburst" or streak artifacts during image reconstruction
  - Materials with lower x-ray attenuation coefficients produce less artifactual distortions
    - Plastic (best) < titanium < < tantalum < stainless steel < cobalt chrome (worst)
  - Metal composition, mass, orientation + position of implant are important factors that determine magnitude of image artifact
- Trade-off must occur in choice of metal
  - Titanium wires exhibit least artifact on CT when compared to cobalt chrome or stainless steel, but are more susceptible to failure
  - Titanium screws + cages produce fewer artifacts than tantalum, but may not have desirable biologic properties
- Use of high peak voltage (kilovolts peak), high tube charge (milliampere-seconds), narrow collimation, and thin sections helps reduce metal-related artifacts
  - Caution should always be exercised, particularly in children, young adults, and patients undergoing multiple examinations
  - Cone beam artifacts caused by geometry of multichannel CT scanners ↓ by using narrower x-ray beam collimation and low-pitch setting
- Reduce metal-related artifacts
  - Thick sections, lower kernel values (similar to standard reconstruction algorithm), and extended CT scale
- MR: Potential safety and biologic considerations
  - Stainless steel is safe, but produces severe artifacts (especially with low nickel content)
  - Titanium = tantalum implant artifact; much less relative to stainless steal
- MR general methods to minimize metal artifact
  - Fast spin-echo > conventional spin-echo > gradient-echo
  - Larger field of view
  - Increase transmit bandwidth
    - Increased specific absorption rate
  - Increase receive bandwidth
    - Decreased signal to noise ratio (SNR)
  - Smaller voxel size
  - Frequency encoding direction should be chosen along long axis of hardware (so artifact projects on hardware)
  - Lower magnet field strength
  - Short inversion time inversion recovery (STIR) imaging is effective alternative method of fat signal suppression and is less dependent on homogeneity of main magnetic field
- Location
  - Intervertebral disc level related to fusion cages, anterior plates + screws, iatrogenic metal
  - Pedicles related to pedicle screws
  - Dorsal elements related to dorsal stabilization rods, interspinous process wiring
- Size
  - Variable
- Morphology
  - Central low signal, with indistinct margins, spatial mismapping of signal giving peripheral curvilinear high signal

Radiographic Findings

- Radiography
  - Visualize hardware malposition and alignment

CT Findings

- NECT
  - Missing data from metal attenuation cause classic "starburst" or streak artifacts

MR Findings

- T1WI
  - Focal central signal loss with peripheral "halo" of ↑ signal related to spatial mismapping
- T2WI
  - Focal central signal loss with peripheral "halo" of ↑ signal related to spatial mismapping
  - Artifact minimized with FSE technique

- T2* GRE
  - Blooming of susceptibility artifact with gradient-echo techniques, worse with increasing echo time

Nonvascular Interventions

- Myelography
  - May be necessary if extensive hardware precludes adequate MR examination
  - Fluoroscopic positioning will obtain most favorable projection with overlapping hardware

Imaging Recommendations

- Best imaging tool
  - MR best sequence choice: Fast spin-echo > conventional spin-echo > gradient-echo
- Protocol advice
  - CT: Thin-section spiral imaging has improved quality compared to conventional discrete slices
  - MR: Optimum sequence should not contain gradient-echoes
    - Preferably FSE technique
    - Optimum FSE should maintain short echo spacing (short echo train less critical)
    - Single shot FSE sequences with half-Fourier (HASTE) useful
    - Hybrid imaging sequences that use both gradient-echo and spin-echo components should not be used
    - Frequency selective fat saturation yields poor image quality with metal implants
    - Orienting frequency encoding direction along long axial of pedicle screws minimizes artifact (except in area just beyond tip of implant)

DIFFERENTIAL DIAGNOSIS

Bone/Osteophyte

- Well-defined low signal on all sequences; may show fatty marrow ↑ T1 signal

Gas

- No protons, no signal
- Iatrogenic epidural or subarachnoid gas
- Vacuum phenomenon with degenerative disc disease

Hemorrhage

- Low signal deoxyhemoglobin on T2WI

Disc Herniation

- Desiccation or calcification giving low signal
- Gas from adjacent vacuum phenomenon

PATHOLOGY

General Features

- Etiology
  - In anterior cervical discectomies, sufficient metals to produce artifacts are deposited by contact of metal drill bits + suction tips
  - Post cervical discectomy/fusion, metallic susceptibility artifacts are produced by microscopic amounts of nickel, copper, and zinc

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Typically asymptomatic, ancillary finding of surgical procedure

Demographics

- Age
  - Any age
- Gender
  - No gender predilection
- Epidemiology
  - 5% of postoperative cervical discectomy cases have sufficient metal artifact to obscure thecal sac

DIAGNOSTIC CHECKLIST

Consider

- Following anterior cervical discectomy/fusion, small amount of metal artifact is typical at fusion site
  - Contact between drill burr and suction tip produced artifacts from metal flakes in soft tissues
- MR pedicle screw artifact size correlates with reduction in ratio of field of view to number of pixels in frequency encoding direction

Image Interpretation Pearls

- Minimize pedicle screw artifact by orienting frequency encoding gradient parallel to screw long axis, using FSE technique
- Slice thickness 3-4 mm is adequate, thinner sections yield little artifact reduction

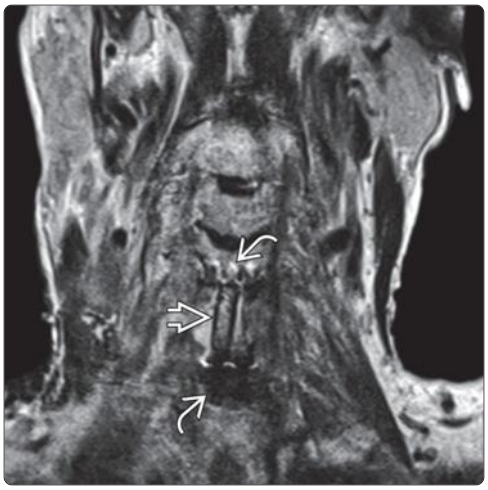
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(Left) *Mild artifact related to an artificial cervical disc prosthesis is shown. Misregistration effect is especially exaggerated in the direction of frequency encoding.* **(Right)** *This patient shows susceptibility artifact from vertebral cage. Factors influencing production of metal-related artifacts include composition (nonferromagnetic means less severe artifacts), size (larger produce more obtrusive artifacts), and orientation of metallic object with regard to the direction of the external magnetic field.*





(Left) *C5 corpectomy with a fibular strut graft shows no susceptibility artifact. Corporal screws at adjacent levels have geometric distortion. The artifact size increases proportionally with an increase in the angle between the long axis of the screw and the main magnetic field direction.* **(Right)** *Suboccipital craniectomy with occipitocervical fusion plate is shown. Small field of view, high-resolution matrix, thin section, and high gradient strength can help reduce metal-related artifacts.*





(Left) *On SE/FSE susceptibility artifact (signal loss and distortion) projects along frequency encoding direction.* **(Right)** *To minimize artifact, frequency encoding direction should be along the long axis of the hardware (so artifact projects on hardware). With pedicle screws, frequency encoding should be anterior-posterior. Increasing the receiver bandwidth, maximizing the echo train length, and decreasing the slice thickness and echo time will also diminish susceptibility artifact.*

Occipitocervical Fixation

KEY FACTS

PRE-PROCEDURE

- Indicated for occipitocervical instability, C1-C2 instability where C1-2 arthrodesis not possible or failed
- Trauma: Occipitoatlantal dislocations, atlantoaxial subluxation
- Rheumatoid arthritis: Cranial settling with brainstem or cord compression
- Tumors
- Infection: Instability and destruction of occipitocervical junction
- Congenital (basilar invagination)

PROCEDURE

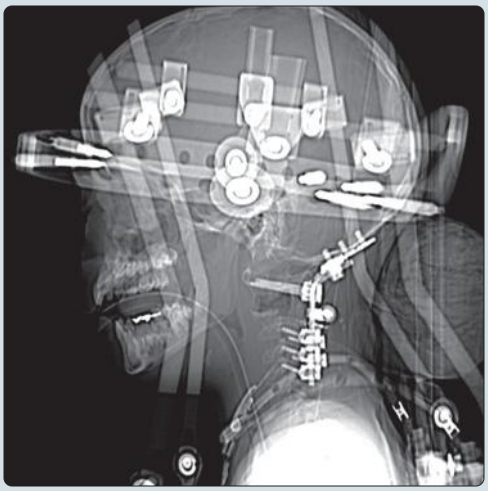
- Posterior wiring/rods, screws/rods, hooks/rods, screws/plates, wires/plates, posterior wiring with onlay graft

POST-PROCEDURE

- Overall fusion rate for occipitocervical fusion > 90%

OUTCOMES

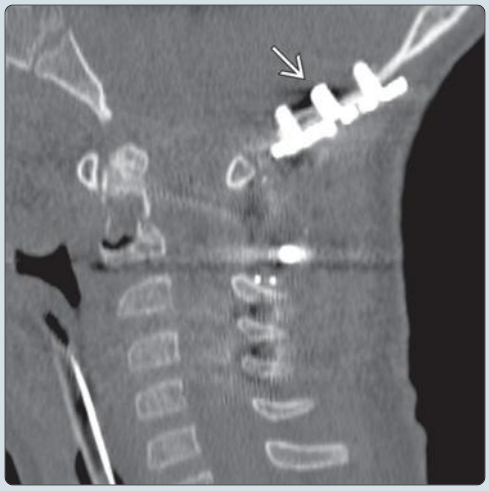
- Neurologic complications
  - Injury to cord, cranial nerve 12, C1 and C2 roots
  - Incidence of cord injury in OCF very low
- Vertebral artery (VA) injury primarily from screw placement (4%)
  - If intraoperative injury to dominant VA occurs, attempts should be made to control bleeding and repair injury
  - Normal immediate VA angiogram does not preclude later pseudoaneurysm formation
  - Bilateral VA injury can be fatal, so no screw placed contralateral to VA injury
  - Bony and VA variations seen in 18-23%
    - ↑ risk of VA injury
    - If high-riding VA at C2, C2 pars becomes very narrow and limits safe screw placement





(Left) *This is a resection of a C2 aneurysmal bone cyst. Lateral CT localizer shows halo instrumentation and occiput-C5 fixation. Three screws fix the occipital plate with bilateral C1 lateral mass screws and C3-C5 lateral mass screws.* **(Right)** *Postoperative sagittal NECT in the same patient shows that a rib allograft ⇨ was used as a scaffold for bony fusion. Occipital hardware is partially visible ⇨, and gas is present in resection site ⇨.*





(Left) *Axial NECT in the same patient shows that the left pars screw extends ventrally into the vertebral foramen ⇨. The left vertebral artery (VA) was coil embolized after verifying that the right VA was patent.* **(Right)** *Sagittal NECT following resection of a giant cell tumor of the C2 shows the occipital part of the hardware construct with 3 screws extending through the inner table ⇨.*

TERMINOLOGY

Abbreviations

- Occipitocervical fixation (OCF)

Synonyms

- Occipitoatlantoaxial fixation

PRE-PROCEDURE

Indications

- OC instability, C1-C2 instability where C1-2 arthrodesis not possible or failed
  - Trauma: Occipitoatlantal dislocations, atlantoaxial subluxation
  - Rheumatoid arthritis: Cranial settling with brainstem or cord compression
  - Tumors
  - Infection: Instability and destruction of occipitocervical junction
  - Congenital (basilar invagination)
- Symptoms
  - Cord and brainstem compression with respiratory distress, pain, cranial nerve dysfunction, paresis and paralysis, sudden death

Contraindications

- Comorbidities that preclude use of general anesthesia
- Lack of sufficient occipital bone (prior suboccipital craniectomy)

PROCEDURE

Procedure Steps

- Multiple methods and hardware types
  - Posterior wiring/rods, screws/rods, hooks/rods, screws/plates, wires/plates, posterior wiring with onlay graft
  - Modern screw-based constructs
    - Have generally supplanted contoured loops or rods connected with sub- or interlaminar wires/cables
  - Strong rigid fixation obtained with occipital plate and screws, longitudinal rods, C1 screws (pedicle or lateral mass), and C2 screws (pedicle, pars, C1-C2 transarticular)
  - C2 or transarticular screws placed first, followed by C1 lateral mass screws
  - Occipital screws placed last, with location dictated by specific implants utilized, quality of occipital bone, and placement of more caudal screws
  - Graft types variable, but iliac crest bone graft is gold standard
    - Rib autograft should be anchored with cranial and other wiring techniques

POST-PROCEDURE

Expected Outcome

- Overall fusion rate for occipitocervical fusion > 90%
- Posterior screw/rod constructs show ↓ postop adverse events and better neurologic improvement compared to posterior wiring/rod, screw/plate, and onlay techniques

OUTCOMES

Complications

- Immediate/periprocedural complication(s)
  - Neurologic
    - Injury to cord, cranial nerve 12, C1 and C2 roots
      - Incidence of cord injury in OCF very low
      - Hypoglossal nerve is anterior to C1 cortical ring; may be injured by C1 lateral mass screw that is too long or C1-C2 transarticular screw
      - C1 and C2 roots may be injured at screw starting point or screw placement
    - Injury of cerebellum or venous sinus from deep penetration of occipital screw
  - Alignment
    - Head position should be neutral
    - Flexed position can result in narrowing of pharynx
  - Vascular
    - Injury to vertebral artery (VA) or internal carotid artery (ICA)
    - VA injury rates from OCF (0-4%)
    - VA injury primarily from screw placement
      - If intraoperative injury to dominant VA occurs, attempts should be made to control bleeding and repair injury
      - Normal immediate VA angiogram does not preclude later pseudoaneurysm formation
      - Bilateral VA injury can be fatal, so no screw placed contralateral to VA injury
    - Bony and VA variations seen in 18-23%
      - ↑ risk of VA injury
      - If high-riding VA at C2, C2 pars becomes very narrow and limits safe screw placement
    - Posterior ponticulus (15%)
      - May be mistaken for broad C1 lamina, and placing screw might breach VA
    - Persistent 1st intersegmental artery (4.7%)
      - VA courses below C1 arch, making lateral mass screws dangerous
    - ICA courses anterior to C1 lateral mass
      - C1 lateral mass screws that breach anterior cortex of C1 at risk of causing ICA injury
- Delayed complication(s)
  - Hardware failure with nonunion (7%)

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KEY FACTS

TERMINOLOGY

- Instrumentation in fusion surgery designed to stabilize, not replace spinal bony elements

IMAGING

- Postoperative imaging: Assess progress of osseous fusion, confirm correct positioning and integrity of instrumentation, detect suspected complications (e.g., infection or hematoma), and detect new disease or disease progression
  - Assess progress of osseous fusion
  - Confirm correct positioning and integrity of instrumentation
  - Detect suspected complications (e.g., infection or hematoma)
  - Detect new disease or disease progression
- Radiographs
  - Noninvasive, look for hardware break or peri-implant lucency
- CT
  - Modality of choice for imaging bony detail to enable accurate assessment of degree of osseous fusion
- MR
  - Useful for detecting and monitoring infection or postoperative collections

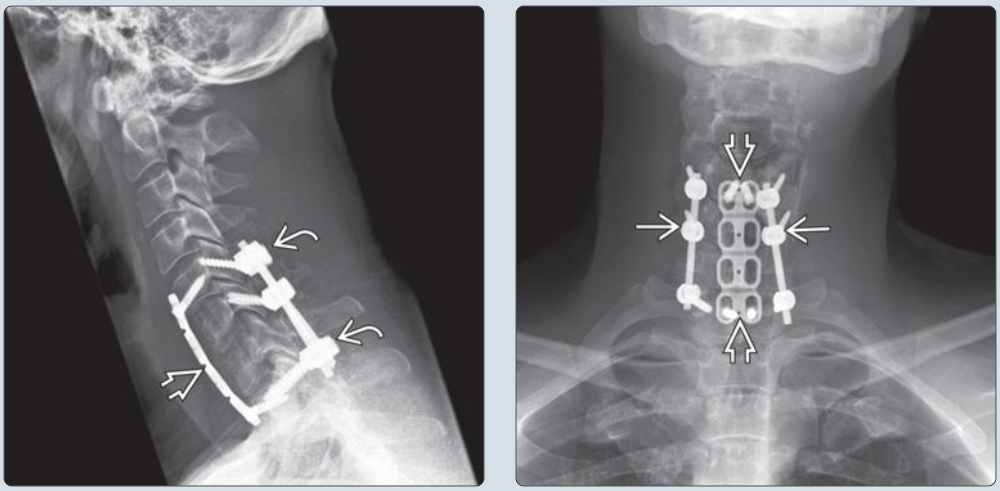
CLINICAL ISSUES

- Complications
  - Pseudoarthrosis, perihardware fractures
  - Nerve or vascular injury, dural tear
  - Infection
  - Hardware misplacement

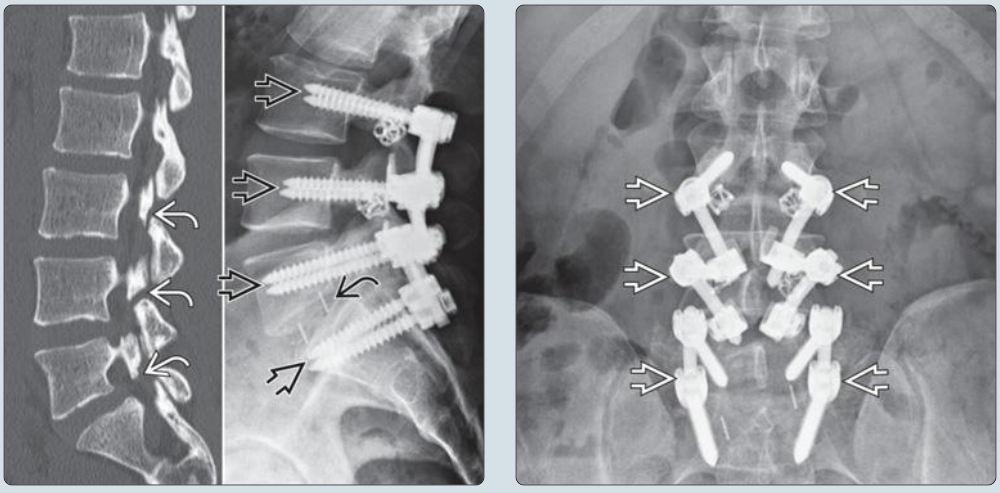
DIAGNOSTIC CHECKLIST

- Compare current radiographs with multiple previous studies

(Left) *Lateral radiograph shows multilevel anterior cervical discectomy and fusion (ACDF) spanning C5-T1. Lateral mass fixation is the gold standard in the presence of post-laminectomy instability and incompetency of posterior elements.* **(Right)** *Dynamic plates allow some motion at screw-plate interface. By allowing graft to subside, higher fusion rate is obtained. Lateral mass plating resists rotational and extension forces more effectively than posterior cervical wiring techniques.*



(Left) *Pars interarticularis defects at L3-L4, L4-L5, and L5-S1. Repetitive hyperextension contributes to spondylolysis; ↑ incidence in certain sports. Note the repair of multiple-level spondylolysis by pedicle screws. Interbody disc space is noted.* **(Right)** *Patients with spondylolysis show higher incidence of spondylolisthesis or DDD at level of pars defects and at upper adjacent level. Pedicle screw-rod-hook constructs are most stable.*



TERMINOLOGY

Synonyms

- Spine instrumentation, spine fusion surgery

Definitions

- Instrumentation in fusion surgery designed to stabilize, not replace spinal bony elements
  - Intact osseous fusion is crucial

IMAGING

General Features

- Location
  - Cervical
    - Anterior cervical discectomy and fusion (ACDF) is standard treatment for degenerative disease
    - Posterior lateral mass plating, laminoplasty
  - Thoracic
    - Pedicle screws connected by single or dual rods
  - Lumbar
    - Pedicle screws connected by plates or rods spanning single or multiple vertebral segments ± crossbars for additional strength

Radiographic Findings

- Noninvasive, most common for assessment of fusion
  - Hardware break or peri-implant lucency
  - Interspinous distance ≥ 2 mm on dynamic radiographs is more reliable indicator of pseudarthrosis than angular motion of 2° based on Cobb angle measurements

CT Findings

- CT is more accurate than radiographs
  - Modality of choice for imaging bony detail to enable accurate assessment of degree of osseous fusion

MR Findings

- Best demonstrates intraspinal contents, presence of infection
- Susceptibility artifact problematic, particularly with stainless steel devices
  - Implants made of titanium alloys less ferromagnetic → less severe susceptibility artifacts

PATHOLOGY

Staging, Grading, & Classification

- Cervical
  - ACDF
    - Inherent stability of bone graft in interspace
  - Posterior
    - Prevention or presence of postop instability
    - Laminoplasty developed for decompression in patients with ossification of the posterior longitudinal ligament
- Lumbar
  - For multilevel fusion, rods preferred over plates → rods can be individually cut and molded to facilitate maintenance of sagittal alignment
    - Tips of pedicle screws should be embedded in vertebral bone, should not breach anterior cortex
  - Sacral screws may be anchored in anterior cortex for additional stability
  - Translaminar facet screws: Alternative form of posterior instrumentation when posterior spinal elements are left intact
  - PLIF: Bilateral partial laminectomies, discectomy, interbody spacer(s), packing disc space with graft material
    - Posterior instrumentation provides rigid support until bone fusion occurs
  - ALIF: Remove disc material, replace disc height with cages, supplemented by screws and rods or plates
    - At L5-S1 vertebrae, anterior fusion supplemented by posterior instrumentation because iliac crests limit lateral access

CLINICAL ISSUES

Presentation

- Complications
  - Pseudoarthrosis
  - Metal ion release secondary to corrosion occurs in implants, even those with successful fusion
  - Cervical anterior surgical approach risks
    - Graft migration, injury to superior or recurrent laryngeal nerves, esophageal perforation, persistent dysphagia, dysphonia, odynophagia, dural tear
  - Infection
  - Hardware misplacement
  - Perihardware fractures
    - Pedicle, facet breach

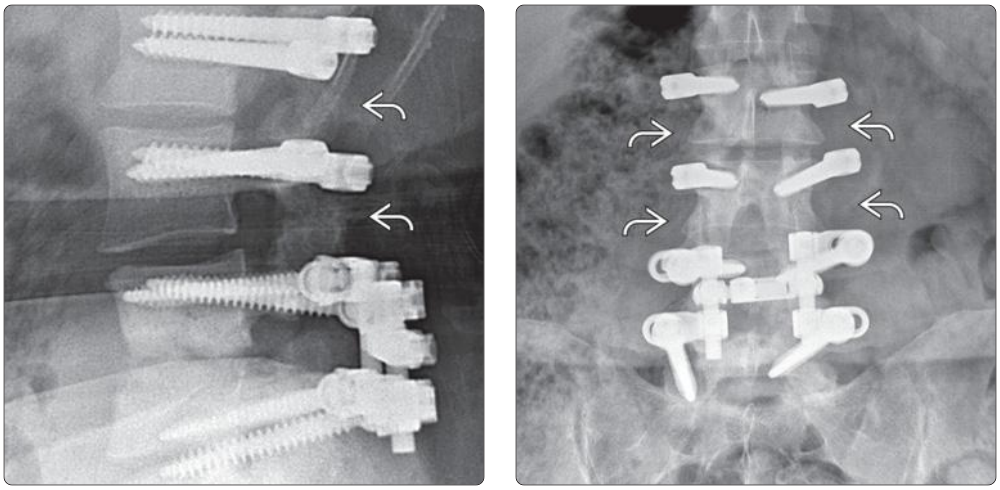
Natural History & Prognosis

- Rate of fusion after anterior cervical intervertebral disc removal and fusions reported to be 97%
  - Average follow-up was 5 ± 3.3 years
  - Complete pain relief in 78%, partial in 18%, and little or no pain relief in 4%
- Long-term radiographic follow-up in patients with cervical ACDF → hypermobility and degenerative changes in nonfused segments: Disc space narrowing, endplate sclerosis, and osteophyte formation
- Neurological outcome laminoplasty = laminectomy

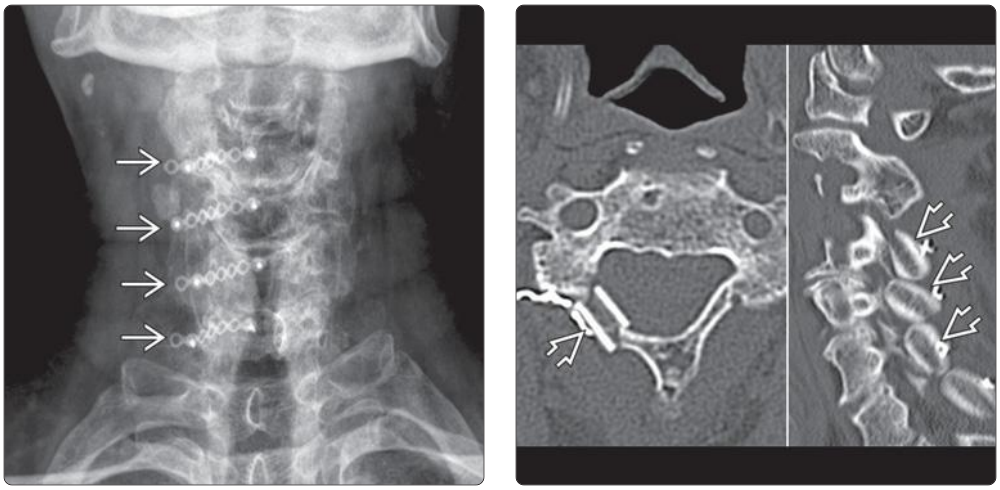
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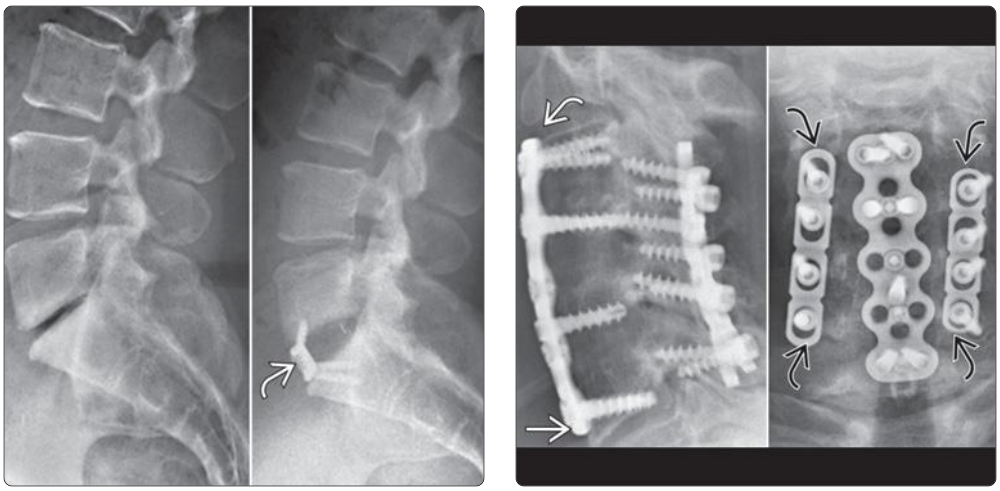
(Left) *Lateral radiograph shows dynamic stabilization device with radiolucent posterior rods. By altering load-bearing & controlling abnormal motion, stabilization helps limit stress placed on segment adjacent to level of fusion & thus helps prevent progressive degeneration.* **(Right)** *Dynamic stabilization devices may be grouped, according to their design: (a) pedicle screws and artificial ligaments, (b) interspinous process decompression devices, and (c) posterior element replacement systems.*



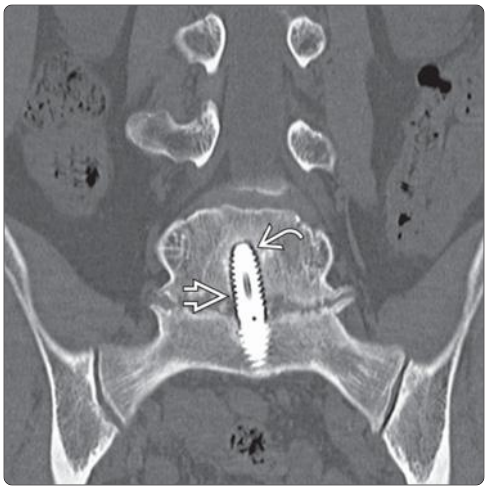
(Left) *Cervical laminoplasty decompresses canal in patients with multilevel anterior compression due to posterior longitudinal ligament ossification or spondylosis. Laminoplasty can worsen alignment in ~ 35% with development of kyphosis in ~ 10% of patients at long-term follow-up. Cervical ROM is decreased substantially after laminoplasty.* **(Right)** *Laminoplasty with intralaminar grafts may be alternative procedure for cervical myelopathy due to disc herniation.*



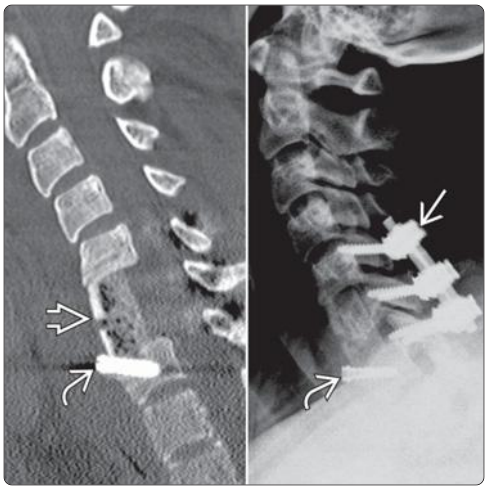
(Left) *Lateral view shows a synfix cage fixed with screws to adjacent vertebral bodies to obviate further posterior instrumentation.* **(Right)** *Anterior cervical plate subsides 1-1.5 mm per segment after procedure and may overlap disc space. Rigid pedicle screw instrumentation with fixation plates ↑ probability of solid fusion but may be associated with ↑ postop morbidity caused by disruption of posterior musculature. ↑ fusion rate is not associated with ↑ clinical success.*







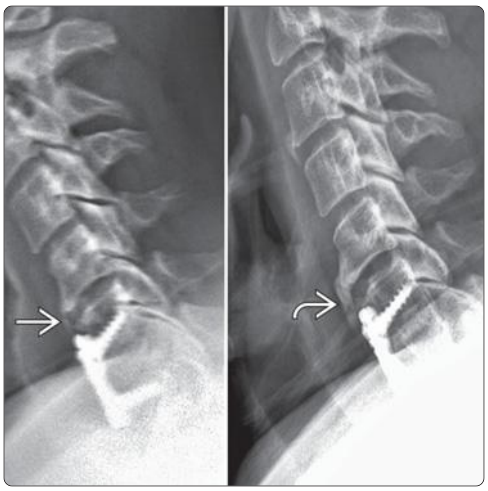
(Left) *Sagittal CT reconstruction shows posterior nonsegmental fusion with bilateral facet screws. Screw fixation of the facet joint has been reported to stabilize the lumbar spine and facilitate spinal fusion.* **(Right)** *Coronal CT reconstruction shows L5-S1 anterior lumbar interbody fusion with interbody screw implant soaked in bone marrow aspirate. Lucency and sclerosis along margins of screw are concerning for loosening and motion at this segment.*





(Left) *Sagittal CT shows C6 corpectomy with intervening allograft, secured in place by an interference screw.Lateral radiograph demonstrates posterior fusion.* **(Right)** *Translaminar facet (TLF) joint screw fixation is a low-profile construct that achieves stabilization when posterior bony elements are preserved. This can restrict the facet joint and intervertebral motions when compared with the intact spine. Paired interbody spacers are observed.*





(Left) *Sagittal T1WI MR shows posterior osteophyte formation at level of ACDF at C3-C4 indicating motion.* **(Right)** *Lateral radiographs of cervical spine in 2007 and in 2009 demonstrate increase in heterotopic bone formation due to suboptimal placement of anterior fusion plate close to the superior intervertebral disc space. Plates should be placed at least half a vertebral body height from adjacent disc space to prevent adjacent level ossification.*

Cages

KEY FACTS

TERMINOLOGY

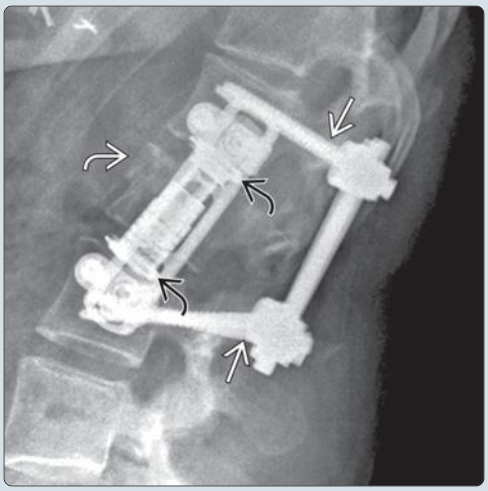
- Cages used to reconstruct anterior spinal column after corpectomy
  - In vivo expansion of cage permits optimal fit and correction of deformity

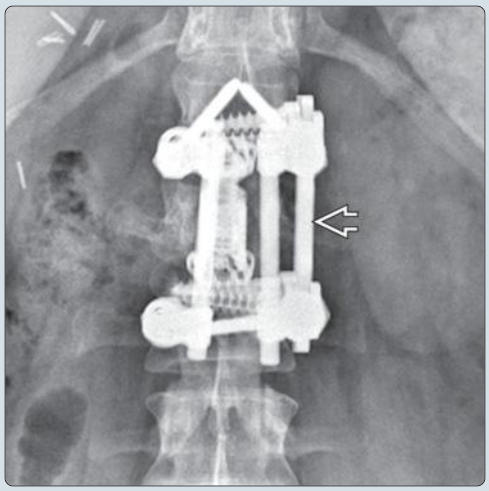
IMAGING

- Cylindrical metallic prosthesis along craniocaudal span of vertebral body (VB)
- Radiography and CT
  - Difficult to assess fusion
  - Assess stability of expandable cages (EC)
  - Determine EC-related complications
- MR
  - May be obscured by metal artifact
  - Best to evaluate thecal sac and neural elements

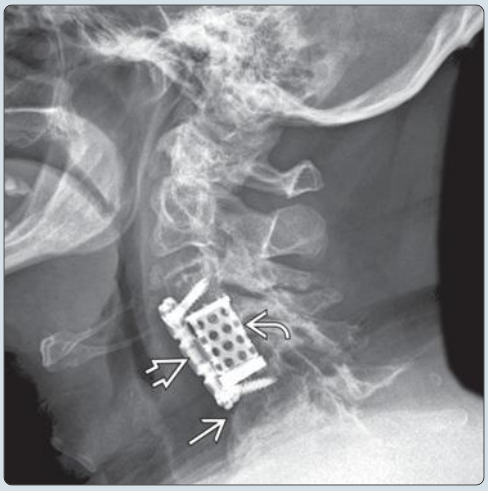
CLINICAL ISSUES

- Indications for corpectomy or vertebrectomy: Tumors, trauma, degenerative disease, infection, deformity
  - Cage provides immediate segmental stability, correction of sagittal plane deformity, and restoration of anterior vertebral support
- Not designed as stand-alone device
  - Anterior instrumentation ± posterior instrumentation
- Complications
  - Migration, subsidence, structural failure of cage
  - EC associated with higher rates and risk of subsidence in comparison with static cages
    - Cages with prongs may limit subsidence





(Left) *Lateral radiograph displays a L2 burst fracture. A Synex expandable cage (EC) extends from L1 to L3, and there are bilateral L1 and L3 pedicle screws. The goals of surgical intervention include decompressing neural elements, restoring vertebral body height, correcting angular deformity, and stabilizing columns of spine.* **(Right)** *AP radiograph shows the EC and lateral construct, which provides effective intervertebral distraction, ↑ anterior column height, and ↓ additional stress across pedicle screw constructs.*





(Left) *Lateral radiograph shows a nonexpandable cage spanning the C4 and C5 levels. ACDF is also noted from C3 to C5. Such titanium mesh cages have to be adjusted to the endplates and to the height of the defect. There is collapse of C5 vertebral body with subsidence of anterior fusion plate and cage.* **(Right)** *Coronal radiograph and coronal CT exhibit paired Pyramesh cages spanning the L2 level for vertebral collapse. Methylmethacrylate is noted surrounding the cages.*

TERMINOLOGY

Synonyms

- Expandable cage (EC), vertebral body (VB) replacement cage, VB prosthesis, distractible VB replacement

Definitions

- Cages used to reconstruct anterior spinal column after corpectomy

IMAGING

General Features

- Best diagnostic clue
  - Cylindrical metallic prosthesis along craniocaudal span of a VB
- Location
  - Cervical, thoracic, lumbar
- Size
  - Variable depending on spinal segment

Radiographic Findings

- Difficult to assess fusion
- May assess stability of EC and related complications
- Important to determine degree of correction of kyphotic angle between pre- and postoperative phase
- Polyetheretherketone (PEEK) and carbon fiber cages are radiolucent
  - Upper and lower titanium pins identify cage position

CT Findings

- Due to beam hardening artifact, difficult to assess fusion
- Assess stability of EC and related complications

MR Findings

- Best to evaluate thecal sac and neural elements
  - May be obscured by metal artifact
  - Little to no implant artifact with PEEK cages
- Helpful to evaluate for postoperative collections

DIFFERENTIAL DIAGNOSIS

Interbody Fusion Devices

- Fusion across disc space to increase stability of operated segment(s)
- Substrate for fusion includes autograft, allograft, bone graft substitutes, and osteogenic factors

Plates and Screws

- Postop imaging to assess progress of osseous fusion, confirm correct positioning and integrity of instrumentation, and detect suspected complications

PATHOLOGY

General Features

- Following neural decompression, interbody replacement and bone fusion are main goals of spine surgery
  - Increase height and cross-sectional area of foramina

Staging, Grading, & Classification

- Most interbody fusion cages are made of titanium
- PEEK: Hard radiolucent plastic ± carbon fiber reinforcement
  - Provides strength and stiffness in disc space
  - Induces cell attachment and fibroblast proliferation and increases protein content of osteoblasts

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Indications for corpectomy or vertebrectomy: Tumors, trauma, degenerative disease, infection, deformity

Natural History & Prognosis

- Immediate segmental stability, correction of sagittal plane deformity, and restoration of anterior vertebral support
- Complications
  - Migration, structural failure of cage
  - Subsidence of cage into vertebral body leading to instrumentation failure
    - EC associated with higher rates and risk of subsidence in comparison with static cages
      - Overexpansion of cage appears to be likely cause
    - Cages with prongs may limit subsidence
  - Extensive distraction for cage placement may end up causing radicular pain due to stretching of nociceptive fibers in joint capsule
  - Anterior approach: Vessel injury (5.8% reported) and internal organ injury
  - Retroperitoneal approach: Injury to ureter, lumbosacral plexus, or sympathetic chain
  - Adjacent-level vertebral body fractures can occur in coronal plane

Treatment

- Reconstruction of vertebral body with allograft, carbon fiber, mesh cages, tricortical autograft, expandable cages
  - Autograft: Gold standard, but morbidity
  - Allograft: Long fusion times, possible immunological rejection
  - Carbon fiber: Radiolucent, inflammatory reaction, brittleness → breakage
  - Mesh cages: Must be cut to a perfect size
    - Sharp edges anchor into adjacent VB → torsional stability
  - Expandable cages: Expand to appropriate size after placement, reduce kyphotic deformity, and restore height loss
- EC
  - Require anterior approach, may be extracavitary
  - Graft material can be used to fill EC
  - Not designed as stand-alone device → anterior plate or posterior rod construct

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Interbody Fusion Devices

KEY FACTS

TERMINOLOGY

- Fusion across disc space to increase stability of operated segment(s)

IMAGING

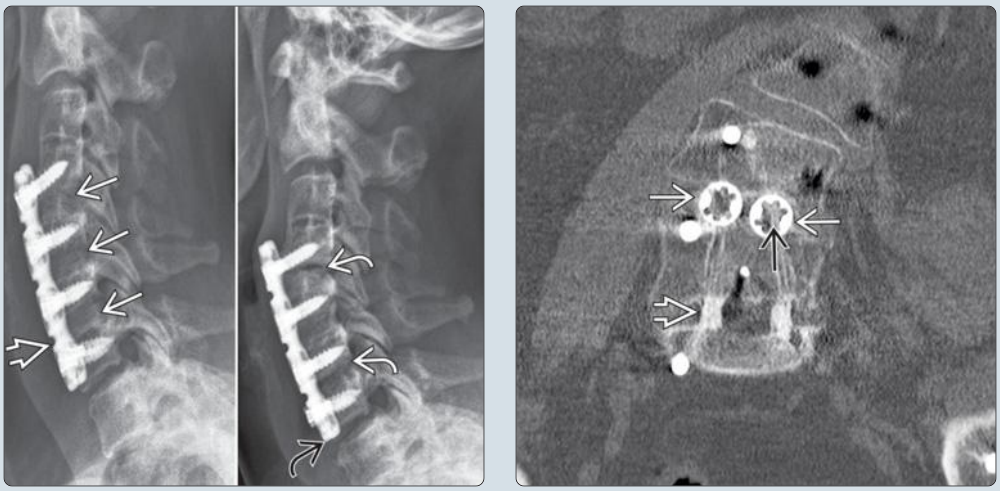
- Radiography and CT
  - Assess fusion level(s), the approach, type and number of fusion devices, change in position of interbody fusion device (IFD), peri-implant lucencies, vertebral alignment, disc disease at levels other than those fused
- Small radiodense markers in radiolucent IFD
- CT preferred method of assessing IFD
  - Thin section coronal and sagittal reformations show fusion maturation and bone growth

PATHOLOGY

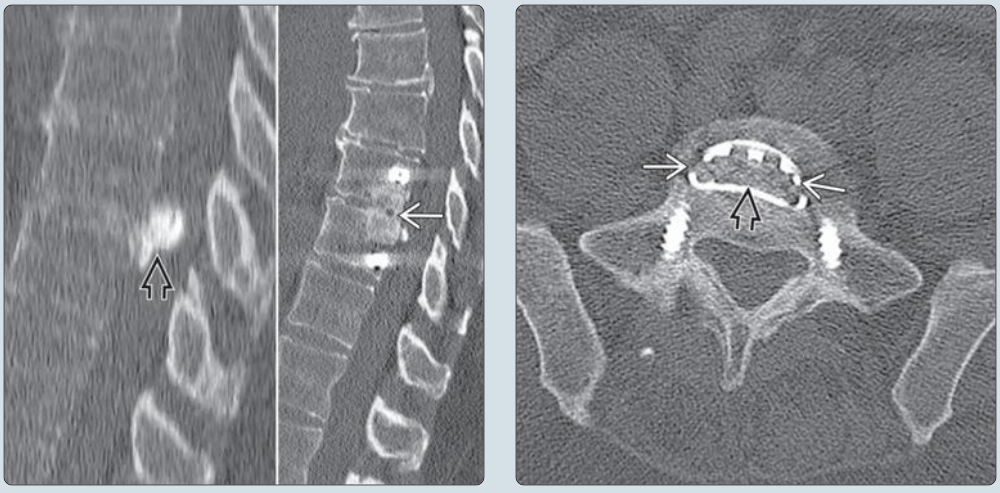
- IFD made of various materials
  - Substrate for fusion includes autograft, human or animal allograft, bone graft substitutes, and osteogenic factors
  - Choice of IFD affects radiographic/CT appearance
- Solid fusion seen radiographically in ~ 95% of patients, but clinical success in only 60–80%

CLINICAL ISSUES

- Primary clinical goal is relief of neural compromise through adequate decompression
- Solid bony fusion
- Complications
  - Subsidence
  - Adjacent segment degeneration (ASD) in 2.9-11.1% of patients with fusion, actuarial 10-year risk of 25.6%
  - Failure of fusion, pseudoarthrosis, in up to 20%



(Left) *Lateral radiograph initially after anterior cervical discectomy with fusion (ACDF) depicts allografts from C4 to C7. The anterior plate helps stabilize multilevel spondylosis. At 12-month follow-up, lucencies at the vertebral-graft interface are concerning for pseudoarthrosis. Subsidence is also noted.* **(Right)** *Coronal CT shows paired Ray titanium cages, which are filled with bone graft material, and have increased surface area for bone growth. An allograft interbody fusion device is in the adjacent disc space.*



(Left) *Sagittal CT reconstruction (left) shows a calcified disc extrusion in the midthoracic spine. The 2nd image (right) illustrates partial corpectomy with resection of the disc extrusion and interbody fusion with an osseous dowel.* **(Right)** *Axial CT reconstruction shows a crescentic metal interbody spacer. Hollow center is filled with bone graft material to promote fusion. Keeping postoperative L4–L5 segmental lordotic angle at ≥ 20° is important for prevention of clinical adjacent segment degeneration (ASD).*

TERMINOLOGY

Synonyms

- Interbody fusion device (IFD), implant, cage, spacer

Definitions

- Fusion across disc space
  - Restore and maintain disc space height and normal sagittal contours
  - Increase stability of operated segment(s)
- Cervical
  - Anterior cervical discectomy with fusion (ACDF) ± plating is standard treatment of cervical degenerative disc disease (DDD)
- Lumbar
  - Lumbar fusion for back pain ± leg pain associated with DDD is the gold standard

IMAGING

General Features

- Best diagnostic clue
  - Radiodense material within intervertebral disc space
- Location
  - Cervical, lumbar, and less commonly thoracic

Radiographic Findings

- Radiolucent polyetheretherketone (PEEK) and JAGUAR I/F cages, BOOMERANG II devices identified by radiodense markers
- Difficult to assess bony bridging
  - Radiolucent graft and increased radiodensity with de novo bone formation
- Criteria for arthrodesis
  - Bridging bone across graft-host interface
    - Identification of bone trabeculation on **static** radiographs considered less reliable indicator of cervical arthrodesis than on **dynamic** films
  - Measurement of interspinous distance on dynamic radiographs of ≥ 2 mm is more reliable indicator of pseudoarthrosis than angular motion of 2° based on Cobb angle measurements

CT Findings

- Assess fusion level(s), the approach, type and number of fusion devices, change in position of IFD, subsidence, peri-implant lucencies, vertebral alignment, disc disease at levels other than those fused
- Thin section CT with coronal and sagittal reformations show fusion maturation and bone growth
- Can identify failure of device fixation and nonunion, but not as sensitive in identifying true arthrodesis
- Metal devices (e.g., BAK cage) can produce severe beam hardening and streak artifacts
- Composite cages are radiolucent
- No imaging artifacts with bony devices

MR Findings

- Metal IFDs produce significant susceptibility artifact, degrading anatomic detail
- Detect infection or postoperative collections
- Marrow edema or inflammation are nonspecific in determining fibrous union vs. early arthrodesis

Imaging Recommendations

- Best imaging tool
  - CT is preferred method of assessing IFD
  - Radiographs: True lateral and AP projections important for adequate evaluation of intervertebral space
    - Weight bearing, flexion/extension, lateral bending
- Protocol advice
  - Some authors suggest CT at 3, 6, 12 months
  - Compare to multiple prior studies to assess gradual changes

Nuclear Medicine Findings

- SPECT imaging shows increased focal uptake at level of pseudoarthrosis

DIFFERENTIAL DIAGNOSIS

Cages

- Provides immediate segmental stability, correction of sagittal plane deformity, and restoration of anterior vertebral support after corpectomy

Lumbar Artificial Disc

- Prosthetic disc replacement to treat DDD, restoring normal mobility of diseased segments and improving clinical outcomes by decreasing risk of adjacent segment degeneration (ASD)

Cervical Artificial Disc

- Prosthetic total disc replacement after anterior decompression to preserve normal range and type of intervertebral motion

PATHOLOGY

General Features

- Substrate for fusion includes autograft (e.g., iliac crest), human or animal allograft, bone graft substitutes, and osteogenic factors
- **Ligamentotaxis** (stretching of motion segment via annulus and supporting ligaments) provides biomechanical stability
  - Indirect decompression of neural foramina

Staging, Grading, & Classification

- IFD made of various materials
  - Titanium
  - PEEK
  - Human allograft bone dowels
  - Bioabsorbable polymers
    - Immediate stability, relative radiolucency, gradual transfer of stress to fusion site, and breakdown products have no mutagenic or immunogenic properties
- Lumbar IFDs have varying geometric configurations and wall thicknesses
  - Metal: BAK cage, Ray threaded fusion cage, lumbar tapered fusion device, interfix/rp threaded fusion device, Harms cage
  - Composite: PEEK cage, JAGUAR I/F cage, BOOMERANG II device
  - Biologic: Cortical bone dowels

- Bone graft substitutes: INFUSE bone graft composed of recombinant human bone morphogenetic protein (rhBMP)
  - Stimulates mesenchymal stem cell differentiation into osteoblasts and initiates bone formation
- Cervical IFD for arthrodesis after 1- or 2-level ACDF
  - Autograft bone harvested from iliac crest, allograft bone from cadaveric iliac crest or fibula
    - Concerns with donor site morbidity
  - Titanium cages and rectangular fusion devices, ± autologous graft or substitute
  - PEEK cages, carbon fiber cage, polymethyl-methylmethacrylate (PMMA), rhBMP
    - PMMA does not lead to fusion
- Choice of IFD affects radiographic/CT appearance
  - Solid fusion seen radiographically in ~ 95%, but clinical success in only 60–80%

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Primary clinical goal is relief of neural compromise through adequate decompression
  - Solid bony fusion
- Complications
  - Failure of fusion, **pseudoarthrosis**, in up to 20%
    - Increasing or persistent pain, neurologic deficit, spinal deformity
    - Up to 30% of patients may not require surgery
  - Subsidence
    - Fusion device sinks into adjacent vertebral bodies
    - Increased incidence of failed fusion 2° loss of mechanical structural stability
  - ASD in 2.9-11.1% of patients with fusion, actuarial 10-year risk of 25.6%
    - ASD progresses in cases of fusion for trauma and nontraumatic cases → not solely the natural history of DDD
      - Results from alterations in normal biomechanics of spine: ↑ intradiscal pressure, ↑ facet loading, ↑ mobility at adjacent levels
      - Risk factors: Preexisting facet degeneration, age > 50 years
      - Incidence of **distal** ASD < that of **proximal** ASD
      - Higher segmental displacement values at cephalad motion segment in flexion/extension, lateral bending, and axial rotation
      - Loss of lumbar lordosis suggested cause of ASD radiographically ± symptomatic
  - Infection of allograft
  - Particulate debris from articulating surfaces induce immune inflammatory response → osteolysis and prosthetic loosening

Natural History & Prognosis

- 5 stages of arthrodesis
  - Inflammation → formation of granulation tissue
  - Vascularization → nutrients and osteoprogenitor cells at grafted site
    - Autologous graft → no immune reaction
    - Allograft → acute immune response is mounted against foreign body or cells
  - Osteoinduction
    - Process of new bone formation → arrival of progenitor cells
  - Osteoconduction → incorporation of graft
  - Remodeling → graft is transformed into stable, weight-bearing bone
- Cervical fusion: Excellent clinical and radiologic outcomes in 85-95% of patients
  - To minimize failure rates, use of anterior plate fixation to provide immediate stability, intervertebral cages, or spacers introduced
  - Higher rate of graft collapse and pseudoarthrosis in multilevel procedures with allograft
  - Expected autograft fusion rate for noninstrumented single-level fusions better than 80% and for 2-level fusion better than 70%
  - With allograft, expected fusion rate for noninstrumented single-level fusion is better than 80% and is better than 50% for 2-level fusion
- Lumbar fusion: 6-12 months for solid arthrodesis from time of implantation of IFD

Treatment

- Annular window, total discectomy to achieve clean bleeding endplates
- Lumbar: Posterior lumbar interbody fusion (PLIF), anterior lumbar interbody fusion (ALIF), transforaminal lumbar interbody fusion (TLIF)
  - ± segmental posterior instrumentation

DIAGNOSTIC CHECKLIST

Consider

- Dynamic views (e.g., flexion and extension) helpful for evaluating motion at operated segment

Image Interpretation Pearls

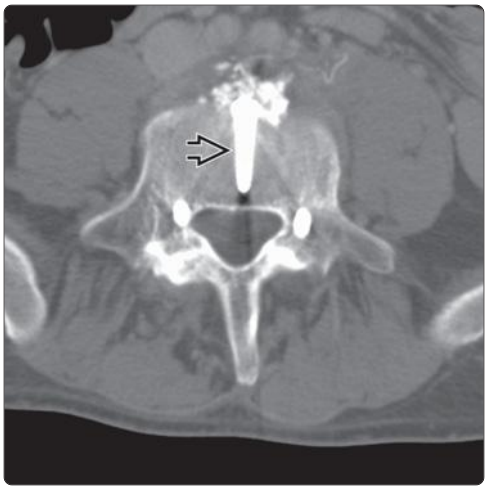
- Date of surgery important to evaluate appropriate appearance of osseous bridging
- Check operative report for type of IFD

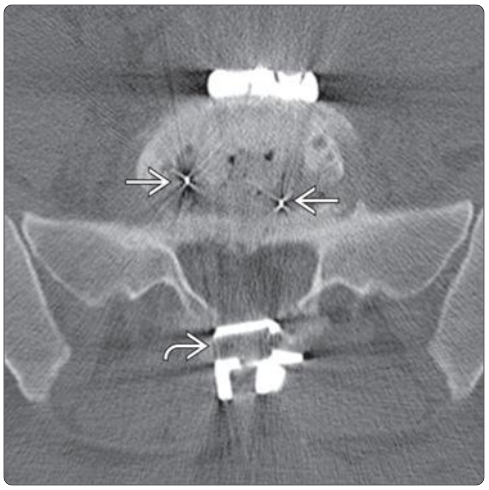
Reporting Tips

- Evaluate other degenerative changes, especially ASD

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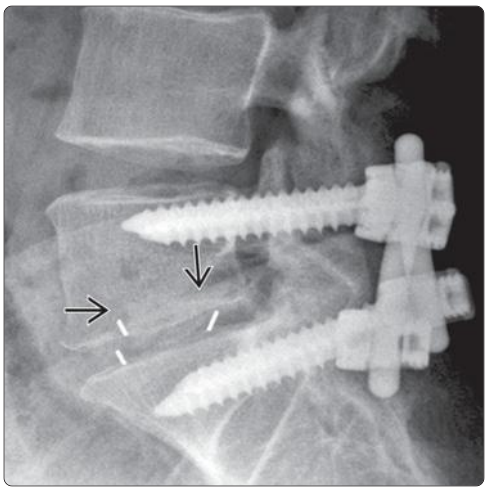
(Left) *Axial NECT shows a small titanium fragment screw ⇨ in the L5 vertebral body, which serves as an interference screw for the interbody graft.* **(Right)** *Axial bone CT illustrates anterior interbody fusion with plate and screw fixation, as well as posterior fusion with interspinous process spacing device ⇨. Small foci of high attenuation reflect markers for the interbody grafts ⇨. Outlines of radiolucent cages become increasingly apparent as adjacent bone graft consolidates.*





(Left) *Coronal CT demonstrates recent placement of radiolucent IFD and graft material ⇨. At 3 months, perihardware lucencies indicate loss of fixation, & subsidence reflects loss of structural stability. At 6 months, assess for bridging bone lateral to & within IFD. Cystic lucencies adjacent to IFD or linear defect through bridging bone indicates delayed union.* **(Right)** *Coronal CT reconstruction at a 12-month follow-up shows osseous bridging within ⇨ and along the lateral margin ⇨ of the implant.*





(Left) *Axial CT displays radiodense markers ⇨ of radiolucent IFD. At 12 months, there should be mature trabecularization. If arthrodesis is not present at 12 months, CT can be performed at 24 months. Lucencies at IFD margins indicates a nonunion.* **(Right)** *Small metallic markers ⇨ identify the radiolucent PEEK interbody spacer. The marker should be located at least 2 mm anterior to the posterior vertebral body margin, which provides reassurance that the ramp is not protruding into the spinal canal.*

Interspinous Spacing Devices

KEY FACTS

TERMINOLOGY

- Interspinous spacing device (ISD) places stenotic segment in slight flexion while preventing extension
  - Dynamic stabilization alters movement and load transmission of spinal motion segment without fusion of segment

IMAGING

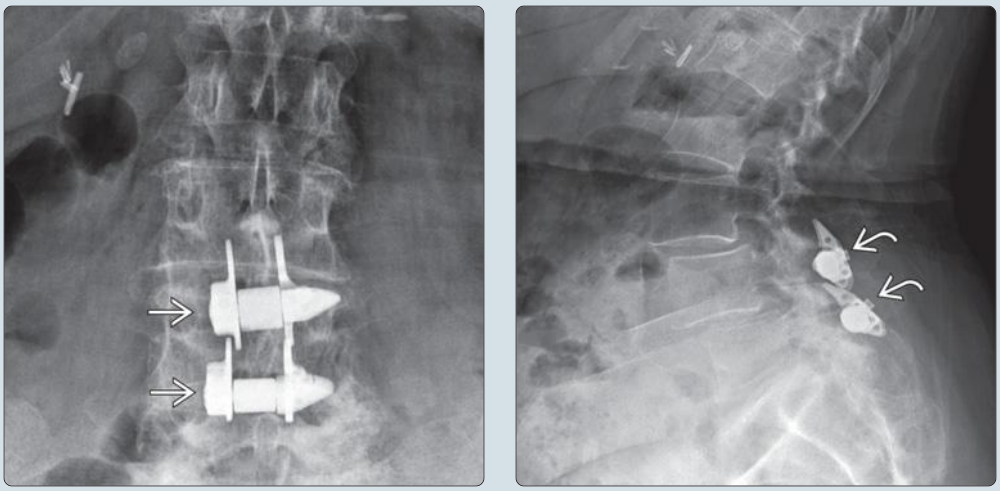
- Radiographs and CT helpful to evaluate interspinous location
  - Assess for fractures

PATHOLOGY

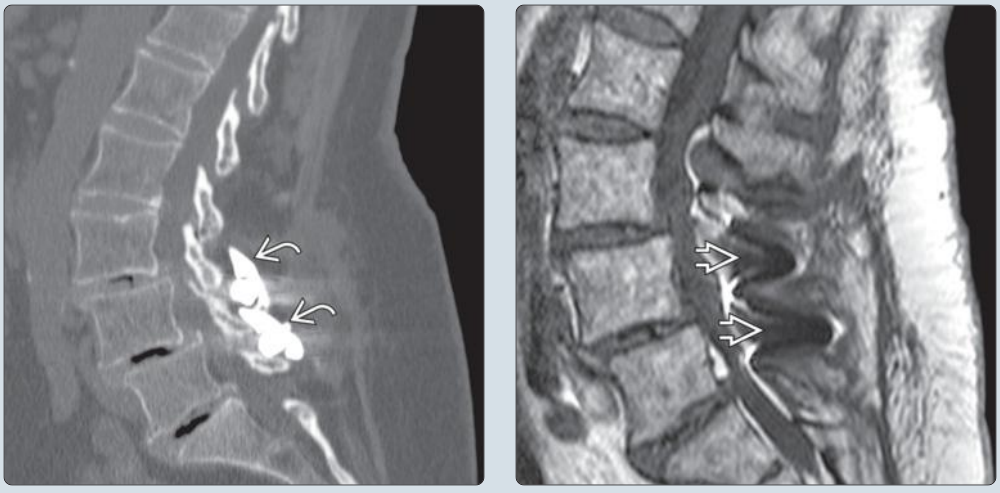
- Lumbar neurogenic claudication symptoms are exacerbated during extension, relieved during flexion
  - Dimensions of canal and foramen ↑ in flexion, ↓ in extension
  - Facet loading ↑ during extension, ↓ during flexion

CLINICAL ISSUES

- ISD potentially helpful for treatment of spinal stenosis with neurogenic claudication
  - Causes focal flexion at applied level resulting in increased canal and foramen dimensions
- Complications
  - ISD malposition, migration, dislocation, spinous process fracture
- Indications
  - Lumbar stenosis ± facet joint hypertrophy and subarticular recess stenosis, foraminal stenosis
  - 1 to 2 level lumbar stenosis from L1-L5 in patients with at least moderate impairment in function
- Patients' anatomy may influence outcomes
  - Decreased accessible distance between laminospinous plane and tip of spinous process
- Osteoporosis is contraindication
- **Precise role of ISD vs. standard surgical treatment of spinal stenosis (laminectomy) is controversial**



(Left) *X-Stop implants ⇨ at L4-L5 and L5-S1 are titanium alloy devices placed between spinous process (SP) to reduce spinal canal (SC) and NF narrowing that occurs in extension; therefore, reducing the symptoms of neurogenic intermittent claudication.* **(Right)** *DDD with loss of disc height can cause the superior facet to move anteriorly and superiorly into the foramen. Foraminal area may be further compromised by ligamentum flavum hypertrophy and osteophytes. Flexion created by ISD ⇨ can increase foraminal area by about 12%.*



(Left) *ISD ⇨ increases foraminal area, width, and height, as well as SC diameter in extension. ISD can improve recurrent facet joint pain in clinical short- and midterm settings, but does not exceed the outcome of denervated patients.* **(Right)** *Sagittal T1 MR shows the susceptibility artifact from the ISDs ⇨. X-Stop is the most commonly used ISD in patients with neurogenic intermittent claudication due to lumbar stenosis. Studies report improved visual analog and Oswestry disability index scores.*

TERMINOLOGY

Synonyms

- Interspinous posterior decompression (IPD), interspinous distraction device (IDD), interspinous spacer device (ISD), interspinous implant

Definitions

- Interspinous implant places stenotic segment in slight flexion while preventing extension
  - Dynamic stabilization alters movement and load transmission of spinal motion segment without fusion of segment

IMAGING

General Features

- Location
  - Interspinous soft tissues of lumbar spine
    - Extradural
- Morphology
  - Varies depending on type of ISD

Radiographic Findings

- "Free-floating" metallic prosthesis in interspinous region

CT Findings

- Implant is not rigidly attached to bony anatomy
  - Restricted from migrating posteriorly by supraspinous ligament, anteriorly by laminae, cranially and caudally by spinous process (SP), and laterally by 2 wings or stops on implant

MR Findings

- Susceptibility artifact may obscure detailed delineation of hardware

Imaging Recommendations

- Best imaging tool
  - Radiograph helpful to evaluate interspinous location
    - Flexion and extension views useful to detect abnormal motion of hardware
  - CT to assess vertebral bodies and posterior elements for fractures

DIFFERENTIAL DIAGNOSIS

Plates and Screws

- Plates are along vertebral margin, anterior or lateral
- Screws can be corporal or transpedicular

PATHOLOGY

General Features

- Lumbar neurogenic intermittent claudication symptoms are exacerbated during extension and relieved during flexion
  - Multiple theories including ischemic, mechanical compression, and stagnant anoxia
  - Radicular pain, often bilateral, exacerbated by standing, walking, and other positions that place lumbar spine (LS) in extension
    - Dimensions of spinal canal (SC) and neural foramen (NF) increase in flexion and decrease in extension
    - During flexion, there is decreased epidural pressure and nerve root compression
- Facet loading increases during extension and decreases during flexion
  - Collapsed disc → increased motion segment mobility → increased facet loading → low back pain
  - Degenerative disc disease (DDD) → loss of disc height → facet hypertrophy and buckling of ligamentum flavum, particularly during extension, contribute to narrowing of thecal sac
- ISD is inserted between SP of affected level and places motion segment in slight flexion, while simultaneously preventing extension
  - ± reduced range of motion during flexion/extension at instrumented level
    - Only restricts motion in 1 direction, allowing axial rotation and lateral bending of instrumented segment
    - Places stenotic segment in about 2° of flexion when unloaded and may reduce flexion and extension of that segment
  - Studies show that kinematics of adjacent levels during flexion/extension, axial rotation, and lateral bending are not significantly affected

Staging, Grading, & Classification

- Numerous devices available
  - X-Stop: Titanium oval spacer with 2 lateral wings to prevent lateral migration
  - Minns device: Dumbbell-shaped, made from silicone
  - Interspinous U (coflex)
  - Aperius PercLID system
  - Diam: Silicone interspinous spacer covered by polyethylene coat, secured in place with 2 ligatures around superior and inferior SPs
  - Wallis implant: Polyetheretherketone (PEEK) spacer placed between SPs and secured with 2 Dacron ligaments wrapped around SPs
    - Good 13-year clinical outcome in 1 study, obviated arthrodesis in 80% of patients
    - Current indications
      - Following discectomy for large herniated disc in which there is significant loss of disc material
      - Redo discectomy for recurrent herniation
      - Discectomy for herniation of transitional disc with sacralization of L5
      - Degenerative disc adjacent to fused segment
      - Isolated Modic 1 lesion attributable to chronic low back pain

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - **Precise role of ISD vs. standard surgical treatment of spinal stenosis (laminectomy) is controversial**
    - Standard surgery may have extensive soft tissue dissection for laminectomy, longer surgical time, and more blood loss
  - ISD helpful for treatment of spinal stenosis with neurogenic claudication
    - Causes focal flexion at applied level resulting in increased SC diameter even in attempted extension

- In some studies, compared with conservative treatment, ISD results in significantly better pain relief with effect lasting up to 2 years
  - ISD relieves disc pressure at implanted level and does not alter disc pressure at adjacent levels
  - Cadaveric studies show decreased facet loading in extension at treated level with no effect on facet loading at adjacent levels
  - Newer ISD can be placed percutaneously
- Recent systematic review showed higher rate of complications with ISC, such as spinous process fracture, implant dislocation, and heterotopic ossification
  - Also higher rate of reoperation
- Osteoporotic fractures may result only in loss of height or stooped posture
  - With spinal stenosis and narrowed lateral recess and foramina, extension of back from brace and axial loading from weight of standing causes nerve root compression
  - Extension of lumbar spine shown to decrease cross-sectional area of NF by about 15%

- Complications
  - Postoperative complication rate 10.1% and reoperation rate 6-7.2%
    - ISD malposition
    - Migration: Device remains between SP, function is preserved, and patient is asymptomatic
    - Dislocation: Device moved outside interspinous area, but contained by detached supraspinous ligament or associated with ruptured supraspinous ligament
      - Nonfunctional with recurring symptoms requiring revision surgery
    - SP fissures and fractures
    - Rare stress fracture of facet joints

Natural History & Prognosis

- Clinical success reported in 63.4% of X-Stop device-treated patients (compared to 12.9% of controls)
- 9.2% reoperation rate because of persistent or recurrent symptoms, with no clearly reported device-related complications

Treatment

- Indications
  - Lumbar stenosis ± facet joint hypertrophy and subarticular recess stenosis, foraminal stenosis, ± stable grade I spondylolisthesis or equivalent retrolisthesis
  - Limited to use in 1 or 2 level lumbar stenosis from L1-L5 in patients with at least moderate impairment in function
  - Dynamic stabilization, compared to fusion, attractive option for younger patients who would bear greater burden on adjacent segments during their prolonged follow-up
- Generally, osteoporosis is contraindication
  - Increased likelihood of further vertebral body compression
- Exclusion criteria in some studies
  - Unable to sit for prolonged periods without pain
  - Unremitting spinal pain in any position
  - Cauda equina syndrome
  - Pathologic fractures of vertebrae
  - Active infection
  - Paget disease at involved segments or spinal metastases
  - Spinal anatomy, such as ankylosing spondylitis or fusion at affected level
- ISD shown to have extremely high failure rate, defined as surgical reintervention, after short-term follow-up in patients with spinal stenosis caused by degenerative spondylolisthesis
- Patients' anatomy may influence outcomes
  - Interspinous degenerative changes, V-shaped interspinous area, SP morphology, facet hypertrophy (decreases accessible distance between laminospinous plane and tip of SP)
- Implants are not favorable, nor currently recommended, for use at L5-S1
- ISD does not restrict or eliminate any potential future therapeutic options that are currently being developed, such as arthroplasty

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

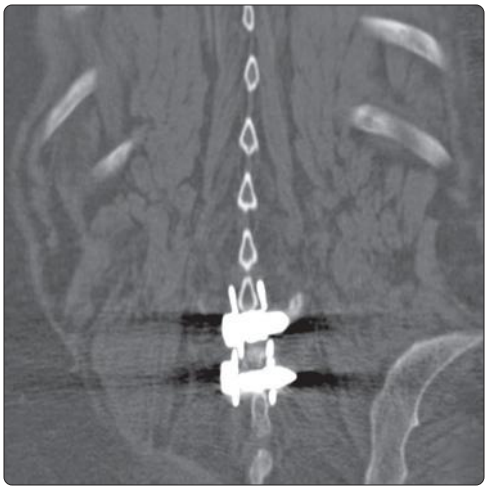
- Developmentally small spinal canal

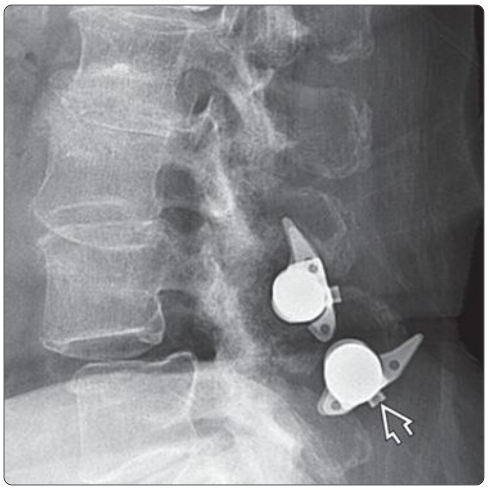
Reporting Tips

- Look for fractures: SP, facets, pars interarticularis
  - Vertebral compression fractures, especially in osteoporotic patients

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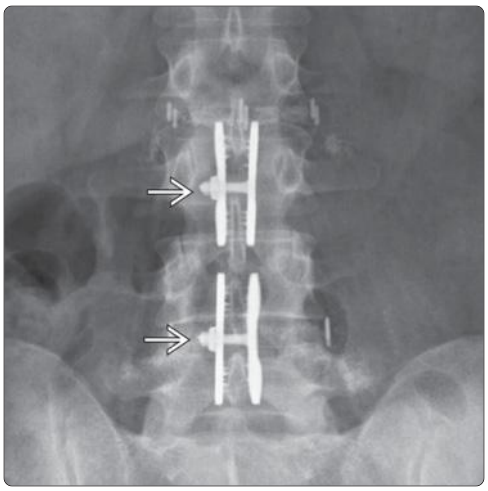


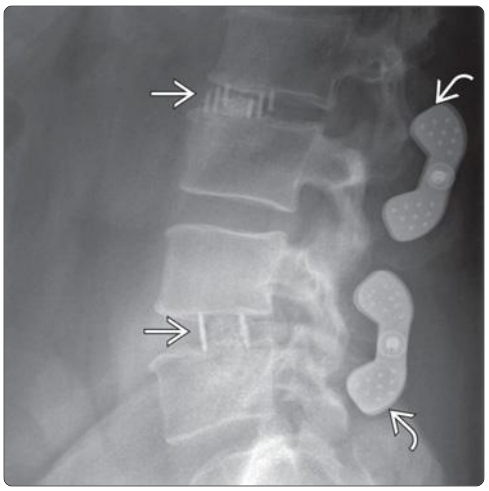
(Left) *Important anatomic factors considered in placement of ISD include, accessible distance between the laminospinous plane and tip of the SP, particularly in elderly patients with hypertrophic facet joints or spondylolisthesis; SP morphology; and kissing spine–like conditions, which may be a contraindication in elderly, less elastic spines.* **(Right)** *Dislocated X-Stop device is seen at the L5-S1 level. V-shaped posterior interspinous area may be a risk factor for ISD dislocation.*





(Left) *SP fracture is a potential complication of ISD. Because ISD results in flexion at the inserted level, there may be increased likelihood of further vertebral body compression in an osteoporotic patient.* **(Right)** *Failed ISD with recurrent symptoms required anterior interbody fusion with plate and screw fixation. Posterior fusion was performed with an interspinous process spacing device, which contains a hollow center allowing for placement of bone graft material.*





(Left) *NuVasive Affix SP plates at L2-L3 and L4-L5 are interspinous clamps used for further stabilization of the fusion construct. Correlation with operative report is essential in correctly reporting spinal hardware.* **(Right)** *Lateral radiograph shows 2-level posterior fusion constructs composed of SP clamps, resembling ISD. Interbody fusion grafts are seen at the L2-L3 and L4-L5 levels.*

Cervical Artificial Disc

KEY FACTS

TERMINOLOGY

- Artificial disc replacement, total cervical disc replacement (TCDR), artificial intervertebral disc arthroplasty (AIDA)
- Insertion of prosthetic total disc replacement after anterior decompression with aim of preserving normal range and type of intervertebral motion
- Preventing complications associated with rigid arthrodesis and subsequent segmental loss of motion

IMAGING

- Radiographs with flexion and extension views
- CT to assess for adjacent level disease and heterotopic ossification
- Postoperative MR may be helpful in cases of persistent pain to assess for inadequate decompression or myelopathy

CLINICAL ISSUES

- Ideal patient has single-level soft disc herniation causing neuro signs/symptoms, motion at involved segment, no evidence of osteoporosis or infection
- Worldwide indication is 1-2-levels radiculopathy or myelopathy; however, FDA indication is limited to single-level disease in levels C3 to C7
- Candidates: Those who failed conservative therapy with evidence of symptomatic nerve root ± spinal cord compression
- Complications
  - Device wear, subsidence or displacement, segmental hypermobility, and excessive disc space distraction to accommodate prosthesis and consequent facet joint separation and screw breakage
  - Facet arthrosis, adjacent level degeneration
  - Paravertebral heterotopic ossification may be prevented with NSAIDs





(Left) *Sagittal T2WI MR shows a C5-C6 focal disc protrusion. Literature suggests TCDR is a treatment option for cervical myelopathy in young patients with a focal central disc herniation, but without significant degenerative disc or facet disease.* **(Right)** *Lateral radiograph illustrates a ProDisc-C cervical disc. It has cobalt-chromium endplates with a central keel for anchorage to the VB and a locking core of ultra-high molecular weight polyethylene that provides a ball-and-socket articulation.*





(Left) *TCDR is placed with the same surgical approach as for anterior cervical discectomy and fusion. Porous titanium spray and hydroxyapatite support long-term fixation. TCDR can be used in patients presenting with neurologic deficit, radiculopathy, or myelopathy.* **(Right)** *Lateral radiograph shows a grade 2 heterotopic ossification, a complication of TCDR. Ossification along the lateral aspect of device may result in fusion, causing reduced movement, whereas anterior ossification results in continued mobility.*

TERMINOLOGY

Synonyms

- Artificial disc replacement, total cervical disc replacement (TCDR), artificial intervertebral disc arthroplasty (AIDA), cervical artificial disc replacement (CADR)

Definitions

- Insertion of prosthetic total disc replacement after anterior decompression with aim of preserving normal range and type of intervertebral motion
  - Prevent complications associated with rigid arthrodesis and subsequent segmental loss of motion
  - Transmit axial load from vertebral body above 1 below
  - Shock absorption
  - Decreased perioperative morbidity
    - Instrumentation
    - Pseudoarthrosis
    - Exposure
    - Bone graft morbidity
    - Postoperative immobilization
  - Early return to activities

IMAGING

General Features

- Best diagnostic clue
  - Metal alloy and polymer prosthesis in intervertebral disc space
- Location
  - Intervertebral disc space from C3-C7

Radiographic Findings

- Features useful for differentiating among devices
  - Presence and appearance of endplate keel, fixation spikes, and inlay may help identify individual devices
- Assess for heterotopic anterior paravertebral ossification

CT Findings

- Evaluate adjacent vertebral bodies, particularly to detect fractures and degenerative changes at adjacent disc space and facet levels
- Detect heterotopic bone formation and small metallic fragments from device wear

MR Findings

- Devices containing nontitanium metals (cobalt/chrome/molybdenum alloys in PCM and ProDisc-C) cause susceptibility artifact, preventing accurate assessment at surgical and adjacent levels
- Titanium devices, ± polyethylene (Bryan disc or Prestige LP), allow for satisfactory monitoring of adjacent and operated levels
- Evaluate adjacent vertebral bodies, particularly in detecting fractures and degenerative changes at adjacent disc space and facet levels
- Postoperative assessment of patients with persistent pain to determine adequacy of decompression and patients with myelopathy

Imaging Recommendations

- Best imaging tool
  - Radiographs with flexion and extension views
  - CT helpful to evaluate for paravertebral ossification, fractures, and adjacent degenerative changes

DIFFERENTIAL DIAGNOSIS

Interbody Fusion Devices

- Fusion across disc space to increase stability of operated segment(s)

Plates and Screws

- Instrumentation in fusion surgery to stabilize not replace spinal bony elements

PATHOLOGY

General Features

- Studies show relationship between solid fusion and development of adjacent-level disc disease
  - Fusion of 2 vertebral segments eliminates spinal motion segment with increased stress/strain at levels adjacent to fused segment
    - Thought to result from increased stress on, or hypermobility of, adjacent segments
- Normal motion between 2 vertebral bodies occurs around instantaneous center of rotation, generally situated in posterior 1/2 of upper portion of inferior vertebral body
  - Location of prothesis mimics natural situation

Staging, Grading, & Classification

- Materials: Metal (titanium, steel, chromium), ceramic, polymer (ultra-high molecular weight polyethylene or polyurethane)
  - Material properties: Strength, modulus, corrosion resistance, biocompatibility, and wear resistance
  - Many systems provide porous system for permanent attachment by bone ingrowth
  - Types currently available: Bryan, Prestige, ProDisc-C, Cervicore, Porous Coated Motion (PCM), Cervidisc, Mobidisc Cervical
- Various ball-and-socket-type designs of cervical artificial discs
  - Motions at implant level increase following TCDR
  - No major kinematic differences among disc designs
  - Implant stresses higher in spherical designs
  - Facet loads lower for designs with inferior ball component
    - Capsule ligament strains lower for oval design with inferior ball component
  - Oval design with inferior ball component, produced motion, facet loads, implant stresses, and capsule ligament strains closest to intact spine → long-term implant survival

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Ideal patient has single-level soft disc herniation causing neuro signs/symptoms, motion at involved segment, no evidence of osteoporosis or infection
    - Normal sagittal alignment without focal/global kyphosis

  - Neck pain is not a widely accepted indication, although clinical experience suggests decreased neck pain after cervical arthroplasty
- Other signs/symptoms
  - Significant improvement in visual analog scale and neck disability index
  - Complications
    - Cerebrospinal fluid leak, esophageal injury, and wound hematomas were reported in European studies, with overall complication rate of 6.3% per operated level
    - Device wear, subsidence or displacement, segmental hypermobility, excessive disc space distraction to accommodate prosthesis and consequent facet joint separation, screw breakage
    - Extrusion of polyethylene inlay, vertical fractures are device specific
    - Microscopic particulate debris from articulating surfaces can induce immune inflammatory response that leads to osteolysis and prosthetic loosening (particle disease)
      - May be less in cervical fibrocartilaginous joints as compared to proinflammatory synovial joints
    - Paravertebral heterotopic ossification (HO), may be prevented with NSAIDs
      - Studies report from 1.7-7.7% HO; others report higher rate (only 33.8% did not show HO)
      - Fusion of cervical arthroplasty
    - Facet arthrosis, adjacent-level degeneration
    - Persistent neurologic symptoms secondary to inadequate foraminal decompression at initial surgery, clinical failure with need to convert to fusion
      - Reoperation rates reported in 2.5-3% of patients within 1st 2 years following implantation
    - Infection

Demographics

- Age
  - Skeletally mature patients
- Indications
  - Worldwide: 1-2-levels radiculopathy or myelopathy
  - Current Food and Drug Administration investigational device exemption (IDE) investigations limited to single-level disease in levels C3-C7
  - Management of axial pain due to disc degeneration not expected to be better than fusion
  - Candidates are those who have failed conservative therapy and have evidence of symptomatic nerve root ± spinal cord compression
- Contraindications
  - Instability
  - Significant facet arthrosis
  - Osteomyelitis
  - Osteoporosis and osteopenia or other bone metabolic diseases
  - Severe myelopathy due to posterior vertebral body spinal cord compression
  - Tumor
  - Pertinent metallic allergies

Natural History & Prognosis

- Clinical experience with currently available devices is limited to 2-4 years post surgery with success equal or better to fusion and only rare mechanical failures
  - Early favorable clinical results reflect appropriate patient selection, adequate neural decompression, and short-term safety device
  - Longer follow-up necessary to determine whether motion preservation translates into decreased adjacent segment disease
- Studies show in early phase after TCDR, ROM of entire neck, as well as functional and adjacent segments decreased, but return to preop state in late phase
  - Adjacent segmental motion shown to be saved by TCDR as compared to interbody cage fusion
    - Studies show > 2.9% of adjacent intervertebral discs degenerate after anterior decompression and interbody fusion
  - Segmental degenerative kyphosis corrected after TCDR
- TCDR at level adjacent to prior cervical fusion is subject to increased biomechanical forces, potentially leading to higher risk of failure, but is tolerated in short term

Treatment

- At current time, cervical radiculopathy is inclusion criterion for major FDA IDE investigations of cervical arthroplasties

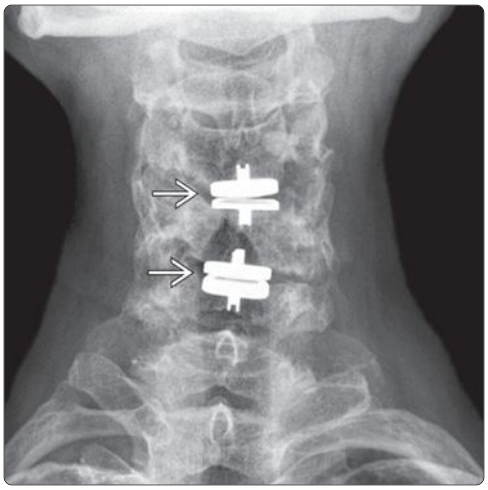
DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Radiography generally is used to evaluate position and integrity of TCDR devices
- CT helpful for detection of heterotopic bone formation and small metallic fragments from device wear
- CT has useful role in evaluating adjacent vertebral bodies, particularly in detecting fractures and degenerative changes at adjacent disc space and facet levels
- MR is beneficial in monitoring development of degenerative disc disease in adjacent motion segments

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(Left) *AP radiograph shows a ProDisc-C at 2 levels ⇨. These can provide near-normal mobility at both levels without destabilizing the implanted segments or affecting motion of adjacent segments, suggesting that single or multilevel cervical TCDR may be advantageous when compared to fusion.* **(Right)** *Lateral radiograph of a ProDisc-C shows a semiconstrained metal-polyethylene design restoring segmental motion, foraminal height, and spinal balance. Rotary motion may stress the facet joints.*





(Left) *Lateral radiograph shows a Bryan cervical disc ⇨, which is composed of porous coated, clamshell-shaped titanium endplates and a polycarbonate, polyurethane core. This prosthesis provides elasticity and compressibility, allowing for unconstrained motion and translation through normal ROM, but can be complicated by anterior and posterior migration, endplate kyphosis, and failure to maintain motion.* **(Right)** *Sagittal T2WI MR shows mild susceptibility artifact at C5-C6 due to a Bryan disc ⇨.*





(Left) *Lateral radiograph shows a Prestige LP TCDR ⇨. This titanium-ceramic device is composed of an articulating ball-and-trough components. Screws attach it to the cervical vertebrae, and there is a porous titanium-plasma spray coating on the endplate surface to facilitate bone growth and long-term fixation.* **(Right)** *Sagittal CT shows a Prestige LP metal-on-metal prosthesis ⇨. It allows AP translation motion of up to 2 mm. CT can be helpful to assess for paravertebral ossification.*

Lumbar Artificial Disc

KEY FACTS

TERMINOLOGY

- Treatment of degenerative disc disease (DDD) restoring normal mobility of diseased segments and improving clinical outcomes by decreasing risk of adjacent-level DDD

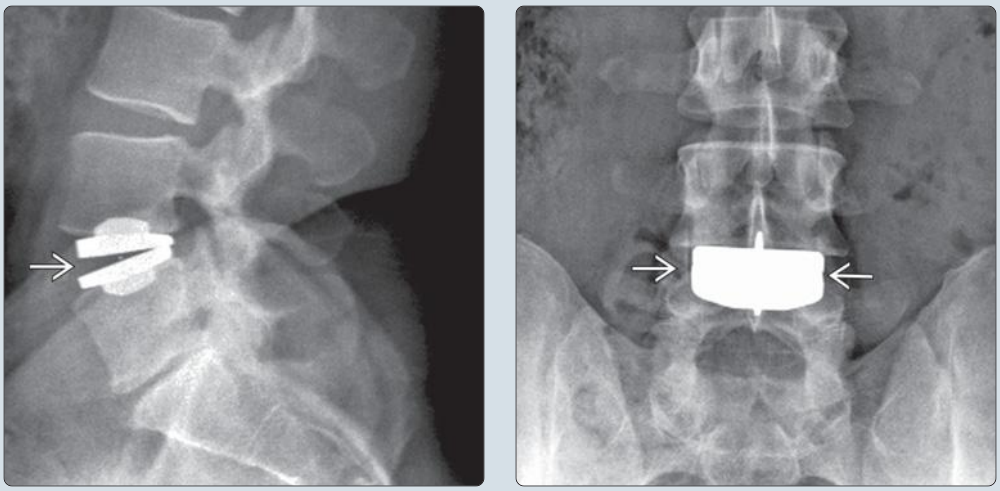
IMAGING

- Endplate keel, fixation spikes, and inlay may help identify individual devices
- Radiography helpful to identify midline placement, AP positioning, and degree of vertebral body penetration or subsidence
- CT best for evaluating extent of vertebral body penetration
- CT and MR helpful to evaluate adjacent vertebral bodies, particularly in detecting fractures and degenerative changes at adjacent disc space and facet levels

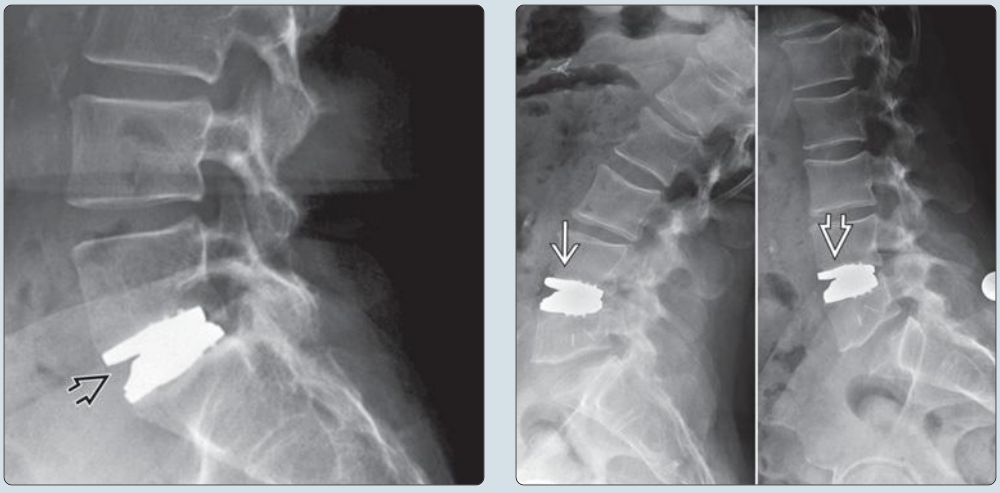
CLINICAL ISSUES

- Lumbar disc arthroplasty indicated for 1- or 2-level discogenic mechanical back pain primarily in absence of radiculopathy
- Complications
  - Heterotopic ossification (HO) incidence in 1.4-15%
  - Adjacent level degenerative disease, facet arthrosis
  - Device migration, extrusion of inlay
  - Subsidence: 3-10% incidence
  - Segmental lordosis alterations
  - Vertebral fractures
- Indications
  - Skeletal maturity
  - Degenerative disc disease at 1 level from L4 through S1
  - No relief of pain after 6 months or more of nonsurgical treatment
- Use is controversial, with some studies showing insufficient evidence to support superiority or routine use of lumbar arthroplasty for symptomatic lumbar degenerative disc disease, even in highly selected populations that meet exclusion criteria

(Left) *Lateral radiograph shows an Activ-L lumbar artificial disc. Arthrodesis is the gold standard for surgical treatment of lumbar degenerative disease; solid fusion may increase motion in segments adjacent to the fused level. This may initiate &/or accelerate adjacent-segment disease process.* **(Right)** *Activ-L allows for maintenance/restoration of physiologic movement at affected segments. Restoring and maintaining normal motion of the segment reduces stresses and loads on adjacent levels.*



(Left) *Lateral radiograph shows the FlexiCore TLDR. Current indications for lumbar disc arthroplasty are young, nonosteoporotic patients with 1- or 2-level symptomatic disc degeneration without severe facet arthropathy, segmental instability, or neural element compression requiring a posterior decompression.* **(Right)** *Extension and flexion views show relatively preserved range of motion. The normal spinal mobility and biomechanics are thought to reduce adjacent spinal deterioration.*



TERMINOLOGY

Synonyms

- Total lumbar disc replacement (TLDR), arthroplasty

Definitions

- Treatment of degenerative disc disease (DDD) by restoring normal mobility of diseased segments and decreasing risk of adjacent-level disc degeneration
- Removal of diseased disc and insertion of disc prosthesis
  - Main goals: Restoration and maintenance of normal or near-normal disc space motion during flexion and extension, right and left lateral bending, and axial rotation

IMAGING

General Features

- Best diagnostic clue
  - Intervertebral radiodense implant
- Location
  - Midline intervertebral placement
- Morphology
  - Endplate keel, fixation spikes, and inlay may help identify individual devices

Radiographic Findings

- Evaluate whether device in proper position
  - Midline placement, anteroposterior (AP) positioning, and degree of vertebral body penetration or subsidence
- No significant difference between conventional radiography and CT in estimation of midline placement
- Charité: Inlay surrounded by metal wire, 3 small spikes positioned on anterior and posterior aspects of both superior and inferior endplates to enhance stability
- Activ-L: Inlay has small radiopaque dot at center

CT Findings

- Ideal location is midline between 2 pedicles
- Should be no penetration of endplate
  - CT better at evaluating extent of vertebral body penetration
- Center of rotation should be located in posterior half of disc space, but implant should not extend beyond posterior vertebral body line
- CT good for detection of heterotopic bone formation and small metallic fragments

MR Findings

- Limited by metal-related artifacts

Imaging Recommendations

- Best imaging tool
  - Radiography helpful to identify midline placement, AP positioning, and degree of vertebral body penetration or subsidence
  - CT best for evaluating extent of vertebral body penetration
- Protocol advice
  - Flexion, extension, and lateral bending views helpful to evaluate range of motion

DIFFERENTIAL DIAGNOSIS

Interbody Fusion Devices

- Fusion across disc space to increase stability of operated segment(s)
  - Solid bony fusion to restore and maintain disc space height and normal sagittal contours

Cages

- Used to reconstruct anterior spinal column after corpectomy

PATHOLOGY

Staging, Grading, & Classification

- Multiple different types and vendors
  - Charité: 2 cobalt-chromium (Co-Cr) endplates with polyethylene inlay or core
  - ProDisc II: 2 cobalt chrome alloy (Co-Cr-molybdenum) endplates and radiolucent polyethylene inlay
    - Endplates have ridged metallic keel inserted into adjacent vertebral body to increase stability
  - FlexiCore: Co-Cr-Mo metal-on-metal device with ball-and-socket mechanism, **no** radiolucent inlay, 3 fixation spikes along lateral margins of endplates
  - Maverick: Ball-and-socket, keel that is embedded within adjacent vertebral bodies to maintain positioning, **no** radiolucent inlay
  - Mobidisc: Keel for stabilization and has 3-part design, with radiolucent polyethylene inlay between the 2 endplates
  - Kineflex: Trials in USA, keel for stability and has 3-part design with a metal inlay
  - Activ-L: Variable design with keel ± spikes for stability, 3-part device with polyethylene inlay

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Lumbar disc arthroplasty is indicated for 1- or 2-level discogenic mechanical back pain primarily in absence of radiculopathy
  - Radiculopathy is exclusion criterion for prospective investigational device exemption (IDE) Food and Drug Administration (FDA) studies on lumbar disc replacement
- Complications
  - Heterotopic ossification (HO) incidence in 1.4-15%
    - Ossification along lateral aspect of device results in fusion, which causes reduced movement of device, whereas anterior ossification results in continued mobility
    - Grade 0: No HO
    - Grade 1: HO presents as island of bone without affecting intervertebral space
    - Grade 2: HO with new bone formation in disc space without blocking or articulating between adjacent endplates
    - Grade 3: Bridging ossifications resulting in limited range of motion of prosthesis
    - Grade 4: Complete bony ankylosis without movement of TLDR in flexion and extension

- Such ossification is most likely to develop after 5th postimplantation year
- Adjacent level degenerative disease
  - Studies report between 4.6-28.6% rate
  - Range of 5° of motion may represent protective threshold against adjacent-level degeneration
- Facet arthrosis
  - Device placement too far anteriorly may generate facet loads up to 2.5x greater in posterior segment compared to loads generated by optimal positioning placement
  - Associated with subsidence
- Device migration
  - Reported with Charité and ProDisc
  - Attributed to oversized device and placement too far anterior within disc space
  - Prosthesis that has migrated should be carefully evaluated for encroachment on aorta and iliac vessels
- Polyethylene inlay may extrude
  - Reported with Charité and ProDisc
  - Increased shear forces created by motion in presence of device may result in pedicle fractures with subsequent dislocation of inlay
- Wear particulate implant debris can induce inflammatory response mediated by various cytokines
  - Pain, osteolysis, pannus formation, prosthetic loosening
- Neurologic complications ≤ than that for interbody fusion with cages or posterior instrumentation
- Anterior approach: Retrograde ejaculation
- Subsidence: 3-10% incidence
  - Implanted device sinks into adjacent vertebral body
  - Endplate loading results in reactive endplate changes, which could lead to pain and ultimate failure
  - Occurrence linked to use of undersized prosthesis
- Segmental lordosis alterations and increase in load through facet joints for both semiconstrained and unconstrained artificial discs
  - Semiconstrained devices may share a greater part of the load, protecting surrounding biological structure from overloading and possible early degeneration
    - May be more susceptible to wear
- Vertebral fractures
  - Maverick and ProDisc require surgical groove into adjacent vertebral body endplates, which may cause weakening and consequent fractures
- Metallosis in adjacent soft tissues, thought to result from impingement of broken marker rings between endplates

Natural History & Prognosis

- Cadaveric studies have shown that artificial disc either maintains or reduces adjacent level motion and intradiscal pressure compared with intact spine
- Currently no evidence to suggest that use of disc arthroplasty results in better short- or long-term functional outcomes than fusion in properly selected patients

Treatment

- Use is controversial, with some studies showing insufficient evidence to support superiority or routine use of lumbar arthroplasty for symptomatic lumbar degenerative disc disease, even in the highly selected populations that meet exclusion criteria
- Indications
  - Skeletal maturity
  - Degenerative disc disease at 1 level from L4 through S1
  - Spondylolisthesis of ≤ 3 mm
  - No radiculopathy
  - No relief of pain after 6 months or more of nonsurgical treatment
- Contraindications
  - Active systemic infection or infection localized to degenerative disease site
  - Osteoporosis, osteopenia
  - Lumbar vertebral stenosis
  - Allergy or sensitivity to materials used in prosthesis manufacture
  - Isolated radicular compression syndrome, especially when due to disc herniation
  - Pars defect
  - Facet arthrosis

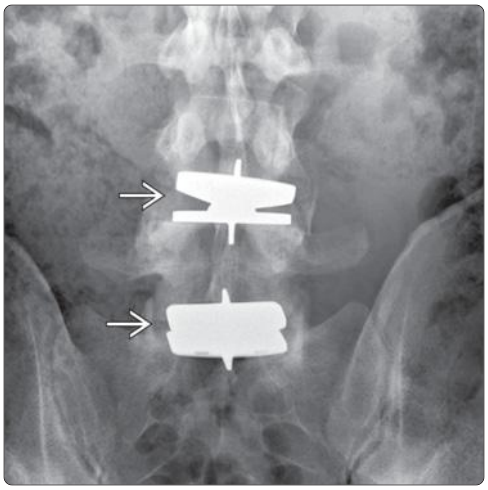
DIAGNOSTIC CHECKLIST

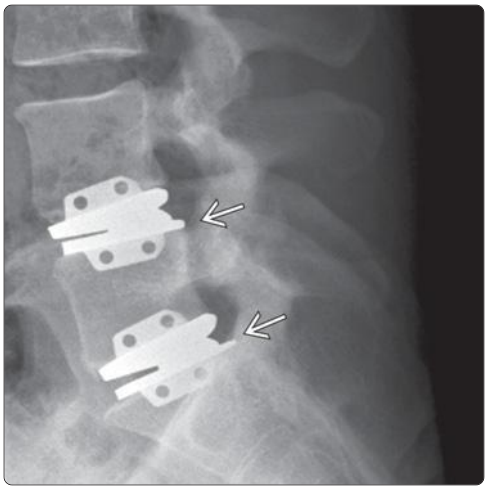
Image Interpretation Pearls

- Evaluate whether device is in proper position
  - Midline placement, AP positioning, and degree of vertebral body penetration or subsidence
- Look for complications: Device migration, polyethylene inlay extrusion, heterotopic bone, wear particulate implant debris, adjacent level disease, facet arthrosis, vertebral fractures

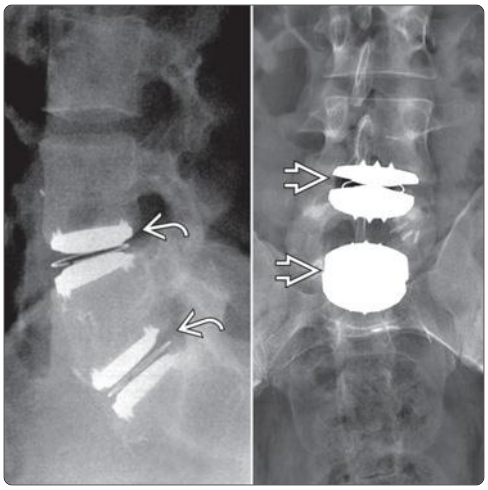
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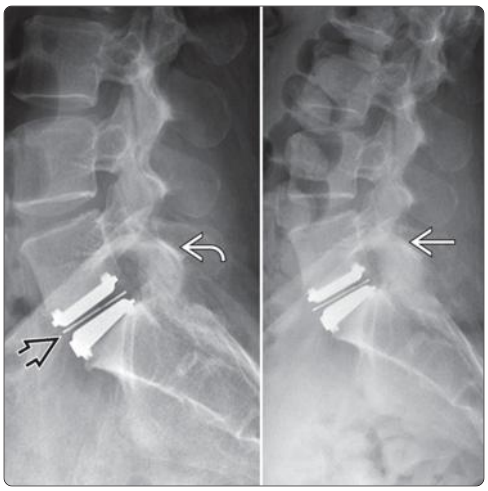


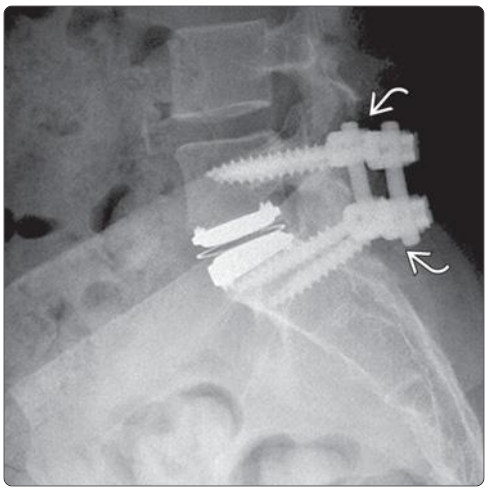
(Left) *Two-level TLDR with Maverick-type of prosthesis is shown. Cadaveric studies show that 2-level lumbar arthroplasty maintains a more favorable biomechanical environment at adjacent segments compared with conventional transpedicular fixation technique.* **(Right)** *Two-level TLDR with Maverick-type prosthesis is shown. Use of 2-level TLDR may have a higher incidence of complications than in cases with 1-level replacement. Two-level TLDR may be associated with progression of facet degeneration.*





(Left) *Lateral and AP x-rays show 2-level Charité prosthesis. Literature shows ~ 9% of arthroplasty patients have progression of asymptomatic radiographic degeneration of adjacent functional spinal unit and 1% of symptomatic adjacent-level degenerative disease. 2-level annulus lumbar resection required for multilevel Charité implantation can add destabilizing effect, resulting in 140-160% ROM increase over intact condition.* **(Right)** *Lateral CT shows a right pedicle fracture due to an increase in posterior stress.*





(Left) *Lateral radiograph shows L5-S1 facet joint in the early postoperative period after placement of Charité TLDR. Increased stress on L5-S1 facet joints indicates progression of arthropathy. Patient had initial improvement of symptoms but experienced gradual recurrence of low back pain.* **(Right)** *Several years after Charité TLDR, this patient had recurrent low back pain and correlative findings on discogram at L5-S1. Lateral radiograph shows revision with posterior transpedicular fusion.*

Hardware Failure

KEY FACTS

TERMINOLOGY

- Mechanical breakdown or malfunction of hardware

IMAGING

- Fractured or malpositioned metallic implant
- Lucency ± sclerosis along implant or at vertebral body-graft interface
- Adjacent levels may experience biomechanical stresses with resulting marrow edema and accelerated degenerative changes
- Plain films excellent in evaluating vertebral alignment, hardware integrity, fusion status
- Flexion-extension views if hardware failure present
- CT evaluation if implant breakage suspected, but not definitive on radiography
  - Especially with complex constructs &/or in osteopenic patients
  - Enables accurate assessment of degree of osseous fusion; however, surgical exploration remains reference standard for evaluating fusion
- Soft tissue complications, marrow edema (suggesting segmental instability), or spinal cord injury

PATHOLOGY

- Fibrous union may provide satisfactory stability in absence of radiographic osseous fusion

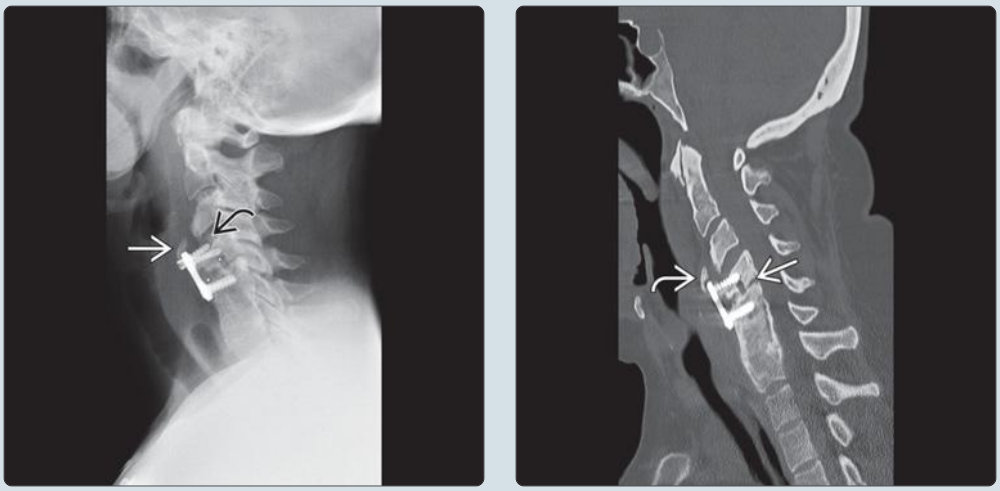
CLINICAL ISSUES

- Pain, tenderness, radiculopathy

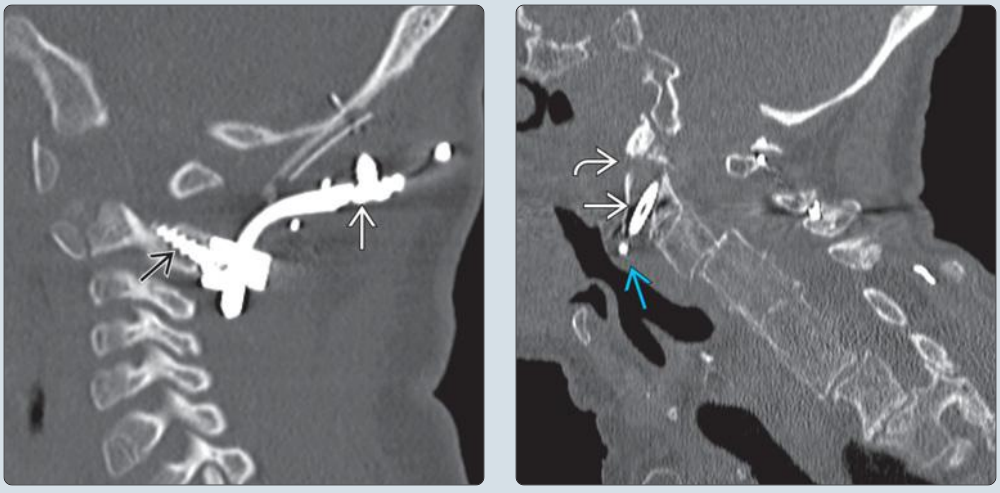
DIAGNOSTIC CHECKLIST

- Compare with multiple prior studies to identify subtle progressive changes
- Failed fusion may indicate unsuspected ligamentous injury in trauma or tumor recurrence or progression

(Left) *Radiograph shows lucency around and displacement of the C4 corporal screws. There is a positive association between adjacent-level ossification following anterior cervical plate procedures and plate-to-disc distance, especially if < 5 mm.* **(Right)** *In this case, there is lack of fusion of the C4-C5 disc space. Adjacent-level ossification within the 1st 12 postoperative months has a substantial likelihood of progression to advanced ossification by 24 months.*



(Left) *Sagittal NECT (bone window) in this child with Hurler syndrome shows that the fixation screws/plate have pulled away from the occipital bone. The proximal fixation screw in the C2 lateral mass is in expected position.* **(Right)** *Sagittal NECT (bone window) shows a failed single screw fixation of a type II odontoid fracture with screw loosening, as well as the screw backing out and extending into the retropharyngeal soft tissues.*



TERMINOLOGY

Synonyms

- Implant or prosthesis failure

Definitions

- Mechanical breakdown or malfunction of hardware
  - Pseudoarthrosis, subsidence

IMAGING

General Features

- Best diagnostic clue
  - Fractured or malpositioned metallic implant
  - Lucency ± sclerosis along implant or at vertebral body-graft interface
- Location
  - Any spinal segment where instrumentations occur
    - Adjacent levels may experience biomechanical stresses with resulting marrow edema and accelerated degenerative changes

Radiographic Findings

- Radiography
  - Cervical spine
    - Lucency along cannulated screw bridging type II odontoid fracture
    - Broken, extruded screw
    - Fractured, ventrally displaced plate
    - Dislodged allograft
    - Broken, detached posterior cervical wire
  - Thoracolumbar and lumbosacral spine
    - Broken sublaminar or subpars wire
    - Bent, loose, or fractured pedicle screw
    - Disengaged hook
    - Broken, dislodged rod
    - Intervertebral cage/allograft migration
  - Pseudoarthrosis
    - Lucency between bone graft and adjacent vertebra
    - Sclerosis and rounding of unfused bones
  - Development or progression of vertebral malalignment
  - Progressive graft collapse and increasing kyphosis

Fluoroscopic Findings

- Pseudoarthrosis on flexion/extension views
  - 4 mm of translation or > 10° of angular motion between adjacent vertebrae
    - Up to 3 mm of translation considered normal
  - ≥ 2 mm change in interspinous distance on flexion and extension

CT Findings

- Bone CT
  - Peri-implant lucency (suggests loosening)
  - Overextension beyond posterior vertebral cortex by cervical bicortical screws
  - Medial cortex penetration by suboptimally placed lumbar pedicle screw
  - Occult osseous fractures
    - Altered biomechanics with fusion may result in adjacent level stress reaction
  - Osseous nonunion, pseudoarthrosis

MR Findings

- MR generally does not depict hardware location or integrity well
  - Nondiagnostic with extensive artifact from steel hardware
  - Titanium hardware produces less artifact
- MR useful for demonstrating impact on surrounding soft tissue and cord morphology
  - T2/STIR hyperintensity can be seen in adjacent level posterior elements due to stress reaction

Nuclear Medicine Findings

- Bone scan
  - Increased uptake at fusion site suggests nonunion
    - Nonspecific until 1 year after surgery
    - Fused segment should be "cold" after 6–12 months
    - Also useful for detecting infection

Imaging Recommendations

- Best imaging tool
  - Plain films excellent in evaluating vertebral alignment, hardware integrity, fusion status
    - Cost effective
    - Flexion/extension views
    - Cannot be used to reliably exclude presence of bone metastases or of cauda equina compression
  - CT evaluation if implant breakage suspected, but not definitive on radiography
    - Especially with complex constructs &/or in osteopenic patients
    - Enables accurate assessment of degree of osseous fusion; however, surgical exploration remains reference standard for evaluating fusion
  - MR to identify soft tissue complications, marrow edema, suggesting segmental instability, or spinal cord injury
- Protocol advice
  - MR techniques to minimize susceptibility artifact
    - Low magnetic field strength minimizes metal artifact
    - Fast spin-echo technique; avoid gradient-echo sequences
      - Higher receiver bandwidths, short TE, small voxel
    - SE/FSE susceptibility artifact (signal loss and distortion) projects along frequency encoding direction
      - Frequency encoding direction should be chosen along long axis of hardware
  - Increasing kilovolt peaks/milliamperes to reduce beam hardening on CT

PATHOLOGY

General Features

- Etiology
  - Excessive stress loading on implant
    - Implant malpositioning at surgery
    - Gross spinal instability
    - Failed fusion with pseudoarthrosis
  - Poor bone quality
    - Peri-implant bone resorption
    - Osteoporosis
    - Osteomyelitis

  - Residual or recurrent neoplasm
- Multisegmental constructs
- Unconstrained cervical fusion plates: Orozco, Casper
  - Screw not locked to plate with risk of screw extrusion
- Risks of pseudoarthrosis
  - Comorbidities: Old age, smoking, obesity, diabetes
  - Multiple spine surgeries
  - Multilevel fusions
  - ≥ grade III anterolisthesis
- Particle disease with disc prostheses
  - Particles resulting from wear debris cause macrophage activation and phagocytosis → joint loosening and implant failure
  - Well-defined osteolytic lesion-mimicking tumor
- Bone morphogenic protein (BMP) in anterior cervical fusion procedures associated with higher complication rate
  - Primary increases seen in wound-related complications and dysphagia or hoarseness
- BMP, particularly in transforaminal lumbar interbody fusion and posterior lumbar interbody fusion, associated with severe postoperative radiculitis
  - Proinflammatory reaction ± ectopic bone formation

- Associated abnormalities
  - Pseudoarthrosis, instability, fractures
  - Dural laceration, nerve injury
- Hardware intended to stabilize fusion construct while awaiting successful osseous fusion
  - All hardware eventually fails if fusion does not occur in timely fashion
  - Fusion rate improved with direct current electrical stimulation
  - Fusion occurs after 6-9 months, up to 18 months
- Fibrous union may provide satisfactory stability in absence of radiographic osseous fusion
  - Best confirmed with dynamic flexion-extension views

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be incidental finding
- Other signs/symptoms
  - Pain, weakness, paresthesia, radiculopathy
  - Dysphagia/esophageal perforation due to cervical plate migration
- Clinical profile
  - Hardware failure in early postoperative period
    - Indication of continued gross spinal instability
  - Development or progression of neurologic symptoms
    - Osseous nonunion &/or hardware failure should be suspected

Demographics

- Epidemiology
  - 2-45% reoperation rate for implant failure
  - Hardware failure in scoliosis surgery
    - 31% with anterior approach
    - 1% with posterior approach
  - Unconstrained cervical fusion system
    - 22-46% failure rate vs. 18% in constrained system
  - Lumbar fusion with unconstrained pedicle screw system
    - 22% failure rate
      - 75% due to pseudoarthrosis

Natural History & Prognosis

- Osseous fusion may be expected even if hardware fails
  - Broken hardware need not be removed if spine clinically, radiographically stable
- Fibrous union without radiographic osseous fusion may be satisfactory
  - Best demonstrated on dynamic flexion-extension views
- Repeat surgical fusion may be necessary
  - Especially if hardware failure occurs early

Treatment

- Conservative observation
- Surgical revision to prevent nonunion and instability

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

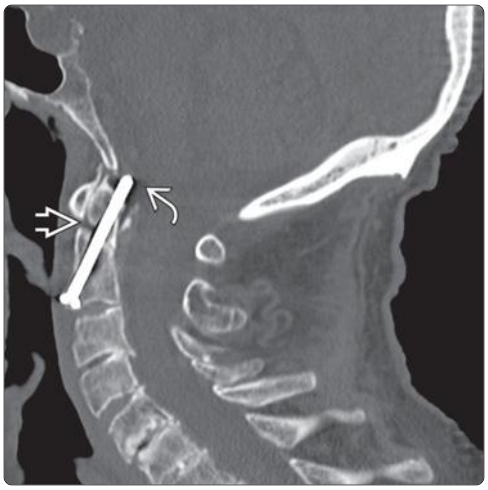
- Important not only to evaluate hardware failure, but also to look for complicating instability or osseous fx
- Important to consider original indication for fusion in suspected implant failure
  - Failed fusion for trauma may indicate unsuspected ligamentous injury
  - Failed fusion for neoplasm may indicate tumor recurrence or progression

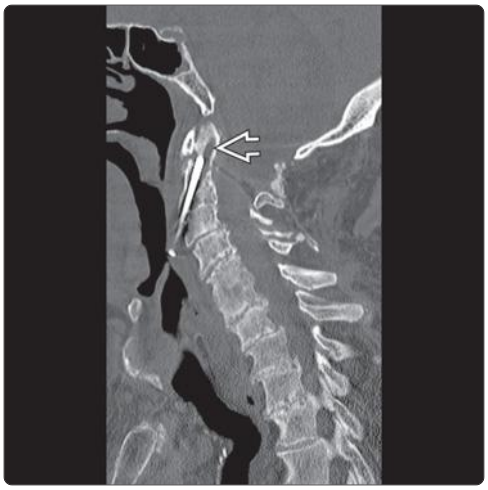
Reporting Tips

- Important to compare current radiographs with multiple previous studies to identify subtle progressive changes (e.g., in spinal alignment and in position of hardware devices)

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(Left) *Sagittal reformatted CT shows an anterior odontoid screw fixation of a type II odontoid fracture that provides spinal stability and preserves normal atlantoaxial rotation. The screw tip is fully engaged into the apical cortex. Extension by few mm into the apical ligaments is safe.* **(Right)** *Lucency along odontoid screw suggests persistent motion. Acute odontoid fractures treated with anterior screw fixation have a fusion rate from 73-90%. In chronic odontoid fractures, however, there is a high chance of nonunion.*





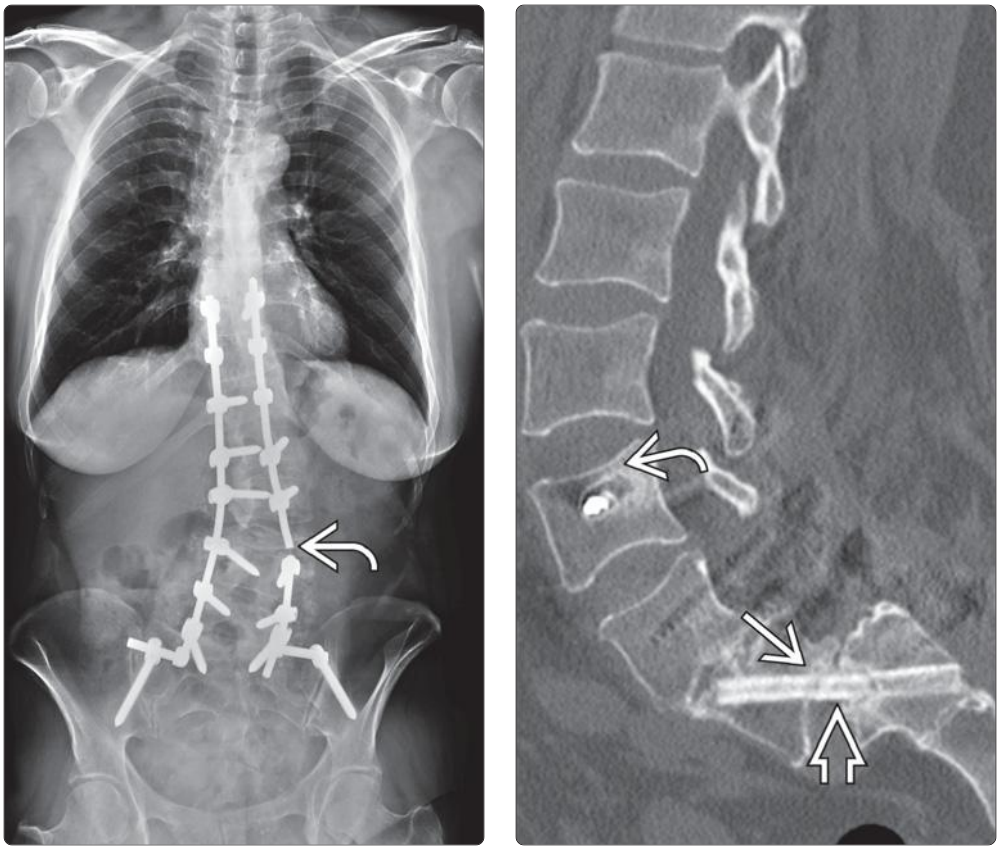
(Left) *Lateral radiograph shows break of the right L4 pedicle screw. Early hardware failure may result from technical insertion stress or failure related to screw/rod design. Posterior metallic marker of the interbody spacer should be located at least 2 mm anterior to the posterior vertebral body margin.* **(Right)** *AP radiograph shows a fractured right L4 pedicle screw. Poor fusion mass, segment motion, lucency around screws, and screw fracture are compatible with failed fusion.*



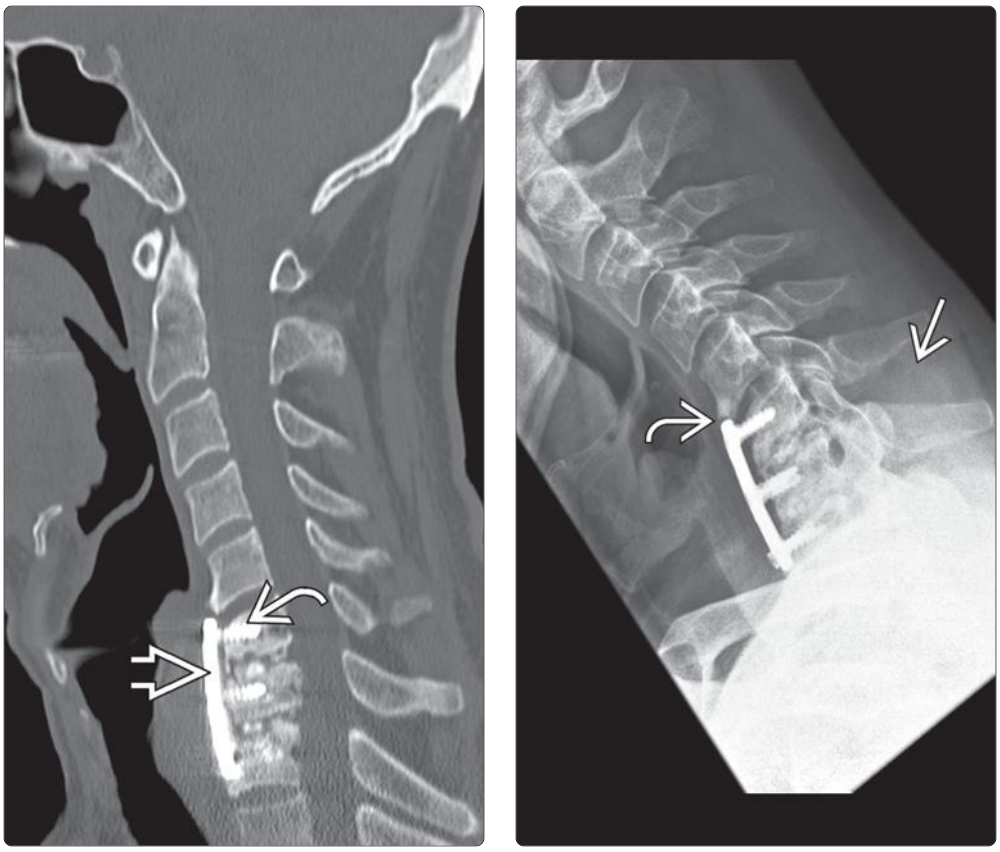


(Left) *Lateral radiograph shows anterior cervical discectomy and fusion at the C5-C6 level. A radiolucent stripe at the vertebral bone-graft interface is observed. There is slight reversal of normal cervical lordosis in the neutral position.* **(Right)** *Lateral radiograph shows that, upon flexion, there is > 2 mm interspinous widening at the C5-C6 fused level, suggesting abnormal motion. There is no subluxation with flexion. The reference standard for diagnosis of pseudoarthrosis is surgical exploration.*

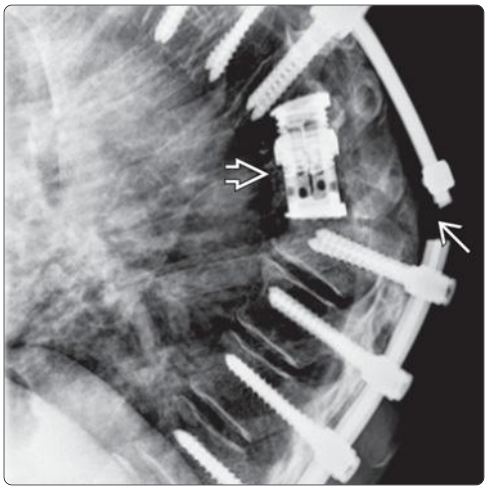
(Left) *Anteroposterior radiograph shows segmental pedicle fusion for levoscoliosis and sacroiliac fusion. There is a fracture of the left posterior rod at the L3-L4 level. Rod breakage signifies pseudoarthrosis (incidence: 5%). Surgeons agree that the fusion should be extended to the sacrum if there is L5–S1 spondylolisthesis, if there has been a previous L5–S1 laminectomy, if there is any form of L5–S1 stenosis, if there is an oblique takeoff at L5–S1, or if there is severe L5–S1 disc degeneration.* **(Right)** *Lucency and sclerosis along the L3 pedicle screw represent osteolysis secondary to screw motion, which develops with bone resorption. An interbody fibular fusion graft and grade 3 anterolisthesis are seen at L5-S1. Linear irregular lucency through new bone formation within the disc space indicates delayed fusion.*



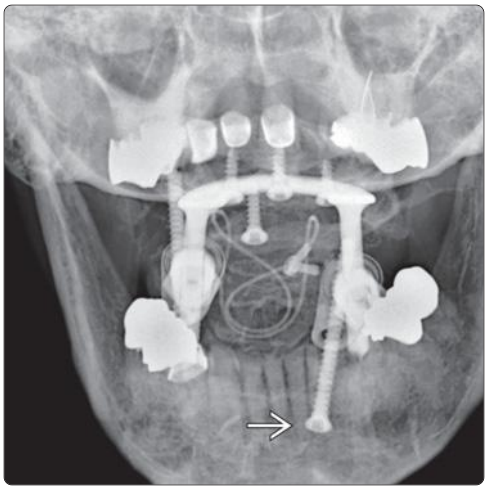
(Left) *Sagittal CT shows anterior cervical discectomy and fusion from C6-T1. Cystic changes along the adjacent endplates represent osteolysis secondary to device motion, which develops with bone resorption. Linear defects in intradiscal new bone within or adjacent to the fusion device parallel to the endplates can indicate failed fusion.* **(Right)** *Cervical spine in flexion shows anterior cervical discectomy and fusion (ACDF) at C6-T1. The superior margin of the fusion plate overlaps the C5-C6 disc space. The fixation plate impacts the C5 inferior anterior endplate. Ossification can occur cephalad and caudal to anterior cervical plates if the margins are within 5 mm of the adjacent disc spaces. There is irregularity along the endplates, lucency along the graft-vertebral interface, and abnormal widening of the interspinous distance.*







(Left) *Axial NECT shows lucency and sclerosis along bilateral pedicle screw tracts ⇨. Lucency suggests movement at the operated level and loosening of the screw. Such lucency is associated with delayed or failed fusion.* **(Right)** *Lateral radiograph demonstrates an expandable cage spanning a fractured midthoracic vertebral body ⇨. There is a break in the posterior interconnecting rod ⇨ and exaggerated focal kyphosis. A potential complication of cages is subsidence into the vertebral body.*





(Left) *Anteroposterior radiography in a patient with C0-C2 fusion shows asymmetric screw placement ⇨.* **(Right)** *Lateral radiography of patient with C0-C2 fusion demonstrates dorsal extrusion of left transarticular screw ⇨ from C1 and C2 lateral masses.*





(Left) *An expandable cage spanning the L2 vertebral body ⇨ was placed for spinal reconstruction after osseous metastasis resection. Posterior fusion is also observed ⇨.* **(Right)** *Sagittal CT myelography shows further collapse of the L3 vertebral body due to a lytic renal cancer metastasis. The metastatic lesion results in collapse of the fusion construct. There is retropulsion of bone with a soft tissue component causing severe thecal sac narrowing and a myelographic block at the L2-L3 level ⇨.*

Bone Graft Complications

KEY FACTS

TERMINOLOGY

- Synonyms: Graft migration, graft displacement, graft extrusion
- Abnormal alignment, position, placement of graft or hardware ± associated neurologic deficit, instability, infection

IMAGING

- Plain films essential for rapid intraoperative/postoperative evaluation of graft position
- MDCT with reformats shows graft position, endplate integrity, graft/vertebral body fractures
- Autogenous graft may show ↑ T1 signal with fatty marrow or ↓ signal due to edema
  - Allograft generally low signal
- T1WI C+ little use in acute phase for graft malposition or epidural hemorrhage; mandatory if question of infection

TOP DIFFERENTIAL DIAGNOSES

- Abscess
- Hemorrhage
- Infarction
- Pseudoarthrosis

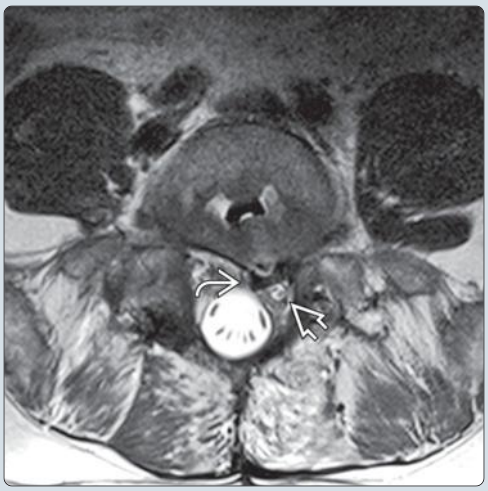
PATHOLOGY

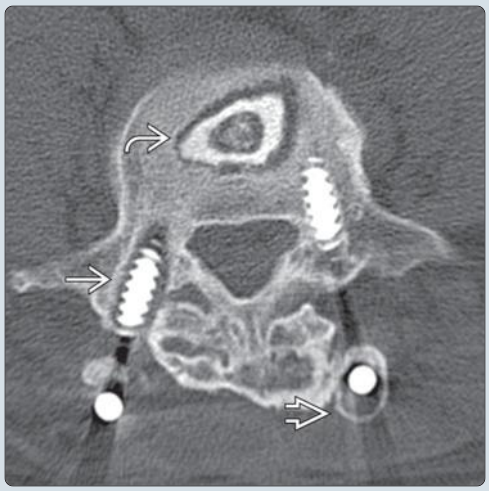
- Graft migration rate increased with more levels of fusion
- Increased graft migration with strut graft compared to interbody graft
- Graft migration occurs in 7% of cases after anterior cervical corpectomy

CLINICAL ISSUES

- May require reoperation for graft migration/extrusion/collapse

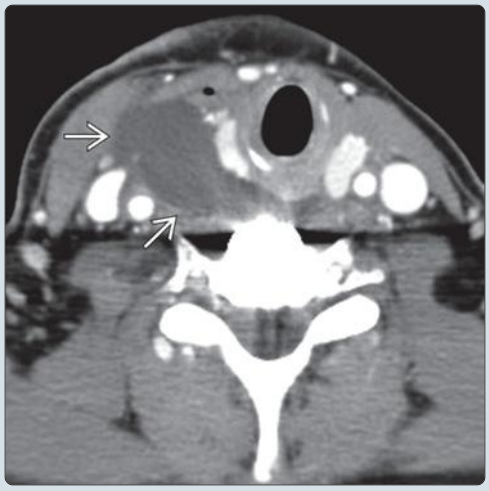
(Left) *Axial T2WI MR in a patient following laminectomy and posterior lumbar interbody fusion (PLIF) shows posterior positioning of the lumbar interbody graft at the L4-L5 level. The posterior margin of the graft contacts the dural margin and the exiting left L5 root.* **(Right)** *Axial NECT in nonunion shows prominent lucency surrounding bone graft and pedicle screws in mid and distal aspects. There is lucency and bony remodeling surrounding the junction with the longitudinal rod.*





(Left) *Axial NECT shows effect of rhBMP-2 osseous remodeling with focal vertebral body bone resorption. This appearance does not imply failure of fusion and generally will progress to solid fusion.* **(Right)** *Axial CECT following surgery shows CSF leak as a large fluid collection extending from the discectomy and graft site into the prevertebral space and then medial to the carotid and jugular vein.*





TERMINOLOGY

Synonyms

- Graft migration, graft displacement, graft extrusion

Definitions

- Abnormal alignment, position, placement of graft or hardware ± associated neurologic deficit, instability, infection

IMAGING

General Features

- Best diagnostic clue
  - Abnormal position of graft material
- Morphology
  - Well-defined cortical graft material displaced with respect to interbody fusion site

Radiographic Findings

- Radiography
  - Essential for rapid intraoperative/postoperative evaluation of graft position

Fluoroscopic Findings

- Not used acutely; useful for late nonfusion evaluation

CT Findings

- NECT
  - MDCT with reformats shows graft position, endplate integrity, graft/vertebral body fractures

MR Findings

- T1WI
  - Variable appearance of graft material
    - Autogenous graft may show ↑ T1 signal with fatty marrow → ↓ signal (edema)
    - Allograft generally low signal
- T1WI C+
  - Little use in acute phase for graft malposition or epidural hemorrhage; mandatory if question of infection

Imaging Recommendations

- Best imaging tool
  - Plain films define graft position, ventral or dorsal migration, collapse
- Protocol advice
  - MR: T1WI, T2WI, GRE axial + sagittal

DIFFERENTIAL DIAGNOSIS

Abscess

- Typical clinical and laboratory findings of infection (elevated sedimentation rate, C-reactive protein)

Hemorrhage

- Nonspecific T1 signal if acute → early subacute age (no increased signal)

Infarction

- Focal ↑ signal on T2WI in slightly expanded cord

Pseudoarthrosis

- Abnormal low T1 signal extending through disc, posterior elements, and ligaments

PATHOLOGY

General Features

- Etiology
  - Graft migration rate increased with more levels of fusion
  - Graft migration rate increased with fusion ending at C7
  - Increased graft migration with strut graft compared to interbody graft
- Graft migration rates vary widely depending on series, type
  - Graft migration occurs in 7% of cases after anterior cervical corpectomy
- Graft nonunion
  - Strut autograft (5-27%), allograft (40%)
- Increased nonunion rates in multilevel interbody grafting than in corpectomy with strut grafting when more vertebral levels involved
- Similar fusion and complication rates for 2-level discectomy or single-level corpectomy with plates

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be asymptomatic, with only radiographic abnormality
  - New postoperative pain/focal deficit
  - Rare respiratory distress

Demographics

- Age
  - Adult
- Gender
  - M = F

Natural History & Prognosis

- ↑ fusion rate after corpectomy/strut grafting (90%) than after multilevel discectomy/interbody graft (66%)
- Fusion less successful when anterior cervical arthrodesis is performed adjacent to prior fusion

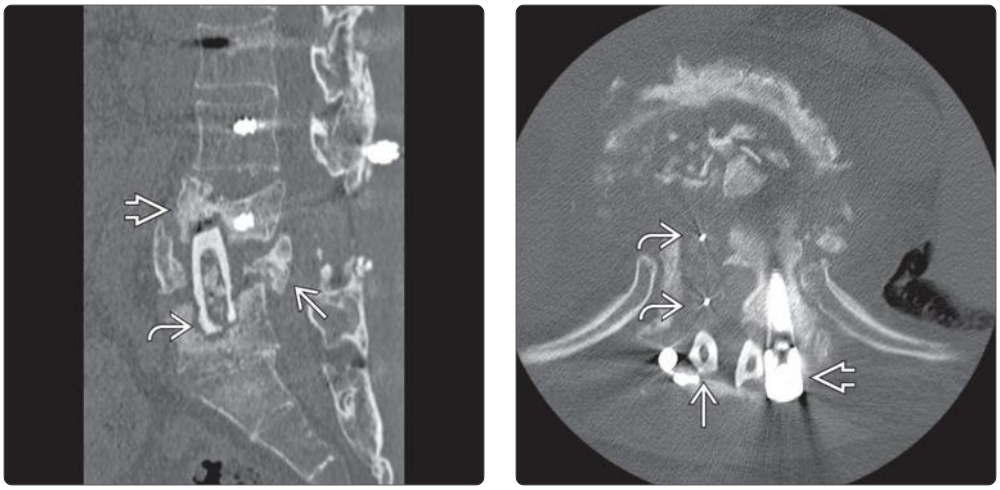
Treatment

- May require reoperation for graft migration/extrusion/collapse

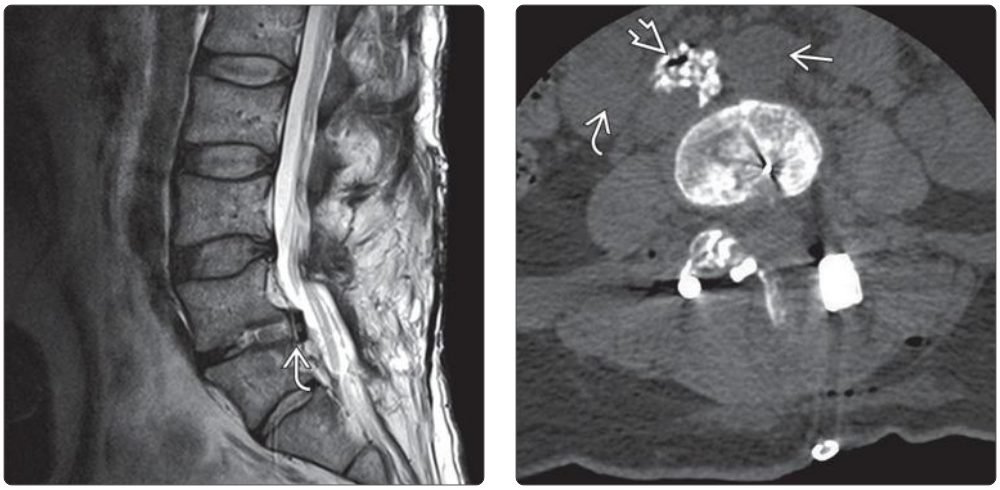
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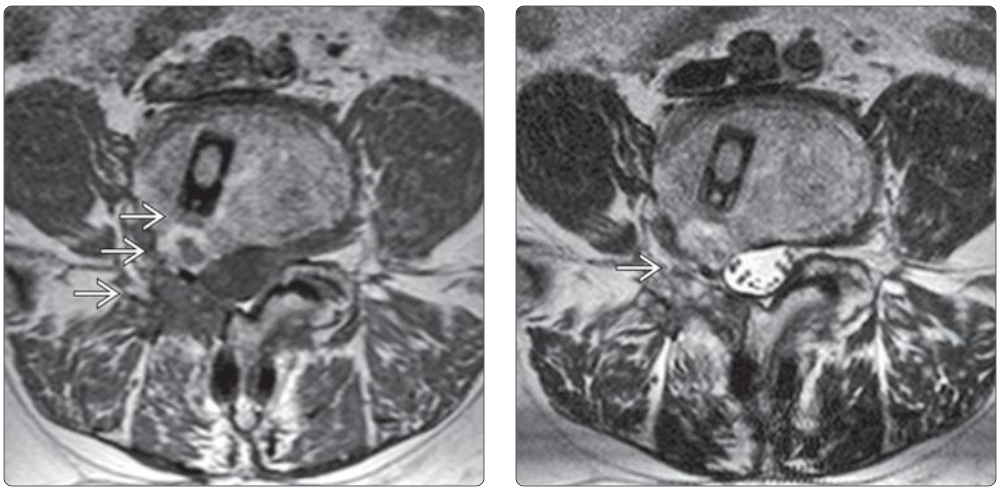
(Left) *Sagittal NECT shows typical pattern of graft nonunion with lucency surrounding the graft placed at the L4 corpectomy site. There is retropulsion of fracture fragment at L4 and collapse of the L3 body with sclerosis.* **(Right)** *Treatment of chronic pseudoarthrosis with instability (Charcot joint) is shown. There is posterior displacement of the interbody fusion graft into the spinal canal. Note posterior graft material and the left pedicle screw.*



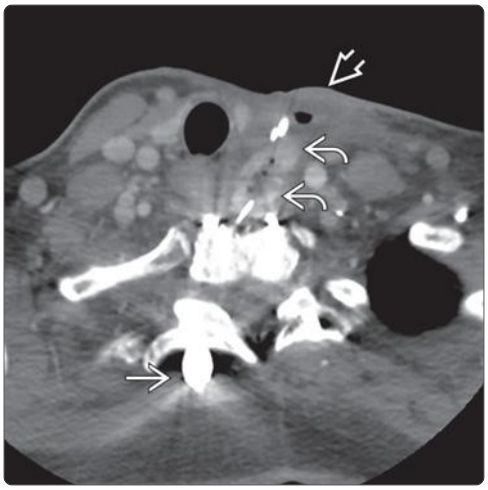
(Left) *Sagittal T2WI MR shows abnormal posterior positioning of the interbody graft at L4-L5 level, deforming the ventral thecal sac and extending into the region of the exiting L5 root.* **(Right)** *Axial CT shows bone graft that has been pushed too far, winding up anterior to the intervertebral disc space (where it should be placed). The graft is positioned between the aorta and inferior vena cava. This was an unexpected postoperative finding.*



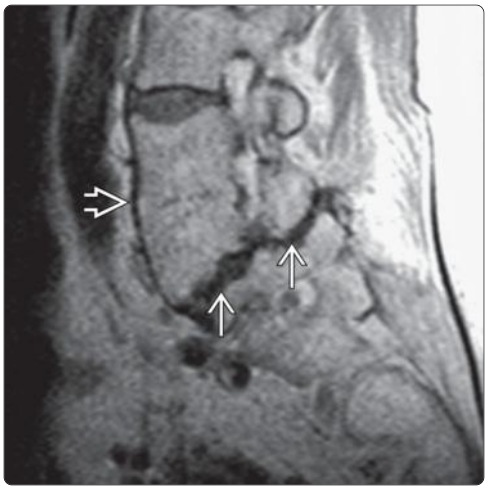
(Left) *Axial T1WI MR shows severe right foraminal stenosis from heterotopic bone formation along the trajectory of prior facetectomy and posterior interbody fusion.* **(Right)** *Axial T2WI MR in the same patient shows the trajectory of prior facetectomy and posterior interbody fusion where the heterotopic bone formation occurred and caused severe right foraminal stenosis.*

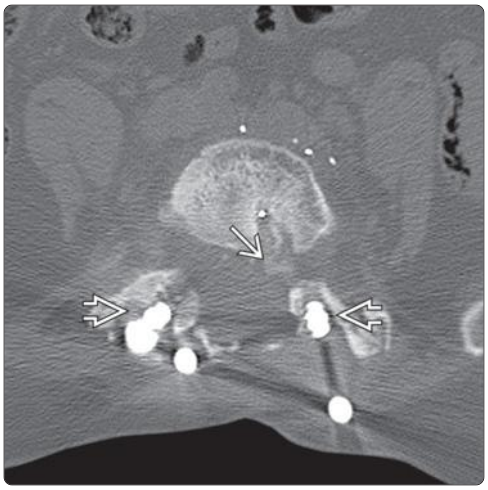






(Left) *Chronic infection complicating cervical fusion with strut graft, with graft extrusion into soft tissues. Initial CECT 2 years prior to latest admission shows C5-C7 fusion with polymer synthetic graft with central core of bony fusion mass.* **(Right)** *Axial CECT 2 years later in the same patient with chronically draining wound on the left neck shows horizontally extruded graft, which now lies within the soft tissues of the anterior neck. Posterior hardware has maintained alignment.*





(Left) *Sagittal T1WI MR shows pseudoarthrosis through L5-S1 level with linear low signal from disc level through posterior elements. Note the solid fusion at L4-L5.* **(Right)** *Axial NECT shows abnormal posterior positioning of graft associated with posterior lumbar interbody fusion. Graft extends into the medial neural foramen. Note the bilateral lucencies about the pedicle screws due to loosening.*





(Left) *Sagittal T2WI MR in a patient with postoperative infection shows collapse of graft at C5-C6, with kyphosis and subluxation with cord compression. Note the substantial prevertebral edema.* **(Right)** *Sagittal T1WI MR in the same patient shows subsequent C5-C6 graft collapse and destruction, with focal kyphosis due to infection.*

rhBMP-2 Complications

KEY FACTS

TERMINOLOGY

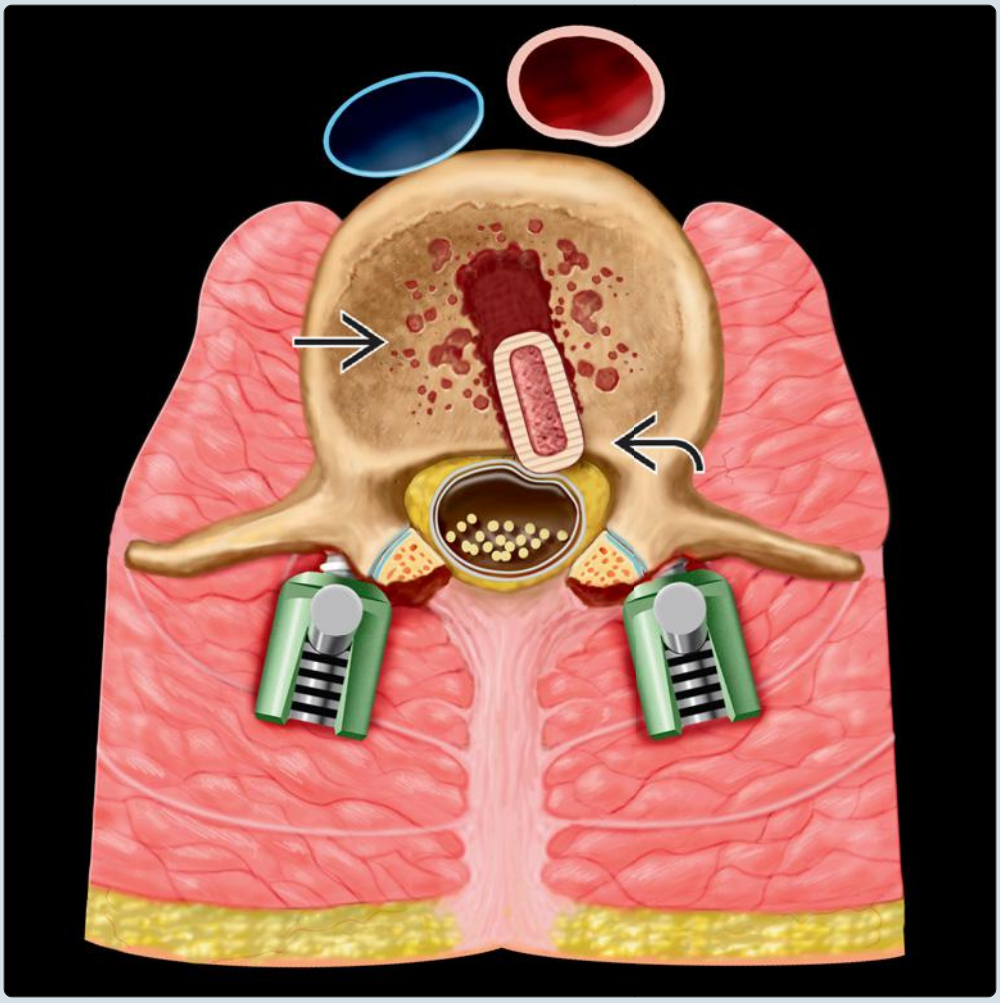
- Recombinant human bone morphogenetic protein 2 (rhBMP-2)
  - Used in surgery to decrease time of graft union and to reduce morbidity from nonunion

IMAGING

- Osteolysis seen as well-defined lucencies at vertebral endplates
- MR shows rounded low T1 signal, high T2 signal lesions along endplates
  - May mimic appearance of early postoperative infection
  - Usually involves adjacent vertebral bodies

TOP DIFFERENTIAL DIAGNOSES

- Heterotopic bone formation
  - Extradiscal, ectopic, and heterotopic bone production
  - Presumably occurs with BMP leakage from carrier into epidural space → canal or foraminal compression
- Resorption/osteolysis
  - Varying rates of occurrence of osteolysis (18-70%)
  - Generally lytic features resolve by 24 months, most by 9 months
- Subsidence/cage migration
  - Maximum osteolysis in 1st 12 weeks after surgery → cage at greatest risk for migration
- Hematomas/seromas
  - 5% of cases of posterolateral fusion in 1 series needed reoperation for exploration of painful seroma
- Soft tissue inflammation
  - rhBMP-2 not recommended for use in anterior cervical approach
  - > 20% of patients with neck swelling



Axial graphic shows lysis of endplates ⇨ associated with rhBMP-2 use in a posterior lumbar interbody fusion. There has been posterior migration of the interbody graft into the epidural space ⇨. Maximum osteolysis occurs in the first 12 weeks after surgery.

TERMINOLOGY

Abbreviations

- Recombinant human bone morphogenetic protein 2 (rhBMP-2)

Definitions

- rhBMP-2
  - Member of transforming growth factors-β super family of proteins
  - Used in surgery to decrease time of graft union and to reduce morbidity from nonunion
  - These types of proteins have roles in cell proliferation, differentiation, apoptosis, and embryologic development
  - BMP induces differentiation of undifferentiated mesenchymal cells into osteogenic cells and enhances function of osteoblasts
  - BMP-induced endochondral ossification → undifferentiated mesenchymal cells develop into chondrocytes
    - Chondrocytes lay down cartilage, which is later resorbed by chondroblasts
    - Allows osteoblasts to begin new bone formation
  - BMPs also have role in regulation of osteoclastogenesis
    - This regulation can lead to stimulation of bone resorption
  - rhBMP-2 commercially supplied in USA as INFUSE bone graft (Medtronic; Minneapolis, MN)
    - Delivered on absorbable collagen sponge
    - FDA approved for use with LT-CAGE (Medtronic; Minneapolis, MN) device for anterior lumbar interbody fusion
  - rhBMP-7 commercially available as OP-1 putty (Olympus Biotech; Hopkinton, MA)

IMAGING

General Features

- Best diagnostic clue
  - Osteolysis seen as well-defined lucencies at vertebral endplates
    - May mimic appearance of early postoperative infection
  - MR shows rounded low T1 signal, high T2 signal lesions along endplates
    - Usually involves adjacent vertebral bodies
    - MR appearance more worrisome for tumor, but adjacent endplate involvement allows for correct diagnosis

DIFFERENTIAL DIAGNOSIS

Heterotopic Bone Formation

- Extradiscal, ectopic, and heterotopic bone production
- Presumably occurs with BMP leakage from carrier into epidural space → canal or foraminal compression
  - > 75% of cases in some series
  - Most prominent with posterior approaches, such as TLIF
- Most are asymptomatic
  - Symptomatic bone production has been reported following transforaminal lumbar interbody fusion

Resorption/Osteolysis

- Normal part of remodeling process that leads to fusion
- May also result in mechanical failure
  - Cage migration
  - Endplate subsidence (6-22%)
  - Fracture
- Varying rates of occurrence of osteolysis (18-70%)
  - Generally lytic features resolve by 24 months, most by 9 months
  - May be extensive with large regions of vertebral bodies involved

Subsidence/Cage Migration

- Maximum osteolysis in 1st 12 weeks after surgery → cage at greatest risk for migration
  - Cage migration generally not going to occur after 6 months
  - Cage migration/subsidence ~ 30% of cases

Hematomas/Seromas

- 5% of cases of posterolateral fusion in 1 series needed reoperation for exploration of painful seroma
- Mean time to reoperation was 7.7 days
- Erythema, tenderness, swelling at wound site
- No clear dose response

Soft Tissue Inflammation

- rhBMP-2 not recommended for use in anterior cervical approach
- > 20% of patients with neck swelling
- Edema and inflammation → not hematoma
- Increased dysphagia with rhBMP use
- Increased expense with no clear improvement in outcomes

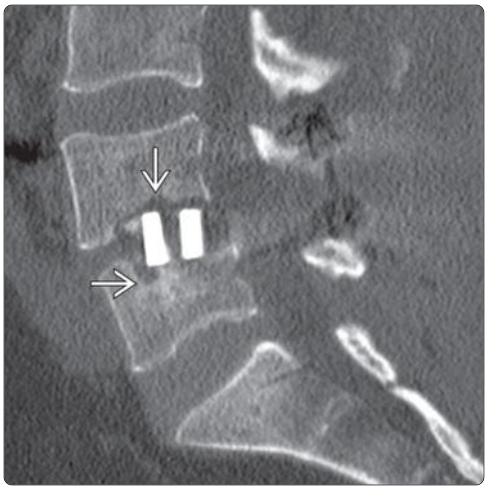
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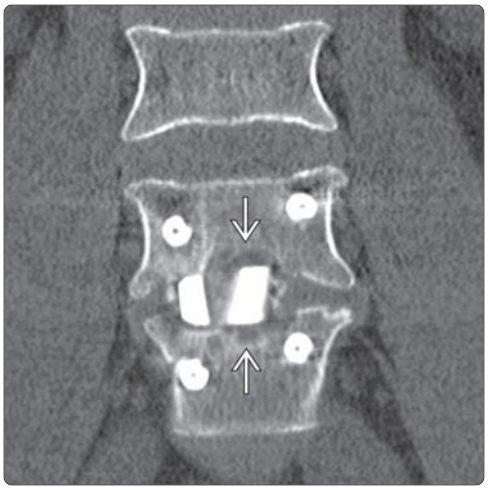
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Complications of rhBMP-2

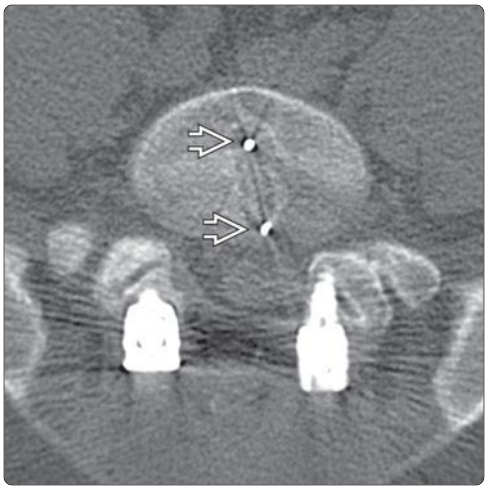
Ranges and Rates of Complications*		
Type	**Range**	**Mean, Rate**
Resorption/osteolysis	0-100%	44%
Cage migration	0-35%	27%
Graft subsidence	0-62%	25%
Radiculitis		11%
Heterotopic bone formation	0-75%	8%
Retrograde ejaculation		7.9%
Dysphagia	3.8-85%	5.8%
Hematoma/seroma	1-9%	4%
Wound dehiscence	0-17%	3%
Antibody formation	0-5%	1%
Complications Related to Specific Procedures**		
Application	**FDA and Literature Assessment**	
Posterolateral fusion	Potential for ↑ risk of malignancy at high doses	rhBMP-2 may be associated with ↑ risk of cancer, with double number of new cancer cases compared with iliac crest bone graft***
	Pain and impairment = harvesting ICBG	
	↑ early back and leg pain	↑ early postsurgical pain compared to iliac crest bone graft***
	Wound problems (2-5x ↑)	
ALIF	↑ osteolysis/subsidence/implant loosening	
	Retrograde ejaculation (6-9%)	
	Urinary retention	
	↑ delayed wound infections	
PLIF	Pain and impairment = harvesting ICBG	
	Ectopic/heterotopic bone formation (6x ↑)	
	Osteolysis/subsidence/implant migration (50-70%)	
	Radiculitis	
	↑ reoperation rate	
ACDF	↑ perioperative mortality	
	↑ perioperative life threatening event	
	↑ perioperative wound problems, difficulty swallowing	
	Prolonged dysphagia (2%)	
	Endplate resorption/subsidence/loss of alignment (> 50% of treated patients)	

*ALIF = anterior lumbar interbody fusion; PLIF = posterior lumbar interbody fusion; ACDF = anterior cervical discectomy and fusion; ICBG = iliac crest bone graft. *Mroz TE et al: Complications related to osteobiologics use in spine surgery: a systematic review. Spine (Phila Pa 1976). 35(9 Suppl):S86-104, 2010. **Carragee EJ et al: A critical review of recombinant human bone morphogenetic protein-2 trials in spinal surgery: emerging safety concerns and lessons learned. Spine J. 11(6):471-91, 2011.*** Simmonds M et al: Safety and effectiveness of recombinant human bone morphogenetic protein-2 for spinal fusion: A meta-analysis of individual-participant data. Ann Intern Med. 158:877-889, 2013*



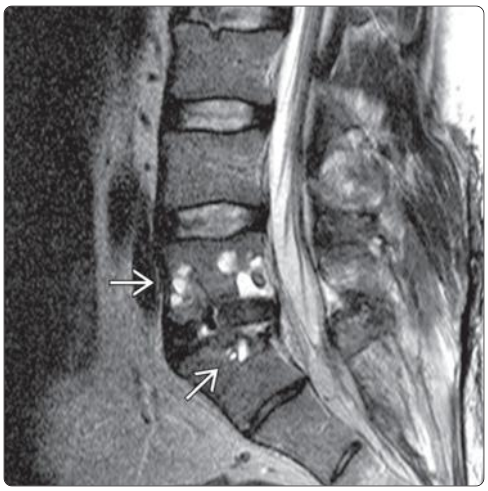


(Left) *Sagittal CT shows lucency surrounding the graft ⇨ in osteolysis related to bone rhBMP-2. This lucency might be concerning for infection or destruction from abnormal motion. However, there is no sclerosis or soft tissue mass to suggest infection, and the patient is asymptomatic.* **(Right)** *Coronal CT in the same patient shows well-defined lucency surrounding the graft ⇨. Lysis is at maximum in 1st 12 weeks after surgery when risk of graft migration/subsidence is greatest.*





(Left) *Axial NECT shows rhBMP-2 osteolysis in PLIF. Graft material is outlined by radiopaque markers ⇨.* **(Right)** *Axial NECT in the same patient just inferior to graft shows well-defined lucencies within the endplates and bodies at the L5-S1 fusion site ⇨. The edge of the graft is outlined by radiopaque marker ⇨.*





(Left) *rhBMP-2 osteolysis is seen on this T2WI MR as multiple small, cystic-appearing lesions with T2 hyperintensity involving the L5 and S1 bodies ⇨. Note the smooth margins, with abrupt transition to more normal marrow signal, and lack of adjacent marrow edema.* **(Right)** *Sagittal T1WI C+ MR in the same patient shows enhancement within several of the areas of lysis ⇨, confirming their solid nature despite the T2 cystic appearance.*

Heterotopic Bone Formation

KEY FACTS

TERMINOLOGY

- Definition: Bone development in an abnormal location
- Occurs in 3 main scenarios
  - rhBMP-2 related
  - TDR related
  - ACDF adjacent segment related

IMAGING

- Thin-section CT: High-attenuation bony osteophytes extending into canal or neural foramen with narrowing
- MR for definition of neural/thecal sac compression
- May be difficult to identify on T1W images if contains fatty marrow mimicking adjacent fat signal

TOP DIFFERENTIAL DIAGNOSES

- Graft migration or extrusion
- Adjacent level degenerative change
- Myositis ossificans

PATHOLOGY

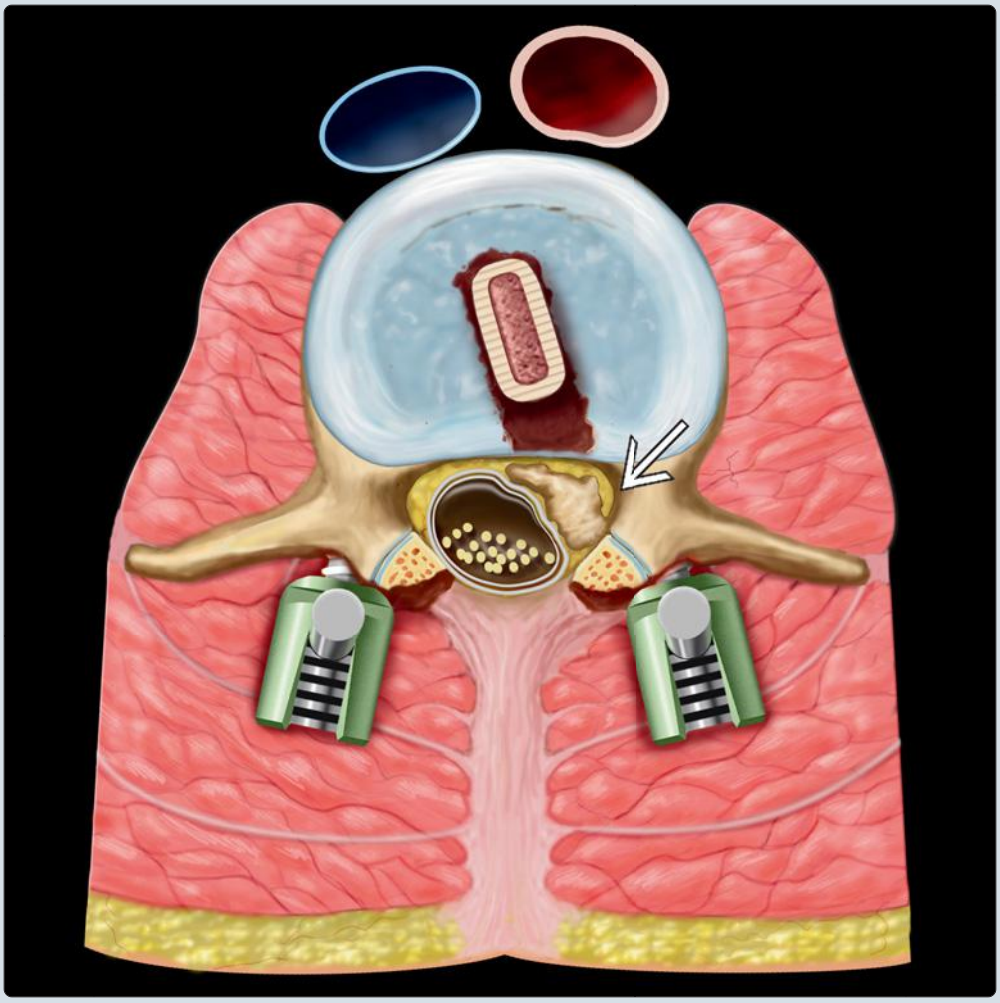
- **Osteobiologic related**: **Lumbar surgery**
  - Extradiscal/ectopic/heterotopic bone formation presumably occurs when BMP leaks from carrier into epidural space
  - 75% incidence in 1 series of single-level PLIF
- **Osteobiologic related**: **Cervical surgery**
  - 11% of rhBMP-2 group vs. 6% in control group

CLINICAL ISSUES

- Majority of HO appears asymptomatic but symptomatic foraminal stenosis reported sporadically
- HO can be the cause of new back pain and radiculopathy

DIAGNOSTIC CHECKLIST

- HO along PLIF or TLIF path may be difficult to identify on axial images since it may be mistaken for normal pedicle
- Look closely at sagittal CT to identify bone projecting from disc level



Axial graphic shows heterotopic bone formation in a patient who has undergone a posterior lumbar interbody fusion (PLIF). Bone formation is seen within the epidural space ➔ with mass effect upon the thecal sac. Heterotopic bone formation in PLIF surgery is highly correlated with the use of rhBMP-2 to enhance bony fusion.

TERMINOLOGY

Abbreviations

- Recombinant human bone morphogenetic protein 2 (rhBMP-2)
- Total disc replacement (TDR)
- Transforaminal lumbar interbody fusion (TLIF)
- Posterior lumbar interbody fusion (PLIF)

Synonyms

- Heterotopic ossification (HO)
- Ectopic bone formation
- Ectopic ossification
- Extradiscal ossification

Definitions

- Bone development in an abnormal location
- Occurs in 3 main scenarios
  - Osteobiologic (rhBMP-2) related
  - Total disc replacement (TDR) related
  - Anterior cervical discectomy and fusion (ACDF) adjacent segment related

IMAGING

General Features

- Best diagnostic clue
  - Thin-section CT: High-attenuation bony osteophytes or excrescences extending into canal or neural foramen with narrowing
- Location
  - Lumbar
  - Along TLIF/PLIF tract in ventrolateral epidural space
  - Arises from facet complex with variable amount of narrowing of neural foramen
- Size
  - Few mm to few centimeters
- Morphology
  - Variable from round and smooth margin to irregular osteophyte-like

CT Findings

- NECT
  - Bone density extending into canal or neural foramen in nonanatomic pattern
  - Must distinguish HO from extruded bone graft or osteophyte
  - May be difficult to identify on axial images since may merge with pedicle bone density
    - Look closely at sagittal images to see if abnormal bone is extending posteriorly from disc level

MR Findings

- T1WI
  - Look for narrowing of thecal sac and neural foramen
  - May be difficult to identify on T1W images if fatty marrow present within HO
- T2WI
  - Variable signal ranging from low to high depending upon composition of bone/fatty marrow

Imaging Recommendations

- Best imaging tool
  - CT for definition of extent of HO
  - MR for definition of neural/thecal sac compression

DIFFERENTIAL DIAGNOSIS

Graft Migration or Extrusion

- Look for radiopaque markers on ends for identification
- Lack of graft at fusion site

Adjacent Level Degenerative Change

- Osteophytes related to adjacent level degeneration

Calcified Disc Herniation

- Focal, rounded, &/or partially calcified lesion arising from disc space

Myositis Ossificans

- Related to prior muscle trauma
- Epicenter of calcification is within muscle
  - Not present in epidural space

Exostosis

- Cortical margin of lesion merges with cortex of posterior elements
- Cartilaginous cap (not always visible)

Fracture Fragment

- Related to intraoperative trauma
- Particularly during realignment of spinal column

Primary Bone-Producing Tumor

- Aggressive lesion with bone destruction and bone proliferation

PATHOLOGY

General Features

- Etiology
  - **Osteobiologic related**: **Lumbar surgery**
    - Extradiscal/ectopic/heterotopic bone formation presumably occurs when rhBMP-2 leaks from carrier into epidural space
      - Potential for central and foraminal stenosis
    - 75% incidence in 1 series of single-level PLIF
      - Cage placement within 2 mm of margin of posterior vertebral cortex associated with bone formation in canal in 77%
      - No clear relationship to symptomatology
    - 21% incidence in 1 series of PLIF and TLIF
      - Foraminal bone formation occurred only in TLIF group
      - No clear relationship to symptomatology
    - Lumbar spine patients who developed heterotopic bone received ALIF, PLIF, or TLIF
      - 4.0-12.0 mg BMP/level
    - Notably, literature shows **no** series of heterotopic bone in lumbar interbody fusions without use of rhBMP-2
  - **Osteobiologic related**: **Cervical surgery**
    - Bone formation anterior to segments adjacent to ACDF

- 11% of rhBMP-2 group vs. 6% in control group
- 2% rate in another study

– Cervical spine ACDF amounts vary from 0.6-7.0 mg BMP/level

- **Total disc replacement**
  - Cervical: Prodisc™ C (Synthes; West Chester, PA)
    - Significant HO in 45% of implants
    - Segmental ankylosis in additional 18% at 4 years
    - No clinical consequences
  - Cervical: Bryan® disc (Spine Experts; Los Angeles, CA)
    - 60% fusion rates 5 years after implantation
    - No new morbidity
    - Strong argument against theory of adjacent segment protection with TDR
  - Cervical: Mobi-C® (LDR; Troyes, France)
    - 8% HO rate

Staging, Grading, & Classification

- **HO grading**
  - Grade 0: No new HO formation after prosthesis implantation
  - Grade I: Segments with new HO formation not reaching intervertebral space
  - Grade II: HO reaches intervertebral space, but segmental movement is not limited
  - Grade III: Bridging ossification with limited, but possible, movement
  - Grade IV: Segmental fusion

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Majority of HO appears asymptomatic
  - Several series where symptomatic bone production does occur
    - Wong et al reported 5 patients with ectopic bone after PLIF/TLIF
      - Had neurologic complaints and 3/5 underwent revision surgery
    - Chen et al reported 4 patients with symptomatic HO following lumbar TLIF
      - Onset of new symptoms 23-51 months following surgery
      - Back pain and radiculopathy

Treatment

- Varies from symptomatic to reoperation with resection of heterotopic bone

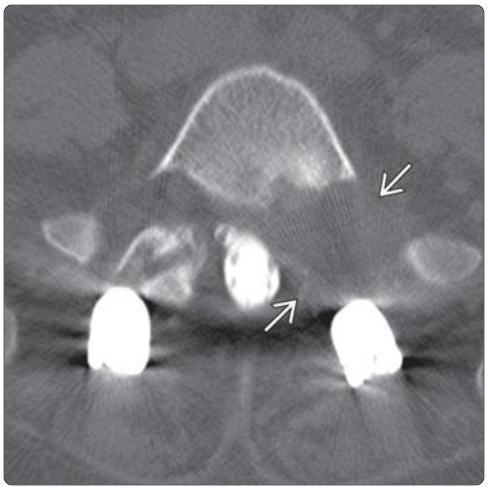
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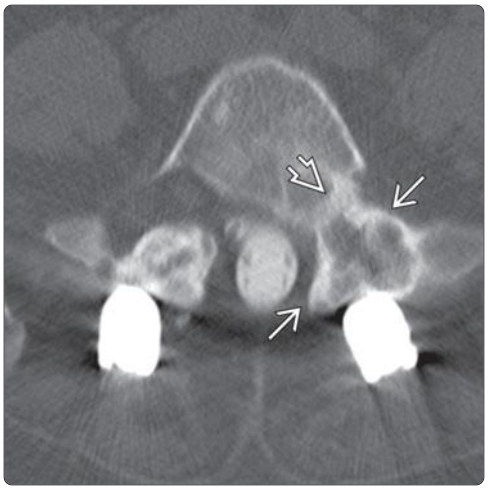
Consider

- HO containing fatty marrow easy to miss on T1W images since epidural and foraminal fat can have equivalent signal
- HO along PLIF or TLIF path may be difficult to identify on axial images since it may be mistaken for normal pedicle
  - Look closely at sagittal CT images to see abnormal bone projecting posteriorly from intervertebral disc level

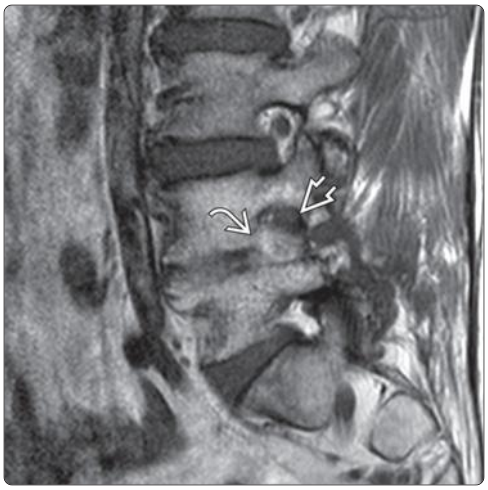
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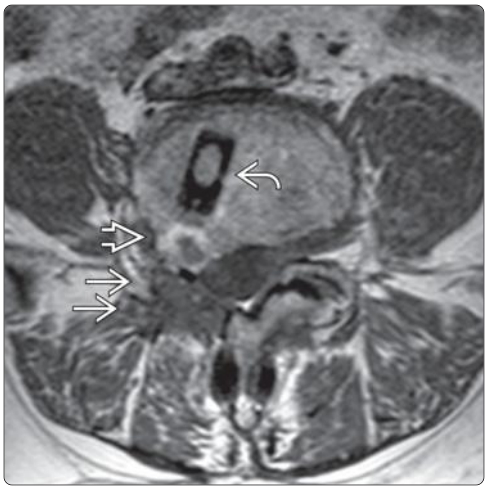
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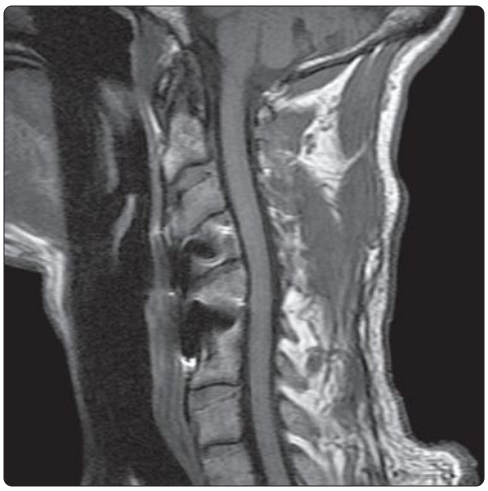


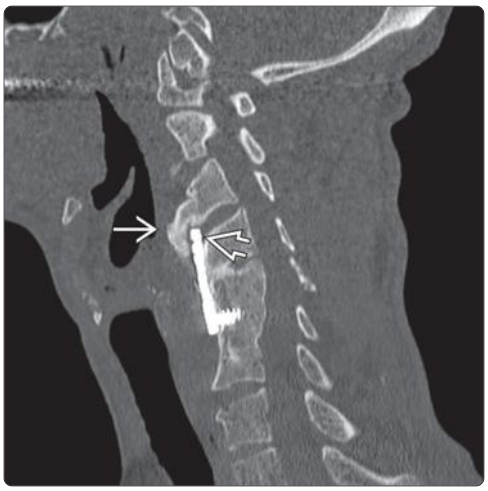
(Left) *Heterotopic bone formation mimics the appearance of facet joint in this patient status post facetectomy and posterior fusion. Initial CT myelogram study shows a large facetectomy defect on the left at L5* ⇨. **(Right)** *CT myelogram in the same patient, performed 2 years later, shows new heterotopic bone filling in the operative defect* ⇨*, which has the outline of the prior facet joint margin. There is left foraminal stenosis* ⇨ *from the heterotopic bone.*





(Left) *Severe foraminal stenosis from heterotopic bone formation is shown. Sagittal T1WI MR through the foramen demonstrates marked bony foraminal narrowing at L4-L5 on the right* ⇨ *with compression of the exiting right L4 root* ⇨*.* **(Right)** *Axial T1WI MR (same patient) shows the trajectory of prior facetectomy and posterior interbody fusion* ⇨*, graft site* ⇨*, and heterotopic ossification with foraminal stenosis* ⇨*. The ossification shows increased T1 signal due to fatty marrow within the bone.*





(Left) *Heterotopic ossification located superior to fusion site effectively extends the fusion cephalad 1 level. Sagittal T1 MR shows prior hardware from anterior fusion spanning C4-C6. There is loss of cervical lordosis; otherwise the fusion sites appear unremarkable. Metal artifact often obscures this abnormality on MR.* **(Right)** *Sagittal NECT (same patient) shows heterotopic ossification extending inferiorly from the C3 body* ⇨ *to cover superior aspect of the C4 plate. This superior margin of the plate is malpositioned & extends to the disc level* ⇨*.*

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PART VII
SECTION 3

Post Radiation and Chemotherapy Complications



Radiation Myelopathy	**1154**
Post-Irradiation Vertebral Marrow	**1158**
Anterior Lumbar Radiculopathy	**1162**

Radiation Myelopathy

KEY FACTS

TERMINOLOGY

- Chronic progressive radiation myelitis (CPRM)
- Delayed radiation myelopathy (DRM)

IMAGING

- Spindle-shaped cord swelling with irregular, focal rind of enhancement (early)
- Focal cord atrophy (late)
- Clinical signs may reflect longer segment of damage than demonstrated on MR

TOP DIFFERENTIAL DIAGNOSES

- Transverse myelitis
- Multiple sclerosis
- Spinal cord infarct
- Astrocytoma
- Syrinx

PATHOLOGY

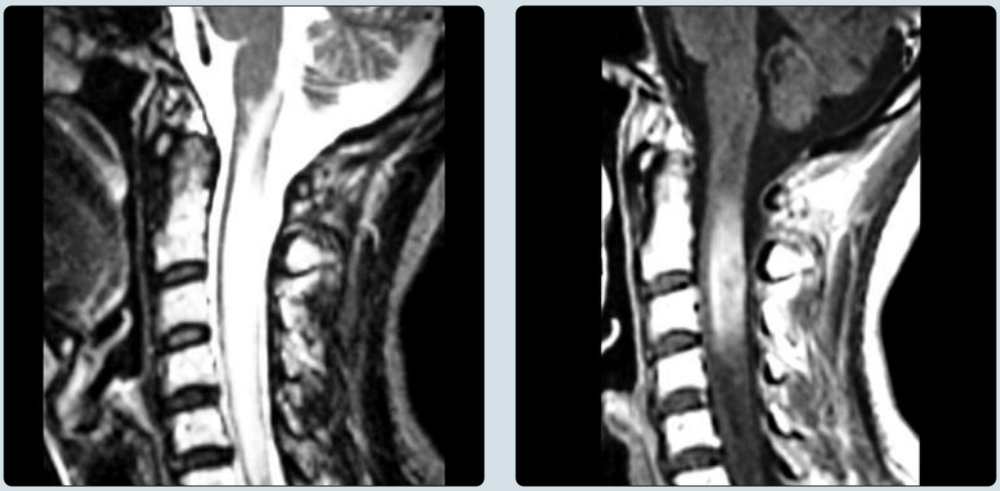
- Demyelination, lipid-laden microphages, swollen astrocytes, endothelial damage, necrosis, local Ca++ deposition, hyalinosis of intramedullary vessel walls

CLINICAL ISSUES

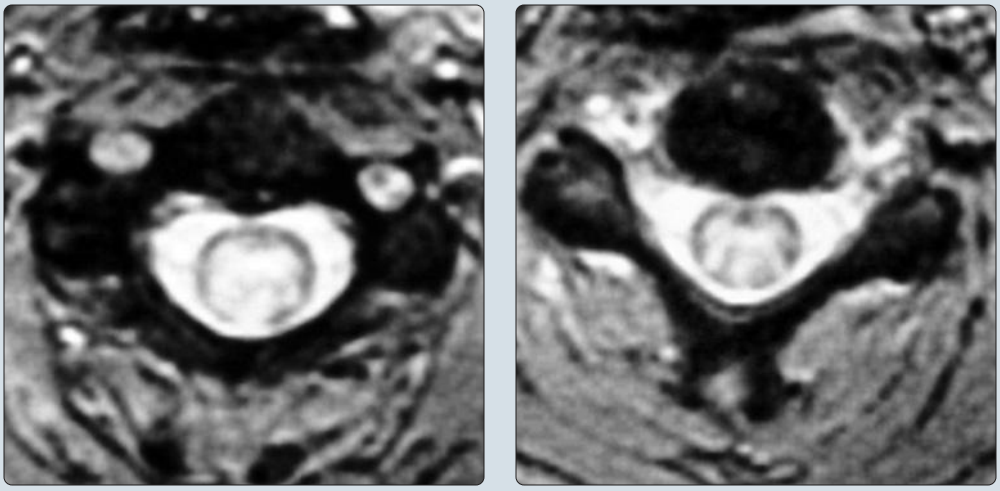
- Onset of progressive numbness and weakness ± sphincter dysfunction 1 month → several years after fractionated radiotherapy
- Relentless progression without significant improvement in most cases

DIAGNOSTIC CHECKLIST

- Late focal atrophy at sites of previous enhancement
- Fatty marrow replacement in treatment field provides clue to etiology
- Pathologic cord changes not always visible on MR imaging



(Left) *Sagittal T2WI MR in a patient following radiation therapy for head and neck squamous cell carcinoma demonstrates abnormal intramedullary T2 hyperintensity in conjunction with fatty marrow replacement, characteristic of postradiation vertebral and spinal cord changes.* **(Right)** *Sagittal T1WI C+ MR confirms abnormal spinal cord parenchymal enhancement in the area of greatest radiation myelopathic injury. Note also the marked T1 shortening in the vertebral marrow reflecting fatty replacement.*



(Left) *Axial T2* GRE MR obtained within the area of greatest cord expansion and cord injury confirms that the majority of abnormal signal resides within spinal cord central gray matter.* **(Right)** *Axial T2* GRE MR obtained at a different level shows less cord expansion probably reflecting mostly regional edema within the gray matter.*

TERMINOLOGY

Abbreviations

- Chronic progressive radiation myelitis (CPRM)
- Delayed radiation myelopathy (DRM)
- Radiation necrosis (RN)

Definitions

- Damage of neural tissue in spinal cord following therapeutic radiation of intrinsic or adjacent neoplastic disease

IMAGING

General Features

- Best diagnostic clue
  - Cord swelling + intramedullary enhancement
- Location
  - Mainly in white matter of lateral spinothalamic tracts, dorsal columns
  - Occurs within radiation field
    - Radiation port often demarcated by geographic fatty marrow changes
- Size
  - Dependent on dimensions of radiation field
  - Clinical signs may reflect longer segment of damage than demonstrated on MR imaging
- Morphology
  - Spindle-shaped cord swelling with irregular, focal rind of enhancement (early)
  - Focal cord atrophy (late)

Radiographic Findings

- Radiography
  - Noncontributory

CT Findings

- NECT
  - ± spinal cord swelling
  - Insensitive for demonstration of radiation myelopathy
- CECT
  - ± spinal cord swelling
  - ± cord enhancement

MR Findings

- T1WI
  - Expanded spinal cord, ↓ intramedullary signal intensity
  - Focal cord atrophy in late stage
- T2WI
  - Fusiform cord expansion, ↑ intramedullary signal intensity
  - Focal cord atrophy, ↑ intramedullary signal in late stage
- STIR
  - Similar to T2WI
  - Suppressed signal intensity within postradiation fatty replaced vertebral marrow
- DWI
  - Diffusion restriction indicates ischemic cord injury
- T1WI C+
  - ± ring or irregular enhancement surrounded by edematous cord
  - Enhancing focus becomes site of focal atrophy in subsequent serial scans

Nonvascular Interventions

- Myelography
  - Indirect demonstration of spinal cord injury
    - Swollen spinal cord
    - Cord atrophy in late stage

Imaging Recommendations

- Best imaging tool
  - Multiplanar MR imaging, including T1 C+ MR
- Protocol advice
  - Fat-saturated T1 C+ increases conspicuity of cord lesions

DIFFERENTIAL DIAGNOSIS

Transverse Myelitis

- More rapid onset of myelopathy
- Clinically relevant predisposing factors of recent viral infection or vaccination

Multiple Sclerosis

- Multifocal lesions ± enhancement
- Patchy high signal foci on T2WI
- Sensory symptoms at onset
- Waxing, waning signs and symptoms

Spinal Cord Infarct

- Acute onset of paralysis
- Gray matter involvement predominates

Astrocytoma

- Very slow onset and progression

Syrinx

- More insidious onset
- Discrete cavity, frequently eccentric

PATHOLOGY

General Features

- Etiology
  - Radiation damages spinal cord parenchyma
  - Typically occurs with doses > 50 gray (Gy)
    - However, may see in cases < 50 Gy
  - Fractionation schedule > 2 Gy per treatment
  - Concurrent chemotherapy may be predisposing factor
    - Especially intrathecal administration route
- Associated abnormalities
  - Fatty vertebral marrow replacement within radiation port

Gross Pathologic & Surgical Features

- Swollen, edematous, enlarged spinal cord

Microscopic Features

- Demyelination worst in lateral, dorsal tracts
- Lipid-laden microphages
- Swollen astrocytes
- Endothelial damage
- Hyalinosis of intramedullary vessel walls
- Necrosis
- Local calcium deposits

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Progressive numbness and weakness
- Other signs/symptoms
  - Sphincter dysfunction
- Clinical profile
  - Onset of weakness, numbness after 1 month → several years following fractionated radiotherapy
  - Radiotherapy for lung or nasopharyngeal cancer; mantle lymphoma therapy (especially with intrathecal methotrexate) higher risk

Demographics

- Age
  - Any age of onset
- Gender
  - No predilection
- Epidemiology
  - Unusual complication

Natural History & Prognosis

- Progressive without significant improvement in most cases

Treatment

- Hyperbaric oxygen helpful in some early cases

DIAGNOSTIC CHECKLIST

Consider

- Carefully consider radiation field, fractionation schedule (especially if > 2 Gy per treatment)

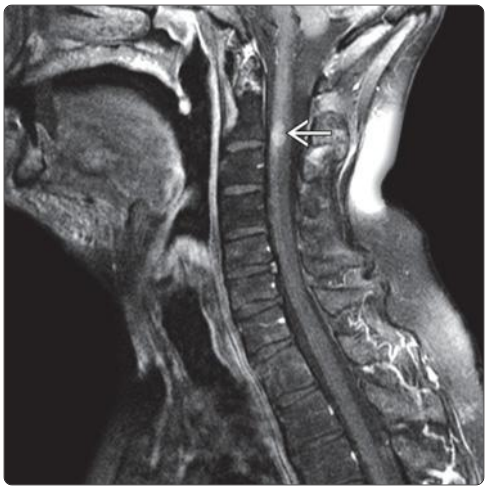
Image Interpretation Pearls

- Late stage shows focal atrophy at sites of previous contrast enhancement
- Fatty replacement of marrow in vertebral bodies within treatment field provides clue to etiology
- Pathologic changes in cord not always visible on MR imaging

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(Left) *Sagittal T2WI FS MR following radiation treatment of oral carcinoma shows abnormal fusiform enlargement and hyperintensity of cervical cord at the C2-C3 level. Note also diffuse suppression of postradiation fatty marrow changes producing abnormally dark marrow.* **(Right)** *Sagittal T1WI C+ FS MR shows focal cervical spinal cord enhancement at C2 following radiation therapy for oral carcinoma. Also, note signal suppression of diffuse postradiation fatty marrow changes.*





(Left) *Sagittal T2WI MR shows diffusely enlarged hyperintense cervical spinal cord in a patient treated previously with radiation therapy for nasopharyngeal cancer.* **(Right)** *Sagittal T1WI C+ MR following radiation therapy for nasopharyngeal carcinoma shows patchy enhancement within a long segment of abnormally enlarged cord. Also, note abnormal hypointense intramedullary edematous low signal intensity rostral and caudal to the enhancing cord lesions.*





(Left) *Sagittal T1WI MR shows extensive spinal cord radiation myelopathy changes with marked diffuse fusiform expansion of the lower cervical and thoracic cord. The radiation therapy port is demarcated by fatty marrow replacement of C6, C7, and upper thoracic vertebral bodies.* **(Right)** *Sagittal T2WI MR following radiation treatment for melanoma shows extensive radiation myelopathy involving the cervical and thoracic cord with diffuse cord hyperintensity.*

Post-Irradiation Vertebral Marrow

KEY FACTS

TERMINOLOGY

- Transformation of cellular vertebral marrow into fatty marrow following therapeutic irradiation

IMAGING

- Sharp demarcation between irradiated and untreated marrow (margins of radiation port)
  - Corresponds to site of irradiation and extent of radiation field
- Marrow signal intensity (SI) within radiotherapy portal similar to subcutaneous fat on T1WI
- May demonstrate associated compression fractures

TOP DIFFERENTIAL DIAGNOSES

- Normal fatty marrow
- Vertebral hemangioma

PATHOLOGY

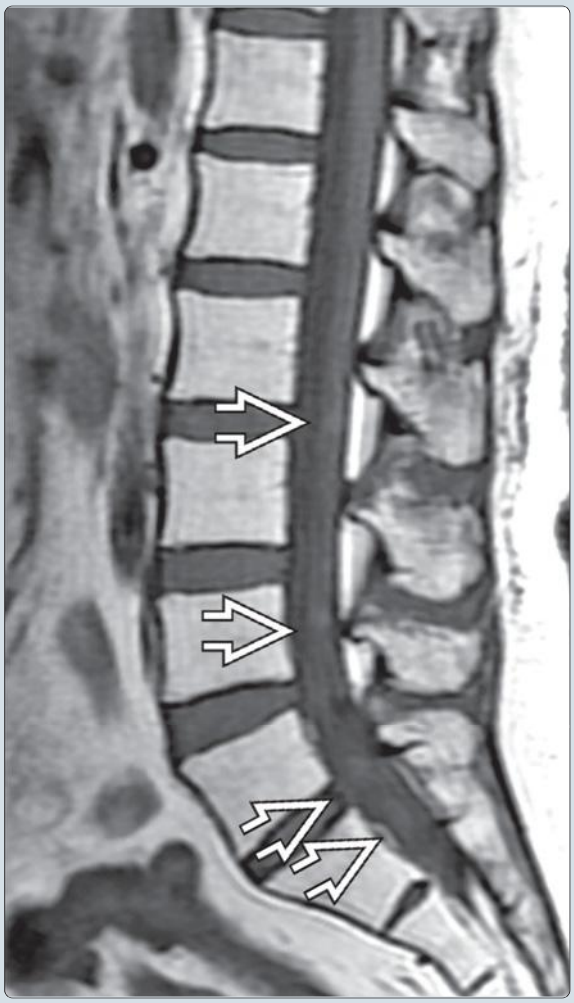
- Marrow changes on MR dependent on radiation dose, fractionation, and time elapsed after treatment
- Complications include osteopenia, osteoporotic compression fracture, osteonecrosis, residual/recurrent metastases, and post-radiation myelitis

CLINICAL ISSUES

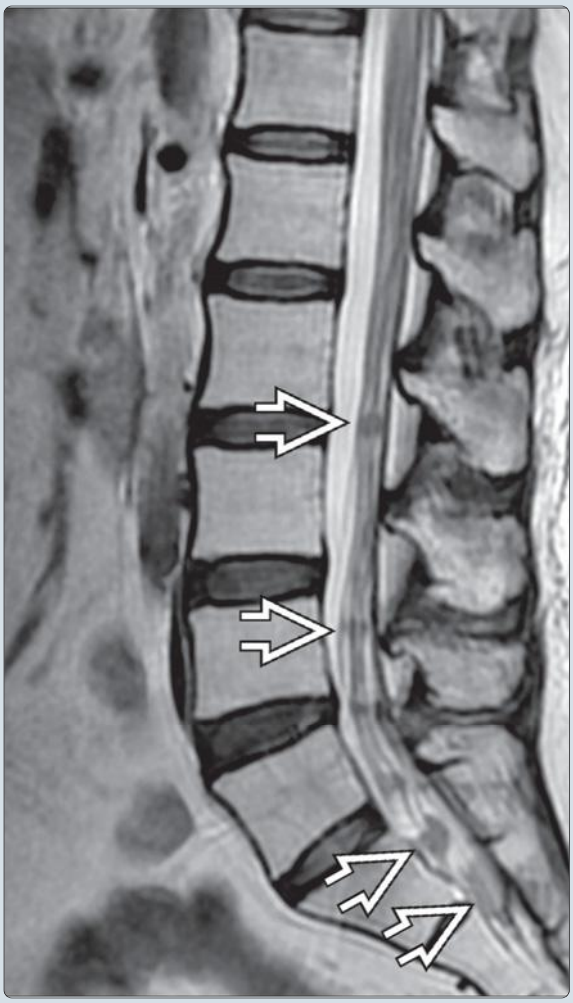
- Usually asymptomatic
- Development or worsening of pain and neurologic complaints worrisome for radiation necrosis, insufficiency fracture, or tumor recurrence or progression

DIAGNOSTIC CHECKLIST

- Homogeneous fatty marrow sharply delineated from untreated vertebral marrow diagnostic of post-irradiation change
- MR is modality of choice for marrow imaging
  - T2WI FS MR or STIR and T1WI C+ FS MR to evaluate for residual or recurrent marrow disease



Sagittal T1WI (intracranial glioblastoma) shows typical post-radiation fatty marrow conversion following whole-spine radiation. Multiple intradural metastatic nodules ⇨ involving the cauda equina are subtle.



Sagittal T2WI (whole spine radiation therapy for glioblastoma intradural metastases) demonstrates diffuse vertebral fatty marrow replacement and multiple metastatic nodules within the cauda equina ⇨.

TERMINOLOGY

Definitions

- Transformation of cellular vertebral marrow into fatty marrow following therapeutic irradiation

IMAGING

General Features

- Best diagnostic clue
  - Marrow signal intensity (SI) within radiotherapy portal similar to subcutaneous fat on T1WI
- Location
  - Corresponds to site of irradiation
- Size
  - Corresponds to extent of radiation field
- Morphology
  - Homogeneous hyperintense marrow SI
    - Heterogeneity suspicious for residual or recurrent marrow disease
  - Sharp demarcation between irradiated and untreated marrow (margins of radiation port)

Radiographic Findings

- Radiography
  - ± osteopenia, compression fractures

CT Findings

- Bone CT
  - ± compression fractures
  - Not particularly useful for detecting fatty marrow content

MR Findings

- T1WI
  - Very high marrow signal intensity (follows fat)
- T2WI
  - Intermediate SI, follows fat
- PD/intermediate
  - Intermediate SI, follows fat
- STIR
  - High signal in acute phase (edema)
  - Low signal thereafter (fatty replacement)
- T1WI C+
  - Post-gadolinium enhancement in acute phase
    - Probably 2° to vascular congestion
  - No enhancement thereafter
    - Decreased vascularity, fibrosis
- 1st 3 weeks
  - No change or early marrow T1 hyperintensity
    - Hyperintensity suggests hemorrhage ± fatty replacement
  - Hyperintensity on STIR
    - Marrow edema and necrosis
    - Peak incidence 9 days after irradiation, subsequent gradual SI decrease
- 3-6 weeks
  - Heterogeneous mottled pattern on T1WI
  - Increasing fat intensity within central vertebral marrow surrounding basivertebral vein
- After 6 weeks
  - Diffuse and homogeneous hyperintensity on T1WI
  - Band pattern of peripheral intermediate SI
    - Surrounding central hyperintense marrow
    - May represent regenerating hematopoietic marrow
  - > 90% of treated marrow has this appearance

Nuclear Medicine Findings

- Bone scan
  - Sharply delineated focal or diffuse diminished radiotracer uptake representing loss of vascularity
- PET
  - Decreased marrow metabolic activity with FDG-18
- Tc-99m sulfur colloid scan
  - Sharply demarcated focal or diffuse photopenic region corresponds to radiation therapy portal

Other Modality Findings

- Dual energy CT study
  - Bone mass may be reduced

Imaging Recommendations

- Best imaging tool
  - MR is modality of choice for marrow imaging
- Protocol advice
  - T1WI is most sensitive for detecting fatty replacement
  - STIR or FSE T2WI with fat saturation best assesses for recurrent or residual marrow disease
  - Pre- and post-gadolinium T1WI with fat suppression increases lesion conspicuity

DIFFERENTIAL DIAGNOSIS

Normal Fatty Marrow

- Age related
- Predominantly fat intensity, often some heterogeneity present
- No clear demarcation
- ± compression fractures
- No abnormal uptake on bone scan or PET

Vertebral Hemangioma

- Well-circumscribed, rounded lesion
- Typically hyperintense on T1WI and T2WI 2° to presence of fatty stroma
- Avid contrast enhancement

PATHOLOGY

General Features

- Etiology
  - Radiation destroys marrow hematopoietic elements (highly radiosensitive)
  - Concurrent reduction of bone mass
  - Marrow changes on MR dependent on radiation dose, fractionation, and time elapsed after treatment
    - Fatty marrow seen on MR as early as 2 weeks after therapeutic irradiation
      - > 90% will show fatty marrow after 2 months
    - No change in marrow SI with radiation dose of 1.25 Gy
    - At 50 Gy, persistent fat SI even after 9 years
      - Complete and irreversible eradication of cellular elements

- Between 20-30 Gy, return to normal marrow pattern after long-term follow-up: > 10 years
  - Partial recovery between 2-9 years
- Marrow reconversion in pediatric patients occurs 11-30 months after radiation treatment

- Genetics
  - No genetic predisposition
- Associated abnormalities
  - Osteopenia
  - Osteoporotic compression fractures
  - Osteonecrosis
    - Sharp border between normal and infarcted marrow
    - MR
      - Hyperintense on STIR or T2WI FS MR
      - Hypointense on T1WI
      - ± intravertebral fluid collection
      - ± enhancement after intravenous gadolinium contrast
    - Radiography, CT
      - ± linear transverse intravertebral gas, sclerosis
    - Bone scan
      - No uptake unless vertebral collapse
      - May be difficult to distinguish from recurrent metastasis
      - Biopsy indicated
  - Residual or recurrent metastases
    - Multifocal discrete lesions
    - Paradural and paravertebral soft tissue extension
    - Pathologic fractures
    - Hypointense on T1WI surrounded by fatty marrow
    - Hyperintense on STIR or T2WI FS MR
    - Treated lesions may not enhance
  - Post-radiation myelitis
    - Diffuse spinal cord T2 hyperintensity within radiation treatment field
    - Mildly hypointense on T1WI
    - Cord enlargement ± irregular foci of enhancement (necrosis)

Microscopic Features

- Based on rat model after single dose of irradiation at 20 Gy
- Initially decreased cellular elements and disrupted sinusoids
  - Associated edema, hemorrhage
- Early influx of cells to repopulate marrow
  - Concomitant increase in fatty marrow
- Subsequent depletion of cellularity and sinusoids
  - Fibrosis with further increase in fatty marrow
- Eventual regeneration of hematopoietic elements and sinusoids

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
- Other signs/symptoms
  - Pain (fracture)
  - Myelopathy or radiculopathy from compression fracture or tumor infiltration
- Clinical profile
  - Development or worsening of pain and neurologic complaints worrisome for radiation necrosis, insufficiency fracture, or tumor recurrence or progression

Demographics

- Age
  - Children and adults
- Gender
  - M = F
- Ethnicity
  - No racial predilection
- Epidemiology
  - Expected finding on all post-radiotherapy patients

Natural History & Prognosis

- Return to normal marrow pattern on long-term follow-up: > 10 years
- Dependent on underlying disease for which radiation therapy is employed

Treatment

- Expected finding following radiation therapy; no treatment necessary in absence of complications
  - Supportive measures for compression fracture
  - Surgical intervention for cord compression or spinal instability

DIAGNOSTIC CHECKLIST

Consider

- T2WI FS MR or STIR and T1WI post-gadolinium with fat suppression to evaluate for residual or recurrent marrow disease

Image Interpretation Pearls

- Homogeneous fatty marrow sharply delineated from untreated vertebral marrow diagnostic of post-irradiation change

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(Left) *Sagittal T1WI MR of the cervicothoracic spine demonstrates smooth marrow hyperintensity in the thoracic vertebrae with distinct signal intensity transition at T1-T2, the upper boundary of the radiotherapy field.* **(Right)** *Sagittal T1WI MR reveals homogeneous post-irradiation fatty marrow in the thoracic spine, with clear cranial and rostral transitional demarcation, corresponding to the radiotherapy portal margins.*





(Left) *Sagittal T1WI MR depicts fatty replacement of inferior T5 and entire T6-T8 vertebral bodies and posterior elements, as compared to the T9 vertebral body, which is nearly completely replaced by a metastasis.* **(Right)** *Sagittal T2WI MR confirms hyperintense fatty replacement of inferior T5 and entire T6-T8 vertebral bodies and posterior elements. T9, in contrast, shows a relatively hypointense metastasis replacing the marrow.*





(Left) *Sagittal STIR MR demonstrates that both normal marrow and irradiated marrow display nearly the same low signal intensity after fat suppression, but the metastasis maintains hyperintense signal intensity.* **(Right)** *Sagittal T1WI C+ FS MR confirms signal suppression and lack of enhancement in fatty replaced marrow of inferior T5 and T6-T8 vertebral bodies and posterior elements. The T9 vertebral body is nearly completely replaced by enhancing metastasis.*

KEY FACTS

TERMINOLOGY

- Transient or permanent paraplegia uncommon complication following intrathecal chemotherapy
- Initially described following intrathecal methotrexate (MTX) in pediatric acute lymphoblastic leukemia patients

IMAGING

- Smooth linear enhancement of conus pia, anterior cauda equina nerve roots
- Unenhanced T1W, T2W images normal

TOP DIFFERENTIAL DIAGNOSES

- Guillain-Barré syndrome
- CSF disseminated metastases
- Postradiation radiculopathy

PATHOLOGY

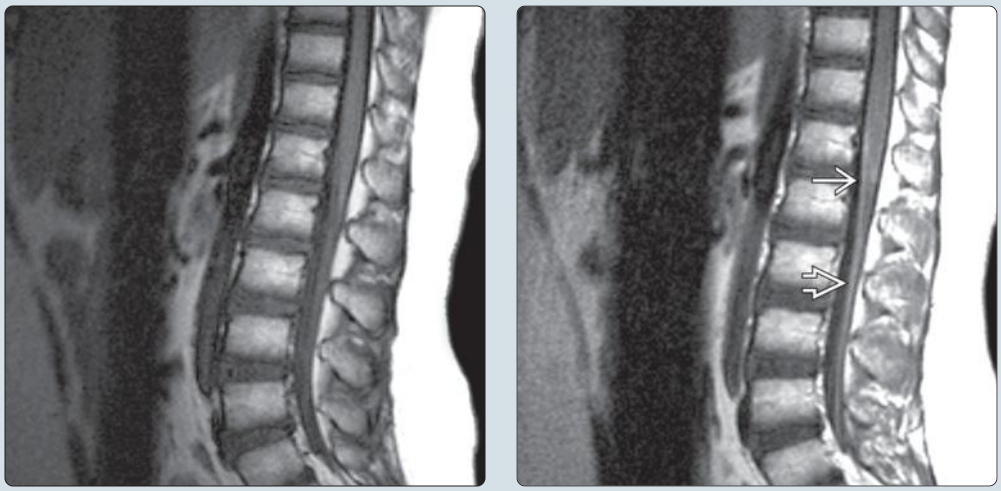
- Intrathecal MTX, arabinoside (Ara-C) standard prophylaxis and treatment agents for pediatric CNS leukemia
- Mechanism of neural toxicity not well understood

CLINICAL ISSUES

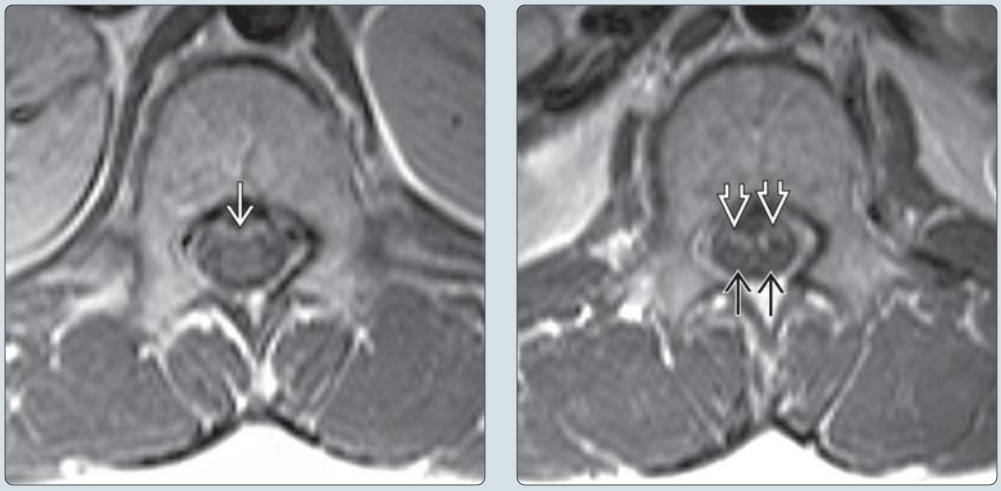
- Lower extremity flaccid weakness, most often without sensory deficit
- ↑ CSF protein, mononuclear pleocytosis, CSF immunoglobulin G synthesis
- Most commonly occurs after > 1 intrathecal chemotherapy dose
- Abnormal electrodiagnostic studies correlate with nerve root enhancement
- Most patients are children, but rare reports in adults
- Extent of clinical recovery is variable

DIAGNOSTIC CHECKLIST

- Correlation with clinical information critical to establishing correct diagnosis
- Contrast administration essential to detect cauda equina abnormality



(Left) *Sagittal T1WI MR in a leukemia patient with leg weakness following intrathecal chemotherapy shows normal appearance of the conus and cauda equina.* **(Right)** *Sagittal T1WI C+ MR in a leukemia patient with leg weakness following intrathecal chemotherapy reveals abnormal smooth, thin, linear enhancement of the ventral conus pia and cauda equina.*



(Left) *Axial T1WI C+ MR through the conus medullaris confirms smooth, thin, linear enhancement of the ventral conus pia and cauda equina.* **(Right)** *At the level of the cauda equina, axial T1WI C+ MR demonstrates characteristic smooth thin linear enhancement of the ventral cauda equina nerve roots but not the dorsal cauda equina nerve roots.*

TERMINOLOGY

Abbreviations

- Methotrexate (MTX), cytosine arabinoside (Ara-C)

Definitions

- Transient or permanent paraplegia
- Uncommon complication following intrathecal chemotherapy

IMAGING

General Features

- Best diagnostic clue
  - Smooth linear enhancement of conus pia and ventral cauda equina following intrathecal methotrexate chemotherapy
- Location
  - Ventral cauda equina and conus pia

MR Findings

- T1WI
  - Nearly always normal; ± mild diffuse cauda equina nerve root thickening
- T2WI
  - Nearly always normal; ± mild diffuse cauda equina nerve root thickening
- T1WI C+
  - Anterior lumbosacral nerve root, conus pial enhancement

Imaging Recommendations

- Best imaging tool
  - Multiplanar contrast-enhanced lumbar MR
- Protocol advice
  - Sagittal and axial T1WI C+ MR key sequences

DIFFERENTIAL DIAGNOSIS

Guillain-Barré Syndrome

- Similar imaging appearance to anterior lumbar radiculopathy
- Clinical history suggests correct diagnosis

CSF-Disseminated Metastases

- Leukemia/lymphoma, primary brain neoplasms most common source
- Nodular and irregular nerve root thickening and enhancement

Post Radiation Radiculopathy

- Smooth enhancement of cauda equina; usually involves both ventral and dorsal roots
- Clinical history of prior radiation therapy

PATHOLOGY

General Features

- Etiology
  - Intrathecal MTX or Ara-C are standard prophylaxis and treatment approach to pediatric CNS leukemia
    - Concomitant use of intravenous or intrathecal Ara-C with intrathecal MTX may increase neurotoxicity
    - Most symptomatic patients have had > 1 intrathecal injection
  - Mechanism of neural toxicity not well understood
  - Proposed theories include
    - Direct MTX or indirect preservative-based neurotoxicity
    - Selective ventral nerve root vulnerability to intrathecal chemotherapy toxicity
    - Gravity-dependent ↑ drug concentration around ventral nerve roots
    - Poorly understood selective autoimmune process
    - Local folate depletion 2° to MTX therapy may ↑ toxicity risk

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Lower extremity flaccid weakness, most often without sensory deficit
  - Most commonly after > 1 intrathecal chemotherapy dose
- Other signs/symptoms
  - ↑ CSF protein, mononuclear pleocytosis, CSF immunoglobulin G synthesis
  - Electrodiagnostic studies (nerve conduction, EMG) → altered sensory and motor conduction in lower extremities
    - Usually correlates with nerve root enhancement

Demographics

- Age
  - Most patients are children, but rare reports in adults

Natural History & Prognosis

- Extent of clinical recovery is variable

Treatment

- Intravenous immunoglobulin
- Concurrent use of hydrocortisone to diminish MTX CNS toxic effects of unconfirmed utility

DIAGNOSTIC CHECKLIST

Consider

- Correlation with clinical information critical

Image Interpretation Pearls

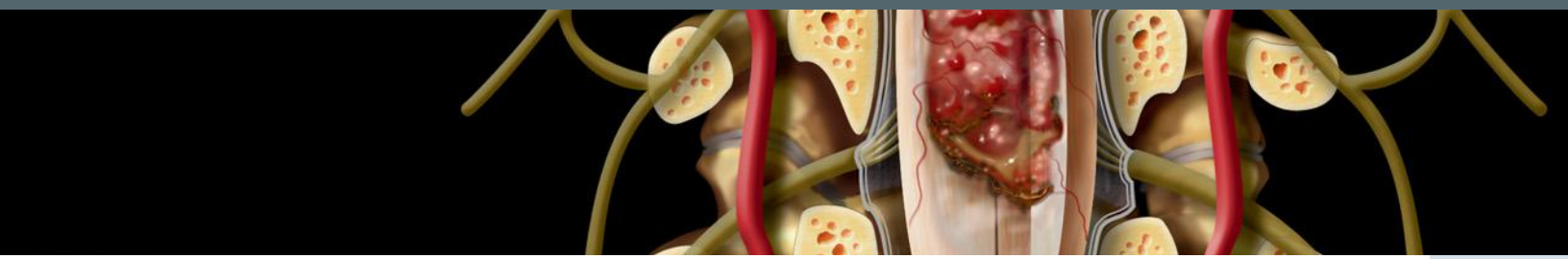
- Contrast administration essential to make imaging diagnosis

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