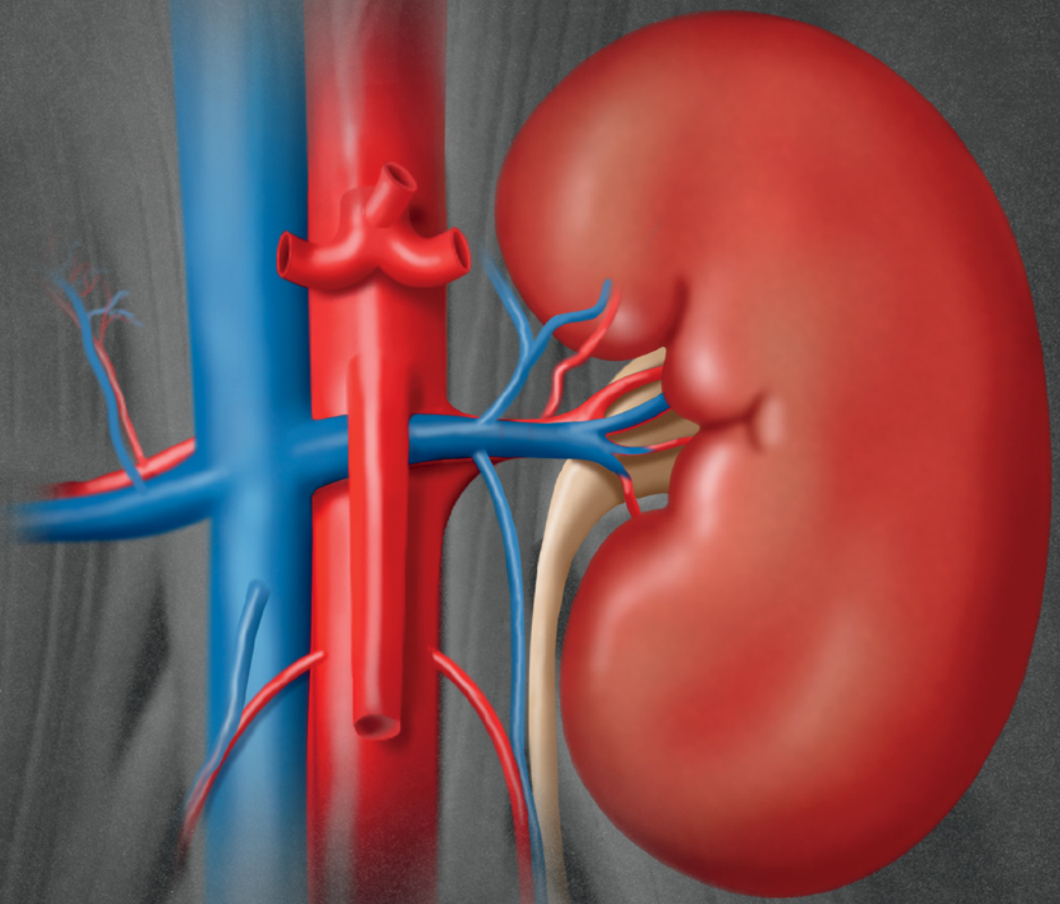
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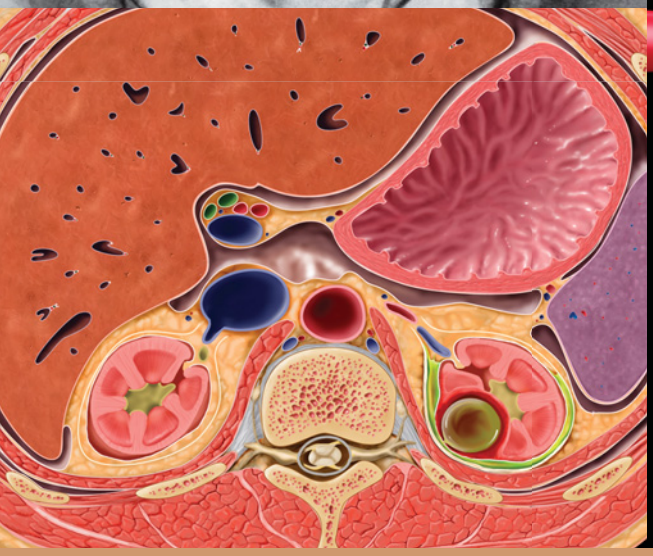
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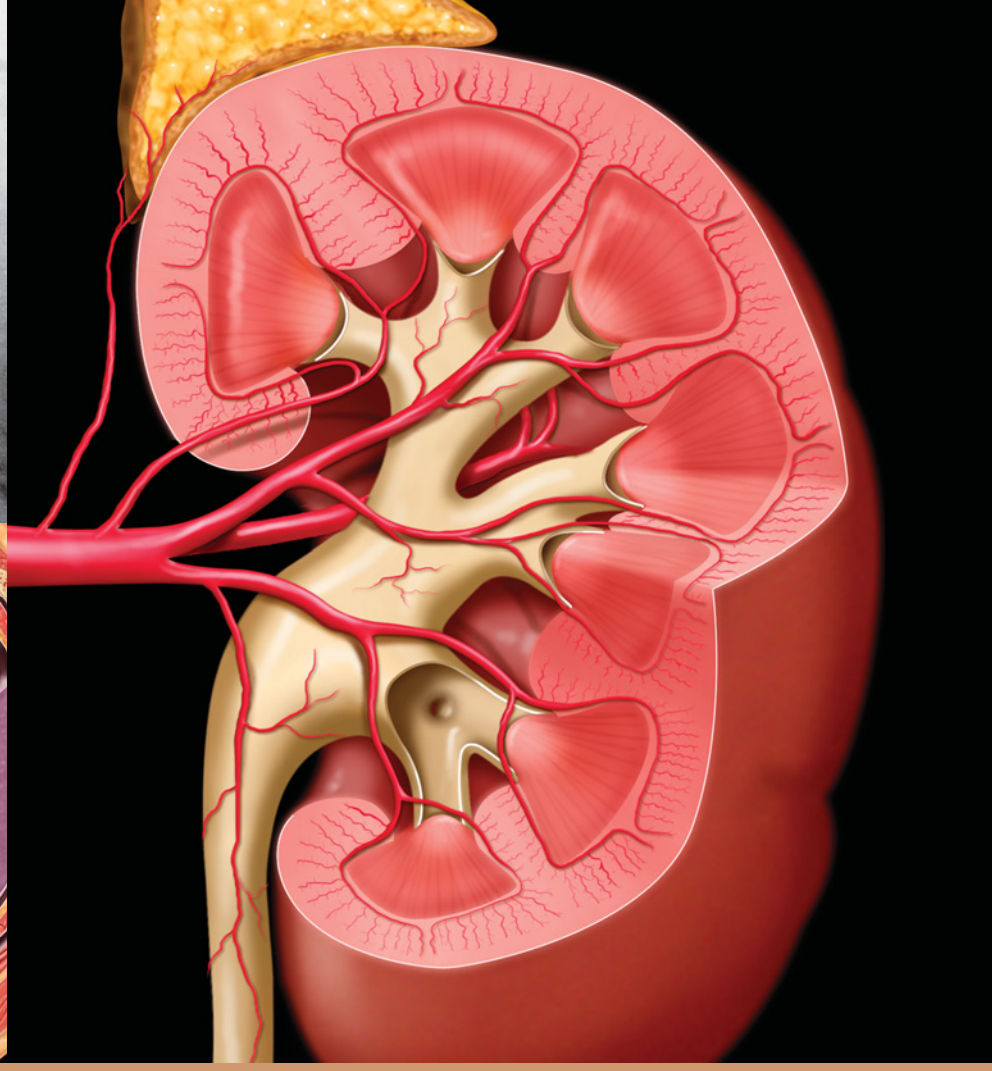
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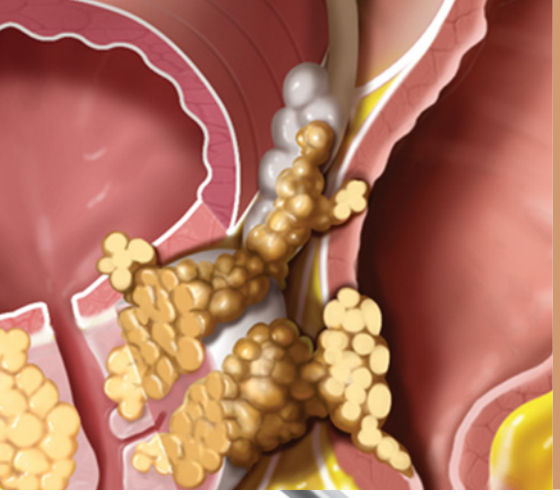




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Dedications

To the home team: My wonderful wife, Mary, and our sons, Daniel, Josh, and Andrew. The Tublin mantra: Good things happen with drive, commitment, and integrity...but it's really your love and support that make everything possible.

MT

I would like to dedicate this book to my wonderful wife, Goli, and to my beloved parents who have supported me in every second of my life.

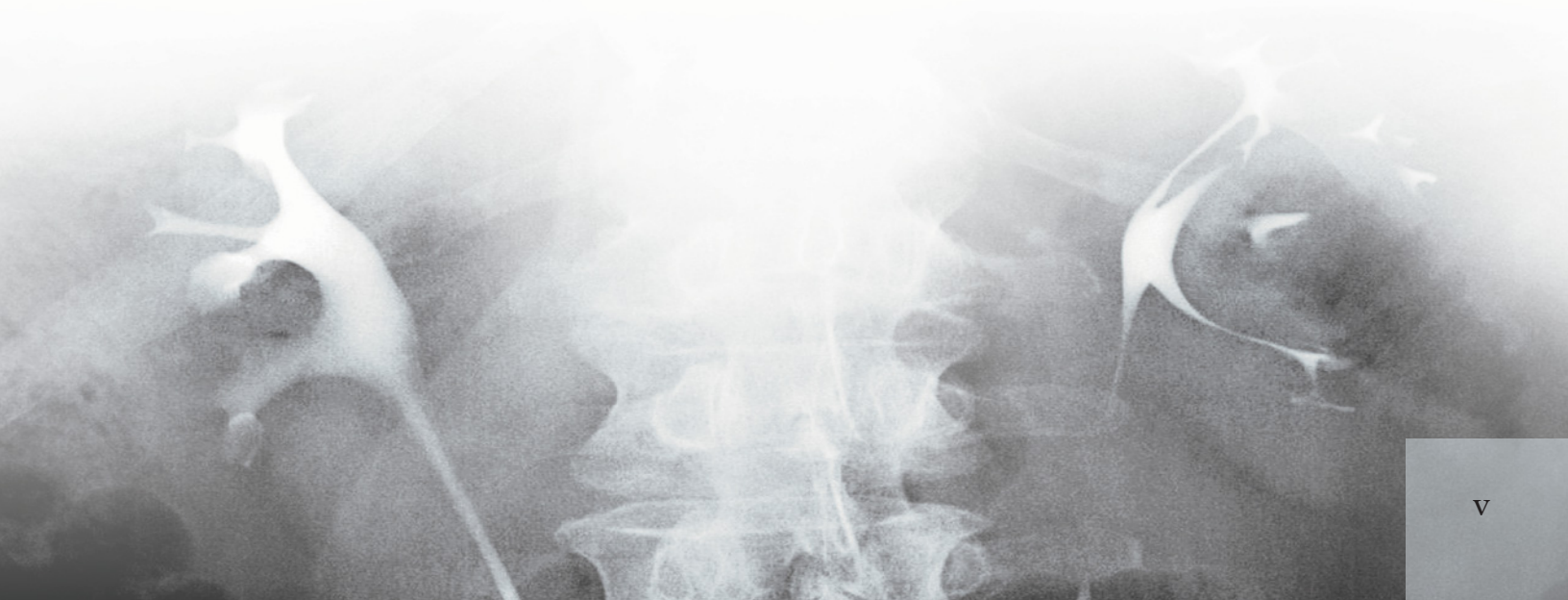
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To Liz, for her patience, support, and love.

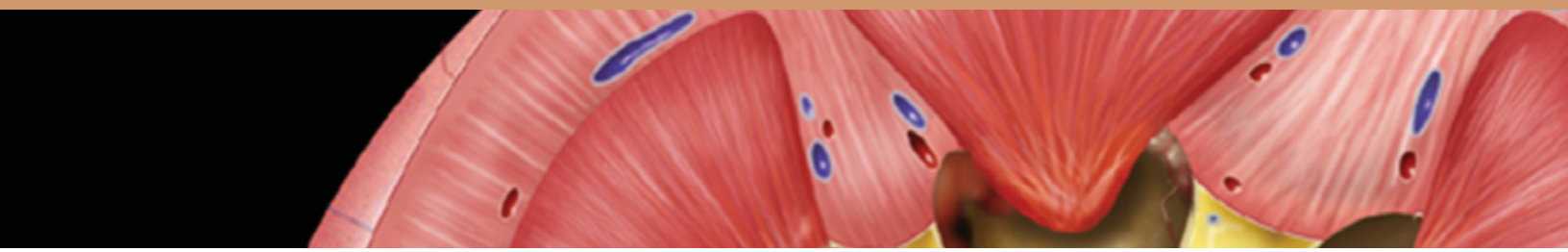
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To Chiara, Sofia, and Oscar for their unwavering love and support.
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To my parents, Michelene and Tom.

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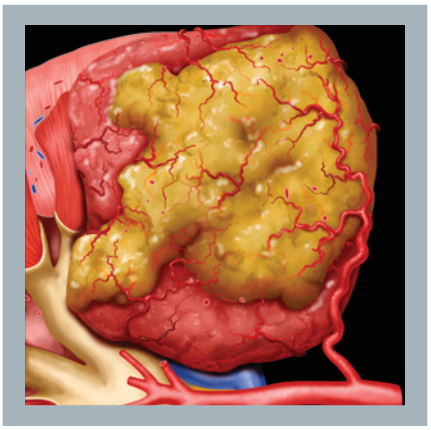
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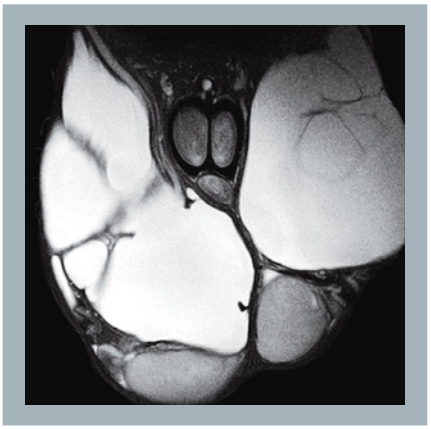
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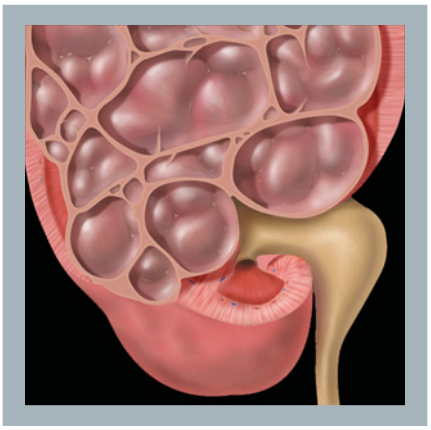
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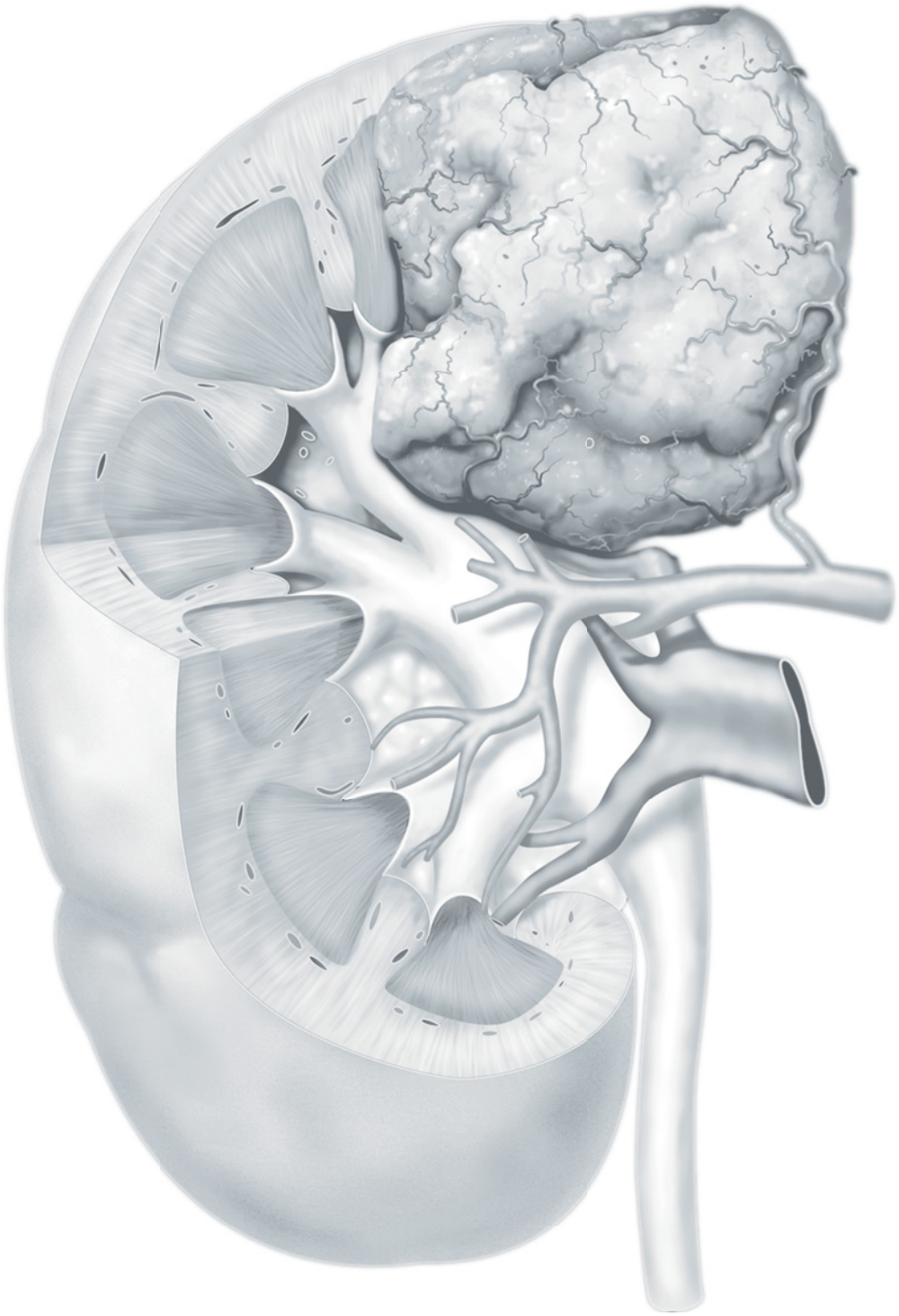
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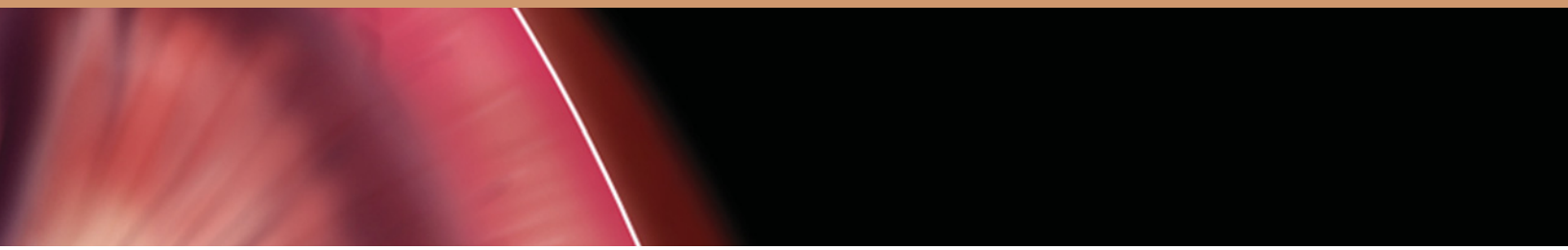








Preface



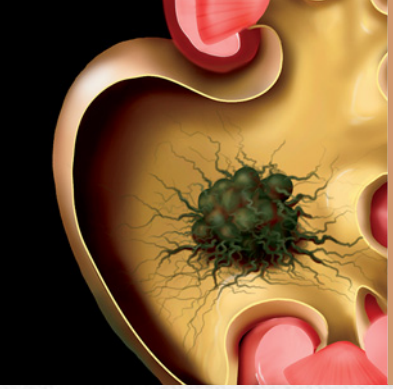
Abdominal imagers recognize that a formal rigid split between GU and GI radiology no longer works with the rapid advancements in cross-sectional imaging that have occurred over the past generation. Nonetheless, when we began planning for the third edition of *Diagnostic Imaging: Abdomen*, we realized that it was no longer possible to encompass the imaging and management of the entire spectrum of abdominal disorders in one text. Thus we decided to separate diagnoses considered to be largely genitourinary (highlighted in this updated and expanded text) from topics judged to be gastrointestinal (covered in a corresponding, companion book).

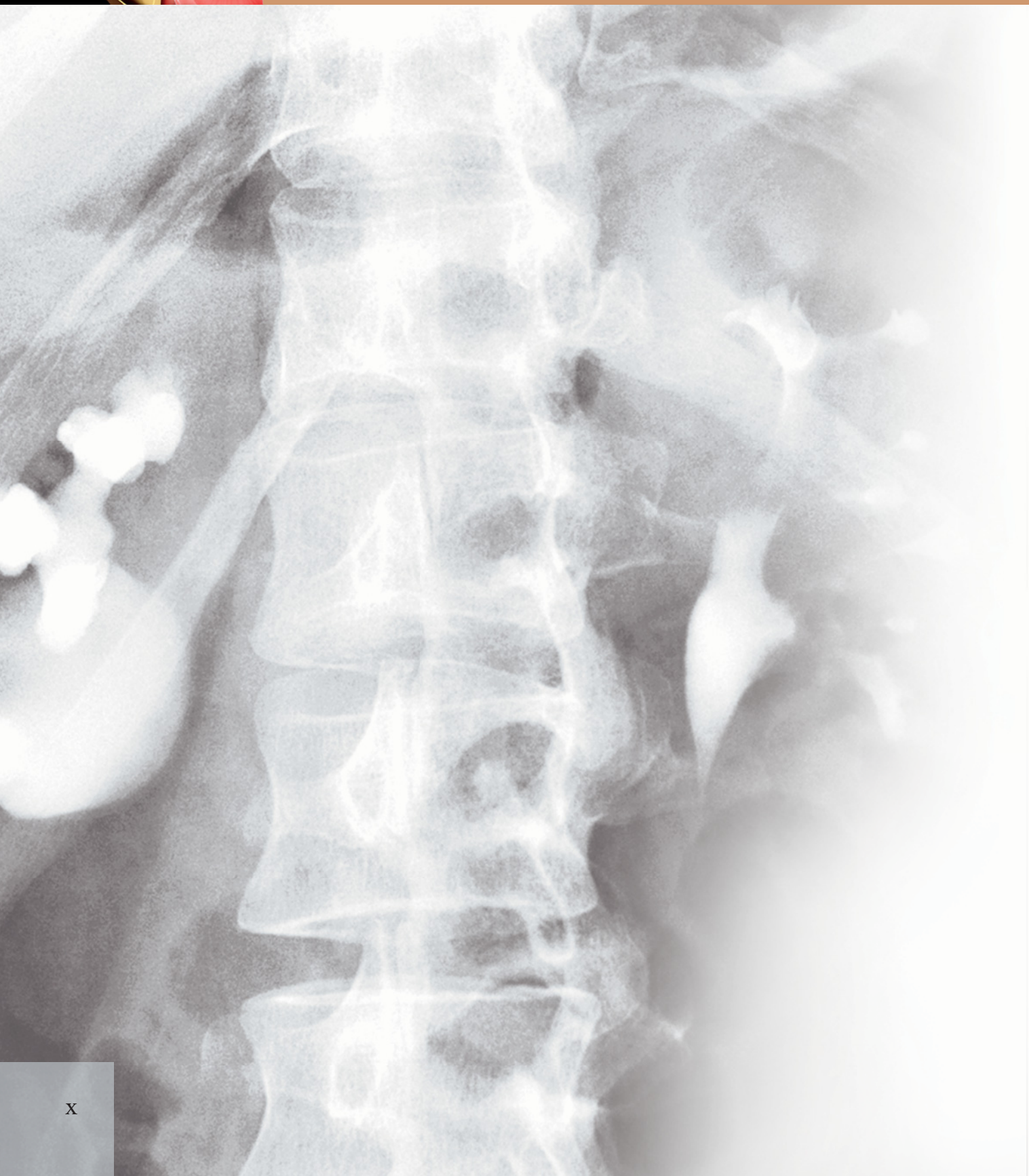
The Amirsys format of bulleted text — including key facts, imaging features, exam protocols, pathology, clinical manifestations/treatment, and image interpretation pearls — is still employed. Succinct but exhaustive outlined descriptions of GU diagnoses are supplemented by image galleries that emphasize the pearls and pitfalls of modern GU imaging. The number of genitourinary diagnoses has been significantly expanded, and the utility of state of the art cross-sectional imaging is emphasized in updated image panels. The approach mirrors current practice: Examples of traditional imaging techniques utilized by GU radiologists decades ago (retrograde, intravenous urography, angiography, etc.) have largely been replaced by their cross-sectional counterparts (MDCT, MR, CTA, MRA, CT urography, MR urography) employed by present day abdominal imagers. Introductory narrative overview chapters describe relevant GU anatomy, physiology, imaging protocols and work-up. GU oncology staging and interventional technique chapters —crucial material for current abdominal imagers—have also been added. Finally, additional images and references are included in the Elsevier Expert Consult eBook that accompanies the print version of *Diagnostic Imaging: Genitourinary, Third Edition*.

This new edition of *Diagnostic Imaging: Genitourinary* was compiled by members of the Abdominal Imaging Division of the University of Pittsburgh. The division has a long history of excellent collaboration with one another, and with referring clinical services. This collaboration has resulted in a legacy of clinical and academic success, and will be even more important in a healthcare environment that appropriately emphasizes value over volume. We are confident that this updated edition will be a go-to, readily accessible, clinically relevant resource for abdominal imagers and trainees for years to come.

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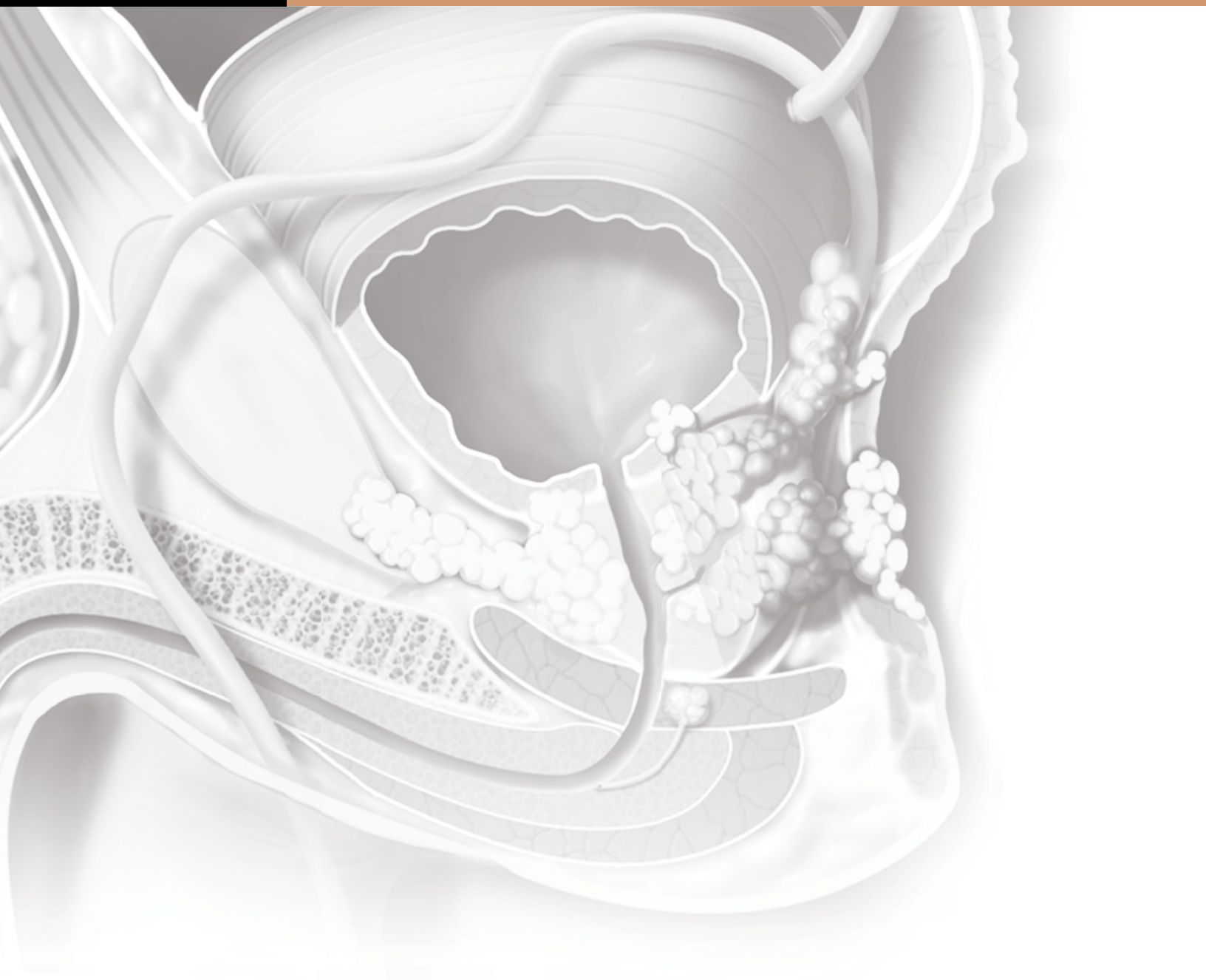
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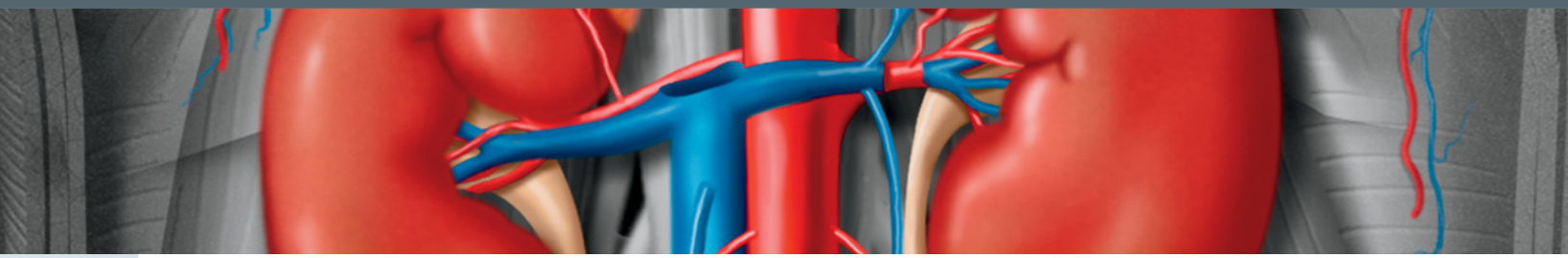
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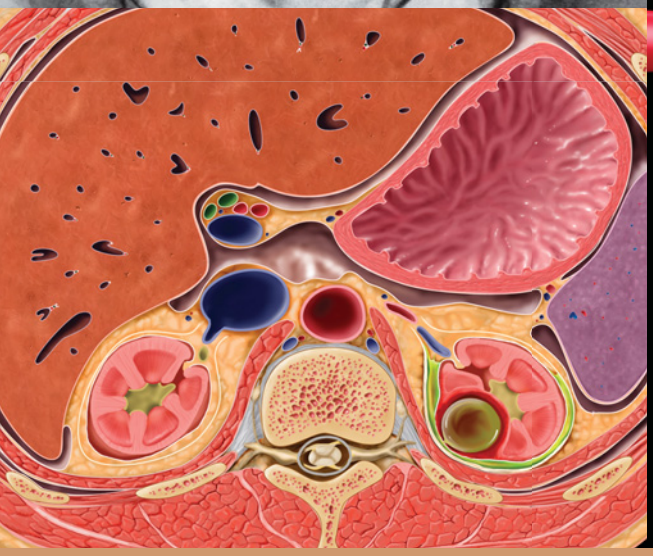
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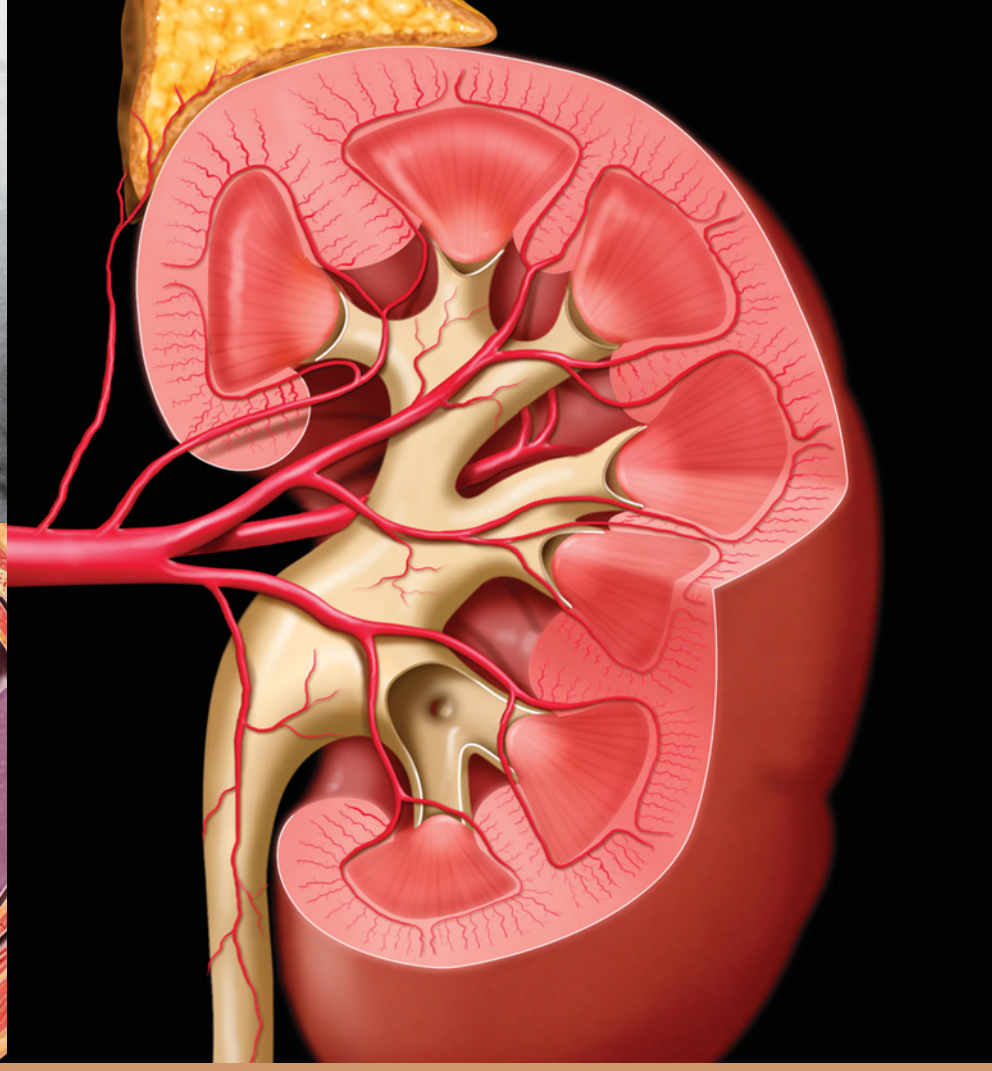
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Diagnostic Imaging

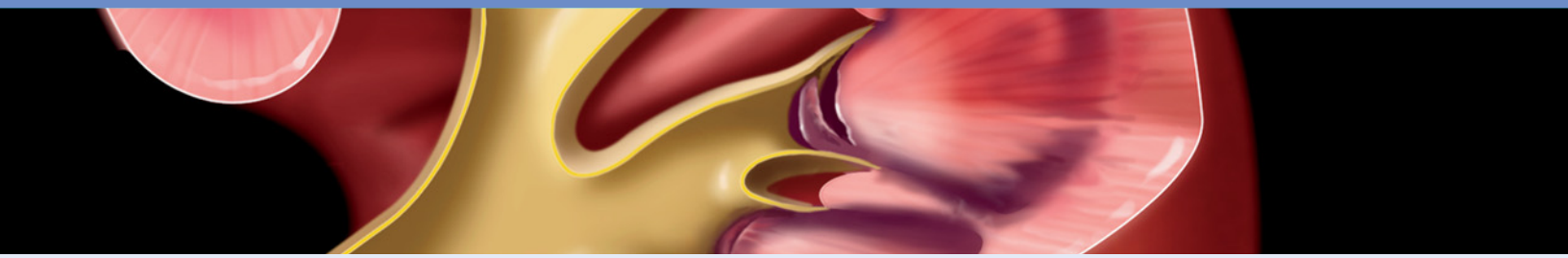
Genitourinary

THIRD EDITION

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SECTION 1

Overview and Introduction



Imaging Approaches 4

Renal Mass Evaluation

CT: Although rapid improvements in scanner technology have resulted in dramatic increases in spatial and temporal resolution, the imaging marker for potential neoplasia (enhancement of soft tissue components of renal masses after contrast administration) has not changed over several generations of CT scanners. Typical renal mass protocols consist of NECT of the kidneys followed by contrast-enhanced images obtained during nephrographic and excretory phases (roughly defined as 85-120 seconds and 3-5 minutes, respectively, post-contrast injection). Corticomedullary phase images (25-70 seconds post contrast) may also be obtained, though small, central, endophytic tumors may not be perceived given the lack of medullary enhancement during this phase. An increase in soft tissue attenuation of soft tissue components post-contrast administration of > 10-20 Hounsfield units (HU) is classically considered to be indicative of true enhancement, though "pseudoenhancement" of small lesions may result in false-positive examinations, and slowly enhancing lesions (e.g., papillary carcinomas) may be missed if delayed images are not acquired. Despite these caveats, if such a protocol is employed, most renal masses can be accurately characterized as potential surgical lesions (i.e., enhancing: Benign/malignant neoplasia), or leave-alone entities (either fat-containing angiomyelolipomas or simple-/high-attenuation cysts). Assessment of cyst complexity at CT, utilizing features described by Bosniak a generation ago, also aids in the triage of this particularly problematic category of renal masses. Suffice to say that increasing levels of complexity (septa thickness, wall/nodularity enhancement, and calcification) increase the likelihood of malignancy.

Optimized protocols should utilize the advantages of multidetector technology. Thin (.625-1.25 mm) source images are acquired and thicker (e.g., 2.5 mm) axial, coronal images are reconstructed. Such protocols allow for diagnostic coronal reconstructed image sets with isotropic or near isotropic resolution; these are particularly helpful for staging of renal cell carcinoma. The potential utility of dual energy for the characterization of renal masses is a topic of current intense investigation. These platforms may permit assessment of enhancement by calculating iodine concentration in the mass, though criteria for meaningful changes have not been universally accepted. The reproducibility of post-processed HU calculations will also need to be validated.

MR: Lesion enhancement is the feature that is also primarily assessed with tailored renal MR protocols. MR offers several unique advantages: Numerous fat-saturation T1-weighted dynamic enhanced phases can be obtained directly in a coronal plane, and dedicated Gadolinium (Gd)-enhanced delayed coronal imaging is the standard of care for identification and characterization of renal vein-caval thrombus. Chemical shift imaging and frequency selective fat-saturation technique may be utilized to identify the fat that is characteristic of angiomyelolipomas. Assessment of T2 and T1 signal intensity improves diagnostic accuracy: Enhancing T2-hyperintense lesions are likely renal cell carcinomas, whereas enhancing T2-hypointense lesions are typically either nonfat-containing AMLs or papillary renal cell carcinoma. Diffusion-weighted imaging assessment may also be employed, though the apparent diffusion coefficient (ADC) value overlap between benign and malignant renal lesions has limited the utility of this technique. A final caveat regarding MR assessment of renal masses is that gadolinium enhancement of renal masses on MR remains a largely qualitative biomarker of neoplasia (though one that is better assessed by evaluating subtracted images).

Ultrasound: Despite marked improvements in ultrasound platforms over the past 40 years, the role of ultrasound in the renal mass imaging algorithm has not changed: Ultrasound is typically performed to determine if a lesion is cystic or solid. Technique should be optimized: Compound and harmonic imaging should be routinely employed to reduce artifacts and to obtain sufficient penetration. Color Doppler may be utilized to help identify pseudolesions (e.g., column of Bertin, renal scars) and rarely, to identify flow within solid or complex cystic masses. Microbubble contrast agents have been employed in several centers to increase the accuracy of ultrasound assessment of renal masses: Unlike CT and MR, enhancement of masses may be observed at continuous real-time examination using low mechanical index protocols, although standards for enhancement are not uniform, and FDA approval for this indication is still forthcoming.

Hematuria (and Potential Urothelial Malignancy) Evaluation

CT urography (CTU): CTU has supplanted conventional intravenous urography as the noninvasive imaging gold standard for the evaluation of patients with hematuria when nephrologic causes (e.g., urinary tract infection, glomerulonephritis) have been excluded. Nonetheless, societies differ on age cutoffs for its use and whether ultrasound may be employed as a primary screen in selected, typically younger, populations. Guidelines for optimizing CTU also widely differ. The basic CTU study includes an initial NECT (to assess for calculi and as a baseline for assessing lesion enhancement if ultimately present), a nephrographic phase for identification of potential renal lesions, and an excretory phase for depicting upper and lower tracts. Coverage and imaging technique may vary, though 64-slice MDCT should be employed to obtain thin-slice source images for multiplanar and MIP reconstructions. Approaches to minimize radiation dosing include multiple new iterative reconstruction algorithms, selective coverage, and split-bolus protocols. CTU performed with dual energy scanners may be a theoretically attractive option to decrease dosing because a virtual NECT may be obtained, though punctate calculi are sometimes obscured. Finally, several options to optimize ureteral opacification (oral hydration, saline administration, low-dose diuretics, prone positioning, and compression) have been employed, but comparison of efficacy between studies is difficult, results have varied, and the logistics of several of these techniques can be daunting. Many centers continue to use simple oral hydration with 1 liter of water 30-60 minutes prior to imaging, and a 3-phase protocol with satisfactory results.

MR urography (MRU): MRU is employed in some centers for evaluation of upper tract malignancy and for identification of urinary tract anomalies. A typical MR urogram includes static fluid heavily T2-weighted sequences and excretory T1-weighted images after gadolinium administration. Like with CTU, saline &/or diuretic administration may improve contrast distribution throughout upper tracts. The renal parenchyma is screened with T1-, T2-weighted images and nephrographic phase Gd-enhanced MR. An abbreviated static T2 MRU protocol may be performed in patients with renal insufficiency to avoid the potential risk of nephrogenic system sclerosis.

Such an approach may also be a viable alternative for pregnant patients with potential renal colic. Use of dedicated phase array coils improves resolution, though it should be noted that both static T2-weighted MR and excretory T1-weighted MR are prone to motion artifact and lack the spatial resolution of CTU; identification of small calculi may be particularly problematic.

Intravenous urography, retrograde urography: Intravenous urography is rarely performed; it includes tomographic images obtained approximately 1 minute post-contrast administration and delayed views of the kidneys, ureters, and bladder. Problematic areas may be evaluated at fluoroscopy and compression improves upper tract distention. Retrograde urography is invasive, though it provides a necessary road map for cystoscopic procedures and may be helpful for confirmation of equivocal lesions suggested at either CTU or MRU.

Renal Dysfunction Evaluation

Ultrasound: Ultrasound is the modality typically employed during the initial assessment of the patient with renal insufficiency. Collecting system dilatation, renal size, symmetry, and echogenicity are easily assessed. The utility of ultrasound in nonselected patients in the acute setting has been called into question, however; the likelihood of bilateral ureteral obstruction (manifested by hydronephrosis at ultrasound) in patients without a prior history of hydronephrosis, abdominal/pelvic malignancy, or pelvic surgery is extremely low. An additional limitation of the modality is that it is an anatomic test; it is impossible to differentiate between obstructive and nonobstructive pelvicaliectasis based upon ultrasound alone.

Preliminary work in the radiology literature, subsequently built upon by other disciplines, has suggested a role for Doppler in assessing the changes in renal hemodynamics that occur with varying causes of renal dysfunction. Most papers have employed the resistive index (RI) (peak systolic velocity-end diastolic velocity/peak systolic velocity) as a surrogate for renal vascular resistance. Recent work has shown, however, how changes in segmental renal arterial RIs are due to tissue compliance (not vascular resistance) and that the RI is neither a specific nor sensitive parameter for the many causes of renal failure. Advocates for contrast-enhanced ultrasound have recently proposed a more elegant ultrasound approach for assessing renal physiology: Tissue perfusion (not flow velocities assessed by Doppler) may be calculated by analyzing replenishment kinetics after bubbles are transiently destroyed by a high mechanical index pulse. However, these techniques have not been adopted by practices outside of select research centers given the lack of FDA approval of microbubble agents and the logistics involved with bubble studies. Despite these limitations, the low cost, portability, and lack of radiation associated with ultrasound have solidified its central longstanding role in the evaluation of the patient with renal dysfunction. Ultrasound is also used for renal biopsy guidance in those patients in whom renal dysfunction is unexplained and persistent.

CT: Like with ultrasound, traditional CT largely evaluates gross anatomic causes for renal failure. It may be helpful for depicting obstructing pelvic malignancies or infiltrating renal neoplastic or inflammatory processes. Several centers have assessed the potential role of MDCT for extracting functional parameters (renal perfusion, renal blood flow, and glomerular filtration) from renal time-density curves obtained during intravenous contrast administration. Patlak modeling of renal contrast kinetics has shown promise in evaluating the pathophysiology of a variety of renal diseases, but the need for central venous line placement, radiation exposure, and (most importantly) large IV contrast doses for renal contrast kinetic studies has prevented its use in patients with preexisting renal dysfunction.

MR: Gd contrast kinetics may also be analyzed to assess split renal function and global glomerular filtration rate (GFR), although like with CT function studies, these approaches have not been validated in large-scale studies. The length of the examination, reproducibility issues, motion artifact, and the potential of Gd agents to induce nephrogenic systemic fibrosis in patients with renal disease have limited the utility of Gd-enhanced renal function studies. Nonenhanced functional MR approaches may ultimately be incorporated into clinical practice; however, diffusion-weighted MR has been proposed as a noninvasive exam for identifying renal fibrosis, and blood oxygen level-dependent (BOLD) MR techniques may assess changes in renal and cortical oxygen tension (and, by extension, renal blood flow).

Nuclear medicine: Various nuclear medicine techniques are still employed to assess renal function. Tc-99m MAG3 is the agent of choice for dynamic radionuclide renal imaging at most centers. Clearance of this agent is primarily via tubular secretion. Renograms can be evaluated to assess urinary uptake, transit, excretion, and split renal function. Lasix renography may be employed to help differentiate between obstructive and nonobstructive pelvicaliectasis. Changes in renogram curves after administration of captopril (an angiotensin-converting enzyme inhibitor) may be helpful for detecting renovascular hypertension and assessing the impact of surgical or angiographic intervention. Tc-99m DMSA is still the preferred radiopharmaceutical for static parenchymal imaging. This agent concentrates within the renal cortex and best shows functioning tubular mass. As such, it is useful for assessing renal scarring (due to urinary tract infections or reflux nephropathy) and calculating relative renal function. Finally, a variety of radionuclide techniques for GFR estimation may be employed, although in routine clinical practice, estimates of GFR are typically made using equations that incorporate serum creatinine, weight, and multiple additional readily measured parameters. It should be noted that, despite their widespread use, all of these equations (including the most commonly employed Modification of Diet in Renal Disease [MDRD] formula) are limited in patients with unstable renal function. Estimates in certain populations (e.g., cirrhotic or pregnant patients) may be particularly prone to error.

Adrenal Mass Evaluation

CT: The high incidence of incidental adrenal masses has prompted a variety of imaging approaches for their characterization. The work-up of these often asymptomatic lesions (or adrenal "incidentalomas") remains imaging intensive, despite large-scale retrospective studies that have shown that the vast majority of small (< 4 cm), incidental adrenal lesions identified at cross-sectional imaging are benign (i.e., adrenal adenomas, cysts, myelolipomas).

Two basic approaches for characterizing adrenal masses at CT are utilized. The attenuation of the adrenal lesion at NECT is assessed. An attenuation of < 10 HU is highly specific for an adenoma (due to the high lipid content of most adenomas).

Attenuation of < 10 HU might also suggest a true endothelial or post-traumatic pseudocyst (another typically benign lesion). Although differentiation between simple cysts and adenomas is usually not clinically relevant, thin wall calcification is a common feature of cysts.

The small percentage of adenomas that do not contain much intracellular lipid may be identified by evaluating contrast kinetics. The washout of contrast from so-called lipid-poor adenomas is typically brisk, as opposed to metastases. Delayed absolute or relative washout of iodinated contrast may be calculated using readily available web-based calculators. Brisk washout may also be seen in a small percentage of pheochromocytomas and vascular metastases (e.g., renal cell carcinoma, hepatocellular carcinoma), although in these cases, an appropriate endocrine evaluation and clinical history may help avoid an errant diagnosis of an adenoma. Finally, the incidental large (> 4 cm) but imaging benign adrenal mass remains a management dilemma. Current dogma continues to suggest that in appropriate surgical candidates, these lesions should be resected given the concern for adrenal cortical carcinoma.

MR: Like NECT, chemical shift MR is employed to identify the lipid content of adrenal adenomas. Relative percent signal suppression at out-phase imaging may be used to increase diagnostic confidence, but qualitative assessment often suffices. Early MR studies suggested a role of MR for identifying pheochromocytomas, but the classic "light bulb" T2-bright appearance is neither a sensitive nor specific feature of these lesions.

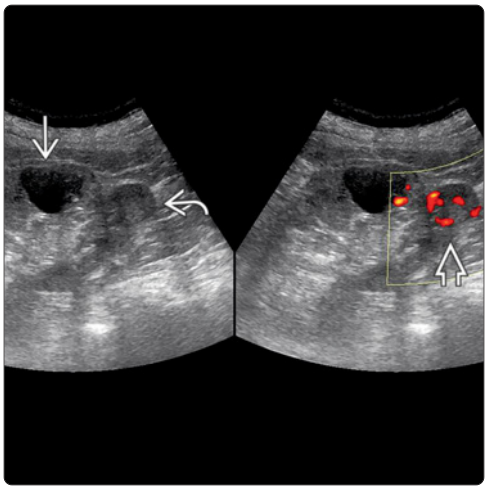
Bladder Mass Evaluation

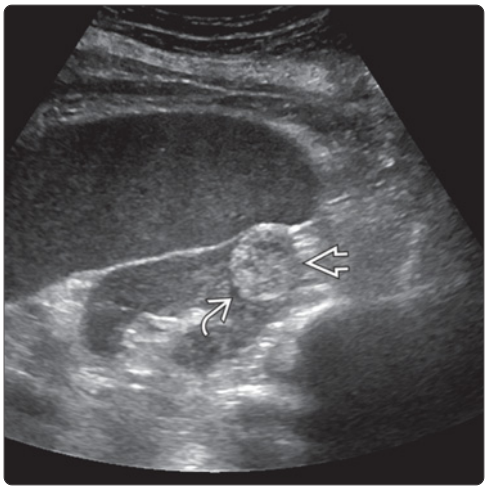
CT: The sensitivity and specificity of CTU for the diagnosis of bladder cancer in patients with hematuria is over 90%. MDCT is readily available, and the recommendations of multiple societies have highlighted its effectiveness in assessing visceral and nodal metastases pre and post therapy. Even well-performed, state of the art MDCT often fails with local T staging, however. The depth of muscle invasion is frequently under- or overestimated, though CT performs better with higher T score tumors. Thus, cystoscopy remains in every algorithm of the investigation of hematuria. Imaging-suspected or occult bladder lesions are directly visualized, and if present, biopsy for diagnosis and depth of invasion is performed. Similarly, size and shape remain the only imaging criteria for nodal metastases, and like with other GU malignancies, low-volume nodal disease will not be identified. Recent work has also unfortunately suggested that PET/CT adds little beyond conventional MDCT to local N staging.

MR: The role of MR for the local staging of bladder cancer continues to evolve. The superior soft tissue contrast resolution of MR using standard T1-, T2-weighted imaging sequences allows for better differentiation between bladder wall layers. Multiple studies have shown that compared to CT, MR performs better for identifying intramural tumor invasion and perivesical spread. Nonetheless, MR often fails, and muscle invasion may be under or over called. Multiparametric imaging (combining multiplanar T1/T2 image sets, diffusion-weighted MR, and dynamic contrast-enhanced MR) may improve staging accuracy, though this approach is currently employed at select centers. Preliminary work has also suggested the utility of ultra small super paramagnetic iron oxide (USPIO) particles and diffusion MR for node characterization, though for the meantime, radical cystectomy and extended template node dissection remain the standard of care for the staging and therapy of confirmed muscle invasive tumor.

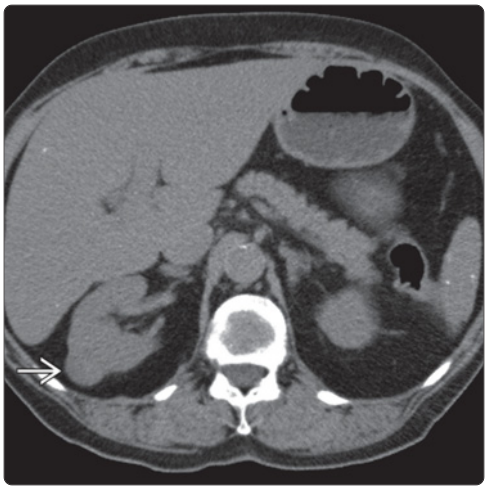
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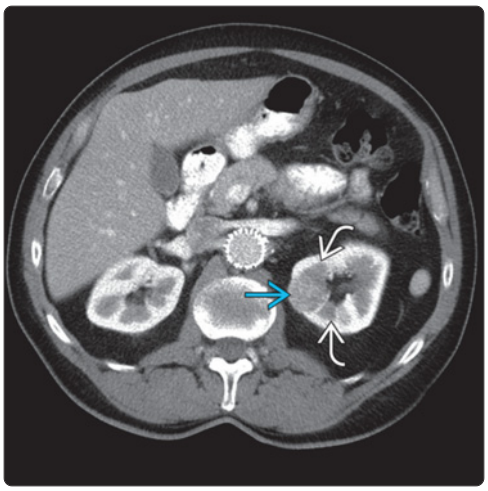


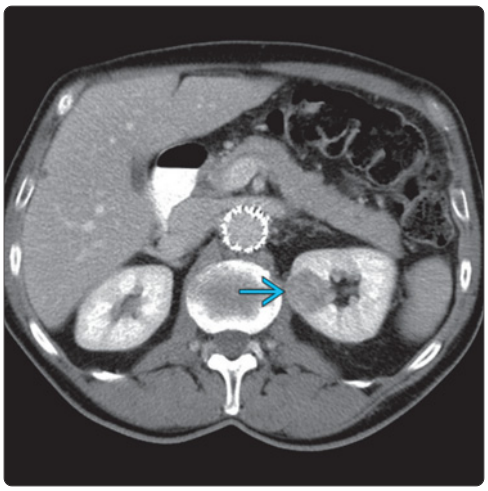
(Left) *Grayscale and power Doppler renal ultrasound performed on a patient with renal insufficiency shows a cyst and an incidental isoechoic lower pole renal cell carcinoma (RCC). Ultrasound is typically performed to differentiate solid from cystic lesions. Intralesional Doppler flow helps confirm malignancy.* **(Right)** *Ultrasound shows an echogenic RCC. Differentiating between an echogenic RCC and AML may be difficult on ultrasound, but a thin halo and the lack of weak shadowing favor RCC.*





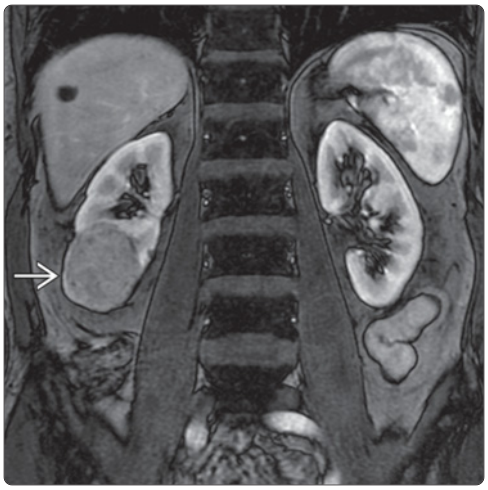
(Left) *NECT shows an exophytic solid attenuation right renal lesion. Either ultrasound or CECT is needed to differentiate between a high-attenuation cyst and solid neoplasia.* **(Right)** *CECT of the same patient performed during the nephrographic phase shows that the lesion enhances by 40 HU. Enhancement is the hallmark imaging marker of neoplasia. Papillary renal cell carcinoma was confirmed at laparoscopic partial nephrectomy.*

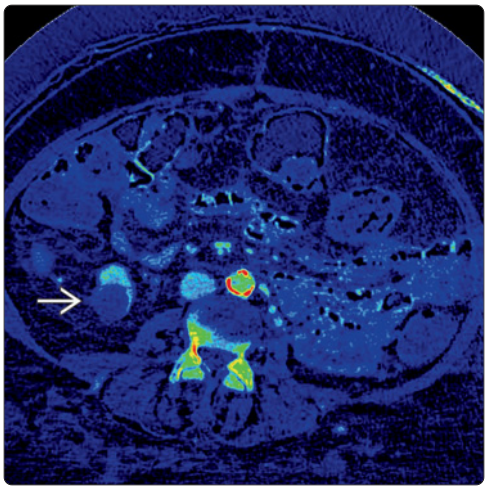




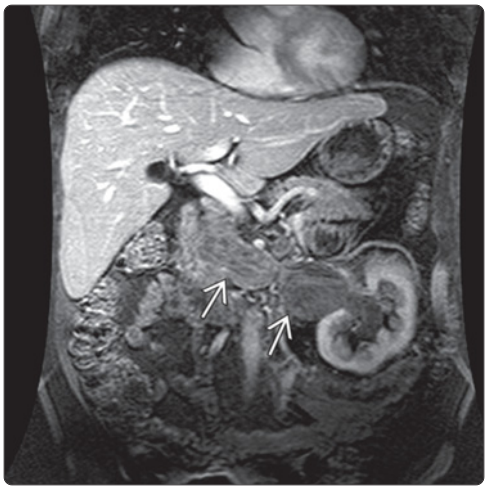
(Left) *CECT performed during the corticomedullary shows a subtle, largely non-border-deforming left upper pole renal lesion. Its attenuation is similar to minimally enhanced surrounding medulla.* **(Right)** *CECT of the same patient during the nephrographic phase shows an obvious, now relatively low-attenuation left upper pole mass (confirmed clear cell RCC). Endophytic renal masses are often imperceptible on the corticomedullary phase. De-enhancement of masses over time is an additional imaging marker of neoplasia.*

(Left) *Gd-enhanced T1WI MR shows an exophytic right lower pole RCC ➡. Assessment of contrast enhancement at MR is largely qualitative; subtraction images may be helpful in problematic cases.* **(Right)** *DECT spectral data is used to generate an iodine-specific overlay map. Qualitative assessment confirms no iodine uptake in a high-attenuation right renal cyst ➡. Early work has suggested that direct quantification of iodine concentration may compete with HU assessment of neoplasia enhancement.*



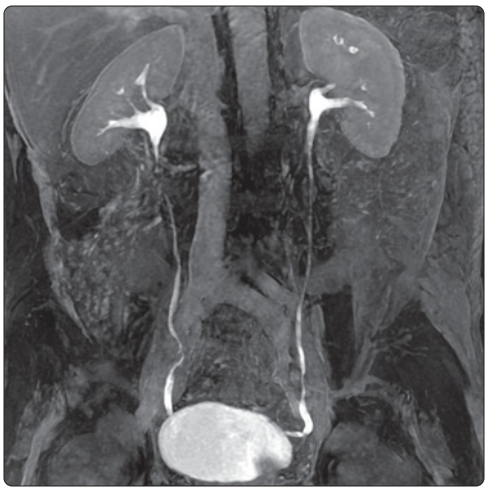


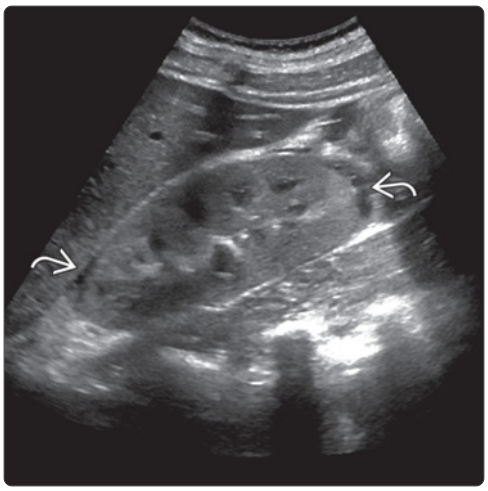
(Left) *Gd-enhanced T1WI MR shows expansile, enhancing tumor thrombus ➡ within the left renal vein in a patient with an adjacent RCC (not shown). Coronal imaging is particularly helpful for the preoperative evaluation of T3 RCC.* **(Right)** *CT urogram in an elderly man with hematuria shows an infiltrating right renal urothelial cell carcinoma ➡. Recent consensus statements advocate CTU for the evaluation of non-nephrologic causes of hematuria, though age guidelines and the utility of ultrasound are still debated.*



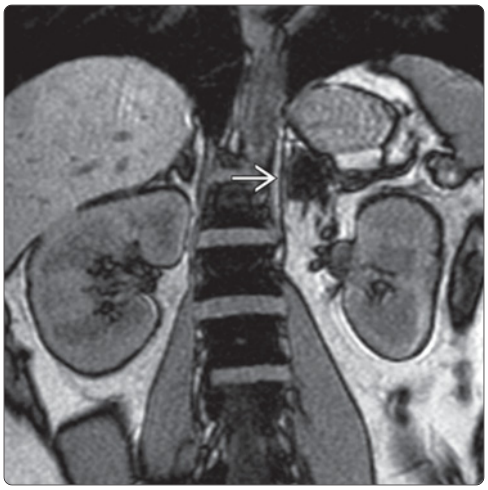


(Left) *Gd-enhanced T1W urogram shows normal upper tracts. Both Gd-enhanced and heavily T2-weighted sequences are part of typical MRU protocols, though image resolution is less than that obtained by CT urography and calculi may not be identified.* **(Right)** *Ultrasound of a patient with acute renal failure shows a normal-sized echogenic kidney (a nonspecific finding of medical renal disease) and perirenal edema ➡. The primary role of ultrasound in this setting is to assess renal size and collecting system dilatation.*

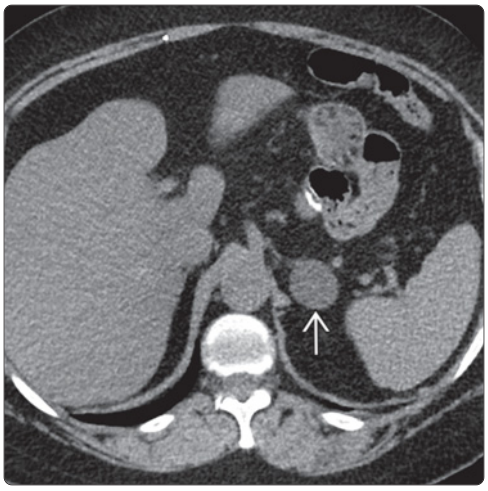


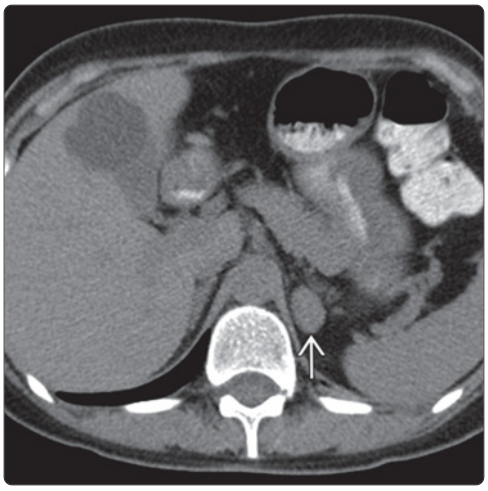




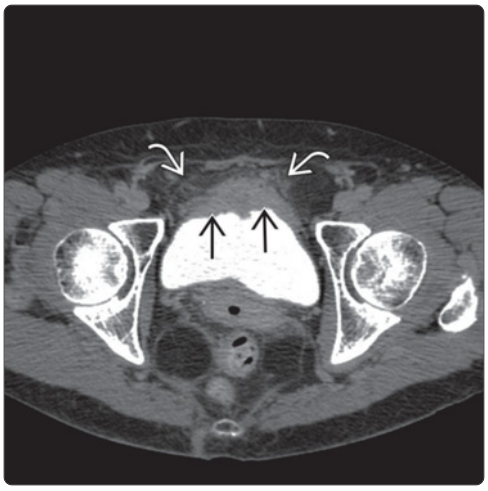


(Left) *In phase T1WI MR in a 37-year-old woman with Cushing syndrome shows an intermediate signal intensity left adrenal lesion ➡. (Right) Out of phase T1WI MR of the same patient shows uniform signal loss of a lipid-rich adenoma ➡. Chemical shift imaging exploits resonance differences between lipid and water to detect the intracytoplasmic fat characteristic of lipid-rich adenomas. Adrenal to spleen suppression ratios may be calculated for problematic cases, but visual assessment typically suffices.*





(Left) *NECT of a patient with Cushing syndrome shows a left adrenal lesion ➡ measuring -2 HU. NECT is an effective screening modality for characterizing lipid-rich adenomas, but the utility of intensive imaging for incidental adrenal lesions has been questioned.* **(Right)** *Baseline NECT shows an incidental, indeterminate, left adrenal mass ➡. Relative and absolute washout percentages on a dedicated adrenal CT (not shown) confirmed a lipid-poor adenoma. Web-based programs are readily available for calculations.*





(Left) *Delayed CECT shows a large, sessile, anterior bladder urothelial carcinoma ⇨. Prevesical fat infiltration ➡ suggests T3 tumor (i.e., tumor extending beyond detrusor). CT is utilized for preoperative evaluation of nodal and visceral metastases, but it is typically not helpful for assessing tumor depth (T stage).* **(Right)** *Sagittal T2WI MR shows a likely muscle-confined (T2) bladder dome urachal adenocarcinoma ⇨. MR performs better than CT for assessing tumor depth, but cystoscopic biopsy remains the standard of care for T staging.*

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SECTION 2

Retroperitoneum



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Relevant Anatomy and Embryology

The parietal peritoneum separates the peritoneal cavity from the retroperitoneum. The retroperitoneum contains all the abdominal contents located between the parietal peritoneum and the transversalis fascia. It is divided into 3 compartments by 2 well-defined fascial planes: The renal and lateroconal fasciae.

The **perirenal space** contains the kidney, adrenal, proximal ureter, and abundant fat and it is enclosed by the renal fascia, which is also referred to as Gerota fascia. The 2 perirenal spaces do not communicate across the abdominal midline.

The **anterior pararenal space** contains the pancreas, duodenum, colon (ascending and descending), and a variable amount of fat.

The **posterior pararenal space** contains fat but no organs; it is contiguous with the properitoneal fat along the flanks.

The anterior renal fascia separates the perirenal space from the anterior pararenal space, and the posterior renal fascia separates the perirenal space from the posterior pararenal space.

The lateroconal fascia separates the anterior from the posterior pararenal space and marks the lateral extent of the anterior pararenal space.

The renal fascia joins and closes the perirenal space resembling an inverted cone with its tip in the iliac fossa. Caudal to the perirenal space, in the pelvis, the anterior and posterior pararenal spaces merge to form a single infrarenal retroperitoneal space, which communicates directly with the pelvic prevesical space (of Retzius). Due to an opening in the cone of the renal fascia caudally, the perirenal space communicates with the infrarenal retroperitoneal space. Thus, all 3 retroperitoneal compartments communicate with each other within the lower abdomen and pelvis. All of the pelvic retroperitoneal compartments, such as the perivesical and perirectal spaces, communicate with each other, which is evident and clinically relevant in cases of pelvic hemorrhage or tumor as well as with extraperitoneal rupture of the urinary bladder.

The renal and lateroconal fascia are laminated planes, which can split to form potential spaces as pathways of spread for rapidly expanding fluid collections or inflammatory processes, such as hemorrhage or acute pancreatitis. Splitting of the anterior renal fascia creates a "retromesenteric plane" that communicates across the midline; splitting of the posterior renal fascia creates a "retrorenal plane," which also communicates across the midline and anteriorly. Knowing this principle is crucial to understanding how disease originating in the anterior pararenal space, such as acute pancreatitis, can extend posterior to the back of the kidney or how fluid collections within the posterior pararenal space or retrorenal plane can extend around the lateral or even anterior abdominal wall.

Imaging Techniques and Indications

Multiplanar CT and MR are ideally suited to display the anatomy and pathology of retroperitoneal disease processes. Use of intravenous contrast material allows easier recognition of fascial plane landmarks and pathology and should be used unless contraindicated.

Approach to Retroperitoneal Abnormalities

Perirenal Space

Disease within the perirenal space is usually the result of diseases of the kidney. Common disease states include hemorrhage, infection, inflammation, and neoplasia.

The renal fascia is very strong and is usually effective in containing most primary renal pathology within the perirenal space. Similarly, it usually excludes most other processes from invading or involving the perirenal space.

The perirenal space is divided irregularly and inconsistently by perirenal bridging septa that often result in loculation of perirenal fluid, which may be misinterpreted as subcapsular in location. The perirenal septa also act as conduits for fluid or infiltrative disease, including tumor, to enter or leave the perirenal space.

Perirenal fluid may represent blood, urine, or pus or may be simulated by inflammation of the perirenal fat. Hemorrhage is often due to trauma, but may occur due to anticoagulation, rupture of a renal tumor, or vasculitis. Pus or inflammation usually originates from acute pyelonephritis, which may be associated with an abscess. Perirenal urine ("urinoma") may result from trauma with laceration through the renal collecting system, but it usually resolves rapidly unless there is an obstruction to the flow of urine to the bladder. Acute urine extravasation may also accompany ureteral obstruction by a calculus due to forniceal rupture.

Renal cell carcinoma is common, and the renal fascia usually confines the tumor, preventing invasion of contiguous structures. Spread to lymph nodes or hematogenous spread through the renal vein and inferior vena cava may occur and constitute important elements of the imaging and staging of this tumor.

Anterior Pararenal Space

Disease within the anterior pararenal space is common. For example, acute pancreatitis results in peripancreatic infiltration ± fluid collections that spread throughout the anterior pararenal space, often affecting the duodenum and ascending and descending colon segments that share this anatomic compartment. The spread of inflammation is usually limited posteriorly by the anterior renal fascia, and laterally by the lateroconal fascia. Thickening of these planes is a reliable clue as to the presence of pancreatitis, which might otherwise be occult on imaging. The perirenal space is usually not involved in acute pancreatitis, sometimes resulting in a striking appearance of a perirenal "halo" of fat density while other retroperitoneal spaces and planes are infiltrated. Ventral (anterior) spread of inflammation or tumor from the anterior pararenal space is not limited by any fascial boundary, but only by the posterior parietal peritoneum. The root of the mesentery and transverse mesocolon originate from just ventral to the 3rd portion of duodenum and pancreas, and disease originating in these organs may easily dissect into the mesentery without crossing any anatomic boundaries. Some refer to the spaces enclosed by the mesenteric layers as the "subperitoneal space," emphasizing that there is no inviolate separation between the intraperitoneal and retroperitoneal spaces.

A duodenal ulcer may perforate and result in extraluminal gas and fluid that occupy 1 or more spaces, including the anterior pararenal, intraperitoneal (as the duodenal bulb is an intraperitoneal structure), and even the perirenal space since

the latter is open at the renal hilum and communicates with the anterior pararenal space.

Posterior Pararenal Space

Disease originating within the posterior pararenal space is uncommon, essentially limited to hemorrhage and tumor.

"Retroperitoneal hemorrhage" is a misnomer since most spontaneous, coagulopathic hemorrhage originates within the abdominal wall, the iliopsoas compartment, or the rectus sheath. Only when hemorrhage extends beyond these fascial boundaries does it enter the retroperitoneum. Rectus sheath hematomas enter the extraperitoneal pelvic spaces through a defect in the caudal (infraumbilical) portion of the sheath. Iliopsoas hemorrhage often extends into any or all of the retroperitoneal compartments, predominantly along the main fascial planes. The hallmarks of coagulopathic hemorrhage are: Bleeding out of proportion to trauma, multiple sites of bleeding, and the presence of the "hematocrit" sign, a fluid-cellular debris level within the hematoma.

Retroperitoneal sarcomas, most commonly liposarcoma, often originate within 1 of the retroperitoneal compartments, and the site of origin can be determined by the relative mass effect on various organs and structures such as the kidneys, colon, and great vessels. Most liposarcomas have some identifiable fat within them and seem to be encapsulated, allowing for excision, although recurrent disease is common.

If retroperitoneal nodes are included in the discussion, the most common retroperitoneal tumor is non-Hodgkin lymphoma (NHL). NHL often results in massive lymphadenopathy. This characteristically involves the mesenteric and retroperitoneal nodes that are confluent and anteriorly displace the aorta and inferior vena cava from the spine. Retroperitoneal nodes are also frequently involved by malignancies originating in pelvic organs, such as the prostate, rectum, and cervix.

The other large, though uncommon group of primary retroperitoneal tumors are of neurogenic origin, including nerve sheath tumors, ganglioneuroma, neuroblastoma, and others. These often share the characteristics of appearing as well-defined, moderately enhancing masses that do not appear to arise from nodes nor abdominal viscera. Many, in fact, arise along the sympathetic nerve trunks, while others are part of a syndrome such as neurofibromatosis that may involve multiple nerves in a paraspinal or presacral distribution.

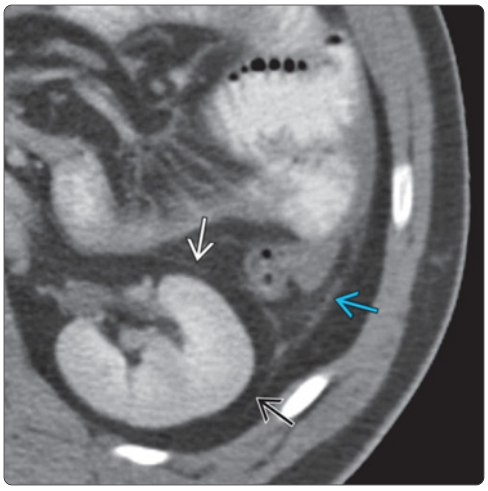
The great vessels, the aorta and inferior vena cava (IVC), are located in the retroperitoneum and are usually depicted as lying within the retromesenteric plane. Although primary disease of the IVC is rare, it may be the site of primary tumor (sarcoma) or the site of spread from a renal or adrenal carcinoma. More common are anomalies of the embryologic development of the IVC. Some 10% of the population have some anomaly of the embryologic sub- and supracardinal veins, usually at or below the level of the renal veins, resulting in variations such as duplicated IVC and retro- and circumaortic renal vein. While these are uncommonly of clinical significance (limited to affecting surgical and interventional procedures), they may be mistaken for pathologic conditions, most commonly enlarged retroperitoneal lymph nodes.

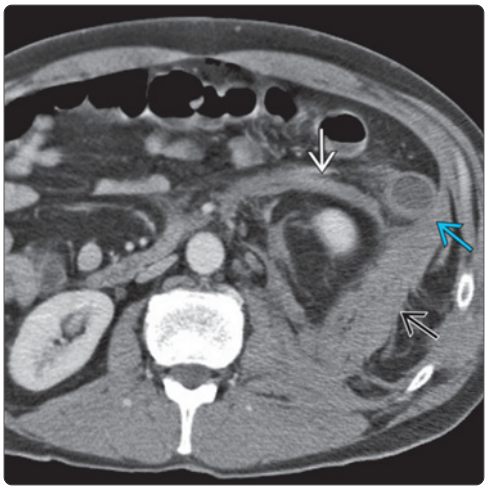
Abdominal aortic aneurysm is a major health concern, and rupture is usually fatal. Accurate diagnosis and precise mapping of the size and shape of an aneurysm allows effective, minimally invasive prophylactic treatment with endovascular stenting.

Retroperitoneal fibrosis is an inflammatory disorder that may be misinterpreted as a malignant process, as it envelops the aorta and IVC, often causing displacement and encasement of the ureters. It may occur as an isolated process or as part of a multisystem autoimmune disorder.

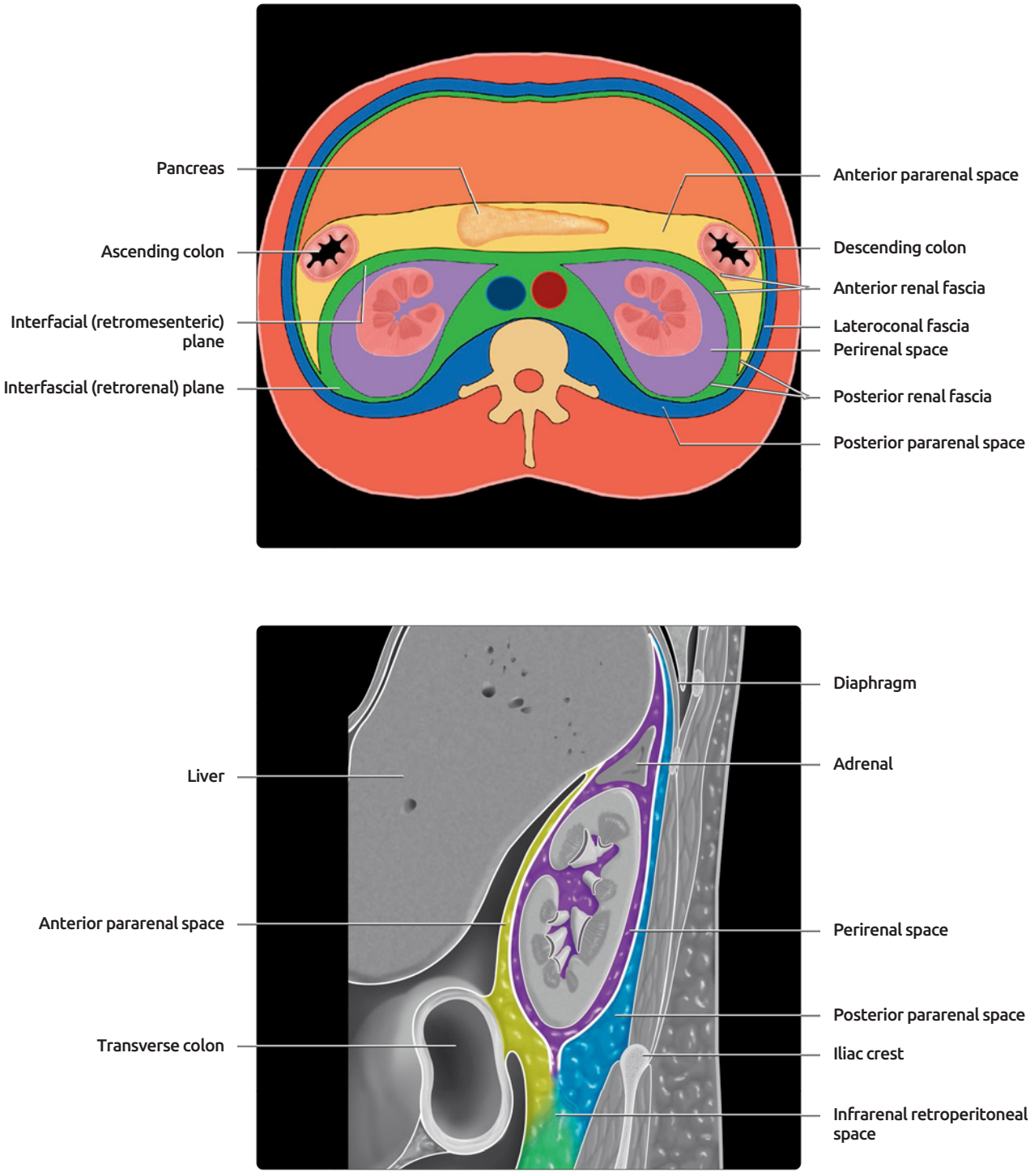
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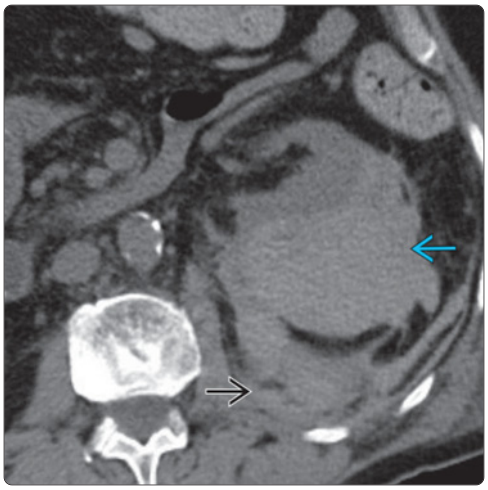


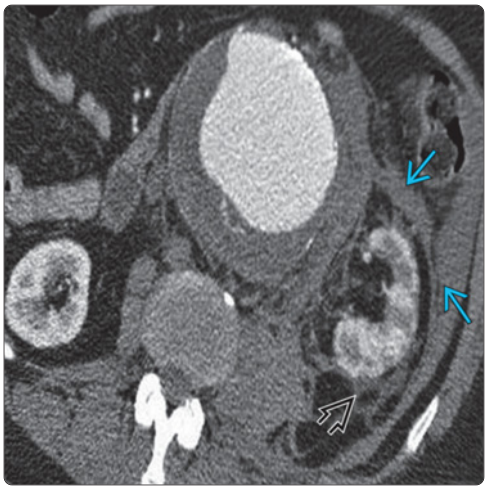


(Left) *Axial CECT shows the normal anterior & posterior renal fascia which fuse to form the lateroconal fascia. Note that the normal fascia are extremely thin.* **(Right)** *Axial CECT demonstrates fluid from pancreatitis dissecting along the retromesenteric plane, formed by the layers of the anterior renal fascia and the retrorenal plane, formed by layers of the posterior pararenal space. Fluid also extends along the lateral conal fascia.*

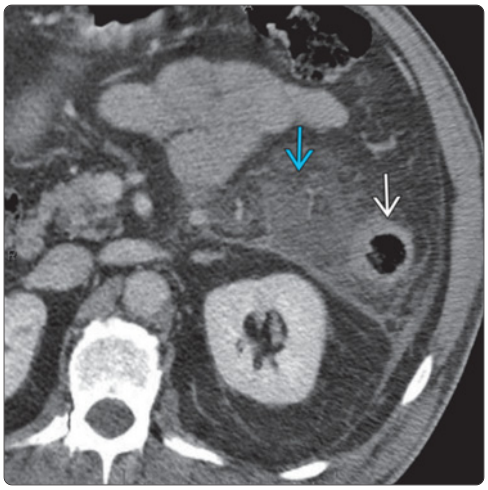


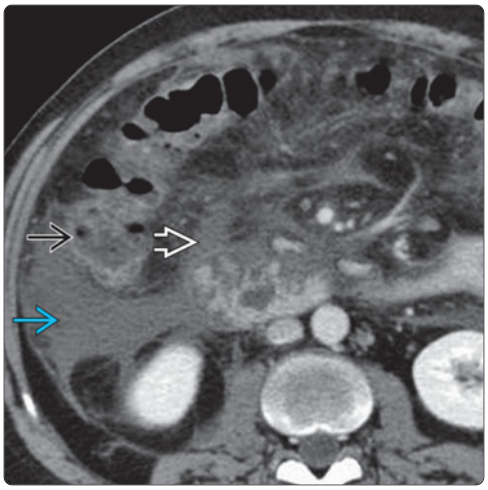
(Top) *The 3 main compartments of the retroperitoneum are the anterior pararenal space (yellow), perirenal space (purple), & posterior pararenal space (blue). The interfascial planes (green) are potential spaces created by inflammatory processes that separate the double laminated layers of the renal and lateroconal fasciae. The posterior pararenal space is synonymous with the properitoneal fat that extends along the lateral & anterior abdominal wall.* **(Bottom)** *Sagittal graphic through the right kidney shows the 3 retroperitoneal compartments. Note the confluence of the anterior & posterior renal fasciae at about the level of the iliac crest. Caudal to this, there is only a single infrarenal retroperitoneal space.*



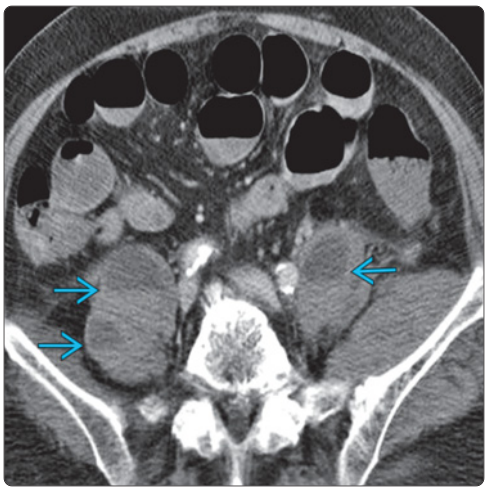


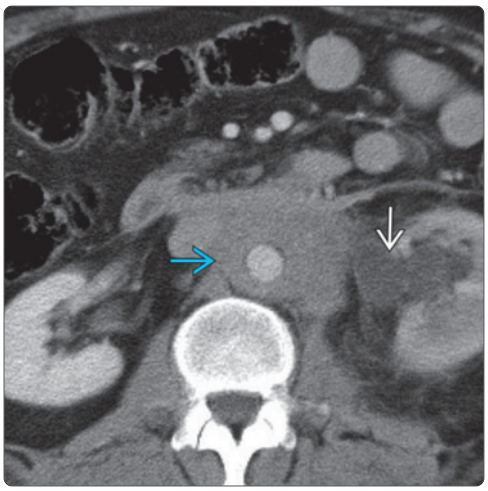
(Left) *Axial NECT in a patient following flank trauma shows that the left kidney is compressed & displaced due to a large subcapsular renal hematoma. There is also hematoma in the posterior pararenal space. Note the lack of a fluid-hematocrit level, a finding that is associated with anticoagulation hemorrhage.* **(Right)** *Axial CECT during the arterial phase reveals hemorrhage dissecting along the left interfascial planes & into the perirenal space due to contained rupture of an abdominal aortic aneurysm.*





(Left) *Axial CECT demonstrates fluid from subacute pancreatitis tracking into the left anterior pararenal space. Note that the descending colon also resides in the anterior pararenal space & is partially surrounded by fluid.* **(Right)** *Axial CECT demonstrates fluid from acute pancreatitis dissecting through the right aspect of the anterior pararenal space. Fluid abuts the ascending colon (also in the anterior pararenal space) & dissects along the proximal aspect of the transverse mesocolon.*





(Left) *Axial CECT in a patient with decreased hematocrit reveals hematomas within both psoas muscles. Note the fluid-hematocrit levels within the hematomas due to anticoagulant hemorrhage.* **(Right)** *Axial CECT shows left hydronephrosis & delayed nephrogram due to obstruction of the proximal ureter. The obstruction results from retroperitoneal fibrosis (RPF) & is shown as a periaortic soft tissue mantle; unlike lymphoma, RPF does not displace the aorta from the spine.*

Duplications and Anomalies of IVC

KEY FACTS

TERMINOLOGY

- Congenital anomalies of inferior vena cava (IVC)

IMAGING

- Duplication of IVC (prevalence ~ 1-3%)
  - Left- and right-sided IVC are present inferior to renal veins
  - Left IVC typically drains into left renal vein, which crosses anterior to aorta to join right IVC
  - Recognition important prior to IVC filter placement
- Left IVC (prevalence ~ 0.2-0.5%)
  - Typically drains into left renal vein, which crosses anterior to aorta to join normal right suprarenal IVC
  - Important variant in repair of abdominal aortic aneurysm and transjugular placement of IVC filter
- Azygos continuation of IVC (prevalence ~ 2-9%)
  - Absence of suprarenal IVC
  - Blood flow enters azygous vein and enters thorax posterior to diaphragmatic crus
  - Enlarged azygos vein empties into SVC normally in right peribronchial location
  - Hepatic veins drain directly into right atrium
  - Important variant in planning cardiopulmonary bypass
- Circumaortic left renal vein (prevalence ~ 2-3.5%)
  - Important variant in nephrectomy planning
  - Rare occurrences of hematuria and hypertension

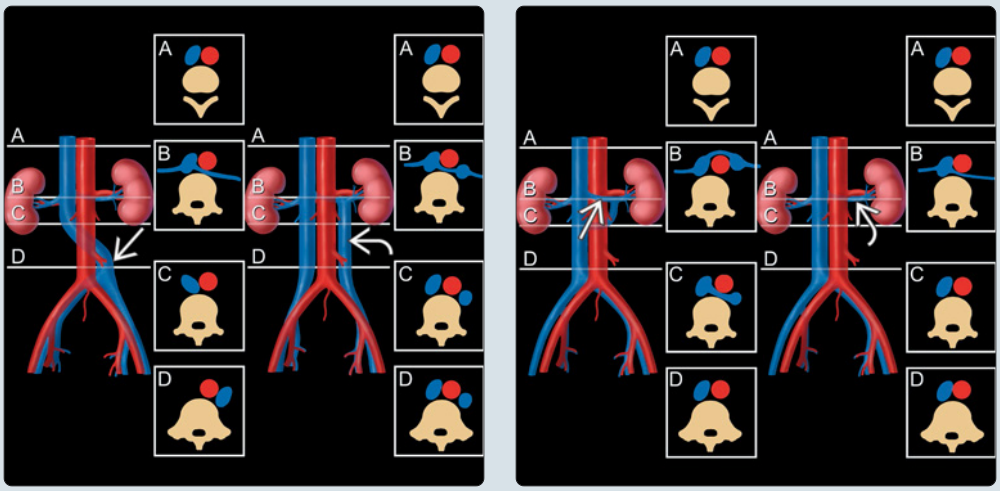
TOP DIFFERENTIAL DIAGNOSES

- Retroperitoneal lymphadenopathy
- Varices/collaterals
- Gonadal vein

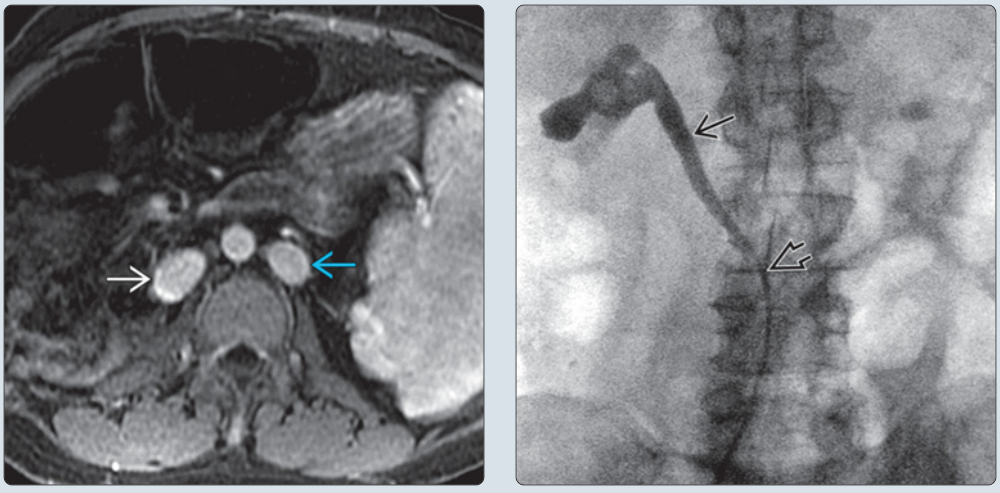
DIAGNOSTIC CHECKLIST

- Preoperative imaging may be important in planning abdominal surgery, liver or kidney transplantation, or interventional vascular procedures
  - e.g., IVC filters, varicocele sclerotherapy, venous renal sampling

(Left) *Graphic shows transposition of the IVC and its more common duplications. The duplicated IVC originates as the left iliac vein caudally and empties into the left renal vein. The boxes labeled "A" through "D" refer to the respective levels of the axial sections.* **(Right)** *Graphic shows a circumaortic left renal vein with a smaller ventral vein crossing cephalad to the dorsal vein.*



(Left) *Post-contrast axial T1 C+ FS MR with fat suppression shows a normal IVC and an additional cylindrical structure to the left of the aorta, consistent with a duplicated IVC.* **(Right)** *AP view during retrograde pyelography shows medial deviation of the right ureter due to its retrocaval course; note mild ureteral dilatation upstream from it coursing posterior to the IVC. The left ureter (not shown) had a normal course.*



TERMINOLOGY

Definitions

- Congenital anomalies of inferior vena cava (IVC)

IMAGING

General Features

- Best diagnostic clue
  - Malposition or duplication of IVC
- Morphology
  - Types of IVC anomalies
    - Duplicated IVC
    - Left-sided IVC
    - Azygos continuation of IVC
    - Circumaortic left renal vein
    - Retroaortic left renal vein
    - Duplication of IVC with retroaortic right renal vein and hemiazygos continuation of IVC
    - Duplication of IVC with retroaortic left renal vein and azygos continuation of IVC
    - Circumcaval or retrocaval ureter
    - Absence of infrarenal or entire IVC

CT Findings

- Duplication of IVC
  - Left- and right-sided IVC inferior to renal veins
  - Left IVC typically drains into left renal vein, which crosses anterior to aorta in normal fashion to join right IVC
  - Duplicated IVCs may have significant asymmetry (left usually smaller)
- Left IVC
  - Drains into left renal vein, which crosses anterior to aorta in normal fashion to unite with right renal vein and form normal right suprarenal IVC
  - ↑ enhancement of right renal vein relative to left renal vein
    - Due to dilution from unenhanced venous return from lower extremities into left renal vein
- Azygos continuation of IVC
  - Absence of suprarenal IVC
  - Infrarenal IVC continues azygous vein, which enters thorax posterior to diaphragmatic crus
  - Enlarged azygos vein empties into SVC normally in right peribronchial location
  - Hepatic veins drain directly into right atrium
  - Enlarged azygos vein has similar attenuation to superior vena cava (SVC)
  - Gonadal veins drain to ipsilateral renal veins
- Circumaortic left renal vein
  - 2 left renal veins
    - Superior renal vein joined by left adrenal vein and crosses anterior to aorta
    - Inferior renal vein, located 1-2 cm caudal to superior renal vein, joined by left gonadal vein and ultimately crosses posterior to aorta
- Retroaortic left renal vein
  - Solitary left renal vein that crosses posterior to aorta
- Duplication of IVC with retroaortic right renal vein and hemiazygos continuation of IVC
  - Left and right IVC inferior to renal veins
  - Right IVC drains into right renal vein, which crosses posterior to aorta to join left IVC
  - Suprarenal IVC passes posterior to diaphragmatic crus to enter thorax as hemiazygos vein
  - In thorax, collateral pathways for hemiazygos vein include these features
    - Crosses posterior to aorta at T8-T9 to join azygos vein
    - Continues superiorly to join coronary vein of heart via persistent left SVC
    - Accessory hemiazygos continuation to left brachiocephalic vein
- Duplication of IVC with retroaortic left renal vein and azygos continuation of IVC
  - Combination of findings previously mentioned
- Circumcaval ureter
  - Proximal ureter courses posterior to IVC, emerges to right of aorta, and lies anterior to right iliac vessels
- Absence of infrarenal or entire IVC
  - External and internal iliac veins join to form enlarged ascending lumbar veins
  - Venous return from lower extremities to azygos and hemiazygos vein via anterior paravertebral collateral veins
  - ± suprarenal IVC formed by left and right renal veins
  - May be acquired abnormality following thrombosis of IVC

MR Findings

- Flow voids or flow-related enhancement may distinguish aberrant vessels from masses or lymph nodes

Ultrasonographic Findings

- Infrahepatic IVC at level of renal veins
  - Suprarenal IVC continues as azygous or hemiazygos veins
- Hepatic veins drain directly into right atrium
- Right renal artery crossing anteriorly to azygos vein; may demonstrate azygos continuation of IVC

Angiographic Findings

- Inferior vena cavography: Highly accurate method

Imaging Recommendations

- Best imaging tool
  - CECT or MR with reformations in coronal plane

DIFFERENTIAL DIAGNOSIS

Retroperitoneal Lymphadenopathy

- Malignancy: Metastases, lymphoma
- Granulomatous disease
- Left-sided paraaortic adenopathy; may mimic duplication of IVC or left IVC
  - Differentiate by renal vein drainage or contrast enhancement of IVC
- Retrocrural adenopathy; may mimic enlarged azygos vein in retrocrural space
  - Differentiate by tubular structure of azygos vein extending from diaphragm to azygos arch
  - Retrocrural adenopathy enhances less than vessels
- Retroperitoneal adenopathy; may mimic circumaortic left renal vein

Varices/Collaterals

- Due to cirrhosis or IVC obstruction

Gonadal Vein

- May appear as paraaortic soft tissue "mass" or mimic left-sided IVC
- Follows inferiorly; does not "join" or replace left iliac vein

PATHOLOGY

General Features

- Etiology
  - Congenital
  - Risk factor: 1st-degree relatives
  - Pathogenesis
    - Duplication of IVC
      - Persistence of both supracardinal veins
    - Left IVC
      - Regression of right supracardinal vein with persistence of left supracardinal vein
    - Azygos continuation of IVC
      - Failure to form right subcardinal–hepatic anastomosis, resulting in atrophy of right subcardinal vein
    - Circumaortic left renal vein
      - Persistence of dorsal limb of embryonic left renal vein and of dorsal arch of renal collar
    - Retroaortic left renal vein
      - Persistence of dorsal arch of renal collar and regression of ventral arch
    - Duplication of IVC with retroaortic right renal vein and hemiazygos continuation of IVC
      - Persistence of left lumbar and thoracic supracardinal vein and left suprasubcardinal anastomosis + failure to form right subcardinal–hepatic anastomosis
    - Duplication of IVC with retroaortic left renal vein and azygos continuation of IVC
      - Persistence of left supracardinal vein and dorsal limb of renal collar with regression of ventral arch + failure to form subcardinal-hepatic anastomosis
- Embryology
  - 6th-8th gestational week
    - Infrahepatic IVC develops from appearance and regression of 3 paired embryonic veins
      - Postcardinal, subcardinal, and supracardinal veins
  - Normal IVC comprised of hepatic, suprarenal, renal, and infrarenal segments
  - Hepatic segment develops from vitelline vein
  - Suprarenal segment develops from subcardinal-hepatic anastomosis
  - Renal segment develops from right suprasubcardinal and postsubcardinal anastomoses
  - Infrarenal segment develops from right supracardinal vein
  - In thorax, supracardinal veins form to azygos and hemiazygos veins
  - In abdomen, subcardinal and supracardinal veins progressively replace postcardinal veins
  - In pelvis, postcardinal veins form common iliac veins

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most patients are asymptomatic
  - Circumcaval ureter: Partial right ureteral obstruction or recurrent urinary tract infections
  - Absence of infrarenal or entire IVC: Venous insufficiency of lower extremities or idiopathic deep venous thrombosis
- Diagnosis
  - Usually diagnosed incidentally by imaging

Demographics

- Age
  - Congenital anomalies
- Epidemiology
  - Prevalence
    - Duplication of IVC: ~ 1-3% of general population
    - Left IVC: ~ 0.2-0.5%
    - Azygos continuation of IVC: ~ 0.6%
    - Circumaortic left renal vein: ~ 2-9%
    - Retroaortic left renal vein: ~ 2-3%

Natural History & Prognosis

- Prognosis: Very good

Treatment

- Usually no treatment required
- Circumcaval ureter
  - Surgical relocation of ureter anterior to IVC

DIAGNOSTIC CHECKLIST

Consider

- Duplication of IVC should be suspected in recurrent pulmonary embolism following IVC filter placement

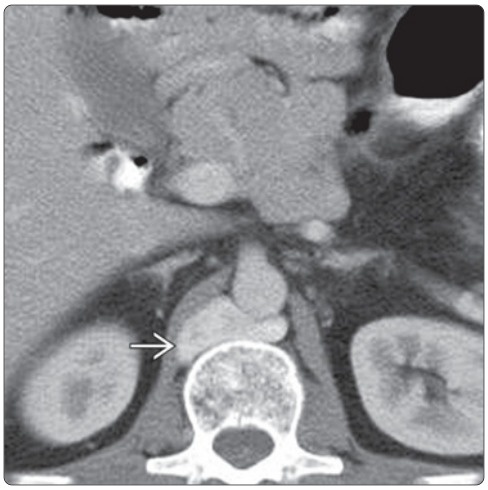
Image Interpretation Pearls

- Preoperative imaging may be important in planning abdominal surgery, liver or kidney transplantation, or interventional procedures
  - e.g., IVC filters, varicocele sclerotherapy, venous renal sampling, cardiopulmonary bypass
- Anomalies of IVC are easily confused with retroperitoneal adenopathy, especially on NECT or if veins are thrombosed

SELECTED REFERENCES

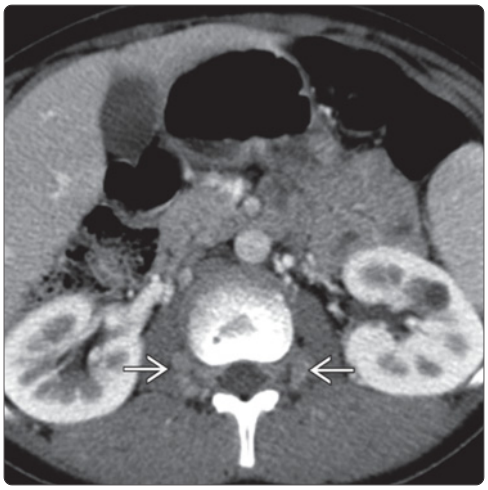
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(Left) *Axial CECT shows azygos continuation of the IVC. Note the renal veins drain into the infrahepatic IVC, which also receives tributaries from the hemiazygos vein coursing posterior to the aorta.* **(Right)** *Axial CECT of a more cephalad section in the same patient shows absence of the intrahepatic portion of the IVC. Instead, the azygous vein returns all the venous blood from the lower body to the heart via its thoracic connections. The hepatic veins drain directly into the right atrium.*





(Left) *Axial CECT more superiorly in the same shows absence of the intrahepatic portion of the IVC. Instead, blood flow from the lower body returns to the heart via the enlarged azygous vein, which eventually drains into the SVC. The hepatic veins (not shown on current image) drain directly into the right atrium.* **(Right)** *Axial CECT shows complete absence of the IVC and numerous paraspinal collaterals.*





(Left) *Post-contrast axial T1 C+ FS MR with fat suppression shows that the anterior component of a circumaortic left renal vein is located cephalad to the posterior component.* **(Right)** *Post-contrast axial T1 C+ FS MR with fat suppression in the same patient demonstrates that the posterior component (retroaortic) of the circumaortic left renal vein is located more caudally.*

Retroperitoneal Fibrosis

KEY FACTS

IMAGING

- Irregular, periaortic soft tissue mass extending from renal vessels to iliac bifurcation
  - Aorta not typically displaced from spine in primary RPF
  - Soft tissue can surround IVC and medially displace ureters
  - Soft tissue may extend to renal hila and into pelvis
- NECT: Isoattenuating to muscle
- CECT or MR: Enhancement varies with stage of disease
  - Early/active disease: Avid enhancement
  - Late/chronic disease: Minimal to no enhancement
- T1WI: Low signal intensity
- T2WI: Signal intensity varies with stage of disease
  - Early/active disease: High signal intensity
  - Late/chronic disease: Low signal intensity
- Prognosis: Decrease in T2 signal intensity, enhancement or size reflects favorable response to treatment

TOP DIFFERENTIAL DIAGNOSES

- Chronic periaortitis
- Retroperitoneal metastases and lymphoma
- Coagulopathic ("retroperitoneal") hemorrhage
- Inflammatory abdominal aortic aneurysms

PATHOLOGY

- Primary (idiopathic): 2/3 of cases
  - Manifestation of systemic autoimmune or inflammatory diseases
- Secondary: 1/3 of cases
  - Most commonly due to medications, neoplasms

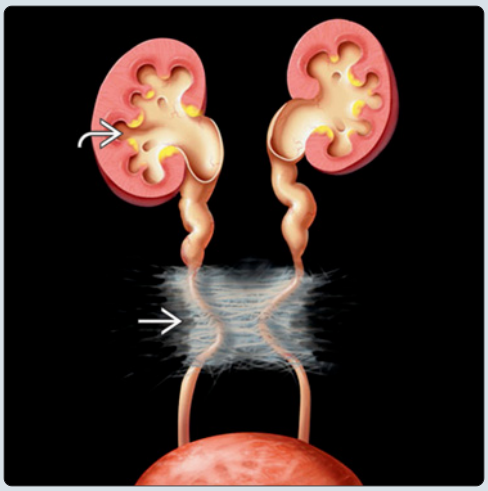
CLINICAL ISSUES

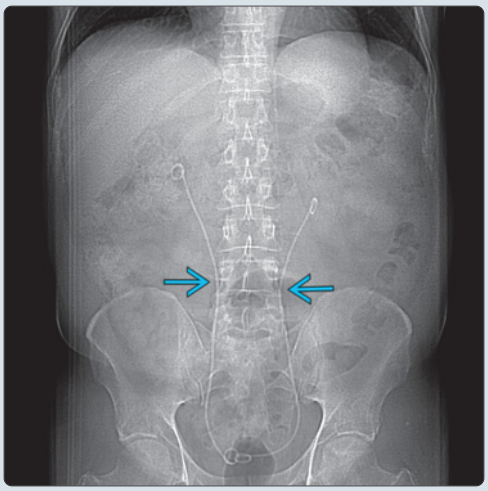
- Difficult diagnosis: Insidious, nonspecific symptoms

DIAGNOSTIC CHECKLIST

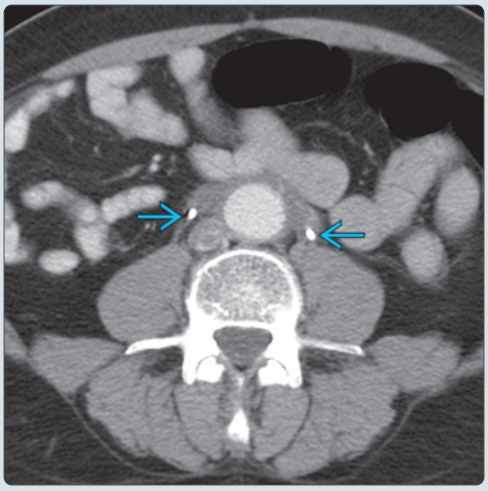
- Surgical biopsy may required for definitive diagnosis and exclusion of malignancy

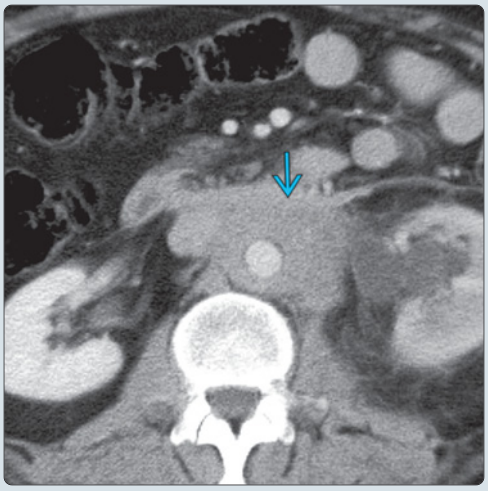
(Left) *Graphic shows encasement and medial displacement of mid ureters by a band of fibrous tissue. Note the bilateral hydroureteronephrosis. The fibrous tissue is usually limited to the inferior lumbar region.* **(Right)** *AP radiograph shows bilateral ureteral stents that are medialized in their mid portion due to RPF.*





(Left) *Axial CECT in the same patient shows stents within the medialized ureters that are surrounded by a mantle of soft tissue due to RPF. Note that the aorta is also encased by the soft tissue, but is not displaced from the aorta.* **(Right)** *Axial CECT shows an unusually massive retroperitoneal fibrosis in this patient with an obstructed left kidney.*





TERMINOLOGY

Abbreviations

- Retroperitoneal fibrosis (RPF)

Synonyms

- Ormond disease, periaortitis

Definitions

- Spectrum of diseases resulting in proliferation of fibroinflammatory tissue in infrarenal portion of retroperitoneum

IMAGING

General Features

- Best diagnostic clue
  - Irregular, periaortic soft tissue mass
- Location
  - Typically extends from renal vessels to iliac bifurcation
  - May extend to renal hila and into pelvis
- Other general features
  - 2 basic types based on etiology
    - Primary (idiopathic): 2/3 of cases
    - Secondary: 1/3 of cases
  - Classifications based on pathology and radiology
    - Malignant vs. benign
      - Malignancy: ~ 10% of cases
      - Medications: ~ 12% of cases
      - Other causes: Infection, trauma, surgery
    - Limited type (common)
      - Periaortic soft tissue limited to L4-L5 level
    - Extensive type (rare)
      - May involve mesenteric root and adjacent organs
      - Fibrosing mediastinitis: Via diaphragmatic crura
      - Pelvis: Inferior ureters, iliac vessels, pelvic viscera
  - 15% of cases associated with fibrosis elsewhere

Radiographic Findings

- Intravenous urography (IVU) triad
  - Hydroureteronephrosis (superior to L4-L5) and delayed nephrogram
  - Medial deviation of mid ureters
  - Gradual tapering of ureters, extrinsic compression
- Retrograde pyelography (RGP)
  - Hydroureteronephrosis, medial deviation of ureters
  - Valuable for assessing extent and severity of ureteral obstruction
- Fluoroscopic-guided barium enema
  - Variable degree of extrinsic compression, displacement of rectosigmoid

CT Findings

- Irregular fibroinflammatory soft tissue mass of variable thickness
  - Most commonly affects periaortic tissues between renal arteries and proximal iliac vessels
    - Soft tissue may extend from crus of diaphragm into pelvis
  - Usually envelopes aorta, IVC and ureters; ± caudal extension to iliac vessels
    - Uncommonly displaces structures anteriorly or laterally
    - May narrow, but rarely invade aorta, IVC, and ureters
    - Loss of tissue plane between fibrosis and muscles
  - Degree of contrast enhancement varies with stage of fibrosis
    - ↑ enhancement: Early/active fibroinflammatory process
    - ↓ enhancement: Chronic/inactive fibrosis
- Localized, adjacent adenopathy in 25% of primary RPF

MR Findings

- T1WI
  - Homogeneous low signal intensity (SI)
- T2WI
  - SI varies with stage of fibrosis
    - ↑ T2 SI: Early/active fibroinflammatory process
    - ↓ T2 SI: Chronic/inactive fibrosis
- T1WI C+
  - Similar to CECT: Enhancement varies with stage of fibrosis

Ultrasonographic Findings

- Grayscale ultrasound
  - Iso- to hypoechoic irregular soft tissue mass
    - Variable degrees of hydronephrosis

Nuclear Medicine Findings

- PET
  - Not useful as primary diagnostic tool due to low specificity
  - Allows detection of remote sites of disease
  - May help guide biopsy and assess treatment response

Imaging Recommendations

- Best imaging tool
  - MR or CECT with multiplanar reformations

Therapeutic Response

- Temporal decrease in size, T2WI SI, enhancement or FDG avidity reflect favorable treatment response

DIFFERENTIAL DIAGNOSIS

Aortitis

- Severe form is known as perianeurysmal fibrosis
- Presumed etiology: Hypersensitivity to antigens in atheromatous plaques
- Pathology + imaging are similar to idiopathic RPF

Retroperitoneal Metastases and Lymphoma

- Retroperitoneal metastases
  - e.g., prostate, cervix, breast, lung carcinoma
  - May cause extensive desmoplastic reaction resulting in RPF
  - Additional pelvic + retroperitoneal nodes usually seen
  - Usually more discrete or asymmetrical
  - More heterogeneous and higher in signal on T2WI
- Lymphoma
  - Typically more cephalic location in retroperitoneum
  - ↑ nodularity of mass + mass effect on vessels/viscera
  - Rarely obstructs ureters

Coagulopathic ("Retroperitoneal") Hemorrhage

- Most commonly due to over-anticoagulation
  - Hematocrit sign (cellular-fluid level), involvement of iliopsoas ± rectus sheath
- Abdominal aortic aneurysm
  - 2nd most common cause of retroperitoneal bleed
  - Acutely ill and hypotensive patient
- Spontaneous hemorrhage from neoplasm
  - Kidney: Renal cell carcinoma and angiomyolipoma
  - Adrenal: Carcinoma or myelolipoma
- CT findings
  - Acute: High-attenuation fluid/hematoma
  - Chronic: Low-attenuation fluid collection
  - Associated renal/adrenal tumors or aortic aneurysm
- MR findings
  - Varied signal intensity (evolution of blood products)
  - Hyperacute phase: Oxyhemoglobin
    - Isointense on T1WI and T2WI
  - Acute phase (1-2 days): Deoxyhemoglobin
    - T1WI: Iso- to hypointense; T2WI: Hypointense
  - Early subacute (2-7 days): Intracellular methemoglobin
    - T1WI: Hyperintense; T2WI: Hypointense
  - Late subacute (7-14 days): Extracellular methemoglobin
    - Hyperintense on on T1WI and T2WI
  - Chronic phase (> 14 days): Intracellular hemosiderin ↓ signal (T1WI); ↑ signal (T2WI)
    - Hypointense on T1WI and T2WI

PATHOLOGY

General Features

- Etiology
  - Primary (idiopathic): Probably autoimmune disease with antibodies
    - Ormond speculation: Retroperitoneal fibrosis is similar to collagen vascular disease, supported by coexistence with other inflammatory processes
  - Secondary
    - Drugs: Methysergide, β-blockers, hydralazine, ergotamine, LSD
    - Diseases that stimulate desmoplastic reaction
      - Malignant tumors, metastases, Hodgkin lymphoma, carcinoid tumor, hematoma, radiation, retroperitoneal injury, surgery, infection, urinary extravasation
- Associated abnormalities
  - Other inflammatory fibrotic processes
    - Pseudotumor of orbit; Riedel thyroiditis
    - Sclerosing cholangitis; chronic fibrosing mediastinitis
    - Autoimmune pancreatitis (IgG4-related disease)

Gross Pathologic & Surgical Features

- Mass of woody fibrous tissue; whitish-gray
- Encases vessels and ureters

Microscopic Features

- Early phase: Cellular (lymphocytes, plasma cells, fibroblasts)
- Late phase: Acellular, inactive fibrosis (dense hyalinized collagen)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific pain: Back, flank, abdomen
  - Renal insufficiency, hypertension, leg edema, anemia
- Laboratory data: No specific or definitive test
  - ↑ ESR, C-reactive protein; ↓ hematocrit; ± azotemia
  - ± rheumatoid factor, various antibodies

Demographics

- Age
  - Usually 40-65 years
- Gender
  - M:F = 1:1
- Epidemiology
  - Prevalence: 1.3:100,000 population

Natural History & Prognosis

- Complications
  - Ureteral obstruction → renal failure
  - Obstruction of great vessels (aorta and IVC)
- Prognosis
  - Good for idiopathic/benign form
  - Poor for malignant form (3-6 months survival)

Treatment

- Withdrawal of possible causative agent
- Immunosuppression: Corticosteroids
- Surgery: Ureteral stent &/or ureterolysis/transposition

DIAGNOSTIC CHECKLIST

Consider

- Surgical biopsy if atypical location or clinical suspicion for malignancy

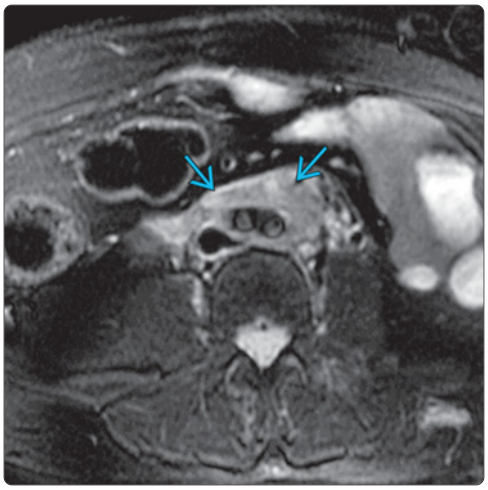
Image Interpretation Pearls

- Irregular retroperitoneal soft tissue encasing great vessels and ureters

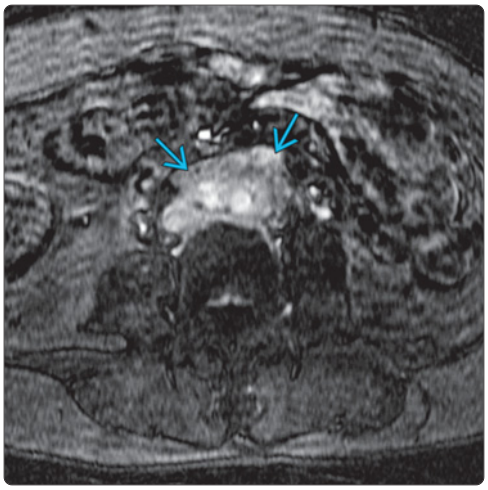
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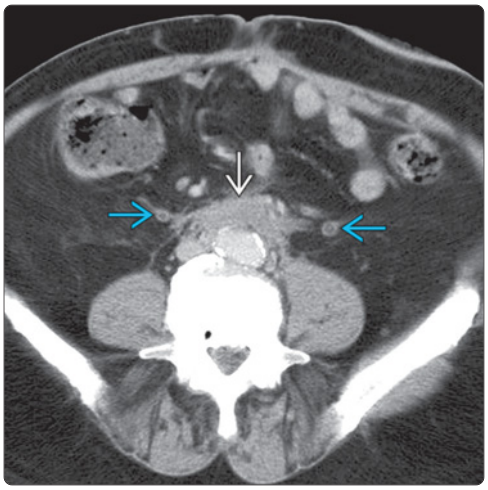
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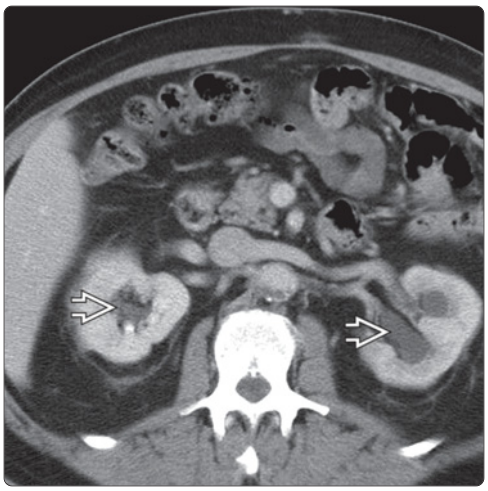


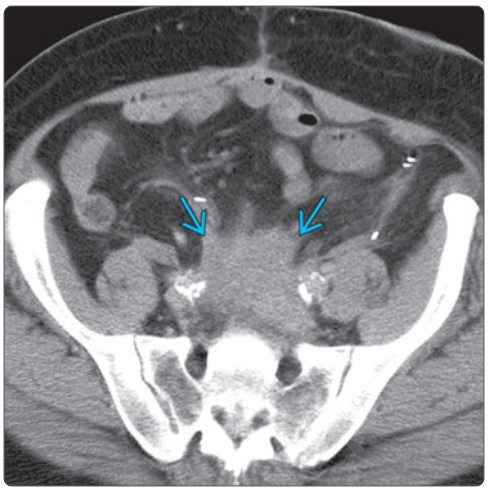
(Left) *Axial T1WI MR shows a mantle of soft tissue ⇨ surrounding the anterior and lateral aspects of the iliac bifurcation. The soft tissue has low to intermediate signal intensity.* **(Right)** *Axial T2WI MR with fat suppression in the same patient demonstrates high signal intensity of the soft tissue mass ⇨ that surrounds the iliac bifurcation, consistent with active fibrosis.*





(Left) *Post-contrast T1WI MR with fat suppression in the same patient shows avid enhancement of the soft tissue mass ⇨ surrounding the iliac bifurcation, consistent with active fibrosis.* **(Right)** *Axial CECT shows a mantle of soft tissue ➡ surrounding the anterolateral aspects of the aorta and inferior vena cava. The mantle caused medial displacement and mild dilatation of both ureters ⇨.*





(Left) *Axial CECT in the same patient reveals bilateral hydronephrosis ➡ due to the fibrotic soft tissue mass shown in the previous image.* **(Right)** *Axial CECT in the same patient shows atypical, caudal extension of the fibrotic process ⇨ into the pelvis along the iliac vessels.*

Pelvic Lipomatosis

KEY FACTS

TERMINOLOGY

- Uncommon progressive benign overgrowth of mature, unencapsulated fat in perirectal and perivesical spaces

IMAGING

- Symmetrically distributed, nonencapsulated fat surrounding pelvic organs
  - No soft tissue component or enhancement of fat
- Superior displacement of bladder, prostate, seminal vesicles, sigmoid colon
- Inverted pear- or teardrop-shaped bladder
  - Dilated, medially displaced ureters
- Elongated, narrowed rectum due to smooth, concentric extrinsic compression
  - Elongated, straight, narrowed rectosigmoid colon

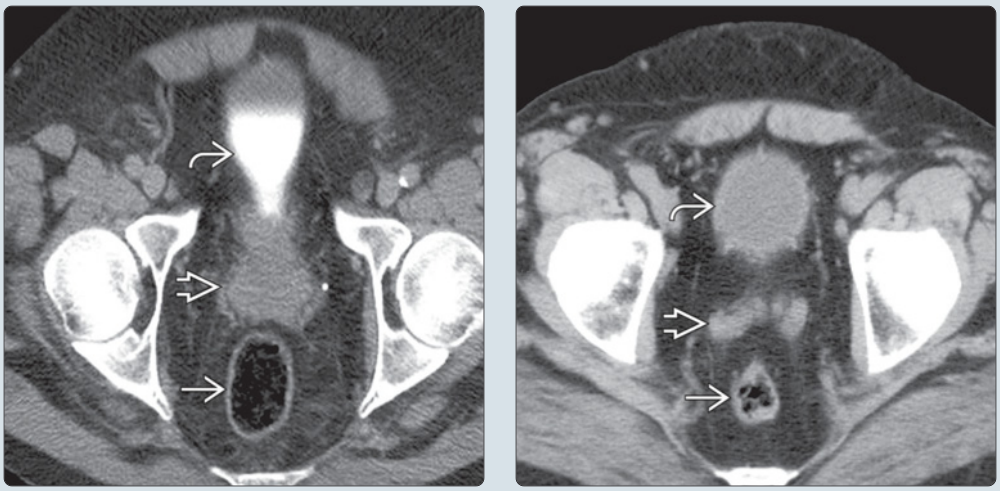
TOP DIFFERENTIAL DIAGNOSES

- Normal variant
  - Large pelvic muscles with narrow bony pelvis
- Proctitis
  - e.g., radiation, lymphogranuloma venereum
- Ulcerative colitis
  - Rectosigmoid: ↑ narrowing, thickness, symptoms
- Postoperative colon
  - e.g., proctosigmoid resection with reanastomosis

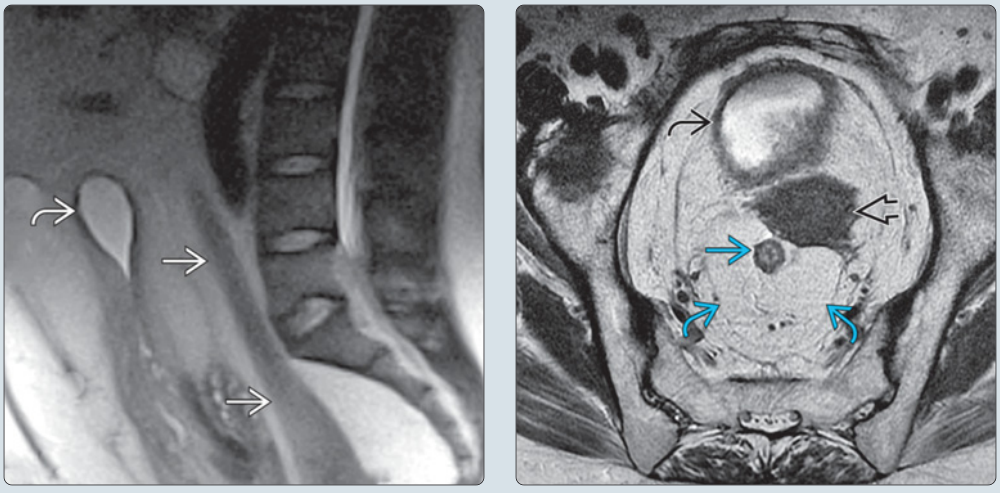
CLINICAL ISSUES

- Compressed bladder (e.g., ↑ frequency, dysuria, nocturia, and hematuria)
- Compressed rectum (e.g., constipation, rectal bleeding, tenesmus, ribbon-like stools, nausea)
- Compressed veins (e.g., leg edema, thrombosis)
- Complications: Hydronephrosis, urolithiasis, renal failure, colon obstruction, venous thrombosis, bladder adenocarcinoma
  - Frequent cystoscopy for those with proliferative cystitis
  - Radical cystoprostatectomy with urinary diversion to relieve obstruction

(Left) *Axial CECT shows the inverted pear- or teardrop-shaped appearance of the bladder. The prostate is displaced cephalad and the rectum is surrounded and separated from other viscera by the lipomatosis. No soft tissue nodule or enhancement was identified within the fat.* **(Right)** *Axial NECT in a 72-year-old man with minimal symptoms of pelvic lipomatosis shows the rectum, bladder, and seminal vesicles to be compressed and displaced from each other by the proliferation of pelvic fat.*



(Left) *Sagittal T2 MR demonstrates the inverted pear- or teardrop-shaped bladder that is displaced superoanteriorly. The rectum is compressed and elongated due to the lipomatosis.* **(Right)** *Axial T2 MR (same patient) shows proliferation of the perirectal fat, narrowing of the rectum, and superoanterior displacement of the prostate and bladder.*



TERMINOLOGY

Abbreviations

- Pelvic lipomatosis (PL)

Definitions

- Uncommon progressive overgrowth of benign, mature white fat in perirectal and perivesical spaces of pelvis

IMAGING

General Features

- Best diagnostic clue
  - Overabundance of symmetrically distributed fat in perivesical and perirectal spaces
  - Compression, elevation, and elongation of bladder and rectosigmoid colon by extensive fat

Radiographic Findings

- Radiography
  - AP view: Extensive lucent areas overlying pelvis
  - Lateral view: Increased size of presacral space

Fluoroscopic Findings

- Air enema
  - Elongated, smoothly narrowed rectum

CT Findings

- Symmetrically distributed, nonencapsulated fat surrounding pelvic organs
- Homogeneous fat attenuation (-80 to -120 HU)
  - Fat may contain occasional fibrous strands
  - No soft tissue component or enhancement of fat
- Superior displacement of bladder, prostate, seminal vesicles, sigmoid colon
- Inverted pear- or teardrop-shaped bladder
  - Medial displacement ± obstruction of ureters
- Elongated, straight, narrowed rectosigmoid colon

MR Findings

- Homogeneous signal intensity follows fat
- No enhancement or restricted diffusion

Ultrasonographic Findings

- Pear-shaped, nondistensible bladder compressed by extensive echogenic perivesical fat

Imaging Recommendations

- CECT or MR

DIFFERENTIAL DIAGNOSIS

Normal Variant

- Large iliopsoas muscles may cause pear-shaped bladder

Proctitis

- e.g., radiation, lymphogranuloma venereum

Ulcerative Colitis

- Rectosigmoid: ↑ narrowing, ↑ thickening, ↑ symptoms

Postoperative Rectosigmoid Colon

- Proctosigmoid resection with re-anastomosis
- Colon appears stretched; bladder not affected

PATHOLOGY

General Features

- Etiology
  - Unknown
- Associated abnormalities
  - Proliferative cystitis: Cystitis glandularis, cystitis cystica
    - Affects ~ 75% of patients with PL
    - Multiple filling defects in contrast-filled bladder
    - Premalignant → adenocarcinoma

Gross Pathologic & Surgical Features

- Fat: Mature, whitish-yellow, lobular, greasy

Microscopic Features

- Metaplasia: Mature cells, thin septa, lobular growth

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Compressed bladder: ↑ frequency, dysuria, nocturia, and hematuria)
  - Compressed rectum: Constipation, rectal bleeding, tenesmus, ribbon-like stools
  - Compressed veins: Leg, scrotal edema

Demographics

- Age
  - Mean: 48 years; peak: 25-60 years
- Gender
  - M:F = 18:1
- Ethnicity
  - Predilection toward African Americans (~ 67%)
- Epidemiology
  - Rare: Incidence of 1.7/100,000

Natural History & Prognosis

- Complications: Hydronephrosis, urolithiasis, renal failure, colon obstruction, venous thrombosis, proliferative cystitis

Treatment

- Ureteral stenting or surgical diversion for obstruction

DIAGNOSTIC CHECKLIST

Consider

- Monitor renal function, proliferative cystitis

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Coagulopathic (Retroperitoneal) Hemorrhage

KEY FACTS

TERMINOLOGY

- Can be misnomer: Retroperitoneal hemorrhage often only in posterior abdominal wall musculature

IMAGING

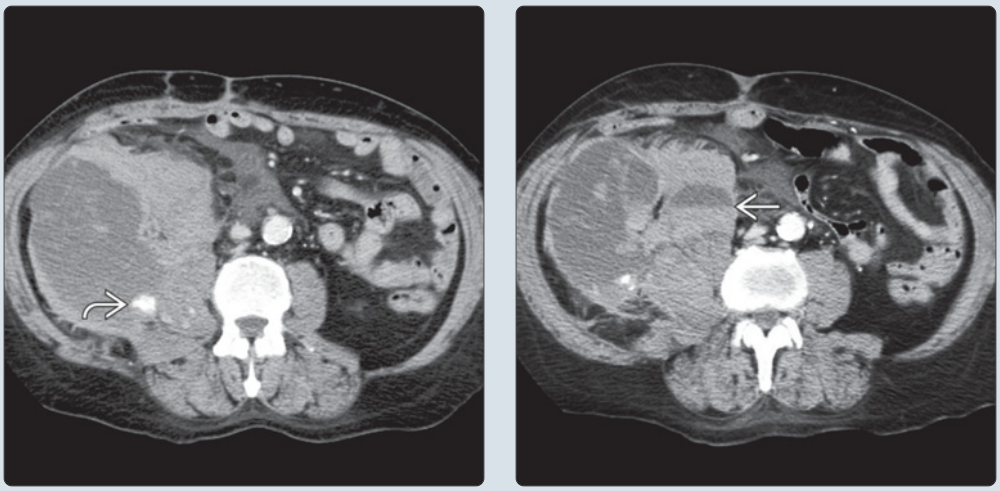
- Major causes and CT findings
  - Coagulopathy or anticoagulation; high-density collection in retroperitoneum or body wall, with cellular-fluid level (hematocrit sign)
  - Ruptured abdominal aortic aneurysm; large eccentric aneurysm, with blood ± active extravasation contiguous with aorta
  - Renal tumors
  - Trauma
  - Vasculitis
- CT appearance of hemorrhage: Heterogeneous
  - Active: Linear, flame-like shape, isodense to vessels
  - Sentinel clot sign: High attenuation (60-80 HU)
  - Initially accumulates near site of bleeding
  - Chronic hematoma: Lower density (20-40 HU)
- MR: Variable signal due to phase of blood products
  - Hyperacute: T1WI and T2WI: Isointense
  - Acute: T1WI: Iso/hypointense; T2: Hypointense
  - Early subacute: T1WI: hyperintense; T2: hypointense
  - Late subacute: T1WI and T2WI: Hyperintense
  - Chronic: T1WI + T2WI: Hypointense rim
    - Center: T1WI: Isointense; T2WI: Hyperintense
- US: Variable echogenicity due to age of hematoma
  - Acute/subacute: Echogenic

TOP DIFFERENTIAL DIAGNOSES

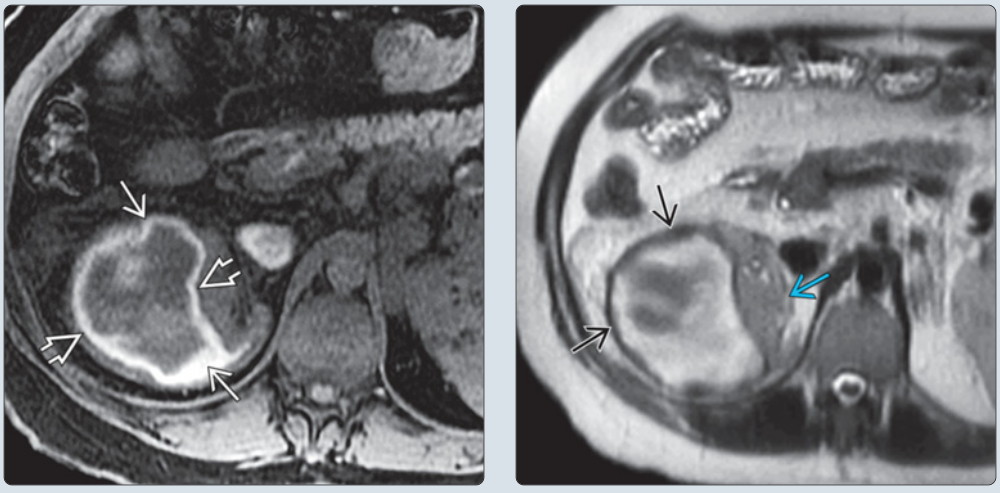
- Retroperitoneal abscess
- Retroperitoneal sarcoma
- Asymmetrical muscles

DIAGNOSTIC CHECKLIST

- Spontaneous perirenal hemorrhage: Consider underlying tumor, vasculitis, or coagulopathy



(Left) *Axial CECT shows a large, heterogeneous, "retroperitoneal" mass centered in the iliopsoas compartment in a patient with coagulopathic hemorrhage. Active extravasation of contrast is usually venous in this setting and does not typically require embolization.* **(Right)** *Axial CECT in the same patient shows the presence of the hematocrit sign, which is consistent with an acute, coagulopathic hemorrhage.*



(Left) *Axial T1WI MR with fat saturation shows a heterogeneous mass in the right perinephric space. Note there is a thin, low signal intensity outer rim surrounding a high signal intensity inner rim, both of which surround an intermediate signal intensity center. Findings are consistent with a subacute hematoma.* **(Right)** *Axial T2WI MR in the same patient better demonstrates a low signal intensity outer rim. Note that the right kidney is compressed and displaced medially.*

TERMINOLOGY

Definitions

- Hemorrhage occurring in retroperitoneal compartments due to various etiologies
- Term retroperitoneal hemorrhage may sometimes be a misnomer
  - Many cases occur primarily in posterior abdominal wall musculature

IMAGING

General Features

- Best diagnostic clue
  - Coagulopathic hemorrhage
    - High-attenuation collection in retroperitoneum or body wall with cellular-fluid level (hematocrit sign)
  - Ruptured abdominal aortic aneurysm (AAA)
    - Large eccentric aneurysm, with hematoma ± active extravasation contiguous with aorta
  - Trauma
    - Traumatic hemorrhage often confined to retroperitoneal interfascial planes
    - Often associated with renal injury

Major Causes of Bleeding and Relevant CT Findings

- **Coagulopathic hemorrhage**
  - Hematocrit sign
    - Cellular-fluid level within collection
    - Highly sensitive and specific for coagulopathy
  - Classically originates in posterior pararenal space
  - May affect several spaces (e.g., body wall, muscles)
  - Bleeding out of proportion to injury
- **Abdominal aortic aneurysm**
  - CT signs of high risk of impending rupture
    - Large aneurysm (> 5 cm, especially if recently increased)
    - Discontinuity of circumferential mural calcifications
    - Hyperattenuating crescent sign within mural thrombus
    - Draped aorta sign (indistinct, eccentric posterior aortic wall)
  - CT signs of ruptured AAA
    - Hematoma contiguous with long segment of aorta (> 3 cm)
      - May dissect through retroperitoneum or into peritoneum
    - Active extravasation from aortic lumen
    - Focal aortic contour deformity may suggest contained rupture
- **Renal tumors**
  - Heterogeneous mass replacing part of kidney
  - Renal cell carcinoma
    - Most common neoplastic cause of retroperitoneal hemorrhage
    - Typically heterogeneous, hypervascular, exophytic mass
  - Angiomyolipoma
    - Contains visible fat and soft tissue components in most cases
    - Adrenal tumors may be obscured by hemorrhage or distorted anatomy
  - Adrenal carcinoma, pheochromocytoma, or metastases
- **Trauma**
  - Hemorrhage classically affecting retroperitoneal interfascial planes
    - Large hematomas may spread to multiple compartments
  - Often associated with renal parenchymal or vascular injury
  - Iatrogenic causes include biopsy, ablation, and lithotripsy
- **Vasculitis**
  - Consider when protean symptoms or recurrent abdominal pain in young patient
  - Commonly affects kidneys and bowel
  - Kidneys: Wedge or striated nephrogram ± subcapsular or perirenal hemorrhage
  - Bowel: Long segment of small bowel with mural thickening or intramural hemorrhage

CT Appearance of Blood

- **Active bleeding**
  - Extravasation of vascular contrast (~ 150-400 HU)
  - Linear, flame-like, or amorphous appearance that is isodense to enhanced vessels
- **Acute**
  - "Sentinel clot" sign
    - Heterogeneous, high attenuation (60-80 HU)
    - Acute clot accumulates 1st near site of bleeding
    - Adjacent spaces typically have lower attenuation, more homogeneous hematoma
- **Chronic, lower density (10-40 HU)**
  - Due to lysis and evolution of clot
- **Acute and chronic**
  - Mixed-attenuation collections
- Hemorrhage can arise within, or spread into, almost any abdominal space
  - e.g., intraperitoneal, perirenal, pelvic
- ± mass effect: Displacement of kidney anteriorly &/or toward midline

MR Findings

- Varied signal intensity depending on phase of blood products
- Hyperacute (first 24 hours: Intracellular oxyhemoglobin)
  - T1WI and T2WI: Isointense
- Acute (~ 1-2 days: Intracellular deoxyhemoglobin)
  - T1WI: Iso to slightly hypointense
  - T2WI: Hypointense
- Early subacute (~ 2-7 days: Intracellular methemoglobin)
  - T1WI: Hyperintense
  - T2WI: Hypointense
- Late subacute (~ 7-28 days: Extracellular methemoglobin)
  - T1WI + T2WI: Hyperintense
- Chronic (> 14-28 days)
  - Periphery (intracellular hemosiderin): T1WI + T2WI: Hypointense
  - Center: T1WI → isointense; T2WI: Hyperintense

Ultrasonographic Findings

- Echogenicity varies with phase of blood products

- Acute/subacute: Echogenic; mass-like
- Chronic: Hypo- to anechoic fluid collection

Imaging Recommendations

- Best imaging tool
  - CT ± IV contrast; contrast may identify active hemorrhage
- Protocol advice
  - CECT
    - Power injection (≥ 3 mL/second; ~ 70 second delay)
    - Diaphragm to symphysis using < 5 mm collimation
      - Consider multiplanar reformations

DIFFERENTIAL DIAGNOSIS

Retroperitoneal Abscess

- Less likely to extend far from site of origin vs. intraperitoneal abscesses
- Perforation of retroperitoneal portion of bowel or colon
  - e.g., appendicitis, diverticulitis, duodenal ulcer, trauma
- CECT: Low-attenuation contents with enhancing rim ± gas

Retroperitoneal Sarcoma

- CT or MR
  - Usually located in peri-/pararenal spaces
  - Large (> 10 cm) heterogeneous mass
    - May contain fat, soft tissue attenuation ± calcification
  - Poorly or sharply marginated and encapsulated mass
  - Displacement, compression, distortion of adjacent structures (kidneys, bowel, colon)

Asymmetrical Muscles

- Posterior abdominal wall
  - e.g., polio or amputation
- CT: Asymmetrical, enlarged iliopsoas muscle may be mistaken for hemorrhage within muscle

PATHOLOGY

General Features

- Etiology
  - Iatrogenic (most common)
    - Medications: Warfarin, heparin, antiplatelet medications
    - Femoral vein cannulization or arteriography
    - Percutaneous nephrostomy, renal biopsy, extracorporeal shock wave lithotripsy, tumor ablation
    - Translumbar aortography
  - Aortic aneurysm and other arterial aneurysms
    - Abdominal aortic aneurysm (2nd most common)
    - Iliac artery aneurysm
    - Renal, hepatic, splenic artery aneurysm
    - Inferior pancreaticoduodenal artery aneurysm
    - Ovarian or uterine artery aneurysm
  - Visceral causes (3rd most common)
    - Renal or adrenal tumors
      - Renal cell carcinoma, angiomyolipoma, cyst
      - Adrenal carcinoma, pheochromocytoma, myelolipoma, metastasis
  - Trauma
    - Abdominal wall, renal or adrenal trauma
    - Posterosuperior region of segment 7 (liver bare area)
  - Other diseases
    - Vasculitis (e.g., polyarteritis or Wegener vasculitis)
    - Pancreatitis (hemorrhagic pancreatitis or ruptured pseudoaneurysm)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Abdominal, back or flank pain; radiating to groin/thigh
  - Hypotension, shock, nausea, vomiting
  - Abdominal distention, peritoneal signs
  - Weakness and ↓ sensation in lower extremities

Demographics

- Age
  - Any; increased likelihood with older age
- Gender
  - M > F

Natural History & Prognosis

- Complications: Paresis, shock, and death
- Prognosis: Poor, but dependent on cause

Treatment

- Stop medication ± protamine for coagulopathic hemorrhage
- Infusion of blood products (i.e., plasma, red blood cells)
- Angiographic embolization for active bleeding
- Surgery (e.g., ruptured aneurysm, tumor, organ injury)

DIAGNOSTIC CHECKLIST

Consider

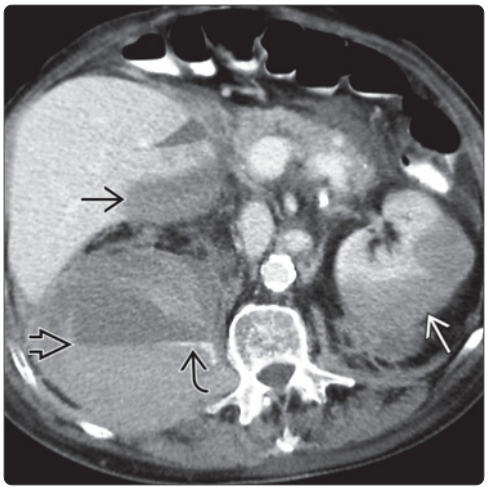
- Most common cause is iatrogenic

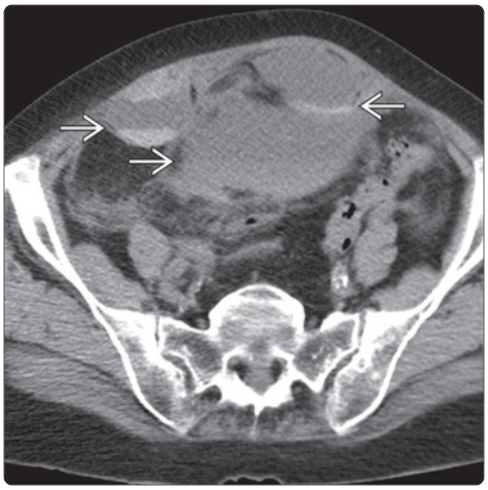
Image Interpretation Pearls

- Hematocrit sign and bleeding into several spaces indicates anticoagulation
- Spontaneous perirenal hemorrhage: Consider underlying tumor, vasculitis or coagulopathy
  - Repeat imaging after resolution to detect occult lesion

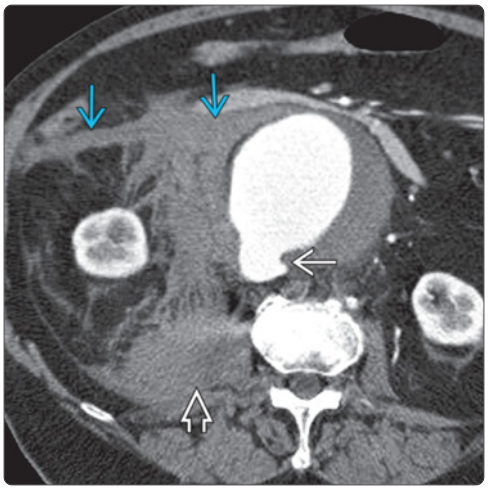
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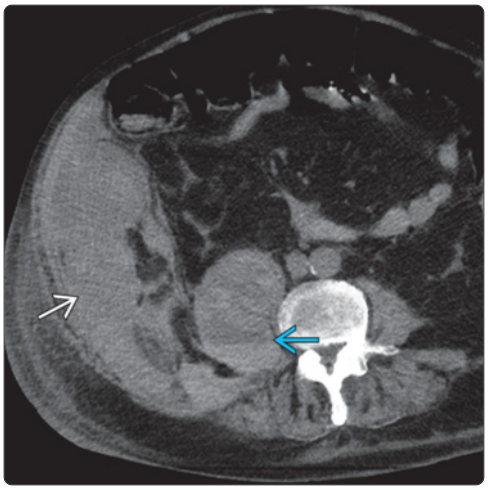
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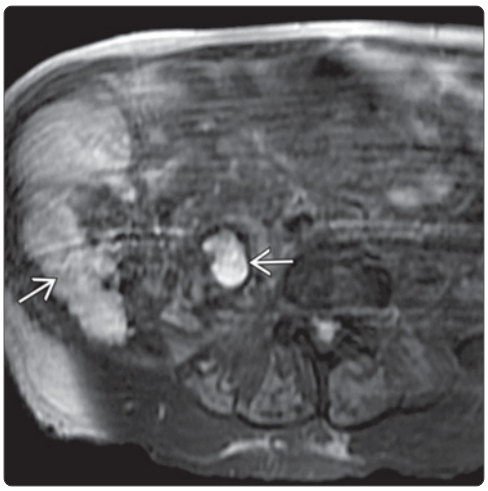
(Left) *Axial CECT shows classic findings of coagulopathic hemorrhage at multiple sites, including the left perirenal ➡, right iliopsoas and intraperitoneal ⇨ spaces. Note the hematocrit sign ⇨ and active (venous) bleeding ⇨ in the right iliopsoas muscle. Active extravasation from coagulopathy was due to venous bleeding and stopped with reversal of the anticoagulation.* **(Right)** *Axial NECT shows enlargement of both rectus sheaths and hematocrit signs ➡ indicating acute hemorrhage due coagulopathy.*





(Left) *Axial CECT shows a large aneurysm extending along the aorta and recent rupture into the right retroperitoneum ⇨ and iliopsoas muscle ➡. The aorta is "draped" over the spine and there is a mural contour defect ➡ along the dorsal aspect of the aortic lumen at the site of extravasation.* **(Right)** *Axial NECT show acute hematoma in the right retroperitoneum ➡ and a hematocrit sign ⇨ in the psoas muscle due to coagulopathic hemorrhage.*





(Left) *Axial T1WI MR in the same patient (16 days after the initial NECT) shows a reduction in size and high signal intensity of the retroperitoneal hematoma ➡ due to the late subacute phase of the blood products.* **(Right)** *Axial T2WI MR in the same patient (16 days after the initial NECT) shows a reduction in size and high signal intensity of the retroperitoneal hematoma ➡ due to the late subacute phase of the blood products.*

Postoperative Lymphocele

KEY FACTS

IMAGING

- Well-marginated, simple cystic lesion with thin wall in extraperitoneal pelvis or retroperitoneum of postoperative patient
- Commonly located along iliac vessels, inguinal region, and paraaortic retroperitoneum
- CECT: Typically unilocular but may contain few, thin septa; no mural nodule or enhancement
  - Mural calcification and internal fat are rare findings
- MR: Homogeneous low SI on T1WI; high SI on T2WI; no contrast enhancement or restricted diffusion
  - MR lymphangiography: Allows specific diagnosis of lymphocele
- US: Marginated, anechoic lesion with through transmission; occasional septa
  - May have internal echoes or layering debris if infected

TOP DIFFERENTIAL DIAGNOSES

- Other pelvic cystic masses
  - Urinoma
  - Bladder diverticulum
  - Lymphangioma

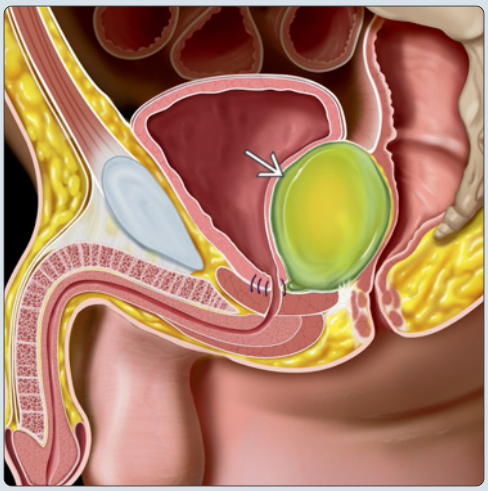
PATHOLOGY

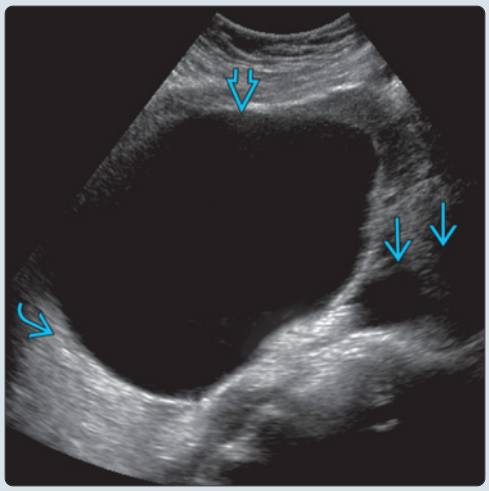
- Occurs in up to ~ 30% of patients undergoing radical pelvic surgery

CLINICAL ISSUES

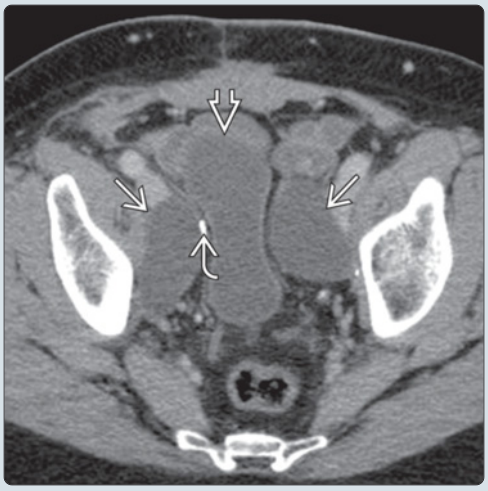
- Most are small, asymptomatic, and resolve spontaneously
- Large lymphoceles may be symptomatic due to mass effect
- Lymphadenectomy is most significant risk factor
- Presentation varies weeks to months after surgery
- Treatment: Drainage, sclerotherapy, or surgery

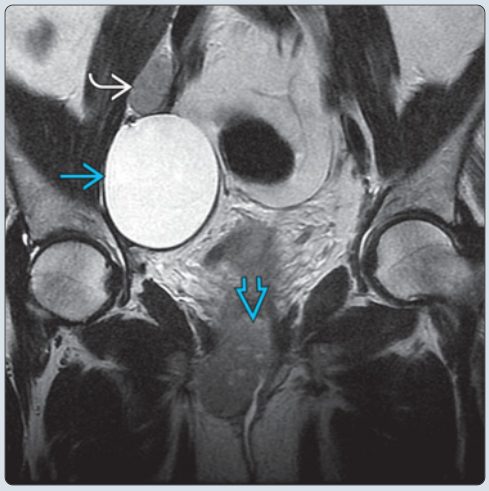
(Left) *Graphic shows a male patient who has had a radical prostatectomy, which includes extensive pelvic node dissection. A lymphocele has developed within the pelvis and exerts mass effect on the bladder and rectum.* **(Right)** *Transverse endovaginal US in a patient who has undergone hysterectomy and node dissection shows a large, anechoic cystic lesion adjacent to the iliac vessels, representing a postoperative lymphocele. Note the thin wall and through transmission.*





(Left) *Axial CECT post prostatectomy shows bladder compression by bilateral lymphoceles. Note the surgical clip from node dissection.* **(Right)** *Coronal SS FSE T2W MR post hysterectomy and node dissection due to cervical cancer reveals that a sharply marginated lymphocele has high T2 signal intensity. Note local tumor recurrence adjacent to the urethra and a metastatic lymph node.*





TERMINOLOGY

Synonyms

- Lymphatic cyst or pseudocyst; lymphocyst

Definitions

- Lymph-filled cystic space that lacks epithelial lining

IMAGING

General Features

- Best diagnostic clue
  - Well-defined, cystic lesion in extraperitoneal space of patient who has undergone pelvic surgery
- Location
  - Extraperitoneal pelvis; paraaortic retroperitoneum
- Morphology
  - Typically unilocular with thin wall; ovoid

CT Findings

- NECT
  - Near water attenuation cyst when uncomplicated
  - Small amount of internal fat (chyle) is rare finding
  - Rare, mural calcification when chronic
- CECT
  - Thin wall; no mural enhancement or nodule

MR Findings

- Homogeneous low signal intensity (SI) on T1WI; high SI on T2WI
  - T2WI with fat suppression: No loss of high SI
  - DWI: Low SI
- No enhancement following IV contrast
  - MR lymphangiography: Allows specific diagnosis

Ultrasonographic Findings

- Grayscale ultrasound
  - Anechoic with through transmission; occasional septa
  - Internal echoes/layering debris if complicated
- Color Doppler
  - No flow

Imaging Recommendations

- Protocol advice
  - CECT or MR: Excretory imaging (> 10 minutes)

DIFFERENTIAL DIAGNOSIS

Pelvic Cystic Lesions

- Urinoma: May fill with contrast after excretory phase
- Abscess: Enhancing thick, irregular wall
- Hematoma/endometrioma: Complex, blood products

Bladder Diverticulum

- Abuts bladder; ± identification of thin neck to bladder
- Opacification after excretory phase imaging

Lymphangioma

- Multiloculated mass with thin septa; ± calcification
- Often involves > 1 retroperitoneal compartment

Pseudomyxoma Retroperitonei

- Multicystic, thick walls, calcifications; right lower quadrant

PATHOLOGY

General Features

- Radical pelvic surgeries (hysterectomy, prostatectomy)
  - Lymphadenectomy in ~ 30%; renal transplant in ~ 18%

Gross Pathologic & Surgical Features

- Cystic lesion with thin, tan to yellowish-brown fluid

Microscopic Features

- Lymphocytes, few red blood cells, variable protein/fat

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Minority (< 10%) of patients are symptomatic
  - Distention, pain, infection, mass effect
  - Renal transplant compression
    - Vascular: Altered hemodynamics; unilateral leg edema
    - Ureteral: Elevated creatinine, hydronephrosis

Demographics

- Any age or gender

Treatment

- Options, risks, complications
  - Drainage or sclerotherapy: Variable success (~ 50-90%)
  - Laparoscopic marsupialization or surgical fenestration

DIAGNOSTIC CHECKLIST

Consider

- Review images/records for radical surgery involving nodal dissection in pelvis or retroperitoneum

Image Interpretation Pearls

- Cystic lesion near surgical clips following node dissection
- Mural thickening and enhancement suggest superinfection

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Retroperitoneal Neurogenic Tumor

KEY FACTS

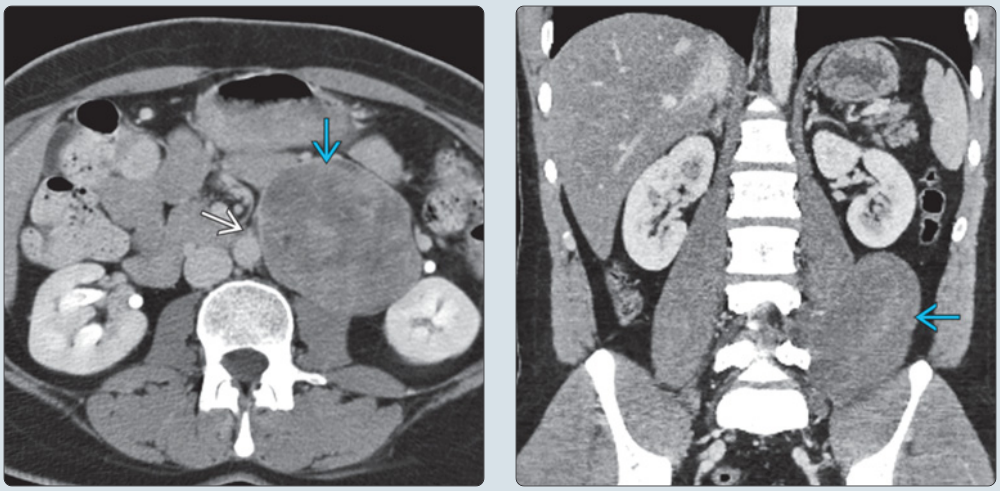
TOP DIFFERENTIAL DIAGNOSES

- Ganglioneuroma
  - Rare, benign tumor of sympathetic ganglia or adrenal
  - Well circumscribed; oval or lobulated
  - Excellent prognosis with rare recurrence
- Ganglioneuroblastoma
  - Malignant sympathetic tumor affecting 2-4 year olds
  - Heterogeneous mass: Ranges from solid to cystic
- Neuroblastoma
  - Malignant tumor of sympathetic nerves or adrenal
  - Unencapsulated, irregular, lobulated
  - Necrosis, hemorrhage, coarse calcifications
  - Invades adjacent structures + metastases
- Paraganglioma ("extraadrenal pheochromocytoma")
  - Benign or malignant; hormonally active in ~ 40%
  - Very avid enhancement due to hypervascularity
  - T2W: "Light bulb bright" in ~ 80%
  - Usually syndromic (e.g., von Hippel-Lindau) and benign
- Schwannoma (neurilemmoma)
  - Usually benign tumor of peripheral nerve sheaths
  - Usually paravertebral, but can arise in any nerve
  - Typically < 5 cm, round, homogeneous
    - ↑ size: ± cystic degeneration, calcifications
- Neurofibroma
  - Typically solitary, benign nerve sheath tumor
    - ~ 10% are multiple, often due to NF1
  - Well marginated, round, homogeneous
  - Malignant degeneration → neurofibrosarcoma
    - ~ 2-20% in those with NF1
    - Large, heterogeneous, infiltrative mass

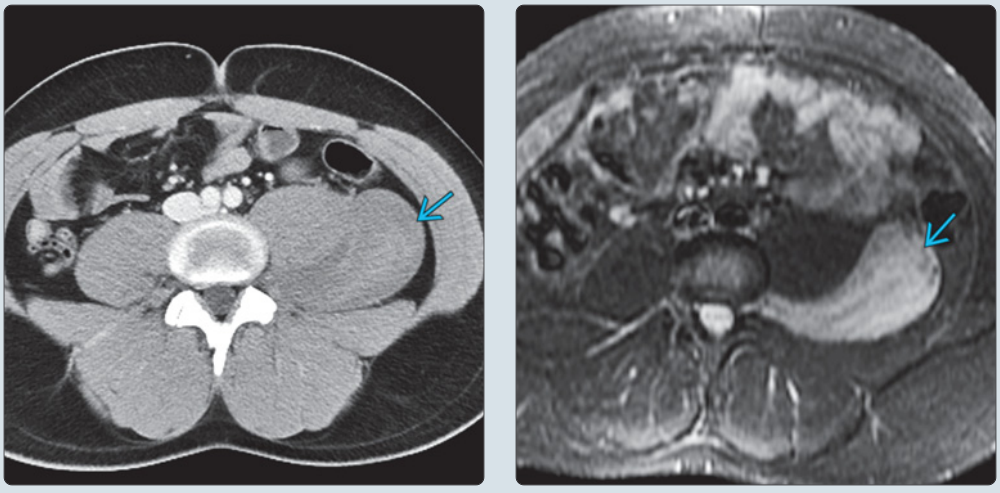
DIAGNOSTIC CHECKLIST

- Review history for syndromes (NF1 and von Hippel-Lindau)
- Most common scenario for neurogenic tumor
  - Child to young adult
  - Solid, enhancing, retroperitoneal mass
  - Mass neither contains fat nor arises from viscera

(Left) *Axial CECT reveals a large, heterogeneously enhancing mass in the left aspect of the retroperitoneum at the level of the origin of the inferior mesenteric artery. Note that the mass abuts and mildly displaces adjacent viscera, but does not arise from them. Pathology was consistent with a paraganglioma.* **(Right)** *Coronal CECT reveals a large, heterogeneously enhancing ovoid mass arising between the spine and left psoas muscle.*



(Left) *Axial CECT in the same patient demonstrates the heterogeneously enhancing mass posterior to the left psoas muscle and contiguous with the adjacent neural foramen.* **(Right)** *Axial T2WI MR with fat suppression in the same patient reveals slightly heterogeneous, high signal intensity within the left retroperitoneal mass. Surgical pathology was consistent with benign schwannoma.*



TERMINOLOGY

Definitions

- Primary retroperitoneal tumors arising from ganglion cells, parasympathetic system, or nerve sheaths

Overview

- Neurogenic tumors: ~ 10% of primary retroperitoneal tumors
- Neurogenic tumors can arise from
  - Ganglionic cells
  - Paraganglionic cells
  - Nerve sheath
- Neurogenic tumors affect younger population and have better prognosis compared to retroperitoneal mesenchymal tumors

DIFFERENTIAL DIAGNOSIS

Ganglioneuroma

- Rare, benign tumor arising from sympathetic ganglia
- Location
  - Retroperitoneum > mediastinum > cervical region
    - In retroperitoneum: Paravertebral sympathetic ganglia > adrenal medulla
- NECT
  - Well circumscribed; oval or lobulated mass
  - Homogeneous, hypoattenuating compared to muscle
  - Punctate calcifications in ~ 20-30%
- CECT
  - Variable, heterogeneous enhancement
  - May surround vessel without narrowing lumen
  - Rare hemorrhage or necrosis
- MR
  - T1WI: Homogeneously ↓ SI
  - T2WI: Variable SI, depending on amounts of myxoid, collagen, cellular components
    - ↑ T2 SI (due to myxoid matrix)
    - Bands of ↓T2 SI = bundles of Schwann cells
      - Results in whorled appearance
- Pathology
  - Composed of Schwann cells, ganglion cells, nerve fibers
- Demographics
  - No sex predilection; 3rd-4th decades
- Clinical manifestations
  - Asymptomatic > abdominal pain or mass
  - Hormonally active form is rare
    - Symptoms: Diarrhea, hypertension, virilization
- Prognosis
  - Excellent; extremely rare recurrence after resection

Ganglioneuroblastoma

- Rare tumor of sympathetic origin
- Location
  - Paravertebral
- CT or MR
  - Heterogeneous: Solid, cystic, or combination
    - Variable enhancement
  - Encapsulated mass with granular calcification
- Pathology
  - Malignant tumor containing primitive neuroblasts and mature ganglion cells
  - Intermediate grade containing mixed malignant and benign components
- More favorable prognosis than neuroblastoma
- Demographics
  - No sex predilection; 2-4 years old

Neuroblastoma

- Malignant tumor of sympathetic origin
- Location
  - Adrenal (~ 66%) > paravertebral sympathetic chain (~ 33%)
- CT or MR
  - Irregular margins, lobulated, unencapsulated
  - Heterogeneous (due to necrosis + hemorrhage)
  - Coarse, amorphous calcifications in ~ 80-90%
  - Invades adjacent structures ± distant metastases
  - Encases vessels and attenuates lumen
- Pathology
  - Primitive neuroblasts ± glial or ganglionic differentiation
- Natural history
  - Tends to metastasize to bone, liver, and skin
  - 70% have disseminated disease at time of diagnosis
- Demographics
  - M > F; 1st decade
- Nuclear scintigraphy
  - Meta-iodobenzylguanidine (MIBG)
    - High sensitivity for localization

Paraganglioma (Extraadrenal Pheochromocytoma)

- Tumor of neural crest origin (benign > malignant) occurring in parasympathetic ganglia
  - Associated syndromes: von Hippel-Lindau, multiple endocrine neoplasia (MEN), neurofibromatosis type 1 (NF1)
  - ~ 40% secrete catecholamines
- Location
  - In retroperitoneum: Most commonly at organ of Zuckerkandl
    - Near origin of inferior mesenteric artery
- CECT
  - Large, well defined, round or lobulated
  - Heterogeneous: Necrosis, hemorrhage, calcification
  - Avid enhancement due to hypervascularity
- MR
  - T1WI: Generally ↓ SI
    - ↑ SI in areas of hemorrhage
    - Signal voids on spin-echo sequences
  - T2WI: ↑ SI, but heterogeneous
    - "Light bulb bright" is rare due to complexity of tumor
- Nuclear scintigraphy
  - FDG PET (fluorodeoxyglucose)
  - Meta-iodobenzylguanidine (MIBG)
    - Typically strong uptake in paragangliomas
      - High sensitivity for localization
- Pathology
  - Benign vs. malignant tumors have identical histology
  - Clinical behavior best predicts prognosis
- Demographics

Classification & Types of Neurogenic Tumors

Neurogenic Origin	Benign Tumor	Malignant Tumor
Ganglionic cells (sympathetic)	Ganglioneuroma, ganglioneuroblastoma	Neuroblastoma
Paraganglionic cells (chromaffin)	Paraganglioma, pheochromocytoma	Malignant paraganglioma or pheochromocytoma
Nerve sheath	Schwannoma, neurofibroma	Malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma

- No sex predilection; 3rd-4th decades
- Clinical manifestations
  - ~ 40% hormonally active
    - Headache, palpitations, sweating, ↑ urine metanephrine or vanillylmandelic acid levels
  - Rupture & hemorrhage can occur in large tumors

Schwannoma (Neurilemmoma)

- Usually benign tumor of peripheral nerve sheaths
- Location: Paravertebral > perirenal > presacral
  - However, can affect any nerve
    - May mimic solid organ mass (i.e., pancreas)
- NECT
  - Usually < 5 cm; well circumscribed, round, homogeneous
  - ↑ size: Ovoid, ± cystic degeneration, calcifications
    - Calcifications: Punctate, mottled, curvilinear
- CECT
  - Enhancement: varies by cystic vs. solid components
  - Nerve of origin can be difficult to identify at imaging
- Demographics
  - F > M; 3rd-6th decades
- Clinical manifestations
  - Asymptomatic > abdominal mass or pain
- Pathology
  - Encapsulated tumor extending along course of nerve
  - Malignant transformation is rare
  - Ancient schwannoma: Hemorrhage, cysts, hyaline
- MR
  - T1WI: Typically ↓ SI
  - T2WI: Variable due to size and amount of cystic/solid components
    - ↑ SI in cystic components
    - ↓ SI on densely cellular regions

Neurofibroma

- Typically benign nerve sheath tumor
  - Solitary (90%) > multiple (10%)
    - ~ 30% solitary and 100% multiple neurofibromas are associated with neurofibromatosis (NF1)
- NECT
  - Well circumscribed, round, homogeneous
  - Hypoattenuating: Lipid-rich Schwann cells + adipocytes
    - Typically ~ 20-25 HU
  - May widen neural foramina and have dumbbell shape
    - May cause scalloping of vertebral body
- CECT
  - Homogeneous, low-grade (~ 30-50 HU) enhancement
    - Small cystic areas rarely seen
  - Occasional "target-like" enhancement
  - Malignant: Large, heterogeneous, infiltrative
- MR
  - T1WI: Relatively higher central SI due to neural tissue
    - ↓ SI peripherally
  - T2WI: ↑ SI peripherally due to myxoid degeneration
    - ↓ SI centrally due to highly cellular neural tissue
- Pathology
  - Solid, unencapsulated tumor that expands nerve
  - Comprised of collagen bundles and nerve sheath cells
  - Malignant type transformation → neurofibrosarcoma
    - ~ 2-20% of those with NF1; rare in solitary tumors
- Demographics
  - M > F; 3rd-5th decades

Malignant Nerve Sheath Tumors

- Types
  - Malignant schwannoma, neurogenic sarcoma, neurofibrosarcoma
- Etiology
  - ~ 50% arise spontaneously
  - ~ 50% due to transformation of neurofibroma, ganglioneuroma, or due to radiation exposure
- Demographics
  - No sex predilection; 3rd-6th decades
- CT or MR
  - Enlarging, heterogeneous, ill-defined, infiltrative mass
- Clinical manifestations
  - Pain, enlarging palpable mass

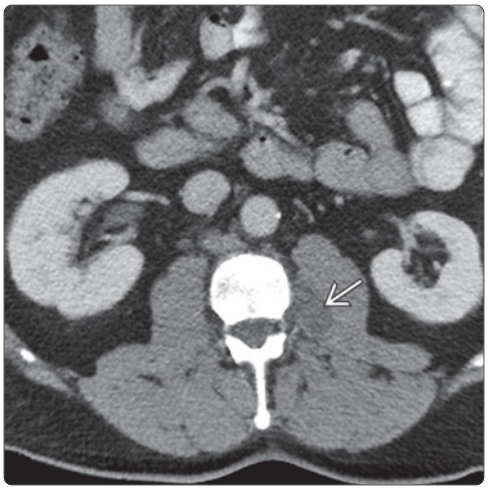
DIAGNOSTIC CHECKLIST

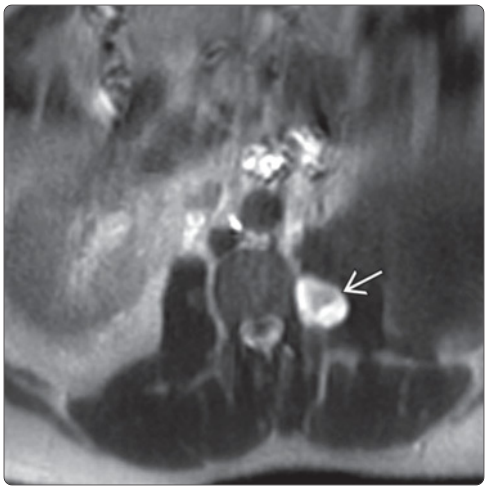
Image Interpretation Pearls

- Consider neurogenic tumor for enhancing retroperitoneal mass in child or young adult that neither arises from organ nor contains fat
- Extension along other neural paths may cause masses in pancreas, porta hepatis, etc.
- Review history for syndromes (NF1 and von Hippel-Lindau)

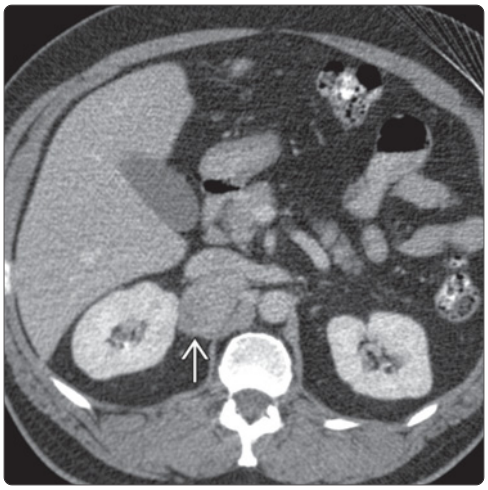
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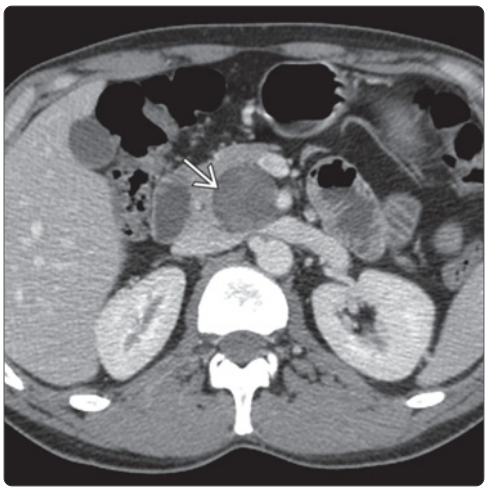
(Left) *Axial CECT shows a well-marginated, hypoattenuating mass ➡ medial to the left psoas muscle and adjacent to a neural foramen.* **(Right)** *Axial T2W MR in the same patient reveals high signal intensity in the periphery of the left paravertebral lesion ➡ due to myxoid matrix and central low signal intensity due to cellular neural tissue. Pathology was consistent with a solitary neurofibroma.*





(Left) *Axial CECT shows an enhancing retrocaval mass ➡. A radionuclide MIBG scan confirmed uptake within this lesion. This is a paraganglioma, which is equivalent to an extraadrenal pheochromocytoma.* **(Right)** *Coronal MIBG (iodine-131-meta-iodobenzylguanidine) scintigraphy shows focal uptake ➡ within the retrocaval soft tissue mass shown on prior CT. Pathology revealed a paraganglioma.*





(Left) *Axial post-contrast T1WI MR with fat suppression shows heterogenous enhancement of a left perirenal soft tissue mass ➡. Pathology proved ganglioneuroma.* **(Right)** *Axial CECT shows a well-marginated, heterogeneous mass ➡ arising from the posterior border of the pancreatic head. Surgical pathology was consistent with pancreatic schwannoma.*

Retroperitoneal Sarcoma

KEY FACTS

IMAGING

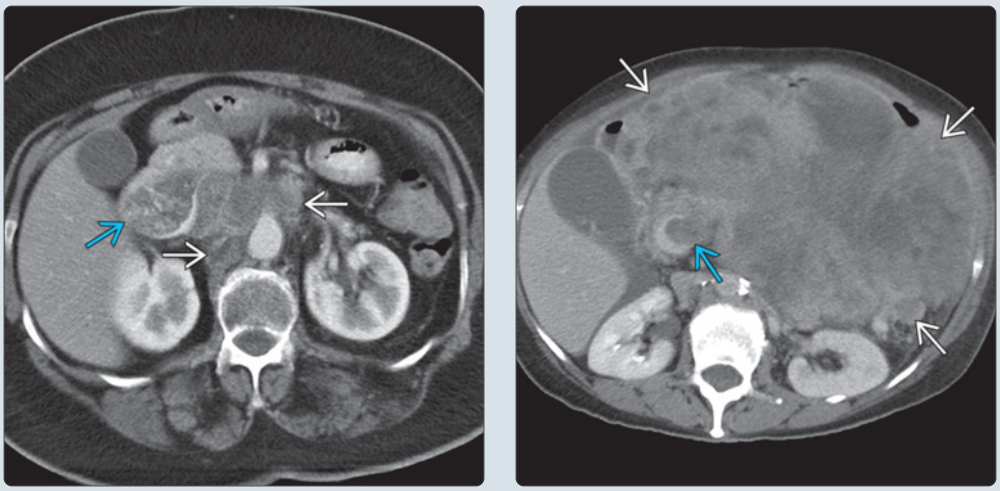
- 80% of RP tumors are malignant
  - Typical appearance: Large heterogeneous mass of fat + soft tissue attenuation displacing retroperitoneal structures or viscera
- Most malignant retroperitoneal tumors are of mesodermal origin
- Liposarcoma and leiomyosarcoma account for > 90%
- Liposarcoma: 3 CT patterns based on amount and distribution of fat in tumor (often coexist)
  - Discrete fatty areas < -20 HU; other soft tissue foci > +20 HU
  - Solid pattern: Attenuation values > +20 HU (mostly myxoid tissue)
  - Pseudocystic pattern: Homogeneous density between +20 and -20 HU
  - Can be mistaken for fluid collection
- Leiomyosarcoma
  - Extravascular (62%)
  - Large retroperitoneal mass ± necrotic or cystic degeneration
  - Intravascular mass (6%)
  - Solid mass within IVC and dilatation or obstruction of lumen

TOP DIFFERENTIAL DIAGNOSES

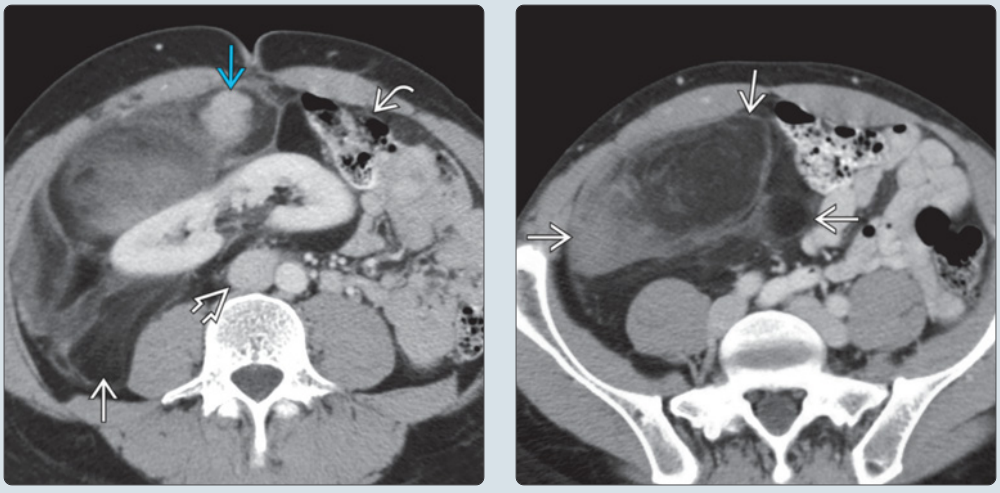
- Renal angiomyolipoma
  - Renal parenchymal defect ("notch" or "claw" sign) and enlarged vessels favor angiomyolipoma (AML)
- Adrenal myelolipoma
- Retroperitoneal hemorrhage

CLINICAL ISSUES

- Most retroperitoneal sarcomas are asymptomatic until sufficiently large
- Signs/symptoms include: Pain (abdomen, back, flank), palpable abdominal mass, lower extremity edema
- Differentiate from renal and adrenal tumors



(Left) *Axial CECT reveals a heterogeneously enhancing mass that expands the IVC. Tumor extends into paraaortic and prevertebral spaces of the retroperitoneum. Pathology revealed leiomyosarcoma that originated in the IVC.* **(Right)** *Axial CECT demonstrates a huge heterogeneous mass that arose in the retroperitoneum and displaced peritoneal structures anteriorly and laterally. Tumor thrombus is shown within the IVC. Pathology was consistent with a malignant fibrous histiocytoma.*



(Left) *Axial CECT shows a large right-sided retroperitoneal mass that displaces the kidney, IVC, and ascending colon. The mass has foci of fat and soft tissue attenuation, the latter representing areas of myxoid degeneration within a liposarcoma.* **(Right)** *Axial CECT in the same patient shows that the more caudal aspect of this large mass displaces, but does not invade, retroperitoneal structures and viscera. The mass was resected, but recurred within 1 year.*

TERMINOLOGY

Definitions

- Malignant primary retroperitoneal (RP) tumor arising from various elements of primitive mesenchyme, urogenital ridge, or embryonic remnants

IMAGING

General Features

- Best diagnostic clue
  - Large mass with heterogeneous attenuation (± fat ± soft tissue) that displaces RP structures or viscera
- Size
  - Usually large at diagnosis (> 10 cm)
- Key concepts
  - 80% of RP tumors are malignant
    - Lymphomas > sarcomas
    - Secondary tumors: Usually metastasize to nodes or perirenal space
  - Most malignant RP tumors are of mesodermal origin
    - Liposarcoma and leiomyosarcoma comprise > 90%
  - Classified into 4 types based on tissue of origin
    - Liposarcoma
    - Leiomyosarcoma
    - Fibrosarcoma and malignant fibrous histiocytoma (MFH)
    - Rhabdomyosarcoma
  - **Liposarcoma**: Adipose tissue tumor
    - Most common primary retroperitoneal malignant tumor
    - 2nd most common adult soft tissue sarcoma after MFH
    - 15% of all soft tissue tumors, 10-20% of them in retroperitoneum
    - Malignant from inception; rarely arise from lipomas
    - Indolent growth allows large size before detection
    - Histologically: 5 types
      - Well differentiated (almost pure fat)
      - Myxoid: Most common (40-50%)
      - Round cell
      - Pleomorphic
      - Mixed
  - **Leiomyosarcoma**: Smooth muscle tumor
    - 2nd most common primary RP tumor after liposarcoma
    - Accounts for 11% of all retroperitoneal malignant tumors
    - Leiomyosarcomas are more common than benign leiomyomas
  - **Fibrosarcoma and MFH**: Connective tissue tumors
    - Fibrosarcoma: 3% of all retroperitoneal malignant tumors
    - MFH: Most common soft tissue sarcoma in adults
      - Most commonly occurs in extremities
  - **Rhabdomyosarcoma**: Striated muscle tumor
    - Embryonal rhabdomyosarcomas: 60% of striated muscle tumors

CT Findings

- Liposarcoma
  - 3 CT patterns based on amount of fat in tumor
    - Mixed pattern (most common)
      - Discrete fatty areas < -20 HU; other soft tissue foci > +20 HU
    - Solid pattern: Attenuation values > +20 HU (mostly myxoid tissue)
    - Pseudocystic pattern: Homogeneous density between +20 and -20 HU
      - Can be mistaken for fluid collection
  - Encapsulated mass ± calcification
  - Displacement, compression, distortion of adjacent structures (kidneys, bowel, colon)
  - ± invasion of adjacent structures
  - CECT: Hetero-/homogeneous enhancement; usually lacks prominent vessels
- Leiomyosarcoma
  - Extravascular (62%)
    - Large retroperitoneal mass ± necrotic or cystic degeneration
    - Liver metastases (necrotic or cystic)
  - Intravascular mass (6%)
    - Solid mass within IVC and dilatation or obstruction of lumen
    - Proximal IVC mass: Show dilated hepatic veins and portal radicles
    - CECT: Heterogeneous enhancement of tumor
  - Extra- and intravascular mass (33%)
    - Solid and necrotic extraluminal tumor
    - Enhancing intravascular component
  - Intramural tumor: Extremely rare

MR Findings

- Variable signal intensities (SI) and enhancement depending on amount of fat; solid, cystic, necrotic, hemorrhagic components

Ultrasonographic Findings

- Grayscale ultrasound
  - Liposarcoma: Large, well-defined solid mass with echogenic (fat) and soft tissue components
  - Leiomyosarcoma: Large solid mass + hypoechoic cystic and necrotic content

Angiographic Findings

- Conventional
  - Show hypo-/hypervascularity; displacement of great vessels
  - Liposarcoma: Hypovascular tumor
  - Leiomyosarcoma: Hypervascular + feeding vessels
- Inferior venacavography
  - For primary or secondary involvement of IVC

Imaging Recommendations

- CECT or MR

DIFFERENTIAL DIAGNOSIS

Renal Angiomyolipoma

- Composed of blood vessels, muscle, and fat tissue
- Hamartomatous lesion (benign tumor)
- 80% of tuberous sclerosis patients have renal angiomyolipomas (AML)

- Large angiomyolipoma may simulate RP liposarcoma (both contain fat)
  - Renal parenchymal defect and enlarged vessels favor AML
  - Smooth compression of kidney and extension beyond perirenal space favor liposarcoma

Adrenal Myelolipoma

- Discrete suprarenal mass containing fat and myelogenous components
- Large adrenal mass may simulate liposarcoma
- Myelolipoma usually smaller; often contains foci of calcification

Retroperitoneal Hemorrhage

- Most are iatrogenic (over-anticoagulation)
- Abdominal aortic aneurysm rupture is 2nd most common cause of RP bleed
- Spontaneous tumoral bleed: 3rd most common cause
  - Kidney: Renal cell carcinoma, AML
- CT findings
  - Acute: High-density fluid collection or hematoma
  - Chronic: Low density (seroma)
  - Associated renal, adrenal tumors or aortic aneurysm
  - Coagulopathic hemorrhage
    - Hematocrit sign (cellular-fluid level)
    - Multiple sites of bleeding
    - Involvement of body wall muscles (iliopsoas and rectus)
- MR findings
  - Phases of variable SI (evolution of blood products)
  - Hyperacute: Isointense on T1WI and T2WI
  - Acute: T1WI: Iso- to hypointense; T2WI: Hypointense
  - Early subacute: T1WI: Hyperintense; T2WI: Hypointense
  - Late subacute: Hyperintense on T1WI and T2WI
  - Chronic phase: Periphery: Hypointense on T1WI and T2WI
    - Center: T1WI: Isointense; T2WI: Hyperintense

PATHOLOGY

General Features

- Embryology/anatomy
  - Mesodermal origin: Most adult malignant RP tumors

Gross Pathologic & Surgical Features

- Liposarcoma
  - Large encapsulated mass
  - White or yellow glistening material
- Leiomyosarcoma
  - Lobulated, encapsulated tumor
  - Cystic, necrotic, blood, and Ca++ components

Microscopic Features

- Liposarcoma
  - Mostly myxoid (mucinous and fibrous tissue); fat < 10%
- Leiomyosarcoma
  - Smooth muscle and atypical giant cells
  - Invasion of vessels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Abdominal, back, flank, radicular pain
  - Palpable mass; GI and urinary tract symptoms
  - Leg edema, varicosities, hypoglycemia

Demographics

- Age
  - 40-60 years
- Gender
  - Liposarcoma and MFH: M > F
  - Leiomyosarcoma: F > M
- Epidemiology
  - 1:11,800 hospital admissions

Natural History & Prognosis

- Liposarcoma
  - 5-year survival rate of 32%; less over time
    - Original mass can usually be resected
    - Tumors recur and become more aggressive
- Leiomyosarcoma
  - High mortality within 5 years
  - Local recurrence: 40-70% cases

Treatment

- Complete resection, radiotherapy, chemotherapy
- Follow-up imaging and excision of local recurrence

DIAGNOSTIC CHECKLIST

Consider

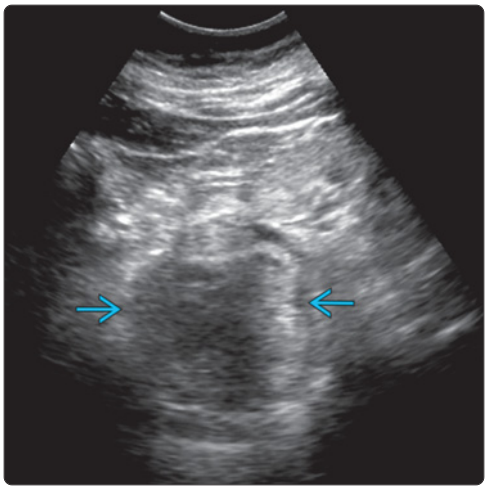
- Differentiate from adrenal and renal tumors

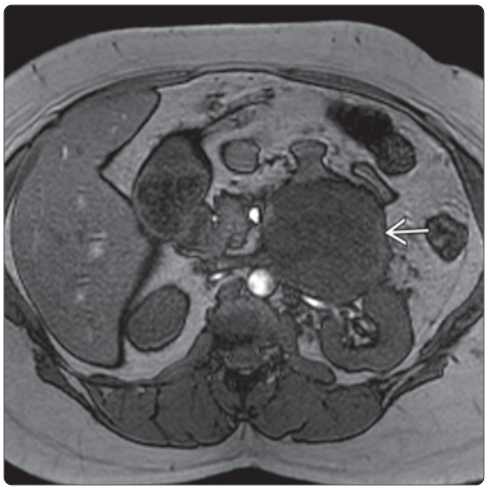
Image Interpretation Pearls

- Large mixed-attenuation mass of fat and soft tissue density displacing RP structures

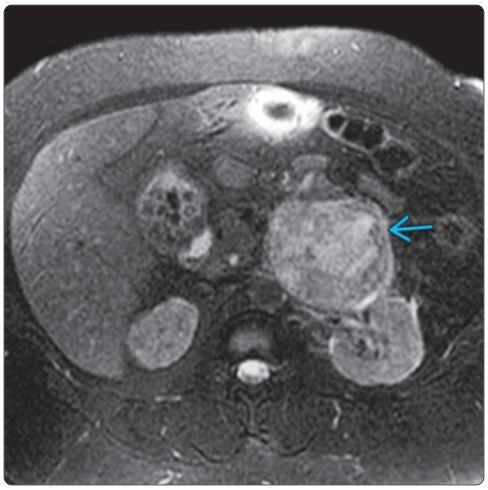
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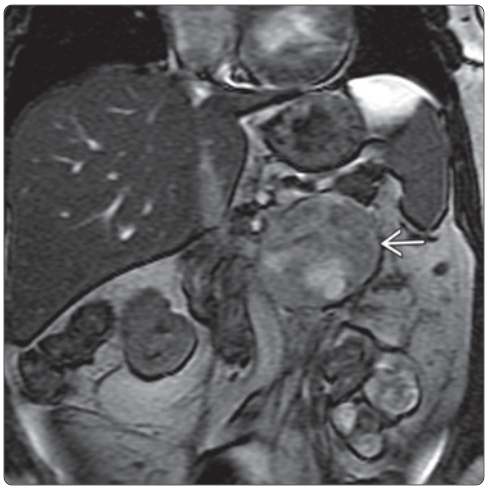
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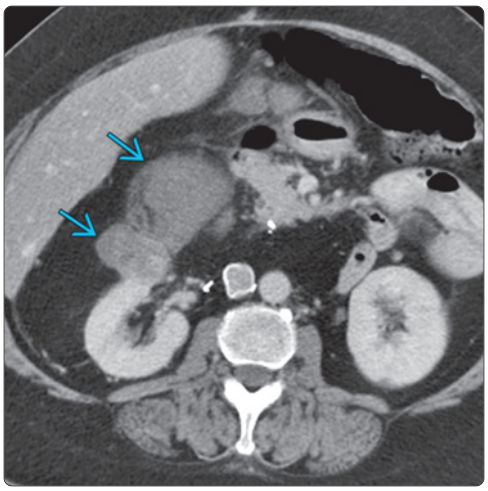


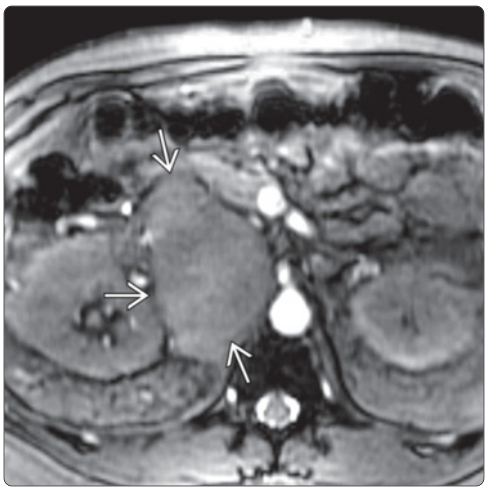
(Left) *Transverse US reveals a lobulated soft tissue mass in the left upper quadrant of an asymptomatic patient.* **(Right)** *Axial T1WI out-phase MR in the same patient reveals a soft tissue mass of intermediate soft tissue signal intensity. Compared to the in-phase sequence (not shown), there was no loss of signal during the out-phase sequence. Surgical resection was consistent with a malignant fibrous histiocytoma (MFH).*





(Left) *Axial T2W MR with fat suppression in the same patient demonstrates heterogeneous signal intensity throughout the MFH. Central high signal intensity was due to necrosis.* **(Right)** *Coronal SS FSE T2W MR in the same patient reveals heterogeneous signal intensity in the MFH. There was no invasion of any adjacent structures. MFH is more commonly an extremity soft tissue tumor, but it can also arise in the retroperitoneum and peritoneum.*





(Left) *Axial CECT shows a heterogeneously enhancing mass spanning the anterior pararenal and perirenal spaces. The mass consists of regions of fat and soft tissue attenuation and was proven to be a liposarcoma.* **(Right)** *Axial T2* GRE MR in a patient with leiomyosarcoma of the IVC shows a large, oblong mass with intermediate soft tissue signal intensity that occupies the expected position of the IVC.*

KEY FACTS

TERMINOLOGY

- Generally classified as non-Hodgkin lymphoma (NHL) vs. Hodgkin disease (HD)
  - However, WHO recognizes over 80 types of lymphoma

IMAGING

- Discrete or confluent soft tissue nodules in mesentery or paraaortic spaces
- Enlarged nodes (generally ≥ 1 cm in short axis) for mesenteric and retroperitoneum
  - Retrocrural nodes enlarged if ≥ 6 mm
- Confluent soft tissue mantle fills paraaortic spaces in advanced cases
  - Displacement of aorta from spine (unusual for benign etiologies)
- Nodes due to NHL typically larger than HD
  - Also more likely to be noncontiguous in distribution in NHL
  - Mesenteric adenopathy more common in NHL (> 50%) than HD (< 5%)
- HD: Typically involves superior paraaortic nodes initially without skip areas of involvement
- Sensitivity of 90-95% for PET compared to 80-85% for CECT
  - Useful to determine early response to chemotherapy
  - Often helpful to distinguish residual lymphoma vs. fibrosis postradiation

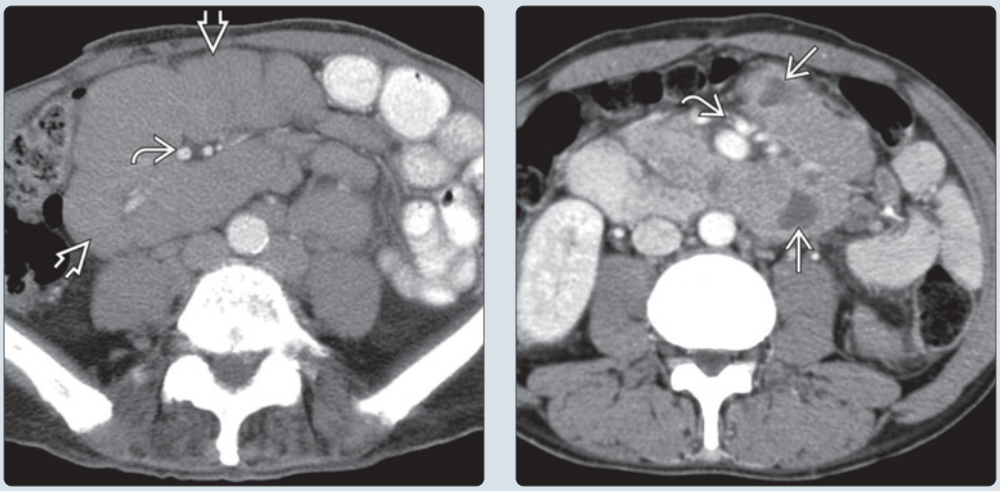
TOP DIFFERENTIAL DIAGNOSES

- Retroperitoneal metastases
- Testicular metastases
- Retroperitoneal sarcoma
- Retroperitoneal neurogenic tumor
- Retroperitoneal fibrosis
- Abdominal manifestations of tuberculosis

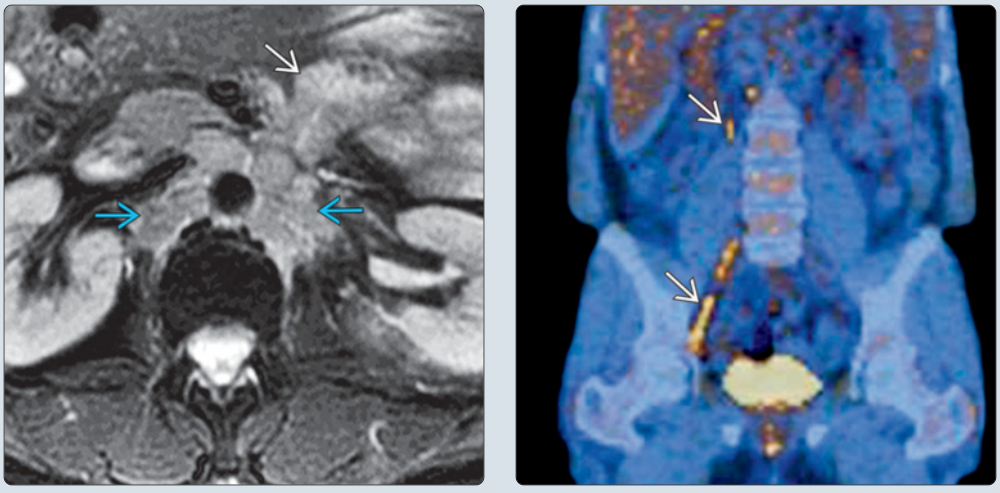
DIAGNOSTIC CHECKLIST

- Tissue sampling: Several core biopsies for flow cytometry

(Left) *Axial CECT in a 34-year-old man with AIDS and non-Hodgkin lymphoma (NHL) shows extensive mesenteric and paraaortic adenopathy. Note the "sandwich" sign created by the mesenteric nodes ➡ surrounding the vessels ➡. Mesenteric adenopathy is common in patients with NHL, but rare in Hodgkin disease (HD).* **(Right)** *Axial CECT shows multifocal soft tissue masses with foci of necrosis ➡ "sandwiching" the mesenteric vessels ➡ due to non-Hodgkin lymphoma.*



(Left) *Axial T2WI MR with fat suppression shows high signal intensity in confluent paraaortic ➡ and mesenteric ➡ adenopathy due to non-Hodgkin lymphoma.* **(Right)** *Fused coronal PET/CT in a patient with Hodgkin disease shows a long contiguous line of FDG-avid right paraaortic and iliac nodes ➡ that are normal in size and would be missed on CT as not involved by tumor.*



TERMINOLOGY

Synonyms

- Non-Hodgkin lymphoma (NHL) > Hodgkin disease (HD)

Definitions

- Malignancy of blood cells that develop from lymphocytes

IMAGING

General Features

- Location
  - Mesentery: Often adjacent to, or surrounding, mesenteric vessels
  - Retroperitoneum: Paraaortic (includes several subgroups) and along iliac vessels
- Size
  - Typically nodes ≥ 1 cm in short axis for mesenteric + paraaortic nodes
    - Enlarged retrocrural nodes: ≥ 6 mm
- Morphology
  - Round or irregular contour
  - Distortion or loss of normal fatty hilum
  - Discrete soft tissue nodules within mesentery or filling paraaortic spaces

CT Findings

- CECT
  - Enlarged nodes (generally ≥ 1 cm in short axis) for mesenteric and paraaortic nodes
    - Retrocrural nodes enlarged if ≥ 6 mm
  - Confluent soft tissue mantle may surround mesenteric vessels and aorta/IVC
  - Displacement of aorta from spine (unusual for benign etiologies)
  - Nodes enhance and attenuation is similar to muscle
  - In untreated patients, nodes are rarely calcified (< 1%) or cystic
  - Nodes due to NHL typically larger than HD
    - NHL nodes also more likely to have noncontiguous distribution
    - Mesenteric adenopathy more common in NHL (> 50%) than HD (< 5%)
  - Peripancreatic nodes due to NHL may mimic pancreatic mass
  - 25% of newly diagnosed patients with HD have positive paraaortic nodes at presentation, compared to 50% with NHL
  - HD: Typically involves superior paraaortic nodes initially without skip areas of involvement
  - HD nodes may be normal in size, but contain microscopic tumor leading to false-negative CT

MR Findings

- T1WI
  - Typically low signal adenopathy
- T2WI
  - Typically high signal intensity adenopathy
  - Low signal intensity may suggest fibrosis from treated disease
- DWI
  - Typically restricted diffusion
- T1WI C+
  - Nodes without necrosis enhance similar to muscle
  - Heterogeneous enhancement in necrotic or partially treated nodes

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic to heterogeneous nodes
    - Rounded or irregular shape
    - Distortion/loss of normal fatty hilum

Nuclear Medicine Findings

- PET
  - Hypermetabolic nodes show avid FDG uptake
  - Sensitivity of 90-95% for PET, compared to 80-85% for CECT
  - More sensitive for thoracic lymphoma (> 90%) than abdominal or pelvic disease (> 75%)
  - Allows ↑ detection of tumor in obscure sites (e.g., bone, muscle, distant/nonenlarged nodes)
    - May change stage in 10-40% of cases
    - Modifies therapy in 25% of cases
  - Useful to determine early response to chemotherapy
    - Decreased FDG uptake precedes decreased size
  - Helpful to distinguish residual lymphoma vs. fibrosis post radiation
  - Low sensitivity for B-cell lymphoma and mucosa-associated lymphoid tissue (MALT)

Imaging Recommendations

- Best imaging tool
  - PET/CT, CECT, MR
- Protocol advice
  - PET/CT: Highly sensitive examination
  - CECT: Coronal reformations helpful for node assessment
  - MR: T2 + DWI + post-contrast T1WI with fat saturation

DIFFERENTIAL DIAGNOSIS

Retroperitoneal Metastases

- Enlarged nodes that are discrete, but typically not "confluent"
- Occasional calcification for some tumors (i.e., mucinous adenocarcinoma)

Testicular Metastases

- Nodes enlarged along gonadal vein as it drains into IVC (right) or renal vein (left)
- Enlarged nodes ipsilateral to testicular tumor

Retroperitoneal Sarcoma

- Unilateral large, heterogeneous retroperitoneal mass
- Organ and vessel displacement
- Heterogeneous fat attenuation in liposarcoma

Retroperitoneal Neurogenic Tumor

- Paraganglioma, ganglioneuroma, schwannoma
- Paraspinous location + widening of neural foramen

Abdominal Manifestations of Tuberculosis

- Low-attenuation nodes; loculated ascites

Retroperitoneal Fibrosis

- Usually limited to mantle of tissue surrounding aorta and IVC from midlumbar level through common iliac

Miscellaneous

- Kaposi sarcoma: Hypervascular retroperitoneal nodes in AIDS patient
- Castleman disease (angiofollicular lymphoid hyperplasia): Hypervascular retroperitoneal nodes
- Growing teratoma syndrome: Water- or fat-density nodal mass due to development of mature teratoma following chemotherapy for testicular nonseminomatous tumor
- Sarcoidosis: Adenopathy, pulmonary manifestations, hepatosplenic lesions

PATHOLOGY

General Features

- Etiology
  - Unknown; numerous risk factors reported
- Genetics
  - Several types of NHL associated with oncogenes Bcl 1, 2, 3, and 6

Staging, Grading, & Classification

- WHO recognizes over 80 forms of lymphoma
- Ann Arbor staging classification
  - Stage I: Single lymph node region/extralymphatic site
  - Stage II: 2 or more nodal regions on same side of diaphragm (including single extralymphatic site)
  - Stage III: Nodal group involvement above and below diaphragm
  - Stage IV: Diffuse involvement of 1 or more extralymphatic sites (liver, lungs, marrow)

Gross Pathologic & Surgical Features

- Excisional or core biopsy preferred for subtyping

Microscopic Features

- HD
  - Few malignant cells within background of normal T lymphocytes and inflammatory cells
  - Malignant cells: Reed-Sternberg cells and variants
    - Lacunar cells, mononuclear and pleomorphic variants
  - Distorted nodal architecture
  - Histologic subtypes: Nodular sclerosing, lymphocyte predominant, mixed cellularity, lymphocyte depleted
- NHL
  - Classification typically based on WHO criteria
  - Cell type origin determines subtype (B cell, T cell, etc.)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, night sweats, weight loss
  - Other signs/symptoms: Axillary, neck or groin mass
- Clinical profile
  - Young patient with palpable nodal mass

Demographics

- Age
  - HD bimodal; 2 peak incidences
    - 20-24 years
    - 80-84 years
  - NHL: ↑ incidence with ↑ age
- Gender
  - HD: Slightly increased in males
  - NHL: Before age 45 years, more common in males
    - After age 45 years, no gender difference
- Ethnicity
  - HD: Higher in Caucasians
- Epidemiology
  - ~ 5% of newly diagnosed cancer in USA
    - HD: ~ 8,000 cases/year in USA
    - NHL: ~ 50,000 cases/year in USA
  - ↑ incidence due to AIDS-related NHL and organ transplantation
  - 3rd most common cancer death

Natural History & Prognosis

- HD: 80% cure with chemoradiation or stem cell transplant
- NHL: Depends on grade + stage
  - NHL prognosis typically worse than HD
  - Worst prognosis: AIDS-related NHL
  - Poor prognosis if no response to initial therapy

Treatment

- Options, risks, complications
  - HD: Initial chemotherapy or radiation; stem cell transplant for treatment failure or relapse
  - NHL: Initial treatment with chemotherapy; stem cell transplant for failure or recurrence

DIAGNOSTIC CHECKLIST

Consider

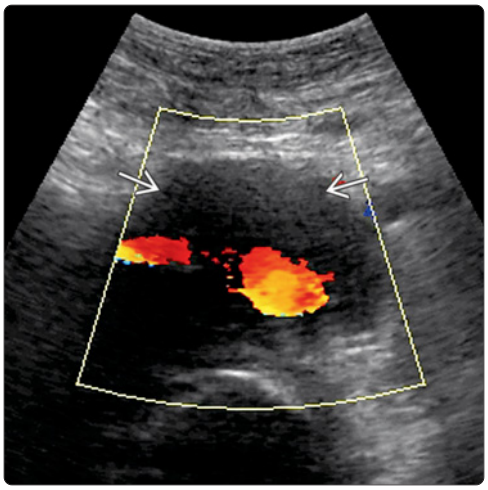
- Metastatic nodes from carcinomas

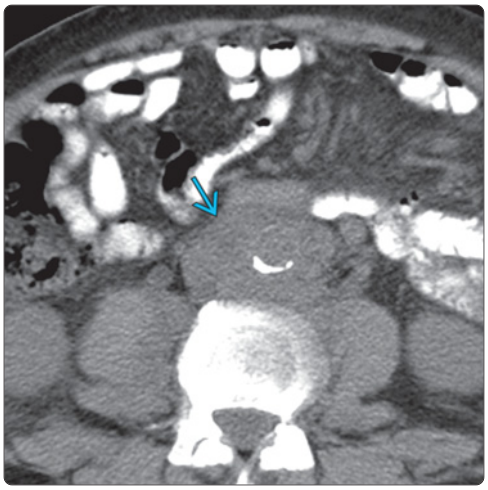
Image Interpretation Pearls

- Discrete soft tissue nodules or confluent soft tissue in mesentery &/or paraaortic spaces

SELECTED REFERENCES

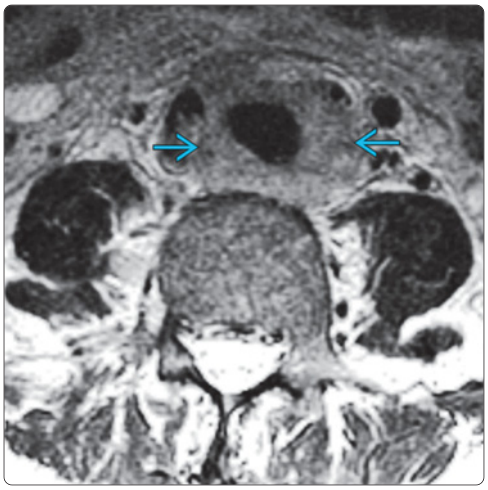
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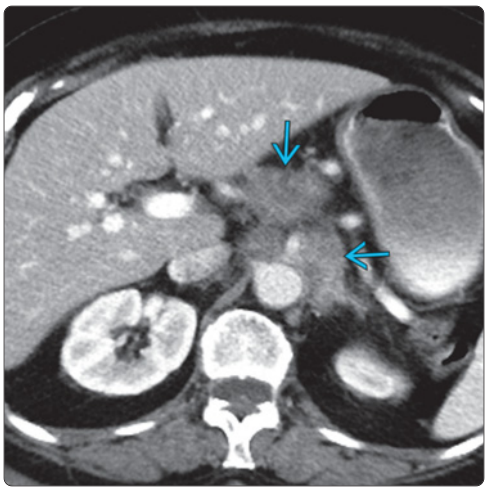


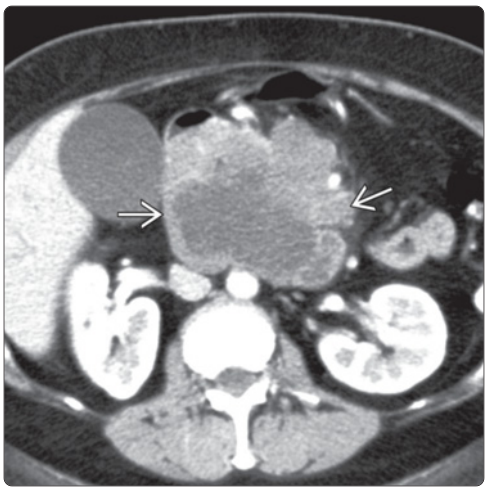
(Left) *Transverse US shows a soft tissue mass ➡ surrounding the aorta and IVC due to non-Hodgkin lymphoma.* **(Right)** *Axial NECT in the same patient shows that the adenopathy ⇨ from NHL fills the paraaortic spaces and displaces the aorta from the spine.*





(Left) *Axial PET/CT in the same patient demonstrates that the NHL adenopathy ➡ is FDG-avid.* **(Right)** *Axial T2WI MR SS FSE in the same patient shows that the periaortic mass ⇨ has intermediate to high signal intensity.*





(Left) *Axial CECT shows confluent paraceliac adenopathy ⇨ due to NHL.* **(Right)** *Axial CECT shows pancreatic non-Hodgkin lymphoma as a bulky mass ➡ in the head of pancreas region. The lymphoma occupies the peripancreatic nodal group, but the appearance may simulate a primary pancreatic tumor.*

Retroperitoneal Metastases

KEY FACTS

IMAGING

- Soft tissue masses or abnormal nodes in retroperitoneum
  - Masses displace aorta + IVC from spine
- Retroperitoneal adenopathy: Size criteria
  - Paraaortic, pelvic adenopathy: ≥ 10 mm short axis
  - Retrocrural nodes: ≥ 6 mm short axis diameter
  - Nodal size not reliable criterion alone (yet still most used)
- Retroperitoneal adenopathy: Morphology
  - Round, irregular margins
- Retroperitoneal adenopathy: Architecture
  - Distortion or loss of normal fatty hilum
- Retroperitoneal adenopathy: Enhancement
  - Variable due to type of primary tumor
  - Similar enhancement suggests same grade and aggressiveness
- Etiologies of metastases
  - Lymphoma and anaplastic tumors tend to cause confluent nodal masses
    - Other nodal metastases tend to be discrete masses
  - Testicular seminomas: Bulky retroperitoneal nodes
    - Nonseminomatous germ cell tumors: Small, clustered nodes
  - Extranodal RP metastases: Melanoma, renal cell, anaplastic, breast, small cell lung cancers

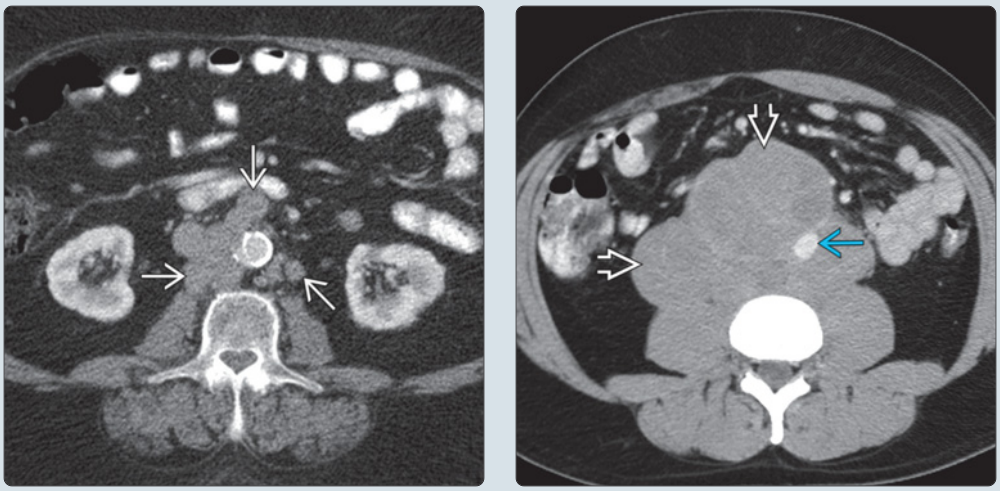
TOP DIFFERENTIAL DIAGNOSES

- Lymphoma
- Infectious
- Primary retroperitoneal tumors
- Inflammatory
- Retroperitoneal fibrosis

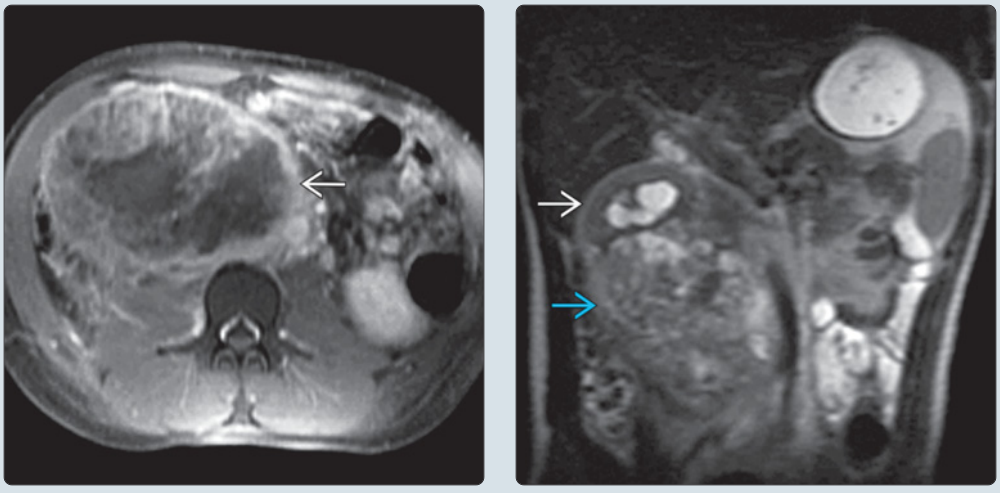
PATHOLOGY

- Depending on primary tumor site: Nodal metastases may constitute N1 (regional), N2, or M1 (distant metastases) spread of disease

(Left) *Axial CECT shows several discrete, enlarged nodes ➡ in the paraaortic spaces (measuring 1-1.5 cm) typical of malignant adenopathy. These lymph node metastases were due to prostate cancer.* **(Right)** *Axial CECT shows extensive, confluent paraaortic adenopathy ➡ due to metastatic testicular seminoma. Note that the aorta ⇨ is displaced from the lumbar spine by the mantle of soft tissue; a finding that is not seen in typical retroperitoneal fibrosis.*



(Left) *Axial post-contrast T1WI MR with fat saturation reveals a large, retroperitoneal mass ➡ that is partially necrotic. Its location to the right of midline reflects that the metastasis is ipsilateral to the primary tumor in the right testis (not shown).* **(Right)** *Coronal T2WI MR in the same patient shows superolateral displacement and hydronephrosis of the right kidney ➡ due to mass effect from the right retroperitoneal mass ⇨.*



TERMINOLOGY

Abbreviations

- Retroperitoneal (RP) metastases (METS)

Definitions

- Spread of malignancy to retroperitoneal lymph nodes and tissues

IMAGING

General Features

- Best diagnostic clue
  - Enlarged, architecturally distorted or FDG-avid nodes in patient with known malignancy
  - Cluster of subcentimeter RP nodes in patient with nonseminomatous testicular malignancy
- Location
  - Paraaortic nodes (numerous subgroups)
    - Testicular cancers: Lymph drainage follows gonadal vessels; nodal metastases most commonly at ipsilateral renal hilum
    - Pelvic cancers: Obturator nodes may be 1st site
  - Iliac chain nodes (common, internal, external)
  - Retrocrural nodes
  - Perinephric space
- Size
  - Paraaortic, iliac chain nodes ≥ 10 mm short axis
    - ↑ concern for metastases in patient with malignancy
  - Size alone is not reliable criterion for malignancy
    - Nodes containing malignancy often < 10 mm
    - Nodes ≥ 10 mm may be benign (i.e., infection)
  - Retrocrural nodes: ≥ 6 mm short axis diameter
- Morphology
  - Round or irregular contour
  - Distortion or loss of fatty hilum
  - Confluent or infiltrating masses

CT Findings

- NECT
  - Calcification in metastatic nodes from ovarian, bladder, and mucin-producing tumors
    - Nonspecific: Can also be seen in tuberculosis + treated lymphoma/seminoma
- CECT
  - Discrete or confluent nodal masses
    - Lymphoma and anaplastic tumors tend to have confluent nodal masses
  - Low-attenuation nodes can be seen in untreated testicular cancer and lymphoma
    - Enlarging low-attenuation RP mass in treated nonseminomatous germ cell tumors (GCT): Residual teratoma
  - Testicular cancer: Bulky RP nodes in seminomas; small, clustered nodes in nonseminomatous GCT
    - Usually asymmetric: Larger nodes ipsilateral to tumor
  - Masses displacing aorta and IVC from spine are usually malignant
    - Vessels not displaced in typical retroperitoneal fibrosis
  - Metastases to extranodal RP spaces
    - Melanoma, renal cell, breast, small cell lung cancers

MR Findings

- T1WI
  - Typically low to intermediate signal intensity (SI) nodes/masses
  - Metastatic melanoma may have ↑ SI
- T2WI
  - Typically ↑ SI
  - ↓ SI following treatment may suggest fibrosis
- DWI
  - ↑ SI: Metastatic nodes/masses with high cellularity
  - ↓ SI: Necrotic nodes/masses (low cellularity)
- T1WI C+
  - Often mimics enhancement of primary tumor
    - Indicates same grade and aggressiveness
- Nanoparticle-enhanced MR
  - Use of iron oxide particles to assess phagocytic function
    - Normal nodes: ↓ SI
    - Malignant nodes: ↑ SI

Ultrasonographic Findings

- Grayscale ultrasound
  - Round or irregular nodes with ratio longitudinal/transverse diameter < 2
  - Distorted or absent echogenic fatty hilum
  - Profoundly hypoechoic or heterogeneous echotexture
- Color Doppler
  - Variable flow due to tumor vascularity (i.e., ↑ in melanoma)
  - Displacement of normal nodal hilar vessels

Nuclear Medicine Findings

- PET
  - PET/CT: Often more sensitive and specific than CT alone
    - However, well-differentiated nonseminomatous GCTs not very FDG avid

Imaging Recommendations

- Best imaging tool
  - CECT or MR; PET/CT for FDG-avid tumors
- Protocol advice
  - Oral contrast may be helpful in thin patients to distinguish bowel from RP nodes

DIFFERENTIAL DIAGNOSIS

Lymphoma

- Enlarged nodes or confluent soft tissue mantle
- Soft tissue often displaces aorta anteriorly from spine

Infection: Tuberculosis, AIDS, Mononucleosis

- TB: Frequently low-attenuation adenopathy
- AIDS: Adenopathy usually not massive
- Mononucleosis: Hepatosplenomegaly

Primary Retroperitoneal Tumors

- Mesenchymal tumors: Liposarcoma, leiomyosarcoma, malignant fibrous histiocytoma
- Typically large (~ 10-20 cm), heterogeneous

Retroperitoneal Fibrosis

- Mantle of soft tissue encasing inferior aorta/IVC

- ~ 10% of cases are due to metastatic foci inciting desmoplastic response

Retroperitoneal Hemorrhage

- Multiple foci, hematocrit sign, involvement of iliopsoas and rectus sheath most common
- Clinical setting of anticoagulation, trauma, &/or dropping hematocrit

Sarcoidosis

- Abdominal adenopathy in ~ 30%
- Pulmonary disease ± hepatosplenic granulomas

Lymphangioleiomyomatosis

- Smooth muscle proliferation in lymph vessels causes obstruction
- Lobulated, hypoattenuating masses surround/displace RP vessels

PATHOLOGY

General Features

- Etiology
  - Spread of malignancy to RP nodes and tissues
    - Most common source: Primary pelvic tumor
      - Testicular GCT: Nodes at ipsilateral renal hilum
      - Ovary, bladder, prostate, uterus, colon: Pelvic + paraaortic nodes
    - Various other primary malignancies (breast, melanoma, lymphoma)
  - Malignant retroperitoneal fibrosis: Desmoplastic reaction to cancer cells
- Associated abnormalities
  - Ureteral obstruction due to bulky tumor
- Mode of spread
  - Lymphatic or hematogenous spread: Testicular cancer, melanoma, ovarian, prostate, lung, breast
  - Direct extension from primary intraabdominal neoplasms: Pancreas, GI tract cancers

Staging, Grading, & Classification

- Depends on site of primary tumor and size of nodes
  - Nodal metastases may constitute N1 (regional), N2 or M1 (distant) disease

Gross Pathologic & Surgical Features

- Seminoma: May have bulky retroperitoneal masses
- Nonseminomatous GCTs and gynecologic malignancies
  - Often have metastases within normal-sized nodes

Microscopic Features

- Testicular GCTs: Metastases may have different histology from primary testicular lesion
  - Embryonal carcinoma: May show teratomatous histology in nodes
  - Teratoma may show choriocarcinoma in nodes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most commonly asymptomatic
  - Other signs/symptoms
    - Back pain, hematuria, obstructive uropathy
- Clinical profile
  - RP adenopathy: Lymphoma, testicular, renal cell, cervical, prostate, or ovarian cancer
  - Non-nodal RP metastases: Melanoma, renal cell, breast, small cell lung cancers

Natural History & Prognosis

- Testicular cancer: Excellent prognosis even with RP adenopathy

Treatment

- Options, risks, complications
  - Primary pelvic and retroperitoneal tumors
    - Usually resect primary tumor + regional nodes
  - If primary tumor is not in retroperitoneum or pelvis, RP lymphadenopathy is considered stage IV disease
    - Commonly treated with chemotherapy
  - Testicular germ cell tumors: 3 possibilities for residual masses
    - Residual cancer: Treat with chemotherapy
    - Residual benign teratoma following chemotherapy ("growing teratoma" syndrome): Resection
    - Fibrosis: No treatment needed

DIAGNOSTIC CHECKLIST

Consider

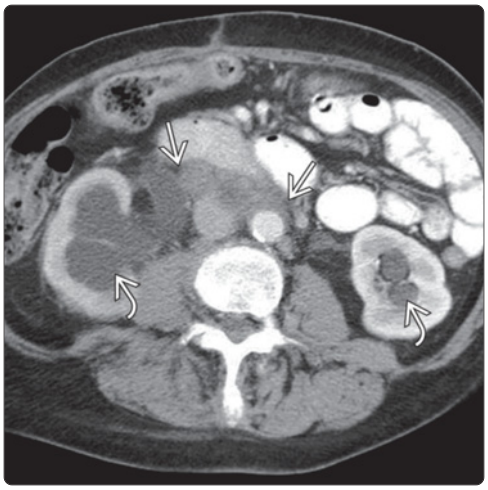
- Nodal size alone is unreliable criterion
  - Especially with primary pelvic malignancies

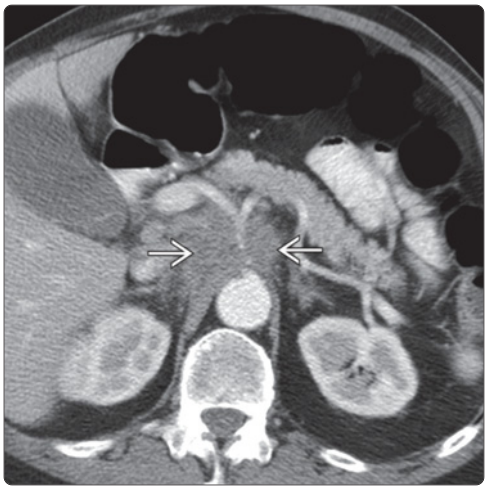
Image Interpretation Pearls

- Nonseminomatous GCTs: Any detectable nodes at ipsilateral renal hilum should be considered suspicious regardless of size

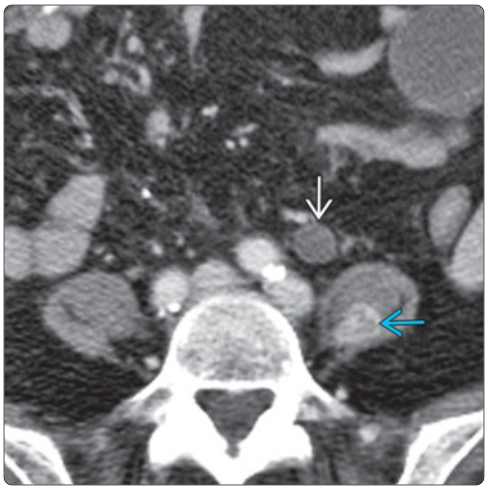
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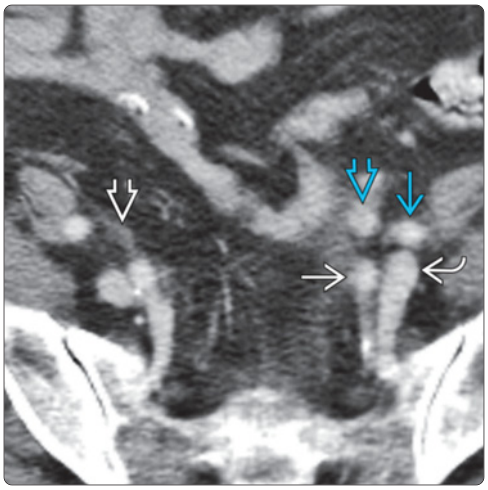
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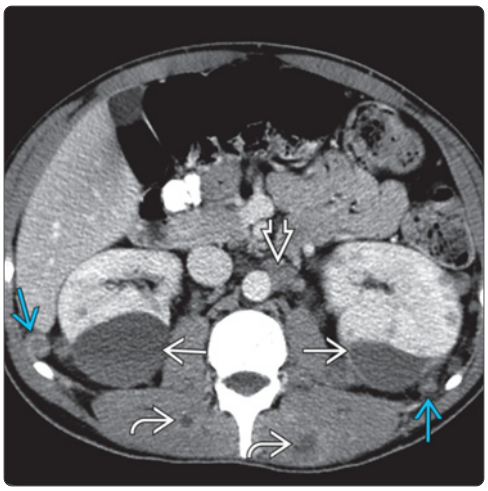


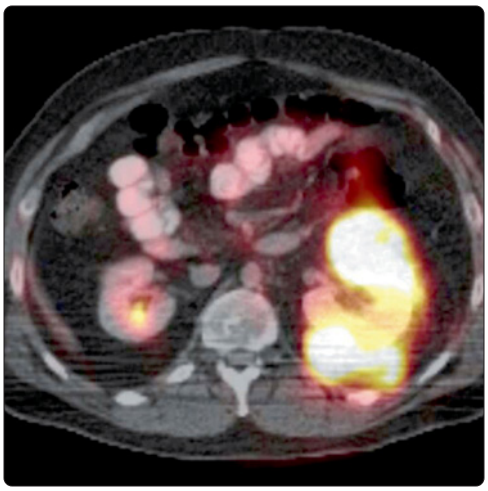
(Left) *Axial CECT demonstrates bilateral hydronephrosis due to confluent retroperitoneal nodal metastases resulting in ureteral obstruction.* **(Right)** *Axial CECT shows confluent nodes encasing the celiac axis due to metastatic esophageal carcinoma. Cancer of the distal esophagus often metastasizes to upper abdominal nodes prior to liver or lungs.*





(Left) *Axial CECT shows left hydroureter and an enhancing metastatic lesion in the left psoas muscle.* **(Right)** *Axial CECT in the same patient shows an enhancing metastasis obstructing the left ureter. The left common iliac artery and vein and left gonadal vein are identified. The normal right ureter is shown for comparison.*





(Left) *Axial CECT shows bilateral renal subcapsular hematomas, perinephric soft tissue nodules, RP adenopathy, and muscular lesions due to metastatic melanoma in a 21-year-old male patient.* **(Right)** *FDG-avid tumor is present within the left kidney and perirenal space, representing hematogenous metastases from melanoma to the retroperitoneum. For tumors that are FDG avid, PET/CT is the ideal imaging tool, because it often detects metastases in unexpected sites and normal-sized nodes.*

Hemangiopericytoma

KEY FACTS

TERMINOLOGY

- Hemangiopericytomas are a group of neoplasms that arise from pericytes in walls of capillaries and can occur in various tissues
  - Renal hemangiopericytoma is rare, vascular neoplasm of variable clinical behavior

IMAGING

- No specific imaging features allow diagnosis
- US
  - Solid renal mass of variable echogenicity
  - Flow may be detected with color Doppler
- CECT
  - Heterogeneously enhancing soft tissue mass
- MR
  - T1WI: Typically low signal intensity
  - T2WI: Heterogeneous intermediate to high signal intensity

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma
- Oncocytoma
- Renal angiosarcoma

PATHOLOGY

- Most renal hemangiopericytomas are benign
  - However, clinical behavior is variable
  - Therefore, long-term follow-up is recommended

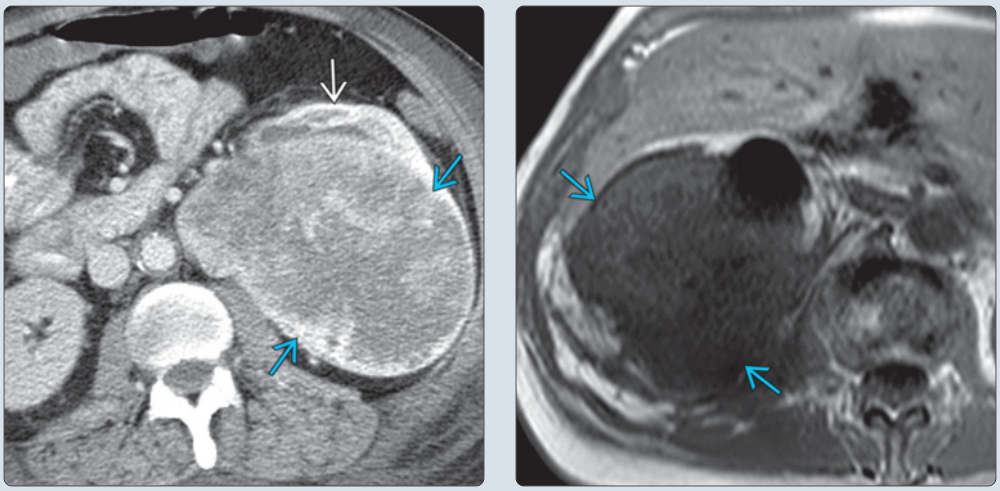
CLINICAL ISSUES

- Most common symptoms
  - Painless mass > flank pain > hematuria > hypoglycemia > hypertension

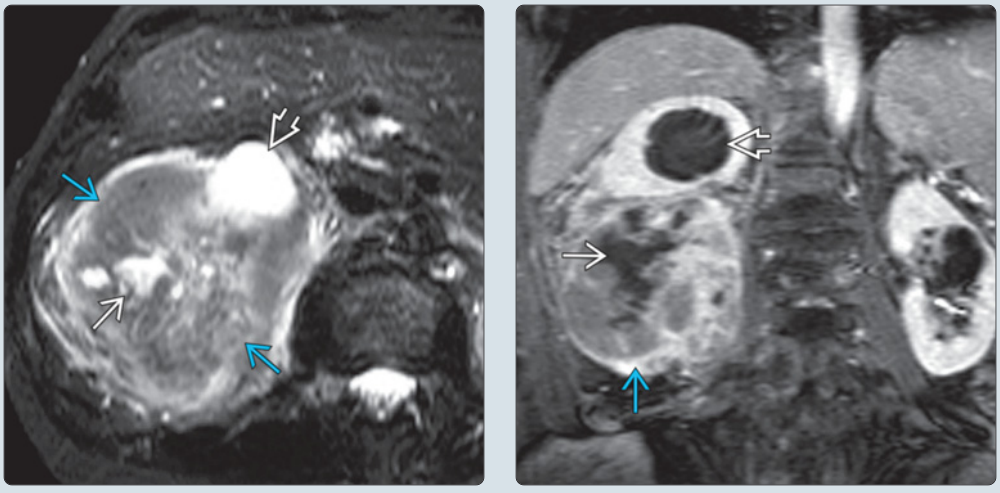
DIAGNOSTIC CHECKLIST

- No specific imaging or clinical findings to facilitate diagnosis
- Diagnosis of renal hemangiopericytoma is one of exclusion, requiring immunohistochemistry and electron microscopy

(Left) *Axial CECT demonstrates a large, ovoid, heterogeneously enhancing mass in the left kidney. A small amount of normal renal parenchyma is located anteriorly. The left renal vessels were displaced, but not invaded, and no adenopathy was present. Resection specimen was consistent with renal hemangiopericytoma.* **(Right)** *Axial T1WI MR reveals a large mass replacing the majority of the right kidney. The mass has homogeneously low signal intensity and displaces, but does not invade, the renal vessels.*



(Left) *Axial T2WI MR with fat suppression in the same patient shows intermediate, heterogeneous signal intensity throughout the mass & a small area of necrosis. An incidental cyst is shown anteriorly.* **(Right)** *Contrast-enhanced coronal T1WI MR with fat suppression in the same patient reveals an area of necrosis within the mass. An incidental cyst is shown in the superior pole of the right kidney. Differential considerations for the mass include various primary renal neoplasms. Pathology revealed renal HPC.*



TERMINOLOGY

Abbreviations

- Hemangiopericytoma (HPC)

Synonyms

- Myopericytoma, infantile myofibromatosis, solitary fibrous tumor, and HPC-like lesions are heterogeneous groups of neoplasms that have been referred to as "hemangiopericytomas"
  - All share pericytic (myoid) differentiation

Definitions

- Vascular neoplasm that arises from pericytes and mostly occurs in extremities, pelvis, head/neck, and meninges; renal involvement is very rare

IMAGING

General Features

- Location
  - Most HPCs arise in extremities or retroperitoneum, but can affect any tissue in body
  - Renal HPCs can arise anywhere in kidney
    - Rarely reported to arise in renal pelvis
- Size
  - Variable, ranging from 2-25 cm
- Morphology
  - Round or oval

CT Findings

- NECT
  - Soft tissue attenuation renal mass
- CECT
  - Heterogeneously enhancing renal mass
    - ± necrosis, calcification
    - + mass effect, but typically not vascular invasion

MR Findings

- T1WI
  - Typically low signal intensity
- T2WI
  - Heterogeneous intermediate to high signal intensity

Ultrasonographic Findings

- Grayscale ultrasound
  - Solid renal mass of variable echogenicity
- Color Doppler
  - Flow may be identified in solid components

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR
- Protocol advice
  - Renal mass protocol (multiphasic imaging)

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- Metastases and vascular invasion are more common with RCC

Oncocytoma

- Heterogeneously enhancing solid mass with nonspecific features

Renal Angiosarcoma

- Large aggressive mass with metastases and vascular invasion

PATHOLOGY

General Features

- Etiology
  - Idiopathic

Gross Pathologic & Surgical Features

- Thinly encapsulated, vascular soft tissue mass

Microscopic Features

- Arise from pericyte cells in capillaries
  - Function to moderate blood flow and permeability
- Variable cell size, morphology, connective tissue
- Variable amount of intercellular connective tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless mass > flank pain > hematuria > hypoglycemia > hypertension
    - May also be asymptomatic and incidentally discovered

Demographics

- M = F; most common in 5th decade

Natural History & Prognosis

- Most renal HPCs are benign
  - Follow-up imaging to exclude long-term recurrence
- Metastases: Hematogenous > lymphogenous
- Lung is most common site of metastasis for renal HPC
- Prognosis correlates with histology
  - Inversely related to mitoses, tumor size, necrosis

Treatment

- Wide surgical excision is mainstay of treatment
  - Adjuvant chemoradiation not effective

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Perivascular Epithelioid Cell Tumor (PEComa)

KEY FACTS

TERMINOLOGY

- Rare group of mesenchymal neoplasms associated with blood vessel walls

IMAGING

- CECT
  - Variable enhancement
    - Hypovascular to avid enhancement
  - Necrosis in large tumors
  - Well circumscribed
  - Rare calcification
- MR
  - T1WI: Hypo- to isointense
  - T2WI: Heterogeneously hyperintense
- Ultrasound
  - Heterogeneous echotexture

TOP DIFFERENTIAL DIAGNOSES

- Renal origin
  - Renal cell carcinoma
  - Transitional cell carcinoma
  - Lymphoma
- Retroperitoneal origin
  - Various sarcomas

PATHOLOGY

- Typically express melanocytic and smooth muscle markers

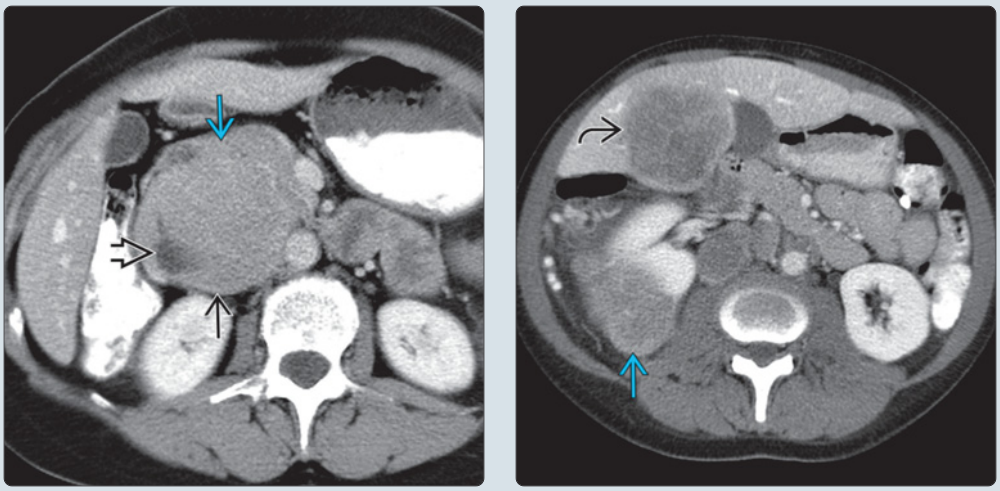
CLINICAL ISSUES

- Usually present as incidentally discovered mass
- Most PEComas are benign
  - Minority are malignant and metastasize

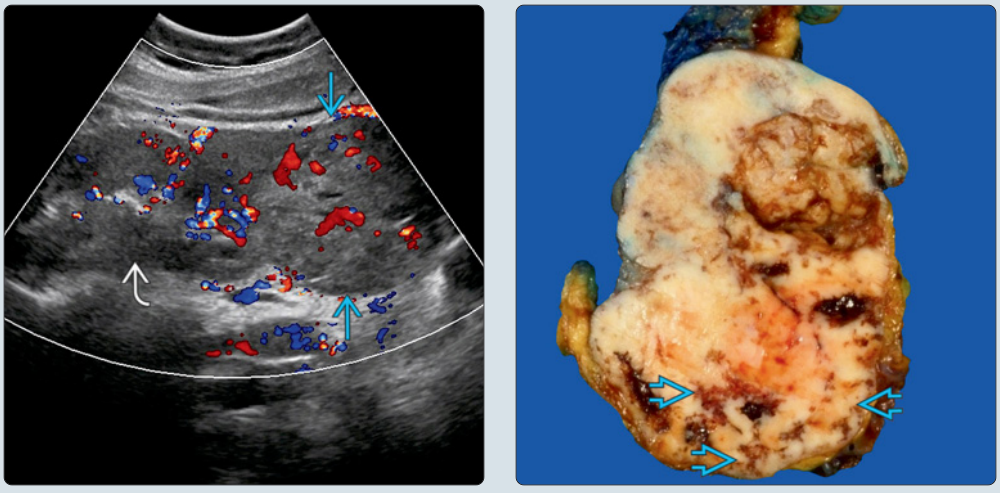
DIAGNOSTIC CHECKLIST

- Consider PEComa for retroperitoneal masses that do not contain fat
- Consider PEComa for renal masses that lack adenopathy and vascular invasion

(Left) *Axial CECT reveals a large, heterogeneously enhancing mass arising in the right aspect of the retroperitoneum. The mass results in compression and posterior displacement of the IVC. A small area of necrosis is shown along the lateral aspect of the mass.* **(Right)** *Axial CECT shows an enhancing mass arising from the lateral cortex of the right kidney. A similar appearing metastasis is within hepatic segment 5. Only a minority of PEComas are malignant and metastasize.*



(Left) *Longitudinal color Doppler US demonstrates a solid mass extending from the inferior pole of the left kidney. There is extensive color flow within the exophytic mass. Although not shown on this image, there was no invasion of the renal vessels or adenopathy.* **(Right)** *Photograph of the resection specimen reveals innumerable small vessels coursing through the mass, demonstrating the often vascular nature of PEComas. Pathology confirmed PEComa.*



TERMINOLOGY

Abbreviations

- Perivascular epithelioid cell tumor (PEComa)

Synonyms

- Subgroups of PEComa
  - Angiomyolipoma, clear cell "sugar" tumors, clear cell myomelanocytic tumors, sarcoma of perivascular cells, lymphangioleiomyomatosis, and pigmented melanotic tumors

Definitions

- Mesenchymal tumor comprised of perivascular epithelioid cells arranged radially around a vascular lumen

IMAGING

General Features

- Best diagnostic clue
  - PEComas can arise in many tissues
    - Kidney: Indistinguishable from renal neoplasms and lymphoma
    - Retroperitoneum: Usually indistinguishable from sarcomas
      - However, PEComas do not contain fat
- Location
  - Most arise in genitourinary tract, retroperitoneum, or gastrointestinal tract
- Size
  - Large (average size ~ 11 cm)
- Well marginated, round or oval

CT Findings

- NECT
  - Hypo- to isoattenuating to skeletal muscle
  - Rare calcification
- CECT
  - Variable enhancement, ranging from hypovascular to avid
  - Invasion of vascularity and adjacent structures is rare
  - Necrosis observed in large tumors

MR Findings

- T1WI
  - Hypo- to isointense to skeletal muscle
- T2WI
  - Heterogeneously hyperintense

Ultrasonographic Findings

- Heterogeneous echotexture; necrosis in large tumors

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- More commonly associated with vascular invasion and adenopathy

Transitional Cell Carcinoma (TCC)

- More likely to have ill-defined, infiltrative borders

Lymphoma

- More likely to have systemic or regional adenopathy

PATHOLOGY

General Features

- Associated abnormalities
  - Characteristic mutations in tuberous sclerosis complex (TSC)
    - Deletions of short arm of chromosome 16p (*TSC2* gene)

Staging, Grading, & Classification

- Characterized by perivascular location

Microscopic Features

- Express melanocytic and smooth muscle markers

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually incidentally discovered mass

Demographics

- Age
  - Most common in 6th decade
- Gender
  - F:M = 2-3:1

Natural History & Prognosis

- Most PEComas are benign
  - However, minority are malignant and metastasize
    - Of these, most (~ 75%) of metastasize within 1 year
    - Metastases: Liver > lung > omentum > skeletal

Treatment

- Surgical resection

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- None; no specific imaging features to differentiate from other neoplasms

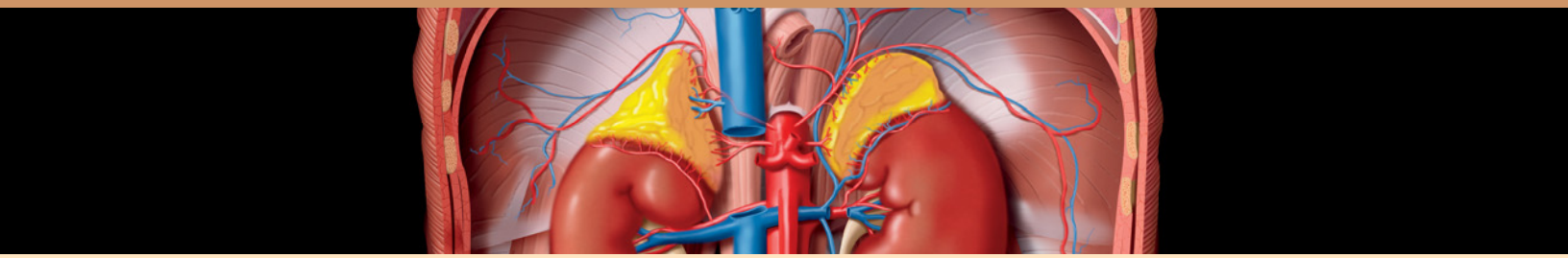
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SECTION 3

Adrenal



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Anatomy, Embryology, and Physiology

The adrenal (suprarenal) glands lie in the perirenal space, usually cephalic to the kidneys. The right adrenal gland lies above the kidney, lateral to the crus of the diaphragm, medial to the liver, and touches the posterior aspect of the inferior vena cava. The left adrenal gland usually lies anterior to the upper pole of the left kidney, posterior to the splenic vein and pancreas. The splenic vein is a useful landmark in identifying the source of left upper quadrant masses that may be of uncertain origin. Adrenal masses generally displace the splenic vein forward, while pancreatic or gastric masses displace it posteriorly.

The adrenal cortex and medulla are essentially different organs that lie within the same structure. The cortex is an endocrine gland, secreting primarily cortisol, aldosterone, and androgenic steroids, all of which are derived from cholesterol esters. This contributes to the high lipid content that is characteristic of most adrenal cortical adenomas.

The medulla is derived from the neural crest and secretes epinephrine and norepinephrine.

Adrenal physiology is controlled by an elaborate interaction between the hypothalamus, pituitary, and adrenals. Stress results in the release of epinephrine and cytokines, which cause the pituitary to secrete adrenocorticotrophic hormone (ACTH), which in turn stimulates the adrenals to secrete cortisol. Cortisol and epinephrine are often regarded as the "stress hormones." Elevated serum cortisol has a suppressing effect on the hypothalamus and pituitary, reducing further release of ACTH.

Cushing syndrome, due to excess cortisol, is characterized by truncal obesity, hirsutism, amenorrhea, hypertension, weakness, and abdominal striae. It is usually caused by adrenal hyperplasia (75-80% of cases) but may also result from an adrenal adenoma (20-25%), adrenal carcinoma (< 5%), or exogenous corticosteroid medication.

Conn syndrome is the eponym sometimes used for excess aldosterone and is characterized by hypertension, hypokalemia, alkalosis, muscle weakness, and cardiac dysfunction. Unlike Cushing syndrome, Conn syndrome is usually caused by an adrenal adenoma (65-70%). Adrenal hyperplasia accounts for most of the remaining (25-30%) cases, while adrenal carcinoma is a rare cause.

Because patients with Cushing syndrome are symptomatic, the diagnosis of either pituitary or adrenal-induced cortisol excess is typically made biochemically, prior to imaging. Imaging (either MR or CT) performed after diagnosis usually easily localizes the pituitary or adrenal adenoma responsible for Cushing syndrome, thus obviating catheter angiography or other invasive tests. Conn adenomas, however, may be quite small, and traditional imaging may be problematic in the setting of elevated aldosterone.

Addison syndrome or disease refers to adrenal insufficiency and is characterized by hypotension, weight loss, and altered pigmentation. Among the causes of "slow onset" adrenal insufficiency are autoimmune disease, which is the most common cause in developed countries. Tuberculosis and other infections remain common causes of Addison syndrome in the developing world, while metastatic disease and AIDS are encountered in all populations.

The most common cause of abrupt onset adrenal insufficiency is adrenal hemorrhage, which may result from sepsis, shock, anticoagulation, or vasculitis. Abrupt withdrawal of corticosteroid medication and postpartum pituitary necrosis, known as Sheehan syndrome, also result in acute adrenal insufficiency. Prompt recognition and treatment of these conditions is critical.

Imaging Techniques and Protocols

Adrenal Protocol, CT

NECT is often sufficient to diagnose an adrenal adenoma, seen as a homogeneously low-attenuation (< 10 HU) nodule. If the lesion is indeterminate or if only a contrast-enhanced scan was obtained, determine the attenuation of the mass on CECT, then repeat a series of thin sections through the adrenals after a delay of 15 minutes and measure the attenuation of the adrenal mass. Calculate the relative or absolute washout.

Adrenal Protocol, MR

Lipid-rich adrenal adenomas can be diagnosed with confidence on T1WI GRE sequences that include both in-phase and opposed-phase imaging. Loss of signal from an adrenal mass on opposed-phase imaging is essentially diagnostic of adenoma. Lipid-poor adenomas may remain indeterminate on MR and are best evaluated by adrenal protocol CT.

Approach to the Adrenal Mass

Is it an Adenoma?

Adrenal adenomas are extremely common, though most are "nonfunctional" or, more accurately, "not hyperfunctioning." It is estimated that some 2% of the general population has an adrenal adenoma, making this the most common cause of an adrenal mass, even in patients with a known extraadrenal malignancy. The high prevalence of adrenal adenomas in the general population, and large scale studies showing that the vast majority of adrenal "incidentalomas" are benign findings of no clinical significance, have prompted some authors to advocate a much more conservative approach to the small incidental adrenal lesion. Nonetheless, most professional societies continue to advocate an imaging and clinically intensive approach to incidental adrenal lesions depicted at CT. Most adenomas are rich in intra- and intercellular lipid, which creates a characteristic appearance as a homogeneous near-water-density (-5 to +10 HU) on a nonenhanced CT scan. The same lipid-rich adenomas have a characteristic appearance on MR as well. For the minority of adrenal adenomas that lack sufficient lipid, the best imaging test is an "adrenal protocol" CT scan. Adenomas are distinguished from metastases and most other adrenal masses by their characteristic "washout" of contrast, quickly returning to near baseline attenuation after the IV administration of contrast medium.

It's not an Adenoma, What is it?

While other adrenal masses may share overlapping imaging characteristics, it is usually not problematic to make an accurate diagnosis when taking into account the imaging features and clinical and laboratory data in a particular patient. Both pheochromocytoma and metastases may result in a large, heterogeneous adrenal mass; however, correlation with the medical history, endocrine lab, and physical findings will usually obviate extensive or expensive additional imaging evaluation.

Are all Fat-Containing Adrenal Masses Adenomas?
Adenomas rarely contain macroscopic fat.

While the lipid in adrenal adenomas is microscopic fat, the presence of macroscopic deposits of fat characterizes a less common adrenal neoplasm, the myelolipoma (not to be confused with a renal angiomyolipoma, which is different in every regard). Myelolipoma is an uncommon benign adrenal neoplasm that can usually be diagnosed with confidence on CT or MR by the presence of mature, macroscopic fat deposits, with or without calcification and soft tissue elements.

How Should We Image for Suspected Pheochromocytoma?
Before initiating an expensive imaging work-up, it is recommended to do the appropriate history, physical exam, and appropriate biochemical screening. In most patients, this will establish the presence or absence of this tumor.

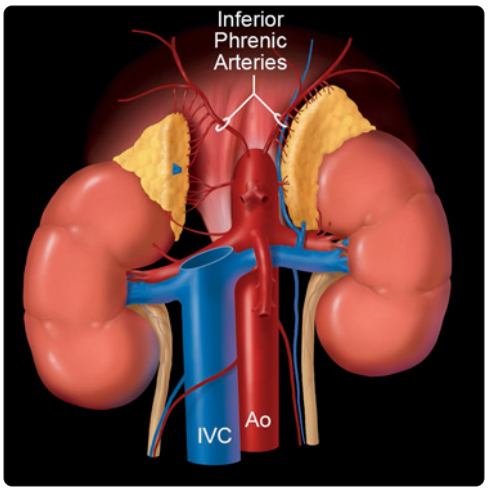
Either CT or MR will easily detect most symptomatic pheochromocytomas, as these are typically 3-5 cm in diameter. While the IV administration of contrast media is often not necessary due to the large size of these lesions, it is safe to do so if needed as there have been no reported adverse effects of IV administration of nonionic contrast media in patients with pheochromocytoma.

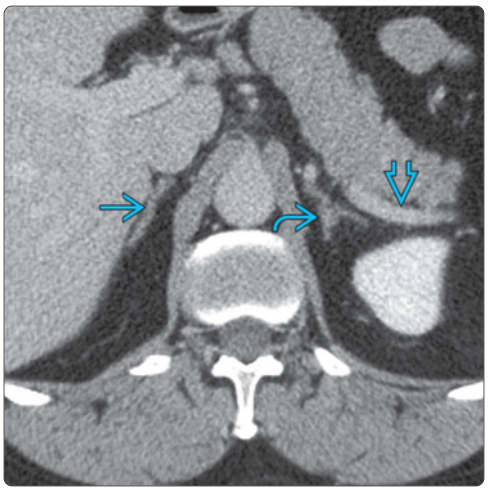
How Should We Image for Suspected Adrenal Metastases?
While the adrenal glands are common sites of metastatic disease (and lymphoma), they are rarely the only site. Remember that an isolated small adrenal lesion in a cancer patient is still most likely to represent an adrenal adenoma, especially if it is of homogeneous low density.

PET/CT is being used more frequently in the staging of patients with cancer and lymphoma, and most adrenal metastases are FDG avid. The expense of PET/CT makes it more appropriate to use as part of an approved cancer staging evaluation, rather than as a specific test to further evaluate an adrenal lesion that was detected by another imaging modality.

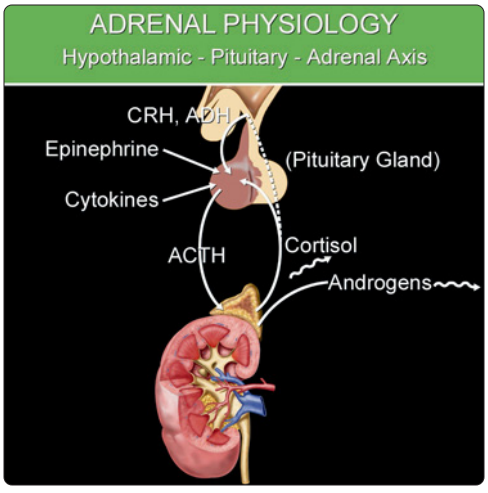
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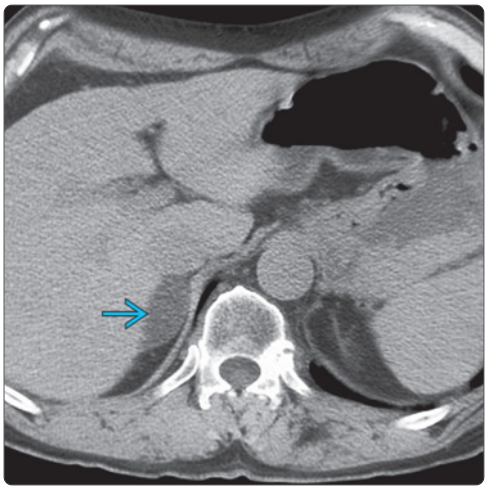
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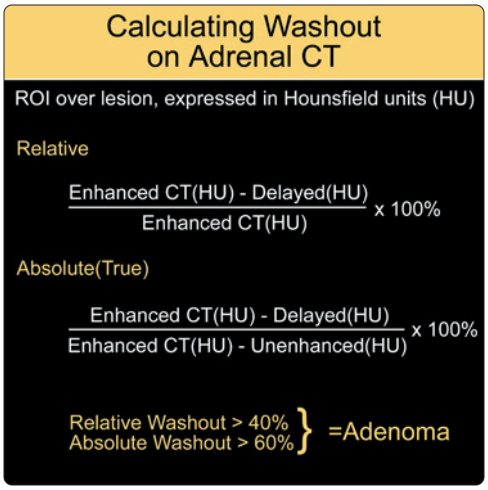
(Left) *Graphic shows typical adrenal anatomy. Note the multiple arterial sources. The right adrenal vein drains directly into the IVC, while the left enters the renal vein. Ao = aorta.* **(Right)** *The right adrenal gland abuts the IVC and lies lateral to the right diaphragmatic crus and medial to the liver. The left adrenal gland usually lies ventral to the upper pole of the left kidney and behind the splenic vein. The left adrenal appears as an inverted "Y," while the right is more like an inverted "V."*

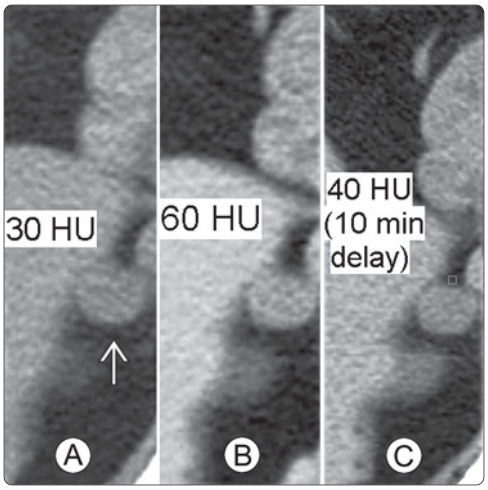




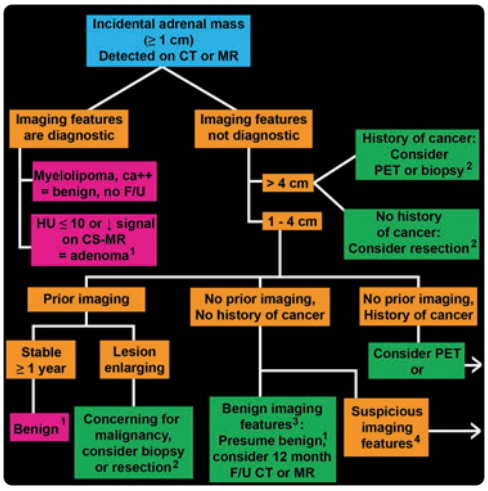
(Left) *Graphic shows stimulation of the anterior pituitary by epinephrine and cytokines or by hypothalamic release of corticotropin-releasing hormone (CRH) or antidiuretic hormone (ADH), resulting in the release of ACTH, causing the adrenal secretion of cortisol. Excess circulating cortisol inhibits further pituitary secretion.* **(Right)** *Axial NECT shows an incidental low-attenuation right adrenal mass, which measures < 10 HU. This feature has > 90% specificity for the diagnosis of a lipid-rich adenoma.*

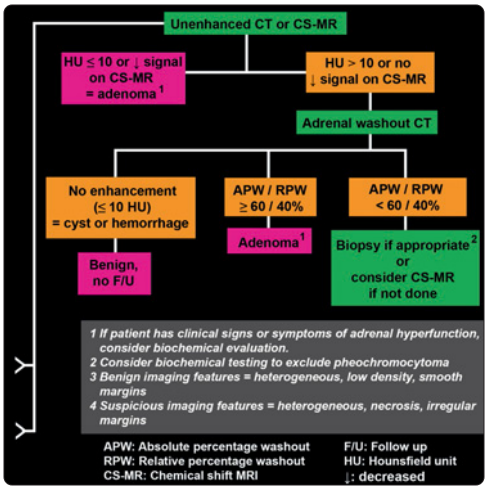
(Left) *Dedicated washout adrenal CT may be performed for indeterminate (> 10 HU) adrenal lesions at NECT to identify potential lipid-poor adenomas. The graph shows the formula for calculating adrenal washout on CT. Web-based calculators are readily available. ROI = region of interest.* **(Right)** *Adrenal CT protocol is shown. NECT (A) shows an adrenal mass ➔ measuring 30 HU. The mass measures 60 HU on CECT (B) and 40 HU on delayed CT (C). The absolute washout of 67% is diagnostic of a lipid-poor adenoma.*



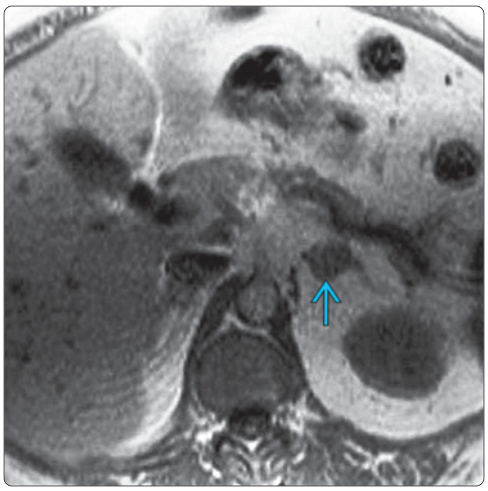


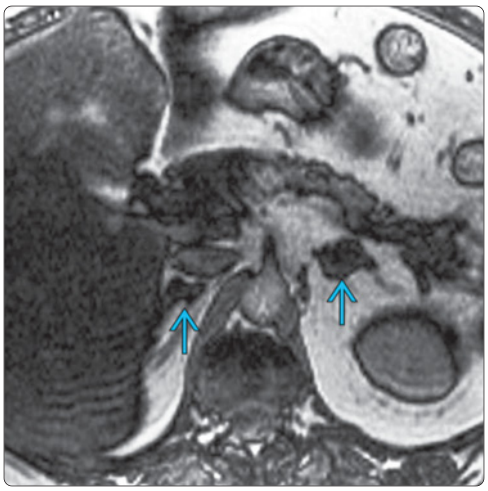
(Left) *Graph shows an imaging approach to the incidental adrenal mass. History and prior studies are crucial. Most adrenal lesions are benign although characterization may be critical in the oncology population. F/U = follow-up.* **(Right)** *Assessment of cortisol or epinephrine excess is also key for adrenal mass characterization (Adapted with permission L. Berland, MD, JACR.)*

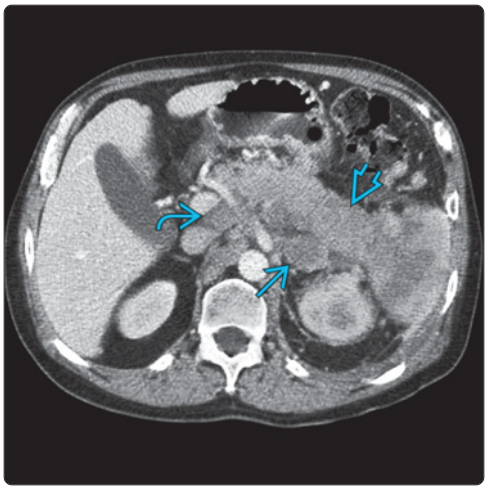


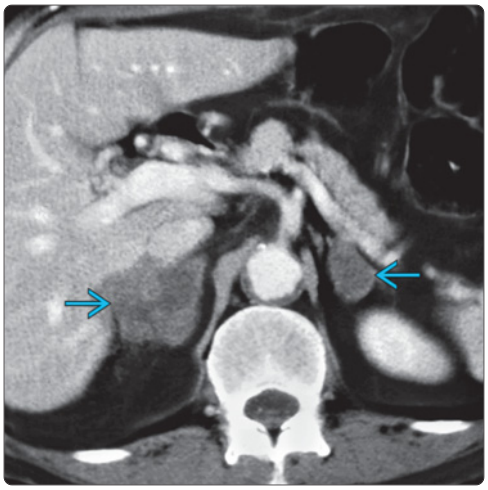


(Left) *On this in-phase T1W GRE image in a patient with bilateral adrenal nodules or small masses, only the left adrenal nodule is evident ⇨.* **(Right)** *On opposed-phase T1WI GRE in the same patient, there is signal dropout from both adrenal nodules ⇨, indicating a mixture of lipid and water protons. This is diagnostic of lipid-rich adenomas.*

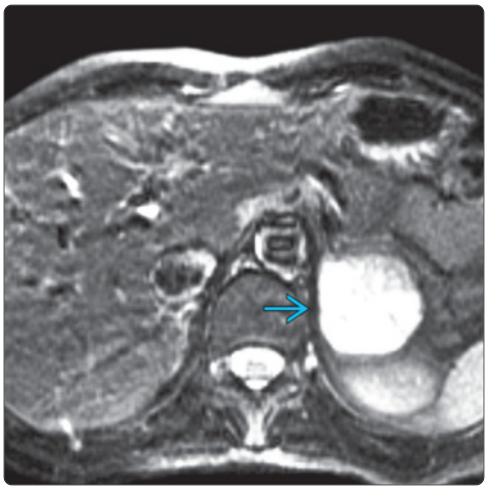


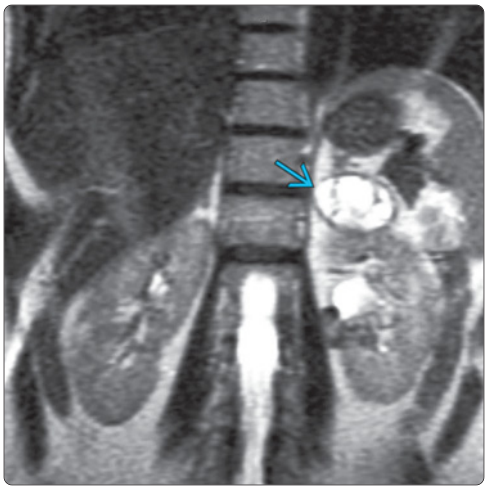




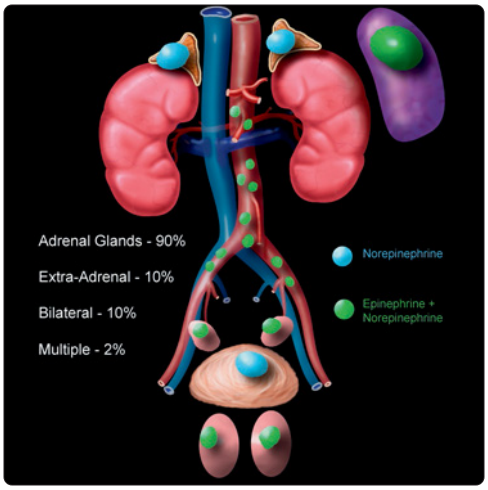


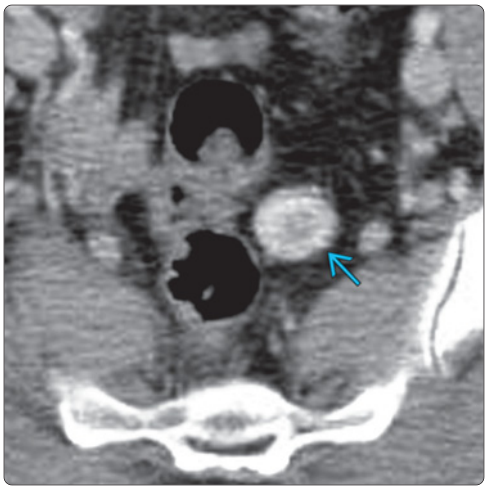
(Left) *Axial CECT in a patient with adrenal (and disseminated) lymphoma shows a left adrenal mass as well as soft tissue density masses that infiltrate the pancreas, spleen, and nodes.* **(Right)** *CECT in a patient with adrenal metastases and esophageal carcinoma shows bilateral heterogeneous lobulated adrenal masses. PET/CT was performed prior to planned esophagectomy. These lesions were FDG avid, resulting in a cancellation of the planned curative esophagectomy.*





(Left) *Axial T2WI MR in a patient with pheochromocytoma shows a large left adrenal mass that is heterogeneously hyperintense on T2WI and was hypointense on T1WI. The appearance is typical, although not pathognomonic, of a pheochromocytoma. The diagnosis was confirmed at resection.* **(Right)** *Coronal T2WI in the same patient shows the heterogeneously hyperintense mass, typical of a pheochromocytoma.*





(Left) *Graphic indicates potential sites for pheochromocytoma (blue) and paraganglioma (green). Note the organ of Zuckerkandl at the aortic bifurcation.* **(Right)** *Axial CECT shows a hypervascular mass in the pelvis. This was a paraganglioma with clinical symptoms identical to those of pheochromocytoma.*

Adrenal Tuberculosis and Fungal Infection

KEY FACTS

IMAGING

- Bilateral > unilateral adrenal involvement
- Acute
  - Homogeneous or heterogeneous masses
  - Mild to marked symmetrical enlargement of adrenal glands with preserved contour
  - Central areas of low attenuation (caseous necrosis)
  - Look for enlarged abdominal nodes
- Chronic
  - Atrophic glands with calcifications

TOP DIFFERENTIAL DIAGNOSES

- Adrenal adenoma
  - Mass with low attenuation (< 10 HU), usually unilateral
  - Contrast washout kinetics helpful for diagnosis
- Pheochromocytoma
  - > 3 cm, hypervascular mass
  - Usually clinically evident
  - May be bilateral (consider multiple endocrine neoplasia)
- Adrenal metastases and lymphoma
  - Clinical history paramount
  - Often in setting of widespread metastases (or adenopathy)
- Adrenal hemorrhage
  - Clinical scenario: Shock or sepsis
  - High-attenuation adrenal masses: Adreniform shape maintained

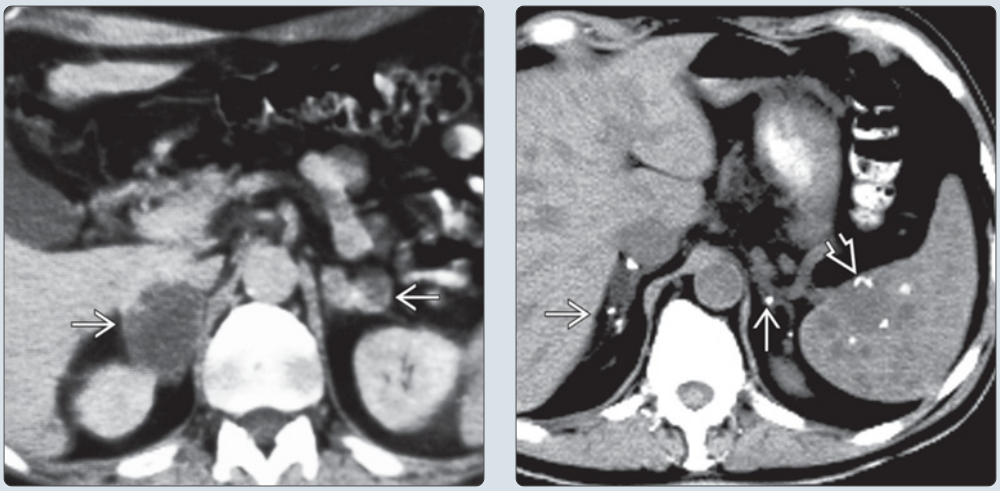
PATHOLOGY

- Tuberculosis
  - Most common in underdeveloped countries
- Histoplasmosis
  - Most common in southeastern, south-central USA

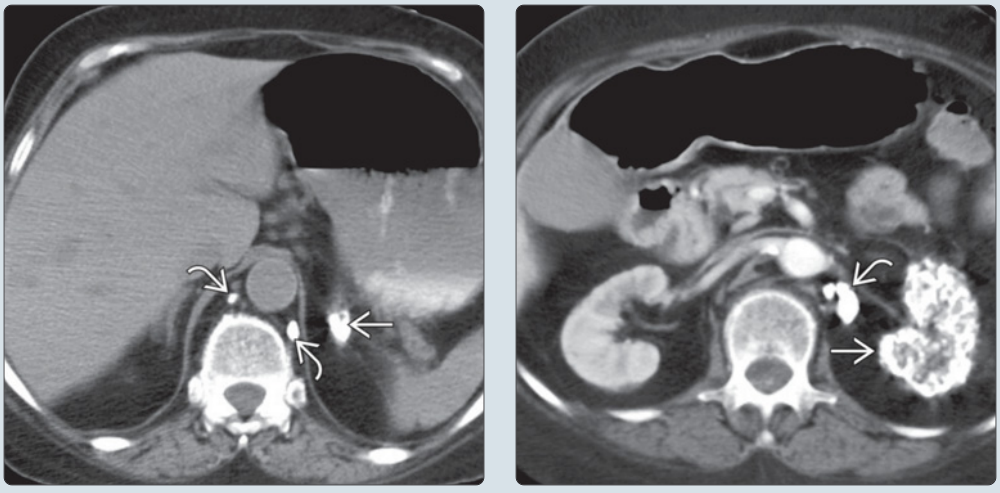
CLINICAL ISSUES

- May cause adrenal insufficiency (most common cause in undeveloped countries)

(Left) *Axial CECT shows bilateral adrenal masses, including 1 large mass in the right adrenal that shows areas of low density due to necrosis or degeneration in this patient with an acute or subacute blastomycosis.* **(Right)** *Axial NECT shows calcifications in both adrenal glands. Also note the calcified granulomas in the spleen, along with low-density splenic foci that may represent noncalcified granulomas. This may be the result of prior disseminated histoplasmosis.*



(Left) *Axial NECT shows calcified foci in the retroperitoneal lymph nodes and the left adrenal gland, incidental findings in this elderly woman. The right adrenal gland is normal. It is unusual to have only unilateral granulomatous infection of the adrenals.* **(Right)** *Axial CECT in the same patient shows that the left kidney is heavily calcified and nonfunctional, a typical "putty" kidney due to infection by tuberculosis. Note the additional calcified nodes.*



IMAGING

General Features

- Best diagnostic clue
  - Adrenal calcification (chronic phase)
- Location
  - Bilateral > unilateral adrenal involvement

CT Findings

- Acute
  - Homogeneous/heterogeneous masses
  - Symmetrical adrenal enlargement; preserved adreniform shape
  - Central areas of low attenuation (caseous necrosis)
  - Peripheral enhancement
  - Calcifications
  - Regional adenopathy
- Chronic: Atrophic glands with calcifications

MR Findings

- T1WI
  - Signal intensity similar to spleen
  - Granulomas show little enhancement
- T2WI
  - Nonspecific ↑ T2 signal adrenal masses

Ultrasonographic Findings

- Grayscale ultrasound
  - Homogeneous to heterogeneous, hypoechoic masses (acute)
  - Shadowing suprarenal calcifications (chronic)

Nuclear Medicine Findings

- Markedly FDG-avid adrenal masses during acute phase
  - May mimic metastases
  - Uptake diminishes with successful treatment

Imaging Recommendations

- Best imaging tool
  - CT

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- Low-attenuation (< 10 HU) mass on NECT, usually unilateral
- Enhancing mass that "de-enhances" rapidly

Pheochromocytoma

- Hypervascular mass > 3 cm
- Diagnosis with clinical history (hypertension) and ↑ urinary catecholamines

Adrenal Metastases and Lymphoma

- Metastases
  - Unilateral or bilateral, invasive, enhancing masses
- Lymphoma
  - Usually bilateral, triangular masses
  - Clinical history, and adenopathy elsewhere permit diagnosis

Adrenal Hemorrhage

- Hyperdense, heterogeneous, unilateral or bilateral mass(es)
- Setting: Following shock or sepsis

PATHOLOGY

General Features

- Etiology
  - Tuberculosis (most common in underdeveloped countries)
  - Histoplasmosis (most common in southeastern, south-central USA)
    - Blastomycosis (South America, North America)
    - Coccidioidomycosis
    - Cryptococcosis
    - Paracoccidioidomycosis
  - Risk factor: Immunosuppression
- Associated abnormalities
  - Pulmonary or other disseminated infections

Gross Pathologic & Surgical Features

- Calcifications, caseous necrosis

Microscopic Features

- Granulomatous or fungal organisms

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, night sweats, weight loss, lethargy, weakness
  - May cause adrenal insufficiency (Addison syndrome)
- Diagnosis
  - Tuberculosis: Positive purified protein derivative skin test
  - Percutaneous aspiration with cytology (e.g., acid-fast bacilli staining) for definitive diagnosis

Natural History & Prognosis

- May cause adrenal insufficiency (most common cause in undeveloped countries)

Treatment

- Antituberculous or antifungal medications

DIAGNOSTIC CHECKLIST

Consider

- Infection in patients with acute adrenal insufficiency

Image Interpretation Pearls

- Bilateral enlargement with preserved adreniform shape in acute infection
- Adrenal calcification in chronic phase

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Adrenal Hyperplasia

KEY FACTS

IMAGING

- Enlarged limbs of 1 or both adrenal glands > 10 mm width on CT
  - Glands may appear normal sized or multinodular
  - Adrenal gland shape is maintained
- Abdominal CT best for imaging adrenals
  - Contiguous < 3 mm axial images with multiplanar reformations
- Congenital adrenal hyperplasia
  - Causes most cases of adrenogenital syndrome (increased secretion of androgenic steroids)
  - Should be diagnosed in neonatal period; not an imaging diagnosis
- Cushing syndrome (hypercortisolism)
  - Usually caused by pituitary adenoma
- Conn syndrome (primary hyperaldosteronism)
  - 80% of cases due to adrenal adenoma

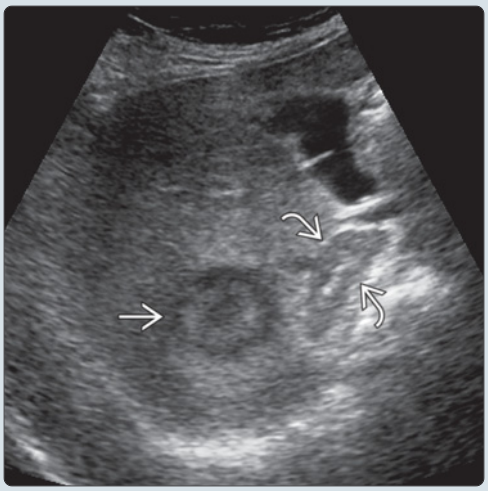
TOP DIFFERENTIAL DIAGNOSES

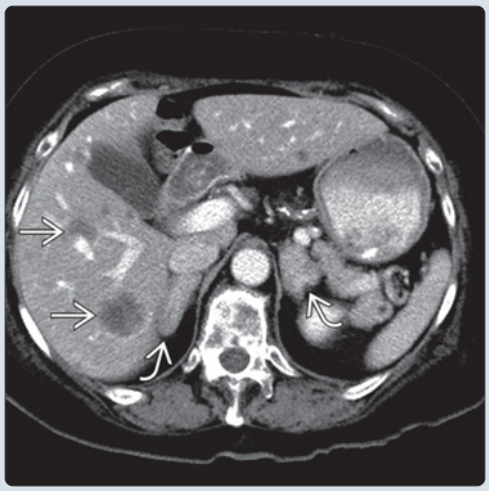
- Adrenal adenoma
- Adrenal metastases and lymphoma
- Adrenal hemorrhage
- Pheochromocytoma

DIAGNOSTIC CHECKLIST

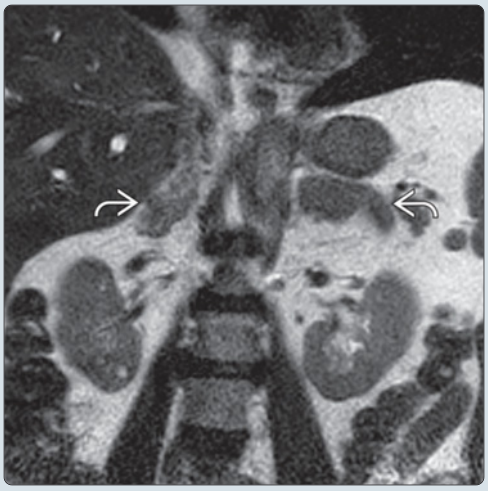
- Correlate clinical, biochemical, and imaging findings
- "Prominent" adrenals may be normal or temporary response to stress
- Multinodular hyperplasia is difficult to distinguish from adenoma by CT
- Most other causes of bilateral disease cause spherical masses in adrenals

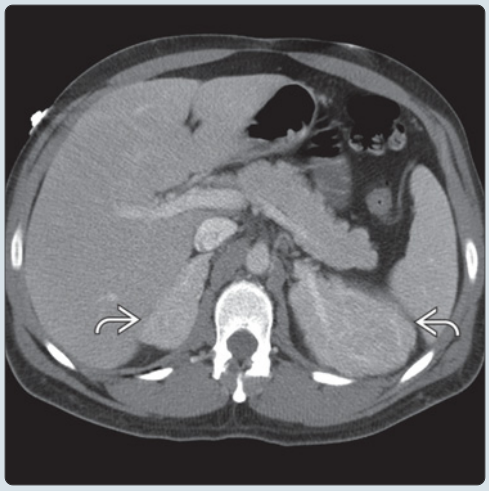
(Left) *Right upper quadrant ultrasound performed on a cushingoid patient shows a right lobe metastasis and a diffusely enlarged right adrenal gland.* **(Right)** *CECT of the same patient shows diffusely enlarged adrenal glands and multiple hepatic metastases. Work-up for ectopic ACTH production revealed a small cell lung primary. Symmetric enlargement of the adrenal glands is the classic imaging manifestation of ACTH-dependent Cushing syndrome and adrenal hyperplasia.*





(Left) *T2 TSE of a patient with Cushing syndrome shows enlarged, nodular adrenal glands. A pituitary MR was negative, and an endocrine work-up ultimately revealed ectopic, but imaging occult, ACTH production. Although ACTH-dependent hyperplasia typically results in diffusely enlarged adrenals, the adrenals may also appear nodular or normal at imaging.* **(Right)** *CECT of a young male shows massively enlarged adrenals, a characteristic feature of congenital adrenal hyperplasia (CAH).*





TERMINOLOGY

Definitions

- Bilateral adrenal gland hyperfunction ± enlargement

IMAGING

General Features

- Best diagnostic clue
  - Enlarged limbs of 1 or both adrenal glands > 10 mm width on CT
- Key concepts
  - Width of adrenal gland limbs > 10 mm is consistent with (but not diagnostic of) hyperplasia
  - Glands may appear normal sized or multinodular
  - Adrenal gland shape is maintained
- **Bilateral adrenocortical hyperplasia: Subtypes**
  - Congenital adrenal hyperplasia (CAH)
  - Adrenocorticotrophic hormone (ACTH)-dependent Cushing syndrome
  - Primary pigmented nodular adrenocortical hyperplasia (PPNAH) or dysplasia (PPNAD)
  - ACTH-independent macronodular adrenocortical hyperplasia (AIMAH)
- **Congenital adrenal hyperplasia**
  - Autosomal recessive; due to enzyme deficiencies
    - Usually 21-hydroxylase deficiency
  - Decreased secretion of cortisol, aldosterone, or both, with compensatory increase in ACTH
  - Most common cause of **adrenogenital syndrome** (increased secretion of androgenic steroids)
    - Usually presents in children, rarely in adults
    - Virilization of infant girls; precocious puberty of boys
    - Should be diagnosed in neonatal period; not imaging diagnosis
    - **Markedly elevated levels of 17-hydroxyprogesterone**
  - Associated with testicular and ovarian "tumors" that arise from ectopic adrenal cortical rests
- **Cushing syndrome (hypercortisolism)**
  - ACTH-dependent (80-85%)
    - Usually caused by **pituitary (not adrenal) adenoma**
      - Referred to as Cushing disease
    - Ectopic ACTH-secreting tumors (15%)
    - Hypothalamic tumor → ↑ corticotropin releasing factor → ↑ pituitary ACTH (rare)
  - ACTH-independent Cushing syndrome (15-20%)
    - Adrenal adenoma or, less often, carcinoma
    - Rarely caused by PPNAH or PPNAD & AIMAH
- **Conn syndrome (primary hyperaldosteronism)**
  - 20% of cases are due to adrenal gland hyperplasia
  - 80% of cases are due to **adrenal adenoma**
- Bilateral adrenal medulla hyperplasia
  - Rare: Nodular or diffuse
  - May cause hypertensive symptoms similar to pheochromocytoma
  - Associated with multiple endocrine neoplasia (MEN2B and MEN3), duodenal carcinoid

CT Findings

- ACTH-dependent Cushing syndrome (80-85% of cases)
  - Adrenal glands often symmetrically enlarged
  - Limbs of adrenal gland > 10 mm supports this diagnosis
  - Usually both adrenal glands involved
  - No discrete mass or nodule seen as rule
    - Cushing syndrome can show nodular hyperplasia
  - Up to 30% of cases may have normal adrenal glands
- Conn syndrome (primary hyperaldosteronism)
  - Usually caused by adrenal adenoma, not hyperplasia
  - Adrenal glands may show nodular hyperplasia
- PPNAH or PPNAD
  - Normal-sized adrenal glands
  - Small discrete nodules between 2-5 mm in size
  - Moderate enhancement of nodular glands
- AIMAH
  - Massively enlarged nodular adrenal glands
  - Normal adrenal shape is retained
  - Adrenal limb width and nodule size is 30 mm
  - Marked enhancement of hyperplastic nodular glands, predominantly at periphery

MR Findings

- Adrenal hyperplasia due to Cushing, Conn syndromes
  - Adds no new information compared to CT for adrenals
- MR demonstrates pituitary adenoma, if present

Nuclear Medicine Findings

- NP-59 traditionally advocated for adrenal hyperplasia imaging
- Ga-88 DOTATATE and In-111 Octreotide utilized for ectopic ACTH-producing tumors
- Marked uptake reported with FDG-PET/CT of AIMAH

Imaging Recommendations

- Abdominal CT best tool for imaging adrenals
  - Contiguous < 3 mm axial images with multiplanar reformations

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- Intracellular lipid makes CT density -20 to +10 HU
- Focal mass, not diffuse enlargement
- Uncommonly bilateral

Adrenal Metastases and Lymphoma

- Adrenal metastases
  - Heterogeneous enhancing mass
  - Loss of adrenal shape; uni- or bilateral
- Adrenal lymphoma
  - Usually spread to adrenal gland from retroperitoneal tumor
  - Bilateral primary lymphoma (non-Hodgkin) can simulate hyperplasia; usually more mass-like

Adrenal Hemorrhage

- Adults
  - Septicemia, burns, trauma, severe stress, or hypotension
  - Excessive anticoagulation, thrombocytopenia
  - Disseminated intravascular coagulation, antiphospholipid antibody syndrome
- Neonates: Meningococci, pneumococci, & gonococci
- Adrenal hematomas usually appear round in shape

- Some poorly marginated with periadrenal stranding
- CT
  - Acute: High-density fluid collection (40-60 HU)
  - Chronic: Low density (clot of 20-30 HU)

Pheochromocytoma

- Tumor of adrenal medulla and usually > 3 cm
- Highly vascular; prone to hemorrhage & necrosis
- Bilateral spherical adrenal tumors in MEN2A and MEN2B syndromes

PATHOLOGY

General Features

- Etiology
  - Congenital (due to enzyme deficiencies)
  - Cushing disease: Usually pituitary adenoma → ↑ ACTH → adrenal gland hyperplasia
  - Cushing syndrome: Uncommonly adrenal adenoma/carcinoma, ectopic ACTH production
    - Cushing syndrome due to exogenous steroids is not an imaging diagnosis
  - Conn syndrome (primary hyperaldosteronism)
    - ↑ aldosterone causes ↑ retention of sodium and loss of potassium at renal tubular level
    - ↑ sodium → water retention → more extracellular volume → hypertension → less renin production
    - Hypokalemia causes hypokalemic alkalosis
- Associated abnormalities
  - Pituitary adenoma in ACTH-dependent hyperplasia
  - PPNAH associated with Carney complex
    - Cushing disease, spotty skin pigmentation
    - Cutaneous and cardiac myxomas
    - Sertoli cell tumors of testis in males
    - Multiple myxoid fibroadenomas of breast, females
  - Adrenal medulla hyperplasia: MEN2B or MEN3, duodenal carcinoid

Gross Pathologic & Surgical Features

- Bilateral diffusely enlarged adrenal glands
- Nodular, macronodular, or pigmented nodular glands

Microscopic Features

- Thickened zona glomerulosa, fasciculata, reticularis
  - Fasciculata may show excess lipid, lipid-depleted, or atypical cells with hyperchromatic nuclei in nodules

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic incidental CT finding
  - Congenital adrenal hyperplasia
    - Neonatal or childhood evidence of virilization ± electrolyte disturbances
  - Cushing syndrome (hypercortisolism)
    - Moon facies, truncal obesity, buffalo hump
  - Conn syndrome (hyperaldosteronism)
    - Hypertension and hypokalemia
- Lab data: ↑ levels of ACTH, cortisol, aldosterone, or other steroids (in congenital syndrome)
- Diagnosis: Clinical, biochemical, imaging, histology

Demographics

- Age
  - Depends on clinical syndrome
    - Congenital adrenal hyperplasia: Neonatal period, though patients with CAH may live to adulthood with improved treatment regimens
    - Cushing syndrome: 25-40 years
    - Conn syndrome: 30-50 years
- Gender
  - Cushing syndrome (M:F = 1:5)
    - Conn syndrome (M:F = 1:2)

Natural History & Prognosis

- Complications: Untreated adrenal crisis → death
- Prognosis: Usually good with treatment

Treatment

- Congenital adrenal hyperplasia: Glucocorticoid and mineralocorticoid supplementation
- Adrenal hyperplasia without symptoms: Follow-up
- Adrenal hyperplasia with symptoms: Surgical resection of pituitary adenoma or source of hormone

DIAGNOSTIC CHECKLIST

Consider

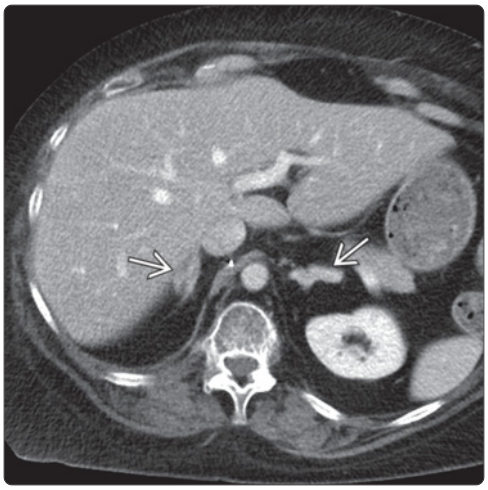
- Correlate clinical, biochemical, and imaging findings
  - "Prominent" adrenals may be normal or temporary response to stress

Image Interpretation Pearls

- Enlarged limbs of adrenal glands (> 10 mm) on CT
- Multinodular hyperplasia difficult to distinguish from adenoma by CT
- Most other causes of bilateral disease cause spherical masses in adrenals

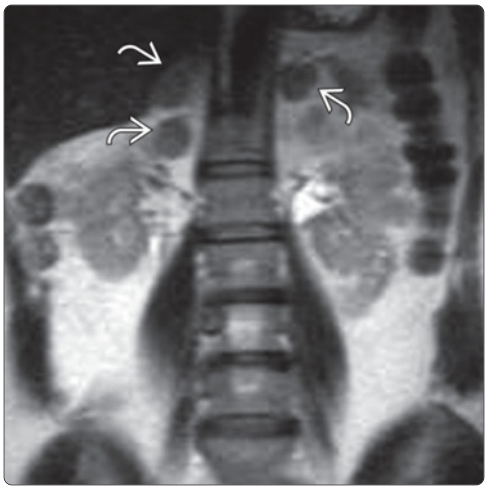
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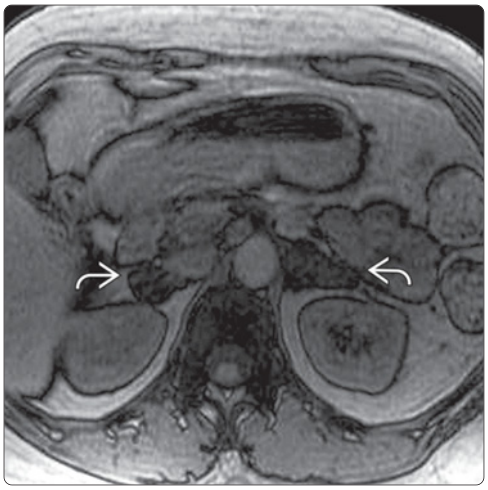
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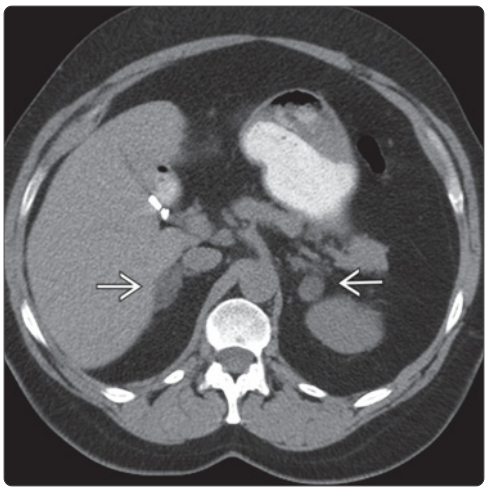


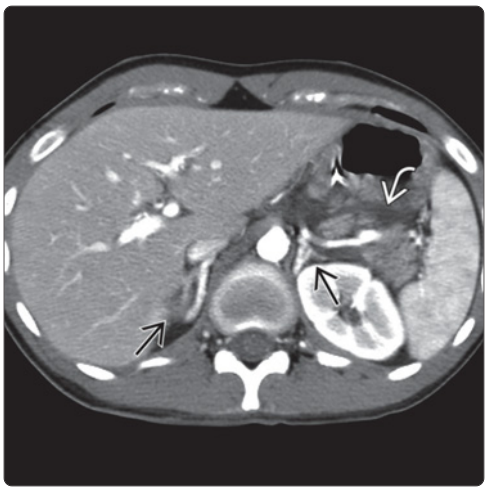
(Left) *CECT of a young male with Cushing syndrome shows hyperstimulated adrenals due to ectopic (nonpituitary) ACTH production. Lung carcinoma is the most common ectopic ACTH source, although other sources include thymic, bowel, and pancreatic neuroendocrine tumors.* **(Right)** *CECT of a 44-year-old man with CAH shows enlarged adrenals. Improved management with gluco/mineralocorticoid replacement has increased the lifespan of these patients. (Courtesy S. Challinor, MD.)*





(Left) *Coronal T2 TSE of a cushingoid patient shows thickened, nodular, adrenal glands.* **(Right)** *Out-of-phase T1WI MR of the same patient shows signal suppression of enlarged, nodular adrenal glands. The work-up for causes of cortisol excess (exogenous steroids, ACTH-dependent and -independent Cushing syndrome) includes patient history, a battery of lab tests, and, potentially, imaging. In this case, cortisol excess was due to a pituitary adenoma.*





(Left) *NECT of an elderly female with Cushing syndrome, a suppressed ACTH, and abnormal dexamethasone suppression shows enlarged, nodular adrenal glands. Laparoscopic adrenalectomy confirmed ACTH-independent macronodular hyperplasia.* **(Right)** *CECT of a young male in shock, after blunt trauma, shows prominent hyperenhancing adrenals and peripancreatic edema. Adrenal hyperenhancement is a feature of the hypoperfusion complex. Mild adrenal enlargement may also be seen with systemic stress.*

Adrenal Insufficiency

KEY FACTS

TERMINOLOGY

- Addison disease
- Inadequate secretion of corticosteroids resulting from partial or complete destruction of adrenal glands

IMAGING

- Acute: Addisonian crisis, adrenal apoplexy
  - Heterogeneous, hyperdense, enlarged (hemorrhagic) adrenals
- Subacute
  - Enlarged glands with decreased density (caseation or necrosis)
- Chronic
  - Small, atrophic glands (autoimmune)
  - Dense, chunky calcifications (infection)

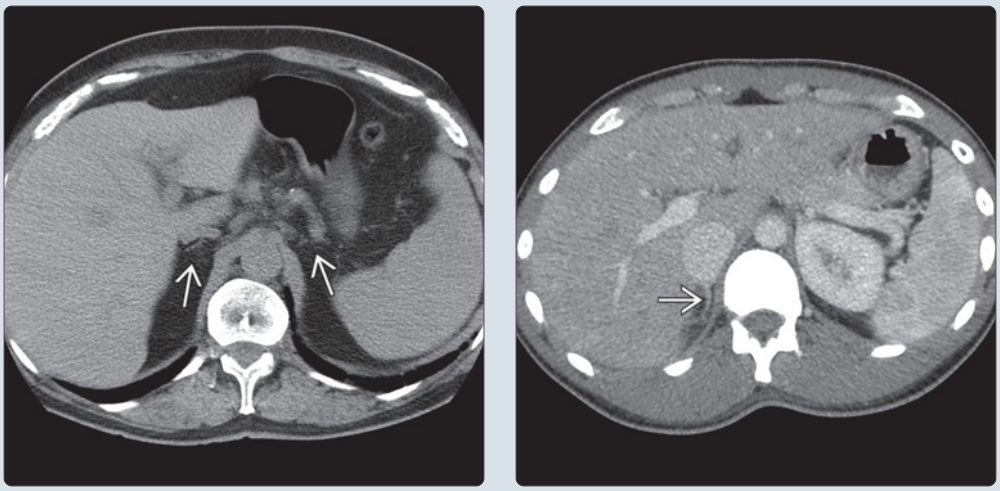
PATHOLOGY

- Idiopathic autoimmune disorders (80% of cases in developed countries)
- Granulomatous diseases: Tuberculosis (most common cause in underdeveloped nations), sarcoidosis
- Neoplasms: Metastases (e.g., lung, ovary, breast, kidney), lymphoma, leukemia
- Adrenal hemorrhage, necrosis, or thrombosis: Stress after surgery, shock, anticoagulation, sepsis (e.g., Waterhouse-Friderichsen syndrome), coagulation disorders, antiphospholipid syndrome
- Central adrenal insufficiency due to ↓ production/action of ACTH (from either pituitary or hypothalamic disease)

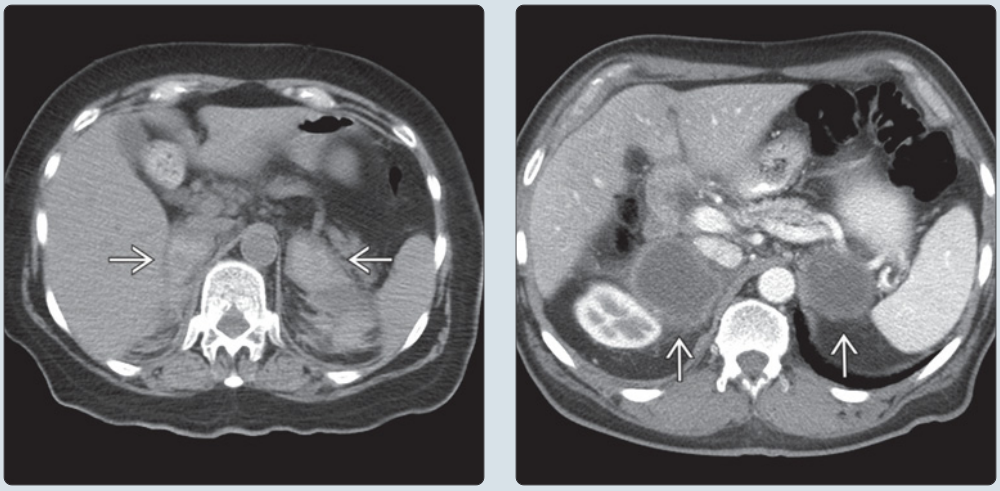
CLINICAL ISSUES

- Chemistry: Hyponatremia, hyperkalemia, azotemia, hypercalcemia, hypoglycemia

(Left) *NECT of a 71-year-old woman with autoimmune polyendocrine syndrome (adrenal insufficiency, hypothyroidism) shows diminutive adrenals ⇒. Interestingly, this patient also had a longstanding history of sarcoid, an additional cause for both primary and secondary (pituitary, hypothalamic) adrenal insufficiency.* **(Right)** *CECT of a young male with acute adrenal insufficiency shows a barely perceptible right adrenal gland ⇒. A serum cortisol after cosyntropin stimulation was undetectable.*



(Left) *NECT of an elderly female performed for evaluation of hypotension, hyponatremia, hypoglycemia and sepsis post knee replacement shows bilateral adrenal hemorrhage ⇒, a rare cause for acute adrenal insufficiency.* **(Right)** *CECT of an elderly male with lung carcinoma shows large necrotic adrenal metastases ⇒. Lab data (↓ Na, ↑ K) and an ACTH stimulation test confirmed profound adrenal insufficiency, which was treated with steroid maintenance therapy.*



TERMINOLOGY

Synonyms

- Addison disease

Definitions

- Inadequate secretion of corticosteroids resulting from partial or complete destruction of adrenal glands

IMAGING

General Features

- Location
  - Bilateral
- Other general features
  - Imaging findings depend on course
    - Acute, subacute (< 2 years), chronic

CT Findings

- Acute: Addisonian crisis, adrenal apoplexy
  - Heterogeneous, hyperdense, enlarged (hemorrhagic) adrenals
- Subacute
  - Enlarged glands with decreased density (caseation or necrosis)
- Chronic
  - Small, atrophic glands (autoimmune)
  - Dense, chunky calcifications (infection)

Imaging Recommendations

- Best imaging tool
  - CT

DIFFERENTIAL DIAGNOSIS

Adrenal Metastases and Lymphoma

- Focal masses, usually without diffuse enlargement

Adrenal Hemorrhage

- Heterogeneous, enhancing mass (40-80 HU)

Adrenal TB and Fungal Infection

- Unilateral or bilateral enlarged adrenal glands (acute)

Adrenal Adenoma

- May be bilateral

PATHOLOGY

General Features

- Primary adrenal insufficiency (disease intrinsic to adrenal cortex)
  - Idiopathic autoimmune disorders (80% of cases in developed countries)
  - Granulomatous diseases: Tuberculosis (most common cause in underdeveloped nations), sarcoidosis
  - Neoplasms: Metastases (e.g., lung, ovary, breast, kidney), lymphoma, leukemia
  - Adrenal hemorrhage, necrosis, or thrombosis: Stress after surgery, shock, anticoagulation, sepsis (e.g., Waterhouse-Friderichsen syndrome), coagulation disorders, antiphospholipid syndrome
  - Systemic fungal infections: Histoplasmosis (most common infection in southeastern, south-central USA), cryptococcosis, blastomycosis
  - Acquired immune deficiency syndrome (AIDS): Opportunistic infections (e.g., cytomegalovirus), neoplasms
- Central adrenal insufficiency: Impaired production or action of ACTH (pituitary or hypothalamic disease)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute: Fever, abdominal or back pain, hypotension, weakness
  - Chronic: Progressive lethargy, weakness, cutaneous pigmentation, weight loss
- Lab data
  - Chemistry: Hyponatremia, hyperkalemia, azotemia, hypercalcemia, hypoglycemia
  - Adrenocorticotrophic hormone (ACTH) stimulation test: Cortisol level fails to rise

Demographics

- Gender
  - Autoimmune type is 2x as common in women
- Epidemiology
  - 50 per 1 million persons

Natural History & Prognosis

- Complications: Shock, death (acute)
- Prognosis: Good if recognized and treated

Treatment

- Acute
  - Glucocorticoid therapy
  - Volume and electrolytes replacement
  - Correct any etiology (e.g., antibiotics for infections)
- Chronic
  - Glucocorticoid & mineralocorticoid replacement

DIAGNOSTIC CHECKLIST

Consider

- Bilateral adrenal masses may occur from infection, hemorrhage, or tumors ± adrenal insufficiency

Image Interpretation Pearls

- Cannot diagnose adrenal hyper- or hypofunction by imaging alone

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Adrenal Hemorrhage

KEY FACTS

IMAGING

- Relatively uncommon condition but potentially catastrophic event seen in patients of all ages
- More common in neonates than children and adults
- Nontraumatic hemorrhage (often bilateral): Causes are classified into 5 categories
  - Stress, hemorrhagic diathesis or coagulopathy, neonatal stress, adrenal tumors, idiopathic
- Manifest with adrenal insufficiency when 90% of adrenal tissue is destroyed
- Round or oval mass of high attenuation (50-90 HU)
  - Asymmetric enlargement of adrenal glands

PATHOLOGY

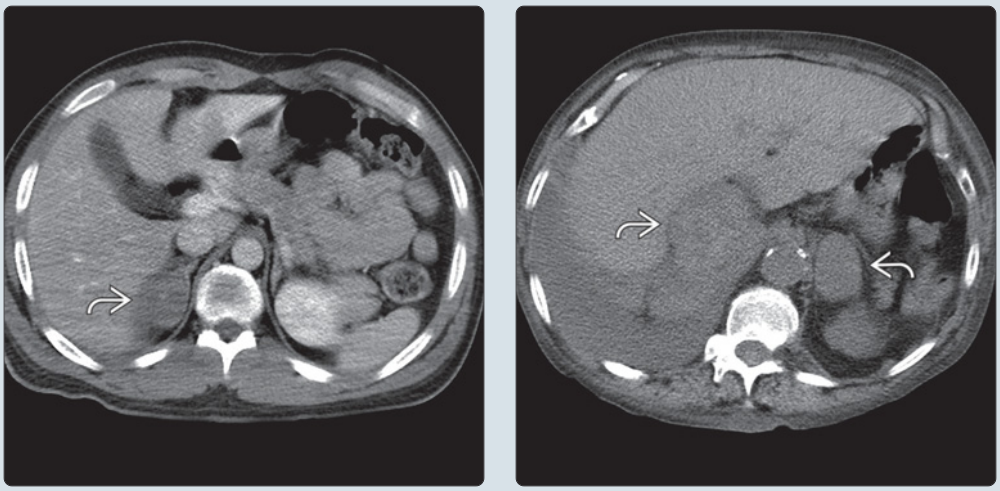
- Bilateral adrenal hemorrhage
  - Anticoagulation therapy (most common)
  - Stress: Surgery, sepsis, burns, hypotension, steroids
- Unilateral adrenal hemorrhage
  - Blunt abdominal trauma (right gland > left gland)
- Pathogenesis (nontraumatic)
  - Stress or adrenal tumor → increased ACTH → increased arterial blood flow + limited venous drainage → hemorrhage

CLINICAL ISSUES

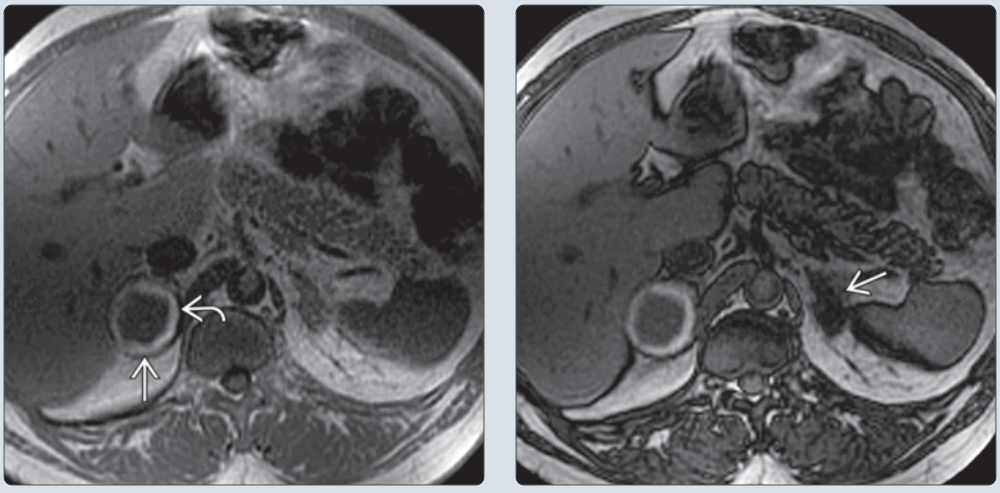
- Waterhouse-Friderichsen syndrome: Skin rash, cough, headache, dizziness, arthralgias, and myalgias
  - Due to septicemia, especially with meningococcal organisms

DIAGNOSTIC CHECKLIST

- Check for history of trauma, anticoagulant therapy, coagulopathies, malignancies, stress, adrenal tumor
- NECT: Hyperdense lesion within adrenal gland
- MR: Varied signal intensity based on age of hematoma



(Left) *CECT of a young male post blunt trauma shows an isolated right adrenal hematoma. Traumatic adrenal hemorrhage is typically associated with other abdominal visceral injuries and high injury severity scores. The presumed mechanism for this adrenal hemorrhage was an acute rise in adrenal venous pressure.* **(Right)** *NECT of a hypotensive female with known metastatic non-small cell carcinoma shows bilateral adrenal hematomas. Small preexisting adrenal metastases were shown on a prior staging CT.*



(Left) *T1WI in-phase MR of a patient on coumadin, with known bilateral adrenal adenomas and a 3-week history of right flank pain, shows a subacute to chronic right adrenal hematoma. Note the varying signal intensities of blood. Peripheral high T1 signal methemoglobin is rimmed by T1-hypointense hemosiderin.* **(Right)** *Out-of-phase image of the same patient shows signal suppression of a contralateral lipid-rich adrenal adenoma. The right adrenal hematoma resolved at follow-up CT.*

TERMINOLOGY

Abbreviations

- Adrenal hemorrhage (AH)

Definitions

- Hemorrhage within adrenal gland or tumor

IMAGING

General Features

- Best diagnostic clue
  - Hyperdense lesion within adrenal gland on NECT
- Key concepts
  - Relatively uncommon but potentially catastrophic condition in patients of all ages
    - More common in neonates than children and adults
  - Secondary to traumatic and nontraumatic causes
    - Traumatic more common than nontraumatic
    - May be unilateral or bilateral
  - Traumatic hemorrhage: Blunt abdominal trauma
    - Unilateral in 80% of cases: Right (85%), left (15%)
    - Bilateral in 20% of cases
  - Nontraumatic hemorrhage (often bilateral): Causes are classified into 5 categories
    - Stress, hemorrhagic diathesis or coagulopathy, neonatal stress, adrenal tumors, idiopathic
  - Bilateral AH in 15% of individuals who die of shock
  - Manifest with adrenal insufficiency when 90% of adrenal tissue is destroyed
  - Neonatal adrenal hemorrhage
    - Most common cause of adrenal mass in infancy
    - Usually seen during 1st week of life
    - Incidence ranges from 1.7-3% per 1,000 births
    - Infant adrenal is hypervascular and very large relative to adult

CT Findings

- Unilateral or bilateral adrenal hematomas
- ± associated adrenal or renal vein thrombosis
- Acute or subacute hematoma
  - Round or oval mass of high attenuation (50-90 HU)
  - Asymmetric enlargement of adrenal glands
  - Homogeneous and no enhancement with contrast
  - Inflammatory stranding of periadrenal fat
  - Thickening of adjacent diaphragmatic crura
  - ± periadrenal hemorrhage + perinephric extension
  - ± upper abdominal traumatic findings
    - Pneumothorax, hydropneumothorax, rib fracture
    - Contusion of lung, liver, spleen, or pancreas
- Chronic hematoma
  - Mass with hypoattenuating center (pseudocyst)
  - Lack of enhancement confirms cystic nature of mass
  - Calcifications (usually seen after 1 year in adults)
- Hematomas ↓ in size, attenuation over period of time
- ± underlying large adrenal mass (cyst, myelolipoma)
  - Intracystic or intratumoral hemorrhage
    - Adrenal cyst: Mass of water density
    - Adrenal myelolipoma: Heterogeneous fatty mass

MR Findings

- T1 and T2WI: Varied signal based on age of hematoma
- Acute hematoma (< 7 days after onset)
  - T1WI: Isointense or slightly hypointense
  - T2WI: Markedly hypointense
  - Due to high concentration of intracellular deoxyhemoglobin → T2 proton relaxation
- Subacute hematoma (7 days to 7 weeks after onset)
  - T1WI: Hyperintense
    - Due to free methemoglobin (Fe^{3+}), produced by oxidation of hemoglobin (Fe^{2+}) as hematoma ages
  - T2WI: Markedly hyperintense
    - Due to serum and clot lysis products
  - Large hematoma: Varied signal (irregular clot lysis)
    - Multilocular, fluid-fluid levels
- Chronic hematoma (> 7 weeks after onset)
  - T1 and T2WI: Hyperintense hematoma
    - Due to persistence of free methemoglobin
  - T1 and T2WI: Hypointense rim
    - Due to hemosiderin deposition in fibrous capsule
- Adrenal or renal vein thrombosis
  - Clot: ↑ signal on both T1 and T2WI
- Extension of thrombus into IVC can be seen on MR
- Underlying large adrenal mass (cyst or myelolipoma)
  - Varied signal based on content of mass lesion
- Gradient-echo imaging
  - Demonstrate "blooming" effect (magnetic susceptibility) due to hemosiderin deposition
    - Detect blood and monitor hemorrhage as it progresses from methemoglobin to hemosiderin
    - Useful in large hematomas, which exhibit slower clot evolution

Ultrasonographic Findings

- Grayscale ultrasound
  - Acute hematoma: Hyperechoic mass-like lesion
  - Subacute hematoma
    - Mixed echogenicity + central hypoechoic area
  - Chronic hematoma
    - Anechoic and cyst-like lesion; wall calcification

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- NECT: Lipid-rich adenoma (< 10 HU)
- T1WI out of phase
  - Marked signal "drop out" (lipid-rich adenoma)

Adrenal Metastases and Lymphoma

- Adrenal metastases
  - Lung cancer: Hemorrhagic, enhancing adrenal mass
  - Malignant melanoma: Hypervascular metastases
- Adrenal lymphoma
  - Primary (rare), secondary (non-Hodgkin, common)
  - Often bilateral; retroperitoneal disease usually seen

Adrenal Hyperplasia

- Adrenal glands are often symmetrically enlarged
- Width of adrenal gland limbs > 10 mm (diagnostic)
- No discrete mass or nodule seen as rule

Adrenal TB and Fungal Infection

- Granulomatous infection: Tuberculosis, histoplasmosis
- Adrenal calcification seen on CT imaging

Adrenal Carcinoma

- Rare, unilateral, invasive, and enhancing mass
- > 6 cm when initially diagnosed

PATHOLOGY

General Features

- Etiology
  - Bilateral adrenal hemorrhage
    - Anticoagulation therapy (most common)
    - Stress: Surgery, sepsis, burns, hypotension, steroids
    - Pheochromocytoma
    - Trauma: Rare
    - Metastases: Lung cancer and malignant melanoma
  - Unilateral adrenal hemorrhage
    - Blunt abdominal trauma (right gland > left gland)
    - Adrenal vein thrombosis with liver transplantation; adrenal tumor
  - Neonates
    - Difficult labor or delivery, ± renal vein thrombosis
    - Asphyxia or hypoxia; hemorrhagic disorders
    - Meningococcal septicemia (Waterhouse-Friderichsen syndrome)
  - Pathogenesis (nontraumatic)
    - Stress or adrenal tumor → increased ACTH → increased arterial blood flow + limited venous drainage → hemorrhage
    - Coagulopathies → venous stasis → thrombosis → hemorrhage
- Associated abnormalities
  - Adrenal or renal vein thrombosis
  - Adrenal tumor, hemorrhagic disorders

Microscopic Features

- Necrosis of all 3 cortical layers + medullary cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Nonspecific: Abdominal, lumbar, thoracic pain
  - Fever, tachycardia, hypotension
  - Acute adrenal insufficiency
    - Fatigue, anorexia, nausea, and vomiting
  - Acute abdomen
    - Guarding, rigidity, rebound tenderness
  - Confusion, disorientation, shock in late phase
  - Symptoms of associated underlying conditions (e.g., tumor)
  - Rarely asymptomatic; may be incidental finding (imaging)
  - Waterhouse-Friderichsen syndrome: Skin rash, cough, headache, dizziness, arthralgias, and myalgias
    - Due to septicemia, especially with meningococcal organisms
- Lab data
  - ↓ hematocrit or hemoglobin; ↑ WBC
  - Hyponatremia, hyperkalemia, and prerenal azotemia
  - ↓ serum cortisol, aldosterone, androgens, and ↑ ACTH

Demographics

- Age
  - Any age group
    - More common in neonates than children and adults
  - Nontraumatic (40-80 years), traumatic (20-30 years)
- Gender
  - M:F = 2:1
- Epidemiology
  - 15% of individuals who die of shock
  - 2% of orthotopic liver transplantation cases

Natural History & Prognosis

- Complications
  - Prerenal azotemia, adrenal abscess, shock
- Prognosis
  - Prognosis depends on etiology rather than extent of adrenal hemorrhage
  - Unilateral adrenal hemorrhage (e.g., from blunt trauma or liver transplantation)
    - Rarely of clinical concern; resolves on its own and adrenal functions normally
  - Overall, AH is associated with 15% mortality rate
    - Waterhouse-Friderichsen syndrome (55-60%)
  - Adults: Adrenal crisis
  - Neonate: Death (from blood loss)

Treatment

- Medical
  - Correct fluid, electrolytes, and treat underlying cause
- Surgical: Adrenalectomy (open or laparoscopic)
  - Surgery not required, except in adrenal tumors

DIAGNOSTIC CHECKLIST

Consider

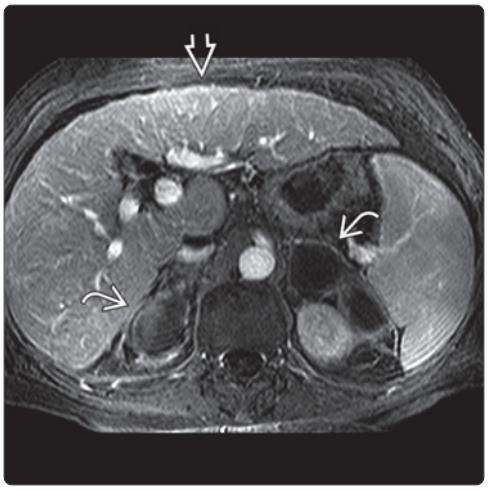
- Check for history of trauma, anticoagulant therapy, coagulopathies, malignancies, stress, adrenal tumor

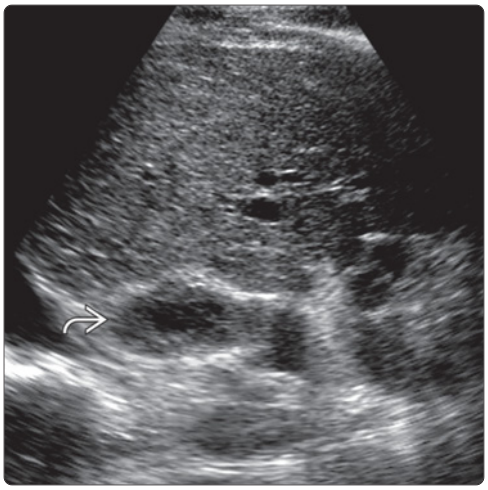
Image Interpretation Pearls

- NECT: Hyperdense lesion within adrenal gland
- MR: Varied signal intensity based on age of hematoma

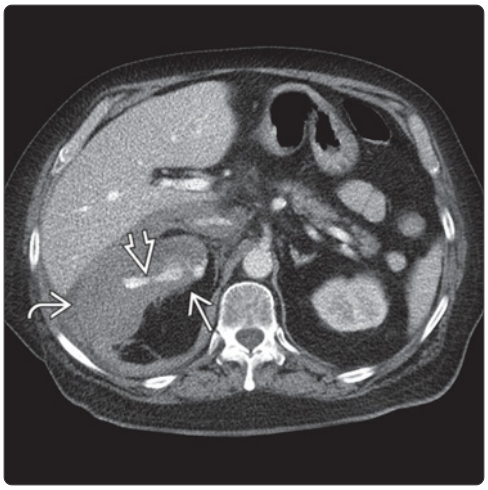
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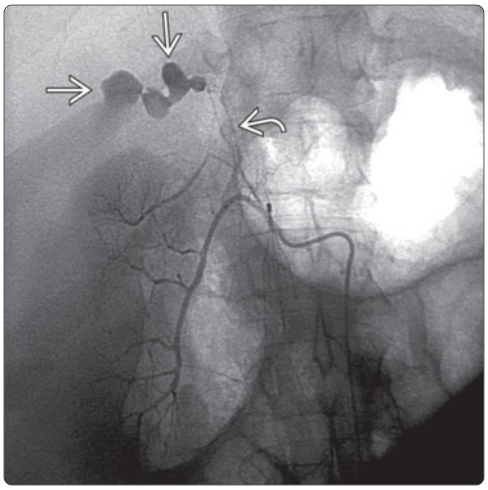
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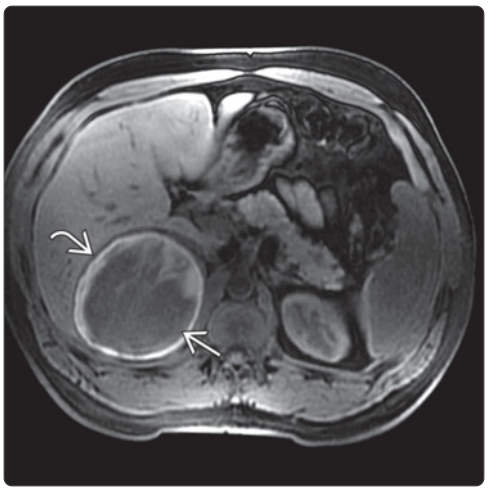


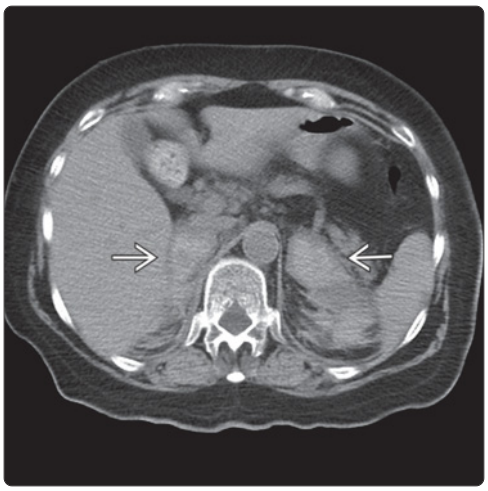
(Left) *T1WI gadolinium-enhanced MR of a female with catastrophic lupus anticoagulant, admitted with hypotension and adrenal insufficiency, shows nonenhancing bilateral adrenal hemorrhage. Note a cirrhotic-appearing liver.* **(Right)** *Ultrasound of the right upper quadrant in the same patient shows a hypoechoic right adrenal hematoma. The ultrasound appearance of adrenal hematomas (like most adrenal lesions) is nonspecific. Clinical history and additional imaging characterization are required.*





(Left) *CECT of an anticoagulated patient shows right adrenal hemorrhage, surrounding perinephric hemorrhage, and active extravasation. Although most adrenal hemorrhage can be managed conservatively, active extravasation and hypotension prompted angiography.* **(Right)** *Selective right renal angiogram shows extravasation from the inferior suprarenal artery. Embolization was successful. No underlying lesion was shown at follow-up CT.*





(Left) *T1WI with fat saturation image of a 40-year-old male with acute flank pain shows a large right adrenal hematoma. Peripheral high signal is due to methemoglobin; central hypointensity suggests fairly acute hemorrhage and deoxyhemoglobin. Although an endocrine work-up was negative, the mass was resected given its size. No underlying malignancy was identified.* **(Right)** *NECT of an elderly female with adrenal insufficiency and overwhelming pneumococcal sepsis shows bilateral adrenal hemorrhage.*

Adrenal Cyst

KEY FACTS

IMAGING

- "Adrenal cyst" is a descriptive term, not a pathological diagnosis
- True adrenal cysts
  - Majority are endothelial cysts (lymphangiomas)
  - Epithelial cysts exceedingly rare
  - Simple, or minimally complex adrenal cyst, thin rim calcification, no enhancement
- Pseudocysts
  - Prior hemorrhage inferred
  - Nonenhancing, but complex contents and wall calcification
  - Relevant history (extraadrenal malignancy, rapid growth), biochemical evaluation (cortisol, metanephrines): Consider underlying adrenal neoplasm
- Parasitic (echinococcal) cyst

TOP DIFFERENTIAL DIAGNOSES

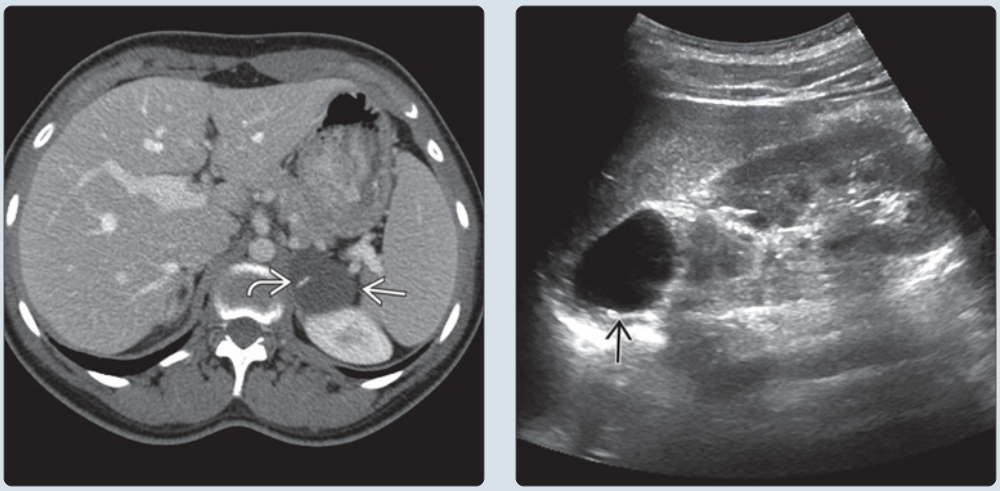
- Adrenal adenoma
  - CECT: Enhancing mass without visible wall or peripheral calcifications
- Gastric diverticulum
  - Air-, fluid-, or contrast-filled mass with no enhancement of contents
- Adrenal myelolipoma
  - Macroscopic fat
- Necrotic adrenal tumor
- Retroperitoneal bronchogenic cyst

CLINICAL ISSUES

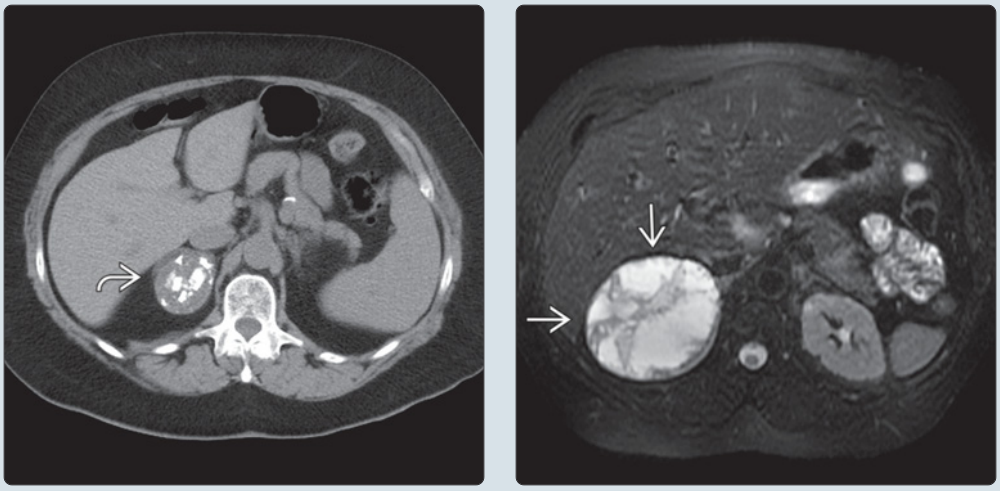
- No treatment required usually
- Imaging surveillance performed, although intensity and length of follow-up not defined

DIAGNOSTIC CHECKLIST

- Complicated cyst has high attenuation, thick enhancing wall, &/or septations



(Left) *Axial CECT in a 28-year-old woman with abdominal pain shows an incidental left adrenal cystic lesion. Note thin cyst septation, a finding characteristic of an endothelial adrenal cyst.* **(Right)** *Ultrasound in the same patient confirms an anechoic left suprarenal-adrenal cyst. The simple appearance of the cyst and the lack of additional relevant clinical history (malignancy, HTN, etc.) prompted surveillance rather than resection for this incidental, benign endothelial cyst.*



(Left) *Axial NECT in a 71-year-old woman shows a 4 cm complex cystic right adrenal mass containing coarse calcifications. Adrenalectomy (performed given lesion complexity and size) confirmed a hemorrhagic pseudocyst.* **(Right)** *Axial T2WI fat-suppressed MR in a 54-year-old woman with left flank pain shows a 7 cm complex, cystic right adrenal lesion with a ↓ signal hemosiderin ring. A pseudocyst was resected. Signal intensity of pseudocysts varies depending on the age of hemorrhage.*

TERMINOLOGY

Definitions

- "Adrenal cyst" is a descriptive term: Can mean true cyst, pseudocyst, or cystic mass

IMAGING

General Features

- Best diagnostic clue
  - Well-defined, nonenhancing, water density adrenal mass ± calcifications
- Location
  - Suprarenal
  - Unilateral > bilateral (8-10% of cases)
- Size
  - < 5 cm (50%), up to 20 cm

CT Findings

- NECT
  - Unilocular or multilocular mass
  - Well-defined, round to oval, homogeneous mass usually with water (0 HU) or near water density
    - Higher or mixed-attenuation mass (hemorrhage, intracystic debris, crystals)
  - Wall usually very thin
    - ↑ wall thickness, up to 3 mm for complex cysts
  - Calcifications
    - Rim-like or nodular (51-69%)
    - Centrally in intracystic septation (19%)
    - Punctate within intracystic hemorrhage (5%)
- CECT
  - No central enhancement ± wall enhancement
  - Coronal reformats helpful to determine organ of origin if large cyst

MR Findings

- T1WI
  - Homogeneous, hypointense mass
  - Hyperintense mass (hemorrhage)
- T2WI
  - Hyperintense mass

Ultrasonographic Findings

- Simple or septated suprarenal cyst
- Shadowing from calcification
- Real-time examination helpful to differentiate adrenal cyst from adjacent (renal, pancreatic) cyst

Imaging Recommendations

- Best imaging tool
  - Enhanced CT or MR (ultrasound for confirmation)

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- NECT: Lipid-rich adenoma (< 10 HU) mimics adrenal cyst
  - Peripheral or septal calcification favors adrenal cyst
- CECT: **Enhancing mass** without visible wall or peripheral calcifications
  - Assess wash-out kinetics to diagnose lipid-poor adenoma
- MR: Signal suppression at out of phase, chemical shift imaging
- Ultrasound: Solid adrenal lesion

Gastric Diverticulum

- May simulate left adrenal cyst
- Air-, fluid-, or contrast-filled suprarenal mass
- No enhancement
- Normal adjacent adrenal gland

Adrenal Myelolipoma

- Fat (not fluid) attenuation mass

Necrotic Adrenal Tumor

- Primary (pheochromocytoma or carcinoma) or metastatic
  - Clinical history, biochemical evaluation, lesion complexity suggests correct diagnosis
  - Enhancing soft tissue components

Retroperitoneal Bronchogenic Cyst

- Rare benign suprarenal fluid or soft tissue attenuation lesion
- Adjacent to, but separate from adrenal gland

Renal Cyst

- Coronal MR/CT or ultrasound useful to determine organ of origin of large retroperitoneal cystic lesions

PATHOLOGY

General Features

- Etiology
  - Congenital (endothelial, epithelial) cysts
  - Acquired (post hemorrhagic, inflammatory) pseudocysts
  - Cystic, hemorrhagic degeneration of underlying adrenal neoplasm

Staging, Grading, & Classification

- Accepted classification scheme
  - Pseudocyst
    - Most common type of cystic adrenal lesion in surgical series
    - No epithelial or endothelial lining: Fibrous cyst wall
    - Potentially as complication of prior trauma or hemorrhage though history of such often not elicited
    - May be associated with underlying adrenal neoplasm (pheochromocytoma, adrenal carcinoma, myelolipoma)
    - Attenuation and complexity at imaging varies depending upon hemorrhagic component
    - Wall and septal calcification common
  - Endothelial cyst
    - Subtypes: Lymphangiomatous and hemangiomatous
    - True cyst: Endothelial lining
    - Originate from preexisting vascular malformation or obstructed, ectatic lymphatic channels
    - Thin rim calcification typical
  - Epithelial cyst
    - Extremely rare: No acinar structures within normal adrenal gland
    - Mesothelial origin suggested (mesothelial cells potentially incorporated within adrenal gland during embryogenesis)

- Parasitic (hydatid) cyst

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Typically asymptomatic
  - Larger cysts may be symptomatic
    - Abdominal pain
    - Hemorrhage
  - Clinical history (malignancy, hypertension) elicited
    - May indicate cystic degeneration of underlying adrenal neoplasm (e.g., metastasis, pheochromocytoma)
- Diagnosis
  - Usually incidental finding at imaging
  - Endocrine-biochemical evaluation performed to exclude underlying functional adrenal tumor

Demographics

- Age
  - Any age, though 20-50 years old most common
- Gender
  - M:F = 1:3
- Epidemiology
  - Uncommon entity: Autopsy incidence 0.064-0.18%
  - Accounts for 1% of incidental adrenal lesions in large imaging series

Natural History & Prognosis

- Complications
  - Hypertension, infection, rupture, hemorrhage
- Excellent prognosis for vast majority of incidental, benign adrenal cysts
- Prognosis for pseudocysts secondary to adrenal neoplasm depends upon tumor histology

Treatment

- No treatment required usually
  - Imaging follow-up typically performed
    - Intensity and length of surveillance not defined
  - Cysts may enlarge over time
  - Endocrine evaluation (cortisol, metanephrine, etc.) performed
- Surgical resection if symptomatic, underlying adrenal neoplasm
  - Laparoscopic resection preferred

DIAGNOSTIC CHECKLIST

Consider

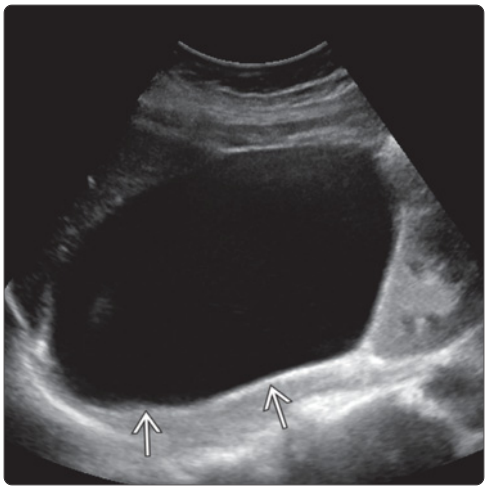
- Complicated cyst may suggest underlying adrenal neoplasm
- Clinical history, biochemical evaluation, prior imaging helpful

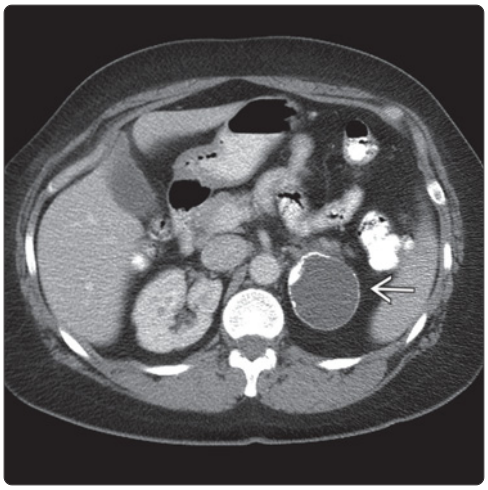
Image Interpretation Pearls

- Simple adrenal cyst: Scant septation, no enhancement, thin rim calcification
  - Likely benign endothelial cyst or pseudocyst
- Coronal imaging helpful to determine organ of origin (and exclude exophytic renal or pancreatic cyst)
- Complicated cyst: High attenuation, thick enhancing wall, &/or septations
  - Complexity may suggest underlying adrenal neoplasm and secondary pseudocyst

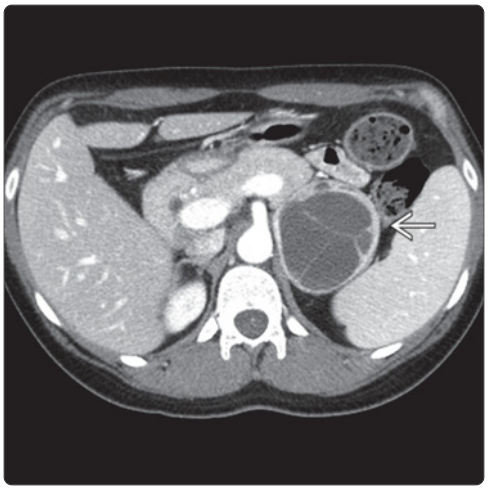
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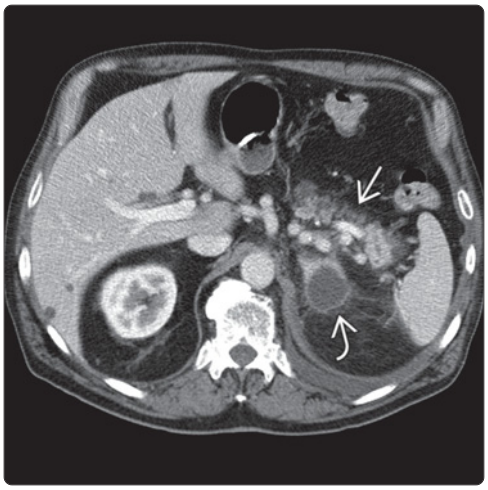
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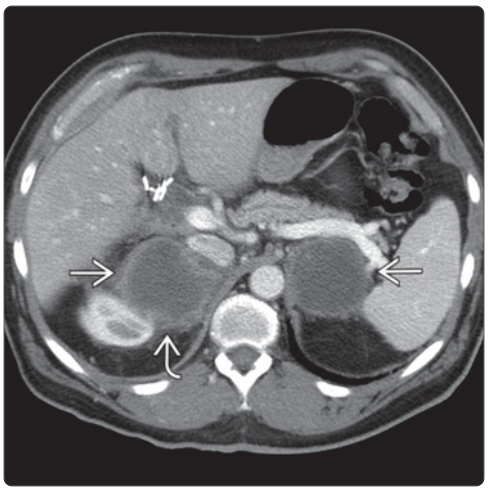


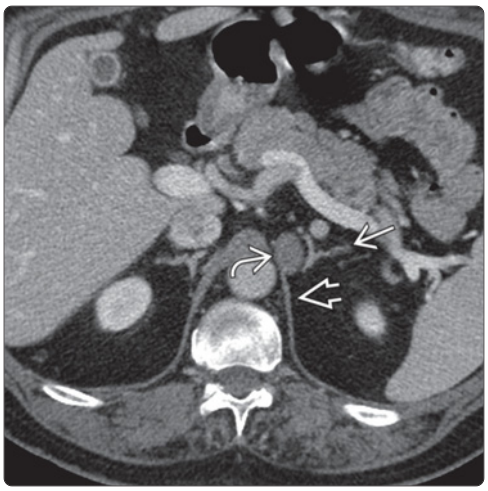
(Left) *Ultrasound in a 35-year-old woman shows a > 10 cm right suprarenal cyst. Real-time examination and a follow-up MR (not shown) confirmed a simple extrarenal-adrenal cyst that was subsequently resected. An epithelial cyst (a rare subtype of adrenal cysts) was shown at histology.* **(Right)** *Axial CECT of a 51-year-old woman shows an incidental peripherally calcified left adrenal cyst. Coarse calcification suggests a pseudocyst. Pseudocysts may be due to prior trauma/hemorrhage but this history is often absent.*





(Left) *Axial CECT in a hypertensive 36-year-old woman shows a peripherally enhancing, septated 7 cm left adrenal pheochromocytoma. Enhancing soft tissue should prompt testing for an underlying adrenal neoplasm.* **(Right)** *Axial CECT of a 72-year-old man with pancreatitis shows peripancreatic infiltration and a 3 cm adrenal pseudocyst. The pseudocyst resolved on a CT follow-up. Imaging surveillance is advocated for asymptomatic, probable pseudocysts.*





(Left) *Staging CT examination in a 66-year-old man with metastatic lung carcinoma shows large bilateral necrotic adrenal metastases. The clinical history and an enhancing rind prevent an erroneous diagnosis of benign adrenal cysts.* **(Right)** *Axial CECT of a 62-year-old man shows a 2 cm incidental left suprarenal lesion. Location adjacent to the left crus, and separate from the left adrenal indicate a retroperitoneal bronchogenic cyst, a mimic of adrenal cyst.*

Adrenal Adenoma

KEY FACTS

IMAGING

- Well-circumscribed, uniform, low-attenuation, small adrenal mass
  - Low attenuation due to abundant intracytoplasmic lipid
  - Imaging features of typical lipid-rich adenomas
    - NECT: < 10 HU (71% sensitivity, 98% specificity)
    - MR: Significant decrease in signal intensity at out of phase T1W imaging (due to intravoxel lipid and water)
  - Lipid-poor adenomas (10-40% cases): Utilize relative or absolute CT contrast washout kinetics for diagnosis
- Accounts for vast majority of adrenal "incidentalomas"
  - Imaging intensive algorithm suggested for incidental adrenal lesions, though overwhelming majority are benign and hormonally inactive
  - Primary hyperaldosteronism (Conn syndrome): 80% due to unilateral, typically small (< 2 cm) adenoma
  - Cushing syndrome: 80-85% due to adrenal hyperplasia

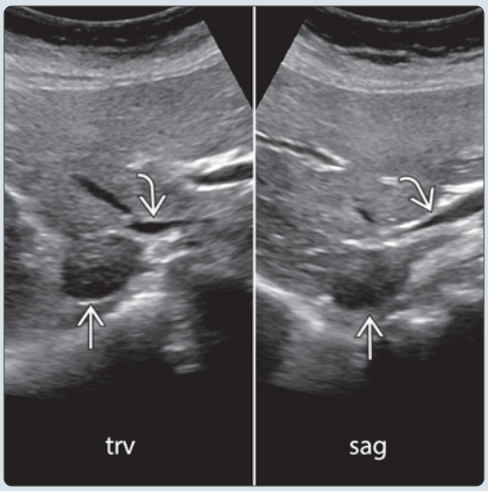
TOP DIFFERENTIAL DIAGNOSES

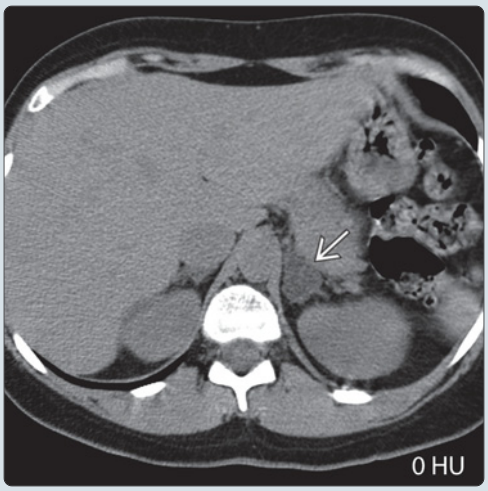
- Adrenal metastases and lymphoma
- Adrenal myelolipoma
- Adrenal (macronodular) hyperplasia
- Pheochromocytoma
- Adrenal carcinoma
- Gastric diverticulum
- Adrenal cyst

DIAGNOSTIC CHECKLIST

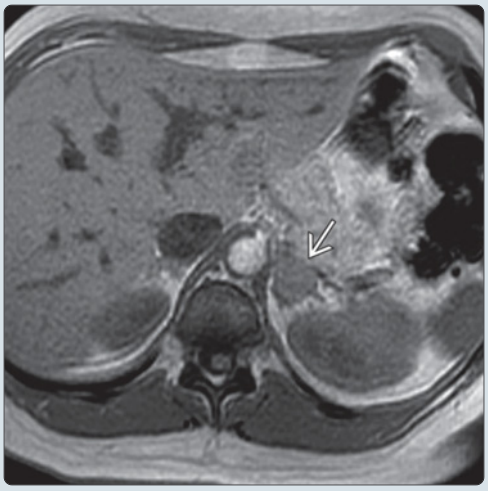
- Asymptomatic mass: Usually nonfunctioning adenoma, even in patients with known cancer
- NECT and MR are equally accurate for diagnosis of lipid-rich adenoma
- Utilize dedicated CECT adrenal protocol for diagnosis of potential lipid-poor adenomas

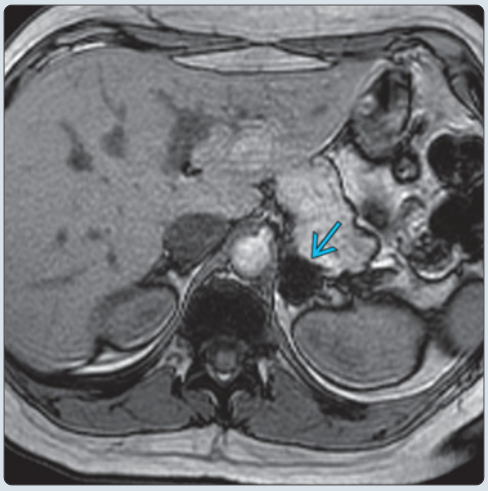
(Left) *Transverse and sagittal ultrasound of a 70-year-old woman with elevated liver function tests show an incidental solid right adrenal lesion ➡ posterior to the IVC ➡. Although statistically an adenoma, the ultrasound appearance is nonspecific. NECT confirmed a lipid-rich adenoma.* **(Right)** *NECT of a 45-year-old man shows a uniform 2 cm low-attenuation (0 HU) left adrenal lesion ➡. NECT is typically the initial imaging modality of choice for adrenal mass characterization, although a follow-up MR was performed.*





(Left) *T1 in-phase MR performed on the same patient shows a well-circumscribed, intermediate signal intensity left adrenal lesion ➡.* **(Right)** *The corresponding out of phase T1WI MR shows uniform signal suppression of the left adrenal gland ➡. Low attenuation (< 10 HU) at NECT and signal dropout at chemical shift MR are characteristic features of lipid-rich adenomas.*





TERMINOLOGY

Definitions

- Benign adrenal cortical tumor

IMAGING

General Features

- Best diagnostic clue
  - Imaging strategies target typical adenoma histology: Abundant intracytoplasmic lipid
    - Low attenuation (< 10 HU) at NECT
    - Significant loss of signal at out of phase T1W imaging (intravoxel fat and water)
- Size
  - Cushing syndrome adenoma: Varies from 2-5 cm
  - Conn syndrome adenoma: Classically < 2 cm (20% < 1 cm)
  - Vast majority of incidental, hormonally inactive adrenal adenomas are small (< 2 cm)
- Morphology
  - Usually round to oval suprarenal mass
- Key concepts
  - Most common adrenal cortex tumor (10% bilateral)
    - Accounts for > 90% of all "incidentalomas"
  - May occur in up to 9% of general population
  - Lipid-rich adrenal adenoma: 60-90% of all adenomas
  - Lipid-poor adrenal adenoma: 10-40% of adenomas
  - Increased incidence in patients with diabetes and hypertension
  - NECT (or MR): Study of choice to diagnose incidental adrenal masses
  - Classified into 2 types based on function
    - Nonhyperfunctioning: Normal hormone levels
    - Hyperfunctioning: Primary hyperaldosteronism, Cushing syndrome, hyperandrogenism
  - **Cushing syndrome**
    - 15-25% of cases are due to autonomous adrenal adenoma
    - 80-85% of cases are due to **adrenal hyperplasia**
    - Adenomas usually > 2 cm
  - **Primary hyperaldosteronism (Conn syndrome)**
    - 80% of cases are due to adrenal adenoma
    - 20% of cases are due to adrenal hyperplasia
    - Adenomas are often small (< 2 cm)

CT Findings

- NECT
  - Smooth, well defined, round or oval in shape
  - Homogeneous soft tissue mass of 0-20 HU
  - **Lipid-rich adrenal adenoma** (60-90% of cases)
    - Uniform low attenuation
      - Meta-analysis of < 10 HU threshold: Sensitivity 71%, specificity 98%
      - Sensitivity may increase to almost 90% with histogram analysis (identify negative pixels), though variable results and scanner dependent
  - **Lipid-poor adrenal adenoma** (10-40% of cases)
    - Attenuation varies from 10-30 HU
    - Difficult to differentiate from metastases on NECT
  - Cushing syndrome due to adrenal adenoma
    - Remainder of ipsilateral gland and contralateral adrenal gland may be atrophic due to ↓ ACTH levels
    - ↑ cortisol: Feedback inhibition on pituitary ACTH
  - ACTH independent macronodular hyperplasia: Multiple, bilateral, functioning adrenal adenomas
  - Conn syndrome due to adrenal adenoma
    - Remainder of ipsilateral gland and contralateral adrenal gland appear normal
  - Large adenomas
    - More heterogeneous than small adenomas
    - ± hemorrhage, cystic degeneration, calcification
      - Growth should raise suspicion for malignancy
- CECT
  - Enhancing adrenal mass that "de-enhances" rapidly
    - Dedicated adrenal CT exam incorporates initial dynamic enhanced phase (~ 70-second scan delay) and 15-minute delay
    - Relative % washout = dynamic enhanced (HU) - delayed (HU) / dynamic enhanced HU
      - Relative % washout > 40%: Sensitivity of 96% and specificity of 100%
    - Absolute % washout (if NECT available) = dynamic enhanced (HU) - delayed (HU) / dynamic enhanced (HU) - unenhanced (HU)
      - Absolute % washout > 60%: Sensitivity of 86-88% and specificity of 92-96%
    - 10-minute delay utilized by some centers, but shorter delay may decrease sensitivity
    - Adrenal washout calculators readily available on multiple websites
    - Utilize technique for indeterminate, potentially lipid-poor adenomas
    - Clinical context critical: Rapid washout of pheochromocytomas, adrenal and renal cell carcinomas reported
  - Dual-energy CT and iodine subtraction techniques can generate virtual unenhanced images
    - Approach may identify lipid-rich adenomas and obviate need for additional imaging

MR Findings

- T1WI and T2WI
  - Low to intermediate signal intensity
- Chemical shift (in and out of phase) imaging
  - Mainstay of MR diagnosis
  - Sensitivity and specificity equivalent to NECT
  - Signal loss at out of phase T1W imaging due to intravoxel water and fat protons
  - Inverse relationship between % of lipid-rich cells and relative ↓ signal intensity at out of phase imaging
    - May not identify lipid-poor adenomas
  - Visual inspection of signal intensity in phase (SIP) and out of phase (SOP), though quantitative analysis may be helpful
    - Adrenal to spleen CSI (chemical shift imaging) ratio: Lesion:spleen SIP/lesion:spleen SIOP
      - < .71 = adenoma
    - Adrenal signal intensity index: 100 x (SIP - SIOP) / SIP
      - > 1.5% = adenoma
  - Beware technical pitfalls
    - Sampling of 1st echo pair at 3T is challenging

  - India ink artifact mimics signal loss, particularly in small adrenal lesions
  - Other primary or secondary adrenal lesions may contain lipid
  - Adenomas and metastases may coexist in same gland (collision tumor)
- T1 C+
  - Rapid, uniform enhancement and de-enhancement
- Ancillary MR techniques
  - Diffusion MR: Disappointing results (ADC overlap between adenomas and metastases)
  - MR spectroscopy: Choline:creatinine and choline:lipid ratio discriminatory threshold ratios may aid in adrenal lesion characterization, though larger studies needed

Ultrasonographic Findings

- Grayscale ultrasound
  - Nonspecific solid suprarenal mass
    - Right suprarenal mass seen more clearly than left side due to acoustic window provided by liver

Angiographic Findings

- Conventional
  - Adrenal arteriography
    - Catheterization of renal or inferior adrenal arteries shows vascular supply of adrenal tumors
    - Adenomas are usually hypovascular to moderately vascular
    - No arterial encasement, or venous laking or puddling, which are malignant vascular features
  - Adrenal venography
    - Most commonly to obtain adrenal vein samples
      - Advocated for patients with primary hyperaldosteronism triaged to adrenalectomy
      - Technically difficult study, but may confirm laterality of small aldosterone-secreting adenoma
      - Technical approach and criteria for positive study varies; ACTH stimulation may increase accuracy
    - Adrenal adenoma is seen as filling defect within adrenal gland displacing adjacent vessels
    - Circumferential vein frequently seen around adrenal adenoma

Nuclear Medicine Findings

- PET/CT
  - Utilized as part of malignancy staging
  - Markedly increased FDG-18 uptake characteristic of metastases
  - Adenomas may also accumulate FDG-18
  - SUV thresholds published, but adenomas typically less intense than liver
    - Potential false-negatives: Metastases from primary carcinomas that are non-FDG avid (e.g,. neuroendocrine tumors)

Imaging Recommendations

- NECT is initial study of choice to confirm diagnosis of lipid-rich adrenal adenoma
  - ROI should encompass lesion: Attenuation < 10 HU is diagnostic
- MR with in and out of phase imaging equivalent to NECT for lipid-rich lesions
  - Signal "dropout" on T1WI out of phase: Qualitative assessment typically suffices
- CECT including 15-minute delayed phase used for potential lipid-poor adenomas: Calculate either relative or absolute washout

DIFFERENTIAL DIAGNOSIS

Adrenal Metastases and Lymphoma

- Adrenal metastases
  - Unilateral or bilateral masses ± central necrosis, hemorrhage
  - Usually known to have malignancy elsewhere
  - NECT: Metastases mimic lipid-poor adenoma
  - CECT: Hypo- or hypervascular and prolonged washout pattern
- Adrenal lymphoma
  - Usually spread to adrenal gland from retroperitoneal tumor
    - Unilateral or bilateral masses
  - Unilateral primary lymphoma (non-Hodgkin) can mimic adenoma
  - Hypovascular; moderate enhancement with contrast

Adrenal Myelolipoma

- Small or large, asymptomatic adrenal mass
- Intramural macroscopic fatty elements on imaging

Adrenal Hyperplasia

- Adrenal glands are often symmetrically enlarged
- Width of adrenal gland limbs > 10 mm (diagnostic)
- No discrete mass or nodule seen as rule
- Dominant macronodule of macronodular hyperplasia mimics small adrenal adenomas
  - Cortisol-secreting adenoma: Remainder of ipsilateral and contralateral glands, atrophic (↓ ACTH)
  - Macronodular hyperplasia: Both glands are enlarged (due to elevated ACTH levels)
- No obvious enhancement and washout pattern seen

Pheochromocytoma

- Tumor > 3 cm in most cases; T2WI classically hyperintense
- Highly vascular tumor prone to hemorrhage, necrosis
- Bilateral adrenal tumors in multiple endocrine neoplasia syndromes

Unilateral Adrenal Hemorrhage

- Chronic hematoma: Well-defined, round, low-density, mass-like lesion simulating adenoma

Adrenal Carcinoma

- Rare, unilateral invasive and enhancing mass
- > 6 cm when initially diagnosed

Gastric Diverticulum

- Abnormal rounded soft tissue lesion in left suprarenal area; mimics adrenal mass
- Diverticular contents do not enhance; adenomas do
- Distend stomach with gas and fluid; scan in prone position to distend diverticulum

Ganglioneuroma

- Younger patients; mean age: 27 years

- Larger mass; average tumor size: 8 cm

Adrenal Cyst

- Attenuation similar to lipid-rich adenoma
- Lack of enhancement, rim calcification may suggest diagnosis

PATHOLOGY

General Features

- Etiology
  - Exact etiology unknown
- Associated abnormalities
  - Multiple endocrine neoplasia (MEN) syndromes
  - Most adrenals with adenoma: Normal function
  - Occasionally adenoma causes adrenal hyperfunction
  - Normal adrenocortical secretory hormones
    - Adrenal cortex: Cortisol, aldosterone, androgens

Gross Pathologic & Surgical Features

- Well-delineated, tan-yellow, ovoid mass

Microscopic Features

- 70% of adenomas: High % of intracytoplasmic lipid
- 30% of adenomas: Lower % of intracytoplasmic lipid

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic incidental CT imaging finding
  - Conn syndrome: Hypertension and weakness
  - Cushing syndrome: Moon facies, truncal obesity, purple striae, and buffalo hump
  - Virilization in women
  - Lab data: ↑ aldosterone, cortisol, &/or androgens
  - Diagnosis: Clinical, biochemical, imaging, histology

Demographics

- Age
  - Prevalence of adenoma increases with age
    - Peak at 60-69 years, decreasing thereafter
- Epidemiology
  - Most common adrenal tumor of all incidentalomas
  - ↑ incidence in patients with diabetes or HTN
  - Occurs in up to 9% of population (postmortem data)

Natural History & Prognosis

- Prognosis: Excellent when incidental and nonhyperfunctioning

Treatment

- No treatment when asymptomatic incidental finding
- Laparoscopic removal of gland if hyperfunctioning

DIAGNOSTIC CHECKLIST

Consider

- Asymptomatic mass: Usually nonhyperfunctioning adenoma, even in patient with known cancer

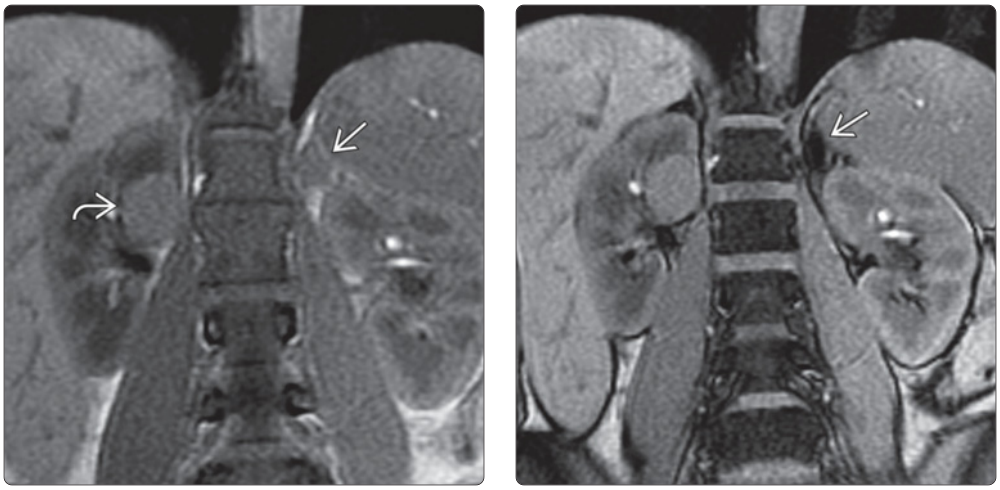
Image Interpretation Pearls

- Well-defined, low-density (< 10 HU) suprarenal mass
- Enhances + washout pattern of > 50% within 15 minutes
- T1WI out of phase: Signal "dropout" lipid-rich mass

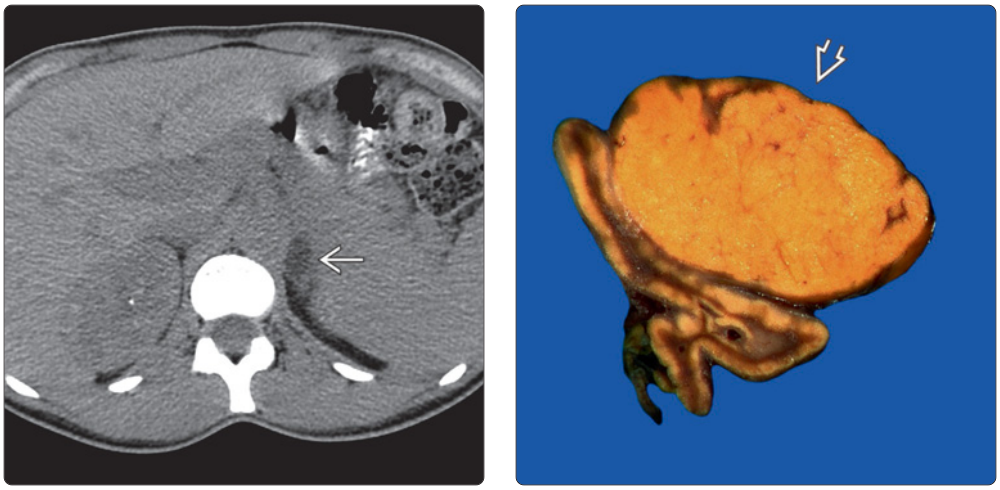
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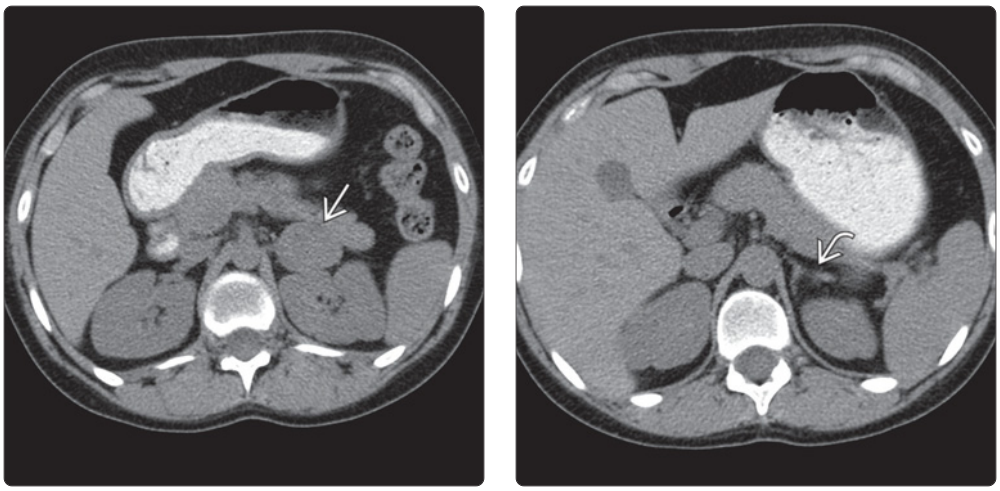
(Left) *Coronal phase T1WI MR of a 34-year-old woman with hypertension and hypokalemia shows an intermediate signal intensity left adrenal lesion. Note incidental high T1 signal right renal cyst.* **(Right)** *Marked signal suppression of the left adrenal lesion on the corresponding out of phase MR confirms a clinically functional, imaging apparent, lipid-rich adenoma. Signal suppression is due to intravoxel cytoplasmic lipid and water. Aldosterone-secreting (Conn) adenomas are typically small (< 2 cm).*

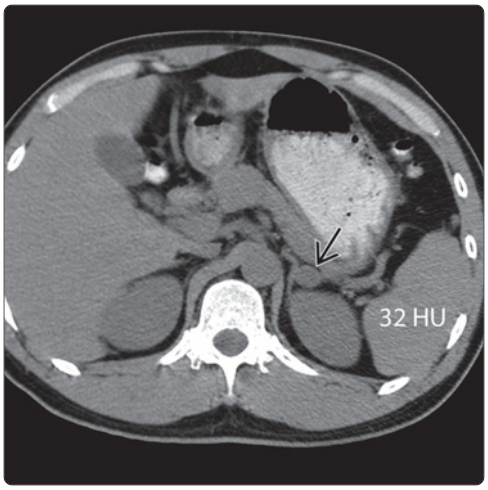


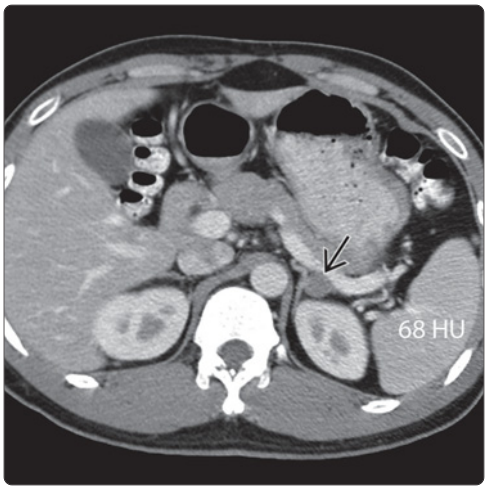
(Left) *Axial NECT of the same patient shows a 1.5 cm low-attenuation (5 HU), lipid-rich, left adrenal adenoma.* **(Right)** *Adrenal vein sampling was subsequently performed, which confirmed an aldosterone-secreting left adrenal lesion. Photograph of the laparoscopically resected left adrenal gland shows an exophytic, well-circumscribed, adrenocortical adenoma.*



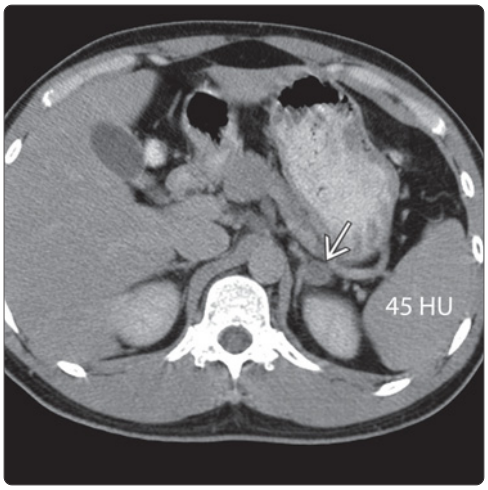
(Left) *NECT of a 20-year-old woman with Cushing syndrome shows a 4 cm solid (42 HU) left adrenal mass. Diffuse adrenal hyperplasia is typically shown in patients with Cushing symptomatology, though cortisol excess may be due to an adenoma (as in this case), ACTH-independent macronodular hyperplasia, or adrenal carcinoma.* **(Right)** *More superior axial image obtained in the same patient shows a thin ipsilateral left adrenal gland due to autonomous cortisol secretion and a feedback ↓ in ACTH.*

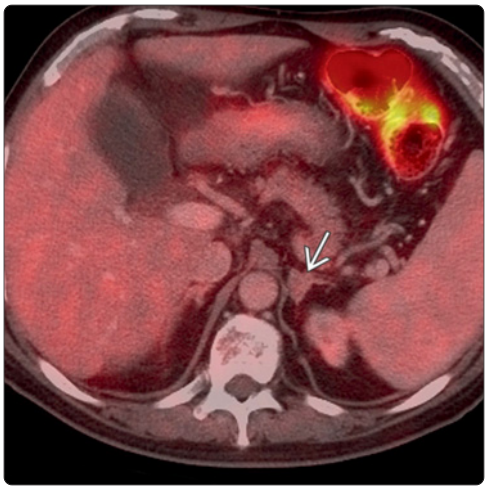




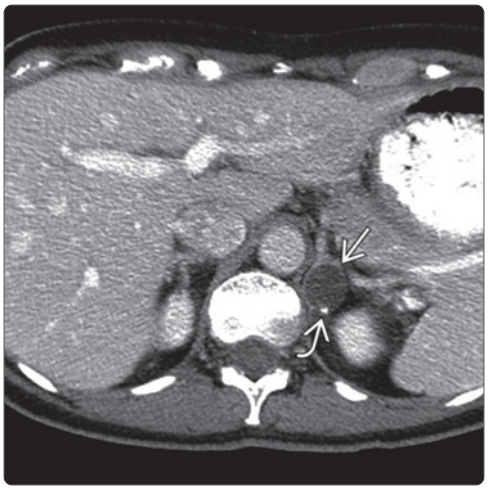


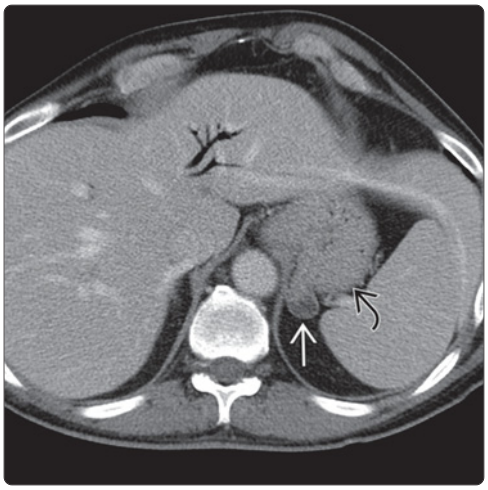
(Left) *A dedicated adrenal wash-out CT examination was performed in this 28-year-old man with an incidental, 1.5 cm, left adrenal nodule. The attenuation of the nodule pre contrast administration was 32 HU. (Right) The attenuation of the left adrenal nodule increased to 68 HU during the portal venous phase of enhancement.*





(Left) *Axial image of the same patient obtained 15 minutes post contrast administration shows nodule de-enhancement to 45 HU. The absolute washout of > 60% is diagnostic of a lipid-poor adenoma. Adrenal CECT is utilized for indeterminate lesions at NECT or chemical shift MR.* **(Right)** *PET/CT examination of a 54-year-old man with a history of lymphoma shows a 1.8 cm left adrenal lesion with FDG uptake similar or less than the liver, a characteristic appearance of an incidental adrenal adenoma.*





(Left) *CECT shows a 3 cm low-attenuation (2 HU) left adrenal mass. No enhancement was shown relative to a baseline NECT. Note rim calcification. Both are features of an adrenal (pseudo) cyst, a mimic of a lipid-rich adenoma.* **(Right)** *CECT shows a low (6 HU) left suprarenal lesion. Its location adjacent to the gastric fundus indicates a gastric diverticulum. Identification of gas or contrast within the "mass" and coronal images may prevent erroneous diagnosis of lipid-rich adenoma.*

Adrenal Myelolipoma

KEY FACTS

TERMINOLOGY

- Uncommon benign tumor composed of mature fat tissue and hematopoietic elements (myeloid and erythroid cells)

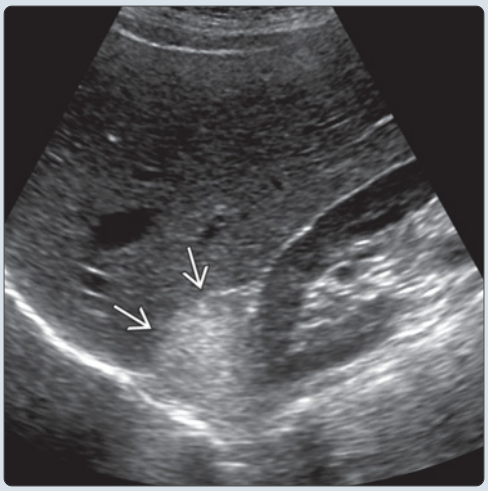
IMAGING

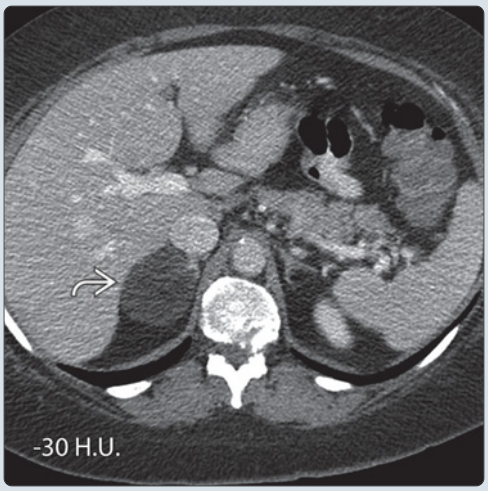
- Uncommon benign, nonfunctioning adrenal tumor
- Accounts for 7-15% of incidental adrenal masses, usually in older population
- Typically unilateral and very rarely bilateral
- Large tumors can mimic retroperitoneal lipomas, liposarcomas
- Asymptomatic, though larger tumors may hemorrhage
- CT
  - Lesion containing fat attenuation (-30 to -90 HU): Fat within tumor is diagnostic
  - Usually well-defined mass with recognizable pseudocapsule (remaining adrenal)
  - Punctate calcifications seen (24% of cases)
  - Coronal reconstruction helpful to differentiate from exophytic renal angiomyolipoma
- MR
  - Tumor with major fat component
  - T1WI in phase: Typically hyperintense
  - Fat-suppression sequences: Loss of signal

TOP DIFFERENTIAL DIAGNOSES

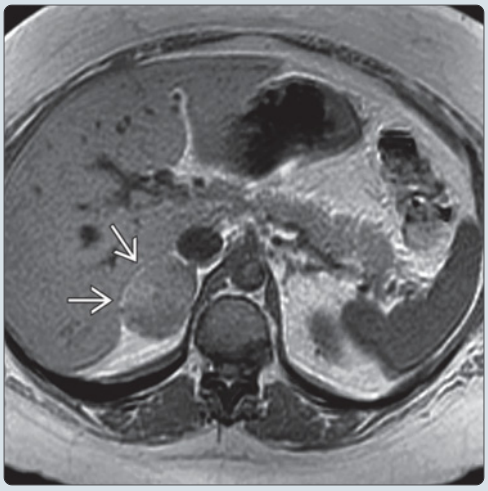
- Adrenal adenoma
  - Intracellular lipid vs. macroscopic fat
- Adrenal metastases and lymphoma
- Retroperitoneal liposarcoma
- Pheochromocytoma
- Adrenal carcinoma
- Renal angiomyelolipoma
  - Coronal CT reconstruction or MR useful to determine organ of origin

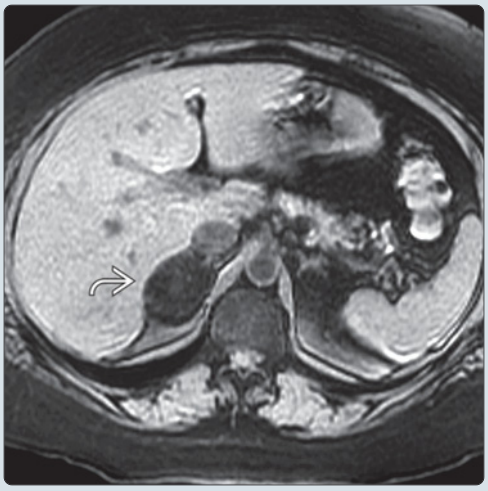
(Left) *Ultrasound of a 52-year-old woman with right upper quadrant pain shows a subtle echogenic right suprarenal mass. Even large myelolipomas may not be perceived at ultrasound given their isoechogenicity relative to retroperitoneal fat.* **(Right)** *Axial CECT of the same patient shows a fat-attenuation right adrenal lesion. Macroscopic fat is the hallmark of myelolipomas though the fat may be interspersed with soft tissue myeloid elements and hemorrhage.*





(Left) *Follow-up T1-weighted MR shows a relatively intense right adrenal lesion. No significant signal loss was demonstrated at out-phase imaging (not shown).* **(Right)** *Axial T1WI MR with fat saturation confirms that the right adrenal lesion is composed of fat, a finding pathognomonic of a myelolipoma. Signal suppression with fat saturation, and persistent signal at out-phase imaging indicate macroscopic fat, rather than the intracellular lipid characteristic of adrenal adenomas.*





TERMINOLOGY

Definitions

- Uncommon benign tumor composed of mature adipose tissue and hematopoietic elements

IMAGING

General Features

- Best diagnostic clue
  - Suprarenal mass containing fat
- Location
  - Suprarenal
    - Rare extraadrenal myelolipomas (presacral, retroperitoneal)
- Size
  - Usually 2-10 cm, rarely 10-20 cm
- Key concepts
  - Uncommon benign neoplasm of adrenal gland
  - Autopsy prevalence rate of 0.2-0.4%
  - Accounts for 7-15% of adrenal "incidentalomas"
  - Usually unilateral incidental finding in elderly patient
  - Larger tumors can bleed spontaneously
  - Nonfunctioning (does not secrete hormones)
  - Large myelolipomas can mimic retroperitoneal lipoma or liposarcoma

CT Findings

- CT appearance depends on histologic composition
  - Most tumors are heterogeneous adrenal masses composed of varying percentages of fat
    - Low-attenuation suprarenal lesion containing fat density (-30 to -90 HU)
      - Average nonenhanced CT attenuation value of tumor: -74 HU in one series
      - Interspersed soft-tissue attenuation components: Myeloid elements, hemorrhage
    - Presence of macroscopic fat within tumor is diagnostic
  - Punctate calcifications seen (24% of cases)
  - Usually well-defined mass with recognizable pseudocapsule (remnant adrenal)
  - Coronal reconstructions may help determine organ of origin: Adrenal myelolipoma vs. exophytic renal angiomyelolipoma

MR Findings

- MR appearance depends on histologic composition
  - Tumor with major fat component
    - T1WI in-phase: Typically hyperintense
    - T1WI out-phase: Persistent hyperintensity of macroscopic fat
    - T1WI fat saturation: Confirmatory suppression of signal
  - Bone marrow elements (myeloid and erythroid cells)
    - Low signal on T1WI, moderate signal on T2WI
  - Hemorrhage: Varying T1, T2 signal depending upon age of blood

Ultrasonographic Findings

- Grayscale ultrasound
  - Well-defined, echogenic mass (↑ fat tissue)
    - Often overlooked: Lack of mass effect and isoechogenicity relative to retroperitoneal fat
  - Heterogeneous mass (↑ myeloid cells)

Angiographic Findings

- Conventional
  - Differentiate myelolipoma from retroperitoneal liposarcoma by determining origin of blood supply and vascularity of tumors

Nuclear Medicine Findings

- Typically not metabolically active, though uptake reported at FDG PET

Imaging Recommendations

- Helical NECT or MR with fat-suppression sequence

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- Lipid-rich adenoma: ↓ attenuation (< 10 HU) at NECT
- No true fat density, unlike adrenal myelolipoma
- CECT: Washout 15 minutes post injection > 50%
  - Relative washout: > 40%
  - Absolute washout > 60%

Metastases and Lymphoma, Adrenal

- Bilateral lesions: Clinical history paramount
  - Metastases: Soft tissue attenuation (signal)
  - Lymphoma: May maintain adreniform shape, adjacent retroperitoneal adenopathy

Liposarcoma, Retroperitoneal

- Retroperitoneal primary sarcoma involving perirenal space may simulate adrenal (or renal) fatty tumor

Pheochromocytoma

- Highly vascular, prone to hemorrhage and necrosis
- Hyperintense on T2WI, bilateral in MEN syndromes
- Clinical history (labile hypertension) and urinary catecholamines

Adrenal Carcinoma

- Rare, unilateral, invasive, and enhancing mass
- Venous invasion, distant metastases
- May contain fat: Engulfed retroperitoneal fat vs. lipomatous metaplasia

Renal Angiomyelolipoma

- Exophytic upper pole angiomyolipoma may mimic
- Coronal reconstruction/MR helpful to determine organ of origin

PATHOLOGY

General Features

- Etiology
  - Unknown
  - Best hypothesis: Reticuloendothelial cell metaplasia of capillaries in adrenal (stress/infection/necrosis)
  - Secondary hypothesis: Myelolipoma represents site of extramedullary hematopoiesis
- Associated abnormalities

- Adrenal collision tumors (coexistent myelolipoma and adenoma typical)
- Large bilateral myelolipomas reported with longstanding, poorly treated congenital adrenal hyperplasia

Gross Pathologic & Surgical Features

- Cut section: Fat, soft tissue components

Microscopic Features

- Mature fat cells and megakaryocytes; no malignant cells
- Calcification
- Hemorrhage within larger lesions

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
    - Usually incidental finding on CT, MR
    - Typically biochemically nonfunctioning
  - Symptomatic
    - Acute abdomen: Flank pain due to rupture and hemorrhage
    - Case reports of hormonally active tumors: Cushing, Conn syndromes, virilization
- Diagnosis: Pathognomonic MR/CT features
  - Biopsy reserved for larger, atypical lesions, though prone to sampling error

Demographics

- Age
  - Usually elderly: 50-70 years old
- Epidemiology
  - Autopsy incidence 0.2-0.4%

Natural History & Prognosis

- Complication: Rupture with hemorrhage (rare)
- Prognosis: Excellent

Treatment

- When diagnosis is certain, surgery not needed for "smaller" (< 5-7 cm) lesions
- Surgery reserved for larger, symptomatic, or atypical lesions
  - Surgical series have confirmed utility of laparoscopic resection

DIAGNOSTIC CHECKLIST

Consider

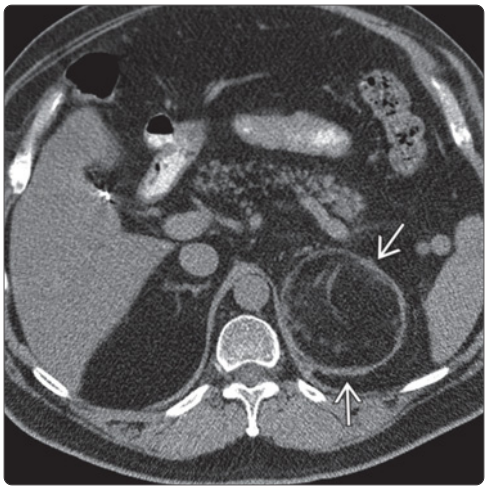
- Differentiate from other tumors (lipid-rich adenoma)
- Key is presence of imaging-apparent adipose tissue; avoid further work-up for incidental mass

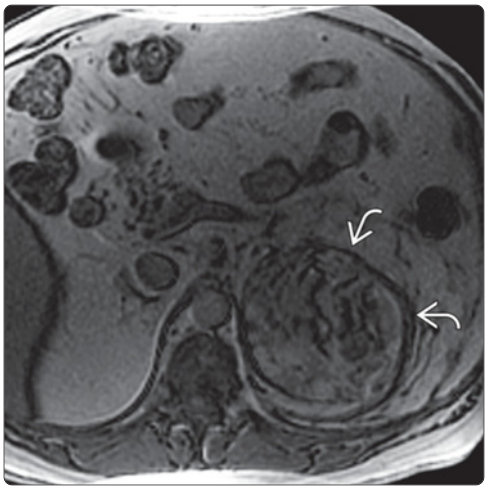
Image Interpretation Pearls

- Well-defined, heterogeneous, fat-attenuation tumor on CT
- T1WI hyperintense, signal loss with fat suppression

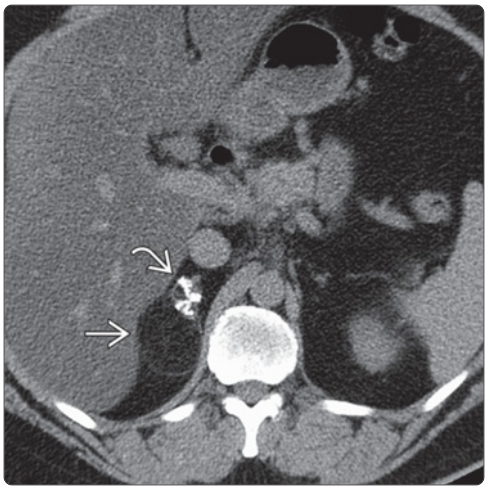
SELECTED REFERENCES

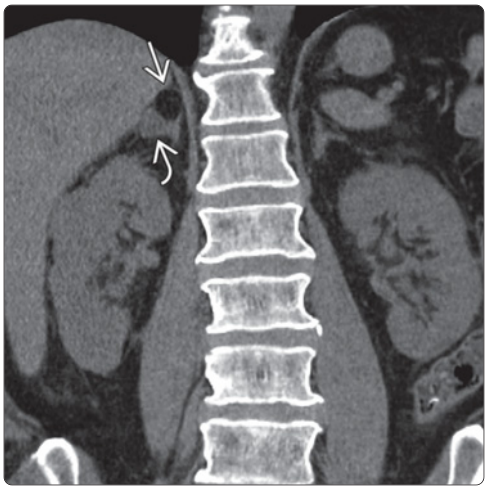
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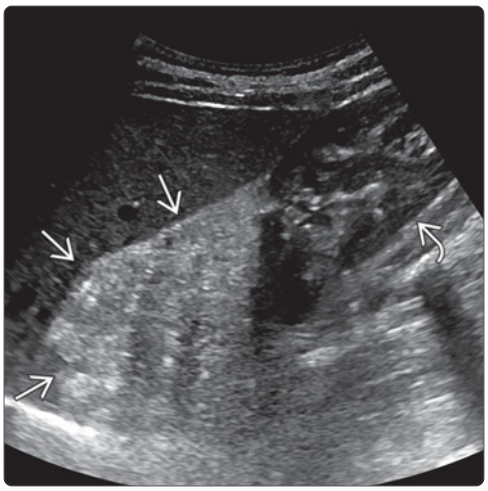


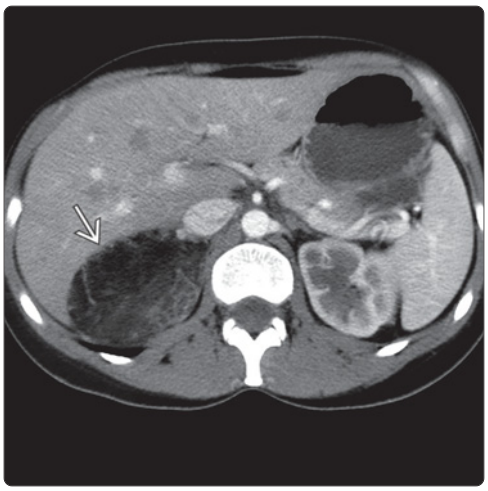
(Left) *Axial CECT performed on a 55-year-old man with left flank pain shows a large, fat attenuation mass, a soft tissue attenuation rim, and perinephric infiltration.* **(Right)** *Axial out-phase MR in the same patient shows a low signal rim (due to hemosiderin) and perinephric infiltration. Most myelolipomas are asymptomatic, but larger tumors may hemorrhage, as in this patient. An adrenalectomy was performed given the large size of the tumor and evidence of repeated hemorrhage.*





(Left) *Axial CECT of a 53-year-old man shows an incidental myelolipoma containing calcifications.* **(Right)** *NECT performed on a 71-year-old man with left flank pain shows an incidental collision tumor composed of a myelolipoma and an adenoma.*





(Left) *Ultrasound of a 44-year-old woman with abdominal pain shows a large, echogenic, suprarenal mass that displaces the kidney. A CT was recommended given the size of the lesion.* **(Right)** *Axial CECT in the same patient confirms a fat attenuation suprarenal mass. An extrarenal angiomyolipoma was ultimately resected. The differential diagnosis of large myelolipomas includes retroperitoneal liposarcomas, angiomyolipomas, and adrenal carcinomas with lipomatous metaplasia.*

Pheochromocytoma

KEY FACTS

TERMINOLOGY

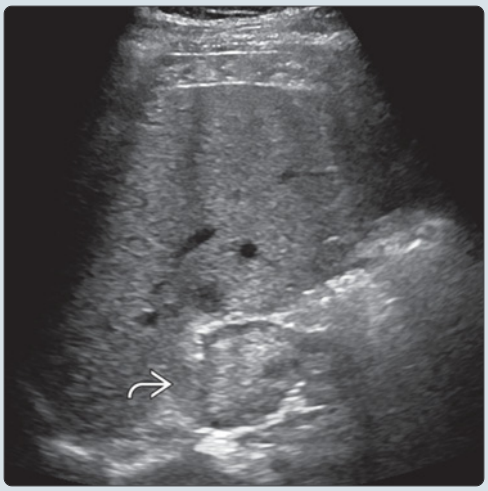
- Tumor arising from chromaffin cells of adrenal medulla or extraadrenal paraganglia

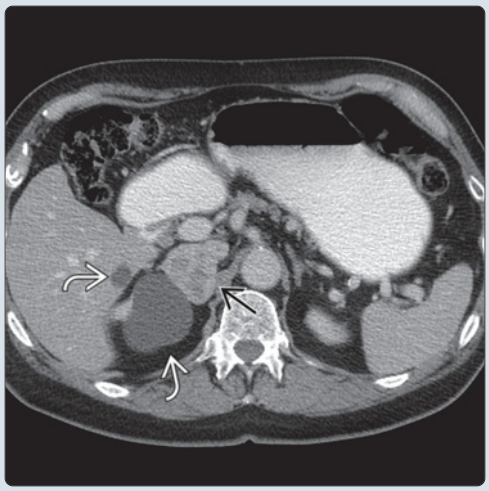
IMAGING

- Adrenal medulla (90%)
- Extraadrenal (10%)
  - Along sympathetic chain: Anywhere from neck to urinary bladder
  - Subdiaphragmatic (98%); thorax (1-2%)
  - Organ of Zuckerkandl and near urinary bladder are relatively common sites
- Hereditary pheochromocytomas
  - Small, bilateral adrenal lesions in younger patient
- Sporadic pheochromocytoma
  - Large (> 3 cm) unilateral adrenal mass in older patient
- Ultrasound: Hypoechoic suprarenal lesion
  - Cystic components may be identified
- NECT: > 10 HU
- CECT
  - Delayed washout kinetics typically similar to adrenal carcinoma and metastases, but rapid washout (like adenoma) possible
  - Heterogeneous enhancement: Necrosis, cystic degeneration, hemorrhage
- MR
  - Traditional classic imaging feature: T2 ("light bulb") hyperintensity
    - Not specific or sensitive: Variable signal intensity due to hemorrhage, necrosis
  - Hypervascular solid components
- MIBG: For ectopic, recurrent, and metastatic tumors

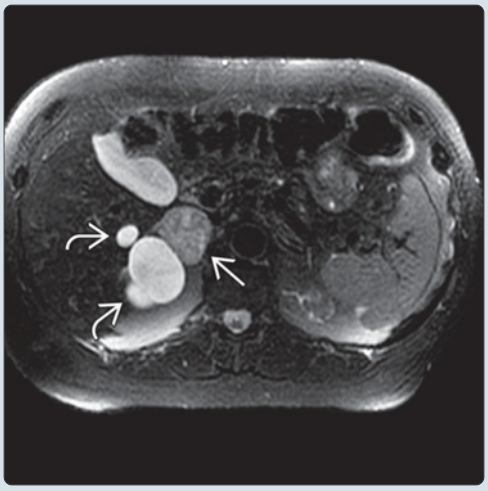
DIAGNOSTIC CHECKLIST

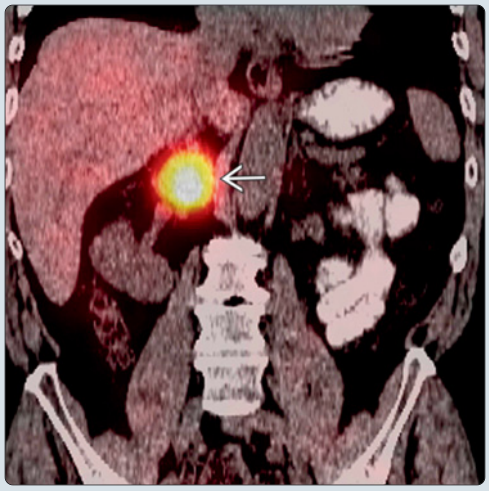
- Pheochromocytoma is not distinguished from other tumors by imaging appearance alone
  - Clinical history and lab values are necessary for diagnosis





(Left) *A right upper quadrant ultrasound performed on a 72-year-old man with vague right upper quadrant pain shows an isoechoic right suprarenal lesion. A dedicated adrenal CECT was recommended.* **(Right)** *CECT in the same patient shows a vascular right adrenal lesion. Vascularity and minimal washout prompted an evaluation for a pheochromocytoma. Note the adjacent right renal and hepatic cysts.*





(Left) *T2WI MR in the same patient shows a slightly T2-intense right adrenal lesion and adjacent renal/hepatic cysts. Elevated 24-hour urinary metanephrines indicated a (sporadic) unilateral pheochromocytoma, although the imaging appearance is nonspecific.* **(Right)** *I-123 MIBI CT-SPECT exam (performed to exclude metastases) confirms a solitary 4 cm right adrenal pheochromocytoma.*

TERMINOLOGY

Abbreviations

- Pheochromocytoma (pheo)

Definitions

- Tumor arising from chromaffin cells of adrenal medulla or sympathetic nervous system
  - Extraadrenal pheochromocytoma: Paraganglioma, ganglioneuroma

IMAGING

General Features

- Best diagnostic clue
  - Adrenal mass with appropriate clinical history and supporting biochemical studies
  - Characteristic T2 hyperintensity described in very early literature, though imaging appearance varies at MR (and other modalities)
- Location
  - Adrenal medulla (90%); extraadrenal (10%)
  - Extraadrenal, along sympathetic chain: Neck to urinary bladder
    - Subdiaphragmatic (98%); thorax (1-2%)
    - Organ of Zuckerkandl and near bladder are relatively common sites
- Size
  - Usually larger (> 3 cm) in sporadic (nonsyndromic) or clinically silent cases
- Morphology
  - Well-circumscribed, encapsulated tumor
    - Solitary (sporadic); multiple (familial)
- Key concepts
  - Classic teaching: "Rule of 10s" (or 10% tumor)
    - 10% extraadrenal: Paragangliomas
    - 10% bilateral
    - 10% malignant
    - 10% extraabdominal
    - 10% familial
    - 10% pediatric
    - 10% silent
    - 10% have autosomal dominant transmission
  - Demographics changing
    - Increased detection of incidental, clinically silent adrenal lesions (including pheochromocytomas) at CT
      - Large imaging series: 23-58% of pheochromocytomas clinically occult
      - Prompts aggressive biochemical evaluation of incidental adrenal lesion
    - Larger percentage of syndromic tumors likely: More aggressive screening for predisposing hereditary conditions
      - Multiple endocrine neoplasia 2
      - Neuroectodermal disorders: Neurofibromatosis, Sturge-Weber syndrome, Carney triad
      - 25% of patients with apparent sporadic pheochromocytomas are carriers of gene mutations
  - Extraadrenal tumors arise from sympathetic ganglia
    - Neck, mediastinum, pelvis, or urinary bladder
    - Aortic bifurcation (organ of Zuckerkandl): Ganglia at origin of inferior mesenteric artery
  - Imaging: Difficult to distinguish benign from malignant
    - Distant metastases indicate malignancy

CT Findings

- NECT
  - Attenuation ranges from low density to soft tissue attenuation
  - Attenuation almost always > 10 HU, though rare intracellular lipid-containing pheochromocytomas reported
  - ± areas of ↑ density (hemorrhage)
  - ± areas of ↓ density (cystic degeneration, necrosis)
  - ± areas of curvilinear or mural calcification
- CECT
  - Heterogeneous enhancement due to tissue necrosis, cystic degeneration, hemorrhage
  - Solid components briskly enhance
    - Hyperenhancement (> 110 HU) on late arterial phase may suggest pheochromocytoma
    - Early, marked enhancement also possible with vascular metastases (HCC, RCC)
  - Initial literature suggested pheochromocytoma washout characteristics similar to adrenal carcinoma and metastases
    - Recent work suggests large percentage of pheochromocytomas with washout percentages similar to adenomas
  - Theoretical risk of induction of hyperadrenergic symptoms with iodinated contrast administration discounted by retrospective series where nonionic material utilized

MR Findings

- T1WI
  - Isointense to muscle and hypointense to liver
  - Heterogeneous signal intensity
    - Due to areas of hemorrhage and necrosis
  - ± areas of increased signal intensity
    - Due to acute or subacute hemorrhage
- T2WI
  - Markedly hyperintense ("light bulb")
    - Due to ↑ water content as result of necrosis, cystic degeneration
    - Thought to be characteristic in early MR series: Discounted in follow-up literature
    - T2 heterogeneity typical; ↓ T2 signal does not exclude pheochromocytoma
- DWI
  - Utility for differentiating between malignant and benign pheochromocytomas suggested (ADC lower in malignant lesions)
- T1WI C+
  - Characteristic "salt and pepper" pattern (due to increased tumor vascularity)
    - "Salt": Represents enhancing parenchyma
    - "Pepper": Represents flow void of vessels
  - Can show marked early, as well as prolonged, contrast enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Variable echogenicity: Majority iso- to hypoechoic
    - Intralesional hemorrhage may be echogenic
  - Round and well-circumscribed mass
  - Cystic components may be identified

Nuclear Medicine Findings

- I-131 or I-123 metaiodobenzylguanidine (MIBG)
  - Most common and available technique
  - MIBG is norepinephrine analogue: Uptake proportional to number of neurosecretory granules within lesion
  - I-123 MIBG has largely replaced I-131 MIBG (lower radiation dose, improved image quality)
  - I-123 sensitivity: 77-90%; specificity: 95-100%
  - Hybridized MIBG SPECT/CT improves diagnostic accuracy
  - Particularly useful for extraadrenal paraganglioma detection, malignant pheochromocytoma staging
- Indium In-111 pentetreotide (somatostatin analog): Potentially useful for dedifferentiated pheochromocytoma
- PET: 18F-fluorodopamine, 18F-dihydroxyphenylalanine
  - Utility currently being investigated in research settings

Imaging Recommendations

- Helical NE + CECT
  - Hypertensive crises not documented with IV administration of nonionic contrast material
    - Routine **premedication** (α and β blockade) is **not recommended**
- MR ± contrast
- MIBG: For ectopic, recurrent, and metastatic tumors

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- NECT
  - Well-defined mass of < 10 HU (lipid rich)
- CECT: Enhancing mass that de-enhances rapidly
  - Early hyperenhancement and marked de-enhancement of pheochromocytomas are reported and may mimic adenomas
- Adenoma with hemorrhage or necrosis may resemble pheochromocytoma

Adrenal Carcinoma

- Rare; usually unilateral
- Large, unilateral adrenal mass with invasive margins
  - ± calcification (30% of cases); variable enhancement
  - Metastatic tumor spread: Lungs, liver, nodes, and bone
  - Inferior vena cava (IVC) tumor thrombus

Adrenal Metastases and Lymphoma

- Adrenal metastases
  - e.g., lung, breast, renal cell carcinoma, and melanoma
  - Unilateral or bilateral; central necrosis ± hemorrhage
  - History: Patient usually known to have malignancy
- Adrenal lymphoma
  - Usually as part of diffuse disease
    - Rarely limited to adrenals
  - Non-Hodgkin most common; usually bilateral
  - CECT: Mild enhancement (hypovascular)

Adrenal Myelolipoma

- Rare benign tumor (fat + hematopoietic elements)
- Unilateral fatty adrenal tumor (-100 to -30 HU)
- T1WI: Typically hyperintense; size varies (2-10 cm)
- Signal loss at T1WI with fat suppression

Adrenal Hemorrhage

- Etiology: Septicemia, burns, trauma, stress, hypotension, and hematological abnormalities
- CT findings
  - Usually bilateral
  - Old hemorrhage: Soft tissue attenuation (20-35 HU)
  - Recent hemorrhage: ↑ attenuation values
- MR findings: T1- and T2WI
  - Varied signal depending on hematoma age
  - Subacute phase: Usually ↑ signal (methemoglobin)
  - Perilesional dark ring (hemosiderin or ferritin)

Adrenal Tuberculosis and Fungal Infection

- e.g., tuberculosis, histoplasmosis, other fungal diseases
- Usually bilateral, heterogeneous, poorly enhancing (acute)
- Chronic: Small and calcified adrenals
- Diagnosis: Clinical history and lab data

PATHOLOGY

General Features

- Etiology
  - Chromaffin cells of adrenal medulla or extraadrenal paraganglioma
    - Adrenal medulla: Pheochromocytoma
    - Extraadrenal: Paraganglioma
- Associated abnormalities
  - Classic associated syndromes
    - von Hippel-Lindau (VHL) syndrome
      - Pheo may be only manifestation of VHL or occur along with other tumors
    - Neurofibromatosis type 1
    - Multiple endocrine neoplasia syndromes (MEN) types 2A and 2B
    - Tuberous sclerosis; Sturge-Weber syndrome
    - Carney syndrome
      - Pulmonary chondroma, gastric leiomyosarcoma, pheochromocytoma
  - Adage that only 10% of pheochromocytomas are hereditary discounted with recent advances in molecular studies
  - 25% of patients with previously considered sporadic pheochromocytomas are carriers of gene mutations
- Embryology/anatomy
  - Neoplasm of chromaffin cells derived from neural crest or neuroectoderm

Gross Pathologic & Surgical Features

- Round, tan-pink to violaceous encapsulated mass
- ± cystic, mucoid, serosanguineous hemorrhage

Microscopic Features

- Large cells: Granular cytoplasm and pleomorphic nuclei
- Chromaffin reaction: Cells stained + chromium salt

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Symptoms may be episodic or paroxysmal
  - Crisis: Headaches, hypertension, palpitations, sweating, tremors, arrhythmias, pain
  - Classic triad: Headache, palpitations, sweating
    - 90% specific, but uncommon presentation (10-36%)
  - Atypical: Labile HTN, myocardial infarction, stroke
  - Often clinically silent
- Clinical profile
  - Young patient with paroxysmal attacks of headache, palpitations, sweating, and tremors
- Lab data
  - 24-hour urine-fractionated metanephrine evaluation initial biochemical test at most institutions
    - Excretion of metanephrine, normetanephrine, and 3-methoxytyramine (dopamine metabolite) measured
    - Sensitivity: 90-97%; specificity: 69-98%
  - Plasma-fractionated metanephrines also measured, though low positive predictive value and ↓ specificity results in high false-positive rates

Demographics

- Age
  - Sporadic pheochromocytoma: Older patient (mean: 44 years)
  - Hereditary pheochromocytoma: Younger patient (mean: 25 years)
  - Pheochromocytomas are exceedingly rare in pediatric patients
    - Higher genetic predisposition and malignancy incidence
- Gender
  - F > M
- Epidemiology
  - Incidence
    - 0.13% in autopsy series; accounts for 0.1-0.5% of patients with hypertension
    - Prevalence likely underestimated

Natural History & Prognosis

- Complications: During hypertensive crisis
  - Cerebrovascular accidents (CVA)
  - Pregnancy + pheochromocytoma: Mortality (48%)
  - Malignancy in 2-14% cases
- Prognosis
  - Noninvasive and nonmetastatic: Good prognosis
  - Malignant and metastatic: Poor prognosis
    - 5-year survival rate: < 50%

Treatment

- Medical therapy: Before, during, and after surgery
  - α-adrenergic blockers
    - Phenoxybenzamine, phentolamine
  - β-adrenergic blocker: Propranolol
- Surgical resection: Benign and malignant
  - Laparoscopic resection preferred
    - Posterior retroperitoneoscopic adrenalectomy associated with decreased morbidity compared to transabdominal laparoscopic approach
    - Partial adrenalectomy may be performed with bilateral pheochromocytomas
- I-131 MIBG and tumor debulking for metastatic, incompletely resected malignant pheochromocytomas
- Combination chemotherapy: Cyclophosphamide + vincristine + dacarbazine

DIAGNOSTIC CHECKLIST

Consider

- Clinical history, supporting biochemical data ultimately drive diagnosis

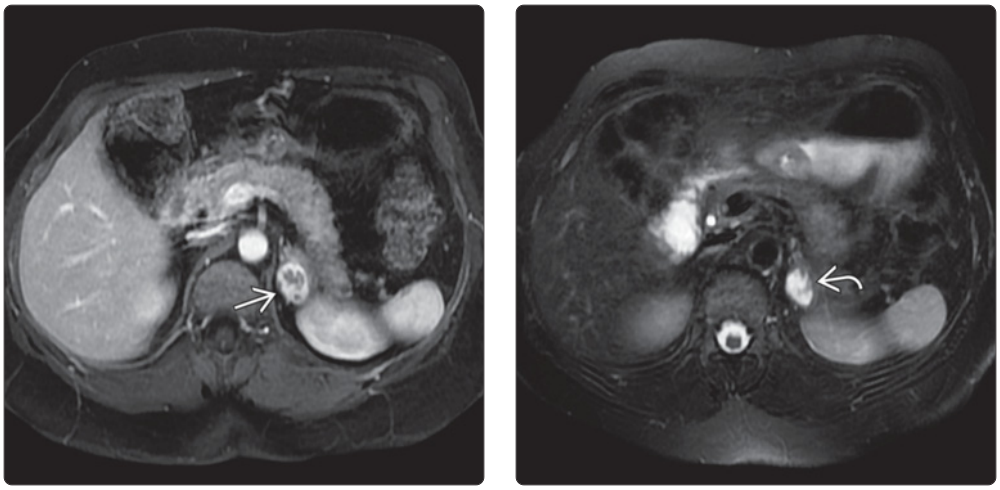
Reporting Tips

- CT and MR features (vascularity, cystic change, T2 hyperintensity) may suggest pheochromocytoma, but overlap precludes definitive diagnosis solely by imaging
- Possibility of pheochromocytoma should be raised, but lab analysis confirms or excludes diagnosis

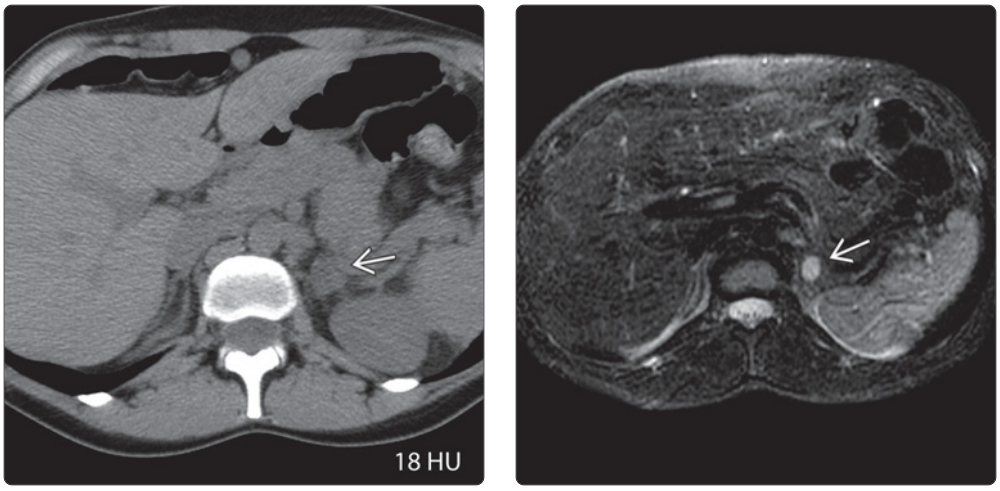
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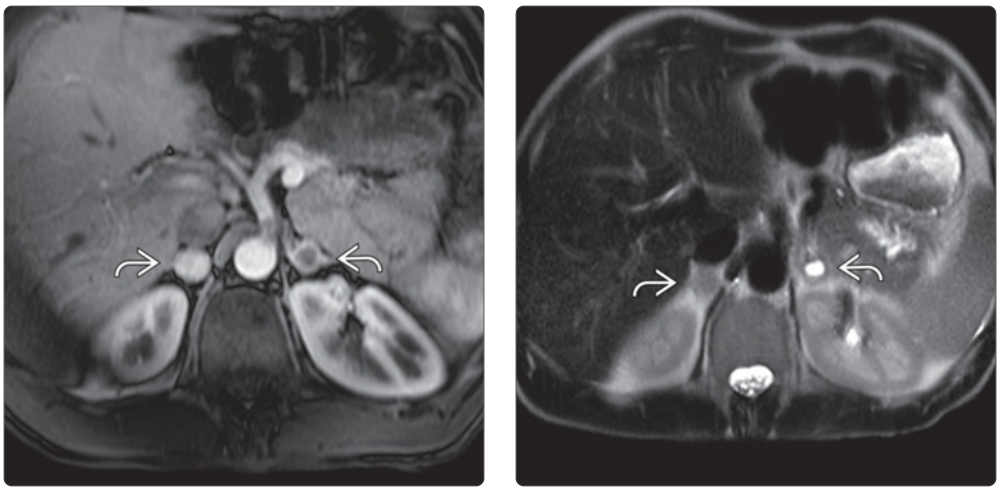
(Left) *T1WI contrast-enhanced MR performed to evaluate an incidental adrenal lesion previously identified on CT in this 52-year-old woman shows a 2 cm vascular left adrenal mass* ➡. **(Right)** *T2WI MR in the same patient shows a T2-hyperintense ("light bulb") left adrenal lesion* ➡. *Although this appearance was historically thought to be characteristic of pheochromocytomas, it is neither sensitive nor specific. Elevated 24-hour urine metanephrines confirmed a pheochromocytoma.*

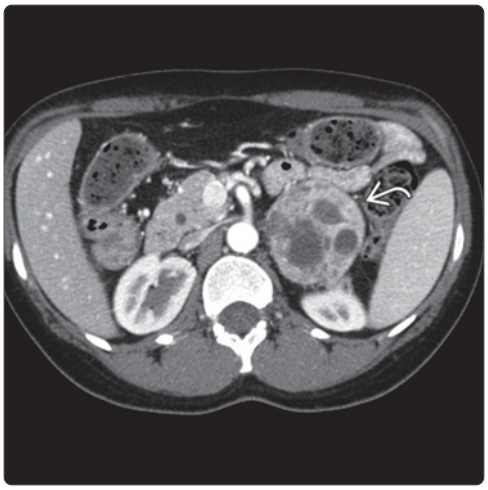


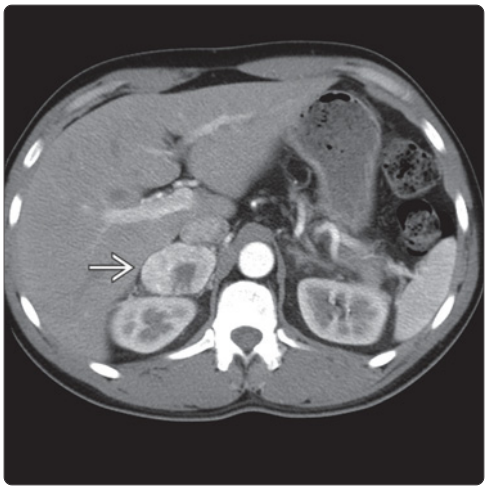
(Left) *NECT of a 32-year-old woman shows an incidental, indeterminate (18 HU) left adrenal lesion* ➡. *A family history of von Hippel-Lindau was elicited, and ↑ 24-hour urine metanephrines confirmed a hereditary pheochromocytoma.* **(Right)** *T2WI MR in the same patient shows a slightly T2-intense left adrenal pheochromocytoma* ➡. *The signal intensity of pheochromocytomas is dependent upon the extent of cystic degeneration, necrosis ± hemorrhage, which is characteristic of these lesions.*



(Left) *T1 FS MR in a 47-year-old man with MEN2a syndrome, metastatic medullary thyroid carcinoma, and ↑ 24-hour urine metanephrines shows small bilateral adrenal pheos* ➡. *Intralesional necrosis and hemorrhage accounts for the relatively diminished enhancement of the left lesion.* **(Right)** *T2WI MR of the same patient shows small (< 2 cm) bilateral pheochromocytomas* ➡. *Hereditary pheos are typically small and often bilateral.*

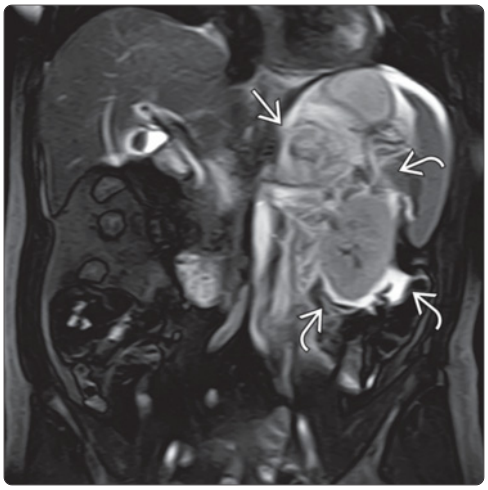




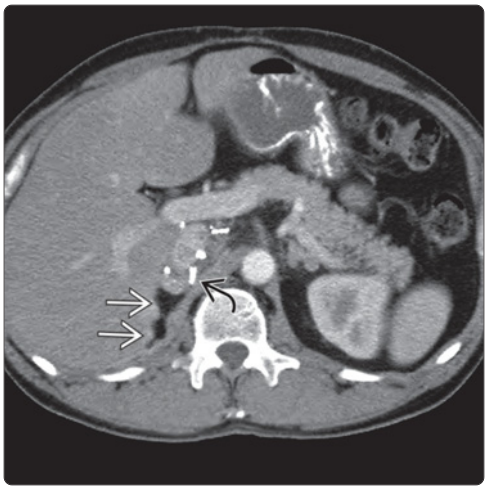


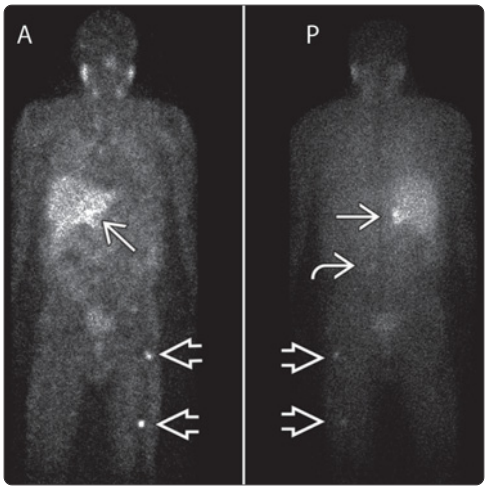
(Left) *CECT of a 36-year-old woman shows a 7 cm heterogeneous left adrenal mass. The differential includes adrenal carcinoma, pheo, or a metastasis. Urinary metanephrines were ↑, and laparoscopic resection (after a and β blockade) confirmed a hemorrhagic pheochromocytoma.* **(Right)** *CECT of a 26-year-old man shows a 4 cm right adrenal pheochromocytoma. Genetic testing, prompted by the patient's young age, confirmed an atypically large hereditary pheochromocytoma.*





(Left) *NECT of a 66-year-old woman, admitted with marked hypertension, flank pain, and an acute MI, shows retroperitoneal hemorrhage and a left adrenal mass. ATN after coronary angiography is suggested by dense nephrograms.* **(Right)** *T2WI of the same patient shows perinephric hemorrhage and a mixed signal, hemorrhagic left adrenal lesion. Increased urinary metanephrines confirmed a spontaneous hemorrhage of a pheo, a rare complication of these vascular lesions.*





(Left) *CECT of 49-year-old man, 3 years post right adrenal pheochromocytoma resection and retroperitoneal node dissection, shows recurrent perihepatic surface and retrocaval tumor.* **(Right)** *Anterior (A) and posterior (P) planar MIBG images of the same patient show locally recurrent tumor, retroperitoneal, and left femur metastases. MIBG imaging is useful for staging the small subset of patients with metastatic pheochromocytoma.*

Adrenal Carcinoma

KEY FACTS

IMAGING

- Best tool: CT or MR, including multiplanar views
- CECT shows most important aspects of tumor
  - Large, solid, unilateral adrenal mass with invasive margins (bilateral in 10%)
  - Functioning tumors: Usually 5 cm at presentation
  - Nonfunctioning tumors: ≥ 10 cm
  - Washout kinetics similar to adrenal metastases
- May contain hemorrhagic or cystic areas
  - Calcification within tumor seen in 30% of cases
- ± venous invasion, renal vein and IVC
- ± invasion into adjacent renal parenchyma
- Metastases to lung bases, liver, or nodes
  - Best shown on CT
- MR optimal for characterizing abdominal extent
  - T1 C+: Heterogeneous enhancement (tumor necrosis)
  - Renal vein, IVC, and adjacent renal parenchymal invasion well depicted on MR
  - Sagittal imaging helps to evaluate IVC invasion
  - Delineate tumor-liver interface if tumor on right
- PET/CT: Marked FDG uptake

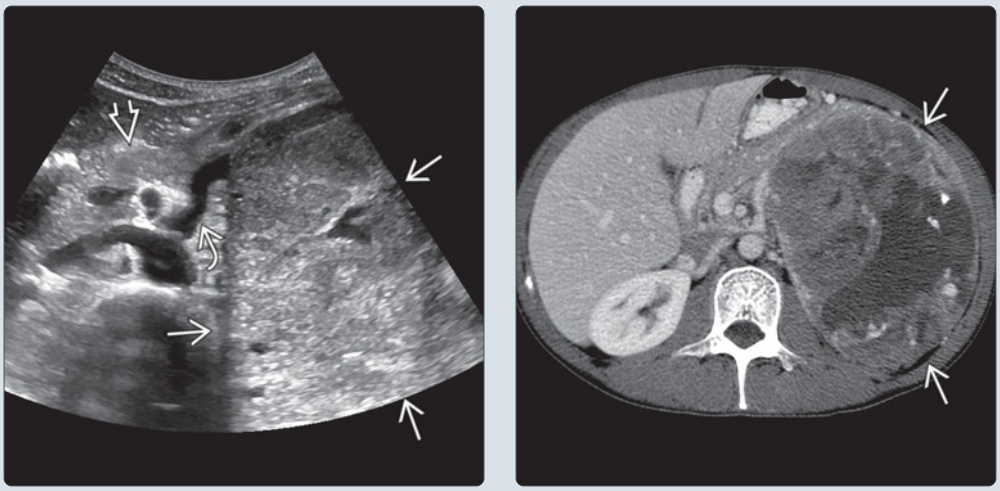
TOP DIFFERENTIAL DIAGNOSES

- Adrenal adenoma
- Adrenal metastases and lymphoma
- Adrenal myelolipoma
- Pheochromocytoma
- Upper pole of renal cell carcinoma (RCC)
- Adrenal hemorrhage (hematoma)
- Ganglioneuroma

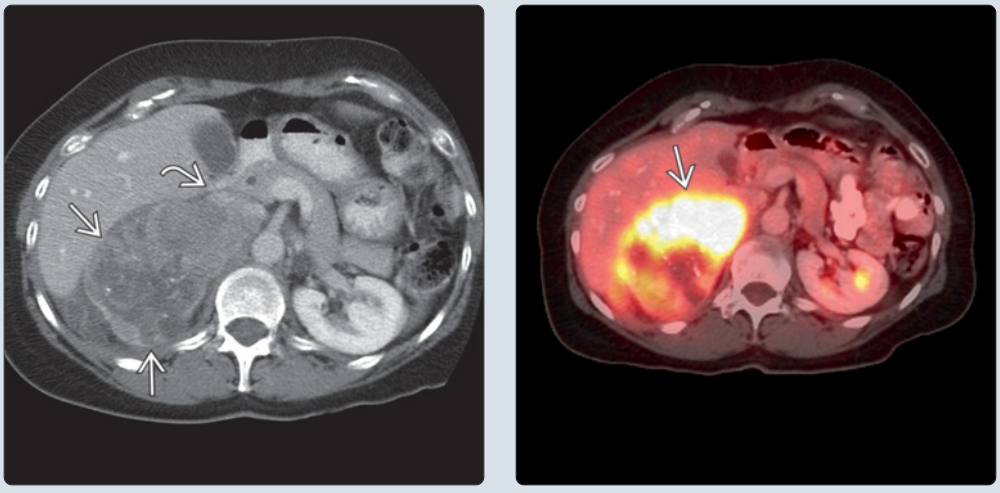
DIAGNOSTIC CHECKLIST

- Typical: Large, unilateral adrenal mass with invasive margins and venous nodal invasion, and lung metastases

(Left) *Ultrasound performed on a 34-year-old man shows a huge solid left retroperitoneal mass that displaces the left renal vein and pancreas anteriorly. The mass was separate from the left kidney at real-time sonography.* **(Right)** *CECT of the same patient shows a huge vascular and necrotic left upper quadrant mass. An endocrine evaluation was negative, and an oncocytic adrenal carcinoma was resected. Nonfunctional adrenal carcinomas are typically large and invasive at presentation.*



(Left) *CECT of a 55-year-old woman shows a 13 cm vascular, necrotic, right suprarenal mass that displaces and is inseparable from the IVC. An endocrine work-up was positive for ↑ urinary cortisol and metanephrines.* **(Right)** *PET/CT examination of the same patient shows marked FDG uptake in the nonnecrotic portions of the mass. An adrenal carcinoma was resected. Marked FDG uptake and venous invasion are characteristic features of this aggressive tumor.*



TERMINOLOGY

Synonyms

- Adrenocortical carcinoma, adrenal cancer

IMAGING

General Features

- Best diagnostic clue
  - Large, solid, unilateral adrenal mass with invasive margins (bilateral in 10%)
- Location
  - Suprarenal, usually unilateral
- Size
  - Functioning tumors: Usually 5 cm at presentation
  - Nonfunctioning tumors: ≥ 10 cm
- Morphology
  - Large suprarenal invasive lesion
  - Usually contain hemorrhagic, cystic, & calcified areas
- Key concepts
  - Rare & highly malignant neoplasm of adrenal cortex
  - Accounts for 0.05-0.2% of all cancers in USA
  - Usually unilateral; bilateral in up to 10% of cases
  - Rarely symptomatic incidental imaging finding
  - Local spread: Renal vein, inferior vena cava (IVC)
  - Metastatic spread: Lungs, liver, nodes, bone
    - 20% of cases metastatic at presentation
  - Most patients at stage III or IV at time of diagnosis
    - Due to early invasion of renal vein of IVC
  - Functioning (< 50%); nonfunctioning (> 50%)
  - Clinical syndromes with functioning tumors
    - Cushing syndrome most common (30-40%)
    - Virilization in females (20-30%)
    - Conn syndrome, feminization (males) < common
  - Accounts for only 0.002% of all childhood cancers
    - Mostly functional; virilization seen in 95% cases

CT Findings

- Solid, well-defined suprarenal mass + invasive margins
- Usually unilateral; may be bilateral in 10% of cases
- ± areas of necrosis, hemorrhage, rarely fat
- Calcification within tumor seen in 30% of cases
- Variable enhancement (necrosis and hemorrhage)
- ± venous invasion, renal vein and IVC
- ± invasion into adjacent renal parenchyma
- Metastases to lung bases, liver, or nodes
- Washout kinetics similar to adrenal metastases

MR Findings

- T1WI
  - Hypointense adrenal mass compared to liver
- T2WI
  - Hyperintense adrenal mass compared to liver
- T1 C+: Heterogeneous enhancement (tumor necrosis)
- Multiplanar contrast-enhanced imaging
  - Renal vein, IVC, and adjacent renal parenchymal invasion well depicted on MR
  - Sagittal, coronal imaging helps to evaluate IVC invasion
  - Delineate tumor-liver interface if right-sided lesion

Ultrasonographic Findings

- Grayscale ultrasound
  - Variable appearance depending on size & contents
  - Large tumors: Mixed heterogeneous echo pattern (due to areas of necrosis & hemorrhage)

Angiographic Findings

- Enlarged adrenal arteries: minimal neovascularity
- Arterial supply may aid in defining organ of origin for problematic cases
- Inferior venacavography: Confirms tumor invasion

Nuclear Medicine Findings

- FDG PET
  - Marked FDG uptake

Imaging Recommendations

- Best imaging tool
  - Either CT or MR, including multiplanar views
- NECT + CECT: Study of choice to exclude adenoma
  - Thin section acquisition: Coronal/sagittal reconstructions for preoperative planning

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- Well-defined soft tissue adrenal mass of 0-30 HU
  - Lipid-rich adenoma: Low-density (< 10 HU) mass
  - Lipid-poor adenoma: Varied density of 10-30 HU
- Enhancing adrenal mass that "de-enhances" rapidly
- Washout of adenoma: 15 min post injection (> 60% absolute or > 40% relative washout)

Adrenal Metastases and Lymphoma

- Adrenal metastases
  - Unilateral or bilateral masses ± necrosis, hemorrhage
  - Mostly hypervascular with prolonged washout pattern
  - Usually known to have malignancy elsewhere
- Adrenal lymphoma
  - Primary (rare); secondary (non-Hodgkin common)
  - Often bilateral; retroperitoneal disease usually seen
  - Discrete or diffuse mass, shape is maintained

Adrenal Myelolipoma

- Intramural macroscopic fatty elements

Pheochromocytoma

- Tumor > 3 cm in most cases
- Highly vascular tumor prone to hemorrhage, necrosis
- Tumors may be hyperintense on T2WI
- Clinical presentation and lab data critical for diagnosis

Upper Pole of Renal Cell Carcinoma (RCC)

- Large upper pole RCC mimics large adrenal carcinoma
- Angiography
  - Selective injection of renal artery: Downward displacement of kidney with tumor vascularity
  - Selective injection of superior or middle adrenal arteries: Opacifies tumor and confirms adrenal etiology

Adrenal Hemorrhage (Hematoma)

- Acute: High-density fluid collection (40-60 HU)
- Chronic: Large, well-defined, low-density hematoma

- No enhancement of hematoma on CECT

Ganglioneuroma

- Younger patients (mean age: 27 years)
- Larger mass with average tumor size 8 cm

PATHOLOGY

General Features

- Etiology
  - Unknown for sporadic adrenal carcinoma
- Genetics
  - Usually shows loss of heterozygosity at some loci
  - More likely to be aneuploid or tetraploid
  - Genetic syndromes may ↑ incidence of tumor
- Associated abnormalities
  - May be associated with genetic syndromes
    - Beckwith-Wiedemann, Li-Fraumeni, Carney, and MEN type 1

Staging, Grading, & Classification

- Staging of adrenal carcinoma
  - T1: Diameter ≤ 5 cm without local invasion
  - T2: Diameter > 5 cm without local invasion
  - T3: Any tumor size with local invasion but not involving adjacent organs
  - T4: Any tumor size with local invasion and extension into adjacent organs

Gross Pathologic & Surgical Features

- Usually large and heterogeneous
- Necrotic, hemorrhagic, calcific, lipoid, and cystic areas

Microscopic Features

- Well-differentiated to markedly anaplastic cells

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Presentation with non-hormonally-active malignancy
    - Abdominal pain, fullness, or palpable mass
    - Incidentally discovered mass on imaging exam
    - Metastatic disease in lung, liver, ± bone (20% at presentation)
    - 54% of tumors are nonfunctioning at presentation
- Presentation with hormonally active malignancy
  - Cushing syndrome (30-40%): ↑ cortisol
    - Moon facies, truncal obesity, purple striae, and buffalo hump
  - Virilization in females (20-30%): ↑ androgens
    - 95% of children with functioning adrenal carcinoma present with virilization
  - Conn syndrome (primary hyperaldosteronism)
    - Hypertension and weakness
  - Feminization in males: ↑ androgens
- Other clinical syndromes at presentation
  - Hypoglycemia, polycythemia, and non-glucocorticoid-related insulin resistance

Demographics

- Age
  - Bimodal distribution
    - 1st peak < age 5
    - 2nd peak in 4th-5th decades
- Gender
  - Overall, M < F
    - Females account for 65-90% of all cases
    - Functioning tumors: More common in females
    - Nonfunctioning tumors: More common in males
- Epidemiology
  - 0.05-0.2% of all cancers
  - 2 new cases per million annually
  - 1 per 1,500 adrenal tumors are malignant
  - 20% have metastatic disease at presentation

Natural History & Prognosis

- Rapid growth with local invasion and distant metastases
- Tumor thrombus: IVC and renal vein
- Mean survival: 18 months; children better than adults
- 5-year survival for stage III disease < 30%
- Stages I and II: Good prognosis after surgical removal
- Stage III and IV: Poor prognosis ± treatment

Treatment

- Small lesions: Laparoscopic adrenalectomy
- Large lesions with extension: Radical resection of ipsilateral kidney, adrenal gland, adjacent structures
- Metastatic sites also resected as possible
- Chemotherapy: Mitotane, cisplatin, 5-FU, & suramin

DIAGNOSTIC CHECKLIST

Consider

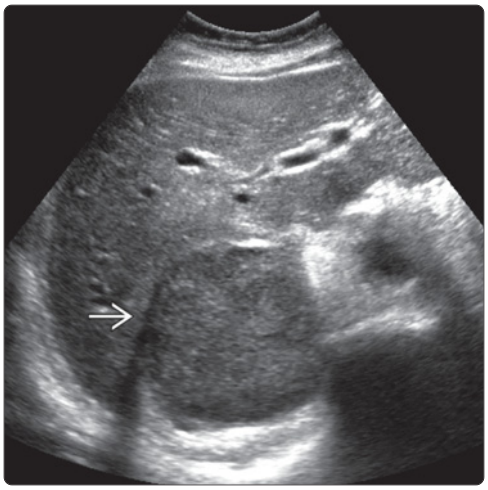
- Rule out other adrenal tumors, especially adenoma

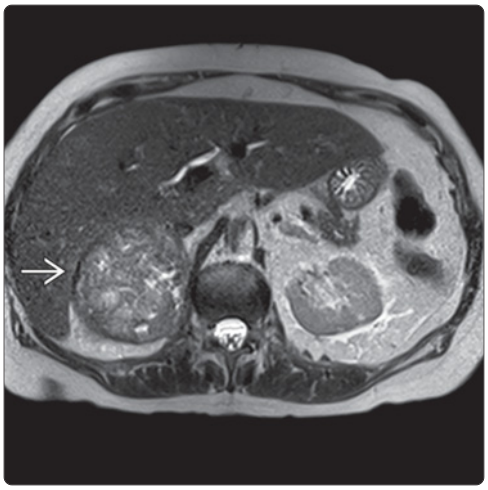
Image Interpretation Pearls

- Large, unilateral adrenal mass with invasive margins and venous or nodal invasion and distant metastases
- Precise definition of cephalad extension of tumor venous thrombus is essential for surgical resection

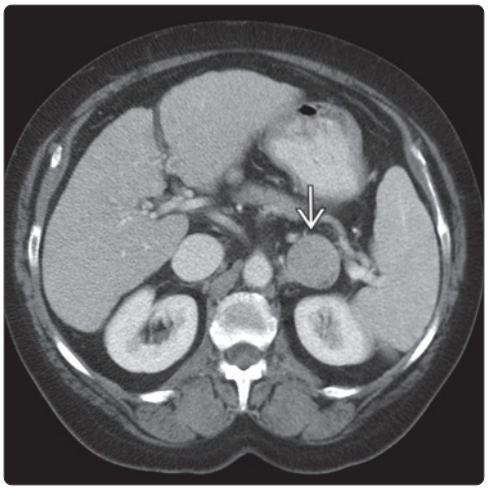
SELECTED REFERENCES

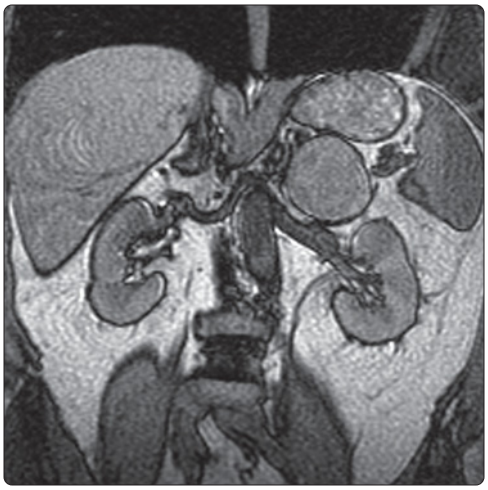
1. Fassnacht M et al: Update in adrenocortical carcinoma. J Clin Endocrinol Metab. 98(12):4551-64, 2013
2. Bharwani N et al: Adrenocortical carcinoma: the range of appearances on CT and MRI. AJR Am J Roentgenol. 196(6):W706-14, 2011
3. Bilimoria KY et al: Adrenocortical carcinoma in the United States: treatment utilization and prognostic factors. Cancer. 113(11):3130-6, 2008
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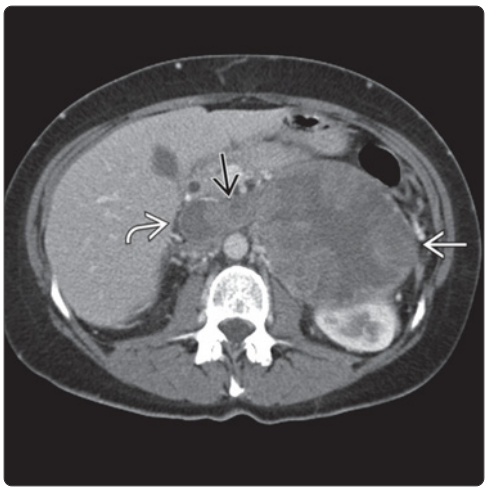


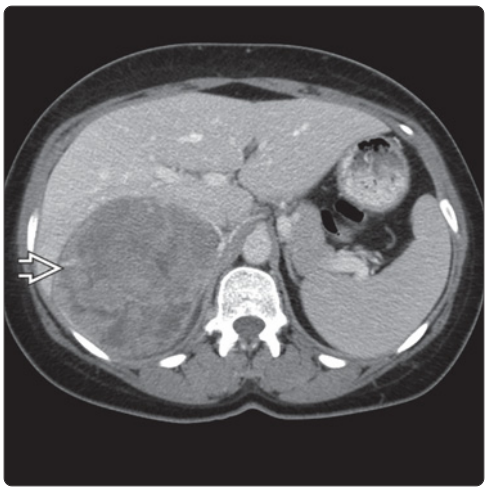
(Left) *Ultrasound shows a solid right suprarenal mass. The DDx of large, hypoechoic solid adrenal lesions is nonspecific but it includes metastases, pheochromocytomas, and adrenal carcinomas.* **(Right)** *T2 HASTE MR of the same patient shows a mixed-intensity right adrenal mass. An adrenal carcinoma adherent to the liver was identified during a debulking attempt. The prognosis for most adrenal carcinomas is dismal, and chemotherapeutic options are limited.*





(Left) *Surveillance CECT of a 59-year-old woman with cirrhosis showed growth of an enhancing 4 cm indeterminate left adrenal mass. A follow-up adrenal CT showed that relative washout of the lesion was < 40%.* **(Right)** *T1 MR, opposed-phase MR of the same patient shows no loss of signal from the left adrenal lesion. Growth and imaging features ruling out either a lipid-rich or -poor adenoma prompted resection. A low-grade T1 stage adrenal carcinoma with vascular invasion was resected.*





(Left) *CECT shows a huge left suprarenal mass, with tumor thrombus in the left renal vein and cava. RCC was suspected at both imaging and surgery, but invasive adrenal carcinoma was identified at pathology.* **(Right)** *CECT shows a huge right suprarenal mass. A nonfunctioning adrenal carcinoma was suspected at both CT and surgery, but clear cell RCC was identified within the en-bloc specimen. Both cases demonstrate the difficulty of differentiating adrenal carcinoma from upper-pole RCC.*

T | Primary Tumor

TNM	Definitions
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor ≤ 5 cm in greatest dimension, no extraadrenal invasion
T2	Tumor > 5 cm, no extraadrenal invasion
T3	Tumor of any size with local invasion but not invading adjacent organs; (adjacent organs include kidney, diaphragm, great vessels, pancreas, spleen, and liver)
T4	Tumor of any size with invasion of adjacent organs

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastases
N1	Metastasis in regional lymph node(s)

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis

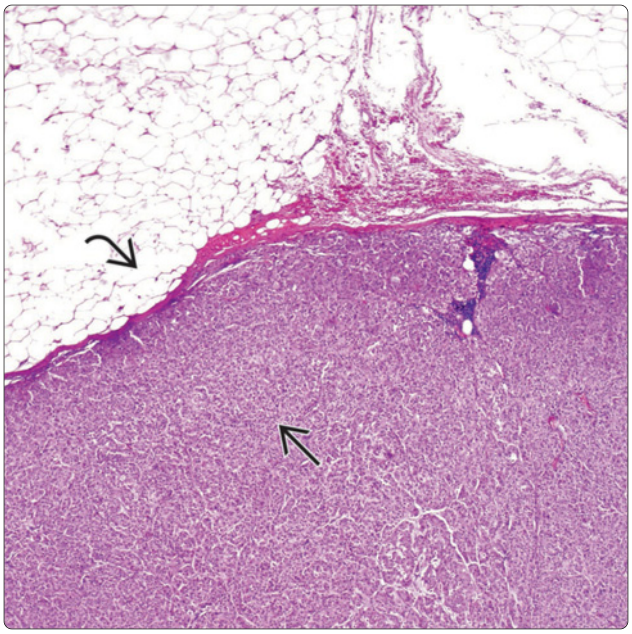
Adapted from 7th edition AJCC Staging Forms (2010).

AJCC | Stages/Prognostic Groups

Stage	T	N	M
I	T1	N0	M0
II	T2	N0	M0
III	T1	N1	M0
	T2	N1	M0
	T3	N0	M0
IV	T3	N1	M0
	T4	N0	M0
	T4	N1	M0
	Any T	Any N	M1

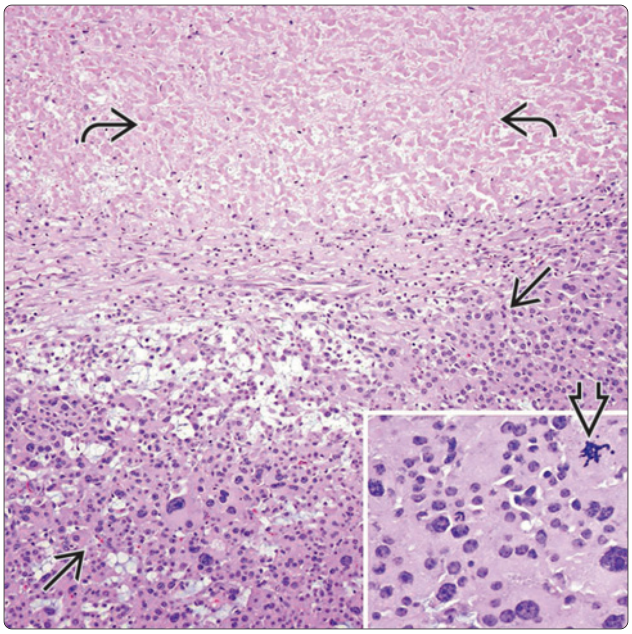
Adapted from 7th edition AJCC Staging Forms (2010).

T1



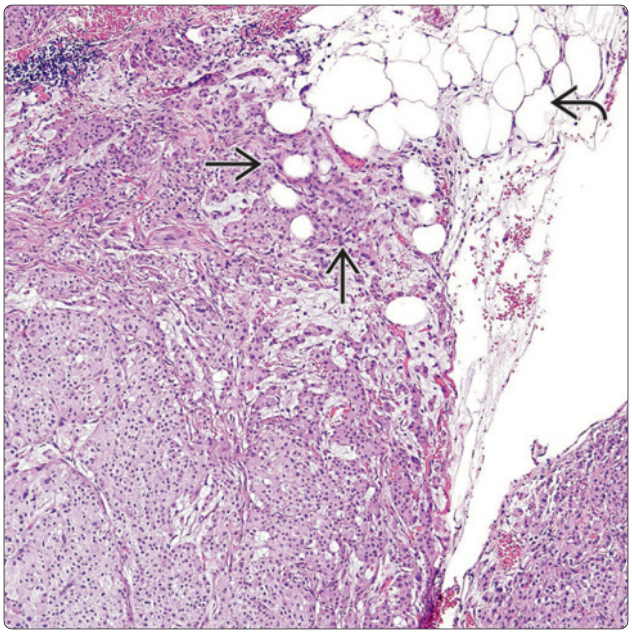
Low-power magnification of an H&E section shows sheets of adrenal cortical neoplastic cells ⇨ that are limited to the adrenal gland (T1). Adipose tissue ⇨ surrounding the adrenal gland is negative for tumor. (Original magnification 40x.)

T2



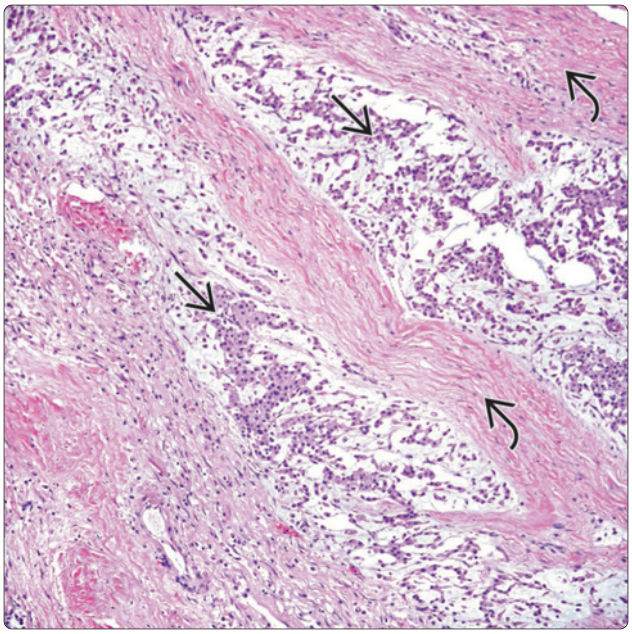
Photomicrograph shows an H&E section from adrenal cortical carcinoma ⇨ with large areas of necrosis in the upper aspect of the slide ⇨. The inset shows the particularly striking nuclear pleomorphism (large vs. small nuclei) as well as a mitotic figure ⇨. (Original magnification 400x.)

T3



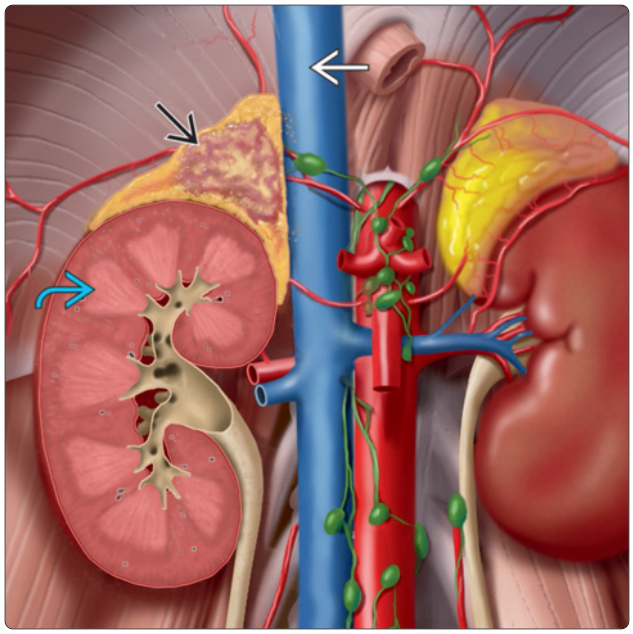
H&E stain shows tumor with local invasion but no extension into adjacent organs, consistent with T3 disease. The adrenal cortical carcinoma invades ⇨ locally into the surrounding adrenal fat ⇨. (Original magnification 100x.)

T4



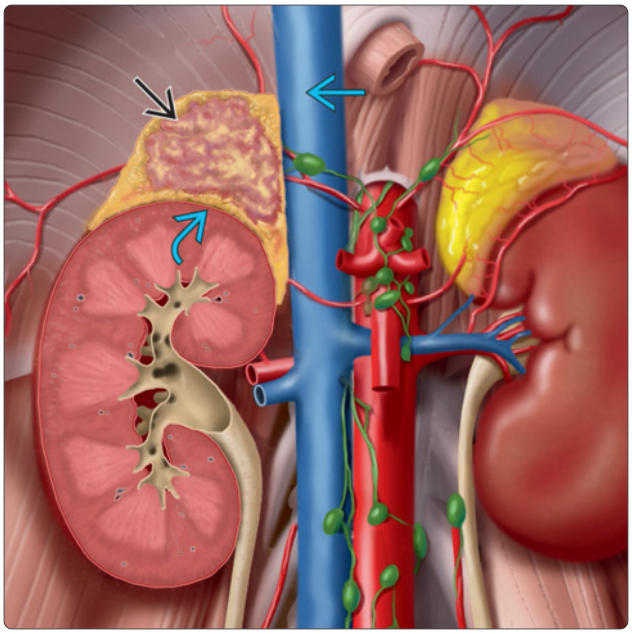
In T4 disease, tumor invades into adjacent organs. This tumor extends to involve the diaphragm. This photomicrograph shows the neoplastic tumor cells ⇨ extending into surrounding fibrotic connective tissue ⇨. (Original magnification 400x.)

T1



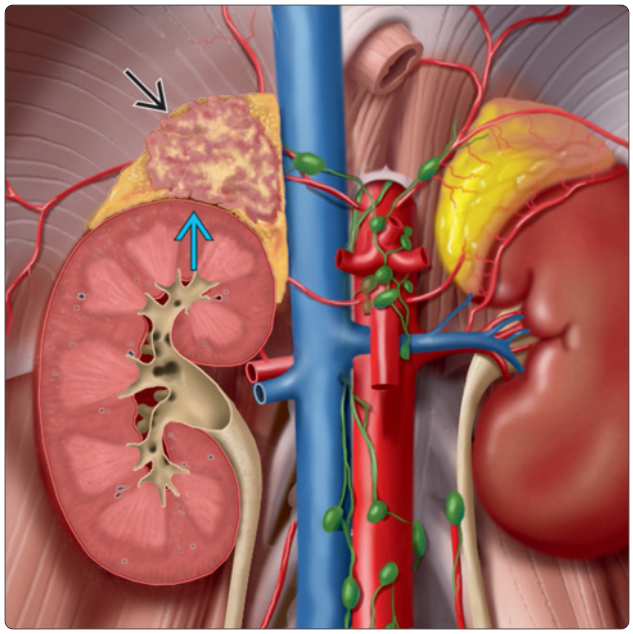
Coronal graphic demonstrates T1 disease. The primary tumor is ≤ 5 cm in greatest dimension, without invasion of adjacent organs, including kidney or inferior vena cava.

T2



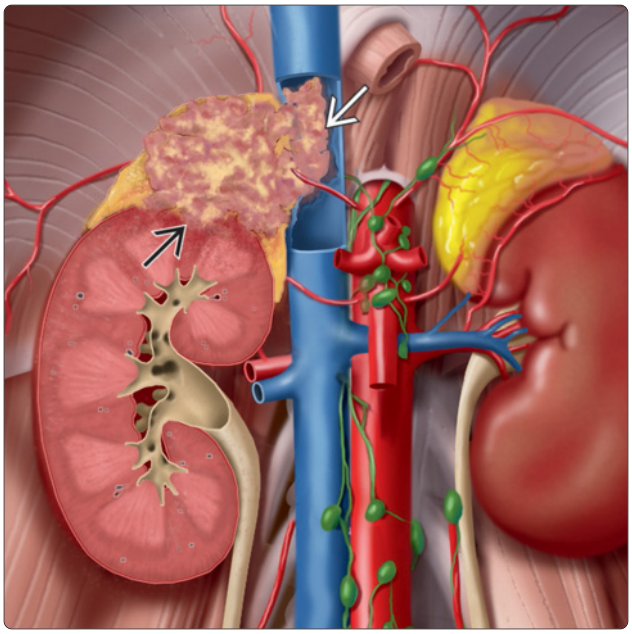
Coronal graphic demonstrates T2 disease. The primary tumor is > 5 cm in greatest dimension, without invasion of adjacent organs, including kidney or inferior vena cava.

T3



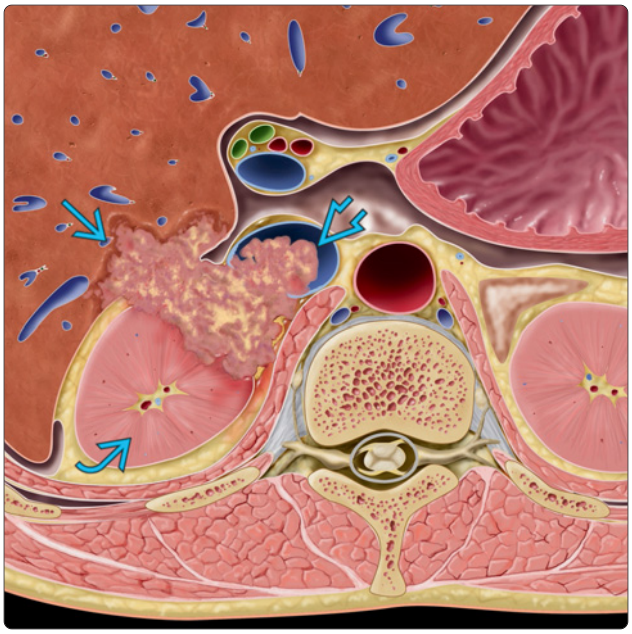
Coronal graphic demonstrates T3 disease. The primary tumor may be any size, with local invasion beyond the confines of the adrenal capsule, shown in the superolateral margin, but no involvement of adjacent organs such as the kidney.

T4



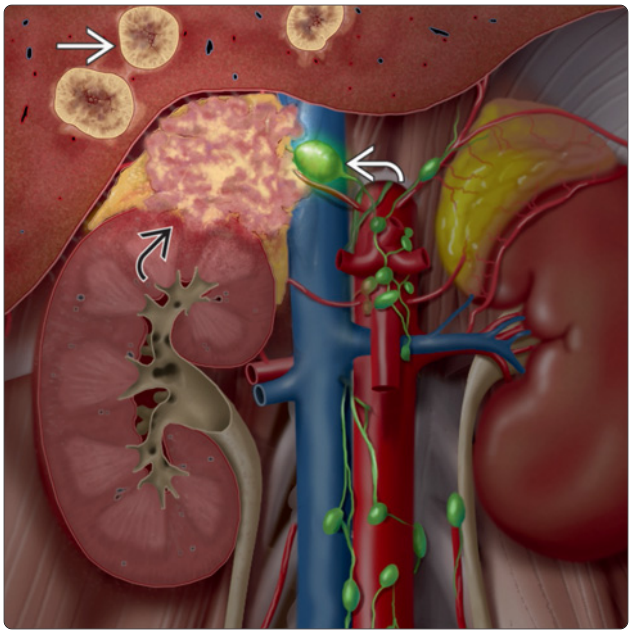
Coronal graphic demonstrates T4 disease. The primary tumor can be any size, with local invasion beyond the confines of the adrenal capsule and into adjacent organs, including the kidney. Direct extension into the inferior vena cava is illustrated.

T4



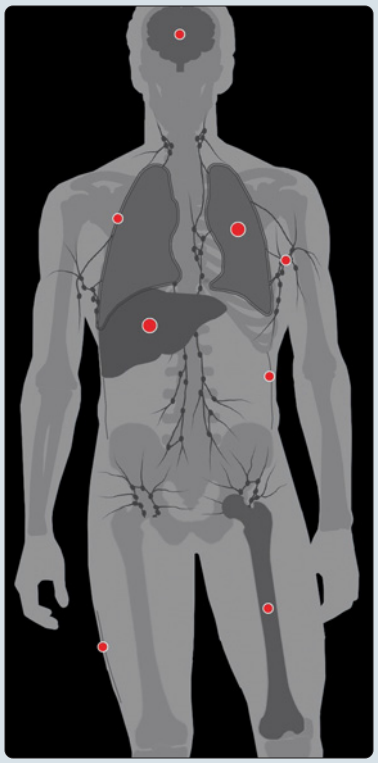
Axial graphic demonstrates right-sided adrenal cortical carcinoma invading adjacent organs, including the right kidney, the liver, and the inferior vena cava.

N1 and M1



Coronal graphic shows primary adrenal cortical carcinoma with invasion into adjacent kidney. N1 and M1 disease with an enlarged paraaortic lymph node and multifocal hepatic metastases are also illustrated.

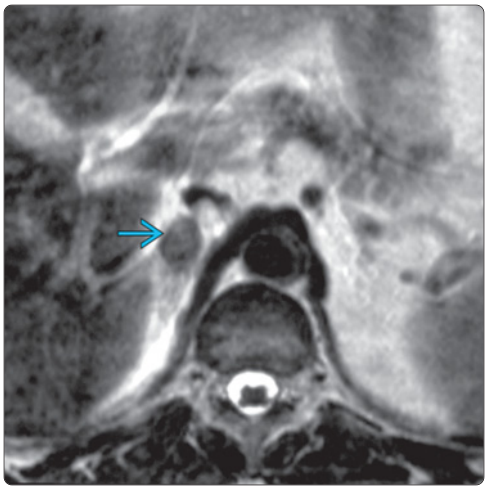
Metastases, Organ Frequency



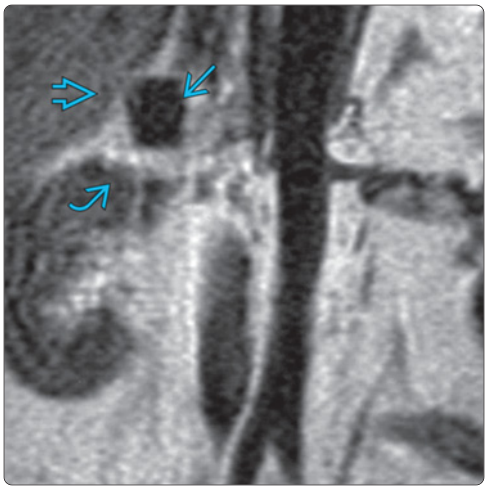
Liver	*11%*
Lung	*9%*
Bone	*3%*
Distant lymph nodes	*2%*
Peritoneum	*1%*
CNS	*< 1%*
Pleura	*< 1%*
Skin	*< 1%*

Data from Bilimoria KY et al: Adrenocortical carcinoma in the United States: treatment utilization and prognostic factors. Cancer. 113(11):3130-6, 2008.

Stage I (T1 N0 M0)

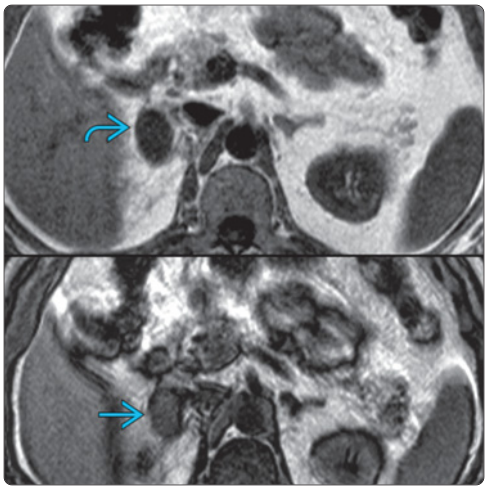


Stage I (T1 N0 M0)

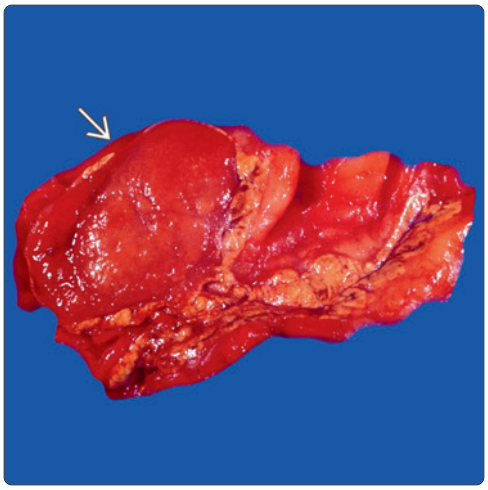


(Left) *Axial T2WI MR demonstrates a well-circumscribed T2 dark adrenal nodule, measuring < 3 cm.* **(Right)** *Coronal T1WI MR shows a T1 dark, well-circumscribed right adrenal nodule. There is a clear fat plane between the nodule and the adjacent kidney and liver.*

Stage I (T1 N0 M0)

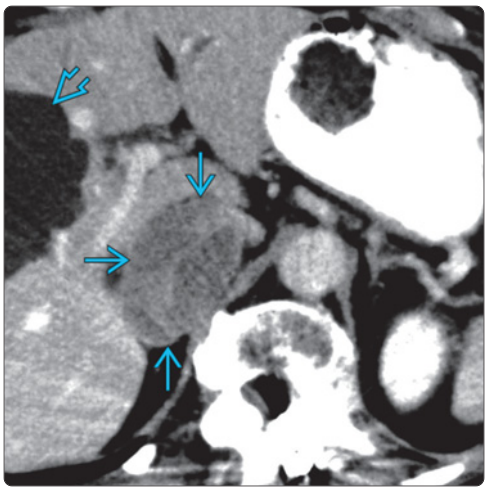


Stage I (T1 N0 M0)

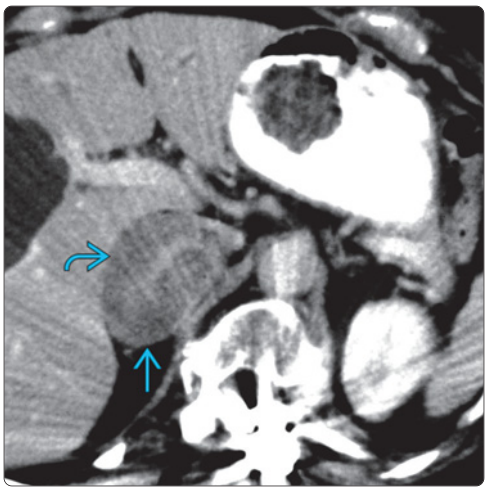


(Left) *Axial T1WI MR in the same patient demonstrates that the nodule gains signal intensity on the out of phase imaging (bottom) compared to the in-phase imaging (top). A benign adrenal nodule would lose signal on the out of phase imaging.* **(Right)** *Gross image shows the well-circumscribed rubbery adrenal mass corresponding to the lesion seen in the MR images in the same patient.*

Stage I (T1 N0 M0)

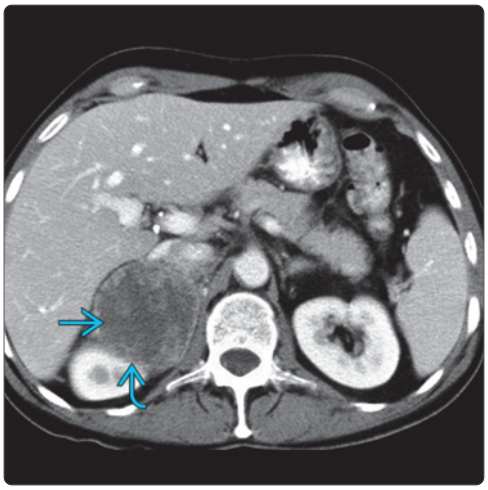


Stage I (T1 N0 M0)

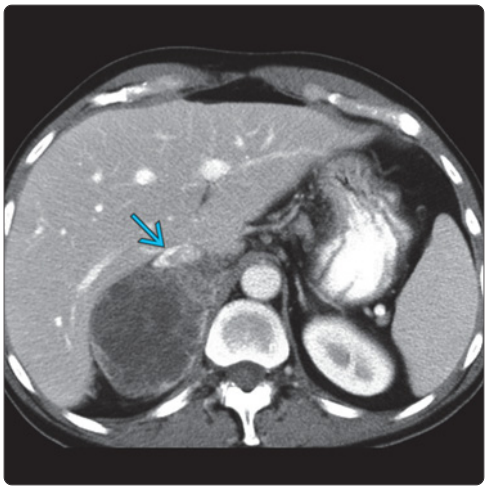


(Left) *Axial CECT shows a 4.5 cm, moderate-sized, heterogeneous right adrenal tumor. This typical appearance of an adrenal cortical carcinoma overlaps with the appearance of metastases and pheochromocytoma on CT. An incidental hepatic cyst is seen.* **(Right)** *A more inferior image in the same patient at the level of the portal vein shows that the tumor is confined within the adrenal capsule and that an intact plane exists between the mass and the liver.*

Stage II (T2 N0 M0)

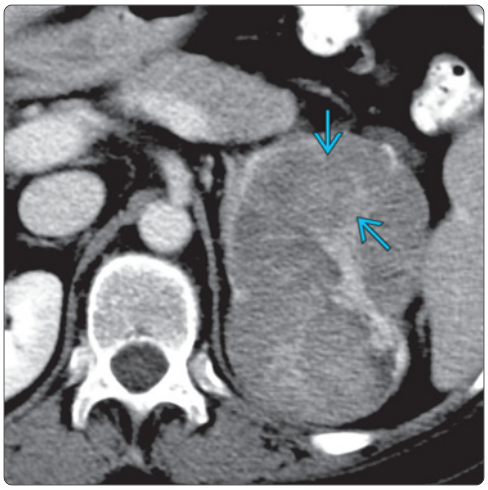


Stage II (T2 N0 M0)

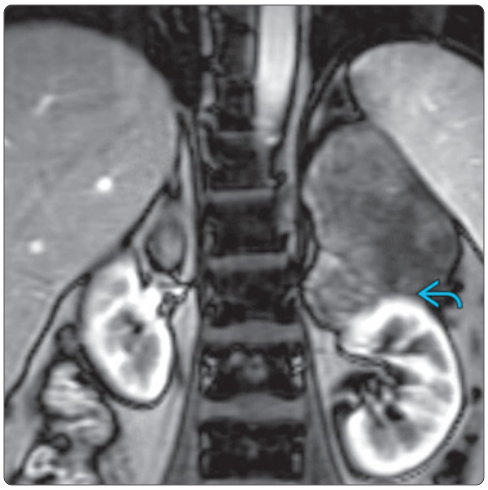


(Left) *Axial CECT shows a large heterogeneous right adrenal mass with areas of low density. Note preservation of distinct margin of right kidney, denoting that the mass neither arises from nor invades the kidney. The mass size > 5 cm makes the lesion T2.* **(Right)** *Axial CECT in the same patient more cranially shows intact planes between the mass and adjacent organs, including liver. The inferior vena cava is compressed but uninvolved.*

Stage II (T2 N0 M0)

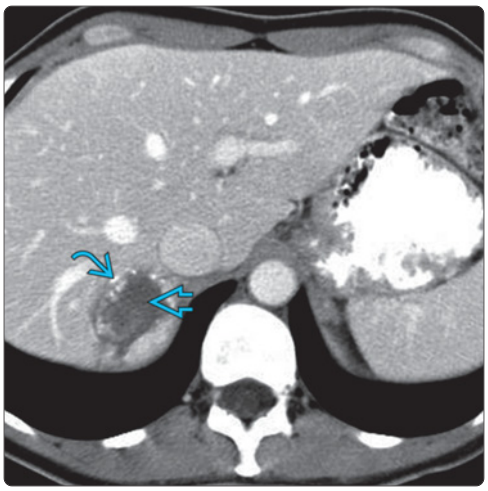


Stage II (T2 N0 M0)

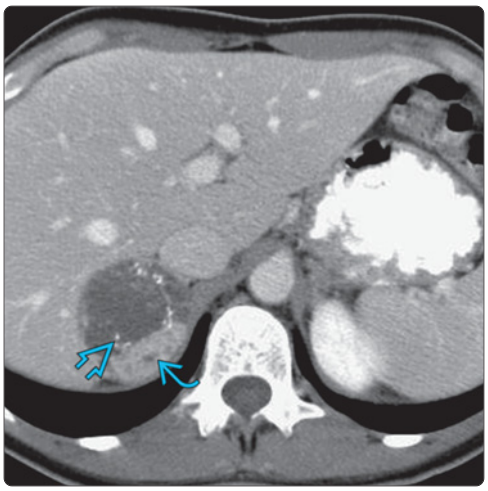


(Left) *Multimodality imaging can be very helpful for characterizing tumor margins for preoperative staging. This CECT demonstrates a large (> 5 cm), heterogeneous left adrenal mass that appears very well circumscribed in a 30-year-old woman presenting with Cushing syndrome.* **(Right)** *Coronal T1WI C+ MR in the same patient suggests that an intact plane is present between the mass and the adjacent kidney. This was confirmed on resection.*

Stage III (T3 N0 M0)

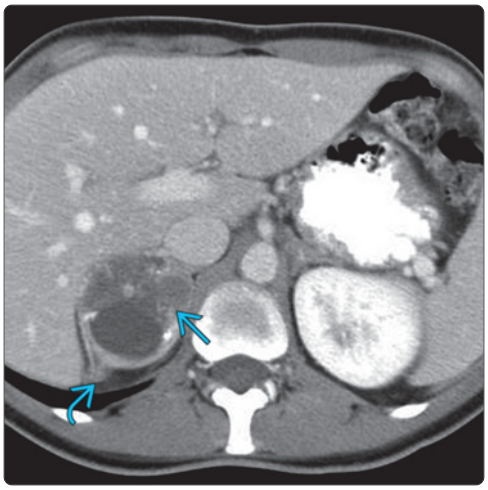


Stage III (T3 N0 M0)

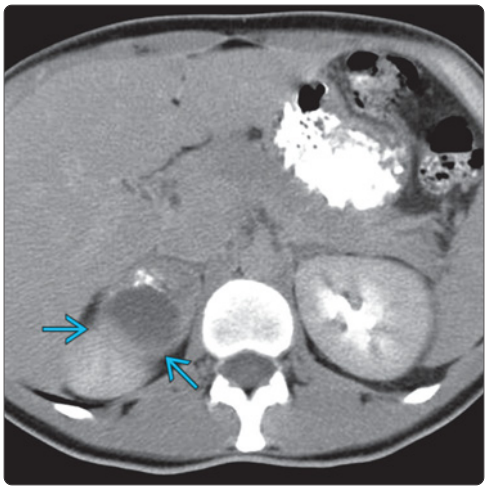


(Left) *Axial CECT demonstrates a right adrenal mass with areas of calcification as well as necrosis.* **(Right)** *A more caudal image in the same patient again shows the mass demonstrating extensive necrosis with calcification and areas of heterogeneous enhancement. On this image, an intact tissue plane is suggested between the mass, liver, diaphragm, and inferior vena cava.*

Stage III (T3 N0 M0)

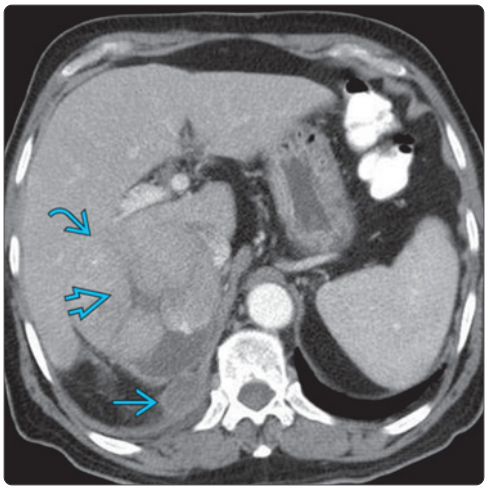


Stage III (T3 N0 M0)

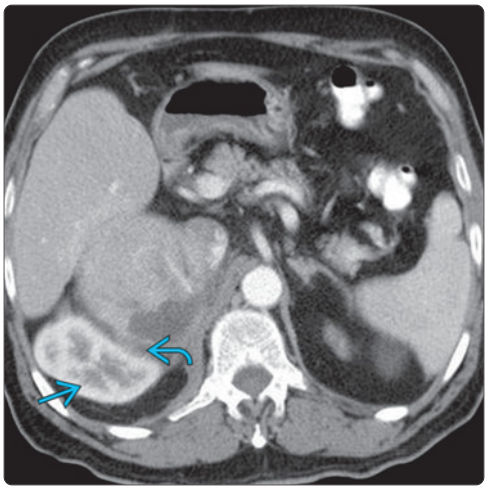


(Left) *Axial CECT in the same patient shows a suggestion of soft tissue nodularity and infiltration in the adjacent fat, which would indicate T3 disease. An intact plane is difficult to appreciate, but it is present between the mass and the liver.* **(Right)** *Delayed CECT in the same patient illustrates the difficulty of documenting an intact plane between the mass and the kidney, especially on delayed CECT.*

Stage IV (T4 N0 M0)

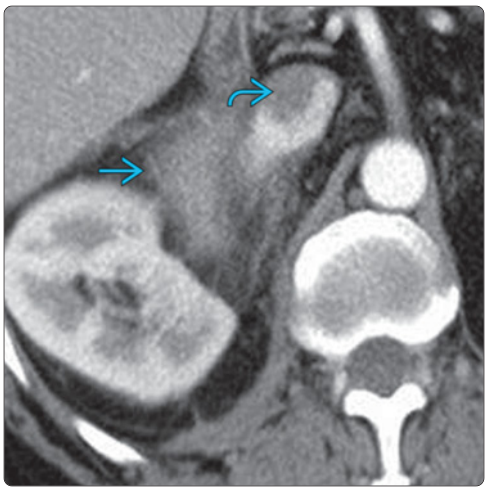


Stage IV (T4 N0 M0)

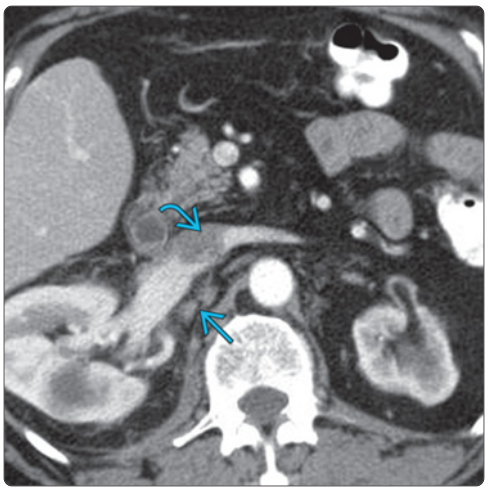


(Left) *Axial CECT shows a very large, partially necrotic, heterogeneous adrenal mass with infiltration beyond the adrenal capsule into adjacent perinephric space. The infiltration into the right diaphragm makes this T4 disease. No distinct plane is seen between the mass and adjacent liver.* **(Right)** *Axial CECT in the same patient shows the infiltrated fat between the mass and the kidney. There is no frank invasion into the adjacent kidney.*

Stage IV (T4 N0 M0)

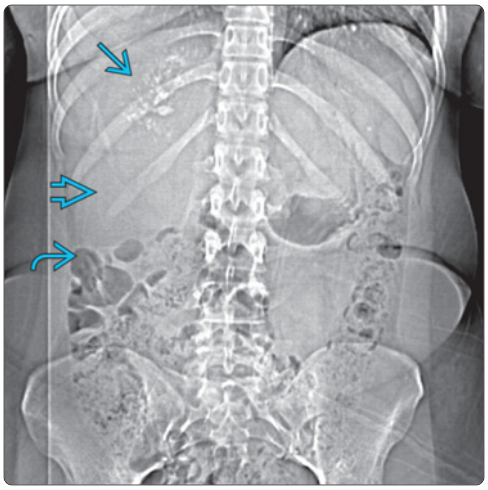


Stage IV (T4 N0 M0)

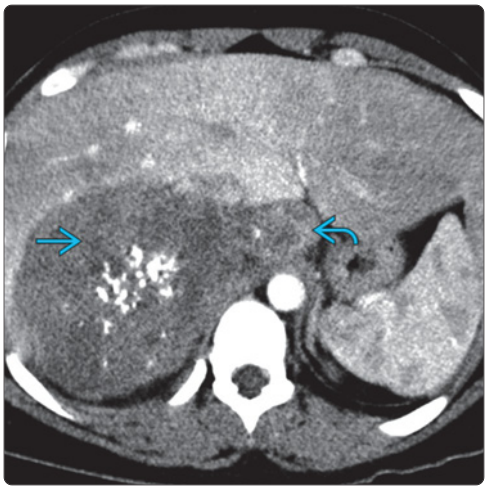


(Left) *Axial CECT in the same patient shows the infiltration of fat between mass and kidney. Additionally, intraluminal tumor thrombus is seen within the inferior vena cava.* **(Right)** *More caudal axial CECT in the same patient demonstrates inferior extension of intraluminal tumor thrombus within the inferior vena cava. Small nodes do not meet CT criteria for pathologic enlargement.*

Stage IV (T3 N1 M0)

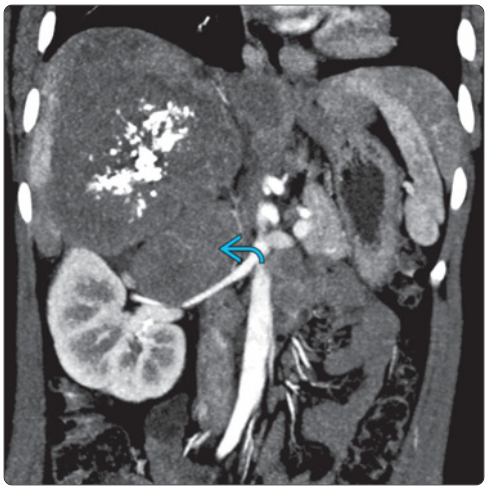


Stage IV (T3 N1 M0)

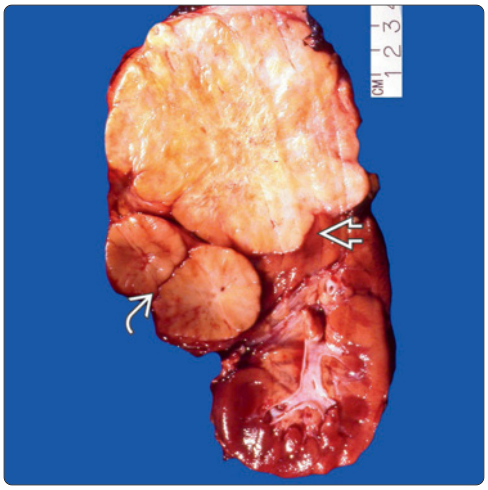


(Left) *Scout image from CT demonstrates calcifications in a right upper quadrant mass. Hepatic flexure is inferiorly displaced by the inferior displacement of the right kidney.* **(Right)** *Axial arterial phase CECT demonstrates the very large, well-circumscribed right adrenal mass with central calcifications. An enlarged paraaortic lymph node is consistent with N1 disease, upstaging this patient's disease to stage IV.*

Stage IV (T3 N1 M0)

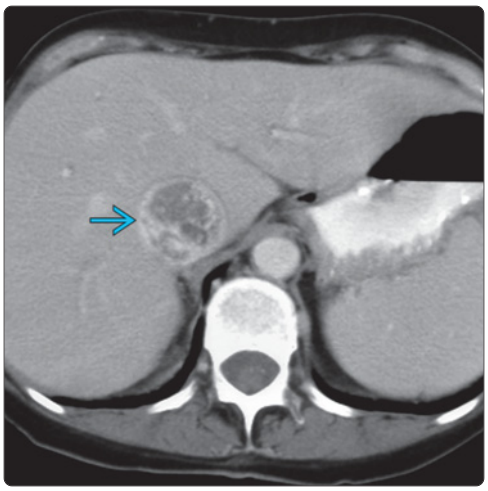


Stage IV (T3 N1 M0)

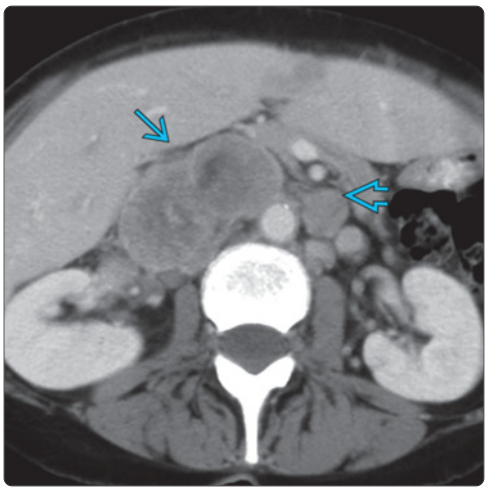


(Left) *Coronal MIP image in the same patient demonstrates the inferior displacement of the right kidney. Intact planes are present between the mass and adjacent organs, but large local nodal metastases are present.* **(Right)** *Gross image demonstrates the clean plane between the mass and adjacent kidney, as well as the metastatic lymph nodes.*

Stage IV (T4 N1 M0)

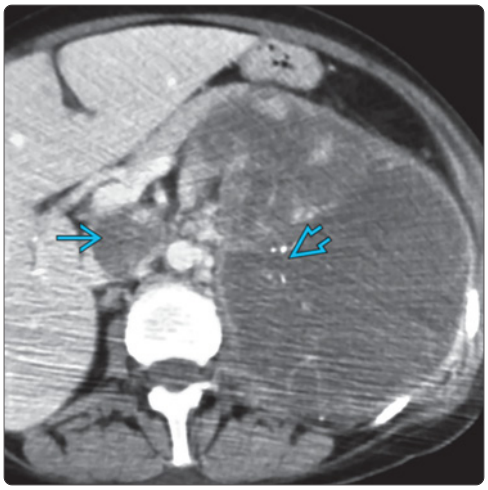


Stage IV (T4 N1 M0)

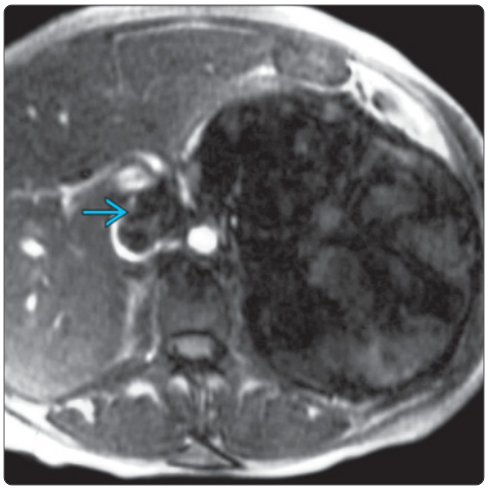


(Left) *Axial CECT demonstrates heterogeneous tumor thrombus in the hepatic inferior vena cava.* **(Right)** *More caudal axial CECT in the same patient demonstrates the markedly enlarged and abnormal paraaortic lymph nodes, both ipsilateral and contralateral to the primary tumor.*

Stage IV (T4 N1 M0)

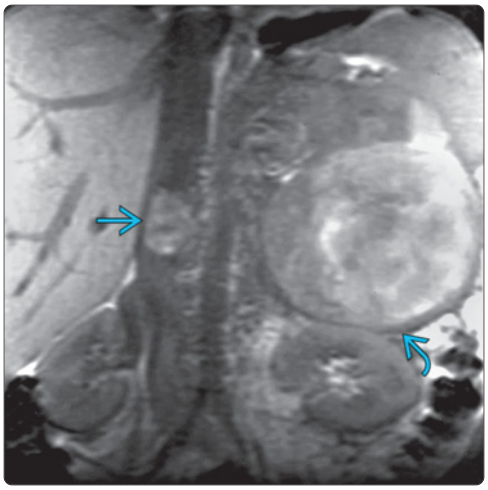


Stage IV (T4 N1 M0)

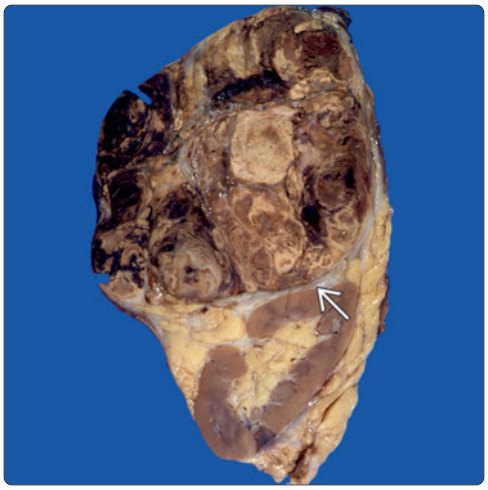


(Left) *This axial CECT demonstrates a large tumor in the left pararenal space. The center of the mass is low attenuation, with calcifications. Tumor thrombus in the infrahepatic inferior vena cava makes this a T4 lesion.* **(Right)** *Axial T1WI C+ MR in the same patient demonstrates the large, heterogeneously low T1 SI mass in the left perirenal space, with IVC thrombus of the same low signal intensity.*

Stage IV (T4 N1 M0)

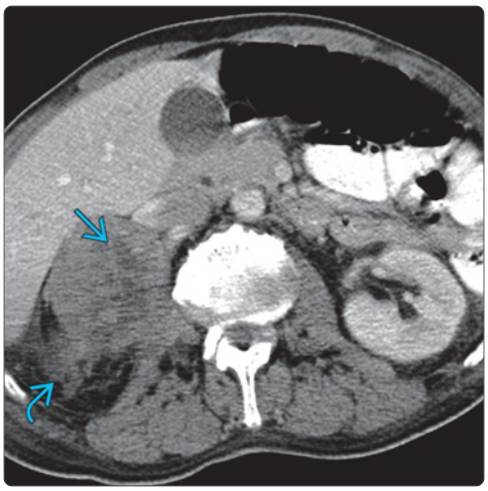


Stage IV (T4 N1 M0)

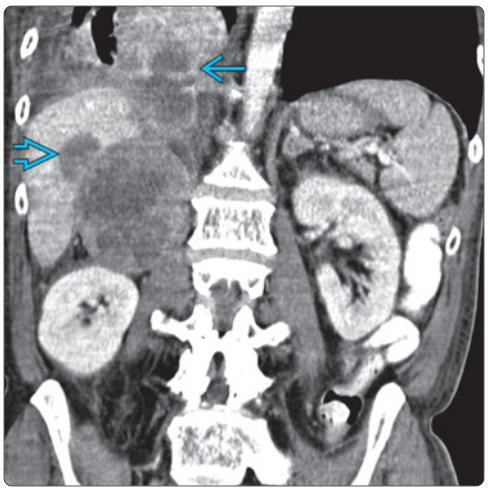


(Left) *Coronal T1WI MR in the same patient shows, more convincingly, the tumor thrombus in the IVC. The left kidney is displaced inferiorly; however, there appears to be an intact plane between the mass and the kidney.* **(Right)** *Gross image shows the heterogeneously solid and necrotic left adrenal carcinoma. A fibrous capsule separates the mass from the adjacent kidney, corresponding to the findings on MR. The IVC invasion was confirmed at resection.*

Stage IV (T4 N1 M1)

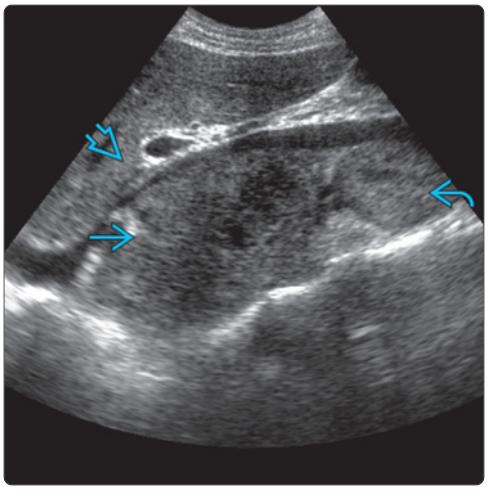


Stage IV (T4 N1 M1)

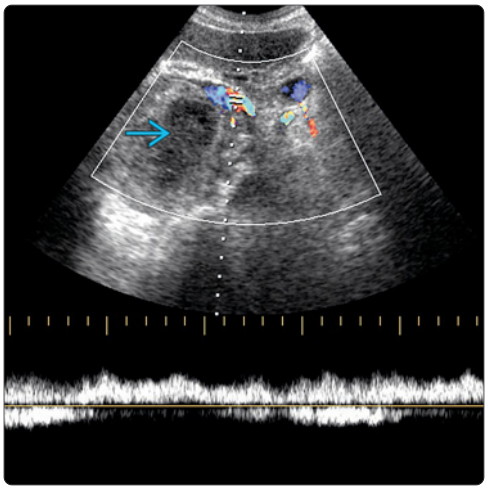


(Left) *Axial CECT reveals a large, heterogeneous right adrenal mass with infiltration of the surrounding fat well demonstrated on the CT.* **(Right)** *Coronal image shows both the hepatic metastasis that may have arisen from direct tumor invasion and invasion of the tumor through the diaphragm to involve the right lung base. This constitutes unresectable disease.*

Stage III (T3 NX MX)

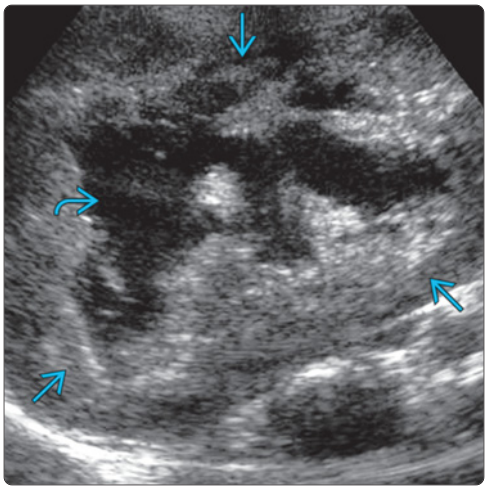


Stage III (T3 NX MX)

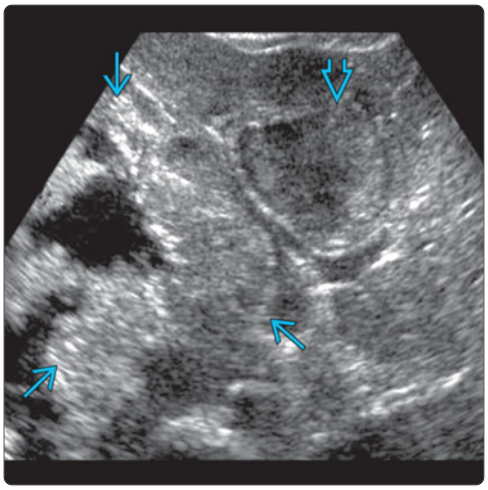


(Left) *Grayscale transabdominal ultrasound shows a > 5 cm adrenal carcinoma. The disruption of the adrenal capsule (local invasion/T3 disease) is not well demonstrated on US. There is no clear invasion of adjacent organs, including the inferiorly displaced right kidney and liver, that would upstage the tumor to T4 disease.* **(Right)** *On color Doppler, the carcinoma shows an internal hypoechoic avascular component consistent with necrosis.*

Stage IV (T4 NX M1)

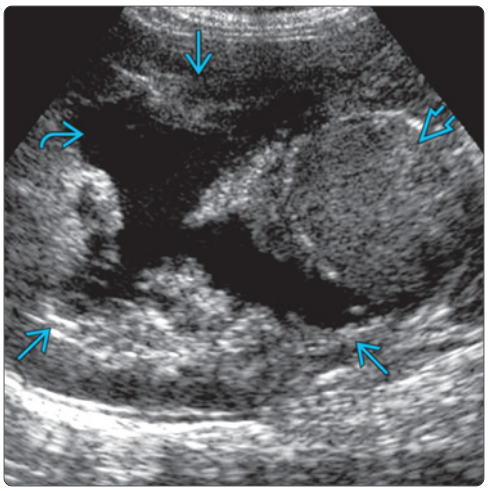


Stage IV (T4 NX M1)

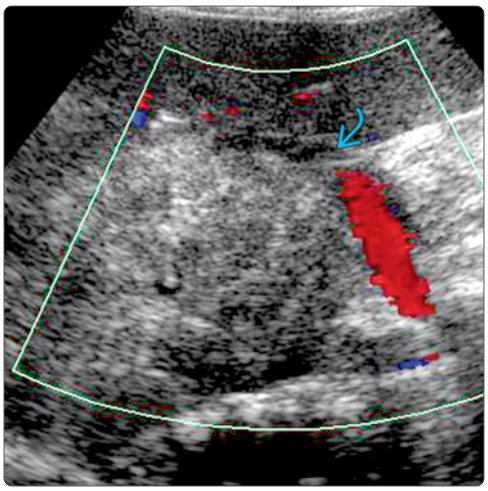


(Left) *Longitudinal transabdominal grayscale ultrasound shows a large adrenal carcinoma with central necrosis mimicking an abscess.* **(Right)** *Transverse transabdominal ultrasound in the same patient demonstrates both the large, centrally necrotic adrenal carcinoma and a well-defined hepatic metastasis adjacent to the carcinoma.*

Stage IV (T4 NX M1)



Stage IV (T4 NX M1)



(Left) *Oblique transabdominal ultrasound in the same patient shows the large, centrally necrotic adrenal carcinoma. A discrete tissue plane between the primary carcinoma and the adjacent hepatic metastasis is difficult to appreciate.* **(Right)** *Longitudinal color Doppler ultrasound in the same patient reveals the extrinsic compression of the inferior vena cava.*

Adrenal Lymphoma

KEY FACTS

IMAGING

- CT: Mild enhancement of bilateral adrenal masses
- MR: T2 intense, T1 isointense (to muscle), bulky adrenal masses
- Ultrasound: Hypoechoic suprarenal lesions
- PET/CT: Hypermetabolic adrenal tumor

TOP DIFFERENTIAL DIAGNOSES

- Adrenal metastases
  - Heterogeneous adrenal masses
  - Clinical history, identification of primary lesion aids diagnosis
  - Adrenal insufficiency rare despite size
- Adrenal hemorrhage
  - High attenuation at NECT in acute setting
  - Predisposing history: Sepsis, anticoagulation
- Adrenal adenoma
  - Typically incidental finding
  - Attenuation < 10 HU at NECT, contrast kinetics (rapid washout)
- Adrenal tuberculosis and fungal infection
  - May also present with adrenal insufficiency
  - Necrotic adrenal masses in acute setting
  - Chronic infection: Adrenal calcification

PATHOLOGY

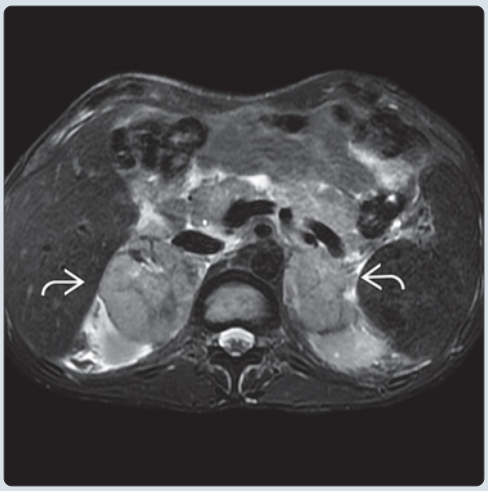
- Majority are diffuse large B-cell lymphoma
- Little or no extraadrenal visceral, nodal involvement

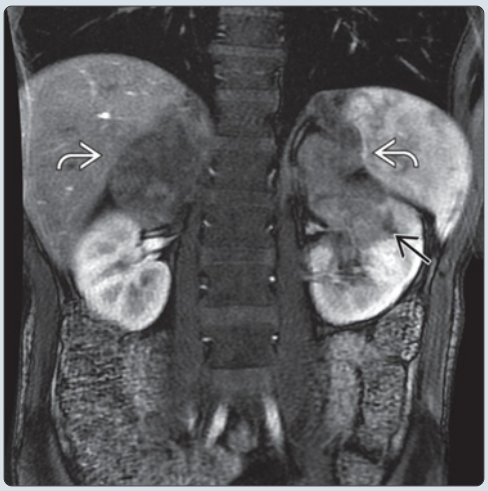
CLINICAL ISSUES

- B-cell symptoms, adrenal insufficiency
- Poor prognosis

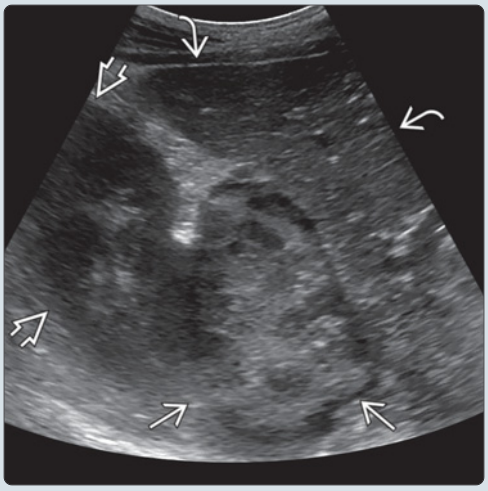
DIAGNOSTIC CHECKLIST

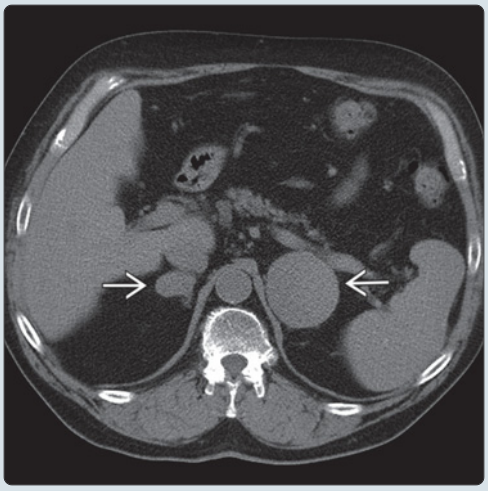
- Consider if large bilateral adrenal masses, in elderly male with adrenal insufficiency, and no known primary malignancy





(Left) *Axial T2 TSE performed on a 36-year-old male with malaise and adrenal insufficiency shows bilateral bulky T2-intense adrenal masses. (Right) Coronal GD-enhanced T1 C+ MR in the same patient shows both adrenal masses mildly enhanced. Note the direct extension of tumor into the upper pole of the left kidney. The differential diagnosis includes adrenal metastases; although, adrenal insufficiency, and a lack of a history of an extrarenal primary suggest an alternative diagnosis.*





(Left) *Ultrasound performed prior to biopsy on the same patient shows a large mixed echogenicity mass adjacent to the kidney and liver. The biopsy revealed B-cell lymphoma. Complete remission was achieved after CHOP and rituximab.* **(Right)** *NECT performed on an elderly male with fatigue and adrenal insufficiency shows large adrenal masses. No additional nodal or visceral disease was identified. A diagnosis of primary large B-cell lymphoma was confirmed after ultrasound-guided biopsy.*

TERMINOLOGY

Definitions

- Histologically confirmed lymphoma involving 1 or both adrenal glands
  - No prior history of lymphoma
  - If other visceral, nodal involvement, adrenal disease is dominant

IMAGING

General Features

- Best diagnostic clue
  - Isolated adrenal masses in setting of adrenal insufficiency
- Location
  - Both adrenal glands typically involved, though unilateral cases reported
- Size
  - Large soft tissue masses, often > 6 cm

CT Findings

- Mild enhancement of uniform, solid attenuation adrenal masses
- Isolated adrenal lesions or if other visceral/nodal disease, adrenal involvement clearly dominant
  - Distinguish from secondary involvement of adrenals in setting of widespread diffuse, extranodal lymphoma

MR Findings

- Iso-/hypointense T1, hyperintense T2 adrenal masses
- Uniform mild enhancement (similar to CECT)

Ultrasonographic Findings

- Bilateral hypoechoic adrenal masses

Nuclear Medicine Findings

- Hypermetabolic at FDG PET/CT

Imaging Recommendations

- Best imaging tool
  - Dedicated adrenal CT: Localization of tumor within adrenal glands, and excludes other visceral, nodal involvement

DIFFERENTIAL DIAGNOSIS

Adrenal Metastases

- Clinical history, identification of primary aids diagnosis
- Adrenal insufficiency rarely reported

Adrenal Hemorrhage

- Predisposing history (sepsis, anticoagulation)
- Acute hemorrhage: ↑ attenuation at NECT, perinephric space stranding

Adrenal Adenoma

- Attenuation at NECT: < 10 HU
- Assess washout kinetics at delayed CT

Adrenal Tuberculosis and Fungal Infection

- May also present with adrenal insufficiency
- Necrotic adrenal masses in acute setting
- Adrenal calcification with chronic infection

PATHOLOGY

General Features

- Vast majority: Diffuse large B-cell lymphoma
- Rare entity: Biology, pathophysiology poorly understood
- Postulated mechanisms for PAL
  - Preexisting autoimmune adrenalitis (controversial)
    - Analogous to primary thyroid lymphoma in setting of chronic autoimmune Hashimoto thyroiditis
    - May help explain high incidence of adrenal insufficiency with PAL
  - Hematopoietic tissue rests within adrenal glands

Staging, Grading, & Classification

- WHO classification based upon percutaneous biopsy, adrenalectomy
- Classified as stage 1E (primary extranodal lymphoma) in Ann Arbor classification
  - Should be differentiated from stage 4 (disseminated extranodal dz), which has worse prognosis

Gross Pathologic & Surgical Features

- Large soft tissue adrenal mass, may infiltrate into ipsilateral kidney

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - B-cell symptoms (fever, night sweats, fatigue, weight loss)
  - Adrenal insufficiency (fatigue, anorexia, hyperpigmentation)

Demographics

- Elderly males more commonly affected

Natural History & Prognosis

- Poor prognosis: Largest PAL study (n = 31), after 6 chemotherapy cycles
  - 2-year survival, progression-free survival rate: 68%/51%
  - Complete remission, partial remission: 55%/32%

Treatment

- First-line therapy: CHOP
- Rituximab also employed in first-line or for salvage

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Large bilateral adrenal masses in patient with adrenal insufficiency and no extraadrenal primary malignancy

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Adrenal Metastases

KEY FACTS

IMAGING

- Likelihood of adrenal metastasis at imaging depends upon clinical history
  - Increased likelihood of adrenal metastasis if history of extrarenal malignancy
  - Imaging needed to differentiate between isolated benign adrenal lesion (typically adenoma) and metastasis during initial staging and surveillance
    - NECT attenuation < 10 HU or signal loss at out-phase GRE MR: Lipid-rich adenoma
    - Rapid washout at CECT (> 60% at 15 minutes): Likely lipid-poor adenoma
      - Caveat: Rapid washout observed with hypervascular adrenal metastases (HCC, RCC)
    - FDG PET/CT: Uptake in adrenal mass significantly greater than liver suggests metastasis
  - Comparison to prior studies critical: New adrenal mass in patient with known (active) extraadrenal malignancy indicates metastasis
  - Biopsy rarely needed for diagnosis but may be performed for equivocal lesions, particularly if surgery or ablation considered for potential isolated adrenal metastasis
- Incidental adrenal lesions ("incidentaloma") are rarely metastases

TOP DIFFERENTIAL DIAGNOSES

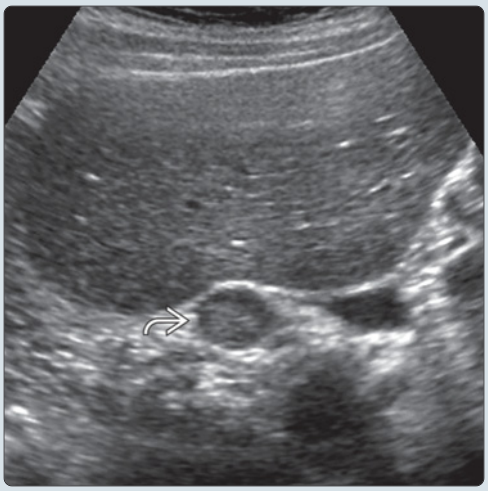
- Adrenal adenoma
- Adrenal carcinoma
- Adrenal hemorrhage

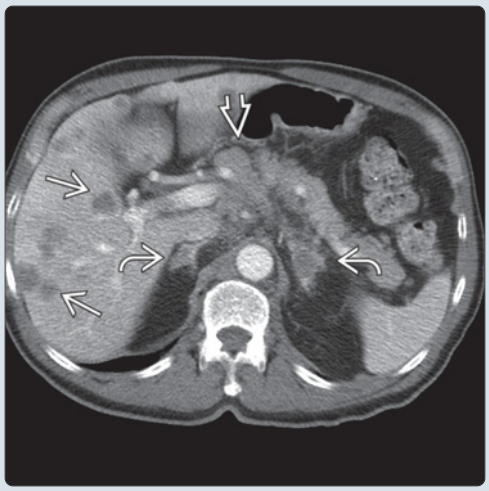
CLINICAL ISSUES

- Common target organ for metastases from lung, breast, melanoma
- Typically implies late-stage disease
- Adrenal insufficiency if large, bilateral metastases

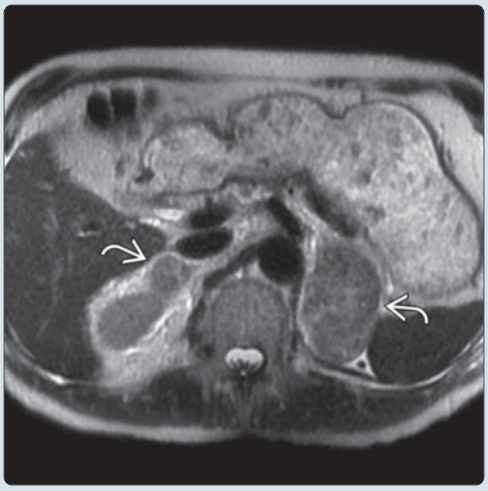
DIAGNOSTIC CHECKLIST

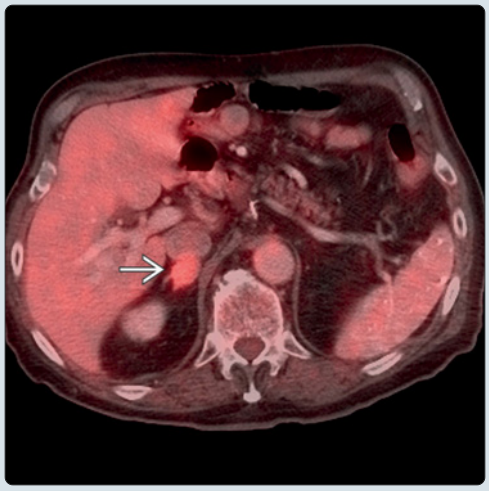
- Clinical setting usually indicates correct diagnosis





(Left) *Axial ultrasound of a patient with lung carcinoma shows a solid right adrenal lesion. The appearance is nonspecific, but the history suggests adrenal metastasis. Biopsy performed because of equivocal findings at PET/CT confirmed metastasis.* **(Right)** *Axial CECT shows hepatic metastases, abdominal adenopathy, and adrenal lesions in a patient with esophageal carcinoma. The imaging appearance of the adrenal lesions is nonspecific, but clinical history and widespread disease confirm adrenal metastases.*





(Left) *Axial T2WI MR of an elderly male with a history of non-small cell lung carcinoma shows large bilateral adrenal masses. Rapid growth from a prior staging CT (not shown) and the clinical history indicate adrenal metastases.* **(Right)** *Axial PET/CT shows an isolated FDG-avid right adrenal metastasis in a patient with a history of T2, N0 esophageal carcinoma, post esophagectomy. An adrenalectomy was performed given the initial stage, and absence of other metastases.*

IMAGING

General Features

- Best diagnostic clue
  - Discrete or diffuse soft tissue attenuation suprarenal masses in patient with history of extraadrenal malignancy
- Key concepts
  - 4th most common site of hematogenous metastases after lungs, liver, and bone
    - Most common primary sites
      - Malignant melanoma: 50% metastasize to adrenal glands
      - Lung and breast cancers: 30-40% metastasize to adrenal glands
      - Renal and GI tract malignancies: 10-20% metastasize to adrenal glands
    - Indicates stage IV distant metastatic disease
    - Mostly discrete intraparenchymal masses
      - Unilateral or bilateral; small or large
    - Direct extension into adrenals from tumors in adjacent organs
      - Kidney, pancreas, stomach, liver, retroperitoneum
  - True prevalence of secondary adrenal metastases difficult to define
    - Up to 30% of adrenal lesions may be metastatic in oncologic population
    - Incidental adrenal lesions extremely unlikely to be metastatic in patients with no history of an extraadrenal malignancy

CT Findings

- Small metastases
  - Well-defined, round or oval in shape
  - Adrenal gland contour may be maintained
  - Homogeneous soft tissue attenuation masses
  - Unilateral or bilateral
  - Necrosis, hemorrhage, and calcification are rare
  - May mimic lipid-poor adrenal adenoma on NECT
- Large metastases
  - Unilateral or bilateral adrenal masses
  - Heterogeneous density (necrosis, hemorrhage)
  - Distortion of normal contour of adrenal gland
  - Lobulated or irregular in shape
  - ± central necrosis, hemorrhage, calcification
  - May have thick enhancing rims
  - Hypervascular or hypovascular
  - Uncommon invasion of contiguous organs
  - Prolonged washout pattern on CECT
    - Absolute washout value of < 60% or relative washout value of < 40% after 15 minutes suggests metastasis or other "nonadenoma"
    - Beware: Hypervascular adrenal metastases (HCC, RCC) may have rapid washout pattern similar to lipid-poor adenomas

MR Findings

- Without necrosis and hemorrhage
  - T1WI: Usually homogeneous and hypointense
  - T2WI: Relatively hyperintense
  - No signal loss on opposed-phase GRE (unlike lipid-rich adenoma)
- With necrosis and hemorrhage
  - ↑ signal at T1WI suggests blood products
  - ↑ signal at T2WI due to necrosis: May mimic pheochromocytoma
  - ↑ signal at T1WI out-phase, and T1WI fat-saturation sequences excludes adenoma and myelolipoma
- No role for diffusion weighted imaging: ADC overlap between benign and malignant adrenal masses
- MR spectroscopy advocated, though technique difficult to apply to small adrenal lesions and not widely accessible

Ultrasonographic Findings

- Grayscale ultrasound
  - Nonspecific appearance: Solid, relatively hypoechoic suprarenal mass(es)

Angiographic Findings

- Hypervascular: Renal cell carcinoma or sarcoma metastases
- Hypovascular: Squamous cell cancer metastases

Nonvascular Interventions

- Image-guided biopsy for definite proof
  - Usually unnecessary; imaging findings and clinical setting suffice

Nuclear Medicine Findings

- FDG PET
  - Hypermetabolic adrenal mass: Activity greater than liver
  - FDG PET meta analysis: Pooled sensitivity and specificity of 97% and 91% for distinguishing between benign and malignant adrenal masses
  - FDG PET/CT
    - Improved spacial, anatomic localization
    - Combination of PET analysis (SUV ratio of adrenal/liver) and NECT attenuation measurement improves accuracy
      - False-negative: Small (< 8 mm) metastases, neoplasms with low FDG avidity (neuroendocrine tumor, bronchoalveolar cancer), tumor necrosis or hemorrhage
      - False-positive: Adrenal adenoma, pheochromocytoma, inflammatory/granulomatous disease

Imaging Recommendations

- Best imaging tool
  - Helical NE + CECT usually suffice
    - Typically identified during initial staging
  - PET/CT for initial staging and evaluation of treatment response
- Protocol advice
  - CT: < 3 mm thick section at 3 mm intervals or less
  - MR: T1WI GRE in- and out-phase to exclude adenoma

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- NECT
  - Lipid-rich adenoma: Low attenuation (< 10 HU)
    - Characteristic and diagnostic of adenoma
  - Lipid-poor adenoma: Attenuation varies (10-30 HU)

    - Simulate metastases on NECT
  - Large adenoma: Heterogeneous density
    - Due to hemorrhage, cystic degeneration, Ca++
- CECT
  - Lipid-rich and -poor adenomas
    - Absolute washout of adenoma: 15 minutes post IV > 60%
    - Washout pattern: Typically diagnostic for adenoma, though overlap with hypervascular metastases (HCC, RCC)
- T1WI GRE out-phase
  - Lipid-rich adenoma: Marked signal "dropout"
  - Lipid-poor adenoma: Minimal signal loss

Adrenal Carcinoma

- Rare, unilateral, invasive, and enhancing mass
- > 6 cm when initially diagnosed

Adrenal Hemorrhage

- Acute: High-density fluid collection (40-60 HU)
- Chronic: Low-density collection (20-30 HU)
  - May mimic metastases or lymphoma
- Size and density evolve rapidly, unlike tumors

Pheochromocytoma

- Highly vascular; prone to hemorrhage and necrosis
- Very hyperintense on T2WI
- Bilateral adrenal tumors in MEN 2A and 2B syndromes, or von Hippel-Lindau

Adrenal Myelolipoma

- Tumor composed of fat and hematopoietic elements
- Usually heterogeneous, fatty adrenal masses
- Tumor with ↑ fat content
  - Fat-suppression sequence: Hypointense
  - Marked signal "dropout" at fat/soft tissue interfaces
- Rarely bilateral

PATHOLOGY

General Features

- Associated abnormalities
  - Extraadrenal primary malignancy usually known when adrenal metastasis is imaged
  - Adrenal collision tumor
    - Metastases and adenoma in same adrenal gland
- Normal anatomy of adrenal glands
  - Located between vertebrae T11 and L2, lateral to body of L1
  - Gland has anteromedial ridge and 2 limbs
  - Maximum body width: 0.79 mm (right), 0.6 mm (left)
  - Length of adrenal limbs varies: May be up to 4 cm
  - Width of limbs: < 1 cm (right thinner than left)

Gross Pathologic & Surgical Features

- Discrete or diffuse; unilateral or bilateral
- ± cystic, necrotic, hemorrhagic, calcific areas

Microscopic Features

- Depends upon primary lesion

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Almost always clinically silent
  - Large masses: Adrenocortical insufficiency (Addison disease) when 90% of tissue is damaged
  - Addison disease: Weakness, weight loss, anorexia, nausea, vomiting, hypotension, skin pigmentation
- Clinical profile
  - Patient with history of malignancy elsewhere and manifestations of Addison disease
- Lab data
  - Adrenocortical insufficiency
    - ↓ cortisol, aldosterone, androgens; ↑ ACTH
    - ↓ Na+ and Cl-; ↑ K+ levels
- Diagnosis: Imaging and rarely percutaneous needle biopsy

Demographics

- Age
  - Any age group; more common in elderly
- Gender
  - M = F

Natural History & Prognosis

- Complications: Adrenocortical insufficiency
- Prognosis: Usually poor

Treatment

- Chemotherapy and follow-up
- Resection or ablation may be considered in selected patients with isolated adrenal metastases

DIAGNOSTIC CHECKLIST

Consider

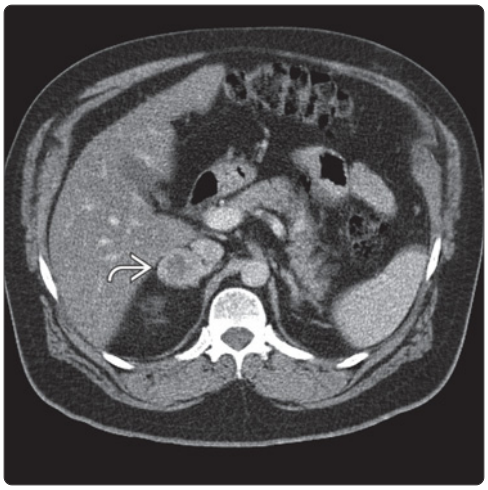
- Check for history of primary cancer

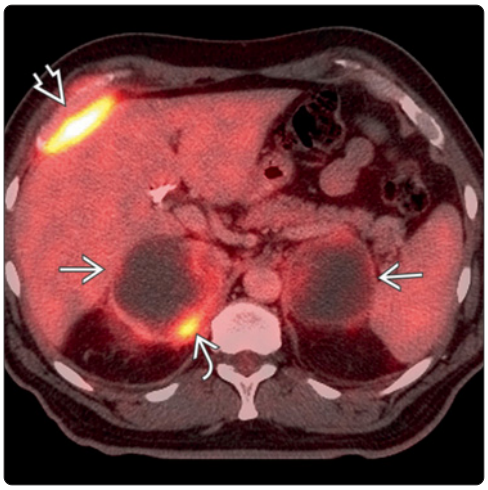
Image Interpretation Pearls

- Clinical setting usually indicates correct diagnosis
- Comparison to prior studies paramount: New adrenal lesion in oncologic patient almost universally indicates metastasis
- Imaging important to suggest and stage malignancy
- PET/CT has supplanted most other imaging and biopsy in patients with adrenal mass(es) and known cancer

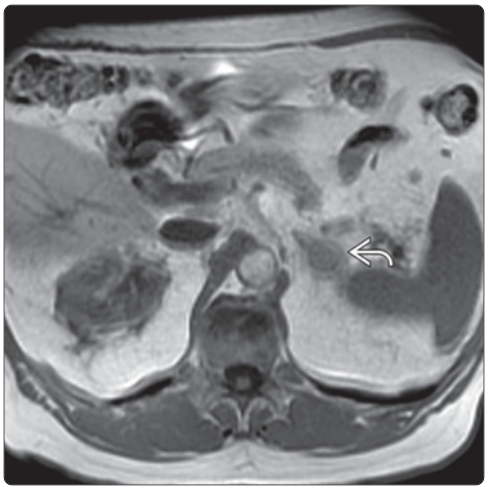
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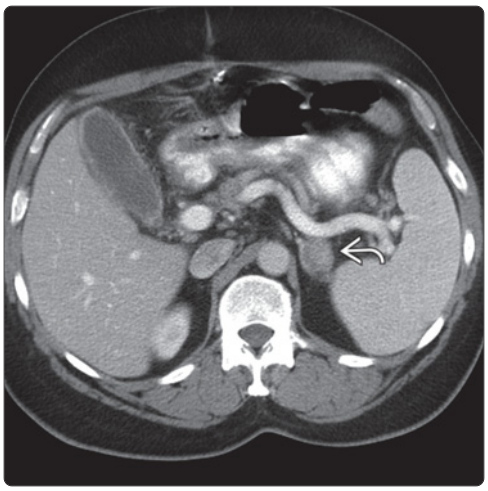


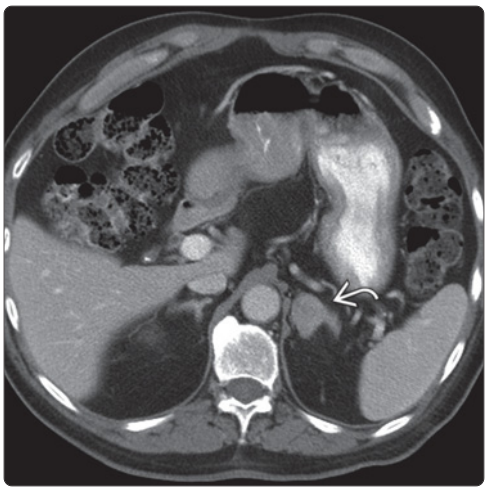
(Left) *Axial CECT of a patient with a history of RCCs post nephrectomy shows a new hypervascular right adrenal mass. The lesion was not FDG avid and rapid washout was shown on a delayed phase. Despite these results, growth and patient history indicated a metastasis (confirmed at biopsy and adrenalectomy).* **(Right)** *FDG PET/CT of a patient with lung cancer shows huge necrotic adrenal metastases. Note scant residual uptake within the periphery of the right lesion & an FDG-avid right chest wall metastasis.*





(Left) *Axial SPGR in-phase MR of a patient with a history of multifocal HCC post liver resection, TACE, and right adrenal metastasectomy shows a small left adrenal mass.* **(Right)** *Axial SPGR out of phase MR of the same patient shows no significant loss of signal from the adrenal lesion. MR imaging alone might still suggest a lipid-poor adenoma, but the history and growth over time indicate a metastasis.*





(Left) *Axial CECT of a patient with colon carcinoma shows a smooth, uniform left adrenal metastasis. Taken out of clinical context, an adenoma is statistically most likely, but the history, liver metastases (not shown), and growth indicate a metastasis.* **(Right)** *Axial CECT of a patient with a history of lung cancer shows a new, isolated, adrenal mass. Although a relative washout of 42% might suggest a lipid-poor adenoma, an adrenalectomy, performed given the absence of disease elsewhere, confirmed a metastasis.*

Adrenal Collision Tumor

KEY FACTS

TERMINOLOGY

- Refers to 2 adjacent but histologically different tumors within same adrenal gland (no histologic admixture at interface)
  - Not a composition tumor (i.e., tumor containing admixture of 2 different cell types)
  - Adenoma and myelolipoma reported to be most common adrenal collision tumor, though adenomas may contain small foci of macroscopic fat

IMAGING

- Both tumors may be malignant, both may be benign, or 1 malignant and 1 benign
- FDG-avid mass suggests malignant tumor
- CT to recognize adrenal mass
- PET/CT to characterize mass
  - Most practical noninvasive means of making diagnosis
  - Histological verification by percutaneous biopsy with imaging guidance

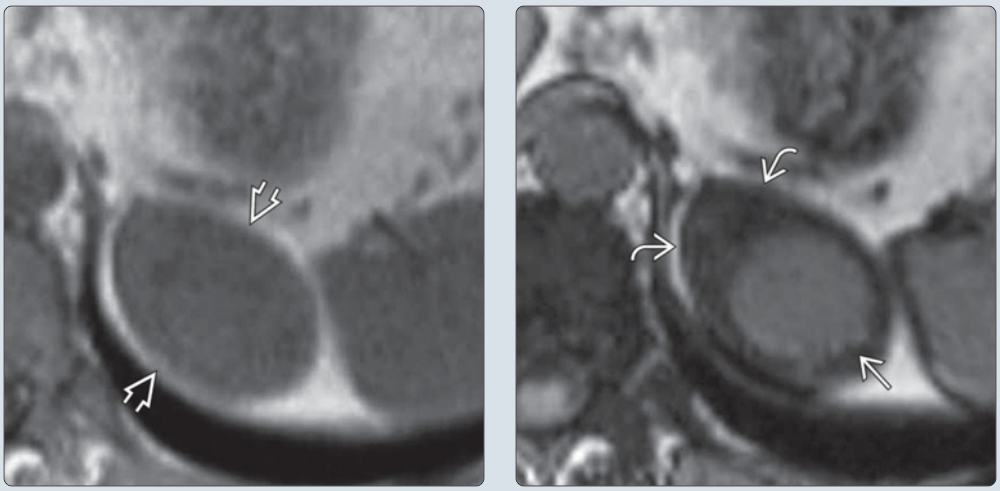
TOP DIFFERENTIAL DIAGNOSES

- Adrenal adenoma
- Adrenal carcinoma
- Adrenal metastases and lymphoma
- Adrenal myelolipoma
- Adrenal hemorrhage

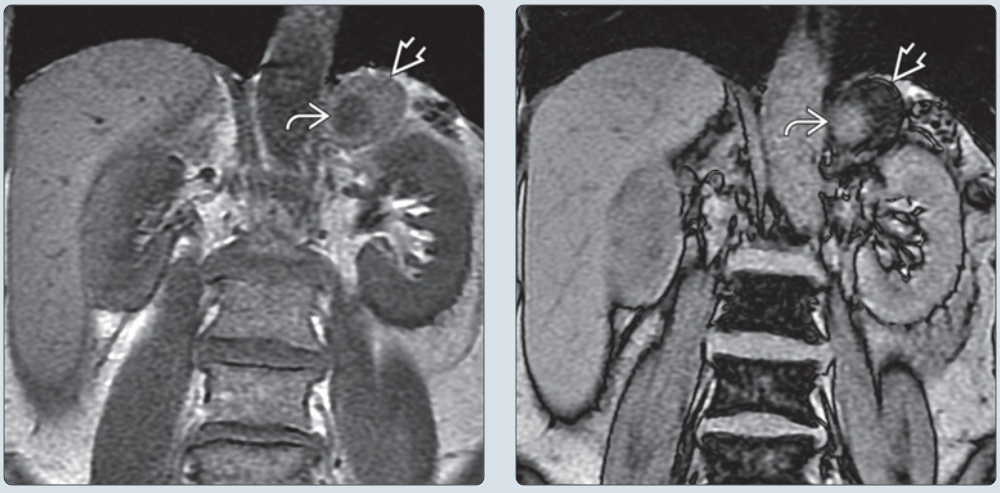
DIAGNOSTIC CHECKLIST

- Adenomas may contain foci of fat and be indistinguishable from adenoma/myelolipoma collision tumor
- Metastasis and adenoma collision tumor is most clinically relevant
- 2 distinct masses with differing attenuation or signal attenuation
- PET/CT useful for diagnosis of adenoma-metastasis collision tumor

(Left) *Axial in-phase T1WI MR shows an indeterminate 4 cm left adrenal lesion. (Right) Corresponding out of phase MR shows that the adrenal mass is actually composed of 2 distinct lesions. Signal suppression of the peripheral lesion indicates a lipid-rich adenoma. The higher signal intensity central lesion is a metastasis. Collision tumors such as this may be due simply to chance, given the high prevalence of adenomas and metastases in the oncologic patient population. (Courtesy E. Caoili, MD.)*



(Left) *Coronal T1WI in-phase MR performed to evaluate an adrenal lesion in an elderly woman with pulmonary fibrosis shows a 3 cm left adrenal mass. Note the well-circumscribed, ↓ signal central component. (Right) Out of phase image again shows peripheral signal suppression (indicating a lipid-rich adenoma) and a separate, indeterminate lesion. Growth of the mass prompted resection. A collision tumor composed of an adenoma and a schwannoma (a rare benign adrenal lesion) was shown at pathology.*



TERMINOLOGY

Abbreviations

- Adrenal collision tumor (ACT)

Definitions

- Coexistence of 2 contiguous but histologically different tumors within same adrenal gland
- Not a composition tumor (i.e., tumor with intermixed, differing cell types)

IMAGING

General Features

- Best diagnostic clue
  - Adrenal gland containing 2 distinct, separate tumors
  - PET/CT is most practical noninvasive means of making diagnosis
  - Histological verification by percutaneous biopsy with imaging guidance
- Unilateral: Both tumors may be malignant, both may be benign, or 1 malignant and 1 benign

CT Findings

- 2 lesions with differing attenuation
  - Macroscopic fat: Myelolipoma
  - NECT attenuation < 10 HU: Lipid-rich adenoma (or cyst)
  - NECT attenuation > 10 HU: Lipid-poor adenoma, metastasis, pheochromocytoma, etc.
    - CECT with washout kinetics may help differentiate
  - Myelolipoma and adenoma reported as most common ACT, although adenomas may have small macroscopic fat components

MR Findings

- 2 lesions with differing signal
- Chemical shift MR (in phase and out of phase imaging) for identification of lipid-rich adenoma

Nuclear Medicine Findings

- PET/CT
  - FDG-avid mass suggests coexistent malignant tumor

Imaging Recommendations

- Best imaging tool
  - CT to recognize adrenal ACT
    - PET/CT to characterize mass

DIFFERENTIAL DIAGNOSIS

Adrenal Adenoma

- Signal dropout on opposed-phase GRE; < 10 HU on NECT
- Rapid washout pattern of contrast material on CECT

Adrenal Carcinoma

- Large (usually ≥ 5 cm); central necrosis and hemorrhage
- Lung metastases, lymphadenopathy, venous invasion

Adrenal Metastases and Lymphoma

- Metastases: Unilateral or bilateral; known malignancy
  - Hyperintense on T2WI; delayed contrast washout
  - > 20 HU and heterogeneous on NECT
- Lymphoma: Usually bilateral, hypovascular

Adrenal Myelolipoma

- Macroscopic fat

Adrenal Hemorrhage

- Usually in setting of trauma or stress

PATHOLOGY

General Features

- Associated abnormalities
  - Unusual variant of multiple endocrine neoplasia syndrome; ACT as component

Gross Pathologic & Surgical Features

- Enlarged adrenal containing 2 distinct masses
- Laparoscopic resection reported for imaging indeterminate, growing collision tumors

Microscopic Features

- 2 distinct tumors abutting or within close proximity to each other
- Not a composite tumor (i.e., 2 different histologic types intermixed within same tumor)

CLINICAL ISSUES

Demographics

- Epidemiology
  - Extremely rare; prevalence unknown
  - May be due to chance (high prevalence of adrenal adenomas and adrenal metastases) although carcinogenic stimulus providing fertile ground for tumors also proposed

Natural History & Prognosis

- Variable, depending on histology of each component

DIAGNOSTIC CHECKLIST

Consider

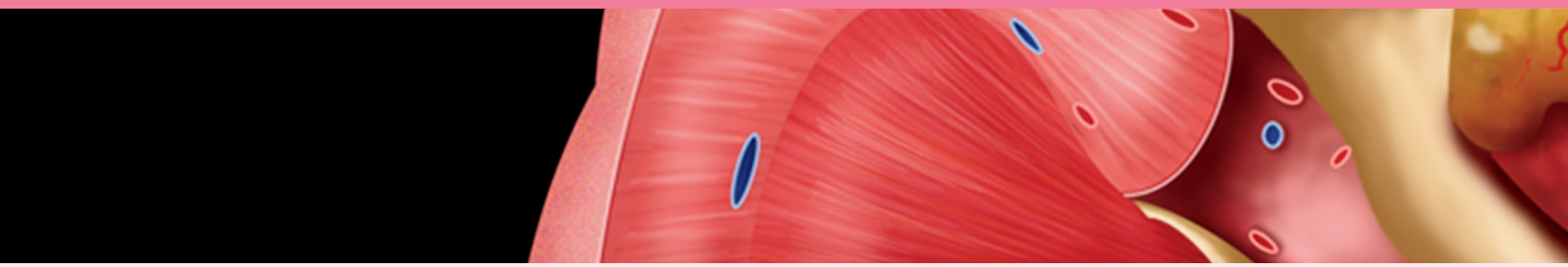
- Adenomas may contain foci of fat and be indistinguishable from adenoma/myelolipoma collision tumor
- Metastasis and coexistent adenoma is most clinically important scenario
- Clinical history (known primary), and coexistent adrenal lesions with differing attenuation or signal attenuation should raise index of suspicion
- PET for initial evaluation; directed percutaneous biopsy of nonadenoma component for problematic cases

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SECTION 4

Kidney and Renal Pelvis



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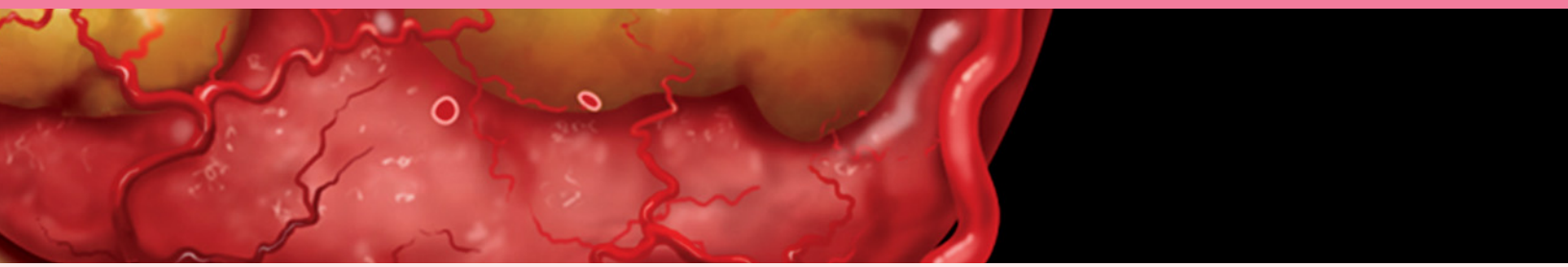
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Principles of Renal Physiology

The main functions of the kidneys are the maintenance of homeostasis, removal of metabolic waste products, and production of urine. The nephron is the functional unit of the kidney. The functions are carried out with a combination of glomerular filtration, tubular reabsorption, and tubular secretion. Filtration is conducted in the renal corpuscle (glomerulus, Bowman capsule) in the cortex, while reabsorption and secretion occur in the different tubular components (proximal, distal convoluted tubule, loop of Henle, collecting ducts) located in the cortex and medulla.

Glomerular filtration rate (GFR): Volume of fluid filtered in the glomerulus per unit of time. It is determined by the net filtration rate, capillary permeability, and surface area of the capillary bed. The normal GFR in adult is 125 mL/minute.

- Renal function according to eGFR is categorized as stage 1 (eGFR > 90 mg/mL/1.73m²), normal renal function; stage 2 (eGFR 60-89 mg/mL/1.73m²), mildly reduced renal function; stage 3 (eGFR 30-69 mg/mL/1.73m²), moderately reduced renal function; stage 4 (eGFR 15-29 mg/mL/1.73m²), severely reduced renal function; and stage 5 (eGFR < 15 mg/mL/1.73m²), end-stage renal failure

Iodinated Contrast Media

Iodinated contrast media (ICM) are hydrophilic chemical compounds excreted from the kidneys by glomerular filtration alone. Enhancement (i.e., increased attenuation) after injection of ICM is based on several agent-related (e.g., concentration, volume, rate of injection), patient-related (weight, height, cardiac output), and technical-related (scan delay for CT acquisition) factors. ICM are described based on their dissociation in solution (ionic, nonionic), osmolarity compared to plasma (high-, iso-, low-osmolar), and number of triiodinated benzene groups (monomer, dimer).

Types of contrast media

- Ionic: High-osmolar contrast media (HOCM); high osmolarity (1,550-2100 mOsm/kg) (diatrizoate, metrizoate)
- Nonionic monomers: Low-osmolar contrast media (LOCM); lower osmolarity (500-880 mOsm/kg) (iopamidol, iohexol, ioxilan, iopromide)
- Nonionic dimers: Iso-osmolar contrast media (IOCM); iso-osmolar to serum (290 mOsm/kg) (Iodixanol)

Contrast-induced nephropathy (CIN): Acute kidney injury (absolute increase in serum creatinine of 0.5 mg/dL or relative 25% increase from baseline value) occurring within 24-48 hours following intravascular administration of contrast material. The most important risk factor for CIN is underlying renal dysfunction (GFR < 60 mL/min/1.73m²). The American College of Radiology recommends obtaining a baseline creatinine in all patients with suspected renal dysfunction. Other risk factors include decreased renal perfusion (e.g., heart failure), age > 70 years, diabetes mellitus, concomitant use of nephrotoxic drugs (e.g., NSAIDs), multiple myeloma, large volume of contrast media, and high-osmolar contrast media. In patients at risk for CIN but still requiring injection of contrast media, the following prophylactic measures should be taken: Intravenous hydration, lower dose of hypo- or iso-osmolar contrast media, and no nephrotoxic drugs.

MR Contrast Media

Gadolinium-based MR contrast agents (GBCA) are paramagnetic compounds that increase signal intensity (i.e., enhancement) by increasing the T1 relaxation rate. The evaluation of the genitourinary tract is conducted using extracellular agents, preferably with high relaxivity.

Approved extracellular GBCAs include gadoterate meglumine, gadobutrol, gadopentate dimeglumine, gadodiamide, gadoversetamide, gadoteridol, and gadobenate dimeglumine. Extracellular GBCAs are 100% excreted by the kidneys, with the exception of gadobenate dimeglumine, which has a 95% renal excretion and a 5% hepatobiliary excretion.

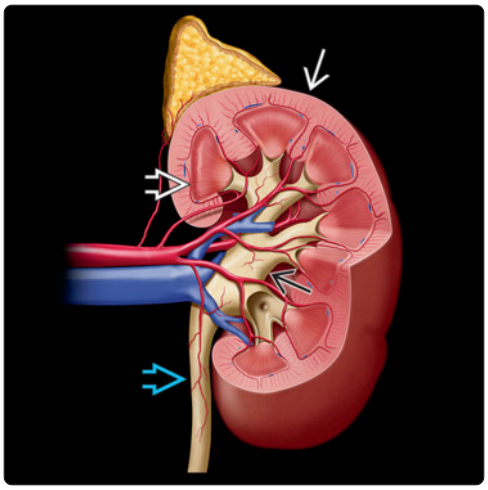
Nephrogenic systemic fibrosis (NSF): Systemic fibrosing disorder occurring in patients with renal dysfunction. An association with exposure to GBCA is accepted although the exact mechanism remains unknown. The disease has been reported weeks to months after contrast injection and no effective treatment is available. The major risk factors are renal dysfunction, type of GBCA (highest risk for gadodiamide), and cumulative dose of contrast. The incidence has significantly decreased after implementation of guidelines for contrast injection in patients at risk. According to the American College of Radiology (ACR) Committee on Drugs and Contrast Media, the renal function of patients at risk for NSF should be assessed by calculating the eGFR values. Recommendations on the injection of contrast are based on eGFR according to the ACR guidelines.

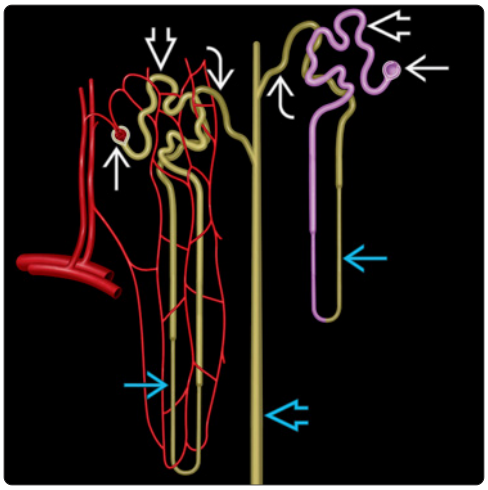
Adverse Reactions to Contrast Media

Adverse reactions to contrast media can be classified as allergic-like and physiologic and have an overall low incidence rate. The most significant risk factor for an allergic-like reaction is a documented allergic reaction to prior exposure. The majority of the reactions can be classified as mild and are non-life-threatening (e.g., limited urticaria, nausea/vomiting). Moderate (e.g., facial edema, vasovagal reaction) and severe (e.g., laryngeal edema with hypoxia, hypertensive emergency) adverse reactions need to be recognized and managed promptly. For prophylaxis measures and management of adverse reactions to contrast media, consult the most recent guidelines (version 9, 2013) from the ACR.

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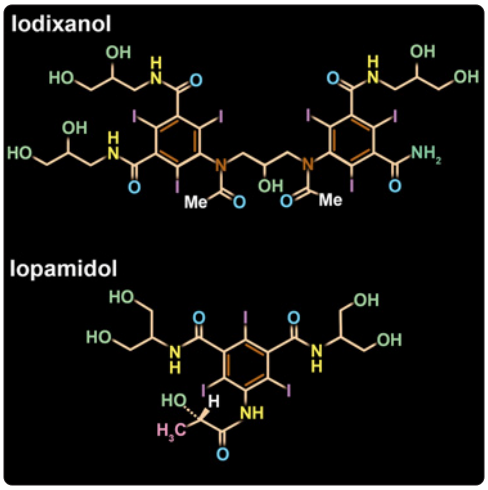
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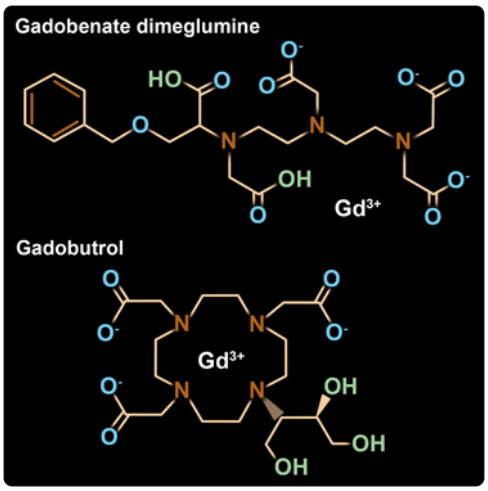
(Left) *Graphic shows renal anatomy and its 2 major components: Cortex (outer portion) and medulla (inner portion). The renal filtrate from each pyramid of the medulla is collected into minor and major calyces and passes into the renal pelvis and ureter.* **(Right)** *Graphic shows the nephron structure. The nephron is the functional unit of the kidney and it is composed of a renal corpuscle (glomerulus and Bowman capsule), proximal and distal convoluted tubule, loop of Henle, and collecting duct.*

First Generation: Ionic High-osmolar Contrast Media		
Name	Iodine content	Osmolality
Diatrizoate *(Hypaque 50)*	300 mg/ml	1550 mOsm/kg
Metrizoate *(Isopaque 370)*	370 mg/ml	2100 mOsm/kg
Second Generation: Non-ionic Low-osmolar Contrast Media		
Name	Iodine content	Osmolality
Iopamidol *(Isovue 370)*	370 mg/ml	800 mOsm/kg
Iohexol *(Omnipaque 350)*	350 mg/ml	880 mOsm/kg
Iopromide *(Ultravist 370)*	370 mg/ml	770 mOsm/kg
Ioxilan *(Oxilan 350)*	350 mg/ml	700 mOsm/kg
Third Generation: Non-ionic Iso-osmolar Contrast Media		
Name	Iodine content	Osmolality
Iodixanol *(Visipaque 320)*	320 mg/ml	290 mOsm/kg



(Left) *Table summarizes different generations of iodinated contrast media with their corresponding iodine content and osmolarity.* **(Right)** *Graphic shows the chemical structure of 2 iodinated contrast media with different osmolarity and number of triiodinated benzene groups. Iopamidol is an example of nonionic monomer, low-osmolar contrast medium, while Iodixanol is a nonionic dimer, iso-osmolar contrast medium.*

Group	Agent name	Structure	Charge
I: Largest number of NSF cases reported	· Gadodiamide · Gadoversatamide · Gadopentate dimeglumine	Linear Linear Linear	Non-ionic Non-ionic Ionic
II: Few, if any, unconfounded NSF cases reported	· Gadobenate dimeglumine · Gadoterate meglumine · Gadoteridol · Gadobutrol	Linear Macrocyclic Macrocyclic Macrocyclic	Ionic Ionic Non-ionic Non-ionic
III: Only recently introduced in the market (limited data available)	· Gadofosveset · Gadoxetate disodium	Linear Linear	Ionic Ionic



(Left) *Table summarizes the approved gadolinium-based contrast media. Agents are grouped according to risk of nephrogenic systemic fibrosis (ACR manual on contrast media, version 9).* **(Right)** *Graphic shows 2 gadolinium-based contrast media with different chemical structures. Gadobenate dimeglumine (Gd-BOPTA) is an example of an agent with linear ligand, while Gadobutrol (Gd-DO3A-butrol) is an example of a macrocyclic agent where gadolinium (Gd^{3+}) is positioned within the frame of a cyclic molecule.*

Imaging Anatomy

The kidneys are bean-shaped, paired, retroperitoneal organs measuring 10-15 cm in length and 5 cm in width. The main renal functions include the maintenance of homeostasis, removal of product of metabolism, hormone excretion, and urine production.

The renal parenchyma has 2 major components: The cortex (outer portion) and the medulla (inner portion). The renal parenchyma is divided in multiple lobes (8-18), each of which is composed of medulla (pyramid) and cortex. Each pyramid drains into a minor calyx. Multiple (2-3) minor calyces converge into a major calyces. The renal pelvis, an extension of the ureter, collects urine from multiple calyces. The renal pelvis may be located outside the renal hilum. The so-called extrarenal pelvis is an anatomic variant.

The functional unit is the nephron. It is composed of the renal corpuscule (glomerulus and Bowman capsule) and tubules (proximal and distal convoluted tubules and loop of Henle). The filtrate from the distal convoluted ducts passes into collecting ducts that extend to the tip of the medulla (papilla).

Renal arteries branch from the abdominal aorta and the renal veins drain into the vena cava. Accessory renal vessels are common, and correct identification and description is important for surgical planning. The renal lymphatic drainage is into arto-caval lymph nodes.

Embryology and Congenital Malformations

Early in embryologic development, the mesoderm on either side of the dorsal body wall develops into a succession of primitive kidneys. The metanephroi are the definitive functioning kidneys and are induced to form when the ureteric buds penetrate the metanephric blastema, a specialized mesoderm at the caudal end of the embryo. The ureteric bud then bifurcates, which in turn, induces the metanephric blastema to form lobes. As the lobes form, there is a reciprocal inductive effect on the branching of the ureteric bud forming the calyces. As the renal parenchyma develops, it will differentiate into an outer cortex, containing the glomeruli, and the medulla, containing the collecting ducts. The next important event in the development of the kidneys is the ascent from the sacral region, where they are formed to either side of the lumbar spine.

The complexity of the interaction of the metanephric blastema with the ureteric bud, coupled with the ascent of the kidneys, leads to a host of potential congenital renal anomalies, including persistent fetal lobation, prominent columns of Bertin, horseshoe kidneys, renal ectopia, renal duplications, ureteropelvic junction obstruction, and megacalyces.

Imaging Techniques and Indications

Ultrasound

Ultrasound is an important imaging modality for the assessment of the kidneys. In patients with acute or chronic kidney injury, ultrasound represents the imaging modality of choice to exclude obstruction (hydronephrosis) and assess renal size.

It is ideally suited for differentiating cystic from solid masses. A simple cystic lesion is anechoic with a sharp posterior border and increased through transmission. While ultrasound can confidently diagnose simple cysts, complicated cysts should be further evaluated with dedicated contrast-enhanced CT and MR.

A potential pitfall is renal lymphoma, which can have a "pseudocystic" appearance. Most renal cell carcinomas (RCC) have a heterogeneous hypo- to mildly hyperechoic appearance; however, 20% may have a more uniform hyperechoic appearance and be confused with an angiomyolipoma (AML). It is imperative that all solid masses be further evaluated with either CT or MR to differentiate AML from RCC.

CT

Contrast-enhanced, multiphase CT is the preferred imaging modality for the characterization of renal lesions.

Unenhanced scan: An unenhanced scan should be performed routinely in a renal mass work-up to evaluate for enhancement. For example, if a mass measures 50 HU after the administration of IV contrast, it is unclear if the lesion is a hyperdense cyst or an enhancing mass. Without knowing the attenuation value of the mass prior to the administration of IV contrast, this question remains unknown (unless delayed scans are performed). Renal calcifications are also best evaluated before contrast is administered. Fat can be confidently detected within a lesion when the attenuation is < - 10 HU. Finally, unenhanced images are helpful to detect hemorrhage.

Corticomedullary phase (20-80 seconds post-contrast injection): While this phase is helpful for evaluation of renal vasculature, it is fraught with pitfalls for the evaluation of renal masses. In this phase, the difference in enhancement between the renal cortex and medulla can be as much as 100 HU. Due to this difference, rapidly enhancing or high-attenuation masses can be missed in the cortex, while slowly enhancing or low-attenuation masses can be missed in the medullary portion of the kidney. This is particularly relevant in the setting of potential RCC. RCC has multiple different histologic types, several of which enhance slowly (e.g., papillary) and others which enhance early (e.g., clear cell).

Nephrographic phase (80-120 seconds post-contrast injection): In this phase, the kidney has an homogeneous uniform nephrogram. This phase may be delayed in patients with altered renal function. This is the best phase for the detection and evaluation of renal masses.

Excretory phase (3-5 minutes post-contrast injection): In this phase the contrast is excreted into the renal collecting system. With the advent of multidetector scanning, there has been a precipitous decline in the utilization of conventional excretory urography. 3D reconstructions in the excretory phase allow evaluation of the collecting system, including the renal pelvis, ureter, and bladder.

MR

MR imaging can be used as an alternative to CT or as a problem-solving tool to characterize indeterminate lesions detected on CT. MR has been proven particularly helpful for confirming the cystic nature of small lesions. The renal mass protocol usually consists of the following sequences.

T2-weighted imaging (T2WI): Simple cysts have high, homogenous signal intensity (SI) on T2WI, similar to the cerebrospinal fluid, whereas complicated, hemorrhagic cysts vary in SI depending on the stage of hemorrhage. Among solid lesions, papillary RCC and fat-poor AML have low SI on T2WI,

whereas clear cell and chromophobe RCC as well as oncocytoma usually show high SI on T2WI.

T1-weighted imaging (T1WI): In-phase and opposed-phase gradient-echo (GRE) ("chemical shift") imaging and frequency selective fat-suppressed imaging are helpful for the detection of intralesional fat. Intralesional "bulk fat" is a diagnostic feature of classic AML and manifests as a loss of SI on frequency selective fat-suppressed images and "India ink artifact" (signal loss at the interface of lesion and surrounding renal parenchyma) on opposed-phase T1-weighted images. Loss of SI within the lesion on opposed-phase images compared with in-phase images indicates a small amount of fat (intravoxel), which can be seen with few fat cells of fat-poor AML or intracellular fat of clear cell-type RCC. Renal lesions that are hyperintense on T1WI usually indicates proteinaceous or hemorrhagic cysts; in these lesions, it is important to confirm the lack of enhancement to exclude hemorrhagic solid lesion, such as RCC or AML.

3D fat-saturated GRE T1WI: This sequence is performed in the axial or coronal plane before and after the intravenous administration of an extracellular gadolinium-chelate-based contrast medium to assess organ and lesion enhancement. Similar to CT, images are obtained during the corticomedullary, nephrographic, and excretory phase. Enhancement can be assessed quantitatively or qualitatively.

Diffusion-weighted imaging: The application of this functional imaging technique to renal diseases is under investigation and has shown promising results in characterization of renal lesions and in the assessment of diffuse renal disease.

Approach to the Renal Mass

There are 4 basics questions that should be asked when evaluating a renal mass. Although there is certainly overlap within each of these groupings, by going through this algorithm, a reasonable differential can be formulated.

1) Is the mass cystic or solid?
Knowing if a mass is cystic or solid is the single most important question facing the radiologist when evaluating a renal mass. The key imaging feature for the differentiation is the presence of enhancement. A cystic mass is a lesion composed of nonenhancing fluid.

Cystic lesions: The vast majority of simple unilocular cystic lesions are benign cysts. Simple cysts are common with 50% of people over the age of 50 having 1 or more cysts. Complex cysts may still be benign, but a cystic RCC must always be considered. The Bosniak classification groups cystic lesions into 5 categories based on the CT imaging appearance; the more complex the lesion, the higher the risk of malignancy. The classification is used to determine management of cystic lesion. While simple or minimally complicated cysts (Bosniak class I and II) are considered benign and need no treatment or imaging follow-up, complicated cysts with thick septations or enhancing nodules (Bosniak class III and IV) are usually considered surgical lesions. Bosniak class IIF category includes complicated cysts requiring imaging follow-up. Clinical information is also helpful in further differentiating cystic masses. For example, a multilocular cystic nephroma (a benign cyst renal neoplasm) is seen in young boys and middle-aged women. Hematoma and abscess should also be suspected based on clinical history.

Lesions measuring < 20 HU or > 70 HU on unenhanced CT are considered benign, representing simple and hemorrhagic cysts, respectively. Lesions measuring 20-70 HU on unenhanced CT (i.e., hyperdense lesions) are indeterminate and require contrast administration to assess for enhancement. If the patient cannot tolerate IV contrast, ultrasound or MR can be used for characterization. At ultrasound, hyperdense lesions on CT may often appear as anechoic, benign cysts.

Solid lesions: An enhancing mass is considered a solid lesion. On CT, enhancement is considered present when there is at least 20 HU increase in attenuation after contrast injection. An increment of 10-20 HU is considered indeterminate and requires further evaluation with additional imaging, biopsy, or imaging follow-up. "Pseudoenhancement" refers to the artificial increase in attenuation of simple fluid-containing cystic lesion on post-contrast images. The phenomenon is mainly related to beam-hardening artifacts and to the reconstruction algorithm used to correct it, and it is accentuated in small (< 2 cm) lesions located within the renal parenchyma. On MR, subtraction images (post-contrast minus pre-contrast) are commonly used to qualitatively assess intralesional enhancement. A 15% increase in SI after contrast administration has been proposed as a sensitive threshold for the detection of renal lesion enhancement. While clear cell and chromophobe RCC usually show avid enhancement, papillary RCC is characterized by minimal enhancement. Benign renal lesions, such as oncocytoma and fat-poor AML, may show avid enhancement. Biopsy is often required to diagnose an indeterminate lesion.

2) What is the growth pattern; expansile or infiltrative ("balls" or "beans")?
If a mass is solid, the next step is to evaluate the growth pattern. Renal masses tend to have 2 basic growth patterns. They can have an expansile, circumferential growth pattern ("ball" shape) and are frequently exophytic, extending beyond the renal capsule and distorting the renal contour. Alternatively, masses can have an infiltrative growth pattern. These often start in the renal hila or medullary portion of the kidney. The kidney is often enlarged and diffusely abnormal, but the external contour retains its normal bean-shaped appearance.

Over 90% of RCCs have a spherical, expansile, ball-shaped growth pattern and commonly extend beyond the cortex of the kidney. Other solid lesions, including oncocytomas and AMLs, also have this growth pattern.

An infiltrative ("bean") pattern can be seen in both inflammatory and neoplastic conditions, and hence, clinical history is important in differentiating this group. In the acute setting, pyelonephritis and renal infarction can appear as an enlarged abnormally enhancing kidney. In a patient with hematuria, neoplasm should be considered; the most classic is an invasive transitional cell carcinoma. A minority of RCCs will have an infiltrative growth pattern, usually with aggressive histologic tumor types, including medullary carcinoma and sarcomatoid tumors. Finally, renal lymphoma may appear with either growth pattern.

3) Does the mass contain fat or just soft tissue?
If a solitary renal mass measures soft tissue density, the diagnosis is RCC until proven otherwise. Features, such as a central scar, may suggest oncocytoma as an alternative

diagnosis, but RCC cannot be excluded without histologic evaluation.

A fat-containing renal mass is overwhelmingly likely to be an AML, although RCC may rarely contain fat. This generally occurs when large tumors have engulfed fat or have undergone osseous metaplasia with the production of both dense calcification, similar to cortical bone, and fatty marrow elements. The presence of dense calcification and fat within a renal mass is highly suspicious for RCC.

A small percentage of AMLs (~ 5%) contain only a few fat cells and are named fat-poor AMLs and cannot be confidently differentiated from other solid lesions. The small amount of fat is not confidently detected at CT, and these lesions commonly appear homogeneously hyperattenuating on unenhanced CT. On MR, intralesional loss of SI can be seen on opposed-phase T1WI, though this is not a specific feature, as this signal loss can also be seen in clear cell RCC.

4) Is the mass solitary or are there multiple masses?
When multiple solid renal masses are seen, one should consider syndromes. Multiple RCCs and cysts occur in von Hippel-Lindau disease. Multiple AMLs should raise the index of suspicion for tuberous sclerosis. Lymphoma and metastases can also present with multiple masses.

If numerous cysts are seen, one should also consider autosomal dominant polycystic kidney disease, von Hippel-Lindau disease, or cystic changes secondary to long-term dialysis.

Approach to Renal Calcifications

Unenhanced scans (or plain films) are best for evaluating calcifications. Calcifications may occur in the collecting system (nephrolithiasis) or in the renal parenchyma (nephrocalcinosis). Nephrocalcinosis can be categorized into 3 large groups depending on appearance and location: Dystrophic calcification, cortical nephrocalcinosis, or medullary nephrocalcinosis.

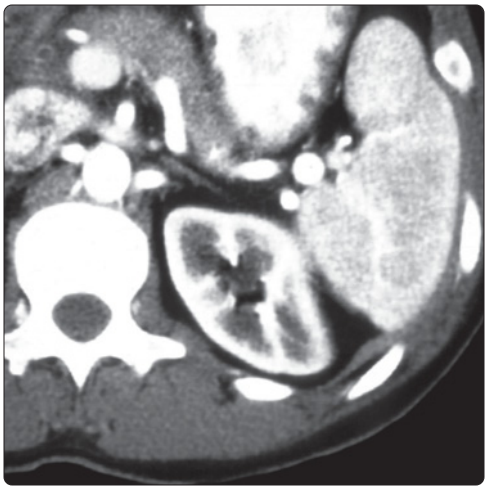
Dystrophic calcification: Occurring within abnormal or damaged tissue, dystrophic calcification is often focal and unilateral. It is most often seen in tumors (RCC) and infection (classically tuberculosis).

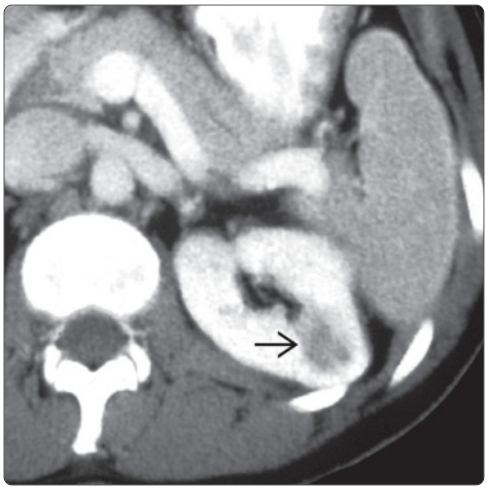
Cortical nephrocalcinosis: Described as appearing as eggshell calcification, cortical nephrocalcinosis features kidneys that are often small and nonfunctioning. Cortical nephrocalcinosis is most commonly seen in the setting of chronic glomerulonephritis but can also be the sequelae of acute cortical necrosis or chronic transplant rejection.

Medullary nephrocalcinosis: This typically occurs in otherwise normal-appearing kidneys and may result from hypercalcemia/hypercalcuria from any cause (e.g., hyperparathyroidism), medullary sponge kidney, or distal renal tubular acidosis. Calcifications precipitate within the renal tubules and are frequently associated with nephrolithiasis.

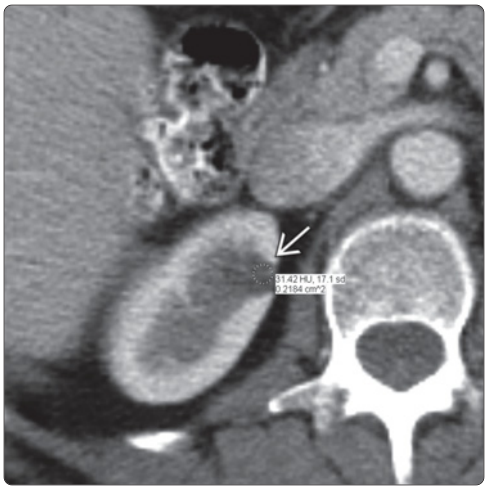
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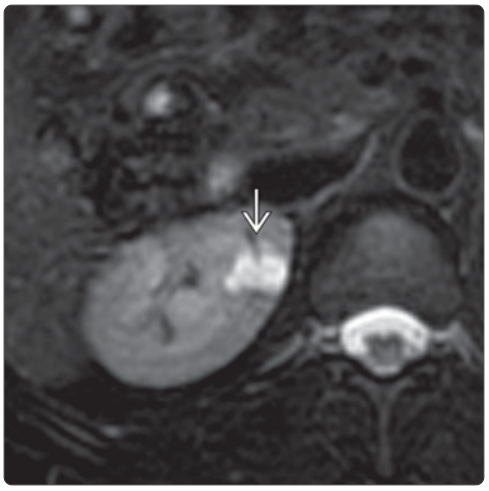
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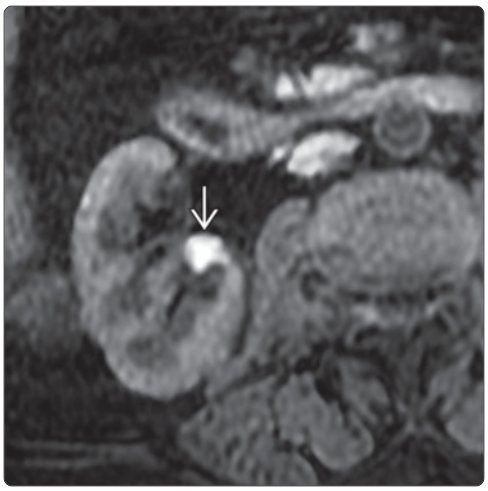


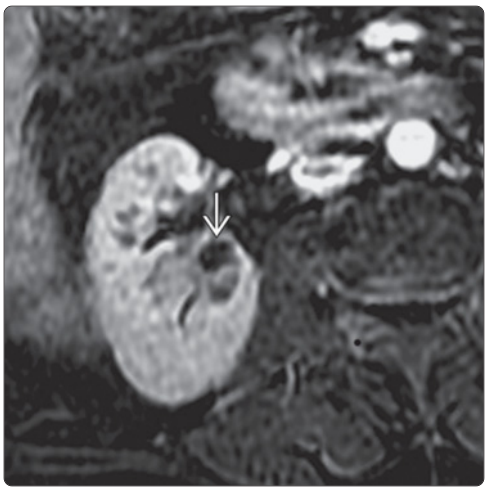
(Left) *Axial CECT acquired in the corticomedullary phase shows a normal left kidney.* **(Right)** *Axial CECT in the same patient acquired in the nephrographic phase shows an abnormal hypoenhancing area ⇨ in the medullary portion of the kidney. This patient had an elevated white count and pyuria, and this proved to be a focal area of pyelonephritis. Low-attenuation abnormalities in the medullary portion of the kidney are easily missed in the corticomedullary phase.*





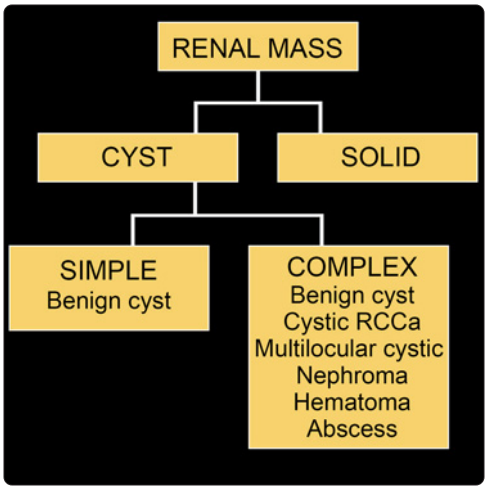
(Left) *Axial CECT shows a small, low-attenuation lesion ➡ in the cortex of the right kidney. The lesion measures 31 HU, and it is therefore indeterminate. It may represent a small cyst with "pseudoenhancement" or a small solid lesion.* **(Right)** *Axial T2 FS MR in the same patient shows a simple-appearing high-signal intensity (similar to the cerebrospinal fluid) lesion ➡ proving the cystic nature of the indeterminate right renal lesion seen on CT. The increased attenuation of the lesion on CECT is related to "pseudoenhancement."*

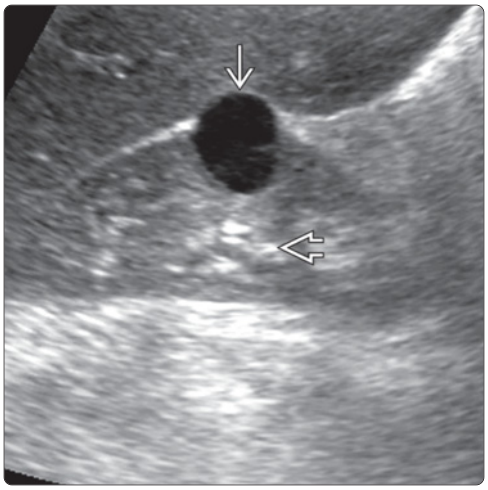




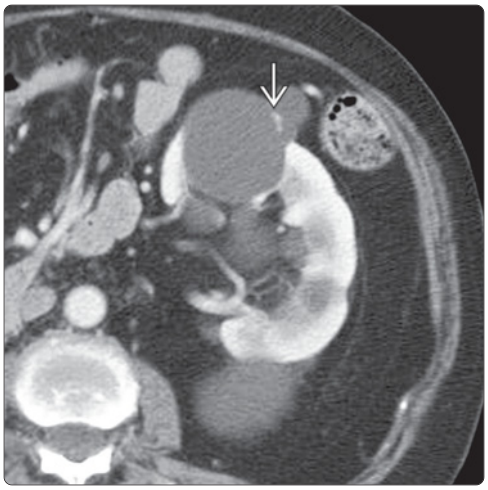
(Left) *Axial fat-suppressed T1WI shows a high signal intensity lesion ➡ in the cortex of the right kidney.* **(Right)** *Axial T1 subtraction (post-contrast minus pre-contrast) MR confirms the lack of enhancement within the lesion ➡. Renal lesions hyperintense on T1WI are usually proteinaceous or hemorrhagic lesions; the lack of enhancement confirms the cystic nature.*

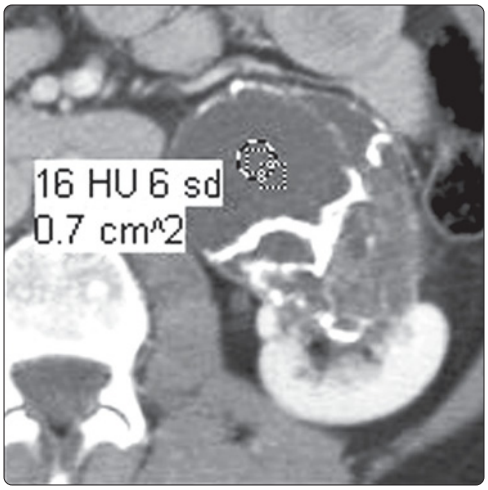
(Left) *An algorithm for analyzing a renal mass begins with deciding if it is cystic or solid. If cystic, then it must be determined if it is simple or complex . If multiple cysts are present, consider a syndrome such as autosomal dominant polycystic kidney disease, von Hippel-Lindau (may have both cystic and solid renal masses), or acquired cystic disease from dialysis.* **(Right)** *Longitudinal ultrasound of the right kidney shows an anechoic lesion ⇨ with imperceptible walls and posterior acoustic enhancement ⇨, compatible with a simple renal cyst.*



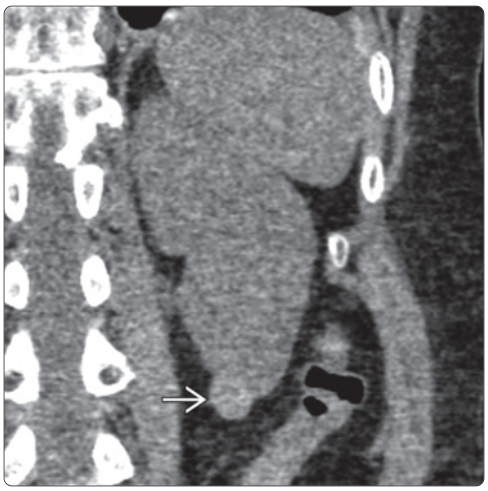


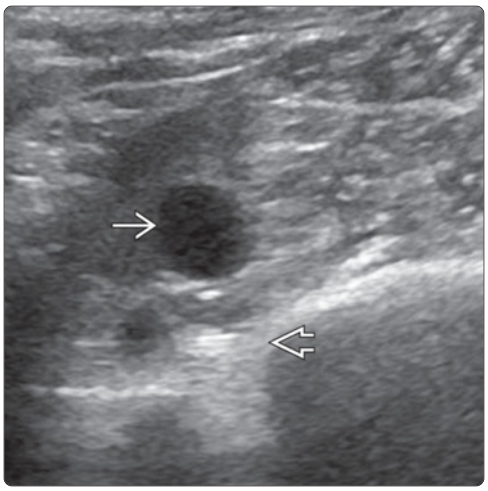
(Left) *Axial CECT shows a mildly complex benign cyst with a thin, partially calcified septum ⇨ (Bosniak class II).* **(Right)** *Axial CECT shows Bosniak class III cystic mass (multilocular cystic nephroma) with thick, calcified septa. The Bosniak classification groups cystic lesions into 5 categories based on the CT imaging appearance; the more complex the lesion, the higher the risk of malignancy.*

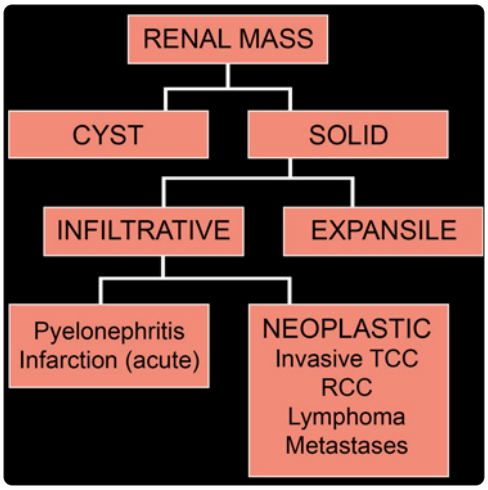


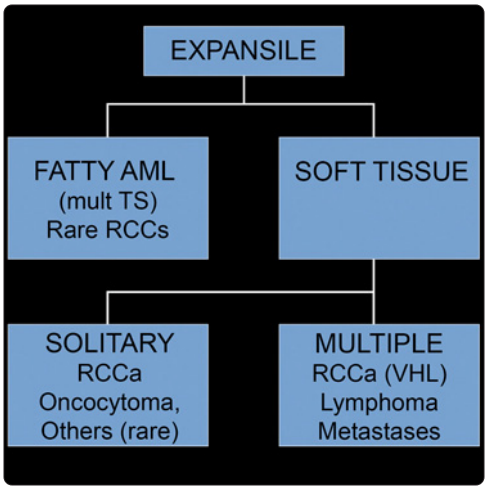


(Left) *Coronal NECT shows a small, exophytic, and hyperattenuating (65 HU) lesion ⇨ in the lower pole of the left kidney.* **(Right)** *Transverse ultrasound of the left kidney in the same patient shows the same lesion ⇨ as anechoic with imperceptible walls and posterior acoustic enhancement ⇨, confirming it as a simple cyst. At ultrasound, hyperdense lesions on CT may often appear as anechoic, benign cysts.*

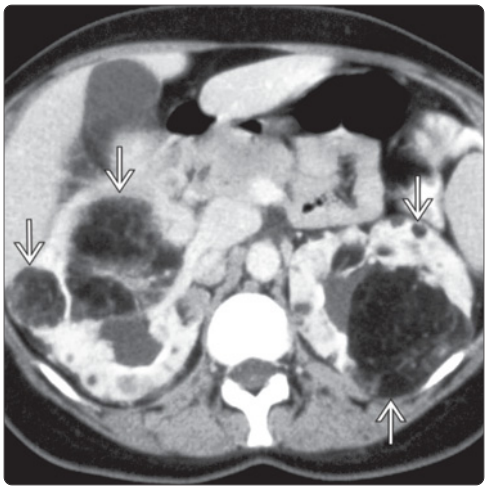






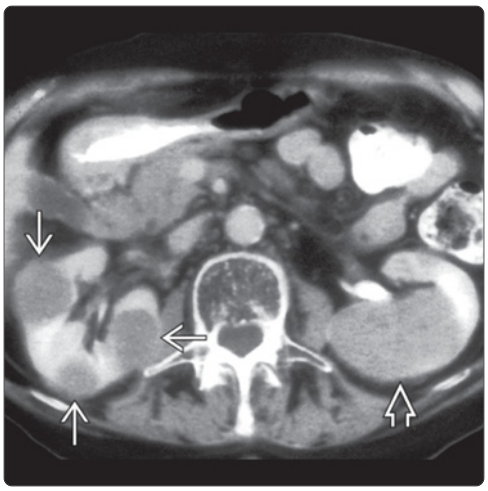


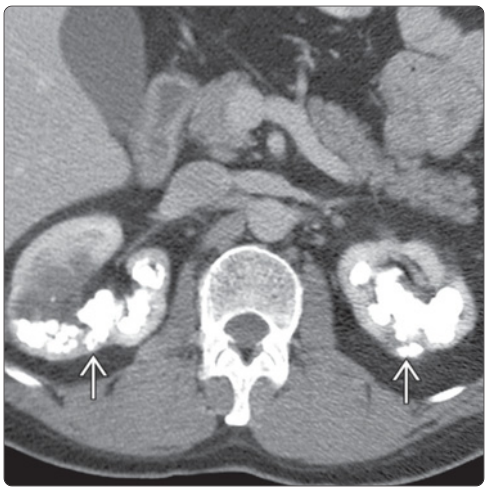
(Left) *If a mass is solid (i.e. enhancing), the next question is, What is the pattern of growth? This graphic shows the primary differential considerations for an infiltrative mass.* **(Right)** *If a mass grows in a spherical expansile pattern, the next question is the fat content. The vast majority of fat-containing renal lesions are angiomyolipomas (AML). A solitary, soft tissue density mass is a renal cell carcinoma (RCC) unless proven otherwise. RCCs may be multiple, but lymphoma and metastases should also be considered.*





(Left) *Axial CECT shows multiple bilateral renal fat-containing masses ➡ of varying size, consistent with multiple classic (fat-rich) angiomyolipomas in a patient with tuberous sclerosis.* **(Right)** *Axial T1WI subtraction image (post-contrast minus pre-contrast) shows enhancing, partially exophytic right renal mass ➡ which was proven as clear clear cell renal cell carcinoma at resection. Subtraction images are commonly used to qualitatively assess lesion enhancement on MR imaging.*





(Left) *Axial CECT shows multiple renal lesions with 2 patterns of growth. Within the right kidney are 3 well-defined, round, ball-shaped masses ➡, while on the left there is a larger infiltrating mass ➡. Note the left kidney retains a normal renal contour ("bean" shape). When there are multiple masses displaying both types of growth, lymphoma is the primary consideration.* **(Right)** *Axial CECT shows extensive, bilateral medullary nephrocalcinosis ➡ in this case of primary oxalosis.*

Renal Fetal Lobation

KEY FACTS

TERMINOLOGY

- Synonym: Fetal lobulation, persistent fetal lobulation
- Normal variant anatomy with indentations of renal surface caused by incomplete fusion of renal lobes

IMAGING

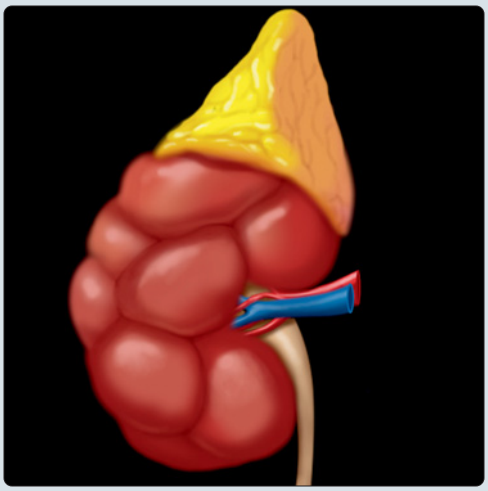
- US
  - Color Doppler shows normal flow and cortical thickness separated by thin echogenic cleft
  - Lobations are clefts of echogenic capsule and perirenal fat invaginating between normal-thickness renal cortex
- CECT during corticomedullary phase shows normal thickness cortex, invaginated by thin, hypodense surface cleft
- Morphology
  - Sharp, shallow indentations along renal surface
  - Indentations extend between renal pyramids and calyces

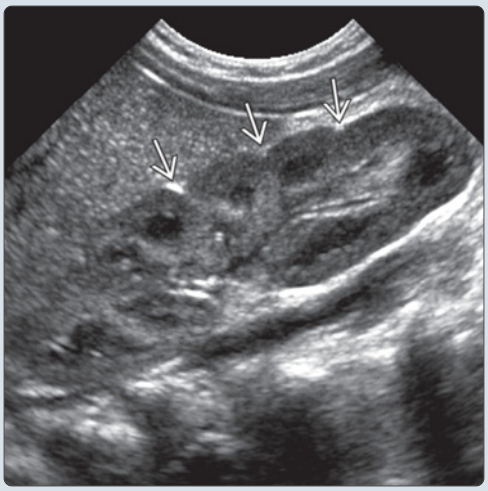
TOP DIFFERENTIAL DIAGNOSES

- Cortical scar
  - Loss or thinning of cortex due to ischemia or infection
  - Often **overlies** distorted calyx (lobation clefts are between calyces)

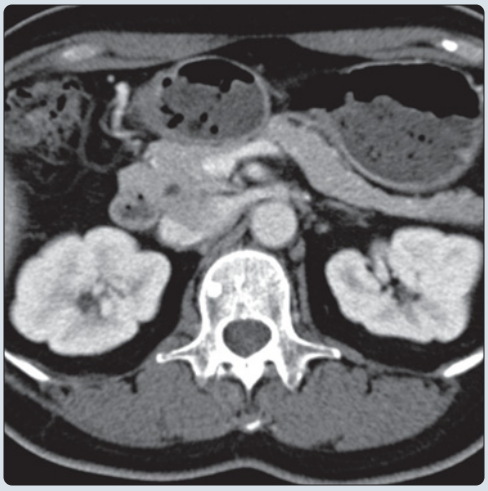
PATHOLOGY

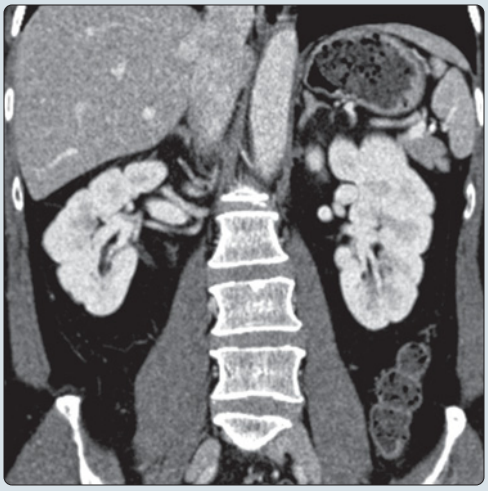
- Fetal human kidney consists of numerous separate renal lobes (reniculi)
  - Each lobe consists of 1 renal pyramid surrounded by cortex and supplied by single calyx, artery, and vein
  - Each lobe marked by surface indentations (lobations)
- Lobes usually disappear by end of childhood, resulting in smooth surface of most adult kidneys
  - Fetal lobation present in > 10% of children, 4% of adults
  - Lobation is often prominent on sonography of newborns





(Left) *This graphic depicts the typical lobulated appearance of the kidney in fetal life, reflecting the development of the kidney from numerous lobes, each consisting of a renal pyramid and its associated cortex. Note that the adrenal gland is relatively large compared to the kidney in fetal life and childhood.* **(Right)** *Sagittal US in this 6-week-old infant shows fetal lobation (clefts ⇨). Note echogenicity of the cortex and paucity of renal sinus fat, which are normal findings in infants and children.*





(Left) *Axial CECT in the corticomedullary phase shows classic bilateral fetal lobation. Note that each surface cleft or indentation extends into a column of normal-thickness renal cortex.* **(Right)** *Coronal reformation of CECT in the same patient shows classic bilateral fetal lobation, with each renal lobe and its cortex demarcated by surface clefts.*

TERMINOLOGY

Synonyms

- Fetal lobulation
- Persistent fetal lobulation

Definitions

- Normal variant anatomy with indentations of renal surface caused by incomplete fusion of renal lobes

IMAGING

General Features

- Morphology
  - Shallow indentations along renal surface
    - Extending into columns of cortical tissue (columns of Bertin)
  - Usually bilateral
  - More common in children

Ultrasonographic Findings

- Multiple hyperechoic indentations along renal surface
  - Lobations are clefts of echogenic capsule and perirenal fat invaginating between normal-thickness renal cortex
- Lobation is often prominent on sonography of newborn infants
  - Tends to diminish with age
  - US appearance of kidneys of neonates/infants is different from adults
    - Renal cortex is more echogenic than in adults
    - Medullary pyramids are hypoechoic
    - Less renal sinus fat
  - In adults, renal cortex is relatively less echogenic and renal sinus fat is more prominent

CT Findings

- Sharp, shallow indentations **between** renal pyramids and calyces
  - Deeper indentation often seen between upper and lower poles in duplex kidney
- Indentations contain hypodense perirenal fat and can extend into columns of cortical tissue (columns of Bertin)
- Corticomedullary phase offers better delineation of cortex and its thickness

Imaging Recommendations

- Best imaging tool
  - US: Color Doppler shows normal flow and cortical thickness separated by thin, echogenic cleft
- Protocol advice
  - CECT during corticomedullary phase shows normal-thickness cortex, invaginated by thin, hypodense surface cleft

DIFFERENTIAL DIAGNOSIS

Cortical Scar

- Loss or thinning of cortex due to ischemia or infection
- Often overlies distorted calyx (lobation clefts are **between** calyces)

PATHOLOGY

General Features

- Fetal human kidney consists of numerous, separate renal lobes (known as reniculi)
  - Similar to those present in lower animals
  - Each lobe consists of 1 renal pyramid surrounded by cortex and supplied by single calyx, artery, and vein
- Each lobe is marked by surface indentations (lobations)
  - These usually disappear by end of childhood, resulting in smooth surface of most adult kidneys
  - Failures of fusion are common, resulting in
    - Persistent lobation of surface (fetal lobation)
    - Columns of cortical tissue extending deeply into renal sinus (prominent column of Bertin)
    - Pseudotumors, cortical dysmorphism
  - Deep surface clefts (junctional parenchymal defect) and prominent columns of Bertin are most common between upper and mid 1/3 of kidney

CLINICAL ISSUES

Demographics

- More common in infants and children
  - Present in > 10% of children
  - Seen in 4% of adults
- No gender predilection

Natural History & Prognosis

- Normal anatomical variant with no clinical significance

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1. Mazziotti S et al: Usefulness of contrast-enhanced ultrasonography in the diagnosis of renal pseudotumors. Abdom Imaging. 35(2):241-5, 2010
2. Bhatt S et al: Renal pseudotumors. AJR Am J Roentgenol. 188(5):1380-7, 2007
3. Dalla Palma L et al: Radiological anatomy of the kidney revisited. Br J Radiol. 63(753):680-90, 1990
4. Patriquin H et al: Fetal lobation. An anatomo-ultrasonographic correlation. J Ultrasound Med. 9(4):191-7, 1990

Junctional Cortical Defect

KEY FACTS

TERMINOLOGY

- Normal-variant line at plane of embryologic fusion of renunculi (embryologic elements forming kidneys)

IMAGING

- Classically seen in anterosuperior aspect of kidney (at junction of upper and middle thirds of kidney)
  - Less commonly seen in posteroinferior aspect (at junction of middle and lower thirds of kidney)
- R > L (~ 2x more common on right side)
- Runs obliquely from anterosuperior toward posteroinferior surface; usually extends to renal sinus
- Better seen on US, due to higher spatial resolution and multiplanar imaging (better seen on parasagittal planes)
- US
  - Echogenic line running obliquely from anterosuperior to posteroinferior surface of kidney (junctional line)
  - Peripheral wedge-shaped echogenicity along plane of fusion of renunculi (caused by Invagination of perirenal fat)

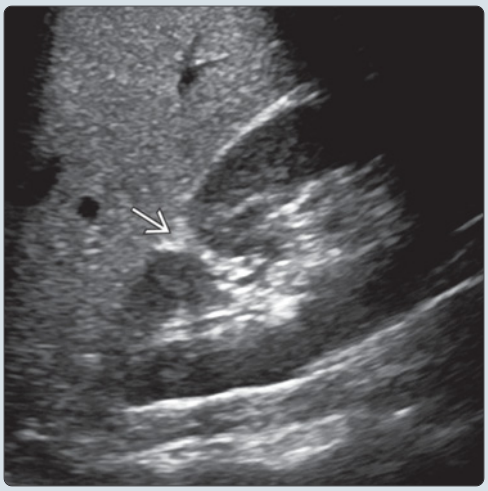
TOP DIFFERENTIAL DIAGNOSES

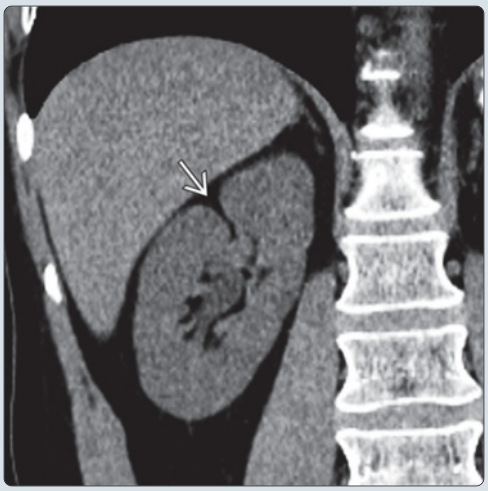
- Renal scar
  - Lack of cortical thinning and classic location of JPD helps to differentiate these entities
- Angiomyolipoma (AML)
  - Classic location, preservation of renal contour, and extension toward renal sinus help to differentiate JPD from AML

PATHOLOGY

- Based on concept that kidneys develop from fusion of 2 sub kidneys (renunculi)
  - Posterosuperior and anteroinferior renunculi fuse to form kidney
- Invagination of perirenal fat into defect results in its wedge-shaped appearance

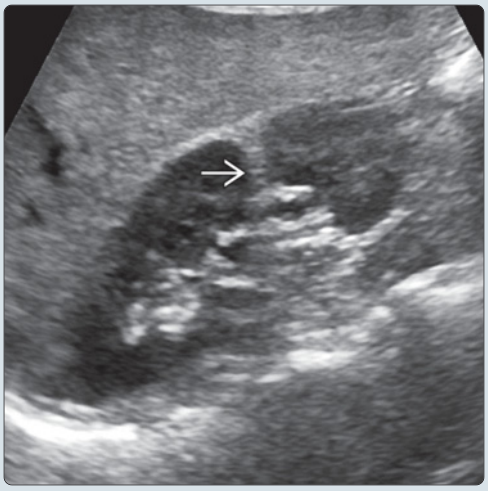
(Left) *Sagittal oblique US through the right kidney in 55-year-old man shows triangular echogenicity ➡ in the anterior aspect of the upper pole, which extends into the renal sinus, compatible with junctional parenchymal defect.* **(Right)** *Coronal CT in the same patient better shows the fat-density notch ➡ and the defect through the upper pole.*





(Left) *Sagittal oblique US through the right kidney in a 48-year-old woman shows an echogenic wedge-shaped defect ➡ in the anterior aspect of the lower pole.* **(Right)** *A slightly different section through the right kidney in the same patient shows extension of the defect ➡ into the renal sinus. The anterior aspect of the lower pole is a less common location for junctional parenchymal defect. Note increased hepatic echogenicity due to steatosis.*





TERMINOLOGY

Abbreviations

- Junctional parenchymal defect (JPD)

Synonyms

- Junctional parenchymal defect, interrenicular septum, junctional line

Definitions

- Normal-variant line at plane of embryologic fusion of renunculi (embryologic elements forming kidneys)
  - Invagination of perirenal fat through this defect results in wedge-shaped appearance

IMAGING

General Features

- Location
  - Classically seen in anterosuperior aspect of kidney (at junction of upper and middle thirds of kidney)
    - Less commonly seen in postero-inferior aspect (at junction of middle and lower thirds of kidney)
  - R > L (~ 2x more common on right side)
  - Runs obliquely from anterosuperior toward posteroinferior surface; usually extends to renal sinus

CT Findings

- Fat-containing notch in anterosuperior (or less commonly in posteroinferior) aspect of kidney, which may or may not extend to renal sinus

MR Findings

- Defect along the anterosuperior, or less commonly in posteroinferior, aspect of kidney (at the junction of middle and lower thirds of kidney) with signal characteristics similar to the perirenal fat
- The defect may or may not extend to renal sinus
- Better seen on coronal images

Ultrasonographic Findings

- Echogenic line running obliquely from anterosuperior to posteroinferior surface of kidney (junctional line)
- Peripheral wedge-shaped echogenicity along plane of fusion of renunculi (caused by Invagination of perirenal fat)

Imaging Recommendations

- Best imaging tool
  - Better seen on US, due to higher spatial resolution and multiplanar imaging
- Protocol advice
  - Best seen on parasagittal planes

DIFFERENTIAL DIAGNOSIS

Renal Scar

- Has associated cortical thinning and is usually in polar regions
  - Lack of cortical thinning and classic location of JPD helps to differentiate these entities

Angiomyolipoma (AML)

- Round JPD can mimic solid mass
  - Classic location, preservation of renal contour, and extension toward renal sinus help to differentiate JPD from AML

PATHOLOGY

General Features

- Based on concept that kidneys develop from fusion of 2 sub kidneys (renunculi)
  - Posterosuperior and anteroinferior renunculi fuse to form kidney
  - Incomplete fusion of renunculi results in spectrum of normal variants including junctional line, JPD, and column of Bertin; these normal variants are highly associated with each other
  - Invagination of perirenal fat into defect results in its wedge-shaped appearance

CLINICAL ISSUES

Demographics

- In study of 100 children, 1/2 of participants and 1/3 of kidneys had JPD
- No age or sex predilection reported

Natural History & Prognosis

- Normal anatomic variants with no clinical significance

SELECTED REFERENCES

1. Pedrosa I et al: MR imaging of renal masses: Correlation with findings at surgery and pathologic analysis. RadioGraphics 28:985-1003; 2008
2. Currarino G et al: The Oddono's sulcus and its relation to the renal "junctional parenchymal defect" and the "interrenicular septum". Pediatr Radiol. 27(1):6-10, 1997
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5. Hoffer FA et al: The interrenicular junction: a mimic of renal scarring on normal pediatric sonograms. AJR Am J Roentgenol. 145(5):1075-8, 1985
6. Dalla Palma L et al: Advances in radiological anatomy of the kidney. Br J Radiol. 55(654):404-12, 1982
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Column of Bertin

KEY FACTS

TERMINOLOGY

- Hypertrophic bands of cortical tissue that separate pyramids of renal medulla
- Synonyms: Hypertrophied or prominent column of Bertin, septal cortex

IMAGING

- Normally enhancing renal cortex protruding into renal sinus
  - Between upper and middle calyces
  - More commonly unilateral and on left side
  - Splaying (abnormal separation) of upper and lower pole of collecting system
  - Most evident on corticomedullary phase
- Similar echogenicity/density/intensity to normal cortex
- Normal renal contour
- Coronal reconstructions helpful if any question on axial images

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma and other renal masses including metastases, oncocytoma, and angiomyolipoma
  - Usually not isoechoic to renal cortex
  - Often distort contour of kidney
- Fetal lobation and renal scarring
  - Abnormal renal contour

PATHOLOGY

- Results from unresorbed polar parenchyma of 1 or both sub-kidneys (renunculi) during embryogenesis

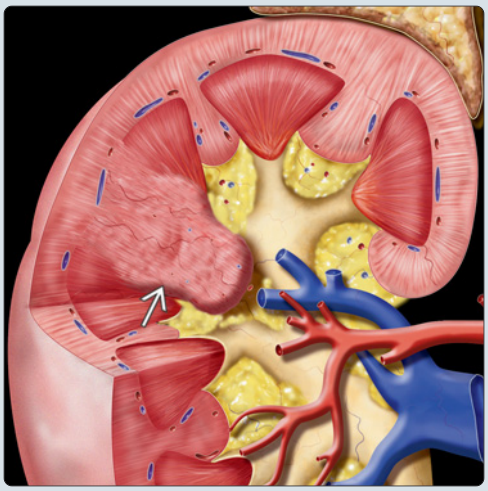
CLINICAL ISSUES

- Normal variant of no clinical significance
- Do not mistake for tumor

DIAGNOSTIC CHECKLIST

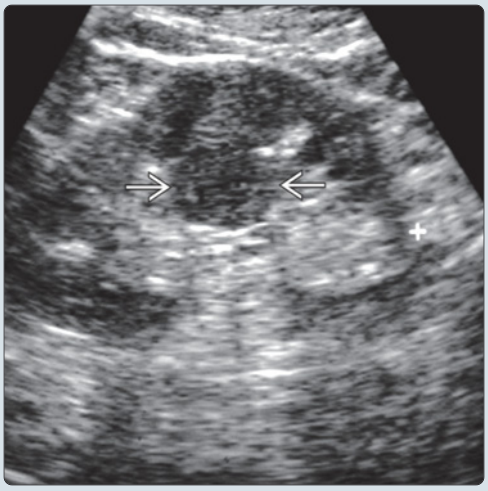
- CECT in corticomedullary phase is diagnostic

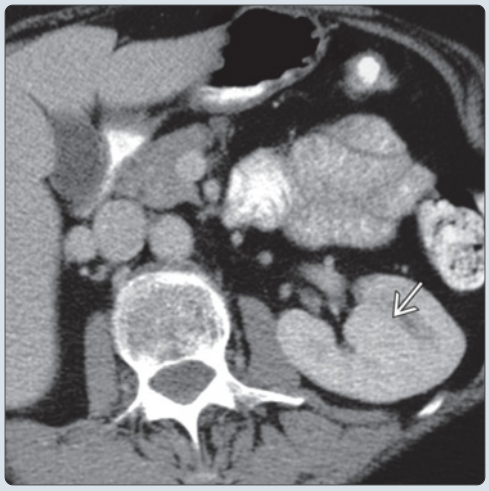
(Left) *Graphic shows a mass-like extension of the renal cortex ⇨ between the upper and middle calyces.* **(Right)** *Axial CT in the corticomedullary phase of enhancement shows the dorsal half of the interpolar kidney ⇨ enhancing in a similar manner to renal cortex ⇨. Subsequent phases showed homogeneous enhancement of the kidney.*





(Left) *Longitudinal ultrasound shows a central hypoechoic "mass" ⇨ that protrudes into the renal pelvis.* **(Right)** *Axial CECT in the same patient shows that the "mass" ⇨ is normal renal cortex that has invaginated into the renal pelvis between the calyces of the upper pole and interpolar kidney. Note the external contour of the kidney is normal. A column of Bertin is more commonly unilateral and on the left, as in this case. It can be bilateral in 18% of cases.*





TERMINOLOGY

Synonyms

- Septal cortex, hypertrophied or prominent column of Bertin, focal cortical hyperplasia, benign cortical rest, cortical island, focal renal hypertrophy, junctional parenchyma

Definitions

- Hypertrophic bands of cortical tissue that separate pyramids of renal medulla

IMAGING

General Features

- Best diagnostic clue
  - Normally enhancing renal cortex protruding into renal sinus
- Location
  - Between upper and middle calyces
  - Left side > right side
  - Unilateral > bilateral (18% of cases)

Radiographic Findings

- IVP
  - Mass effect on pelvicalyceal system, always at level of emerging renal vein
  - Splaying (abnormal separation) of upper and lower pole of collecting system

CT Findings

- NECT
  - Continuous with renal cortex
  - Lateral indentation of renal sinus
  - Deformed adjacent calyces and infundibula
- CECT
  - Absence of mass
  - Enhancement similar to normal renal cortex
    - Most evident on corticomedullary phase
    - Coronal reconstructions helpful if any question on axial images

MR Findings

- Hypertrophied column with signal intensity and enhancement pattern, similar to the cortex on all sequences

Ultrasonographic Findings

- Grayscale ultrasound
  - Normal renal contour
  - Typically isoechoic relative to renal cortex
  - Color and power Doppler show flow similar to cortex
  - Splaying of central sinus echoes

Nuclear Medicine Findings

- In equivocal cases, renal scintigraphy can differentiate normal renal parenchyma from pathologic mass

Imaging Recommendations

- Best imaging tool
  - Multiphasic CT or MR
- Protocol advice
  - CT: Corticomedullary phase is best

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma

- Heterogeneous renal mass with enhancement different from surrounding parenchyma
- Conventional type (clear-cell type) is hypervascular, although hypodense to kidney on nephrographic phase
- Mass is usually round or oval and usually causes contour deformity

Other Renal Masses

- Including renal metastases and lymphoma, oncocytoma, and angiomyolipoma
- Usually not isoechoic to renal cortex
- Often distort contour of kidney

Fetal Lobation

- Persistent cortical lobulation, usually seen in young children
- Individual lobes with centrilobar cortex located around calyces

Renal Scarring

- Focal scarring from chronic reflux nephropathy, surgery, trauma, or infarction
- Nodular compensatory hypertrophy of areas of unaffected tissue

PATHOLOGY

General Features

- Unresorbed polar parenchyma of 1 or both sub-kidneys (renunculi) that fuse to form normal kidney

CLINICAL ISSUES

Natural History & Prognosis

- Normal variant of no clinical significance

DIAGNOSTIC CHECKLIST

Consider

- Pseudotumor, not pathological disease

Image Interpretation Pearls

- CECT in corticomedullary phase is diagnostic

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Horseshoe Kidney

KEY FACTS

TERMINOLOGY

- Congenital anomaly in which kidneys are fused by isthmus at lower poles

IMAGING

- CT: Defines structural abnormalities, complications, and associated abnormalities
- 2 types of fusion
  - Midline or symmetrical fusion (90% of cases)
  - Lateral or asymmetrical fusion
- Variant arterial supply
  - Multiple, bilateral renal arteries
  - Arteries arising from aorta or iliacs

TOP DIFFERENTIAL DIAGNOSES

- Renal ectopia
- Renal displacement

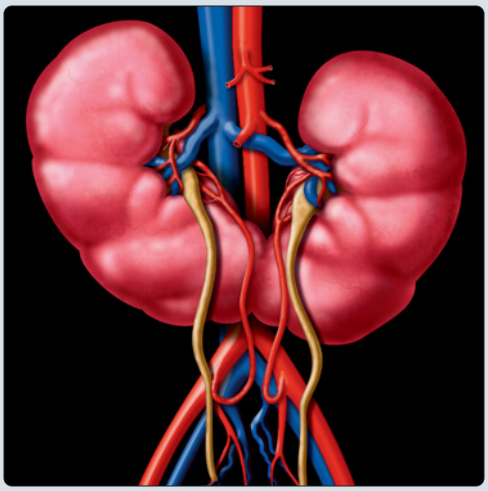
PATHOLOGY

- Other anomalies of kidneys and other organs are common, such as
  - Ureteropelvic junction (UPJ) obstruction
  - Vesicoureteral reflux

CLINICAL ISSUES

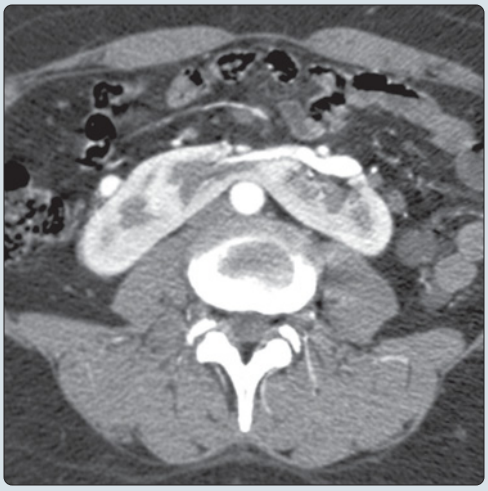
- Complications
  - Traumatic injury
  - Recurrent infections: Vesicoureteral reflux and UPJ obstruction
  - Urolithiasis: 75% metabolic calculi, 25% struvite calculi
  - In children, Wilms tumor 2-8x more common in patients with horseshoe kidney
  - Primary renal cell carcinoma: ↑ prevalence

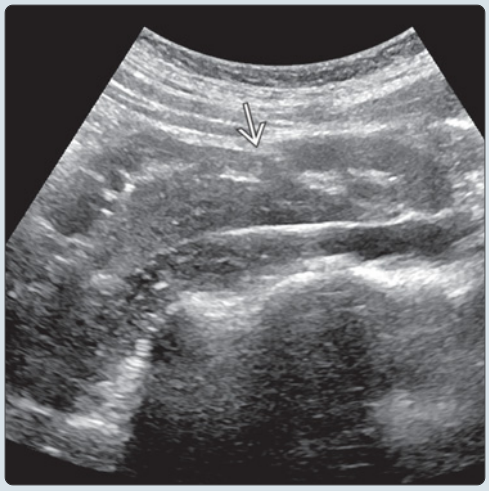
(Left) *Graphic shows horseshoe kidney. Note multiple renal arteries arising from the aorta and iliac arteries.* **(Right)** *IVP shows horseshoe kidney. Note the lower calyces descending toward midline near isthmus, and the U-shaped nephrogram.*





(Left) *Axial CECT shows fusion of the lower poles of the kidneys across the midline and low position of kidneys. The isthmus contains functioning enhancing parenchyma. Note the abnormal forward-facing calyces.* **(Right)** *US shows horseshoe kidney with isthmus ⇨ in contiguity with lower poles.*





TERMINOLOGY

Definitions

- Congenital anomaly in which kidneys are fused by isthmus at lower poles

IMAGING

General Features

- Best diagnostic clue
  - 2 kidneys on opposite sides of body with lower poles fused in midline
- Location
  - Ectopic, lies lower than normal kidneys
  - Isthmus usually anterior to aorta and caudal to confluence of iliac veins
  - Rarely, isthmus is anterior to aorta and dorsal to inferior vena cava (IVC)
- Morphology
  - 2 types of fusion
    - Midline or symmetrical fusion (90% of cases)
    - Lateral or asymmetrical fusion

Radiographic Findings

- Radiography
  - Kidney appears too close to spine
  - Vertical long axis of kidney may be seen, lower poles lie closer to spine
- IVP
  - Midline fusion
    - "Hand-holding" calyces: Lower calyces descend toward midline near isthmus
    - Nephrogram is U-shaped
    - Rarely, calyces from opposing kidneys join to form common renal pelvis with 1 ureter
  - Lateral fusion
    - Lower calyces cross midline and drain part of renal parenchyma of opposite kidney
    - Nephrogram is L-shaped
    - 1 part crosses midline and lies in transverse position, renal pelvis lies anteriorly or laterally
    - Remaining part lies in vertical position, renal pelvis lies anteriorly or medially
  - Large and extrarenal renal pelvis
    - Renal pelvis often large and flabby; ureter inserts abnormally high in renal pelvis
  - Rarely, kidney is fused at upper poles (5%)
  - Ipsilateral lower calyces medial to ureter, may simulate renal malrotation without fusion
  - Ureteropelvic (more common) or ureterovesical junction obstruction with delayed clearing of contrast
  - "Flower vase" appearance: Each ureter crosses isthmus, curves laterally, and continues medially, assuming normal course distally
  - Bifid or double ureters drain kidneys
- Voiding cystourethrography
  - Vesicoureteral reflux often seen

CT Findings

- CECT
  - Defines structural abnormalities
    - Degree and site of fusion: Midline or lateral fusion
    - Shows degree of renal malrotation
    - Renal parenchymal changes (e.g., scarring, cystic disease)
    - Collecting system abnormalities (e.g., duplex system, hydronephrosis)
  - Differentiate composition of isthmus between fibrous or normal parenchyma
    - CT almost always shows functioning parenchyma in isthmus
- CTA
  - Variant arterial supply
    - Multiple, bilateral renal arteries
    - Arteries arising from aorta or common iliac, internal iliac, external iliac, or inferior mesenteric arteries
    - Inferior mesenteric artery always crosses isthmus

Ultrasonographic Findings

- Grayscale ultrasound
  - Isthmus lies anterior to spine and is contiguous with lower poles of opposite kidneys
  - Curved configuration, elongation, and poorly defined lower poles
  - Inverted triangular or pyriform shape (longitudinal scan)

Angiographic Findings

- Catheter angiography shows variant arteries

Nuclear Medicine Findings

- Demonstrates fusion with functional parenchymal tissue
  - Can detect regional loss of function due to obstruction and inflammation

Imaging Recommendations

- Best imaging tool
  - CECT with multiplanar and 3D reconstructions
  - US for diagnosis in utero
- Protocol advice
  - CTA: Use 3D volume-rendered CT to better define vessels

DIFFERENTIAL DIAGNOSIS

Renal Ectopia

- Kidney congenitally in abnormal position
- Ipsilateral or simple ectopia: Kidney and its ureter on appropriate side
  - Abdominal: Kidney lies above iliac crest but below L2
  - Iliac: Kidney is located in iliac fossa
  - Pelvic (sacral): Kidney in true pelvis
- Crossed renal ectopia: 2 kidneys on same side of body (R > L)
  - With fusion (90%)
    - 2 fused kidneys lie on same side of spine; ureter of crossed kidney crosses midline to insert into bladder
  - Without fusion
    - 2 kidneys lie on same side of spine without fusion; ureter of crossed kidney crosses midline to insert into bladder
  - Solitary
    - 1 kidney arises on wrong side; ureter crosses midline to insert into bladder

- Bilateral
  - Left and right kidneys arise on wrong side; both ureters cross midline to insert into bladder

Renal Displacement

- Thoracolumbar gibbous deformity: Alters renal axis → pseudohorseshoe kidney
- Displacement by mass: Forcing 1 kidney to opposite side of body or into pelvis
  - Retroperitoneal masses
  - Giant hydronephrosis secondary to ureteropelvic junction (UPJ) obstruction
  - Giant renal cyst (e.g., autosomal dominant polycystic kidney disease)

PATHOLOGY

General Features

- Genetics
  - Reported in identical twins, but no clear evidence
- Associated abnormalities
  - Congenital disorders
    - Chromosomal abnormalities: Turner syndrome, trisomy 18
    - Hematological abnormalities: Fanconi anemia, dyskeratosis congenita with pancytopenia
    - Laurence-Biedl-Moon syndrome
    - Thalidomide embryopathy
  - Anomalies (most common to least common)
    - Ureteropelvic junction obstruction
    - Vesicoureteral reflux
    - Unilateral or bilateral duplication
    - Megaureter
    - Ectopic ureter
    - Unilateral triplication
    - Renal dysplasia
    - Retrocaval ureter
    - Supernumerary kidney
    - Anorectal malformation
    - Esophageal atresia
    - Rectovaginal fistula
    - Omphalocele
    - Cardiovascular, vertebral, neurological, peripheral skeletal, or facial anomalies

Gross Pathologic & Surgical Features

- Isthmus is composed of normal parenchyma (more common) or connective tissue

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic or associated abnormalities
  - Vague abdominal pain, radiating to back
    - Ureteral obstruction
  - Palpable midline abdominal mass

Demographics

- Age
  - Any age
  - Stillbirths > infants > children > adults; ↓ with age because many are diagnosed based on associated abnormalities
- Gender
  - M:F = 2:1
- Epidemiology
  - 1 in 400 people
  - Most common renal fusion anomaly

Natural History & Prognosis

- Complications
  - Traumatic injury
    - Isthmus lies anteriorly without protection by ribs → split by hard blow to abdomen
  - UPJ obstruction: High "insertion" of ureter
  - Recurrent infections: Vesicoureteral reflux and UPJ obstruction
  - Urolithiasis: 75% metabolic calculi, 25% struvite calculi
  - In children, Wilms tumor is 2-8x more common in patients with horseshoe kidney
  - Primary renal cell carcinoma: ↑ prevalence
- Prognosis
  - Poor, with associated abnormalities causing significant morbidity and mortality
  - Good, without other abnormalities

Treatment

- Surgical separation in symptomatic patients

DIAGNOSTIC CHECKLIST

Consider

- Associated abnormalities and other complications in imaging, treatment, and prognosis

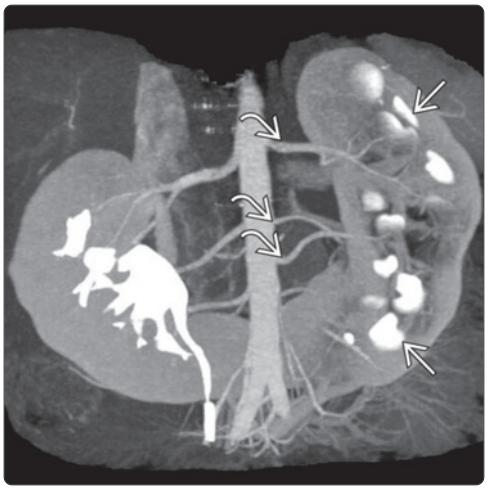
Image Interpretation Pearls

- Kidney appears U-shaped with isthmus in midline

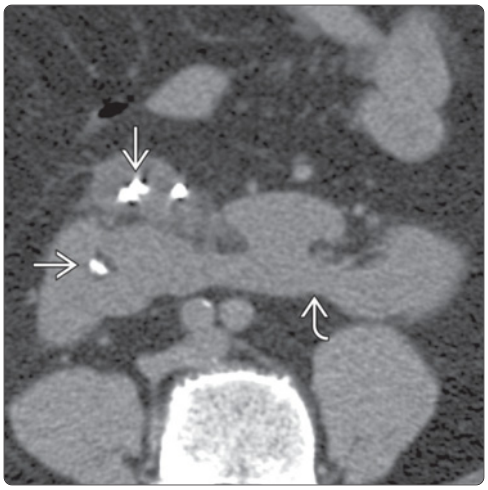
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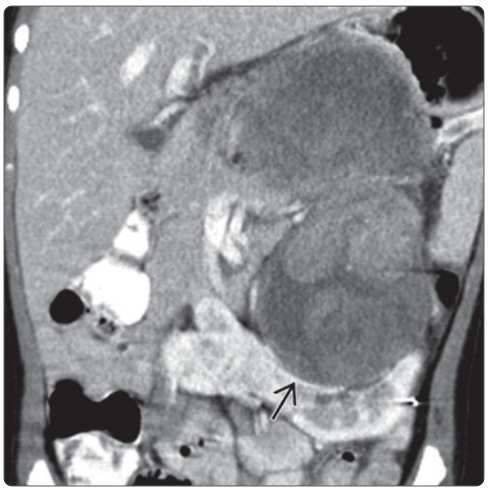
(Left) *Axial delayed CECT demonstrates horseshoe kidney with dilated left-sided calyces. Multiple renal arteries cross the isthmus and renal pelvis and may contribute to obstruction.* **(Right)** *Coronal CECT volumetric reformation in the same patient demonstrates multiple renal arteries, some of which have probably crossed and partially obstructed the infundibula on the left side. The left side calyces are dilated, and the cortex is thinned.*





(Left) *Axial NECT shows horseshoe kidney with multiple calculi and cortical scarring.* **(Right)** *Axial CECT shows large laceration of the left moiety of horseshoe kidney with surrounding hematoma.*





(Left) *Axial CECT shows a large mass replacing most of the isthmus and left moiety of horseshoe kidney. The mass proved to be renal cell carcinoma.* **(Right)** *Coronal CECT in a 3-year-old boy shows a large, heterogeneous mass arising from the left moiety of horseshoe kidney. The mass proved to be Wilms tumor.*

Renal Ectopia and Agenesis

KEY FACTS

TERMINOLOGY

- Abnormal kidney location due to developmental anomaly

IMAGING

- Ipsilateral renal ectopia: Kidney on same side of body as orifice of its attendant ureter
  - Usually lies caudal to its normal position
- Crossed renal ectopia: Kidney located on opposite side of midline from its ureteral orifice
  - With fusion (85-90%), without (10-15%)
- Ectopic kidneys often have errors of rotation
  - Renal pelvis pointed anteriorly, laterally, posteriorly
  - Often impairs urinary drainage
- Renal agenesis
  - Congenital anomaly often associated with other genitourinary and systemic anomalies

TOP DIFFERENTIAL DIAGNOSES

- Renal allograft
- Renal autotransplantation
- Post nephrectomy
- Horseshoe kidney
- Acquired renal displacement
- Acquired renal atrophy

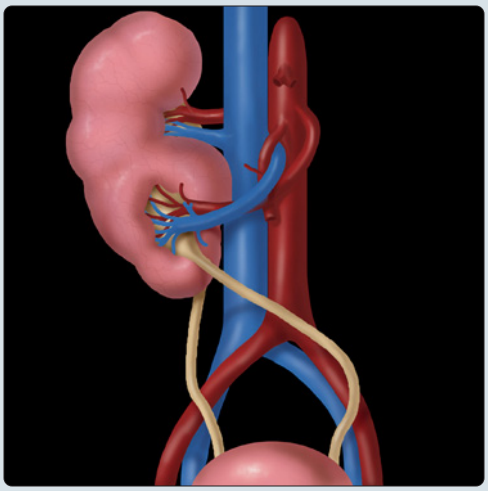
PATHOLOGY

- VACTERL: Associated anomalies of vertebrae, anorectum, cardiovascular, tracheoesophageal, renal, and limb
- MURCS: Müllerian ducts, renal, cervicothoracic spine
- Ectopic kidneys prone to trauma, urinary stasis, infection, stones

CLINICAL ISSUES

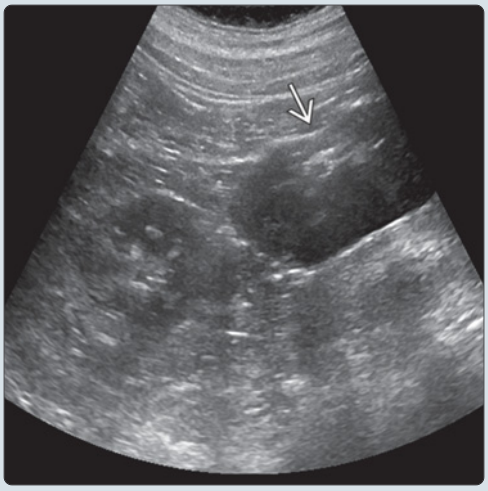
- Complications
  - Obstruction, urolithiasis, reflux, infection
  - Aberrant arteries may cross and obstruct ureter
  - Ectopic kidneys more prone to blunt traumatic and iatrogenic injury

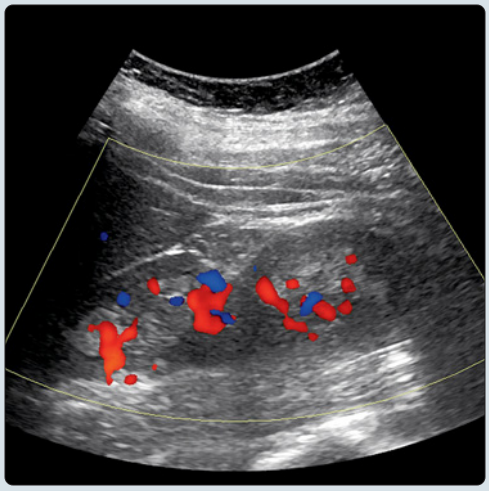
(Left) *Graphic shows crossed inferior fused renal ectopia. Note that the left kidney lies on the side opposite its ureteral orifice, findings that define "crossed" ectopia.* **(Right)** *Axial CECT shows the left kidney fused to the lower pole of the right kidney in this patient with crossed fused ectopia. The hilum of the left kidney is rotated anteriorly, and both renal moieties are abnormally low in position. The left ureter entered the bladder at the normal site of the left trigone.*





(Left) *Sagittal ultrasound of the right flank in a patient with crossed renal ectopia without fusion shows that the right kidney (RK) has proper position and rotation; the ectopic left kidney is inferior to right kidney and malrotated. Note the separation between the two kidneys.* **(Right)** *Sagittal color Doppler ultrasound of the right flank in a patient with crossed renal ectopia with fusion shows lack of separation between the two kidneys.*





TERMINOLOGY

Abbreviations

- Renal ectopia (RE)

Definitions

- Abnormal kidney location due to developmental anomaly

IMAGING

General Features

- Best diagnostic clue
  - Abnormal location or congenital absence of kidney
- Location
  - 1st-3rd lumbar vertebrae normal site of kidneys
  - Ipsilateral RE: Kidney on same side of body as orifice of its attendant ureter
    - Cranial (superior RE): Above normal position, intrathoracic or below eventrated diaphragm
    - Caudal (simple RE): Below normal position, abdominal, iliac, or pelvic
    - Abdominal: Kidney lies above iliac crest, below L2
    - Iliac: Kidney located in iliac fossa
    - Pelvic (sacral): Kidney located in true pelvis, below iliopectineal line
  - Crossed RE: Kidney located on opposite side of midline from its ureteral orifice
- Other general features
  - Caudal RE
    - Unilateral (more common), bilateral is rare
    - Affecting solitary kidney (least common)
  - Crossed RE
    - With fusion (85-90%), without fusion (10-15%)
  - Classification of unilateral fused kidney or crossed fused RE
    - Superior: Kidney crosses over midline, lies superior to resident kidney (rare)
    - Sigmoid (S-shaped): Crossed kidney lies inferiorly
    - Unilateral lump kidney: Both kidneys completely fused; large, irregular lump
    - Unilateral L-shaped: Crossed kidney inferior and transverse; resident kidney normally oriented
    - Unilateral disk ("pancake"): Kidneys fused to each other along medial concave border
    - Inferior RE: Crossed kidney inferior to resident; upper pole fused to lower pole of resident kidney
      - Most common form of crossed ectopia
  - Renal agenesis
    - Congenital anomaly often associated with other genitourinary and systemic anomalies

Radiographic Findings

- IVP
  - Cranial renal ectopia
    - Kidney lies partially or completely in thorax
    - Attendant ureter longer than normal
  - Abdominal or iliac RE: Kidney in either abdominal or iliac area
    - Can be simple or crossed
    - Kidney usually smaller and ureter shorter than normal
    - Bizarre pattern of calyces; extrarenal calyces (common)
    - Rotational anomalies: Incomplete, reverse, or nonrotation
  - Pelvic kidney
    - Left (70%) > right; if bilateral, left usually inferior to right kidney and generally fused
    - May see anomalies of rotation
    - Ureter frequently too high as it exits renal pelvis ("high insertion")
    - May see ectopic ureter, extrarenal calyces
  - Crossed ectopia: Kidney located opposite side of midline from its ureteral orifice
    - Both pelvises rotated anteriorly in superior, unilateral lump and inferior RE
    - Sigmoid (S-shaped): Resident kidney pelvis is medial; lateral in crossed kidney
    - Unilateral disc: Resident kidney pelvis is anteromedial; pelvis of other is anterolateral
  - Bilateral crossed RE: Both kidneys on wrong side, but their attendant ureters arise normally

CT Findings

- Cranial RE: Kidney residing in thorax, differentiate from mediastinal mass
  - Adrenal gland may lie above, behind, or below ectopic kidney
- Abdominal or iliac RE
  - Adrenal gland in normal place, appears linear on CT
  - Colonic flexures, duodenum, loops of small bowel, spleen, tail of pancreas, in abnormal (medial) position
- Pelvic RE: Differentiate RE from various pelvic masses
- May show separation between apparently fused kidneys
- Renal agenesis: Ipsilateral vertical colon segment lies medial to its normal position
  - Does not occur after nephrectomy (although small bowel may occupy renal fossa)

Ultrasonographic Findings

- Grayscale ultrasound
  - Cranial RE
    - Kidney lies just below eventrated diaphragm
    - Or, passing through defect in diaphragm
  - Caudal RE (abdominal, iliac, or pelvic)
    - Renal sinus echo complex: Eccentric or absent
  - Crossed RE
    - Separation of kidneys may be demonstrated
    - Kidneys may move separately during respiration
  - Crossed fused RE
    - 1 or 2 anterior or posterior notches in renal parenchyma
    - Renal sinuses lie in different planes, run in different directions, & reflect echoes differently

Angiographic Findings

- Conventional
  - Cranial RE
    - Elongated renal artery from aorta at normal level
    - Occasionally, accessory RA from thoracic aorta
  - Abdominal or iliac RE
    - Renal arteries arise lower in aorta than normal

  - Multiple renal arteries (common)
- Pelvic RE
  - Often multiple renal arteries
  - Usually arise from distal end or aortic bifurcation
- Crossed RE: Anomalous blood supply arising from vessels in vicinity

Nuclear Medicine Findings

- Tc-99m DMSA or Tc-99m glucoheptonate scan
  - Detects ectopic kidney by outlining kidney shape
  - Crossed fused renal ectopia: Isotope excretion or localization by kidney, with no contralateral isotope excretion or localization

Imaging Recommendations

- Best imaging tool
  - Multiplanar CECT
- CECT with multiplanar reformations, CT urography

DIFFERENTIAL DIAGNOSIS

Renal Allograft

- Transplanted kidney in iliac fossa
- Renal vessels anastomosed to external or internal iliac artery and vein
- Ureter reimplanted into bladder via submucosal tunnel; variable axis of renal pelvis

Renal Autotransplantation

- Surgical repositioning patient's own kidney

Post Nephrectomy

- Ipsilateral colon will remain in its normal position

Horseshoe Kidney

- Fusion of lower poles of kidneys in low mid-abdomen

Acquired Renal Displacement

- Due to large liver, splenic, or any retroperitoneal tumor or mass

Acquired Renal Atrophy

- May be mistaken for renal agenesis; may be source of symptoms

PATHOLOGY

General Features

- Etiology
  - Cranial RE: Kidney herniated into thorax through lumbocostal triangle or foramen of Bochdalek
  - Caudal RE: Diminished ureteral growth; umbilical arteries block cranial ascent of kidney; asymmetry in level of development of 2 kidneys
  - Crossed RE: Mesonephric ducts and ureteral buds may stray from normal course
  - RE inherited as autosomal recessive trait, reported in monozygotic twins
- Associated abnormalities
  - Extremely common association of other anomalies and various syndromes
  - **VACTERL**: Associated anomalies of vertebrae, anorectum, cardiovascular, tracheoesophageal, renal, and limb
  - **MURCS**: Müllerian ducts, renal, cervicothoracic spine
  - Genitourinary (50%): Malrotation, hypospadias, high insertion of ureter into renal pelvis, ectopic ureter, extrarenal calyces, calyceal diverticula, bladder extrophy
  - Skeletal (40%): Anomalies of ribs, vertebral bodies, skull asymmetry and absence of radius
  - Cardiovascular (40%): Valvular and septal defects
  - Gastrointestinal (33%): Anorectal malformations, malrotation
  - Ears, lips, palate (33%): Low-set or absent ears, hare lip, cleft palate
  - Hematopoietic (7%): Fanconi anemia

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be asymptomatic, incidental finding, or present with signs and symptoms of obstruction, urolithiasis, reflux, or infection

Demographics

- Gender
  - Cross-fused ectopia, M < F
- Epidemiology
  - Bilateral renal agenesis: 3.5 in 100,000 (lethal to fetus)
  - Unilateral agenesis: 1 in 1,000 people, M > F, L > R
  - Crossed fused ectopia: ~ 1 in 7,000 births
  - Anomalies of position (simple ectopia): ~ 1 in 500

Natural History & Prognosis

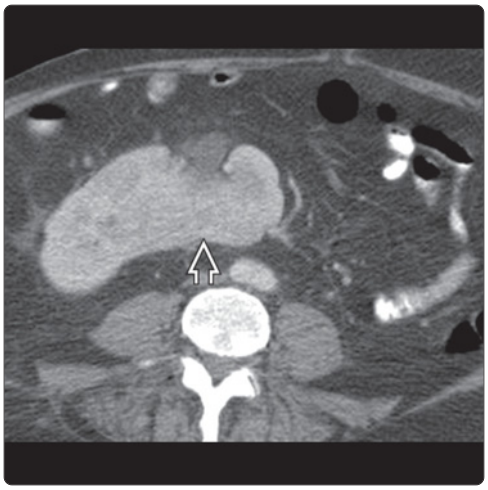
- Complications
  - Obstruction, urolithiasis, reflux, infection
  - Pelvic kidneys: ↓ function; may obstruct labor
  - Aberrant arteries may cross and obstruct ureter
  - Abdominal and iliac ectopic kidneys more prone to blunt traumatic and iatrogenic injury (e.g., during vascular surgery)
- Prognosis
  - Good for simple ectopia; fatal for bilateral agenesis; variable for remainder

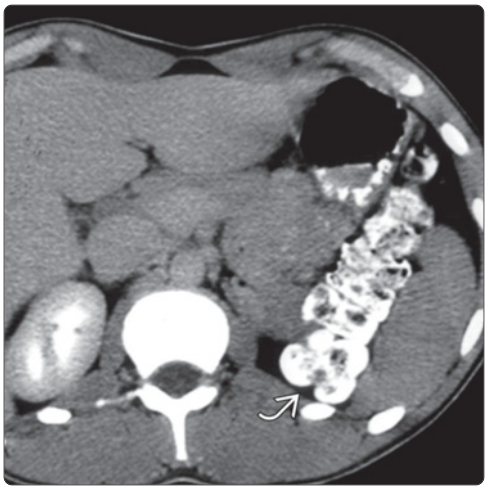
Treatment

- Treat complications of renal ectopia, such as ureteral obstruction and stones

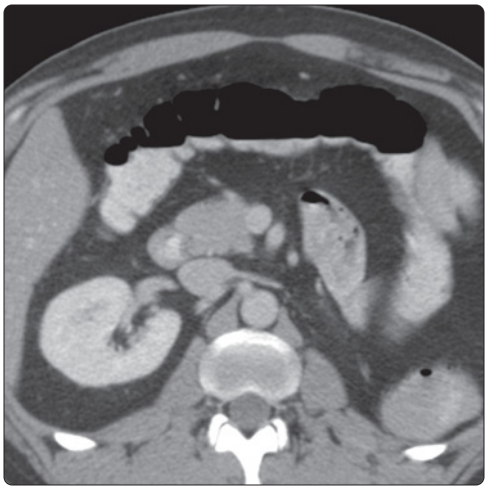
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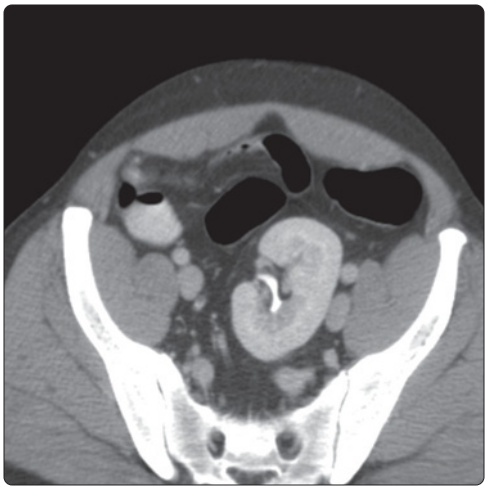
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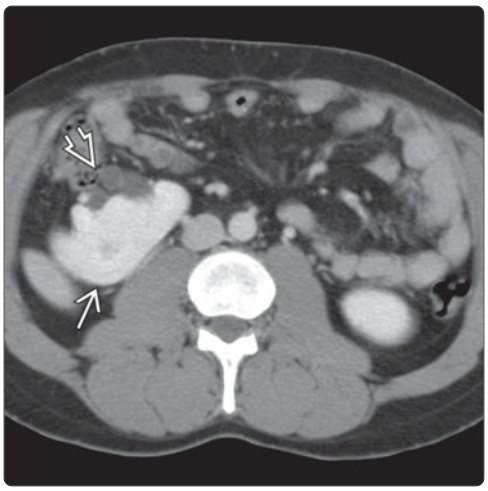


(Left) *Axial CECT of crossed fused ectopia shows the absence of a kidney in the left renal fossa, with colon and small bowel in its place. The mid left kidney is fused to the lower pole of the right, resulting in a "pancake" kidney.* **(Right)** *Axial CECT in a patient with unilateral renal agenesis shows absence of the left kidney and evidence that this was congenital as the descending colon lies in an unusually medial position.*





(Left) *Axial CECT in a patient with caudal simple ectopia (iliac kidney) shows the right kidney in a normal position but no kidney in the left renal fossa. Unilateral ectopia is far more common than bilateral anomalies. Ectopic kidneys are almost always abnormally low in position, representing a failure to "ascend" from their pelvic position in early fetal life.* **(Right)** *Axial CECT in the same patient shows the left kidney deep in the pelvis but functioning normally. There were no associated anomalies.*





(Left) *Axial CECT shows caudal ectopia with malrotation. The left kidney is normal, but the right kidney is abnormally caudal and malrotated, with the renal hilum facing anterior. The renal pelvis is dilated, suggesting impairment of urinary drainage.* **(Right)** *In the same patient, the abnormally rotated and dilated right renal pelvis is better seen on this caudal section. Anomalies of position and rotation often result in impairment of drainage and may result in chronic renal injury.*

Ureteropelvic Junction Obstruction

KEY FACTS

TERMINOLOGY

- Functional or anatomic obstruction to urine flow from renal pelvis into ureter at their anatomic junction
  - If left untreated, results in symptoms, renal damage, or both

IMAGING

- Pyelocaliectasis with abrupt UPJ narrowing
- Imaging recommendations
  - Neonates and children: Ultrasonography
  - Adults: CT urography and CTA ± diuresis renography
  - Depiction of relation of UPJ obstruction to vessels is important for planning endoluminal surgery
  - Diuretic renography: Tc-99m labeled mercaptoacetyltriglycine (MAG3) to quantitate relative obstruction and function

TOP DIFFERENTIAL DIAGNOSES

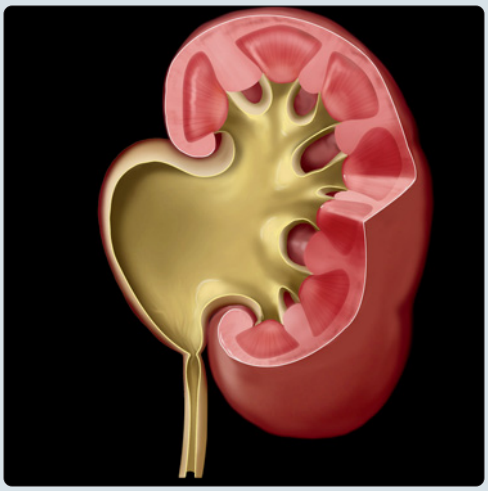
- Extrarenal pelvis
  - No caliectasis or delayed excretion
- Urolithiasis (renal calculi)
  - Can obstruct at UPJ; easily seen on CT and MR
- Tumor
  - Intrinsic or extrinsic; usually seen on CT and MR
- Extrinsic indentation
- Retrocaval ureter
- Renal pelvis blood clot

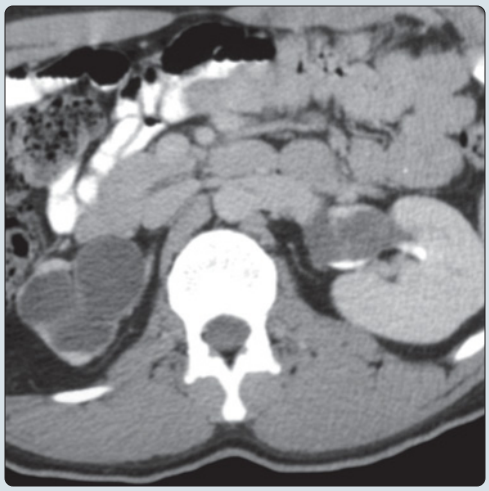
CLINICAL ISSUES

- Most common signs/symptoms
  - Neonates: Prenatal screening; palpable mass
  - Children and adults: Abdominal pain, hematuria
- Demographics
  - Any age; less commonly presents in adults

DIAGNOSTIC CHECKLIST

- CT and MR defines altered morphology
- Diuretic renogram defines function





(Left) *Graphic shows a congenital ureteropelvic junction (UPJ) obstruction with abrupt narrowing and angulation at the UPJ with resulting pelvocaliectasis and mild cortical atrophy.* **(Right)** *Axial CECT shows a dilated renal pelvis, caliectasis, and parenchymal atrophy of the right kidney, presumed to be due to a congenital UPJ obstruction that was never treated. For comparison, an extrarenal pelvis is seen in the left kidney. Note the absence of caliectasis or renal parenchymal injury.*





(Left) *Axial CECT of bilateral UPJ obstruction shows marked bilateral hydronephrosis and cortical atrophy with contrast-opacified urine in the dependent portions of the dilated calyces. The ureters were of normal caliber, and there was no obstructing mass or other abnormality.* **(Right)** *Upright radiograph in the same patient, with 30-minute delay after CECT, shows bilateral pelvocaliectasis and delayed clearance of contrast from the collecting systems. The normal-caliber ureters are poorly visualized.*

TERMINOLOGY

Abbreviations

- Ureteropelvic junction (UPJ) obstruction

Definitions

- Functional or anatomic obstruction to urine flow from renal pelvis into ureter at anatomic junction
  - If left untreated, results in symptoms, renal damage, or both

IMAGING

General Features

- Best diagnostic clue
  - Pyelocaliectasis with abrupt UPJ narrowing and delayed excretion
- Location
  - Left kidney > right kidney (2:1)
  - Unilateral (70-90%), bilateral obstruction (10-30%)

Radiographic Findings

- IVP
  - Marked pyelocaliectasis; UPJ narrowing
  - Increased or decreased renal size
    - Chronic, severe obstruction → renal atrophy
  - Nephrogram and pyelogram patterns
    - "Soap bubble" nephrogram: Parenchymal thinning → opacified, thin, overlapping rims of parenchyma
    - "Negative pyelogram": Early films; delayed opacification of collecting system
    - Calyceal crescents: Thin, semilunar, opacified, collecting tubules in periphery of dilated calyces
    - "Puddles" on pyelogram: Contrast settles in dependent portions of dilated calyces
  - Delayed clearing of contrast from collecting system
- Voiding cystourethrography
  - Exclude severe vesicoureteral reflux in infants
- Retrograde pyelography
  - Assess ureter if not visualized in other studies
  - Jet of contrast squirting through narrow UPJ into nonopacified, urine-filled renal pelvis

CT Findings

- NECT
  - Displays level of obstruction and degree of hydronephrosis
  - Shows possible etiologies (e.g., renal calculi, horseshoe or duplex kidney)
- CECT
  - Displays level of obstruction and degree of hydronephrosis
  - Shows any associated anomalies (e.g., aberrant vessels)
  - Distinguish from acquired lesions that obstruct (e.g., retroperitoneal fibrosis, tumor)
- CTA
  - 3D and multiplanar reformations (CT urography)
    - Optimally includes axial, MPRs, volume-rendered 3D displays
      - Shows relationship of renal vessels to UPJ
      - Useful in planning endoscopic or surgical pyloroplasty

MR Findings

- MR and MRA
  - Alternative to CT and CTA for depiction of anatomy and possible congenital or acquired abnormalities
- MR urography as alternative to CT urography

Ultrasonographic Findings

- Grayscale ultrasound
  - Use US in both prenatal and postnatal evaluation
  - Prenatal findings
    - Pelvic or pyelocalyceal dilatation
      - Anteroposterior (AP) pelvic diameter > 10 mm, pelvic:AP renal diameter ratio > 0.5, caliectasis: Fetal hydronephrosis
      - Mild pyelectasis (pelvic diameter 4-10 mm in < 20 weeks and 5-10 mm in 20-24 weeks gestation): 10-15% are obstructed at neonatal evaluation
    - Oligohydramnios, polyhydramnios, or normal fluid
  - Postnatal findings
    - Hydronephrosis
    - Large, medial, sonolucent area (renal pelvis) surrounded by smaller, rounded, sonolucent areas (dilated calyces)
    - Advanced parenchymal atrophy in severe cases
    - Hypertrophy of normal kidney contralateral to hydronephrotic kidney

Nuclear Medicine Findings

- Diuretic renography
  - Tc-99m-MAG3 (mercaptoacetyltriglycine) as preferred agent
  - Separates obstructive from nonobstructive dilatation
  - Localize level of obstruction
  - Assess renal function, often preoperatively

Imaging Recommendations

- Best imaging tool
  - Neonates and children: Ultrasonography
  - Adults: CT urography and CTA ± diuresis renography
- Protocol advice
  - CTA and CT urography: Multiplane displays to better define vessels and their relation to UPJ (preoperative evaluation)
  - Serial US should be done several days postnatally due to relative neonatal oliguria
  - Diuresis renography: Tc-99m labeled MAG3 is preferred due to lower radiation burden

DIFFERENTIAL DIAGNOSIS

Extrarenal Pelvis

- No caliectasis, delayed excretion, or renal atrophy
- Diuresis renography rules out delayed excretion and decreased function

Retrocaval Ureter

- Ureter courses behind and medial to inferior vena cava
- Obstruction at mid-ureter → dilatation of proximal ureter and pelvis

Urolithiasis (Renal Calculi)

- Dense ureteral filling defects

- Easily detected on CT

Tumor

- Benign or malignant retroperitoneal mass may cause ureteral obstruction; use CT to evaluate
- Primary transitional cell carcinoma near UPJ may cause obstruction

Renal Pelvis Blood Clot

- e.g., anticoagulated patient, trauma
- May obstruct UPJ

PATHOLOGY

General Features

- Etiology
  - Congenital (most common)
    - **Intrinsic stenosis due to collagen or muscle abnormality**
      - Most likely cause (controversial)
    - Adynamic segment
    - Valves, folds, kinks are probably secondary findings, not causative
    - High "insertion" of ureter
    - Adhesions, bands, and crossing vessels are probably not causative
  - Acquired
    - Scarring: Prior surgery, trauma, calculi
    - Vesicoureteral reflux
    - Malignant neoplasm: Transitional cell carcinoma, squamous cell carcinoma, metastasis
    - Benign neoplasm: Polyp, mesodermal tumor
    - Intraluminal lesion: Stone, clot, sloughed papilla, fungus ball, etc.
    - Herniation of kidney: Through lumbar hernia or Bochdalek hernia
- Genetics
  - Familial occurrences in some cases
- Associated abnormalities
  - Cystic renal dysplasia, primary megaureter
  - Lower or upper segment of duplex kidney
  - Ectopic, malrotated, pelvic, and horseshoe-shaped kidneys
  - Complex congenital anomaly: VATERL (vertebral, anus, tracheoesophageal, renal, and radial)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Neonates
    - Asymptomatic, diagnosed by prenatal screening
    - Palpable, sometimes visible abdominal mass
  - Children and adults
    - Intermittent abdominal or flank pain, nausea, vomiting
    - Hematuria, renovascular hypertension (rare)
- Lab data: Microhematuria, pyuria, urinary tract infection

Demographics

- Age
  - Any age; less commonly presents in adults
- Gender
  - Overall, M:F = 2:1
  - In infants, M:F = 5:1
- Epidemiology
  - Causes 40% of all significant neonatal hydronephrosis (1:500 pregnancies)

Natural History & Prognosis

- Complications include failure to thrive, renal insufficiency, urinary tract infection, urolithiasis, gastroduodenal obstruction, traumatic or spontaneous kidney rupture
- Prognosis: Good, after treating unilateral obstruction

Treatment

- Indicated when patient has symptoms, stones, infection, or when renal function is impaired or at risk
  - Open pyeloplasty
  - Endoscopic or laparoscopic pyeloplasty (pyelotomy)

DIAGNOSTIC CHECKLIST

Consider

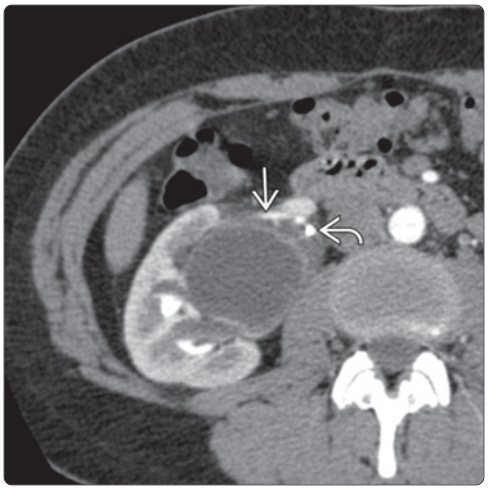
- CT and MR urography comprehensively shows pelvocalyceal anatomy and any potentially associated abnormalities

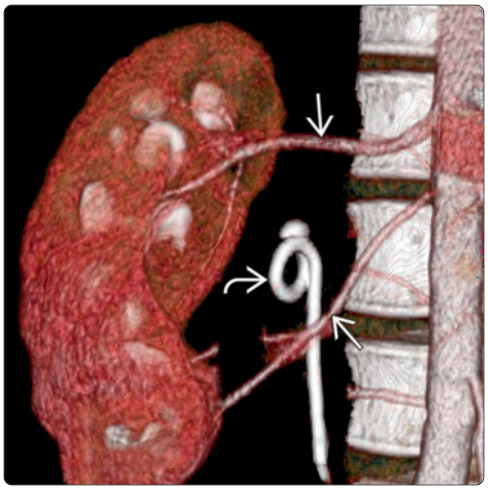
Image Interpretation Pearls

- CT and MR defines altered morphology and etiologies
- Diuretic renogram defines function

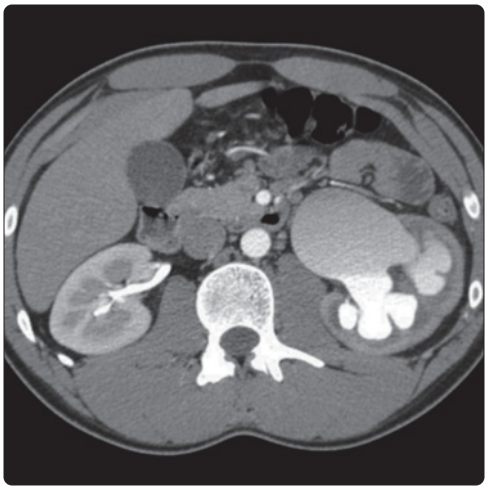
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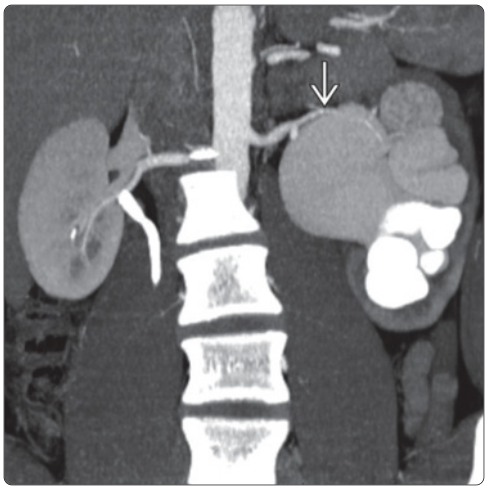
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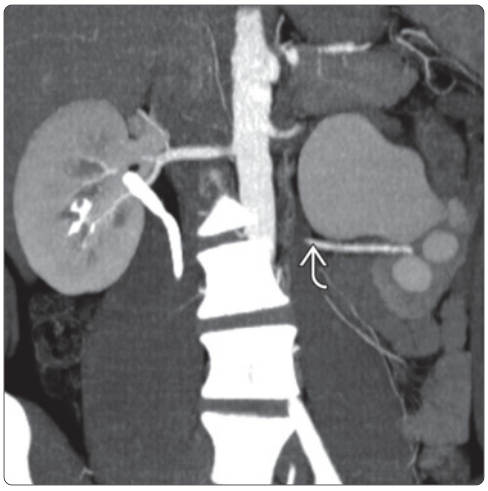


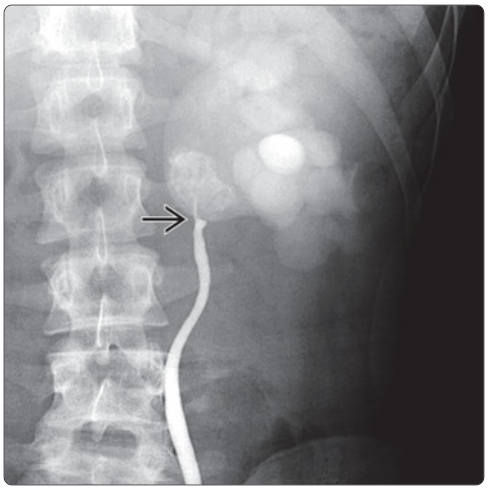
(Left) *Axial CECT section shows the dilated renal calyces and pelvis with a ureteral stent in place. The relationship of the dilated renal pelvis to a renal artery branch is shown in this plane.* **(Right)** *Volume rendered 3D CT in the same patient shows the spatial relationship between renal arteries and renal pelvis. This display is important in evaluating patients for possible endoureteral surgery, which may be contraindicated if the UPJ abuts arterial branches.*





(Left) *Axial CECT shows marked left renal hydronephrosis, a delayed nephrogram, and slight cortical thinning. The left ureter was normal, and no intrinsic or extrinsic masses or obstructing processes were identified. The relationship among the renal hilar structures is not displayed optimally.* **(Right)** *Coronal MIP CECT in the same patient demonstrates left side hydronephrosis and a renal arterial branch to the upper pole.*





(Left) *Coronal MIP CT in the same patient shows an "accessory" lower pole renal artery that appears to lie close to the UPJ. Vessels are rarely the cause of UPJ obstruction, but their presence must be considered when planning for surgical or endoluminal repairs to the UPJ.* **(Right)** *Retrograde pyelogram in the same patient shows an abrupt transition from normal-caliber ureter to the dilated renal pelvis at the UPJ. Note the dilution of injected contrast as it streams into the pelvis as a jet.*

Congenital Megacalyces and Megaureter

KEY FACTS

TERMINOLOGY

- Megacalyces: Dilatation of calyces with normal size renal pelvis and ureter
- Megaureter: Enlarged ureter (diameter > 7 mm)
  - Primary obstructed megaureter: Dilatation due to functional obstruction at ureterovesical junction (absent peristalsis)

IMAGING

- Megacalyces
  - Caliectasis, normal renal pelvis, no evidence of obstruction
  - Narrow, crescent-shaped medulla; absent papillae
  - Numerous calyces, polygonal (not usual cup shape)
- Megaureter
  - Dilated ureter (> 7 mm) tapers to normal caliber just proximal to bladder
  - Left side more common
  - Varying degrees of hydronephrosis

TOP DIFFERENTIAL DIAGNOSES

- Obstructive hydronephrosis
- Postobstructive atrophy
- Vesicoureteral reflux
- Renal papillary necrosis

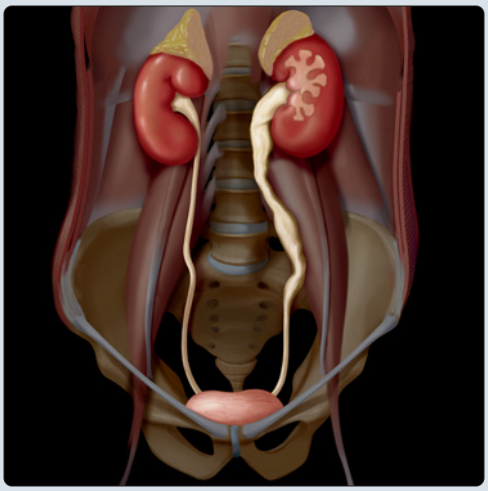
PATHOLOGY

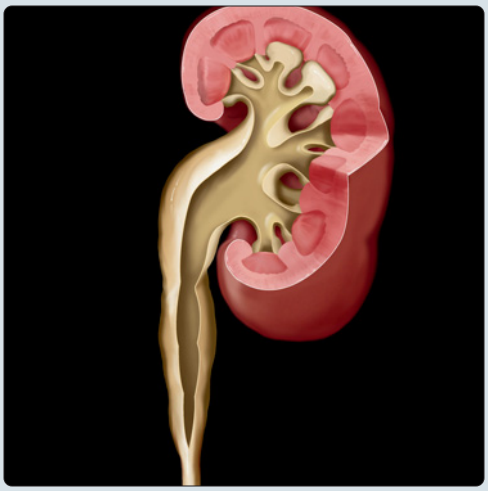
- Megacalyces
  - Hypoplastic medullary pyramids
- Megaureter
  - Adynamic distal ureteral segment, which has normal caliber (primary obstructed)

CLINICAL ISSUES

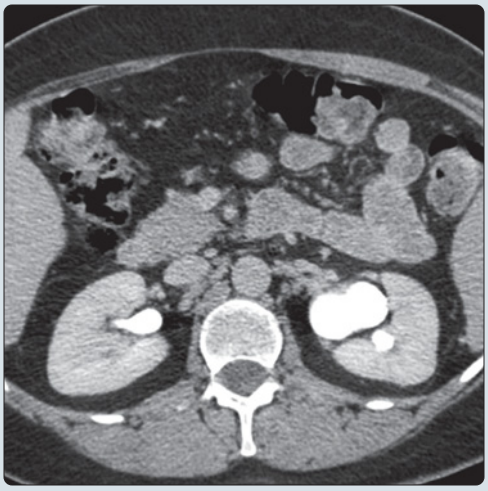
- Ranges from mild stable ureterectasis to severe progressive hydroureteronephrosis
- May even improve with time (10-30% of cases)
- Prophylactic antibiotic is indicated in children with urinary stasis in upper ureter and kidney

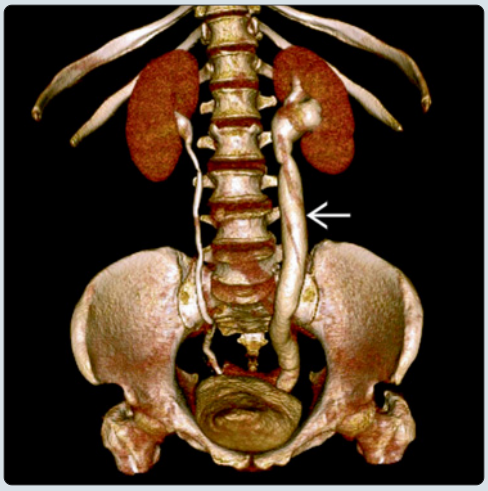
(Left) *Graphic shows a left-sided megaureter with gross dilation of the proximal 2/3 of the ureter and moderate caliectasis. Renal morphology and function are nearly normal. By definition, there is no mechanical obstruction of the ureter.* **(Right)** *This graphic shows megacalyces. The calyces and proximal ureter are grossly dilated, and the calyces are increased in number in some cases. There is only mild atrophy of the papillae and renal cortex.*





(Left) *Axial CT obtained in the excretory phase shows a dilated renal pelvis and calyces but normal renal cortical thickness in the left kidney. The right kidney is normal.* **(Right)** *Coronal volume-rendered CECT in the same patient shows pelvocaliectasis and a dilated left ureter ➡ but no renal parenchymal scarring or significantly delayed excretion. No intrinsic or extrinsic obstructing process was found. A radionuclide renogram showed almost normal and equal function in both kidneys.*





TERMINOLOGY

Synonyms

- Megacalyces = megacalycosis

Definitions

- Megacalyces: Dilatation of calyces with normal-sized renal pelvis and ureter
- Megaureter: Enlarged ureter (diameter > 7 mm)
  - Primary obstructed megaureter: Dilatation due to functional obstruction at ureterovesical junction (absent peristalsis)

IMAGING

Radiographic Findings

- IVP
  - Megacalyces
    - Caliectasis, normal renal pelvis, no evidence of obstruction
    - Narrow, crescent-shaped medulla; absent papillae
    - Numerous calyces, polygonal (not usual cup shape)
  - Megaureter
    - Dilated ureter (> 7 mm) tapers to normal caliber just proximal to bladder
    - Left side more common

Ultrasonographic Findings

- Megaureter
  - Dilated ureter tapering in distal portion
  - Varying degrees of hydronephrosis

CT Findings

- Dilated ureter with curved distal narrowing
- Varying degrees of hydronephrosis
- Best appreciated on CT urography

Nuclear Medicine Findings

- Tc-99m DTPA renography → early stasis pattern, prompt washout after diuresis
- Tc-99m DMSA → normal (symmetric) cortical uptake

DIFFERENTIAL DIAGNOSIS

Obstructive Hydronephrosis

- Both medulla and cortex become atrophic; no polycalyces
- Evidence of obstruction, intrinsic (e.g., stones, tumor) or extrinsic (e.g., retroperitoneal fibrosis or tumor)

Postobstructive Atrophy

- Residual dilatation after chronic obstruction
- History helps to differentiate from megacalyces/ureter

Vesicoureteral Reflux

- Grades 3-5 reflux may cause irreversible dilatation of ureters and calyces
- Tortuous dilated ureter; no "adynamic" juxtavesical segment

Renal Papillary Necrosis

- Usually 1 to several sloughed papillae
- Club-shaped calyces

PATHOLOGY

General Features

- Etiology
  - Megacalyces
    - Hypoplastic medullary pyramids
  - Megaureter
    - Adynamic distal ureteral segment, which has normal caliber (primary obstructed)
    - Excess collagen and fibrosis; deficient or disordered muscle in wall of distal ureter
- Associated abnormalities
  - Megaureter and megacalyces may coexist; medullary sponge kidney, horseshoe kidney, duplex kidney
  - Absent or dysplastic contralateral kidney (10-15%), ipsilateral ureteropelvic junction (UPJ) obstruction (2-8%)

Staging, Grading, & Classification

- Megaureter
  - Primary: Anomaly at vesicoureteral junction
  - Secondary: Abnormality at bladder or urethra

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - May be asymptomatic
  - Urinary tract infections, flank pain
- Clinical profile
  - Normal glomerular filtration rate (GFR), normal renal plasma flow

Demographics

- Age
  - May be diagnosed prenatally (severe megaureter) or as incidental finding in adult
- Gender
  - M:F = 2-6:1

Natural History & Prognosis

- Ranges from mild stable ureterectasis to severe progressive hydroureteronephrosis; rarely progresses to renal insufficiency
  - May even improve with time (10-30% of cases)

Treatment

- Stable cases (no pyelocaliectasis, no symptoms) should be followed up by ultrasound every few years
  - Prophylactic antibiotic is indicated in children with urinary stasis in upper ureter and kidney
- Surgery performed in cases of recurrent UTIs or worsening renal function

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Renal Lymphangiomatosis

KEY FACTS

TERMINOLOGY

- Developmental malformation resulting in cystic mass in perirenal and peripelvic area

IMAGING

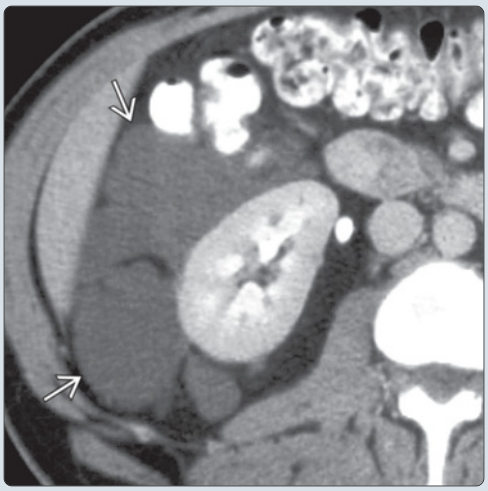
- Uni- or multilocular cystic mass in perirenal or peripelvic area
- Usually bilateral
- Best imaging tool: CT or MR
- CT and MR imaging findings
  - Subcapsular mass with fluid attenuation on CT and high signal intensity on T2WI MR
  - Displaces renal parenchyma and adjacent structures
  - ± peripheral and septal enhancement
  - Calcification may be seen
- Ultrasonographic findings
  - Cystic mass, uni- or multilocular

TOP DIFFERENTIAL DIAGNOSES

- Renal and perirenal abscess
  - Complex fluid collections with enhancing septa at CECT
- Urinoma
  - CECT findings: Fluid collection filling with contrast on delayed images
- Perinephric hematoma
  - High-attenuation collection; fluid-fluid levels
- Renal cell carcinoma
  - Uni- or multilocular cystic mass with thick wall
  - Enhancing, smooth or nodular septa on CECT
- Perinephric lymphoma
  - CECT findings: Soft tissue perinephric mass with mild homogeneous enhancement

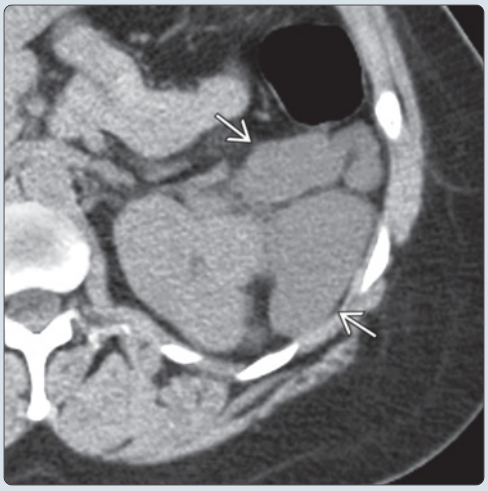
PATHOLOGY

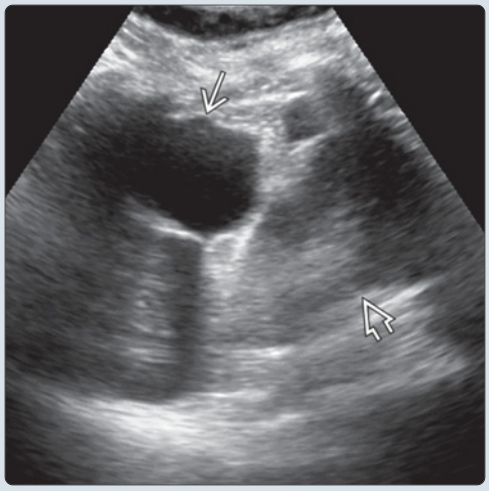
- Failure of development of normal lymphatic tissue in perirenal &/or peripelvic area





(Left) *Axial CECT shows a fluid-attenuation multilocular mass ➡ within the right perirenal space, abutting the renal capsule. The fluid attenuation and multilocular appearance make lymphangioma the most likely diagnosis.* **(Right)** *Gross photograph from the same patient shows multiple, well-defined cysts, correlating with the CT appearance and compatible with renal lymphangiomatosis.*





(Left) *Axial noncontrast CT shows multiple low-attenuation lesions ➡ in the left perinephric space.* **(Right)** *Transverse US of the left kidney ➡ in the same patient shows that the lesions ➡ in the left perinephric space are anechoic, thus confirming the cystic nature and a diagnosis of renal lymphangiomatosis.*

TERMINOLOGY

Synonyms

- Renal lymphangioma, hygroma renalis, renal lymphangiectasia

Definitions

- Developmental malformation of lymphatics resulting in perirenal and peripelvic cystic lesions

IMAGING

General Features

- Best diagnostic clue
  - Multilocular cystic mass in perirenal area with thin septa
- Location
  - Perirenal, peripelvic; usually bilateral
- Size
  - Variable, can become very large
- Morphology
  - Uni- or multilocular cystic mass

Imaging Recommendations

- Best imaging tool
  - CT or MR

CT Findings

- Perinephric fluid attenuation mass
- Displaces renal parenchyma and adjacent structures
- Honeycombing may be seen
- Unilocular or multilocular
- ± peripheral and septal enhancement
- Occasionally calcification may be seen

MR Findings

- T1WI: Usually low signal intensity
  - ↑ signal intensity on T1WI is seen when ↑ proteinaceous material in cyst
- T2WI: High signal intensity

Ultrasonographic Findings

- Anechoic, cystic mass
- Unilocular or multilocular
- Fine internal echoes can be seen with debris

DIFFERENTIAL DIAGNOSIS

Renal and Perirenal Abscess

- Usually presenting with fever, flank pain, leukocytosis
- Complex fluid collections with enhancing septa at CECT and MR

Urinoma

- CECT findings: Fluid collection filling with contrast on delayed images

Perinephric Hematoma

- High-attenuation collection; fluid-fluid levels

Renal Cell Carcinoma

- Cystic renal cell carcinoma
- Uni- or multilocular cystic mass with thick wall
- Calcification of septa or tumor capsule
- Enhancing, smooth or nodular septa on CECT
- Diffuse hypoechoic mass (hemorrhage and necrosis) with fluid-debris levels on ultrasound

Perinephric Lymphoma

- US findings: Solid, hypoechoic mass in the perinephric space
- CECT findings: Soft tissue perinephric mass with mild homogeneous enhancement
- MR findings: Low signal intensity on T1WI and T2WI

PATHOLOGY

General Features

- Etiology
  - Failure of development of normal lymphatic tissue in perirenal &/or peripelvic area resulting in dilatation of lymphatic channels

Gross Pathologic & Surgical Features

- Contains clear fluid with high globulin and lymphocyte content

Microscopic Features

- Thin-walled cysts lined by single layer of endothelium

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic, hypertension, obstructive uropathy
- Other signs/symptoms
  - Dull flank pain, palpable mass, hematuria

Demographics

- Seen in any age
- Same incidence in men and women

Natural History & Prognosis

- ↑ in size during pregnancy with ↑ risk of rupture

Treatment

- Conservative management in asymptomatic cases
- Percutaneous drainage
- Injection of sclerosing agents
- Resection of mass

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Acute Pyelonephritis

KEY FACTS

TERMINOLOGY

- Acute infection of renal parenchyma

IMAGING

- Best diagnostic clue: Wedge-shaped, striated areas of decreased enhancement and renal swelling on CECT
- CT findings
  - Wedge-shaped or rounded areas of poor/streaky enhancement
    - Best identified on nephrographic &/or excretory phase
    - Inflammatory changes in perirenal fat
- MR findings
  - ↑ signal intensity on T2WI and DWI of affected areas
  - Wedge-shaped or rounded areas of poor/streaky enhancement
- Ultrasound findings
  - Often negative
  - ↓ perfusion in areas of pyelonephritis at color Doppler and power Doppler evaluation

TOP DIFFERENTIAL DIAGNOSES

- Renal infarction
- Renal cell carcinoma
- Renal lymphoma
- Vasculitis

PATHOLOGY

- Ascending infection (most common)
  - *Escherichia coli* (*E. coli*)

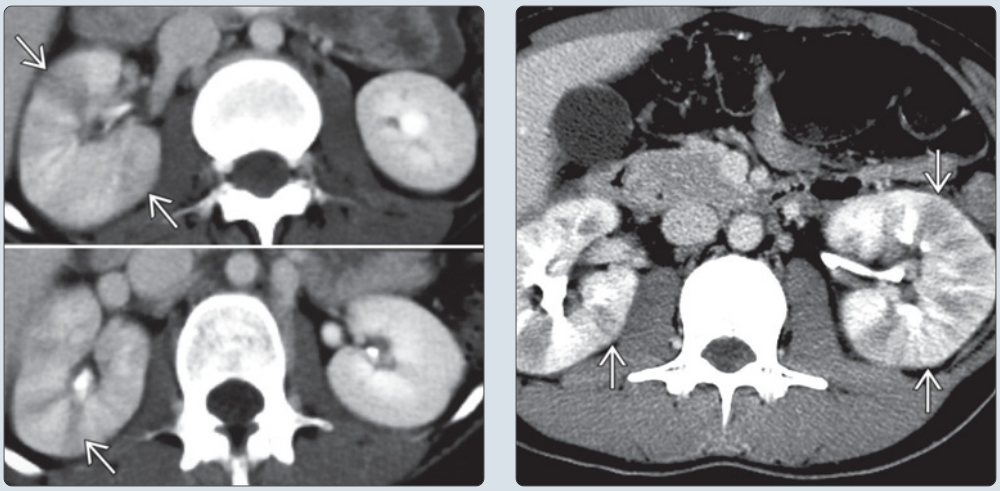
CLINICAL ISSUES

- ↑ incidence in women < 40 years and men > 65 years
- Fever, flank pain, costovertebral angle pain, nausea, vomit

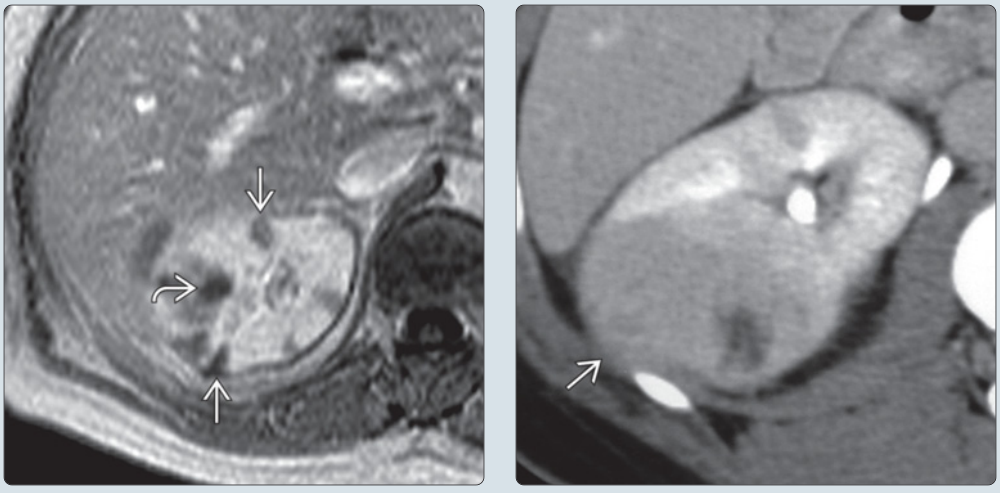
DIAGNOSTIC CHECKLIST

- Diagnosis of acute pyelonephritis is usually clinical, based on combination of clinical and laboratory presentation

(Left) *Axial CECT obtained during the excretory phase shows a swollen right kidney with a "striated nephrogram" ➡. (Right) Axial CECT shows bilateral wedge-shaped areas of renal parenchymal hypoenhancement ("striated nephrograms") from pyelonephritis ➡.*



(Left) *Axial gadolinium-enhanced T1WI MR through the right kidney shows multiple wedge-shaped areas of hypoenhancement ➡ compatible with pyelonephritis. Small intraparenchymal round focus of hypoenhancement suggests a developing microabscess ➡. (Right) Axial CECT shows focal severe pyelonephritis with mass-like swelling and a striated nephrogram ➡.*



TERMINOLOGY

Definitions

- Acute infection of renal parenchyma

IMAGING

General Features

- Best diagnostic clue
  - Wedge-shaped, striated areas of decreased enhancement and renal swelling on CECT
- Morphology
  - The affected area is typically wedge-shaped; can be rounded and mass-like

CT Findings

- CECT
  - Localized or generalized swelling
  - Unilateral or bilateral (less common)
  - Wedge-shaped or rounded areas of poor or streaky enhancement
    - Best identified on nephrographic and excretory phase
    - Small areas of pyelonephritis may be missed on corticomedullary phase images
  - Occasionally mass-like; may distort normal renal contour
  - Findings may persist after clinical improvement
  - Perinephric inflammatory changes
    - Thickening of Gerota fascia
    - Perinephric fat stranding and fluid
    - Renal, perirenal abscess

MR Findings

- T2WI
  - ↑ signal intensity in affected areas of renal parenchyma
  - Perinephric inflammatory changes
- DWI
  - ↑ signal intensity on images acquired with high b-value, and ↓ signal intensity on ADC map
- T1WI C+
  - ↓ enhancement with streaky appearance, similar to CECT

Ultrasonographic Findings

- Grayscale ultrasound
  - Localized or generalized swelling
  - Unilateral renal enlargement may be only indicator
  - Poor corticomedullary differentiation
  - Focal areas of ↑ or ↓ echogenicity
  - No significant findings in > 50% of cases
- Color Doppler
  - ↓ perfusion in areas of pyelonephritis
- Power Doppler
  - ↓ perfusion in areas of pyelonephritis

Nuclear Medicine Findings

- Renal scintigraphy
  - Tc-99m-DMSA (dimercaptosuccinic acid)
  - ↓ accumulation of renal cortical agents in wedge-shaped distribution pointing toward renal hilum
  - Difficult to differentiate acute lesions from prior scars
  - Pinhole collimation, SPECT imaging improve diagnostic sensitivity and accuracy

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or gadolinium-enhanced MR
- Protocol advice
  - Striated nephrogram appears more conspicuous on CT images acquired during the nephrographic (60-100 seconds post contrast injection) or excretory phase (2-3 minutes post injection)

DIFFERENTIAL DIAGNOSIS

Renal Infarction

- Wedge-shaped pattern of absent perfusion
- Cortical rim sign on CECT (enhancement from capsular artery)
  - Emboli, vasculitis, trauma, aortic dissection, atherosclerosis

Renal Cell Carcinoma

- Enhancing mass bulging the renal contour
- Invasion of perirenal space, renal vein
- Retroperitoneal lymphadenopathy

Renal Lymphoma

- Multiple imaging presentations
- Multiple low attenuation renal masses
- More commonly bilateral
- Lymphadenopathy

Vasculitis

- PAN, SLE, scleroderma, drug abuse

PATHOLOGY

General Features

- Etiology
  - Ascending infection (most common)
    - Risk factors
      - Vesicoureteral reflux
      - Neuropathic bladder
      - Prostatic hypertrophy
    - Pathogens
      - *Escherichia coli* (*E. coli*) (> 80%)
      - Others: *Klebsiella*, *Proteus mirabilis*, *Enterococcus faecalis*
  - Hematogenous infection
    - Risk factors
      - Immunosuppression, IV drug abuse
    - Pathogens
      - *Staphylococcus aureus*, *E. coli*
- Associated abnormalities
  - Benign prostatic hypertrophy, vesicoureteral reflux, urinary tract obstruction

Gross Pathologic & Surgical Features

- Enlarged kidney
- Suppurative abscesses visible on cortical surface

Microscopic Features

- Tubulointerstitial nephritis
- Abundant neutrophilic infiltrate
- Patchy distribution

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, flank pain, costovertebral angle pain, nausea, vomiting, dysuria
- Laboratory studies
  - Leukocytosis
  - Occasionally positive blood cultures
  - Urine analysis
    - Pyuria
  - Urine culture
    - Positive urine culture (24%)
    - *E. coli* most common

Demographics

- Gender
  - M:F = 1:5
  - ↑ incidence in women < 40 years, men > 65 years
- Epidemiology
  - Incidence: 250,000 cases/year in USA
  - Hospitalization: 100,000/year
  - Higher incidence in obstruction, duplicated kidneys, other anomalies

Natural History & Prognosis

- Generally excellent prognosis unless complications or recurrent infections
- Complications
  - Renal abscess
  - Perirenal abscess
  - Renal scarring
  - Recurrent infections
  - End-stage renal disease

Treatment

- Antibiotic therapy

DIAGNOSTIC CHECKLIST

Consider

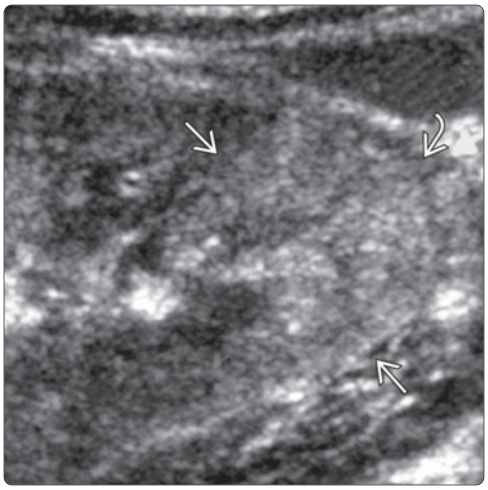
- Diagnosis of acute pyelonephritis is usually clinical, based on a combination of clinical and laboratory presentation
- Imaging for atypical presentation, complicated cases, lack of response to antibiotic treatment
- Distinction of pyelonephritis from vasculitis or renal infarction often requires clinical correlation
- Exclude underlying congenital anomaly in children

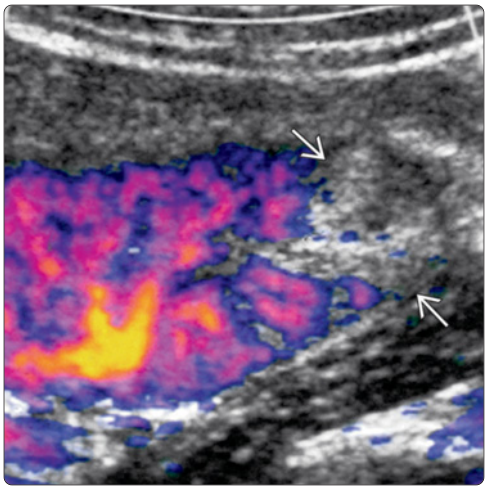
Image Interpretation Pearls

- Contrast-enhanced CT and gadolinium-enhanced MR: Lesion may be poorly visualized if scanned too early (corticomedullary phase)
  - "Striated nephrogram" better visualized on nephrographic &/or excretory phase of enhancement
  - Hypoenhancing areas may be poorly visualized on corticomedullary phase

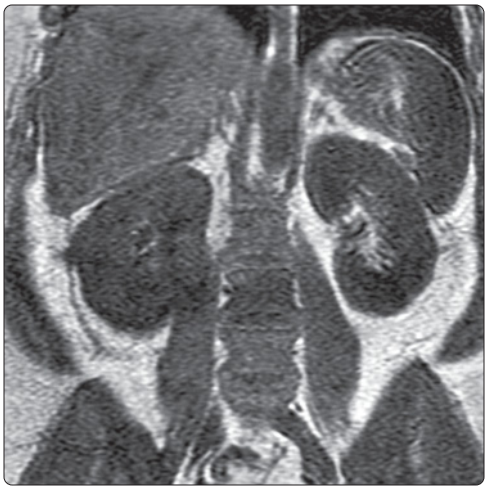
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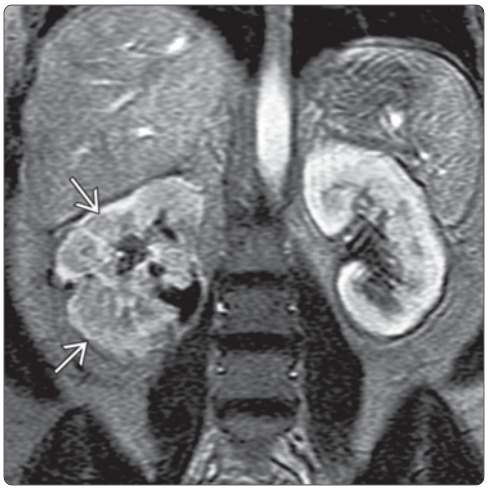
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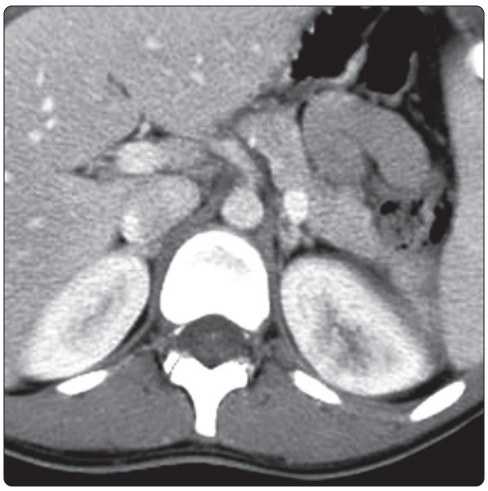


(Left) *Sagittal grayscale sonogram of acute hemorrhagic pyelonephritis shows the wedge-shaped area of increased echogenicity within the lower pole* ⇒ *and the thin rim of adjacent perinephric fluid* ⇒. **(Right)** *Sagittal power Doppler ultrasound in the same patient demonstrates absence of flow in the area of pyelonephritis* ⇒.





(Left) *Coronal T1WI MR of the kidneys demonstrates no clear abnormality.* **(Right)** *Coronal gadolinium-enhanced T1WI MR in the same patient reveals diminished perfusion throughout the right kidney from severe pyelonephritis* ⇒.





(Left) *Axial CECT of the left kidney during corticomedullary phase demonstrates no abnormality.* **(Right)** *Axial CECT in the same patient during excretory phase shows decreased perfusion to the upper pole of the left kidney from pyelonephritis* ⇒.

Chronic Pyelonephritis/Reflux Nephropathy

KEY FACTS

TERMINOLOGY

- Renal scarring and shrinkage secondary to multiple episodes of acute pyelonephritis during early childhood
  - Most cases are secondary to vesicoureteral reflux

IMAGING

- Deep cortical scars overlying deformed calyces
- Scarring is often seen in poles of kidney
  - Upper pole > lower pole
- Typically asymmetric, segmental, and unilateral
- Hypertrophy of residual normal tissue
  - Can mimic mass
- Blunting of calyces
  - Secondary to retraction of papilla
- Dystrophic calcification
- Compensatory hypertrophy of contralateral kidney when unilateral

TOP DIFFERENTIAL DIAGNOSES

- Renal tuberculosis
- Renal infarction
- Radiation nephritis
- Chronic renal failure

PATHOLOGY

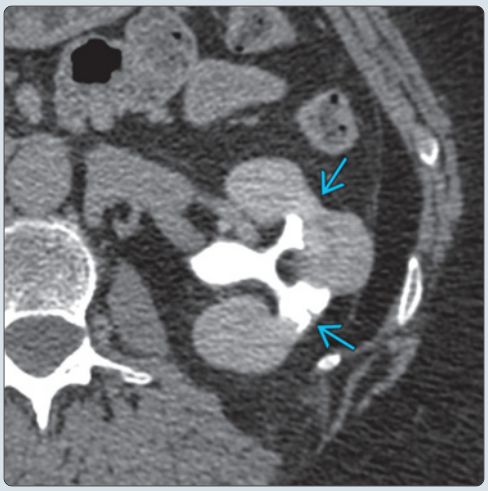
- Etiology: Reflux of infected urine from bladder > urinary obstruction, calculi, foreign body
- In some cases, disease may progress in absence of any bacteriuria or obstruction
- Acute pyelonephritis in adults resolves and does not lead to scarring

CLINICAL ISSUES

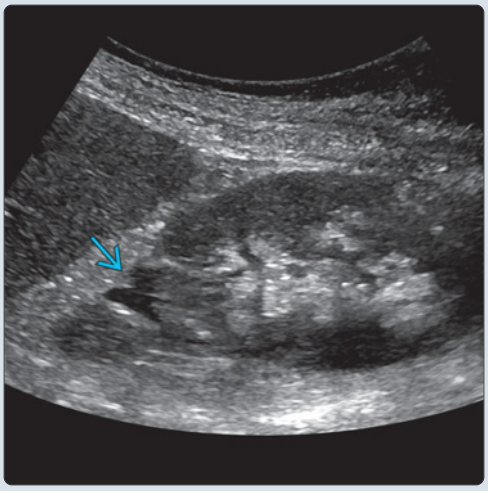
- Time of resolution of VUR depends on grade
- Management of VUR includes prophylactic antibiotics and surgery

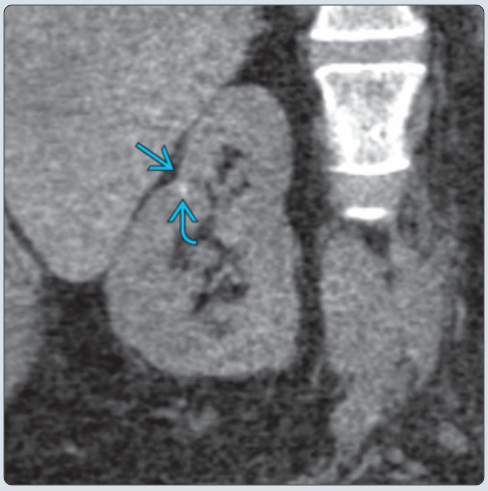
(Left) *Graphic shows the grading system for vesicoureteric reflux, showing progressive levels of reflux, calyceal blunting, and renal cortical injury from grade 1 on the left to grade 5 on the right.* **(Right)** *Axial CECT obtained during the excretory phase shows marked and deep cortical scarring consistent with chronic pyelonephritis.*





(Left) *Longitudinal ultrasound of the right kidney shows deep cortical scar associated with deformed and dilated calyx in the upper pole.* **(Right)** *Coronal NECT in the same patient shows cortical scar in the upper pole of the right kidney associated with small calcification. In chronic pyelonephritis scarring typically affects the poles of the kidney (upper > lower).*





TERMINOLOGY

Synonyms

- Chronic atrophic pyelonephritis

Definitions

- Renal scarring and shrinkage secondary to multiple episodes of acute pyelonephritis during early childhood
  - Most cases secondary to vesicoureteral reflux (VUR)

IMAGING

General Features

- Best diagnostic clue
  - Deep cortical scars overlying deformed calyces, typically in upper pole
- Location
  - Scarring typically in poles of kidney (upper > lower)
- Morphology
  - Typically asymmetric, segmental, and unilateral
  - Compensatory hypertrophy of contralateral kidney when unilateral

Radiographic Findings

- Voiding cystourethrogram
  - Useful in children for grading reflux

CT Findings

- Segmental thinning of cortex and medulla (typically in poles)
- Hypertrophy of residual normal tissue (can mimic mass)
- Blunting of calyces (secondary to retraction of papilla)
- Dystrophic calcification

Ultrasonographic Findings

- Small kidney with segmental loss of parenchyma
- ↑ echogenicity of scarred area
- Extension of central renal sinus echoes to periphery in areas of scarring

DIFFERENTIAL DIAGNOSIS

Renal Tuberculosis

- Multiple cortical scars ± calcification
- Strictures of collecting system and ureter

Renal Infarction

- Localized or global area of ischemic necrosis without caliectasis
- Nonenhancing wedge-shaped area with enhancing cortical rim

Radiation Nephritis

- Linear area of ↓ attenuation corresponding to irradiated portion of kidney
- Global atrophy and nonfunctioning in cases of whole kidney irradiation

Chronic Renal Failure

- Small kidneys; cortical thinning
- Dilatation of collecting system if cause is obstruction

PATHOLOGY

General Features

- Etiology
  - Most cases due to reflux of infected urine from bladder
    - Some cases lack documented infection or reflux
  - Acute pyelonephritis in adults resolves and does not lead to scarring
- Pathophysiology: Proposed mechanisms of injury include
  - Recurrent episodes of acute pyelonephritis
  - Stable changes from remote single episode of acute pyelonephritis

Staging, Grading, & Classification

- Reflux (VUR) grading
  - Grade 1: Reflux into ureter only
  - Grade 2: Reflux into ureter, pelvis, and calyces without dilation
  - Grade 3: Mild dilation of ureter and slight blunting of calyceal fornices
  - Grade 4: Moderate dilation and loss of sharp calyceal fornices
  - Grade 5: Gross dilation of ureter and calyces

Gross Pathologic & Surgical Features

- Segmental deep scars involving both cortex and medulla
- Typically involves poles

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Failure to thrive, flank pain, hypertension, dysuria
  - Rate of progression from VUR to chronic kidney injury and end-stage renal disease is controversial
- Other signs/symptoms
  - Fever, nausea, vomiting

Demographics

- Age: Onset in early childhood (< 4 years old)
- Gender: M:F = 1:2
- Epidemiology: 3x more common in Caucasians

Natural History & Prognosis

- Spontaneous resolution in the majority of cases; time of resolution of VUR depends on grade

Treatment

- Prophylactic antibiotics
- Surgery

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Xanthogranulomatous Pyelonephritis

KEY FACTS

TERMINOLOGY

- Chronic infection of kidney and surrounding tissues characterized by destruction of renal parenchyma and replacement by lipid-laden macrophages

IMAGING

- Best diagnostic clues
  - Enlarged kidney with preserved reniform morphology
  - Low-attenuation renal lesions
  - Diffuse or focally absent nephrogram
  - Centrally obstructing calculus; staghorn
  - Contracted pelvis; dilated calyces
  - Perinephric extension

TOP DIFFERENTIAL DIAGNOSES

- Renal abscess
- Pyonephrosis
- Renal cell carcinoma (RCC)
- Transitional cell carcinoma (TCC)
- Renal metastases and lymphoma

PATHOLOGY

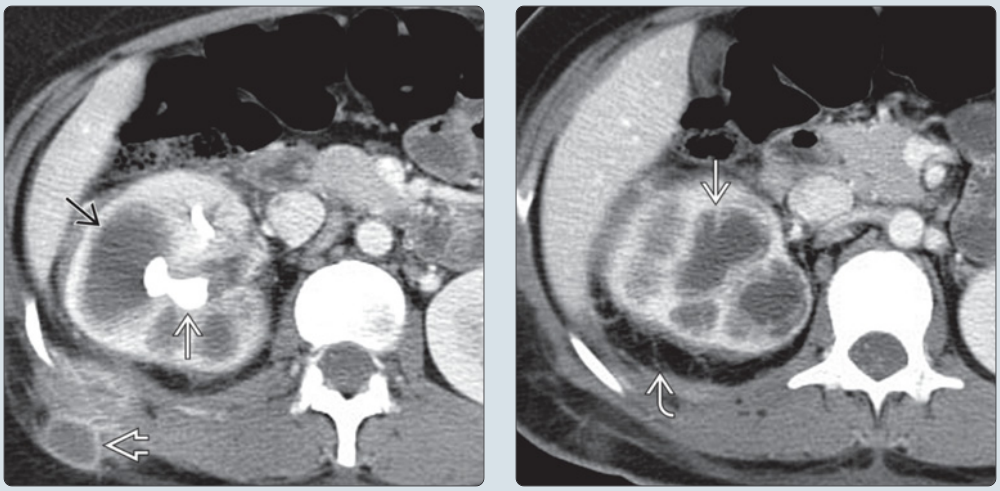
- Accumulation of lipid-laden macrophages (xanthoma cells) and granulomatous infiltrate

CLINICAL ISSUES

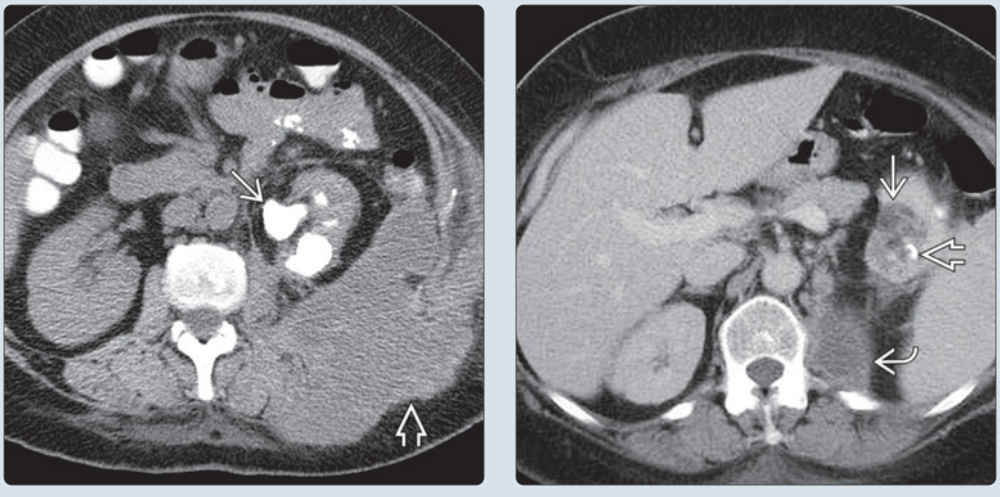
- More frequent in women
- Age: 45-65 years
- Symptoms/signs: Dull flank pain, fever, palpable mass
- Treatment: Nephrectomy usually required
- Good prognosis

DIAGNOSTIC CHECKLIST

- Some variation possible (small kidney, lack of calculi); difficult to distinguish from other infections or neoplasms
- Histologic diagnosis required, not based solely on imaging studies



(Left) *Axial CECT in a 65-year-old woman who presented with pyuria, fever, and right flank pain demonstrates a central stone in the renal pelvis, with surrounding low-attenuation lesions. Note the extension of the inflammatory process through the perirenal space, resulting in an abdominal wall abscess.* **(Right)** *Axial CECT in the same patient reveals perirenal stranding and multiple low-attenuation, necrotic renal lesions corresponding to debris-filled calyces and xanthoma collections.*



(Left) *Axial NECT reveals abscess formation in the setting of XGP. Note the large staghorn calculus in the collecting system of the left kidney with a collection involving the left retroperitoneum, the psoas muscle, and the posterior abdominal wall musculature.* **(Right)** *Axial CECT shows focal areas of low density in posterior aspect of upper pole of left kidney due to focal XGP. Note calculi and large paraspinal/posterior pararenal abscess.*

TERMINOLOGY

Abbreviations

- XGP or XGPN

Definitions

- Chronic infection of kidney and surrounding tissues characterized by destruction of renal parenchyma and replacement by lipid-laden macrophages

IMAGING

General Features

- Best diagnostic clue
  - Enlarged, nonfunctioning kidney
  - Large, central calculus
  - Multiple low-attenuation masses throughout kidney
- Location
  - 2 forms of XGP
    - Diffuse (83-90%)
      - Involves entire kidney
    - Focal or tumefactive (10-17%)
      - Secondary to obstructed single infundibulum; 1 moiety of duplex system
  - Most often unilateral
- Size
  - 2.5-5.8 cm; mean 3.8 cm
- Morphology
  - Well-circumscribed mass with global or focal renal enlargement

Fluoroscopic Findings

- Intravenous pyelography
  - Diffuse or focally absent nephrogram
  - Contracted pelvis; dilated calyces
  - Centrally obstructing calculus; staghorn (75%)
- Retrograde pyelography
  - Complete obstruction at ureteropelvic junction, infundibulum, proximal ureter
  - Contracted renal pelvis; dilated deformed calyces with nodular filling defects
  - Irregular parenchymal masses with cavitation

Ultrasonographic Findings

- Grayscale ultrasound
  - Multiple anechoic or hypoechoic masses
  - Acoustic shadowing from central staghorn calculus
  - Parenchymal thinning and hydronephrosis
  - Echoes in dilated collecting system
  - Perinephric fluid collection
  - Focal XGP
    - Nonspecific findings mimicking abscess and RCC

CT Findings

- NECT
  - Diffuse XGP
    - Enlarged kidney
    - Preserved reniform shape
    - Multiple, low-attenuation masses scattered throughout kidney
    - Large central calculus, often staghorn
    - No pelvic dilatation
    - Obliterated renal sinus fat
  - Focal XGP
    - Low-attenuation mass associated with calculus
- CECT
  - Rim enhancement of collections secondary to inflammation
  - Poor/absent excretion of contrast into collecting system
- Perinephric extension
  - Perirenal space, pararenal space, ipsilateral psoas muscle, colon, spleen, diaphragm, posterior abdominal wall, skin
- Rare findings: Gas, renal atrophy, no calculi

MR Findings

- T1WI
  - Intermediate to high signal intensity of solid component
    - Low signal intensity on fat-suppressed images
  - Low signal intensity of content of collections
- T2WI
  - Intermediate- to high-signal intensity of fluid within collections and dilated collecting system
    - Heterogeneous signal intensity (fluid levels) reflecting debris/pus
  - Defects within dilated collecting system (renal calculi)
- T1WI C+
  - Rim enhancement of collection

Imaging Recommendations

- Best imaging tool
  - MDCT

DIFFERENTIAL DIAGNOSIS

Renal Abscess

- Solitary or multiple, round, well-marginated, low-attenuation mass
- Wall enhancement without central enhancement
- Perinephric stranding
- ± presence of gas

Pyonephrosis

- Pus-filled collecting system may simulate XGP

Renal Cell Carcinoma (RCC)

- Solitary, soft tissue density mass (30-50 HU) with central necrosis
- Hypervascular mass at renal cortex

Transitional Cell Carcinoma (TCC)

- Renal infiltration → calyceal invasion, renal enlargement, poorly defined margins without change in shape
- Renal pelvic filling defect, irregular narrowing of collecting system
- Encasement of pelvicalyceal system → hypovascular tumor

Renal Metastases and Lymphoma

- Metastases: Lung, breast, colon, malignant melanoma
  - Usually hypovascular with infiltrative growth
- Lymphoma
  - Usually multiple or bilateral with infiltrative growth
  - Hypoechoic, hypovascular, solitary, intrarenal mass ± adenopathy

PATHOLOGY

General Features

- Etiology
  - Pathogenesis
    - Chronic renal obstruction and infection with failure of local host immunity
  - Pathogens
    - *Escherichia coli*
    - *Proteus mirabilis*
    - Other: *Staphylococcus aureus, Klebsiella, Pseudomonas*
  - Risk factors
    - Recurrent or chronic urinary tract infections
    - Diabetes, immunocompromised patients
    - Abnormal lipid metabolism

Staging, Grading, & Classification

- 3 stages
  - Stage 1: Confined to kidney
  - Stage 2: Extension to perirenal space
  - Stage 3: Spread to pararenal spaces ± abdominal wall

Gross Pathologic & Surgical Features

- Diffuse
  - Renal enlargement
  - Dilated renal pelvis with staghorn calculus
  - Yellow nodules replacing corticomedullary junction
- Focal
  - Yellowish-white, solid or semisolid renal mass

Microscopic Features

- Lipid-laden macrophages (xanthoma cells)
- Diffuse infiltration by plasma cells and histiocytes
- Peripheral granulation tissue

CLINICAL ISSUES

Presentation

- Symptoms/signs
  - Dull flank pain, fever
  - Palpable mass, weight loss
- Lab data
  - Urinalysis: Microscopic hematuria, proteinuria, pyuria
  - Urine culture
    - Specific bacterial species
    - Negative in up to 40% of cases
  - ↑ erythrocyte sedimentation rate (ESR)
  - Abnormal liver function

Demographics

- Age
  - Most common: 45-65 years
  - Rare in childhood
  - Focal form more common in children
- Gender
  - M:F = 1:3-4
- Epidemiology
  - Incidence: 1% of all renal infections

Natural History & Prognosis

- Symptomatic 6 months prior to diagnosis (40% of cases)
- Complications
  - Extrarenal extension
  - Fistulas (pyelocutaneous, ureterocutaneous)
  - Hemorrhage
  - Hepatic dysfunction (reversible)
- Good prognosis
- Rare mortality, but morbidity can be substantial

Treatment

- Antibiotic treatment prior to surgical intervention
- Nephrectomy usually required
  - Radical in diffuse XGP, partial in focal XGP

DIAGNOSTIC CHECKLIST

Consider

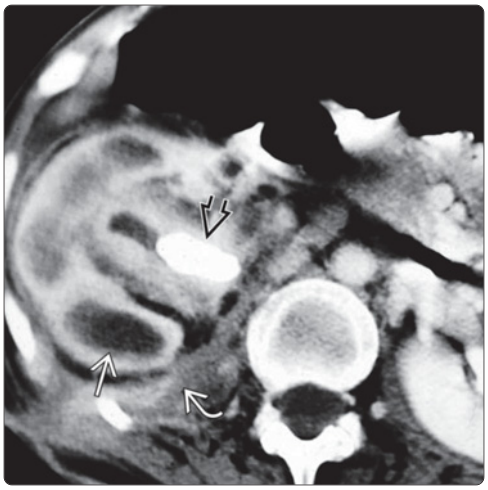
- Some variation possible (small kidney, lack of calculi)
  - Difficult to distinguish from other infections or neoplasms
- Histologic diagnosis required, not based solely on imaging studies

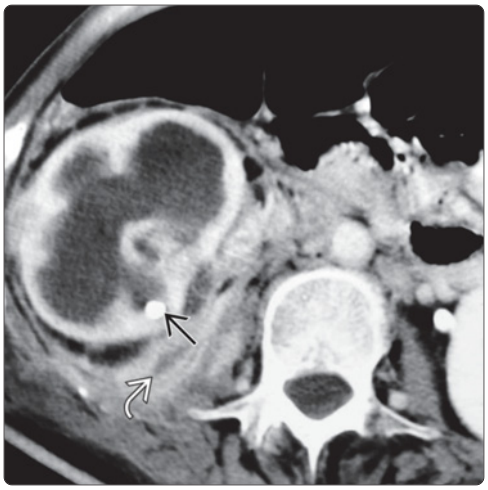
Image Interpretation Pearls

- Enlarged kidney; poor/absent contrast excretion from kidney; large/staghorn calculi

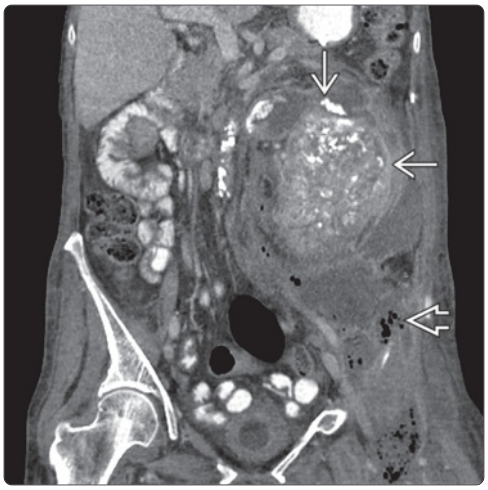
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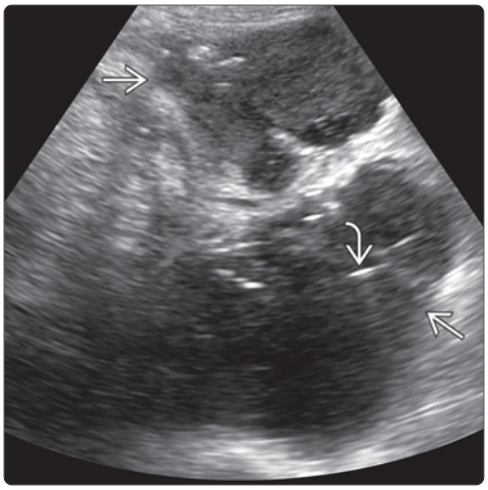
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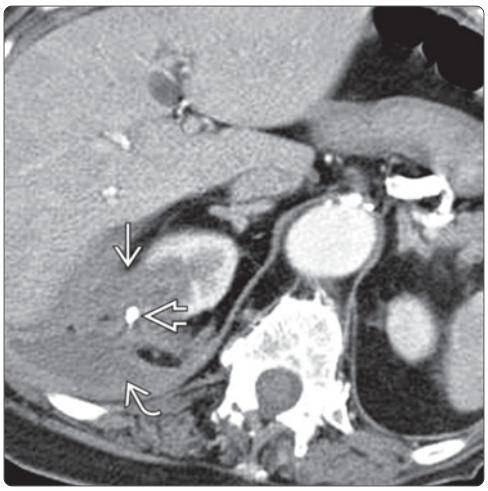


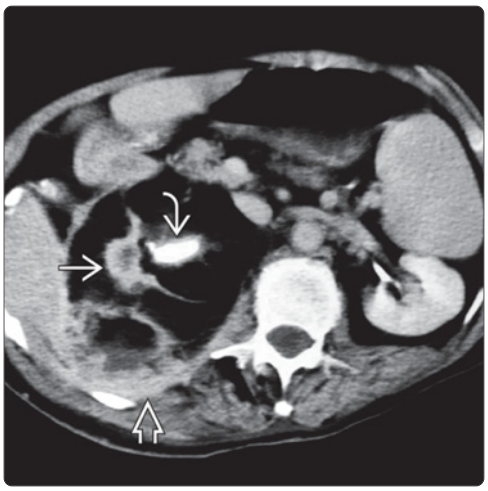
(Left) *Axial CECT in an 81-year-old man who presented with long history of right flank pain demonstrates an obstructed collecting system due to a large central calculus in the renal pelvis. Note the inflammatory changes in the perinephric space.* **(Right)** *Axial CECT in the same patient again illustrates the inflammatory changes in the perinephric space, and smaller renal calculi. The right kidney is enlarged with preserved reniform shape.*





(Left) *Coronal CECT of a case of suspected XGP shows a markedly enlarged and dysmorphic left kidney with multiple obstructing pelvic stones. Note the large perinephric abscess extending to the left iliopsoas muscle.* **(Right)** *Transverse ultrasound in the same patient shows the perinephric collection as hypoechoic with internal fine echoes, and high-amplitude echoes representing gas. Drainage was achieved with ultrasound-guidance. Analysis of drained fluid revealed Proteus mirabilis.*





(Left) *Axial CECT shows a low-attenuation, nonenhancing mass in the upper pole of the right kidney associated with calcification and perinephric inflammatory changes and abscess. Imaging findings compatible with focal XGP.* **(Right)** *Axial CECT demonstrates a large stone in the right renal pelvis and atrophic kidney. Note fat proliferation surrounding the kidney and extension of the inflammatory process through the retroperitoneum and musculature of the back.*

Emphysematous Pyelonephritis

KEY FACTS

TERMINOLOGY

- Emphysematous pyelonephritis (EPN)

IMAGING

- Best imaging tool: CT
- CT findings
  - Intraparenchymal gas ± perirenal extension
  - ± renal or perirenal fluid collections
  - Classification systems (Wang et al.; Huang and Tseng) based on distribution of gas and fluid collections
- Ultrasound findings
  - Nondependent echogenic foci in renal parenchyma &/or collecting system
  - "Dirty shadowing"; ring-down artifacts

TOP DIFFERENTIAL DIAGNOSES

- Emphysematous pyelitis
- Perforated duodenal ulcer
- Iatrogenic

PATHOLOGY

- Most common pathogen
  - (*Escherichia coli*) *E. coli*

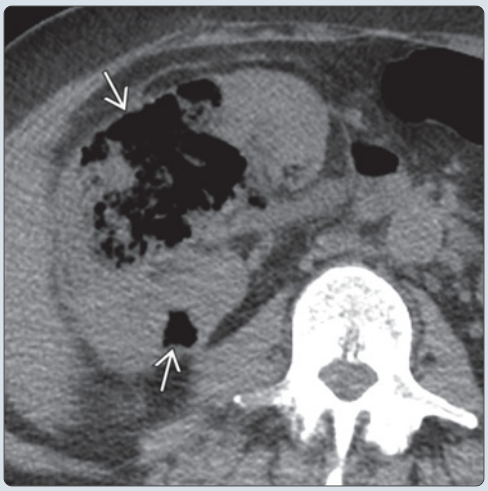
CLINICAL ISSUES

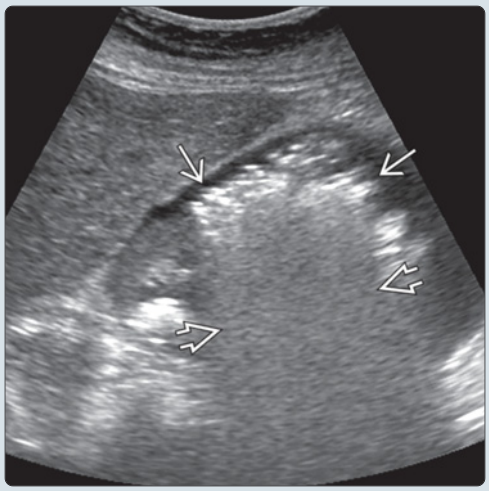
- Life-threatening infection
- Prognosis affected by clinical/lab presentation and gas extension
- Overall mortality: 11-50%
- Highest mortality rate in type 1 (36-69%) and class 4 (50%)
- Treatment
  - Fluid and electrolyte resuscitation; antibiotic therapy
  - Percutaneous drainage
  - Nephrectomy

DIAGNOSTIC CHECKLIST

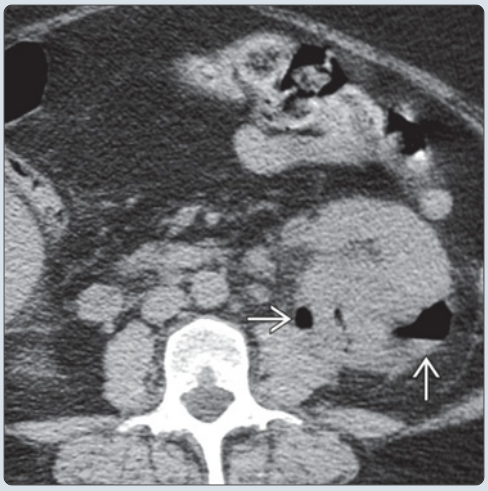
- Best diagnostic clue: Intraparenchymal, intracalyceal, intrapelvic gas on CT

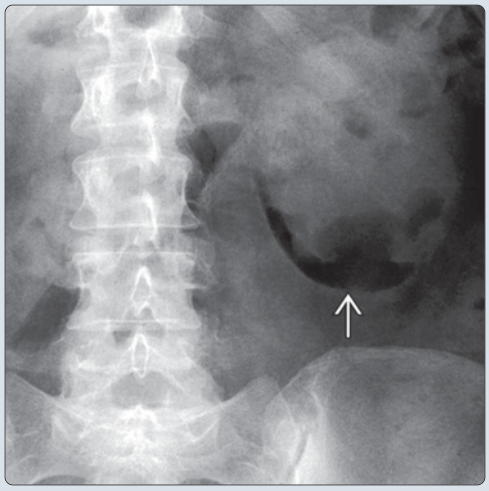
(Left) *Axial NECT shows large amount of gas ⇨ within the right kidney, compatible with emphysematous pyelonephritis. No fluid collections are present. CT is the best imaging modality for diagnosis and assessment of gas extension.* **(Right)** *Sagittal ultrasound of the right kidney in the same patient reveals nondependent echoes within the renal parenchyma ⇨ associated with "dirty shadowing" ⇨.*





(Left) *Axial NECT in a 69-year-old diabetic woman who presented with a fever and flank pain demonstrates large gas-fluid collections in the left renal parenchyma ⇨, diagnostic of a gas-forming abscess from emphysematous pyelonephritis.* **(Right)** *Anteroposterior radiography in the same patient demonstrates a curvilinear gas collection ⇨ outlining the lower pole of the left kidney.*





TERMINOLOGY

Abbreviations

- Emphysematous pyelonephritis (EPN)

Definitions

- Necrotizing, gas-forming renal infection

IMAGING

General Features

- Best diagnostic clue
  - Gas in renal parenchyma on CT

Radiographic Findings

- Radiography
  - Gas in renal parenchyma ± perinephric space

CT Findings

- NECT
  - Intraparenchymal, intracalyceal, intrapelvic renal gas
  - Classification according to Wan et al
    - Type 1: Parenchymal destruction without fluid
      - Streaky/mottled gas in renal parenchyma; ± subcapsular or perinephric gas
    - Type 2: Renal or perirenal fluid collections
      - Bubbly gas pattern; ± gas within renal pelvis
  - Classification according to Huang and Tseng
    - Class 1: Gas only within the collecting system (emphysematous pyelitis)
    - Class 2: Gas limited to the renal parenchyma
    - Class 3A: Gas/abscess in the perinephric space
    - Class 3B: Gas/abscess in the pararenal space
    - Class 4: Bilateral EPN
- CECT
  - Decreased enhancement of affected renal parenchyma

Ultrasonographic Findings

- Grayscale ultrasound
  - Nondependent echogenic foci in renal parenchyma &/or collecting system
  - "Dirty shadowing"; ring-down artifacts

Imaging Recommendations

- Best imaging tool
  - CT for diagnosis and classification

DIFFERENTIAL DIAGNOSIS

Emphysematous Pyelitis

- Gas in renal pelvis and calyces, not parenchyma
- Seen in diabetes (50%) and urinary tract obstruction
- Better prognosis than EPN

Perforated Duodenal Ulcer

- Occasionally gas "outlines" kidney

Iatrogenic

- Diagnostic (e.g., retrograde pyelography, cystoscopy) or therapeutic procedures (e.g., nephrostomy, embolization)

PATHOLOGY

General Features

- Etiology
  - Most common pathogens
    - *Escherichia coli*: 69%
  - Risk factors
    - Diabetes mellitus (> 90%)
    - Urinary tract obstruction

Staging, Grading, & Classification

- Classification schemes based on distribution of gas at imaging

Gross Pathologic & Surgical Features

- Necrotizing renal parenchyma and perirenal tissue; abscesses

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, chills, flank pain, lethargy, confusion
  - Complications: Septic shock
  - Lab data: Hyperglycemia, thrombocytopenia, pyuria

Demographics

- Age
  - Median: 60 years
- Gender
  - More common in women

Natural History & Prognosis

- Life-threatening disease
- Overall mortality: 11-50%
- Highest mortality rate in type 1 (36-69%) and class 4 (50%)

Treatment

- Fluid and electrolyte resuscitation; antibiotic therapy
- Percutaneous drainage
- Nephrectomy

DIAGNOSTIC CHECKLIST

Consider

- CT for diagnosis and management

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Renal Abscess

KEY FACTS

TERMINOLOGY

- Localized collection of pus due to suppurative necrosis in kidney

IMAGING

- Best diagnostic clue: Spherical nonenhancing renal mass with perinephric stranding on CECT
- CT findings
  - ± gas within collection
  - "Rim" or "ring" sign: Enhancement of abscess wall (subacute or chronic)
  - No central enhancement of lesion; enhancement of normal renal tissue

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma (RCC)
- Metastases and lymphoma
- Infected or hemorrhagic cyst

PATHOLOGY

- Urinary tract infection → ascends to kidney → acute pyelonephritis or acute focal bacterial nephritis → liquefaction → sequestration → renal abscess

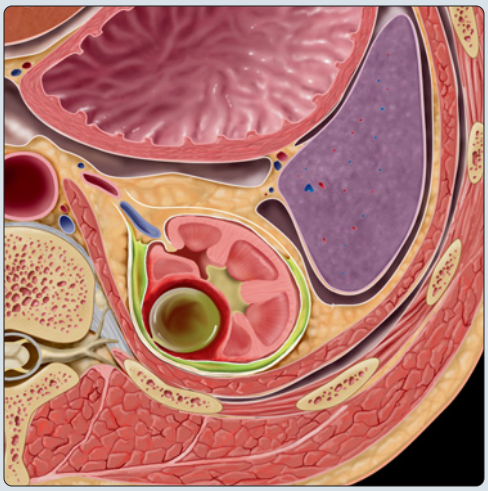
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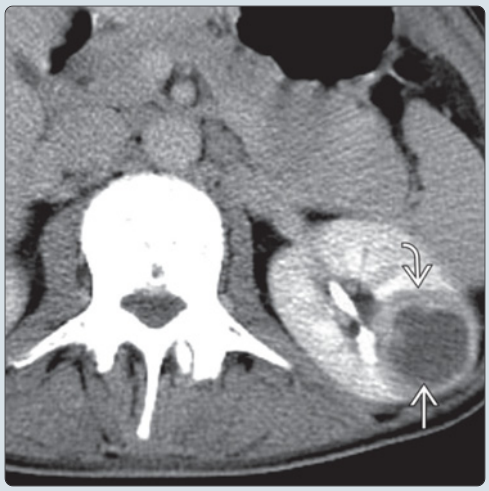
- Most common signs/symptoms: Fever, flank or abdominal pain, chills, dysuria
- Treatment
  - Antibiotic therapy
  - Antibiotic therapy + percutaneous drainage via US or CT
  - Surgery

DIAGNOSTIC CHECKLIST

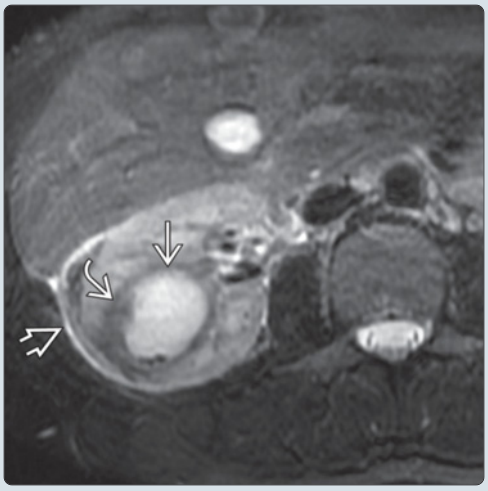
- Consider clinical history and urinalysis to diagnose and differentiate from malignancy
- Imaging pearls: "Rim" sign; absence of lesion enhancement; perinephric stranding

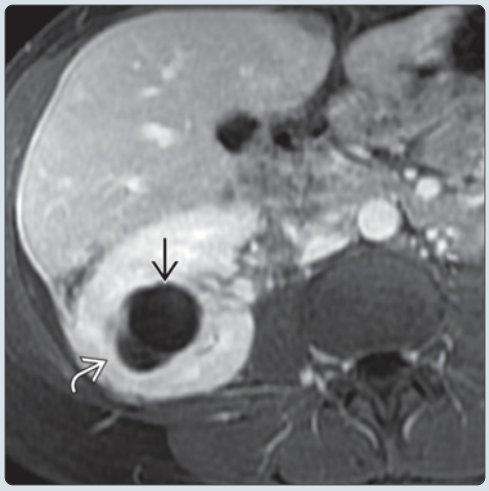
(Left) *Graphic demonstrates a left renal abscess with a thick peripheral rim and adjacent infected perinephric fluid.* **(Right)** *Axial CECT in a 46-year-old woman with a history of diabetes, now presenting with left flank pain and fever, demonstrates a nonenhancing fluid collection in the left kidney compatible with an abscess. Note the thick, irregular wall of the abscess capsule.*





(Left) *Axial T2 FS MR through the right kidney shows a large lesion with a central hyperintense component (fluid) and an irregular, peripheral hypointense rim. Note the rim of perinephric fluid.* **(Right)** *Axial gadolinium-enhanced T1 C+ FS MR through the right kidney in the same patient shows a lesion with no central enhancement and mild peripheral rim enhancement, compatible with abscess.*





TERMINOLOGY

Definitions

- Localized collection of pus secondary to suppurative necrosis in kidney

IMAGING

General Features

- Best diagnostic clue
  - Spherical nonenhancing renal mass with perinephric stranding on CECT

Fluoroscopic Findings

- IVP
  - Impaired excretion
    - Delayed appearance time, ↓ contrast density, ↓ nephrogram
    - ± absence of nephrogram and calyceal opacification
  - Heterogeneous nephrogram
  - Single or multiple, well-defined, round or irregular lucent mass
  - Calyceal or pelvic effacement
  - ± calyceal, pelvic, or ureteral dilatation

Ultrasonographic Findings

- Grayscale ultrasound
  - Anechoic or hypoechoic fluid collection blending with normal echogenic fat within Gerota fascia
  - Mass within or displacing kidney
  - Round, thick- or smooth-walled complex mass
  - ± internal septations or loculations
  - Low-level internal echoes (internal debris)
  - "Ring-down" artifacts, "dirty shadowing"
- Color Doppler
  - Peripheral flow (hyperemia)
  - Central flow absent or within septa

CT Findings

- NECT
  - Single (more common) or multiple; unilateral or bilateral
  - Round, well-marginated, low-attenuation masses
  - ± gas within collection
- CECT
  - Enlarged kidney with focal areas of hypoattenuation (acute)
  - "Rim" or "ring" sign: Enhancement of abscess wall (subacute or chronic)
  - No central enhancement of lesion
  - Normal renal tissue enhances
  - Obliteration of renal sinus or calyceal effacement
  - Thickened walls, mild dilatation of renal pelvis and ureter
  - Perinephric reaction or extension
    - Altered renal contour, indistinct renal outline, renal displacement
    - Edema or obliteration of perinephric fat
    - Thickened Gerota fascia and perinephric septa

MR Findings

- T1WI
  - Hypointense mass
- T2WI
  - Hyperintense mass, ↑ signal surrounding mass (perilesional edema)
- T1WI C+
  - Rim enhancement (lesion < 1 cm may enhance homogeneously)

Nuclear Medicine Findings

- WBC scan
  - ↑ uptake of indium-111 labeled leukocytes noted within renal ± perinephric abscess
  - Possible false-negative leukocyte scans: Prior antibiotic therapy, walled-off abscesses, poorly developed inflammatory response

Imaging Recommendations

- Best imaging tool
  - MDCT before and after contrast to distinguish abscess from tumor

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- CECT: Enhancing mass
- Often incidental finding on CT or US
- Often presenting with metastases
- Symptoms rare, usually asymptomatic
- Clinical history and urinalysis can differentiate

Metastases and Lymphoma

- Metastases (lung, breast, gastrointestinal, malignant melanoma)
  - Almost all develop via hematogenous spread
  - Lung, breast, and colon carcinoma are occasionally large and solitary; difficult to differentiate from RCC
  - Multifocal, small, enhancing nodules on CT; widespread nonrenal metastases
  - Asymptomatic (most common), or flank pain and hematuria
  - CT- or US-guided biopsy for pathologic confirmation
- Lymphoma
  - Multiple patterns: Multiple distinct masses, direct invasion from enlarged retroperitoneal nodes, solitary mass, diffuse infiltration, predominantly perinephric involvement
  - Asymptomatic (most common); fever, weight loss, flank pain, hematuria, renal failure

Infected or Hemorrhagic Cyst

- Solitary, nonenhancing lesion
- CT: Absence of perinephric stranding, "rim" sign, shaggy wall, hyperintense mass
- May be indistinguishable on imaging

PATHOLOGY

General Features

- Etiology
  - Sequelae of acute renal infections
    - Acute pyelonephritis or focal bacterial nephritis
    - Usually seen 1-2 weeks after infection
  - Ascending urinary tract infections
    - Calculi, obstruction, renal anomalies, urinary reflux (diabetes or pregnancy)

  - Iatrogenic intervention (catheterization)
  - Gram-negative organisms (*Escherichia coli*, *Proteus* species, *Klebsiella* species)
- Hematogenous spread
  - IV drug users, skin infection
  - Seeding from other infected sites (valvular heart disease, prosthesis)
  - Iatrogenic intervention (cyst aspiration, embolization of kidney vessels)
  - Gram-positive and gram-negative organisms (*Staphylococcus aureus*, *Streptococcus*, or Enterobacteriaceae)
- Risk factors
  - Diabetes mellitus
  - Immunosuppression
  - Urinary tract obstruction
- Pathogenesis
  - Urinary tract infection → ascends to kidney → acute pyelonephritis/acute focal bacterial nephritis → liquefaction → sequestration → renal abscess

- Associated abnormalities
  - Urinary tract obstruction
  - Urinary tract abnormalities (e.g., VUR) especially in pediatric subjects

Gross Pathologic & Surgical Features

- Well-defined, round, thick- or smooth-walled mass

Microscopic Features

- Infected and necrotic tissue; ± gas

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, flank or abdominal pain, chills, dysuria
  - Symptoms > 2 weeks
  - Costovertebral angle tenderness
  - Palpable flank mass
- Lab data
  - Urinalysis: ↑ WBC, positive bacterial culture
  - Blood tests: ↑ erythrocyte sedimentation rate (ESR), positive bacterial culture

Demographics

- Age
  - All ages
- Gender
  - M = F
- Epidemiology
  - Incidence: Renal abscess (0.2%), perinephric abscess (0.02%)

Natural History & Prognosis

- Complications
  - Rupture into perinephric space (perinephric abscess) → beyond Gerota fascia (paranephric abscess) → psoas or transversalis muscles → anterior peritoneal cavity → subdiaphragmatic or pelvic abscess
  - Rupture → renal collecting system → pyonephrosis
  - Compression or obstruction → hydronephrosis → renal atrophy
  - Necrosis and cavitation
- Prognosis
  - Good if early diagnosis and treatment
  - Poor if delayed diagnosis and treatment

Treatment

- Antibiotic therapy
- Antibiotic therapy + percutaneous drainage via US or CT (depending on abscess size)
- Surgery
  - Failure of medical therapy or drainage
  - Complicated cases not amenable to percutaneous drainage
- Follow-up
  - Imaging to confirm abscess resolution
  - Evaluate for underlying urinary tract abnormalities

DIAGNOSTIC CHECKLIST

Consider

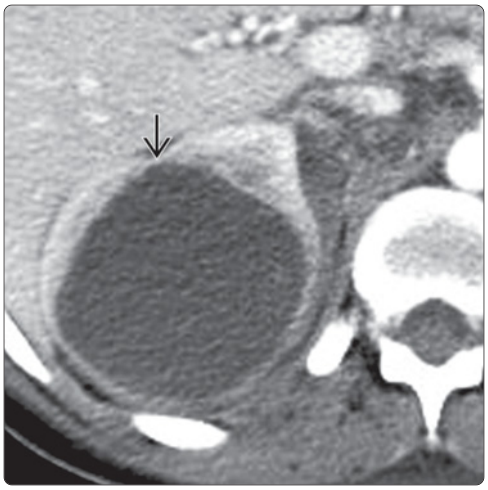
- Clinical history and urinalysis to diagnose and differentiate from malignancy

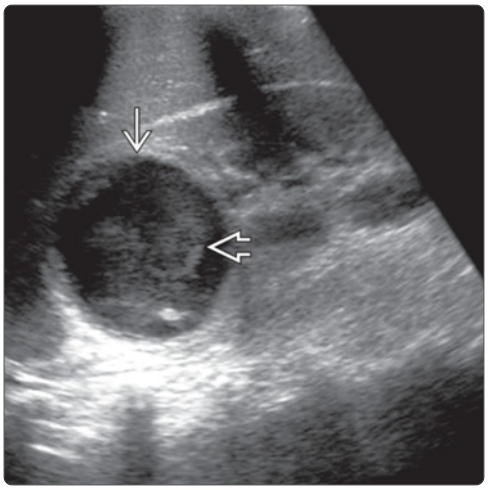
Image Interpretation Pearls

- CECT
  - Low-attenuation lesion with no central enhancement
  - "Rim" enhancement
  - Perinephric stranding

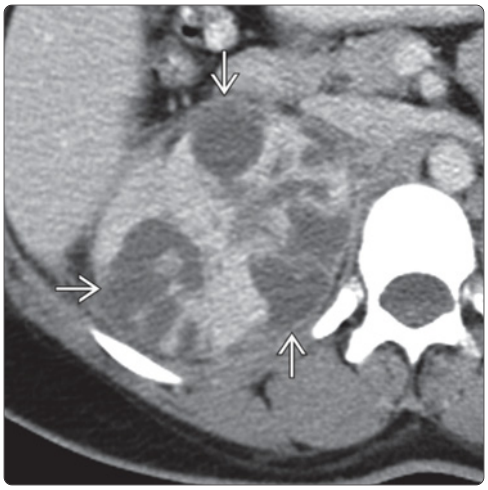
SELECTED REFERENCES

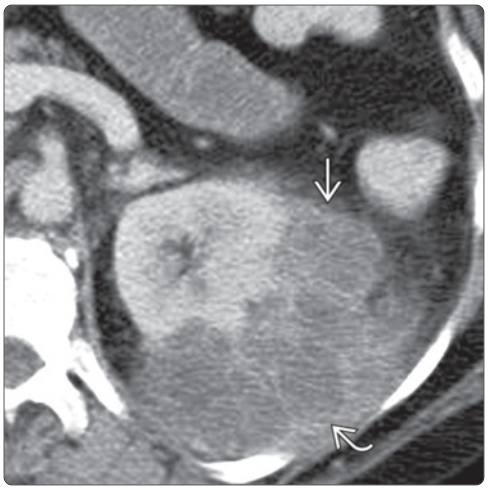
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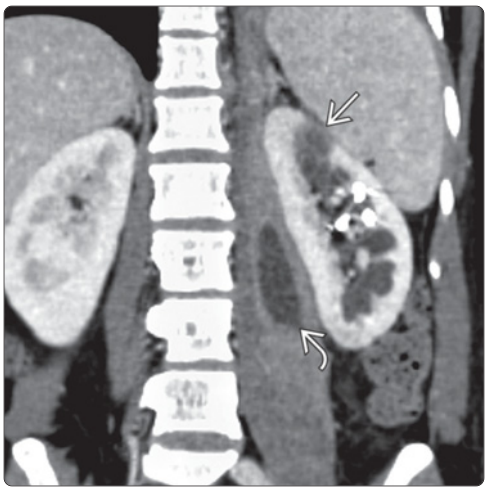


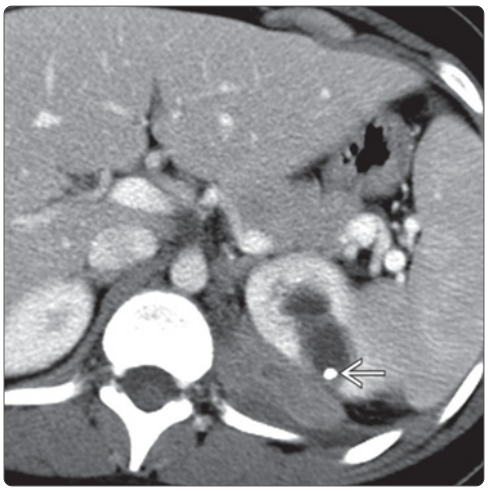
(Left) *Axial CECT through the right kidney shows a large, encapsulated, nonenhancing renal mass ⇨ with an attenuation of 20 HU, suggestive of abscess.* **(Right)** *Sagittal ultrasound of the right kidney in the same patient shows a round, anechoic mass ➡ with low-level internal echoes ➡ and posterior acoustic enhancement. A history of fever and image findings indicate an abscess.*





(Left) *Axial CECT shows an enlarged right kidney with multiple irregular and nonenhancing collections ➡ compatible with multifocal abscesses.* **(Right)** *Axial CECT shows multiloculated, rim-enhancing renal ➡ and perirenal abscesses ➡.*





(Left) *Coronal CECT in a 32-year-old woman, presenting with flank pain, fever, and blood cultures that were positive for E. coli, reveals hematogenous Staphylococcus abscesses in the left kidney ➡ and the left psoas muscle ➡.* **(Right)** *Axial CECT in the same patient shows a percutaneous drainage catheter ➡ within the renal abscess. The patient made a full recovery following drainage.*

Pyonephrosis

KEY FACTS

TERMINOLOGY

- Infection of obstructed renal collecting system

IMAGING

- CT
  - Dilated renal collecting system
  - Fluid-debris level ± gas in renal pelvis
  - Thickened urothelial lining of renal pelvis and proximal ureter
- Ultrasound
  - Dilated renal collecting system
    - Low-level echoes
    - Fluid-debris levels

TOP DIFFERENTIAL DIAGNOSES

- Hydronephrosis
- Transitional cell carcinoma

PATHOLOGY

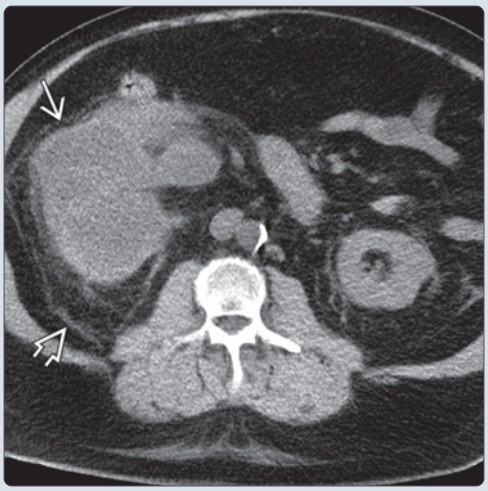
- Purulent fluid in dilated renal collecting system
  - Causes of renal collecting system obstruction
    - Stone, tumor, postsurgical stricture, congenital anomalies

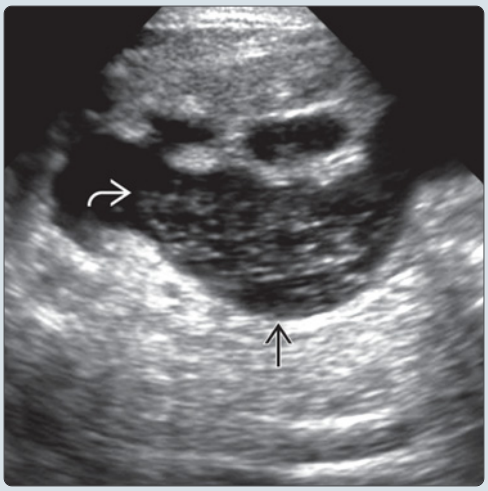
CLINICAL ISSUES

- Medical emergency
- If left untreated → sepsis → death
- Must be treated emergently
  - Antibiotics + urgent drainage of pus
  - Percutaneous nephrostomy or ureteral catheterization

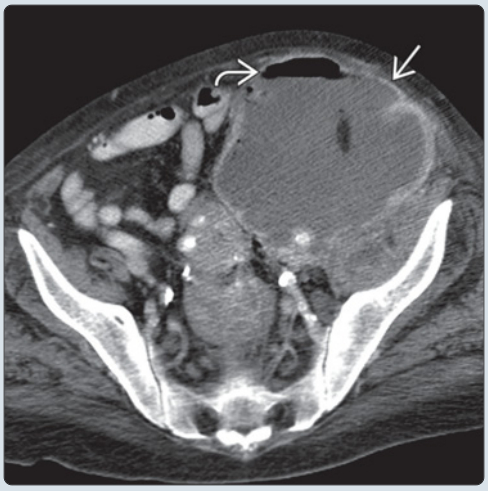
DIAGNOSTIC CHECKLIST

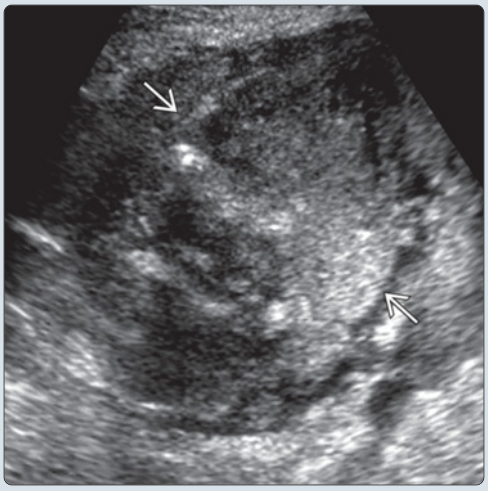
- Consider ultrasound or CT-guided aspiration of fluid in dilated collecting system to confirm diagnosis





(Left) *Axial NECT in pyonephrosis shows severe dilatation of the right renal collecting system. The attenuation of the fluid within the dilated renal pelvis is higher than water. Note perinephric inflammatory changes. Pyonephrosis is a medical emergency.* **(Right)** *Renal ultrasound shows a dilated collecting system filled with low-level echoes; the combination of imaging findings is diagnostic for pyonephrosis.*





(Left) *Axial CECT of the pelvis in a patient with fever and a left lower quadrant renal transplant demonstrates severe pyonephrosis of the renal allograft. Note severe dilatation of the renal pelvis with gas, and fluid with attenuation higher than water.* **(Right)** *Longitudinal ultrasound through the left lower quadrant renal allograft in the same patient with severe pyonephrosis reveals severe dilatation of the renal collecting system with echogenic material. Nephrostomy drainage was performed emergently.*

TERMINOLOGY

Definitions

- Infection of obstructed renal collecting system

IMAGING

General Features

- Best diagnostic clue
  - Ultrasound
    - Dilated renal collecting system with low-level echoes
  - CT
    - Dilated renal collecting system with fluid-debris levels ± gas

CT Findings

- NECT
  - Dilated renal collecting system
  - Fluid-debris level ± gas in renal pelvis
  - Attenuation of fluid within dilated renal pelvis may be higher than water
  - Perinephric inflammatory changes
- CECT
  - Thickened urothelial lining of renal pelvis and proximal ureter
  - Layer of contrast material over infected fluid (rare)

MR Findings

- T2WI
  - Dilated renal collecting system
  - ↑ signal intensity of fluid within dilated collecting system
    - Fluid-debris levels
- DWI
  - Under investigation
    - Lower ADC values of fluid in dilated collecting system in pyonephrosis compared to hydronephrosis

Ultrasonographic Findings

- Grayscale ultrasound
  - Dilated renal collecting system
    - Low-level echoes
    - Fluid-debris levels
    - Occasionally anechoic fluid

Imaging Recommendations

- US &/or MDCT

DIFFERENTIAL DIAGNOSIS

Hydronephrosis

- Ultrasound
  - Dilated, anechoic renal collecting system
  - Acoustic enhancement; absent internal echoes

Transitional Cell Carcinoma

- Soft tissue, enhancing mass dilating renal collecting system

PATHOLOGY

General Features

- Etiology
  - Causes of renal collecting system obstruction in adults
    - Stones
    - Tumor
    - Strictures
    - Congenital anomalies (e.g., UPJ obstruction)
  - Pathogens
    - *Escherichia coli, Proteus mirabilis,* etc.

Gross Pathologic & Surgical Features

- Purulent fluid in dilated renal collecting system

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, flank pain, dysuria
  - Bacteriuria, pyuria, leukocytosis
- Other signs/symptoms
  - Low-grade fever, weight loss
  - No symptoms (15%)

Natural History & Prognosis

- Medical emergency
- Poor prognosis if left untreated
- Complications: Sepsis and death

Treatment

- Must be treated emergently
- Antibiotics (gram-negative coverage) + urgent drainage of pus
  - Percutaneous nephrostomy
  - Ureteral catheterization
- Surgery (radical nephrectomy)

DIAGNOSTIC CHECKLIST

Consider

- Ultrasound or CT-guided aspiration of fluid in dilated collecting system to confirm diagnosis

Image Interpretation Pearls

- US: Low-level echoes within dilated pelvis

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KEY FACTS

TERMINOLOGY

- Renal involvement by unusual infections in immunocompromised or debilitated host

IMAGING

- CT or ultrasound

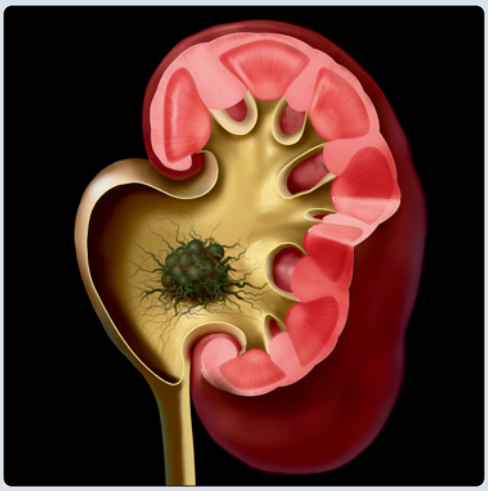
TOP DIFFERENTIAL DIAGNOSES

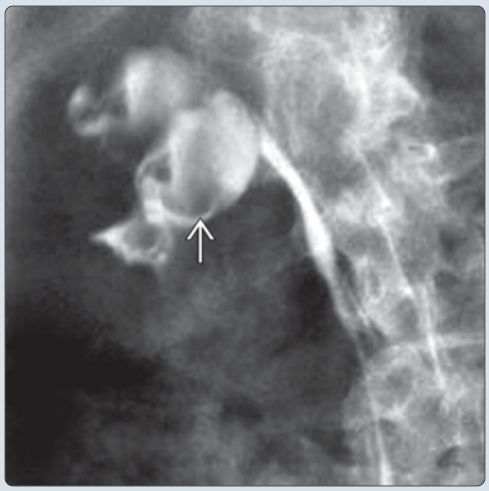
- Candidiasis
  - Multiple, low-attenuation, nonenhancing lesions with hydronephrosis on CT
  - Nonenhancing filling defect within collecting system on CT
- Pneumocystis carinii
  - Punctate cortical calcifications in healing phase
  - Multiple focal areas of ↑ echogenicity in cortex and medulla
- Aspergillosis
  - Immunocompromised patients
  - Filling defects ("fungus balls") within dilated collecting system
  - Renal aspergilloma (pseudotumor)
- Tuberculosis
  - Hematogenous spread of *Mycobacterium tuberculosis*
  - 4-8% of patients with pulmonary disease
  - Strictures (ureter, infundibulum, pelvis)
  - Caliectasis; hydronephrosis
  - Calcifications (punctate, linear, curvilinear, diffuse)
- Mucormycosis
  - Renal infarction
  - Thrombosis of renal vessels

DIAGNOSTIC CHECKLIST

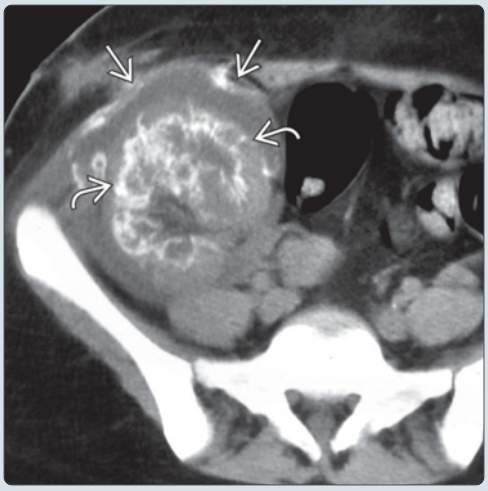
- Serologic studies are very helpful to establish diagnosis as imaging findings are often nonspecific
- Poorly defined, noncalcified filling defects within hydronephrotic kidney of debilitated patient

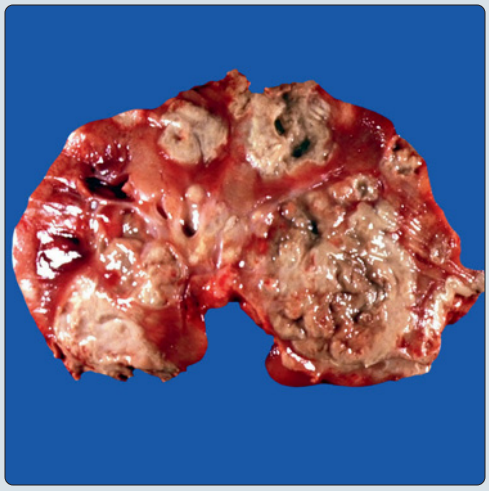
(Left) *Graphic shows a hydronephrotic kidney with a "fungus ball" in the dilated pelvis. Renal fungal infections may result from Candida albicans or Aspergillus and usually occur in debilitated patients. They may result from or cause ureteral obstruction.* **(Right)** *Retrograde pyelogram of the right kidney shows filling defects in mildly dilated collecting system corresponding to a "fungus ball" in a patient with Candida.*





(Left) *NECT of a young patient with a failed transplant and fever shows amorphous calcification throughout the allograft and a rim of perinephric fluid. Aspergillus infection was confirmed after biopsy and nephrectomy. The diagnosis of renal aspergillosis is difficult given its rarity and nonspecific appearance, but a history of immunosuppression is typical.* **(Right)** *Gross pathology image of the renal allograft shows multiple abscesses.*





TERMINOLOGY

Definitions

- Renal involvement by unusual infections in immunocompromised or debilitated host

IMAGING

Imaging Recommendations

- Best imaging tool
  - CT or ultrasound

DIFFERENTIAL DIAGNOSIS

Candidiasis

- Imaging finding
  - Renal parenchymal infection
    - Focal abscess/microabscess within parenchyma; may extend into perirenal space
    - Papillary necrosis (rare)
  - Collecting system involvement
    - Hydronephrosis due to ureteral obstruction
    - Nonenhancing, soft, nonopaque filling defect within dilated collecting system on CT, urography, or sonography
    - Fungal material has lower attenuation compared to calculus on CT
    - "Fungus ball": *Candida* is most common organism
- Epidemiology
  - Kidney is major target organ in candidemia
- Clinical manifestation
  - Pyelonephritis, flank pain, fever
  - Renal colic secondary to "fungus ball"
- Treatment
  - Systemic antifungal
  - Percutaneous nephrostomy with extraction

Pneumocystis carinii

- Most common opportunistic infection in HIV patients
- Opportunistic infections in patients with HIV/AIDS seen with CD4 < 200
- Renal involvement is uncommon
  - Almost always associated with *Pneumocystis* pneumonia
- CT: Punctate cortical calcification in healing phase
  - Diffuse or ill-defined hypodense lesions in acute phase
- US: Multiple focal areas of ↑ echogenicity in cortex and medulla
- Pathology: Areas of tubular destruction with focal calcifications in cortex

Aspergillosis

- Rare
- Immunocompromised subjects
- Etiology: *Aspergillus fumigatus* and *Aspergillus flavus*
- Imaging findings
  - Filling defects within dilated collecting system ("fungus balls")
  - Renal aspergilloma (pseudotumor)
  - Uni- or bilateral
  - Multiple abscesses within kidney, with possible extension into perirenal space
- Clinical manifestations
  - Flank pain, flank tenderness, fever
  - Obstructive uropathy

Tuberculosis

- Hematogenous spread of *Mycobacterium tuberculosis*
- 4-8% of patients with pulmonary disease
- Imaging findings
  - Necrotic lesions
  - Strictures (ureter, infundibulum, pelvis)
  - Caliectasis; hydronephrosis
  - Calcifications (punctate, linear, curvilinear, diffuse)
  - "Putty kidney": End-stage infection associated with diffuse calcifications (autonephrectomy)
- Clinical manifestations
  - Nonspecific symptoms
  - Hematuria; culture negative pyuria

Mucormycosis

- More common in patients with diabetes, chronic kidney disease, and after hematogenic malignancies
- Renal infarction
- Thrombosis of renal vessels
- Clinical manifestations
  - Obstructive uropathy
  - Acute renal infection

DIAGNOSTIC CHECKLIST

Consider

- Serologic studies are very helpful to establish diagnosis as imaging findings are often nonspecific

Image Interpretation Pearls

- Poorly defined, noncalcified filling defects within hydronephrotic kidney of debilitated patient
  - Strongly suggestive of fungal infection (*Candida* or *Aspergillus*)

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Renal Cyst

KEY FACTS

TERMINOLOGY

- Benign, fluid-filled, nonneoplastic renal lesion

IMAGING

- **Bosniak classification**
  - Imaging-based classification of renal cystic lesions
    - Class 1: Benign cysts
    - Class 2: Minimally complicated cysts; benign
    - Class 2F: Requires CT/MR imaging follow-up
    - Class 3: More complicated cysts; require surgery (biopsy controversial)
    - Class 4: Malignant lesions; require surgery
- Simple cyst
  - Ultrasound: Simple, uncomplicated cyst; spherical or ovoid, anechoic content, sharply defined, imperceptible wall, and acoustical enhancement
  - NECT: Sharply marginated, round, smooth, homogeneous, hypodense (< 20 HU) mass
  - MR : ↓ signal intensity on T1WI and ↑ signal intensity on T2WI
- Neoplastic cystic masses: Enhancing soft tissue component

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma
- Multilocular cystic nephroma
- Renal abscess
- Renal metastases and lymphoma
- Autosomal dominant polycystic kidney disease
- Uremic cystic disease

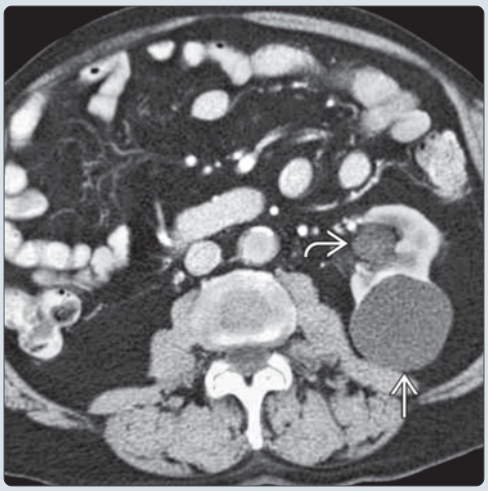
CLINICAL ISSUES

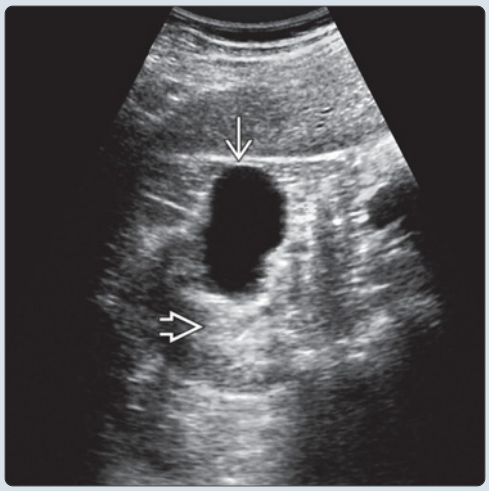
- Asymptomatic or palpable mass and flank pain
- Present in 20-30% of middle-aged adults, incidence increases with age

DIAGNOSTIC CHECKLIST

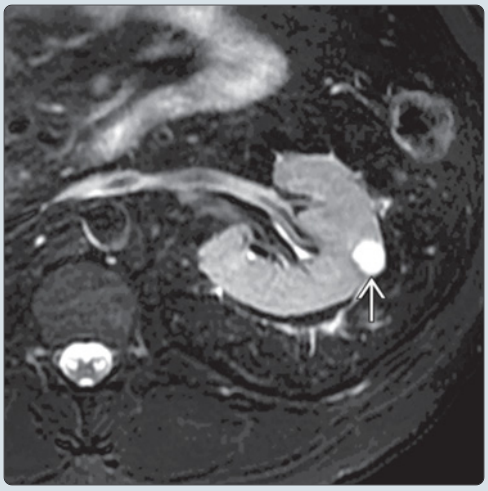
- Image evaluation and classification of cystic masses are key to management

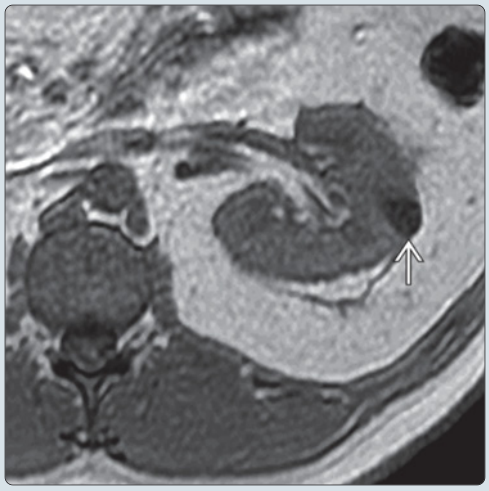
(Left) *Axial CECT shows a spherical, water attenuation mass with no definable wall compatible with a simple cyst (Bosniak class 1). There is also a simple renal sinus cyst, which may be arising from either the medial cortex and projecting into the renal sinus, or is of primary lymphatic origin.* **(Right)** *Transverse ultrasound shows a simple renal cyst displaying the typical imaging features including anechoic content, imperceptible wall, and acoustic enhancement.*





(Left) *Axial T2WI FS MR shows a simple hyperintense cyst in the left kidney.* **(Right)** *Axial T1WI in the same patient shows a simple hypointense cyst in the left kidney. The lesion represents a Bosniak class 1 cyst. Bosniak classification can be used on CT and MR imaging. Cyst class may be upgraded on MR given the higher contrast resolution and increased visualization of wall thickness and internal septations.*





TERMINOLOGY

Definitions

- Benign, fluid-filled, nonneoplastic renal lesion

IMAGING

General Features

- Best diagnostic clue
  - Water density, nonenhancing, spherical lesion with no visible wall on CT
  - Anechoic lesion with through transmission and no visible wall on US
- **Bosniak classification**
  - Imaging-based classification of renal cystic lesions
    - Used with CT and MR imaging
      - Lesion classification may be upgraded on MR images because of ↑ contrast resolution
  - 5 categories
    - Class 1: Benign cysts
      - Well-defined, rounded, homogeneous, lucent (0-20 HU, near water density) mass with thin or imperceptible, nonenhancing wall and contents; no septa or calcifications
    - Class 2: Minimally complicated cysts; benign
      - Thin (< 1 mm) septations, smooth, ± perceived (not measurable) enhancement
      - Calcification of a short segment of wall or septa
      - Hyperdense cyst (> 20 HU), no enhancement, spherical, partially exophytic, usually ≤ 3 cm in diameter
    - Class 2F: Requires CT/MR imaging follow-up
      - Multiple thin septations ± perceived (not measurable) enhancement
      - Minimal wall or septal thickening ± perceived (not measurable) enhancement
      - Thick calcifications
      - Hyperdense, intraparenchymal cysts > 3 cm
      - No enhancing nodules
    - Class 3: More complicated cysts; require surgery (biopsy controversial)
      - Irregular and thickened septa &/or wall + measurable enhancement
      - ± thickened and irregular calcification
      - Benign lesions: Hemorrhagic cysts, renal abscess, mixed epithelial and stromal tumor, complicated benign septated cysts
      - Malignant lesions: Cystic nephroma, multiloculated cystic renal cell carcinoma
    - Class 4: Malignant lesions; require surgery
      - Enhancing component, irregular wall thickening

Ultrasonographic Findings

- Grayscale ultrasound
  - Simple, uncomplicated cyst: Spherical or ovoid, anechoic content, sharply defined, imperceptible wall, and acoustical enhancement
  - Hemorrhagic cyst: Internal echoes (clot); thick calcified wall ± multiloculated (chronic)
  - Infected cyst: Thick wall with scattered internal echoes ± debris-fluid level

CT Findings

- NECT
  - Simple, uncomplicated cyst
    - Sharply marginated, round, smooth, homogeneous, hypodense (< 20 HU) mass
    - Thin, imperceptible, wall
  - Complicated cysts
    - Due to hemorrhage, infection, ischemia, proteinaceous fluid
    - May show ↑ density (> 20 HU on NECT), septations, wall thickening, calcifications
    - Classification and management is based on Bosniak classes
    - Hemorrhagic cyst
      - Hyperdense (60-90 HU), homogeneous content (acute)
      - ± heterogenous content (clot or debris), wall thickening, calcifications (chronic)
    - Infected cyst: Thick wall, septate, heterogeneous enhancing fluid, debris- or gas-fluid level, ± calcification (chronic)
  - Milk of calcium cyst: Dependent, fluid-calcium layer
- CECT
  - Enhancement post administration of IV contrast
    - Should be evaluated on nephrographic phase images (i.e., 100 seconds post contrast administration)
    - Enhanced HU minus unenhanced HU
      - < 10 HU: No enhancement
      - 10-20 HU: Indeterminate
      - ≥ 20 HU: Enhancement
    - "Pseudoenhancement"
      - Refers to artifactual increase in cyst content attenuation (by > 10 HU) on CECT
      - Related to several factors, including partial volume averaging and beam hardening
  - Simple cysts: No enhancement
  - Complicated cysts: Granulation tissue in inflammatory/infected cysts may enhance
  - Neoplastic cystic masses: Enhancing soft tissue component

MR Findings

- T1WI
  - Simple cysts: ↓ signal intensity, round/oval, homogeneous mass
  - Complicated, hemorrhagic cysts: ↑ signal intensity (signal intensity changes according to stage of hemorrhage)
- T2WI
  - Simple cysts: ↑ signal intensity, homogeneous mass with imperceptible wall
  - Complicated, hemorrhagic cysts: ↓ signal intensity (signal intensity changes according to stage of hemorrhage) ± fluid-debris level
- T1WI C+
  - Image subtraction (gadolinium-enhanced images minus unenhanced images) helpful for evaluation of enhancement
  - Simple cysts: No enhancement
  - Neoplastic cystic masses: Enhancing soft tissue component

Imaging Recommendations

- Best imaging tool
  - Management of hyperattenuating cystic lesions on NECT
    - > 70 HU on NECT = hyperdense, benign renal cyst requiring no follow-up or treatment
    - 20-70 HU on NECT: Requires contrast administration to exclude enhancing components; alternatively, US or MR imaging (T1WI, T2WI, DWI) can be used if patient cannot tolerate IV contrast administration
      - At US, hyperdense lesions on CT may often appear as anechoic, benign cysts
- Protocol advice
  - CT: NECT + CECT nephrographic phase (100 seconds after IV contrast administration), section thickness ≤ 5 mm
  - Consider US or MR for cystic lesions with equivocal enhancement at CECT

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- Some RCCs are cystic from inception (not just large "necrotic" tumors)
- Any enhancing mural nodularity should be considered neoplastic

Multilocular Cystic Nephroma

- Septate, encapsulated, multicystic mass
- Herniation into renal sinus

Renal Abscess

- Enhancing capsule, ± perinephric stranding
- Shaggy wall; hyperdense (> water) but nonenhancing contents
- Key to diagnosis: Correlation with clinical signs of infection

Renal Metastases and Lymphoma

- Usually > water density with enhancement of mass
- Lymphoma most common hypovascular tumor that may mimic cyst
  - Often multiple with adenopathy

Autosomal Dominant Polycystic Kidney Disease

- Inherited polycystic kidney disease characterized by progressive cystic growth
- Enlarged kidneys replaced by innumerable cysts
- Extrarenal manifestations: polycystic liver disease, intracranial arterial aneurysm, cysts in other organs, abdominal hernias

Uremic Cystic Disease

- Multiple renal cysts in patients with end-stage renal disease and no history of hereditary cystic disease

PATHOLOGY

Gross Pathologic & Surgical Features

- Uncomplicated cyst
  - Unilocular; arises in cortex and bulge from renal surface, less commonly into renal sinus
  - Smooth, yellow-white, thin, translucent wall with cuboidal or flattened epithelium
- Hemorrhagic cyst: Rust-colored putty-like material ± surrounded by fibrosis and rim of calcification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic or palpable mass and flank pain

Demographics

- Gender
  - M > F
- Epidemiology
  - Present in 20-30% of middle-aged adults, increases with age
    - 39% in 18-49 year olds
    - 63% in 50-75 year olds

Natural History & Prognosis

- Simple cysts grow slowly
- Complications are rare: Hydronephrosis, hemorrhage, infection, rupture
- Risks of renal cell carcinoma
  - Bosniak class 1, 2: 0%
  - Bosniak class 2F: 11-25%
  - Bosniak class 3: 28-54%
  - Bosniak class 4: 90%

Treatment

- Bosniak class 1, 2: Benign → no follow-up or treatment
- Bosniak class 2F: Imaging follow-up
  - CT/MR, interval time: 6, 12 months
  - Concerning imaging features at follow-up: Enhancing solid nodules or thick septations, increased thickness of enhancing wall
    - Cyst growth is not correlated with progression to malignancy
  - Recommended length of follow-up: 4-5 years
- Bosniak class 3, 4: Treatment (according to patient conditions; biopsy is controversial)
  - Surgery

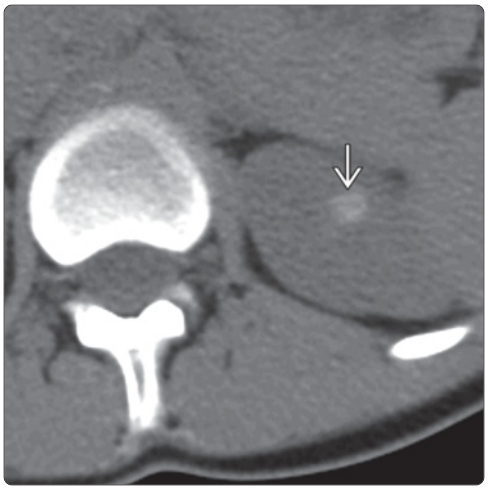
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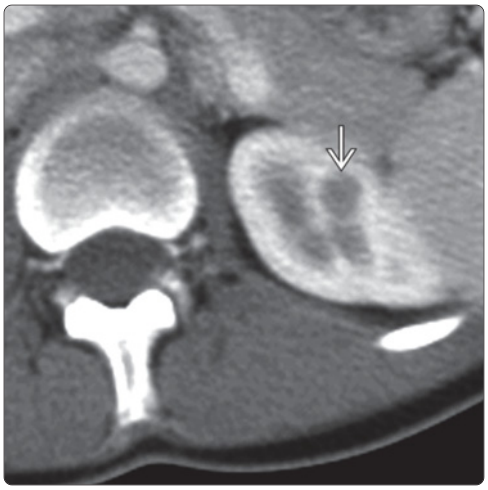
Image Interpretation Pearls

- Image evaluation and classification of cystic masses are key to management

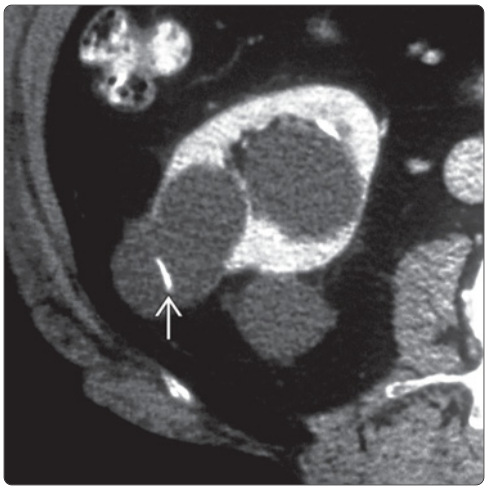
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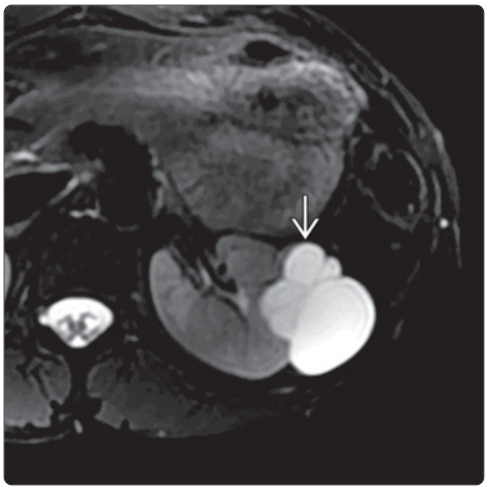
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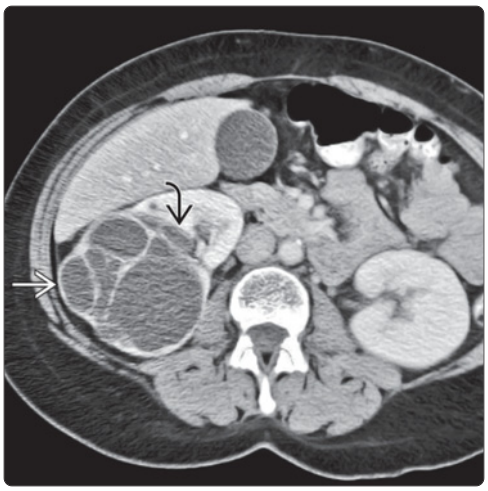


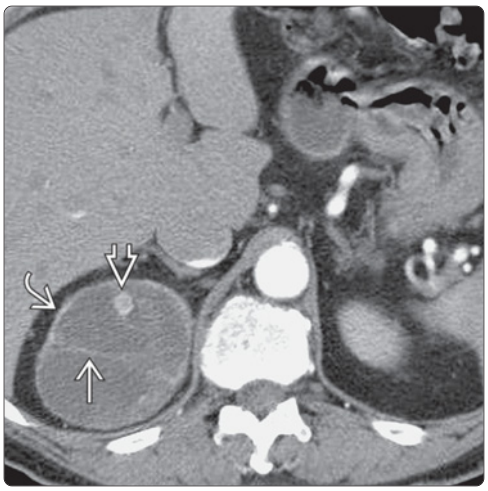
(Left) *Axial NECT shows an hyperdense (100 HU) cyst in the left kidney (Bosniak class 2).* **(Right)** *Axial CECT in the same patient shows no enhancement of the left renal cyst (106 HU). Hyperdense renal cysts measuring > 70 HU on NECT are considered benign. Soft tissue attenuation (20-70 HU) lesions at NECT should be evaluated by ultrasound or dedicated renal CT to differentiate between true solid lesions and high-attenuation cysts.*





(Left) *Axial CECT shows multiple water density cystic lesions, including 1 with a calcified thin septum (Bosniak class 2).* **(Right)** *Axial T2WI FS MR shows a lobulated left renal cyst with multiple internal thin septations. Though thin, the septations qualitatively enhance on Gd-enhanced T1WI (not shown). The lesion is a Bosniak class 2F cyst. Class 2F cysts require imaging follow-up with CT or MR imaging.*





(Left) *Axial CECT shows a multiloculated cystic mass with septa with measurable enhancement (Bosniak class 3). The invagination into the renal sinus, is characteristic of multilocular cystic nephroma.* **(Right)** *Axial CECT shows a cystic right upper pole renal mass with thick septations, enhancing mural nodules, and a thick wall compatible with Bosniak class 4 cyst. This represents a case of cystic renal cell carcinoma. The presence of enhancing nodules is indicative of malignancy.*

Parapelvic (Peripelvic) Cyst

KEY FACTS

TERMINOLOGY

- Synonym: Renal sinus cysts
- Definitions
  - Parapelvic cyst: Renal cysts extending into renal sinus fat
  - Peripelvic cyst: Originating from renal sinus lymphatics

IMAGING

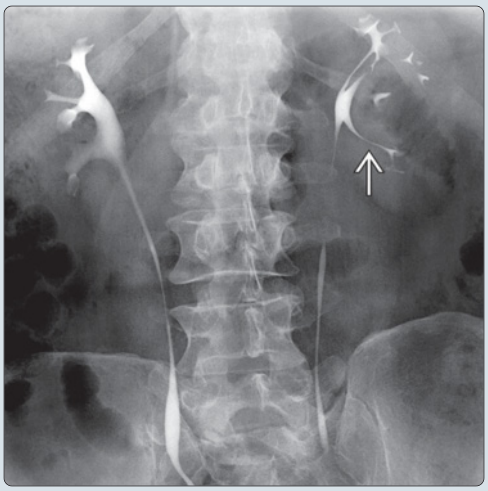
- Parapelvic cysts
  - Usually single and unilateral
- Peripelvic cysts
  - Usually multiple and bilateral
- IVP
  - Compression and displacement of renal calyces and pelvis
- CECT
  - Excretory phase: Water-attenuation cysts separate from enhanced renal collecting system
- Ultrasound
  - Anechoic lesions in renal sinus
  - No connection among cysts
- Best diagnostic clue
  - Contrast-enhanced CT or MR obtained during excretory phase show enhanced collecting system separate from nonenhancing renal cysts

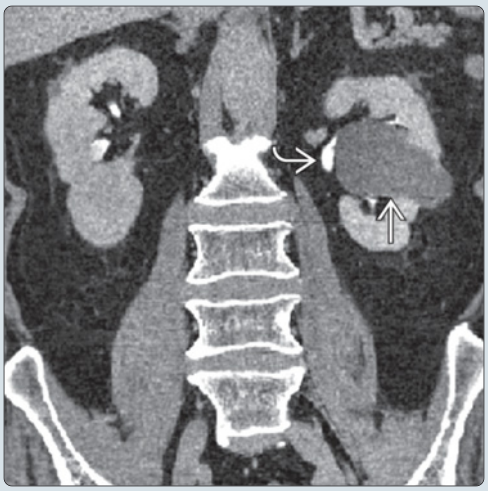
TOP DIFFERENTIAL DIAGNOSES

- Hydronephrosis
- Renal lipomatosis
- Renal lymphangiomatosis

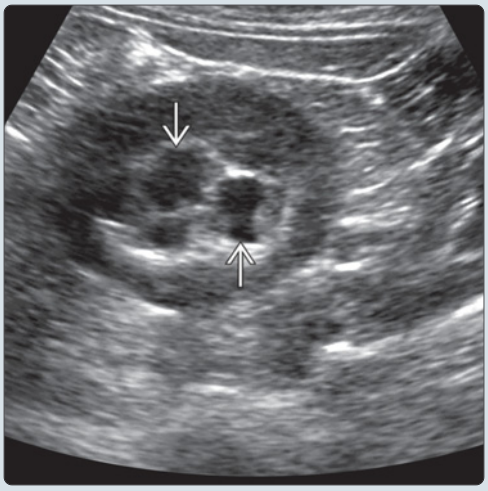
CLINICAL ISSUES

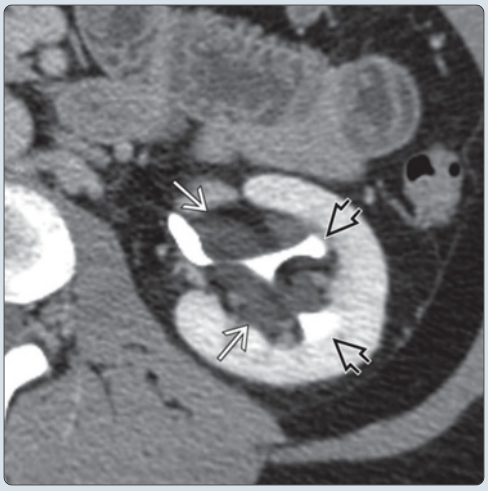
- Most commonly asymptomatic
- Treatment for symptomatic cysts
  - Laparoscopic, ureteroscopic, or percutaneous ablation





(Left) *Retrograde pyelogram shows displaced and compressed left renal collecting system ➡. (Right) Coronal CECT obtained during the excretory phase in the same patient shows a large nonenhancing left parapelvic cyst ➡ displacing the enhancing renal collecting system ➡.*





(Left) *Transverse ultrasound of the left kidney shows multiple anechoic cysts in the renal pelvis ➡. (Right) Axial CECT obtained during the excretory phase in the same patient demonstrates peripelvic cysts ➡ displacing the enhanced renal collecting system ⇨. Excretory phase of contrast-enhanced CT or MR helps in differentiating cysts from enhancing renal collecting system.*

TERMINOLOGY

Synonyms

- Renal sinus cyst

Definitions

- Parapelvic cyst: Renal cysts extending into renal sinus fat
- Peripelvic cyst: Originating from renal sinus lymphatics

IMAGING

General Features

- Best diagnostic clue
  - Contrast-enhanced CT or MR obtained during excretory phase
- Location
  - Renal pelvis
  - Parapelvic cysts
    - Usually single and unilateral
    - Rarely multiple and bilateral
  - Peripelvic cysts
    - Usually multiple and bilateral
- Size
  - Variable

Radiographic Findings

- IVP
  - Compression and displacement of renal calyces and pelvis

CT Findings

- CECT
  - Excretory phase
    - Water-attenuation cysts separate from enhanced renal collecting system
    - Cysts do not communicate with collecting system
    - Thin, nondiscernible wall
    - No enhancement

MR Findings

- ↑ signal intensity on T2WI; ↓ signal intensity on T1WI
- No communication with renal collecting system

Ultrasonographic Findings

- Anechoic lesions in renal sinus
- No connection among cysts
- No connection with ureter

DIFFERENTIAL DIAGNOSIS

Hydronephrosis

- Dilatation of collecting system
- Anechoic spaces in renal sinus communicating among them and with ureter

Renal Lipomatosis

- Fatty tissue proliferation in renal sinus
- Enlarged renal sinus with fat density (-30 to -90 HU)
- + renal parenchymal atrophy and ± centrally located calculus in replacement lipomatosis

Renal Lymphangiomatosis

- Development malformation
- Uni- or multilocular cystic mass in perirenal or peripelvic area

PATHOLOGY

General Features

- Etiology
  - Peripelvic cysts
    - Lymphatic origin
  - Parapelvic cysts
    - Extension of renal cysts into the renal pelvis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Complications of parapelvic cysts (rare)
    - Hydronephrosis
    - Hypertension
    - Infection
    - Hemorrhage

Demographics

- Gender
  - More frequent in men
- Epidemiology
  - Prevalence at autopsy: 1.3-1.5%
  - High prevalence (50%) in patients with Fabry disease

Treatment

- Symptomatic cysts: Laparoscopic, ureteroscopic, or percutaneous ablation

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Excretory phase of contrast-enhanced CT or MR to differentiate cysts from enhancing renal collecting system

SELECTED REFERENCES

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Autosomal Dominant Polycystic Kidney Disease

KEY FACTS

TERMINOLOGY

- Autosomal dominant polycystic kidney disease (ADPKD)
- Inherited polycystic renal disease characterized by progressive cystic growth and multiple systemic manifestations

IMAGING

- Enlarged kidneys replaced by innumerable cysts
  - Simple ± complicated (hemorrhagic, infected) cysts
  - Progressive increase in cyst and renal volume
- Extrarenal manifestations
  - Polycystic liver disease, intracranial arterial aneurysm, cysts in other organs, abdominal hernias

TOP DIFFERENTIAL DIAGNOSES

- Multiple simple cysts
- Uremic cystic disease
- von Hippel-Lindau disease
- Medullary cystic kidney disease

PATHOLOGY

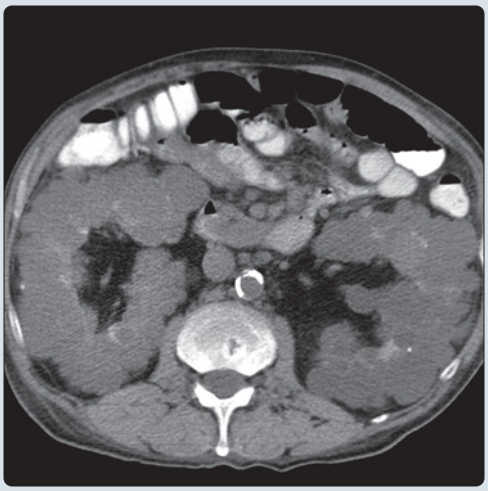
- Autosomal dominant inheritance
  - *PKD1* gene mutation (85% of cases)
  - *PKD2* gene mutation (15% of cases)
- Enlarged kidneys (2-4 Kg; normal kidney = 150 g), multiple cysts of various size in cortex and medulla

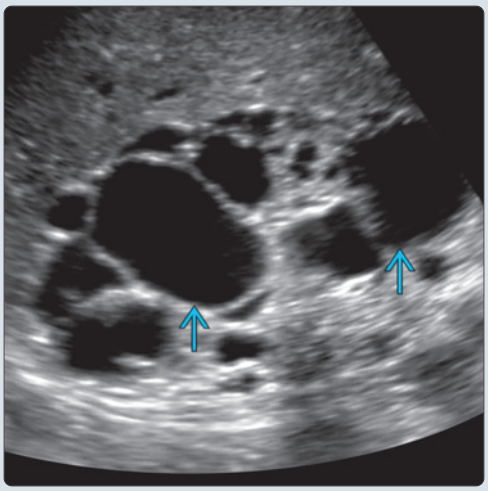
CLINICAL ISSUES

- Incidence in US: 1:400-1000
- Asymptomatic, abdominal pain, hematuria, hypertension
- Complications
  - 50% evolve to end-stage renal disease (ESRD) by 6th decade
  - Renal function is related to kidney volume

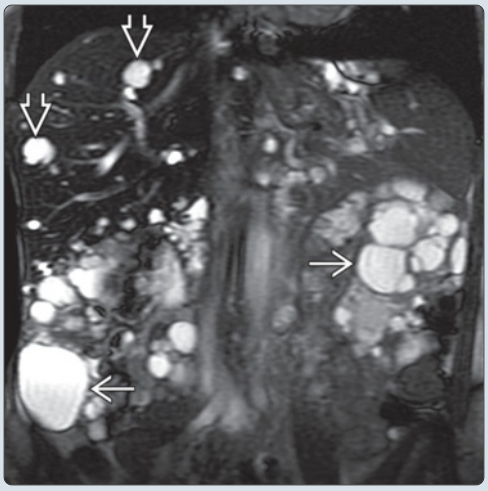
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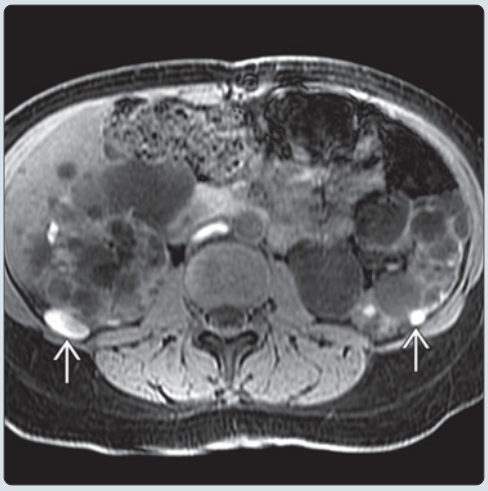
- Enlarged kidneys with multiple, bilateral cysts
- ± polycystic liver disease, and cysts in other organs (pancreas, spleen)





(Left) *Axial NECT in patient with autosomal dominant polycystic kidney disease shows massively enlarged kidneys replaced by innumerable simple cysts.* **(Right)** *Longitudinal ultrasound of the right kidney in patient with autosomal dominant polycystic kidney disease shows multiple varying size simple anechoic cysts that replace and enlarge the kidney.*





(Left) *Coronal T2WI shows enlarged kidneys replaced by innumerable T2-hyperintense cysts and multiple liver cysts. Liver cysts are the most common extrarenal manifestation in patients with autosomal dominant polycystic kidney disease* **(Right)** *Axial T1WI with fat saturation in the same patient shows multiple renal cysts with varying signal intensity. Complicated, hemorrhagic cysts appear hyperintense on T1WI.*

TERMINOLOGY

Abbreviations

- Autosomal dominant polycystic kidney disease (ADPKD)

Definitions

- Inherited polycystic renal disease characterized by progressive cystic growth and multiple systemic manifestations

IMAGING

General Features

- Best diagnostic clue
  - Enlarged kidneys replaced by innumerable cysts
- Location
  - Bilateral

Ultrasonographic Findings

- Grayscale ultrasound
  - Enlarged kidneys
  - Multiple bilateral renal cysts
    - Simple cysts: Anechoic
    - Complex cysts: Hyperechoic content, thick wall, septations
- Diagnostic criteria for at-risk individuals from ADPKD families (Pei et al. 2009)
  - 15-39 years: ≥ 3 renal cysts
  - 40-59 years: ≥ 2 cysts in each kidney
  - ≥ 60 years: ≥ 4 cysts in each kidney

CT Findings

- Multiple bilateral cysts in enlarged kidneys
  - Simple cysts
    - Water attenuation (0-20 HU), imperceptible walls, no enhancement
  - Complicated cysts
    - Hemorrhagic cyst: Hyperdense content (60-90 HU), ± mural calcifications
    - Infected cysts: Thick, irregular wall ± gas, ± wall enhancement

MR Findings

- Multiple bilateral cysts in enlarged kidneys
  - > 10 renal cysts is diagnostic in patients at risk for ADPKD
  - Simple cysts: Signal intensity ↑ on T2WI and ↓ on T1WI
  - Complicated cysts: Signal intensity ↑ on T1WI and variable on T2WI, mural thickening
  - No enhancement on Gd-enhanced images

Other Renal Findings

- Nephrolithiasis and hydronephrosis in case of obstruction
- Perinephric hemorrhage with cyst rupture

Extrarenal Manifestations

- Polycystic liver disease
  - > 20 cysts and > 50% liver involvement
  - Intrahepatic cysts and peribiliary cysts
  - Hepatomegaly
- Cysts in pancreas, spleen, and seminal vesicles
- Intracranial arterial aneurysm, aortic aneurysm
- Aortic and mitral valve abnormalities
- Hernia, colonic diverticula

Imaging Recommendations

- Best imaging tool
  - Ultrasound for screening and diagnosis
  - CT and MR for assessment of complications and renal volume

DIFFERENTIAL DIAGNOSIS

Multiple Simple Cysts

- Less numerous, normal renal size and function

Uremic Cystic Disease

- Multiple cysts in atrophic kidneys; end-stage renal disease or dialysis

von Hippel-Lindau Disease

- Multiple renal cysts, ± renal cell carcinoma, ± other extrarenal manifestations (central nervous system hemangioblastoma, pheochromocytoma, pancreatic cysts)

Medullary Cystic Kidney Disease

- Small, smooth kidneys with cysts in medulla

PATHOLOGY

General Features

- Genetics
  - Autosomal dominant inheritance
    - Complete penetrance
    - 5% of cases spontaneous mutations
  - *PKD1* gene mutation
    - Short arm chromosome 16
    - Responsible for polycystin 1
  - *PKD2* gene mutation
    - Long arm chromosome 4
    - Responsible for polycystin 2
- Associated abnormalities
  - Polycystic liver disease (> 80%)
  - Abdominal wall hernias (45%)
  - Cardiac valve abnormalities (25%)
  - Brain aneurysms (3-7%)
  - Cysts in other organs (pancreas, seminal vesicles)

Staging, Grading, & Classification

- *PKD1* gene mutation
  - 85% of cases
  - More severe presentation
  - Earlier presentation (54 years)
- *PKD2* gene mutation
  - 15% of cases
  - Milder and later presentation (74 years of age)
  - Fewer renal cysts
  - Similar growth rate of renal cysts

Gross Pathologic & Surgical Features

- Enlarged kidneys (2-4 Kg; normal kidney = 150 g)
- Multiple cysts of various size in cortex and medulla
- Compressed renal parenchyma in between cysts
- Bands of fibrotic tissue in late stage of disease

Microscopic Features

- Single layer cyst epithelium

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic, abdominal pain
  - Hematuria, hypertension, cyst infection/pyelonephritis, nephrolithiasis
- Other signs/symptoms
  - End-stage renal disease
    - Usually manifest in 5th/6th decade
    - Decrease in renal function is related to renal volume and rate of kidney growth

Demographics

- Age
  - Childhood to 8th decade
  - Most commonly diagnosed between 3rd and 5th decades
- Gender
  - No gender predilection
- Ethnicity
  - No race predilection
- Epidemiology
  - Most common hereditary kidney disease
  - Incidence in USA: 1:400-1,000

Natural History & Prognosis

- Progressive increase in cyst and renal volume
  - 5.3% increase in renal volume per year
- Complications
  - Renal cyst infection
  - Renal cyst hemorrhage, rupture
  - Nephrolithiasis
- 50% evolve to end-stage renal disease (ESRD) by 6th decade
  - 5% of all ESRD cases
  - Most common cause of mortality
  - Renal function is related to kidney volume
    - Height-adjusted total kidney volume > 600 cc/m → increased risk of renal insufficiency
- No increased risk of renal cell carcinoma
- Prognosis
  - Good after renal transplant: Similar survival rate as non-ADPKD patient

Treatment

- Screening
  - Genetic testing (identifies mutations in 47-58% of cases)
  - Ultrasound
- Pain
  - Conservative medical treatment
  - Laparoscopic cyst decortication, renal denervation
- Hypertension
  - Angiotensin-converting enzyme inhibitors
  - Angiotensin II–receptor blockers
- End-stage renal disease
  - Dialysis
  - Renal transplant

DIAGNOSTIC CHECKLIST

Consider

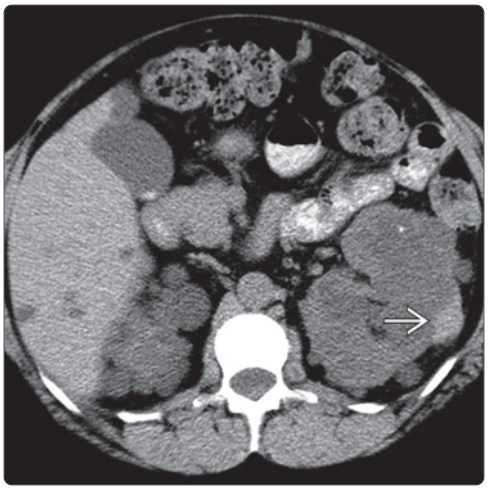
- Screening in patient with family history

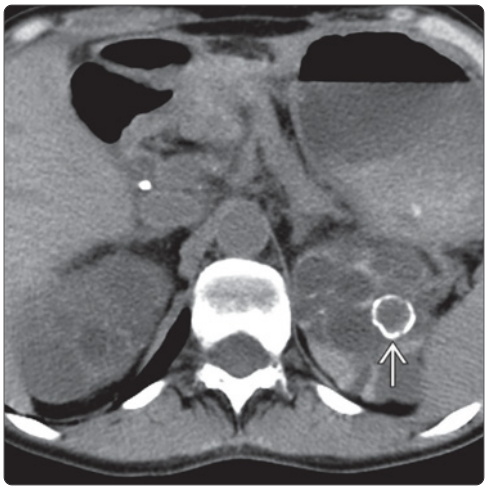
Image Interpretation Pearls

- Enlarged kidneys with multiple, bilateral cysts
- ± polycystic liver disease, and cysts in other organs (pancreas, spleen)
- ± cerebral aneurysms and cardiac valve defects

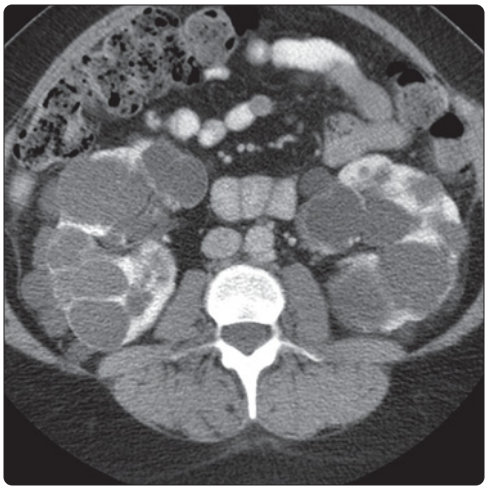
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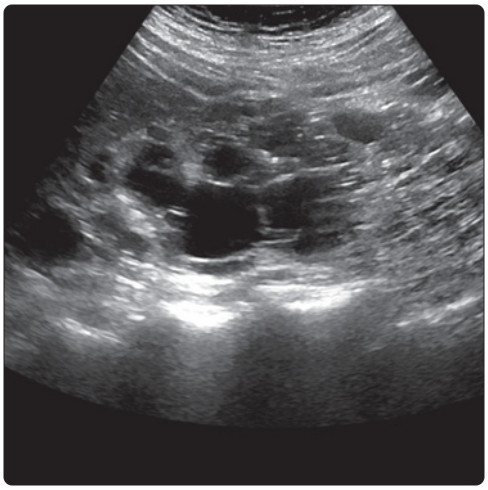
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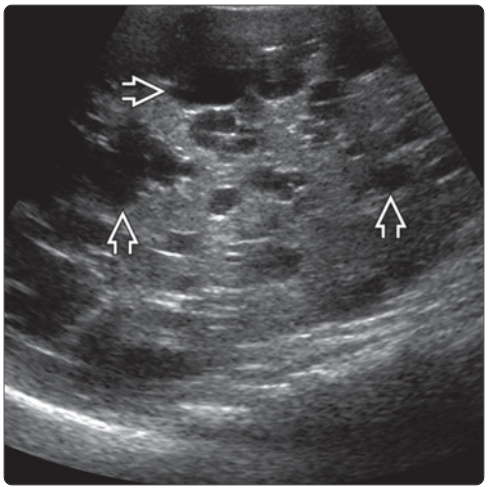
(Left) *Axial NECT shows polycystic involvement of kidneys and liver. While the majority of the renal cysts are hypodense, a peripheral left renal cyst appears hyperdense ⇒ compatible with a hemorrhagic cyst.* **(Right)** *Axial NECT in patient with autosomal dominant polycystic disease reveals multiple renal simple cysts and a complicated cyst with mural calcifications ⇒.*





(Left) *Axial CECT in patient with autosomal dominant polycystic kidney disease reveals multiple bilateral simple renal cysts. Cysts show imperceptible walls and no enhancement.* **(Right)** *Longitudinal ultrasound in the same patient reveals multiple simple, anechoic cysts in an enlarged right kidney. The disease is characterized by a progressive increase in volume of cysts and kidneys.*





(Left) *Axial T2 FS WI shows multiple cysts in left kidney ⇒ and liver ⇒ in this patient with autosomal dominant polycystic kidney disease and polycystic liver disease.* **(Right)** *Transverse ultrasound of the liver in the same patient reveals multiple simple, anechoic hepatic cysts ⇒. Polycystic liver disease is commonly encountered in patients with autosomal dominant polycystic kidney disease. The liver is generally enlarged. Hepatic cysts are intrahepatic and peribiliary.*

Uremic Cystic Disease

KEY FACTS

TERMINOLOGY

- Acquired cystic kidney disease (ACKD)
- ≥ 3 cysts per kidney in patients with end-stage renal disease and no history of hereditary cystic disease

IMAGING

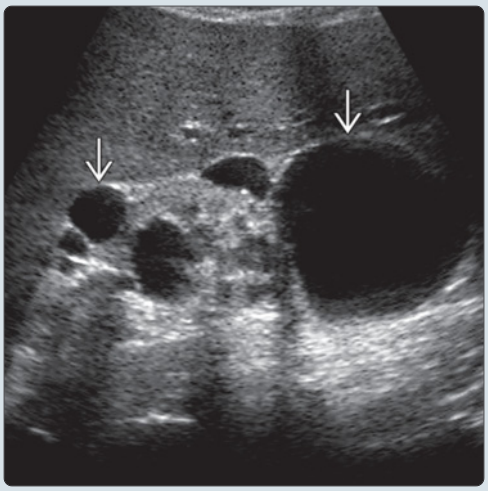
- Imaging findings
  - Atrophic kidneys
  - Multiple cysts
    - Hemorrhagic cysts in 50% of cases
  - Renal cell carcinoma (RCC) in 3-6% of cases
- Best imaging tools
  - Ultrasound: Screening
  - CT or MR: Complications, diagnosis of RCC

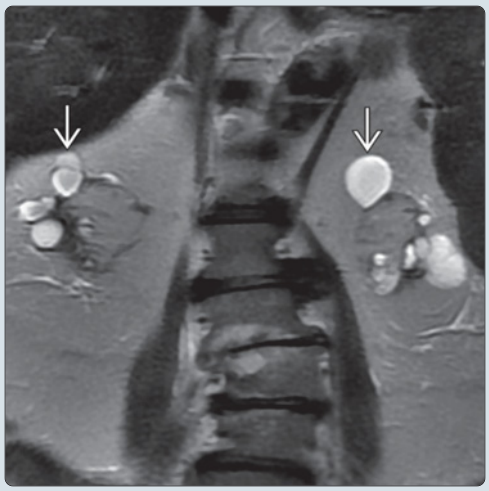
PATHOLOGY

- Multiple cysts of various size
  - Single layer of hyperplastic epithelium
- Histological subtypes of RCC
  - Papillary (most common among the subtypes similar to sporadic RCC), clear cell, chromophobe
  - Acquired cystic disease-associated RCC (most common in ACKD), clear cell papillary renal cell carcinoma of end-stage kidney

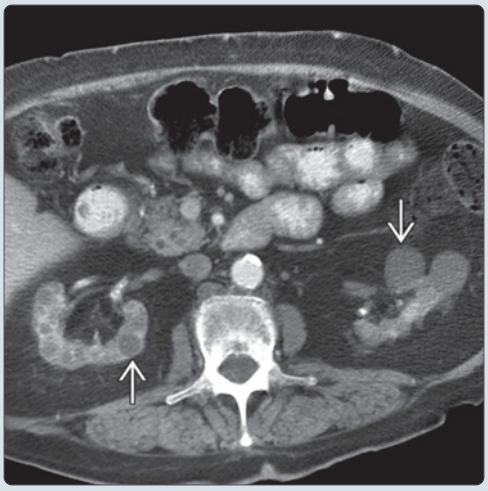
CLINICAL ISSUES

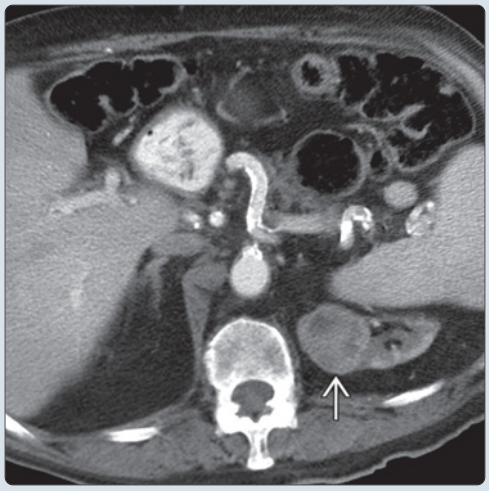
- Usually asymptomatic
  - Pain, hematuria in case of hemorrhage into cyst or cyst rupture
- Incidence increases with duration of dialysis
  - > 90% of patients will develop ACKD after 5-10 years of hemodialysis
- Complications
  - Hemorrhage into renal cysts
  - RCC
    - Prevalence: 3-6%
- Screening with ultrasound (controversial)





(Left) *Longitudinal ultrasound of the right kidney in a patient with end-stage renal disease shows an atrophic echogenic kidney with multiple varying size and anechoic cysts ⇨, an appearance compatible with uremic cystic disease.* **(Right)** *Coronal T2WI MR in a patient with end-stage renal disease and acquired cystic renal disease (ACRD) shows small, atrophic kidneys with multiple simple cysts ⇨.*





(Left) *Axial CECT in a patient with acquired renal cystic disease shows small kidneys replaced by innumerable cysts ⇨.* **(Right)** *Axial CECT in the same patient shows a heterogeneous enhancing mass ⇨ in the upper pole of the left kidney. The lesion is a biopsy-proven chromophobe renal cell carcinoma (RCC). The prevalence of RCC in patients with ACRD is 3-6%.*

TERMINOLOGY

Synonyms

- Acquired cystic kidney disease (ACKD)

Definitions

- ≥ 3 cysts per kidney in patients with end-stage renal disease and no history of hereditary cystic disease

IMAGING

General Features

- Best diagnostic clue
  - Multiple cysts of different size in atrophic kidneys
- Location
  - Bilateral
- Size
  - Kidneys
    - Small
    - Occasionally enlarged because of multiple cysts
  - Cysts
    - 0.5-3.0 cm

CT Findings

- NECT
  - Atrophic kidneys
  - Multiple (≥ 3) cysts in both kidneys
    - Fluid attenuation
    - Increased attenuation in case of hemorrhagic cysts
- CECT
  - Renal cell carcinoma (RCC) in 3-6% of cases
    - Uni- or multifocal
    - Enhancing solid renal mass

MR Findings

- T1WI
  - Simple cysts: ↓ signal intensity
  - Complicated (hemorrhagic) cysts: ↑ signal intensity
- T2WI
  - Simple cysts: ↑ signal intensity
  - Complicated (hemorrhagic) cysts: ↓ signal intensity

Ultrasonographic Findings

- Grayscale ultrasound
  - ↓ renal size
  - ↑ renal echogenicity
  - Multiple cysts
    - Anechoic
    - Internal echoes when complicated by debris/hemorrhage

Imaging Recommendations

- Best imaging tool
  - Ultrasound: Screening
  - CT or MR: Complications, diagnosis of RCC

PATHOLOGY

Gross Pathologic & Surgical Features

- Multiple cysts of various size
  - Single layer of hyperplastic epithelium
- Histological subtypes of RCC
  - Papillary (most common among the subtypes similar to sporadic RCC), clear cell, chromophobe
  - Acquired cystic disease-associated RCC (most common in ACKD), clear cell papillary renal cell carcinoma of end-stage kidney

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
  - Pain, hematuria in case of hemorrhage into cyst or cyst rupture

Demographics

- Gender
  - M:F = 3:1
- Epidemiology
  - Incidence increases with duration of dialysis
    - > 90% of patients will develop ACKD after 5-10 years of hemodialysis
    - 8-13% of end-stage renal disease patients have ACKD before dialysis
    - No correlation with cause of renal failure or method of dialysis

Natural History & Prognosis

- Complications
  - Hemorrhage into renal cysts
    - 50% of cases
    - May result in rupture and perinephric hematoma
  - RCC
    - Prevalence: 3-6%
    - Risk factors: Male gender, length of dialysis, kidney weight > 150 g
    - Metastatic in 15% of cases
  - Ureteral stones
- After transplantation
  - ↓ cystic disease
  - ↑ aggressiveness of RCC (↓ immune status)

Treatment

- Screening with ultrasound (controversial)
- Embolization or surgery for severe bleeding
- Nephrectomy for RCC

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von Hippel-Lindau Disease

KEY FACTS

TERMINOLOGY

- von Hippel-Lindau (VHL)
- Hereditary, autosomal dominant disease affecting several organs and associated with multiple neoplasms

IMAGING

- Renal manifestations
  - Renal cysts (59-63%)
    - Usually multiple and bilateral; may be simple or complex
  - Renal cell carcinoma (RCC) (25-45%)
    - Frequently multiple and bilateral

TOP DIFFERENTIAL DIAGNOSES

- Renal cyst
- Adult polycystic kidney disease
- Acquired cystic kidney disease of uremia
- Medullary cystic disease

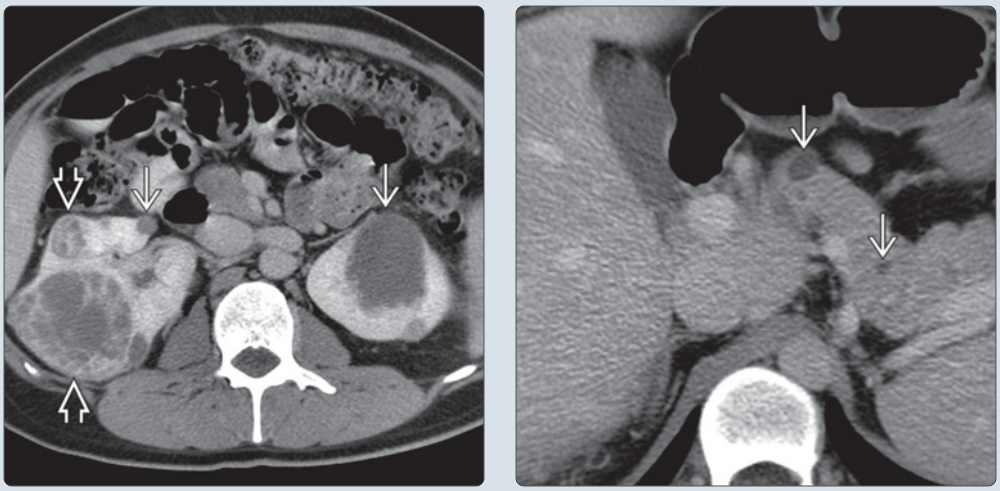
PATHOLOGY

- Rare, autosomal dominant disease with high penetrance
- Mutation of *VHL* tumor-suppressor gene on chromosome 3 (3p25-p26)
- Type 1: Pheocromocytoma absent
- Type 2: Pheochromocytoma present
- Benign cysts, atypical cysts, cystic RCC, solid RCC

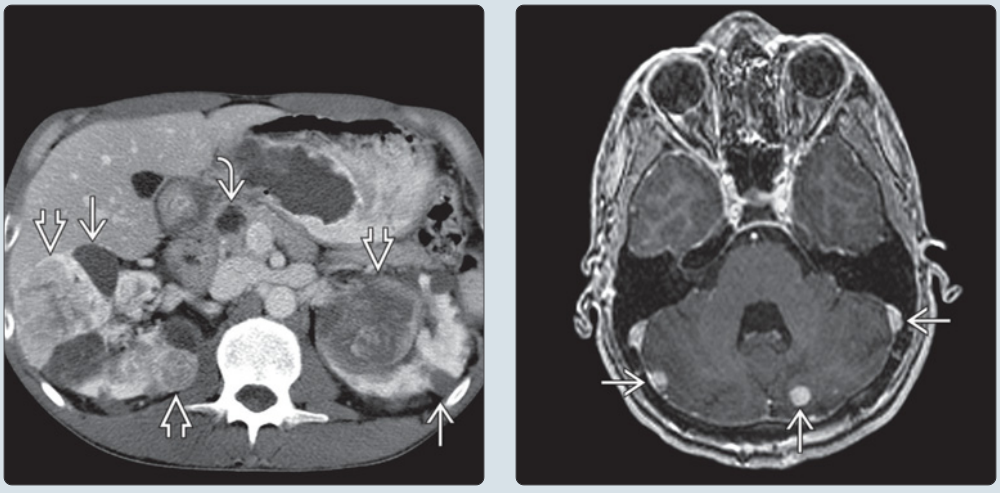
CLINICAL ISSUES

- Age: Infancy to 7th decade (average age 26 years)
- Prevalence: 1:30,000 - 1:50,000
- Natural history and prognosis
  - RCC responsible for 50% of deaths in VHL patients
- Screening for RCC
  - Imaging (ultrasound or MR) every year since age 16
- Treatment of RCC
  - Nephron-sparing surgery for RCC > 3 cm

(Left) *Axial CECT in a patient with von Hippel-Lindau (VHL) disease shows bilateral renal cysts and multiple complex cystic masses with enhancing nodules and septations that are compatible with cystic renal cell carcinomas (RCC). Renal cysts and RCCs are common visceral manifestations of VHL.* **(Right)** *Axial CECT in the same patient shows multiple pancreatic cysts.*



(Left) *Axial CECT in a patient with VHL shows multiple and bilateral renal cysts and RCCs. A cystic neoplasm is noted in the pancreatic head. Pancreatic manifestations of VHL include cysts, cystic neoplasms (microcystic adenoma), and neuroendocrine tumors.* **(Right)** *Axial T1 C+ MR shows multiple cerebellar enhancing lesions compatible with hemangioblastomas.*



TERMINOLOGY

Abbreviations

- von Hippel-Lindau (VHL)

Definitions

- Hereditary, autosomal dominant disease affecting several organs and associated with multiple neoplasms

IMAGING

General Features

- Renal manifestations
  - Renal cysts (59-63%)
  - Renal cell carcinoma (RCC) (25-45%)
- Extrarenal manifestations
  - Hemangioblastomas in central nervous system (13-72%)
  - Retinal hemangioblastoma (45-59%)
  - Endolymphatic sac tumor (2-11%)
  - Pancreatic lesions (17-56%)
  - Pheochromocytomas (0-60%)

CT Findings

- Renal cysts
  - Usually bilateral, multiple, and of various size
  - Simple cysts: Thin-walled, nonenhancing, near water density lesions
  - Complex cysts: Irregular, septate, minimally calcified, ↑ attenuation, no enhancement
- RCC
  - Usually bilateral and multiple
  - Solid enhancing mass or complex cystic mass + mural nodules &/or septa

MR Findings

- Renal cysts
  - Simple cysts: ↑ signal intensity (SI) on T2WI, ↓ SI on T1WI, no enhancement
  - Complex cysts: Varied SI on T1WI and T2WI
- RCC
  - Solid enhancing mass, complex cystic mass + mural nodules &/or septa

Ultrasonographic Findings

- Renal cysts: Usually anechoic
- RCC: Varied echogenicity

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR with renal mass protocol

DIFFERENTIAL DIAGNOSIS

Renal Cyst

- May be present in kidney with RCC without VHL

Adult Polycystic Kidney Disease

- Massively enlarged kidneys with innumerable cysts

Acquired Cystic Kidney Disease of Uremia

- Multiple cysts in small, atrophic kidneys

Medullary Cystic Disease

- Small kidneys with cysts in medulla

PATHOLOGY

General Features

- Genetics
  - Rare, autosomal dominant disease with high penetrance
  - Mutation of *VHL* tumor-suppressor gene on chromosome 3 (3p25-p26)

Staging, Grading, & Classification

- Type 1: Pheocromocytoma absent
- Type 2: Pheochromocytoma present

Gross Pathologic & Surgical Features

- Benign cysts, atypical cysts, cystic RCC, solid RCC
- Cysts
  - 3-30 (mean: 8 per kidney)
  - Usually < 1.5 cm (mean: 0.7 cm)
- RCCs
  - Histology: Clear cell type
  - Location: Bilateral (75%), multicentric (87%)
  - Metastases: 8-32%, in tumor > 3 cm, liver/lung/bone

CLINICAL ISSUES

Presentation

- Diagnostic criteria
  - > 1 hemangioblastoma in CNS or retina
  - 1 hemangioblastoma + visceral manifestation
  - Known family history + 1 of above manifestation

Demographics

- Age
  - Infancy to 7th decade (average: 26 years)
- Gender
  - M = F
- Epidemiology
  - Prevalence: 1:30,000-1:50,000

Natural History & Prognosis

- Common causes of mortality
  - RCC (responsible for 50% of deaths in VHL patients)
  - Neurologic complications

Treatment

- Screening for RCC
  - Imaging (ultrasound or MR) every year after age 16
- Nephron-sparing surgery for RCC

SELECTED REFERENCES

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Medullary Cystic Kidney Disease

KEY FACTS

TERMINOLOGY

- Group of related diseases with progressive renal tubular atrophy, secondary glomerulosclerosis, and medullary cyst formation

IMAGING

- Small, smooth kidneys with cysts in medulla
  - Small cysts: Microscopic to 1.5 cm
- Ultrasonography, coupled with clinical and laboratory findings, is usually sufficient for diagnosis

TOP DIFFERENTIAL DIAGNOSES

- Acquired cystic kidney disease
  - Atrophic kidneys, cysts mostly in cortex, history of dialysis
- Lithium-induced nephropathy
  - Normal-sized kidneys, multiple microcysts in cortex and medulla, history of lithium intake
- Autosomal dominant polycystic kidney disease
  - Large kidneys, multiple cysts of various size

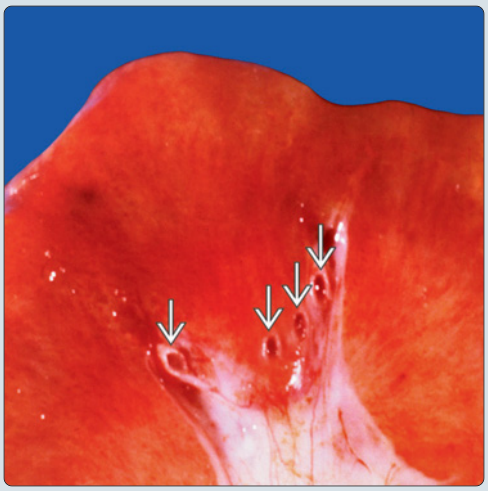
PATHOLOGY

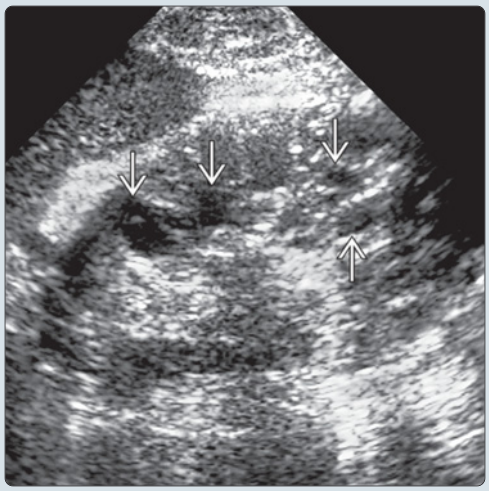
- Juvenile form of nephronophthisis (NPH) (type 1)
  - Onset around age 10
  - Inherited as autosomal recessive
  - Associated with many syndromes in various organ systems
  - Symptoms: Polyuria, polydipsia, growth retardation, anemia
- Medullary cystic kidney disease (MCKD)
  - Onset around age 30
  - Inherited as autosomal dominant trait
  - **Not** associated with extrarenal anomalies other than gout

CLINICAL ISSUES

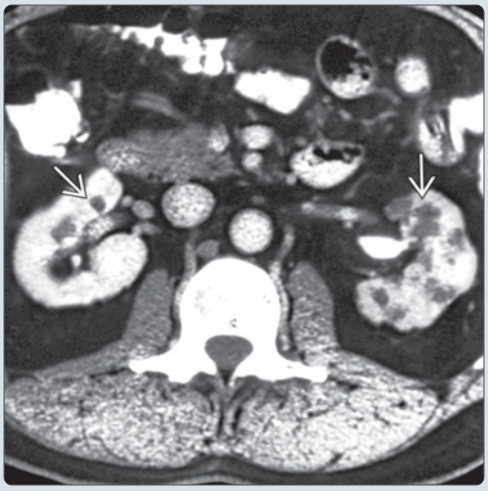
- End-stage renal failure within about 10-15 years
- Treatment: Renal transplantation; disease does not recur

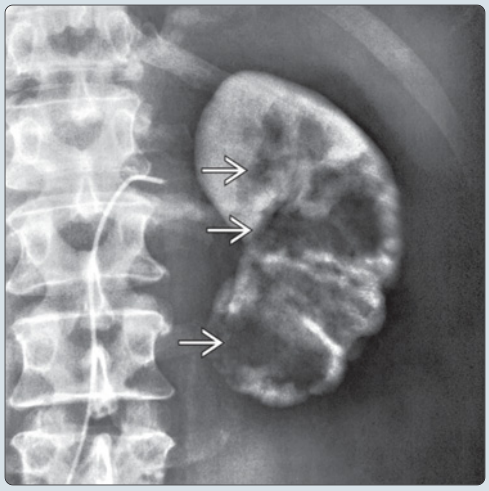
(Left) *Gross pathology section of a renal papilla from a patient with medullary cystic disease shows multiple small cysts ➡. Early in the disease, cysts may not be evident on imaging.* **(Right)** *Longitudinal ultrasound in a 42-year-old woman who presented with renal insufficiency shows a small left kidney with multiple medullary cysts ➡. Patients may present with electrolyte abnormalities (hyponatremia, hypokalemia) secondary to salt-wasting nephropathy.*





(Left) *Axial oblique CECT in a 25-year-old man with the adult form of medullary cystic disease shows a slight decrease of renal size bilaterally, with multiple small cysts ➡ predominantly located within the medulla or corticomedullary junction. The overlying renal cortex is thinned.* **(Right)** *Anteroposterior nephrographic phase of selective renal angiogram in the same patient shows multiple renal medullary cysts ➡ and thin renal cortex. The patient went on to progressive renal failure.*





TERMINOLOGY

Synonyms

- Nephronophthisis (NPH) or nephronophthisis complex
- Medullary cystic kidney disease (MCKD), types 1 and 2
- Familial juvenile nephronophthisis
- Cystic disease of renal medulla

Definitions

- Group of related diseases with progressive renal tubular atrophy, secondary glomerulosclerosis, and medullary cyst formation

IMAGING

General Features

- Best diagnostic clue
  - Small kidneys with cysts in medulla

Imaging Recommendations

- Best imaging tool
  - Ultrasonography, coupled with clinical and laboratory findings, is usually sufficient for diagnosis

CT Findings

- Small to normal-sized kidneys with cysts confined to medulla or corticomedullary junction

MR Findings

- Cysts are small and in medulla
- ↑ signal on T2WI, ↓ signal on T1WI

Ultrasonographic Findings

- Small to normal-sized kidneys
- Diffusely echogenic
- Multiple small cysts in medulla and corticomedullary junction
  - Cysts may be absent before renal failure

DIFFERENTIAL DIAGNOSIS

Acquired Cystic Kidney Disease

- Atrophic kidneys, cysts mostly in cortex, history of dialysis

Lithium-Induced Nephropathy

- Normal-sized kidneys, multiple microcysts in cortex and medulla, history of lithium intake

Autosomal Dominant Polycystic Kidney Disease

- Large kidneys, multiple cysts of various size

PATHOLOGY

General Features

- Genetics
  - Juvenile form of NPH (type 1)
    - Inherited as autosomal recessive trait
  - MCKD
    - Inherited as autosomal dominant trait
    - **Not** associated with extrarenal anomalies other than gout
    - Usually develops in adults aged 30-70
- Associated abnormalities
  - More common in childhood form of disease (NPH)
    - Associated with many syndromes (e.g., Joubert, Meckel-Gruber, and Senior-Loken)
  - Sometimes classified among broad group of diseases: Celiopathies
    - Associated with various diseases in many organ systems, including neurological and renal cystic diseases

Gross Pathologic & Surgical Features

- Early: Small to normal-sized kidneys with smooth contours
  - Progressive and chronic tubulointerstitial nephritis
  - Small cysts (0.1-1.5 cm diameter) in medulla and corticomedullary junction

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Symptoms in children: Polyuria, polydipsia, enuresis, growth retardation, anemia
    - Lethargy, fatigue, bone pain
  - Causes renal failure in children; less commonly presents in adulthood
    - Causes 10-25% of cases of renal failure in children
    - Most common genetic cause of end-stage renal disease in first 3 decades of life
  - Salt-wasting nephropathy: Loss of sodium and potassium in urine

Demographics

- Age
  - Juvenile nephronophthisis
    - Onset of disease at about age 10
  - Adult form (medullary cystic disease)
    - Onset around age 30, sometimes later

Natural History & Prognosis

- End-stage renal failure within about 10-15 years

Treatment

- Renal transplantation: Disease does not recur

SELECTED REFERENCES

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Lithium Nephropathy

KEY FACTS

TERMINOLOGY

- Spectrum of functional and structural renal damage caused by lithium ingestion
- Nephrogenic diabetes insipidus and chronic renal insufficiency are most common presentations

IMAGING

- Numerous nonenhancing, fluid intensity microcysts on MR in patient with history of chronic lithium use
  - Normal-sized kidneys, numerous (few to innumerable) microcysts (1-2 mm in diameter)
- On ultrasound microcysts can manifest as small anechoic lesions or tiny echogenic foci
- Imaging usually not needed for diagnosis

TOP DIFFERENTIAL DIAGNOSES

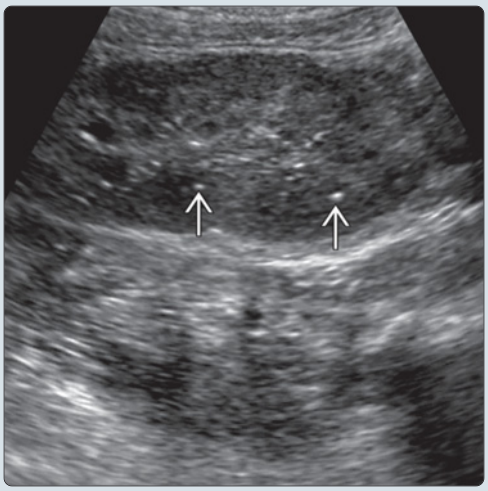
- Autosomal dominant polycystic kidney disease (ADPKD)
- Medullary cystic disease
- Acquired cystic disease of uremia

PATHOLOGY

- Distal tubular dilatation and microcysts (33-62%)
- Chronic interstitial nephritis, tubular atrophy

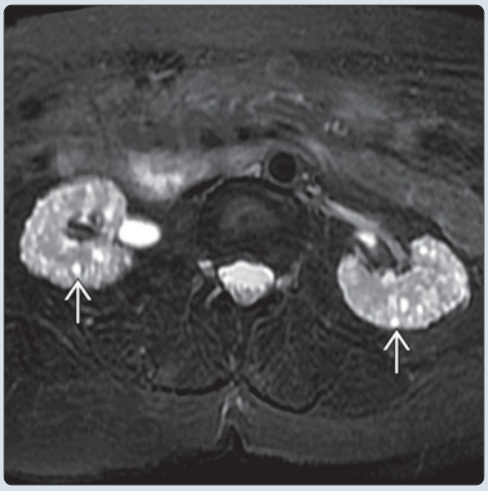
CLINICAL ISSUES

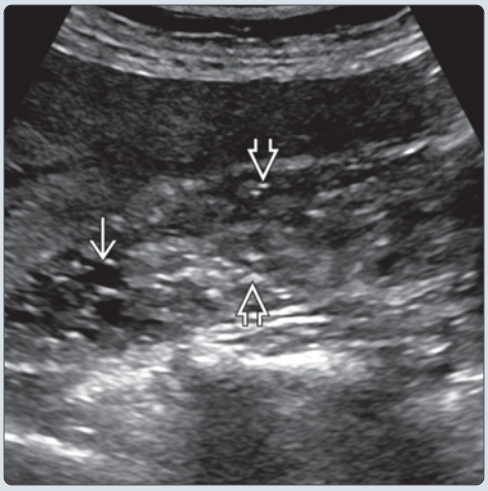
- Nephrotoxicity usually occurs within 1 month of onset of drug
- 20-54% of those taking lithium have urine-concentrating defects
- Renal insufficiency is less common
- Structural damage (microcysts) is correlated with degree of renal failure (↓ glomerular filtration rate)
- Diabetes insipidus is harmless and completely reversible by discontinuing drug
- Acute intoxication requires aggressive correction of fluid and electrolyte abnormalities ± dialysis





(Left) *Longitudinal US shows the right kidney of a 49-year-old woman with a history of long-term lithium intake. Scattered tiny echogenic foci ➡ are seen within the right kidney, compatible with lithium nephropathy. The microcysts may be too small to resolve as fluid-containing structures on ultrasound.* **(Right)** *Axial CECT in patient with lithium nephropathy shows innumerable bilateral tiny renal cysts ➡.*





(Left) *Axial fat-suppressed T2WI shows innumerable, small renal cysts ➡ in a 69-year-old woman with a history of bipolar disease and lithium intake compatible with lithium nephropathy.* **(Right)** *Longitudinal ultrasound of the right kidney in the same patient shows some renal cysts as small anechoic lesions ➡ and multiple other tiny echogenic foci ➡.*

TERMINOLOGY

Synonyms

- Lithium-induced nephropathy
- Lithium nephrotoxicity

Definitions

- Spectrum of functional and structural renal damage caused by lithium ingestion
- Can occur in context of suicidal attempts, elevated lithium level, or therapeutic lithium level
- Nephrogenic diabetes insipidus and chronic renal insufficiency are most common presentations

IMAGING

General Features

- Best diagnostic clue
  - Numerous nonenhancing, fluid intensity microcysts on MR in patient with history of chronic lithium use
- Location
  - Both in cortex and medulla
- Size
  - Normal-sized kidneys, numerous (few to innumerable) microcysts (1-2 mm in diameter)
- Morphology
  - Symmetric distribution of cysts in both kidneys

Imaging Recommendations

- Best imaging tool
  - T2WI MR is most sensitive tool to detect microcysts

MR Findings

- T1WI
  - Numerous nonenhancing hypointense cysts in cortex and medulla
- T2WI
  - Hyperintense cysts with characteristic fluid intensity

Ultrasonographic Findings

- Microcysts can manifest as small anechoic lesions or tiny echogenic foci

CT Findings

- Multiple small hypodense cysts

DIFFERENTIAL DIAGNOSIS

Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- Cysts are variable in size and signal intensity
- Kidneys are usually enlarged
- Cysts in other organs ± family history

Medullary Cystic Disease

- Cysts are located in medulla and corticomedullary junction
- Clinical: Salt-wasting nephropathy and renal insufficiency

Acquired Cystic Disease of Uremia

- Small kidneys with multiple cysts of varying size
- End-stage renal disease or dialysis

PATHOLOGY

General Features

- Etiology
  - Direct tubular damage by lithium
  - Microcysts are caused by dilatation and proliferation of distal tubular cells

Microscopic Features

- Distal tubular dilatation and microcysts (33-62%)
- Chronic interstitial nephritis, tubular atrophy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Polyuria, polydipsia, urine osmolarity < 100 mOsm/kg; normal or ↑ serum osmolarity (diabetes insipidus)
  - Increased risk of hypothyroidism, hyperparathyroidism, weight gain
- Other signs/symptoms
  - ↑ creatinine, ↑ BUN

Demographics

- Epidemiology
  - Nephrotoxicity usually occurs within 1 month of onset of drug
  - 20-54% of those taking lithium have urine-concentrating defects
  - Renal insufficiency is less common
  - Structural damage that causes microcysts is correlated with degree of renal failure (↓ glomerular filtration rate)

Natural History & Prognosis

- Diabetes insipidus is harmless and completely reversible by discontinuing drug
- Chronic renal disease is generally irreversible

Treatment

- Acute intoxication requires aggressive correction of fluid and electrolyte abnormalities ± dialysis
- Diabetes insipidus managed by thiazide diuretics and NSAIDs
- Chronic renal disease: Indication for discontinuing lithium; managed medically

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Localized Cystic Renal Disease

KEY FACTS

TERMINOLOGY

- Benign nonneoplastic proliferation of cysts, usually within normally functioning kidney

IMAGING

- Nonencapsulated collection of cysts of varying sizes separated by normal (or compressed) renal parenchyma
- Always unilateral
- Usually involves only part of kidney but can affect entire kidney
- Renal tissue in between cysts and in uninvolved portions of kidney functions, enhances normally
- Ipsilateral ureter normal

TOP DIFFERENTIAL DIAGNOSES

- Multilocular cystic nephroma
- Autosomal dominant polycystic disease
- Cystic renal cell carcinoma

PATHOLOGY

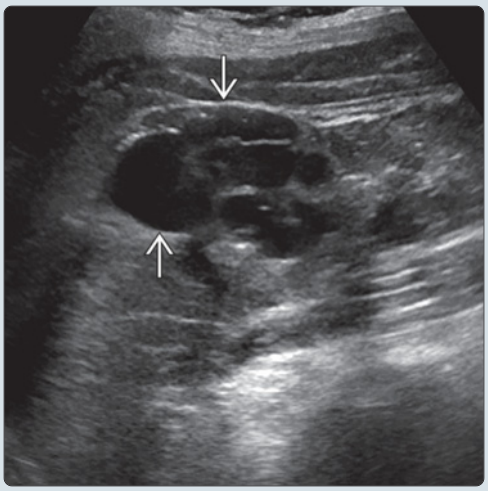
- Simple cysts with normal renal tissue in between

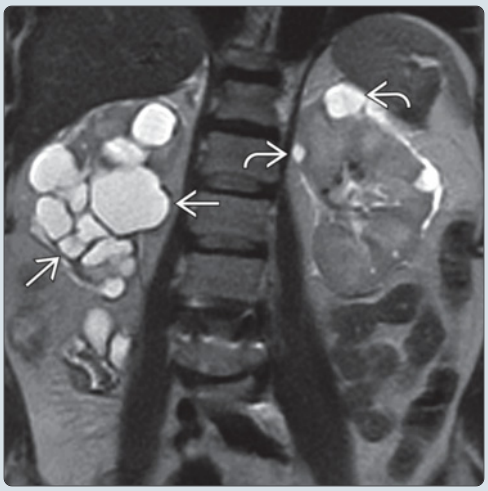
CLINICAL ISSUES

- Usually asymptomatic
- Occasionally flank pain or palpable mass, hematuria, hypertension
- Young adults to elderly (mean age at presentation: 54 years)
- Benign, stable, or slowly progressive
- Conservative management; imaging follow-up

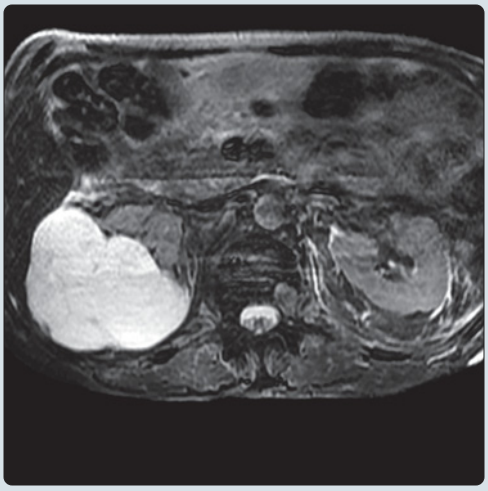
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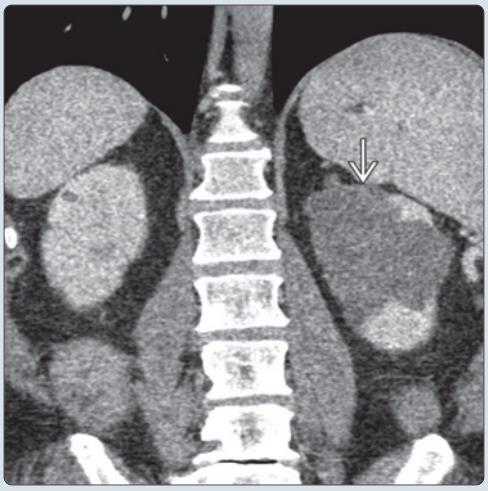
- Important to not label patient as having an inherited, progressive, or neoplastic disease
- Key is to distinguish from multilocular cystic nephroma and cystic renal cell carcinoma
  - Both of these have irregular cystic spaces surrounded by thick capsule





(Left) *Longitudinal ultrasound through the right kidney shows a cluster of multiple cysts. Each cyst has a typical benign appearance.* **(Right)** *Coronal T2WI MR in the same patient shows a cluster of cysts in the lower pole of the right kidney. Absence of a capsule is in keeping with localized cystic renal disease. Note scattered left renal cysts.*





(Left) *Axial fat-suppressed T2WI MR shows a cluster of high signal intensity cysts in the right kidney without a capsule, compatible with localized cystic renal disease.* **(Right)** *Coronal CECT demonstrates the lack of a capsule associated to the collection of cysts, consistent with localized cystic renal disease.*

TERMINOLOGY

Abbreviations

- Localized cystic renal disease (LCRD)

Synonyms

- Segmental cystic disease
- Unilateral cystic disease of kidney

Definitions

- Benign nonneoplastic proliferation of simple cysts, usually within normally functioning kidney

IMAGING

General Features

- Best diagnostic clue
  - Nonencapsulated collection of cysts of varying sizes separated by normal (or compressed) renal parenchyma with normal renal function
- Location
  - Always unilateral
  - Usually involves only part of kidney but can affect entire kidney
- Size
  - Cysts are of varying size; remainder of kidney is of normal size

Imaging Recommendations

- Best imaging tool
  - CECT with multiplanar reformations or MR

CT Findings

- Nonencapsulated collection of numerous cysts, which are individually typical simple cysts
- Renal tissue in between cysts and in uninvolved portions of kidney functions and enhances normally
  - Ipsilateral ureter is normal

MR Findings

- Same as CT, with each cystic component having appearance of simple cysts
  - ↓ signal intensity on T1WI; ↑ signal intensity on T2WI

DIFFERENTIAL DIAGNOSIS

Multilocular Cystic Nephroma

- Neoplasm with thick capsule enclosing multilocular cystic spaces
  - No normal renal tissue between cystic components
- Mass replaces parenchyma, invaginates into renal pelvis
- Cystic spaces do not resemble simple cysts

Autosomal Dominant Polycystic Disease

- Affects both kidneys, causing enlargement and progressive loss of function
- Inherited, multisystem, progressive disease
- Cysts in multiple organs (liver, pancreas, seminal vesicles, etc.)

Cystic Renal Cell Carcinoma

- Encapsulated neoplasm resembling parenchyma
- Cystic spaces do not resemble simple cysts

PATHOLOGY

General Features

- Genetics
  - Acquired, not inherited

Gross Pathologic & Surgical Features

- Simple cysts with normal renal tissue in between
  - No capsule
  - Involve only part of kidney (rarely entire kidney)
  - Cuboidal epithelium lining
- A few cysts may be present in contralateral kidney

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
  - Occasionally flank pain or palpable mass, hematuria, hypertension

Demographics

- Age
  - Young adults to elderly (mean age at presentation: 54 years)
- Gender
  - M:F ratio = 5:4
- Epidemiology
  - Rare

Natural History & Prognosis

- Benign, stable, or slowly progressive

Treatment

- Conservative management; imaging follow-up
- No resection

DIAGNOSTIC CHECKLIST

Consider

- Important to not label patient as having inherited, progressive, or neoplastic disease

Image Interpretation Pearls

- Key is to distinguish from multilocular cystic nephroma and cystic renal cell carcinoma
  - Both have irregular cystic spaces surrounded by thick capsule

SELECTED REFERENCES

1. Wood CG 3rd et al: CT and MR Imaging for Evaluation of Cystic Renal Lesions and Diseases. Radiographics. 35(1):125-41, 2015
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Renal Angiomyolipoma

KEY FACTS

TERMINOLOGY

- Benign renal tumor composed of abnormal blood vessels, smooth muscle, and adipose components

IMAGING

- Most common benign renal neoplasm
- 80% are incidental findings on imaging
- 90% are unilateral and solitary: Usually not associated TSC
- 10% are multiple and bilateral: Usually due to TSC
  - 80% of TSC patients have renal AMLs
- Solid, heterogeneous, renal cortical mass in adult with macroscopic fat is a reliable sign of AML
  - Variable amounts of fat may be present
  - ~ 5% of AMLs contain minimal fat and cannot be reliably diagnosed by imaging
- Lipid-poor AMLs are often hyperdense on NECT

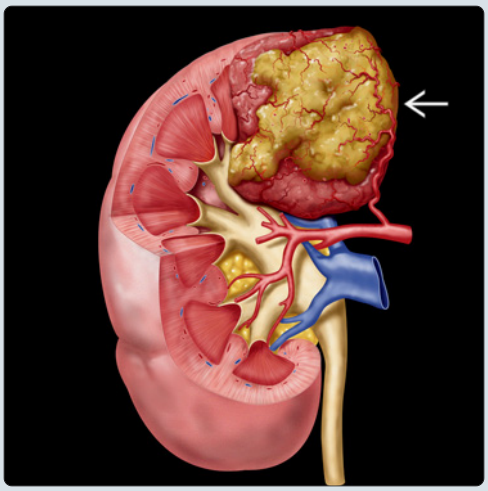
TOP DIFFERENTIAL DIAGNOSES

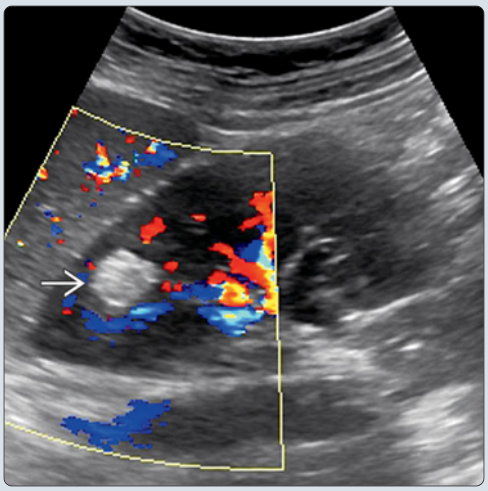
- Renal cell carcinoma (RCC)
  - Rarely reported to contain fat (engulfed sinus or perirenal fat)
  - Calcification/ossification within tumor is highly suggestive of RCC
- Perirenal liposarcoma
  - Smooth compression of kidney and extension beyond perirenal space favor liposarcoma
  - Renal parenchymal defect and enlarged vessels favor angiomyolipoma
- Wilms tumor
  - If child has TSC, mass more likely to be AML
  - Otherwise suspect Wilms tumor
- Other less common considerations: Metastases, lymphoma, oncocytoma, lipoma, teratoma

CLINICAL ISSUES

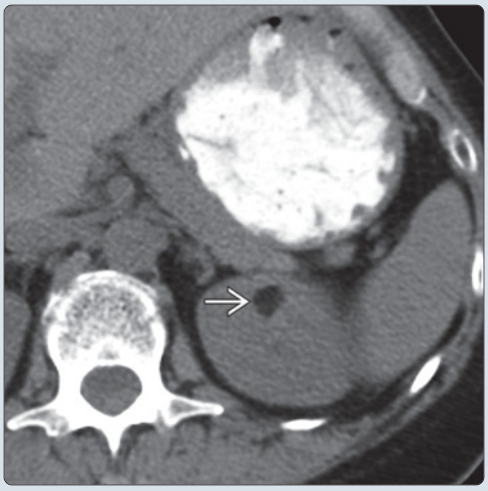
- Most common complication: Hemorrhage
- Small subgroup of AMLs can be angioinvasive, affect local lymph nodes and rarely metastasize

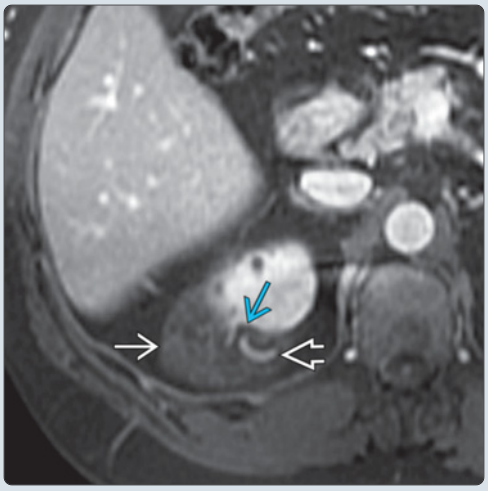
(Left) *Graphic shows a vascular renal mass with prominent fatty and soft tissue components. Note the large "feeding" arteries. This hypervascularity predisposes these tumors to spontaneous hemorrhage.* **(Right)** *Sagittal US shows an echogenic lesion in the superior cortex of the left kidney due to an incidental AML. Color flow was not identified in the lesion.*





(Left) *Axial NECT in the same patient demonstrates a fat attenuation lesion in the superior cortex of the left kidney, consistent with an incidental AML.* **(Right)** *Axial T1 post-contrast MR with fat saturation reveals an AML that extends into the perinephric space. A cortical defect ("notch" sign) identifies the origin of the AML from the kidney. A large vessel also extends into the neoplasm from the kidney.*





TERMINOLOGY

Abbreviations

- AML

Synonyms

- Renal hamartoma or choristoma

Definitions

- Benign renal neoplasm composed of abnormal blood vessels, smooth muscle, and adipose components

IMAGING

General Features

- Best diagnostic clue
  - Solid, heterogeneous, renal cortical mass containing macroscopic fat in an adult
- Size
  - Variable: Few mm to > 25 cm
- Other general features
  - Most common benign tumor of kidney
  - 80% are incidental findings on imaging
  - 90% are unilateral and single: Usually not associated with tuberous sclerosis complex (TSC)
  - 10% are multiple and bilateral: Usually due to TSC
  - 20% of patients with renal AML have TSC
  - 80% of TSC patients have renal AMLs
  - Tuberous sclerosis complex (Bourneville disease)
    - Facial angiofibroma, ungual fibroma
    - Retinal hamartoma
    - Cortical tubers and subependymal nodules
    - Multiple renal angiomyolipomas
    - Lymphangioleiomyomatosis (LAM)
      - Progressive cystic lung disease

Radiographic Findings

- IVP
  - Small tumor: Well-defined radiolucent defect
  - Large tumor: Distortion of collecting system
  - Multiple lucent defects when associated with tuberous sclerosis

CT Findings

- NECT
  - Well-marginated, heterogeneous mass with macroscopic fat arising from renal cortex
    - "Notch" sign: AML originates from triangular or rectangular notch-like defect in cortex
  - Solid renal mass with predominance of macroscopic fat is a reliable sign of AML
    - Tissue attenuation of -10 HU or lower is reliable indicator of fat
      - Specificity of fat detection increases with use of lower attenuation values
    - However, amount of fat is variable
    - ~ 5% of AMLs contain minimal fat and cannot be reliably diagnosed by CT, pixel analysis, or MR
  - Minimal fat AMLs are often hyperattenuating relative to normal renal parenchyma on NECT
    - Not sufficient to allow diagnosis due to overlap with RCCs
    - CT histogram analysis and pixel mapping have not reliably been able to differentiate minimal fat AMLs from RCCs
  - Hemorrhage more likely in large (≥ 4 cm) AMLs
  - Rare calcification; if present, suspect RCC
- CECT
  - Varied enhancement pattern based on tumor composition
  - Without macroscopic fat, enhancement alone does not differentiate AML from renal tumors
    - Minimal fat AMLs typically show more homogeneous and prolonged enhancement than RCCs during nephrographic phase
    - However, differentiation remains difficult and biopsy of small, solid minimal fat lesions should be considered
  - Invasion of renal vein/IVC by AML is extremely rare
  - Hemorrhage from ruptured AML can extend into perinephric space and renal hilum
    - Extensive hemorrhage may obscure AML on initial CT
- CTA
  - Disorganized tumoral vessels, aneurysms, and pseudoaneurysms may be seen within and around tumor

MR Findings

- AML typically heterogeneous signal intensity
  - Variable proportion of vessels, muscle, and fat
  - Macroscopic fat: High signal intensity on T1WI and T2WI
  - Acute hemorrhage
- MR techniques for localization of fat
  - Fat suppression (frequency selective): Loss of signal
  - Opposed-phase imaging
    - Signal loss on out of phase images
      - Not specific; can occur in various types of renal neoplasms
    - India ink artifact: Signal loss at fat-water interfaces
    - Chemical shift artifact: Alternating high and low signal bands on out of phase images
- Caveat: Clear cell RCC also lose signal on out of phase sequence
  - However, clear cell RCCs lack India ink artifact and do not lose signal on conventional T1 fat-suppressed sequences
- T1 C+
  - Variable enhancement due to tumor composition
    - Hypointense compared to renal parenchyma
    - Signal intensity similar to hypovascular RCCs
  - In cases without macroscopic fat, enhancement cannot reliably differentiate AML from malignant and other benign renal lesions
- DWI
  - Not useful for differentiating AML from other renal lesions
- Minimal fat AMLs (~ 5% of all AMLs)
  - Often indistinguishable from RCC
  - Signal decrease with opposed-phase imaging for tissues containing intracellular lipid
    - Nonspecific: Opposed-phase signal decrease also observed in various benign and malignant renal cortical tumors
  - Smaller size and low T2 signal intensity compared to clear cell RCC

Ultrasonographic Findings

- Grayscale ultrasound
  - Classical appearance: Well-defined, markedly hyperechoic mass relative to normal renal parenchyma with acoustic shadowing
    - However, a subset of minimal fat AMLs are isoechoic to slightly hypoechoic and lack shadowing
    - Minimal fat AMLs are typically homogeneously isoechoic
  - Small renal cell carcinoma may also be hyperechoic
    - AML and RCC can be difficult to distinguish US
    - Further evaluation with CT, MR, or biopsy may be considered
- Color Doppler
  - Pseudoaneurysm detection
    - Color-filled ovoid or round structures within or adjacent to AML
    - Color flow appears as swirling or "yin and yang" sign
    - Classic "to and fro" pattern during spectral analysis at neck of pseudoaneurysm
- Power Doppler
  - Most AMLs show "intratumoral focal pattern"
    - Central, internal flow that does not extend to margins of tumor
  - Conversely, most small RCCs have mixed peripheral and penetrating pattern
    - Flow present in center and extends to margins/periphery of tumor

Angiographic Findings

- Tumor with increased vascular component
  - Highly vascular mass with disorganized, long, and tortuous vessels
  - Sacculated pseudoaneurysms
  - Absence of arteriovenous shunts
  - "Sunburst" appearance of capillary nephrogram
  - "Onion peel" appearance of peripheral vessels in venous phase

Imaging Recommendations

- NECT thin sections are best to detect subtle fat within tumor
- Multiplanar CT or MR best display full extent of mass

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- Rarely reported to contain fat
  - May be due to engulfed sinus/perirenal fat, tissue necrosis, or osseous metaplasia
- However, ~ 30% of RCCs < 3 cm mimic AMLs on US
- If AML is fat deficient, often mimic renal cell carcinoma
- Calcification or ossification within a mass is highly suggestive of RCC
- US characteristics favoring RCC over AML
  - Hypoechoic rim
  - Intratumoral cysts
  - Lack of acoustic shadowing
  - Heterogeneity

Perirenal Liposarcoma

- Large, exophytic AML may simulate retroperitoneal liposarcoma
  - Renal cortical defect ("notch" sign) and enlarged vessels favor AML
  - Smooth compression of kidney and extension beyond perirenal space favor liposarcoma
- Very rarely liposarcoma originates within kidney
  - Typically subcapsular in location, lentiform in shape

Wilms Tumor

- Pediatric renal tumor that may contain fat
- If child has tuberous sclerosis, mass more likely to be AML
  - Otherwise suspect Wilms tumor

Renal Metastases and Lymphoma

- Usually multiple soft tissue lesions
- Usually occur in conjunction with extrarenal disease

Renal Oncocytoma

- Well-defined mass of homogeneous soft tissue density
  - Rarely contains foci of fat
- Central stellate scar is suggestive of oncocytoma

PATHOLOGY

General Features

- Etiology
  - Hamartoma: Benign tumor consisting of abnormal amount of tissues that normally occur in organ of origin
  - Choristoma: Benign tumor composed of tissues not normally found in organ of origin
    - For renal AML, choristoma is a more accurate pathologic descriptor than hamartoma
- Associated abnormalities
  - Tuberous sclerosis; lymphangioleiomyomatosis
  - Epithelioid AMLs are a rare, aggressive subgroup
    - Highly cellular tumors comprised of atypical epithelioid cells with little to no fat
    - Typically large (> 9 cm) tumors affecting females (80%)
    - Stain strongly for melanoma-associated markers
    - ~ 33% are locally invasive or metastatic at presentation
      - Larger tumors are more likely to metastasize

Gross Pathologic & Surgical Features

- Spherical, lobulated, yellow-to-gray color secondary to fat content
- Numerous tortuous arteries within tumor
  - Arterial walls have disorganized smooth muscle and lack elastic membrane
  - Arteries are therefore prone to aneurysms and rupture
- Large tumors recruit additional blood supply from adjacent arteries
- Minimal fat AMLs (~ 5% of all AMLs) contain only microscopic fat

Microscopic Features

- Variable amounts of angioid (vascular), myoid (smooth muscle), and lipoid (fatty) components

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often asymptomatic, incidental imaging finding
  - Hematuria, flank pain or palpable mass
  - Acute pain (spontaneous hemorrhage, rupture)
  - Occasionally hypertension and chronic renal failure
    - Usually when most of 1 or both kidneys replaced by tumor
  - Detected while screening tuberous sclerosis patients for renal cell carcinoma

Demographics

- Age
  - Usually > 40 years
- Gender
  - M:F = 1:4
  - AML associated with tuberous sclerosis (M:F = 1:1)
- Epidemiology
  - 0.3-3% in autopsy series
  - 80% isolated (sporadic) AML
  - 20% of AMLs are associated with tuberous sclerosis

Natural History & Prognosis

- Complications
  - When AML ≥ 4 cm, 70-80% of patients experience symptoms
    - Most common: Hemorrhage/rupture, pain, mass effect
  - Typically benign behavior, but rare subset can be aggressive
    - Locally: Renal vein/IVC invasion ± adenopathy
    - Epithelioid AML: ~ 33% are locally aggressive or metastasize
- Prognosis
  - Most AML are indolent and asymptomatic
  - Favorable prognosis with treatment
  - Less favorable prognosis if presenting with hemorrhage, rupture

Treatment

- Debate exists on how to manage asymptomatic patients with AML
- Asymptomatic with tumor size < 4 cm
  - Conservative; imaging follow-up every 6-12 months
- Symptomatic or tumor size ≥ 4 cm
  - Catheter embolization, partial nephrectomy, or enucleation can be considered
  - Success rate of > 90%
  - Patients with TS are more likely to require treatment
  - Embolization materials include ethanol, polyvinyl alcohol particles, embospheres, coils, foam
- Patients presenting with hemorrhage are typically treated with embolization initially
- Postembolization syndrome: Fever, nausea, vomiting, pain
  - Occurs in majority of patients and generally resolves by 72 hours post procedure
- Radiofrequency ablation has been performed in small lesions

DIAGNOSTIC CHECKLIST

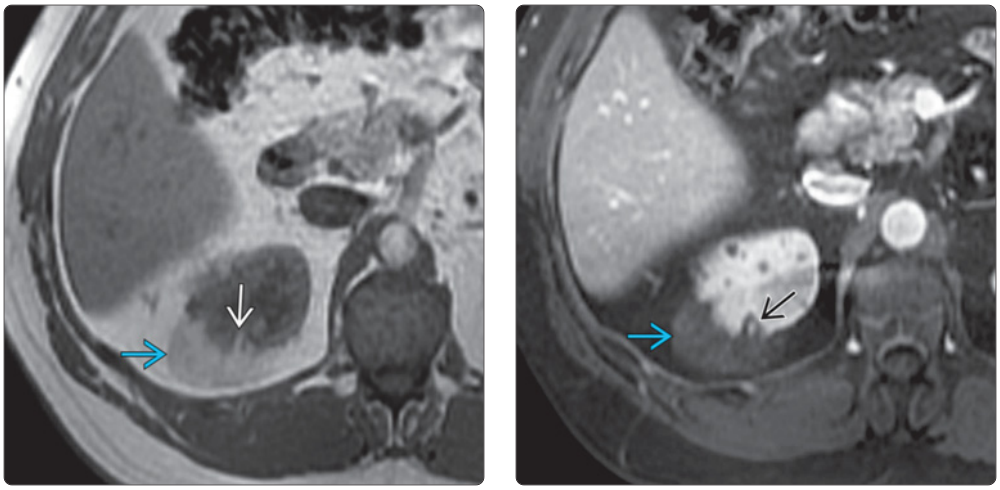
Image Interpretation Pearls

- Well-marginated, heterogeneous renal mass containing macroscopic fat
- "Notch" sign: AML originates from small, geographic defect in renal cortex
- Opposed-phase imaging
  - Signal loss of out of phase sequence
  - India ink artifact at interface with kidney
  - Chemical shift artifact: Alternating high and low signal intensity bands on out of phase images
- Calcification or ossification within tumor highly suggestive of renal cell carcinoma

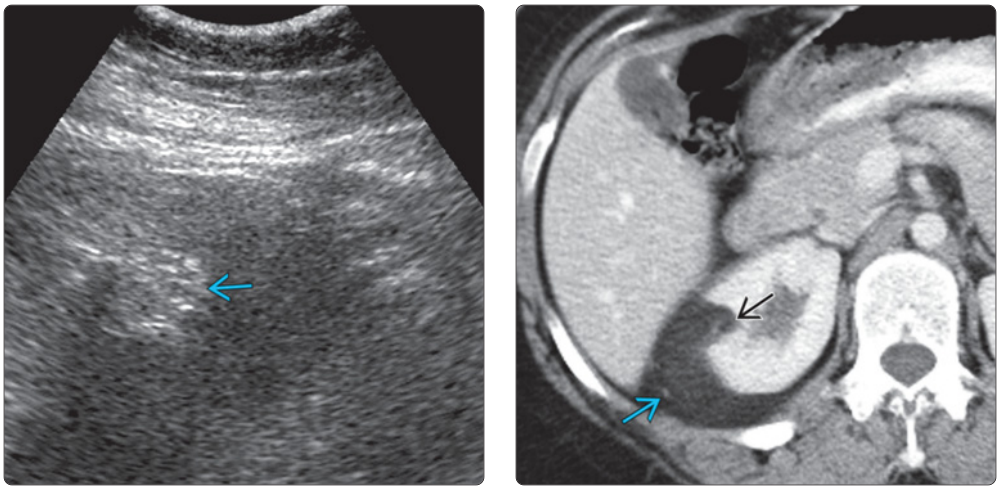
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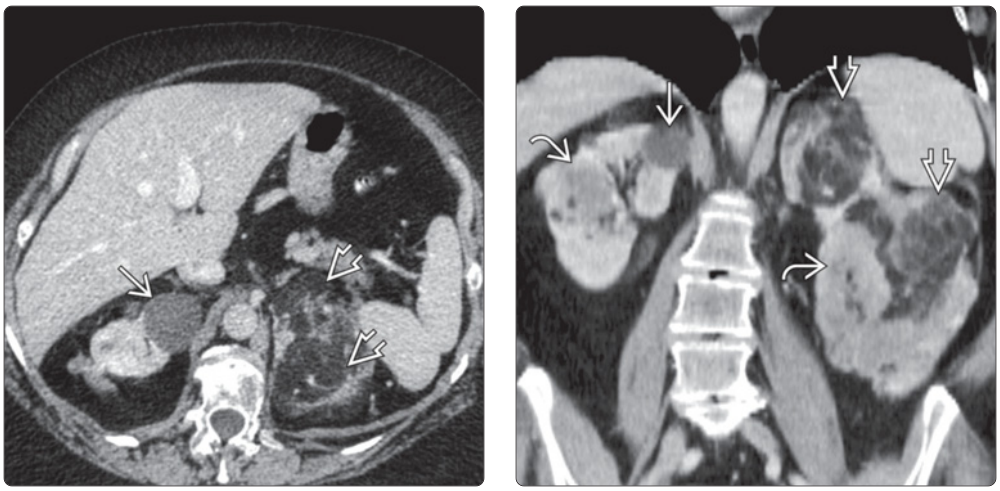
(Left) *Axial T2 MR shows a lesion ⇨ with signal intensity similar to fat arising from the posterior renal cortex via a small defect ➡. (Right) Axial post-contrast T1 MR with fat suppression in the same patient shows that the lesion ⇨ loses signal intensity with application of fat saturation. A cortical defect ("notch" sign) and a feeding vessel ⇨ are shown.*

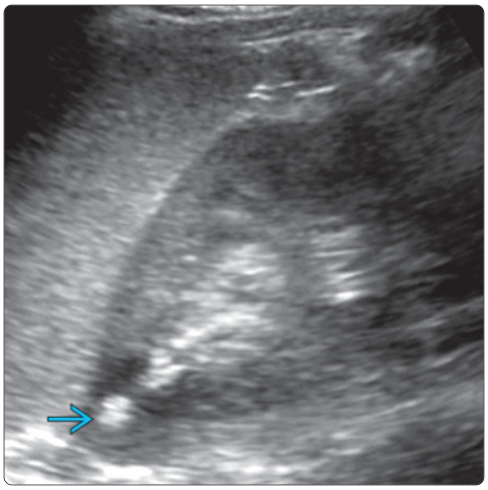


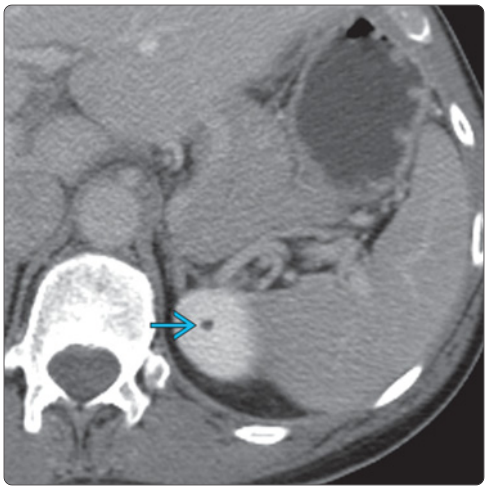
(Left) *Sagittal US of the right kidney shows a round, echogenic lesion ⇨ within the posterior cortex.* **(Right)** *Axial CECT in the same patient shows the cortical defect ⇨ from which the AML ⇨ originates and extends into the perinephric space.*



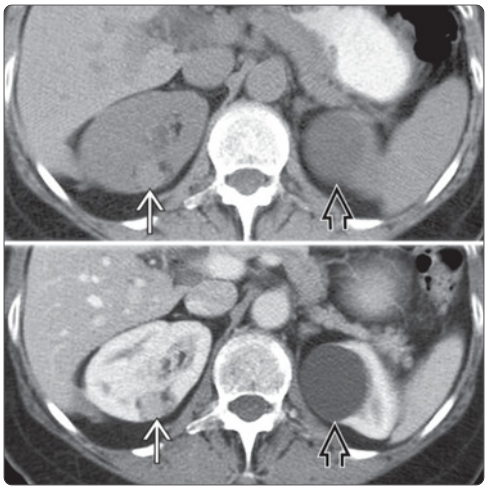
(Left) *Axial CECT in a 61-year-old woman with tuberous sclerosis shows bilateral renal masses, some of which contain obvious fat ➡ (due to AML), while others represent cysts ➡.* **(Right)** *Coronal CECT reformation in the same patient shows multiple bilateral cysts ➡ and solid masses (AMLs) with both fatty ➡ and soft tissue density ➡ components. The presence of multiple and bilateral AMLs is strong evidence for tuberous sclerosis.*

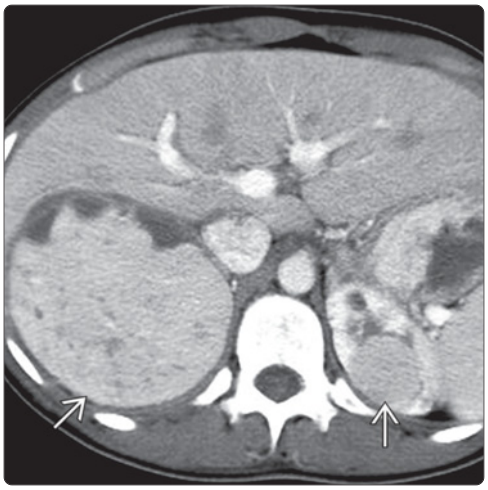




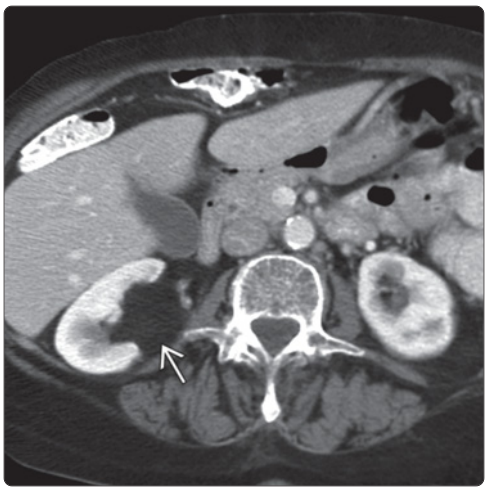


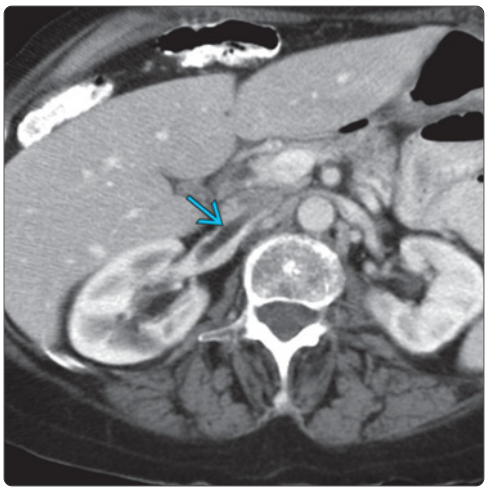
(Left) *Sagittal US shows an echogenic lesion ⇨ at the superior pole of the kidney, consistent with an angiomyolipoma.* **(Right)** *Axial post-contrast CT shows a fat attenuation lesion ⇨ in the superior pole of the left kidney that corresponds to the echogenic lesion noted on prior US, consistent with angiomyolipoma.*





(Left) *Axial NECT and CECT show subtle foci of fat within multiple AMLs in the right kidney ⇨. Note the simple cyst in the left kidney ⇨.* **(Right)** *Axial CECT shows bilateral enhancing renal masses ⇨ in a patient with tuberous sclerosis; AMLs without obvious fat component.*





(Left) *Axial contrast-enhanced CT shows an AML ⇨ in the right renal hilum.* **(Right)** *Axial contrast-enhanced CT in the same patient reveals fat attenuation in the lumen of the right renal vein ⇨, consistent with renal vein invasion by AML.*

Renal Oncocytoma

KEY FACTS

TERMINOLOGY

- Benign renal tumor composed of eosinophilic epithelial cells, arising from collecting ducts

IMAGING

- Solid renal cortical mass lesion ± central stellate scar
- Dynamic post-contrast imaging is highly variable and depends on degree of cellularity and stroma
- Absence of malignant features
  - No invasion of vessels, perinephric fat, or collecting system
  - No adenopathy or metastases
- Other general features
  - 2nd most common benign renal tumor after angiomyolipoma (AML)
  - Accounts for 3-7% of all renal cortical neoplasms
  - ~ 3-16% are multifocal and 4-14% are bilateral

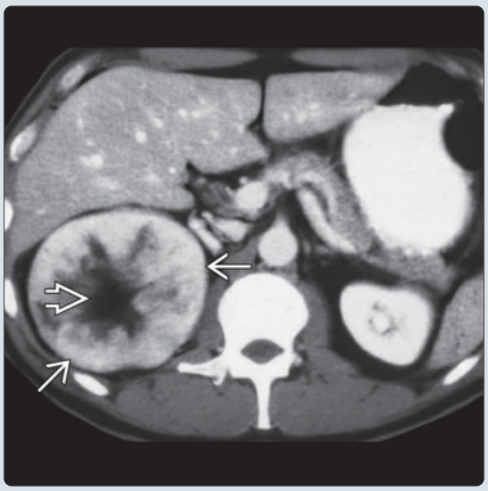
TOP DIFFERENTIAL DIAGNOSES

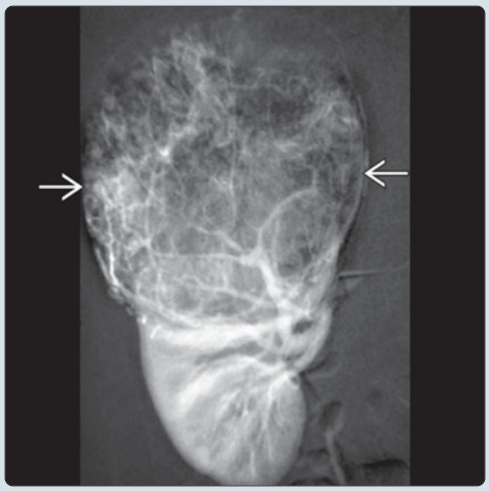
- Renal cell carcinoma
  - Necrotic, calcific, hemorrhagic, cystic components often seen in RCC, but rare or absent in oncocytoma
  - Central necrosis of RCC may mimic central scar of oncocytoma
- Renal angiomyolipoma
  - Well-defined cortical heterogeneous tumor, with foci of fat density (-30 to -100 HU)
  - CTA: Aneurysmal renal vessels may be seen
- Renal metastases and lymphoma
  - Solid, hypoattenuating masses showing moderate enhancement
  - Usually known primary tumor and metastases in other sites strongly suggest renal metastases

DIAGNOSTIC CHECKLIST

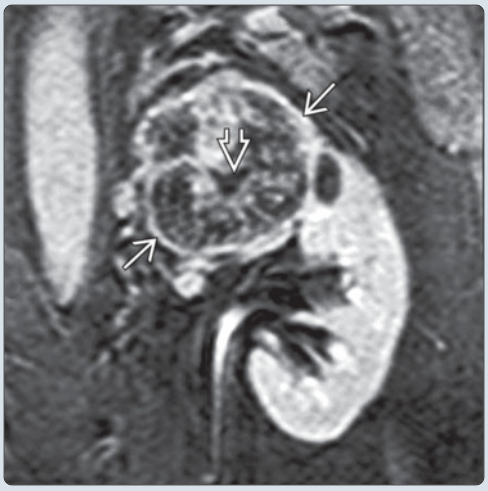
- Not possible to distinguish from RCC on imaging alone
- May suggest diagnosis → nephron-sparing surgery

(Left) *Axial CECT shows an exophytic, solid, enhancing mass ⇨ extending from the superior pole of the right kidney. Note the large central scar ⇨. The imaging and pathologic findings of renal oncocytoma are analogous to those seen with hepatic focal nodular hyperplasia, another benign homogeneous, solid mass with a central scar.* **(Right)** *Digital subtraction angiography in the same patient shows hypervascularity of the large mass ⇨. Note the absence of bizarre tumor neovascularity that is often seen with RCC.*





(Left) *Coronal T1WI C+ MR demonstrates a well-defined, spherical renal mass ⇨ with a pseudocapsule and a central scar ⇨, findings that are characteristic, but not diagnostic, of an oncocytoma. Some variants of renal cell carcinoma share many imaging features and some histologic features with oncocytomas.* **(Right)** *Surgical photograph shows a resected oncocytoma with a central scar ⇨ and thin septa within a homogeneous, sharply marginated, solid mass.*





TERMINOLOGY

Definitions

- Benign renal tumor composed of eosinophilic epithelial cells, arising from intercalated cells of collecting ducts

IMAGING

General Features

- Best diagnostic clue
  - Solid renal cortical mass lesion with central stellate scar
- Size
  - 3-10 cm (average: 7 cm)
- Morphology
  - Encapsulated, well marginated, smooth contour
- Other general features
  - 2nd most common benign renal tumor after angiomyolipoma (AML)
    - Accounts for 3-7% of all renal cortical neoplasms
    - ~ 3-16% are multifocal and 4-14% are bilateral
  - 50-90% of tumors are incidental findings (asymptomatic)
  - Oncocytic tumors also occur in thyroid, parathyroid, adrenal, and salivary glands

CT Findings

- NECT
  - Well-defined, homogeneous, soft tissue mass
  - Isodense or slightly hyperdense relative to kidney
  - Central hypoattenuating area suggestive of scar
    - Seen in 33-54% of cases (in lesions usually > 5 cm)
    - Mimicked by central necrosis of renal cell carcinoma (RCC)
  - Rarely: Calcification, necrosis, cystic change, hemorrhage
  - Occasionally multifocal and bilateral
- CECT
  - Degree and timing of enhancement are variable due to variable cellularity
  - Potential enhancement patterns
    - Intense arterial enhancement then washout
      - Due to high cellularity
    - Segmental enhancement inversion
      - Due to areas of adjacent high and low cellularity
      - "Flip-flop" of arterial and excretory phase enhancement
    - Arterial enhancement persisting in nephrographic phase
      - High vascularity and edematous stroma
    - Hypovascularity with gradual enhancement
      - Low cellularity and edematous stroma
    - Hypovascularity in corticomedullary and excretory phases
      - Cystic changes and hemorrhage
  - Absence of malignant features
    - No adenopathy
    - No vascular, collecting system or perinephric invasion
      - Rare vascular invasion has been reported, but does not affect benign clinical behavior6

MR Findings

- T1WI
  - Iso to low signal intensity (SI)
- T2WI
  - Intermediate SI: Highly cellular components
  - ↑ SI: Edematous stroma, central scar, cystic areas
- DWI
  - ↑ SI in highly cellular components
  - ↑ apparent diffusion coefficient (ADC) values
    - Not useful to differentiate from RCC
- T1WI C+
  - Dynamic post-contrast findings are similar to CECT

Ultrasonographic Findings

- Grayscale ultrasound
  - Well-defined, homogeneous, hypo-/isoechoic mass
  - Central scar: Usually echogenic
- Color Doppler
  - Internal flow ± central radiating vessels

Angiographic Findings

- No angiographic pattern is pathognomonic
- Classic "spoke-wheel" pattern is rare
- Sharp demarcation from kidney
- In contrast to RCC
  - Absence of bizarre neoplastic vessels
  - No arteriovenous shunting or renal vein invasion

Nuclear Medicine Findings

- FDG PET: Less FDG uptake than RCCs
  - Usually isointense to renal tissue

Imaging Recommendations

- Contrast-enhanced CT or MR with multiplanar imaging

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- Most common primary renal malignant neoplasm
- Necrotic, calcific, hemorrhagic, cystic components often seen in RCC; rare or absent in oncocytoma
  - Central necrosis of RCC may mimic central scar of oncocytoma
- CECT
  - Clear cell RCC: Hypervascular mass with thick septa and nodularity
  - ± invasion of vessels perinephric fat, collecting system
  - ± regional lymphadenopathy and hypervascular metastases
  - Chromophobe or papillary RCC: Hypovascular mass
- Histology: Highly variable

Renal Angiomyolipoma

- Tumor composed of abnormal blood vessels, smooth muscle, and fatty components
- NECT
  - Well-defined, cortical heterogeneous tumor with foci of fat density (-30 to -100 HU)
    - Renal mass with fat is usually an AML
  - Calcification rarely seen (if present, suspect RCC)
  - Hemorrhage may be seen in large tumors > 4 cm
  - When multiple AMLs are seen, suspect tuberous sclerosis
- CECT
  - Varied enhancement based on amount of fat and vascular components

- Notch sign: Geographic, triangular cortical defect
- CTA: Aneurysmal renal vessels may be seen
- MR
  - Tumor with increased fat content
    - T1WI: Hyperintense
    - Fat-suppression sequences: Show signal "drop out"

Renal Metastases and Lymphoma

- Renal metastases
  - Multiple hypovascular renal masses
  - When seen as solitary dense mass, may mimic oncocytoma
  - Usually known primary tumor and metastases in other sites strongly suggest renal metastases
- Renal lymphoma
  - Primary is very rare; secondary from generalized spread is more common
  - Bilateral involvement in 40-60% of cases
  - Hypovascular, moderate enhancement with contrast

PATHOLOGY

General Features

- Etiology
  - Exact etiology unknown
- Genetics
  - Deletion of chromosome 1 and sex chromosomes
  - Balanced translocation involving 11q13
- Associated abnormalities
  - Oncocytoma and RCC subtypes may coexist
    - Can be separate, hybrid, or collision tumors
  - Birt-Hogg-Dubé syndrome
    - Rare autosomal dominant disorder with multisystem epithelial neoplasms (skin, thyroid, colon, etc.)
    - Multiple RCCs, renal oncocytomas, and lung cysts
  - Oncocytosis: Oncocytic tumors replace entire kidney
    - Sporadic or due to chronic renal failure/dialysis

Gross Pathologic & Surgical Features

- Spherical/ovoid tumor with pseudocapsule
- Surgical specimen
  - Dark brown color (compared to yellow-orange RCC)
  - Fleshy central scar: More common when tumor > 5 cm
  - Rare necrosis, hemorrhage, calcification (~ 30%)

Microscopic Features

- Sheets of oncocytes packed with mitochondria
  - Large cells with granular eosinophilic cytoplasm
  - Nuclei appear smooth, round, with minimal atypia
- 3 cellularity patterns
  - Organoid: Tightly or loosely packed nests of cells
  - Tubulocystic: Few cells in edematous stroma
  - Mixed: Variable cellularity and stroma ratios
- Differences in cellularity and stroma influence enhancement

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Mostly asymptomatic; rarely flank pain, hematuria
- Laboratory data
  - Immunohistochemical tests
    - Oncocytoma: Cathepsin H positivity
    - Chromophobe RCC: Vimentin positivity
- Pathologic diagnosis
  - Needle biopsy increasingly performed to rule out RCC
    - Sampling error may underepresent tumor makeup
    - Can be difficult to distinguish oncocytoma from chromophobe and oncocytic variants of RCC
  - Histopathology of entire tumor resection is best

Demographics

- Age
  - Peak incidence occurs in 6th and 7th decades
- Gender
  - M:F = 2-3:1
- Epidemiology
  - Unknown: Usually incidental on imaging or autopsy

Natural History & Prognosis

- Benign tumors with excellent prognosis after resection

Treatment

- Surgical resection: Partial or total nephrectomy

DIAGNOSTIC CHECKLIST

Consider

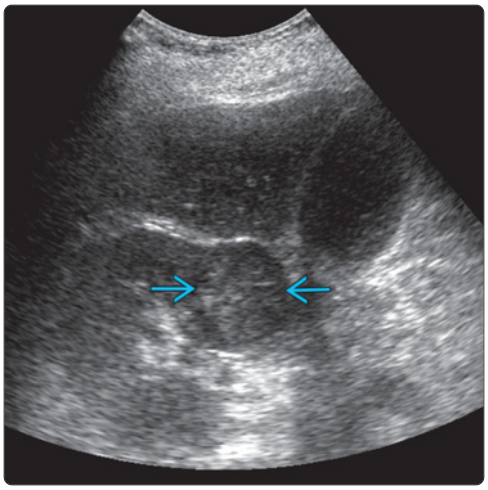
- Not possible to distinguish from RCC on imaging alone
  - May suggest diagnosis → nephron-sparing surgery
- Definitive pathologic diagnosis requires entire tumor to assure no sampling error
  - Primary pathologic differential is oncocytic variant of RCC

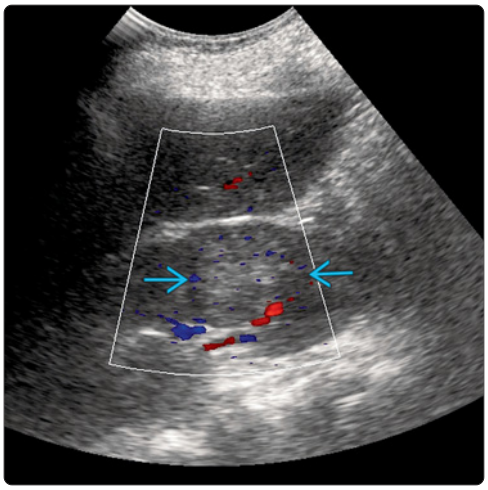
Image Interpretation Pearls

- Well-defined, solid, cortical mass lesion ± central scar

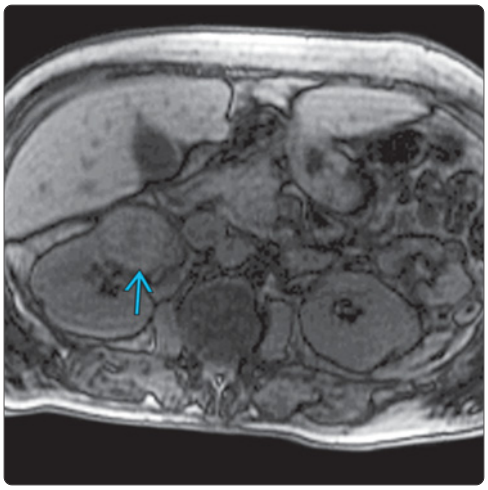
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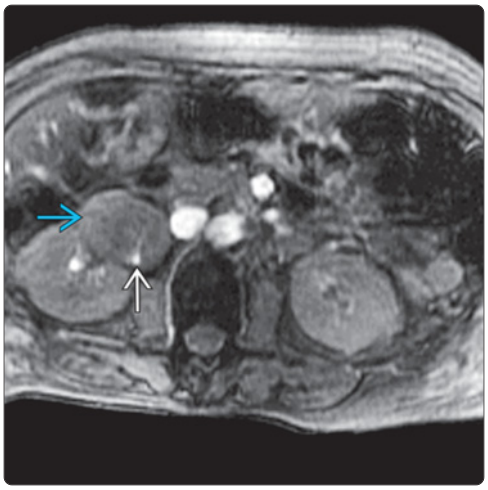


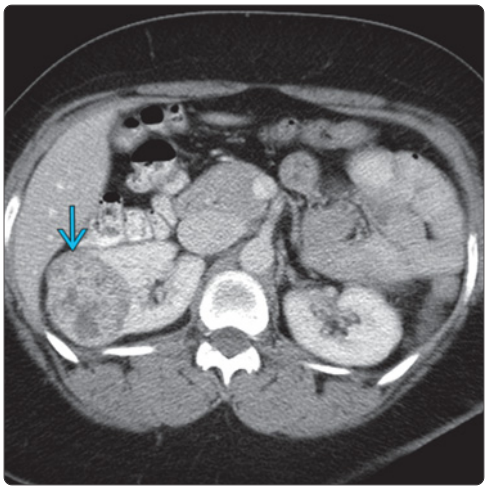
(Left) *Transverse US of the right upper quadrant reveals an incidental solid right renal mass with heterogeneous echotexture.* **(Right)** *Longitudinal color Doppler US in the same patient shows minimal, peripheral, low-level color flow within the right renal mass. These findings are consistent with a renal neoplasm, but further characterization is not possible by US alone. Flow may be better assessed with contrast-enhanced US, CT, or MR. Surgical resection was consistent with oncocytoma.*





(Left) *Axial T1WI out-phase MR shows a solid mass in the anterior cortex of the right kidney with mildly increased signal intensity compared to the renal parenchyma. Compared to the in-phase image, there was no loss of signal.* **(Right)** *Axial T2WI MR in the same patient shows heterogeneous, intermediate signal intensity of the right renal mass.*





(Left) *Axial FIESTA MR in the same patient shows the presence of vessels within the mass that is isointense to the renal parenchyma on this sequence. Although these findings suggest that the mass is due to renal neoplasm, the MR findings of oncocytoma do not allow definitive characterization or differentiation from RCC. Surgical resection was consistent with oncocytoma.* **(Right)** *Axial CECT during the nephrographic phase shows heterogeneous enhancement of a partially exophytic renal mass due to oncocytoma.*

Metanephric Adenoma

KEY FACTS

TERMINOLOGY

- Rare, benign epithelial renal neoplasm

IMAGING

- Ultrasound
  - Solid expansile mass without specific features
  - May range from hypoechoic to echogenic mass
- NECT
  - Soft tissue attenuation mass
    - Frequently hyperattenuating
  - Calcification in ~ 20%
- CECT
  - Well-circumscribed mass
  - Mild, delayed enhancement
  - Large tumors: Heterogeneity due to areas of necrosis and hemorrhage
- MR
  - T1WI: Low signal intensity
  - T2WI: Isointense to slightly hyperintense signal intensity

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma (RCC)
- Oncocytoma
- Wilms tumor (nephroblastoma)

PATHOLOGY

- Epithelial cells with tubulopapillary architecture
- Foci of hemorrhage and necrosis are common
- Immunohistochemical staining facilitates differentiation from papillary RCC

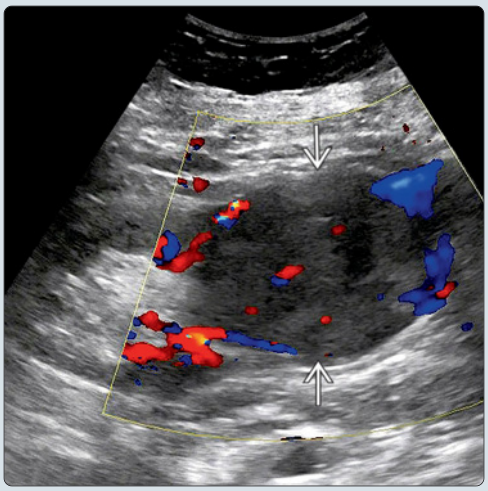
CLINICAL ISSUES

- Most are asymptomatic and discovered incidentally
  - Associated with polycythemia (~ 10-12%)

DIAGNOSTIC CHECKLIST

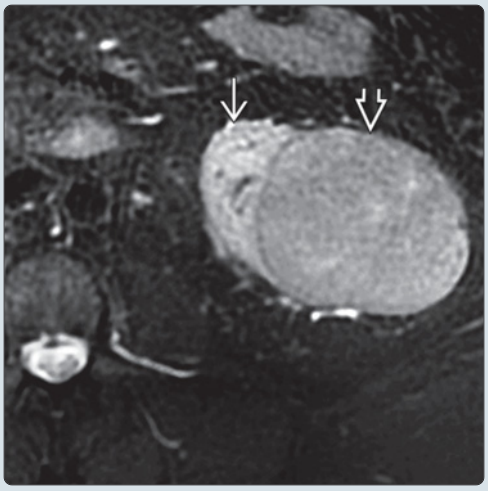
- Consider in middle-aged patient with solitary renal mass that is sightly hyperattenuating on NECT
- Excellent prognosis

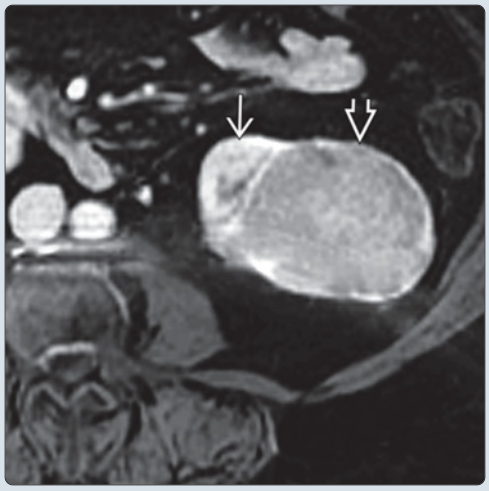
(Left) *Longitudinal US shows a solid, exophytic mass ⇨ extending from the inferior pole of the left kidney. The mass is isoechoic to renal cortex and has internal color flow.* **(Right)** *Axial NECT in the same patient shows that the mass ⇨ is slightly hyperattenuating to the renal parenchyma. Although pathology proved metanephric adenoma, these imaging features are nonspecific and differential considerations can include various other renal neoplasms, such as renal cell carcinoma and oncocytoma.*





(Left) *Axial T2WI MR with fat saturation in the same patient demonstrates that the exophytic left renal mass ⇨ has mildly heterogeneous signal intensity that is slightly less than the adjacent renal parenchyma ⇨.* **(Right)** *Axial post-contrast T1WI MR with fat saturation in the same patient reveals that the mass ⇨ is hypovascular compared to the adjacent normal renal parenchyma ⇨. There was no vascular invasion, metastases, or regional adenopathy. Pathology was consistent with metanephric adenoma.*





TERMINOLOGY

Synonyms

- Metanephric adenoma, nephrogenic nephroma, renal epithelial tumor, embryonal adenoma

Definitions

- Rare, benign epithelial renal tumor

IMAGING

General Features

- Location
  - Arise in renal cortex
  - Usually unilateral and unifocal
    - Few cases of multifocal adenoma reported
- Size
  - Variable, reported to range from 0.3-15 cm
    - Average: ~ 5.5 cm
- Morphology
  - Round to oval; well circumscribed

CT Findings

- NECT
  - Soft tissue attenuation mass
  - Reported to be frequently hyperdense
  - Calcification in ~ 20%
- CECT
  - Well-circumscribed mass
  - Typically only mild, delayed enhancement
    - Large tumors: Heterogeneous enhancement due to vascular stroma intermixed with areas of necrosis and hemorrhage
  - No vascular invasion

MR Findings

- T1WI
  - Low signal intensity
- T2WI
  - Isointense to slightly hyperintense signal intensity

Ultrasonographic Findings

- Grayscale ultrasound
  - Solid expansile mass without specific features
    - May range from hypoechoic to echogenic mass
    - Echogenicity due to psammomatous calcifications and interfaces from numerous small tubules
  - Cystic forms are extremely rare
- Color Doppler
  - Internal flow is often difficult to appreciate

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR
- Protocol advice
  - Perform CT or MR renal mass protocol

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma (RCC)

- Papillary variant typically shows mild, delayed enhancement similar to renal adenoma

Oncocytoma

- Nonspecific, heterogeneously enhancing mass

Wilms Tumor (Nephroblastoma)

- Solid, enhancing mass in pediatric population

PATHOLOGY

General Features

- Epithelial cells with tubulopapillary architecture
- Foci of hemorrhage and necrosis are common

Staging, Grading, & Classification

- Immunohistochemical staining facilitates differentiation from papillary RCC
  - Adenomas: Positive for CD57, AE1, and AE3
  - Adenomas: Negative for cytokeratin 7, WT1

Microscopic Features

- Uniform, small blue embryonal epithelial cells
- Abundant psammoma bodies; variable cytoplasm
- Cells arranged in acinar, tubular, or sheet-like configuration

CLINICAL ISSUES

Presentation

- Most are asymptomatic and discovered incidentally
  - Occasionally present with pain or hematuria
  - Associated with polycythemia (~ 10-12%)

Demographics

- Age
  - Wide variability, affecting children through adults
    - Most common in 5th decade
- Gender
  - F:M = 2:1

Natural History & Prognosis

- Typically considered to be benign with excellent prognosis
  - Few cases with metastatic disease have been reported

Treatment

- Nephrectomy (total or nephron sparing)

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Multilocular Cystic Nephroma

KEY FACTS

IMAGING

- Encapsulated, multilocular, cystic mass herniating into renal hilum
  - Typically unilateral

TOP DIFFERENTIAL DIAGNOSES

- Cystic renal cell carcinoma
  - Enhancing nodules favor RCC over MLCN
  - Cysts in RCC usually not enclosed by capsule
  - Usually occur in adult men more than women
- Cystic Wilms tumor
  - Septa: Numerous/thick in Wilms; thin in MLCN
- Localized cystic renal disease
  - Multiple cysts replace part or all of 1 kidney
  - No surrounding capsule
- Multicystic dysplastic kidney
  - Usually involves entire kidney
  - Segmental MCDK: Mostly associated with ureteral duplication
- Cortical (simple) cysts
  - No surrounding capsule
- Mixed embryonal and stromal tumor of kidney
  - Very rare, recently defined entity
  - Complex solid and cystic renal tumor
  - Occurs almost exclusively in postmenopausal women

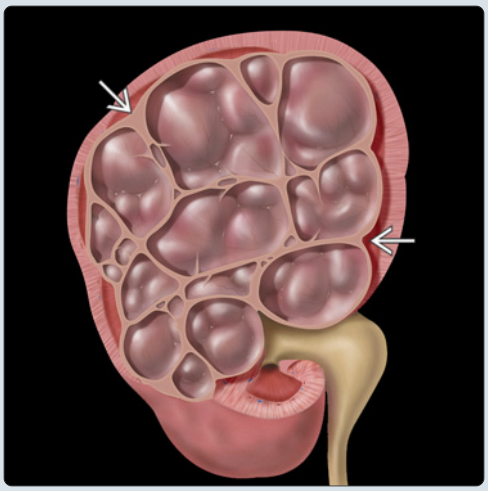
CLINICAL ISSUES

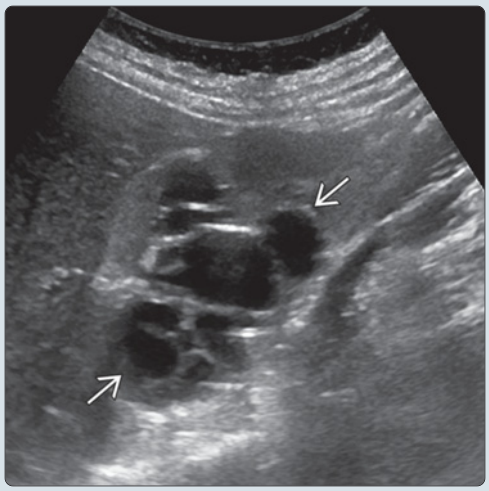
- Biphasic age distribution
  - Males: 3 months to 2 years
  - Females: < 5 years and 40-60 years

DIAGNOSTIC CHECKLIST

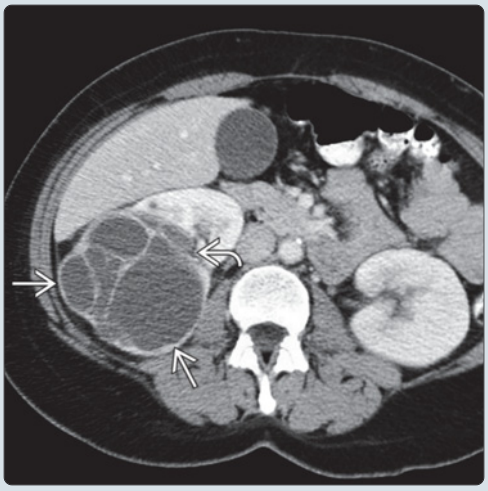
- MLCN qualifies as Bosniak class 3 or 4 complex cystic mass
  - Surgical excision is treatment of choice
- Difficult to distinguish from multilocular cystic RCC
- Imaging features suggestive of MLCN
  - Cystic spaces in MLCN are enclosed within capsule
  - Herniation of cystic spaces into renal hilum

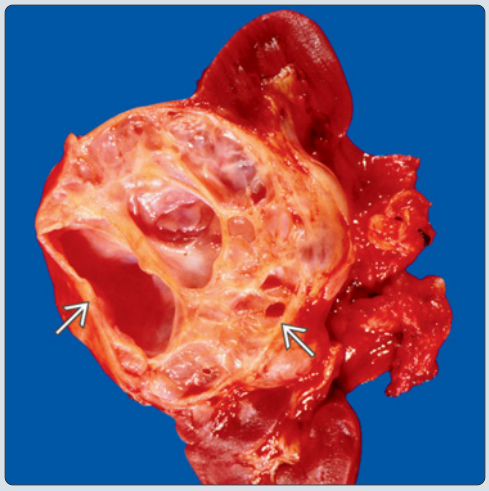
(Left) *Graphic shows an encapsulated ⇨, multilocular, cystic mass that herniates into the renal hilum.* **(Right)** *Sagittal US in a 48-year-old woman shows a multiloculated, cystic mass ⇨ invaginating into the renal hilum.*





(Left) *Axial CECT in a middle-aged woman with flank discomfort shows an encapsulated, multiloculated mass ⇨ with septa that enhances after contrast administration. Note the invagination of the mass into the renal hilum ⇨, a characteristic feature of this tumor.* **(Right)** *Coronal surgical cross section photograph of an excised kidney in the same patient shows a multiloculated cystic mass extending into the renal hilum. Note the presence of a capsule ⇨ completely surrounding the cystic mass.*





TERMINOLOGY

Abbreviations

- Multilocular cystic nephroma (MLCN)

Synonyms

- Multilocular cystic renal tumor, benign cystic nephroma, cystadenoma, Perlman tumor
- Polycystic nephroblastoma, cystic partially differentiated nephroblastoma (CPDN)

Definitions

- Nonhereditary, benign renal neoplasm arising from metanephric blastema

IMAGING

General Features

- Best diagnostic clue
  - Encapsulated, multilocular, cystic mass herniating into renal hilum
- Location
  - Typically unilateral and solitary
  - Occasionally completely replaces kidney
- Size
  - Mass: Few cm to > 30 cm (average: 10 cm)
  - Locules: Few mm to 2.5 cm
- Morphology
  - Well-circumscribed, cystic mass with thick fibrous capsule ± herniation into renal pelvis
- Other general features
  - Usually solitary and unilateral
  - Usually symptomatic in adults and asymptomatic palpable mass in children
  - Variable growth: Slowly over years to rapid over months
  - Histological classification of multilocular cystic renal tumors
    - Multilocular cystic nephroma
    - Cystic partially differentiated nephroblastoma
  - MLCN and CPDN indistinguishable by imaging alone

Radiographic Findings

- Radiography
  - ± soft tissue mass (± displacement of adjacent structures)
  - ± curvilinear or amorphous calcification
- IVP
  - Depending on size and location of mass
    - ± pelvocaliectasis
    - ± tumor herniates into renal pelvis (mimics primary tumor of collecting system)
  - Tomogram of nephrographic phase shows septations

CT Findings

- NECT
  - Large, well-defined, encapsulated, multiloculated, cystic mass
  - CT attenuation: ≥ water (proteinaceous fluid)
  - Small locules (< 1 cm) and myxomatous material within cysts and fibrous septa may appear as solid mass
  - Rare, curvilinear septal calcifications
- CECT
  - Capsule: Usually enhances
  - Septa: Moderate enhancement (thin and linear)
  - Cystic component: No enhancement
    - No communication with collecting system during excretory phase
  - ± distortion of collecting system or herniation of mass into renal hilum ± obstructive changes

MR Findings

- T1WI
  - Typically low SI within cystic components
  - Occasionally higher SI (protein, hemorrhage)
  - Capsule and septa: Low SI (fibrous content)
- T2WI
  - High SI within cystic components
  - Occasionally variable SI (protein, hemorrhage)
  - Capsule and septa: Low SI (fibrous content)
- DWI
  - No restricted diffusion
- T1WI C+
  - Mild enhancement of capsule and thin septa
    - No mural nodule or solid component
  - Herniation of mass into renal pelvis well depicted

Ultrasonographic Findings

- Grayscale ultrasound
  - Large, well-defined, multiloculated, cystic mass
  - Numerous anechoic cysts and thin, echogenic septa
  - Echogenic, fibrous capsule
  - ± echogenicity within cysts may mimic solid component
    - Clustered small cysts cause acoustic interfaces

Angiographic Findings

- Conventional
  - Hypovascular mass (rarely avascular or hypervascular)

Nuclear Medicine Findings

- Tc-99m DMSA
  - Photon defect corresponding to renal mass
  - Occasionally faint uptake by septa noted

Imaging Recommendations

- Multiplanar CT or MR + IV contrast

DIFFERENTIAL DIAGNOSIS

Cystic Renal Cell Carcinoma

- Mimics multilocular cystic nephroma on imaging
- Cysts in RCC usually not enclosed by capsule
- Cystic RCC less often projects into renal sinus
- Enhancing nodules favor RCC over MLCN
- Usually seen in adult men more than women
  - MLCN seen in boys and adult women
- Gross pathology: Simulates MLCN
- Diagnosis: Histology

Cystic Wilms Tumor

- Grows entirely by expansion of large cystic spaces within stroma
- Septa: Numerous/thick in Wilms; thin in MLCN

Localized Cystic Renal Disease

- Multiple cysts replace part or all of 1 kidney

- No surrounding capsule

Multicystic Dysplastic Kidney (MCDK)

- Usually involves entire kidney
- Rarely focal or segmental
- Segmental MCDK: Mostly associated with ureteral duplication
- Segmental MCDK + absence of ureteral duplication may simulate MLCN
  - Both are histologically distinctive
  - Segmental MCDK lacks capsule

Cortical (Simple) Cysts

- Solitary or multiple; unilateral or bilateral
- Most cysts are unilocular and arise in cortex; less commonly from medulla
- May have multiple simple cysts
  - But no surrounding capsule

Mixed Embryonal and Stromal Tumor of Kidney

- Very rare, recently defined entity
- Occurs almost exclusively in postmenopausal women
- Complex solid-cystic mass mimicking array of renal lesions
- Expansile mass with heterogeneous and delayed contrast enhancement

PATHOLOGY

General Features

- Etiology
  - Unknown
  - Pathogenesis: Benign equivalents of nephroblastoma, hamartoma
- Associated abnormalities
  - Sporadic association with other anomalies
  - Coexist with angiomyolipoma; adrenal adenoma
- Embryology/anatomy
  - Tumor arises from metanephric blastema

Gross Pathologic & Surgical Features

- "Honeycomb" cystic areas of varied sizes
- Encapsulated by thick fibrous capsule
  - No communication with renal collecting system
- Noncommunicating locules separated by thin septa
  - CPDN locules separated by thick or irregular septa
- Mostly intraparenchymal tumors

Microscopic Features

- Locules: Lined by flattened or cuboidal epithelium
- Septa contains fibrous tissue + tubular elements
- No capsular, lymphatic, or vascular invasion
- No mitotic figures
- MLCN: Septa have no undifferentiated elements
- CPDN: Septa contain blastemal ± embryonal elements

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Adults
    - Abdominal or flank pain; ± palpable mass
    - Hematuria, urinary tract infection, HTN
  - Children
    - Nonpainful; palpable abdominal or flank mass
    - ± hematuria and urinary tract infection
- Laboratory data
  - Urine analysis may show RBC ± WBC
- Diagnosis: Biopsy and histology

Demographics

- Age
  - Bimodal distribution
  - Males: 3 months to 4 years
  - Females: < 5 years and 40-60 years
- Gender
  - Bimodal distribution
  - Predominantly young boys (2/3 of all cases) and middle-age women (1/3 of all cases)
- Epidemiology
  - Rare tumor

Natural History & Prognosis

- Complications
  - Obstructive uropathy, infection, local recurrence
  - Hemorrhagic when extended into renal pelvis
  - Extremely rare potential for aggressive behavior
- Prognosis
  - Good following nephrectomy

Treatment

- Surgical excision: Nephrectomy
  - Complete or partial with tumor-free margins

DIAGNOSTIC CHECKLIST

Consider

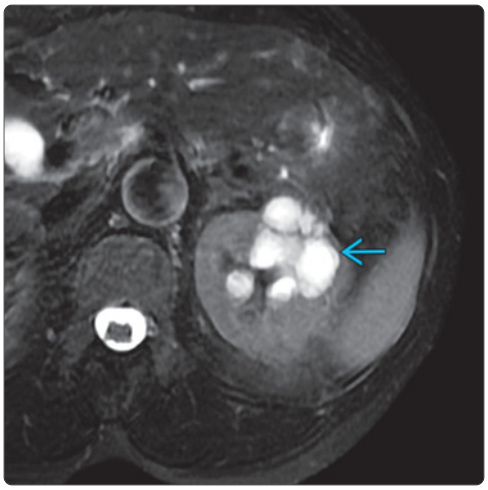
- MLCN appears as Bosniak class 3 or 4 complex cystic mass (surgical excision is treatment of choice)
- Difficult to distinguish from multilocular cystic RCC

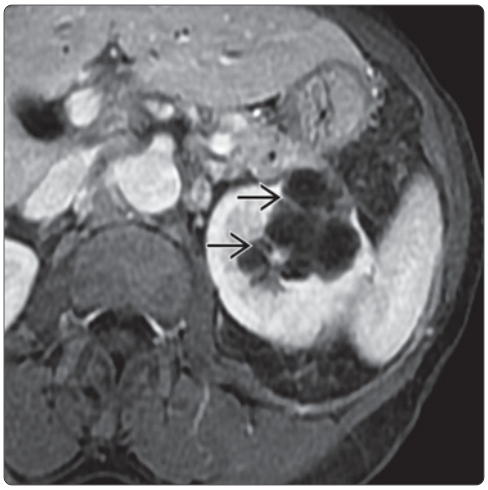
Image Interpretation Pearls

- Cystic spaces in MLCN are enclosed within capsule, unlike almost all other cystic renal masses

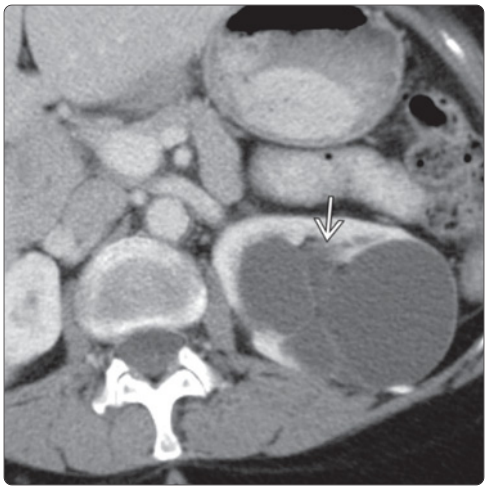
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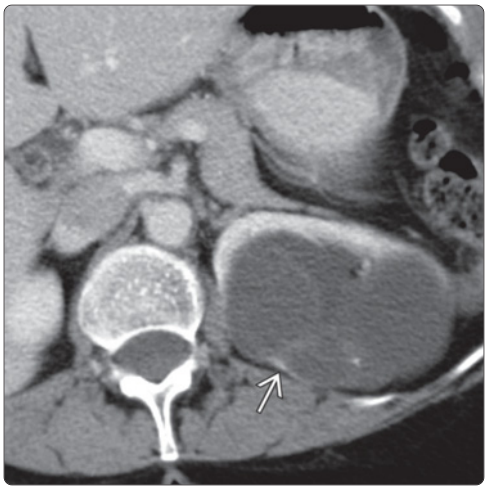
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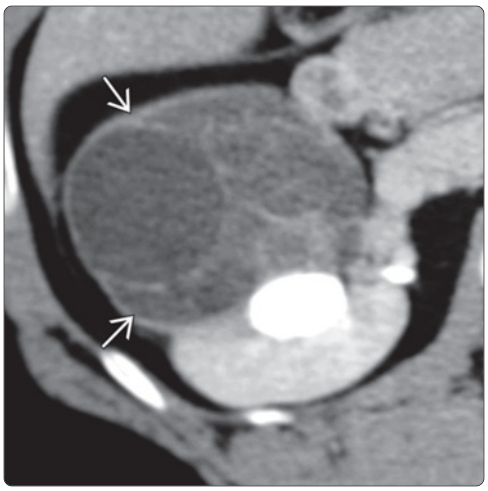


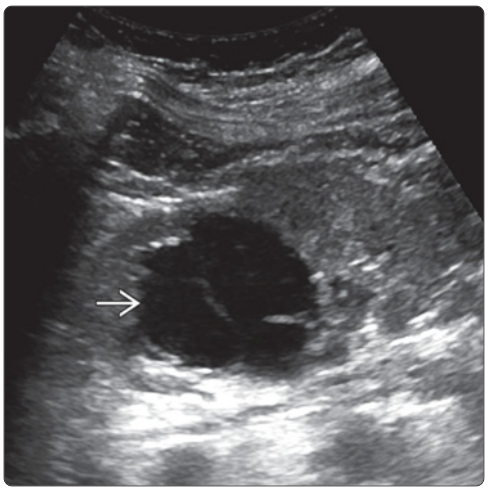
(Left) *Axial T2 MR with fat saturation shows high signal intensity in a multiloculated cystic lesion ⇨ that arises from the superior pole of the left kidney and extends toward the renal sinus.* **(Right)** *Axial post-contrast T1 MR with fat saturation in the same patient shows enhancement within the thin septa ⇨ of the multiloculated cystic mass.*





(Left) *Axial CECT shows a classic multilocular cystic nephroma as a multiseptated cystic mass that herniates into the renal sinus ➡. The kidney functions normally, and there is no evidence of vascular invasion, lymphadenopathy, or other signs of malignancy.* **(Right)** *Axial CECT in the same patient shows a classic, multilocular cystic nephroma as an encapsulated ➡ multicystic mass with herniation into the renal sinus.*





(Left) *Axial CECT shows an encapsulated ➡, multiloculated cystic mass with enhancing septa herniating into the renal hilum. The presence of the capsule encircling the cysts is a key feature that distinguishes multilocular cystic nephroma from other multicystic renal masses.* **(Right)** *Sagittal US demonstrates an encapsulated, multiloculated cystic mass ➡ in the superior pole of the kidney that herniates toward the renal sinus.*

Mixed Epithelial and Stromal Tumor

KEY FACTS

TERMINOLOGY

- Rare, benign cystic renal neoplasm that contains stromal and epithelial components

IMAGING

- Well-marginated, multiseptate cystic and solid mass with delayed contrast enhancement
- Variable appearances due to differential components of cystic and solid components
- Very rare calcification or fatty component

TOP DIFFERENTIAL DIAGNOSES

- Cystic renal cell carcinoma
- Complex renal cyst
- Renal abscess
- Adult cystic nephroma
- Multicystic dysplastic kidney

PATHOLOGY

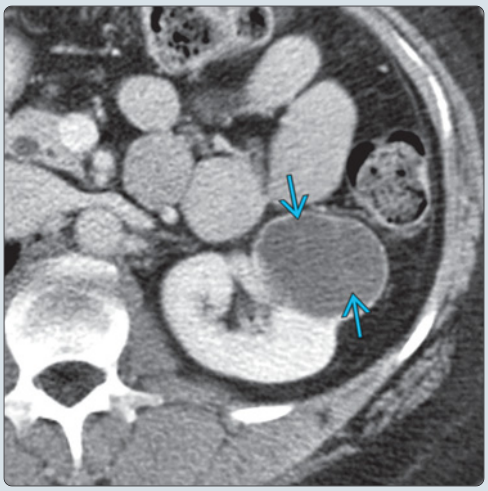
- Stromal elements: Spindle cells that mimic ovarian stroma
- Epithelial elements: Epithelial-lined cysts and microcysts

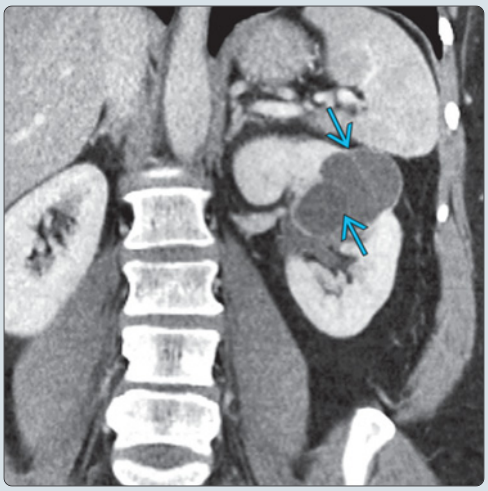
CLINICAL ISSUES

- Almost exclusively found in perimenopausal women
- Common symptoms: Hematuria, flank pain, palpable mass, urinary tract infection
  - ~ 25% are asymptomatic
- ~ 70% of MESTs are Bosniak 4 lesions and resected

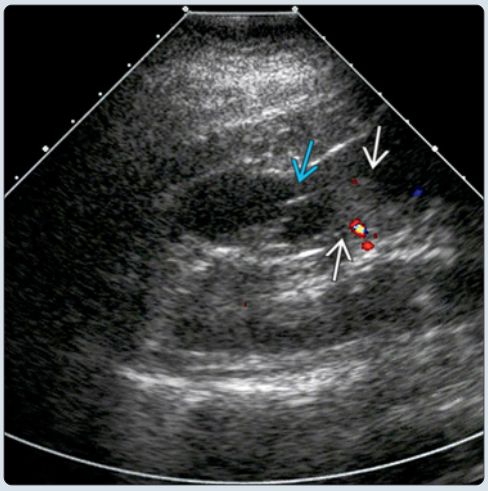
DIAGNOSTIC CHECKLIST

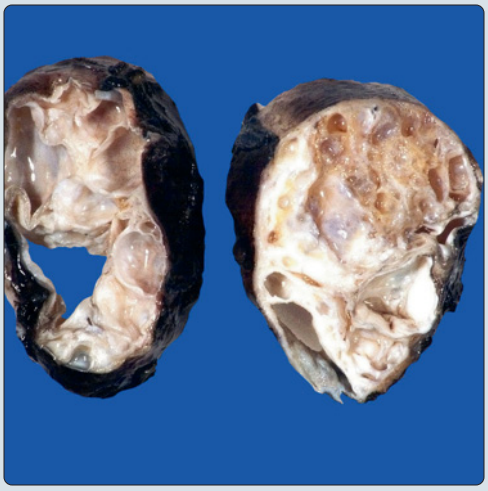
- Consider diagnosis of MEST in
  - Perimenopausal women
  - Patients with chronic estrogen use
  - Cystic tumors with delayed enhancement
  - Renal pelvis tumors with negative urine cytology
  - Obstructed duplicated collecting system





(Left) *Axial CECT shows an ovoid, well-circumscribed mass that is partially exophytic from the left kidney. Note several thin, mildly enhancing septa. Pathology revealed mixed epithelial and stromal tumor (MEST).* **(Right)** *Coronal CECT in the same patient shows the multiple, enhancing thin septa in the MEST to better advantage. Multiplanar reformatted images are often useful for detecting and assessing the thickness of intratumoral septa and for preoperative planning.*





(Left) *Longitudinal color Doppler US of the right kidney in a different patient shows an ovoid, predominantly cystic mass with internal septum and a small solid component that shows color flow. The patient underwent partial nephrectomy. Cystic renal cell carcinoma was suspected at imaging. The pathology revealed MEST.* **(Right)** *Longitudinal bisected gross surgical specimen of MEST demonstrates numerous cysts and microcysts of variable size interspersed between septa and solid components.*

TERMINOLOGY

Abbreviations

- Mixed epithelial and stromal tumor (MEST)

Synonyms

- Formerly referred to as multilocular cyst with ovarian stroma, cystic hamartoma of renal pelvis, adult-type mesoblastic nephroma, leiomyomatous renal hamartoma

Definitions

- Rare, benign cystic renal neoplasm that contains stromal and epithelial components

IMAGING

General Features

- Best diagnostic clue
  - Well-marginated, expansile, multiseptate cystic and solid mass with delayed contrast enhancement
- Size
  - Round or ovoid tumor of variable size

CT Findings

- NECT
  - Cystic component: Near water to intermediate attenuation depending on presence of proteinaceous/hemorrhagic fluid
  - Solid component: Soft tissue attenuation
    - Very rare calcification or fatty components
- CECT
  - Heterogeneous, delayed enhancement of solid components and septa
    - Septa are typically ≥ 5 mm in thickness

MR Findings

- T1WI
  - Cystic component: Low signal intensity (SI)
  - Solid component: Intermediate to high SI
- T2WI
  - Cystic component: High SI
  - Solid component: Low SI
- T1WI C+
  - Same as noted for CECT

Ultrasonographic Findings

- Hyperechoic mass to cystic mass with ± solid component

Imaging Recommendations

- Best imaging tool
  - CT or MR: Multiplanar with post-contrast imaging

DIFFERENTIAL DIAGNOSIS

Cystic Renal Cell Carcinoma (RCC)

- ~ 15% of RCCs have cystic components
- Thick, irregular septa; ± solid, enhancing nodules

Complex Renal Cyst

- Thin, nonenhancing septa; no solid component

Adult Cystic Nephroma

- Multicystic lesion; no solid component; septa < 5 mm

Multicystic Dysplastic Kidney

- Variably sized cysts; minimal enhancement; no excretion

Renal Abscess

- Infiltration of perinephric fat and infectious symptoms

Obstructed Duplicated Collecting System

- Excretory phase: Gradual filling of obstructed system

PATHOLOGY

General Features

- Etiology
  - Unknown; ± association with chronic estrogen therapy

Gross Pathologic & Surgical Features

- Complex solid and cystic renal neoplasm

Microscopic Features

- Epithelial components: Microcysts or epithelial-lined cysts
- Stromal components contain spindle cells
  - Spindle cells mimic ovarian stroma
    - Express estrogen and progesterone receptors

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Hematuria, pain, urinary tract infection, palpable mass
  - ~ 25% are asymptomatic and found incidentally at imaging

Demographics

- Almost exclusively found in perimenopausal women
  - Few case reports in literature describe MEST in men

Natural History & Prognosis

- Almost always follow benign course after resection
  - Few reports of MEST coexisting with sarcoma
    - Questionable true malignant transformation

Treatment

- Partial or radical nephrectomy

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Renal Cell Carcinoma

KEY FACTS

IMAGING

- 2% of sporadic RCC are bilateral, and 16-25% of sporadic RCC are multicentric in same kidney
- Exophytic: Projects from cortical surface, distinct from parenchyma
- Most are hypervascular (not papillary RCC)
- Rarely, small areas of fat attenuation (-80 to -120 HU)
  - Combination of fat and calcification suggests RCC, not renal angiomyolipoma
- Renal venous (23%) and inferior vena cava (IVC) tumor extension (7%)
- Metastases (most to least common): Lung, liver, bone, adrenal, opposite kidney, and brain
- Cystic RCC: Enhancing, smooth or nodular septa
- Multiphase CT
  - Mandatory: Nonenhanced and parenchymal phase
  - Optional: Corticomedullary, excretory phases
- Multiphase MR
  - T1WI: Typically low signal intensity (SI)
  - T2WI: High to heterogeneous SI due to necrosis
  - Post contrast: Heterogeneous due to areas of viable, hypervascular tumor & necrosis

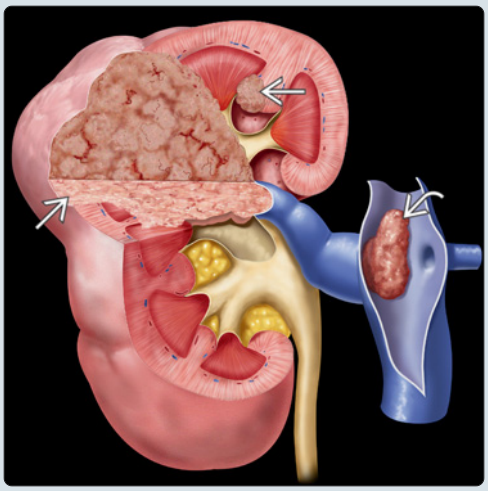
TOP DIFFERENTIAL DIAGNOSES

- Renal oncocytoma
- Renal angiomyolipoma
- Renal transitional cell carcinoma
- Renal metastases and lymphoma
- Renal abscess
- Hemorrhagic renal cyst

PATHOLOGY

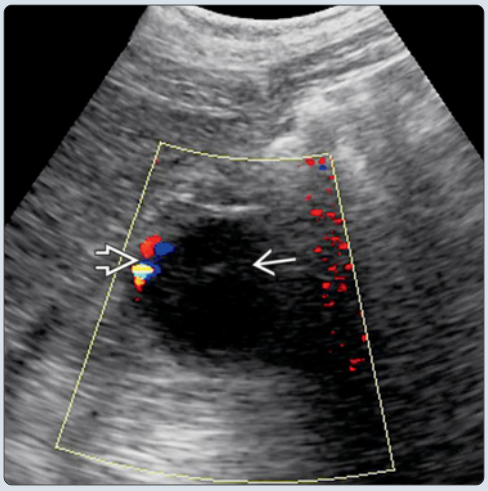
- Clear cell (70%), papillary (10-15%), granular cell (7%), chromophobe cell (5%), sarcomatoid (1.5%), collecting duct (< 1%)

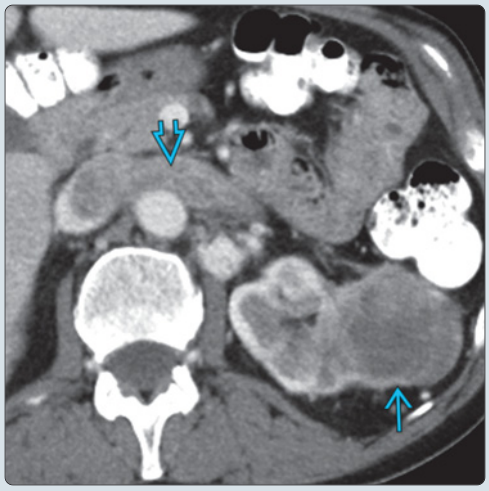
(Left) *Graphic shows a heterogeneous, vascular, expansile mass arising from the renal cortex, invading the renal vein and IVC. The tumor is multicentric, as is the case in 16-25% of sporadic RCCs and in a higher percentage of syndromal or inherited cases.* **(Right)** *Coronal T1WI MR with contrast enhancement shows an expansile mass in the upper pole of the left kidney, extending into the renal vein and IVC. The propensity for venous invasion leads to a high prevalence of pulmonary metastases.*





(Left) *Sagittal color Doppler US shows a cystic lesion in the superior pole of the right kidney. The lesion has a thick septum and mural nodule with internal color flow. Partial nephrectomy was performed, confirming cystic renal cell carcinoma.* **(Right)** *Axial CECT demonstrates a heterogeneously enhancing, solid mass with central necrosis that is exophytic from the lateral cortex of the left kidney. There is enhancing tumor thrombus in the left renal vein and IVC. The tumor was confirmed to be clear cell renal cell carcinoma.*





TERMINOLOGY

Abbreviations

- Renal cell carcinoma (RCC)

Definitions

- Malignant tumor arising from renal tubular epithelium

IMAGING

General Features

- Location
  - 2% of sporadic RCCs are bilateral; 16-25% of sporadic RCC are multicentric in same kidney
  - Renal cortex is most common location
- Morphology
  - Usually solid mass, but occasionally cystic
- Other general features
  - ~ 50% found incidentally on imaging

CT Findings

- NECT
  - Heterogeneous mass ± cystic components
    - High attenuation (hemorrhage)
    - Low attenuation (≤ 20 Hounsfield units) due to necrosis or cystic component
  - Rare calcifications (< 10% of cases)
  - Rare, small areas of fat attenuation (-80 to -120 HU)
    - Usually due to enveloping perirenal or sinus fat
    - Dedifferentiated RCC → fat and calcification or ossification
    - Combination of fat & calcification suggests RCC more than angiomyolipoma
  - Cystic RCC
    - Uni- or multilocular cystic mass with thick wall
    - Calcification of septa or tumor capsule
- CECT
  - Enhancement (attenuation value ↑ by ≥ 20 HU compared to NECT)
    - Less than renal parenchyma on nephrographic & pyelographic phases
    - Small mass may be missed on corticomedullary phase
  - Heterogeneous enhancement (hemorrhage and necrosis)
  - Exophytic: Projects from cortical surface, distinct from parenchyma
  - ± lucent zone surrounding mass (pseudocapsule)
  - ± infiltration of calyces or renal pelvis may simulate transitional cell carcinoma
  - Subcapsular hemorrhage, perinephric hemorrhage, or both
  - Renal venous (23%) and inferior vena cava (IVC) tumor extension (7%)
  - Direct extension to adjacent muscles (e.g., diaphragm, psoas, quadratus lumborum, erector spinae) & viscera (i.e., colon, liver, pancreas, spleen)
  - Metastases to local lymph nodes (≥ 1 cm)
  - Metastases (most to least common): Lung, liver, bone, adrenal, opposite kidney, and brain (often hypervascular)
  - Cystic RCC: Enhancing, smooth or nodular septa

MR Findings

- T1WI
  - Typically isointense (~ 60%) to hypointense
  - ↑ signal intensity (SI) if internal hemorrhage (methemoglobin)
- T2WI
  - Clear cell RCC: Typically ↑ SI
  - Papillary RCC: Typically ↓ SI
    - Appears same as angiomyolipoma with minimal fat
    - Papillary RCCs often have pseudocapsule
- T1WI C+ FS
  - Enhancement is usually less than normal renal tissue
- Hypointense band/rim on T1WI (25%) and T2WI (60%)
- In/out of phase
  - Clear cell RCC can lose signal intensity on out of phase images
    - Tumor cells may contain intracellular lipid
  - Loss of signal on out of phase sequence cannot be reliably used to differentiate RCC from AML

Ultrasonographic Findings

- Grayscale ultrasound
  - Detect 85% of masses > 3 cm; ≤ 60% < 2 cm
  - Hyperechoic (48%), isoechoic (42%), or hypoechoic (10%) renal mass
  - Poor acoustic transmission & lobulated contour
  - Cystic RCC
    - Multiple thick septa
    - Mural thickening, nodules, and calcification
    - Diffuse, hypoechoic mass (hemorrhage and necrosis) with fluid-debris levels
- Color Doppler
  - Most prominent color flow around periphery of mass
  - Variable flow depending on degree of necrosis & cystic components

Imaging Recommendations

- Best imaging tool
  - Multiphasic CT: Diagnosis and staging (corticomedullary, nephrographic, & excretory phases)
  - MR: Equal to or better than CT for staging

DIFFERENTIAL DIAGNOSIS

Renal Oncocytoma

- Enhancing mass that is indistinguishable from RCC
- ± stellate, hypoattenuating scar

Renal Angiomyolipoma (AML)

- Intratumoral, fat-containing mass without calcification
- May be indistinguishable from RCC if no visible fat on CT
  - ~ 5% of AMLs have minimal to no fat
  - Most lipid poor AMLs are hyperattenuating & homogeneously enhancing

Transitional Cell Carcinoma, Kidney

- Renal infiltration → kidney enlargement and poorly defined margins without change in shape
- Renal pelvic filling defect, irregular narrowing of collecting system
- Encasing pelvicalyceal system → hypovascular tumor

Renal Metastases and Lymphoma

- Metastases (e.g., lung/breast/colon cancer, melanoma)
  - Usually hypovascular with infiltrative or exophytic growth
- Lymphoma
  - Usually multiple or bilateral with infiltrative growth
  - Hypoechoic, hypovascular, solitary, intrarenal mass ± adenopathy may simulate RCC

Renal Infection (e.g., Focal Pyelonephritis)

- Abscess: Necrotic mass with renal enlargement
- Differentiate by clinical history & urinalysis

Hemorrhagic Renal Cyst

- Lack of central/internal enhancement

PATHOLOGY

General Features

- Etiology
  - Genetics, smoking, chemicals, idiopathic
- Genetics
  - ~ 4% of RCCs due to genetic diseases
  - + family history: ↑ chance of developing RCC by 2.8x
- Associated abnormalities
  - 6 hereditary RCC syndromes
  - von Hippel-Lindau
    - Clear cell RCC; tumors in brain and spinal cord; adrenal, renal, and pancreatic cysts and tumors
  - Hereditary papillary RCC
    - Autosomal dominant, multiple and bilateral papillary tumors
  - Hereditary leiomyomatosis and RCC
    - RCCs, uterine and cutaneous leiomyomas, adrenal hyperplasia
  - Birt-Hogg-Dubé syndrome
    - Autosomal dominant, benign cutaneous tumors, oncocytomas and chromophobe RCCs, medullary thyroid cancer
  - Succinate dehydrogenase (SDH) associated familial cancer
    - RCCs, oncocytomas, head/neck paragangliomas, pheochromocytomas
  - Tuberous sclerosis complex
    - Facial angiofibromas, renal AMLs, clear cell RCC, lymphangiomyomatosis of lungs

Staging, Grading, & Classification

- TNM staging (7th edition)
  - T (tumor size)
    - T1: Limited to kidney but ≤ 7 cm
      - T1a: ≤ 4 cm
      - T1b: > 4 cm but ≤ 7 cm
    - T2: Limited to kidney but > 7 cm
      - T2a: > 7 cm but ≤ 10 cm
      - T2b: > 10 cm
    - T3: Extension into major veins or perinephric tissues but not into ipsilateral adrenal or beyond Gerota fascia
      - T3a: Spread to renal vein
      - T3b: Spread to infradiaphragmatic IVC
      - T3c: Spread to supradiaphragmatic IVC or invades wall of IVC
    - T4: Into ipsilateral adrenal gland or beyond Gerota fascia
  - N (lymph nodes)
    - N0: No nodal involvement
    - N1: Regional nodal involvement
  - M (metastases)
    - M0: No metastases
    - M1: Distant metastases
  - Stage groupings
    - Stage 1: T1, N0, M0
    - Stage 2: T2, N0, M0
    - Stage 3: T1 or T2, N1, M0; or T3, N0 or N1, M0
    - Stage 4: T4, any N, M0; or any T, any N, M1
- Bosniak classification for management of cystic renal lesions
  - Bosniak 1 cystic lesion
    - Simple cyst: Water attenuation; round; well-marginated; hairline thin wall; no septa, calcification, solid components, or enhancement
    - Malignant: 0%
  - Bosniak 2 cystic lesion
    - Minimally complex: Few hair-line thin septa ± perceived enhancement (not measurable); fine calcifications or short segment of slightly thickened calcification in wall or septa; homogeneously high-attentuation masses (≤ 3 cm) that are sharply marginated and do not enhance
    - Malignant: 0%
  - Bosniak 2F cystic lesion
    - Minimally complex but require follow-up: Multiple hair-line thin septa ± perceived enhancement (not measurable); thick or nodular calcifications; minimal smooth thickening of wall or septa that may show perceived (not measurable) enhancement; intrarenal nonenhancing high-attenuation masses (> 3 cm); no enhancing soft tissue components
    - Work-up: Follow-up imaging (US, CT, MR); frequency is variable, but typically performed at 6 & 12 months, then annually for 5 years
    - Malignant: ~ 5%
  - Bosniak 3 cystic lesion
    - Indeterminate cystic renal lesions: Thickened irregular or smooth walls or septa, with measurable enhancement
    - Work-up: Partial nephrectomy; radiofrequency ablation in elderly or poor surgical candidates
    - Percentage malignant: ~ 54%
  - Bosniak 4 cystic lesion
    - Clearly malignant: Criteria of category 3 but also containing enhancing soft tissue components
    - Work-up: Partial or complete nephrectomy
    - Malignant: 100%
- Management recommendations of incidental solid renal mass
  - Large (> 3 cm): RCC is most likely diagnosis
    - Clinical management: Surgery
    - Caveat: AML with minimal fat, oncocytoma, and other benign lesions may be found at surgery

        - Typically not able to be differentiated by imaging alone
    - Small (1-3 cm): RCC is most likely diagnosis
        - Traditional clinical management: Surgery
            - Active surveillance increasingly employed in elderly, or patients with commodities: Very ow incidence of metastases with small (< 3 cm) RCC
        - Caveat: Benign etiology may be found at surgery
            - If hyperattenuating & homogeneously enhancing, consider MR & percutaneous biopsy to diagnose potential AML with minimal fat
    - Very small (< 1 cm): Difficult to differentiate RCC, AML, oncocytoma
        - Clinical management: observe until 1 cm
        - Thin collimation imaging (≤ 3 mm) helps to confirm enhancement

Gross Pathologic & Surgical Features

- Ranges from cystic to mixed cystic-solid to completely solid mass with irregular lobulated margins

Microscopic Features

- Clear cell (70%), papillary (10-15%), granular cell (7%), chromophobe cell (5%), sarcomatoid (1.5%), collecting duct (< 1%)
    - Papillary RCC: Type 1 & type 2 are not reliably differentiated by any 1 imaging criterion
    - Type 2 papillary RCC tend to present at more advanced stage compared to type 1
- Marked variability among tumor by histopathology
    - Accurate distinction relies on molecular and cytogenetic markers
- Role of image-guided percutaneous biopsy
    - Biopsy can aid in diagnosis of indeterminate renal mass (and is routinely performed prior to percutaneous ablation)
        - e.g., differentiating papillary RCC vs. AML with minimal fat
    - However, tumor heterogeneity & overlap with benign lesions (some RCCs have oncocytic features) may limit role of biopsy in some cases

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
    - Classic triad of symptoms found in < 10%
        - Gross hematuria (60%), flank pain (40%), palpable flank mass (30-40%)
    - Systemic manifestations by paraneoplastic (e.g., erythropoietin secretion, renin secretion), paraendocrine (e.g., hepatopathy), and serological (e.g., amyloidosis) syndromes
    - Distant metastases may cause symptoms of cough, hemoptysis, bone pain, seizures, etc.

Demographics

- Age
    - 50-70 years
- Gender
    - M:F = 2:1
- Epidemiology
    - Accounts for 2-3% of all cancers
    - Incidence: ~ 65,000/year in USA
    - Mortality: ~ 13,500/year in USA

Natural History & Prognosis

- Prognosis
    - 5-year survival rates
        - Stage I: 81%
        - Stage II: 74%
        - Stage III: 53%
        - Stage IV: 8%
    - Survival rates are better for incidentally discovered RCCs
    - Bilateral or multiple RCC have poorer survival rate; solitary RCC or metastasis has better survival rate
    - 20-30% of patients with localized tumor who undergo surgical resection relapse in 15-18 months

Treatment

- Nephrectomy: Radical vs. partial
    - Partial nephrectomy: Stages Ia-Ib
    - Radical nephrectomy: Stages II-III
- Ablation (cryo- or radiofrequency)
    - Requires ≤ 5 cm exophytic tumor & no extrarenal extension
- Targeted therapy
    - Tyrosine kinase inhibitors (e.g., sorafenib, sunitinib)/mTOR inhibitors (e.g., temsirolimus)
- Immunotherapy
    - Interleukin-2 (IL2)/Bevacizumab plus interferon alpha
- Chemotherapy, radiation

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Papillary type RCC is hypovascular; easily mistaken for cyst
    - Papillary RCC often has ↓ SI on T2WI and may be mistaken for lipid-poor AML
- Clear cell RCC may lose signal on out of phase sequences
    - Out of phase sequence cannot be used to reliably differentiate RCC from AML

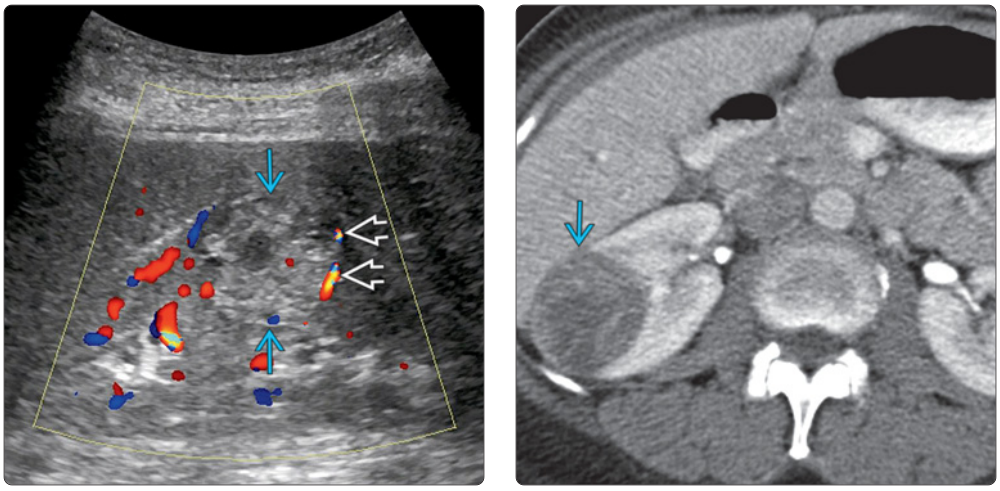
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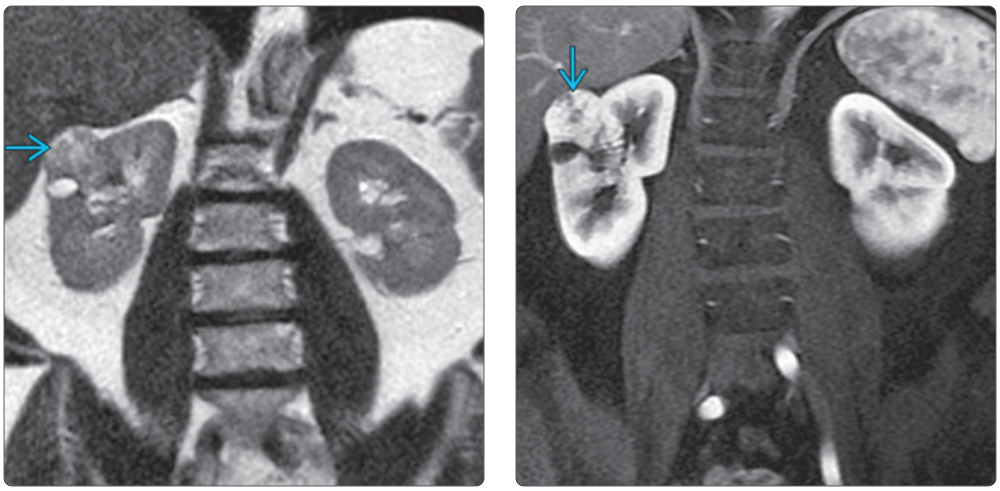
(Left) *Coronal CECT reformation shows a large mass replacing all but the superior pole of the left kidney. The mass is heterogeneous due to areas of viable, enhancing tumor and areas of necrosis.* **(Right)** *Coronal CECT of a different plane in the same patient shows invasion of the left renal vein and IVC. Even patients with invasion of the IVC are often candidates for surgical resection. It is important to determine the complete extent of the venous invasion and whether the wall of the vein is involved.*

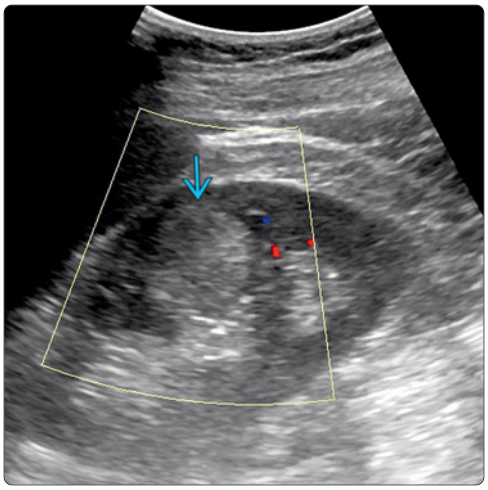


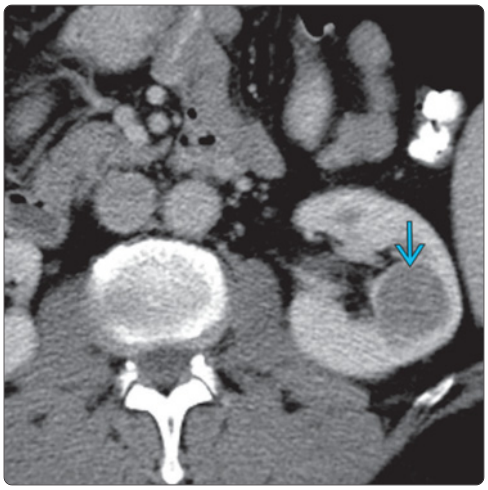
(Left) *Sagittal color Doppler US shows a solid, heterogeneous, and slightly hyperechoic mass within the posterior aspect of the interpolar cortex. The mass results in a mild contour deformity of the posterior renal cortex. Several areas of color flow are present within the mass due to tumor neovascularity.* **(Right)** *Axial CECT in the same patient shows a heterogeneously enhancing mass in the lateral cortex of the right kidney that corresponds to the abnormality found on prior US.*



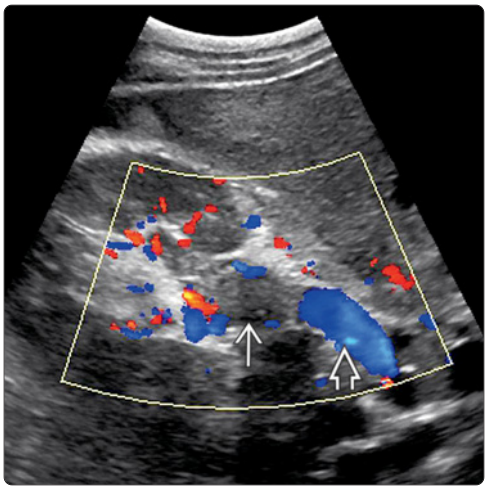
(Left) *Coronal T2WI MR in the same patient shows that the mass has heterogeneous signal intensity and is partially exophytic along the posteroinferior border of the right renal cortex.* **(Right)** *Coronal T1WI MR with fat saturation demonstrates heterogeneous enhancement of the right renal mass due to small areas of central necrosis. There was no vascular invasion, adenopathy, or metastatic disease. Due to these factors and the size and location of the tumor, the patient was cured by partial nephrectomy.*

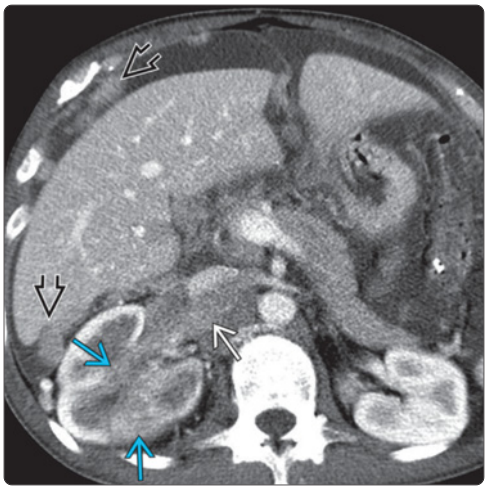




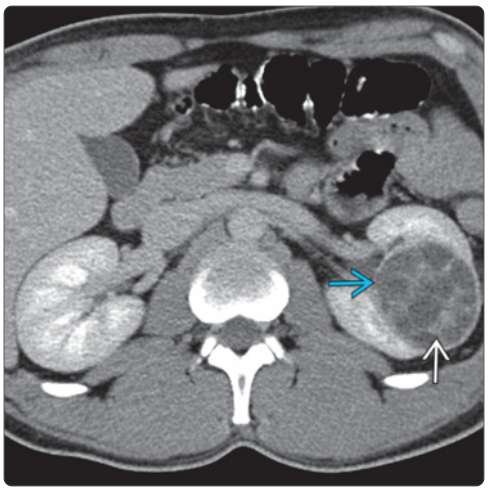


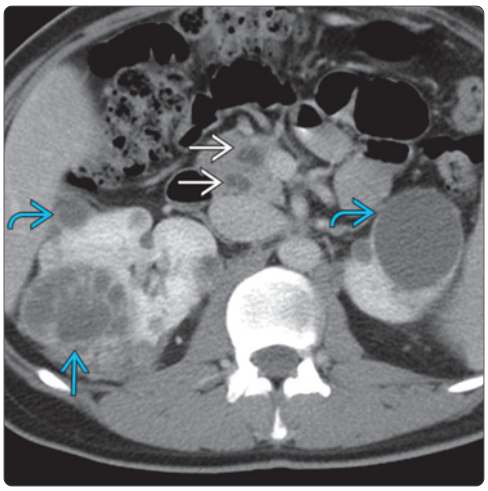
(Left) *Sagittal color Doppler US of the left kidney reveals an incidental round, echogenic mass in the interpolar cortex. No demonstrable color Doppler flow was shown within the mass.* **(Right)** *Axial CECT during the nephrographic phase in the same patient shows a solid, round mass that has minimal enhancement (30 HU above NECT). The nephrectomy specimen was consistent with a papillary RCC. Papillary RCCs are hypovascular compared to clear cell RCCs.*





(Left) *Axial color Doppler US reveals echogenic thrombus in a segment of the left renal vein near the renal hilum. Flow is preserved in the segment of renal vein closest to the IVC.* **(Right)** *Subsequent follow-up axial CECT in the same patient demonstrates tumor thrombus in the right renal vein, peritoneal carcinomatosis, and ill-defined tumor in the right kidney due to infiltrative papillary RCC. Type 2 papillary RCCs typically present at a more advanced stage compared to type 1 tumors.*





(Left) *Axial CECT reveals an incidental heterogeneous, cystic mass in the left kidney. This is a Bosniak 4 cystic lesion due to the numerous thick, enhancing septa and enhancing nodular components. The surgical resection specimen was consistent with a clear cell RCC.* **(Right)** *Axial CECT in a patient with von Hippel Lindau disease demonstrates renal cysts, pancreatic cysts, and a RCC in the right kidney. The patient also had hemangioblastomas in the brain and spinal cord.*

T | Primary Tumor

TNM	Definitions
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor ≤ 7 cm in greatest dimension, limited to kidney
T1a	Tumor ≤ 4 cm in greatest dimension, limited to kidney
T1b	Tumor > 4 cm but ≤ 7 cm in greatest dimension, limited to kidney
T2	Tumor > 7 cm in greatest dimension, limited to kidney
T2a	Tumor > 7 cm but ≤ 10 cm in greatest dimension, limited to kidney
T2b	Tumor > 10 cm, limited to kidney
T3	Tumor extends into major veins or perinephric tissues but not into ipsilateral adrenal gland and not beyond Gerota fascia
T3a	Tumor grossly extends into renal vein or its segmental (muscle containing) branches, or tumor invades perirenal &/or renal sinus fat, but not beyond Gerota fascia
T3b	Tumor grossly extends into vena cava below diaphragm
T3c	Tumor grossly extends into vena cava above diaphragm or invades wall of vena cava
T4	Tumor invades beyond Gerota fascia (including contiguous extension into ipsilateral adrenal gland)

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in regional lymph node(s)

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis

Adapted from 7th edition AJCC Staging Forms (2010).

AJCC | Stages/Prognostic Groups

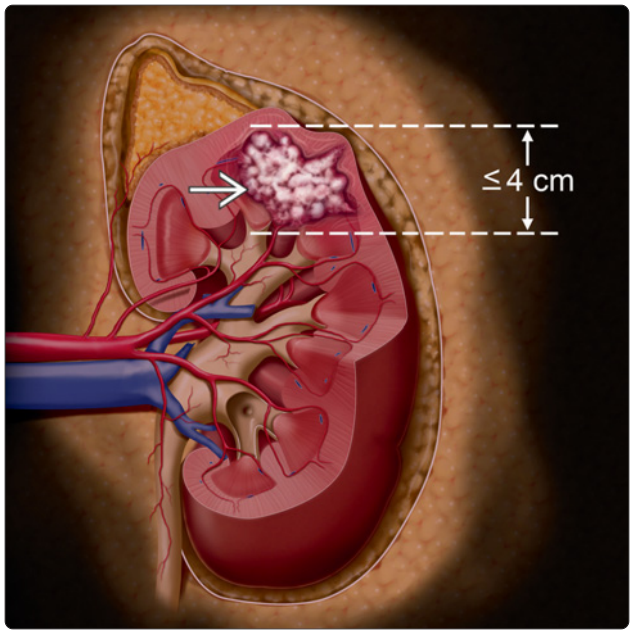
Stage	T	N	M
I	T1	N0	M0
II	T2	N0	M0
III	T1 or T2	N1	M0
	T3	N0 or N1	M0
IV	T4	Any N	M0
	Any T	Any N	M1

Adapted from 7th edition AJCC Staging Forms (2010).

Comparison of Robson and TNM Staging

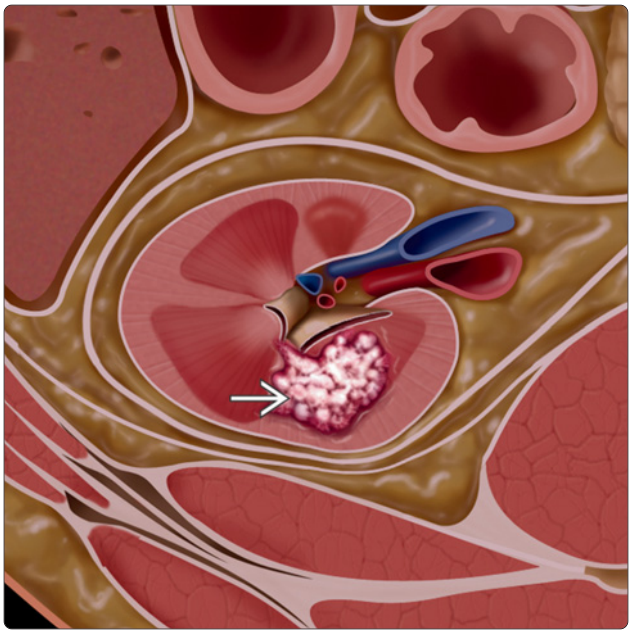
Robson Stage	Definitions	TNM
I	Tumor ≤ 2.5 cm and confined to kidney	T1
	Tumor > 2.5 cm and confined to kidney	T1-T2
II	Tumor extends into perinephric fat or adrenal	T3a if involves perinephric fat, T4 if involves ipsilateral adrenal
IIIA	Tumor invades renal vein	T3a
	Tumor extends into inferior vena cava	T3b below diaphragm
IIIB	Lymph node involvement	N0-N1, M0
IIIC	Involvement of local vasculature and lymph nodes	T3a-c, N0-N1
IVA	Involvement of adjacent organs (except ipsilateral adrenal)	T4
IVB	Distant metastases	M1

T1a



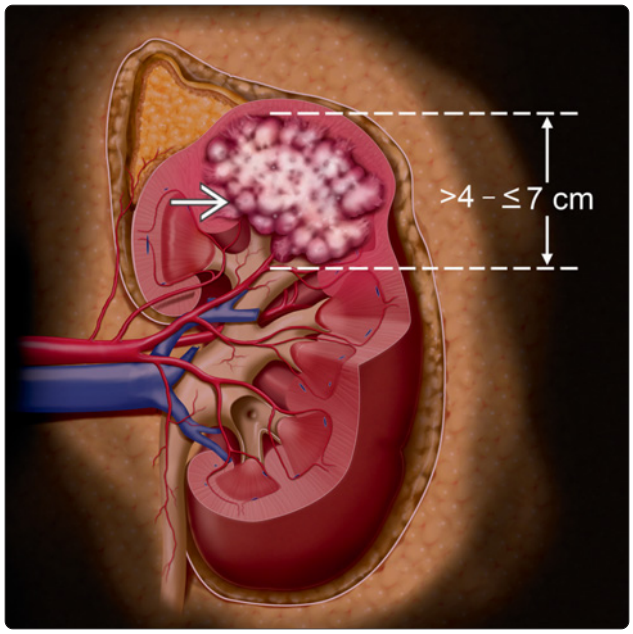
Coronal graphic shows a typical T1a lesion ➡, defined as ≤ 4 cm and confined to the kidney.

T1a



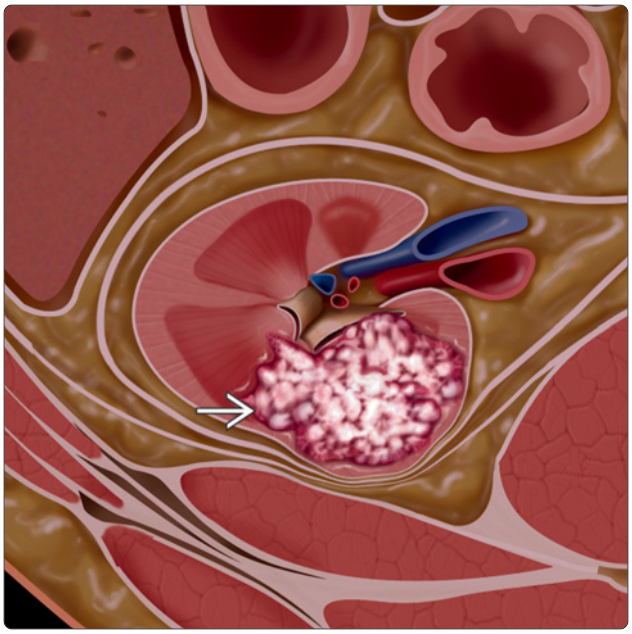
Axial graphic shows a T1a renal cell carcinoma ➡, defined as ≤ 4 cm and confined to the kidney. Lesions may be exophytic or more centrally located, as is the T1a depicted here.

T1b



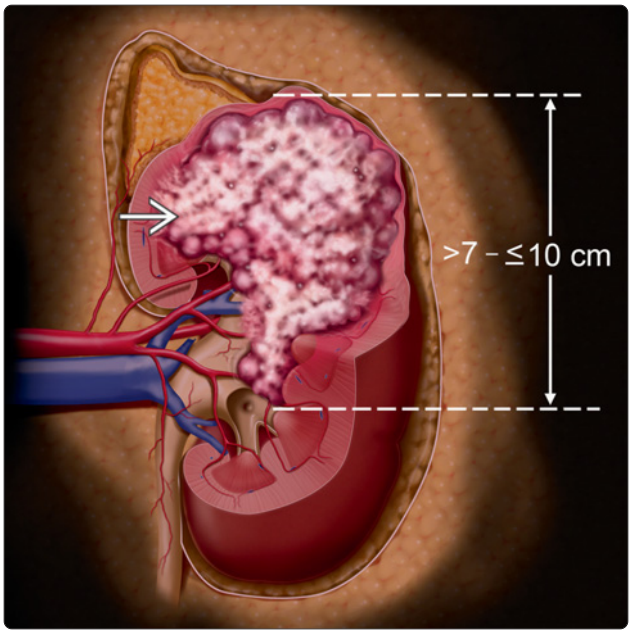
Coronal graphic shows a typical T1b lesion ➡, defined as > 4 cm but ≤ 7 cm and confined to the kidney.

T1b



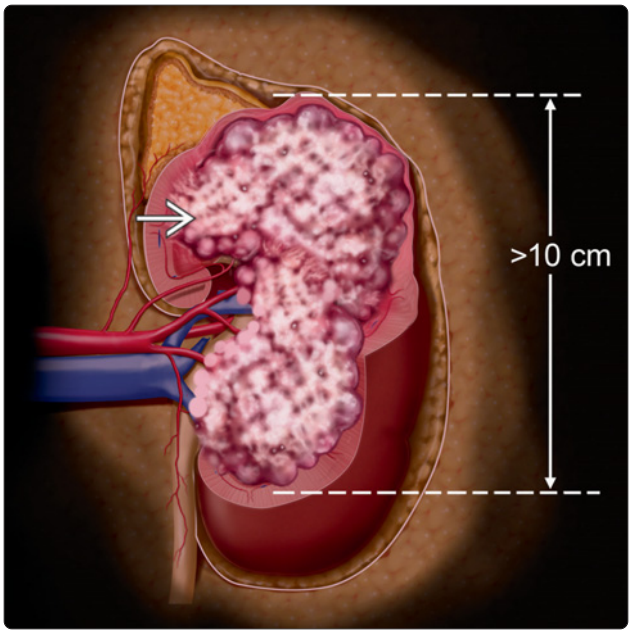
Axial graphic shows a T1b renal cell carcinoma ➡, defined as > 4 cm but ≤ 7 cm and confined to the kidney. These lesions may be exophytic but will still be categorized as T1 as long as there is no extension outside of the kidney.

T2a



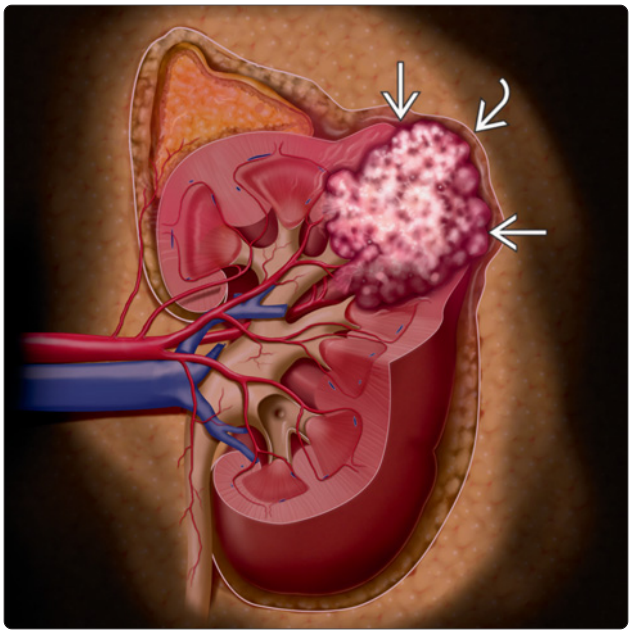
Coronal graphic shows a T2a renal cell carcinoma ➡, defined as > 7 cm but ≤ 10 cm and confined to the kidney. These lesions may also be described as exophytic but do not invade outside of the kidney.

T2b



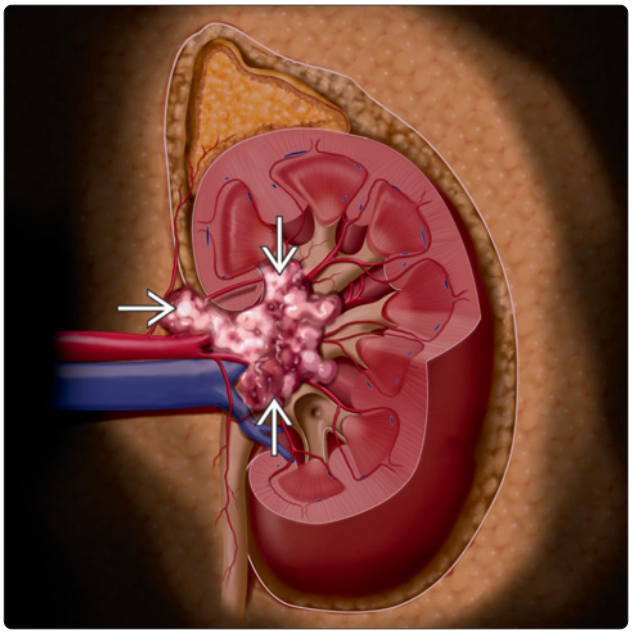
Coronal graphic shows a typical T2b renal cell carcinoma ➡, defined as > 10 cm but confined to the kidney.

T3a



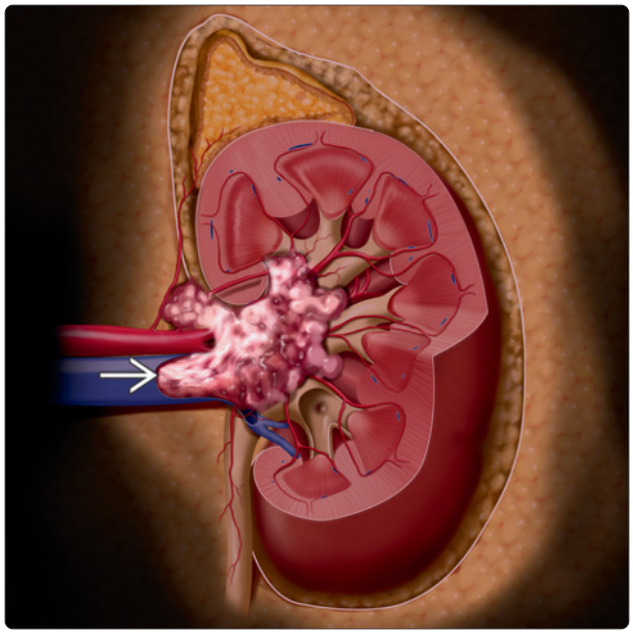
Coronal graphic shows a T3a renal cell carcinoma with tumor extension ➡ into the perirenal fat but not beyond the confines of the Gerota fascia ➡.

T3a



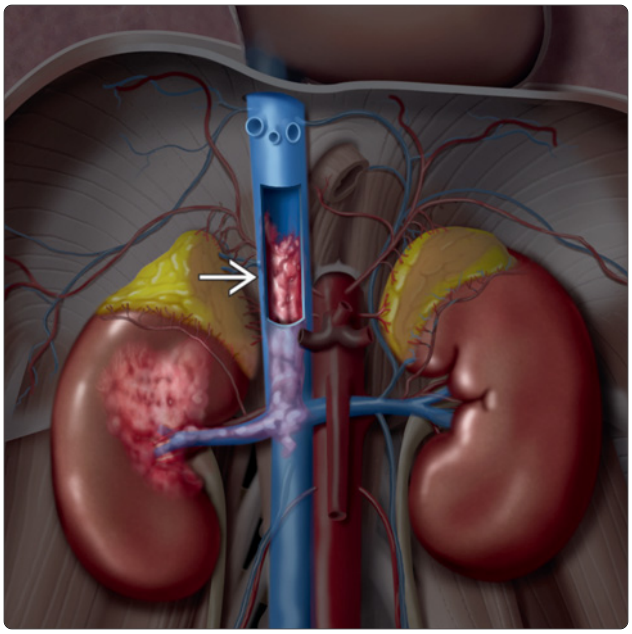
Coronal graphic shows another example of a T3a lesion with extension into the perirenal or renal sinus fat ➡. Again, the tumor is limited by the confines of the Gerota fascia.

T3a



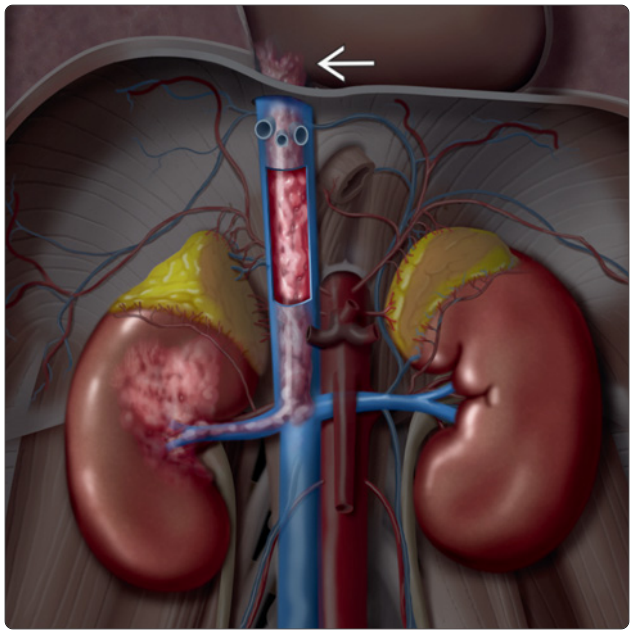
Coronal graphic shows a T3a renal cell carcinoma with extension of tumor into the renal vein ➔. Extension beyond the renal vein into the inferior vena cava would be defined as a T3b lesion.

T3b



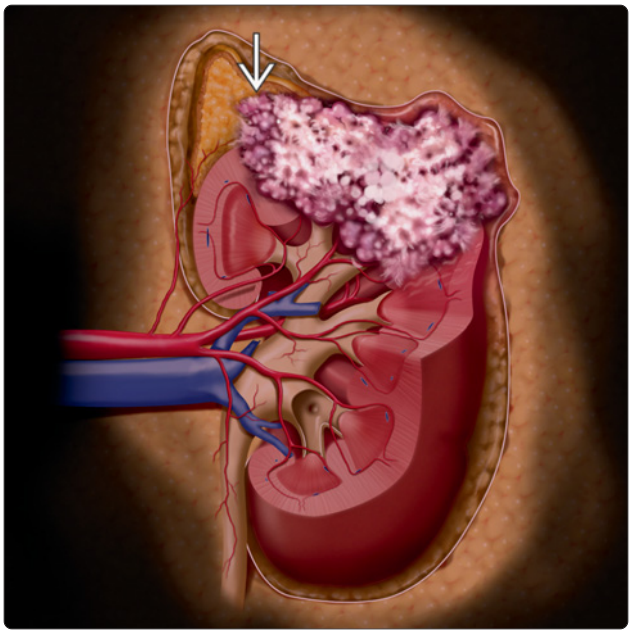
Coronal graphic shows a T3b renal cell carcinoma with extension of tumor not only into the renal vein but also into the inferior vena cava ➔. T3b lesions do not extend above the diaphragm.

T3c



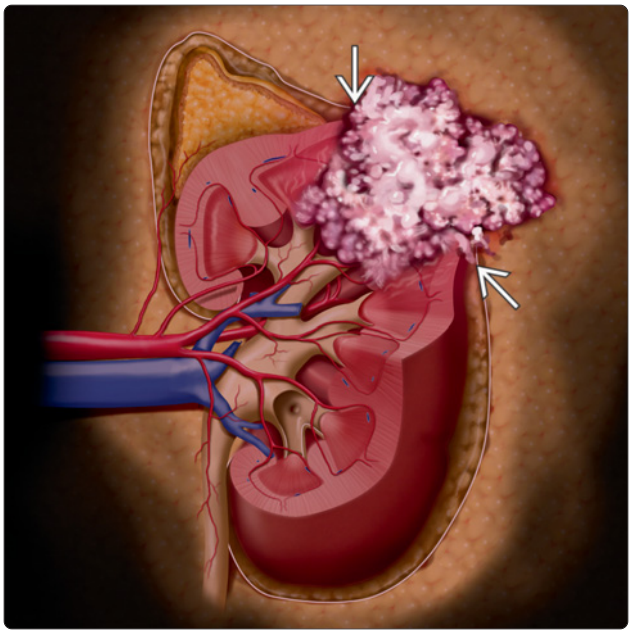
Coronal graphic shows a T3c renal cell carcinoma with involvement of tumor in the renal vein and inferior vena cava, as well as extension of tumor within the inferior vena cava above the diaphragm ➔.

T4



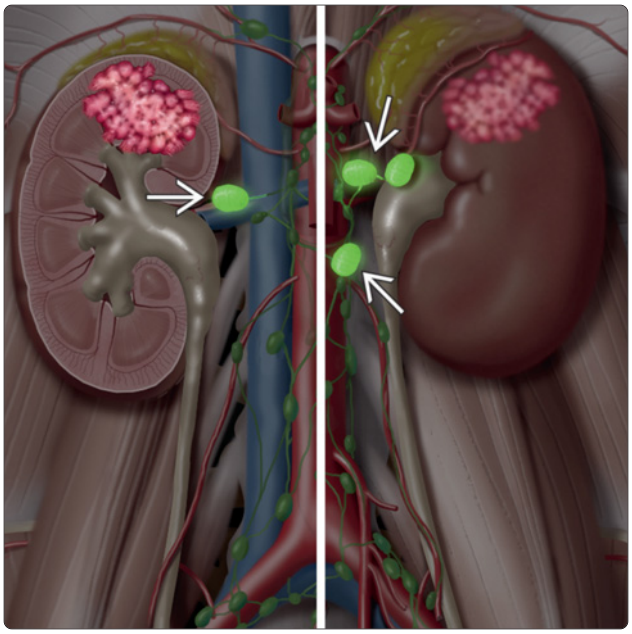
Coronal graphic shows a T4 renal cell carcinoma with extension into the ipsilateral adrenal gland ➔.

T4



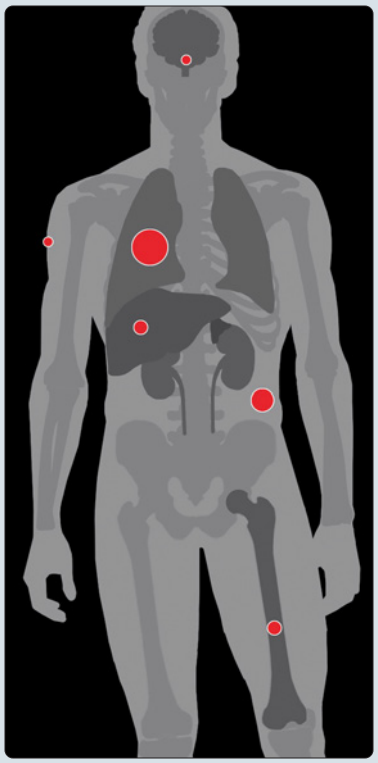
Coronal graphic shows another example of a T4 lesion with invasion into not only the perirenal fat but also with extension beyond the Gerota fascia ➔.

N1 Disease (Stage III or IV)



Coronal graphic shows retroperitoneal adenopathy on both sides of the midline ➔. N0 disease is classified as no regional lymph node metastasis, while N1 disease is classified as metastatic disease in regional lymph nodes.

Metastases, Organ Frequency



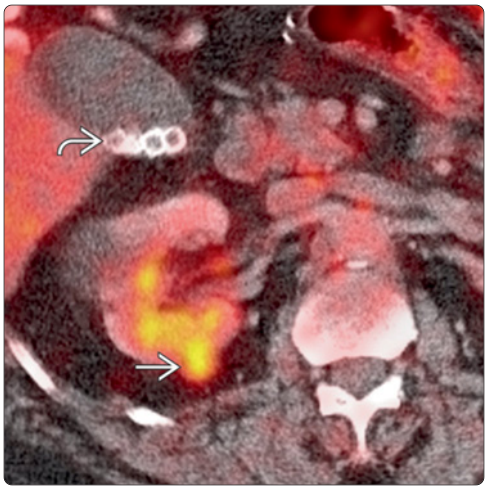
Lung	*75%*
Soft tissue	*35%*
Bone	*20%*
Liver	*20%*
Adrenal gland	*19%*
Cutaneous tissues	*8%*
Central nervous system	*8%*

25-40% of patients present with metastatic disease.

Stage I (T1a N0 M0)

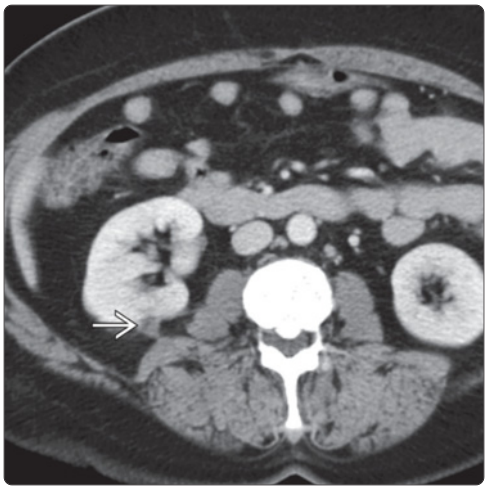


Stage I (T1a N0 M0)

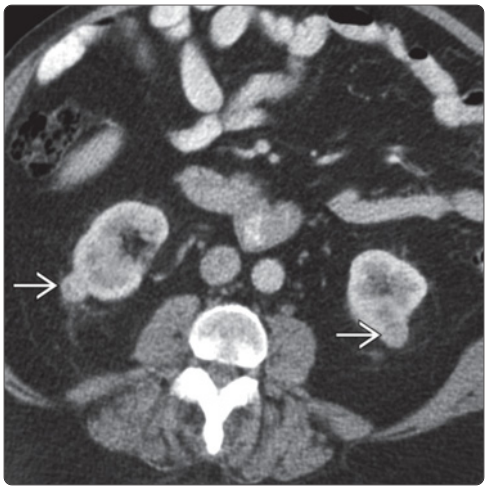


(Left) *Axial CECT shows a small (just over 1 cm) low-attenuation enhancing lesion in the posterior mid-pole of the right kidney. Although nonspecific, the enhancement makes this suspicious for a renal cell carcinoma.* **(Right)** *Axial PET/CT shows focal FDG within the pathologically confirmed T1a renal cell carcinoma in the right kidney. Renal cell carcinomas have variable FDG uptake, but intensely FDG-avid lesions should be viewed as very suspicious. Note the gallstones.*

Stage I (T1a N0 M0)



Stage I (T1a N0 M0)

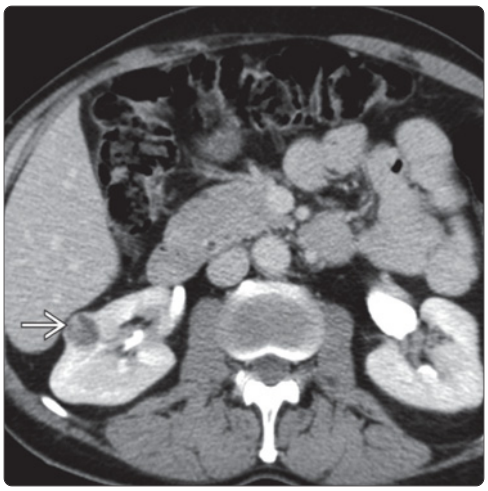


(Left) *Axial CECT shows a 1 cm, incidentally discovered T1a renal cell carcinoma in the posterior mid-pole of the right kidney, subsequently treated with radiofrequency ablation.* **(Right)** *Axial CECT shows bilateral enhancing renal lesions in this patient with von Hippel-Lindau, compatible with bilateral T1a renal cell carcinomas. T1a lesions are those ≤ 4 cm in greatest dimension and limited to the kidney.*

Stage I (T1a N0 M0)



Stage I (T1a N0 M0)

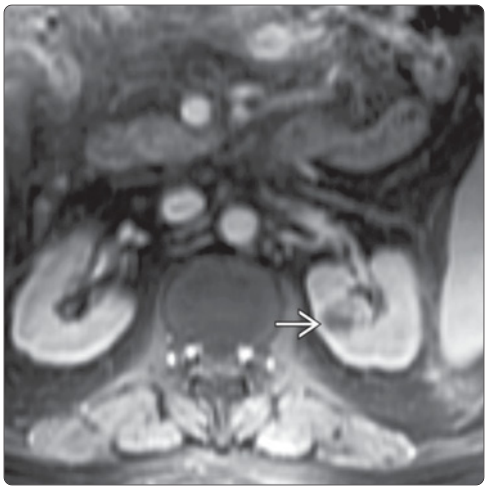


(Left) *Transverse transabdominal ultrasound shows a 1.7 cm T1a renal cell carcinoma (calipers) that is iso- to slightly hyperechoic compared to normal background renal cortical tissue.* **(Right)** *Axial CECT shows a T1a renal cell carcinoma in the lateral aspect of the mid-pole right kidney. The mass is low attenuation with areas of enhancement, characteristic of many renal cell carcinomas.*

Stage I (T1a N0 M0)

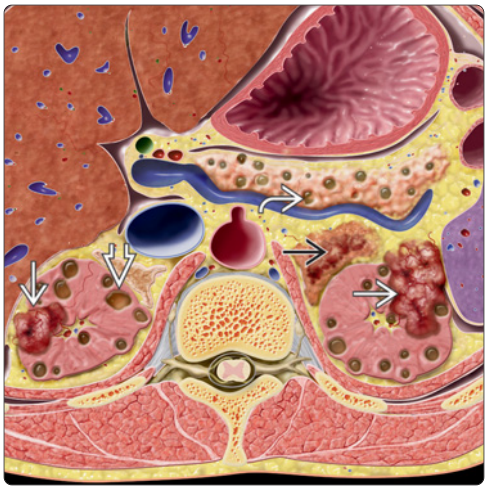


Stage I (T1a N0 M0)

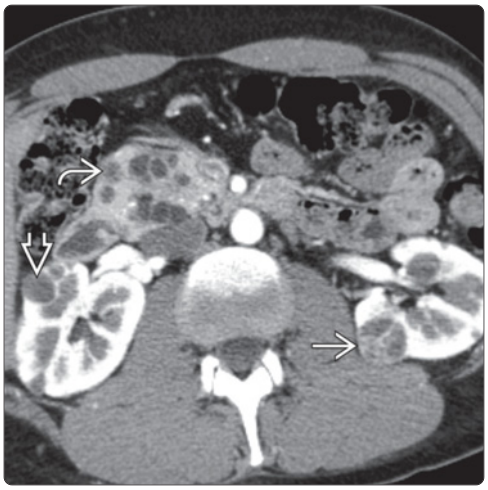


(Left) *Axial CECT shows an enhancing low-attenuation lesion ➡ measuring just over 1 cm in the right kidney. The lesion was discovered incidentally and proved to be a small renal cell carcinoma (T1a) at pathology.* **(Right)** *Axial T1WI C+ FS MR shows a 1.5 cm mass ➡ in the medial aspect of the mid-pole of the left kidney with heterogeneous signal characteristics and some enhancement, compatible with a T1a renal cell carcinoma.*

Stage I (T1a N0 M0)

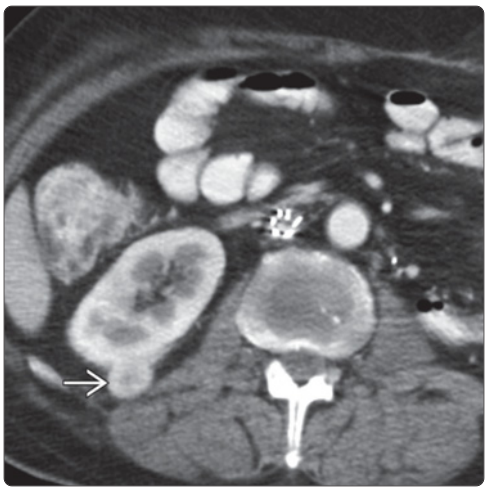


Stage I (T1a N0 M0)

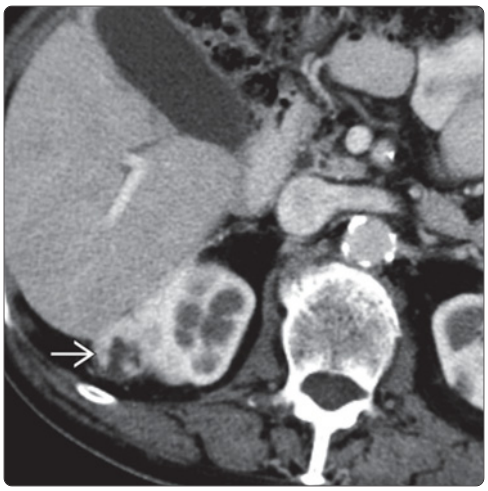


(Left) *Axial graphic shows multiple renal cysts ➡, pancreatic cysts ➡, a pheochromocytoma ⇨, and bilateral renal cell carcinomas ➡ in a patient with von Hippel-Lindau syndrome. As these lesions are confined to the kidney and < 4 cm in their largest dimension, they are all T1a.* **(Right)** *Axial CECT in a patient with von Hippel-Lindau shows multiple renal cysts ➡, pancreatic cysts ➡, and a 2 cm renal cell carcinoma ➡, compatible with a T1a lesion.*

Stage I (T1a N0 M0)

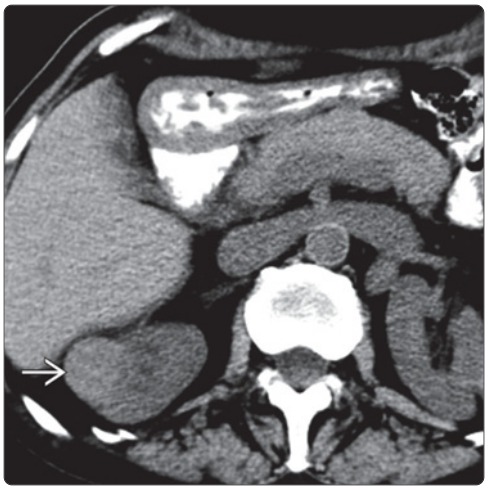


Stage I (T1a N0 M0)

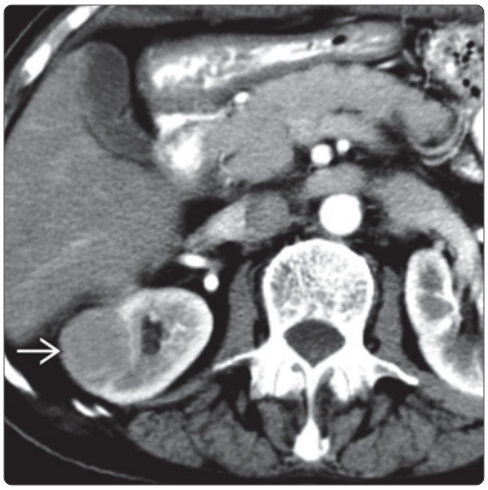


(Left) *Axial CECT shows an exophytic, enhancing, solid mass ➡ in the posterior aspect of the right kidney, measuring approximately 2 cm and compatible with a T1a lesion.* **(Right)** *Axial CECT shows a heterogeneously enhancing exophytic mass ➡ with areas of central low attenuation or necrosis arising off the lateral aspect of the right kidney, compatible with a renal cell carcinoma.*

Stage I (T1a N0 M0)

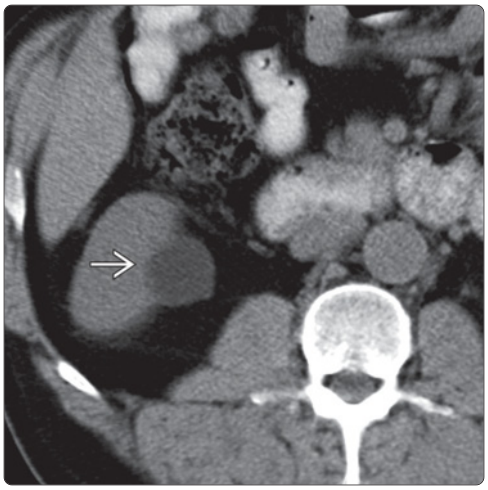


Stage I (T1a N0 M0)

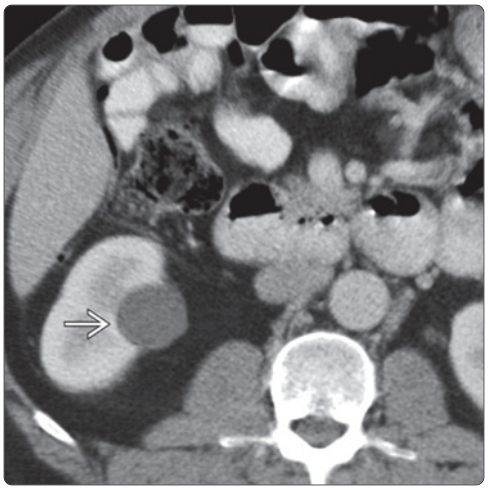


(Left) *Axial NECT in a patient with a papillary renal cell carcinoma shows a solid mass ⇨ arising from the lateral superior pole of the right kidney and measuring approximately 30 Hounsfield units.* **(Right)** *Axial CECT in the same patient shows the region of interest post contrast at 50 Hounsfield units ⇨, indicating mild enhancement. Renal cell carcinomas generally show enhancement, though papillary cell types may enhance only mildly.*

Stage I (T1a N0 M0)

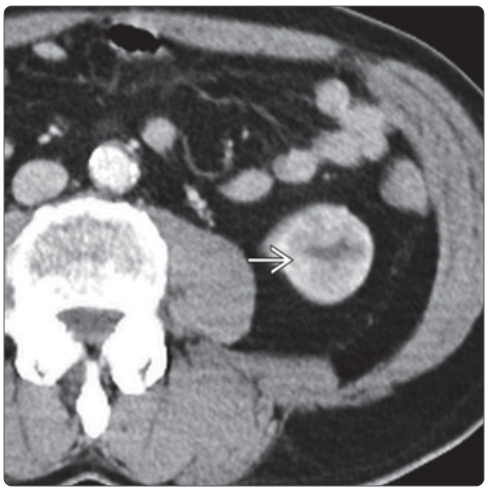


Stage I (T1a N0 M0)

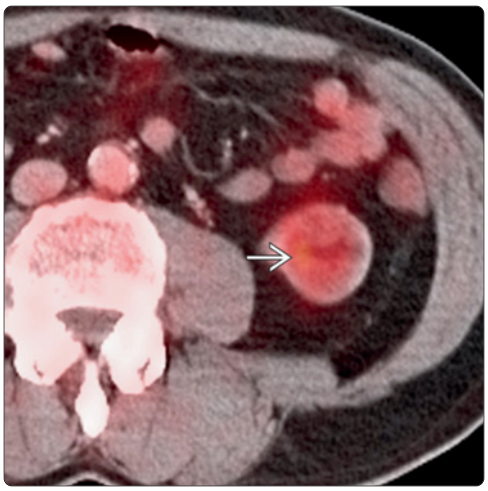


(Left) *Axial NECT shows a solid mass ⇨ arising from the medial inferior pole of the right kidney and measuring 23 HU.* **(Right)** *Axial CECT in the same patient demonstrates the mass ⇨ with post-contrast region of interest showing 39 HU, indicating mild enhancement. Pathology confirmed papillary cell type. Being less than 4 cm, this lesion would be categorized T1a.*

Stage I (T1a N0 M0)

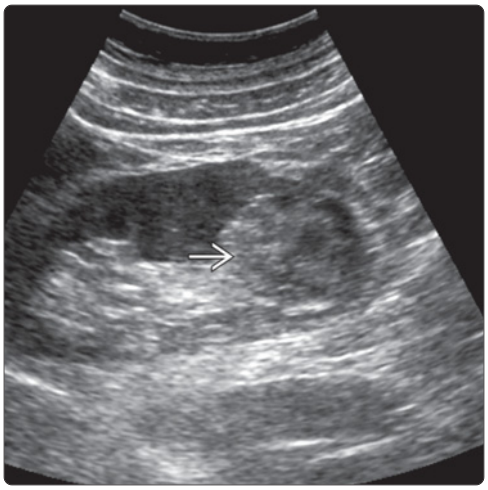


Stage I (T1a N0 M0)

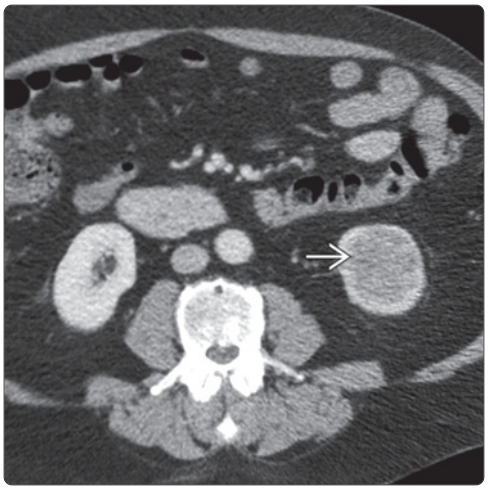


(Left) *Axial CECT shows a 3.1 cm solid enhancing mass ⇨ in the inferior pole of the left kidney with areas of central necrosis, compatible with a T1a renal cell carcinoma.* **(Right)** *Axial fused PET/CT in the same patient shows minimal diffuse FDG activity in the mass with the exception of an area of slightly more increased activity medially ⇨. Renal cell carcinomas tend to have variable FDG activity, but may have almost no increased metabolic activity.*

Stage I (T1a N0 M0)

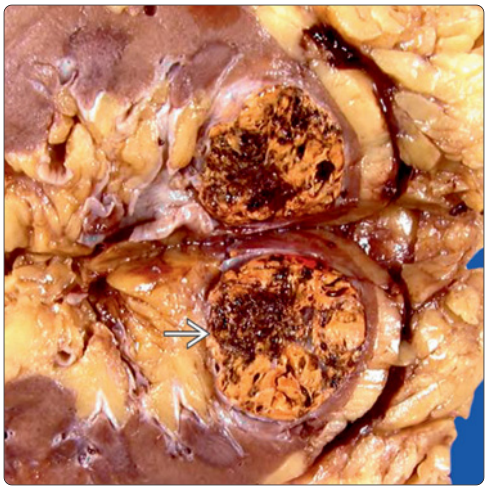


Stage I (T1a N0 M0)

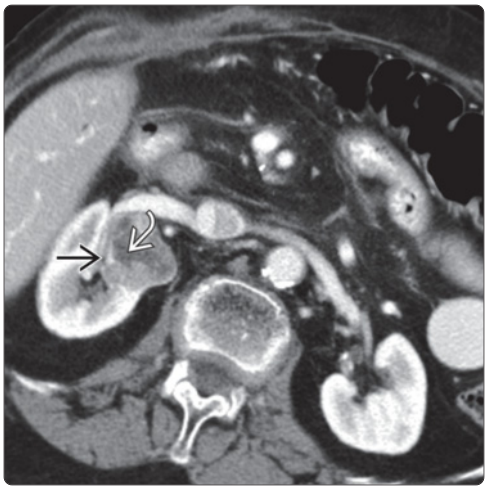


(Left) *Transverse transabdominal ultrasound shows a slightly hyperechoic, almost 3 cm, solid mass in the inferior pole of the left kidney. The ultrasound findings are worrisome for a renal cell carcinoma. A 3 cm mass would be defined as a T1a lesion.* **(Right)** *Axial CECT in the same patient shows a moderately enhancing solid renal mass that is compatible with a renal cell carcinoma. Pathology confirmed a T1a clear cell carcinoma.*

Stage I (T1a N0 M0)

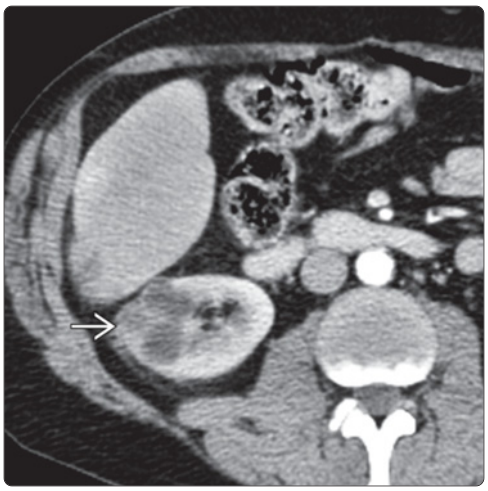


Stage I (T1a N0 M0)

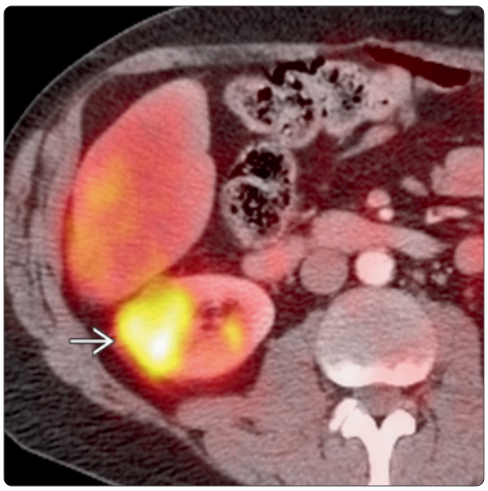


(Left) *Bisected correlative pathologic specimen from the same patient shows the mass in the inferior pole left kidney to be 2.8 cm, confined to the kidney, and without positive nodes, compatible with a stage I (T1a N0 M0) lesion.* **(Right)** *Axial CECT in another patient shows a 3 cm mass in the medial aspect of the mid-pole right kidney, displaying areas of peripheral solid enhancement and central low attenuation, likely indicating areas of necrosis.*

Stage I (T1a N0 M0)

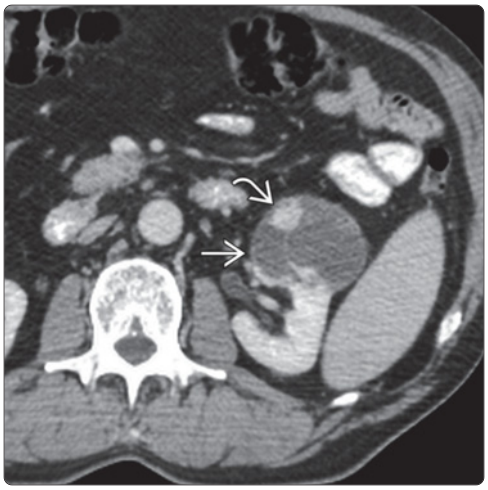


Stage I (T1a N0 M0)

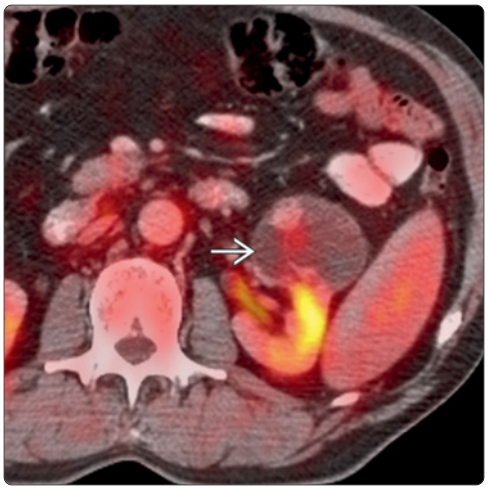


(Left) *Axial CECT shows a complex mass arising from the lateral aspect of the mid-pole of the right kidney. The low attenuation areas were solid, making this suspicious for a renal cell carcinoma.* **(Right)** *Axial fused PET/CT in the same patient shows intense FDG activity in the complex renal cortical mass, compatible with a T1a renal cell carcinoma. In contrast to the prior case, there is intense FDG activity.*

Stage I (T1b N0 M0)

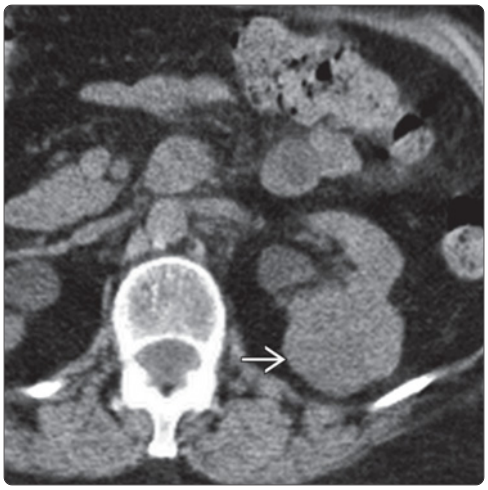


Stage I (T1b N0 M0)

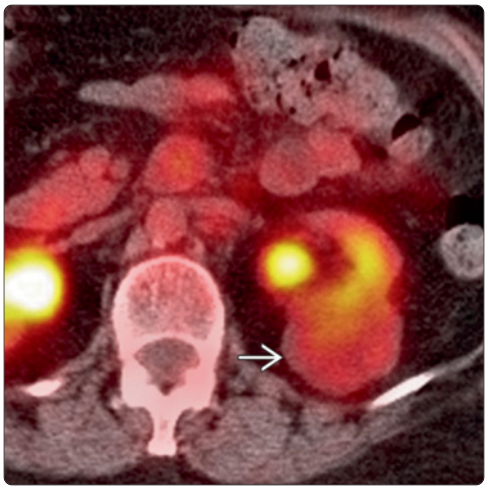


(Left) *Axial CECT shows a 4.3 cm complex cystic mass ➡ arising from the anterior aspect of the mid-pole of the left kidney with enhancing mural nodularity ➡, compatible with a T1b renal cell carcinoma.* **(Right)** *Axial fused PET/CT in the same patient shows almost no increased metabolic activity ➡ within the renal cell carcinoma, a fairly common finding identified on FDG PET in patients with renal cell carcinoma.*

Stage I (T1b N0 M0)

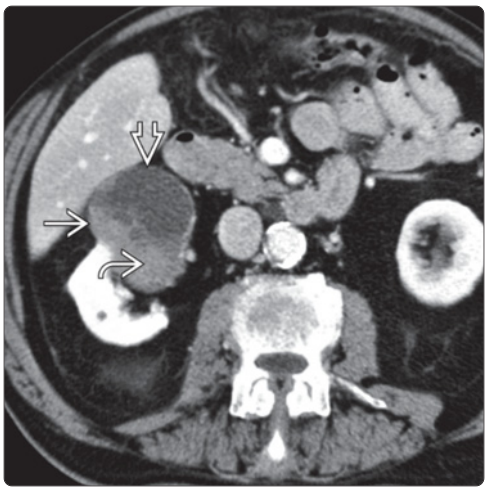


Stage I (T1b N0 M0)

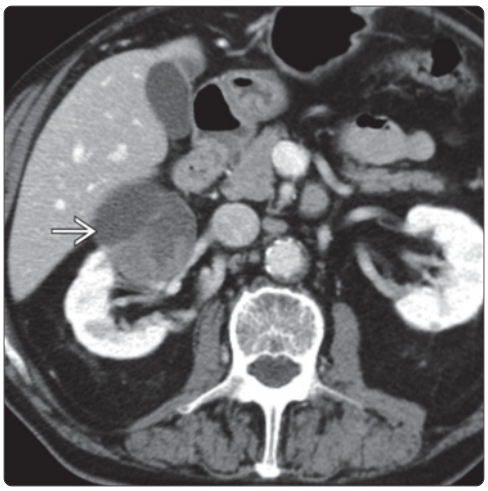


(Left) *Axial NECT shows an exophytic solid renal mass ➡ in the posterior aspect of the mid-pole left kidney, subsequently shown to be a 4.6 cm (T1b) renal cell carcinoma.* **(Right)** *Axial fused PET/CT in the same patient shows very little FDG activity in the renal cell carcinoma ➡. As renal cell carcinomas tend to have variable metabolic activity, the lack of increased metabolic activity does not exclude a renal cell carcinoma.*

Stage I (T1b N0 M0)

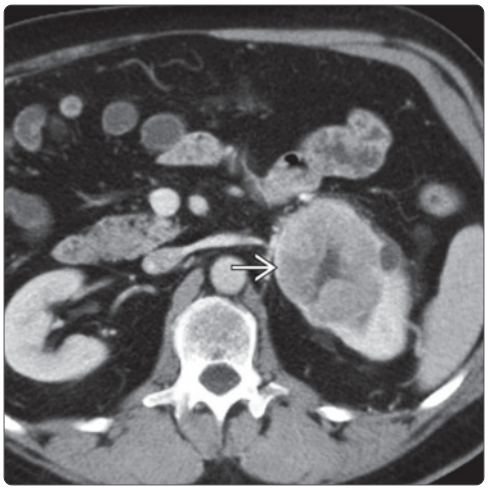


Stage I (T1b N0 M0)

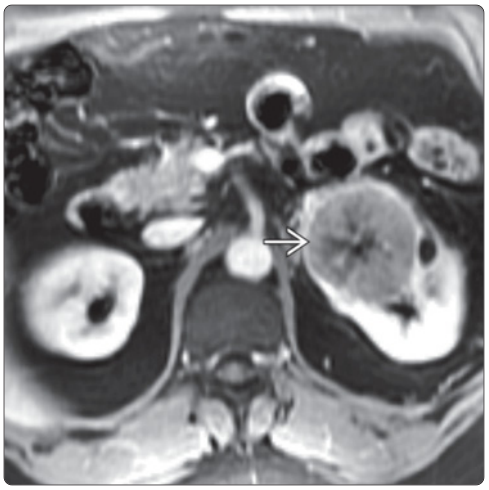


(Left) *Axial CECT shows a complex cystic mass ➡ in the anterior mid-pole of the right kidney with areas of enhancement posteriorly ➡ and more cystic areas anteriorly ➡, worrisome for a renal cell carcinoma.* **(Right)** *Axial CECT in the same patient shows similar findings of a renal mass ➡ with cystic complexity (areas of solid and cystic change). Subsequent nephrectomy showed a 4.7 cm (T1b) renal cell carcinoma.*

Stage I (T1b N0 M0)

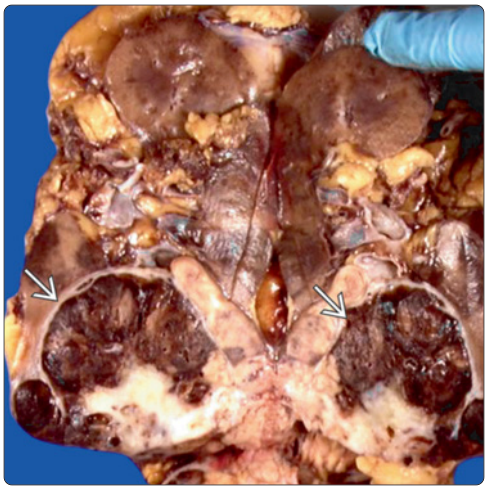


Stage I (T1b N0 M0)

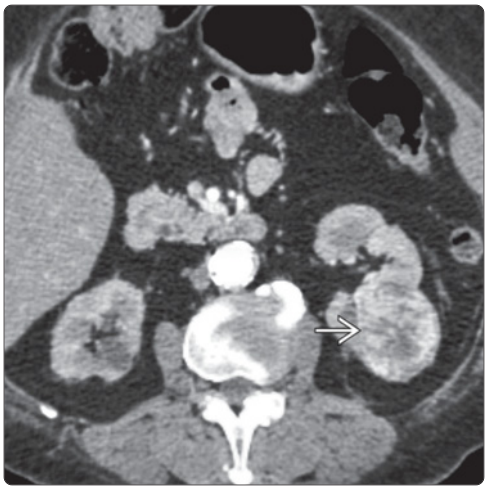


(Left) *Axial CECT shows a 5 cm mass in the left kidney with heterogeneous enhancement and central low attenuation, compatible with a renal cell carcinoma. The differential diagnosis may include an oncocytoma, which also characteristically has a central scar.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows a correlative MR image of an enhancing solid mass in the left kidney, most compatible with a renal cell carcinoma.*

Stage I (T1b N0 M0)



Stage I (T1b N0 M0)

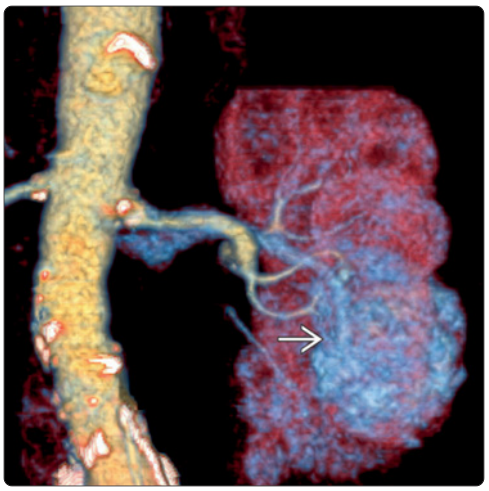


(Left) *Cut gross pathologic specimen from the same patient shows the heterogeneous mass in the left kidney. This proved to be a stage I lesion (T1b).* **(Right)** *Axial CECT in another patient shows a fairly vascular mass in the posterior aspect of the mid-pole of the left kidney. The mass measures over 4 cm and is compatible with a T1b renal cell carcinoma.*

Stage I (T1b N0 M0)



Stage I (T1b N0 M0)

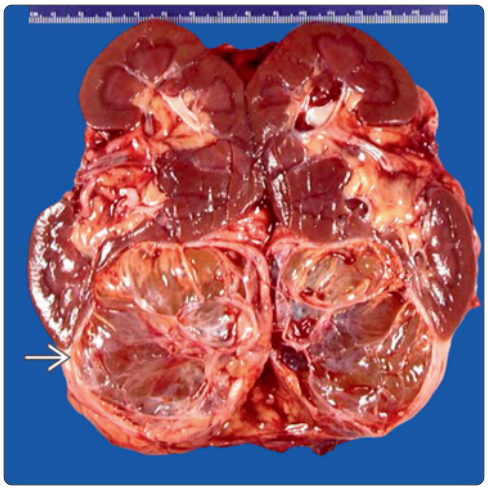


(Left) *Coronal CECT in the same patient shows the renal cell carcinoma to be relatively vascular compared to the background renal parenchyma.* **(Right)** *Reconstructed image from an abdominal CTA in the same patient as the previous 2 images shows the relative vascularity of the inferior pole renal cell carcinoma compared to the background renal vascularity.*

Stage I (T1b N0 M0)



Stage I (T1b N0 M0)

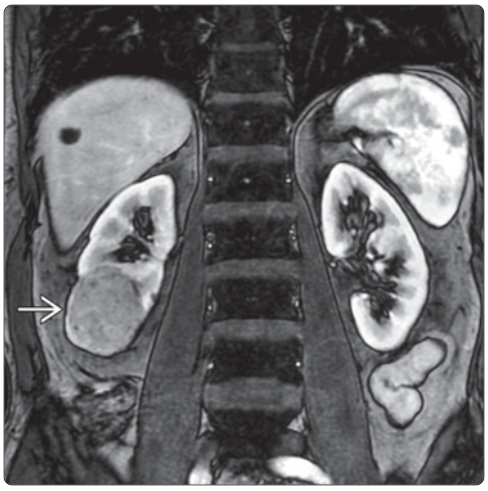


(Left) *Coronal T2WI FS MR shows a heterogeneous mass in the inferior pole of the left kidney, worrisome for a renal cell carcinoma. This lesion measures approximately 5.5 cm and is compatible with a T1b lesion.* **(Right)** *Correlative coronal cut gross pathologic specimen shows the complex inferior pole lesion, which was found to be a clear cell carcinoma of the kidney.*

Stage I (T1b N0 M0)

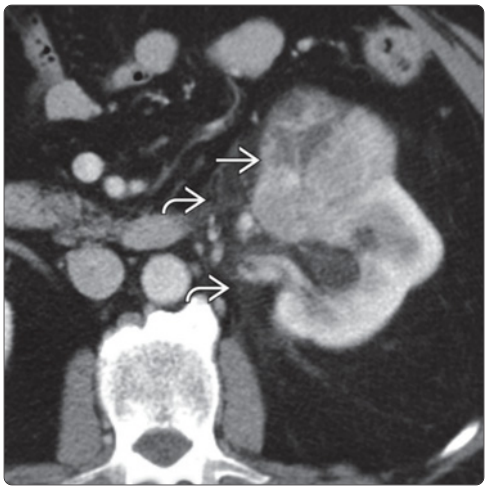


Stage I (T1b N0 M0)

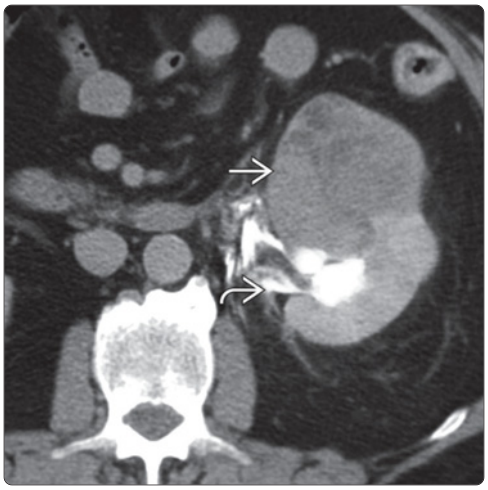


(Left) *Axial T1WI C+ FS MR shows an example of a T1b lesion in the posterior aspect of the inferior pole right kidney. In this case, the margins of the tumor are fairly well circumscribed, and the internal signal of the lesion is somewhat complex.* **(Right)** *Coronal T1WI C+ FS MR shows a 5.5 cm enhancing mass in the inferior pole of the right kidney, subsequently confirmed pathologically as a renal cell carcinoma.*

Stage I (T1b N0 M0)

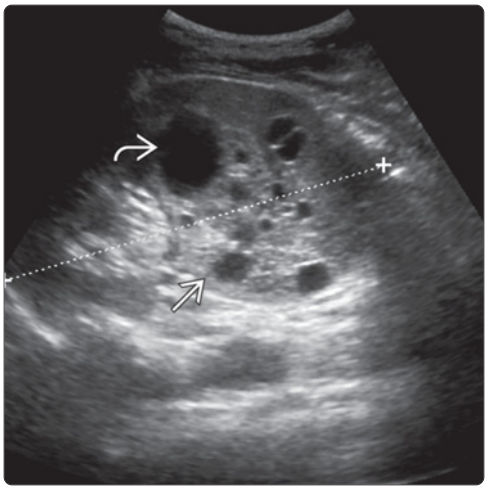


Stage I (T1b N0 M0)

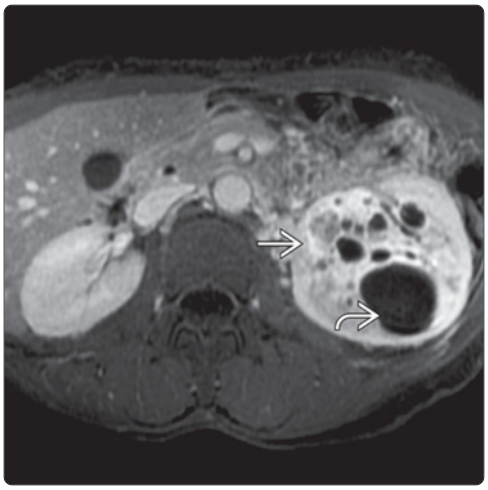


(Left) *Axial CECT shows a heterogeneously enhancing mass arising in the left kidney. The mass measures 5.6 cm and is compatible with a T1b lesion. The etiology of the perinephric stranding medially is unclear on this image.* **(Right)** *Axial CECT in the same patient with a 12-minute delay shows contrast extravasation from a ruptured calyx secondary to obstruction and possible direct tumor invasion.*

Stage I (T1b N0 M0)

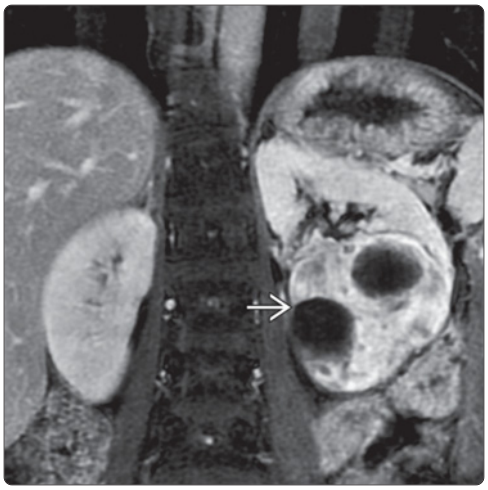


Stage I (T1b N0 M0)

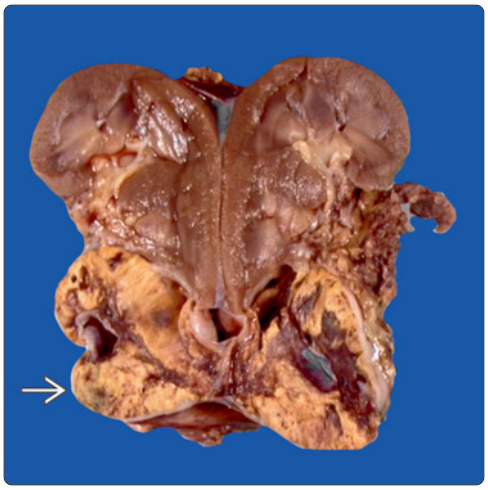


(Left) *Longitudinal transabdominal ultrasound shows a 5.8 cm (T1b) mass arising from the inferior pole of the left kidney with heterogeneous echotexture and areas of cystic change.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows a correlative MR image of the complex inferior pole renal cell carcinoma in the left kidney with central cystic areas.*

Stage I (T1b N0 M0)

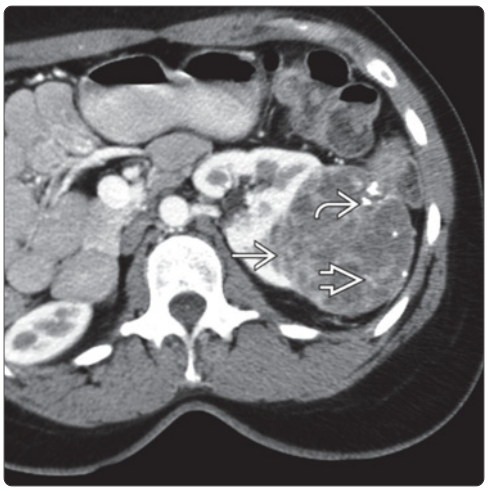


Stage I (T1b N0 M0)

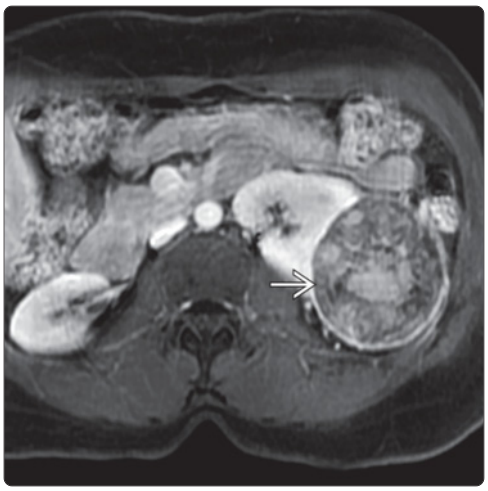


(Left) *Coronal T1WI C+ FS MR in the same patient shows the mass prior to nephrectomy, again displaying complex internal signal representing areas of solid and cystic components. No regional adenopathy was identified.* **(Right)** *Cut gross pathologic specimen from the same patient shows the renal cell carcinoma as a yellowish-red lesion with areas of internal cystic change. The final pathology was stage I.*

Stage I (T1b N0 M0)

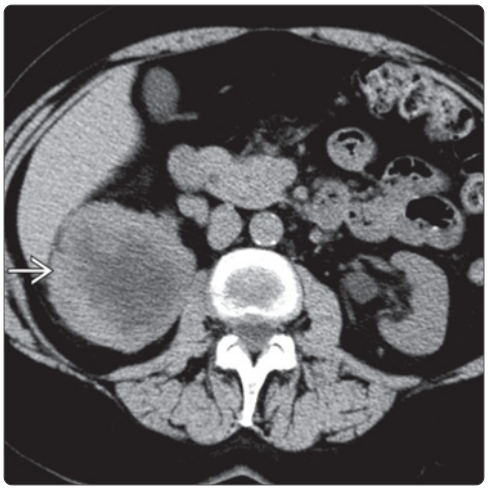


Stage I (T1b N0 M0)



(Left) *Axial CECT shows a 6 cm complex mass in the left kidney. The mass has internal components characteristic of renal cell carcinoma, including multiple enhancing septations and calcifications. The size is compatible with a T1b renal cell carcinoma.* **(Right)** *Axial T1WI C+ FS MR shows a 6.2 cm solid mass arising from the posterior aspect of the mid-pole of the left kidney with complex internal signal. These findings are compatible with a T1b renal cell carcinoma.*

Stage I (T1b N0 M0)



Stage I (T1b N0 M0)

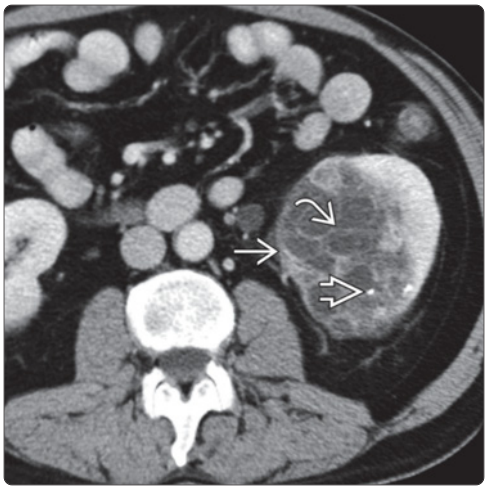


(Left) *Axial NECT shows a 6.4 cm mass arising from the superior pole of the right kidney that is suspicious for a primary renal cell carcinoma (T1b), although it is incompletely evaluated on this NECT.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows a large complex mass in the right kidney with areas of abnormal enhancement, compatible with a primary renal cell carcinoma.*

Stage I (T1b N0 M0)

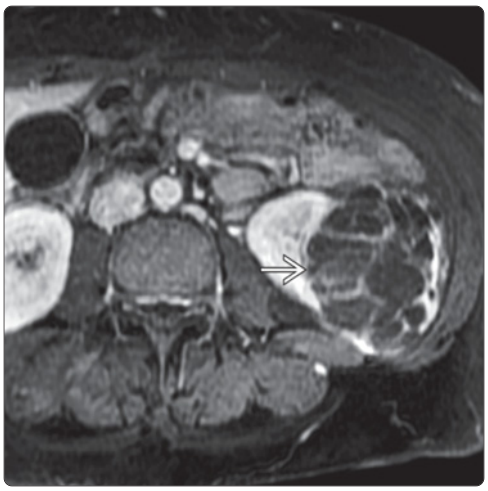


Stage I (T1b N0 M0)

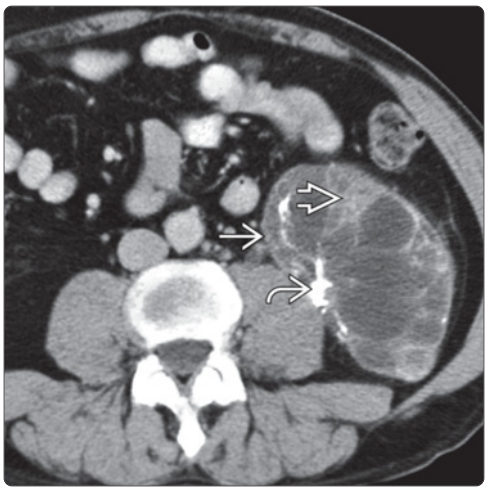


(Left) *Coronal T1WI C+ FS MR in the same patient shows the complex mass arising from the superior pole right kidney. Subsequent nephrectomy demonstrated a stage I renal cell carcinoma.* **(Right)** *Axial CECT in another patient shows a 6.6 cm complex cystic mass arising from the medial aspect of the left kidney with multiple enhancing internal septa and focal calcifications. The imaging characteristics are compatible with a renal cell carcinoma.*

Stage I (T1b N0 M0)



Stage II (T2a N0 M0)



(Left) *Axial T1WI C+ FS MR shows a similar-appearing, 6.7 cm, complex cystic mass with a similar appearance in the left kidney of a different patient, compatible with a T1b cystic renal cell carcinoma.* **(Right)** *Axial CECT shows a complex cystic mass arising from the left kidney with multiple enhancing septa and somewhat linear areas of calcification. Subsequent nephrectomy proved a stage II renal cell carcinoma with a 7.3 cm (T2) lesion.*

Stage II (T2a N0 M0)

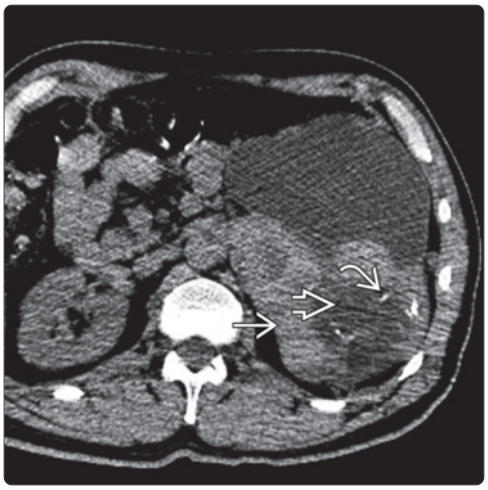


Stage II (T2a N0 M0)

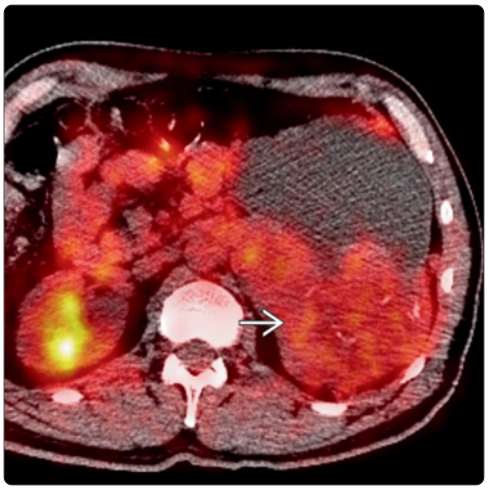


(Left) *Axial T1WI C+ FS MR shows a 7.9 cm mass in the left kidney with complex internal signal, the overall appearance of which is very worrisome for renal cell carcinoma.* **(Right)** *Coronal T1WI C+ FS MR in the same patient shows the large exophytic mass arising from the inferior pole of the left kidney. Subsequent nephrectomy revealed a stage II (T2a N0 M0) renal cell carcinoma.*

Stage II (T2a N0 M0)

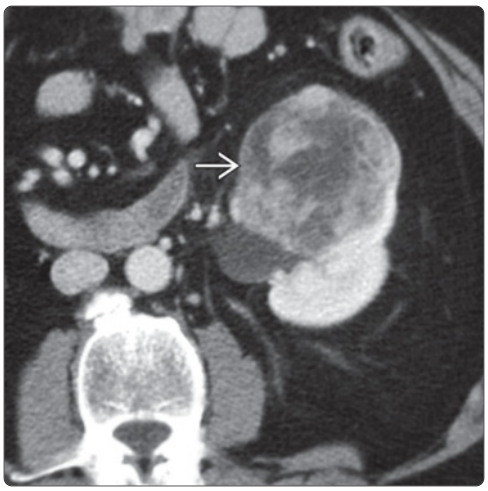


Stage II (T2a N0 M0)

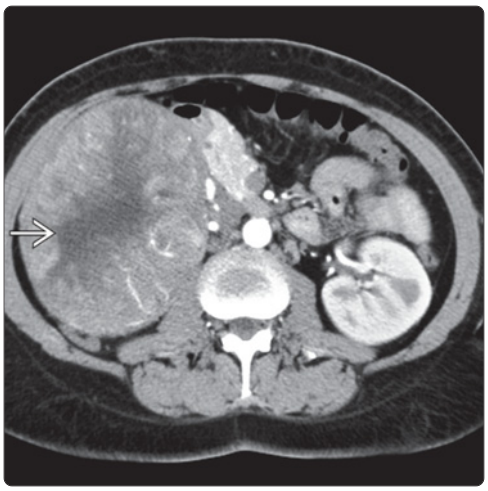


(Left) *Axial NECT performed as part of a PET/CT examination shows an 8 cm complex solid mass arising from the left kidney with internal areas of low attenuation and focal/linear calcifications, compatible with a renal cell carcinoma.* **(Right)** *Axial fused PET/CT in the same patient shows almost no increased metabolic activity within the large left renal mass. At nephrectomy, this proved to be a stage II renal cell carcinoma.*

Stage II (T2a N0 M0)



Stage II (T2a N0 M0)



(Left) *Axial CECT shows an 8.5 cm (measured in the coronal plane) enhancing mass in the anterior aspect of the mid-pole left kidney. The mass was shown to be a stage II (T2a N0 M0) lesion following nephrectomy.* **(Right)** *Axial CECT shows a well-circumscribed large mass arising from the right kidney with a central area of low attenuation and thick rind of peripheral enhancing tissue, compatible with a renal cell carcinoma.*

Stage II (T2b N0 M0)

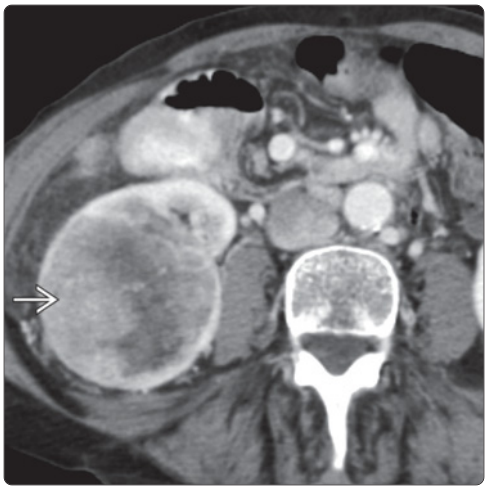


Stage II (T2b N0 M0)

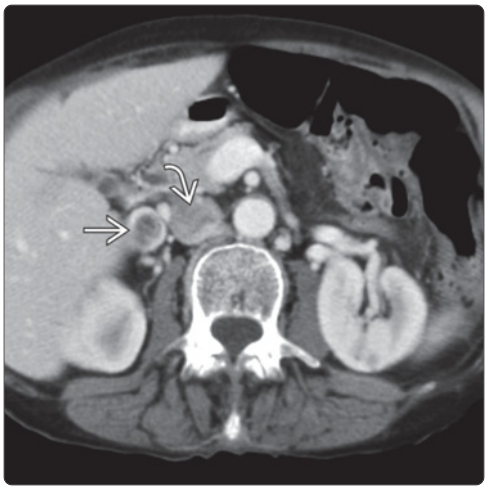


(Left) *Coronal CECT in the same patient shows a 13 cm (T2b), primarily solid, enhancing mass ⇨ arising from and confined to the right kidney, compatible with a renal cell carcinoma. Despite the large size of the primary lesion, no enlarged regional lymph nodes were identified.* **(Right)** *Gross pathologic specimen in the same patient as the previous 2 images shows the large resected mass ⇨. Final pathology demonstrated a stage II (T2b N0 M0) lesion.*

Stage III (T3a N0 M0)

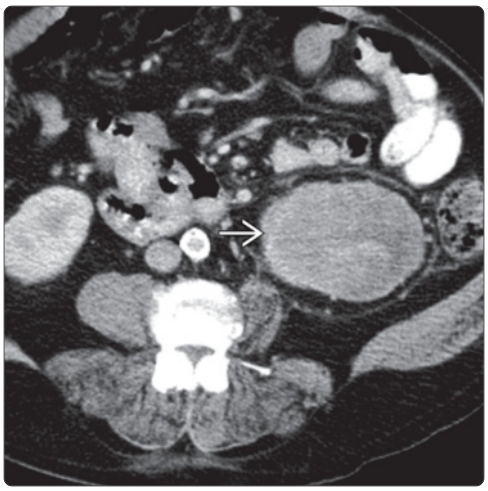


Stage III (T3a N0 M0)

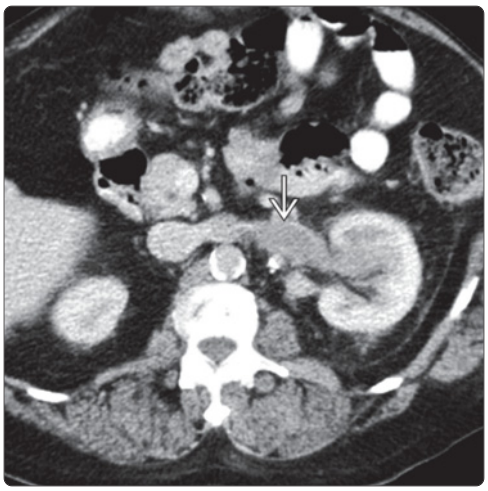


(Left) *Axial CECT shows a renal cell carcinoma ⇨ in the right kidney.* **(Right)** *The tumor is invading the right renal vein ⇨. A tumor extending into the renal vein or its segmental branches constitutes a T3a lesion; extension of the tumor in the IVC is staged as T3b; extension above the diaphragm constitutes a T3c lesion. The low attenuation within the IVC ⇨ is the result of mixing of opacified with nonopacified blood and should not be mistaken for tumor invasion.*

Stage III (T3a N0 M0)

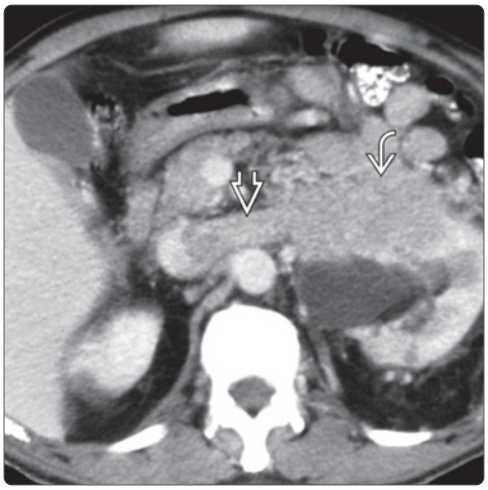


Stage III (T3a N0 M0)

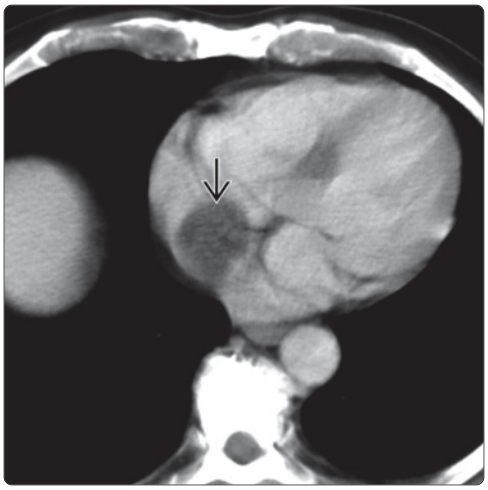


(Left) *Axial CECT shows a solid enhancing renal mass ⇨ in the left kidney, compatible with renal cell carcinoma.* **(Right)** *Axial CECT in the same patient shows enlargement of the left renal vein ⇨ secondary to tumor infiltration, making this a T3a lesion.*

Stage III (T3c N0 M0)

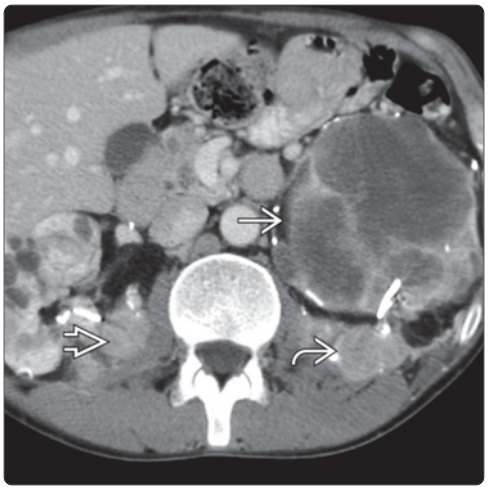


Stage III (T3c N0 M0)

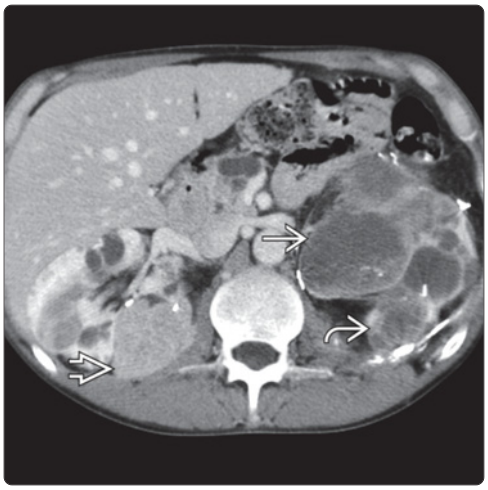


(Left) *Axial CECT shows a left renal cell carcinoma with invasion of the left renal vein. (Right) Axial CECT at the level of the heart in the same patient shows tumor thrombus within the right atrium. Extension of tumor thrombus above the diaphragm makes this a T3c tumor and has important implications for the surgical approach.*

Stage IV (T4 N0 M0)



Stage IV (T4 N0 M0)

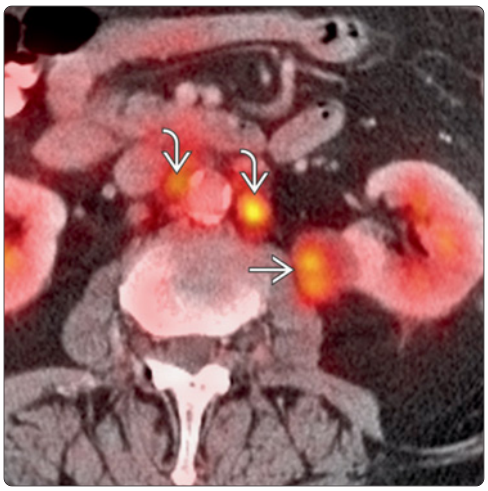


(Left) *Axial CECT in a patient with von Hippel-Lindau syndrome shows a cystic renal cell carcinoma in the left kidney with perirenal/posterior abdominal wall involvement on the ipsilateral side, and tumor in the perirenal fat on the contralateral side from another renal cell carcinoma.* **(Right)** *Axial CECT shows there are multiple contralateral renal cell carcinomas, in addition to the cystic renal cell carcinoma seen on the prior image, with involvement of the perirenal fat.*

Stage IV (T1a N1 M0)

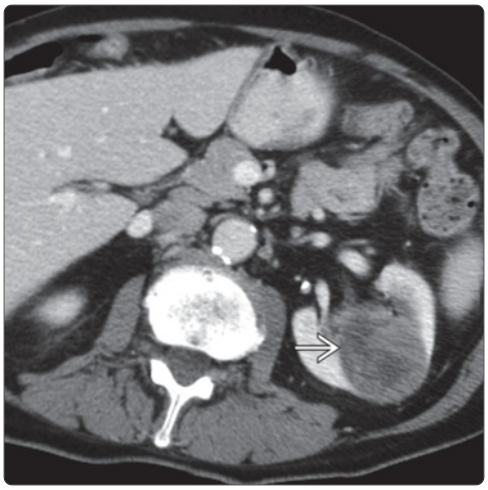


Stage IV (T1a N1 M0)

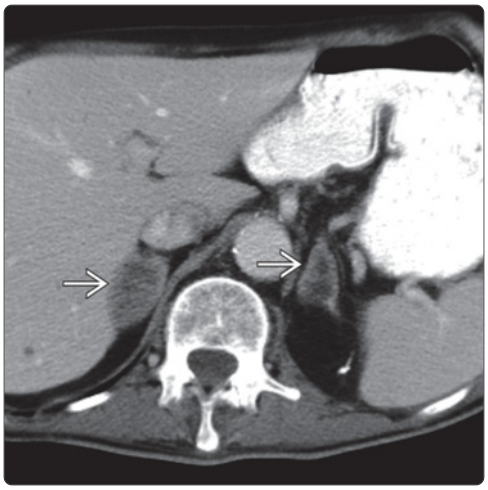


(Left) *Axial CECT shows a low-attenuation but enhancing mass within the left kidney.* **(Right)** *Axial fused PET/CT in the same patient shows moderate increased FDG activity along the medial border of this renal cell carcinoma. In addition, the left paraaortic and aortocaval nodes demonstrate increased metabolic activity compatible with metastatic nodes, making this N1 disease.*

Stage IV (T1b N0 M1)



Stage IV (T1b N0 M1)

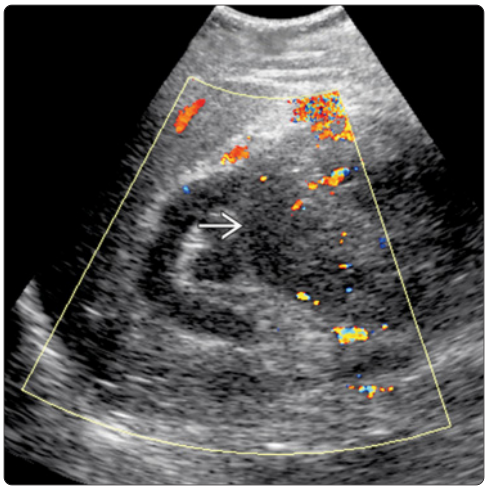


(Left) *Axial CECT shows a 4.3 cm mixed attenuation and heterogeneously enhancing mass in the left kidney, compatible with a T1b lesion.* **(Right)** *Axial CECT in the same patient shows bilateral adrenal masses. These masses are indeterminate on this single contrast-enhanced CT. The patient was referred for a CT-guided biopsy of the right adrenal gland mass, which showed metastatic renal cell carcinoma.*

Stage IV (T1b N0 M1)

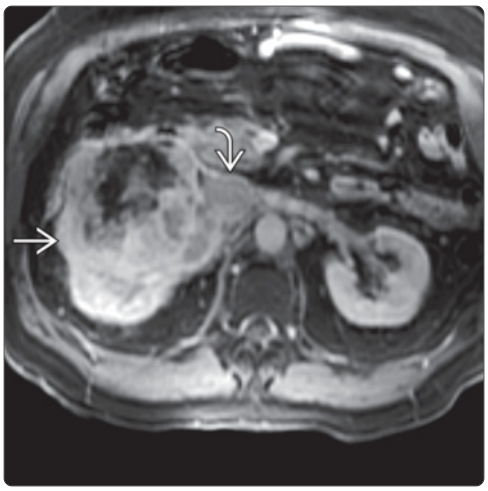


Stage IV (T3a N0 M1)

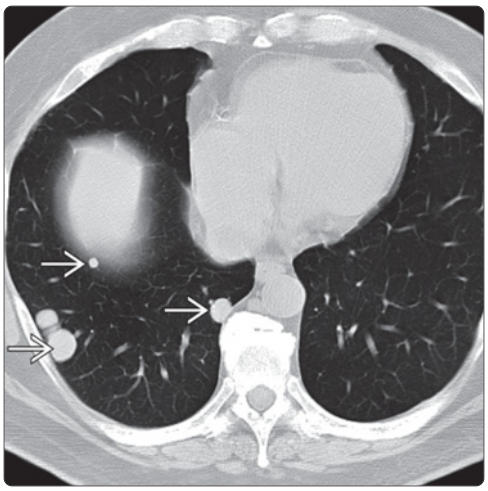


(Left) *Coronal posterior bone scan in a patient who underwent a recent nephrectomy for a 6 cm renal cell carcinoma (T1b) shows multiple abnormal foci of increased tracer activity throughout the spine, compatible with metastatic disease.* **(Right)** *Longitudinal color Doppler ultrasound in a patient with a stage IV renal cell carcinoma shows a fairly vascular mass within the right kidney.*

Stage IV (T3a N0 M1)

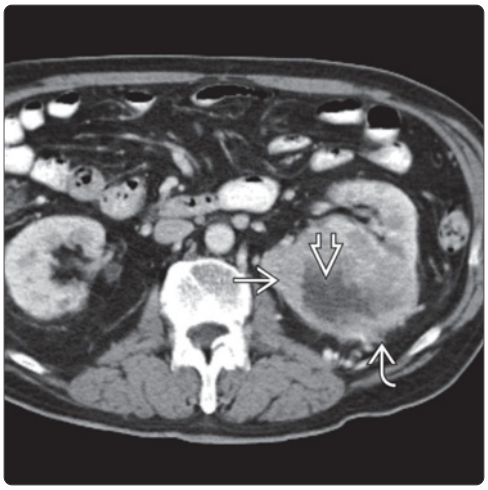


Stage IV (T3a N0 M1)

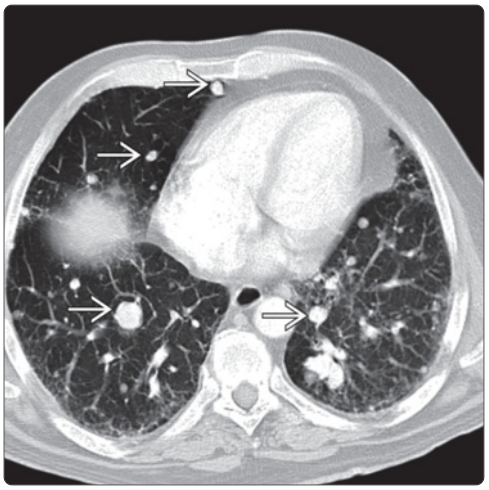


(Left) *Axial T1WI C+ FS MR the same patient shows a heterogeneously enhancing mass in the right kidney with tumor extension into the right renal vein.* **(Right)** *Axial CECT in the same patient shows multiple well-circumscribed, lower lobe pulmonary nodules on the right, compatible with M1 or stage IV disease.*

Stage IV (T3a N0 M1)

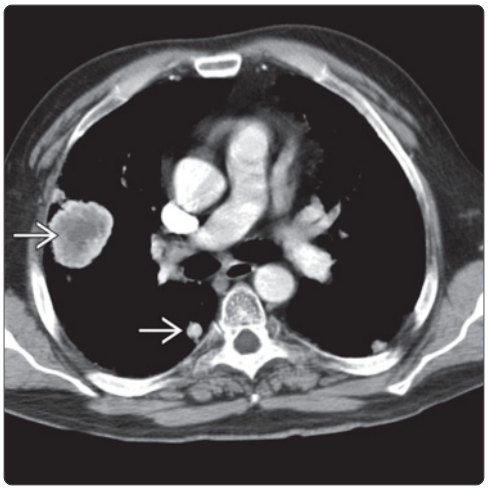


Stage IV (T3a N0 M1)

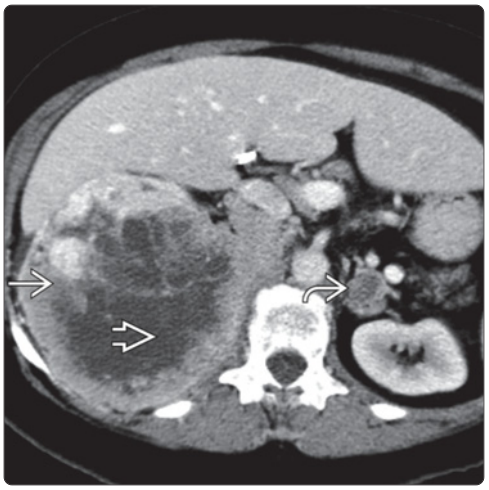


(Left) *Axial CECT shows a large, diffusely enhancing mass arising from the posterior aspect of the superior pole left kidney with central necrosis and early extension of tumor into the perirenal space.* **(Right)** *Axial CECT in the same patient shows multiple well-circumscribed, bilateral, noncalcified pulmonary nodules, compatible with pulmonary metastases.*

Stage IV (T3a N0 M1)

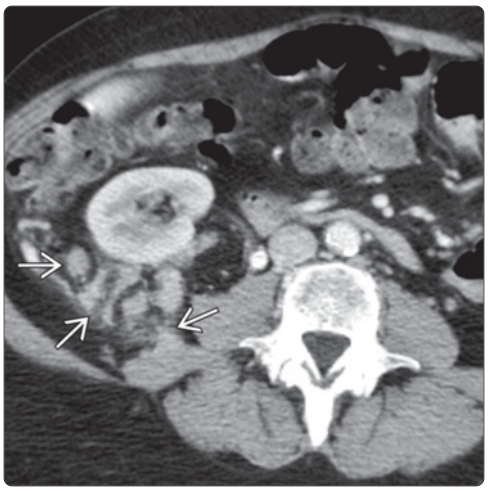


Stage IV (T4 N0 M1)

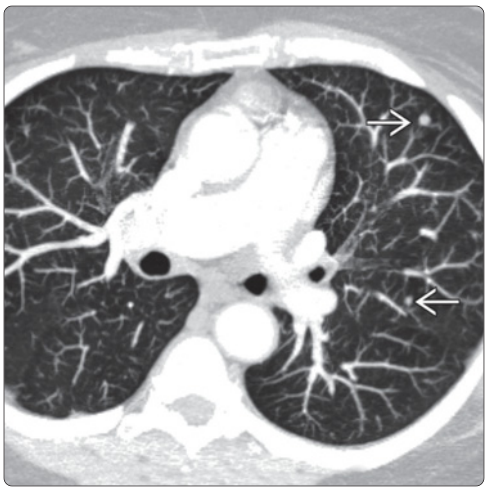


(Left) *Axial CECT in the same patient as the previous 2 images shows additional pulmonary metastases with soft tissue windows. The largest lesion in the right upper lobe shows significant central necrosis. Multiple pulmonary metastases signify stage IV disease.* **(Right)** *Axial CECT in a different patient shows a large, enhancing, right renal mass with central necrosis and a contralateral adrenal metastasis.*

Stage IV (T4 N0 M1)

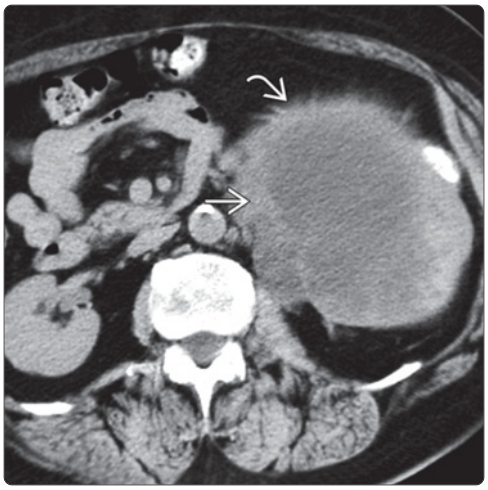


Stage IV (T4 N0 M1)

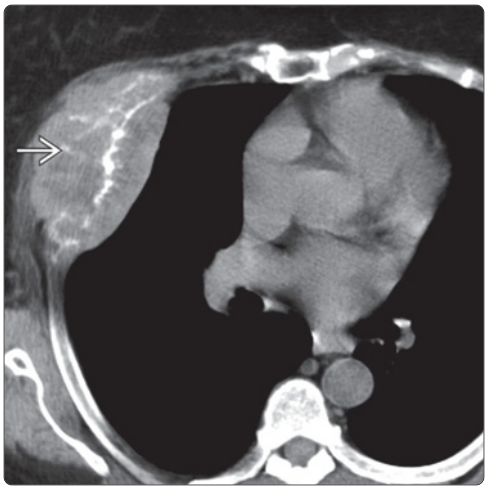


(Left) *Axial CECT in the same patient shows extensive infiltration and nodularity of the perirenal fat on the right that extended beyond the Gerota fascia (T4).* **(Right)** *Axial CECT MIP in the same patient as the previous 2 images shows 2 small, well-circumscribed, left upper lobe nodules that were not present on a CT scan of the chest 2 years earlier. These are most likely metastatic lesions. With adrenal metastasis and 2 lung nodules, this is M1 or stage IV disease.*

Stage IV (T4 N0 M1)

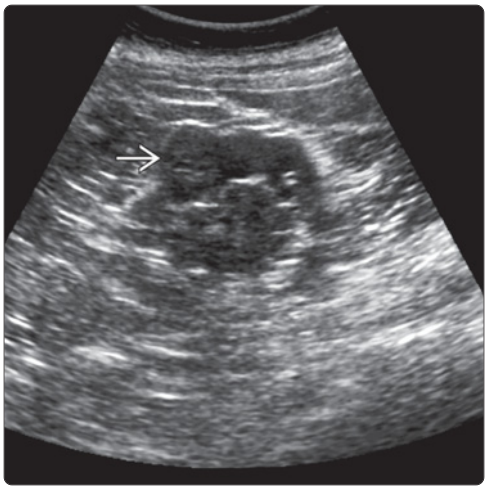


Stage IV (T4 N0 M1)

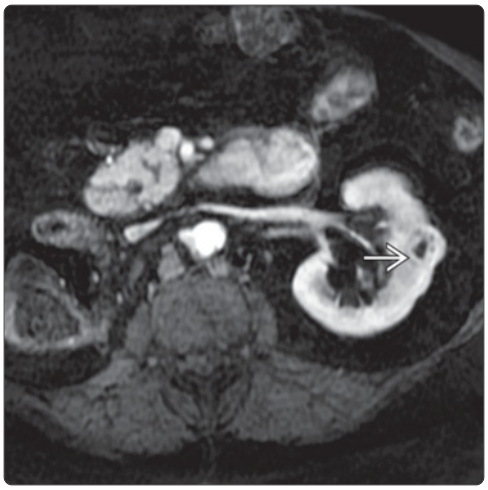


(Left) *Axial NECT shows a large heterogeneous mass arising in the left kidney with spiculated and ill-defined borders anteriorly and medially, signifying extension of tumor into the perirenal fat and breaching the Gerota fascia (T4).* **(Right)** *Axial NECT in the same patient shows a large destructive metastasis involving a right anterolateral rib, compatible with M1 disease.*

Stage IV (T1a N1 M1)

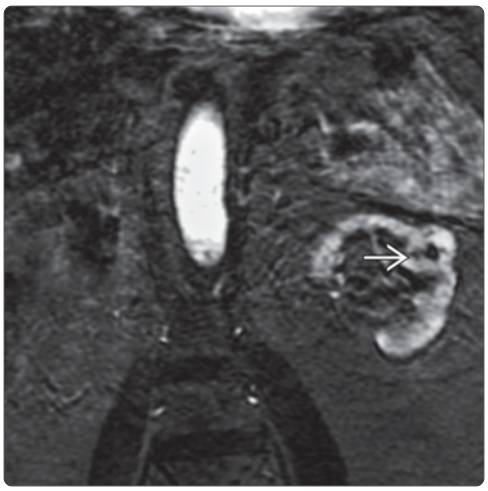


Stage IV (T1a N1 M1)

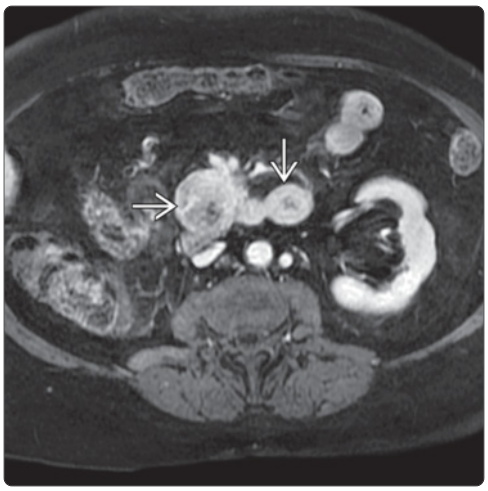


(Left) *Transverse ultrasound shows a nearly isoechoic, slightly exophytic, solid mass in the mid-pole of the left kidney. As this lesion is not completely cystic, it is worrisome for a renal cell carcinoma.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows a primarily solid renal mass, worrisome for renal cell carcinoma. The mass measures approximately 2 cm and would be compatible with a T1a lesion.*

Stage IV (T1a N1 M1)

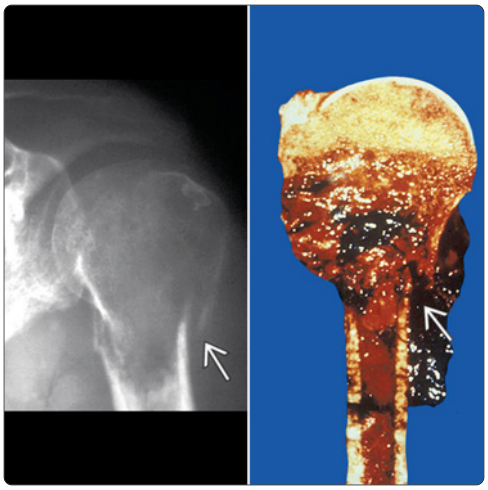


Stage IV (T1a N1 M1)

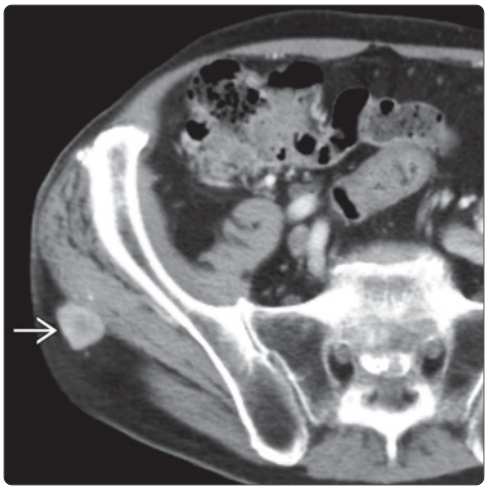


(Left) *Coronal MR in the same patient shows the same primary solid left renal mass in a different plane.* **(Right)** *Axial T1WI C+ FS MR in the same patient as the previous 3 images shows enlarged nodal metastases to the left and to the right of the aorta, despite the relatively small size of the primary tumor, that is compatible with N1 disease. The patient also had pulmonary metastases (M1) or overall stage IV disease.*

Stage IV (T3 N1 M1)

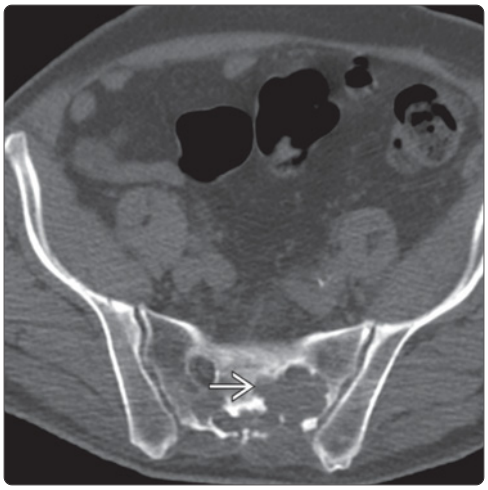


Stage IV Recurrent RCC

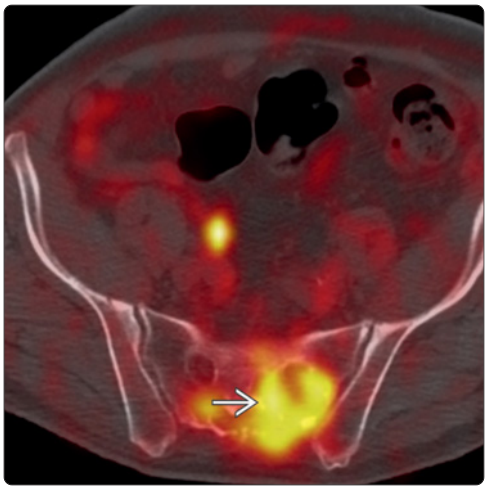


(Left) *Composite image shows a pathologic fracture ⇨ of the humerus with the corresponding specimen at autopsy. The patient presented with acute arm pain after a minor trauma. The work-up showed a T3 tumor of the kidney with extensive nodal disease and widespread metastases.* **(Right)** *Axial CECT shows a solid enhancing lesion ⇨ in the subcutaneous fat in this patient with a history of a large renal cell carcinoma, compatible with recurrent disease.*

Stage IV Recurrent RCC

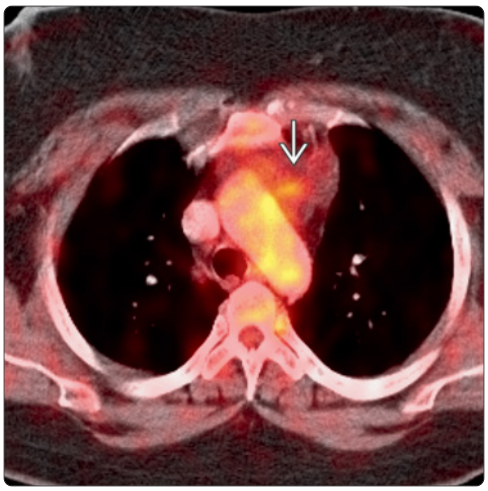


Stage IV Recurrent RCC

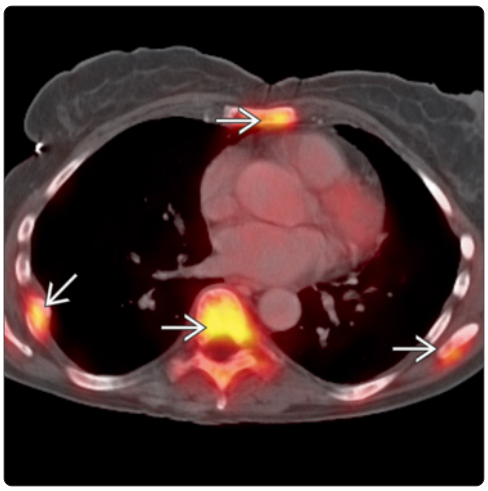


(Left) *Axial NECT in this patient with a history of renal cell carcinoma shows a lytic lesion ⇨ involving the left hemisacrum, worrisome for recurrent metastatic disease.* **(Right)** *Axial fused PET/CT in the same patient shows intense FDG activity ⇨ corresponding to the lytic lesion of the left hemisacrum, compatible with recurrent renal cell carcinoma. Metastases from renal cell carcinoma may have variable FDG uptake.*

Stage IV Recurrent RCC

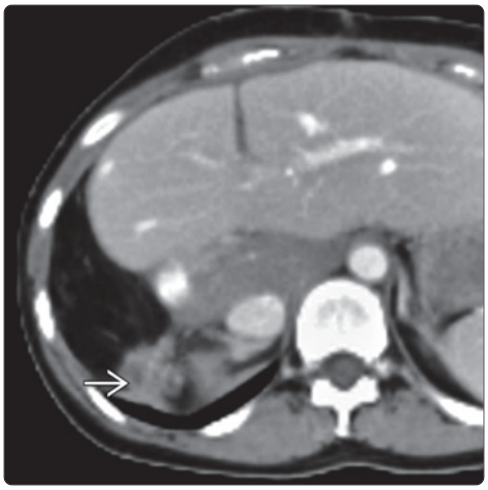


Stage IV Recurrent RCC

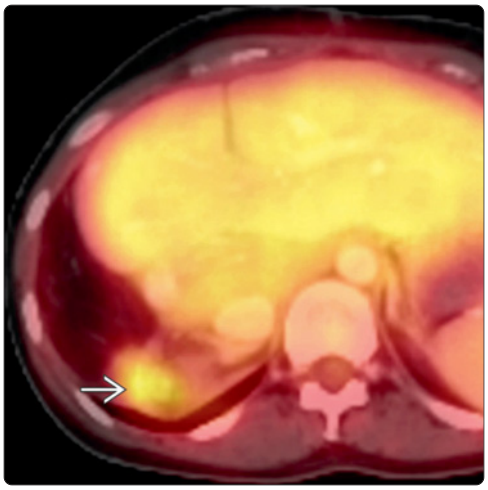


(Left) *Axial fused PET/CT in a patient with a history of renal cell carcinoma shows abnormal soft tissue ⇨ in the prevascular space with patchy areas of increased metabolic activity, compatible with metastatic disease.* **(Right)** *Axial fused PET/CT in the same patient shows multiple hypermetabolic bone lesions ⇨, compatible with osseous metastases from renal cell carcinoma.*

Stage IV Recurrent RCC

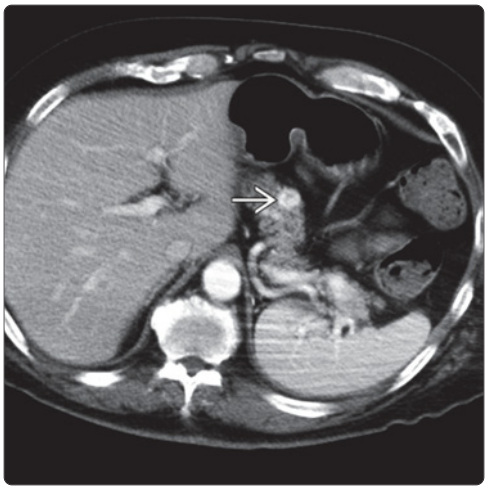


Stage IV Recurrent RCC

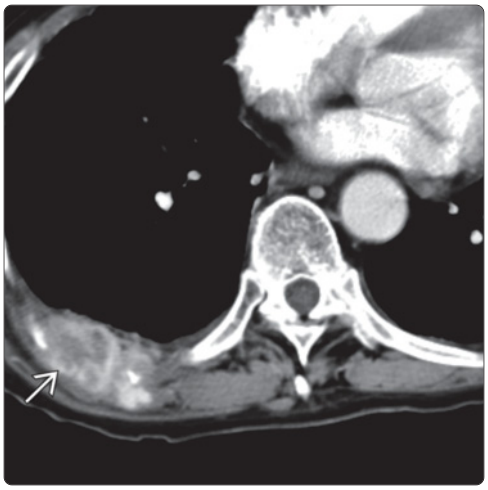


(Left) *Axial CECT in a patient with a history of renal cell carcinoma shows abnormal soft tissue in the nephrectomy bed ~ 1 year following nephrectomy, which is worrisome for recurrent disease.* **(Right)** *Axial fused PET/CT in the same patient shows moderately increased FDG activity corresponding to the soft tissue in the nephrectomy bed, compatible with recurrent disease.*

Stage IV Recurrent RCC

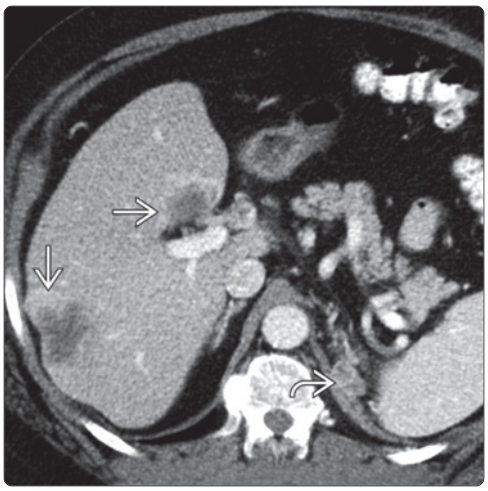


Stage IV Recurrent RCC

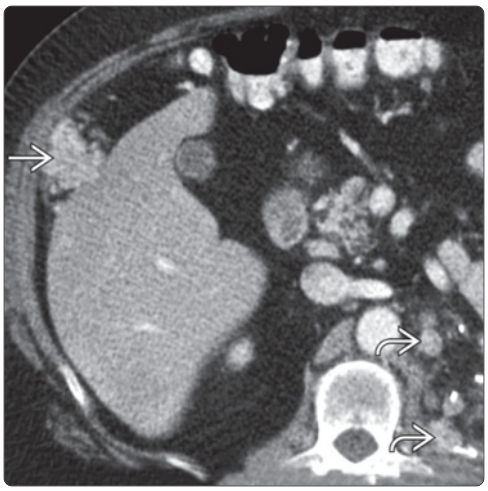


(Left) *Axial CECT in this patient with a history of renal cell carcinoma shows an enhancing lesion in the body of the pancreas, compatible with recurrent metastatic disease.* **(Right)** *Axial CECT in this patient with a history of renal cell carcinoma shows a destructive osseous lesion involving the posterior chest wall/rib, compatible with recurrent metastatic disease.*

Stage IV Recurrent RCC

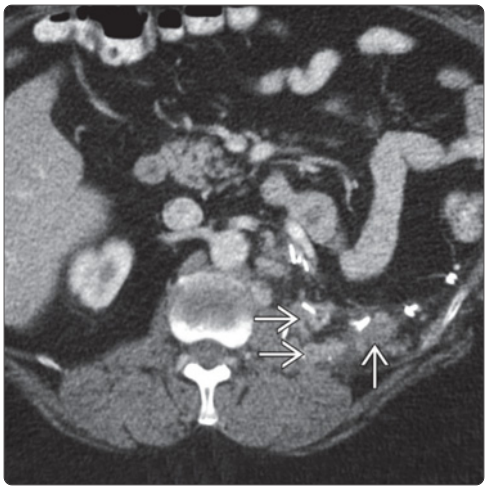


Stage IV Recurrent RCC

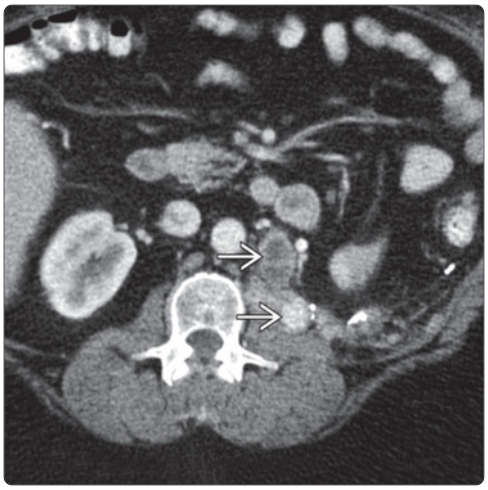


(Left) *Axial CECT shows multiple low-attenuation lesions in the liver in this patient with a history renal cell carcinoma, compatible with recurrent stage IV disease. In addition, there is recurrent disease in the left renal fossa.* **(Right)** *Axial CECT shows a metastatic implant anterior to the liver in this patient with a history of renal cell carcinoma, compatible with recurrent disease. Also note smaller nodular implants in the left nephrectomy bed.*

Stage IV Recurrent RCC

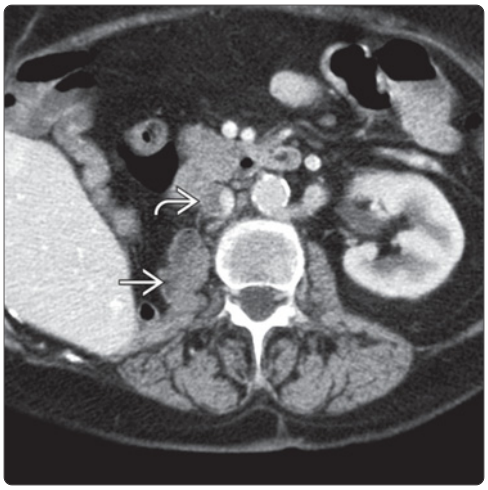


Stage IV Recurrent RCC

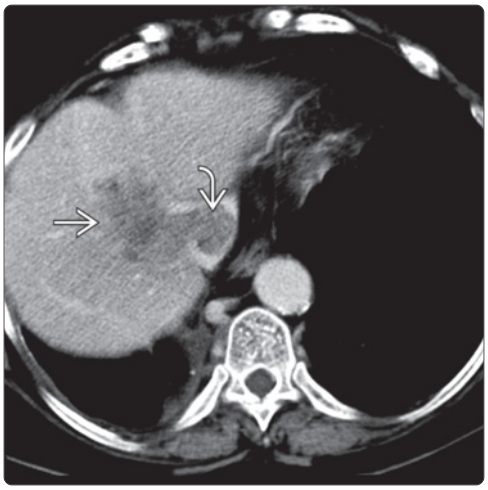


(Left) *Axial CECT shows multiple soft tissue nodular implants in this patient with a history of a left-sided renal cell carcinoma status post nephrectomy, findings compatible with local recurrent disease.* **(Right)** *Axial CECT in the same patient shows additional nodular metastatic lesions, again compatible with recurrent renal cell carcinoma.*

Stage IV Recurrent RCC

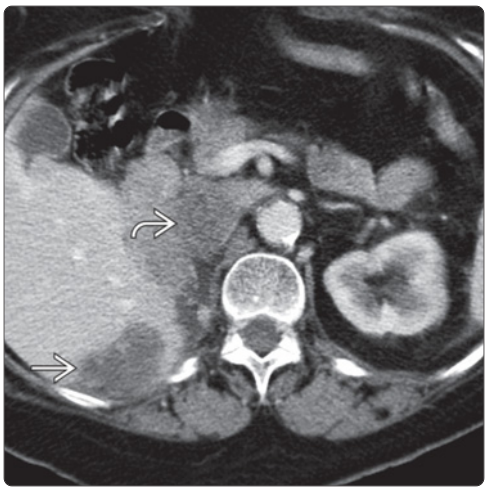


Stage IV Recurrent RCC

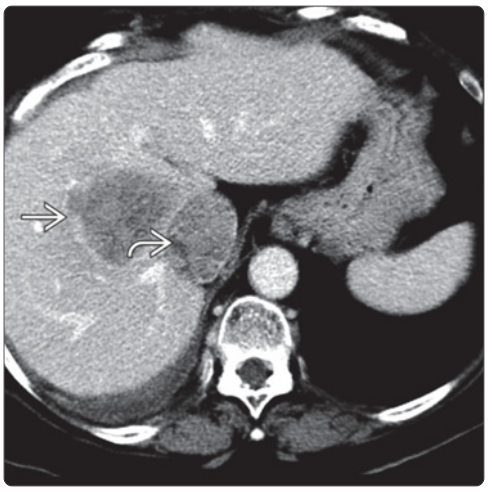


(Left) *Axial CECT in a patient with right-sided renal cell carcinoma status post right nephrectomy shows multiple soft tissue nodular lesions in the nephrectomy bed, compatible with recurrent malignancy. Also note partial thrombosis of the inferior vena cava.* **(Right)** *Axial CECT in the same patient shows extensive metastatic disease involving the liver with direct extension of tumor into the inferior vena cava.*

Stage IV Recurrent RCC



Stage IV Recurrent RCC



(Left) *Axial CECT in the same patient shows additional areas of metastatic involvement in the posterior segment of the liver and expansion of the vena cava, compatible with tumor thrombus.* **(Right)** *Axial CECT in the same patient shows an additional view of the expanded inferior vena cava, compatible with tumor thrombus as well as the metastatic lesion in the liver.*

Medullary Carcinoma

KEY FACTS

TERMINOLOGY

- Highly aggressive, infiltrative malignancy arising in renal medulla

IMAGING

- CECT
  - Ill-defined, infiltrative central renal mass
  - Hypovascular; heterogeneous enhancement
  - Caliectasis without pelviectasis
  - Necrosis and hemorrhage are common
- MR
  - T1WI: Low signal intensity
  - T2WI: Low to intermediate signal intensity
  - DWI: Restricted diffusion in areas of viable tumor
- Ultrasound
  - Mixed echogenicity central mass
  - Loss of normal renal architecture

TOP DIFFERENTIAL DIAGNOSES

- Transitional cell carcinoma (TCC)
- Lymphoma
- Renal cell carcinoma (RCC)
- Pyelonephritis

PATHOLOGY

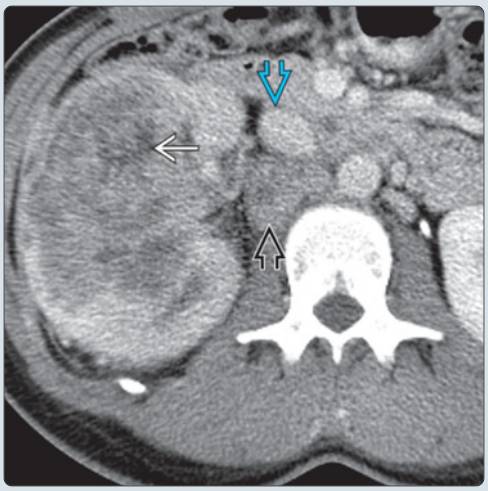
- Sheets of poorly differentiated, eosinophilic cells

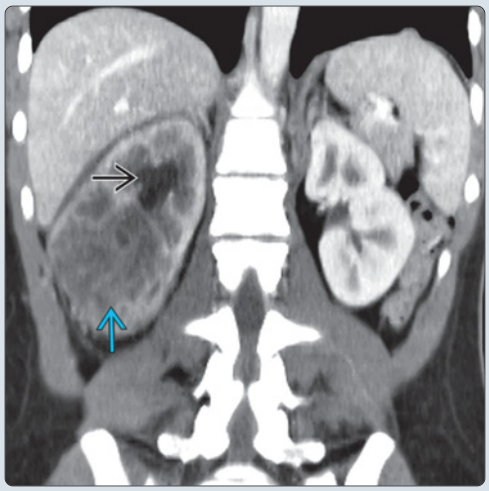
CLINICAL ISSUES

- Most commonly affects young African American males with sickle cell trait
- Most common symptoms: Flank pain, hematuria, weight loss
- Poor prognosis: Mean survival ~ 15 months

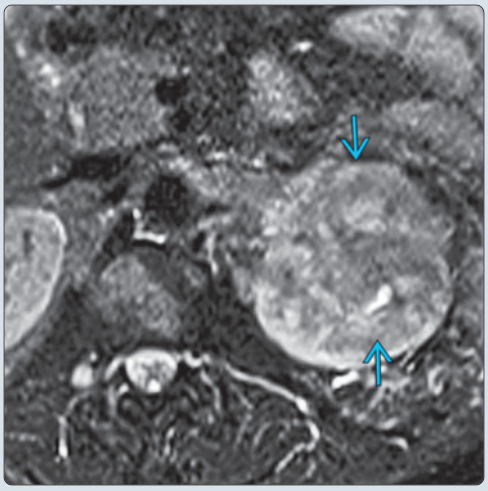
DIAGNOSTIC CHECKLIST

- Suspect in young male with sickle cell trait
- Adenopathy and metastases are common at presentation





(Left) *Axial CECT in a patient with sickle cell trait who presented with right flank pain and hematuria shows an ill-defined, infiltrative mass in the right kidney. The mass enhances heterogeneously due to areas of necrosis. Note the retrocaval adenopathy resulting in anterior displacement of the IVC.* **(Right)** *Coronal CECT in the same patient reveals that the ill-defined mass results in caliectasis in the superior collecting system but no pelviectasis. Pathology was consistent with medullary carcinoma.*





(Left) *Axial T2WI MR with fat saturation shows an ill-defined mass that infiltrates the renal cortex, medulla, and collecting system. The mass has heterogeneous low to intermediate signal intensity.* **(Right)** *Axial T1 C+ MR with fat saturation slightly more inferiorly in the same patient shows heterogeneous enhancement of the infiltrative, hypovascular mass. The tumor also extends into the expanded left renal vein.*

TERMINOLOGY

Synonyms

- 7th sickle cell nephropathy

Definitions

- Highly aggressive, infiltrative primary renal malignancy

IMAGING

General Features

- Best diagnostic clue
  - Infiltrative renal mass with metastases in young patient with sickle cell trait
- Location
  - Originating in renal medulla but can invade cortex and renal sinus
- Size
  - Variable; often difficult to accurately measure due to ill-defined borders
- Morphology
  - Infiltrative and ill-defined

CT Findings

- NECT
  - Infiltrative soft tissue mass
  - Loss of normal renal architecture
- CECT
  - Ill-defined, infiltrative, heterogeneously enhancing renal mass
  - Areas of necrosis and hemorrhage are common
  - Regional adenopathy and metastases common at presentation
    - Metastases: Lymph nodes, lung, liver, adrenal glands, pleura, omentum
  - Caliectasis without pelviectasis

MR Findings

- T1WI
  - Typically ill-defined low signal intensity
  - ± areas of high signal intensity due to hemorrhage
- T2WI
  - Typically low to intermediate signal intensity
  - Large areas of high signal intensity due to necrosis
  - Heterogeneity due to blood products of varying age
- DWI
  - Restricted diffusion in areas of viable tumor
- T1WI C+ FS
  - Typically hypovascular tumors with heterogeneous enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Ill-defined mass with heterogeneous echotexture
- Color Doppler
  - Hypovascular tumors; color flow typically not detected

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR

DIFFERENTIAL DIAGNOSIS

Transitional Cell Carcinoma (TCC)

- Similar imaging appearance as medullary carcinoma, but TCC occurs in older population

Lymphoma

- Less likely to present as infiltrative mass

Renal Cell Carcinoma (RCC)

- Typically well-defined, encapsulated mass

Pyelonephritis

- ± perinephric stranding; + signs/symptoms of infection

PATHOLOGY

General Features

- Etiology
  - Sickle cell trait → chronic medullary hypoxia
    - Hypoxia → transitional cell proliferation of terminal collecting ducts and papillary epithelium

Gross Pathologic & Surgical Features

- Central location in kidney; ± satellite lesions in cortex

Microscopic Features

- Sheets of poorly differentiated, eosinophilic cells
- Inflammatory, edematous stroma
- + mucin production

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Abdominal/flank pain, hematuria, weight loss

Demographics

- Age
  - Most common: < 25 years (typically between ~ 10-40 years)
- Gender
  - M:F = 2-3:1
    - No gender predilection in patients > 25 years old
- Ethnicity
  - Most common in African Americans with sickle cell trait

Natural History & Prognosis

- Poor prognosis: Mean survival ~ 15 weeks

SELECTED REFERENCES

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2. Srigley JR et al: The International Society of Urological Pathology (ISUP) Vancouver Classification of Renal Neoplasia. Am J Surg Pathol. 37(10):1469-89, 2013
3. Raman SP et al: Beyond renal cell carcinoma: rare and unusual renal masses. Abdom Imaging. 37(5):873-84, 2012
4. López Revuelta K et al: Kidney abnormalities in sickle cell disease. Nefrologia. 31(5):591-601, 2011
5. Prasad SR et al: Common and uncommon histologic subtypes of renal cell carcinoma: imaging spectrum with pathologic correlation. Radiographics. 26(6):1795-806; discussion 1806-10, 2006
6. Prasad SR et al: Neoplasms of the renal medulla: radiologic-pathologic correlation. Radiographics. 25(2):369-80, 2005

Collecting Duct Carcinoma

KEY FACTS

TERMINOLOGY

- Rare, aggressive neoplasm arising in renal medulla
- Synonym: Duct of Bellini carcinoma

IMAGING

- US
  - Ill-defined, hyperechoic mass
  - Obscuration of normal renal architecture
- CT
  - Infiltrative, solid mass centered in renal medulla
    - Invasion of renal sinus ~ 90-95%
  - Hypovascular to heterogeneous enhancement
  - Vascular invasion in ~ 25%
  - Preservation of renal contour > expansile appearance
- MR
  - T1WI: Hypo- to isointense relative to renal parenchyma
  - T2WI: Hypointense relative to renal parenchyma

TOP DIFFERENTIAL DIAGNOSES

- Transitional cell carcinoma (TCC)
- Renal cell carcinoma (RCC)
- Lymphoma
- Renal medullary carcinoma
- Pyelonephritis

PATHOLOGY

- Arises from collecting ducts in renal medulla
- Columnar cells with variable pleomorphism

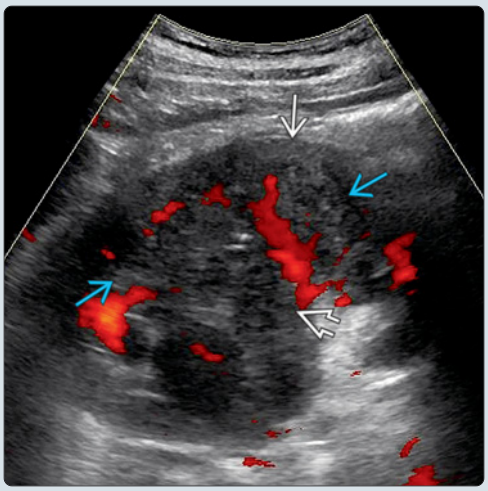
CLINICAL ISSUES

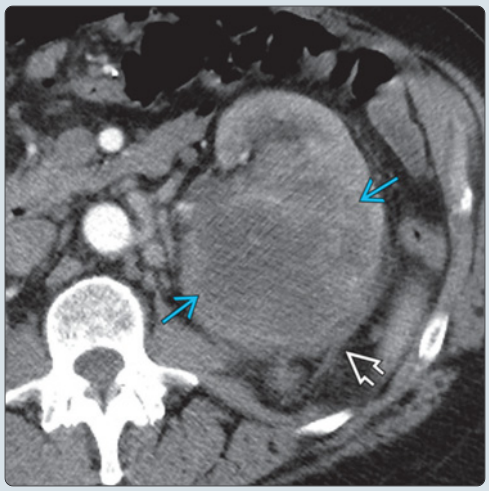
- ~ 40% of patients present with metastasis at diagnosis
- Poor prognosis: 2-year survival rate is only ~ 33%
- Presentation: Hematuria, flank pain, palpable mass

DIAGNOSTIC CHECKLIST

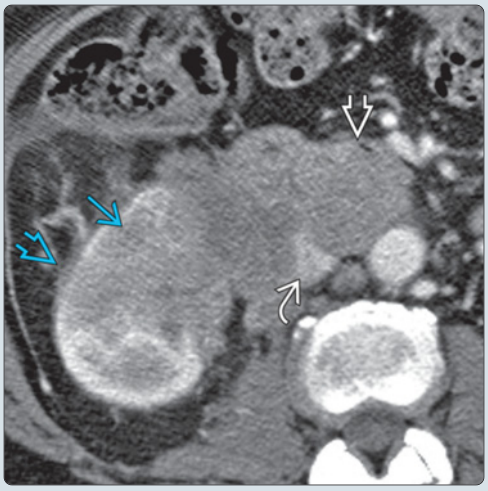
- Consider CDC in middle-aged patient with infiltrative renal mass centered in renal medulla

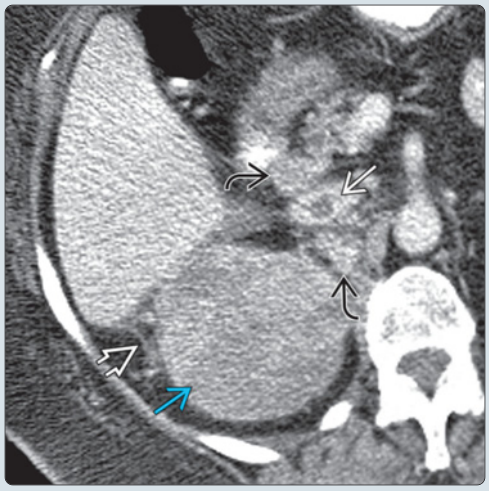
(Left) *Longitudinal power Doppler US demonstrates ill definition of the central kidney with obscuration of the normal anatomic landmarks due to an infiltrating mass, which invades the renal sinus, medulla, and cortex. Flow is evident throughout the abnormal soft tissue.* **(Right)** *Axial CECT in the same patient confirms an infiltrating, hypovascular mass. The mass originates in the medulla, but extends through the cortex into the perinephric fat. Pathology proved collecting duct carcinoma.*





(Left) *Axial CECT during the corticomedullary phase shows an ill-defined, hypovascular mass that infiltrates from the collecting system into the perinephric fat. Tumor also extends medially to surround and compress the IVC and fills the aortocaval space. Pathology revealed collecting duct carcinoma.* **(Right)** *Axial CECT shows a hypovascular mass replacing the entire superior pole of the right kidney. The mass infiltrates into the perinephric fat. Note metastatic pericaval adenopathy and tumor thrombus in the IVC.*





TERMINOLOGY

Abbreviations

- Collecting duct carcinoma (CDC)

Synonyms

- Duct of Bellini carcinoma; distal renal tubular carcinoma

Definitions

- Rare neoplasm arising from renal collecting ducts

IMAGING

General Features

- Best diagnostic clue
  - Large, ill-defined, infiltrative mass centered in renal medulla
- Location
  - Epicenter in renal medulla
- Size
  - Variable, but generally ~ 6-8 cm
- Morphology
  - Ill defined, infiltrative

CT Findings

- NECT
  - Hypo- to isoattenuating relative to renal parenchyma
  - Perinephric stranding: ~ 55%
  - Rare calcification
- CECT
  - Infiltrative, solid mass centered in renal medulla
    - Invasion of renal sinus: ~ 90-95%
  - Hypovascular to heterogeneous enhancement
  - Preservation of renal contour > expansile appearance
    - Expansile component due to ductal dilatation
  - Vascular invasion: ~ 25%
  - Cystic component: ~ 50%

MR Findings

- T1WI
  - Hypo- to isointense relative to renal parenchyma
- T2WI
  - Hypointense relative to renal parenchyma

Ultrasonographic Findings

- Hyperechoic, infiltrative soft tissue mass
- Obscuration of normal renal architecture

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR

DIFFERENTIAL DIAGNOSIS

Transitional Cell Carcinoma (TCC)

- Typically indistinguishable from CDC by imaging

Renal Cell Carcinoma (RCC)

- More commonly discrete mass

Medullary Carcinoma

- Associated with sickle cell trait

Lymphoma

- Commonly associated with systemic adenopathy

Pyelonephritis

- + signs/symptoms of infection; ± perinephric stranding

PATHOLOGY

General Features

- Associated abnormalities
  - Family history of other malignancies

Staging, Grading, & Classification

- CDC is adenocarcinoma with 2 parts
  - Papillary tumor with fibrovascular core
  - Glandular elements with desmoplastic reaction

Gross Pathologic & Surgical Features

- Centrally located, firm, grayish-white, infiltrating

Microscopic Features

- Arises from distal segment of collecting ducts of Bellini in renal medullary pyramid
- Columnar tumor cells with variable pleomorphism
  - Presence of hobnail cells highly suggestive of CDC

CLINICAL ISSUES

Presentation

- Hematuria, flank pain, palpable mass

Demographics

- Age
  - Average: 55 years
- Gender
  - M:F = 2:1

Natural History & Prognosis

- ~ 40% of patients present with metastatic disease
- Poor prognosis: ~ 33% survival at 2 years after diagnosis

DIAGNOSTIC CHECKLIST

Consider

- CDC may be indistinguishable from TCC at imaging
  - Pathologic distinction important for treatment
    - CDC → nephrectomy
    - TCC → nephroureterectomy

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Renal Transitional Cell Carcinoma

KEY FACTS

IMAGING

- Bladder: Site of 90% of all cases of TCC
  - Kidney (8%): Extrarenal part of renal pelvis more common than infundibulocalyceal
  - Ureter and proximal 2/3 of urethra (2%)
- Most are low-grade, superficial, papillary masses within renal collecting system
  - Sessile, flat, or polypoid solid mass
- ~ 15% are aggressive and invade renal parenchyma, renal sinus, etc.
  - Maintains bean-shaped renal contour
- Best clue: Irregular filling defect in renal pelvis with tumor infiltration of parenchyma
- CT urography
  - Axial NECT through abdomen and pelvis; CECT (nephrographic, 80-140 second delay) through kidneys
  - Excretory phase (5-10 minute delay) through abdomen and pelvis
  - Provides "IVP"-type image plus conventional CT views
- Ultrasound
  - Solid mass in collecting system that may efface renal sinus fat and invade into renal parenchyma
- MR
  - Iso- to slightly ↓ SI on T1W; iso- to mildly ↑ SI on T2WI

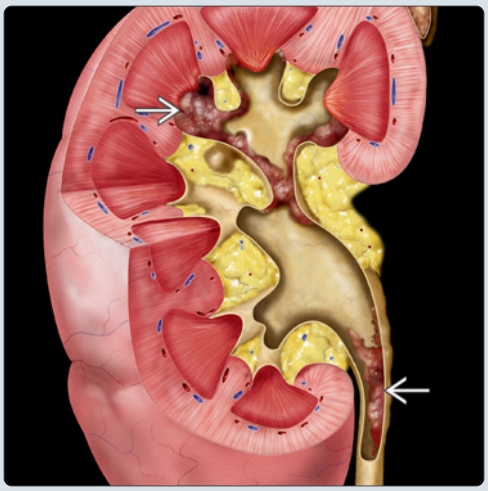
TOP DIFFERENTIAL DIAGNOSES

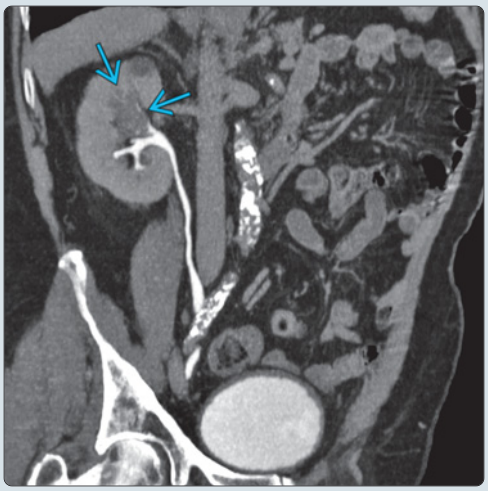
- Urolithiasis (renal calculi)
- Blood clot
- Infection (e.g., tuberculosis)
- Papillary necrosis
- Renal cell carcinoma
- Xanthogranulomatous pyelonephritis

DIAGNOSTIC CHECKLIST

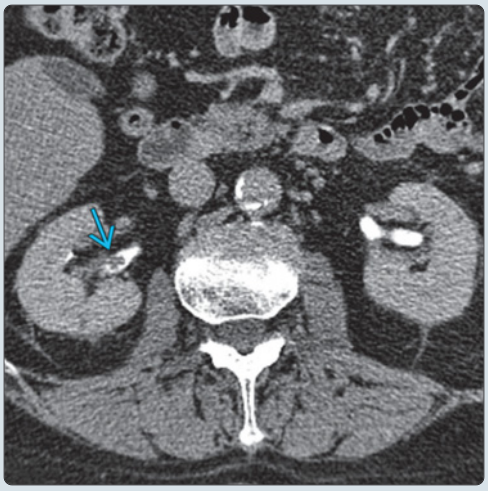
- Synchronous or metachronous TCC
- Cystoscopy still necessary to diagnose bladder cancer

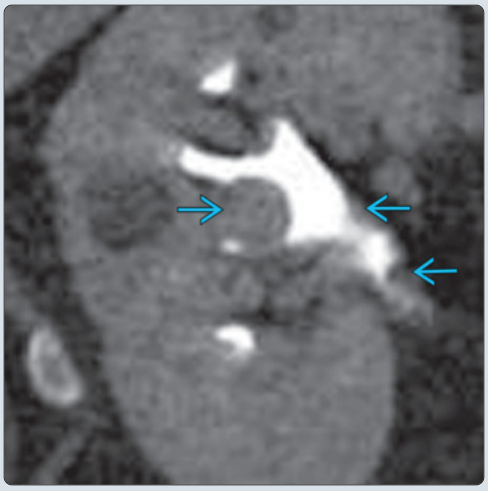
(Left) *Graphic shows multifocal transitional cell carcinoma ➡ involving the proximal ureter and superior pole calyces. Partial obstruction of the upper pole infundibulum can result in hydronephrosis of a superior calyx with filling defects ("oncocalyces").* **(Right)** *Coronal CECT during the excretory phase shows a soft tissue mass ⇨ filling the superior pole collecting system due to transitional cell carcinoma.*





(Left) *Axial CECT during the excretory phase in the same patient shows the transitional cell carcinoma as a filling defect ⇨ in the right renal collecting system.* **(Right)** *Coronal CECT in a different patient shows papillary projections of the tumor ⇨ at multiple sites along the renal pelvis and proximal ureter. There was no infiltration of the renal parenchyma (best evaluated on nephrographic phase images, not shown).*





TERMINOLOGY

Abbreviations

- Transitional cell carcinoma (TCC)

IMAGING

General Features

- Best diagnostic clue
  - Irregular filling defect in renal pelvis with tumor infiltration of parenchyma
- Location
  - Bladder: Site of 90% of all cases of TCC
  - Kidney (8%): Extrarenal part of renal pelvis more common than infundibula & calyces
  - Ureter & proximal 2/3 of urethra (2%)
- Morphology
  - Most are low-grade, superficial, papillary masses within renal collecting system
    - ~ 15% are aggressive and invade renal sinus and parenchyma

Radiographic Findings

- IVP
  - Nonfunctioning parenchymal segments due to tumor invasion or obstructed drainage
    - Global nonfunction: Obstructed ureteropelvic junction (UPJ) or ureter
  - Single or multiple discrete filling defects; surface is irregular, stippled, serrated, or frond-like
  - Mass may be sessile with minimal or no intraluminal intrusion (nonpapillary) or have pedicle
  - Renal pelvis
    - "Oncocalyx": Dilated tumor-filled calyx
    - "Phantom calyx": Unopacified calyx from obstruction of infundibulum
    - Stipple sign: Contrast within interstices of tumor
  - Ureter
    - Normal or delayed excretion (partial obstruction)
    - Hydronephrosis ± hydroureter
    - Fixation of ureter with irregular narrowing of lumen and nontapering margins
- Retrograde pyelography
  - Renal pelvis
    - Pyelotumoral backflow: Contrast in interstices
    - Opacification of phantom calyces; irregular papillary or nodular mucosa
  - Ureter
    - Goblet or champagne glass sign: Cup-shaped contrast collection distal to intraluminal filling defect of ureter (intraluminal growth of tumor)
    - Ovoid filling defect from wall: Short stalk
    - Smooth or frond-like surface; brush border or stippled appearance

CT Findings

- NECT
  - Isodense to slightly hyperdense (~ 30-50 HU) mass relative to urine & kidney
- CECT: Renal pelvis
  - Sessile or polypoid solid mass
  - Rare calcification (2% of renal pelvis TCC)
  - Focal renal pelvic wall thickening
  - Crust-like rims: Contrast in curvilinear calyceal spaces around periphery of tumor
  - Coarse striations or diminished nephrogram (partial obstruction)
  - Oncocalyx: Dilated, tumor-filled calyx
  - ± hydronephrosis
- Aggressive type: Compression or invasion of renal sinus fat and parenchyma
  - Maintains normal renal contour
  - Variable enhancement: Based on growth pattern, vascularity, and obstruction
  - ± metastases (nodes, retroperitoneum, lungs, bones)
- Ureter
  - Ureter may be dilated upstream from tumor
  - Intraluminal soft tissue mass (30-60 HU)
  - ± periureteral extension; ± enhancement
  - Eccentric or circumferential wall thickening

MR Findings

- T1WI
  - Tumor is iso- or hypointense to renal parenchyma
    - ↑ SI on T1WI due to intratumoral hemorrhage rare in TCC
- T2WI
  - Iso- to slight ↑ SI relative to renal parenchyma
  - Generally, lack cystic and necrotic components
- DWI
  - Typically restricted diffusion
  - ADC values generally lower than renal cell carcinoma
- T1WI C+
  - Typically mild, heterogeneous enhancement resulting in hypoattenuation of tumor relative to adjacent renal parenchyma
    - TCC enhances less than clear cell renal cell carcinoma
- MR urography
  - ≥ 1 filling defects in renal collecting system or ureter

Ultrasonographic Findings

- Grayscale ultrasound
  - Solid, heterogeneous mass with variable echogenicity
  - Mass may distort renal sinus fat and invade renal parenchyma
- Color Doppler
  - Often hypovascular with minimal internal flow

Imaging Recommendations

- Best imaging tool: CT urography (CECT with excretory phase)
  - CT urography
    - Axial NECT followed by CECT (nephrographic, 80-140 second delay)
      - Excretory phase (10-12 minute delay) through abdomen and pelvis
      - Alternatively, split bolus technique allows single acquisition of post-contrast images by combining nephrographic and excretory phases

DIFFERENTIAL DIAGNOSIS

Urolithiasis (Renal Calculi)

- IVP: Ureteral filling defects; often opaque on radiography
- Retrograde pyelography: No goblet sign
- CT: Most calculi are hyperattenuating (> 200 HU); differentiate from tumors

Blood Clot

- IVP: "Stipple" sign simulates TCC
- CT: Denser than renal parenchyma (50-90 HU), no enhancement; evolve over time
- US: Often similar echogenicity compared to neoplasms; mobile and evolve over time

Infection (e.g., Tuberculosis)

- IVP: Amputated calyx
- CT: Heterogeneous parenchymal mass ± calcifications

Renal Papillary Necrosis

- Destruction of apex of pyramid → irregular cavitation and sinus formation between papilla and calyx
- Retrograde pyelography: Fills 1 or 2 irregular cavities
- CT: Ring or triangular calcification of sloughed papilla

Renal Cell Carcinoma

- Usually seen as exophytic, heterogeneous lesion
- May also cause infiltrative mass and extend into renal pelvis

Xanthogranulomatous Pyelonephritis

- Can also infiltrate renal parenchyma
- Usually associated with large obstructing calculus and enlarged, nonfunctioning kidney

PATHOLOGY

General Features

- Urothelial cancers
  - TCC: 90% of renal pelvic and 97% of ureteral cancers
  - Squamous cell carcinoma: 5-10% of renal pelvic and rare in ureteral cancers
  - Adenocarcinoma: Very rare in both
- Synchronous or metachronous (multicentricity)
  - Metachronous upper tract TCC (11-13%)
  - Metachronous ipsilateral upper tract TCC (14-30%)
  - Synchronous bilateral renal pelvic TCC (1-2%), ureteral (2-9%)
  - Synchronous bladder TCC and renal pelvic TCC (24%), ureteral (39%)
  - Upper tract and bladder TCC at some time (30-75%)
  - Upper tract TCC in 10%, 26%, and 34% at 5, 5-10, and > 10 years of follow-up of bladder TCC

Staging, Grading, & Classification

- T0: No tumor; Tis: Carcinoma in situ; Ta: Tumor confined to mucosa
- T1: Invades lamina propria
- T2a: Invades inner muscularis; T2b: Outer muscularis
- T3a: Microscopic perivesical fat invasion; T3b: Macroscopic perivesical fat invasion
- T4a: Invades adjacent organs; T4b: Invades pelvic or abdominal wall
- N1-3: Pelvic nodes; N4: Above renal bifurcation
- M1: Distant metastases

Gross Pathologic & Surgical Features

- ≥ 85% papillary (low grade); ± infiltrative (high)

Microscopic Features

- Transitional epithelium: In renal pelvis, ureter, bladder, and proximal 2/3 of urethra
- Epithelial atypia or dysplasia; abnormal fibrovascular core of lamina propria

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Gross hematuria (70-80%), dull or colicky pain (50%)
- Laboratory data
  - Urinalysis: Hematuria, ± pyuria and bacteriuria

Demographics

- Age
  - > 60 years of age
- Gender
  - M:F = 3:1
- Epidemiology
  - Risk factors: Smoking, chemicals, cyclophosphamide therapy, human papilloma virus, analgesic abuse

Natural History & Prognosis

- Renal pelvis and ureteral
  - ≤ T1: 5-year survival rate (77-80%)
  - T2: 5-year survival rate (44%)
  - ≥ T3: 5-year survival rate (0-25%)
- Bladder: Overall 5-year survival rate (30%)

Treatment

- Renal pelvis and ureteral
  - Total nephroureterectomy and bladder cuff excision
  - Metastases: Chemotherapy &/or radiation

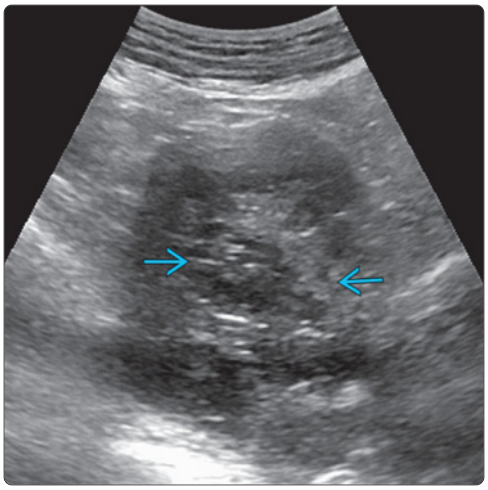
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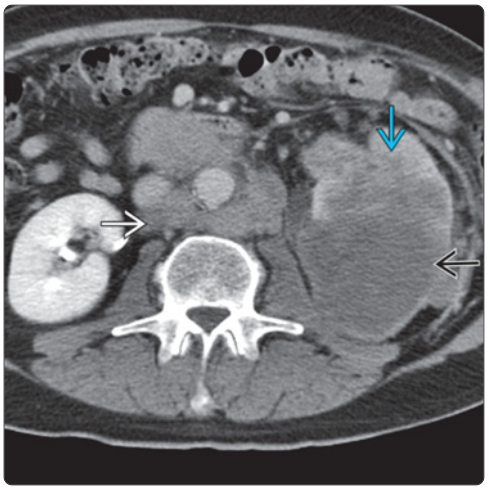
Consider

- Synchronous or metachronous TCC
- Lifetime surveillance essential to detect subsequent tumors

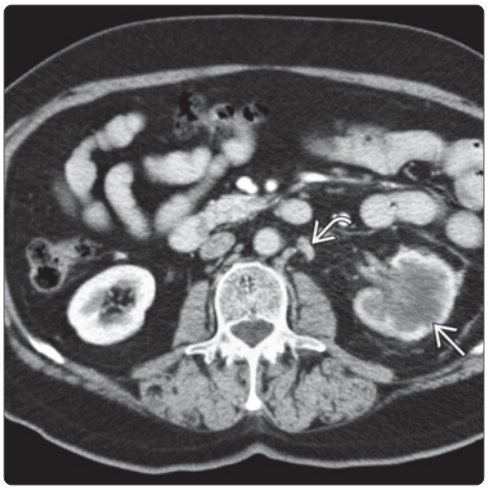
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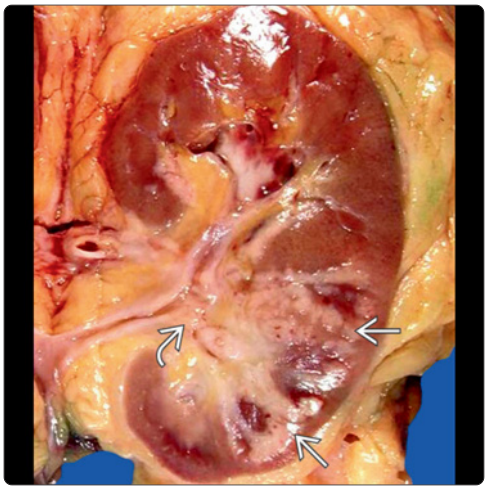
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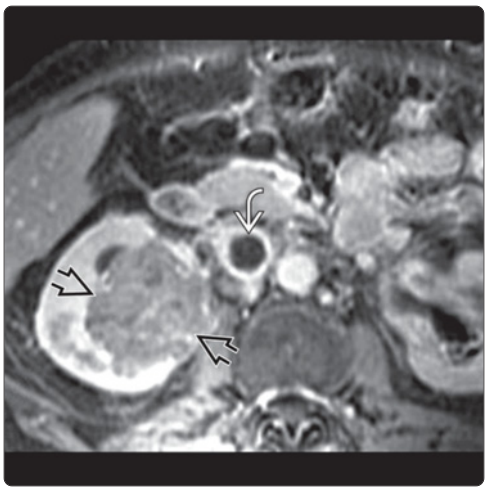


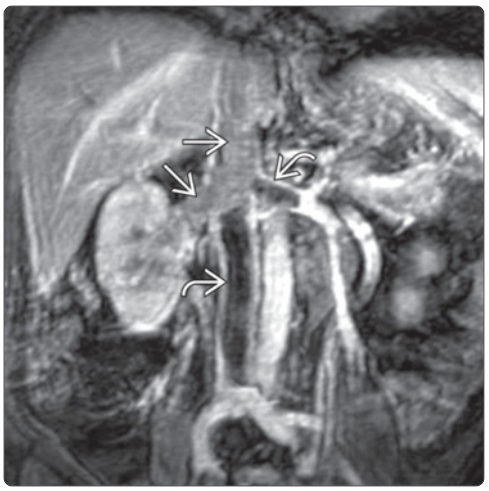
(Left) *Transverse ultrasound of the left kidney shows replacement of the normal sinus fat with an ill-defined, soft tissue mass due to transitional cell carcinoma.* **(Right)** *Axial CECT during the nephrographic phase in the same patient reveals a heterogeneously enhancing mass effacing the renal sinus and extending posteriorly into the renal parenchyma. Note metastatic retroperitoneal adenopathy.*





(Left) *Axial CECT shows infiltration of the renal parenchyma by an aggressive TCC from an inferior pole calyx that was "amputated" on pyelographic phase images. Renal contour is preserved. Nodal metastases were better shown on adjacent images.* **(Right)** *Gross pathology of the bivalved resected kidney (coronal plane) in the same patient shows a tumor along the surface of the inferior pole collecting system and invading into the parenchyma with preservation of the normal renal contour.*





(Left) *Axial T1 C+ FS MR shows a tumor arising from the renal pelvis and infiltrating the renal parenchyma, without altering the renal contour. Note the thrombus in the IVC.* **(Right)** *Coronal T1 C+ FS MR shows an enhancing tumor thrombus within the right renal vein and suprarenal IVC, along with diffuse infiltration of the kidney but preservation of its the renal contour. Thrombus is present in the infrarenal IVC and left renal vein.*

T | Primary Tumor

TNM	Definitions
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Ta	Papillary noninvasive carcinoma
Tis	Carcinoma in situ
T1	Tumor invades subepithelial connective tissue
T2	Tumor invades muscularis
T3	
Renal pelvis only	Tumor invades beyond muscularis into peripelvic fat or the renal parenchyma
Ureter only	Tumor invades beyond muscularis into periureteric fat
T4	Tumor invades adjacent organs or through kidney into perinephric fat

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in single lymph node, ≤ 2 cm in greatest dimension
N2	Metastasis in single lymph node, > 2 cm but ≤ 5 cm in greatest dimension; or multiple lymph nodes, none > 5 cm in greatest dimension
N3	Metastasis in lymph node, > 5 cm in greatest dimension

Laterality does not affect the N classification.

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis

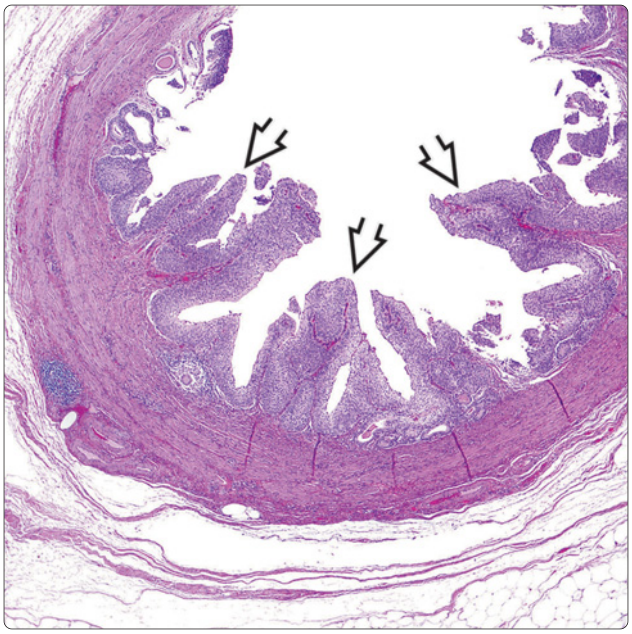
Adapted from 7th edition AJCC Staging Forms (2010).

AJCC | Stages/Prognostic Groups

Stage	T	N	M
0a	Ta	N0	M0
0is	Tis	N0	M0
I	T1	N0	M0
II	T2	N0	M0
III	T3	N0	M0
IV	T4	N0	M0
	Any T	N1	M0
	Any T	N2	M0
	Any T	N3	M0
	Any T	Any N	M1

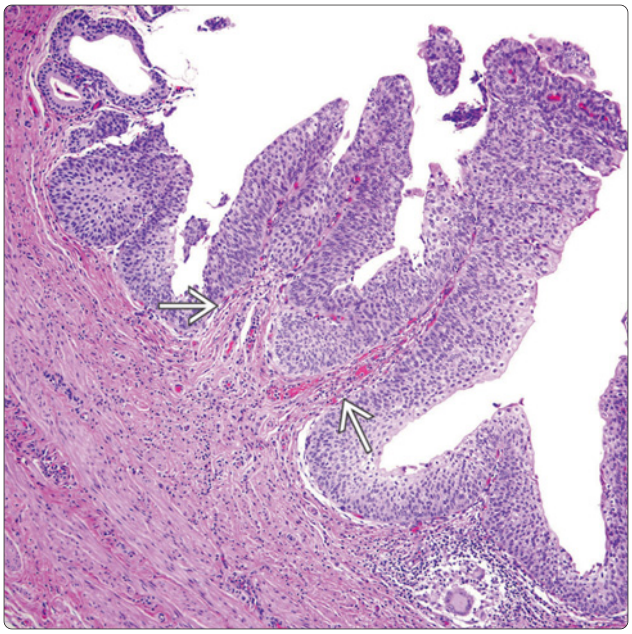
Adapted from 7th edition AJCC Staging Forms (2010).

Ta



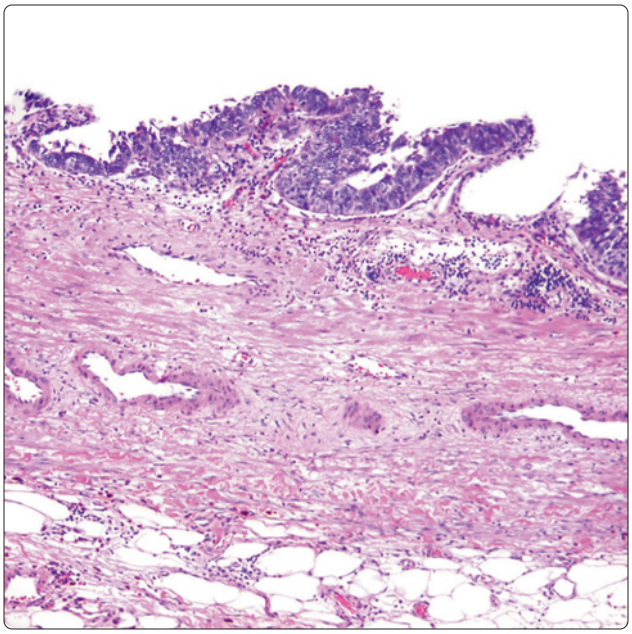
Low-power magnification (H&E stain) shows a cross section of proximal ureter with papillary noninvasive urothelial carcinoma. Note the multiple papillary fronds ⇨ lined by variably thickened epithelium extending into the lemon.

Ta



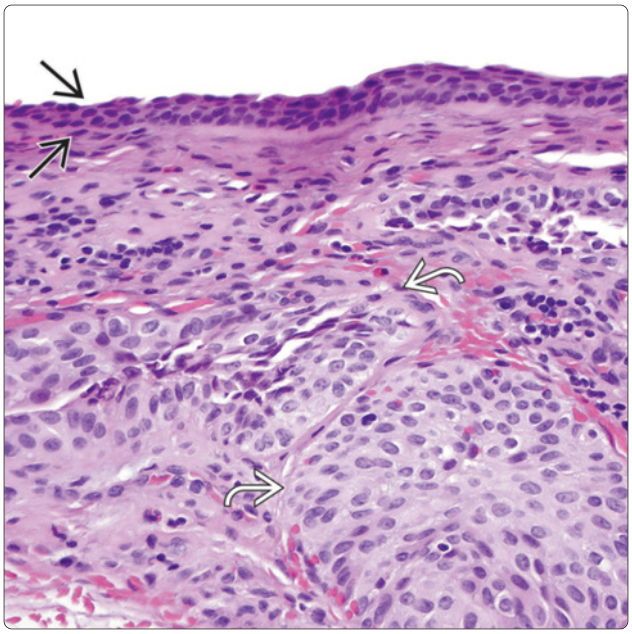
Higher power magnification of the previous image shows fibrovascular cores ➡ surrounded by an increased number of cells compared to normal urothelium. Cells are oriented perpendicular to basement membrane and lack cytologic atypia and pleomorphism.

Tis



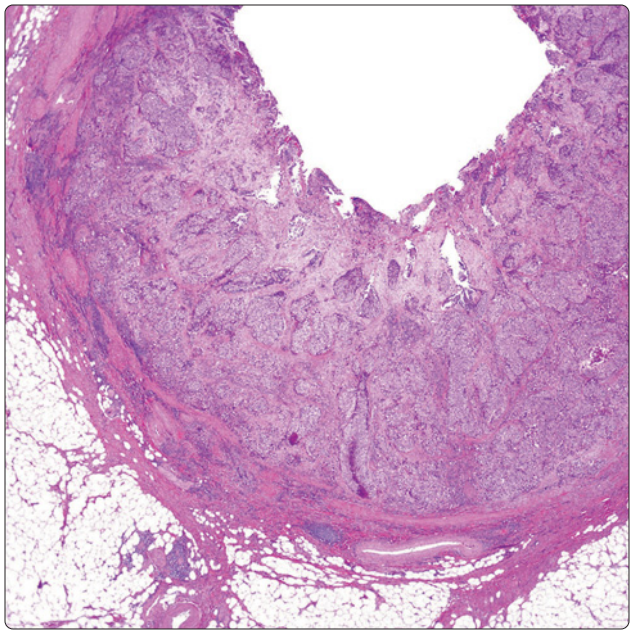
H&E shows that, in contrast to cells in the previous images, the neoplastic cells are markedly pleomorphic and hyperchromatic. The nuclei lack polarity to basement membrane. Note also the propensity of these cells to become detached from basement membrane focally.

T1



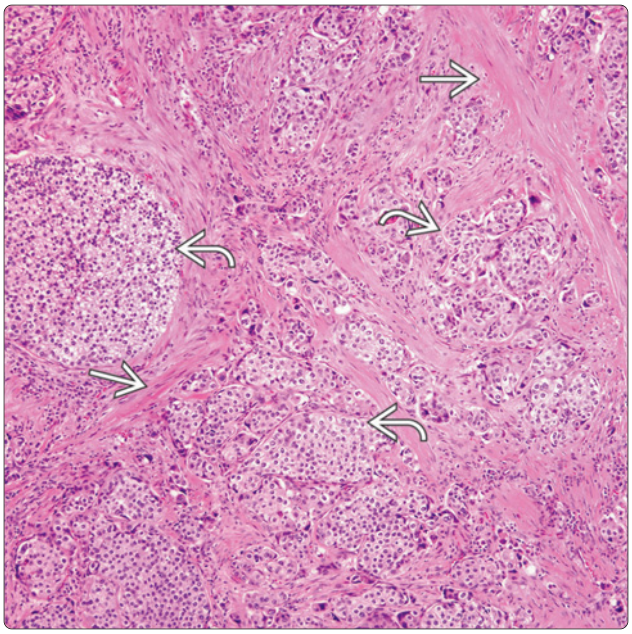
H&E stain demonstrates uninvolved superficial urothelium ⇨ with underlying nests of neoplastic urothelium ➡ that invade the subepithelial connective tissue.

T2



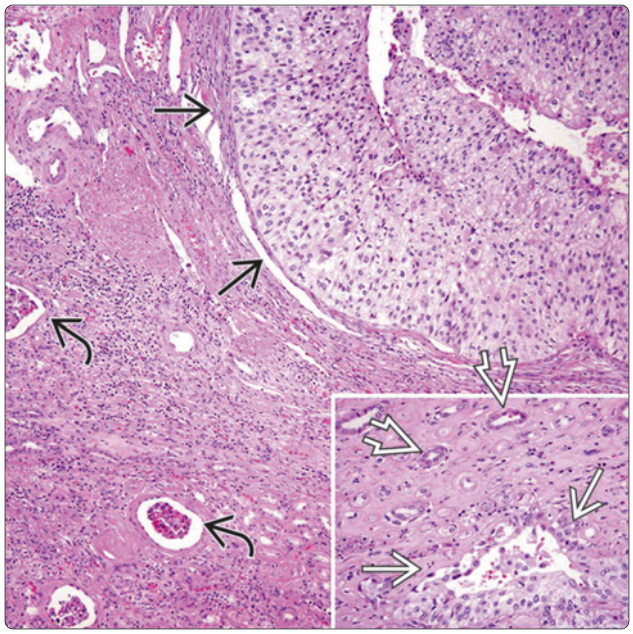
H&E stain shows a cross section of a ureter with invasive urothelial carcinoma. Tumor nests invade almost the entire thickness of the muscularis propria but do not extend into the periureteric fat.

T2



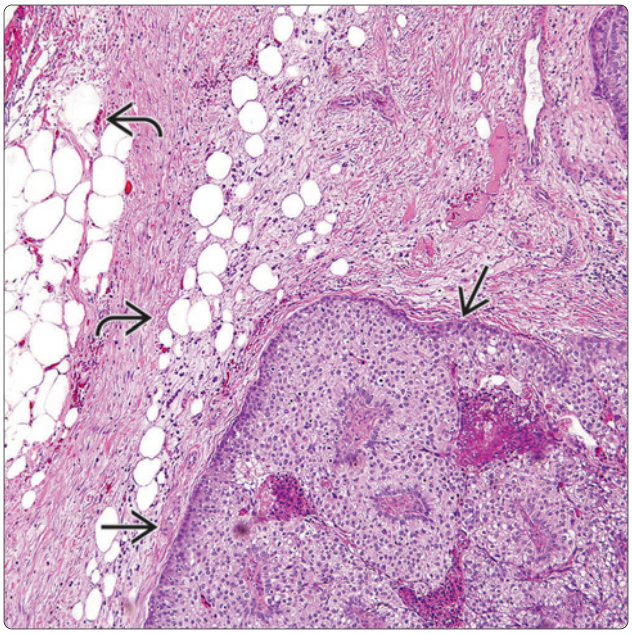
High-power magnification of the previous image shows nests of urothelial carcinoma ⇨ invading and disrupting the muscularis propria (pink bundles of smooth muscles) ⇨.

T3



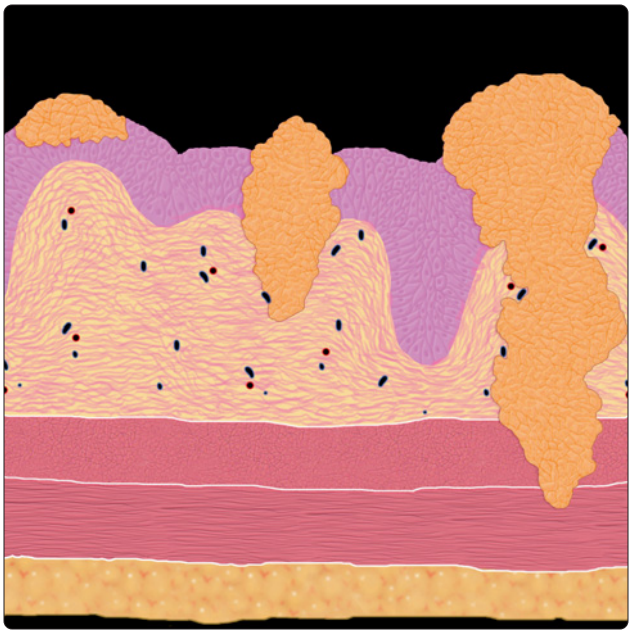
H&E stain shows a nest of urothelial carcinoma ⇨ invading into renal parenchyma; note the renal glomeruli ⇨. The inset shows a nest of urothelial carcinoma ⇨ invading renal parenchyma in proximity to renal tubules ⇨.

T4



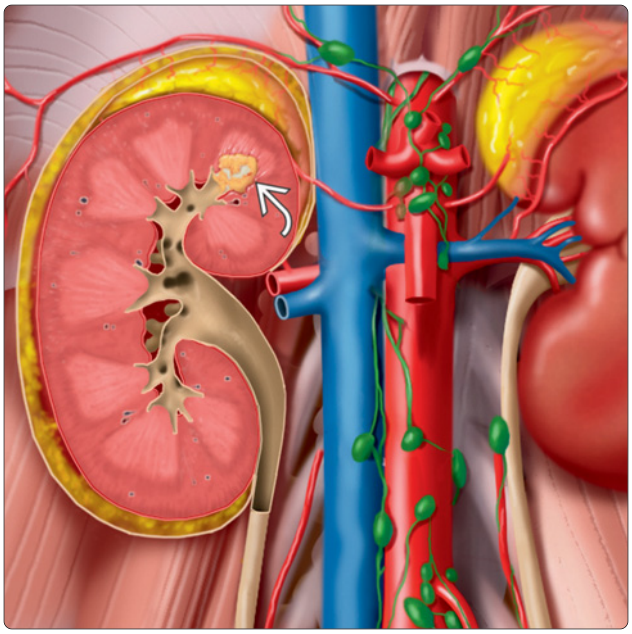
Urothelial carcinoma invades through the kidney to perinephric fat. The large nests of urothelial carcinoma ⇨ infiltrate the perinephric fat (white round cells) ⇨ with prominent pink desmoplastic reaction.

Ta, T1, and T2



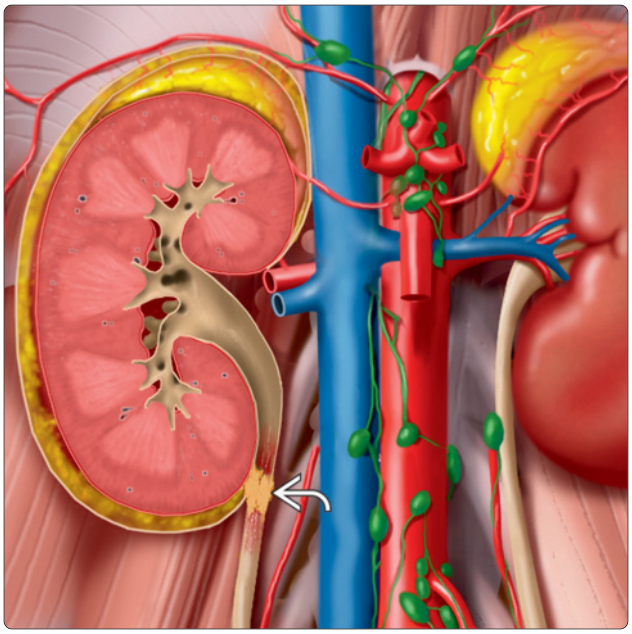
Graphic shows local extent of early stages of ureteric carcinoma: Ta is papillary noninvasive carcinoma, T1 is tumor invading subepithelial connective tissue, and T2 is tumor invading the muscularis.

T3: Renal Pelvis and Calyceal Carcinoma



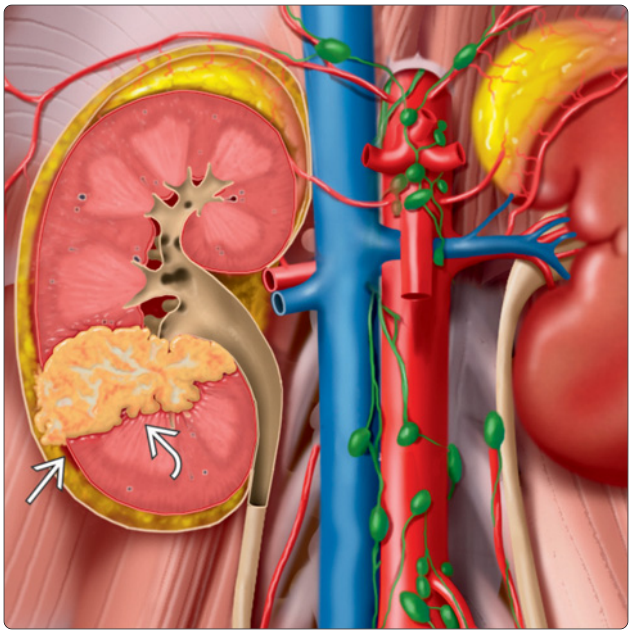
Graphic shows T3 tumor of the upper calyx with extension through the muscularis layer into the peripelvic fat or renal parenchyma.

T3: Ureteral Carcinoma



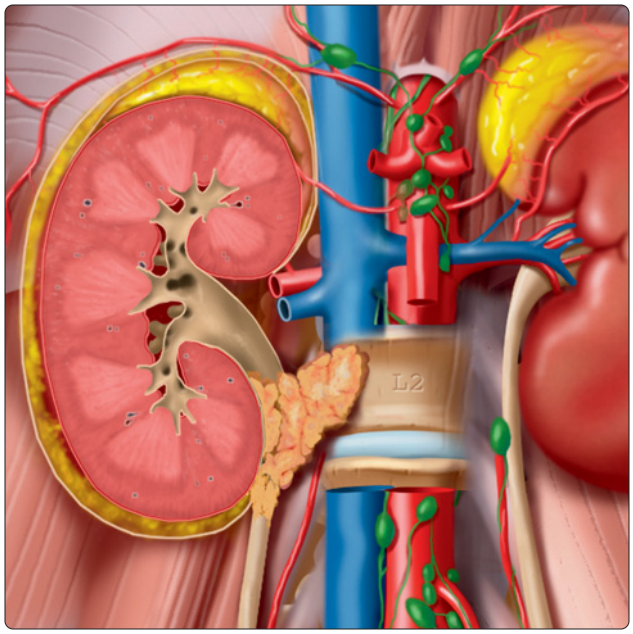
Graphic shows T3 tumor of the ureter with extension through the muscularis layer into the periureteric fat without invasion of surrounding organs or structures.

T4: Renal Pelvis and Calyceal Carcinoma



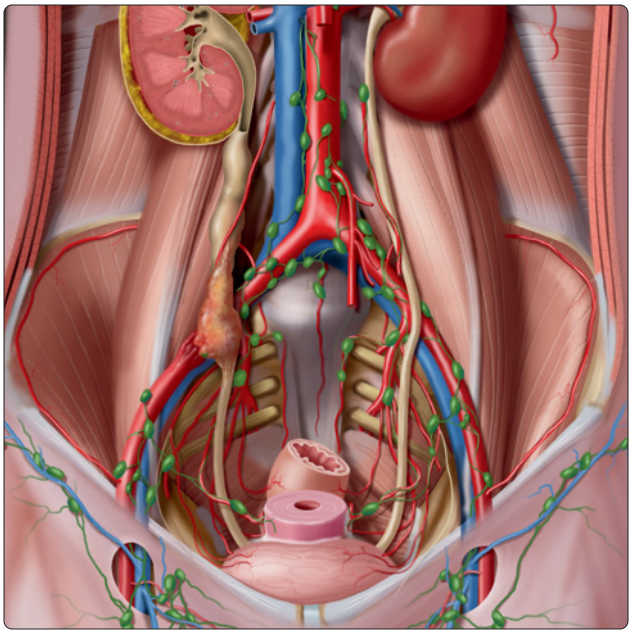
Graphic shows T4 tumor of the lower calyx with invasion through the renal parenchyma into the perinephric fat.

T4: Ureteral Carcinoma



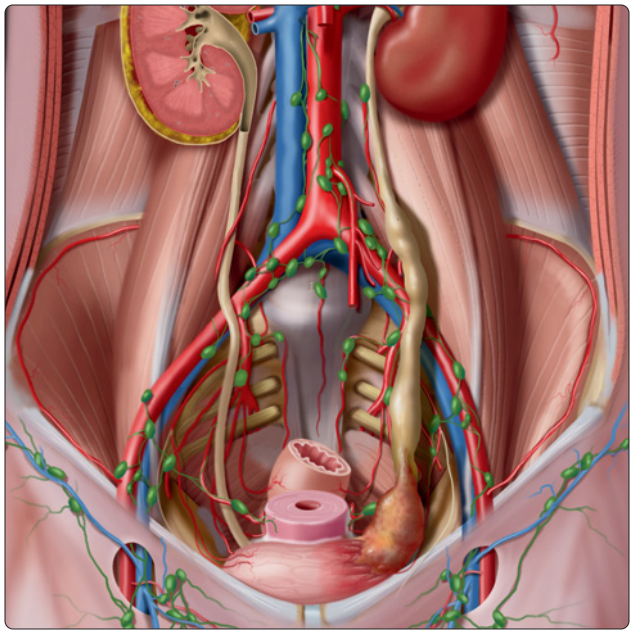
T4 ureteric tumors invade through the surrounding organs and structures. Graphic shows a T4 tumor of the upper ureter invading L2 vertebral body.

T4: Ureteral Carcinoma



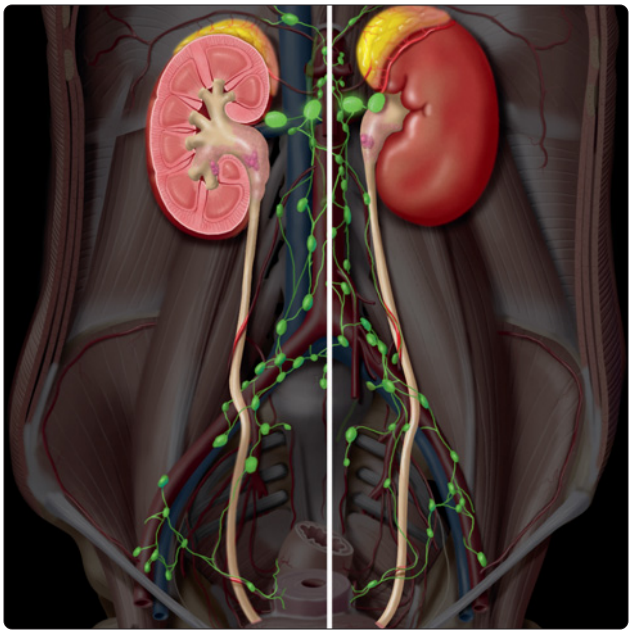
T4 ureteric tumors invade through the surrounding organs and structures. Graphic shows a T4 tumor of the middle ureter invading the right iliac vessels.

T4: Ureteral Carcinoma



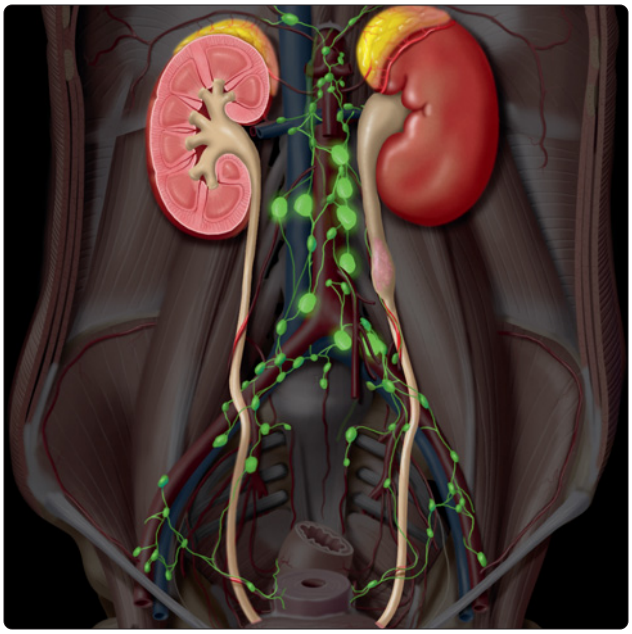
T4 ureteric tumors invade through the surrounding organs and structures. Graphic shows a T4 tumor of the lower ureter invading the urinary bladder.

Nodal Drainage of Renal Pelvis and Calyces



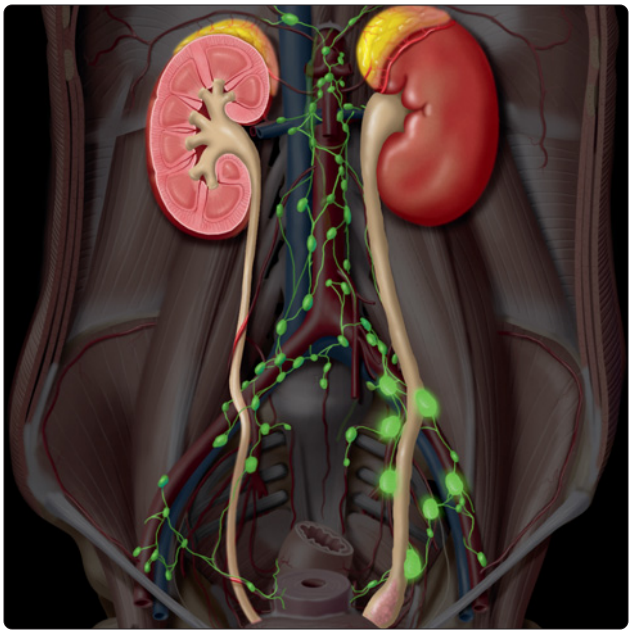
Right renal pelvis tumors spread to renal hilar, paracaval, and retrocaval nodes, whereas left renal pelvic tumors spread to renal hilar and paraaortic nodes.

Nodal Drainage of Upper 2/3 of Ureters



Tumors of the upper 2/3 of the right ureter spread to retrocaval and aortocaval nodes, and tumors of the upper 2/3 of the left ureter spread to paraaortic nodes, nodes at the origin of the inferior mesenteric artery, and common iliac nodes.

Nodal Drainage of Lower 1/3 of Ureters



Tumors of the distal 1/3 of either ureter spread to common iliac, internal iliac, external iliac, obturator, and presacral nodes.

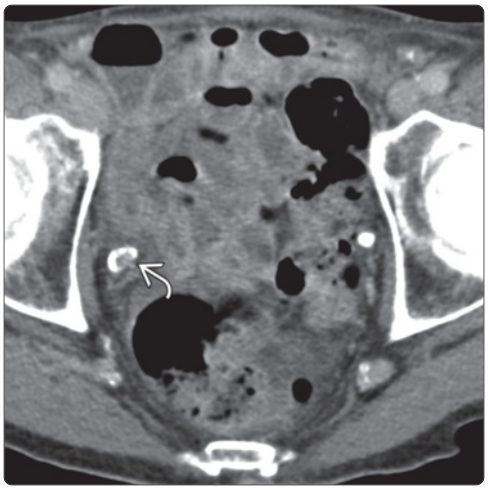
Metastases, Organ Frequency



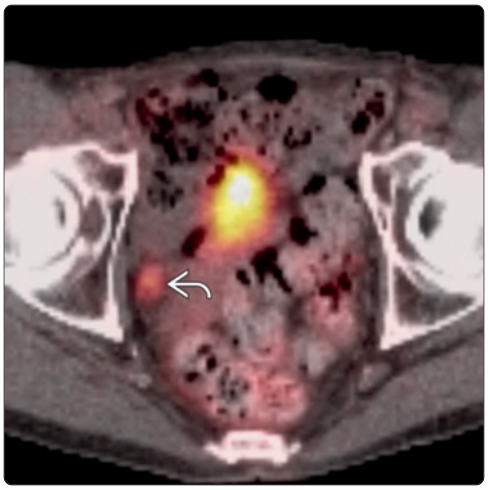
Liver	*63%*
Lung	*58%*
Bone	*43%*
Gastrointestinal tract	*20%*
Peritoneum	*18%*
Adrenal gland	*15%*
Ovary	*13%*
Uterus	*10%*

Data from Batata MA et al: Primary carcinoma of the ureter: a prognostic study. Cancer. 35:1626-32, 1975.

Stage 0a (Ta N0 M0)

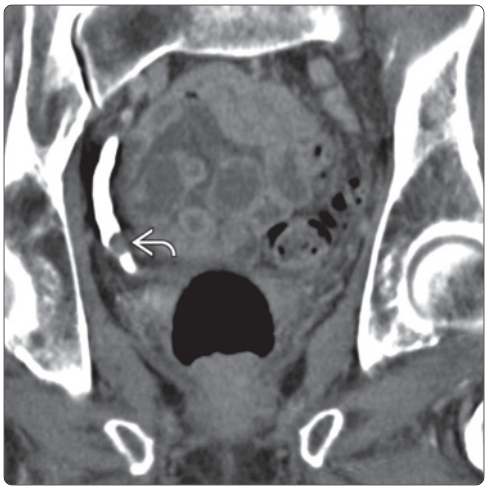


Stage 0a (Ta N0 M0)

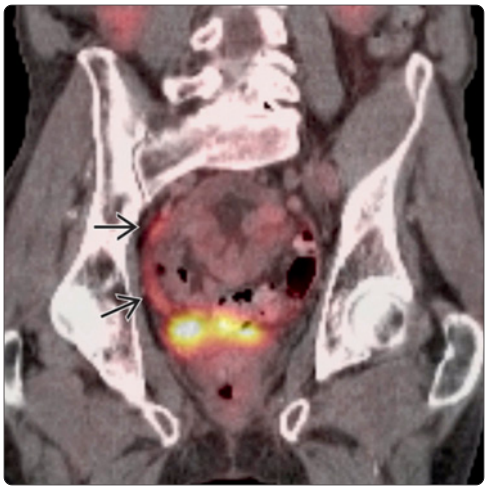


(Left) *Axial CT IVP in a patient with a history of lymphoma and hematuria shows a polypoid filling defect within the distal right ureter. At surgery, this proved to be a noninvasive carcinoma.* **(Right)** *Axial PET/CT in the same patient performed 2 days later for staging of lymphoma shows increased metabolic activity in the distal right ureter due to renal excretion. It is very difficult to detect urothelial tumor on PET/CT due to urinary excretion of FDG.*

Stage 0a (Ta N0 M0)

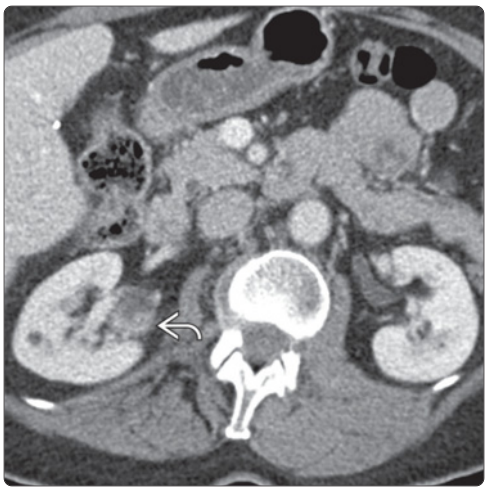


Stage 0a (Ta N0 M0)

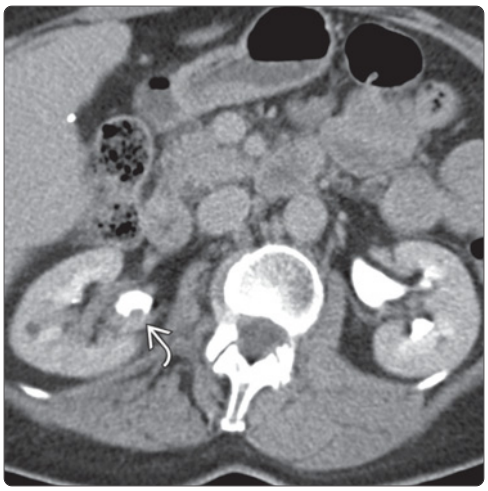


(Left) *Coronal CT IVP in the same patient shows a well-defined filling defect within the distal right ureter.* **(Right)** *Coronal PET/CT in the same patient shows a long segment of increased activity within the distal right ureter. Urinary excretion of FDG masks the increased activity resulting from urothelial tumor. If a lesion is suspected, forced diuresis will help wash out excreted FDG from the ureter.*

Stage 0a (Ta N0 M0)

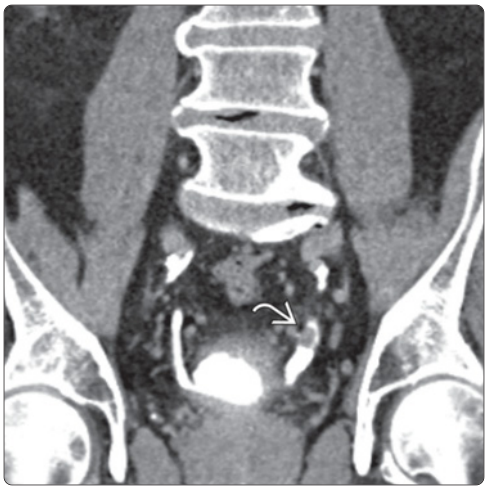


Stage 0a (Ta N0 M0)

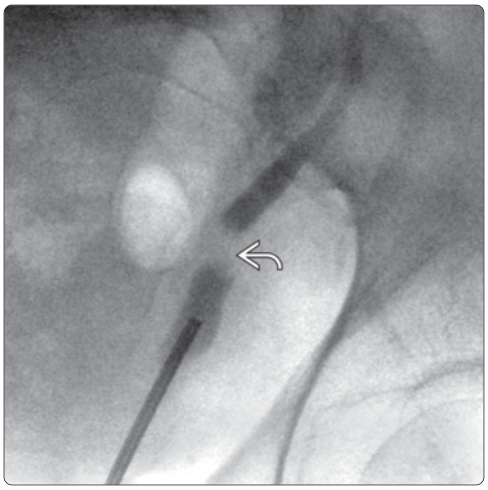


(Left) *Axial CECT in another patient obtained during the nephrographic phase of contrast enhancement shows a small enhancing polypoid lesion of the posterior wall of the right renal pelvis.* **(Right)** *Axial CECT in the same patient obtained during the pyelographic phase of contrast enhancement shows the right renal pelvis polypoid lesion as a filling defect against the contrast-filled renal pelvis. This was found to be papillary noninvasive carcinoma.*

Stage I (T1 N0 M0)

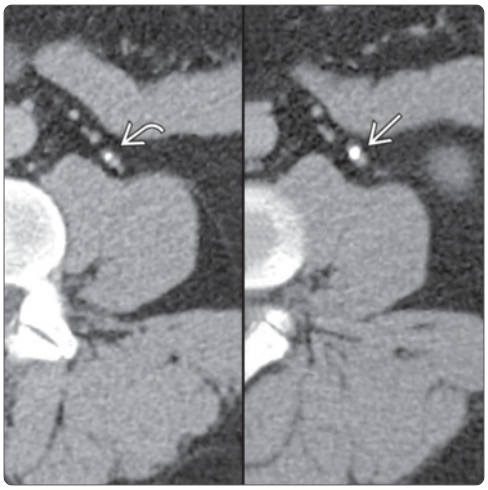


Stage I (T1 N0 M0)

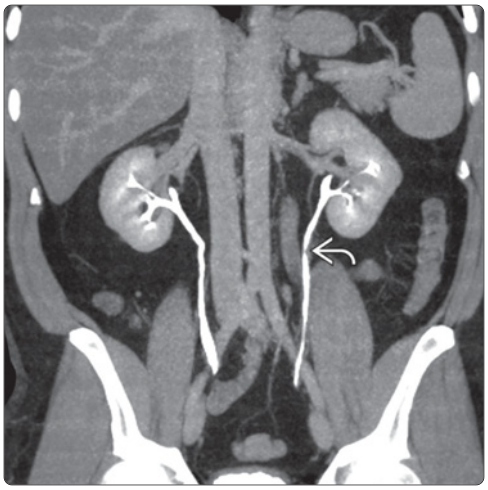


(Left) *Coronal CT IVP shows an intraluminal mass in the distal part of the left ureter. There is no evidence of periureteric invasion. CT is not helpful in differentiating early stage ureteric tumors.* **(Right)** *Frontal view of a retrograde pyelography in the same patient shows the mass almost obstructing the left ureter.*

Stage I (T1 N0 M0)

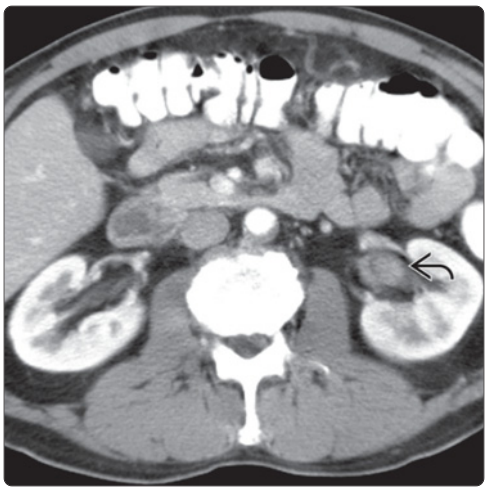


Stage I (T1 N0 M0)

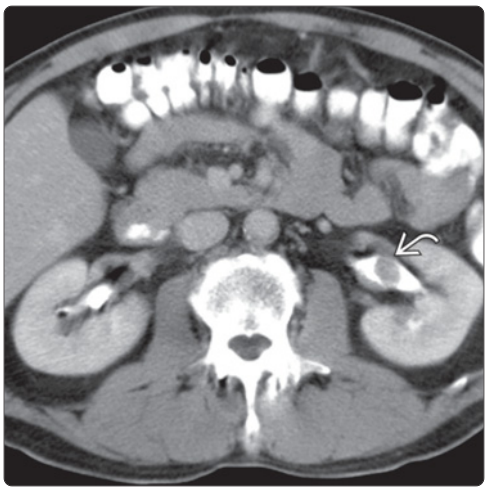


(Left) *Axial CT IVP at 2 levels shows subtle narrowing of the ureter with normal caliber above that level.* **(Right)** *Coronal CT IVP MIP reformatted image in the same patient shows a subtle eccentric filling defect at the junction between the upper and middle 1/3 of the left ureter. This was found to be a TCC that invaded into the subepithelial connective tissue. The use of coronal reformats and wide window width allow detection of subtle ureteric lesions.*

Stage I (T1 N0 M0)



Stage I (T1 N0 M0)

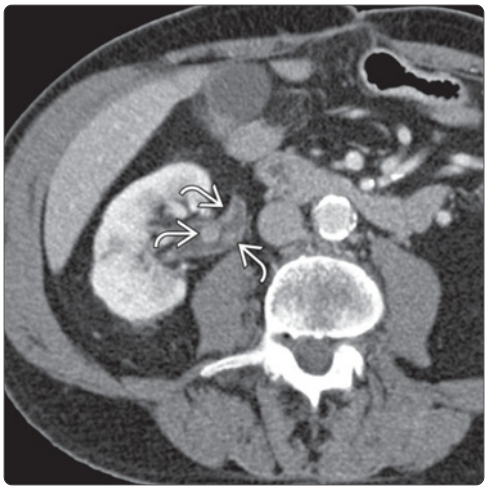


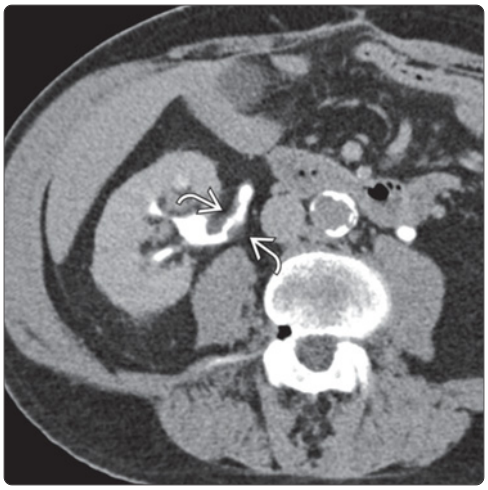
(Left) *Axial CT IVP obtained during the nephrographic phase of contrast enhancement shows an enhancing mass slightly distending the left renal pelvis. Ureteric or renal pelvis distension provides a clue to the presence of a mass, even on a nonenhanced CT.* **(Right)** *Axial CT IVP in the same patient obtained during the pyelographic phase of contrast enhancement clearly shows a filling defect within the left renal pelvis.*

Stage I (T1 N0 M0)

Stage I (T1 N0 M0)

(Left) *Axial CECT during the nephrographic phase of contrast enhancement shows multiple enhancing polypoid lesions within the right renal pelvis.* **(Right)** *Axial CECT in the same patient during the pyelographic phase of contrast enhancement shows multiple filling defects in the contrast-filled renal pelvis. Unilateral multicentricity does not affect staging of urothelial carcinoma.*

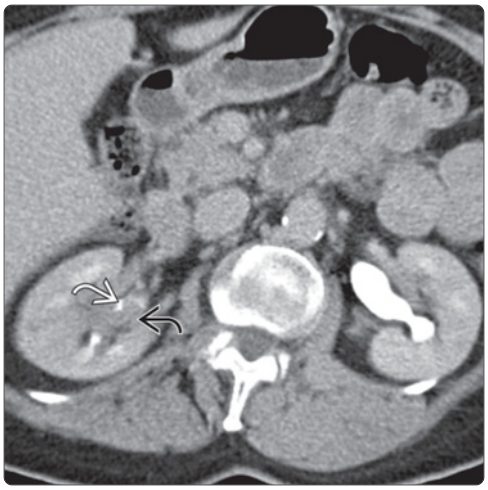




Stage II (T2 N0 M0)

Stage II (T2 N0 M0)

(Left) *Axial CECT shows an enhancing lesion expanding the right renal pelvis. Note also focal calcifications.* **(Right)** *Frontal retrograde pyelogram in the same patient shows a filling defect within the renal pelvis and extending into the superior calyx. The mass was found at surgery to invade the muscularis without invading the peripelvic fat, representing T2 tumor.*

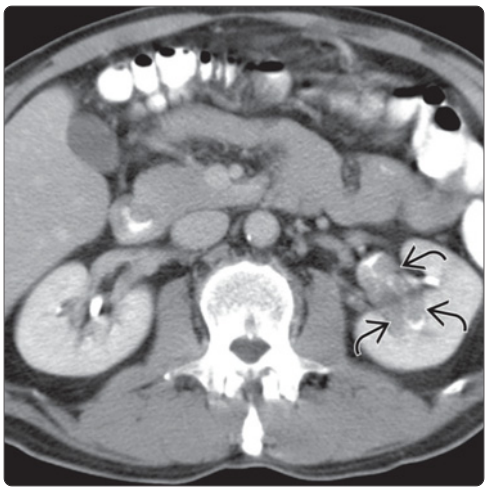


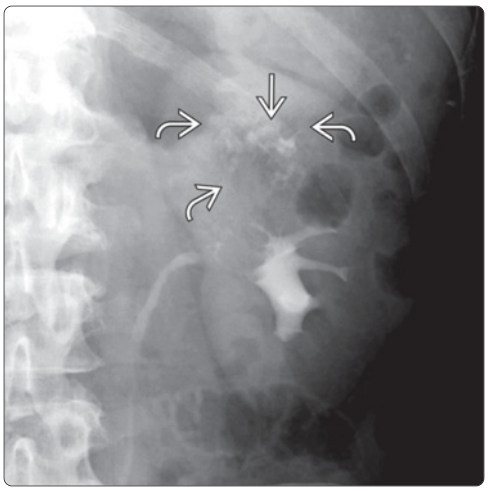


Stage II (T2 N0 M0)

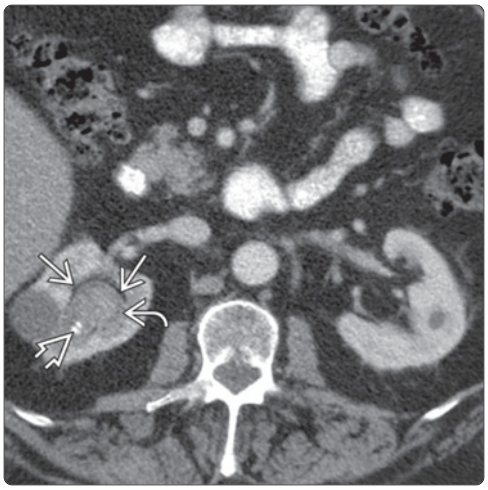
Stage II (T2 N0 M0)

(Left) *Axial CT IVP shows a mass filling the left renal pelvis and upper calyces without involvement of the renal sinus fat or renal parenchyma.* **(Right)** *Frontal excretory urography in the same patient shows a large, stippled filling defect involving the renal pelvis and upper calyces of the left kidney. The stipple sign refers to tracking of contrast material into the interstices of a papillary lesion.*

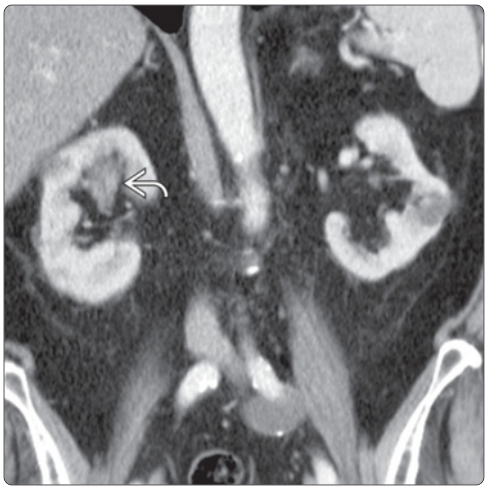




Stage II (T2 N0 M0)

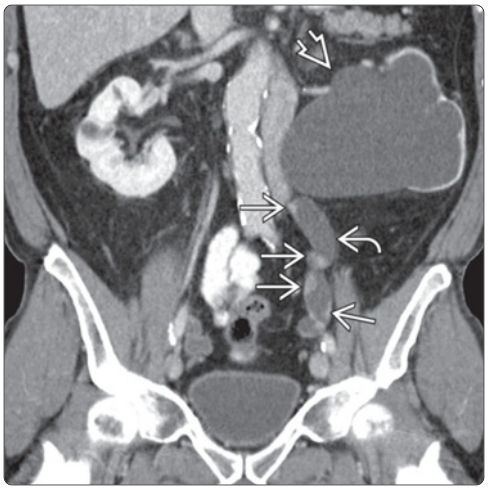


Stage II (T2 N0 M0)

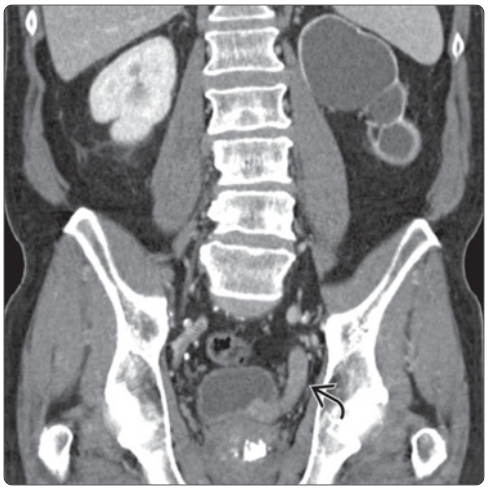


(Left) *Axial CT IVP shows a mass filling and distending the right upper calyx. Note the compressed renal sinus fat around the distended calyx. Calcifications in the mass are a common feature of TCC, which allow detection of tumors on a nonenhanced CT performed for evaluation of hematuria.* **(Right)** *Coronal CT IVP in the same patient shows a mass filling the upper calyx (oncocalyx) without involvement of the renal sinus fat or renal parenchyma.*

Stage II (T2 N0 M0)



Stage II (T2 N0 M0)

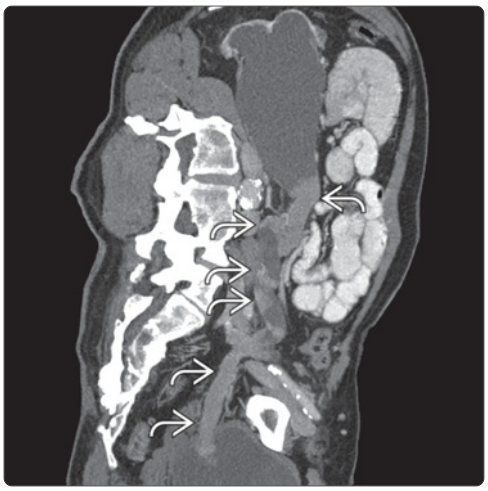


(Left) *Coronal CECT shows severe hydronephrosis and hydroureter. Multiple ureteric masses are seen within the ureter. It is difficult to show the extent of ureteric involvement on a single axial or coronal image. No contrast was seen in the renal pelvis or ureter, even an hour after contrast injection.* **(Right)** *Coronal CECT in the same patient shows that the distal ureter is filled with a large mass extending to the ureterovesical junction.*

Stage II (T2 N0 M0)

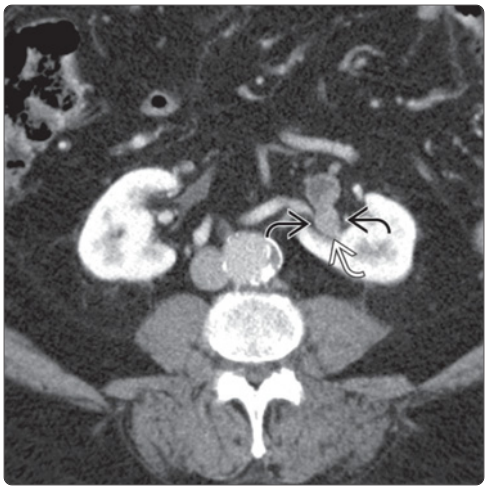


Stage II (T2 N0 M0)



(Left) *Axial CECT in the same patient shows the distal ureteric mass protruding into the urinary bladder lumen.* **(Right)** *Curved multiplanar reformatting along the course of the ureter shows the extent of ureteric involvement with multiple masses. The ureteric masses were all confined to the ureter without extraureteric extension. This case illustrates a common pathway of tumor spread through seeding along the ureter.*

Stage II (T2 N0 M0)

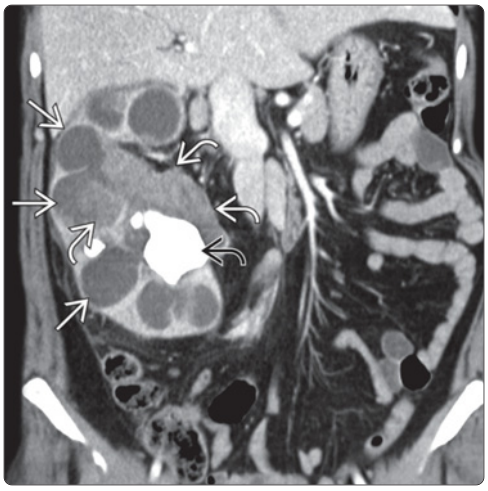


Stage II (T2 N0 M0)

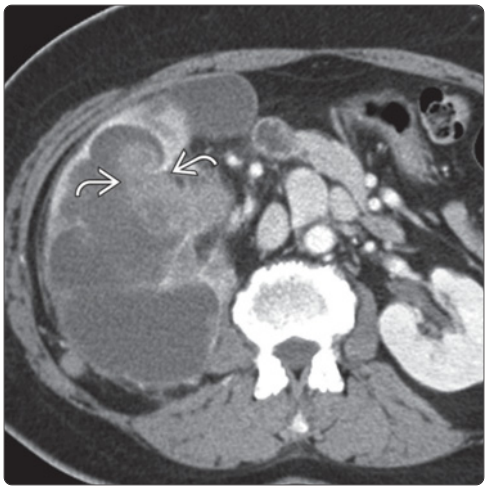


(Left) *Axial CECT in a patient with horseshoe kidney shows a mass filling left posterior calyx without renal invasion. Note the clear fat plane around the distended calyx.* **(Right)** *Axial CECT in the same patient shows extension of the tumor to the renal pelvis, again with preservation of peripelvic fat. The relative risk of TCC in a patient with horseshoe kidney is increased 3-4x.*

Stage II (T2 N0 M0)

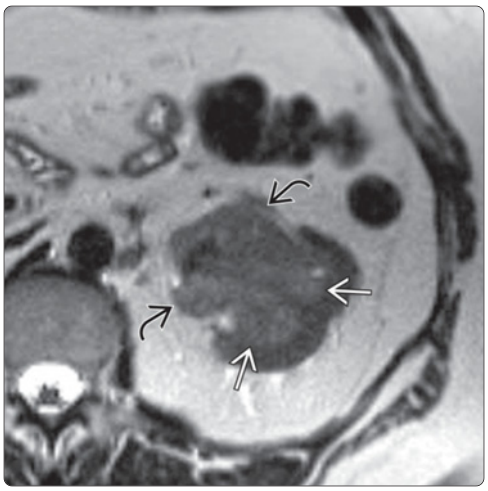


Stage II (T2 N0 M0)



(Left) *Coronal CECT shows an enlarged kidney with a staghorn stone in the renal pelvis and replacement of renal parenchyma by multiple low-density, fluid-filled areas surrounded by a thin rim of residual renal parenchyma, due to xanthogranulomatous pyelonephritis. Abnormal enhancing soft tissue fills the renal pelvis around the stone.* **(Right)** *Axial CECT in the same patient shows tumor extending into the dilated calyces. Biopsy specimen revealed squamous cell carcinoma.*

Stage II (T2 N0 M0)



Stage II (T2 N0 M0)

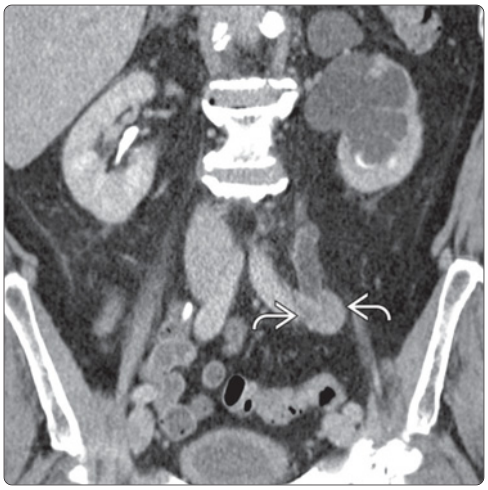


(Left) *Axial T2WI MR shows a left renal pelvis mass with extension to the renal calyces. There is no evidence of invasion into the peripelvic fat or into the renal parenchyma.* **(Right)** *Axial T1WI C+ FS MR shows a large mass filling the renal pelvis and calyces. The mass is hypoenhancing relative to the kidney. Although the mass is distending the renal pelvis, there is a complete circle of ureteric enhancement, indicating that the peripelvic fat is not invaded by tumor.*

Stage II (T2 N0 M0)

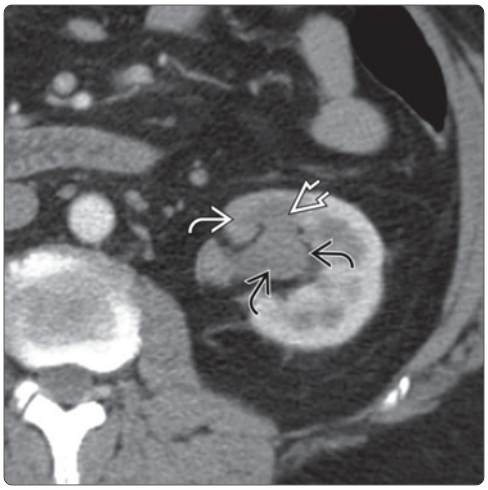


Stage II (T2 N0 M0)

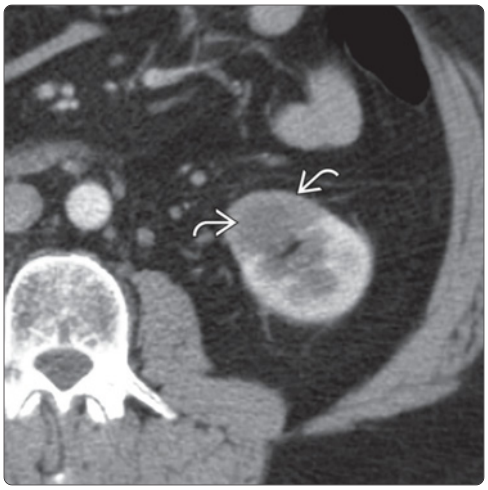


(Left) *Axial CT IVP shows a mass centered on the left ureter. Although the mass is large and appears to extend beyond the contour of the ureter, pathological evaluation of the resected specimen showed that the tumor invaded the muscularis and was surrounded by a concentric mass of fibrous tissue.* **(Right)** *Coronal CT IVP in the same patient shows the exophytic ureteral mass causing hydronephrosis and dilatation of the proximal ureter.*

Stage III (T3 N0 M0)

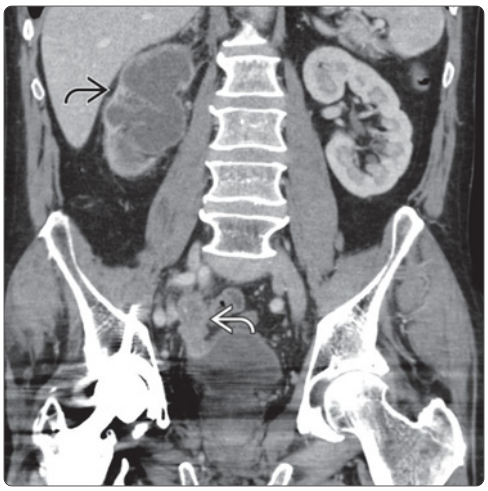


Stage III (T3 N0 M0)

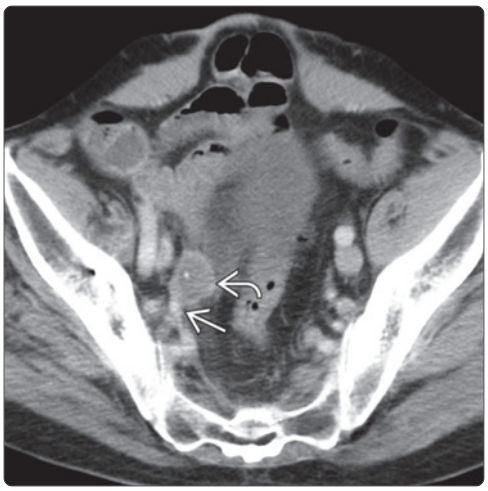


(Left) *Axial CT IVP shows a soft tissue mass distending the renal pelvis and extending into an anterior calyx. The mass also invades into the renal parenchyma; the tumor enhances less than the normal parenchyma.* **(Right)** *Axial CT IVP in the same patient shows renal parenchymal invasion. The involved kidney is hypoenhancing compared to the normal enhancing kidney. There is no evidence of perinephric fat involvement.*

Stage IV (T4 N0 M0)



Stage IV (T4 N0 M0)



(Left) *Coronal CECT shows right side hydronephrosis and a right ureteric mass involving the ureterovesical junction.* **(Right)** *Axial CECT in the same patient shows the right ureteric mass invading into the periureteric fat to involve the right internal iliac artery.*

Stage IV (T2 N1 M0)

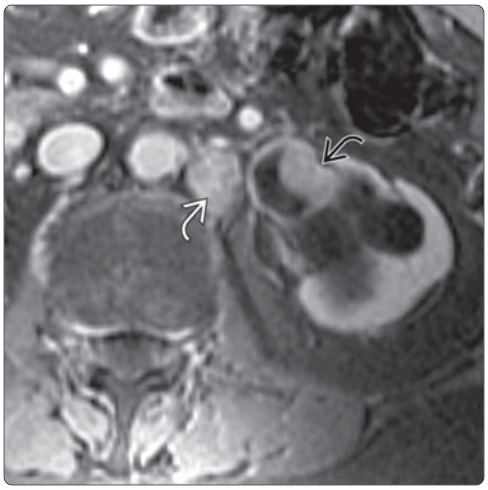


Stage IV (T2 N1 M0)

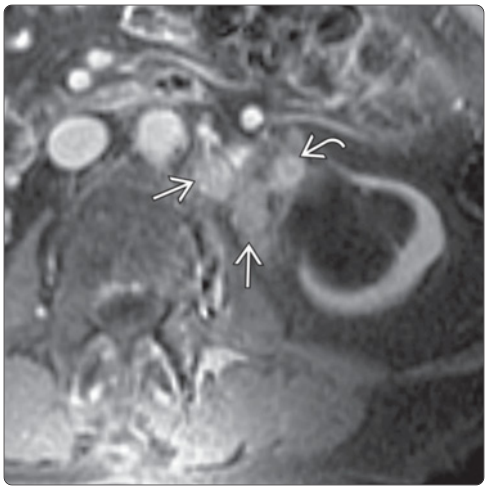


(Left) *Axial T2WI MR shows a polypoid mass of intermediate signal intensity within the left renal pelvis. The mass does not infiltrate into the renal sinus fat or the kidney.* **(Right)** *Axial T2WI MR in the same patient shows an intermediate signal intensity renal pelvis mass infiltrating the upper ureter and causing hydronephrosis. Multiple paraaortic lymph nodes, each < 2 cm, are present.*

Stage IV (T2 N1 M0)

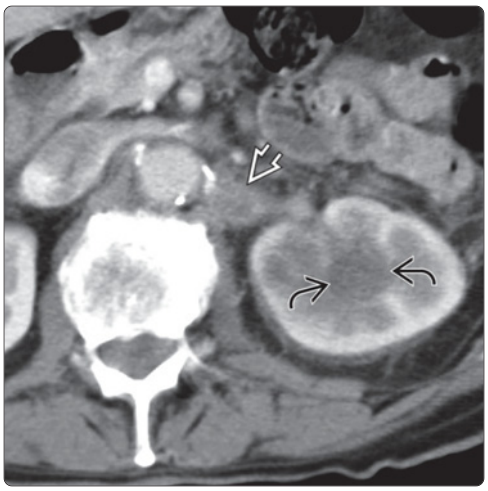


Stage IV (T2 N1 M0)



(Left) *Axial T1WI C+ FS MR in the same patient shows an enhancing renal pelvis mass with no evidence of invasion of renal parenchyma. There is an enhancing paraaortic lymph node that measures just < 2 cm.* **(Right)** *Axial T1 C+ FS MR in the same patient shows involvement of the upper ureter and multiple paraaortic lymph nodes.*

Stage IV (T2 N1 M0)

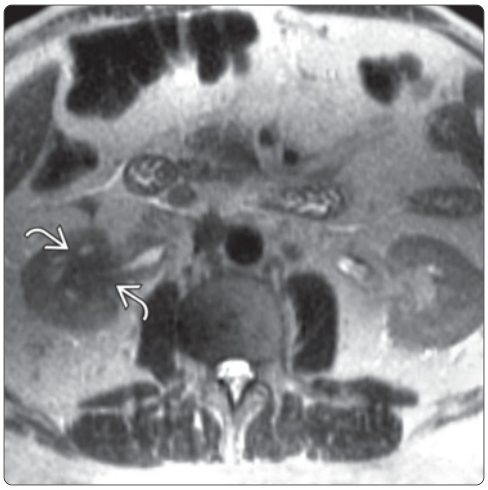


Stage IV (T2 N1 M0)

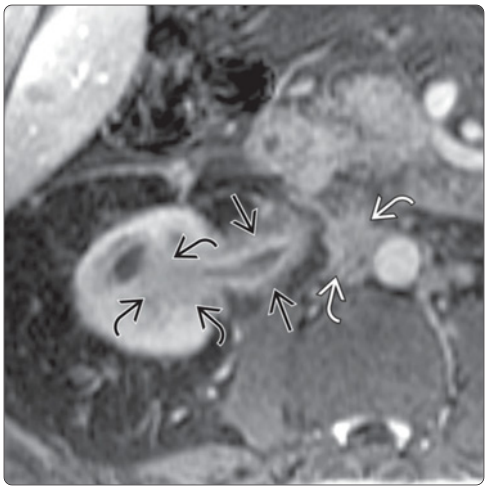


(Left) *Axial CECT in a 76-year-old man who presented with hematuria shows an enhancing soft tissue mass filling and expanding the left upper calyx (oncocalyx). An ill-defined, left, paraaortic, 1.8 cm lymph node is also present.* **(Right)** *Retrograde pyelogram in the same patient shows amputation of the left upper calyx.*

Stage IV (T3 N1 M0)

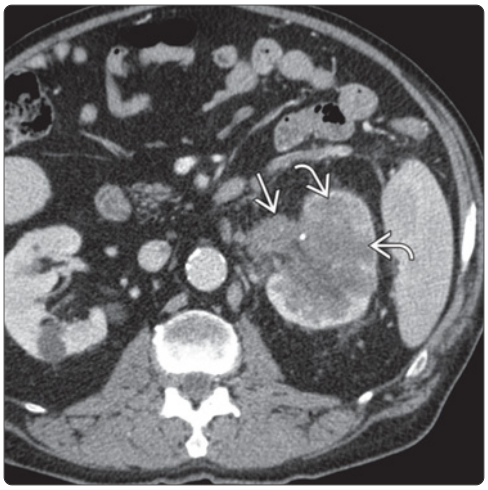


Stage IV (T3 N1 M0)

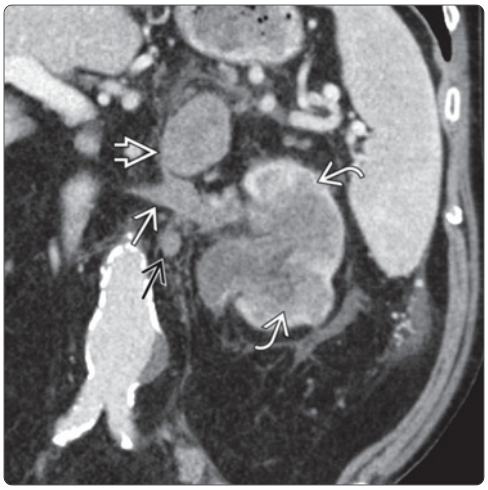


(Left) *Axial T2WI MR shows a low signal intensity mass obliterating the right renal sinus fat.* **(Right)** *Axial T1 C+ FS MR shows an enhancing mass obliterating the renal sinus fat. Note thickening and mucosal enhancement of the renal pelvis, which can be due to submucosal tumor spread or associated inflammatory changes. Also note the infiltrative enhancing nodal mass occluding the inferior vena cava.*

Stage IV (T4 N1 M0)

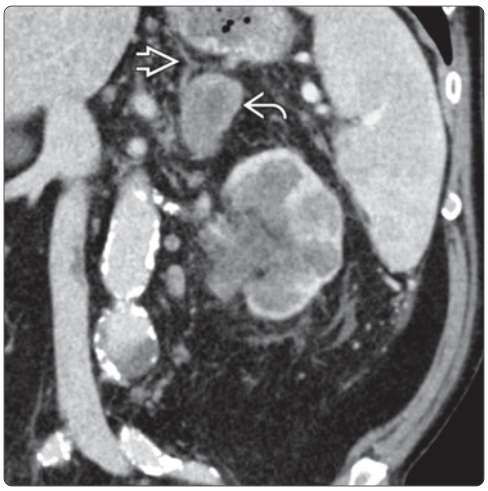


Stage IV (T4 N1 M0)

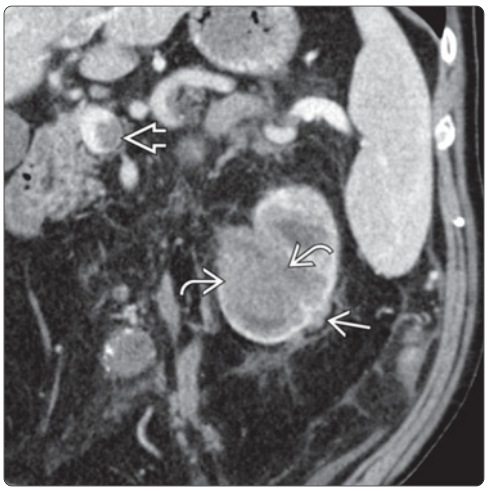


(Left) *Axial CECT shows a mass filling the left renal pelvis and calyces, as well as invading the renal parenchyma. The mass also infiltrates the renal vein.* **(Right)** *Coronal CECT in the same patient shows the mass infiltrating most of the kidney. There is involvement of the left renal vein and left adrenal vein, forming a mass that is separate from the adrenal gland. There is also a 2 cm enlarged lymph node.*

Stage IV (T4 N1 M0)

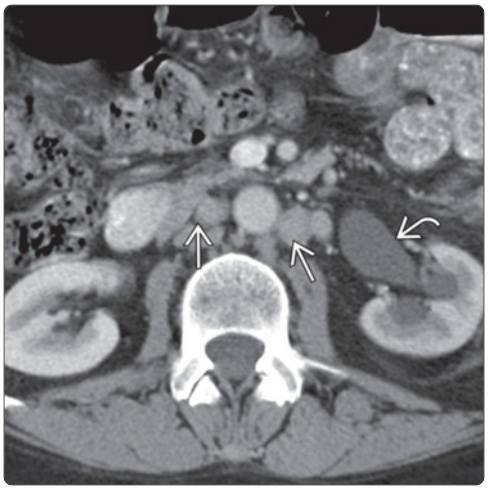


Stage IV (T4 N1 M0)

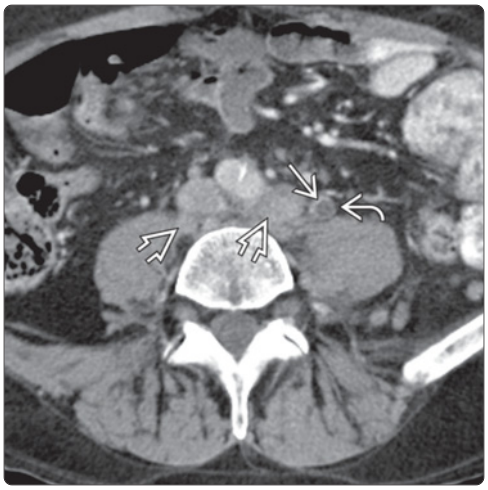


(Left) *Coronal CECT in the same patient shows the mass involving the left adrenal vein separate from the normal adrenal gland.* **(Right)** *Coronal CECT in the same patient shows the infiltrative mass involving almost the entire left kidney with violation of the renal capsule and extension into the perinephric fat. An incidental finding was thrombosis of the portal vein.*

Stage IV (T2 N2 M0)

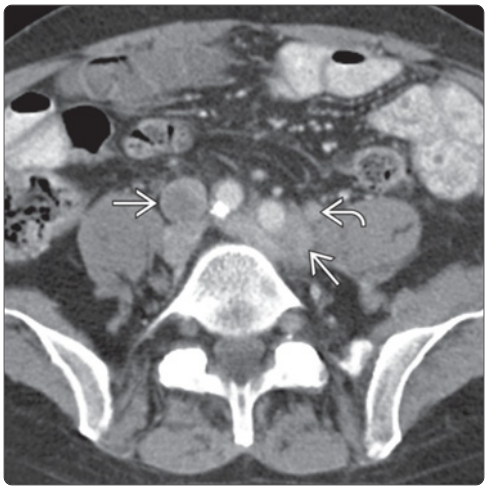


Stage IV (T2 N2 M0)



(Left) *Axial CECT in a 56-year-old man who presented with hematuria shows left side hydronephrosis with delayed left renal contrast excretion. Multiple paraaortic lymph nodes are present.* **(Right)** *Axial CECT in the same patient shows left side hydroureter just above the level of obstruction with a rim of enhancing ureteral wall thickening due to tumor extension. Multiple paraaortic lymph nodes are present.*

Stage IV (T2 N2 M0)

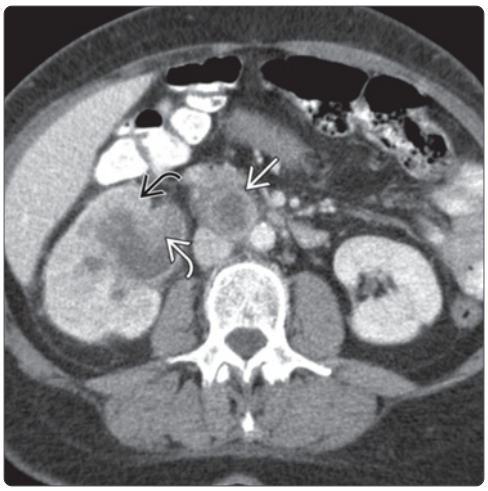


Stage IV (T2 N2 M0)

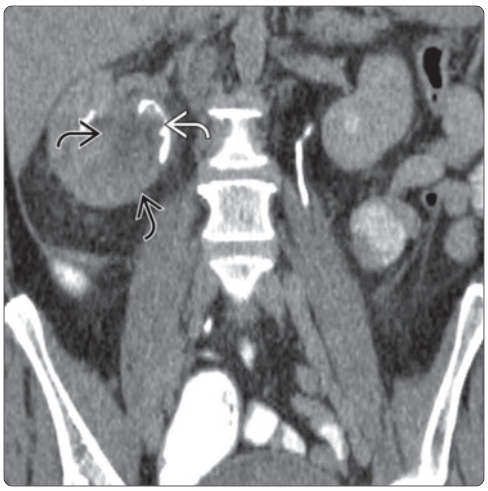


(Left) *Axial CECT in the same patient shows an enhancing mass obstructing the ureter, as well as multiple retroperitoneal lymph nodes.* **(Right)** *Coronal CECT in the same patient shows an enhancing ureteric soft tissue mass with dilatation of the proximal ureter and multiple paraaortic lymph nodes. The cystic lesion in the left side of the pelvis is a lymphocele resulting from previous pelvic nodal dissection for prostate carcinoma.*

Stage IV (T3 N2 M0)

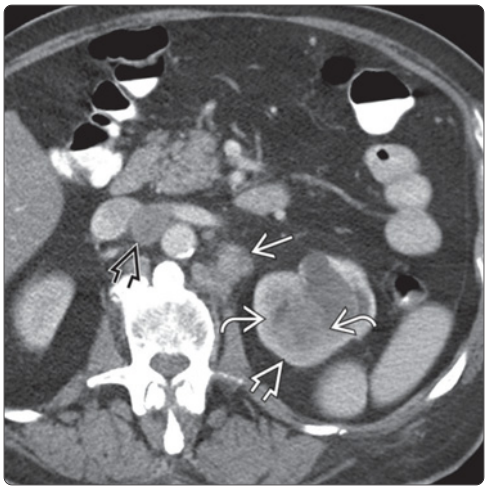


Stage IV (T3 N2 M0)

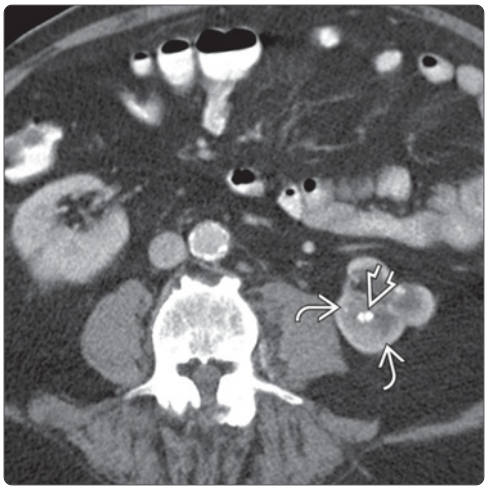


(Left) *Axial CECT shows a right renal pelvis mass that invades into the renal parenchyma but not through the kidney into the perinephric fat. There is also a single enlarged necrotic aortocaval lymph node that measures 3 cm.* **(Right)** *Coronal CECT in the same patient shows the right renal pelvis mass invading into the renal parenchyma. There is no invasion of the perinephric fat.*

Stage IV (T3 N2 M1)

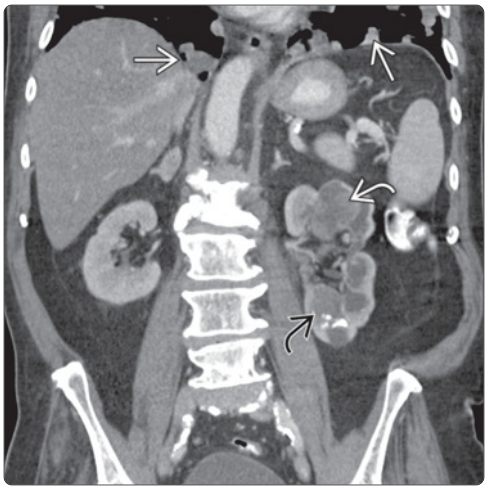


Stage IV (T3 N2 M1)

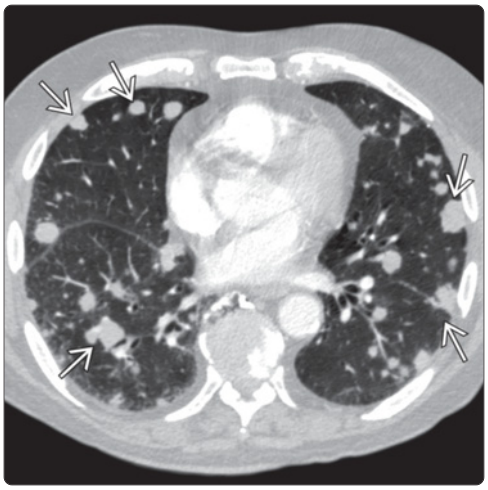


(Left) *Axial CECT in a 77-year-old man who presented with gross hematuria shows an upper pole infiltrative renal mass that extends to, but not beyond, the renal capsule. Multiple paraaortic and aortocaval lymph nodes are also present.* **(Right)** *Axial CECT in the same patient shows a lower pole renal mass that contains nodular calcifications.*

Stage IV (T3 N2 M1)

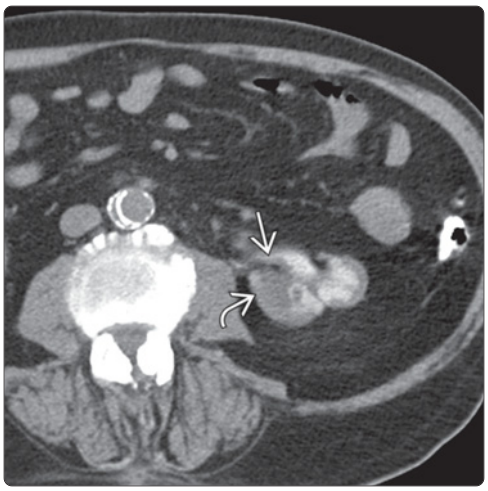


Stage IV (T3 N2 M1)

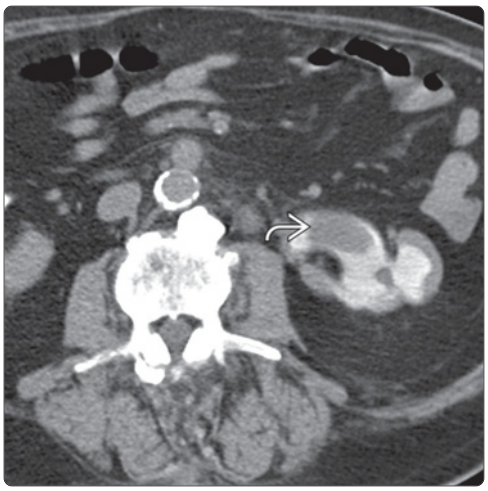


(Left) *Coronal CECT in the same patient shows 2 renal masses, 1 in the upper pole and 1 in the lower pole. It is difficult to determine whether these masses are renal cell carcinomas or transitional cell carcinomas arising from the collecting system. Note also numerous pulmonary metastases.* **(Right)** *Axial CECT in the same patient shows numerous pulmonary nodules due to parenchymal pulmonary metastases.*

Stage IV (T3 N2 M1)



Stage IV (T3 N2 M1)



(Left) *Axial CECT in the same patient 1 day later shows high-density contrast within the collecting system. The lower pole mass extends to involve the adjacent part of the renal pelvis.* **(Right)** *Axial CECT in the same patient shows a large filling defect that changed position after the patient assumed a prone position, behavior consistent with a floating blood clot.*

Renal Lymphoma

KEY FACTS

IMAGING

- CECT
  - Multiple bilateral renal masses (40-60%)
  - Homogeneous, mild enhancement (10-20 HU)
  - Retroperitoneal adenopathy (25%), splenomegaly, or lymphadenopathy at other sites
  - Infiltration of renal parenchyma or sinus with maintenance of reniform shape; may simulate transitional cell carcinoma
  - Extranodal sites: Bowel, brain, liver, bone marrow
- MR
  - T1: Iso- to slightly hypointense
  - T2: Typically hypointense
  - T1WI C+ FS: Mild enhancement
- US
  - Solid, hypoechoic lesions relative to renal parenchyma
  - ± internal flow; power Doppler improves sensitivity

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma
- Transitional cell carcinoma
- Infection

PATHOLOGY

- Renal involvement of non-Hodgkin lymphoma to Hodgkin lymphoma ratio is 10:1

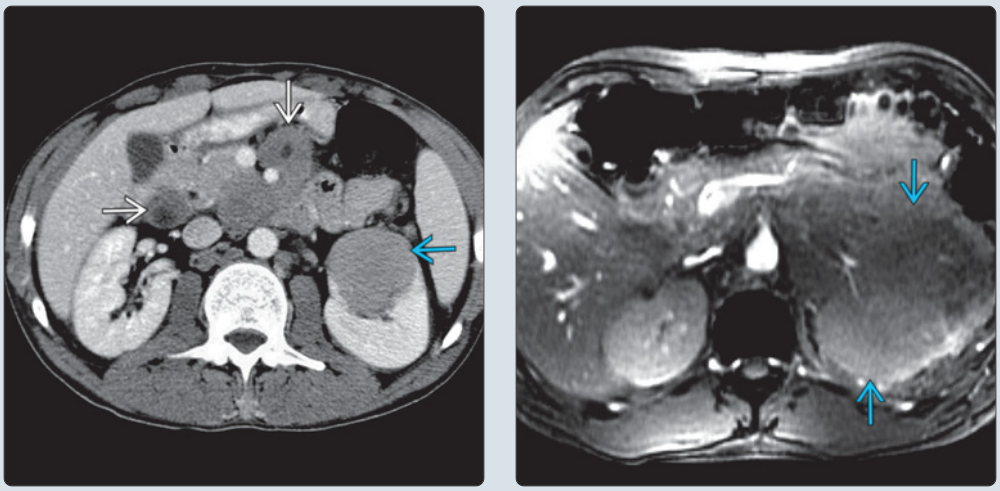
CLINICAL ISSUES

- Usually asymptomatic
- ± flank pain, fever, weight loss

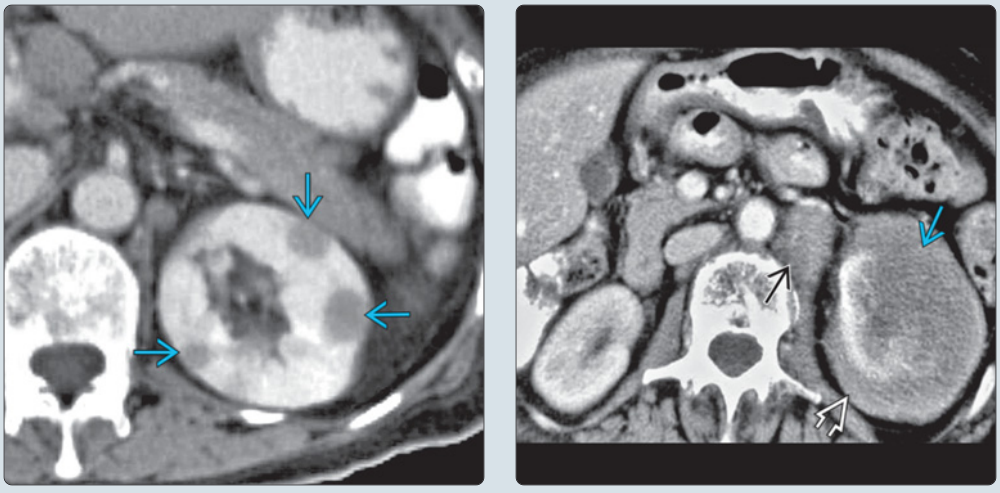
DIAGNOSTIC CHECKLIST

- CECT or MR is best for evaluation of indeterminate renal masses
- Consider renal lymphoma when there are discreet or infiltrative renal masses in setting of systemic adenopathy
- Biopsy often required for diagnosis

(Left) *Axial CECT in a 34-year-old man with AIDS shows a well-circumscribed, mildly enhancing mass in the left kidney. There are also several enlarged lymph nodes with hypoattenuating areas of necrosis due to non-Hodgkin lymphoma.* **(Right)** *Axial T2* GRE MR shows an ill-defined tumor infiltrating the left kidney and perirenal space. The patient also had systemic adenopathy that is not shown on this image. Biopsy of the renal mass revealed B-cell non-Hodgkin lymphoma.*



(Left) *Axial CECT shows multiple, small, homogeneous, hypoattenuating masses in a patient with systemic non-Hodgkin lymphoma. Renal lymphoma may present as numerous hypoattenuating lesions or as an infiltrating mass that invades the kidney and perinephric space.* **(Right)** *Axial CECT shows ill-defined tumor infiltrating the left kidney and extending into the perirenal space. An adjacent retroperitoneal node is enlarged. Pathology revealed non-Hodgkin lymphoma.*



TERMINOLOGY

Definitions

- Malignant tumor of B-cell lymphocytes

IMAGING

General Features

- Best diagnostic clue
  - Discreet or infiltrative renal masses in setting of systemic adenopathy
- Location
  - Bilateral > unilateral
- Size
  - Variable; multiple small mass vs. large, infiltrative masses

Radiographic Findings

- Angiography Findings
  - Hypovascular to avascular
  - Attenuation of segmental and interlobar arteries

CT Findings

- NECT
  - Multiple bilateral masses (50-60%)
- CECT
  - Homogeneous, mild enhancement (10-20 HU)
  - Adenopathy (25%), splenomegaly
  - Infiltration of parenchyma/sinus; normal contour
    - May simulate transitional cell carcinoma
  - Direct extension into hilum or pericapsular region from retroperitoneal lymph nodes (25%)
  - Diffuse renal enlargement (10%)
    - Attenuated collecting systems
  - ± renal vein and inferior vena cava thrombus
  - Extranodal sites: Bowel, brain, liver, bone marrow

MR Findings

- T1WI
  - Iso- to slightly hypointense
- T2WI
  - Typically hypointense
- DWI
  - Usually restricted diffusion
- T1WI C+ FS
  - Mild enhancement

Ultrasonographic Findings

- Grayscale ultrasound
  - Solid, hypoechoic lesions relative to renal parenchyma
- Color Doppler
  - ± internal flow; power Doppler improves sensitivity

Imaging Recommendations

- Best imaging tool
  - Contrast enhanced CT or MR

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma

- Solitary, heterogeneously enhancing mass
- ± central necrosis, vascular invasion, adenopathy

Transitional Cell Carcinoma

- Poorly circumscribed, infiltrative, hypovascular mass
- Filling defect within collecting system ± obstruction

Infection

- Pyelonephritis ± abscess
- Imaging: Infiltrative vs. necrotic mass/collection
- Diagnosis aided by clinical/laboratory findings

PATHOLOGY

General Features

- Etiology
  - Immunosuppression: Iatrogenic (e.g., transplant) or acquired (e.g., AIDS, autoimmune)
- Most renal lymphoma is secondary to systemic disease
  - Hematogenous (90%) vs. direct nodal extension

Staging, Grading, & Classification

- Renal involvement of NHL:HL = 10:1

Microscopic Features

- Lymphoepithelial lesions
- NHL: Large cell lymphoma is most common
- AIDS: Small cell lymphoma is most common

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic
- Other signs/symptoms
  - ± hematuria, flank pain, fever, weight loss

Demographics

- No age, gender, or ethnic predilection
- Systemic lymphoma: 6% involve kidneys at presentation

Natural History & Prognosis

- 57% have complete remission after treatment
- Fatal lymphoma: 33% have renal involvement

Treatment

- Chemotherapy ± radiation therapy
- ± debulking or ureteral stenting if obstruction

DIAGNOSTIC CHECKLIST

Consider

- Multiple renal masses with widespread systemic adenopathy: Consider lymphoma

Image Interpretation Pearls

- CT or US-guided biopsy often needed for diagnosis

SELECTED REFERENCES

1. Allen BC et al: Characterizing solid renal neoplasms with MRI in adults. Abdom Imaging. 39(2):358-87, 2014
2. Davarpanah AH et al: MR imaging of the kidneys and adrenal glands. Radiol Clin North Am. 52(4):779-98, 2014
3. Ganeshan D et al: Imaging of primary and secondary renal lymphoma. AJR Am J Roentgenol. 201(5):W712-9, 2013
4. Sheth S et al: Imaging of renal lymphoma: patterns of disease with pathologic correlation. Radiographics. 26(4):1151-68, 2006

Renal Metastases

KEY FACTS

IMAGING

- Renal metastases often found in setting of other visceral and nodal metastases
- CT
  - Multiple solid, hypoattenuating renal masses
  - Typically located in renal cortex or at corticomedullary junction
  - Variable enhancement (depending on primary tumor type)
    - Hypervascular: Melanoma, breast, neuroendocrine
- MR
  - T1WI: Typically iso- to hypointense
  - T2WI: Typically hyperintense
- Best advice for evaluation of indeterminate renal lesions
  - Contrast-enhanced CT or MR
  - PET/CT for staging of malignancy
- US
  - Multiple solid, hypoechoic lesions at grayscale
  - Variable color Doppler flow (depending on tumor type)

TOP DIFFERENTIAL DIAGNOSES

- Renal cell carcinoma
- Transitional cell carcinoma
- Lymphoma
- Infection (pyelonephritis, abscess)

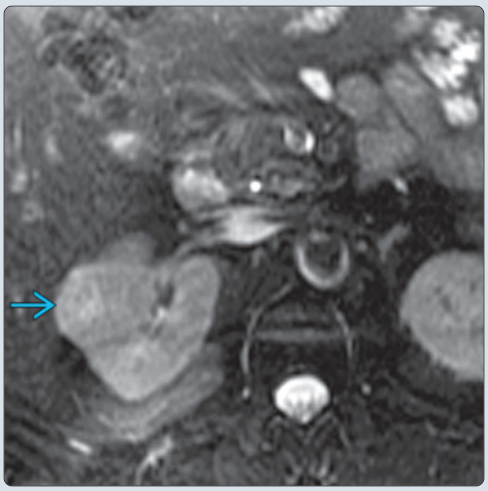
PATHOLOGY

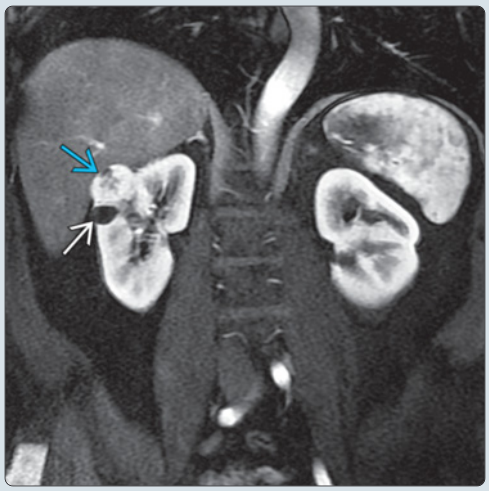
- Common etiologies for renal metastases
  - Bronchogenic carcinoma
  - Breast carcinoma
  - Melanoma
  - Gastric carcinoma
  - Contralateral renal cell carcinoma

DIAGNOSTIC CHECKLIST

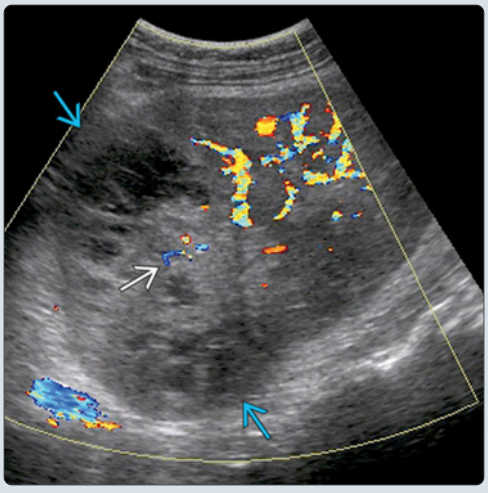
- Imaging features of renal metastases often nonspecific
- Percutaneous biopsy often required for definitive diagnosis

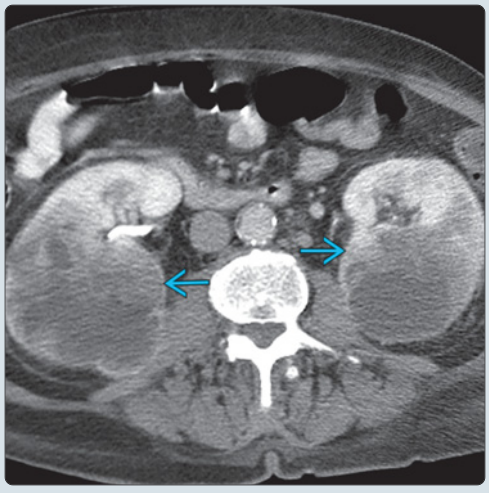
(Left) *Axial T2 MR with fat suppression reveals a solid mass ⇨ centered in the lateral cortex of the right kidney that has intermediate to slightly increased signal intensity relative to the normal adjacent renal parenchyma.* **(Right)** *Post contrast coronal T1W MR with fat suppression in the same patient shows a hypervascular mass ⇨ with central necrosis in the right kidney. Pathology was consistent with metastatic breast carcinoma. An incidental cyst ➡ is inferior to the metastasis.*





(Left) *Sagittal color Doppler shows a large, heterogeneous mass ⇨ replacing the superior pole of the right kidney due to metastatic non-small cell lung carcinoma. Note internal color flow ➡ within the metastasis. There was a similar appearing mass in the contralateral kidney (not shown).* **(Right)** *Subsequent axial CECT in the same patient demonstrates bilateral, heterogeneously enhancing renal masses ⇨ with areas of necrosis due to metastatic non-small cell lung carcinoma.*





TERMINOLOGY

Definitions

- Metastases from primary cancer of other sites

IMAGING

General Features

- Best diagnostic clue
  - Multiple renal masses in patient with primary malignancy
    - ± perirenal & retroperitoneal lesions
  - Hematogenous > direct extension
- Location
  - Bilateral > unilateral
  - Cortical or corticomedullary junction
- Size
  - Variable; often relatively uniform size
- Morphology
  - Typically round; rarely irregular or wedge-shaped
    - Rarely deform renal contour or capsule

CT Findings

- NECT
  - Multiple iso- to hypoattenuating solid renal lesions
- CECT
  - Multiple hypoattenuating lesions
  - Typically in cortex or corticomedullary junction
  - Variable enhancement (depending on tumor type)
    - Hypervascular: Melanoma, breast, neuroendocrine

MR Findings

- T1WI
  - Variable signal intensity (SI) (depending on tumor type)
  - Most will be hypo- to isointense relative to renal parenchyma
  - Hemorrhagic metastases (melanoma, breast) can have high SI
  - High SI can also be due to melanin
- T2WI
  - Typically high SI
  - Hemorrhage: Variable SI (depending on age of blood products)
- DWI
  - Often high SI, but variable due to tumor type & degree of cellularity

Ultrasonographic Findings

- Grayscale ultrasound
  - Multiple solid, hypoechoic lesions
- Color Doppler
  - Variable internal flow
    - Power Doppler increases sensitivity

Nuclear Medicine Findings

- PET/CT
  - Variable FDG avidity depends on tumor type
    - FDG avid: Melanoma, breast, neuroendocrine

Imaging Recommendations

- Best imaging tool
  - Contrast-enhanced CT or MR for renal lesions
- Protocol advice
  - Staging aided by full-body PET/CT

DIFFERENTIAL DIAGNOSIS

Renal Cell Carcinoma

- Solitary mass with heterogeneous enhancement
- ± central necrosis, vascular invasion, adenopathy

Transitional Cell Carcinoma

- Poorly circumscribed, infiltrative, hypovascular mass
- Filling defect within collecting system ± obstruction

Lymphoma

- Solid, hypovascular renal masses

Pyelonephritis

- Infiltrative to necrotic mass/collection
- Diagnosis aided by clinical signs/symptoms & laboratory

PATHOLOGY

General Features

- At autopsy, ~ 2x more common than primary renal tumors
- Often too small to be identified by imaging
- Etiology
  - Bronchogenic carcinoma (19.8-23.3%)
  - Breast carcinoma (12.3%)
  - Melanoma
  - Gastric carcinoma
  - Renal cell carcinoma of contralateral kidney

Microscopic Features

- Varies based on primary cancer

CLINICAL ISSUES

Presentation

- Usually asymptomatic; ± hematuria or flank pain

Demographics

- Can affect any age, gender, ethnicity

Natural History & Prognosis

- Prognosis: Typically very poor
- Complications: Hemorrhage, obstruction

Treatment

- Systemic therapy or palliative treatment
- Consider nephrectomy if isolated to 1 kidney

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Percutaneous biopsy often needed for diagnosis

SELECTED REFERENCES

1. Allen BC et al: Characterizing solid renal neoplasms with MRI in adults. Abdom Imaging. 39(2):358-87, 2014
2. Davarpanah AH et al: MR imaging of the kidneys and adrenal glands. Radiol Clin North Am. 52(4):779-98, 2014
3. Sahni VA et al: Imaging management of incidentally detected small renal masses. Semin Intervent Radiol. 31(1):9-19, 2014
4. Roy A et al: Common and uncommon bilateral adult renal masses. Cancer Imaging. 12:205-11, 2012
5. Taneja R et al: Common and less-common renal masses and masslike conditions. Radiol Clin North Am. 50(2):245-57, v-vi, 2012

Nephrocalcinosis

KEY FACTS

IMAGING

- Renal parenchymal sites of calcinosis
  - Medullary nephrocalcinosis (95%)
  - Cortical nephrocalcinosis (5%)
- Scattered punctate calcifications in renal medulla
  - Common in medullary sponge kidney (MSK)
- Dense, confluent medullary calcification
  - Renal tubular acidosis > hyperparathyroidism
- "Tram line" calcification in renal cortex
  - Especially in acute cortical necrosis

TOP DIFFERENTIAL DIAGNOSES

- Renal papillary necrosis
- Renal tuberculosis
- Extrapulmonary *Pneumocystis carinii* or *Mycobacterium avium-intracellulare* infection in AIDS patients

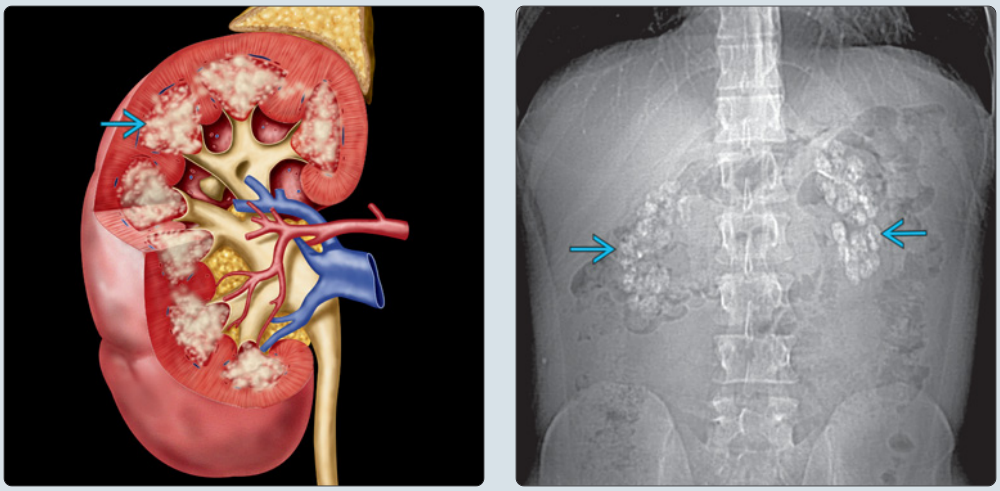
PATHOLOGY

- Medullary nephrocalcinosis
  - Hyperparathyroidism > renal tubular acidosis > medullary sponge kidney
- Cortical nephrocalcinosis
  - Chronic glomerulonephritis > cortical necrosis > rejected renal allograft
- 3 primary mechanisms for calcium deposition
  - Metastatic: Metabolic abnormality leads to calcium deposition in normal kidneys
  - Dystrophic: Calcium deposition in damaged renal parenchyma
  - Urinary stasis: Calcium salts precipitate in dilated tubules (medullary sponge kidney)

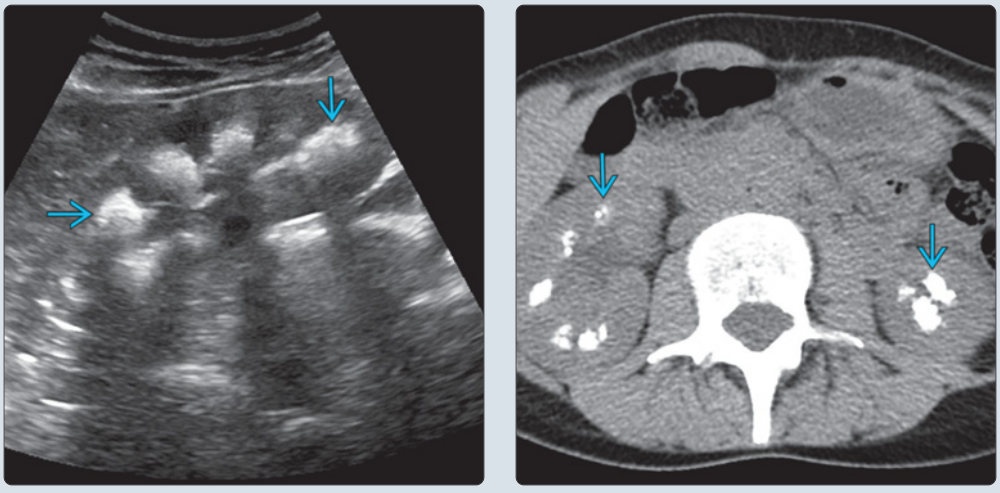
CLINICAL ISSUES

- Nephrocalcinosis is most often asymptomatic
- Increased intestinal absorption of calcium
  - Sarcoidosis and milk-alkali syndrome

(Left) *Graphic shows calcification in the renal pyramids, representing nephrocalcinosis. Severe, diffuse calcification of this type would be typical of renal tubular acidosis.* **(Right)** *AP radiograph shows extensive, dense calcifications in the renal medulla due to medullary nephrocalcinosis in a patient with hyperparathyroidism, the most common etiology of medullary nephrocalcinosis.*



(Left) *Sagittal US in a patient with renal tubular acidosis shows increased echogenicity of the renal pyramids due to extensive nephrocalcinosis. Note that there is shadowing posterior to the annotated pyramids while others do not show shadowing. Medullary nephrocalcinosis can also produce ring-like echogenicity on US.* **(Right)** *Axial NECT in the same patient demonstrates the calcifications in the renal medullae bilaterally. The calcifications can erode through the renal papillae and enter the collecting system.*



TERMINOLOGY

Synonyms

- Medullary nephrocalcinosis, cortical nephrocalcinosis

Definitions

- Radiologically detectable diffuse calcium deposition within renal parenchyma

IMAGING

General Features

- Best diagnostic clue
  - Renal parenchymal calcification identified on CT, radiography, or US
- Location
  - Renal parenchymal sites of calcinosis
    - Medullary nephrocalcinosis (95%)
    - Cortical nephrocalcinosis (5%)
    - Both cortical and medullary (rare)
- Size
  - Kidneys often normal in size and contour
    - Kidneys affected by longstanding disease processes may show variable degree of atrophy
- Morphology
  - Variable patterns of calcification
    - Scattered punctate calcifications in renal medulla
      - Common in medullary sponge kidney
    - Dense, confluent medullary calcification
      - Common in renal tubular acidosis > hyperparathyroidism
    - "Tram line" calcification or punctate calcifications in renal cortex
      - Especially in cortical necrosis

Radiographic Findings

- Radiography
  - Fine, stippled calcification in renal pyramids
  - Coarse, confluent calcification
  - Punctate or "tram line" cortical calcification
- IVP
  - Medullary: Calcification in renal pyramids on preliminary radiographs
  - May see linear striations and cystic spaces in papillae in patients with underlying medullary sponge kidney (MSK)

CT Findings

- NECT
  - Stippled or confluent calcifications in renal parenchyma
  - Ring-like pattern due to relatively increased calcification at corticomedullary junction

Ultrasonographic Findings

- Grayscale ultrasound
  - Medullary: Echogenic renal pyramids
    - May have ring-like pattern with increased echogenicity along periphery of pyramid
    - Posterior shadowing is variable depending on amount and density of calcification
  - Cortical nephrocalcinosis
    - Hyperechoic peripheral renal cortex with acoustic shadowing

Imaging Recommendations

- Best imaging tool
  - Noncontrast CT
- Protocol advice
  - Detection of nephrocalcinosis on radiographs is optimized by low kV technique

MR Findings

- Calcification has low signal intensity on T1WI and T2WI
- No enhancement of densely calcified portions of parenchyma

DIFFERENTIAL DIAGNOSIS

Technical Artifacts

- NECT: Residual contrast material

Renal Papillary Necrosis

- Common in analgesic nephropathy
- Curvilinear, ring-shaped, or triangular calcification in sloughed papillae
- Calcifications arranged in arc conforming to positions of papillary tips
- May be indistinguishable from nephrocalcinosis

Renal Tuberculosis

- Focal amorphous calcification within kidney at sites of caseating infection
- Diffuse: Dense calcification throughout autonephrectomized kidney ("putty" kidney)

Extrapulmonary *Pneumocystis carinii* or *Mycobacterium avium-intracellulare* Infection in AIDS Patients

- May have both cortical and medullary punctate calcification

PATHOLOGY

General Features

- Etiology
  - Medullary nephrocalcinosis
    - Due to hyperparathyroidism (40%)
    - Renal tubular acidosis type 1 (20%)
    - Medullary sponge kidney (20%)
  - Cortical nephrocalcinosis
    - Chronic glomerulonephritis
    - Renal cortical necrosis
    - Transplanted kidney: Chronic rejection
  - 3 primary mechanisms for calcium deposition
    - Metastatic: Metabolic abnormality leads to calcium deposition in medullae of morphologically normal kidneys
    - Dystrophic: Calcium deposition in damaged renal tissue
    - Urinary stasis: Calcium salts precipitate in dilated collecting ducts containing static urine
  - Entities causing medullary metastatic calcification
    - Renal tubular acidosis type 1 (distal)
    - Hyperparathyroidism
    - Hypercalcuria
    - Hyperoxaluria (oxalosis)
  - Nephrocalcinosis due to urinary stasis

- Medullary sponge kidney: Cystic or fusiform dilation of collecting ducts in renal pyramids
- Nephrocalcinosis due to dystrophic calcification
  - Acute cortical necrosis secondary to shock, placental abruption, nephrotoxins
  - Cortical: Chronic glomerulonephritis
- Genetics
  - Type 1 renal tubular acidosis (RTA): Familial form
    - Autosomal dominant inheritance pattern most common
    - May be due to defect in chloride-bicarbonate exchange gene *AE1*
  - Hyperoxaluria: Familial form (oxalosis)
    - Autosomal recessive
- Associated abnormalities
  - Urolithiasis when calculi formed in renal medulla erode into collecting system
    - Common in medullary sponge kidney > hyperparathyroidism
    - Calcium stones grow on papillae
      - Most break loose and enter collecting system → urolithiasis
      - If calcium stones remain in place → medullary nephrocalcinosis

Gross Pathologic & Surgical Features

- Depends on underlying etiology of nephrocalcinosis

Microscopic Features

- Calcium deposition in interstitium, tubule epithelial cells, along basement membranes
  - Also within lumina of tubules

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most often asymptomatic
  - Other signs/symptoms
    - Flank pain, hematuria if associated with urolithiasis
- Clinical profile: Cortical nephrocalcinosis
  - Acute cortical necrosis
    - Acute vascular insult (shock, placental abruption)
    - Nephrotoxic drugs (ethylene glycol, methoxyflurane anesthesia, amphotericin B)
  - Chronic glomerulonephritis
  - Alport syndrome: Hereditary nephritis and nerve deafness
- Clinical profile: Medullary nephrocalcinosis
  - Skeletal deossification
    - Primary and secondary hyperparathyroidism
    - Bony metastases
    - Prolonged immobilization
  - Increased intestinal absorption of calcium
    - Sarcoidosis
    - Milk-alkali syndrome
  - Medullary sponge kidney
  - Hyperoxaluria
    - Hereditary type (oxalosis)
    - Acquired: Secondary to small bowel disease or bariatric surgery
  - RTA type 1 (distal RTA)
    - May be primary or secondary to other systemic disease (Sjögren, lupus, others)
    - Distal tubule unable to secrete hydrogen ions
    - Metabolic acidosis with urinary pH > 5.5
    - Type 2 (proximal) RTA never causes nephrocalcinosis

Demographics

- Age
  - Any
- Epidemiology
  - Incidence: 0.1-6%
  - Medullary sponge kidney: Seen in 0.5% of excretory urograms

Natural History & Prognosis

- Depends on underlying cause of nephrocalcinosis

Treatment

- Options, risks, complications
  - Medullary nephrocalcinosis often complicated by urolithiasis
  - Parathyroid resection recommended in patients with hyperparathyroidism
    - Even if serum calcium is normal, especially if they have renal calculi
      - Prevalence of stone disease is ↑ 4x in patients with hyperparathyroidism

DIAGNOSTIC CHECKLIST

Consider

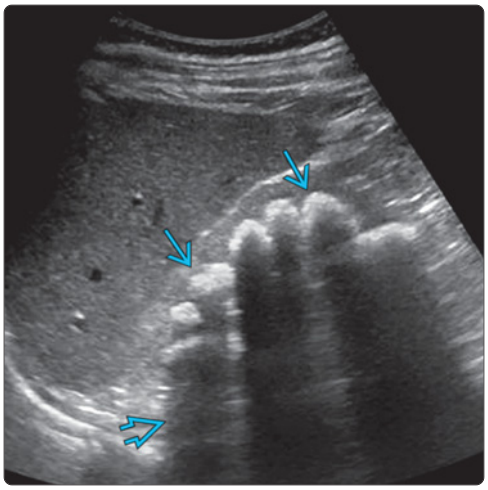
- Focal areas of dystrophic calcification in masses or infection are not considered nephrocalcinosis

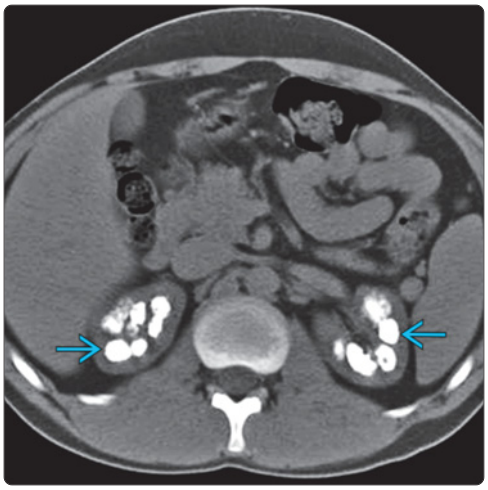
Image Interpretation Pearls

- Massive, dense, medullary nephrocalcinosis usually due to RTA type 1
- Unilateral or segmental medullary multifocal nephrocalcinosis: Medullary sponge kidney

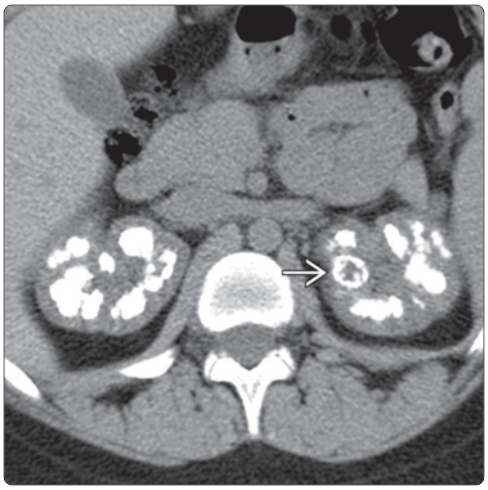
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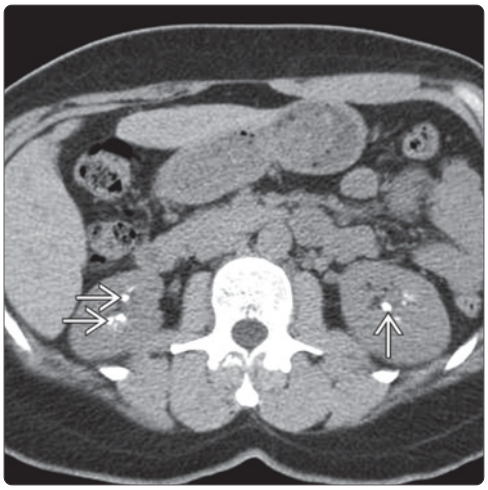
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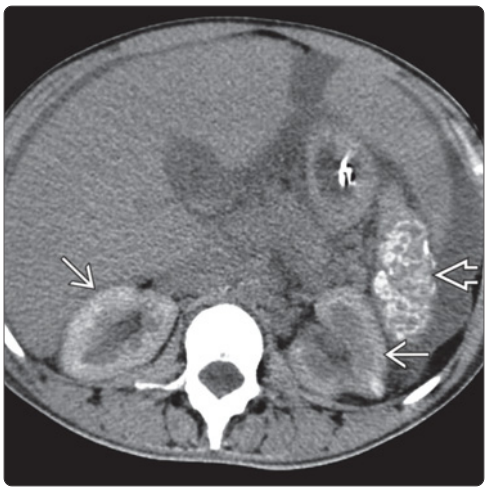
(Left) *Sagittal US demonstrates dense calcification ⇨ in the renal medulla due to extensive medullary nephrocalcinosis in a patient with renal tubular acidosis. Note the extensive posterior shadowing ⇨ that obscures the posterior aspect of the kidney.* **(Right)** *Axial NECT in the same patient reveals the extensive, dense calcifications ⇨ in the renal medulla bilaterally. Unlike medullary sponge kidney, medullary nephrocalcinosis due to hyperparathyroidism and renal tubular acidosis is usually very dense.*





(Left) *Axial NECT in a patient with medullary nephrocalcinosis due to sarcoidosis shows densely calcified renal pyramids. Note the ring-like pattern ➡ at the corticomedullary junction.* **(Right)** *Axial NECT in a patient with medullary sponge kidney (MSK) disease shows multiple discrete calcifications ➡ in the medulla of both kidneys. MSK usually results in multifocal, but not diffuse, calcification in the medullae near the tips of the renal pyramids.*





(Left) *Anteroposterior radiograph demonstrates cortical nephrocalcinosis in a patient with chronic glomerulonephritis. Note that the patient has not received any intravenous contrast material. The cortical calcification is dystrophic due to longstanding inflammation.* **(Right)** *Axial NECT in a young woman with sickle cell anemia and cortical nephrocalcinosis shows diffuse calcification in both renal cortices ➡ and diffuse calcification of a chronically infarcted and atrophic spleen ➡.*

Urolithiasis

KEY FACTS

IMAGING

- Calculi are uniformly dense, except matrix and indinavir stones
- Indinavir calculi: Soft tissue density; deduced from secondary findings (obstruction)
  - Imaging pearl: In AIDS patient with flank pain and obstructed ureter without visible calculi, consider indinavir calculus
- CT findings
  - Dense (several hundred Hounsfield unit) foci in calyces, renal pelvis, ureter, or bladder
  - Soft tissue rim sign: Ureteral wall edema surrounds stone
  - Perinephric stranding: Inflammation/fluid in perinephric space
  - Hydroureteronephrosis if full or partial obstruction
- US findings
  - Round or ovoid echogenic foci
  - Posterior shadowing: Hypoechoic posterior band
  - "Twinkle" artifact: Disorganized, alternating color band posterior to calculus

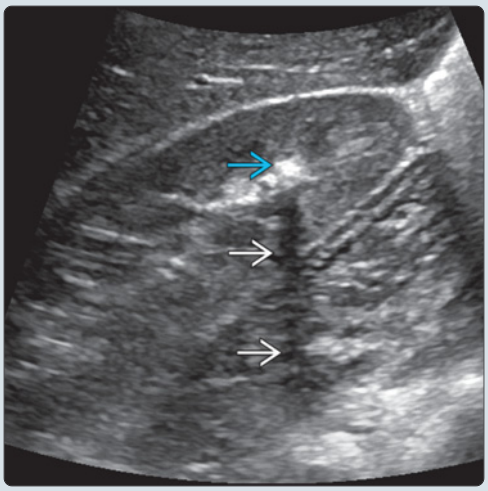
TOP DIFFERENTIAL DIAGNOSES

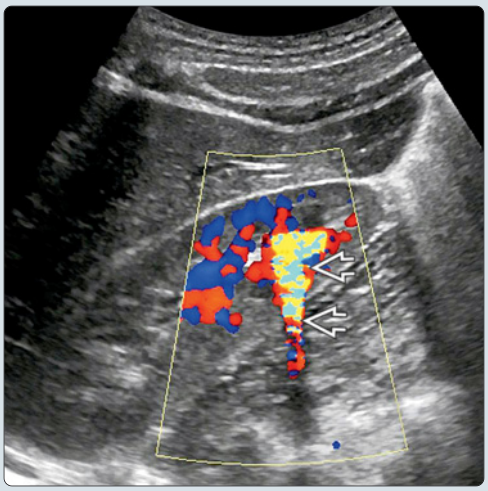
- Renal artery calcification
  - Radiography: Curvilinear parallel lines of calcification in extrarenal, intrarenal, or pelvic arteries
- Nephrocalcinosis
  - Parenchymal calcification: Medulla > cortex
  - May erode and cause nephrolithiasis
- Phlebolith
  - Pelvic phleboliths usually caudal to ureterovesical junction (UVJ) (level of cervix or seminal vesicles)
  - May require excretory phase imaging to differentiate
- Congenital megacalyces and megaureter

CLINICAL ISSUES

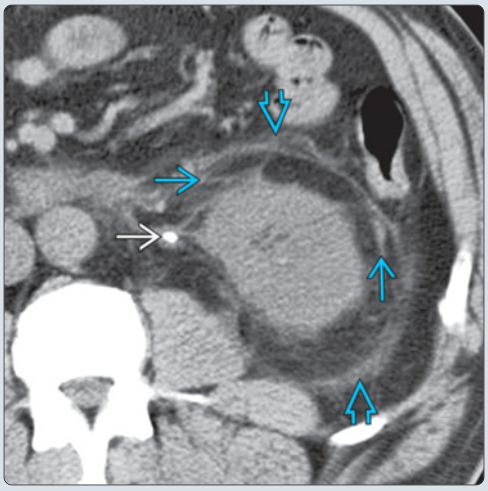
- Small, "nonobstructing" renal calculi can be symptomatic

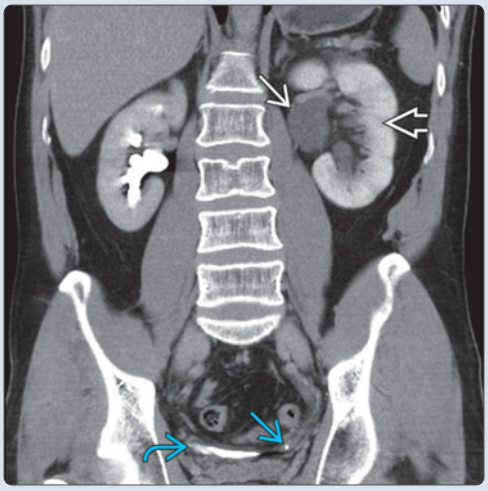
(Left) *Sagittal US image shows an echogenic calculus within a superior pole calyx and posterior shadowing shown by a linear hypoechoic band. In the absence of the shadowing artifact, it may be difficult to differentiate this calculus from sinus fat.* **(Right)** *Sagittal color Doppler US in the same patient reveals "twinkle" artifact due to the highly refractive calculus; the "twinkle" artifact appears as a linear band or tail of disorganized, rapidly changing color posterior to the calculus. Other areas of color flow are due to blood vessels.*





(Left) *Axial NECT shows a proximal ureteral stone with a "rim" of soft tissue, which represents the ureteral wall. Note the moderate degree of perinephric stranding and fascial thickening.* **(Right)** *Coronal CECT during the excretory phase shows moderate left hydronephrosis and delayed nephrogram due to a 3 mm calculus in the distal left ureter. Note the small volume of contrast in the urinary bladder due to excretion from the right kidney.*





TERMINOLOGY

Synonyms

- Kidney, renal, or ureteral "stones"; calculus disease

Definitions

- Concretions within urinary system

IMAGING

General Features

- Best diagnostic clue
  - Small dense focus in ureter with proximal hydronephrosis and perinephric stranding
- Location
  - Upper urinary tract (UT): Calyceal, renal pelvis, or ureteropelvic junction (UPJ)
  - Ureteral calculi: Ureter or ureterovesical junction (UVJ)
  - Lower UT: Bladder, urethral
- Other general features
  - Types of calculi
    - Calcium stones (75-80%): Oxalate &/or phosphate
    - Struvite calculi (15-20%): Magnesium ammonium phosphate (struvite), magnesium ammonium phosphate, and calcium phosphate (triple phosphate)
    - Uric acid calculi (5-10%)
    - Cystine calculi (1-3%)
    - Matrix calculi (rare): Mucoproteins
    - Xanthine calculi (extremely rare)
    - Protease inhibitor calculi: Indinavir induced
    - Milk of calcium: Calcium carbonate & calcium phosphate (carbonate apatite)

Radiographic Findings

- Radiography
  - Based on CT correlation, radiography misses majority of calculi
    - Due to small size, insufficient radiopacity, overlying bones, bowel, etc.
  - Calcium oxalate or phosphate calculi
    - Usually very opaque; variable shape
  - Struvite and cystine calculi
    - Staghorn calculi: Conform to pelvocalyceal system
    - Usually opaque
  - Uric acid and xanthine calculi
    - Usually small, smooth, disk-shaped
    - Rarely opaque or detectable on radiograph
  - Milk of calcium
    - Liquid suspension of calcium salts that are "trapped" in calyceal diverticulum or ureterocele
  - Protease inhibitor calculi
    - Nonopaque on radiography (and sometimes CT)
- IVP
  - Ureteral calculi
    - Enlarged, hydronephrotic kidney
    - Delayed ("obstructive," peak at 6 hours); prolonged (> 24 hours) nephrogram
    - Dense, striated, or absent nephrogram
    - Contrast extravasation; ± forniceal rupture

CT Findings

- NECT
  - Calculi are all uniformly dense except matrix and indinavir stones
  - Radiopacity: Calcium oxalate &/or phosphate > cystine > struvite > uric acid
  - Calcium calculi: 400-600 HU
  - Uric acid and cystine calculi: 100-300 HU
  - Matrix calculi
    - Soft tissue attenuation (pure, rare)
    - Laminated peripheral calcification, diffuse ↑ density or round faintly opaque nodules with densely calcified center (when mixed with calcium salts)
  - Milk of calcium: Layered opaque suspension; stone movement in calyceal diverticulum
  - Indinavir calculi: Not or faintly opaque; deduced from secondary findings (obstruction)
  - Ureteral calculi
    - **Soft tissue rim sign**: Ureteral wall edema at calculus
    - Pseudoureterocele: UVJ edema around calculus
    - Hydroureteronephrosis: Perinephric stranding
- CECT
  - Lucent filling defects (matrix and indinavir stones) during excretory phase
- Dual-energy CT (DECT)
  - Calculi have different attenuation values when imaged at low and high energies
  - Analysis of energy-dependent changes in attenuation allow determination of chemical composition
    - Stone analysis is compared to referenced material-specific attenuation curves
  - Specificity of chemical composition decreases with
    - Small (< 3 mm) calculi
    - Large patient body habitus

MR Findings

- Calculi appear as signal voids (due to lack of mobile protons)
- Secondary signs
  - Renal edema: ↑T2 signal intensity (SI), ↓ T1 SI of parenchyma
  - Perinephric stranding: ↑ T2 SI of perinephric fat
  - Ureter obstruction: Abrupt caliber change of ureteral lumen

Ultrasonographic Findings

- Grayscale ultrasound
  - Focal echogenic foci
    - Not dependent on chemical consistency
  - Posterior shadowing: Linear hypoechoic artifact
- Color Doppler
  - "Twinkle" artifact: Linear band of disorganized, rapidly changing color deep to a highly refractive surface
    - a.k.a. "color comet tail" artifact
  - Lack of urine jet from UVJ into bladder is associated with obstructing ureteral calculus

DIFFERENTIAL DIAGNOSIS

Renal Artery Calcification

- Radiography: Curvilinear parallel lines of calcification in extrarenal, intrarenal, or pelvic arteries
- US
  - Echogenic foci within renal sinus with acoustical shadowing; indistinguishable from < 1 cm calculi
  - Curvilinear appearance; detect > 1 cm calculi
- Differentiate by correlation with radiography or CT

Nephrocalcinosis

- Calcification within parenchyma: Cortex and medulla (most common); can erode and evolve into urolithiasis
- CT and US: Indistinguishable except by location

Phlebolith

- Stone within vein (pelvic, gonadal)
- Radiography: Multiple, round, central lucency
- CT
  - Lack of soft tissue rim sign
  - "Comet" or "tail" sign
    - Adjacent eccentric, tapering soft tissue attenuation (noncalcified portion of pelvic vein)
- Distinguishing ovarian vein phlebolith from ureteral calculus is difficult
  - Follow course of vessel and ureter; look for signs of ureteral obstruction
  - Pelvic phleboliths usually caudal to UVJ (level of cervix or seminal vesicles)

Congenital Megacalyces and Megaureter

- Rare congenital anomaly causes dilated ureters

PATHOLOGY

General Features

- Etiology
  - Calcium calculi
    - Idiopathic (85%): Idiopathic hypercalciuria
    - Acquired (15%): Hyperparathyroidism, sarcoidosis, renal tubular acidosis, hyperoxaluria, steroids, Cushing syndrome, immobilization, ↑ vitamin D
  - Struvite calculi: Urinary tract infections (UTI) (*Proteus*, *Klebsiella*, *Pseudomonas*)
  - Uric acid calculi: Hyperuricosuria (25% with gout), ileostomy, chemotherapy, acidic and concentrated urine, adenine phosphoribosyltransferase deficiency
  - Cystine calculi: Cystinuria (autosomal recessive)
  - Matrix calculi: Chronic UTI, urine stasis, obstruction
  - Xanthine calculi: Xanthine oxidase deficiency
  - Milk of calcium in calyceal diverticula, ureteroceles
  - Risk factors
    - Environment: Warm climates, summer
    - Medications: Acetazolamide, indinavir
    - Anatomical abnormalities: UPJ obstruction (horseshoe or ectopic kidney), PC diverticula, tubular ectasia, urinary diversion
- Most are mixed composition; > 50% contain calcium salts

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Upper UT: Asymptomatic, flank pain, fever
  - Ureteral calculi
    - Acute colicky flank pain radiating to groin
  - Lower UT: Asymptomatic, dysuria, dull/sharp pain radiating to penis, buttocks, perineum, or scrotum
- Laboratory data
  - Urinalysis: Hematuria, crystals ± bacteruria
- Diagnosis: Ureteral calculi by CT
  - ≤ 4 mm with soft tissue rim sign (91% sensitive)
  - Hydroureteronephrosis and perinephric stranding (97% positive and 93% negative predictor value)

Demographics

- Gender
  - Overall, M:F = 3:1
    - Struvite and matrix stone: M:F = 1:2-3
- Epidemiology
  - Prevalence: 2-3%; 40-60 years of age
  - Incidence: 1-2 per 1,000; peak at 20-40 years of age

Natural History & Prognosis

- Spontaneous passage through ureter: 80% (< 4 mm), 50% (4-6 mm), 20% (> 8 mm)
- Complications: Obstruction, infection, abscess, and renal insufficiency
- Prognosis: Recurrence without treatment: 10% at 1 year, 35% at 5 years, 50% at 10 years

Treatment

- ↑ hydration (2 L urine/day), restrict diet (protein, sodium, calcium) and give drugs (thiazides or allopurinol)
- Extracorporeal shock wave lithotripsy, nephrostolithotomy, endoscopic retrieval, or suprapubic cystolithotomy
- Follow-up recurrence only: 4-6 weeks after treatment, 24-hour urine

DIAGNOSTIC CHECKLIST

Consider

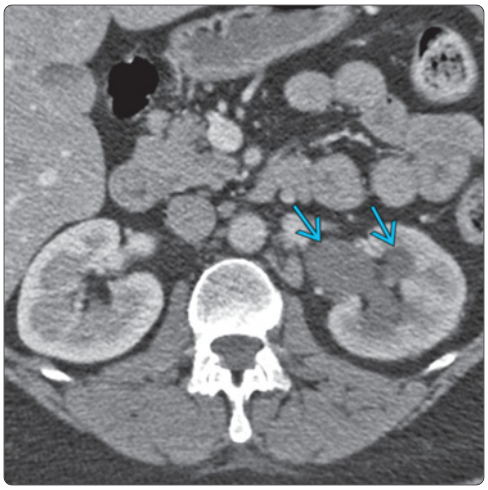
- Avoid repeat CT in young patients with known or clinically evident calculi (excessive radiation)
- In pregnant patients with renal colic symptoms, begin imaging work-up with US
  - If US is inconclusive, consider MR before CT

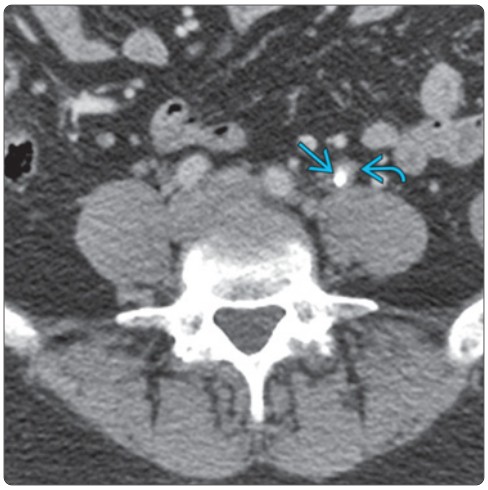
Image Interpretation Pearls

- Small dense focus in course of ureter with perinephric stranding ± hydronephrosis

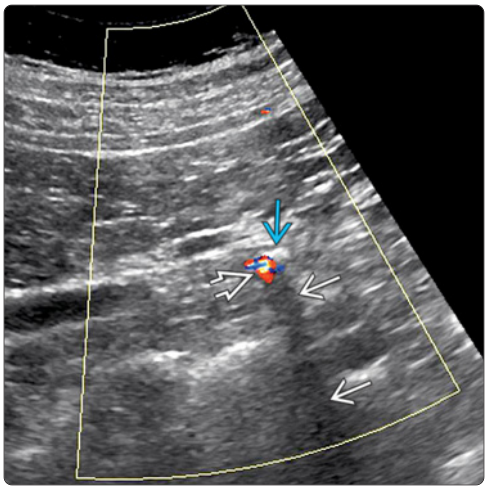
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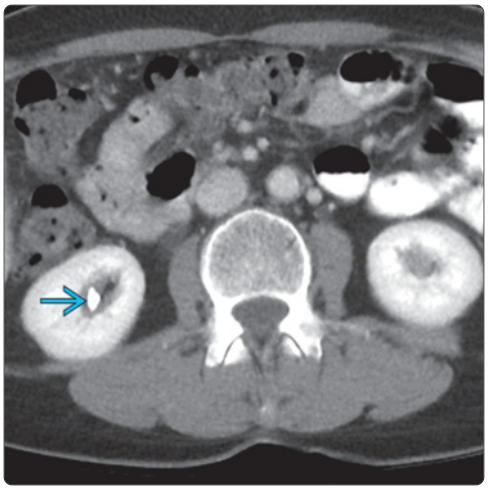
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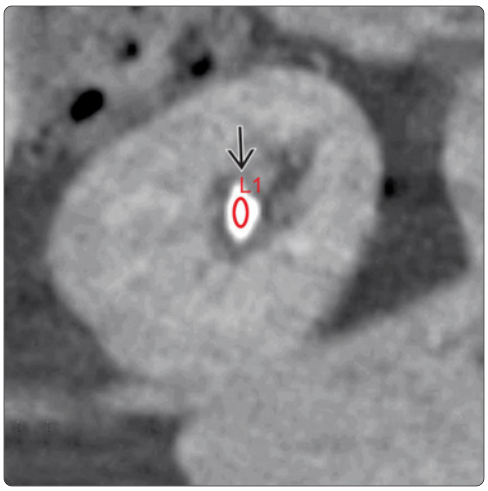


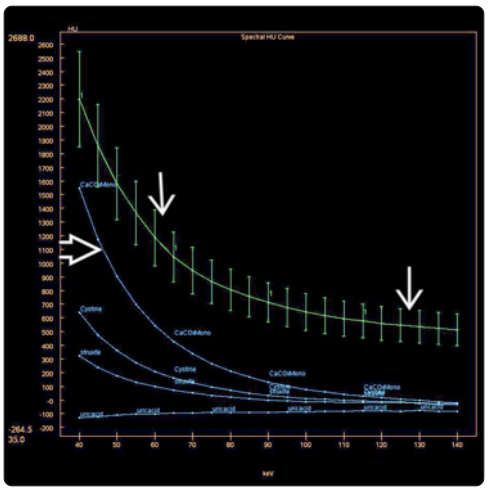
(Left) *Axial CECT obtained during the corticomedullary phase reveals moderate left hydronephrosis. On the excretory phase (not shown), the left nephrogram was delayed.* **(Right)** *Axial CECT more inferiorly in the same patient during the corticomedullary phase reveals a calculus in the lumen of the mid left ureter. Note the "soft tissue rim" sign due to the edematous ureteral wall surrounding the calculus. Recognition of this sign allows differentiation of ureteral calculi from phleboliths.*





(Left) *Sagittal color Doppler US in the same patient shows the echogenic calculus in the ureteral lumen; its location can be aided by shadowing & "twinkle" artifact, shown by the focus of disorganized color Doppler signal posterior to the calculus.* **(Right)** *Axial CECT using dual-energy technique shows a nonobstructing calculus in a right inferior calyx. Dual-energy CT can be used to characterize the composition of renal calculi by exploiting the change in attenuation when images are acquired at different energies.*





(Left) *Axial virtual noncontrast CT shows a region of interest (ROI) drawn over the right renal calculus.* **(Right)** *Spectral attenuation curve representing a plot of attenuation in Hounsfield units (HU, y-axis) vs. energy level (keV, x-axis) has been generated from the ROI. The curve of the calculus most closely approximates the curve of calcium oxalate dihydrate, 1 of several included attenuation profiles for calculi of known composition. The effective Z value of 13 was also consistent with calcium oxalate dihydrate.*

Paroxysmal Nocturnal Hemoglobinuria

KEY FACTS

TERMINOLOGY

- Rare, acquired stem cell disorder that results in predisposition to complement-mediated hemolysis
  - Commonly abbreviated as PNH

IMAGING

- CT
  - No specific CT findings
  - ± ↑ attenuation of renal cortices on NECT
- MR
  - Low signal intensity (SI) in renal cortices is characteristic but not pathognomonic
  - T1W: Reverse of normal pattern of corticomedullary SI
    - ↓ SI in renal cortices; medulla not affected, so appears as relatively ↑ SI
  - T2W: ↓ SI in renal cortices
  - Liver, spleen, and bone marrow usually have normal SI

TOP DIFFERENTIAL DIAGNOSES

- Low MR signal intensity in renal cortices
  - Malfunctioning prosthetic heart valves
  - Sickle cell anemia
  - Diffuse renal cortical calcification

PATHOLOGY

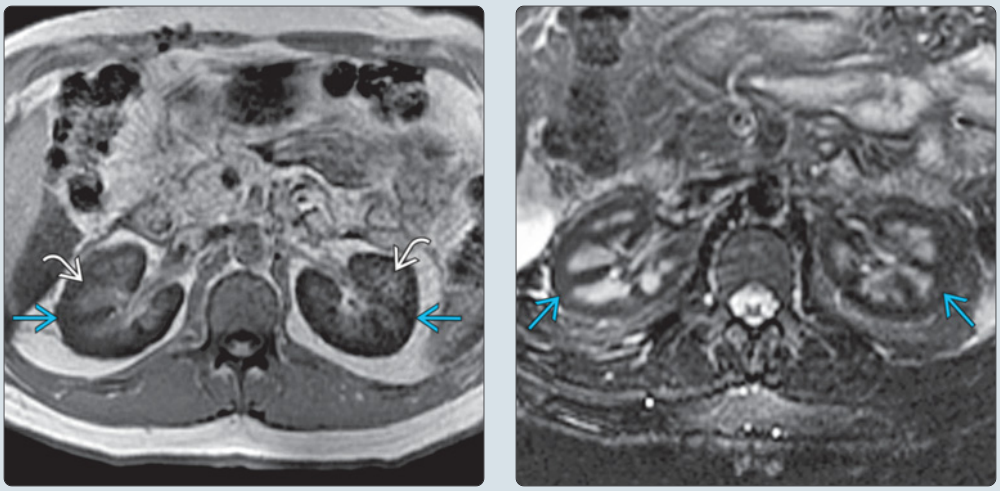
- Characterized by acute and chronic intravascular hemolysis
- Hemosiderin deposition in epithelial cells of proximal convoluted tubules in renal cortex

CLINICAL ISSUES

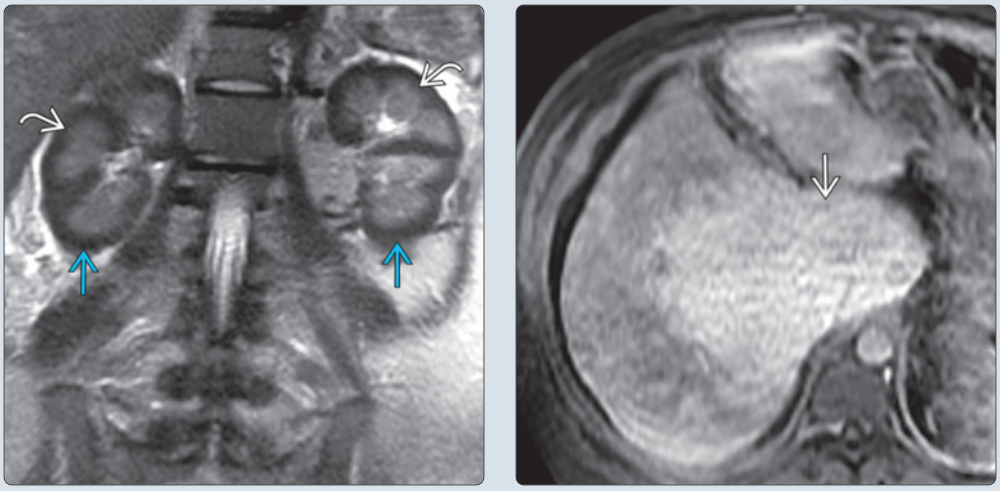
- Venous thrombosis (hepatic, sagittal sinus, mesenteric)

DIAGNOSTIC CHECKLIST

- Important to consider PNH as secondary cause of Budd-Chiari due to treatment implications



(Left) *Axial T1W in-phase MR shows low signal intensity in the renal cortices due to hemosiderin deposition from PNH. Note that the medulla is not affected by PNH and has a relatively higher signal intensity relative to the cortices. This is the reverse of the normal corticomedullary signal intensity.* **(Right)** *Axial T2W MR with fat saturation in the same patient shows low signal intensity in the renal cortices. The more profoundly low signal intensity on the T2W sequence occurs because there is more susceptibility of ferric iron with a longer TE.*



(Left) *Coronal T2 MR in the same patient demonstrates low signal intensity in the renal cortices and normal signal intensity in the medulla.* **(Right)** *Post-contrast T1W MR with fat saturation in the same patient reveals preferential early enchantment of the enlarged caudate lobe and nonvisualization of the hepatic veins due to Budd-Chiari syndrome.*

TERMINOLOGY

Abbreviations

- Paroxysmal nocturnal hemoglobinuria (PNH)

Definitions

- Rare, acquired stem cell disorder resulting in predisposition to complement-mediated hemolysis

IMAGING

General Features

- Best diagnostic clue
  - Low signal intensity in renal cortices on MR
    - Characteristic but not pathognomonic
- Location
  - Renal cortices (renal medulla is spared)

CT Findings

- No specific CT findings
  - ± ↑ attenuation of renal cortices on NECT
- CECT may show secondary signs (venous thrombosis)

MR Findings

- T1WI
  - Reverse of normal pattern of corticomedullary signal intensity (SI)
    - ↓ SI in renal cortices; medulla not affected, so appears as relatively ↑ SI
    - Lower SI with gradient and in phase T1W compared to out of phase T1W
    - Due to ↑ susceptibility from ferric iron with longer TE
- T1WI FS
  - Enhancement and excretion are not affected unless renal function is impaired
- T2WI
  - Low SI in renal cortices
  - Medulla is not affected, so appears relatively ↑ SI
- Liver, spleen, and bone marrow usually have normal SI
  - However, coexistent transfusional siderosis can lower SI

Ultrasonographic Findings

- No specific US findings for PNH

Imaging Recommendations

- Contrast enhanced MR

DIFFERENTIAL DIAGNOSIS

Malfunctioning Prosthetic Heart Valves

- Results in intravascular hemolysis

Hemolytic Anemias (e.g., Sickle Cell)

- Most are characterized by extravascular hemolysis
- Hemosiderin deposition occurs primarily in spleen (and may affect liver and bone marrow)
  - However, acute hemolytic crises or severe disease can also result in hemosiderin deposition in renal cortices
- Unlike PNH, other hemolytic anemias are congenital and usually manifest symptoms during childhood

Cortical Nephrocalcinosis

- Multifactorial process; differentiate from PNH by identifying cortical calcification with CT, US, or radiography

PATHOLOGY

General Features

- Etiology
  - Due to acute and chronic intravascular hemolysis
    - Direct release of hemoglobin in plasma
    - Resorption and storage by proximal convoluted tubule
    - Excess hemoglobin passes into urine
- Associated abnormalities
  - Venous thrombosis, bone marrow hypoplasia, infections, aplastic anemia, leukemia

Staging, Grading, & Classification

- RBC flow cytometry can be used to test for PNH phenotype

Microscopic Features

- Hemosiderin deposition in epithelial cells of proximal convoluted tubules of renal cortices

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Venous thrombosis (hepatic, mesenteric)
  - Hemoglobinuria, anemia
  - ± chronic renal failure

Demographics

- Primarily affects adults; no gender predilection

Natural History & Prognosis

- Highly variable clinical course
- Mean survival in 1 series was 10 years

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Important to consider PNH in patients with Budd-Chiari, as treatment is systemic (bone marrow transplant) rather than hepatic (surgical decompression vs. transjugular intrahepatic portosystemic shunt creation)

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Hydronephrosis

KEY FACTS

TERMINOLOGY

- Dilatation of collecting system

IMAGING

- Best diagnostic clue
  - Pyelocalyceal ± ureteral dilatation
- Best imaging tool
  - US for initial diagnosis
  - CECT for problem solving; e.g., distinguishing hydronephrosis from renal sinus cysts
    - Identification of cause of obstruction

TOP DIFFERENTIAL DIAGNOSES

- Congenital megacalyces and megaureter
- Parapelvic (peripelvic) cyst
- Pyonephrosis
- Extrarenal pelvis

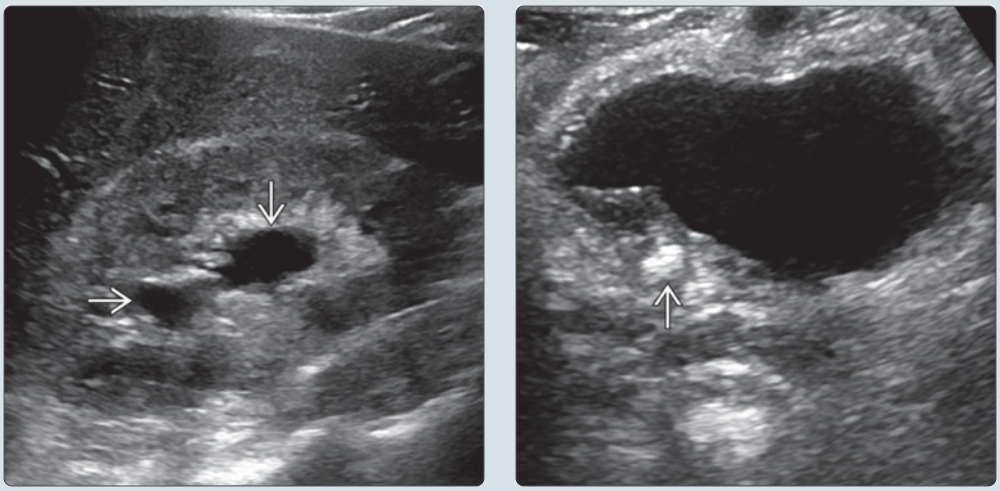
PATHOLOGY

- Etiology
  - Obstructive hydronephrosis: Partial or complete obstruction anywhere from proximal ureter to urethral meatus
    - Most common etiologies
      - Young adult: Calculi
      - Children: Ureteropelvic junction obstruction
      - Men: Benign prostatic hypertrophy (BPH)
      - Women: Gynecological cancers and pregnancy

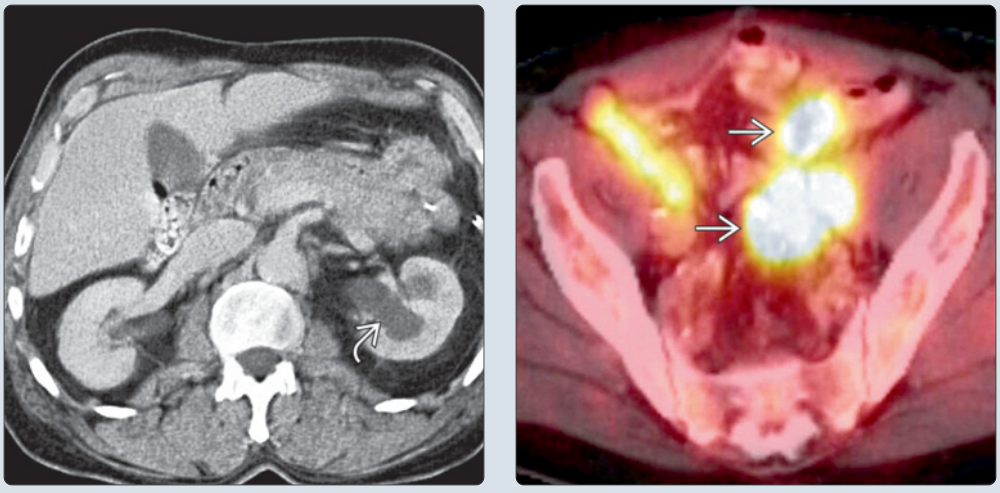
CLINICAL ISSUES

- Acute: Flank pain, ↑ pain with fluids, nausea, vomiting
- Obstruction represents 10-15% of acute kidney injury cases
- Treatment
  - Correct underlying cause (obstruction)
  - If complicated by pyonephrosis (infected collecting system with sepsis) must be treated emergently

(Left) *Longitudinal ultrasound of the right kidney shows mild hydronephrosis in a patient presenting with right flank pain.* **(Right)** *Transverse ultrasound of the pelvis in the same patient shows an echogenic stone impacted at the right ureterovesical junction causing obstructive hydronephrosis.*



(Left) *Axial CECT shows a left-sided hydronephrosis in this patient with ureteral obstruction due to recurrent colon cancer.* **(Right)** *Axial PET CT in the same patient shows the left ureter encased by a mass that is FDG-avid on PET CT, representing recurrent cancer.*



TERMINOLOGY

Definitions

- Dilatation of collecting system

IMAGING

General Features

- Best diagnostic clue
  - Pyelocalyceal ± ureteral dilatation

Imaging Recommendations

- Best imaging tool
  - US for initial diagnosis
  - CECT for problem solving; e.g., distinguishing hydronephrosis from renal sinus cysts

Ultrasonographic Findings

- Dilated pyelocalyceal system
- In obstructive hydronephrosis
  - ± absent ureteric jet at ureterovesical junction
  - ± ↑ resistive index (RI > 0.7)
- Grading
  - Mild: Mild dilatation of renal pelvis ± dilatation of calyces
  - Moderate: Moderate dilatation of renal pelvis and dilatation of calyces
  - Severe: Severe dilatation of renal pelvis and calyces and parenchymal thinning

CT Findings

- Dilatation of pyelocalyceal system
- Parenchymal thinning (chronic)
- Associated lesion causing obstruction may be seen

DIFFERENTIAL DIAGNOSIS

Congenital Megacalyces and Megaureter

- Enlargement of calyces &/or ureter in absence of obstruction or reflux

Parapelvic (Peripelvic) Cyst

- Renal sinus cysts
  - Parapelvic cysts: Renal cysts extending into renal sinus fat
  - Peripelvic cysts: Originating from renal sinus lymphatics
- May mimic hydronephrosis on US
  - Easy to diagnose on CECT in excretory phase
    - Water-attenuation cysts separate from enhanced renal collecting system

Pyonephrosis

- Infection of obstructed renal collecting system
- Ultrasound: Dilated renal pelvis with low-level echoes ± fluid-debris levels
- CT: Dilated renal pelvis with fluid-debris level ± gas, thickened urothelial lining

Extrarenal Pelvis

- Normal anatomy of calyces and papillae

PATHOLOGY

General Features

- Etiology
  - Obstructive hydronephrosis: Partial or complete obstruction anywhere from proximal ureter to urethral meatus
    - Ureter
      - Congenital: Stricture, ureterocele
      - Neoplastic: Primary and metastatic ureteral cancers
      - Inflammatory: Tuberculosis, schistosomiasis
      - Miscellaneous: Calculi, retroperitoneal fibrosis
    - Bladder and urethra
      - Congenital: Posterior urethral valves, hypospadias
      - Neoplastic: Bladder or prostate cancer
      - Miscellaneous: Benign prostatic hyperplasia, neurogenic bladder
    - Most common etiologies
      - Young adult: Calculi
      - Children: Ureteropelvic junction obstruction
      - Women: Gynecological cancers and pregnancy
      - Men: Benign prostatic hypertrophy (BPH)
  - Nonobstructive hydronephrosis
    - Diabetes insipidus
    - Vesicoureteral reflux
    - Pregnancy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute: Flank pain, ↑ pain with fluids, nausea, vomiting
  - Chronic: Asymptomatic
- Other signs/symptoms
  - Acute kidney injury

Demographics

- Gender
  - < 20 years: M = F
  - 20-60 years: F > M
  - > 60 years: M > F

Natural History & Prognosis

- Acute: Full recovery of renal function after correction of underlying lesion
  - Obstruction represents 10-15% of acute kidney injury cases
- Chronic: Loss of function is irreversible

Treatment

- Correct underlying cause (obstruction)
- If complicated by pyonephrosis (infected collecting system with sepsis) must be treated emergently

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Glomerulonephritis

KEY FACTS

TERMINOLOGY

- Inflammation and proliferation of glomerular tissue

IMAGING

- Nonspecific imaging findings
  - Enlarged, diffusely echogenic kidneys: Acute GN
  - Small kidneys: Chronic GN
- US useful to rule out hydronephrosis and to document signs of chronic vs. acute renal failure

TOP DIFFERENTIAL DIAGNOSES

- Bilateral smooth renal enlargement
  - Amyloidosis, multiple myeloma
  - Acute tubular necrosis
  - Acute interstitial nephritis
- Bilateral small kidneys with smooth contour
  - Arteriosclerosis, nephrosclerosis

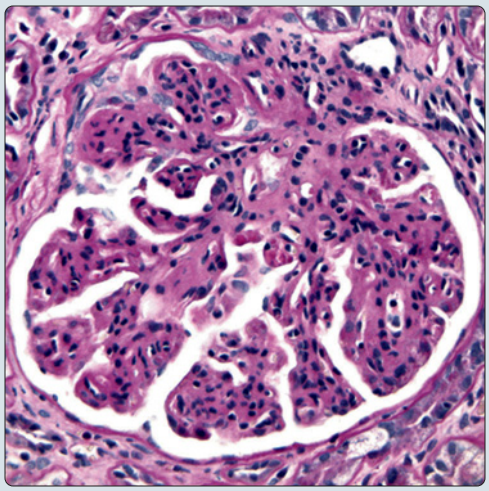
PATHOLOGY

- Primary GN: Confined to kidney
  - Systemic features secondary to renal dysfunction
  - Most are immune mediated (post *Streptococcus* infection)
- Secondary: Part of multisystem disorder
  - Many autoimmune disorders (e.g., systemic lupus)
- Associated abnormalities
  - Renal vein thrombosis
  - Hemolytic uremic syndrome

CLINICAL ISSUES

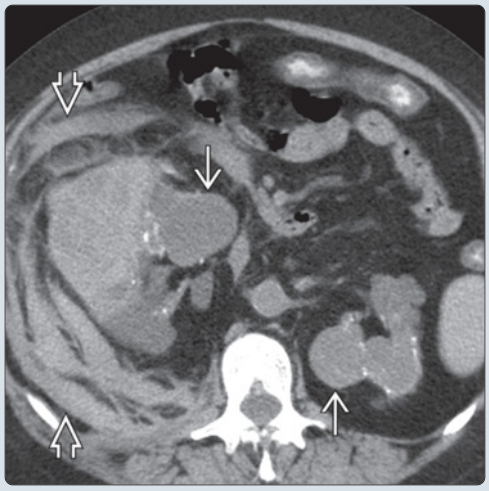
- Major presentations of glomerular disease
  - Acute nephritic or nephrotic syndrome
  - Asymptomatic urinalysis abnormalities





(Left) *Coronal oblique US shows a markedly echogenic kidney in a patient with acute renal failure due to glomerulosclerosis. There is nothing specific about the US appearance of the kidney to suggest a diagnosis other than general "acute medicorenal disease." (Courtesy H. Harvin, MD.)* **(Right)** *Micropathology shows a diffusely enlarged glomerulus with increased cellularity and thickening of the glomerular capillary walls in membranoproliferative glomerulonephritis associated with hepatitis C virus.*





(Left) *Axial NECT shows punctate parenchymal calcification in a patient on dialysis with chronic GN and secondary hyperparathyroidism. Also note the acquired cystic renal disease.* **(Right)** *Axial CECT shows kidneys that are small and nonfunctional with innumerable cysts ⇨, characteristic of acquired cystic disease of uremia. There is an extensive hemorrhage ⇨ into the right perirenal and other retroperitoneal spaces. The cause of renal failure was chronic GN.*

TERMINOLOGY

Abbreviations

- Glomerulonephritis (GN)

Definitions

- Inflammation and proliferation of glomerular tissue

IMAGING

General Features

- Size
  - Enlarged, diffusely echogenic kidneys: Acute GN
  - Small kidneys: Chronic GN
- Morphology
  - Smooth renal contour
- Imaging findings are nonspecific

CT Findings

- NECT
  - Acute GN: Normal or bilateral renal enlargement
  - Chronic GN: May have cortical calcification

Ultrasonographic Findings

- Grayscale ultrasound
  - Acute GN
    - Normal to increased renal echogenicity
    - Bilateral normal or enlarged kidneys
    - No hydronephrosis to explain renal failure
  - Chronic GN
    - Increased echogenicity
    - Small, smooth kidneys
- Color Doppler
  - May see renal vein thrombosis

Imaging Recommendations

- Protocol advice
  - US useful to rule out hydronephrosis and to document signs of chronic vs. acute renal failure

DIFFERENTIAL DIAGNOSIS

Bilateral Smooth Renal Enlargement

- Amyloidosis
- Multiple myeloma
- Acute tubular necrosis
- Acute interstitial nephritis

Bilateral Small Kidneys With Smooth Contour

- Arteriosclerosis, nephrosclerosis

PATHOLOGY

General Features

- Etiology
  - Primary GN: Confined to kidney
    - Systemic features secondary to renal dysfunction
    - Most are immune mediated
      - Common: Post *Streptococcus* infection
  - Secondary GN: Part of multisystem disorder
    - Many autoimmune disorders (e.g., systemic lupus)
- Associated abnormalities
  - Renal vein thrombosis, hemolytic uremic syndrome
  - GN is one of several intrinsic causes of acute renal failure

Staging, Grading, & Classification

- Morphologic criteria
  - Proliferative: Increased glomerular cell number
  - Crescentic: Half-moon collection of cells in Bowman space
  - Membranous: Expansion of glomerular basement membrane with immune deposits
- Extent within kidney
  - Focal: Involves < 50% of glomeruli
  - Diffuse: Involves majority of glomeruli
- Glomerular involvement
  - Global: Involves whole of glomerular tuft
  - Segmental: Involves part of glomerular tuft

Microscopic Features

- Hypercellularity
- Glomerular basement membrane thickening
- Hyalinization: Deposition of amorphous proteinaceous material
- Sclerosis: Obliteration of glomerular tuft

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Major presentations of glomerular disease
    - Acute nephritic or nephrotic syndrome
    - Asymptomatic urinalysis abnormalities
  - Acute GN: Develops over days
    - Proteinuria, hematuria, red cell casts
    - Hypertension, edema, oliguria
  - Rapidly progressive GN: Weeks → months
  - Chronic GN: Develops over months-years
- Clinical profile
  - Post *Streptococcus* infection: Follows pharyngitis or impetigo
  - Secondary: Autoimmune diseases, other entities

Demographics

- Epidemiology
  - Causes of acute renal failure
    - Prerenal (renal hypoperfusion): ~ 55%
    - Intrinsic renal disease: ~ 40%
    - Postrenal (obstruction): ~ 5%

Natural History & Prognosis

- May progress to end-stage renal disease

Treatment

- Immunosuppressive therapy
- Treatment of underlying systemic disorder

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Acute Tubular Necrosis

KEY FACTS

TERMINOLOGY

- Renal cause of acute kidney injury (AKI) characterized by tubular epithelial cell damage from toxins or ischemia

IMAGING

- Ultrasound
  - To assess renal morphology and echogenicity
  - Exclude other causes of AKI (i.e., obstruction)
  - Renal length: Normal or ↑
  - Renal echogenicity: Normal or ↑
- NECT
  - Persistent patchy nephrogram (following prior contrast administration)

TOP DIFFERENTIAL DIAGNOSES

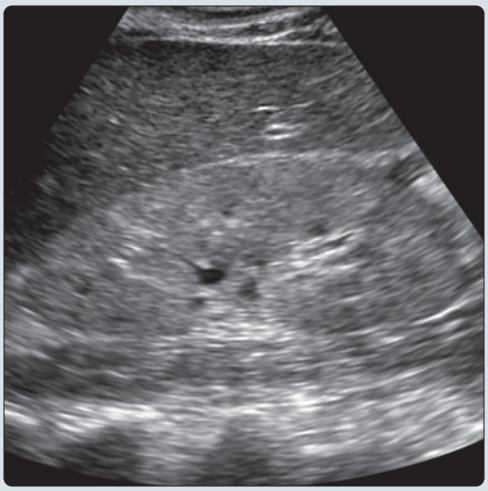
- Acute glomerulonephritis
- HIV nephropathy
- Acute interstitial nephritis

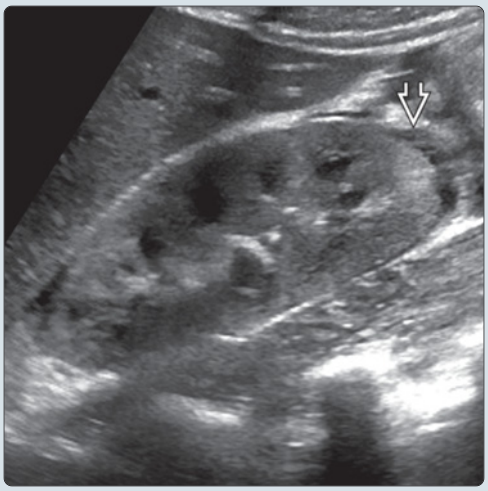
PATHOLOGY

- Ischemia: Hypotension (most common cause)
- Toxins
  - Exogenous: Iodinated contrast media, antibiotics, chemotherapeutic drugs, organic solvents, heavy metals
  - Endogenous: Hemolysis, rhabdomyolysis, uric acid, oxalate

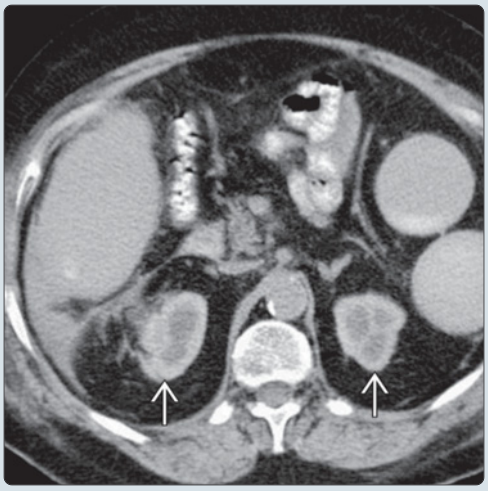
CLINICAL ISSUES

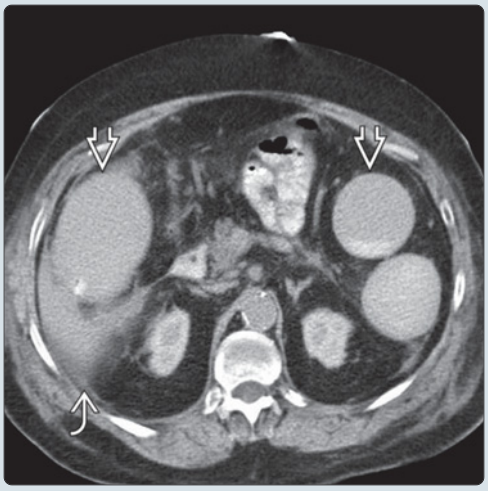
- Most common form of reversible AKI
- Renal failure develops quickly and lasts 10-20 days
- Mortality rates ↑ in ICU patients (50-80%)
- Presentation
  - Oliguria/anuria, water/salt overload, azotemia, ↑ serum potassium, metabolic acidosis
- Treatment
  - Correction of volume depletion or overload, electrolyte imbalance, metabolic acidosis
  - Dialysis





(Left) *Ultrasound of a patient with acute kidney injury (AKI) shows an enlarged, echogenic right kidney; findings commonly seen in the setting of medical renal disease. Note the absence of collecting system dilatation.* **(Right)** *Ultrasound of a patient with acute tubular necrosis (ATN) following cardiac arrest shows an enlarged and echogenic right kidney. Note an extracapsular hypoechoic rim ("kidney sweat") ⇨, a sign often seen in patients with renal failure.*





(Left) *Axial NECT in a patient with shock following myocardial infarction shows persistent nephrograms ⇨ many hours after coronary angiography, indicating ATN.* **(Right)** *Axial NECT in the same patient shows colon distended with fluid and diluted enteric contrast medium ⇨. Moderate ascites ⇨ is also present measuring 45 HU in attenuation. This is an example of opacification of ascites by the IV administration of iodinated contrast, a process more marked in patients with renal failure.*

TERMINOLOGY

Abbreviations

- Acute tubular necrosis (ATN)
- Acute kidney injury (AKI)
  - Preferred term over acute renal failure (ARF)

Definitions

- Renal cause of AKI characterized by tubular epithelial cell damage from toxins or ischemia

IMAGING

Imaging Recommendations

- Best imaging tool
  - Ultrasound
    - To assess renal morphology and echogenicity
    - To exclude other causes of AKI (i.e., obstruction)

Ultrasonographic Findings

- Grayscale ultrasound
  - Morphology
    - Renal length: Normal or ↑
    - Cortical thickness: Normal or ↑
  - Echogenicity
    - Normal or ↑
    - Usually normal in ischemic ATN, and ↑ in toxic ATN
  - Perirenal hypoechoic rim ("kidney sweat")
- Color Doppler
  - Resistive index: Normal or ↑ (> 0.7)

CT Findings

- NECT
  - Persistent patchy nephrogram (following prior contrast administration)

MR Findings

- Functional renal imaging
  - Under investigation
  - Blood oxygen level dependent (BOLD) MR for renal oxygenation
  - Arterial spin labeling (ASL) MR for renal perfusion

DIFFERENTIAL DIAGNOSIS

Glomerulonephritis

- Normal to increased renal echogenicity
- Normal or enlarged kidneys
- History, labs, and biopsy distinguish it from ATN

HIV Nephropathy

- HIV-1 infection associated with focal segmental glomerulosclerosis
- Enlarged, echogenic kidneys on US

Acute Interstitial Nephritis

- History of drug hypersensitivity to antibiotics or other medications
- Normal or enlarged echogenic kidneys
- White blood cell casts on urinalysis

PATHOLOGY

General Features

- Etiology
  - Ischemia: Hypotension (most common cause), hypovolemia, renal vasoconstriction, DIC
  - Toxins
    - Exogenous: Iodinated contrast media, antibiotics, chemotherapeutic drugs, organic solvents, heavy metals
    - Endogenous: Hemolysis, rhabdomyolysis, uric acid, oxalate
  - Sustained prerenal azotemia is most common predisposing factor for acute tubular necrosis

Microscopic Features

- Multiple areas of focal tubular epithelial necrosis with large skip areas, rupture of basement membrane, tubular occlusion by casts, interstitial edema
- In toxic ATN, necrosis most obvious in proximal convoluted tubules

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Oliguria/anuria, water/salt overload, azotemia, ↑ serum potassium, metabolic acidosis

Demographics

- Epidemiology
  - Most common form of reversible AKI

Natural History & Prognosis

- Renal failure develops quickly and lasts 10-20 days
- Mortality rates ↑ in ICU patients (50-80%)

Treatment

- Correction of volume depletion (IV fluid) or overload (diuretics)
- Correction of electrolyte imbalance and metabolic acidosis
- Dialysis

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Renal Cortical Necrosis

KEY FACTS

TERMINOLOGY

- Rare form of acute renal injury characterized by necrosis of renal cortex

IMAGING

- NECT: Calcifications in renal cortex and columns of Bertin (chronic cortical necrosis)
- CECT
  - Acute setting: ↓ enhancement of renal cortex
  - Preserved enhancement of thin rim of subcapsular cortical tissue
  - Preserved medullary enhancement
  - ↓ excretion of contrast media into collecting system
- MR: ↓ signal intensity inner cortex and columns of Bertin

TOP DIFFERENTIAL DIAGNOSES

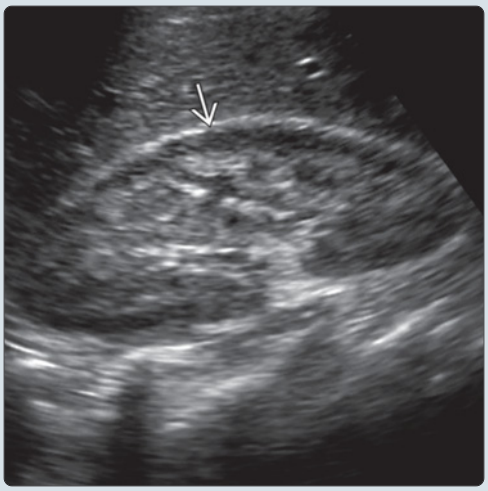
- Acute tubular necrosis
- Renal infarction
- Renal vein thrombosis

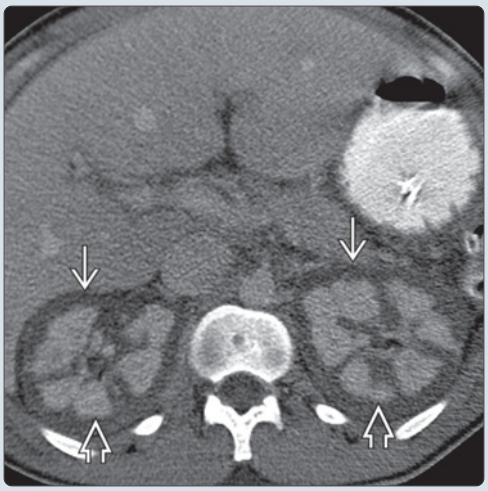
PATHOLOGY

- Obstetrical complication (> 50% of cases)
  - Abruptio placentae, septic abortion, severe eclampsia
- Other causes: Hemolytic uremic syndrome, disseminated intravascular coagulopathy, shock, sickle cell anemia, renal allograft rejection

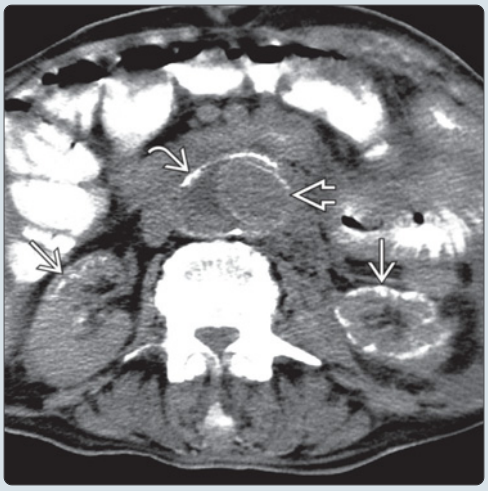
CLINICAL ISSUES

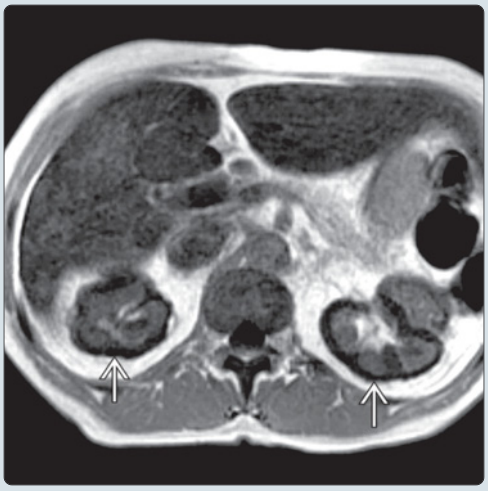
- Rare cause of acute kidney injury in developed countries
  - Incidence higher in developing countries
- Symptoms/signs: Prolonged anuria/oliguria; azotemia, hyperkalemia, metabolic acidosis
- Poor prognosis
  - Evolution toward chronic renal failure and end-stage renal disease
  - Mortality > 50% if untreated
- Treatment
  - Dialysis; transplantation





(Left) *Longitudinal ultrasound of the right kidney in a patient with sickle cell anemia developing renal cortical necrosis shows decreased renal cortical echogenicity ➡ compared to medulla.* **(Right)** *Axial CECT in the same patient shows bilateral lack of renal cortical enhancement ➡ and preserved medullary enhancement ➡, compatible with bilateral renal cortical necrosis.*





(Left) *Axial NECT shows subcapsular calcifications ➡ in both kidneys, indicative of renal cortical necrosis following shock caused by a ruptured abdominal aortic aneurysm. Note the aortic wall ➡ and endograft ➡.* **(Right)** *Axial T1WI GRE in a patient with sickle cell anemia shows reversal of the usual corticomedullary pattern, with the cortex ➡ being hypointense to the medulla. This may be due to hemosiderin deposition or cortical necrosis, both of which can be seen in patients with sickle cell disease.*

TERMINOLOGY

Definitions

- Rare form of acute renal injury characterized by ischemic necrosis of renal cortex

IMAGING

General Features

- Best diagnostic clue
  - CECT: ↓ enhancement of renal cortex with preserved enhancement of medulla and thin rim of subcapsular cortical tissue best shows pathophysiology
- Location
  - Usually extensive and bilateral

Imaging Recommendations

- Best imaging tool
  - Sonography for exclusion of collecting system dilatation
  - Characteristic ultrasound appearance (hypoechoic cortical rim) suggests acute cortical necrosis in appropriate clinical context

Radiographic Findings

- Radiography
  - Cortical calcification: Dual linear opacities paralleling corticomedullary line ("tram line" sign), which is usually seen after 4 weeks

CT Findings

- NECT
  - Calcifications in renal cortex and columns of Bertin may indicate prior cortical necrosis
- CECT
  - Acute necrosis: ↓ enhancement of renal cortex
  - Preserved enhancement of thin rim of subcapsular cortical tissue due to separate capsular blood supply
  - Preserved medullary enhancement
  - ↓ excretion of contrast media into collecting system

MR Findings

- T1WI
  - ↓ signal intensity inner cortex and columns of Bertin
- T2WI
  - ↓ signal intensity inner cortex and columns of Bertin
  - ↑ signal intensity subcapsular rim

Ultrasonographic Findings

- Acute cortical necrosis: ↓ renal cortical echogenicity
- In later stages, diffuse ↑ cortical echogenicity and acoustic shadow due to cortical calcification

DIFFERENTIAL DIAGNOSIS

Acute Tubular Necrosis

- Persistent nephrogram on NECT (after prior contrast administration)
- US: ↑ renal size and echogenicity

Renal Infarction

- CECT: Nonenhancing wedge-shaped area of renal parenchyma with enhancing capsular rim

Renal Vein Thrombosis

- Occluded renal vein with renal enlargement and delayed renal function
- MR: Low signal intensity in outer medulla

PATHOLOGY

General Features

- Etiology
  - Obstetrical complication (> 50% of cases)
    - Abruptio placentae, septic abortion, eclampsia
  - Hemolytic uremic syndrome, disseminated intravascular coagulopathy, shock, sickle cell anemia, renal allograft rejection

Microscopic Features

- Complete cortical necrosis
  - Global ischemic necrosis of cortex with preservation of thin subcapsular rim of tissue
- Patchy cortical necrosis
  - Multiple contiguous areas of necrosis involving up to 1/2 of cortex

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Prolonged anuria/oliguria; azotemia, hyperkalemia, metabolic acidosis

Demographics

- Epidemiology
  - Rare cause of acute kidney injury (AKI) in developed countries
    - 2% of all cases of AKI; 15-20% of AKI in pregnancy
  - Incidence higher in developing countries
    - 7% of all cases of AKI
  - More common in infants and young women
  - Obstetric complications account for > 50% of cases

Natural History & Prognosis

- Poor prognosis
- Evolution toward chronic renal failure and end-stage renal disease
- Mortality > 50% if untreated

Treatment

- Treatment of underlying cause
- Dialysis; transplantation

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Renal Papillary Necrosis

KEY FACTS

TERMINOLOGY

- Necrosis of renal papilla within medulla secondary to interstitial nephritis or ischemia

IMAGING

- Bilateral disease: Analgesics and diabetes
- Unilateral disease: Obstruction, infection, venous thrombus
- Triangular or bulbous cavitation adjacent to calyx on IVP, CT urogram, or retrograde pyelogram
- Subtle streak of contrast extending from fornix parallel to long axis of papilla
- Calyx
  - Club-shaped or saccular (sloughed papilla)
  - Widened fornix (necrotic shrinkage of papilla)
- Sloughed papilla
  - Triangular filling defect within calyx, pelvis, ureter
  - May have ring calcification

TOP DIFFERENTIAL DIAGNOSES

- Hydronephrosis
- Medullary sponge kidney
- Congenital megacalyces and megaureter

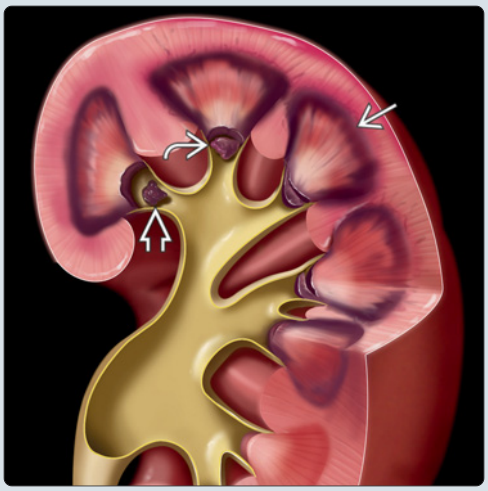
PATHOLOGY

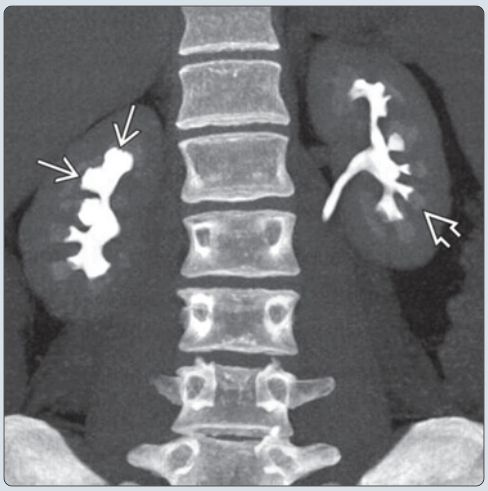
- Analgesic nephropathy, diabetes, sickle cell
- UTI and obstruction, TB, kidney transplantation, alcoholism

CLINICAL ISSUES

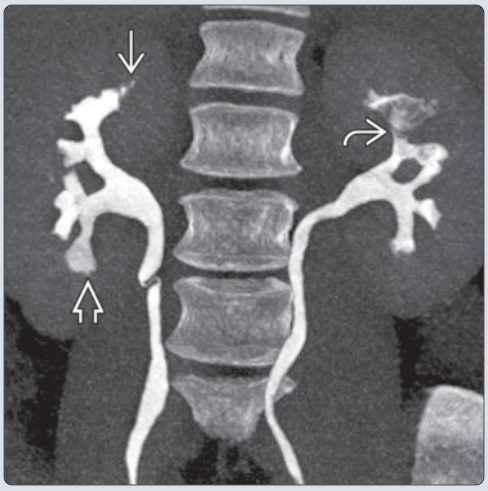
- Complications
  - Obstruction, infection, renal failure, transitional cell carcinoma
- Prognosis
  - Early stage (good), advanced stage (poor)

(Left) *Graphic shows ischemic injury of some of the renal pyramids with sloughing of several papillae, 1 of which lies free within the collecting system.* **(Right)** *Coronal CECT urography in a 33-year-old man with history of IV drug use shows loss of the normal renal pyramids in the upper pole of the right kidney with blunted, rounded calyces. Compare these with the cupped, normal calyces on the left and the pyramids, with their rays of contrast-opacified tubules.*





(Left) *Coronal CECT urography in a 46-year-old woman with sickle cell disease shows clubbing of the calyces, contrast pooling into the site of sloughed papillae, and amorphous debris within the upper pole calyces of the left kidney. These are all classic findings indicating papillary necrosis.* **(Right)** *Coronal CECT MIP in the same patient shows 1 of the sloughed papillae along with the more amorphous debris within the collecting system.*





TERMINOLOGY

Abbreviations

- Renal papillary necrosis (RPN)

Definitions

- Necrosis of renal papilla within medulla secondary to interstitial nephritis or ischemia

IMAGING

General Features

- Best diagnostic clue
  - Triangular or bulbous cavitation adjacent to calyx on IVP, CT urogram, or retrograde pyelogram
- Location
  - Bilateral: Analgesics and diabetes
  - Unilateral: Obstruction, infection, venous thrombus
- Morphology
  - Papillary swelling, tract formation, cavitation, and sloughing

IVP, CT Urography Findings

- Subtle streak of contrast extending from fornix parallel to long axis of papilla
- Triangular or bulbous cavitation of papilla
- Widened fornix (necrotic shrinkage of papilla)
- Calyx: Club-shaped or saccular (sloughed papilla)
- Sloughed papilla: Triangular filling defect within calyx, pelvis, ureter
  - Sloughed papilla may have ring calcification

CT Findings

- Normal, enlarged, or small contracted kidneys
- Ring-shaped medullary calcification
- Hematoma, lobar infarct, scarring (sickle cell)
- Contrast-filled clefts in renal parenchyma
- Filling defects: Renal pelvis/ureter (sloughed papillae)

Ultrasonographic Findings

- Grayscale ultrasound
  - Early stage
    - Necrotic renal papillae: Seen as echogenic foci
  - Advanced stage
    - Single or multiple cystic cavities in medullary pyramids continuous with calyces ± calcification

DIFFERENTIAL DIAGNOSIS

Hydronephrosis

- Dilated pelvicalyceal system and ureter; ↓ renal function

Medullary Sponge Kidney

- Multiple small cystic cavities or tubular ectasia; medullary nephrocalcinosis
- Excretory or CT urography
  - Linear densities ("brush" appearance) in pyramids

Congenital Megacalyces and Megaureter

- Large and wide calyceal system with normal renal morphology and function

PATHOLOGY

General Features

- Etiology
  - Analgesic nephropathy, diabetes, sickle cell disease
  - UTI and obstruction, TB, kidney transplantation, alcoholism

Gross Pathologic & Surgical Features

- Mild cases: Normal or enlarged kidneys
- Severe cases: Small, smooth or scarred

Microscopic Features

- Analgesics: RPN, centrilobar cortex atrophy, septal cortex hypertrophy
- Sickle cell disease: Vessel occlusion, lobar infarcts, tubular obliteration, fibrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Flank pain, dysuria, fever, ureteral colic
  - Pyuria, hematuria, acute oliguric renal failure
- Lab: ± ↑ WBC, proteinuria, impaired function

Demographics

- Middle-aged women (analgesic nephropathy)

Natural History & Prognosis

- Complications: Obstruction, infection, renal failure, transitional cell carcinoma
- Prognosis: Early stage (good), advanced stage (poor)

Treatment

- Early stage: Symptomatic treatment
- Advanced stage (obstruction, infection, failure)
  - Ureteral stent, surgical treatment

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Club-shaped calyx, triangular cavitation of papillae, or filling defect of sloughed papillae with ring shadow

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HIV Nephropathy

KEY FACTS

TERMINOLOGY

- HIV-associated nephropathy (HIVAN)
- Progressive chronic renal disease in patients with HIV infection histologically characterized by focal and segmental glomerulosclerosis (collapsing glomerulopathy)

IMAGING

- Ultrasound
  - Increased renal echogenicity
  - Decreased corticomedullary differentiation
  - Urothelial thickening
  - Decreased renal sinus fat
  - No hydronephrosis

TOP DIFFERENTIAL DIAGNOSES

- Glomerulonephritis (GN)
- Acute tubular necrosis (ATN)
- Opportunistic renal infections

PATHOLOGY

- Focal and segmental glomerulosclerosis (collapsing glomerulopathy)

CLINICAL ISSUES

- Proteinuria; progressive renal failure
- Higher incidence in African Americans (> 90% of reported cases)
- ↓ incidence after introduction of combination antiretroviral therapy (cART)

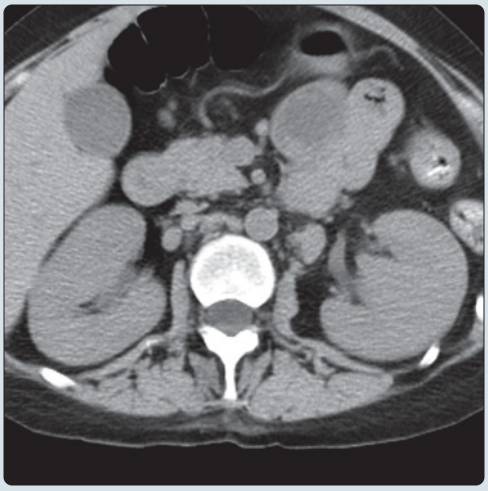
DIAGNOSTIC CHECKLIST

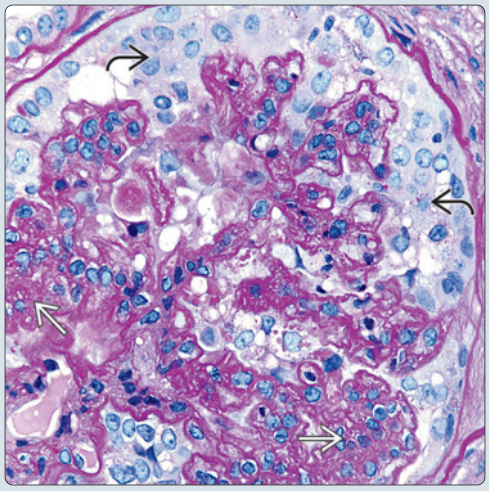
- Final diagnosis is made at histology
- Other causes of renal disease in HIV-infected patients, including
  - Antiretroviral nephrotoxicity
  - Opportunistic infections
  - Urolithiasis
  - Neoplasms





(Left) *Longitudinal ultrasound of the left kidney in patient with biopsy-proven HIV-associated nephropathy (HIVAN) shows an enlarged, echogenic kidney with no hydronephrosis.* **(Right)** *Longitudinal ultrasound of the right kidney in the same patient reveals an affected enlarged echogenic kidney. HIVAN typically affects both kidneys. Note the increased echogenicity of the renal parenchyma compared with the adjacent liver and the decreased corticomedullary differentiation.*





(Left) *Axial NECT of the abdomen in an HIV-positive patient shows enlarged kidneys with loss of corticomedullary differentiation and decreased pelvic sinus fat suggesting HIVAN.* **(Right)** *Classic appearance of HIV-associated nephropathy (HIVAN) with glomerul collapse ➡ and proliferation of overlying podocytes ⇨ is well demonstrated by periodic acid-shift (PAS) stain in this HIV-positive man. (From DP: Kidney.)*

TERMINOLOGY

Abbreviations

- HIV-associated nephropathy (HIVAN)

Definitions

- Progressive chronic renal disease in patients with HIV infection histologically characterized by focal and segmental glomerulosclerosis (collapsing glomerulopathy)

IMAGING

General Features

- Best diagnostic clue
  - Ultrasound: Enlarged, echogenic kidneys
- Location
  - Bilateral
- Size
  - Normal or increased

CT Findings

- NECT
  - Loss of corticomedullary differentiation
- CECT
  - Striated nephrogram

MR Findings

- Nonspecific imaging appearance
- Loss of corticomedullary differentiation

Ultrasonographic Findings

- Grayscale ultrasound
  - Increased renal echogenicity
  - Decreased corticomedullary differentiation
  - Urothelial thickening
  - Decreased renal sinus fat
  - No hydronephrosis

Imaging Recommendations

- Best imaging tool
  - Ultrasound

DIFFERENTIAL DIAGNOSIS

Glomerulonephritis (GN)

- Normal to increased renal echogenicity
- Normal or enlarged kidneys in acute GN
- Small kidneys in chronic GN

Acute Tubular Necrosis (ATN)

- Most common cause of reversible acute renal failure
- Ultrasound appearance of hypotension-related ATN is often normal
- Ultrasound appearance of ATN caused by drugs, heavy metals, or solvent exposure results in enlarged echogenic kidneys

Opportunistic Renal Infections

- Pneumocystis carinii
  - Most common opportunistic infection in HIV patients
  - Punctate cortical calcifications

PATHOLOGY

General Features

- Etiology
  - HIV-1 infection

Gross Pathologic & Surgical Features

- Focal and segmental glomerulosclerosis (collapsing glomerulopathy)
- Dilated tubules, interstitial inflammation

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Proteinuria; progressive renal failure
- Other signs/symptoms
  - Absence of significant edema and hypertension

Demographics

- Ethnicity
  - Higher incidence in African Americans (> 90% of reported cases)
- Epidemiology
  - ↓ incidence after introduction of combination antiretroviral therapy (cART)

Natural History & Prognosis

- Common evolution to end-stage renal disease

Treatment

- Antiretroviral therapy
- Angiotensin-converting enzyme (ACE) inhibitors, corticosteroids
- Transplantation

DIAGNOSTIC CHECKLIST

Consider

- Final diagnosis is made at histology
- Other causes of renal disease in HIV-infected patients, including
  - Antiretroviral nephrotoxicity
  - Opportunistic infections
  - Urolithiasis
  - Neoplasms

Image Interpretation Pearls

- Enlarged echogenic kidneys at ultrasound

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Chronic Renal Failure

KEY FACTS

TERMINOLOGY

- Chronic kidney disease (CKD)
- Kidney damage or decreased renal function for ≥ 3 months

IMAGING

- Ultrasound
  - ↓ renal length
  - ↓ cortical thickness
  - ↑ echogenicity
  - Multiple cysts (uremic cystic disease)
- NECT
  - Small kidneys; cortical thinning
  - Dilatation of collecting system if cause is obstruction

TOP DIFFERENTIAL DIAGNOSES

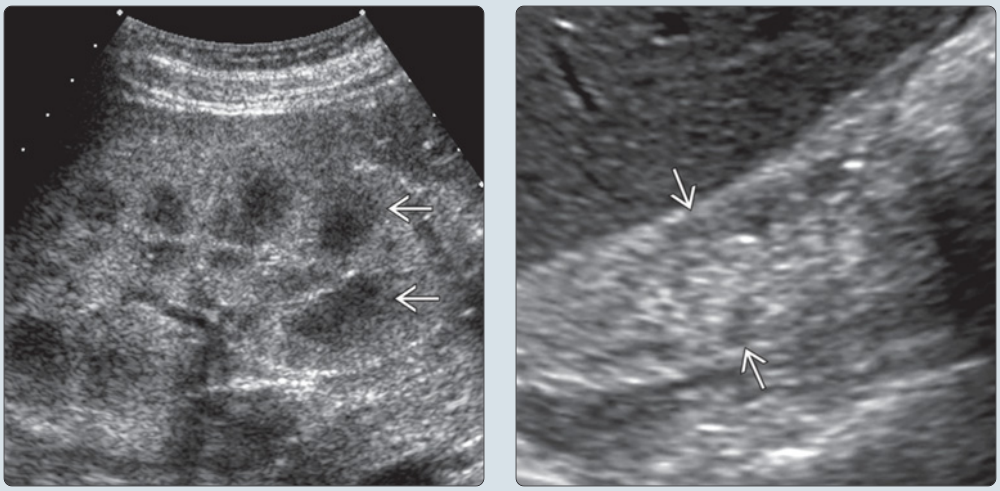
- Acute tubular necrosis
- Renal cortical necrosis

PATHOLOGY

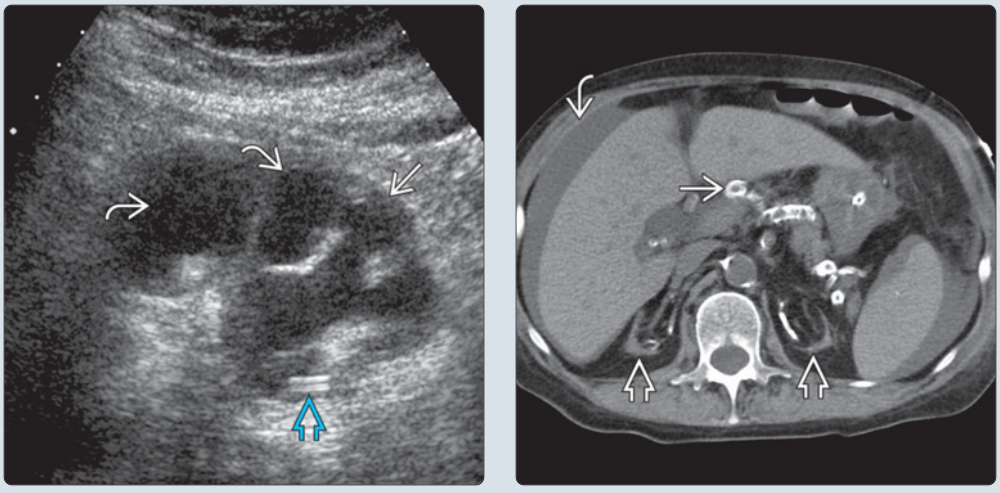
- Etiology
  - Diabetic nephropathy, chronic glomerulonephritis, hypertensive nephropathy, polycystic renal disease, chronic pyelonephritis, renal calculi
- Severity stratified by glomerular filtration rate (GFR)
  - Stage 1: GFR ≥ 90 mL/min/1.73 m²
  - Stage 2: GFR 60-89 ml/min/1.73 m²
  - Stage 3: GFR 30-59 mL/min/1.73 m²
  - Stage 4: GFR 15-29 mL/min/1.73 m²
  - Stage 5: GFR < 15 mL/min/1.73 m²

CLINICAL ISSUES

- Incidence in the USA: 449 cases per million population
- Symptoms and signs
  - Hypertension, fluid overload, malaise, pericarditis, encephalopathy
- Treatment of underlying etiologies, dialysis, transplantation



(Left) *Longitudinal grayscale ultrasound of end-stage diabetic nephropathy in a 74-year-old man presenting with type 2 diabetes and uremia demonstrates echogenic kidneys with prominent hypoechoic pyramids.* **(Right)** *Longitudinal ultrasound of the right kidney shows a small kidney with decreased cortical thickness and increased echogenicity compatible with chronic kidney injury.*



(Left) *Longitudinal grayscale ultrasound of end-stage hydronephrosis in an 82-year-old man with prostate cancer and longstanding ureteral obstruction shows marked cortical thinning and hydronephrosis. Note the ureteral stent in the proximal renal pelvis.* **(Right)** *Axial NECT shows shrunken, end-stage kidneys, with calcifications of large- and medium-sized abdominal arteries, findings almost unique to patients with diabetic vasculopathy. Note ascites, a complication of chronic renal failure.*

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TERMINOLOGY

Synonyms

- Chronic kidney disease (CKD)
- End-stage renal disease (ESRD)

Definitions

- Kidney damage or decreased renal function for ≥ 3 months

IMAGING

General Features

- Best diagnostic clue
  - Small (< 9 cm) echogenic kidneys at ultrasound

Imaging Recommendations

- Best imaging tool
  - Ultrasound

Ultrasonographic Findings

- Grayscale ultrasound
  - Morphology
    - ↓ renal length
    - ↓ cortical thickness
  - Echogenicity
    - ↑; may be similar to surrounding fat
    - Prominent hypoechoic pyramids
  - Multiple cysts (uremic cystic disease)
  - Dilatation of the collecting system if obstructive uropathy
- Pulsed Doppler
  - Increased intrarenal (segmental, interlobar) arterial resistive index (RI)
    - Native kidney threshold: > 0.7
    - Nonspecific finding of renal dysfunction
    - Elevated RI largely due to decreased tissue compliance (not renal vascular resistance)
    - Elevated pulse pressure also contributes

CT Findings

- Small kidneys
- Cortical thinning
- Dilatation of the collecting system if obstructive uropathy

DIFFERENTIAL DIAGNOSIS

Acute Tubular Necrosis

- Renal cause of acute kidney injury characterized by tubular epithelial cell damage from toxins or ischemia
- Ultrasound
  - Normal or ↑ renal length
  - Normal or ↑ renal echogenicity
  - Elevated resistive indices
  - No hydronephrosis
- NECT
  - Persistent patchy nephrogram (following prior contrast administration)

Renal Cortical Necrosis

- CECT: No cortical enhancement with preservation of subcapsular zone
- ↓ cortical echogenicity

PATHOLOGY

General Features

- Etiology
  - Diabetic nephropathy, chronic glomerulonephritis, hypertensive nephropathy, polycystic renal disease, chronic pyelonephritis, renal calculi
- Associated abnormalities
  - Congestive heart failure, arrhythmias due to hyperkalemic pericarditis, secondary hyperparathyroidism, renal osteodystrophy

Staging, Grading, & Classification

- Severity stratified by glomerular filtration rate (GFR)
  - Stage 1: GFR ≥ 90 mL/min/1.73 m²
  - Stage 2: GFR 60-89 ml/min/1.73 m²
  - Stage 3: GFR 30-59 mL/min/1.73 m²
  - Stage 4: GFR 15-29 mL/min/1.73 m²
  - Stage 5: GFR < 15 mL/min/1.73 m²

Gross Pathologic & Surgical Features

- Small, atrophic kidneys; marked hydronephrosis if obstruction

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Hypertension, fluid overload
- Other signs/symptoms
  - Symptoms of uremia: Malaise, pericarditis, encephalopathy

Demographics

- Epidemiology
  - Incidence in the USA: 449 cases per million population

Natural History & Prognosis

- Poor prognosis unless continued dialysis or transplantation

Treatment

- Treat underlying conditions, such as diabetes or hypertension; hemodialysis
- Transplantation

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Small (< 9 cm), echogenic kidneys on US ("medical renal disease," "renal parenchymal disease")

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Renal Lipomatosis

KEY FACTS

TERMINOLOGY

- Fatty tissue proliferation in renal sinus (renal sinus lipomatosis) that in extreme cases can replace renal parenchyma (renal replacement lipomatosis)

IMAGING

- Best imaging tool
  - CECT
- Renal sinus lipomatosis
  - Bilateral process, mild associated atrophy
  - Enlarged sinus with fat density (-30 to -90 HU)
- Replacement lipomatosis
  - Unilateral process
  - Enlarged kidney with reniform shape
  - Focal or diffuse renal parenchymal atrophy
  - Fibrofatty proliferation in renal sinus and perirenal space
  - Centrally located calculus in 70%
  - ± hydronephrosis

TOP DIFFERENTIAL DIAGNOSES

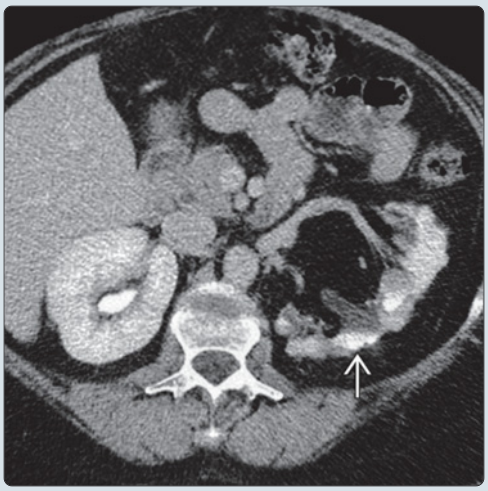
- Xanthogranulomatous pyelonephritis
  - Signs, symptoms, and pathology overlap with replacement lipomatosis
  - Often extends into tissues of posterior abdominal wall
- Renal angiomyolipoma
  - Spherical mass of mostly fat and blood vessels
  - Mass effect on collecting system
  - Normal renal function; no stones or inflammation

CLINICAL ISSUES

- Renal sinus lipomatosis
  - Normal finding in elderly and obese patients with good prognosis
- Renal replacement lipomatosis
  - Worse prognosis as it is associated with underlying chronic inflammation
  - May require nephrectomy

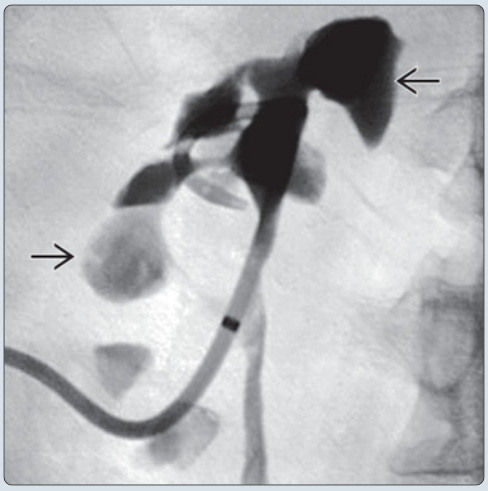
(Left) *Longitudinal ultrasound of the right kidney shows mild cortical thinning and proliferation of sinus fat compatible with renal sinus lipomatosis.* **(Right)** *Axial CECT in a patient with renal replacement lipomatosis shows left side renal cortical atrophy, dilated calyces ⇨, and markedly proliferated renal sinus fat. The caliectasis, renal scarring, and lipomatosis were the result of multiple, chronic episodes of renal and ureteral calculi and infection.*





(Left) *Axial NECT in a patient with replacement lipomatosis shows a small left kidney, a large renal stone ⇨, and dilated calyces with marked cortical thinning and fibrofatty proliferation in the renal sinus and perirenal space.* **(Right)** *Percutaneous nephrostogram in the same patient shows dilated calyces ⇨. Renal replacement lipomatosis was the result of chronic partial obstruction by the large stone and chronic infection and inflammation of the kidney.*





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TERMINOLOGY

Synonyms

- Renal sinus lipomatosis (RSL), renal replacement lipomatosis (RRL)

Definitions

- Fatty tissue proliferation in renal sinus (renal sinus lipomatosis) that in extreme cases can replace renal parenchyma (renal replacement lipomatosis)

IMAGING

General Features

- Best diagnostic clue
  - Fatty infiltration of renal sinus and thin rim of renal parenchyma
- Morphology
  - Bilateral and symmetrical involvement in RSL
  - Involvement is usually unilateral and asymmetrical in RRL
  - Uncommonly, RRL may be focal process

Imaging Recommendations

- Best imaging tool
  - CECT

Radiographic Findings

- Radiography
  - Large or staghorn calculus associated with RRL
- IVP
  - Renal sinus lipomatosis
    - Elongation and stretching of infundibula; no caliectasis
    - Enlarged radiolucent renal sinus
  - Renal replacement lipomatosis
    - Enlarged radiolucent sinus; stretched infundibula
    - Thin rim of parenchyma with poor enhancement and excretion

CT Findings

- Renal sinus lipomatosis
  - Bilateral process; mild atrophy
  - Enlarged sinus with fat attenuation (-30 to -90 HU)
- Replacement lipomatosis
  - Unilateral process
  - Enlarged kidney with reniform shape
  - Severe parenchymal atrophy
  - Fibrofatty proliferation in sinus and perirenal space
  - Centrally located calculus in 70%
  - ± hydronephrosis

MR Findings

- Enlargement of renal sinus with fat signal on T1WI and T2WI
  - Signal suppression on fat-saturation imaging

Ultrasonographic Findings

- Expansion of echogenic central sinus complex ± calculus
- Reniform shape

DIFFERENTIAL DIAGNOSIS

Xanthogranulomatous Pyelonephritis

- Chronic infection of kidney and surrounding tissue
- Destroyed parenchyma, replaced by lipid-laden macrophages
- Often extends into tissues of posterior abdominal wall

Renal Angiomyolipoma

- Benign renal tumor composed of abnormal blood vessels, smooth muscle, and fatty components
- Normal renal function, no stones or inflammation
- Mass effect on collecting system

PATHOLOGY

General Features

- Etiology
  - RSL: Normal finding in aged and obese
    - Can also be seen with endogenous or exogenous steroid excess
  - RRL: Sequela of severe renal atrophy or inflammation, usually due to chronic calculous disease
    - Chronic inflammation → renal parenchymal loss → replacement by fibrofatty tissue
- Associated abnormalities
  - Hydronephrosis, pyonephrosis (mostly in RRL)
  - Calculus is associated with 70% of cases of RRL

Gross Pathologic & Surgical Features

- RRL: Enlarged kidney with proliferation of fibrofatty tissue and loss of parenchyma

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Patients with RSL are typically asymptomatic
  - Patients with RRL are often symptomatic with chronic inflammation and decreased renal function

Demographics

- Age
  - RSL is common in elderly
  - RRL can be seen in younger patients but commonly occur in 6th-7th decades
- Epidemiology
  - Prevalence is 1%, higher in elderly

Treatment

- RRL is associated with ↓ renal function and may require nephrectomy

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KEY FACTS

TERMINOLOGY

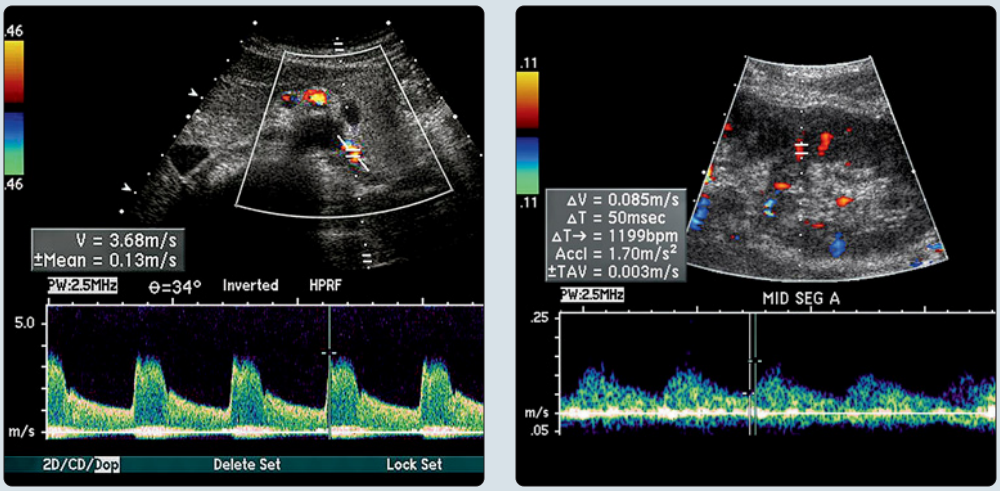
- Renal artery stenosis (RAS)

IMAGING

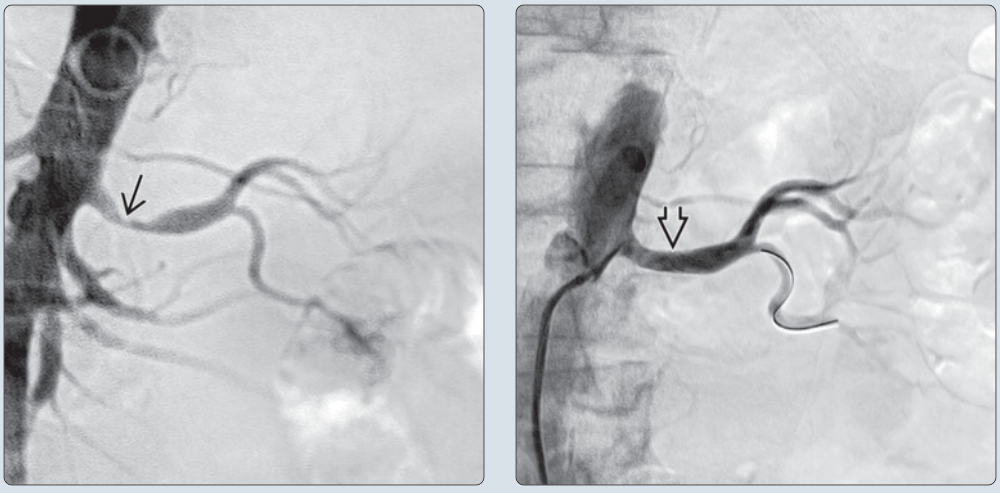
- Best diagnostic clue: Focal or segmental luminal narrowing of renal artery (RA)
- Atherosclerotic RAS: Involves ostium or proximal 2 cm
- Fibromuscular dysplasia (FMD): Involves mid and distal segments of RA ± intrarenal arteries; right > left
- Affected kidney can be normal sized or atrophic
- Doppler findings
  - Direct signs (proximal criteria)
    - ↑ peak systolic velocity **> 200 cm/sec** (> 60% stenosis)
    - ↑ renal/aortic peak velocity ratio **> 3-3.5** (> 60% stenosis)
    - ↑ velocity gradient across RA: > 2.7
    - Poststenotic turbulent flow with spectral broadening and color aliasing
  - Indirect signs (distal criteria): Much less reliable and shouldn't be used as sole criteria for diagnosing RAS
    - Pulsus tardus-parvus, ↑ acceleration time, ↓ resistive index

PATHOLOGY

- **Atherosclerosis**
  - Most common cause of RAS (60-90%); more common in elderly
- **Fibromuscular dysplasia (FMD)**
  - 2nd most common cause of RAS (10-30%); more common in young and middle-aged women
  - 2/3 of cases are bilateral
- **Pathophysiology**
  - Decreased flow to affected kidney → activation of renin-angiotensin-aldosterone axis → production of angiotensin II → systemic vasoconstriction and increased level of aldosterone (accounting for fluid retention)



(Left) *Spectral Doppler sampling of the proximal left renal artery in 48-year-old man with history of intractable hypertension shows high velocity flow (368 cm/sec) indicating severe renal artery stenosis. The renal/aortic peak velocity ratio was also elevated (~ 4).* **(Right)** *Spectral Doppler tracing of the left interpolar segmental artery in the same patient shows parvus tardus waveform with increased acceleration time and borderline low resistive index (0.55).*



(Left) *Anteroposterior catheter angiography shows a variant appearance of renal artery stenosis, as the stenotic region ⇨ is distal to the ostium. Note the focal narrowing of the left renal artery ~ 2 cm from the origin of the renal artery.* **(Right)** *The patient underwent balloon angioplasty and placement of a stent ⇨, with marked improvement of narrowing. Angiographic intervention is very effective (80% success rate) in treating stenoses distal to the renal ostium.*

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TERMINOLOGY

Abbreviations

- Renal artery stenosis (RAS)
- Fibromuscular dysplasia (FMD)

IMAGING

General Features

- Best diagnostic clue
  - Focal or segmental luminal narrowing of renal artery (RA)
- Location
  - Atherosclerotic RAS
    - Renal artery ostium or within proximal 2 cm
  - Fibromuscular dysplasia: (Right > left)
    - Commonly involves mid or distal main RA ± intrarenal arteries

Ultrasonographic Findings

- Grayscale ultrasound
  - Affected kidney can be normal sized or atrophic
- Color Doppler
  - Direct signs (proximal criteria)
    - ↑ peak systolic velocity (PSV): Best predictor of RAS
      - 120-200 cm/sec: Mild, < 60% stenosis
      - **> 200 cm/sec**: Severe, 60-99% stenosis
    - ↑ renal/aortic PSV ratio **> 3-3.5** (severe, > 60% stenosis)
    - ↑ velocity gradient across RA: > 2.7
    - Poststenotic turbulent flow with spectral broadening and color aliasing
    - Complete occlusion with absence of flow
  - Indirect signs (distal criteria)
    - **Much less reliable and their absence does not exclude RAS**; hence, indirect should be used in combination with direct signs
    - Pulsus tardus-parvus: Dampened Doppler waveform during systole
      - ↑ acceleration time: > 0.05-0.08 sec
    - Resistive index (RI): > 0.05-0.07 difference between RI of affected kidney and contralateral normal kidney
  - Administration of captopril
    - ↑ detection when degree of RAS is > 50%

Angiographic Findings

- Findings on CTA, MRA, or conventional angiography
  - Atherosclerotic lesions
    - Focal/segmental, eccentric/concentric, ± Ca++
    - Location: Ostium or proximal 2 cm
  - FMD (medial fibroplasia subtype)
    - Stenosis, string of beads pattern, focal aneurysms
    - Location: Mid or distal long segment of main RA ± intrarenal arteries
    - Conventional catheter angiography has higher sensitivity to show these changes
  - FMD (intimal fibroplasia or medial hyperplasia subtypes)
    - Focal concentric narrowing of distal main RA and intrarenal branches
  - Secondary signs of RAS
    - Poststenotic dilatation ± collateral vessels
    - Decreased, delayed, or absent nephrogram
    - Normal sized or atrophic kidney

Nuclear Medicine Findings

- Renal scintigraphy
  - Baseline and ACE-inhibited phases
  - Baseline: Normal symmetric uptake and excretion of radiotracer (Tc-99m MAG3); activation of renin-angiotensin-aldosterone axis compensate for RAS
  - ACE-inhibited phase: Impaired function of affected kidney with delayed uptake and delayed excretion of radiotracer

Imaging Recommendations

- Color Doppler US (3.5 MHz curvilinear probe)
  - Excellent screening modality
  - Need to follow-up with another modality for better morphologic detail of etiology, location, and severity of disease
  - Beware of extrarenal and systemic conditions affecting spectral waveforms (such as valvular heart disease, extensive atherosclerosis, etc.)
- CT angiography (CTA)
  - MIP, volume-rendering techniques (complimentary)
- MR angiography (MRA)
  - Multiplanar, contrast enhanced, 3D, maximum intensity projection, fast gradient echo sequences
- Digital subtraction catheter angiography (DSA)
  - Usually in anticipation of angiographic intervention

DIFFERENTIAL DIAGNOSIS

Arterial Dissection

- Aortic dissection extending into renal artery
- Most frequently seen in elderly people (M > F)

Vasculitis

- Polyarteritis nodosa, Takayasu arteritis
  - Takayasu: Inflammation of medium-large arteries
  - Most common in Asian women age 15-45 years
- Fibrous thickening of wall of aorta, narrowing orifices of major branches
- Occasionally distal part of aorta and its branches are involved

Extrinsic Compression of Renal Artery

- Caused by
  - Abdominal aortic aneurysm
  - Retroperitoneal tumors
  - Retroperitoneal fibrosis

PATHOLOGY

General Features

- Etiology
  - **Atherosclerosis**
    - Most common cause of RAS (60-90%); more common in elderly
    - Stenosis due to atherosclerotic plaque and Ca++ deposition in ostium or proximal 2 cm of RA
    - 30-40% of cases are bilateral
  - **Fibromuscular dysplasia (FMD)**

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- 2nd most common cause of RAS (10-30%); more common in young and middle-aged women
- Medial fibroplasia subtype usually accounts for stenosis; involves mid and distal segments of RA
- 2/3 of cases are bilateral
- Subtypes
  - Medial fibroplasia (75-80%): Most common type
  - Intimal fibroplasia (10%)
  - Perimedial (subadventitial) fibroplasia (10%)
  - Medial hyperplasia (1-2%)
  - Adventitial fibroplasia (rare)
- Congenital RAS
  - Due to intimal fibroplasia of distal 2/3 of RA and its branches
- Rare causes of RAS
  - Thromboembolic, arterial dissection, infrarenal aortic aneurysm, postradiation
  - Buerger and Takayasu disease, polyarteritis nodosa
  - Neurofibromatosis, retroperitoneal fibrosis

- Pathophysiology
  - Decreased flow to affected kidney → activation of renin-angiotensin-aldosterone axis → production of angiotensin II → systemic vasoconstriction and increased level of aldosterone (accounting for fluid retention)

Gross Pathologic & Surgical Features

- Atherosclerotic RAS
  - Eccentric plaque in ostium or proximal RA
- FMD
  - String of beads pattern in mid or distal RA
- Preserved renal size in mild cases; renal atrophy in moderate to severe cases
- Poststenotic dilatation of main RA ± collateral vessels

Microscopic Features

- Atherosclerotic RAS
  - White-yellow intimal plaque of vessel wall
  - Mixture of fatty, fibrous, and calcium components
- FMD
  - Focal muscular thinning of media + fibrous ridges

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - New-onset severe hypertension (> 180 mmHg systolic or > 120 mmHg diastolic) after 55 years of age
  - Hypertension that is difficult to control with medication
  - Unexplained deterioration of renal function following antihypertensive therapy with ACE inhibitors
  - Epigastric or flank bruit
  - Symptoms of increased intracranial pressure
    - Papilledema, headache, vomiting
- Lab data
  - Positive captopril test
    - Exaggerated ↑ in plasma renin activity (PRA) following administration of captopril
  - ± impaired renal function tests

Demographics

- Age
  - Atherosclerotic RAS: Older adult (> 50 years); less common in African Americans
  - FMD: Young and middle-aged patients
- Gender
  - Atherosclerotic RAS: M > F
  - FMD: F > M (F:M = 3:1)
- Epidemiology
  - RAS is most common cause of secondary hypertension
  - Accounts for < 5% of hypertension cases in adults

Natural History & Prognosis

- Complications
  - Severe hypertension: Accelerated heart disease, cerebrovascular attack, azotemia
- Prognosis
  - Transluminal angioplasty
    - Success rate: Nonostial (80%), ostial lesions (25-30%)
  - Surgical revascularization
    - Success rate: 80-90% for any lesion and location
    - FMD: ↓ incidence of restenosis and improved glomerular filtration rate

Treatment

- Majority of cases can be controlled with antihypertensive drugs
- Transluminal angioplasty ± stenting
  - Stents are not commonly employed for FMD due to good response to angioplasty alone
- Surgical revascularization
- **Large randomized clinical trial (ASTRAL) comparing medical therapy alone with revascularization plus medical therapy in patients with atherosclerotic RAS did not show any benefit for revascularization**

DIAGNOSTIC CHECKLIST

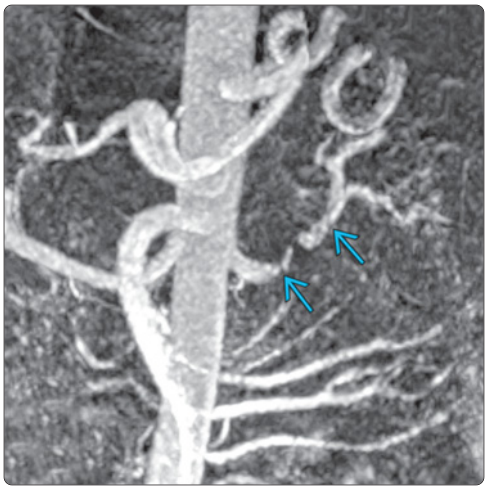
Consider

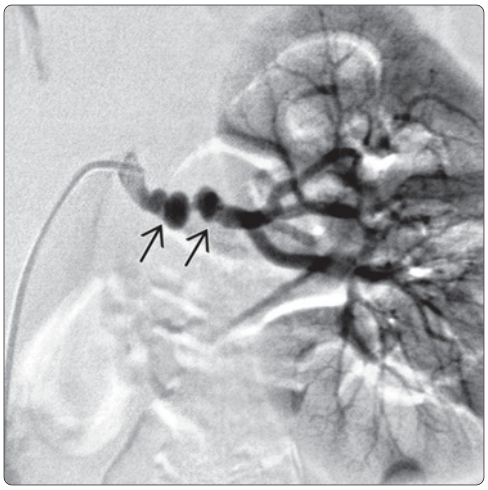
- RAS in patient with high or sudden ↑ in blood pressure + unexplained impairment of renal function
- Rule out other causes of RAS, especially dissection and vasculitis

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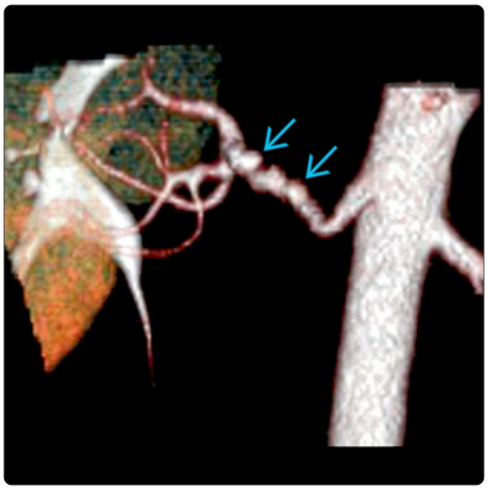
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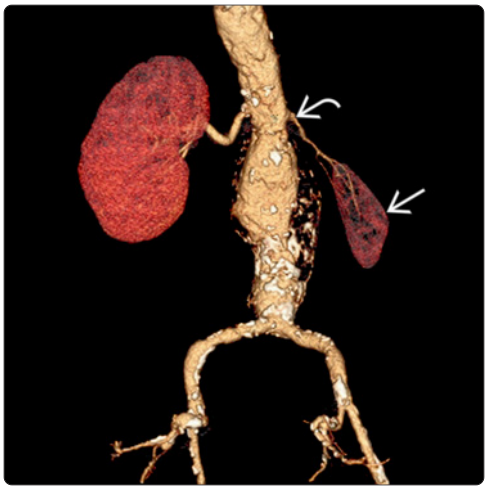


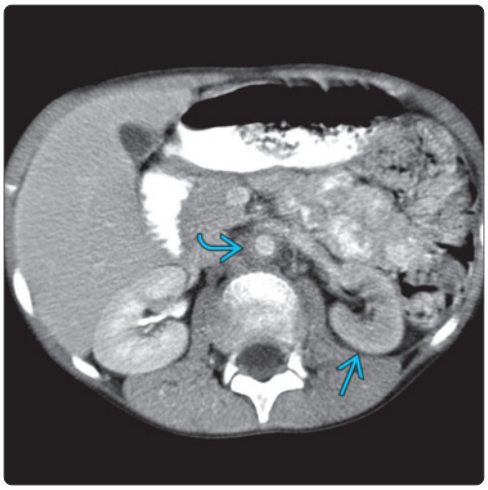
(Left) *Coronal oblique MRA in a young woman with hypertension shows a beaded appearance ⇨ of the left renal artery, findings characteristic of fibromuscular dysplasia.* **(Right)** *The "beaded" stenoses ⇨ in the same patient are best shown on this selective renal arterial catheterization. This was treated successfully with balloon angioplasty (not shown).*





(Left) *3-D rendered reconstruction from CTA shows a "beaded" appearance of the right renal artery ⇨ due to fibromuscular dysplasia.* **(Right)** *Axial CECT in a 41-year-old female with worsening hypertension shows global atrophy of the right kidney with delayed nephrogram, and a beaded renal artery ⇨ due to fibromuscular dysplasia (FMD).*





(Left) *Volume-rendered CTA shows an infrarenal abdominal aortic aneurysm and extensive atherosclerotic calcification. Note the atrophic left kidney ⇨ that was the result of chronic ischemia due to atherosclerotic renal artery stenosis ⇨.* **(Right)** *Axial CECT in 10-year-old boy with hypertension shows a small left kidney ⇨ with delayed nephrogram. Note the collar of soft tissue surrounding the aorta ⇨, which accounted for renal artery narrowing. A diagnosis of Takayasu arteritis was confirmed.*

Renal Infarction

KEY FACTS

TERMINOLOGY

- Localized or global area of ischemic necrosis in kidney secondary to interrupted vascular supply

IMAGING

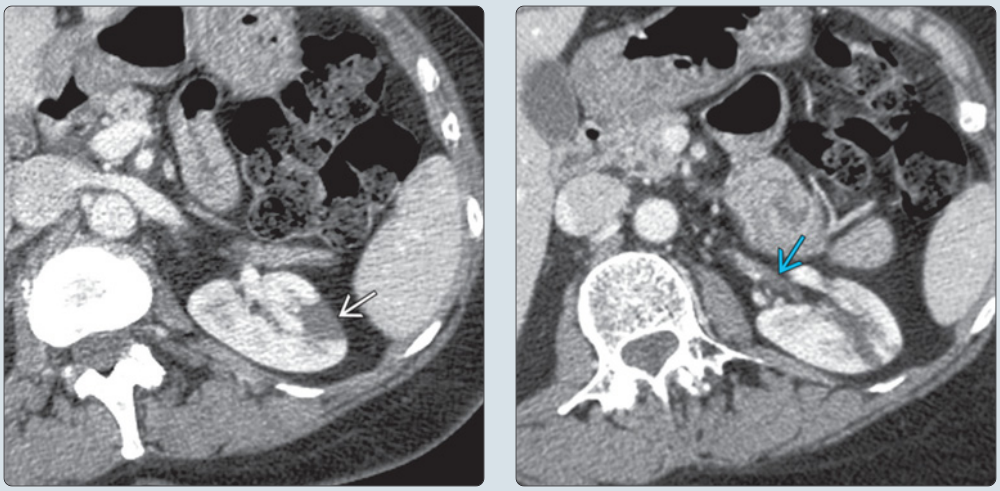
- Best diagnostic clue: Nonenhancing peripheral wedge-shaped area with enhancing cortical rim
- Imaging manifestations are variable based on etiology and chronicity
  - Thromboembolic: Usually multifocal and bilateral
  - In situ thrombotic: Unilateral (segmental or global)

TOP DIFFERENTIAL DIAGNOSES

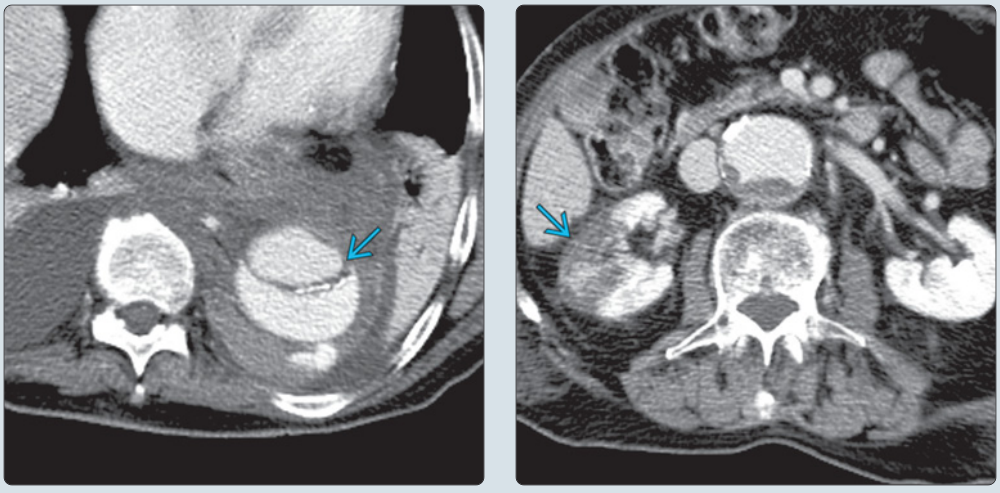
- Pyelonephritis
- Renal mass
- Multiple cortical scars
- Renal laceration or hematoma
- Vasculitis

PATHOLOGY

- Majority of cases are due to sudden occlusion of main renal artery or segmental branch artery
  - Venous infarction is seen in pediatric population (secondary to venous thrombosis); extremely rare in adults
- No causes can be found (idiopathic) in up to 1/3 of patients
- **Arterial infarction**
  - Thromboembolism: From cardiac or aortic source
  - In situ thrombosis: Secondary to trauma, vasculitis, hypercoagulable states, and iatrogenic injury
  - Dissection
- **Venous infarction (rare in adults)**
  - Dehydration is the most common risk factor in children
  - Other causes include: Nephrotic syndrome, hypercoagulable states, and renal cell carcinoma
- Torsion (rare)



(Left) *Axial CECT in a 57-year-old woman who presented with a known history of antiphospholipid syndrome and acute-onset left flank pain and hematuria illustrates a wedge-shaped area of nonenhancement in the lateral aspect of the left upper pole.* **(Right)** *Axial CECT in the same patient reveals a thrombus within the left main renal artery.*



(Left) *Axial CECT in an 82-year-old hypertensive man who presented with severe right-sided back pain demonstrates a type B aortic dissection. Note the intimal flap.* **(Right)** *Axial CECT in the same patient reveals a right renal infarct as the aortic dissection extended into the right renal artery, which was supplied by the false lumen of the aorta; infarction ensued due to low flow.*

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TERMINOLOGY

Definitions

- Localized or global area of ischemic necrosis in kidney secondary to interrupted vascular supply
  - Majority of cases are secondary to sudden occlusion of arterial supply

IMAGING

General Features

- Best diagnostic clue
  - Nonenhancing peripheral wedge-shaped area with enhancing cortical rim
- Location
  - Varies depending on etiology
    - Thromboembolic
      - Usually multifocal and bilateral
    - In situ thrombotic
      - Unilateral (segmental or global)
    - Venous infarction
      - More common on left side (possibly due to longer course of left renal vein)
  - Bilateral involvement seen in 15% of cases
- Morphology
  - Varies depending on chronicity
    - Acute
      - Wedge-shaped; normal to slightly increased size ± subcapsular fluid collection and perinephric stranding
    - Chronic
      - Wedge shaped; atrophy of infarcted segment with retraction of capsule

Fluoroscopic Findings

- IVP
  - Segmental or subsegmental infarction
    - Focal absent or decreased nephrogram
  - Global infarction
    - Complete absence of nephrogram and excretion

CT Findings

- CT manifestations are variable based on etiology and chronicity
  - Thromboembolic: Usually multifocal and bilateral
    - Infarction of other organs (spleen) might be seen as well
  - In situ thrombotic: Unilateral (segmental or global)
- Acute infarction
  - More commonly is segmental or subsegmental
  - Segmental/subsegmental infarction
    - **Sharply demarcated**, dorsal or ventral segmental (or subsegmental) area of decreased enhancement
    - Straight-line demarcation between normal enhancing and abnormal nonenhancing parenchyma
    - Base of wedge pointed toward renal capsule, apex toward hilum
  - Global infarction
    - Normal or enlarged kidney; smooth contour; ± subcapsular fluid collection
    - Absent or decreased nephrogram
    - ± medullary striations: "Spoke-wheel" enhancement (collateral circulation)
      - **Renal artery (RA) avulsion**: Total absence of renal enhancement, large perinephric hematoma
      - **RA thrombosis**: Total absence of renal enhancement, no excretion, no perinephric hematoma, preserved renal contour
  - Cortical rim sign
    - Preserved capsular/subcapsular enhancement
    - Seen in 50% cases of infarction (intact perinephric collateral circulation)
    - Usually seen 6-8 hours after infarction
- Chronic infarction
  - Absent/diminished enhancement in infarcted area; no cortical rim sign
  - Small size of infarcted area with capsular retraction; smooth or irregular contour

MR Findings

- T1WI
  - Variable; commonly T1 hyperintense (compared to noninfarcted renal parenchyma)
- T2WI
  - Depending on age of infarction, T2 signal can be variable
    - Usually iso- to hyperintense in acute phase
    - Hypointense to background renal parenchyma in chronic phase
- DWI
  - Acute infarction shows restricted diffusion (though not part of routine protocols)
- T1WI C+
  - Absence of enhancement in infarcted area
  - Sharp demarcation from enhancing noninfarcted parenchyma

Ultrasonographic Findings

- Grayscale ultrasound
  - Depending on age of infarct, affected areas may have different echogenicities; detection on grayscale US might be difficult
  - Areas of chronic infarction are usually hyperechoic
  - Renal vein thrombosis: Echogenic intraluminal filling defect
- Color Doppler
  - Absence of blood flow in infarcted area
  - Might be helpful to delineate level of vascular occlusion

Angiographic Findings

- Conventional angiography has both diagnostic and therapeutic role
  - Better delineates level of arterial occlusion &/or dissection
    - Helps to diagnose the underlying pathology (such as FMD, vasculitis, etc.)
  - Shows absence of perfusion and enhancement of infarcted area

Nuclear Medicine Findings

- SPECT imaging with Tc-99m DMSA
  - Lack of radiotracer uptake in infarcted area

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Imaging Recommendations

- Best imaging tool
  - MDCT with contrast; color Doppler sonography; selective renal angiography

DIFFERENTIAL DIAGNOSIS

Pyelonephritis

- Clinical presentation and underlying disease are helpful to differentiate between the 2 conditions
- Acute
  - Cortical wedge-shaped or striated nephrogram
    - May simulate segmental or subsegmental infarction
  - Loss of normal corticomedullary differentiation
  - Renal enlargement, focal swelling
  - Thickening of Gerota fascia and perinephric stranding
  - Calyceal effacement, dilated renal pelvis and ureter
- Chronic
  - Small kidney with cortical scarring over dilated calyces
  - Unilateral with compensatory hypertrophy of contralateral kidney

Renal Mass

- Acute infarction can rarely mimic renal mass (especially on US)
- Lack of enhancement, presence of cortical rim enhancement, wedged shape, and linear demarcation are helpful to differentiate the 2

Renal Tuberculosis

- Multiple cortical scars
- Parenchymal calcification
- Strictures of collecting system and ureter
  - "Putty" kidney of end-stage calcification

Renal Laceration or Hematoma

- Irregular linear or segmental nonenhancing tissue; subcapsular or perinephric hematoma

Vasculitis

- Polyarteritis nodosa, systemic lupus erythematosus, scleroderma, drug abuse (cocaine)
- Usually bilateral, multifocal, and subsegmental
- Microaneurysm dilatation of small vessels

PATHOLOGY

General Features

- Etiology
  - Majority of cases are due to sudden occlusion of main RA or a segmental branch artery
    - Venous infarction is seen in pediatric population (secondary to venous thrombosis); extremely rare in adults
  - In up to 1/3 of patients, no causes can be found (idiopathic)
  - **Arterial infarction**
    - Thromboembolism
      - From cardiac or aortic source
      - Common causes: Atrial fibrillation, endocarditis, myocardial infarction, prosthetic valve, aortic dissection
    - In situ thrombosis
      - Secondary to trauma, vasculitis, hypercoagulable states, and iatrogenic injury
      - Vasculitis (polyarteritis nodosa, etc.) and cocaine use are uncommon causes
      - Thrombosis of renal allograft vessels can be seen in setting of rejection
    - Dissection
      - Secondary to trauma, spontaneous dissection, vasculitis, or iatrogenic injury
      - Spontaneous dissection is seen in setting of fibromuscular dysplasia (FMD), Ehlers-Danlos syndrome, etc.
  - **Venous infarction (rare in adults)**
    - Dehydration is the most common risk factor in children
    - Other risk factors include
      - Nephrotic syndrome, hypercoagulable states, and renal cell carcinoma
    - Renal vein thrombosis in adults **rarely** results in infarction
  - Torsion (rare)
    - Rare complication of intraperitoneal renal transplantation
    - Extraperitoneal transplantation technique or prophylactic nephropexy are performed to avoid this complication
- Associated abnormalities
  - Cardiac abnormalities, hypercoagulable state, aortic aneurysm or dissection, splenic infarction

Staging, Grading, & Classification

- Classification based on onset
  - Acute, subacute, chronic
- Classification based on anatomy and vascular distribution
  - Segmental or subsegmental (cortex ± medulla); more common
    - Solitary or multiple, frequently associated with other renal injuries
  - Global

Gross Pathologic & Surgical Features

- Acute
  - Wedge-shaped area of coagulative necrosis, white or pale in color
  - Preserved immediate subcortical zone (equivalent or cortical rim sign)
  - RA thrombus or traumatic avulsion
- Chronic
  - Wedge-shaped area of atrophy

Microscopic Features

- Focal or global
- Acute
  - Coagulative necrosis of glomeruli and tubules with surrounding area of hyperemia and ischemia
- Chronic
  - Wedge-shaped areas of atrophy and scarring

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CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute onset flank or abdominal pain, nausea, vomiting
  - Leukocytosis, hematuria
  - Symptoms/signs are nonspecific and only < 50% of cases are diagnosed promptly

Demographics

- Age
  - Affects any age group
  - In 1 cohort, mean age was 53 years
- Gender
  - M = F
- Epidemiology
  - Rare (1.5% prevalence on autopsies)
  - Majority of cases are clinically undiagnosed, or diagnosed with delay, due to nonspecific symptoms

Natural History & Prognosis

- Complications (uncommon)
  - Infection, abscess formation
- Prognosis
  - Focal infarction: Good
  - Global infarction: Poor

Treatment

- Indicated in acute cases
  - Anticoagulation
  - Percutaneous endovascular therapy
    - Thrombolysis, thrombectomy, angioplasty
  - Nephrectomy for irreversible traumatic global infarction
- Antihypertensives (ACE inhibitors) in selected cases of hypertension caused by renal infarction

DIAGNOSTIC CHECKLIST

Consider

- Correlate with history: Trauma, cardiac/aortic disease, thromboembolic phenomena

Image Interpretation Pearls

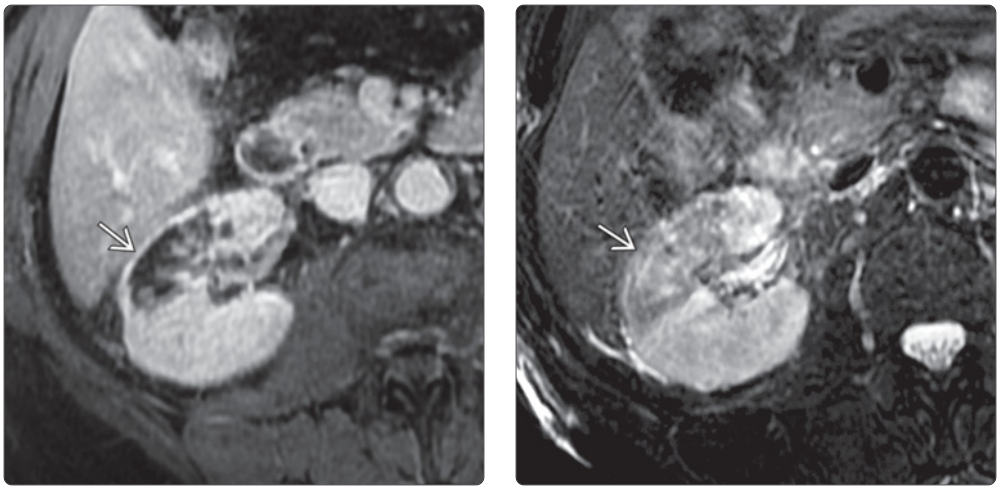
- Pyelonephritis and acute infarction may have similar appearance
- Straight-line demarcation and cortical rim sign favor infarction
- Perinephric stranding favors pyelonephritis

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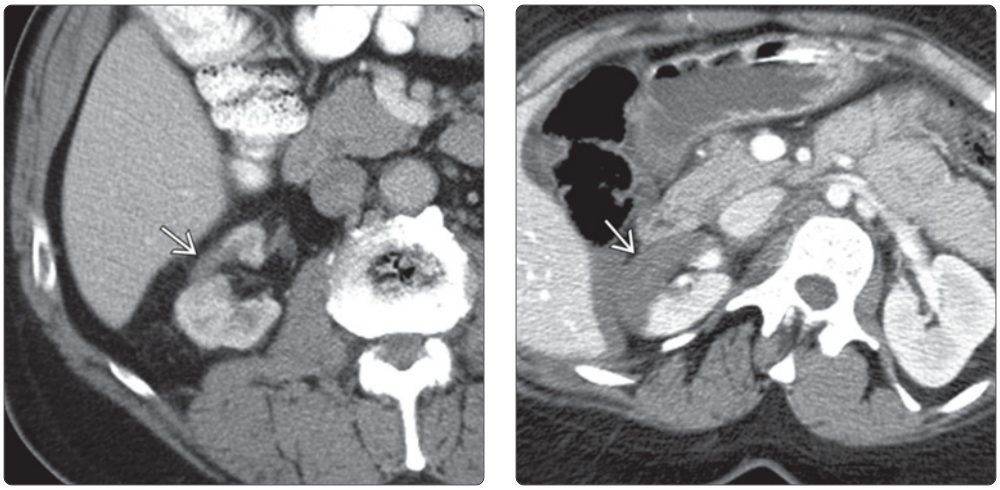
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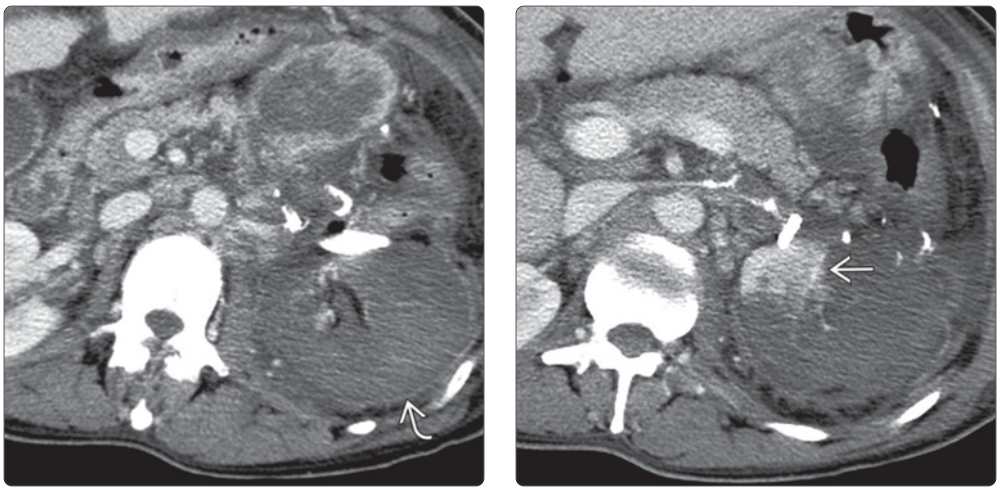
(Left) *Axial post-contrast T1WI MR in a 55-year-old woman with history of atrial fibrillation who was being worked up for a renal mass demonstrates a wedge-shaped area of nonenhancement in the right kidney. Presence of a cortical rim of enhancement ⇒ and preserved renal contour suggest that infarction is acute to subacute.* **(Right)** *Corresponding T2WI MR in the same patient shows decreased T2 signal in the infarcted area ⇒.*



(Left) *Follow-up axial CT in the same patient 1 year later shows evolution of infarction with atrophy and contour deformity in the infarcted area ⇒.* **(Right)** *Axial CECT in a 47-year-old man with acute onset of right flank pain after a motorcycle accident illustrates a traumatic right renal infarct ⇒ due to blunt trauma.*

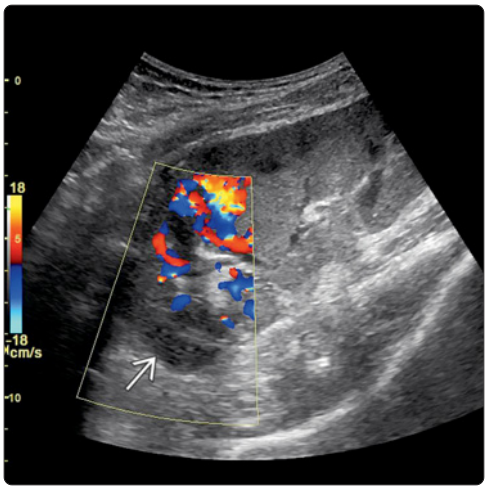


(Left) *Axial CECT in a 38-year-old man who sustained a blunt abdominal trauma following a motor vehicle crash demonstrates an extensive renal infarct with lack of enhancement. Note the faint cortical rim enhancement ⇒.* **(Right)** *Axial CECT in the same patient reveals only a small area of preserved enhancement in the left kidney ⇒. The left renal artery thrombosis occurred as a result of traumatic dissection.*

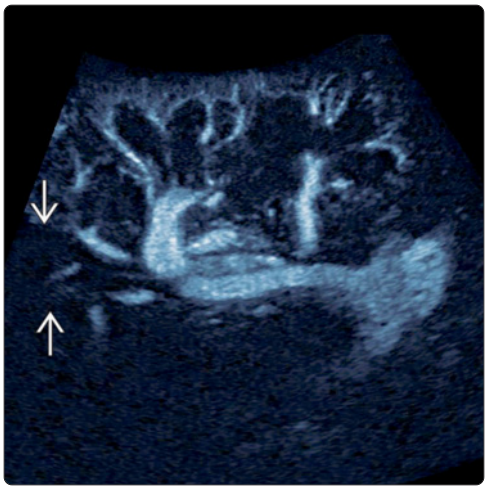


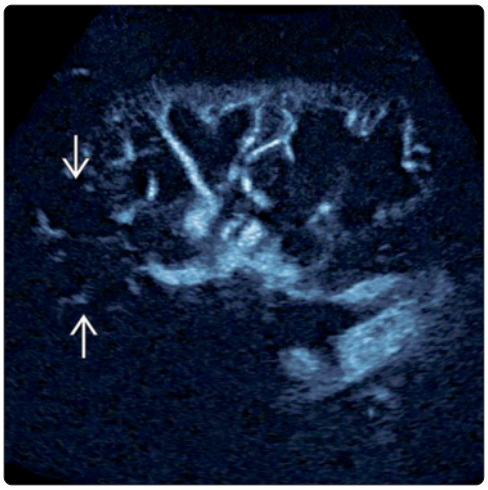
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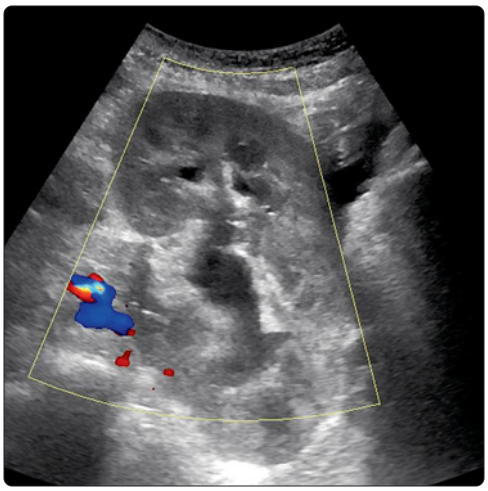


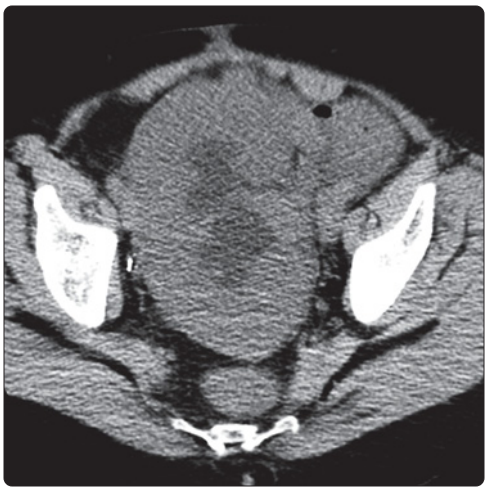
(Left) *Grayscale US in a woman with history of renal transplant shows a wedge-shaped area of decreased echogenicity ⇨ in the superior pole of the allograft.* **(Right)** *Corresponding color Doppler US of the same patient shows shows no flow within the upper pole ⇨. Infarcts are often poorly perceived at grayscale examination.*





(Left) *Arterial B-flow image shows a region of presumed infarction ⇨.* **(Right)** *The lack of vascularity ⇨ is more obvious on this venous B-flow image. The patient had a renal biopsy on the same day, which showed a large area of infarction presumably due to vasculitis and chronic rejection. (Courtesy G. Frey, RDMS.)*





(Left) *Grayscale US in a 60-year-old woman with history of kidney and pancreas transplantation and presenting with lower abdominal pain shows lack of intrarenal flow. Note the mild collecting system dilatation.* **(Right)** *Axial CT in the same patient shows marked enlargement and displacement of the renal allograft (transplanted in the LLQ). The patient underwent immediate surgical exploration, which revealed torsion of the intraperitoneal allograft.*

Renal Vein Thrombosis

KEY FACTS

TERMINOLOGY

- Obstruction of renal vein (RV) by thrombus

IMAGING

- RV thrombus with renal enlargement and delayed renal function
- Renal enlargement in 75% of cases
- Doppler findings
  - Reversal of diastolic flow in intrarenal arteries
  - Absent, or markedly reduced, venous flow
  - Turbulence and ↑ velocity of flow at level of obstruction (in subocclusive renal vein thrombosis [RVT])
  - Classic arterial Doppler findings are only seen in setting of acute RVT
- Best imaging tool: CECT, gadolinium-enhanced MR

TOP DIFFERENTIAL DIAGNOSES

- Other causes of unilateral renal enlargement
  - Ureteral obstruction
  - Infiltrating tumor
  - Pyelonephritis

PATHOLOGY

- Nephrotic syndrome and underlying malignancy (usually RCC) are the most common etiologies in adults
- Dehydration and sepsis are the most common etiologies in children

CLINICAL ISSUES

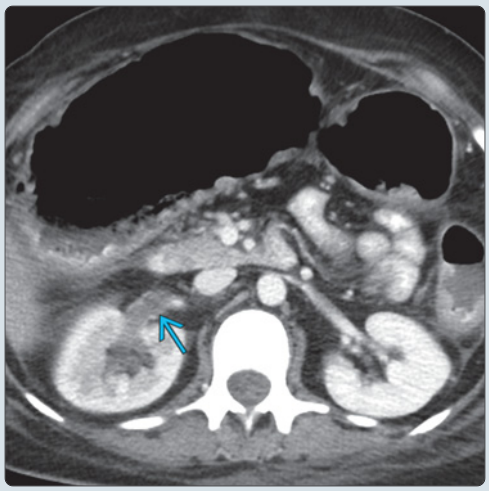
- More common in adults, but also seen in patients < 2 years
- Acute signs/symptoms: Flank pain, nausea, vomiting, palpable kidney, hypertension
- Chronic signs/symptoms: Asymptomatic or thromboembolic disease; fever, edema
- Treatment: Anticoagulation therapy

DIAGNOSTIC CHECKLIST

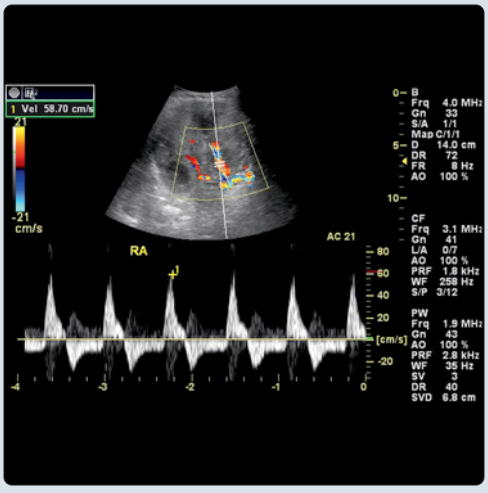
- Adequate return of renal circulation may prevent renal function deterioration

(Left) *Coronal post-contrast T1WI in 49-year-old man with history of CML and nephrotic syndrome shows bilateral renal venous thrombosis.* **(Right)** *Axial CECT in a 30-year-old man with a history of ulcerative colitis shows right renal vein thrombosis with associated delayed enhancement of the right kidney. Note the marked dilatation of the transverse colon with mucosal denudation and pseudopolyposis compatible with toxic megacolon.*





(Left) *Spectral Doppler tracing of a renal allograft artery shows reversal of diastolic flow. The allograft was noted to be enlarged. There was no detectable flow in the renal vein.* **(Right)** *Longitudinal section through the explanted renal allograft in the same patient demonstrates renal enlargement with areas of hemorrhagic infarction. The renal vein was noted to be completely thrombosed.*





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TERMINOLOGY

Abbreviations

- Renal vein thrombosis (RVT)

Definitions

- Obstruction of renal vein (RV) by thrombus

IMAGING

General Features

- Best diagnostic clue
  - Filling defect in renal vein with associated renal enlargement and delayed renal function
- Location
  - Unilateral > bilateral
    - Bilateral more common in children
  - Left > right
    - Thought to be due to longer length of left renal vein
- Size
  - Renal enlargement in 75% of cases
  - Small shrunken kidney (rare)
- Morphology
  - Mass in RV ± extension to inferior vena cava (IVC) ± right atrium

Fluoroscopic Findings

- IVP
  - Dense, prolonged nephrogram (partial)
  - Little or no nephrographic opacification (complete)
  - ↓ amount of opaque urine in renal calyces, infundibula, and pelvis
  - Opacified veins in perinephric space
  - Venous notching or ureteral indentation by tortuous collateral veins
  - Rarely alternating radiopaque and radiolucent striations in renal cortex

CT Findings

- CECT
  - Low-attenuation filling defect within RV
  - ↓ nephrographic attenuation: Persistent parenchymal opacification; no corticomedullary differentiation
  - Delayed corticomedullary junction time, contrast excretion into renal calyces and pelvis
  - Thickened Gerota fascia and perinephric "whiskering" (edema or hemorrhage)
  - Opacified periureteral and perinephric collaterals ("cobwebs")
- CTA
  - Tortuous and dilated collateral veins close to ureters
  - Venous collateralization: Retrograde flow in dilated superficial epigastric veins

MR Findings

- Filling defect in RV (best seen on post-contrast T1WI)
- Renomegaly, obliteration of renal sinus fat, compression of renal collecting systems
- Prolonged transit time of renal cortex and medulla
- Absence of corticomedullary differentiation
- Multiple perinephric collateral veins
- Gonadal vein dilatation

Ultrasonographic Findings

- Grayscale ultrasound
  - Renal edema
  - ↓ echogenicity (acute), ↑ echogenicity after 10-14 days
  - ± accentuated corticomedullary differentiation
  - Intraluminal filling defect with RV dilatation proximal to occlusion; more visible on left
    - Acute thrombosis: Anechoic or hypoechoic
    - Chronic thrombosis: Echogenic
  - Thrombus or tumor in IVC (≤ 20%)
- Color Doppler
  - Renal artery (RA) sampling
    - Reversal of, or absence of, diastolic flow
    - Increased resistive indices (RI) of intrarenal arteries
    - Classic arterial Doppler findings only seen in setting of acute RVT; as collateral circulation opens, Doppler findings become less characteristic.
      - Due to lack of collateral circulation in transplanted kidney, classic Doppler changes last longer
    - Normal arterial waveform does not exclude RVT
  - Renal vein (RV) sampling
    - Absent or markedly reduced venous signal
    - ↑ velocity and turbulence at level of obstruction in cases of subocclusive thrombosis

Angiographic Findings

- Filling defect in RV (during venous phase)
- Venous collateralization

Nuclear Medicine Findings

- Delayed or absent renal perfusion

Imaging Recommendations

- Best imaging tool
  - CECT, gadolinium-enhanced MR

DIFFERENTIAL DIAGNOSIS

Ureteral Obstruction

- Similar to RVT, ureteral obstruction can result in renal enlargement and delayed nephrogram
- Hydronephrosis, hydroureter, perinephric or periureteral stranding

Infiltrating Tumor

- Transitional cell carcinoma (TCC), renal cell carcinoma (RCC), renal metastases, and lymphoma are among other causes of unilateral renal enlargement
- Preserved shape of kidney with distorted internal architecture
- Heterogeneous mass ± calcification
- Enhancing mass with soft tissue attenuation

Pyelonephritis

- Differentiate by clinical history and urinalysis

PATHOLOGY

General Features

- Etiology
  - **Nephrotic syndrome**

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    - Membranous or membranoproliferative glomerulonephritis, focal sclerosis, amyloidosis, lipoid nephrosis
  - Other primary renal disease
    - Other glomerulonephritis, pyelonephritis, vasculitis
  - **Renal hypoperfusion by hypovolemia or vascular stasis**
    - Dehydration, sepsis, hemorrhage
    - Congestive heart failure, aortic insufficiency, constrictive pericarditis
  - **Tumor extension**
    - RCC, renal angiomyolipoma, TCC, Wilms tumor, metastases
  - Iatrogenic
    - Drugs (oral contraceptive pills, estrogens)
    - Abdominal surgery, renal biopsy
    - Renal transplant rejection
  - Other hypercoagulable states
    - Pregnancy, septic abortion, malignancy, hereditary hypercoagulable states
  - Abdominal trauma
  - Thrombus extension
  - Mechanical compression
- Genetics
  - Inherited hypercoagulable states (such as antithrombin 3, protein S, protein C deficiency)
- Nephrotic syndrome and underlying malignancy (usually RCC) are the most common etiologies in adults
- Dehydration and sepsis are the most common etiologies in children

Gross Pathologic & Surgical Features

- Renal fibrosis, hemorrhage, necrosis, calcification

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute (more common in children)
    - Flank pain, nausea, vomiting, palpable kidney, hypertension
  - Chronic (more common in adults)
    - Asymptomatic or thromboembolic disease, fever, edema
- Clinical profile
  - RVT is more often chronic
  - Acute RVT (which is more common in children) is most often due to trauma, generalized hypercoagulable state, and dehydration (especially in infants)
- Lab data
  - Acute: Gross or microscopic hematuria on urinalysis
  - Chronic: Proteinuria on urinalysis

Demographics

- Age
  - More common in adults
    - Also seen in patients < 2 years old
- Epidemiology
  - Nephrotic syndrome present in 16-42% of patients with RVT

Natural History & Prognosis

- Effects of RVT depend on site of origin, time to occlusion, collateral circulation, and extent of recanalization
- Complications
  - Pulmonary embolism
  - Widespread thrombosis; renal infarction, hemorrhage, and renal failure
- Prognosis: Good; frequent spontaneous recovery

Treatment

- Anticoagulation therapy
  - IV heparin → oral warfarin
- Thrombolytic therapy: Bilateral RVT, RVT extension into IVC, massive clot, pulmonary emboli, severe flank pain, or failed anticoagulation therapy
- Surgical thrombectomy or nephrectomy for failed medical treatment, tumor thrombus
- Suprarenal vena cava filter

DIAGNOSTIC CHECKLIST

Consider

- Adequate return of renal circulation may prevent renal function deterioration

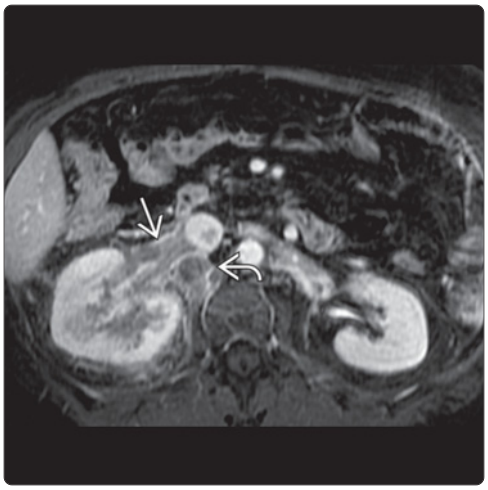
Image Interpretation Pearls

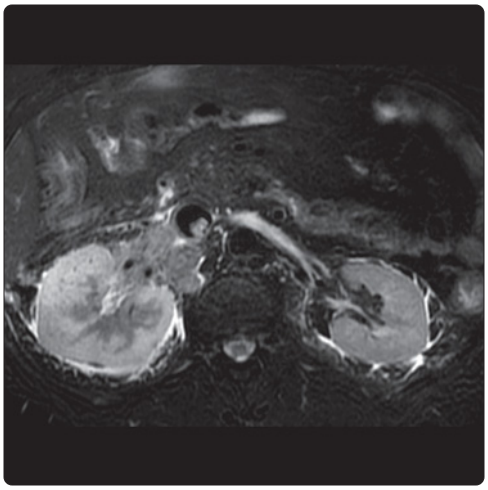
- Filling defect within RV; venous collaterals

SELECTED REFERENCES

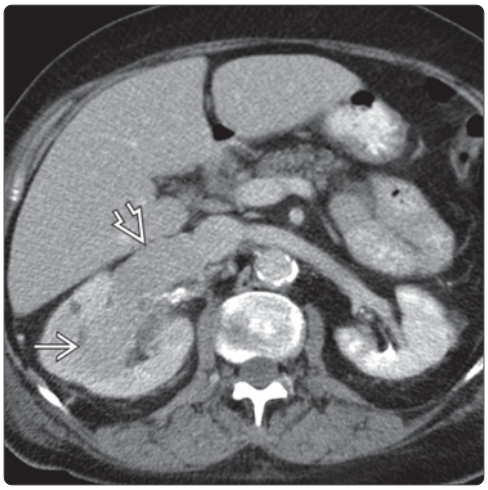
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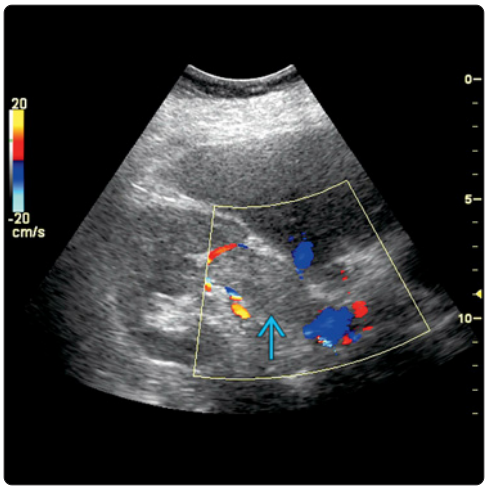
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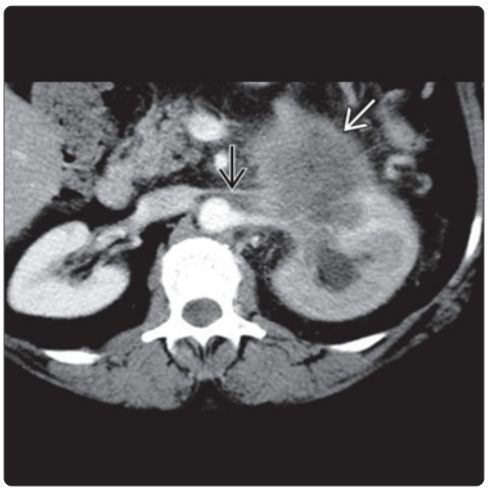
(Left) *Axial post-contrast T1WI in a 59-year-old man with history of right renal mass shows expansile thrombosis of the right renal vein with extension into the IVC and a lumbar branch. Note the enlargement of the right kidney and delayed nephrogram secondary to venous thrombosis.* **(Right)** *Axial T2WI in the same patient shows altered T2 signal in the right kidney secondary to venous thrombosis and congestion. Pathology was consistent with clear-cell renal cell carcinoma.*





(Left) *Axial CECT in a 60-year-old woman shows an enhancing mass in the interpolar region of the right kidney with tumor thrombosis in the right renal vein. Note the marked expansion of the renal vein.* **(Right)** *Color Doppler US in the same patient demonstrates expansile echogenic clot within the right renal vein.*





(Left) *Axial CECT in a 46-year-old woman who presented with vague abdominal pain and a history of protein S deficiency demonstrates a thrombus within the inferior vena cava and the left renal vein.* **(Right)** *Axial CECT in a patient presenting with weight loss and flank pain shows a pancreatic tail mass extending to and encasing the left renal vein with resultant renal vein thrombosis. Note the delayed nephrogram and hydronephrosis.*

Renal Trauma

KEY FACTS

IMAGING

- Best diagnostic clue: Renal parenchymal defect with perirenal hemorrhage ± extravasation of blood/urine
- CT findings
  - Laceration: Linear, hypoattenuating defect
  - Segmental renal infarct: Sharply demarcated, wedge-shaped area of decreased enhancement
  - Global infarction (nonenhancement) and no perinephric hematoma: Renal artery thrombosis
  - Global infarction (nonenhancement) and perinephric hematoma: Renal artery avulsion
- Protocol advice: If renal laceration is evident, obtain 10-12 minute delayed scans

TOP DIFFERENTIAL DIAGNOSES

- Hemorrhage from renal tumor
- Hemorrhage from vasculitis

PATHOLOGY

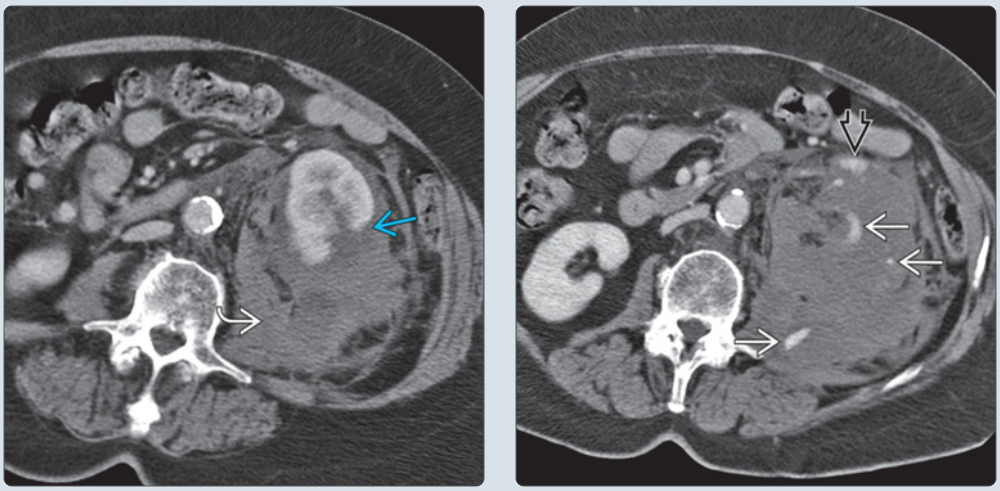
- Blunt, penetrating, and deceleration injuries
- Serious renal injuries usually associated with multiorgan involvement

CLINICAL ISSUES

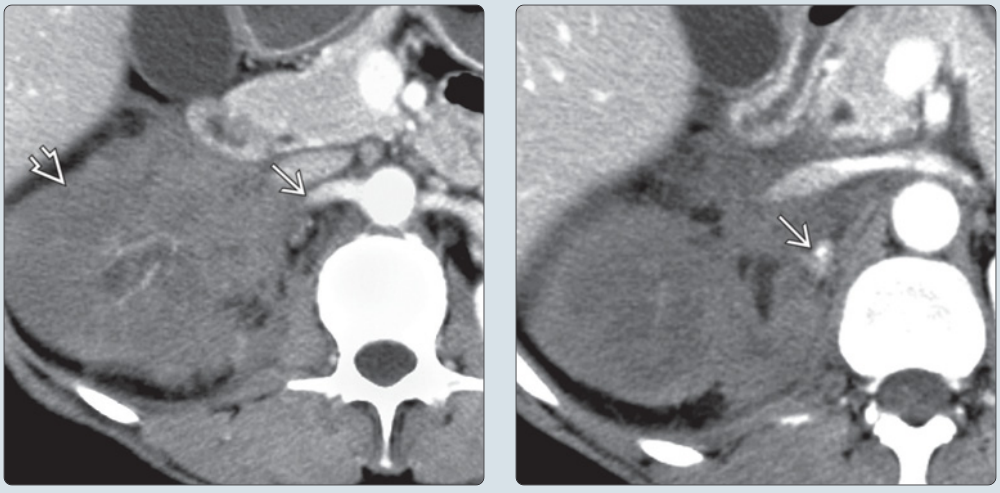
- Flank pain, ecchymosis, hematuria, shock
- Poor correlation between degree of hematuria and severity of injury
- AAST classification correlates well between surgical and MDCT findings

DIAGNOSTIC CHECKLIST

- Consider possibility of underlying tumor if bleeding seems disproportionate to degree of trauma
- Arterial extravasation usually requires catheter embolization
- Urinary extravasation often requires ureteral stent ± catheter drainage of urinoma



(Left) *Axial CECT following motor vehicle crash shows a contour deformity of the posterior aspect of the left kidney due to fracture (the shattered fragment is not shown on this image). Note the large perinephric hematoma.* **(Right)** *More inferior axial CECT in the same patient reveals multiple sites of active arterial extravasation & a fragment of the fractured kidney, consistent with a grade V injury. Due to the extensive parenchymal damage & active bleeding, an emergent nephrectomy was performed.*



(Left) *Axial CECT in a 19-year-old male who sustained blunt abdominal trauma in a motorcycle crash reveals a traumatic renal artery occlusion due to dissection. Nonenhancement of the right kidney is consistent with global infarction, consistent with grade V injury.* **(Right)** *Axial CECT in the same patient reveals an additional small area of active bleeding from the lumbar artery, which accounted for bleeding into the perirenal and psoas compartments.*

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IMAGING

General Features

- Best diagnostic clue
  - Renal parenchymal defect with perirenal hemorrhage ± extravasation of blood/urine

Radiographic Findings

- The most commonly used classification system for renal trauma was set forth by the American Association for the Surgery of Trauma (AAST)
  - AAST classification consists of 5 injury classifications
    - ~ 80-85% of renal injuries are classified as grade I
  - AAST classification strongly correlates with surgical and MDCT findings
  - AAST classification of injury corresponds to clinical outcome

CT Findings

- AAST classification of kidney injuries
- Grade I Injury
  - Contusion
    - Poorly defined region of decreased parenchymal enhancement
    - NCCT: Contusions may be iso- to hyperattenuating relative to adjacent normal renal parenchyma
  - Hematoma
    - Nonexpanding, subcapsular collection that has crescentic shape and follows renal contour when small
    - When large, subcapsular hematomas are convex and exert mass effect on kidney
    - No associated laceration in grade I injuries
- Grade II Injury
  - Hematoma
    - Nonexpanding hematoma confined to perinephric space
    - Blood dissecting along perinephric septa results in "stranding" or "cobwebbing"
    - Acute to subacute hematomas are iso- to hyperattenuating (~ 35-70 HU)
    - Chronic hematomas evolve and may approach near water attenuation (0-15 HU) as blood products become resorbed
  - Laceration
    - Superficial cortical defect (≤ 1 cm)
      - Typically linear and hypoattenuating
    - No associated collecting system injury
- Grade III Injury
  - Laceration
    - > 1 cm depth
    - Laceration does not affect collecting system
- Grade IV Injury
  - Laceration
    - Extends through renal cortex, medulla, and collecting system
  - Vascular
    - Injury of main renal artery or vein with contained hematoma
    - Segmental infarctions due to tear, dissection, thrombosis segmental vessels
      - Sharply marginated, wedge-shaped areas of nonenhancement
- Grade V Injury
  - Laceration
    - Shattered kidney
      - Pulverized renal parenchyma shown as separated islands of vascularized or devascularized tissue
    - Avulsion of ureteropelvic junction
      - Large volume urine leak/urinoma; confirmed by extravasation of contrast-opacified urine during excretory phase
  - Vascular
    - Avulsion or thrombosis of main renal artery or vein with renal devascularization
      - Avulsion: Brisk extravasation of contrast and massive hematoma
      - Complete avulsion of renal vessels is often fatal
      - Thrombosis: Abrupt termination of renal vessel
      - Devascularization results in no enhancement of renal parenchyma; persistent enhancement (rim sign) of renal capsule is due to numerous small collateral vessels that perforate through perinephric fat
      - Rim sign usually not apparent until ~ 8 hours after injury
- AAST classification does not specifically address active hemorrhage
  - Active hemorrhage is more commonly associated with high-grade injuries (grades IV and V) but can occur with lower grade injuries (grade II and III)

Imaging Recommendations

- Protocol advice
  - Multiphasic CECT is preferred modality
  - If renal laceration is evident on CT, mandatory 10-12 minute delayed scan (to evaluate for urine extravasation)
- Retrograde pyelogram: Assess ureteral and renal pelvic injuries

MR Findings

- MR typically plays no role in evaluation of acute renal injury
- MR can be useful for detection of complications
- Hematoma: Varies with age of blood products
  - Acute: ↑ signal intensity (SI) on T1WI; ↓ SI on T2WI
  - Subacute: ↑ SI on T1WI and T2WI
  - Chronic: ↓ SI on T1WI and T2WI
- Urinoma: ↓ SI on T1WI; ↑ SI on T2WI
- Pseudoaneurysm: Contained, smoothly marginated contour deformity that follows arterial enhancement

Angiographic Findings

- Active hemorrhage: Ill-defined blush of extravasated contrast material
- Pseudoaneurysm: Contained, smoothly marginated outpouching of contrast
- Dissection: Linear, intraluminal flap due to intimal injury
- Occlusion: Lack of flow through lumen due to traumatic dissection or embolus
- Arteriovenous fistula: Early opacification of vein during arterial phase injection

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Ultrasonographic Findings

- While ultrasound (US) is useful for detection of hemoperitoneum in the acute setting, it typically does not play a significant role for acute renal injury
- Similar to MR, US is useful for evaluation of complications of renal trauma
- Hematoma: Varies with age of blood products
  - Acute: Echogenic fluid collection
  - Chronic: Resorption of blood product leads to anechoic collection
- Pseudoaneurysm: Swirling of blood ("yin yang" sign)
  - To and fro spectral pattern at neck of pseudoaneurysm
- Arteriovenous fistula: Turbulent, low-resistance waveform (high diastolic flow)

DIFFERENTIAL DIAGNOSIS

Renal Tumor

- Spontaneous bleed/rupture may be seen
- Perinephric fluid collection of blood density
- Underlying renal mass lesion
  - Renal cell carcinoma (RCC)
    - Solid renal mass, usually hypervascular
    - Possible renal vein/IVC extension
  - Angiomyolipoma (AML)
    - Vascular, smooth muscle, and fat components
    - Renal mass with variable amount of fat is diagnostic

Vasculitis

- Polyarteritis nodosa, systemic lupus erythematosus, scleroderma, drug abuse
- Wedge-shaped defects or striated nephrogram
- Microaneurysms of small vessels

PATHOLOGY

General Features

- Etiology
  - Motor vehicle accidents, falls, assault
    - Blunt, penetrating, and deceleration injuries
    - Renal trauma identified in 8-10% of patients with abdominal injuries
      - Overall, blunt trauma accounts for ~ 80% of all renal injuries
- Associated abnormalities
  - Significant renal injuries associated with polytrauma

Gross Pathologic & Surgical Features

- Contusion, laceration, hematoma, infarction, vascular, or ureteropelvic injury

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Flank pain, tenderness, ecchymosis
  - Hematuria, anuria, uremia, shock
  - Poor correlation between degree of hematuria and severity of injury
- Clinical profile
  - History of vehicular accident, flank pain, hematuria
- Lab data: Hematuria (> 5 RBCs per high-power field)

Demographics

- Any age and gender

Natural History & Prognosis

- Complications
  - Early: Urinoma, perinephric abscess, sepsis, arteriovenous fistula, pseudoaneurysm
  - Late: Hydronephrosis, hypertension, chronic pyelonephritis, renal failure, atrophy
  - Delayed hemorrhage: ~ 2-3 weeks after initial injury

Treatment

- 98% of renal injuries are low grade and managed conservatively
- Grade I and II injuries: Conservative therapy
  - Typically self-limited
- Grade III Injuries: Typically conservative therapy
  - Associated with renal scarring but minimal impact to renal function
  - Active urinary extravasation: ± ureteral stent
- Grade IV-V Injuries: More likely to require intervention
- Surgical or angiographic indications
  - Active hemorrhage
  - Vascular (renal pedicle) injury
  - Shattered kidney
  - Expanding or pulsatile hematoma
  - Shocked polytrauma patient
  - Ureteropelvic junction tear: Observation ± stent
  - Ureteropelvic avulsion: Surgical repair

DIAGNOSTIC CHECKLIST

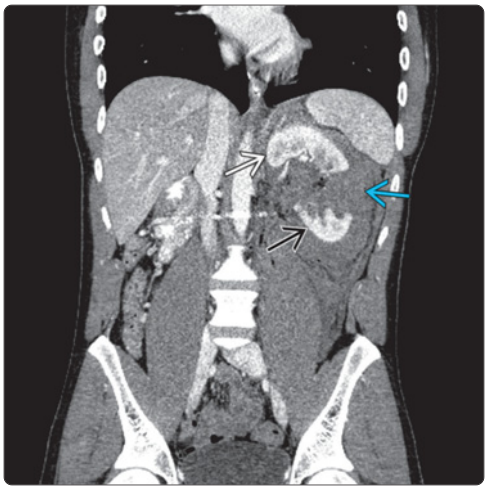
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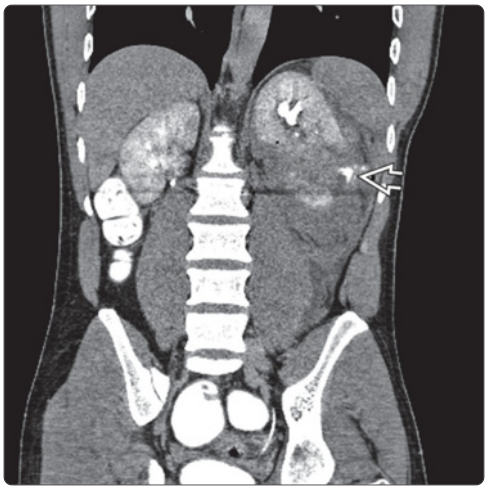
- Possibility of underlying tumor if bleeding seems disproportionate to degree of trauma

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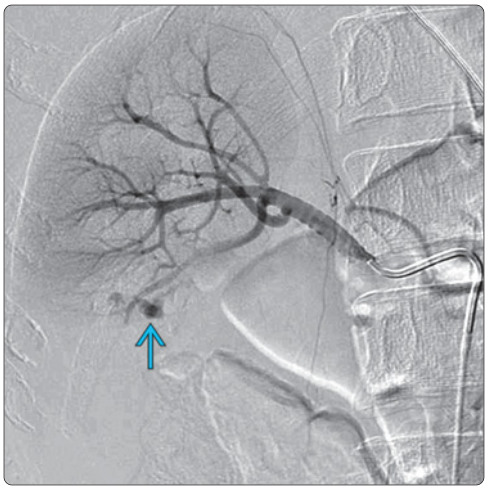
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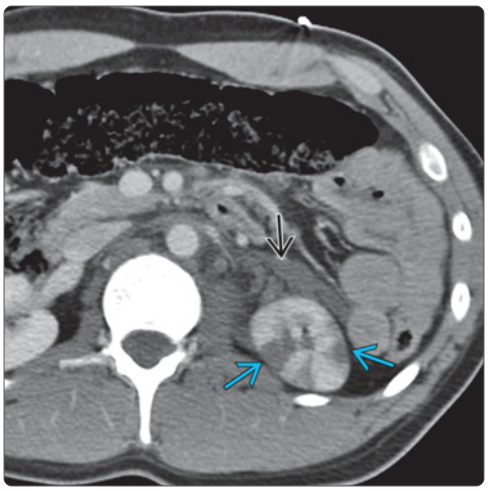


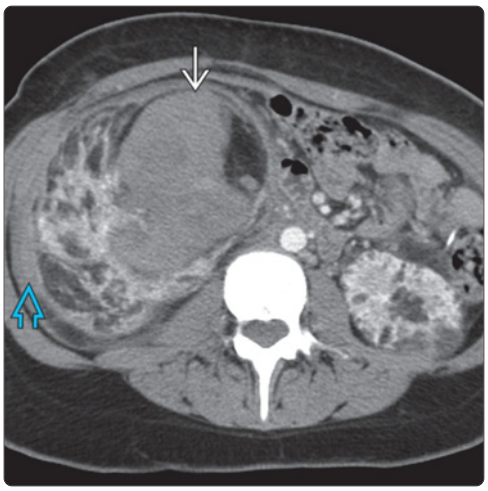
(Left) *Coronal CECT during the nephrographic phase following gunshot wound through the left flank reveals fracture of the left kidney, consistent with grade V injury; the superior ➡ and inferior ⇨ fragments are perfused but are separated and surrounded by intermediate attenuation 'fluid' ⇨.* **(Right)** *Coronal CECT during the excretory phase in the same patient reveals extravasation ➡ of contrast-opacified urine due to disruption of the renal pelvis.*





(Left) *Coronal CECT during the corticomedullary phase demonstrates fracture ➡ of the right kidney and a traumatic pseudoaneurysm ⇨ within the hematoma that separates the superior and inferior fragments.* **(Right)** *AP view during digital subtraction angiography shows a catheter in the right renal artery and the pseudoaneurysm ⇨ adjacent to the avascular plane of the renal fracture. The pseudoaneurysm was subsequently treated with coil embolization.*





(Left) *Axial CECT following trauma reveals hematoma ⇨ in the left perinephric space and multiple peripheral areas of nonenhancement due to sites of contusion ⇨. The patient was treated conservatively and did not experience any long-term sequelae.* **(Right)** *Axial CECT was performed after a patient developed right flank pain following minor trauma. There is a large hematoma ➡ within an angiomyolipoma (AML) and a small amount of perinephric hematoma ⇨. The patient had bilateral AMLs due to tuberous sclerosis.*

Urinoma

KEY FACTS

TERMINOLOGY

- Loculated collection of urine from urinary leak

IMAGING

- Location
  - Along course of urinary tract: Perinephric space, retroperitoneum along ureter, extraperitoneal pelvis adjacent to bladder
  - Pelvic intra- or extraperitoneal location secondary to bladder rupture
- Best imaging tool: CECT or MR; cystogram if bladder leak suspected
  - Typically appears as well-marginated, homogeneous fluid collection
  - Thin rim enhancement with near water attenuation interior
  - ↓ SI on T1W; ↑ SI on T2W
  - Consider US for assessment and image-guided drainage
- Protocol advice
  - CECT or MR with additional imaging during excretory phase (10-minute delay)
  - Cystogram: Distend bladder with at least 250 mL of contrast to visualize leak from bladder dome

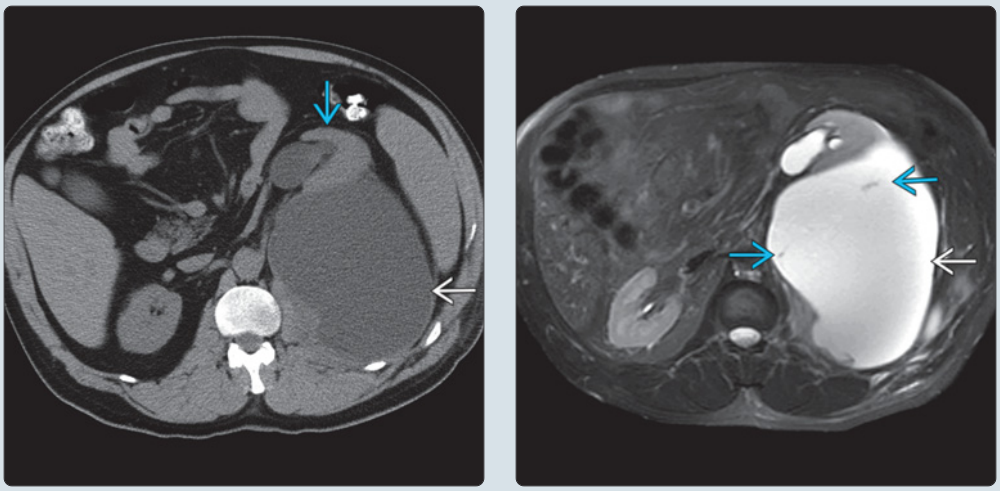
TOP DIFFERENTIAL DIAGNOSES

- Hematoma
- Abscess
- Lymphocele

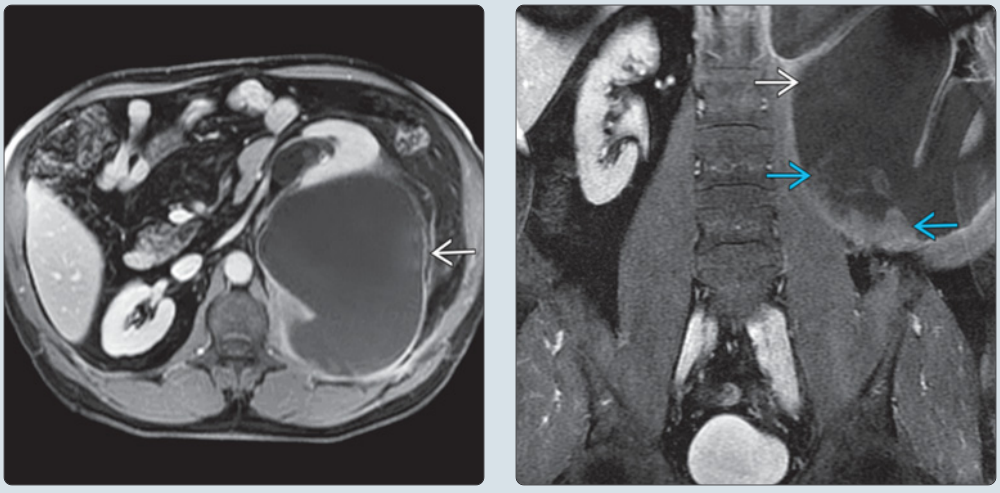
CLINICAL ISSUES

- Most common signs/symptoms: Pain due to mass effect; fever if infected
  - Consider superinfection if imaging appearance is complex
- Other signs/symptoms: Ileus, electrolyte imbalance
- Small urinomas typically resolve spontaneously
- Large urinomas require percutaneous or surgical drainage

(Left) *Axial NECT following partial nephrectomy. A large urinoma is encapsulated and has homogeneous near-water attenuation. Note anterior displacement of the left kidney and hydronephrosis due to mass effect.* **(Right)** *Axial T2W MR with fat saturation shows homogenous high signal intensity within the urinoma; a few thin septa are noted.*



(Left) *Axial post-contrast T1W MR with fat saturation shows enhancement of the rim of the urinoma.* **(Right)** *Coronal T1W MR with fat saturation during the excretory phase shows accumulation of contrast material layering within the urinoma. Excretory phase imaging is the key to differentiating urinomas from other fluid collections since excreted contrast will accumulate in the urinoma. The excretory phase is acquired ~ 8-12 minutes after the initial contrast injection.*



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TERMINOLOGY

Definitions

- Loculated collection of urine from urinary leak

IMAGING

General Features

- Best diagnostic clue
  - Fluid collection containing extravasated contrast material from renal collecting systems, ureters, or bladder during excretory phase CT/MR or contrast extravasation during cystogram
- Location
  - Along course of urinary tract: Perinephric space, retroperitoneum along ureter, extraperitoneal pelvis adjacent to bladder
  - Pelvic intra- or extraperitoneal location secondary to bladder rupture

Imaging Recommendations

- Best imaging tool
  - CECT with excretory phase imaging; cystogram if bladder leak suspected
- Protocol advice
  - Obtain images at 10-minute delay during excretory phase of intravenous pyelogram or CECT
  - Cystogram: Distend bladder with at least 250 mL of contrast to visualize leak from bladder dome

Ultrasonographic Findings

- Hypoechoic fluid collection with thin wall in perirenal space, retroperitoneum, or pelvis
  - Few, thin septa variably present
  - Increasing complexity (thick septa/wall, layering debris/internal echoes) may indicate superinfection

CT Findings

- NECT: Water attenuation fluid collection in retroperitoneum or pelvis
- CECT: Excretory phase shows leakage of contrast into urinoma
  - Variable enhancement of thin rim during venous phase

MR Findings

- Fluid collection with homogeneous low signal intensity (SI) on T1WI and high SI on T2WI
- Enhancement characteristics are similar to CT

Radiographic Findings

- IVP
  - Excretory phase may demonstrate leakage of contrast into urinoma

DIFFERENTIAL DIAGNOSIS

Hematoma

- US: Echogenicity varies with age and size of clot
- NECT: Typically > 30 HU
- MR: Variable SI depending on age of blood products, but often heterogeneous

Abscess

- US: Thick/irregular wall with variable color flow, thick septations, low-level internal echoes, ± fluid-fluid level
- CECT: Enhancing thick/irregular wall, heterogeneous internal debris ± fluid-fluid level, &/or internal gas locules
- MR: Rim-enhancing fluid collection with variable internal SI depending on contents and protein content

Lymphocele

- History of lymph node dissection; may be bilateral
- US/CT/MR: Defined cystic lesion with imaging characteristics of water found in distribution of node dissection
  - May contain several thin septa

PATHOLOGY

General Features

- Etiology
  - Disruption of collecting system or ureter
  - Forniceal rupture (e.g., acute obstruction from stone)
  - Intra- or extraperitoneal bladder rupture
- Associated abnormalities
  - Renal trauma with deep laceration or fracture

Gross Pathologic & Surgical Features

- Rare fibrocapillary capsule; mural calcification if chronic

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Pain due to mass effect; fever if infected
- Other signs/symptoms
  - Ileus, electrolyte imbalance

Demographics

- Any age, M = F

Natural History & Prognosis

- Small urinomas typically resolve spontaneously

Treatment

- Large urinomas require percutaneous or surgical drainage

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Excretory phase CECT or MR (10-minute delay)

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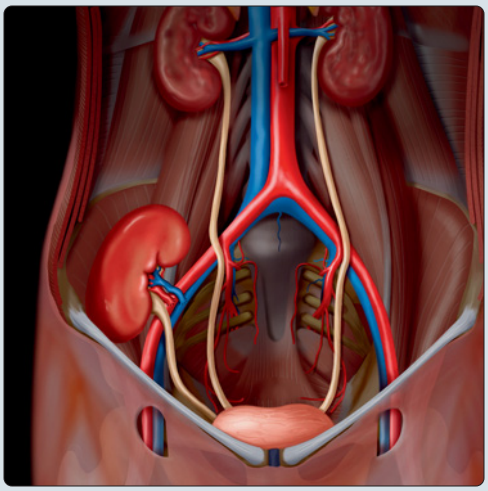
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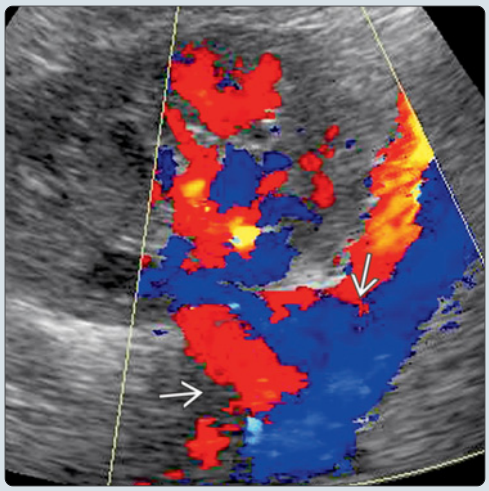
KEY FACTS

IMAGING

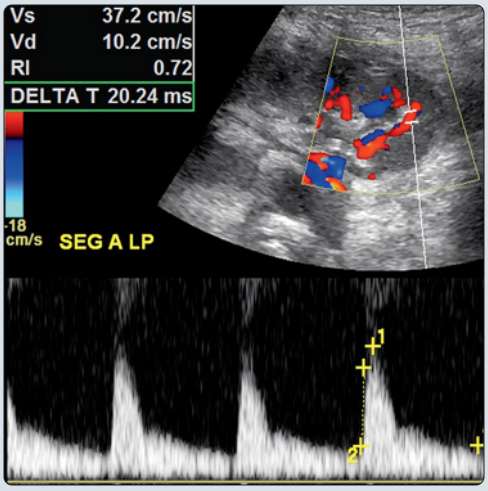
- CTA is modality of choice for preoperative evaluation of potential renal donors
- Doppler ultrasound: Frontline modality for identification of potentially correctable causes of transplant dysfunction
  - Parenchymal disease (rejection, ATN, drug toxicity pyelonephritis, etc.): Clinical correlation and biopsy often needed given nonspecific ultrasound appearance
  - Collecting system dilatation
    - Mild nonobstructive pelvicaliectasis common: Reflux and denervated collecting system
    - Obstructive pelvicaliectasis due to stricture at ureteroneocystostomy, calculi, clot, fungus balls
  - Perinephric fluid collections
    - Hematoma, urinoma, lymphocele, abscess: Time interval post transplantation helps differentiate
    - Appearance often nonspecific and aspiration typically necessary for definitive diagnosis
  - Renal artery stenosis
    - Most common renal vascular complication: Unexplained graft dysfunction, hypertension
    - Color aliasing at renal arterial anastomosis: Renal arterial/external iliac artery velocity gradient (> 1.8)
  - Renal vein thrombosis
    - Anuria, graft swelling in immediate postoperative period
    - Doppler: Reversal of intrarenal arterial diastolic flow and absent renal venous flow
  - Pseudoaneurysm
    - Contained biopsy injury of intrarenal artery: "Yin-yang" color flow within intrarenal cyst; to-and-fro flow within neck
  - Arteriovenous fistula
    - Iatrogenic communication between intrarenal artery and vein post biopsy
    - Focal color flurry, high-velocity, "low-resistance" flow within feeding artery and pulsatile flow within draining vein

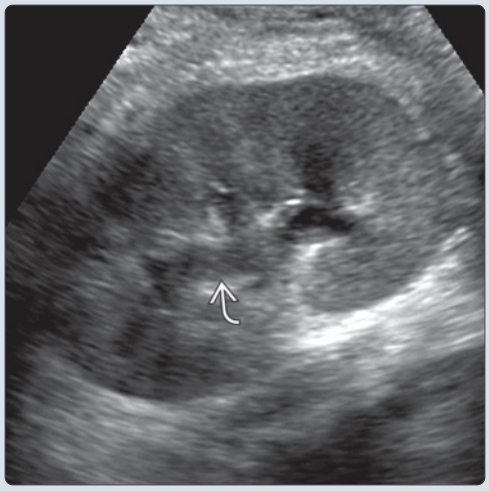
(Left) *Graphic shows typical renal transplant anatomy. The kidney is placed in the right iliac fossa in a retroperitoneal position. The ureter is tunneled through the dome of the bladder. The renal artery and vein are anastomosed end-to-side with the external iliac vessels of the recipient.* **(Right)** *A color Doppler map of vascular anatomy is initially created to identify potential vascular complications. Flow disturbances (aliasing) are quantified with pulse wave Doppler. Note end-to-side arterial and venous anastomoses ➔.*





(Left) *Resistive indices are obtained from segmental arteries. An ↑ RI (> .8) is a nonspecific finding of renal dysfunction due to ↓ tissue/vessel compliance (and not vascular resistance as implied in early publications).* **(Right)** *Ultrasound shows urothelial thickening ➔ and poor corticomedullary differentiation; features associated with acute rejection. Doppler also showed no diastolic flow. A biopsy confirmed rejection, though the grayscale (and Doppler) findings are neither specific or sensitive.*





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IMAGING

General Features

- Morphology
  - Allograft placed in extraperitoneal location, either iliac fossa
    - Right lower quadrant preferred: Easier dissection and longer, straighter course of right external iliac vein
    - Left lower quadrant utilized if 2nd transplantation, prohibitive RLQ vascular disease, or simultaneous pancreas-renal transplantation
    - Intraperitoneal placement rarely performed in pediatric population, or if multiple prior failed transplants
  - Arterial anastomosis
    - Cadaveric: End-to-side renal artery to external iliac artery
      - Carrel patch (harvested aortic cuff proximal to donor renal artery/arteries) may simplify arterial reconstruction
    - Living: Shorter donor renal artery typically available
      - End-to-side renal artery to external iliac artery if possible; end-to-end donor renal artery to internal iliac artery otherwise
  - Venous anastomosis
    - End-to-side to recipient external iliac vein
  - Ureter
    - Ureteroneocystostomy: Ureter is anastomosed through tunnel in dome of bladder
  - En bloc transplant
    - Both kidneys from infant donor given to adult recipient
    - Technically challenging, but outcomes are superior

Radiographic Findings

- Multidetector CT modality of choice for preoperative evaluation of potential recipient and donor
- Potential recipient imaging evaluation
  - NECT: Routinely utilized at some centers to qualitatively assess prohibitive iliac artery calcific plaque burden
  - CTA selectively employed to assess arterial inflow and potential flow-limiting aortoiliac stenoses
- Potential renal donor evaluation
  - Vascular (arterial, venous) and collecting system-ureteral anatomy defined
  - Multidetector (64-128) CT: Arterial (bolus tracking), nephrographic (70-second delay) and excretory (5-minute delay) phases obtained
    - Reconstructed axial (1.25-2.5 mm) images reviewed: Curved/straight MPR, MIP, VR 3D reconstructions for display
    - 3D processing: Renal lengths, volumes calculated
  - Parenchymal evaluation: Anomalies, calculi, masses, etc. identified
  - Renal arterial anatomy and variants
    - Number and types (accessor, polar) arteries described, distance between segmental-main renal arteries calculated
    - Potential contraindications to donor nephrectomy
      - > 2 renal arteries (though small polar artery may be sacrificed)
      - Perihilar branching (right renal artery: Retrocaval branching; left renal artery: Segmental branching < 1-1.5 cm from left renal artery origin)
      - Renal arterial disease: Atherosclerosis, fibromuscular dysplasia
  - Renal venous anatomy and variants
    - Number, branching pattern, anomalies (e.g., retroaortic, circumaortic left renal veins) reported
    - Large (> 5 mm) renal vein tributaries (lumbar, gonadal, etc.) identified: Identification critical because of limited visual FOV of laparoscopic nephrectomy
  - Upper urinary tract assessment: Complete or partial ureteral duplication not absolute contraindications for donation, though more technically challenging
- Renal transplant evaluation
  - Doppler ultrasound is premier modality (other modalities as adjunct or confirmatory test)
  - Protocol
    - Grayscale examination: Renal parenchyma and length, collecting system, perinephric fluid collections
      - If collecting system dilatation present, grade (mild, moderate, severe) pre- and post bladder voiding
      - Improvement after voiding implies nonobstructive pelvicaliectasis and reflux
    - Color Doppler vascular (renal: External iliac artery, vein) map and screen
      - Quantify areas of flow disturbance (aliasing) with pulse wave
      - Optimize parameters: Greatest gain without background noise, lowest pulse repetition frequency without aliasing
      - Segmental arterial resistive indices (RI): Peak systolic velocity: End diastolic velocity/peak systolic velocity
  - Renal parenchymal abnormalities
    - Reported grayscale findings of rejection: Globular, enlarged kidney, urothelial thickening, loss of corticomedullary differentiation, effacement of central sinus
      - Not specific or sensitive: Ultrasound-guided biopsy routinely performed to differentiate between causes of transplant dysfunction (acute/chronic rejection, ATN, drug toxicity)
      - Elevated RI (> .8): Nonspecific finding of graft dysfunction, due to diminished tissue/vascular compliance
    - Ultrasound manifestations of renal abscess, renal cell carcinoma similar to native kidneys
    - Renal post-transplantation lymphoproliferative disease: Infiltrating renal parenchyma tumor, often within renal pelvis
  - Collecting system dilatation
    - Mild dilatation common: Denervated collecting system, reflux
    - Correctable causes of obstructive pelvicaliectasis: Ureteroneocystostomy stricture, perinephric fluid collections, calculi, clot, fungus balls
  - Perinephric fluid collections
    - Largely nonspecific appearance, time interval post transplantation may narrow differential diagnosis, though sampling often required

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- Hematoma: Small elliptical hematomas common in immediate postoperative period: Acute hematomas post transplantation/biopsy may be echogenic, inconspicuous despite large size
- Urinoma: Anechoic collection weeks/months post transplant: Nuclear medicine study (DTPA, MAG3) may confirm leak
- Lymphocele: Occurs several months post transplantation, due to disrupted pelvic lymphatics: Anechoic collection, thin septations
- Abscess: Rare, complex perinephric collection: Clinical history (fever, sepsis, leukocytosis) aids diagnosis

- Vascular complications: Large initial series, prevalence rate of 10% reported
  - Renal artery stenosis
    - Most common vascular complication, typically 1-3 months post transplantation
    - Clinical manifestations: Unexplained graft failure, hypertension
    - Stenosis usually at anastomosis, though inflow iliac stenosis (due to clamp injury, atherosclerosis) present similarly
    - Color Doppler screen: Focal color aliasing
    - Pulse wave Doppler: Peak systolic value > 250 cm/sec, renal artery: ipsilateral external iliac artery ratio > 1.8
    - Distal waveform changes (tardus-parvus: Delayed systolic upstroke, resistive index < .5) often absent
    - If normal renal function, may confirm equivocal cases with MRA, CTA
    - Highly suggestive Doppler examination should directly prompt catheter angiography for diagnosis and management (angioplasty, stent)
  - Renal artery thrombosis
    - Rare, typically immediate postoperative complication (kink/torsion renal artery, arterial dissection, rejection, acute tubular necrosis, hypercoagulable state)
    - Absent intrarenal Doppler flow
    - Prompt thrombectomy, arterial revision may be attempted
  - Renal vein stenosis
    - Often incidental Doppler finding: Aliasing at renal venous anastomosis and 2-3 fold velocity gradient
    - Due to kink, perianastomotic fibrosis, perivascular fluid collections
    - Consider venography if unexplained graft dysfunction after biopsy
  - Renal vein thrombosis
    - Rare, immediate postoperative complications: Abrupt anuria, graft swelling
    - Doppler: Reversal of intrarenal diastolic flow and absent renal venous flow (diastolic flow reversal also shown with severe acute rejection, ATN)
  - Pseudoaneurysm (PA)
    - Intrarenal PA typically due to arterial injury post biopsy
    - Often asymptomatic, though may rupture into collecting system, perinephric space: Large, growing PAs treated by coil embolization
    - Color Doppler: "Yin-yang" flow within intrarenal "cyst," to and fro flow within neck
    - Extrarenal PA: Usually perianastomotic and may be mycotic (surgical revision necessary)
  - Arteriovenous fistula (AVF)
    - Iatrogenic communication of artery and vein post biopsy
    - Focal color "flurry" due to tissue vibration
    - High-velocity, "low-resistance" (high diastolic) flow within artery and pulsatile flow within draining vein
    - Large AVFs may lead to diminished perfusion and ischemia of other portions of kidney: Coil embolization required

- Post-transplant malignancies
  - Recipients have much higher prevalence of various malignancies
  - Post-transplant lymphoproliferative disorder (PTLD)
    - Associated with Epstein-Barr viral infection
    - Occurs in 1% of renal allograft recipients
    - Imaging: Lymphadenopathy, masses in solid organs ± bowel
    - May affect allograft itself (especially near hilum)
  - Renal and transitional cell (urothelial) carcinoma
    - Higher prevalence of both in transplant recipients
    - Can occur in allograft or native kidneys
      - Especially papillary RCC in native kidneys with acquired cystic kidney disease of uremia

CLINICAL ISSUES

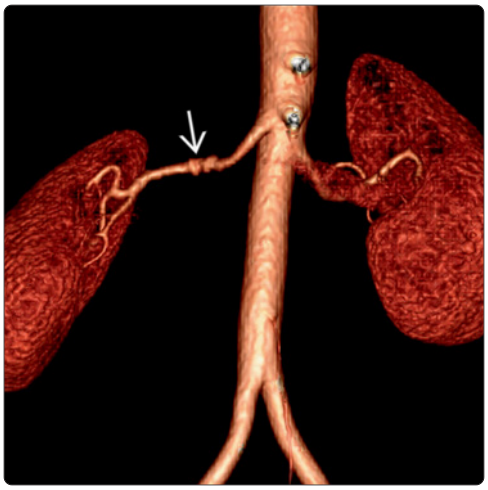
Natural History & Prognosis

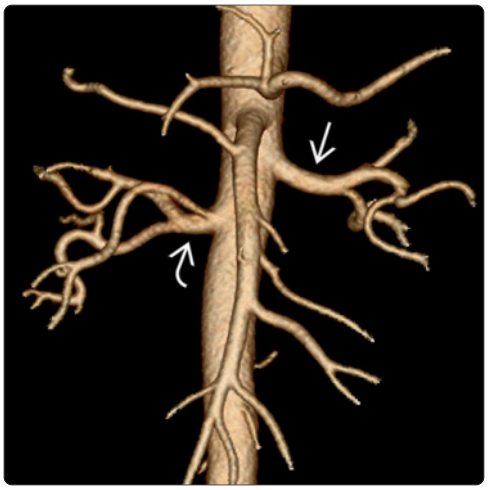
- Renal transplantation is treatment of choice for irreversible renal failure
  - Lower morbidity and mortality than for patients managed with chronic dialysis
  - Advances in surgical techniques, immunosuppressive medications, and identification of potentially correctable cause of transplant function by imaging have improved survival and quality of life
  - OPTN data (4/15): 5-year graft survival of cadaveric and living renal transplants 66.6 and 79.8%, respectively

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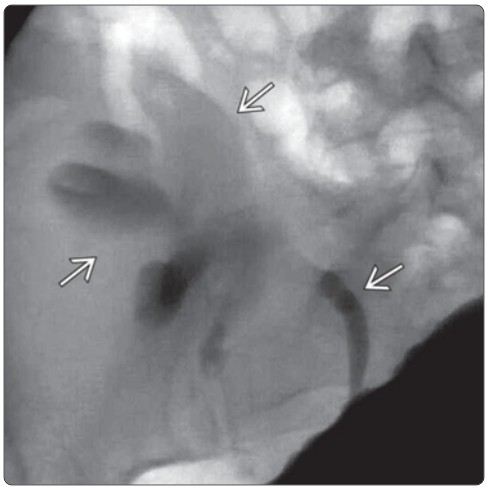
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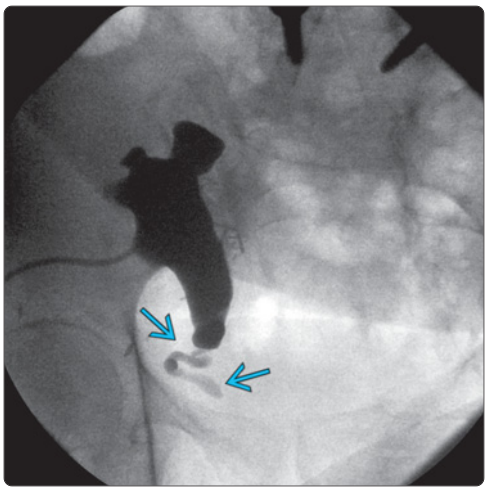
(Left) *Volume rendered renal CT angiogram performed on a young female potential renal donor shows a beaded mid right renal artery, a feature characteristic of fibromuscular dysplasia.* **(Right)** *Volume rendered CTA performed on a potential donor shows a single left renal artery and a perihilar branching pattern of the right renal artery. The number, location, and branching pattern of renal arteries and veins should be detailed so that prohibitive anatomy is not unexpectedly encountered during laparoscopic organ harvesting.*





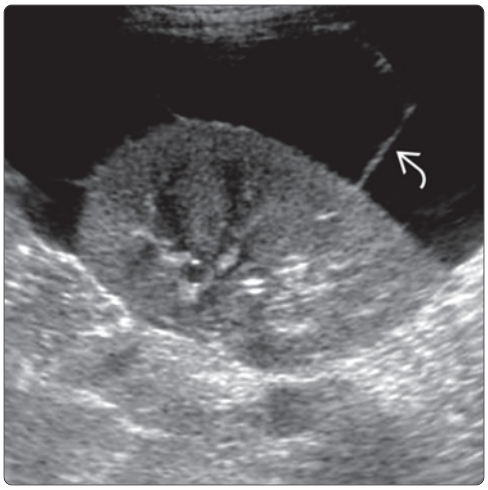
(Left) *Ultrasound performed on a patient with graft dysfunction shows marked hydronephrosis. It is often impossible to differentiate between obstructive and nonobstructive pelvicaliectasis, but improvement in hydronephrosis after bladder emptying suggests reflux (as in this case).* **(Right)** *A cystogram performed on the same patient confirms marked reflux. Mild pelviectasis is often seen post transplantation due to reflux and collecting system denervated.*

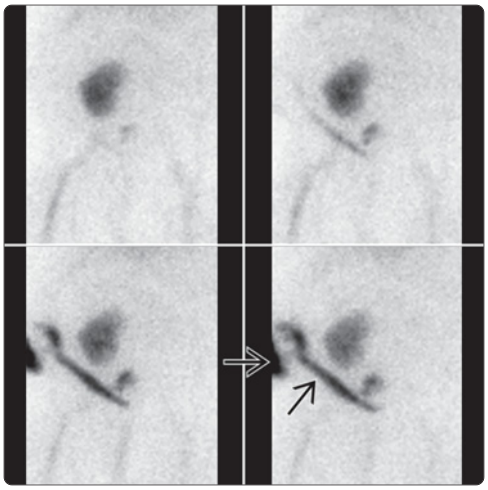




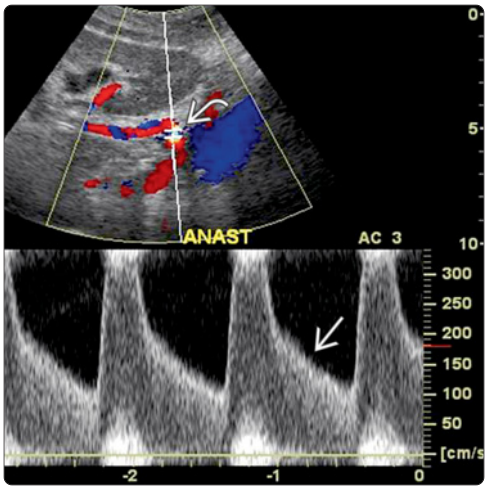
(Left) *Renal transplant ultrasound shows marked hydronephrosis. Collecting system dilatation persisted after bladder voiding. An ↑ RI may be ancillary evidence of true obstruction, but the parameter is not sensitive or specific.* **(Right)** *An antegrade nephrostogram performed on the same patient shows a long stricture of the distal ureter at and proximal to the ureteroneocystostomy, likely due to perianastomotic fibrosis. Other causes of ureteral obstruction include calculi, clots, and periureteral fluid collections.*

(Left) *Ultrasound performed 4 months post transplantation shows a large, anechoic, perinephric fluid collection. The time course and septation suggest a lymphocele, but aspiration is needed for definitive diagnosis.* **(Right)** *Sequential planar Tc-99m MAG3 in a patient with graft dysfunction and a perinephric fluid collection 1 month post transplantation shows tracer activity outside of the bladder, which tracks to the incision. The ureteroneocystostomy was subsequently revised.*



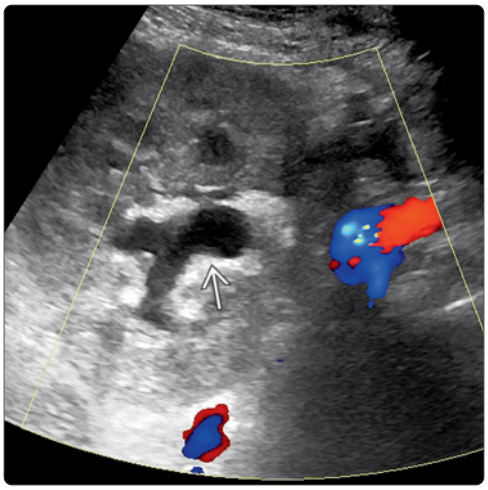


(Left) *Renal Doppler was performed for graft dysfunction and hypertension. The Doppler gate is placed on an area of color aliasing. Pulse wave Doppler shows anastomotic velocities of > 3.5m/s and marked diastolic broadening, features compatible with a high-grade arterial stenosis. Ancillary evidence included ↓ intrarenal RIs (< .5) and tardus parvus spectra.* **(Right)** *DSA performed on the same patient confirms a tight anastomotic stenosis, which was subsequently angioplastied.*

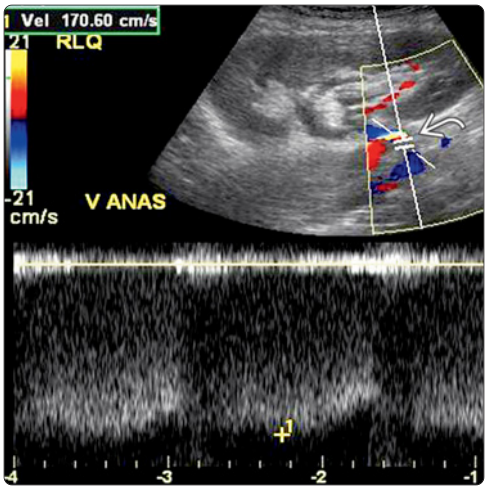


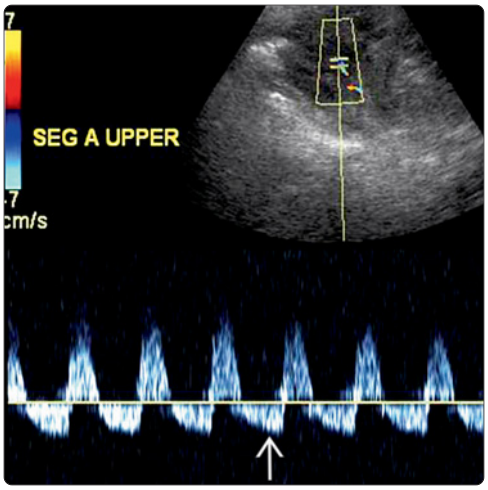


(Left) *Doppler ultrasound performed on an acutely anuric patient 2 years post renal transplantation shows mild hydronephrosis. No transplant flow was identified despite optimized technique; a renal artery thrombosis was suggested despite the time interval post transplantation.* **(Right)** *Emergent CT of the same patient shows an enlarged, transversely oriented allograft. A torsed transplant (a rare transplant complication) was identified at exploration. Complete graft recovery occurred after detorsion.*

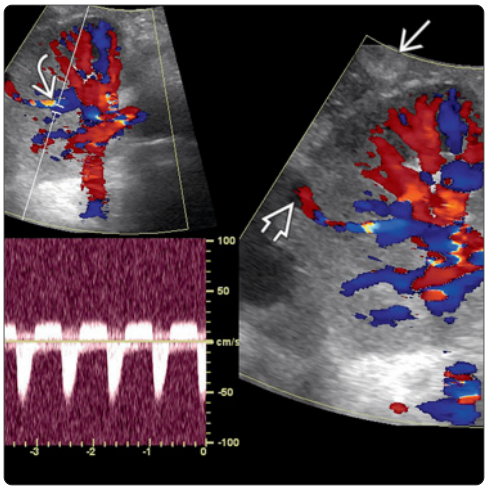






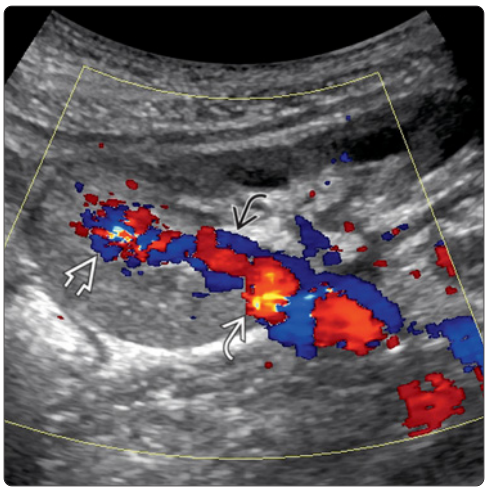


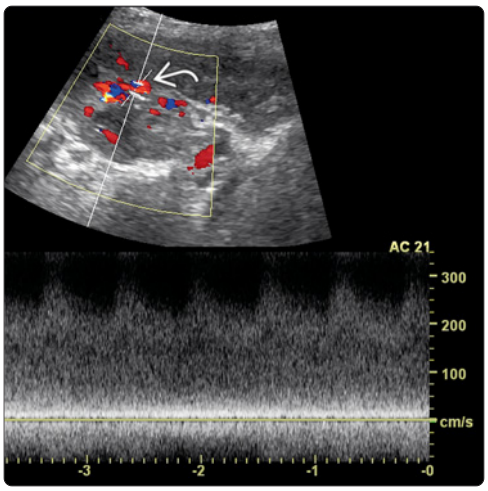
(Left) *Doppler interrogation of focal color aliasing at the venous anastomosis shows markedly ↑ velocities. A 2- to 3-fold gradient suggests a venous stenosis, but this is typically clinically insignificant. Venography can be performed if graft dysfunction is unexplained.* **(Right)** *Doppler interrogation of a segmental artery in an anuric patient 1 day post transplantation shows reversed diastolic flow. Renal venous flow was also absent, and nephrectomy was performed after unsuccessful thrombectomy.*





(Left) *Color Doppler ultrasound performed in a patient with pain and a ↓ hematocrit post biopsy shows echogenic perinephric clot and a small pericapsular pseudoaneurysm (PA). Interrogation of the neck shows to-and-fro flow.* **(Right)** *DSA during an ultimately successful embolization procedure on the same patient shows an extrarenal PA. PAs are typically due to needle biopsy. Small, incidental intrarenal PAs may resolve over time; enlarging or extrarenal PAs should be embolized.*





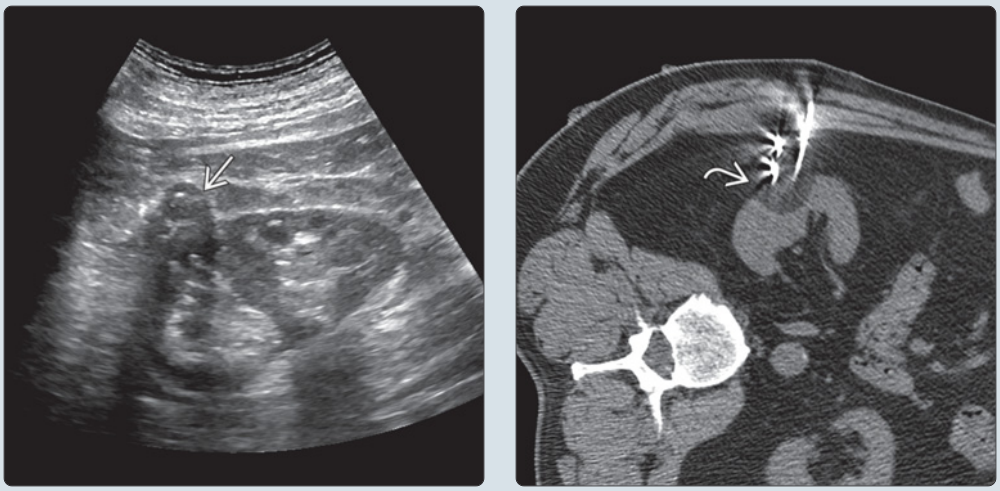
(Left) *Color Doppler ultrasound performed on a patient with renal insufficiency 3 weeks post biopsy shows a focal flurry (due to tissue vibration at an arteriovenous fistula), a feeding artery, and a draining vein.* **(Right)** *Doppler interrogation of the feeding artery shows high-velocity systolic flow and "low-resistance" diastolic flow. Pulsatile flow was shown in the draining vein. Preferential shunting of flow to the AVF may result in tissue ischemia. Embolization was performed because of size of the AVF.*

KEY FACTS

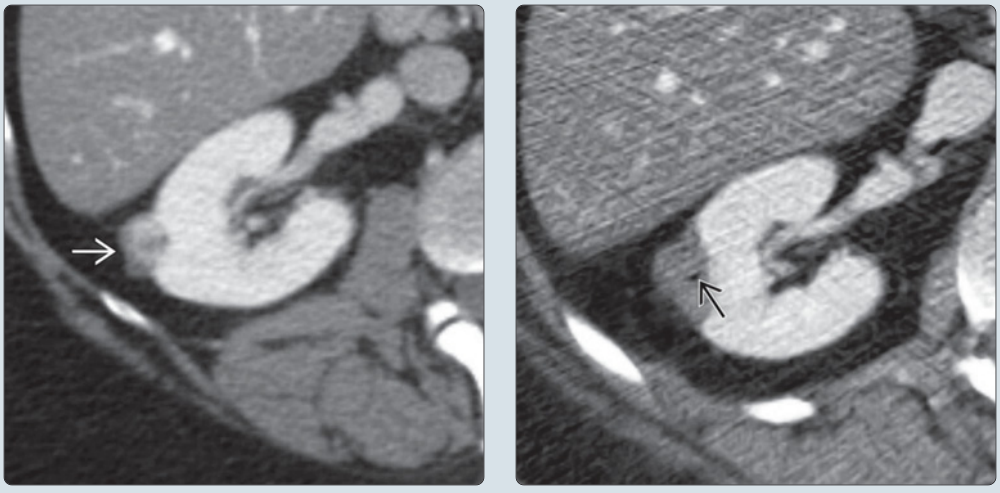
TOP DIFFERENTIAL DIAGNOSES

- **Total nephrectomy**
  - **Radical nephrectomy**: Total nephrectomy with excision of adrenal gland and regional lymph nodes (performed for malignant tumors)
  - Indications
    - High-grade traumatic injury: Such as devascularization, schattered kidney
    - Irreversible kidney damage in setting of chronic infection, symptomatic nephrolithiasis, congenital causes
    - Nonfunctioning kidney resulting in renovascular hypertension
    - Kidney donor
    - Large, locally advanced malignant tumors
- **Partial nephrectomy**
  - Resection of part of kidney, a.k.a. nephron-sparing surgery
  - Performed for both benign and neoplastic conditions
    - **Trauma**: In setting of low-/intermediate-grade traumatic renal injury; to control hemorrhage or urinary leak
    - **Nephrocalcinosis**: To remove large stones and infected tissue
    - **Infection**: Such as xanthogranulomatous pyelonephritis affecting portion of kidney
    - **Benign tumors**: Such as renal angiomyolipoma
    - **Malignant tumors**
  - Site of resection is often packed with fat tissue, which can resemble fat-containing lesion on CT
- **Renal tumor ablation**
  - Radiofrequency (heat) or cryoablation (freezing)
    - Both are effective in eradication of small tumors
  - Morbidity is lower than surgery, although, recurrence rate is slightly higher
- **Urinary catheters and stents**

(Left) *Longitudinal US image through the right kidney demonstrates an exophytic isoechoic mass. The age of the patient and multiple comorbidities prompted biopsy and cryoablation rather than partial nephrectomy. Biopsy was positive for papillary type renal cell carcinoma.* **(Right)** *CT image of the same patient obtained during percutaneous cryoablation procedure shows formation of an ice ball around the cryoablation probes.*



(Left) *Axial CECT though the right kidney shows a small renal mass with characteristics of a renal cell carcinoma. Following confirmation by needle biopsy, this lesion was treated by cryoablation.* **(Right)** *Axial CECT, following cryoablation, shows post-treatment changes with no residual enhancing tissue seen. Note the small bubble of gas at the cryoablation site, which should not be considered indicative of infection in this setting.*



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TERMINOLOGY

Definitions

- Anatomical changes following variety of interventions and operations on kidneys

DIFFERENTIAL DIAGNOSIS

Total Nephrectomy

- Removal of entire kidney
  - **Radical nephrectomy**: Total nephrectomy with excision of adrenal gland and regional lymph nodes (performed for malignant tumors)
- Indications
  - High-grade traumatic injury: Such as devascularization, schattered kidney
  - Irreversible kidney damage in setting of chronic infection, symptomatic nephrolithiasis, congenital causes
  - Nonfunctioning kidney resulting in renovascular hypertension
  - Kidney donor
  - Large, locally advanced malignant tumors
- Compensatory hypertrophy of contralateral kidney is seen
- Renal fossa is filled with adjacent organs
- Complications
  - Hematoma, bowel injury, splenic injury
  - Abdominal abscess
    - Thick and irregular wall enhancement with surrounding stranding
  - Postoperative lymphocele
    - Round or ovoid fluid density without surrounding fat stranding
  - Abdominal wall hernia

Partial Nephrectomy

- Resection of part of kidney, a.k.a. nephron-sparing surgery
- Performed for both benign and neoplastic conditions
  - **Trauma**: In setting of low-/intermediate-grade traumatic renal injury; to control hemorrhage or urinary leak
  - **Nephrocalcinosis**: To remove large stones and infected tissue
  - **Infection**: Such as xanthogranulomatous pyelonephritis affecting portion of kidney
  - **Benign tumors**: Such as renal angiomyolipoma
  - **Malignant tumors**
    - Usually reserved for small, low-stage renal cell carcinoma
      - Particularly in patients with diminished renal function, solitary kidney, or bilateral renal malignancy
    - Imaging findings predictive of favorable outcome following partial nephrectomy
      - Size < 3 cm; peripheral location of tumor with lack of invasion into renal sinus and collecting system
    - In selected cases with large size of tumor or presence of distant metastasis, partial nephrectomy can still be performed
- Site of resection is often packed with fat tissue
  - Packed fat may resemble fat-containing lesions (such as angiomyolipoma) on CT
  - Look for surgical clips or other signs or history of surgery
- Absorbable hemostat material (such as Gelfoam or Surgicel) may be placed at surgical resection site to control bleeding
  - In immediate postoperative period surgical gelatin often appears tightly packed, linear collections of gas bubbles that may be mistaken for abscess
  - Usually with very little fluid and no enhancing rim
  - Correlation with operative history is crucial
- Complications
  - Urinary leak, bleeding, bowel injury, splenic injury
  - Renal insufficiency: Mainly due to acute tubular necrosis (ATN) secondary to clamping of hilar vessels
  - Positive margins with tumor recurrence
  - Chyluria: Rare complication due to connection between lymphatics and collecting system

Renal Tumor Ablation

- Radiofrequency (heat) or cryoablation (freezing)
  - Both are effective in eradication of small tumors
  - Morbidity is lower than surgery, although, recurrence rate is slightly higher
    - Usually preserved with patients who are not surgical candidates
  - Can be performed percutaneously or laparoscopically
  - May cause release of gas at site of ablation from sudden death of tumor mass

Urinary Catheters and Stents

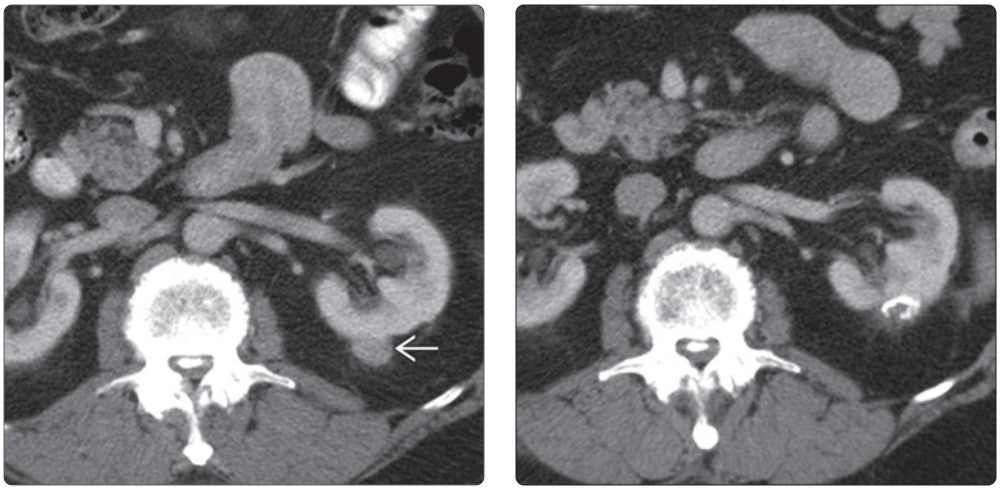
- Ureteral stents
  - Common indications: Ureteral obstruction/stricture; low-grade ureteral injury; following ureteral reconstruction; following lithotripsy
  - May result in injury to ureter, including perforation
  - May introduce gas into renal collecting system, which may be mistaken for infectious emphysematous pyelitis
- Percutaneous nephrostomy
  - Used to treat hydronephrosis or pyonephrosis
  - Used to access renal pelvis for direct lithotripsy
  - May introduce gas &/or hemorrhage into collecting system
    - May be mistaken for emphysematous pyelonephritis or pyelitis

SELECTED REFERENCES

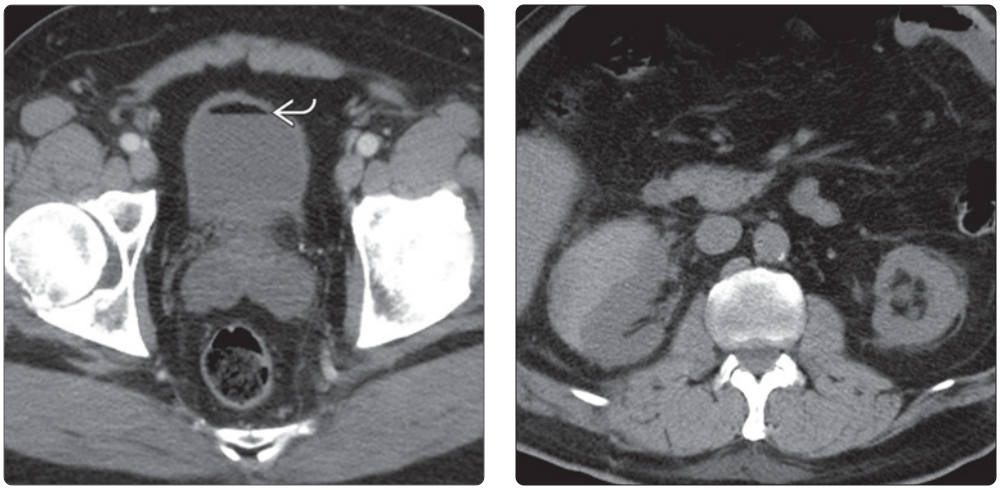
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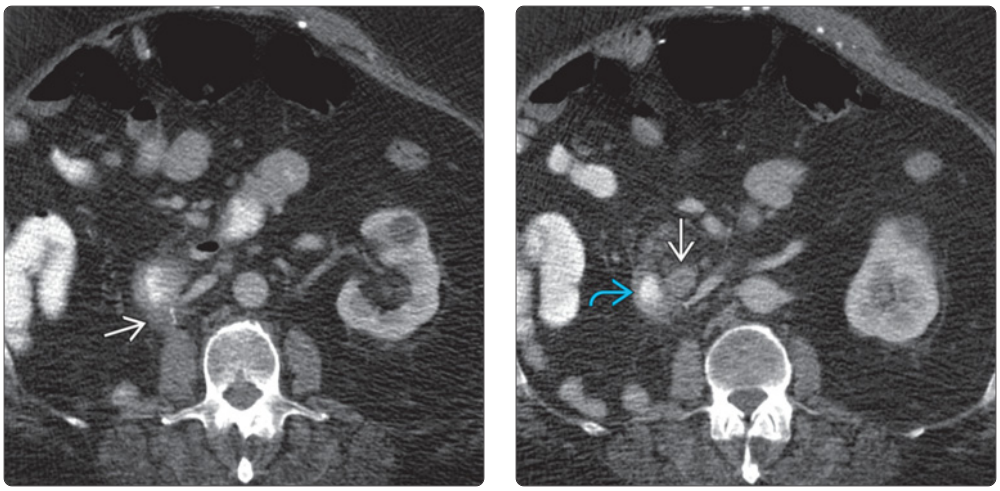
(Left) *Axial CT through the kidneys demonstrates an enhancing exophytic mass in left kidney highly suspicious for renal cell carcinoma.* **(Right)** *The patient underwent laparoscopic nephron-sparing surgery. Note the surgical clips and mild stranding at the resection site.*

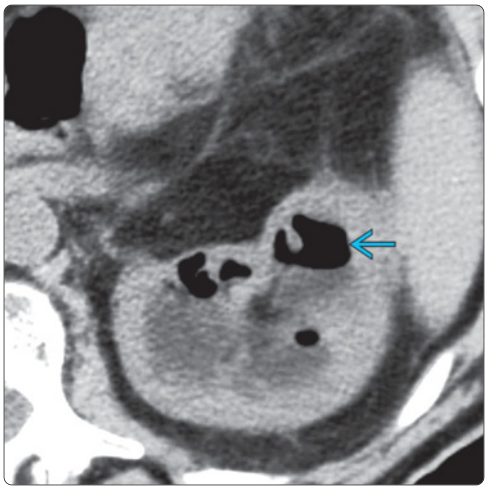


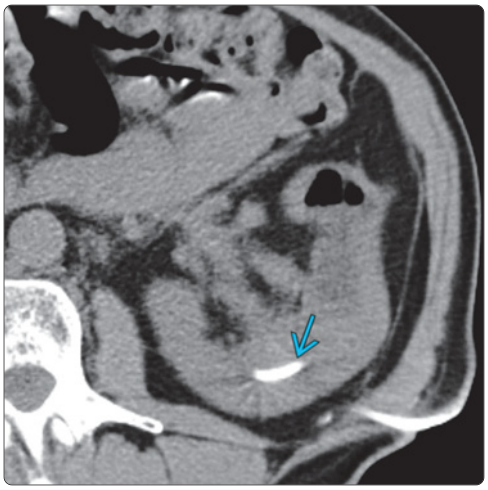
(Left) *Axial CT through the pelvis in the same patient shows fat-fluid level in the urinary bladder compatible with chyluria, an uncommon complication of partial nephrectomy.* **(Right)** *Axial CT through the kidneys, obtained 2 days following right partial nephrectomy for clear-cell type renal cell carcinoma, shows a large subcapsular hematoma with significant mass effect on the kidney. The patient was taken back to the OR for drainage of the hematoma.*



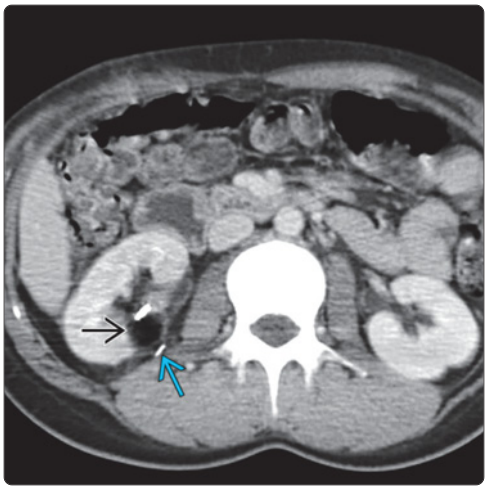
(Left) *Axial CT in 72-year-old man with history of right radical nephrectomy for clear-cell type RCC shows soft tissue adjacent to the oversewn venous stump. This finding is highly suspicious for local recurrence of tumor.* **(Right)** *A higher section CT image shows an enlarged regional precaval lymph node . Note posterior displacement of the duodenum post radical nephrectomy.*





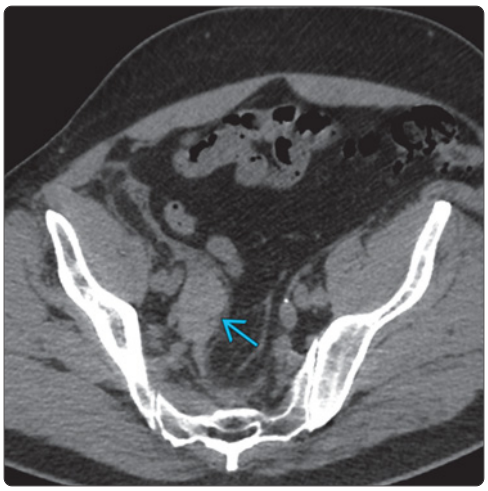


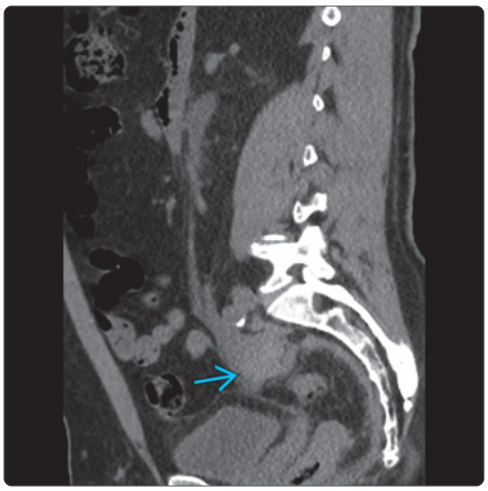
(Left) *Axial NECT shows the left renal collecting system distended with fluid and gas ⇨; a finding which can be seen in the setting of emphysematous pyelitis.* **(Right)** *Axial CECT through a lower section in the same patient shows a percutaneous nephrostomy tube ⇨, which has been placed to treat hydronephrosis. The urine was sterile and the gas had been introduced along with the nephrostomy catheter.*





(Left) *Axial CECT shows a fat-density structure ⇨ within the right kidney. The presence of an adjacent surgical clip ⇨ is a clue that this is an iatrogenic lesion, due to deliberate placement of fat into the renal defect left at the time of surgery to prevent a urine and blood leak.* **(Right)** *Retrograde pyelogram performed during stone extraction shows opacification of the right ureter. A ureteral stent was placed after completion of the procedure. The patient presented with flank pain after removal of stent.*





(Left) *Axial CT images in the same patient shows a moderate-size retroperitoneal hematoma ⇨ along the course of the distal ureter, presumably due to iatrogenic injury to the ureter.* **(Right)** *Sagittal CT image better demonstrates the retroperitoneal hematoma ⇨. A ureteral stent was placed and the patient was managed conservatively.*

Radiation Nephritis

KEY FACTS

TERMINOLOGY

- Renal injury caused by ionizing radiation

IMAGING

- Sharply demarcated area of decreased attenuation and parenchymal atrophy on CECT (corresponding to irradiated portion of kidney)
- Partial kidney radiation
  - Irradiated portion is sharply demarcated and has smooth surface
- Whole kidney radiation
  - Progressive atrophy of kidney (most pronounced in outer cortex)
  - Bilateral radiation nephritis is indistinguishable from other causes of chronic renal failure

TOP DIFFERENTIAL DIAGNOSES

- Pyelonephritis
  - Usually has wedge-shaped or striated nephrogram & perirenal infiltration
- Renal infarction
  - Usually has wedge-shaped or global distribution
  - Cortical rim sign
- Chronic renal failure (other causes)

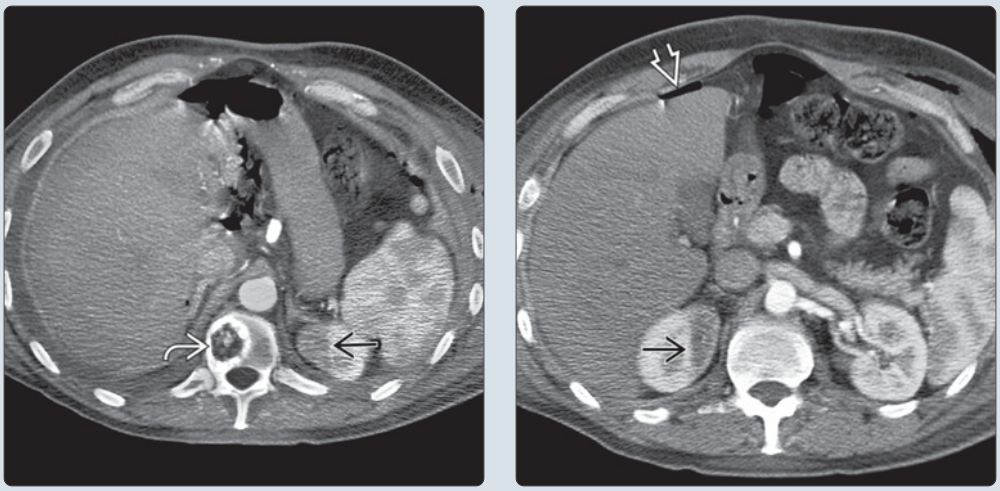
PATHOLOGY

- Etiology: Ionizing radiation → oxidative injury to DNA → cell damage/death
- **Total dose** and **volume** of irradiated kidney are most important predictors of degree of renal injury
- Patients receiving total dose of > 20 Gy (2000 rad) have highest risk (when kidneys included in field of radiation)

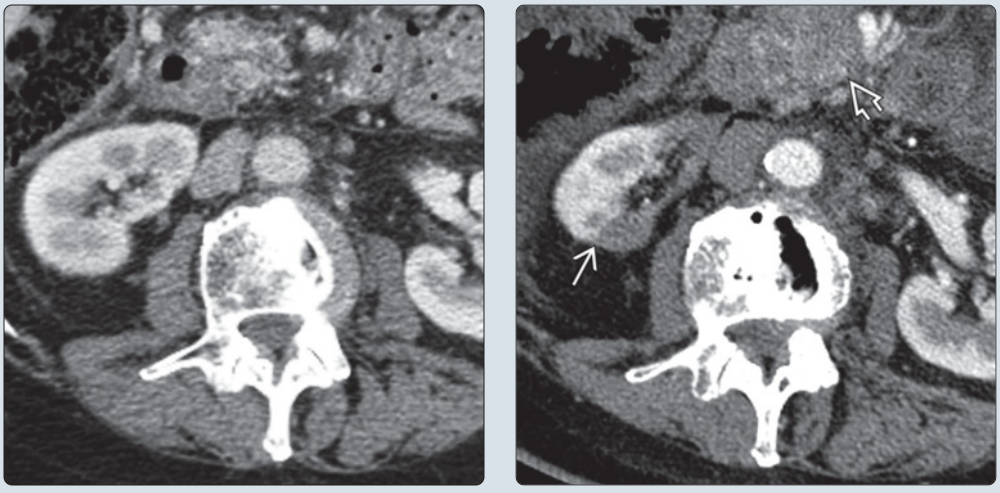
CLINICAL ISSUES

- Only 20% of patients receiving > 25 Gy to kidneys develop renal dysfunction
- Proteinuria, azotemia, and hypertension are the most common signs/symptoms

(Left) *Middle-aged man is shown post recent resection of colon carcinoma and radiation therapy for a vertebral metastasis. Axial CECT shows a sharply demarcated hypoenhancing area in the left kidney. Note the vertebral metastasis that was the target of the radiation therapy.* **(Right)** *A more caudal section in the same patient shows evidence of the radiation nephritis in the right kidney as well. Note the free air related to recent laparotomy.*



(Left) *This 72-year-old woman with history of unresectable pancreatic adenocarcinoma underwent external beam radiation. Axial CECT through the kidneys, 5 months following radiation, does not show any discernible changes.* **(Right)** *Repeat imaging 15 months following external beam radiation shows atrophy and markedly decreased perfusion of the medial aspect of the right kidney along the radiation port. Note sharp demarcation between the normal and affected parts of the kidney. Note the pancreatic mass.*



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TERMINOLOGY

Synonyms

- Radiation nephropathy, radiation-associated renal injury

Definitions

- Renal injury caused by ionizing radiation

IMAGING

General Features

- Best diagnostic clue
  - Sharply demarcated area of decreased attenuation and parenchymal atrophy on CECT (corresponding to irradiated portion of kidney)
- Morphology
  - Partial kidney radiation
    - Irradiated portion is sharply demarcated and has smooth surface
    - Does not correspond to wedge or pyramidal shape of renal lobules
  - Whole kidney radiation
    - Progressive atrophy of kidney (most pronounced in outer cortex)
    - Bilateral radiation nephritis is indistinguishable from other causes of chronic renal failure

Ultrasonographic Findings

- Atrophy and increased echogenicity of irradiated portion (or whole kidney if entire kidney was irradiated)
  - In patients with history of whole body irradiation, both kidneys become progressively small and echogenic; indistinguishable from other causes of chronic renal failure
- Compensatory hypertrophy of nonradiated kidney

CT Findings

- Partial kidney radiation
  - Early phase CECT: Sharply demarcated area with ↓ attenuation corresponding to radiation portal
  - Delayed (pyelographic) phase CECT: Persistent nephrogram in damaged portion
  - Cortical ± medullary atrophy of affected portions of kidneys
- Whole kidney radiation
  - Atrophy and delayed nephrogram in affected kidney

Imaging Recommendations

- Best imaging tool
  - CECT

DIFFERENTIAL DIAGNOSIS

Pyelonephritis

- Usually has wedge-shaped or striated nephrogram
- Often has perirenal infiltration

Renal Infarction

- Usually has wedge-shaped or global distribution
- Cortical rim sign
  - Enhanced rim of cortical and capsular tissue
  - Seen only in setting of subacute renal ischemia or infarction

Chronic Renal Failure (Other Causes)

- Bilateral radiation nephritis, due to total body irradiation, is indistinguishable from other causes of chronic renal failure

PATHOLOGY

General Features

- Etiology
  - Ionizing radiation → oxidative injury to DNA → cell damage/death
- All components (including glomeruli, tubules, vessels, and interstitium) are affected
- Risk factor
  - **Total dose** and **volume** of irradiated kidney are most important predictors of degree of renal injury
  - Patients receiving total dose of > 20 Gy (2000 rad) have highest risk (when kidneys included in field of radiation)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic for renal injury
    - Only 20% of patients receiving > 25 Gy to kidneys develop renal dysfunction
  - Proteinuria, azotemia, hypertension
    - Proteinuria is earliest sign

Demographics

- Mostly seen in patients receiving large therapeutic doses of external beam radiation
  - Also reported in patients receiving low-dose total body radiation prior to bone marrow transplant
  - Less commonly seen in patients treated with systemic radionuclides
- Children are more vulnerable

Treatment

- Conservative management of hypertension and renal failure (similar to other causes of chronic renal failure)
- Prophylaxis
  - Use of minimum effective dose
  - Selective shielding

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Contrast-Induced Nephropathy

KEY FACTS

TERMINOLOGY

- Acute kidney injury (AKI) within 24-48 hours following intravascular administration of contrast material (CM); must exclude other causes of AKI
  - AKI: Absolute increase in serum creatinine of 0.5 mg/dL or relative 25% increase from baseline value

IMAGING

- Delayed nephrogram on subsequent CT or KUB

TOP DIFFERENTIAL DIAGNOSES

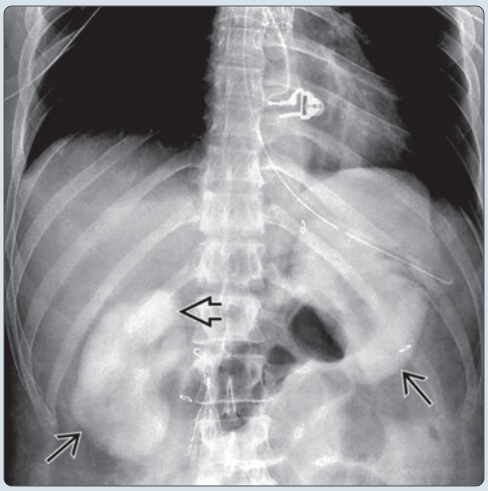
- Acute kidney injury (AKI)
  - Other causes of AKI such as nephrotoxic drugs, prerenal azotemia, urinary obstruction, etc.
  - It may be difficult to recognize exact cause of acute tubular necrosis (ATN) in inpatients since they are exposed to multiple risk factors
- Acute interstitial nephritis
- Renal atheroembolic phenomena

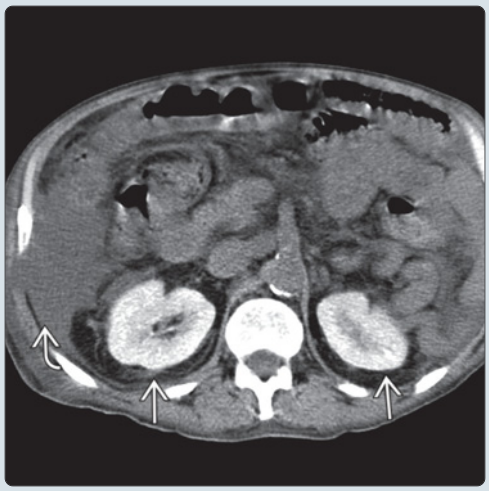
PATHOLOGY

- Animal models have indicated that **ATN** is underlying pathogenesis but mechanism by which ATN occurs is not well understood
  - **Renal vasoconstriction**
  - **Direct tubular injury**
- Risk of CIN is deterministic (dose dependent)

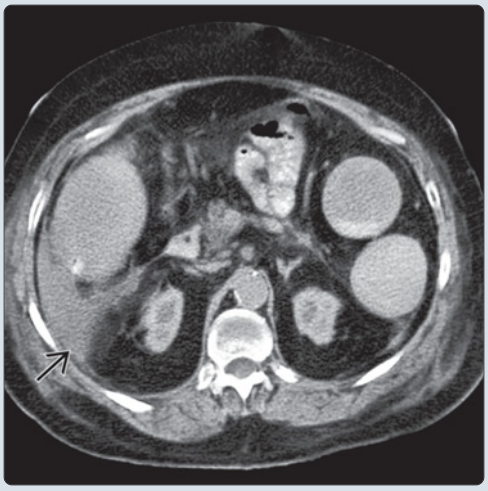
CLINICAL ISSUES

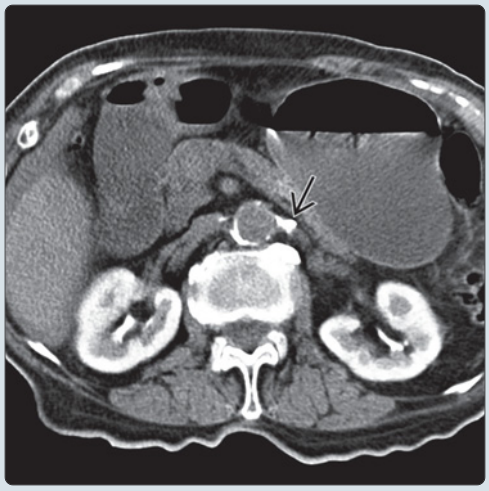
- **Prophylaxis**
  - Consider alternative imaging methods not requiring CM
  - Use lower dose of CM; avoid closely spaced contrast-enhanced studies
    - Hypo-osmolar and iso-osmolar nonionic CM have been shown to be superior to hyperosmolar ionic agents
  - Intravenous hydration
  - N-acetylcysteine, orally or IV
  - Discontinuing nephrotoxic drugs
  - **No** benefit in prophylactic hemodialysis





(Left) *Radiograph taken 18 hours after a coronary angiogram shows persistent nephrograms, a finding indicating acute tubular necrosis. The densely opacified bile within the gallbladder is due to vicarious excretion of contrast through the hepatobiliary system to compensate for the decreased renal excretion.* **(Right)** *Axial NECT shows dense bilateral nephrograms with otherwise normal-appearing kidneys in a patient with acute renal failure following angiography. Ascites is also present.*





(Left) *Axial NECT shows persistent enhancement of the kidneys in a patient with CIN following angiography. The ascites measured 60 HU due to vicarious excretion of contrast medium from the peritoneum to compensate for failed renal excretion.* **(Right)** *Axial NECT shows dense persistent nephrograms in this 92-year-old woman 3 days after a contrast study. She had baseline decreased renal function. Note the marked osteal calcification of renal arteries. Decreased GFR and advanced age are both risk factors for CIN.*

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TERMINOLOGY

Abbreviations

- Contrast-induced nephropathy (CIN)

Definitions

- Acute kidney injury (AKI) within 24-48 hours following intravascular administration of contrast material (CM); must exclude other causes of AKI
  - AKI: Absolute increase in serum creatinine of 0.5 mg/dL or relative 25% increase from baseline value

IMAGING

General Features

- Best diagnostic clue
  - Nonspecific imaging findings
  - Delayed nephrogram on subsequent CT or KUB

DIFFERENTIAL DIAGNOSIS

Acute Kidney Injury (AKI)

- Other causes of AKI, such as nephrotoxic drugs, prerenal azotemia, urinary obstruction, etc.
- It may be difficult to recognize exact cause of acute tubular necrosis (ATN) in inpatients since they are exposed to multiple risk factors
  - In large retrospective study (Newhous et al, AJR 2008) of 32,161 inpatients who were not exposed to CM, > 50% of them developed AKI

Acute Interstitial Nephritis

- History of drug hypersensitivity
- White blood cell casts on urinalysis

Renal Atheroembolic Phenomena

- Decline in renal function after angiography secondary to renal infarction in setting of thromboembolic and atheroembolic phenomena
- Its onset is usually delayed for days to weeks; protracted course with little or no recovery

PATHOLOGY

General Features

- Etiology
  - Animal studies have shown that ATN is critical unifying precipitating event, but mechanism by which ATN occurs is not well understood
  - Hypothesized mechanisms include
  - **Renal vasoconstriction**
    - Renal vasoconstriction → medullary hypoxia (outer medulla is more susceptible to ischemia due to its high metabolic state)
    - Likely mediated through alterations in nitric oxide, endothelin, and adenosine pathways
      - Nitric oxide pathway is impaired in diabetes and heart failure, contributing to increased susceptibility to CIN in these populations
  - **Direct tubular injury**
    - Due to direct cytotoxicity and generation of oxygen free radicals
    - Tubular obstruction by intratubular precipitation of dye crystals
  - **Risk of CIN is deterministic (dose dependent)**
  - CIN almost never occurs if renal function is normal
  - Risk factors
    - **Underlying renal dysfunction** (glomerular filtration rate < 60 mL/min): Most important risk factor
    - Decreased renal perfusion: Heart failure, hypotension, or dehydration
    - Age > 70
    - Diabetes mellitus
    - Concomitant use of nephrotoxic drugs (NSAIDs, aminoglycosides, cyclosporine, etc.)
    - Multiple myeloma
    - **Procedure-related risk factors**
      - Large CM volume
      - High osmolarity ionic (1st generation) CM
      - Type of study: Much higher prevalence with coronary angiography

Microscopic Features

- Focal necrosis of proximal tubules, which is nonspecific for CIN
- Obstructing tubular casts

Types of CM

- Ionic: High-osmolar contrast media (HOCM); high osmolarity (1550-2100 mOsm/kg)
  - Normal serum osmolarity: 280-300 mOsm/kg
  - Diatrizoate, Metrizoate T
- Nonionic monomers: Low-osmolar contrast media (LOCM); lower osmolarity (500-880 mOsm/kg)
  - Iopamidol, iohexol, ioxilan, iopromide
- Nonionic dimers: Iso-osmolar contrast media (IOCM); iso-osmolar to serum (290 mOsm/Kg)
  - Iodixanol

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - ↑ serum creatinine, hyperkalemia, hyperphosphatemia, acidosis
  - Most patients are nonoliguric

Demographics

- Age
  - ↑ incidence in elderly due to ↓ ability to accommodate to oxidative injury
- Epidemiology
  - 3rd leading cause of acute renal failure in hospitalized patients
  - Incidence: Negligible (<1%) in patients without risk factors; ~ 10% in patients > 70 years old; can increase to 50% in patients with multiple risk factors
    - Decreased incidence of CIN in past decade might be due to use of nonionic iso-osmolar and hypo-osmolar CM
  - In general, risk of CIN after coronary angiography is much higher than after CT (due to comorbidities and the higher administered dose)

Natural History & Prognosis

- Serum creatinine rises within 24 hours, peaks in 2-3 days, and usually returns to baseline within 7-10 days
- Majority of cases have mild decline in GFR
  - In prospective study of 353 patients with moderate to severe renal dysfunction who underwent coronary angiography (Brar et al, JAMA 2008), < 1% had severe AKI requiring dialysis
- In retrospective study of 21,346 hospitalized patients (McDonald et al, Radiology 2014), CM was **not** found to be an independent risk factor for AKI, emergent dialysis, or 30-day mortality.

Treatment

- In patients at risk for CIN, serum Cr or estimated GFR should be measured before study and 24-48 hours after administration of CM
- **Prophylaxis**
  - Consider alternative imaging methods not requiring CM
  - Use lower dose of CM; avoid closely spaced contrast-enhanced studies
    - Hypo-osmolar and iso-osmolar nonionic CM have been shown to be less nephrotoxic than hyperosmolar ionic agents
  - Intravenous hydration
    - No consensus regarding optimal solution and rate of administration
      - Isotonic solutions were shown to be superior to hypotonic ones
      - Several studies have suggested that serum alkalinization with sodium bicarbonate may be beneficial
    - Suggested rate: isotonic saline or sodium bicarbonate (1 mL/kg/hr, starting 12 hours before study until 12 hours afterward)
    - No evidence-based benefit to oral hydration with water
  - N-acetylcysteine, orally or IV
    - Theoretically can prevent CIN by ↓ vasoconstriction and ↓ production of oxygen free radicals
    - Conflicting data regarding its effectiveness
    - Due to lack of convincing data, use of IV acetylcysteine is not generally recommended
  - Discontinuing nephrotoxic drugs
  - **No** benefit in prophylactic hemodialysis
- Performing hemodialysis immediately after CM administration does not prevent CIN
- Treatment
  - Supportive care, dopamine agonists, loop diuretics, mannitol
  - Dialysis: In minority of patients

DIAGNOSTIC CHECKLIST

Consider

- American College of Radiology recommends to obtain baseline creatinine in all patients with suspected renal dysfunction or those at risk
  - Patient is considered at risk if
    - History of kidney disease as adult
    - Family history of kidney failure
    - Treatment for diabetes mellitus
    - Paraproteinemia syndromes
    - Collagen vascular disease
    - Certain medications, such as NSAID, metformin, aminoglycosides, etc.

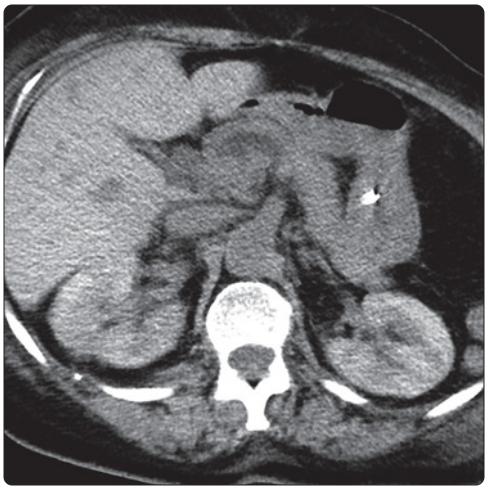
Guidelines for Management of Patients Taking Metformin

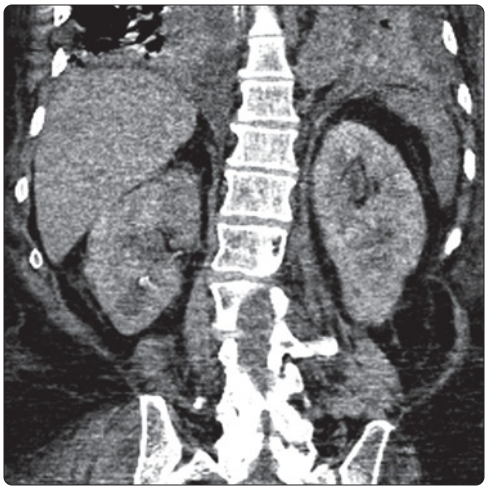
- Metformin-associated lactic acidosis is rare condition with reported morality as high as 50%
- Metformin is primarily cleared through glomerular filtration and tubular excretion; renal dysfunction, among other causes, results in increased half-life of metformin, which is risk factor for lactic acidosis
- CM is not independent risk factor for lactic acidosis and risk of clinically significant CIN is weak in patients with normal renal function
- In patients with underlying renal dysfunction or multiple comorbidities, CM may result in CIN and increased risk of lactic acidosis
- ACR recommendations

SELECTED REFERENCES

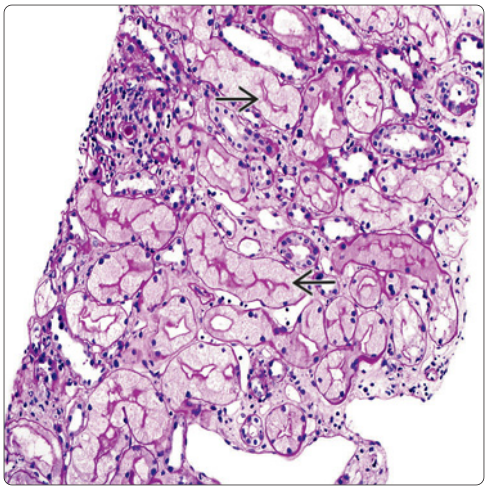
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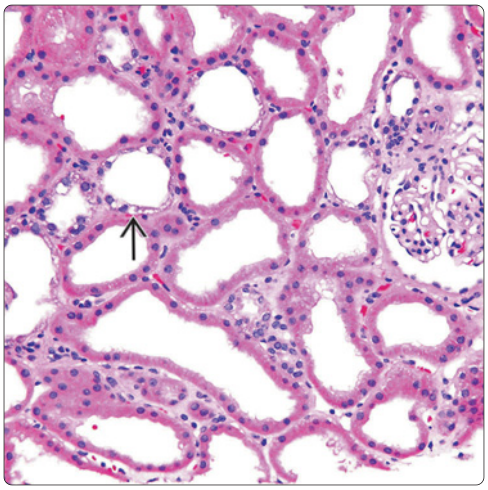
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(Left) *Axial unenhanced CT in a 69-year-old woman with history of septic shock shows delayed patchy enhancement of kidneys.* **(Right)** *Coronal image again depicts delayed patchy enhancement of kidneys. Patient had received iodinated contrast 7 days prior to this test. She had multiple comorbidities and her serum creatinine climbed from 1.5 mg/dL to 3.2 mg/dL due to ATN, which perhaps was multifactorial.*





(Left) *Photomicrograph in patient with osmotic tubulopathy associated with use of iodinated contrast shows tubular profiles that have cytoplasmic swelling, vacuolization, and preservation of the brush borders ⇨. (From DP2: Kidney.)* **(Right)** *Photomicrograph, H&E staining, in patient with ATN shows epithelial attenuation, loss of apical cytoplasm, prominent dilated tubules, and focal, largely distal, tubular epithelial vacuolization ⇨. (From DP: Kidney.)*

1st Generation: Ionic High-Osmolar Contrast Media		
Name	Iodine content	Osmolality
Diatrizoate *(Hypaque 50)*	300 mg/mL	1550 mOsm/kg
Metrizoate *(Isopaque 370)*	370 mg/mL	2100 mOsm/kg
2nd Generation: Nonionic Low-Osmolar Contrast Media		
Name	Iodine content	Osmolality
Iopamidol *(Isovue 370)*	370 mg/mL	800 mOsm/kg
Iohexol *(Omnipaque 350)*	350 mg/mL	880 mOsm/kg
Iopromide *(Ultravist 370)*	370 mg/mL	770 mOsm/kg
Ioxilan *(Oxilan 350)*	350 mg/mL	700 mOsm/kg
3rd Generation: Nonionic Iso-Osmolar Contrast Media		
Name	Iodine content	Osmolality
Iodixanol *(Visipaque 320)*	320 mg/mL	290 mOsm/kg

Category I	
Patients with normal GFR and no known comorbidities*	No need to discontinue metformin
Category II	
Patients with normal GFR who have multiple comorbidities	Discontinue metformin and withheld for 48 hours Depending on comorbidities, GFR might need to be rechecked before restarting metformin
Category III	
Patients with known renal dysfunction	Metformin should be discontinued at time of examination and creatinine level should be followed up closely until metformin can be safely restarted

*Comorbidities: Liver dysfunction, alcohol abuse, cardiac failure, muscle ischemia, rhabdomyolysis, sepsis

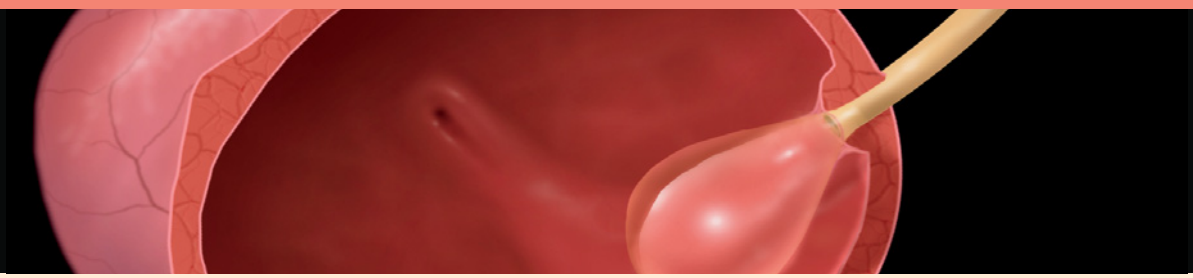
(Left) *Table summarizes different generations of iodinated contrast media with their corresponding iodine content and osmolarity.* **(Right)** *Table summarizes suggested ACR guidelines for discontinuation of metformin after administration of iodinated contrast media.*

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SECTION 5

Ureter



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Congenital

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Inflammation

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Anatomy

The ureter is a muscular tube extending from the ureteropelvic junction (UPJ) to the bladder for a length of 25-30 cm. The proximal ureter is in the perirenal space, passing posterior to the gonadal vessels and traveling along the psoas muscle to cross over the iliac vessels at the pelvic brim.

The ureters continue inferiorly along the pelvic wall to approximately the level of the ischial spines where they curve anteromedially to enter the bladder. This occurs at the level of the seminal vesicles in men and cervix in women. This anatomic proximity has important implications in women with invasive cervical cancer in whom ureteral obstruction is a common and important complication.

At the ureterovesical junction (UVJ), the ureters pass obliquely through the bladder wall, creating a valve effect and therefore preventing reflux. The UVJ, along with the UPJ and pelvic brim, are all physiologic points of narrowing and are often the sites where ureteral stones may be lodged.

Imaging Techniques and Indications

Radiography

With the advent of multidetector CT and advancing MR techniques, excretory urography (also referred to as intravenous pyelography or IVP) has largely been replaced. Modern CT and MR provide a comprehensive assessment of both the renal parenchyma and now, with excretory phase protocols, the urinary tract. Although retrograde urography is still performed, it is usually undertaken in conjunction with a procedure, such as stone retrieval or biopsy of a mass, and less commonly as a primary diagnostic procedure.

CT Urography

There is no universally accepted technique for performing CT urography. An unenhanced phase is necessary for detection of calculi, a nephrographic phase is best for identifying renal masses, and an excretory phase best detects urothelial lesions.

The goal in evaluating the ureter is to view it distended and opacified throughout its entire course, but as in the days of the conventional excretory urogram, peristalsis may result in unopacified ureteral segments. Because the ureters taper as they enter the bladder, the distal portions are particularly difficult to completely opacify.

While many protocols exist, it is clear that the ureter is best evaluated when the patient is well hydrated. Hydration can be achieved orally (750-1,000 mL of water ~ 30 minutes prior to the exam) or by a combination of oral and intravenous approaches (for example 500 mL water ~ 30 minutes prior to the exam and then 250 mL of intravenous 0.9% saline just before scanning the patient). Several investigators have advocated intravenous administration of 10 mg of furosemide 2-3 minutes prior to the procedure to further increase the flow rate of opacified urine.

Excretory phase images are obtained 10-15 minutes after the initial contrast administration with thin beam collimation and low pitch scanning. Since radiation dose is a paramount consideration, particularly in young patients, various protocols including split-bolus and triple-dose techniques have been designed to reduce the number of scans. Inconsistent results, however, with the split-bolus technique have prompted many centers to employ 3-phase protocols for all referred patients or to selectively employ split-bolusing in low-risk, younger patients. Novel reconstruction algorithms (i.e., statistical and model-based iterative algorithms) allow for significant dose reduction without affecting the diagnostic quality of 3-phase exams.

The ureters should be reviewed routinely in both the axial and coronal planes. Further postprocessing reconstructions, including maximum intensity projections (MIP), curved planar reformations, and 3D volume rendering, give the "big picture," simulating the old excretory urogram. The result is an excellent evaluation of ureters, which is equivalent to retrograde urography for the detection of urothelial tumors.

MR Urography

MR is more technically challenging than CT and is primarily used to limit radiation dose in children, younger patients, and pregnant women. Two basics techniques can be used to view the ureter: Static-fluid MR urography and contrast-enhanced excretory MR urography.

Static-fluid MR urography uses heavily T2-weighted images with suppression of background tissues, similar to that done in MR cholangiography. This technique is useful when the collecting system is dilated, but it may be difficult to completely visualize normal ureters. Multiple thick-slab, single-shot, fast spin-echo images can be obtained and viewed in a cine loop to improve visualization of nondilated ureters and avoid mistaking ureteral peristalsis for a stricture of filling defect.

Excretory MR urography is performed after the administration of gadolinium. This technique is technically challenging. Gadolinium initially appears bright on T1-weighted images. As it becomes concentrated, however, the signal intensity of urine decreases due to T2* effects. Administration of a diuretic greatly improves the quality of MR urography by both distending the collecting system and diluting the gadolinium concentration.

Approach to the Abnormal Ureter

Structural abnormalities of the ureter can be divided into 3 simplified categories: **Dilatation**, **stricture**, and **filling defect (masses)**. Clearly there is overlap in these categories.

Ureteral dilatation can be structural (due to downstream obstruction) or functional. **Obstructive etiologies** include ureteral calculi (the most common etiology, particularly in the acute setting), obstructing masses, downstream strictures, extrinsic compression, and ureteroceles. **Functional etiologies** include reflux, primary megaureter, and ureterectasis of pregnancy.

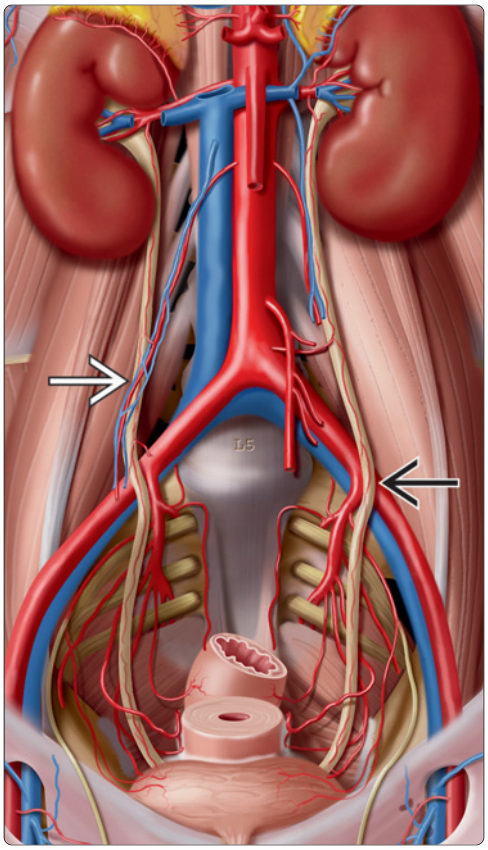
Ureteral strictures may be caused by neoplastic, infectious, or inflammatory processes. Neoplastic strictures include urothelial tumors or encasement by an extrinsic tumor, such as colon or cervical cancer. Infections (TB, schistosomiasis) or inflammatory/infiltrative processes (Crohn disease, endometriosis) also may involve and constrict the ureter. Many of these entities also cause ureteral wall thickening. When wall thickening is profound, always consider lymphoma.

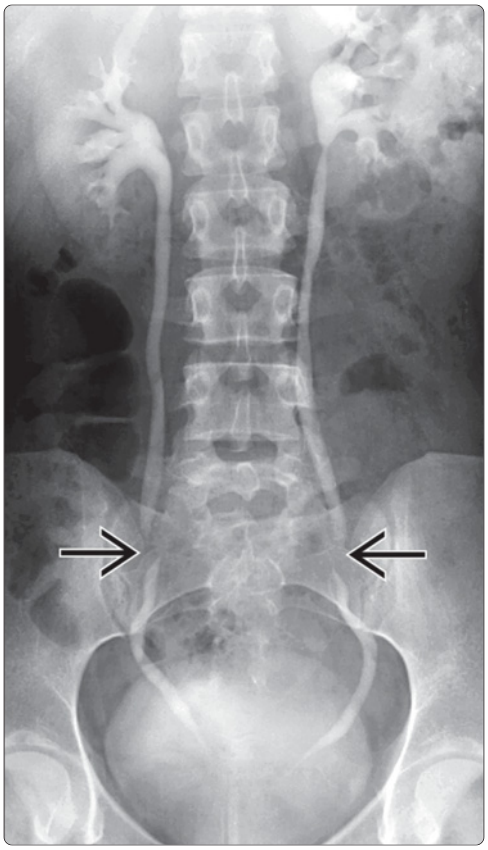
The most common **ureteral filling defects** are calculi and urothelial tumors. Other differential considerations include blood clot, mycetoma, fibroepithelial polyp, papilloma, ureteritis cystica, and malakoplakia.

Selected References

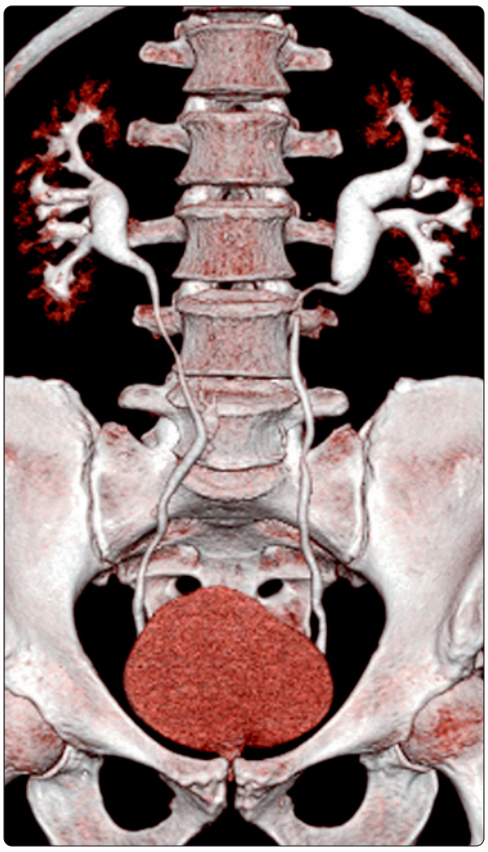
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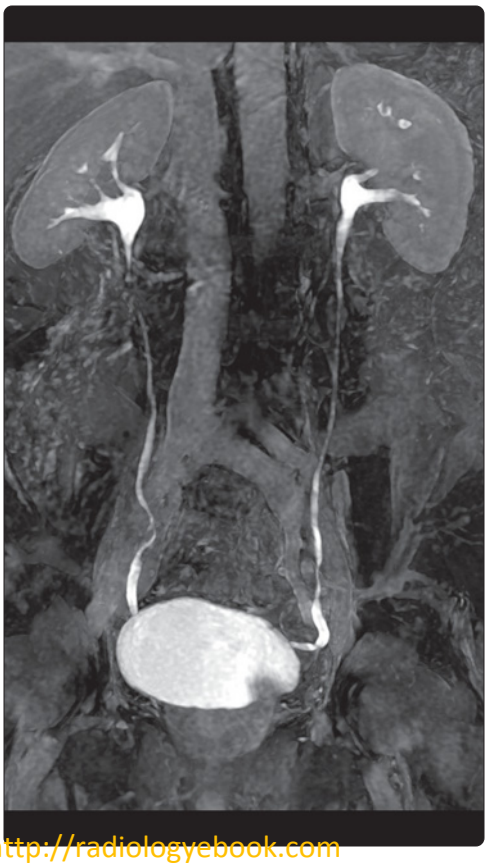
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(Left) *Graphic illustrates the normal course of the ureters, which extend from the renal pelvis to the urinary bladder. The ureters travel along the psoas muscle, passing posterior to the gonadal vessels ➡. Near the pelvic brim, the ureter ⇨ passes over the iliac vessels, which can cause extrinsic impression on the ureter on imaging studies.* **(Right)** *Excretory urogram shows mild extrinsic compression and medial deviation of the ureters ⇨ where they pass over the iliac vessels. Excretory urography has essentially been replaced by CT and MR urography, which provide a comprehensive assessment of the kidney, urinary tract, and surrounding structures.*





(Left) *In this 3D surface-rendered reconstruction of the urinary tract, the windowing has been optimized to display the renal collecting system. The color scale is arbitrary, and in this case, opacified urine is displayed as white. Less dense urine, within the renal tubules of the pyramids and the diluted urine within the bladder, is displayed in shades of red.* **(Right)** *Coronal MIP post-contrast T1W MR (MR urography) in a 48-year-old man with a history of microscopic hematuria shows opacification of renal collecting systems, ureters, and bladder. This method is especially useful in children (for evaluation of congenital anomalies).*

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Duplicated and Ectopic Ureter

KEY FACTS

TERMINOLOGY

- **Ectopic ureter**: Ureteral orifice below normal insertion in trigone; commonly associated with duplex collecting system
- **Duplicated ureter**: 2 ureters drain duplex kidney and remain separate down to bladder insertion or beyond

IMAGING

- Location
  - Males: Insertion of ureter is always above external sphincter
    - Insertion sites: Prostatic urethra (54%), seminal vesicles (28%), vas deferens (10%), ejaculatory duct (8%)
  - Females: Insertion is usually below sphincter resulting in urinary incontinence
    - Insertion sites: Bladder neck and upper urethra (33%), vestibule (33%), vagina (25%), cervix and uterus (5%)
- 75-90% of ectopic ureters occur in setting of duplication (in Western world)
- 85% of duplicated ureters obey Weigert-Meyer rule
- **Weigert-Meyer rule**: Upper pole ureter inserts medial and caudal to lower pole ureter

TOP DIFFERENTIAL DIAGNOSES

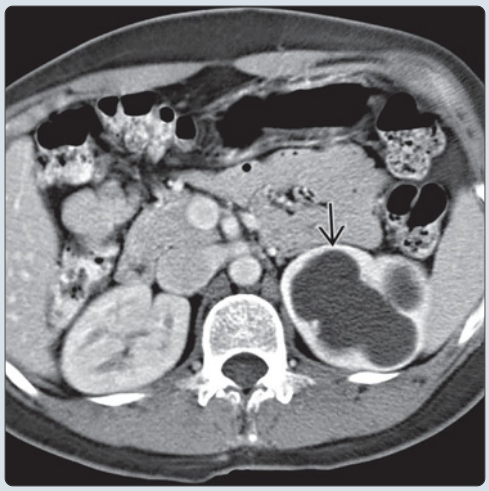
- Bifid ureters
- Ureterocele
- Seminal vesicle cyst
- Gartner duct cysts

CLINICAL ISSUES

- Often asymptomatic in males
- Incontinence is only seen in females due to insertion of upper pole ureteral orifice below bladder sphincter
- Management depends on presence of duplication and extent of function of involved kidney (moiety)

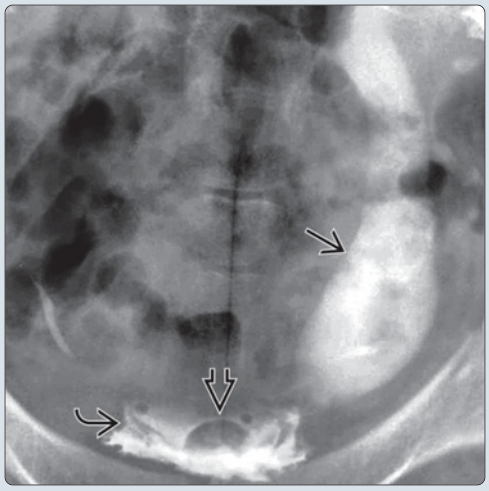
(Left) *Graphic illustrates a duplex left kidney and the Weigert-Meyer principle. The ureter from the upper pole inserts ectopically, medial and caudal to the lower pole ureter, ending in a ureterocele in this case.* **(Right)** *Axial CECT in a 19-year-old woman with chronic left flank pain shows a dilated collecting system in the upper pole, while the lower pole of the kidney was normal. Two ureters could be followed from the left kidney down to the bladder.*





(Left) *Plain film following CECT in the same patient shows dilation of the pelvocalyceal system in the upper pole. Although displaced inferiorly, the lower pole calyces are normal.* **(Right)** *Excretory urography in the same patient shows dilated upper-pole ureter just above its ectopic insertion near the base of the bladder, where it terminates in a ureterocele, seen as a filling defect in the bladder. Note the ureter from the right kidney and its normal insertion in the trigone.*





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TERMINOLOGY

Synonyms

- Double ureters
- Ureteral ectopia

Definitions

- **Ectopic ureter**: Ureteral orifice below normal insertion in trigone; commonly associated with duplex collecting system
- **Duplicated ureter**: 2 ureters drain duplex kidney and remain separate down to bladder insertion or beyond

IMAGING

General Features

- Best diagnostic clue
  - Double (duplex) collecting system with complete separation of ureters down to their insertion
- Location
  - Ectopic ureter inserts along developing mesonephric duct
    - Mesonephric (wolffian) duct: Embryogenic precursor of trigone, epididymis, vas deferens, ejaculatory duct, and seminal vesicles
  - Most ectopic insertions are outside bladder
  - Ectopic insertion within bladder
    - Usually no clinical sequela
  - Ectopic insertion outside bladder
    - Males: Insertion of ureter is always above external sphincter
      - Boys with ureteral duplication and ectopia remain continent of urine
      - Insertion sites: Prostatic urethra (54%), seminal vesicles (28%), vas deferens (10%), ejaculatory duct (8%)
    - Females: Insertion is usually below sphincter resulting in urinary incontinence
      - Insertion sites: Bladder neck and upper urethra (33%), vestibule (33%), vagina (25%), cervix and uterus (5%)
  - 75-90% of ectopic ureters occur in setting of duplication (in Western world)
  - 85% of duplicated ureters obey Weigert-Meyer rule
    - **Weigert-Meyer rule**: Upper pole ureter inserts medial and caudal to lower pole ureter
    - Most commonly, upper pole ureter is ectopic and obstructed (due to ureterocele), and lower pole ureter refluxes
    - In other 15%, upper pole ureter inserts anywhere along mesonephric duct pathway (see above)
  - ~ 10% chance of ureteral ectopia in contralateral ureter
  - ~ 20% chance of ureteral duplication in contralateral side

Radiographic Findings

- IVP or CT urography
  - Duplicated kidney with ectopic ureter insertion
    - Duplex kidney with double ureters
    - Dilated tortuous ureter to level of insertion
    - Poor or no excretion by upper pole of duplex kidney
    - **Drooping lily sign**
      - Hydronephrosis and ↓ function of obstructed upper pole → downward displacement of lower pole calyces
    - **Nubbin sign**
      - Scarring, atrophy, and ↓ function of lower pole moiety
      - May simulate renal mass
    - Fewer calyces and infundibula of lower pole collecting system; shortened upper pole infundibulum
    - Single or diffuse calyceal clubbing, thin overlying parenchyma ± scarring in lower pole
    - ± ureteropelvic junction obstruction of lower pole
    - Ectopic ureter is commonly associated with ureterocele
  - Nonduplicated kidney with ectopic ureter insertion
    - Involved kidney is usually small, dysplastic, nonfunctional
    - Males with ectopic single system often have dilated ipsilateral seminal vesicle (on CT, US, MR)
- Voiding cystourethrogram
  - ± reflux, ureterocele, diverticulum of urethra
  - Useful to locate ectopic insertion if it is within urinary tract

CT Findings

- "Faceless" kidney: No renal sinus or collecting system at junction of upper and lower pole of duplex kidney
- ± obstruction in either pole of duplex kidney
- Abnormal course and insertion of ectopic ureter, and associated ureterocele, are better depicted on delayed-phase CT urography

Ultrasonographic Findings

- Duplex kidney: 2 central echo complexes with intervening renal parenchyma
- Upper pole hydronephrosis
  - May appear as cystic mass in medial upper pole of kidney
- Dilated ureter behind bladder

MR Findings

- MR urography (MRU) is based on fat-saturated T2-weighted and multiphasic post-contrast T1-weighted 3D GRE sequences
  - Furosemide (Lasix) and IV hydration is used for optimal distention of collecting system
  - Bladder catheterization is used in selected patients
- MRU is especially useful in younger population to avoid ionizing radiation
- Morphology and course of renal collecting systems can be nicely depicted on MRU

Nuclear Medicine Findings

- Nuclear cystogram can substitute VCUG for evaluation of reflux
- Renal scintigraphy (with DMSA or MAG-3) is helpful to estimate degree of renal function
  - Variable function of upper pole moiety

Imaging Recommendations

- Best imaging tool
  - US for screening
    - Can identify duplicated obstructed system

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- CT
  - Can locate small, poorly functioning, dysplastic kidney drained by single ectopic ureter
  - CT urography can demonstrate entire spectrum of anomalies
- MR
  - Can also show complete extent of anomalies without radiation
    - Especially useful in children

DIFFERENTIAL DIAGNOSIS

Bifid Ureters

- 2 ureters drain duplex kidney but join before emptying into bladder; can be ectopic
- Ureters usually join above bladder as "V" or "Y" junction
- IVP
  - Ureteroureteral reflux or "yo-yo" phenomenon: Peristalsis down 1 limb of ureter forces urine via reflux up other limb
  - Asymmetric dilatation of 1 ureteral segment

Ureterocele

- Can be seen as an isolated finding without association with ectopic or duplicated ureter

Seminal Vesicle Cyst

- Associated with ipsilateral renal agenesis
- May be mistaken for ectopic insertion of ureter

Gartner Duct Cysts

- Inclusion cyst in lateral wall of vagina
- Remnant of mesonephric (wolffian) duct
- Might be mistaken for ectopic insertion of ureter

PATHOLOGY

General Features

- Etiology
  - Embryology
    - Ureters form from ureteral buds, which arise as diverticula from distal mesonephric (wolffian) duct
    - Ureteral bud grows into metanephric blastemal ridge, which is progenitor of kidney
    - Distal portion of mesonephric duct (between ureteral bud and cloaca) develops into bladder trigone
  - Pathogenesis
    - Abnormal location of ureteral bud results in ureteral ectopia
    - Duplex kidney is caused by duplication of ureteral bud
  - Genetics: Autosomal dominant with low penetrance
- Associated abnormalities
  - Solitary or dysplastic kidney, hypoplastic kidneys, all types of fused kidneys or posterior urethral valves
  - Complex congenital anomalies: VATER, VACTERL (**v**ertebral, **a**nal, **c**ardiovascular, **t**racheo**e**sophageal, **r**enal, and **l**imb)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Often asymptomatic in males
    - May have recurrent epididymitis, urinary tract infection, or flank pain
  - Incontinence is only seen in females due to insertion of upper pole ureteral orifice below bladder sphincter
  - Intermittent or persistent urinary tract infections ± acute pyelonephritis, frequently in females

Demographics

- Gender
  - Female predominance (M:F = 1:10)
    - May be artifactual since males often remain asymptomatic
- Epidemiology
  - In Western world, 75-90% of ectopic ureters occur in setting of duplex collecting system
  - True incidence is unknown, because many patients are asymptomatic
  - Prevalence of duplex renal collecting system in autopsy series is 0.5-1.2%

Natural History & Prognosis

- Complications: Obstruction, urolithiasis, abscess, renal failure

Treatment

- Management depends on presence of duplication and extent of function of involved kidney (moiety)
- Partial nephrectomy and ureterectomy: In patients with ureteral ectopia in setting of duplex system with dysplastic/nonfunctioning upper pole moiety
- Nephroureterectomy: In patients with single system and poorly functioning kidney
- Ureteral reimplantation (to bladder or to lower pole ureter): In patients with normal-functioning kidney (moiety)

DIAGNOSTIC CHECKLIST

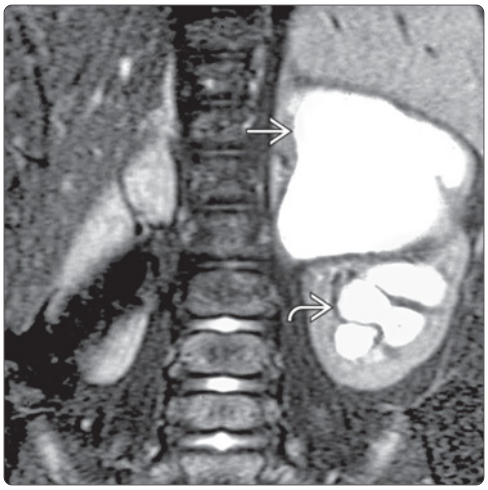
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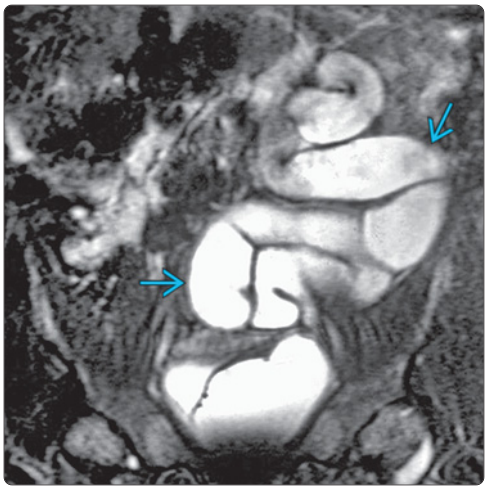
- Young girl with incontinence or recurrent urinary tract infections
- Boy with recurrent epididymitis or urinary tract infections

SELECTED REFERENCES

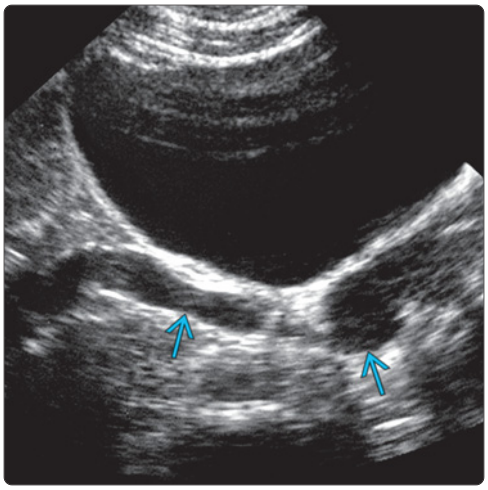
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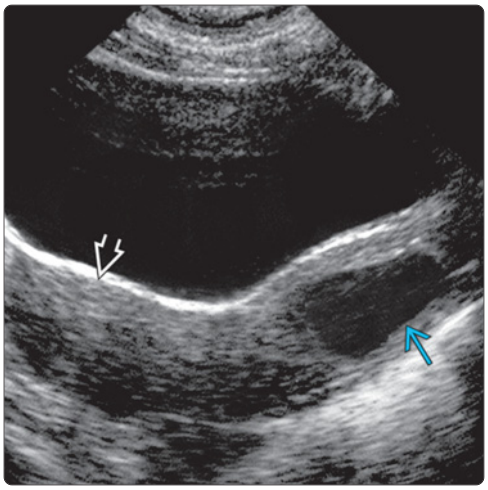
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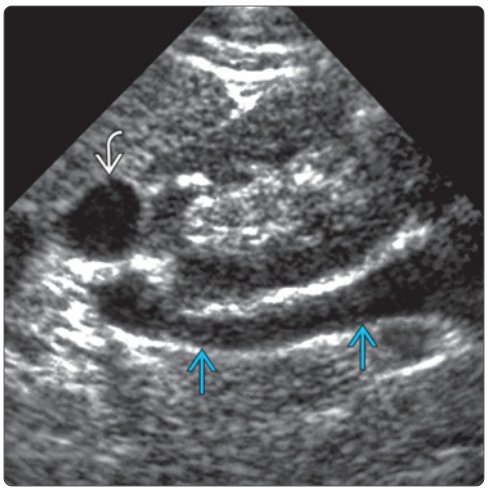


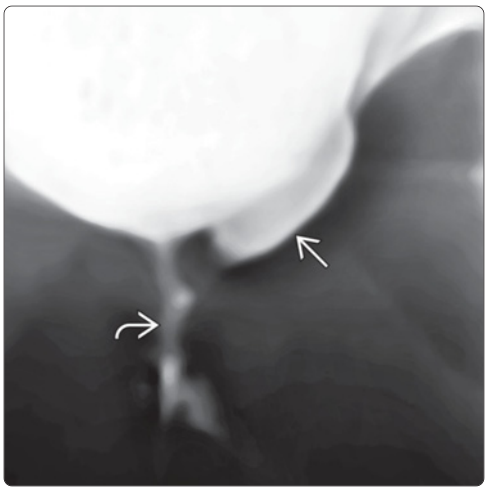
(Left) *Coronal T2W MR shows a duplicated left collecting system with a markedly hydronephrotic upper pole and a moderately hydronephrotic lower pole.* **(Right)** *Coronal T2W MR, in the same patient shows the markedly dilated and tortuous ectopic upper-pole moiety ureter.*





(Left) *Longitudinal ultrasound through the bladder shows a tortuous dilated ureter continuing beyond the expected insertion site at the bladder trigone.* **(Right)** *Longitudinal ultrasound in the same patient shows a cystic mass within the wall of the vagina, representing an ectopic insertion of the dilated, obstructed ureter, which was draining the upper pole of a duplicated system. The uterus is also seen and has a normal appearance.*





(Left) *Longitudinal US through the left kidney shows a dilated ectopic ureter medial and deep to the kidney. Note the cystic dysplasia of the upper pole moiety.* **(Right)** *Voiding cystourethrogram shows reflux of urine into an ectopic left ureter that inserts into the urethra.*

Ureterocele

KEY FACTS

TERMINOLOGY

- Cystic dilatation of terminal ureter
- Several classification schemes exist, more widely accepted scheme divides ureteroceles into intravesical and ectopic
- **Intravesical**: Ureterocele is entirely in bladder
- **Ectopic**: Insertion at bladder neck or in posterior urethra
- ~ 80% of ureteroceles (in pediatric population) are seen in association with duplicated collecting system
- ~ 60% of these pediatric ureteroceles have ectopic insertion

IMAGING

- "Cobra head" or "spring onion" deformity of distal ureter with surrounding radiolucent halo
- Smooth round intravesical filling defect

TOP DIFFERENTIAL DIAGNOSES

- Pseudoureterocele
  - Acquired dilation of terminal ureter due to obstructing mass or calculi
  - Results in "cobra head" appearance on IVP or VCUG; halo around "cobra head" is thicker and indistinct
- Filling defect in bladder at trigone
- Bladder or cervical cancer
- Cystitis
- Distal ureterovesical junction stone or edema

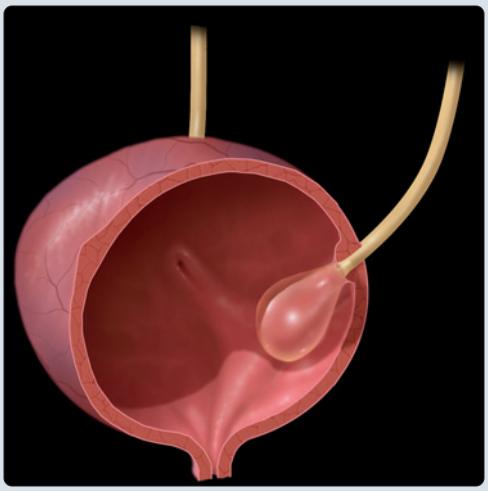
PATHOLOGY

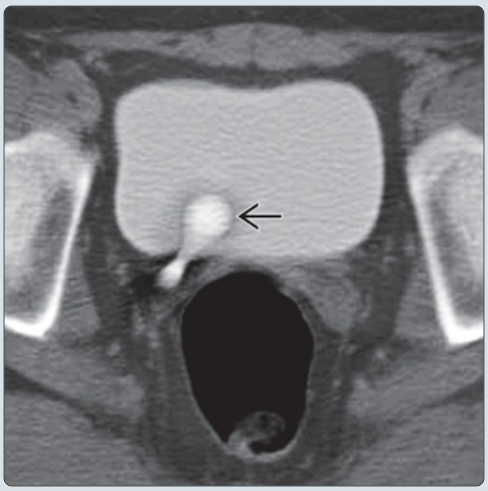
- Most accepted theory proposes stenosis of ureteral orifice as pathogenesis

CLINICAL ISSUES

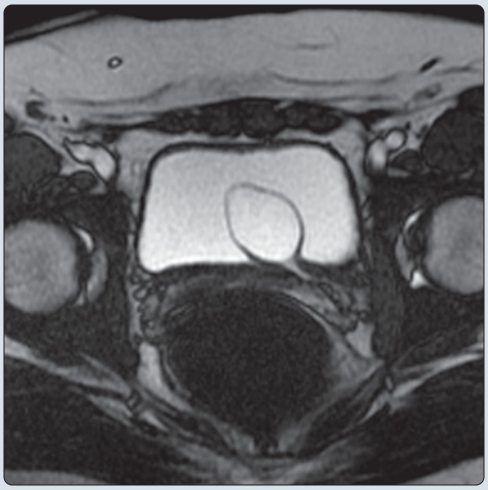
- Most symptomatic cases needed surgery
  - Endoscopic decompression: Most effective for intravesical type
  - Open surgery with ureteral reimplantation

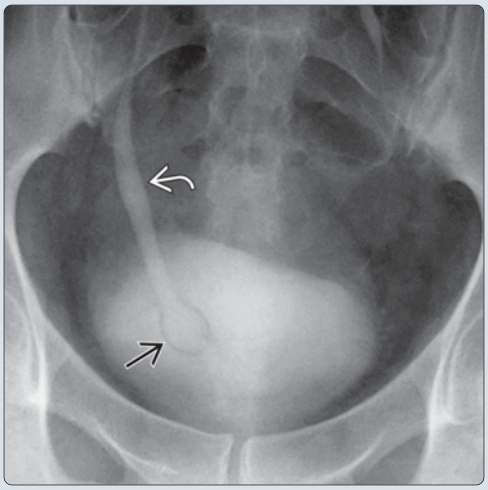
(Left) *Graphic shows an intravesical ureterocele. Note the prolapsing, dilated end of an orthotopic ureter, which inserts normally at the trigone.* **(Right)** *Axial delayed-phase CECT shows the typical appearance of an intravesical ureterocele. The distal ureter is dilated and prolapses into the bladder lumen. The insertion point of the ureter is normal, and there was no significant hydronephrosis in this patient.*





(Left) *Axial steady-state free precession (FIESTA) image through the bladder shows a classic "cobra head" appearance of an intravesical ureterocele. The ureterocele wall is thin and well delineated.* **(Right)** *Frontal projection from an excretory-phase IVP shows a classic spring onion appearance of an intravesical ureterocele with mild hydroureter.*





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TERMINOLOGY

Definitions

- Cystic dilatation of terminal ureter
- Several classification schemes exist, the most commonly used (introduced by the American Academy of Pediatrics [AAP]) divides ureteroceles into intravesical and ectopic
  - **Intravesical**: The ureterocele is entirely in bladder
  - **Ectopic**: Insertion at bladder neck or in posterior urethra
    - More common with duplicated systems
  - Older classification (introduced in 1954) divided ureteroceles into simple (orthotopic) and ectopic; however, this classification has been largely abandoned due to confusion with the word "simple"
    - "Simple" ureterocele can be seen with both simple and duplex collecting systems
- ~ 80% of ureteroceles (in pediatric population) are seen in association with duplicated collecting system
  - ~ 60% of these have ectopic insertion

IMAGING

General Features

- Best diagnostic clue
  - "Cobra head" or "spring onion" deformity of distal ureter with surrounding radiolucent halo
  - Smooth round intravesical filling defect
- Location
  - Intravesical: Entire ureterocele is in bladder
  - Ectopic: Insertion in bladder neck or posterior urethra
    - In males, insertion is always above external sphincter therefore continent
  - Similar frequency on right and left
  - ~ 10% of cases are bilateral
- Size
  - Up to several cm in diameter

Radiographic Findings

- IVP
  - "Cobra head" or "spring onion" deformity of terminal ureter
    - Dilated distal ureter projects into lumen of bladder with surrounding radiolucent halo
      - Wall of ureterocele is thin
    - This configuration is not pathognomonic for ureterocele and can be seen with other obstructing lesions
      - "Pseudoureterocele": Dilation of terminal ureter due to obstructing mass, calculus, or edema at orifice (in setting of recently passed stone)
      - Halo of "pseudoureterocele" is thicker and indistinct
  - Smooth, radiolucent intravesicular mass
- Voiding cystourethrography (VCUG)
  - Smooth, radiolucent intravesicular mass
  - Ureteroceles may temporarily evert during voiding and mimic diverticula ("everted ureterocele")

CT Findings

- CECT
  - Intravesicular mass at ureterovesical junction (UVJ) or in bladder neck/proximal urethra
  - "Mass" opacifies with contrast on delayed-phase images
  - Occasionally, calculi are seen within diverticulum, presumably due to stenotic orifice and urinary stasis
  - Look for duplex collecting system and its complications

MR Findings

- MR Urography (MRU)
  - Heavily T2-weighted sequences and delayed post-contrast urographic-phase sequences offer high spatial and contrast resolution for evaluation of urinary tract
  - Intravesicular "mass" at UVJ or in bladder neck/proximal urethra
  - "Mass" is markedly T2-hyperintense and fills with contrast on delayed urographic phase
  - Look for duplex collecting system and its complications

Ultrasonographic Findings

- Grayscale ultrasound
  - Thin-walled, cystic intravesical mass near insertion of ureter

Imaging Recommendations

- Best imaging tool
  - CT or MR urography
- Protocol advice
  - Need delayed-phase imaging to see opacification of lumen of ureterocele

DIFFERENTIAL DIAGNOSIS

Pseudoureterocele

- Acquired dilation of terminal ureter due to obstructing mass or calculi
- Results in cobra head appearance on IVP or VCUG; halo around "cobra head" is thicker and indistinct

Filling Defect in Bladder at Trigone

- Bladder or cervical cancer
- Cystitis
- Distal UVJ stone or edema
  - Secondary to recent stone passage
- Intravesical stone or clot

PATHOLOGY

General Features

- Etiology
  - Congenital anomaly
  - Exact underlying pathogenesis is unknown
  - Most accepted theory proposes stenosis of ureteral orifice as pathogenesis

Staging, Grading, & Classification

- Initial classification (1954) divided ureteroceles into simple and ectopic
  - The term "simple" caused confusion since "simple" ureteroceles can be seen with both single system and duplex systems
- Most widely used classification (introduced by the AAP) divides ureteroceles into intravesical and ectopic
- Several other classifications based on size of orifice and presence of associated reflux exist
- Classification based on size and location of orifice

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- Stenotic
- Sphincteric
- Sphincterostenotic
- Cecoureterocele

Gross Pathologic & Surgical Features

- Intravesical ureteroceles: Pinpoint orifices, but no significant obstruction
- Ectopic ureteroceles: Often obstructed, with dysplasia of upper pole kidney

Microscopic Features

- Wall: Covered by bladder mucosa and lined by ureteral mucosa

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Intravesical: Usually asymptomatic, incidental finding in adults
  - Ectopic ureteroceles: Urinary tract infection (UTI), incontinence
- Clinical profile
  - Ectopic: Infant or child with UTI or sepsis

Demographics

- Age
  - Ectopic
    - Median age: 3 months at diagnosis
      - Often diagnosed with prenatal ultrasound
- Gender
  - 4-6 times more common in female
- Ethnicity
  - More common in Caucasians
- Epidemiology
  - Clinical incidence is 1:12,000-1:5,000
  - Its prevalence on autopsy has been reported as high as 1:500 in some series

Natural History & Prognosis

- Obstructed ureteroceles may cause stasis and stone formation
- Severe obstruction: Primarily with ectopic ureteroceles

Treatment

- Options, risks, complications
  - Management depends on type of ureterocele, presence or absence of duplex collecting system, function of upper pole moiety (in case of duplex system), and presence or absence of reflux
  - Most symptomatic cases needed surgery
    - Endoscopic decompression: Most effective in intravesical type
    - Open surgery with ureteral reimplantation
    - Partial nephrectomy (for ectopic ureterocele associated with nonfunctioning upper pole moiety)
  - Prophylactic antibiotics if associated vesicoureteral reflux (VUR) exists

DIAGNOSTIC CHECKLIST

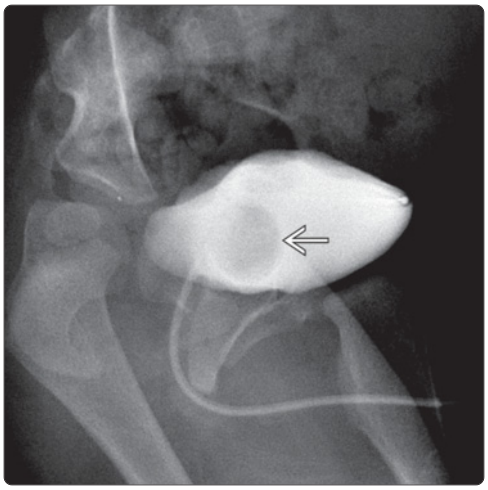
Consider

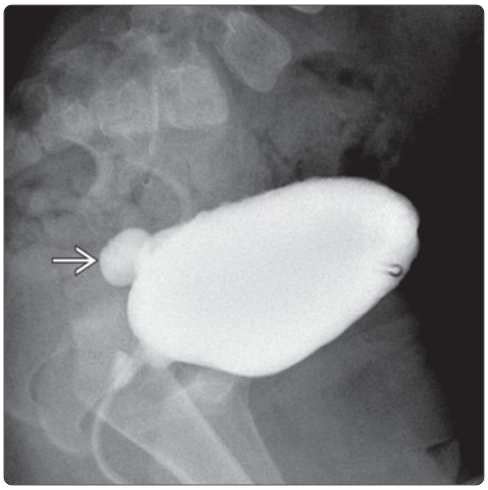
- New classification scheme dividing ureteroceles into intravesical and ectopic subtypes is utilized by urologists and pediatricians
- Look for duplex collecting system and its complications
- Check for mucosal irregularity to rule out tumor

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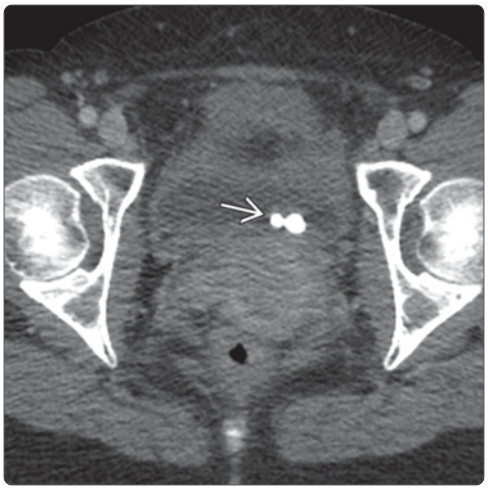
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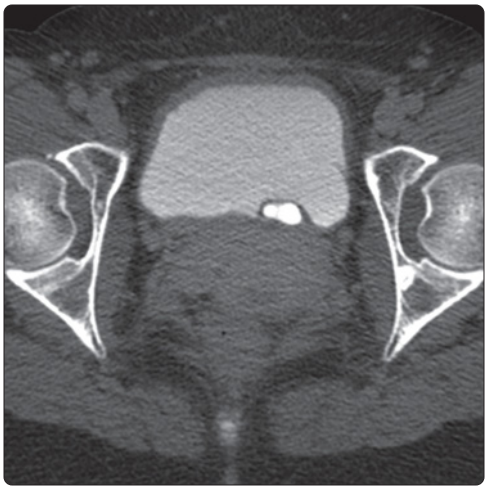
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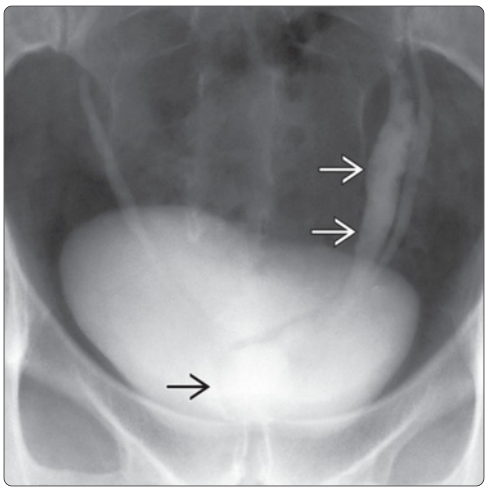


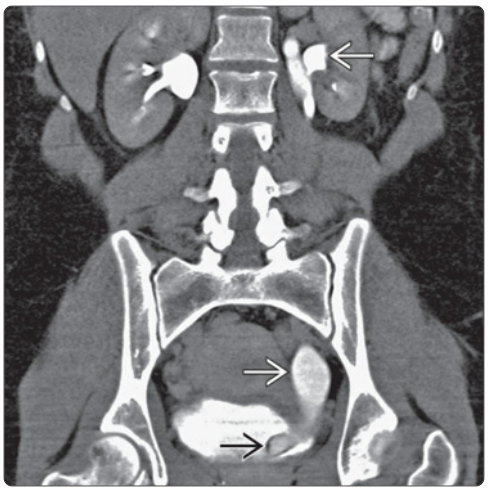
(Left) *Anterior oblique projection from VCUG in a male toddler with a history of urinary tract infection shows a smooth round filling defect within the bladder ➡ compatible with a ureterocele.* **(Right)** *Upon further distention of bladder, the ureterocele everted to outside of bladder lumen ➡. Everted ureteroceles can be mistaken for bladder diverticula.*





(Left) *Axial noncontrast CT through the bladder shows 2 densely calcified stones ➡ in region of the left UVJ.* **(Right)** *Axial image through same section, obtained during a delayed urographic phase, shows that the calculi are confined within a ureterocele. At times, it might be difficult to differentiate a congenital ureterocele containing calculi from acquired dilatation of terminal ureter in the setting of an obstructing calculus (so-called "pseudoureterocele").*





(Left) *Intravenous pyelogram shows complete duplication of left ureters with an ectopic ureterocele at the bladder neck ⇨. Filling defects in the dilated upper moiety ureter ➡ represent ureteritis cystica. (Courtesy M. Nino-Murcia, MD.)* **(Right)** *Coronal CECT shows an intravesical ureterocele ⇨ with associated dilation of the ureter and calyces ➡. Hydronephrosis is relatively uncommon and usually mild in patients with intravesical ureteroceles.*

Ureteritis Cystica

KEY FACTS

TERMINOLOGY

- Reactive proliferative changes of urothelium with formation of multiple small, subepithelial cysts

IMAGING

- More common in proximal 1/3 of ureter
- May also occur in renal pelvis (pyelitis cystica) and urinary bladder (cystitis cystica)
- Ureterogram (intravenous or retrograde/antegrade)
  - Nodular, mural-based, radiolucent small filling defects in ureteral lumen
  - Scalloping of ureteral margins (in profile view)

TOP DIFFERENTIAL DIAGNOSES

- Blood clots, radiolucent stones
- Air bubbles
- Multifocal transitional cell carcinoma or papillary tumors
- Metastatic melanoma
- Impression from crossing vessels
- Chronic infectious ureteritis

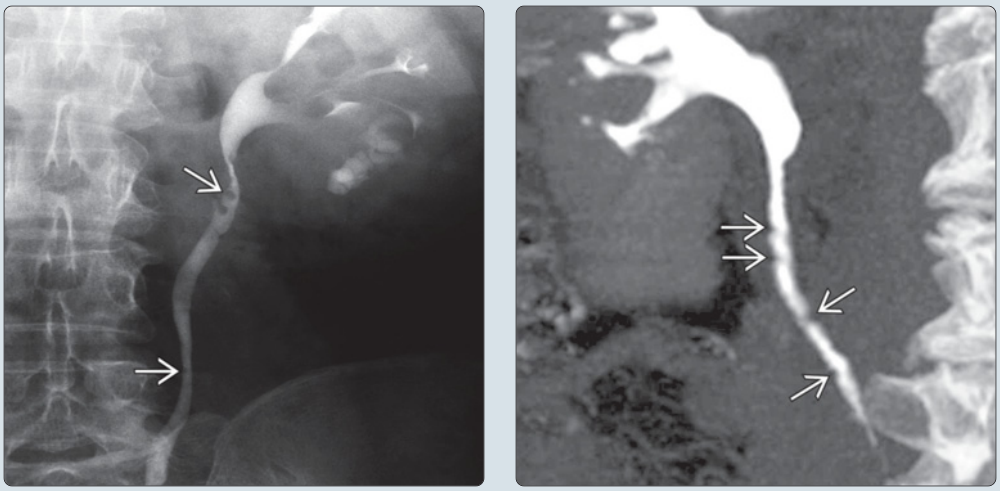
PATHOLOGY

- Likely the response to chronic inflammation or irritation
  - Seen in association with UTI, urolithiasis, chemotherapy, radiation, schistosomiasis, and instrumentation
- Theory of pathogenesis: **von Brunn cell nest**
  - Inflammatory stimulation → proliferation of urothelium → invagination of urothelial cells
  - Cells become isolated within lamina propria → metaplastic transformation into glandular structures (**ureteritis glandularis**) → cystic dilation of von Brunn nests (**ureteritis cystica**)

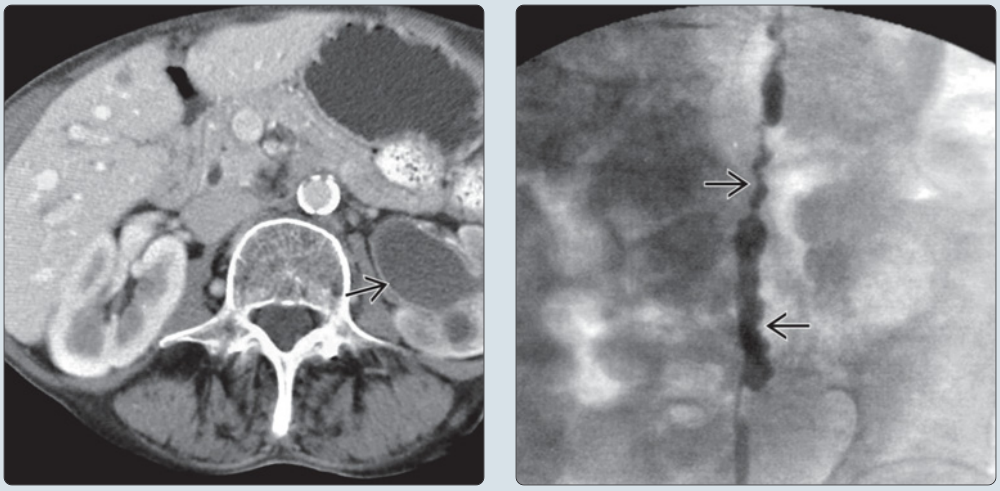
CLINICAL ISSUES

- Usually inconsequential; should rule out UTI
- No risk of malignant degeneration
  - Predisposing factors for ureteritis cystica (such as chronic infection) may act as risk factors for urothelial malignancy

(Left) *Frontal projection from excretory phase IVP shows several small, mural-based filling defects in left proximal and mid ureter, typical findings of ureteritis cystica.* **(Right)** *Coronal MIP reformat from CT urogram shows several smooth filling defects along the right ureter in this elderly man with dysuria and prior infections. Endoscopy confirmed the submucosal location of these lesions, and biopsy specimens were negative for transitional cell carcinoma.*



(Left) *Axial contrast-enhanced CT shows left-sided hydronephrosis and a delayed nephrogram in this elderly man with pyuria and fever. The walls of the left ureter and renal pelvis are thickened and enhance with contrast, indicating inflammation. Pyonephrosis was confirmed at nephrostomy.* **(Right)** *Antegrade ureterography in the same patient shows an irregular contour of the left ureter, compatible with ureteritis. Ureteral spasm may also play a part in this patient, as active infection is present.*



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TERMINOLOGY

Synonyms

- Pyeloureteritis cystica (if renal pelvis involved), proliferative ureteritis, ureteritis cystica et glandularis

Definitions

- Reactive proliferative changes of urothelium with formation of multiple small, subepithelial cysts

IMAGING

General Features

- Location
  - More common in proximal 1/3 of ureter
  - May also occur in renal pelvis (pyelitis cystica) and urinary bladder (cystitis cystica)
  - May be unilateral or bilateral, symmetric or asymmetric
    - ~ 50% are bilateral
- Size
  - Cysts are < 5 mm; typically 2-3 mm
    - Occasionally may be as large as 2 cm

Radiographic Findings

- Ureterogram (IVP, retrograde/antegrade ureterogram)
  - Nodular, mural-based, radiolucent small filling defects in ureteral lumen
  - Scalloping of ureteral margins (in profile view)
  - May have ragged appearance of ureteral wall due to confluent groups of tiny cysts

MR Findings

- MR urography (MRU)
  - By use of heavily T2-weighted sequences and post-contrast urographic-phase imaging, MRU offers high spatial and contrast resolution for evaluation of urothelial lesions
  - Irregular, beaded appearance of ureteral wall with numerous nodular filling defects protruding into lumen

Imaging Recommendations

- Best imaging tool
  - CT urography, or retrograde ureterography

CT Findings

- CT urography
  - Same abnormalities as IVP or ureterography
  - Coronal and sagittal reformats are very helpful for better depiction of lesions in ureteritis cystica

DIFFERENTIAL DIAGNOSIS

Blood Clots, Radiolucent Stones

- Fewer, larger, mobile

Air Bubbles

- From recent instrumentation or infection with gas-forming organisms
- Mobile

Multifocal Transitional Cell Carcinoma or Papillary Tumors

- Usually have more irregular surface
- Not as numerous as ureteritis cystica

Metastatic Melanoma

- Usually larger, fewer nodules

Impression From Crossing Vessels

- Usually not spherical; filling defects not as numerous

Chronic Infectious Ureteritis

- Ureteral involvement by tuberculosis and schistosomiasis results in long-segment or multifocal ureteral stricture
  - Commonly have calcification in ureteral and bladder wall
- Chronic infection/irritation predisposes to ureteritis cystica and these 2 conditions may coexist

PATHOLOGY

General Features

- Etiology
  - Likely the response to chronic inflammation or irritation
    - Seen in association with UTI, urolithiasis, chemotherapy, radiation, schistosomiasis, and instrumentation
  - Theory of pathogenesis: **von Brunn cell nest**
    - Inflammatory stimulation → proliferation of urothelium → invagination of urothelial cells
    - Cells become isolated within lamina propria → metaplastic transformation into glandular structures (**ureteritis glandularis**) → cystic dilation of von Brunn nests (**ureteritis cystica**)
    - These 3 patterns (florid von Brunn nests, ureteritis glandularis, and ureteritis cystica; covered under umbrella term **proliferative ureteritis**) are in continuum and usually present at the same time
- Not premalignant or neoplastic

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
  - Patient might have signs/symptoms due to coexistent UTI or stones

Demographics

- Age
  - ↑ incidence with age

Natural History & Prognosis

- Usually inconsequential; should rule out UTI
- No risk of malignant degeneration
  - Predisposing factors for ureteritis cystica (such as chronic infection) can act as risk factors for urothelial malignancy

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Ureteral Stricture

KEY FACTS

TERMINOLOGY

- Narrowing of ureteral lumen due to intraluminal, intramural, or extrinsic process

IMAGING

- Segmental narrowing of ureter with dilation of upstream ureter on pyelography
- Best imaging tool
  - Retrograde pyelography for best resolution of length and degree of stenosis
- CT or MR for extrinsic processes
  - e.g., retroperitoneal fibrosis or malignancy

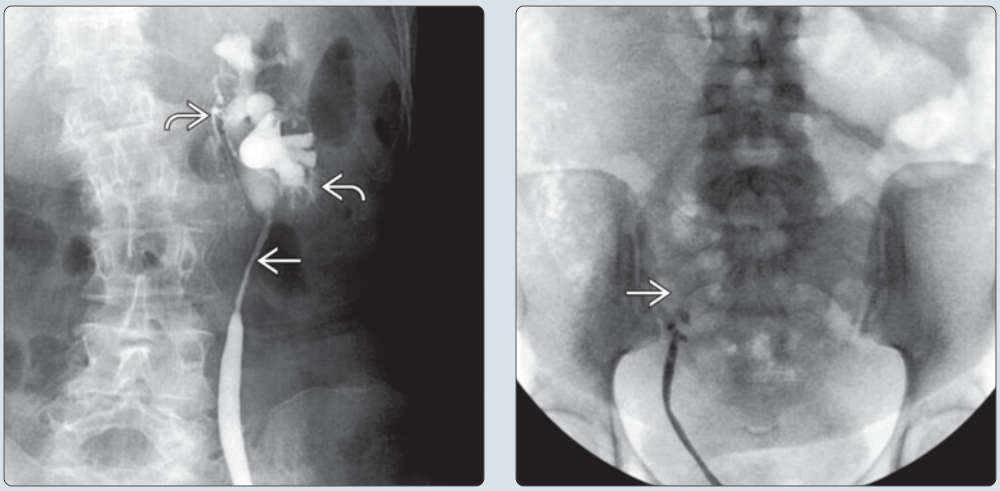
TOP DIFFERENTIAL DIAGNOSES

- Iatrogenic stricture
  - Most common etiology
  - Seen after lithotomy, ureteroscopy, ureteral catheterization, or radiation therapy
- Postoperative stricture
  - Most commonly at site of anastomosis of ureter to bladder, neobladder, or ileal conduit
- Urolithiasis
- Malignant stricture
  - Transitional cell carcinoma
  - Metastases and lymphoma
- Retroperitoneal fibrosis
- Malakoplakia
- Tuberculosis
- Congenital ureteral stricture
- Vascular compression

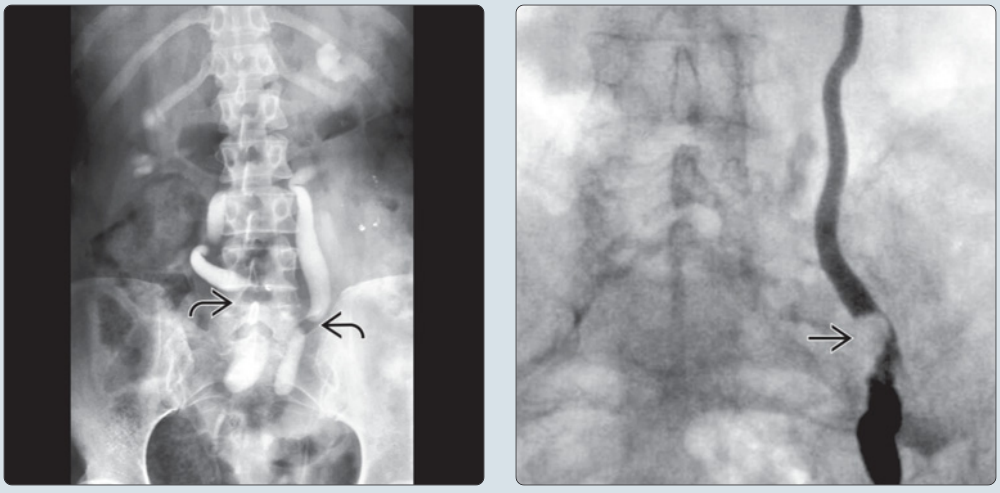
CLINICAL ISSUES

- Most strictures are benign, and are treated by catheter balloon dilation &/or stent placement

(Left) *Retrograde pyelography shows a long segment stricture of the proximal ureter with dilation of the calyces. Note the intravasation of contrast, an indication of overdistention of the collecting system under pressure. This resulted from the surgical extraction of a ureteral stone.* **(Right)** *Retrograde pyelogram shows complete obstruction of the right ureter in the pelvis due to a ureteral injury from gynecologic surgery. The findings were confirmed on antegrade pyelogram (not shown).*



(Left) *Retrograde pyelogram shows medial deviation and partial obstruction of both ureters, a classic feature of retroperitoneal fibrosis. The diagnosis was confirmed on CECT (not shown) as a mantle of tissue surrounding the aorta, inferior vena cava, and distal ureters.* **(Right)** *Retrograde pyelogram shows an eccentric filling defect and stricture in the distal ureter, with both a smooth and an irregular surface component. This proved to be a malignant stricture from transitional cell carcinoma.*



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TERMINOLOGY

Definitions

- Narrowing of ureteral lumen due to intraluminal, intramural, or extrinsic process

IMAGING

General Features

- Best diagnostic clue
  - Segmental narrowing of ureter with dilation of upstream ureter on pyelography

Imaging Recommendations

- Best imaging tool
  - Retrograde pyelography for most detailed depiction of length and degree of stenosis
  - CT or MR for extrinsic processes
    - e.g., retroperitoneal fibrosis or malignancy

DIFFERENTIAL DIAGNOSIS

Iatrogenic Stricture

- Most common etiology
- Seen after lithotomy, ureteroscopy, ureteral catheterization
- Often secondary to edema in immediate postprocedure period
  - Most cases are transient and resolve spontaneously
- Also seen following radiation therapy for pelvic malignancy

Postoperative Stricture

- Most commonly at site of anastomosis of ureter to bladder, neobladder, or ileal conduit
- Also seen following nonurologic abdominal surgery (open or laparoscopic)
  - Often result of ischemic injury to ureter
  - Dissection near or mobilization of ureters can avulse their fragile blood supply
    - → chronic strictures, unlikely to resolve

Urolithiasis (Renal Calculi)

- 25% of patients with prolonged stone impaction (> 2 months) develop stricture
- Most strictures are secondary to edema and are transient, resolving in 6-12 weeks

Malignant Strictures

- Transitional cell carcinoma
  - May have multiple sites of involvement (bladder, renal pelvis, contralateral ureter)
  - Intraluminal filling defect with dilation of upstream ureter
- Retroperitoneal metastases
  - May be to ureters themselves or, more commonly, to retroperitoneal nodes
  - Can encase and obstruct ureters
  - PET CT (plus clinical history): Best means of documenting etiology of malignant strictures
- Lymphoproliferative processes: Lymphoma, Erdheim-Chester disease

Retroperitoneal Fibrosis

- Encases, obstructs, and medially displaces ureters
- CT shows mantle of soft tissue encasing aorta and inferior vena cava throughout lumbar region

Tuberculosis

- Secondary to rupture of renal focus and bacilluria
- Seen in 20% of patients with renal tuberculosis
- More common in proximal ureter
- Multiple foci of nodularity, fibrosis, calcification, wall thickening
- Pyelography: Beaded, corkscrew, or pipestem appearance of ureter
- Calcification may be seen on radiography and CT

Vasculitis

- Etiology: Necrotizing vasculitis, scleroderma, polyarteritis nodosa (PAN), Wegner granulomatosis, dermatomyositis, Henoch-Schönlein purpura, Churg-Strauss
- Mechanism: Vasculitis → ischemia → fibrosis
- Morphology: Short or long segment, single or multiple, unilateral or bilateral
- CT: Ureteral wall thickening; calcification may be seen
- Multiple short-segment strictures ("string of pearls") on pyelography: Suggestive of PAN or necrotizing vasculitis
- Bilateral involvement, middle 1/3 of ureters, calcification: Suggestive of dermatomyositis

Malakoplakia

- Inflammatory disease associated with chronic coliform bacteriuria
- Seen in immunocompromised patients; F > M
- Simultaneous bladder and renal lesions are seen; more common in bladder
- Due to defect of intracellular bacterial digestion
- Pyelography: Multiple smooth nodular filling defects in ureter
- Treatment: Long-term antibiotics, surgery, ureteral stent

Congenital Ureteral Stricture

- Failure to grow at specific ring areas of ureter
- Smooth ring-shaped narrowing on intravenous pyelogram

Vascular Compression

- Compression by aberrant or dilated/thrombosed gonadal vein at level of pelvic brim; more common on right
- Compression by IVC or iliac vessels (seen with aberrant [retroiliac/retrocaval] course of ureter)
- Usually results in mild dilatation of upstream ureter

CLINICAL ISSUES

Treatment

- Most strictures are benign, and are treated by catheter balloon dilation &/or stent placement

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Malakoplakia

KEY FACTS

TERMINOLOGY

- Rare chronic granulomatous condition affecting urinary tract
  - Thought to be due to altered host response and defective phagocytosis

IMAGING

- Bladder is most frequently involved organ (40%) followed by renal parenchyma, upper urinary tract, prostate, and urethra
- Variable appearance ranging from flat plaques to nodules and masses ± ulceration

TOP DIFFERENTIAL DIAGNOSES

- Transitional cell carcinoma
- Leukoplakia
- Ureteritis cystica
- Chronic granulomatous ureteritis (tuberculosis)
- Nephrogenic adenoma

PATHOLOGY

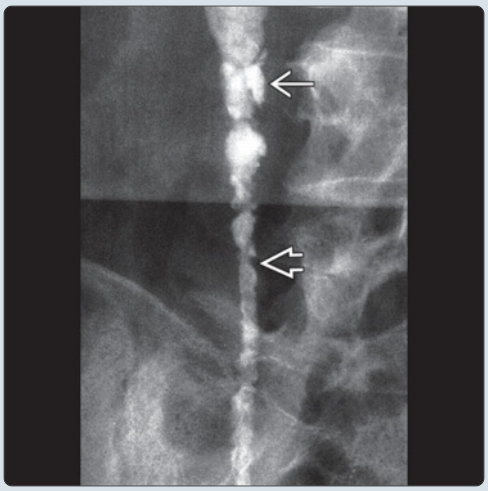
- Thought to be due to impaired host response and defective phagocytosis
- Highly associated with *Escherichia coli* and other gram-negative bacilli
- More common in immunosuppressed and diabetic patients
- Distinctive histologic appearance
  - Mass-forming histiocytic infiltrate with granular eosinophilic cytoplasm (von Hansemann cells)
  - Characteristic basophilic intracytoplasmic inclusions (Michaelis-Gutman bodies)

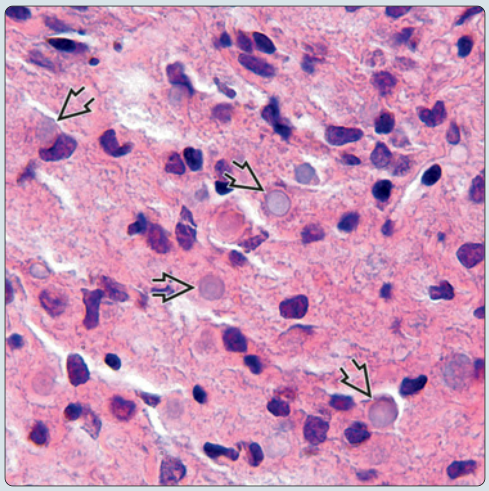
CLINICAL ISSUES

- No malignant potential

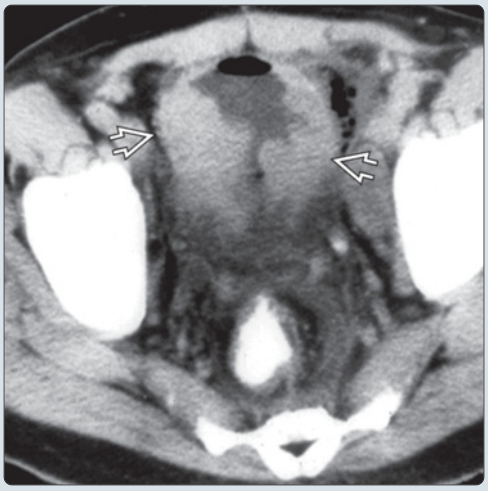
DIAGNOSTIC CHECKLIST

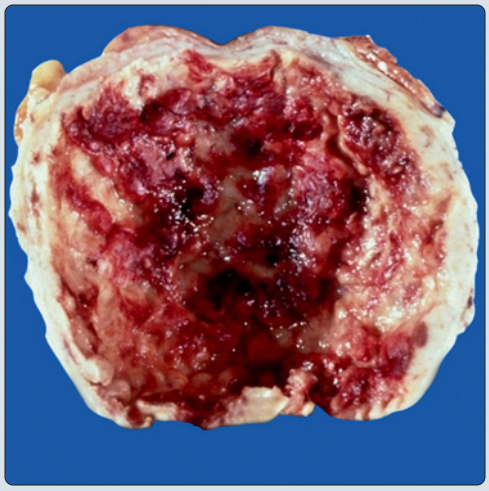
- Imaging manifestations are nonspecific and there is overlap with other inflammatory and neoplastic conditions





(Left) *Coned-down view of the right ureter in a case of malakoplakia shows diffuse irregularity with small plaques and ulcerations. Urinary tract involvement can be quite variable from a focal lesion to diffuse abnormality.* **(Right)** *High-power H&E stain shows sheets of histiocytes (von Hansemann cells) with lamellated intracytoplasmic inclusions (Michaelis-Gutman bodies). Malakoplakia is felt to be the result of defective phagolysosomal activity in patients with recurrent E. coli infections. (From DP: Genitourinary)*





(Left) *CECT at the level of the bladder shows irregular mass-like thickening of the bladder wall. This appearance is indistinguishable from urothelial carcinoma and requires biopsy.* **(Right)** *Photograph of the cut, resected specimen shows a friable, hemorrhagic surface and dramatic wall thickening. The bladder is the most frequent site of malakoplakia followed by the renal parenchyma, upper urinary tract, prostate, and urethra.*

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TERMINOLOGY

Definitions

- Rare chronic granulomatous condition affecting urinary tract; thought to be due to altered host response and defective phagocytosis

IMAGING

General Features

- Location
  - Bladder is most frequently involved organ (40%) followed by renal parenchyma, upper urinary tract, prostate, and urethra
- Size
  - Variable from small plaques to involvement of entire ureter and bladder
- Morphology
  - Variable; ranging from flat plaques (< 5 mm) to nodules and masses ± ulceration

Radiographic Findings

- Ureterogram (excretory or retrograde)
  - Solitary or multiple broad-based plaque-like or polypoid lesion(s)
  - Luminal narrowing and stricture

CT Findings

- Variable findings ranging from small plaque-like or polypoid lesions to large ulcerated solid masses
- Circumferential ureteral wall thickening
- In rare cases, lesion can act aggressively and extends beyond ureter and bladder
- Delayed phase (CT urogram) is extremely useful for better delineation of lesions

DIFFERENTIAL DIAGNOSIS

Transitional Cell Carcinoma, Ureter

- Imaging findings of urothelial carcinoma and malakoplakia overlap

Leukoplakia

- Rare condition manifested by squamous metaplasis of urothelium; more common in bladder
- Exact etiology is unknown; most accepted hypothesis is reactive metaplasia in response to chronic/repetitive urothelial irritation (chronic infection, calculi, etc.)
- Imaging findings are nonspecific: Solitary or multiple plaque-like lesion(s)
- Premalignant lesion with up to 28% chance of malignant transformation

Ureteritis Cystica

- Reactive proliferative changes of urothelium with formation of multiple small subepithelial cysts
- Lesions are numerous and have smooth surface; more common in upper 1/3 of ureter

Chronic Granulomatous Ureteritis (Tuberculosis)

- Results in multifocal or long-segment stricture ± calcification

Nephrogenic Adenoma

- Rare nonneoplastic urothelial lesion caused by urothelial irritation (infection, calculi, instrumentation, etc.)
- Solitary or multiple; plaque-like or papillary/polypoid lesions
- Impossible to distinguish based on imaging alone; biopsy is essential for diagnosis

PATHOLOGY

General Features

- Etiology
  - Thought to be due to impaired host response and defective phagocytosis
  - Highly associated with *Escherichia coli* and other gram-negative bacilli

Gross Pathologic & Surgical Features

- Variable gross features (as seen on cystoscopy)
  - Soft yellow, or yellow-brown, plaques with varying size and number; nodules or polyps
  - Central ulceration can be seen with larger lesions

Microscopic Features

- Mass-forming histiocytic infiltrate with granular eosinophilic cytoplasm (**von Hansemann cells**)
- Histiocytes contain characteristic basophilic intracytoplasmic inclusions (**Michaelis-Guttman bodies**)
  - Michaelis-Guttman bodies represent mineralized poorly digested bacteria

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Hematuria
  - Signs of bladder irritability (dysuria, frequency, hesitancy)

Demographics

- Age
  - Peak incidence in middle age (5th-7th decades)
- Gender
  - Predominantly seen in women (F:M = 4:1)

Natural History & Prognosis

- No malignant potential

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KEY FACTS

TERMINOLOGY

- Injury of ureter from blunt, penetrating, or iatrogenic trauma

IMAGING

- Best imaging tools: CECT with CT urography for global view of abdomen & urinary tract
  - Antegrade or retrograde pyelography for detailed analysis of site & character of injury
- Sites of urine accumulation
  - Perirenal or subcapsular from blunt renal injury
  - Medial to UPJ for proximal ureteral & UPJ leaks
  - May enter peritoneal cavity (after penetrating trauma or laparotomy/laparoscopy)
- Ureteral strictures less accurately diagnosed by CT
  - Difficult to distinguish segments of stricture from ureteral spasm & peristalsis due to stricture
  - Indirect signs of ureteral stricture: Delayed nephrogram, hydronephrosis
- Image-guided aspiration of fluid collection may identify fluid as urine & guide drainage

TOP DIFFERENTIAL DIAGNOSES

- Abdominal abscess
- Postoperative hematoma or seroma
- Postoperative edema

PATHOLOGY

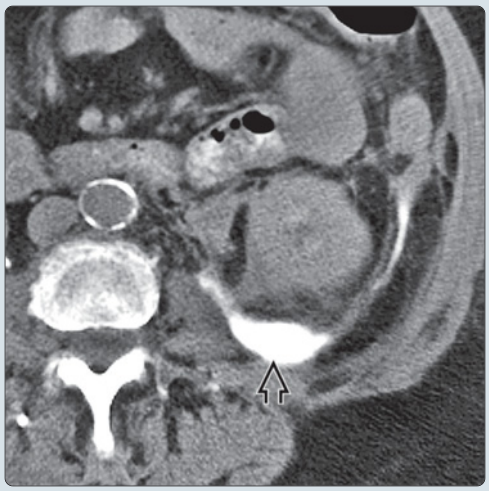
- Iatrogenic trauma accounts for 80-90% of ureteral injuries
  - Open abdominal or gynecologic surgery (> 50%)
  - Laparoscopic abdominal and gynecologic surgery
  - Ureteroscopy and endourologic interventions

CLINICAL ISSUES

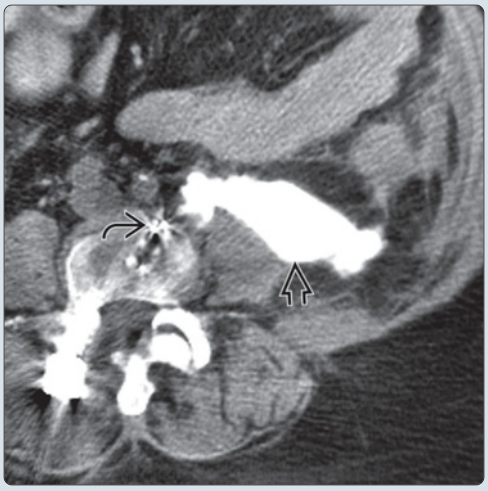
- Frequently delayed diagnosis
- Presents with fever, nausea, flank & low back pain, leukocytosis
- Fistula to skin or vagina

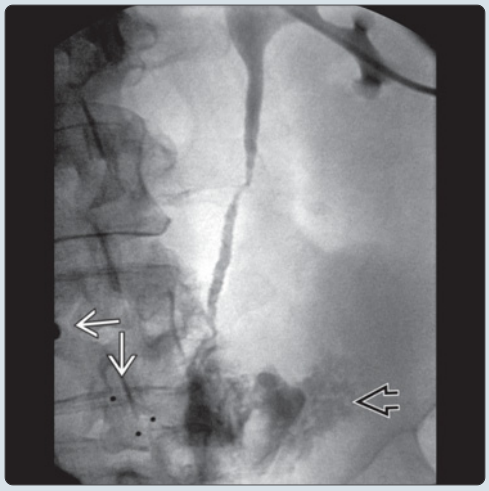
(Left) *Axial CECT in a patient following spine surgery shows fluid ⇨ in the left perinephric space. Differential considerations include hematoma and urinoma. CT was performed due to aberrant spinal screw placement and concern for retroperitoneal hemorrhage.* **(Right)** *Axial CECT during the excretory phase in the same patient demonstrates opacification of the perinephric fluid consistent with extravasated urine ⇨ due to ureteral injury.*





(Left) *More inferiorly, axial CECT during the excretory phase in the same patient shows increased accumulation of the extravasated urine ⇨. Note the metallic spinal hardware ⇨ along the ventral margin of the vertebral body.* **(Right)** *AP image during antegrade nephrostogram demonstrates transection of the distal aspect of the left ureter and extravasation of contrast material ⇨ into the retroperitoneum. Note the spine hardware ⇨ at the same level.*





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TERMINOLOGY

Definitions

- Injury of ureter from blunt, penetrating, or iatrogenic trauma

IMAGING

General Features

- Best diagnostic clue
  - Extravasation of contrast-opacified urine during CT urography or pyelography
- Location
  - > 80% affect inferior 3rd of ureter
- Morphology
  - Avulsion, perforation, or stricture

MR Findings

- MR urography: Can be performed ± IV contrast material
  - Stricture: Focal ureteral narrowing ± upstream dilatation
  - Laceration: Luminal truncation with extravasation of urine
- Urinoma/leak: ↓ signal intensity (SI) on T1WI; ↑ SI on T2WI

Ultrasonographic Findings

- Grayscale ultrasound
  - Perinephric or retroperitoneal fluid or fluid collections in proper clinical setting may be 1st indication of ureteral injury
    - Urinoma: Anechoic retroperitoneal fluid collection with through transmission
      - No internal flow or echoes
    - Ultrasound can be used to guide percutaneous aspiration

Imaging Recommendations

- Best imaging tool
  - CECT with CT urography for global view of abdomen and urinary tract
    - CT urography: Unenhanced, nephrographic, and excretory phases
    - Antegrade or retrograde pyelography for detailed analysis of site and characterization of injury
- Protocol advice
  - Include excretory (10-12 minutes post IV injection) phase images from CECT
    - View in multiplanar reconstructed images; 3D CT urography

Radiographic Findings

- IVP
  - Poor sensitivity (33%) in identifying nature of acute ureteral injuries
  - Better for diagnosis of delayed injuries with well-formed tract & collections (e.g., urinoma, sinus tracts)
  - Hydronephrosis
    - Common but nonspecific sign following any form of ureteral injury, even transient & mild
  - Extravasation of opacified urine
    - May be identified from any site of perforation
    - Avulsion or perforation of ureteropelvic junction (UPJ)
      - Collection of urine medial to UPJ & proximal ureter
      - Complete avulsion: No opacification of distal ureter
- Ureterography (pyelography): Antegrade or retrograde
  - Considered gold standard for confirming presence & characteristics of ureteral injury
  - Antegrade (via nephrostomy); retrograde (via cystoscopy)

CT Findings

- NECT
  - Collection of near water density fluid in perirenal or other retroperitoneal spaces
- CECT
  - Excretory phase images
    - Progressive accumulation of contrast-opacified urine in cases of perforation
    - CT urography may clarify site & extent of perforation
      - Usually requires pyelography for detailed analysis & therapy
    - Sites of urine accumulation
      - Perirenal or subcapsular from blunt renal injury
      - Medial to UPJ for proximal ureteral and UPJ leaks
      - May extend superiorly into mediastinum or caudally into thigh along iliopsoas compartment
      - May enter peritoneal cavity (after penetrating trauma or laparotomy/laparoscopy)
  - Ureteral strictures less accurately diagnosed by CT
    - Difficult to distinguish strictured segments from ureteral spasm or peristalsis
  - Indirect signs of ureteral stricture
    - Delayed nephrogram & hydronephrosis

Nuclear Medicine Findings

- Renal scintigraphy with Tc-99m dimercaptosuccinic acid
  - Sequential dynamic & delayed imaging over abdomen
  - May demonstrate accumulation of radiotracer outside renal collecting system and ureter

Nonvascular Interventions

- Image-guided aspiration of collection may identify fluid as urine
  - May be initial clue in delayed diagnosis of ureteral injury

DIFFERENTIAL DIAGNOSIS

Abdominal Abscess

- Also appears as loculated collection in postoperative patient
- No enhancement of contents on excretory phase CECT or radionuclide scintigraphy
- Percutaneous needle aspiration useful for diagnosis and treatment

Postoperative Hematoma or Seroma

- Hematoma may appear reticulated or heterogeneous (similar to abscess) depending on its age & evolution of blood products
- No enhancement of internal contents

Postoperative Edema

- At site of ureteral anastomoses or near periureteral surgical site
- May mimic ureteral stricture

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- Usually resolves quickly; does not cause progressive hydronephrosis

PATHOLOGY

General Features

- Iatrogenic trauma accounts for 80-90% of ureteral injuries
  - Injury from open or laparoscopic surgery
    - May result in perforation or transection of ureter
      - More often results in ischemic injury (due to stripping of fragile ureteral vessels)
    - Often results in delayed diagnosis (70%)
      - Extravasated urine induces fibrotic reaction & ureteral stricture
      - May result in progressive hydronephrosis & loss of renal function
  - Open abdominal or gynecologic (GYN) surgery
    - GYN surgery (> 50%)
      - Radical hysterectomy > salpingo-oophorectomy > cesarean section
      - 80% of ureteral injuries from GYN surgery not recognized at time of surgery
      - Delayed diagnosis → increased morbidity and mortality
    - Urologic surgery (30%)
      - Open prostatectomy, cystectomy, nephrectomy, etc.
      - 80% of ureteral injuries caused by open urological surgery recognized & treated at time of surgery
      - Anastomotic leaks from ureterovesical junction (renal transplantation, following cystectomy with urinary diversion, etc.)
    - Other abdominal surgery (5-20%)
      - Colectomy > aortic aneurysm repair > aortofemoral bypass surgery > spine surgery
  - Increasing trend toward more injuries from less invasive procedures
    - Laparoscopic abdominal and GYN surgery (e.g., hysterectomy, oophorectomy)
    - Ureteroscopy and endourologic interventions
      - Occurs in 5-10% of procedures
      - Usually recognized at time of procedure
      - May be complication of ureteroscopic stone extraction (most common), repair of UPJ obstruction
      - Small leaks heal spontaneously
      - Larger leaks → urinomas; usually require percutaneous drainage & ureteral stent for several weeks
- Trauma
  - Penetrating (e.g., knife or gunshot)
    - Ureter may be perforated or transected
    - Usually accompanied by other abdominal injuries
    - Usually recognized and treated at time of injury
  - Blunt trauma
    - Rarely results in ureteral injury
    - Children may suffer whiplash injury with avulsion of UPJ
- Radiation therapy
  - External beam or interstitial
  - Usually causes stricture of distal ureter

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Delayed diagnosis frequently
  - Presents with fever, nausea, flank & low back pain, leukocytosis
- Other signs/symptoms
  - Fistula to skin or vagina
    - Urine leakage
  - Urinoma may lead to electrolyte abnormalities
    - May become infected

Demographics

- Age
  - Older adults (iatrogenic injuries from surgery)
  - Younger adults (iatrogenic injuries from stone extractions)

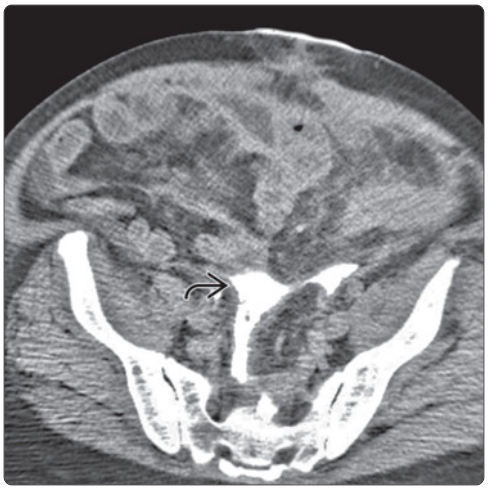
Natural History & Prognosis

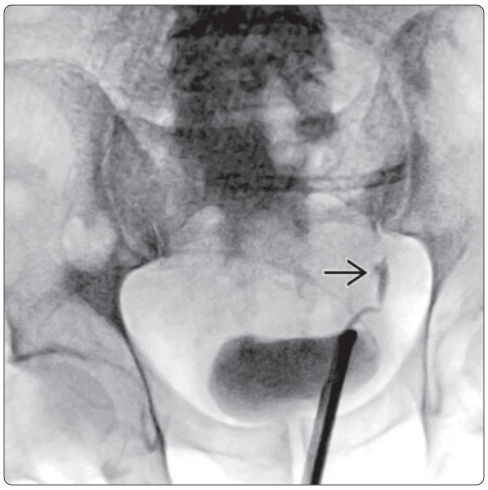
- Small leaks, which are promptly recognized & treated
  - Heals without sequelae
  - May require temporary ureteral stent
- Larger leak with urinoma
  - May require percutaneous drainage of urinoma and prolonged ureteral stenting
- Delayed diagnosis of leak
  - Often need to treat fibrotic stricture & leak
  - Balloon dilation of stricture, followed by long-term stenting
- Ureteral transection or intractable stricture
  - May require surgical reimplantation of ureter into bladder or neobladder (ileal conduit or other variation)
  - Native bladder may need to be mobilized as part of "psoas hitch" procedure to reduce tension at anastomosis with shortened ureter

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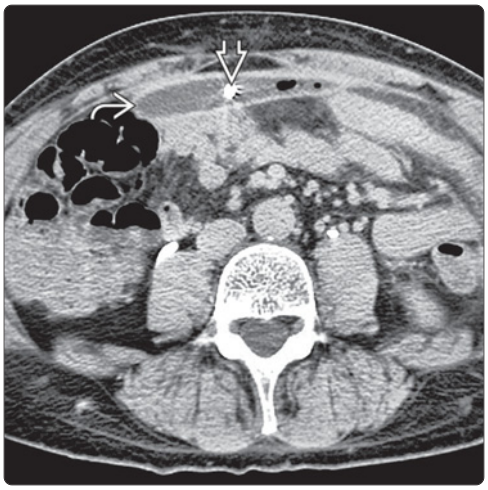
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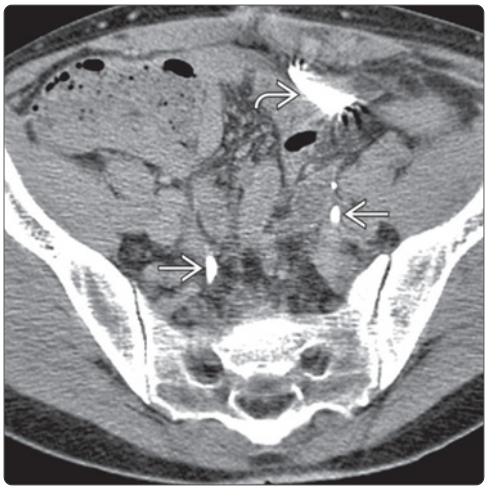
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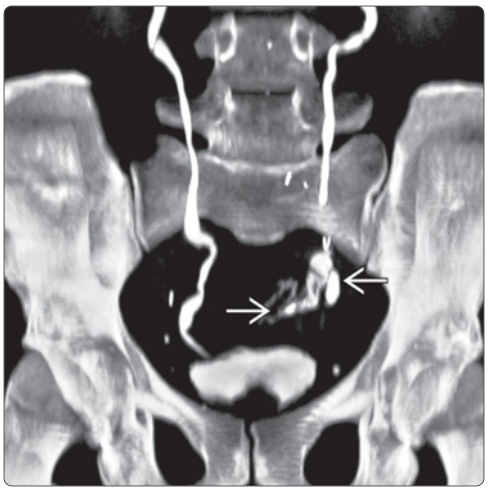


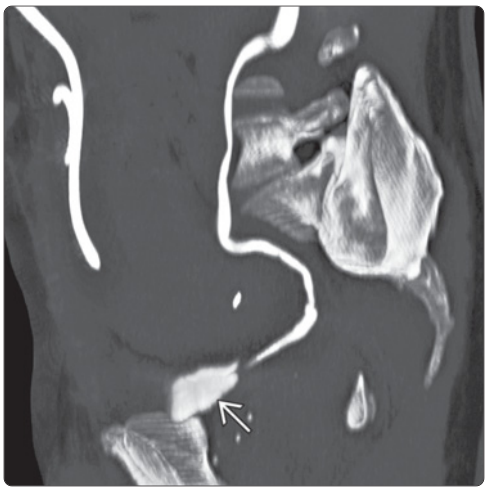
(Left) *Axial CECT during the excretory phase following subtotal colectomy demonstrates extravasation of contrast-opacified urine into a collection near the site of the colonic anastomosis due to ureteral transection.* **(Right)** *Retrograde pyelogram in the same patient shows truncation of the left ureter and subtle extravasation. A ureteral transection was confirmed at reoperation.*





(Left) *Axial CECT shows an elliptical fluid collection in the ventral aspect of the abdomen following partial colectomy. A portion of a percutaneous drainage catheter is shown in the center of the fluid collection; the aspirated contents were suspicious for infected urine.* **(Right)** *Excretory phase CECT shows a more caudal section in the same patient. Note the dense opacification of the ureters and contrast material filling the fluid collection around the catheter.*





(Left) *Coronal multiple intensity projection from excretory phase CECT in the same patient shows extravasation of contrast-opacified urine from the transected left ureter. Note that the more distal ureter is not opacified. The extravasated urine collected within the previously identified elliptical collection.* **(Right)** *Coronal oblique reformatted image from an excretory phase CECT in the same patient clearly shows the extravasation of urine from the distal left ureter.*

Polyps

KEY FACTS

TERMINOLOGY

- Synonym: Ureteral fibroepithelial polyp
- Definition: Benign mesodermal lesion with hyperplastic fibroconnective stromal core and normal urothelial lining

IMAGING

- Elongated soft tissue density intraluminal mass with smooth surface
- Shows enhancement
- CT or US may show protrusion of polyp through ureteral orifice into bladder
- ± hydroureter/hydronephrosis
- Delayed imaging (CT urography) helps to better delineate shape and contour of lesion
- CT is helpful in excluding other causes of obstruction such as stone, etc.

TOP DIFFERENTIAL DIAGNOSES

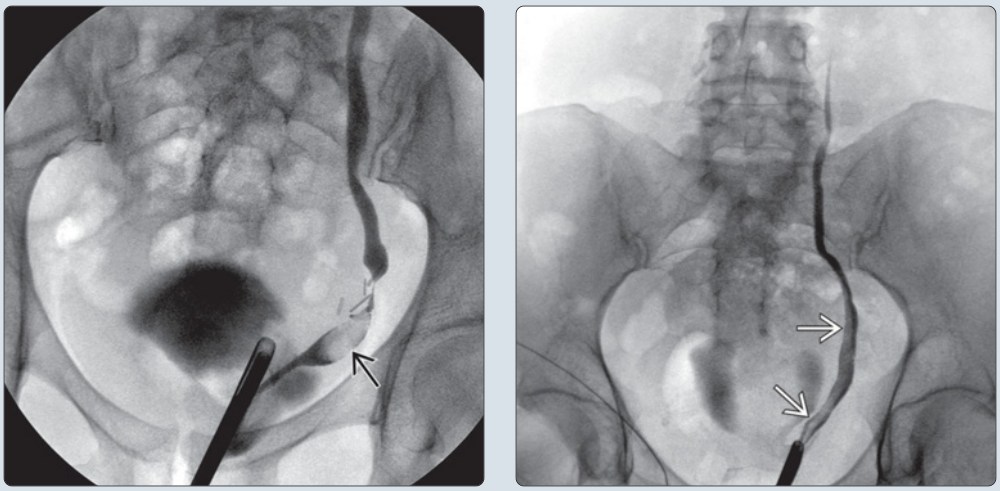
- **Urothelial neoplasms**
  - Papillary urothelial neoplasms have exophytic growth and may have polypoid morphology
  - Papillary urothelial neoplasms are usually smaller and fixed
- **Other mesenchymal ureteral tumors**
  - Leiomyoma, neurofibroma, lymphangioma, hemangioma
- **Intraluminal filling defects**
  - Stone, blood clot, debris, sloughed papillae, fungus ball, and foreign body
  - CT is particularly helpful to differentiate: Evaluate morphology and attenuation

PATHOLOGY

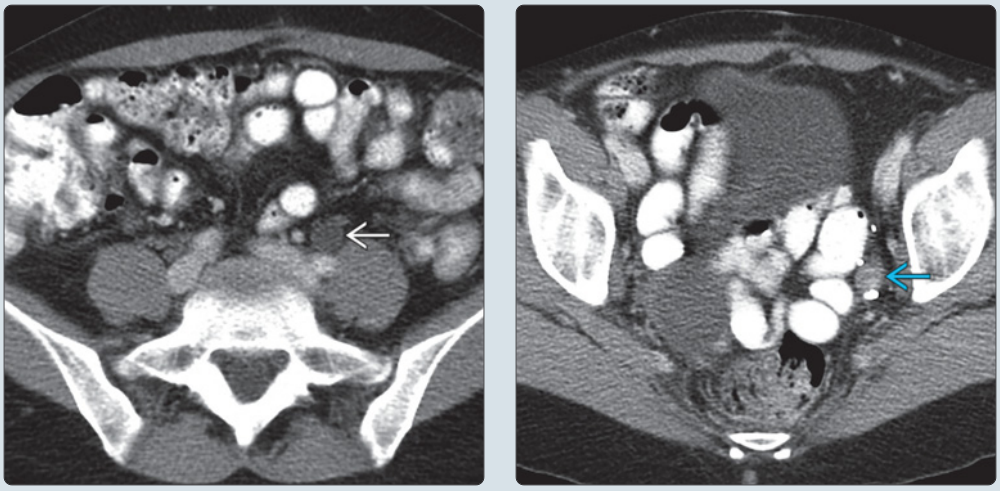
- Exact etiology is unknown
- Congenital factors and chronic inflammation play role

CLINICAL ISSUES

- Transurethral excision



(Left) *AP retrograde urography shows a well-delineated intraluminal filling defect in the distal left ureter. Pathology was consistent with a fibroepithelial polyp. The surgical clips in the left pelvis are related to a prior hysterectomy.* **(Right)** *AP retrograde urogram shows a well-delineated, elongated intraluminal filling defect in the distal left ureter. Cystoscopy showed a polypoid lesion with a long stalk, which intermittently protruded through the ureteral orifice.*



(Left) *Axial CT at the pelvic brim shows moderate dilatation of the left ureter.* **(Right)** *CECT section through the lower pelvis, in the same patient, shows enhancing soft tissue within the distal left ureter. Subsequent retrograde urography and cystoscopy confirmed an intraluminal polypoid lesion. The patient underwent transurethral excision of a polyp and pathology was consistent with a fibroepithelial polyp.*

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TERMINOLOGY

Synonyms

- Ureteral fibroepithelial polyp

Definitions

- Benign mesodermal lesion with hyperplastic fibroconnective stromal core and normal urothelial lining

IMAGING

General Features

- Best diagnostic clue
  - Elongated, soft tissue density intraluminal filling defect with smooth surfaces
- Location
  - More common on left side
  - More common in upper 1/3
- Size
  - Variable; can be as large as 12 cm
- Morphology
  - Elongated intraluminal filling defect with smooth surface

Radiographic Findings

- **Ureterography (intravenous or retrograde)**
  - Long intraluminal filling defect with smooth surface
  - By using supraphysiologic pressure, retrograde ureterography better delineates lesion
  - ± hydroureter/hydronephrosis

CT Findings

- Elongated soft tissue density intraluminal mass with smooth surface
- Shows enhancement
- Thin, slender lesions have corkscrew or vermiform appearance
- CT might show protrusion of polyp through ureteral orifice into bladder
- ± hydroureter/hydronephrosis
- Delayed imaging (CT urography) helps to better delineate shape and contour of lesion
- CT is helpful in excluding other causes of obstruction such as stone, etc.

Ultrasonographic Findings

- Ureteral polyp protruding into bladder is seen as vermiform, mobile lesion on US
- Mobility is helpful feature for exclusion of urothelial lesion (urothelial lesions are fixed and immobile)
- Doppler will show tumoral vascularity (helpful to differentiate from blood clot)

Imaging Recommendations

- Best imaging tool
  - CT urography is the preferred imaging modality for evaluation of obstructing lesions

DIFFERENTIAL DIAGNOSIS

Transitional Cell Carcinoma

- Papillary urothelial neoplasms have exophytic growth pattern and may have polypoid morphology
- Papillary urothelial neoplasms are usually smaller and fixed

Other Mesenchymal Ureteral Tumors

- Leiomyoma, neurofibroma, lymphangioma, hemangioma
- Not typically elongated and intraluminal

Intraluminal Filling Defects

- Stone, blood clot, debris, sloughed papillae, fungus ball, and foreign body
- CT is particularly helpful to differentiate these conditions, by evaluation of morphology and attenuation

PATHOLOGY

General Features

- Etiology
  - Exact etiology is unknown
  - Congenital factors and chronic inflammation play role

Gross Pathologic & Surgical Features

- Polypoid pink tan lesion with smooth surface

Microscopic Features

- Loose vascular fibrous stroma covered with normal transitional epithelium

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Flank pain, hematuria
  - Flank pain is usually intermittent, attributed to transient intussusception, obstruction, or torsion

Demographics

- Age
  - Most cases present in 20-40 years
- Gender
  - Slight male predominance (M:F = 3:2)
- Epidemiology
  - Rare; exact prevalence is unknown
  - Considered most common benign nonepithelial tumor of ureter

Natural History & Prognosis

- Benign lesion with no malignant potential

Treatment

- Transurethral excision

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Ureteral Transitional Cell Carcinoma

KEY FACTS

TERMINOLOGY

- Carcinoma arising from transitional epithelium of ureter
  - 5% of all urothelial malignancies
  - 90% of ureteral tumors are transitional cell carcinoma (TCC)

IMAGING

- Intraluminal soft tissue mass with dilation of ureter upstream
- More common in distal ureter (70%); presumably due to stasis
- CT urography is best noninvasive imaging tool for diagnosis and staging
  - ↑ sensitivity/specificity and ↑ accuracy compared with IVP
- MR urography (MRU) is a noninvasive alternative to CT urography in patients with contraindication for intravenous iodinated contrast
- Bilateral ureteral TCC seen in 2-5%

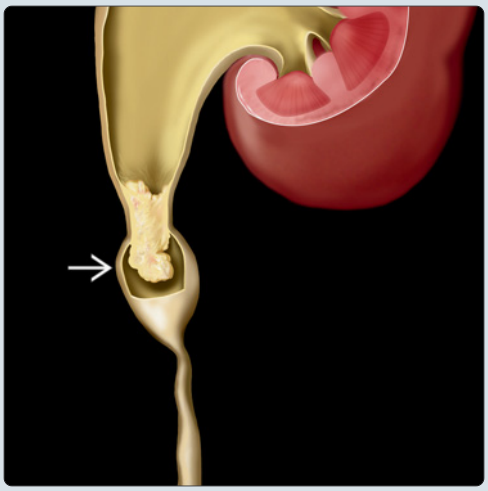
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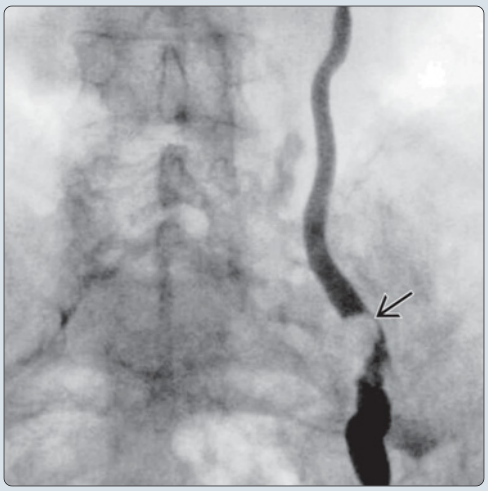
- Urolithiasis
- Blood clot
- Sloughed papilla (papillary necrosis)
- Fibroepithelial polyp
- Ureteritis cystica
- Tuberculosis
- Ureteral malakoplakia

PATHOLOGY

- Risk factors
  - Smoking, dose dependent
  - Occupational exposure: Aniline, benzidine, aromatic amine (used in textile industry)
- ~ 50% of patients with upper urinary tract TCC will develop metachronous bladder TCC
- 12% will develop metachronous TCC of upper urinary tract within 28 months

(Left) *Graphic shows a transitional cell carcinoma of the proximal ureter with dilation of the renal pelvis. Note the focal expansion of the lumen ➡ at the site of the tumor, the origin of the goblet sign seen on pyelography and sometimes on urography as well.* **(Right)** *Retrograde pyelogram in an 87-year-old man shows an eccentric filling defect ⇨ in the distal ureter, with both a smooth and an irregular surface component.*





(Left) *Axial CECT in an elderly man with hematuria shows circumferential wall thickening and luminal narrowing of distal left ureter ➡ just proximal to the ureterovesical junction.* **(Right)** *Volume-rendered 3D image from the excretory phase of CECT (same patient) shows the filling defect ➡ within the distal ureter that proved to be a transitional cell carcinoma of the ureter. Such reformations are complementary to the axial sections, but should not be viewed in isolation.*





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TERMINOLOGY

Abbreviations

- Transitional cell carcinoma (TCC)

Synonyms

- Urothelial cell carcinoma

Definitions

- Carcinoma arising from transitional epithelium of ureter (urothelium)

IMAGING

General Features

- Best diagnostic clue
  - Intraluminal soft tissue mass with dilation of upstream ureter
- Location
  - More common in distal ureter; presumably due to more stasis
    - Upper ureter: 3%
    - Mid ureter: 24%
    - Distal ureter: 73%
  - Bilateral in 2-5%

Radiographic Findings

- Radiography
  - Calcification may be seen in 2-7% of cases, resembling calculus
- IVP
  - Ureteral filling defect or wall thickening
  - Hydronephrosis or nonfunctioning kidney
  - Fixation of ureter with irregular narrowing of lumen (circumferential or eccentric)
- Retrograde pyelography
  - Invasive alternative to CT urography in patients with contraindication to intravenous contrast
  - Mucosal irregularity (frond-like tumoral surface)
  - Tumor adherent to wall, may be pedunculated
  - **Goblet sign**: Dilation of ureteral lumen at the level of tumor

CT Findings

- ↑ sensitivity/specificity and ↑ accuracy compared with IVP
- Intraluminal ureteral soft tissue lesion (30-60 HU)
- Variable enhancement of tumor
- Ureteral dilatation at level of lesion and upstream
  - CT equivalent of goblet sign
- Wall thickening (circumferential or eccentric)
- Periureteral invasion and lymph node involvement
- Also provides information for staging, such as nodal status and presence of metastatic disease

MR Findings

- MR urography (MRU) is noninvasive alternative to CT urography in patients with contraindication to intravenous iodinated contrast
- Spatial resolution of MRU is inferior to CT and sensitivity is lower (63-88%; depending on size of lesion)

Ultrasonographic Findings

- Limited role
- May show hydronephrosis and proximal hydroureter

Imaging Recommendations

- Best imaging tool
  - CT urography with nephrographic and urographic phases

DIFFERENTIAL DIAGNOSIS

Urolithiasis (Renal Calculi)

- Not contiguous with ureteral wall
- More homogeneous and higher attenuation than tumor

Blood Clot

- High attenuation on NECT
- Not contiguous with ureteral wall

Renal Papillary Necrosis

- Nonenhancing sloughed papilla
- Not contiguous with ureteral wall
- Clubbed calyces

Radiation Ureteritis

- Longer segmental thickening of ureteral wall
- Segmental narrowing of ureteral lumen
- Correlate with history of radiation therapy

Fibroepithelial Polyp

- Benign mesodermal lesion with hyperplastic fibroconnective stromal core and normal urothelial lining
- Cannot be confidently distinguished from TCC by imaging alone
- Usually elongated with smooth surface; may protrude through ureteral orifice into the bladder
- Papillary urothelial neoplasms are usually smaller and fixed

Ureteritis Cystica

- Multiple small subepithelial cysts in ureteral wall
- Seen in proximal 1/3 of ureter

Tuberculosis

- Often associated with renal tuberculosis
- Volume loss, renal scarring, caliectasis
- Parenchymal calcifications

Small Cell Carcinoma

- Rare (< 5% of ureter cancers)
  - Typically seen in elderly
  - Worse prognosis
- Associated with chronic inflammation and analgesic abuse

Ureteral Malakoplakia

- Rare chronic inflammatory process presenting as solitary or multiple plaques or polypoid lesions

PATHOLOGY

General Features

- Etiology
  - Field change of urothelium
    - Smoking (increases risk by 100-200%)
    - Cyclophosphamide (lag time ~ 6 years)
    - Structural anomalies with stasis of urine
    - Occupational exposure: Aniline, benzidine, aromatic amine (used in textile industry)

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  - Chronic analgesic abuse, chronic UTI
  - Balkan nephropathy (100-200 times increased risk)
- Associated abnormalities
  - 12% will develop metachronous TCC of upper urinary tract within 28 months
  - ~ 50% of patients with upper urinary tract TCC will develop metachronous bladder TCC

Staging, Grading, & Classification

- Stage 0: Papillary carcinoma and carcinoma in situ
- Stage I: Spread through urothelium and into connective tissue of ureteral wall
- Stage II: Spread though connective tissue into muscle layer
- Stage III: Spread into periureteral fat
- Stage IV: Spread into adjacent organ, lymph nodes, or distant metastases

Gross Pathologic & Surgical Features

- Morphologic classification
  - Grade refers to degree of cellular dysplasia
    - Some pathologists use as many as 7 grades of dysplasia or anaplasia
      - Papillary (most common type, usually of low grade)
      - Infiltrating (usually of high grade)
      - Mixed papillary and infiltrating
      - Carcinoma in situ
- Pathologic staging criteria
  - T1: Tumor invades subepithelial connective tissue
  - T2: Tumor invades muscularis
  - T3: Tumor invades periureteral fat
  - T4: Tumor invades adjacent organs or through kidney into perirenal fat
- Bilateral lesions seen in 2-9%

Microscopic Features

- Urine cytology and brushing from surface of urothelium are standard methods of confirming diagnosis
  - Occasional false-positive and false-negative cytology studies
    - False-positive can result from chronic irritation of urothelium (e.g., by chronic calculus disease) causing cellular atypia

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Gross or microscopic hematuria
  - Dull flank pain

Demographics

- Mean age: 64 years
- More common in men (M:F = 3:1)
- 5% of all urinary tract tumors

Natural History & Prognosis

- Tumor stage and histologic grade are most important prognostic factors
- ~ 85% of ureteral TCCs are low stage and superficial

Treatment

- Endoscopic resection, segmental ureterectomy, nephroureterectomy
- Surveillance for metachronous disease
  - Cystoscopy with urine cytology
  - Urography (CT urogram, IVP, or retrograde pyelography) for surveillance of upper tract metachronous lesions

DIAGNOSTIC CHECKLIST

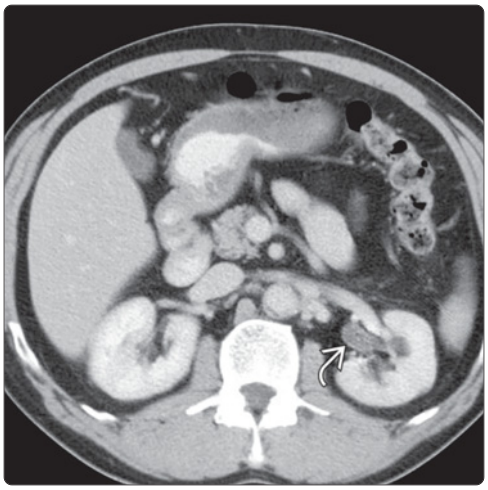
Image Interpretation Pearls

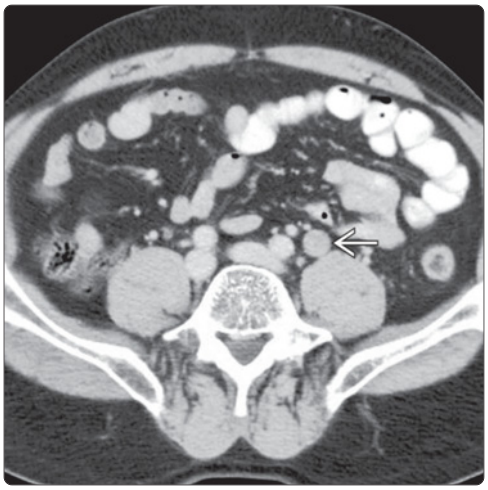
- Always check axial "source" CT images in addition to reformats
  - Can miss circumferential mass if only viewing CT urogram images

SELECTED REFERENCES

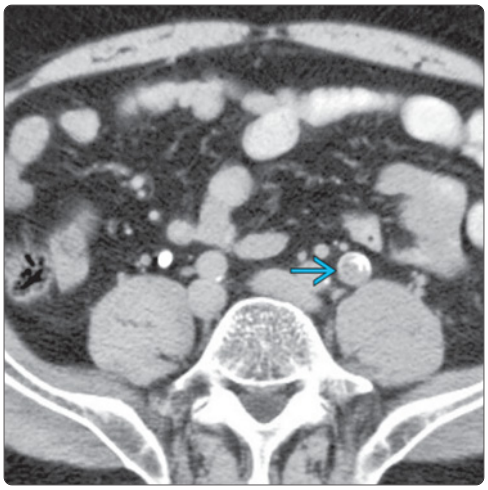
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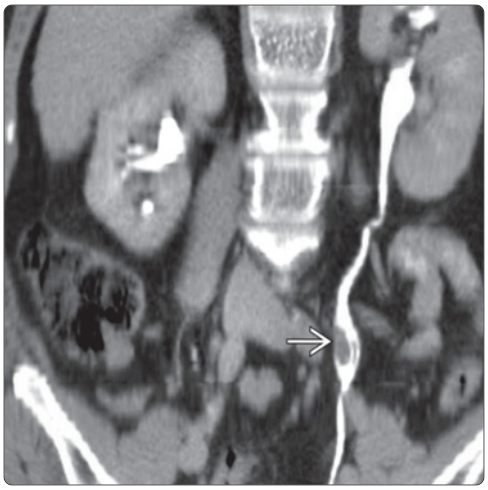
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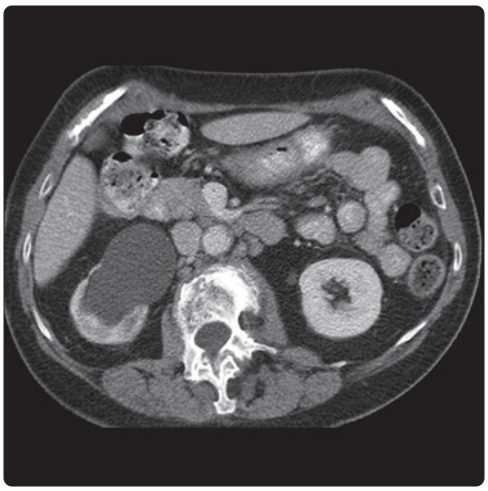


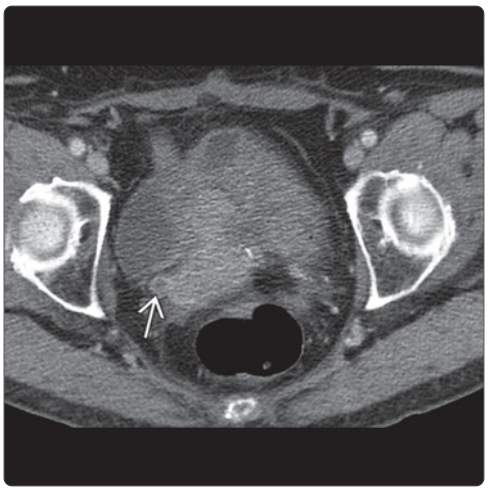
(Left) *Axial CECT shows left-sided hydronephrosis in an elderly man with hematuria and flank discomfort.* **(Right)** *A more caudal section in the same patient shows hydroureter.*





(Left) *Axial CT during delayed excretory phase through the same section shows contrast surrounding an irregular, soft tissue density filling defect within the ureteral lumen.* **(Right)** *Coronal CT urography images better shows the intraluminal filling defect. Note the goblet sign, indicating expansion of the ureter at the point of obstruction.*





(Left) *Axial CECT image through kidneys shows marked right-sided hydronephrosis with delayed nephrogram.* **(Right)** *Image through the bladder in the same patient shows marked hydroureter. Note luminal narrowing and circumferential wall thickening of distal ureter. The patient had history of local and external beam radiation for recurrent vaginal melanoma. Imaging findings in and of themselves are nonspecific. Given the history and distribution, however, these findings were attributed to radiation-induced ureteritis.*

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Ureterectasis of Pregnancy

KEY FACTS

TERMINOLOGY

- Physiologic dilatation of ureter and renal collecting system during pregnancy

IMAGING

- Ureteral dilatation down to point of compression (where ureter crosses iliac vessels)
- Ureter should tapers to normal diameter below pelvic brim; failure to do so is highly suggestive of pathologic obstruction
- Predominantly on right side (**80-90%**)
- Bilateral ureteral jets present
  - Ureteral jet might not be visible in supine position; turning patient into contralateral decubitus position helps to see jet
  - Obstruction is abrupt and is usually above or below level of common iliac arteries
  - ± posterior shadowing, ± twinkling artifact
  - Unilateral absence of ureteral jets

TOP DIFFERENTIAL DIAGNOSES

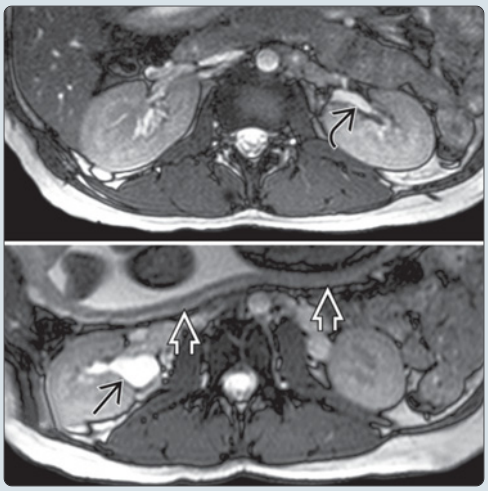
- Urolithiasis (renal calculi)

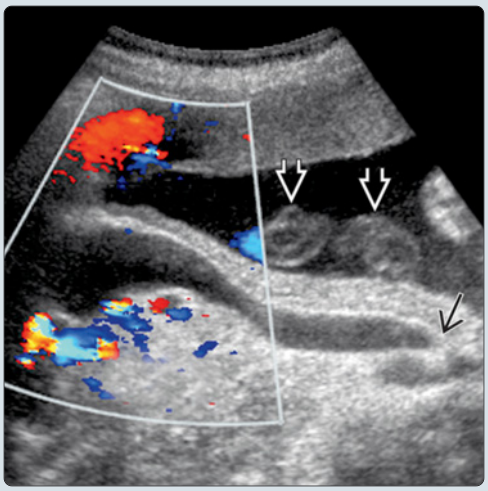
PATHOLOGY

- Caused by combination of extrinsic compression on ureter (more important factor) and intrinsic changes in ureters
- **Extrinsic compression**: Mainly by enlarged gravid uterus and, to lesser extent, by engorged parauterine vessels
- **Intrinsic changes**: ↓ ureteral tone and ↓ peristalsis caused by increased progesterone and gonadotropin levels

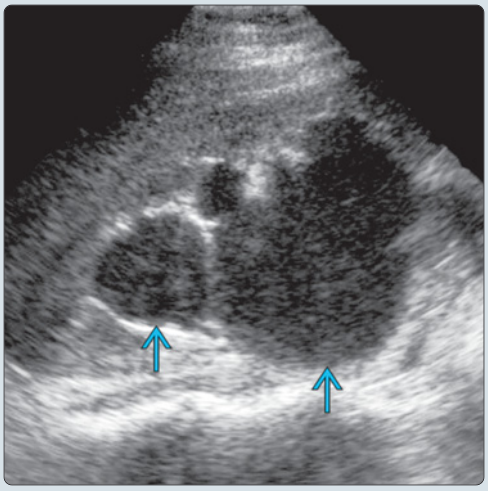
CLINICAL ISSUES

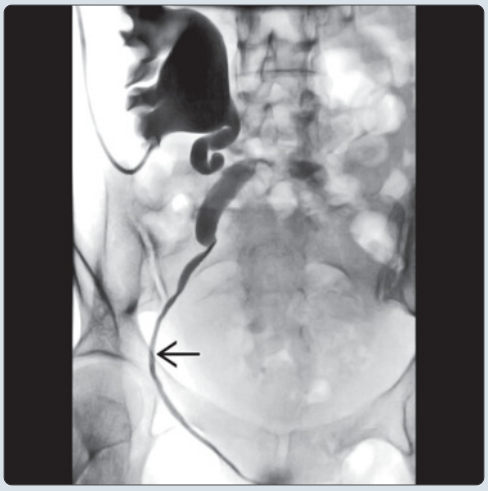
- Seen in ~ 90% of pregnant women in 3rd trimester
- Prevalence and severity increase as pregnancy advances





(Left) *Axial T2WI MR performed for evaluation of a fetal mass shows incidental mild dilatation of the right maternal collecting system, which extended to the pelvic brim. The left collecting system is normal. Note the posterior uterine wall.* **(Right)** *Longitudinal color Doppler US through the right kidney shows significant ureteral dilatation. The ureter tapers where it crosses the iliac vessels. Ureterectasis is more common on the right. Note loops of umbilical cord.*





(Left) *Longitudinal ultrasound through the right kidney shows unusually marked dilation of the right renal pelvis and calyces in a pregnant woman.* **(Right)** *Antegrade nephrostogram performed after delivery in the same patient shows lateral displacement of the distal right ureter by the enlarged uterus. Note the persistent dilation of the proximal right ureter and right collecting system despite placement of a nephrostomy catheter.*

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TERMINOLOGY

Synonyms

- Hydroureter of pregnancy, hydronephrosis of pregnancy, pelvicaliectasis of pregnancy

Definitions

- Physiologic dilatation of ureter and renal collecting system during pregnancy

IMAGING

General Features

- Best diagnostic clue
  - Dilatation of right ureter, tapering where it crosses common iliac arteries
- Location
  - Dilatation of ureter down to pelvic brim (where ureter crosses the common iliac vessels)
    - Pelvic ureter is always spared
  - Predominantly on right side (**80-90%**)
- Morphology
  - Ranges from mild to severe ureterectasis

Ultrasonographic Findings

- Ureteral dilatation down to point of compression (where ureter crosses iliac vessels), with resumption of normal caliber inferiorly
  - Color Doppler is helpful to differentiate ureter from dilated ovarian veins
  - Ureter should tapers to normal diameter below pelvic brim; failure to do so is highly suggestive of pathologic obstruction
    - In selected cases when there is high suspicion for distal ureteral stone, transvaginal or transrectal approaches may be needed to better evaluate distal ureter
    - Color Doppler "twinkle" artifact is helpful for detection of small stones in difficult locations
- Bilateral ureteral jets present
  - Ureteral jet may not visualized in a supine position; turning patient into contralateral decubitus position helps to see jet
  - An absent unilateral ureteral jet is suggestive of high-grade obstruction (usually due to stone)
- Spectral Doppler with calculation of resistive index (RI) of intrarenal arteries has potential role; however, sensitivity and specificity are low

MR Findings

- MR urography (MRU)
  - MRU is based on heavily T2-weighted fast spin-echo and MRCP sequences; **no intravenous contrast should be used**
  - Ureteral course can be delineated and cause and degree of obstruction can be better evaluated
  - Perinephric edema may suggest physiologically significant obstruction

DIFFERENTIAL DIAGNOSIS

Urolithiasis (Renal Calculi)

- Obstruction is abrupt and is usually above or below level of common iliac arteries
- ± posterior shadowing, ± twinkling artifact
- Absent unilateral jet

PATHOLOGY

General Features

- Etiology
  - Caused by combination of extrinsic compression on ureter (more important factor) and intrinsic changes in ureters
  - **Extrinsic compression**: Mainly by enlarged gravid uterus and, to lesser extent, by engorged parauterine vessels
    - Dextroflexion of uterus, course of right ureter (which crosses over iliac vessels with acute angle), and proximity of right ureter to right ovarian vein are hypothesized causes explaining more frequent involvement of right ureter
    - Point of maximum narrowing is at level of pelvic brim, where ureter crosses common iliac vessels
  - **Intrinsic changes**: ↓ ureteral tone and ↓ peristalsis caused by increased progesterone and gonadotropin levels

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most cases are asymptomatic
  - Can present with abdominal/flank pain
  - Urinary stasis predisposes to urinary tract infection

Demographics

- Epidemiology
  - Seen in ~ 90% of pregnant women in 3rd trimester
  - Can occur as early as 10th week of pregnancy
  - Prevalence and severity increase as pregnancy advances

Natural History & Prognosis

- Usually regresses within 8 weeks postpartum

Treatment

- Conservative treatment
- Postural drainage (lateral decubitus on opposite side of pain, knee chest position)
- Ureteral stenting
- Percutaneous nephrostomy
- Antibiotics for treating pyelonephritis indicated if patient is febrile

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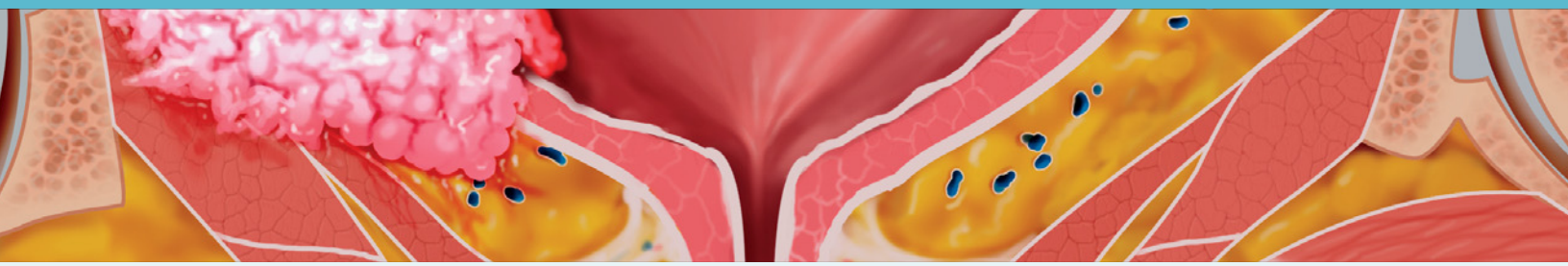
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SECTION 6

Bladder



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Embryology and Anatomy

The bladder is a hollow distensible viscus with a strong muscular wall. Embryologically, it forms from the urogenital sinus, which is contiguous with the allantois (a hindgut diverticulum that extends to the umbilicus). The allantois normally involutes by the 2nd month of gestation, forming the median umbilical ligament. Any persistent segments of the allantoic channel are called urachal remnants.

The distal ureters are incorporated into the posterolateral portion of the bladder wall at the trigone. The urethral orifice forms the distal apex of the trigone at the bladder base.

The bladder wall consists of 4 layers. The lumen is lined by uroepithelium, which consists of 3-7 layers of stratified flat cells. These cells are flexible and can change shape from cuboidal to flattened as the bladder distends (hence the term transitional epithelium). The 2nd layer is the lamina propria, which is very vascular. The 3rd layer is the detrusor muscle (muscularis propria). The detrusor muscle is a complex network of interlacing smooth muscle fibers. The inner and outer muscle fibers tend to be oriented in a longitudinal fashion, but distinct layers are usually not discernible. Fibers from the detrusor muscle merge with the prostate capsule (or anterior vagina in females) and pelvic floor muscles. A 4th adventitial layer is formed by connective tissue. A serosal covering, formed by the peritoneum, is present only over the bladder dome.

The bladder is located within the extraperitoneal space and is surrounded by loose connective tissue and pelvic fat. The perivesical space contains the bladder and urachus. The prevesical space (also called space of Retzius) extends anteriorly to the pubic symphysis and communicates posteriorly with the presacral space. These spaces can expand to contain large amounts of fluid as in an extraperitoneal bladder rupture or hemorrhage from pelvic fractures.

Imaging Techniques and Indications

Conventional Cystogram

Conventional cystography is primarily used to evaluate for bladder leak (either abdominal trauma or iatrogenic injury) or for fistulae (vesicocolic, vesicoenteric, vesicovaginal). Cystography has an accuracy rate of 90-100% for detecting bladder injury if performed properly.

A scout film should be performed in all cases before infusion of contrast. It is imperative that adequate bladder distention is obtained with a minimum of 300 cc of contrast. Small leaks are easily missed if the bladder is not appropriately distended. Ideally, the study should be performed under fluoroscopy with oblique images. However, in practice these patients often have pelvic fractures and are not mobile. Diagnostic images can still be obtained in this situation, but it is imperative to obtain images of a maximally filled bladder and a post-drainage film. The importance of the post-drainage film cannot be overemphasized because in ~ 10% of cases the injury is only seen on the post-drainage film.

Leaks may occur in the extraperitoneal space and appear as focal collections of contrast, or in the intraperitoneal space, in which the contrast is free flowing and outlines bowel loops. Fistulae are also well demonstrated with contrast accumulating in the small bowel, colon, or vagina.

CT Cystogram

CT cystography has an equal sensitivity and specificity to conventional cystography for bladder leaks and can be performed at the time of the initial trauma CT. Note that merely clamping the Foley catheter during a routine abdominopelvic CT is inadequate for evaluating bladder injury.

After routine CT, the bladder should be drained and a minimum of 300 cc of dilute contrast instilled followed by a repeat scan through the bladder. Multiplanar reformations in the coronal and sagittal plane have been shown to be helpful delineating the precise point of injury. Unlike with conventional cystography, a post-drainage film is not required.

CT and MR

Both modalities are suboptimal in evaluating the bladder wall. Underdistention can erroneously simulate wall thickening and small, sessile masses can be obscured with overdistention. The depth of mural invasion by tumors cannot currently be accurately determined with either modality. MR has moderate accuracy in assessment of T stage. Currently, the predominant role of imaging in bladder tumor evaluation is to differentiate organ-confined from non-organ-confined disease and to look for lymphadenopathy and distant metastasis. Continued advancements in multiparametric MR techniques may result in improved accuracy in T staging in the near future.

Ultrasound

The bladder is easily seen with transabdominal imaging, but such imaging is fraught with potential pitfalls. It is easy to overidentify bladder wall thickening unless the bladder is adequately distended. In addition, the anterior wall is poorly evaluated secondary to reverberation artifact. Color Doppler is useful to evaluate the vascularity of masses and presence of ureteral jets. Post-void residuals are also easily calculated.

Approach to the Bladder Mass

Pathologic conditions of the bladder can manifest as either a focal bladder mass or diffuse wall thickening. Focal masses are most commonly neoplastic but may develop secondary to congenital, inflammatory, or infectious processes. Some focal defects, such as ureteroceles, have a very diagnostic appearance. Extrinsic inflammatory diseases such as Crohn disease and diverticulitis may be associated with fistulae to the bladder and cause a focal bladder wall abnormality. In many cases the clinical, macroscopic, and radiologic findings for these masses may overlap; thus, histologic evaluation is often required.

Bladder neoplasms can arise from any of the bladder layers. They are broadly classified as either epithelial or nonepithelial (mesenchymal) neoplasia. Greater than 95% of bladder neoplasms are epithelial. Epithelial tumors that differentiate from normal urothelium are transitional cell carcinomas. The spectrum of neoplasia ranges from benign papilloma, to carcinoma in situ, to invasive carcinoma. Other primary epithelial tumors include squamous carcinoma and adenocarcinoma. Because epithelial masses derive from the most superficial layer of the bladder wall, they often appear as irregular, intraluminal filling defects.

Neoplasms derived from mesenchymal tissue differentiate toward muscle, nerve, cartilage, fat, fibrous tissue, and blood vessels. Benign tumors include leiomyoma, paraganglioma, fibroma, hemangioma, solitary fibrous tumor, neurofibroma, and lipoma. Malignant tumors include rhabdomyosarcoma, leiomyosarcoma, lymphoma, and osteosarcoma. Because

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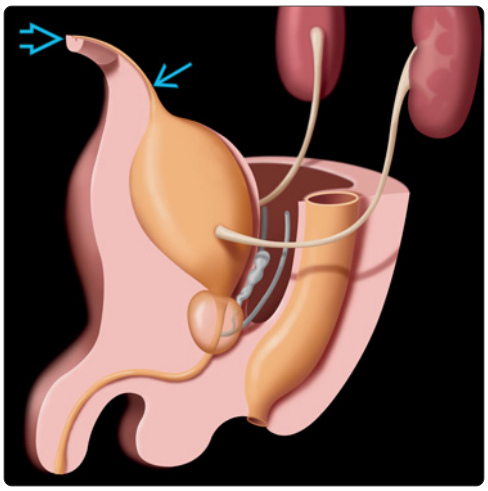
mesenchymal tumors arise from the submucosal portion of the bladder wall, they often appear as smooth intramural lesions. If large, however, they can ulcerate and be confused with a mucosal mass.

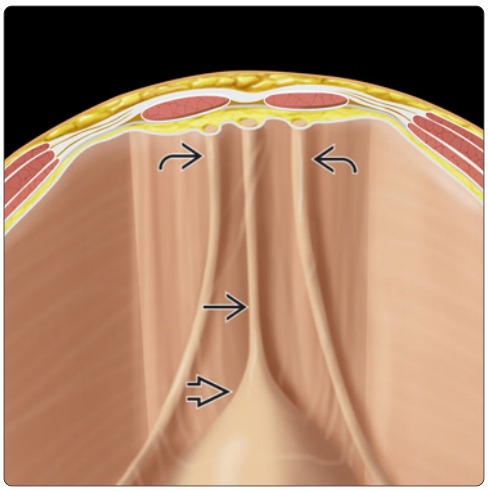
Some neoplasms can cause diffuse wall thickening (adenocarcinoma and lymphoma most commonly), but this is more typically seen in nonneoplastic disorders. Diffuse bladder wall thickening can develop secondary to many conditions, including infection with bacteria or adenovirus, schistosomiasis, tuberculosis, and inflammatory conditions, such as cystitis cystica, cystitis glandularis, or eosinophilic cystitis. Exposure to chemotherapy (particularly with cyclophosphamide) or irradiation also cause diffuse wall thickening. Although the radiologic characteristics of these disorders are less specific, radiologic evaluation is still valuable.

Cystitis appears as a thick-walled bladder with enhancing mucosa and perivesicular inflammatory changes. The various types of cystitis require pathologic diagnosis. Bladder infection with tuberculosis and schistosomiasis produce nonspecific bladder wall thickening and ulceration in the acute phase and should be suspected in patients who are immunocompromised or from regions where these infections are common. In the chronic phase the bladder wall may calcify; wall calcification is a characteristic finding of chronic schistosomiasis. The diagnosis of chemotherapy cystitis and radiation cystitis should be clinically evident, but imaging may be used to determine severity and to assess complications.

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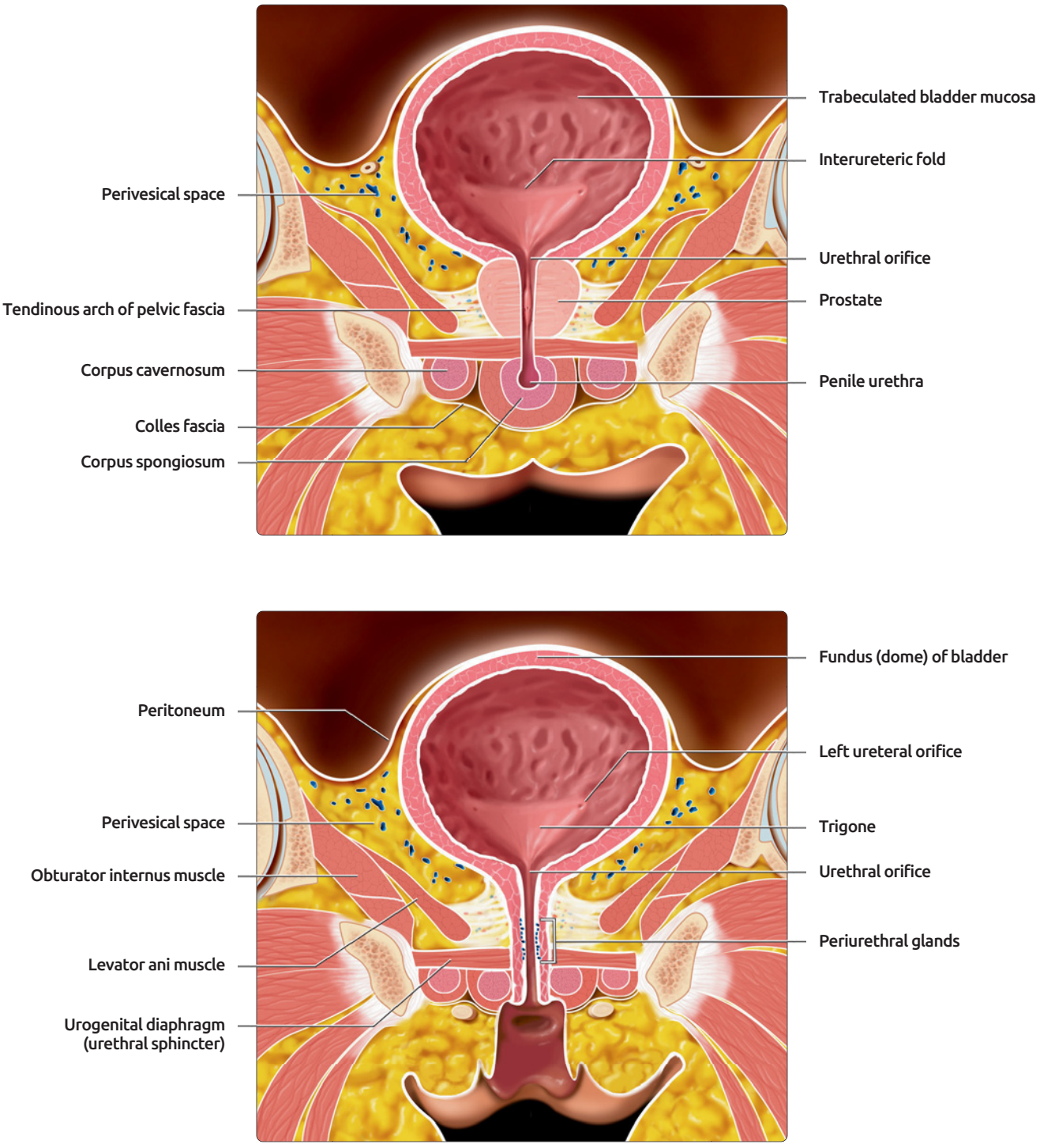


(Left) *Graphic of a developing male fetus shows the urachus ⇨ extending from the bladder dome to the umbilicus ⇨. Normally this involutes to form the median umbilical ligament. Failure to do so leads to a host of urachal anomalies.* **(Right)** *Posterior view of the anterior pelvic wall shows the median umbilical ligament ⇨ as a midline structure extending cephalad from the bladder dome ⇨. On either side are the medial umbilical folds ⇨, which cover the obliterated umbilical arteries.*



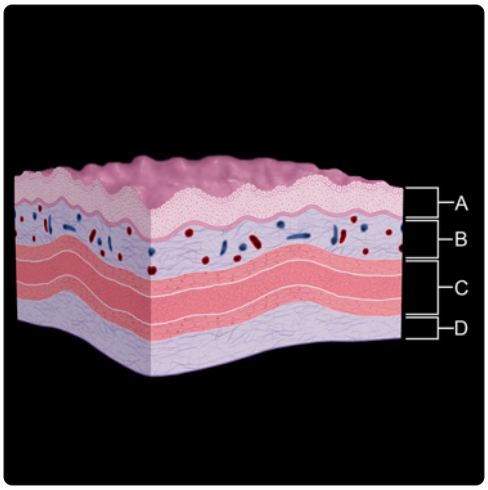


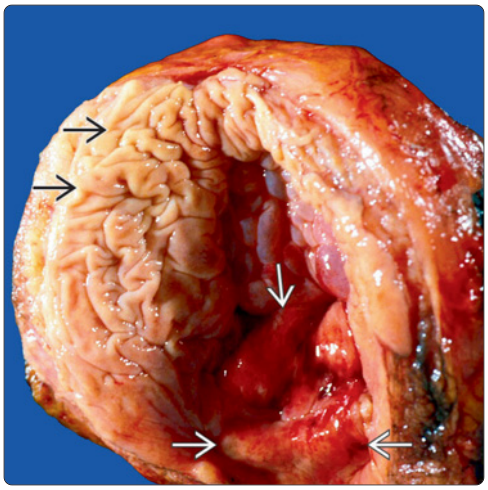
(Left) *Axial NECT shows gas within a small urachal diverticulum ⇨. This diverticulum is outlined by ascites on either side. This continued superiorly to the umbilicus as a fibrous cord, the median umbilical ligament.* **(Right)** *A more inferior NECT image in the same patient shows a triangle-shaped bladder due to the urachal diverticulum.*



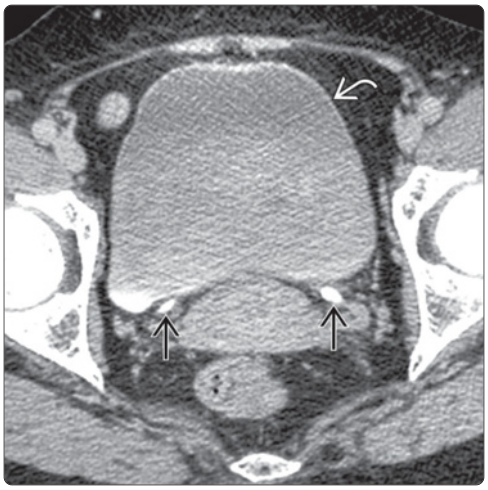
(Top) *A coronal section of the male bladder shows that it rests on the prostate. The bladder wall is muscular, strong, and very distensible. The ureters enter the bladder through an oblique anteromedial course that helps to prevent urinary reflux into the ureters.* **(Bottom)** *A coronal section of the female bladder shows that it rests almost directly on the muscular floor of the pelvis. The dome of the bladder is covered with peritoneum. The trigone is a distinct triangular structure at the base of the bladder whose apices are formed by the ureteral and urethral orifices. The bladder is surrounded by a layer of loose fat and connective tissue (perivesical space) that communicates with the retroperitoneum.*

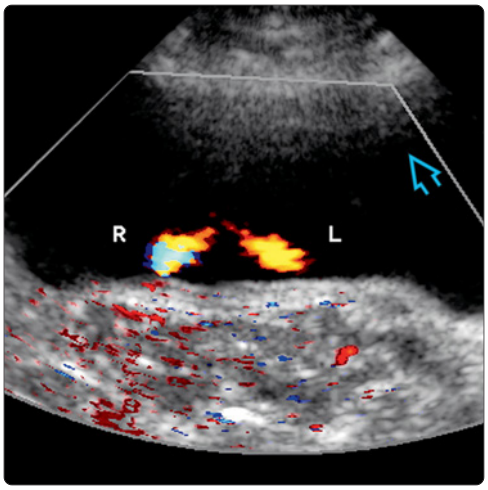
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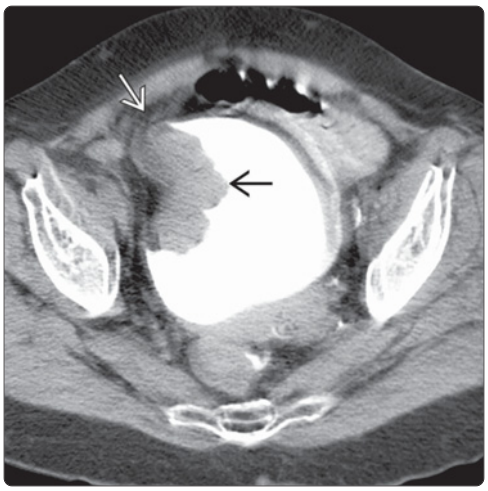


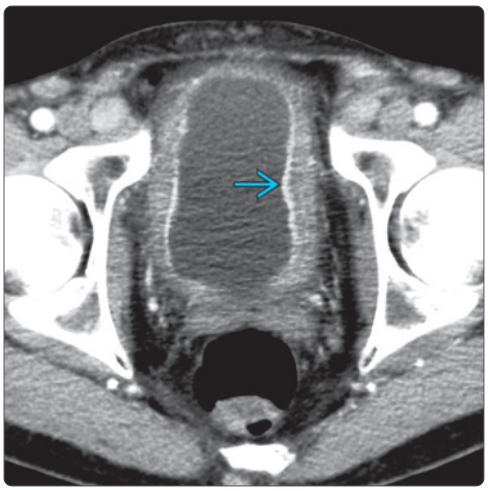
(Left) *Graphic of the 4 layers of the bladder wall shows the urothelium (A), lamina propria (B), muscularis propria (detrusor muscle) (C), and adventitia (D). A 5th layer, the peritoneum, is present only over the dome of the bladder.* **(Right)** *Gross specimen shows the opened bladder. Note the redundancy of the normal urothelium ⇨ in its collapsed state. A hemorrhagic, fungating urothelial tumor (transitional cell carcinoma) ➡ is seen at the bladder base protruding into the lumen.*





(Left) *Axial CECT of a well-distended bladder shows the ureters ⇨ entering the bladder. This marks the level of the bladder trigone. Note the very uniform, thin bladder wall ➡.* **(Right)** *Transverse color Doppler ultrasound shows bilateral ureteral jets indicating patency of the ureters. Note reverberation artifact ⇨ off the anterior bladder wall. This is a major pitfall when using ultrasound to evaluate the bladder.*





(Left) *Axial CT cystogram show an irregular mucosal mass ⇨, which was a transitional cell carcinoma that had subtle invasion ➡ into the adjacent perivesical fat (T3b lesion). Focal mucosal masses are predominately malignant.* **(Right)** *Axial CECT shows diffuse bladder wall thickening with mucosal hyperenhancement ⇨. This was chemotherapy-related cystitis from cyclophosphamide. Diffuse bladder wall thickening is more commonly benign but can be seen with some malignancies.*

Urachal Anomalies

KEY FACTS

TERMINOLOGY

- **Urachal anomalies**: Spectrum of anomalies due to incomplete obliteration of embryonic connection between bladder dome & allantoic duct
- **Patent urachus**: Entire urachal channel fails to close
- **Urachal cyst**: Umbilical & bladder openings close; channel between remains open & fluid filled
- **Urachal sinus**: Dilatation of urachus at umbilical end; no communication with bladder
- **Urachal diverticulum** (a.k.a. urachal remnant): Dilatation of urachus at vesical end; no communication with umbilicus
- Vestigial obliterated urachus (median umbilical ligament)

IMAGING

- Urachal diverticulum
  - Midline cystic lesion at anterosuperior aspect of bladder
- Urachal cyst
  - Midline cyst above bladder dome
  - May have rim calcification
- Urachal carcinoma
  - Midline supravesical soft tissue mass
  - Calcification in 70%

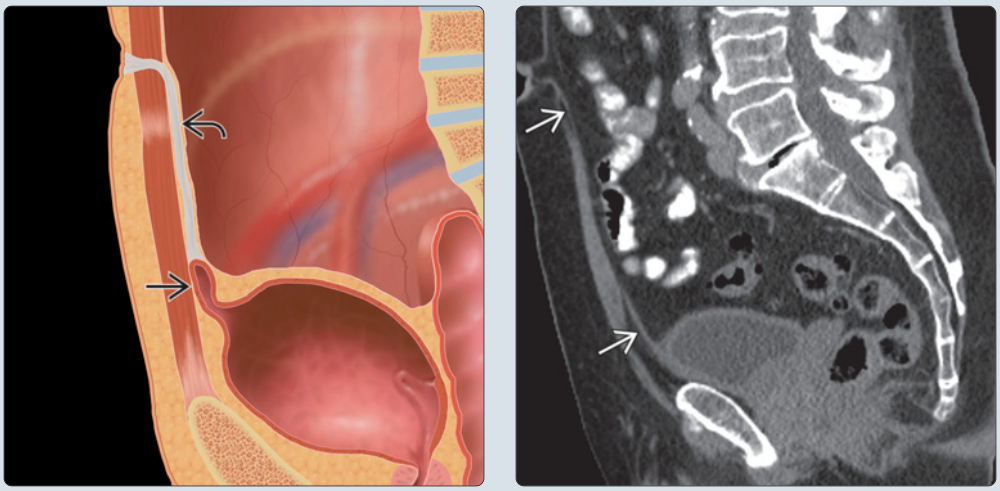
TOP DIFFERENTIAL DIAGNOSES

- Cystic lesions near bladder/umbilicus
  - Vitelline cyst: Failure in complete obliteration of omphalomesenteric duct
  - Mesenteric cyst
  - Abdominal abscess
- Bladder tumor
  - Primary bladder carcinoma at dome
  - Metastasis to bladder

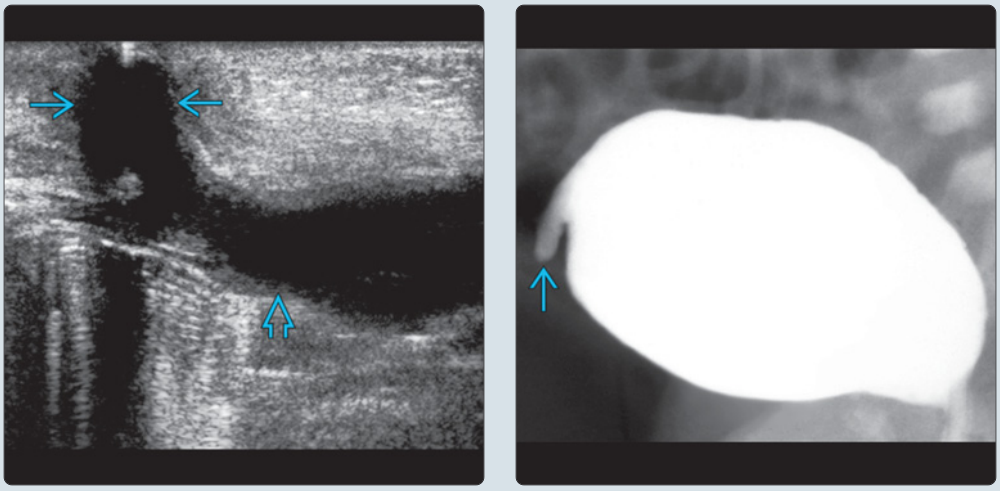
CLINICAL ISSUES

- Complications
  - Stone formation and infection in urachal diverticulum
  - Adenocarcinoma in urachal remnant
    - Poor prognosis related to late presentation

(Left) *Sagittal graphic shows a tubular channel extending from the dome of the bladder in the midline, which represents a urachal diverticulum. The normal obliterated urachus (median umbilical ligament) extends to the umbilicus.* **(Right)** *Sagittal NECT through the midline shows a normal-appearing obliterated urachus, also known as the median umbilical ligament.*



(Left) *Midline sagittal ultrasound through the urinary bladder shows a tubular anechoic channel extending from the bladder dome, which represents urachal diverticulum.* **(Right)** *Lateral view from retrograde cystogram shows a tubular structure arising from the bladder dome and extending toward the umbilicus, which represents a urachal diverticulum. (Courtesy of G. Friedland, MD.)*



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TERMINOLOGY

Definitions

- Spectrum of anomalies due to incomplete obliteration of embryonic connection between bladder dome and allantoic duct
  - **Patent urachus**: Entire urachal channel fails to close
  - **Urachal cyst**: Umbilical and bladder openings close; channel in between remains open and fluid filled
  - **Urachal sinus**: Dilatation of urachus at umbilical end; no communication with bladder
  - **Urachal diverticulum** (a.k.a. urachal remnant): Dilatation of urachus at vesical end; no communication with umbilicus
- Vestigial obliterated urachus, a.k.a. median umbilical ligament

IMAGING

General Features

- Best diagnostic clue
  - Midline cyst or mass between bladder and umbilicus
- Location
  - Between parietal peritoneum and fascia transversalis (space of Retzius)

Radiographic Findings

- Radiography
  - Urachal carcinoma: Stippled, granular, or curvilinear calcifications above bladder outline

Fluoroscopic Findings

- Sinogram: Tract extending inferiorly from umbilicus (sinus)
- Cystogram: Tubular channel arising from bladder dome (diverticulum)

CT Findings

- CECT
  - Urachal diverticulum: Midline cystic lesion at anterosuperior aspect of bladder communicating with bladder
  - Urachal cyst: Midline cyst above bladder dome
    - May have rim calcification
    - Low-density contents due to mucin production
    - No soft tissue component unless complicated
  - Urachal carcinoma: Midline supravesical soft tissue mass; calcification in 70%

Ultrasonographic Findings

- Grayscale ultrasound
  - Urachal diverticulum: Tubular anechoic extension from bladder dome
  - Urachal cyst: Midline cyst above bladder dome
  - Complicated urachal cyst: Mixed echogenicity

Imaging Recommendations

- Best imaging tool
  - Multiplanar CT or MR; cystogram

DIFFERENTIAL DIAGNOSIS

Cystic Lesions Near Bladder/Umbilicus

- Vitelline cyst: Failure in complete obliteration of omphalomesenteric duct
- Mesenteric cyst
- Abdominal abscess

Bladder Tumor

- Primary bladder carcinoma at dome
- Metastasis to bladder

PATHOLOGY

Microscopic Features

- Urachal anomalies: Mucosal lining of transitional epithelium
- Urachal carcinoma: 85-90% are adenocarcinomas resulting from metaplasia of transitional epithelium

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Patent urachus: Urine leakage from umbilicus
  - Complicated urachal cyst: Suprapubic pain, tenderness, fever
  - Urachal carcinoma: Clinically silent until bladder dome invaded; then hematuria &/or mucoid discharge

Demographics

- Age
  - Patent urachus: Diagnosed at birth
  - Urachal carcinoma: 40-70 years
- Gender
  - M:F = 3:1
- Epidemiology
  - Prevalence of urachal anomalies: ~ 1%
    - Patent urachus has reported prevalence 0.25-15 per 10,000
  - Urachal carcinoma: 0.2-0.3% of all bladder cancers; 20-40% of primary bladder adenocarcinomas

Natural History & Prognosis

- Most cases with urachal diverticulum and cyst are asymptomatic
- Complications
  - Stone formation and infection in urachal diverticulum
  - Adenocarcinoma in urachal remnant
    - Urachal carcinoma: 5-year survival ≤ 15%
      - Poor prognosis due to late presentation

Treatment

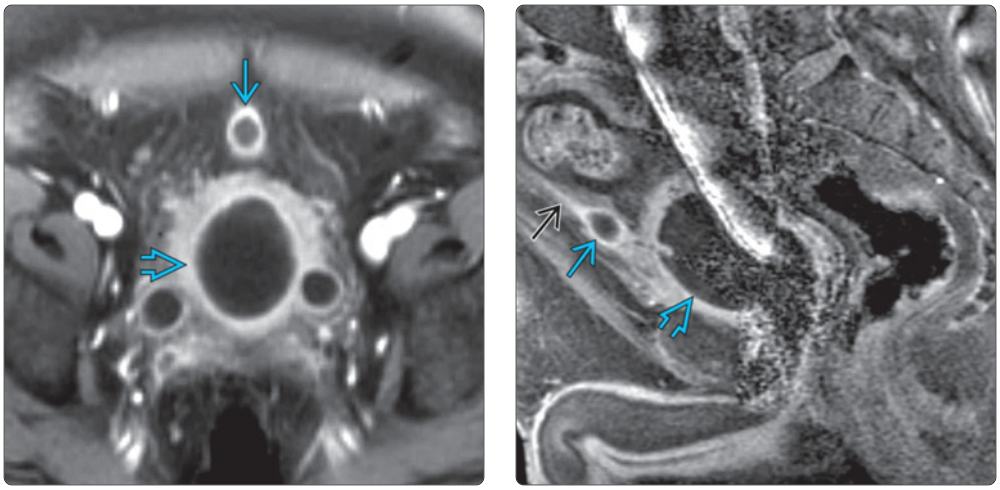
- Options, risks, complications
  - Excision of urachus and accompanying abnormality

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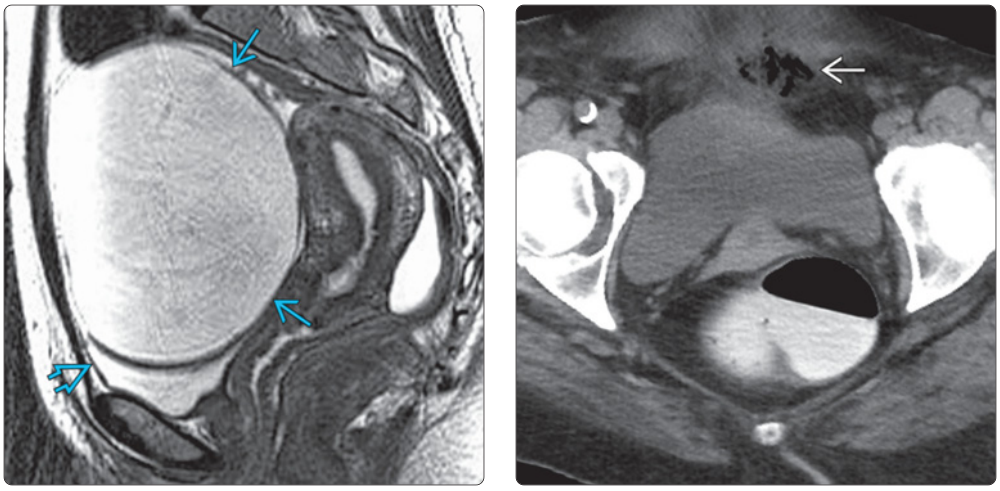
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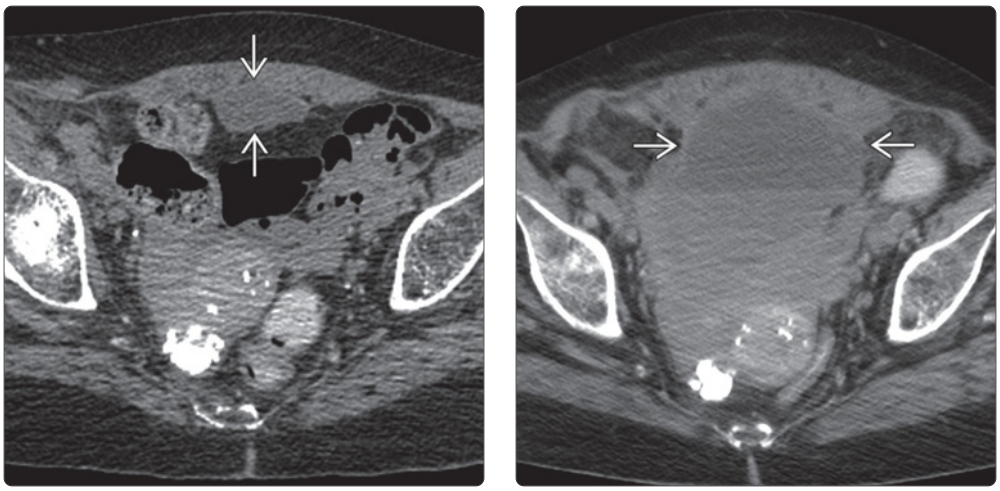
(Left) *Axial contrast-enhanced T1WI MR shows a small cystic structure, anterior and superior to the bladder dome.* **(Right)** *Sagittal contrast-enhanced T1WI MR (same patient) again shows the cystic structure, superior to the bladder dome. Note the median umbilical ligament connecting the bladder dome to the umbilicus. A urachal cyst results when the bladder and umbilical ends of the fetal urachus obliterate while the central part of the tract remains open, as in this case.*



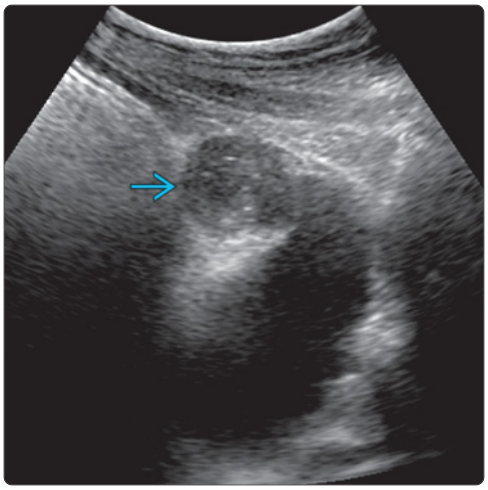
(Left) *Sagittal T2WI MR shows a large urachal cyst depressing the dome of the urinary bladder and displacing the uterus posteriorly. Urachal cysts have no opening to the bladder or the umbilicus, which distinguishes them from urachal diverticula.* **(Right)** *Axial NECT in a 72-year-old female presenting with abdominal pain and sepsis, demonstrates a gas-containing collection anterior to the bladder dome. The patient underwent exploratory laparotomy, which showed infected urachal cyst.*

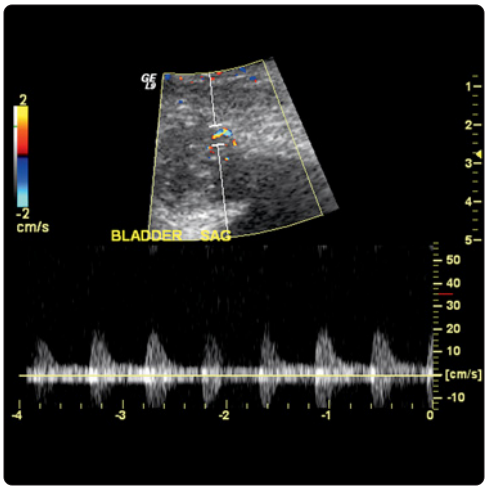


(Left) *Axial NECT through the pelvis shows an incidental cystic structure along the course of medial umbilical ligament, compatible with urachal cyst.* **(Right)** *Repeat NECT through same level of the same patient (presenting with urosepsis and pain 1 year later) shows marked enlargement of the urachal cyst. Complex cyst fluid and extensive surrounding inflammatory changes indicate superinfection. The patient underwent percutaneous drainage of the abscess and subsequent partial cystectomy.*

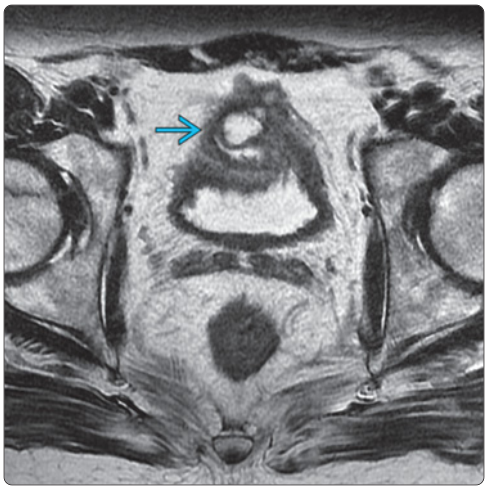


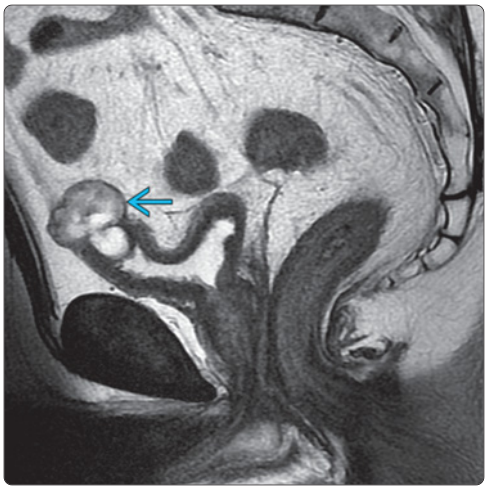
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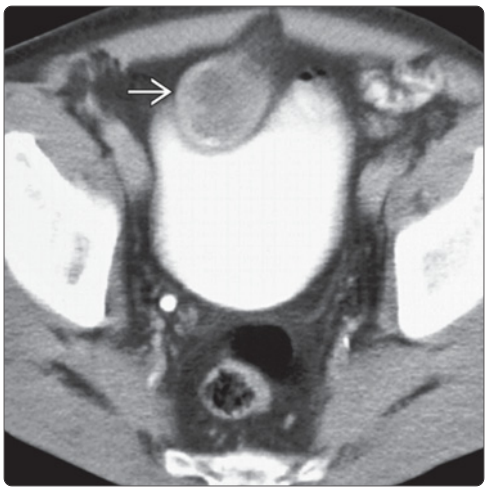


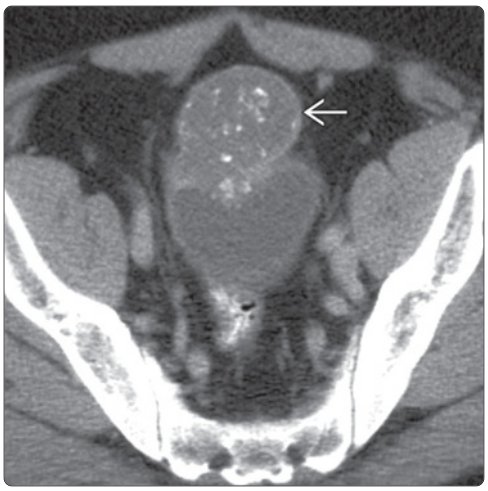
(Left) *Midline sagittal ultrasound of the bladder shows an incidental complex mass ⇨ superior to the dome.* **(Right)** *Spectral Doppler evaluation of the mass shows vascularity of the lesion.*





(Left) *Axial T2 SSFSE MR shows a complex solid/cystic mass ⇨ attached to the bladder dome.* **(Right)** *Midline sagittal T2 SSFSE MR in the same patient again demonstrates the complex solid/cystic mass ⇨ attached to the bladder dome. The patient could not receive IV contrast due to renal failure. Given its morphology, and vascularity on US, this lesion was highly concerning for a carcinoma within the urachal remnant. Patient underwent partial cystectomy and pathology was consistent with adenocarcinoma.*





(Left) *Axial CECT shows a soft tissue mass ➡ adjacent to, but separate from, the bladder. The mass has an enhancing periphery and a cystic core. This is a urachal carcinoma occurring within a urachal remnant.* **(Right)** *Axial CECT shows an exophytic mass ➡ with coarse internal calcification invading the bladder dome, representing urachal adenocarcinoma.*

Cystitis

KEY FACTS

IMAGING

- Acute: Bladder wall thickening, ± hypodense wall (edema), ± perivesical inflammatory changes
- Chronic: Contracted, irregular, thick-walled bladder
- Emphysematous: Gas in bladder wall and lumen

TOP DIFFERENTIAL DIAGNOSES

- Cystitis secondary to adjacent inflammation (such as diverticulitis or salpingitis)
- Bladder underdistention
- Bladder outlet obstruction
- Neurogenic bladder
- Bladder carcinoma

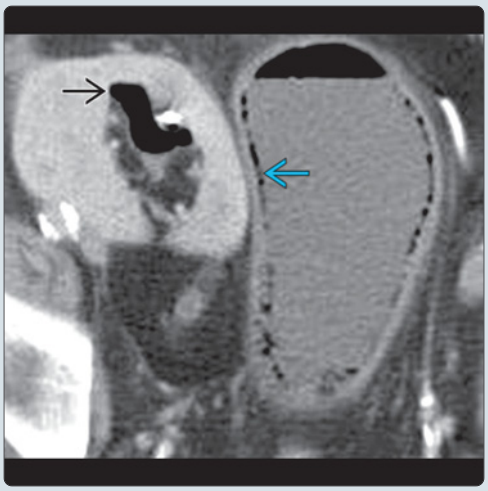
PATHOLOGY

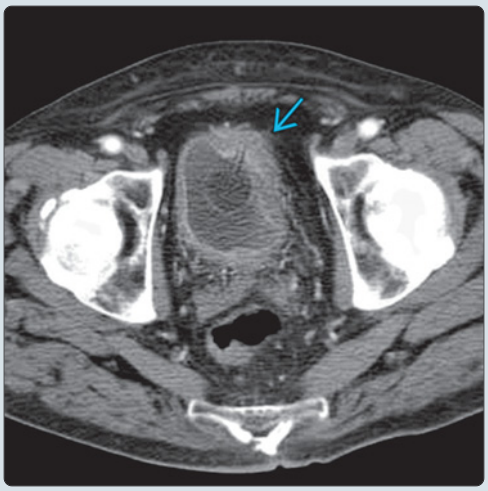
- **Infectious cystitis**
  - Bacterial
  - Schistosomiasis: Inflammatory response to ova deposited in bladder submucosa
  - Emphysematous: Infection with *Escherichia coli*, *Enterobacter aerogenes*, or *Candida* infection; seen in immunosuppressed and diabetic patients
- **Noninfectious cystitis**
  - Mechanical: Local irritation from indwelling catheter, stone, foreign body, etc.
  - Drug related: Cyclophosphamide (hemorrhagic cystitis)
  - Radiation induced
  - Idiopathic interstitial cystitis: Pancystitis causing severe urgency and frequency

CLINICAL ISSUES

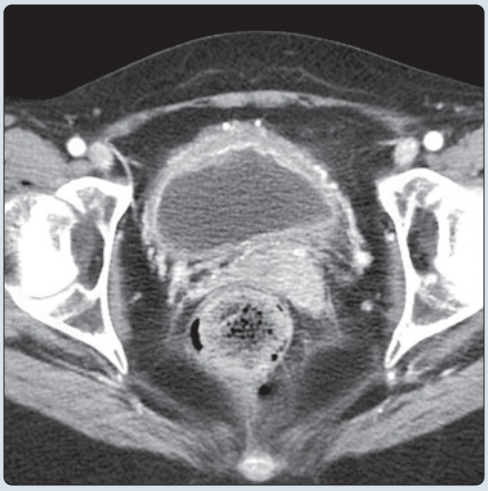
- Less common in men due to longer length of urethra and antibacterial properties of prostatic fluid
- **Risk factors**: Sexual intercourse, spermicide use, urogenital anomalies, bladder outlet obstruction, foreign bodies, history of prior urinary tract infections

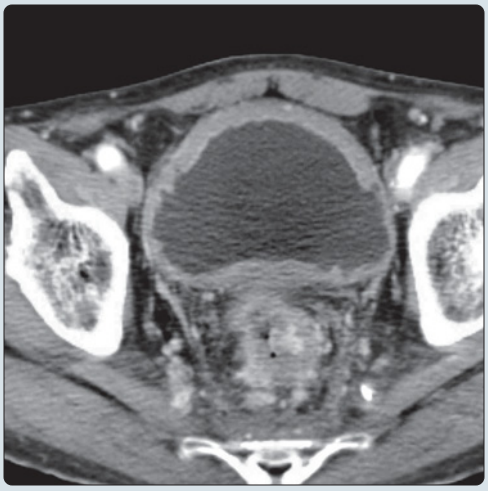
(Left) *Axial CECT shows gas within the bladder lumen and wall ⇨, as well as in the allograft pelvicalyceal system ⇨ compatible with emphysematous cystitis/pyelitis. There was no history of recent instrumentation to account for intravesical/intrarenal gas.* **(Right)** *Axial CECT shows eccentric bladder wall thickening with perivesical inflammatory changes ⇨ compatible with cystitis, likely due to chronic irritation by indwelling catheter. Wall hyperemia suggests cystitis, rather than a neoplasm.*





(Left) *Axial CT shows marked bladder wall thickening and engorged perivesical blood vessels. The patient was receiving cyclophosphamide, which is excreted in the urine, and these changes were felt to be due to chemical cystitis. Cancer patients are at increased risk of infectious cystitis; urinalysis is helpful in differentiating these 2 entities.* **(Right)** *Axial CT in a patient with history of recent external beam radiation for rectal carcinoma shows bladder wall thickening representing acute radiation-induced cystitis.*





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TERMINOLOGY

Definitions

- Inflammation of urinary bladder

IMAGING

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic, thickened bladder wall

CT Findings

- CECT
  - Acute: Diffuse bladder wall thickening, ± hypodensity of bladder wall (edema), ± hyperemia and infiltration of perivesical fat
  - Chronic: Contracted, irregular, thick-walled bladder
  - **Emphysematous cystitis**
    - Gas in bladder wall and lumen

MR Findings

- Thick-walled, hyperemic bladder ± perivesical inflammatory changes

DIFFERENTIAL DIAGNOSIS

Bladder Underdistention

- Bladder underdistention can mimic wall thickening

Secondary Inflammation From Extrinsic Source

- Diverticulitis
- Salpingitis

Bladder Outlet Obstruction

- Bladder wall thickening and trabeculation secondary to long-standing obstruction
- Usually secondary to prostate enlargement; other causes include urethral strictures and urethral valves
- Usually have increased postvoid residue

Neurogenic Bladder

- Spastic-type neurogenic bladder results in marked bladder wall thickening and trabeculation
- Usually seen in patients with spinal cord injury and multiple sclerosis

Bladder Carcinoma

- Usually causes eccentric focal wall thickening ± intraluminal papillary projection
- Soft tissue attenuation mucosal/submucosal tumor (rather than low attenuation of cystitis)

PATHOLOGY

General Features

- Etiology
  - **Infectious cystitis**
    - Bacterial
      - Transurethral contamination of bladder by perineal flora in sexually active women
      - Bladder outlet obstruction and stasis in men
    - Schistosomiasis: Inflammatory response to ova deposited in bladder submucosa
    - Tuberculosis: Descending infection from kidney
    - Emphysematous: Infection with *Escherichia coli*, *Enterobacter aerogenes*, or *Candida* infection; seen in immunosuppressed and diabetic patients
    - Alkaline encrustation: Infection by urea-splitting organism → alkaline urine + focal necrosis → dystrophic calcification
    - Others: Viral, fungal (*Candida*)
  - **Noninfectious cystitis**
    - Mechanical: Local irritation from indwelling catheter, stone, foreign body, etc.
    - Drug related: Cyclophosphamide (hemorrhagic cystitis)
    - Radiation induced
    - Idiopathic interstitial cystitis: pancystitis causing severe urgency and frequency

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Dysuria, frequency, urgency; hematuria, pyuria

Demographics

- Gender
  - M:F = 1:10 (for bacterial cystitis)
  - Less common in men due to longer length of urethra and antibacterial properties of prostatic fluid
- Epidemiology
  - Extremely common; estimated incidence of 0.5-0.7 per person/year in sexually active women

Natural History & Prognosis

- **Risk factors**: Sexual intercourse, spermicide use, urogenital anomalies, bladder outlet obstruction, foreign bodies, history of prior urinary tract infections
- Acute cystitis (including emphysematous type) responds well to antibiotics
- **Complications of chronic cystitis**
  - Hyperplastic uroepithelial cell clusters (von Brunn nests) form in bladder submucosa
    - Fluid accumulation → pseudocyst formation (**cystitis cystica**)
    - Transformation into glands (**cystitis glandularis**)
  - Squamous metaplasia: Transformation of urothelium into keratin-producing squamous cells
    - May see white patches (**leukoplakia**) on foci of squamous metaplasia
  - Chronic infection + deficient macrophage function → granulomatous inflammation (**malakoplakia**)
- Schistosomiasis: Increased risk of bladder carcinoma (especially squamous type)

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Bladder Schistosomiasis

KEY FACTS

TERMINOLOGY

- Bilharziasis, parasitic infection
- Infection of urinary system by parasite *Schistosoma haematobium*

IMAGING

- Imaging findings mirror pathologic course
  - Acute phase: Nodular bladder wall thickening
  - Chronic phase: Contracted, fibrotic, thick-walled bladder with calcifications
- Calcifications of bladder wall
  - ± calcification of ureter (34%), seminal vesicle (late)
  - Calcification best seen on NECT
- Mucosal irregularity, inflammatory pseudopolyps of bladder
- Ureteritis cystica, cystitis cystica, distal ureteric stricture

TOP DIFFERENTIAL DIAGNOSES

- Bladder calculi
- Bladder carcinoma
- Cystitis
- Emphysematous cystitis

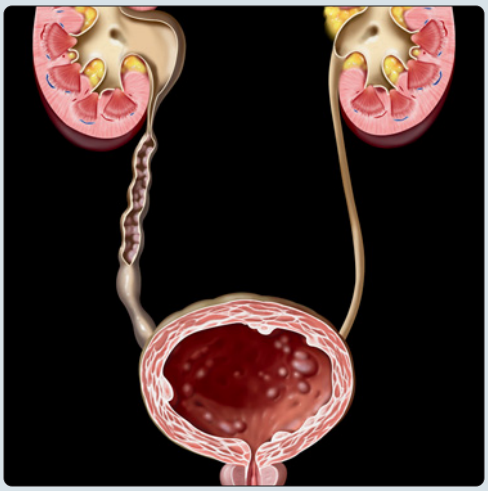
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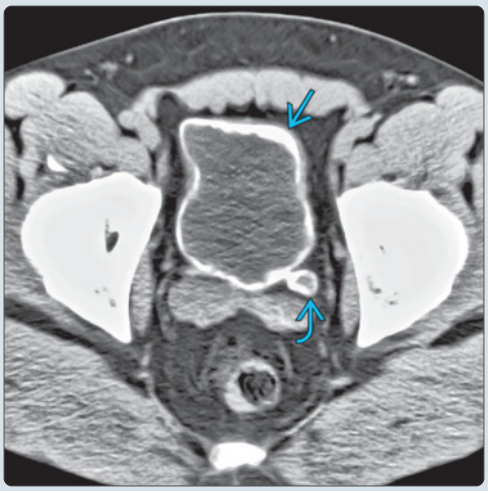
- Clinical manifestations are attributed to host immune response and hypersensitivity reactions
  - Cell-mediated (type IV) immune response: Dominant pathogenetic pathway by forming granulomata (inflammatory and fibrotic phases)
  - Immune-complex-mediated (type III) response: Plays role in glomerulonephritis

CLINICAL ISSUES

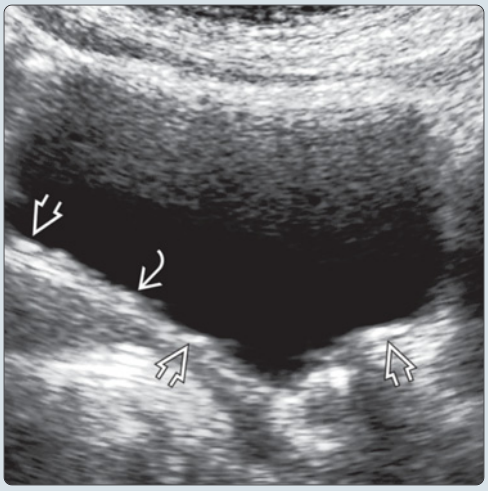
- Endemic in Middle East, India, Africa, Central America, and South America
- Confirm diagnosis by identifying ova of parasite in urine
- Squamous cell carcinoma of bladder is late complication

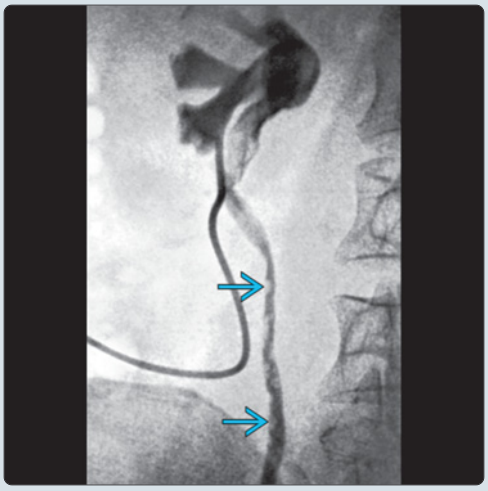
(Left) *Schematic drawing shows a markedly thickened urinary bladder wall with mucosal irregularity and mural calcifications. Changes of ureteritis cystica and cystitis cystica are also seen in the bladder and in the dilated right ureter.* **(Right)** *Axial NECT shows profound bladder wall calcification, which also extends to involve the left ureter. Calcifications are seen in the chronic phase of the infection and represent calcified granulomata.*





(Left) *Longitudinal ultrasound shows multiple echogenic foci of calcifications and mucosal irregularity of the posterior bladder wall. Bladder volume is reduced.* **(Right)** *Frontal projection from antegrade nephrostogram shows the typical appearance of ureteritis cystica caused by infection with Schistosoma haematobium.*





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TERMINOLOGY

Synonyms

- Bilharziasis, parasitic infection

Definitions

- Infection of urinary system by *Schistosoma haematobium*

IMAGING

General Features

- Best diagnostic clue
  - Curvilinear bladder wall calcification in patient from endemic area
- Imaging findings mirror pathologic course
  - Acute phase: Nodular bladder wall thickening
  - Chronic phase: Contracted, fibrotic, thick-walled bladder with calcifications

Radiographic Findings

- Calcifications of bladder wall
  - ± calcification of ureter (34%), seminal vesicle (late)

Fluoroscopic Findings

- Mucosal irregularity, inflammatory pseudopolyps of bladder
- Ureteritis cystica, cystitis cystica, distal ureteric stricture
- Vesicoureteric reflux

CT Findings

- NECT
  - Best delineation of extent of bladder calcification, mural pathology, ureteral involvement

Ultrasonographic Findings

- Noncompliant, thick-walled, small-capacity bladder with significant postvoid residual
- Echogenic calcification within bladder wall
- Hydronephrosis and hydroureter due to distal ureteric stricture

DIFFERENTIAL DIAGNOSIS

Bladder Calculi

- Mobile calcification within dependent portion of bladder lumen
- Majority of cases associated with bladder outlet obstruction (BPH)

Bladder Urothelial Carcinoma

- Irregular wall thickening
- Associated lymphadenopathy ± multiple sites of involvement

Urachal Carcinoma

- Intramural exophytic mass arising from dome of bladder extending toward umbilicus
- May have focal calcification

Cystitis

- Diffuse or focal thickening of bladder wall; no wall calcification
- Clinical, not imaging, distinction

Emphysematous Cystitis

- Acute infection by gas-forming organism
- Echogenic foci within bladder wall can mimic schistosomiasis on US; look for ring-down shadowing
- Gas in bladder wall or lumen easily detected on CT

PATHOLOGY

General Features

- **Pathogenesis**
  - Clinical manifestations are attributed to host immune response and hypersensitivity reactions
    - Cell-mediated (type IV) immune response: Dominant pathogenetic pathway by forming granulomata (inflammatory and fibrotic phases)
    - Immune-complex-mediated (type III) response: Plays role in glomerulonephritis

Microscopic Features

- Encapsulated eggs cause inflammatory granulomatous reaction
- Fibrosis traps ova in muscular layer of bladder wall where ova die and calcify

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Frequency, urgency, dysuria, hematuria
- Clinical profile
  - Confirm diagnosis by identifying ova of parasite in urine

Demographics

- Epidemiology
  - Endemic in Middle East, India, Africa, Central America, and South America

Natural History & Prognosis

- Causal relationship between *S. haematobium* and bladder carcinoma has been shown in animal studies
- Tumor usually remains localized for prolonged period because of occlusion of lymphatic channels by fibrotic changes
- Squamous cell carcinoma is dominant type (~ 70%)
- Reported incidence of bladder cancer as high as 4.5%

Treatment

- Anthelminthic therapy (praziquantel)
- Not beneficial for reversing delayed fibrosis

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Bladder Calculi

KEY FACTS

TERMINOLOGY

- Concretions of mineral salts within bladder lumen
- Classified as migrant, primary (idiopathic), and secondary
- Obstruction accounts for 70% of adult bladder stones

IMAGING

- Round, oval, spiculated, laminated, faceted
- All bladder calculi are radiopaque on CT

TOP DIFFERENTIAL DIAGNOSES

- Other bladder calcifications
  - Urachal carcinoma: Punctate or coarse calcification in mass at bladder dome
  - Schistosomiasis: Calcification in bladder wall
- Other pelvic calcifications
  - Prostate calcification, calcified fibroids
- Filling defect in bladder
  - Blood clot, fungus ball, ureterocele, tumor

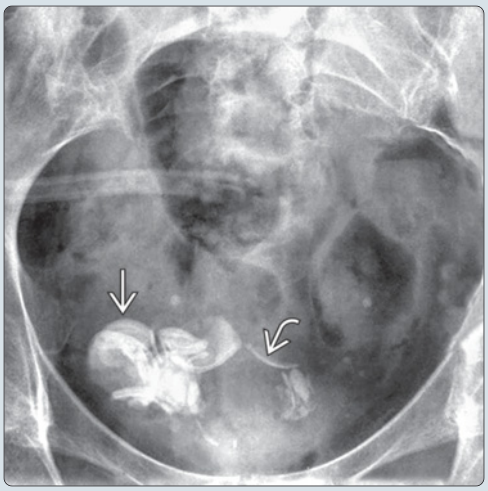
PATHOLOGY

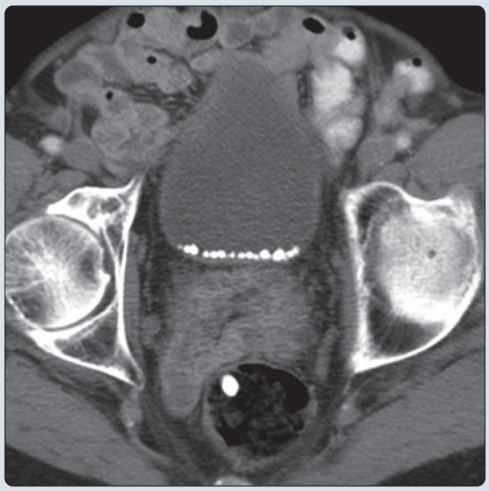
- **Stasis**: Bladder outlet obstruction, neurogenic bladder, bladder diverticula
- **Infection**, especially with *Proteus mirabilis*
- **Foreign bodies**: Nidus for crystal growth
- **Bladder augmentation**: Metabolic derangement and stasis
- Most are mixture of calcium oxalate and calcium phosphate

CLINICAL ISSUES

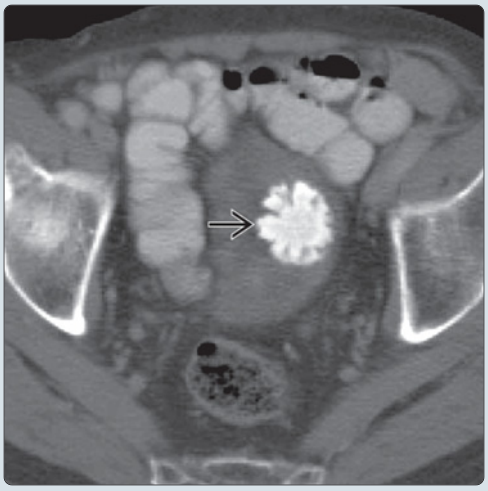
- Most are asymptomatic
  - Some have suprapubic pain, microhematuria
- Clinical scenarios
  - Elderly man with bladder outlet obstruction
  - Spinal cord injury patient with indwelling Foley catheter
  - Bladder augmentation with ileal or colonic segments
- Complication: Malignant bladder tumors in patients with stones from indwelling Foley catheters

(Left) *Plain film frontal radiograph in a 37-year-old woman with paraplegia and microhematuria shows multiple calculi within the bladder. Some have a peculiar curvilinear shape, suggesting that they may have formed around foreign bodies* **(Right)** *Axial CECT through the urinary bladder shows multiple rounded calcified stones layering in the dependent portion of the bladder. The patient had longstanding urinary obstruction in the setting of benign prostatic hyperplasia (BPH).*





(Left) *Axial CECT shows the typical appearance of a "jackstone" bladder calculus, usually composed of calcium oxalate dihydrate. They usually form within the bladder, rather than the upper urinary system.* **(Right)** *Axial T1WI MR through the bladder shows innumerable round signal void foci in the dependent portion of the bladder, representing calculi in a man with marked prostatic enlargement. Bladder calculi are hypointense on T1W and T2W images due to the diamagnetic property of calcium salts.*





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TERMINOLOGY

Synonyms

- Bladder stones, vesical calculi, cystolithiasis

Definitions

- Concretions of mineral salts within bladder lumen
- Classified as migrant (migrated from upper urinary tract), primary (idiopathic), and secondary (in setting of stasis, infection, or nidus)

IMAGING

General Features

- Best diagnostic clue
  - Smooth round calcification in bladder
- Location
  - Bladder lumen: Usually midline with patient supine
    - Eccentric if within bladder augmentation or diverticulum
- Morphology
  - Round, oval, spiculated, laminated, faceted
    - Stones with multiple spicules: "Jackstones"
    - Stones with lobulated margins: "Mulberry stones"

Radiographic Findings

- Radiography
  - Solitary or multiple calcifications in low pelvis
  - Most are radiopaque, but opacity is variable
- IVP
  - Filling defect or radiopacity, depending on relative density of stone vs. opacified urine

CT Findings

- NECT
  - All bladder calculi are radiopaque on CT

MR Findings

- Signal void(s) in bladder (on all pulse sequences)
  - Due to diamagnetic property of stones

Ultrasonographic Findings

- Grayscale ultrasound
  - Mobile echogenic foci in bladder with posterior acoustic shadowing

Imaging Recommendations

- Best imaging tool
  - CT

DIFFERENTIAL DIAGNOSIS

Other Bladder Calcifications

- Urachal carcinoma: Punctate or coarse calcification in mass at bladder dome
- Schistosomiasis (calcification in bladder wall)

Other Pelvic Calcifications

- Prostate calcification, calcified fibroids

Filling Defect in Bladder

- Blood clot, fungus ball, ureterocele, bladder neoplasm

PATHOLOGY

General Features

- Etiology
  - Stasis: Bladder outlet obstruction, neurogenic bladder, bladder diverticula
  - Infection, especially with *Proteus mirabilis*
  - Foreign bodies: Nidus for crystal growth
    - Renal calculus passing into bladder
    - Suture material, stents, migrated intrauterine devices, or other gynecologic appliances
    - Pubic hairs introduced by catheterization
  - Bladder augmentation: Metabolic derangement and stasis
  - Nutritional deficiencies in children in developing countries
- Most are a mixture of calcium oxalate and calcium phosphate
- Infection stones: Magnesium ammonium phosphate (struvite)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most are asymptomatic
  - Other signs/symptoms
    - Suprapubic pain, microhematuria
- Clinical profile
  - Spinal cord injury patient with indwelling Foley catheter
  - Elderly man with bladder outlet obstruction
  - Older woman with pelvic prolapse
  - Bladder augmentation with ileal or colonic segments

Demographics

- Gender
  - M > F
- Epidemiology
  - 5% of all urinary tract calculi in Western nations
  - Idiopathic: Endemic in underdeveloped areas, especially in children

Natural History & Prognosis

- Complication: Malignant bladder tumors in patients with stones from indwelling Foley catheters

Treatment

- Options, risks, complications
  - Prevention and eradication of underlying cause
  - Cystoscopic fragmentation and removal
  - Open cystolithotomy for large, hard stones

DIAGNOSTIC CHECKLIST

Consider

- Obstruction accounts for 70% of adult bladder stones

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Bladder Diverticulum

KEY FACTS

TERMINOLOGY

- Sac formed by herniation of bladder mucosa and submucosa through muscular wall
- **Hutch diverticulum**: Congenital form due to weakness in detrusor muscle anterolateral to ureteral orifice

IMAGING

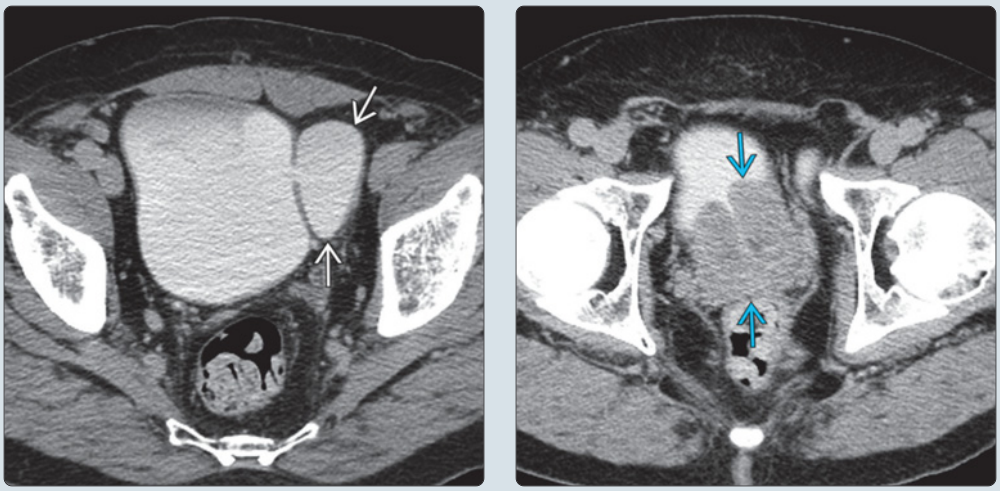
- Perivesical cystic mass with connection to bladder
- Usually multiple; smooth wall
- May contain debris, calculi, or tumor
- US: Anechoic outpouching from bladder with visible neck
  - Change in size post urination and color jet connecting to bladder are useful signs for distinguishing diverticulum from other masses
  - Check for emptying on postvoid imaging as an indicator of degree of stasis
- VCUG: Preferred modality in young children to look for associated VUR

TOP DIFFERENTIAL DIAGNOSES

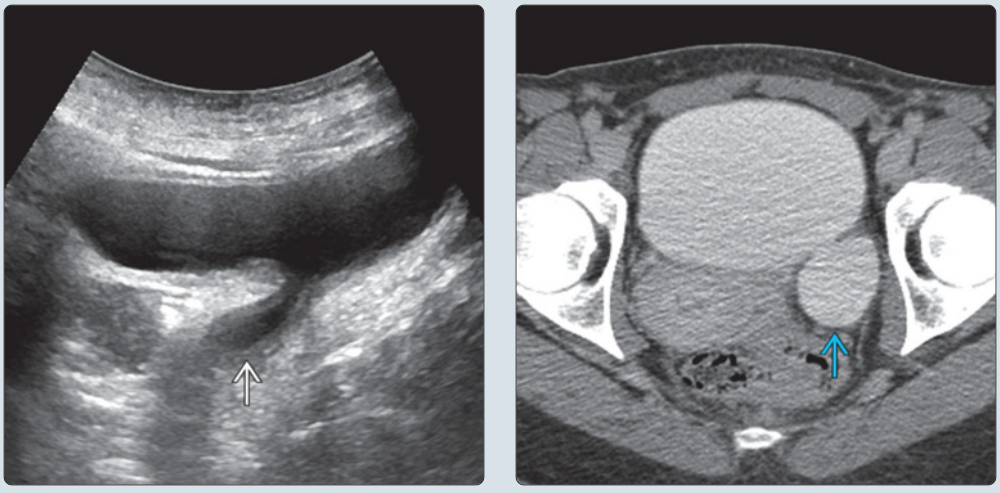
- Abdominal abscess
  - Multiseptate tender mass in febrile patient
- Postoperative lymphocele
  - Adjacent to extraperitoneal vessels; does not fill with contrast; multiseptate
- Everted ureterocele
- Ovarian cystic lesions

PATHOLOGY

- Predisposing conditions: Bladder outlet obstruction, neurogenic bladder, congenital weakness of detrusor muscle
- Lack of muscular wall results in stasis, which has pivotal role in development of complications
- **Complications**: Infection, stone, neoplasm
  - Transitional cell carcinoma is most prevalent type
  - Lack of muscularis propria leads to more rapid local spread and poor prognosis



(Left) *Axial CT urography image in 78-year-old male with history of hematuria shows a large diverticulum ⇒, with a relatively narrow neck and with smooth surface, along the left lateral aspect of bladder.* **(Right)** *Axial CT through a more inferior plane in the same patient demonstrates marked enlargement of the transitional zone of the prostate gland ⇨ suggesting urinary outlet obstruction as the etiology of diverticulum.*



(Left) *Sagittal US in 50-year-old female with history of hematuria shows a moderate-size diverticulum ⇒ along the posterior wall of the bladder.* **(Right)** *The patient underwent CT urography for further work-up of hematuria. CT image better shows the diverticulum ⇨ along the posterolateral wall of bladder, in close proximity to UVJ. The patient had no known cause of bladder obstruction or neurogenic bladder. Although rare, this finding can represent a congenital (Hutch) diverticulum.*

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TERMINOLOGY

Synonyms

- Vesicular diverticulum
- Bladder pseudodiverticulum

Definitions

- Sac formed by herniation of bladder mucosa and submucosa through muscular wall
- **Hutch diverticulum**: Congenital form secondary to weakness in detrusor muscle anterolateral to ureteral orifice

IMAGING

General Features

- Best diagnostic clue
  - Perivesical cystic mass with connection to bladder lumen
- Location
  - Can occur anywhere in bladder
  - Common locations are
    - Near ureterovesical junction (UVJ)
    - Bladder dome
      - Consider urachal remnant if solitary, midline, and extending along median umbilical ligament
- Size
  - Small to very large
    - Can exceed size of bladder
  - Size can vary depending on degree of bladder distension
- Morphology
  - Usually multiple
    - Congenital form is usually solitary
  - Smooth wall
  - Wide or narrow neck
  - May contain debris, calculi, or tumor

Radiographic Findings

- IVP
  - Medial deviation of ipsilateral ureter
  - Usually fills with contrast unless obstructed
  - Diverticulum may contain stones, debris, or tumor
- Cystogram
  - Oblique films may show configuration of diverticulum neck
- VCUG
  - Preferred modality in young children to look for associated vesicoureteral reflux (VUR)

CT Findings

- CECT
  - Water-attenuation outpouching from bladder
  - Usually fills with contrast on delayed urographic phase
    - Debris, stone, or tumor may be seen within diverticulum

MR Findings

- Homogeneous low signal on T1WI; high on T2WI
  - May see loss of signal on T2WI due to dephasing with motion of urine between diverticulum and bladder lumen

Ultrasonographic Findings

- Grayscale ultrasound
  - Anechoic outpouching from bladder with visible neck (connection)
  - Size changes post voiding
- Color Doppler
  - Color jet connecting to bladder is very useful in distinguishing diverticulum from other masses

Imaging Recommendations

- Best imaging tool
  - Ultrasound, cystogram
- Protocol advice
  - Delayed contrast-enhanced CT or MR
  - Check for degree of emptying on postvoid imaging

DIFFERENTIAL DIAGNOSIS

Abdominal Abscess

- Multiseptate tender mass in febrile patient
- Not communicating with bladder

Postoperative Lymphocele

- Adjacent to extraperitoneal vessels
- Does not fill with contrast
- Multiseptate

Everted Ureterocele

- Continuous with ureter

Ovarian Cystic Lesions

- No communication with bladder

PATHOLOGY

General Features

- Etiology
  - Acquired (most common type)
    - Secondary to bladder outlet obstruction
      - Common causes in adults: Prostatic enlargement, neurogenic bladder, large ureterocele, urethral stricture
      - Common causes in children: Posterior urethral valves, prune belly syndrome
  - Congenital, known as **Hutch diverticulum** (rare)
    - Congenital weakness in detrusor muscle anterolateral to ureteral orifice
- Associated abnormalities
  - Hutch diverticulum can be seen in association with Ehlers-Danlos syndrome
  - Significant subset of pediatric patients with bladder diverticulum have congenital syndromes
- **Complications**
  - Lack of muscular wall results in stasis, which has pivotal role in development of complications
    - Infection
    - Stone formation
    - Neoplasm
      - Incidence of neoplasm in bladder diverticulum has been reported in range of 0.8-13%
      - Most common type is transitional cell carcinoma

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- Incidence of squamous cell carcinoma arising in diverticulum (17%) is higher than in bladder
- Tumors in diverticula have poor prognosis; lack of muscularis propria leads to more rapid local spread

Microscopic Features

- Chronic inflammation
- Cystitis glandularis
- Mucosal denudation
- Squamous metaplasia

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Usually asymptomatic in adults
    - If symptomatic, urinary tract infection and hematuria are most common symptoms
  - Prevalence of clinical symptoms is higher in children
    - Urinary tract infection, hydronephrosis, hematuria, and nephrolithiasis are common symptoms
    - Hutch diverticulum is commonly associated with vesicoureteral reflux (VUR), due to altered UVJ anatomy
      - High-grade VUR results in hydronephrosis
- Clinical profile
  - Older man with benign prostatic hypertrophy
  - Spinal cord injury patient
  - Young boy with symptoms of recurrent urinary tract infection

Demographics

- Age
  - Mostly in older age; 6th and 7th decades
  - Mean age of presentation in children is 8 years
- Gender
  - M:F = 9:1
  - Hutch diverticulum occurs almost exclusively in boys
- Epidemiology
  - 1.7% estimated prevalence in children
  - Much more common in adults, although exact prevalence is unknown (since most are asymptomatic)

Natural History & Prognosis

- Wide-neck diverticula empty readily with bladder
- Narrow-neck diverticula often drain poorly → urinary stasis → chronic infection and stone formation
- Secondary inflammation predisposes to metaplasia/dysplasia and development of carcinoma within diverticulum
- Tumors in diverticula have poor prognosis; lack of muscularis propria leads to more rapid local spread

Treatment

- Options, risks, complications
  - Surgery may be indicated for persistent infection, stone formation, or ureteral obstruction

DIAGNOSTIC CHECKLIST

Consider

- Large diverticulum may be confused with bladder, especially if bladder is contracted

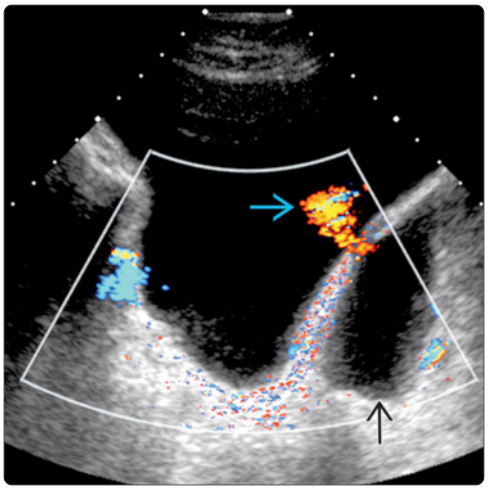
Image Interpretation Pearls

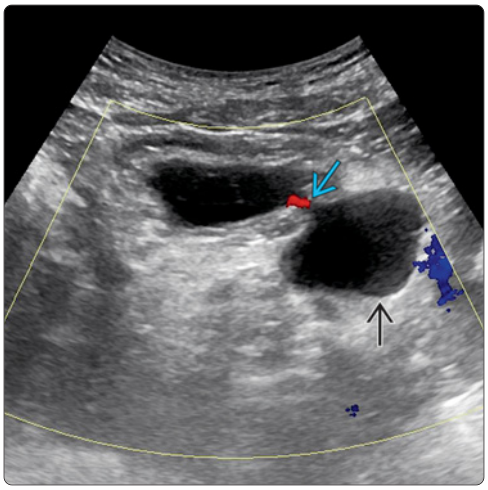
- Continuity with urethra distinguishes bladder from diverticulum

SELECTED REFERENCES

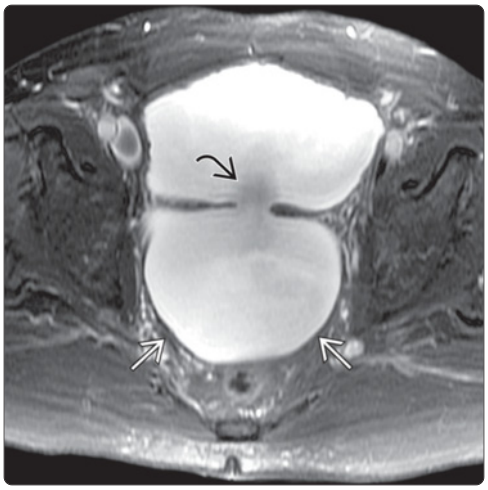
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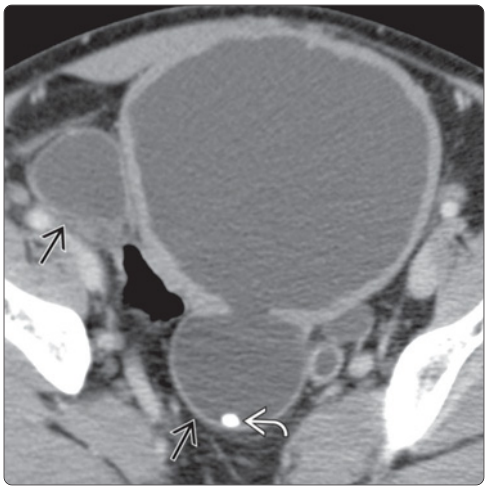
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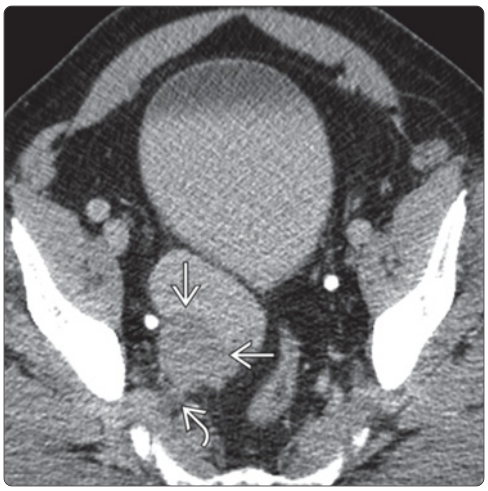


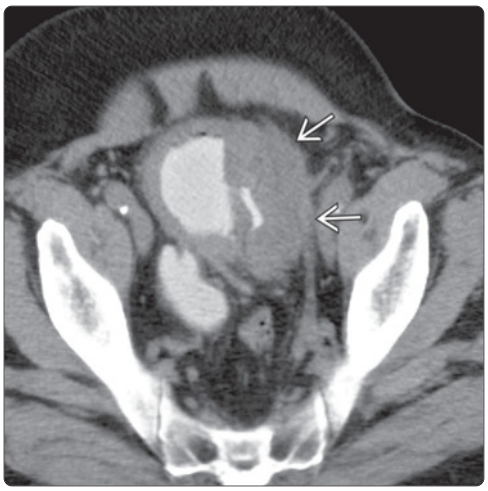
(Left) *Color Doppler ultrasound shows a large bladder diverticulum ⇨ posterior to the bladder. Note the color jet ⇨ traversing the neck of the diverticulum and the connection to the bladder lumen.* **(Right)** *Axial duplex US image in 65-year-old male shows a large diverticulum ⇨ in the left posterolateral aspect of the bladder with jet flow through its neck ⇨ due to the differing specific gravity of urine within the bladder and diverticulum.*





(Left) *Axial T2WI MR shows a large posterior bladder diverticulum ➡ connected to the bladder via a broad neck. Signal loss in the connecting channel ⇨ is due to the turbulent motion of urine and resultant dephasing.* **(Right)** *Axial CECT shows multiple bladder diverticula ⇨. The diverticulum along the posterior bladder wall contains a calcified calculus ➡. The lack of detrusor muscle in diverticula results in stasis, which can predispose to stone formation.*





(Left) *Axial CECT shows a large enhancing mass ➡ inside the diverticulum. Bulging outside the diverticulum and stranding in perivesical fat ➡ are findings highly suspicious for extramural extension. Pathology showed poorly differentiated carcinoma. Patient developed metastases to liver and lung.* **(Right)** *Axial CECT in a different patient shows marked irregular wall thickening ➡ of one of the diverticula, which was found to be the site of an invasive carcinoma.*

KEY FACTS

TERMINOLOGY

- Nonanatomic communication between bladder and skin or adjacent viscus

IMAGING

- Cystography and contrast enemas
  - Identify fistula in < 50% of cases
- CT: Good overall test for fistula and underlying etiology
  - Enterovesical fistula
    - Gas in bladder (90%)
    - CT may depict tract itself, especially if it is in plane of CT section (< 50%)
  - Vesicovaginal
    - Contrast-opacified urine in vagina
- MR: High sensitivity/specificity for depiction of colovesical fistula
  - Majority (~ 70%) of cases have associated abscess
  - Focal bladder wall thickening, air in bladder, and loss of perivesical fat plane are sensitive findings on MR

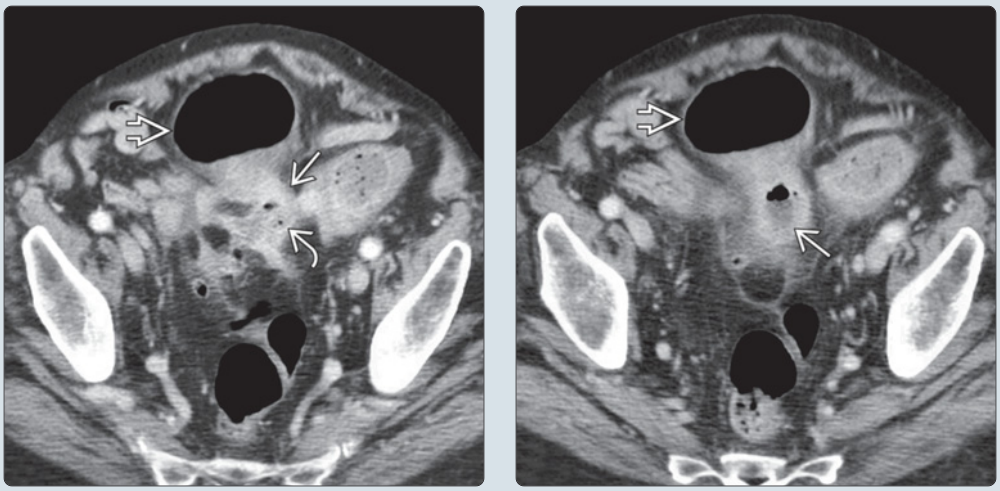
TOP DIFFERENTIAL DIAGNOSES

- Cystitis (with gas-forming bacteria)
- Emphysematous cystitis
- Presence of indwelling or recent bladder catheter

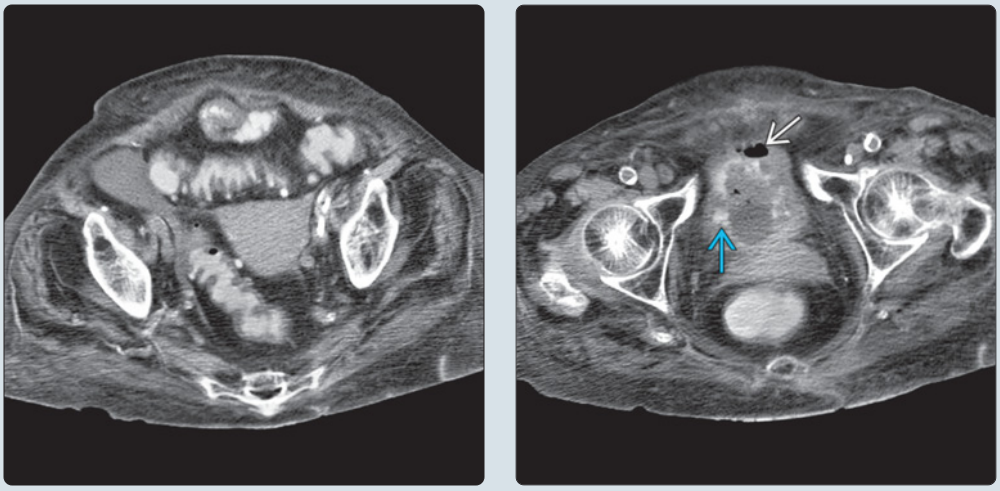
PATHOLOGY

- Colovesical fistula
  - Diverticulitis (most common cause: 80% of cases)
- Small bowel-bladder fistula
  - Crohn disease: Most common cause
- Vesicocutaneous fistula
  - Usually due to surgical complication or trauma
- Vesicovaginal
  - Gynecologic surgery (hysterectomy) most common cause
- Colovaginal fistula
  - Diverticulitis or complication of hysterectomy

(Left) *Axial CECT in a patient with colovesical fistula from diverticulitis shows extensive sigmoid diverticulosis and inflammation of the surrounding tissues. The bladder contains a large amount of gas. Note the pericolonic collection of gas and fluid tracking directly to the bladder dome. The fistula is seen as a contrast-enhancing track.* **(Right)** *Axial CECT in the same patient shows the gas within the bladder lumen and an abscess interposed between the sigmoid colon and the bladder dome.*



(Left) *Axial NECT in an elderly woman with pyuria and flank pain shows extensive sigmoid diverticular disease. Diagnosis was colovesical fistula from diverticulitis.* **(Right)** *Axial CECT in the same patient shows particulate matter, gas, and enteric contrast medium within the bladder lumen. An antegrade pyelogram showed hydronephrosis and yielded grossly infected urine as a result of a colovesical fistula.*



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TERMINOLOGY

Definitions

- Nonanatomic communication between bladder and skin or adjacent viscus
- Types include enterovesical, colovesical, appendicovesical, uterovesical, vesicovaginal, vesicocutaneous, etc.

IMAGING

General Features

- Best diagnostic clue
  - Enteric contrast medium within bladder lumen

Radiographic Findings

- Radiography
  - Gas in bladder lumen
- IVP
  - Bladder wall irregularity ± mass effect
  - Fistula tract is rarely seen
- Cystography
  - Enterovesical
    - May only see bladder wall irregularity
    - Opacification of tract and bowel seen in < 50% of cases
    - Tract may be unidirectional (e.g., bowel into bladder)
  - Vesicovaginal
    - Contrast opacifies vagina posteriorly
    - Postvoid and lateral films can be helpful to demonstrate vaginal contrast opacification
- Retrograde urethrography
  - Best for fistulas involving urethra
- Contrast enema
  - Demonstrates only 35% of colovesical fistulas
  - Can demonstrate signs of diverticulitis and other potential etiologies
  - CT may detect small amount of barium in bladder after enema
    - Too subtle to detect on radiograph
- Fistulography
  - Cannulation and contrast injection through cutaneous opening
  - Best means of documenting vesicocutaneous fistula
- Vaginography (contrast injection into vagina)
  - Limited role, mainly used for rectovaginal fistula

CT Findings

- CECT
  - Enterovesical fistula
    - Gas in bladder (seen in 90%)
    - Mural thickening of bladder and bowel
    - Enteric contrast material ± feces within bladder lumen
    - May depict tract itself, especially if it is in plane of CT section (< 50%)
    - If CT performed following cystography, opacified urine can be seen in lumen of bowel
    - Often identifies indirect signs of etiology of fistula
      - Diverticulitis: Presence of diverticula, colonic wall thickening, effaced fat plane between colon and bladder, ± adjacent abscess
      - Inflammatory bowel disease: Uniform long segment bowel wall thickening, mesenteric hyperemia
      - Primary bladder origin: May see calcification of bladder wall (schistosomiasis or bladder cancer)
  - Vesicovaginal
    - Gas in bladder
    - Contrast-opacified urine in vagina (in delayed phase or if CT performed following cystography)
  - Colovaginal fistula
    - Surgically absent uterus (hysterectomy is most common etiology)
      - May have inflammatory mass between colon and vagina
    - Diverticulitis signs (2nd most common etiology)
      - Abscess between colon and vagina

Ultrasonographic Findings

- Limited role in evaluation of fistulae
- Color Doppler US (± microbubble contrast medium) may show communication between bladder and vagina
- May be helpful in delineation of tract in vesicocutaneous fistula

MR Findings

- MR has high sensitivity and specificity for depiction of colovesical fistula
  - Majority (~ 70%) of cases have associated perivesical abscess
- Focal bladder wall thickening, air in bladder, and loss of perivesical fat plane are sensitive findings on MR
- Fistulous tract and abscess show high signal on T2WI and marked peripheral enhancement
- Post-contrast T1WI allows differentiation of abscess from inflammation and granulation tissue

Imaging Recommendations

- Best imaging tool
  - Depends on nature and site of suspected fistula
- Protocol advice
  - CT cystogram technique or CT with rectal contrast administration may improve ability to visualize tract
  - Multiplanar reformations may be helpful to demonstrate course of fistula
    - Especially important to image in sagittal plane for suspected colovesical, colovaginal, or vesicovaginal fistulas
  - Postvoid or postevacuation scans may improve fistula detection

DIFFERENTIAL DIAGNOSIS

Gas in Bladder Lumen

- Cystitis (with gas-forming bacteria)
- Emphysematous cystitis
  - Gas often present in bladder wall
- Presence of indwelling or recent bladder catheter
  - Do not expect much gas in bladder (inquire about evidence of pyuria or symptoms of fistula)
  - Gas should float on top of urine, not be suspended within it
- Recent bladder instrumentation (e.g., cystoscopy)

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Thick-Walled Bladder

- Cystitis
  - Infectious, radiation induced, chemotherapy induced, etc.
- Bladder carcinoma

Contrast Medium in Vagina

- Incontinence: Leaking urine may enter vagina via perineum

PATHOLOGY

General Features

- Etiology
  - Enterovesical fistula
    - Colovesical fistula
      - Diverticulitis (most common cause in Western industrialized countries; causes 80% of cases)
      - Rectosigmoid adenocarcinoma
      - Crohn disease
      - Radiation therapy
    - Rectovesical fistula
      - Almost always due to tumor or trauma
    - Small bowel-bladder fistula
      - Crohn disease: Most common cause
      - Foreign body (e.g., ingested chicken bone): Rare
    - Inflammatory causes: Diverticulitis, Crohn disease, granulomatous infection of bladder with TB or schistosomiasis
    - Neoplastic causes: Colon, bladder, or gynecologic cancers
    - Radiation therapy: Usually occurs 12-18 months after therapy
  - Vesicocutaneous fistula
    - Usually due to complication of surgery or trauma
    - Usually at site of prior suprapubic cystostomy after catheter removal
      - Most close spontaneously if there is no bladder outlet obstruction or wound infection
    - May follow radiation therapy (rare)
  - Vesicovaginal
    - Gynecologic surgery (usually hysterectomy): Most common cause in developed countries
    - Obstetric complications: Most common in undeveloped countries
    - Following creation of "neobladder" after radical cystectomy
      - Fistula between ileal pouch and vagina
    - Penetrating trauma and iatrogenic injury
  - Vesicourethral fistula
    - Results from urethral injury (trauma or pelvic surgery)
  - Colovaginal fistula
    - Diverticulitis or complication of hysterectomy are most common etiologies
    - Neoplasms, radiation, inflammatory bowel disease, infection, trauma
      - All uncommon etiologies
  - Rectovaginal fistula
    - Uncommon, usually result of obstetric complication
  - Ureterorectal fistula
    - Congenital in children
      - In association with imperforate anus
    - Acquired in adults
      - Complication of prostate surgery (most common)
      - Others: Neoplasms, tuberculosis, radiation therapy, instrumentation

Gross Pathologic & Surgical Features

- Rectosigmoid colon is bowel segment most frequently involved
- Cystoscopy and sigmoidoscopy show inflammatory changes but usually not fistula itself

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most common: Nonspecific cystitis symptoms
  - Fecaluria or pneumaturia present in 40-70% of cases
  - Persistent urinary tract infections (UTIs)
  - Vesicovaginal fistula: Painless dribbling incontinence of urine through vagina
- Clinical profile
  - Vesicovaginal fistula: Post gynecologic surgery
    - Preoperative irradiation predisposes to fistula
  - Enterovesical fistula: Patient with Crohn disease
  - Colovesical fistula: Patient with diverticulitis or colon cancer

Demographics

- Epidemiology
  - 0.5-1% frequency of bladder injury with total abdominal hysterectomy
    - 75% of vesicovaginal fistulas occur following hysterectomy for benign disease
  - Obstetrical trauma most common cause of vesicovaginal fistula in developing countries

DIAGNOSTIC CHECKLIST

Consider

- Enterovaginal fistulas are harder to detect than vesicovaginal fistulas

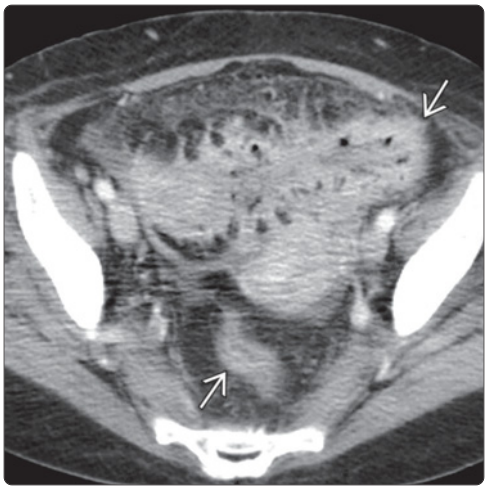
Image Interpretation Pearls

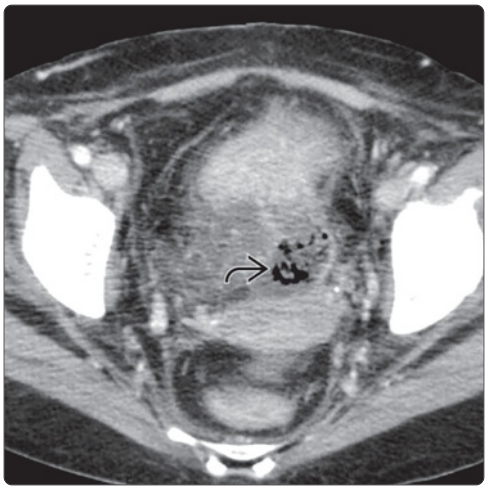
- Bladder wall irregularity may be sole clue to presence of fistula

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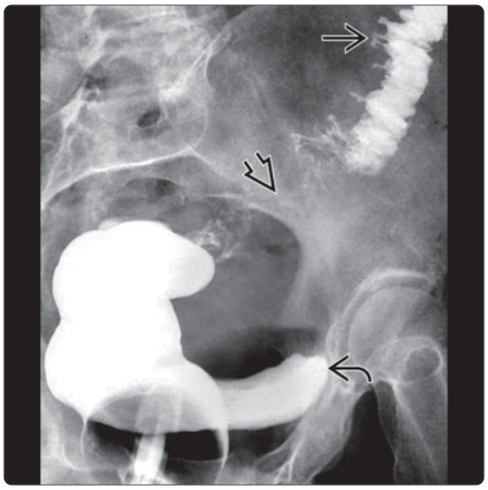
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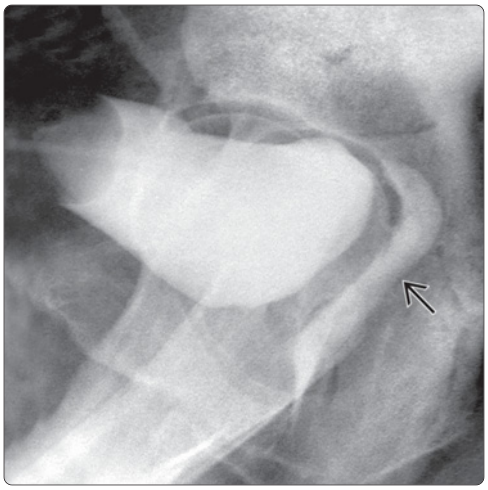
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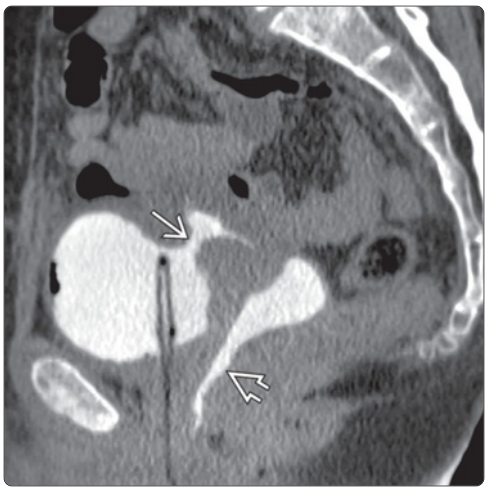


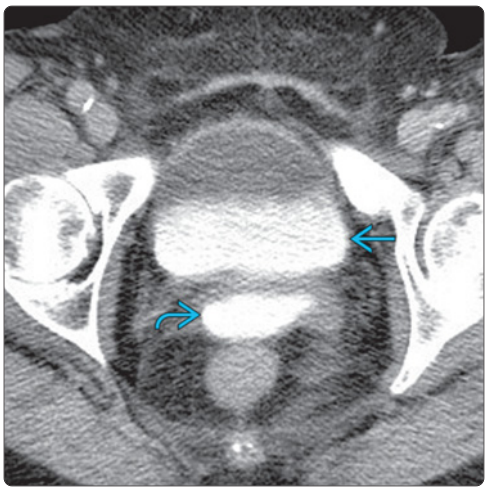
(Left) *CECT in a patient with history of colovesical fistula from granulomatous colitis shows marked inflammatory thickening of the rectosigmoid colon and mesenteric hyperemia.* **(Right)** *CECT in the same patient shows an extraluminal collection of gas and fluid between the sigmoid colon and the dome of the bladder. Gas was seen within the bladder, and a retrograde cystogram (not shown) confirmed a fistulous tract from the bladder to the colon.*





(Left) *Contrast enema in a patient with history of colovesical fistula from diverticulitis shows several colonic diverticula, spasm of the sigmoid colon, and contrast filling of the bladder, indicating a colovesical fistula.* **(Right)** *Lateral view from a cystogram performed via a suprapubic catheter in patient with history of vesicovaginal fistula following hysterectomy shows filling of the vagina posteriorly.*





(Left) *Sagittal CT cystogram in a patient with history of vesicouterine fistula following cesarean section shows a fistulous connection between the bladder, which has an indwelling catheter, and the uterus. There is subsequent filling of the vagina.* **(Right)** *Delayed image from CECT in the same patient shows opacified urine within the bladder and the vagina. It is sometimes necessary to distend the bladder or vagina with direct injection of contrast material in order to demonstrate fistulas.*

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KEY FACTS

IMAGING

- **Suprapontine** (above pontine center) lesions → detrusor hyperreflexia
  - Rounding of bladder shape and serration of mucosa above trigone (detrusor contractions)
  - ± trabeculations and ↑ wall thickness
- **Suprasacral** (above S2-4) lesions → detrusor hyperreflexia with detrusor-sphincter dyssynergia
  - Christmas tree or pine cone shape (severe): Elongated and pointed with pseudodiverticula
- **Peripheral** (below S2-4) lesions → detrusor areflexia
  - Large atonic bladder
- **Sensory** lesions → inability to sense bladder fullness
  - Rounding of bladder shape and serration of mucosa
- Secondary bladder and upper tract abnormalities
  - Trabeculation, dilated upper tracts, pseudodiverticula, vesicoureteral reflux
- Best imaging test: Cystourethrography at rest and during voiding
- Findings on US vary depending on type and level of neurological damage

TOP DIFFERENTIAL DIAGNOSES

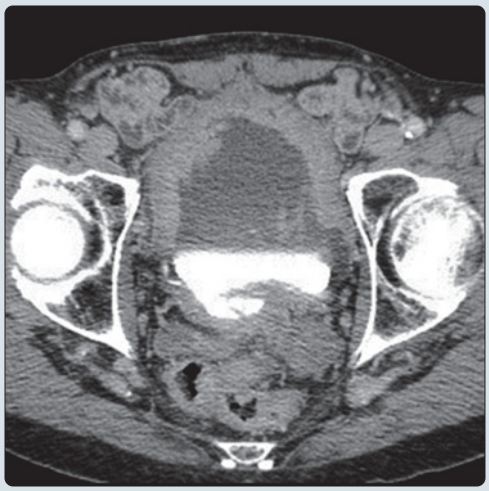
- Bladder outlet obstruction
- Bladder carcinoma

PATHOLOGY

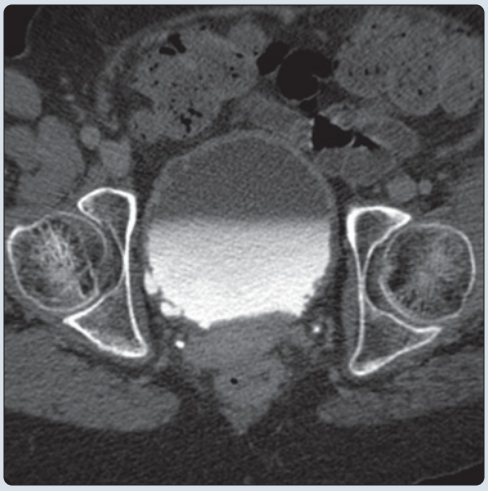
- Suprapontine: Stroke, arteriosclerosis, brain tumor, multiple sclerosis
- Suprasacral: Trauma, tumor, or multiple sclerosis
- Epiconal: Myelomeningocele, tumor, or trauma
- Peripheral (pelvic ± pudendal nerve): Pelvic surgery, cauda equina
- Sensory: Diabetes mellitus, pernicious anemia, or tabes dorsalis

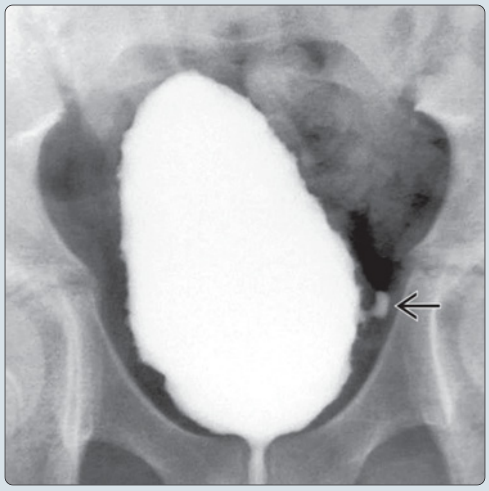
(Left) *Oblique cystogram in a young man with a history of spina bifida demonstrates an elongated, pointed, and mildly trabeculated bladder, as well as a distended posterior urethra ⇨: Features associated with suprasacral type (above S2) neurogenic bladder.* **(Right)** *Axial CECT in a young woman with a traumatic spinal cord injury shows a markedly thickened and trabeculated bladder wall compatible with neurogenic bladder.*





(Left) *Axial CECT in patient with history of spina bifida shows moderate bladder wall thickening and trabeculation.* **(Right)** *Cystogram in the same patient shows a pine cone appearance of the bladder, characterized by an elongated and pointed shape and pseudodiverticula ⇨.*





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TERMINOLOGY

Definitions

- Bladder dysfunction caused by neurologic damage
  - Categorized based on level of neurologic damage into suprapontine, spinal (suprasacral, epiconal), peripheral, and sensory types

IMAGING

General Features

- Best diagnostic clue
  - Contracted or distended bladder

Radiographic Findings

- Cystourethrography or cystography findings
  - Suprapontine (above pontine center) lesions → detrusor hyperreflexia
    - Rounding of bladder shape and serration of mucosa above trigone (detrusor contractions)
    - ± trabeculations and ↑ wall thickness
  - Suprasacral (above S2-4) lesions → detrusor hyperreflexia with detrusor-sphincter dyssynergia
    - Beak of contrast (slightly open posterior urethra)
    - Contrast extends to external sphincter/urogenital diaphragm (distended posterior urethra)
    - Christmas tree or pine cone shape (severe): Elongated and pointed with pseudodiverticula
  - Epiconal (in and around S2-4) lesions → weak detrusor hyperreflexia (storing), detrusor areflexia (voiding), detrusor-sphincter dyssynergia
    - Upper and lower motor neuron features; pine cone
  - Peripheral (below S2-4) lesions → detrusor areflexia
    - Large atonic bladder: No signs of detrusor contraction during voiding and poor emptying
    - Downward displacement of bladder base and vagina → functional infravesical obstruction (females)
  - Sensory lesions → inability to sense bladder fullness
    - Voiding is preserved early on
    - Rounding of bladder shape and serration of mucosa
    - Large atonic and smooth bladder: Indistinguishable from peripheral nerve lesions (chronic)
  - Secondary bladder and upper tract abnormalities
    - Trabeculation, dilated upper tracts, pseudodiverticula, vesicoureteral reflux
    - Indistinguishable between detrusor-sphincter dyssynergia and functional infravesical obstruction

CT Findings

- Bladder wall thickening, trabeculation, and pseudodiverticula
- Hydroureter/hydronephrosis and renal atrophy in severe chronic cases

Ultrasonographic Findings

- Grayscale ultrasound
  - Findings vary depending on type and level of neurological damage
  - Small contracted or large atonic bladder; wall thickening; vesicoureteric reflux during voiding; increased postvoid residual urine

DIFFERENTIAL DIAGNOSIS

Bladder Outlet Obstruction

- Due to urethral strictures, benign or malignant enlargement of prostate, congenital urethral valves
- Trabeculation of detrusor muscle and urine retention
- Detrusor hyperreflexia (early) → areflexia (chronic)

Bladder Carcinoma

- Bladder wall thickening in setting of carcinoma is focal and eccentric

PATHOLOGY

General Features

- Etiology
  - Suprapontine: Stroke, arteriosclerosis, brain tumor, Parkinson disease, Shy-Drager syndrome, multiple sclerosis
  - Suprasacral: Trauma, tumor, multiple sclerosis
  - Epiconal: Myelomeningocele, tumor, or trauma
  - Peripheral (pelvic ± pudendal nerve): Pelvic surgery, cauda equina
  - Sensory: Diabetes mellitus, pernicious anemia, or tabes dorsalis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Frequency, nocturia, urgency, retention, incontinence
  - Hyper- or hyporeflexia; impaired or no sensation

Natural History & Prognosis

- **Complications**: Infection, reflux, hydronephrosis, urolithiasis, sexual dysfunction, autonomic dysreflexia
- Good prognosis if treated at early stage
- Poor prognosis if not treated early; leads to permanent renal damage

Treatment

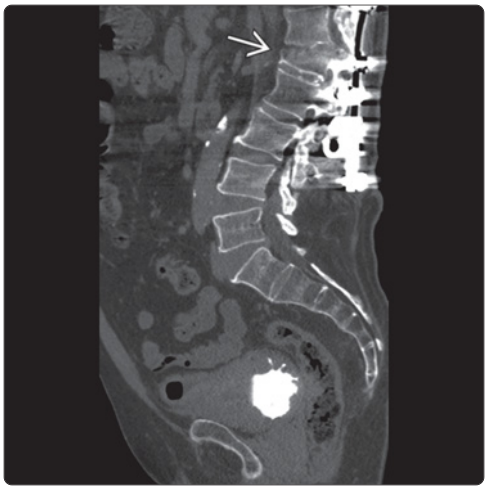
- Hyperreflexia
  - Increasing bladder volume: Bladder augmentation cystoplasty, muscular or fascial slings, parasympatholytic drugs, botulinum type a toxin
  - Increasing voiding frequency: Catheterization, transurethral sphincterotomy
- Hyporeflexia
  - Bladder training, catheterization, bladder neck resection/denervation, parasympathomimetic drugs

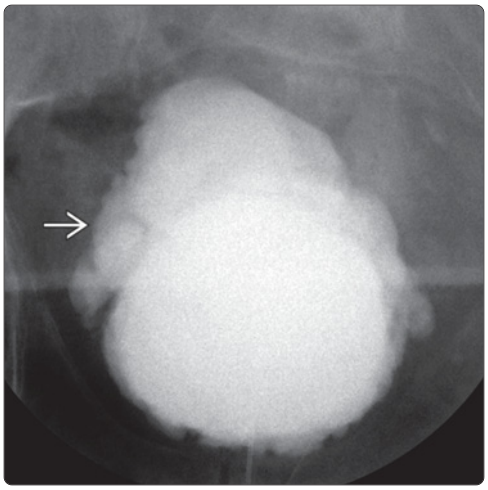
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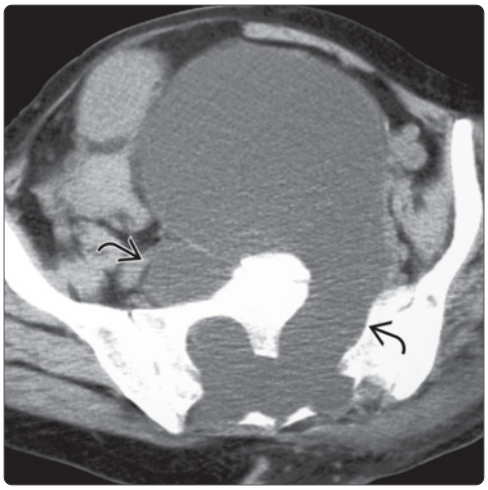
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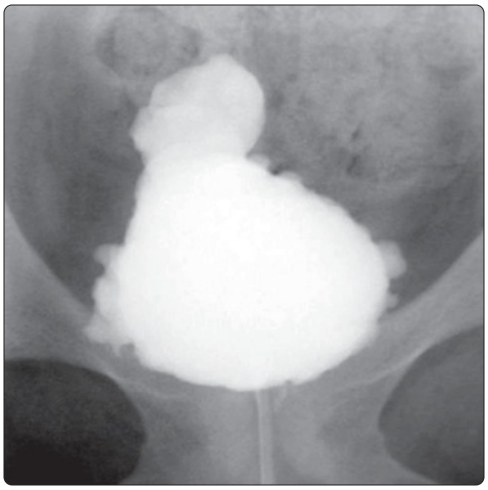
(Left) *Sagittal CECT image obtained during urographic phase shows a markedly thickened and trabeculated bladder in this patient with history of a neurogenic bladder due to traumatic spine injury. Note compression fracture of the L1 vertebral body.* **(Right)** *Retrograde cystogram in the same patient better delineates the trabeculae. This appearance can also be seen with chronic urinary outlet obstruction.*



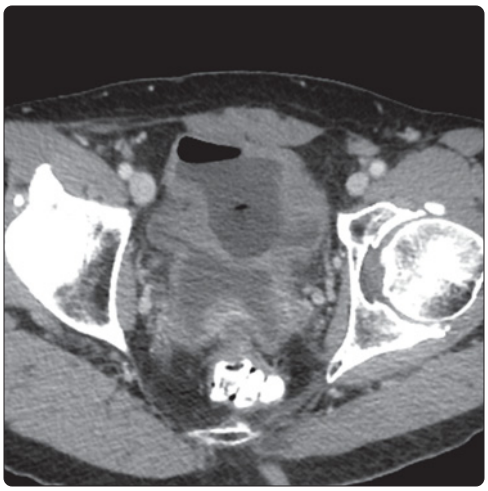


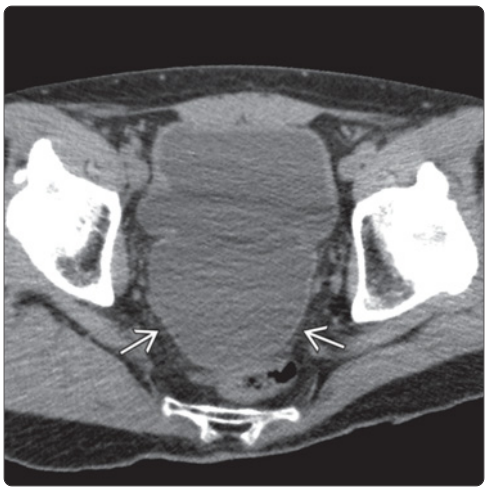
(Left) *Axial CECT in a patient with spina bifida shows a distended neurogenic bladder and anterior meningocele.* **(Right)** *Frontal cystogram shows a markedly trabeculated bladder, resembling a Christmas tree, often associated with detrusor hyperreflexia. This is usually associated with suprasacral lesions.*

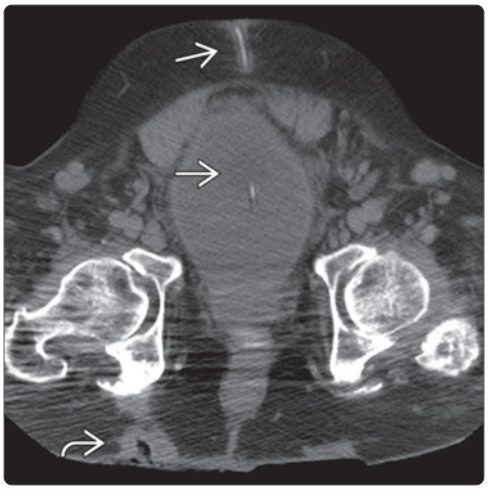


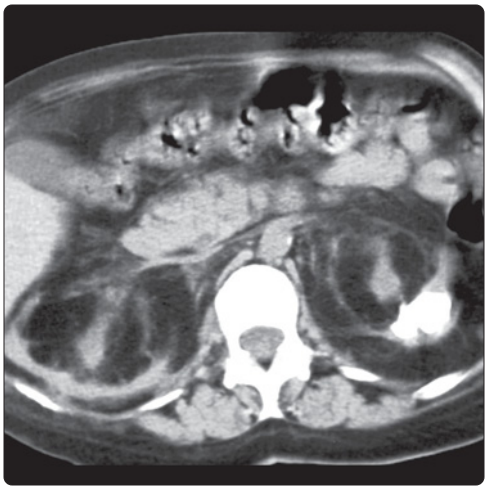


(Left) *Axial CT in patient with history of stroke demonstrates marked bladder wall thickening and trabeculation.* **(Right)** *Another CT in the same patient better delineates the large diverticulum along the posterior bladder wall. These findings are compatible with suprapontine type of neurogenic bladder.*

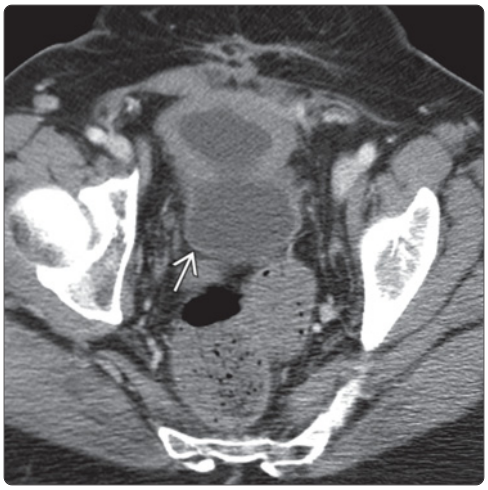


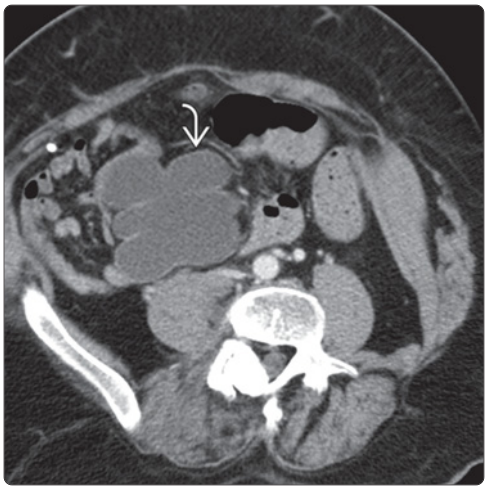




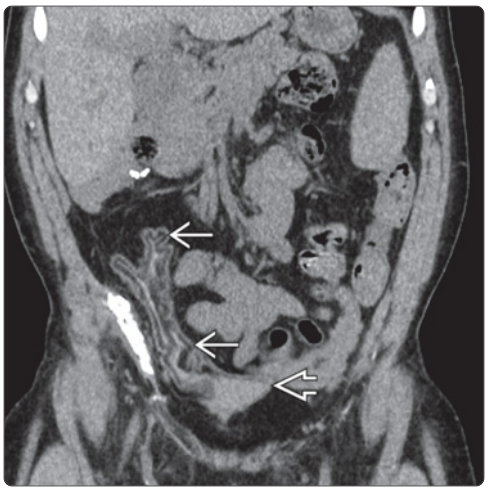


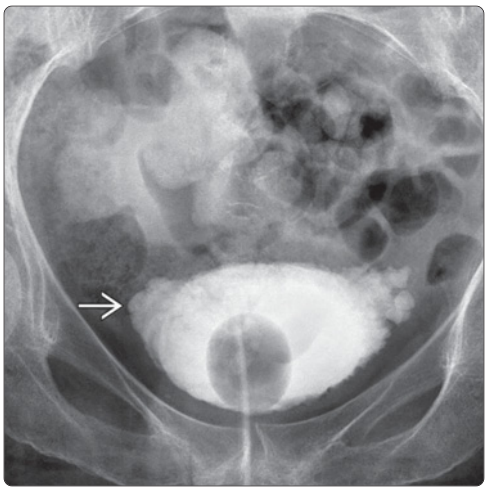
(Left) *Axial NECT in a patient with history of gunshot wound injury to the lower thoracic spine shows a distended bladder with a suprapubic catheter in place. Note the decubitus ulcer extending to the ischium in this paralyzed patient.* **(Right)** *Axial NECT in a patient with neurogenic bladder shows bilateral renal cortical atrophy and parenchymal calcification due to chronic reflux.*





(Left) *Axial CECT in a patient with history of neurogenic bladder in the setting of a spine tumor shows marked bladder wall thickening and a large diverticulum along the posterior bladder wall.* **(Right)** *Axial CT in the same patient shows postsurgical changes of bladder augmentation with use of a segment of ileum.*





(Left) *Coronal unenhanced CT shows an augmented bladder in a patient with a history of a neurogenic bladder. Note submucosal fat deposition in the ileal loop used for augmentation. Excreted contrast is seen in a small-capacity urinary bladder.* **(Right)** *Frontal projection from a retrograde cystogram in a patient with a history of multiple sclerosis shows bladder trabeculation.*

Bladder Trauma

KEY FACTS

TERMINOLOGY

- Injury due to blunt, penetrating, or iatrogenic trauma

IMAGING

- Extravasation of contrast-opacified urine at cystography (CT or conventional): Almost 100% accuracy
- Contusion (type 1)
  - Ecchymosis of localized segment of bladder wall
- Intraperitoneal rupture (type 2)
  - Opacified urine in peritoneal spaces (pouch of Douglas), outlining bowel loops & intraperitoneal viscera
  - Requires surgical repair
- Interstitial injury (type 3)
  - Intramural tear with intact serosa
- Extraperitoneal rupture (type 4)
  - Perforation by bony spicules of pelvic fractures
  - **"Molar tooth" sign**: Extravasated perivesical urine assumes rounded configuration cranially & pointed contours inferolaterally
  - Usually managed nonoperatively
  - Complex (type 4B): Extravasation extends beyond perivesical space; thigh, scrotum, etc.
- Combined injury (type 5): Intra- and extraperitoneal rupture

TOP DIFFERENTIAL DIAGNOSES

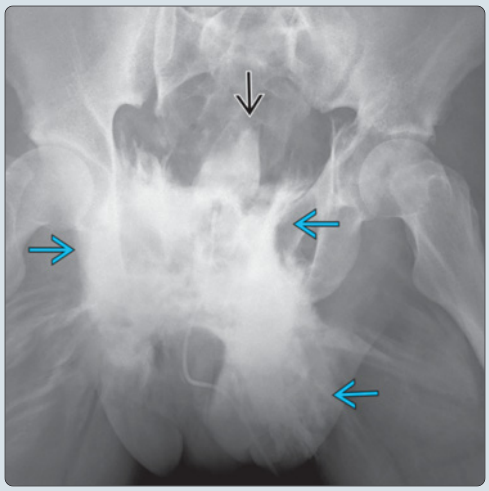
- Male urethral injury
  - Extravasated urine can extend into perivesical spaces
- Pelvic or abdominal bleeding
  - Active hemorrhage may simulate bladder rupture
- Blood clot in bladder

PATHOLOGY

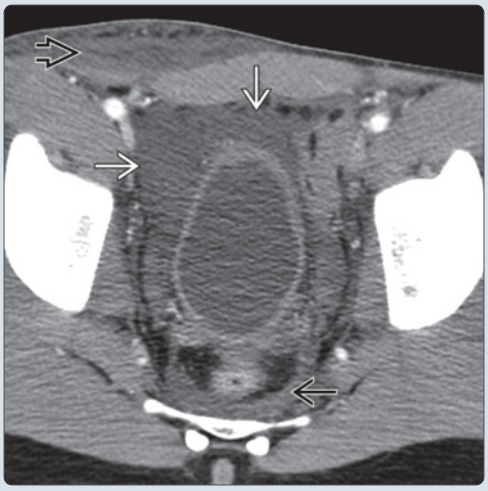
- Location of extravasation depends on anatomy
  - Anterosuperior rupture: Extravasation to intraperitoneal, Retzius space, or both
  - Injury below peritoneal reflection → extraperitoneal (paravesical, presacral spaces)

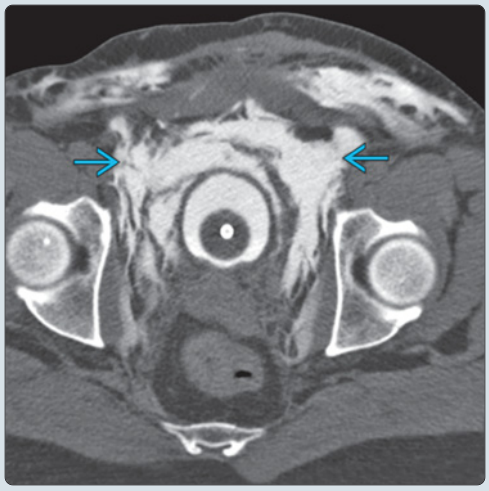
(Left) *Axial CECT reveals diastasis of the pubic symphysis and fluid within the ventral soft tissues; it is difficult to determine if this is due to urine, hematoma, or a combination of both.* **(Right)** *AP view from a follow-up cystogram in the same patient shows extravasation of contrast from the urinary bladder into the extraperitoneal tissues surrounding the bladder and along the left superior thigh and scrotum. A small volume of contrast is within the urinary bladder.*





(Left) *Axial CECT during the initial trauma evaluation reveals pericystic fluid for which differential considerations include urine and blood. Mild hemorrhage resides in the ventral subcutaneous tissues and presacral space.* **(Right)** *Axial CT cystogram in the same patient reveals that the pericystic fluid was from extravasated contrast-opacified urine due to extraperitoneal bladder rupture. Note the "molar tooth" configuration of the extraperitoneal contrast in the pericystic spaces.*





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TERMINOLOGY

Definitions

- Bladder injury due to blunt, penetrating, or iatrogenic trauma

IMAGING

General Features

- Best diagnostic clue
  - Extravasation of contrast-opacified urine at cystography (CT or conventional)
- Other general features
  - Classification of bladder injury after blunt trauma
    - Type 1: Bladder contusion
    - Type 2: Intraperitoneal rupture
    - Type 3: Interstitial injury
    - Type 4A: Simple extraperitoneal rupture
    - Type 4B: Complex extraperitoneal rupture
    - Type 5: Combined injury

Radiographic Findings

- Cystography
  - Bladder contusion (type 1)
    - Pear-shaped bladder: Extrinsic compression of bladder (symmetrical extraperitoneal hematoma)
  - Intraperitoneal rupture (type 2)
    - Intraperitoneal contrast: Outlines small bowel loops & intraperitoneal viscera; layers in dependent peritoneal recesses (pouch of Douglas, Morrison's pouch)
    - ± pelvic fractures
  - Interstitial injury (type 3)
    - Focal mural defect along wall of bladder
  - Extraperitoneal rupture (type 4)
    - Simple (type 4A)
      - Flame-shaped extravasation around bladder
    - Complex (type 4B)
      - Extravasation extends beyond pelvis
    - Extravasation often best seen on post-drainage films
  - Combined rupture (type 5)
    - Features of intra- and extraperitoneal ruptures
  - Penetrating injury
    - Foreign bodies (e.g., metallic fragments from bullet) along course of penetration
    - Extravasation ± foreign body (e.g., bullet) best seen on post-drainage films

CT Findings

- CT cystography
  - Bladder contusion (type 1)
    - Normal findings
  - Intraperitoneal rupture (type 2)
    - Opacified urine fills peritoneal spaces (pouch of Douglas, Morrison's pouch), outlines bowel loops & intraperitoneal viscera
  - Interstitial injury (type 3)
    - Intramural & submucosal extravasation of contrast without transmural extension
  - Extraperitoneal rupture (type 4)
    - Perforation by bony spicules of pelvic fractures
    - "Knuckle" of bladder: Trapped bladder by displaced fracture of anterior pelvic arch
    - Simple (type 4A): Extravasation is confined to perivesical space
    - Complex (type 4B): Extravasation extends beyond perivesical space; thigh, scrotum, penis, perineum, anterior abdominal wall, retroperitoneum (pararenal or perirenal spaces), or hip joint
    - **"Molar tooth" sign**: Extravsated perivesical contrast assumes rounded contour cranially & pointed contours inferolaterally
  - Combined injury (type 5)
    - Features of intra- & extraperitoneal ruptures

Ultrasonographic Findings

- Grayscale ultrasound
  - "Bladder within bladder": Fluid around bladder
  - Intravesical hematoma
  - Fluid in abdomen & pelvis (e.g., pouch of Douglas)

Imaging Recommendations

- Best imaging tool
  - Cystography (CT or conventional): 95-100% sensitivity
- Protocol advice
  - Cystography
    - Exclude urethral injury in males prior to cystography; if positive, use suprapubic cystostomy tube
    - Post-drainage film to check for extravasation hidden by contrast
    - CT cystography: Instill diluted contrast (10 mL contrast, 300 mL saline)
      - Multiplanar reformation aids visualization of leak

DIFFERENTIAL DIAGNOSIS

Simple Male Urethral Injury

- Contrast extravasation into base of penis or thigh may result from urethral or extraperitoneal bladder injury
- Blood in urethral meatus suggestive of urethral injury; avoid "blind" catheterization of bladder

Hemoperitoneum

- Active extravasation of enhanced blood may simulate extravesical urine from bladder rupture
  - Compare with density of blood & urine
- CT cystography usually diagnostic

Pelvic Bleeding

- Bleeding from other pelvic trauma can appear similar to contrast extravasation from bladder
- Synchronous occurrence with bladder trauma likely; complete pelvic assessment needed

Blood Clot in Bladder

- Blood from renal or bladder injury may cause irregular filling defect within bladder

PATHOLOGY

General Features

- Etiology
  - Blunt trauma: Seat belt or steering wheel injury
  - Penetrating injury: Gunshot or knife wounds

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- Iatrogenic injury: Pelvic, urological, obstetrical, & gynecological procedures
- Location of extravasation depends on anatomy
  - Anterosuperior rupture: Extravasation to intraperitoneal, Retzius space, or both
  - Posterosuperior rupture: Extravasation to intraperitoneal, extraperitoneal, or both
  - Injury below peritoneal reflection → extraperitoneal (paravesical, presacral spaces)
  - Intact superior fascia of urogenital diaphragm prohibit extravasation from escaping pelvis
  - Intact inferior fascia of urogenital diaphragm prohibit extravasation into perineum
- Bladder contusion (type 1)
  - Incomplete or partial tear of bladder mucosa; ecchymosis of localized segment of bladder wall
  - Most common minor bladder injury
  - Diagnosed by exclusion of more serious injury
- Intraperitoneal rupture (type 2)
  - Direct blow to lower abdomen with distended bladder
  - ↑ bladder pressure → horizontal tear along bladder wall; at dome of bladder covered by peritoneum
  - 25% of major bladder injuries
  - More common in children due to intraabdominal bladder
- Interstitial injury (type 3)
  - Intramural or partial thickness laceration with intact serosa
  - Incomplete perforation; of either intra- or extraperitoneal portion of bladder
  - Rare type of bladder injury
- Extraperitoneal rupture (type 4)
  - Classic mechanism: Anterolateral laceration at base of bladder by bony spicules (anterior pelvic arch fractures)
  - Other mechanism: Stress applied to hypogastric wing or puboprostatic ligaments → laceration; direct blow to distended bladder
  - 62% (most common) of major bladder injuries
- Combined rupture (type 5)
  - Both intra- & extraperitoneal bladder rupture
  - 12% of bladder ruptures, can result from both blunt and penetrating trauma
- Penetrating injury
  - Results in intraperitoneal, extraperitoneal, or combined bladder injury
  - ↑ incidence of vascular injuries → high mortality

Gross Pathologic & Surgical Features

- Asymmetrical compression, congestion, laceration, or fill-thickness tear

Microscopic Features

- Mural defect; contusion or clot within bladder mucosa

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Gross hematuria (84-95%)
  - Suprapubic pain and tenderness, anuria, fever
  - Bladder contusion (type 1): Hematuria
  - Intraperitoneal rupture (type 2)
    - No bowel sounds; acute abdomen
- Laboratory data
  - Urinalysis: Hematuria
  - Intraperitoneal rupture (type 2)
    - Chemistry: Hypernatremia, hyperkalemia, uremia, and acidosis (reabsorption of urine)
    - Peritoneal lavage: Urinary ascitic fluid
    - Catheterization: No urine

Natural History & Prognosis

- Complications: Fistulas, sinuses, abscess, sepsis, bladder calculi, hematoma, hemorrhage, or shock
- Minor injuries: Good, spontaneous healing within 10-14 days
- Major injuries: Poor, if not corrected surgically and with complications
  - Bladder trauma including penetrating injury: 12-22% mortality
  - Penetrating injury: 12% mortality

Treatment

- Bladder contusion (type 1) and interstitial injury (type 3): Self limiting, no treatment
- Intraperitoneal rupture (type 2): Open surgical correction via transperitoneal approach; postoperatively, use suprapubic and urethral catheters
- Extraperitoneal rupture (type 4): Antibiotics and urethral catheter drainage (10 days); large blood clots or bladder neck injuries require surgery
- Penetrating injury: Exploratory laparotomy with surgical correction
- Follow-up: Cystography 10 days after conservative or surgical treatment

DIAGNOSTIC CHECKLIST

Consider

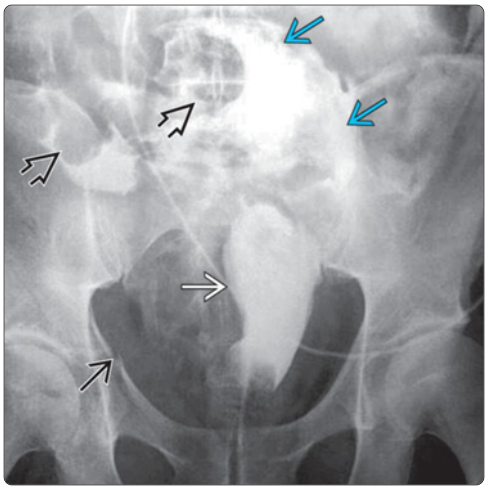
- Cystography ± CT still procedure of choice
- Bloody urethral discharge requires urethrogram prior to catheterization of bladder

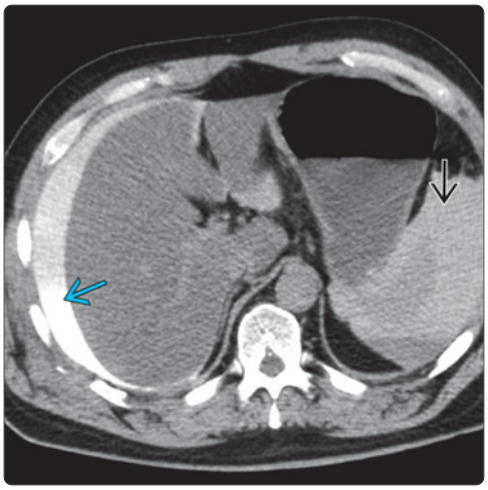
Image Interpretation Pearls

- CT only after IV contrast may miss bladder rupture
- CT cystogram best single study

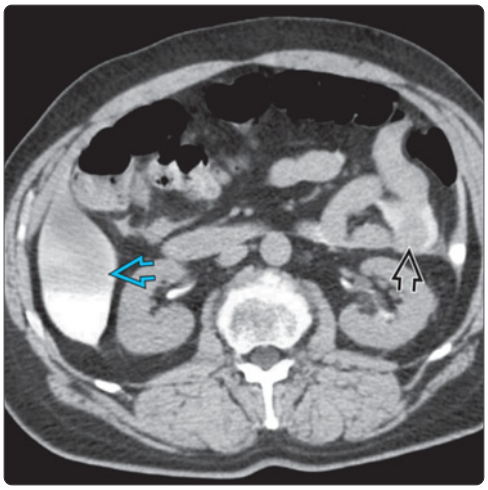
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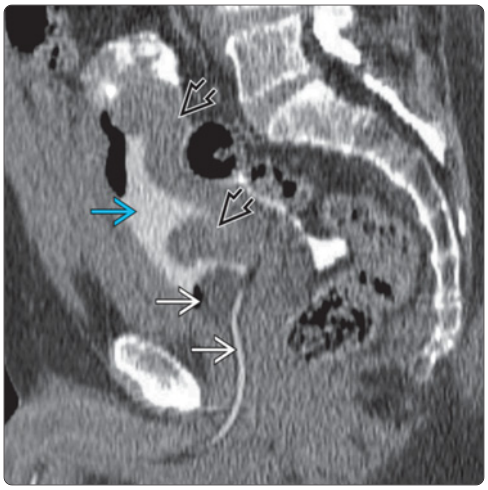
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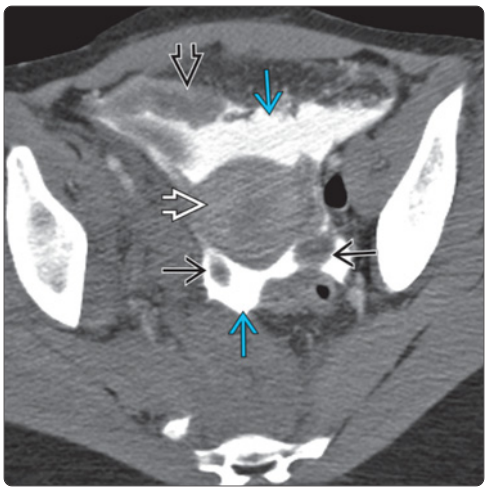


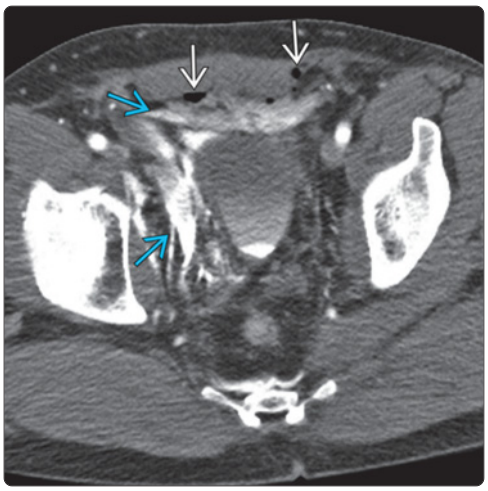
(Left) *AP view during cystogram reveals bladder displacement due to hematoma, fracture, and intraperitoneal extravasation of urine; note that bowel loops are outlined by the extravasated urine.* **(Right)** *Axial NECT in the same patient shows opacified urine layering in the perihepatic space. The density of the fluid around the spleen is mixed, due to a splenic laceration with hematoma and intermixed extravasated urine.*





(Left) *Axial NECT in the same patient shows the kidneys excreting contrast-opacified urine due to absorption of contrast from the intraperitoneal fluid. Intraperitoneal opacified urine outlines small bowel loops and distends Morrison's pouch.* **(Right)** *Sagittal NECT reformation in the same patient shows the collapsed bladder due to a Foley catheter and intraperitoneal extravasation of urine surrounding bowel loops.*





(Left) *Axial CECT following cystogram demonstrates extravasated, contrast-opacified urine surrounding the uterus, ovaries, and small bowel loops, consistent with intraperitoneal bladder rupture.* **(Right)** *Axial CECT following cystogram shows extravasated contrast-opacified urine in the perivesical spaces due to extraperitoneal bladder rupture. A few locules of gas deep to the rectus abdominis musculature were due to traumatic soft tissue injuries & pelvic fractures.*

Postoperative State, Bladder

KEY FACTS

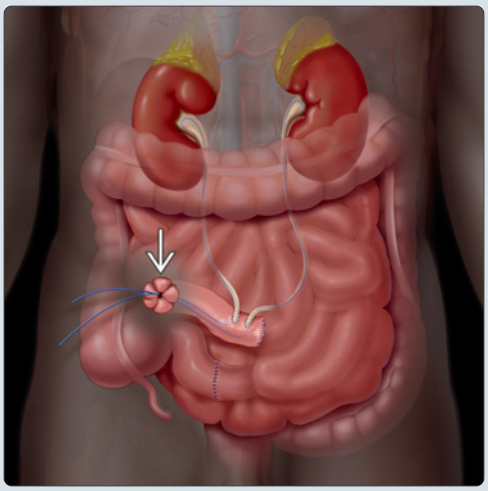
TERMINOLOGY

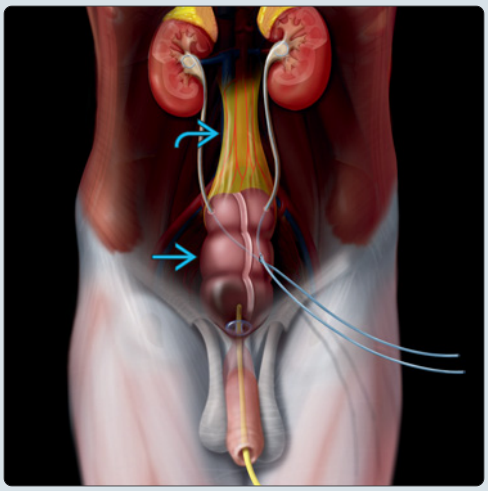
- **Segmental cystectomy**
  - Removing full-thickness portion of bladder wall
  - Used for benign and malignant lesions of bladder
- **Simple cystectomy**
  - Total resection of bladder
- **Radical cystectomy**
  - Resection of bladder with its peritoneal covering, perivesical fat, lower ureters, urachal remnant, regional lymph nodes, and adjacent structures
- **Urinary diversion techniques**
  - **Incontinent** diversion techniques
    - Cutaneous ureterostomy, Ileal conduit creation
    - Usually performed in patients with poor prognosis
  - **Continent** diversion techniques
    - Cutaneous diversion, orthotopic neobladder reconstruction
    - More complicated techniques. Method of choice in patients with better prognosis
- **Bladder augmentation**
  - Procedure to increase bladder capacity
  - For patients who lack adequate bladder capacity or detrusor compliance
- **Psoas-hitch ureterocystostomy**
  - Technique used for ureteral reimplantation if ureter is not long enough to be sutured without tension

CLINICAL ISSUES

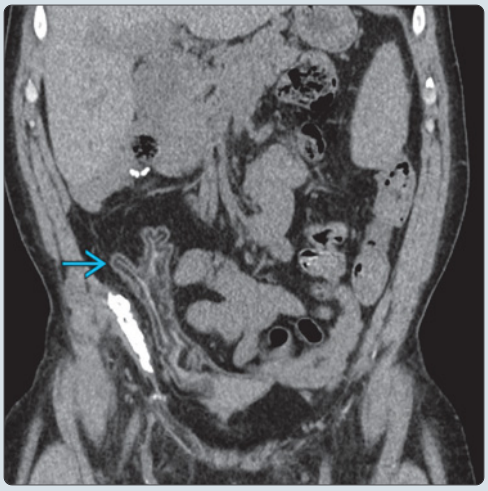
- **Complications**
  - Early complications (< 1 month)
    - Ileus, obstruction, urinary leak, urinary obstruction, fluid collection (urinoma, hematoma, lymphocele, abscess)
  - Late complications
    - Ureteral stenosis, infection, parastomal herniation, lithiasis, tumor recurrence, metabolic acidosis

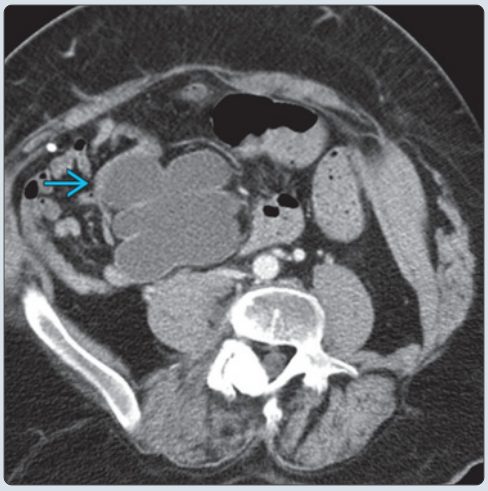
(Left) *Graphic shows an ileal conduit diversion in which a segment of ileum is mobilized and serves as a reservoir and conduit for urine. The cutaneous stoma ➡ is at one end and the ureters are anastomosed to the other end with temporary stents in place.* **(Right)** *Graphic shows a simplified variation of a "neobladder" created by mobilizing the ascending colon ⇨ and its mesentery ⇨. The ureters are anastomosed to the colon and are stented temporarily. A Foley catheter stents the anastomosis to the urethra.*





(Left) *Coronal CT in a patient with history of a neurogenic bladder shows bladder augmentation ⇨ using a segment of ileum. Note the submucosal fat deposition in the ileal loop used for augmentation.* **(Right)** *Axial CT in a patient with a history of a neurogenic bladder, in the setting of a spine tumor, shows bladder augmentation ⇨ utilizing a segment of ileum.*





TERMINOLOGY

Definitions

- Anatomical alterations resulting from bladder resection or modification
- **Segmental cystectomy**
  - Also called partial cystectomy
  - Used for benign and malignant lesions of bladder
  - Removing full-thickness portion of bladder wall
  - Benign indications
    - Colovesical or vesicovaginal fistula (e.g., from diverticulitis)
    - Bladder diverticulum
    - Ulcerative interstitial cystitis
    - Cavernous hemangioma of bladder wall
    - Localized endometriosis of bladder
    - Poor surgical risk for more aggressive procedures
  - Malignant indications
    - High-grade cancer that invades bladder wall but does not involve bladder trigone, vesical neck, or posterior urethra
    - Inability to resect these tumors transurethrally
  - Bladder contour defects are most evident soon after surgery
    - Bladder gains its normal capacity and contour over time
- **Simple cystectomy**
  - Total resection of bladder
  - Indications
    - Radiation cystitis
    - Interstitial cystitis
    - Noncorrectable fistula to bowel or other structure
    - Intractable bleeding from bladder
- **Radical cystectomy**
  - Resection of bladder with its peritoneal covering, perivesical fat, lower ureters, urachal remnant, regional lymph nodes, and adjacent structures
    - Men: Prostate, seminal vesicles, distal vas deferens
    - Women: Uterus, anterior vaginal wall, ovaries, fallopian tubes, urethra, and external meatus
    - Some form of urinary diversion and reservoir must be provided
    - Indications
      - Infiltrating bladder cancer without evidence of metastasis or with low-volume resectable locoregional metastases
      - Superficial bladder tumors refractory to cystoscopic resection
      - Stage T1, grade III tumors unresponsive to intravesical BCG vaccine therapy
      - Primary adenocarcinoma, squamous cell carcinoma, or sarcoma
      - Palliation for pain, bleeding, or urinary frequency
- **Urinary diversion techniques**
  - Urinary diversion techniques can be divided into **incontinent** and **continent** techniques
    - Incontinent diversion techniques
      - Cutaneous ureterostomy
      - Ileal conduit creation
      - Usually performed in patients with poor prognosis
    - Continent diversion techniques
      - Cutaneous diversion (Kock, colonic reservoir)
      - Orthotopic neobladder reconstruction (Studer, Hautmann, Padovan, Mainz, Mayo techniques)
      - More complicated techniques; method of choice in patients with better prognosis
      - Based on creation of a low-pressure, low-residue reservoir using segment of bowel to maintain continence
  - **Cutaneous ureterostomy**
    - Ureters are directly anastomosed to anterior abdominal wall
    - Rarely performed, due to high chance of ureteral stenosis at stoma
  - **Ileal conduit diversion (Bricker procedure)**
    - 15-20 cm ileal segment is isolated, and both ureters are anastomosed to its proximal end
    - Easiest procedure to perform
      - Continence and voluntary voiding are lost (plus altered body image)
      - Usually performed in patients with poor prognosis
    - Distal end provides urinary drainage to cutaneous stoma (usually in right side of abdomen)
    - Preserves 15-20 cm-long segment of distal/terminal ileum to maintain absorption of bile salts and vitamins
    - Trace ileal conduit back from stoma to identify it on imaging studies
      - Will appear as small bowel segment with water-density contents, except on delayed phase enhanced imaging (when urine is opacified)
  - **Cutaneous diversion with Kock technique**
    - 70 cm length of ileum is isolated
    - Reservoir is created by detubularization of central 50 cm segment
    - Remaining proximal and distal segments are then used to create 2 valves to prevent reflux
      - 1 valve provides cutaneous egress
      - Other valve is anastomosed to ureters
  - **Cutaneous diversion with colonic reservoir**
    - Segment of cecum is used to create reservoir
    - Continence is provided by catheterizable tunnel and stoma created by appendix with cuff of cecum **(Mitrofanoff technique)**
  - **Orthotopic bladder replacement ("neobladder")**
    - 40-60 cm segment of ileum **(Studer technique)**, segment of cecum and ileum **(Mainz technique)**, or entire right colon **(Mayo technique)** is used to construct neobladder
      - CT will show stapled anastomosis (bowel to bowel) and reservoir
    - Neobladder will be anastomosed to remaining part of bladder or directly to the urethra
- **Bladder augmentation**
  - Also called augmentation cystoplasty
  - Procedure to increase bladder capacity
  - Utilized for patients who lack adequate bladder capacity or detrusor compliance
    - Decreased bladder capacity or abnormal compliance may manifest as debilitating urgency, frequency, incontinence, recurrent urinary tract infections, or progressive renal insufficiency

- Indications
  - Interstitial cystitis, radiation cystitis, chronic cystitis
  - Detrusor instability, cloacal extrophy
  - Spinal cord injury, multiple sclerosis, myelodysplasia
- Isolated loop of ileum &/or segment of colon is used to augment bladder
- Enteric portion is attached to superior surface of bladder
- Attached bowel folds may be seen as irregular wall thickening on CT and cystography
- Asymmetric superior border of bladder on imaging

- **Psoas-hitch ureteroneocystostomy**
  - Technique used for ureteral reimplantation if ureter is not long enough to be sutured without tension
  - Used after segmental resection of ureter
  - Ipsilateral side of bladder is mobilized superiorly and sutured to psoas muscle to release tension
  - Asymmetric bladder on imaging
    - Ipsilateral side of bladder is tilted superiorly
    - Contralateral side is displaced inferiorly

IMAGING

Imaging Recommendations

- Protocol advice
  - Multidetector, multiphase CT urography
    - All phases obtained as thin sections from costovertebral angles to pelvic floor
    - Start by having patient drink 500-750 mL of water for hydration and as "oral contrast"
      - Unenhanced phase to detect urinary calculi
      - Nephrographic phase (80-second delay) for fluid collections, tumor recurrence
      - Excretory phase (10-minute delay) to detect urinary leaks and recurrent urothelial tumor
    - Reconstruct 2D and 3D multiplanar MIP and volume rendered images

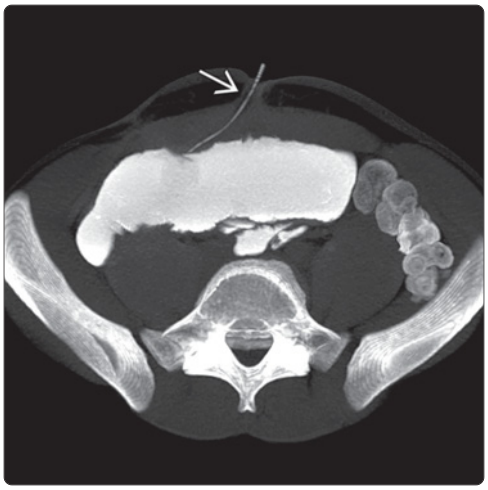
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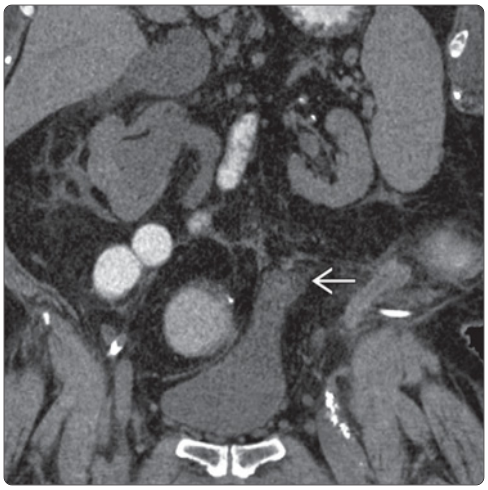
Natural History & Prognosis

- **Complications**
  - Early complications (< 1 month)
    - Ileus, obstruction, urinary leak, urinary obstruction, fluid collection (urinoma, hematoma, lymphocele, abscess)
  - Late complications
    - Ureteral stenosis, infection, parastomal herniation, lithiasis, tumor recurrence, metabolic acidosis
  - Ileus
    - Common; affects 18-25% of patients after urinary diversion
    - Usually resolves within 1 week
  - Bowel obstruction
    - Adhesive bowel obstruction near enteroenteric anastomosis is most common
    - CT shows transition from dilated to normal bowel near stapled anastomosis
  - Urinary leakage
    - Occurs in ~ 5% of patients, usually at anastomosis of ureters to reservoir
    - Collection of opacified urine seen on delayed CECT
  - Hematoma
    - Heterogeneous increased attenuation on CT in early hematoma
  - Lymphocele
    - Round/ovoid, fluid-density lesion
    - Very thin wall &/or septa
    - Usually found near surgical clips in cystectomy-node dissection site
    - No peripheral fat stranding
  - Abscess
    - Thick/irregular wall, septation, internal debris, air, peripheral fat stranding, wall enhancement
  - Urinary obstruction
    - Stricture at ureteroenteric anastomosis most commonly
    - Due to ischemia (more common) or recurrent tumor
    - May require surgical revision
  - Metabolic acidosis
    - Occurs to some degree in all continent bladder replacement surgeries
    - Due to absorption of urea and ammonia by mucosa lining reservoir
  - Fistula
    - Entero-urinary or enterocutaneous
    - Seen in ~ 2% of diversion cases
      - Higher rate when cystectomy was followed by radiation therapy to pelvis
  - **Recurrence of tumor**
    - Late complication (months)
    - Occurs in 3-16% (higher in advanced stage of original tumor)
    - Delayed-phase CT or MR urography are paramount for surveillance in patients with history of urothelial carcinoma
    - Soft tissue mass, wall asymmetry, enlarged lymph nodes, distant metastasis
    - Differential diagnosis includes scar tissue and radiation fibrosis
  - Urinary tract infection
    - Stasis of urine in reservoir is associated with high prevalence of infection
    - Bacteria in urine (85%); pyelonephritis (15%)
  - Calculi
    - Upper tract calculi occur in 10% of patients with urinary diversion

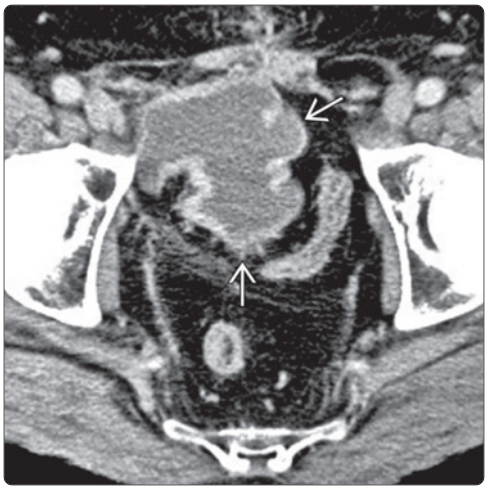
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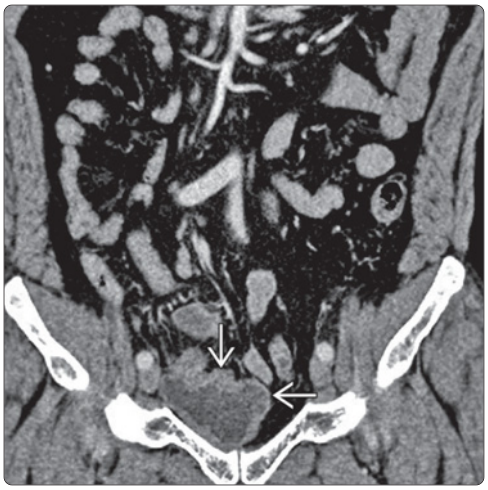
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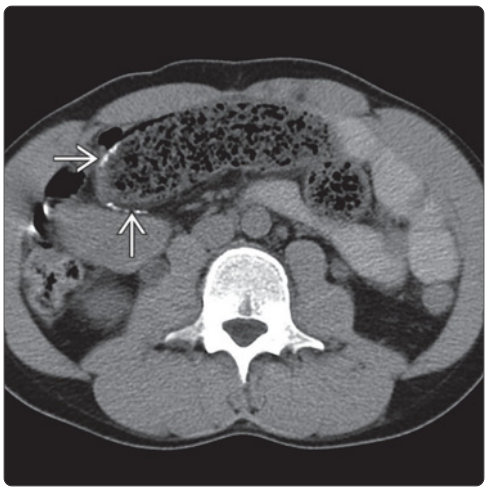


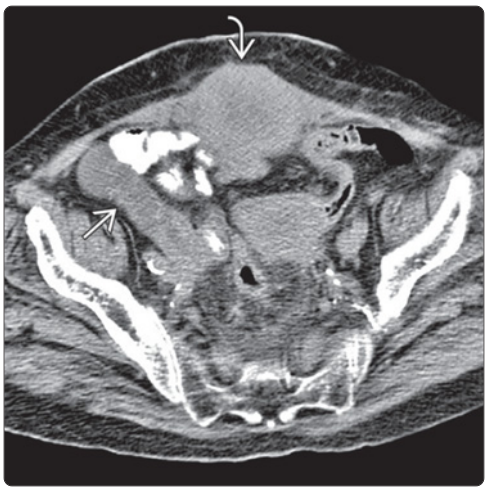
(Left) *Axial MIP CT in patient with history of a neurogenic bladder shows a continent ileal conduit that utilizes the appendix (Mitrofanoff technique). The appendiceal conduit opens to the umbilicus and is catheterized.* **(Right)** *Coronal NECT in a patient with history of psoas-hitch ureteroneocystostomy shows the bladder mobilized and sutured to the left psoas muscle, resulting in asymmetric appearance of bladder dome. The patient was post distal ureteral resection for retroperitoneal fibrosis.*





(Left) *Axial CECT shows a "neobladder" constructed from an ileal segment that was anastomosed to the ureters and urethra. This is an example of orthotopic bladder replacement (Studer technique).* **(Right)** *Coronal CECT in the same patient shows the "neobladder" constructed from an ileal segment that was anastomosed to the ureters and urethra. On other sections, the surgical staple line was seen at the site from which the ileal segment was harvested.*





(Left) *Axial NECT in patient with history of ileal conduit formation shows dilatation of ileum and small bowel feces sign reflective of chronic bowel obstruction, presumably related to a stricture at the anastomosis site.* **(Right)** *Axial NECT in a patient with history of radical cystectomy and ileal conduit diversion for bladder cancer shows a soft tissue attenuation mass that represents recurrent carcinoma.*

Mesenchymal Bladder Neoplasms

KEY FACTS

TERMINOLOGY

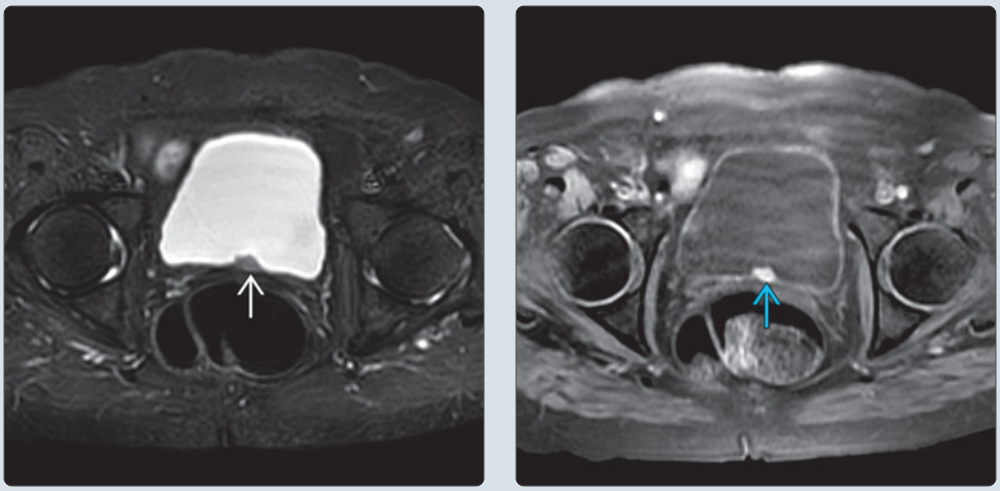
- Primary neoplasms of urinary bladder deriving from nonepithelial elements
- Mesenchymal bladder tumors are rare; altogether account for < 5% of bladder tumors

IMAGING

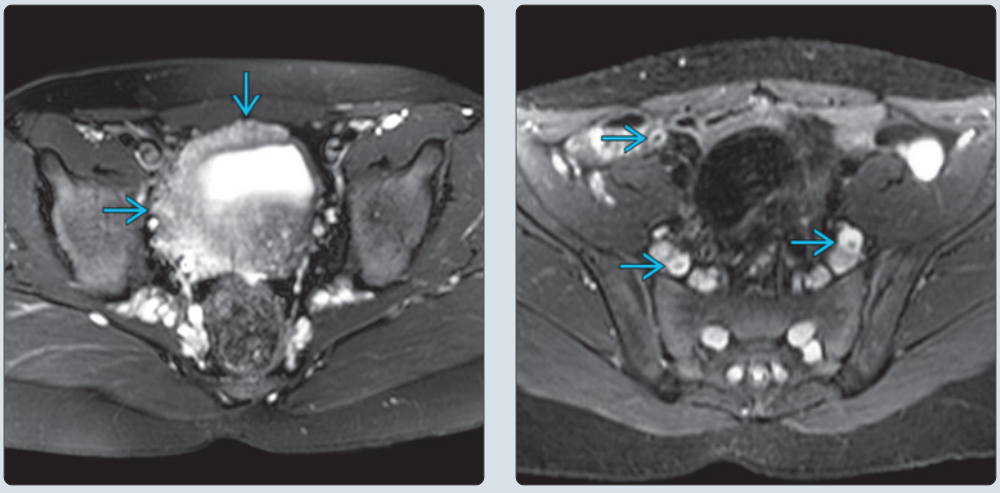
- **Leiomyoma**
  - Most common mesenchymal tumor showing smooth muscle differentiation
  - Soft tissue density intramural lesion with avid enhancement; larger lesions tend to be heterogeneous with nonuniform enhancement
  - Low/intermediate signal on T1W and **low signal on T2W MR;** areas of degeneration show ↑ T2 signal and do not enhance
- **Paraganglioma**
  - Round or oval-shaped soft tissue density mass with broad base
  - Usually have ↑ T1 and intermediate T2 signal on MR
  - Majority of reported cases are symptomatic and functional
- **Neurofibroma**
  - Nodular bladder wall thickening with low density (due to myxoid matrix) and nonuniform enhancement
  - Low-to-intermediate signal on T1W MR and ↑ signal (**target sign**) on T2W; + enhancement
- **Hemangioma/Vascular Malformation**
  - Solitary, small (< 3 cm), intramural mass bulging into bladder lumen
  - Highly vascular and enhancing (similar to hemangioma and vascular malformation in other parts)

TOP DIFFERENTIAL DIAGNOSES

- Bladder urothelial carcinoma
  - Imaging findings may overlap
- Bladder metastasis



(Left) *Axial fat-suppressed STIR MR in a 68-year-old woman with a history of hematuria shows a broad-base filling defect ➡ at the midline bladder trigone. The lesion has intermediate T2 signal.* **(Right)** *Axial T1W MR in the same patient demonstrates inherent T1 hyperintensity of the lesion ⇨. The patient underwent partial cystectomy and the pathology was consistent with paraganglioma.*



(Left) *Axial T2W MR with fat saturation shows marked nodular bladder wall thickening ⇨ in this 9-year-old boy with a history of neurofibromatosis type 1. Note additional plexiform neurofibromas along the pelvic sidewalls and in the perirectal region.* **(Right)** *Axial T2W with inversion recovery (STIR) in the same patient shows the target sign in multiple neurofibromas ⇨. The target appearance is due to central fibrosis surrounded by T2 hyperintense myxoid stroma.*

TERMINOLOGY

Definitions

- Primary neoplasms of urinary bladder derived from nonepithelial elements
  - Leiomyoma, paraganglioma, fibroma, hemangioma, neurofibroma, plasmacytoma
  - Malignant counterparts (sarcomas) are exceedingly rare
- Mesenchymal bladder tumors are rare; altogether account for < 5% of bladder tumors

IMAGING

Leiomyoma

- **Definition**: Most common mesenchymal tumor showing smooth muscle differentiation
- **Demographics**: Accounts for < 0.5% of bladder tumors
  - Mean age at detection: 44 (range: 11-75); some reports showed slight female predilection
- **Imaging findings**
  - Intramural mass arising in submucosa or muscularis propria (detrusor muscle); may show intraluminal (65%) or extravesical (30%) growth
  - **Cystography**: Smooth surface; makes obtuse angle with bladder lumen (similar to any other intramural lesions)
  - **US**: Usually hypoechoic; larger lesions tend to be heterogeneous, due to degeneration
  - **CT**: Soft tissue density intramural lesion with avid enhancement; larger lesions tend to be heterogeneous with nonuniform enhancement (also due to degeneration)
  - **MR:** Low/intermediate signal on T1W and **low signal on T2W**
    - Smaller lesions are avidly enhancing; larger lesions tend to be heterogeneous with nonuniform enhancement
    - Areas of degeneration show ↑ T2 signal and do not enhance
- **Clinical presentation**: Most cases are small, asymptomatic, and discovered serendipitously
  - Larger tumors present with obstructive symptoms, irritative symptoms, or hematuria
- **Microscopic features**: Interlacing fascicles of spindle cells with prominent eosinophilic cytoplasm
  - Covered by intact urothelium
- **Treatment**: Histologic examination is required to differentiate it from well-differentiated leiomyosarcoma or other mesenchymal tumors
  - Smaller lesions can be treated with transurethral resection (TURBT); larger tumors may require partial cystectomy
  - Preoperative suspicion is important to avoid radical surgery

Paraganglioma

- **Definition**: Exceedingly rare tumor arising from chromaffin cells of bladder sympathetic system
  - Also known as bladder pheochromocytoma
- **Demographics**: Accounts for < 0.5% of bladder tumors
  - ~ 6% of extraadrenal paragangliomas occur in bladder
  - ~ 10% have malignant behavior (i.e., metastasis)
  - Mean age at detection: 43 (range: 11-84); no gender predilection
- **Imaging findings**
  - Usually along posterior or lateral wall of urinary bladder
  - Mean tumors size: 3.9 cm (range: 1-9 cm)
  - **Cystography**: Broad-based intravesical filling defect
  - **US**: Well-defined, immobile echogenic mass with broad-base attachment to bladder wall
    - Findings are nonspecific and indistinguishable from papillary urothelial carcinoma
    - Color Doppler useful to document vascularity
  - **CT**: Round or oval-shaped soft tissue density mass with broad base
    - Typically has avid homogeneous enhancement
      - Large, malignant lesions are more heterogeneous due to tumoral necrosis
    - CT urography augments sensitivity for depiction of small, broad base lesions
  - **MR**: Usually have ↑T1 and intermediate T2 signal
    - Intense enhancement
      - Subtraction images might be necessary for detection of enhancement, since lesions are usually inherently T1 hyperintense
    - Restricted diffusion
      - Wang et al (EJR 2011) reported mean ADC value of 973 x 10^{-6} mm^2/s (range 723-1110 x 10^{-6} mm^2/s)
  - **Scintigraphy**: Metaiodobenzylguanidine (MIBG) and somatostatin receptor scintigraphy may show radiotracer uptake by tumor
    - Sensitivity of these tests for detection of bladder paraganglioma is less than for adrenal pheochromocytoma
- **Microscopic features**: Nest-like cluster of uniform, round-to-polygonal cells surrounded by delicate richly vascular tissue and sustentacular cells (**Zellballen**)
  - Surface urothelium remains intact
  - Express neuroendocrine markers (such as synaptophysin, chromogranin-A, and CD56)
- **Clinical presentation**: According to a large meta-analysis (Beilan et al 2013), majority of reported cases were symptomatic and majority of tumors were functional
  - Hypertension (55%), micturition attacks (53%), headache (48%), hematuria (47%)
    - Voiding, bladder distention, defecation, or sexual intercourse can trigger release of catecholamines
  - ↑ urinary/serum VMA, metanephrine, catecholamines
  - With widespread use of cross-sectional imaging more cases are detected serendipitously and at early stage
    - Many of these incidental cases are asymptomatic and nonfunctional, likely due to their small size
- Classified into localized, regional, and metastatic forms (NCI classification)
  - Features associated with malignancy
    - Large tumor size, extensive tumor necrosis
    - ↑ number of mitoses, DNA aneuploidy
- ~ 25% of paragangliomas occur in setting of hereditary syndromes
  - MEN type 2, NF type 1, vHL disease
  - Hereditary paraganglioma syndrome: Caused by mutation in gene encoding succinate dehydrogenase (SDH) enzyme

- **Treatment**
  - Partial or radical cystectomy are treatment of choice
    - Due to nonepithelial intramural nature of tumor, recurrence rate after transurethral resection of bladder tumor (TURBT) is high
  - Pelvic lymph node dissection is reserved for cases with regional and metastatic disease
  - Tissue biopsy should be avoided (due to risk of severe hypertension) unless no biochemical evidence of catecholamine hypersecretion exists or patient has been alpha-blockaded

Neurofibroma

- **Definition**: Nerve sheath tumor arising from vesical/perivesical neural plexus
  - May be isolated or, more commonly, in association with neurofibromatosis type 1 (NF-1)
    - Localized, diffuse, or plexiform subtypes; plexiform type more common
  - Intramural process resulting in marked bladder wall thickening; overlying urothelium is usually intact
- **Imaging findings**
  - Diffuse, nodular bladder wall thickening
  - Plexiform neurofibromas may also extend to adjacent prostate, seminal vesicles, uterus, vagina, and pelvic sidewalls
  - **CT**: Nodular bladder wall thickening with low density (due to myxoid matrix) and nonuniform enhancement
  - **MR**: Low-to-intermediate signal on T1W and ↑ signal on T2W (**target sign)**; + enhancement
    - Target sign: ↓ intensity central dot (fibrosis) surrounded by ↑ intensity myxoid stroma
- **Treatment:** Conservative management unless intractable symptoms or suspicion for malignant degeneration (such as rapid increase in size of tumor)
  - Radical cystectomy is common surgical technique

Hemangioma/Vascular Malformation

- May occur in isolation or in association with Sturge-Weber or Kleppel-Trénaunay-Weber syndromes
  - Majority of lesions are vascular malformations (no endothelial proliferation)
- Seen in both adult and pediatric populations; mean age in adults: 58 years (range: 19-76); more common in men
- Arise in submucosal layer and bulge into bladder lumen; overlying urothelium is intact
- Seen as lobulated bluish red lesion on cystoscopy
- **Imaging findings**
  - Solitary, small (< 3 cm), intramural mass bulging into bladder lumen; diffuse and multifocal forms are rare
  - Highly vascular and enhancing (similar to hemangioma and vascular malformation in other parts)
  - Most lesions have ↓ T1 and ↑ T2 signal on MR
- **Treatment**: Biopsy (to exclude other lesions) and fulguration; partial cystectomy might be performed for larger lesions

Sarcoma

- **Leiomyosarcoma** is most common malignant primary mesenchymal tumor in adults
  - Exceedingly rare; ↑ prevalence in patients with prior radiation or chemotherapy; more common in men (M:F=3:1)
  - Imaging features of well-differentiated leiomyosarcoma and leiomyoma overlap
  - Sarcomas are usually larger (mean diameter of 7 cm) and more heterogeneous at presentation
    - Usually present with hematuria or obstructive/irritative symptoms
  - Treatment: Radical cystectomy ± adjuvant chemoradiation
- **Rhabdomyosarcoma** is most common malignant bladder tumor in children
  - Exceedingly rare in adults
  - Mean age at presentation: 4 years; more common in boys (M:F=3:1)
  - Large heterogeneous bladder mass with common invasion into prostate
  - Usually has polypoid intraluminal growth resulting in grape-like appearance (sarcoma botryoides)
  - Treatment: Surgical resection &/or chemotherapy
- Other sarcoma (such as angiosarcoma, osteosarcoma, pleomorphic undifferentiated sarcoma, etc.) are extremely rare
  - Imaging findings are usually nonspecific and tissue biopsy is required to establish the diagnosis

DIFFERENTIAL DIAGNOSIS

Bladder Urothelial Carcinoma

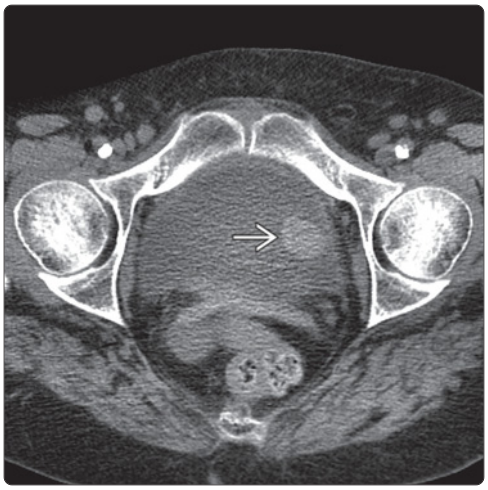
- Imaging findings of papillary urothelial carcinoma and mesenchymal tumors may overlap
- Mesenchymal tumors are intramural and overlying urothelium remains intact
  - Smooth surface and obtuse angles with bladder lumen are clues to diagnosis
- Several mesenchymal tumors have quasi-characteristic features on MR
  - Such as ↓ T2 signal in leiomyoma, ↑ T1 signal in paraganglioma, and target sign in neurofibroma

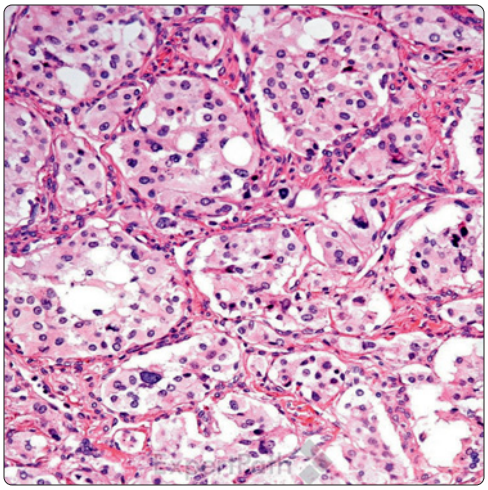
Bladder Metastasis

- Metastatic lesions from renal cell cancer and melanoma can present as hypervascular intramural lesions
- Clinical history and identification of additional metastases aid diagnosis

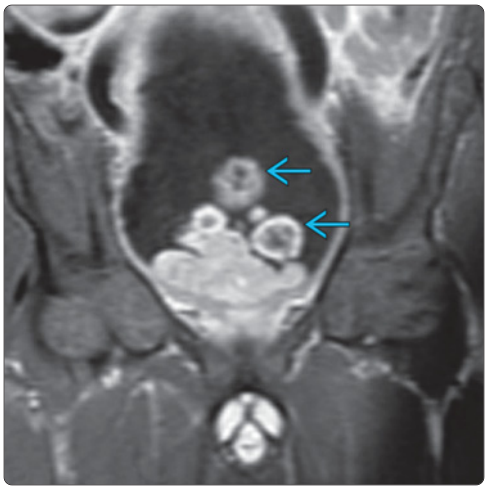
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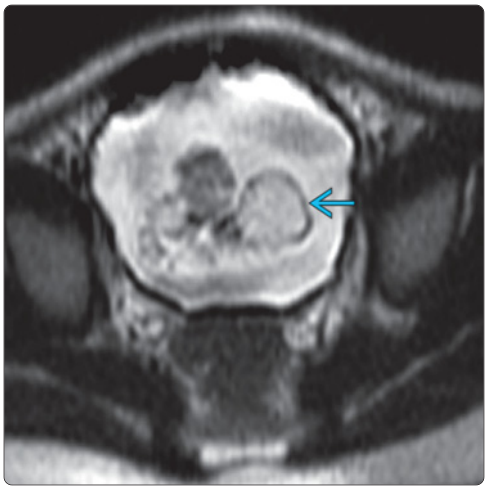
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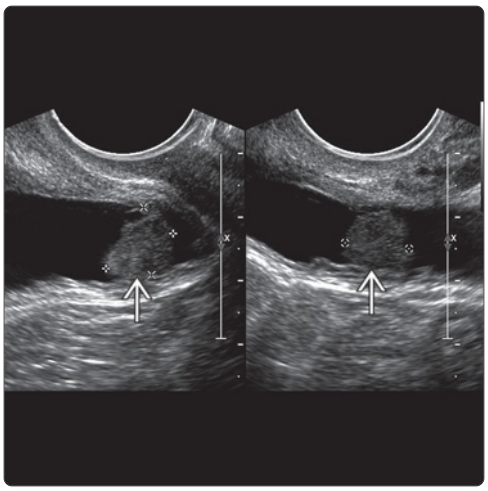


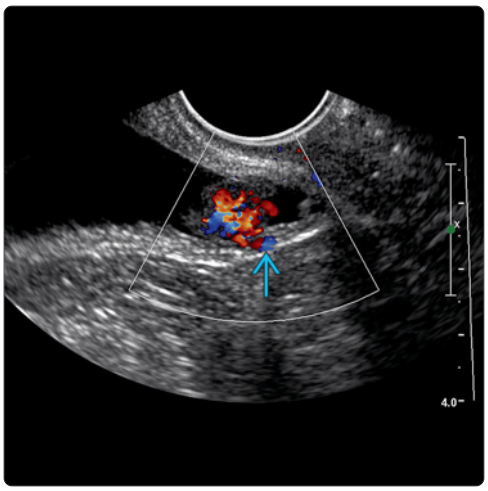
(Left) *Axial unenhanced CT in a 74-year-old man with no urinary symptoms shows an incidental pedunculated lesion ➡ along the left lateral bladder wall. The patient underwent transurethral resection of the tumor. Pathology was consistent with a paraganglioma.* **(Right)** *Photomicrograph with H&E staining shows classic nested, or Zellballen, architecture of paraganglioma.*





(Left) *Coronal post-contrast T1W MR in 3 year-old boy shows a large enhancing bladder mass with polypoid intraluminal growth ⇨, typical for bladder rhabdomyosarcoma (sarcoma botryoides).* **(Right)** *Axial T2W MR in the same patient shows high T2 intensity of the intraluminal polypoid projections ⇨, which is attributed to myxoid stroma.*





(Left) *Transverse and longitudinal US images through the bladder in a 66-year-old woman show a polypoid lesion ➡ at the inferior aspect of the bladder.* **(Right)** *Color Doppler shows that the lesion is vascular. Note the lesion's pedicle ⇨, which arises from posterior wall. The patient underwent transurethral resection of the tumor. Pathology was compatible with a bladder paraganglioma.*

Bladder Inflammatory Pseudotumor

KEY FACTS

TERMINOLOGY

- Polypoid bladder mass caused by nonneoplastic proliferation of myofibroblasts and inflammatory cells
- Depending on predominant inflammatory cell in lesion, it might have different names

IMAGING

- Polypoid mass projecting into lumen of bladder
- More common in bladder dome and posterior wall
  - Tends to spare trigone
- Ring enhancement and calcified surface may be seen

TOP DIFFERENTIAL DIAGNOSES

- Bladder urothelial carcinoma
- Bladder rhabdomyosarcoma
- Bladder myxoid leiomyosarcoma

PATHOLOGY

- Unclear etiology
  - May be secondary to inflammation following infection, minor trauma, surgery, or malignancy
  - Immune-mediated mechanism may play role

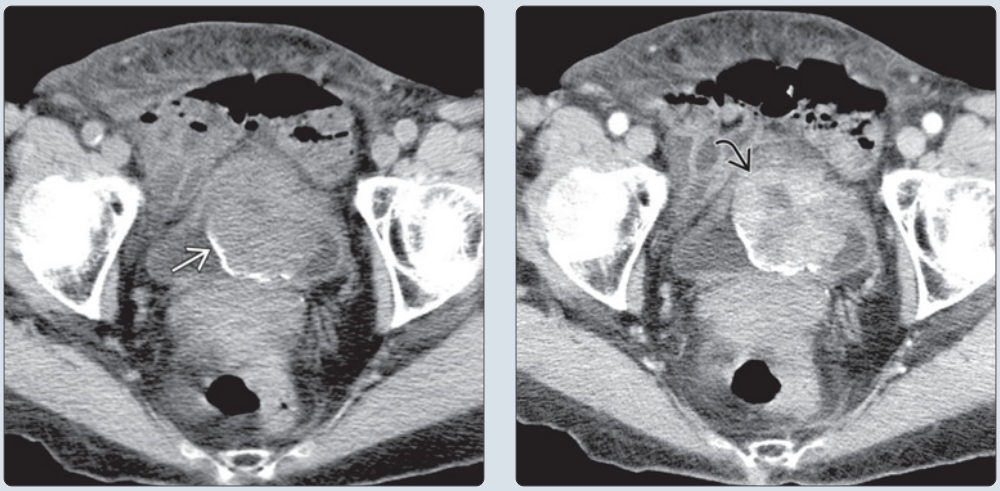
CLINICAL ISSUES

- Presenting symptoms: Painless hematuria, dysuria, frequency, lower abdominal pain
- More common in adults; mean age: 39 years
- Locally aggressive tumor; may invade through bladder wall
  - Local recurrence after resection is unusual (< 5%)
- Treatment
  - Surgical resection (transurethral resection of bladder tumor [TURBT] or partial cystectomy)
  - Medical management with long-term antibiotics &/or anti-inflammatory medications (steroidal and nonsteroidal)

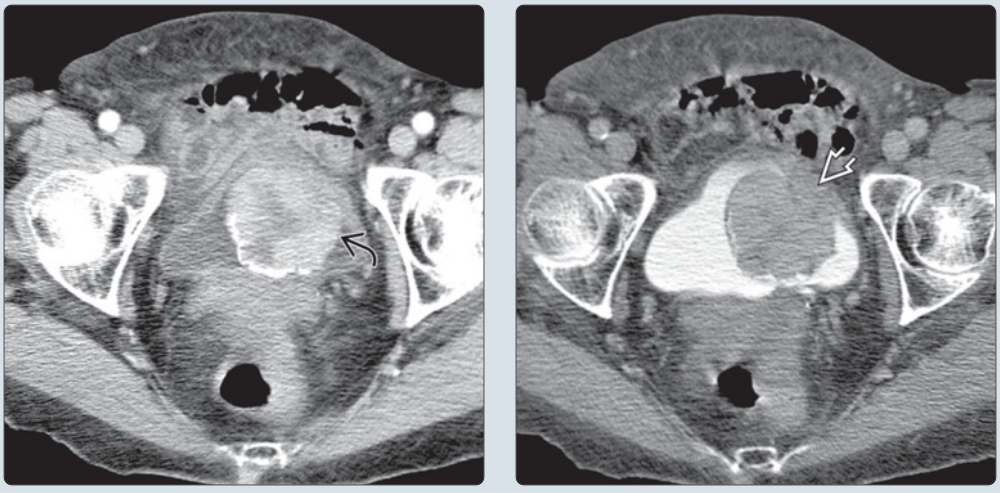
DIAGNOSTIC CHECKLIST

- Tissue biopsy is needed to establish diagnosis

(Left) *Axial NECT shows a large bladder mass with a partially calcified surface* ⇨ *in this 76-year-old woman with gross hematuria and pelvic pain.* **(Right)** *Axial CECT in the same patient shows brisk enhancement of the mass* ⇨. *The perivesical fat planes adjacent to the mass are indistinct, suggesting transmural extension of this process.*



(Left) *More caudal axial CECT section in the same patient shows brisk enhancement of the mass* ⇨ *with a central zone of necrosis.* **(Right)** *Delayed phase image CECT in the same patient shows the polypoid mass as a filling defect within the contrast-opacified urine. There is a broad base attachment to the bladder wall* ⇨. *At surgery, the mass was very hard and difficult to extract. The frozen section diagnosis favored sarcoma, but the final diagnosis was inflammatory pseudotumor of the bladder.*



TERMINOLOGY

Synonyms

- Inflammatory myofibroblastic tumor
- Pseudosarcomatous fibromyxoid tumor
- Plasma cell granuloma

Definitions

- Polypoid bladder mass caused by nonneoplastic proliferation of myofibroblasts and inflammatory cells
- Depending on predominant inflammatory cell in lesion, it might have different names

IMAGING

General Features

- Best diagnostic clue
  - Enhancing polypoid mass projecting into lumen of bladder
- Location
  - More common in bladder dome and posterior wall; tends to spare trigone
  - May invade through bladder wall
- Size
  - Mean size: 4.5 cm
- Morphology
  - Exophytic or polypoid; may be ulcerated
  - Solid and cystic variants may occur

CT Findings

- Polypoid mass projecting into lumen of bladder
- Heterogeneous attenuation and enhancement: Ring enhancement may be seen
- May have calcified surface

MR Findings

- T1WI
  - Heterogeneous, iso- to hypointense
- T2WI
  - Central hyperintense component correlating with central areas of necrosis
  - Peripheral hypointense component
- T1WI C+ FS
  - Heterogeneous enhancement; may show peripheral enhancement

DIFFERENTIAL DIAGNOSIS

Bladder Urothelial Carcinoma

- Indistinguishable from pseudotumor based on imaging

Bladder Rhabdomyosarcoma

- Indistinguishable from pseudotumor based on imaging
- Usually diagnosed in children

Bladder Myxoid Leiomyosarcoma

- Indistinguishable from pseudotumor based on imaging

PATHOLOGY

General Features

- Etiology
  - Unclear
  - May be secondary to inflammation following infection, minor trauma, surgery, or malignancy
  - Immune-mediated mechanism may play role
    - Inflammatory mediators (e.g., IL-1) → proliferation of fibroblasts/myofibroblasts

Gross Pathologic & Surgical Features

- Broad-based polypoid mass, edematous and ulcerated surface
- Luminal clot surrounding surface may be seen

Microscopic Features

- Acute and chronic inflammatory cells within fibrous background (vimentin-positive spindle cells)
  - May have predominantly myxoid, vascular, compact spindle cell, fibrous hypocellular, or mixed pattern
- Central areas of necrosis may be seen

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless hematuria
  - Dysuria, frequency
  - Lower abdominal pain

Demographics

- Age
  - More common in adults (mean age: 39 years)
- Gender
  - No gender predominance

Natural History & Prognosis

- Locally aggressive tumor
  - May invade through bladder wall
- Local recurrence after resection is unusual (< 5%)

Treatment

- Surgical resection
  - Transurethral resection of bladder tumor (TURBT)
  - Partial cystectomy
- Medical management with long-term antibiotics &/or anti-inflammatory medications (steroidal and nonsteroidal)

DIAGNOSTIC CHECKLIST

Consider

- Impossible to distinguish from bladder cancer based on imaging
- Tissue biopsy is needed to establish diagnosis

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Bladder and Ureteral Intramural Masses

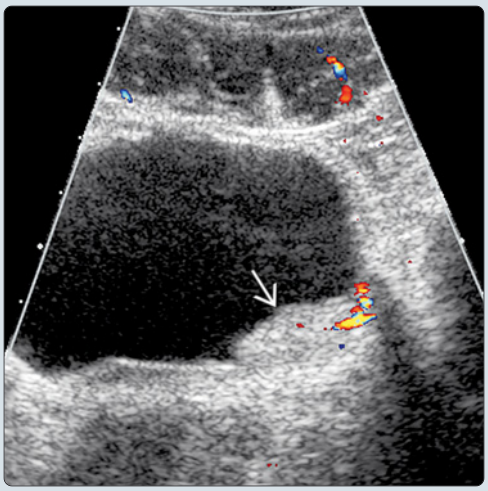
KEY FACTS

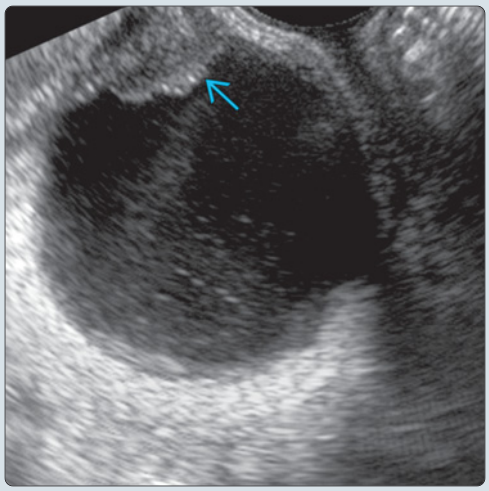
IMAGING

- Mass centered in bladder wall causing impression on bladder lumen; makes obtuse angles with bladder lumen
- Usually have both intravesical and extravesical growth and makes smooth interface with bladder lumen

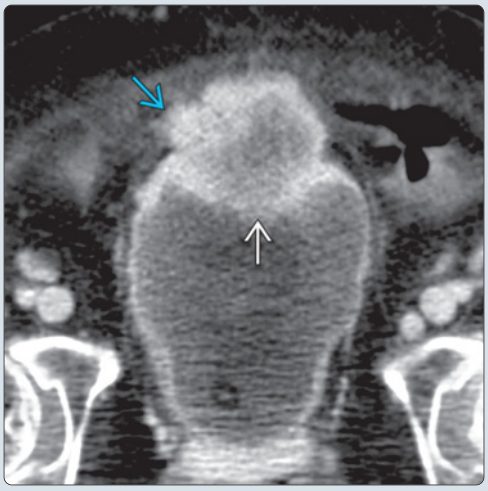
TOP DIFFERENTIAL DIAGNOSES

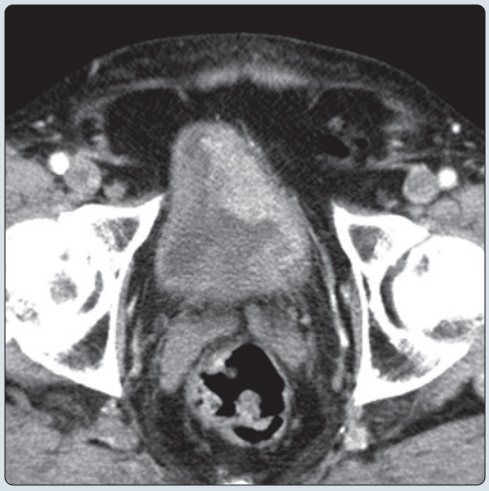
- **Leiomyoma**
  - **Most common** mesenchymal tumor of bladder
  - Low signal on T1W and T2W MR; shows different degrees on enhancement
  - Areas of cystic degeneration are T2 hyperintense
- **Leiomyosarcoma**
  - **Most common malignant** mesenchymal tumor of bladder
  - Imaging findings overlap with leiomyoma; usually larger and more heterogeneous containing areas of necrosis
- **Rhabdomyosarcoma**
  - Most common bladder tumor in pediatric population
- **Neurofibroma**
  - Typically low signal on T1W MR with **target sign** on T2W MR
- **Paraganglioma**
  - Show high T1 signal and restricted diffusion on MR
- **Lymphoma**
  - Most often secondary involvement rather than primary
- **Endometriosis**
  - Almost always along the posterior aspect of bladder (dependent) and inseparable from adjacent uterus
- **Metastasis**
  - Melanoma, breast, and gastric adenocarcinoma are most frequent primaries
- **Extrinsic masses (mimic)**
  - External compression on bladder can mimic intramural process on antegrade/retrograde cystography and on cystoscopy





(Left) *Longitudinal color Doppler US of the urinary bladder in a 64-year-old woman with symptoms of cystitis reveals a smooth isoechoic intramural mass in the bladder base and no significant flow. Endoscopic biopsy proved this to be a leiomyoma.* **(Right)** *Longitudinal transvaginal grayscale US of the bladder in a 72-year-old woman with frequent urination shows a hypoechoic mass in the bladder dome with obtuse angles to the lumen. Endoscopic biopsy confirmed a leiomyoma.*





(Left) *Axial CECT of a distended urinary bladder in a 48-year-old woman with pelvic pain shows a heterogeneous soft tissue mass infiltrating the bladder wall; note the extramural tumor invasion into surrounding perivesicular fat. Endoscopic biopsy identified the tumor as a leiomyosarcoma.* **(Right)** *Axial CECT of a distended urinary bladder reveals an infiltrating soft tissue mass involving the lateral wall of the bladder, identified by an endoscopic biopsy as a B-cell lymphomatous tumor.*

TERMINOLOGY

Definitions

- Primary or metastatic tumors centered within bladder wall involving either lamina propria, muscularis propria, or adventitial layers ± intravesical growth

IMAGING

General Features

- Best diagnostic clue
  - Mass centered in bladder wall causing impression on bladder lumen
  - Makes obtuse angles with bladder lumen
- Location
  - Differs based on pathology
  - Endometriosis is almost always in posterior wall (dependent portion) along uterovesical recess
- Size
  - Variable, 1-10 cm
- Morphology
  - Usually have both intravesical and extravesical growth and makes smooth interface with bladder lumen

Imaging Recommendations

- Best imaging tool
  - CECT with delayed phase; MR
- Protocol advice
  - Hydrate patient to distend bladder, scan with IV contrast

Radiographic Findings

- Antegrade/retrograde cystography
  - Smooth-surface filling defect
  - Impression on bladder lumen making obtuse angles with bladder lumen

CT Findings

- Intramural soft tissue mass with variable enhancement

MR Findings

- MR allows better delineation of bladder wall layers and helps to better localize lesion
- Signal characteristics differ based on pathology and are often nonspecific
  - Leiomyoma/leiomyosarcoma have low signal on both T1W and T2W (except for areas of cystic degeneration and necrosis, which show intermediate-to-high T2 signal)
  - Endometriosis shows high T1 signal (due to blood by-products)
  - Neurofibroma may show classic target sign on T2W MR (described below)

Ultrasonographic Findings

- Isoechoic or hypoechoic soft tissue mass involving bladder wall

DIFFERENTIAL DIAGNOSIS

Leiomyoma

- **Most common** mesenchymal tumor of bladder
- Most are small and found incidentally
  - Large tumors can present with irritative urinary symptoms, hematuria, or obstruction
- Tumor originates in muscular layer and shows extravesical &/or intraluminal growth
- Smaller tumors are homogeneous; large tumors are more likely to be heterogeneous due to cystic degeneration
- **Low signal** on T1W and T2W MR; shows different degrees of enhancement
  - Areas of cystic degeneration are T2 hyperintense
- Benign tumor with no malignant potential
- Treatment: Focal excision

Leiomyosarcoma

- **Most common malignant** mesenchymal tumor of bladder
- Rare tumor; more common in older men
- Associated with prior radiation or cyclophosphamide treatment
- 80% are high grade; 5-year survival rate is ~ 60%
- Imaging findings overlap with leiomyoma; usually larger and more heterogeneous containing areas of necrosis
- Treatment: Radical cystectomy ± chemoradiation

Rhabdomyosarcoma

- Arises from primitive muscle cells in bladder and prostate
- Most common bladder tumor in pediatric population
  - 3x more common in Caucasian children than African American children
- Presents with hematuria, dysuria, or urinary infection
- Grape-like polypoid masses from sarcoma botryoides variant

Neurofibroma

- Rare; bladder is most common genitourinary site
- May be isolated or associated with neurofibromatosis type 1
- Lesions arise from intramural neural plexus near bladder trigone
- Neurofibromas may be focal, plexiform, or diffuse
  - May result in diffuse nodular thickening of bladder wall
- Hypodense on NECT; shows heterogeneous enhancement
- Typically low signal on T1W MR with **target sign** on T2W MR
  - Low signal (fibrosis) surrounded by high signal myxoid stroma
  - Stromal elements avidly enhance on contrast-enhanced MR images

Paraganglioma

- Rare intramural/subepithelial tumor arising from chromaffin cells of bladder sympathetic system
  - 1% of all pheochromocytomas
- Purely intramural tumors present as indentation on bladder wall with obtuse angle (similar to other intramural tumors)
- Majority of cases, however, show intraluminal growth as well and may be indistinguishable from papillary urothelial carcinoma on imaging
- Avid homogeneous enhancement (similar to paragangliomas in other body regions)
- High T1 signal and restricted diffusion on MR
- Majority of cases are symptomatic and functional

Lymphoma

- Most often secondary involvement rather than primary
- Occurs in 10-25% of all lymphoma patients

- Typically low-grade B-cell or mucosa-associated lymphoid tissue (MALT) tumor
- Well-defined masses in bladder wall
- Good prognosis with either chemotherapy or radiation

Endometriosis

- Bladder involvement in endometriosis is rare
- Caused by direct implantation of endometrium or following pelvic surgery
- Endometrial implantation typically occur at uterovesical pouch and the mass grows through detrusor muscle into submucosa
- Almost always along posterior aspect of bladder (dependent) and inseparable from adjacent uterus
- Imaging features are nonspecific; presents as indentation along posterior bladder wall with obtuse angles (similar to other intramural processes)
  - Less commonly, endometriosis can grow through epithelium and present as polypoid intraluminal mass
- Avid enhancement (usually heterogeneous due to presence of hemorrhagic and fibrotic foci)
- MR shows areas of hemorrhage (high signal on T1W MR)

Metastasis

- Rare
- Melanoma, breast, and gastric adenocarcinoma are most frequent primaries
- Usually manifest as focal lesions
  - May also present as diffuse bladder wall thickening; differentiation from benign causes of cystitis may be very difficult based on imaging alone
- Usually intramural/submucosal with intravesical growth

Extrinsic Masses (Mimic)

- External compression on bladder can mimic intramural process on antegrade/retrograde cystography and on cystoscopy
- Invasive infiltrative processes, such as rectal/genitourinary malignancies and endometriosis, start extrinsic to bladder and subsequently extend into bladder wall
- US, CT, and MR can differentiate between extrinsic and intramural processes

PATHOLOGY

General Features

- Genetics
  - Paragangliomas and neurofibromas may be associated with familial syndromes

Staging, Grading, & Classification

- Relate to depth of mural invasion, nodal and distant metastasis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Small intramural lesions are usually asymptomatic and incidentally found
  - Larger lesions can present with hematuria, irritative symptoms, or obstruction
  - Bladder paraganglioma can be functional

Demographics

- Epidemiology
  - Intramural bladder tumors are rare
    - < 5% of all bladder tumors

Treatment

- Depends on histology of lesion
  - Leiomyoma: Surgical excision
  - Lymphoma: Chemotherapy and radiation
  - Rhabdomyosarcoma: Surgery, chemotherapy, and radiation
  - Paraganglioma: Surgery

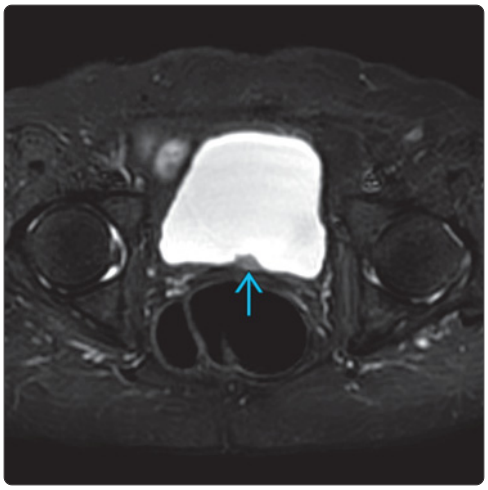
DIAGNOSTIC CHECKLIST

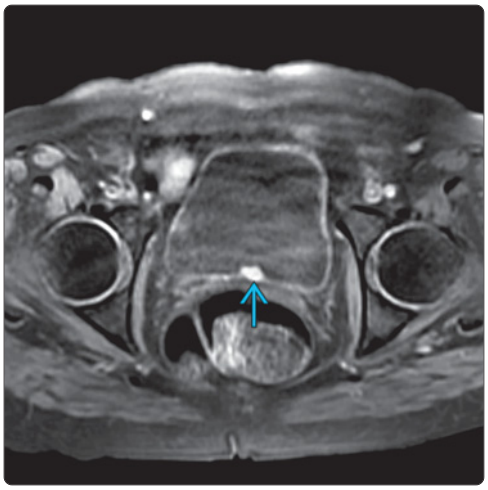
Image Interpretation Pearls

- Overlying mucosa is usually intact, mass has obtuse angles with bladder lumen

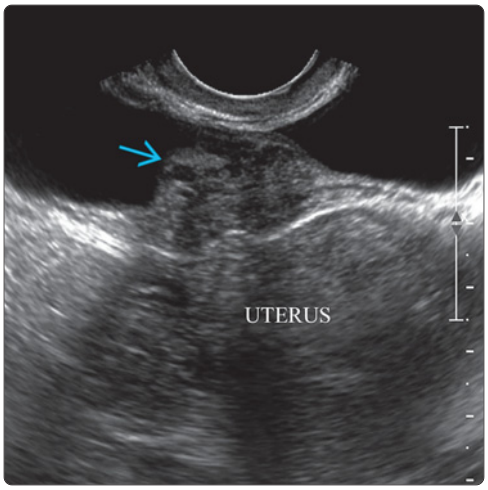
SELECTED REFERENCES

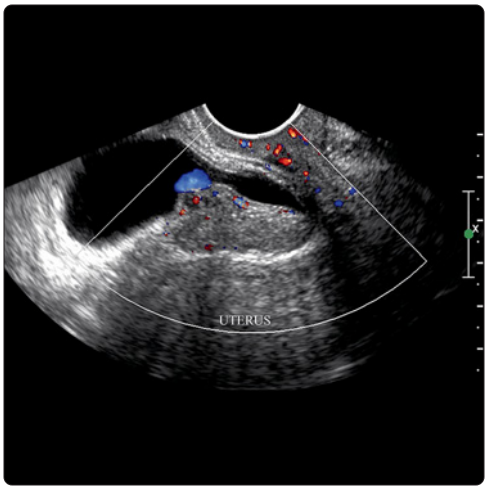
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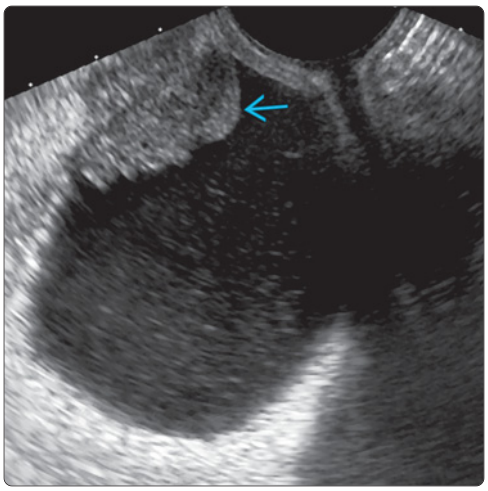


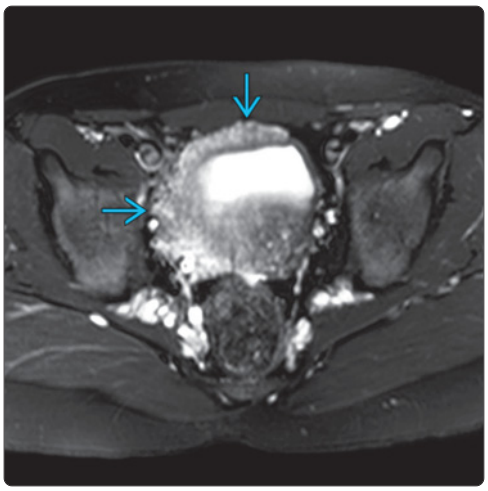
(Left) *Axial fat-suppressed STIR MR, in 68-year-old woman with a history of hematuria shows a T2-intermediate, broad-base filling defect ⇨ with obtuse angles to the bladder.* **(Right)** *Axial T1W MR, in the same patient, demonstrates inherent T1 hyperintensity of the lesion ⇨. The patient underwent partial cystectomy and the pathology result was consistent with a paraganglioma.*





(Left) *Transverse US through the bladder in a 45-year-old woman shows a heterogeneous, hypoechoic intramural mass ⇨ along the posterior bladder wall (centered within uterovesical recess).* **(Right)** *Longitudinal color Doppler US, in the same patient, better shows the relationship between the mass, uterus, and bladder. Note the smooth surface of the mass and the obtuse angle formed between the mass and bladder wall. The mass was confirmed to be endometriosis.*





(Left) *Longitudinal transvaginal grayscale US in a 65-year-old woman who presented with symptoms of cystitis demonstrates a hypoechoic mass ⇨ near the dome of the bladder. Cystoscopic biopsy confirmed this to be a leiomyoma.* **(Right)** *Axial T2W MR with fat suppression shows marked nodular bladder wall thickening ⇨ in this 9-year-old boy with a history of neurofibromatosis type 1. Note the additional plexiform neurofibromas along pelvic sidewalls and in perirectal space.*

T | Primary Tumor

TNM	Definitions
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Ta	Noninvasive papillary carcinoma
Tis	Carcinoma in situ: "Flat tumor"
T1	Tumor invades subepithelial connective tissue
T2	Tumor invades muscularis propria
pT2a	Tumor invades superficial muscularis propria (inner half)
pT2b	Tumor invades deep muscularis propria (outer half)
T3	Tumor invades perivesical tissue
pT3a	Tumor invades perivesical tissue microscopically
pT3b	Tumor invades perivesical tissue macroscopically (extravesical mass)
T4	Tumor invades any of the following: Prostatic stroma, uterus, vagina, pelvic wall, abdominal wall
T4a	Tumor invades prostatic stroma, uterus, vagina
T4b	Tumor invades pelvic wall, abdominal wall

N | Regional Lymph Nodes

TNM	Definitions
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Single regional lymph node metastasis in true pelvis (hypogastric, obturator, external iliac, or presacral lymph node)
N2	Multiple regional lymph node metastases in true pelvis (hypogastric, obturator, external iliac, or presacral lymph node)
N3	Lymph node metastasis to common iliac lymph nodes

Regional lymph nodes include both primary and secondary drainage regions. All other nodes above the aortic bifurcation are considered distant lymph nodes.

M | Distant Metastasis

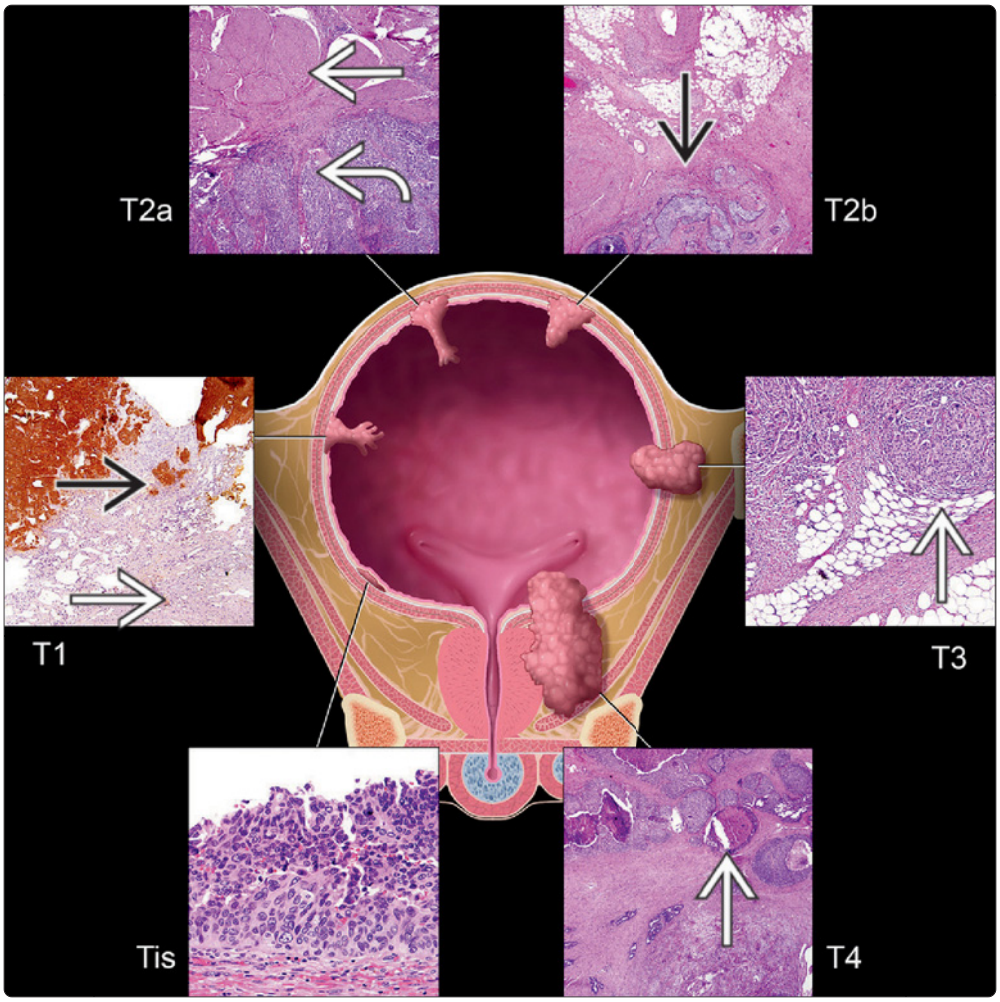
TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis

AJCC | Stages/Prognostic Groups

Stage	T	N	M
0a	Ta	N0	M0
0is	Tis	N0	M0
I	T1	N0	M0
II	T2a	N0	M0
	T2b	N0	M0
III	T3a	N0	M0
	T3b	N0	M0
	T4a	N0	M0
IV	T4b	N0	M0
	Any T	N1-3	M0
	Any T	Any N	M1

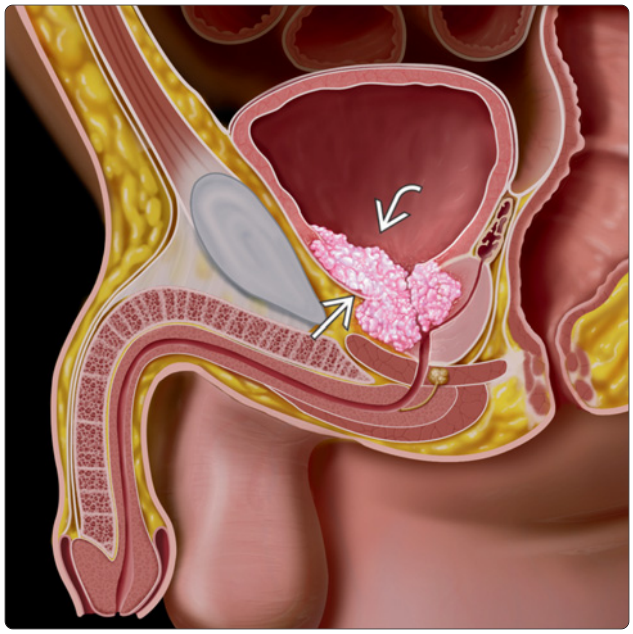
Adapted from 7th edition AJCC Staging Forms (2010).

T Staging of Urinary Bladder Carcinoma



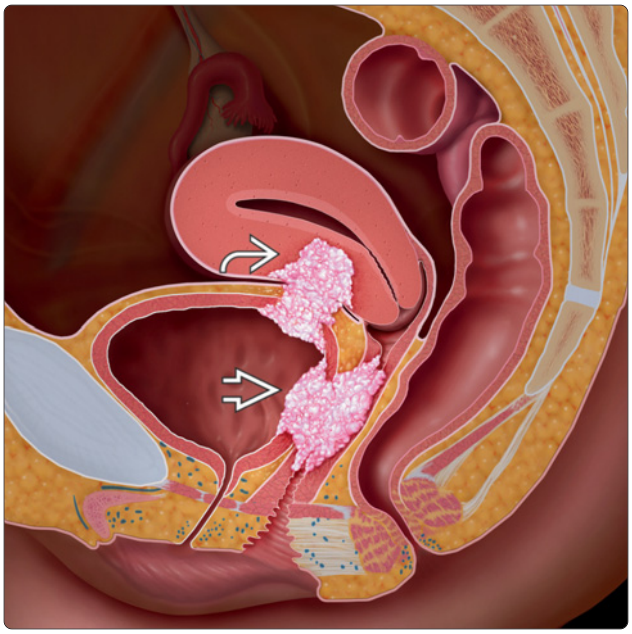
Graphics show T stages of urinary bladder carcinoma with histologic correlation. Tis or carcinoma in situ refers to nonpapillary (flat) mucosa in which the normal urothelium has been replaced by cancer cells that have not invaded through the basement membrane. Carcinoma in situ is a high-grade lesion and essentially recognized because of cytologic abnormalities similar to those noted in high-grade papillary tumors. Neoplastic cells are pleomorphic, hyperchromatic, and occupy a portion of the thickness of the urothelium. They lack polarity in relation to the basement membrane. (Original magnification 400x.) T1 describes urothelial tumor invasion through the basement membrane into the subepithelial connective tissue. In this photomicrograph, cytokeratin immunohistochemical stain is used to highlight the tumor ⇨ invading into subepithelial connective tissue but not to muscularis propria ➡. (Original magnification 200x.) T2 designates a tumor that invades muscularis propria. T2a: H&E stain shows tumor cell invading the superficial/inner 1/2 of muscularis propria ➡. Note that outer 1/2 of muscularis propria is not involved ➡. (Original magnification 100x.) T2b: H&E stain shows tumor cells ⇨ invading the outer 1/2 of the muscularis propria. (Original magnification 100x.) T3 applies to a tumor invading perivesical tissue. H&E stain shows tumor cells ➡ invading perivesical fat tissue. (Original magnification 200x.) Tumor is considered T3a if perivesical fat involvement is microscopic (not evident by imaging) and T3b if macroscopic (potentially detected by imaging). T4 describes a tumor invading any of the following: Prostate, uterus, vagina, pelvic wall, or abdominal wall. It is T4a when the tumor invades prostate, uterus, and vagina; tumor is T4b when it invades pelvic wall and abdominal wall. H&E stain demonstrates neoplastic transitional cell carcinoma cells ➡ invading prostate. (Original magnification 100x.)

T4a: Male



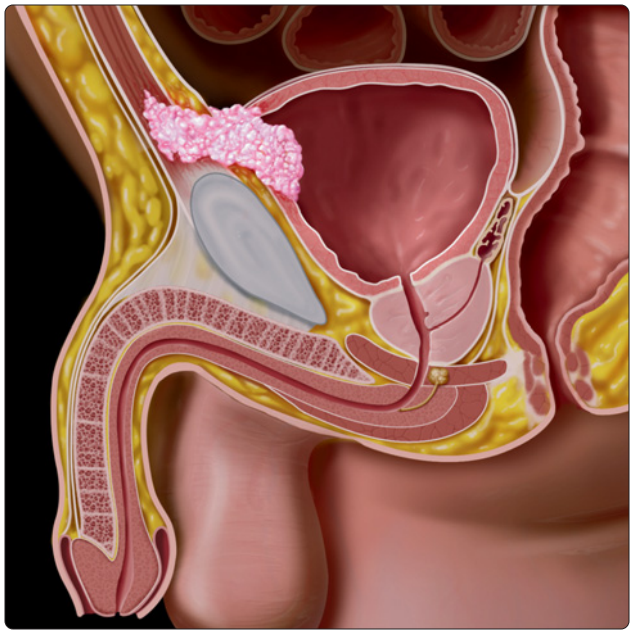
Graphic shows a bladder neck tumor in a male patient; the tumor invades the prostate, constituting stage T4a. To be considered T4a disease, the tumor should have prostatic stromal invasion directly from the bladder tumor. Subepithelial invasion of prostatic urethra will not constitute T4 staging status.

T4a: Female



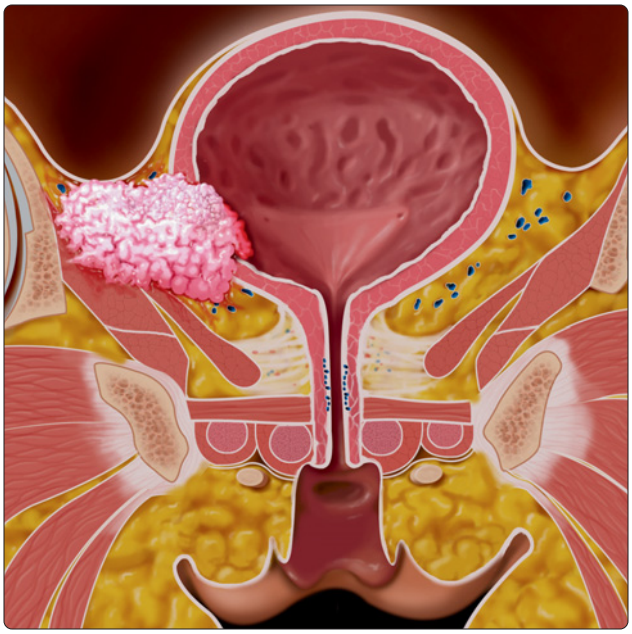
Graphic shows posterior wall bladder tumors in a female patient invading the uterus and vagina. Invasion of the uterus &/or vagina constitutes stage T4a.

T4b: Male



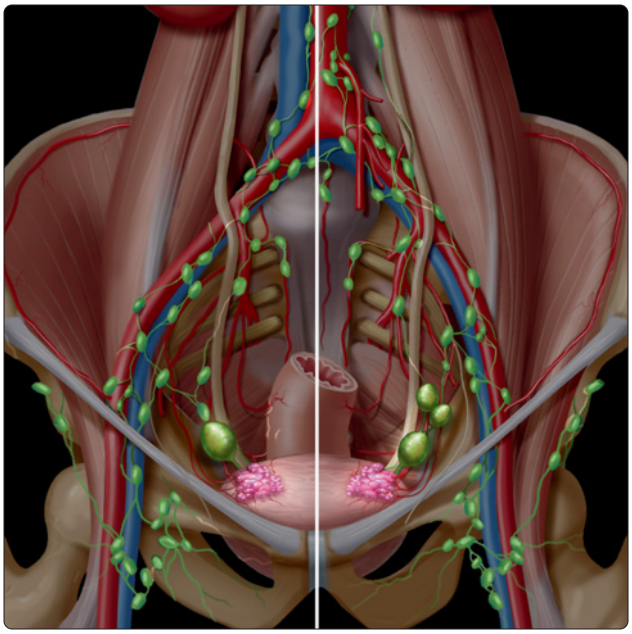
Graphic shows a tumor of the anterior wall of the urinary bladder that invades into the extraperitoneal prevesical fat (space of Retzius) and eventually the muscles of the anterior abdominal wall, constituting stage T4b.

T4b: Female



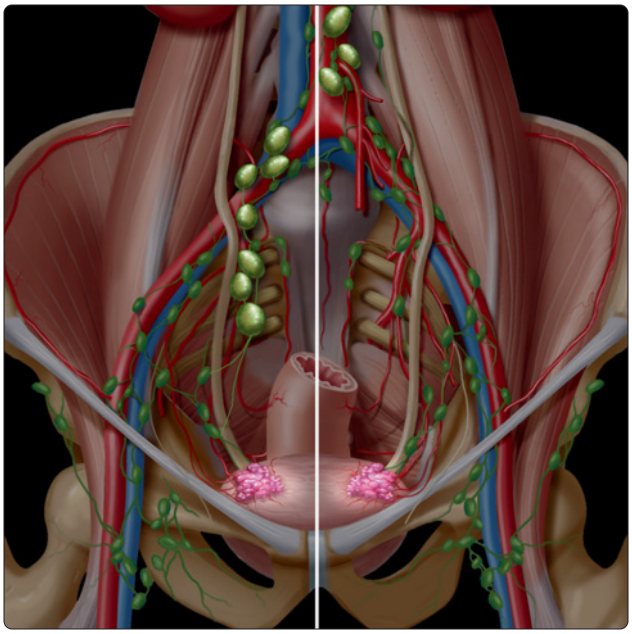
Graphic shows a lateral wall bladder tumor invading the muscles of the lateral pelvic wall, constituting stage T4b.

N1 and N2



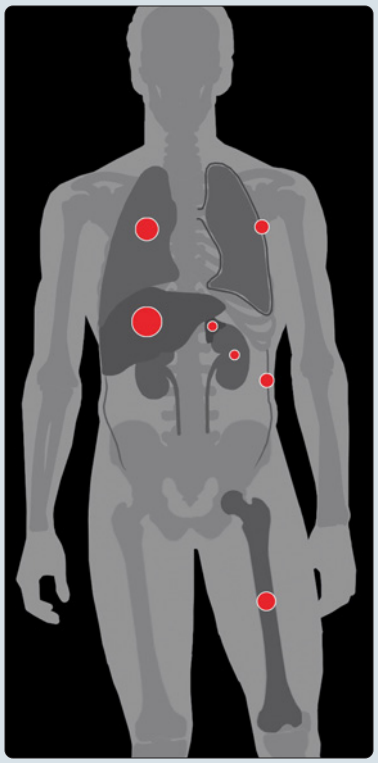
Graphics show N1 and N2 disease in urinary bladder carcinoma. Both classifications describe nodal metastases confined to the true pelvis. N1 (left) is a single metastatic pelvic lymph node, whereas N2 (right) is defined as multiple metastatic pelvic lymph nodes.

N3 and Distant Nodal Metastases



The graphic on the left shows N3 disease, which describes involvement of the common iliac lymph nodes. The graphic on the right shows involvement of paraaortic lymph nodes, which constitute distant metastases and M1 staging in the 7th edition of the AJCC Cancer Staging Manual.

Metastases, Organ Frequency



Liver	*47%*
Lung	*45%*
Bone	*32%*
Peritoneum	*19%*
Pleura	*16%*
Kidney	*14%*
Adrenal gland	*14%*

Adapted from Wallmeroth A et al: Patterns of metastasis in muscle-invasive bladder cancer (pT2-4): an autopsy study on 367 patients. Urol Int. 62:69-75, 1999.

OVERVIEW

General Comments

- Most common urinary tract malignancy

Classification

- Uroepithelial (95%)
  - Transitional cell carcinoma (TCC) (90%)
  - Squamous cell carcinoma (6-8%)
  - Adenocarcinoma (2%)
    - Urachal origin
    - Nonurachal origin (usually chronic irritation)
  - Neuroendocrinal (1%)
  - Mixed
- Mesenchymal (5%) includes
  - Neurofibrosarcoma
  - Pheochromocytoma
  - Lymphoma
  - Angiosarcoma
  - Leiomyosarcoma
  - Rhabdomyosarcoma
  - Liposarcoma
  - Chondrosarcoma
  - Osteosarcoma
  - Plasmacytoma

PATHOLOGY

Routes Spread

- Local spread
  - Extension through layers of bladder wall into perivesical fat
  - Invasion of local pelvic organs
    - Seminal vesicles
    - Prostate
    - Uterus
    - Ovaries
    - Rectum
    - Perineum
  - Later spread to pelvic side wall or anterior abdominal wall
- Lymphatic spread
  - Depends on depth of invasion of bladder wall
    - Superficial tumors (< T2b): Rarely spread to local nodes
    - Deep muscle invasion (T2b): 30% risk of nodal metastases
    - Perivesical extension (T3): 50-60% risk of nodal metastases
  - Initially to perivesical, presacral, and sacral nodes
  - Later to internal iliac, obturator, and external iliac nodes, and eventually to common iliac and paraaortic nodes
  - Regional lymph node metastases are those confined to true pelvis (hypogastric, obturator, external iliac, or presacral lymph node)
    - N1: Single regional lymph node
    - N2: Multiple regional lymph nodes
  - Common iliac nodes are defined as N3 disease
  - All other nodes above aortic bifurcation are considered distant metastases (M1 disease)
- Hematogenous spread
  - Occurs late and with recurrent disease
  - Common sites are bones, lung, brain, and liver
    - Bone metastases occur mainly to pelvic bones and spine (perivesical venous plexus → Batson paravertebral plexus → vertebral bodies)
    - Direct extension to pelvic bones is also common

General Features

- Etiology
  - Direct prolonged contact with excreted carcinogens
    - Smoking is most important risk factor
      - Smokers' risk is 2x that of nonsmokers
    - Industrial carcinogens (aniline, benzidine, arylamine)
    - High level of arsenic in drinking water
  - Recurrent urinary tract infections and stone disease
    - Squamous cell carcinoma
  - *Schistosoma hematobium* infection
    - Squamous cell carcinoma
- Epidemiology cancer
  - Most common tumor of urinary system
    - 4th most common cancer in men
    - 10th most common cancer in women
  - Median age at diagnosis is 73 years
  - Incidence differs by sex and race
    - 4x more common in men than women
    - 2x more common in whites than in African Americans
  - Age-adjusted incidence of 21.2/100,000
    - Estimated 70,980 new cases in USA in 2009
  - Age-adjusted death rate of 4.3/100,000
    - Estimated 14,330 deaths in USA in 2009
- Associated diseases
  - Metaplastic changes can involve extensive areas of bladder epithelium
  - 30% present with multifocal urinary bladder disease
    - Often widespread bladder squamous metaplasia and carcinoma in situ
  - 50% of patients presenting with upper urinary tract carcinoma will develop metachronous tumors in urinary bladder
  - 5% of patients presenting with urinary bladder carcinoma will develop metachronous tumors in upper urinary tract
    - More likely in patients with multiple bladder lesions

Gross Pathology

- Urothelial neoplasms of urinary bladder may be subdivided into
  - Papillary
    - Papillomas
    - Low malignant potential papillary tumors
    - Papillary carcinoma
  - Nonpapillary
    - Urothelial carcinoma in situ
    - Invasive carcinoma
      - Invasive urothelial carcinoma may present as polypoid, sessile, ulcerated, or infiltrative lesion

Microscopic Pathology

- H&E
  - Carcinoma in situ (CIS)

- Malignant urothelial cells within nonpapillary urothelial lining
  - Characterized by extensive (often full thickness) replacement of urothelium by cells demonstrating severe cytologic atypia
- Papillary lesions
  - Microscopic appearance
    - Contain well-defined fibrovascular cores
    - Lining urothelium may vary from indistinguishable from normal (papilloma) to markedly anaplastic (high-grade urothelial carcinoma)
  - Papillary urothelial neoplasia of low malignant potential (PUNLMP)
    - Papillary tumor characterized by cytologically bland, yet thickened, epithelium when compared to papilloma
    - Lower rate of recurrence than low-grade urothelial carcinoma
  - Papillary lesions with cytological atypia
    - Low-grade papillary urothelial carcinoma (LGPUC)
    - High-grade papillary urothelial carcinoma (HGPUC)
- Invasive carcinoma
  - Neoplastic cells invade bladder wall as nests, cords, trabeculae, small clusters, or single cells that are usually separated by desmoplastic stroma

IMAGING FINDINGS

Detection

- **Cystoscopy** is considered gold standard for detection of bladder cancer
- **Intravenous urography (IVP)**
  - Traditionally modality of choice in evaluating hematuria
  - Largely replaced by CT IVP
  - Used primarily to access upper urinary tract + cystoscopy
  - Only 60% of bladder cancers are detected by IVP
    - Small tumors may be obscured in contrast-filled urinary bladder
  - Tumors appear as sessile or pedunculated filling defects
- **Ultrasound**
  - Bladder tumors may be found incidentally or during work-up for hematuria
  - Sonographic features include
    - Focal hypoechoic or mixed echogenicity nonmobile mass projecting into bladder lumen
    - No posterior shadowing
    - Focal bladder wall thickening
    - Color Doppler shows ↑ vascularity (differentiate bladder masses from blood clots)
  - Sonographic detection of bladder tumors depends on size and location
    - Difficult to detect tumors < 5 mm in size and tumors located in neck or dome of bladder
  - Transrectal US can be useful in differentiating bladder neck from prostatic tumor
- **NECT**
  - Modality of choice in patients presenting with painful hematuria to assess for stones
  - Polypoid bladder tumors, isodense to bladder wall, may be seen against background of fluid density urine
    - Changing window settings can make it easier to detect bladder tumors
  - Factors that can make tumor easier to detect against isodense urine
    - Presence of tumor calcifications
      - Tumoral calcifications in approximately 5% of cases of transitional cell carcinoma
      - Calcification typically encrusts tumor surface
    - Presence of high-density hemorrhage in bladder lumen
- **CT IVP**
  - Modality of choice in evaluating patients presenting with painless hematuria
  - Evaluation of upper urinary tract for synchronous tumors in patients with known or suspected carcinoma
  - Consistent performance in detection of bladder tumors
    - Sensitivity (79%)
    - Specificity (94%)
    - Accuracy (91%)
    - Positive predictive value (75%)
    - Negative predictive value (95%)
  - Imaging features
    - Tumors appear as polypoid or flat lesions of similar attenuation compared to bladder wall
    - Larger tumors tend to be more heterogeneous with areas of low density due to necrosis
    - Urachal carcinoma
      - Midline mass anterosuperior to bladder dome
      - Low-attenuation components, which represent pools of mucin at pathologic examination
      - Peripheral calcifications in masses of soft tissue attenuation occur in 50–70% of cases (pathognomonic for urachal adenocarcinoma)
- **MR**
  - Not used routinely for tumor detection
    - Tumors may be detected during pelvic imaging for other indications
  - As good as CT IVP for detection of bladder tumors
    - Sensitivity and positive predictive value > 90%
  - Imaging features
    - T1WI
      - Intermediate signal intensity, isointense to bladder wall, higher than urine and lower than perivesical fat
    - T2WI
      - Slightly hyperintense to bladder wall and hypointense to urine
    - Dynamic gadolinium-enhanced T1WI
      - Earlier and more pronounced enhancement than bladder wall
    - Diffusion-weighted imaging (DWI) under free breathing is very promising in detection of urinary bladder carcinoma
      - ADC values of urinary bladder carcinoma are lower compared to urine, normal bladder wall, prostate, and seminal vesicles
      - Sensitivity (98.1%); specificity (92.3%); positive predictive value (100%); negative predictive value (92.3%); and accuracy (97.0%)

- Dynamic contrast-enhanced MR parameters: Peak time enhancement in 1st minute (E[max/1]) after contrast administration and steepest slope
  - E(max/1) and steepest slope correlate with histologic grade

Staging

- Determined by depth of invasion of bladder wall during cystoscopic examination, which includes
  - Deep biopsy, encompassing all layers of bladder wall
  - Examination under anesthesia to assess
    - Size and mobility of palpable masses
    - Degree of induration of bladder wall
    - Presence of extravesical extension or invasion of adjacent organs
- Unfortunately, clinical staging is not perfect
  - Errors reported in 25-50% of cases
- Patients shown to have muscle-invasive disease following cystoscopy are referred for imaging, usually CT or MR, for complete staging
- **Imaging techniques for local staging**
  - **MR** is imaging modality of choice in local staging of urinary bladder carcinoma
    - Sensitivity and positive predictive value > 90% and overall diagnostic accuracy of 62-75%
    - Inherent high soft tissue contrast
    - Multiplanar capabilities
    - Nonnephrotoxic contrast
    - Superior to CT in assessing depth of muscular invasion
    - Diffusion-weighted MR improves specificity for detection of invasive urinary bladder tumors
  - **CT** is useful to distinguish tumors confined to bladder wall from those spreading into perivesical fat
    - Cannot determine depth of wall invasion to differentiate T2a from T2b disease
  - **Transabdominal US** is not useful for staging
  - **Transurethral US** can be used for staging
    - Can distinguish between T2 and T3 disease
    - Invasive
    - Does not provide more staging information than CT or MR
  - **FDG PET** has limited role in local staging
    - Increased tumor uptake may be obscured by radioisotope excretion into bladder
    - Early results suggest promising role of tracer 11C-choline in evaluation of bladder cancer because of its minimal urinary excretion
- **Local staging**
  - Urinary bladder carcinoma stages
    - Not possible to differentiate Ta from T1 by imaging
    - **T1**: Intraluminal filling defect with normal underlying wall
    - **T2**: Localized wall thickening and retraction indicate muscle involvement
    - **T3a**: Microscopic perivesical invasion cannot be resolved with CT or MR
    - **T3b**: Loss of clear interface between bladder wall and perivesical fat → perivesical fat stranding and nodularity
    - **T4**: Tumor invading adjacent organs, abdominal or pelvic wall, and perineum → distortion and irregularity between tumor and adjacent organs
      - T4 disease includes prostatic stromal invasion; subepithelial invasion of prostatic urethra does not constitute T4 disease
  - Presence of ureteral obstruction strongly suggests muscle invasion
  - MR imaging tends to overestimate local extent of tumor
    - Improved detection of perivesical fat stranding, which may be reactive or inflammatory rather than metastatic
  - MR is particularly helpful to differentiate muscle-invasive from noninvasive disease
    - T2WI
      - Interrupted low signal intensity muscle layer → muscle invasion (T2 disease)
    - Dynamic gadolinium-enhanced T1WI
      - Can differentiate T2a from T2b by showing depth of muscle invasion
    - Improved accuracy of T-stage diagnosis from 67% for T2WI alone to 88% for T2W and DW images
  - Staging CT or MR should be delayed > 7 days after transurethral resection (TUR) because focal wall thickening and perivesical fat stranding can be seen following TUR → overstaging
  - Tumors arising in bladder diverticula are challenging from staging point of view
    - Tend to invade perivesical fat early due to absence of muscle layer in diverticular wall
    - Thinner wall makes accurate staging difficult
    - Omission of T2 has been suggested
      - Tumor either confined to diverticulum (T1) or extradiverticular tumor (T3)
- **Nodal metastases**
  - Nodal size and morphology criteria
    - Nodal metastases in oval nodes > 10 mm and round nodes > 8 mm
  - CT and routine MR have similar accuracy for detection of nodal disease
    - Reported accuracy (73-92%)
    - Sensitivity (83%) and specificity (98%)
  - Improved detection of nodal metastases using ultrasmall superparamagnetic iron oxide (ferumoxtran-10) particles
    - Can detect metastases in normal-sized nodes and exclude metastases in enlarged reactive nodes
    - Normal nodal tissue shows contrast material uptake → ↓ signal intensity on T2- or T2*-weighted images
    - Nodal metastases lack ferumoxtran-10 uptake and retain high-signal intensity on ferumoxtran-10-enhanced images
  - PET can be used for detection of nodal metastases
- **Distant metastases**
  - Lung metastases are common and can present as
    - Solitary nodule
    - Multiple nodules
    - Cavitary nodules
    - Diffuse multinodular opacities
  - Bony metastases are usually lytic, although sclerotic and mixed lesions are also common

- Bone scan may be used if there is suspicion of bone metastases
- T1WI MR is useful to assess pelvic bone marrow involvement

- Brain metastases
  - Rare manifestation of metastatic bladder carcinoma
  - Usually occurs with advanced metastatic disease
  - May present as single or multiple enhancing parenchymal masses
  - Can rarely present as leptomeningeal carcinomatosis
- PET can be used for detection of distant metastases

Restaging

- Routine imaging follow-up is **not** indicated for patients with superficial TCC and no additional risk factors
  - Cystoscopy every 3 months for 2 years, then every 6 months for 2 years, and then yearly thereafter
- Patients with invasive TCC, especially those with risk factors, should have CT IVP every 1-2 years
- Patients requiring cystectomy for invasive bladder cancer should have
  - Abdominal and pelvic CT (or MR) at 6, 12, and 24 months
    - Local recurrence can present as
      - Pelvic lymphadenopathy
      - Well-defined or poorly defined pelvic soft tissue masses
      - Tumor invading adjacent structures, such as vagina, urethra, pelvic sidewall, anterior abdominal wall, seminal vesicle, and spermatic cord
  - Chest x-ray at 6, 12, 18, 24, 36, 48, and 60 months postoperatively

CLINICAL ISSUES

Presentation

- Macroscopic painless hematuria
  - 80% of cases of bladder carcinoma present with painless hematuria
  - Bladder carcinoma is detected in up to 13-28% of patients presenting with macroscopic hematuria
- Urinary frequency, urgency, and dysuria
- Urinary tract infections
- Urinary obstruction
- Urinary bladder rupture is rare; can result
  - Spontaneously
  - Following biopsy
  - From endovesical chemotherapy with mitomycin C
  - As long-term complication of radiotherapy

Cancer History

- Approximately 70% of newly diagnosed cases of bladder cancer represent superficial disease
  - High risk of local recurrence
  - Rarely progress to invasive or metastatic disease
- Risk of disease relapse following radical cystectomy may be as high as 70%
- As many as 50% of patients who have muscle-invasive tumors will have occult metastases that will present within 5 years of diagnosis
- 78% of patients who developed metastases did so within 1 year of cystectomy
  - Suggests that metastases must be present at time of cystectomy
- Presence of ≥ 1 of the following risk factors ↑ likelihood of recurrent or metastatic disease in TCC
  - Extent of bladder wall invasion
  - Tumor size
    - Tumors > 3 cm have up to 35% chance of progression
  - Pathological tumor grade (i.e., degree of differentiation)
    - 5-year survival of patients with grade I tumors is 94% and only 40% for patients with grade III tumors
    - < 10% of grade I tumors, 50% of grade II tumors, and > 80% of grade III tumors are invasive at time of initial diagnosis
  - Adjacent or remote bladder mucosal changes
    - Carcinoma in situ (CIS) in patients with low-grade, low-stage lesions may be associated with progression to muscle invasion (> 80% within 4 years of diagnosis)
  - Multiplicity of foci
    - Recurrence rate is almost 1/3 higher in patients with multiple lesions than in patients with single lesions
  - Upper tract obstruction
    - 5-year survival: 31% in patients with bilateral hydronephrosis
    - 5-year survival: 45% in patients with unilateral hydronephrosis
    - 5-year survival: 63% in patients with no hydronephrosis
  - Lymphatic invasion in lamina propria
    - Very poor prognostic sign
    - Most patients die within 6 years
  - Involvement of prostate
    - Increased risk of urethral recurrence
- 70-80% present with superficial bladder tumors (i.e., stage Ta, Tis, or T1)
  - Complete cure is expected
- 5-year survival rate for bladder cancer by stage
  - Stage 0 (95%)
  - Stage I (85%)
  - Stage II (55%)
  - Stage III (38%)
  - Stage IV (16%)

Treatment Options

- Options by stage
  - **Stage 0 and I**
    - Transurethral resection (TUR) and fulguration
    - TUR with fulguration followed by intravesical bacille Calmette-Guérin (BCG)
    - TUR with fulguration followed by intravesical chemotherapy
    - Segmental cystectomy (rarely indicated)
    - Radical cystectomy in selected patients with extensive or refractory superficial tumor
    - Interstitial implantation of radioisotopes ± external beam radiation therapy (EBRT)
  - **Stage II and III**
    - Radical cystectomy ± pelvic lymph node dissection
    - Urinary diversion becomes necessary and can be accomplished through
      - Cutaneous ureterostomy
      - Ileal conduit with ureteroileocutaneostomy

        - Ureterosigmoidostomy
        - Orthotopic neobladder reconstruction with ileal or ileocecal segments
      - Neoadjuvant platinum-based combination chemotherapy followed by radical cystectomy
      - EBRT ± concurrent chemotherapy
      - Interstitial implantation of radioisotopes before or after EBRT
      - TUR with fulguration (in selected patients)
      - Segmental cystectomy (in selected patients)
    - Locally advanced stage IV
      - Radical cystectomy + pelvic lymph node dissection
      - EBRT
      - Urinary diversion or cystectomy for palliation
      - Chemotherapy as adjunct to local treatment
    - Stage IV with distant metastases
      - Chemotherapy ± adjunct to local treatment
      - EBRT for palliation
      - Urinary diversion or cystectomy for palliation

REPORTING CHECKLIST

T Staging

- Depth of bladder wall invasion
  - MR to evaluate muscle layer invasion
  - Ureteric orifice involvement strongly suggests muscle invasion
  - Focal retraction of outer bladder contour suggests muscle invasion
- Perivesical fat invasion
  - Microscopic perivesical fat invasion cannot be resolved by imaging
- Invasion of surrounding pelvic organs
- Tumors arising in diverticula and involving wall are at least T3 disease

N Staging

- N1: Single lymph node metastasis confined to true pelvis
- N2: Multiple lymph nodes metastases confined to true pelvis
- N3: Lymph node metastases confined to common iliac lymph nodes

M Staging

- Common sites include bones, lungs, and liver
  - Bone metastases can be lytic, mixed, or sclerotic
- Nodal metastases above aortic bifurcation are classified as M1 disease

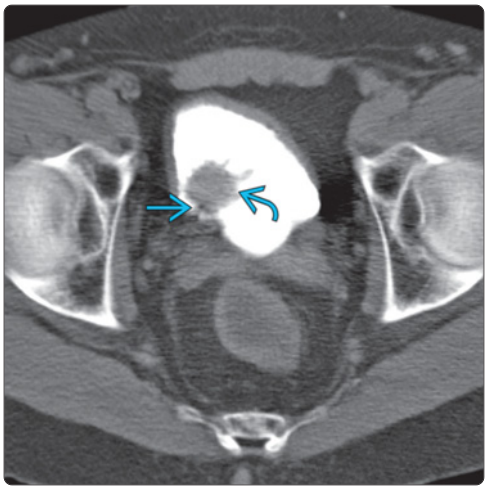
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Stage 0a (Ta N0 M0)

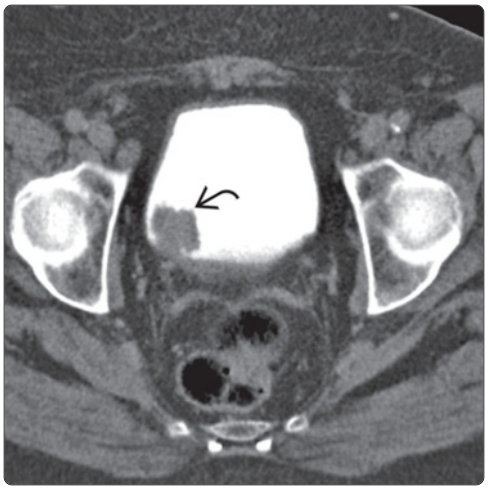


Stage 0a (Ta N0 M0)

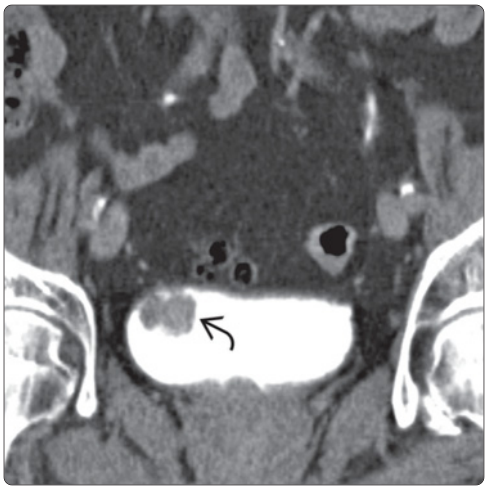


(Left) *Right posterior oblique intravenous pyelography in a patient who presented with gross hematuria shows a papillary mass projecting into the contrast-filled lumen of the urinary bladder.* **(Right)** *Axial CECT in the same patient shows a pedunculated papillary mass with a narrow stalk attached to the right posterolateral aspect of the urinary bladder. The tumor was found to be a noninvasive papillary carcinoma.*

Stage I (T1 N0 M0)

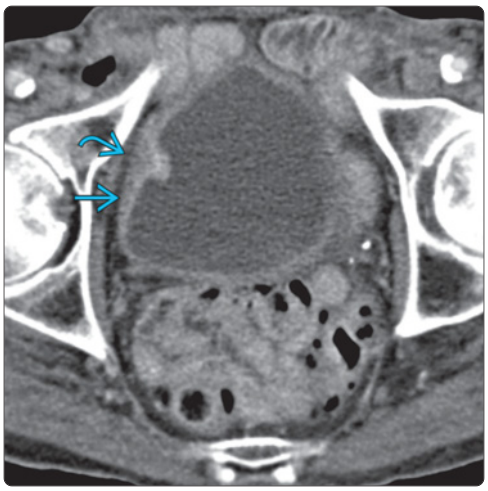


Stage I (T1 N0 M0)

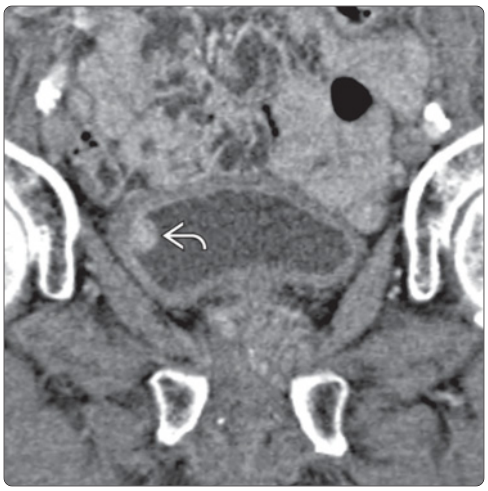


(Left) *Axial CECT shows a polypoid mass involving the right side of the posterior wall of the bladder lateral to the ureteric orifice. The tumor was found to invade into the subepithelial connective tissue without extension into the muscle layer.* **(Right)** *Coronal CECT in the same patient shows the polypoid tumor attached to the superior aspect of the right posterior bladder wall.*

Stage II (T2a N0 M0)

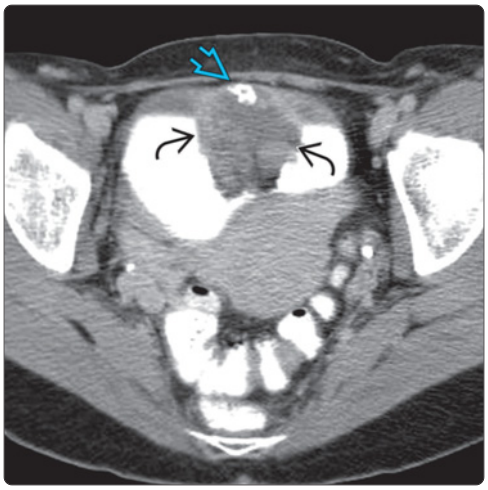


Stage II (T2a N0 M0)

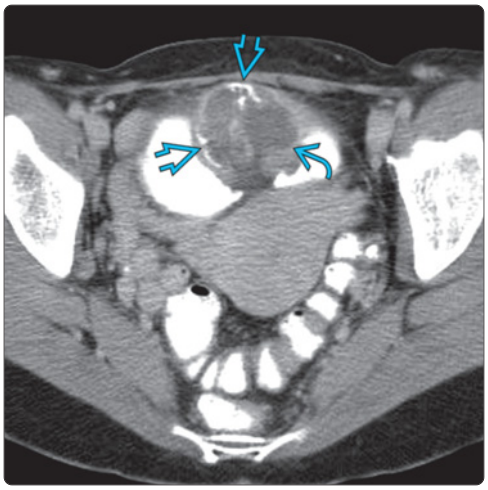


(Left) *Axial CECT shows an enhancing polypoid mass arising from the right lateral wall of the urinary bladder. There is associated thickening of the adjacent bladder wall.* **(Right)** *Coronal CECT in the same patient shows the enhancing urinary bladder mass. Enhancement is limited to the mass without involvement of the underlying wall, suggesting the absence of muscle invasion, although CT is not adequate to definitely make this distinction. This was found to be a T2a tumor.*

Stage II (T2a N0 M0)

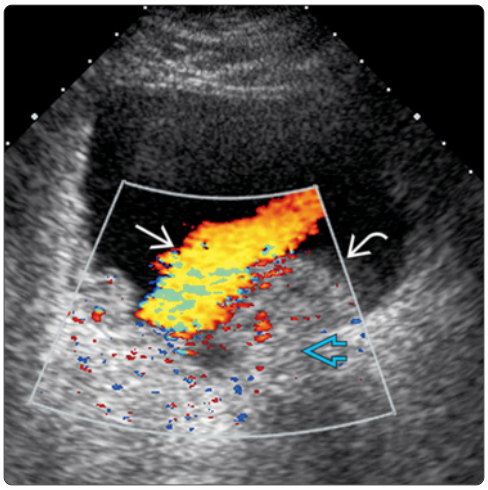


Stage II (T2a N0 M0)

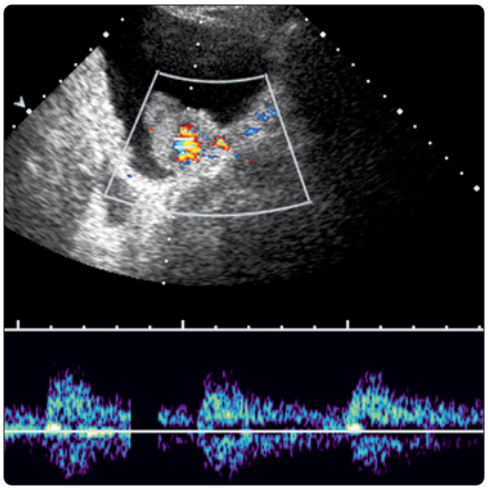


(Left) *Axial CECT in a 38-year-old woman who presented with repeated urinary tract infection and gross hematuria shows a heterogeneous mass in the anterior aspect of the dome of the urinary bladder with focal calcifications. There is no evidence of extravesical extension.* **(Right)** *Axial CECT in the same patient shows the bladder dome mass with peripheral curvilinear calcifications, which is pathognomonic for urachal adenocarcinoma.*

Stage II (T2b N0 M0)

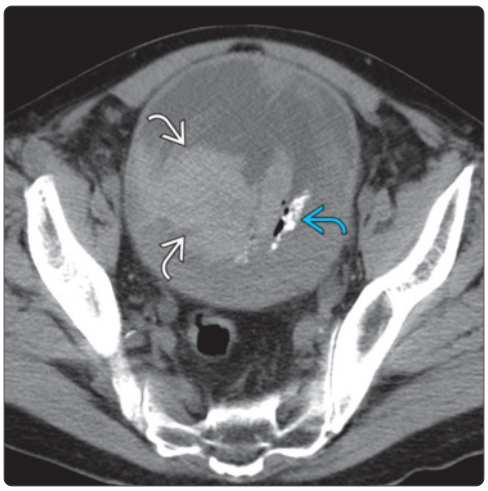


Stage II (T2b N0 M0)

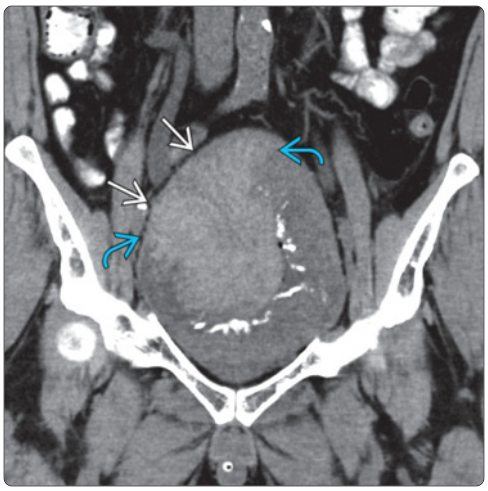


(Left) *Transverse ultrasound image of the urinary bladder shows an echogenic mass centered on and obstructing the left ureteric orifice. Note the normal right side ureteric jet. Ultrasound is not useful for staging bladder carcinoma but can help in tumor detection and evaluation of ureteric orifice.* **(Right)** *Longitudinal duplex Doppler ultrasound in the same patient shows blood flow in the mass, which helps differentiate a solid mass from blood clot.*

Stage II (T2b N0 M0)

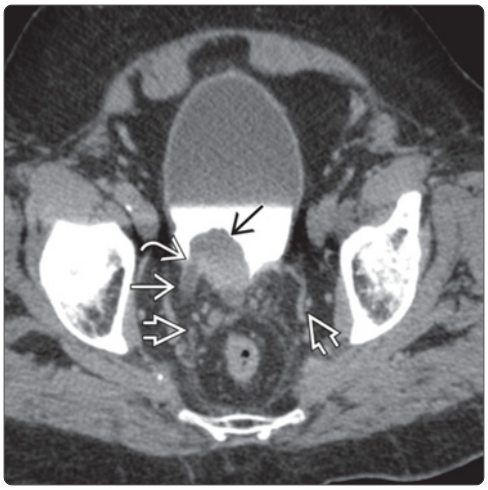


Stage II (T2b N0 M0)

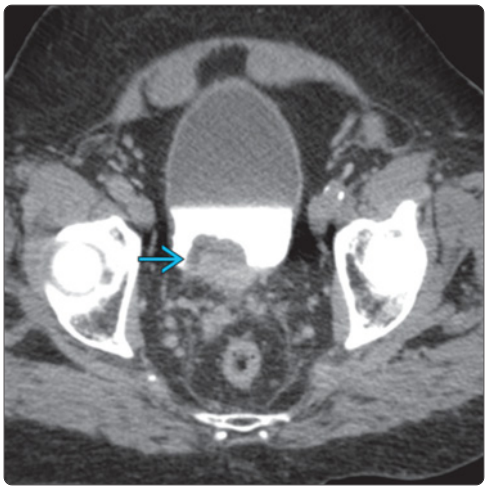


(Left) *Axial CECT shows a large mass filling the bladder without evidence of perivesical infiltration. Calcifications are seen on the surface of the mass.* **(Right)** *Coronal CECT in the same patient shows that the mass is attached to the superior wall of the bladder. Note the smooth outer contour of the bladder at the site of tumor attachment, indicating the absence of perivesical infiltration despite the large tumor size.*

Stage II (T2b N0 M0)

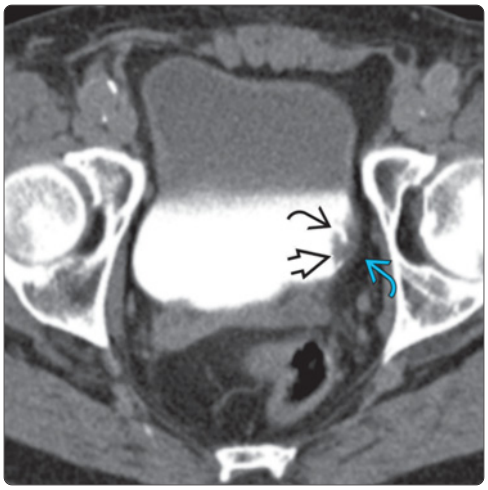


Stage II (T2b N0 M0)

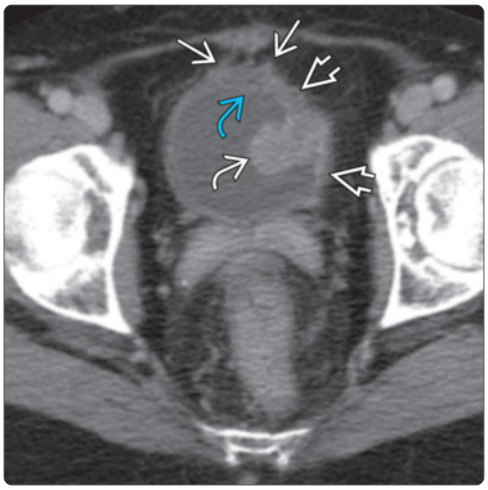


(Left) *Axial CECT in a patient who presented with hematuria shows a posterior wall bladder mass that involves the ureteric orifice. The right ureter is slightly dilated. No regional adenopathy is present. Small nodular densities in the perivesical fat represent dilated veins.* **(Right)** *Axial CECT in the same patient shows involvement of the right ureteric orifice. Ureteric involvement is a strong indication of muscle-invasive (T2) disease.*

Stage II (T2b N0 M0)

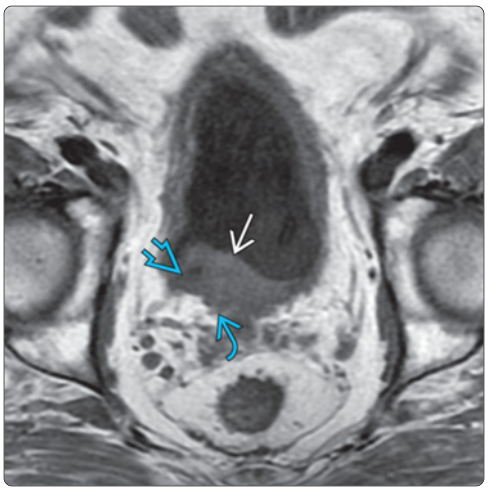


Stage III (T3b N0 M0)

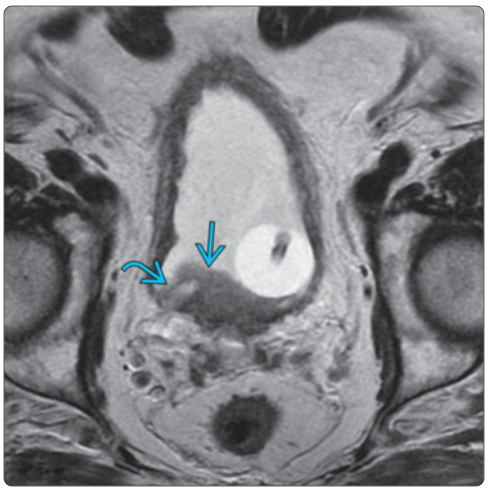


(Left) *Axial CECT shows a polypoid lesion arising from the left lateral wall of the urinary bladder. Contrast is extending into a superficial ulcer. There is no perivesical tumor extension.* **(Right)** *Axial CECT shows thickening of the anterior and left lateral wall of the urinary bladder with a large polypoid mass projecting into the bladder lumen. There is stranding of the perivesical fat almost reaching to the anterior abdominal wall.*

Stage III (T3b N0 M0)

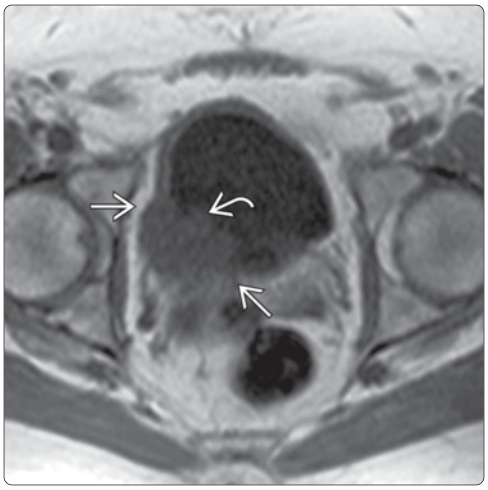


Stage III (T3b N0 M0)

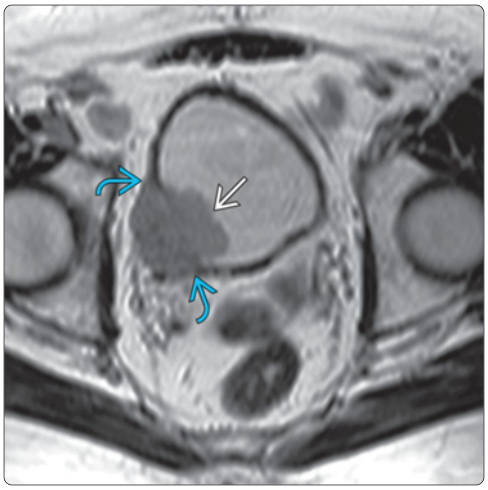


(Left) *Axial T1WI MR shows an intermediate signal intensity posterior wall bladder mass that is hyperintense to urine, hypointense to fat, and isointense to muscle. Tumor nodules extend into high signal perivesical fat. The ureteric orifice is embedded in the mass.* **(Right)** *Axial T2WI MR in the same patient shows an intermediate signal bladder mass that is hypointense to the urine and perivesical fat. Note also involvement of the ureteric orifice.*

Stage III (T3b N0 M0)

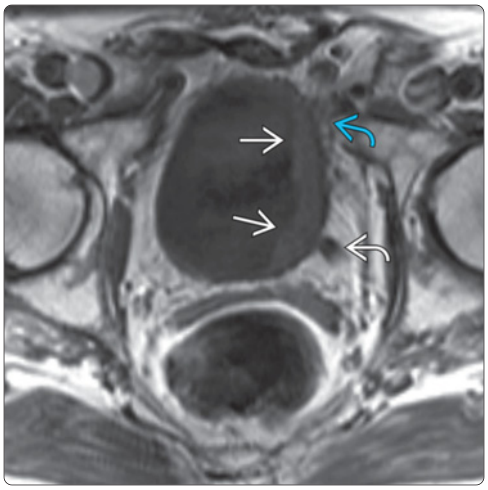


Stage III (T3b N0 M0)

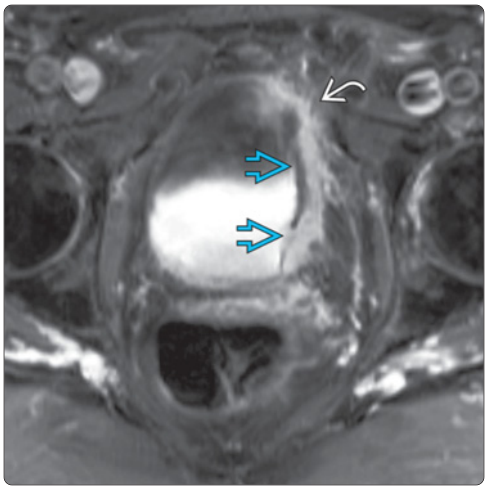


(Left) *Axial T1WI MR shows a right posterolateral urinary bladder mass that has a large extravesical component. The mass is isointense to the bladder wall on T1WI MR.* **(Right)** *Axial T2WI MR shows a bladder mass that is hyperintense relative to the dark urinary bladder wall and hypointense relative to urine and perivesical fat. Note the interruption of the dark urinary bladder wall signal where the mass extends into the perivesical fat.*

Stage III (T3b N0 M0)

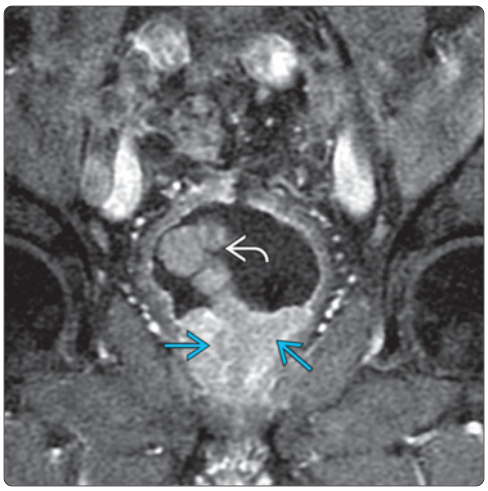


Stage III (T3b N0 M0)

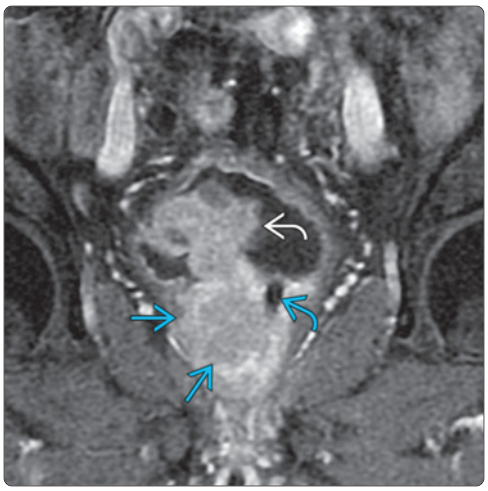


(Left) *Axial T1WI MR shows diffuse thickening of the left lateral urinary bladder wall. Note focal disruption of the smooth outer contour of the bladder and tumor extension into the high signal perivesical fat. The left ureter is dilated, which suggests muscle-invasive disease.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows tumor enhancement and confirms the extension of enhancing tumor tissue into the low signal perivesical fat.*

Stage III (T4a N0 M0)

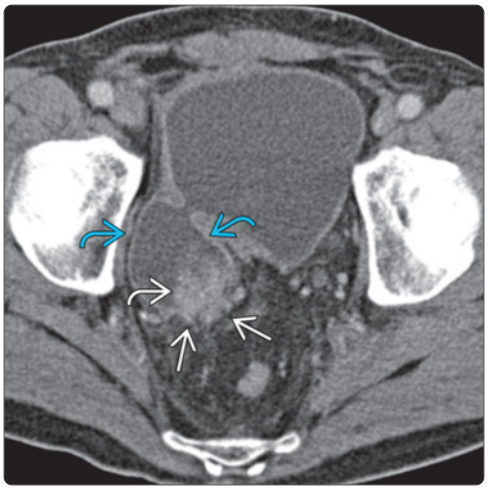


Stage III (T4a N0 M0)

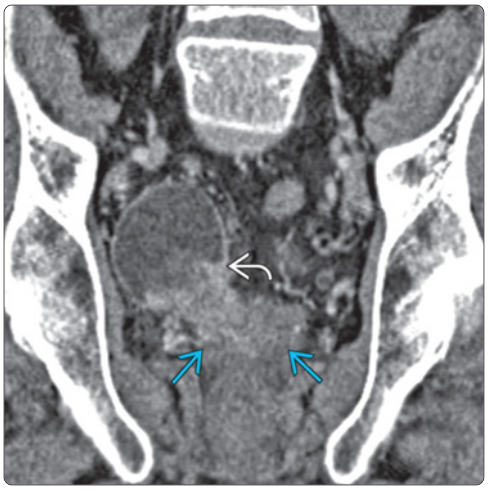


(Left) *Coronal T1WI C+ FS MR in the same patient shows a polypoid bladder mass that infiltrates into the prostate.* **(Right)** *Coronal T1WI C+ FS MR in the same patient again shows the bladder mass invading into the prostate and displacing the Foley catheter toward the left side. It can be difficult to differentiate primary bladder masses invading the prostate from prostate carcinoma invading the neck of the urinary bladder.*

Stage III (T4a N0 M0)

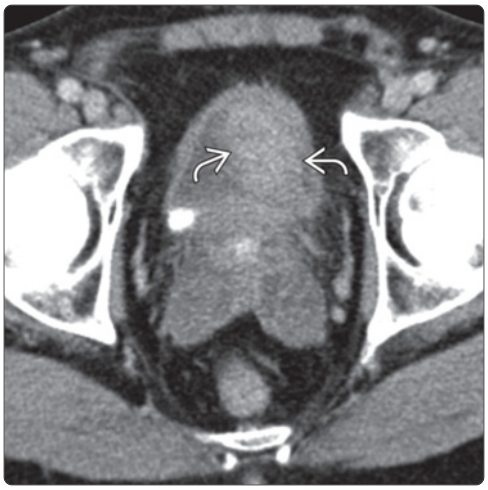


Stage III (T4a N0 M0)

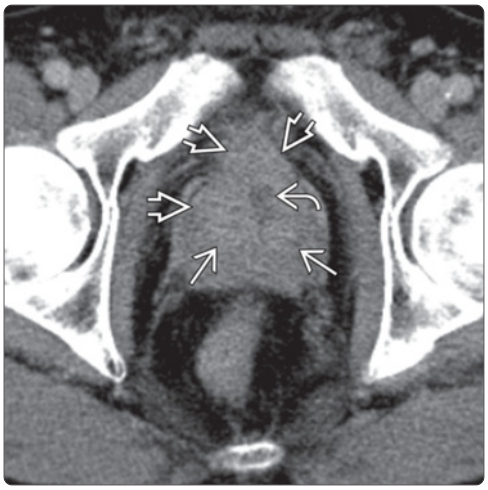


(Left) *Axial CECT shows a right posterolateral bladder diverticulum with a polypoid mass arising within the diverticulum and infiltrating into the perivesical fat. Pathology confirmed the diagnosis of squamous cell carcinoma.* **(Right)** *Coronal CECT in the same patient shows that the mass within the diverticulum invades into the prostate. Because bladder diverticula lack a muscle layer, tumors arising within a diverticulum and invading the wall are at least T3.*

Stage III (T4a N0 M0)

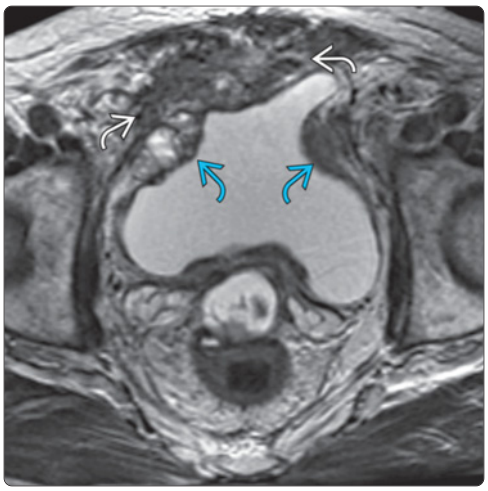


Stage III (T4a N0 M0)

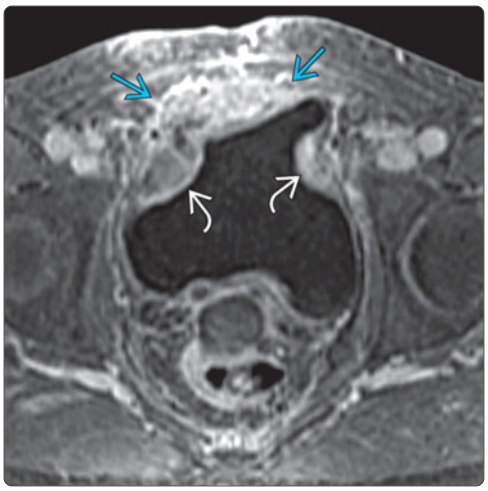


(Left) *Axial CECT shows an enhancing mass in the region of the neck of the urinary bladder.* **(Right)** *Axial CECT in the same patient at the level of the prostate shows tumor surrounding the urethra and invading into the prostate. It can be difficult to differentiate prostatic tumor invading the bladder neck from bladder tumors invading the prostate. The presence of prostate invasion makes this T4a disease.*

Stage IV (T4b N0 M0)

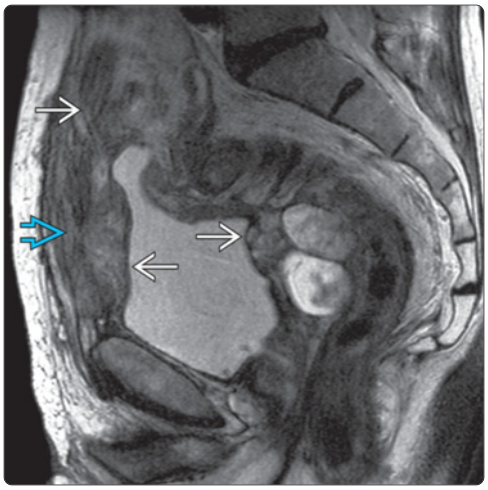


Stage IV (T4b N0 M0)

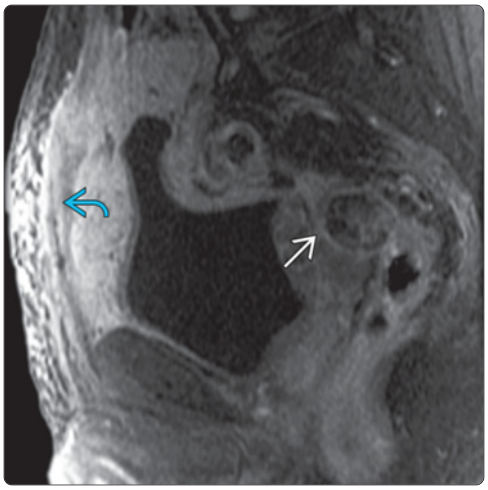


(Left) *Axial T2WI MR shows extensive infiltrative tumor involving mainly the anterior bladder wall with focal invasion of the rectus abdominus muscle. The tumor shows heterogeneous T2 signal intensity.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows heterogeneously enhancing tumor of the anterior wall of the bladder. Enhancing tumor fills the prevesical space and invades the rectus muscles.*

Stage IV (T4b N0 M0)

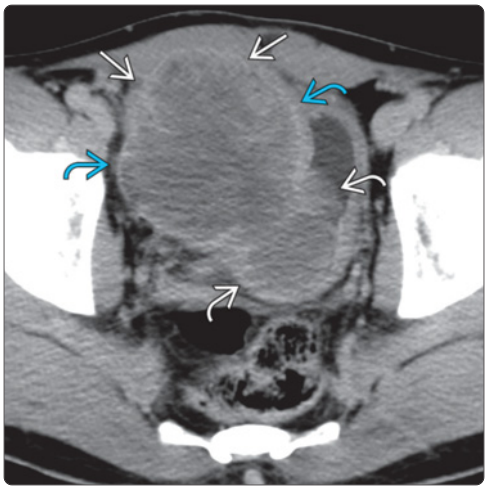


Stage IV (T4b N0 M0)

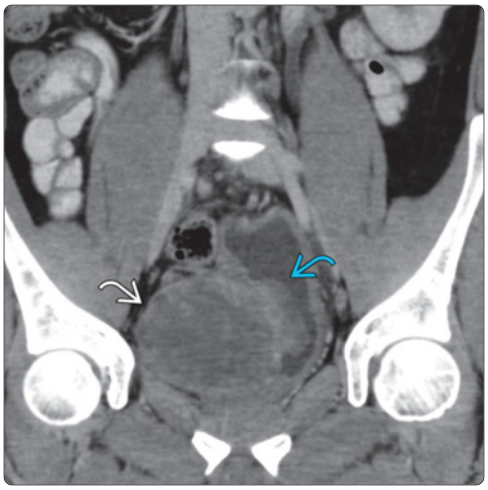


(Left) *Sagittal T2WI MR in the same patient shows marked circumferential thickening of the wall of the urinary bladder, as well as invasion of the rectus muscles anteriorly.* **(Right)** *Sagittal T1WI C+ FS MR in the same patient shows diffuse enhancement of the bladder wall with tumor extension into the wall of the rectum and the anterior abdominal wall muscles.*

Stage IV (T4b N0 M0)

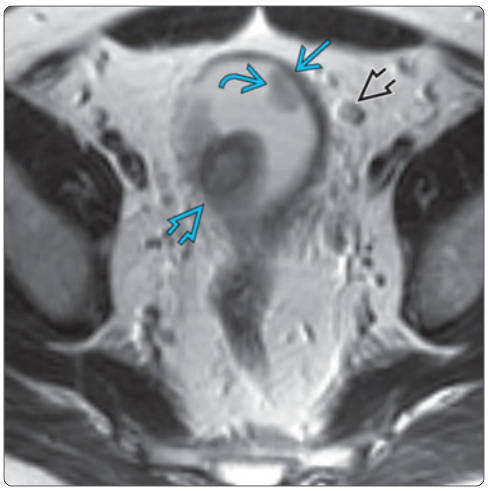


Stage IV (T4b N0 M0)

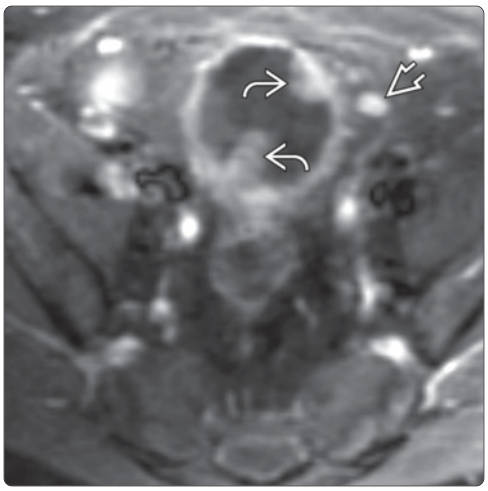


(Left) *Axial CECT shows a large dumbbell-shaped polypoid mass that has a large intraluminal component and a large extraluminal component. The extraluminal component invades into the anterior abdominal wall.* **(Right)** *Coronal CECT in the same patient shows the intraluminal and large extraluminal components. This lesion was initially thought to be a mesenchymal mural tumor but was found at surgery to be transitional cell carcinoma.*

Stage IV (T2b N1 M0)

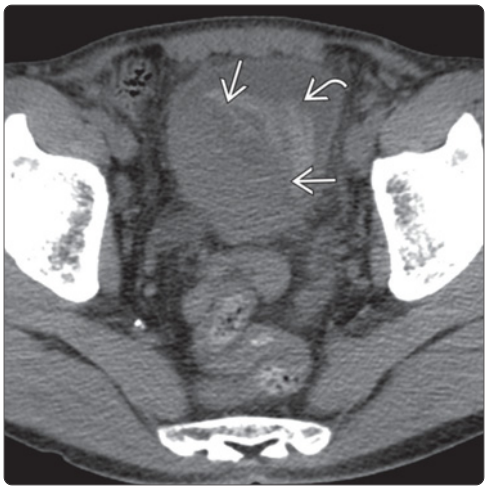


Stage IV (T2b N1 M0)

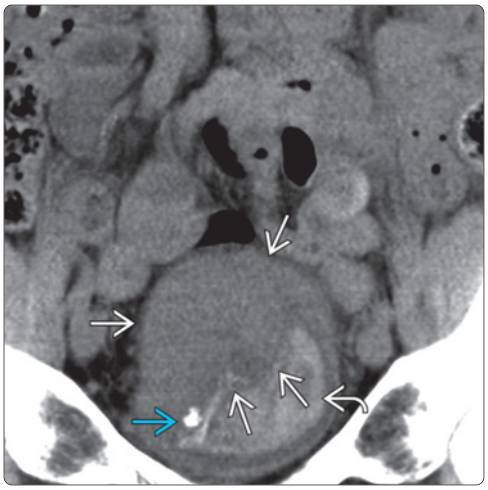


(Left) *Axial T2WI MR shows 2 urinary bladder masses. The 1st mass is superficial and does not invade through the muscle layer (uniform low signal intensity muscle layer at the base of the lesion). The 2nd lesion invades and interrupts the muscle layer. A perivesical lymph node is present.* **(Right)** *Axial T1WI C+ FS MR in the same patient shows enhancement of both lesions, as well as similar enhancement of the perivesical lymph node.*

Stage IV (T2b N1 M0)

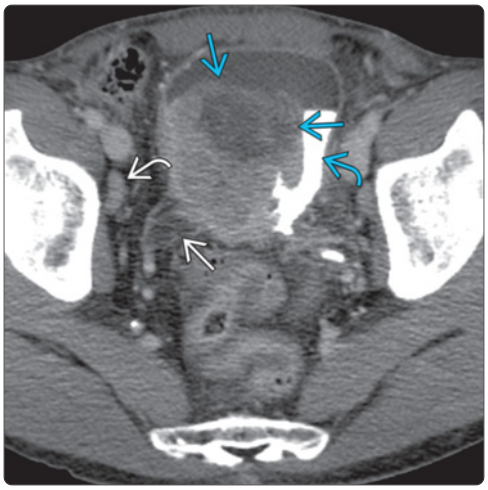


Stage IV (T2b N1 M0)

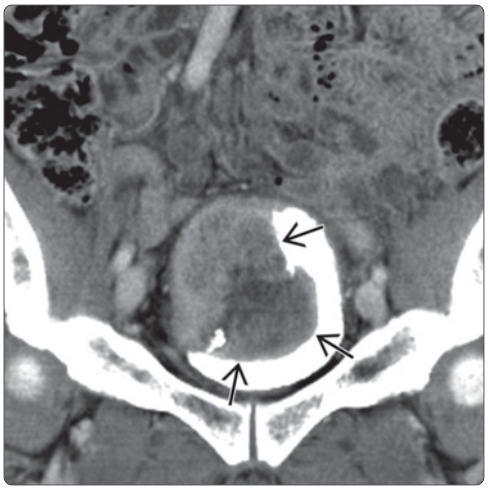


(Left) *Axial NECT in a patient who presented with gross hematuria shows high-density blood within the urinary bladder. The intraluminal blood outlines a large urinary bladder tumor that is otherwise isodense to urine.* **(Right)** *Coronal NECT in the same patient shows intravesical blood outlining the bladder tumor. There are focal tumoral calcifications. The bladder has a smooth outer contour, indicating the absence of perivesical invasion.*

Stage IV (T2b N1 M0)

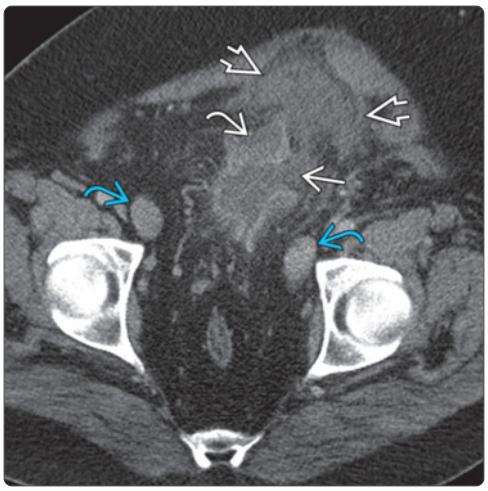


Stage IV (T2b N1 M0)

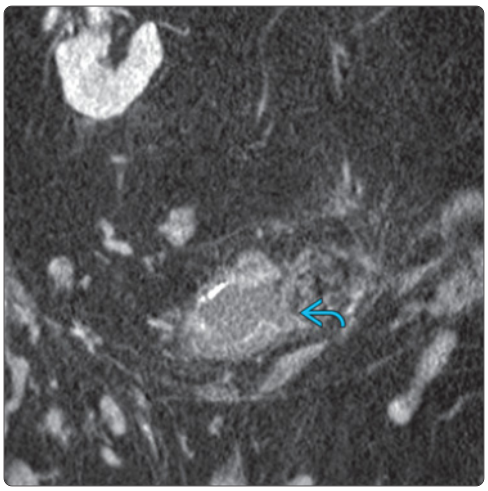


(Left) *Axial CECT in the same patient shows contrast material within the urinary bladder outlining the large necrotic bladder mass. Note involvement of the ureteric orifice and mild ureteric dilatation. There is a single obturator lymph node, which constitutes N1 disease (stage IV).* **(Right)** *Coronal CECT in the same patient shows the tumor outlined by intravesical contrast.*

Stage IV (T2b N2 M0)

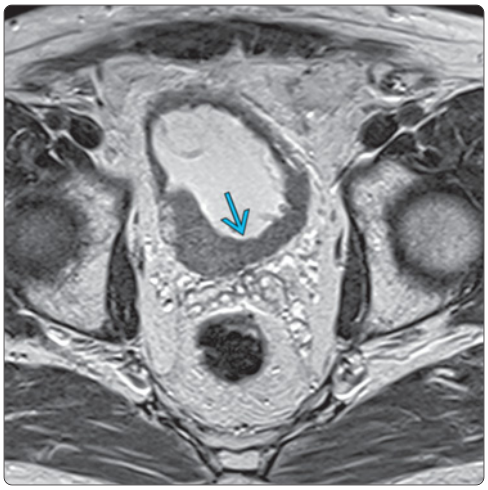


Stage IV (T2b N2 M0)

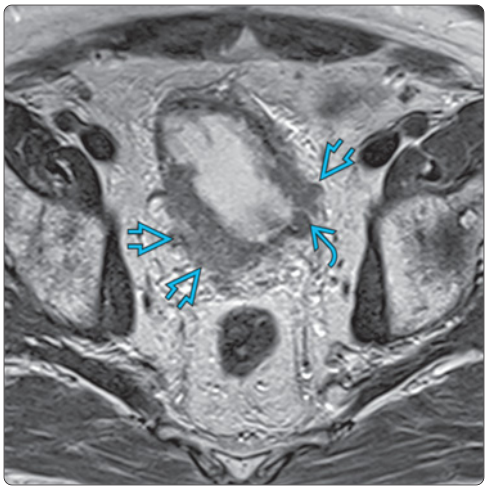


(Left) *Axial CECT in a morbidly obese patient with pathologically proven T2b disease, who presented with severe lower abdominal pain, shows focal thickening of the anterior wall of the urinary bladder with a large defect of the bladder wall and fluid collection in the prevesical space due to bladder rupture. Bilateral enlarged external iliac nodes are seen.* **(Right)** *Coronal CECT in the same patient shows the bladder defect.*

Stage IV (T3b N2 M0)

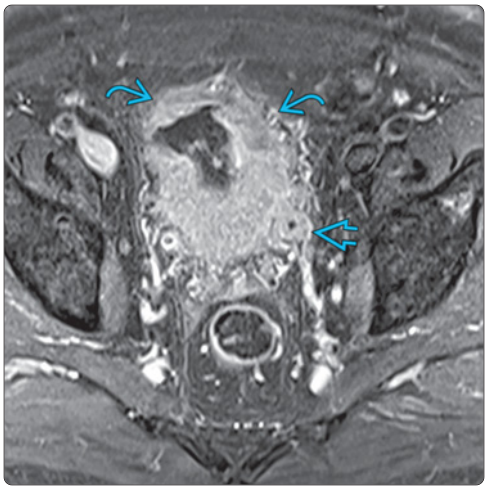


Stage IV (T3b N2 M0)

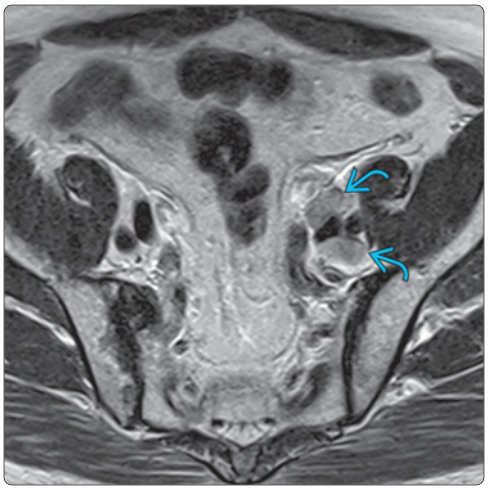


(Left) *Axial T2WI MR in a 72-year-old man who presented with painless hematuria shows posterior urinary bladder wall thickening with loss of the low signal bladder wall indicating muscle invasion.* **(Right)** *Axial T2WI MR in the same patient shows thickening of the wall of the distal left ureter due to tumor invasion. Irregular outer contour of the urinary bladder due to gross invasion of the perivesical fat, constituting T3b disease, is also shown.*

Stage IV (T3b N2 M0)

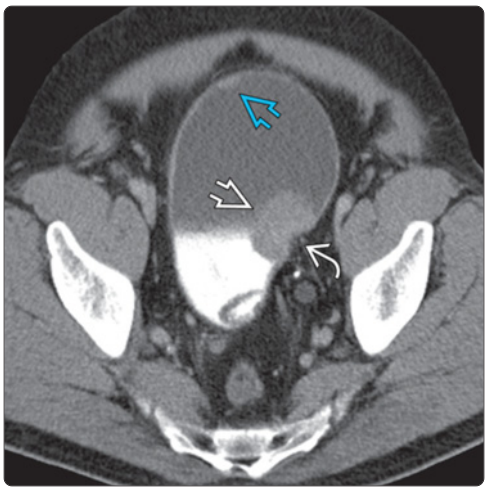


Stage IV (T3b N2 M0)

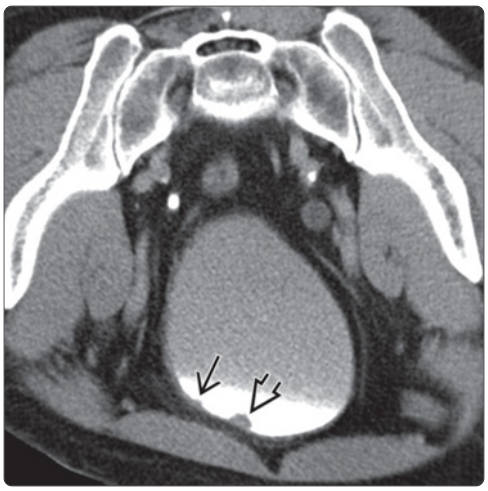


(Left) *Axial T1WI C+ FS MR obtained at a higher level in the same patient shows diffuse enhancing wall thickening involving the dome of the urinary bladder with circumferential thickening of the distal left ureter due to tumor invasion.* **(Right)** *Axial T2WI MR in the same patient shows 2 enlarged left external iliac lymph nodes. The presence of multiple lymph nodes metastases within the true pelvis constitutes N2 disease.*

Stage IV (T4a N2 M0)

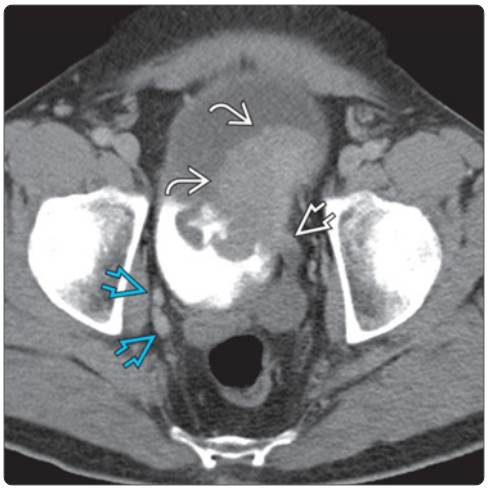


Stage IV (T4a N2 M0)

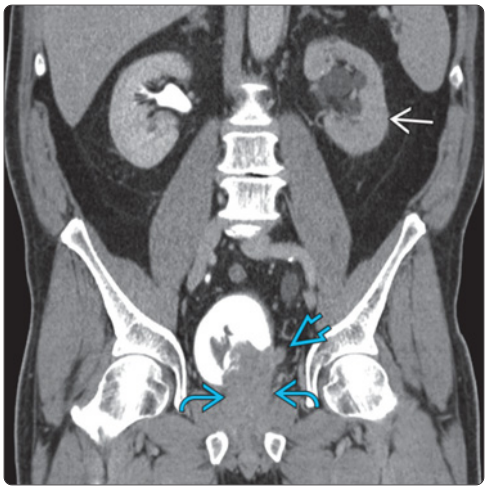


(Left) *Axial CECT in a 54-year-old man shows a large polypoid lesion with indentation of the urinary bladder wall at the site of tumor attachment indicating muscle invasion. Note the subtle nodular lesion of the anterior bladder wall.* **(Right)** *Axial CECT in the same patient obtained in the prone position confirms the presence of the anterior wall polypoid lesion and shows another subtle flat lesion, which was pathologically proven to be carcinoma in situ.*

Stage IV (T4a N2 M0)

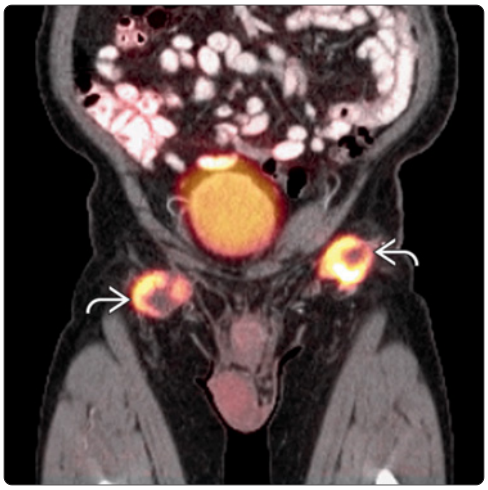


Stage IV (T4a N2 M0)

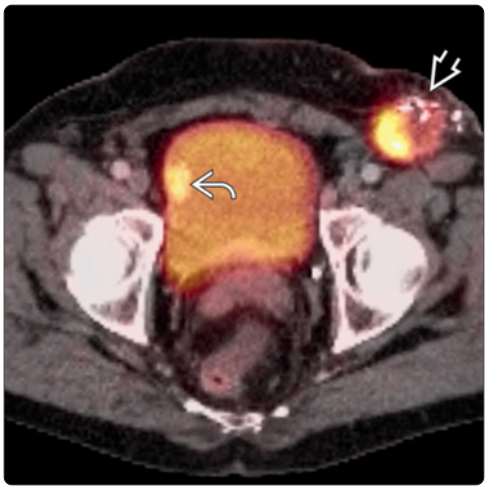


(Left) *Axial CECT in the same patient shows the large polypoid mass invading the left ureter. Two metastatic obturator lymph nodes are present. The presence of multiple metastatic lymph nodes within the true pelvis constitute N2 disease.* **(Right)** *Coronal CECT in the same patient shows invasion of the prostate and the left ureteric orifice. The left kidney shows delayed nephrogram due to left ureteric obstruction.*

Stage IV (T2b N0 M1)

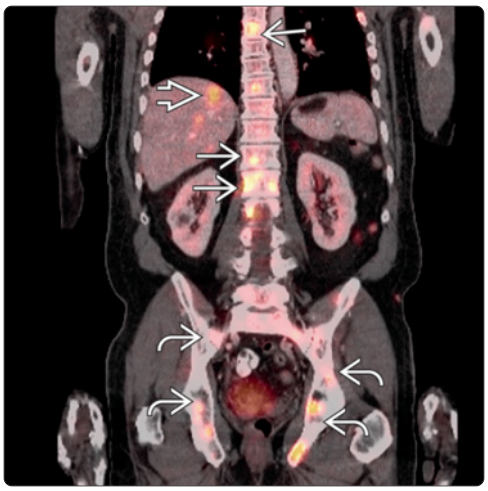


Stage IV (T2b N0 M1)

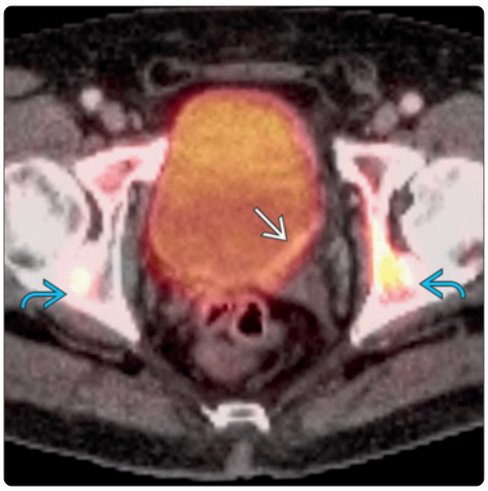


(Left) *Coronal PET/CT was obtained after forced diuresis in a patient who presented with bilateral inguinal lymphadenopathy that was pathologically proven to be metastatic transitional cell carcinoma. Bilateral FDG-avid inguinal lymph nodes are seen.* **(Right)** *Axial PET/CT in the same patient shows inguinal lymphadenopathy. A polypoid bladder mass is seen, representing the primary tumor that was found to be T2b tumor at surgery.*

Stage IV (T3b N0 M1)



Stage IV (T3b N0 M1)



(Left) *Coronal PET/CT shows liver metastases, multiple spine metastases, as well as multiple pelvic bony metastases. Spine and bony pelvic metastases are common due to tumor spread through the Batson venous plexus.* **(Right)** *Axial PET/CT in the same patient shows uptake along the left posterolateral wall of the urinary bladder following resection of a muscle-invasive tumor and bilateral acetabular uptake due to pelvic bony metastases.*

Squamous Cell Carcinoma

KEY FACTS

TERMINOLOGY

- Malignant epithelial neoplasm with pure squamous cell phenotype

IMAGING

- CT with delayed phase imaging (CT urography) modality of choice
- Enhancing plaque-like or sessile polypoid lesion
- Tumor has predilection for trigone and lateral walls
  - May occur in bladder diverticula
- Invasion through the serosal layer (T3b) manifested by soft tissue and stranding in perivesical fat
- Differentiation between malignant and reactive nodes can be very difficult
- Tumor has intermediate T1 and T2 signal intensity on MR imaging and shows early enhancement

TOP DIFFERENTIAL DIAGNOSES

- Invasive urothelial carcinoma
  - Differentiation between urothelial carcinoma and SCC almost impossible based on imaging alone
- Rare bladder neoplasms
- Chronic granulomatous cystitis

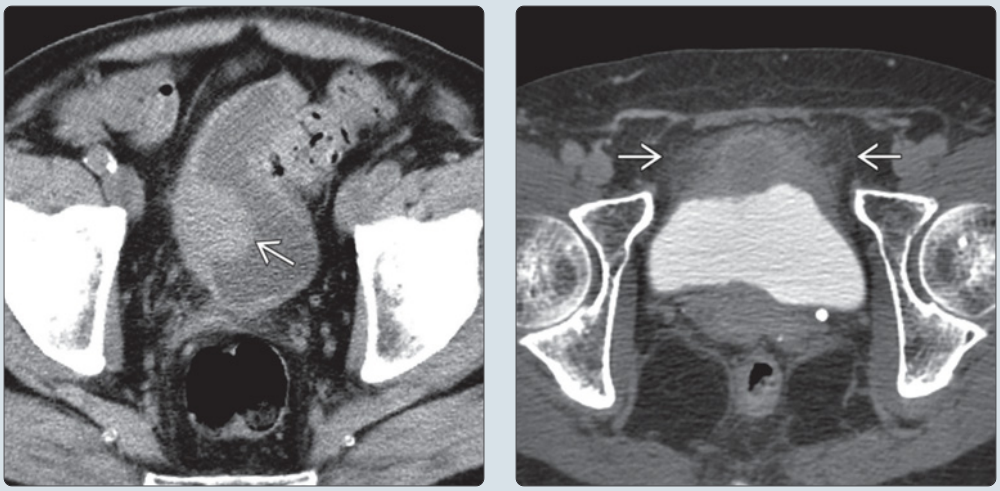
PATHOLOGY

- Chronic mucosal irritation and inflammation play major role
  - Chronic/repetitive injury → squamous metaplasia → dysplasia → carcinoma
- Staging of bladder SCC follows same TNM staging scheme used for urothelial bladder carcinoma

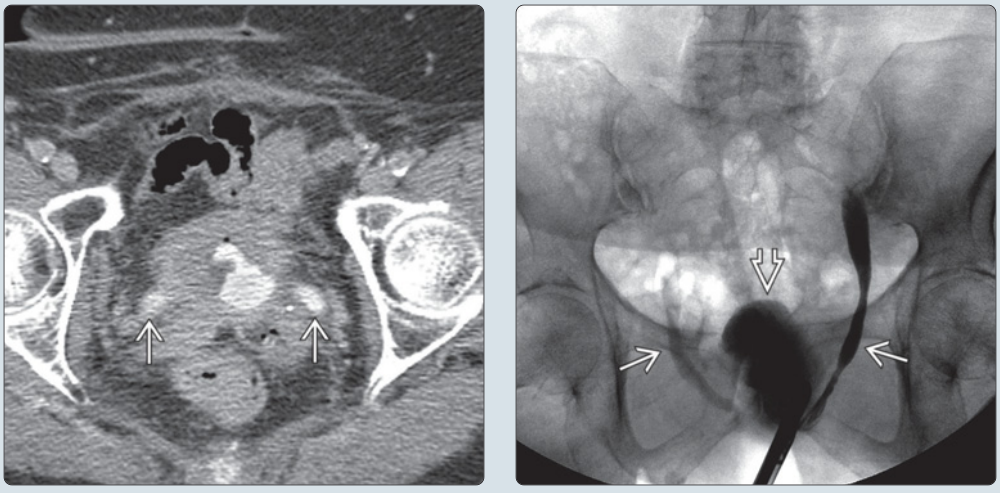
CLINICAL ISSUES

- Common symptoms: Painless microscopic hematuria
- Nonschistosomal forms usually present in advanced stage (80% have detrusor muscle invasion and up to 10% are metastatic at presentation)
  - Schistosomiasis-related forms, on the other hand, have relatively low prevalence of nodal and distant metastasis
- Radical cystectomy treatment of choice

(Left) *Axial NECT in a patient with a history of hematuria shows a soft tissue-attenuation polypoid mass ➡ along lateral bladder wall. Patient underwent transurethral resection of bladder tumor (TURBT). Pathology was compatible with squamous cell carcinoma.* **(Right)** *CECT shows marked, irregular wall thickening of anterior bladder wall. Note perivesical fat stranding ➡, suggestive of tumor extension into serosa (T3 disease). Patient underwent radical cystectomy; pathology was compatible with T3N1 disease.*



(Left) *Axial delayed phase CECT in patient with a history of remote radiation for anal cancers shows marked, irregular circumferential bladder wall thickening. There was extension of infiltrative tumor to the ureterovesical junctions resulting in bilateral hydroureteronephrosis ➡.* **(Right)** *Frontal retrograde cystogram in the same patient shows irregular contour and decreased distensibility of the urinary bladder ➡. Note bilateral hydroureter ➡.*



TERMINOLOGY

Abbreviations

- SCC: Squamous cell carcinoma

Definitions

- Malignant epithelial neoplasm with pure squamous cell phenotype

IMAGING

General Features

- Best diagnostic clue
  - Bladder mass or eccentric bladder wall thickening
- Location
  - Predilection for trigone and lateral walls
  - May occur in bladder diverticula
- Morphology
  - Plaque-like, polypoid, or infiltrative

CT Findings

- Enhancing plaque-like or sessile polypoid lesion
- ± calcification
- Invasion through serosal layer (T3b) manifested by soft tissue and stranding in perivesical fat

MR Findings

- Tumor has intermediate T1 and T2 signal intensity
- Shows early enhancement
- Fat-suppressed post-contrast sequences extremely useful for detection of perivesical extension (T3b)

Ultrasonographic Findings

- Hypoechoic plaque-like or polypoid mass
- Internal flow may be seen on Doppler

Imaging Recommendations

- Best imaging tool
  - CT with delayed phase imaging (CT urography)

DIFFERENTIAL DIAGNOSIS

Invasive Urothelial Carcinoma

- Differentiation between urothelial carcinoma and SCC almost impossible based on imaging alone
- SCC (nonschistosomal type) usually more locally advanced at time of presentation

Rare Bladder Neoplasms

- Adenocarcinoma, small cell tumor
- Cystoscopy with tissue sampling necessary for diagnosis

Chronic Granulomatous Cystitis

- Differentiation between changes of chronic granulomatous cystitis and SCC in patients with schistosomiasis can be difficult
- Urine cytology plays important role

PATHOLOGY

General Features

- Etiology
  - Chronic mucosal irritation and inflammation play major role
    - Chronic/repetitive injury → squamous metaplasia → dysplasia → carcinoma
  - Common causes of chronic injury: Chronic urinary tract infection, intravesical calculi, long-term indwelling catheter, infestation with *Schistosoma*, cyclophosphamide
  - Reported increased risk associated with smoking

Staging, Grading, & Classification

- Staging of bladder SCC follows same TNM staging schema used for urothelial bladder carcinoma

Microscopic Features

- Keratinized islands ("squamous pearls") composed of concentric cell aggregations with varying degree of differentiation
- Differentiation from poorly differentiated urothelial carcinoma with squamous differentiation might be difficult
  - By definition, to be considered pure SCC no component of urothelial carcinoma should be present

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless microscopic hematuria
  - Irritative symptoms (frequency, dysuria)

Demographics

- Age
  - Elderly (usually in 7th decade of life)
  - Cases related to schistosomiasis present in younger age
- Epidemiology
  - Accounts for 3-7% of bladder cancers in United States
    - Has much higher prevalence in endemic regions for schistosomiasis, such as Egypt and Sudan

Natural History & Prognosis

- Nonschistosomal forms usually present in advanced stage (80% have detrusor muscle invasion and up to 10% have metastasis at presentation)
  - Schistosomiasis-related forms, on the other hand, have relatively low prevalence of nodal and distant metastasis
    - Likely related to lymphatic and capillary fibrosis in setting of schistosomiasis and lower histologic grade of tumor in this population

Treatment

- Radical cystectomy is treatment of choice
- Transurethral resection of bladder tumor (TURBT) can be performed for smaller polypoid lesions

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2. Mouli S et al: Imaging features of common and uncommon bladder neoplasms. Radiol Clin North Am. 50(2):301-16, vi, 2012
3. Wong-You-Cheong J et al: Neoplasms of the urinary bladder: Radiologic-pathologic correlation. RadioGraphics 26: 553-580',2006
4. Wong JT et al: Bladder squamous cell carcinoma. Radiographics. 24(3):855-60, 2004

Adenocarcinoma

KEY FACTS

TERMINOLOGY

- Primary gland-forming carcinoma of urinary bladder
- 1/3 are associated with urachal remnant (urachal type)

IMAGING

- Predilection for bladder base and urachus (urachal remnant)
- Tumor may present as a heterogeneous mass or irregular focal or diffuse bladder wall thickening
- Most tumors are mixed solid and cystic
  - Cystic component due to mucin (which is T2 hyperintense on MR)
- ~ 70% of cases have calcification
- Extravesical extension is common
- The bulk of tumor in urachal adenocarcinoma is extravesical
- Urachal adenocarcinomas are usually large at time of presentation (mean size: 6 cm)

TOP DIFFERENTIAL DIAGNOSES

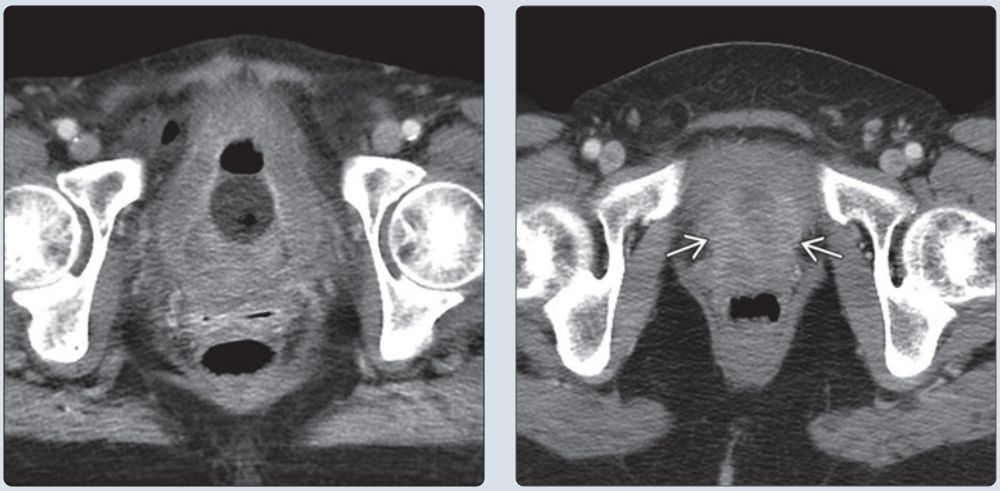
- Urothelial carcinoma
  - Urothelial carcinoma usually shows papillary intravesical growth, which is unusual in adenocarcinoma
  - Presence of cystic spaced (mucin) and calcification are clues for adenocarcinoma
- Other uncommon bladder neoplasms
- Cystitis

PATHOLOGY

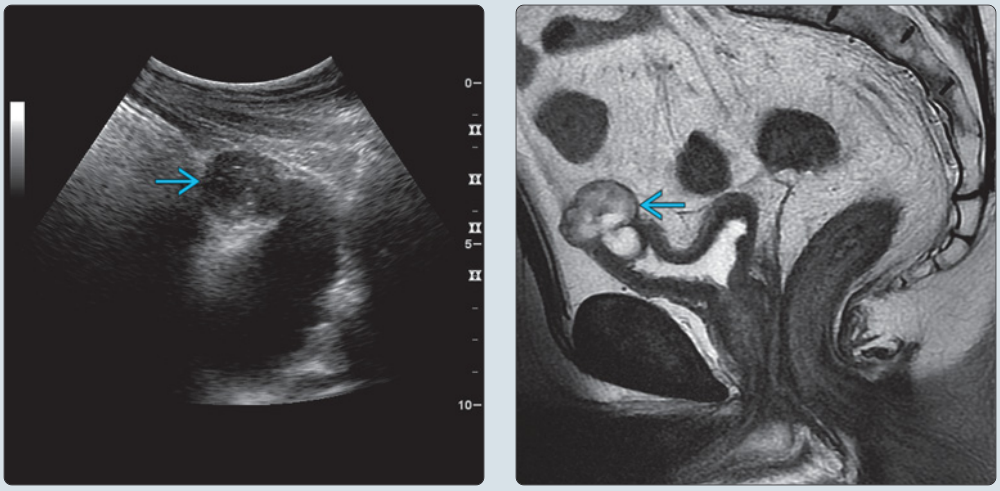
- Proposed pathogenesis: Chronic mucosal irritation → intestinal metaplasia → adenocarcinoma
- Risk factors: Bladder exstrophy, **urachal remnant,** chronic mucosal irritation/inflammation, cystitis glandularis (such as with pelvic lipomatosis), endometriosis

CLINICAL ISSUES

- Rare; < 2% of bladder malignancies
- Usually in advanced stage at the time of presentation



(Left) *Axial CECT in a 63-year-old female shows marked circumferential bladder wall thickening. Tissue biopsy was compatible with clear cell carcinoma. There was extension into urethra and invasion into anterior vaginal wall (T4a lesion). The patient underwent pelvic exenteration.* **(Right)** *Axial CECT shows a heterogeneously enhancing mass at the bladder base, which was shown to be bladder adenocarcinoma (nonurachal type). The patient underwent radical cystectomy.*



(Left) *Longitudinal US image of the bladder shows an incidental complex mass superior to the bladder dome.* **(Right)** *Sagittal T2W MR in the same patient shows a complex solid/cystic mass attached to the bladder dome. The T2 hyperintense areas are related to mucin content. The patient underwent partial cystectomy and pathology was consistent with urachal adenocarcinoma.*

TERMINOLOGY

Definitions

- Primary gland-forming carcinoma of urinary bladder
- 1/3 are associated with urachal remnant (urachal type)

IMAGING

General Features

- Location
  - Predilection for bladder base and urachus (urachal remnant)
- Size
  - Urachal adenocarcinomas are usually large at time of presentation (mean size: 6 cm)
- Morphology
  - Urachal: Large midline supravesical/infraumbilical mass with calcification
  - Nonurachal: Could present as mass or marked bladder wall thickening

Radiographic Findings

- Antegrade/retrograde cystography
  - Filling defect in bladder base
  - Contour deformity and external compression in bladder dome (urachal form)

CT Findings

- Irregular focal or diffuse bladder wall thickening
- Most tumors are mixed solid and cystic
  - Cystic component due to mucin
- ~ 70% of cases have calcification
  - Usually peripheral
- Extravesical extension is common
  - Perivesical fat stranding
  - Regional or distant lymphadenopathy
  - Peritoneal carcinomatosis
- Bulk of tumor in urachal adenocarcinoma is extravesical

MR Findings

- Enhancing mass with heterogeneous signal on T1WI and T2WI
- Contains areas of high T2 signal intensity corresponding to mucin
- Extravesical extension (T3 tumor) manifests as perivesical fat stranding and enhancement

Ultrasonographic Findings

- Heterogeneous intravesical mass
  - Shadowing calcification
- Complex solid/cystic mass superior to bladder dome (urachal type)

DIFFERENTIAL DIAGNOSIS

Urothelial Carcinoma

- Urothelial carcinoma usually shows papillary intravesical growth, which is unusual in adenocarcinoma
- Presence of cystic spaces (mucin) and calcification are clues for adenocarcinoma
- Adenocarcinoma is most common urachal neoplasm

Other Uncommon Bladder Neoplasms

- Such as squamous cell carcinoma, small cell tumor, carcinoid, etc.
- Differentiation between these conditions based on imaging alone is difficult

Cystitis

- Adenocarcinoma can present as diffuse bladder wall thickening and mimic cystitis

PATHOLOGY

General Features

- Etiology
  - Risk factors: Bladder exstrophy, urachal remnant, chronic mucosal irritation/inflammation, cystitis glandularis (such as with pelvic lipomatosis), endometriosis
  - 80% of urachal carcinomas are adenocarcinoma
    - Urachal adenocarcinoma is thought to arise from rests of embryonic hindgut epithelium

Microscopic Features

- Important to differentiate primary from secondary adenocarcinoma (invasion from adjacent organs or distant metastasis)
- Gland-forming with vacuolated cytoplasm

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Hematuria
  - Irritative symptoms (dysuria, frequency)

Demographics

- Age
  - Peak incidence in 6th decade
  - Patients with urachal type present ~ 10 years earlier
- Gender
  - More common in male (M:F=2.6:1)
  - No gender predilection in urachal type
- Epidemiology
  - Rare; < 2% of bladder malignancies

Natural History & Prognosis

- Usually in advanced stage at time of presentation
  - > 90% of cases show invasion into detrusor muscle (T2)
  - ~ 50% of cases have distant metastasis at time of presentation
- Poor prognosis; 5-year survival:18-47%

Treatment

- Radical cystectomy ± adjuvant chemoradiation
- En bloc resection of urachal tumor and adjacent abdominal wall in cases of advanced urachal adenocarcinoma

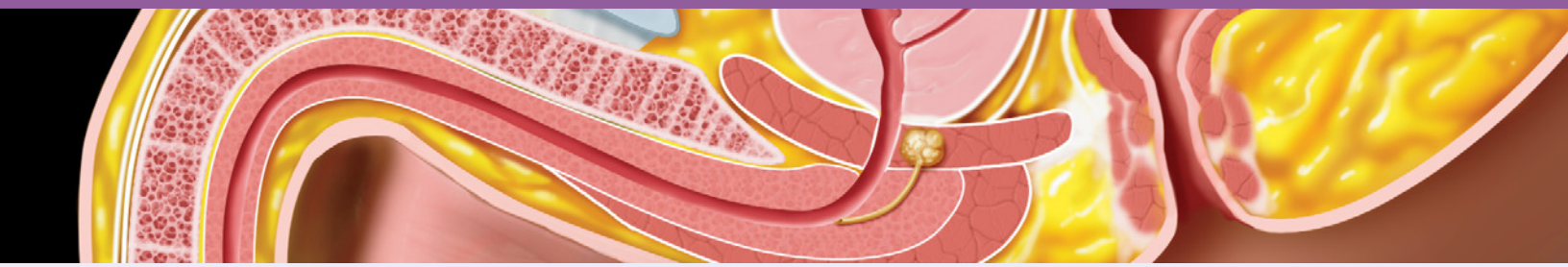
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2. Verma S et al: Urinary bladder cancer: role of MR imaging. Radiographics. 32(2):371-87, 2012
3. Wong-You-Cheong J et al: Neoplasms of the urinary bladder: Radiologic-pathologic correlation. RadioGraphics 26: 553-580',2006

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SECTION 7

Urethra/Penis



Introduction to the Urethra 422

Neoplasms

Urethral Carcinoma 424

Infection

Urethral Stricture 436

Urethral Diverticulum 438

Trauma

Urethral Trauma 442

Erectile Dysfunction 444

Anatomy

Female Urethra

The female urethra is ~ 4 cm in length and follows an oblique course anteroinferiorly from the internal urethral meatus at the bladder neck to the external urethral meatus anterior to the vaginal opening. The urethra passes through the pelvic diaphragm, which is formed by the levator ani and coccygeus muscles and fascia. The levator ani separates the pelvic cavity from the perineum. Inferior to the levator ani, the urethra is surrounded by the urogenital diaphragm, which is formed by the sphincter urethrae and deep transverse peroneus muscles and their fascia. Due to its short course, traumatic injury isolated to the female urethra is extremely rare.

Male Urethra

The male urethra is ~ 18-20 cm in length and has 2 major divisions: The **anterior** and **posterior urethra**, each of which can be further subdivided into 2 parts. The posterior urethra is composed of the prostatic and membranous portions, and the anterior urethra is composed of the bulbous and penile portions.

The **prostatic urethra** begins at the bladder neck and extends to the apex of the prostate gland. The **membranous urethra** traverses the urogenital diaphragm. It is the shortest portion (~ 1 cm) of the urethra but the area most vulnerable to injury, especially in the setting of lower abdominal blunt trauma resulting in pelvic fractures. The **bulbous urethra** extends from the inferior border of the urogenital diaphragm to the penoscrotal junction. The bulbous urethra passes beneath the inferior margin of the symphysis pubis and may be disrupted with a straddle injury. The **penile urethra** is distal to the penoscrotal junction and travels through the pendulous portion of the penis within the corpus spongiosum. It widens into the fossa navicularis at the distal glans.

The **verumontanum** is a 1 cm ovoid mound of smooth muscle along the posterior wall of the prostatic urethra. The prostatic utricle (an embryologic remnant of the Müllerian system) enters in the center of the verumontanum, along with the ejaculatory ducts, which are just distal on either side.

Cowper glands are located within the urogenital diaphragm, but their ducts course distally ~ 2 cm to enter the bulbous urethra. Multiple small mucosal glands (glands of Littré) line the mucosa of the anterior urethra.

Imaging Techniques and Indications

Retrograde Urethrogram

A retrograde urethrogram (RUG) is the study of choice for evaluating the male urethra and is most often used in the setting of acute trauma. It is also the study of choice for evaluating for the sequelae of trauma including strictures and fistulae formation.

Proper positioning is very important when performing a RUG. The penis should be imaged in an oblique position to visualize the full length of the urethra, avoid overlapping structures, and optimize evaluation of the membranous urethra (the most common site of urethral injury in the setting of blunt abdominal trauma). If the clinical history is a straddle injury, the examination should be focused on the bulbous portion of the urethra.

It is also imperative to get adequate distention of the urethra. Inadequate distention may result in the failure to diagnose a urethral tear. This is particularly true when a partial tear is present.

Voiding Cystourethrogram

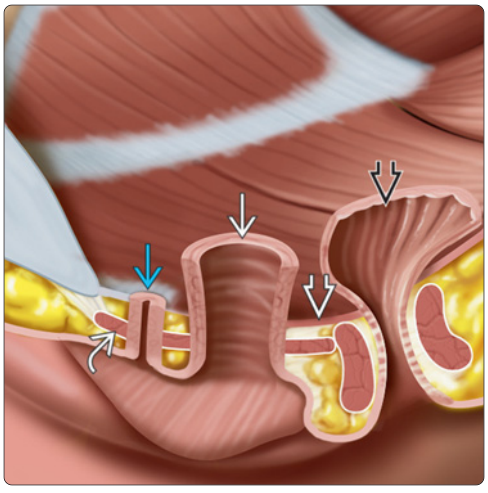
The urethra can also be evaluated with a voiding cystourethrogram (VCUG). This is most often used to evaluate for diverticula of the female urethra and obstructing valves in the male urethra.

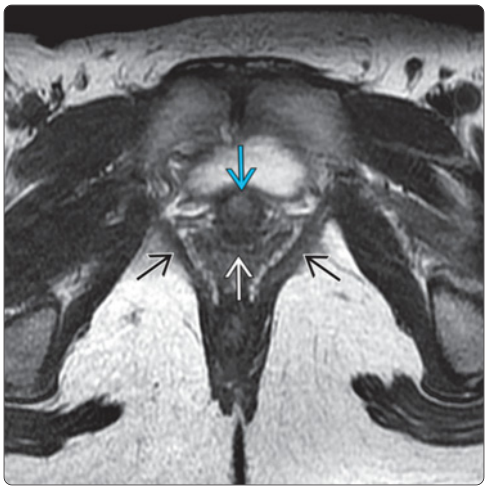
If a patient has a suprapubic catheter placed for trauma, a combined cystogram and RUG can be performed prior to reparative surgery to determine the length of injured urethra.

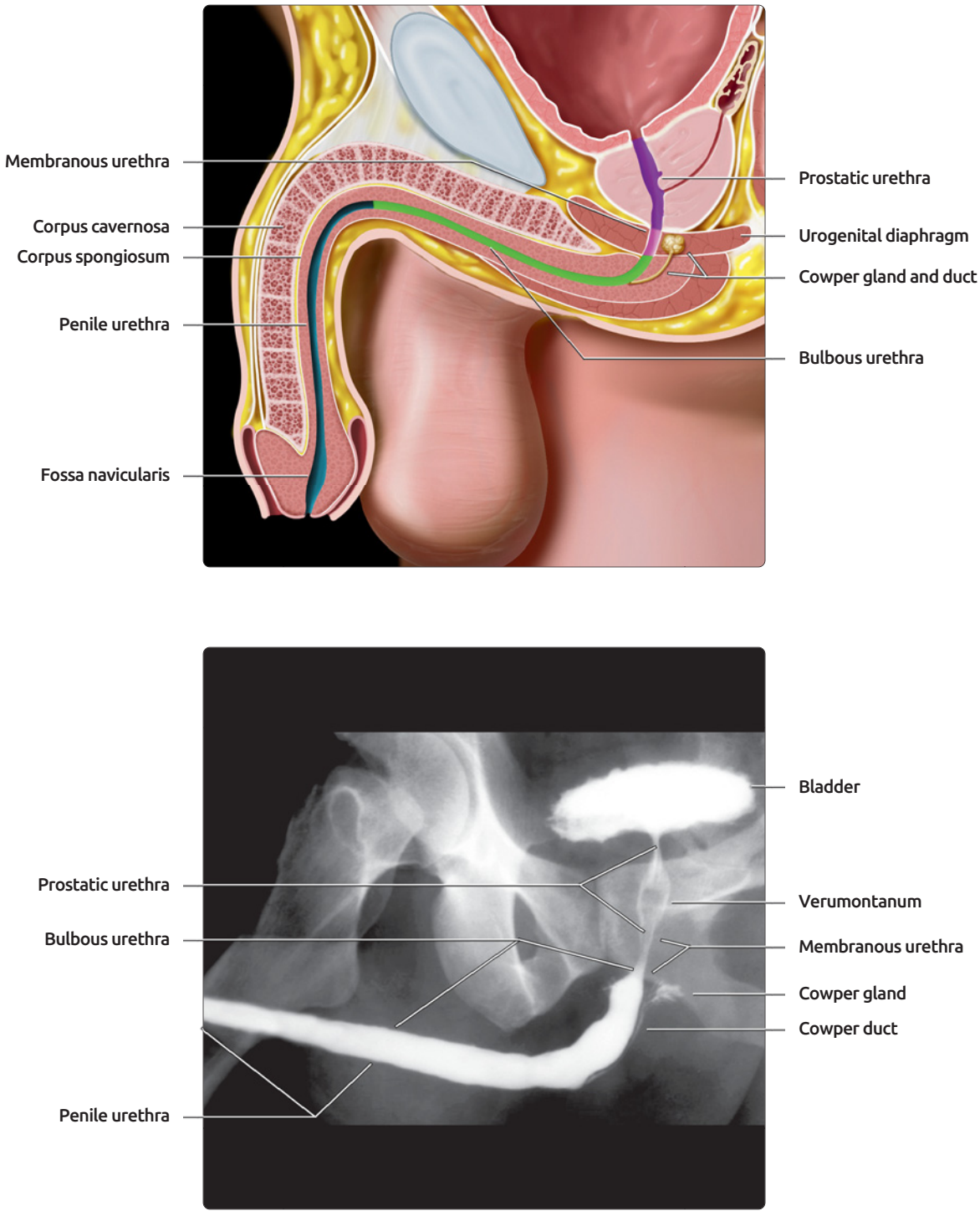
MR

T2WI MR sequences provide the best images of the urethra and are particularly helpful for demonstrating the female urethra, as it is poorly evaluated on other imaging studies. T2WI with fat saturation is the best technique when evaluating for urethral diverticula. T2WI and post-contrast T1WI with fat suppression are key sequences for evaluating local invasion of urethral carcinoma.

(Left) *Sagittal graphic of the female pelvis shows the urethra, vagina, and rectum passing through the pelvic diaphragm, which separates the pelvic cavity from the perineum. The urethra and vagina then pass through the muscular urogenital diaphragm.* **(Right)** *Axial T2WI MR shows the female urethra as a round, intermediate signal intensity structure anterior to the vagina. The levator muscles are shown well laterally.*







(Top) *The male urethra is divided into 2 large segments: The posterior urethra, which is composed of the prostatic (purple) and membranous (pink) portions, and the anterior urethra, which is composed of the bulbous (green) and penile (blue) portions. The membranous urethra is the shortest segment; it passes through the urogenital diaphragm, and is a vulnerable site for injury.* **(Bottom)** *Retrograde urethrogram shows the segments of the urethra. The verumontanum is seen as a smooth filling defect along the posterior wall of the prostatic urethra. The paired Cowper glands lie within the urogenital diaphragm, but their ducts extend ~ 2 cm distal to enter the bulbous urethra.*

T | Primary Tumor

TNM	Definitions
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Ta	Noninvasive papillary, polypoid, or verrucous carcinoma
Tis	Carcinoma in situ
T1	Tumor invades subepithelial connective tissue
T2	Tumor invades any of the following: Corpus spongiosum, prostate, periurethral muscle
T3	Tumor invades any of the following: Corpus cavernosum, beyond prostatic capsule, anterior vagina, bladder neck
T4	Tumor invades other adjacent organs
Urothelial (Transitional Cell) Carcinoma of the Prostate	
Tis pu	Carcinoma in situ, involvement of prostatic urethra
Tis pd	Carcinoma in situ, involvement of prostatic ducts
T1	Tumor invades urethral subepithelial connective tissue
T2	Tumor invades any of the following: Prostatic stroma, corpus spongiosum, periurethral muscle
T3	Tumor invades any of the following: Corpus cavernosum, beyond prostatic capsule, bladder neck (extraprostatic extension)
T4	Tumor invades other adjacent organs (invasion of bladder)

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in single lymph node ≤ 2 cm in greatest dimension
N2	Metastasis in single node > 2 cm in greatest dimension or in multiple nodes

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis

Adapted from 7th edition AJCC Staging Forms (2010).

G | Histologic Grade

TNM	Definitions
World Health Organization/International Society of Urologic Pathology (WHO/ISUP) Grading System	
LG	Low grade
HG	High grade
General Histologic Grading System	
GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

Adapted from 7th edition AJCC Staging Forms (2010).

AJCC | Stages/Prognostic Groups

Stage	T	N	M
0a	Ta	N0	M0
0is	Tis	N0	M0
	Tis pu	N0	M0
	Tis pd	N0	M0
I	T1	N0	M0
II	T2	N0	M0
III	T1	N1	M0
	T2	N1	M0
	T3	N0	M0
	T3	N1	M0
IV	T4	N0	M0
	T4	N1	M0
	Any T	N2	M0
	Any T	Any N	M1

Adapted from 7th edition AJCC Staging Forms (2010).

Incidence of Primary Urethral Carcinoma

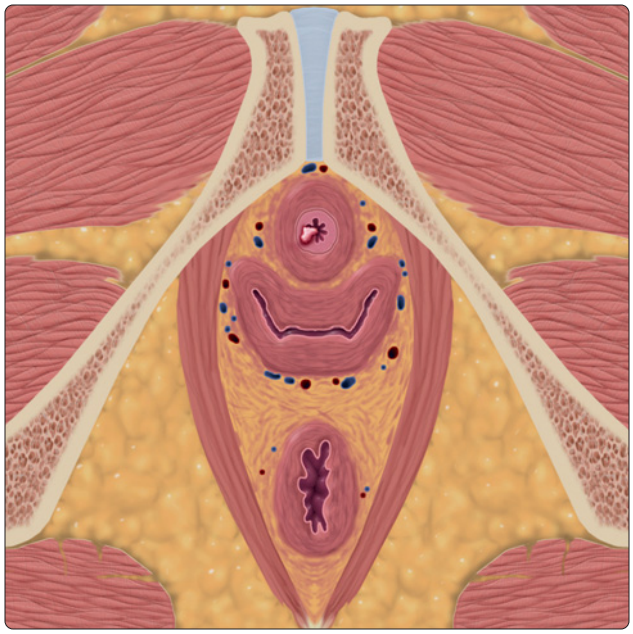
Histologic Subtype	Incidence in Males	Incidence in Females
Transitional cell carcinoma	70.2%	33.9%
Squamous cell carcinoma	18.4%	31.1%
Adenocarcinoma	11.4%	35.0%

Observed and Overall 1- and 5-Year Survival Rates

Stage	1-Year Survival	5-Year Survival
0a	97%	79%
0is	93%	62%
I	90%	59%
II	82%	51%
III	79%	28%
IV	59%	22%

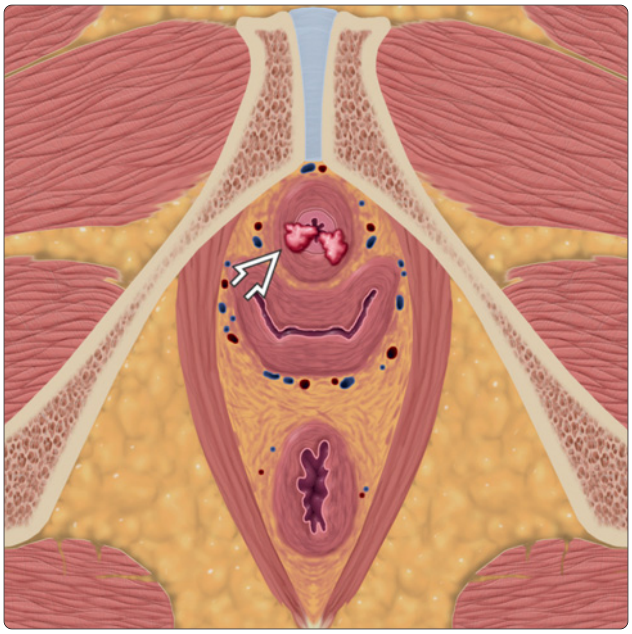
Adapted from 7th edition AJCC Staging Forms (2010).

T1: Female



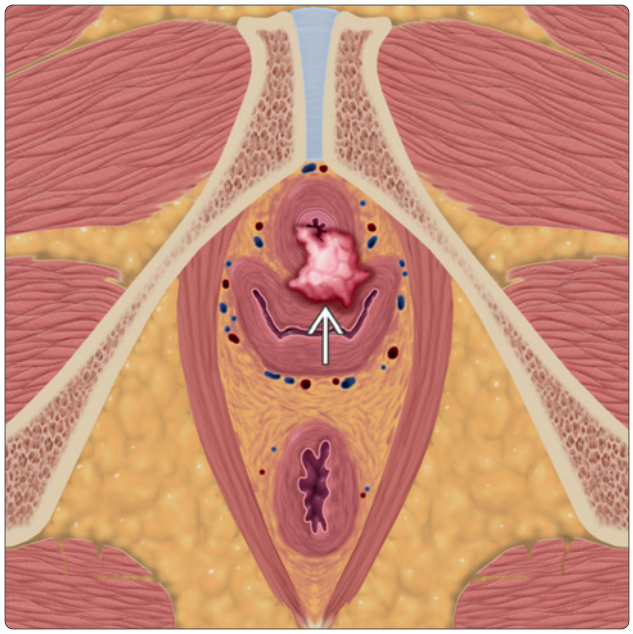
Axial illustration demonstrates T1 disease, which is limited to focal tumor with submucosal invasion. Periurethral tissue is not involved. It is unlikely that this minimal disease burden would be visible on imaging.

T2: Female



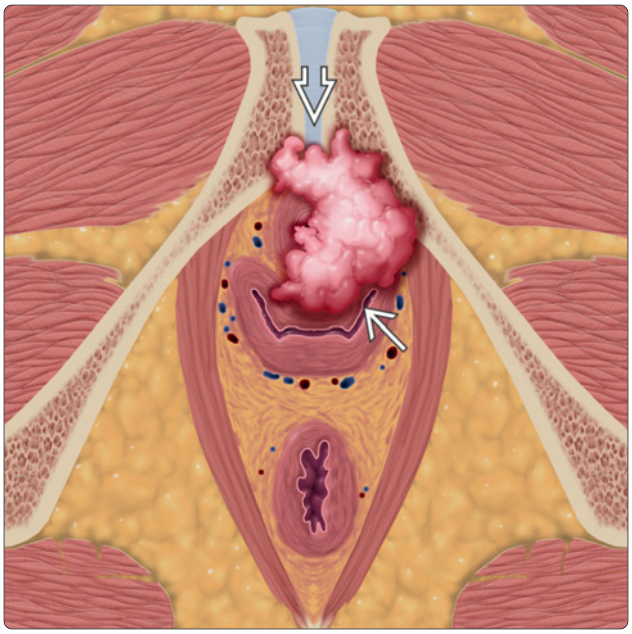
Axial illustration demonstrates T2 disease, which is centered in the urethra and extends through the subepithelial tissue into the periurethral muscle ➪ layer.

T3: Female



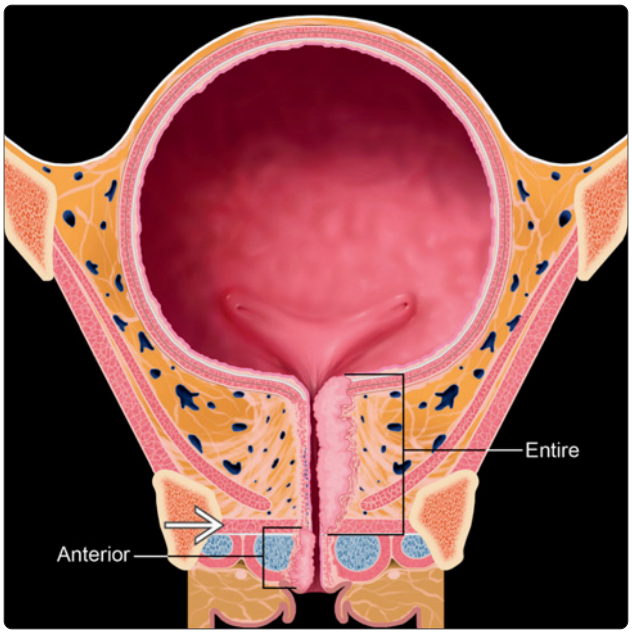
Axial illustration demonstrates T3 disease, which extends through the periurethral muscle into the adjacent anterior vagina ➪.

T4: Female



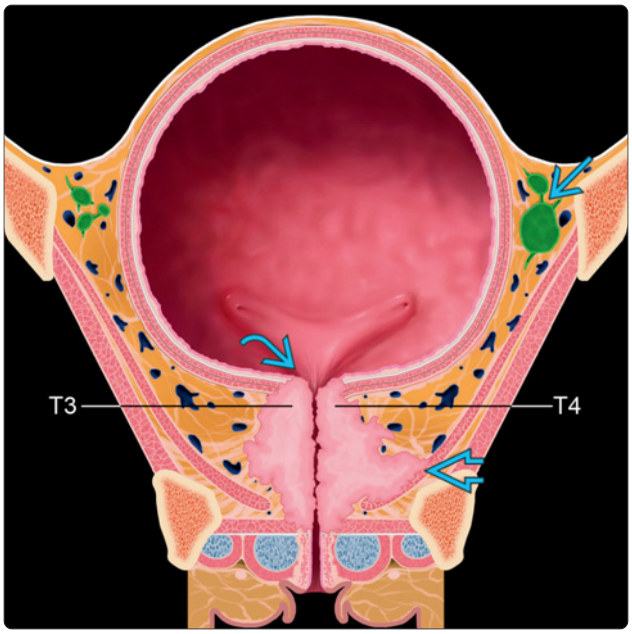
Axial illustration demonstrates T4 disease that is locally advanced and involves both the anterior vagina ➪ and the pubic symphysis ➪.

T1 and T2: Female



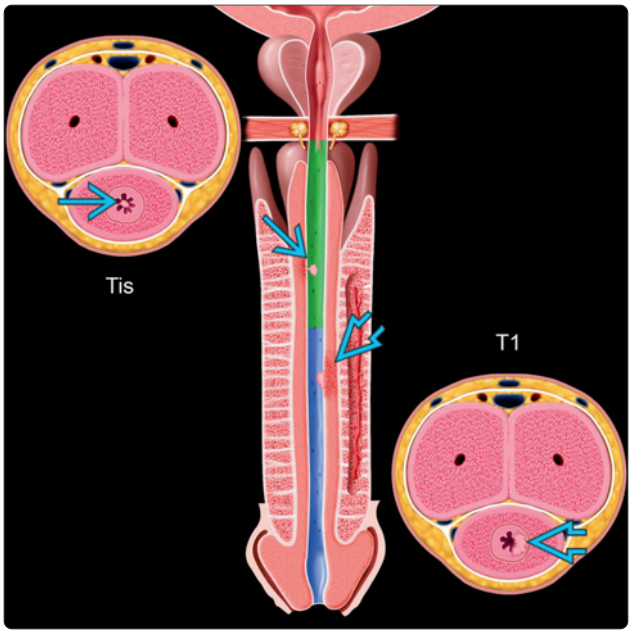
Coronal illustration of the bladder shows primary urethral cancer. The distribution of disease is divided into the anterior and entire urethra. Anterior urethral cancer is limited to the distal 1/3 of the urethra, external to the urogenital diaphragm ➡. Entire urethral cancer is usually high grade and locally advanced.

T3 and T4: Female



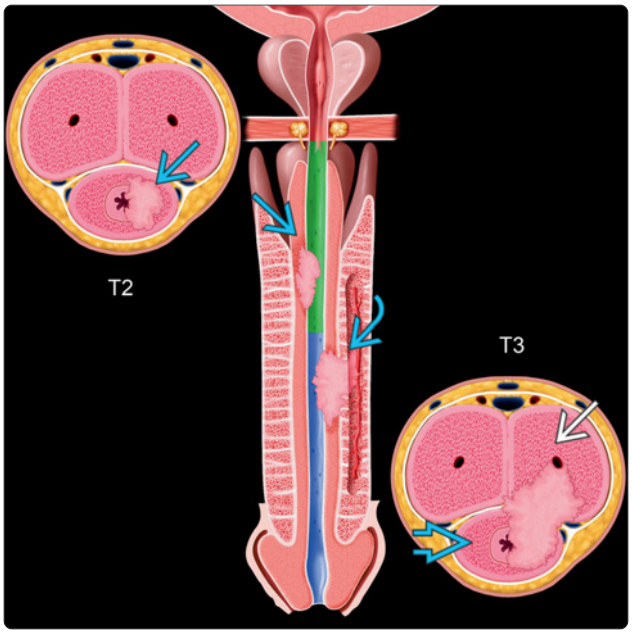
Coronal illustration of the bladder shows T3 disease on the left. It extends into the periurethral soft tissues and may involve the bladder neck ⇨. T4 disease (right) involves the entire urethra and periurethral muscles ⇨. N1 disease is illustrated by the enlarged regional lymph node ⇨, which measures < 2 cm.

Tis and T1: Male



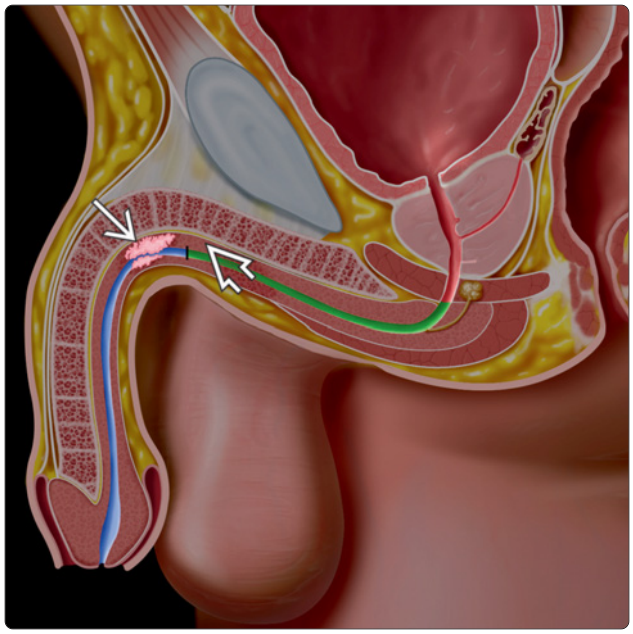
Primary urethral cancer in a male is divided into anterior (penile urethra in blue) and posterior (bulbomembranous urethra in green) distributions. On the left, Tis disease ⇨ is papillary and localized, without submucosal invasion. On the right, T1 disease ⇨ extends into the submucosal layer.

T2 and T3: Male



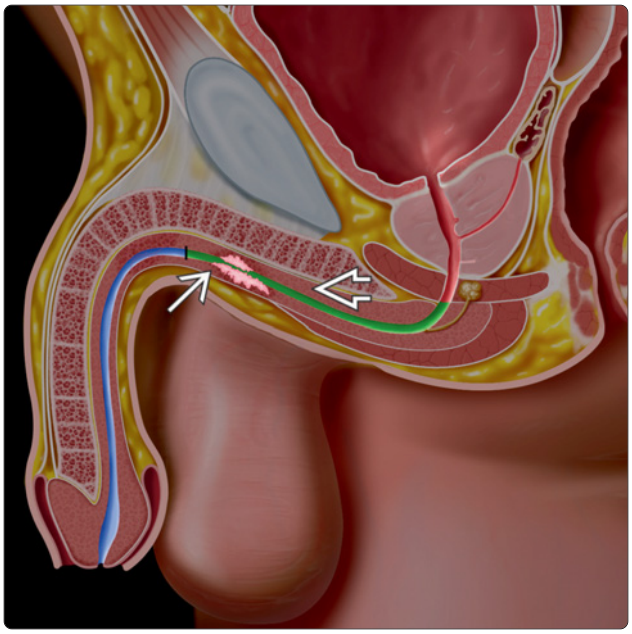
On the left, T2 disease extends into the corpus spongiosum ⇨. On the right, T3 disease ⇨ extends beyond the corpus spongiosum ⇨ into the corpus cavernosum ➡. If prostatic urethra is involved, the etiology is more often prostatic or bladder cancer extending into prostatic urethra.

T2: Male



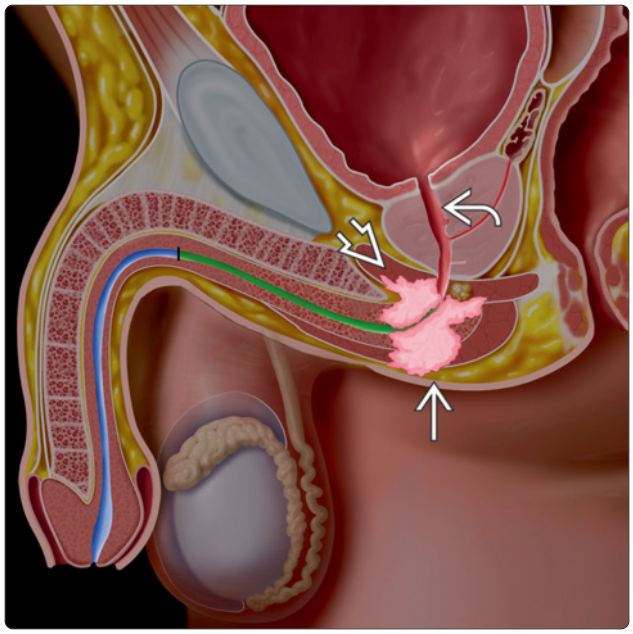
Sagittal illustration through the bladder and penis demonstrates anterior disease ➡ in the penile urethra (blue). Disease extends into the corpus spongiosum ➡, making this at least T2/stage II disease.

T2: Male



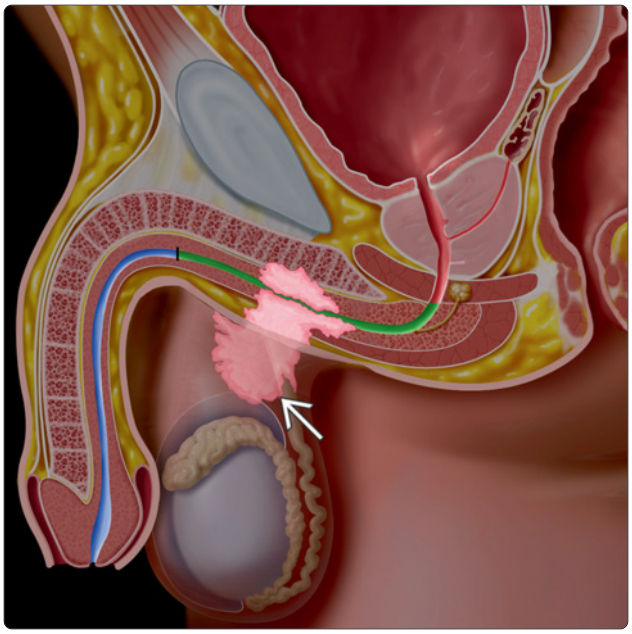
Sagittal illustration through the bladder and penis demonstrates posterior disease ➡ centered in the bulbous urethra (green). Again, there is disease demonstrated that extends into the corpus spongiosum ➡, resulting in at least T2/stage II classification.

T3: Male



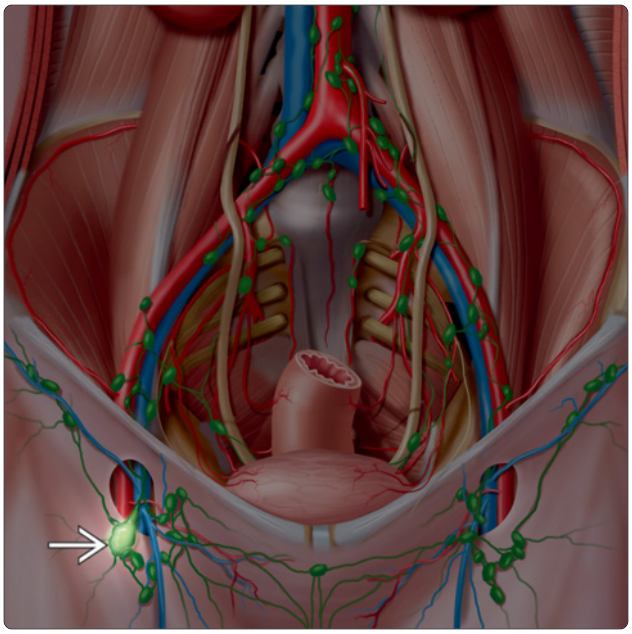
Locally advanced disease in the bulbomembranous urethra (green) is shown. Tumor invades periurethral muscle, urogenital diaphragm ➡, and perineal tissues ➡. Note the normal prostatic urethra ➡, which is consistent with the tumor arising primarily from the urethra rather than from bladder or prostate.

T4: Male



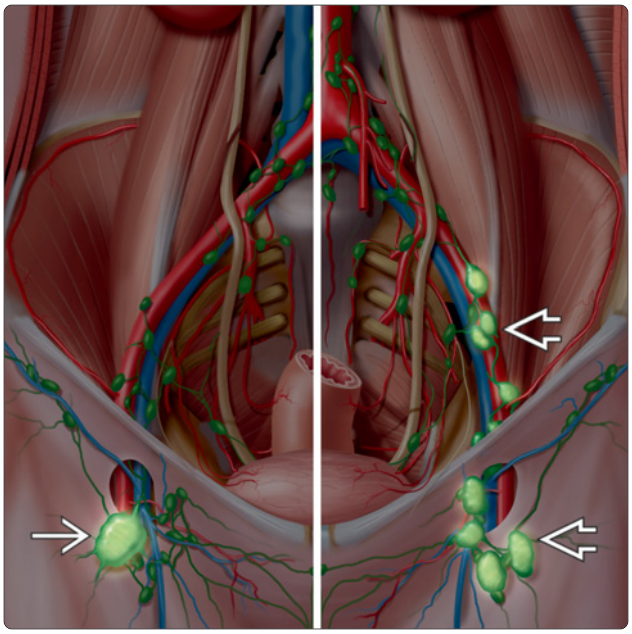
Sagittal illustration through the bladder and perineum demonstrates locally advanced posterior disease in the bulbomembranous urethra (green). This lesion extends beyond the corpus spongiosum to the scrotum ➡. A patient may present with a nonhealing scrotal ulcer in such cases.

N1



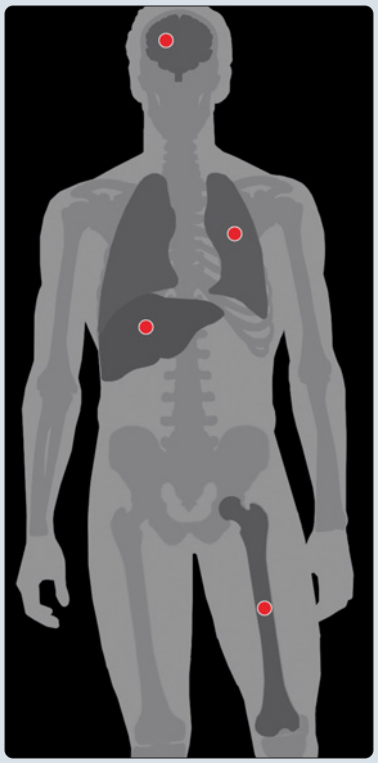
A single enlarged superficial inguinal node ➔ is present that measures ≤ 2 cm. The presence of N1 disease upgrades disease severity to stage III.

N2



On the left, a single enlarged superficial inguinal node ➔ measures more than 2 cm. On the right, multiple nodes and multiple nodal stations are involved ➔. These are both examples of N2 disease. The presence of N2 disease upgrades disease severity to stage IV.

Metastases, Organ Frequency



Liver	*Unknown*
Lung	*Unknown*
Bone	*Unknown*
Brain	*Unknown*

Urethral carcinoma is so uncommon that the frequency distribution of metastases is unknown. Most metastatic disease is to local lymph nodes.

Urethral Carcinoma

OVERVIEW

Classification

- Women: Location is **anterior** or **entire**
  - Anterior tumors exclusively in distal 1/3 of urethra
    - 46% of urethral tumors
    - Squamous cell carcinoma most common
  - Entire urethral tumors usually high grade and locally advanced
- Men: Location is **posterior** or **anterior**
  - Posterior: Prostatic urethra (10%)
  - Posterior: Bulbomembranous urethra (60%)
  - Anterior: Penile urethra (30%)

PATHOLOGY

Routes Spread

- Local spread
  - In women: Local extension
    - Bladder neck, vagina, vulva
  - In men: Infiltrate via vascular spaces of corpus spongiosum and periurethral tissues
    - Anterior tumors may infiltrate to skin surface
    - Posterior/bulbomembranous tumors spread locally to perineum
- Lymphatic metastases to regional nodes
  - Anterior urethral cancer nodal distribution
    - Superficial and deep inguinal nodes
    - Occasionally external iliac nodes
  - Posterior urethral cancer nodal distribution
    - Pelvic nodes
  - Palpable inguinal nodes occur in 20%
    - Usually indicative of metastatic disease
  - Female cancer often spreads systemically without regional nodal disease
- Hematogenous spread uncommon except in
  - Primary transitional cell carcinoma (TCC) of prostatic urethra
  - Advanced local disease

General Features

- Comments
  - Prognosis depends on anatomic location and depth of invasion
    - Histology less important predictor
  - Superficial tumors may be resectable
- Etiology
  - Urethral stricture
    - Present in 1/2 of patients with primary urethral carcinoma
      - Insidious onset of disease
      - Requires high index of clinical suspicion
  - History of sexually transmitted diseases
    - Approximately 1/4 of patients with primary urethral carcinoma
  - Chronic urinary tract infections
  - Age > 60 years
  - History of bladder cancer
    - If cystectomy done for bladder carcinoma → 10% will develop urethral cancer
    - Present in 30% of cystoprostatectomy specimens for bladder TCC
- Epidemiology cancer
  - Uncommon, < 1% of all urothelial malignancies
    - Associated with poor outcomes
  - M:F = 1:4
    - Only urologic malignancy more common in women
  - Usually occurs after 50 years of age

Gross Pathology

- Anterior urethral lesions more amenable to surgical resection
- Localized, early stage tumor may present as papillary mass expanding urethra
- Large lesions may ulcerate

Microscopic Pathology

- H&E
  - Most common histologic subtypes are transitional cell carcinoma (TCC), squamous cell carcinoma (SCCa), and adenocarcinoma (AdenoCa)
    - In males
      - TCC most common in prostatic urethra
      - SCCa more common in bulbomembranous and penile urethra
      - Undifferentiated subtype associated with bulbomembranous urethra
      - AdenoCa from glands of Littré or Cowper
    - In females
      - SCCa > TCC > AdenoCa
      - Clear cell AdenoCa most common subtype in urethral diverticulum
      - Uncommon tumors include undifferentiated, sarcoma, melanoma, and metastases

IMAGING FINDINGS

Detection

- **Primary evaluation: Examination under anesthesia**
  - Cystoscopy
  - Bimanual examination
    - External genitalia, urethra, rectum, perineum
    - Flexible sigmoidoscopy if rectal involvement suspected
  - Transurethral or needle biopsy
- **Retrograde cystourethrography**
  - Standard imaging technique for morphologic and functional urethral evaluation
  - Tumor may be incidental finding during evaluation of stricture disease
    - Infiltrating lesions associated with tight stenoses
    - Often difficult or impossible to opacify urethral lumen above lesion
- **Voiding cystourethrogram**
  - Urethral strictures or filling defects
- **MR**
  - Best for depicting local extent of disease due to superior soft tissue resolution
- **Ultrasound**
  - Transperineal, transvaginal, or transrectal
    - Endourethral US with catheter-based transducer

- Hypo- to isoechoic
- Irregularly marginated urethral mass

Staging

- **CECT**
  - Limited utility for local disease
    - Difficult to differentiate urethra from vagina/bladder base
    - May appear as homogeneously or heterogeneously enhancing mass
  - Local soft tissue extension
  - Pelvic lymph node involvement
  - Extension into adjacent bony structures
- **MR**
  - T1WI
    - Low signal intensity mass, difficult to differentiate from urethra
  - T2WI
    - Urethral wall: Signal intensity similar to muscle
    - Urethral cancer
      - Relatively high signal intensity mass disrupting female urethral "target-like" zonal anatomy; high signal intensity mass invading corpora cavernosa
  - T1WI C+
    - Variable enhancement
    - Best for urethra and periurethral tissue evaluation

CLINICAL ISSUES

Presentation

- Urethral bleeding, serosanguineous discharge
- Palpable urethral mass
- Obstructive voiding symptoms
- Urethral fistula, periurethral abscess
- Perineal pain

Cancer History

- Female: Anterior urethral cancer
  - Much better prognosis, even with local nodal METS
- Female: Entire urethral cancer
  - Commonly locally invasive with pelvic nodal METS
  - Prospects for cure low unless lesions small
    - If < 2 cm, 5-year survival (60%)
    - If > 4 cm, 5-year survival (13%)
- Male: Anterior urethral cancer
  - Potential for cure high with superficial disease
- Male: Posterior urethral cancer
  - Prospects for cure low, high local recurrence rates
  - 5-year survival from 15-20%

Treatment Options

- Alternatives
  - Surgical treatment
    - Options depend on location and extent of disease
    - In men, no demonstrated benefit from prophylactic inguinal lymph node dissection
  - Adjunctive or neoadjuvant radiation therapy
  - Chemotherapy: In clinical trials for metastatic urethral cancer
    - Urethral TCC may respond to same chemotherapy regimens as advanced bladder TCC
- Options by stage
  - Female: Anterior urethral cancer
    - Stage 0/Tis, Ta: Open excision, **or** electroresection & fulguration, **or** laser vaporization-coagulation
    - Stage T1 & T2: External beam radiation therapy, **or** interstitial radiation, **or** combination, **or** surgical resection of distal 1/3 of urethra
    - T3/recurrent lesions: Anterior exenteration & urinary diversion ± preoperative radiation & urinary diversion
    - Palpable inguinal nodes: Frozen section tumor confirmation **then** ipsilateral node dissection
  - Female: Entire urethral cancer
    - Preoperative radiation, anterior exenteration & urinary diversion with bilateral pelvic node dissection ± inguinal node dissection
    - If < 2 cm in greatest dimension, radiation alone, **or** nonextensive surgery alone, **or** combination may be sufficient
  - Male: Anterior urethral cancer
    - Stage 0/Tis, Ta: Open excision, **or** electroresection & fulguration, **or** laser vaporization-coagulation
    - T1, T2, T3: Partial penectomy, negative margins to 2 cm proximal to tumor
    - T1, T2, T3 (penectomy refused): Radiation
    - Palpable inguinal nodes: Frozen section tumor confirmation **then** ipsilateral node dissection
  - Male: Posterior urethral cancer
    - Preoperative radiation, cystoprostatectomy, urinary diversion, & penectomy with bilateral pelvic node dissection ± inguinal node dissection
  - Female/male: Recurrent urethral cancer
    - Local recurrence after radiation: Surgical excision
    - Local recurrence after surgery alone: Combination radiation & wider surgical resection

REPORTING CHECKLIST

T Staging

- Female
  - Assess for tumor beyond periurethral muscle; assess for tumor in relation to bladder base
- Male
  - Assess tumor extension beyond corpus spongiosum
  - Determine relation of tumor to prostatic urethra
    - Tumor in prostatic urethra more likely prostate than urethral cancer

N Staging

- Size and number of nodes most significant

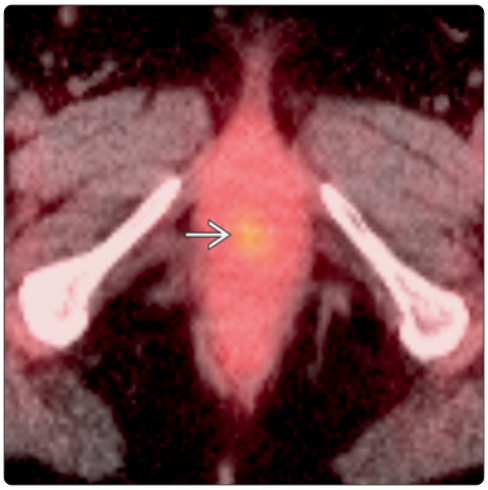
M Staging

- Lung, liver, bone, brain

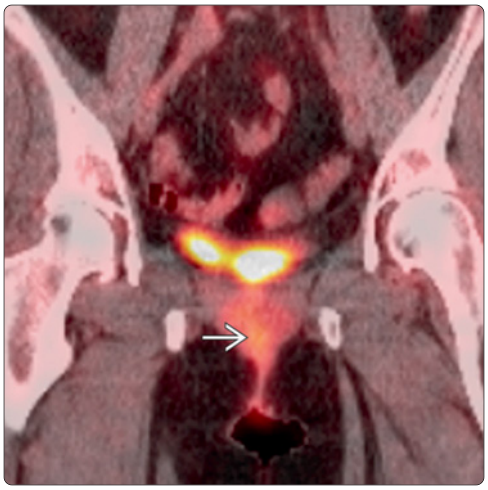
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4. Kawashima A et al: Imaging of urethral disease: a pictorial review. Radiographics. 24 Suppl 1:S195-216, 2004

Stage I (T1 N0 M0)

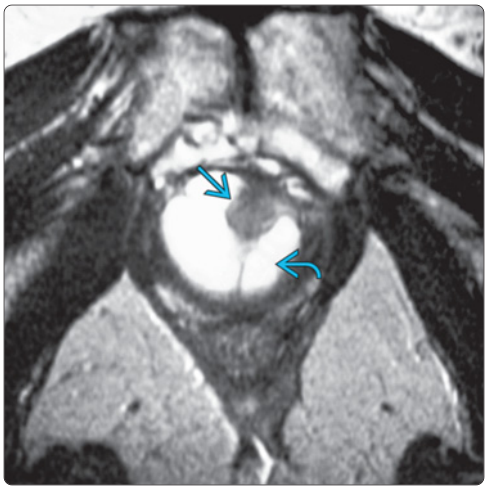


Stage I (T1 N0 M0)

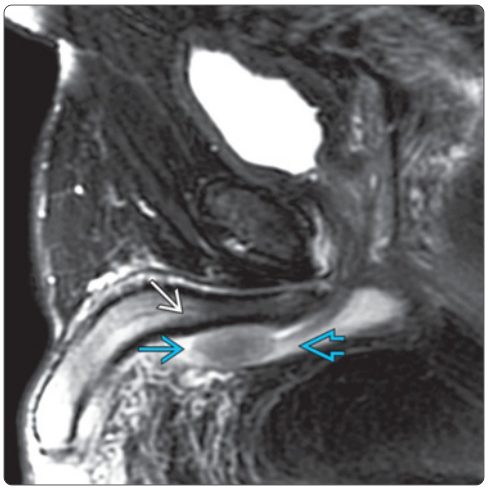


(Left) *Axial PET/CT demonstrates minimal increased activity in the urethra. Although this finding can be seen in normal mucosa, this patient had a < 1 cm anterior urethral mass, which was easily removed by local excision.* **(Right)** *Coronal PET/CT demonstrates minimally increased activity in the urethra, corresponding to the known site of disease. No discrete lesion was identified on imaging, nor were additional sites of disease identified.*

Stage I (T1 N0 M0)

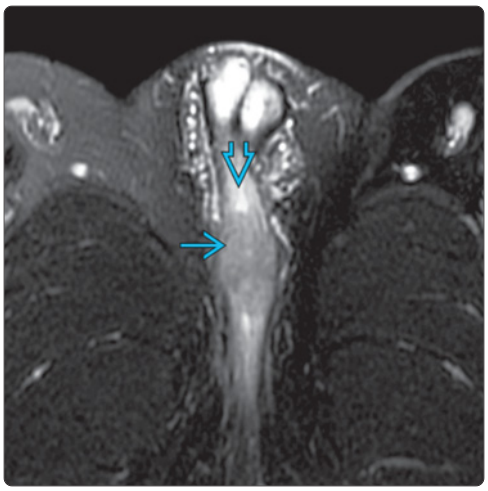


Stage II (T2 N0 M0)

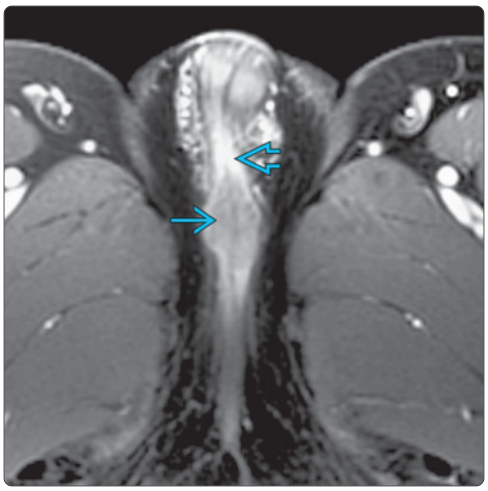


(Left) *Axial T2WI MR demonstrates a soft tissue nodule within a urethral diverticulum. The depth of invasion is difficult to assess. On resection the lesion depth was limited to the subepithelial connective tissue layer.* **(Right)** *Sagittal T2WI FS MR in a male patient demonstrates a low T2 SI expansile ureteral mass in the bulbous portion of the urethra. It involves the corpus spongiosum, but the corpus cavernosum is spared. (Courtesy M. Lockhart, MD, MPH.)*

Stage II (T2 N0 M0)

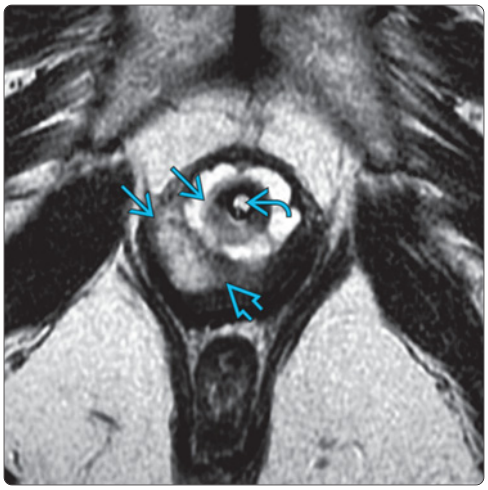


Stage II (T2 N0 M0)

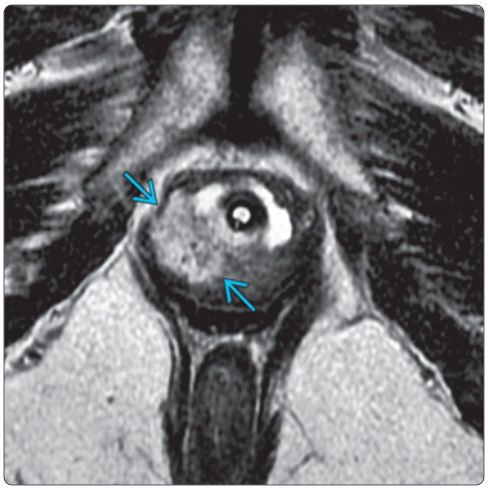


(Left) *Axial T2WI FS MR in the same patient demonstrates a hypointense mass centered in the bulbous urethra with expansion into the surrounding corpus spongiosum. (Courtesy M. Lockhart, MD, MPH.)* **(Right)** *Axial T1WI C+ FS MR demonstrates relatively poor enhancement of the tumor compared to the adjacent highly vascularized corpus spongiosum. (Courtesy M. Lockhart, MD, MPH.)*

Stage II (T2 N0 M0)

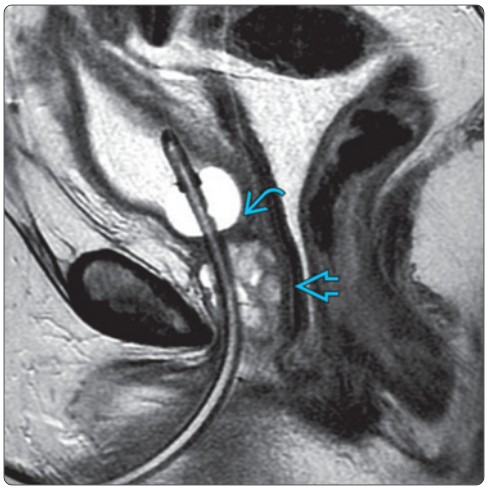


Stage II (T2 N0 M0)

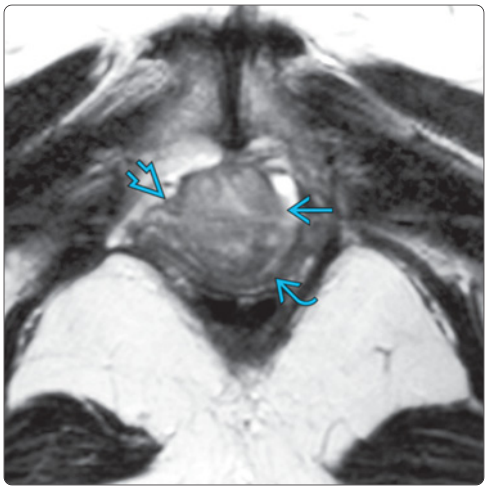


(Left) *Axial T2WI MR demonstrates a relatively low T2 SI mass arising in a female urethral diverticulum, extending from the urethra into the periurethral tissues. A Foley catheter is in place.* **(Right)** *More caudal image in the same patient better demonstrates the extent of the relatively low T2 SI mass in the diverticulum.*

Stage II (T2 N0 M0)

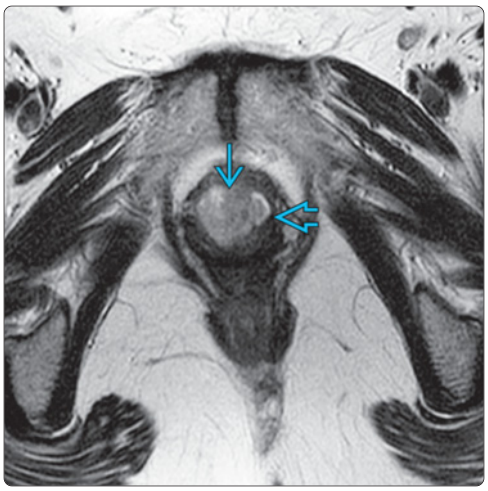


Stage II (T2 N0 M0)

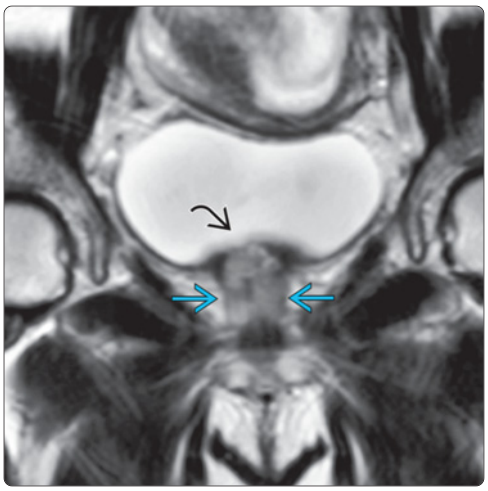


(Left) *Sagittal T2 MR in the same patient demonstrates the confinement of the mass to the urethral tissues with sparing of the bladder base and anterior vagina.* **(Right)** *Axial T2WI MR in a different woman with urethral carcinoma shows a heterogeneously hypointense urethral mass with periurethral extension on the right. The normally low T2 SI urethral wall appears to be intact adjacent to the vagina.*

Stage III (T3 N0 M0)



Stage III (T3 N0 M0)

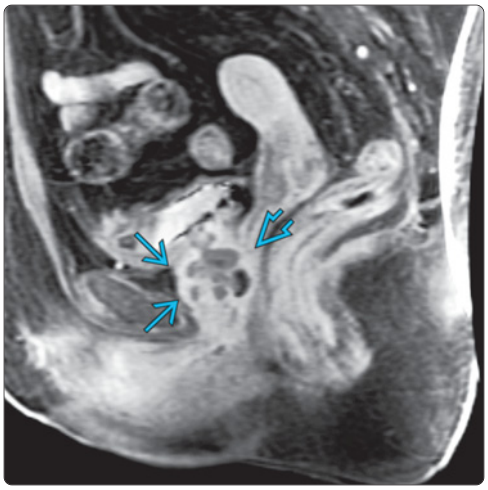


(Left) *Axial T2WI MR shows an intermediate signal intensity urethral mass that expands the urethra. The low T2 SI urethral muscular wall remains intact.* **(Right)** *Coronal T2 MR shows that the entire urethra is expanded by the mass. Superior extension of the mass into the bladder neck upstages this to T3 disease.*

Stage III (T3 N0 M0)

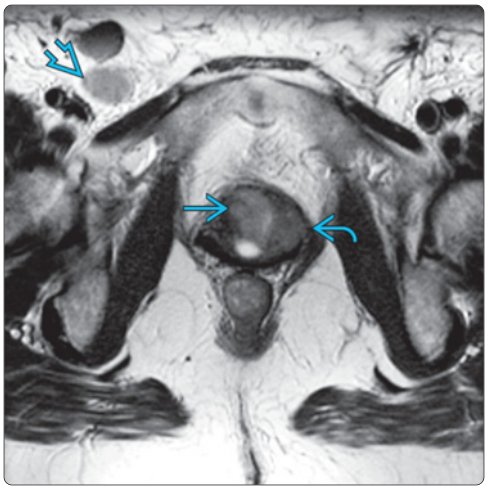


Stage III (T3 N0 M0)

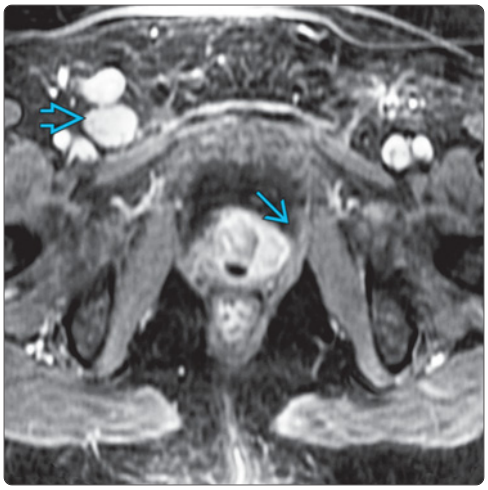


(Left) *Sagittal T2WI MR in the same patient shows an intermediate signal intensity urethral mass that expands the urethra. The superior extension into the bladder neck is well depicted.* **(Right)** *Post-contrast sagittal T1WI FS MR demonstrates a heterogeneously enhancing urethral mass, with increased concern for involvement of the anterior vagina.*

Stage III (T2 N1 M0)

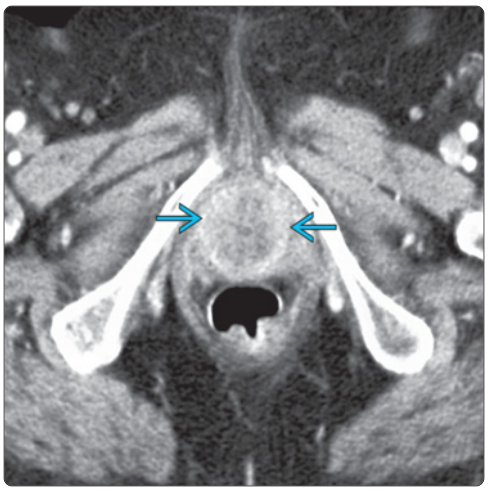


Stage III (T2 N1 M0)

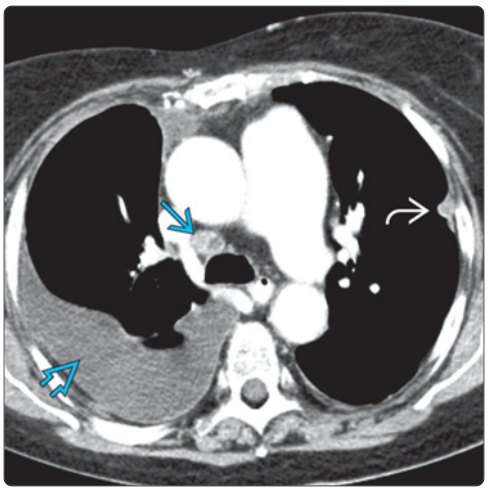


(Left) *Axial T2WI MR demonstrates an intermediate signal intensity urethral mass expanding the female urethra. Periurethral muscle is thinned on the left. Deep periurethral muscle invasion is not seen. An enlarged right inguinal node is present.* **(Right)** *Axial T1WI C+ FS MR shows the heterogeneously enhancing tumor contacting, but not involving, the left periurethral muscle. Metastatic right inguinal adenopathy upstages the disease from stage II to stage III.*

Stage IV (T2 N0 M1)

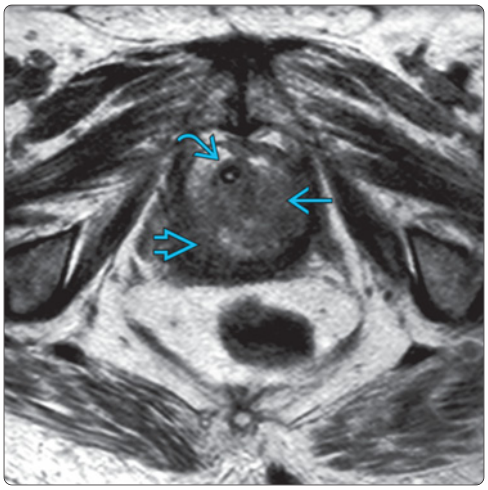


Stage IV (T2 N0 M1)

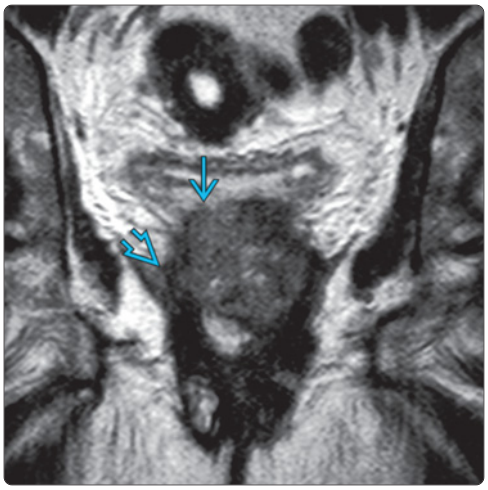


(Left) *Axial CECT shows a heterogeneously enhancing urethral mass; however, the integrity of periurethral tissue planes cannot be determined. Indeed, CT is of limited utility in assessing urethral T stage.* **(Right)** *Axial thoracic CECT in the same patient demonstrates an enlarged enhancing precarinal node, a left pleural nodule, and a right pleural effusion. The extensive thoracic metastatic disease is best demonstrated on CT.*

Stage IV (T4 N0 M0)

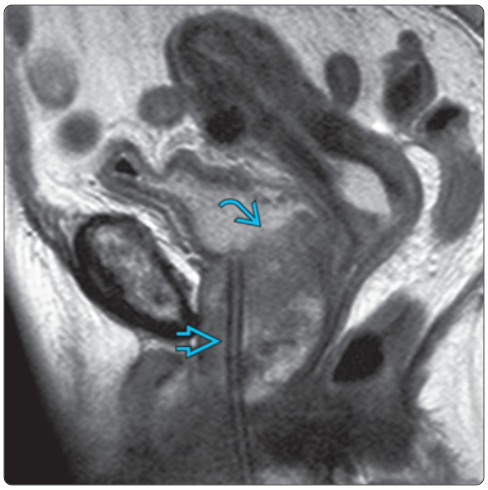


Stage IV (T4 N0 M0)

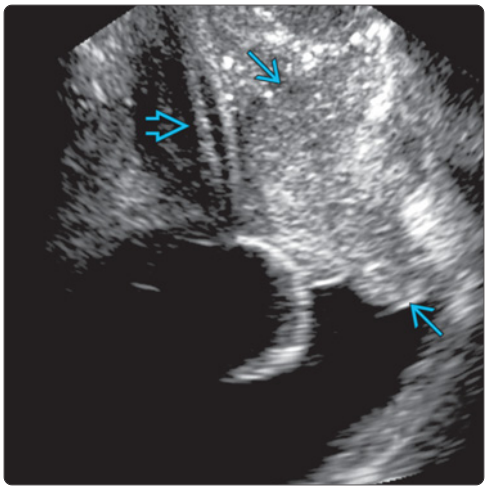


(Left) *Axial T2WI MR demonstrates a heterogeneously hypointense mass expanding a female urethra with loss of a distinct high T2 SI plane in the vesicovaginal space. A Foley catheter is in place.* **(Right)** *Coronal T2WI MR in the same patient demonstrates infiltration beyond the periurethral tissue into the adjacent levator ani muscle complex on the right. Additionally, the mass is seen invading the bladder base.*

Stage IV (T4 N0 M0)

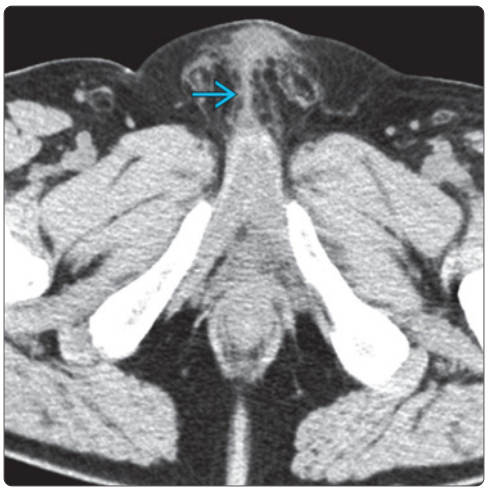


Stage IV (T4 N0 M0)



(Left) *Sagittal T2WI MR in the same patient better demonstrates the heterogeneous low T2 SI mass involving the entire urethra and extending cranially to invade the bladder base. A Foley catheter is present.* **(Right)** *Transperineal ultrasound in the same patient demonstrates the polypoid urethral mass extending into the bladder. A Foley catheter is well demonstrated within the urethra, with the catheter balloon expanded in the bladder.*

Postoperative Appearance



Postoperative Recurrence



(Left) *Axial CECT shows the postoperative appearance of a partial penectomy for anterior stage II disease. At this time, no local recurrence is present. Long-term survival has been reported in subjects who undergo this therapy.* **(Right)** *Axial NECT in a different patient demonstrates a local recurrence after partial penectomy. Recurrence developed along the bulbomembranous urethra.*

Urethral Stricture

KEY FACTS

TERMINOLOGY

- Narrowing of urethra due to scar tissue or tumor

IMAGING

- Strictures have variable appearance
  - Focal, asymmetric indentations
  - Long segmental, tubular, or beaded constrictions
  - Proximal urethral dilatation on urethrography in chronic phase
- Filling of glands of Littré (mucous-secreting glands of urethra)
  - Highly associated with infection or inflammation
- Iatrogenic trauma
  - Located in penoscrotal junction and membranous portion of urethra (most common)
  - Other sites: Meatus and junction with bladder neck
- Retrograde urethrogram and voiding urethrography combined to evaluate proximal and distal stricture

TOP DIFFERENTIAL DIAGNOSES

- Urethral trauma
  - Stretch injury with similar appearance to stricture but in acute setting
- Urethral carcinoma

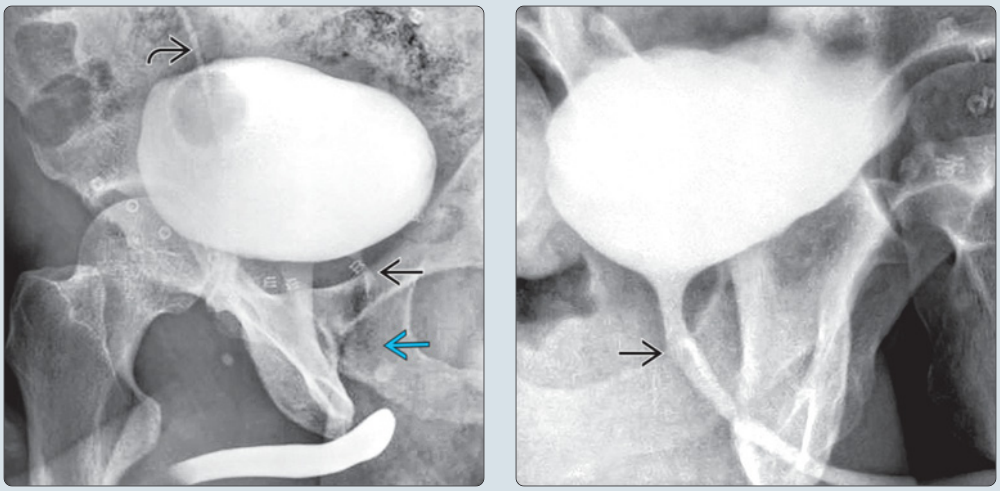
PATHOLOGY

- Etiology: Trauma (most common etiology)
  - Iatrogenic trauma (most common traumatic cause)
  - Infection: Gonorrhea, syphilis (2nd most common), tuberculosis

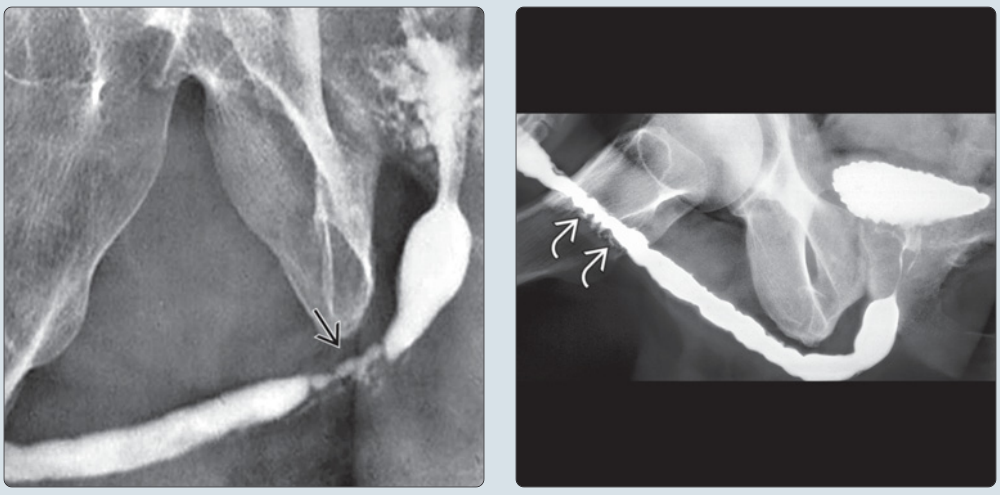
CLINICAL ISSUES

- Treatment: Use 1 or more procedures
  - Dilatation with catheters or balloons
  - Urethrotomy, surgical reconstruction, endoluminal stents
- Complications: Obstruction, abscess, fistula

(Left) *Combined retrograde urethrogram (RUG) & cystogram through a suprapubic catheter was performed 1 month after the patient sustained pelvic trauma. There is filling of the proximal aspect of the prostatic portion of the urethra followed by a severe stricture of the remainder of the posterior urethra.* **(Right)** *Postoperative RUG in the same patient, following the repair of the severe stricture, shows a normal urethra. Trauma is 1 of the leading causes of urethral strictures.*



(Left) *RUG shows an irregular segment of stricture in the bulbous portion of the urethra due to transitional cell carcinoma. Unlike most iatrogenic strictures, malignant strictures have an irregular appearance.* **(Right)** *RUG in a patient with gonococcal urethritis shows irregularity of the penile & bulbous urethra, with filling of the periurethral glands of Littré. This sign is highly associated with an infectious or inflammatory etiology, which frequently progress to stricture formation.*



TERMINOLOGY

Definitions

- Narrowing of urethra due to scar tissue or tumor

IMAGING

General Features

- Best diagnostic clue
  - Focal narrowing of urethra with proximal dilation on urethrography

Radiographic Findings

- Voiding cystourethrography
  - Useful to evaluate entire posterior urethra proximal to severe stricture

Fluoroscopic Findings

- Voiding cystourethrogram
  - Evaluates posterior urethra proximal to severe stricture
- Retrograde urethrogram
  - Focal, asymmetric indentations
  - Long segmental, tubular, or beaded constrictions
  - Marked narrowing to slight irregularity of urethral wall closely proximal &/or distal to main stricture
    - Acute: Dilatation proximal and distal to stricture
    - Chronic: Dilatation proximal to stricture, not distal
  - Filling of glands of Littré (mucous-secreting glands of urethra)
    - Highly associated with infection or inflammation
    - Small amount of reflux into Cowper gland duct
  - Thick-walled bladder (reflects bladder outlet obstruction)
  - Iatrogenic
    - Most commonly located at penoscrotal junction and membranous portion of urethra
    - Other sites: Meatus and junction with bladder neck
    - Rarely entire urethra; diffuse or multifocal
  - Gonorrhea: Located in bulbous portion of urethra or immediately proximal to meatus; multifocal
  - Reiter syndrome: Involves entire penile urethra

Ultrasonographic Findings

- Sonourethrography
  - Used mainly in evaluating anterior strictures
  - Good accuracy in depicting stricture length
  - Staging of complex or preoperative strictures
    - Calculi, urethral hair, false passage, stent encrustation

Imaging Recommendations

- Best imaging tool
  - Retrograde urethrogram ± voiding urethrography
    - Combine to evaluate severe proximal and distal strictures

DIFFERENTIAL DIAGNOSIS

Urethral Trauma

- Acute: Stretching or narrowing; ± pelvic fractures
  - ± extravasation from partial or complete tear
- Blunt trauma → partial or complete urethral tears → post-traumatic stricture

Urethral Carcinoma

- Long, irregular narrowing ± fistula
- Intraluminal filling defect (rare)
- Primary neoplasm (rare): Squamous cell carcinoma from bulbar or bulbomembranous portions
- Secondary neoplasm in or near urethra

PATHOLOGY

General Features

- Etiology
  - Trauma (most common etiology)
    - Iatrogenic trauma (most common traumatic cause)
      - Instrumentation: Cystoscopy
      - Urethral catheters: Straight, indwelling, rubber
      - Surgery: Transurethral resection of prostate, urethroplasty
      - Irradiation: Treat uterine cervical cancer
  - Infection: Gonorrhea, syphilis (2nd most common), tuberculosis
  - Inflammatory disease: Reiter syndrome, amyloidosis
  - Chemical trauma: Injection of corrosive substances
  - Idiopathic

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Obstructive symptoms: Diminished force of stream, ↓ urine flow rate, prolonged voiding, frequency

Demographics

- Age
  - Any age, but incidence ↑ with age
- Gender
  - M > F

Natural History & Prognosis

- Complications: Obstruction, abscess, fistula
- Prognosis: Very good after treatment

Treatment

- Use 1 or more procedures
  - Dilatation with catheters or balloons
  - Internal or external urethrotomy
  - Surgical reconstruction
  - Endoluminal stents

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Urethral Diverticulum

KEY FACTS

TERMINOLOGY

- Focal outpouching of urethra into adjacent fascia
  - Most arise from posterolateral wall of middle 1/3 of urethra

IMAGING

- MR
  - Periurethral cystic lesion that may abut or surround urethra
  - T1WI: Typically low signal intensity (SI)
    - ↑ SI with proteinaceous or hemorrhagic contents
  - T2WI: Typically ↑ SI
    - ↓ SI with proteinaceous or inflammatory debris
  - T1WI post contrast
    - Inflammation/infection: Enhancement of diverticular wall and periurethral tissues
    - Neoplasm: Enhancement of mural nodule or mass
- Voiding cystourethrogram
  - Opacification of diverticulum during voiding of contrast instilled into bladder

TOP DIFFERENTIAL DIAGNOSES

- Bartholin gland cyst
- Gartner cyst
- Skene duct cyst
- Urethral caruncle

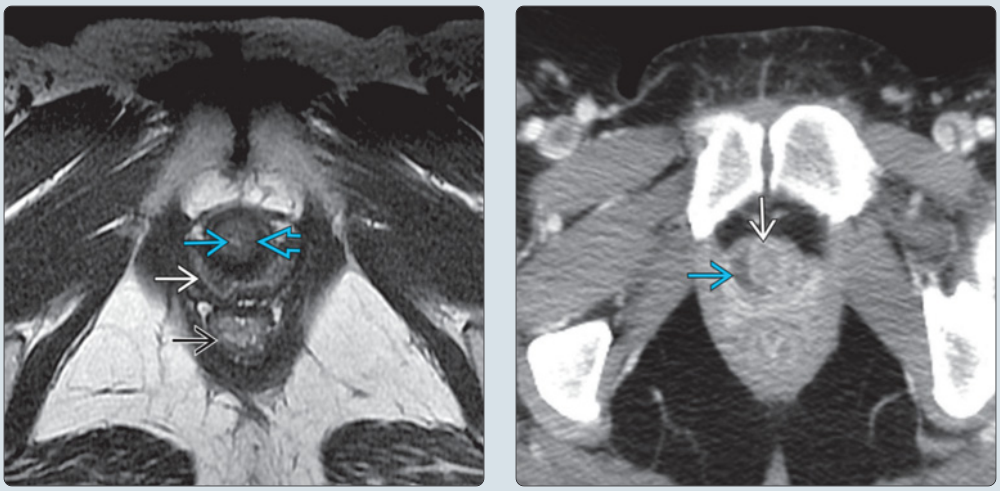
CLINICAL ISSUES

- Nonspecific signs/symptoms: Recurrent/chronic UTI, dysuria, incontinence
- Classic triad of dysuria, dyspareunia, and post-void dribbling is less common
- Intraluminal calculi and neoplasms are rare
- Treatment = diverticulectomy

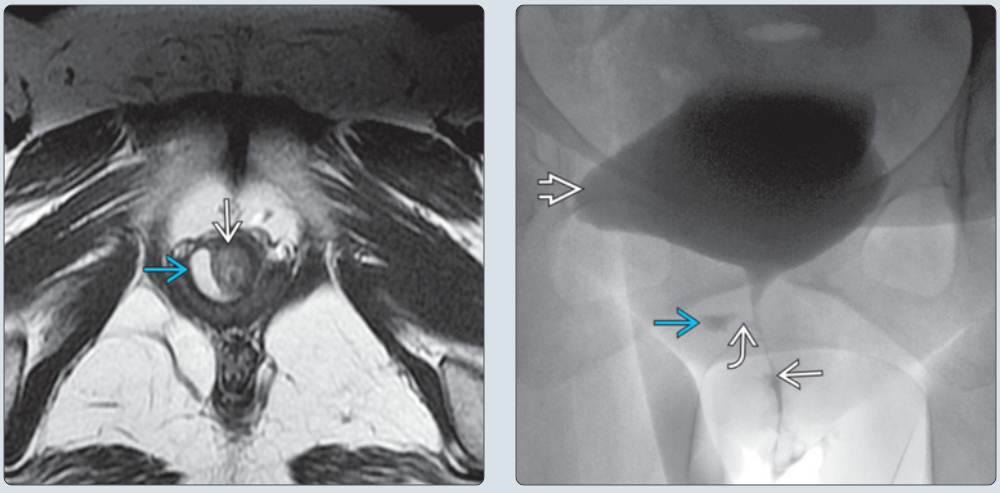
DIAGNOSTIC CHECKLIST

- Describe location, size, number, complexity (septa, mural nodules, calculi)

(Left) *Axial single-shot fast spin-echo T2WI MR at the level of the pubic symphysis demonstrates the normal appearance of the female urethra; the central high signal intensity is due to the mucosa ⇨ while the surrounding intermediate signal intensity is due to the muscular wall ⇨. More posteriorly, the vaginal ➡ and anal ⇨ canals are evident.* **(Right)** *Axial CECT reveals a crescentic, fluid-attenuation structure ⇨ along the right lateral aspect of the urethra ➡ that is due to urethral diverticulum.*



(Left) *Axial single-shot fast spin-echo T2WI MR in the same patient demonstrates homogeneous, high signal intensity within the diverticulum ⇨ that extends along the right lateral aspect of the urethra ➡.* **(Right)** *Anteroposterior view during a voiding cystourethrogram in the same patient shows that the orifice ➡ of the diverticulum ⇨ arises from the right posterolateral wall of the urethra ➡. The catheter used to instill contrast material into the urinary bladder ➡ had been removed prior to voiding.*



TERMINOLOGY

Definitions

- Focal protrusion of urethra into adjacent fascia

IMAGING

General Features

- Best diagnostic clue
  - Cystic lesion adjacent to urethra with high signal intensity on T2WI
- Location
  - Majority of diverticula occur in middle 1/3 of urethra and arise from posterolateral wall
- Size
  - Variable, but generally 1-3 cm
- Morphology
  - Variable: Round, ovoid, U-shaped, circumferential
  - Most diverticula are unilocular
    - Approximately 1/3 are compound: Multiseptated or associated with multiple other diverticula

CT Findings

- NECT
  - Cystic mass at level of pubic symphysis
  - Intraluminal calcifications = stones
- CECT
  - Variable thickness and enhancement of wall, depending on degree of inflammation
  - Enhancing, solid mass suggests malignancy

MR Findings

- T1WI FS
  - Typically, ↓ signal intensity (SI) due to contents of urine or simple fluid
  - ↑ SI may result from proteinaceous, hemorrhagic, or inflammatory contents
- T2WI
  - Typically, ↑ SI due to being filled with urine or simple fluid
  - Internal inflammatory debris, proteinaceous contents, or intraluminal tumor may have ↓ SI
- T1WI C+
  - Brisk enhancement of diverticular wall and adjacent tissues suggests inflammation or infection
  - Enhancement of solid mass suggests malignancy
- MR can be performed with phased-array torso coil or endoluminal coil
  - Endoluminal coils allow increased sensitivity but are also invasive

Ultrasonographic Findings

- Grayscale ultrasound
  - May be performed via transabdominal, transperineal, or transluminal approach
    - Transabdominal and transperineal approaches are insensitive for small (< 1.5-2 cm) diverticula
  - Periurethral cystic structure
    - Anechoic with through transmission when simple
    - Internal echoes, septa, shadowing (calculi) when complex
- Color Doppler
  - Flow can be detected in thickened septa
  - Flow within mural nodule or solid component suggests neoplasm

Other Modality Findings

- Voiding cystourethrography
  - Filling of diverticulum while voiding in standing position
  - Diagnostic only for diverticula with patent orifice
  - May underestimate size and extent if only partially opacified
- Double-balloon urethrography
  - Performed by injecting contrast agent under positive pressure into closed urethral lumen
    - Urethral lumen is closed by dual balloons
    - Injected contrast fills diverticular cavity and its neck
  - Higher sensitivity compared to voiding cystourethrography
  - Not commonly performed any longer
    - Discomfort: Invasive technique with cannulation of urethra
    - Technical difficulty: Leakage of contrast around balloons
- CT voiding urethrography and virtual urethroscopy
  - Performed with high-resolution MDCT during voiding after bladder is distended with contrast
  - Post-processing allows creation of 2D and real-time 3D surface rendering images
  - Useful in preoperative planning prior to diverticulectomy

Imaging Recommendations

- Best imaging tool
  - MR is more sensitive than urethrography and fiberoptic urethroscopy
- Protocol advice
  - Multiplanar MR with pelvic phased-array coil using fat-saturated T2WI and post-contrast sequences

DIFFERENTIAL DIAGNOSIS

Bartholin Gland Cyst

- No communication with urethra
- Located in posterolateral wall of vaginal introitus, medial to labia minora
- Located at, or inferior to, level of pubic symphysis

Gartner Cyst

- No communication with urethra
- Located in anterolateral wall in proximal 3rd of vagina
- Located superior to inferior most border of pubic symphysis

Skene Duct Cysts

- Located lateral to external urethral meatus
- Located inferior to level of pubic symphysis

Epidermal Inclusion Cysts

- No communication with urethra
- Located in inferoposterior or lateral vaginal wall at sites of prior trauma or surgery

Urethral Caruncle

- Due to distal urethral prolapse
- Located at posterior margin of external urethral meatus

- T2WI: ↑ SI of rim of tissue surrounding posteroinferior aspect of urethra

Müllerian Cysts

- No communication with urethra
- Small (< 1 cm), cysts in anterolateral wall of vagina

Solid Urethral Masses

- Lesions with minimal to no cystic component
- Variable enhancement
- Examples include endometrioma, adenoma, carcinoma, rhabdomyoma

Periurethral Collagen Injection

- Used to treat stress incontinence
- Typically ↓ SI on T2WI
- ↑ attenuation on CT may mimic calculi within diverticulum

PATHOLOGY

General Features

- Etiology
  - Most common theory: Obstruction of paraurethral gland leads to abscess, which ruptures into urethral lumen and results in diverticulum
    - Most commonly associated infections are *E. coli* and chlamydia
  - Diverticula can also result from obstetric trauma or urethral or vaginal surgery
  - Congenital diverticula are rare and may develop from cloacogenic rests, Gartner duct cysts, and müllerian duct cysts

Gross Pathologic & Surgical Features

- Focal outpouching of urethra consisting of various sizes
- Most diverticula and associated orifices are situated posterolaterally
- Only small minority have orifices located anteriorly or anterolaterally

Microscopic Features

- Inflamed transitional epithelial mucosa
- Reactive changes due to chronic cystitis, cystitis cystica, cystitis glandularis
- Foci of squamous and adenomatous metaplasia

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most commonly, signs and symptoms are nonspecific and include: Recurrent or chronic lower urinary tract infections, urinary incontinence, dysuria, urgency, frequency, pain, post-void dribbling
- Other signs/symptoms
  - Classic triad of dysuria, dyspareunia, and post-void dribbling is less common
- Clinical profile
  - Physical examination rarely aids in diagnosis because diverticula
    - May not be palpable or tender
    - May mimic several vaginal wall lesions
    - May present as discreet or diffuse periurethral lesion

Demographics

- Age
  - Can occur at any age
    - Most common in 3rd-5th decades
- Gender
  - F >> M
- Epidemiology
  - Approximately 0.6-6% of women develop diverticula

Natural History & Prognosis

- Prognosis and recovery excellent following surgery
- If untreated, diverticula are prone to complications
  - Recurrent infection
    - Recurrent UTI reported in ~ 30-50% of patients with diverticula
  - Urinary incontinence
    - Associated with ~ 60% of diverticula
  - Calculus formation
    - Found in up to ~ 10% of diverticula
    - Due to stagnation of infected urine
    - Most calculi due to calcium oxalate or calcium phosphate
  - Intraluminal neoplasms
    - Adenocarcinoma (~ 60%) > squamous cell > transitional cell carcinomas
    - Diverticular neoplasms are typically late stage at presentation
    - High rate of recurrence and poor prognosis

Treatment

- Transvaginal and transvaginal diverticulectomy are preferred treatments
- Potential postoperative complications include
  - Recurrence/incomplete resection (1-29%)
  - Stress incontinence (1.7-12.5%)
  - Urethrovaginal fistula (0.9-5%)

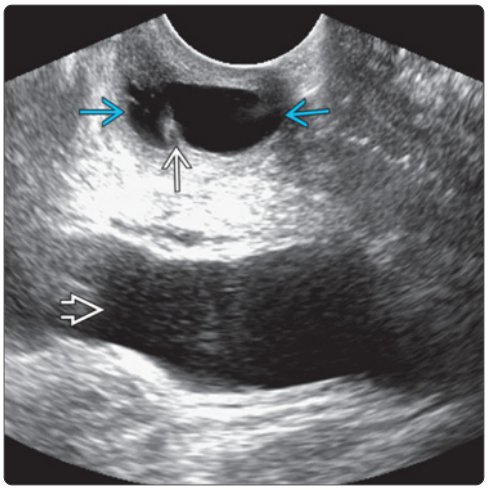
DIAGNOSTIC CHECKLIST

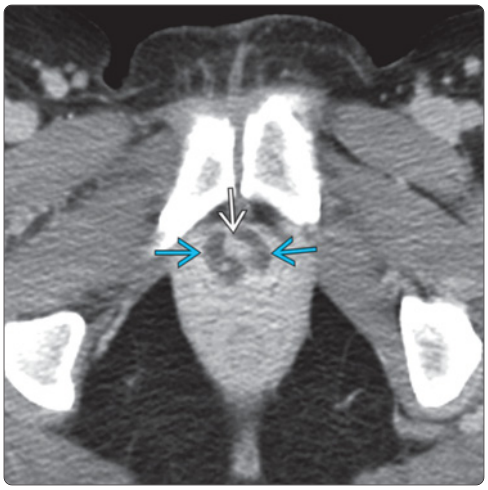
Consider

- In women with recurrent UTIs

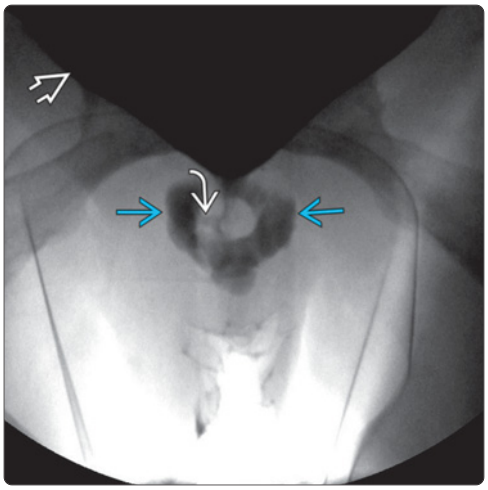
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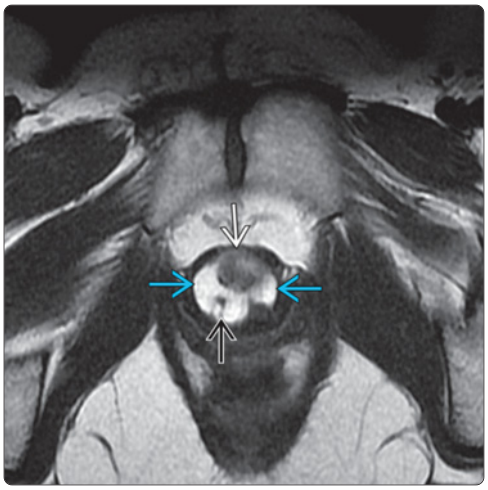
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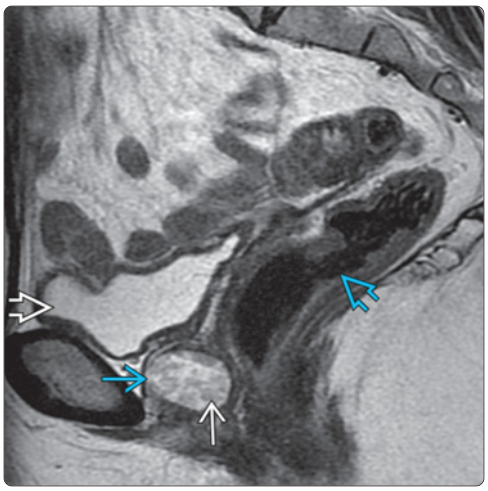


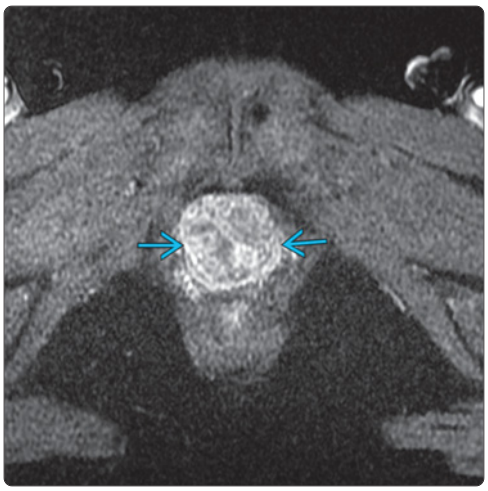
(Left) *Coronal, angled endovaginal US shows an ovoid, cystic structure ⇨ surrounding the urethra ➡. The cystic structure is anechoic and shows posterior acoustic enhancement. More superoposterior, the urinary bladder ➡ is shown.* **(Right)** *Axial CECT at the level of the pubic symphysis in the same patient shows a rounded, cystic structure ⇨ encircling the urethra due to a urethral diverticulum. The enhancement of the urethra and adjacent tissues ➡ is normal. The patient had a history of post-void dribbling.*





(Left) *AP view during voiding cystourethrogram in the same patient demonstrates contrast material filling circumferential urethral diverticulum ⇨. The orifice ➡ of the diverticulum is shown to arise posterolaterally on right side. Contrast material is within the urinary bladder ➡.* **(Right)** *Axial single-shot fast spin-echo T2WI MR in the same patient reveals high signal intensity in the diverticulum ⇨ that surrounds the urethra ➡. A thin, partial septum ⇨ is shown along the right posterolateral aspect of the diverticulum.*





(Left) *Sagittal single-shot fast spin-echo T2WI MR reveals an ovoid diverticulum ⇨ arising from the midurethra. The diverticulum has a heterogeneous appearance due to high signal intensity from fluid and intermediate signal intensity from numerous soft tissue nodules ➡. The urinary bladder ➡ & rectum ⇨ are shown.* **(Right)** *Axial T1WI with fat saturation after contrast injection in the same patient shows heterogeneous enhancement throughout the diverticulum ⇨ due to adenocarcinoma within the diverticular lumen.*

Urethral Trauma

KEY FACTS

TERMINOLOGY

- Classification schemes vary
  - Important to determine whether urethra is intact or partially or completely disrupted
  - If ruptured, determine site (relative to urogenital diaphragm) and whether bladder is intact

IMAGING

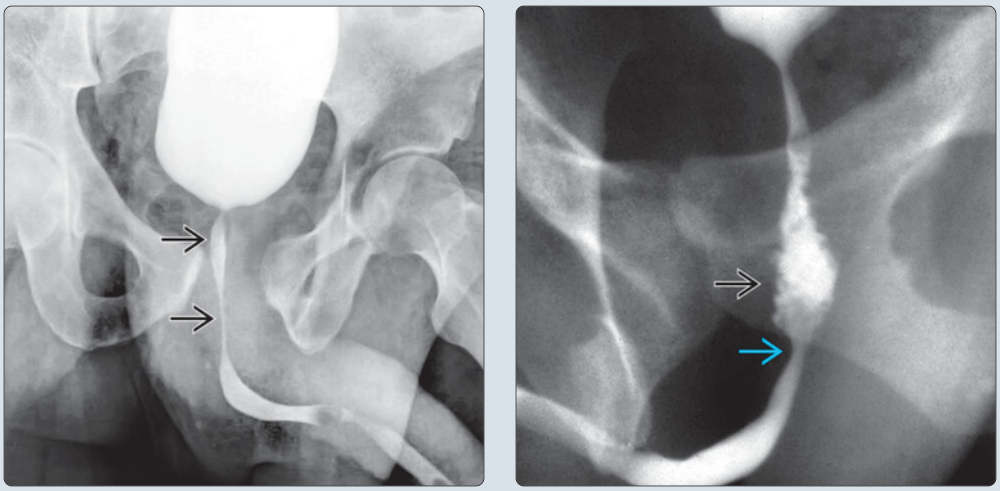
- Goldman classification is most commonly used to describe urethral injuries
- Urethral injuries often accompany extraperitoneal bladder injuries and pelvic fractures
  - Posterior urethral injury is more common
- Retrograde urethrogram (RUG) is modality of choice
- Best diagnostic clue
  - Contrast extravasation from urethra
  - Stretched proximal urethra

PATHOLOGY

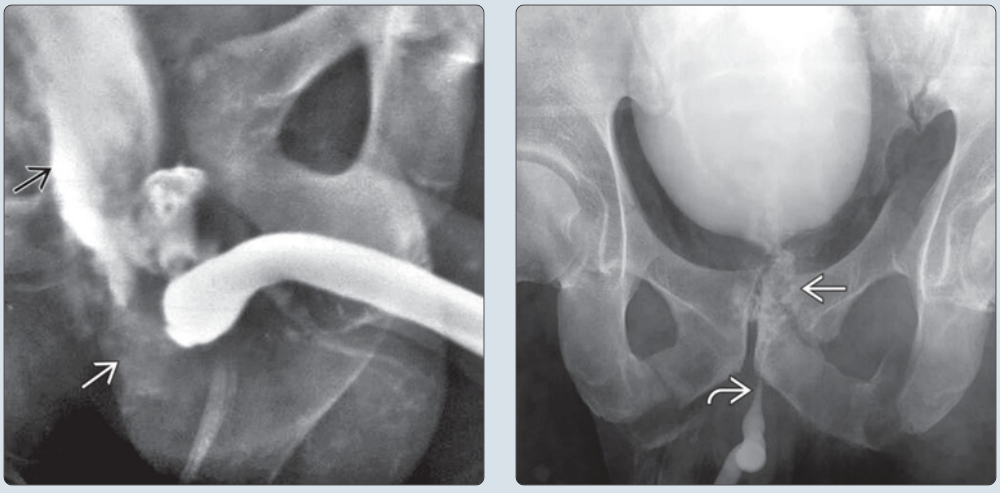
- Most cases are due to blunt trauma
  - Posterior urethral injuries caused by crushing force to pelvis
  - Anterior urethral injuries caused by straddle pelvic injuries
- 20% of posterior urethral injuries have associated bladder laceration
- Anterior urethral injuries often isolated
- Urethral injuries in females often associated with vaginal/rectal injuries

CLINICAL ISSUES

- Urethral injury is seen in 24% of men with pelvic fracture (6% in women)
- Type 3 injury (disruption of urogenital diaphragm) is most common form
- Treatment includes endoscopic stent placement and surgical reconstruction



(Left) *Retrograde urethrogram (RUG) shows a stretched, but intact, posterior urethra, consistent with a type 1 injury in the Goldman classification. According to the AAST classification, this is a type 2 injury. Note the diastasis of the pubic symphysis.* **(Right)** *RUG shows extravasation of contrast material from the posterior urethra; note that the extravasated contrast is located superior to the level of the urogenital diaphragm, consistent with a type 2 injury in the Goldman classification.*



(Left) *RUG shows a Goldman classification type 3 injury, with extravasated contrast superior and inferior to the level of the urogenital diaphragm.* **(Right)** *RUG shows contrast extravasation from a bladder neck laceration that extended into the proximal urethra (Goldman type 4 injury). Note the symmetrical coned appearance of the proximal bulbar urethra demarcating the approximate junction of the anterior and posterior aspects of the urethra and the level of the urogenital diaphragm.*

TERMINOLOGY

Definitions

- Stretching or tearing of urethral mucosa

Classification Schemes Vary

- Main point: Determine integrity of urethra
  - If ruptured, determine site (relative to urogenital diaphragm)

IMAGING

General Features

- Best diagnostic clue
  - Contrast extravasation from urethra during retrograde urethrogram (RUG)

Imaging Recommendations

- Best imaging tool
  - RUG

CT Findings

- CT cystography shows intraperitoneal and extraperitoneal bladder ruptures well
  - Generally insufficient for detailed depiction of urethral injuries
- Urethral injuries often accompany extraperitoneal bladder injuries and pelvic fractures
  - Evaluate for extravasated urine in perivesical space and in scrotum

Fluoroscopic Findings

- Retrograde urethrogram
  - Findings based on Goldman classification
  - Type 1: Stretched posterior urethra but no extravasation
  - Type 2: Contrast extravasation superior to level of urogenital diaphragm
    - Urogenital diaphragm remains intact
  - Type 3: Contrast extravasation surrounding membranous and proximal bulbous urethra into perineum
    - Disruption of urogenital diaphragm
  - Type 4: Bladder neck injury extending into proximal urethra
    - Type 4a: Injury at base of bladder (not involving bladder neck) with periurethral extravasation simulating true type 4 urethral injury
      - Type 4 and 4a indistinguishable on urethrogram
  - Type 5: Contrast extravasation from anterior urethra
    - Injury is isolated to anterior urethra
  - In partial ruptures, contrast fills bladder
    - In complete ruptures, contrast fails to fill bladder
  - Urogenital diaphragm can be identified immediately superior to symmetrical cone-shaped tip of bulbar urethra

PATHOLOGY

General Features

- Etiology
  - Most cases are due to blunt trauma
    - Posterior urethral injuries caused by crushing force to pelvis (motor vehicle accident or fall)
    - Anterior urethral injuries caused by straddle pelvic injuries
  - Penetrating urethral injuries are rare and almost always affect anterior urethra
    - 50% of penetrating penile injuries affect urethra
- Associated abnormalities
  - Posterior urethral injuries
    - Almost always associated with pelvic fracture
    - 20% have associated bladder laceration
  - Anterior urethral injuries often isolated
    - 38% of penile fractures have urethral injury
  - Urethral injuries in females are rare
    - Often associated with vaginal (75%) and rectal (33%) injuries

Staging, Grading, & Classification

- Alternative classification from American Association for Surgery of Trauma (AAST)
  - Class 1: Contusion
  - Class 2: Stretch injury (intact lumen)
  - Class 3: Partial disruption
  - Class 4: Complete disruption (< 2 cm separation)
  - Class 5: Complete disruption (> 2 cm separation)

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Blood at meatus (50%), gross hematuria, inability to void
- Other signs/symptoms
  - Swelling of perineum, "high riding" prostate on digital rectal examination

Demographics

- Gender
  - M > F
- Epidemiology
  - Urethral injury is seen in 24% of men with pelvic fracture (6% in women)
  - Type 3 (Goldman classification) injury is most common

Natural History & Prognosis

- Complications: Impotence, incontinence, stricture, fistula
- Incontinence: More common after type 2 and 3 injuries

Treatment

- Immediate vs. delayed primary reconstruction
- Endoscopic stent placement
- Penetrating injuries need immediate surgical exploration

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Erectile Dysfunction

KEY FACTS

TERMINOLOGY

- Persistent inability to achieve or maintain penile erections of sufficient value to engage in satisfactory sexual activity

IMAGING

- Doppler ultrasound
  - Spectral Doppler interrogation of cavernosal arteries following injection of stimulant (e.g., prostaglandin derivative) should show ↑ peak systolic velocity and ↓ end diastolic velocity
  - Arterial insufficiency threshold range: PSV < 25-35 cm/s
  - Venous insufficiency threshold range: EDV > 5-7 cm/s
- Grayscale ultrasound
  - Peyronie disease: Hyperechoic plaques in corpora cavernosa
  - Penile fracture: Focal, linear defect in corpora cavernosa
- Measurement of penile: Branchial index
  - Measure ratio of peak systolic blood pressures
  - Abnormal is < 0.7
- Angiography
  - 2nd line technique to evaluate for stenosis or occlusion in branches of internal pudendal artery

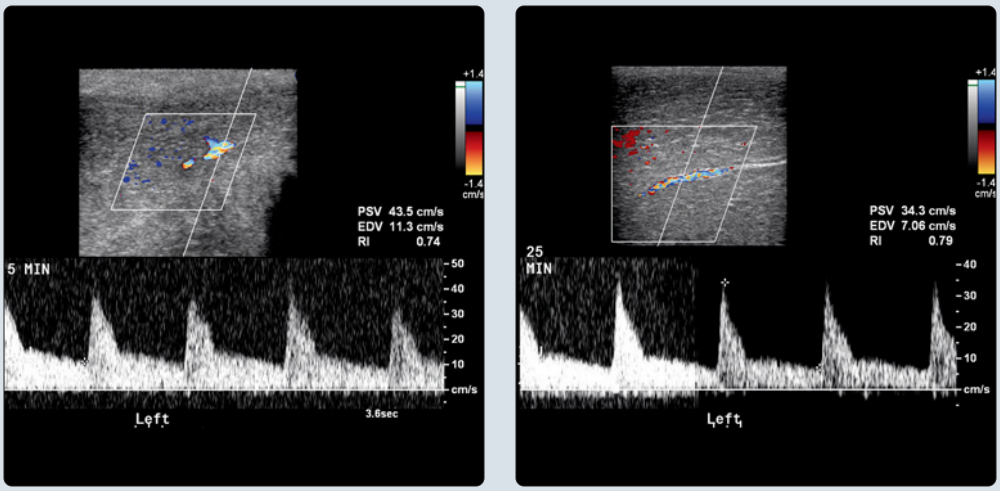
PATHOLOGY

- Multifactorial etiologies
  - Vascular
  - Anatomic
  - Neurological
  - Hormonal
  - Medications
  - Psychological

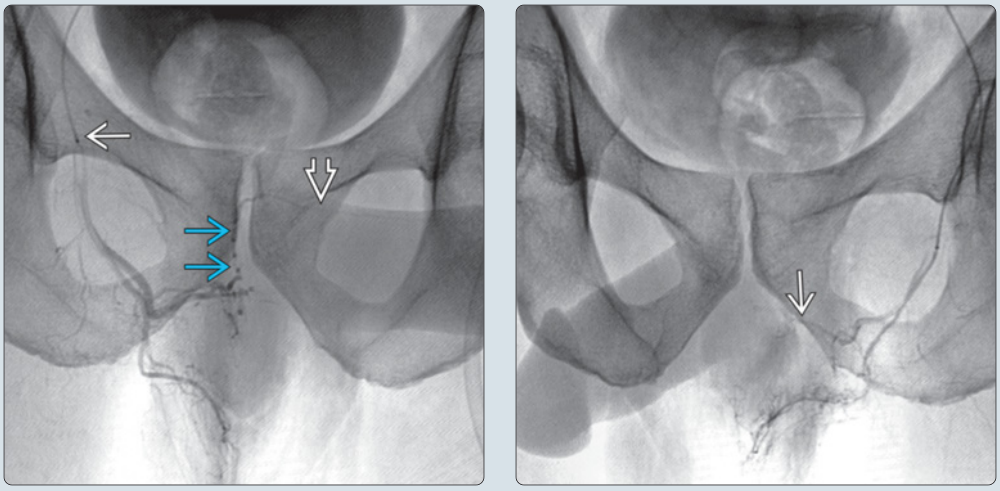
CLINICAL ISSUES

- Patients typically given trial of phosphodiesterase inhibitors prior to any imaging
- Treatment
  - Medical: Phosphodiesterase inhibitors
  - Surgical: Revascularization vs. penile pump prosthesis

(Left) *Transverse Doppler US at the left penile base obtained 5 minutes past intracavernosal injection shows a normal peak systolic velocity of 44 cm/s and an end diastolic velocity of 11 cm/s within the left cavernosal artery. (Courtesy M. Lockhart, MD.)* **(Right)** *Sagittal Doppler US in the same patient was obtained 20 minutes later at the left aspect of the penile base and shows that the end diastolic velocity has only decreased to 7 cm/s, consistent with venous insufficiency. (Courtesy M. Lockhart, MD.)*



(Left) *Anteroposterior view during angiography shows the catheter tip in the right internal pudendal artery. The penile artery is irregular and shows multiple areas of narrowing. The dorsal penile artery is diminutive, while the cavernosal artery could not be identified.* **(Right)** *AP view during left internal pudendal arteriogram in the same patient reveals that the penile artery is diminutive and no flow was observed in the dorsal penile artery or cavernosal artery in this patient with advanced atherosclerosis.*



TERMINOLOGY

Abbreviations

- Erectile dysfunction (ED)

Definitions

- Persistent inability to achieve or maintain penile erections of sufficient value to engage in satisfactory sexual activity

IMAGING

General Features

- Best diagnostic clue
  - Abnormal arterial waveforms and velocities on spectral Doppler US
- Location
  - Penile and cavernosal arteries

MR Findings

- T1WI
  - Normal corpora cavernosa and spongiosum have intermediate to high signal intensity (SI)
  - Cavernosal fibrosis and Peyronie disease: ↓ SI
- T2WI
  - Normal corpora cavernosa and spongiosum have high SI
  - Cavernosal fibrosis and Peyronie disease: ↓ SI

Ultrasonographic Findings

- Grayscale ultrasound
  - Peyronie disease
    - Localized benign connective tissue disorder
    - Hyperechoic thickened plaques of penile tunica albuginea
      - ± calcified plaques
    - Hyperperfusion near plaques in early phase
  - Penile fracture
    - May result in focal fibrosis or arterial and venous insufficiency
    - Focal, linear hyperechoic lesion in corpora cavernosa
- Pulsed Doppler
  - Arterial waveform during erection is multiphasic
    - Normal waveforms show ↑ peak systolic velocity (PSV), narrowing of systolic peak after injection of pharmacostimulant, and ↓ end diastolic velocity (EDV)
  - Normal PSV is > 30-35 cm/s
    - Arterial insufficiency threshold: PSV < 25 cm/s
      - Borderline if <30 cm/s
    - PSV < 25 cm/s correlates to severe arterial disease
  - Normal EDV if negative or close to 0 cm/s
    - Venous insufficiency threshold: EDV > 5 cm/s
- Measurement of penile-brachial index
  - Noninvasive screening test for vasculogenic erectile dysfunction
  - Systolic pressures are measured in both cavernosal arteries with Doppler probe and compared to systolic pressure in brachial artery
  - Abnormal is < 0.7

Angiographic Findings

- Performed as 2nd line technique after US
- 2 main approaches
  - Internal pudendal arteriography
    - Evaluate arterial branches for stenosis or occlusion
  - Cavernosography
    - Intracavernosal contrast injection to evaluate for veno-occlusive disease
    - Not commonly performed currently

Imaging Recommendations

- Best imaging tool
  - Penile Doppler ultrasonography
- Protocol advice
  - Doppler sonography: Performed every 5 minutes after intracavernosal injection of stimulant agent (e.g., prostaglandin E1 derivative) during 20-30 minute period
  - Cavernosal arterial waveforms are sampled until peak systolic and end diastolic velocities are reached

PATHOLOGY

General Features

- Etiology
  - Vascular (atherosclerosis, diabetes, smoking)
  - Anatomic (Peyronie disease, penile fibrosis)
  - Neurological (brain injury, cerebrovascular disease)
  - Hormonal (central hypogonadism, Cushing disease)
  - Medications (beta blockers, antidepressants)
  - Psychological (sole cause in only ~ 10-20%)

CLINICAL ISSUES

Presentation

- Inability to achieve or maintain erection

Demographics

- Age
  - Affects ~ 50% of men between 40-70 years of age
    - ~ 10% are severely affected

Treatment

- Medications
  - Phosphodiesterase type 5 (PED5) inhibitors
- Surgery
  - Arterial bypass/revascularization
  - Penile pump insertion
    - Less commonly used due to effective medical therapy
    - Complications include cavernosal perforation, reservoir malpositioning, infection

DIAGNOSTIC CHECKLIST

Consider

- Imaging after failed trial of phosphodiesterase inhibitors

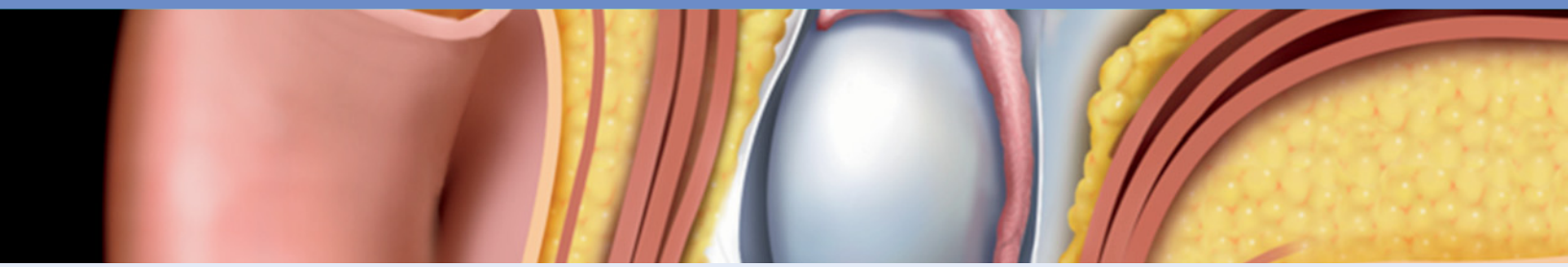
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SECTION 8

Testes



Nonneoplastic Conditions

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Epidermoid Cyst **486**

Introduction

High-frequency transducer sonography using gray-scale along with pulsed and color Doppler is the imaging modality of choice for evaluating patients presenting with scrotal pathology. Scrotal ultrasound (US) is often requested in an emergency setting in a case of acute scrotal pain. The leading differential diagnoses in such a scenario includes testicular torsion, acute epididymo-orchitis, and traumatic injury (in the setting of preceding trauma). In a nonacute setting, scrotal ultrasound is often requested for evaluation of chronic testicular pain or a palpable scrotal mass. The leading differential diagnoses in the nonacute setting when a mass is palpated includes a testicular neoplasm (benign or malignant) and extra testicular masses, including epididymal lesions and inguinal hernias.

Clinical correlation with history and symptoms is an extremely important aspect of scrotal sonography.

Ultrasound Technique for Scrotal Evaluation

Scrotal US is performed with the patient in the supine position and the scrotum supported by a towel placed between the thighs. Optimal results are obtained with a 10-14MHz high-frequency, linear-array transducer. Scanning is performed with the transducer in direct contact with the skin with copious amounts of gel; if necessary, a stand-off pad can be used for evaluation of superficial lesions.

The testes are examined in at least two planes (i.e., the longitudinal and transverse axes). The size and echogenicity of each testis and epididymis are compared with those on the opposite side. Scrotal skin thickness is evaluated for symmetricity as well as focal or diffuse edema. Color Doppler and pulsed Doppler parameters are optimized to display low-flow velocities and to demonstrate blood flow in the testes and surrounding scrotal structures. Color Doppler ultrasound should include comparison of right and left testicular spectral Doppler tracings. Power Doppler US may also be used to demonstrate intratesticular blood flow in patients with an acute scrotum, particularly if torsion is considered.

When evaluating patients presenting with an acute scrotum, the asymptomatic side should be scanned first in order to optimize gray-scale and color Doppler gain settings. This allows for comparison with the affected side (though a caveat is that testicular torsion can be a bilateral process in 2% of patients). Transverse images with portions of each testis on the same image should be acquired in gray-scale and color Doppler modes. Additional techniques, such as use of the Valsalva maneuver or upright positioning, can be used as needed for venous evaluation. Power Doppler sonography uses the integrated power of the Doppler signal to depict the presence of blood flow. Power Doppler sonography may be slightly more sensitive than standard color Doppler for detecting low-flow states and may provide essential information in the diagnosis of potential complete testicular torsion.

Clinical Perspective

Acute Scrotal Pain

In an emergency setting, acute scrotal pain should prompt an assessment of epididymo-orchitis, testicular torsion, and traumatic injury. Ultrasound features are correlated with history and physical examination findings. Although acute epididymo-orchitis and acute testicular torsion present similarly with acute onset of unilateral testicular pain, epididymo-orchitis may be accompanied with fever &/or a Prehn sign (relief of pain with elevation of scrotum above the level of pubic symphysis). Moreover, the sonographic appearances for both diagnoses are very distinct: Edema with increased vascularity in epididymo-orchitis and edema with absent or reduced flow in torsion. A potential pitfall for testicular torsion is the torsion-detorsion syndrome, wherein the testis may detorse spontaneously: Detorsion results in reactive hyperemia, which can appear sonographically similar to epididymo-orchitis. An appropriate history preceding the clinical presentation may lead one to the correct diagnosis. Torsion-detorsion syndrome will typically present with intermittent symptoms of pain and discomfort. Epididymo-orchitis will present with constant or worsening pain. Associated findings of inflammation in the ipsilateral scrotum, such as scrotal skin thickening and pyocele, also favor epididymo-orchitis.

The diagnosis of a traumatic scrotal injury is usually not difficult with a prior history of trauma (blunt or penetrating). The purpose of performing an ultrasound after trauma is to determine whether surgical exploration is required. Most cases with testicular rupture (tunica albuginea disruption) will need to be surgically repaired. In addition, large hematoceles or large hematomas resulting in testicular ischemia may require surgical evacuation &/or debridement. The presences of color Doppler flow in the testis helps determine viability. It is extremely important to follow all such cases to resolution, as some hematomas may become infected and form an abscess.

Chronic/Nonacute Scrotal Pathology

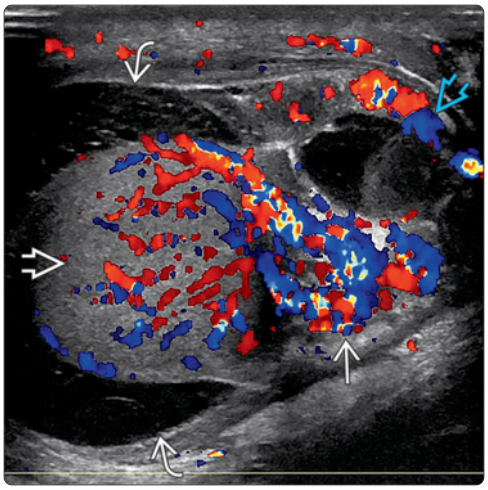
In a nonacute setting, patients often present with an enlarged scrotum, palpable mass, or mild scrotal pain. An enlarged scrotum may be related to a hydrocele, spermatocele, scrotal wall edema, or inguinal hernia. In the setting of a palpable mass, ultrasound is extremely sensitive in detecting the presence of an intratesticular neoplasm, which is a malignancy until proven otherwise. Mild scrotal pain is often related to a varicocele, which can easily be diagnosed with color Doppler and Valsalva maneuvers.

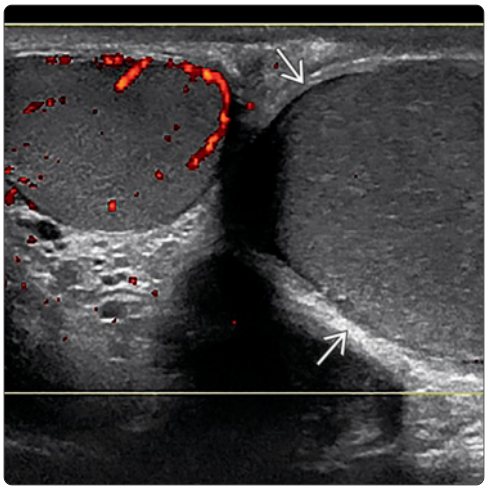
Summary

The ability of ultrasound to diagnose the pathogenesis of the acute scrotum is unsurpassed by any other imaging modality. Ultrasound is the first-line imaging study performed in patients with acute scrotum. Knowledge of the normal and pathologic sonographic appearance of the scrotum and an understanding of the proper technique are essential for accurate diagnosis of an acute scrotum. High-frequency transducer sonography combined with color and power Doppler sonography provides information essential to reach a specific diagnosis in patients with testicular torsion, epididymo-orchitis, and scrotal trauma.

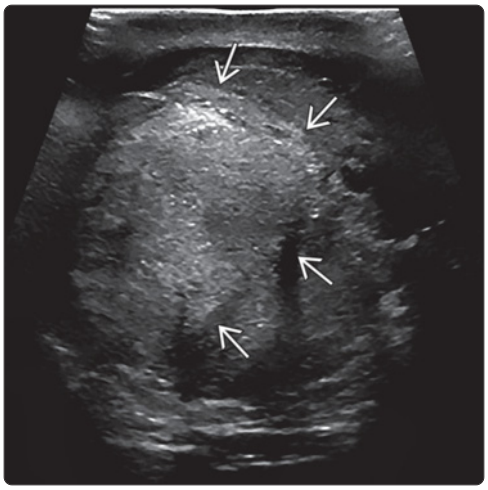
Selected References

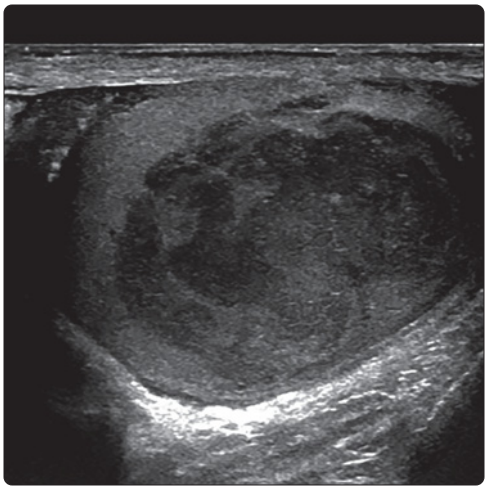
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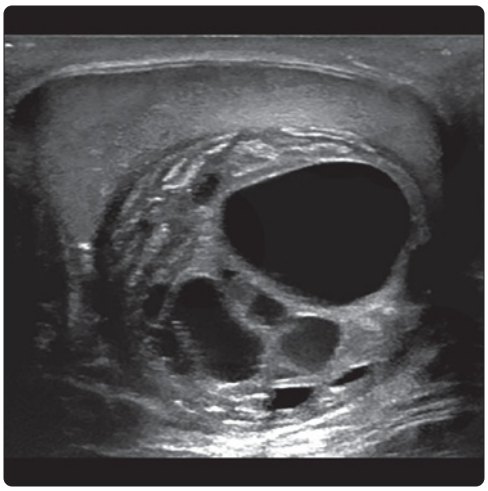


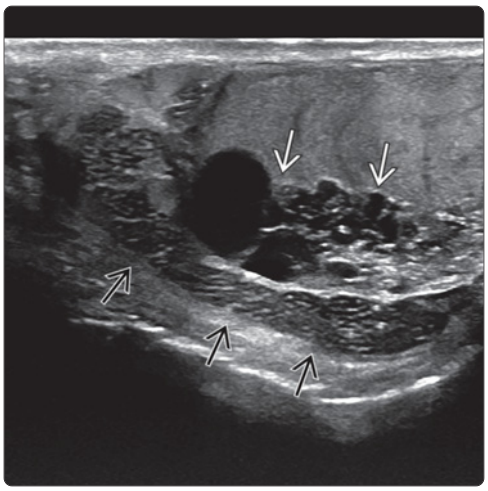
(Left) *Color Doppler ultrasound of an elderly male with acute scrotal pain and a history of urinary tract infections shows a hyperemic, epididymis and testis (an appearance compatible with epididymo-orchitis). Note the adjacent pyocele and debris filled epididymal cyst.* **(Right)** *Power Doppler ultrasound of a 35-year-old man with acute left testicular torsion shows no flow within the left testis. Technique should be optimized to detect slow flow when torsion is suspected; comparison to the asymptomatic testis is critical.*





(Left) *Ultrasound performed on a young male, post blunt scrotal injury, shows a wide testicular fracture. No flow was shown at color Doppler and the testis was not viable at exploration. The goal of ultrasound in these settings is to assess testicular flow and tunica integrity.* **(Right)** *Ultrasound shows a solid hypoechoic testicular mass (a finding indicating malignancy until proven otherwise). Most testicular tumors are either seminomas or mixed germ cell tumors, although lymphoma was confirmed in this HIV positive male.*





(Left) *Scrotal ultrasound of a 40-year-old man with a palpable mass shows a mixed cystic and solid testicular mass (a confirmed mixed germ cell tumor). Ultrasound is the imaging modality of choice for evaluation of scrotal contents.* **(Right)** *Ultrasound performed for evaluation of scrotal discomfort shows an elongated cystic mass composed of tubular components. Location within the mediastinum testis and the ultrasound appearance indicate dilated rete testes (a leave alone lesion). Note the post vasectomy epididymis.*

Cryptorchidism

KEY FACTS

TERMINOLOGY

- Incomplete descent of testis into scrotum

IMAGING

- Located anywhere from kidney to inguinal canal
  - Inguinal canal most common (80%)
  - Bilateral in 10%
- Ultrasound: Study of choice for inguinal testis
  - Ovoid, homogeneous, well-circumscribed mass
  - Smaller than normally descended testis
  - Echogenic line of mediastinum testis helps differentiate from other soft tissues masses
- MR: Study of choice for intraabdominal testes
  - 94% accuracy for detection and characterization of undescended testes
  - T1WI: Low signal intensity ovoid mass
  - T2WI: High signal intensity ovoid mass but often not as bright as normal testis
  - Decreased T2 signal secondary to fibrotic changes
  - Focal T2 low signal mass suspicious for tumor
- Role of imaging discounted though in recent (2014) AUA guidelines: Rarely impacts decision making

TOP DIFFERENTIAL DIAGNOSES

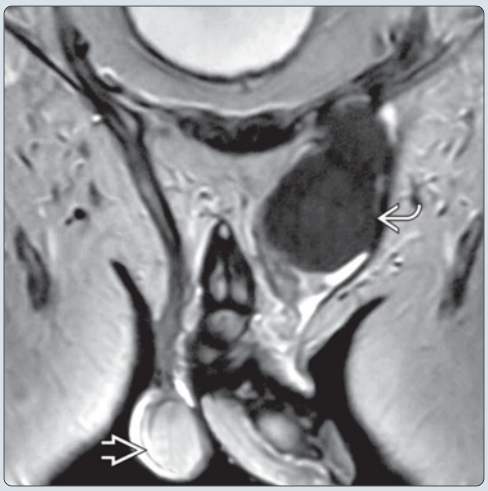
- Absent testis (anorchia)
  - Congenital (rare) or prior resection
- Inguinal masses
  - Lymphadenopathy, hernia

CLINICAL ISSUES

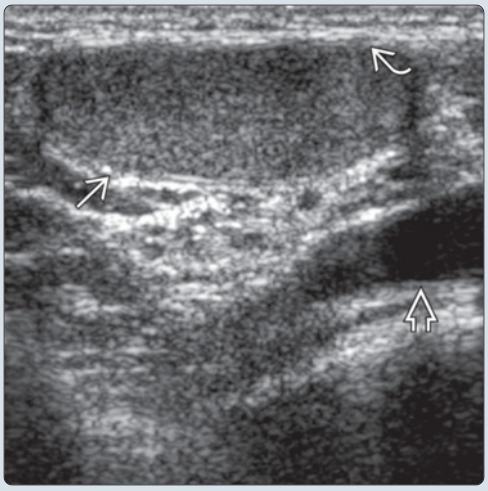
- ↑ risk of malignant neoplasm in both cryptorchid testis and contralateral testis
  - Increased risk likely from abnormal embryogenesis affecting both testes
  - 3.5-14.5% of testicular cancer patients have history of cryptorchidism
- ↑ incidence of infertility

(Left) *Coronal T2WI MR shows an empty scrotal sac and bilateral undescended testes. The gubernaculum, a ligament which extends from the lower pole of the testis to the scrotal sac, is well demonstrated.* **(Right)** *Coronal T2WI MR shows a low signal tumor within an undescended testis. Compare this signal to the normally descended testis on the right. It is important to know that the increased risk of carcinoma does not just apply to the undescended side but also to the normally descended testis.*





(Left) *Oblique ultrasound along the inguinal canal shows an ovoid, hypoechoic testis deep to the subcutaneous tissue and fascia. Note the femoral vessels. The inguinal canal is the most common location for an undescended testis and may present as a palpable groin mass.* **(Right)** *Axial CECT shows ovoid soft tissue mass within the retroperitoneum just lateral to the kidney. This could easily be confused with other soft tissue masses or adenopathy, so history is essential. Lower images showed no spermatic cord.*





TERMINOLOGY

Definitions

- Incomplete descent of testis into scrotum

IMAGING

General Features

- Location
  - Anywhere from kidney to inguinal canal
  - Inguinal canal most common (80%)
  - Bilateral in 10%

Ultrasonographic Findings

- Ovoid, homogeneous, well-circumscribed mass
  - Iso- or hypoechoic compared to normal testis
  - Smaller than normally descended testis
- Echogenic line of mediastinum testis helps differentiate from other soft tissues masses
- Best for identifying testis in prescrotal region, just beyond external inguinal ring, or in inguinal canal

CT Findings

- Low-attenuation mass above scrotum in path of testicular descent
- Absent spermatic cord in inguinal canal

MR Findings

- T1WI: Low signal intensity ovoid mass
- T2WI: High signal intensity ovoid mass but often not as bright as normal testis
  - Decreased signal secondary to fibrotic changes
  - Focal low signal mass suspicious for tumor

Imaging Recommendations

- Ultrasound for inguinal testis
- MR study of choice for intraabdominal testes: 94% accuracy for detection and characterization of undescended testes
- Recent (2014) AUA guidelines do not recommend imaging for localization
  - Rarely assists decision making
  - Early exploration and inguinal/scrotal orchiopexy advocated if no spontaneous testicular descent after 6 months of age

DIFFERENTIAL DIAGNOSIS

Absent Testis (Anorchia)

- Congenital (rare)
  - Testis is absent rather than undescended in 5% of cases when testis is nonpalpable
- Prior resection

Inguinal Masses

- Lymphadenopathy
  - Hypoechoic mass, usually multiple
  - Large benign nodes have echogenic hilum: Look for color Doppler flow within hilum
- Hernia
  - "Gut signature": Alternating hypo- and hyperechoic layers of bowel wall
- Varicocele
  - Distended serpiginous veins

PATHOLOGY

General Features

- Associated abnormalities
  - Renal agenesis/ectopia, prune belly syndrome, epispadias
  - Wolffian duct anomalies
    - Seminal vesical cyst or agenesis
  - Infertility: Risk increases if bilateral cryptorchidism, &/or delayed orchiopexy
- Risk factor for developing testicular carcinoma
  - Pathophysiology not completely clear but thought to be manifestation of generalized embryogenesis defect in testicular development
  - Theory supported by several observations
    - Risk of developing carcinoma not limited to cryptorchid testis, also increased in contralateral testis
    - Orchiopexy only modestly decreases risk of developing tumor
    - Risk of carcinoma increases with degree of ectopy, with intraabdominal testes at greatest risk
      - Higher testis location correlates with earlier defect in embryogenesis

CLINICAL ISSUES

Demographics

- 9.2-30% in premature infants
- 3.4-5.8% in full-term infants
- 0.8% by age 3 months
  - Spontaneous descent after birth

Natural History & Prognosis

- ↑ risk of malignant neoplasm in both cryptorchid testis and contralateral testis
  - 3.5-14.5% of testicular cancer patients have history of cryptorchidism
- ↑ incidence of infertility

Treatment

- Orchiopexy: Optimal time has been debated
  - 6-12 months currently recommended
    - Better postoperative growth of testis than if performed later
  - 95% success rate
- Surgical removal post puberty

DIAGNOSTIC CHECKLIST

Consider

- Increased risk of developing testicular carcinoma not just limited to cryptorchid testis; opposite side also at risk

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Testicular Torsion

KEY FACTS

TERMINOLOGY

- Spontaneous or traumatic twisting of testis and spermatic cord within scrotum, resulting in vascular occlusion/infarction

IMAGING

- Absent or decreased abnormal testicular blood flow on color Doppler US
- Findings vary with duration and degree of rotation of cord
- Unilateral in 95% of patients
- Role of spectral Doppler is limited; may be helpful to detect partial torsion; in partial torsion of 360° or less, spectral Doppler may show diminished diastolic arterial flow
- Spiral twist of spermatic cord cranial to testis and epididymis causes torsion knot or whirlpool pattern of concentric layers

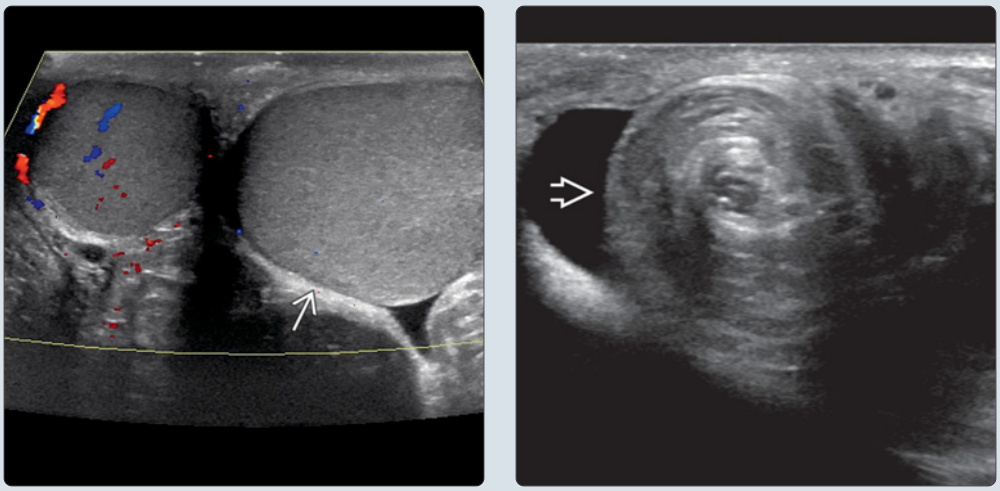
PATHOLOGY

- Varying degrees of ischemic necrosis and fibrosis depending on duration of symptoms
- Undescended testes have an increased risk of torsion
- Intravaginal torsion: Common type, most frequently occurs at puberty

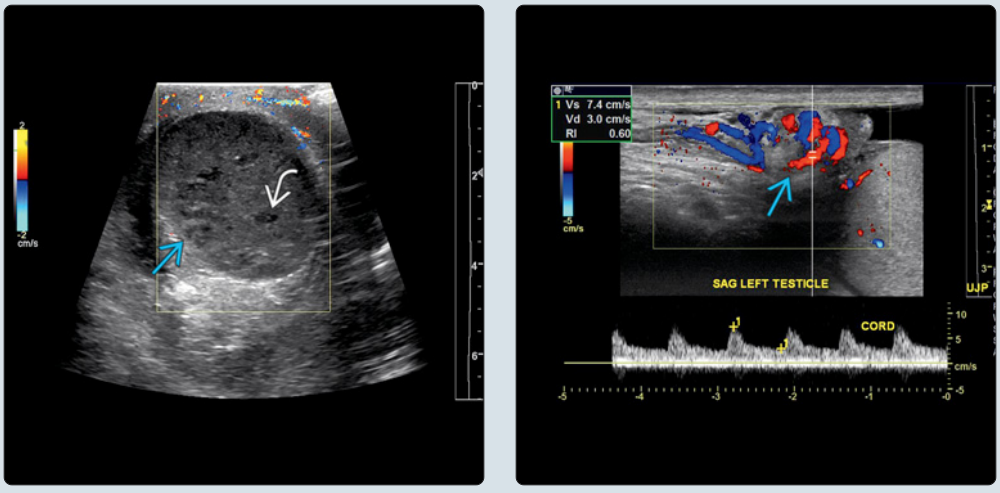
CLINICAL ISSUES

- Acute scrotal/inguinal pain; swollen, erythematous hemiscrotum without recognized trauma
- Reducing time lag between onset of symptoms and time of surgical or manual detorsion is of utmost importance in preserving viable testis
- Nonviable testicle usually removed; higher risk of subsequent torsion on contralateral side
- Venous obstruction occurs 1st, followed by obstruction of arterial flow, which leads to testicular ischemia

(Left) *Color Doppler ultrasound performed on a young male with acute scrotal pain shows no flow within a relatively enlarged left testis. Doppler parameters should be optimized to detect slow flow and comparison to the contralateral (asymptomatic) testis is critical.* **(Right)** *Ultrasound performed above the left testis in the same patient shows the "whirl" sign due to twisting of the spermatic cord. Direct visualization of the twisted cord may increase exam accuracy, particularly in cases of partial torsion.*



(Left) *Transverse color Doppler ultrasound of the testis in a young male with an acute painful scrotum for 48 hours shows a heterogeneous avascular testis with cystic areas. The time course and ultrasound features indicate frank necrosis and a nonviable testis.* **(Right)** *Sagittal color Doppler ultrasound superior to the left testis in a young male with acute painful scrotum shows a whirlpool sign secondary to a twisted spermatic cord. The testis was manually detorsed and definitive orchiopexy was performed thereafter.*



TERMINOLOGY

Definitions

- Spontaneous or traumatic twisting of testis and spermatic cord within scrotum, resulting in vascular occlusion/infarction

IMAGING

General Features

- Best diagnostic clue
  - Absent or decreased testicular blood flow on color Doppler US
- Location
  - Unilateral in 95% of patients
- Morphology
  - Complete/incomplete torsion
  - Types according to location of torsion knot
    - Intravaginal torsion: Torsion knot within tunica vaginalis
    - Extravaginal torsion: Torsion knot outside tunica vaginalis

Ultrasonographic Findings

- Grayscale ultrasound
  - Testicular torsion
    - Findings vary with duration and degree of rotation of cord
    - Grayscale appearance during acute phase (critical phase) may be normal
    - Spiral twist of spermatic cord cranial to testis and epididymis causing torsion knot or whirlpool pattern of concentric layers
    - Enlarged testis and epididymis (heterogeneous echotexture, most often decreased echogenicity)
    - Edema of scrotal wall, secondary hydrocele
    - Intratesticular necrosis, hemorrhage, or fragmentation if delayed diagnosis missed (chronic) torsion
    - Hypoechoic areas associated with testicular infarction may have striated appearance (due to accentuation of septa within the testis)
    - Normal testicular echogenicity is strong predictor of testicular viability; decreased and heterogeneous testicular echogenicity correlates with worse prognosis and may indicate nonviability
    - Subacute phase (1-10 days): Testis echogenicity decreases in initial 4-5 days, then may show focal or diffuse infarction; epididymis may remain echogenic
    - Chronic phase: Atrophic, hypoechoic testis; relatively enlarged echogenic epididymis
  - Testicular infarction
    - Diffusely hypoechoic small testis or focal wedge-shaped hypoechoic area in infarcted testis
    - Hyperechoic regions (hemorrhage/fibrosis), focal infarctions may have striated appearance
- Color Doppler
  - Critical for establishing diagnosis of testicular torsion
  - In acute torsion sensitivity, 80-90%; 10% of patients with early or partial torsion may have normal exam
    - Optimize parameters to detect slow flow: Low pulse repetition frequency, low wall filter, high Doppler gain
  - Absent or decreased or abnormal flow
  - Comparison with contralateral spermatic cord and testis is mandatory
  - Subacute or chronic torsion: No flow in testis and increased flow in paratesticular tissues, including epididymis-cord complex and dartos fascia
  - Role of spectral Doppler is limited; may be helpful to detect partial torsion: In partial torsion of 360° or less, spectral Doppler may show diminished diastolic arterial flow
  - Use of intravascular ultrasound contrast agents may improve sensitivity for detecting blood flow in testes
  - Increased flow may be seen in the affected testis if torsion detorsion: Correlation with clinical history of preceding intermittent ipsilateral pain is helpful clue

Nuclear Medicine Findings

- Tc-99m pertechnetate: Dynamic flow imaging at 2-5 second intervals for 1 minute (vascular phase); 5 minute intervals for tissue phase; sensitivity 80-90%
  - Can detect reduced or absent testicular flow in 94-99%
  - In testicular torsion, rounded cold area and halo of dartos perfusion is seen

Imaging Recommendations

- Best imaging tool
  - US with high-frequency linear transducer and color/power Doppler
- Protocol advice
  - Compare grayscale, color Doppler appearance to contralateral normal testis

DIFFERENTIAL DIAGNOSIS

Testicular Trauma

- Hematocele, irregular contours, heterogeneous parenchymal echogenicity
- Avascular fracture plane

Testicular Abscess (Epididymo-Orchitis)

- Thick-walled, hypoechoic focus with low-level internal echoes, thickened tunica albuginea
- Enlarged hypoechoic epididymis with increased flow on color Doppler

Testicular Tumor

- Focal hypoechoic mass with heterogeneous regions
- Abnormal vascularity within mass

Torsion of Appendix Testis or Appendix Epididymis

- Presents with acute scrotum
- Small firm nodule that is palpable on superior aspect of testis that exhibits bluish discoloration through overlying skin called "blue dot" sign
- Hyperechoic mass with central hypoechoic area adjacent to testis or epididymis, reactive hydrocele, skin thickening, and peripheral hyperemia

PATHOLOGY

General Features

- Etiology
  - Most occur spontaneously; rarely occurs due to traumatic etiology

- Varying degrees of ischemic necrosis and fibrosis depending on duration of symptoms
- Undescended testes have increased risk of torsion
- Exocrine and endocrine function is substandard in men with history of unilateral torsion; the following 3 theories explain the contralateral disease noted in torsion
  - Unrecognized or unreported repeated injury to both testes
  - Preexisting pathologic condition predisposing both testes to abnormal spermatogenesis and torsion of spermatic cord
  - Induction of pathologic changes in contralateral testis by retention of injured testis

Staging, Grading, & Classification

- Intravaginal torsion: Common type, most frequently occurs at puberty
  - Torsion occurs within tunica vaginalis
  - 2 predisposing conditions
    - Long stalk of mesentery or spermatic cord leading to anomalous suspension of testis
    - Bell clapper deformity where tunica vaginalis completely encircles epididymis, distal spermatic cord and testis rather than attaching to posterolateral aspect of testis
  - Anomalous testicular suspension is bilateral in 50-80%
  - 10x increased incidence of torsion in undescended testis after orchiopexy
- Extravaginal torsion: Exclusively in newborns
  - No bell clapper deformity
  - Torsion occurs outside tunica vaginalis when testes and gubernacula are not fixed and are free to rotate
  - Infarcted and necrotic testis at birth

Gross Pathologic & Surgical Features

- Purple, edematous, ischemic testicle, may rapidly reperfuse when manually detorsed

Microscopic Features

- Hemorrhage, interstitial edema; necrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute scrotal/inguinal pain; swollen, erythematous hemiscrotum without recognized trauma
  - Pain not relieved by elevation of scrotum
  - Absent cremasteric reflex
- Clinical profile
  - Young male with acute scrotal pain

Demographics

- Epidemiology
  - Infant and adolescent boys most often affected

Natural History & Prognosis

- Testis usually turns medially up to 1,080° (3 full revolutions)
  - Venous obstruction occurs 1st, followed by obstruction of arterial flow, which leads to testicular ischemia
  - Diminished blood flow in testis with twist of 180° or less
  - Testicular viability depends on degree of torsion and duration of symptoms
- Surgical emergency: Testicular infarction if not treated promptly

Treatment

- Surgical exploration; detorsion; bilateral orchidopexy if viable testicle
  - Nonviable testicle usually removed; higher risk of subsequent torsion on contralateral side
- Delaying surgical intervention worsens intraoperative testicular salvage rate and extent of subsequent testicular atrophy
- Reducing time lag between onset of symptoms and time of surgical or manual detorsion is of utmost importance in preserving viable testis
- Salvage rate of testis vs. time interval between onset of pain and surgery
  - 80-100% after < 6 hours
  - 76% after 6-12 hours
  - 20% after 12-24 hours
  - 0% after > 24 hours

DIAGNOSTIC CHECKLIST

Consider

- Normal US (grayscale and Doppler) does not exclude early or partial torsion
  - Repeat examination at 1-4 hour intervals if conservatively managed

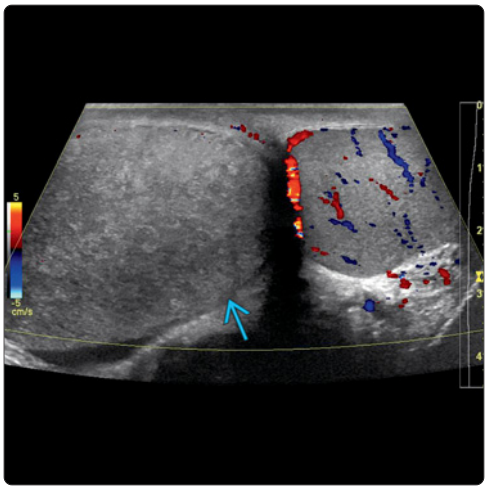
Image Interpretation Pearls

- Decreased or absent flow on Doppler ultrasound

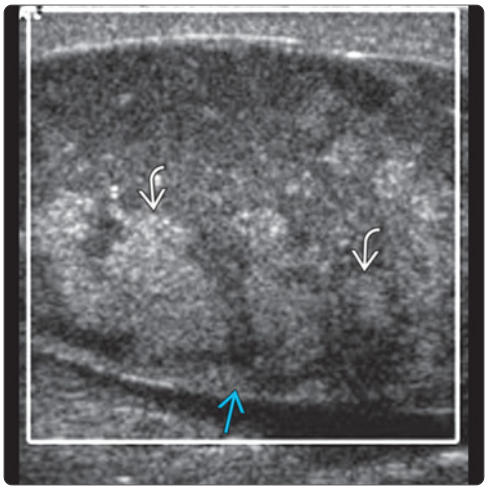
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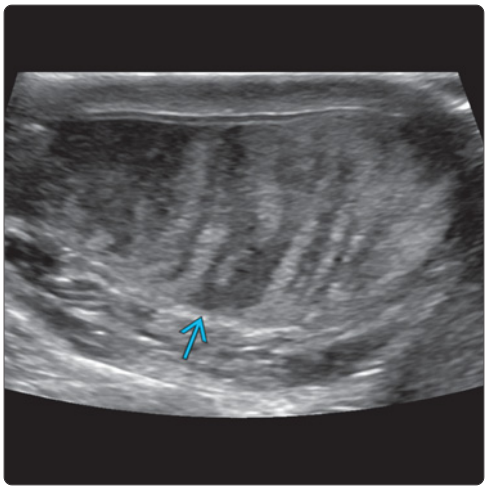
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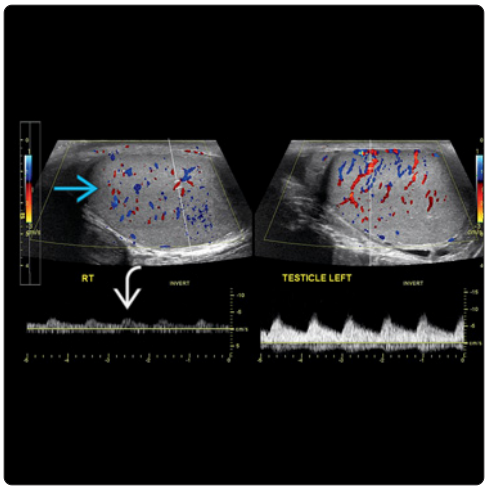


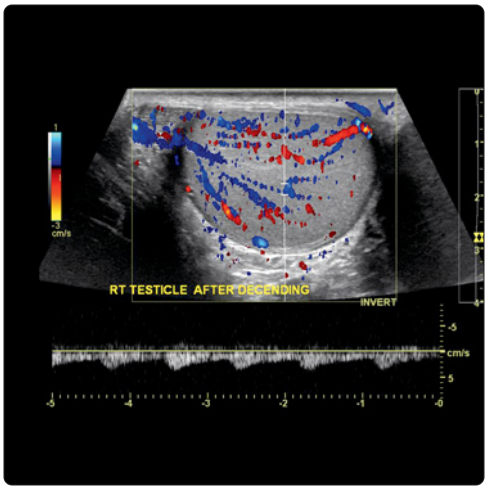
(Left) *Color Doppler ultrasound of the left testis of a male with 2 hours of acute scrotal pain shows completely absent intratesticular flow. Note the normal grayscale appearance of the testis. The time course and grayscale appearance suggest a likely viable testis (confirmed at orchiopexy).* **(Right)** *Color Doppler ultrasound of a male with right scrotal pain lasting over 24 hours shows an enlarged, heterogeneous and avascular right testis. As expected, the testis was not viable and an orchiectomy was performed.*





(Left) *Color Doppler ultrasound of the left testis shows an enlarged, heterogeneous, avascular testis with multiple hyperechoic regions. Torsion > 360° was shown at exploration, and pathology confirmed hemorrhagic necrosis.* **(Right)** *Sagittal grayscale ultrasound of right testis shows an enlarged, heterogeneous right testis with a striated appearance suggestive of partial infarcts. The patient had a history of partial testicular torsion.*





(Left) *Sagittal color Doppler ultrasound of both testes in a young male with right testicular pain shows an asymmetric blood flow with reduced flow to the right testis, as seen on color flow and spectral waveform. This was surgically confirmed to be partial (180°) right testicular torsion.* **(Right)** *Transverse color Doppler ultrasound of the right testis of the same patient after detorsion shows characteristic Doppler hyperemia.*

Segmental Infarction

KEY FACTS

IMAGING

- Rare lesion, but characteristic appearance suggests diagnosis in acute setting
  - Ultrasound: Elongated, geographic "mass" within superior aspect of testis
    - May be isoechoic in acute setting; hypoechoic lesion with peripheral echogenic rim is more typical
    - Avascular or hypovascular at color Doppler, peripheral hyperemic rim may be identified
  - MR: Utilize for confirmation, or if ultrasound equivocal
    - Well-defined, geographic, peripheral "mass" with peripheral enhancement

TOP DIFFERENTIAL DIAGNOSES

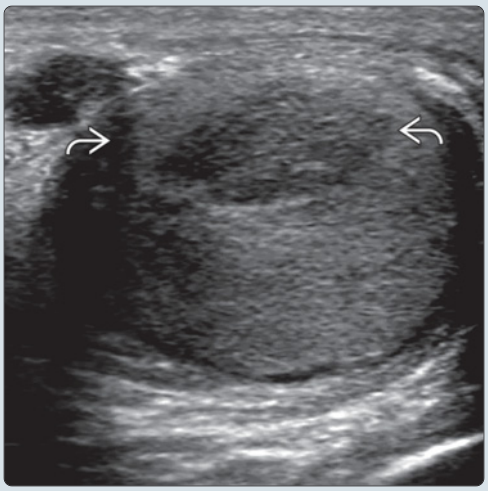
- Testicular neoplasm
  - Round mass, rarely presents in acute setting
- Testicular hematoma
  - Hypovascular, hypoechoic mass: Typically post trauma
- Testicular abscess
  - History (prior epididymo-orchitis, fever, leukocytosis, pyuria) aids diagnosis
- Testicular lymphoma and leukemia
  - Uni- or multifocal hypoechoic mass, post cessation of maintenance chemotherapy

CLINICAL ISSUES

- Rare cause for acute scrotum
  - Reported predisposing history: Vasculitis, sickle cell disease, hypercoagulable states, herniorrhaphy, varicocelectomy
  - Often idiopathic

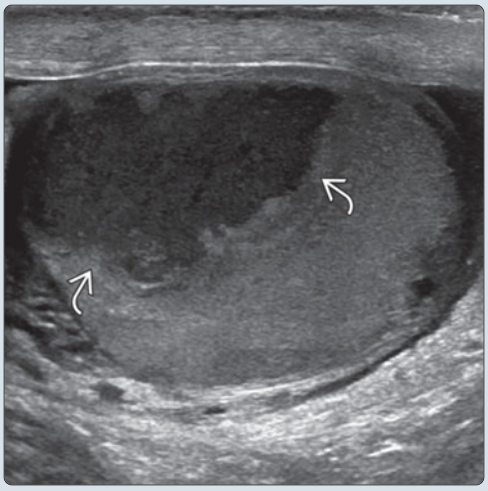
DIAGNOSTIC CHECKLIST

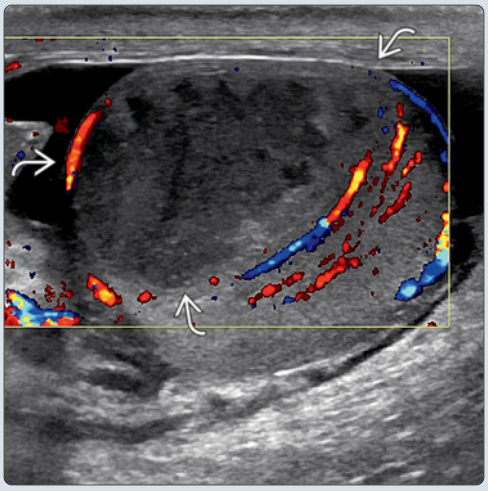
- Ultrasound (and clinical) follow-up needed to confirm imaging impression (and exclude testicular tumor)
  - MR may increase diagnostic confidence





(Left) *Transverse ultrasound performed on a 45-year-old man with acute scrotal pain shows an elongated, peripheral hypoechoic mass. Although the appearance strongly suggests segmental infarction, as is often the case, no predisposing history was elicited. Ultrasound follow-up and a confirmatory MR were recommended.* **(Right)** *Axial gadolinium-enhanced T1 C+ FS MR performed on the same patient shows a triangular, avascular mass with peripheral rim enhancement, characteristic features of segmental infarction.*





(Left) *Sagittal US performed on a 47-year-old man with 2 weeks of scrotal pain shows a peripheral, wedge-shaped hypoechoic mass. A prior US performed at the onset of pain was normal.* **(Right)** *Color Doppler US (same patient) shows the mass is avascular. Configuration of the lesion suggests segmental infarction, a diagnosis that prompted a work-up for vasculitis. Diagnosis of drug-induced (minocycline) vasculitis was ultimately confirmed, and symptoms resolved with steroids.*

TERMINOLOGY

Definitions

- Localized testicular infarct

IMAGING

General Features

- Best diagnostic clue
  - Elongated, geographic, iso- to hypoechoic, avascular testicular "mass" in 20- to 40-year-old man with acute scrotal pain
- Location
  - Typically upper portion of testis (less collateral blood supply)
- Size
  - Mean size: 9 mm (range: 3-19 mm)
- Peripheral, geographic lesion
  - Atypical round segmental infarcts may mimic tumor

MR Findings

- T2WI
  - Well-defined, peripheral, lesion
  - T2 intensity likely depends upon age of infarct, though majority present with ↓ T2 signal
- T1WI C+ FS
  - Nonenhancing geographic lesion, with peripheral enhancement
- MR advocated as confirmatory test for patients with equivocal ultrasound findings &/or confounding history

Ultrasonographic Findings

- Grayscale ultrasound
  - Peripheral, elongated lesion
    - May be isoechoic and barely perceptible at initial acute setting
    - Borders often better perceived as lesion becomes hypoechoic over time
      - Echogenic rim
- Color Doppler
  - Avascular or hypovascular peripheral lesion, perilesional hyperemia
    - Utility of microbubble contrast enhanced ultrasound for follow-up reported

Imaging Recommendations

- Best imaging tool
  - Acute setting: Color Doppler scrotal sonography
    - Scrotal MR for confirmation of equivocal cases

DIFFERENTIAL DIAGNOSIS

Testicular Neoplasm

- Round mass, rarely presents as acute scrotum
- Vascularity not useful for differentiating between subtypes

Testicular Hematoma

- Avascular mass, typically post trauma, though hemorrhage into preexisting tumor possible
- Ultrasound surveillance critical for confirmation

Testicular Abscess

- Clinical history (prior epididymo-orchitis, fever, leukocytosis, pyuria) should be elicited

Testicular Lymphoma and Leukemia

- May be multifocal, typically post cessation of maintenance chemotherapy ("blood-gonad barrier")

PATHOLOGY

Gross Pathologic & Surgical Features

- Acute: Acute coagulative necrosis, red cell and fibrin extravasation
- Chronic: Sclerosed seminiferous tubules, and hyalinized interstitial fibrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute scrotal pain

Demographics

- Age
  - Onset later than "classic" testicular infarction: Typically 2nd-4th decades

Natural History & Prognosis

- Dependent upon predisposing condition (if present)

Treatment

- Symptomatic (analgesia)

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Elongated, peripheral, avascular mass in patient with acute scrotal pain

Reporting Tips

- Ultrasound follow-up to confirm slow regression (and exclude tumor)

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Tubular Ectasia

KEY FACTS

TERMINOLOGY

- Dilated rete testis
- Cystic transformation of rete testis

IMAGING

- Frequently bilateral
  - Usually asymmetric involvement
- Branching tubules converging at mediastinum testis
  - Dilated tubules create lace-like or fishnet appearance
- Adjacent parenchyma is normal
- Associated ipsilateral spermatoceles are common
- Tubules are avascular and fluid filled
  - No flow on color Doppler imaging
- MR performed for confirmation if cystic malignant neoplasm cannot be ruled out

TOP DIFFERENTIAL DIAGNOSES

- Testicular carcinoma
  - Mixed germ cell tumors with teratomatous components will often have cystic areas
  - Does not form network of tubules
- Intratesticular varicocele
  - Characteristic color flow on Doppler
- Testicular infarct
  - Avascular wedge-shaped area with sharp borders

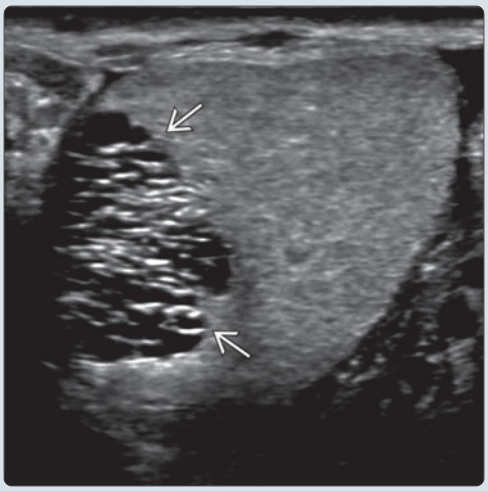
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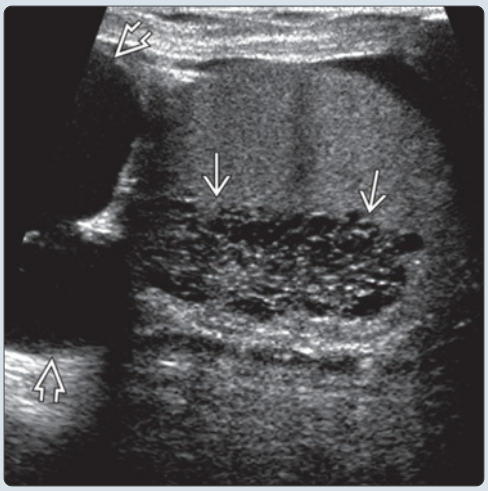
- Generally nonpalpable and asymptomatic
- May be found when ultrasound performed for a related issue, such as an epididymal cyst

DIAGNOSTIC CHECKLIST

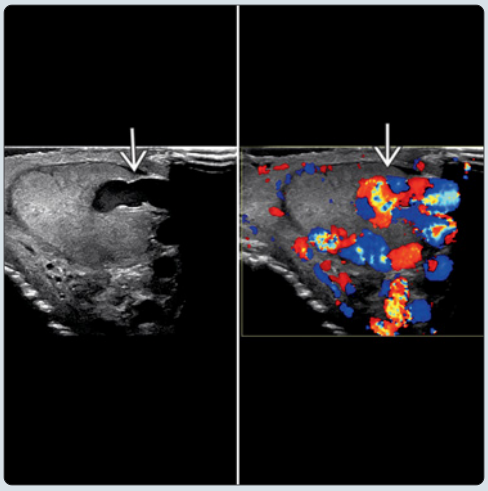
- Important to distinguish tubular ectasia from malignancy to prevent unnecessary orchiectomy

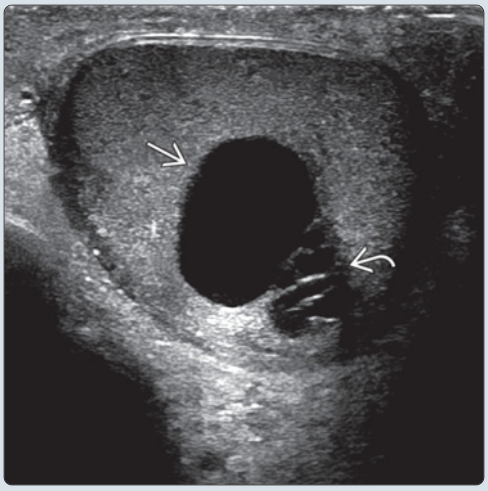
(Left) *Transverse ultrasound of the left testis shows serpiginous, anechoic tubules oriented along the mediastinum testis, a finding pathognomonic for dilated rete testis. The configuration and location should prompt a definitive imaging diagnosis of this benign entity.* **(Right)** *Sagittal ultrasound performed on an elderly man shows dilated rete testes and large spermatoceles. Both entities are associated with efferent duct and epididymal duct obstruction.*





(Left) *Sagittal ultrasound of an elderly man with severe right heart dysfunction shows a tubular anechoic structure within the mediastinum testis. Color Doppler confirms a huge varicocele with an intratesticular component, a potential mimic for dilated rete testis.* **(Right)** *Ultrasound of 31-year-old man with cystic fibrosis shows dilated rete testis and a simple testicular cyst, potentially as a result of seminal vesicle agenesis and the abnormal epididymal fluid transport that is a characteristic feature of this patient population.*





TERMINOLOGY

Synonyms

- Dilated rete testis
- Cystic transformation of rete testis

IMAGING

General Features

- Best diagnostic clue
  - Variably sized network of dilated tubules near mediastinum testis with no flow on color Doppler
- Location
  - Posterior near mediastinum testis
  - Frequently bilateral
- Size
  - Can replace large portion of normal parenchyma
  - Variably sized cystic spaces
- Morphology
  - Branching tubules converging at mediastinum testis
  - Characteristic appearance and location make it possible to distinguish this benign condition from malignancy

Ultrasonographic Findings

- Grayscale ultrasound
  - Longitudinal plane shows branching tubular structures along mediastinum
    - Dilated tubules create lace-like or fishnet appearance
  - Adjacent parenchyma is normal
    - Differentiates it from cystic malignant masses, which have abnormal rind of parenchyma
  - Associated ipsilateral spermatoceles are common
    - May also see intratesticular cysts
- Color Doppler
  - Tubules are avascular and fluid filled, hence no color flow
  - Normal flow in adjacent testicular parenchyma

MR Findings

- MR performed for confirmation if cystic malignant neoplasm cannot be ruled out
- T1 and proton density weighted Images: Hypointense to testis
- T2WI: Iso- to hyperintense to testis: Nearly invisible
  - Malignant testicular neoplasms: Solid portions hypointense on T2WI; have dark fibrous capsule

Imaging Recommendations

- Best imaging tool
  - High-resolution US (≥ 7.5 MHz) is imaging modality of choice
- Protocol advice
  - Longitudinal plane shows morphology of tubules far better than transverse plane
    - Appears more mass-like and has greater likelihood of causing confusion when viewed in transverse plane
  - Always use color Doppler to look for areas of abnormal parenchymal flow

DIFFERENTIAL DIAGNOSIS

Testicular Carcinoma

- Mixed germ cell tumors with teratomatous components will often have cystic areas
- Does not form network of tubules
- Surrounding parenchyma is abnormal
  - Will have abnormal flow on color Doppler

Intratesticular Varicocele

- Multiple intratesticular anechoic serpiginous tubules with characteristic color flow on Doppler, particularly during Valsalva maneuver
- Will have associated extratesticular varicocele

Testicular Infarct

- Avascular hypoechoic mass, as sequelae of any previous vascular insult
- Sharp, linear borders demarcate area

PATHOLOGY

General Features

- Etiology
  - Partial or complete efferent ductule obstruction → ectasia → eventually cystic transformation
  - May be associated with epididymal obstruction due to inflammation or trauma
- Associated abnormalities
  - Spermatocele
  - Epididymal cyst
  - Intratesticular cyst

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Generally nonpalpable and asymptomatic
  - May be found when doing ultrasound for a related issue, such as epididymal cyst or spermatocele

Demographics

- Age
  - Middle-aged to elderly men most commonly affected

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Important to distinguish tubular ectasia from malignancy to prevent unnecessary orchiectomy

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Testicular Microlithiasis

KEY FACTS

TERMINOLOGY

- Microcalcifications located within spermatic tubules

IMAGING

- Discrete, punctate, nonshadowing echogenic foci scattered within testicular parenchyma
- Majority are idiopathic; previous infection or trauma may also be responsible
- Adjacent hypoechoic foci, if seen, may represent neoplasia
- High resolution US (≥ 7.5 MHz) is modality of choice

TOP DIFFERENTIAL DIAGNOSES

- Large cell calcifying Sertoli cell tumor
- Testicular granuloma
- Testicular vascular calcification
- "Burned-out" testicular cancer

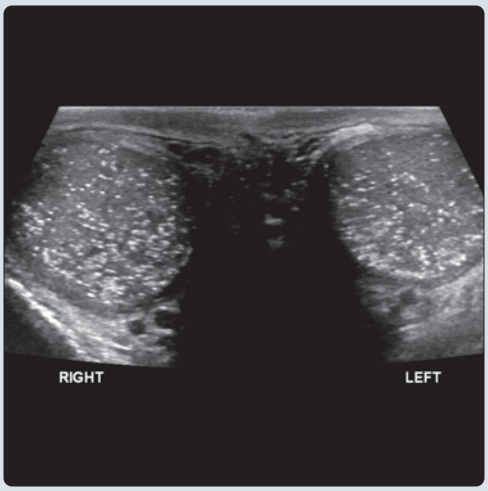
PATHOLOGY

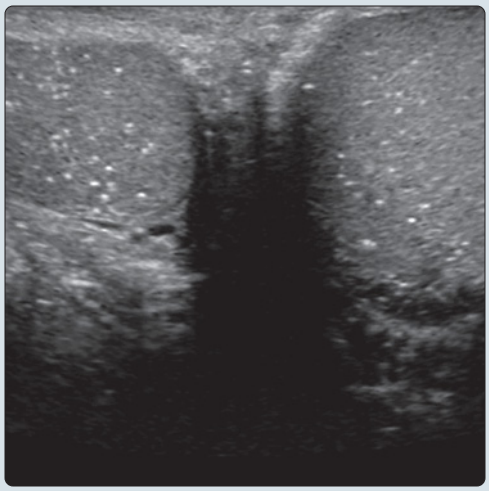
- Coexistent germ cell tumor in 18-75% of symptomatic males referred for ultrasound
- Associated with intratubular germ cell neoplasia (IGCN): Germ cell version of breast carcinoma in situ

CLINICAL ISSUES

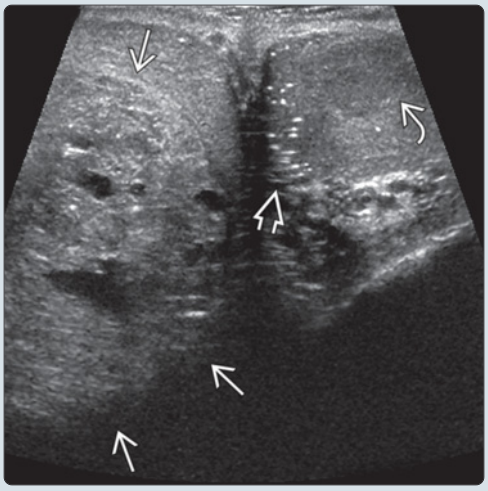
- Presence of microlithiasis alone in absence of other risk factors is not indication for further sonographic surveillance or biopsy
- US is recommended in follow-up of patients with ↑ risk of germ cell tumor: Personal/family history of germ cell tumor, maldescent or undescended testes, orchidopexy, testicular atrophy

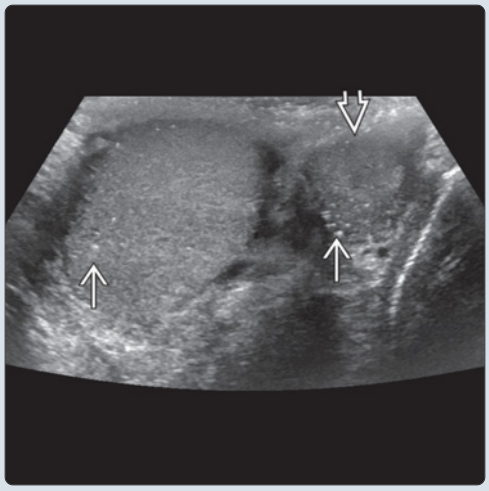
(Left) *Transverse grayscale ultrasound of both testes demonstrates extensive microlithiasis. Microliths measure 1-3 mm and do not shadow.* **(Right)** *Ultrasound performed on a 31-year-old man with scrotal pain shows bilateral microlithiasis. Microlithiasis is strictly defined as ≥ 5 microliths per image. Without additional risk factors for germ cell tumor, follow-up by physical examination suffices.*





(Left) *Ultrasound performed on a 43-year-old man with palpable masses and ↑ βHCG and aFP shows a huge right mixed germ cell tumor, a left seminoma, and surrounding microlithiasis. Background intratubular germ cell neoplasia was also shown at pathology.* **(Right)** *Transverse grayscale ultrasound of both testes demonstrates an atrophic left testis with bilateral microlithiasis. Annual screening ultrasound is indicated due to an increased risk for cancer in an atrophic testis with microlithiasis.*





TERMINOLOGY

Definitions

- Testicular microlithiasis (TML): 5 or more nonshadowing echogenic testicular foci measuring 1-3 mm

IMAGING

General Features

- Best diagnostic clue
  - Discrete, punctate, nonshadowing echogenic foci scattered throughout testes
- Location
  - Unilateral or bilateral
- Size
  - 1-3 mm
- Morphology
  - Asymmetrically distributed, peripheral predominance, not palpable
  - Microcalcifications composed of hydroxyapatite formed within spermatic tubule lumina
  - Multilayered envelope, composed of stratified collagen fibers: Matrix is thought to be cause for absent acoustic shadowing

Ultrasonographic Findings

- Grayscale ultrasound
  - Small hyperechoic foci diffusely scattered throughout testicular parenchyma
    - 1-3 mm echogenic foci, no shadowing: May occasionally see "comet tail" artifact
    - May be peripheral or segmental
    - Clusters of microliths may represent testicular tumor (carcinoma in situ) without soft tissue mass
      - Clusters of microliths adjacent to solid mass suggest germ cell tumor

Imaging Recommendations

- Best imaging tool
  - High resolution US (≥ 7.5 MHz)

DIFFERENTIAL DIAGNOSIS

Large Cell Calcifying Sertoli Cell Tumor

- Gonadal stromal tumor, often bilateral and multifocal
- Most common cause of intratesticular macrolithiasis, mass may be almost completely calcified

Testicular Granuloma

- TB epididymo-orchitis may produce intrascrotal calcifications and scrotal sinus tract

Testicular Vascular Calcification

- Linear echogenicity, history of diabetes

"Burned-Out" Primary Testicular Cancer

- Ill-defined remnant testicular scar, macro/microcalcifications and retroperitoneal adenopathy

PATHOLOGY

General Features

- Etiology
  - Exact cause is unknown
    - Sertoli cell dysfunction postulated
- Genetics
  - Associated with Klinefelter syndrome, Down syndrome, male pseudohermaphroditism, cryptorchidism, McCune-Albright syndrome, Peutz-Jeghers syndrome

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic, typically incidental ultrasound finding

Demographics

- Age
  - Usually seen in older boys and adolescents
  - Rare in boys younger than 2 years
- Epidemiology
  - Prevalence rates vary depending upon study population: Prevalence of 5.6% reported in large asymptomatic screening study of 17- to 35-year-old males

Natural History & Prognosis

- ↑ prevalence of microlithiasis in early retrospective ultrasound studies of testicular carcinoma suggested causation and prompted imaging intensive surveillance
- Strength of association subsequently questioned: Testicular carcinoma is relatively rare, and microlithiasis is common in asymptomatic males
- Meta-analysis: Presence of microlithiasis without other risk factors is not indication for routine sonographic screening or biopsy (self-exam sufficient)
- Follow-up US recommended in patients with risk factors: Personal/family history of germ cell tumor, maldescent or undescended testes, orchidopexy, testicular atrophy

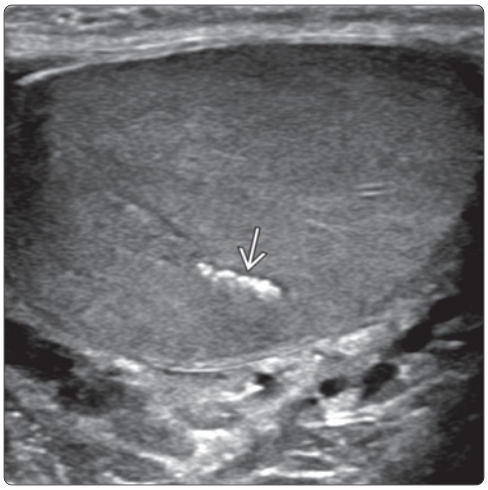
Treatment

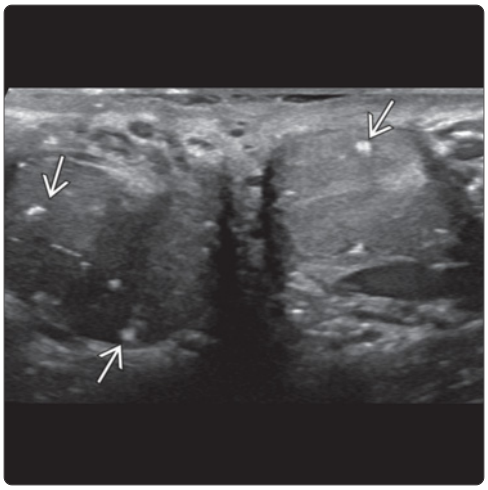
- No treatment or follow-up or biopsy in absence of risk factors of testicular malignancy
- If associated with hypoechoic mass, surgical biopsy or orchiectomy

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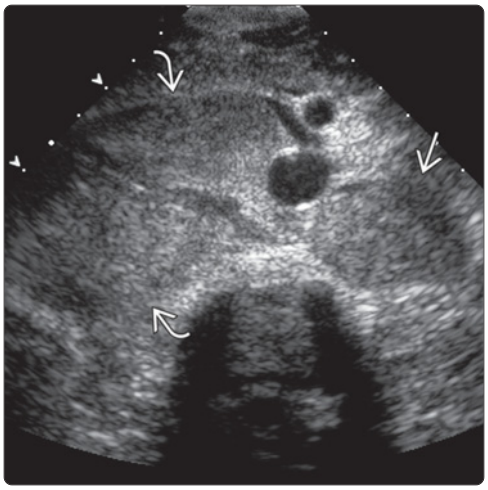
(Left) *Testicular ultrasound performed on a 37-year-old man with diabetes shows linear echogenicity ⇨ oriented along the mediastinum testis. Location and configuration suggest vessel calcification rather than isolated microlithiasis.* **(Right)** *Scrotal ultrasound of a 21-year-old man with Carney complex shows multiple macrocalcifications ⇨, a characteristic appearance (and syndromic association) of large cell calcifying Sertoli cell tumors. These extremely rare sex-cord stromal tumors are usually benign.*



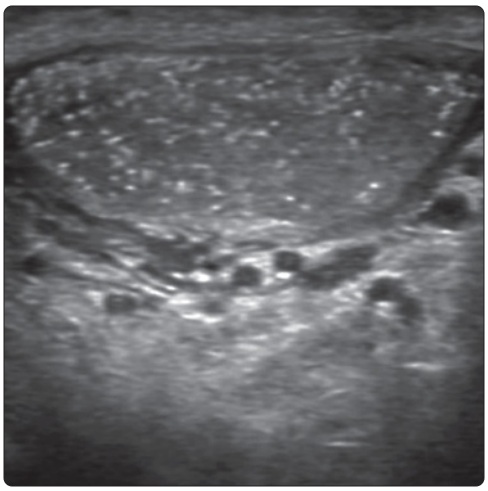


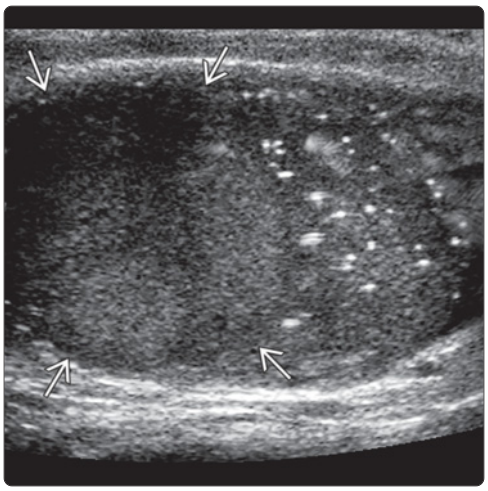
(Left) *Ultrasound of a young male with scrotal discomfort and fatigue shows clustered macrocalcifications ⇨ and microcalcifications ⇨ and vague background hypoechogenicity ⇨.* **(Right)** *Scrotal findings prompted retroperitoneal ultrasound, which showed massive paraaortic ⇨ and paracaval adenopathy ⇨. Clustered testicular calcifications and ill-defined hypoechogenicity are typical features of a burned-out germ cell tumor (GCT) (subsequently confirmed in this patient).*

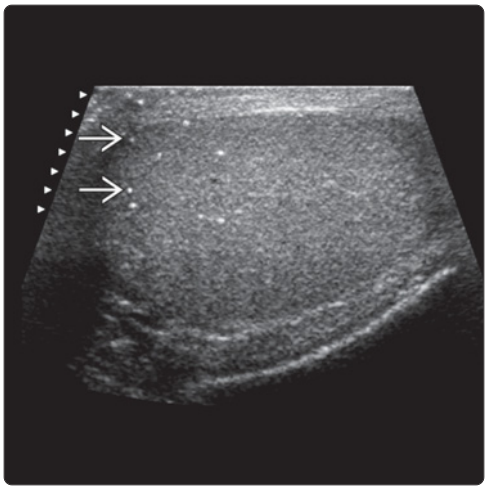


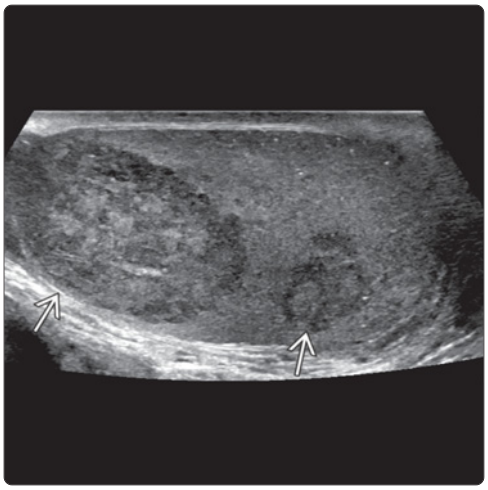


(Left) *Ultrasound of a young male with a history of a contralateral GCT shows an atrophic testis post orchiopexy with innumerable microliths. Ultrasound surveillance was performed because of the history of prior GCT and cryptorchidism and not microlithiasis per se.* **(Right)** *Surveillance ultrasound of a male post orchiopexy shows a uniform isoechoic seminoma ⇨ and background microlithiasis. A study 3 years prior was negative.*

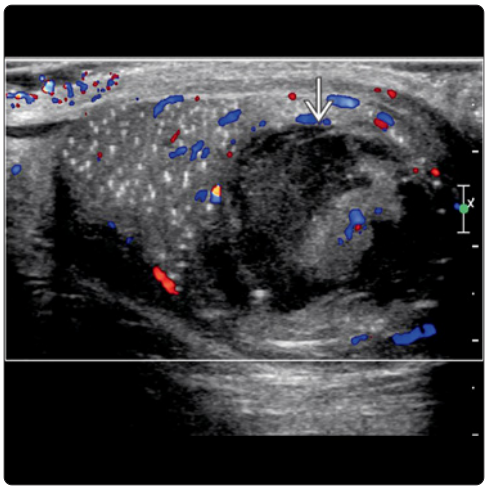


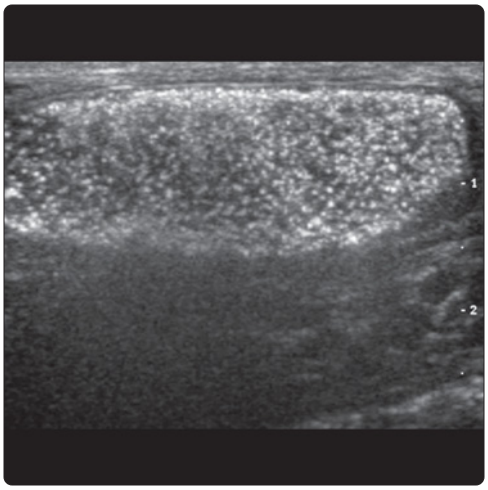




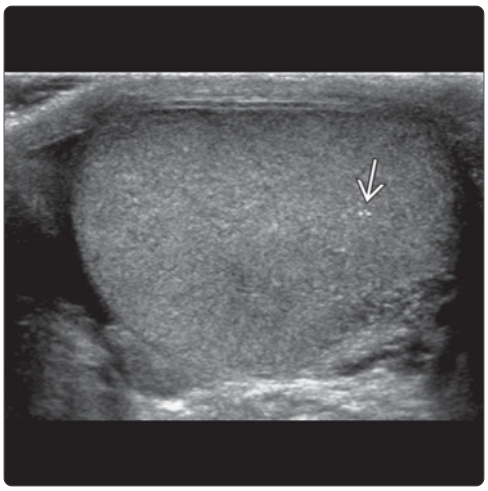


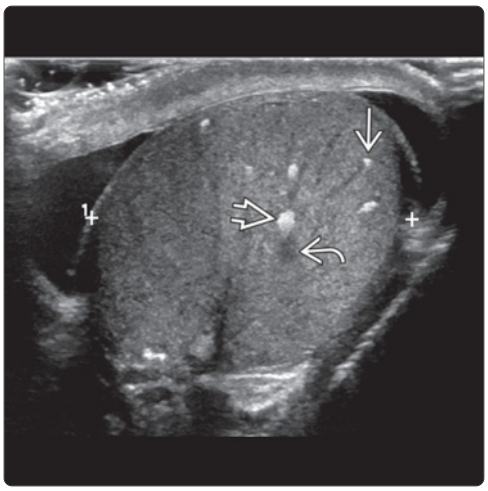
(Left) *Transverse grayscale ultrasound of the left testis demonstrates clustered microlithiasis. The patient had a history of right orchiectomy for GCT and was being followed-up by ultrasound on an annual basis.* **(Right)** *Sagittal grayscale ultrasound follow-up on the same patient after 11 years demonstrates 2 new hypoechoic masses. Pathology confirmed multifocal seminoma.*





(Left) *Sagittal color Doppler ultrasound image of the right testis demonstrates a large hypoechoic mass in the background of extensive microlithiasis. Pathology confirmed a classic seminoma.* **(Right)** *Sagittal grayscale ultrasound of the right testis demonstrates diffuse extensive microlithiasis. This limits adequate assessment of the testicular parenchyma for tumor, hence these must be referred to specialist centers for alternate methods of future screening.*





(Left) *Transverse grayscale ultrasound of the left testis demonstrates an isolated microlith. This does not meet the definition of testicular microlithiasis and is likely a sequela of prior infection or trauma.* **(Right)** *Transverse grayscale ultrasound of the left testis demonstrates micro and macrocalcifications. Note shadowing posterior to the macrocalcification. Patients with intratesticular macrocalcifications should be considered to be at higher risk of a testicular malignancy.*

Germ Cell Tumors

KEY FACTS

IMAGING

- Most common neoplasm in males aged 15-34 years
- Mostly unilateral; contralateral tumor develops eventually in 8%
- Seminoma is most common pure germ cell tumor of testis
- On ultrasound, seminomas are usually well defined, hypoechoic, solid without calcification or tunica invasion
- Tumor < 1.5 cm is commonly hypovascular, and tumors > 1.6 cm are more often hypervascular
- Embryonal cell carcinomas are aggressive tumors, may invade tunica albuginea and distort testicular contour
- US is used to identify and characterize scrotal mass; CT or MR for metastatic staging; PET to evaluate post-treatment residual masses
- Lymph nodes < 1 cm are suspicious if located in typical drainage areas; left renal hilus and right retrocaval in location

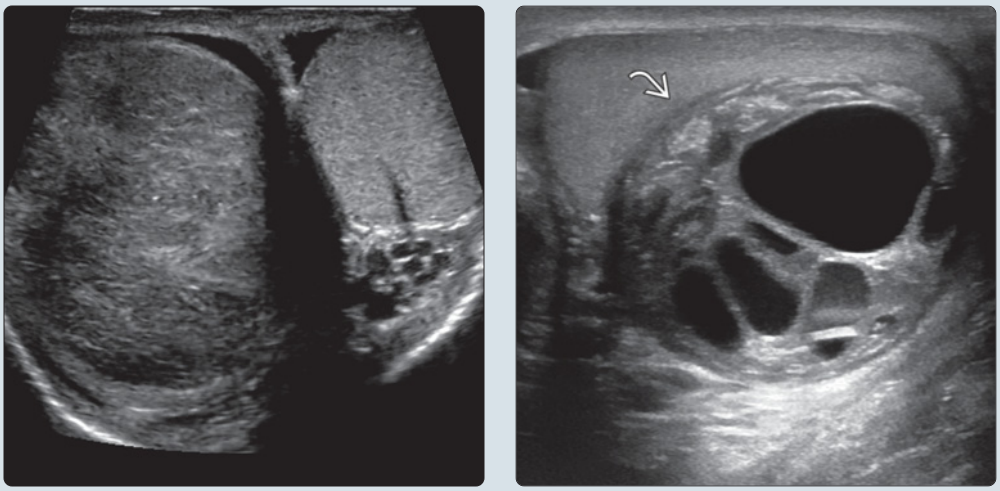
PATHOLOGY

- Associated with cryptorchidism, previous contralateral cancer; possible association with mumps orchitis, microlithiasis, and family history of tumor

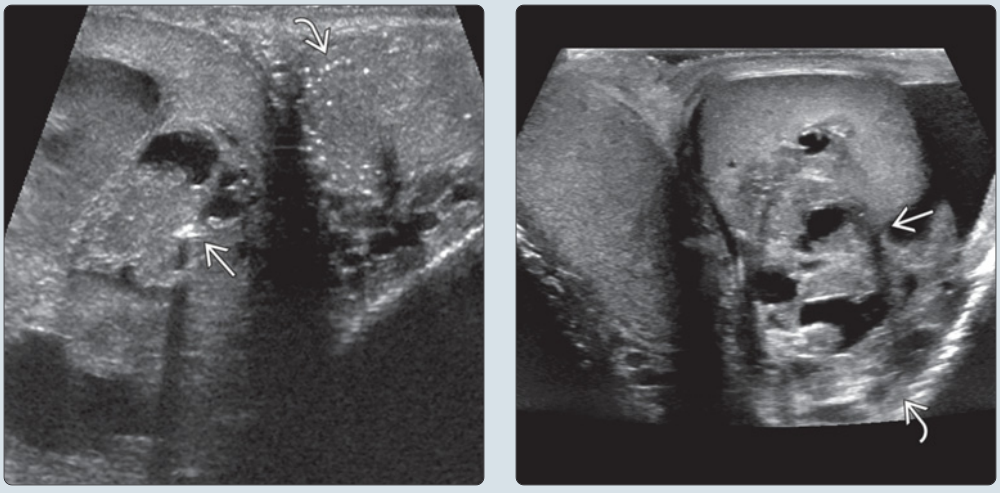
CLINICAL ISSUES

- Beta hCG is elevated in pure or mixed embryonal carcinoma or choriocarcinoma; also in 15-20% of those with advanced seminoma
- Elevated AFP levels above 10,000 µg/L are found almost exclusively in patients with nonseminomatous germ cell tumors (NSGCTs) (not seen with pure seminomas) and hepatocellular carcinoma
- LDH has independent prognostic significance: Increased levels reflect tumor burden, growth rate, and cellular proliferation

(Left) *US of a 27-year-old man shows a hypoechoic mass that replaces & enlarges the right testis. The tunica was intact despite the size of the mass. Orchiectomy confirmed a pure seminoma.* **(Right)** *US shows a cystic & solid testicular mass (orchiectomy-confirmed MGCT composed of 90% teratoma and 10% embryonal tumor). Most testicular cancers are seminomas or MGCTs. Cystic change suggests nonseminomatous tumors. Tumor markers may ↑ specificity, but ultimate diagnosis occurs with orchiectomy.*



(Left) *US of a 43-year-old man shows a large mixed cystic and solid right testicular mass containing calcifications. Note the left microlithiasis. Bilateral orchiectomy was performed for what was ultimately shown to be a tunica invasive right MGCT and small left seminoma (not shown).* **(Right)** *US of a 19 year old with back pain and a retroperitoneal mass shows a mixed cystic and solid left testicular lesion. It extends through tunica and abuts up to scrotal wall. A MGCT was confirmed at orchiectomy.*



TERMINOLOGY

Abbreviations

- Germ cell tumor (GCT)
- Mixed germ cell tumor (MGCT)

Definitions

- Malignant germ cell tumor of testis

IMAGING

General Features

- Best diagnostic clue
  - Discrete hypoechoic or mixed echogenic testicular mass, ± vascularity
- Morphology
  - Most common neoplasm in males aged 15-34 years
  - Mostly unilateral; contralateral tumor develops eventually in 8%
  - Seminoma is most common pure germ cell tumor of testis
    - Bilateral in 1-3%, almost always asynchronous

Ultrasonographic Findings

- Grayscale ultrasound
  - Seminoma
    - Typically well-defined, homogeneously hypoechoic, solid mass without calcification or tunica invasion
      - With high-resolution US, some lesions have heterogeneous echo pattern, ± lobulation
    - May be solitary or multifocal masses
    - May rarely undergo necrosis and appear partly cystic
  - Teratoma/teratocarcinoma
    - Well-defined, anechoic/complex heterogeneous cystic mass
    - Cystic areas, calcification (cartilage, immature bone) ± fibrosis
  - Embryonal cell carcinoma
    - Heterogeneous, predominantly solid mass, of mixed echogenicity
    - Poorly marginated, 1/3 have cystic necrosis
    - Coarse calcification infrequently seen
    - Embryonal cell carcinoma are aggressive tumors, may invade tunica albuginea and distort testicular contour
  - Yolk sac tumor
    - Most common testicular neoplasm in pediatric population
    - Seen in pure form in children, while almost always admixed with other germ cell elements in adults
  - Choriocarcinoma
    - Mixed echogenicity, heterogeneous mass
    - Cystic areas with calcification common
    - Hemorrhage with focal necrosis is typical feature of choriocarcinoma
    - Usually rare in pure form; occurs with mixed GCT in 8%
    - Highly malignant
- Color Doppler
  - Tumor < 1.5 cm is commonly hypovascular, and tumors > 1.6 cm are more often hypervascular
  - Disorganized flow is typical
  - Cystic areas are avascular

CT Findings

- CECT
  - Useful for staging retroperitoneal, nodal, and pulmonary metastases
  - Low-attenuation nodes
  - Even nodes < 1 cm suspicious if located in typical drainage areas; left renal hilus and right retrocaval in location
  - Helpful to identify retroperitoneal recurrence &/or "growing teratoma" syndrome

MR Findings

- T2WI
  - Useful for identifying nodal metastases
  - Moderate high signal intensity lymphadenopathy in retroperitoneum
  - Seminoma is multinodular, hypointense
  - Intratumoral low signal, band-like structures representing fibrovascular septa

Nuclear Medicine Findings

- PET
  - Helpful to reduce false-negative CT study
    - May aid in differentiating residual tumor from scar in treated patients

Imaging Recommendations

- Best imaging tool
  - US to identify and characterize scrotal mass; CT or MR for metastatic staging; PET to evaluate post-treatment residual masses
- Protocol advice
  - High-frequency (≥ 10 MHz) linear array transducer

DIFFERENTIAL DIAGNOSIS

Epidermoid Cyst

- Cystic cavity lined by stratified squamous epithelium
- Onion skin appearance due to alternating layers of keratin and desquamated squamous cells
- May have calcified rim, no enhancement on MR

Lymphoma

- Older age group, most common tumor of testes in men > 60 years
- 50% of cases bilateral, often multiple lesions, associated with lymphadenopathy, masses elsewhere
- Hypoechoic and hypervascular on color Doppler

Subacute Hematoma

- History of trauma, associated hematocele
- Hypoechoic on US

Segmental Infarct

- Acute pain, no palpable mass
- Infarct is typically hypoechoic, focal avascular area on color Doppler

Focal Orchitis

- Irregular hypoechoic area within testis, enlarged epididymis
- Increased vascularity on color Doppler without displacement of vessels

- Reactive hydrocele with low-level echoes, scrotal wall thickening

PATHOLOGY

General Features

- Etiology
  - Associated with cryptorchidism, previous contralateral cancer; possible association with mumps orchitis, microlithiasis, and family history of tumor
- Genetics
  - Family history increases risk
- Associated abnormalities
  - Gynecomastia, prepubescent virilization
- 95% of testicular tumors are malignant germ cell tumors
- Single histologic subtype in 65% of tumors (seminoma most common)
- Multiple subtypes in 35%

Staging, Grading, & Classification

- Stage I (A): Tumor confined to testis
- Stage II (B): Tumor metastatic to nodes below diaphragm
- Stage IIA (B1): Retroperitoneal node enlargement < 2 cm (5 cm^3)
- Stage IIB (B2): Retroperitoneal node enlargement > 2 cm x < 5 cm (10 cm^3)
- Stage IIC (B3): Retroperitoneal node enlargement > 5 cm
- Stage III (C): Tumor metastatic to lymph nodes above diaphragm
- Stage IIIA (C1): Metastases confined to lymphatic system
- Stage IIIB or IV: Extranodal metastases

Gross Pathologic & Surgical Features

- Solid or solid/cystic intratesticular mass
- 10-15% have epididymis or spermatic cord involvement
- Bilateral in 2-3% of cases

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Palpable mass in testis, painless enlarging mass
  - Dull pain (27%)
  - Acute pain (10%)
- Other signs/symptoms
  - Gynecomastia, virilization
- Clinical profile
  - Young male with palpable testicular mass, elevated serum tumor markers
    - 3 serum tumor markers have established roles in management of men with testicular germ cell tumors
    - Beta subunit of human chorionic gonadotropin (beta-hCG)
      - Elevated in pure or mixed embryonal carcinoma or choriocarcinoma; also in 15-20% of those with advanced seminoma
    - Alpha fetoprotein (AFP)
      - Elevated levels above 10,000 µg/L are found almost exclusively in patients with nonseminomatous germ cell tumors (NSGCTs) (not seen with pure seminomas)
      - Elevated AFP is most often seen with yolk sac tumor and less often embryonal tumors
    - Lactate dehydrogenase (LDH)
      - Elevated in 40-60% of men with testicular GCTs

Demographics

- Age
  - Seminomatous tumor: Average 40.5 years
  - Nonseminomatous tumor: 20-30 years
  - Endodermal sinus tumor/teratoma: 1st decade
- Ethnicity
  - Increased incidence in Caucasian and Jewish males
- Epidemiology
  - Most common cancer in men aged 15-34
  - 1% of all cancers in men, 4-6% of all male genitourinary tumors, 4th most common cause of death from malignancy in men aged 15-34 years
  - Seminomas most common in men 35-39 years old, most common tumor in undescended testis
  - Seminomas rare before 10 years and after 60 years

Natural History & Prognosis

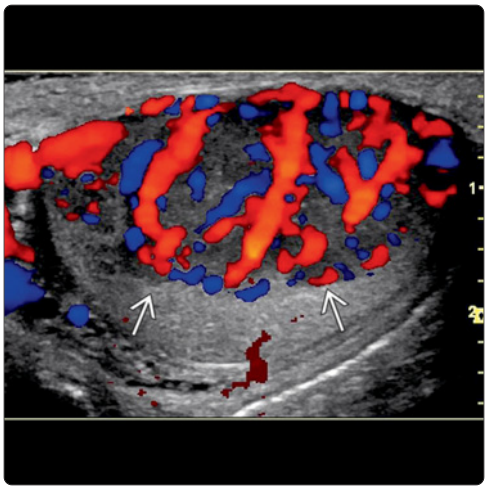
- 95% 5-year survival rate overall
- Metastases at presentation is seen in 4-14% of individuals
  - Distant spread occurs along testicular lymphatics
  - Hematogenous dissemination (usually late) to lung, bone, brain
  - Choriocarcinoma has a proclivity for early hematogenous spread especially to brain, death usually within 1 year of diagnosis
- Growing teratoma syndrome: Evolution of mixed germ cell tumor into mature teratoma after chemotherapy (in 40%) followed by interval growth despite maintaining a benign histologic type

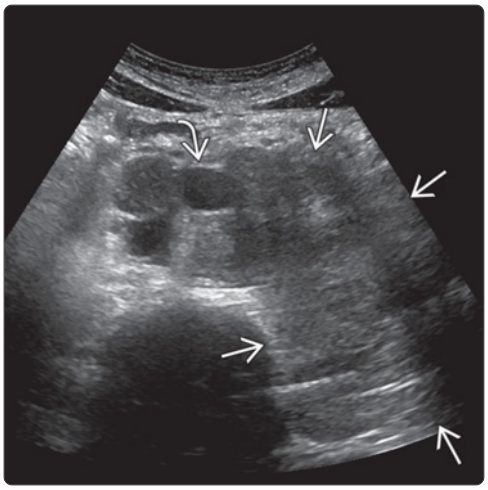
Treatment

- Seminoma very sensitive to radiotherapy ± chemotherapy
- Radical orchiectomy; retroperitoneal node dissection for nonseminomatous tumor
- Radiotherapy or chemotherapy for metastatic disease

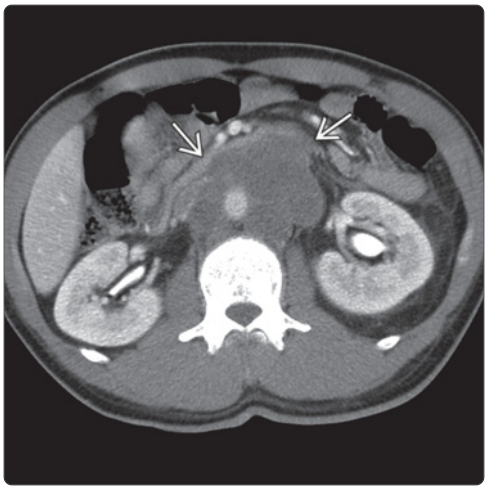
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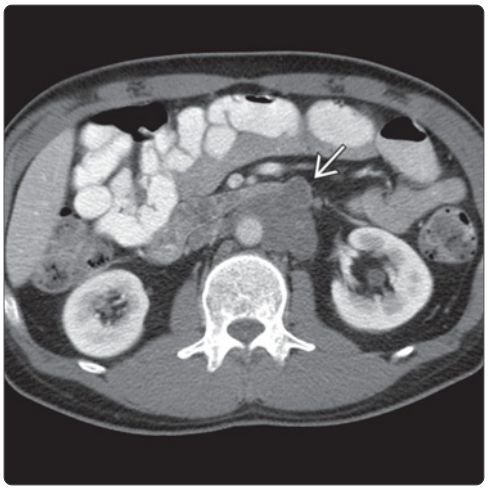
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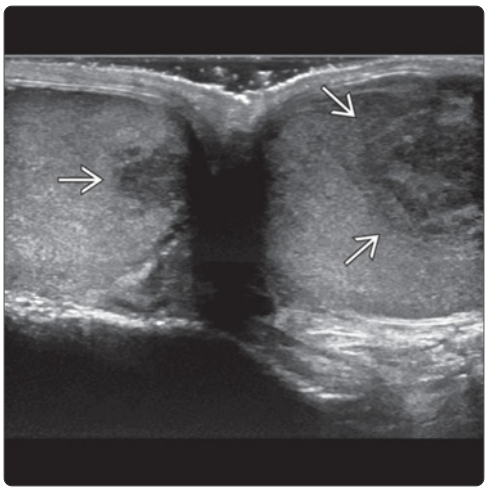


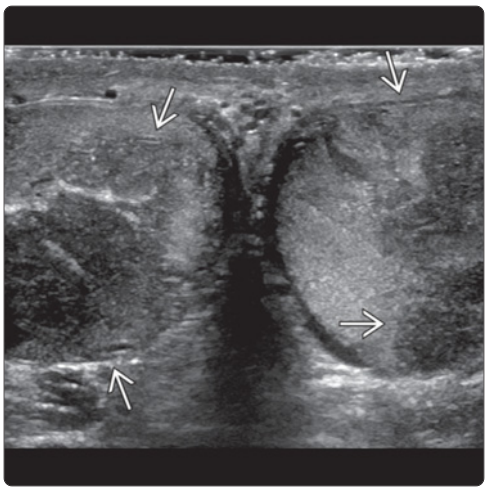
(Left) *Scrotal color Doppler ultrasound of a young male with lung, liver, and brain metastases and ↑ LDH and βHCG shows a vascular left testicular mass. The Doppler appearance is nonspecific, but the elevated tumor markers indicate a MGCT.* **(Right)** *Transabdominal ultrasound of the same patient shows bulky paraaortic adenopathy. The aorta is displaced anteriorly by tumor.*





(Left) *CECT of the same patient confirms bulky retroperitoneal adenopathy. Testicular carcinoma is routinely staged with CT: Awareness of patterns of nodal spread is critical, and even small nodes in characteristic locations should be viewed with concern.* **(Right)** *Post-chemotherapy CT of the same patient shows diminished enhancement of a smaller conglomerate nodal mass. Residual mature teratoma was resected at retroperitoneal lymph node dissection.*





(Left) *Ultrasound of a young black male shows bilateral testicular masses. The differential diagnosis included multifocal GCT and lymphoma, but sarcoidosis was strongly suggested given characteristic mediastinal adenopathy on a prior CT. Sarcoid was (unfortunately) confirmed at orchiectomy.* **(Right)** *Ultrasound of an infertile male shows bilateral hypoechoic masses. The same differential as the prior patient may be considered, but a history of congenital adrenal hyperplasia indicates adrenal rests.*

T | Primary Tumor

TNM	Definitions
pTX	Primary tumor cannot be assessed (if no radical orchiectomy has been performed, TX is used)
pT0	No evidence of primary tumor (e.g., histologic scar in testis)
pTis	Intratubular germ cell neoplasia (carcinoma in situ)
pT1	Tumor limited to testis and epididymis without vascular/lymphatic invasion; tumor may invade into tunica albuginea but not tunica vaginalis
pT2	Tumor limited to testis and epididymis with vascular/lymphatic invasion, or tumor extending through tunica albuginea with involvement of tunica vaginalis
pT3	Tumor invades spermatic cord ± vascular/lymphatic invasion
pT4	Tumor invades scrotum ± vascular/lymphatic invasion

Except for pTis and pT4, extent of primary tumor is classified by radical orchiectomy. For this reason, a pathologic stage is usually assigned. TX may be used for other categories in the absence of radical orchiectomy.

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
Clinical	
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis with a lymph node mass ≤ 2 cm in greatest dimension; or multiple lymph nodes, none > 2 cm in greatest dimension
N2	Metastasis with a lymph node mass > 2 cm but ≤ 5 cm in greatest dimension; or multiple lymph nodes, any 1 mass > 2 cm but ≤ 5 cm in greatest dimension
N3	Metastasis with a lymph node mass > 5 cm in greatest dimension
Pathological	
pNX	Regional lymph nodes cannot be assessed
pN0	No regional lymph node metastasis
pN1	Metastasis with a lymph node mass ≤ 2 cm in greatest dimension and ≤ 5 nodes positive, none > 2 cm in greatest dimension
pN2	Metastasis with a lymph node mass > 2 cm but ≤ 5 cm in greatest dimension; or > 5 nodes positive, none > 5 cm; or evidence of extranodal extension of tumor
pN3	Metastasis with a lymph node mass > 5 cm in greatest dimension

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis
M1a	No regional nodal or pulmonary metastasis
M1b	Distant metastasis other than to nonregional lymph nodes and lungs

Adapted from 7th edition AJCC Staging Forms (2010).

S | Serum Tumor Markers

TNM	Definitions
SX	Tumor marker studies not available or not performed
S0	Tumor marker study levels within normal limits
S1	LDH < 1.5x normal **and** β-hCG < 5,000 IU/L **and** AFP < 1,000 ng/mL
S2	LDH 1.5-10x normal **or** β-hCG 5,000-50,000 IU/L **or** AFP 1,000-10,000 ng/mL
S3	LDH >10x normal **or** β-hCG > 50,000 IU/L **or** AFP > 10,000 ng/mL

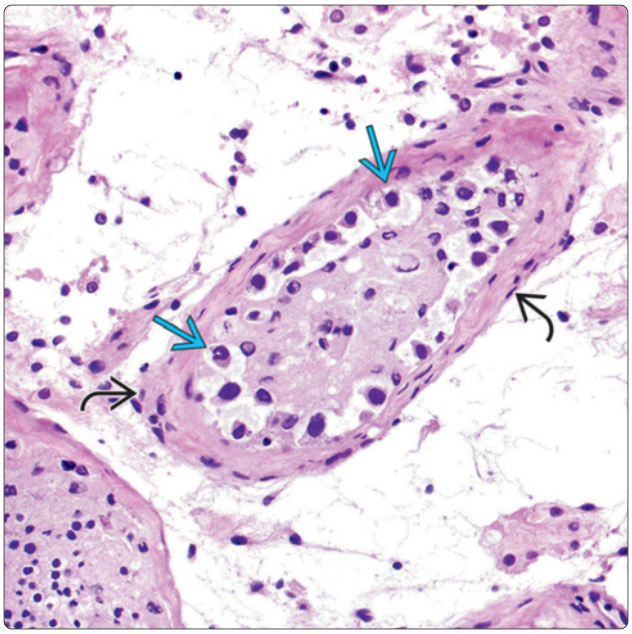
Serum tumor markers are used as part of tumor stage grouping in testicular cancer. LDH = lactate dehydrogenase, hCG = human chorionic gonadotropin, AFP = α-fetoprotein.

AJCC | Stages/Prognostic Groups

Stage	T	N	M	S (Serum Tumor Markers)
0	pTis	N0	M0	S0
I	pT1-4	N0	M0	SX
IA	pT1	N0	M0	S0
IB	pT2	N0	M0	S0
	pT3	N0	M0	S0
	pT4	N0	M0	S0
IS	Any pT/TX	N0	M0	S1-3 (measured post orchiectomy)
II	Any pT/TX	N1-3	M0	SX
IIA	Any pT/TX	N1	M0	S0
	Any pT/TX	N1	M0	S1
IIB	Any pT/TX	N2	M0	S0
	Any pT/TX	N2	M0	S1
IIC	Any pT/TX	N3	M0	S0
	Any pT/TX	N3	M0	S1
III	Any pT/TX	Any N	M1	SX
IIIA	Any pT/TX	Any N	M1a	S0
	Any pT/TX	Any N	M1a	S1
IIIB	Any pT/TX	N1-3	M0	S2
	Any pT/TX	Any N	M1a	S2
IIIC	Any pT/TX	N1-3	M0	S3
	Any pT/TX	Any N	M1a	S3
	Any pT/TX	Any N	M1b	Any S

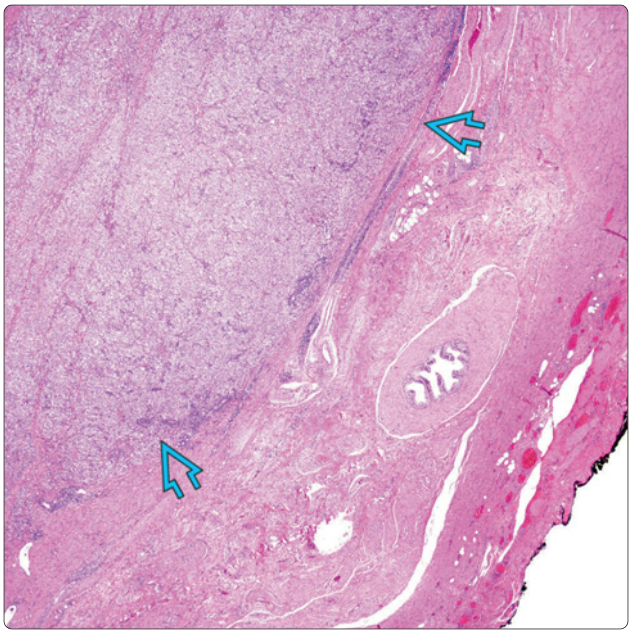
Adapted from 7th edition AJCC Staging Forms (2010).

pTis (Intratubular Germ Cell Neoplasia)



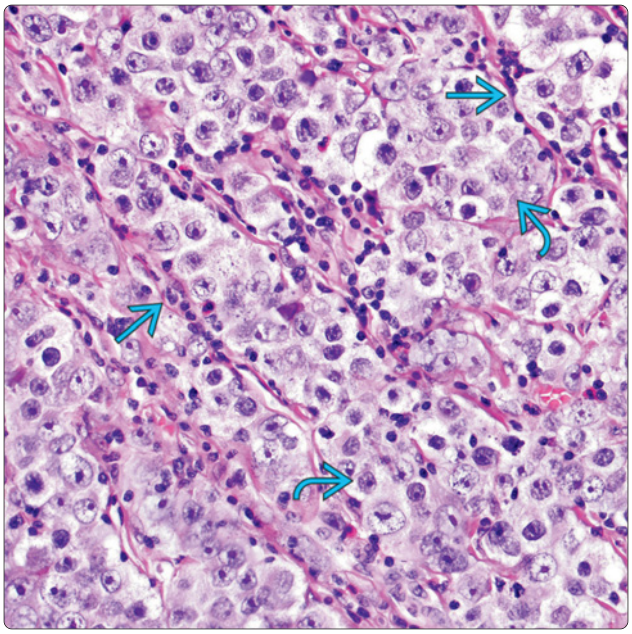
High-power magnification of an H&E stain shows a seminiferous tubule with neoplastic intratubular germ cells. The neoplastic cells are large, pleomorphic with abundant vacuolated cytoplasm and arranged in a single layer along the basement membrane.

pT1



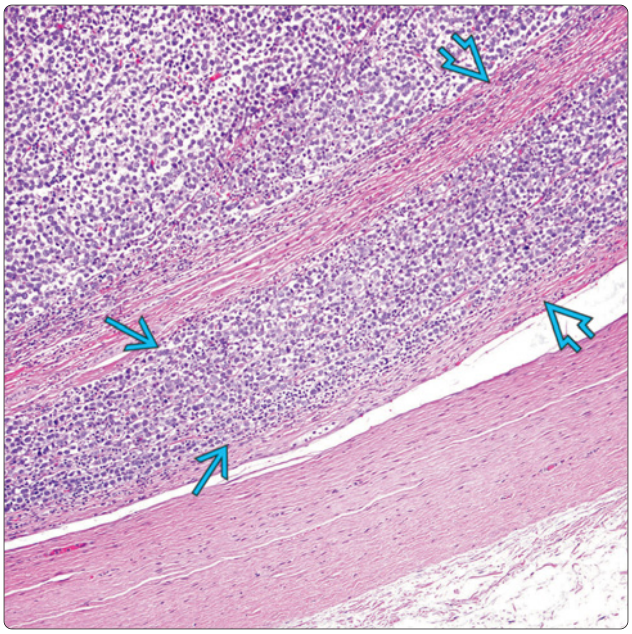
Low-power magnification of H&E stain shows sheets of seminoma that is replacing the testicular tissue but is limited to the testis without vascular or lymphatic invasion.

pT1



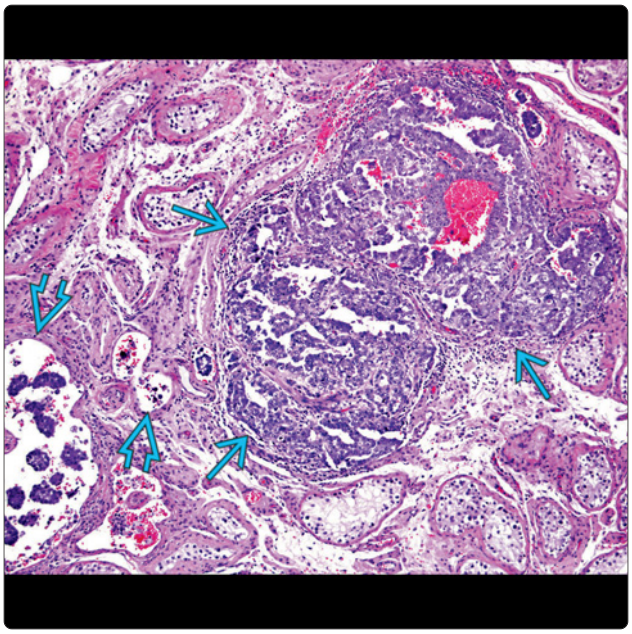
High-power magnification of an H&E stain of seminoma shows diffuse sheets of tumor cells with intervening branching fibrous septa. The neoplastic cells are polyhedral and have pale to clear cytoplasm with round to oval nuclei and 1-2 prominent nuclei.

pT1



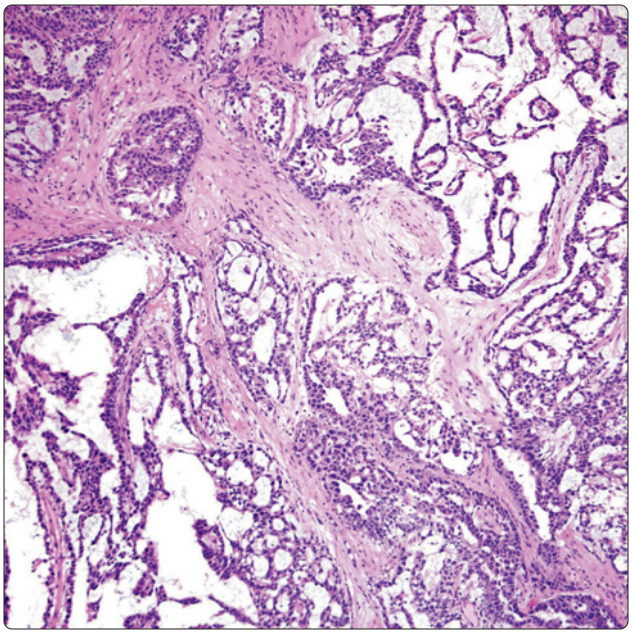
Intermediate-power magnification of H&E stain shows seminoma tumor cells that focally infiltrate into the tunica albuginea without involvement of the tunica vaginalis.

pT2



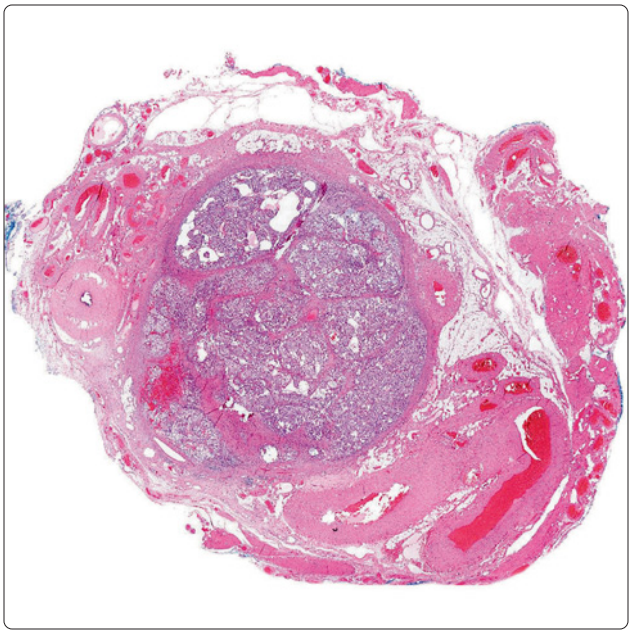
Intermediate-power magnification of H&E stain shows embryonal carcinoma component of germ cell tumor ⇨ infiltrating the seminiferous tubules with intravascular invasion in lymphatic/blood vessel spaces ⇨.

pT2



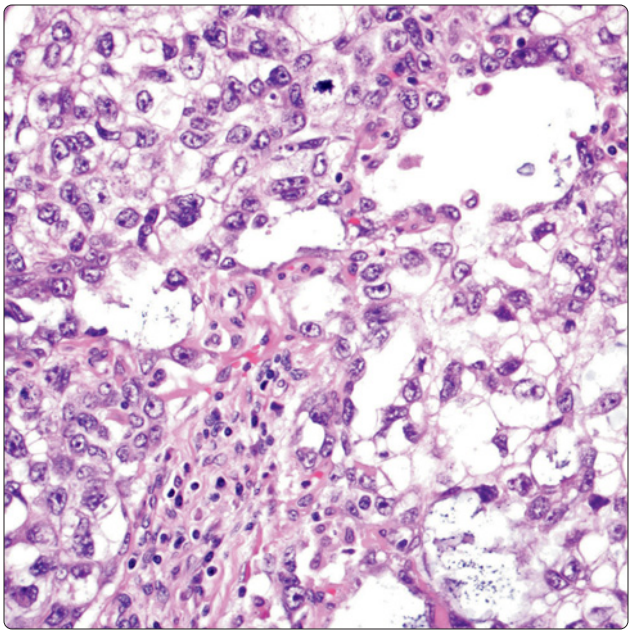
In an H&E-stained specimen from the same patient, other areas of the mixed germ cell tumor show yolk sac component.

pT3



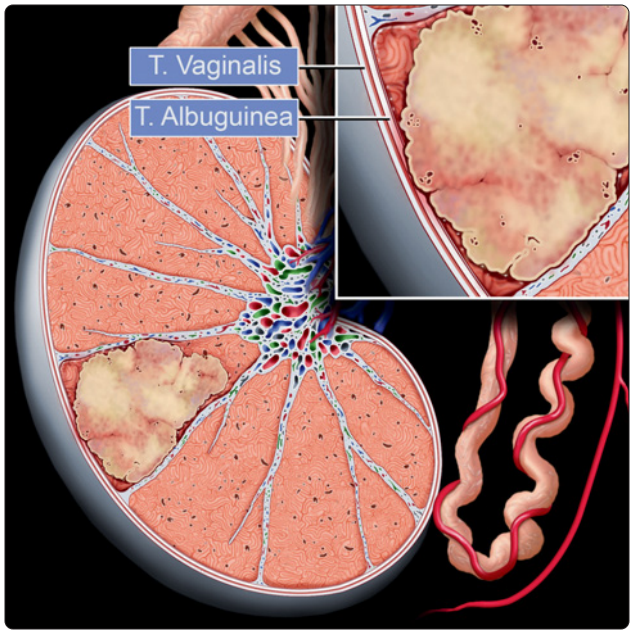
H&E stain of a cross section from a spermatic cord shows spermatic cord involvement by a testicular tumor.

pT3



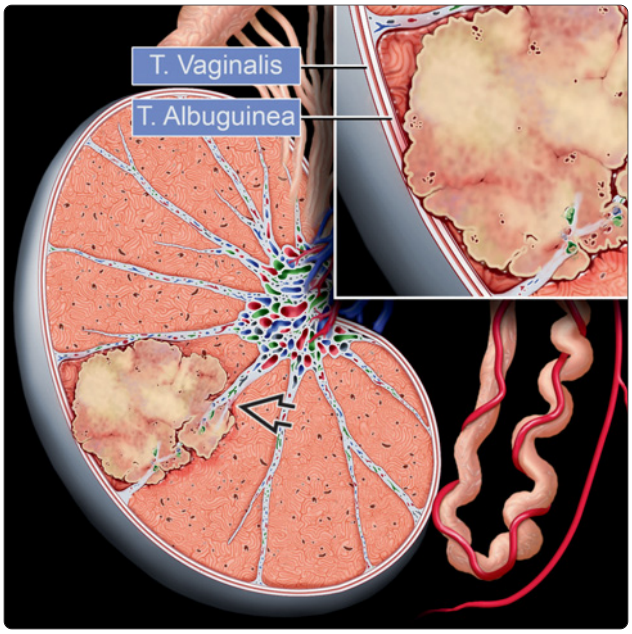
High-power magnification of the H&E stained section shows the yolk sac tumor, which expands the spermatic cord with a microcystic (most common) pattern giving a reticular or lace-like appearance.

T1



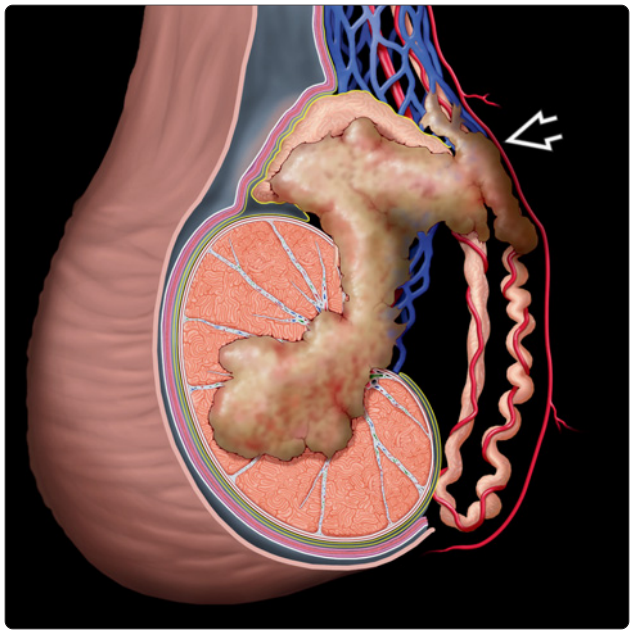
Graphic illustrates tumor limited to the testis and epididymis without vascular/lymphatic invasion. The tumor may invade into the tunical albuginea but not the tunica vaginalis (as seen in the inset). Both are classified as T1 disease.

T2



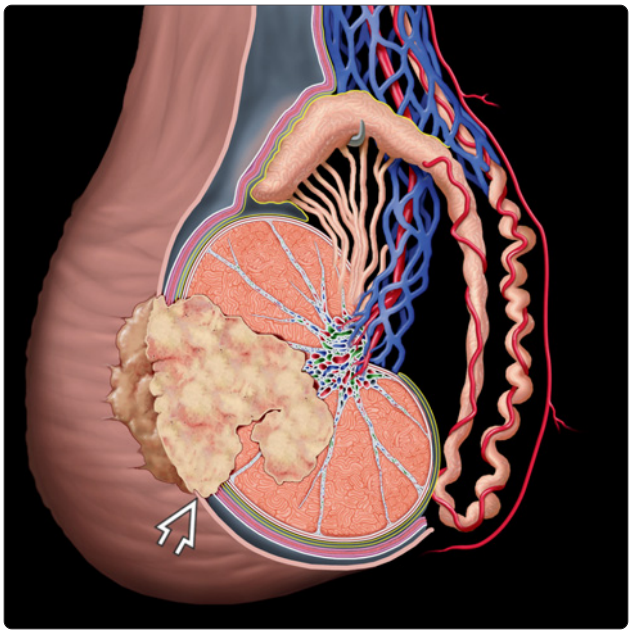
Graphic illustrates tumor limited to the testis and epididymis with vascular/lymphatic invasion ⇨. Tumor extending through tunica albuginea and involving tunica vaginalis (as seen in the inset) is also classified as T2 disease.

T3



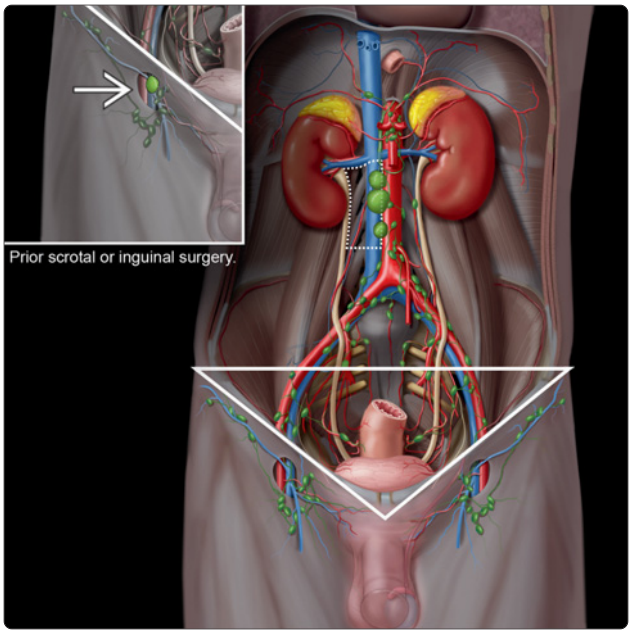
Graphic illustrates tumor invading the spermatic cord ⇨ (± vascular/lymphatic invasion), compatible with T3 disease.

T4



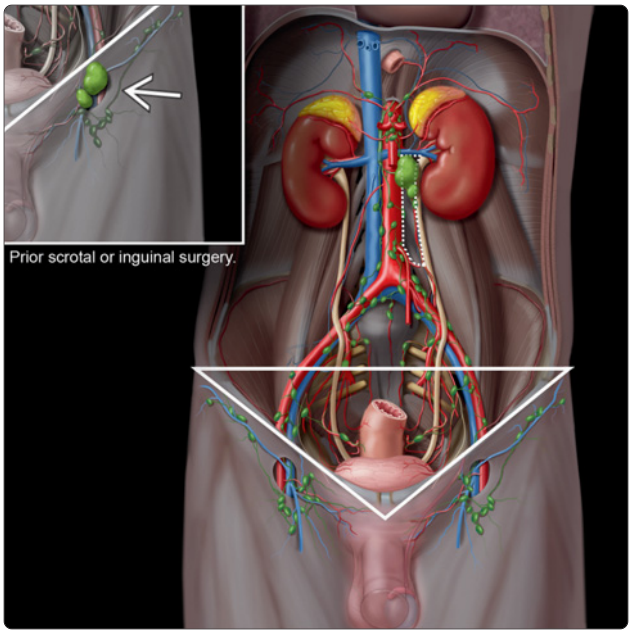
Graphic illustrates tumor invading the scrotum ⇨ (± vascular/lymphatic invasion), which is considered T4 disease.

N Staging



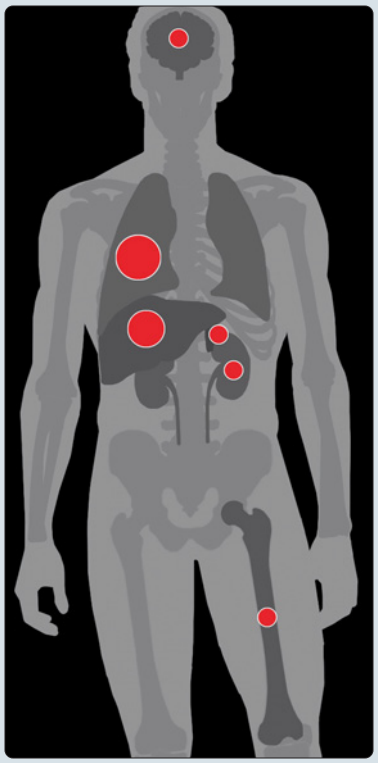
Right testicular nodal spread initially involves nodes around the IVC, including retroperitoneal, aortocaval, and paracaval nodes (dotted white line). If there has been disruption of lymphatics from prior scrotal or inguinal surgery or tumor invasion of scrotum, then spread is to inguinal nodes ➔.

N Staging



Left testicular spread involves retroperitoneal area bounded by renal vein, aorta, ureter, and IVC (dotted line). If lymphatics are disrupted by prior scrotal or inguinal surgery or tumor invasion of scrotum, then spread is to inguinal nodes ➔. Size of nodes determines N stage: < 2 cm = N1, 2-5 cm = N2, > 5 cm = N3.

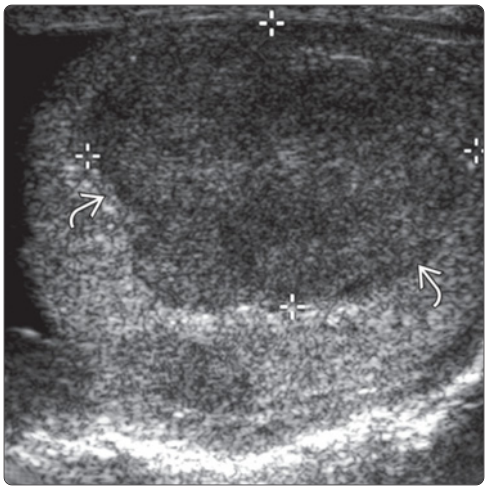
Metastases, Organ Frequency



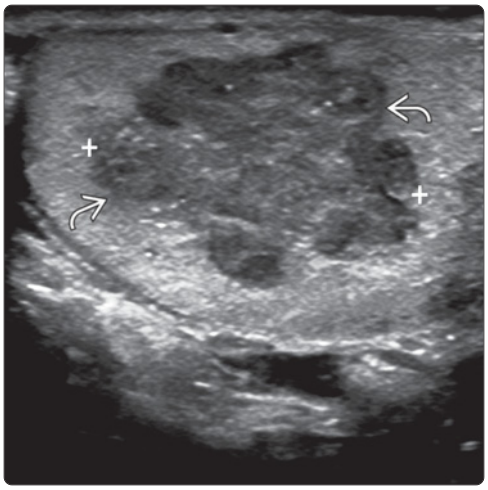
Lung	*89%*
Liver	*73%*
Brain	*31%*
Bone	*30%*
Kidney	*30%*
Adrenal gland	*29%*

Data from Bredael JJ et al: Autopsy findings in 154 patients with germ cell tumors of the testis. Cancer. 50(3):548-51, 1982.

Stage I (T1 N0 M0)

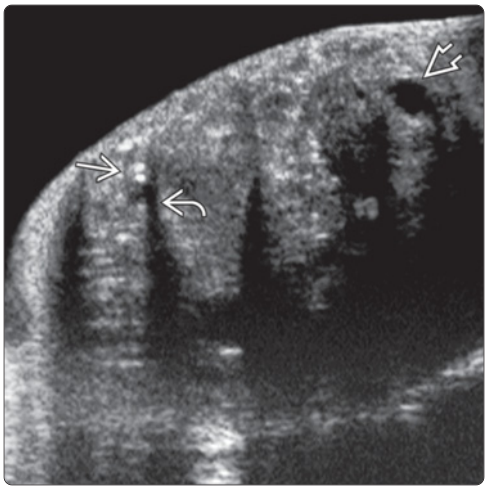


Stage I (T1 N0 M0)

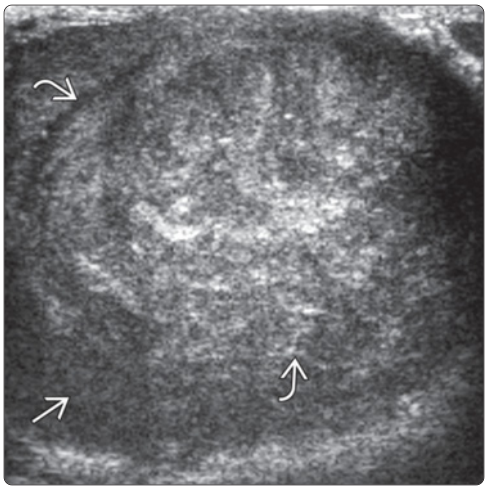


(Left) *Longitudinal grayscale ultrasound shows circumscribed, homogeneously hypoechoic mass found to be seminoma. This is a characteristic ultrasound appearance of seminoma. Ultrasound is not sensitive for evaluating tunica vaginalis, spermatic cord, or scrotal invasion.* **(Right)** *Longitudinal grayscale ultrasound shows lobulated, heterogeneous, predominately hypoechoic mass. The sonographic appearance is nonspecific and proved to be embryonal cell carcinoma.*

Stage I (T1 N0 M0)

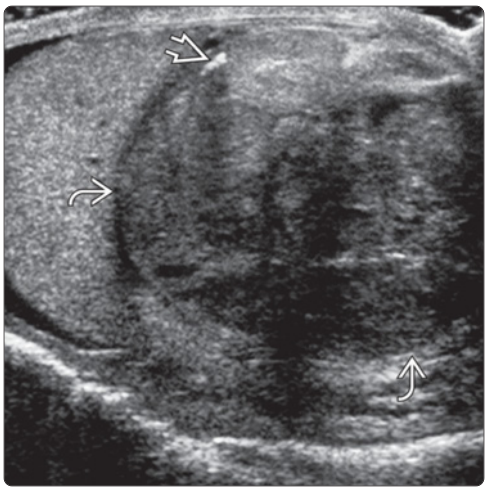


Stage I (T1 N0 M0)

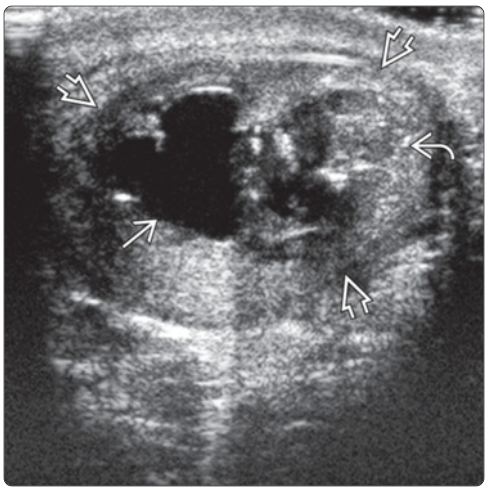


(Left) *Longitudinal grayscale ultrasound demonstrates heterogeneous, poorly defined mass. Mass contains foci of calcification with shadowing and cystic areas. Pathology revealed mature teratoma without invasion of the tunica or spermatic cord.* **(Right)** *Longitudinal grayscale ultrasound shows a poorly circumscribed heterogeneous testicular mass. Pathology revealed a yolk sac tumor. It can be difficult to differentiate tumor from normal testicle.*

Stage I (T1 N0 M0)

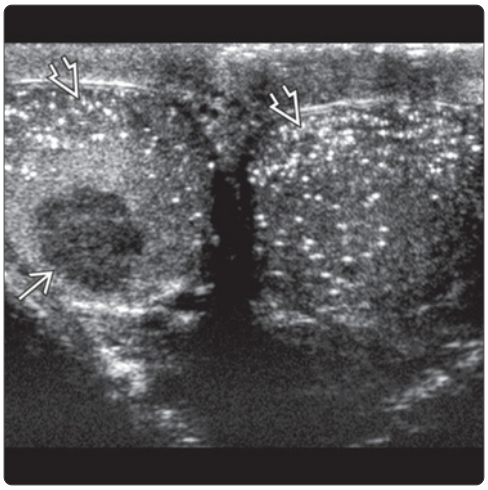


Stage I (T1 N0 M0)

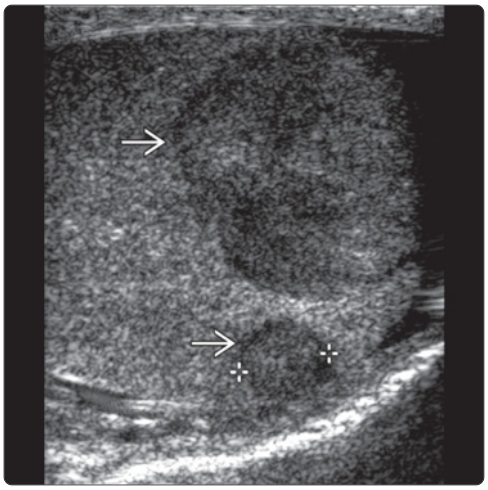


(Left) *Longitudinal grayscale ultrasound shows circumscribed mass with foci of calcification. This mass proved to be Sertoli cell tumor, although imaging findings are nonspecific.* **(Right)** *Longitudinal grayscale ultrasound demonstrates a heterogeneous mass with solid and cystic components. Pathology revealed mixed germ cell tumor with seminoma and embryonal cell components that developed in this patient with cryptorchidism.*

Stage I (T1 N0 M0)

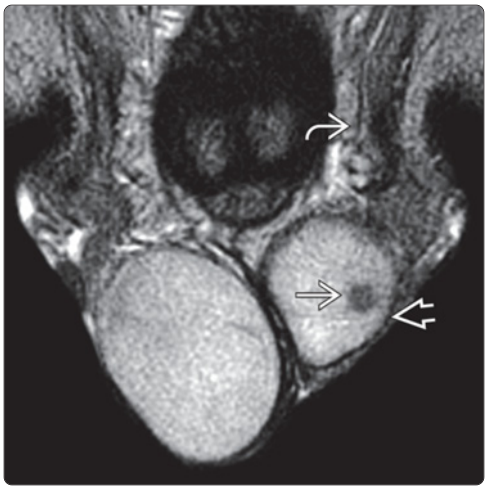


Stage I (T1 N0 M0)



(Left) *Longitudinal grayscale ultrasound shows a circumscribed, homogeneous, hypoechoic right testicular mass in a patient with bilateral testicular microlithiasis. Mass proved to be seminoma at orchiectomy.* **(Right)** *Longitudinal grayscale ultrasound shows 2 homogeneous, hypoechoic, circumscribed masses found to be multifocal seminoma.*

Stage I (T1 N0 M0)



Stage I (T1 N0 M0)

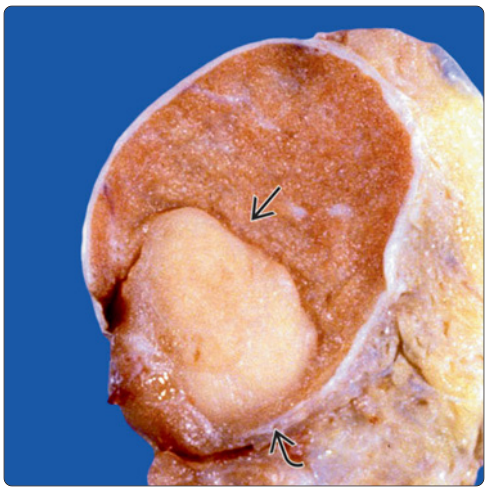


(Left) *Coronal T2WI MR shows a circumscribed, T2 homogeneously hypointense mass in the left testicle. MR is best imaging modality to evaluate for tunica and spermatic cord involvement, neither of which were involved in this case. Pathology revealed seminoma.* **(Right)** *Axial T1WI C+ MR in the same patient shows homogeneous enhancement in left testicular mass with sparing of the low signal tunica.*

Stage I (T1 N0 M0)

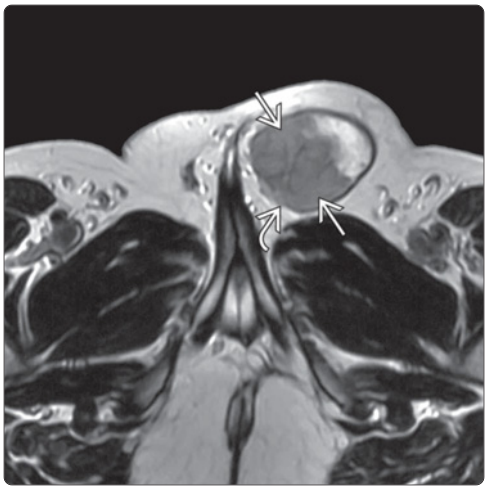


Stage I (T1 N0 M0)

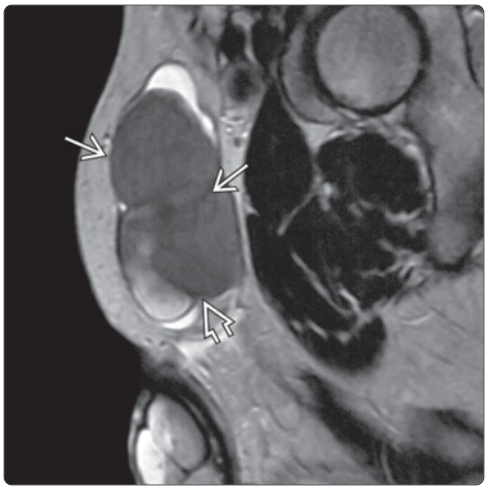


(Left) *Axial T2WI MR shows a circumscribed, mildly heterogeneous, hypointense mass in left testicle. Tunica vaginalis is not invaded.* **(Right)** *Gross pathology in sagittal plane from the same patient demonstrates a testicular mass, which was pure seminoma. Tunica vaginalis is intact, corresponding to imaging findings.*

Stage IB (T2 N0 M0)

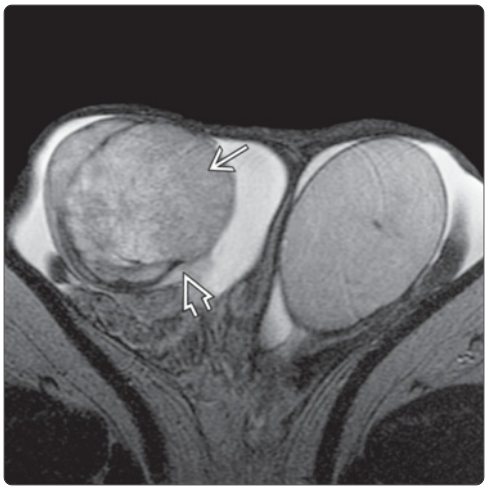


Stage IB (T2 N0 M0)

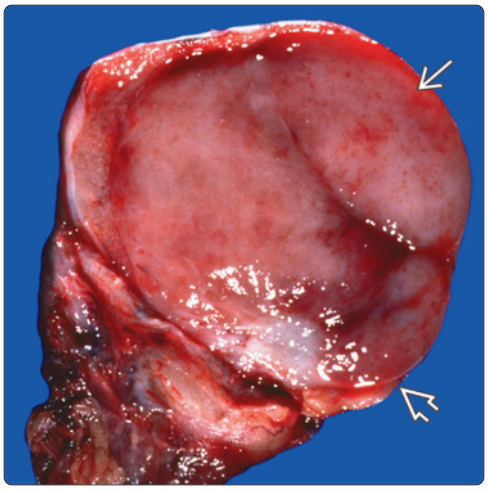


(Left) *Axial T2WI MR shows a heterogeneous hypointense mass replacing the majority of normal testicular tissue of this left side undescended testicle. Tunica vaginalis has been invaded.* **(Right)** *Sagittal T2WI MR in the same patient shows a large hypointense mass replacing the majority of normal testicular tissue in this undescended testicle. Disruption of tunica vaginalis is again demonstrated.*

Stage IB (T2 N0 M0)

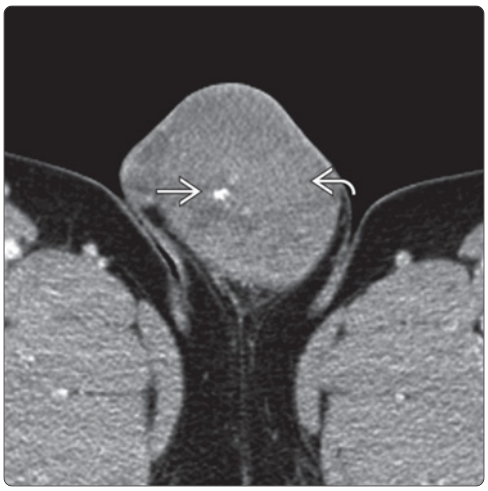


Stage IB (T2 N0 M0)

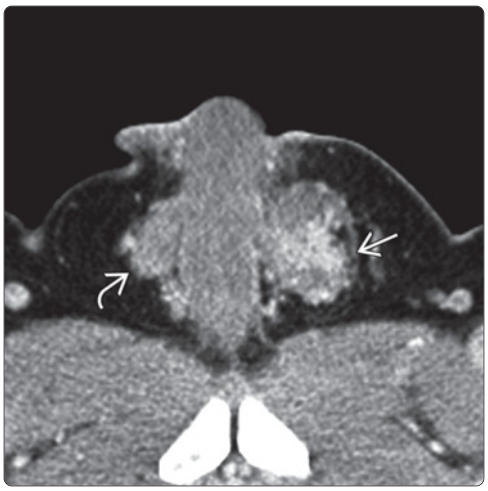


(Left) *Axial T2WI MR shows a large, heterogeneous, predominately isointense mass replacing the majority of the normal right testicle. Tunica vaginalis is invaded.* **(Right)** *Gross pathology from the same patient correlates to imaging findings showing mass replacing the majority of testicular tissue as well as invasion through tunica vaginalis.*

Stage IB (T3 N0 M0)

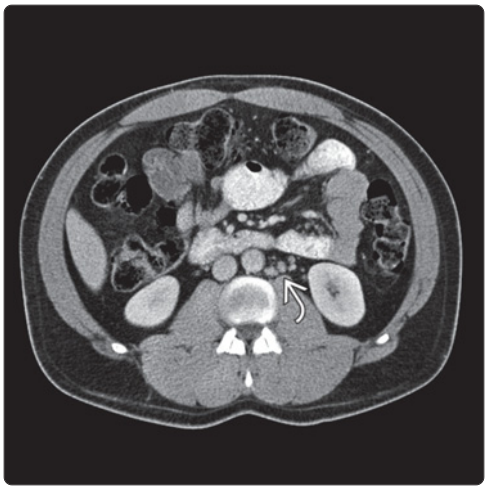


Stage IB (T3 N0 M0)

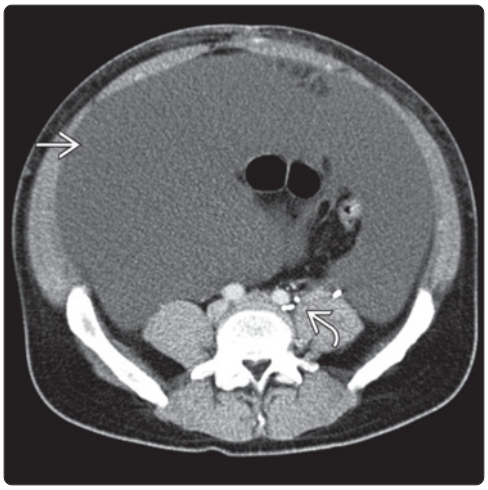


(Left) *Axial CECT shows a heterogeneous left testicular mass with foci of calcification. Pathology demonstrated invasion of tunica vaginalis.* **(Right)** *Axial CECT in the same patient shows tumor involvement in the left spermatic cord. Compare the appearance to that of the normal right spermatic cord.*

Stage IIA (T3 N1 M0)



Stage IIA (T3 N1 M0)

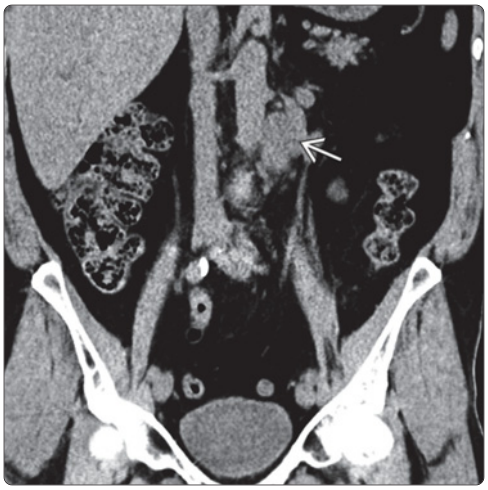


(Left) *Axial CECT in a patient with a mixed germ cell tumor shows left-sided paraaortic adenopathy. Nodes measure less than 2 cm in size.* **(Right)** *Axial CECT in the same patient status post orchiectomy and retroperitoneal lymph node resection reveals chylous ascites, which developed post surgery.*

Stage IIB (T2 N2 M0)

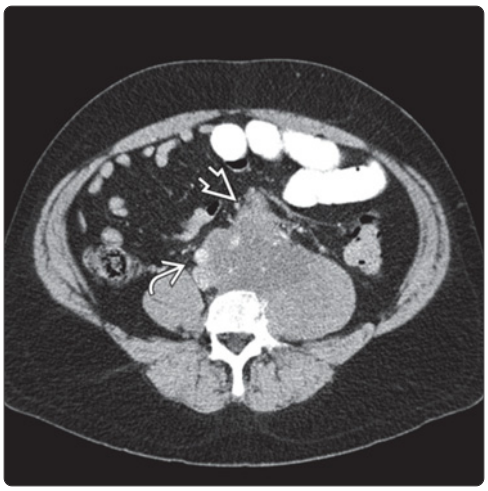


Stage IIB (T2 N2 M0)

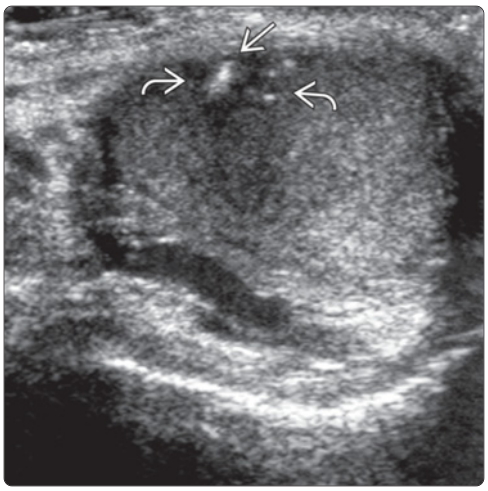


(Left) *Axial NECT in a patient with left side seminoma shows left paraaortic lymph node with hazy border, concerning for extranodal extension. Pathology post orchiectomy demonstrated vascular and lymphatic invasion, making this testicular mass T2 disease.* **(Right)** *Coronal NECT in the same patient demonstrates left paraaortic lymph node conglomeration measuring up to 4 cm, making this N2 disease.*

Stage IIC (T1 N3 M0)



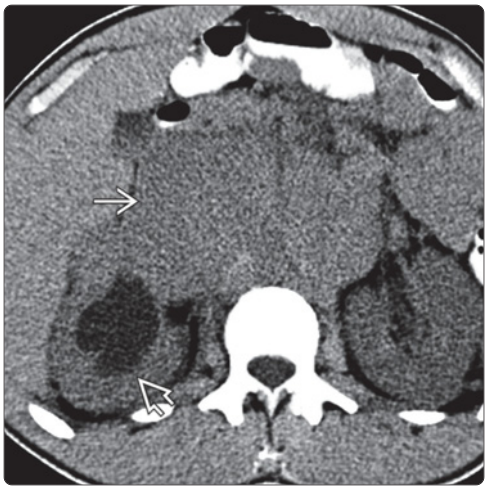
Stage IIC (T1 N3 M0)



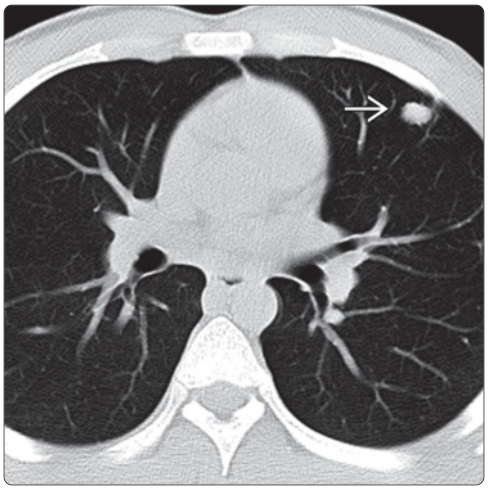
(Left) *Axial CECT shows a retroperitoneal mass with displacement of infrarenal abdominal aorta. Biopsy revealed poorly differentiated carcinoma. Tumor staining indicated positive germ cell markers. Retroperitoneal mass is greater than 5 cm, making this N3 stage.* **(Right)** *Subsequent ultrasound in the same patient shows foci of calcification within a small hypoechoic testicular mass due to a "burned out" germ cell tumor.*

(Left) *Axial NECT in a patient with testicular seminoma shows a large retroperitoneal mass. Right hydronephrosis is related to ureteric obstruction by the nodal mass.* **(Right)** *Axial NECT in the same patient shows pulmonary parenchymal metastatic lesion in the chest, making this M1a stage.*

Stage IIIB (T2 N3 M1a)

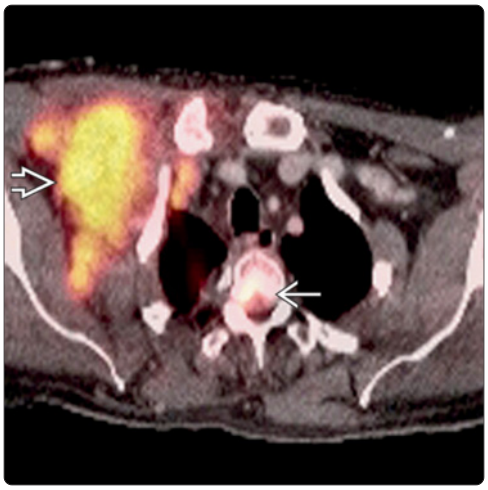


Stage IIIB (T2 N3 M1a)



(Left) *Axial fused PET/CT demonstrates FDG-avid supraclavicular adenopathy and FDG-avid vertebral body lesion in a patient with testicular seminoma.* **(Right)** *Coronal PET/CT in the same patient shows supraclavicular adenopathy and left lower lobe lung lesion.*

Stage IIIC (T2 N3 M1b)

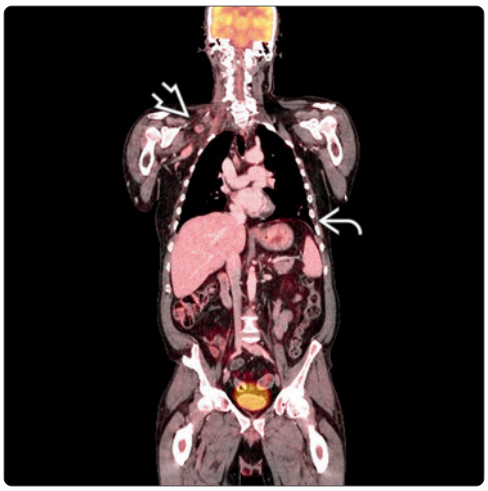


Stage IIIC (T2 N3 M1b)

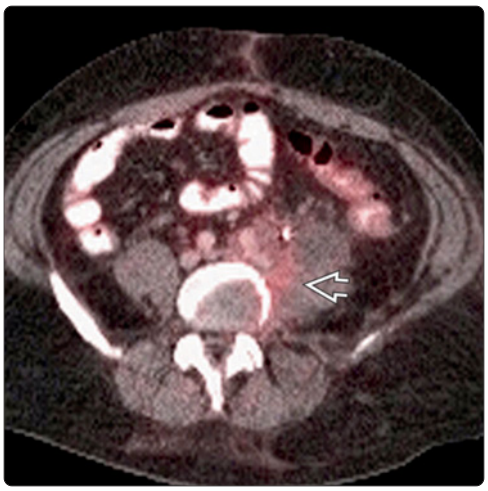


(Left) *Coronal PET/CT for restaging in the same patient after treatment shows significant decrease in supraclavicular adenopathy and resolution of lung nodule.* **(Right)** *Axial fused PET/CT for restaging in a patient with "burned out" germ cell tumor post tumor resection and chemotherapy demonstrates residual retroperitoneal mass with FDG uptake. This is consistent with residual viable tumor.*

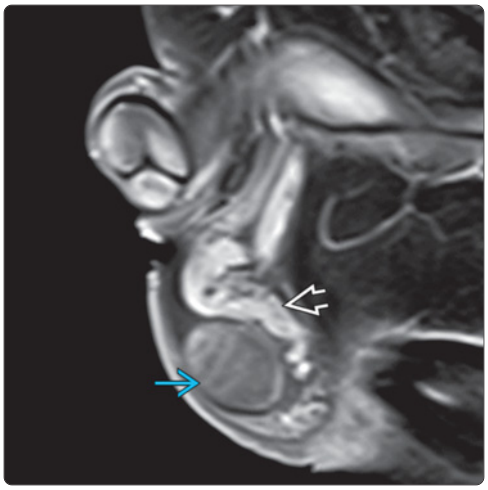
Stage IIIC (T2 N3 M1b)



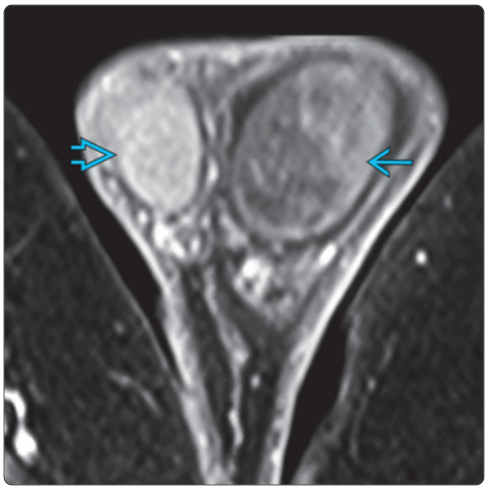
Stage IIC (T1 N3 M0)



Stage IIIC (T1 N3 M1b)



Stage IIIC (T1 N3 M1b)



(Left) *Sagittal T1WI C+ MR shows mild irregular heterogeneous enhancement throughout an enlarged left testicle. No focal mass lesion is identified. Left spermatic cord is spared.* **(Right)** *Axial T1WI C+ MR in the same patient shows an enlarged, heterogeneous, enhancing left testicle compared to a normal right testicle.*

Stage IIIC (T1 N3 M1b)

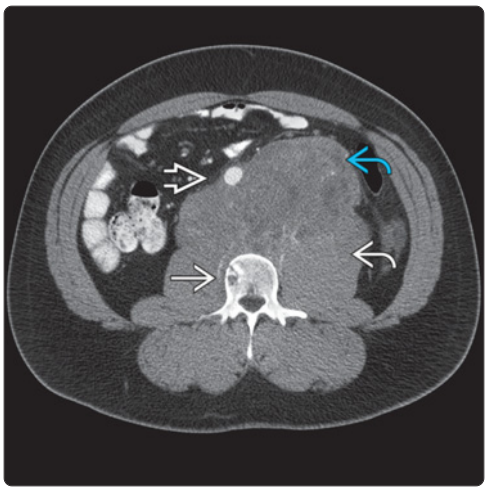


Stage IIIC (T1 N3 M1b)

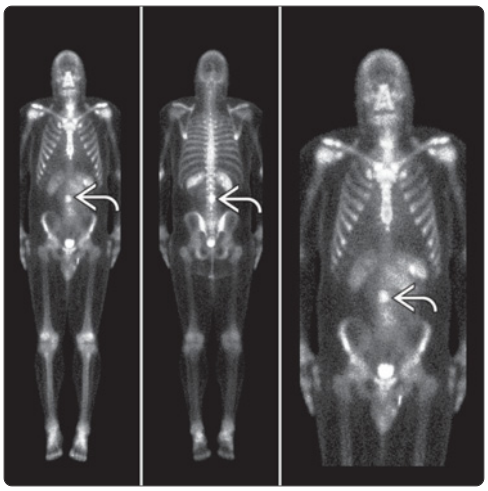


(Left) *Axial T1WI in phase MR in the same patient shows heterogeneous retroperitoneal mass. Left psoas muscle was invaded.* **(Right)** *Axial T1WI opposed-phase MR in the same patient shows a heterogeneous retroperitoneal mass. Signal drop on the opposed-phase image in comparison to the in-phase MR sequence corresponds with fat elements in the mass.*

Stage IIIC (T1 N3 M1b)



Stage IIIC (T1 N3 M1b)



(Left) *Axial CECT in the same patient demonstrates a large heterogeneous retroperitoneal mass displacing aorta and invading left psoas muscle. Lytic lesion is seen in L3 vertebral body.* **(Right)** *Coronal bone scan in the same patient demonstrates uptake in L3 vertebral lytic lesion, consistent with metastatic disease.*

Stromal Tumors

KEY FACTS

TERMINOLOGY

- Gonadal stromal tumors arise from non-germ cell elements

IMAGING

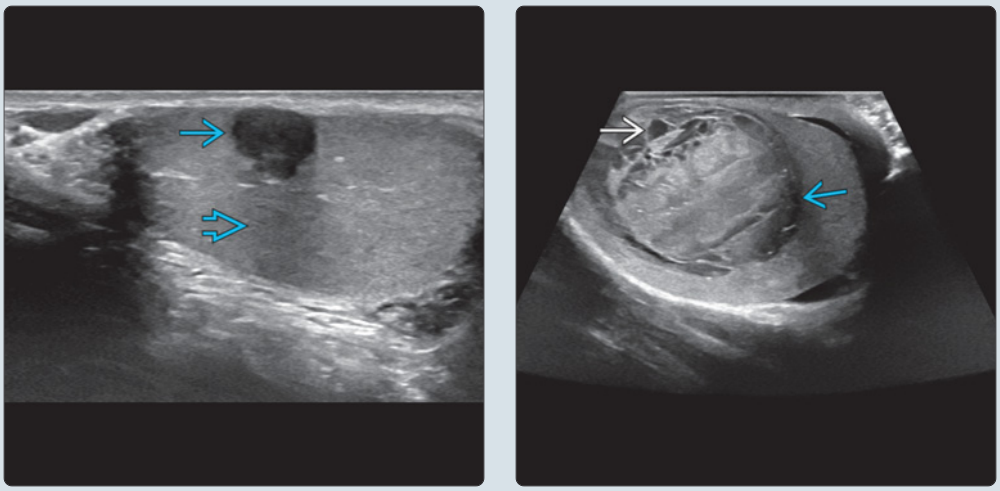
- May be indistinguishable from germ cell tumors on grayscale ultrasound
  - Leydig cell tumors: Small, solid, hypoechoic intratesticular mass
  - Sertoli cell tumors: Small, hypoechoic mass with occasional hemorrhage, which may lead to heterogeneity and cystic components
  - Gonadoblastoma: Stromal tumor in conjunction with germ cell tumor, usually mixed sonographic features
- High-resolution US (≥ 7.5 MHz) is best imaging tool for detection of gonadal stromal neoplasms

CLINICAL ISSUES

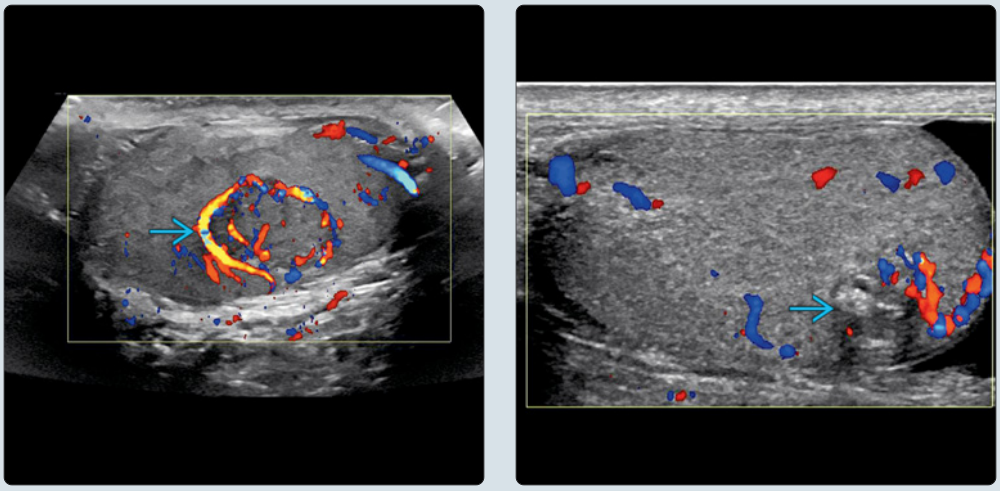
- 30% of patients with gonadal stromal tumors have endocrinopathy secondary to testosterone or estrogen production by tumor
  - Children: Precocious puberty and gynecomastia
  - Adults: Impotence, ↓ libido
- Majority of these tumors are benign
- Orchidectomy is preferred treatment

DIAGNOSTIC CHECKLIST

- Consider stromal tumor in any patient with endocrinopathy and testicular mass



(Left) *Sagittal grayscale ultrasound of the right testis in a 16-year-old male demonstrates a well-defined hypoechoic mass ⇨ with mild posterior acoustic shadowing ⇨. Pathology after orchiectomy confirmed it to be a benign sex cord-stromal tumor (unclassified type).* **(Right)** *Sagittal grayscale ultrasound in a 16-year-old male demonstrates a well-defined, predominantly solid mass ⇨ with interspersed cystic areas ➡. Pathology confirmed it to be a granulosa cell tumor (adult type).*



(Left) *Sagittal color Doppler ultrasound of the right testis in this 33-year-old male demonstrates a well-defined, hypoechoic solid mass with marked internal vascularity ⇨. Pathology confirmed it to be a Leydig cell tumor.* **(Right)** *Sagittal color Doppler ultrasound shows the right testis with a partially calcified mass ⇨ and minimal to no internal vascularity. Pathology confirmed a Sertoli cell tumor.*

TERMINOLOGY

Abbreviations

- Gonadal stromal tumors (GST): Also called non-germ cell tumors, interstitial cell tumors, or sex cord tumors

Definitions

- Leydig cell tumor (LCT): Arises from interstitial cells
- Sertoli cell tumor (SCT): Arises from sustentacular cells lining seminiferous tubules
- Granulosa cell tumor (GCT): Rare sex cord tumor
- Gonadoblastoma: Contain both stromal and germ cell elements

IMAGING

General Features

- Location
  - Bilateral in 3%
- Size
  - Benign tumors: Usually < 3 cm; malignant tumors: Usually > 5 cm
- Morphology
  - Well circumscribed, round/lobulated

MR Findings

- T2WI: Low signal intratesticular mass ± high signal fibrous capsule rim and high signal intensity foci internally secondary to central scars

Ultrasonographic Findings

- Grayscale ultrasound
  - May be indistinguishable from germ cell tumors
  - LCT: Small, solid, hypoechoic intratesticular mass
    - Larger tumors: Hemorrhage or necrosis leads to heterogeneous echo pattern
    - May occasionally show cystic change
  - SCT: Small, hypoechoic mass, hemorrhage may lead to heterogeneity
    - Solid and cystic components
    - ± punctate calcification; large calcified mass in large-cell calcifying Sertoli cell tumor
  - GCT: Well-defined hypoechoic mass with solid and cystic components
  - Adult testicular GCTs are rare sex cord-stromal tumors

Imaging Recommendations

- Best imaging tool
  - High-resolution US (≥ 7.5 MHz)

DIFFERENTIAL DIAGNOSIS

Testicular Germ Cell Tumors

- May be indistinguishable from stromal tumors on US

Testicular Metastases, Lymphoma, Leukemia

- Often multiple; otherwise indistinguishable

Intratesticular Hematoma

- Scrotal trauma, no internal color flow in hematoma

PATHOLOGY

General Features

- Associated abnormalities
  - Leydig cell tumor: Klinefelter syndrome
  - SCT: Peutz-Jeghers syndrome and Carney syndrome
- Leydig cell tumors: 3% of all testicular tumors
  - 90% benign, 3% bilateral
  - Most common, may produce testosterone
- Sertoli cell tumors (SCT): 1% of all testis tumors
  - 85-90% benign
  - May produce estrogen/müllerian inhibiting factor
- Granulosa cell tumor: Rare
  - Juvenile GCTs are typically benign; 20% of testicular adult GCTs have been reported to be malignant

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless testicular enlargement
  - 30% of patients have endocrinopathy secondary to testosterone or estrogen production by tumor
    - Gynecomastia, impotence, ↓ libido in adults

Demographics

- Age
  - LCT: 30-60 years; 25% occur before puberty
  - SCT: All age groups; 1/3 < 12 years
  - GCT: Juvenile type in infants; adult type in men aged 16-77 years

Natural History & Prognosis

- Malignant tumors metastasize in same pattern as testicular germ cell tumors

Treatment

- Orchidectomy or testis-sparing surgery

DIAGNOSTIC CHECKLIST

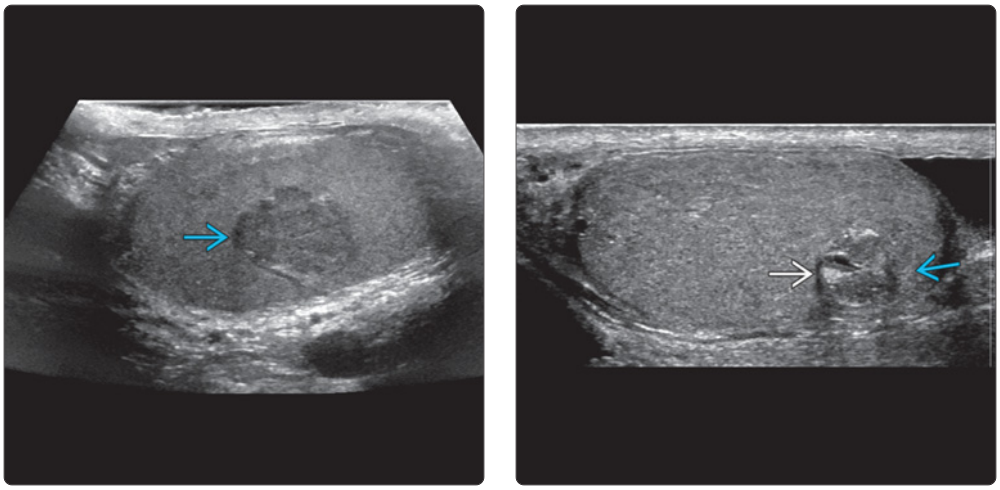
Image Interpretation Pearls

- Consider stromal tumor in any patient with endocrinopathy and testicular mass

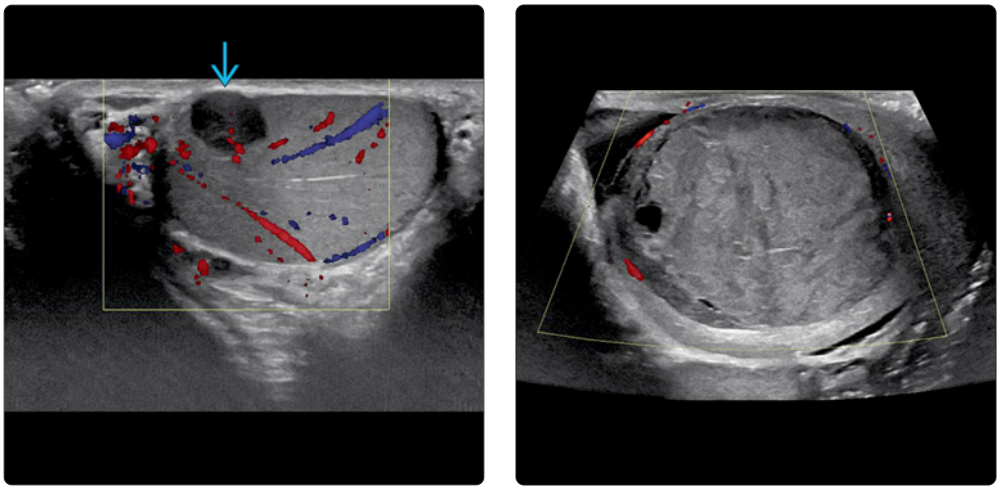
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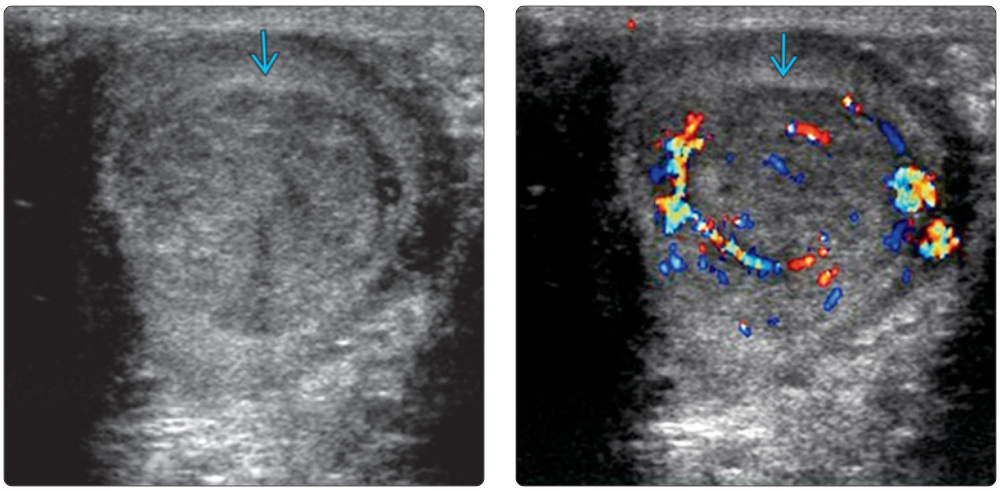
(Left) *Sagittal grayscale ultrasound of the right testis in this 33-year-old male demonstrates a well-defined, hypoechoic solid mass. Pathology confirmed a Leydig cell tumor.* **(Right)** *Sagittal grayscale ultrasound of the right testis in a young male demonstrates a solid mass with scant cystic components and echogenic regions with calcification. Pathology confirmed a Sertoli cell tumor.*

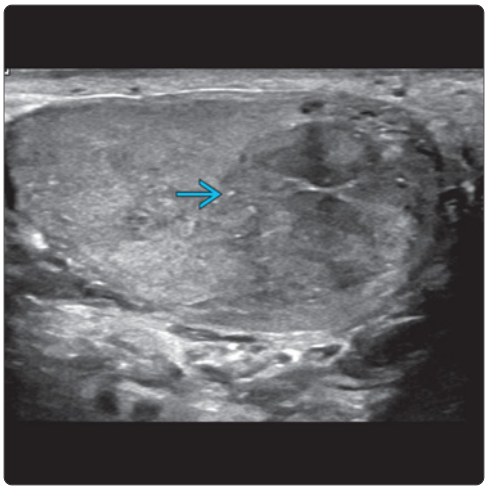


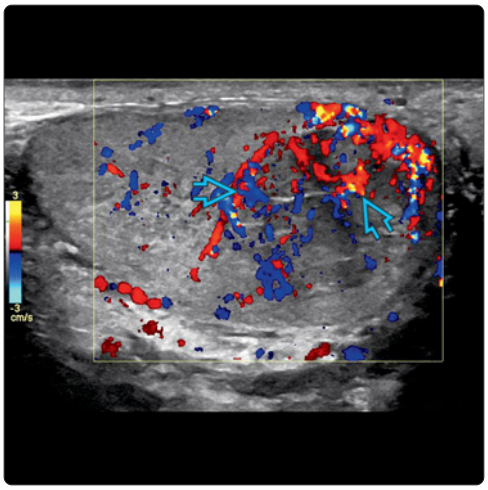
(Left) *Transverse color Doppler ultrasound of the right testis in this 16-year-old male demonstrates a well-defined, hypoechoic solid mass with minimal internal vascularity. Pathology confirmed a benign sex cord-stromal tumor (unclassified type).* **(Right)** *Sagittal color Doppler ultrasound of a hypoechoic testicular mass shows no internal vascularity. Pathology confirmed it to be an adult granulosa cell tumor.*



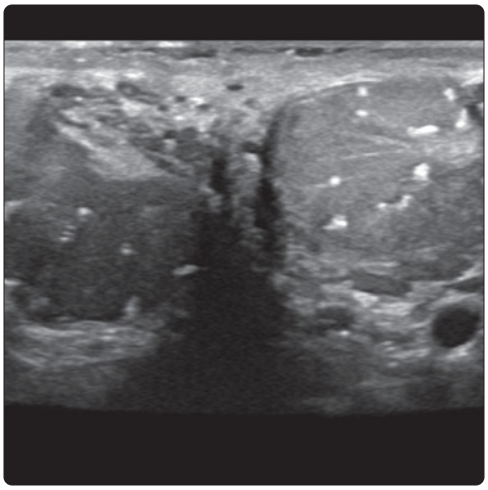
(Left) *Transverse grayscale ultrasound of the testis in a 12-year-old male demonstrates a well-defined, heterogeneous, hypoechoic solid mass.* **(Right)** *Corresponding transverse color Doppler ultrasound of the testis demonstrates internal vascularity in this well-defined, heterogenous, hypoechoic solid mass. Pathology confirmed a Leydig cell tumor.*

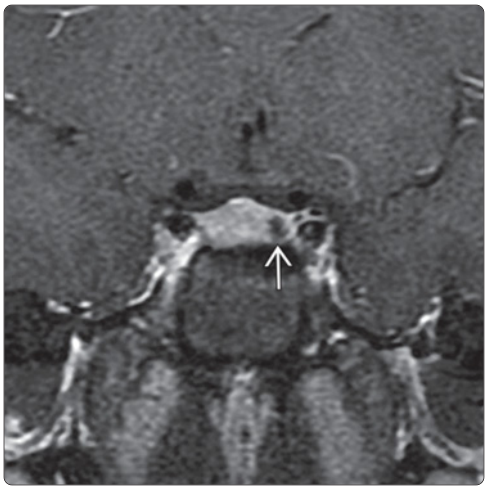




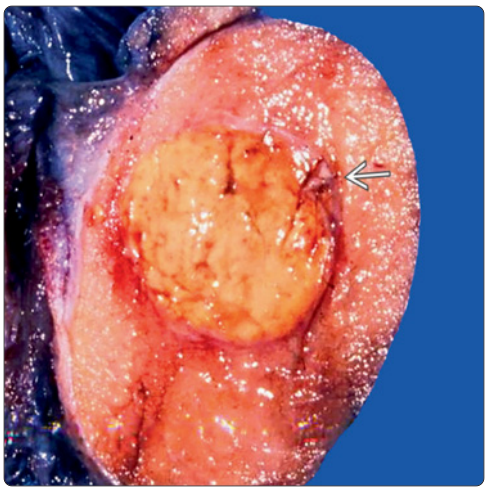


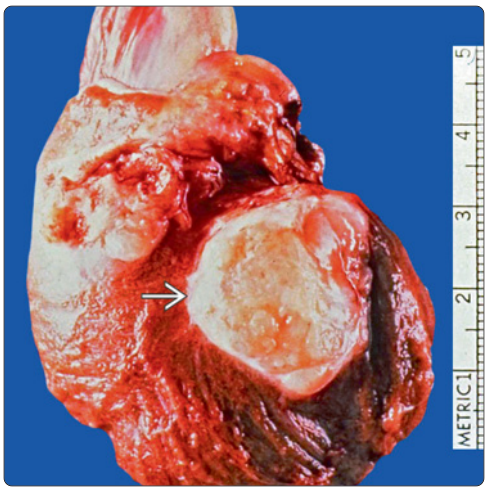
(Left) *Sagittal grayscale ultrasound of the left testis in this 18-year-old male demonstrates a hypoechoic solid mass ⇨. (Right) Corresponding color Doppler ultrasound of the left testis in the same patient shows marked internal vascularity ⇨. Pathology confirmed a Leydig cell tumor.*





(Left) *Surveillance ultrasound performed on a 20-year-old male with a history of Carney complex shows innumerable echogenic foci throughout both testes. The appearance was stable over many years. Though exceedingly rare, the patient's clinical history and stability at surveillance imaging indicate large-cell calcifying Sertoli cell tumors.* **(Right)** *T1 C+ MR in the same patient shows a pituitary microadenoma ➡. Another tumor associated with the Carney complex, an atrial myxoma, was resected previously.*





(Left) *Leydig cell tumor (LCT) is often a well-circumscribed mass with a homogeneous yellow-tan cut surface. Focal cystic change is present ➡. Hemorrhage or necrosis is lacking. LCTs often do not replace the entire testis.* **(Right)** *A well-circumscribed Sertoli cell tumor with a tan-white firm cut surface ➡ is shown. The gross finding is different from Leydig cell tumor, which is usually tan-brown to yellow due to high lipid content.*

Testicular Lymphoma and Leukemia

KEY FACTS

TERMINOLOGY

- Infiltrative neoplasm of testis in which tumor cells surround and compress seminiferous tubules and normal testicular vessels

IMAGING

- Bilateral, solid, hypoechoic, hypervascular nodules/masses
- Diffuse hypoechoic testis with hypervascularity
- Striated pattern
- Testicular shape not altered
- Normal testicular vessels with straight course crossing through lesions
- Comparison of both testes to look for asymmetry in sizes and echogenicity

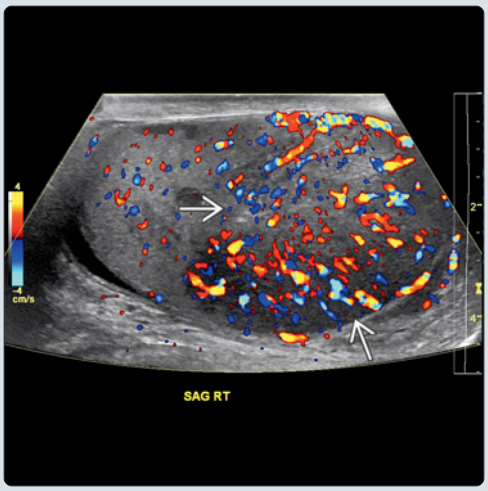
PATHOLOGY

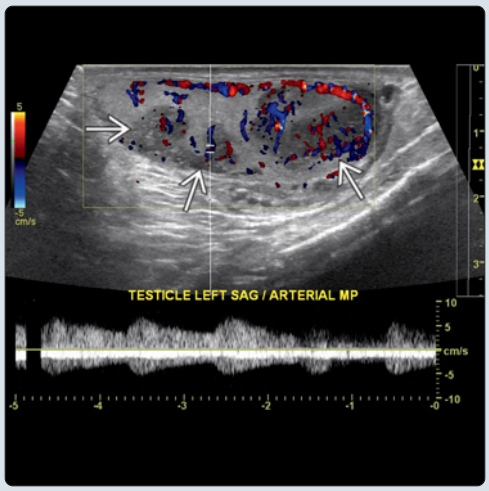
- Most commonly secondary lymphomatous involvement of testis; rarely primary
- Lymphoma behaves similar to leukemia with abnormal cells diffusely infiltrating interstitium with compression of seminiferous tubules without causing their destruction
- Testis is "sanctuary organ": Blood-gonad barrier limits accumulation of chemotherapeutic agents

CLINICAL ISSUES

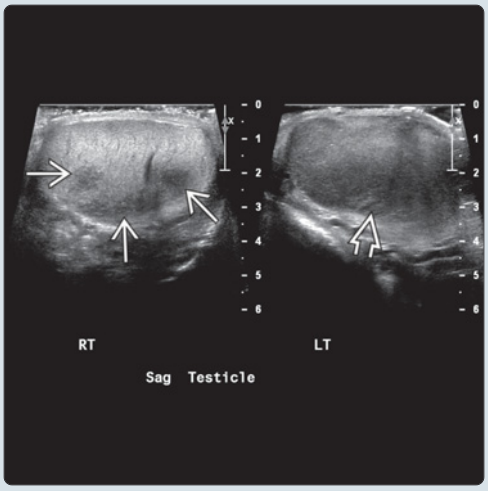
- Stages IE and IIE: Orchidectomy
- Stage IIIE and IVE: Systemic chemotherapy using cyclophosphamide, doxorubicin, vincristine, and prednisolone
- Radiation in symptomatic and bulky deposits
- Lymphoma accounts for ~ 5% of all testicular tumors
- Most common bilateral testicular tumor

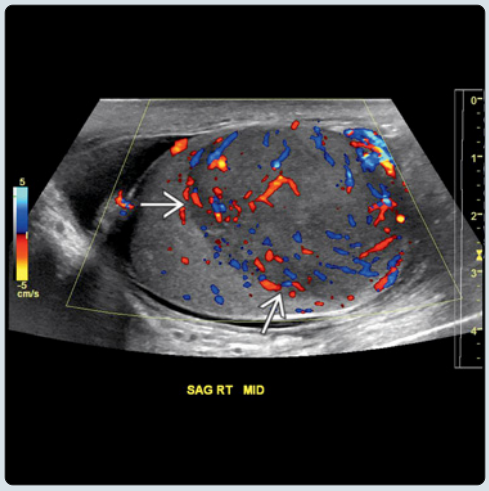
(Left) *Sagittal color Doppler ultrasound of the right testis in a 56-year-old man with non-Hodgkin lymphoma shows a hypervascular focal mass ➡ in the inferior pole with focal enlargement of testis. Note that the shape of the testis is maintained. Pathology confirmed lymphoma.* **(Right)** *Sagittal color Doppler ultrasound of the left testis in a 65-year-old man shows multiple hypervascular hypoechoic masses ➡ in the testis. Pathology confirmed lymphoma.*





(Left) *Sagittal grayscale ultrasound of the testes in a 60-year-old man with non-Hodgkin lymphoma shows bilateral hypoechoic masses, multifocal in the right ➡ and diffusely infiltrating in the left ➡.* **(Right)** *Sagittal color Doppler ultrasound of the right testis in a 51-year-old man with known history of acute myeloid leukemia shows a hypervascular focal hypoechoic mass ➡ in the inferior pole. Pathology confirmed acute myeloid leukemia.*





TERMINOLOGY

Synonyms

- B-cell lymphoma of testis

Definitions

- Infiltrative neoplasm of testis in which tumor cells surround and compress seminiferous tubules and normal testicular vessels

IMAGING

General Features

- Best diagnostic clue
  - Bilateral, solid, hypoechoic, hypervascular nodules/masses
  - Diffuse hypoechoic testis: Relatively increased Doppler flow

Ultrasonographic Findings

- Grayscale ultrasound
  - Unifocal or multifocal hypoechoic lesions
  - Diffusely enlarged, hypoechoic testis
  - Striated pattern
  - Testicular shape not altered
- Color Doppler
  - Hypervascular lesions
  - Normal testicular vessels with straight course crossing through lesions

Imaging Recommendations

- Best imaging tool
  - Ultrasound
- Protocol advice
  - Comparison of both testes to look for asymmetry in size and echogenicity

Nuclear Medicine Findings

- Increased FDG uptake on PET

DIFFERENTIAL DIAGNOSIS

Epididymo-Orchitis

- Enlarged and heterogeneous testis and epididymis
- Hypervascular
- Associated findings: Complex hydrocele, skin thickening
- Clinical findings of acute scrotum at time of presentation

Primary Testicular Neoplasm

- Solid or mixed solid-cystic discrete mass; ± calcifications
- May have increased vascularity
- May have invasion of tunica albuginea (embryonal carcinoma)

Granulomatous Orchitis (Tuberculosis)

- Multiple hypoechoic nodules
- Testis and epididymis involved

PATHOLOGY

General Features

- Etiology
  - Most commonly secondary lymphomatous involvement of testis; rarely primary
  - Testis is a "sanctuary organ": Blood-gonad barrier limits accumulation of chemotherapeutic agents
    - Most commonly described in children with acute lymphoblastic leukemia
- Associated abnormalities
  - May have enlarged lymph nodes in other parts of body

Gross Pathologic & Surgical Features

- Solid, tannish-white mass
- Lymphoma behaves similar to leukemia with abnormal cells diffusely infiltrating interstitium with compression of seminiferous tubules without causing their destruction

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless testicular mass/enlargement
- Other signs/symptoms
  - Weight loss, night sweats, fever, anorexia

Demographics

- Age
  - Lymphoma: 20-80 years
    - Most common malignant tumor of testis in elderly
  - Leukemia: Seen in childhood
- Epidemiology
  - Most common bilateral testicular tumor

Natural History & Prognosis

- Primary lymphoma: 5-year survival rate of 12%

Treatment

- Stages IE and IIE: Orchidectomy
- Stage IIIE and IVE: Systemic chemotherapy using cyclophosphamide, doxorubicin, vincristine, and prednisolone
- Radiation of symptomatic and bulky deposits
- CNS prophylaxis should be considered
- Lymphoma accounts for ~ 5% of all testicular tumors

DIAGNOSTIC CHECKLIST

Consider

- Primary testicular neoplasm
- Orchitis if pain and fever present

Image Interpretation Pearls

- Bilateral, uni- or multifocal, or infiltrative hypoechoic mass(es) with increased vascularity
- Maintained testicular contour

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Epidermoid Cyst

KEY FACTS

TERMINOLOGY

- Rare, benign, keratin-containing lesion of controversial origin

IMAGING

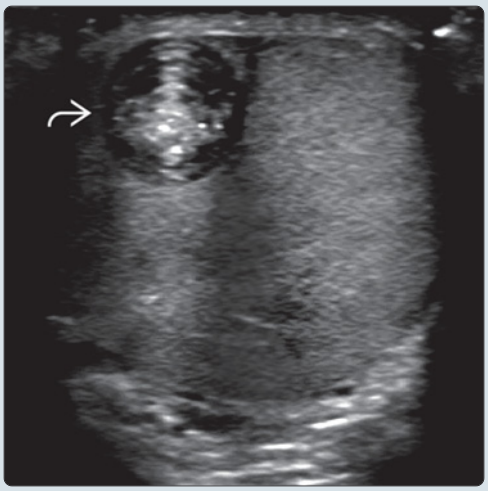
- High-resolution US (≥ 7.5 MHz) is imaging modality of choice
- **Grayscale US**
  - Characteristic onion skin or target/bull's-eye appearance of avascular testicular "mass"
  - Unilocular cyst containing keratin; fibrous wall
  - Sharply circumscribed, encapsulated round "mass"
- **Color Doppler US**
  - Avascular, no blood flow demonstrable
- Most commonly intratesticular, but rarely may be extratesticular

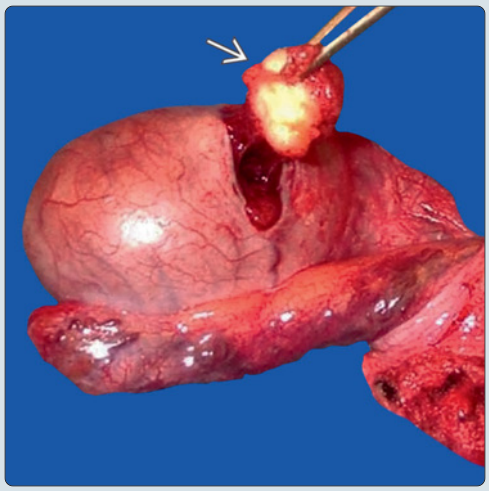
TOP DIFFERENTIAL DIAGNOSES

- Tunica albuginea cyst
  - Located within tunica: Solitary, unilocular, and anechoic
- Germ cell tumor
  - Heterogeneous mass with vascularity seen on Doppler
- Testicular granuloma
  - Most probably due to TB, usually multiple

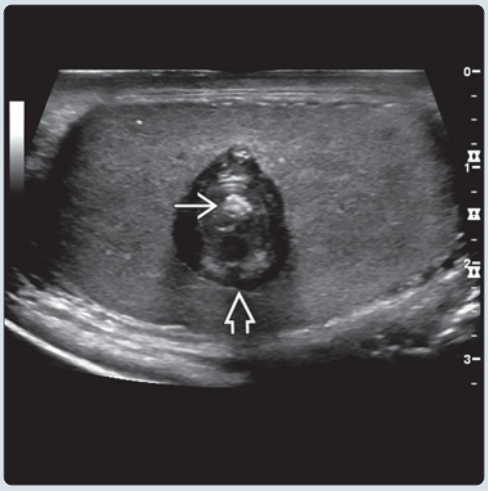
CLINICAL ISSUES

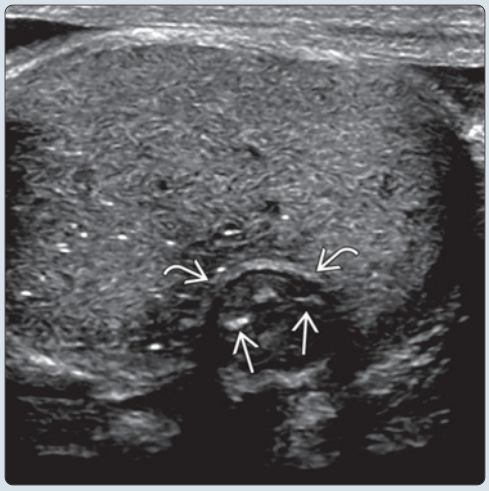
- May occur at any age; 2nd-4th decades most common
- 1-2% of all testicular tumors
- No malignant potential
- Enucleate 1st if lesions < 3 cm with characteristic US appearance and no color flow; testis can be spared if
  - Frozen sections of lesion are consistent with epidermoid cyst
  - No evidence of malignancy within or surrounding lesion
  - Negative tumor markers (AFP, β-HCG)





(Left) *Scrotal ultrasound of a young male shows a testicular mass composed of concentric rings of alternating echogenicity and an echogenic core. The "onion skin" appearance is due to lamellated keratin layers and strongly suggests an epidermoid cyst.* **(Right)** *Tumor markers were negative and the mass was enucleated. Frozen sectioning confirmed an epidermoid cyst. Recognition of classic sonographic features should prompt an attempt at testis-sparing surgery.*





(Left) *Ultrasound of the testis demonstrates a well-demarcated intratesticular epidermoid cyst. Central echogenicity is due to keratin.* **(Right)** *Scrotal ultrasound performed on a different patient to assess for a residual abscess post scrotal wall incision and drainage shows a solid mass with an echogenic rim and several internal echogenic reflectors, an appearance suggestive of an epidermoid cyst. Intraoperative frozen sectioning was inconclusive. Orchiectomy confirmed a foreign body granuloma.*

TERMINOLOGY

Synonyms

- Monodermal dermoid, keratin/cyst of testis

Definitions

- Rare, benign, keratin-containing lesion of controversial origin

IMAGING

General Features

- Best diagnostic clue
  - Characteristic onion skin or target/bull's-eye appearance of avascular testicular "mass"
- Location
  - Most commonly intratesticular, but rarely may be extratesticular
- Size
  - 0.5-10.5 cm in diameter
- Morphology
  - Unilocular cyst containing keratin; fibrous wall

Ultrasonographic Findings

- Grayscale ultrasound
  - Sharply circumscribed, encapsulated round "mass"
  - 4 ultrasound appearances varying with maturation, compactness, and amount of keratin present
    - Type 1: Classic onion skin appearance (concentric hypoechoic and hyperechoic rings)
    - Type 2: Densely calcified echogenic mass with posterior acoustic shadowing
    - Type 3: Target/bull's-eye appearance (cystic-appearing lesion with echogenic center; secondary to compact keratin/calcification)
    - Type 4: Mixed pattern
- Color Doppler
  - Avascular, no blood flow demonstrable

MR Findings

- Can have "bull's-eye" or "onion skin" appearance similar to what is seen on ultrasound; however
  - What appears bright on ultrasound (central keratin and fibrous capsule) has low signal on both T1WI and T2WI
  - What appears dark on ultrasound (desquamated cellular debris with ↑ water and fat content) has high signal on T1WI and T2WI
- Does not enhance → differentiates cyst from neoplasm

Imaging Recommendations

- Best imaging tool
  - High-resolution US (≥ 7.5 MHz)

DIFFERENTIAL DIAGNOSIS

Tunica Albuginea Cyst

- Located within tunica: Solitary, unilocular, and anechoic

Germ Cell Tumor

- Heterogeneous mass with vascularity seen on Doppler
- Testicular teratoma: Appearance may be similar to dermoid, but are typically larger

Testicular Granuloma

- Most probably due to TB, usually multiple

PATHOLOGY

General Features

- Etiology
  - Controversial etiology
  - Prevailing theory: Monodermal teratoma composed entirely of ectoderm
  - Alternate theories
    - Metaplasia of seminiferous tubules or rete testis
    - Due to abnormal closure of neural groove

Microscopic Features

- Unilocular cyst surrounded by fibrous wall composed at least partially of squamous epithelium
- Contains keratin and desquamated material

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Incidental, painless tumor with negative tumor markers: May cause diffuse testicular enlargement

Demographics

- Age
  - May present at any age; 2nd-4th decades most common
- Epidemiology
  - 1-2% of all testicular tumors
  - 3% of all pediatric tumors

Natural History & Prognosis

- No malignant potential

Treatment

- Enucleate 1st if lesions < 3 cm with characteristic US appearance and no color flow
- Testis can be spared if
  - Frozen sections of lesion are consistent with epidermoid cyst
  - No evidence of malignancy within or surrounding lesion
  - Negative tumor markers (AFP, β-HCG)

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- "Onion skin" appearance in well-circumscribed testicular mass on US
- Avascular benign mass on color Doppler

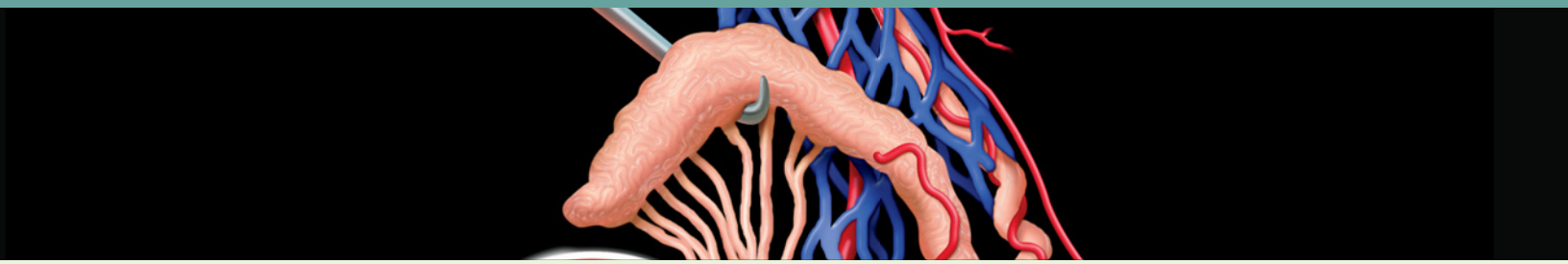
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SECTION 9

Epididymis



Epididymitis	**490**
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Spermatocele/Epididymal Cyst	**496**
Sperm Granuloma	**498**

Epididymitis

KEY FACTS

TERMINOLOGY

- Inflammation of epididymis &/or testis

IMAGING

- Test of choice: Color Doppler US; high-frequency transducers (≥ 10 MHz)
- Diffuse or focal hyperemia in body and tail of epididymis ± increased vascularity of testis (compare with contralateral testis if subtle)
- Starts within tail of epididymis → body → testis
- Orchitis is usually secondary, occurring in 20-40% of epididymitis due to contiguous spread of infection
  - Can cause vascular compromise → ischemia → testicular infarction → sonographic features indistinguishable from testicular torsion
  - Reversal of arterial diastolic flow of testis is ominous finding associated with testicular infarction

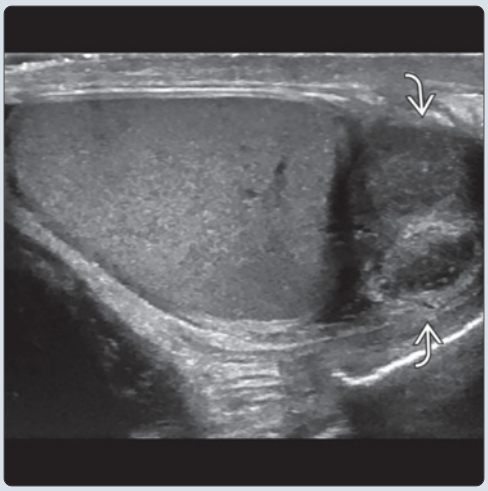
TOP DIFFERENTIAL DIAGNOSES

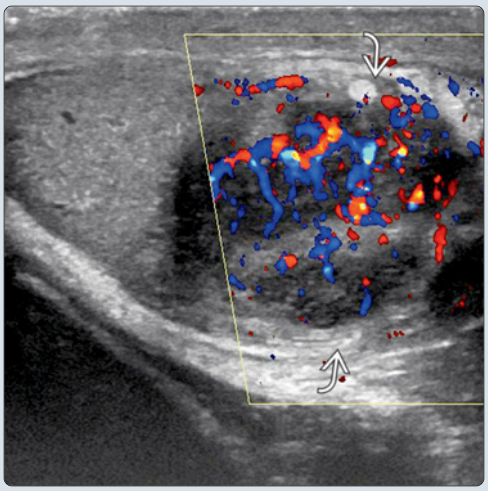
- Testicular torsion
- Testicular lymphoma
- Testicular trauma

CLINICAL ISSUES

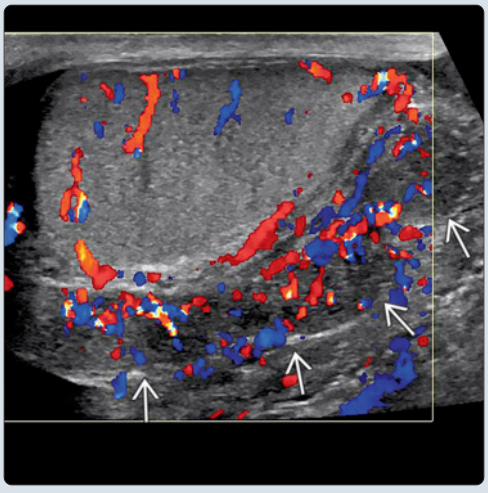
- Most common cause of acute scrotal pain in adolescent boys and adults (15-35 years)
- Males 14-35 years of age: Most commonly caused by *Neisseria gonorrhoeae* and *Chlamydia trachomatis*
- Scrotal swelling, erythema; fever; dysuria
  - Scrotal pain due to epididymitis usually relieved after elevation of testes (scrotum) over symphysis pubis (Prehn sign)
  - Associated lower urinary tract infection and its symptoms, urethral discharge
- Prognosis excellent if treated early with antibiotics; follow-up scans to exclude abscess if no improvement

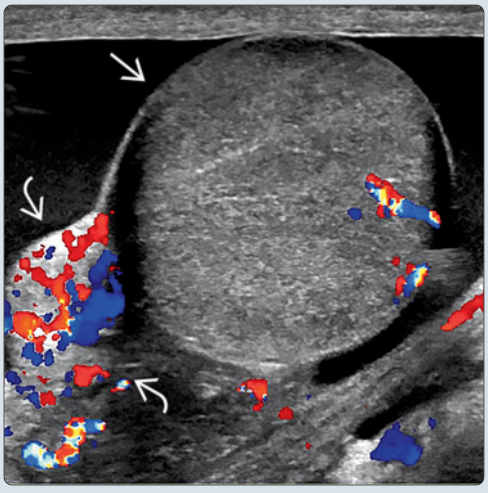
(Left) *US performed on a 39-year-old man with subacute scrotal pain shows a relatively enlarged, hypoechoic left epididymal tail.* **(Right)** *Color Doppler US performed on the same patient shows that the epididymal tail mass is hyperemic. The patient ultimately reported a recent history of treated sexually transmitted disease. Clinical symptoms and vascularity improved after a course of antibiotics. The clinical history and Doppler appearance are classic for early, uncomplicated epididymitis.*





(Left) *Color Doppler US performed on an elderly man with persistent severe scrotal pain despite antibiotic therapy for epididymitis following a prostate biopsy shows a persistently enlarged, hyperemic right epididymis.* **(Right)** *Color Doppler of the same patient shows a hyperemic left epididymis, and minimal flow within a heterogeneous testis. Aggressive or poorly treated epididymitis may result in testicular venous infarction (confirmed in this case, post orchiectomy).*





TERMINOLOGY

Synonyms

- Acute scrotum, orchitis, epididymo-orchitis

Definitions

- Infectious inflammation of epididymis &/or testis

IMAGING

General Features

- Best diagnostic clue
  - Enlarged, hyperemic epididymis &/or testis on color Doppler US
- Location
  - Early epididymitis often involves tail of epididymis
  - Orchitis is usually secondary, occurring in 20-40% of epididymitis due to contiguous spread of infection
    - Primary orchitis is caused by mumps and is usually bilateral
- Size
  - Epididymis typically 2-3x larger than normal
- Morphology
  - Focal enlargement of tail or diffuse enlargement of entire epididymis

Ultrasonographic Findings

- Grayscale ultrasound
  - Epididymitis: Primarily involved in epididymo-orchitis
    - Acute: Enlarged epididymis, ↓ echogenicity, coarse heterogeneous echotexture due to edema and hemorrhage
    - Chronic epididymitis: Enlarged hyperechoic epididymis
  - Orchitis: Follows in 20-40% of epididymitis due to contiguous spread of infection
    - Focal or diffuse
      - Focal: Hypoechoic area adjacent to enlarged portion of epididymis
      - Diffuse: Testis is diffusely enlarged with heterogenous echotexture, thickening of tunica albuginea (in severe infection)
    - Edematous, congested testes contained within rigid tunica albuginea → ↑ intratesticular pressure
      - Heterogeneous parenchymal echogenicity (predominantly hyperechoic initially and hypoechoic later) and septal accentuation visible as hypoechoic bands
      - May lead to venous occlusion
      - Can cause vascular compromise → ischemia → testicular infarction → sonographic features indistinguishable from testicular torsion
  - Spermatic cord may be inflamed and may appear hypoechoic with associated hyperechoic fat within
  - Reactive hydrocele containing low-level internal echoes, septa, thickening of tunical layers ± skin edema; all represent changes of periorchitis
- Color Doppler
  - Diffuse or focal hyperemia in body and tail of epididymis ± increased vascularity of testis → highly sensitive and specific for epididymo-orchitis
  - Inflammation of epididymis and testis is associated with ↓ vascular resistance compared with healthy individuals
    - Resistive index (RI) < 0.5 (normal RI in testis ≥ 0.5)
  - Signs of infarction in cases with severe epididymo-orchitis
    - Relatively avascular areas within hyperemic testis or epididymis suggest focal infarction
    - Reversal of arterial diastolic flow of testis (seen with obstruction of venous outflow/venous infarction) is ominous finding associated with testicular infarction

Nuclear Medicine Findings

- Tc-99m: 90% accurate in differentiating torsion from epididymitis
  - Increased flow within testicular vessels and vas deferens on flow study
  - Markedly increased perfusion through spermatic cord vessels (testicular + deferential arteries)
  - Curvilinear increased activity laterally in hemiscrotum on static images (also centrally if testis is involved)

Imaging Recommendations

- Best imaging tool
  - Color Doppler US; high-frequency transducers (≥ 10 MHz)
- Protocol advice
  - Comparison with contralateral testis is useful when increase in vascularity is subtle

DIFFERENTIAL DIAGNOSIS

Testicular Torsion

- Absent or diminished color Doppler flow, "twist" of spermatic cord in inguinal region
- Epididymis may be enlarged but not hyperemic on color Doppler US
- Clinical differentiation results in 50% false-positive rate and unnecessary surgical exploration

Testicular Lymphoma

- Often large in size at time of diagnosis, commonly occurs in association with disseminated disease
- Heterogeneous echopattern; often bilateral; involvement of epididymis and spermatic cord is common; hemorrhage and necrosis are rare

Testicular Trauma

- History of trauma; acute pain
- Focal hypoechoic avascular area on color Doppler; rupture of tunica albuginea associated with hematocele

PATHOLOGY

General Features

- Etiology
  - Ascending genitourinary tract infection
    - In males 14-35 years of age, disease most frequently caused by *Neisseria gonorrhoeae* and *Chlamydia trachomatis*
    - In prepubertal boys, men over 35 years of age, and men who practice anal intercourse, disease most frequently caused by *Escherichia coli*
    - Other causes: *Pseudomonas*, *Klebsiella*, *Proteus mirabilis*, *Staphylococcus aureus*, *Mycobacterium tuberculosis*, mumps virus

- Bacterial seeding occurs directly in cases with GU anomaly and presumably hematogenously in cases without demonstrable anomaly
- Primary orchitis is rare and caused by mumps (usually bilateral)
- Traumatic epididymitis: Similar findings as infectious epididymitis
  - However, patient will have preceding history of scrotal trauma ± additional traumatic features such as hematocele &/or testicular injury
  - Conservative management: Antibiotics not needed
  - Should not be confused with infectious epididymitis
- Drugs such as amiodarone hydrochloride may cause chemical epididymitis

Staging, Grading, & Classification

- Epididymitis: Isolated epididymitis, focal or diffuse
  - Acute/chronic epididymitis
- Orchitis or combined epididymitis and orchitis
  - Primary: Isolated orchitis (may be seen in boys with mumps)
  - Secondary: Infection spread from adjacent epididymis
  - Acute/chronic orchitis or epididymo-orchitis

Gross Pathologic & Surgical Features

- Treated surgically only if abscess forms despite antibiotic treatment

Microscopic Features

- Inflammatory infiltrate of testis and epididymis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Most common cause of acute scrotal pain in adolescent boys and adults
  - Scrotal swelling, erythema; fever; dysuria
    - Prehn sign: Scrotal pain due to epididymo-orchitis usually relieved after elevation of scrotum over symphysis pubis → may help clinically to differentiate from torsion
  - Associated lower urinary tract infection and its symptoms, urethral discharge
- Other signs/symptoms
  - Pyuria (95%), prostatic tenderness (infrequent)
  - ↑ C-reactive protein (CRP) can help distinguish epididymo-orchitis from testicular torsion (sensitivity 96%, specificity 94%)
  - Positive urinalysis for WBC and bacteria; may have elevated WBC

Demographics

- Age
  - Most commonly 15-35 years
- Epidemiology
  - Most frequently seen in sexually active young men; also seen in infants and boys

Natural History & Prognosis

- Prognosis excellent if treated early with antibiotics
- Complications
  - Abscess formation (epididymal abscess: 6%, testicular abscess: 6%), microabscesses usually seen in low-grade infection such as tuberculosis and in immunocompromised host
  - Testicular infarction
    - Venous infarction: Due to venous outflow obstruction
    - Thrombosis of main testicular artery or its branches secondary to chronic inflammation
    - Gangrene is rare but known complication
  - Gonadal vein thrombosis
  - Pyocele
  - Late testicular atrophy (21%)
  - Recurrent infection may lead to infertility

Treatment

- Antibiotic therapy; follow-up scans to exclude abscess if no improvement
- Work-up for GU anomalies in younger children and recurrent cases
- Bed rest, scrotal elevation, analgesics

DIAGNOSTIC CHECKLIST

Consider

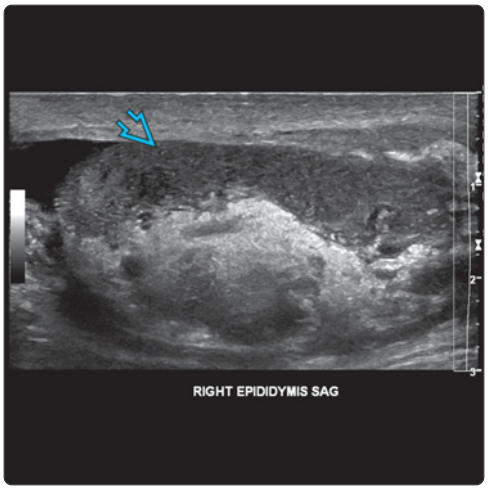
- Torsion if low or absent flow within testis

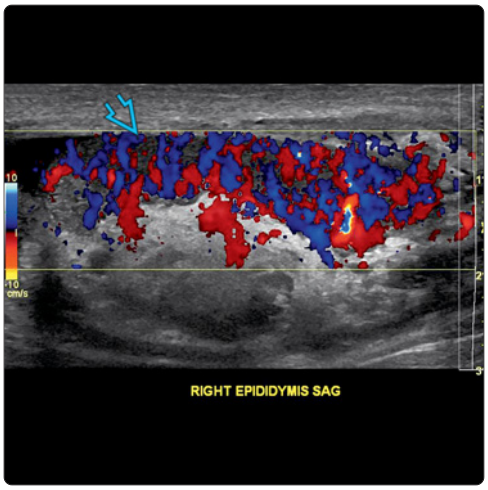
Image Interpretation Pearls

- Hyperemic and enlarged epididymis &/or testis

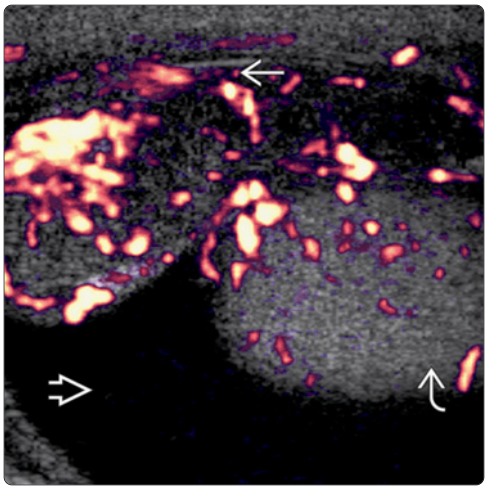
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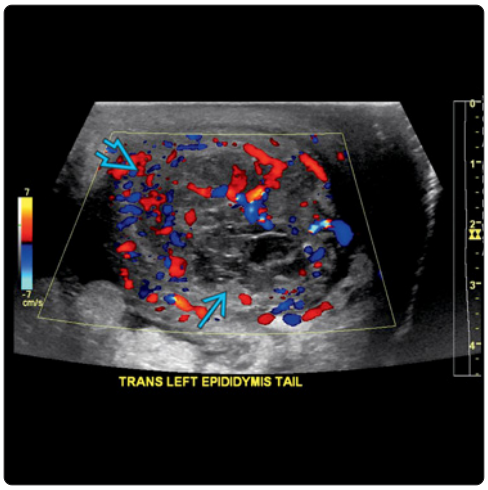
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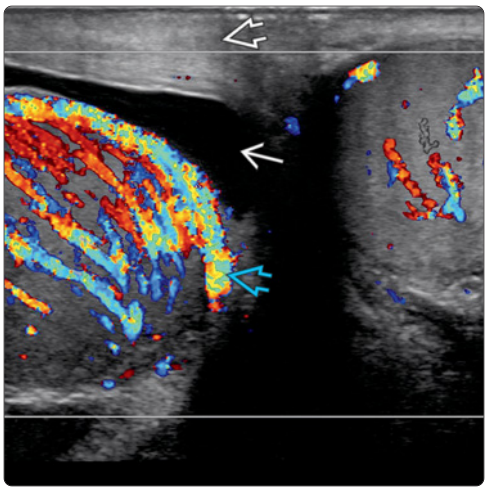


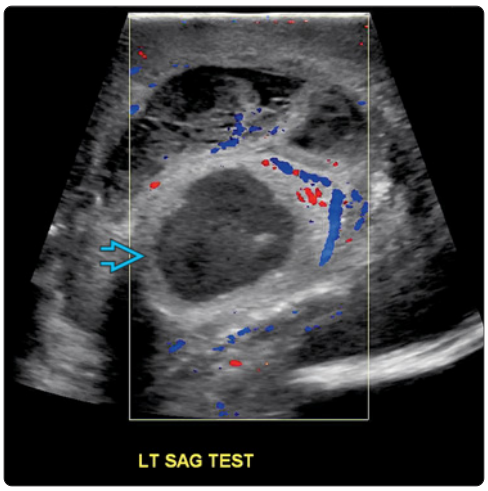
(Left) *Sagittal grayscale US of the right scrotum demonstrates a thickened heterogeneous and hypoechoic epididymis.* **(Right)** *Color Doppler of the epididymis in the same patient reveals marked hyperemia of the epididymis consistent with acute epididymitis. Side-to-side grayscale and Doppler comparison is useful for more equivocal cases.*





(Left) *Oblique power Doppler US shows a markedly enlarged, hypoechoic epididymis with increased vascularity, suggesting acute epididymitis. Note normal testis and hydrocele.* **(Right)** *Color Doppler of the left epididymis reveals a markedly hyperemic tail with a central heterogenous hypoechoic lesion. The findings are consistent with epididymitis complicated by an epididymal abscess.*





(Left) *Transverse color Doppler image of bilateral testicles shows the right testicle is markedly hypervascular in appearance compared with the left, consistent with right-sided orchitis. In addition, there are associated findings of scrotal wall thickening and a small hydrocele.* **(Right)** *Color Doppler of the left testis in a patient who presented with acute epididymo-orchitis reveals a hypoechoic, thick-walled lesion within the testicle consistent with a testicular abscess.*

Adenomatoid Tumor

KEY FACTS

TERMINOLOGY

- Benign solid paratesticular tumor of mesenchymal origin

IMAGING

- Solid intrascrotal mass, usually extratesticular
- Location
  - Epididymis: Most common location overall
  - May arise in tunica albuginea
  - Rarely intratesticular or other locations, such as spermatic cord and prostate
- Imaging appearance
  - Rounded or ovoid
  - Well circumscribed
  - Varying echogenicity
  - Gentle transducer pressure may show mass can move independent of testis
  - Refractive edge shadows on grayscale US
  - Hypovascular or avascular on color Doppler US
  - Size: 5 mm to 5 cm

TOP DIFFERENTIAL DIAGNOSES

- Leiomyoma
- Lipoma
- Cystadenoma

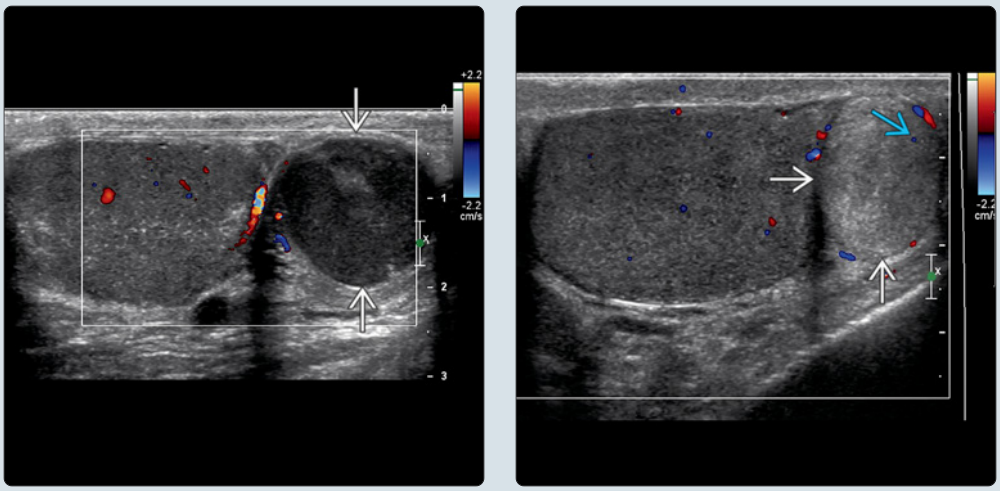
CLINICAL ISSUES

- Most common solid mass in epididymis
  - 36% of all paratesticular tumors
- Slowly enlarges over years
- Most surgically excised to confirm diagnosis
- Some urologists and patients elect surveillance
- Age: 20 years and older
  - Mean age: 36
  - Rarely seen in boys

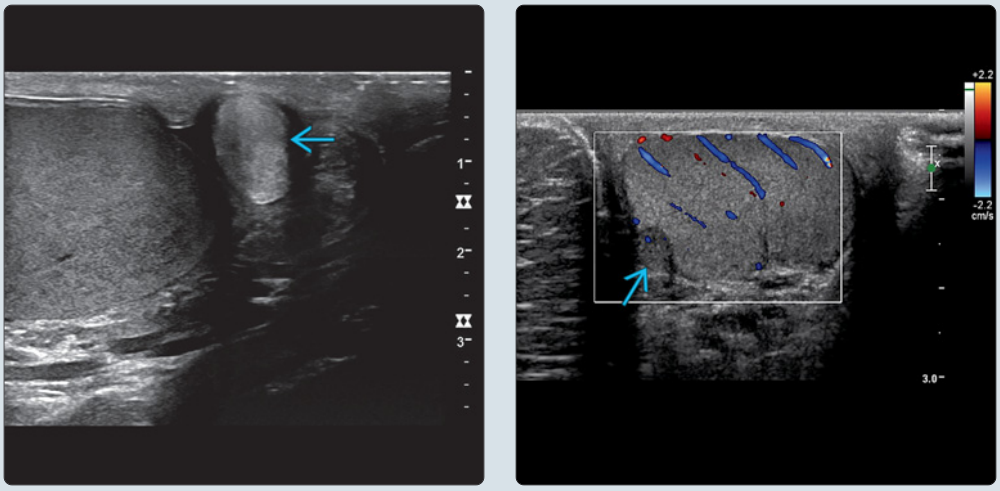
DIAGNOSTIC CHECKLIST

- Consider leiomyoma

(Left) *Longitudinal US in a 41-year-old male with a history of a palpable scrotal mass shows a hypoechoic adenomatoid tumor, with no internal flow, within the epididymal tail.* **(Right)** *Ultrasound of a 50-year-old man with 5 years of ultrasound surveillance for a presumed adenomatoid tumor shows an ovoid, well-circumscribed mass in the tail of the epididymis that is isoechoic to testis and shows minimal internal vascularity. Note the very different echogenicity compared to the otherwise similar patient shown previously.*



(Left) *Longitudinal US in a 37-year-old man shows an incidental hyperechoic ovoid mass in the epididymal tail. The tumor was resected and was compatible with an adenomatoid tumor.* **(Right)** *Transverse US shows an adenomatoid tumor of the tunica albuginea that was incidentally noted on a pelvic MR image (not shown) in a 12-year-old boy. The lesion is equally endophytic and exophytic to the testis, as it is centered in the tunica. Note minimal internal vascularity.*



TERMINOLOGY

Definitions

- Benign solid paratesticular tumor of mesenchymal origin

IMAGING

General Features

- Best diagnostic clue
  - Solid intrascrotal mass, usually extratesticular
- Location
  - Epididymis: Most common location overall
    - Most commonly in tail
  - Tunica albuginea
  - Rarely intratesticular or other locations, such as spermatic cord and prostate
- Size
  - 5 mm to 5 cm
- Morphology
  - Rounded or ovoid
  - Well circumscribed

Imaging Recommendations

- Best imaging tool
  - US
- Protocol advice
  - Grayscale US with color Doppler imaging

Ultrasonographic Findings

- Solid mass within epididymis or tunica, varying echogenicity
  - Gentle transducer pressure may show mass can move independent of testis
- Hypovascular or avascular on color Doppler US
- Refractive edge shadows on grayscale US

DIFFERENTIAL DIAGNOSIS

Leiomyoma

- Solid or cystic, may contain calcifications
- Most often located in epididymal head

Lipoma

- 45% of all paratesticular masses
- Composed of mature fat cells
  - Homogeneous and hyperechoic on US
  - High signal on T1-weighted MR

Cystadenoma

- Reported in 60% of men with von Hippel-Lindau disease
- May occur sporadically, typically in middle age
- May be primarily cystic, or mixed solid and cystic on US

PATHOLOGY

General Features

- Etiology
  - Unknown
  - Believed to be mesothelial in origin

Gross Pathologic & Surgical Features

- Solid, homogeneous, yellowish nodule with smooth surface
  - Most common location is tail of epididymis
- Rarely may occur in tunica, testis, or spermatic cord
- Generally not encapsulated

Microscopic Features

- Lesion comprised of irregular tubules lined with flattened and cuboid epithelial or endothelial cells
- Keratin positive on immunoperoxidase stains
- Cells are positive for mesothelial related markers (calretinin, HMBE1)
- Increasing recognition that FNA can provide accurate diagnosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Many patients are asymptomatic
  - Slowly growing palpable mass
- Other signs/symptoms
  - 5% of patients present with pain

Demographics

- Age
  - 20 years and older
    - Mean age: 36
  - Rarely seen in boys
- Gender
  - Only male patients have scrotal adenomatoid tumors
  - Masses of similar histology may arise in female genital tract as well
- Epidemiology
  - Most common solid mass in epididymis
  - 36% of all paratesticular tumors

Natural History & Prognosis

- Slowly enlarges over years

Treatment

- Most surgically excised to confirm diagnosis, usually testis-sparing
- Some urologists and patients elect surveillance
- Emerging role for FNA

DIAGNOSTIC CHECKLIST

Consider

- Leiomyoma

Image Interpretation Pearls

- Solid paratesticular mass
- Hypovascular or avascular on color Doppler US

SELECTED REFERENCES

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Spermatocele/Epididymal Cyst

KEY FACTS

TERMINOLOGY

- Spermatocele: Retention cyst of rete testis or epididymis containing spermatozoa
- Epididymal head cyst: Collection of simple fluid

IMAGING

- Protocol advice: Grayscale US with color Doppler imaging
- US findings
  - Simple or complex cystic mass: May be multiseptated; may contain diffuse, low-level echoes due to spermatozoa
  - "Falling snow" sign: 2° color Doppler mechanically moving particular matter within cyst

PATHOLOGY

- Etiology: Unknown, may reflect scarring 2° either epididymitis, trauma, or vasectomy
- Associated abnormalities: Dilation of rete testis within mediastinum of testis

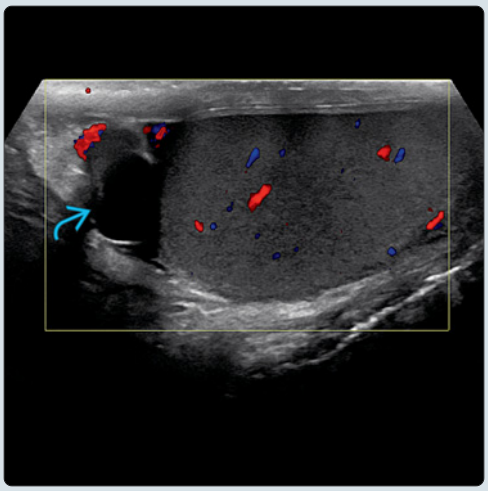
CLINICAL ISSUES

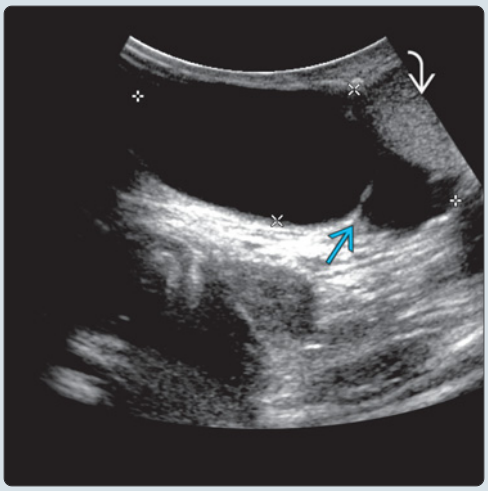
- Most common signs/symptoms: Painless scrotal mass
  - Rare hypofertility or torsion
- Treatment: Most can be observed if asymptomatic
  - Surgery (spermatocelectomy) only if sufficiently large to cause discomfort

DIAGNOSTIC CHECKLIST

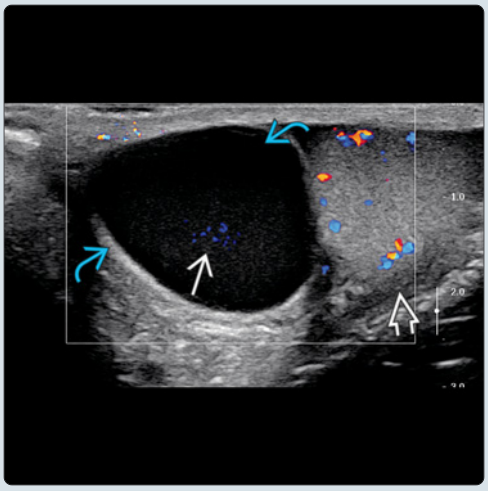
- Image interpretation pearls: Cystic mass in head of epididymis, either simple or with falling snow sign on color Doppler US
- Cystic extratesticular lesions almost always benign; if no vascularized soft tissue elements, not neoplasm

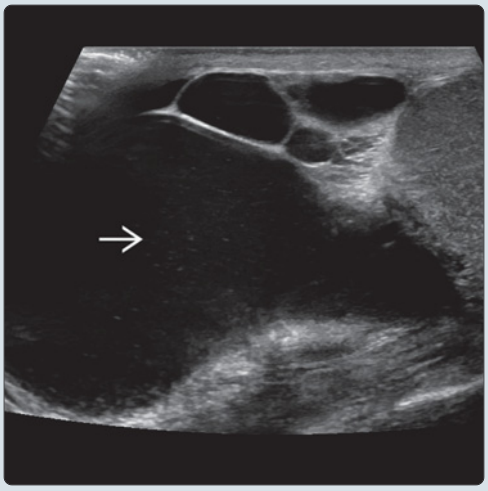
(Left) *Typical appearance of an incidentally discovered and clinically unimportant epididymal cyst is shown. Note the well-circumscribed, anechoic, avascular lesion in the epididymal head.* **(Right)** *Large epididymal cyst (calipers) is shown with a single thin septation in the right hemiscrotum, displacing the testis inferiorly. Note that an abdominal probe rather than a high-frequency linear probe was needed to capture this large lesion.*





(Left) *Large spermatocele superior to right testis contains punctate low-level echoes with corresponding Doppler artifact. These punctate foci were mobile in real time.* **(Right)** *Longitudinal grayscale ultrasound demonstrates a complex, multiseptate cystic mass with numerous locules containing debris, characteristic findings for a spermatocele.*





TERMINOLOGY

Definitions

- Spermatocele: Retention cyst of rete testis or epididymis containing spermatozoa
- Epididymal head cyst: Collection of simple fluid

IMAGING

General Features

- Best diagnostic clue
  - Simple or minimally complex cystic mass within head of epididymis
- Location
  - Head of epididymis
- Size
  - 3 mm to 10 cm
- Morphology
  - Epididymal head cysts are anechoic, thin walled, and avascular
  - Spermatoceles may contain linear septations and internal debris

Imaging Recommendations

- Best imaging tool
  - US
- Protocol advice
  - Grayscale US with color Doppler imaging

Ultrasonographic Findings

- Simple or complex cystic mass: May be multiseptated; may contain diffuse; low-level echoes due to spermatozoa
- Falling snow sign: 2° to color Doppler mechanically moving particulate matter within cyst

DIFFERENTIAL DIAGNOSIS

Hematocele

- History of trauma
- Complex fluid collection within tunica vaginalis
- Contains echogenic clot and fibrin strands

Pyocele

- Associated with epididymo-orchitis, with hyperemia and enlargement of epididymis &/or testis
- Complex fluid collection located within tunica vaginalis
- Contains white cell debris and fibrinous strands producing linear septations and low-level echoes

Hydrocele

- Simple or complex intrascrotal fluid collection within tunica vaginalis

Cyst of Tunica Albuginea

- Usually small (2-5 mm) simple cyst between layers of tunica albuginea

PATHOLOGY

General Features

- Etiology
  - Scarring 2° to either epididymitis, trauma, or vasectomy
- Associated abnormalities
  - Dilation of rete testis within mediastinum of testis

Gross Pathologic & Surgical Features

- Complex cystic mass containing simple fluid or milky fluid and spermatozoa, depending on type

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Painless scrotal mass
    - Rare hypofertility or torsion
  - Cystic masses can be transilluminated like hydroceles at physical exam

Demographics

- Age
  - Any age can be affected
- Epidemiology
  - Very common; present in 20-40% of men

Treatment

- Most can be observed if asymptomatic
- Surgery (spermatocelectomy) only if sufficiently large to cause discomfort

DIAGNOSTIC CHECKLIST

Consider

- Abscess: Correlate with recent epididymitis and ongoing hyperemia
- Inguinal hernia: Should vary with Valsalva and exhibit expected gut signature or echogenic fat

Image Interpretation Pearls

- Cystic mass in head of epididymis with falling snow sign on color Doppler US
- Cystic extratesticular lesions almost always benign; if no vascularized soft tissue elements, not neoplasm

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Sperm Granuloma

KEY FACTS

IMAGING

- Ultrasound: Solid, well-circumscribed hypoechoic epididymal lesion
- Nonspecific appearance, though clinical history, vasectomy, or other causes of epididymal tubular obstruction (trauma, infection) may prompt diagnosis

TOP DIFFERENTIAL DIAGNOSES

- Adenomatoid tumor
  - Solitary, varying echogenicity lesion: Most common solid epididymal tumor
- Chronic epididymitis
  - Enlarged, heterogeneous epididymis: Elicit history of prior bacterial epididymitis
  - Other causes (drug induced, traumatic) possible
- Leiomyoma
  - 2nd most common solid epididymal tumor

PATHOLOGY

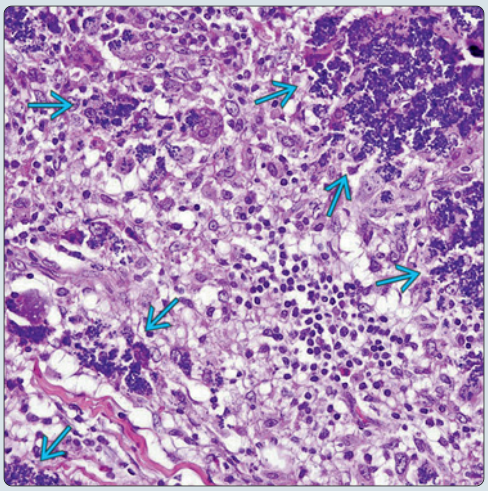
- Foreign body giant cell reaction to extravasated sperm after epididymal obstruction, most commonly post vasectomy

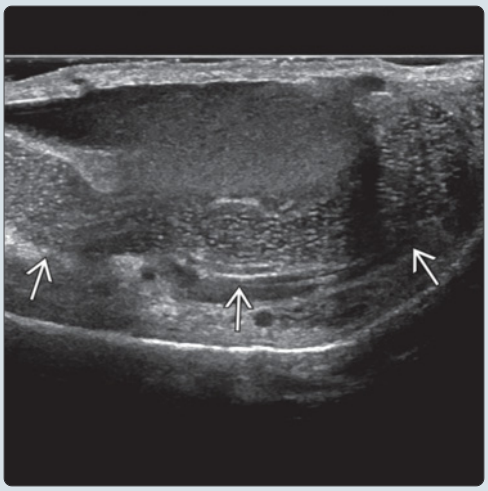
CLINICAL ISSUES

- A benign inflammatory process: Typically asymptomatic palpable mass
- Pathophysiology of sperm granulomas (tubular obstruction) may lead to post vasectomy pain syndrome in 10% of males, 5-7 years post procedure

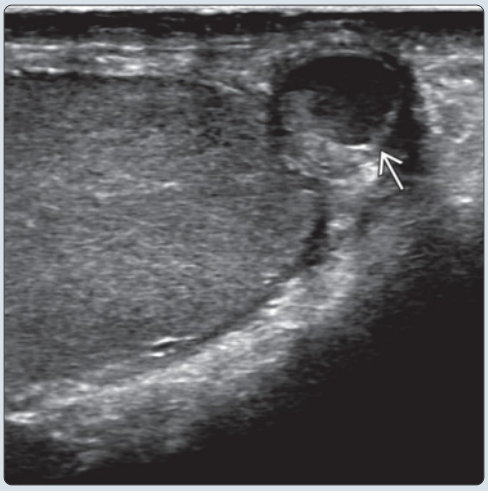
DIAGNOSTIC CHECKLIST

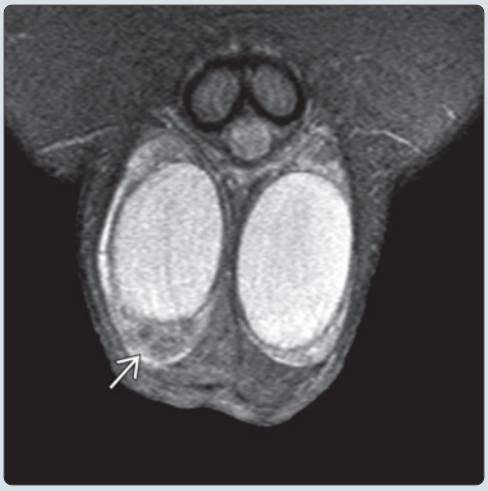
- Consider if small, solid, hypoechoic epididymal mass, post vasectomy
- Physical examination &/or ultrasound follow-up may suffice





(Left) *H&E stain of an epididymal mass shows aggregates of extravasated sperm surrounded by inflammatory cells, typical of a sperm granuloma. (From DP: Genitourinary.)* **(Right)** *Ultrasound of the left testis in a man with pain post vasectomy shows an enlarged epididymis and tubular ectasia. Obstruction after vasectomy (or due to trauma, infection) results in sperm extravasation and localized foreign body reactions (sperm granuloma).*





(Left) *Ultrasound performed to evaluate a palpable mass 4 years post vasectomy shows a well-circumscribed hypoechoic right epididymal tail mass. Epididymectomy performed because of lesion growth showed fibrosis, granulation, and a sperm granuloma.* **(Right)** *Preoperative coronal FSE T2 MR of the same patient shows a corresponding T2 hypointense epididymal tail lesion. Scrotal MR is typically reserved for evaluation of large masses, though ↑ epididymal T1 intensity may suggest a poor vasectomy reversal outcome.*

TERMINOLOGY

Synonyms

- Epididymitis nodosa

Definitions

- Type of chronic epididymitis, commonly seen post vasectomy

IMAGING

General Features

- Best diagnostic clue
  - Well-defined hypoechoic epididymal nodule post vasectomy
- Location
  - Classic: Ends of severed vas deferens post vasectomy
  - May occur anywhere along ductal system
- Size
  - Microscopic to 4 cm; usually < 1 cm
- Morphology
  - Well circumscribed, may be multiple

MR Findings

- T1WI
  - ↑ epididymal signal may reflect proteinaceous material with epididymal tubules (postulated precursor for granuloma formation)
  - May predict poor outcome for standard vasovasostomy

Ultrasonographic Findings

- Grayscale ultrasound
  - Post vasectomy epididymis
    - Globally enlarged epididymis
    - Epididymal tubular ectasia
  - Sperm granuloma: Well-circumscribed hypoechoic epididymal nodule(s)

Imaging Recommendations

- Best imaging tool
  - High-frequency (> 7.5 MHz) ultrasound
- Protocol advice
  - Patient directed linear probe positioning over palpable nodule

DIFFERENTIAL DIAGNOSIS

Adenomatoid Tumor

- Most common epididymal tumor: Benign
- Asymptomatic, palpable mass: Variable echogenicity

Chronic Epididymitis

- Heterogeneous, enlarged epididymis
- Elicit history of prior bacterial epididymitis
- Other causes (drug induced, traumatic) possible

Leiomyoma

- 2nd most common epididymal neoplasm: Nonspecific US appearance

Inflammatory Pseudotumor

- Rare paratesticular lesion: Inflammatory, fibrous proliferation secondary to prior trauma or inflammation

PATHOLOGY

General Features

- Early post vasectomy: ↑ intraluminal fluid pressure within obstructed epididymis: dilatation of efferent ductules, spermatoceles
- Intermediate post vasectomy: Macrophage infiltration and clearance of sperm/fluid
- Delayed post vasectomy changes: Extravasation of sperm into epididymal interstitium → foreign body giant cell reaction (sperm granuloma)

Gross Pathologic & Surgical Features

- Definitive diagnosis at partial epididymectomy, though FNA of chronic (likely benign) epididymal lesions advocated

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Palpable, nonpainful, paratesticular (epididymal) mass
  - Typically post vasectomy but may occur with other causes of epididymal obstruction (trauma, infection)

Demographics

- Autopsy series: Prevalence of up to 42% of men post vasectomy (2.5% of general male population)

Natural History & Prognosis

- Lesion itself is benign, although pathophysiology of sperm granuloma formation (obstructed tubules, epididymal congestion, perineural inflammation) may lead to post vasectomy pain syndrome years after procedure

Treatment

- Physical examination follow-up if appropriate clinical history typically suffices
- Partial epididymectomy if equivocal history and growth
- Post vasectomy pain syndrome: Oral analgesia, local analgesia, steroid injection, and rarely epididymectomy

DIAGNOSTIC CHECKLIST

Consider

- Majority of solid epididymal lesions are benign

Image Interpretation Pearls

- Nonspecific appearance (solid, hypoechoic epididymal lesion), but suggest sperm granuloma if history of vasectomy and post vasectomy changes (enlarged epididymis, epididymal tubular ectasia)

Reporting Tips

- Follow-up for stability by physical examination &/or ultrasound

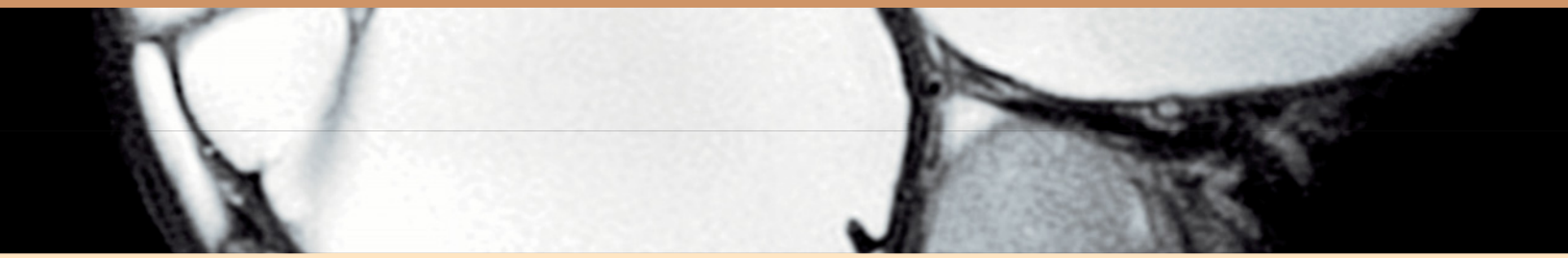
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SECTION 10

Scrotum



Hydrocele	**502**
Varicocele	**504**
Pyocele	**506**
Hernia	**508**
Fournier Gangrene	**512**
Scrotal Trauma	**514**

Hydrocele

KEY FACTS

TERMINOLOGY

- Congenital or acquired serous fluid contained within layers of tunica vaginalis

IMAGING

- Best diagnostic clue: Scrotal fluid collection surrounding testis except for "bare area" where tunica vaginalis does not cover testis and is attached to epididymis
- Grayscale US: Crescentic anechoic fluid collection surrounding testis
- Avascular on color Doppler US
- Best imaging tool: Ultrasound

TOP DIFFERENTIAL DIAGNOSES

- Pyocele
- Spermatocele
- Hematocele
- Scrotal hernia

PATHOLOGY

- Congenital or communicating hydrocele secondary to failure of processus vaginalis to close
- Secondary condition in adults due to epididymitis or surgery for varicocele

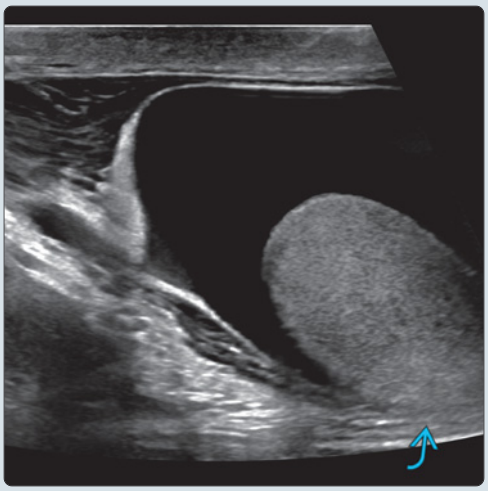
CLINICAL ISSUES

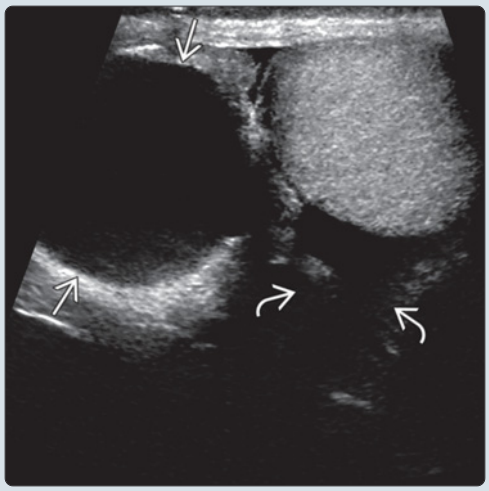
- Excellent prognosis with surgical repair
  - Surgical resection of hydrocele sac with oversewing of edges

DIAGNOSTIC CHECKLIST

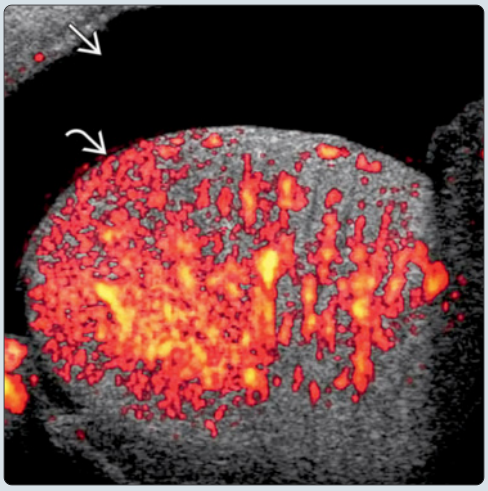
- Consider spermatocele
- Image interpretation pearl
  - Anechoic fluid collection along anterolateral aspect of testis

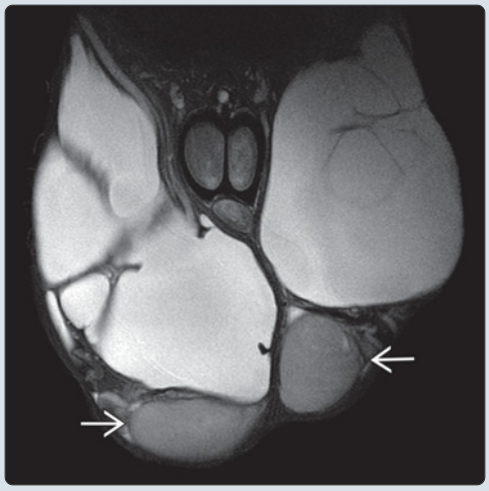
(Left) *Scrotal ultrasound of a 28-year-old male with scrotal discomfort shows a simple hydrocele. Note how with the exception of the "bare" area, the fluid envelopes the testis. Most secondary hydroceles in this age group are due to prior epididymitis.* **(Right)** *Ultrasound performed on a 45-year-old male with a palpable left scrotal mass shows a large epididymal cyst and a small adjacent hydrocele. The cyst displaces the testis rather than enveloping it.*





(Left) *Power Doppler ultrasound of a 27-year-old male who presented with a fever and scrotal pain shows a simple hydrocele adjacent to the testis, which is hyperemic due to orchitis. Note the numerous engorged vessels radiating from the mediastinum testis.* **(Right)** *T2 MR of a male with scrotal swelling shows huge, septated (communicating) hydroceles that conform to the scrotal sac but displace the testes. A prior CT showed complex ascites and carcinomatosis due to peritoneal mesothelioma.*





TERMINOLOGY

Definitions

- Congenital or acquired serous fluid contained within layers of tunica vaginalis

IMAGING

General Features

- Best diagnostic clue
  - Scrotal fluid collection surrounding testis except for "bare area" where tunica vaginalis does not cover testis and is attached to epididymis
- Location
  - Tunica vaginalis, located at anterolateral aspect of testis
- Morphology
  - Simple fluid collection

MR Findings

- T1WI
  - Low signal fluid collection
- T2WI
  - High signal, consistent with serous fluid

Ultrasonographic Findings

- Grayscale ultrasound
  - Crescentic anechoic fluid collection surrounding testis
- Color Doppler
  - Avascular

Imaging Recommendations

- Best imaging tool
  - Ultrasound

DIFFERENTIAL DIAGNOSIS

Pyocele

- Septated fluid with low-level echoes
- Associated with epididymitis, intrascrotal abscess, clinical signs of inflammation

Spermatocele

- Retention cyst of tubules connecting rete testis to head of epididymis
- Cystic mass in head of epididymis
- Low-level echoes and septations if large
- Inferior displacement of testis

Hematocele

- Complex echogenic fluid in tunica vaginalis on US
- Associated with trauma, torsion, infarct

Scrotal Hernia

- Bowel or echogenic omentum seen within scrotum secondary to indirect inguinal hernia

PATHOLOGY

General Features

- Etiology
  - Embryology-anatomy
    - Congenital or communicating hydrocele secondary to failure of processus vaginalis to close
    - Congenital in 6% of male term infants
    - Secondary condition in adults due to epididymitis or surgery for varicocele
- Simple serous fluid collection within tunica vaginalis
- Chronic: Thickened layers of tunica, septations

Staging, Grading, & Classification

- Congenital: 2° incomplete closure of tunica vaginalis
- Acquired: Epididymitis, torsion, trauma, rarely tumor

Gross Pathologic & Surgical Features

- Serous fluid collection

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic scrotal mass
- Other signs/symptoms
  - Mass "transilluminates" when evaluated with light source
- Clinical profile
  - Painless scrotal mass, transilluminating fluid collection
  - Chronic hydrocele with thick walls may not transilluminate

Demographics

- Age
  - All ages
  - Pediatric patients invariably have congenital hydroceles with persistent communication with peritoneal cavity
- Epidemiology
  - 10% of testicular tumors have associated hydrocele

Natural History & Prognosis

- Excellent with surgical repair

Treatment

- Surgical resection of hydrocele sac with oversewing of edges

DIAGNOSTIC CHECKLIST

Consider

- Spermatocele

Image Interpretation Pearls

- Anechoic fluid collection along anterolateral aspect of testis

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Varicocele

KEY FACTS

TERMINOLOGY

- Dilatation of veins of pampiniform plexus > 2-3 mm in diameter secondary to retrograde flow in gonadal (testicular) vein

IMAGING

- Best diagnostic clue
  - Dilated serpiginous veins behind superior pole of testis on color Doppler US
  - Prominent color flow within vessels with Valsalva secondary to retrograde flow
- Protocol advice: Resting and Valsalva color Doppler images of epididymis
- Distention secondary to retrograde flow with Valsalva

TOP DIFFERENTIAL DIAGNOSES

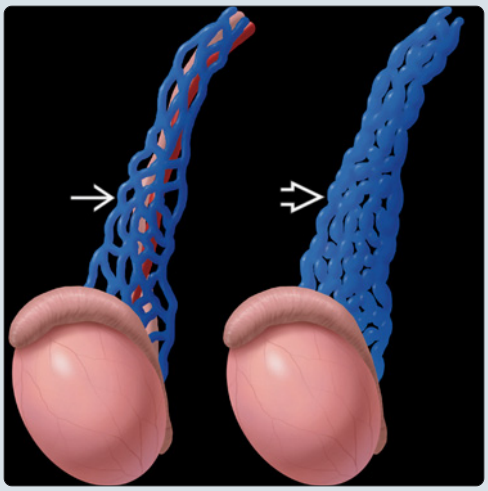
- Tubular ectasia/rete testis
- Hematocele/pyocele
- Epididymitis
- Hernia

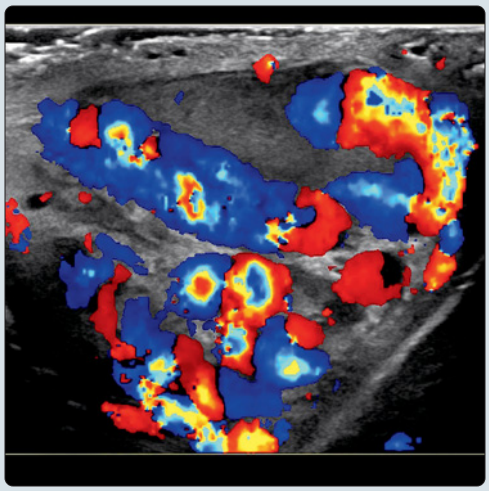
CLINICAL ISSUES

- Most common signs/symptoms
  - Vague scrotal discomfort or pressure, primarily when standing
- Majority (80%) are left sided, bilateral in 15% of patients
- Most frequent cause of male infertility
- Results from treatment to increase fertility mixed, may be less effective than previously thought

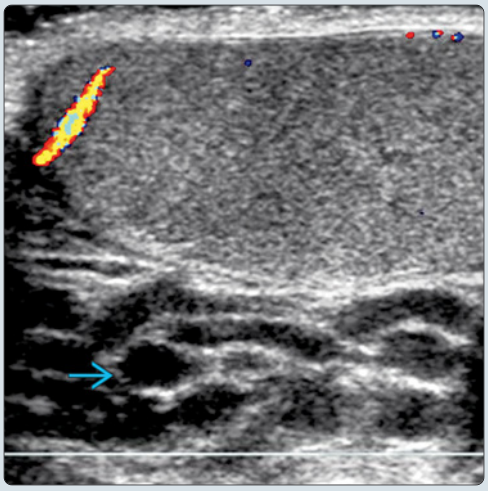
DIAGNOSTIC CHECKLIST

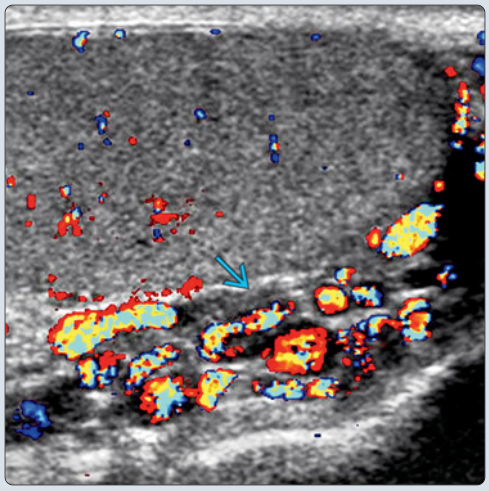
- Consider renal vein occlusion/compression by retroperitoneal tumor in elderly male
- Check retroperitoneum, particularly if isolated right-sided varicocele
- Valsalva essential for diagnosis of small varicoceles





(Left) *Graphic of the veins of the pampiniform plexus depicts the normal appearance on the left, while the depiction on the right demonstrates the abnormally dilated veins characteristic of a varicocele. (Right) Color Doppler ultrasound performed on an elderly male with scrotal discomfort shows a huge left varicocele. The late onset prompted imaging of the retroperitoneum, which demonstrated marked dilatation of the cava and renal veins due to severe right heart dysfunction.*





(Left) *Longitudinal color Doppler ultrasound without Valsalva maneuver demonstrates little flow in the dilated pampiniform veins.* **(Right)** *Longitudinal color Doppler ultrasound obtained while the patient performed the Valsalva maneuver illustrates a marked increase in flow within the varicocele, the characteristic finding in the setting of a varicocele. The visualization of flow on color Doppler US during the Valsalva maneuver is particularly beneficial in diagnosing small varicoceles.*

TERMINOLOGY

Definitions

- Dilatation of veins of pampiniform plexus > 2-3 mm diameter secondary to retrograde flow in internal spermatic vein

IMAGING

General Features

- Best diagnostic clue
  - Dilated serpiginous veins behind superior pole of testis on color Doppler US
  - Distention secondary to retrograde flow with Valsalva
- Location
  - Dilated veins in cremasteric plexus, vein of vas deferens, internal spermatic vein
- Size
  - Multiple veins in pampiniform plexus > 2-3 mm, ↑ size with Valsalva
- Morphology
  - Tortuous vascular channels (dilated veins)

Ultrasonographic Findings

- Grayscale ultrasound
  - Tubular serpiginous vessels posterior to testis
- Color Doppler
  - Prominent color flow within vessels with Valsalva secondary to retrograde flow

Imaging Recommendations

- Best imaging tool
  - US with color Doppler
- Protocol advice
  - Resting and Valsalva color Doppler images of epididymis

DIFFERENTIAL DIAGNOSIS

Tubular Ectasia/Rete Testis

- Dilated seminiferous tubules within mediastinum testis, often associated with epididymal cyst
- No flow on color Doppler (distinguish from intratesticular varicocele)

Hematocele/Pyocele

- Complex fluid collection enveloping testis
- Conforms to testis, no color Doppler flow
- Clinical history aids diagnosis (trauma, fever, scrotal pain)

Epididymitis

- ↑ hypoechoic epididymis, ↑ flow on color Doppler

Hernia

- Echogenic fat (mesentery) &/or bowel displaces testis
- May be accentuated with Valsalva, no flow at color Doppler
- Check for gut signature, bowel peristalsis

PATHOLOGY

General Features

- Etiology
  - Primary: Incompetent venous valve
    - Vast majority left sided: Due to entrance of left testicular vein into left renal vein at 90% angle (right testicular vein drains directly into IVC)
  - Secondary: Compression of testicular venous drainage
    - Retroperitoneal tumor (RCC, lymphoma), retroperitoneal fibrosis
    - Compression of left renal vein by SMA ("nutcracker syndrome")
- Associated abnormalities
  - Low sperm count, infertility
    - Elevated testicular venous pressures: Testicular congestion, diminished oxygenation, increased temperatures result in reduced sperm quality and quantity, and testicular atrophy over time
  - Secondary (reactive) varicoceles caused by retroperitoneal mass obstructing testicular veins

Staging, Grading, & Classification

- Primary: Idiopathic (incompetent valves); 98% left sided
- Secondary: Obstruction/compression of renal vein or IVC by retroperitoneal mass or fibrosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Vague scrotal discomfort or pressure, primarily when standing; infertility
- Clinical profile
  - 80% left sided; bilateral in 15% of patients
    - Isolated right-sided varicocele should raise suspicion of retroperitoneal tumor

Demographics

- Epidemiology
  - 10-15% of men in USA have varicoceles
  - Most frequent cause of male infertility
  - Subclinical varicocele in 40-75% of infertile men

Natural History & Prognosis

- Treatment increases fertility with mixed results, may be less effective than previously thought

Treatment

- Catheter embolization if symptomatic or causing low sperm count vs. surgical ligation

DIAGNOSTIC CHECKLIST

Consider

- Left renal vein, caval compression or invasion by tumor in elderly male

Image Interpretation Pearls

- Valsalva essential for diagnosis of small varicoceles

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Pyocele

KEY FACTS

TERMINOLOGY

- Scrotal abscess, complication of epididymo-orchitis

IMAGING

- Best imaging tool: Ultrasound with color Doppler
- Complex fluid collection within tunica vaginalis on ultrasound
  - Linear septations due to fibrinous strands
  - Low-level echoes from pus, white cell debris
- Color Doppler findings
  - Hyperemia of epididymis and testis 2° to epididymo-orchitis
  - Avascular complex paratesticular fluid collection
- Power Doppler findings
  - Increased flow to epididymis and testis
  - Avascular fluid and internal septations

TOP DIFFERENTIAL DIAGNOSES

- Hematocele
- Spermatocele
- Hydrocele
- Epididymal cyst
- Fournier gangrene

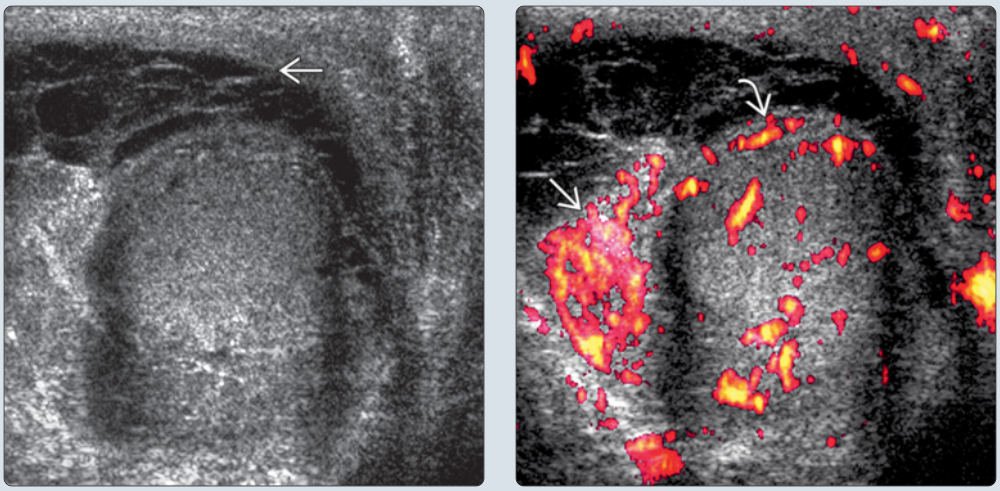
CLINICAL ISSUES

- Most common signs/symptoms: Acute scrotal pain
- Other signs/symptoms: Fever, dysuria, scrotal skin thickening
- Most respond to antibiotic therapy
  - Empiric antibiotic choice based upon patient age
    - Prepubescent, middle-aged males: Coliform bacteria
    - Patients 14-35 years of age: Gonococcus/chlamydia

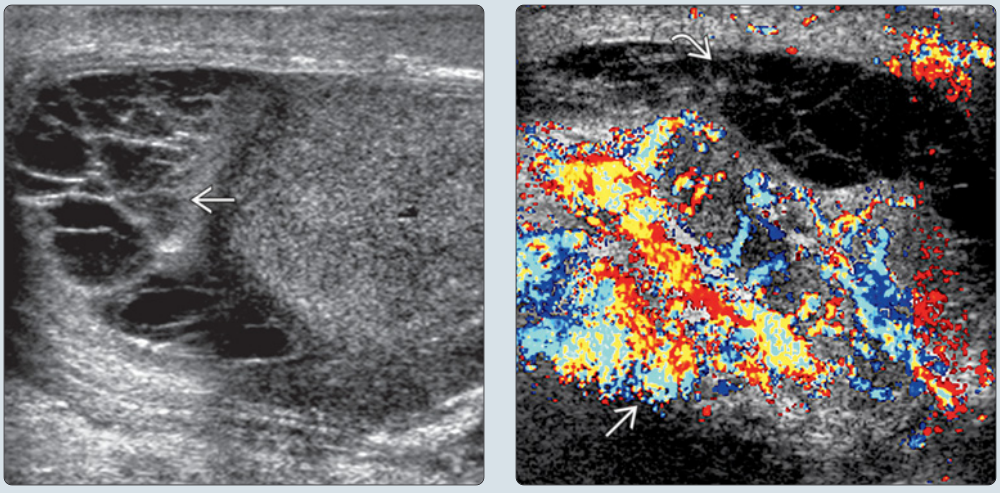
DIAGNOSTIC CHECKLIST

- Consider repeat ultrasound if no clinical improvement with empiric antibiotic therapy
- Assess for complications (testicular abscess, infarction) or atypical infection (mycobacterium)

(Left) *Sagittal grayscale ultrasound of the scrotum in a 19-year-old male presenting with fever and acute scrotal pain demonstrates linear strands of pus within the tunica vaginalis, consistent with a pyocele.* **(Right)** *Sagittal power Doppler ultrasound in the same patient reveals epididymal and testicular hyperemia (hallmarks of epididymo-orchitis).*



(Left) *Sagittal grayscale ultrasound in a 37-year-old man presenting with acute scrotal pain, fever, and dysuria demonstrates a large pyocele with numerous septations.* **(Right)** *Sagittal color Doppler ultrasound of epididymis in the same patient illustrates hyperemia from epididymitis, as well as the adjacent pyocele.*



TERMINOLOGY

Definitions

- Scrotal abscess, complication of epididymo-orchitis

IMAGING

General Features

- Best diagnostic clue
  - Complex fluid collection containing linear strands and low-level echoes within tunica vaginalis
- Associated with epididymo-orchitis

Imaging Recommendations

- Best imaging tool
  - Ultrasound with color Doppler

Ultrasonographic Findings

- Grayscale ultrasound
  - Complex fluid collection within tunica vaginalis
    - Linear septations due to fibrinous strands
    - Low-level echoes from pus, white-cell debris
- Color Doppler
  - Hyperemia of epididymis and testis secondary to epididymo-orchitis
  - Avascular complex fluid collection
- Power Doppler
  - Increased flow to epididymis and testis
  - Avascular fluid and internal septations

DIFFERENTIAL DIAGNOSIS

Hematocele

- History of trauma
- Heterogeneity of testis due to fracture
- Complex fluid collection comprised of clot, blood, and fibrin strands within tunica vaginalis

Spermatocele

- Often asymptomatic scrotal mass
- Thin-walled complex cyst within epididymal head
  - Cyst comprised of spermatozoa, fat sediment, and cellular debris
- "Streaming" of particulate debris on US: "Falling snow" sign

Hydrocele

- Simple fluid collection within tunica vaginalis
- Most patients asymptomatic

Epididymal Cyst

- Simple cyst usually located in head of epididymis
- Most patients asymptomatic

Fournier Gangrene

- Gas within scrotal wall, perineum
- Reactive, complex hydrocele

PATHOLOGY

General Features

- Etiology
  - Epididymo-orchitis
  - Spontaneous rupture of testicular abscess
- Associated abnormalities
  - Epididymitis, orchitis
  - Testicular or epididymal abscess if inadequately treated

Gross Pathologic & Surgical Features

- Thick-walled fluid collection
  - Pus and cellular debris

Microscopic Features

- Bacteria, WBC debris

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute scrotal pain
- Other signs/symptoms
  - Fever, dysuria, scrotal skin thickening

Demographics

- Age
  - 0-50 years
- Gender
  - Male

Natural History & Prognosis

- Most respond to antibiotic therapy
  - Empiric therapy typically employed based upon likely pathogens
    - Patients 14-35 years of age: Gonococcus/chlamydia
    - Patients < 14 or > 35 years of age: Enteric organisms (coliform bacteria)
    - Repeat ultrasound employed if no clinical improvement: Assess for complications (testicular abscess, infarction) or atypical infection (mycobacterium)
- Rarely drainage for refractory cases

Treatment

- Antibiotics
- Surgery

DIAGNOSTIC CHECKLIST

Consider

- Repeat ultrasound if no improvement with empiric antibiotic therapy
  - Assess for complications (testicular abscess, testicular infarction) or atypical infection (mycobacterium)

Image Interpretation Pearls

- Complex fluid collection with internal septations on ultrasound

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Hernia

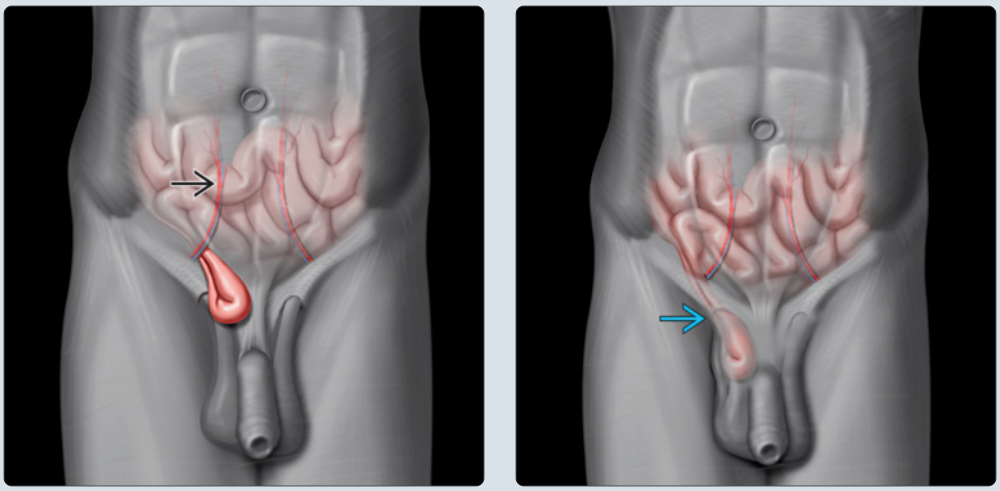
KEY FACTS

TERMINOLOGY

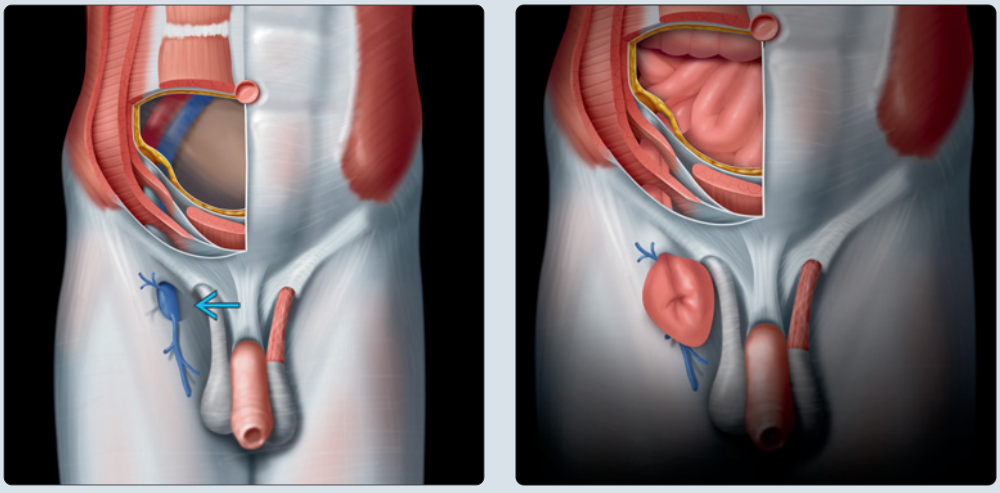
- **Incarcerated (irreducible) hernia**: Entrapment of hernia content within hernia sac
- **Strangulated hernia**: Compromised blood flow to hernia content resulting in ischemia/necrosis

IMAGING

- **Direct inguinal hernia**
  - Acquired hernia caused by weakness of fascia transversalis
  - Hernia sac emerges **superomedial** to origin of inferior epigastric vessels and protrudes through Hesselbach triangle
  - **Lateral crescent sign**: Compression, flattening, and lateral displacement of inguinal canal content
- **Indirect inguinal hernia**
  - Most common type of groin hernia; more common in male
  - Hernia sac protrudes lateral to Hesselbach triangle, **superolateral** to course of inferior epigastric vessels
  - Inguinal canal is expanded and its content might not be well visualized
- **Femoral hernia**
  - Due to narrow neck, it has high rate of complications (obstruction, strangulation)
  - Femoral hernia neck is **inferior** to inguinal ligament and inferior epigastric vessels
  - Hernia sac runs craniocaudal course and is closely associated with femoral vein & causing flattening of vein
- **Obturator hernia**
  - Rare; usually seen in older females
  - Hernia sac extends through obturator canal and commonly protrudes between obturator externus (posteriorly) and pectineus muscle (anteriorly)
  - High risk of obstruction; ~ 90% of patients present with bowel obstruction



(Left) *Graphic shows a direct inguinal hernia protruding through the roof of the inguinal canal. Note the location of the hernia sac above the inguinal ligament and medial to the inferior epigastric vessels. Direct inguinal hernia results in flattening of inguinal canal.* **(Right)** *Graphic of indirect inguinal hernia shows hernia sac extending into the inguinal canal and exiting through the superficial inguinal ring into the scrotum.*



(Left) *Graphic shows normal anatomy of femoral canal, which lies medial to the femoral vein and inferior to the inguinal ligament. The femoral canal normally contains fat and lymphatics.* **(Right)** *Graphic of femoral hernia shows close association between the hernia sac and femoral vessels, commonly resulting in compression and concavity of the vein. The femoral hernia sac originates below the inguinal ligament and below the origin of the inferior epigastric artery.*

TERMINOLOGY

Definitions

- Protrusion of organ (or tissue) outside its physiologic cavity
- **Incarcerated (irreducible) hernia**: Entrapment of hernia content within hernia sac
- **Strangulated hernia**: Compromised blood flow to hernia content resulting in ischemia/necrosis

IMAGING

Relevant Anatomy

- **Inguinal canal**
  - Narrow, oblique canal extending from internal (deep) inguinal ring to external (superficial) ring
  - Anterior wall: Aponeuroses of external and internal oblique muscles
  - Floor: Inguinal ligament (a.k.a. ligament of Poupart; thickened and folded inferior portion of external oblique aponeurosis extending from anterior superior iliac spine to pubic tubercle)
  - Posterior wall: Conjoint tendon (internal oblique and transversalis fascia)
  - Roof: Aponeurosis of internal oblique and transversus abdominis muscles
  - Deep inguinal ring: Round opening through transversalis fascia; superior to inguinal ligament and lateral to inferior epigastric vessels
  - Superficial inguinal ring: V-shaped opening through external oblique aponeurosis; medial and superior to pubic tubercle
  - Contents: Spermatic cord (in male); round ligament (in female); ilioinguinal nerve
- **Femoral canal**
  - Medial-most compartment of femoral sheath containing lymphatics and fat
  - Anterosuperior border: Inguinal ligament
  - Posterior border: Pectineal ligament
  - Medial border: Lacunar ligament
  - Lateral border: Femoral vein
- **Obturator canal**
  - Uncovered superior aspect of obturator foramen connecting pelvis and adductor compartment of thigh
  - Contains obturator vessels and obturator nerve

Direct Inguinal Hernia

- Acquired hernia caused by weakness of fascia transversalis
- More common in men; less risk of incarceration/strangulation
- Hernia sac protrudes through Hesselbach triangle
  - Borders of Hesselbach triangle are inguinal canal (inferiorly), inferior epigastric vessels (laterally), and lateral edge of rectus abdominis muscle (medially)
- Hernia sac emerges **superomedial** to origin of inferior epigastric vessels
- Hernia sac is anterior to pubic tubercle; causes compression and flattening of inguinal canal content
  - **Lateral crescent sign**: Compression, flattening, and lateral displacement of inguinal canal content
- Small hernias may reduce when patient in supine position and are thus missed on CT
  - Some authors have advocated performing CT with Valsalva maneuver to increase sensitivity for detection of hernias

Indirect Inguinal Hernia

- Most common type of groin hernia; more common in male
- Congenital (patent processus vaginalis) or acquired (weakness of internal inguinal ring)
- Hernia sac protrudes lateral to Hesselbach triangle, **superolateral** to course of inferior epigastric vessels
  - Inguinal canal is expanded and its content might not be well visualized

Femoral Hernia

- Less common than inguinal hernia (~ 2-4% of groin hernia); 4x more common in females
- Due to narrow neck, has high rate of complications (obstruction, strangulation)
- Femoral hernia neck is **inferior** to inguinal ligament and inferior epigastric vessels
- Hernia sac runs craniocaudal course and is closely associated with femoral vein & causing flattening of vein
  - Hernia sac is posterior to pubic tubercles

Obturator Hernia

- Rare; usually seen in older females
- Hernia sac extends through obturator canal and commonly protrudes between obturator externus (posteriorly) and pectineus muscle (anteriorly)
- High risk of obstruction; ~ 90% of patients present with bowel obstruction
- CT: Hernia sac within obturator canal and extending between obturator externus and pectineus muscles; associated bowel obstruction is commonly seen
- Treatment: Sac resection and surgical repair of defect with sutures or prosthetic mesh; can be done via transabdominal or extraabdominal approaches

Perineal Hernia

- Rare; usually seen in elderly females: Due to pelvic floor weakness
- Posterior type: Hernia sac protrudes through defect in levator ani muscle or between levator and coccygeus muscles
- Anterior type: Hernia sac protrudes through urogenital diaphragm

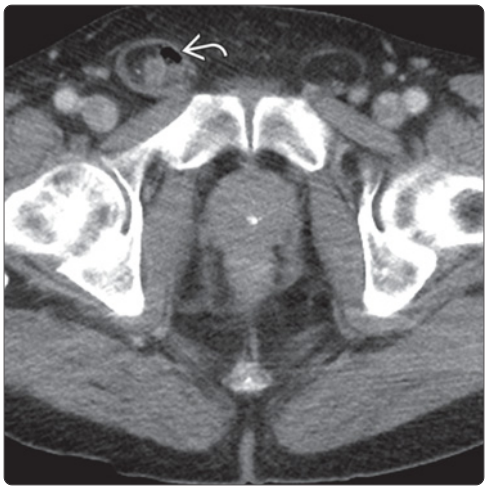
Imaging Recommendations

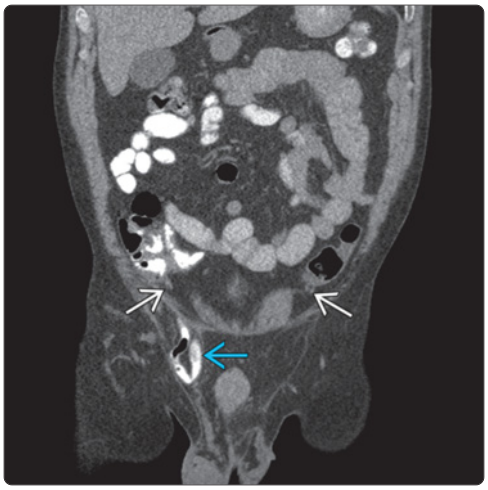
- Best imaging tool
  - CT is modality of choice for evaluation of hernias and their complications
  - US has wide sensitivity range (30-100%) with advantage of real-time scanning (with patient in upright position or during Valsalva)
- Protocol advice
  - Some authors suggest CT with Valsalva maneuver to increase sensitivity for detection of hernias

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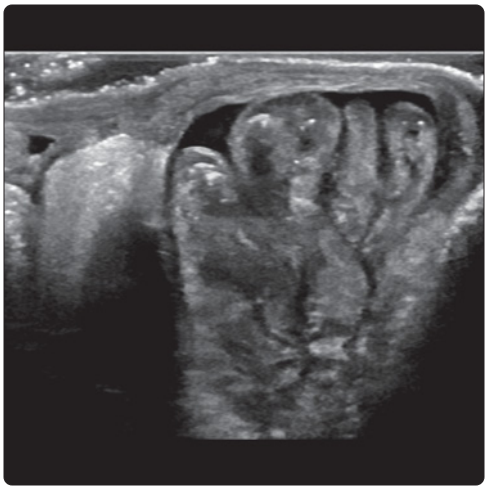
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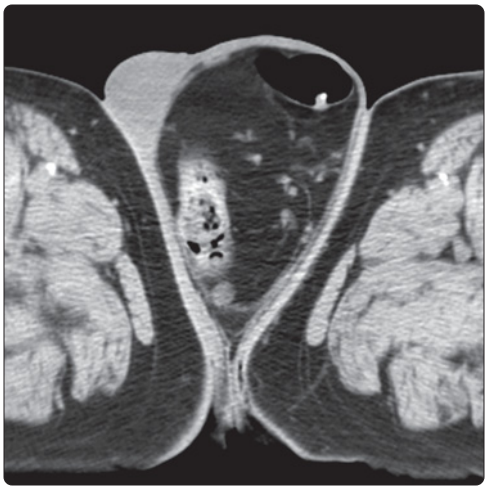
(Left) *Axial CECT in this elderly man shows bilateral indirect inguinal hernias with the right hernia sac containing the bowel* ➡. **(Right)** *Coronal NECT in the same patient shows that the hernia sac contains the appendix* ⇨. *This type of hernia is termed an Amyand hernia. The hernia sac originated lateral to the inferior epigastric vessels* ➡.



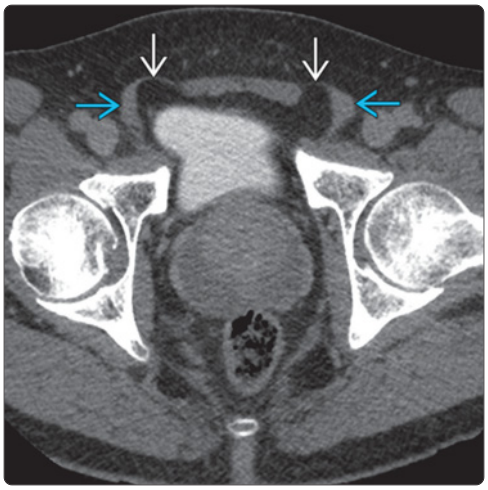


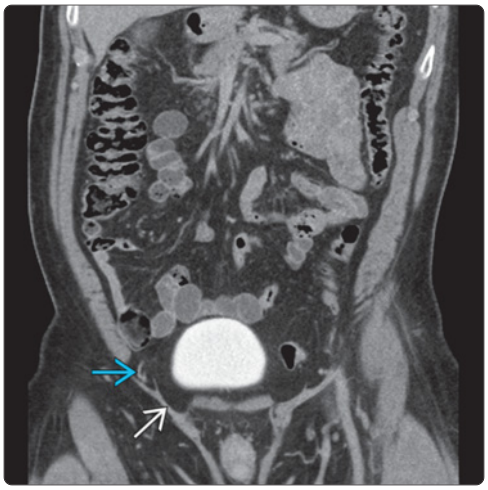
(Left) *Grayscale US in a patient with history of scrotal mass shows multiple bowel loops and free fluid within the left hemiscrotum. Real-time scan also showed peristalsis.* **(Right)** *Axial NECT in the same patient confirms large scrotal hernia containing bowel loops and mesenteric fat.*

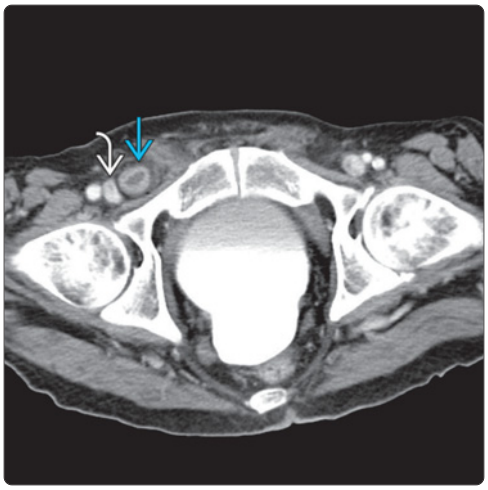


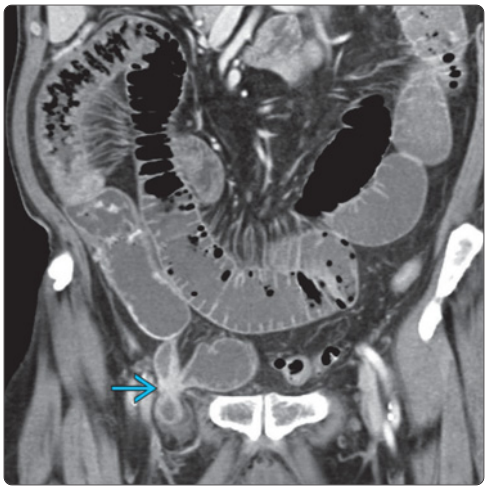


(Left) *Axial CECT shows bilateral direct inguinal hernia* ➡ *containing part of the bladder and perivesical fat. Note the lateral crescent sign* ⇨, *which represents flattened inguinal canal.* **(Right)** *Coronal CECT in the same patient shows that the hernia sac* ➡ *originates medial to the inferior epigastric vessels* ⇨.

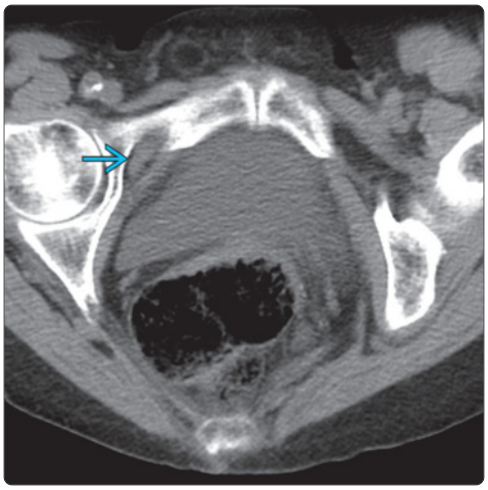


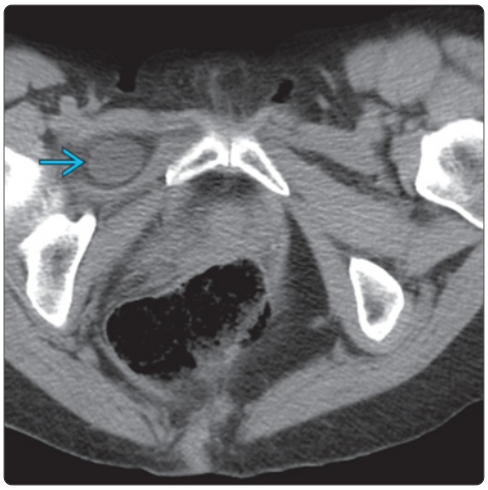




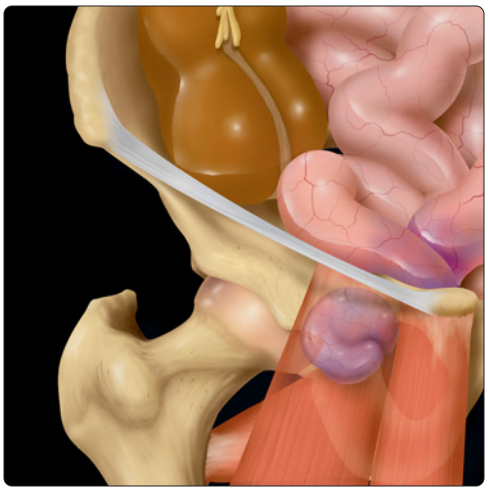


(Left) *Axial CECT shows a loop of thickened, hyperemic bowel herniating into the femoral canal, medial to the femoral vein. Notice that the femoral vein is compressed. The herniated bowel lies posterolateral to the pubic tubercle. (From DI3: GI.)* **(Right)** *Coronal CECT in the same patient shows multiple dilated small bowel loops with a transition point at the hernia neck. This thickened, hyperenhancing bowel within the hernia sac was found to be ischemic at surgery. (From DI3: GI.)*





(Left) *Axial NECT in this elderly female shows bowel herniation through the obturator canal. The hernia sac lies lateral to the obturator internus muscle.* **(Right)** *Axial NECT through a more inferior section in the same patient shows caudal extension of hernia into the adductor space between the pectineus muscle (anteriorly) and obturator externus muscle (posteriorly).*





(Left) *Graphic shows location of obturator hernia deep to the pectineus muscle.* **(Right)** *Axial NECT in this elderly woman shows a defect in the left posterolateral aspect of levator ani muscle with herniation of bowel and mesenteric fat into left ischioanal fossa. Perineal hernias are extremely rare and are usually seen in elderly females.*

Fournier Gangrene

KEY FACTS

TERMINOLOGY

- Life-threatening, polymicrobial, necrotizing soft tissue infection of perineal, perianal, or genital regions

IMAGING

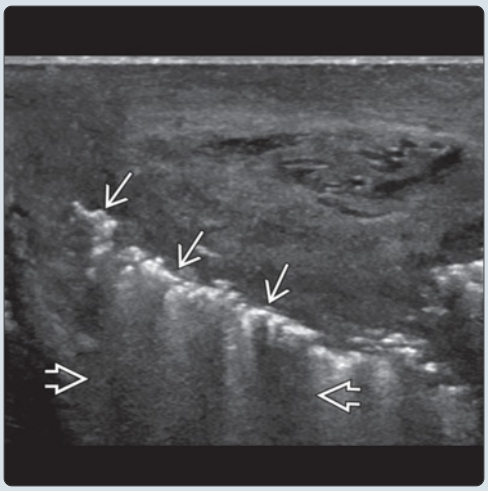
- NECT: Most useful modality for depicting extent of soft tissue infection and gas in emergent setting
  - Fascial thickening, subcutaneous fat stranding, subcutaneous gas correlates with extent of affected tissue at debridement
  - Infection spreads along superficial pelvic fascial planes to scrotum, anterior abdominal wall, buttocks
  - Untreated, overwhelming infection may extend into deep pelvic extraperitoneal space
- Ultrasound may be employed as initial test for males with acute scrotal pain and early (subclinical) manifestations of Fournier gangrene
  - Scrotal wall edema
  - Scrotal wall gas: Echogenic foci with posterior "dirty" shadowing

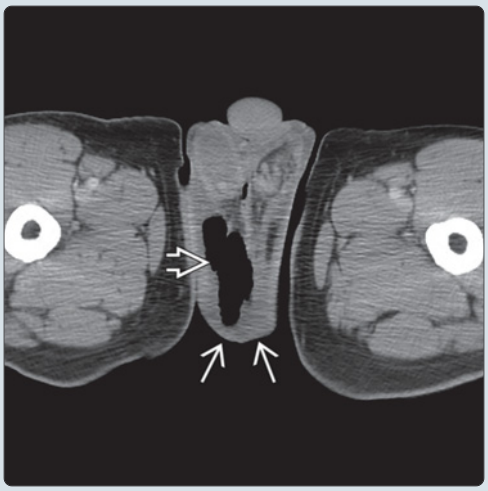
TOP DIFFERENTIAL DIAGNOSES

- Scrotal edema
  - Reflection of 3rd spacing: Local, systemic manifestations of inflammation absent
- Scrotal cellulitis
  - Less aggressive, localized soft tissue infection
  - Subcutaneous gas absent (though absence does not entirely exclude Fournier gangrene)
- Perianal fistula/abscess

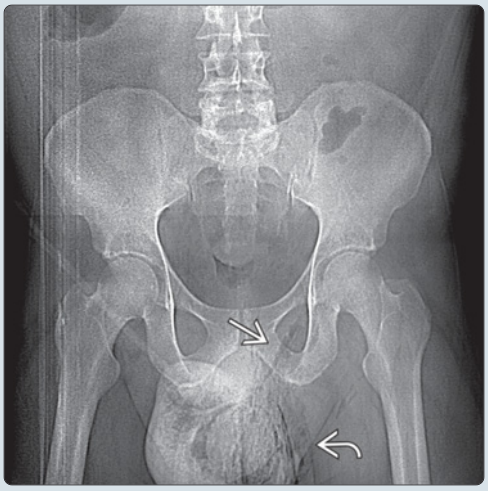
CLINICAL ISSUES

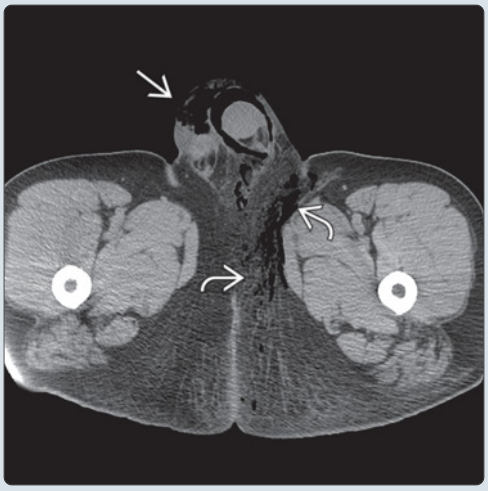
- Predisposing factors: Impaired host immunity and local tissue hypoxia (diabetes)
- High mortality rate even with aggressive, rapid treatment: Sepsis, multisystem organ failure
- CT guides extent of often repeated debridements





(Left) *Transverse ultrasound performed on a 54-year-old diabetic man with a 2-day history of scrotal pain and erythema shows posterior scrotal wall echogenicity and "dirty" posterior shadowing, a characteristic appearance of soft tissue gas.* **(Right)** *Axial CECT of the same patient shows scrotal wall edema and extensive gas. Fournier gangrene is a surgical emergency. This patient was successfully treated with broad spectrum antibiotics, partial scrotectomy, and ultimately flap reconstruction.*





(Left) *Digital scout image of a 43-year-old man with scrotal pain, erythema, and leukocytosis shows scrotal and inguinal subcutaneous emphysema.* **(Right)** *Axial NECT performed on the same patient shows scrotal wall and left medial buttock subcutaneous gas. Serial NECT studies guided the extent of multiple debridements. Predisposing causes of Fournier gangrene include impaired immunity and local tissue hypoxia. The inciting event for overwhelming infection may be minor local trauma.*

TERMINOLOGY

Definitions

- Rapidly progressing, life-threatening, fulminant, necrotizing polymicrobial fasciitis of perineal, genital, and perianal regions

IMAGING

General Features

- Best diagnostic clue
  - Fascial thickening, subcutaneous infiltration, fluid collections
  - Imaging hallmark: Subcutaneous emphysema
- Location
  - Perineal, perianal, genital regions
  - May extend into pelvic extraperitoneal space, anterior abdominal wall

Radiographic Findings

- Radiography
  - Perineal, scrotal soft tissue swelling and gas
  - Soft tissue gas may be identified before crepitus detected at physical examination

CT Findings

- NECT
  - Modality of choice for depicting location of soft tissue infection/gas
  - Fascial thickening, fat stranding, subcutaneous gas correlates with extent of affected tissue at debridement
  - Infection spreads along superficial pelvic fascial planes to scrotum, anterior abdominal wall, buttocks
  - Untreated Fournier gangrene may progress to pelvic extraperitoneal (perivesical, posterior interfascial) space
- CECT
  - Suggested as modality for identifying frank tissue necrosis but typically not employed in emergent setting, particularly if multiorgan system failure

Ultrasonographic Findings

- Scrotal wall thickening, edema
- Scrotal wall gas: Hyperechoic foci with posterior "dirty" shadowing
- Reactive hydrocele, normal testes/epididymis
- Blood flow to testes preserved (different blood supply than scrotum)

Imaging Recommendations

- Best imaging tool
  - NECT: Readily available and best shows extent of soft tissue infection in emergent setting

DIFFERENTIAL DIAGNOSIS

Scrotal Edema

- Generalized 3rd spacing (liver, heart, renal failure)
- Absent local/systemic manifestations of fulminant soft tissue infection

Scrotal Cellulitis

- Less severe, localized soft tissue infection
- Absent scrotal wall emphysema does not entirely exclude Fournier gangrene

Perianal Fistula

- May be inciting event for Fournier gangrene

PATHOLOGY

General Features

- Fulminant, polymicrobial, necrotizing soft tissue infection of perineum
  - Common organisms: *Escherichia coli*, *Bacteroides*, streptococcal species, *Staphylococcus* (common perineal flora)
- Predisposing conditions: Impaired host immunity &/or local tissue hypoxia
  - Diabetes most commonly associated comorbid condition
- Initial event: Local trauma or extension of urinary tract/perianal infection

Staging, Grading, & Classification

- Fournier gangrene severity index utilized to assess disease severity

Microscopic Features

- Bacterial infection → vascular thrombosis with soft tissue (dermal, subcutaneous, fascial) necrosis, polymorphonuclear cell infiltration

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Tenderness, pain, swelling, erythema at effected site: Pain out of proportion to physical findings
  - Crepitus, foul discharge
- Other signs/symptoms
  - Fever, leukocytosis, sepsis, multisystem organ failure

Demographics

- M >> F, mean age: 51 years

Natural History & Prognosis

- Fulminant necrotizing infection with mortality rates up to 34%

Treatment

- Fluid resuscitation, antimicrobial therapy, repeated surgical debridements, organ support, hyperbaric oxygen

DIAGNOSTIC CHECKLIST

Image Interpretation Pearls

- Hallmark of potentially fatal infection: Perineal, perianal, scrotal infiltration, and subcutaneous gas at CECT

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KEY FACTS

IMAGING

- Heterogeneous testicular parenchyma or contour abnormality of testis in setting of scrotal trauma
  - Disruption of tunica albuginea is very specific for testicular rupture
- Extratesticular hematocele: Most common finding in scrotum after blunt injury
- US appearance of hematoma depends on time elapsed since trauma
- Intra- or extratesticular gas in scrotum, missile track &/or foreign bodies (bullet/pellet) is indicative of penetrating injury
- Spermatic cord hematoma appears as heterogeneous avascular mass superior to testis
- Color Doppler is useful to determine viable portions of testis

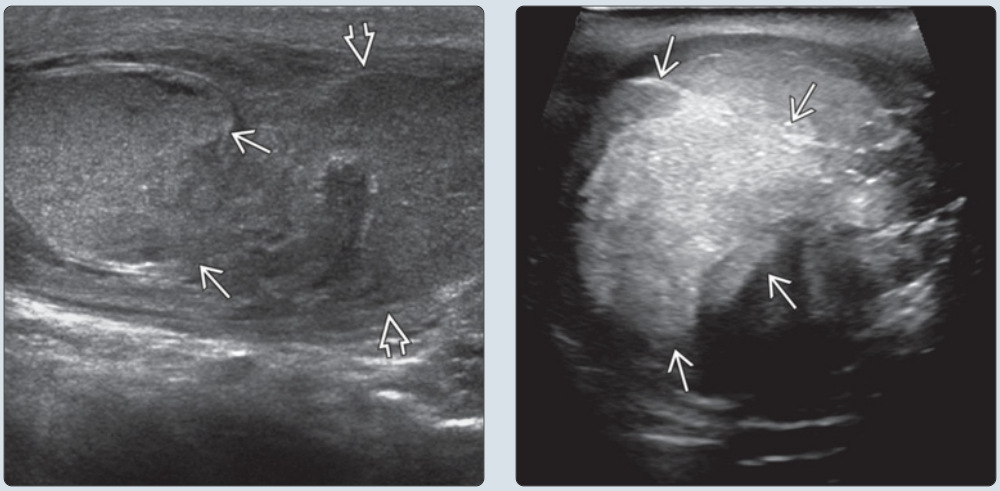
PATHOLOGY

- Sports injuries (> 50%), vehicular and ballistic trauma, and iatrogenic

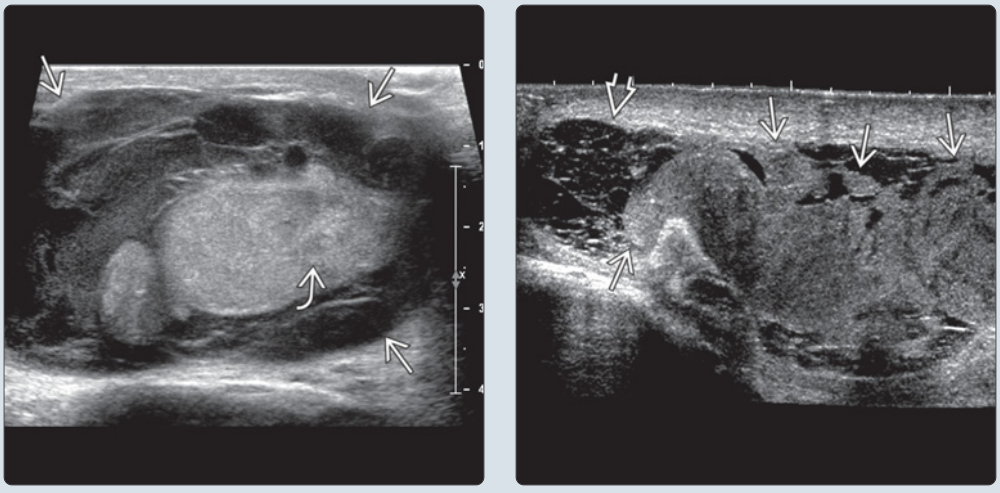
CLINICAL ISSUES

- Unless repaired within 72 hours, salvage rate only 45%
- Follow-up of conservatively treated testicular hematomas is essential due to increased risk of infection, which may obviate orchiectomy
- Orchiectomy (total or partial) is performed for nonviable testis
- Surgical exploration and drainage must be performed for large intratesticular hematomas

(Left) *Scrotal US performed on a 23-year-old man post-MVA shows widely disrupted lower pole tunica ➡ and adjacent hypoechogenicity ➡ (a combination of extruded seminiferous tubules and acute clot). Flow within the upper pole (not shown) suggested residual viable testis and emergent partial orchiectomy was performed.* **(Right)** *Transverse ultrasound shows a mid testicular fracture ➡ post blunt trauma. The testis was entirely necrotic despite emergent exploration, and an orchiectomy was performed.*



(Left) *Ultrasound post trauma shows a complex extratesticular fluid collection ➡ with an appearance consistent with a hematocele. Mild testicular heterogeneity ➡ also suggests contusion/hematoma.* **(Right)** *Ultrasound performed 1 week post MVA shows a deformed heterogenous testis, extruded parenchyma ➡, and a hematocele ➡. The combination of testicular heterogeneity and tunica disruption is both sensitive and specific for rupture. The testis was not viable at exploration.*



TERMINOLOGY

Definitions

- Rupture of tunica albuginea, extrusion of testicular tissue into scrotal sac, collection of blood in tunica vaginalis or scrotal wall

IMAGING

General Features

- Best diagnostic clue
  - Heterogeneous testis or contour abnormality of testis and history of scrotal trauma
- Morphology
  - Irregularity of testicular contour and focal or diffuse testicular heterogeneity &/or epididymis
  - Extratesticular hematocele: Most common finding in scrotum after blunt injury

Ultrasonographic Findings

- Grayscale ultrasound
  - Hematocele: Hemorrhage contained within layers of tunica vaginalis
  - Testicular rupture
    - Disruption of tunica albuginea
    - Contour abnormality of testis due to testicular extrusion
    - Heterogeneous testicular echotexture and echogenicity
  - Testicular fracture
    - Discrete linear or irregular fracture plane within testis (17%)
  - Testicular dislocation
    - Unilateral > > bilateral
    - Most commonly from high-impact injury against fuel tank in motorcycle accidents
  - Testicular torsion
    - Trauma-induced torsion in 5-8% cases
  - Testicular hematoma
    - US appearance depends on time interval post trauma
  - Penetrating trauma
    - Intra- or extratesticular gas in scrotum, missile track &/or foreign bodies (bullet/pellet)
  - Epididymal injury: Focal epididymal enlargement with ↓ echogenicity & abnormal position relative to testis
  - Scrotal wall hematoma: Focal wall thickening or loculated collection within wall
  - Spermatic cord hematoma: Heterogeneous avascular mass superior to testis
- Color Doppler
  - Distorted intratesticular vascularity with interruption of vessels in area of injury due to injury to tunica vasculosa
    - Useful to determine viable portions of testis

Imaging Recommendations

- Best imaging tool
  - High-resolution US (≥ 7.5 MHz)

DIFFERENTIAL DIAGNOSIS

Testicular Torsion

- ↓ overall vascularity compared to normal testis

Epididymo-Orchitis

- Acute or chronic pain without history of trauma, enlarged hypoechoic epididymis with increased vascularity in epididymis and testis on color Doppler

Testicular Abscess

- Focal ill-defined, thick-walled mass and necrotic center

PATHOLOGY

General Features

- Etiology
  - Sports injuries (> 50%), vehicular and ballistic trauma, and iatrogenic
  - Blunt trauma impales scrotal contents to symphysis pubis or pubic rami, pelvic fracture
  - Penetrating trauma includes sharp objects such as knives, bullets, animal bites, self mutilation; gunshot injuries most common
  - Iatrogenic injuries from inguinal herniorrhaphy and orchiectomy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute scrotal hematoma following blunt trauma

Natural History & Prognosis

- Unless repaired within 72 hours, salvage rate only 45%
- Follow-up of conservatively treated testicular hematomas is essential due to increased risk of infection, which may ultimately obviate orchiectomy

Treatment

- Drainage: Large hematocele
- Orchiectomy (total or partial): Nonviable testis in testicular rupture
- Surgical exploration and drainage: Large intratesticular hematomas

DIAGNOSTIC CHECKLIST

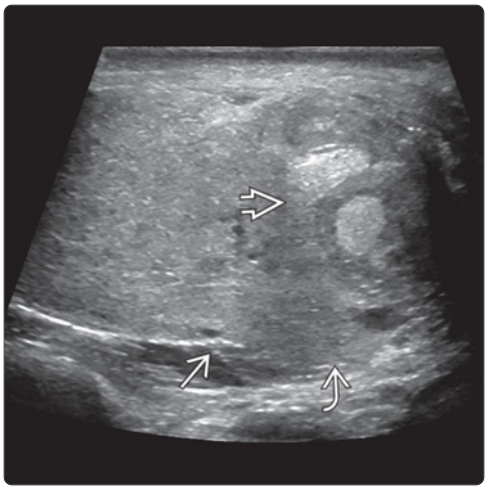
Image Interpretation Pearls

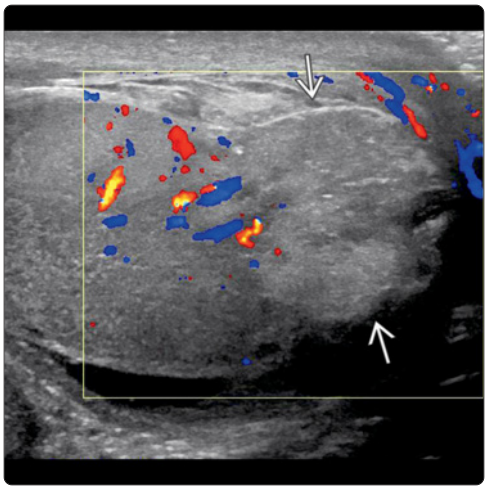
- Irregularity of testicular contour, heterogeneous parenchyma and echogenic collection in tunica vaginalis

SELECTED REFERENCES

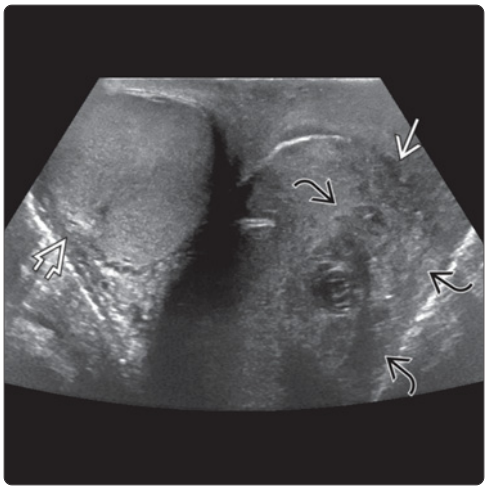
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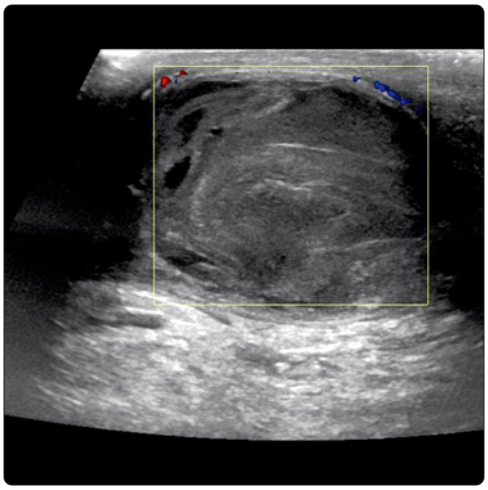
(Left) *Ultrasound of the testis after a lacrosse ball injury demonstrates a disrupted tunica albuginea, testicular heterogeneity, and a contour abnormality, a constellation of findings sensitive and specific for testicular rupture.* **(Right)** *Corresponding color Doppler ultrasound in the same patient shows avascular, nonviable extruded testicular parenchyma. Salvage of perfused viable testis may be possible immediately post injury.*



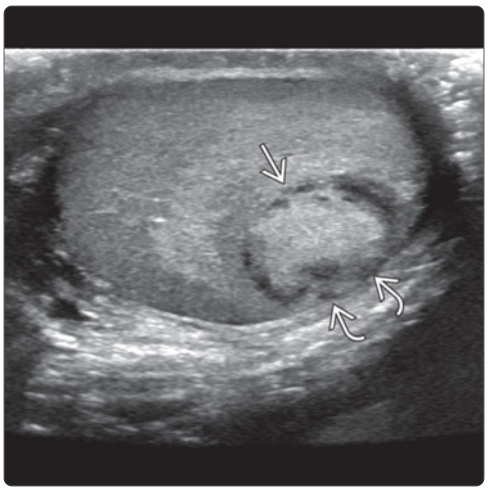


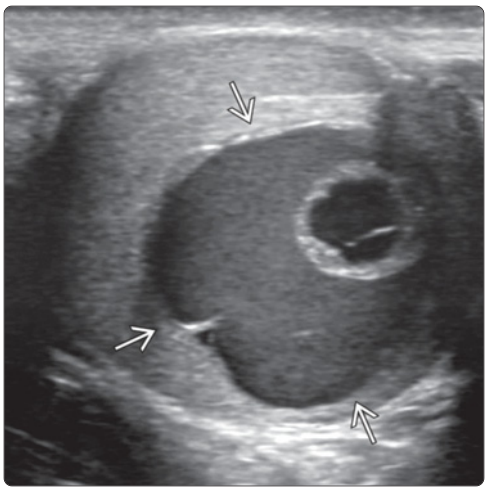
(Left) *Ultrasound post trauma shows a normal right testis, interrupted left tunica, and amorphous testicular/paratesticular echogenicity, reflecting testicular contusion, extruded seminiferous tubules, and adjacent acute clot.* **(Right)** *Color Doppler ultrasound of the scrotal cavity in the same patient with testicular rupture demonstrates a complex extratesticular fluid collection, consistent with an associated hematocele.*

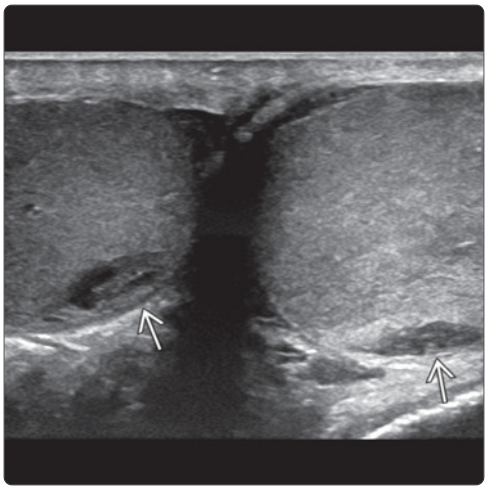


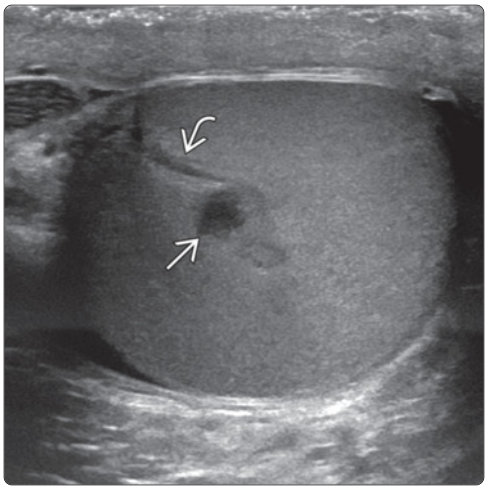


(Left) *Ultrasound performed post blunt trauma shows a well-defined intraparenchymal lesion and intact adjacent tunica. Resolution of the "lesion" at follow-up ultrasound confirmed the impression of a contained testicular hematoma.* **(Right)** *Ultrasound performed on a patient with a history of scrotal trauma and 3-week history of scrotal pain shows a complex cystic testicular mass. Lack of change at short-term surveillance imaging prompted orchiectomy for what was confirmed to be a mixed germ cell tumor.*

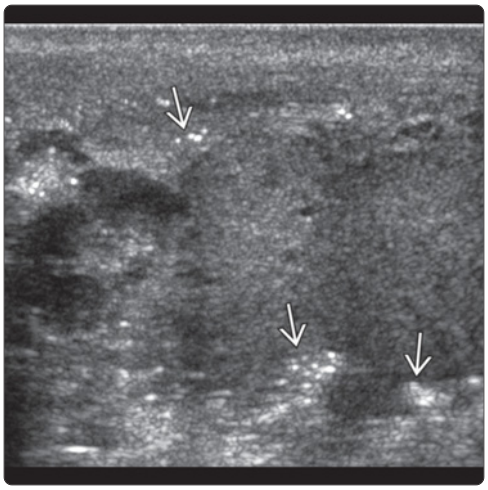


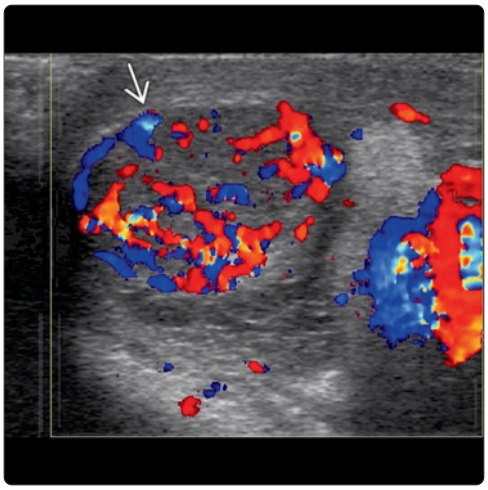




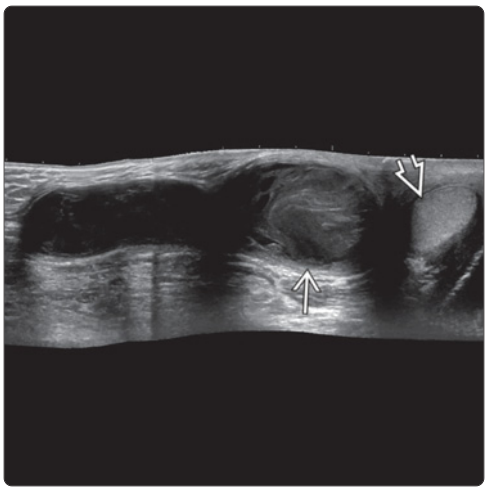


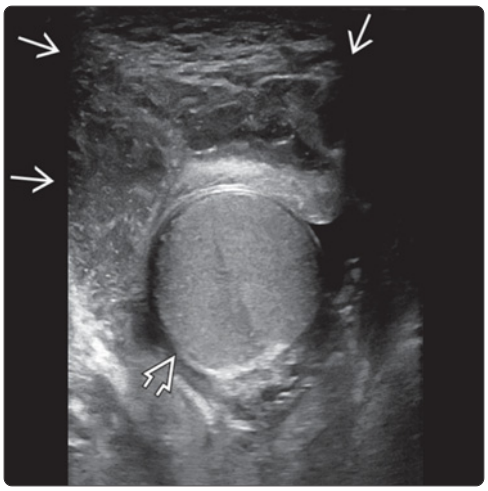
(Left) *Scrotal ultrasound performed on a 31-year-old man with scrotal pain post MVA shows small, peripheral, lentiform testicular hematomas. The patient was treated conservatively and the hematomas resolved at follow-up sonography.* **(Right)** *Ultrasound performed on a male with azoospermia and scrotal pain 1 week post thin needle testicular sperm extraction (TESE) shows a small intratesticular hematoma. Note adjacent transmediastinal artery.*





(Left) *Ultrasound of the testis in a patient with gunshot injury to the thigh shows multiple echogenic foci in the scrotum suggestive of gas. Surgery confirmed an associated testicular rupture.* **(Right)** *Color Doppler ultrasound after blunt injury to the scrotum shows an enlarged, heterogeneous hyperemic epididymis. The constellation of findings suggests traumatic epididymitis.*



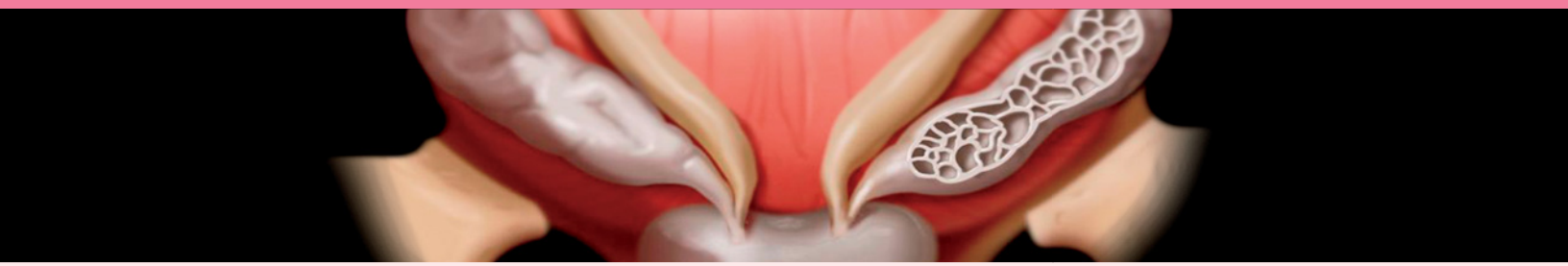


(Left) *Sagittal panoramic ultrasound of the right scrotum after inguinal herniorrhaphy shows a large, complex fluid collection extending from the superior pole of the testis into the groin. Surgery confirmed a spermatic cord hematoma.* **(Right)** *Ultrasound performed on a patient with multiple pelvic fractures and extensive extraperitoneal hemorrhage shows an intact testis and a large scrotal wall hematoma.*

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SECTION 11

Seminal Vesicles



Congenital Lesions **520**

Acquired Seminal Vesicle Lesions **522**

KEY FACTS

TERMINOLOGY

- SV: Seminal vesicle

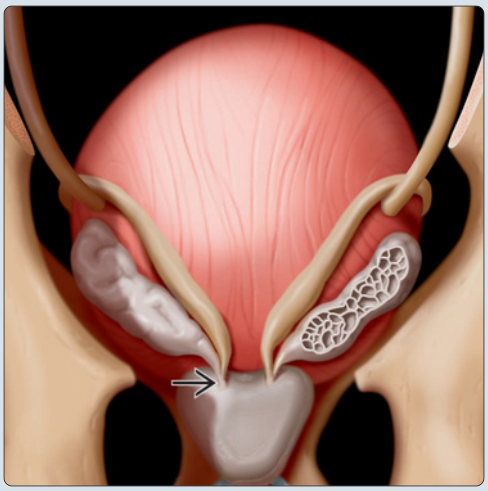
IMAGING

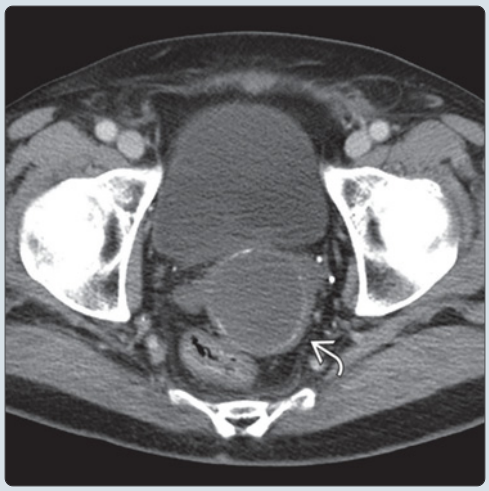
- **Embryology**
  - Development of SV, vas deferens, and urinary system closely associated to each other
  - **Mesonephric (Wolffian) duct** gives rise to SV and vas deferens
- **SV agenesis/hypogenesis**
  - Absent, or markedly small, SV
  - Agenesis more common than hypogenesis
  - Usually unilateral
  - Due to interruption of normal SV development (occurs before 7th week of gestation)
  - High association with renal and vas deferens anomalies
  - Bilateral SV agenesis strongly associated with *CFTR* gene mutation (64-73%)
    - Considered a genital form of cystic fibrosis
- **Congenital SV cyst**
  - Can be isolated or associated with other genitourinary anomalies
  - Bilateral SV cysts have high association with autosomal dominant polycystic kidney disease (44-60%)
  - SV cysts usually < 5 cm
  - MR: ↑ T2, variable T1 signal
    - SV cyst may show hight T1 signal due to proteinaceous or hemorrhagic cyst content

TOP DIFFERENTIAL DIAGNOSES

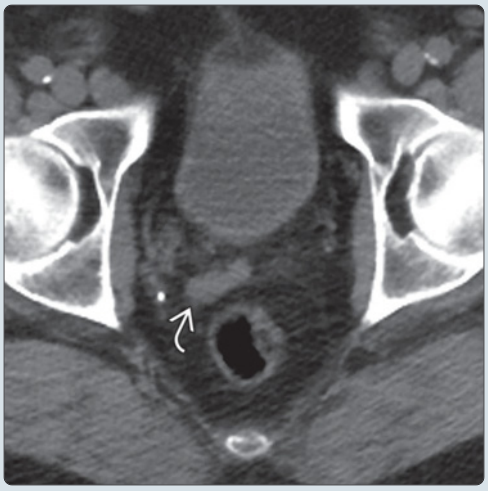
- **Acquired seminal vesicle cysts**
  - Any obstructive process in SV, ejaculatory duct, or vas deferens can result in acquired SV cyst
- **Müllerian duct cyst**
  - Usually in midline prostate
- **Cyst-like intrapelvic structures (mimics)**
  - Ureterocele, dilated ectopic ureter, bladder diverticulum, urethral diverticulum

(Left) *Coronal posterior graphic of the bladder shows the relationship between the seminal vesicles, vas deferens, and prostate gland. The vas deferens and the excretory duct of the seminal vesicle join each other to form the ejaculatory duct ⇨, which, through the verumontanum, drains into prostatic urethra.* **(Right)** *Axial CECT in a patient with a history of left renal agenesis shows a large, rim-calcified, left seminal vesicle cyst ⇨.*





(Left) *Axial NECT in patient with a history of left renal agenesis shows an absent ipsilateral, left seminal vesicle. Note the normal right SV ⇨.* **(Right)** *Axial NECT obtained more superiorly in the same patient shows the left vas deferens agenesis as well. The right vas deference is annotated ⇨. Because of the common pathway of development, there is a high association between anomalies of these organs.*





TERMINOLOGY

Abbreviations

- SV: Seminal vesicle

IMAGING

Normal SV

- US: Normal SV appears as elongated septate cystic structure on transrectal US
  - SV cannot adequately be evaluated by transabdominal US due to its depth
- CT: Due to convoluted nature of SV, its cystic nature is not well appreciated on CT
  - SVs appear as paired soft tissue density structures with bowtie configuration
- MR: ↓T1 and ↑T2 with thin septations
  - SV wall and septations have ↓T1 and ↓T2 and show mild enhancement

SV Agenesis/Hypogenesis

- Absent, or markedly atrophic, SV
  - Agenesis more common than hypogenesis
  - Usually unilateral
- Due to interruption of normal SV development; occurs before 7th week of gestation
- High association with renal and vas deferens anomalies
  - Ipsilateral renal agenesis (~ 80%) or other renal anomalies (~ 10%)
  - Usually associated with agenesis of ipsilateral vas deferens
- Bilateral SV agenesis strongly associated with *CFTR* gene mutation (64-73%)
  - Considered a genital form of cystic fibrosis
  - SV agenesis believed to be secondary to obstruction of vas deferens by thick secretions

Congenital SV Cyst

- Can be isolated or associated with other genitourinary anomalies
  - Ipsilateral renal agenesis/dysgenesis (~ 70%)
  - Agenesis of ipsilateral vas deferens
  - Ectopic ureteral insertion
  - Zinner syndrome: Triad of SV cyst, renal agenesis, and ejaculatory duct obstruction
- Bilateral SV cysts have high association with autosomal dominant polycystic kidney disease (44-60%)
- SV cysts usually < 5 cm
- US: High resolution transrectal ultrasound useful for identification and evaluation of SV cysts
- CT: SV cyst may simply be seen as SV enlargement if cyst is small; larger cysts present as rounded near water attenuation mass in retrovesical space
- MR: ↑ T2, variable T1 signal
  - SV cyst may show hight T1 signal due to proteinaceous or hemorrhagic cyst content

DIFFERENTIAL DIAGNOSIS

Acquired Seminal Vesicle Cysts

- Any obstructive process in SV, ejaculatory duct, or vas deferens (such as infection, scarring, tumor, etc.) can result in acquired SV cyst
- SV abscess can occur in setting of prostatitis and seminal vesiculitis (rare)

Müllerian Duct Cyst

- Cyst arising from remnants of müllerian duct
  - Anywhere along the path of müllerian duct
  - Usually in midline prostate
- Differentiation between large, eccentric müllerian cysts and SV cysts may be difficult

Cyst-Like Intrapelvic Structures (Mimics)

- Intrapelvic cysts and cyst-like structures can mimic SV cysts
  - Ureterocele, dilated ectopic ureter, bladder diverticulum, urethral diverticulum

PATHOLOGY

Embryology

- Development of SV, vas deferens, and urinary system closely associated
- **Mesonephric (Wolffian) duct** gives rise to SV and vas deferens
  - Ureteric buds also arise from distal aspect of Mesonephric duct and give rise to ureters
  - Growth of ureters induces formation of kidneys from metanephric blastomas

Anatomy

- SVs are paired **extraperitoneal** convoluted tubular structures which secret seminal fluid (stabilizing pH and necessary for function and nourishment of spermatozoa)
- SV forms as outpouching from lateral wall of ampulla of vas deferens
- **Excretory duct** (very distal aspect of SV) joins vas deferens to form **ejaculatory duct**
  - Ejaculatory ducts course through substance of prostate and open through verumontanum into the prostatic urethra

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic; often incidental finding on imaging
- Other signs/symptoms
  - Azoospermia (in bilateral SV agenesis/hypogenesis)
  - Hematospermia, hematuria, prostatitis, epididymitis, urinary tract infection

SELECTED REFERENCES

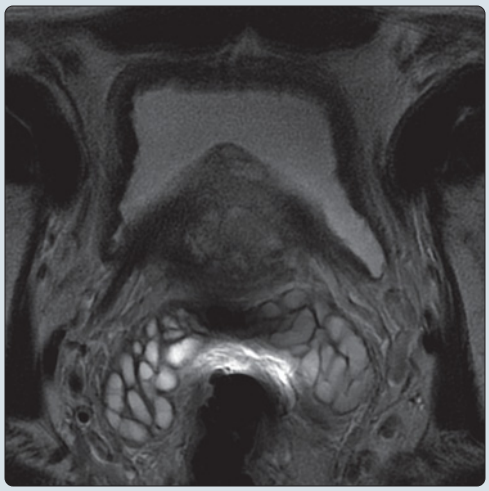
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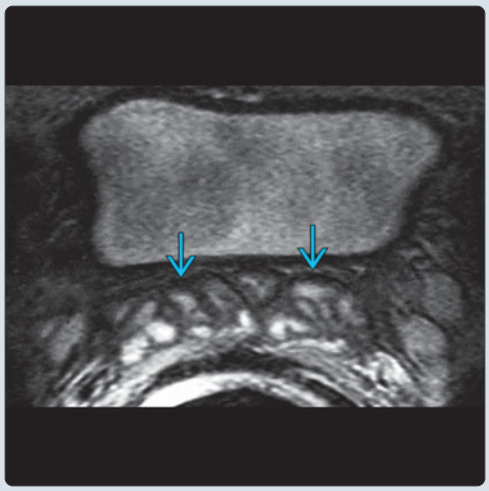
KEY FACTS

IMAGING

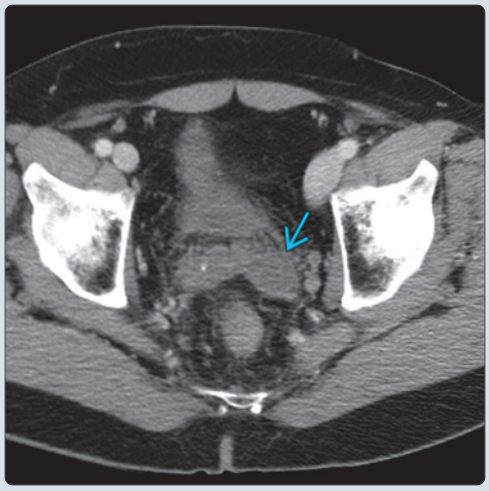
- **Infectious/inflammatory conditions**
  - **Acute seminal vesiculitis**
    - Usually caused by bacterial infection and seen in association with prostatitis &/or epididymitis
    - Imaging: Enlarged seminal vesicles (SVs); diffuse wall thickening with diffuse enhancement; complex fluid content; surrounding inflammatory changes
  - **Chronic seminal vesiculitis**
    - Rare entity caused by chronic or repetitive bacterial infection of SV, usually in association with chronic prostatitis
    - Imaging: Enlargement of SVs or atrophy and loss of convolutions; thickening (± enhancement) of SV septations and walls
  - **Amyloidosis**
    - Localized SV amyloidosis, common finding in elderly; usually an incidental finding with no clinical significance
    - Imaging: **Nodular** (more common) or diffuse (less common) wall thickening with markedly ↓ T2 signal of convolutions; ± SV hemorrhage
    - May mimic tumoral invasion by prostate cancer
- **Degenerative processes**
  - **Acquired SV cyst**
    - Obstruction at level of excretory duct or ejaculatory duct can result in acquired cystic dilatation of SV
  - **SV calcification**
- **Neoplastic lesions**
  - **Primary neoplasms**: Exceedingly rare
    - Benign: Cystadenoma, papillary adenoma, leiomyoma, teratoma
    - Malignant: Adenocarcinoma, leiomyosarcoma, cystosarcoma phyllodes, angiosarcoma, seminoma, carcinoid
  - **Secondary neoplasms**
    - Secondary involvement by local invasion or metastasis

(Left) *Axial T2WI MR, with use of endorectal coil, shows the normal appearance of seminal vesicles with thin-walled convolutions. Note the bladder wall thickening, which is presumably secondary to prostatomegaly.* **(Right)** *Axial T2WI MR, with use of endorectal coil, shows diffuse seminal vesicular wall thickening. In the absence of clinical findings of acute seminal vesiculitis, these findings were thought to be due to chronic seminal vesiculitis or amyloidosis. (Courtesy M. Heller, MD.)*





(Left) *Axial CECT in 52-year-old man with history of perineal pain shows asymmetric enlargement and hypodensity of left seminal vesicle with stranding in rectovesical & perirectal spaces.* **(Right)** *Axial CECT in the same patient shows prostatomegaly (volume of 65 cc) and periprostatic stranding that were new since a prior CT. The patient had new elevated PSA, CRP, and WBC. He was diagnosed with acute prostato-vesiculitis. PSA returned to normal after treatment with systemic antibiotics.*





TERMINOLOGY

Abbreviations

- Seminal vesicle (SV)

Definitions

- Noncongenital conditions affecting seminal vesicles

IMAGING

Normal SV

- US: Normal SV appears as elongated septate cystic structure on transrectal US (TRUS)
  - SV can not be adequately evaluated by transabdominal US due to its depth
- CT: Due to convoluted nature of SV, its cystic nature is not well appreciated on CT
  - SVs appear as paired soft tissue-density structures with bow tie configuration
- MR: ↓ T1 and ↑ T2 with thin septations
  - SV wall and septations are ↓T1 and ↓T2 and show mild enhancement on post-contrast T1W sequences

Infectious/Inflammatory Conditions

- **Acute seminal vesiculitis**
  - Usually caused by bacterial infection and seen in association with prostatitis &/or epididymitis (so-called prostato-vesiculitis and prostate-vesiculo-epididymitis)
    - Some authors use the umbrella term of "male accessory gland infection" (MAGI) to refer to this spectrum
    - *Escherichia coli*, *Neisseria gonorrhea*, *Ureaplasma urealyticum*, *Staphylococcus epidermidis*, *Pseudomonas*, and *Chlamydia trachomatis* are common microbial agents cultured in these patients
  - Patients present with lower abdominal/perineal pain
  - Imaging: Enlarged SVs; diffuse wall thickening with diffuse enhancement; complex fluid content; surrounding inflammatory changes
    - MR, with use of endorectal coil, has superior contrast resolution for evaluation of SVs
    - Proteinaceous and hemorrhagic fluid contents appear as fluid-fluid level with ↑ T1 and variable T2 signal
    - Findings of associated prostatitis, such as prostatomegaly and periprostatic stranding, may also be seen
    - Acute seminal vesiculitis can rarely be complicated by abscess formation
      - Abscess is more common in diabetics and in patients with history of GU instrumentation
  - Treatment: Systemic antibiotics; large abscess might need surgical drainage
- **Chronic seminal vesiculitis**
  - Rare entity caused by chronic or repetitive bacterial infection of SV
    - *E. coli*, *S. epidermidis*, *Pseudomonas*, and *N. gonorrhea* are common microbial agents cultured in these patients
    - Tuberculosis and schistosomiasis are other infectious etiologies, in endemic regions
  - Usually seen in association with chronic prostatitis ("prostato-vesiculitis")
  - Patients usually present with chronic hematospermia, pyospermia, and perineal pain
  - Imaging: Enlargement of SVs or atrophy and loss of convolutions; thickening (± enhancement) of SV septations and walls
    - Imaging findings are nonspecific and are usually indistinguishable from acute seminal vesiculitis or amyloidosis
  - Treatment: Systemic antibiotics

Degenerative Processes

- **Amyloidosis**
  - Localized SV amyloidosis, common finding in elderly, is a senile process caused by localized amyloid deposition in lamina propria of SV convolutions
  - Usually incidental finding with no clinical significance
  - Imaging: **Nodular** (more common) or diffuse (less common) wall thickening with markedly ↓T2 signal of convolutions; ± SV hemorrhage
  - SV amyloidosis may mimic tumoral invasion on T2WI MR in patients with prostate cancer
    - Areas of amyloid deposit do not show enhancement following contrast administration
- **Acquired SV cyst**
  - Obstruction at level of excretory duct or ejaculatory duct can result in acquired cystic dilatation of SV
  - Obstruction is caused by scarring (sequela of prior infection) or tumor
  - Acquired SV cysts are indistinguishable from congenital cysts on imaging
    - Look for associated findings and absence of renal anomalies
- **SV calcification**
  - Calcification of SV and vas deferens is highly associated with diabetes (unclear etiology)
    - Other less common causes are hyperparathyroidism, senile calcification, and postinflammatory dystrophic calcification
  - SV calcification in setting of diabetes is intramural and symmetric
- **SV calculi**
  - Rare entity, similar to prostatic and vas deferens calculi, that is thought to be caused by urine reflux &/or stasis
  - Usually incidental finding on imaging; patient may present with hematospermia
  - Imaging: SV calculi have typical imaging characteristics of urinary stones
    - Echogenic with posterior shadow on TRUS; discrete and hyperdense on CT; ↓ signal on T1WI and T2WI MR

Neoplastic Lesions

- **Primary neoplasms**
  - Primary SV tumors are exceedingly rare
    - Benign: Cystadenoma, papillary adenoma, leiomyoma, teratoma
    - Malignant: Adenocarcinoma, leiomyosarcoma, cystosarcoma phyllodes, angiosarcoma, seminoma, carcinoid
  - **Adenocarcinoma**: Most common primary SV tumor
    - Age in reported cases ranged 13-90; most patients presented with obstructive urinary symptoms and hematospermia

  - Tumors are usually large at the time of presentation and show cystic and necrotic areas; ± local invasion into adjacent structure
    - In advanced infiltrative cases, differentiation from locally advanced prostate or rectal cancer might be difficult based on imaging
    - Tissue sampling with immunohistochemical staining are required to establish primary source
  - Surgical excision ± radiation therapy is treatment of choice; prognosis is poor
- **Cystadenoma**: Most common benign primary SV tumor
  - Age in reported cases ranged 37-66; patients were asymptomatic or presented with pain, hematospermia, or irritative urinary symptoms
  - Cystadenomas are usually large and have mixed solid and cystic appearance
- **Secondary neoplasms**
  - Secondary neoplastic involvement of SV, by local invasion or metastasis, is much more common than primary neoplasm
  - **Local invasion**
    - Seen with **prostate**, rectal, and bladder cancers
    - MR, with endorectal coil, has high specificity and moderate sensitivity for detection of SV invasion by prostate cancer
    - Loss of normal SV architecture, SV wall thickening (with ↓ T2 signal, restricted diffusion, ± enhancement), and obliteration of SV-prostate angle are clues to SV invasion by prostate cancer
    - Ohori et al (1993) classified different pathways of SV invasion by prostate cancer
      - SV invasion can be "internal" (through plane around ejaculatory ducts), transcapsular (with extension of tumor from base of prostate through prostatic capsule and then into SV), or metastatic (noncontiguous tumor deposits remote from primary tumor)
    - Detection of SV invasion has significant impact on treatment strategy and outcome
    - Localized SV amyloidosis might mimic SV tumoral invasion on MR
  - **Metastases**
    - Metastatic involvement from distant organs is extremely rare
    - Noncontiguous tumor deposits in SV in setting of prostate cancer is considered SV invasion rather than metastasis (T3b lesion)
    - Metastases appear as focal soft tissue mass or diffuse enlargement of SV

PATHOLOGY

Anatomy

- SVs are paired **extraperitoneal** convoluted tubular structures that secrete seminal fluid (stabilizing pH and necessary for function and nourishment of spermatozoa)
- SV forms as outpouching from lateral wall of ampulla of vas deferens
- **Excretory duct** (very distal aspect of SV) joins vas deferens to form **ejaculatory duct**
  - Ejaculatory ducts course through substance of prostate and open through verumontanum into prostatic urethra

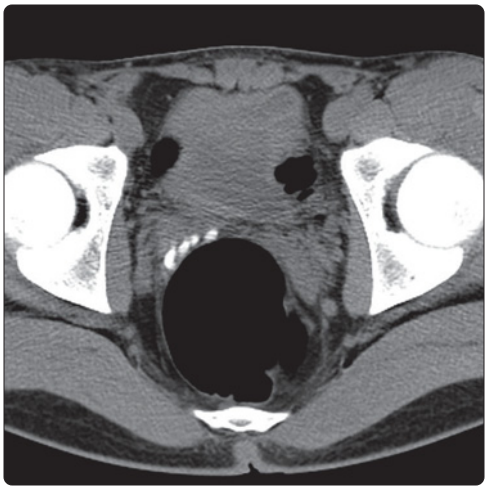
CLINICAL ISSUES

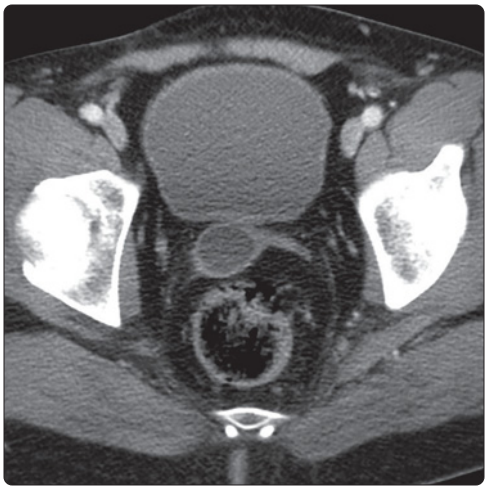
Presentation

- Most common signs/symptoms
  - Inflammatory SV lesions present with perineal pain, hematospermia, and irritative urinary symptoms
  - Small SV tumors are usually asymptomatic; large tumors result in urinary irritative/obstructive symptoms (due to mass effect) and hematospermia

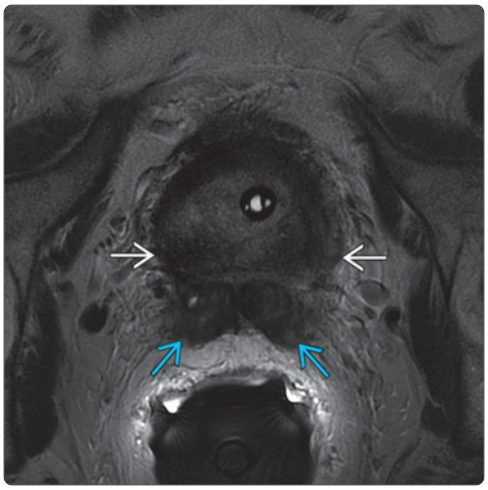
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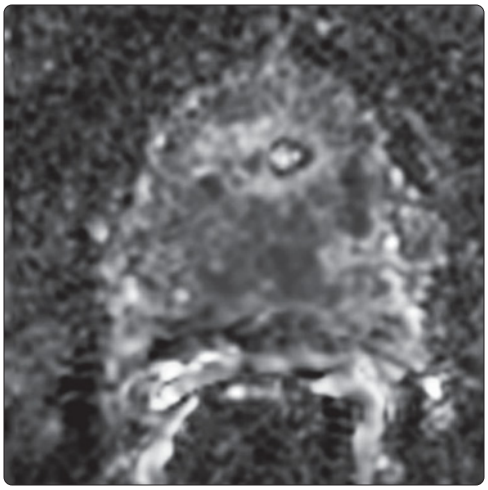
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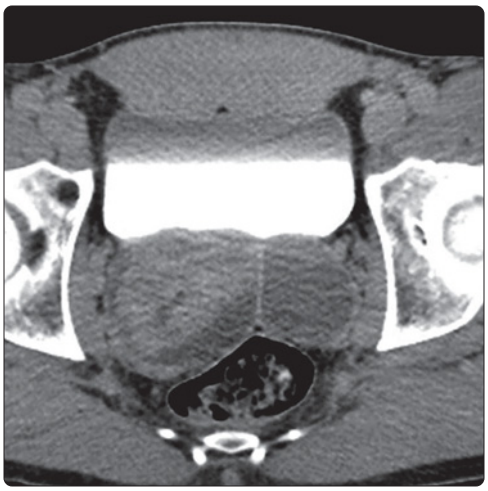


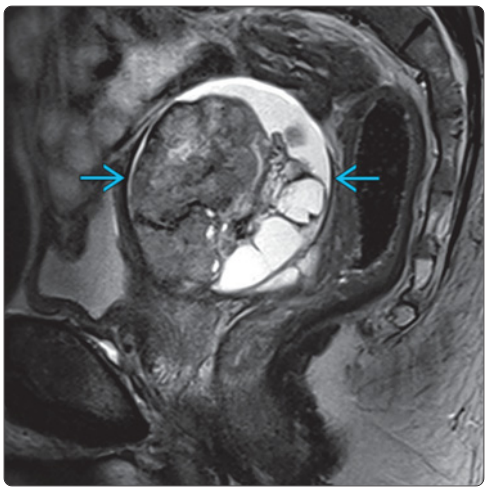
(Left) *NECT through seminal vesicles shows discrete calcified calculi in distribution of right vas deferens. Similar to prostate calculi, the majority of vas deferens and seminal vesicle calculi have urinary composition and are thought to be due to reflux of urine and stasis.* **(Right)** *Axial CECT in 40-year-old male with history of repeated epididymitis shows a right seminal vesicle cyst. Given the history and absence of renal anomalies, this cyst was felt to be sequela of prior infections. Note the marked atrophy of left seminal vesicle.*





(Left) *Axial T2WI MR in 77-year-old patient with history of biopsy-proven Gleason 8 prostate cancer shows abnormal signal in prostate base peripheral zone ➡ as well as distortion and signal alteration of seminal vesicles ⇨.* **(Right)** *ADC map in the same patient shows corresponding restricted diffusion in areas of tumoral invasion. Patient had extracapsular extension and involvement of neurovascular bundles (not shown).*





(Left) *Axial NECT in 52-year-old patient with history of hematospermia shows a large mixed solid and cystic mass in rectovesical space.* **(Right)** *Sagittal T2WI MR in the same patient better shows mixed solid/cystic nature of tumor ⇨. There was no invasion to adjacent organs. The mass was resected and pathology result was consistent with low-grade sarcoma. (Courtesy P. Woodward, MD.)*

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SECTION 12

Prostate



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Prostate Carcinoma	**534**
Prostate Carcinoma Staging	**540**

Prostatitis and Abscess

KEY FACTS

TERMINOLOGY

- NIH classified into 4 types
  - Type 1: Acute bacterial prostatitis
  - Type 2: Chronic bacterial prostatitis
  - Type 3: Chronic prostatitis/chronic pelvic pain syndrome (CP/CPPS)
  - Type 4: Asymptomatic inflammatory prostatitis

IMAGING

- Prostatitis is clinical diagnosis
- Imaging generally reserved to evaluate for abscess formation
  - Rim-enhancing, unilocular or multilocular, low-attenuation mass in prostate
- Focal or diffuse low signal intensity of peripheral zone on T2WI MR

TOP DIFFERENTIAL DIAGNOSES

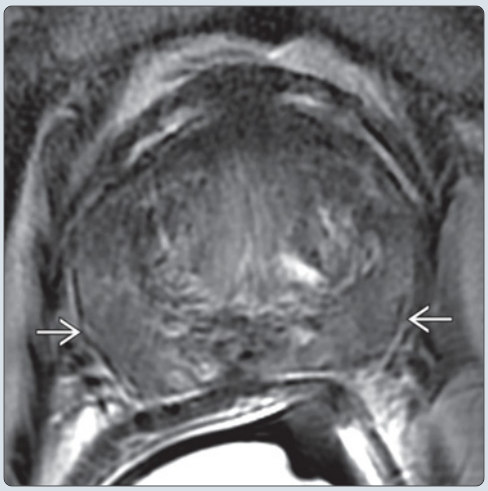
- Congenital prostatic cyst
- Cystic degeneration of benign prostatic hypertrophy
- Prostate carcinoma

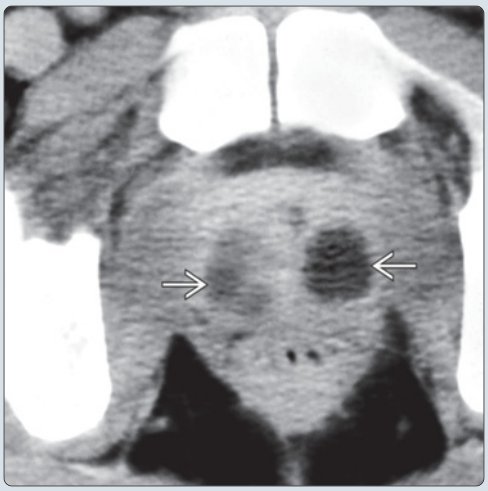
PATHOLOGY

- Acute bacterial prostatitis
  - Ascending colonization of urinary tract or post biopsy
- Predisposing risk factors
  - Bladder outlet obstruction due to benign prostatic hyperplasia
  - Prior instrumentation
  - Chronic indwelling bladder catheters
  - Immunocompromised states, such as diabetes

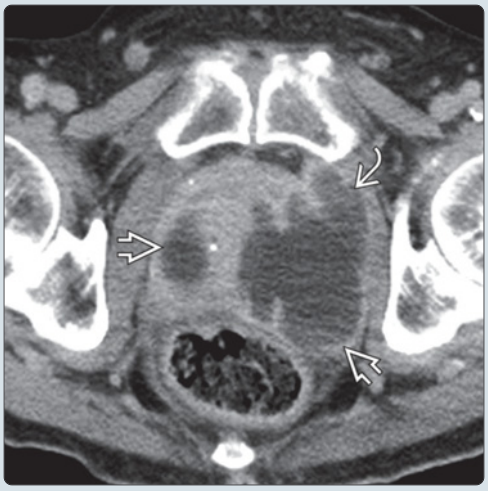
CLINICAL ISSUES

- Treatment
  - Antibiotics
  - Abscess drainage may be necessary





(Left) *Axial T2WI MR shows diffuse low signal intensity of the peripheral zone in this patient with prostatitis. When the inflammatory process affects the peripheral gland focally, it may be difficult to distinguish from cancer, and a biopsy may be required.* **(Right)** *Axial CECT shows multiple rim-enhancing low-density regions within the prostate consistent with abscesses. Medical treatment is usually sufficient, but occasionally prostatic abscesses need drainage.*





(Left) *Axial CECT shows multiple abscesses within an enlarged prostate. The abscess on the left has extended beyond the capsule into the periprostatic tissues.* **(Right)** *Axial CECT shows gas and fluid within the soft tissues of the buttock and thigh, as well as multiloculated fluid collections within the prostate. This was part of an extensive infectious process that involved the entire perineum and portions of the pelvis.*

TERMINOLOGY

Definitions

- NIH classified into 4 types
  - Type 1: Acute bacterial prostatitis
  - Type 2: Chronic bacterial prostatitis
  - Type 3: Chronic prostatitis/chronic pelvic pain syndrome (CP/CPPS)
  - Type 4: Asymptomatic inflammatory prostatitis

IMAGING

General Features

- Prostatitis is clinical diagnosis
- Imaging generally reserved to evaluate for abscess formation

CT Findings

- Prostatitis
  - Diffusely enlarged, edematous gland
  - Peripheral zone most commonly involved
- Abscess
  - Rim-enhancing, unilocular or multilocular, low-attenuation mass in prostate
    - More commonly in peripheral portion of gland
    - Can be central following transurethral prostatectomy
  - May extend through capsule into periprostatic tissues, seminal vesicles, peritoneum

MR Findings

- Prostatitis is a mimicker of cancer on multiparametric MR
- Distribution: Diffuse, lobar, focal
- T2WI
  - Focal: Linear, wedge-shaped lesion with ill-defined margins and low signal intensity
  - Diffuse: Low signal intensity of peripheral zone
- DWI: Low ADC values may overlap with those of prostate cancer

Ultrasonographic Findings

- Transrectal ultrasound
  - Focal hypoechoic areas indistinguishable from prostate cancer
  - Hypoechoic rim surrounding gland
  - Hypoechoic periurethral region
  - Discrete fluid collection suggests abscess formation

DIFFERENTIAL DIAGNOSIS

Congenital Prostatic Cyst

- Usually in or near midline, or along course of ejaculatory duct

Cystic Degeneration of Benign Prostatic Hypertrophy (BPH)

- Small cysts in background of BPH nodules
- Located in transitional zone

Prostate Carcinoma

- Prostatitis is a common mimicker of prostate cancer at multiparametric MR
- Round/oval lesion in peripheral zone with low signal intensity on T2WI MR suggests cancer

PATHOLOGY

General Features

- Etiology
  - Acute bacterial prostatitis
    - Ascending colonization of urinary tract or post biopsy
  - Chronic bacterial prostatitis
    - Recurrent episodes of prostatitis from same uropathogen
  - CP/CPPS
    - Etiology poorly understood
    - May result from infectious or inflammatory process leading to neurologic injury
  - Asymptomatic inflammatory prostatitis
    - May be incidentally found during evaluation of infertility or prostate cancer

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute bacterial prostatitis
    - Fever, chills, dysuria, frequency, and pain
    - Swollen, warm, and tender prostate on digital rectal exam
  - Chronic bacterial prostatitis
    - Dysuria, urgency, pain
    - Greater than 10x bacterial growth in expressed prostatic secretions
  - CP/CPPS
    - Chronic pelvic pain
    - Urinary and ejaculatory dysfunction

Demographics

- Age
  - Most common in men aged 40-70 years
- Epidemiology
  - Worldwide prevalence: 2-10%

Treatment

- Antibiotics
- Abscess drainage may be necessary

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Benign Prostatic Hypertrophy

KEY FACTS

TERMINOLOGY

- Benign prostatic hypertrophy (BPH)

IMAGING

- Best diagnostic clue: Enlarged prostate, nodular hypertrophy in transition/periurethral zones
- IVP: Extrinsic impression on bladder base, "J-hooking"/"fish hooking" of distal ureters
- CT: Enlarged prostate with extrinsic impression on base of bladder
- Ultrasound: 80% demonstrate hypoechoic nodular enlargement of transition and periurethral zones; 10-20% of hyperplastic nodules are isoechoic or echogenic
- T2WI MR: Enlarged transition zone with multiple nodules of different signal intensity

TOP DIFFERENTIAL DIAGNOSES

- Prostate carcinoma
- Prostatic abscess
- Bladder carcinoma

PATHOLOGY

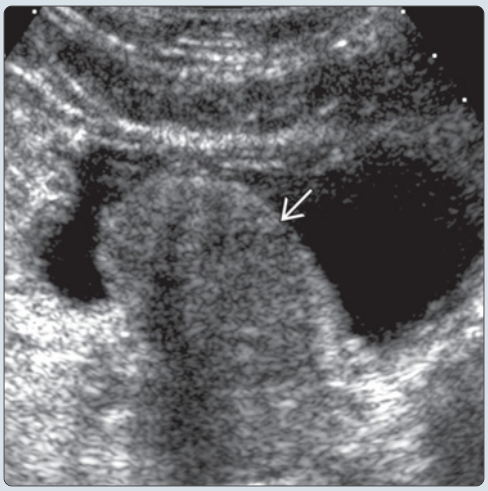
- Enlargement of prostate from benign hyperplastic nodule (fibromyoadenomatous nodule)
- Location: Transition zone and periurethral zone proximal to verumontanum

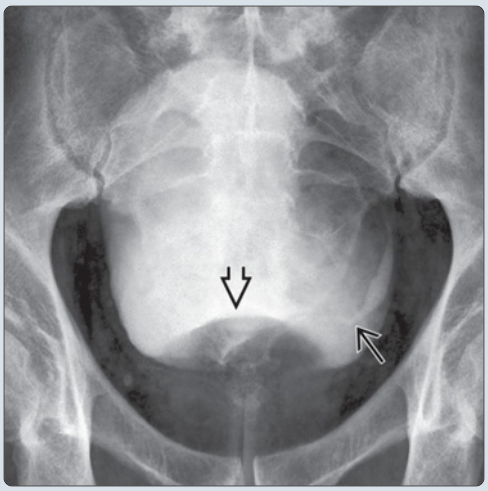
CLINICAL ISSUES

- Prostate volume peaks in 6th/7th decade
- Symptoms of retention and distended bladder, nocturnal dribbling, and poor urethral stream
- May lead to urinary infection, gross hematuria
- Medical therapy (e.g., α_1 blockers) and surgery (e.g., transurethral resection)

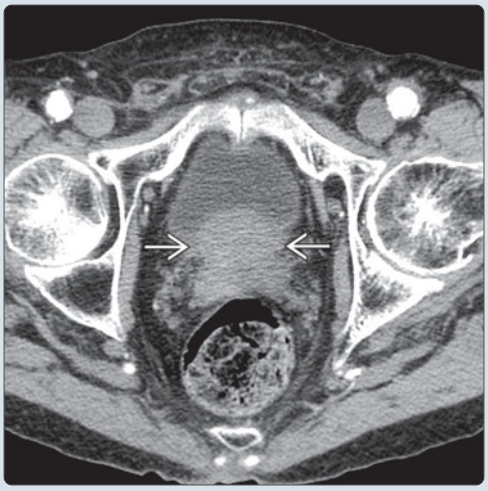
DIAGNOSTIC CHECKLIST

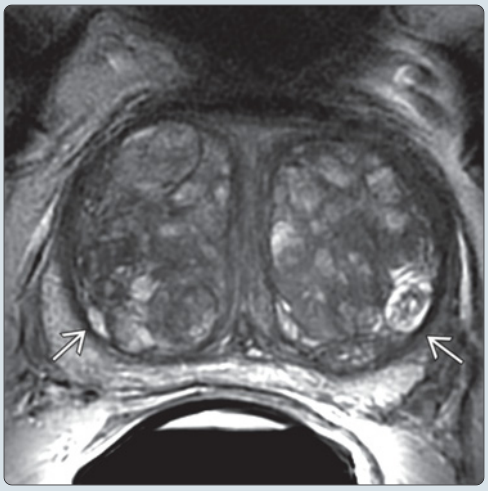
- Consider prostate carcinoma
- Morphology features on T2WI are helpful to differentiate benign from malignant nodule





(Left) *Transverse grayscale ultrasound in an 82-year-old man presenting with symptoms of bladder outlet obstruction reveals marked enlargement of the prostate due to benign prostatic hypertrophy.* **(Right)** *IVP shows an extrinsic impression on the base of the bladder with "fish hooking" of the left ureter in this patient with benign prostatic hypertrophy.*





(Left) *Axial CECT in a 78-year-old man presenting with urinary frequency demonstrates an enlarged prostate gland protruding into the bladder lumen.* **(Right)** *Axial T2WI shows enlarged central gland with multiple nodules of different signal intensity, consistent with benign prostatic hypertrophy. T2WI is helpful for distinguishing benign BPH nodules from tumor originating in the transition zone.*

TERMINOLOGY

Abbreviations

- Benign prostatic hypertrophy (BPH)

Definitions

- Enlarged prostate secondary to stromal and glandular hyperplasia

IMAGING

General Features

- Best diagnostic clue
  - Enlarged prostate, nodular hypertrophy in transition/periurethral zones
- Location
  - Transition/periurethral zone proximal to verumontanum
- Size
  - Central gland and whole prostate volume increases with age

Radiographic Findings

- IVP
  - Extrinsic impression on bladder base, "J-hooking" or "fish hooking" of distal ureters

CT Findings

- NECT
  - Enlarged prostate ± calcifications
- CECT
  - Enlarged prostate, extrinsic impression on bladder base

MR Findings

- T2WI
  - Enlarged transition zone with multiple nodules
  - Signal intensity: ↑ glandular BPH, ↓ stromal BPH
  - Morphology of nodules: Round/oval; usually with capsule

Ultrasonographic Findings

- Grayscale ultrasound
  - Hypoechoic nodular enlargement of transition/periurethral zones
  - Isoechoic or echogenic hyperplastic nodules (10-20%)
  - Large residual urine volume post void

DIFFERENTIAL DIAGNOSIS

Prostate Carcinoma

- Transition zone cancers on T2WI: Lenticular/amorphous shape, low signal intensity, ill-defined margins

Prostatic Abscess

- Low-attenuation fluid collection causing capsule to bulge

Bladder Carcinoma

- Posterior polypoid mass may simulate enlarged median lobe

PATHOLOGY

General Features

- Etiology
  - Aging; hormones (dihydrotestosterone)
- Associated abnormalities
  - Bladder wall hypertrophy, trabeculation, and diverticula

Gross Pathologic & Surgical Features

- Enlarged, firm gland at prostatectomy

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Retention/distended bladder symptoms
  - Enlarged prostate on rectal exam
  - May have increased prostate-specific antigen (PSA)

Demographics

- Epidemiology
  - Prostate volume peaks in 6th/7th decade

Natural History & Prognosis

- May progress to bladder outlet obstruction; hydronephrosis if severe
- May lead to urinary infection, gross hematuria

Treatment

- Options, risks, complications
  - Medical therapy: e.g., α_1 blockers
  - Surgery: Open surgery if gland > 80 g; transurethral resection if smaller

DIAGNOSTIC CHECKLIST

Consider

- Prostate carcinoma

Image Interpretation Pearls

- Morphology features on T2WI are helpful to differentiate benign from malignant nodule

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Prostatic Cyst

KEY FACTS

TERMINOLOGY

- Spectrum of cystic lesions in prostate
  - May be either congenital or acquired

IMAGING

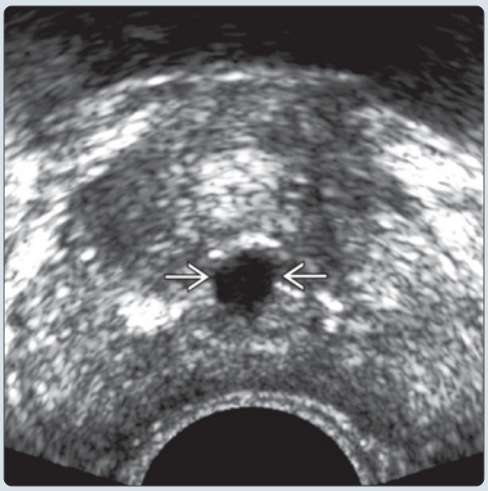
- Best evaluated with transrectal ultrasound (TRUS)
- Subtypes
  - Degenerative cyst
    - Most common prostatic cyst
    - Cystic degeneration of benign prostatic hyperplasia
    - Small cysts in background of hyperplastic nodules
  - Müllerian duct cyst
    - Typically large, extending above prostate
    - Oval or teardrop-shaped
  - Prostatic utricle cyst
    - Arises from midline and rarely extends beyond prostate
    - Commonly communicates with urethra
  - Ejaculatory duct cyst
    - Located along course of ejaculatory duct
  - Retention cyst
    - Obstruction of prostatic gland ducts
    - Away from midline

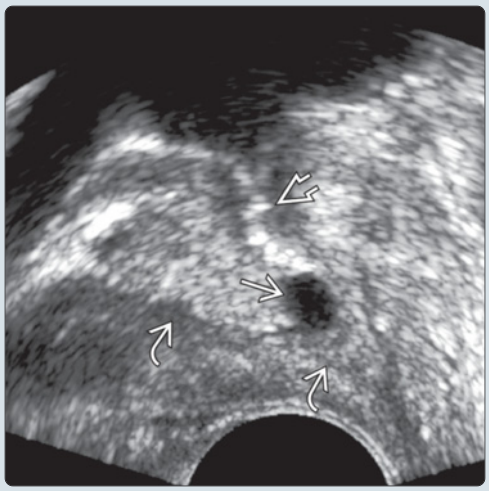
TOP DIFFERENTIAL DIAGNOSES

- Cavitary prostatitis
  - Multiple cysts of varying size
  - Swiss cheese appearance on US
- Prostatic abscess
  - Cystic lesion with thick walls and septa

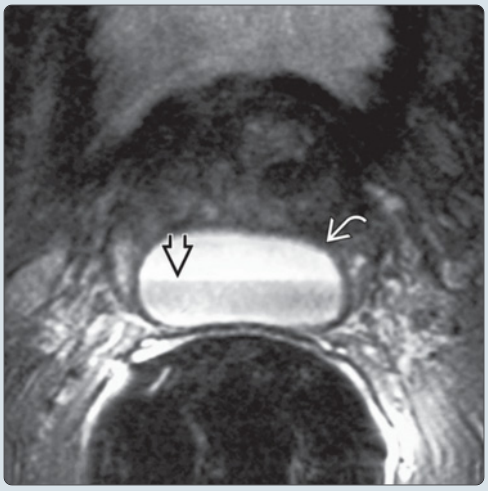
CLINICAL ISSUES

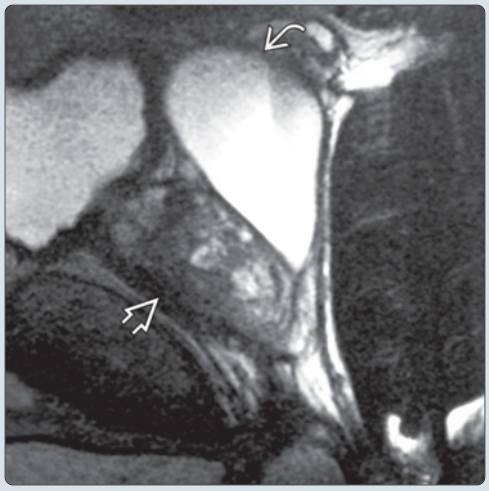
- Prostatic cysts reported in 7.6% of asymptomatic men
- May be symptomatic
  - Pelvic pain, hematuria, rectal pain, dysuria, ejaculatory pain, hematospermia
- Often no treatment; may drain and inject with sclerosing agent if symptomatic





(Left) *Axial transrectal ultrasound shows a midline cyst. To distinguish this from other cysts, it is important to examine the relationship of the cyst to the urethra.* **(Right)** *Sagittal transrectal ultrasound in the same patient shows the cyst anterior to, but separate from, the ejaculatory duct and communicating with the urethra. These features favor a utricle cyst.*





(Left) *Axial T2WI MR shows a hemorrhagic midline cyst within the peripheral zone of the prostate gland. Note the fluid-fluid level created by the layering blood products.* **(Right)** *Sagittal T2WI MR shows the same teardrop-shaped cyst extending cephalad to the prostate gland. Müllerian duct cysts are typically midline and large, extending above the base of the prostate, and only rarely communicate with the urethra. This patient presented with acute pain from the intracystic hemorrhage.*

TERMINOLOGY

Definitions

- Spectrum of cystic lesions in prostate
  - May be either congenital or acquired

IMAGING

General Features

- Location
  - Midline: Müllerian duct cysts, utricle cysts
  - Away from midline: Ejaculatory duct cyst, retention cyst
- Size
  - Variable
  - Müllerian duct cysts tend to be larger
- Morphology
  - Single cyst: Müllerian duct cyst, utricle cyst, ejaculatory duct cyst, retention cyst
  - Multiple cyst: Degenerative cysts, cavitary prostatitis
- Subtypes
  - Degenerative cyst
    - Most common prostatic cyst
    - Cystic degeneration of benign prostatic hyperplasia
    - Multiple small cysts of varying sizes in background of hyperplastic nodules
    - Located in transitional zone of prostate
  - Müllerian duct cyst
    - Remnant of müllerian ducts
    - Arises from verumontanum in midline and extends superiorly
      - Typically large, extending above prostate
      - Oval or teardrop-shaped
      - Rarely communicates with urethra
    - May develop into carcinoma (rare)
  - Prostatic utricle cyst
    - Cystic dilatation of prostatic utricle
    - Arises from midline and rarely extends beyond prostate
      - Usually 8-10 mm in diameter
      - Pear-shaped
      - Commonly communicates with urethra
    - Associated with other congenital genitourinary malformations (e.g., hypospadias, unilateral agenesis)
  - Ejaculatory duct cyst
    - Secondary to congenital or inflammatory obstruction of ejaculatory duct
    - Located along course of ejaculatory duct
      - Paramedian at base, midline at verumontanum
    - Intracystic calculi common
  - Retention cyst
    - Obstruction of prostatic gland ducts
      - Away from midline
      - Usually 1-2 cm in diameter
    - May be indistinguishable from degenerative cyst in benign prostate hyperplasia

CT Findings

- Nonenhancing cystic lesion

MR Findings

- T1WI
  - Generally low signal intensity
  - High signal intensity in case of infection/hemorrhage
- T2WI
  - High signal intensity

Ultrasonographic Findings

- Transrectal ultrasound (TRUS)
  - Well-defined anechoic or hypoechoic lesion with increased through-transmission

DIFFERENTIAL DIAGNOSIS

Cavitary Prostatitis

- Multiple cysts of varying size (Swiss cheese appearance on US)
- Seen in setting of chronic prostatitis

Prostatic Abscess

- Cystic lesion with thick walls and septa
- Clinical presentation aids diagnosis

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Asymptomatic
    - Prostatic cysts reported in 7.6% of asymptomatic men
  - Obstructive urinary symptoms
    - Müllerian duct cyst, utricle cyst
  - Ejaculation pain, hematospermia
    - Ejaculatory duct cyst
  - Urinary tract infection
    - Müllerian duct cyst, utricle cyst
  - Pelvic pain, hematuria, rectal pain, dysuria
    - Potentially any prostatic cyst

Demographics

- Age
  - Utricle cysts: 1st and 2nd decades
  - Müllerian duct cysts: 3rd and 4th decades
  - Retention and degenerative cysts: 5th and 6th decades

Treatment

- Often none; may drain and inject with sclerosing agent if symptomatic

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Prostate Carcinoma

KEY FACTS

IMAGING

- 70% of prostate carcinomas occur in peripheral zone
- Transrectal ultrasonography (TRUS)
  - Hypoechoic lesion in peripheral zone
- Multiparametric MR
  - T1WI
    - Persistent blood from biopsy shows high signal intensity
  - T2WI
    - Peripheral zone: Low signal intensity, round, or ill-defined lesion in peripheral zone
    - Transition zone: Ill-defined low signal intensity lesion
    - Extracapsular extension: Breech of capsule, obliteration of rectoprostatic angle, invasion into neurovascular bundle and seminal vesicles
  - DWI
    - Focal area hyperintense on DWI (b value > 800 s/mm²) and hypointense on ADC maps
  - DCE-MR
    - Rapid wash-in and wash-out
  - MRS
    - ↓ citrate level; ↑ choline level
    - ↑ choline + creatine:citrate ratio

TOP DIFFERENTIAL DIAGNOSES

- Benign prostatic hypertrophy (BPH)
- Bladder carcinoma
- Prostatitis

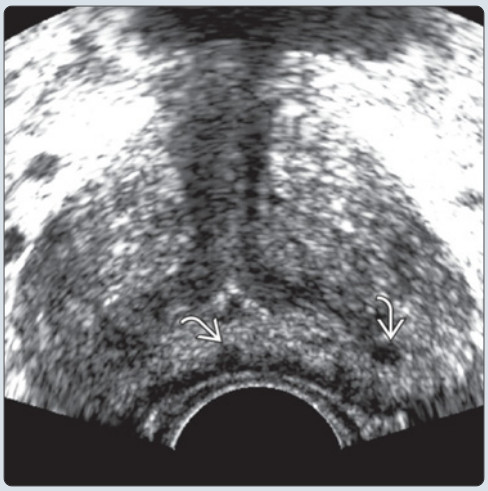
PATHOLOGY

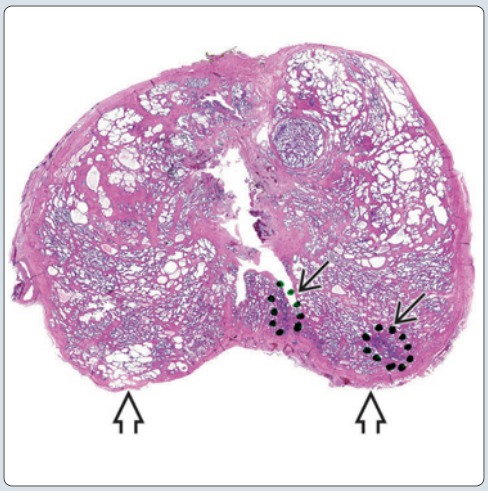
- > 95% of tumors are adenocarcinoma

CLINICAL ISSUES

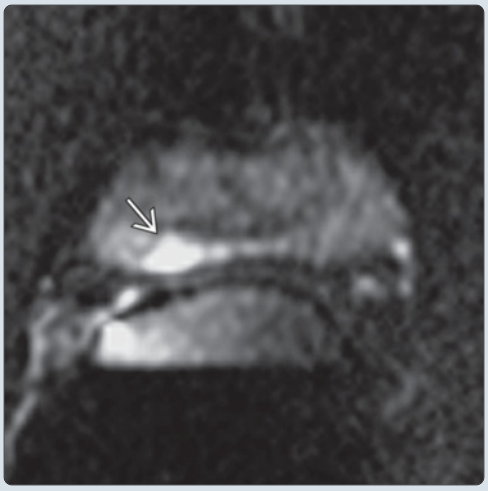
- Most common noncutaneous cancer in American men
- 2nd most common cause of cancer deaths in men after lung cancer
- Most important factor affecting choice of treatment is presence or absence of extracapsular extension

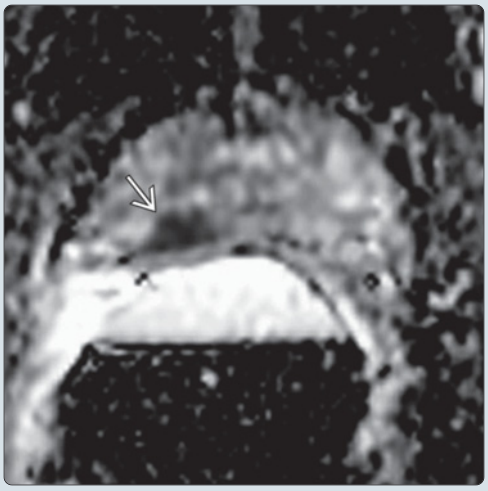
(Left) *Transverse TRUS shows a mildly enlarged prostate with small, hypoechoic lesions in the peripheral zone. The prostate-specific antigen (PSA) level was elevated (14.2 ng/mL). Biopsy of the lesions confirmed carcinoma.* **(Right)** *Resected prostate (whole mount section) shows the tumor inked. The prostatic capsule is well demonstrated.*





(Left) *Axial DWI MR (b = 1,000 s/mm²) shows a round hyperintense focus in the right postero-medial peripheral zone of the mid-gland, corresponding to biopsy-proven prostate adenocarcinoma.* **(Right)** *Axial ADC map shows the cancer in the right peripheral zone as a low signal intensity lesion. A focal lesion in the peripheral zone hyperintense on high b-value DWI and hypointense on ADC is highly suspicious for cancer.*





TERMINOLOGY

Synonyms

- Prostate cancer (PC)

Definitions

- Malignancy of prostate gland
- > 95% of tumors are adenocarcinoma

IMAGING

General Features

- Best diagnostic clue
  - Transrectal ultrasound: Hypoechoic lesion in peripheral zone (PZ)
  - Multiparametric MR (mpMR): Low signal intensity (SI) focal lesion in PZ
- Location
  - 70% in PZ, 20-25% in transition zone (TZ), 5-10% in central zone (CZ)

Ultrasonographic Findings

- **Transrectal ultrasonography (TRUS)**
  - Hypoechoic lesion in PZ
  - Primary use
    - Assessment of prostate size
    - Guide for biopsy
    - Brachytherapy seed placement
  - Limitations
    - Limited accuracy (62%)
      - False-negative: > 40% of cancers are isoechoic; 1-5% are hyperechoic
      - False-positive: Prostatitis may appear hypoechoic
  - Color Doppler and power Doppler ↑ accuracy

CT Findings

- Limited role in detecting and staging PC
  - Poor soft tissue resolution makes it impossible to define zonal anatomy
- Primary use: Evaluation of advanced disease
  - Lymphadenopathy
  - Bone metastases (osteoblastic)

MR Findings

- T1WI
  - Normal appearance of prostate gland
    - Low SI with poor differentiation of PZ and TZ
  - PC appearance: Isointense to surrounding prostate
  - Advantage: High SI of post-biopsy hemorrhage (methemoglobin)
- T2WI
  - Normal appearance of prostate gland
    - PZ
      - 70% volume of gland
      - High and homogeneous SI
    - TZ
      - 5% volume of gland; surrounding proximal prostatic urethra
      - Enlarged in BPH; heterogeneous SI: "Organized chaos"
    - CZ
      - 25% volume of gland
      - Low SI cone-shaped structure surrounding ejaculatory ducts
  - PC appearance
    - PZ
      - Low SI lesion confined to prostate
      - Low SI mass with extracapsular extension (ECE)
    - TZ
      - Ill-defined, lenticular shape, homogeneous low SI lesion (erased charcoal drawing sign)
      - Invasion of anterior fibromuscular stroma
  - Advantages
    - High sensitivity for tumor detection
    - Assessment of ECE
      - Bulging prostatic capsule
      - Disruption of prostatic capsule
      - Obliteration of rectoprostatic angle
      - Asymmetry/encasement of neurovascular bundle
      - Asymmetric low SI in seminal vesicles
  - Limitations
    - False-positives: Biopsy-related hemorrhage, prostatitis, fibrosis, radiotherapy
    - Tumors in TZ are difficult to differentiate from surrounding gland
- DWI
  - Normal appearance of prostate gland
    - PZ: Homogeneous high SI on ADC map
  - PC appearance: Focal area of high SI on DWI (b value > 800/1,000 s/mm²) and low SI on ADC map
  - Advantages
    - Information on tumor aggressiveness (ADC values correlate with Gleason score)
    - Specificity of T2WI + DWI > T2WI
  - Limitations
    - Poor spatial resolution; susceptible to motion and magnetic field inhomogeneities
    - ADC values depend on field strength and b value
- T1WI C+
  - PC appearance
    - Qualitative (visual) analysis: Early hyperenhancement; rapid washout
    - Semiquantitative analysis: Type III curve (increase followed by decline) most suspicious for cancer
    - Quantitative analysis: ↑ Ktrans (forward volume transfer constant); ↑ Kep (reverse reflux rate constant)
  - Advantages
    - Improved diagnostic accuracy when combined with T2WI and DWI
    - Improved assessment of ECE when combined with T2WI
    - Detection of tumor recurrence
  - Limitations
    - False-positive: Prostatitis in PZ, and BPH in TZ
    - Lack of standardization for acquisition and analysis
- MRS
  - Dominant compounds in prostate gland
    - Citrate (2.60 ppm)
    - Creatine (3.04 ppm)
    - Choline (3.20 ppm)
  - PC appearance

- – ↓ citrate level; ↑ choline level
- – ↑ choline + creatine:citrate ratio
- Advantages
  - – Increased specificity
  - – Information on tumor aggressiveness
- Limitations
  - – Increased total acquisition time
  - – High expertise required

Nuclear Medicine Findings

- Bone scan
  - ↑ uptake in bone metastases
  - Location: Pelvis, thoracic and lumbar spine, ribs
  - Indications
    - – Gleason score ≥ 8; prostate-specific antigen (PSA) ≥ 20 ng/mL; T ≥ 3
    - – Advanced prostate cancer and bone pain
    - – Post-treatment follow-up
- PET
  - 18F-FDG
    - – Limited for detection and staging of tumor (poor tumor uptake; interference from excretion into bladder)
  - 11C-choline; 18F-fluorocholine
    - – Role in restaging patients with biochemical failure (PSA relapse) after treatment
- In-111 Capromab pendetide (ProstaScint scan)
  - Staging: Potential role in detecting lymph node metastases
  - Detection of recurrent disease

Imaging Recommendations

- Best imaging tool
  - mpMR
    - – Detection (PZ cancer): Sensitivity 81%, specificity 91%
    - – Staging: NPV for ECE 95%
    - – Active surveillance
    - – Guidance for biopsy
    - – Post-treatment follow-up
- Protocol advice
  - Multiplanar mpMR
    - – High-resolution T2WI +
      - ▫ DWI
      - ▫ Dynamic contrast-enhanced MR (DCE-MR)
      - ▫ MR spectroscopy (MRS [optional])
    - – Pelvic phased array coil + endorectal coil
    - – 1.5T-3.0T magnet
    - – > 6 weeks post biopsy

DIFFERENTIAL DIAGNOSIS

Benign Prostatic Hypertrophy (BPH)

- Stromal BPH: Round nodules with discrete margins; low SI on T2WI

Bladder Carcinoma

- Tumor involving base of bladder may mimic PC

Prostatitis

- Low SI on T2WI: Linear/wedge-shape morphology; ill-defined margins.
- Distribution: Lobar; diffuse

PATHOLOGY

General Features

- Etiology
  - Age, hormones, environmental and genetic factors

Staging, Grading, & Classification

- **T (tumor) stage (AJCC 7th edition)**
  - T0: No evidence of tumor
  - T1: Clinically inapparent
    - – T1a: Incidental finding in < 5% of tissue resected
    - – T1b: Incidental finding in > 5% of tissue resected
    - – T1c: Identified by needle biopsy
  - T2: Confined to prostate
    - – T2a: < 1/2 of 1 lobe
    - – T2b: > 1/2 of 1 lobe
    - – T2c: Both lobes
  - T3: Extension beyond prostate capsule
    - – T3a: Unilateral or bilateral extracapsular extension
    - – T3b: Invasion of seminal vesicle
  - T4: Invasion of adjacent structures
    - – Bladder, rectum, levator ani, pelvic sidewall
- **N (node)**
  - N0: No regional lymph node metastasis
  - N1: Metastasis in regional lymph node
    - – Regional lymph nodes: Hypogastric, obturator, iliac, sacral
- **M (metastasis)**
  - M0: No distant metastasis
  - M1: Distant metastasis
    - – M1a: Nonregional lymph node
    - – M1b: Bone
    - – M1c: Other site
- **Gleason score**
  - Score 1-5 (5 being worst) for 2 most common areas of tumor
    - – 2-6: Well differentiated with good prognosis
    - – 7: Variable prognosis
    - – 8-10: Aggressive tumor with high risk of recurrence

Gross Pathologic & Surgical Features

- > 95% of PC are adenocarcinoma
- Tumor spread
  - Hematogenous
    - – Prostatic venous plexus drains into internal iliac veins and communicates with vertebral venous plexus
    - – Sacrum and lumbar spine most common site of bone metastases (osteoblastic)
  - Lymphatic
    - – Primary drainage to internal iliac nodes then to retroperitoneal nodes
  - Local
    - – Seminal vesicles, bladder, rectum

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Symptoms
    - – Asymptomatic

- Urinary symptoms: Hesitancy, urgency, increased frequency
- Bone pain from metastases
- Abnormal digital rectal exam (DRE)

- **Laboratory data**
  - Increased PSA level
    - Normal levels: < 4.0 ng/mL (NPV 85%)
    - Elevated PSA not specific for prostate cancer
    - PSA velocity: Rate of increase of PSA over time
    - PSA density: Serum PSA/volume of prostate
- **Screening**
  - PSA screening has greatest benefit in men 55-69 years old
    - Shared decision making recommended
  - Screening interval of 2 years is preferred

Demographics

- Age
  - ↑ risk with age; rare < 50 years
  - Median age at diagnosis: 68 years old
- Ethnicity
  - African Americans > Caucasians
- Epidemiology
  - Lifetime risk: 1 in 6 men (developed countries)
  - Most common noncutaneous malignancy
  - 2nd most common cause of cancer death in men after lung cancer

Natural History & Prognosis

- Complications
  - Bladder outlet and rarely rectal obstruction
  - Obstructive uropathy, uremia, pathological fractures
- Prognosis
  - Classified in 1 of 3 risk groups (low, intermediate, high) based on PSA, Gleason score, and clinical stage

Treatment

- Options, risks, complications
  - Surgery
    - Low- and intermediate-risk localized PC
    - Radical prostatectomy; pelvic lymphadenectomy
    - Risks: Erectile dysfunction, urinary incontinence, death
  - Radiation therapy
    - High-risk locally advanced PC
    - External beam radiotherapy; brachytherapy
    - Risks: Erectile, bladder dysfunction
  - Cryotherapy; high intensity focused ultrasound (HIFU)
  - Hormonal therapy; chemotherapy (advanced disease)
- **Most important factor affecting choice of treatment is presence or absence of ECE**

DIAGNOSTIC CHECKLIST

Consider

- mpMR for PC staging

Image Interpretation Pearls

- T2WI: Low SI lesion on PZ

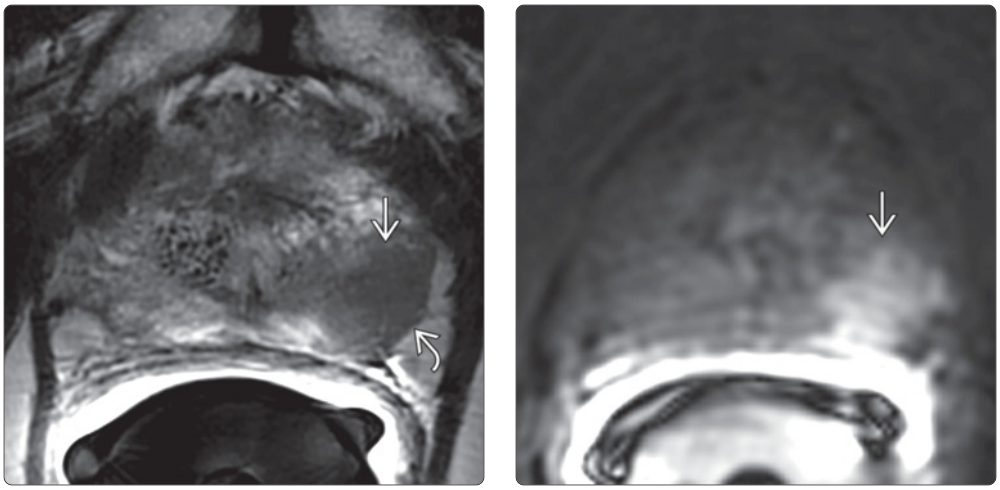
Reporting Tips

- Prostate Imaging Reporting and Data System (PI-RADS)
  - Evolving standardization system for acquisition, interpretation, and reporting of mpMR

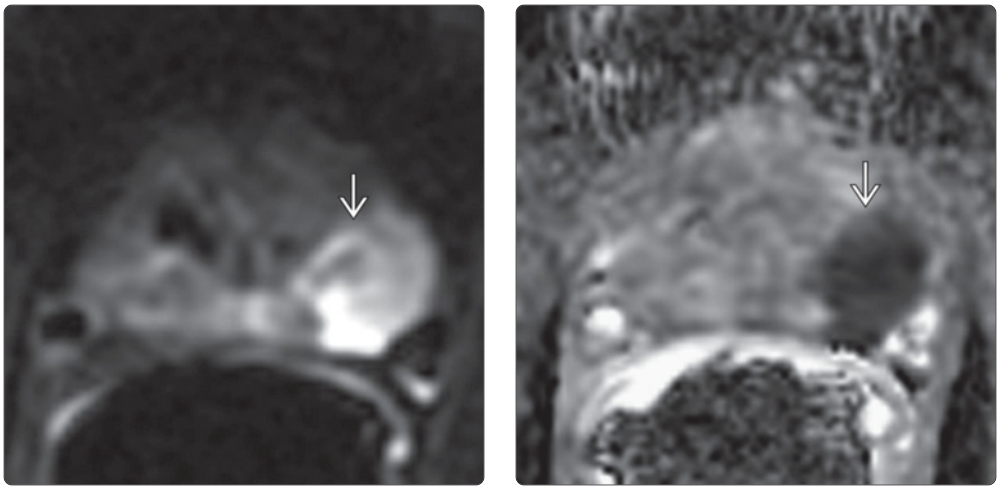
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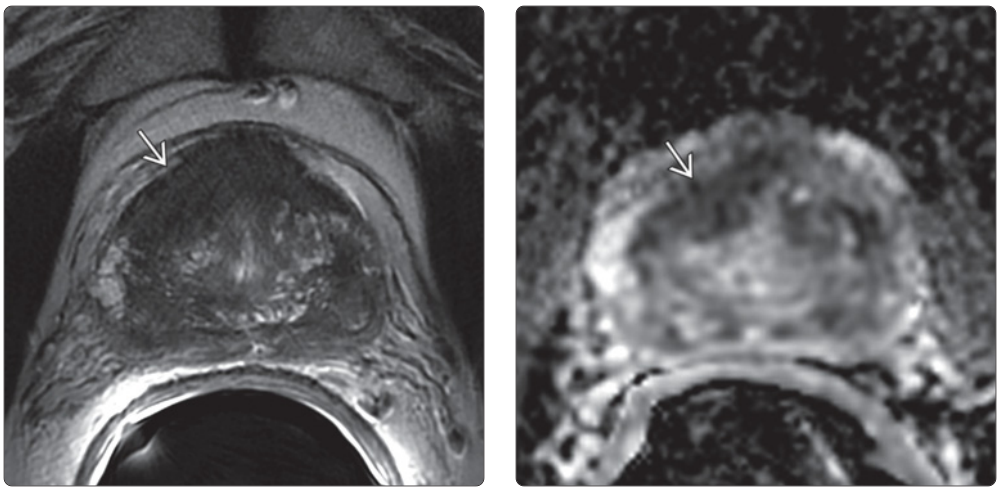
(Left) *Axial T2WI through the prostate apex shows a large hypointense lesion in the left peripheral zone. The lesion shows a broad-base contact and bulging of the capsule.* **(Right)** *Axial T1WI MR acquired post gadolinium injection (DCE-MR) shows focal early enhancement corresponding to the lesion demonstrated on T2WI. Prostate cancer usually shows rapid wash-in greater than surrounding gland because of tumor angiogenesis.*

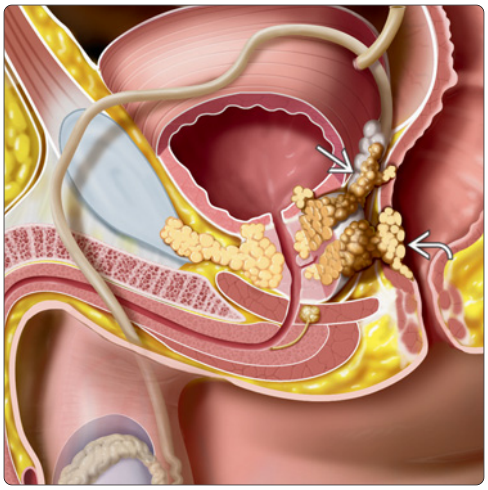


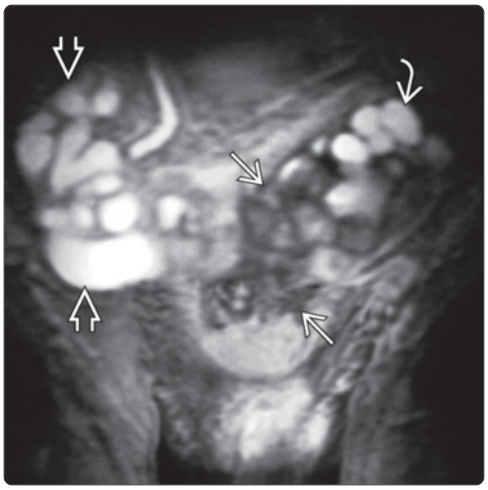
(Left) *Axial DWI MR (b = 1,500 s/mm²) demonstrates the mass in the left peripheral zone as markedly hyperintense.* **(Right)** *Axial ADC map shows the mass in the left peripheral zone as markedly hypointense. The mass is a biopsy-proven Gleason score 7 (3+4) prostate adenocarcinoma. Extracapsular extension was demonstrated at radical prostatectomy.*



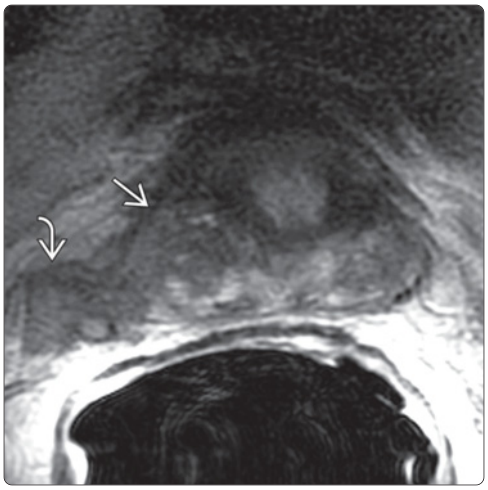
(Left) *Axial T2WI through the mid-gland shows a lenticular, hypointense mass with ill-defined borders (erased charcoal drawing sign) in the anterior right transition zone, corresponding to biopsy proven prostate adenocarcinoma. The transition zone is the origin of 20-25% of prostate cancers, which may be difficult to differentiate from benign prostatic hypertrophy nodules.* **(Right)** *Axial ADC map shows the tumor in the transition zone as hypointense mass.*

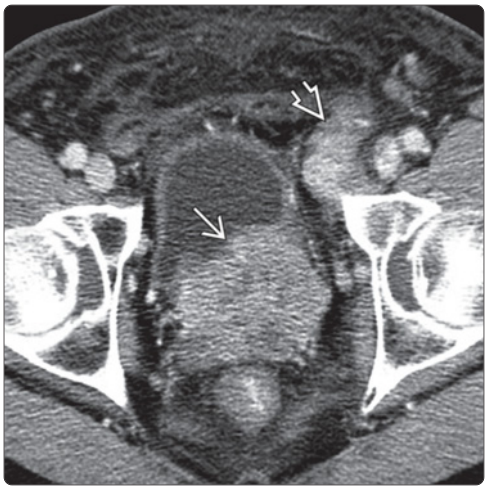




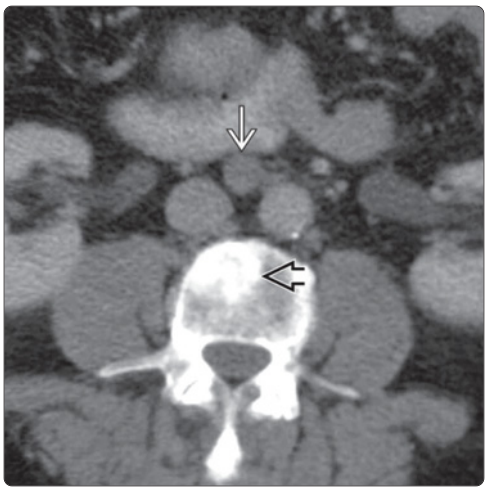


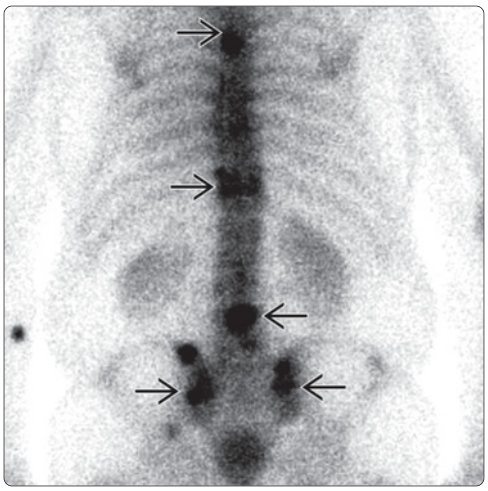
(Left) *Sagittal graphic of the prostate gland shows tumor invasion into the surrounding tissues, including the rectum and seminal vesicles.* **(Right)** *Coronal T2WI MR shows the normal serpiginous, high signal intensity, right seminal vesicle. Although there is invasion of low signal tumor into the left seminal vesicle, a short segment of uninvolved left seminal vesicle remains.*





(Left) *Axial T2WI MR shows a large prostate cancer in the right peripheral zone as a low signal intensity mass. Note associated extracapsular extension with encasement of the right neurovascular bundle. The assessment of extracapsular extension is important for proper treatment planning.* **(Right)** *Axial CECT of advanced disease shows an enhancing mass within the prostate with obliteration of the fat plane between the mass and the bladder. There is also inguinal adenopathy.*





(Left) *Axial CECT shows retroperitoneal lymphadenopathy and sclerotic vertebral metastasis.* **(Right)** *Bone scan shows multiple metastases, predominately in the lumbar spine and pelvis. Because there is drainage from the prostatic venous plexus to the vertebral venous plexus, the lower spine is a common site of involvement. Bone metastases are generally osteoblastic.*

T | Primary Tumor

TNM	Definitions
Clinical	
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Clinically inapparent tumor neither palpable nor visible by imaging
T1a	Tumor incidental histologic finding in ≤ 5% of tissue resected
T1b	Tumor incidental histologic finding in > 5% of tissue resected
T1c	Tumor identified by needle biopsy (e.g., because of elevated PSA)
T2	Tumor confined within prostate[1]
T2a	Tumor involves ≤ 1/2 of 1 lobe
T2b	Tumor involves > 1/2 of 1 lobe but not both lobes
T2c	Tumor involves both lobes
T3	Tumor extends through the prostate capsule[2]
T3a	Extracapsular extension (unilateral or bilateral)
T3b	Tumor invades seminal vesicle(s)
T4	Tumor is fixed or invades adjacent structures other than seminal vesicles, such as external sphincter, rectum, bladder, levator muscles, &/or pelvic wall
Pathologic[3]	
pT2	Organ confined
pT2a	Unilateral, ≤ 1/2 of 1 side
pT2b	Unilateral, involving > 1/2 of 1 side but not both sides
pT2c	Bilateral disease
pT3	Extraprostatic extension
pT3a	Extraprostatic extension or microscopic invasion of bladder neck[4]
pT3b	Seminal vesicle invasion
pT4	Invasion of rectum, levator muscles, &/or pelvic wall

[1]Tumor found in 1 or both lobes by needle biopsy, but not palpable or reliably visible by imaging, is classified as T1c. [2]Invasion into the prostatic apex or into (but not beyond) the prostatic capsule is classified not as T3 but as T2. [3]There is no pathologic T1 classification. [4]Positive surgical margin should be indicated by an R1 descriptor (residual microscopic disease). PSA = prostate-specific antigen.

Adapted from 7th edition AJCC Staging Forms (2010).

N | Regional Lymph Nodes

TNM	Definitions
Clinical	
NX	Regional lymph nodes were not assessed
N0	No regional lymph node metastasis
N1	Metastasis in regional lymph node(s)
Pathologic	
pNX	Regional nodes not sampled
pN0	No positive regional nodes
pN1	Metastases in regional node(s)

Adapted from 7th edition AJCC Staging Forms (2010).

M | Distant Metastasis

TNM	Definitions
M0	No distant metastasis
M1	Distant metastasis
M1a	Nonregional lymph node(s)
M1b	Bone(s)
M1c	Other site(s) ± bone disease

When > 1 site of metastasis is present, the most advanced category is used (pM1c is most advanced).

Adapted from 7th edition AJCC Staging Forms (2010).

G | Histologic Grade

TNM	Definitions
GX	Gleason score cannot be processed
Gleason ≤ 6	Well differentiated (slight anaplasia)
Gleason 7	Moderately differentiated (moderate anaplasia)
Gleason 8-10	Poorly differentiated/undifferentiated (marked anaplasia)

Adapted from 7th edition AJCC Staging Forms (2010).

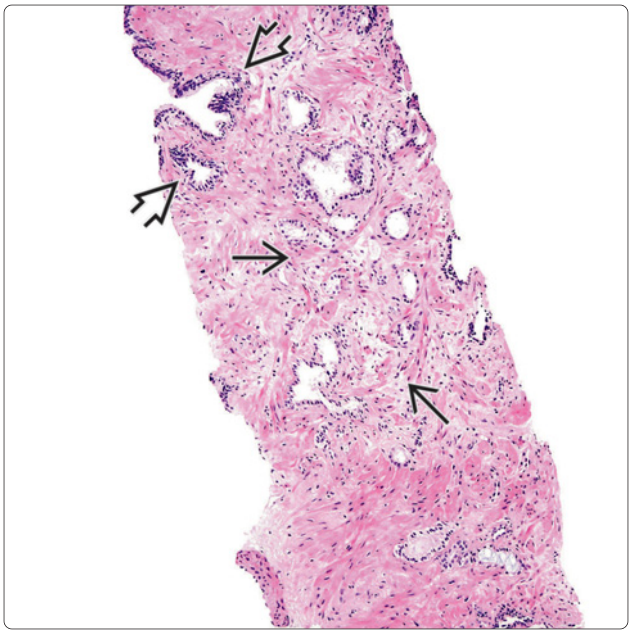
AJCC | Stages/Prognostic Groups

Stage	T	N	M	PSA	Gleason
I	T1a-c	N0	M0	PSA < 10	Gleason ≤ 6
	T2a	N0	M0	PSA < 10	Gleason ≤ 6
	T1-2a	N0	M0	PSA X	Gleason X
IIA	T1a-c	N0	M0	PSA < 20	Gleason 7
	T1a-c	N0	M0	10 ≤ PSA < 20	Gleason ≤ 6
	T2a	N0	M0	PSA < 20	Gleason ≤ 7
	T2b	N0	M0	PSA < 20	Gleason ≤ 7
	T2b	N0	M0	PSA X	Gleason X
IIB	T2c	N0	M0	Any PSA	Any Gleason
	T1-2	N0	M0	PSA ≥ 20	Any Gleason
	T1-2	N0	M0	Any PSA	Gleason ≥ 8
III	T3a-b	N0	M0	Any PSA	Any Gleason
IV	T4	N0	M0	Any PSA	Any Gleason
	Any T	N1	M0	Any PSA	Any Gleason
	Any T	Any N	M1	Any PSA	Any Gleason

When either PSA or Gleason is not available, grouping should be determined by T stage &/or either PSA or Gleason as available.

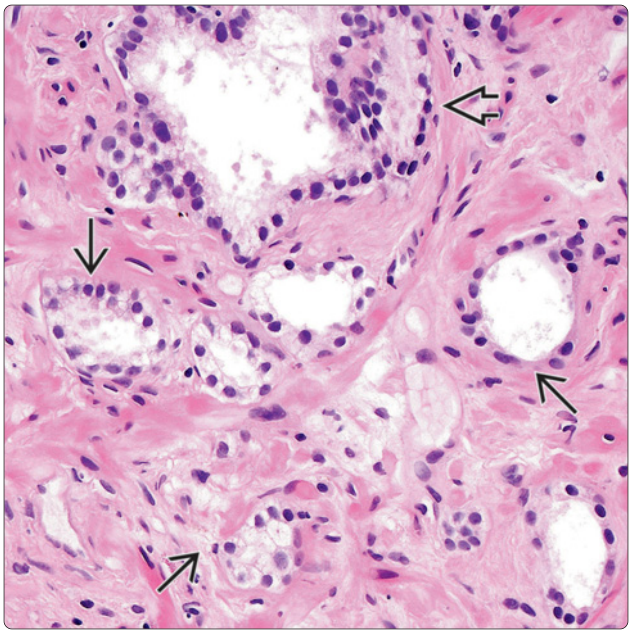
Adapted from 7th edition AJCC Staging Forms (2010).

T1 (Gleason Score 3 + 3 = 6)



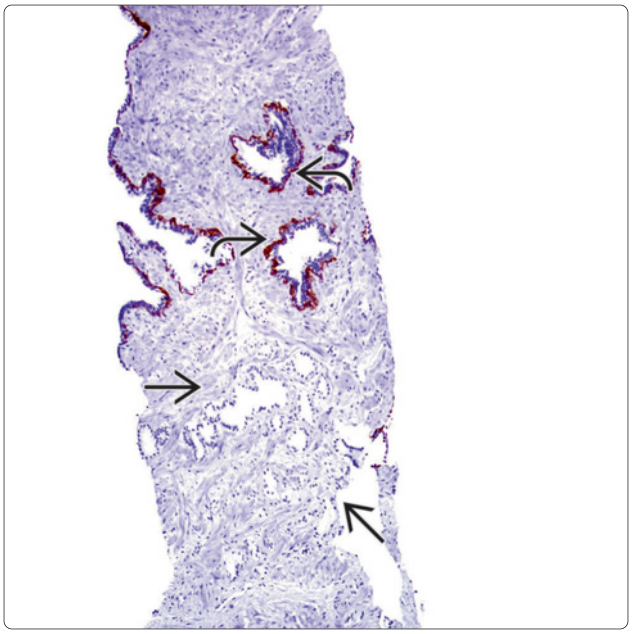
Low-power H&E stain from a core needle prostate biopsy sample shows a small focus of cancer ⇨. Normal prostatic glands are also present ⇨. The tumor represented < 5% of both the core biopsy and the final prostatectomy specimen. (Original magnification, 100x)

T1 (Gleason Score 3 + 3 = 6)



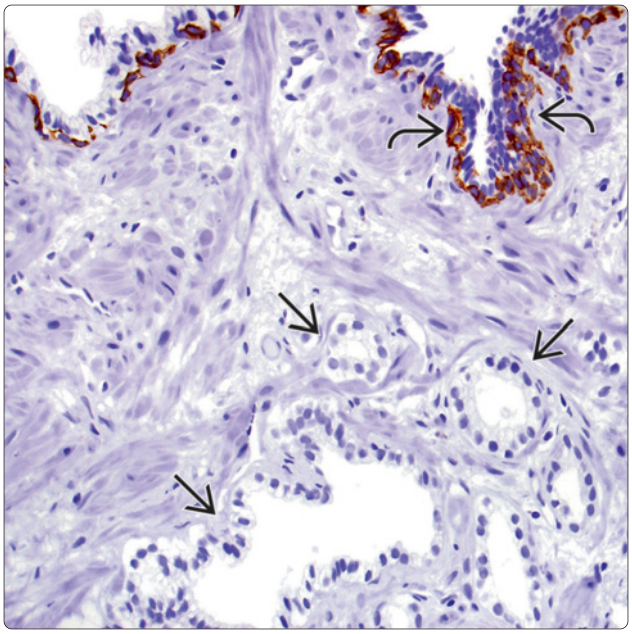
Higher magnification of the same sample shows the numerous invasive neoplastic glands ⇨ lacking basal epithelial cells. Normal prostatic glandular tissue ⇨ has normal benign luminal epithelial cells as well as basal epithelial layer. (Original magnification, 400x)

T1 (Gleason Score 3 + 3 = 6)



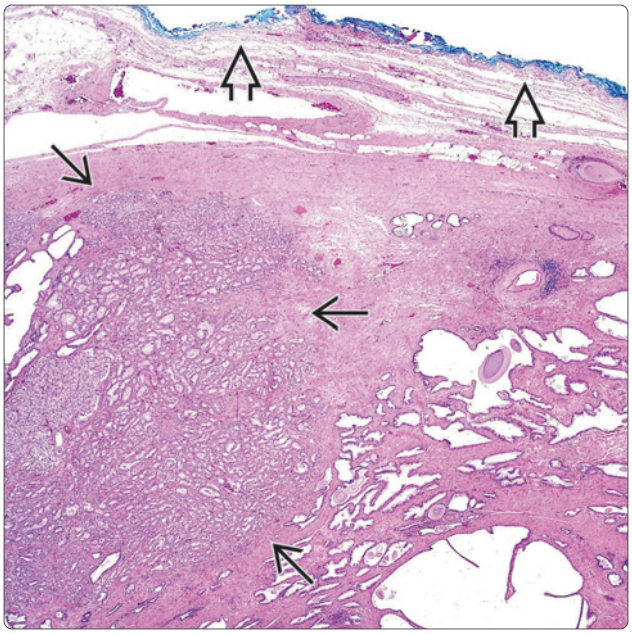
High molecular weight cytokeratin immunostain in the same case demonstrates absence of basal cell layer in the atypical glands ⇨, supporting the diagnosis of adenocarcinoma. The benign glands ⇨ show positive staining, which indicates intact basal cell layer. (Original magnification, 100x)

T1 (Gleason Score 3 + 3 = 6)



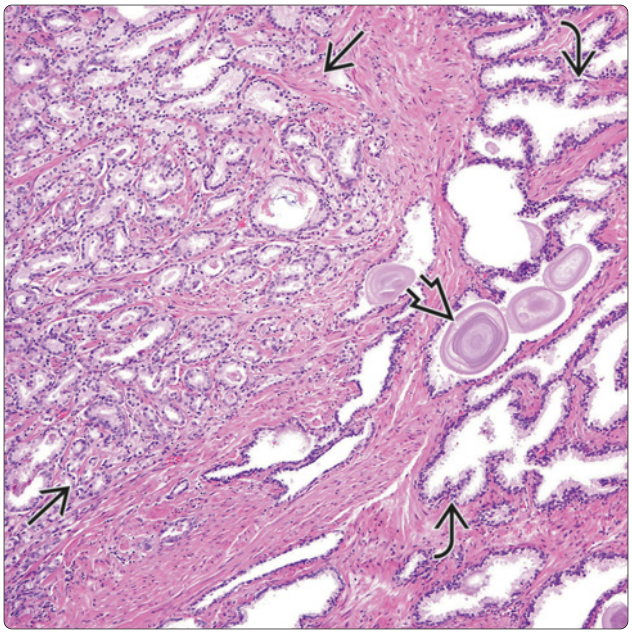
Higher magnification shows malignant glands ⇨ and total absence of staining with keratin antibody. The basal cell layer is intact (brown staining) in benign glands ⇨. (Original magnification, 600x)

T2b (Gleason Score 3 + 4 = 7)



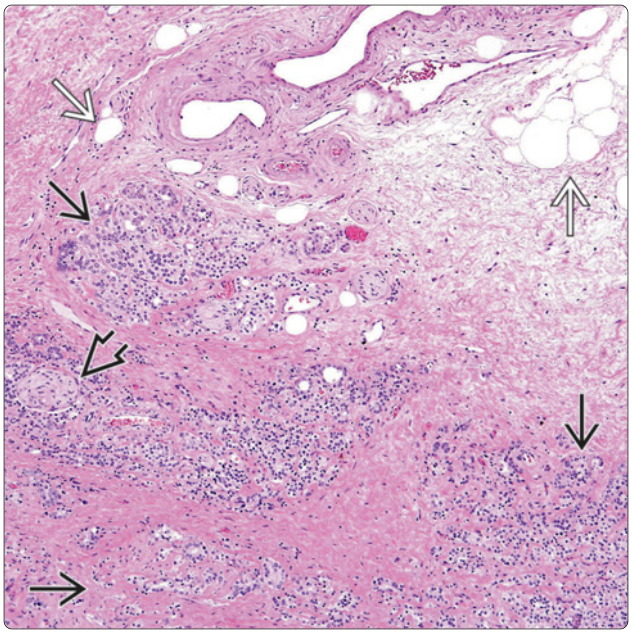
Low-power H&E-stained section from a prostatectomy specimen shows sheets of invasive prostatic adenocarcinoma ⇨ on the left. The adenocarcinoma is confined to the prostate tissue and is away from the prostatic capsule or the blue-inked resection margin ⇨. (Original magnification, 40x)

T2b (Gleason Score 3 + 4 = 7)



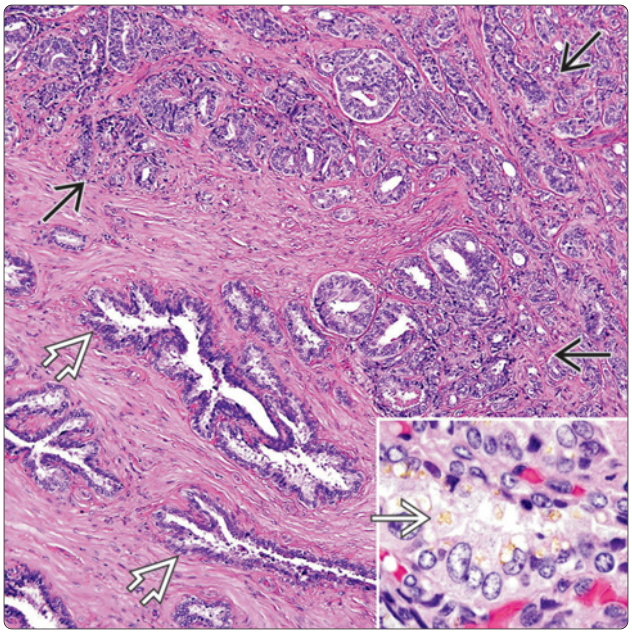
High-power view of the previous slide shows sheets of neoplastic glands in the left upper corner ⇨ and benign prostatic glands ⇨ with numerous pink round structures ⇨, (corpora amylacea) representing laminated concretions of prostatic secretions within glandular lumina. (Original magnification, 400x)

T3a (Gleason Score 4 + 3 = 7)



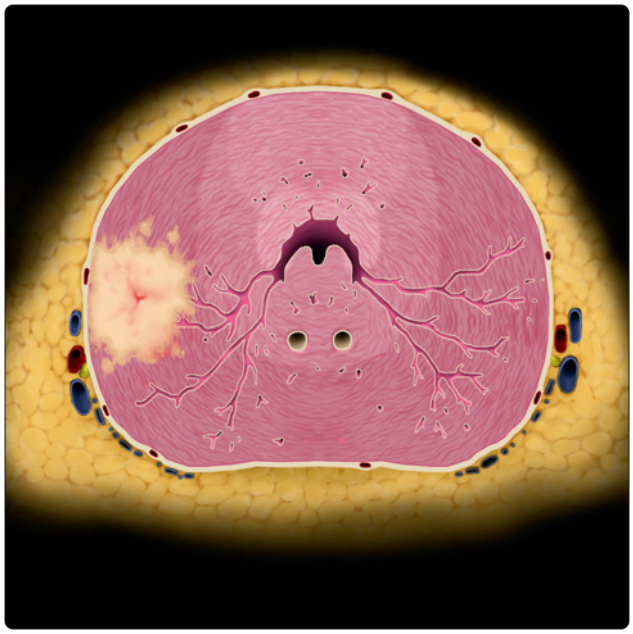
Low-power H&E stain from prostatectomy specimen shows the tumor ⇨ extending into the periprostatic tissue in proximity to the periprostatic fat ➡. Note also the perineural invasion (tumor surrounding nerve tissue) ⇨. (Original magnification, 40x)

T3b (Gleason Score 3 + 4 = 7)



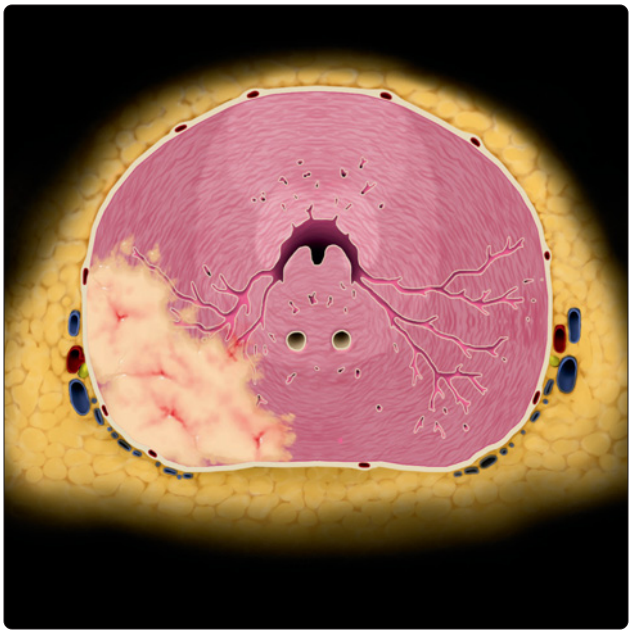
H&E stain of a tumor invading seminal vesicles shows the tumor ⇨ and seminal vesicle tissue ➡. (Original magnification, 400x) The inset highlights the golden yellow pigment ➡ (lipofuscin) that is characteristically found abundantly in seminal vesicle tissue.

T2a



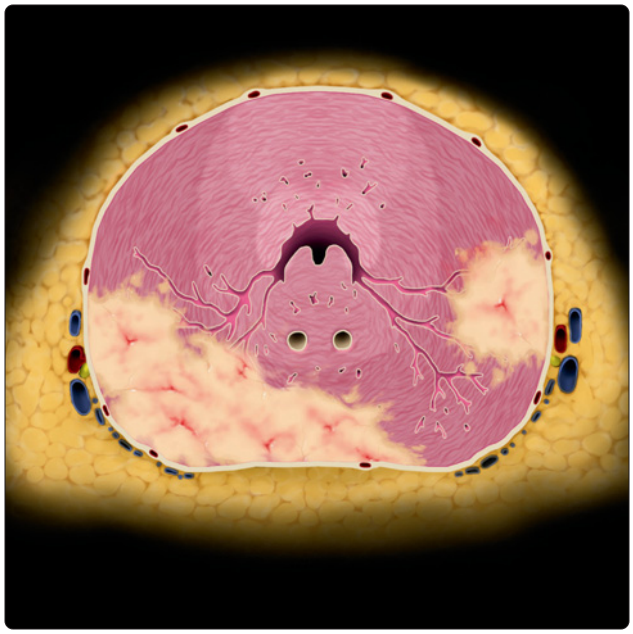
Axial graphic demonstrates a localized focus of peripheral zone tumor that involves less than 1/2 of 1 lobe of the prostate. This is consistent with T2a disease. T1 disease is neither clinically apparent nor visible on imaging.

T2b



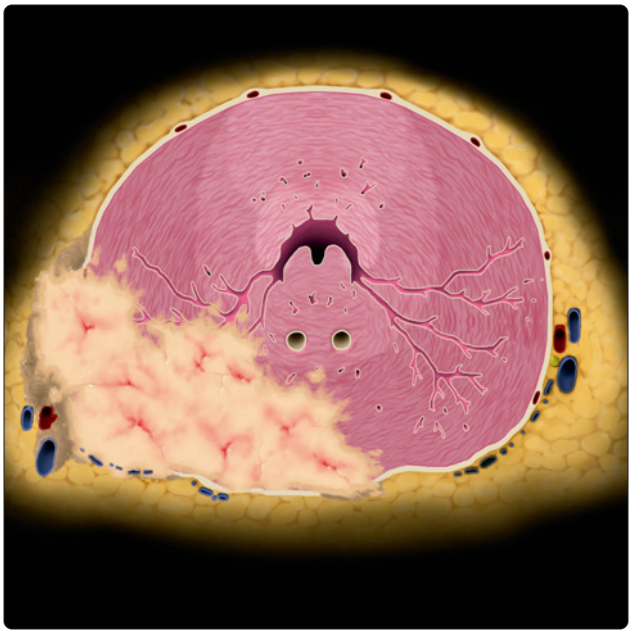
Axial graphic demonstrates a larger focus of peripheral zone tumor that involves more than 1/2 of 1 lobe of the prostate but does not cross the midline. The prostatic capsule is intact, and the neurovascular bundle is unaffected by tumor. This is consistent with T2b disease.

T2c



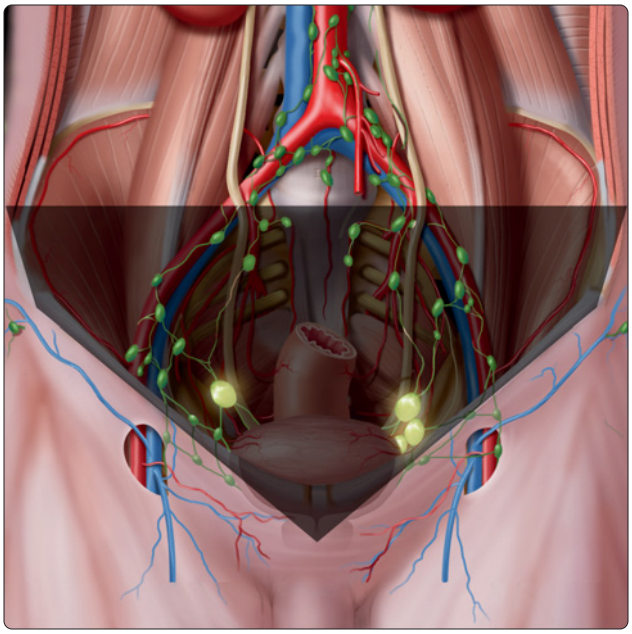
Axial graphic demonstrates a larger focus of peripheral zone tumor that involves more than 1/2 of 1 lobe of the prostate and crosses the midline. An additional focus of tumor is present on the contralateral side of the gland. Both of these findings fulfill the criteria for T2c disease.

T3a



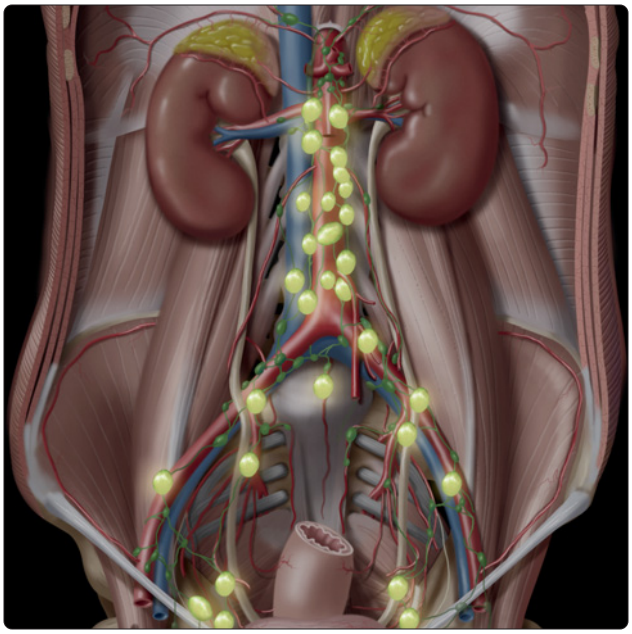
Axial graphic demonstrates a larger peripheral zone tumor with focal bulging of the prostatic contour posteriorly. Additionally, in the posterolateral margin, the capsule is invaded and tumor spills into the surrounding periprostatic fat and surrounds the neurovascular bundle. This is consistent with T3a disease.

N1



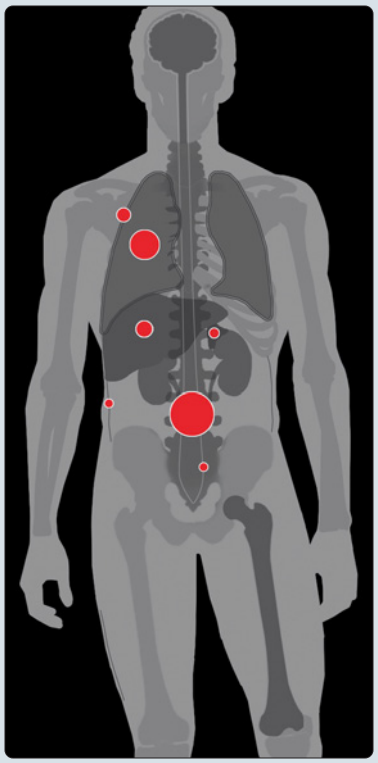
Coronal graphic shows regional lymph nodes shaded in black. Enlarged bilateral internal iliac nodes are present, indicating N1 disease. N1 disease is considered stage IV, regardless of the T stage.

M1a



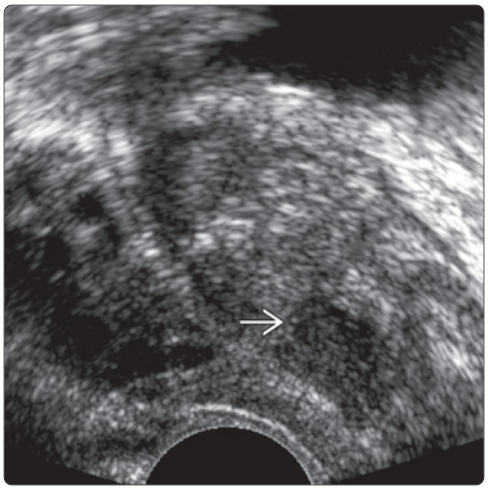
Coronal graphic shows enlarged regional lymph nodes in the pelvis and enlarged nonregional nodes in the common iliac and paraaortic lymph node stations. The presence of nonregional lymph node metastases upgrades disease to M1a.

Metastases, Organ Frequency

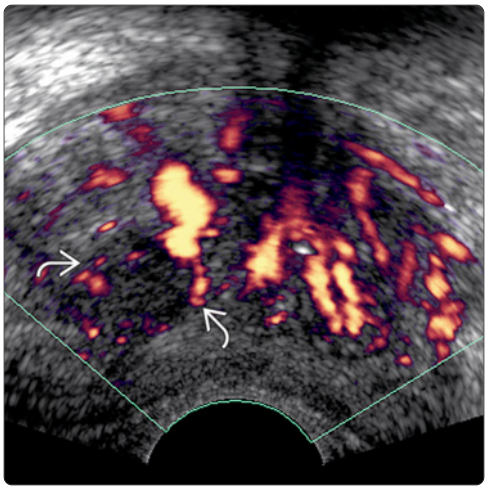


Bone	*90%*
Lung	*46%*
Liver	*25%*
Pleura	*21%*
Adrenal gland	*13%*
Peritoneum	*7%*
Meninges	*6%*

T2



T2

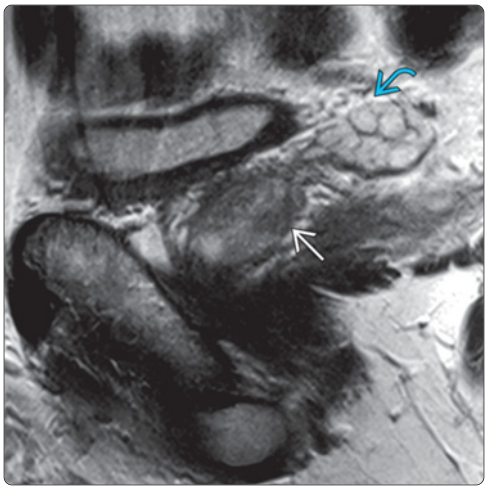


(Left) *Grayscale transrectal ultrasound (TRUS) shows an ill-defined, hypoechoic, heterogeneous area in the right peripheral zone. Tumor appears confined to the limits of the prostatic capsule. TRUS is not sensitive enough to exclude extracapsular extension (ECE) or seminal vesicle invasion.* **(Right)** *In the same patient, the transducer is rotated to bring the nodule to the left side of the image. This power Doppler ultrasound shows neovascularization around the mass. A guided biopsy confirmed prostatic carcinoma.*

Stage I (pT2a N0 M0, PSA 6, Gleason 6)



Stage I (pT2a N0 M0, PSA 6, Gleason 6)

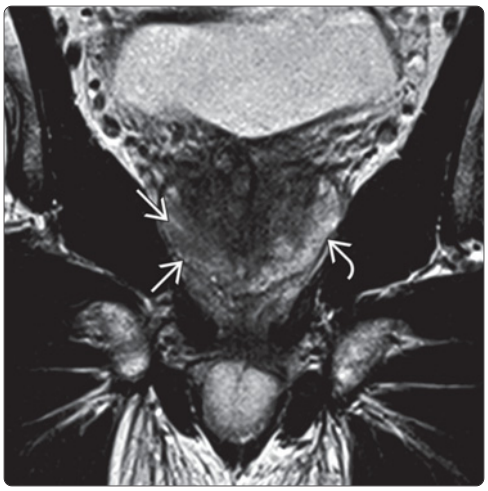


(Left) *Axial T2WI MR without ER coil at 3.0T demonstrates a 9-mm focus of low T2 SI in the right peripheral zone, with an intact, sharply demarcated T2 dark fibrous band overlying the abnormal focus, suggesting an intact capsule. The normal heterogeneously low T2 SI of the central gland is well demonstrated.* **(Right)** *Sagittal T2WI MR in the same patient shows the tumor focus, although not as well. Normal high T2 SI is present in the right seminal vesicle.*

Stage IIA (T2b N0 M0)

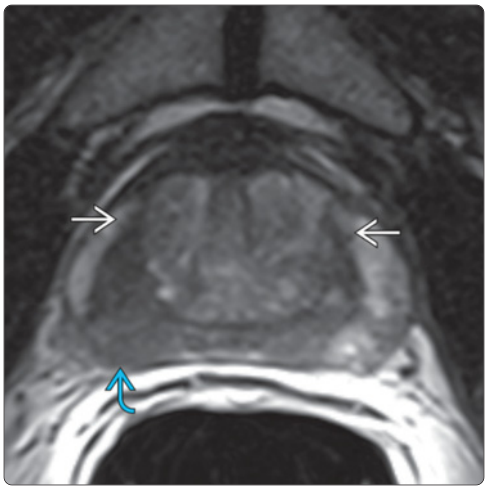


Stage IIA (T2b N0 M0)

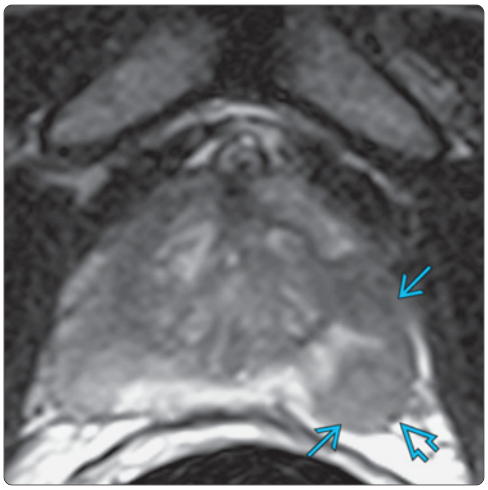


(Left) *Axial T2WI MR without ER coil at 3.0T demonstrates low T2 SI in the right peripheral zone (PZ) mid gland spanning from 7:00 to 10:30. This area spans more than 1/2 of the left lobe, with normal heterogeneously high T2 SI in the right PZ. An intact neurovascular bundle is present.* **(Right)** *Coronal T2WI MR from the same patient demonstrates the PZ asymmetry, with abnormal low T2 SI in the right PZ and normal high T2 SI in the left PZ.*

Stage IIB (T2c N0 M0)



Stage IIB (T2c N0 M0)

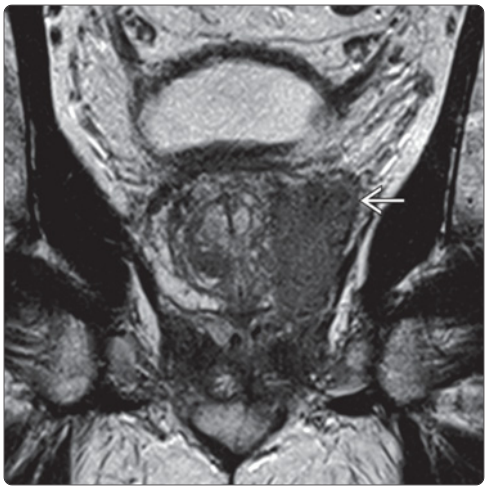


(Left) *Axial T2WI MR with ER coil at the prostate base shows a continuous band of low PZ T2 SI from 1:00 to 11:00. The band abuts both the prostate central zone (CZ) and the T2 dark fibrous capsule. The capsular margin appears intact.* **(Right)** *At the mid gland level in the same patient, a more focal nodule of disease is present on the left with a bulging rectoprostatic angle. This is concerning for ECE. Right-sided low-SI tumor is also seen at this level.*

Stage III (pT3a N0 M0, PSA 6, Gleason 7)

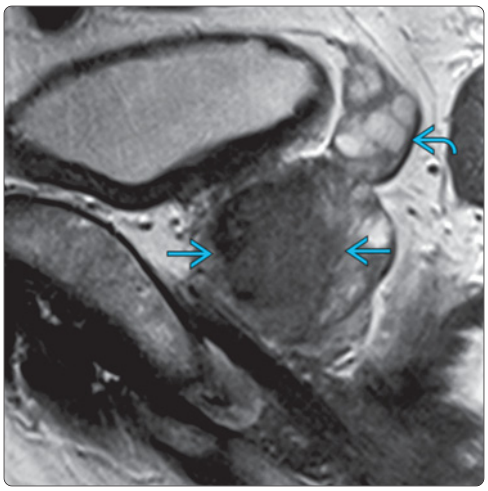


Stage III (pT3a N0 M0, PSA 6, Gleason 7)

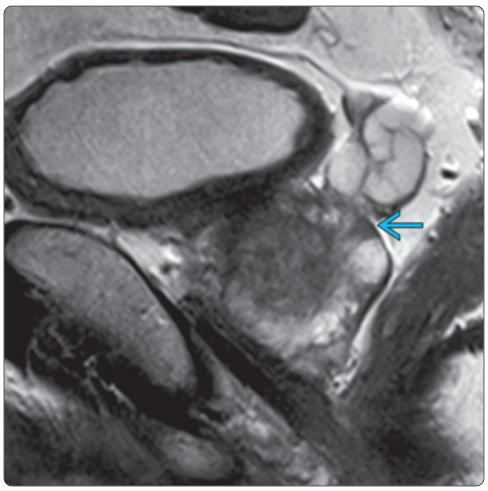


(Left) *Axial T2WI MR without ER coil demonstrates large, homogeneously low T2 SI tumor in the left gland from 2:00 to 4:00. The mass involves the CZ and extends beyond the prostatic capsule. Because the bulk of the tumor is in the anterior portion of the gland, the neurovascular bundle may be spared.* **(Right)** *Coronal T2WI MR shows the focal bulge at the lateral base well. This is consistent with ECE, which was confirmed at resection.*

Stage III (pT3a N0 M0, PSA 6, Gleason 7)

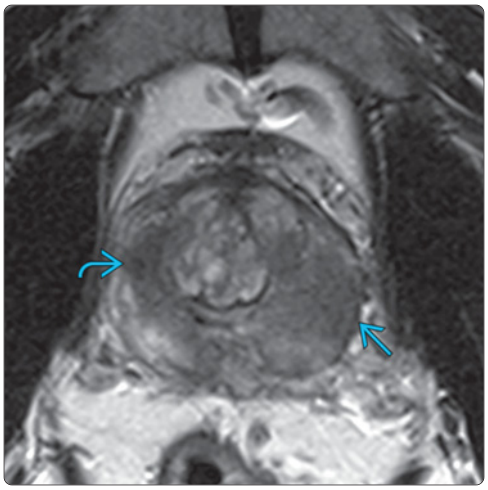


Stage III (pT3a N0 M0, PSA 6, Gleason 7)

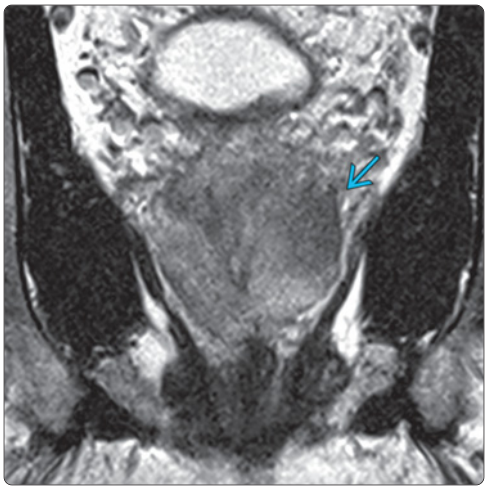


(Left) *Sagittal T2WI MR in the same patient shows the focal low T2 SI area corresponding to tumor on the left. No definite low SI is present in the seminal vesicle. The bulk of the tumor is anterior in the prostate, decreasing the likelihood of direct seminal vesicle invasion.* **(Right)** *Sagittal T2WI MR on the right in the same patient shows the heterogeneously high PZ T2 SI. A focus of low T2 SI near the prostate base is concerning for an additional site of disease.*

Stage III (T3a N0 M0)



Stage III (T3a N0 M0)

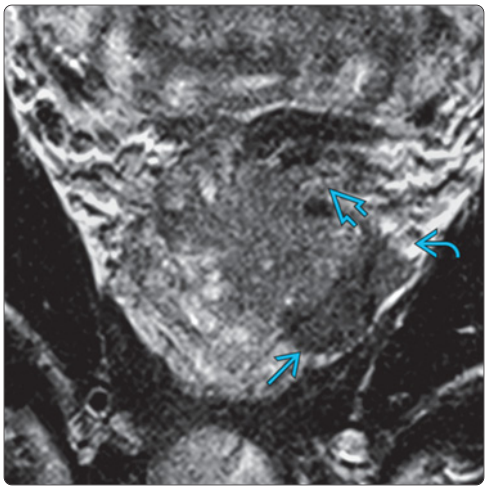


(Left) *Axial T2WI MR shows diffuse low T2 SI throughout the PZ on the left, with a rim of abnormal signal in the right transition zone. The capsule appears intact, but the low T2 SI area in the PZ contacts > 10 mm of the prostate margin.* **(Right)** *Coronal T2WI MR in the same patient shows the left-sided low T2 SI nodule with focal bulging and irregularity at the lateral base. This is evidence of unilateral ECE. Seminal vesicles were uninvolved.*

Stage III (T3b N0 M0)

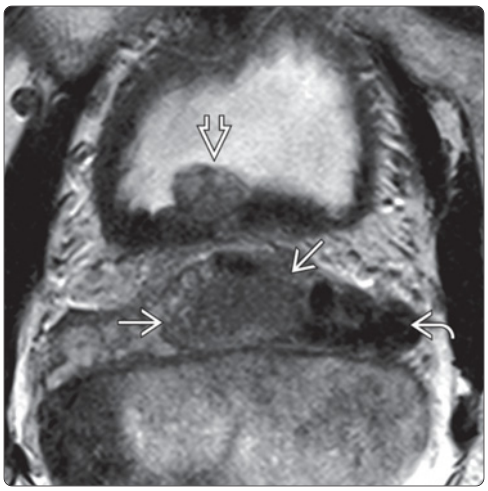


Stage III (T3b N0 M0)

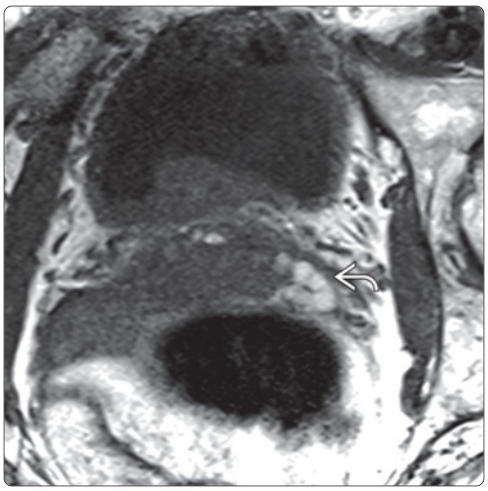


(Left) *Axial T2WI MR shows a focal bulging low T2 SI mass in the left PZ. This is palpable on digital rectal exam as a firm nodule.* **(Right)** *Coronal T2WI MR in the same patient shows the focal area of very low T2 SI tumor extending from the left mid gland to the left base. ECE is seen at the base, where the tumor extends beyond the capsular margin. Seminal vesicle invasion is present with heterogeneously low T2 SI extending into the seminal vesicle.*

Stage III (T3b N0 M0)



Stage III (T3b N0 M0)

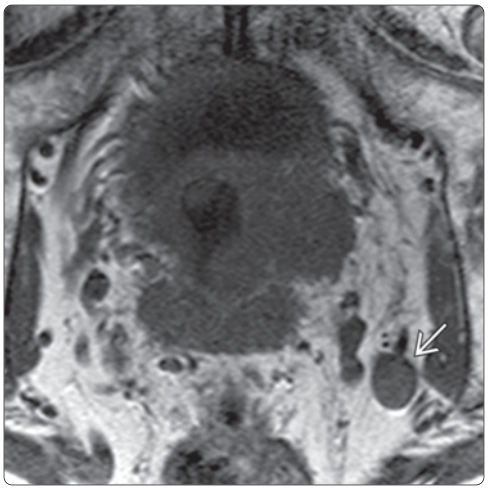


(Left) *Axial T2WI more cranially at the level of the prostate base in the same patient shows the low T2 SI extending into the seminal vesicles related to patient's BPH. The tubules of the seminal vesicles on the left demonstrate very dark T2 SI. A prostatic nodule from patient's BPH protrudes into the bladder base.* **(Right)** *Axial T1WI MR at the same level confirms that the low T2 signal in the seminal vesicles on the left is due to hemorrhage, which is T1 bright.*

Stage IV (T3b N1 M0)

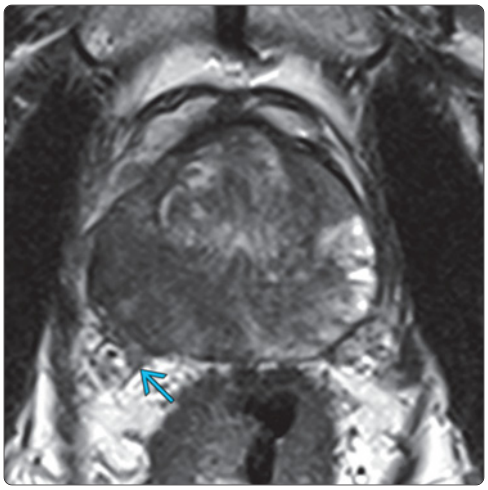


Stage IV (T3b N1 M0)

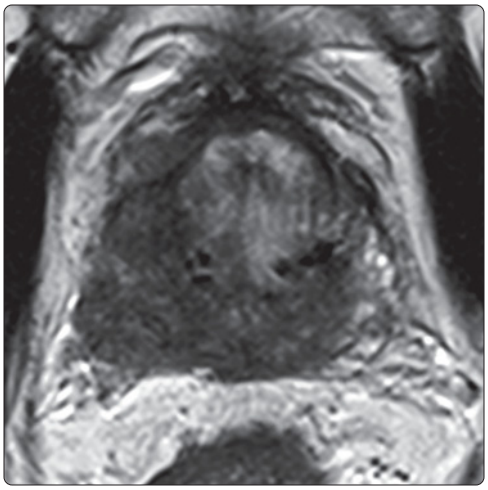


(Left) *Coronal T2WI MR in a patient who had previously undergone a transurethral resection of the prostate (TURP) shows a large heterogeneous low T2 SI mass extending from the left side of the remaining prostate into the periprostatic fat, encasing vessels. The surgical defect from the TURP is noted.* **(Right)** *Axial T1WI MR in the same patient shows an enlarged left obturator node, making this N1 disease.*

Stage IV (T3b N1 M0, PSA 22, Gleason 9)

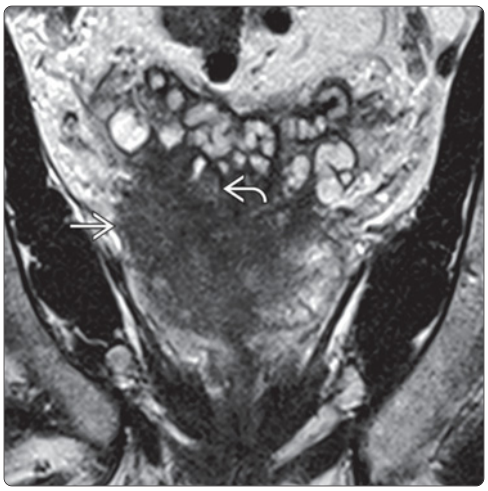


Stage IV (T3b N1 M0, PSA 22, Gleason 9)

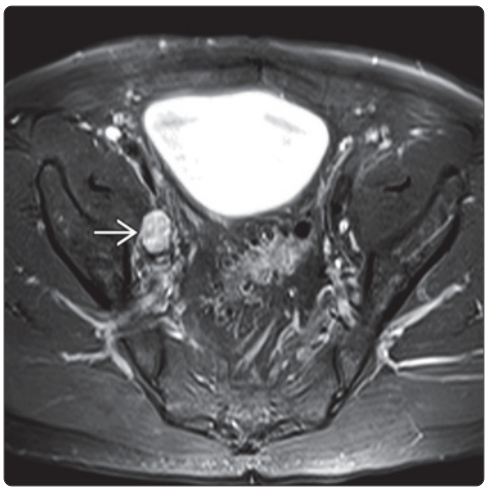


(Left) *Axial T2WI MR at a mid gland level shows diffuse low T2 SI throughout the PZ. The margin between the PZ and CZ is obscured. The asymmetric bulge on the right, with low T2 SI infiltrating into the region of the neurovascular bundle, is consistent with ECE and neurovascular bundle involvement.* **(Right)** *Axial T2WI MR shows mass-like confluent low T2 SI involving nearly the entire base of the prostate, with definite angularity at the right prostatic angle, evidence of ECE.*

Stage IV (T3b N1 M0, PSA 22, Gleason 9)



Stage IV (T3b N1 M0, PSA 22, Gleason 9)

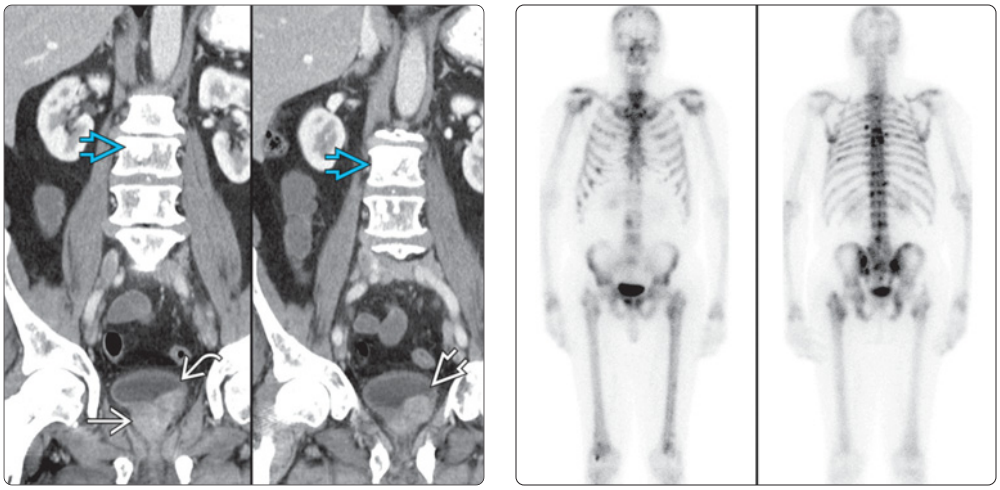


(Left) *Coronal T2WI MR in the same patient shows the abnormal low T2 SI infiltrating from the left side of prostate into the left seminal vesicles, consistent with seminal vesicle invasion.* **(Right)** *Axial T2WI FS MR in the same patient shows an abnormally enlarged right external iliac node. The metastasis to a regional lymph node is N1 disease. Surgery is not indicated for this stage of disease.*

Stage IV (T4 N0 M1b)

Stage IV (T4 N0 M1b)

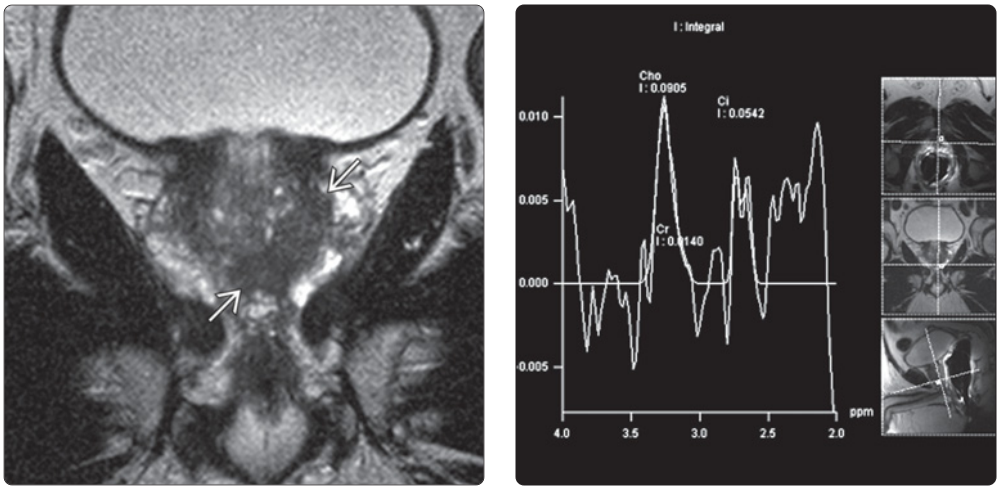
(Left) *Coronal CECT images demonstrate a mass extending from the prostate into the bladder base. The right image shows the intraluminal bladder component well. Sclerotic lesions are seen in the lumbar spine.* **(Right)** *Whole-body bone scan in the same patient shows diffuse increased uptake in the spine and bony pelvis, as well as foci of increased uptake in many ribs. With little renal or soft tissue activity, this study approaches a "superscan."*



Stage IV (T2b N0 M1b, PSA 16.5, Gleason 7)

Stage IV (T2b N0 M1b, PSA 16.5, Gleason 7)

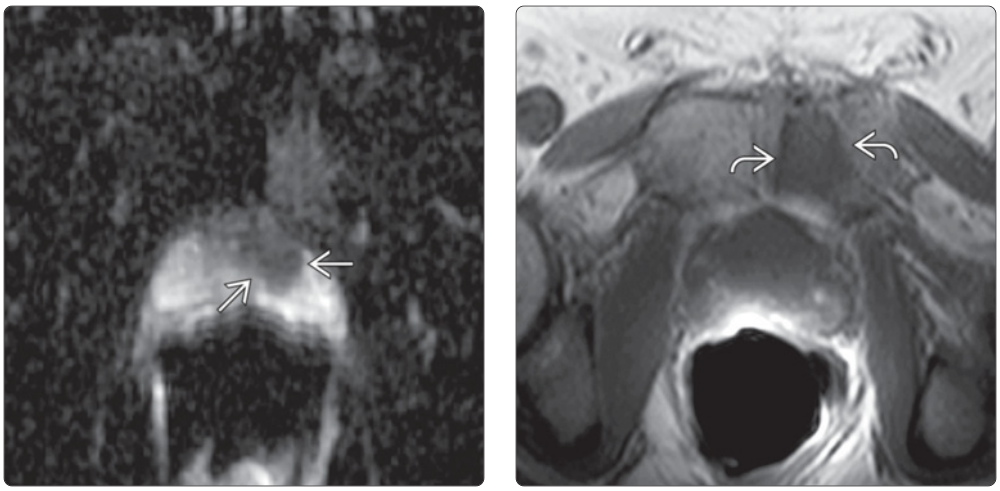
(Left) *Coronal T2WI MR shows a band of low T2 SI extending from the apex to base on the left. (Courtesy S. Eberhardt, MD.)* **(Right)** *Single-voxel MR spectroscopy was performed in the same patient by placing a voxel over the area of tumor. This demonstrates a metabolic spectra of elevated choline + creatine/citrate. This spectra is consistent with malignancy. (Courtesy S. Eberhardt, MD.)*



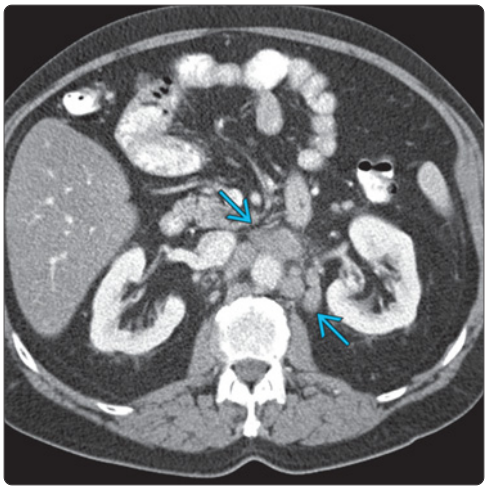
Stage IV (T2b N0 M1b, PSA 16.5, Gleason 7)

Stage IV (T2b N0 M1b, PSA 16.5, Gleason 7)

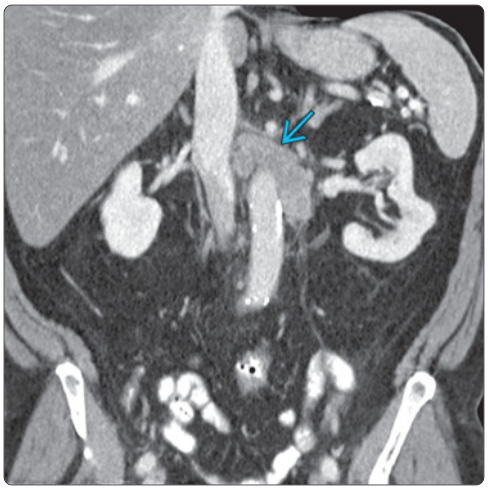
(Left) *Axial ADC map from diffusion-weighted imaging in the same patient shows restricted diffusion corresponding to the area of decreased T2 SI. (Courtesy S. Eberhardt, MD.)* **(Right)** *Coned-down axial T1WI MR from the same patient demonstrates a focus of low T1 SI in the left pubis. This is consistent with an osteoblastic bony metastasis. (Courtesy S. Eberhardt, MD.)*



Stage IV (TX N1 M1a)

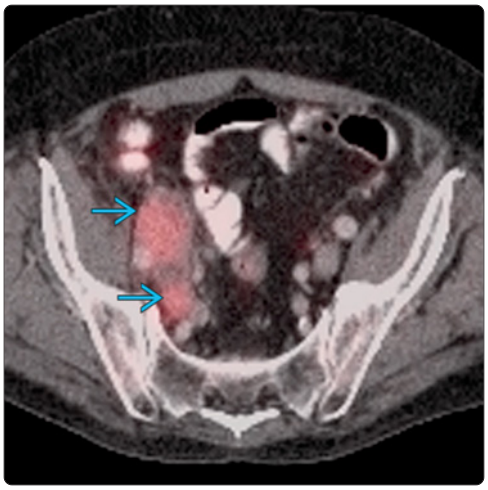


Stage IV (TX N1 M1a)

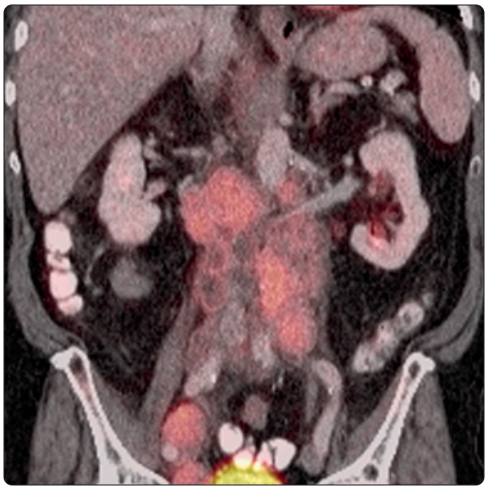


(Left) *Axial CECT at the level of the right renal vein demonstrates significant periaortic lymphadenopathy* ⇨. **(Right)** *Coronal CECT in the same patient demonstrates the confluent adenopathy* ⇨ *crossing anterior to the aorta and between the aorta and inferior vena cava. Metastases to the paraaortic lymph nodes are the most common site of nonregional lymphatic spread of disease. These upstage the patient to M1a, while pelvic regional adenopathy is N1 disease.*

Stage IV (TX N1 M1a)

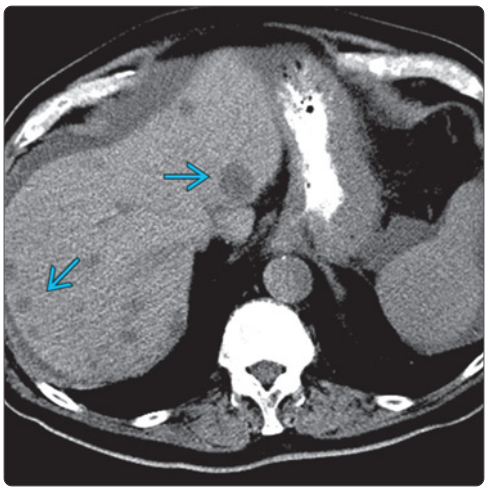


Stage IV (TX N1 M1a)

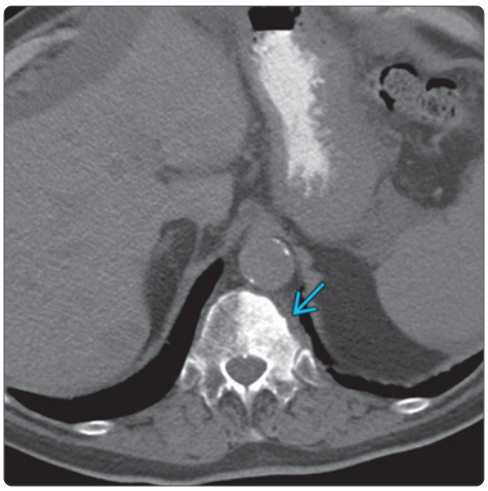


(Left) *Axial fused PET/CT in the same patient shows the internal and external iliac adenopathy on the right* ⇨. *This regional adenopathy is considered N1 disease.* **(Right)** *Coronal fused PET/CT in the same patient shows the extensive regional and distant lymphadenopathy. No bone lesions were identified on the PET.*

Stage IV (TX NX M1c)



Stage IV (TX NX M1c)

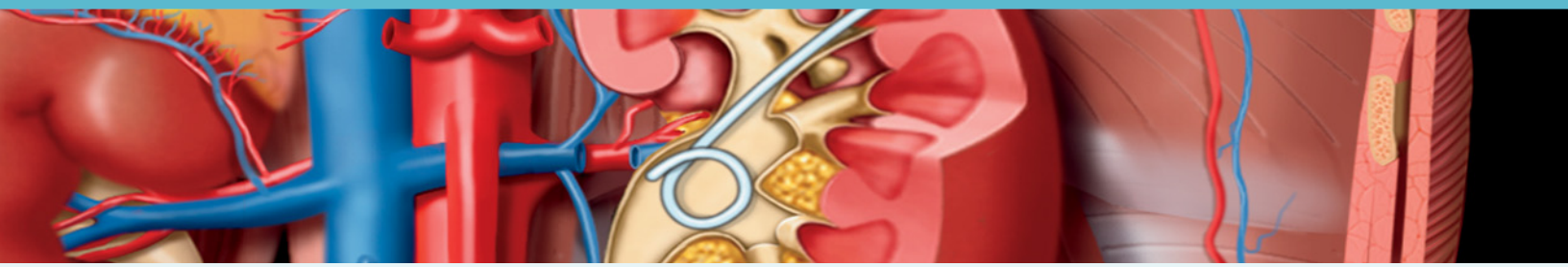


(Left) *Axial NECT in a patient with a history of prostate cancer, treated more than 10 years prior who had a CT of his abdomen after abnormal liver function tests were found, shows multiple hypoattenuating hepatic lesions* ⇨. *On percutaneous CT-guided biopsy, these were proven to be metastases from prostate cancer.* **(Right)** *Axial NECT shows that, in addition to the hepatic lesions, a sclerotic bone lesion in the T10 vertebral body* ⇨ *is identified.*

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SECTION 13

Procedures



Percutaneous Genitourinary Interventions	**554**
Kidney Ablation/Embolization	**566**
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Venous Sampling and Venography (Renal and Adrenal)	**590**
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KEY FACTS

PREPROCEDURE

- Indications
  - Upper tract decompression
  - Nephrolithotomy
  - Urinary diversion
  - Whitaker test for evaluation of obstruction
- Preprocedure imaging
  - Assess degree of hydronephrosis
  - Evaluate position/anatomy of kidney

PROCEDURE

- US-guided initial access in hydronephrosis
- Direct fluoroscopic guidance can be used for
  - Retrograde contrast injection
  - Target calculus or indwelling stent
  - IV contrast ± Lasix (helpful in nondilated system)
  - Direct puncture using anatomic landmarks
- 1-puncture technique if targeted calyx visualized/accessed at outset of procedure; otherwise, 2-puncture technique
- Avascular plane of Brödel: 20-30° off sagittal plane
  - Decreases likelihood of vascular injury

POST PROCEDURE

- Technical success rate in hydronephrosis: 95-98%
  - 80-85% success in nondilated systems
- Transient hematuria common after placement
  - Often resolves within 24-48 hours

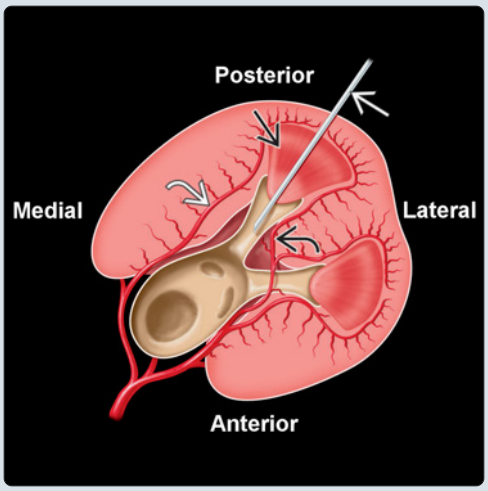
OUTCOMES

- Complications
  - Vascular injury and gross hematuria
  - Consider angiography/embolization if persists
  - Bowel transgression with suprapubic tube
- Follow-up imaging for urolithiasis can include
  - Abdominal and pelvic radiography
  - Ultrasound (US)
  - Nonenhanced CT (NECT)

Percutaneous Nephrostomy

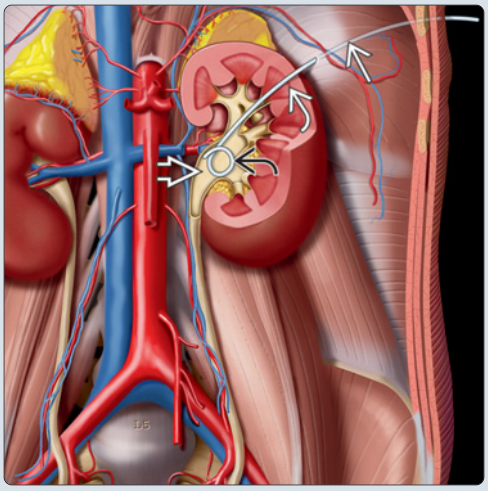


Preferred Needle Position (Avascular Plane of Brödel)

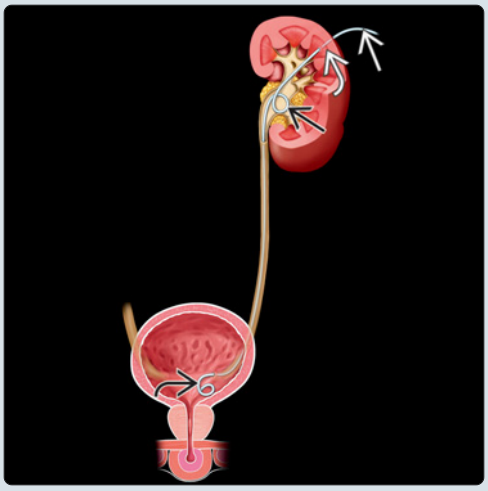


(Left) *Clinical photograph shows the typical sagittal position of the US probe during percutaneous nephrostomy. The US probe is cloaked in a sterile cover for use in the sterile field.* **(Right)** *Graphic shows a needle entering the renal collecting system via a posterior calyx along the relatively avascular plane of Brödel. This plane is located between the anterior & posterior divisions of the renal artery & lies 20-30° from the sagittal plane. Access via this plane minimizes bleeding and potential vascular injury.*

Nephrostomy Catheter



Nephroureteral Catheter



(Left) *Graphic shows that a nephrostomy catheter has been percutaneously introduced into the left renal collecting system via a superior pole calyx. The pigtail coil of the catheter is positioned in the renal pelvis.* **(Right)** *Graphic shows that a nephroureteral catheter has been percutaneously placed into the left renal collecting system via a posterior calyx; the distal pigtail coil of the catheter is in the bladder, and the proximal pigtail is in the renal pelvis.*

TERMINOLOGY

Definitions

- **Percutaneous nephrostomy (PCN)**: External drainage catheter placement into renal collecting system
- **Nephroureteral catheter**: PCN with catheter also extending from renal collecting system to bladder
- **Ureteral stent**: Internalized catheter extending from renal pelvis to bladder
- **Suprapubic cystostomy (SPT)**: Percutaneous drainage catheter that enters bladder superior to pubic symphysis

PREPROCEDURE

Indications

- Obstructive hydronephrosis/pyonephrosis
- Provide percutaneous access for
  - Antegrade ureteral stent placement
  - Percutaneous nephrolithotomy
    - Renal pelvis calculi > 2 cm
    - Staghorn calculus
    - Inferior pole calculi > 1-2 cm
  - Endopyelotomy
  - Administration of antifungal agents
  - Foreign body retrieval
- Urinary diversion in cases of
  - Ureteral fistulas
  - Hemorrhagic cystitis
  - Ureteral injury and urinoma
    - Urinoma requires separate drain
- Whitaker test
  - Gold standard in evaluating urinary obstruction between renal pelvis & bladder such as
    - Pediatric hydronephrosis/hydroureter
      - In absence of vesicoureteral reflux
    - Hydronephrosis/hydroureter
      - Persisting despite urologic intervention (e.g., ureteral reimplantation, pyeloplasty)
      - With inconclusive noninvasive studies (e.g., Lasix renogram)
- Suprapubic cystostomy
  - Bladder outlet obstruction
    - Prostate disease
  - Urinary incontinence
  - Neurogenic bladder
  - Fistula involving bladder
    - e.g., vesicocolic, vesicouterine, vesicovaginal
  - Urethral trauma

Contraindications

- Uncorrectable coagulopathy

Preprocedure Imaging

- US or CT
  - Assess presence & degree of hydronephrosis
  - Evaluate position of kidney & optimal approach
  - Normal anatomy
    - Superior pole usually more medial than inferior pole
    - Kidneys oriented 30-50° to coronal plane
    - Average: ~ 14 calyces in each kidney
      - Anterior calyces project side-on; appear cup-like
      - Posterior calyces project end-on; appear circular
    - Posterior plane of Brödel
      - Relatively avascular plane between anterior/posterior divisions of renal artery
      - Lies 20-30° from sagittal plane
      - Reduced bleeding risk if used for needle entry
  - Is there an anatomic variant?
    - e.g., horseshoe and malrotated kidneys
  - Transplant kidney
    - Right or left iliac fossa location

Getting Started

- Preprocedural considerations
  - Pertinent clinical history & physical examination
    - Procedure indications
    - Current medications
      - Any anticoagulant or antiplatelet agents
    - Allergies
    - Is patient uroseptic?
      - Appropriate antibiotic coverage
      - Hemodynamic support as necessary
      - May require anesthesiology assistance
  - Laboratory parameters
    - Complete blood count (CBC)
      - Platelet > 50,000/μL
    - Coagulation profile
      - International normalized ratio (INR) ≤ 1.5
      - Normal prothrombin time (PT), partial thromboplastin time (PTT)
    - Serum electrolytes
      - Correct any renal failure-associated hyperkalemia
- Medications
  - Anesthesia
    - Local anesthetic: 1% lidocaine
    - Procedural sedation (if hemodynamically stable)
      - Typically fentanyl, midazolam
    - Low threshold for anesthesiology assistance
      - Due to risk of sepsis/septic shock during procedure
  - Prophylactic antibiotics
    - Many possibilities
      - Ampicillin (1 g) & gentamicin (80 mg)
      - Levaquin (500 mg)
      - Other agents, based on Gram stain/culture data
- Equipment list
  - **General**
    - Ultrasound equipment
      - 3.5 MHz transducer
      - Sterile ultrasound probe cover/gel
    - Fluoroscopy unit
    - CT equipment
      - Radiopaque grid
  - **1-puncture technique**
    - 21-22-gauge Chiba needle
    - 0.018 inch mandril or nitinol guidewire
    - 5-6 Fr coaxial dilator-sheath assembly
    - 0.035 inch 3-J guidewire
    - 5 Fr directional catheter (e.g., Kumpe catheter)
    - 8-10 Fr dilator as appropriate
    - 8-10 Fr locking pigtail catheter (PCN)
  - **2-puncture technique**

- Same materials as for 1-puncture technique
- 18-gauge Longdwell needle (e.g., Ring needle)

- **Nephroureteral catheter**
  - 0.035 inch Amplatz guidewire
  - 8 Fr 20-24 cm nephroureteral catheter
- **Ureteral stenting**
  - 0.035 inch Amplatz guidewire
  - 9-10 Fr peel-away sheath
  - 8 Fr 20-24 cm double-J ureteral stent
- **Whitaker test**
  - 18- to 22-gauge needle
  - Manometer or pressure transducer
  - Power injector/infusion pump
  - Bladder catheter (e.g., Foley)
  - 2 individual 3-way stopcocks
  - Connecting tubing
- **Suprapubic cystostomy**
  - Transurethral catheter if possible
    - 19-gauge needle if not possible
  - **Trocar technique**
    - Foley catheter (8-16 Fr)
    - Stiffening inner cannula
    - Sharp trocar
  - Seldinger technique
    - Longdwell needle
    - 0.035 inch Amplatz wire
    - Sequential dilators, as appropriate
    - 8-14 Fr locking pigtail catheter

PROCEDURE

Patient Position/Location

- Best procedure approach
  - **General**
    - Avoid injury to renal vessels, viscera, pleura
      - Subcostal approach preferred
    - Review prior imaging
      - Renal anatomy
      - Possible interposed viscera (e.g., spleen, colon)
      - Structural relationships may change when patient prone vs. supine
  - **Patient position**
    - Native kidney
      - Prone position preferred
      - Prone-oblique (ipsilateral side up) if unable to lie prone or if pregnant
    - Transplant kidney
      - Supine
  - **Needle entry**
    - Access via avascular plane of Brödel: Requires positioning 20-30° off sagittal plane using
      - Ultrasound
      - C-arm fluoroscopy
      - Fluoroscopy with wedge under ipsilateral side
    - Inferior pole posterior calyx generally used for external drainage
    - Mid or superior pole access may facilitate negotiation to ureteropelvic junction
      - Helpful for endoureteral interventions
      - May require intercostal approach; risks traversing pleura
    - Transplant kidney
      - Consider lateral approach; reduced risk of transperitoneal puncture
    - Nephrolithotomy
      - Select calyx providing best access to stone(s)
  - **Suprapubic cystostomy**
    - Supine
    - Midline or paramedian access above pubic symphysis
      - Avoid lateral approach
      - May injure inferior epigastric vessels

Procedure Steps

- **General**
  - Imaging guidance for initial access depends on clinical scenario
    - Ultrasound-guided access in hydronephrosis
    - Direct fluoroscopic guidance can be used with
      - Intravenous nephrogram with IV contrast ± Lasix (helpful in nondilated system)
      - Target visible stone/proximal pigtail of nonfunctioning ureteral stent
      - Retrograde contrast injection of cystoscopically placed externalized ureteral stent
      - Direct puncture using anatomic landmarks (review prior imaging)
    - CT-guided access (rarely used)
      - May be helpful in malrotated or horseshoe kidney, severe kyphoscoliosis
      - Also consider if high risk of injuring interposed viscera when US/fluoroscopy used
  - Discuss best nephrolithotomy access with urologist
  - Cleanse skin with preparatory solution
  - Place sterile drapes to isolate sterile field
- **Ultrasound-guided initial access**
  - **General**
    - Place sterile cover on transducer
      - 1st apply gel on ultrasound probe
      - Apply sterile gel externally over probe cover
  - **1-puncture technique**
    - Image posterior lower pole calyx with US
    - Administer 1% lidocaine as local anesthetic
    - 2-3 mm skin incision with #11 blade scalpel
    - Advance Chiba needle into calyx under US
    - Remove needle stylet; aspirate urine
      - Aspirate urine volume > planned contrast volume; reduces risk of overdistention that may aggravate bacteremia/sepsis
      - Send urine sample for Gram stain/culture
    - Inject contrast; perform antegrade nephrostogram
      - Assess needle entry site into collecting system
      - Prefer papillary tip or fornix of calyx
      - Avoid infundibular access; risk of vascular injury
      - Transition to 2-puncture technique if initial puncture is suboptimal
    - Advance 0.018 inch guidewire into renal collecting system using fluoroscopy guidance
    - Exchange needle for 5-6 Fr dilator-sheath assembly

- Once assembly in collecting system, unscrew & advance sheath over guidewire
- Remove guidewire & dilator

– Place 0.035" 3-J wire in renal collecting system
– Exchange sheath for 5 Fr directional catheter
- Negotiate guidewire & catheter into renal pelvis or proximal ureter for stability

– Dilate tract as appropriate
– Load pigtail catheter onto stiffening cannula
– Advance into renal collecting system
– Unscrew inner stiffener; stabilize stiffener so it does not advance with catheter
- Advance catheter further into collecting system

– Remove stiffener and guidewire
– Form/lock pigtail; position catheter in renal pelvis
– Confirm position with contrast injection

o **2-puncture technique**
– Use ultrasound to determine target of initial puncture
- Posterior calyx of inferior pole often used but not necessary; operator preference

– **1st puncture**: For antegrade nephrostogram
- Performed similarly to 1-puncture technique
- After gaining access, obtain antegrade nephrostogram; use to plan 2nd puncture

– **2nd puncture**: For PCN placement
- Select desired calyx (e.g., posterior lower pole)
- May inject air/CO_2 to better delineate posterior calyces (antidependent in prone patient)

– Rotate C-arm 20-30° from sagittal plane toward ipsilateral side
- Facilitates access via plane of Brödel
- If C-arm unavailable, place wedge under ipsilateral side for positioning

– Select skin entry site, which overlies selected calyx
– Administer 1% lidocaine as local anesthetic
– 2-3 mm skin incision with #11 blade scalpel
– Advance 18-gauge Longdwell needle along fluoroscopic axis
- If in renal parenchyma, needle usually moves with diaphragmatic excursion

– Return C-arm to posteroanterior orientation
– Advance needle into calyx
- May see "tenting" of contrast-filled calyx as needle indents calyceal margin

– Advance sheath slightly & remove needle
– In obstructed systems, may see efflux of urine
– If no urine aspirated, advance 0.035 inch 3-J guidewire into sheath
- Contrast injection of sheath discouraged; any extracalyceal contrast may obscure visualization

– Slowly retract sheath while alternatively advancing/retracting guidewire until it coils in space conforming to renal collecting system
– Readvance sheath over guidewire
– Remove guidewire from sheath
- Perform contrast injection to confirm location

– Reinsert 3-J guidewire; exchange sheath for 5 Fr directional catheter
– Negotiate wire/catheter into renal pelvis/proximal ureter
– Dilate tract as appropriate
– Advance 8 Fr locking pigtail catheter loaded onto stiffening cannula into collecting system
– Unscrew inner stiffener; stabilize stiffener so it does not advance with catheter
- Advance catheter further into collecting system

– Remove stiffener & guidewire
– Form/lock pigtail; position catheter in renal pelvis
– Confirm position with contrast injection

• **Fluoroscopically-guided initial access**
o Ultrasound guidance may not be possible or needed in following scenarios
– Nondilated system
– Externalized cystoscopically placed ureteral stent for nephrolithotomy

o Can use fluoroscopic guidance for access if able to adequately opacify renal collecting system
– Administer IV contrast ± Lasix
- Need sufficient renal function for contrast excretion/opacification of collecting system
- Advance 21- or 22-gauge needle as per 1st stage of 2-puncture technique
- 1st access necessary to allow continued opacification of system via injection of needle, as IV contrast-induced opacification is transient
- Once initial access obtained, proceed to 2nd stage of 2-puncture technique

– Retrograde injection of externalized ureteral stent
- Usually performed in setting of nephrolithotomy
- Procedure performed as per 2nd stage of 2-puncture technique
- 2nd puncture directed at chosen calyx as discussed with urologist

o Fluoroscopically guided access without renal collecting system opacification
– Access collecting system using anatomic landmarks
- Review prior CT for renal pelvis location
- Place 21- or 22-g Chiba needle vertically into renal pelvis using posterior approach

– Visible stone or nonfunctioning ureteral stent
- Direct puncture using 21- to 22-g Chiba needle directed at stone/proximal pigtail of stent

– Once access obtained, inject contrast for antegrade nephrostogram
– Proceed with 2nd stage of 2-puncture technique

• **Nephroureteral catheter placement**
o May be preferred in
– Ureteral stenosis/injury
– Patients prone to dislodgement of PCN

o In septic patient may place PCN 1st; delay nephroureteral catheter until improved clinically
o Nephrostomy access performed as above
o 3-J wire is coiled in renal pelvis or ureter
o Using 5 Fr directional catheter, negotiate wire & catheter through ureter into bladder
– Exchange for hydrophilic guidewire if unable to cross an area of stenosis
– 4 Fr glidecatheter may also be helpful
– May consider leaving PCN for several days to allow inflammation to improve

- Remove guidewire
- Perform contrast injection to confirm location
- Advance 0.035 inch Amplatz guidewire into urinary bladder; remove catheter
- Dilate subcutaneous tract as needed
- Choose appropriate nephroureteral catheter
  - Catheter has pigtail at urinary bladder end; 2nd pigtail situated in renal pelvis
  - 8-10 Fr catheter; lengths of 20-28 cm (distance between pigtails)
  - May estimate length based on patient height
- Load catheter onto stiffening cannula & advance over guidewire into urinary bladder
- Retract wire so that tip is just within catheter
- Unscrew inner cannula at hub
- Withdraw guidewire and inner cannula slowly
  - Pigtail will form in urinary bladder
- Remove guidewire and cannula
- Form 2nd pigtail by pulling on string, positioning it in renal pelvis
  - Slowly withdraw catheter if proximal pigtail is located in ureter
- Lock pigtail & perform contrast injection to confirm location

- **Antegrade ureteral stent placement**
  - Same steps as nephroureteral catheter placement until Amplatz wire extends to bladder
  - Advance 9-10 Fr peel-away sheath over wire into kidney
  - Prepare ureteral stent
    - Select appropriate 8 Fr double pigtail stent
      - Lengths of 20-28 cm
    - Load stent onto stiffening cannula
    - Attach pusher to proximal end
    - Straighten string attached at proximal end
  - Advance stent so distal tip extends well into bladder
    - Radiopaque marker on distal aspect of pusher should be in renal pelvis
  - Retract guidewire so tip is just within stent
  - Unscrew inner cannula
  - Withdraw inner cannula and wire as unit
    - Distal pigtail will form in urinary bladder
  - Slowly retract wire/inner cannula to proximal ureter
  - If stent too far distal (i.e., proximal pigtail in ureter)
    - Advance guidewire
    - Pull gently on string/assembly to retract stent
  - Maintain wire and cannula position in proximal stent; keep forward pressure on pusher
    - Cut and remove string
  - Withdraw pusher to deploy stent
  - Advance guidewire into renal pelvis
  - Load 8 Fr pigtail catheter onto stiffening cannula
  - Advance over guidewire into renal pelvis and deploy as safety PCN catheter
    - Confirm position/patency with contrast injection
- **Pregnancy**
  - Obstructive stone disease not uncommon
  - Conservative management successful in 80%
    - IV hydration
    - Pain management
    - Bed rest
  - If conservative management fails, ureteroscopic removal unless
    - Sepsis
    - Large stone burden, (> 1 cm)
    - Lack of available expertise/equipment
  - Place patient prone-oblique, ipsilateral side up
  - Radiation-sparing maneuvers
    - 1-puncture technique; US for all wire manipulations
    - If fluoroscopy required
      - Place lead shield below pelvis
      - Pulsed fluoroscopy at lowest possible pulse rate that provides sufficient image quality
      - Maximize source-to-image receptor distance
      - Minimize patient-to-image receptor distance
      - Minimize fluoroscopy time/magnification
- **Whitaker test**
  - Several techniques described
    - 1-puncture technique is safest
      - Lower risk of hemorrhage
    - 2-puncture technique requires placement of 2 needles into renal collecting system
      - 1 needle for contrast injection
      - 1 needle for pressure measurement
  - Place patient in prone position
  - Perform primary PCN with needle/use existing PCN
    - If accessing renal collecting system
      - 18- to 23-g needle; prefer access into large calyx
  - Attach needle/PCN to 3-way stopcock
    - Attach 3-way stopcock to both a pressure transducer and power injector/pump
    - Place transducer at approximate level of kidney
  - Place Foley catheter in urinary bladder
  - Attach Foley catheter tubing to 3-way stopcock
    - Attach 3-way stopcock to pressure transducer & to tubing for urinary drainage bag
    - Place transducer at approximate level of bladder
  - Transducer reading measured in cm of H_2O as baseline opening pressure
  - Open renal 3-way stopcock to injector/pump
  - Open urinary bladder 3-way stopcock to transducer
  - Begin power injection/pump of 1:1 contrast diluted with sterile saline into needle/PCN
    - Adult flow rate: 10 mL/min
    - Small children: 5 mL/min
  - Use intermittent fluoroscopy to assess
    - Distention of renal pelvis and ureter
    - Possible point of obstruction
  - Intermittently measure pressures via both transducers during saline infusion
    - Turn off infusion
    - Open renal 3-way stopcock to transducer
  - Stop injection once complete distention achieved
  - Open renal 3-way stopcock to transducer
    - Measure renal pelvis & bladder pressures
  - Gradient pressure = (renal pelvis pressure - baseline) - (bladder pressure - baseline)
    - Gradient pressure < 15 cm H_2O: Normal
    - Gradient pressure > 22 cm H_2O: Obstruction
    - Gradient pressure of 15-22 cm H_2O: Indeterminate

        - Increasing infusion rate to 20 mL/min may elicit abnormal gradient in such cases
    - In obstructed system, may convert access to PCN
- **Suprapubic cystostomy**
    - Distend urinary bladder
        - Administer saline via transurethral catheter
        - If transurethral catheter not possible
            - Administer 1% lidocaine as local anesthetic
            - Advance 19-gauge needle into bladder with US guidance
            - May need CT guidance if interposed bowel
            - Watch for efflux of urine from needle
            - Fluoroscopically confirm position with contrast
            - Distend with saline or dilute contrast
            - 150-500 mL volume usually sufficient
    - **Trocar technique**
        - Use ultrasound guidance
            - Midline or slight paramedian approach
        - Administer local anesthetic 2-4 cm above pubic symphysis
        - 2-3 mm skin incision with #11 blade scalpel
        - Perform blunt dissection with hemostat
        - Insert inner stiffener into Foley catheter
        - Load Foley-cannula assembly onto trocar
        - Advance assembly into bladder under US guidance
            - Bladder wall may tent considerably
        - Remove trocar from Foley-cannula assembly
        - Disengage inner stiffener; advance Foley catheter into urinary bladder
        - Inflate Foley balloon with sterile water
        - Confirm position with US
            - Use contrast injection if fluoroscopy available
    - **Seldinger technique**
        - Advance Longdwell needle into distended bladder under US
            - Watch for efflux of urine from needle
            - May inject contrast to confirm position
        - Advance 0.035 inch Amplatz wire into bladder
        - Dilate tract as appropriate
        - Advance 8-14 Fr locking pigtail catheter over wire into bladder
        - Inject contrast to confirm position
        - May change/upsize to 16 Fr Foley after 6 weeks

Findings and Reporting

- Describe findings of antegrade nephrostogram
    - Filling defects (e.g., renal calculi)
    - Ureteral stricture
    - Variant anatomy (e.g., duplicated system)
- Describe consistency of urine
    - Clear, purulent

Alternative Procedures/Therapies

- Other
    - Retrograde ureteroscopic ureteral stent placement
    - Renal calculus removal
        - Extracorporeal shock wave lithotripsy (ESWL)
        - Ureteroscopic removal

POST PROCEDURE

Expected Outcome

- Technical success rate in hydronephrosis: 95-98%
    - 80-85% in nondilated systems
- Transient hematuria common after placement
    - Often resolves within 24-48 hours

Maintenance Considerations

- Flush catheter with 10 mL normal saline q 8 hours
    - 5 mL toward kidney and 5 mL toward drainage bag
- Regular catheter exchange every 4-12 weeks
    - More frequent if high urinary sediment/clogging
    - May cease regular flushing if no hematuria/clogging
    - Prophylactic antibiotics prior to every change
        - Tailor to prior cultures
        - Minimize contrast injection volume/guidewire manipulations
            - Reduces risk of sepsis
- Nephrolithotomy
    - Common management for calculi that are large, staghorn, & associated with anatomic anomalies (i.e., horseshoe kidney)
    - PCN may be performed in operating room immediately before nephrolithotomy
        - Requires intraoperative C-arm use
    - After nephrolithotomy, urologist may leave safety nephrostomy catheter in place
        - PCN injection next day
        - If patent ureter, usually may remove PCN
- Ureteral stent
    - Consider clamping safety PCN next day
    - Clamp trial may extend 1-7 days
        - Unclamp PCN if flank pain or fever develops
    - Contrast injection at end of trial
        - If patent stent, may remove PCN
    - Regular retrograde stent changes by urology

Issues to Consider

- Beware access with increased risk of complication
    - Supracostal access
        - Pneumo-/hydro-/hemothorax/empyema
    - PCN access through infundibulum or renal pelvis
        - Vascular injury, hematuria

Post Procedure Imaging

- Antegrade nephrostogram
    - May not fully evaluate ureter at initial PCN placement due to sepsis/obstruction
        - Risk of aggravating sepsis from overdistention
        - Perform when clinical condition improves
- Follow-up of calculus disease may include
    - Abdominal and pelvic radiography
    - US
    - NECT
- No current consensus guidelines for follow-up imaging for residual urolithiasis
    - Follow-up imaging typically pursued when complete stone clearance required, such as
        - Infectious calculi
        - Recurrent calculi

- NECT is modality with highest sensitivity & specificity for residual of urolithiasis
- Dual-energy CT (DECT) has evolved to allow characterization of chemical composition of calculi
- DECT can also be optimized to differentiate residual calculi fragments from adjacent ureteral catheter
  - Ureteral stents are homogeneous at DECT & readily differentiated from calculi
  - However, pitfalls exist with some catheter types
    - Nephrostomy tubes have internal bands & tips made of materials different from the remainder of the catheter
    - These portions can potentially be mischaracterized as calculi fragments
    - In problematic cases, in vitro scanning of the catheter can be performed
  - Sensitivity & specificity of DECT are decreased for small calculi (< 3 mm) & in patients with large body habitus

OUTCOMES

Problems

- Catheter dislodgement
  - May rescue within 1-2 days if well-established tract
    - Otherwise, may require new PCN access
- Frequent clogging of PCN
  - Increase frequency of catheter flushing
  - Increase frequency of catheter exchanges
  - Upsize catheter

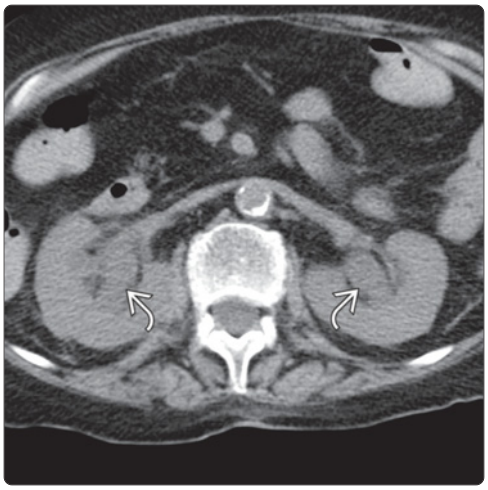
Complications

- Most feared complication(s)
  - Iatrogenic vascular injury and gross hematuria
    - Various vascular injuries possible
      - Pseudoaneurysm formation
      - Arteriovenous (AV) fistula
      - Arterial laceration
      - Hemorrhage & perinephric hematoma
    - May tamponade with catheter in place
      - Venous injuries usually self-limiting
      - Small AV fistula may be self-limiting
    - Angiography and embolization if persistent
  - Visceral transgression (rare)
    - Retrorenal colon
    - Spleen
    - Liver
    - Pleura
      - Pneumo-/hydro-/hemothorax/empyema
    - Bowel injury with suprapubic cystostomy
- Immediate/periprocedural complication(s)
  - Transient hematuria
  - Sepsis
    - Higher risk in pyonephrosis
- Delayed complication(s)
  - Pseudoaneurysm with gross hematuria
  - Urinoma
- Other complications
  - Bladder irritation/pain from nephroureteral catheter or ureteral stent
    - May consider cutting nephroureteral catheter
      - Distal tip terminates in ureter
    - May require conversion to PCN
  - Ureteral stent occlusion
    - Treat with stent exchange
  - Encrusted ureteral stent
    - Unable to remove cystoscopically
    - May require primary PCN
  - Migrated or fractured stent
    - Ureteroscopic removal
    - Percutaneous snare retrieval
  - Retained calculus fragment
    - Even small fragments (< 5 mm) can cause symptoms
      - Nidus for infection
      - Dislodgement & obstruction
      - Increased size of calculus leading to obstruction

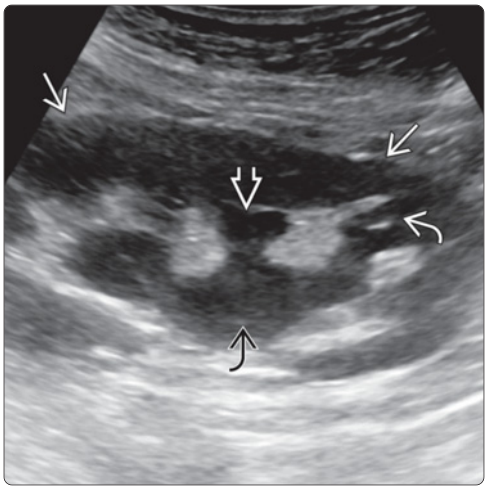
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Percutaneous Nephrostomy (Initial CT Imaging)

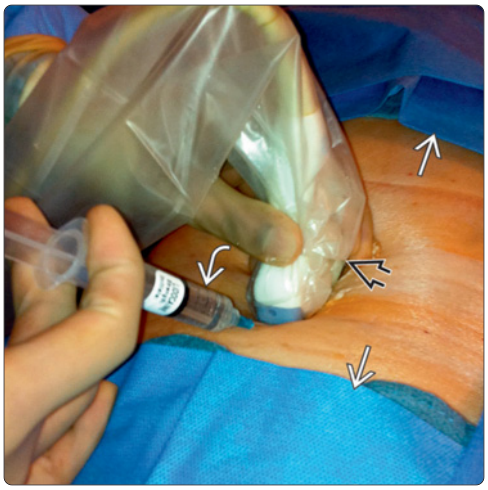


Percutaneous Nephrostomy (Preliminary Ultrasound Planning)

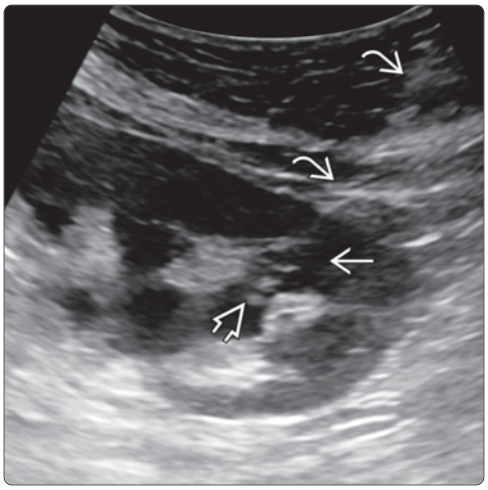


(Left) *Axial NECT obtained in a 68-year-old man who presented with fever & leukocytosis following radical cystectomy & ileal conduit formation shows moderate bilateral hydronephrosis, which was due to recurrent tumor at the ureteral-conduit anastomosis.* **(Right)** *Sagittal US of the left kidney demonstrates moderate dilatation of the calyces & the renal pelvis. A posterior calyx of the inferior pole has been selected for percutaneous nephrostomy access.*

Percutaneous Nephrostomy (Preparation of Access Site)



Percutaneous Nephrostomy (US-Guided Needle Placement)

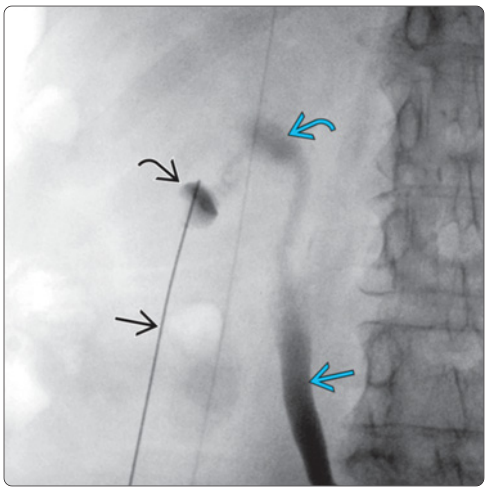


(Left) *After the skin has been sterilely prepared & draped, 1% lidocaine is administered as a local anesthetic. Using a US probe that has been wrapped within a sterile probe cover, sonography of the kidney is performed in the sagittal plane.* **(Right)** *Sagittal US of the kidney shows that a 22-gauge Chiba needle has been advanced into a posterior calyx of the inferior pole using continuous, real-time US guidance. The tip of the needle is well visualized within the hypoechoic collecting system.*

Percutaneous Nephrostomy (US-Guided Needle Placement)

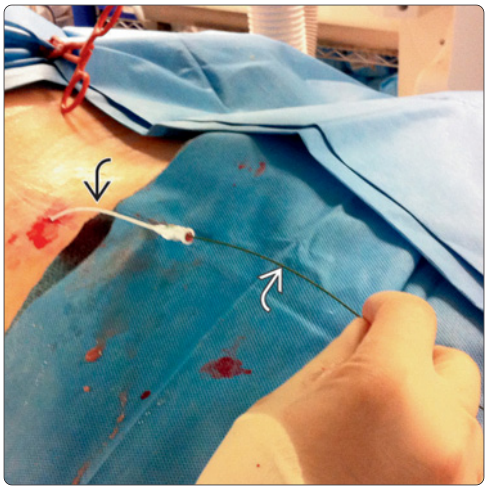


Percutaneous Nephrostomy (Antegrade Nephrostogram)

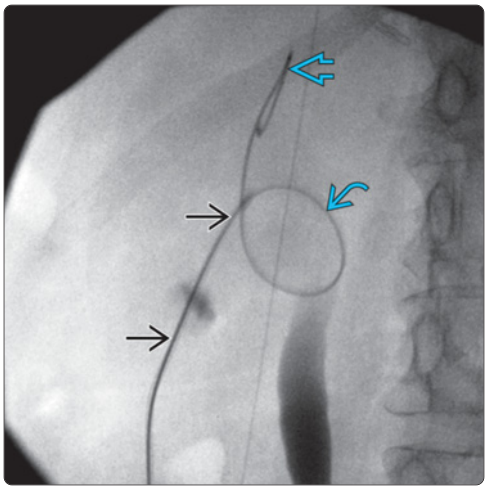


(Left) *After advancing the Chiba needle into the targeted calyx, the inner stylet is removed & efflux of urine is noted, thereby confirming access into the collecting system.* **(Right)** *Fluoroscopic AP image shows that contrast injected via the Chiba needle confirms access in the inferior pole calyx & propagates into the renal pelvis & proximal ureter. A 1-puncture technique or 2-puncture technique (using a Ring needle) may be used to access a calyx under fluoroscopic guidance.*

Percutaneous Nephrostomy (Transition to Larger Guidewire)

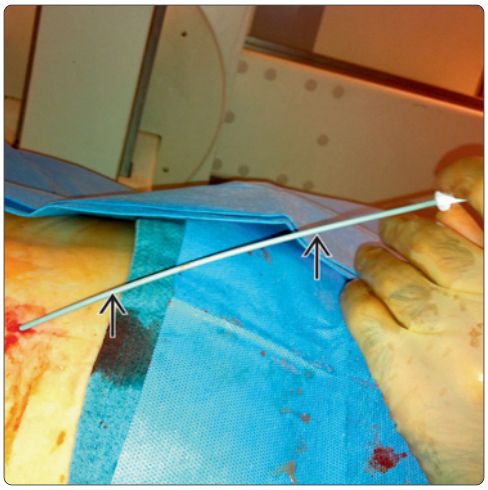


Percutaneous Nephrostomy (Guidewire Placement in Renal Pelvis)

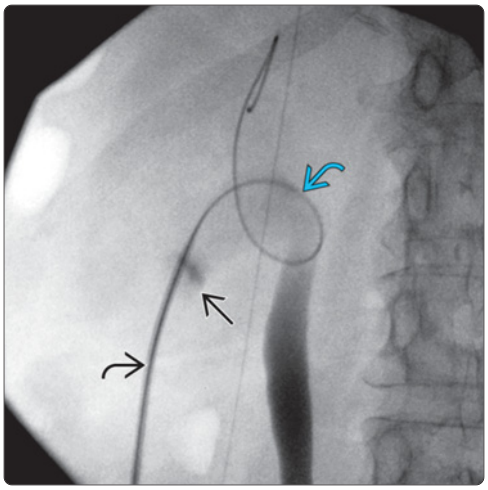


(Left) *Photograph shows that a 0.035" 3-J guidewire has been introduced into the sheath & is being advanced into the renal collecting system. The larger guidewire is needed to provide more stability for nephrostomy catheter placement.* **(Right)** *Fluoroscopic radiograph shows that the 0.035 inch 3-J guidewire has been advanced through the 6 Fr sheath using fluoroscopic guidance & has been allowed to form a loop within the renal pelvis. The tip of the guidewire is in a superior pole calyx.*

Percutaneous Nephrostomy (Dilatation of Tract)

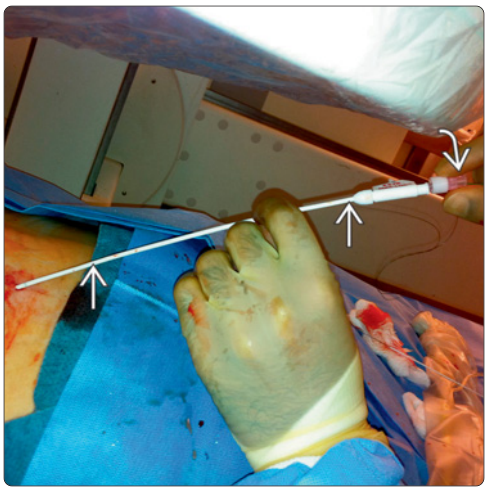


Percutaneous Nephrostomy (Dilatation of Tract)

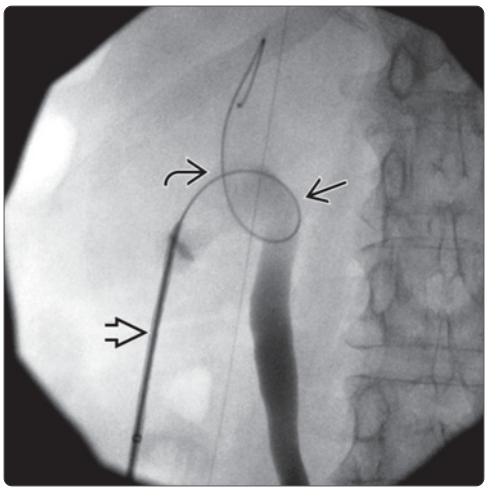


(Left) *The access sheath is exchanged for an 8 Fr dilator to expand the soft tissue tract. The fascial dilator is advanced over the 3-J guidewire into the renal collecting system. Care is taken not to kink the guidewire during tract dilation.* **(Right)** *Fluoroscopic AP image obtained during tract dilatation shows that the 8 Fr dilator has been advanced over the 3-J guidewire, so that the dilator tip is within the lower pole calyx. The guidewire position should remain stable during all exchanges.*

Percutaneous Nephrostomy (Nephrostomy Catheter Placement)

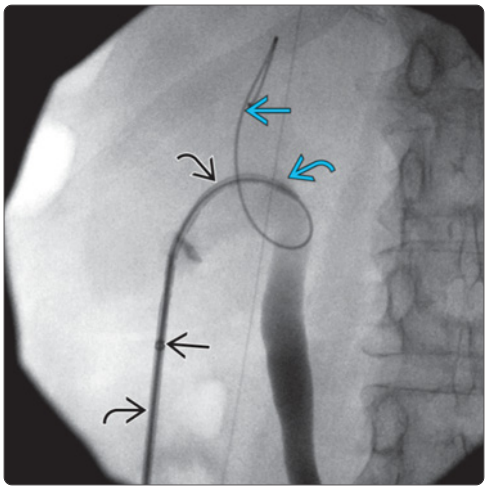


Percutaneous Nephrostomy (Nephrostomy Catheter Placement)



(Left) *The dilator is removed, & an 8 Fr locking nephrostomy catheter, which is mounted on an inner stiffener, is advanced. Once within the renal collecting system, the inner stiffener is unscrewed & held still while the catheter is advanced over the wire further into the collecting system.* **(Right)** *Fluoroscopic AP image shows that the 8 Fr nephrostomy catheter has been advanced into the collecting system. Once a turn in the guidewire is reached, the inner stiffener is unscrewed.*

Percutaneous Nephrostomy (Nephrostomy Catheter Placement)

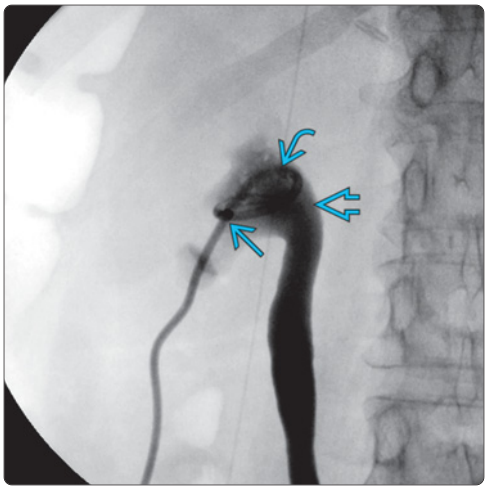


Percutaneous Nephrostomy (Nephrostomy Catheter Placement)



(Left) *After the inner stiffener is unscrewed, it is held in a stationary position, & the catheter is advanced over the guidewire further into the collecting system. A radiopaque marker demarcates the last sidehole of the nephrostomy catheter, which should reside within the collecting system.* **(Right)** *After the nephrostomy catheter has been satisfactorily positioned under fluoroscopy, the guidewire is removed & the string is pulled to form the distal pigtail of the catheter.*

Percutaneous Nephrostomy (Nephrostomy Catheter Placement)

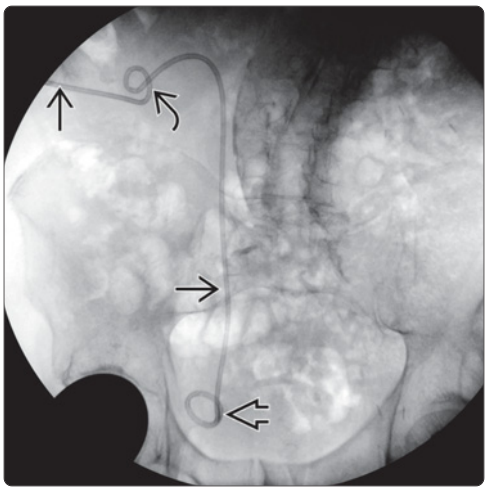


Percutaneous Nephrostomy (Aspiration of Urine)

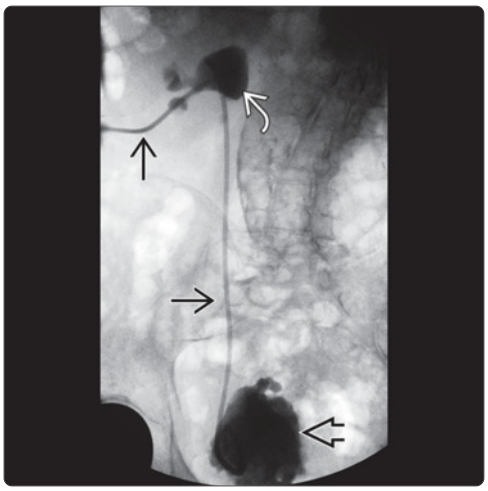


(Left) *Fluoroscopic image demonstrates the final position of the nephrostomy catheter. The pigtail portion of the catheter & the radiopaque marker lie within the renal pelvis. Injection of contrast confirms positioning in the collecting system.* **(Right)** *Photograph shows that a syringe has been attached to the nephrostomy catheter, allowing aspiration & decompression of the collecting system. In this example, purulent urine was present, reflecting clinically suspected urosepsis.*

Nephroureteral Catheter (Appearance After Placement)



Nephroureteral Catheter (Contrast Injection Evaluation)

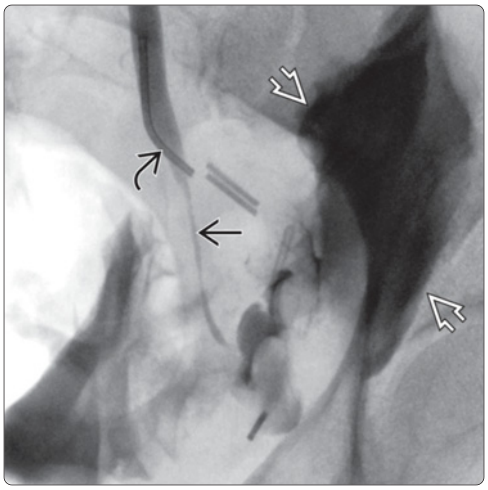


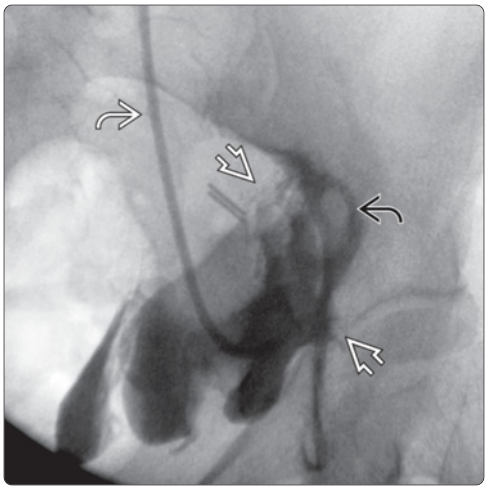
(Left) *Fluoroscopic AP image prior to catheter exchange shows the typical course of a nephroureteral catheter. The proximal pigtail is in the upper quadrant (renal pelvis), & the distal pigtail is in the pelvis (urinary bladder). Nephroureteral catheters are useful in patients prone to catheter dislodgement or in cases of ureteral stenosis or injury.* **(Right)** *Contrast injection demonstrates catheter patency. There is opacification of the renal pelvis & the urinary bladder.*

Evaluation of Cause of Obstruction (Distal Ureteral Stricture)

Traversing Cause of Obstruction (Distal Ureteral Stricture)

(Left) *Fluoroscopic AP image shows contrast injection via a 5 Fr Kumpe catheter that has been advanced into the distal ureter. The injected contrast reveals a distal ureteral stricture that results in ureteral obstruction in this patient who has undergone radical cystectomy & ileal conduit formation.* **(Right)** *The distal ureteral stricture was crossed with a 0.035 inch angled hydrophilic guidewire followed by an 8 Fr nephroureteral catheter with the distal pigtail placed into the conduit.*

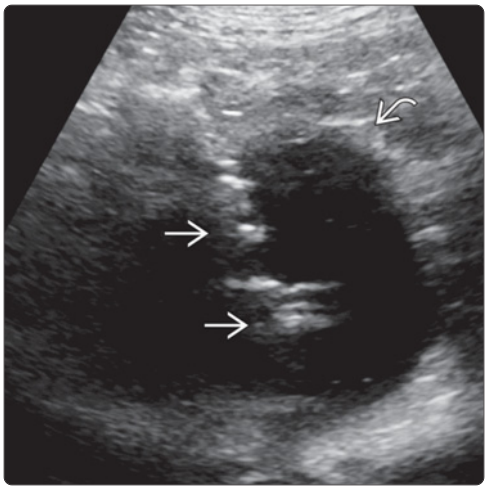


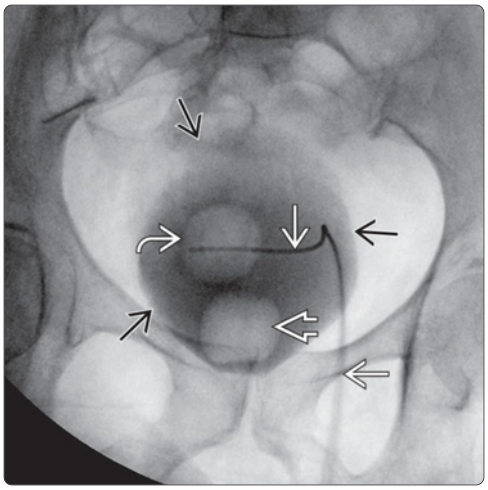


Suprapubic Cystostomy (Trocar Technique)

Suprapubic Cystostomy (Trocar Technique)

(Left) *Transverse US reveals a urinary bladder that has been distended by saline administered via a transurethral catheter. A trocar mounted with a 14 Fr Foley catheter is advanced into the bladder. A strong thrust is used to puncture, rather than "tent," the bladder wall.* **(Right)** *Fluoroscopic AP image shows contrast injected through the suprapubic cystostomy catheter, confirming location of the catheters in the bladder. The Foley balloons of the suprapubic & transurethral catheters are seen.*

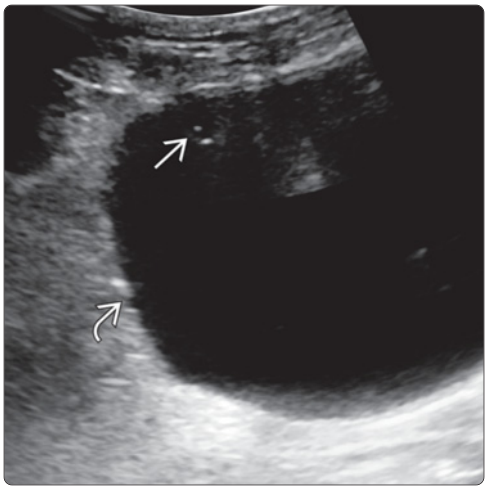


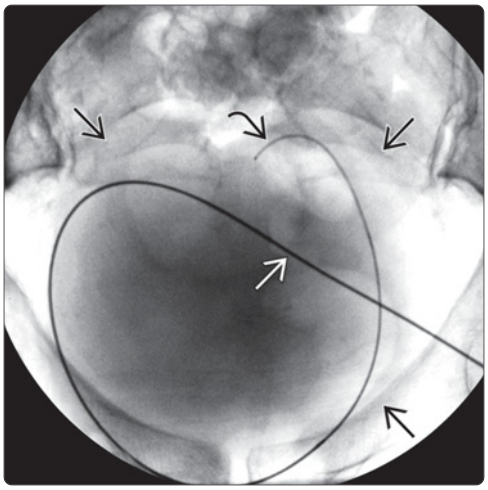


Suprapubic Cystostomy (Seldinger Technique)

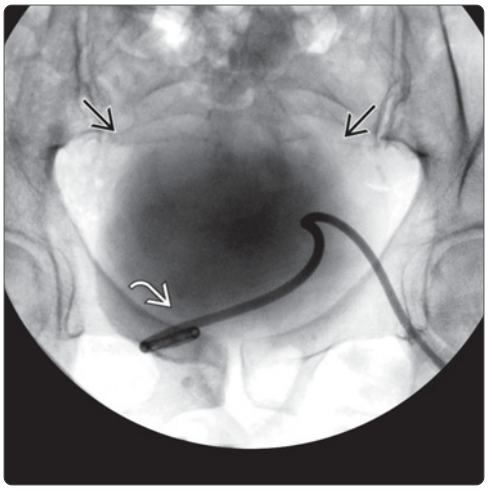
Suprapubic Cystostomy (Seldinger Technique)

(Left) *Transverse US shows that a 19-g ultrathin Chiba needle has been advanced into a distended bladder. A Longdwell needle could also have been used to access the bladder.* **(Right)** *Fluoroscopic image shows that after accessing the urinary bladder contrast is injected via the needle to confirm positioning. A 0.035" Amplatz guidewire is then advanced via the Chiba needle & allowed to coil in the bladder. The needle is removed & the tract is dilated, followed by suprapubic catheter placement.*

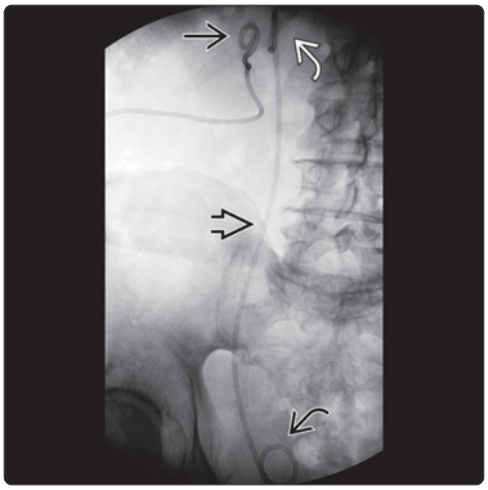




Suprapubic Cystostomy (Seldinger Technique)

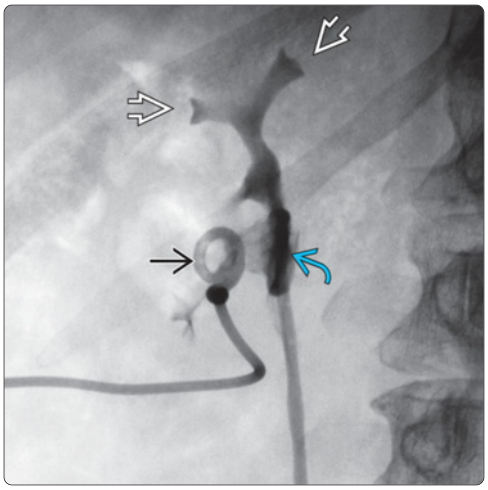


Antegrade Ureteral Stent (Appearance After Placement)

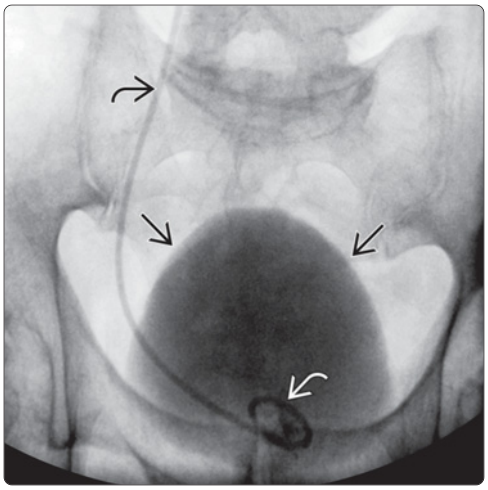


(Left) *Fluoroscopic radiograph shows that, after advancing the suprapubic cystostomy catheter over the guidewire, contrast is injected through the catheter. This shows the 12 Fr locking pigtail catheter and confirms a satisfactory location of the catheter in the urinary bladder.* **(Right)** *Fluoroscopic image obtained prior to contrast injection of a percutaneous nephrostomy (PCN) tube demonstrates a ureteral stent that was placed using an antegrade approach. The proximal and distal pigtails are visualized.*

Antegrade Ureteral Stent (Injection of Safety PCN)

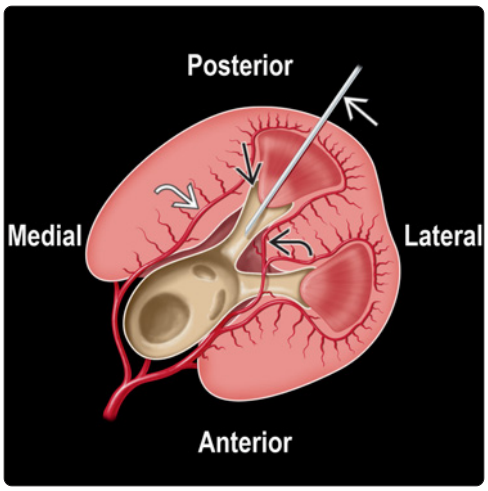


Antegrade Ureteral Stent (Injection of Safety PCN)

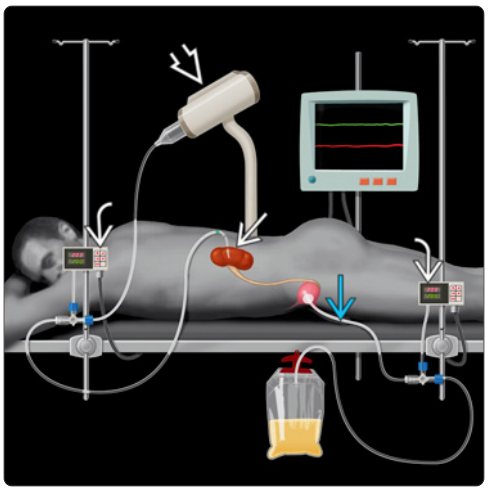


(Left) *Fluoroscopic radiograph shows injection of contrast via the nephrostomy catheter to assess the ureteral stent prior to PCN removal. The renal pelvis & calyces are opacified, & the proximal pigtail of the ureteral stent is seen.* **(Right)** *Fluoroscopic image of the pelvis following contrast injection of the safety nephrostomy catheter shows an opacified bladder containing the distal pigtail of the ureteral stent. As this confirmed stent patency, the safety PCN was removed.*

Preferred Needle Course (Avascular Plane of Brödel)



Whitaker Test Schematic (Evaluation of Ureteral Obstruction)



(Left) *Graphic shows that a needle has been placed into a posterior calyx along the relatively avascular plane of Brödel, located between the anterior & posterior divisions of the renal artery; this plane lies 20-30° from the sagittal plane. This approach minimizes potential vascular injury.* **(Right)** *Whitaker test schematic shows that a needle in the renal pelvis is infused with saline via an injector. Pressures are measured there & in the bladder via a Foley catheter, using transducers, to evaluate for ureteral stasis vs. obstruction.*

Kidney Ablation/Embolization

KEY FACTS

PREPROCEDURE

- **Indications for ablation**
  - Small (< 3 cm) exophytic, posterior renal mass ideal
  - Traditional indications: Elderly patient, comorbidities
  - Evolving management paradigm: Favorable safety profile, lower long-term costs, recurrence rates comparable to open/laparoscopic nephron-sparing surgery
  - Approach also changing with recognition of biology of incidental "small" (< 3 cm) renal mass
  - > 20% small (≤ 3) renal masses are benign
  - Very low chance of metastases of renal cell carcinoma (RCC) < 3 cm
  - Biology prompts less aggressive approach
    - Active surveillance
    - Percutaneous ablation, particularly if growth
- **Indications for embolization**
  - Angiomyolipoma (AML) > 4 cm; nonruptured aneurysms > 5 mm
- Review prior imaging
  - Thermal ablation injury risk if tumor near collecting system; consider pyeloperfusion
  - Determine vascularity of AML before treating

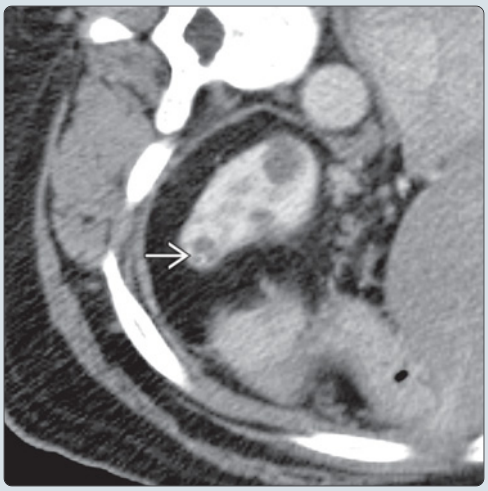
PROCEDURE

- **Ablation**: CT guidance used for most renal ablations
  - Goal: Ablation zone completely encompasses tumor plus 0.5-1 cm margin
- **Embolization**: Perform as selectively as possible
  - Devascularize AML; preserve normal renal tissue

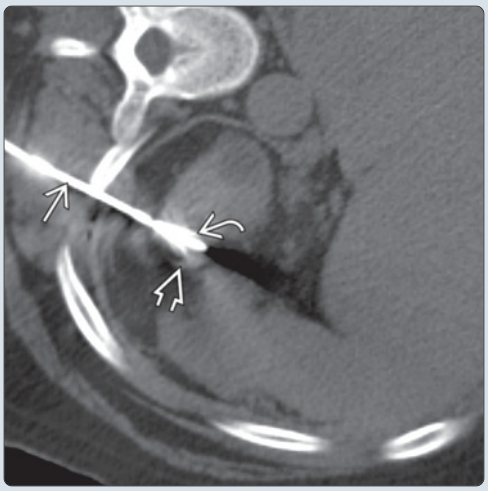
POST PROCEDURE

- **Ablation**: ≥ 90% complete necrosis in mass ≤ 4 cm
  - CT/MR nonenhancement correlates with necrosis
- **AML embolization**: Success rate of 90-100%
  - Tumor does not disappear; decreases by 40-66%

Radiofrequency Ablation of RCC: Small Target Lesion Identified at CECT

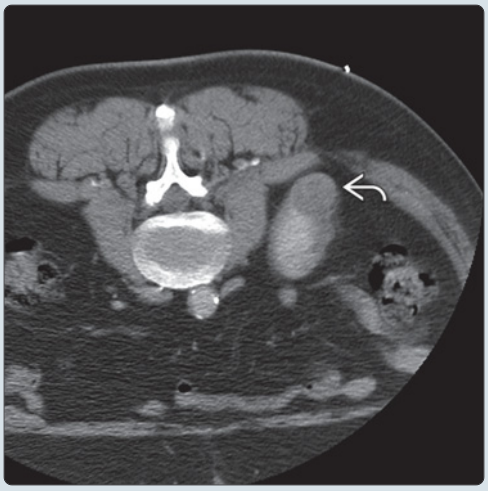


Radiofrequency Ablation of RCC: Targeting via NECT

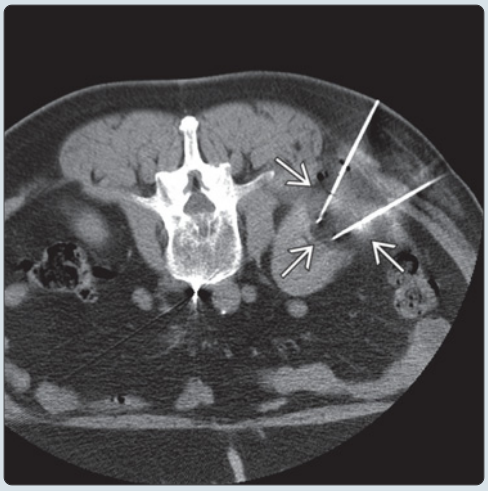


(Left) *Axial CECT shows a 1.4 cm solid renal mass in a patient who is not a surgical candidate. Renal masses < 4 cm in size are particularly amenable to percutaneous thermal ablation, with a clinical success rate > 90%.* **(Right)** *A multitined radiofrequency ablation (RFA) electrode, also known as an RFA probe, has been advanced into the lesion using CT guidance. The tines are deployed and the probe position is confirmed before ablation.*

Cryoablation of RCC: Targeting via CECT



Cryoablation of RCC: Ice Ball Monitor



(Left) *CECT used to target a biopsy confirmed papillary RCC imperceptible at NECT and ultrasound. Cryoablation was chosen given the favorable location and growth of the mass and prior retroperitoneal surgery.* **(Right)** *Three cryoablation probes were positioned. The low attenuation "ice ball" roughly corresponds to a lethal ablation zone, but multiple overlapping probes are necessary for successful ablation. Analgesia requirements for cryoablation are typically less than that of RFA.*

TERMINOLOGY

Definitions

- **Ablation**: Imaging directed induction of thermal/nonthermal cellular death
  - **Energy-based ablation**
    - Radiofrequency ablation (RFA): Heat based
      - Most common modality used for renal ablation
    - Cryoablation: Freezing/thawing cycle
      - Also frequently used modality for renal ablation
    - Microwave ablation: Heat-based cytotoxicity
      - Offers faster ablation times; less susceptibility to thermal heat-sink effect
    - High-intensity focused ultrasound (HIFU): Coagulation necrosis caused by high-intensity ultrasound beam focused on small tissue volume
      - Respiratory motion/overlying ribs problematic
    - Irreversible electroporation (IRE): Electrical disruption of integrity of cell membrane
      - Promising preliminary animal/human data
  - **Chemical ablation**
    - Inconsistent cellular necrosis in renal tumors
      - Not routinely used for renal ablation
- **Embolization**:Transcatheter introduction of intravascular agent for devascularization/occlusion
  - Bland embolization: Transcatheter arterial embolization without chemotherapeutic agent
  - Transcatheter ethanol injection: Denatures cellular proteins/causes small vessel thrombosis
- **Thermal heat-sink effect**: Thermal energy needed to achieve cytotoxicity is dissipated
  - Convective heat transfer to adjacent structures
    - Blood flow in vessels abutting tumor prevents adequate heat generation for effective ablation

PREPROCEDURE

Indications

- **Renal cell carcinoma (RCC)**
  - Traditional impetus for percutaneous ablation
    - Nonsurgical candidate: Elderly patient, comorbidities
    - von Hippel-Lindau syndrome, multiple renal lesions
    - Solitary kidney
    - Renal insufficiency
  - Evolving management paradigm: Favorable safety profile, diminished long-term costs, recurrence rates comparable to open/laparoscopic nephron sparing surgery
  - Approach also changing with recognition of biology of incidental "small" (< 3 cm) renal mass
    - > 20% small (3 cm or less) renal masses are benign
    - Very low chance of metastases if < 3 cm
    - Biology prompts less aggressive approach
      - Active surveillance
      - Percutaneous ablation, particularly if growth
  - Anatomy also drives treatment algorithm
    - Best percutaneous ablation results with masses < 4 cm
    - Exophytic, posterior lesions more easily targeted
- **Angiomyolipoma (AML)**
  - **Elective/prophylactic treatment**
    - Tumor > 4 cm
      - Size is crude measure of rupture potential (large/predominantly fatty tumor unlikely to bleed)
      - 1 bleeding episode/3.4 pt-y if AML > 4 cm
    - Nonruptured aneurysms > 5 mm
      - More predictive of rupture than tumor size
    - Symptomatic patient
      - Pain/tenderness/mass effect/hemorrhage
  - **Emergent treatment**
    - Acute rupture/hemorrhage
      - Ruptured AML aneurysm may present in shock
      - Rupture spontaneously or after minimal trauma
      - Successful intervention possible even if unstable

Contraindications

- **General**
  - Uncorrectable coagulopathy
  - Lack of safe access to lesion
- **Ablation**
  - Lesion with high risk of incomplete ablation
    - Large size (≥ 4 cm)
    - Proximity to central vascular structures
      - Produces heat-sink effect
  - Lesion near structures at risk of thermal injury
    - Renal hilar structures
      - e.g., vessels, renal collecting system
- **Embolization**

Preprocedure Imaging

- **CT/MR**
  - Determine lesion location
    - Least challenging location: Posterior exophytic
    - Most challenging location: Central hilar
  - Ascertain lesion size and margins
    - Ideal lesion ≤ 3 cm but good results if < 4 cm
    - Risk of incomplete ablation rises with lesion size
  - 3D margins/tumor volume should be determined
    - Multiplanar reformatted images helpful
  - Evaluate lesion proximity to collecting system
    - Pyeloperfusion of cool D5W through externalized ureteral stent may protect ureter
  - Evaluate lesion proximity to vascular structures
    - Consider additional treatment in areas at risk
  - Plan site/path of probe insertion
- **Computed tomography angiography (CTA)**
  - Very useful prior to transcatheter embolization
    - May show dominant vascular supply to tumor
    - May demonstrate arteriovenous shunting
      - Implications for safety of embolization
- **Ultrasound**
  - Must document satisfactory visualization of lesion with sonography if US guidance considered
  - Combined approach (US for probe placement, CT for ablation monitoring) may be employed
- **Digital subtraction angiography (DSA)**
  - Evaluation of tumoral hemodynamics
    - Potential impact on choice of embolic agent/safety profile of transcatheter embolization
  - Imaging guidance during transcatheter embolization

Getting Started

- Things to check
  - Procedure indications
  - Current medications
    - Any anticoagulant or antiplatelet agents
  - Allergies
  - Pacemaker/defibrillator
    - Pre-RFA electrophysiologist evaluation
  - Laboratory parameters
    - Complete blood count (CBC)
      - Platelet > 50,000/μL
    - Coagulation profile
      - International normalized ratio (INR) ≤ 1.5
      - Normal prothrombin time (PT), partial thromboplastin time (PTT)
    - Estimated glomerular filtration rate (eGFR)
      - Reduced eGFR may impact prophylactic antibiotic regimen/ability to give IV contrast
  - Biopsy
    - Greater than 20% of renal masses < 3 cm benign
- Medications
  - Anesthesia
    - 1% lidocaine local anesthetic
    - Adequate procedural sedation
      - Fentanyl, midazolam, Demerol typically used
      - Analgesia requirements of cryoablation may be less than RFA
    - Anesthesiology assistance if necessary
  - Prophylactic antibiotics
    - Administer if pyeloperfusion required
    - Regimens vary
      - Ampicillin (1 gm), gentamicin (80 mg)
      - Levofloxacin (500 mg)
- Equipment list
  - **General**
    - CT/MR/DSA equipment for guidance
      - Most renal ablations performed with CT
      - MR guidance possible but availability limited
    - US equipment for guidance; US may aid in initial probe placement followed by CT
      - 3-5 MHz transducer
      - Sterile US probe cover/gel
  - **Hydrodissection**
    - D5W used as hydrodissection agent
    - Longdwell sheathed needle (e.g., Ring)
  - **Radiofrequency ablation**
    - Radiofrequency generator/probe
    - Grounding pads (if monopolar probe)
    - If internally cooled probe
      - Bucket containing ice water
  - **Cryoablation**
    - Specialized gases (e.g., argon, helium)
    - Cryoprobes
  - **Microwave ablation**
    - Microwave generator
    - Microwave antenna and cable
  - **Embolization**
    - Appropriate catheters/guidewires/embolic agents

PROCEDURE

Patient Position/Location

- Best procedure approach
  - **Ablation**: Most comfortable position that also facilitates procedural access
    - Ipsilateral decubitus
      - Optimal for upper pole masses
      - Lung displaced, reducing risk of pneumothorax
      - Ipsilateral kidney moves less
    - Prone
      - Suitable for lower pole masses
      - Subcostal approach; angle gantry if necessary
    - 30° oblique (ipsilateral side up)
      - Suitable for anterior masses
      - Allows movement of bowel from ablation zone
    - Supine
      - Suitable for anterior/lateral masses
      - Liver, spleen, colon should not be in trajectory
  - **Embolization**: Transfemoral arterial access

Equipment Preparation

- Hydrodissection
  - Consider nonionic contrast diluted in D5W
    - e.g., 5-10 mL Isovue-370 in 500 mL D5W
    - May improve hydrodissection margin delineation
- Pyeloperfusion
  - Requires urology to place externalized ureteral stent
  - Connect 1 L bag of cool D5W to ureteral stent
    - Start slow drip infusion (e.g., rate of 1-2 drops/second)

Procedure Steps

- **General**
  - Perform preliminary CT and localize target
    - Verify stent position for pyeloperfusion
    - Reconfirm planned access route is appropriate
  - Reassess any other ancillary requirements
    - e.g., hydrodissection/change in patient position
  - Sterilely prepare/drape skin
  - Administer 1% lidocaine local anesthetic
    - Dermatotomy with #11 scalpel at skin entry site
  - If needed, may perform biopsy prior to ablation
    - Consider biopsy in a separate prior session
      - Exclude possibility of benign mass
      - "Trial run" for ablation procedure
- **Hydrodissection**
  - Insert Longdwell needle into plane between planned ablation margin and structure at injury risk
  - Connect sheath to extension tubing/3-way stopcock
  - Instill 10 mL hydrodissectant
    - Should flow easily; if resistance encountered, needle tip may be intraparenchymal; adjust tip
  - Perform CT to evaluate effect of hydrodissection
- **Ablation**
  - **General**
    - Ablation zone should completely encompass tumor with additional 0.5-1 cm margin
    - If lesion imperceptible on noncontrast scan
      - Place needle/probe using anatomic landmarks
      - Perform intravenous contrast enhanced scan

    - Adjust probe placement accordingly
  - At end of ablation, obtain postprocedure imaging
    - Assess for hemorrhage/other complications
- **RFA/microwave ablation**
  - Place grounding pads on legs; both at same level
    - Risk of skin burn if not same level
  - Place RF probe(s)/microwave antenna(s) in lesion
    - Confirm position with repeat imaging
  - Activate generator; activate ablation system
    - Some patients may sense pain during ablation
  - At end of ablation cycle, reposition probe/antenna
    - Overlapping ablations may be needed to achieve desired ablation volume
    - Image after each applicator repositioning
- **Cryoablation**
  - Advance cryoprobes into lesion
  - Multiple cryoprobes usually necessary
    - Smaller ablation zone per probe than RFA
    - Position probes 1 cm from tumor margin
    - Perform scan after each probe placement to verify position; generally space 1.5 cm apart
  - Ablation zone visible on intraprocedural CT/MR
    - Allows more confident tumor margin coverage than RFA; no need for IV contrast to verify
    - Less risk of collecting system injury reported
  - Activate freeze cycle
    - Duration: 10-15 minutes
    - Scan every 2-5 minutes; assess ice ball size
    - Ice ball formation is hypodense on CT
  - Goal is ice ball margin at least 5 mm past desired ablation margin
    - Ice ball margin represents 0° C isotherm (insufficient for cell death)
    - -20° C temperature 3 mm deep to ice ball margin; lethal zone ~ 5 mm deep
  - Ice ball should be 2 cm larger in diameter than tumor to achieve adequate margin
    - Consider additional cryoprobes if small ice ball
  - Activate passive thaw cycle (8-10 minutes)
  - Repeat freeze cycle
    - At end of 2nd freeze cycle, active thaw
  - Remove cryoprobes gently with twisting motion
    - Aggressive removal risks parenchymal fracture
  - May note backbleeding requiring manual pressure
    - Cryoablation lacks coagulative effects of RFA
  - Perform postprocedure scan
- **Embolization**
  - Obtain arterial access
  - Introduce access sheath/selective catheter
  - Catheterize artery most likely supplying tumor
  - Obtain/analyze DSA images
    - Determine appropriate/optimal embolic agent
    - Significant arteriovenous shunting may influence choice/appropriateness of particles
  - Selectively catheterize dominant arterial branch feeding tumor; may use coaxial microcatheter
  - Obtain DSA; proceed with embolization
    - Embolize as selectively as possible
    - Embolization agent: Particles or alcohol
    - Endpoint: Stasis within target circulation

Alternative Procedures/Therapies

- Surgical
  - Open/laparoscopic thermal ablation
  - Open/laparoscopic partial/complete nephrectomy

POST PROCEDURE

Expected Outcome

- **Ablation**
  - Complete necrosis in masses ≤ 4 cm: ≥ 90%
  - Reported residual/recurrent tumor incidence
    - Single radiofrequency ablation: 13.4%
    - Single cryoablation: 3.9%
    - After repeat ablation (both modalities): 4.2%
  - Overall 2-year survival: 82.5%
- **Embolization**
  - Success rate of AML embolization: 90-100%
  - Tumor does not disappear; decreases by 40-66%
    - Angiomyogenic component decreases > fat
    - Fatty tissue relatively insensitive to embolization
  - Minimal loss of surrounding normal parenchyma

Things to Avoid

- Thermal injury to adjacent structures (ablation)
  - Renal collecting system/ureter
  - Bowel/solid organs (e.g., pancreas/spleen/liver)
- Nontarget embolization (transcatheter embolization)

Postprocedure Imaging (Ablation)

- Need serial follow-up imaging to assess success
- Imaging interval institution-dependent
  - 1 algorithm: 1, 3, 6, 9, 12 months; then annually
- Normal postablation protocol and findings
  - NECT/CECT (nephrographic/excretory phases)
    - Subtraction images helpful to assess enhancement; multiplanar reformations helpful to assess margins
    - Hypodense ablation zone; may be wedge-shaped
    - Hyperdense areas due to coagulated proteins
    - Ablation zone larger than index lesion
      - May see enlargement for up to 6 months
      - Later enlargement suspicious for progression
  - MR
    - Lesion variable in intensity on T1WI
    - Lesion hypointense on T2WI
- CT/MR nonenhancement correlates with tumor necrosis
- Residual disease or local progression manifested by
  - Nodular type: Peripheral nodules of enhancement
  - Peripheral enhancing crescent
  - Morphologic changes/growth on serial imaging
- Benign ancillary findings
  - Thin rim (< 5 mm) of periablational enhancement
    - MR may show T2 hyperintense rim
    - Most prominent at 1 month; may last 6 months
  - Perirenal or pararenal fat stranding
    - May coalesce over serial scans into dominant halo oriented parallel to surface of kidney
    - May displace axis of kidney
      - May complicate comparison across studies
  - Fatty replacement of zone of ablation

- Extracalyceal contrast on delayed images; not important if nonobstructed distal collecting system
- Liquefactive necrosis
  - Thermal ablation generally causes coagulative necrosis; rarely, may see liquefactive necrosis
    - Hyperintense on T2WI
    - Fat-fluid levels/internal debris
  - Distinguish from recurrent tumor by lack of enhancement
  - Consider biopsy in challenging cases

OUTCOMES

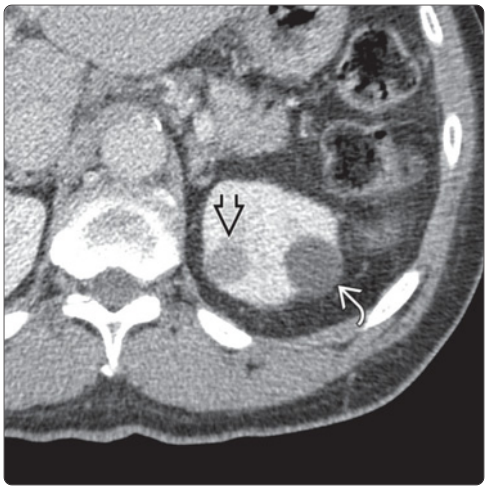
Complications

- Most feared complication(s)
  - Ureteral stricture
    - May require nephrostomy, ureteral stent
    - Urinoma may require separate drain
  - Bowel perforation
- Immediate/periprocedural complication(s)
  - **Ablation**
    - Hemorrhage
    - Pleural effusion/pneumothorax
    - Skin burn
      - Overlying treatment site, at grounding pad site
      - Shave skin/ensure good contact to reduce risk
    - Cutaneous nerve injury (paresthesias)
  - **Embolization**
    - Periprocedural aneurysm rupture in AML
    - Nontarget embolization/renal parenchymal loss
      - If superselective embolization not possible
    - Postembolization syndrome (AML embolization)
      - Inflammatory response to necrotic tissue
      - Affects 90% of AML embolization patients
      - Symptoms: Pain, nausea, vomiting, fever
      - Treat with IV fluids, analgesics, antipyretics, antiemetics, tapered dose of prednisone
    - Arterial access site complications
- Delayed complication(s)
  - Tract seeding (exceedingly rare)
  - Pseudoseeding
    - Inflammatory nodules along applicator tract; consider biopsy if fails to resolve on serial imaging
  - Postablation syndrome

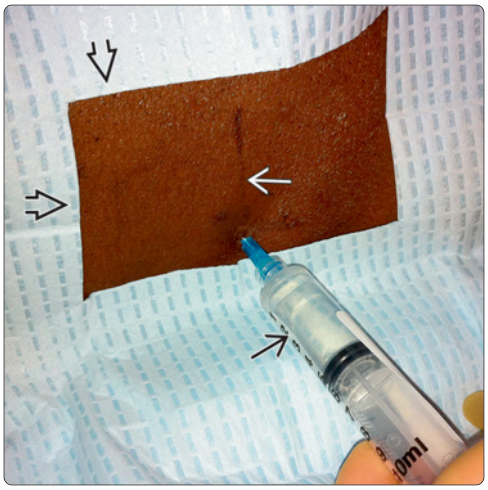
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Radiofrequency Ablation of RCC (Initial CECT Imaging Evaluation)

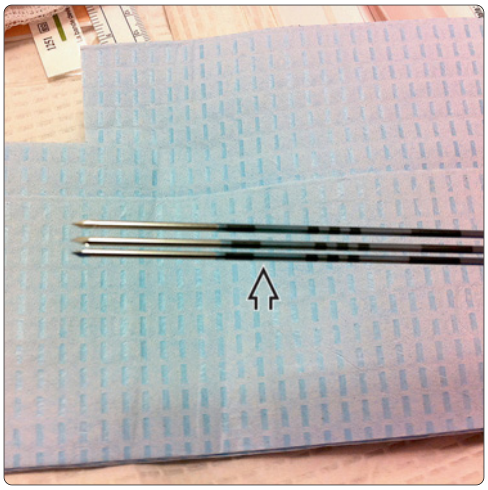


Radiofrequency Ablation of RCC (Access Site Preparation & Anesthesia)

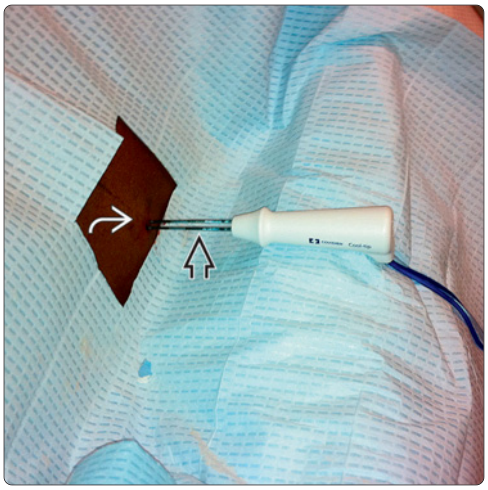


(Left) *Axial CECT demonstrates a hypodense renal mass ⇨ medially at the same level as a simple cyst ⇨. Ultrasound confirmed that the hypodense mass was solid. Review of prior imaging is important in planning access to the lesion. Imaging also shows that hydrodissection is unnecessary, as there are no adjacent structures at risk of nontarget thermal injury.* **(Right)** *The skin is marked ⇨ after a preliminary CT with a radiopaque grid. After sterile cleansing and draping ⇨ the skin, 1% lidocaine local anesthetic ⇨ is administered.*

Radiofrequency Ablation of RCC (Cluster Configuration RFA Electrode)

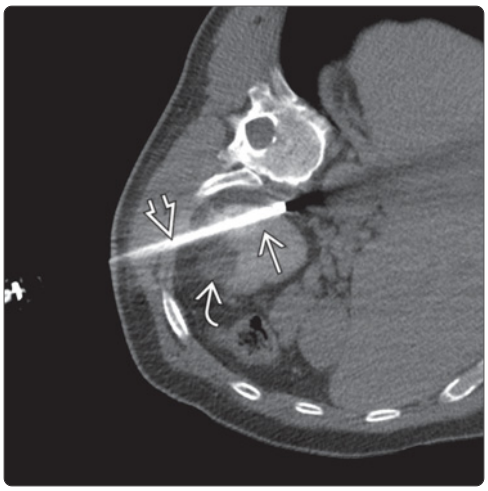


Radiofrequency Ablation of RCC (Cluster Electrode Placement)

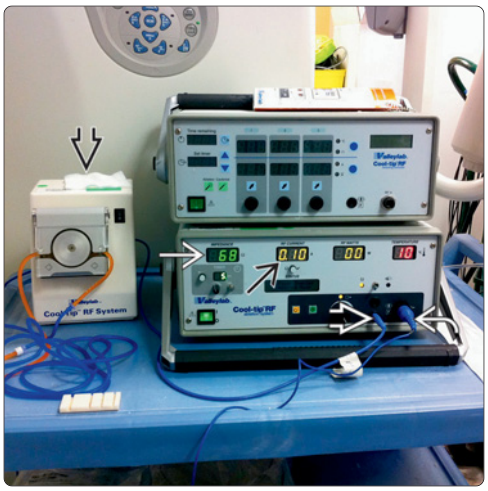


(Left) *Photograph shows a cluster RFA probe ⇨. The probe consists of 3 prongs attached to a single hub. The design allows a larger area of thermal ablation when compared to a single needle design. Internal cooling of the probes reduces the impedance of the surrounding tissue, enabling a larger ablation.* **(Right)** *The probe ⇨ is inserted through the skin ⇨ to a predetermined depth as measured by CT. The prongs are kept parallel but separate; prongs too close or too far apart may yield a smaller burn.*

Radiofrequency Ablation of RCC (Cluster Electrode Placement)



Radiofrequency Ablation of RCC (Radiofrequency Generator Operation)

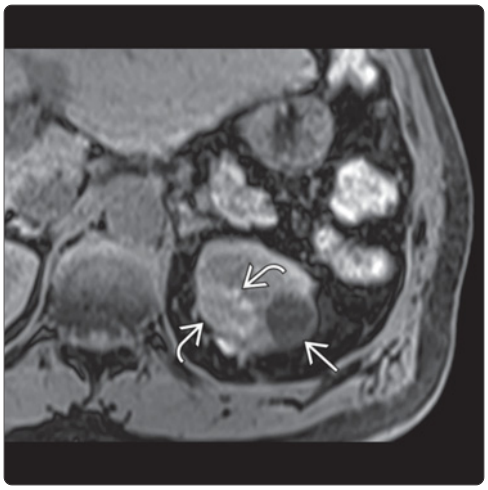


(Left) *Axial NECT confirms that there is satisfactory probe ⇨ placement through the lesion ⇨. Note the simple cyst ⇨ at the same level as the mass.* **(Right)** *The probe ⇨ and grounding pad ⇨ cables are attached to the generator. There are displays for impedance ⇨ and for current and temperature ⇨. During active ablation, the temperature is low as a result of internal cooling. At the end of RFA, the cooling is shut off and the actual temperature of the ablation is displayed. The electric pump ⇨ for providing internal cooling is also seen.*

Radiofrequency Ablation of RCC (Post-Treatment NECT Imaging)

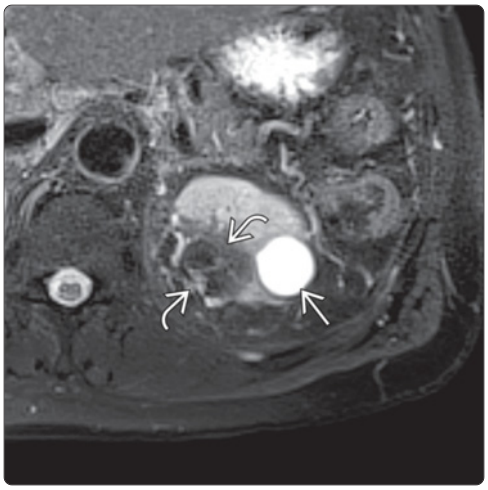


Radiofrequency Ablation of RCC (Post-Treatment MR at 1 Month)



(Left) *Two overlapping ablations were performed with adjustment and repeat NECT confirmation of the probe position between each treatment. A postprocedure NECT demonstrates a small posterior collection of perirenal blood adjacent to the ablation zone, which is a typical finding.* **(Right)** *Axial T1WI FS MR performed 1 month post RFA demonstrates T1 hyperintensity in the ablation zone, which is compatible with coagulation necrosis. Note the T1-hypointense simple renal cyst.*

Radiofrequency Ablation of RCC (Post-Treatment MR at 1 Month)

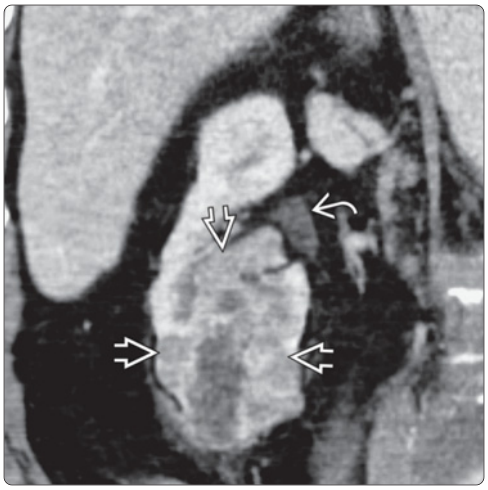


Radiofrequency Ablation of RCC (Post-Treatment MR at 1 Month)

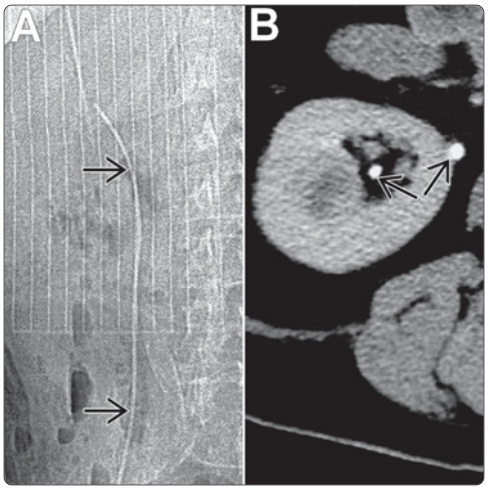


(Left) *T2 FSE image 1 month post RFA shows predominantly ↓ T2 signal in the ablation zone, which is an expected finding. Note the ↑ T2 signal cyst.* **(Right)** *Gd-enhanced T1WI FS MR shows no enhancement in the ablation zone, which is the best imaging correlate of tumor necrosis. Comparison to the pre-RFA CECT shows that the ablation zone encompasses the lesion and a surrounding margin. A thin rim (< 5 mm) of enhancement may occasionally be seen early on, which is a benign ancillary finding.*

Radiofrequency Ablation Planning (Potential Risk of Thermal Injury)

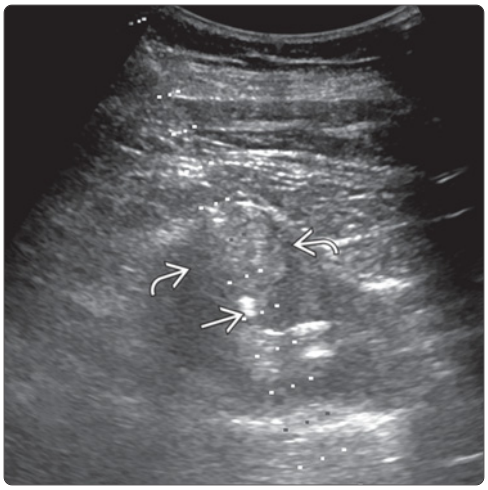


Radiofrequency Ablation Planning (Need for Pyeloperfusion)

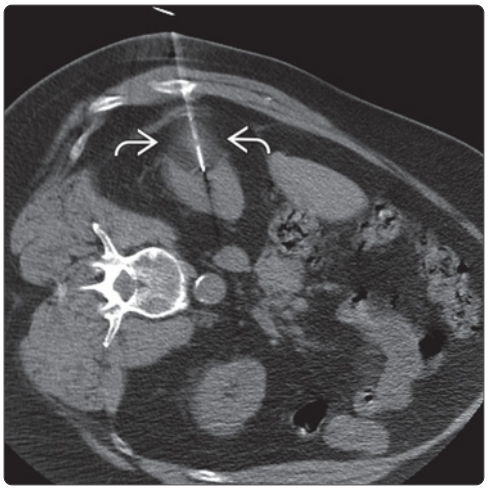


(Left) *Coronal CECT shows a large lower pole renal mass. Note that the mass extends centrally, close to the ureter and the renal pelvis. These structures are at risk of nontarget thermal injury during ablation, which could result in a ureteral stricture &/or urinoma.* **(Right)** *(A) Scout and (B) axial images from a preliminary NECT for RFA show an externalized ureteral stent placed immediately prior to RFA. Pyeloperfusion with D5W protects the renal collecting system from thermal injury during ablation.*

Cryoablation of RCC: Ultrasound Guidance

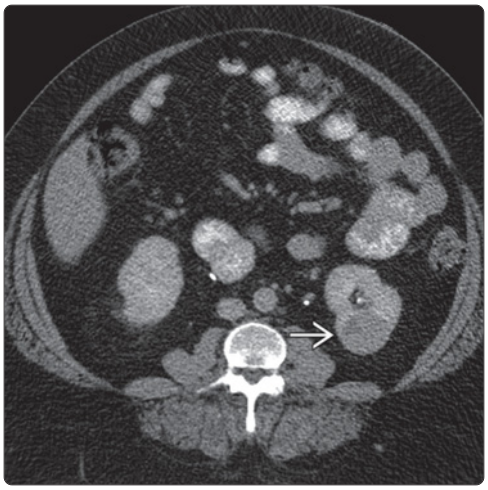


Cryoablation of RCC: NECT Ice Ball Monitor

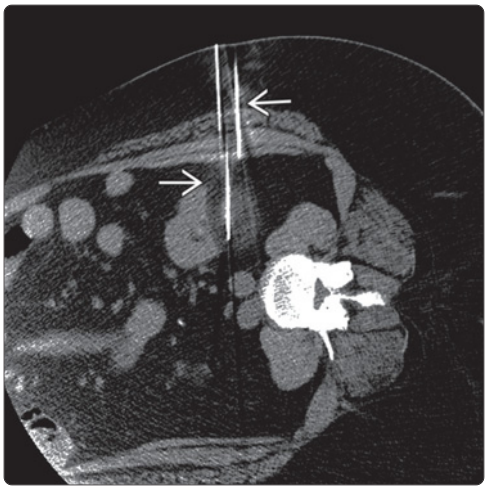


(Left) *Ultrasound guidance may be used for biopsy and initial placement of ablation probes. The distal aspect of 1 of 2 overlapping 17 gauge cryoprobes is shown within the posterior aspect of a biopsy confirmed (echogenic) clear cell RCC. (Right) NECT of the same patient shows a low attenuation ablation zone. Although probes may be placed under ultrasound guidance, ice ball shadowing makes it impossible to monitor the ablation.*

Cryoablation of RCC: Baseline CECT

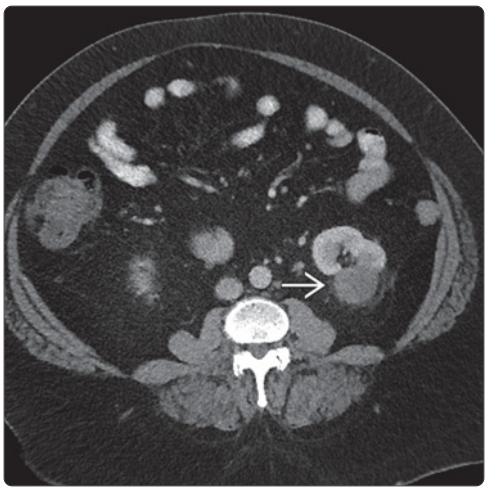


Cryoablation of RCC: Probe Positioning



(Left) *Excretory phase CECT performed on a 57-year-old obese woman with diabetes shows a 2.5 cm non-border deforming renal mass. A biopsy confirmed clear cell RCC.* **(Right)** *The patient was referred for percutaneous cryoablation given her significant comorbidities. The lesion was not visualized at either ultrasound or NECT. Overlapping cryoablation probes were positioned after administration of IV contrast.*

Cryoablation of RCC: Early CECT Surveillance

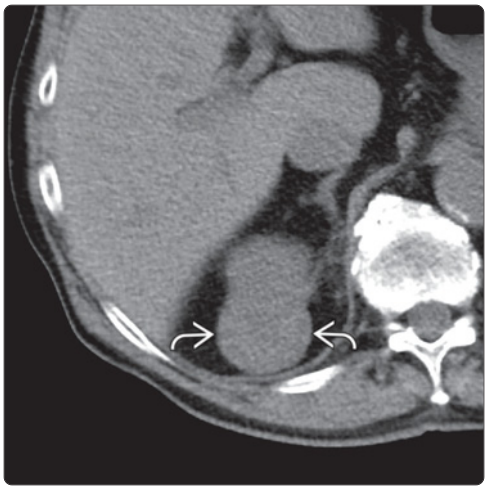


Cryoablation of RCC: Long-Term Surveillance

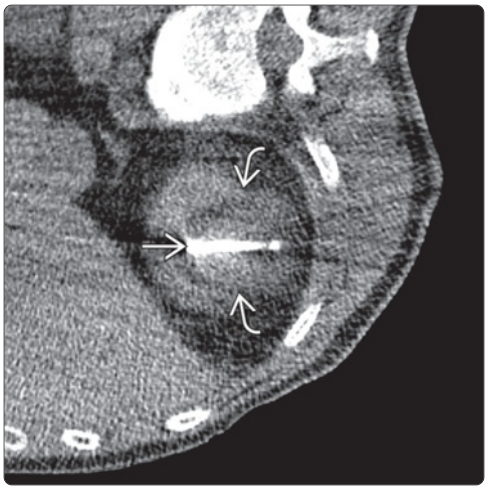


(Left) *Surveillance CECT performed 3 months post treatment on the same patient shows a nonenhancing ablated lesion that is smaller than on previous imaging.* **(Right)** *2 year surveillance CT shows a significantly smaller, nonenhancing ablated lesion, and a partial peripheral fat halo. The renal halo sign is a characteristic feature of renal tumors after RFA, though it may be seen after cryoablation as well.*

Cryoablation of Renal Cell Carcinoma (Initial NECT Imaging Evaluation)

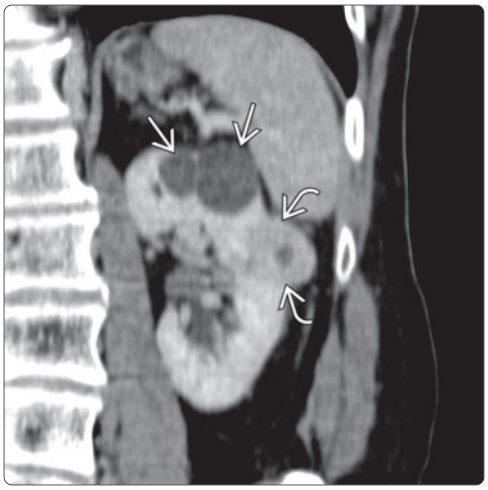


Cryoablation of Renal Cell Carcinoma (Hypodense Ice Ball During Freeze Cycle)

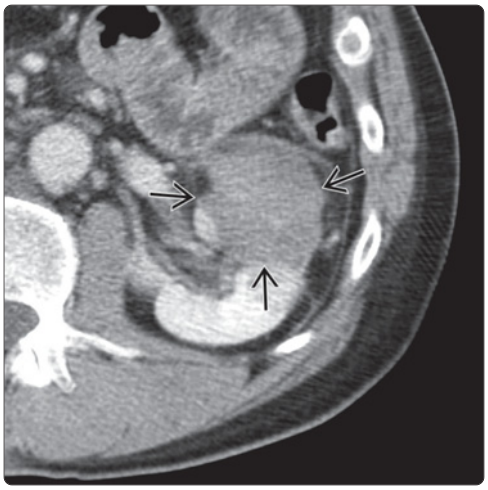


(Left) *Axial NECT shows a renal mass located posteriorly in the upper pole of the right kidney. The patient was not a surgical candidate due to comorbidities, and the renal mass was an appropriate size for ablation.* **(Right)** *NECT obtained 10 minutes into the cryoablation freeze cycle shows 1 of the cryoprobes, as well as the hypodense ice ball encompassing the lesion. The lethal zone is 5 mm internal to the ice ball margin. Typically, a freeze-passive thaw-freeze cycle is performed. Cryoablation may cause less pain than RFA.*

Residual Disease After RFA (CECT Prior to Initial Ablation)

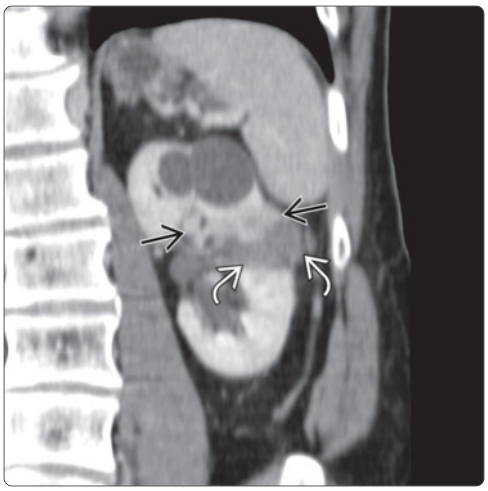


Residual Disease After RFA (CECT After Initial Ablation)

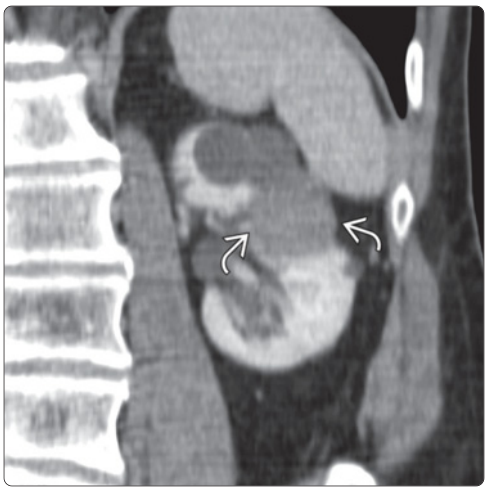


(Left) *Coronal CECT reconstruction prior to planned RFA demonstrates a solid renal mass in the interpolar region of the left kidney. A pair of renal cysts are also evident. Using a cluster electrode, several overlapping RFA treatments were performed.* **(Right)** *Axial CECT obtained 3 months after the RFA demonstrates a nonenhancing zone of ablation, which appears both to completely encompass the lesion and to extend beyond it with a generous surrounding margin.*

Residual Disease After RFA (Coronal CECT After Ablation)

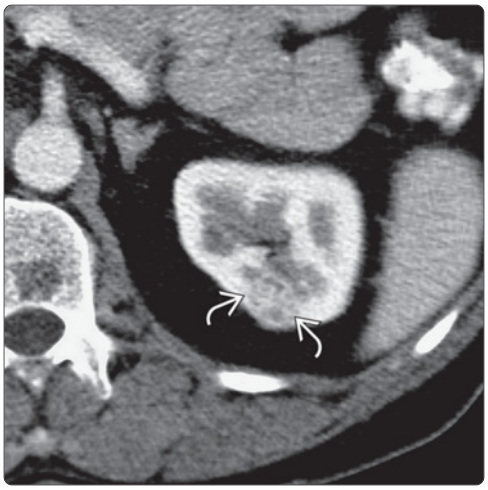


Residual Disease After RFA (Coronal CECT After Retreatment)



(Left) *Although the axial images showed what appeared to be a satisfactory result, a coronal image from the same CECT shows irregular enhancement along the superior margin of the ablation zone. Review of multiplanar reformats may aid assessment of ablation zone margins and is useful in avoiding volume averaging effects.* **(Right)** *Repeat RFA of the residual disease was performed. CECT 1 month later shows no enhancement in the ablation zone, an appearance consistent with tumor necrosis.*

Benign Post-Ablation Ancillary Findings (Initial CECT Evaluation Before RFA)

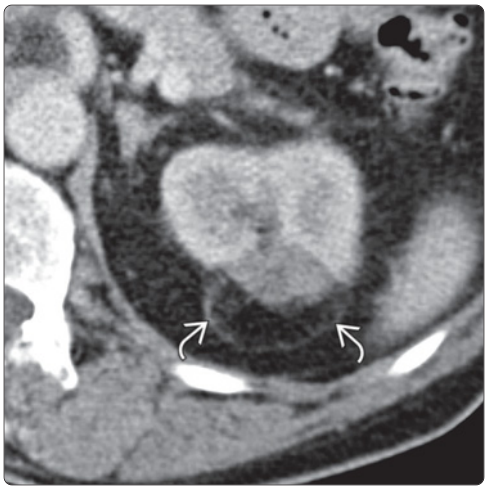


Benign Post-Ablation Ancillary Findings (CECT One Month After Treatment)

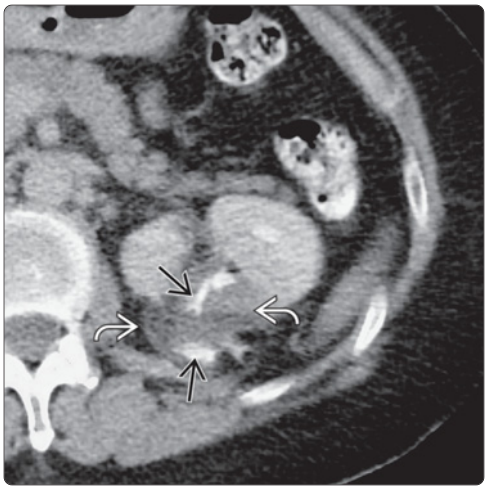


(Left) *Axial CECT obtained prior to a planned RFA demonstrates a small enhancing mass in the posterior aspect of the left kidney. The patient subsequently underwent RFA.* **(Right)** *Axial CECT 1 month post RFA shows a satisfactory lack of enhancement in the ablation zone. However, there is soft tissue stranding in the perirenal fat oriented parallel to the renal contour. This is a typical benign finding that occurs after ablation and should not prompt concern for tract seeding.*

Benign Post-Ablation Ancillary Findings (Fat Halo on CECT 9 Months After RFA)

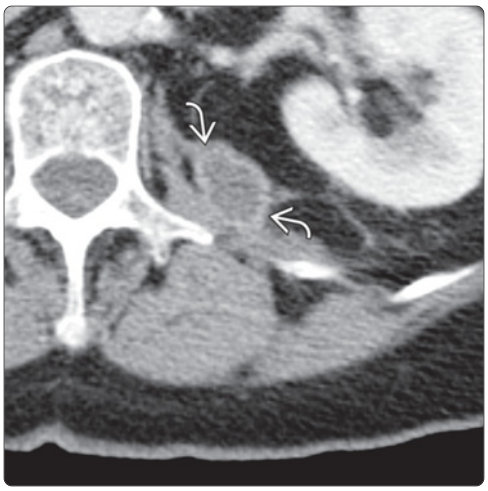


Benign Post-Ablation Ancillary Findings (Extracalyceal Contrast Accumulation)

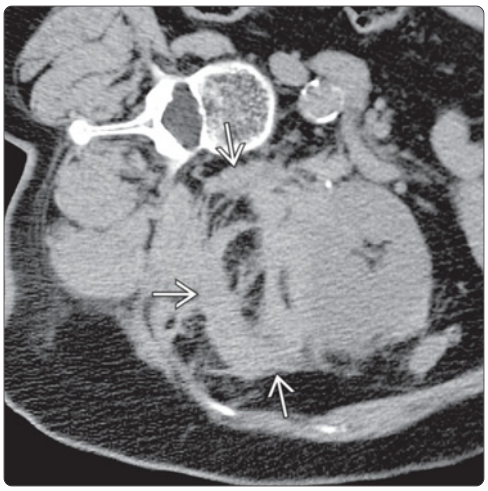


(Left) *Axial CECT image obtained 9 months after RFA shows that there has been coalescence of the perirenal stranding into a fat halo. The ablation zone and fat halo may shrink over serial imaging.* **(Right)** *Axial CECT image obtained during the excretory phase demonstrates strands of extracalyceal contrast in the ablation zone. This may be seen on serial CECT exams but is present only in the excretory phase. In the absence of downstream obstruction in the renal collecting system, this is a benign ancillary finding.*

Benign Post-Ablation Ancillary Findings (Probe Tract Pseudoseeding After RFA)

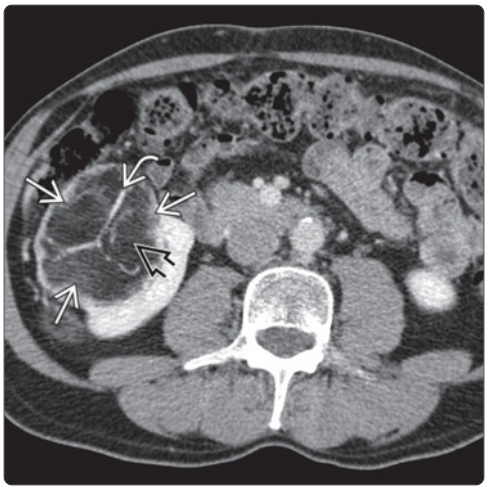


Complication after RFA (Perirenal Hematoma Formation)

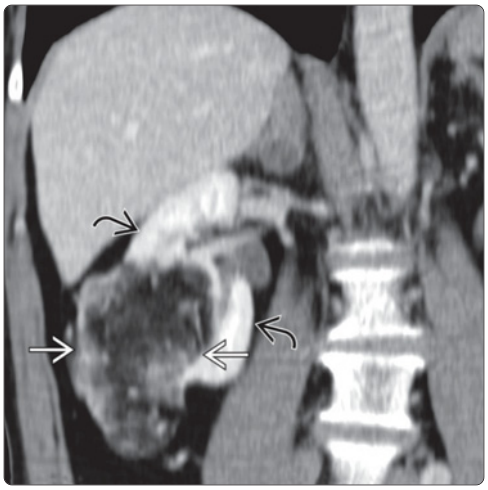


(Left) *CECT obtained 1 month after RFA shows a rim-enhancing density posterior to the kidney along the RFA probe tract. This abnormality involuted over serial CT studies. While true tumor seeding is exceedingly rare, enlarging or persistent suspicious soft tissue may prompt biopsy.* **(Right)** *NECT obtained immediately after RFA shows a posterior perirenal hematoma. Mild perirenal hemorrhage can be expected after ablation. Rarely is blood loss significant enough to require or prolong hospital admission.*

Angiomyolipoma Embolization (Initial CECT Imaging Evaluation)

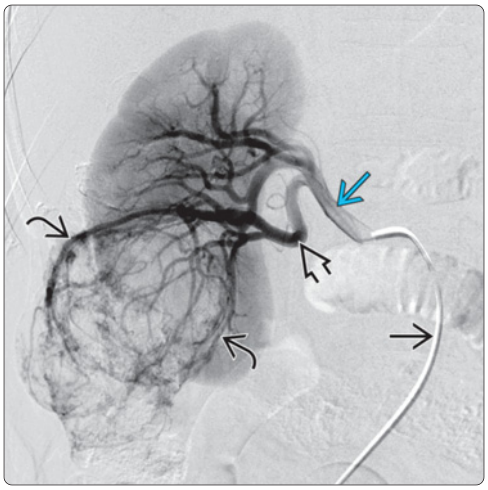


Angiomyolipoma Embolization (Initial CECT Imaging Evaluation)



(Left) *Axial CECT shows a large, right lower pole angiomyolipoma (AML). The mass has a large fat component but also contains numerous enhancing vascular structures, which is typical of AMLs.* **(Right)** *Coronal CECT shows the large right renal mass and its relationship to the normal renal parenchyma. Size is a rough measure of the rupture potential of an AML, and tumors > 4 cm are often prophylactically embolized, especially if they are vascular.*

Angiomyolipoma Embolization (Initial DSA Imaging Evaluation)

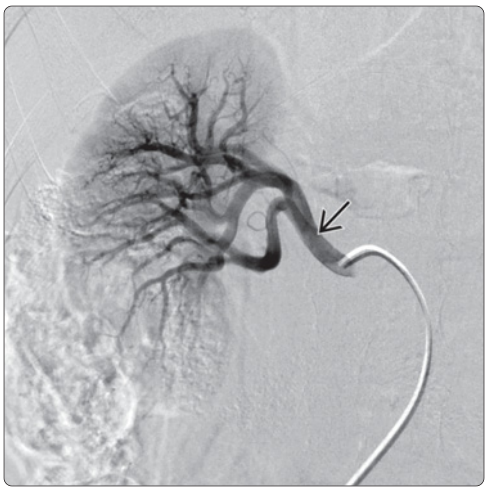


Angiomyolipoma Embolization (Superselective DSA via Microcatheter)

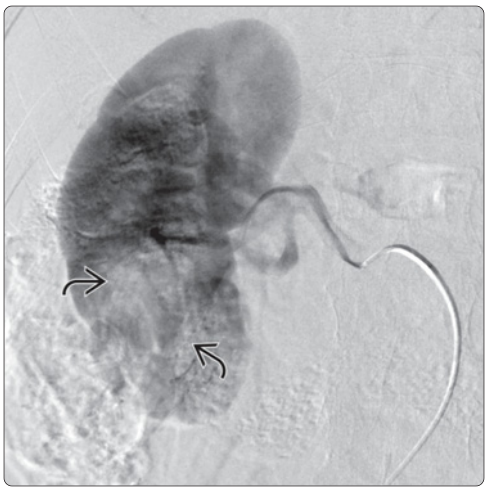


(Left) *Selective DSA via a Cobra catheter placed in the right renal artery shows there is a prominent vascular component to the AML and that the tumoral blood supply arises from an enlarged lower pole arterial branch.* **(Right)** *Superselective DSA via a coaxial microcatheter prior to embolization shows typical intratumoral vascularity but no arterial aneurysms. The goal of AML embolization is tumor devascularization to limit or prevent growth &/or spontaneous rupture.*

Angiomyolipoma Embolization (Renal DSA After Embolization)

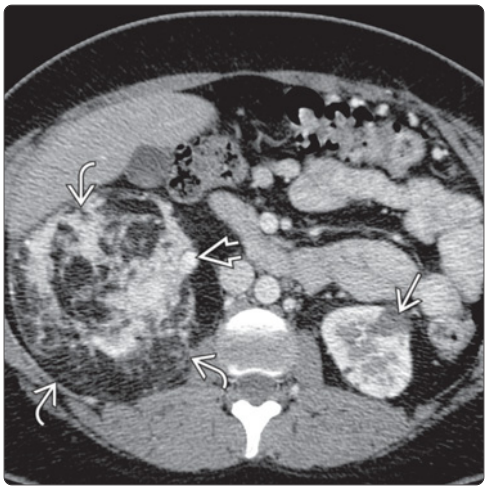


Angiomyolipoma Embolization (Renal DSA After Embolization)



(Left) *After embolization with 300-500 µm particles via the microcatheter, a repeat renal DSA was performed. The arterial phase shows that the AML has been successfully devascularized, as no tumor vascularity is seen. Absolute alcohol mixed with lipiodol has also been utilized for AML embolization.* **(Right)** *Delayed parenchymal phase image from the selective renal DSA shows preservation of normal renal tissue with an area of slight hypoperfusion corresponding to the treated AML.*

Massive AML Embolization (Initial Evaluation with CECT)

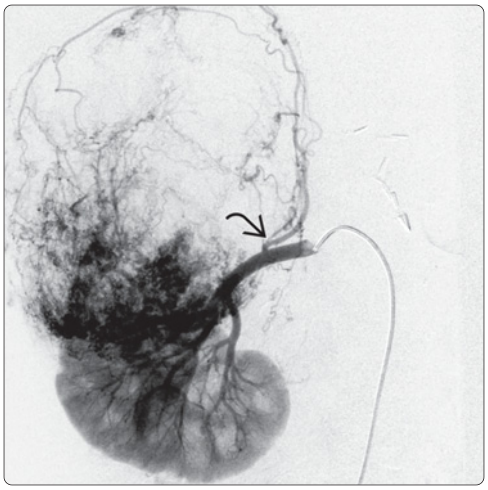


Massive AML Embolization (Initial Evaluation with CECT)

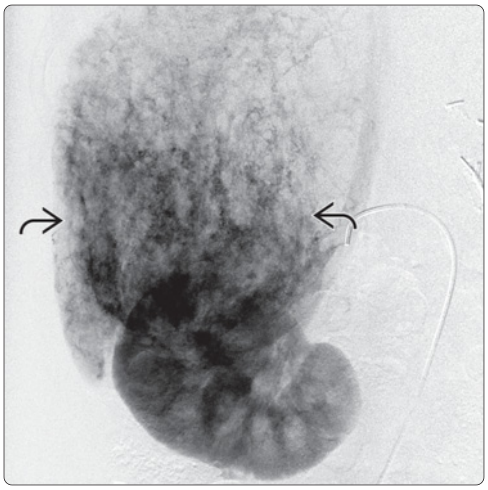


(Left) *Axial CECT obtained in a patient with tuberous sclerosis shows an extremely large, vascular AML. A much smaller 2nd AML is seen in the left kidney. The relationship of the main renal artery to the mass is evident.* **(Right)** *Coronal CT reconstruction shows the massive size of the AML and the extent of residual normal renal parenchyma. It is important to preserve normal renal tissue during embolization, particularly given the number of AMLs.*

Massive AML Embolization (Initial DSA Imaging Evaluation)

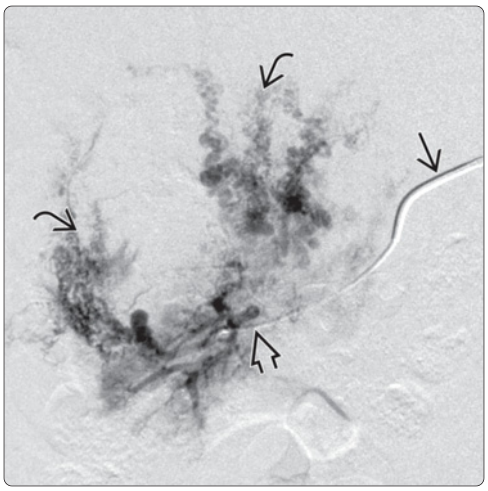


Massive AML Embolization (Initial DSA Imaging Evaluation)

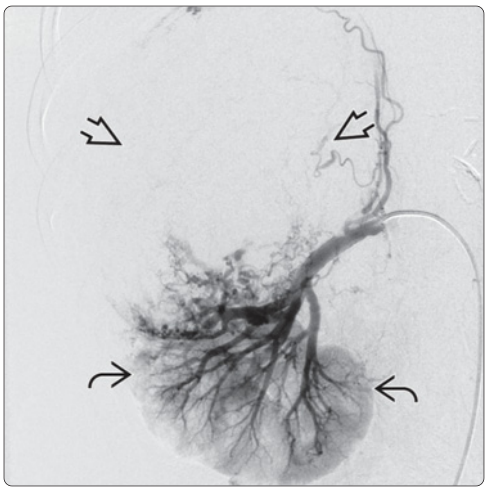


(Left) *Arterial phase renal DSA provides important information regarding the arterial supply and serves as a road map for embolization. The main supply to the AML is through middle and upper pole renal branches, but the AML is also supplied by hypertrophied capsular branches.* **(Right)** *The parenchymal phase from the DSA shows marked vascularity of the AML. Although increased size is a predictor of the rupture potential of an AML, tumor composition is important, as predominantly fatty tumors are unlikely to bleed.*

Massive AML Embolization (Superselective DSA via Microcatheter)



Massive AML Embolization (Renal DSA After Embolization)



(Left) *A microcatheter was coaxially introduced through a Cobra catheter in the main right renal artery and has been positioned more selectively beyond the arterial supply to the residual kidney. DSA shows only tumor vascularity at this catheter position. Particle embolization was performed and additional branches supplying the tumor were also embolized.* **(Right)** *Right renal DSA following embolization shows marked tumor devascularization and preservation of residual normal renal parenchyma.*

Post Kidney Transplant Procedures

KEY FACTS

TERMINOLOGY

- Transplant renal disease processes
  - Transplant renal artery stenosis (TRAS): Luminal narrowing involving or near anastomosis
  - Inflow aortoiliac disease ("pseudo-TRAS")
  - Renal artery/vein thrombosis
  - Arteriovenous fistula/pseudoaneurysm
  - Graft intolerance syndrome: Flu-like syndrome due to renal transplant rejection
  - Urinary obstruction/leak
  - Perigraft fluid collection (e.g., hematoma, abscess)

PREPROCEDURE

- Review procedure indications; prior imaging
- Review operative history to determine anatomy of arterial/venous/ureteral anastomoses

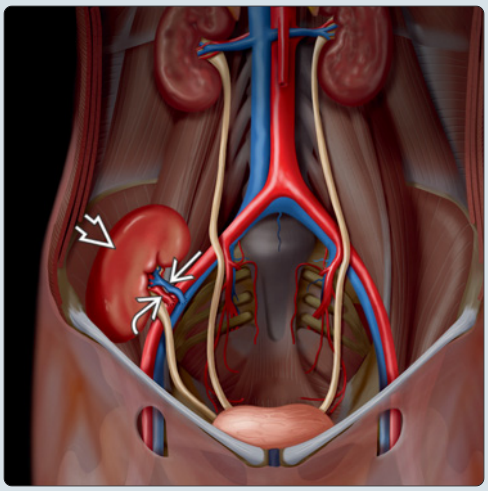
PROCEDURE

- Transplant renal artery stenosis: PTA ± stent
- Aortoiliac inflow disease: Stent placement
- AV fistula/pseudoaneurysm: Coil embolization
- Graft intolerance syndrome: Ethanol ablation
- Urinary obstruction: Percutaneous nephrostomy/stent; ureteral balloon dilatation
- Perigraft collection: Aspiration/drainage

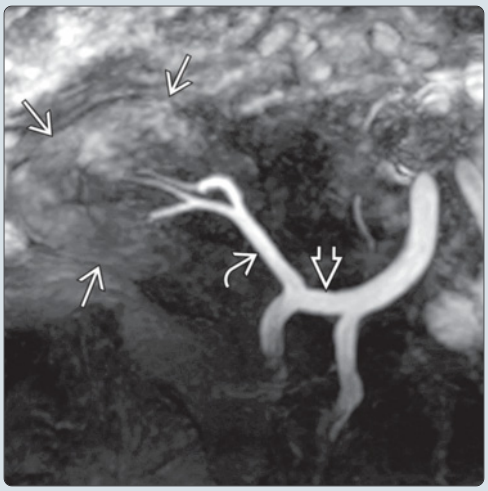
POST PROCEDURE

- Clinical success
  - Transplant renal artery stenosis: 82%
  - AV fistula/pseudoaneurysm: 57-88%
  - Ureteral balloon dilatation/stenting: 73-100%
  - Lymphocele drainage/sclerosis: 68-100%

Renal Transplantation: Normal Anatomy

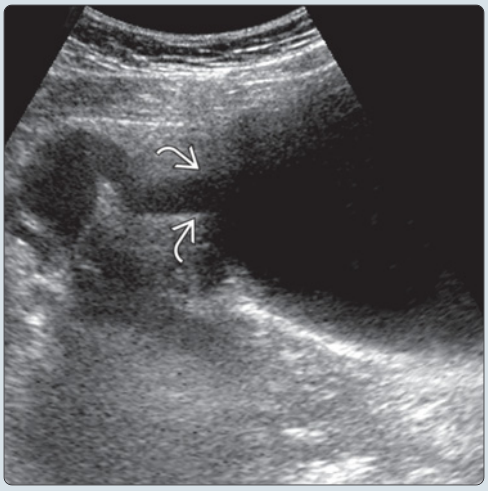


MRA: Normal Transplant Arterial Anatomy

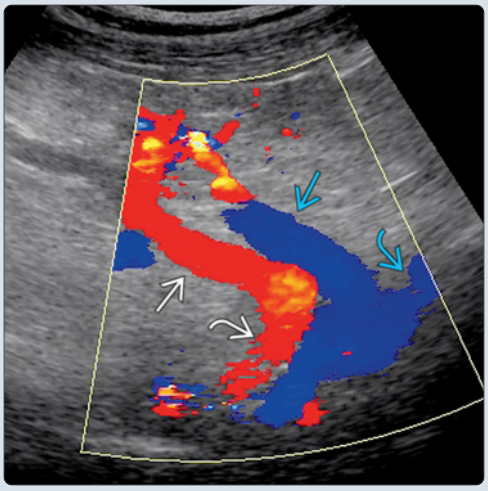


(Left) *Graphic shows a renal allograft in the right iliac fossa in a retroperitoneal location, with the ureter tunneled into the dome of the bladder. The renal artery and vein are anastomosed to the recipient external iliac vessels.* **(Right)** *Coronal noncontrast MRA shows normal arterial anatomy of a right renal allograft. The donor renal artery is anastomosed to the recipient's right external iliac artery, with no evidence of an anastomotic stenosis.*

Normal Transplant Anatomy: Ureter



Renal Transplant Anatomy: Color Doppler Map



(Left) *A ureteroneocystostomy at the bladder dome is particularly well shown in this patient because of severe reflux. Complications at the ureteroneocystostomy include reflux (despite tunneling techniques), obstruction, and leaks. Assessment of collecting system dilatation after voiding helps to differentiate between reflux and true obstruction.* **(Right)** *Color Doppler screen shows normal renal vascular anatomy. Note the end-to-side renal artery to external iliac artery and the renal vein to external iliac vein anastomoses.*

TERMINOLOGY

Definitions

- **Transplant renal artery stenosis (TRAS)**: Hemodynamically significant narrowing of donor renal artery; often juxtaanastomotic in location; incidence of 3-12%
  - Clinical manifestations
    - Refractory hypertension
    - Renal insufficiency
  - Causes
    - Surgical arterial injury (e.g., anastomotic stenosis)
    - Extrinsic compression by perigraft fibrosis
    - Arterial kinking
  - Proximal "pseudostenosis" of transplant renal artery
    - Actually due to inflow aortoiliac atherosclerosis
      - Stenosis proximal to transplant artery
      - Incidence up to 2.4%; increases with age
- **Renal graft thrombosis**: Acute thrombotic occlusion of transplant renal artery/vein (incidence up to 6%)
  - **Transplant renal artery thrombosis**
    - Etiologies
      - Transplant renal artery stenosis
      - Rejection
    - 80% occur in 1st month
      - Most common cause of graft loss in 1st week
    - Surgical therapy generally preferred
  - **Transplant renal vein thrombosis**
    - Etiologies
      - Iliofemoral deep vein thrombosis (DVT)
      - Compression by perigraft collection
    - Treated with anticoagulation &/or surgery
- **Arteriovenous fistula (AVF)/pseudoaneurysm (PSA)**
  - **AVF**: Abnormal arteriovenous communication
    - Usually involves injury to both artery and vein
    - Acquired abnormality in renal transplant
      - Typically occurs after transplant biopsy
    - Clinical features: Hematuria, renal insufficiency, hypertension, high-output failure, bruit
    - May be asymptomatic; may spontaneously resolve
      - Endovascular therapy if symptomatic/enlarging
  - **PSA**: Contained leak from injured artery with connecting tract to sac-like blood collection contained in perivascular space
    - Incidence: 1-18% after biopsy
    - Clinical features: Hematuria
    - Coil embolization if symptomatic/enlarging
    - AVF and PSA may coexist
- **Graft intolerance syndrome**
  - Syndrome due to graft rejection
    - Fever, flu-like syndrome, local pain
  - Consider percutaneous/transcatheter ablation of renal transplant with ethanol prior to nephrectomy to reduce bleeding
- **Urinary obstruction/leak**
  - Incidence: 3-10%
  - Etiologies
    - Often due to distal ureteral ischemia
      - Ureteral vascular supply arises from renal hilum
    - Obstructing calculus
    - Extrinsic ureteral compression
  - Can diagnose leak by creatinine level in urinoma
    - High infection risk if immunocompromised
    - May require both percutaneous nephrostomy (PCN) and urinoma drain for treatment
- **Perigraft fluid collection**
  - Occurs in up to 50% of patients
    - 15-20% clinically significant due to pain or transplant dysfunction (vascular/ureteral compression)
  - Etiologies in early postoperative period: Abscess, hematoma, urinoma
  - Etiology after 1st month: Lymphocele

PREPROCEDURE

Indications

- Angioplasty/stenting
  - Transplant renal artery stenosis
  - Aortoiliac inflow stenosis
    - Compromised transplant renal artery inflow
  - Arterial dissection
- Thrombolysis
  - Renal artery thrombosis
    - Low clot burden
    - Poor surgical candidate
- Embolization
  - Arteriovenous fistula/pseudoaneurysm
- Percutaneous transplant renal biopsy
  - Tissue evaluation for suspected rejection
- PCN/ureteral stent
  - Ureteral stenosis/urine leak-urinoma
- Ureteral balloon dilation
  - Ureteral stenosis
- Aspiration/drainage
  - Perigraft fluid collection

Contraindications

- Lack of safe access for intervention
- Uncorrectable coagulopathy
- Arterial kinking
  - Better suited to surgical management
- Renal artery thrombosis with high clot burden or occurring within first 2 weeks after surgery
  - Surgical exploration preferred
  - Risk to anastomosis in 1st 2 weeks

Preprocedure Imaging

- **Ultrasound (US)**
  - Arterial stenosis
    - Elevated peak systolic velocity > 200-250 cm/s
    - Low pulsatility index, tardus parvus waveform, prolonged (> 100 ms) acceleration time
    - 2-fold gradient between systolic velocities at, and proximal to, stenosis
  - Arteriovenous fistula
    - Abnormal artery/vein communication
    - High velocity, low "resistance" (i.e., relatively high diastolic) flow within feeding artery
    - Pulsatile flow within draining vein
  - Pseudoaneurysm
    - Pulsatile color Doppler flow within PSA sac

  - Swirling flow/turbulence/"yin-yang" sign in sac
  - Tract/neck connects PSA and feeding vessel
  - Biphasic arterial Doppler signal in tract/neck
- Renal artery thrombosis
  - Lack of main or intrarenal artery flow
- Renal vein thrombosis
  - Lack of renal venous flow, reversed intrarenal diastolic flow
  - Enlarged kidney (allograft edema)
- Perigraft fluid collection
  - Abscess
    - Hypoechoic/mixed echogenicity collection
  - Hematoma
    - Echogenic components if acute
    - Hypoechoic or anechoic while resolving
  - Lymphocele
    - Rounded anechoic collection ± septa

- **Magnetic resonance (MR)/computed tomography (CT)**
  - May demonstrate location of stenosis
    - Involving or adjacent to arterial anastomosis
    - Aortoiliac inflow disease
    - Tortuous/kinked artery may be well demonstrated
  - **Magnetic resonance angiography (MRA)**
    - May be performed ± gadolinium
      - Glomerular filtration rate (eGFR) important
      - Risk of nephrogenic systemic fibrosis (NSF) if severe renal dysfunction and gadolinium given
    - Susceptible to artifact from metallic stents
  - **Computed tomography (CT)**
    - Good for evaluating hydronephrosis
    - Characterization of perigraft fluid collection
      - Abscess may contain gas
      - Hematoma may contain hyperdense areas
  - **Computed tomographic angiography (CTA)**
    - Requires iodinated contrast
    - Risk of contrast-induced nephropathy
- **Digital subtraction angiography (DSA)**
  - Gold standard for diagnosis/endovascular treatment of transplant renal artery stenosis
    - Issues: Invasive, ionizing radiation, contrast-induced nephropathy risk (consider CO2 angiography)
  - Provides guidance for intervention
- **Renal nuclear scintigram**
  - Confirm urinary obstruction/leak
- **Retrograde pyelogram**
  - If ureteroureterostomy/ureteropyelostomy present
  - Evaluate urinary obstruction/leak

Getting Started

- Things to check
  - Transplant operative history
    - Cadaveric or living donor
    - Arterial anastomosis
      - End-to-side of common/external iliac artery
      - End-to-end of internal iliac artery (living donor)
    - Venous anastomosis
      - End-to-side of common/external iliac vein
    - Ureteral anastomosis
      - Ureteroneocystostomy (most common)
      - Ureteroureterostomy
      - Ureteropyelostomy
    - Children < 20 kg
      - Transplant more cephalad in location
      - End-to-side anastomoses to distal aorta and IVC
  - Pertinent clinical history and physical examination
    - Procedure indications; prior imaging
    - Current medications (anticoagulants, antiplatelet agents)
    - Allergies
  - Laboratory parameters
    - Complete blood count (CBC)
      - Platelet > 50,000/μL
    - Coagulation profile
      - International normalized ratio (INR) ≤ 1.5
      - Normal prothrombin time (PT), partial thromboplastin time
    - Estimated eGFR
      - Reduced eGFR may impact procedure
    - Cyclosporine level
      - Is graft dysfunction due to toxicity
- Medications
  - Anesthesia
    - 1% lidocaine local anesthetic
    - Adequate procedural sedation
      - Fentanyl, midazolam typically used
    - Anesthesiology assistance if necessary
  - Prophylactic antibiotics
    - Percutaneous nephrostomy
    - Perigraft collection aspiration/drainage
      - e.g., ampicillin (1 g), gentamicin (80 mg)
  - Heparin
    - Administered prior to crossing arterial stenosis
  - Vasodilator (e.g., nitroglycerin)
    - Typical bolus dose of 100 μg nitroglycerin
    - Prevents/treats catheter-induced vasospasm
  - Thrombolytic agent
    - e.g., tissue plasminogen activator (tPA)
- Equipment list
  - **General**
    - Ultrasound equipment: 3-6 MHz curved transducer, sterile ultrasound probe cover/gel
    - Fluoroscopy equipment (C-arm)
    - Computed tomography equipment: Radiopaque skin grid
  - **Arterial revascularization/exclusion**
    - Micropuncture access set
    - Catheters/sheaths
      - 5-6 Fr arterial access sheath
      - Flush catheter (e.g., pigtail, Omni Flush)
      - Selective catheter (e.g., Cobra-2)
      - Guiding catheter/sheath; permits contrast injection around stent during placement
      - Microcatheter (primarily for embolization)
    - Guidewires
      - Initial 0.035" guidewire
      - Hydrophilic guidewire
      - Floppy tip guidewire for crossing stenosis
      - 0.014-0.018" microwire
      - Stiff guidewire (e.g., Rosen, McNamara)

- Angioplasty (PTA) balloon
  - Match balloon and lesion lengths
  - Over-the-wire vs. rapid exchange system
- Intravascular stents
  - Prefer high radial force balloon-mounted stent
  - Introduced coaxially via guiding catheter; rapid exchange system
- Embolic agents
  - Embolization coils
- Iso-osmolar iodinated contrast
  - Use small volumes of dilute contrast
  - Consider CO_2 contrast use if renal dysfunction

- **Thrombolysis**
  - Infusion catheter
- **Biopsy**
  - 16 or 18g spring-loaded core biopsy gun
- **Percutaneous nephrostomy/ureteral stent**
  - Percutaneous access needles
    - 21-22-g Chiba needle
    - 18-g Longdwell needle (e.g., Ring) if 2-stick technique used
  - Guidewires
    - 0.018" mandril or nitinol guidewire
    - 0.035" 3-J guidewire
  - Catheters/sheaths
    - 5-6 Fr coaxial dilator-sheath assembly
    - 5 Fr directional catheter (e.g., Kumpe)
    - 8 Fr locking pigtail nephrostomy catheter
  - Ureteral stent
    - 9-10 Fr peel-away sheath
    - 8 Fr double J ureteral stent
- **Ureteral balloon dilatation**
  - 0.035" stiff guidewire (e.g., Amplatz)
  - Angioplasty balloon (5-8 mm diameter)
- **Aspiration/drainage**
  - Direct trocar technique
    - 8-14 Fr trocar-mounted locking pigtail catheter
  - Tandem-trocar technique
    - 20-g Chiba guiding needle
    - 8-14 Fr locking pigtail catheter
  - Seldinger technique
    - Access needle (e.g., 18-g Longdwell needle)
    - 0.035" 3-J guidewire
    - 8-14 Fr dilators as appropriate
    - 8-14 Fr locking pigtail catheter

PROCEDURE

Equipment Preparation

- Flush catheters/sheaths with heparinized saline

Procedure Steps

- **Arterial revascularization for stenosis**
  - Obtain arterial access; place access sheath
  - Perform nonselective CO_2 aortoiliac DSA
    - Position flush catheter above aortic bifurcation
    - Assess for inflow atherosclerotic stenosis
  - If TRAS shown by other imaging (e.g., US), proceed directly to selective DSA of inflow/target artery
  - Perform common/external/internal iliac DSA
    - Target artery depends on anastomosis location
    - Usually requires multiple projections
    - Stenosis typically near anastomosis
  - Place guiding catheter/sheath in target artery
  - Advance 4-5 Fr coaxial catheter proximal to stenosis
    - Inject contrast; obtain DSA images
      - Use either CO_2 or small volume of dilute iodinated contrast medium
  - Evaluate length and severity of stenosis
    - Criteria
      - Luminal narrowing > 50%
      - Pressure gradient > 10% of peak systolic blood pressure
  - Administer IV heparin
    - 70 U/kg bolus or empiric 2,500-5,000 units
      - Goal: Activated clotting time (ACT) 250-300 s
  - Consider intraarterial nitroglycerin (100-200 µg)
    - Administer on "as needed" basis for vasospasm
  - Traverse stenosis
    - Advance floppy-tip guidewire through stenosis
      - Stop advancing if resistance encountered
      - Assess if guidewire passed subintimally (avoid dissection)
    - Advance selective catheter over guidewire
    - Exchange for stiff guidewire (e.g., Rosen/McNamara)
  - Proceed to angioplasty &/or stenting
  - **Percutaneous transluminal angioplasty (PTA)**
    - Match balloon size to normal artery diameter
      - Consider using coaxial rapid-exchange system; smaller caliber system, less traumatic
    - Maintain guidewire across lesion
    - Center angioplasty balloon with lesion
    - Use insufflator for balloon inflation
      - Inflate with 50/50 contrast/saline mix
    - Balloon "waist" typically created by stenosis
      - With continued inflation, "waist" should release
    - Deflate and remove angioplasty balloon catheter
      - Maintain guidewire across lesion: Avoid recrossing lesion with guidewire (dissection risk)
    - Inject contrast via guiding catheter/sheath
      - Document angioplasty result
      - Technical success = residual stenosis < 20-30%
  - **Intravascular stenting**
    - Indications
      - Aortoiliac inflow disease
      - > 30% residual stenosis after angioplasty of TRAS
      - Flow-limiting dissection
    - Select appropriate balloon-mounted stent
      - Consider using guiding catheter and coaxial rapid-exchange system
    - Position stent
      - Stent should encompass entire stenosis
      - Inject guiding catheter/sheath during deployment; confirm position
    - Retract guiding catheter/sheath
    - Fully inflate balloon to deploy stent
    - If self-expanding stent used
      - Retract outer constraining sheath of delivery system while maintaining stent position
      - Perform post-deployment angioplasty

- Perform completion DSA arteriogram: Determine technical success and document results
- Remove catheters/sheath; obtain hemostasis

- **Thrombolysis of renal artery thrombosis**
  - Selectively catheterize thrombosed target artery
    - Advance guidewire through thrombosed lumen
  - Administer IV heparin
  - Exchange over guidewire for infusion catheter
  - Place tip of infusion catheter beyond anastomosis
  - Begin infusion with thrombolytic agent (e.g., tPA)
    - Usually requires continuous infusion of thrombolytic agent
  - Admit to ICU for appropriate monitoring
  - Obtain DSA after thrombolysis; assess for TRAS
    - Treat underlying stenosis with angioplasty/stent
    - Arterial kinking may require surgical correction
- **Arteriovenous fistula/pseudoaneurysm exclusion**
  - **General**
    - Obtain DSA of transplant kidney vasculature
    - Introduce microcatheter into target artery
    - Selectively catheterize branch leading to PSA/AVF
    - Perform coil embolization
  - **Pseudoaneurysm**
    - Position catheter tip within pseudoaneurysm sac
    - Tightly deposit coils in sac to thrombose
    - May sometimes need to place coils in arterial lumen proximal/distal to pseudoaneurysm, particularly if wide-mouthed PSA
    - Also consider stent-assisted coil embolization
  - **Arteriovenous fistula**
    - High arterial flow risks nontarget embolization
      - Consider using detachable coils; more precise
    - May require simultaneous arterial/venous catheterization
      - Position balloon in venous side
      - Inflate balloon to reduce flow
      - Deposit coils along fistula from arterial side
  - Perform completion angiogram
    - May be unable to completely obliterate PSA/AVF with coil embolization, particularly if large/high flow lesions
- **Ethanol ablation of rejected transplant**
  - Selectively catheterize transplant renal artery
  - Perform renal DSA; exclude any other abnormality
  - Exchange diagnostic for balloon occlusion catheter
  - Inflate balloon to occlude proximal renal artery
  - Inject contrast to visualize renal branch vessels
    - Note contrast volume used to visualize branches
  - Slowly inject similar volume of absolute ethanol
  - Repeat contrast injection; confirm ablated vessels
- **Biopsy**
  - Single-needle technique usually appropriate
  - Use real-time US guidance; advance needle to target
  - Orient trajectory toward upper or lower pole, avoid hilum
  - Enter cortex; maintain trajectory parallel to cortex: Obtain biopsy
  - May repeat if additional samples desired
  - Coaxial technique may also be considered
  - Perform postbiopsy ultrasound
    - Assess for perirenal hematoma
  - Use CT guidance if poor US visualization (rare)
- **Percutaneous nephrostomy for urinary obstruction/leak**
  - Visualize transplant kidney with US
  - Select lateral calyx
  - Use 1- or 2-stick technique
  - **Nephroureteral catheter placement**
    - Avoid in septic patient; place percutaneous nephrostomy, delay nephroureteral catheter placement until clinically improved
  - **Antegrade ureteral stent placement**
    - Same steps as nephroureteral catheter placement until Amplatz guidewire extends to bladder
    - Select appropriate length 8 Fr double pigtail stent (pediatric lengths available)
  - **Ureteral balloon dilation**
    - Indicated in high-grade anastomotic stricture
    - Advance Amplatz wire across stenosis
      - May use combination of hydrophilic guidewire and 5 Fr directional catheter to cross stenosis
    - Select balloon size: Should match diameter of normal ureter
    - Advance balloon catheter over guidewire: Position balloon across stenosis
    - Attach insufflator to balloon catheter
    - Insufflate 10-12 atm for 30-120 s
      - May see "waist" at stenosis
      - Continued inflation should eliminate "waist"
    - Repeat dilatation until "waist" disappears (may upsize balloon if "waist" does not disappear)
    - Inject contrast; assess for residual stenosis
    - Place nephroureteral catheter or antegrade stent/safety nephrostomy catheter as previously detailed
- **Aspiration/drainage**
  - Can aspirate/drain most perigraft collections using ultrasound guidance
  - **Aspiration**
    - Localize fluid collection with ultrasound
    - Use Doppler to avoid any intervening vessels
    - Cleanse skin with preparatory solution: 1% lidocaine local anesthetic
    - Nick skin with #11 scalpel blade
    - Advance spinal needle or 7 Fr curved-tip catheter loaded on trocar into collection using real-time US
      - Use needle if diagnostic aspiration only
      - Use catheter if therapeutic/complete aspiration
      - Once catheter tip in collection, unscrew hub and advance catheter into collection
    - Connect to extension tubing and 3-way stopcock
    - Submit specimens for
      - Gram stain, culture, and sensitivity
      - Creatinine assay
      - Cell count
    - Ultrasound to assess residual collection
    - Loculated component may require separate catheter placement
  - **Drainage**
    - Select appropriate catheter size
      - 8-14 Fr generally available
      - 8-10 Fr used in most instances

        - More viscous collections may benefit from 12-14 Fr catheters
    - Direct trocar technique
        - Load locking pigtail catheter over trocar
        - Advance catheter assembly into collection using real-time US guidance
        - Unscrew trocar from catheter once tip of catheter assembly is in collection
        - Stabilize trocar so it no longer advances; advance catheter further into collection
        - Remove trocar; form/lock pigtail
        - Connect to extension tubing, 3-way stopcock, and drainage bag
        - Demonstrate pigtail loop in collection with US
        - Aspirate fluid
        - Confirm interval drainage by US
    - Seldinger technique
        - US and fluoroscopic guidance are complementary
        - Advance access needle (e.g., 18-g) into collection using US guidance
        - Aspirate small amount of fluid
        - May confirm position in collection by contrast injection under fluoroscopic monitoring
        - Inspect fluid for color, purulence, viscosity
        - Select catheter size based on fluid viscosity
        - Advance 0.035" stiff guidewire into collection
        - Remove needle/ring sheath
        - Dilate tract as appropriate
        - Load catheter on stiffening cannula
        - Advance assembly over wire into collection
        - Once assembly tip in collection, unscrew hub
        - While stabilizing cannula so it does not advance, advance catheter over guidewire further into collection
        - Remove stiffener and wire
        - Form/lock pigtail
        - Connect drainage assembly as previously detailed with direct trocar technique
    - CT-guidance rarely required
        - Tandem trocar or Seldinger technique
- Drainage and sclerotherapy of lymphoceles
    - Place drainage catheter into collection
        - Aspirate contents of collection
    - Inject contrast via drainage catheter to exclude communication with renal collecting system
    - Various sclerosants available (e.g., ethanol, fibrin sealant, sodium tetradecyl sulfate, bleomycin)
    - Inject volume of sclerosant < volume of cavity
    - May clamp catheter; allow sclerosant to indwell up to 15 minutes
    - Unclamp catheter and aspirate contents
    - May require several treatments

Findings and Reporting

- Transplant renal artery stenosis intervention
    - Length and severity of stenosis
    - Technical success
        - Degree of residual stenosis
    - Size/type/location of intravascular stent
    - Volume of contrast used and any complications
- Arteriovenous fistula/pseudoaneurysm
    - Location/size of AVF/PSA
    - Type of intervention used for treatment
    - Completion angiogram appearance
- Biopsy
    - Biopsy device used
    - Presence/absence of hematoma after biopsy
- Percutaneous nephrostomy/ureteral intervention
    - PCN/nephroureteral catheter/stent type used
    - Location/length/degree of stenosis of collecting system
- Aspiration/drainage
    - Color/opacity/viscosity of fluid drained
    - Laboratory analyses requested
    - Type of drainage catheter used

Alternative Procedures/Therapies

- Surgical
    - Surgical revascularization
        - Arterial kinking
    - Surgical marsupialization of lymphocele

POST PROCEDURE

Expected Outcome

- Transplant renal artery stenosis
    - Technical success rate reported as high as 94%
    - 95% graft survival post-PTA at 1 year; 82% at 2 years
    - 82% clinical success (defined as)
        - > 15% reduction in serum creatinine
        - > 15% reduction diastolic blood pressure with no change in antihypertensive medications
        - > 10% reduction diastolic blood pressure with reduction in antihypertensive medications
- Arteriovenous fistula/pseudoaneurysm
    - Technical success: 71-100%
    - Clinical success: 57-88%
        - May repeat procedure
    - Minor infarcts (involving < 30% kidney) may occur
        - Usually clinically insignificant
        - Recommend using coils as higher risk of major infarct with particles; risk allograft loss
- Urinary obstruction
    - Clinical success with balloon dilatation/stent placement: 73-100%
- Urine leak
    - Clinical success of urinary diversion: 36-100%
- Aspiration/drainage
    - Abscess: > 90%
    - Lymphocele
        - 80-90% recurrence with aspiration alone
        - 68-100% clinical success when drainage combined with sclerotherapy

Things to Do

- Arterial revascularization
    - Intravenous hydration; monitor blood pressure
    - Medications after stenting
        - Clopidogrel 75 mg daily for 3 months/aspirin 81 mg or 325 mg lifetime
- Percutaneous nephrostomy/nephroureteral catheter
    - Flush catheter with 10 mL normal saline q 8 h

- 5 mL to kidney; 5 mL to drainage bag
- Regular catheter exchanges every 4-12 weeks
  - More frequent if high urinary sediment/clogging
  - May cease regular flushing if no hematuria/clogging of catheter
  - Prophylactic antibiotics prior to every change: Tailor to previous culture results
  - Minimize contrast injection volume/guidewire manipulations
    - Reduces risk of sepsis
- Ureteral stent
  - Consider clamping safety PCN next day
  - Clamp trial may extend 1-7 days
    - Unclamp PCN if pain or fever develops
  - Contrast injection at end of trial
    - If patent stent, may remove PCN
  - Regular retrograde stent changes by urology

- Drainage
  - Record drain output q shift
  - Flush catheter with 10 mL normal saline q 8-12 h
    - 5 mL to cavity; 5 mL to drainage bag via 3-way stopcock
  - Always flush after aspiration
  - Catheter removal criteria
    - Resolution of fever/leukocytosis and hemodynamic stability
    - Drainage catheter output < 10-20 mL/day
    - No fistula or large cavity on catheter injection

Post Procedure Imaging

- Stenosis/arteriovenous fistula/pseudoaneurysm
  - Follow-up duplex Doppler sonography
- Percutaneous nephrostomy/ureteral stent
  - Antegrade nephrostogram when clinically improved

OUTCOMES

Problems

- Restenosis
  - Repeat angioplasty, stent placement
  - In-stent stenosis (13-25% incidence)
    - Consider angioplasty of stent/new stent
    - Cutting balloon angioplasty
- Failure to improve hypertension or eGFR
  - Rejection, technical failure, irreversible intrinsic renal damage

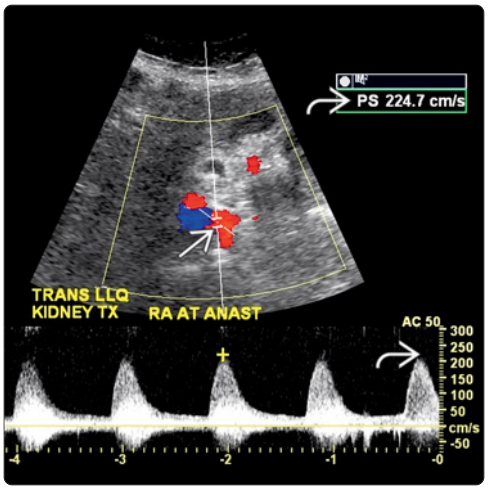
Complications

- Most feared complication(s)
  - Loss of renal allograft
  - Flow-limiting renal artery dissection
    - Maintain guidewire position across dissection
    - Place intravascular stent
  - Renal artery perforation/rupture
    - Maintain guidewire position across lesion
    - Reverse anticoagulation
    - Inflate angioplasty balloon across perforation for arterial tamponade
    - Place covered stent bridging injury
    - Use coil embolization for distal rupture
- Immediate/periprocedural complication(s)
  - Hemorrhage (may see transient hematuria after biopsy or PCN)
  - Infection/sepsis
  - Arteriovenous fistula/pseudoaneurysm
- Delayed complication(s)
  - In-stent restenosis
- Other complications
  - Vascular access complications
  - Bladder irritation/pain from nephroureteral catheter or ureteral stent
    - May require conversion to percutaneous nephrostomy
  - Ureteral stent occlusion
    - Treat with stent exchange
  - Postembolization syndrome

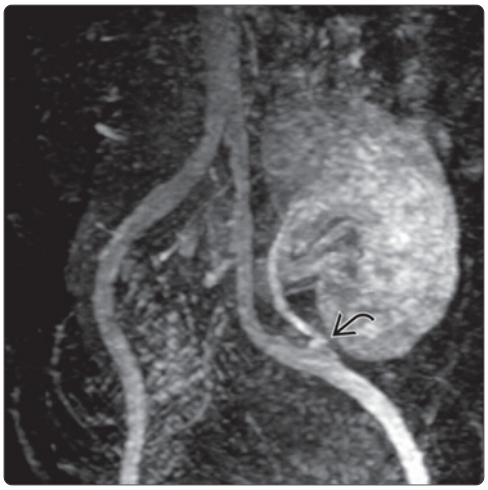
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Transplant Kidney RAS (Ultrasound Evaluation)



Transplant Kidney RAS (Initial MRA Evaluation)



(Left) *Color Doppler US, with the gate ➔ at the anastomosis of the donor renal artery to the left external iliac artery, shows markedly elevated peak systolic velocities ➔, reflecting a high-grade renal artery stenosis. Common causes of stenoses include anastomotic strictures and vascular clamp injuries.* **(Right)** *Oblique MIP from an MRA shows an angulation ⇨ of the donor renal artery near the anastomosis to the external iliac artery, which is the area of the suspected renal artery stenosis.*

Transplant Kidney RAS (Initial MRA Evaluation)

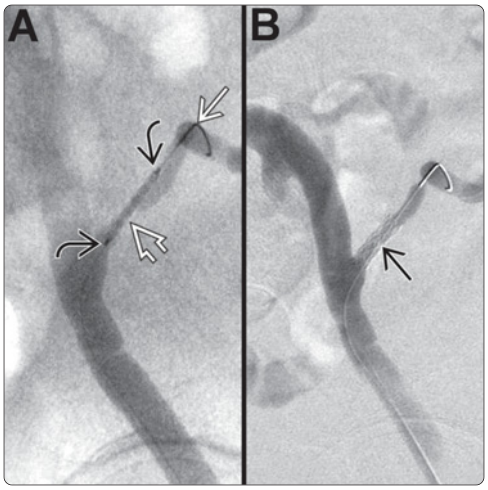


Transplant Kidney RAS (Diagnostic DSA Evaluation)



(Left) *Oblique Gd-enhanced MRA shows a stenosis ➔ of the proximal transplant renal artery in the area of acute angulation near the anastomosis. Based upon the imaging and clinical findings, further evaluation with DSA and potential transcatheter intervention was recommended.* **(Right)** *A catheter was introduced from a left common femoral approach and the tip ➔ was placed in the external iliac artery proximal to the renal artery anastomosis ⇨. The DSA confirms a focal severe renal artery stenosis ➔.*

Transplant Kidney RAS (Intravascular Stent Placement)

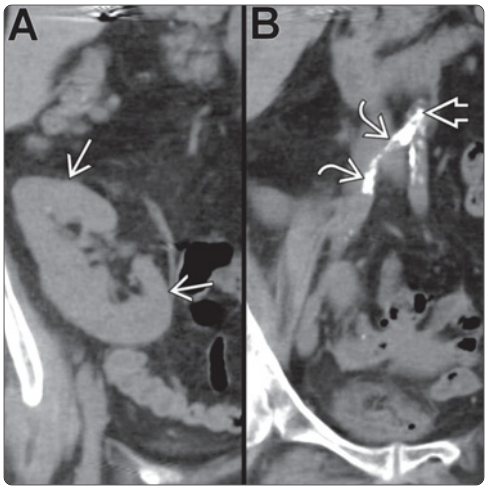


Transplant Kidney RAS (Post-Treatment DSA)

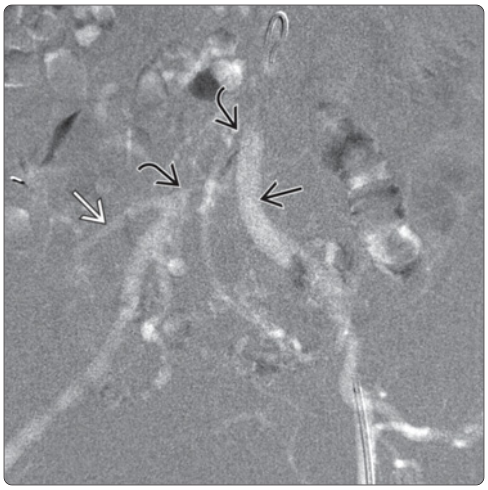


(Left) *Fluoroscopic spot radiographs during stent placement show (A) a balloon-mounted stent ⇨ advanced over a guidewire ➔ that bridges the stenosis ➔. (B) After expanding the stent ⇨, the stenosis has been eliminated.* **(Right)** *A DSA performed after removal of the stent delivery system and guidewire shows an appropriately positioned stent ➔ and no residual stenosis. Intrarenal branches ⇨ fill appropriately, and subsequent images showed no dissection or distal emboli.*

Transplant Recipient Arterial Disease (Evaluation with CT Imaging)

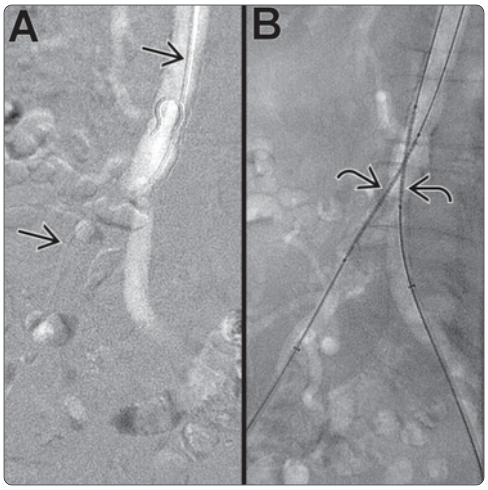


Transplant Recipient Arterial Disease (Evaluation with CO_2 DSA Imaging)

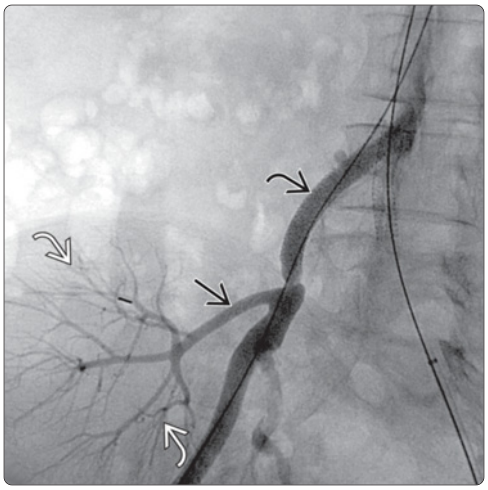


(Left) *Coronal NECT in a failing renal transplant (A) shows a transplant kidney in the right iliac fossa, which was anastomosed end-to-side to the common iliac artery. (B) Extensive calcified plaque in the right common iliac artery extends into the distal abdominal aorta.* **(Right)** *DSA from a left femoral access was performed using CO_2 contrast, given the patient's poor renal function. This shows a patent left common iliac artery and an absent (occluded) right common iliac artery. The transplant renal artery is patent.*

Transplant Recipient Arterial Disease (Stenting of Iliac Artery Occlusion)



Transplant Recipient Arterial Disease (Post-Intervention DSA Imaging)

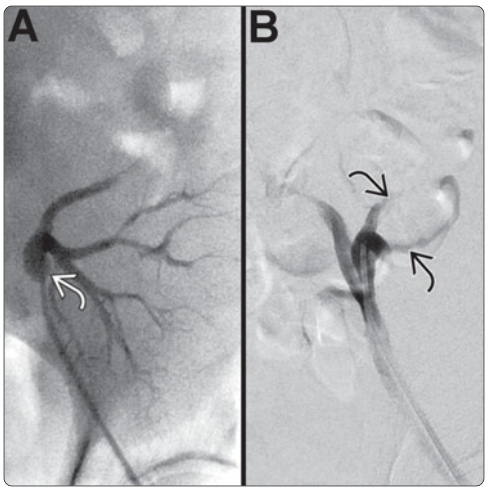


(Left) *(A) The right femoral artery was accessed, and a guidewire and catheter were used to traverse the occluded right common iliac artery. (B) Balloon-mounted stents were introduced bilaterally and were positioned in the common iliac arteries, extending slightly into the distal aorta. They were simultaneously deployed using a "kissing" stent technique.* **(Right)** *DSA after "kissing" stents shows that the right common iliac artery is widely patent. The transplant renal artery and intrarenal branches now fill normally.*

Transcatheter Transplant Ablation (Diagnostic DSA Evaluation)

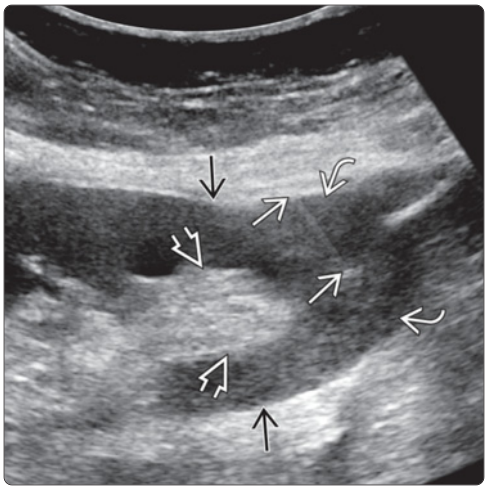


Transcatheter Transplant Ablation (Embolization With Absolute Alcohol)

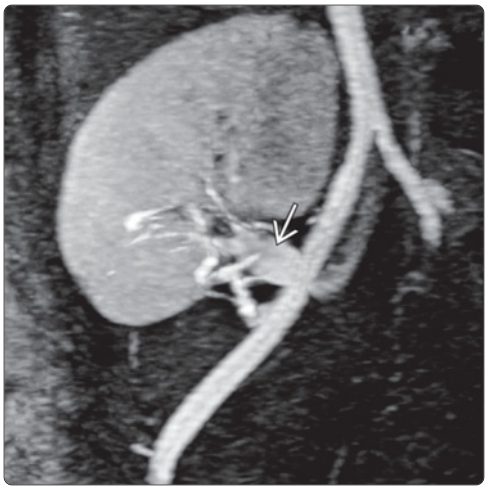


(Left) *DSA in a case of acute transplant rejection and severe hematuria shows a catheter in the left external iliac artery. The transplant renal artery and intrarenal branches are patent, and no mass is seen.* **(Right)** *Transcatheter transplant ablation was performed before nephrectomy to control bleeding. (A) After placing a balloon occlusion catheter in the renal artery, contrast was injected, noting the volume used. (B) DSA after injecting a similar volume of alcohol shows nonfilling of branch vessels, ablation.*

Acquired Arteriovenous Fistula (Nonfocal Renal Biopsy)

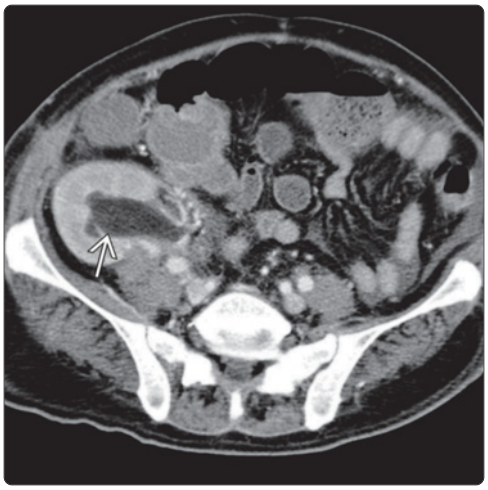


Acquired Arteriovenous Fistula (MR Angiography Evaluation)

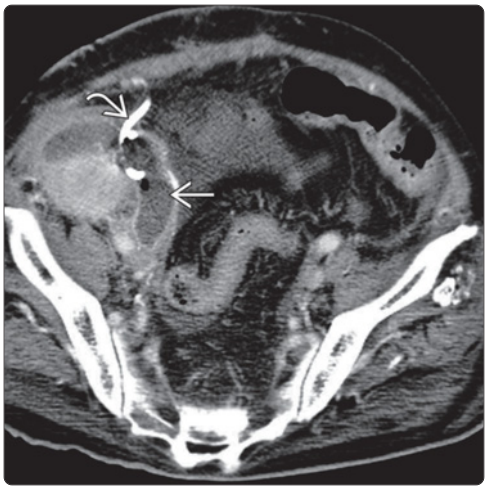


(Left) *Sagittal US obtained during biopsy of a right lower quadrant transplant shows the needle of a biopsy device within the lower pole renal cortex. Note the needle trajectory is oriented away from the hilum and is parallel to the cortex.* **(Right)** *Arterial phase MRA shows premature filling of the renal vein due to an arteriovenous fistula. Both pseudoaneurysms and arteriovenous fistulas can occur as a result of a renal biopsy that lacerates branches of the artery and vein within the transplant kidney.*

Perigraft Abscess Drainage (Associated Hydronephrosis on CECT)

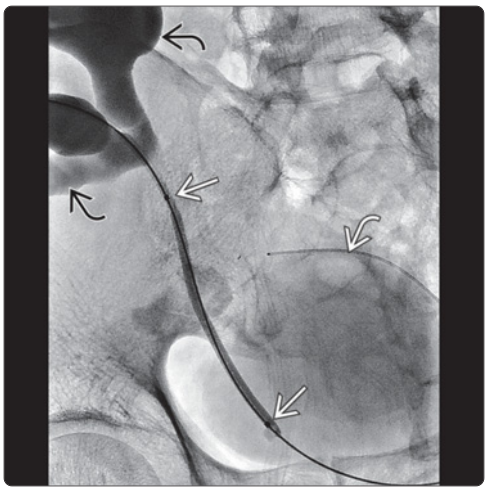


Perigraft Abscess Drainage (Drainage Catheter in Abscess)

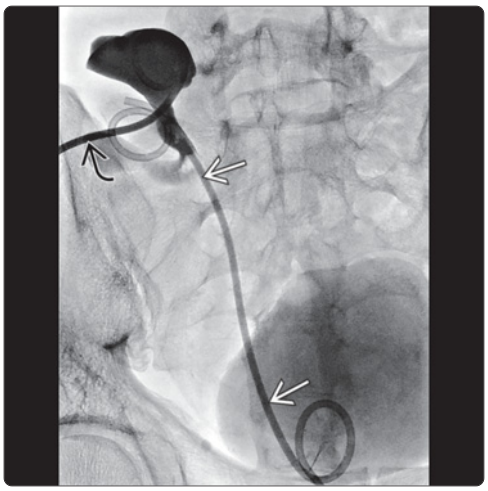


(Left) *Axial CECT obtained in a patient with a perigraft abscess demonstrates hydronephrosis caused by compression of the ureter by a peritransplant fluid collection.* **(Right)** *A more caudal axial CECT shows a percutaneously placed drainage catheter within a medial perinephric fluid collection. Loculation and gas suggest an abscess. Peritransplant fluid collections may come to clinical attention because of ureteral compression and resultant urinary obstruction.*

Ureteral Stricture Causing Obstruction (Ureteral Balloon Dilatation)

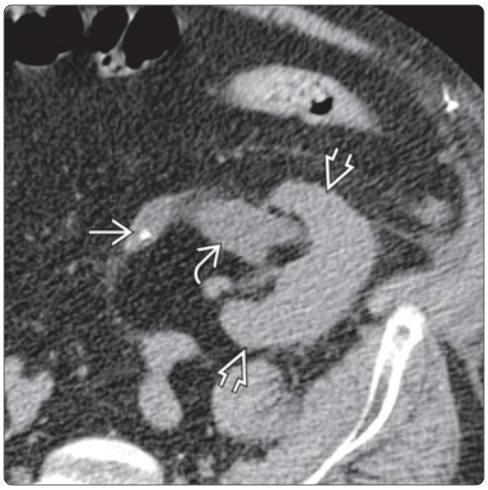


Ureteral Stricture Causing Obstruction (Antegrade Ureteral Stent Placement)

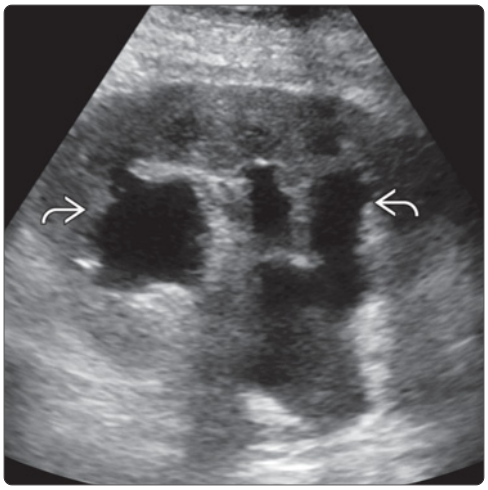


(Left) *Fluoroscopic spot radiograph obtained after percutaneous access to the transplant renal collecting system and contrast injection shows marked hydronephrosis due to a long ureteral stricture. A balloon was introduced over a guidewire and was inflated to dilate the stricture. Ureteral ischemia is the main cause of these strictures.* **(Right)** *Following balloon dilatation, a double pigtail ureteral stent was introduced and was placed across the stricture. Note the safety nephrostomy catheter.*

Renal Transplant Urinary Obstruction (Initial NECT Imaging Evaluation)

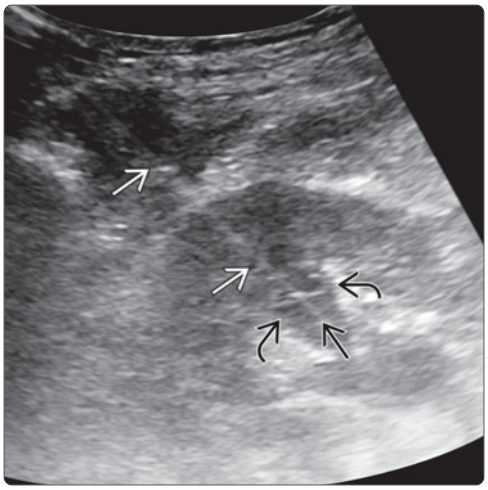


Renal Transplant Urinary Obstruction (Initial Ultrasound Evaluation)

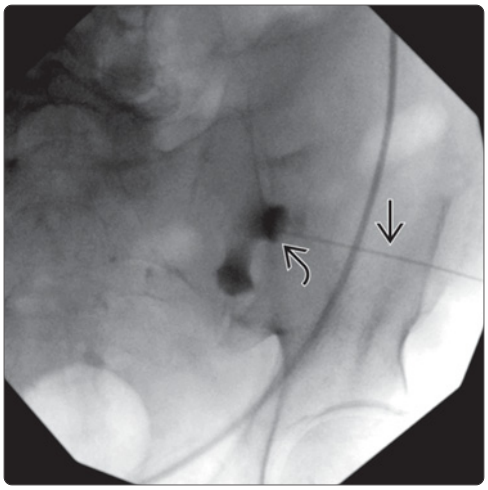


(Left) *Axial NECT of the left hemipelvis demonstrates an obstructing calculus in the proximal ureter of a renal transplant. Dilatation of the renal pelvis is evident, indicating hydronephrosis. Ureteral stenosis/ischemia is a more common cause of obstruction than calculus.* **(Right)** *Sagittal US prior to nephrostomy shows marked hydronephrosis in the transplant kidney. After selecting a lateral calyx for access, local anesthetic is administered using 1% lidocaine and a dermatotomy is made with a #11 scalpel.*

Renal Transplant Urinary Obstruction (US Guidance During Nephrostomy)

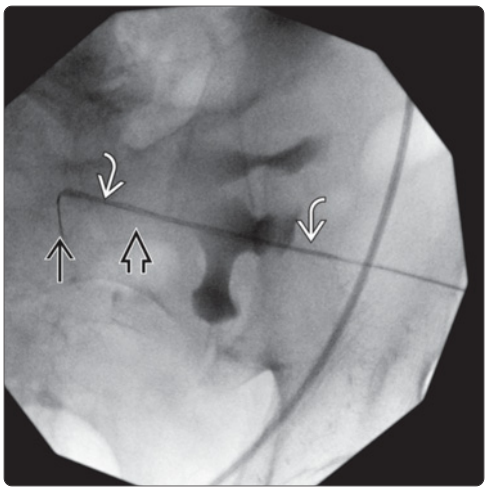


Renal Transplant Urinary Obstruction (Fluoroscopy During Nephrostomy)

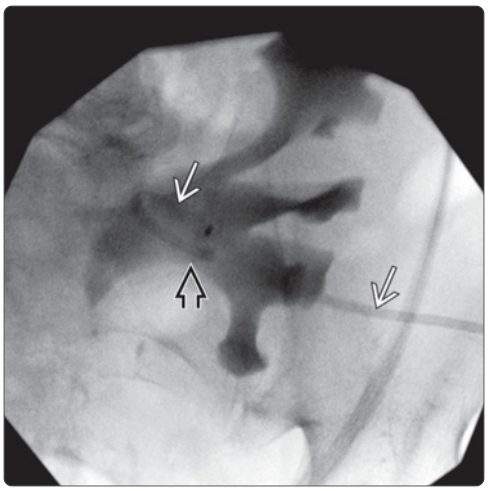


(Left) *Ultrasound obtained during primary access for percutaneous nephrostomy catheter placement demonstrates a 21-gauge Chiba needle with the tip within a lateral calyx of the transplant kidney.* **(Right)** *Fluoroscopic spot image obtained during nephrostomy after contrast injection through the Chiba needle confirms that the tip is in a lateral calyx. Nephrostomy catheter placement now will proceed using the 1-stick technique. The injected contrast volume is limited to reduce sepsis risk.*

Renal Transplant Urinary Obstruction (One-Stick Access Technique)

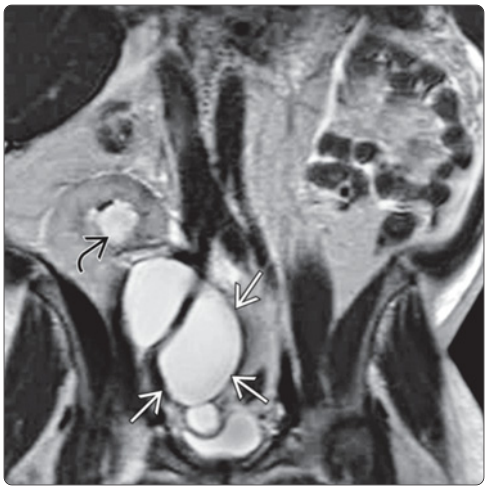


Renal Transplant Urinary Obstruction (Completion Nephrostogram)

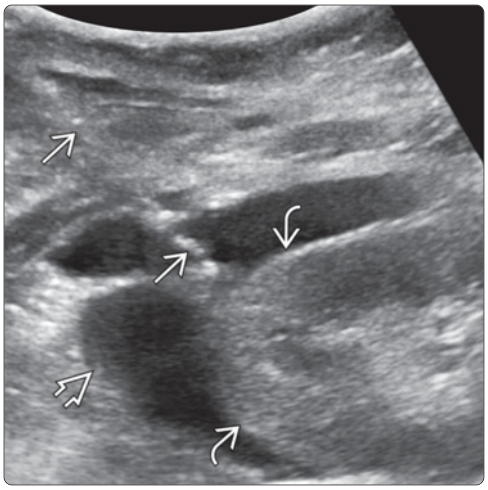


(Left) *A 0.018" guidewire is advanced through the needle using fluoroscopic guidance. The needle is exchanged for a 6 Fr coaxial dilator-sheath assembly, and the sheath is advanced over the guidewire into the renal pelvis. The dilator is removed, the microwire exchanged for a 0.035" guidewire, and the tract dilated to 8 French.* **(Right)** *Over the 0.035" guidewire, an 8 Fr pigtail nephrostomy catheter is advanced into the collecting system. Contrast injection confirms location within the renal pelvis.*

Paratransplant Lymphocele (Initial MR Evaluation)

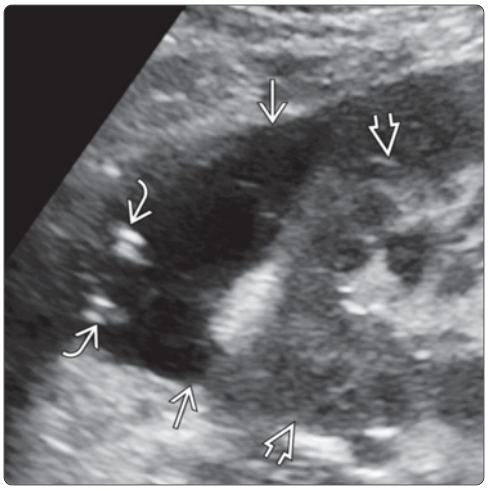


Paratransplant Lymphocele (Ultrasound-Guided Drainage)

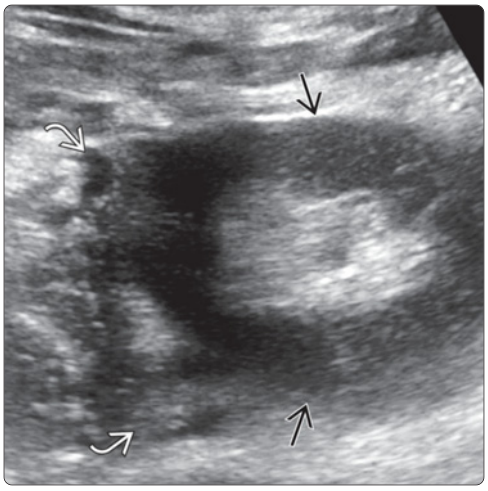


(Left) *Coronal T2WI MR shows transplant hydronephrosis, caused by extrinsic ureteral compression by a surrounding lymphocele. The lymphocele has a typical appearance with multiple septations.* **(Right)** *US shows an anechoic perinephric collection. An 8 Fr locking pigtail catheter loaded on a trocar has been advanced into the collection. Once the tip of the assembly is in the collection, the hub is unscrewed, the inner trocar is held still, and the catheter is advanced further into the collection.*

Paratransplant Lymphocele (Persistence After Drainage)

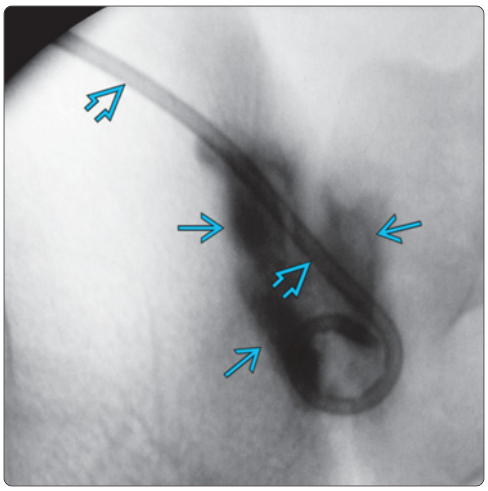


Paratransplant Lymphocele (Post-Drainage Ultrasound)

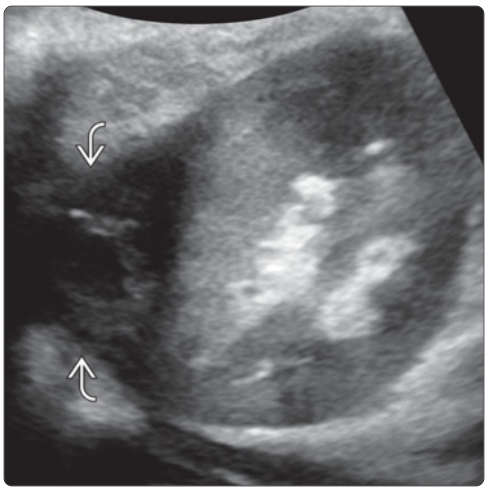


(Left) *Drainage persisted 1 week post catheter placement. US shows the pigtail loop within the persistent collection adjacent to the kidney. Lymphoceles are frequently refractory to simple drainage and sclerotherapy may ultimately be necessary.* **(Right)** *Postprocedural ultrasound shows significant decompression of the collection that previously surrounded the inferior aspect of the kidney. Only a minimal residual collection remains. A drainage catheter was left indwelling.*

Paratransplant Lymphocele (Lymphocele Sclerotherapy)



Paratransplant Lymphocele (Lymphocele Sclerotherapy)



(Left) *In preparation for lymphocele sclerosis, contrast was injected via the drainage catheter, showing a persistent cavity but no fistula to the collecting system. Sclerotherapy was subsequently performed using a fibrin sealant. 15 mL of sealant were injected into the lymphocele through the catheter.* **(Right)** *Ultrasound performed 1 week post sclerotherapy shows interval collapse of the collection. The patient was pain free, there was no reaccumulation of lymphatic fluid, and the hydronephrosis resolved.*

Venous Sampling and Venography (Renal and Adrenal)

KEY FACTS

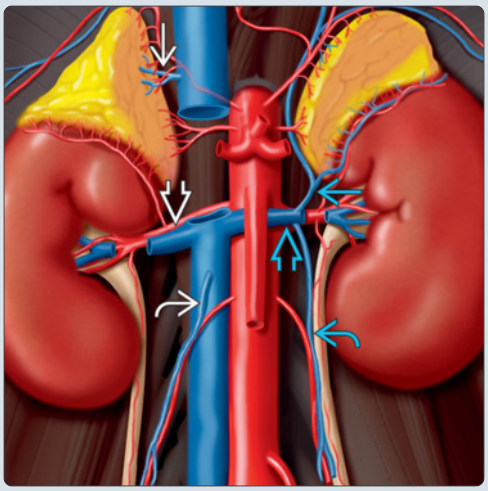
PREPROCEDURE

- **Indications**
  - **Adrenal venous sampling**
    - Differentiating etiology of primary aldosteronism
    - Used to confirm adrenal gland as source of excess cortisol in patient with Cushing syndrome and adrenal mass on imaging
    - Adrenal venous sampling for pheochromocytomas and paragangliomas
  - **Renal vein sampling**
    - Determining if renal artery stenosis is the etiology of hypertension (renovascular hypertension)
  - **Renal venography for diagnosis &/or treatment**
    - "Nutcracker" syndrome
      - Pressure gradient > 2 mmHg between renal vein and IVC confirms diagnosis
    - Pelvic congestion syndrome
    - Varicocele

PROCEDURE

- **Adrenal vein sampling**
  - Selective catheterization of each adrenal vein
  - Right adrenal vein more difficult to catheterize than left; should be catheterized/sampled 1st
  - Obtain samples from each adrenal vein before and after adrenocorticotrophic hormone (ACTH) administration
  - High aldosterone:cortisol ratio before and after ACTH, lateralizing to 1 side is confirmatory of adrenal adenoma
  - Elevated cortisol lateralizing to adrenal vein in presence of adrenal mass on CT/MR is confirmatory of endogenous Cushing syndrome
- **Renal venography and renin sampling**
  - Discontinue antihypertensive β blockers and ACE inhibitors several days prior to sampling
  - Captopril stimulation (1 mg/kg body weight) 60-90 minutes before sampling increases accuracy

(Left) *Graphic shows that the right adrenal, renal, and gonadal veins drain directly into the IVC; the left adrenal and gonadal veins drain into the left renal vein, which courses anterior to the aorta.* **(Right)** *Axial CECT shows bilateral adrenal masses in a patient with hypokalemia and diastolic hypertension, suspected of primary aldosteronism. Bilateral adrenal vein sampling was performed for further evaluation.*

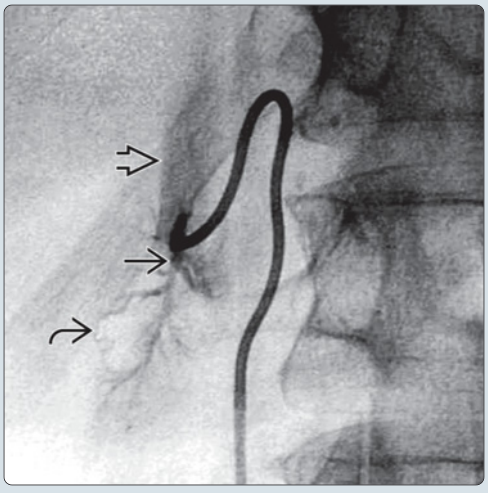


Adrenal Mass

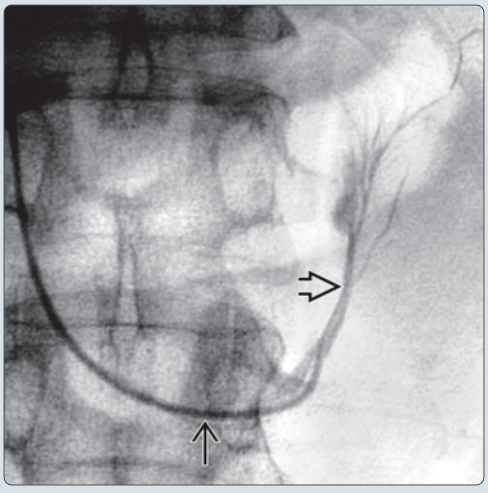


(Left) *Right adrenal venogram shows a short main adrenal vein trunk draining to the IVC. Note filling of branches within the adrenal gland.* **(Right)** *Left adrenal venogram shows the catheter entering the left renal vein, through which a microcatheter has been coaxially introduced and positioned in the left adrenal vein. After confirming a correct catheter position with gentle contrast injection, venous sampling is performed following cortisol stimulation.*

Adrenal Vein Sampling



Adrenal Vein Sampling



TERMINOLOGY

Synonyms

- Renal vein sampling (RVS), renal vein renin sampling (RVRS)

Definitions

- **Primary aldosteronism** (a.k.a. primary hyperaldosteronism): Hypersecretion of aldosterone by adrenal glands
  - Adrenal adenoma (**Conn syndrome**): Cause in 2/3 of cases
  - Bilateral idiopathic adrenal hyperplasia: Cause in remaining 1/3 of cases
- **Cushing syndrome**: Clinical complex resulting from prolonged inappropriate exposure to glucocorticoids
  - Most frequently caused by administration of exogenous glucocorticoids or adrenocorticotrophic hormone (ACTH)
  - Endogenous causes include ACTH-secreting pituitary tumor, adrenal neoplasm (benign or malignant), or ectopic ACTH secretion by tumor

Adrenal Vein Anatomy

- In most individuals, each gland has single main vein
  - Multiple adrenal veins occasionally occur
  - Adrenal veins communicate with retroperitoneal and renal capsular veins
- Right adrenal vein drains directly into right posterolateral aspect of IVC
  - Typically located 2-4 cm above right renal vein
  - Small accessory hepatic vein may rarely drain into right adrenal vein or vice versa (10% incidence)
- Left adrenal vein drains into superior aspect of left renal vein
  - Typically located 3-5 cm from left renal vein orifice
  - May very rarely drain directly into IVC

Renal Vein Anatomy

- Usually each kidney is drained by single renal vein
  - Renal veins rarely have valves
  - Communicate with other retroperitoneal veins (e.g., lumbar, azygos, gonadal veins)
- Left renal vein courses anterior to aorta and posterior to SMA to join IVC
  - Left renal vein drains somewhat anteriorly into IVC
- Right renal vein is shorter, drains into IVC at ~ L2 vertebral level
- Frequent variations in renal venous anatomy

PREPROCEDURE

Indications

- **Adrenal vein sampling (AVS)**
  - Differentiating etiology of primary aldosteronism
    - AVS is gold standard for distinguishing adenomas from bilateral adrenal hyperplasia
      - Aldosterone-producing adenoma results in lateralization of aldosterone hypersecretion
    - Has impact on clinical management: Bilateral adrenal hyperplasia is treated medically; aldosterone-producing adenoma is treated surgically (adrenalectomy)
  - Used to confirm adrenal gland as source of excess cortisol in patient with Cushing syndrome and adrenal mass on imaging
    - AVS is useful to localize independently functioning adrenal mass
    - Resection of functioning unilateral adrenal mass is curative
  - AVS for pheochromocytomas and paragangliomas
    - With advances in MR and CT, adrenal vein sampling is infrequently needed
    - Used in localizing occult pheochromocytoma when imaging is equivocal or negative
    - Detection of bilateral tumors in familial syndromes, such as von Hippel-Lindau, when imaging shows only unilateral disease
- **RVS**
  - Determining if renal artery stenosis is etiology of hypertension (renovascular hypertension)
    - RVRS in evaluation/treatment of renovascular hypertension has decreased, now infrequently performed
- **Renal venography for diagnosis &/or treatment**
  - "Nutcracker" syndrome
    - Left renal vein compression between SMA and aorta
      - Results in left renal venous hypertension; may produce chronic pain, hematuria, and proteinuria
      - May result in varicoceles or female pelvic congestion syndrome
    - Pressure gradient > 2 mmHg between renal vein and IVC confirms diagnosis
    - Treatment with stent has shown varying degrees of success
  - Pelvic congestion syndrome
    - Left ovarian vein drains directly into left renal vein
    - Left renal venography may demonstrate reflux into dilated ovarian vein, indicating venous valvular incompetency
  - Varicocele
    - Left internal spermatic (gonadal) vein drains directly into left renal vein
    - Left renal venogram may show reflux into gonadal vein; indicating venous valvular incompetency

Getting Started

- Things to check
  - **RVRS**
    - Discontinue antihypertensive β blockers and ACE inhibitors several days prior to sampling
- Medications
  - **AVS**: Cosyntropin (Cortrosyn) for stimulation; synthetic subunit of ACTH
  - **RVRS**: Captopril (1 mg/kg body weight) 60-90 minutes before sampling
- Angiographic sheaths and catheters
  - Vascular sheath (5-7 Fr) for femoral vein access
  - Angiographic catheters
    - Selective catheters should have sidehole near tip
    - Simmons, SOS, or Mikaelsson catheters are typical choices for right adrenal vein
    - Cobra, Simmons catheters typical choices for left adrenal vein and for either renal vein

PROCEDURE

Patient Position/Location

- For both renal and adrenal venography/sampling
  - Introduction of selective catheter from common femoral vein access sheath

Procedure Steps

- **Adrenal venography and sampling**
  - Obtain venous access from common femoral vein puncture; introduce 6 or 7 Fr vascular sheath
  - Place selective catheter in IVC; sequentially cannulate/sample right followed by left adrenal vein
    - Unnecessary to simultaneously sample veins
      - Use reverse-curve catheter to selectively catheterize right adrenal vein
      - Use Cobra/Simmons catheter to cannulate left adrenal vein; may require coaxial microcatheter
  - Confirm catheter position; gentle contrast injection
    - Patient may experience back pain during contrast injection into adrenal vein, particularly on right
  - Venous sampling from each adrenal vein and peripheral source (e.g., femoral vein)
    - Obtain sample from adrenal vein 15 minutes after administration of cosyntropin (0.25 mg bolus followed by infusion of 0.15-0.20 mg/hr)
  - Label all tubes, with attention to right vs. left
  - Submit samples for aldosterone and cortisol assays
    - Appropriate cortisol level confirms correct catheter position within adrenal vein
- **RVRS**
  - Captopril stimulation (1 mg/kg body weight) 60-90 minutes before sampling increases accuracy
  - Obtain venous access from common femoral vein puncture; introduce 5 Fr vascular sheath
  - Catheterize right/left main renal vein with 5 Fr Cobra or Simmons catheter (sidehole at tip)
    - On left, catheter should be beyond gonadal vein
  - Be certain there are no accessory/aberrant veins that also require venous sampling
  - Obtain 5-10 mL blood sample from each renal vein
    - Sample quickly; renin levels vary with time
  - Obtain samples from IVC above and below renal veins

Findings and Reporting

- **AVS for primary aldosteronism**
  - Appropriate cortisol level confirms correct catheter position within adrenal vein
    - Cortisol production should be same for both adrenal glands
  - Sampling may distinguish between functioning adrenal adenoma and idiopathic adrenal hyperplasia
    - Adrenal adenoma has high aldosterone:cortisol ratio before and after ACTH; lateralizes to affected side
    - Lateralization generally considered > 4:1 ratio difference between glands
    - In bilateral hyperplasia, no lateralization of ratios before or after ACTH occurs, but aldosterone:cortisol ratio is higher than in IVC
- **AVS for Cushing syndrome**
  - Obtain samples from both adrenal veins, suprarenal and infrarenal IVC with no cortisol stimulation
    - Elevated cortisol lateralizing to 1 adrenal vein in presence of adrenal mass on CT/MR is confirmatory
- **RVRS**
  - Renin levels from 1 kidney that are > 1.5x that of contralateral kidney is positive result
  - Rise in renin from infrarenal to suprarenal IVC is further evidence for renovascular hypertension

POST PROCEDURE

Expected Outcome

- AVS is most accurate means of diagnosing cause of hyperaldosteronism

Things to Do

- When obtaining venous samples, note time obtained and label all tubes with attention to right vs. left
- Collect samples in appropriate containers for laboratory analysis; conform to lab requirements for accurate sample analysis (e.g., refrigerated sample)

Things to Avoid

- Adrenal veins are small, weak, and prone to rupture; avoid forceful contrast injections
- Usually tenuous selective catheter positions in adrenal veins; avoid excessive catheter movement when obtaining samples to prevent dislodgement

OUTCOMES

Problems

- Most common difficulty is failure to cannulate right adrenal vein due to challenging anatomy
- Selective catheterization of hepatic caudate lobe venous drainage often mistaken for right adrenal vein
- Adrenal vein characteristics
  - Triangle-/delta-shaped angiographic blush
  - Central vein communicating with smaller veins in stellate/spiculated pattern
  - Communicating retroperitoneal collaterals

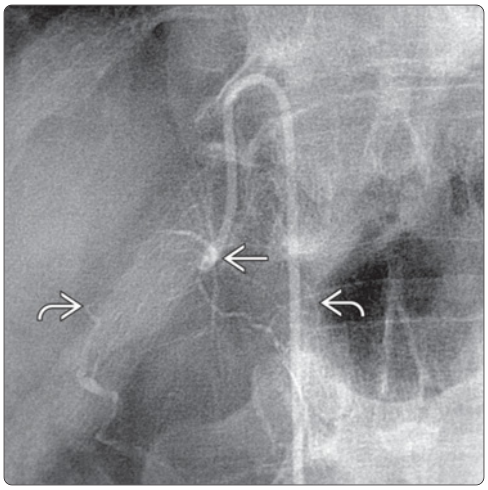
Complications

- Most feared complication(s)
  - Adrenal vein dissection/rupture (0.5%), adrenal hemorrhage or infarction (0.5%)
- Other complications
  - Contrast reaction or contrast-induced nephropathy
  - Access site hematoma, venous thrombosis, infection

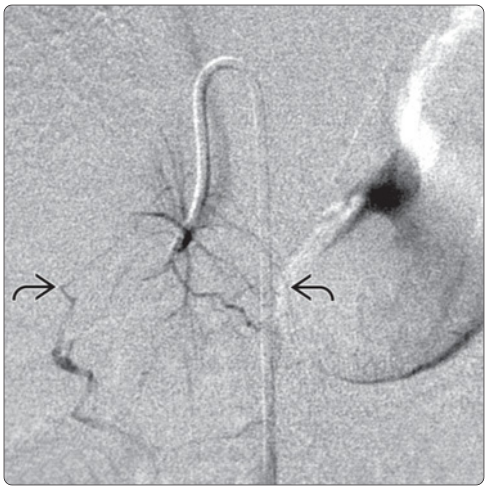
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Adrenal Vein Sampling

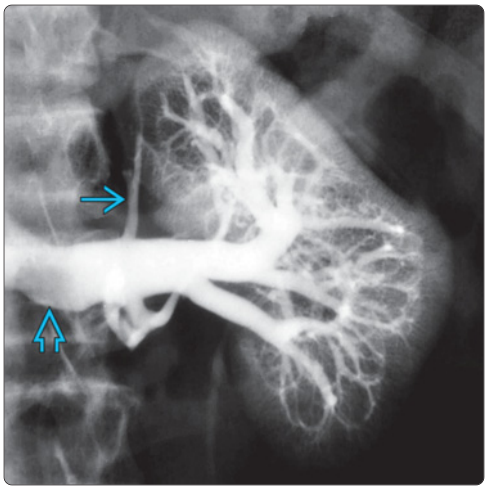


Adrenal Vein Sampling

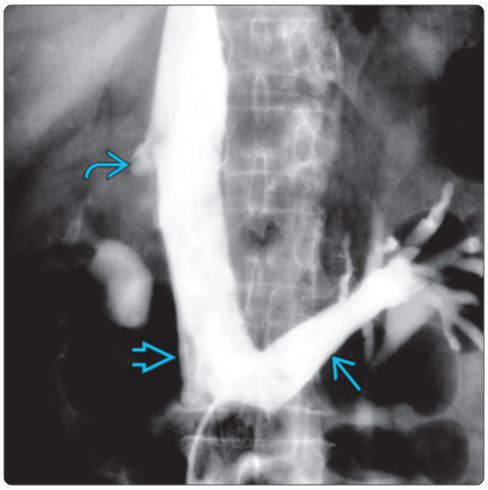


(Left) *Frontal projection from right adrenal venography shows selective injection of the main trunk of the right adrenal vein with a Simmons-1 catheter. The glandular veins are draped around a mass, which was seen on cross-sectional imaging in this patient with Cushing disease. Sampling confirmed this was the source of excess cortisol.* **(Right)** *Digital subtraction angiography (DSA) image better depicts draping of adrenal veins around the adrenal mass.*

Normal Left Renal Venogram

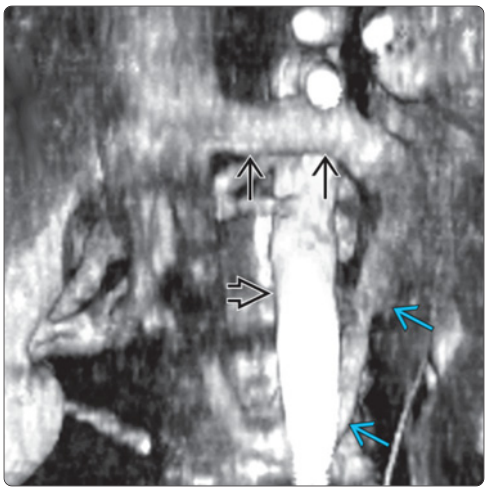


Retroaortic Left Renal Venogram

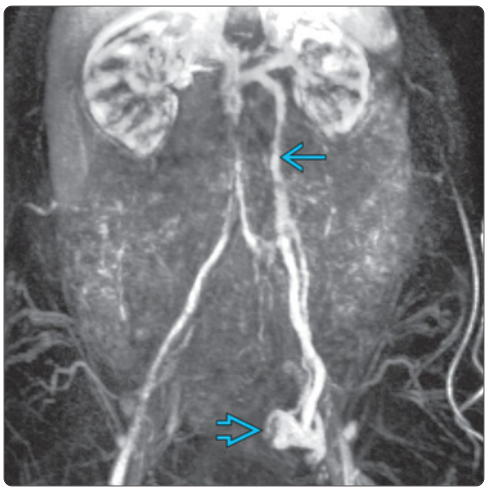


(Left) *A normal left renal venogram shows multiple intrarenal branches draining into the main left renal vein. The left adrenal vein drains directly into the left renal vein.* **(Right)** *Venogram shows a nonconventional course of the left renal vein as it joins the IVC. This appearance is typical of a retroaortic left renal vein, which is a well-known anatomic variant. A small portion of the right renal vein is seen entering the IVC at the expected location.*

Circumaortic Left Renal Vein CTV



Nutcracker Syndrome



(Left) *3D CTA/CTV of a circumaortic left renal vein shows a normal course of the superior renal vein anterior to the aorta while the inferior vein has a retroaortic course. This anatomy has implications for IVC filter placement.* **(Right)** *Coronal MIP from MRV of suspected "nutcracker" syndrome shows a dilated left gonadal vein and pelvic varicosities. Compression of the left renal vein between the aorta and SMA causes renal venous hypertension and gonadal vein reflux.*

KEY FACTS

TERMINOLOGY

- **Hysterosalpingography**: Contrast injection into endometrial cavity with fluoroscopy/imaging
- **Selective salpingography**: Direct selective catheterization of fallopian tubal ostium
  - Contrast injection via catheter during imaging
- **Fallopian tube recanalization**: Guidewire/catheter passage through occluded FT to reestablish patency
- **Fallopian tube occlusion**: Placement of mechanical occluding device into fallopian tubes

PREPROCEDURE

- Selective salpingography indications
  - Differentiate spasm from true obstruction
  - Inadequate fallopian tube opacification by HSG
- Fallopian tube recanalization indications
  - Tubal occlusion despite selective salpingography
- Fallopian tube occlusion indications
  - Prophylaxis against unwanted pregnancy

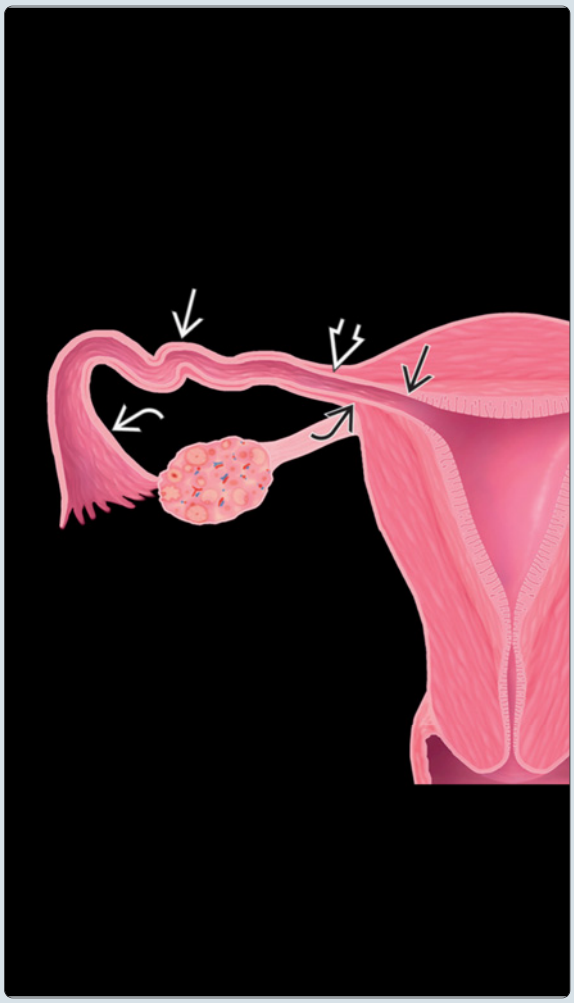
PROCEDURE

- **Selective salpingography**
  - Advance 4-5-French catheter over 0.035" guidewire; wedge tip in uterine cornual region
  - Gently inject contrast medium; obtain images
- **Fallopian tube recanalization**
  - Advance catheter/guidewire through obstruction
- **Fallopian tube occlusion**
  - Place device in most challenging side 1st
  - Engage 5-French catheter working port in tubal ostium
  - Advance device until 3rd radiopaque marker at tubal ostium; deploy Essure coil

POST PROCEDURE

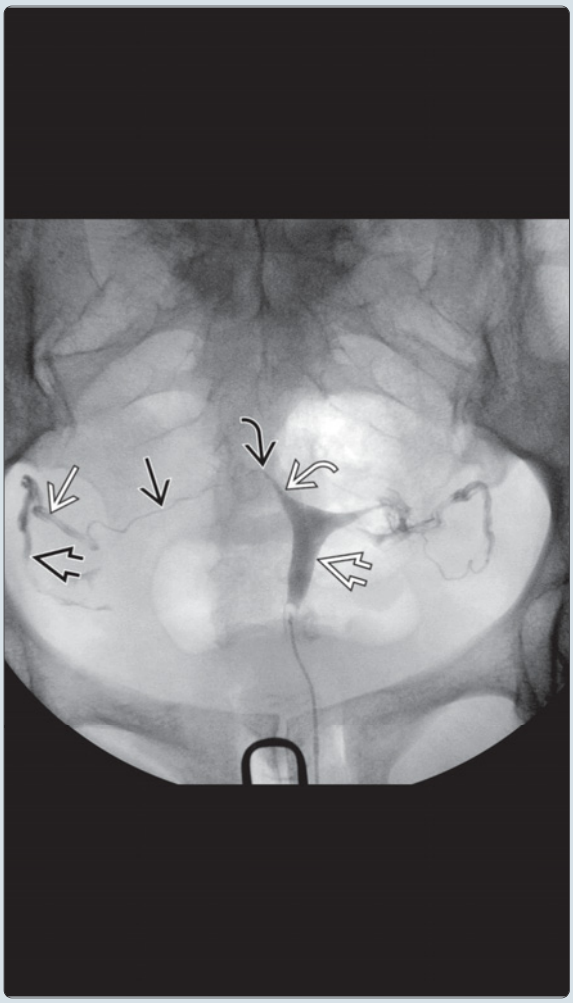
- Fallopian tube recanalization
  - Procedural success: 71-92%
- Fallopian tube occlusion
  - Efficacy rate of Essure preventing pregnancy: 99.8%

Fallopian Tube Anatomy



Graphic shows a normal fallopian tube. From the ostium ⇨, the interstitial segment ⇨ transitions to the isthmic segment at the uterotubal junction ⇨. The remaining segments are termed ampullary ⇨ and infundibular ⇨.

Normal Hysterosalpingogram



Hysterosalpingogram shows a normal endometrial cavity ⇨, the tubal ostium ⇨, uterotubal junction ⇨, and the isthmic ⇨, ampullary ⇨, and infundibular ⇨ segments. Both fallopian tubes are widely patent.

TERMINOLOGY

Definitions

- **Hysterosalpingography (HSG)**: Contrast injection via cervical canal into endometrial cavity with fluoroscopic monitoring/imaging
  - Normally shows filling of uterine cavity
  - Should also show bilateral fallopian tube (FT) filling
    - Tubal patency confirmed if contrast spills from tubes into abdominal cavity
  - Sensitivity and specificity in detecting pathology
    - Bilateral tubal pathology: 46% and 95%
    - Any fallopian tube pathology: 53% and 87%
- **Selective salpingography**: Direct selective catheterization of fallopian tubal ostium
  - Contrast injection via catheter during imaging
    - Provides direct opacification of fallopian tube
- **Fallopian tubal anatomy**
  - 4 tubal segments
    - Interstitial (intramural)
    - Isthmic
    - Ampullary
    - Infundibular
  - Uterotubal junction located at transition of intramural to isthmic segment of fallopian tube
    - Narrowest portion of fallopian tube
  - Average tubal diameter: 0.8-2.0 mm
    - Increases as fallopian tube courses toward ovaries
  - Average tubal length: 11 cm (range: 7-16 cm)
- **Fallopian tube recanalization (FTR)**:Guidewire &/or catheter passage through occluded fallopian tube to reestablish tubal patency
  - Fallopian tube disease accounts for up to 35% of infertility cases; various causes
    - Infection (chlamydia)
    - Postsurgical
    - Salpingitis isthmica nodosa
      - Inflammation of isthmic segment; characterized by nodular thickening causing obstruction
    - Peritubal adhesions
  - Proximal tubal occlusion often caused by mucus plug/inflammatory debris; also caused by spasm
    - Involves interstitial segment/uterotubal junction
    - Responds fairly well to recanalization
  - Less favorable outcomes with more distal occlusions
    - More often due to scarring/fibrosis
- **Fallopian tube occlusion (FTO)**: Placement of mechanical occluding device into fallopian tubes
  - Type of bilateral tubal sterilization
    - Hysteroscopic transcervical fallopian tube access
    - Placement of Essure device (Conceptus; Mountain View, CA) into fallopian tubes
      - Combined outer nitinol/inner stainless steel coil elicits tissue growth; occludes fallopian tubes
      - During first 3 months device not fully effective
      - Alternate birth control needed during this time

PREPROCEDURE

Indications

- **Selective salpingography**
  - Differentiate spasm from true tubal obstruction
  - Inadequate fallopian tube opacification by HSG
- **Fallopian tube recanalization**
  - Persistent tubal occlusion after selective salpingography
- **Fallopian tube occlusion**
  - Prophylaxis against unwanted pregnancy
  - Medical conditions aggravated by pregnancy

Contraindications

- **Selective salpingography**
  - Active infection
- **Fallopian tube recanalization**
  - Active infection
- **Fallopian tube occlusion**
  - Desire to maintain fertility
  - Pregnancy; recent or current infection
  - Essure device-related contraindications
    - Prior tubal ligation
    - Pregnancy termination/child delivery < 6 weeks before intended placement
    - Immunosuppression
    - Menorrhagia/metrorrhagia of unknown etiology
    - Allergy to nickel

Getting Started

- Things to check
  - Detailed clinical and physical evaluation
  - Allergies
  - Negative qualitative urinary β-hCG (day of procedure)
  - Selective salpingography: Review images/anatomy
  - Fallopian tube recanalization: Document fallopian tubal occlusion
  - Fallopian tube occlusion: Correct procedure timing
    - Perform in follicular phase (day 7-14) of menses
- Medications
  - Preprocedural ketorolac (30 mg)
  - Antibiotic prophylaxis
  - Intraprocedural conscious sedation
  - Paracervical block anesthesia
- Equipment list
  - Speculum
  - Transcervical access catheter/device for hysterosalpingography (various options)
    - Thurmond-Rösch Hysterocath (Cook Medical; Bloomington, IN)
    - 9-French sheath in endometrial cavity
    - 12-French transcervical catheter with balloon
  - Essure device
    - 4.3-French outer diameter delivery system
    - Placed via 5-French catheter working port
  - Catheters for selective FT catheterization
    - 4-French angled-tip catheter
    - 5-French Kumpe catheter
  - Catheter for fallopian tube recanalization
    - Thurmond-Rösch fallopian tube catheterization set (Cook Medical; Bloomington, IN)

PROCEDURE

Patient Position/Location

- Patient in lithotomy position

Equipment Preparation

- Dilute contrast medium to 30%

Procedure Steps

- **Cervical cannulation**
  - Sterilely prepare/drape vulvar and perineal area
  - Introduce speculum; visualize cervix
  - Place 9-12-French occlusive catheter and fix it with tenaculum, external cervical suction, or endocervical/uterine balloon
  - Place 5-French diagnostic catheter: Inject contrast to opacify uterine cavity
- **Selective salpingography**
  - Advance 4-5-French catheter over 0.035" guidewire
  - Gently inject contrast medium
  - Perform contralateral fallopian tube catheterization
- **Fallopian tube recanalization**
  - 3-French tapered catheter preloaded over 0.014-0.018" guidewire
  - Advance catheter/guidewire through obstruction
  - Persistent obstruction
    - Cross with 0.014-0.018" diameter floppy-tip/0.035" hydrophilic guidewire
    - Use firm traction on cervix to straighten uterus
- **Fallopian tube occlusion**
  - Obtain HSG images of both fallopian tubes
  - Place device in most challenging side 1st; should abort procedure if bilateral placement not possible
  - Engage 5 French working port in tubal ostium
  - Advance device through working port into fallopian tube until 3rd radiopaque marker at tubal ostium
  - Deploy Essure coil
  - Perform contralateral device placement

Findings and Reporting

- **Selective salpingography**
  - Selective tubal catheterization: Technically successful in up to 90% of cases
  - Opacification of normal tube in 20-30% of cases with peritoneal passage of contrast
- **Fallopian tube recanalization**
  - Improved tubal diagnosis in 90% of cases
  - Restoration of tubal patency in 40% of cases
- **Fallopian tube occlusion**
  - HSG criteria for grading coil placement and tubal occlusion (3 months follow-up after FTO)
    - Grade I
      - Expulsion of coil or > 50% of coil inner length trails into uterine cavity
      - Tubal occlusion at cornua
    - Grade II
      - < 50% of inner coil length trails into uterine cavity or proximal end of inner coil is < 30 mm into tube from tubal ostium
      - Contrast in tube, not past any portion of coil
    - Grade III
      - Coil inner length proximal end > 30 mm distal to tubal ostium or coil within peritoneal cavity
      - Contrast tracks past coil or into peritoneal cavity
    - Coil placement must be grade II and tubal occlusion must be grade I or II for sterility

POST PROCEDURE

Expected Outcome

- Fallopian tube recanalization
  - Procedural success: 71-92%
  - Pregnancy typically occurs within 6 months of FTR
- Fallopian tube occlusion
  - Efficacy rate of Essure preventing pregnancy: 99.8%
  - Vaginal spotting, pelvic cramping expected afterward

Post Procedure Imaging

- Fallopian tube recanalization
  - Repeat HSG in patients who fail to achieve pregnancy
- Fallopian tube occlusion
  - Follow-up in 3 months with hysterosalpingogram

OUTCOMES

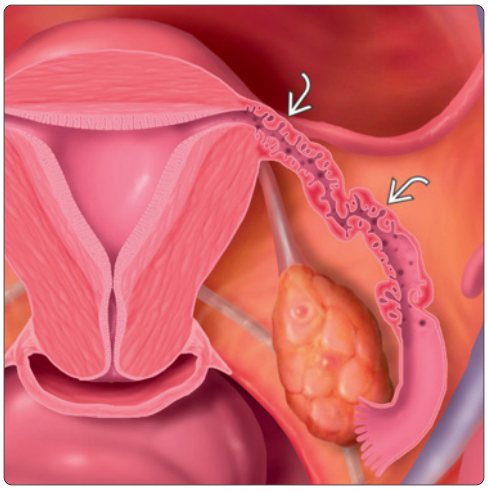
Complications

- Immediate/periprocedural complication(s)
  - Tubal perforation: 2% of cases
    - Often actually submucosal guidewire passage
  - Contrast intravasation
    - Can occur if catheter poorly positioned
  - Infection (extremely rare)
    - Peritonitis: Incidence < 1%
    - Pyosalpinx, endometritis
- Delayed complication(s)
  - Extrauterine (ectopic) pregnancy
    - Tubal pregnancies reported in up to 3% of women after FTR
    - Higher risk after FTR but less than after microsurgical tubal intervention
- Other complications
  - Radiation exposure
    - Ovarian exposure ~ 1 rad (10 mGy)
    - Perform procedures in follicular phase of ovulatory cycle to ensure patient not pregnant

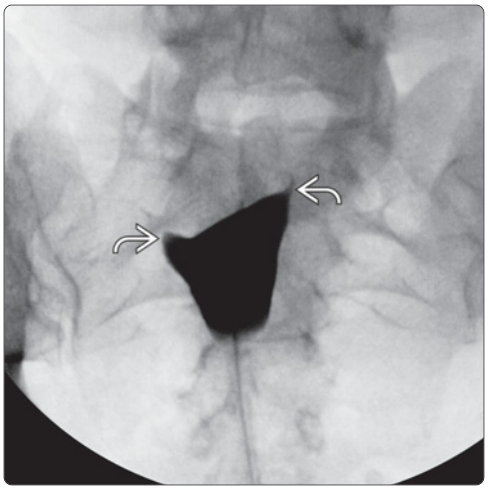
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Fallopian Tube Recanalization (Tubal Occlusion From Salpingitis)

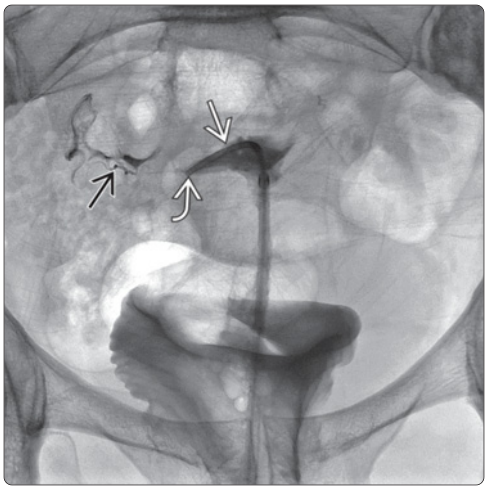


Fallopian Tube Recanalization (Initial Hysterosalpingogram)

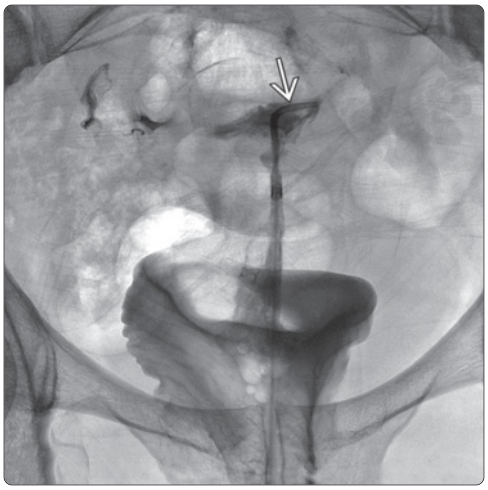


(Left) *Fallopian tube disease accounts for up to 35% of infertility cases with various causes for tubal occlusion. This graphic shows a diseased left fallopian tube due to salpingitis, which is often due to chlamydia.* **(Right)** *Initial evaluation is via hysterosalpingography (HSG). After accessing the cervix and endometrial canal, contrast medium is injected and images are obtained. In this case, there is bilateral fallopian tube occlusion at the ostia.*

Fallopian Tube Recanalization (Selective Right Salpingography)

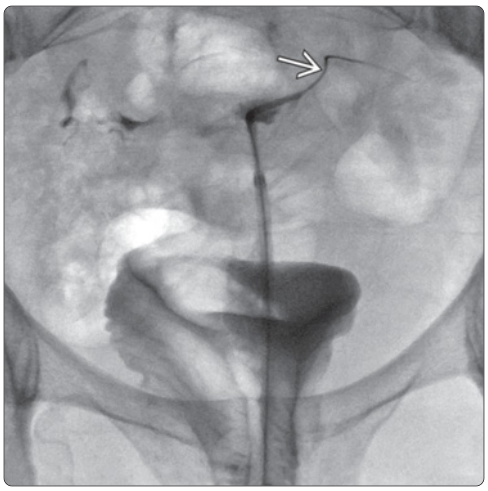


Fallopian Tube Recanalization (Selective Left Salpingography)

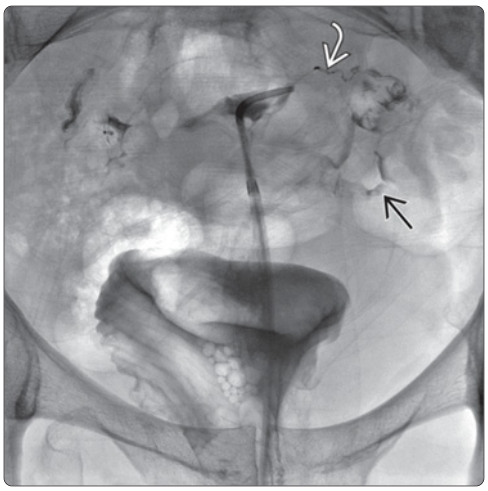


(Left) *The tip of an angled catheter has been placed at the right tubal ostium and selective salpingography performed showing a patent fallopian tube. It is not uncommon that proximal occlusion, as seen on a prior HSG, is no longer present (reported in up to 40% of patients).* **(Right)** *The angled catheter has been placed in the left cornu where contrast injection fails to fill the fallopian tube, indicating tubal occlusion.*

Fallopian Tube Recanalization (Guidewire Passage on Left)

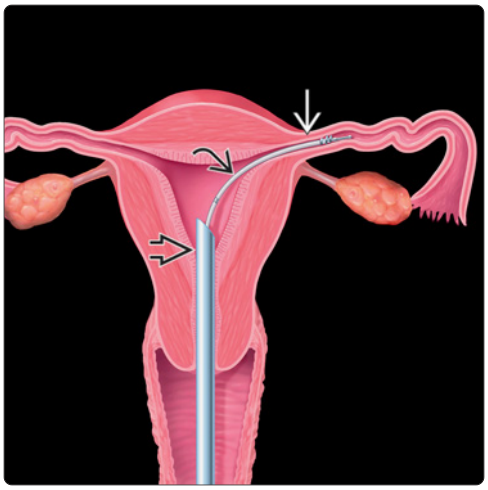


Fallopian Tube Recanalization (Final Hysterosalpingography)

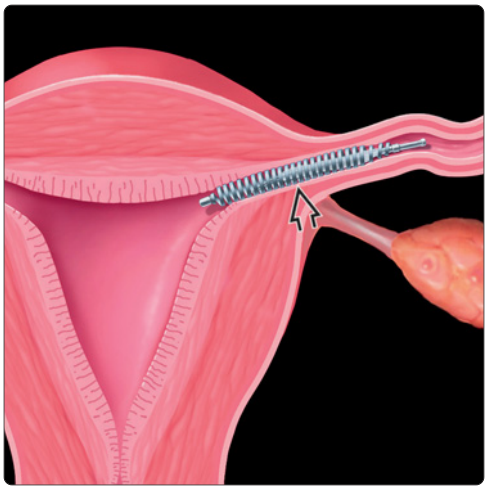


(Left) *After cannulating the fallopian tube orifice, a 0.018" guidewire has been advanced into the interstitial tubal segment in an attempt to dislodge any occluding debris. More distal occlusions are far less responsive to recanalization.* **(Right)** *Left salpingogram after guidewire passage shows the fallopian tube is patent, with free contrast spillage into the peritoneal cavity. There is a high technical success rate for recanalization, but conception rates are unfortunately much lower and are generally reported at around 30%.*

Fallopian Tube Occlusion (Cannulation of Tubal Ostium)

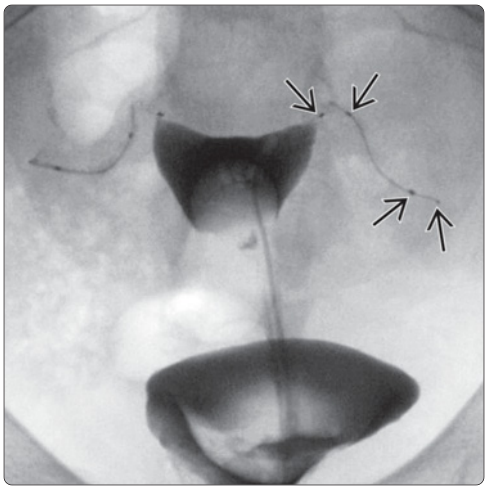


Fallopian Tube Occlusion (Essure Device Placement)

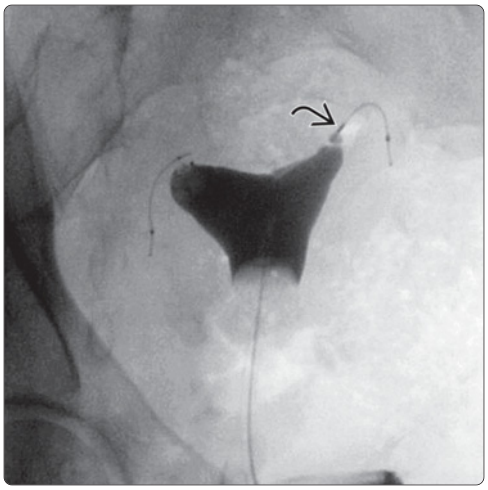


(Left) *Graphic shows a transcervical cannula in place through which the 4.3-French diameter delivery system for the Essure device has been introduced. The left fallopian tube ostium has been selectively cannulated, and the device has been advanced so that it will span the uterotubal junction once it is deployed.* **(Right)** *Graphic shows the deployed Essure device in place. The combined outer nitinol and inner stainless steel coils elicit tissue ingrowth, which will occlude the fallopian tubes over a period of 3 months.*

Fallopian Tube Occlusion (Successful Tubal Occlusion)



Fallopian Tube Occlusion (Successful Tubal Occlusion)

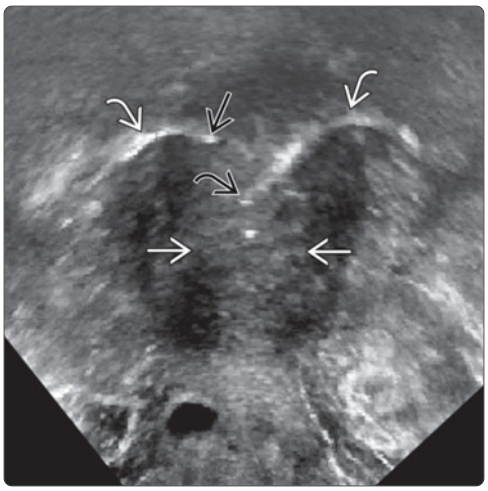


(Left) *Partial fill view of the uterine cavity from a hysterosalpingogram obtained 3 months after bilateral Essure placement shows both devices are in satisfactory position. Each device has a series of 4 radiopaque markers for determining proper positioning.* **(Right)** *Magnified oblique view of the left uterine cornu shows the 3rd radiopaque marker of the device at the tubal ostium, the desired position. The device stimulates tissue ingrowth, causing permanent tubal occlusion.*

Fallopian Tube Occlusion (Incomplete Tubal Occlusion)

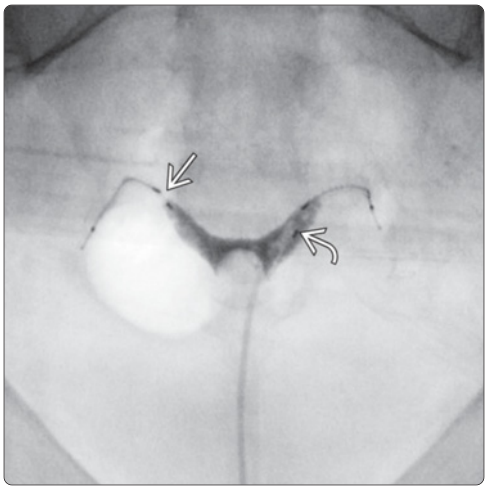


Fallopian Tube Occlusion (Malpositioned Essure Device)

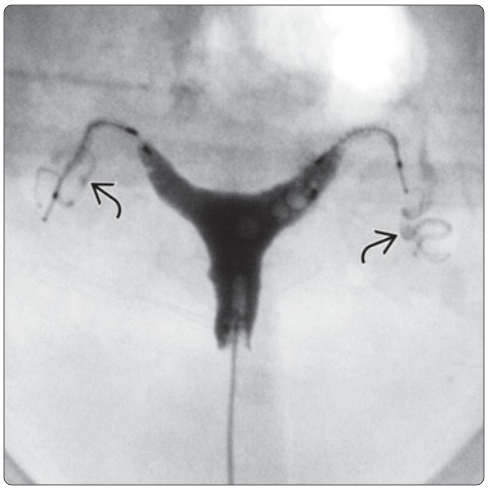


(Left) *Hysterosalpingogram 3 months after bilateral Essure placement shows both devices in place but contrast tracking proximally along each into the tubes. This indicates incomplete tubal occlusion, as contrast should not extend beyond any portion of the coil.* **(Right)** *Transvaginal uterine ultrasound shows the endometrial cavity and bilateral Essure devices. The tip of the right device is in good position, extending slightly into the cornu. The left device is malpositioned, with the tip extending into the uterine cavity.*

Fallopian Tube Occlusion (Malpositioned Essure Device)

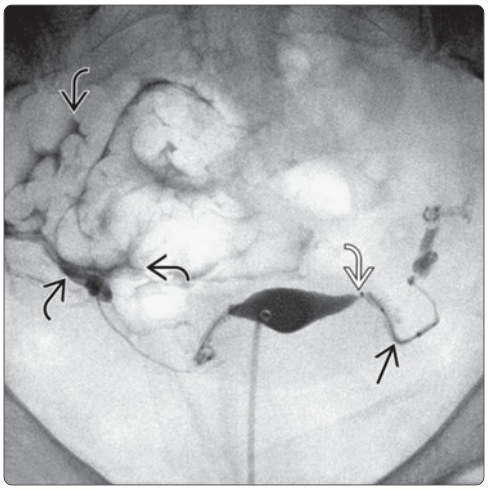


Fallopian Tube Occlusion (Malpositioned Essure Device)

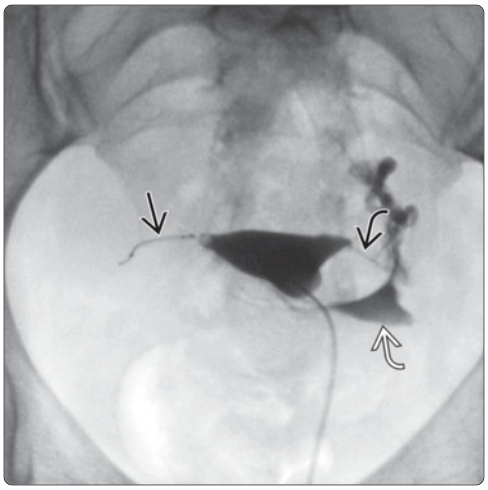


(Left) *Minimal fill view from a hysterosalpingogram 3 months after fallopian tube occlusion procedure shows that the left Essure device tip extends into the uterine cavity, while the right device is optimally positioned. Less than 50% of the inner coil length should trail into the uterine cavity.* **(Right)** *Total fill view of the uterine cavity in the same patient shows that in addition to a malpositioned left Essure device, contrast fills both fallopian tubes, indicating tubal patency despite the procedure 3 months earlier.*

Fallopian Tube Occlusion (Patent Tubes Despite Essure)

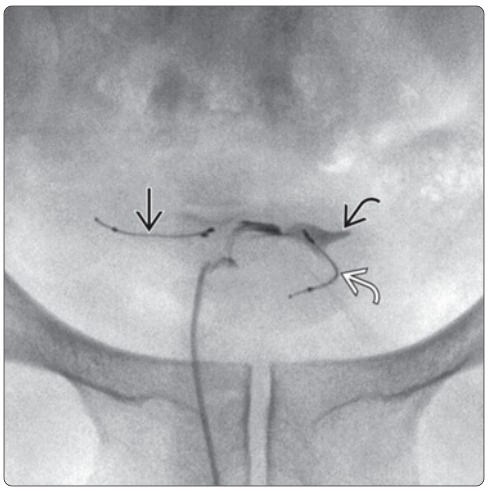


Fallopian Tube Occlusion (Absent Left Essure Device)

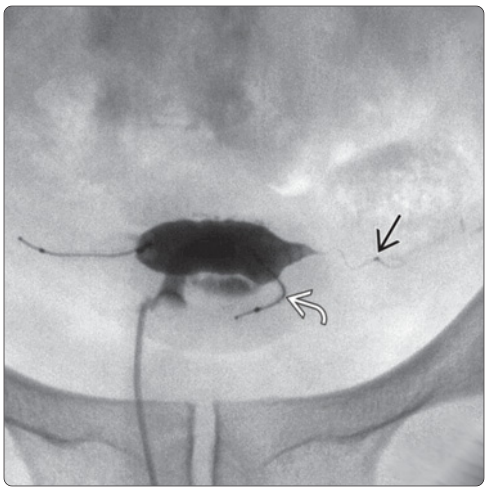


(Left) *Spot radiograph shows a left Essure device positioned distally in the fallopian tube, with proximal marker beyond the cornu. Contrast opacifies the fallopian tubes and spills into the peritoneal cavity, indicating tubal patency.* **(Right)** *Spot radiograph shows a right Essure device in good position, with no fallopian tube filling. There is no device identified on the left, the tube is patent with free peritoneal spillage. The left device was likely malpositioned and expelled.*

Fallopian Tube Occlusion (Malpositioned Left Essure Device)



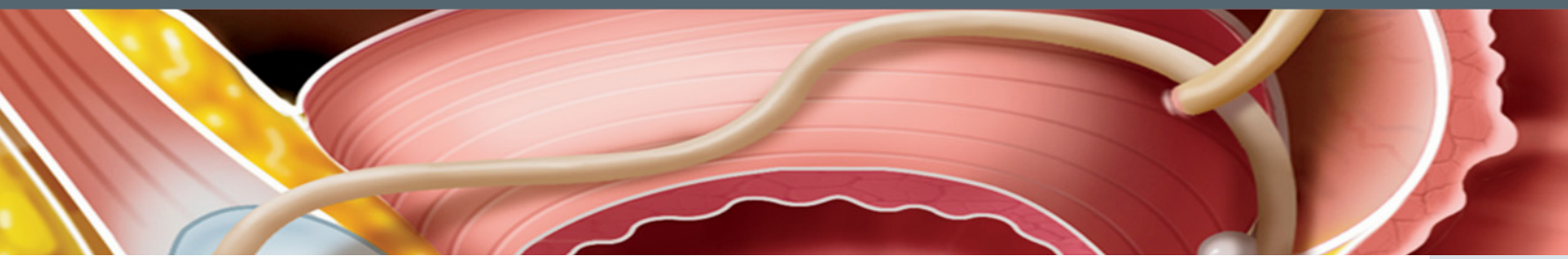
Fallopian Tube Occlusion (Malpositioned Left Essure Device)



(Left) *Minimal fill view from a hysterosalpingogram 3 months after Essure placement shows the left device extending through the uterine wall, not into the fallopian tube (contrast opacifies the cornu). The right device is well positioned.* **(Right)** *Total fill hysterosalpingogram view shows a patent left fallopian tube. During the fallopian tube occlusion procedure, the left Essure device was advanced into a portion of the endometrial cavity mistaken for the uterine cornu.*

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