CT and MR Imaging for Evaluation of Cystic Renal Lesions and Diseases[1]

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Abbreviations: ADPKD = autosomal dominant polycystic kidney disease, GCKD = glomerulocystic kidney disease, MCDK = multicystic dysplastic kidney, MEST = mixed epithelial and stromal tumor, VHL = von Hippel–Lindau disease

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SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Use the Bosniak classification system to categorize renal cysts.
- Describe CT, US, and MR imaging findings of various types of cystic renal disease and the clinical data necessary for diagnosis.
- Discuss the clinical implications of the various types of cystic renal disease.

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Cystic renal lesions are commonly encountered in abdominal imaging. Although most cystic renal lesions are benign simple cysts, complex renal cysts, infectious cystic renal disease, and multifocal cystic renal disease are also common phenomena. The Bosniak classification system provides a useful means of categorizing cystic renal lesions but places less emphasis on their underlying pathophysiology. Cystic renal diseases can be categorized as focal, multifocal, or infectious lesions. Diseases that manifest with focal lesions, such as cystic renal cell carcinoma, mixed epithelial and stromal tumor, and cystic nephroma, are often difficult to differentiate but have differing implications for follow-up after resection. Multifocal cystic renal lesions can be categorized as acquired or heritable. Acquired entities, such as glomerulocystic kidney disease, lithium-induced nephrotoxicity, acquired cystic kidney disease, multicystic dysplastic kidney, and localized cystic renal disease, often have distinct imaging and clinical features that allow definitive diagnosis. Heritable diseases, such as autosomal dominant polycystic kidney disease, von Hippel–Lindau disease, and tuberous sclerosis, are usually easily identified and have various implications for patient management. Infectious diseases have varied imaging appearances, and the possibility of infection must not be overlooked when assessing a cystic renal lesion. A thorough understanding of the spectrum of cystic renal disease will allow the radiologist to make a more specific diagnosis and provide the clinician with optimal recommendations for further diagnostic testing and follow-up imaging.

Introduction

Cystic renal lesions are a common entity seen by the radiologist on a daily basis. The vast majority of these lesions are benign simple cysts, but complex and multifocal cystic renal lesions are also relatively common. The differential diagnosis for a complex cystic lesion is wide. Although the Bosniak classification system provides a useful tool for categorizing and managing complex renal cysts, radiologists should also recognize the imaging appearances of specific types of cystic lesions and understand their pathophysiology. Cystic renal diseases can be categorized as focal, multifocal, or infectious.

TEACHING POINTS

- CT and MR imaging findings of lithium-induced nephrotoxicity typically include normal-sized kidneys with abundant and uniformly distributed renal microcysts.
- Renal malignancy will develop in 3%–7% of patients with acquired cystic kidney disease.
- The presence of intervening normally enhancing renal parenchyma and the absence of a capsule around the cysts help to distinguish localized cystic renal disease from cystic nephroma and multiloculated cystic renal cell carcinoma.
- Renal aspergillosis occurs in immunocompromised patients, including patients with diabetes or human immunodeficiency virus infection and those undergoing corticosteroid therapy.
- About 50% of individuals with ADPKD will develop end-stage renal disease.

Multifocal lesions are further categorized as acquired or heritable entities. A working knowledge of the pathophysiology and imaging characteristics of these various entities allows the radiologist to narrow the differential diagnosis and assist the clinician in tailoring the patient's care more precisely.

Bosniak Classification of Renal Cysts

Over 50% of people older than 50 years have a renal mass or masses. Although the vast majority of these lesions are benign cysts (1), a small percentage are malignant. The Bosniak criteria were introduced to allow the use of specific computed tomographic (CT) findings to help separate nonsurgical from surgical cystic masses and guide patient management (2). These criteria use five separate categories (I, II, IIF, III, and IV) to help communicate the appropriate management to radiologists, urologists, and internists (1). Cystic renal masses are characterized and differentiated by attenuation, contrast material enhancement (perceived vs measurable), and the presence of calcifications and septations (Table).

Bosniak I Cyst

A Bosniak I cyst is a simple cyst. At CT, these cysts have fluid attenuation (0–20 HU) and a hairline-thin wall, do not contain calcifications or septations, and do not demonstrate contrast enhancement. These cysts are benign and require no further evaluation (1).

Bosniak II Cyst

Bosniak II cysts are minimally complicated. They may demonstrate fine calcifications or a short segment of slightly thickened calcification in the cyst wall or septa. They may contain a few hairline-thin septa, in which there may be perceived (as opposed to measurable) enhancement (Fig 1) (1). Homogeneously hyperattenuating (>20 HU) nonenhancing lesions measuring 3 cm or less that are not completely intrarenal are also included in this category (Fig 2). Ideally, at least one-fourth of the hyperattenuating cyst wall should extend outside the renal parenchyma for verification that the cyst wall is smooth (2).

Some studies have reported that homogeneously hyperattenuating lesions that measure at least 70 HU on nonenhanced CT images have a greater than 99.9% likelihood of being benign (3–5). This may obviate further assessment of hyperattenuating cysts found at nonenhanced CT. Bosniak II cysts are considered benign and require no further evaluation (1).

Bosniak IIF Cyst

A Bosniak IIF cyst is a cyst that requires follow-up imaging to determine whether it is benign. These cysts contain more complex elements than Bosniak II cysts but are not believed to be complex enough to warrant surgical intervention. These lesions may show several thin internal septations without measurable enhancement, a few nodular calcifications, and a smooth thickening of the cyst wall. Nonenhancing hyperattenuating cysts that are larger than 3 cm or completely intrarenal are included in this category (1).

According to one study, 95% of these lesions are benign and therefore have a low probability of being small renal cell carcinoma and a low chance of metastasis (6). A more recent study reported a malignancy rate of 25% in surgically excised Bosniak IIF lesions, but the patients with resected Bosniak IIF lesions in that series were highly selected and had a high number of risk factors associated with malignancy, such as a coexisting Bosniak IV lesion or a history of renal cell carcinoma. No patients in this series developed locally advanced or metastatic disease from a Bosniak IIF lesion (7). Bosniak IIF lesions are followed up with CT or MR imaging to assess for increasing complexity that may indicate malignancy. Variable recommendations for the duration of follow-up range from 1–2 years to more than 4 years, depending on the complexity of the lesion (8).

Bosniak III Cyst

A Bosniak III cyst may contain thick irregular walls and/or septa that demonstrate measurable enhancement. Septations are more numerous than in a Bosniak II cyst. The cyst wall and septa may also contain thick nodular calcifications (Fig 3) (1).

These cysts have a reported 30%–100% chance of malignancy, depending on the case series (1). Aside from renal malignancy, the

Bosniak Classification of Renal Cysts

Lesion Type	Morphology	Calcification	Septations	Cyst Wall	Management
Bosniak I	Simple cyst with fluid attenuation (0–20 HU)	None	None	Thin and smooth	Benign; no follow-up needed
Bosniak II	Minimally complex cyst or a well-marginated, uniformly hyperattenuating cyst; diameter ≤3 cm; partially outside the kidney	Fine or minimally thick calcifications in wall or septa*	A few hairline-thin septa without measurable enhancement*	Thin and smooth	Benign; no follow-up needed
Bosniak IIF	More complex elements than a Bosniak II cyst but fewer than a Bosniak III cyst, or a uniformly hyperattenuating cyst that does not meet Bosniak II criteria	May contain a few small nodular calcifications*	Multiple thin internal septations without measurable enhancement*	May be mildly thickened, without measurable enhancement	Follow-up CT or MR imaging to assess for increasing complexity, which may indicate malignancy
Bosniak III	Complex cyst with enhancing septations or wall	Variable	May be thick or irregular, with measurable enhancement	May be thick or irregular, with measurable enhancement	30%–100% chance of malignancy; resection recommended
Bosniak IV	Cystic mass with enhancing soft-tissue components	Variable	Clearly enhancing nodule in septa	Clearly enhancing nodule in wall	Malignant until proven otherwise; resection recommended

Note.—MR = magnetic resonance.
*Hyperattenuating Bosniak II and IIF cysts do not have septations or calcifications.

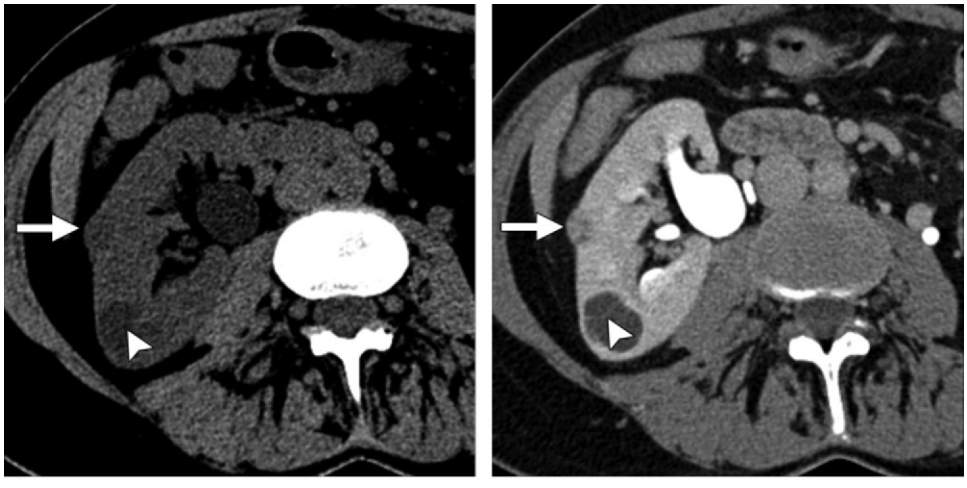

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Figure 1. Bosniak II lesion. **(a)** Axial nonenhanced CT image shows a cyst with a subtle septation (arrowhead) in the right kidney, as well as an ipsilateral renal lesion (arrow). **(b)** Axial contrast-enhanced CT image shows perceptible but not measurable enhancement within the cyst (arrowhead). The ipsilateral solid enhancing renal lesion is also seen (arrow). Both lesions were surgically removed, and the cystic lesion was found to be a benign multiloculated cyst.

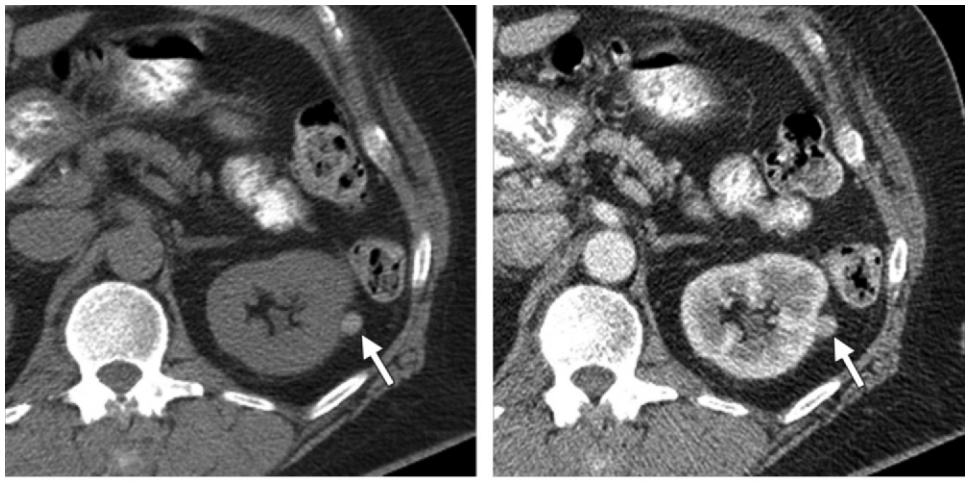

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Figure 2. Bosniak II lesion. **(a)** Axial nonenhanced CT image shows a hyperattenuating (82-HU) structure (arrow) arising from the upper pole of the left kidney. **(b)** Axial contrast-enhanced CT image shows no enhancement (87 HU) in the structure (arrow). The lesion was removed, along with an ipsilateral solid renal lesion (not shown), and was found to be a benign cyst with hemorrhage.

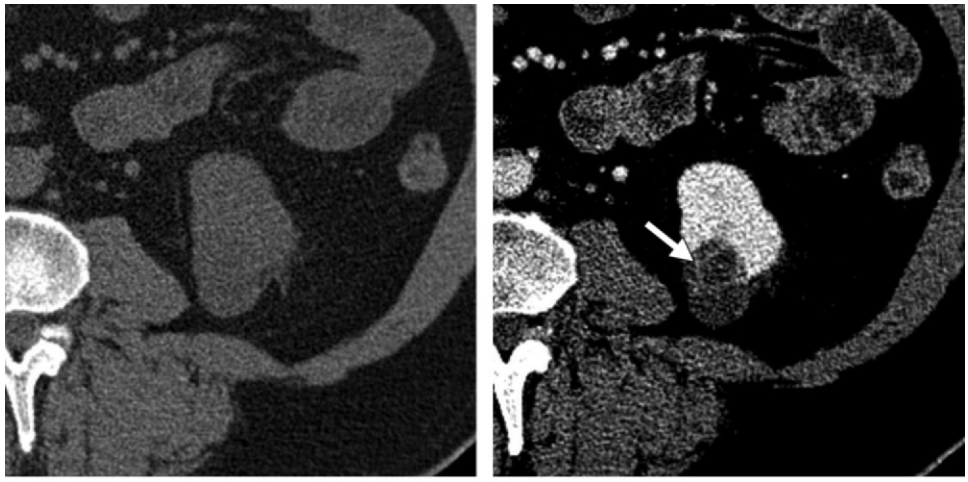

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Figure 3. Bosniak III lesion. **(a)** Axial nonenhanced CT image shows a low-attenuating (18-HU) mass arising from the left kidney. **(b)** Axial contrast-enhanced CT image shows multiple measurably enhancing (45- to 75-HU) septations in the mass (arrow). The lesion was resected and was found to be cystic renal cell carcinoma.

differential diagnosis for a Bosniak III cyst includes cystic nephroma, mixed epithelial and stromal tumor (MEST), benign multiloculated cyst, hemorrhagic cyst, and renal abscess. Patient management can be difficult because of the broad differential diagnosis, which contains both benign and malignant entities. Given the risk for malignancy, these patients usually undergo surgical resection. However, the risks and benefits of surgery in a particular patient must be weighed against the possibility that the lesion may be benign. Some authors recommend lesion biopsy (9). However, the relatively small volume of tumor cells within the cyst may increase the risk for a false-negative biopsy result (1).

Bosniak IV Cyst

A Bosniak IV cyst contains enhancing nodular soft-tissue components and is considered malignant until proven otherwise (Fig 4). A small percentage of these lesions have a benign pathology (eg, a MEST). Surgical resection is recommended unless factors such as patient comorbidities or a limited life expectancy militate against this treatment strategy.

Focal Cystic Renal Disease

Mixed Epithelial and Stromal Tumor

MEST is a relatively recently described pathologic entity with fewer than 100 reported cases in the

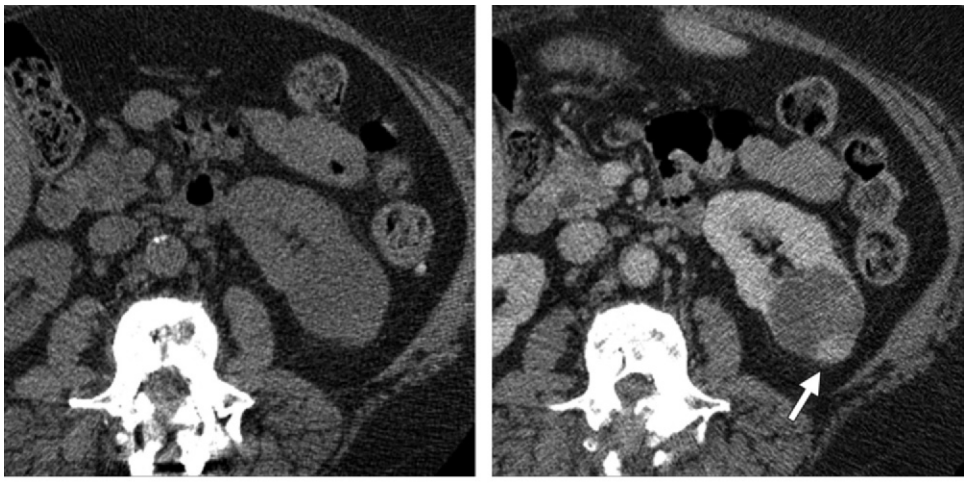

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Figure 4. Bosniak IV lesion. **(a)** Axial nonenhanced CT image shows a low-attenuating (22-HU) cystic lesion arising from the left kidney. **(b)** Axial contrast-enhanced CT image shows an enhancing (114-HU) peripheral nodule (arrow) with subtle enhancing septations. The lesion was resected and was found to be cystic renal cell carcinoma.

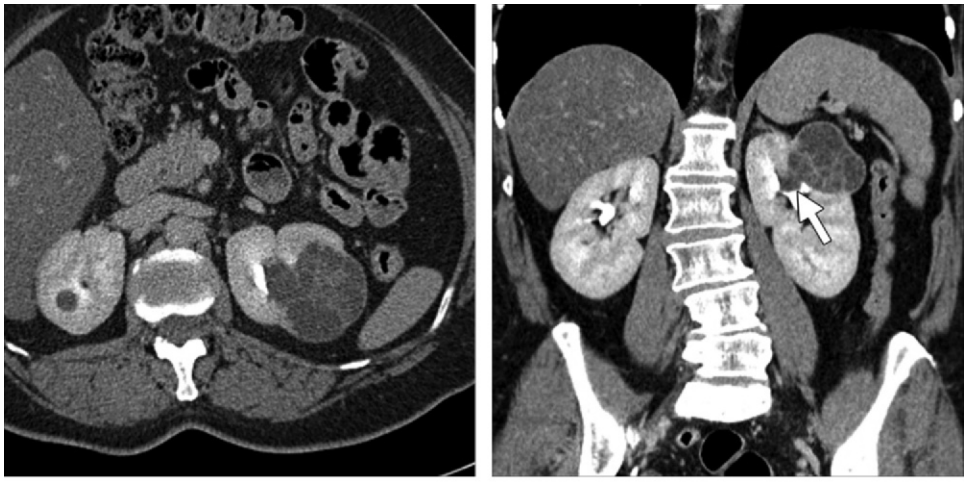

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Figure 5. Axial **(a)** and coronal **(b)** contrast-enhanced CT images show a cystic and solid mass in the left kidney, with numerous enhancing septations. The mass extends into the renal pelvis (arrow in **b**) and was confirmed to be MEST at histologic analysis.

literature. These complex cystic and solid masses are characterized histologically by the presence of stromal elements that resemble ovarian stroma, as well as an epithelial component consisting of cysts with an epithelial lining (10). MEST is most frequently encountered in perimenopausal women (female-to-male ratio, 11:1), with an average age at presentation of 56 years (range, 17–84 years) (11). The use of oral estrogen supplements or contraceptives is frequently reported in patients with MEST. Common presenting symptoms include hematuria, flank pain, or a flank mass, although 25% of patients are asymptomatic (12). The imaging appearance is nonspecific, and MEST cannot be confidently differentiated from cystic renal cell carcinoma at imaging (10). Typically, MEST manifests at imaging as an expansile multiloculated cystic mass that may herniate into the renal pelvis (Fig 5). The intervening septa show variable contrast enhancement, and an enhancing mural nodule or mural calcifications may be seen. Delayed enhancement is often noted. Intralesional hemorrhage may be seen and is suggested by the presence of hyperattenuating components at CT or hyperintense foci at T1-weighted MR imaging (10). Because MEST cannot be confidently distinguished from cystic renal cell carcinoma, most cases are treated surgically.

Cystic Nephroma

Cystic nephroma is a rare benign tumor that typically occurs in boys younger than 4 years

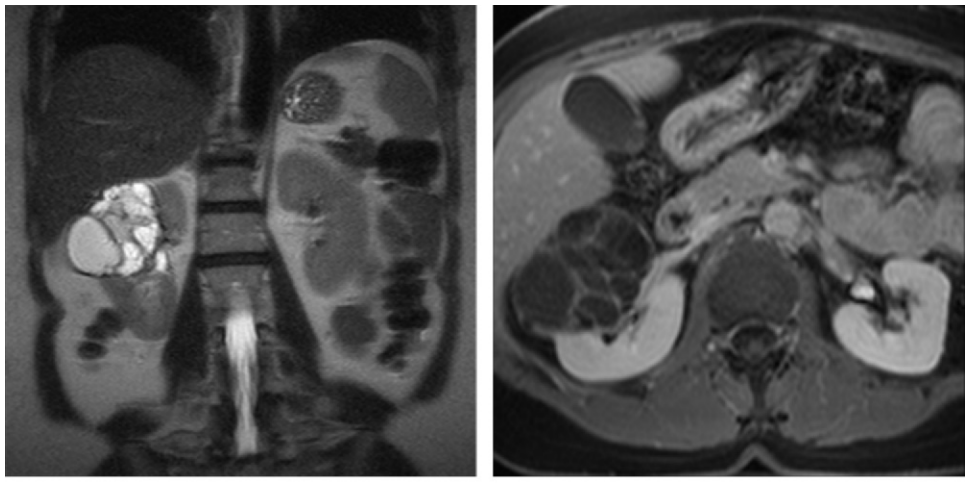

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Figure 6. Cystic nephroma. Coronal T2-weighted MR image **(a)** and axial contrast-enhanced fat-saturated T1-weighted gradient-recalled-echo MR image **(b)** show a multiseptated cystic mass arising from the upper pole of the right kidney and extending into the renal pelvis. In **a**, variable signal intensity in the contents of the cyst locules represents areas of simple and complex fluid, and signal hypointensity in the surrounding capsule and septa of the lesion reflects their fibrous content. Enhancement of the thin septa is seen in **b**.

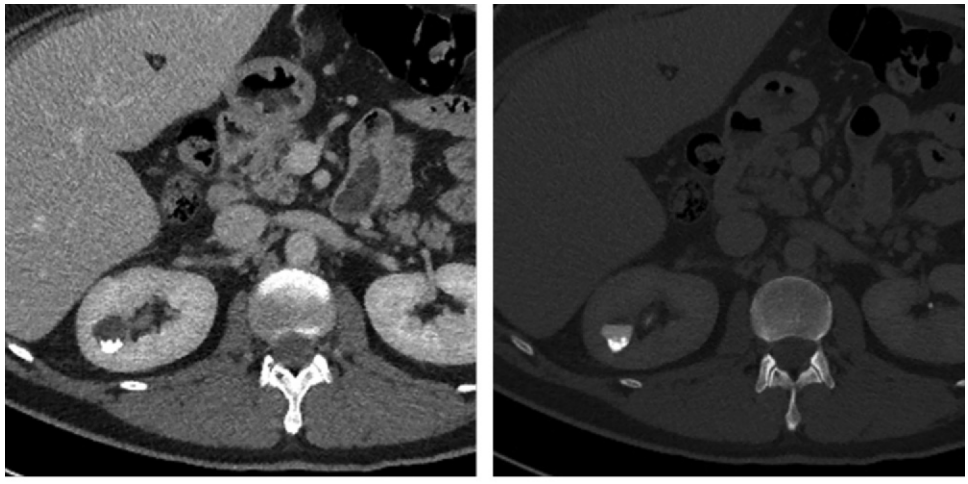

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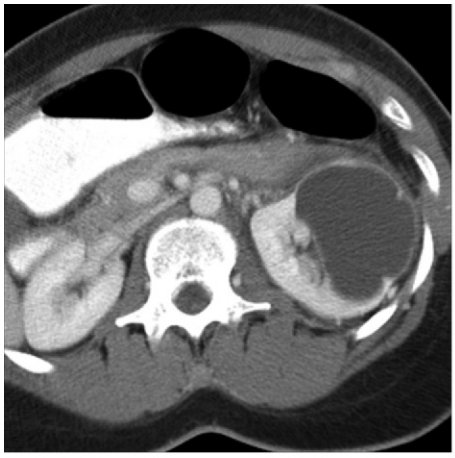
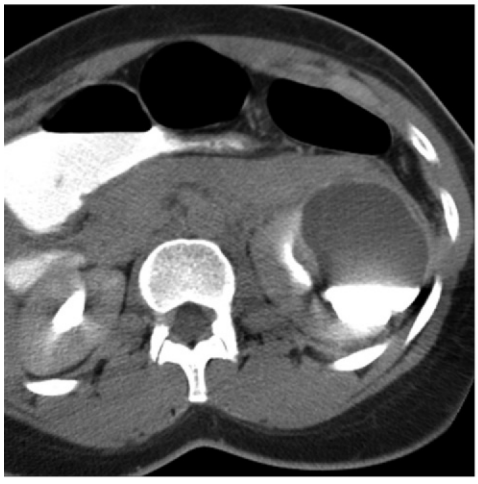
Figure 7. Pyelocalyceal diverticulum. **(a)** Axial contrast-enhanced nephrographic phase CT image shows calcifications in a cystic renal structure. **(b)** Axial contrast-enhanced excretory phase CT image shows layering of excreted contrast material in the cystic structure, a finding that proves communication with the collecting system.

or women aged 40–60 years. *Cystic nephroma* is a general term that encompasses a wide pathologic spectrum of benign cystic masses. It has been postulated that cystic nephromas and MESTs may be part of the same spectrum of stromal epithelial lesions (13,14). Children often present with a painless abdominal mass, while adults are more likely to present with abdominal pain or hematuria but may be asymptomatic (13). On cross-sectional images, cystic nephroma typically appears as a well-circumscribed, encapsulated, multicystic mass with variably enhancing septa. When compared with simple fluid, the contents of the mass may have similar or slightly higher attenuation at CT and higher signal intensity at T1-weighted MR imaging. The lesions often herniate into the renal pelvis or proximal ureter. Because of their fibrous composition, the septa and capsule of the lesion are hypointense on both T1- and T2-weighted MR images (Fig 6) (13,14). Nonetheless, cystic nephroma may mimic a variety of benign and malignant renal lesions, including complex renal cysts and cystic renal cell carcinoma, along with MEST and localized cystic renal disease. Because it is challenging to distinguish cystic nephroma from cystic renal cell carcinoma, many cases are treated surgically.





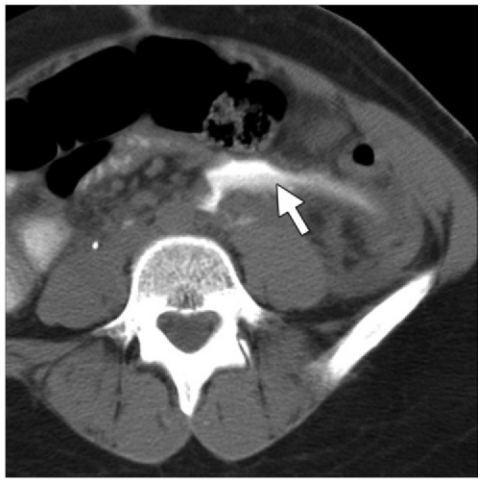


Figure 8. Ruptured pyelocalyceal diverticulum in a woman who presented with left flank pain. **(a)** Axial contrast-enhanced CT image shows a cystic lesion in the left kidney. **(b, c)** Axial contrast-enhanced delayed phase CT images show filling of the cystic structure with excreted contrast material, a finding that confirms connection with the collecting system. Excreted contrast material tracks from this structure into a retroperitoneal fluid collection (arrow in **c**), a finding that confirms a urinoma from a ruptured pyelocalyceal diverticulum.

Pyelocalyceal Diverticulum

Pyelocalyceal diverticulum is a urine-filled cavity within the renal cortex that communicates with the renal collecting system. It is lined with transitional epithelium. There are two types of pyelocalyceal diverticulum: type 1, the more common type, communicates with a minor calyx, and type 2 connects with a major calyx or the renal pelvis. Pyelocalyceal diverticulum is relatively rare and is identified at 2.1–4.5 per 1000 intravenous urographic evaluations (15). The etiology of pyelocalyceal diverticulum is controversial; however, most cases are thought to be congenital. The most common theory is that pyelocalyceal diverticulum results from failure of regression of the third- and fourth-generation ureteric buds. A smaller percentage of cases of pyelocalyceal diverticulum may be acquired, resulting from obstructing stones or infection. Fifty percent of cases of pyelocalyceal diverticulum are complicated by calculi and milk of calcium. This is likely due to a combination of urinary stasis and repeated infection (15).

A simple pyelocalyceal diverticulum without stones has an appearance similar to that of a simple cyst at ultrasonography (US), MR imaging, and nonenhanced or nephrographic phase contrast-enhanced CT. In the excretory phase of imaging, the structure fills with contrast material because of communication with the renal collecting system, and layering of contrast material is seen within the cystlike structure (16). A large pyelocalyceal diverticulum may be difficult to distinguish from a hydrocalyx due to calyceal obstruction from benign or malignant causes.

At US, a pyelocalyceal diverticulum complicated by calculi or milk of calcium appears as an anechoic cyst with layering, mobile, echogenic material. At nonenhanced CT, it similarly appears as a cyst with layering high-attenuating material. On excretory phase CT images, excreted contrast material layers within the cyst (Fig 7) (15).

Complications of pyelocalyceal diverticulum include hemorrhage, infection, and cyst rupture (Fig 8). Malignancy in a pyelocalyceal diverticulum is exceedingly rare, with only four cases reported in the literature (17).

Acquired Multifocal Cystic Renal Disease

Glomerulocystic Kidney Disease

Glomerulocystic kidney disease (GCKD) is a rare form of cystic renal disease characterized histologically by uniform cystic dilatation of the Bowman capsule (18). These cysts are located predominantly in the subcapsular area of the renal cortex and spare the tubular portions of the nephron (19). The cortical distribution helps distinguish GCKD from other cystic renal diseases, such as autosomal recessive polycystic kidney disease, autosomal dominant polycystic kidney disease (ADPKD), multicystic dysplastic kidney (MCDK), and cystic renal dysplasia (20).

Most often, GCKD occurs in neonates and young children. In many instances, mild renal failure with accompanying infantile congenital malformations is seen. Although an autosomal dominant inheritance pattern has been demonstrated in families with GCKD, several sporadic cases have also been reported (19,21).

The predominant imaging finding is small renal cysts with a primarily cortical subcapsular distribution (Fig 9). The kidneys may be normal in size or hypoplastic (22). The cysts may be difficult to define at US, and cortical hyperechogenicity may be seen instead of the typical anechoic appearance of simple cysts (21). Because patients typically present with symptoms of renal failure, nonenhanced CT is typically performed but is of limited value in establishing the diagnosis (20). Nonenhanced T2-weighted MR imaging is the preferred imaging modality.

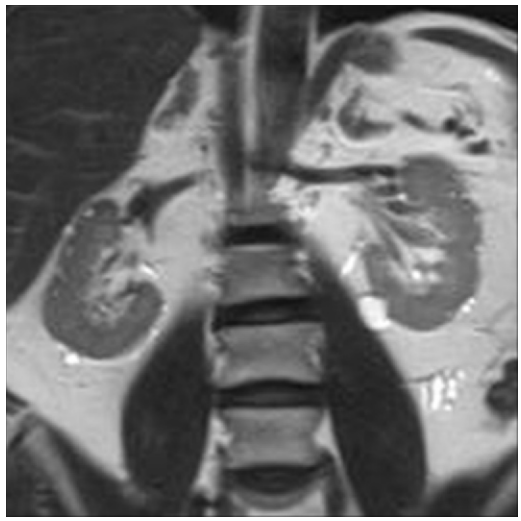


Figure 9. Coronal T2-weighted MR image shows numerous hyperintense lesions predominantly in the subcapsular portions of the renal cortex, a finding indicative of simple cysts. This distribution is a classic finding in GCKD. (Courtesy of Peter Humphrey, MD, Northwest Imaging, Kalispell, Mont.)

Lithium-induced Nephrotoxicity

Nephrotoxicity due to long-term lithium therapy is a well-described entity and is divided into three main categories: acute intoxication, nephrogenic diabetes insipidus, and chronic renal disease. Nephrogenic diabetes insipidus is the most common complication of lithium-induced nephrotoxicity and typically is reversible with drug cessation (23–25). However, patients who undergo long-term lithium therapy may develop chronic focal interstitial nephritis that results in progressive, nonreversible, chronic renal insufficiency. In the setting of progressive renal dysfunction and lithium therapy, clinical and laboratory data typically are sufficient to establish the diagnosis. However, if a renal biopsy is performed, it will show tubular atrophy, glomerulosclerosis, interstitial fibrosis, and distal tubular dilatation with microcyst formation (24).

CT and MR imaging findings of lithium-induced nephrotoxicity typically include normal-sized kidneys with abundant and uniformly distributed renal microcysts (26). Microcysts have been reported in 33%–62% of patients who are undergoing lithium therapy (24). These cysts are found in the cortex and medulla and typically are 1–2 mm in diameter. At CT, the cysts appear as multiple tiny hypoattenuating lesions (Fig 10). MR imaging best depicts the number and size of cystic lesions (27). Cysts are best appreciated on T2-weighted MR images, which will depict multiple hyperintense foci (26,27) (Fig 11). The differential diagnosis includes ADPKD, GCKD, medullary cystic kidney disease, and acquired cystic kidney disease. These entities can usually be differentiated from lithium-induced nephro-

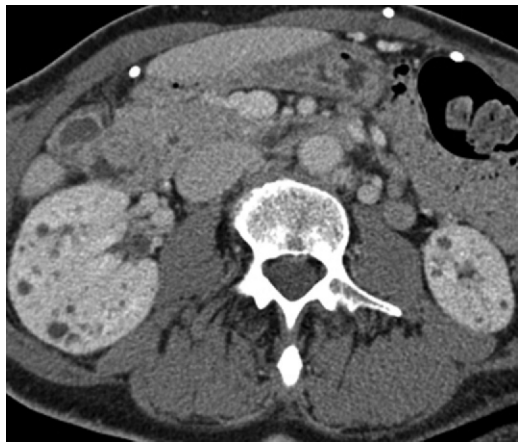


Figure 10. Lithium-induced nephrotoxicity in a patient undergoing long-term lithium therapy. Axial contrast-enhanced CT image shows multiple hypoattenuating lesions scattered throughout the cortex and medulla of both kidneys. The findings are consistent with microcysts.

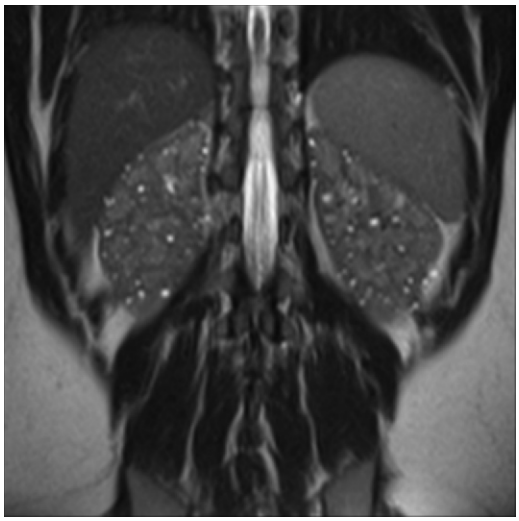


Figure 11. Lithium-induced nephrotoxicity in a patient undergoing long-term lithium therapy. Coronal T2-weighted MR image shows multiple tiny hyperintense lesions uniformly distributed throughout both kidneys. The findings are consistent with microcysts.

toxicity on the basis of clinical history as well as the size and distribution of cysts at imaging (27).

Acquired Cystic Kidney Disease

Acquired cystic kidney disease refers to the development of renal cysts in patients with end-stage renal disease who do not already have a heritable cystic renal disease. Because renal cysts are a common phenomenon, diagnostic criteria require the presence of at least three cysts in each kidney (28). The cysts are usually small (0.5–3 cm in diameter). Approximately 8%–13% of patients with end-stage renal disease have acquired cystic kidney disease before they undergo dialysis. The incidence increases with the length of time on dialysis, with approximate rates of 13%, 50%, and 87% after 2, 6, and 9 years of dialysis, respectively (28). After 10 years of dialysis, the incidence is believed to approach 100%. The male-to-female ratio is 3:1. There is no correlation with race, cause of renal failure, or method of dialysis (ie, hemodialysis or peritoneal dialysis) (28).

Acquired cystic kidney disease is thought to be multifactorial in cause. The initiating factor is thought to be compensatory hypertrophy of functional nephrons in response to adjacent tissue destruction. Renal tubular obstruction and expansion caused by interstitial fibrosis, epithelial hyperplasia, and increased fluid secretion are thought to lead to cyst development (28).

Complications include cyst hemorrhage, ureteral stones, and the development of renal cell carcinoma. Cyst hemorrhage occurs in up to half of cases and may proceed to cyst rupture, with resultant perinephric and retroperitoneal hemorrhage. In rare cases, hemorrhage may be life threatening (28).

At US, acquired cystic kidney disease manifests as atrophic echogenic kidneys with cysts of varying size and complexity. CT and MR imaging similarly demonstrate small kidneys with cystic lesions that vary in size and complexity (Fig 12).

Renal malignancy will develop in 3%–7% of patients with acquired cystic kidney disease (28). Risk factors include male sex and an increased length of time on dialysis (28). After renal transplant, the cysts usually regress; however, patients continue to have an increased risk for malignancy in the native kidney. In patients who have undergone renal transplant, these malignancies tend to be more aggressive because of the immunosuppression therapy necessary to prevent graft rejection (29).

Recommendations vary for screening asymptomatic patients with acquired cystic kidney disease for malignancy. Patients with end-stage renal disease, especially those undergoing dialysis, are already quite ill and have high mortality from other causes. Screening of asymptomatic patients should be based on the expected benefit in light of the patient's overall health and life expectancy (29).

Multicystic Dysplastic Kidney

MCDK is a nonheritable developmental disorder in which the kidney is replaced by nonfunctioning noncommunicating cysts. It occurs in approximately one of 4300 live births (28). Although there are several theories about the etiology of MCDK, the most accepted is that it results during embryogenesis from the failure of the ascending ureteric bud to induce the metanephric blastema to differentiate into nephrons and the various stromal elements of the kidney. Without the chemical signals from the ureteric bud, the metanephric blastema differentiates into a mass of cartilage, cysts, and unorganized tubules. Less-accepted theories implicate

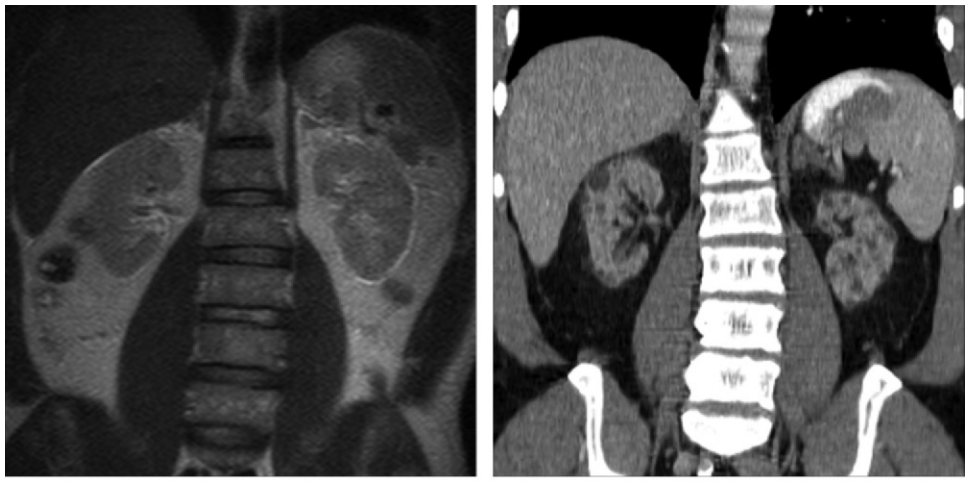


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Figure 12. Acquired cystic renal disease. **(a)** Coronal T2-weighted MR image shows no renal lesions. **(b)** Coronal contrast-enhanced CT image obtained in the same patient 6 years later, after 5 years of dialysis, shows numerous small cysts throughout both kidneys.

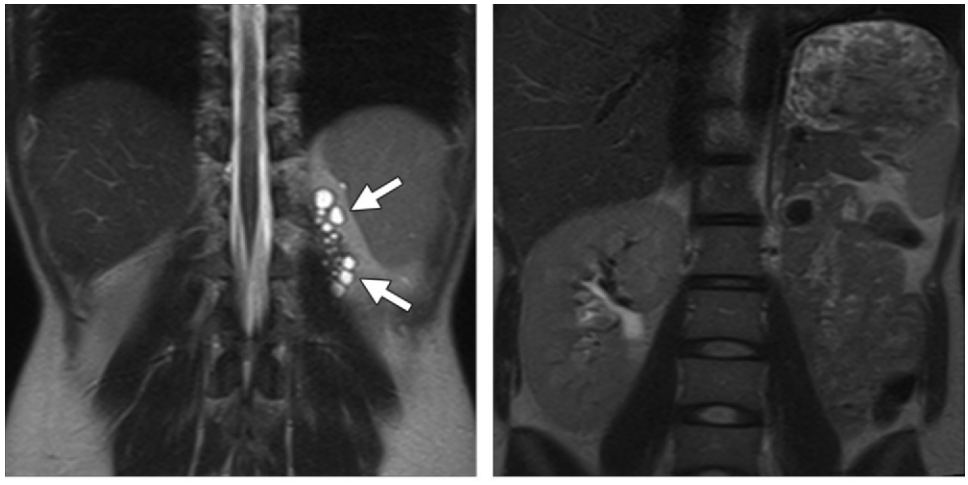

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Figure 13. Coronal T2-weighted MR images show a small collection of hyperintense cysts in the left renal fossa (arrows in **a**), a finding that represents MCDK. Compensatory hypertrophy of the otherwise normal right kidney is seen in **b**.

urinary tract obstruction or teratogens such as viral infections or medication as the cause of MCDK (30).

The imaging appearance of MCDK varies by age. Often it is diagnosed in utero. In children, it is most commonly assessed with US and appears as a large cluster of noncommunicating cysts that replaces the renal parenchyma. In some cases, the cysts may replace only a portion of the renal parenchyma (28). In children diagnosed with MCDK, it is important to assess the contralateral renal collecting system because there is vesicoureteral reflux in 5%–43% of cases (30).

In most cases, the dysplastic kidney will completely regress by adulthood. However, in a minority of cases, a small residual mass may be seen in the renal fossa at imaging. Typical imaging features include a central region of soft tissue; peripheral noncommunicating cysts; and absence or severe atrophy of the ipsilateral ureter, renal collecting system, and renal vasculature (Fig 13) (28). Occasionally, the cysts may be calcified and identifiable at radiography (31). There may be compensatory hypertrophy of the contralateral kidney (30).

In adults with residual MCDK, the mass may regress or follow an indolent course but is typically not a source of clinical concern (28).

Localized Cystic Renal Disease

Localized cystic renal disease is a rare condition in which a cluster of tightly spaced, variably sized cysts replaces a portion of or the entire renal parenchyma of one kidney (Fig 14). Normal renal parenchyma is seen between the cysts. The re-

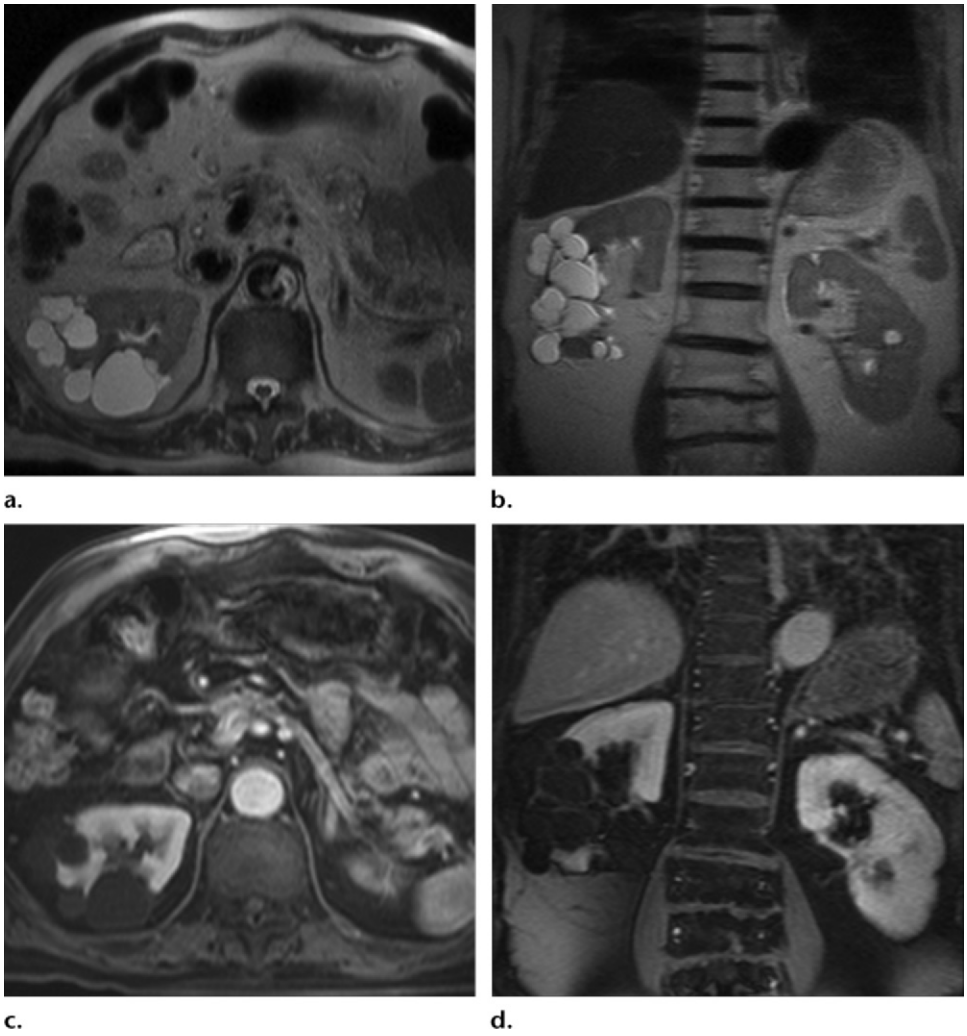


Figure 14. Localized cystic renal disease. Axial **(a)** and coronal **(b)** T2-weighted MR images and axial **(c)** and coronal **(d)** contrast-enhanced fat-saturated T1-weighted gradient-recalled-echo MR images show a cluster of nonenhancing cysts replacing the middle of the right kidney. No capsule is seen around the cysts, and normal renal parenchyma is seen between the cysts.

maining renal parenchyma in the affected kidney as well as in the contralateral kidney may contain a few scattered cysts (32). Unlike ADPKD, localized cystic renal disease is unilateral, and other organs do not have cystic involvement. In contrast to cystic nephroma, no capsule is seen around the cysts (32). Localized cystic renal disease is a rare nonheritable form of cystic renal disease and is not progressive. The pathogenesis is unclear, with an acquired developmental origin postulated. At gross pathologic analysis, the cysts are lined with a single layer of flattened epithelium (33).

Patients with localized cystic renal disease may be asymptomatic. Common presenting symptoms include abdominal pain, a flank mass, hematuria, and hypertension. There is no impairment of renal function (28).

At imaging, localized cystic renal disease manifests as a conglomeration of cysts in one kidney. The presence of intervening normally enhancing renal parenchyma and the absence of a capsule around the cysts help to distinguish localized cystic renal disease from cystic nephroma and multiloculated cystic renal cell carcinoma (32). However, the intervening renal parenchyma may be stretched and attenuated, which makes differentiation of thick septations from intervening parenchyma difficult. Tight spacing of the cysts may also limit detection of a capsule. However, close inspection of axial CT images and the use of multiplanar reformatted CT and multiplanar MR imaging should allow accurate assessment of the presence or absence of these features (32). Hemorrhage, a finding typically seen in the setting of trauma, has been reported within the cysts (32). Although localized cystic renal disease is a benign entity, surveillance imaging may be appropriate in equivocal cases and in symptomatic patients.

Cysts of the Renal Pelvis

Cysts involving the renal pelvis are classified as parapelvic or renal sinus cysts according to their site of origin. Parapelvic cysts are cortical cysts that extend into the renal sinus fat. Histologically, they are lined with a single epithelial layer. They usually are single and unilateral but may be multiple. Although most are asymptomatic, parapelvic cysts may cause hydronephrosis if they compress the collecting system. They may rarely cause hypertension if they compress the renal artery. In addition, they may become infected and may hemorrhage. They also may cause pain or hematuria. At US, CT, and MR imaging, parapelvic cysts are simple cysts that extend into the renal sinus fat (34). Rarely, parapelvic cysts have complex features at imaging.

Renal sinus cysts (sometimes referred to as *peripelvic cysts*) are asymptomatic cysts that arise from the lymphatics of the renal hilum. They usually are multiple, small, and bilateral. Their appearance is often difficult to distinguish from hydronephrosis. However, the distinction is readily apparent on contrast-enhanced excretory phase CT images, where the high-attenuation contrast agent–filled collecting system can easily be distinguished from fluid-attenuation renal sinus cysts (Fig 15) (34).

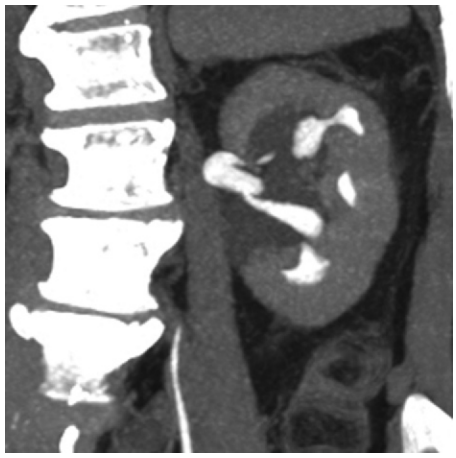


Figure 15. Peripelvic cysts. Coronal excretory phase maximum intensity projection CT image of the abdomen shows the contrast material–filled collecting system surrounded by multiple cysts in the renal pelvis.

Renal Infection

Pyogenic Abscess

Renal abscesses are typically seen as a complication of acute pyelonephritis. They often occur as the result of untreated or incompletely treated pyelonephritis or ascending urinary tract infections. Less commonly, renal abscesses may be caused by hematogenous seeding in patients with bacteremia. Immunocompromised, pregnant, and diabetic patients have an increased risk for abscess formation.

At radiology, a renal abscess may mimic a renal cyst; however, differentiation can typically be made on the basis of the patient's clinical scenario. US most commonly demonstrates an anechoic to hypoechoic complex cystic mass with posterior acoustic enhancement. Internal echoes, septations, and loculations may also be seen at US (35). CT and MR imaging are more sensitive modalities for detecting a renal abscess. Contrast-enhanced CT demonstrates a well-defined, hypoattenuating, round mass, often with a thick rim or halo (Fig 16). Perinephric extension of the abscess may be seen. MR imaging can also be used to evaluate renal abscesses, particularly in cases where it is important to limit ionizing radiation exposure. MR imaging typically shows a heterogeneous thick-rimmed lesion that is hypointense on T1-weighted images and hyperintense on T2-weighted images, with restricted diffusion. The presence of gas will result in blooming artifact on gradient-recalled-echo MR images.

Renal Apergillosis

Renal aspergillosis is an uncommon renal infection that manifests as a complex cystic renal mass. Renal aspergillosis occurs in immunocompromised patients, including patients with diabetes or human immunodeficiency virus infection and those undergoing corticosteroid therapy (36). Renal aspergillosis is acquired in one of three ways: hematogenous dissemination, ascending infection, or *Aspergillus* casts in the renal pelvis (36). Hematogenous dissemination is the most common mechanism of acquisition. The radiologic appearance of renal aspergillosis mimics that of a complex renal cyst or abscess. CT demonstrates a hypoattenuating mass with a thick enhancing wall and internal septations (Fig 17). The surrounding renal parenchyma may demonstrate features of focal pyelonephritis and delayed enhancement (37). The diagnosis of renal aspergillosis is typically made after urinalysis or aspiration of the lesion.

Renal Echinococcosis

Echinococcosis is a zoonosis caused by the larvae of the *Echinococcus* tapeworm (*Echinococcus granulosus*). Humans can be an intermediate host when infected through contact with the definitive host or through contaminated water or food. The human liver serves as the first line of defense and is the most commonly involved organ (75%). The lungs are the second most common site of involvement (15%). However, hematogenous dissemination can occur to almost any anatomic

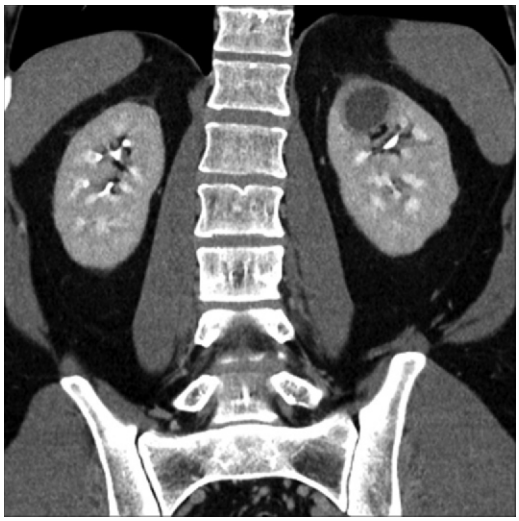
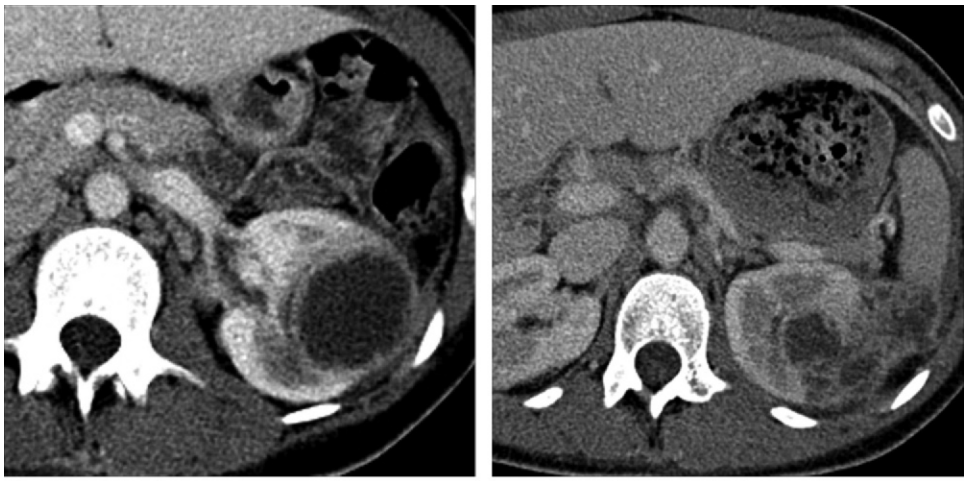


Figure 16. Renal abscess in a patient with a 1-week history of fever, chills, and left flank pain who had recently been treated for pyelonephritis. Coronal contrast-enhanced CT image shows a hypoattenuating mass with a thickened enhancing rim in the left kidney, a finding consistent with a renal abscess.



a. b.

Figure 17. Renal aspergillosis in a patient with acquired immunodeficiency syndrome and known aspergillosis who presented with flank pain and fever. **(a)** Axial contrast-enhanced CT image shows a large unilocular cystic mass arising from the left kidney, with thick rim enhancement. **(b)** Axial contrast-enhanced CT image obtained 4 months later shows progression to a multilocular, complex, inflammatory mass that extends into the perinephric space.

location (38) and may rarely involve the kidneys. Renal echinococcosis is seen in 2%–3% of patients with echinococcal disease (38). In patients with renal dissemination, hydatid cysts form but typically are asymptomatic for many years. Patients with cyst growth can become symptomatic and may present with a palpable mass, pain, and dysuria. Large cysts can rupture and may result in a severe antigenic immune response, such as anaphylaxis (39).

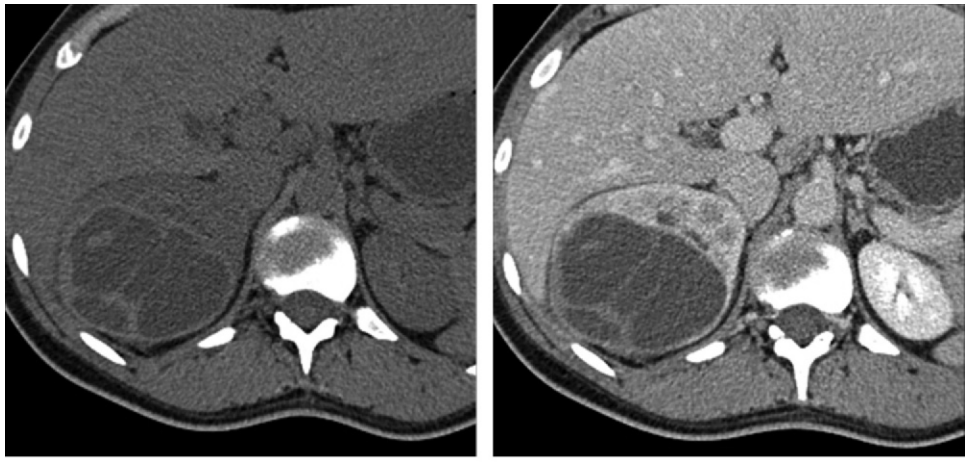
Renal hydatid cysts typically are unilateral single lesions that are found in the upper or lower poles of the kidney (40). Imaging findings vary according to the stage of cyst growth. Renal hydatid cysts initially appear as unilocular cystic masses. In the intermediate stage, the cysts appear as a multilocular cystic mass because of the development of multiple daughter cysts (Fig 18). These cystic masses demonstrate a thick enhancing wall. Internal septations, when present, typically demonstrate enhancement. With quiescence or death of the parasite, the lesion becomes calcified.

Heritable Multifocal Cystic Renal Disease

Autosomal Dominant Polycystic Kidney Disease

ADPKD is a heritable cystic renal disease that causes cysts along the nephron. The disease occurs in approximately one of 500–1000 live births and is the most common heritable renal disorder (41).

There are two types of ADPKD. The more common type is responsible for 85% of cases and is associated with a mutation in the *PDK1* gene, which is located on the short arm of chromosome



a. b.

Figure 18. Renal echinococcal (hydatid) disease. **(a)** Axial nonenhanced CT image shows a complex cystic mass arising from the right kidney. **(b)** Axial contrast-enhanced CT image shows the thick enhancing wall of the mass, with enhancing internal septations.

16. The less common type is associated with mutations in the *PDK2* gene, which is located on the long arm of chromosome 4. The *PKD2* mutation results in a milder form of disease than the *PKD1* mutation and also has a later age of onset (42).

The radiologic appearance of ADPKD includes enlarged kidneys with multiple cysts (Fig 19). Extrarenal manifestations may also be seen and most commonly include hepatic cysts. Cysts are less commonly seen in the pancreas, spleen, ovaries, and testes. Patients are at an increased risk for cardiac valve abnormalities (26%) and intracranial aneurysms (10%–15%) (43).

Renal complications of ADPKD include cyst hemorrhage, cyst infection, cyst rupture, and nephrolithiasis. About 50% of individuals with ADPKD will develop end-stage renal disease. Unlike in some other congenital cystic kidney diseases, there is no increased risk for renal cell carcinoma unless the patient is undergoing prolonged dialysis (28).

Imaging plays a crucial role in the diagnosis of ADPKD in patients with a first-degree relative with the disease. Imaging is especially important in the screening of individuals who plan to donate a kidney to a parent with the disease. Although diagnosis of ADPKD in its most severe form is relatively straightforward, younger patients and those with the *PKD2* mutation often have a milder form of the disease (41), and imaging findings may overlap with those seen in an unaffected patient with incidentally noted renal cysts. Genetic testing identifies only 70% of individuals with ADPKD (42).

Criteria for diagnosing ADPKD in patients at high risk for the disease (ie, those with a first-degree relative with the disease) were published by Ravine et al (44) in 1994. These worked well

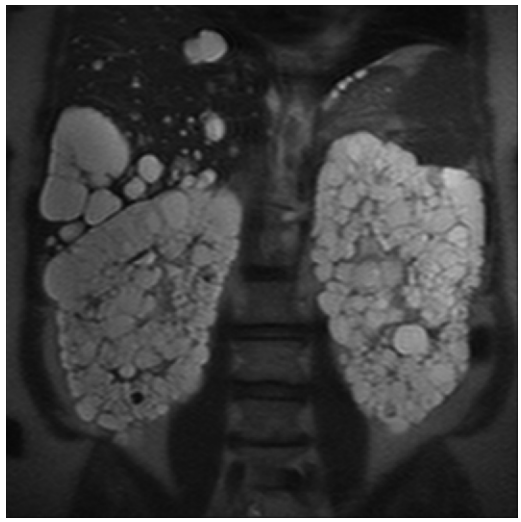
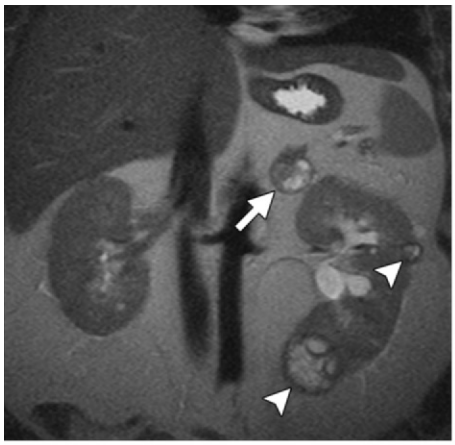


Figure 19. Coronal T2-weighted MR image shows the kidneys expanded by innumerable cysts, a finding consistent with ADPKD. Several hepatic cysts are also seen.

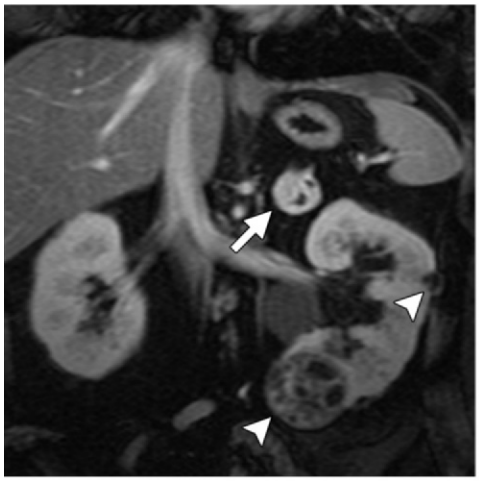
in patients with the *PKD1* mutation but were found to have a high false-negative rate in patients with the *PKD2* mutation. In 2009, Pei et al (41) published unified US criteria for diagnosis of ADPKD in families with an unknown genotype. They proposed the following criteria: *(a)* In high-risk patients aged 15–39 years, the presence of three renal cysts would confirm the diagnosis of ADPKD; *(b)* in high-risk patients aged 40–59 years, at least two cysts in each kidney would be necessary for diagnosis; and *(c)* in high-risk patients aged 60 years or older, at least four cysts in each kidney would be necessary for diagnosis.

Von Hippel–Lindau Disease

Von Hippel–Lindau disease (VHL) is an autosomal dominant disease that is linked to a tumor-



a.



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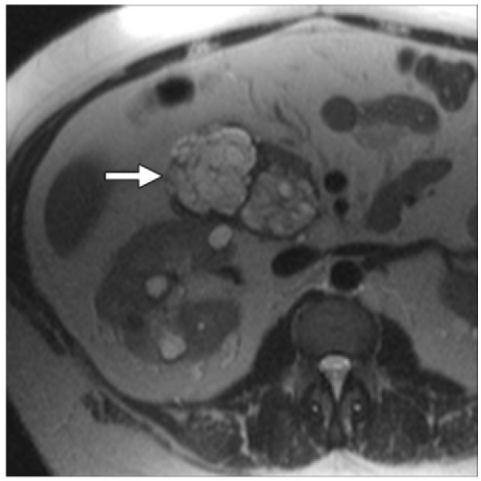


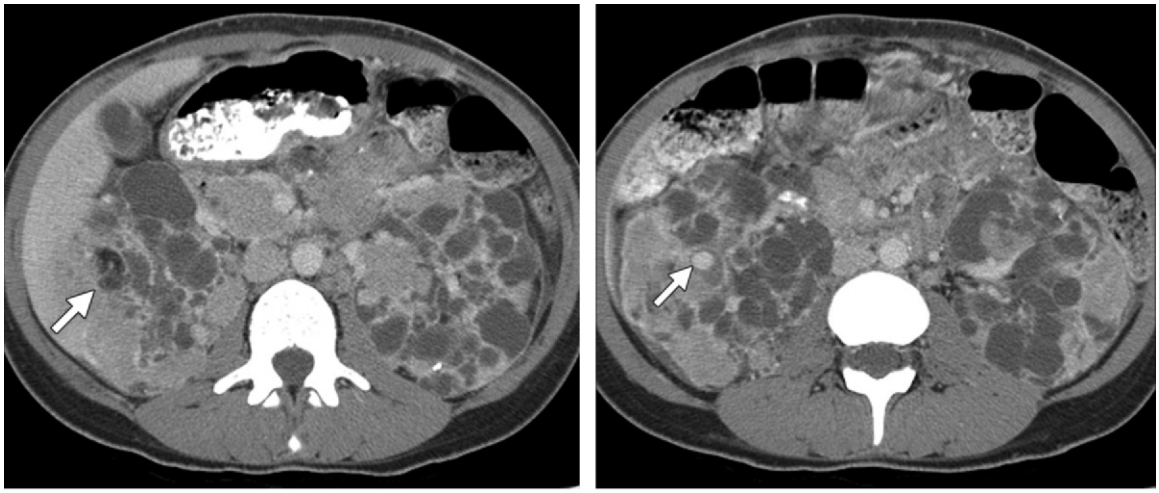
c.

Figure 20. Visceral manifestations of VHL. **(a, b)** Coronal T2-weighted MR image **(a)** and coronal contrast-enhanced fat-saturated T1-weighted gradient-recalled-echo MR image **(b)** show two complex enhancing cystic lesions (arrowheads) in the left kidney, findings compatible with cystic renal cell carcinomas. Several additional less complex cystic renal lesions are also seen. An enhancing solid and cystic lesion seen in the left adrenal gland (arrow) is a finding compatible with pheochromocytoma. **(c)** Axial T2-weighted MR image shows two cystic pancreatic lesions (arrow), findings compatible with a serous cystadenoma of the pancreas.

suppression gene on chromosome 3. It occurs in approximately one of every 31,000–53,000 births (45). Although the gene responsible for VHL has high penetrance, it has variable expression. As a result, any given individual with VHL may develop a different subset of 40 types of lesions in 14 different organs associated with the disease. The disease-defining tumor, however, is almost always a hemangioblastoma, which may occur anywhere in the central nervous system and is seen in almost all patients with the disease.

The most common visceral manifestations of VHL are renal cysts (59%–63% of patients), renal cell carcinoma (24%–45%), pheochromocytoma (0%–60%), pancreatic cysts (50%–91%), and pancreatic neoplasms such as serous cystadenomas (12%) and neuroendocrine tumors of the pancreas (5%–17%) (Fig 20). Of these, renal cell carcinoma is a significant source of mortality (45). It is thought that in patients with VHL, macroscopically normal–appearing renal tissue may contain up to 600 microscopic foci of tumor per kidney (46). However, in patients with VHL, renal cell carcinoma has been noted to be slower growing, with a higher 10-year patient survival rate, than in patients without VHL (47). In addition, metastatic renal cell carcinoma in patients with VHL is rare when the largest solid renal mass is smaller than 3 cm (47). Although some renal cell carcinomas in patients with VHL arise as de novo solid lesions, many arise in preexisting cysts. In patients with VHL, cysts that appear simple at imaging often contain small foci of renal cell carcinoma. As time progresses, these small foci may grow, forming a solid mass as the original cyst involutes. Therefore, in a patient with VHL, no cyst can be confidently dismissed at imaging as free of tumor cells (46).

Various screening and management strategies have been developed to assess for renal cell carcinoma in patients with VHL. Most are centered on following solid lesions on a 6- to 12-month basis with US, CT, or MR imaging until the largest lesion is 3 cm. At that point, nephron-sparing surgery is typically advocated to remove the largest solid lesion and as many smaller solid and cystic lesions in that kidney as possible. The idea is to "reset the clock" for the development of renal cell carcinoma in that kidney and to continue surveillance until another 3-cm solid tumor develops (47).



a. b.

Figure 21. Tuberous sclerosis. Axial contrast-enhanced CT images show the kidneys to be enlarged and replaced by numerous cysts and soft-tissue masses. In the right kidney, a focus of fat (arrow in **a**) and a small intralesional aneurysm (arrow in **b**) are findings consistent with angiomyolipomas. The right kidney was subsequently removed because of hemorrhage and was found to be replaced by cysts and angiomyolipomas. The angiomyolipomas were composed mostly of smooth muscle elements and very little fat.

Tuberous Sclerosis

Tuberous sclerosis complex is the second most common phakomatosis after neurofibromatosis type 1 and occurs in approximately one of every 6000 people (48). Tuberous sclerosis is an autosomal dominant neurocutaneous syndrome that causes hamartomas in the kidneys, lungs, liver, brain, heart, eyes, and skin. It is caused by mutations in the genes *TSC1* and *TSC2*, which encode for hamartin and tuberin, respectively (49). The most common renal manifestation of tuberous sclerosis is angiomyolipoma, which occurs in 55%–75% of patients (50). The next most commonly seen masses associated with tuberous sclerosis are simple renal cysts (Fig 21). It is unclear whether patients with tuberous sclerosis are at increased risk for renal cell carcinoma. The results of early studies that suggested an increased risk for renal cell carcinoma have been called into doubt with the subsequent identification of a malignant epithelioid form of angiomyolipoma that has a similar histologic appearance to that of renal cell carcinoma but may be differentiated at immunohistochemical staining (51,52). However, the average age of patients with tuberous sclerosis and renal cell carcinoma is 28 years, which is 25 years younger than the average age of patients with renal cell carcinoma in the general population (50).

At imaging, differentiation of lipid-poor angiomyolipoma from renal cell carcinoma is difficult. Rapid growth and the presence of calcification are two imaging findings that suggest renal cell carcinoma. However, a biopsy may be necessary for differentiation (53). As patients with tuberous sclerosis age, their risk for end-stage renal disease increases because of the mass effect of cysts and angiomyolipomas on the renal parenchyma. Angiomyolipomas also can rupture and cause substantial hemorrhage. Other common nonrenal conditions associated with tuberous sclerosis include facial angiofibromas, multiple hypopigmented macules, cortical tubers, subependymal nodules, subependymal giant cell astrocytomas, retinal nodular hamartomas, hepatic angiomyolipomas, lymphangioleiomyomatosis of the lung and retroperitoneum, and cardiac rhabdomyomas.

Conclusion

Cystic renal disease is a common entity with a wide differential diagnosis. The Bosniak classification of cystic renal disease provides a useful guide to managing cystic renal lesions. An in-depth knowledge of the various diseases that can manifest as cystic renal disease and their associated imaging findings will allow the radiologist to provide useful information to clinicians and positively affect patient care.

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