A 4-YEAR-OLD girl with no noteworthy medical history presented to the emergency department complaining of right arm pain after falling off a swing. A nondisplaced transverse fracture of the proximal right humerus was discovered on initial radiographs. On subsequent physical examination, a large abdominal mass was palpated. A plain radiograph of the abdomen demonstrated a masslike density in the left upper quadrant with inferior displacement of the transverse colon and medial displacement of the stomach bubble. Calcifications were noted on the plain radiograph. A computed tomographic scan of the abdomen was immediately obtained (Figure 1 and Figure 2).
Although usually a solid lesion, Wilms tumors can have unusual radiographic manifestations. Wilms tumor can appear as a multilocular cystic neoplasm and can occasionally mimic other cystic lesions of the kidney.1,2 Wilms tumor is a triphasic embryonic neoplasm containing epithelial, blastemal, and stromal elements. Wilms tumors can also contain various components of muscle, cartilage, bone, fibrous tissue, and even fat. Calcification seen on roentgenographic studies is uncommon. Wilms tumors have even been reported in extrarenal locations.3

Typically, Wilms tumors appear on computed tomographic (CT) scan as a rounded renal mass of low attenuation with inhomogeneous contrast enhancement. The tumor is usually solid and may appear to be separte because of fibrous stroma.2 There may be evidence of hemorrhage or cystic necrosis on CT scan in less than 10% of cases and rarely is it predominantly cystic.3 Calcifications are seen on plain x-ray film in approximately 5% of cases and on CT scan in approximately 10% to 15% of cases.2,4 These calcifications are thought to be dystrophic and may be curvilinear or amorphous in appearance radiographically.

The differential diagnosis of multilocular cystic masses of the kidney in children includes multilocular cystic nephroma, CPDN, cystic clear cell carcinoma, cystic renal cell carcinoma, multicystic dysplastic kidney, and cystic Wilms tumor.2,3 Other less common cystic-appearing entities seen in children that can mimic Wilms tumor include adult polycystic kidney disease, obstruction in a duplicated collecting system, abscess, xanthogranulomatous pyelonephritis, organizing hematoma, teratoma, and lymphangioma.2,3

Multilocular cystic renal tumors is a term used to describe cystic nephroma and CPDN.3 Cystic nephroma is a segmental, purely cystic mass characterized by multiple septations composed entirely of differentiated tissues, without blastemal elements. The CPDN is also a multiloculated lesion without nodular solid elements, but its septa contain embryonal cells. It has been proposed that cystic nephroma, CPDN, and solid Wilms tumor represent benign and malignant ends of a spectrum, similar to the way ganglioneuroma, ganglioneuroblastoma, and neuroblastoma are related to each other.3 The exact relationship between these entities, however, remains controversial.

Cystic nephroma and CPDN appear as large, solitary, sharply circumscribed masses composed entirely of fluid-filled loculi separated by thin septa. Unequivocal demonstration of solid nodular elements, which are generally seen in cystic Wilms tumors, excludes cystic nephroma and CPDN from consideration.3

Clear cell sarcoma may appear cystic secondary to hemorrhage or necrosis and may be difficult to distinguish from Wilms tumor radiographically, although they are distinct histologically. No associated somatic abnormalities have been recognized as seen with Wilms tumor. Clear cell sarcoma has a proclivity to metastasize to bone and carries a poor prognosis.3

Renal cell carcinoma is relatively rare in childhood and tends to occur in older children (mean age, 10 years).3 On CT, renal cell carcinoma is a nonenhancing solitary mass and may have areas of hemorrhage and necrosis. Calcification is seen in as many as 75% of cases.5

Multicystic dysplastic kidney is a lesion of the neonatal period, usually associated with a nonfunctioning kidney. Uncommonly, multicystic dysplastic kidney can occur in a segmental form, in which case, the focal cystic mass can appear as a multilocular cystic neoplasm.3 In summary, multilocular cystic Wilms tumor is uncommon. Differentiation from benign cystic lesions relies on identifying solid tumor nodules in the septa. In many cases, differentiation may be difficult. Therefore, Wilms tumor should be considered in the differential diagnosis of multilocular cystic lesions of the kidney.

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REFERENCES


Submissions

The Editors welcome contributions to Pathological Case of the Month, Picture of the Month, and Radiological Case of the Month. Those who wish to contribute should send their manuscripts to Dr Gilbert Barness (Pathological Case of the Month), Department of Pathology, Tampa General Hospital, University of South Florida, Davis Island, Tampa, FL 33606; Dr Tunnessen (Picture of the Month), The American Board of Pediatrics, 111 Silver Cedar Ct, Chapel Hill, NC 27514-1651; or Dr Wood (Radiological Case of the Month), Department of Radiology, Childrens Hospital Los Angeles, 4650 Sunset Blvd, Los Angeles, CA 90027. Articles and photographs accepted for publication will be acknowledged by name. There is no charge for reproduction and printing of color illustrations.