Benign Multilocular Cyst

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Benign multilocular cyst is a rare benign multicystic renal tumor. It usually involves the kidneys unilaterally. There is no generally accepted theory concerning its pathogenesis. The usual clinical presentation is an asymptomatic abdominal mass in children and nonspecific symptoms such as abdominal pain, hematuria, and urinary tract infection in adults. This report presents a case of benign multilocular cyst in an 18-months-old boy admitted with abdominal distension and a palpable mass in his right loin. The patient underwent right total nephrectomy, and histological findings were compatible with benign multilocular cyst.

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INTRODUCTION

A multilocular cystic lesion in a child's kidney may be a benign multilocular cyst, a multilocular cyst with partially differentiated Wilms tumor, a multilocular cyst with nodules of Wilms tumor, or a cystic Wilms tumor.[1] In 1956, Boggs and Kimmelstiel first described the true neoplastic nature of the lesions, proposing the term *benign multilocular cystic nephroma* for this condition.[2] In 1989, Joshi and Beckwith modified the existing terminology, emphasizing a neoplastic rather than a developmental or hamartomatous origin.[3] They defined the septa of a benign multilocular cyst composed of fibrous tissue in which well-differentiated tubules may be present, but poorly differentiated tissues and blastemal cells are not present.[3] Joshi and Beckwith suggested that the term *cystic partially differentiated nephroblastoma* be used to denote a predominantly cystic lesion and in which the septa contain blastemal or other embryonal elements. Furthermore, they proposed that both terms be used as subsets of the category term *multilocular cystic renal tumor*.[3] The origin of benign multilocular cyst is uncertain, and only about 200 cases have been reported to date.[4] Hereby, I present a case of benign multilocular cyst in an 18-month old boy with abdominal distention.

CASE REPORT

An 18-month-old boy was admitted to our center for abdominal distension and a palpable mass in his right loin. His parents mentioned that abdominal distension and vomiting had begun since the previous 2 months. He did not have any history of delivery problems and prenatal ultrasonography had revealed no abnormality. There was no fever, diarrhea, constipation, poor feeding, or urinary retention. The patient had no history of previous admission. His parents were not related and his sibling was healthy.

Upon admission, he weighted 11 kg and his height was 80 cm. On physical examination, he had abdominal distension and a palpable mass in the right upper quadrant region that extended to the right loin. Abdominal examination revealed a large, firm, well-defined, nontender mass in the right flank. His blood pressure on admission was 100/70 mm Hg. On laboratory evaluation showed hemoglobin level of 10.5 g/dL, leukocyte count of 8.6×10^9/L; and platelet count of 388×10^9/L. Serum biochemistry results were as follows: blood urea nitrogen, 7.3 mg/dL; creatinine, 0.5 mg/dL; sodium, 135 mEq/L; potassium, 4.2 mEq/L; prothrombin time, 12.5 sec; and partial prothrombin time, 35 sec. Other laboratory finding consisting of urinalysis, urine culture, and plasma renin activity were unremarkable. Abdominal radiography revealed displacement of the bowel and adjacent structures without calcification. Chest radiography revealed no abnormality. Ultrasonographic examinations

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of the abdomen showed a multiseptated cystic mass of the right kidney (Figure 1). There were multiple circumscribed sonolucent cysts that had no communication with each other or with the renal pelvis. Computed tomography showed multiple hypodense round or oval cysts with thin intervening septa. No calcification, hemorrhage, or fluid debris level were seen. No contrast enhancement was seen in the cysts, but minimal enhancement was seen in the septa. Dimercaptosuccinic acid renal scintigraphy demonstrated a defect corresponding to the renal mass in the right kidney.

Preoperative diagnosis of multilocular cystic nephroma and a cystic Wilms tumor were considered. The patient underwent a right total nephrectomy. No adhesions were found between the tumor and the surrounding tissue. The cut section showed an encapsulated mass composed of multiple discrete cysts filled with clear fluid with variable dimension. There was no solid tissue (Figure 2). Pathologic examination of a specimen from kidney biopsy showed renal tissue with numerous cysts of variable size and shape lined by single flat or cuboidal epithelial cells. In some of the cysts, lining epithelium had a distinctive "hobnail" appearance. The cellular spindle cell stroma set between the cysts had a fibroblastic nature without smooth muscle, cartilage, or immature blastematous elements (Figures 3 and 4). There was no evidence of malignancy, and scattered inflammatory cells infiltration with tubular atrophy was shown in various sections.

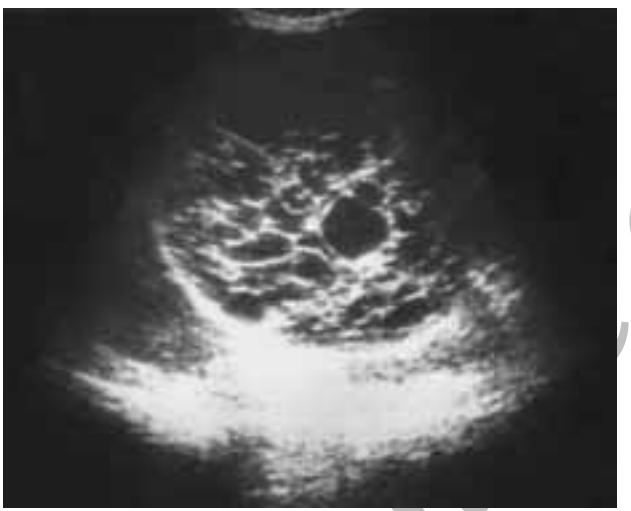

Figure 1. Ultrasonography of the abdomen shows a multiloculated cystic mass of the right kidney. The locules are noncommunicating with each other and thin septa separate them.

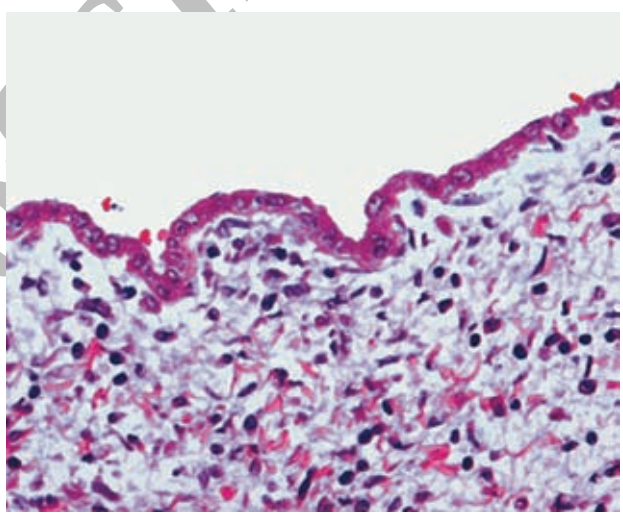

Figure 3. Cysts lined with flat-to-columnar epithelial cells occasionally producing a "hobnail" appearance due to the bulging of the nucleus toward the interior of the cyst.

Figure 2. Cut gross specimen reveals noncommunicating loculation in the right kidney.

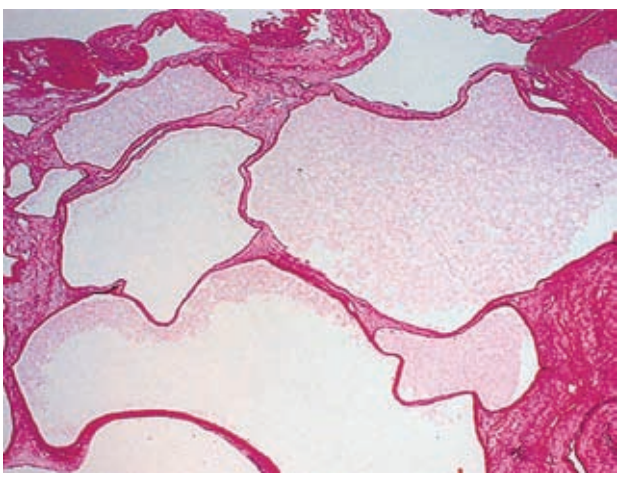

Figure 4. Multiple cystic spaces with fibrous tissue septa

This histologic finding was compatible with benign multilocular cyst.

DISCUSSION

Benign multilocular cystic renal tumor is characterized as a solitary, well-circumscribed, multiseptated mass of noncommunicating fluid-filled loculi that are surrounded by a thick fibrous capsule and compressed renal parenchyma. The most commonly reported origin of this tumor is the lower pole, although tumors have been known to originate from other areas.[1,5] Multilocular cystic renal tumors are rare. Therefore, determining the true prevalence and the age and sex distributions is difficult. Benign multilocular cyst has no racial predilection,[1] and is usually unilateral although rarely bilateral involvement have been reported.[1,3] Castillo and colleagues found that two-thirds of the tumors occurred in children aged 3 months to 2 years, with a male-to-female ratio of 2:1.[2] Madewell and coworkers found that in childhood, 73% of cases occurred in boys younger than 4 years. In patients older than 4 years, 89% of cases occurred in girls.[6]

Anatomically defined by Joshi and Beckwith, the septa of a benign multilocular cyst is composed of fibrous tissue in which well-differentiated tubules may be present, but poorly differentiated tissues and blastemal cells are not present.[3] The lesion may extend beyond the renal capsule into the renal pelvis or the perinephric space. The cyst contains clear-to-yellow fluid. Although complications are unusual, cyst herniation into the renal pelvis has been described as occurring most frequently. On gross inspection, hemorrhage and necrosis is usually not found. Calcification is also uncommon.[8]

Benign multilocular cysts in children are predominantly manifested as asymptomatic painless flank and abdominal masses, while in an adults, the most common presentations are flank mass, abdominal pain, and hematuria. Bleeding is secondary to herniation of the cyst through the transitional epithelium into the renal pelvis.[1,9] Less frequent findings include hypertension because of high renin activity and urinary traction infection.[1,8,9] On plain radiography, a soft tissue haze is seen with displacement of bowel loops and calcification is uncommon.[1,6,10,11]

Ultrasonography is the first radiologic examination performed for the evaluation of any abdominal mass. Ultrasonography can provide images necessary for diagnosing multilocular cystic nephroma. Diagnosis can be confirmed by computed tomography, and together, ultrasonography and computed tomography may be the studies of choice because they enable the evaluation of cystic lesions, stromal tissue, and the perfusion of this stroma.[1,6,12,13] Imaging findings correlate with the cyst size and the amount of stromal tissue. In most cases, a large multiloculated mass is identified. The septa highly echogenic with intravenous contrast medium due to their vascularity and calcification are rarely visible.[2] Banner and colleagues reported one case of dense calcium rings in multiple cysts.[14]

Surgery is the treatment of choice for benign multilocular cyst. The extent of surgery reported in literature has ranged from partial nephrectomy to complete excision and radical nephrectomy.[6,12,13] Although no evidence of local recurrence or metastatic disease has been reported in the literature, long-term follow-up is encouraged.[1,8]

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CONFLICT OF INTEREST

None declared.

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