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A CASE WITH A MULTILOCULAR CYST OF THE KIDNEY

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ABSTRACT

A case with a multilocular cyst of the kidney diagnosed by ultrasound and computerized tomography is documented and the related literature is reviewed. Specific pre-operative diagnosis of multilocular cyst obviates the need for extensive staging or radical surgery.

Multilocular cyst of the kidney is an uncommon lesion with less than 200 reported in the literature.[1~31] *Until the advent of cross-sectional imaging, correct preoperative differentiation from a malignant tumor could not be made with any confidence.*[5~10] We report on a case in which tomography provided evidence of distinctive morphologic characteristics of multilocular cyst that allowed a preoperative diagnosis.

CASE REPORT

A 7-month-old boy was noted to have a right abdominal mass during the course of an examination for an upper respiratory tract infection. Blood pressure on admission was 96/46 mmHg. Blood count and urinalysis were normal (Table. 1). Ultrasound scanning showed a right sonar lucent mass with numerous curvilinear internal echoes (Fig. 1). IVP showed a normal left urinary tract but a large right renal mass (Fig. 2). CT revealed a well defined mass which contained numerous low density areas irregularly separated by internal septations (Fig. 3). 99mTcDMSA renal scan showed decreased excretion of the radionuclide in the lower pole of the right kidney (Fig. 4). The diagnosis

Table 1. Laboratory findings on admission (April, 25th 1983)

CBC			Blood chemistry		
RBC		4770000/mm³	GOT		24IU/L
Hb		12.9g/dl	GPT		12IU/L
WBC		11100/mm³	LDH		426IU/L
Stab	1.0%		BUN		12mg/dl
Seg	25.0%		Creatinine		0.6mg/dl
Lymp.	63.0%		Na		140mEq/L
Mono	10.0%		K		4.1mEq/L
Eo	1.0%		Cl		110mEq/L
Plat		386000/mm³	α-fetoprotein		13ng/ml
Ret	10%				
			Serum electrophoresis		
Urinalysis			TP		6.6g/dl
Protein		(−)	Alb	75.3%	
Sugar		(−)	α_1	2.2%	
Acetone		(−)	α_2	8.0%	
Sediment			β	8.0%	
γ	6.5%				
RBC		0-1/hpf			
WBC		2-3/hpf	IgG		586mg/dl
Epi		2-3/hpf	IgA		20.7mg/dl
Cast		(−)	IgM		71.6mg/dl
Cast		(−)			
VMA		(−)			
ESR		8/1hr 22/2hr	SIgA		50μg/dl
CRP		(−)			

was multilocular cyst. Right total nephrectomy was performed.

Pathological Findings

The specimen consisted of a 210 g kideney which was mostly replaced by a well-encapsulated 10 by 6.5 by 4.5 cm mass consisting of multiple cysts. Microscopically, the cysts were lined by a flat to cuboidal epithelium supported by fibrous stroma. The stroma were composed mostly of loosely proliferated spindle to ovoid cells. However, in some areas, small tubular structures were present (Fig. 5). At the periphery of the mass, glomeruloid structures resembling fetal kidney tissue were found (Fig. 6). The surrounding renal parenchyma was normal.

Discussion

Multilocular cyst of the kidney is a unilateral well encapsulated multicystic lesion containing dysontogenic renal tissue.

The pathogenesis of multilocular cyst of the kidney has been debated for many years. *Coleman*[4] proposed that multilocular cysts were a differentiated and cystic form of

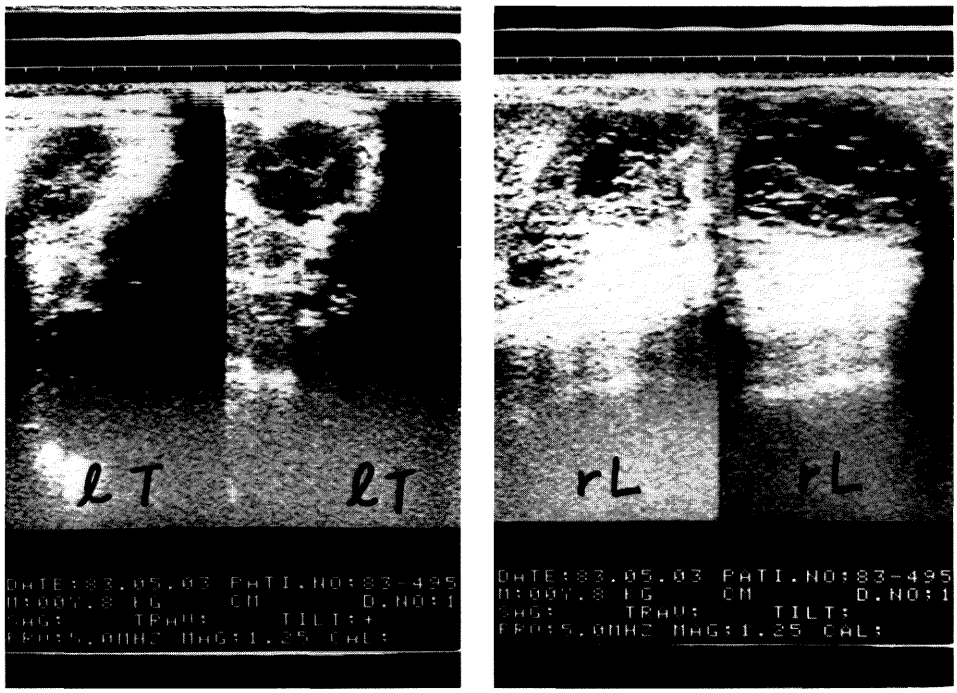

Fig. 1. Ultrasound scanning (USG). Ultrasound scanning showed a right sonar lucent mass with numerous curvilinear internal echoes.

Wilms' tumor and he defined three possibly interrelated lesions: (1) Wilms' tumors showing cystic differentiation; (2) Lesions macroscopically indistinguishable from multilocular cyst containing variably differentiated nephroblastomatous foci. All such cases have been described in infants; (3) Typical multilocular renal cysts, which have been described in children and adults.

Akhtar[1] on the other hand, divided cases of multilocular cysts in pediatric age groups into two subgroups base on the presence or absence of embryonic tissue within the cysts. The first group includes multilocular cysts, which are entirely composed of epitherial lined locules separated by fibrous septa. The second group includes multilocular cysts which are similar to those comprising the first group in every other aspect, except for the fact that these contain variable amounts of organoid embryonic tissue. The cysts included in the second group are unique to the pediatric age group. He described the prognosis of the cysts comprising the first group as identical to that of cysts in the second group.

Multilocular cysts occur from three months of age to adulthood. *There is no family history of renal cystic disease or apparent predilection for race.*[3~5] Males are almost always under four years of age.

The presenting symptoms vary with the patient's age.[4,5,11,13~17] *The most common symptom in children is a nonpainful abdominal mass.*[12] Our case was noted to have a

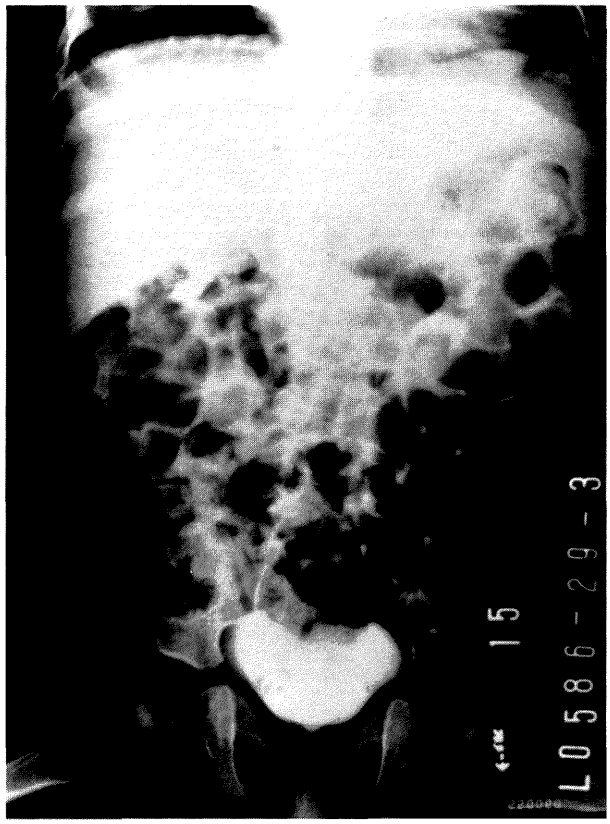

Fig. 2. Excretory urogram (IVP). IVP showed a normal left urinary tract, but a large right renal mass.

right abdominal mass during the course of an examination for an upper respiratory tract infection. In the adult the most common presentations are abdominal pain and hematuria.

Plain radiographs of the abdomen show a mass. *Calcifilation is uncommon and is usually seen in older patients.*[13,16]

The excretory urogram demonstrates a well-defined, intrarenal mass in a normally functioning kidney.[3]

On angiographic examination, multilocular cyst shows no specific features. *These nonspecific angiographic features do not allow preoperative differentiation of multilocular cyst from renal cell carcinoma or nephroblastoma.*[8]

The sonogram will demonstrate a cluster of echo-free masses separated by intense echoes, when the cysts are large enough.[19,20] When the locules are small, the sonogram will demonstrate a nonspecific, complex internal masses.

Multilocular cyst appears on CT scans as a well marginated, rounded, or polycystic cortical mass.[13,20] The septa are enhanced by intravenous contrast medium due to their

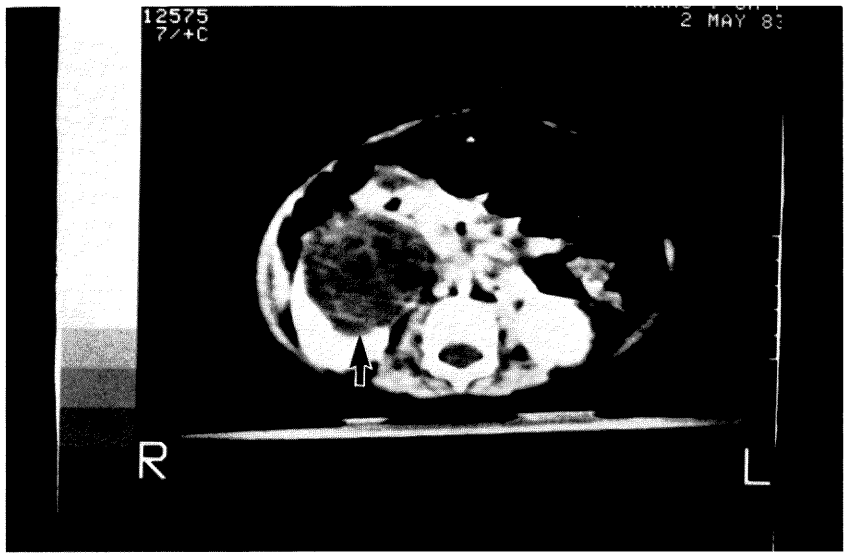

Fig. 3. Computerized tomography (CT). CT revealed a well difined mass which contained numerous low density areas irregularly separated by internal septations.

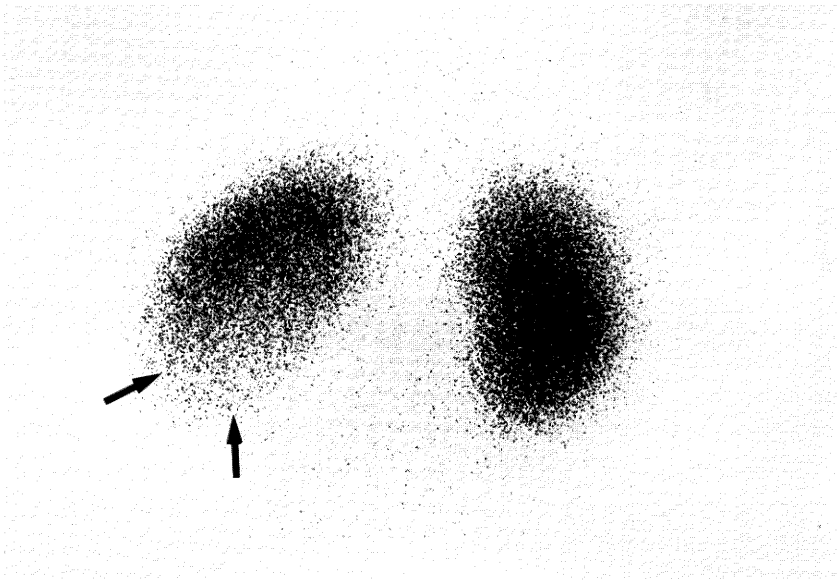

Fig. 4. 99mTc DMSA renal scan. 99mTc renal scan showed excretion of the radionuclide in the lower pole of the right kidney.

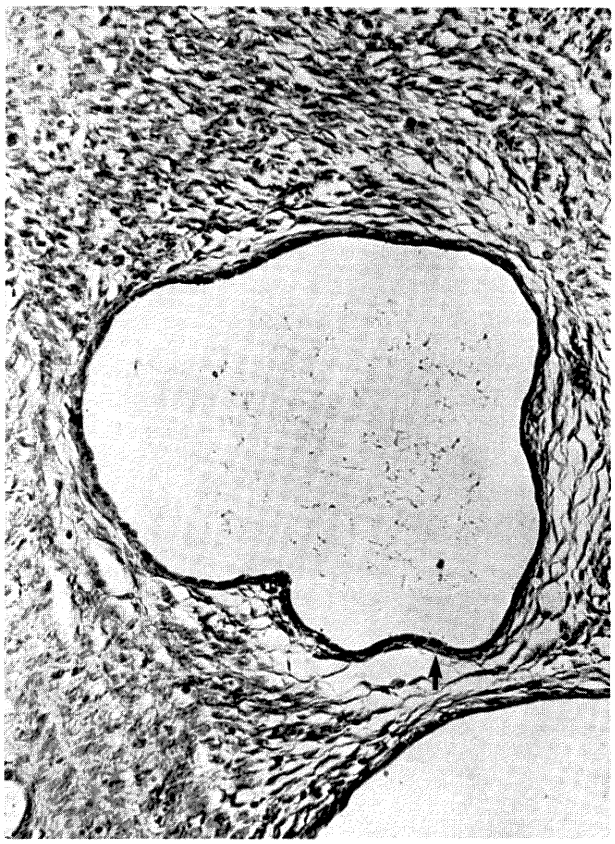

Fig. 5. Microcysts lined by flat to cuboidal epithelium in the multilocular cyst.

vascularity. The locules are not enhanced. When the locules are small or filled with thick mixomatous material, the CT appearance is indistinguishable from other solid masses.

Nuclear medicine renal scans show decreased excretion of the radionuclide in the area of the mass.

In our case, the sonogram demonstrated right sonar lucent mass with numerous curvilinear internal echoes (Fig. 1). IVP showed a normal left urinary tract but a large right renal mass (Fig. 2). CT revealed a well defined mass which contained numerous low density areas irregularly separated by internal septations (Fig. 3). 99mDMSA renal scan showed decreased excretion of the radionuclude in the lower pole of the right kidney (Fig. 4).

There are no reported cases of multilocular cyst of the kidney associated with cystic changes in other organs,[4] *or with congenital anomalies in the urinary tract or other systems.*[3]

Multilocular cysts are seen on either side with approximately equal frequency and

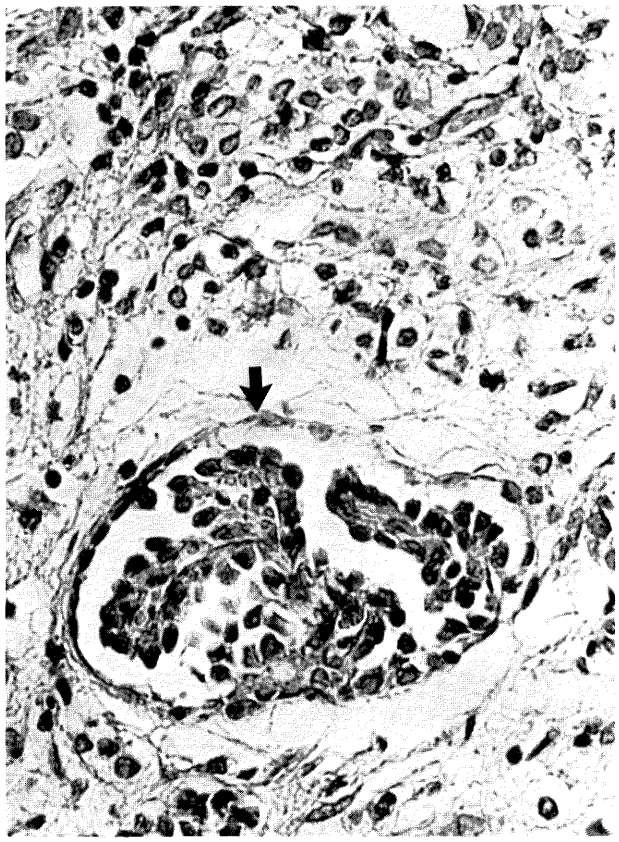

Fig. 6. Glomeruloid structures found in the multilocular cyst.

occasionally involvement will be bilateral. Although they may arise anywhere within the renal parenchyma, origin from the lower pole is most common. Our case was located in the lower pole of the right kidney.

On gross examination the multilocular cyst is well encapsulated and it frequently compresses the adjacent renal parenchyma. The masses vary in size from about 3×4 cm to 18×34 cm. Rates of growth may be quite slow or very rapid. Recurrent growth after local excision appears to be related to inadequate excision.

Criteria for the diagnosis of multilocular cysts were established in 1951 by *Powel* et al. In 1956, *Boggs and Kimmelstiel*[31] suggested certain modifications and applied the follwing criteria: (1) The lesion must be multilocular; (2) the cysts must, for the most part, be lined by epithelium. (3) the cysts must not communicate with the pelvis, (4) the residual tissue should be essentially normal, except for pressure atrophy; (5) fully developed, mature nephra or portions of such should not be present within the septa of the cystic lesion. The last criterion recognizes that in some cases, variably developed tubular structures are present within the septa of multilocular cysts, and sometimes

immature or embryonal elements including glomeruloid structures, may also be found. Our case satisfied all of these criteria.

The differential diagnosis of multilocular cyst of kidney is age dependent. *In children, multiloclar cysts are most often misdiagnosed as Wilms' tumor, hamartoma, or multicystic and polycystic kidneys.*[13]

Multilocular cyst is most frequently treated by nephrectomy or occasionally by local excision.[7,8,11,15] *Local resection is the prefered method of treatment. Metastasis is uncommon.*[3] If there is a significantly large component of nephroblastoma or sarcomatous stroma the patient should be followed closely for local reccurence or distant metastasis. The presence of embryonic tissue does not warrant postoperative radiation therapy or chemotherapy. *Akhtar*[1] designated these cases multilocular cysts with embryonic tissue and reported that the presence of embryonic renal tissue within the multilocular cysts does not adversely affect their prognosis. Our case was deemed to be one of the multilocular cysts comprising Akhtar's second group and postoperative radiotherapy or chemotherapy were not necessary. In rare cases, the multilocular cyst may be malignant and may also later involeve the contralateral kidney. We recommended that our case be closely followed for at least two years.

Conclusion

We report a case with multilocular cyst of the right kidney in a child diagnosed by ultrasound and computerized tomography. Right total nephrectomy was performed. Microscopically, embronic tissue was present and the case was considered to be one of the multilocular cysts comprising Akhtar's second group. We recommended that the case be followed closely for local recuurrence or distant metatasis.

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