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Quiz case of the month

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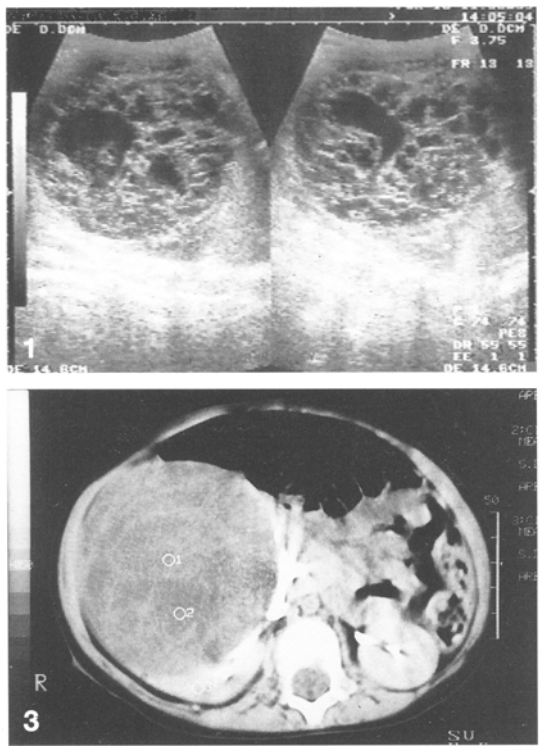
Case report

A 1-year-old female with a palpable mass in her right upper quadrant was presented. The size of the mass had increased with time. An abdominal US, color-Doppler imaging, excretory urogram, and abdominal CT were performed.

Ultrasound showed multiple circumscribed sonolucent cysts that had no connection with each other and renal pelvis, and none exceeded 3.1 cm in diameter. The average thickness of the septations between the cysts was approximately 5 mm. The total dimension of the mass was $9 \times 10 \times 8$ cm (Fig. 1).

Color-Doppler imaging showed no flow at the capsule and the septa. Excretory urogram showed soft tissue density in right renal region, delayed excretion of contrast medium, minimal pelvicaliectasis, and displacement of the ureter medially. The mass showed no calcification (Fig. 2).

Computed tomography showed that a multilocular, well-defined renal mass had a CT number that was slightly greater than that of water density. No contrast enhancement was observed in the cysts, but very slightly in the septa (Fig. 3). The renal parenchyma outside the lesion was normal, even though it was stretched.



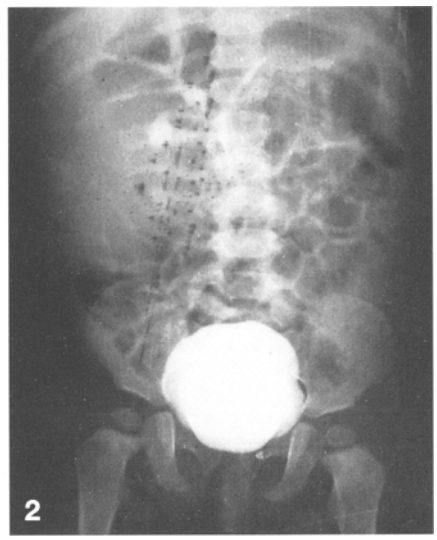


Fig. 1. Multiple circumscribed sonolucent cysts that have no connection with each other and pelvis are depicted at US

Fig. 2. On excretory urogram soft tissue density is seen in right renal region. Renal pelvis is quite dilated. Left kidney is not seen because of gas distention

Fig. 3. Multiple hypodense oval or round cysts and septa are seen at CT. There is no contrast enhancement in the cysts, but slightly in the septa. Enhancing renal parenchyma is seen medial and posterior to the mass

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Diagnosis

The pathological diagnosis was of a multilocular cystic nephroma (MLCN).

Discussion

The MLCN was first described in 1892 by Edmunds. He called the lesion "cystadenoma of the kidney" [1]. Since then, there have been less than 102 cases reported in the literature [1–6]. It is an uncommon, nonfamilial, and benign renal neoplasm, characterized by a well-circumscribed encapsulated mass that contains multiple noncommunicating fluid-filled spaces lined by epithelium and separated by a distinct stroma.

The tumor generally appears as an abdominal mass in children [1, 4]. Because of its rarity and clinical presentation, it is often thought to be nephroblastoma. In childhood, 73 % of cases appear in boys under 4 years of age. Over 4 years of age, 89 % of cases are female and present as an abdominal mass [1, 5, 7].

The MLCNs are generally unilateral, and seen on either side in approximately equal frequency, but origin from the lower pole is the most common location as in this case [1, 4]. On plain radiography, usually a mass lesion is detected. Calcification is uncommon, but may be seen in older patients as curvilinear and peripheric localization [1, 4, 7].

On excretory urogram, kidneys function normally with MLCN. Depending on the localization of the tumor, displacement of other structures, extension of the tumor to the renal pelvis, and delayed excretion or no excretion is demonstrated

Ultrasound findings depend on the amount of stroma and size of the locules. When the cysts are large, US demonstrates anechoic fluid-filled spaces separated by hyperechoic septa. This pattern is suggestive of multilocular cystic nephroma [4]. However, when the locules are small, sonography demonstrates echogenic solid mass [1, 7].

On angiographic examination, MLCN is generally hypovascular, but it may be avascular or hypervascular. Thus, angiographic features are not specific for differential diagnosis, but may be helpful in preoperative vascular anatomy evaluation [4].

With color-Doppler US, it is possible to evaluate lesion vascularity noninvasively. Tumor vascularity in malignant renal lesions gives rise to abnormally high-velocity Doppler-shifted signals [8–10]. We think these findings may be helpful in differential diagnosis, but needs further evaluation in large benign renal lesions series.

As in US, CT findings change with cyst size and amount of stromal tissue [4]. In most cases a large multilocular mass is identified. The septa among the locules enhance with contrast medium due to their vascularity. The cystic spaces do not enhance and have CT numbers that are slightly greater than the CT numbers of water.

Multicystic dysplastic kidney, cystic partially differentiated nephroblastoma, malignant necrotic and hemorrhagic masses (renal cell carcinoma) and Wilms' tumor, and cystic mesoblastic nephroma should be included in differential diagnosis. Multicystic dysplastic kidney shows no function on excretory urogram and is characterized by multiple noncommunicating cysts of variable size. Contralateral genitourinary abnormalities, such as vesicoureteral reflux and ureteropelvic-junction obstruction, are seen in up to 41 % of patients [7]. Multicystic dysplastic kidney is generally detected in the newborn period and antenatally.

The differential diagnosis with cystic partially differentiated nephroblastoma is not possible based on imaging findings. The only differentiation between these two lesions is histological [2]. Cystic partially differentiated nephroblastoma contains blastema [7].

Wilms' tumor is the most common renal neoplasm in childhood (mean age of presentation 3 years) and shows pelvicaliceal deformation on excretory urogram. Cystic necrosis is seen on CT scans and US images in less than 10 % of cases. It is a rare predominantly cystic [7].

Renal cell carcinoma is rare in children and therefore does not pose a problem in that age group. When it occurs, hemorrhagic and necrotic degeneration in these tumors may mimic MLCN.

Mesoblastic nephroma is congenital and the most common renal neoplasm in infancy. The cellular subtype of mesoblastic nephroma tends to be larger and cystic because of necrosis and hemorrhage [7].

References

1. John EM, Stanford MG, Charles JD Jr, David SH (1983) Multilocular cystic nephroma: a radiographic pathologic correlation of 58 patients. Radiology 146: 309–321
2. Vijay VJ, Bruce B (1989) Multilocular cyst of the kidney (cystic nephroma) and cystic partially differentiated nephroblastoma. Terminology and criteria for diagnosis. Cancer 64: 466–479
3. Dikengil A, Benson M, Sanders L, Newhouse JH (1988) MRI of multilocular cystic nephroma. Urol Radiol 10: 95–99
4. Banner MP, Pollack HM, Chatten J, Witzleben C (1981) Multilocular renal cysts: radiologic–pathologic correlation. Am J Roentgenol 136: 239–247
5. Carlson DH, Carlson D, Simon H (1978) Benign multilocular cystic nephroma. Am J Roentgenol 131: 621–625
6. Abara OE, Liu P, Churhill BM et al. (1990) Magnetic resonance imaging of cystic partially differentiated nephroblastoma. Urology 38: 424–427
7. Agrons GA, Wagner BJ, Davidson AJ, Suarez ES (1995) Multilocular cystic renal tumor in children: radiologic–pathologic correlation. Radiographics 15: 653–669
8. Isabel MR, Kenneth JWT, Ruben K, Peter NB, Daniel PS, Darrly C (1988) Tumor vascular signals in renal masses: detection with Doppler US. Radiology 168: 633–637
9. Ruben K, Kenneth JWT, Ann LF, Isabel MR (1990) Renal masses: characterization with Doppler US. Radiology 176: 703–707
10. Dirkjan K, Rob J (1989) Renal masses: differential diagnosis with pulsed Doppler US. Radiology 170: 59–60