CASE REPORT

doi: 10.4274/jus.81
Journal of Urological Surgery, 2014; 1: 39-41

Pediatric Multilocular Cystic Nephroma Extending into the Renal Pelvis and Ureter

Çocuk Hastada Renal Pelvis ve Üretere Uzanan Multilokuler Kistik Nefroma

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ABSTRACT

Multilocular cystic nephroma (MCN) is a rare tumor at the most benign end of the spectrum of the multilocular cystic neplasms of kidney. Nephrectomy is curative for MCN. In this case-report, we present a 16-month-old girl with a 10x15 cm multilocular cystic renal tumor extending into the renal pelvis and proximal ureter on the right side demonstrated on magnetic resonance imaging. Nephrectomy was performed. The pathology was completely consistent with MCN.

Key Words

Pediatric, kidney, cyst, multilocular cystic nephroma, nephrectomy

ÖZET

Multiloküler kistik nefroma (MKN), böbreğin multiloküler kistik neoplazmları içerisinde spektrumun en iyi huylu tarafında yer alan nadir bir tümördür. Nefektomi küratif olmaktadır. Bu olgu sunumunda, 16 aylık bir kız çocuğunda manyetik rezonans görüntüleme yöntemi ile tanısı konulan, sağ böbreğin renal pelvisinden proksimal üretere kadar uzanan 10x15 cm'lik multiloküler kistik böbrek tümörü sunulmaktadır. Hastada nefrektomi uygulanmış olup patolojik tanısı tamamen MKN ile uyumlu gelmiştir.

Anahtar Kelimeler

Çocuk, böbrek, kist, multiloküler kistik nefroma, nefrektomi

Introduction

Multilocular cystic nephroma (MCN) is a very rare neoplasm of kidney. It was first described in 1892 and within 100 years Castillo and colleagues collected about 200 cases (1,2). It constitutes less than 1% of the patient population even in a very large international Wilms' tumor (WT) trial (3). This case report aims to present clinical, radiological and histological features of a patient with a final diagnosis of MCN.

Case Presentation

A 16 month-old girl was referred to our institute with a non-tender palpable flank mass on her right side. The parents mentioned that the mass became palpable within the last 2 months. The patient was unable to gain weight and had difficulty in feeding. Physical examination revealed a palpable mass filling the right upper quadrant and expanding to the right lower quadrant. Blood pressure was normal.

Prior to referral, patient underwent ultrasonography (US) and computed tomographic (CT) scan. The US was repeated and magnetic resonance (MR) imaging was performed to further delineate the anatomic details in our hospital. US showed a well-circumscribed, 10x15x10 cm mass consisting of multiple anechoic cysts separated by thin echogenic septa and there was no vascular involvement. On CT and MR imaging, the mass replaced most of the renal parenchyma of the upper pole and herniating into the renal pelvis, some calyces of the lower pole and the proximal ureter with moderate dilatation of the renal collecting system (Figure 1). The capsule and internal septa of the mass were enhanced with contrast medium; no solid components or nodules were seen within the lesion. The remaining

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renal parenchyma excreted the contrast material symmetrically with the normal left kidney. The radiological findings were consistent with multilocular cystic renal tumor. The urinalysis showed microscopic hematuria, leukocyturia and proteinuria. She had a hemoglobin level of 9.3 g/dl where the serum creatinine was 0.4 mg/dl. Coagulation parameters and chest X-ray was normal. Serum lactate dehydrogenase was normal and tests for hydatid cyst were negative.

The operation was performed in a 45 degrees lateral decubitis position with a slight extension. The incision was a transverse lumbal incision starting from 2 cm. cranial to umbilicus and extending laterally. The mass was mobile and no significant adhesion was present. The mass was originating from the right kidney and was compressing it to iliac fossa. Access to the kidney has been possible after the birth of the mass outside through the incision and the tumor was taken out en-block with kidney (Figure 2). There were six hilar lymph nodes which were smooth and soft with palpation. After local hemostasis, a hemovac drain was placed to the operation area and layers were closed regardingly. The postoperative course was uneventful and the patient was discharged at postoperative fourth day.

The pathological evaluation revealed a diagnosis of MCN which shows multiple cysts in different sizes with fibrous tissue septa (Figure 3). The cyst walls were comprised of single layered flattened benign cuboidal epithelium, neither a blastema nor atypical epithelial cells were found in the septa between the cysts. All the lymph nodes were negative.

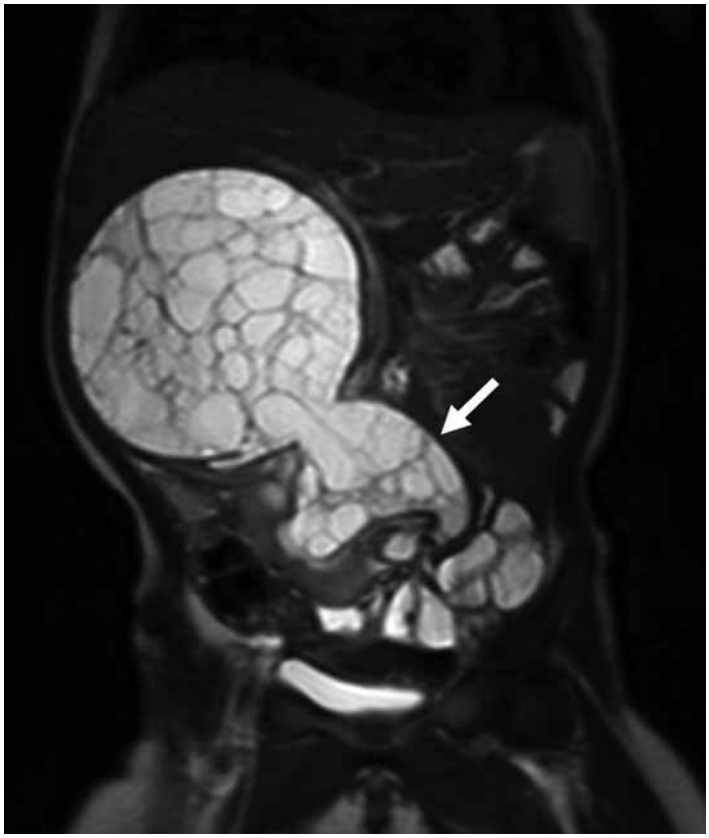

Figure 1. Coronal T2-weighted MR image demonstrates the mass extending into the dilated renal pelvis and proximal ureter (arrow)

Discussion

The differential diagnosis of cystic renal lesions includes several benign and malign pathologies. Multicystic dysplastic kindey (MCDK) is the most common cause amongst these pathologies. However, most of the cases are detected prenatally and the patient had no positive history in this aspect. In MCDK, which results from ureteral atresia, the kidney has a shape of 'bunch of grapes' and radiologically there is no significant parenchyme around the cysts (4). In our patient, there was a significant amount of renal parenchyma which was compressed to iliac fossa by the mass effect of the giant multilocular cyst. Additionally, it is extremely rare that a MCDK grows to a size which requires nephrectomy. It was shown that about 1% of MCDK gets larger after a 5 years period of follow-up (5).

Wilms' tumor (WT) is another renal pathology which should be considered in a young child with a palpable renal mass with a cystic structure. Less

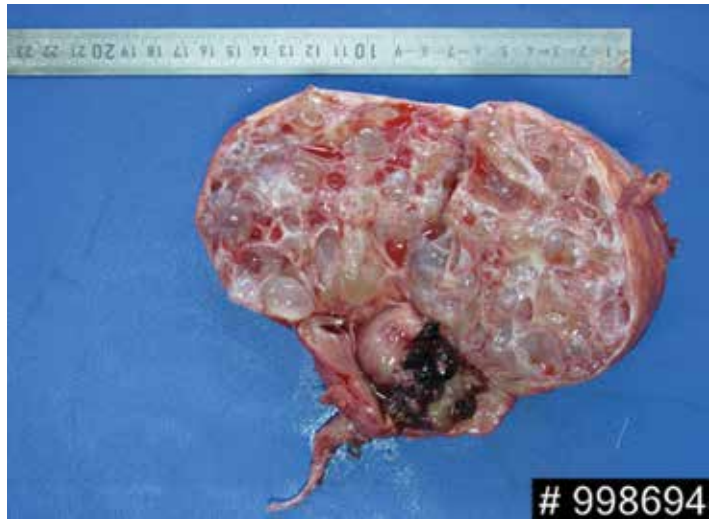

Figure 2. Macroscopic appearance of the tumor which is composed of multiple cysts in different sizes those completely filling the renal cortex and medulla and extending into the renal pelvis and proximal ureter

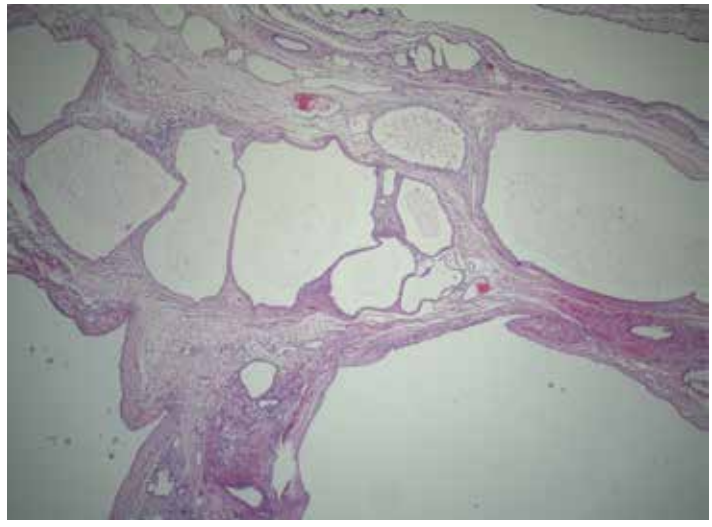

Figure 3. Microscopic appearance of the tumor which is composed of cysts in different sizes separated with fibrous septa. Cysts are lined by single layered flattened benign cuboidal cells

than 10% of WT has cystic component in imaging tests. However, both radiologic tests and gross appearance show a presence of necrotic tumor with multiple cystic spaces filled with hemorrhage (6). Therefore, our preoperative diagnosis was far away from cystic WT. The other less probable pathologies for differential diagnosis can be counted as clear cell sarcoma, mesoblastic nephroma and renal cell carcinoma those none have a clear cystic fluid neither by gross appearance nor radiologically (6).

The most probable diagnosis in a child with such a palpable mass originating from kidney with the mentioned features are MCN and cystic partially differentiated nephroblastoma (CPDN). Despite the controversies, CPDN may be regarded as a transition point between MCN and cystic WT, the benign and malign ends of the spectrum (6). MCN and CPDN are indistinguishable by means of radiologic tests and gross appearance (6). The distinction is made by histological examination. In MCN, the cysts are lined by flattened, cuboidal, or hobnail epithelium. The septa consist of fibrous tissue, which may contain well-differentiated renal tubules but no poorly differentiated tissues or blastemal cells. CPDN, on the other hand, has the histologic features of cystic nephroma with the addition of septa blastemal cells in any amount, with or without other embryonal stroma or epithelial cell types (4). In our case, the clinical and histologic findings were fully consistent with Powell's criteria for MCN: 1) unilateral involvement, 2) solitary lesion, 3) multilocular lesion, 4) noncommunication with the renal pelvis, 5) noncommunication of cysts with each other, 6) loculi lined by epithelium, 7) intralocular septa devoid of renal parenchyma, and 8) if residual renal tissue is present it should be normal (7). Two thirds of multilocular cystic renal tumors are mostly detected under 2 years age and equally distributed between MCN and CPDN. In this age group a male predominance of 2:1 is present and the palpable mass in the flank region is the most common finding (6).

Nephrectomy is always curative for MCN if totally excised. There are some reports on the use of partial nehrectomy in children with MCN. However, partial nephrectomy may be indicated in bilateral cases which is absolutely a rare occurrence and when radiographic evaluation indicates a well-demarcated lesion with thin-walled cysts and clear fluid permitting to preserve a significant renal parenchyma (2,8,9). Herniation of parts of the cystic mass into the renal collecting system is not uncommon which can also explain the microscopic hematuria as in the present patient [10,11,12,13,14]. However, in our case, although preoperative imagings revealed a multilocular cystic renal lesion and a completely normal contralateral kidney, it was not feasible to perform a partial nehprectomy because of the herniation of the cysts to renal pelvis and proximal ureter and nephrectomy has been performed regarding the concerns about leaving residual tumor.

Conclusion

Multilocular cystic nephroma is a rare disease of childhood. The patients are mostly diagnosed with a palpable flank mass and approach should be based on a preoperative proper radiological and clinical evaluation. Although, the mass can get very huge volumes, the surgery is not difficult. Although, partial nephrectomy can be an option for selected cases, nephrectomy is always curative.

Conflicts of Interest

There are no conflicts of interest.

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