Multicystic Renal Cell Carcinoma

A Rare kidney Tumor in Children

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INTRODUCTION

Renal cell carcinoma (RCC) is the most common (85%) primary malignancy of the renal parenchyma, and accounts for about 3% of all adult neoplasms.(1) It is well known that kidney tumors are rarely seen in pediatric age in contrast to adults. Until today, only about 450 patients with RCC under 21 years of age have been reported.(2) Multicystic RCC (MCRCC) is an extremely rarely seen variety of kidney tumors. Here, we report a case of MCRCC in a pediatric patient. To the best of our knowledge, this is the second reported case of MCRCC in a child in English literature.

CASE REPORT

A 14-year-old male child presented with a history of flank and abdominal pain, macroscopic hematuria, and abdominal mass for about two months. During the past two weeks, these symptoms had worsened. On physical examination, right costovertebral angle tenderness and palpable mass were detected, and systemic examination was unremarkable. Past medical history was normal, and there were no findings related to tuberous sclerosis or von Hippel-Lindau disease.

Routine biochemical and hematologic studies were within normal ranges. Ultrasonography demonstrated a lesion which had solid and cystic components localized in the mid and the lower pole of the right kidney. Computed tomography (CT) scan confirmed the observation of the ultrasonography, and revealed a heterogeneous lesion in the lower pole of the right kidney with hydronephrosis (Figure 1). There was no invasion into the perirenal tissues. Other intraperitoneal organs and contralateral retroperitoneal region were normal. Intrave-

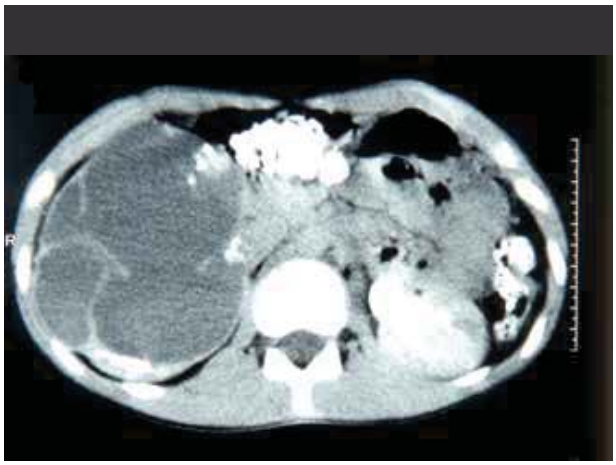

Figure 1. Computed tomography showing a heterogeneous cystic lesion in the lower pole of the right kidney.

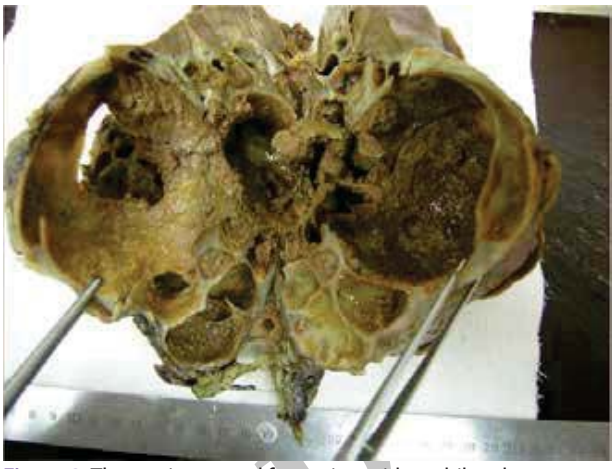

Figure 2. The cystic tumoral formation with multilocular appearance is seen in macroscopic section. The normal kidney tissue is seen in a small area in the upper part.

nous pyelography revealed a right hydronephrotic kidney with nonfunctional mid and lower segments.

With the pre-operative diagnosis of hydronephrosis and a complicated cyst, exploration was performed. During operation, it was noted that Gerota's fascia and perirenal tissues were very tight and adherent. Therefore, we performed radical nephrectomy with regional lymphadenectomy. Gross appearance of the nephrectomy specimen was as follows; the specimen consisted of one kidney and attached perirenal fat measuring 18 × 16 × 12 cm. There were prominent distortions in renal contours. Sectioning revealed multilocular cystic lesion, and lumen of cystic cavities were filled with necrotic, granular material. The uninvolved normal renal tissue was noticed as a narrow area at one side of the multilocular cystic lesion (Figure 2).

Cystic renal cell carcinoma was diagnosed on the basis of microscopic and macroscopic findings (Figures 3 and 4). According to Fuhrman nuclear grading scheme, the nuclear grade was 2. Tumor was restricted since intrarenal and capsular invasion were not observed. Two months after the surgery, the patient was alive without any recurrence of the tumor.

DISCUSSION

Childhood cancers are much less common than adult cancers. Leukemia and brain and spinal cord tumors are the most common type of cancers found in children. Other infrequent childhood cancers include retinoblastoma, Wilms' tumor, muscle or bone cancers, lymphoma, and RCC. In the literature, it is shown that RCC corresponds to 1.4% of renal tumors in patients under 4 years, 15.2% between 5 and 9 years, and 52.6% between 10 and 15 years old.[3]

Although RCCs may include cystic or solid structures, cystic forms constitute only about 4% to 10% of all RCCs. However, solitary cystic RCC is extremely rare in adults.[4] The average age of the children with RCC changes between 10.6 and 14 years. In pediatric kidney tumors, male-to-female ratio has been reported between 1 and 3/1.

Abdominal pain, hematuria, and fever are the most common symptoms of CRCC. Even if palpable mass occurs in 38%, hematuria in 38%, and abdominal pain in 50% of cases, classic symptom triad can be determined in only 6% of patients presented with CRCC.[5]

Multilocular CRCC is also known as multilocular clear cell RCC, which constitutes 1% to 4% of RCCs.[6] Multilocular CRCC was accepted as a different subtype of clear cell RCC in the World Health Organization (WHO) classification (2004). Comparison between a multilocular CRCC and a solid renal tumor has shown that these cystic tumors contain fewer malignant cells (less than 25%), which may proliferate at a slower rate.[4]

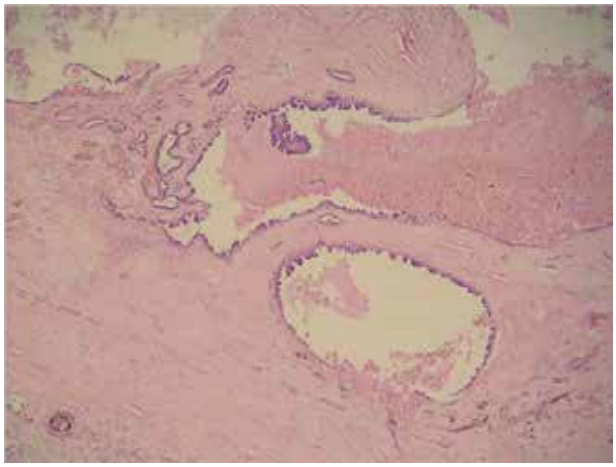

Figure 3. Histolological sections revealed multiple cysts seperated by fibrous hypocellular septa. Small papillary structures are seen among cystic areas. The cysts are filled with necrotic material (Hematoxylin and Eosin stain, ×10).

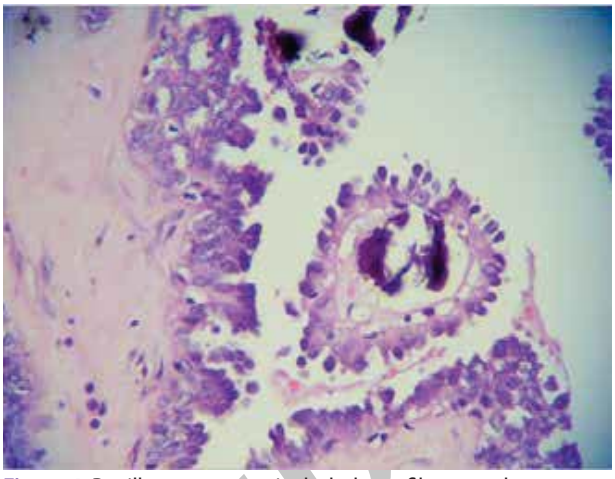

Figure 4. Papillary structures included true fibrovascular cores, and lining cells had mild nuclear pleomorphism and hyperchromasia. Some of the atypical cells showed eosinophilic cytoplasm. Psammomatous calcification and the appearence of atypical cells in high magnification (Hematoxylin and Eosin stain, ×30).

Multicystic RCC is also an unusual entity in children and to the best of our knowledge, only one such case has been reported previously in the literature.[7] Because of the rarity of these neoplasms, their exact characteristics, including pathologic features, natural history, clinical behavior, and prognosis remain uncertain. There are no treatment protocols agreed upon for pediatric MCRCC due to limited knowledge. Most reports include management of adolescent or adult patients in the literature.[8] These tumors are not radiosensitive, and chemotherapy results have been ineffective. However, ideal treatment consists of radical nephrectomy.

Since cases are mostly with relatively low Fuhrman grade, stage, and average tumor size less than 5 cm at diagnosis, it seems that MCRCC has better prognosis than non cystic RCC.[9] Owing to the favorable prognosis with a 5-year disease-free interval close to 100%, nephron-sparing surgery should be performed, especially in younger patients and in smaller and exophytic tumors, provided this is technically feasible intra-operatively.[10] Although it is specified in the literature that MCRCC has better prognosis, malignant potential of this disease should be considered and management should be handled according to this point.

To conclude, we can say that because MCRCC of the kidney is extremely rarely seen in pediatric ages, it must be taken into account in the differential diagnosis of the kidney tumors in childhood, especially when the radiological studies show a multicystic heterogeneous lesion.

CONFLICT OF INTEREST

None declared.

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