Case report

Cystic nephroma of childhood: A case report and review of the literature

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A B S T R A C T

Cystic nephroma of childhood is a rare renal tumor. It is generally considered as a benign end of the spectrum with cystic partially differentiated nephroblastoma and nephroblastoma (Wilms tumor). Most of the cases are unilateral and sporadic. However, bilateral and familial cases have also been reported. The clinical presentation is usually as an asymptomatic abdominal mass. It is almost impossible to differentiate cystic nephroma from cystically partially differentiated nephroblastoma or other cystic renal tumor on radiological studies. The final diagnosis depends on the histopathologic examination. We present a case of an 8-month-old female baby with a left renal tumor. The patient accepted left total nephrectomy after physical examinations and radiological studies. Microscopically, the tumor was composed of variable-sized cysts separated by fibrous septa without blastemal elements. A diagnosis of cystic nephroma was made.

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1. Introduction

Cystic nephroma of childhood is defined as a renal tumor composed of multilocular cysts without blastemal or undifferentiated elements. It is considered to be related to cystically partially differentiated nephroblastoma and nephroblastoma (Wilms tumor). The clinical significance is its benign nature, which should be differentiated from other renal neoplasms of children. In this article, we present a case of a solitary cystic nephroma in a female baby and review the clinical presentation, radiological findings, and histopathologic differential diagnosis.

2. Case Report

An 8-month-old female baby was found to have an abdominal mass by her parents for about 1 week. No other symptoms such as vomiting, diarrhea, fever, or hematuria were observed. She was brought to the Department of Pediatrics of our hospital for management in August 2013. Physical examination showed a left-sided abdominal mass. Ultrasonographic examination revealed that it was composed of multiple anechoic spaces separated by hyperechoic septa. The following computed tomography examination revealed a demarcated, multilocular, and hypodense mass arising from the upper pole of the left kidney (Fig. 1). No separated nodule or other structural abnormality was found. Considering the large size of the tumor and the possibility of malignancy, total nephrectomy through anterior subcostal incision was performed.

Grossly, the tumor was well circumscribed with a smooth surface. It was located at the upper pole of the left kidney and measured $9.4 \times 8.5 \times 7.4$ cm$^3$ in size. The cut surface revealed variable-sized cysts separated by thin intervening septa. The cysts contained clear fluid. No solid component was found in it. The tumor did not communicate with the renal pelvis. The remaining renal parenchyma was compressed. Microscopically, the tumor was composed of multilocular and variable-sized cysts separated by fibrous septa (Fig. 2) and lined by a single row of low cuboid or flattened cells (Fig. 3). The epithelial cell lining had relatively uniform nuclei with a focal hobnail appearance (Fig. 4). No blastemal element was found in the septa. The renal parenchyma revealed normal development. A final diagnosis of left kidney cystic nephroma was made.

3. Discussion

Cystic nephroma is a rare, benign, renal tumor. Edmunds first reported this tumor in 1892 as “cystic adenoma of the kidney”. Cystic nephroma has two incidence peaks, one in the childhood and...
the other in the adult population. Most of the childhood cases have been recorded in children between 3 months and 4 years of age, with a male predominance. The adult cases occur mostly between the 5th decade and 6th decade of life, with a female predominate.2 The prevalence is difficult to determine due to its rarity. Most authors regard cystic nephroma of the childhood as a benign end of the spectrum with cystic partially differentiated nephroblastoma and nephroblastoma (Wilms tumor). The same terminology used in adult cases has the similar histopathologic features, but is considered to have a different origin and to be not associated with nephroblastoma or nephrogenic rests.

Fig. 1. Computed tomography reveals a multilocular cystic tumor occupying the left abdominal space.

Cystic nephroma of childhood was first considered as a unilateral and sporadic neoplasm. However, bilateral and familial cases associated with pleuropulmonary blastoma have also been reported in recent decades.3 The pathogenesis is uncertain. The DICER1 mutations may be the major genetic event in the development of cystic nephroma.4

The clinical presentation is usually as a palpable abdominal mass incidentally found by parents or caretakers, or during routine physical examination. Ultrasonographic findings are multiple anechoic spaces separated by thin septa. On computed tomography, it is a circumscribed and multilocular cystic tumor. However, the entire, or portions of, tumor may appear solid on both ultrasonography and computed tomography, because of the aggregation of small-sized cysts.5 It is difficult to distinguish cystic nephroma from cystic partially differentiated nephroblastoma or other renal tumors with cystic change on radiological studies. The final diagnosis depends on the pathologic examinations.

Grossly, cystic nephroma is a well-circumscribed tumor with a smooth surface. The cut surface reveals variable-sized cysts separated by fibrous septa. The cysts contain clear or yellow fluid. They may be herniated into the renal pelvis but do not communicate with it. No solid nodule should be found. Tumor necrosis, hemorrhage, and calcification are extremely rare. The histologic diagnosis criteria were first established by Boggs and Kimmelstiel6 in 1956 and modified by Joshi and Beckwith7 in 1989. Microscopically, the cysts are lined by flattened, cuboidal, or hobnailed epithelium. The

Fig. 2. The tumor is composed of variable-sized cysts separated by fibrous septa (H&E 40×). H&E = hematoxylin and eosin.

Fig. 3. Cysts are lined by a single layer of low cuboid cells (H&E 400×). H&E = hematoxylin and eosin.

Fig. 4. Focal hobnail appearance (H&E 400×). H&E = hematoxylin and eosin.
septa are composed of fibrous tissue without poorly differentiated or blastemal elements. Mature tubules may or may not be present. The residual renal tissue should be essentially normal.

The differential diagnosis of other childhood neoplasms includes cystic partially differentiated nephroblastoma and solid tumors with cystic change such as nephroblastoma, clear cell sarcoma, congenital mesoblastic nephroma, and renal cell carcinoma. Occasionally, developmental disorder such as cystic renal dysplasia may also be mistaken as a cyst tumor.

Cystic partially differentiated nephroblastoma has a similar structure to cystic nephroma. However, blastemal elements, nephroblastomatous epithelial elements, and undifferentiated or differentiated mesenchyme are present in the septa. By definition, no expansive solid nodule exists in the tumor. Cystic partially differentiated nephroblastoma has a low malignant potential for local recurrence. Surgery is curative in most cases. By contrast, nephroblastoma is at the most malignant end of the spectrum with cystic nephroma and cystically partially differentiated nephroblastoma. It is the most common renal malignant neoplasm of childhood and has the ability to metastasize and recur. Pre- and postoperative chemotherapy may be needed. Although hemorrhage or tumor necrosis can make cystic change, residual solid expansive areas can still be found. Triphasic patterns composed of blastemal, epithelial, and stromal elements are the most characteristic pathologic findings. Sometimes, biphasic or monophagic tumor can also be observed. The prognosis depends on the stage and whether the anaplasia area is present or not.

Congenital mesoblastic nephroma may undergo cystic change, especially cellular type. Unlike cystic nephroma, the stroma is composed of interlacing fascicles of spindle cells with a much higher cellularity. Tumor cells dissect and entrap renal parenchyma with an infiltration border. Congenital mesoblastic nephroma has an excellent prognosis after complete excision. However, recurrence and metastasis have also been reported.

Clear cell sarcoma is a rare pediatric renal tumor with a propensity to metastasize to bone. Cystic change secondary to hemorrhage and necrosis has been reported. Tumor cells can be epithelioid or spindled. Production of extracellular myxoid material that mimics clear cytoplasm is the characteristic of this tumor. Treatment requires nephrectomy followed by chemotherapy.

Renal cell carcinoma occasionally occurs in children. The mean age at diagnosis is older than that of cystic nephroma. Similar to the malignant tumors described earlier, hemorrhage and necrosis can also make cystic change in renal cell carcinoma. However, it is usually not difficult to make a differential diagnosis of cystic nephroma based on the cytology and solid growth areas.

Cystic renal dysplasia is a disorder that occurs during embryo development. Most of the cases are unilateral. The affected kidney consists of irregular cysts surrounded by primitive mesenchyme. Cartilage and adipose tissue can be found. Glomeruli and renal tubules are scattered in the interstitium without an ordered distribution.

In conclusion, the clinical significance of cystic nephroma is its benign behavior, and it should be distinct from other more common pediatric malignant neoplasms. Neither clinical presentations nor radiological studies can predict the histologic pictures of cystic renal tumor of childhood. Surgical intervention and histopathologic examination are necessary for the final diagnosis. Noninvasive follow-up is recommended after the complete resection.

Conflicts of interest
The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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