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CASE REPORT

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Cystic nephroma: a rare benign renal tumor

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Abstract We report two boys aged 1 year and 2 years 2 months, respectively, with cystic nephromas. Both presented with a painless abdominal mass. Computed tomography showed a homogeneous, multicystic tumor of the lower pole of the kidney in both cases with thin septa without solid parts. Macroscopically, the surface of the tumor was smooth. Both patients underwent a renal-sparing procedure; histology confirmed the diagnosis of cystic nephroma.

Key words Renal tumor · Cystic nephroma · Multilocular renal cyst · Child

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Introduction

Cystic lesions of the kidney are common, whereas cystic tumors of the kidney are rare and may present a diagnostic challenge. Cystic renal neoplasms in childhood represent a spectrum of clinicopathologic entities with cystic nephroma and cystic, partially-differentiated nephroblastoma on the benign end and polycystic Wilms' tumor on the malignant end. We report two boys with a preoperatively suspected diagnosis of cystic nephroma treated by a renal-sparing procedure.

Case reports

Case 1 A 1-year-old boy presented with a painless abdominal mass on the left side. Radiologic work-up with ultrasound (US) and computed tomography (CT) showed a multicystic tumor in the left lower renal pole about 8 cm in diameter with thin septa without solid parts (Fig. 1a and b). Intraoperatively, the tumor presented as a homogeneous cystic tumor with thin septa that was clearly cut off from the regular renal parenchyma (Fig. 2). Therefore, a renal-sparing procedure, i.e., resection of the cystic tumor, was performed. Histology confirmed the diagnosis of cystic nephroma (Fig. 3).

Case 2 The mother of this 2-year-2-month-old boy noted a painless mass in the right hemiabdomen. US and CT showed a multicystic tumor of the lower pole of the right kidney about 10 cm in diameter. No solid parts were encountered. A smooth tumor surface was noted macroscopically, suggesting a cystic nephroma, and a renal-sparing excision was performed. Histology revealed the diagnosis of cystic nephroma.

Discussion

Cystic nephroma, formerly called multilocular renal cyst, is a very rare benign tumor of the kidney presenting mainly as a painless abdominal mass, abdominal or flank pain, or hematuria [3]. There are only about 200 case reports in the literature [5]. Shimokama and Watanabe [6] suggested on the basis of scanning and transmission electron microscopic observations that the pathogenesis of this tumor is a neoplasm probably originating from the ureteral bud.

Cystic nephroma may be seen at all ages, but occurs predominantly during the first 2 years of life and mainly in boys, as in our cases [3]. It usually affects only one kidney, but bilateral tumors have been reported [4]. US shows a multicystic lesion, but the cyst walls in septa may mimic solid parts. Intravenous urography simply shows an extended processes, eventually with obstruction, and is not very helpful. CT is the most reliable diagnostic tool and is usually distinctive, showing multiple fluid-filled cysts separated by thin septa [7]. Absence of normal renal tissue or solid parts in the septa distinguishes the tumor clearly from other renal cystic lesions, especially cystic nephroblastoma. Macroscopically, the tumor presents with a

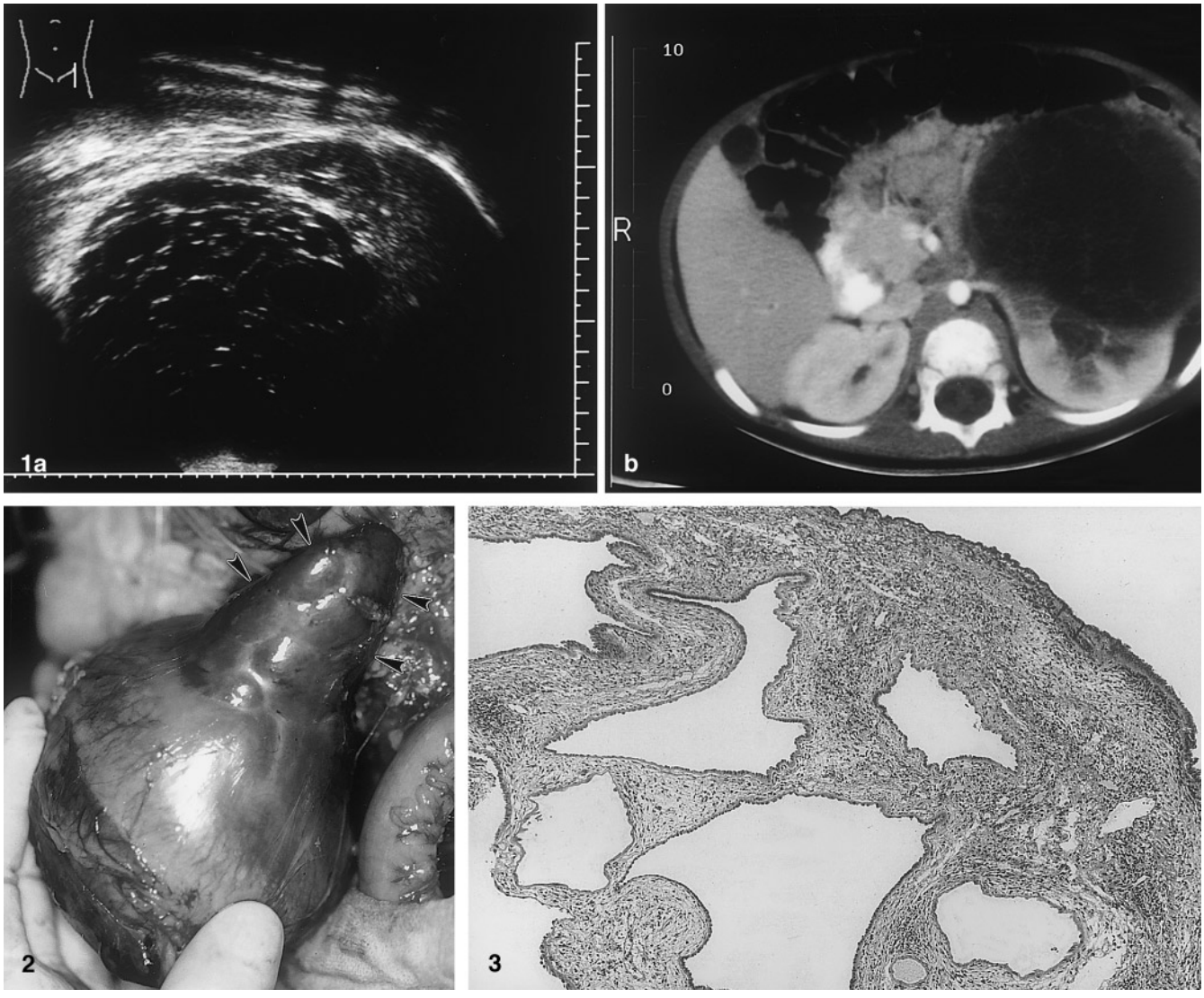

Fig. 1 **a** US scan of left kidney demonstrating multicystic tumor of lower pole. **b** CT scan showing cystic tumor of left kidney with thin septa without solid tumor

Fig. 2 Macroscopic view of tumor with smooth surface and adjacent regular renal parenchyma (*arrowheads*)

Fig. 3 Cystic nephroma containing large cysts with epithelial lining. Solid components are made of loose connective tissue that includes focal inflammatory cells, but never renal tissue or blastema (H & E, ×50)

smooth external surface and usually clear demarcation from the regular renal parenchyma. We noted an absence of tumor vessels in contrast to cystic Wilms' tumors.

The criteria for diagnosis of cystic nephroma were laid down by Boggs and Kimmelstiel [2] in 1956 and are listed in Table 1. Although most authors have performed a nephrectomy, some strongly advocate a renal-sparing procedure when the characteristic radiologic findings are present [3, 4] Our experience in two cases with grossly smooth external tumor surface and regular renal parenchyma, i.e., a distinctly benign appearance, in addition to well-defined radiologic findings, supports the use of a renal-sparing procedure. If the pathomorphology of the resected specimen shows undifferentiated cellular elements between cysts, the diagnosis would be a cystic, partially-differentiated nephroblastoma, a very rare low-grade malignant tumor. There are no reports of metastases or other malignant behavior of this tumor, and therefore, chemotherapy is not indicated [3].

Table 1 Histologic criteria of cystic nephroma

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 renal pelvis
4.	The residual renal 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 lesions

Accordingly, an uncertain preoperative diagnosis of cystic nephroma versus cystic, partially-differentiated nephroblastoma is of little importance, since chemotherapy and radiotherapy are no longer used in treating the latter [1].

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