MULTILOCULAR CYSTIC NEPHROMA: A CLINICAL CASE

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Summary.- OBJECTIVE: To describe the clinical presentation, preoperative diagnostic possibilities, and treatment of cystic nephroma.

METHODS: We describe a case of cystic nephroma in an adult male and show that, both in our patient and in the literature, a definitive diagnosis can only be obtained postoperatively, even when there is a reasonable clinical suspicion.

CONCLUSIONS: Cystic nephroma is rare, and some authors consider it a questionable entity. However, it should be distinguished from renal cystic neoplasms, which can resemble the condition.

Keywords: Cystic Wilms tumor. MEST. Cystic clear-cell carcinoma. Partial nephrectomy.

INTRODUCTION

Multilocular cystic nephroma was first described in 1892 by Walter Edmunds, who called it cystic adenoma (1).

Over 200 cases have been published, but the number is most certainly higher, given the high variability in the terms used to identify the condition. The tumor has been called a benign cystic nephroma, multilocular renal cyst, cystic hamartoma, cystadenoma, and Perlman tumor, as well as multilocular cystic nephroma, the term used most often at present (2,3).

PRESENTATION

Cystic nephroma is a rare, usually unilateral renal mass composed of congenital, nonhereditary cysts of unknown etiology. We present a clinical case recently seen in our department.

CASE REPORT

A 48-year-old man with primary biliary cirrhosis was hospitalized for febrile syndrome and hematuria and diagnosed with lower respiratory tract disease. Imaging studies showed a right renal cystic mass.
The renal ultrasound revealed a multilobular lesion in the lower pole of the right kidney, with echogenic content in its interior of 3x3 cm (Figure 1).

The computed tomography (CT) scan showed a multilobular cystic image in the lower pole of the right kidney that measured 3.5x3 cm, had internal partitions and a small, somewhat more solid area in its medial portion. Post-contrast enhancement was minimal (Figure 2).

Contrast magnetic resonance imaging (MRI) of the kidney revealed a well-circumscribed mass in the lower pole of the right kidney that measured 3.5x3 cm. The mass was multilobular, had hyperintense content in the contrast-enhanced T2-weighted sequences and hypointense in the T1-weighted sequences. The mass was cystic and multiseptated, but showed no evidence of solid nodules or images of fatty intensity in its interior. There was slight contrast uptake at the septal level (Figure 3).

These findings were initially consistent with renal neoplasms, although the differential diagnosis included cystic renal cell carcinoma, cystic nephroma and multilocular cyst.

The patient underwent lower pole nephrectomy via a right lumbotomy. A macroscopic observation of the surgical specimen showed a wedge of renal tissue that measured 4x1.5x3 cm and presented a raised multilobular lesion on the surface. A cross-section of the specimen revealed a multilocular cystic lesion with clear brown content (Figure 4).

Histology showed a well-delimited mass composed of variable-sized cystic formations comprising a single row of flattened epithelial cells, some with clearer cytoplasm, without mitosis, separated by fine fibrous septa formed of spindle-shaped cells, but no clear-cell, muscle- or adipose-tissue aggregates and no immature or blastemal components.

**DISCUSSION**

Cystic nephroma is a benign condition, and only one case in which renal cell carcinoma cystic coexisted has been reported (4). The tumor tends to be single, unilateral, and composed of noncommunicating cysts, although several bilateral cases were published (5). In 1956 Boggs and Kimeshtshel established the criteria for the diagnosis of multilocular cyst based on Powell’s 1951 criteria. These criteria were reformulated by Joshi and Beckwith as follows (6,7):

a) the lesion is composed entirely of cysts and their septa;

b) it forms a discrete mass, well demarcated from the noncystic renal parenchyma;

c) the septa are the only solid portion of the tumor, conforming to the outlines of the cysts without solid expansive nodules;

d) the cysts are lined by flattened, cuboidal, or hobnail epithelium; and

e) the septa are composed of fibrous tissue in which well-differentiated tubules may be present.

Eble and Bonsib (8) advocate similar criteria and add that it is a mass surrounded by a fibrous pseudocapsule.
and that the septa can contain epithelial structures similar to mature renal tubules, but should not contain epithelial cells with clear cytoplasm.

There are two well-defined age groups in terms of presentation (7):

- children younger than 4 years of age, almost always boys.
- children older than 4 years of age, almost always women (with a peak between age 4 and 20, and another between age 40 and 60).

In children younger than 4 years, the tumor often presents as a palpable mass whereas in adults, it presents as an incidentaloma or nonspecific clinical symptoms.

The congenital forms tend to appear in childhood, and the male-to-female ratio is 2:1. In adult patients, the ratio is 1:9.

Several authors consider that although congenital cystic nephromas in children and adults share morphological characteristics, they are different entities due to their different biological behavior (9). In fact, some consider that the term “cystic nephroma” should only be used in adult cases and that childhood renal neoplasms composed of epithelium-lined cysts and separated by septa without expansive nodules should be diagnosed with cystic partially differentiated nephroblastoma, regardless of the presence or absence of immature tissue (8).

Recent evidence suggests that tumors that were initially diagnosed as cystic nephroma in the adult population represent a genetically and histologically distinct entity called MEST (mixed epithelial and stromal tumor of the kidney), i.e., a benign, biphasic tumor with an epithelial and stromal component (3,10). Both lesions affect adults, women in particular, and some morphological and clinical characteristics are the same; both are well circumscribed, although some authors believe that cystic nephroma in the adult is an inconclusive or rare condition (10). MEST is usually composed of multiple cysts and septa with solid mesenchymal areas, whereas cystic nephroma is composed entirely of multiple cysts and septa without solid portions. According to Antic (3), cystic nephroma and MEST would be at the opposite ends of the spectrum for the same lesion. Although cystic nephroma can be seen in patients of both sexes who have no history of estrogen therapy or obesity, MEST is a tumor that occasionally shows cells similar to ovarian stroma, probably as a result of hormonal influences.

The actual origin is unknown; in some cases the pathogenesis of the tumor has been related to hormones because it is more likely to affect women, particularly those receiving oral estrogen therapy. At least one published case has been associated with hormonal treatment manipulation in prostate cancer. In some cases, the presence of estrogen and progesterone receptors in the stroma of these tumors support this hypothesis (9).

Immunohistochemical studies in both lesions (MEST and cystic nephroma) reveal the epithelial components which are diffusely positive for pan-keratins and focally positive for CD10, CK7 and high-molecular-weight keratins and negative for CK203.

The clinical symptoms are nonspecific: the condition can manifest as urinary symptoms, abdominal or lower

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**Figure 3.** Magnetic resonance image showing a multilobular mass of cystic characteristics in the lower pole of the right kidney.

**Figure 4.** Multilocular cystic lesion separated by fibrous septa and compressing the adjacent renal parenchyma.
back pain, hematuria or hypertension in adults and as a palpable mass in children.

The differential diagnosis should include cystic Wilms tumor/cystic partially differentiated nephroblastoma in childhood and with cystic clear-cell renal cell carcinoma and multilocular cystic carcinoma in adulthood. The differential diagnosis should also include other processes or syndromes in which cysts and neoplasms can coexist in the kidneys (e.g., polycystic kidney, renal lymphangiomatosis, segmental multicystic dysplastic kidney, complex tuberous sclerosis, von Hippel-Lindau disease, cystic clear-cell sarcoma of the kidney). Complex tuberous sclerosis consists of multiple angiomylipomas and cysts lined with large eosinophil cells, whereas in renal lymphangiomatosis, there are multiple perirenal and parapelvic lymphatic cysts (5). In these last two conditions and in polycystic kidney, radiological studies will aid diagnosis, but in other cases, no imaging studies are able to provide an accurate diagnosis. Imaging studies are not definitive; pyelography can show a mass effect with displacement or compression of the pelvis or calyces. In some patients, tumor prolapse is observed inside the renal pelvis. The sign has been considered characteristic of these lesions and is the result of protrusion by one or more cysts into the collector system. In the CT scan, the appearance of a smooth mass introduced or invaginated by digitations into the pelvis and/or calyces has also been described (6,11). The CT scan and the MRI in particular allow better visualization of the septa and in some cases, can rule out the presence of solid components. However, an ultrasound study or CT or MRI scan will usually show a multilocular cystic renal mass and the radiological reports will suggest the presence of a “complex renal cyst”. For this reason, the Bosniak classification is used most often to indicate the treatment.

The definitive diagnosis is histological, based on a microscopic examination of the surgical specimen. Cystic nephroma and cystic partially differentiated nephroblastoma are macroscopically indistinguishable. Septa in the cystic nephroma may be fibrous and contain well-differentiated tubular structures, should not contain poorly differentiated tissues or blastema cells, and is uncommon before age 30 years. Cystic partially differentiated nephroblastoma contains blastema cells with or without other embryonic stromal cells, but without solid portions, and can also contain skeletal muscle fibers; the tumor is rare after 2 years of age (5,6,8,11,12). Cystic nephroma has no solid components, which differentiate it from Wilms tumor and other renal neoplasms. The histological difference with clear-cell renal cell carcinoma is based on the absence of clear-cell aggregates or nests in the septa and absence of atypias.

Fine-needle aspiration biopsy of the lesion has low sensitivity and is of little diagnostic benefit because representative samples of most cystic septa cannot be taken.

Because no diagnostic method can prove whether the condition is benign or not, the Bosniak classification should be applied in the case of a cystic renal lesion that does not meet the radiological criteria of a simple cyst. Multilocular cystic renal cell carcinoma, cystic nephroma, and MEST are usually classified as Bosniak categories II and III. The course tends to be benign in all patients (although one case of malignant transformation originating in the solid mesenchymal component of the MEST has been reported (13)) and, therefore, the indication is partial nephrectomy (10,13). On follow-up, most cases of multilocular cystic nephroma have a favorable, benign course and, therefore, optimal treatment is conservative nephroma surgery, i.e., partial nephrectomy or tumorectomy with intraoperative biopsies of the surgical specimen and surgical bed.

**CONCLUSIONS**

Cystic nephroma is a rare, benign condition that is hard to diagnose preoperatively and ideally treated by partial nephrectomy.

**REFERENCES AND RECOMMENDED READINGS**

*of special interest, **of outstanding interest


**AA TYPE PRIMARY AMYLOIDOSIS OF THE URINARY BLADDER: CLINICAL CASE AND BIBLIOGRAPHIC REVIEW**

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### Summary

**OBJECTIVE:** Amyloidosis is a disease characterised by deposition of eosinophilic hyaline material in different tissues. Urinary bladder involvement is uncommon with less than 200 cases of the primary form published in the literature. We present a new case of primary AA type amyloidosis of the urinary bladder (typical of secondary forms).

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