

Cystic Nephroma: Report of a Case and Review of the Literature

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Abstract. *Cystic nephroma is a rare benign cystic renal tumor, which has been only recently recognized as an exclusively adult histological entity. We present a case of cystic nephroma, together with clinical, radiological, histopathological and immunohistochemical findings. The histopathological differential diagnosis and immunohistochemical features that are potentially useful for refining this tumor are discussed.*

Cystic nephroma is a rare benign multicystic renal tumor. This uncommon lesion, also termed multilocular cyst and multilocular cystic nephroma, has been thoroughly defined by Eble and Bonsib as "an entirely cystic lesion with no grossly recognizable expansible stroma" (1). These authors also determined this lesion to be an exclusively adult entity. Approximately 200 cases of cystic nephroma have been described in the literature to date since the first original report in 1892 (2). We present a case of cystic nephroma, together with clinical, radiological, histopathological and immunohistochemical findings. The histopathological differential diagnosis and immuno-histochemical features that are potentially useful for refining this tumor are discussed.

Case Report

The patient, a 38-year-old man, was referred to the Surgery Department of the Second University of Naples with signs of right-sided abdominal pain. Ultrasonography and CT revealed a multicystic mass with thin septae and apparently no solid component, occupying the middle and upper portions of the right kidney (Figure 1A). CT of the chest was found to be normal. Right radical nephrectomy was performed and the resected specimens weighed 565 g. No

adjunct therapy was administered. The patient is free of any recurrence two years after the surgery.

Gross examination of the tumor mass revealed a capsulated large multilocular cystic tumor originating from the middle and upper parts of the kidney. The mass was clearly demarcated from the surrounding normal renal tissue.

The excised biopsy tumor specimens were fixed in 10% buffered-formalin and paraffin embedded. Sections of 5 µm were stained with haematoxylin-eosin, haematoxylin-van Gieson and PAS-haematoxylin. For immunohistochemistry, the avidin-biotin complex (ABC) method was applied as described elsewhere (3). We used a panel of monoclonal antibodies for the following markers: Vimentin at 1:500, CK19 at 1:100, CK AE1/AE3 at 1:100, CD34 at 1:100 and smooth muscle actin at 1:200 (all the antibodies were from DAKO, Carpinteria, CA, USA). Appropriate controls were tested simultaneously.

Results

Microscopic examination showed that the cyst walls were lined with a layer of flattened or hobnail cuboidal epithelium (Figure 1B). Between the cysts, there was loose stroma with spindle-shaped fibroblasts and cells resembling smooth muscle cells. The septae were mainly composed of fibrous tissue and contained dilated vessels. In some areas, occasional tubular structures or focal aggregates of lymphocytes were found. No blastema was identified and there were no mitotic figures. No microscopic capsular, lymphatic, or vascular invasion was identified. A small portion of the lower pole of the kidney had normal-appearing renal parenchyma. Immunohistochemistry showed that the tumor cells were strongly positive for CK19 and CK AE1/AE3 (Figure 1C and D), while the stroma was strongly positive for vimentin, CD34 and smooth muscle actin (data not shown).

Discussion

Cystic nephroma is an uncommon, non-heritable, unilateral, benign tumor of the kidney in adult patients (1). Cystic nephromas are commonly found incidentally on radiographic

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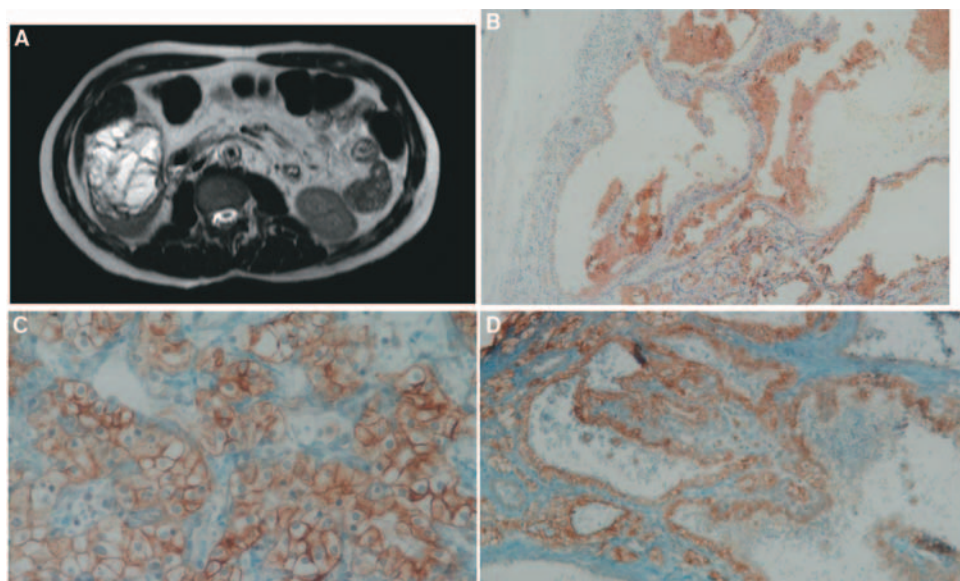


Figure 1. Cystic nephroma in a 38-year-old male patient. A) CT-scan appearance of the neoplastic mass; B) Histological appearance of the neoplasm, showing cystic spaces limited by cuboidal cells (haematoxylin/eosin, original magnification x20); C) The cuboidal tumor cells express CK 19 (ABC, original magnification x40); D) The cuboidal tumor cells express CK AE1/AE3 (ABC, original magnification x20).

studies, but may present as an abdominal mass found on routine physical examination. The clinical, radiological and histological features of cystic nephroma are well defined (1-4). Cystic nephroma is actually classified as a multilocular cystic renal tumor of adult patients, with no relation to the very similar mature forms of Wilms tumor occurring in children (1). The immunohistochemical features of cystic nephroma have not been studied in great detail and very few reports are available in the literature (2, 5-7). Our immunohistochemical results indicate that the epithelial component of the neoplasm expresses cytokeratins, while the stroma was positive for vimentin, CD34 and smooth muscle actin.

The differential diagnosis of cystic nephroma includes multicystic dysplastic kidney, a very necrotic or hemorrhagic Wilms tumor, clear cell sarcoma, or a cystic variant of mesoblastic nephroma (8). Differentiation between these entities has prognostic and diagnostic significance. To label a lesion as a cystic nephroma, the CT-scan appearance of the tumor is important as is the absence of blastemal or poorly differentiated cells on microscopic examination.

Treatment of cystic nephroma consists of surgical excision, which is curative. In our case, the patient had an uneventful recovery after undergoing complete resection of the cystic nephroma.

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