CLINICAL CASE

Cystic nephroma: a benign tumor with a malignant appearance


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Abstract
Cystic nephroma is a renal lesion belonging to the group of mixed epithelial and mesenchymal tumors, together with other infrequent tumors, such as synovial sarcoma and mixed epithelial and stromal tumor. It is considered one of the rarest renal tumors, and up to the present date, fewer than 250 cases have been reported in the international medical literature. A 24-year-old homosexual man with a smoking index of 2 had a past history of condylomatous perianal lesion resection. He presented with left renal colic that was medically managed. Physical exploration revealed left costovertebral angle percussion. Urinalysis reported a pH of 5.5, blood ++, and a scant quantity of yeasts. The patient was HIV negative. Tomography scan identified an incidental left renal lesion with a Bosniak III multicystic component (25.3 mm x 28.2 mm). Magnetic resonance imaging showed a complex cystic lesion. A left open partial nephrectomy was performed and the histopathologic study reported cystic nephroma. The patient had adequate postoperative progression and was released on the 4th day after surgery. Cystic nephromas are rare lesions that are commonly diagnosed in the postoperative period because their radiologic characteristics are similar to complex cysts suggesting malignancy. Treatment is surgical and nephron-sparing surgery is preferred in young patients with a small lesion. Cystic nephromas are rare tumors and it is a challenge to differentiate them from malignant renal tumors and thus manage them by sparing functioning renal units whenever possible. © 2015 Sociedad Mexicana de Urología. Published by Masson Doyma México S.A. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
Introduction

Cystic nephromas are uncommon tumors of benign behavior grouped together with the mixed epithelial and stromal tumors in accordance with the World Health Organization classification. Around 250 cases have been reported in the literature and the large majority of reported cases are lesions documented as tomographic findings during the evaluation protocol of some other disease. Their radiologic characteristics suggest malignant neoplasia of the kidney, meriting a more complete diagnostic protocol.

Case presentation

A 24-year-old homosexual man had a past history of smoking and perianal condylomata acuminata resection 2 years before his present evaluation. The patient’s illness began 3 months earlier, with a colicky pain located in the right renal fossa that irradiated toward the contralateral lumbar region with no other associated symptoms.

Physical examination revealed a conscious and oriented patient with adequate coloration of the skin and teguments, an ectomorphic body type, head and neck with no alterations, no apparent cardiopulmonary involvement, a flat, soft, nonpainful, depressible abdomen with no visceromegaly, and no technical complications. Surgery duration was 3 h, warm ischemia time was 30 min and a specimen measuring 4.5 x 3.5 x 3 cm with negative surgical margins was obtained.

Laboratory tests reported: hemoglobin 14.7 mg/dl, hematocrit 42.2%, leukocytes 5,500/μl, neutrophils 57.5%, platelets 216,000/μl, glucose 93 mg/dl, BUN 19 mg/dl, urea 40.66 mg/dl, creatinine 0.79 mg/dl, sodium 139 mEq/l, potassium 4.1 mEq/l, chloride 109 mEq/l, PT 15.8 sec, PTT 40.66 mg/dl, creatinine 0.79 mg/dl, sodium 139 mEq/l, potassium 4.1 mEq/l, glucose 93 mg/dl, BUN 19 mg/dl, urea 40.66 mg/dl, creatinine 0.79 mg/dl, sodium 139 mEq/l, potassium 4.1 mEq/l, chloride 109 mEq/l, PT 15.8 sec, PTT 28.2 sec, total bilirubin 0.7 mg/dl, direct bilirubin 0.0 mg/dl, indirect bilirubin 0.7 mg/dl, AST 33 IU/l, ALT 35 IU/l, total proteins 7.0 g/dl, albumin 4.2 g/dl, globulin 2.8 g/dl, lactate dehydrogenase 129 IU/l, alkaline phosphatase 70 IU/l, GGT 22 IU/l, urinalysis with blood ++, proteins, bacteria, nitrites or negative cylinders. The laboratory tests were within normal parameters, with the exception of microhematuria.

CT-urography revealed data of a left, 28 x 25 mm, irregular, exophytic, cortical renal tumor located in the middle third, with a 30 HU density and no contrast medium enhancement. It had a multiloculated, cystic aspect with solid areas and discreet septa in its interior (Bosniak III) (fig. 1).

The diagnostic protocol was complemented with magnetic resonance imaging that corroborated the solid areas of the lesion with the discreet cystic component (fig. 2).

A preoperative diethylenetriaminepentaacetic acid kidney scintigram identified a total glomerular filtration rate of 116 ml/min, a left kidney glomerular filtration rate of 63.85 ml/min (54.69%), and a right kidney glomerular filtration rate of 52.91 ml/min (45.31%).

The initial decision was to perform tumor enucleation, calculating the R.E.N.A.L. Nephrometry Score and identifying a low complexity lesion (6 points). Finally, an open partial nephrectomy was carried out on the left kidney with no technical complications. Surgery duration was 3 h, warm ischemia time was 30 min and a specimen measuring 4.5 x 3.5 x 3 cm with negative surgical margins was obtained (fig. 3).

An irregularly shaped ovoid sample (fig. 4) that had a brown surface with hard, exophytic lesions was sent to the pathology department and the histopathology report stated cystic nephroma (fig. 5).

At the follow-up consultation at 3 months the patient was asymptomatic, with creatinine of 0.6 ml/min and no alterations. A postoperative follow-up diethylenetriaminepentaacetic acid kidney scintigram was taken that identified a total...
glomerular filtration rate of 119.35 ml/min, a left kidney glomerular filtration rate of 57.75 ml/min (48.39%), and a right kidney glomerular filtration rate of 61.6 ml/min (51.61%). Adequate functioning of the left kidney was verified 3 months after the partial nephrectomy.

Discussion

Cystic nephroma is a rare, non-familial tumor with a bimodal distribution of age and sex. It shows a 75% predilection in the pediatric population for the male sex, whereas in later stages of life, females are typically more affected between the fifth and sixth decades of life. 4

Diagnosis is often incidental and discovered during urologic or non-urologic abdominal imaging studies. Cystic nephroma rarely causes symptomatology, but when present, it can manifest as lumbar pain, hematuria, or recurrent urinary infections. When there are symptoms, the lesion is usually located close to the renal pelvis.

Diagnostic accuracy is histopathologic, through biopsy or surgical excision of the lesion, which are essential, given that in imaging studies (tomography, magnetic resonance) the lesions are indistinguishable from renal carcinoma, particularly the clear cell type with a cystic component.5

The radiologic findings of cystic nephroma can include a cyst associated with calcifications (Bosniak III or IV) that are initially highly suggestive of renal cell carcinoma.6

The histopathologic characteristics upon the slicing of the macroscopic sample tend to be lesions delimited by simple epithelium and fluid in their interior with 2 distinctive microscopic characteristics:7

1. Nail-head morphology. The epithelium that surrounds the nucleus of the cyst is projected into the interior of the cyst.

2. Stroma resembles ovarian stroma with basophilic cytoplasm and cell morphology. In addition, cystic nephromas have an immunologic pattern similar to that
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Figure 4 A. Macroscopic specimen after preparation. B and C. Pathologic cross-section.

Figure 5 Photomicrography x40. Hematoxylin-Eosin stain. Microscopic characteristics of the cystic nephroma. Nail-head morphology.

of the ovarian stroma with positivity for estrogen and progesterone receptors. They usually show positivity for CD-10 receptors, as well as inhibin and calretinin.

The differential diagnosis should be made with other renal tumors, mainly those of malignant behavior, including partially differentiated or standard cystic nephroblastoma, mesoblastic nephroma, clear cell carcinoma with a cystic component, and other renal cysts. 8

Habitual treatment is almost always partial or radical nephrectomy, and sometimes with lymphadenectomy in the initial management, if technically feasible, when renal tumor is suspected. Once the surgery is performed and the definitive pathologic diagnosis of cystic nephroma is made, no adjuvant treatment is required and disease course is benign.9

In regard to follow-up, no cases of recurrence or metastasis in these tumors have been reported.10

Conclusions

Cystic nephromas are very rare lesions. Their incidental finding and radiologic characteristics suggestive of malignancy require invasive treatment in the majority of cases. Definitive diagnosis is made through histopathology and at present is retrospective in nature. Greater evidence and more complete diagnostic protocols are needed to differentiate this lesion from malignant neoplasias, so that patients with this diagnosis are spared from unnecessarily undergoing invasive procedures, such as partial nephrectomy, or in the worst of cases, radical nephrectomy.

Ethical responsibilities

Protection of persons and animals. The authors declare that the procedures followed conformed to the ethical standards of the responsible committee on human experimentation and were in accordance with the World Medical Association and the Declaration of Helsinki.

Data confidentiality. The authors declare that they have followed the protocols of their work center in relation to the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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Conflict of interest

The authors declare that there is no conflict of interest.

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