

Adult Multiloculated Cystic Nephroma: A Case Report

Eriřkin Tip Multiloküle Kistik Nefroma: Olgu Sunumu

Orkun Batmaz[®], Murat Uçar[®], Erdem Aktař[®], Bahar Akkaya[®], İsmail Türker Köksal[®]

Olgu Sunumu
Case Report

ABSTRACT

A 71-year old women who underwent radical nephrectomy due to left renal mass is presented. The histopathological examination was reported as adult cystic nephroma, which is very seldom benign tumour of the kidney. The epidemiology, differential diagnosis, histopathological features, and treatment options are discussed.

Keywords: Cystic Nephroma, Benign, Kidney, Pathology

Öz

Sol böbrek kitlesi nedeniyle radikal nefrektomi yapılan 71 yařındaki kadın hastada eriřkin tip multiloküle kistik nefroma olgusu sunulmuřtur. Eriřkin multiloküle kistik nefroma oldukça nadir görölen benign bir tümördür. Olgumuzda hastalığın epidemiyolojisi, histopatolojik özellikleri ve tedavi seçenekleri tartiřılmıştır.

Anahtar kelimeler: Kistik Nefroma, Benign, Böbrek, Patoloji

INTRODUCTION

Multilocular cystic nephroma (MCN) is a rare benign lesion of the kidney which usually presents as a unilateral multicystic renal mass without solid elements and more often seen in bimodal age distribution and adult onset form especially in postmenopausal women. It was first described in 1892 as cystic adenoma of the kidney. Preoperative diagnosis of MCN is difficult and surgery is recommended to exclude malignant cystic lesions of kidney. However, partial removal of the mass can be an option depending on the site and size of the lesion. In this case, we are presenting a 71-year-old woman with MCN treated successfully with total nephrectomy.

CASE REPORT

A 71-year-old woman was admitted to our urology department with a mass in her left kidney. She was investigated for the presence of resistant hypertension in nephrology outpatient clinic. Her systolic, and diastolic blood pressures at rest were 170 mmHg and 100 mmHg, respectively. She was using irbesartan 300 mg and propranolol 50 mg. She also had hyperlipidemia, atrial fibrillation and were using rosuvastatin 20 mg, furosemide 40 mg and apixaban 5 mg per day. Her creatinine (1.07 mg/dL), GFR: (51 mL/min/1.73 m²) and BUN (22.67 mg/dL) levels were as indicated. She had not any flank pain, hematuria or dysuria, history of smoking and family history of renal carcinoma.

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Orkun Batmaz
Tepecik Training and
Research Hospital,
Department of Urology,
İzmir - Turkey

✉ orkun.batmaz@hotmail.com
ORCID: 0000-0003-3924-2325

M. Uçar 0000-0001-6977-7430
E. Aktař 0000-0002-8268-6653
B. Akkaya 0000-0001-8054-294X
İ.T. Köksal 0000-0002-7606-1200
Department of Urology,
Akdeniz University,
Antalya, Turkey

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In her abdominal ultrasonography, there was a multicystic mass in her lower pole of the left kidney resembling hydatid cyst and cystic renal cell carcinoma. There was a multicystic mass the largest one having a diameter of 5.5 cm. There was no obvious vascular signaling in the septae between the cysts in Doppler US scan which looked like type 3 hydatid cysts. However, a stone with 1.5 cm diameter in the middle calyx of left kidney and deterioration of the collection system. After observing this sonographic finding we decided to perform computerized tomography; and 104x86 mm-multicystic mass with multiple contrasted septae considered as cystic renal cell carcinoma was observed (Figure 1).



Figure 1.

Echinococcus granulosus IgG was not detected. These findings forced us to perform renal exploration. We reached to the left kidney transperitonally and palpated the cystic mass in the lower pole of the kidney. There was no invasion into the adjacent tissues. We found one renal artery and vein without any collateral vasculature. We decided to perform left radical nephrectomy. Operation was performed without complication such as bleeding or injuring the adjacent viscera.

Oliguria/anuria, acute impairment of renal functions or anemia did not develop. After the fourth day of the operation we discharged the patient.

DISCUSSION

MCN is a very rarely seen benign entity characterized by multiple septations composed of differentiated tissue without blastemal elements slow-growing mesenchymal and epithelial neoplasm of kidney and there are more than 200 cases reported in the literature to the best of our knowledge ⁽¹⁾. Bimodal age distribution is often seen, accordingly it is mostly seen in children younger than 2 years and in the middle age (40-69 years) adults (male-to-female children ratio, 3:1 and adult female-to-male ratio, 9:1) ⁽²⁾. Pediatric cystic nephroma is a distinct entity. The presence of immature nephroblastic elements excludes the diagnosis of pediatric cystic nephroma and indicates partially differentiated cystic nephroblastoma ⁽³⁾. Keeping in mind the age, clinical presentation and radiological appearance, cystic partially differentiated Wilm's tumour is a possibility. Since MCN has a benign behaviour it must be differentiated from focal cystic neoplastic lesions, including Cystic Partially Differentiated Nephroblastoma (CPDN), which has a low but distinct capability for local recurrence, differentiates from Wilm's tumour with cystic change. Segmental form of unilateral renal dysplasia also needs to be considered in the clinical and radiological differential diagnosis ⁽⁴⁾.

Adults present with nonspecific signs and symptoms, including abdominal pain, hypertension, and urinary tract infection. Microscopic or gross hematuria can occur at all ages ⁽⁵⁾. Our patient was a 71 year-old female presented to us with an incidental renal mass. Radiologically, MCN usually presents as a solitary, centrally located, approximately 6-cm tumor consisting of small noncommunicating small cysts ⁽⁶⁾. Ultrasonography is usually performed as a first-line diagnostic tool and commonly seen in contrast-enhanced CT as a unilateral mass with irregular cysts containing watery fluid and septae of variable thickness ⁽⁷⁾. Sometimes, extension into the renal pelvis may be seen, which results in hydronephrosis and hemorrhage and usually resembled as category

III Bosniak classification⁽⁸⁻⁹⁾. In time, CT became the primary imaging technique for evaluation of MCN but unfortunately can not be used to certainly differentiate other cystic renal masses like renal hydatid cyst and cystic renal cell carcinoma⁽¹⁰⁾. However, number of cases can encourage the performing percutaneous biopsy before surgery or frozen-biopsy intraoperatively and has the potential to avoid a significant number of major surgical procedures⁽¹¹⁾.

In renal ultrasonography there was a multicystic mass in our patient in the lower pole of the left kidney the largest being 5.5 cm in diameter. There was no obvious vascular signaling in the septae between the cysts in Doppler US can and it was like type 3 hydatid cyst. However, there was a stone with a diameter of 1.5 cm in the middle calyx of left kidney and deterioration of the collection system. In CT; 104x86 mm-multicystic mass with multiple contrasted septae considered as cystic renal cell carcinoma was seen (Figure 1). No metastatic nodules or lymphadenopathy was detected in tomographic evaluation of thorax and abdomen.

In this case we did not perform preoperative biopsy due to well-contrast enhancement of the septae on tomographic views. In our opinion renal hydatid cyst and cystic renal cell carcinoma could be possible diagnoses and we studied serum Echinococcus Granulosus IgG level to exclude hydatid cyst which is endemic for our country.

Nephron-sparing surgery is the suggested surgical treatment modality for a small renal mass. Even though surgery remains the standard of care for small renal tumors, cryoablation and radiofrequency ablation have emerged as minimally invasive treatment alternatives^(7-12,13). In our case, due to tumor size, similarity to cystic renal cell carcinoma and renal hydatid cyst, we decided to perform open total nephrectomy.

The resected left kidney measured 11.5 cmx9cmx6

cm without a solid component. A well-capsulated multilocular cyst mass was noted in the upper pole of the left kidney (Figure 2). Microscopically, well-circumscribed, multilocular tumour with non-communicating cysts lined by hobnail columnar or flattened epithelium was seen. The septa consisted of fibrous stroma, often with areas of cellular con-



Figure 2.

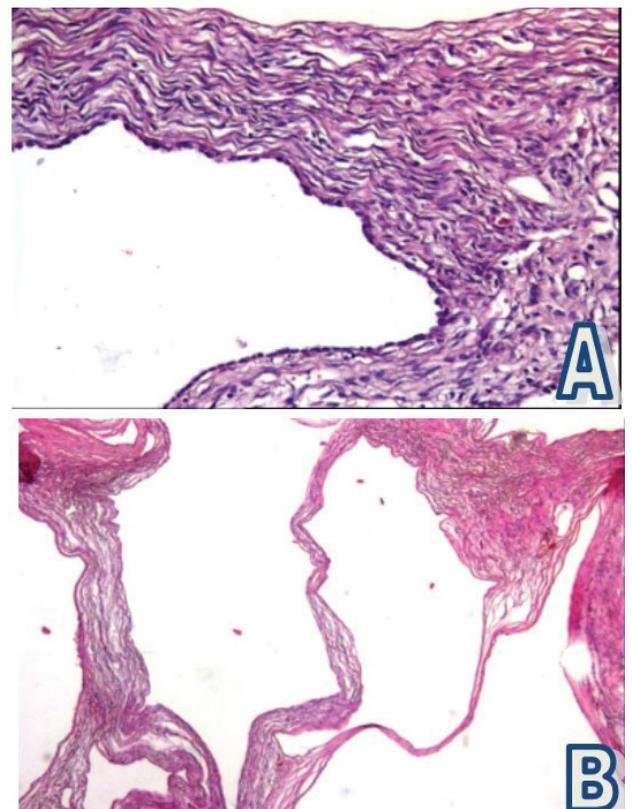


Figure 3.

densation reminiscent of ovarian stroma (Figure 3). Immunohistochemically, tumor stained positively for cytokeratin (CK) 7, CK 19, CK AE1/AE3, vimentin, but it did not stain for estrogen receptor (ER), and progesterone receptor (PR). The final diagnosis of multilocular cystic nephroma was made.

Postoperative prognosis is excellent. However up to now only three cases of recurrent bilateral cystic nephroma have been described in the literature. Other renal lesions may not be differentiated preoperatively from cystic nephroma and thus require surgical exploration. Long-term follow-up is recommended to rule out local recurrence ⁽¹⁴⁾.

Conflict of Interest: None.

Informed Consent: None.

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