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Two Cystic Masses in the Same Kidney: Renal Cell Carcinoma and Cystic Nephroma

Aynı Böbrekte İki Kistik Kitle: Renal Hücreli Karsinom ve Kistik Nefroma

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Abstract

Cystic nephroma (CN) is a rare, multicystic, non-hereditary benign lesion that does not contain solid components. The fact that this entity can not be easily distinguished from other cystic tumors of kidney creates difficulties in diagnosis and treatment. We present a 37-year-old male case with incidentally detected two cystic masses in his left kidney. After nephron-sparing nephrectomy, the histopathology of one of the cystic mass of the patient was reported as renal cell cancer and the other as cystic nephroma. Cystic nephroma is a rare tumor of the kidney. Definitive diagnosis can be made histopathologically rather than using physical examination and imaging methods. We reviewed the literature by presenting our case that had 2 cystic lesions in the same kidney, one of which was renal cell cancer (RCC) and the other was cystic nephroma.

Keywords: cystic nephroma, renal cell cancer, cortical cyst, nephrectomy

Öz

Kistik nefroma, kalıtsal olmayan, solid bileşen içermeyen, böbreğin nadir görülen multikistik benign bir lezyonudur. Böbreğin diğer kistik tümörleri ile kolay ayırt edilmemesi tanı ve tedavide güçlükler yaratmaktadır. İnsidental olarak sol böbreğinde iki adet kistik kitle saptanan 37 yaşında erkek bir olguyu sunuyoruz. Yapılan nefron koruyucu nefrektomi sonrası hastanın kistik kitlesinin birinin patolojisi renal hücreli karsinom (RHK), diğerinin ise kistik nefroma olarak raporlandı. Kistik nefroma, böbreğin nadir görülen bir tümörüdür. Muayene ve görüntüleme yöntemleri ile tanı konulamayıp, kesin tanı histopatolojik olarak konulmaktadır. Aynı böbrekteki 2 kistik lezyondan birinin RHK, diğerinin kistik nefroma çıktığı olgumuzu sunup literatürü gözden geçirdik.

Anahtar Kelimeler: kistik nefroma, renal hücreli kanser, kortikal kist, nefrektomi

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Introduction

Cystic nephroma, first described by Edmunds in 1892, is a non-hereditary, multicystic usually unilateral benign lesion of the kidney that does not contain solid components. Histologically, it contains cysts with flat or cuboidal epithelium and fibrous or stromal septa. It is generally seen in the first 2 years of life and after the third decade. The fact that the kidney has great similarities with other cystic masses, especially cystic renal cell carcinoma, causes diagnostic difficulties, and uncertainty in determining the method of treatment. Histopathological examination is the only way to confirm the diagnosis of multilocular cystic lesion detected by imaging studies [1].

Two cystic masses located side by side in the lower pole of the left kidney of our patient were successfully treated with the "open partial nephrectomy" method. In this study, we presented our case of two cystic lesions in the same kidney, one of which was reported as renal cell carcinoma and the other as cystic nephroma and reviewed the relevant literature.

Case

A 37-year-old male patient without any previously known disease was applied to our clinic. Contrast-enhanced abdominal computed tomography (CT) taken three months before the date of admission, revealed a 33x30 mm Bosniak type 2 cyst in the middle pole of the left kidney, and a 19x16 mm cystic exophytic mass with contrast-enhancing solid content in the medial side of this cystic lesion (**Figure 1**).

The patient did not report any specific complaints in his anamnesis and system query. In summary, there was no history of hematuria, and no remarkable finding was detected in the physical examination. Renal function tests, hemogram and complete urinalysis were requested from the patient. Laboratory examinations did not reveal any pathological findings, including microscopic hematuria. Upper abdominal magnetic resonance imaging (MRI) revealed a 5x4.5 cm cystic lesion in the lower pole of the left kidney and a 33x30 mm cyst in the medial side of this cyst, with early arterial enhancement which was interpreted as a Bosniak type 4 cystic mass with a contrast-enhanced soft tissue component in the late phase MRI examination (**Figure 2-3**).

There was no lymphadenopathy or metastasis on CT and MRI. Cyst excision was planned for the patient's cystic lesion and open nephron-sparing surgery for the medially situated solid lesion. However, during the surgery, it was determined that the mass, which was reported as a complete cyst in previous imaging studies, also had solid contents, and this lesion was also excised during nephron-sparing surgery. The cystic lesion was reported as 3.9x3.8x3.7 cm with multi-millimetric cystic areas on macroscopic and as cystic nephroma on microscopic examination (**Figure 4**). Solid lesion was reported as 2.2x2x1.9 cm clear cell renal cell carcinoma. There was no tumor at the surgical margins and was interpreted as pT1a (**Figure 5**).

Discussion

Cystic nephroma is a rare cystic disease of the kidney. It manifests as a benign, cystic, multilocular and usually unilateral lesion. It consists of epithelial and stromal components. In the literature, many terms such as cystadenoma, solitary multilocular cyst, benign multilocular cyst, and cystic hamartoma have been used to define this lesion [2]. Its etiology is not clearly known. More than two hundred cases have been reported in the literature after Edmunds' definition. There is a bimodal age distribution. It is most common in the first 2 years of life and after the third decade of life. Although there is a slight predominance of males in children, the number of female cases is significantly higher in adults [3]. Diagnostic histological criteria were first defined by Powell in 1951 and modified by Boggs and Kimmelstiel in 1956 [4]. According to this definition diagnostic criteria of cystic nephroma are as follows:

• a well-circumscribed mass with many cysts and septa,

• lack of connection of these cysts with each other and with the renal pelvis,

• the cysts do not contain a solid component, but there may be a solid component in the septa,

• cyst epithelium contains flat, cubic and hobnail cells,

• septa formed from well-differentiated renal tubular and fibrous tissues.

Cystic nephroma is usually asymptomatic and diagnosed incidentally. But it may manifest with abdominal mass in children and with abdominal mass, flank pain, urinary tract infection, and hematuria in adults. Our case was asymptomatic and cystic



Figure 1. Computed tomography image (Blue arrow: cystic nephroma; Red arrow: cystic lesion with solid component)

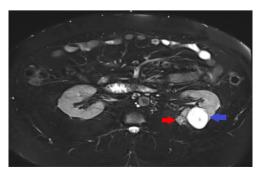


Figure 2. T1-weighted magnetic resonance image (Blue arrow: cystic nephroma; Red arrow: cystic lesion with solid component)

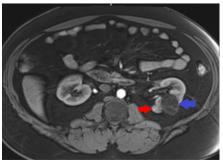


Figure 3. T2-weighted magnetic resonance image (Blue arrow: cystic nephroma; Red arrow: cystic lesion with solid component)

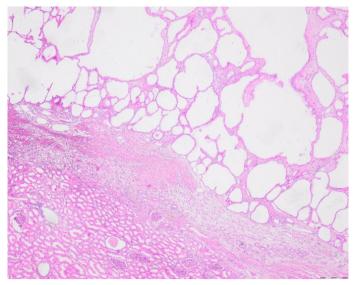


Figure 4. Microscopic examination of surgical specimen; cystic nephroma adjacent to the kidney parenchyma

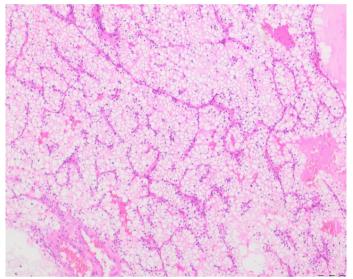


Figure 5. Microscopic examination of surgical specimen; clear cell renal cell carcinoma

nephroma was diagnosed as cortical cyst in CT and MRI taken urgently after a traffic accident. In the differential diagnosis, as seen in our case, non-neoplastic cystic kidney diseases, mixed epithelial-stromal tumor (MEST), multicystic renal cell carcinoma and nephroblastoma should be considered as well as cortical cysts. However, the definitive diagnosis cannot be made by imaging or physical examination. Radiologically, in cystic nephromas, the cyst may herniate into the collecting system [5]. Cystic nephromas are well circumscribed on computed tomography. In magnetic resonance, they show a hypointense signal in T1 and a hyperintense signal in T2-weighted images. Septa are usually hypointense due to its fibrous content. In the MRI examination of our patient, there was a hypointense appearance in T1 and a hyperintense appearance in T2-weighted images. The cyst had a thin capsule, and contained thin septations. The cystic lesion was herniated into the collecting system. There was no solid component. It is very difficult to distinguish cystic nephromas from Bosniak type 2 and 3 cysts by

imaging methods [6]. Therefore, histopathological examination is necessary for definitive diagnosis. Histologically, the cysts are lined with squamous, cuboidal and hobnail epithelium. Stromal content often stains positively with CD10, calretinin, inhibin, estrogen, and progesterone receptors. In cystic nephromas, the cystic component is more prominent than in MEST. In MEST, the cyst wall is thicker [7]. In a study of 7 patients with cystic nephromas seen in the pediatric age group, DICER1 mutation associated with anaplastic renal sarcomas was found in 86% of the patients [8].

Renal cell carcinomas are usually solid, while 4-7% are cystic. Cystic nephroma also gives the impression of a multilocular Bosniak type 3 or 4 cysts which complicates the diagnosis [9]. Although cystic nephroma is not malignant, since it is difficult to differentiate from Bosniak type 3 cyst, radical nephrectomy or nephron-sparing surgery is an option for its diagnosis and treatment [10].

Conclusion

We performed nephron-sparing surgery for the cystic nephroma case with two adjacent lesions in the lower pole of the kidney which could not be diagnosed by imaging methods and required histopathological examination of the surgical specimen for a definitive diagnosis. Our patient differs from other cases in that one of the 2 cystic lesions in the same kidney is renal cell carcinoma and the other is cystic nephroma.

Ethics Committee Approval: N / A.

Informed Consent: An informed consent was obtained from the patient.

Publication: The results of the study were not published in full or in part in form of abstracts.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions: Any contribution was not made by any individual not listed as an author. Concept - U.A., E.C.B.; Design – U.A., E.C.B.; Supervision – U.A., E.C.B., I.S.; Resources - M.O., C.C., B.O.; Materials - M.O., C.C., B.O.; Data Collection and/or Processing - U.A., M.O., C.C., B.O.; Analysis and/or Interpretation - U.A., E.C.B., M.O..; Literature Search - M.O., C.C., B.O.; Writing - U.A., E.C.B.; Critical Review – U.A., E.C.B., I.S.

Conflict of Interest: The authors declare that they have no conflict of interest.

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