Cystic angiomyolipoma of the kidney in a patient with chronic renal failure: a very rare variant

Kronik böbrek yetmezliği olan bir hastada böbreğin kistik anjiyomiyolipomu: çok nadir görülen bir olgu

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ABSTRACT

Angiomyolipomas are one of the most common benign tumors of the kidney and can affect vascular, smooth muscle and adipose tissue. Cystic angiomyolipomas have recently been described as a variant of the classic angiomyolipoma, and fewer than 20 cases have been reported in the English literature. A review of the English literature was conducted using the Pubmed/Medline database concerning cystic angiomyolipoma of the kidney. Cystic angiomyolipomas, or angiomyolipomas with epithelial cysts, are extremely rare, and fewer than 20 cases have been reported in the literature according to the Pubmed/Medline database. Herein, we report a case of cystic angiomyolipoma of the kidney in a 62-year-old male patient. The angiomyolipoma presented components of smooth muscle tissue, blood vessels and mature adipose tissue accompanying the epithelial cyst, which was lined by cuboidal cells. To the best of our knowledge, this is the first case report of cystic angiomyolipoma in Turkey. Cystic angiomyolipoma of the kidney is a very rare variant of classic angiomyolipoma with distinct pathological features.

Key words: Cystic angiomyolipoma; kidney; tumor.

ÖZET

Anjiyomiyolipomlar, böbreğin en sık görülen benign tümörlerinden olup, vasküler, düz kas ve yağ dokusunu kapsayabilmektedir. Kistik anjiyomiyolipomlar yakın zaman önce klasik anjiyomiyolipomların bir çeşidi olarak tanımlanmışlardır. İngilizce literatürde 20'den az sayıda olgu sunumu bildirilmiştir. Pubmed/Medline taranarak kistik anjiyomiyolipom ile ilgili İngilizce literatür araştırılmıştır. Kistik anjiyomiyolipomlar ya da epitelyal kistler içeren anjiyomiyolipomlar, çok nadir görülürler. Pubmed/Medline araştırmamıza göre, İngilizce literatürde 20'den az sayıda olgu bildirilmiştir. Bu makalede 62 yaşındaki bir erkek hastada saptadığımız kistik anjiyomiyolipom olgusu sunulmuştur. Bu anjiyomiyolipomda, küboid hücrelerden oluşan epitelyal kiste eşlik eden düz kas, kan damarları ve matür yağ dokusu komponentleri izlenmiştir. Bilgimize göre, bu olgu ülkemizde bildirilen ilk kistik anjiyomiyolipom olgusudur. Böbreğin kistik anjiyomiyolipomu, klasik anjiyomiyolipomuna göre çok nadir görülen ve kendine özgü patolojik özellikleri olan bir çeşitidir.

Anahtar sözcükler: Böbrek; kistik anjiyomiyolipom; tümör.

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Introduction

Angiomyolipomas are one of the most common types of kidney tumor and can be composed of thick-walled or dysplastic vascular structures, smooth muscle and adipose tissues.[1] Although angiomyolipomas are commonly benign in origin, rare cases of malignant transformation have been reported.[2,3] Angiomyolipomas are generally identified as solid masses in the kidneys, and the detection of fat by computerized tomography (CT) is commonly observed during radiological evaluation of these lesions.[4] However, some angiomyolipomas may not demonstrate fat content by CT, and these types are referred to as atypical or minimal fat angiomyolipomas.[4]

Cystic angiomyolipomas, or angiomyolipomas with epithelial cysts, are extremely rare lesions, and fewer than 20 cases have been reported in the literature according to the Pubmed database.[5-8]

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Herein, we report a very rare case of cystic angiomyolipoma of the kidney and discuss the clinical and pathological findings.

Case report

A 62-year-old male patient was admitted to our department with lower urinary tract symptoms. He had a history of right ureterolithotomy and diabetes, although no history of tuberous sclerosis, lymphangiomyomatosis, renal cancer or renal cyst was present. The physical examination was normal apart from the presence of a right flank incision scar. An enlarged benign prostate was detected upon digital rectal examination, and the patient also had chronic renal failure with increased blood urea nitrogen and creatinine levels. The abdominal ultrasound (USG) and CT results revealed a left kidney with a size of 200x77 mm, grade IV hydronephrosis, severe parenchymal loss and a ureteropelvic stone (18x17 mm). A 13-mm ureteropelvic stone and a 2-cm simple cortical cyst were also detected in the right kidney (Figure 1). Dynamic renal scintigraphy demonstrated a non-functioning left kidney. We performed left retroperitoneal simple nephrectomy and double J catheter insertion on the right kidney. The pathologic evaluation of the left kidney demonstrated cystic angiomyolipoma of the kidney (Figure 2). Currently, this patient is tumor-free without any local recurrence.

Discussion

In general, angiomyolipomas are solid tumors that demonstrate benign behavior. Among the published cases in the literature regarding cystic angiomyolipomas, no cases of malignant transformation have been reported on follow-up, even among patients who had received nephron-sparing surgery.[5-7] Thus, according to the published literature, cystic angiomyolipomas seem to have a benign disease course similar to classic angiomyolipomas.[5-8]

The differential diagnosis of cystic angiomyolipomas involves distinguishing between mixed epithelial and stromal tumors (previously classified as cystic hamartomas of the renal pelvis, adult mesoblastic nephroma, or renal pelvic or cortical hamartomas), cystic nephroma, multilocular cystic renal cell carcinoma, liposarcoma and vascular malformations.[7,8]

Although mixed epithelial and stromal tumors of the kidney are thought to be more common in females (or males with estrogen exposure),[8-10] our patient did not have a history of estrogen exposure. Both cystic angiomyolipomas and mixed epithelial and stromal tumors of the kidney can express estrogen or progesterone receptors, whereas the latter type typically does not express melanocytic markers.[8-10] Cysts lined by epithelium and the surrounding smooth muscle walls are common features of both cystic angiomyolipomas and mixed epithelial and stromal tumors of the kidney.[8]

Previous immunohistochemical studies of cystic angiomyolipomas demonstrated positive staining of the subepithelial stroma with melanocytic markers (HMB-45 and Melan-A), estrogen

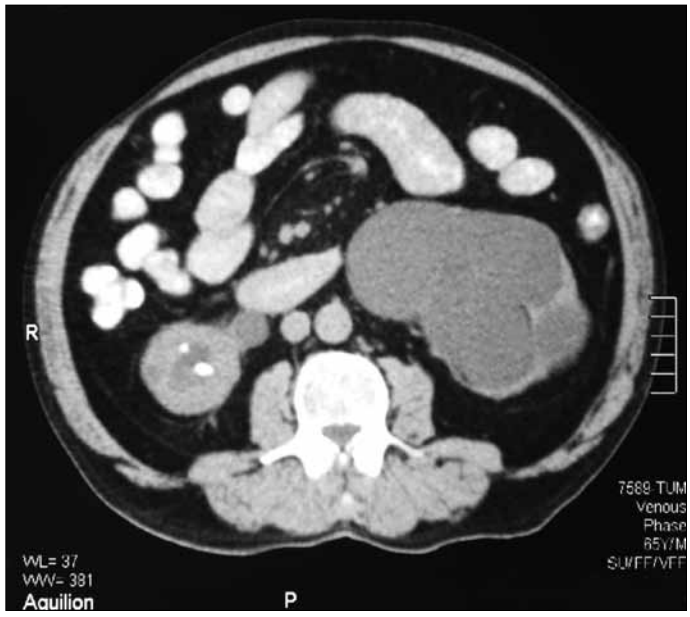

Figure 1. Abdominal computerized tomography showing a left kidney with severe hydronephrosis and parenchymal loss as well as a ureteropelvic stone.

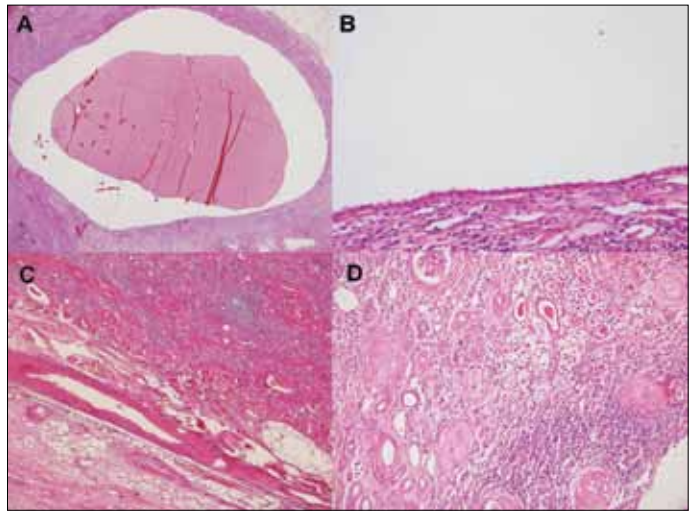

Figure 2. A) A large epithelial cyst in the tumor tissue, B) Epithelial cyst lined by cuboidal cells, C) The three tumor components consisting of smooth muscle tissue, blood vessels and mature adipose tissue, D) Prominent lymphoplasmacytic infiltrate and glomerulosclerosis (Hematoxylin-eosin stain, original magnification, X20, X 400, X100, X400).

and progesterone receptors and CD10.[7,8] Moreover, muscle staining with actin and desmin and cyst lining staining with pancytokeratin were also demonstrated.[7,8] Of the RCC marker antigens, such as inhibin, WT-1, c-kit (CD117), S-100 protein, and CK20, no positive staining was detected for any of the three components of the tumor (i.e., smooth muscle tissue, blood vessels and mature adipose tissue), and a low proliferative index value was detected according to Ki67 labeling.[7] Therefore, no distinct immunohistochemical staining profile has been identified for cystic angiomyolipomas.

Reported symptoms related to cystic angiomyolipomas include hematuria (microscopic, recurrent or gross), intermittent flank pain, retroperitoneal hemorrhage and acute abdomen due to blood vessel rupture.[5-8] In our case, the patient was diagnosed with cystic angiomyolipoma after being admitted to our outpatient clinic with lower urinary tract symptoms mostly relating to prostatic obstruction.

Although the histogenesis of cystic angiomyolipoma remains unclear, some authors have suggested a role for entrapped renal tubular elements in the etiology of this disease.[6] In our case, no cystic renal cell carcinoma or papillary renal cancer growth was detected upon microscopic evaluation.

In addition, a radiologic follow-up evaluation (abdominal USG and CT) was carried out for our patient due to the presence of a 2-cm simple cortical cyst and urinary tract stone disease.

Conflict of interest

No conflict of interest was declared by the authors.

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