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Interventional radiology for hemorrhage due to tuberous sclerosis associated angiolipoma rupture

Tüberoz skleroz ile ilişkili anjiyolipoma rüptürüne bağlı kanamada girişimsel radyoloji

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Abstract

Renal angiomyolipomas are rare tumors that may be seen with tuberous sclerosis and cause massive retroperitoneal hemorrhage. In this paper, we present a case with massive hemorrhage due to angiomyolipoma in the left kidney and treated with selective arterial embolization by interventional radiology. Even in cases of massive bleeding, the angiographic procedure can be successfully performed and thus the kidney can be spared.

Keywords: Angiography, angiomyolipoma, interventional radiology, tuberous sclerosis.

Öz

Renal anjiyomyolipomlar, tüberoz sklerozla birlikte görülebilen, masif retroperitoneal kanamaya neden olabilen nadir bir tümördür. Bu yazıda, sol böbrekte anjiyomyolipom nedeniyle masif hemorajisi olan ve girişimsel radyoloji tarafından selektif arter embolizasyonu ile tedavi edilen bir vakayı sunuyoruz. Masif kanama durumunda dahi anjiyografik girişim başarıyla uygulanabilir ve böylece böbreğin korunması sağlanabilir.

Anahtar Sözcükler: *Anjiyografi, anjiyomyolipom, girişimsel radyoloji, tüberoz skleroz.*

Introduction

Renal angiolipoma (RAML) is a rare tumor, it has a good prognosis in most cases and its radiological and histological features have been well characterized (1). Large RAMLs can develop micro and macro aneurysms that may be ruptured. This condition may be sudden and painful and it is sometimes life threatening. In 10% of cases massive retroperitoneal hemorrhage may lead to hypovolemic shock and it is known as Wunderlich's syndrome (2). We present a case of RAML associated with tuberous sclerosis (TS) and causing retroperitoneal hemorrhage which was managed by angioembolization.

Case Report

A 28-year-old woman was admitted to our university hospital emergency department in January 2016 due to sudden, severe pain in the left lumber region.

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Patient had history of tuberosis sclerosis and she had no admission previously to emergency services for any reason. In physical examination, there was defense in the left upper and lower quadrants of the abdomen. Butterfly patterned facial angiofibromas were seen in the area of the face which are typical in TS.

Normal motor mental development was observed. Vital signs were normal. Hematuria was not detected. Hemoglobin and hematocrit values were detected 8.9 g/dL and 26.8% respectively in routine blood tests and urea, creatinine, and blood electrolyte levels and bleeding profile were found normal. In the whole body computerized tomography both kidneys were observed in vascular and fat rich view with multiple giant angiomyolipomas and complicated hematoma was detected in the left kidney upper pole. Mass lesion was observed neighborhood to the left lower pole which is considered to be a hypervascular angiomyolipoma component. Widespread sclerotic lesions in the vertebral bodies were also found. Axial CT scan shows angiomyolipoma in both kidneys with characteristic fatty components.

Vital signs proceeded stable at follow-up and hemoglobin and hematocrit values fell to 7.4 g/dL and

24.1%, respectively 48 hours after the patient's emergency admission. The patient was transfused two units of packed red blood cells. Hemoglobin value and hematocrit value rose up to 8.9 g/dL and 28% level, respectively.

The patient discharged from the emergency department without any emergency intervention and with no significant change in hemogram profile in the next 48 hours.

The next day the patient was admitted to our urology clinic with angioembolization plan. The patient's left upper pole renal artery embolization procedure was performed by the interventional radiology section (Figure-1).

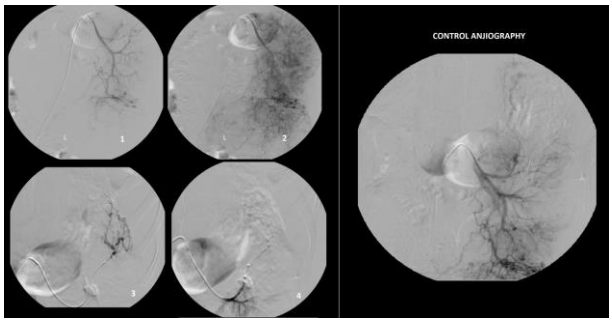

Figure-1. Left upper pole renal artery embolization procedure of the patient.

After the procedure, patient was monitored in the intensive care unit of our clinic. Hemogram values did not decline in 72 hours follow-up period. Patient was discharged to be followed up one month later and afterwards yearly with magnetic resonance imaging.

Written informed consent was obtained from the patient for publishing the individual medical records.

Discussion

RAML is a subset of perivascular epithelioid cell tumors with epithelioid tumor differentiation (3). RAML can be seen together with TS or pulmonary lymphangioleiomyomatosis (LAM) and can also occur sporadically without both. 80% of RAMLs are sporadic and there is no relationship with any genetic syndromes. However, it is thought that TS is found in about 10% of RAML diagnosis (4).

Sporadic RAML is most commonly seen in middle-aged women. The most serious complications are retroperitoneal hemorrhage (Wunderlich syndrome), hematuria and renal function disorder. Wunderlich syndrome can occur classically during pregnancy and hemorrhagic shock can be observed in 20% of patients (4). Sporadic RAML does not usually have symptoms and detected incidentally during renal imaging as a single lesion.

Unlike the sporadic RAML, multiple lesions in both kidneys are seen in RAML associated with the TS (5). Slow-growing sporadic RAML usually does not lead to deterioration of renal function (6). In the differentiation of patients with TS characteristic skin lesions in patients with RAML and the presence of other symptoms of TS should be considered such as benign tumors in multiple organs. Only 10% of RAML patients are associated with TS, but all patients should be evaluated for subclinical or undiagnosed TS. Sporadic RAML, compared with RAML associated with TS usually occurs in older age, being single, rarely causes symptoms and hemorrhages and grows more slowly (6). Active intervention is required more often in TS associated RAML patients compared to sporadic RAML patients.

Surgical treatment is suggested for patients with high suspicion of malignancy with intramural necrosis and / or calcification (7). Available interventions include nephron-sparing surgery (NSS), selective renal artery embolization, total nephrectomy and radiofrequency ablation. Intervention may vary according to the patient. NSS is an important option rather than total nephrectomy in 7-10 cm sized tumors and patients with multiple RAMLs (8). Selective renal artery embolization or total nephrectomy may be a viable option for tumors unsuitable for NSS due to tumor location, size, hemorrhage or urinary fistula risk. Self-limiting postembolization syndrome developed in 35.9% of patients in a study with 524 patients with RAML managed with transarterial embolization. Mortality has not developed in this patient group depending on the embolization. Tumor size reduction of 38.3% (mean 3.4 cm) was determined at average 39-month follow-up after embolization. Unplanned embolization or surgery was necessitated in 20.9% of patients during follow up period. Re-operation etiology includes RAML revascularization (30%), constant or increasing tumor size (22.6%), refractory or recurrent symptoms (16.7%) and the emergence of acute retroperitoneal hemorrhage (14.3%) (9).

According to the European Association of Urology (EAU) guidelines, active surveillance would be the most appropriate approach for RAML [Grade of Evidence (GE): 3].

Selective arterial embolization is recommended as first-line therapy whether active monitoring will be discontinued (GE: 3). If surgical treatment is preferred, many patients can be managed by NSS, complete nephrectomy is indicated in some patients (GE: 3). Radiofrequency ablation can be used as another option. Moreover the tumor volume can be reduced with mTOR inhibitors (sirolimus and everolimus) and surgery may be delayed with this treatment (10).

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