Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2018;43(3):772-774
DOI: 10.17826/cumj.396955

EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Renal angiomyolipoma rupture

Renal anjiomiyolipom rüptürü

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Cukurova Medical Journal 2018;43(3):772-774

Dear Editor,

Renal angiomyolipoma is a rare benign tumour that is rarely seen in the emergency department. Angiomyolipoma is most often seen as a solitary lesion, but it is more likely to be bilateral in some syndromes, especially tuberous sclerosis. However, it can be seen without any cause. The incidence is higher in women. The expansion of the tumour may result in a rupture. It is a life threatening condition and delayed diagnosis and treatment can increase mortality. Patients may be referred to emergency department with severe abdominal pain, bleeding and shock[1,2]. We aimed to present a 32-year-old woman with acute abdominal pain and shock due to renal angiomyolipoma rupture.

A 32-year-old woman presented to the emergency department with fatigue and acute right flank pain. Fever, nausea, vomiting, anorexia, constipation was not present. Only for current pain, she used single dose of non steroidal anti inflammatory drug about four hours ago. Hemodynamic parameters; blood pressure was 80/60 mmHg, pulse was 130/min and O_2 saturation was 92 % in room air. On examination, she seemed pale and a solid, sensitive mass was palpated on the right flank. There were no defence and rebound tenderness. Blood test revealed a haemoglobin level of 7,6 g/dl, a leukocyte count of 12100 /ul, a thrombocyte count of 225.000/ul .Other routine biochemical values and APTT, prothrombin time, INR, fibrinogen values were within normal limits. Emergency performed ultrasonography (USG) showed that a large uniformly hyperechoic mass was found in the upper pole of the right kidney with acoustic shadowing. Intravenous (IV) contrast enhanced computerized tomography (CT), well-demarcated mass of the upper pole of the right kidney and also left kidney with mixed soft tissue and fatty tissue elements was observed. Mass was measured 14,5 x 9,5 x 15,5 cm on the right side and 8,5 x 6 x 10,5 cm on the left side. After administration of contrast massive recent haemorrhage were detected on the right side. Also findings were showed suspicious of a bleeding renal angiomyolipoma (Figure 1).

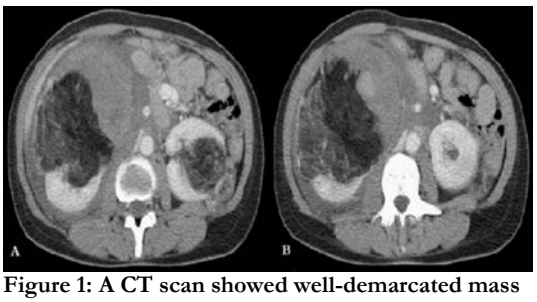

Figure 1: A CT scan showed well-demarcated mass of the right kidney with mixed soft tissue and fatty tissue elements (A) and after administration of contrast massive retroperitoneal haemorrhage were detected on the right side (B).

Supportive therapy was initiated with ventilation, monitoring, and central venous access as well as two large catheters in peripheral lines. Fluid treatment a 0.9% sodium chloride injection solution was initiated. Erythrocyte suspension was requested. When the patient with renal angiomyolipoma bleeding was diagnosed as hemodynamically unstable, selective renal angiography and selective

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Geliş tarihi/Received: 14.09.2017 Kabul tarihi/Accepted: 03.10.2017

embolization were planned. The right common femoral artery approach is used for percutaneous renal intervention. After renal arteriography, selective embolization of the upper pole branch of the renal artery was performed with coils by interventional radiology department. The angiogram after the embolisation procedure showed normal renal parenchyma and remaining embolized tumor vasculature. (Figure 2) later in time, selective embolisation was performed against the left kidney angiomyolipoma.

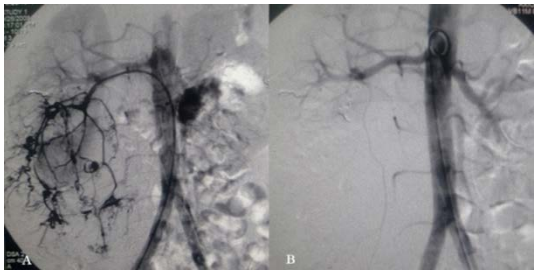

Figure 2. Selective right renal arteriogram shows tumour vessels and active bleeding in the upper portion of the kidney (A) and tumour vessels and active bleeding disappear after embolization (B).

Patient was treated and followed 7 days and then discharged from hospital. She was readmitted about 10-11 weeks later for control. The control CT demonstrated that a decrease in the size of the angiomyolipoma.

Renal angiomyolipoma is a rare benign tumour composed of smooth muscle, fatty tissue and tortuous blood vessels. The most common is the kidney, with less frequent liver, lymph nodes, spleen, lung, and retroperitoneal area. Angiomyolipoma is most often seen as a solitary lesion, but it is more likely to be bilateral in some syndromes, especially tuberous sclerosis. Differential diagnoses include renal cell carcinoma, spontaneous rupture of the kidneys with hydronephrosis, and spontaneous rupture of the aneurysm[1,2,3]. Our patient was young and female in accordance with the literature. Despite bilateral angiomyolipoma, there was no tuberosclerosis on physical examination and computed tomography.

Symptoms of the disease include abdominal pain, palpable mass, and haematuria (Lenck triad). In addition, fever, nausea-vomiting, anaemia, renal insufficiency and hypotension can also be seen. Hemorrhage is common in angiomyolipoma. The risk of bleeding in angiomyolipomas larger than 4 centimetres increases. The wall of vessels in angiomyolipoma is thicker than normal renal veins but less elastic. For this reason they tend to bleed without trauma. Spontaneous rupture in large angiomyolipomas is a serious medical condition. The patient is confronted by sudden onset of abdominal pain, hypotension and shock[4,2]. In our patient was presented as an emergency with a flank pain, hypotension, and hypovolemic shock.

Radiological imaging methods in angiomyolipoma diagnosis is very important. Often angiomyolipomas can be diagnosed by various imaging modalities. The classic ultrasonography image is a hyperechogenic mass and an acoustic shadow. CT is a preferred imaging technique for showing the presence of a renal mass containing perirenal and retroperitoneal bleeding due to spontaneous angiomyolipoma rupture[2]. In our patient, ultrasonography showed large uniformly hyperechoic mass of the upper pole of the right kidney with acoustic shadowing. Abdominal tomographic scans showed well-demarcated mass of the upper pole of the right kidney and also left kidney with mixed soft tissue and fatty tissue elements. After administration of contrast massive recent haemorrhage were detected on the right side

Treatment options for angiomyolipoma rupture include partial or total nephrectomy, cryoablation, radiofrequency ablation, and selective arterial embolization. Selective arterial embolization is an ideal and effective treatment option in the presence of acute bleeding or life-threatening cases[5,6]. The clinical diagnosis of our patient was of right renal angiomyolipoma rupture. The size of this angiomyolipoma markedly reduced due to transcatheter arterial embolization. It is an effective therapy for rupture of renal angiomyolipoma.

Ruptured renal angiomyolipoma is a rare presentation of acute flank pain in the emergency department. In a young patient, if there are flank pain, hypotension and signs of hemorrhagic shock, renal angiomyolipoma rupture and hemorrhage be kept in mind. Late diagnosis and treatment can be fatal.

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