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Case Report / Olgu Sunumu

Wunderlich's syndrome as the fatal complication of a renal angiomyolipoma: a case report and review of the literature

Renal anjiyomiyolipomun ölümcül komplikasyonu olarak Wunderlich sendromu: olgu sunumu ve literatürün gözden geçirilmesi

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ABSTRACT

Wunderlich's syndrome is a rare acute abdominal condition, which is characterized with spontaneous nontraumatic renal hemorrhage into the subcapsular and perirenal spaces. Renal cell carcinoma, angiomyolipoma, renal cysts, vasculitis or blood dyscrasias are among the known risk factors. We reported a case of renal angiomyolipoma presenting as Wunderlich syndrome.

Keywords: Wunderlich's syndrome, angiomyolipoma, computed tomography

ÖZ

Wunderlich sendromu, subkapsular ve perirenal boşluklara spontan travmatik olmayan renal hemorajiyle karakterize nadir görülen akut karın hastalığıdır. Renal hücreli karsinom, anjiyomiyolipom, renal kistler, vaskülit veya kan diskrazileri bilinen risk faktörleri arasındadır. Wunderlich sendromu'na neden olan renal anjiomyolipom olgusunu sunduk.

Anahtar Kelimeler: Wunderlich's sendromu, anjiyomiyolipom, bilgisayarlı tomografi

INTRODUCTION

Wunderlich's syndrome (WS) is a rare acute abdominal condition, which is characterized with spontaneous nontraumatic renal hemorrhage into the subcapsular and perirenal spaces (1). Angiomyolipoma is the most frequent cause of spontaneous renal hemorrhage, due to its rarity incidence is not properly understood (2). We reported a case of renal angiomyolipoma presenting as Wunderlich syndrome.

CASE REPORT

Fifty nine year old woman admitted to our emergency service complaining from sudden right-sided abdominal pain and dizziness. Formerly known angiomyolipoma on the right kidney was noted while medical history was taking. There was no significant past family history. On physical examination pain in the flank was present and blood pressure was measured as 110/70 mmHg. Laboratory findings were normal. Abdominal ultrasonography was performed. A

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heterogeneous, solid mass with a maximum diameter of 19 cm, originating from the right kidney was sonographically detected. Because of the previously known angiomyolipomas of the patient a dynamic abdominal computed tomography (CT) was suggested. The solid lesion, which also contains lipid tissue of about 70 mm in diameter, filling the lower portion of the right kidney was seen on the dynamic examination (**Figure 1**). Prompt arterial enhancement was not detected on dynamic examination. There was also a huge retroperitoneal hematoma extending to the pelvis. Since the hemodynamic parameters of the patient were stable, selective embolization of the renal artery was not performed. Blood pressure, hemogram and sonographic follow-up were applied. on control CT after 5 and 8 months; the hematoma was completely resolved while the lesion size and characteristics were the same (**Figure 2**).

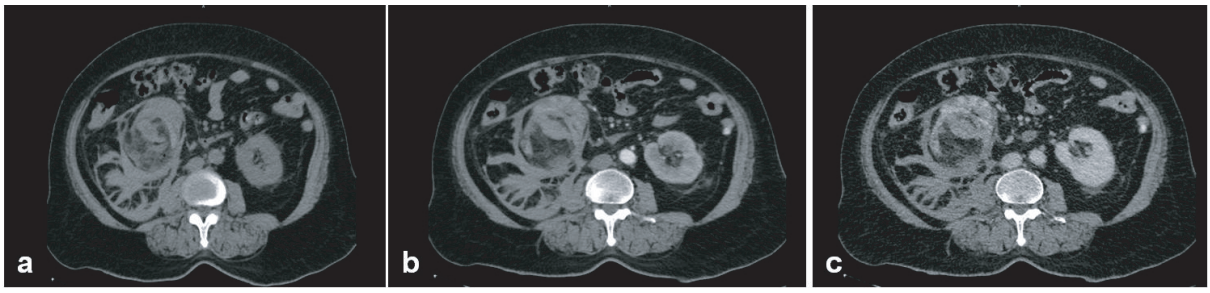

Figure 1. On axial images of the dynamic abdomen CT **(a)** Non-enhanced is revealed that the solid lesion filling the lower portion of the right kidney. There is also a huge retroperitoneal hematoma extending to the pelvis. **(b)** On arterial phase prompt arterial enhancement is not detected. **(c)** On the late phase; no wash- out is seen within the lesion.

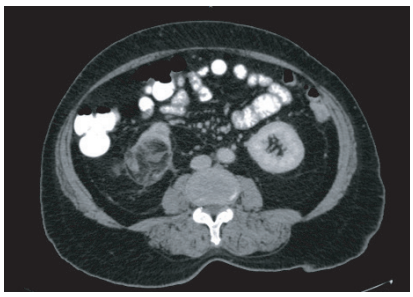

Figure 2. After 8 months axial images of control abdomen CT has shown completely resolved hematoma while the lesion size and characteristics were the same.

DISCUSSION

Spontaneous renal hemorrhage, which is known as WS, may lead to a life-threatening clinical manifestation by causing fulminant hypovolemic shock. Renal cell carcinoma, angiomyolipoma, renal cysts, vasculitis or blood dyscrasias are among the known risk factors. The increased proportion of angiogenic component and whether underlying tuberous sclerosis is an important predisposing for spontaneous hemorrhage in angiomyolipomas, which is the most frequent cause of WS (3). In patients with tuberous sclerosis, angiomyolipoma has a predilection for females in the third and fourth decades of life and lesions generally very large, asymptomatic, multifocal, and bilateral. In sporadic cases, angiomyolipoma is encountered in an elderly woman and is often symptomatic, and unilateral (4). In this case, our patient was older female and sporadic.

Clinical presentation may vary from slight dizziness to fulminant hypovolemic shock according to the degree of the bleeding. side-wall pain, palpable abdominal mass, and hypovolemic shock are seen in only 20% of all cases; which is a characteristic presentation of WS (5). Medda et al (6) reported that there is a higher bleeding risk and the more severe clinical picture is presented as the lesion diameter increased. The majority of angiomyolipoma sonographically seen as hyperechoic lesions secondary to fat component of the lesion. Establishing the angiomyolipoma diagnosis can be really challenging and the lesion may mimic malign tumor, since the hemorrhage occurred within angiomyolipoma in patients with WS (7). The fat component can be seen in unenhanced computed tomography as hypodense areas. With and without fat suppressed MRI sequences are useful in demonstrating a fat component of angiomyolipomas. Angiomyolipomas usually do not contain calcifications different from clear cell renal carcinoma. The presence of macroscopic adipose tissue demonstrated with CT or magnetic resonance imaging and lack of calcification is helpful in diagnosing angiomyolipomas, since calcifications within angiomyolipomas are uncommon. In the setting of WS, a benign mass lesion with a heterogenous density extending perinephric space and retroperitoneal region can tomographically be seen. There is no wash out seen within the hemorrhagic mass lesion on dynamic images. Differential diagnosis includes renal cell carcinomas, vasculitis, renal vein thrombosis, cystic renal diseases, calculus disease, nephri-

tis. Regarding the rarity of WS, varying etiology and fatal consequences; the diagnosis and management of WS can be challenging. It is important to reporting further cases, since it is difficult to differentiate such rare syndromes. Treatment options are conservative or surgical (embolization, nephrectomy or evacuation of hematoma). In our patient, clinical findings limited itself and hemodynamic parameters of the patient were stable, selective embolization of the renal artery was not performed.

In conclusion, WS is a rare, life threading condition leading to high morbidity and mortality. A proper suspicion in the setting of the aforementioned clinical picture and being familiar with this unusual clinical condition is important to obtain an accurate diagnosis.

DECLARATION OF CONFLICTING INTERESTS

The author declared no conflicts of interest with respect to the authorship and/or publication of this article.

ETHICS

Institution and patient approval was obtained.

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