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Case report

Wunderlich syndrome: a rare case in a young woman

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Abstract: We report the case of a 29-year-old woman with Wunderlich syndrome, a rare spontaneous renal hemorrhage into the subcapsular and perinephric space. She presented to our emergency department with a sudden and persistent right flank pain in the abscence of abdominal injury. The onset of the symptoms could be insidious and lead to hypovolemic shock. Computed Tomography helps both in the diagnosis, detecting the renal hemorrhage, and contributes to an optimal patient management. Selective arterial embolization is an efficient technique to stop acute and potential life-threatening hemorrhage and preserve the renal parenchyma. (www.actabiomedica.it)

Keywords: Wunderlich syndrome, Angiomyolipoma, Hemorrhage, Kidney

Introduction

Wunderlich syndrome (WS) is a rare and life-threatening condition characterized by spontaneous renal hemorrhage into the subcapsular and perinephric space in patients with no history of trauma (1). Tumors represent the most common etiologies including renal angiomyolipoma and renal cell carcinoma (1,2). Other causes are vascular abnormalities, renal cystic diseases, inflammatory and infectious processes and in some cases no predisposing condition can be found (3). The onset of the symptoms could be rapid and lead to hypovolemic shock (4). This paper presents a case of WS in a 29-year-old female patient showing the insidiousness of this pathology and its clinical course.

Case presentation

A 29-year-old woman presented to our emergency department with a sudden and persistent (3-4 hours) right flank pain which at first radiated also to the ipsilateral groin. The other associated symptoms were nausea and vomiting. There was neither history of trauma nor significant medical and family history. On physical examination, she was pale with a blood pressure of 112/78 mmHg and tenderness on the right flank was observed. Right renal colic was suspected. Laboratory investigations showed haemoglobin of 10,3 g/dL, red blood cell count of 3,55x10^6mcL, C-reactive protein of 8,9 mg/dL while liver and renal function were in the normal range. The urine analysis showed hematuria and pyuria. Abdomen ultrasonography was performed, revealing a hyperechoic mass in the right renal region (Figure 1). The patient's symptoms worsened thus an urgent abdominopelvic contrast-enhanced Computed Tomography (CT) was requested. It showed a 6,3x5x8 cm mass that compressed the right kidney laterally, characterized by both hypervascularized and adipose areas, referable to a hemorrhagic angiomyolipoma (Figure 2, A,B). The lesion was associated with free fluid in the perirenal space, lateroconal fascia and pelvic region. Moreover, an active extravasation of contrast media was observed (Figure 3). She was diagnosed with a non-traumatic hemorrhage in the perinephric space, thus a WS. Emergency angiography was performed and the branch of the right renal artery, which feeded the angiomyolipoma, was

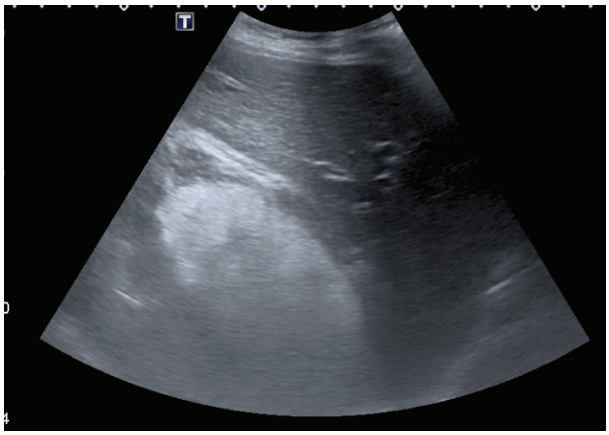

Figure 1. Abdominal ultrasonography which shows a hyperechoic mass in the right renal region

successfully embolized. Following embolization, the patient remained hemodynamically stable and her general condition improved. After 7 days, in the follow-up CT the hemorrhagic mass was reduced in dimensions and no signs of extravasation were observed (Figure 4). She was discharged home the day later.

Discussion

WS is a rare syndrome characterized by spontaneous renal haemorrhage into the subcapsular and perinephric space (1). It was described clinically for the first time by Carl Wunderlich in 1856 (3). The presentation of this syndrome may vary depending on the degree and duration of the bleeding (4). The classic symptomatological presentation is represented by an insidious onset of flank or abdominal pain, a palpable tender mass and symptoms of internal bleeding (Lenk's triad) (5). Other symptoms include vomiting, nausea, hematuria, anemia, hemodynamic instability till hypovolemic shock, rendering WS a life-threatening condition. Renal tumors are the most common cause of WS that account for up to 60% of all cases (6). Other causes include vascular abnormalities, renal cystic diseases, inflammatory and infectious processes and in some cases no predisposing conditions can be found (3). Angiomyolipomas, which are renal tumors composed of abnormal blood vessels, smooth muscle, and mature adipose tissue, are the most common benign cause of WS (2). The major risk factor for bleeding is the tumor size (>4cm) (7) because there is an increase in blood flow, causing vessel dilatation and pseudoaneurysm formation (8) and, thus, a higher risk of rupture. Abdominal ultrasonography is often the initial imaging of choice but findings have to be confirmed with CT (9). Therefore, CT plays an

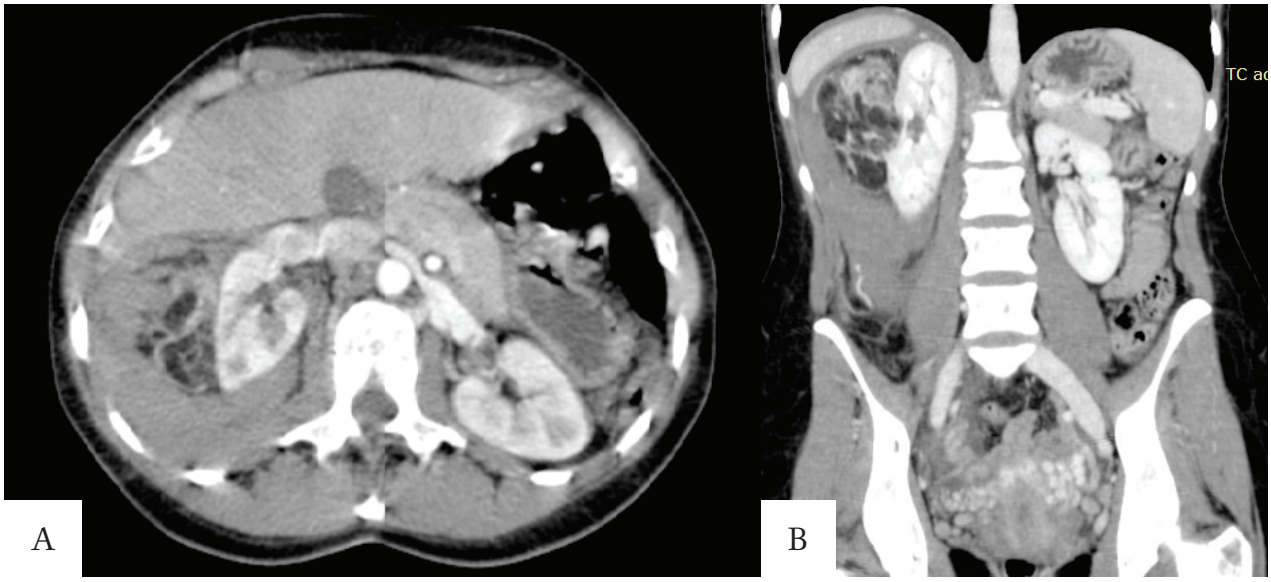

Figure 2. Contrast-enhanced abdominal CT scan in arterial phase with (A) axial and (B) coronal views which show a 6,3x5x8 cm mass that compressed laterally the right kidney characterized by hypervascularized and adipose areas with perirenal hematoma; free fluid in the lateroconal fascia and pelvic region is associated

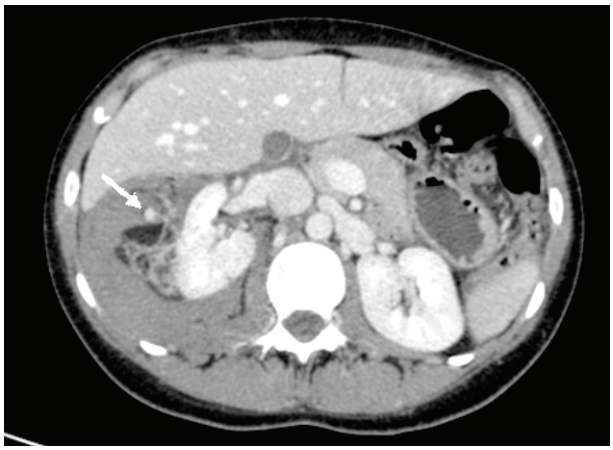

Figure 3. Axial contrast-enhanced abdominal CT scan in portal phase which shows an active extravasation of contrast media (arrowed)

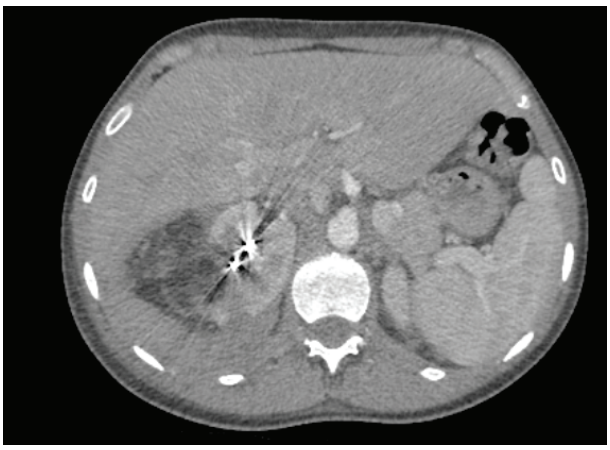

Figure 4. Follow-up axial contrast-enhanced abdominal CT scan which shows the hemorrhagic mass reduced in dimensions without signs of extravasation

important role in the diagnosis of WS and leads to an optimal patient management. Selective arterial embolization is an efficient technique to stop acute and potential life-threatening hemorrhage and preserve the renal parenchyma (10,11). Nevertheless, surgical treatment is preferred in cases of renal malignancy or hemodynamic instability (12). In conclusion, in this report we presented a rare case of a young patient with WS. Considering the insidiousness of this condition, a prompt diagnosis and timely treatment are important to prevent life-threatening complications and improve patient outcome.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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