

Wunderlich Syndrome presenting as Spontaneous renal haemorrhage in the retroperitoneal cavity as the first manifestation of renal cell carcinoma

Zespół Wunderlicha objawiający się samoistnym krwotokiem nerkowym w jamie zaotrzewnowej jako pierwsza manifestacja raka nerkowokomórkowego

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Streszczenie

Wstęp: Zespół Wunderlicha odnosi się do spontanicznego krwotoku okołonerkowego, często związanego z patologią nerek. Najczęstszą przyczyną krwawienia jest naczyńniakomięśniakotłuszczak. Rzadziej występuje w wyniku polekowego zapalenia naczyń, raka nerki, guza chromochłonny czy torbiel nerki.

Opis przypadku: Przedstawiamy przypadek samoistnego pęknięcia nerki w następstwie raka nerkowokomórkowego, objawiającego się ostrym bólem lędźwiowym po stronie prawej.

Wnioski: Wyniki badań obrazowych nie są charakterystyczne. Tomografia komputerowa pozostaje badaniem z wyboru, ponieważ może pokazać leżącą u podstaw patologię.

Słowa kluczowe: zespół Wunderlicha, krwotok zaotrzewnowy, rak nerkowokomórkowy, krwiak nerki, operacja oszczędzająca nerk

Abstract

Background: The Wunderlich Syndrome refers to spontaneous perirenal hemorrhage often associated with underlying renal pathology. The most common cause of bleeding is angiomyolipoma. Less often are drug-induced vasculitis, kidney cancer, pheochromocytoma or renal cyst.

Case Report: We present a case of spontaneous kidney rupture secondary to renal cell carcinoma presenting as an acute right loin pain.

Conclusions: The CT and US images are not characteristic. Computed Tomography remains the investigation of choice, as it might show the underlying pathology.

Key words: Wunderlich syndrome, retroperitoneal haemorrhage, renal cell carcinoma, renal haemorrhage, nephron sparing surgery

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Background

Spontaneous, nontraumatic retroperitoneal hemorrhage or Wunderlich syndrome (WS) is a rare but potential life-threatening condition. It can prove fatal if not recognized and treated aggressively at the appropriate time. A shock caused by massive retroperitoneal or intraperitoneal haemorrhage may develop in isolated cases [1]. The clinical findings are not specific, which is why the differential diagnosis must be made with a multitude of benign and malignant renal masses until the correct diagnosis can be made by the pathologist. The CT and US images are not characteristic. The most common cause of bleeding is angiomyolipoma. Less often are drug-induced vasculitis, kidney cancer, pheochromocytoma or renal cyst. We present the case of 53-year old man with symptoms of renal haemorrhage in the retroperitoneal cavity as the first manifestation of renal cell carcinoma.

Case Report

A 53 year old man, was admitted to Emergency Department in a District Hospital following an acute right loin pain after working within lawnmower few days ago. He was also complaining of generalised abdominal pain. He had no complaints of fever, vomiting or haematuria. He had no history of any specific disease or trauma. He was not taking any anticoagulation. On physical examination, the patient was hemodynamically stable, he had tachycardia (pulse = 124 per minute), blood pressure of 151/110 mmHg without pallor. Per-abdominal examination revealed fullness over right flank with tenderness over right flank, lumbar and iliac regions. Initial investigations showed a low haemoglobin at 12.4 g/dl with haematocrit 33.9%, raised white cell count at $16.8 \times 10^3/\mu\text{l}$, raised C reactive protein at 108 mg/dl, serum creatinine was 0.9 mg/dl, platelet count was $196 \times 10^3/\mu\text{l}$, Na 133.7 mmol/l, K 4.22 mmol/l. Ultrasonography of abdomen revealed: The right kidney poorly visible, partially obscured by gases. Left kidney of normal size and echogenicity, both kidneys without urine retention. Thickening of cortex layer with its simultaneous emphasizing in the area of the right kidney is visible, it can suggest Bertin's column hypertrophy – however, coexistence of pathology cannot be excluded. Additionally, in the middle part of this kidney there is a minor hyperechoic area, which may correspond to calculus. NCCT image of the abdomen and pelvis showed: No presence of free-air in abdominal cavity. Small amount of liquid present on the right prerenal fascia and in the lesser pelvis. Irregular, heterogeneous cortex layer of the right kidney, presence of hyperdense areas within it cannot be excluded. Around the abdominal labium of right kidney a hyperdense fluid rim is visible, measuring up to 30mm thickness –as likely as a blood (Fig. 1, Fig 2).



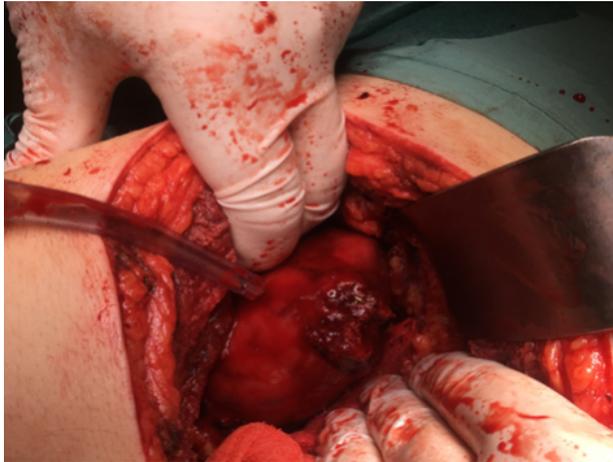
Ryc. 1. Around the abdominal labium of right kidney a hyperdense fluid rim is visible



Ryc. 2. Around the abdominal labium of right kidney a hyperdense fluid rim is visible, as likely as blood

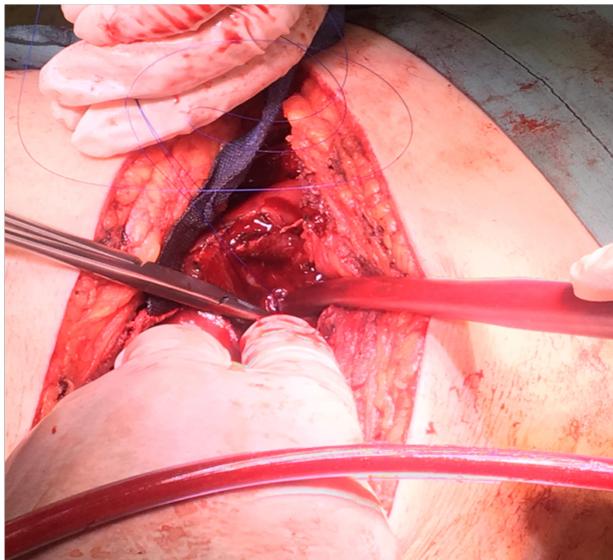
Apart from that, in the retroperitoneal fat there are numerous bands concentrations and a hyperdense fluid stains, these changes extend from diaphragm to groin. The collecting system of both kidneys is not enlarged. Twelve hours later, at the end of the day patient was transferred to Urology Department. He was hemodynamically stable. He remained under observation, conservative treatment was chosen, including the monitoring of vital signs, blood testing and repeated ultrasound scans. He was administered intravenous fluids and a broad-spectrum antibiotic. Because of the significant decrease in hemoglobin 9.5

g/dl and two episodes of high temperature level above 38°C, patient underwent an exploration of right lumbar area (Fig. 3).



Ryc. 3. Hemorrhagic renal mass involving midrenal area revealed after hematoma evacuation from right retroperitoneal area

The haematoma was evacuated from the right retroperitoneal area and a hemorrhagic renal mass involving midrenal area was revealed. The decision was made to treat the patient with nephron sparing surgery, and nearly 4cm tumor was excised (Fig. 4).



Ryc. 4. Image after the tumor excision. Ligation of the blood vessels

The right retroperitoneal area was packed and the wound was closed. In second day post operation, the packing material was outplaced in general anesthesia. Patient did well post operatively and was discharged home one week later. Histological examination showed a variant of RCC — Papillary renal partially mucinous tubular and spindle cell carcinoma Fuhrman II, as the cause of the renal bleeding.

Discussion

As indicated by the data available in the literature, nephrectomy is performed in around 68% of patients with spontaneous perirenal haematoma. The selection of such a procedure reflects the conviction of numerous urologists that the cause of haematoma development may involve an undiagnosed neoplasm [2]. However, conservative management using CT and biopsy may prevent unnecessary nephrectomy in patients with a benign disease and a stable condition [3, 4], permitting the possibility of nephron saving nephrectomy or selective renal arterial embolization if necessary. Management is largely influenced by the haemodynamic stability, aetiology of renal rupture, availability of facilities such as an angiography suite or operating theatre and surgeon's judgement.

Conclusions

Wunderlich syndrome, defined as spontaneous, non-traumatic renal haemorrhage into the subcapsular space, is an uncommon condition which was first reported in 1856 [5] and can only be found in small case series in the literature [6, 7]. Symptoms can range from mild to life-threatening [8]. Symptomatic patients may present with Lenk's triad: flank pain, palpable tender mass, and macrohematuria [9]. Hypovolemic shock occurs later in the course of this disease [10]. The most common cause for WS are neoplasms, less common aetiologies include blood dyscrasias and anticoagulation. Renal Angiomyolipoma (AML) has been reported to be the most common benign renal neoplasm and renal cell carcinoma is the most common malignant neoplasm to cause WS [7]. With reference to AML, larger lesions (>4 cm) have a higher incidence of spontaneous bleeding [11]. In the case of renal carcinoma, there is no relation described in the literature between the tumour size and the advancement of renal haemorrhage [12]. Ultrasound is extremely valuable for the immediate identification of spontaneous retroperitoneal haemorrhage. It enables the imaging of abnormal masses which surround the ruptured kidney, but it fails to clearly define the primary cause of the rupture. Sometimes spontaneous retroperitoneal haemorrhage might be misdiagnosed as a renal tumor or abscess [13]. Therefore Ultrasound findings have to be confirmed on a CT scan. Computed Tomography remains the investigation of choice, as it might show the underlying pathology. CT also gives valuable information about the contralateral kidney [14]. Specificity and sensitivity of diagnosis in cases of renal pathologies in kidneys with haematoma are 0.57 and 0.82, respectively, for CT, and 0.11 and 0.33 for US [2, 15].

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