Wunderlich syndrome – good imaging avoids bad surgery: a case report and short review

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KEY WORDS

kidney rupture ◗ RCC ◗ Wunderlich syndrome ◗ CT

ABSTRACT

Spontaneous renal hemorrhage, known as Wunderlich syndrome, is a rare clinical condition. The most common cause is a tumor – benign or malignant. Other causes are: rupture of a renal cyst, vasculitis, hydronephrosis, pre-eclampsia, and kidney infections. Although rare, this syndrome may cause life-threatening hemorrhagic shock – it requires prompt diagnosis and treatment. Radiologic imaging plays an essential role in the diagnosis, although often misleading and insecure about the underlying entity. We report a case of Wunderlich syndrome caused by a renal cell carcinoma in a patient on anticoagulant treatment. A short review based on the literature is also presented.

INTRODUCTION

Non-traumatic retroperitoneal hemorrhage is an uncommon but dramatic, even life-threatening, clinical entity. It can be due to rupture of an abdominal aortic aneurysm, adrenal bleeding, coagulopathy, hypertension and, rarely, renal disorders [1]. Spontaneous retroperitoneal renal hemorrhage, also known as Wunderlich syndrome [2], is usually associated with underlying diseases of the kidney. It may affect either the collecting system of the kidney or the renal parenchyma. Rupture of the collecting system usually can be managed by renal drainage, while rupture of the kidney parenchyma, accompanied by severe renal hemorrhage, may demand surgical exploration. In patients with ruptured renal cancer, total nephrectomy should be performed. Radiologic imaging is essential for diagnosis, but it is often misleading both in terms of recognition of the rupture and identification of its etiology.

We present a case of Wunderlich syndrome in which the initial radiologic diagnosis of pyonephrosis had to be revised and the therapeutic strategy changed.

CASE REPORT

A 50-year-old man (NLN, medical record 963/10.01.2009) was referred to our hospital with complaints of a right flank pain for a few days, a single episode of macroscopic hematuria, dizziness, fatigue, and fever (up to 38°C). There was no evidence of traumatic injury. The patient reported a history of pulmonary thromboembolism treated in a hospital 4 years ago. He has been taking an indirect anticoagulant drug (acenocoumarol) since then.

On physical examination the patient was pale, cold, and sweating, with a pulse frequency of 120/min and a low blood pressure. He had mild tenderness in the right flank. The abdomen was slightly painful on palpation of the right side, but without any palpable masses. The laboratory tests showed a drop in hemoglobin (Hb) levels from 125 g/l and hematocrit (Ht) from 0.39 to Hb 72 g/l and Ht 0.21 since the last three days prior to admission. White blood cells (WBC) were 15.0 G/l and the erythrocyte sedimentation rate (ESR) was 100/120. Coagulation status: INR = 2.54.

The patient was referred to our clinic for percutaneous drainage after outpatient ultrasonography (USG) (Fig. 1), IVP (Fig. 2), and computerd tomography (CT) (Fig. 3) were performed by the radiologist who proposed the initial diagnosis: "Right hydro-pyonephrosis".

While preparing for nephrostomy we repeated the USG examination and concluded that the presented case was not of an obstructed kidney, but more likely of a tumor that had destroyed its structure and caused a retroperitoneal liquid collection.

Needing further evaluation of this case, we revised the printouts of the CT scan performed prior to admission, which showed the wrong initial diagnosis: the rupture of the right kidney on the late scans (24 hour) was evident and our diagnosis of a spontaneous kidney rupture with perinephric hematoma was given (Fig. 3).

We started preparing the patient for surgical treatment due to the high suspicion of a ruptured tumor as the underlying cause. The Hb levels were brought to satisfactory levels and the patient was operated on the 4th day after admission.

The retroperitoneal space on the right side was explored and a ruptured kidney tumor was found. The tumor was 7 x 6 cm in diameter, without macroscopic invasion to the renal capsule, the perinephric fat, or the renal pelvis. There was no visible lymphadenopathy along the inferior vena cava. Right radical nephrectomy was performed. The histological result (Nos. 520-533/26.01.2009) revealed a poorly differentiated sarcomatoid renal cell carcinoma – $T_1N_0M_0G_3$. The initial histologic diagnosis was verified by immunohistochemistry (Cytokeratin and Vimentin positive staining).

Despite the pre- and perioperative prophylaxis with low molecular heparin (enoxaparine 60 mg daily subcutaneously) and elastic socks of the lower extremities, the patient developed pulmonary thromboembolism in the early postoperative period and

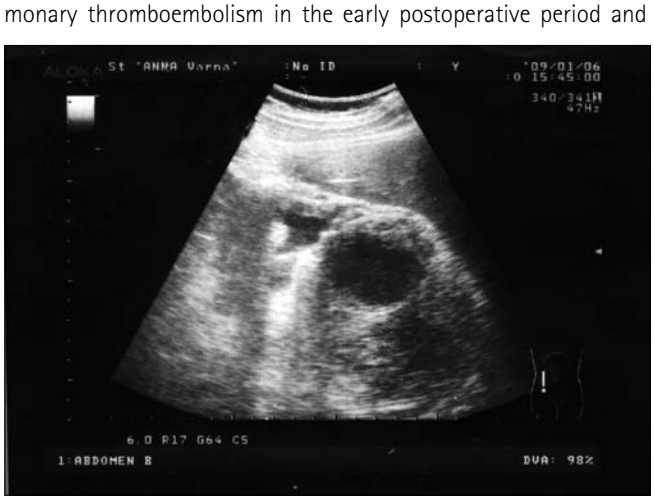

Fig. 1. USG examination demonstrating retroperitoneal liquid collection, initially reported as hydro-pyonephrosis.

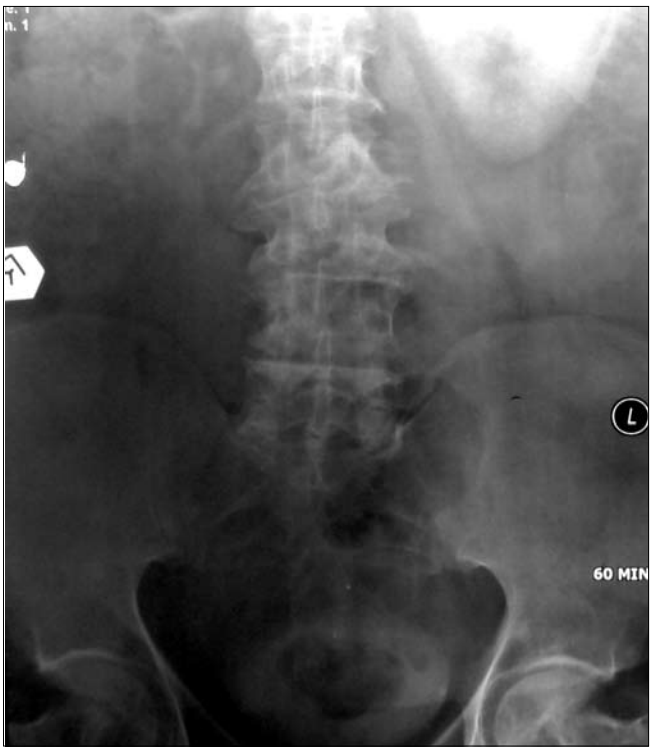

Fig. 2. IVP at 60th minute showing non-functioning kidney to the right. After this result, CT with IVCM was performed.

was transferred to the ICU where he was treated for five days. After successful treatment he was discharged from the hospital on the 16th day after surgery.

The early six months of postoperative follow-up showed no signs of recurrent or metastatic disease during regular check-ups, but, unfortunately, eight months after surgery the patient died due to cerebral infarction, not related to the oncologic disease.

DISCUSSION

The clinical manifestation of spontaneous renal bleeding confined to the subcapsular and perinephric space in patients with no obvious underlying cause was first described by Wunderlich in 1856 [3]. Patients may present with the classic triad of symptoms: acute lumbo-abdominal pain, a palpable tumor mass, and general deterioration of the patient caused by hypovolemic shock [4]. Underlying conditions, described as major causes for Wunderlich syndrome, include: benign and malignant renal tumors, vascular lesions like polyarteritis nodosa, renal infections, nephritis, previously undiagnosed hematological conditions, and anatomical lesions like cysts and hydronephrosis [4]. The most common cause of spontaneous renal hemorrhage in most series is angiomyolipoma, followed by renal cell carcinoma [5].

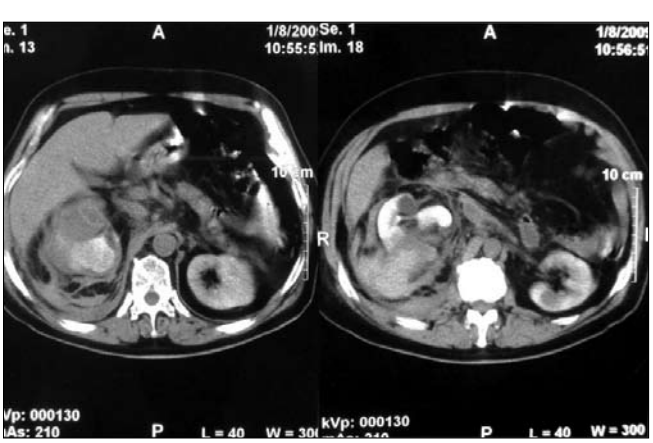

Fig. 3. Late CT scans demonstrating retroperitoneal collection and a contrasting ruptured renal parenchyma.

The appropriate treatment of patients with spontaneous perinephric hemorrhage depends on the prompt diagnosis of a retroperitoneal hemorrhage and on the proper determination of its cause [6]. Radiologic imaging plays an important role in addressing these issues. CT is the method of choice for the demonstration of perirenal hemorrhage, having a sensitivity of 100% [5]. However, a CT performed at the time of hemorrhage is only moderately successful in identifying the renal neoplasm causing the hemorrhage, with a reported sensitivity being 57% [5].

Some studies on spontaneous kidney rupture [7] as well as the current guidelines for the management of traumatic kidney ruptures [8] suggest that conservative treatment is an acceptable option if an accurate diagnosis of the rupture and its extent have been made by the currently available imaging modalities.

On the contrary, some case-based articles [9] show that in spontaneous renal rupture secondary to renal tumors, imaging studies such as CT or magnetic resonance imaging (MRI) often fail to demonstrate the primary lesion and this could be fatal for patients in whom the underlying malignancy progresses. This is the reason why surgical exploration has been accepted in many centers as the primary therapeutic option when the etiology of the renal rupture remains unclear. Surgery also facilitates the final pathological diagnosis. Our conclusion is that if the patient can be medically stabilized during the acute phase of spontaneous perinephric hemorrhage, nephrectomy for RCC or selective arterial embolization for angiomyolipoma can be deferred. In their review on the diagnosis and management of seven cases of Wunderlich syndrome, Cubillana et al. found that conservative management was the most acceptable treatment option, unless a malignant pathology was demonstrated [10].

One particularly interesting point in our case is the anticoagulant treatment as a triggering factor for the development of Wunderlich syndrome. Capitanini et al. [11] also reported a case of a renal hemorrhage in a patient with coagulopathy, but with no malignant cause.

Our case shows that the initial diagnosis is crucial for the appropriate treatment and imaging can be misleading. Our treatment strategy included surgical intervention, but was delayed after clarifying the retroperitoneal hemorrhage and bringing the patient's vital and laboratory parameters to normal.

CONCLUSIONS

Wunderlich syndrome refers to spontaneous non-traumatic renal bleeding into the subcapsular and/or perirenal space. The appropriate treatment of this condition depends on the prompt diagnosis of perinephric hemorrhage and on the accurate determination of its etiology, which requires awareness of the condition. Imaging, therefore, plays an important role in the evaluation of such patients. The above case shows wrong primary imaging diagnosis as a result of the rareness of the condition, which can lead to a wrong therapeutic choice. The trend towards less invasive surgical treatment and more advanced imaging options requires precise diagnosis prior to any surgical intervention.

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