Wunderlich syndrome secondary to cyst rupture and concurrent anticoagulation

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Wunderlich syndrome (WS) is a rare triad of flank pain, flank mass and hypovolemic shock and is classically attributed to angiomyolipomata or neoplasms. Treatment is guided by clinical severity: conservative, selective arterial embolization, or nephrectomy. We report an atypical case of a 69-year old man with a pre-existing 9 cm left renal tumor who developed WS secondary to anticoagulation and simple cyst rupture from his contralateral kidney, complicated by abdominal compartment syndrome with hemodynamic instability despite inotropic support and robust resuscitation. Early recognition and source control via radical nephrectomy were essential in securing a positive outcome..

Key Words: spontaneous renal hemorrhage, simple renal cyst, Wunderlich syndrome

Introduction

Wunderlich syndrome (WS), first described in 1856 as "spontaneous renal capsule apoplexy", is a rare clinical condition characterized by spontaneous atraumatic renal hemorrhage into the subcapsular and perirenal space, and is classically attributed to angiomyolipomata or neoplasms. We report an atypical case of a 69-year old man with a pre-existing 9 cm left renal tumor who developed WS secondary to anticoagulation and simple cyst rupture from his contralateral kidney.

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Case report

A 69-year-old male with no significant history other than well-controlled hypertension initially presented with severe hepatobiliary sepsis from cholangitis secondary to an obstructing common bile duct calculus. He was also found to have portal vein thrombosis and was started on subcutaneous enoxaparin. A computed tomography (CT) scan of his abdomen and pelvis incidentally showed a 4.8 cm right renal upper pole Bosniak 1 cyst and a 9 cm mass in his left renal upper pole consistent with the radiological diagnosis of renal cell carcinoma (RCC) with no evidence of local or distant spread. Our patient was therefore planned for elective left radical nephrectomy following discharge.

Seven days after discharge, he presented with right sided abdominal pain and vomiting. Vital signs were

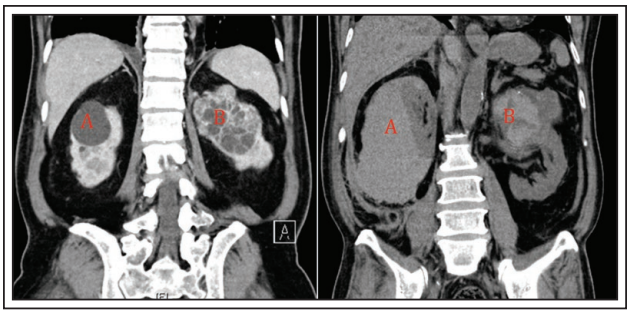

Figure 1. Computed tomography of the abdomen and pelvis (CTAP) showing a simple right upper pole renal cyst **(A)** and left renal tumor **(B)**. Post-WS non-contrasted CTAP shows rupture of the right renal cyst with a large perinephric hematoma but interval stability of the known contralateral renal tumor.

stable at the time of admission. Abdominal examination revealed a tender vaguely palpable mass extending from the right upper quadrant to the right flank. Initial laboratory investigations showed anemia, leukocytosis (hemoglobin 7.9 g/dL, WBC 12.7 x 10^9/L) and raised inflammatory markers (procalcitonin 0.25 ug/L). Liver function was normal. Enoxaparin was withheld pending further investigations. Two hours into his admission, he developed hypovolemic shock with blood pressure 88/50 and sinus tachycardia of 120 beats per minute. Hemoglobin (Hb) when rechecked showed an acute drop to 3.8 g/dL. The patient was resuscitated with intravenous fluids and packed red cell transfusion and transferred to the intensive care unit (ICU) for further management. Urgent non-contrasted CTAP showed a large right perinephric hematoma with extensive perinephric stranding and fascial thickening. The left renal tumor remained stable and did not show evidence of hemorrhage, Figure 1. Though initially planned for angioembolization, our patient continued to deteriorate rapidly, requiring intubation and dual inotropic support with noradrenaline and vasopressin. Thereafter, he developed abdominal compartment syndrome – with a raised intra-abdominal pressure (IAP) of 25 mmH20 – as well as acute renal failure with anuria and severe metabolic acidosis (pH 7.2; BE -11; lactate 10.9), for which he underwent dialysis.

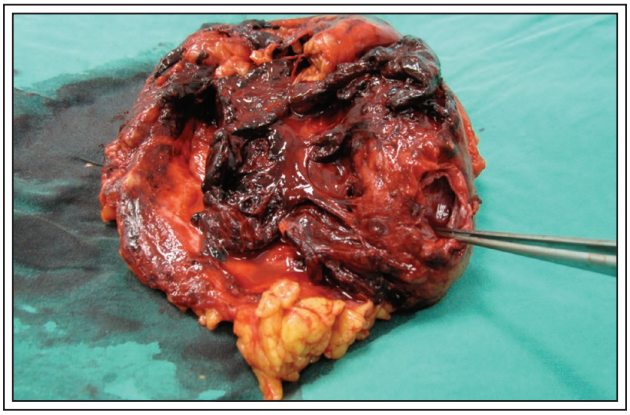

Figure 2. Right kidney with ruptured right upper pole cyst.

The patient was resuscitated and optimized within limits and the decision was made for emergent source control via a right nephrectomy. IAP prior to incision was 47 mmH20. Hemoperitoneum was encountered on entry with the source congruent with that of a ruptured right upper pole cyst. Histopathological examination confirmed rupture of the right upper pole simple cyst with hemorrhage and organized hematoma, Figure 2. Postoperative recovery was turbulent. On day one postoperatively, he developed ventricular fibrillation with a subsequent cardiac arrest but achieved return of spontaneous circulation (ROSC) following 4 cycles of cardiopulmonary resuscitation and intravenous adrenaline. Additionally, he had two episodes of severe cecal bleeding requiring angioembolization. He was finally discharged well 42 days from admission. The outcome of a lengthy discussion between the medical team, the patient, and his family was to forgo the initially planned left radical nephrectomy for his RCC in view of the high morbidity associated with an anephric state and the need for renal replacement therapy long term. Having opted for a management approach centered on quality-of-life rather than cure of his existing renal neoplasm, the patient is now on follow up with urology and palliative care post-discharge.

Discussion

Although WS is classically characterized by Lenk's triad (acute flank pain, flank mass and hypovolemic shock) which was also reflected in our patient, clinical presentation can vary, including nonspecific malaise to gross hematuria.[1,2] Incidence is higher in males, with one meta-analysis reporting a male-to-female ratio of 6:5, wherein the majority of cases occurred in those between 30 and 60 years old.[3]

Conventionally, tumors are the most common cause of WS (56.9%), with ruptured angiomyolipomata accounting for as much as 74.1% of this subgroup.[4] Vascular causes, particularly polyarteritis nodosa, were typically the third most common culprit, although recent literature has demonstrated a shift favoring vasculitis over malignant neoplasms (15.7% versus 14.7%) as the second most common cause.[4] Other causes

include renal artery aneurysm (4.9%), arteriovenous malformation (2.9%), severe pyelonephritis (2.9%), as well as miscellaneous causes including idiopathic (5.9%), pre-eclampsia (1%) and cyst rupture (1%).[4] There were two reported cases of WS caused by simple cyst rupture – one of these cases was associated with concurrent aspirin use, which was then ruled out as a predisposing factor.[5,6] However, a review of the literature demonstrated three more recent cases of WS in anticoagulated patients in the absence of other risk factors, highlighting that the fairly benign concept of routine anticoagulation may indeed require more recognition when considering WS in clinical practice.[7,8] To our knowledge, this is the first case report wherein WS occurred from simple cyst rupture on a background of anticoagulation in the presence of a contralateral large renal tumor.

Imaging can be performed with ultrasound or CT scan. However, CT remains the diagnostic modality of choice especially in an unwell patient as it is 100% sensitive for retroperitoneal hemorrhage.[3] Adequate resuscitation and early embolization where appropriate should be the tenets of treatment. A 2017 systematic review demonstrated that transarterial embolization (TAE) was successful in up to 87% of cases, with the remainder requiring escalation to definitive surgical intervention by way of partial or radical nephrectomy and hematoma evacuation.[4] In our patient, the initial goal of management was to perform TAE to avoid unnecessary organ loss, especially in the context of his planned left nephrectomy for his contralateral renal tumor. Unfortunately, this was limited by his rapid progression of abdominal compartment syndrome contributing to his clinical deterioration and source control with an emergency nephrectomy proved to be a crucial life-saving measure in our patient's management. □

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