Case report

Idiopathic Bilateral Wunderlich Syndrome: a case report

ABSTRACT

Wunderlich Syndrome (WS) is a rare condition characterized by spontaneous bleeding in the kidney without any traumatic event. It usually happens unilaterally, but bilateral cases are rare. The most common causes of WS are renal neoplasms, vascular disorders, infections, renal cystic diseases, and anticoagulation states, with idiopathic cases being uncommon. Here, we present a case of bilateral idiopathic Wunderlich Syndrome.

Keywords: Wunderlich Syndrome, spontaneous bleeding, kidney, imaging studies.

1. INTRODUCTION

Wunderlich Syndrome (WS) is a rare condition where there is spontaneous bleeding in the kidney into the subcapsular, perirenal, and/or pararenal area without traumatic event. People with his condition typically experience sudden flank pain, a mass in the flank, and hypovolemic shock, known as the Lenk triad. The most common causes of WS are angiomyolipomas and renal cell cancer, but it can also be caused by vascular disease, cystic renal diseases, infections, and induced anticoagulation states. Imaging studies, like computer tomography (CT), are used to diagnose this condition. WS usually happens on one side, but there have been cases where both kidneys are affected, like in the case of a young man that we present here.

2. PRESENTATION OF CASE

A 37-year-old man presented to the emergency department with pain in both flanks over the last 4 days. He had a history of asthma since he was 3 years old and was on treatment with montelukast; no traumatic events were reported, and he wasn't taking anticoagulants or platelet antiaggregants. The pain increased, and nausea with vomiting was added. Initial evaluation revealed a temperature of 38.5°C, blood pressure of 142/98 mmHg, pulse of 104 beats per minute, respiratory rate of 12 breaths per minute, oxygen saturation of 93% while breathing ambient air, and bilateral Giordano sign. The results of pulmonary and cardiovascular examinations were normal. Initial laboratory studies were hemoglobin of 17.8 g/dL, white blood cells of 12,400/mcL, platelets of 222,000 cells/mcL, prothrombin time of 15.1 seconds, partial thromboplastin time of 40.0 seconds, serum creatinine of 3.0 mg/dL, and blood urea nitrogen of 35 mg/dL. Urine analysis showed 8-10/c eumorphic blood cells, and 5-6 non-active white blood cells, without proteins. Renal ultrasound (figure 1) shows heterogeneous hypoechoic collections next to both renal capsules. A non-enhanced contrast CT scan (figure 2) was performed due to acute renal disease, bilateral perirenal heterogeneous collections with blood density areas (average 54 HU) were observed. WS diagnosis was established, and conservative treatment was initiated; after five days, renal function was recovered (SCr 0.9 mg/dL). To look for a possible etiology of WS, magnetic resonance imaging with gadolinium enhancement was obtained (figure 3); an echo gradient sequence confirmed hemosiderin deposits in perirenal collections, and neoplasia was ruled out when the renal parenchyma showed homogeneous gadolinium enhancement in fat-sat T1-weighted sequence. No one possible etiology of the WS was identified, then it was

classified as idiopathic. The patient was discharged without symptoms, a 1-month control CT scan showed complete resolution of WS.

3.DISCUSSION

The nontraumatic spontaneous kidney hemorrhage, also known as Wunderlich Syndrome (WS), was first described as “spontaneous renal capsule apoplexy” in 1856 by Carl Reinhold August Wunderlich (1). It is a rare condition characterized by kidney hemorrhage into subcapsular or perirenal spaces without any prior trauma. Most cases reported in the past had unilateral kidney involvement. However, here we present a rarer case of bilateral WS.

The clinical presentation of WS is broad. The most commonly reported symptom is sudden flank pain, a mass in the flank, and hypovolemic shock, collectively known as de Lenk triad. However, this triad is present only in 27% of the patients(2). The most frequent symptom, as in our case, is flank pain or generalized abdominal pain in 67% of the cases, followed by hematuria in 40%, and hypovolemic shock in 27%. Other symptoms that could occur are vomiting, nausea, and anemia. In a few cases, important perirenal hemorrhage has been associated with Page Kidney, which is characterized by systemic arterial hypertension due to activation of the renin-angiotensin-aldosterone system driven by renal extrinsic compression(3).

WS usually affects only one kidney, and bilateral presentation, like in our case, represents about 3% of all reported cases(4). Bilateral WS is more frequently associated with tuberous sclerosis complex and less frequently associated with other neoplasias, vasculitis, pseudoaneurysms, and pregnancy. In approximately 5%–10% of patients with WS, no renal or systemic abnormality is identified at imaging, and these cases are classified as idiopathic WS(5). To the best of our knowledge, this is the first case of idiopathic bilateral WS being reported.

Diagnosis of WS is made through imaging studies. Today, multiple imaging study modalities are available to evaluate spontaneous kidney hemorrhage, such as ultrasonography (US), computer tomography (CT), and magnetic resonance imaging (MRI). These studies establish the diagnosis of WS and identify any possible etiology(6). The first imaging study usually performed in these patients is an ultrasound, which has good sensitivity in identifying perirenal hematomas. However, compared to CT and MRI, ultrasound has limited capacity to identify the underlying cause of WS.

In ultrasound, subcapsular or perinephric hemorrhages are usually seen as iso or hyperechoic collections in the acute stage and hypoechoic in the subacute stage, which may also have septa inside. The main utility of ultrasound in patients with WS lies in interventional radiology for guiding percutaneous drainage of these hematomas and in evaluating their evolution(5). CT is the standard method for diagnosing WS, which, in addition to being useful for identifying hemorrhage, allows for a precise evaluation of its extension and the identification of underlying causes in up to 50% of cases. In unenhanced CT during the acute phase of hemorrhage, it appears as a hyperdense liquid collection (30-79 HU), while in contrast-enhanced CT, the presence of extravasation and pseudoaneurysms suggests active bleeding. MRI is usually performed when, after the tomographic study, a cause has not been found. Intensity abnormalities depend on the state of the blood products. Acute hemorrhage can cause variable changes in the signal intensity of T1-weighted images, usually appearing isointense to hyperintense. In subacute hemorrhage, hemoglobin degradation results in heterogeneous hyperintense signals on T1- and T2-weighted images(5).

Treatment depends on the clinical presentation and the etiology. Cases without hypovolemic shock can be managed conservatively, while those with hypovolemic shock require fluid resuscitation and blood transfusions. In cases of refractory shock or active hemorrhage, management includes open nephrectomy or selective endovascular embolism(7).

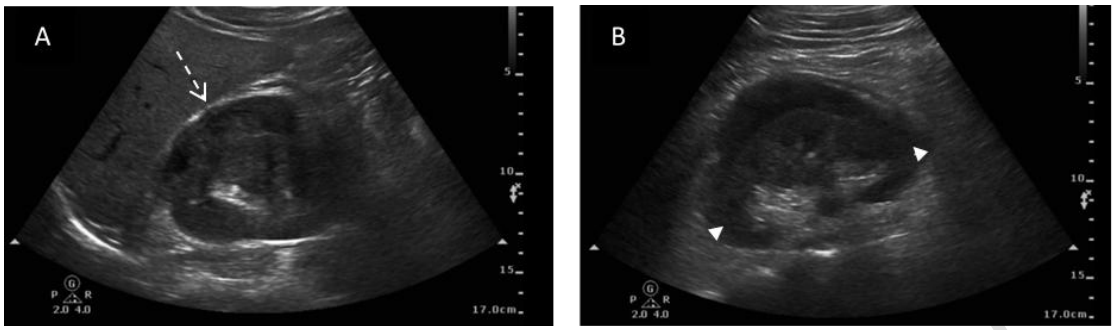

Fig. 1. Renal ultrasound. A) Gray-scale ultrasound image of the right kidney showing a heterogeneous liquid collection with internal echoes (dotted arrow). B) Gray-scale ultrasound image of the left kidney showing that the collection is subcapsular perirenal (arrowheads).

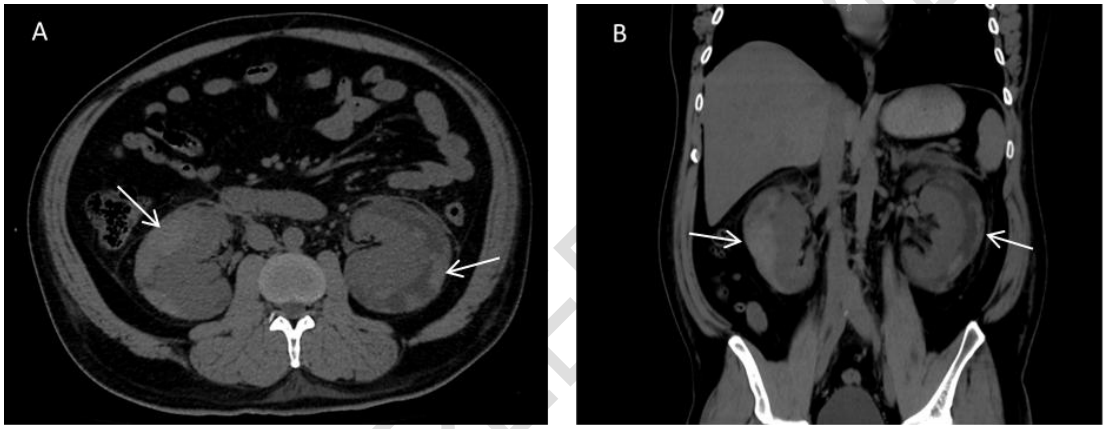

Fig.2. Non enhanced abdominal computed tomography Axial (A) and coronal (B) sections showing both kidneys with subcapsular heterogeneous collections with areas of hemorrhagic density (54 HU) (white arrows).

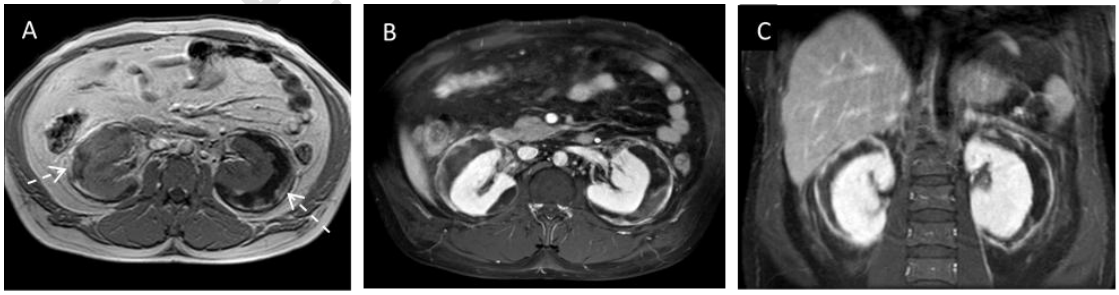

Fig. 3. Abdominal magnetic resonance imaging with contrast. A) Axial gradient echo T1 sequence showing hyperintense areas within the perirenal collection suggesting composition by elements derived from blood degradation (dotted white arrows). B) Axial and C) coronal T1 fat saturation sequences after gadolinium administration demonstrating the absence of occupying lesions in the renal parenchyma.

4. CONCLUSION

Idiopathic bilateral Wunderlich is a rare presentation.

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