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Wunderlich Syndrome in a Patient with Pyelonephritis

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Abstract **Case Report**

Wunderlich syndrome (WS) is a life-threatening emergency characterized by the sudden onset of spontaneous, non-traumatic renal hemorrhage in the sub-capsular and perirenal space, with a distinct clinical and radiological presentation that enables diagnosis. Although most cases require invasive treatments such as surgery or embolization to control the bleeding, our 17-year-old patient admitted to the Mohammed VI hospital in Marrakech was successfully managed with conservative treatment.

Keywords: Wunderlich Syndrome, Spontaneous, Haemorrhage, Pyelonephritis.

INTRODUCTION

Wunderlich syndrome (WS), named after Carl Wunderlich, is a rare condition characterized by an acute, spontaneous renal hemorrhage into the subcapsular, perirenal, and/or pararenal spaces, occurring without any history of trauma. The primary etiology of this hemorrhage is tumors, accounting for approximately 65% of cases, followed by vascular pathologies (20%-30%), infectious causes (12%), and a variety of other etiologies, including cystic renal diseases, nephrosclerosis, and preeclampsia, which collectively constitute about 12.7% of cases [1, 2]. Clinically, WS is classically identified by Lenk's triad, which includes acute flank pain, a palpable flank mass, and hypovolemic shock. However, the presentation can be variable and nonspecific, with patients exhibiting a range of symptoms from general flank or abdominal pain to severe manifestations such as hypovolemic shock. Cross-sectional imaging modalities, such as computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound (including contrast-enhanced ultrasound), are employed to evaluate patients suspected of having WS. These imaging techniques are crucial for detecting perirenal hemorrhage and identifying its underlying cause [3-5]. Interventional radiology plays a vital role in the management of patients with active and significant bleeding.

Herein, we report the case of a 17-year-old male presenting with bilateral pyelonephritis complicated by Wunderlich syndrome.

CASE REPORT

A 17-year-old male with no significant past medical history—including the absence of recent trauma, coagulopathy, or risk factors for renal malignancy or vascular disease—presented to the emergency department (ED) with left flank pain persisting for two weeks, accompanied by irritative lower urinary symptoms. Upon arrival at the ED, he exhibited an altered state of consciousness, tachycardia, tachypnea, and generalized jaundice. Laboratory investigations revealed the following: hemoglobin level of 9 g/dL (reference range: 13-17 g/dL), hematocrit of 24.6% (reference range: 42%-54%), leukocyte count of 38.4 K/µL (reference range: 4.0-10.0 K/µL), blood urea nitrogen of 3.15 g/L (reference range: 0.25-0.48 g/L), and creatinine of 102.6 mg/L (reference range: 7-12 mg/L). Abdominal ultrasonography suggested bilateral pyelonephritis with a left renal abscess (Figure 1). Contrast-enhanced abdominal computed tomography (CT) confirmed bilateral pyelonephritis and identified a subcapsular hematoma in the left kidney, measuring 14 × 6.3 × 5.9 cm with a volume of 260 cc (Figure 2), and showed no active contrast extravasation.

Subsequently, renal angiography was proposed to investigate a potential vascular etiology of the hemorrhage and to perform embolization if necessary. The left selective renal angiogram demonstrated an aberrant renal artery supplying the inferior pole of the kidney, without evidence of vascular malformation or disease (Figure 3). The right selective renal angiogram was unremarkable (Figure 4). Urine and blood cultures

grew Escherichia coli producing beta-lactamase, sensitive to quinolones. Following an emergency hemodialysis session, conservative management was adopted, and antibiotic therapy was initiated with ceftriaxone at a dosage of 1000 mg every 12 hours, alongside fluid replacement and blood product transfusion. The patient experienced a favorable postoperative course and continued treatment with levofloxacin 500 mg every 24 hours for two weeks based on urine culture results. He was discharged after 17 days with a plan for outpatient follow-up.

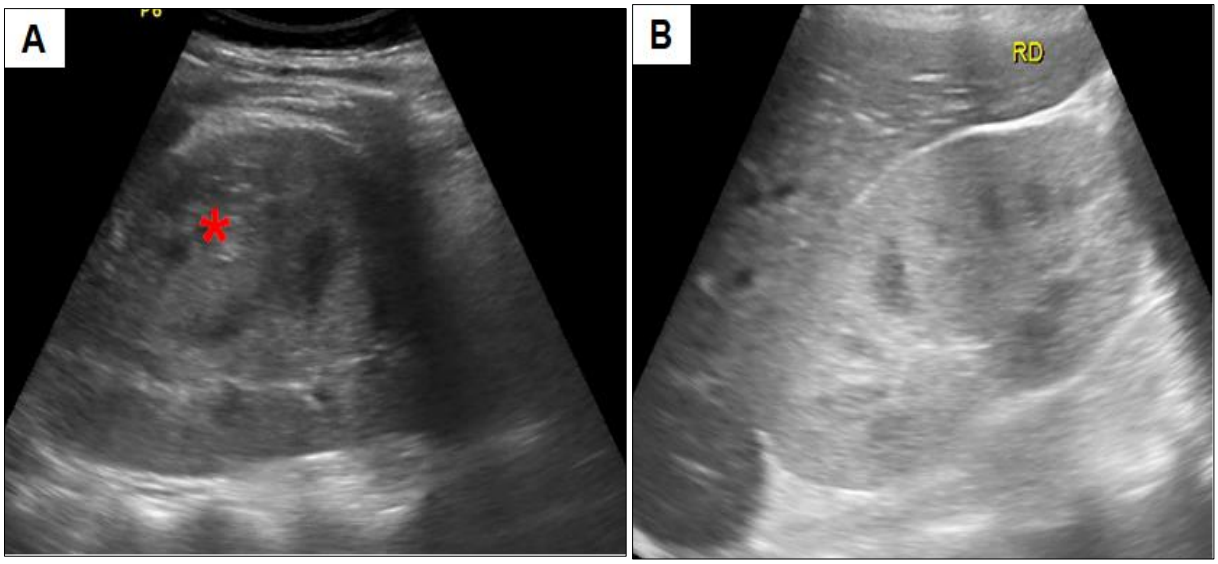

Figure 1: Ultrasound images of the left kidney (A) showing a heterogeneous subcapsular collection (*) initially thought to be a perirenal abscess and a right kidney image (B) showing enlarged kidney with echogenic cortex suggestive of pyelonephritis.

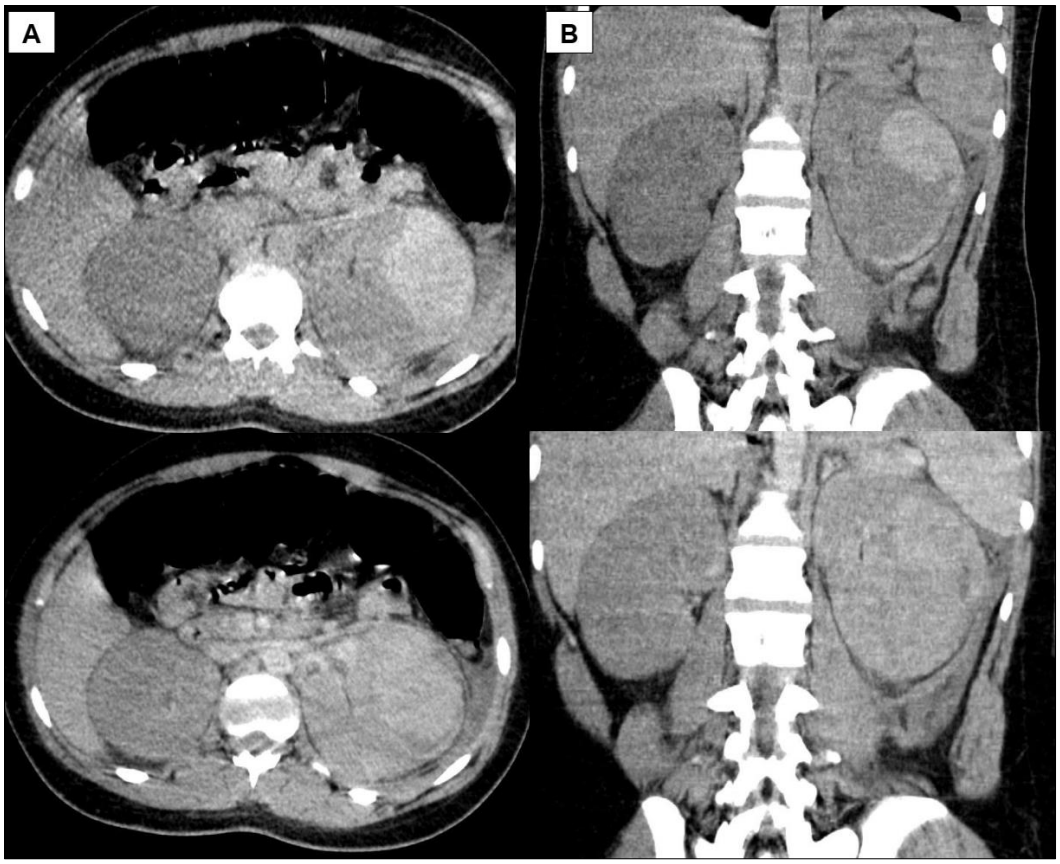

Figure 2: Abdominal-pelvic (A) transverse and (B) coronal computed tomography without and with contrast shows a sub capsular hematoma in the left kidney measuring 14 x 6.3 x 5.9 cm with a volume of 260 cc and bilateral nephromegaly with pale nephrography and infiltration of peri-renal fat in favour of bilateral pyelonephritis

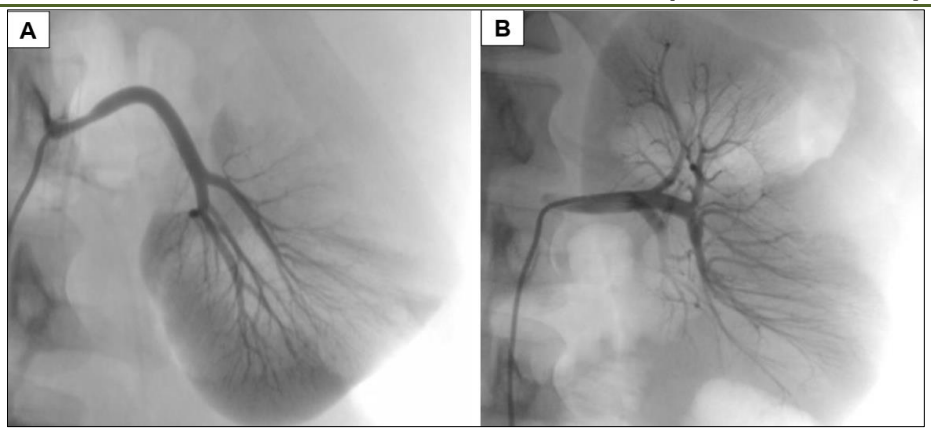

Figure 3: Selective left renal angiogram shows an aberrant renal artery supplying the inferior pole of the kidney (A)

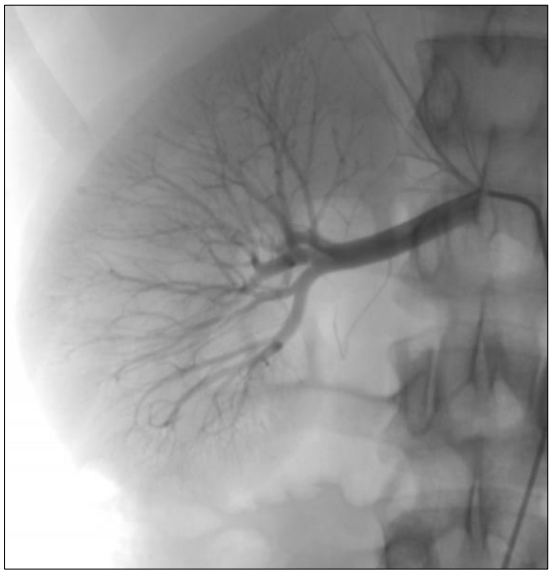

Figure 4: A normal selective right renal angiogram shows unique renal artery

Table 1: Etiologic factors of WS.

Renal neoplasms	angiomyolipoma
	RCC
	Metastases
	Renal sarcomas
	Urothelial carcinoma
	oncocytoma
Renal vascular diseases	Polyarteritis nodosa
	RAAs and pseudoanevrysms
	Renal aerteriovenous malformations
	Renal vein thrombosis
Renal infections	Acute pyelonephritis
	Renal abscesses
	Emphysematous pyelonephritis
Kidney failure	
Hematologic abnormalities	hemophilia
	Anticoagulant drug induced
Renal cystic disorders	
Renal calculus	
Other	Recreational drug use
	Idiopathic WS

DISCUSSION

A variety of causes can lead to spontaneous, non-traumatic, acute renal hemorrhage that extends to the subcapsular, perirenal, and/or pararenal spaces, a condition known as Wunderlich syndrome (WS) (Table). Neoplastic and vascular causes are the main etiology of WS [6, 7]. Angiomyolipomas and clear cell renal cell carcinomas (RCCs) are the most common benign and malignant neoplasms, re-spectively, of the kidneys, with a marked propensity to cause WS (5). Rupture of renal artery aneurysms (RAAs) or pseudo-aneurysms, vascular malformations, and vasculitis syndromes, including polyarteritis nodosa, are among the next most com-mon causes of WS [6-3]. Hereditary and acquired renal cys-tic diseases, kidney failure, pregnancy, and iatrogenic causes (systemic anticoagulation) predispose patients to WS. Renal infections, calculus disease, and coagulation disorders are rare causes of WS [6-3].

Renal infections are an uncommon cause of WS, including acute pyelonephritis, emphysem-atous pyelonephritis, and renal abscesses, renal infections are responsable of 5%–10% of all cases of WS [3-8]. Infection associated with renal parenchymal necrosis and vascular thrombosis of the renal vessels predispose patients to renal parenchymal hemorrhage and subsequent rupture, with the development of WS [4]. Treatment options include conservative management with antibiotics, percutaneous drainage of abscesses or hematomas, and transcatheter arterial embolization (TAE) or surgery in cases of ongoing bleeding

The primary objectives of imaging in patients with Wunderlich syndrome (WS) are to confirm the diagnosis, evaluate the extent and anatomical location of the bleeding (such as subcapsular, perirenal, or pararenal), identify the underlying cause, and provide context for treatment options [3]. Ultrasound is highly sensitive for identification of a perirenal hematoma and is useful as a follow-up imaging to assess the evolution of the hematoma. However CT is the modality of choice for the diagnosis of WS. It allows accurate estimation of the hematoma's volume and its extent, and assists in the the identification of many of the underlying causes of WS [4]. Acute hemorrhage appears as a hyperattenuating fluid collection (30–70 HU) on non-contrast CT images. It is important to mention that large hematomas may obscure small renal cysts or masses. The presence of active contrast material extravasation and pseudoaneurysms on post-contrast CT images suggests ongoing bleeding, necessitating urgent consultation with an interventional radiologist to perform an embolization if necessary [9]. Catheter angiography is useful for detecting the source of bleeding and identifying the vascular causes of Wunderlich syndrome (WS) [8]. Selective renovascular catheterization and embolization have increasingly become the first-line treatment for quickly controlling life-threatening WS and avoiding radical surgery.

Treatment depends on the clinical situation of the patient. In hemodynamically stable patients, treatment is conservative, with abdominal CT scan used for diagnosis and follow-up. However, in unstable patients, a renal angiography is proposed to diagnose and treat a vascular condition, urgent surgical intervention by nephrectomy may be indicated, which is not a first-line treatment because the surgery is associated with high morbidity and mortality [10, 11].

CONCLUSION

Wunderlich syndrome (WS) is an uncommon but potentially life-threatening condition. While a neoplastic origin is the most frequent cause, a wide range of common and uncommon etiologies can predispose patients to WS. Computed tomography (CT) is the preferred method for both diagnosis and follow-up, as it aids in determining the underlying etiology and guides interventional radiology or emergency surgical procedures.

Pyelonephritis, though a rare cause of Wunderlich syndrome, should be considered in patients with renal infection who present with acute severe flank pain, a drop in hemoglobin levels, and hemodynamic instability. Early treatment is crucial for favorable outcomes and should initially be conservative, with continuous assessment of the patient's clinical and hemodynamic status.

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