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(CASE REPORT)

Wunderlich syndrome: Diagnosis and management of a clinical case

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Abstract

Wunderlich syndrome is a rare condition characterized by acute and spontaneous renal hemorrhage in the subcapsular, perirenal, or pararenal spaces, without prior trauma. Symptoms can vary from flank pain to hypovolemic shock. The classic Lenk triad (pain, flank mass, and hypovolemic shock) is observed in approximately 20% of patients. Renal neoplasms, such as angiomyolipomas and renal carcinomas, account for 60 to 65% of cases. Renal vascular diseases (aneurysms, arteriovenous malformations, and vasculitis) represent 20 to 30% of cases. Rare causes include renal infections, cystic diseases, renal stones, renal failure, and coagulation disorders, though Wunderlich syndrome can also be idiopathic. CT (computed tomography) scans and MRIs are essential for detecting and characterizing underlying causes, with follow-up studies sometimes necessary when no clear cause is found. Renal artery embolization may be used as a minimally invasive treatment for acute hemorrhage. Timely diagnosis is crucial to prevent hemodynamic instability, particularly in patients with rare causes such as infections. This article presents the case of a 17-year-old male initially diagnosed with a renal abscess, which progressed into Wunderlich syndrome due to bilateral pyelonephritis.

Keywords: Wunderlich syndrom; hemorrhage; hemodialysis; pyelonephritis; renal failure

1. Introduction

The Wunderlich syndrome is a rare condition characterized by acute and spontaneous renal hemorrhage in the subcapsular, perirenal, or pararenal spaces, without prior trauma. The symptoms can vary, ranging from flank pain to hypovolemic shock (1). The classic triad of Lenk (pain, flank mass, and hypovolemic shock) is observed in approximately 20% of patients. Renal neoplasms, such as angiomyolipomas and renal carcinomas, account for 60 to 65% of cases. Renal vascular diseases (aneurysms, arteriovenous malformations, and vasculitis) represent 20 to 30% of cases. Rare causes include renal infections, cystic diseases, calculi, renal failure, and coagulation disorders. Wunderlich syndrome can rarely be idiopathic. CT scans and MRIs are crucial for detecting and characterizing underlying causes, with follow-up studies sometimes necessary if no cause is identified (2). Renal artery embolization can be used as a minimally invasive treatment in cases of acute hemorrhage. An accurate diagnosis is essential for optimal treatment, whether in emergency or non-urgent care. Furthermore, the importance of identifying Wunderlich syndrome lies in its potential to cause hemodynamic instability, particularly in patients with rare etiologies such as infectious diseases. We present the case of a 17-year-old male with an initial diagnosis of renal abscess that developed into Wunderlich syndrome due to bilateral pyelonephritis.

2. Case report

A 17-year-old patient, from a first-degree consanguineous marriage, with no significant pathological history, presented to the emergency department with diffuse abdominal pain, associated with diarrhea, vomiting, a dry cough, and

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macroglossia. The symptoms had been evolving for 25 days in the context of a general deterioration and fever. On clinical examination, the patient was conscious, with intellectual and mental delay, stable and eupneic, with pale conjunctivae, blood pressure of 140/70 mmHg, no edema in the lower limbs, preserved diuresis, and a negative urine strip.
The urological and nephrological examination was normal, with no pain on lumbar percussion, and a negative Giordano sign.

2.1. Complementary Examinations

The biological tests showed: hypochromic microcytic anemia at 8.5 g/dl, leukocytosis at 32,810/mm^3, normal hemostasis, CRP at 170 mg/l, renal failure with creatinine at 81.41 mg/l and urea at 3.15 g/l. A metabolic acidosis at 9.77 mEq/l, hypoalbuminemia at 31 g/l, and a hepatic panel with ASAT at 181 IU/l and ALAT at 79 IU/l were noted. The macrophage activation syndrome panel showed fibrinogen at 6.68 g/l, lactate dehydrogenase at 468 IU/l, and ferritin greater than 2000. The blood smear showed a reactive appearance. Plasma protein electrophoresis indicated an alpha-1 globulinemia associated with a significant increase in polyclonal gammaglobulins. The profile was suggestive of moderate inflammatory syndrome with an immune reaction. The 24-hour proteinuria was 1.79 g/24h. The urine cytobacteriological examination was sterile. Given the pleuropulmonary symptoms, the patient underwent pleural puncture, which yielded an exudative hemorrhagic fluid, with no germs found in culture. A chest X-ray showed an opacity in the middle and lower left lobe. The abdominal X-ray (without preparation) did not show any hydro-aeric level. A thoracic ultrasound showed a moderate left pleural effusion with some fine echoes and thin and thick septations, measuring 5.3 cm in maximal thickness, with effusion in the parietal pleura. An abdominal ultrasound showed a left kidney measuring 14.6 x 8.8 cm with irregular contours, echogenic cortex, and a hypoechoic cortical-medullary collection in the upper polar region, with fine non-vascularized echoes on Doppler, measuring 6.4 x 6.6 cm, and a focal hypoechoic region of triangular shape in the upper polar region. The right kidney was of normal size, with regular contours and an echogenic cortex. A small, anechoic peritoneal effusion was visible in the pelvic region.
A renal CT scan showed a left kidney measuring 14.6 x 9.5 cm with irregular contours, indicating pale nephropathy. A subcapsular collection causing scolloping on the renal parenchyma in the mid-renal region, extending over its entire length, showed spontaneously hyperdense zones that did not enhance with contrast on Doppler, measuring 63 mm in thickness and extending 14 cm. There was also infiltration of the perirenal fat with thickening of the left perirenal fascia. The right kidney was of normal size with regular contours and pale nephrography.

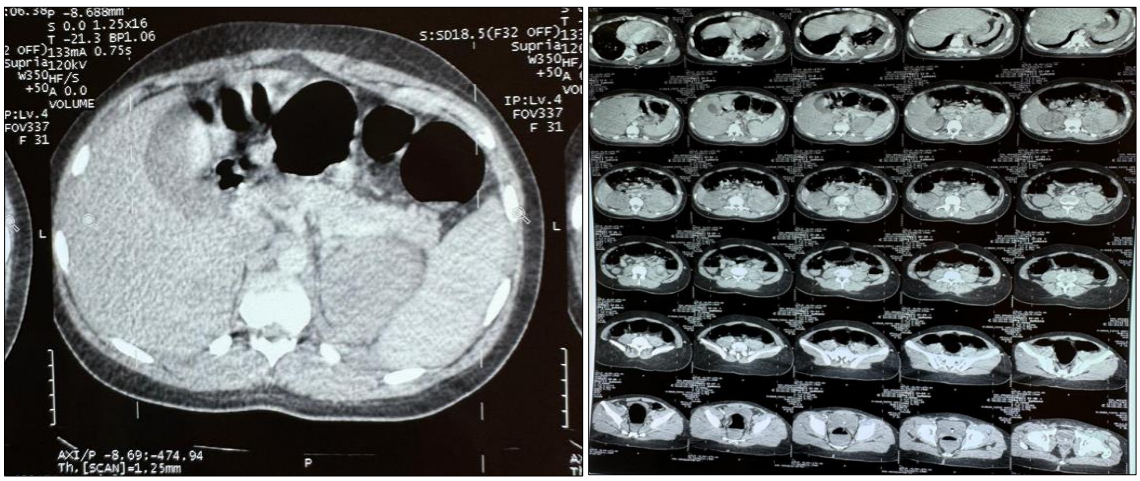

Figure 1 CT scan image of our case

Both kidneys excreted within normal physiological time. Several left latero-aortic lymph nodes of oval shape and sub-centimetric size were present. A small peritoneal effusion was visible in the perisplenic region, and a moderate left pleural effusion with passive atelectasis was noted. The scan findings were suggestive of bilateral pyelonephritis associated with a left subcapsular hematoma undergoing liquefaction, consistent with Wunderlich syndrome. A renal artery angiography showed no abnormalities. The management of our patient included initial stabilization and initiation of hemodialysis due to the clinical-biological uremic syndrome, with 4 hemodialysis sessions. A triple-antibiotic regimen of C3G, gentamicin, and Flagyl was administered for 10 days, along with embolization. The patient showed positive progress with a complete recovery of renal function. However, persistent hypokalemia required potassium supplementation. After stabilization of his condition, he was discharged from the hospital with recommendations for outpatient follow-up to monitor his condition and renal function

3. Discussion

Spontaneous renal hemorrhage was first described by Bonet in 1700, and Wünderlich defined it in 1856 as "spontaneous apoplexy of the renal capsule (1) ." In 1910, Coenen gave this condition the name "Wünderlich syndrome" (1, 5, 6). Wünderlich syndrome can have various causes:

3.1. Tumor-related origin

Renal lesions causing hemorrhages mainly include renal tumors, with angiomyolipomas being the most common (57 to 73% of cases). Angiomyolipoma is the most frequent benign solid renal tumor (3%), composed of smooth muscle fibers, blood vessels, and adipose tissue (8). Although benign in most cases, it can occasionally become malignant. Angiomyolipomas can be asymptomatic, especially the sporadic forms, which primarily occur in middle-aged women. Pregnancy can promote the growth and bleeding of angiomyolipomas, posing risks to both the mother and fetus (12). Several types of angiomyolipomas are noted: fatty predominant, vascular, exclusively muscular, and epithelioid. Histologically, angiomyolipoma is often associated with epithelioid perivascular cells. A diagnosis of angiomyolipoma can be suggested by the presence of fatty content in the renal lesion, evidenced by attenuation values below -20 HU on CT or a high signal on T1 MRI with a signal drop. Other tumors may also contain adipose tissue, including clear cell carcinoma, liposarcoma, atypical nephroblastoma, and teratoma (11). Treatment varies depending on the circumstances: it may include embolization, tumor resection, radiofrequency ablation, cryoablation, or nephrectomy. Follow-up imaging is necessary every 6 to 12 months to monitor for potential complications, such as sarcomatous degeneration (9, 10).

3.2. Vascular origin

Spontaneous renal rupture is more rarely observed in cases of macroscopic vascular lesions (ruptured atheromatous or dysplastic aneurysms, collagenopathies such as Ehlers-Danlos type IV, Marfan syndrome, neurofibromatosis, spontaneous dissection of the renal artery or its branches, renal arteriovenous fistulas, infectious false aneurysms, and also abdominal trauma or percutaneous urological procedures) (7). Another etiological group is represented by systemic vasculitis, particularly polyarteritis nodosa (PAN), microscopic polyangiitis, and sporadically, granulomatosis with polyangiitis (formerly Wegener's disease), vascularitis in IV drug abusers (amphetamines), lead poisoning, and a recent case of W. syndrome during cryoglobulinemia associated with Sjögren's syndrome (necrotizing vasculitis of medium and small caliber arteries) (6).

3.3. Other etiologies

Other cases of spontaneous renal rupture with Wünderlich syndrome have been reported in various situations, notably iatrogenic origin during anticoagulant or antiplatelet therapy, as part of anti-angiogenic targeted treatments in oncology (such as bevacizumab) (5). Infectious etiology remains very rare, mainly in pyelonephritis or infectious complications related to upper urinary tract stones. In about 5% of cases, no identifiable cause is found, which corresponds to idiopathic spontaneous renal rupture, particularly in hemodialysis patients. CT is the reference imaging exam as it allows for assessment of the extent of renal lesions and adjacent structures, and in most cases, it establishes a presumptive etiological diagnosis (2). It is also the primary tool for follow-up in conservatively treated patients. Renal angiography is indicated when a vascular cause is suspected, and embolization can be a therapeutic option in cases of vascular lesions or in patients with benign lesions and contraindications for surgery. Treatment varies depending on the clinical condition of the patient. In hemodynamically stable patients, a conservative approach is favored, using abdominal CT for diagnosis and follow-up (2). However, in unstable patients, urgent surgical intervention, such as nephrectomy, may be required. However, this is not considered the first-line treatment, as surgery carries a high risk of morbidity and mortality (4).

4. Conclusion

Wunderlich syndrome is a rare but potentially serious condition, characterized by spontaneous renal hemorrhages, which can be linked to various etiologies, including renal neoplasms, vascular anomalies, or iatrogenic causes. Its diagnosis relies on clinical evaluation and imaging tests such as CT and MRI, which help to better understand the underlying cause and the extent of the lesions. Treatment varies depending on the patient's hemodynamic stability, ranging from conservative management to more invasive interventions, such as embolization or surgery. Long-term follow-up is essential to prevent complications, especially in cases of benign lesions, such as angiomyolipomas. Our case illustrates the importance of multidisciplinary management, particularly when an initial misdiagnosis may lead to changes in treatment. The favorable outcome of our patient underscores the importance of early and appropriate management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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