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CASE REPORT

Wünderlich syndrome: report of two cases with different presentation modalities

Síndrome de Wünderlich: reporte de dos casos con modalidades diferentes de presentación

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ABSTRACT

Introduction: Wünderlich syndrome is characterized by retropritoneal hemorrhage localized at the renal or perirenal level, which occurs sudden onset. The available evidence on this pathology is scarce because it is a little suspected medical emergency.
Case reports: case 1 is a male patient who presented with left lumbar pain and wanders to the emergency department. Case 2 is a female patient is brought to the emergency department in hypovolemic shock. In both cases, it was decided to perform urgent surgical treatment to correct the hemorrhage due to the progressive deterioration of hemodynamics, the subsequent evolution was favorable.
Conclusion: Wünderlich syndrome presents relatively frequently in emergency departments and represents a diagnostic-therapeutic challenge.

Keywords: Wünderlich Syndrome; Hypovolemic Shock; Retropritoneal Hemorrhage.

RESUMEN

Reporte de caso: el caso 1, es un paciente masculino que presenta dolor lumbar izquierdo y llega deambulando al servicio de urgencias. El caso 2 es una mujer que es trasladada al servicio de urgencias en estado de shock hipovolémico. En ambos se decidió realizar el tratamiento quirúrgico urgente para corregir la hemorragia debido al deterioro progresivo de la hemodinámica, la evolución ulterior fue favorables.
Introducción: el Síndrome de Wünderlich se caracteriza por una hemorragia retropritoneal localizada a nivel renal o perirrenal, que se presenta de forma súbita. La evidencia disponible sobre esta patología es escasa debido a que resulta una urgencia médica poco sospechada.
Conclusión: el Síndrome de Wünderlich se presenta con relativa frecuencia en los servicios de urgencias y representa un reto diagnóstico-terapéutico.

Palabras clave: Síndrome de Wünderlich; Shock Hipovolémico; Hemorragia Retropritoneal.

INTRODUCTION

"Spontaneous renal apoplexy as a result of subcapsular or perirenal hemorrhage," first observed by Bonet in 1700 and described by Carl Wünderlich in 1856, is called Wünderlich syndrome and is characterized by retroperitoneal hemorrhage localized at the renal or perirenal level, which occurs suddenly, without a history of trauma and with multiple etiologies. It is also known as spontaneous renal hemorrhage and is considered a rarely suspected medical emergency.[1] The etiology of this syndrome has been considered to include renal tumors such as angiomyolipomas (40-60 %) and renal cell carcinomas (30-35 %), renal vascular disorders (17 %), infections (5-10 %), coagulation disorders or use of anticoagulants (15 %), and idiopathic causes (5-10 %).[1,2,3] Diagnosis is made based on the clinical picture, which is characterized by Lenck's triad (present in 20 % of patients), consisting of flank pain (67 %), hematuria (40 %), palpable renal mass (27 %), complemented by hypovolemic shock (27 %) and confirmed by imaging studies such as ultrasound, tomography, and nuclear magnetic resonance.[2,3,4] The course of action to be taken in these patients varies, depending mainly on the clinical form in which it presents and its etiology. It may be conservative or surgical emergency treatment after hemodynamic stabilization.[2,5]

Although this disease is considered rare, it is recognized that it is underdiagnosed due to the dissimilar forms of presentation in emergency departments.[1,5] This publication aims to describe two clinical cases of Wünderlich syndrome with different clinical manifestations and outcomes.

CASE REPORT

Case 1

A 79-year-old male patient with a history of type 2 diabetes mellitus presented to the emergency department with sudden onset of severe pain in the left lumbar region, with no history of trauma, which did not radiate and was relieved only slightly by analgesics, forcing the patient to adopt an antalgic posture and making it difficult to walk due to fatigue and muscle weakness.

On physical examination, positive findings included skin and mucous membrane pallor, diaphoresis, sustained tachycardia (102 beats/minute), capillary refill time of 3 seconds, hypotension (90/60 mmHg), exploration of the left lumbar fossa was painful, the fist percussion maneuver was positive, and the pyelonephritis points were tender, with a palpable, movable, and pinchable kidney, with a considerable increase in volume. Hematological studies showed anemia of 9 g/L. Leukocytosis of 11x10(3) /UL with a predominance of neutrophils. An ultrasound scan shows an enlarged left kidney with an irregular edge and the presence of a large heterogeneous fluid collection. A contrast-enhanced computed tomography (CT) scan was performed, revealing a large, hypodense, heterogeneous collection around the left kidney, measuring 85x60x126 mm, with infiltration of the perirenal fat (figure 1).

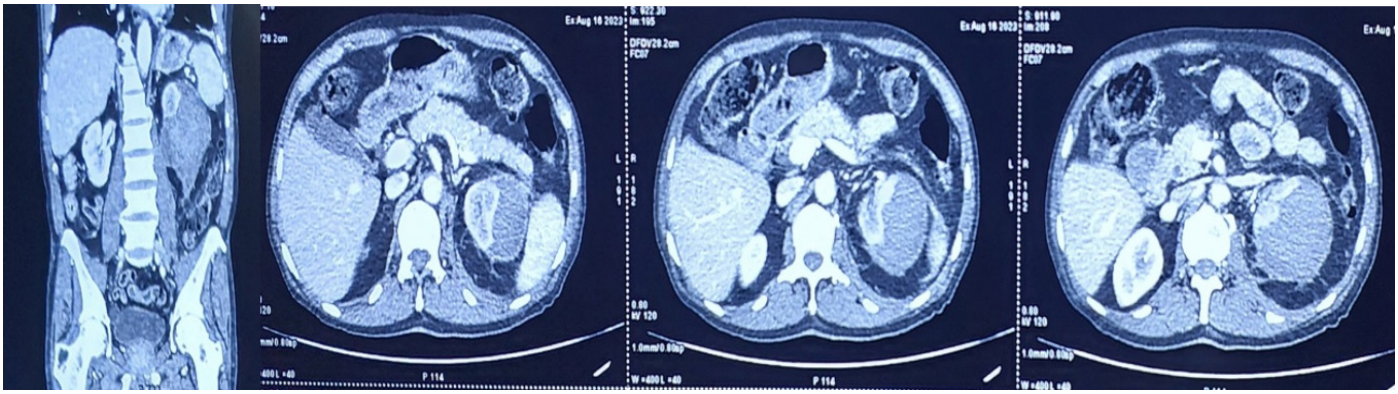

Figure 1. Contrast-enhanced CT scan of the abdomen, coronal reconstruction and axial section, showing a hypodense image bordering the left kidney, which compresses and deforms the kidney due to mass effect. Peri-renal fat alteration is also observed

The diagnosis was left renal and perirenal collection with large hematoma.

With a diagnostic hypothesis of Wünderlich syndrome, the patient was taken to the operating room, where a left lumbotomy was performed and the kidney was observed to be enlarged, with neovascularization, surrounded by a hematoma, adhered to the psoas muscle, lumbar quadratus, and peritoneum. It was freed with difficulty, and during manipulation of the pedicle, the pelvis opened, releasing a large amount of blood, and the renal

cavities were filled with clots (figure 2).

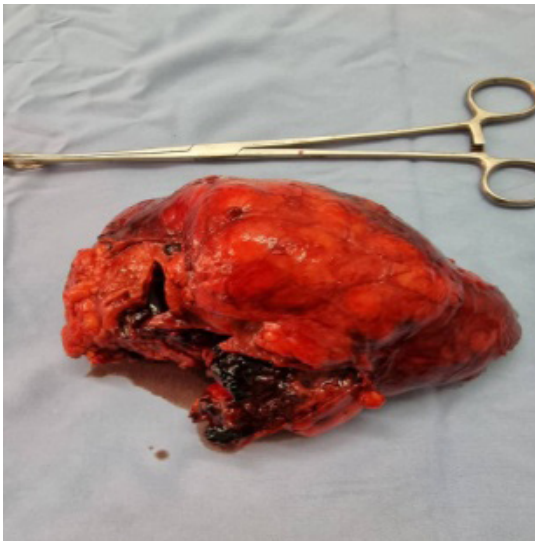

Figure 2. Surgical specimen of the left kidney

A nephrectomy was performed with satisfactory results. The pathological findings revealed renal cell carcinoma.

Case 2

A 25-year-old female patient was transferred to the hospital by the International Medical Emergency System (SIUM) with a condition characterized by loss of consciousness. The family reported no trauma or amenorrhea. Physical examination revealed tachycardia (120 beats/minute), hypotension (80/40 mmHg), signs of tissue hypoperfusion (capillary refill time of 4 seconds), cold skin, a distended abdomen, generalized peritoneal irritation with abdominal tenderness, and absent bowel sounds. Tests show severe anemia, hemoglobin (Hb) 5 g/l. A FAST ultrasound shows abundant free fluid in the abdominal cavity. A puncture of the fundus of the Douglas pouch is performed and blood that does not clot is obtained.

She began to stabilize with volume replacement, transfusions, and the use of vasoactive amines (noradrenaline at a rate of 1,5 mcg/kg/minute). Once this objective was achieved, she was rushed to the operating room with a suspected diagnosis of hypovolemic shock secondary to a possible ruptured ectopic pregnancy. A laparotomy is performed and a double ectopic kidney is observed in the left inguinal fossa, located next to the fallopian tube and immediately above the body of the uterus, surrounded by a large hematoma that drains blood into the peritoneal cavity. A nephrectomy is performed (figure 3). Pathological anatomy revealed a double kidney with hypoplasia of the upper kidney.

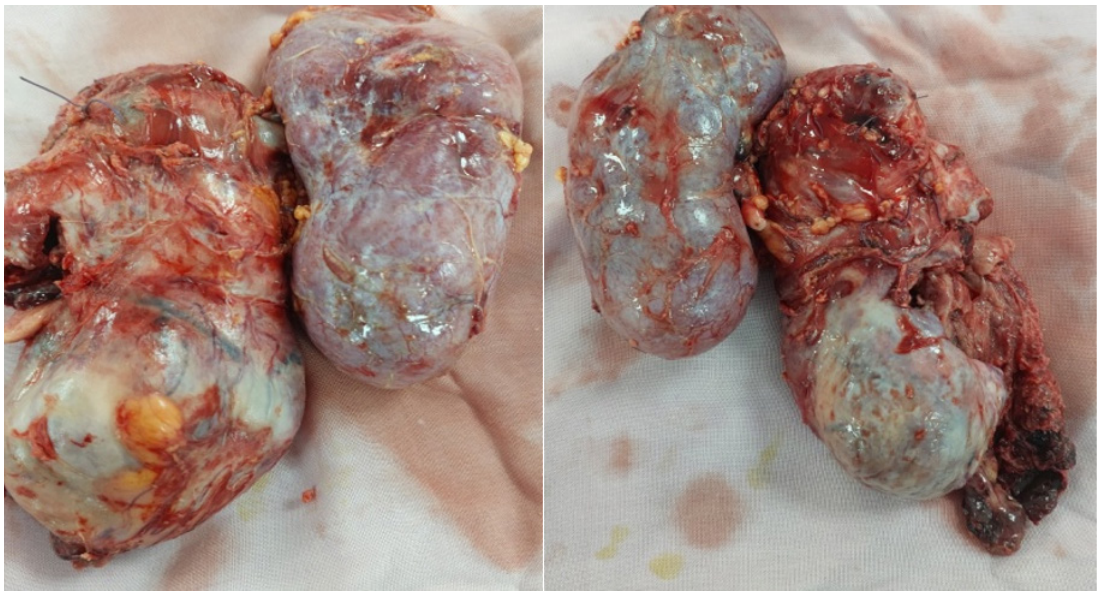

Figure 3. Surgical specimen of the double ectopic kidney

DISCUSSION

The cases described show two forms of presentation of Wünderlich syndrome. The classic clinical picture describes Lenck's triad, which occurs in 20 % of cases, although the manifestations are usually more florid. In the first case, the patient's hemodynamic compromise allowed for a more detailed study, despite evidence of progressive deterioration, while in the second case, hypovolemic shock with the possibility of hemodynamic stabilization prevailed, requiring additional studies essential for diagnosis with transfer to the operating room and surgical correction of the bleeding, which would otherwise have resulted in the patient's death. Both cases were considered medical emergencies.[1, 2, 6]

Emergency medical action is essential due to the course of the disease, where a significant number of cases show hemodynamic deterioration, an aspect that hinders adequate documentation for publication in the scientific literature, resulting in a lack of large case series on the subject.[6]

Etiological diagnosis is difficult in the acute phase, as it requires anatomopathological studies of the kidney. Renal tumors are the most common cause; case 1 was associated with clear cell carcinoma, which accounts for 30-35 % of cases. Case 2 was associated with a congenital malformation characterized by a double kidney with hypoplasia of the upper kidney.[1,6]

The course of action to be taken depends on the patient's condition. Conservative and surgical treatments have been described in the literature, all with satisfactory results. In the two cases presented, invasive treatment was chosen due to the progressive deterioration of the patients' condition.[1,2,3,4,5,6,7,8]

CONCLUSIONS

Although rare in general clinical practice, Wünderlich syndrome represents a real diagnostic and therapeutic challenge in the context of emergency medicine. Its clinical presentation, which is often nonspecific and sudden, together with a diverse etiology ranging from benign processes to malignant entities, requires high clinical suspicion and the timely use of diagnostic tools. Early recognition of this entity not only allows for the initiation of appropriate treatment, but can also be decisive for patient survival, underscoring the importance of its inclusion in the differential diagnosis of acute abdominal or lumbar pain accompanied by hemodynamic instability.

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CONSENT

The patient's consent was obtained for this study.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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