International Journal of Medical Science and Clinical Research Studies

ISSN(print): 2767-8326, ISSN(online): 2767-8342

Volume 03 Issue 11 November 2023

Page No: 2766-2769

DOI: https://doi.org/10.47191/ijmscrs/v3-i11-43, Impact Factor: 6.597

Wünderlich Syndrome Associated with Obstructive Uropathy due to Coralliform Renal Lithiasis: Case Report and Literature Review

Andrea Carolina Torres Ruiz[1], Elizabeth Torres Ruiz[2], Rodolfo Martín Ruiz Ravelo[2], Martín Aquino Ordoñez[1], José de Jesús Fernández Rivera[2], Perla Karina Hernández de Lira[2]

[1] Mexican Social Security Institute – General Hospital of Zone No. 50. Department of Surgery. San Luis Potosí, San Luis Potosí, México

[2] Mexican Social Security Institute – General Hospital of Zone No. 33. Department of Internal Medicine. Monterrey, Nuevo León, México

ABSTRACT

Wünderlich syndrome or non-traumatic spontaneous renal hemorrhage is a rare entity, with less than 500 cases reported to date. It has multiple associated risk factors, the tumoral etiology is the principal one and the angiomyolipoma is the most frequently associated benign tumor. Other causes that are less frequent are systemic diseases such as polyarteritis nodosa, ruptured renal cysts, coralliform renal lithiasis, among others. It´s most common clinical presentation only occurs in 20% of cases and consists of Lenk's triad. Even in cases where the triad is present, the initial diagnosis is by computed tomography (CT) where the retroperitoneal hematoma is observed and sometimes evidencing the underlying etiology as in cases of renal lithiasis. Treatment depends on its severity and extension, ranging from conservative treatment in mild cases to urgent surgery in cases with hemodynamic instability.

We present the case of a 58-year-old male patient with previous inpatient history of acute urinary retention and the finding of right coralliform renal lithiasis, without accepting surgical treatment and was readmitted a few weeks later presenting the characteristic clinical Lenk´s triad he undergoing to simple abdominopelvic computed tomography TC, as initial study and reporting a right perirenal hematoma secondary to obstructive uropathy due to renal lithiasis.

KEYWORDS: Spontaneous renal hemorrhage, Wünderlich syndrome, Spontaneous renal rupture, Spontaneous retroperitoneal hemorrhage, Coralliform lithiasis.

ARTICLE DETAILS

Published On:
16 November 2023

Available on:
https://ijmscr.org/

INTRODUCTION

Wünderlich syndrome or spontaneous retroperitoneal hemorrhage of renal origin is an extremely rarely entity that mostly occurs in unhealthy kidneys and without a history of trauma, distinguished by presenting as a self-limited subcapsular or perirenal blood collection [1,2]. It was first described in 1856 by Carl August Wunderlich, defining it as a spontaneous apoplexy of the renal capsule [3]. Clinically, spontaneous renal rupture occurs mainly in one kidney [2]. Wünderlich syndrome puts the renal function and the patient's life at risk, and it may be underdiagnosed, so is very important the early detection of the clinical signs with the Lenk's triad that consists in sudden onset severe abdominal pain, increased right renal fossa volume and signs or symptoms of hemodynamic shock. The diagnosis is usually made by imaging methods such as CT which evidences the presence of perirenal subcapsular hematoma. However, the CT is unable to determine the etiology of the syndrome, so the histopathologic report is the method of choice to demonstrate the cause of renal hemorrhage [3]. Treatment varies from conservative to surgical management depending on the severity of symptoms and hemodynamic condition, generally in cases without hemodynamic instability patients respond well to conservative management but in case of failure, surgical treatment is indicated. The prognosis is linked to the early recognition and intervention of the pathology, so this review will increase the knowledge of an adequate detection and diagnostic-therapeutic approach

CASE REPORT

We present the case of a 58-year-old male patient with no relevant family history, hypertensive and diabetic of long evolution in therapeutic control for both pathologies, with the following surgical history: appendicectomy in 2007, inguinal plasty in 2018, other history without relevance to the current condition.

He refers to start his symptoms 4 months prior to our evaluation with the presence of colic type pain in right renal fossa with 8/10 intensity, without irradiation, not associated with fever, vomiting or chills, which improved partially after the administration of nonsteroidal anti-inflammatories. However, one week prior to our assessment, he reported a pain aggravation with increasing intensity, which did not yield to previous measures, this is why he decided to go to the emergency department of our hospital, presenting with the following vital signs T/A: 120/70 mmHg, HR: 70 bpm, FR 20 rpm, physical examination highlighted the presence of a palpable bladder, acute urine retention and pain in right renal fossa at renal percussion, rest of abdominal examination without alterations. The following laboratory studies were performed: Leukocytes 6.95 K/ul, Platelets 520 K/ul, Hemoglobin 7.9 gr/dl, Hematocrit 39.2%, Glucose 192 mg/dl, Creatinine 0.92 mg/dl. With suspicion of right urolithiasis, a abdominal CT scan was requested, showing coralliform lithiasis in the right kidney (Fig 1).

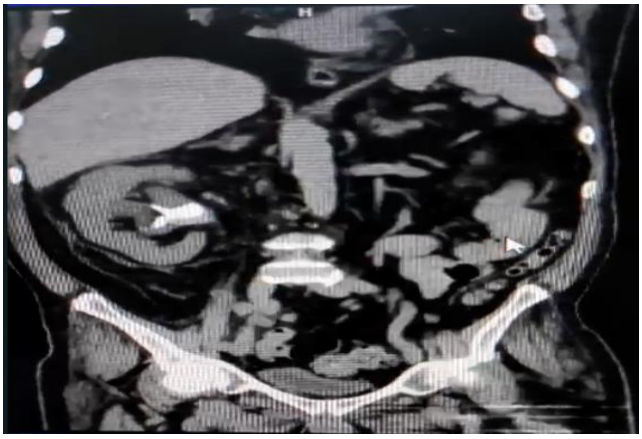

Figure 1. Simple abdominopelvic tomography coralliform lithiasis in the right kidney is observed.

zAcute urinary retention was resolved with urinary urethral catheterization and a protocol for surgical treatment was established; however, due to the patient's refusal, he was discharged.

Two weeks after his voluntary discharge, he presented sudden pain in the right renal fossa with greater intensity 10/10, radiating to the right hypochondrium, asthenia and adynamia, admitted on this occasion without symptoms of hemodynamic instability, vital signs within normal parameters, highlighting on physical examination an increase in right flank volume were observed, new laboratories were taken: Leukocytes 10.79, Hemoglobin 6.8 gr/dl, Hematocrit 22.3%, Platelets 576, Urea 92.8 gr/dl, Creatinine 2.06 gr/dl. Due to the decrease in hemoglobin levels in comparison to previous tests, new CT scan was performed and heterogeneous density right kidney with irregular edges associated with the presence of perirenal collection with homogeneous aspect suggestive of a right perirenal hematoma, left kidney without alterations was observed (Fig 2).

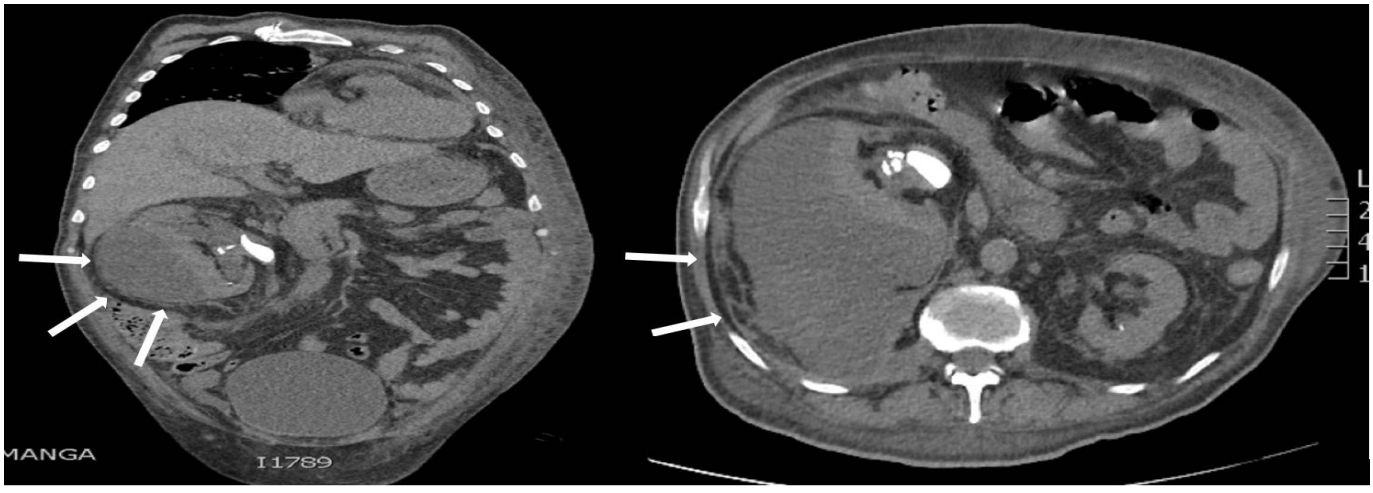

Figure 2. Simple abdominal tomography. A) Coronal plane showing right perirenal hematoma limited by perirenal tissue with adjacent coralliform lithiasis (Arrows). B) Axial plane again showing a contained right perirenal hematoma and coralliform lithiasis (Arrows), the contralateral kidney without apparent alterations.

Apart from the perirenal hematoma, our patient always remained hemodynamically stable, and it was decided, in the absence of surgical emergency data, to treat him conservatively with improvement of general conditions prior to the surgical procedure, transfusing two globular units. However, during his hospitalization he presented sudden deterioration of his respiratory function which had a rapidly progressive course culminating in need of invasive mechanical ventilation and later death of our patient, due this the definitive surgical treatment was impossible.

DISCUSSION

Wunderlich syndrome is defined as a spontaneous renal hemorrhage in the subcapsular, perirenal and/or pararenal space without traumatic antecedent [4].

It´s an extremely rare condition, with less than 500 cases reported to date, so its actual incidence is currently unknown. There are not enough documented records to clarify a specific etiology, it has been determined that it can originate from multiple entities, the tumoral etiology is responsible for 57-63% of total cases, with angiomyolipoma standing out, especially if it is larger than 4 cm.

Vascular conditions such as renal artery aneurysm account for 18-26% of cases, and 11% are due to inflammatory causes [5]. Other rare causes are systemic diseases such as polyarteritis nodosa, rupture of renal cysts, coagulation disorders, infections of the renal parenchyma, lithiasis, hemodialysis, among others [5, 6]. Hypertension, diabetes, obesity, autoimmune diseases, and coagulation disorders have been associated as risk factors for developing perirenal hematoma [2].

Spontaneous kidney rupture can be classified according to its location: Renal parenchymal, collecting system or mixed rupture; the most common being the renal pelvis localization [7]. Parenchymal rupture usually occurs in renal cell carcinoma, angiomyolipoma, renal cysts, arteriovenous malformations, or vascular diseases [8].

There are reports that spontaneous kidney rupture occurs when the normal intrapelvic pressure of 25-75 mmHg is exceeded, being a predisposing factor the urinary obstruction [9], which could have conditioned the renal rupture in our patient due he also presented acute urinary retention associated with his partial coralliform lithiasis. Acute or chronic urinary retention can generate retrograde urine flow into one or both ureters and sometimes into the kidneys, which can lead to irreversible renal failure due to tubulointerstitial nephritis or even spontaneous rupture of the collecting system. Rupture of the collecting system is a rare complication that can be seen in cases of obstructive urinary lithiasis [4], usually located in the distal third of the ureter, and can be associated with urinomas or immediate life-threatening retroperitoneal hemorrhage [8]. They may appear on CT as a perirenal hematoma, hydroureteronephrosis or as an obstructing renal or ureteral calculus [4]. Our patient had this history, which may explain the origin of the spontaneous renal hemorrhage, without being able to determine the complete resolution of urinary retention with the urinary catheter since he requested voluntary discharge.

There is no typical symptom or specific pathognomonic sign, abrupt and intense flank pain is the most frequent symptom, present in 40-75% of cases. There may be hemodynamic instability (25%), fever with leukocytosis (10-23%), and renal failure (2-13%) [6]. The principal characteristic of Wünderlich syndrome is the presence of Lenk's triad, but this has only been reported in 20% of cases, being associated with the progression of the hematoma, which interferes when determining the therapeutic strategy to follow [10]. There may be laboratory alterations such as decreased hemoglobin and hematocrit, leukocytosis, increased ESR, LDH, urea and creatinine [5].

The definitive diagnosis is by imaging tests, abdominal and pelvic CT scan can quickly diagnose the origin of retroperitoneal spontaneous hemorrhage in 92.6% of cases, compared to abdominal ultrasound, which has a 40.9% certainty to define the diagnosis [2, 5]. In addition, it shows the extent of bleeding and the underlying cause. Acute hemorrhage is seen as a hyperattenuating liquid collection of 30-70 HU or in contrast studies as contrast extravasation and pseudoaneurysms. It is for this reason that, when retroperitoneal hemorrhage is suspected, the initial study of choice is CT. Ultrasound is highly sensitive to identify perirenal hematoma and is useful for monitoring the evolution of the hematoma, being able to report as a finding an isodense to hyperdense avascular mass, in the acute stage as a liquid collection or as a heterogeneous hypoechoic or anechoic collection with septum's in subacute and chronic stages [4]. Other useful studies when there is diagnostic doubt are magnetic resonance imaging (MRI) or selective arteriography.

Treatment must be individualized for each case, where the choice will depend entirely on the cause and the hemodynamic status of the patient at the time of initial evaluation. If the patient is stable the choice is for conservative management with volemia replacement, continuous monitoring, and serial radiological assessment of the extent of the hematoma in addition to the treatment of choice for the underlying etiology [10]. If there is sustained hemodynamic instability or the cause can be clearly resolved surgically, urgent exploratory treatment is chosen, with selective arterial embolization, radical or partial nephrectomy, last ones being performed more frequently in cases of Wünderlich syndrome associated with tumors [2]. The treatment for hematomas due to systemic conditions is conservative, unless there is an over added surgical pathology. Selective arterial embolization has proven to be effective and safe in treating renal hemorrhage of any etiology, with significant value in controlling spontaneous

renal hemorrhage, avoiding nephrectomy. Its disadvantages are that there is a risk of requiring a second embolization in uncontained renal hematomas or ischemia of the renal parenchyma with subsequent loss function [2].

The prognosis is generally good in cases with an early detection, so it is important to consider Wünderlich syndrome in the differential diagnosis of acute low back pain in patients with risk factors and provide treatment as soon as possible.

There are no differences in renal function outcomes between high-dose intravenous corticosteroids versus 1 mg/kg/day oral prednisone for 2 to 3 weeks, followed by gradual tapering over 1 to 3 months [16].

Prognosis depends primarily on the cause of ATIN, in combination with treatment for systemic disease, timing of treatment, previous renal function, and clearance of any known offending agents.

CONCLUSION

Spontaneous retroperitoneal hemorrhage of renal origin or Wunderlich Syndrome is a condition that can be life threatening if the diagnosis is not established at time, especially when it presents with hemodynamic instability that doesn´t respond to initial resuscitation measures.

It is important to be aware of the existence of this pathology despite a few reports that have been made to date. The main factors are patients with renal tumors larger than 4 cm, regardless of its benign or malignant behavior, vascular diseases or coagulopathies, as well as systemic diseases and renal lithiasis, among others; that, associated with clinical data or signs of acute abdomen or hemodynamic instability, practically make the diagnosis. Not ignoring its existence and recognizing in time could improve the prognosis of this type of patients and decrease the mortality rates associated with this pathology.

CONFLICT OF INTERESTS

The authors have declared no conflicts of interest.

REFERENCES

I. Wang BH, Pureza V, Wang H. A tale of Wünderlich syndrome. J Surg Case Rep. 2012;11:1-3. https://doi.org/10.1093/jscr/rjs015

II. Tang W, Yang D, Wu T, Liang G. Delayed bilateral spontaneous renal rupture after surgery for unilateral upper ureteral calculi: a case report. Front Med, 2023;10:1-7. https://doi.org/10.3389/fmed.2023.1173386

III. Segura A, Barreda A. Sindrome de Wünderlich (hemorragia renal espontánea). Rev Fac Med Univ Nac Auton Mex, 2021;64(6):26-31.

IV. http://doi.org/10.22201/fm.24484865e.2021.64.6.03

V. Shah JN, Gandhi D, Prasad SR, Sandhu PK, Banker H, Molina R, et al. Wunderlich syndrome: comprehensive review of diagnosis and management. Radiographics, 2023;43(7):E220172. https://doi.org/10.1148/rg.220172

VI. Gimeno V, Bosquet M, Ramirez M, Trassierra M, Arlandis S, Jimenez JF. Hemorragia retroperitoneal espontanea: nuestra experiencia en los ultimos 10 años. Actas Urol Esp, 2007;31(5): 521-527.

VII. Franco D, Hernandez MA. Hemorragia renal no traumática (síndrome de Wünderlich). Informe de un caso. Rev Mex Urol, 2005; 65(4): 284-287.

VIII. Yin G, Pan X, Tian H, Zhou Z, Li J, Tian F, et al. Spontaneous renal rupture due to renal calculi: a case report and literature review. Exp Ther Med. 2022; 24:588. https://doi.org/10.3892/etm.2022.11525

IX. Chiancone F, Meccariello C, Ferraiuolo M, De Marco GP, Fedelini M, Langella NA, Fedelini P. A rare case of spontaneous parenchymal kidney explosion in a patient with ureteral obstruction caused by a single stone. Urologia. 2020;88(4):386-388. https://doi.org/10.1177/0391560320975881

X. Li, Z., Beh, J.C.Y. Spontaneous bilateral renal pelvis rupture during CT in the absence of urinary tract obstruction: case report. BMC Urology. 2020; 20:98. https://doi.org/10.1186/s12894-020-00669-4

XI. Escuadra-Gallegos J, Hernández-Remess H, Heredia-Torres B, Hernández-Quiroz J. Síndrome de Wünderlich. Med Int Méx. 2020;36(5): 735-739. https://doi.org/10.24245/mim. v36i5.4010.