Case Report

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Wunderlich syndrome (spontaneous renal hematoma) as a cause of acute abdomen: a case report

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ABSTRACT

Spontaneous renal hemorrhage, also known as Wünderlich syndrome, is a rare condition that can be life-threatening and consists of the onset of sudden hemorrhage into the subcapsular and perirenal spaces. It can be lethal as it goes unnoticed and requires aggressive treatment. The entity lies mainly in neoplasms, the malignant ones being the most common. We present the case of a 63-year-old female patient with acute abdomen clinic, a diagnostic laparoscopy was performed and renal hematoma was evidenced as the cause of the symptoms. It was managed conservatively without any complications. We emphasize the importance of keeping in mind Wünderlich syndrome as a differential diagnosis to instigate early treatment for a better outcome.

Keywords: Spontaneous renal rupture, Subcapsular haematoma

INTRODUCTION

Wünderlich syndrome was first described in 1856 by Carl Reinhold August Wünderlich, as a spontaneous renal hemorrhage, that dissects the subcapsular and perinephric spaces. It can manifest with the Lenk's triad: acute abdominal pain in flank, hypovolemic shock, and lumbar palpable mass.¹ Clinical presentation is variable depending on the duration and severity of the bleeding, it can present with mild abdominal pain, or with an acute abdomen emulating acute appendicitis or bowel perforation.²

The most frequent cause of spontaneous renal hematoma (SRH) is tumoral bleeding, mainly the angiomyolipoma (30%) and the clear renal cells carcinoma (55-70%).³ There are also vascular causes, the most frequent in this group polyarteritis nodosa, and there is a small group that is idiopathic.

The main relevance of this pathology is its low incidence and the possibility to simulate other pathologies, including acute abdomen. It must be considered as a cause of acute abdomen and depending on the case it may require surgical treatment. We present a case of spontaneous renal hematoma as cause of acute abdomen.

CASE REPORT

A 62 years old female with antecedents of obesity, diabetes mellitus, hypertension, dyslipidemia and ischemic cardiopathy with chronic ingestion of prophylactic aspirin. She presented with abdominal pain in inferior right abdomen, nausea and vomit to emergency department. At the clinical exam was hemodynamically stable, with generalized abdominal pain mostly in inferior right abdomen. McBurney, Von Blumberg, Dunphy and psoas signs were positive. Laboratory reported hyperglycemia and leukocytosis. Case was in charge of general surgery service suspecting of acute appendicitis, a diagnostic laparoscopy was proposed. During the procedure appendix was identified without signs of inflammation, therefore a complete peritoneal cavity revision was performed, a hematoma was identified dissecting below toldt's fascia from renal fossa to cecum. At renal fossa level, we identified nonpulsatile, soft increase in volume blood collection. Urology service was called for an intraoperative consultation, deciding conservative management of the apparent renal hematoma. During post operatory evolution, patient develops respiratory difficulty, decrease in oxygen saturation associated with elevation of d-dimer. Pulmonary thromboembolism is ruled out with an Angio-CT. Low levels of hemoglobin are confirmed, and blood transfusion was performed recovering levels of hemoglobin according world organization health (WHO) standards. Patient remained stable and without hemodynamical compromise. An abdominal CT was obtained to follow up renal hematoma, observing only a sub capsular hematoma without another associated lesion (Figure 1). Patient evolves without complications with ambulatory management and was discharged after 10 days of hospitalization.



Figure 1: Contrasted computed tomography showing the subcapsular hematoma.

DISCUSSION

SRH is an uncommon pathology that was first described by Bonet in 1679; but was Carl Reinhold August Wünderlich, who described it as a "submit stroke of the renal capsulae" in 1856.⁴

Causes of SRH are multiple, in a metaanalysis conducted by Zhang et al, was observed that 61.5% of the cases were caused by neoplasia (31.5% malignant and 29.7%benign), being angiomyolipoma the most frequent .In second place, there are vascular causes (17%), mainly associated with polyarteritis nodosa (PAN), followed by idiopathic cause (6.7%) and infectious causes (2.4%).⁵ SRH has been associated to some medications , mostly antithrombotic as low molecular weight heparin and aspirin, also secondary to shock wave extracorporeal lithotripsy.^{6,7} In our case, patient had no renal lesion and only had the antecedent of chronic aspirin ingestion.

Due to rare nature and mostly case reports in the literature, well described incidence it is not clear, Ahn et al made a metaanalysis from January 2000 to December 2016, where 102 cases were observed, incidence was predominant in women (61.8% vs 38.2%); renal neoplasias were the principal cause with 56.9% of the cases; 74.1% of them were angiomiolipoma, followed by clear cell renal carcinoma in 12.1% of cases.³ In our case, patient had no renal lesion and only had the antecedent of chronic aspirin ingestion.

Clinical presentation of Wünderlich syndrome is diverse and easily can confuse in the initial evaluation of patients. Lenk's triad ocurrs in 20% of cases, it consists in abdominal pain (67%), hypovolemic shock (26.5%) and palpable tumor in flank (30-50%).⁸ SRH can present as acute abdomen simulating an acute appendicitis or bowel perforation.9 It can present with other signs and symptoms like nausea, vomit, hematuria (40%), fever and leukocytosis (10-23%). Because of the low specificity of the symptoms and the low suspicion, the diagnosis is complex. SRH is mostly an exclusion diagnosis, usually it is not considered as the main diagnosis until complementary studies as ultrasound, CT, angiography or MRI are available.¹⁰ In the presented case, SRH was not considered initially as a differential diagnosis, because overlapped with acute appendicitis presentation.

Abdominal ultrasound has a limited utility in the diagnosis of SRH, and CT has been suggested as a better diagnostic method. Zhang et al, found a 56% sensitivity for abdominal ultrasound and 100% sensitivity for CT.⁵ A 57% sensitivity and 82% specificity was reported for cause determination for CT. Both, abdominal ultrasound and CT are the preferred studies in patients with suspicion of renal hematoma, particularly; CT in urography phase is the ideal approach in case of traumatic renal injury suspicion and, could be extrapolated for the SRH.¹¹ Renal angiography has utility in diagnosis and treatment of renal hematoma.¹² In this case, initial diagnostic approach was not useful to suspect SRH, and CT was a useful study in a second time to confirm the diagnosis and follow up.

For the initial management with renal hematoma there are three main approaches: conservative management, embolization and surgical management.³ Conservative management consists in observing the patient and maintaining hemodynamic stability with seriated CT until hematoma resolution or decision of an invasive management. Conservative management is preferred in patients with hemodynamic stability, as in the presented case and it has been more used recently.² Minimal invasive managements with arterial embolization has been the preferred invasive approach. The management with arterial embolization allows to treat patients with a neoplasic cause, in any cases being the definitive treatment.¹³ The surgical management in the past was preferred due to high incidence of neoplasic cause in SRH, it actually has been replaced by spectating and the minimal invasion management.3 The World society of (WSES) and the emergency surgery American association for surgery of trauma (AAST) recommend the management with angioembolization in cases of expansive perirrenal hematoma.¹⁴ In centers with low availability of minimal invasion approaches, in patients with focal lesions or complications as perirrenal abscess, radical or partial nephrectomy are the options to consider.¹⁵ Image guided percutaneous drainage has been reported as a minimal invasion approach for treatment, but it hasn't been formally studied.¹⁶ In the practice, there is no clinical guideline for SRH management, partially because of its low incidence; nevertheless classifications and management of traumatic renal hematoma can be used.¹⁶ The WSES and the AAST have developed classifications for the management of renal traumatic lesion.¹⁴ According to the WSES and the AAST, a renal hematoma in a stable patient corresponding to WSES I and AAST I-II as the described in this case, the recommended management, with 1B level of evidence, is the non-surgical management.14

CONCLUSION

SRH is an uncommon condition, with no specific clinical presentation and it is possible to find it as an acute abdomen etiology. This entity can be suspected in patients with risk factors like use of anticoagulation or aspirin intake, or a related comorbidity. CT is the best initial study in patients with suspected diagnosis. It is important for the surgeon to be familiarized with diagnosis and management of traumatic and nontraumatic renal lesions to guarantee an opportune diagnosis and avoid unnecessary interventions. Conservative management and minimally invasive approach have replaced surgical management of SRH.

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