IJMPR 2023, 7(7), 01-04

International Journal of Modern Pharmaceutical Research

www.ijmpronline.com

ISSN: 2319-5878
IJMPR
Case Study

SJIF Impact Factor: 5.273

WUNDERLICH SYNDROME, A RARE AND INTERESTING CASE WITH ITS RADIOLOGICAL PRESENTATION

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Received on: 12/05/2023
Revised on: 02/06/2023
Accepted on: 22/06/2023

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ABSTRACT

Wunderlich syndrome is a rare but potentially life-threatening clinical condition that presents as an atraumatic spontaneous hemorrhage into renal subcapsular and retroperitoneal region. Its clinical presentation includes a triad of symptoms known as Lenk's triad and comprises of acute onset flank pain, a palpable flank mass and hypovolemic state secondary to internal bleeding. We present the case of a 73-year-old non-compliant hypertensive patient who presented with left upper quadrant pain and vomiting and was found to have perinephric hematoma, secondary to active bleeding from aneurysms in a renal angiomyolipoma. She was managed with selective angioembolization. Post embolization clinical course was uneventful and patient recovered smoothly.

KEYWORDS: hemorrhage, renal, retroperitoneal, Wunderlich syndrome.

INTRODUCTION

One of the most common complaints that patients present in ER with is acute abdominal pain. Although rare, but potentially fatal, Wunderlich syndrome has a similar presentation. It is the atraumatic spontaneous retroperitoneal hemorrhage most often associated with an underlying renal pathology such as malignancy, angiomyolipoma, infection and arteriovenous malformations. It is therefore crucial to reach a prompt diagnosis using appropriate investigations in order for timely management, whether conservative, interventional or surgical.

CASE PRESENTATION

A 73-year-old female presented to our emergency department with complaints of a dull, non-radiating, left upper quadrant abdominal pain, increased urinary frequency, nausea and vomiting for the past 3-4 days. No history of associated fever, hematuria or trauma. She was a known hypertensive and diabetic albeit non-compliant.

Upon arrival, the patient was afebrile, blood pressure was 120/80mmHg, pulse rate was 94bpm, respiratory rate of 18bpm and maintaining oxygen saturation of 98%. Systemic examination was unremarkable except mild tenderness over left upper quadrant. Baseline blood workup revealed hemoglobin of 8.8 g/dL among other normal results.

A baseline ultrasound showed a large predominantly echogenic area measuring approximately 10.0x6.1cm, with minimal vascularity, in the left lumbar region, likely an angiomyolipoma (Figure 1a). Another irregular heterogeneous area with no internal vascularity, measuring approximately 10.3x4.5cm was seen in perinephric region representing hematoma (Figure 1b). Minimal free fluid also noted along hepatorenal and splenorenal angles. All these findings were suggestive of ruptured angiomyolipoma.

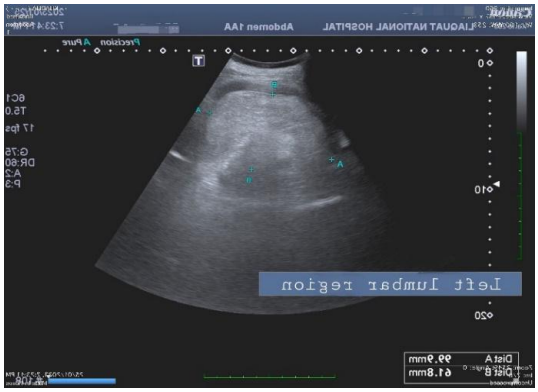

Figure 1a:

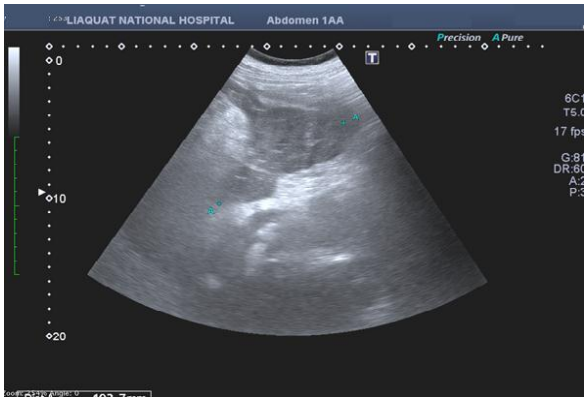

Figure 1b:

Patient was admitted immediately and after the initial workup, the patient's hemoglobin dropped to 4.04 g/dL and an urgent CT scan was performed for further evaluation.

A large exophytic, fat-density lesion was identified arising from the upper pole of left kidney approximately measuring 5.7x6.0cm representing angiomyolipoma (Figure 2a,b). A small high-density area was seen within the angiomyolipoma that raised the possibility of active bleeding site (Figure 3a,b). A large hematoma was seen in the perinephric space measuring about 6.4x7.2cm, extending up the left adrenal region, compressing and indenting the left renal parenchyma and left adrenal gland.

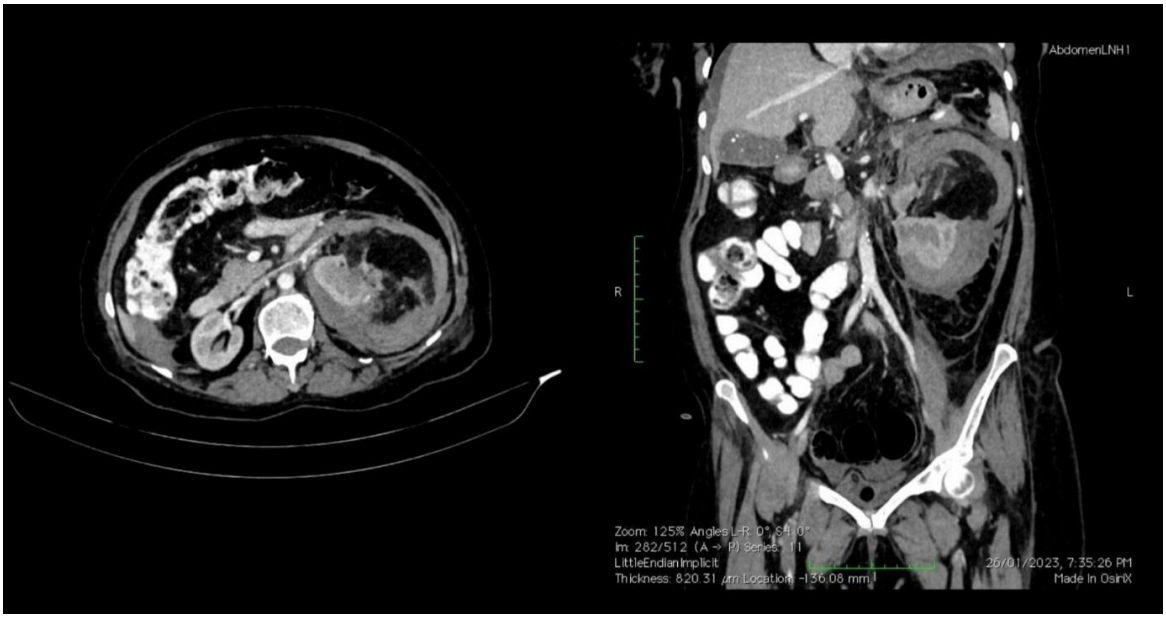

Figure 2a: **Figure 2b:**

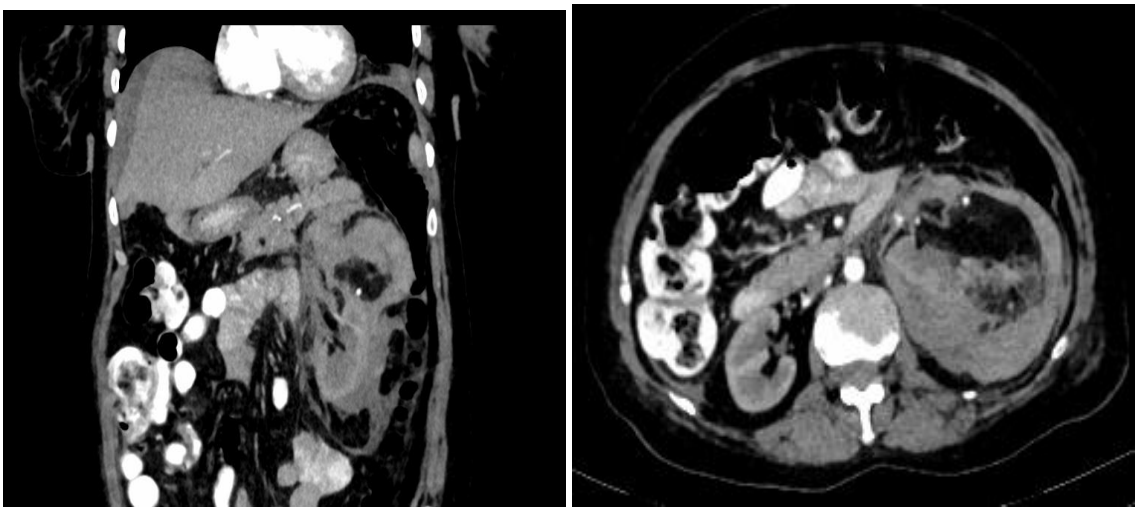

Figure 3a: **Figure 3b:**

Keeping in view the findings of CT Scan, angiography of the patient was planned to look for the possibility of active bleed and its embolization if present.

After written and informed consent, baseline investigations, pre-procedural workup, she was booked for emergency procedure and shifted to cath lab after transfusion of 3 units of PCV.

After all asceptic measures, local anesthesia and fluoroscopic guidance the patient underwent left renal angiography through right femoral artery approach. Tumor blush with multiple aneurysms in the tumor were located at the upper pole of left kidney, which were supplied by superior interpolar artery (Figure 4). It was selectively cannulated, embolization was done using polyvinyl alcohol (Figure 5).

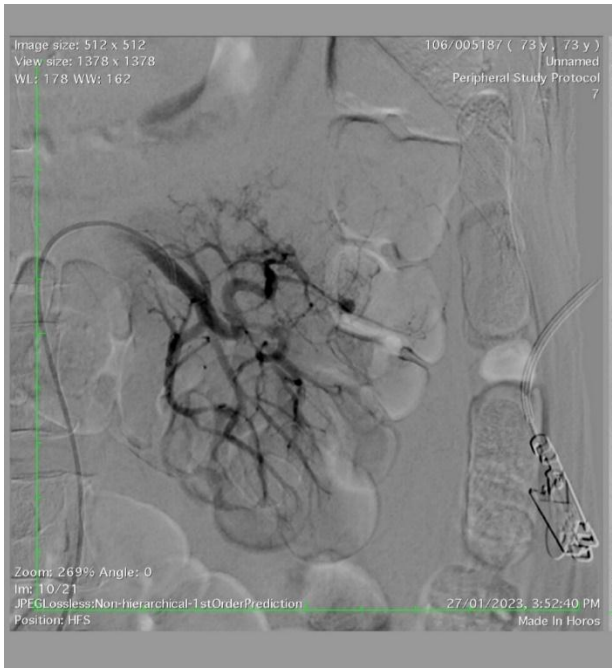

Figure 4:

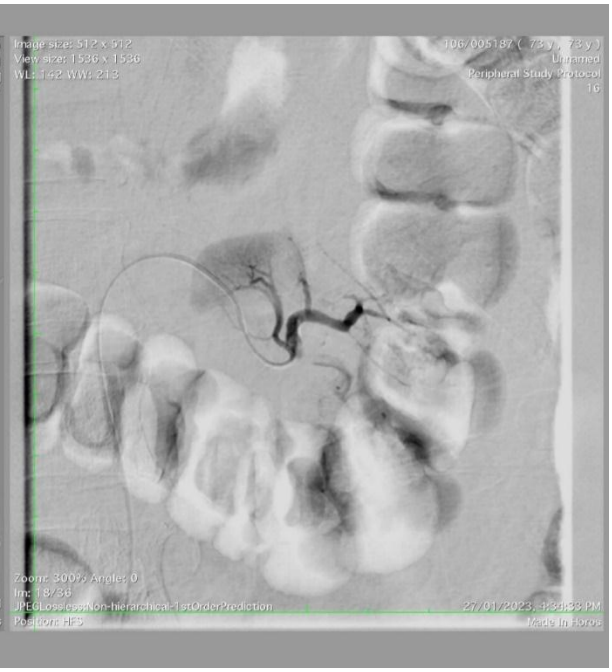

Figure 5:

No immediate post procedural complications were noted.

She was on antibiotics and symptomatic treatment in hospital post-operative course. The recovery was smooth and uneventful. Folley's catheter was removed on third post-operative day and she passed urine normally. She was clinically and vitally stable, hence discharged with outpatient follow-up.

DISCUSSION

Wunderlich syndrome is a rare but potentially life-threatening clinical condition that presents as an atraumatic spontaneous haemorrhage into renal subcapsular and retroperitoneal region.[1] It was first described by Bonet in 1679, although its scientific significance was made famous by Wunderlich in his description in 1856.[2] Its clinical presentation includes a triad of symptoms known as Lenk's triad and comprises of acute onset flank pain, a palpable flank mass and hypovolemic secondary to internal bleeding.[3] It is to be noted, however, that Wunderlich syndrome may typically present with the triad of symptoms in only 20-30% of cases.[4] Other clinical symptoms being fever, hematuria, nausea and vomiting.

The most common underlying pathologies are neoplasms, typically renal angiomyolipoma and renal cell carcinoma (up to 60%).[5] While other possible etiologies are rupture of renal artery, arteriovenous malformation, nephritis, renal calculi, cystic medial necrosis, segmental arterial amyloidosis, polyartritis nodosa, acquired renal cystic disease associated with chronic hemodialysis, cystic rupture, native kidneys of patients who had undergone renal transplantation, coagulopathies or even idiopathic.[6,7] High risk pre-existing clinical conditions for Wunderlich syndrome are diabetes mellitus, hypertension, end-stage renal disease, a history of urinary tract infections, especially pyelonephritis, and renal cystic disease.[8]

It is critical to establish a prompt diagnosis with mindful suspicion on clinical assessment and appropriate investigations, in order to resolve the management dilemma. The differential diagnosis includes rupture of abdominal aortic aneurysm, visceral aneurysm or other visceral bleeding. A POCUS is usually the initial imaging of choice and should be able to evaluate a retroperitoneal hemorrhage with a diagnostic accuracy of 56%, however an abdominal CT is mandatory in hemodynamically unstable patients, as CT has >99% diagnostic accuracy.[9,10,11] CT helps to exclude any pathology in contralateral kidney, as well as other retroperitoneal structures such as the abdominal aorta. MRI may be useful in patients with impaired renal function, which is a contraindication for I/V contrast administration.

Management of the patient depends on the underlying etiology and the hemodynamic status of the patient. Severe hemorrhage requires urgent open surgical intervention and nephrectomy.[10] Angioembolisation plays an important role in securing hemostasis. However, patients with no evidence of active bleeding can be managed conservatively.[12] This approach has mainly been advocated for patients on chronic hemodialysis where the possibility of acquired renal cystic disease and bleeding in the cysts is the likely pathology. This requires accurate and close CT surveillance performed at 3-6 months surveillance to exclude the possibility of underlying renal tumor.[11]

CONCLUSION

Wunderlich syndrome is a potentially fatal clinical entity lest it is handled with great suspicion and immediate intervention. The patient may present with the classical clinical presentation and Ct imaging is the gold standard investigation to confirm the diagnosis and identify the underlying pathology. Determined by the hemodynamic status of the patient, conservative or surgical approaches can be decided.

Explicit informed written consent has been obtained from the patient to share these clinical data for purposes of medical education.

Ethical Review

This case report was written and radiological images and other details were added after taking consent from patient. Patient's identity is not shown here and will be kept confidential.

Conflict of interest

The authors declared, there is no conflict of interest.

Funding

No funding or grant support.

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