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Case Report

Wunderlich Syndrome - A Rare Cause of Hypovolemic Shock

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Abstract

Retroperitoneal haemorrhage (Wunderlich syndrome) is a rare cause of hypovolemic shock in emergency patients; however, it is associated with high morbidity and presents a diagnostic challenge in the Emergency Department. It is more common in female patients older than 50 years, and one of the leading causes is a benign renal tumour-angiomyolipoma (AML). Angiomyolipoma predominantly affects only one kidney. Bilateral AML is strongly associated with a rare autoimmune disorder-tuberous sclerosis and occur in 80% of cases of TS. Symptoms caused by growing renal AML are absent to mild and non-specific (intermittent loin, lower back pains, haematuria). Rupture with retroperitoneal bleeding is a frequent complication of AML and occurs in 20% of all cases. Initial presentation of retroperitoneal haemorrhage is remarkably similar to a ruptured abdominal aortic aneurysm (AAA).

Most of the cases remain clinically silent during the growth phase and present to the hospital with a complication in the form of retroperitoneal bleeding. Risk of bleeding increases with the size of the tumour and AML bigger than 4cm are treated with selective embolisation if found on routine scanning. Many cases of AML are diagnosed when a patient is being scanned for other medical problems. Even in an emergency when AML ruptures is still possible to treat it either with selective embolisation or selective nephrectomy, and the majority of patients are making a successful recovery.

Keywords: *Wunderlich Syndrome (Retroperitoneal Haemorrhage); Renal Angiomyolipoma (AML); Loin Pain with Haematuria; Hypovolemic Shock; Selective Embolisation*

Introduction

Wunderlich syndrome is a rare, life-threatening, non-traumatic renal haemorrhage extending into subcapsular and retroperitoneal space. Lenk's triad of symptoms characterises it as acute flank pain, flank mass and hypovolemic shock. The most common causes are renal tumours and number one is a benign tumour - angiomyolipoma (AML) [1]. Other quite rare causes include malignant urothelial carcinomas, vasculitis and renal vascular malformations and coagulation disorders including warfarin overdose. Angiomyolipoma is a highly vascular locally invasive tumour which consists of thick-walled abnormal blood vessels that lack a developed internal lamina plus spindle-like smooth muscle-like cells and adipose tissue [2]. Estimated prevalence (based on ultrasound screening of healthy volunteers) is 0.1 per cent of men and 0.2 per cent of women [3].

Case Presentation

63 yo female patient was brought to the Emergency Department from her own home with a history of collapse and confusion following a few hours of mid back pain/flank pain. She was brought by "blue light" ambulance from her home, query ruptured aortic aneurysm. She was hypotensive and complained of central chest pain and back pain.

The patient was looking pale and unwell, and due to the confusion, it was challenging to obtain a full history of presenting complaint. At the same time, one of the members of the AE team has spoken to the patient's daughter as she was unable to obtain a reliable history from the patient. Patient's daughter admitted her Mum was seen by her GP several days ago as she was complaining of bilateral loin pain. GP tested her urine sample, which was positive for non-haemolysed blood (++++), slightly positive for leucocytes and protein (+) and negative for glucose, ketones and nitrates. He diagnosed UTI and prescribed the patient three days course of TMP 200 mg BD.

On examination: Initial vital signs were showing GCS 14/15 (E4 V4 M5), blood pressure of 93/45 mmHg, HR-121/min, Sats 95% on room air and T 36.1. On auscultation chest and heart sounds were normal. Abdominal examination showed slightly distended and generally tender abdomen, although painful responses were not apparent. Percussion of both costovertebral angles and flank palpation caused well documented painful reaction, and the patient started to guard against palpation. Bowel sounds were diminished, and there was no palpable abnormal intraabdominal mass. Bedside portable chest X-ray raised suspicion of a widened mediastinum (Figure 1).

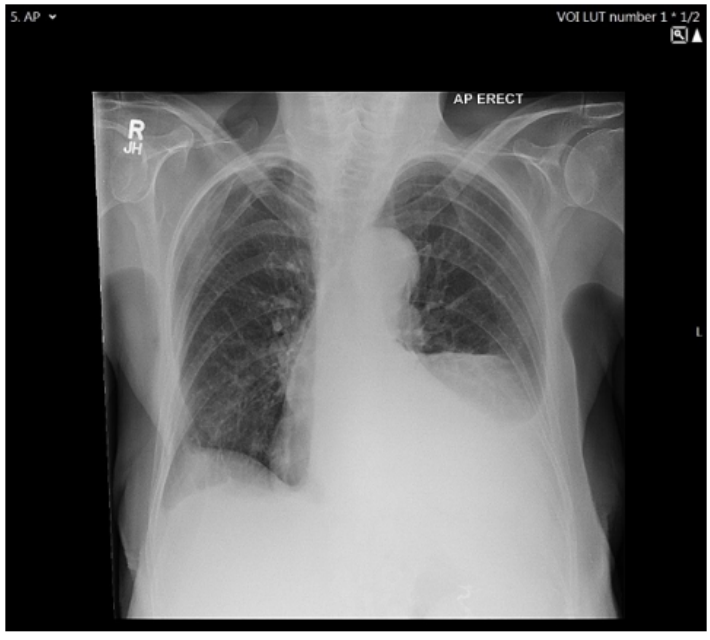

Figure 1: *Portable chest X-ray showing widened mediastinum.*

Treatment was started in AE Resus and the patient was put on high flow O_2, one large-bore cannula was inserted in a peripheral vein with a blood sample sent for FBC, U&E, Group and save and The patient received 1000 mls of 0.9% Saline stat and second 1000 mls bag was started. AE Registrar did bedside FAST and AAA ultrasound scan to check the diameter of the abdominal aorta and asses peritoneal

cavity for free fluid (blood). A scan showed standard size abdominal aorta and no visible free fluid collection in the peritoneal cavity (Figure 2 and 3). The doctor doing FAST scan also commented he had been unable to obtain a clear image of the left kidney (Figure 4).

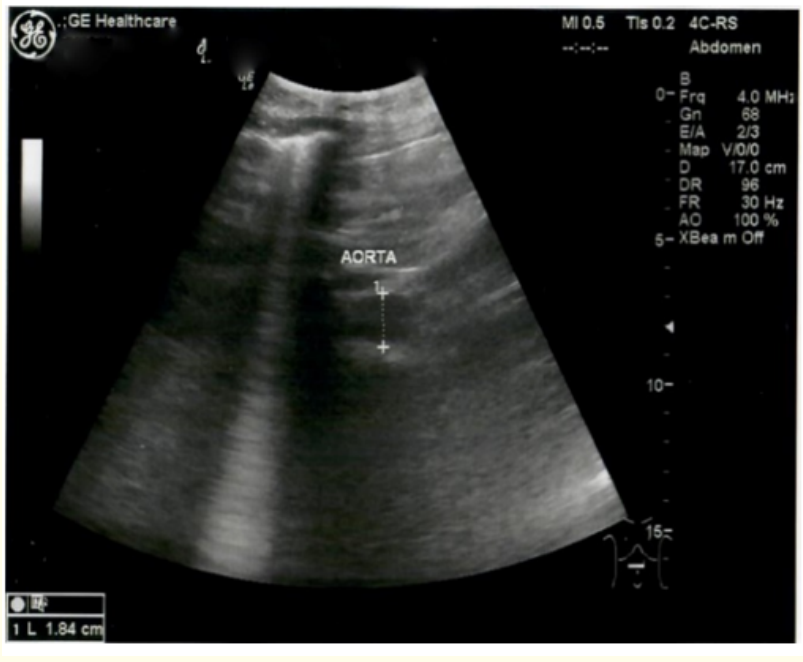

Figure 2: *Bedside USS showing the average size abdominal aorta (1.94 cm).*

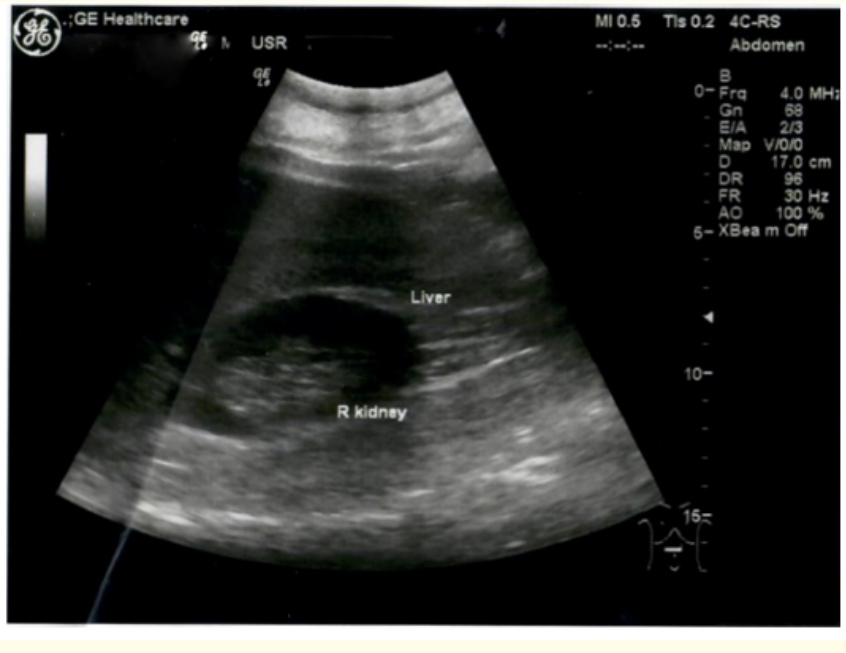

Figure 3: *Normal USS of hepato-renal recess (Morrison's pouch) USS scan.*

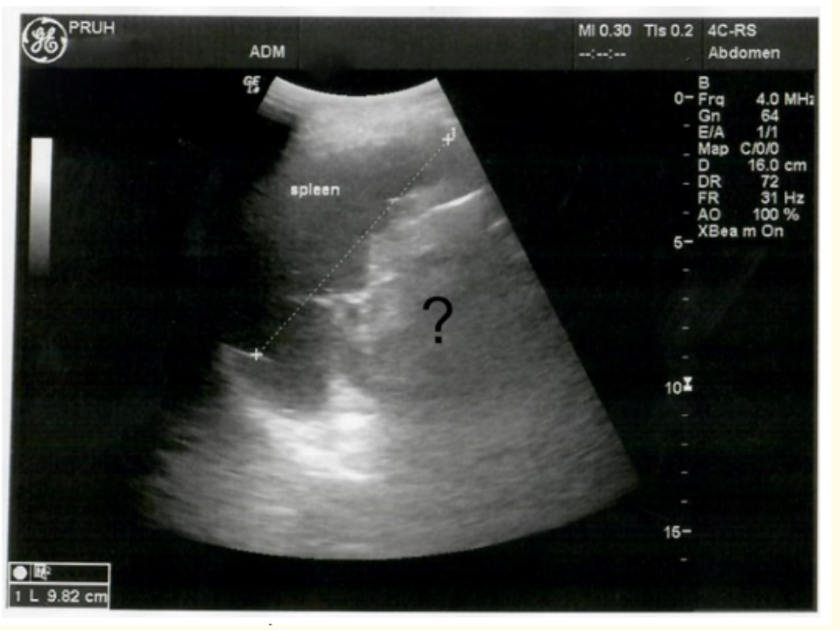

Figure 4: *USS showing spleen hilum and heterogenous mass instead of the left kidney.*

In the meantime, blood test results came back from the lab and showed: HGB 9.1, WBC 12.3, Neutrophils 7.9, PLT 150, Creatinine 150, Urea 8.9, Na 146 and K 4.3 and INR of 1.1.

The patient started to be more lucid, and her vital signs improved with BP 110/74, HR-92 and Sats 98% on room air. Upon discussion with EM Consultant patient was transferred to the Radiology department to have CT aortogram as the working diagnosis was dissecting thoracic aortic aneurysm. Contrast CT aortogram was reported as 12.5 cm heterogeneous mass in left upper quadrant (retroperitoneal space) displacing spleen anteriorly and pushing up left hemidiaphragm - sizeable retroperitoneal haematoma (Figure 5). There was also 15 mm fatty lesion within the upper pole of the left kidney with a prominent arterial vessel, and Consultant Radiologist described the lesion as AML (Figure 6).

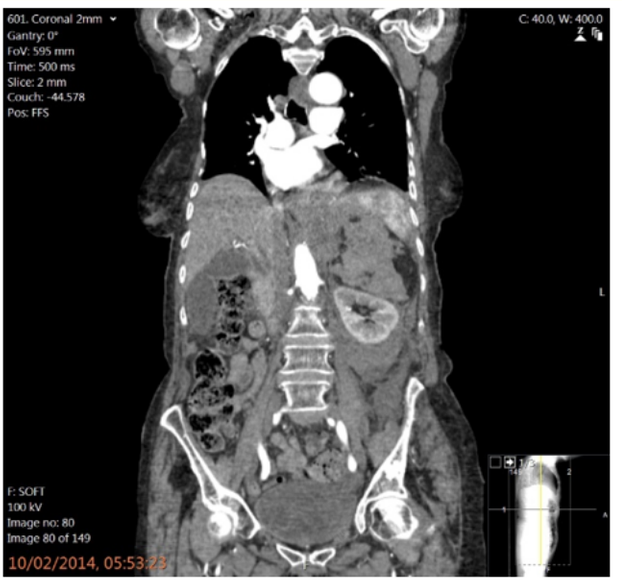

Figure 5: *CT coronal view showing a large retroperitoneal haematoma (white arrow) and causing displacement of the spleen and diaphragm.*

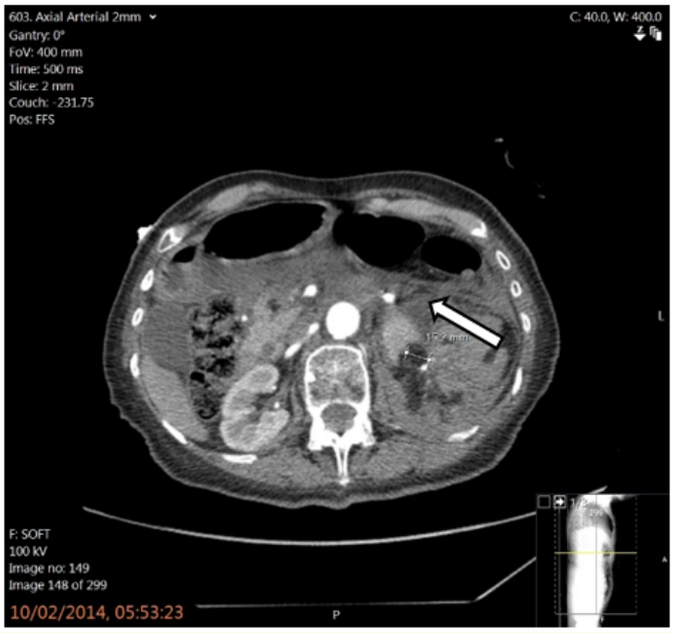

Figure 6: *CT transverse view showing 15mm fatty lesion with arising from an upper renal pole (white arrow).*

Treatment and outcome

The patient was referred and admitted by a local Urology team. Few hours after she was transferred to tertiary referral hospital and had a successful selective embolisation of abnormal arterial vessel, which was also the main supply for the tumour (Figure 7 and 8).

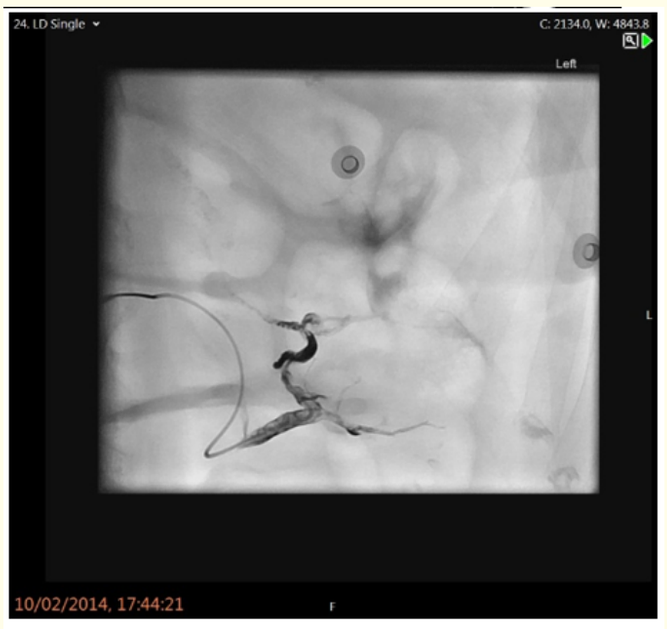

Figure 7: *Visible bleeding (leaking contrast) from an abnormal arterial vessel which was the main blood supply to the tumour.*

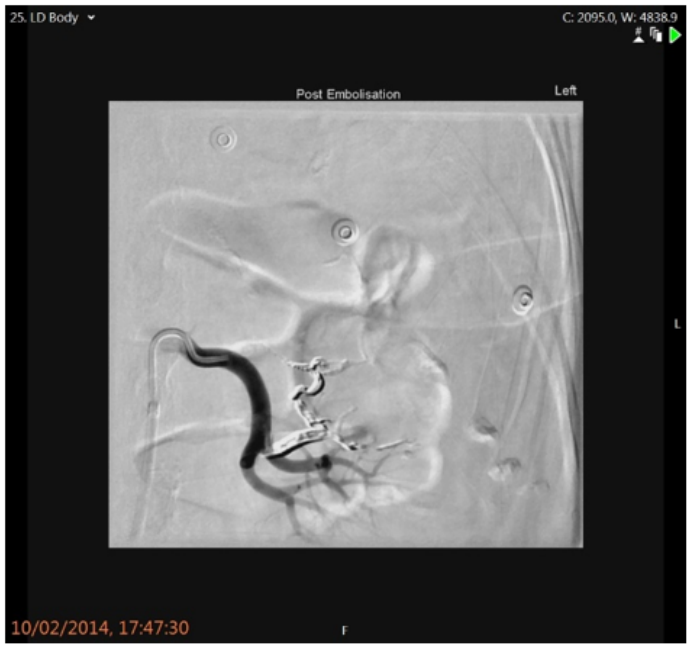

Figure 8: *Showing closed pathological artery and a good contrast flow in remain branches of left renal artery renal artery.*

She was discharged home five days later with no clinical symptoms, mobilising independently, and Creatinine level of 110. Urology team asked her GP to repeat FBC and renal function tests four weeks after the discharge date.

Discussion and Conclusion

Wunderlich syndrome is a rare cause of hypovolemic shock, and presentation resembles abdominal aortic aneurysm rupture. The most common cause of the syndrome is a locally invasive renal tumour -angiomyolipoma (AML). Renal AML is more common in women, and most symptomatic cases are discovered in middle age [4]. Although renal AML is much more common among patients with tuberous sclerosis (75 - 80 per cent) due to the low prevalence of tuberous sclerosis, a spontaneous renal AML is the most common scenario [5]. They possess a diagnostic challenge as the majority of small renal AML are asymptomatic, and patients have a normal renal function, and microscopic haematuria is not common [6]. Symptoms are more likely to occur in renal AML bigger than 4 cm [6]. The most common symptoms are ongoing flank pain and gross or microscopic haematuria. In the case of a spontaneous rupture, the preferred therapy should be renal artery embolisation [7].

However, in this case, the patient underwent open nephrectomy, and it was due to the lack of a hospital bed at the tertiary referral centre hospital. Small or incidentally detected renal AML could be observed with close follow up (at six months) and if stable, then yearly [8]. Asymptomatic lesions bigger than 4 cm are managed by embolisation procedure or partial nephrectomy [7,8].

Learning Points:

When assessing a patient with hypovolemic shock:

- Wunderlich syndrome as a cause of hypovolemic shock is almost twice as common in perimenopausal female patients than in men of the same age.
- During resuscitation, phase perform ultrasound FAST and AAA scan alongside with treatment.
- If FAST and AAA scans are both negative, consider the possibility of retroperitoneal haemorrhage (Wunderlich syndrome) in patients with flank pain or gross haematuria.
- Clinical presentation of retroperitoneal haemorrhage may be confusing as symptoms can be multiple, and assessment can be difficult due to reduced contact with a patient.
- Renal angiomyolipomas smaller than 4 cm are mostly asymptomatic and frequently are detected on renal ultrasound or CT that is obtained for an unrelated reason.

Differential diagnosis of hypovolemic shock and flank pain:

- Ruptured abdominal aortic aneurysm
- Sepsis due to obstructed nephropathy
- Retroperitoneal bleeding due to renal tumours (most common being renal AML)
- Retroperitoneal bleeding due to vasculitis, renal AV malformation, over coagulation with warfarin.

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2. Dr Swetha Rao MBBS, FRCEM Consultant in Emergency Medicine, Princess Royal University Hospital, Orpington (part of Kings College NHS Foundation Trust).

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