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Case Report
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Wunderlich's Syndrome due to Ruptured Renal Angiomyolipoma

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Abstract

Wunderlich's syndrome or spontaneous renal hematoma is rare condition but can be life threatening. Renal tumours are the most common etiology. CT plays a vital role in identifying the perirenal hematoma and diagnosing the underlying cause. Selective arterial embolization is the current treatment of choice in ruptured renal angiomyolipoma, especially in life threatening cases.

Keywords: Wunderlich's syndrome; Angiomyolipoma; Bleeding angiomyolipoma; Renal tumour

Case Report

A 45 year-old Malay female, with underlying hypertension, complained of acute onset, severe left loin pain 1 hour prior to presentation, associated with nausea and an episode of vomiting. No history of trauma, hematuria or abdominal mass prior to presentation. Clinically, she was pink. Her blood pressure was 194/101 mmHg, heart rate was 59 bpm, temperature was 37°C and oxygen saturation was 98%. There were few episodes of hypotension. Her blood pressure normalized after fluid resuscitation. There was tenderness and presence of vague mass at left lumbar region but no signs of peritonism.

Initial blood investigations were: haemoglobin 14.5g/dL, total white cell count $12.39x10^3/mm^3$, platelet count $328x10^9/l$, urea 3.4mmol/L, potassium 4.2mmol/L, sodium 138mmol/L, creatinine 97mmol/L, INR 1.23, aPTT 30.2 secs, PT 15.4 seconds. Urinalysis showed no microscopic hematuria. Urgent abdominal ultrasound reported a large left retroperitoneal lesion, measuring 13.4cm x 7.3cm with poor demarcation with the left kidney. Initial diagnosis was renal cell carcinoma.

However, her haemoglobin level dropped to 6.3g/dL on the next day of hospital admission. Platelet count was 135. Coagulation profile was normal. She was transfused with 2 pints packed cells and 1 DIVC cycle regime. She proceeds with CT abdomen, which showed presence of angiomyolipoma at midpole of left kidney, measuring 3.8cm x 3.9cm x4.7cm. There was large perinephric hematoma with evidence of active bleed. She underwent selective interlobar arterial embolization of the left kidney using Prograte.

Post-embolization, she recovered well and was discharged home. Repeated CT abdomen was performed 1 month later, showed increased size of angiomyolipoma at left kidney, measuring 6.2cm x 3.5cm x 9.9cm. There was still large residual subcapsular hematoma. She refused for surgical intervention. Currently, she is asymptomatic (Figure 1-5).

Discussions

Wunderlich's syndrome or Spontaneous Renal Hematoma was originally reported by Bonet in 1679 [1] and later described by Wunderlich in 1856 [2]. It is a spontaneous non-traumatic renal haemorrhage into the subcapsular and/or perinephric space. It is a rare clinical entity, but potentially life-threatening condition.

There are various etiologies of spontaneous renal hematoma. 61.5% of cases are due to renal neoplasm's with 31.5% malignant and 29.7% benign renal tumours [3]. Other underlying causes are vascular disease (vasculitis, renal artery aneurysm), infection and hypertension [3]. Renal angiomyolipoma (AML) is the most common benign renal tumour causing spontaneous renal hematoma [3]. In this patient, the cause of spontaneous renal haemorrhage is angiomyolipoma.

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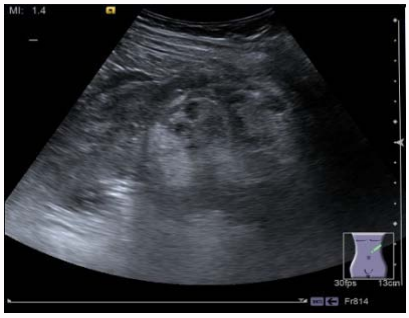

Figure 1: Ultrasound abdomen showed large left retroperitoneal lesion, measuring 13.4cm x 7.3cm with poor demarcation with the left kidney.

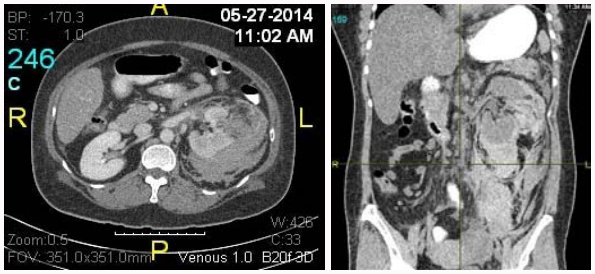

Figure 2: CT abdomen showed angiomyolipoma at midpole of left kidney, measuring 3.8cm x 3.9cm x 4.7cm. There was large perinephric hematoma.

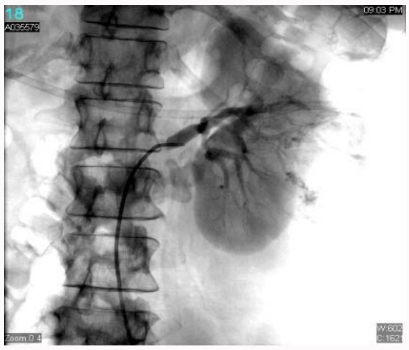

Figure 3: Tumoral blush seen from left interlobar artery which supplied interlobar lobe.

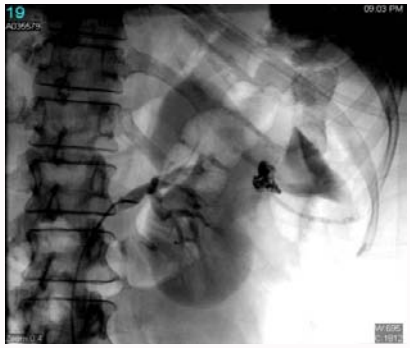

Figure 4: Selective embolization done interlobar artery which supplied interlobar lobe.

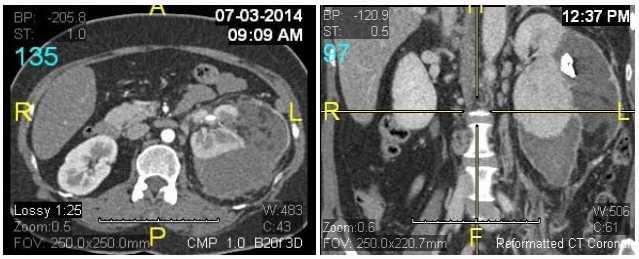

Figure 5: Large angiomyolipoma over the superolateral of left kidney, measuring 6.2cm x 3.5cm x 9.9cm with large subcapsular hematoma at lateral and inferior aspect of left kidney.

It is a benign mesenchymal tumour of kidney, composed of variable amounts of adipose tissue, spindle and epithelioid smooth muscle cells, and abnormal thick-walled blood vessels [4]. The incidence is approximately 1% of surgically removed renal tumours [4], with female preponderance of 80.7% [5]. Most of AML are smaller than 4cm (85%) and are asymptomatic [5]. They usually found incidentally upon radiographic imaging for other causes [5-7]. Tumour greater than 4cm are more often symptomatic and likely to grow faster than do lesions lesser than 4cm in diameter [5]. In this case, the initial size of AML is 4.7cm and later the tumour grows to 9.9cm.

Wunderlich's syndrome occurs in up to 50% of patients with angiomyolipoma larger than 4cm [8,9]. It is one of the most dramatic and life-threatening complications of angiomyolipoma.

Clinical pictures vary depends on the degree and duration of bleeding. The classical presentations include flank pain, a palpable tender mass and signs of internal haemorrhage (such as hypotension, tachycardia, anemia, hematuria), known as Lenk's triad [9,10] as occurred into this patient. In 10% of cases, patients may develop hypovolemic shock due to intense bleeding.

Ultrasound is the first choice of imaging for rapid identification of renal haemorrhage because it is quick and inexpensive [3]. Though, sometimes renal hematoma can be misinterpreted as renal tumour or abscess formation [10,11]. In the case reported, initial diagnosis from the ultrasound was malignant renal neoplasms. CT scan are required to confirm the ultrasonography findings and to identify underlying renal mass [3,11,12]. It is 100% sensitive to detect hematoma [11] and has higher sensitivity and specificity than ultrasound in diagnosing underlying cause. CT scan is able to identify fatty elements in renal mass with perirenal hematoma, which are the features of Wunderlich's syndrome due to angiomyolipoma [12,13]. It also gives added information about the other kidney [11]. Magnetic Resonance Imaging (MRI) is an alternative to CT scan. It can distinguish blood from tumour, and can even identify small tumours that may not be detected during CT scan [3,11].

The decision for the managing patients with Wunderlich's syndrome is based on hemodynamic status and the underlying etiology [10] with the aim to preserve renal function [14]. The treatment choices are either therapeutic embolization or surgery [15]. Angiography with selective embolization is the preferred option in acute haemorrhage due to ruptured AML [3,6,14] to avoid surgery [3,14]. It allows patient's stabilization [14], relieves symptoms and prevents further bleeding [8]. Selective arterial embolization was successful in this patient. Some patients may require nephrectomy if persistent haemorrhage, failed embolization or suspicious of malignancy [14].

Conclusion

Wunderlich's syndrome is one of the most dramatic complications

of renal angiomyolipoma and should be treated promptly and aggressively. Early and accurate diagnosis of this condition is crucial in saving the patients' life. Angiography with selective embolization plays a major role in acute settings of ruptured renal AML.

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