Reminder of important clinical lesson

Wunderlich's syndrome (spontaneous renal haemorrhage)

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Summary

Wunderlich's syndrome is a rare condition characterised by spontaneous non-traumatic renal haemorrhage into the subcapsular and perirenal space. The commonest cause of Wunderlich's syndrome is renal neoplasms, with angiomyolipoma and renal cell carcinoma being the most common benign and malignant lesions, respectively. We report a case of Wunderlich's syndrome in a 46-year-old woman. She presented in the emergency department with the chief complaints of sudden onset of abdominal pain with no significant medical history. Imaging findings and clinical history led to the diagnosis of Wunderlich's syndrome. The patient underwent left partial nephrectomy and recovered without any complications. She is presently doing well.

BACKGROUND

Non-traumatic retroperitoneal haemorrhage is an uncommon but life-threatening clinical entity, which can be due to ruptured abdominal aortic aneurysm, adrenal bleeding, coagulation disorders and, rarely, renal issues. Wunderlich's syndrome is spontaneous non-traumatic renal haemorrhage into the subcapsular and perirenal space in a patient with no obvious underlying cause. The commonest causes for this conditions are renal neoplasms, with angiomyolipoma (AML) and renal cell carcinoma being the most common benign and malignant lesions, respectively.^{1 2} Polyarteritis nodosa and renal infections are the other less common causes.^{1 2} The common symptoms include acute lumbo-abdominal pain, a palpable mass and haemodynamic instability.² Wunderlich's syndrome is one of the most feared complications of renal AML and can be fatal if not managed aggressively. CT is highly recommended to achieve a definitive diagnosis and to determine the approach to follow.

CASE PRESENTATION

A 46-year-old woman presented to the emergency department with a sudden onset of abdominal pain, which was mainly localised around the left flank region. The pain was severe in intensity and not radiating to any other area. There was no history of trauma to the back or abdomen; also there was no significant medical history.

On examination, the patient was anxious, her pulse was weak and thready, pulse rate was 124/min, blood pressure was 84 mm Hg systolic, respiratory rate was 22/min and temperature was 98 °F. Mild pallor was observed. On per abdomen examination there was fullness in the left flank and tenderness was elicited on palpating the left flank region. An examination of the cardiovascular system revealed tachycardia; first and second heart sounds were normal. Respiratory and neurological examinations were within normal limits.

INVESTIGATIONS

The patient's haemoglobin was 8.5 g/dl, total leucocyte count was 9500/μl and differential leucocyte count

showed 78% neutrophils and 22% lymphocytes. Blood urea was 80 mg/dl and serum creatinine was 1.8 mg/dl.

On an ultrasound of the abdomen, a heterogeneous, predominantly echogenic mass lesion was present at the upper pole of the left kidney along with a complex heterogeneous collection in the left perinephric space (figure 1). On colour Doppler imaging, the lesion showed intratumoural vascularity (figure 2). These findings were suggestive of AML with haemorrhage leading to heterogeneous perinephric collection.

To further characterise the lesion and confirm the ultrasound findings, a contrast-enhanced CT scan was done which showed a large, well-defined intensely enhancing soft-tissue mass lesion arising from the upper pole of the left kidney. Areas of fat attenuation were noted within the lesion with no evidence of any calcification, but with evidence of haematoma formation around it (figures 3–5).

Figure 1 An ultrasound of the abdomen showing a heterogeneous, predominantly echogenic, lesion at the upper pole of the left kidney with adjacent complex collection in the left perinephric space.

Figure 2 A colour Doppler ultrasound of the left renal area showing intralesional vascularity, differentiating the echogenic mass from haemorrhage. M, mass; K, lower pole of the kidney.

The size of the lesion was approximately $6.6 \times 4 \times 2.5$ cm. Thus, a diagnosis of ruptured renal AML with perirenal haematoma formation was made.

TREATMENT

On arrival in the emergency department, the patient was resuscitated with a bolus of intravenous fluids; she was shifted to the emergency operation theatre and left partial nephrectomy was done. The histopathology of the renal mass revealed proportions of thick-walled blood vessels intermixed with mature adipose tissue and smooth muscle fibres, consistent with diagnosis of AML.

OUTCOME AND FOLLOW-UP

The patient made an uneventful postoperative recovery. She is under regular follow-up and doing well.

DISCUSSION

The commonest cause of Wunderlich's syndrome (spontaneous renal haemorrhage) is AML.³ In our patient,

Figure 4 An axial contrast-enhanced CT scan of the abdomen at the level of L1 vertebra showing a large, well-defined, soft-tissue mass lesion with areas of fat attenuation arising from the upper pole of the left kidney, with evidence of haematoma formation around it.

the cause for spontaneous perinephric haemorrhage was renal AML. AMLs are benign hamartomas composed of blood vessels, smooth muscle cells, and fatty tissue.⁴ They can be sporadic or in association with tuberous sclerosis.⁵ The sporadic form accounts for 80–90% of cases of AML and is more common in middle-aged women (male:female ratio—1:4). AML in patients with tuberous sclerosis tends to be larger, bilateral and in younger age groups. Our patient had no other features of tuberous sclerosis. The relationship between size and rupture of AML has been well described in the literature. Masses less than 4 cm in diameter are usually asymptomatic, but those larger than 4 cm often cause flank pain, haematuria and anaemia and carry a significant risk of intratumoural or perinephric haemorrhage. On CT, AMLs are well-circumscribed renal masses and

Figure 3 An axial non-contrast CT scan of the abdomen at the level of L1 vertebra showing a soft-tissue mass lesion with areas of fat attenuation arising from the upper pole of the left kidney, with no evidence of any calcification but with evidence of haematoma formation around it.

Figure 5 An axial contrast-enhanced CT scan of the abdomen 1 cm below figure 4 showing a normally enhancing kidney forming a crescent posterior to the mass, with perinephric haematoma.

presence of intratumoural fat (CT attenuation range -20 to -40 HU) is almost diagnostic of AMLs. Wunderlich's syndrome is one of the most feared complications of renal angiomyolipoma and can be fatal if not treated promptly and aggressively. It occurs in up to 50% of patients with tumours larger than 4 cm, as in our case, because of the association with an increased risk of intralesional aneurysm formation and, therefore, a greater possibility of rupture. Radiological imaging plays an important role in establishing diagnosis and cause. Ultrasonography has been found to be only moderately useful in identifying renal haemorrhage and in differentiating the renal mass and clotted blood.^{2 3 6} CT is the method of choice for the demonstration of perirenal haemorrhage (sensitivity of 100%); it has limited sensitivity (~57%) in identifying renal neoplasm when performed at the time of haemorrhage, but has been found to identify all cases of Wunderlich's syndrome due to AML.³ Biopsy is only rarely useful in the diagnosis of renal AML.⁶ Once a patient is diagnosed with spontaneous perinephric haemorrhage due to AML, the treatment options are either surgery or therapeutic embolisation.^{2 6}

Learning points

- ▶ Wunderlich's syndrome should be kept in differential diagnosis of acute abdominal pain with haemodynamic instability.
- ▶ CT is the imaging procedure of choice to diagnose Wunderlich's syndrome.
- ▶ Therapeutic embolisation and nephrectomy are the treatment options for Wunderlich's syndrome.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ahmad M, Arora M, Reddu R, Rizvi I. Wunderlich's syndrome (spontaneous renal haemorrhage). *BMJ Case Reports* 2012;10.1136/bcr-2012-006280, Published XXX

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