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CASE REPORT

Wunderlich's syndrome in pregnancy: a shocking triad

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SUMMARY

Wunderlich's syndrome, non-traumatic renal haemorrhage into the subscapular and perinephric space, in pregnancy, is a very rare clinical entity. We describe a case of Wunderlich's syndrome in a 29-year-old gravida 5 para 4 who presented to our emergency department with sudden onset severe left flank pain. On assessment, she was clinically shocked—hypotensive, tachycardic and perfused poorly peripherally. Ultrasound of the abdomen and pelvis and subsequent MRI of the left kidney revealed a large hypervascular exophytic lesion arising from the left renal pole—appearance consistent with an angiomyolipoma. This specific presentation is clinically characterised as Lenk's triad—acute flank pain, flank mass and hypovolaemic shock. The patient was adequately resuscitated and interventional radiological embolisation of the mass was performed. She went on to have an uneventful pregnancy and delivered vaginally after induction at 38 weeks of gestation.

BACKGROUND

Wunderlich's syndrome, which was described by Wunderlich in 1856, is a rare and life-threatening condition which is characterised by non-traumatic renal haemorrhage into the subscapular and perinephric space.¹ It is a very rare clinical entity in pregnancy and as of yet there is no clear guidance as to how these should be managed in pregnancy.²

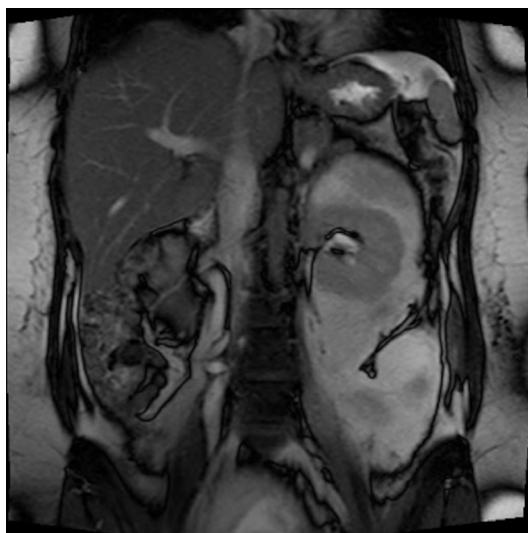


Figure 1 MRI image of abdomen showing left-sided renal mass in the upper renal pole with retroperitoneal haemorrhage.

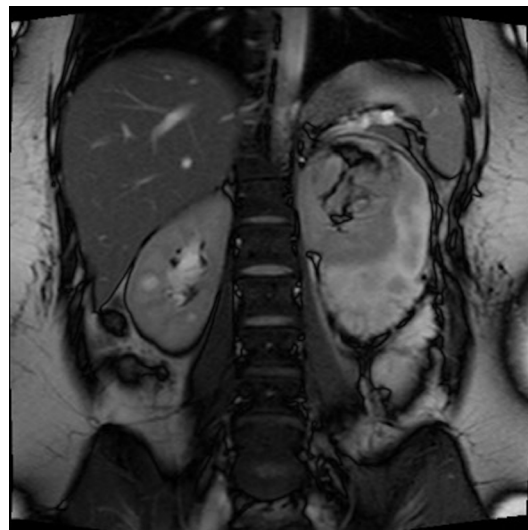


Figure 2 MRI image of abdomen showing the left-sided renal mass in the upper renal pole.

CASE PRESENTATION

A 29-year-old gravida 5 para 4, collapsed at home at 17+3 week's gestation following the sudden onset of severe left-sided flank pain. She had an uneventful pregnancy until this presentation, having booked at 12 weeks of gestation with a singleton intrauterine pregnancy.

She was brought in by ambulance. She gave a history of sudden severe onset of left-sided abdominal pain with no history of any trauma. She had no significant previous medical or surgical history and had no significant family history.

On examination, she was noted to be pale and diaphoretic, apyrexial, blood pressure of 73/38 mm Hg, heart-rate of 95 bpm and an respiratory rate of 14. Her abdomen was soft, with appropriate distension for her gravid state and she had marked left-sided flank tenderness. There was no evidence of ecchymosis.

INVESTIGATIONS

Initial investigations showed a haemoglobin of 87g/L, white cell count of $20.4 \times 10^9/L$, C-reactive protein of 62 and a lactate of 2.23. Urine dipstick showed +3 ketones with no haematuria. A focused obstetric ultrasound showed an intrauterine pregnancy of 17 weeks of gestational age.

An ultrasound of her abdomen and pelvis revealed a 6×7 cm complex mass arising from the left kidney. An MRI scan of her abdomen showed a large hypervascular exophytic lesion arising from the left renal pole, which contained multiple small



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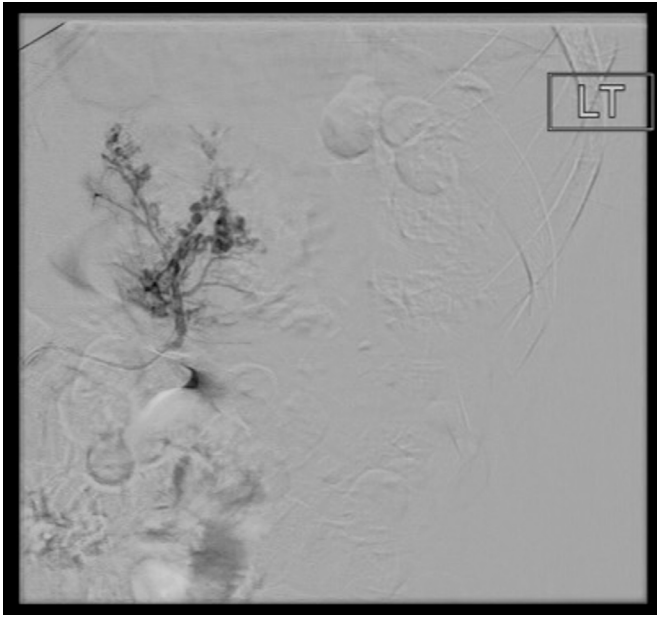


Figure 3 IR fluoroscopy image of left renal angiomyolipoma, prior to embolisation. IR, interventional radiological.

pseudoaneurysms and tortuous abnormal-appearing arteries consistent with an angiomyolipoma (figures 1 and 2).

TREATMENT

The patient was fluid resuscitated and transfused with two units of packed red cells after a repeat haemoglobin of 59g/L. Once

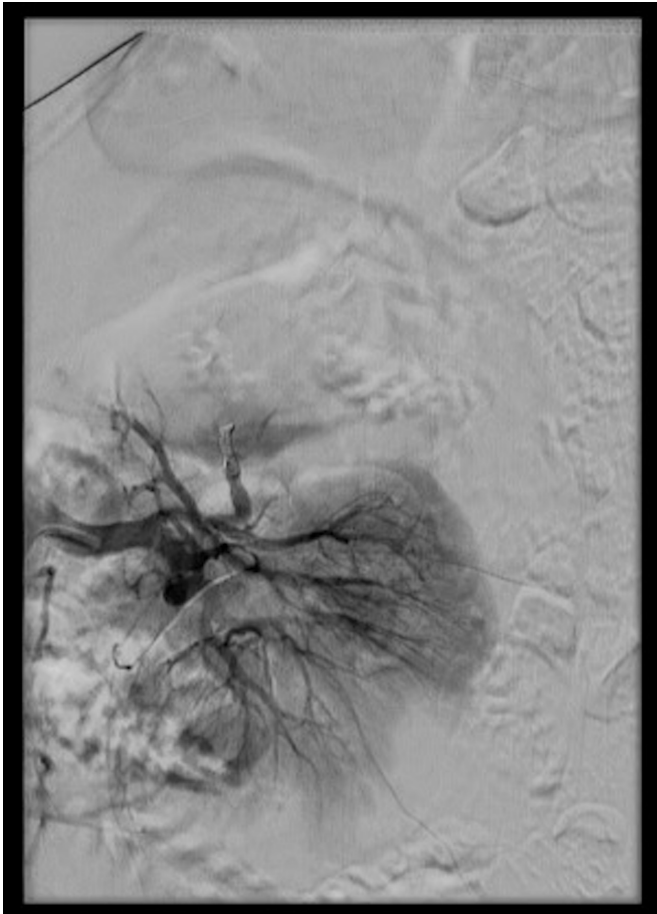


Figure 4 IR fluoroscopy image of left renal AML, postembolisation. IR, interventional radiological.

stabilised and with an improved haemoglobin level, the patient was transferred to a tertiary referral unit, where a urology opinion was obtained. Following review of the patient and the available imaging, the decision was made to recommend interventional radiological embolisation of the tumour's feeding renal artery (figures 3 and 4), in an effort to effectively control haemorrhage while preserving nephrons. The interventional radiologist was aware of the patient's gravid status, and maximal precautions were taken to minimise radiation exposure for the patient and the fetus.

OUTCOME AND FOLLOW-UP

The patient was discharged 4 days postembolisation. She was then reviewed every few weeks in the consultant-led antenatal clinic in our institution. The rest of her antenatal course was uneventful.

She was induced at 38 weeks of gestation and had an uncomplicated spontaneous vaginal delivery. She will be reviewed in the urology outpatient department going forward with post-partum renal imaging, due to her childbearing age and risk of further AML haemorrhage.

DISCUSSION

This is a unique case of Wunderlich's syndrome in pregnancy, as it is the first described case in Ireland and is also one of only four documented cases in pregnancy who went on to have a vaginal delivery with a preserved kidney.

Angiomyolipomas are benign mesenchymal tumours. They are four times more likely in women, and are sometimes associated with tuberous sclerosis (TS). They may have irregular and aneurysmatic blood vessels. The major risk factors for bleeding are tumour size (>4 cm) and the presence of TS. Indications for intervention are pain, bleeding or suspected malignancy. Selective arterial embolisation is an efficient treatment for AML devascularisation and useful in controlling haemorrhage in the acute setting, but often more definitive surgical management is required (eg, partial nephrectomy).³

Wunderlich's syndrome is a feared and sometimes fatal complication of AMLs. As AMLs are hormone sensitive (with ~25% of them displaying progesterone and oestrogen receptors), they are therefore more likely to increase in size during pregnancy and rupture.⁴ For that reason, many urologists favour close observation of women with known AMLs that are of childbearing age.

A literature review has shown that most reported cases during pregnancy had ruptured (80%).⁵

Management of this condition is case dependent as there is a paucity of evidence in the literature to support a definite management pathway in pregnancy. Treatment should be tailored depending on the patient's haemodynamic stability, the size of the tumour and whether they are symptomatic or not.

Learning points

- ▶ The mode of delivery should be dictated by obstetrics indications and not solely by the presence of an AML.
- ▶ Vaginal delivery is safe if there is no obstetric contraindication.
- ▶ Preservation of the kidney is possible if the patient is haemodynamically stable or asymptomatic.
- ▶ It is important to consider renal AML as a differential for a renal mass during pregnancy, as their complications can be catastrophic.
- ▶ In an asymptomatic pregnant patient, a conservative approach, including active surveillance can often be adopted.

In an asymptomatic pregnant patient, a conservative approach, including active surveillance can often be adopted, but close monitoring by urology is recommended.²

Contributors DK—author/obstetrics team—involved in patient’s care. MA—second author/obstetrics team—involved in patient’s care. ATL—third author and urologist—involved in patient care. ME—consultant obstetrician—involved in patient’s care.

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