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Surgical management of haemorrhaging renal angiomyolipoma in pregnancy

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ABSTRACT

INTRODUCTION: Renal angiomyolipoma (AML) is a benign mesenchymal tumour of the kidney with a tendency of aneurysm formation at risk of rupturing. Due to increased maternal circulation and hormonal influences, rupture risk is greater in pregnancy, often leading to a vascular emergency and premature delivery or termination.
PRESENTATION OF CASE: A 24-weeks pregnant woman (45 years old, G6P1) presented with haematuria and flank pain. CT showed AML with acute haemorrhage. The patient became haemodynamically unstable and underwent urgent embolisation and follow-on total radical nephrectomy with the foetus being left in-utero. This involved a multidisciplinary team (urologist, vascular surgeon, interventional radiologist and obstetrician). The procedure was uncomplicated and the pregnancy went to term with a healthy girl delivered at 38 weeks.
DISCUSSION: The incidence of AML is 0.13% in the general population. 21 reports of haemorrhaging AML in pregnancy have been published in the last 35 years. Mean gestational age was 29.6 weeks. Eight were treated conservatively to term, one underwent exploratory laparotomy with evacuation of haematoma only, five were embolised, and seven were managed with nephrectomy. Of the nephrectomy subgroup, one was preceded by vaginal delivery and five underwent concurrent caesarean section (one with pre-op embolisation). There were two associated foetal deaths.
CONCLUSION: This case demonstrates that with a multidisciplinary approach, it is possible to successfully leave a foetus undelivered whilst performing a radical nephrectomy for a large bleeding AML in a woman carrying a late second trimester pregnancy.

1. Presentation of case:

A 45 year-old female in her 24th week of pregnancy (G6P1) presented to a women's specialty hospital with right-sided flank pain and haematuria. Her medical history consisted of four first trimester miscarriages, an emergency caesarean section, non-insulin dependent diabetes mellitus and iron deficiency anaemia.

Abdominal examination revealed a ballotable right upper quadrant mass. Ultrasound and computed tomography (CT) (Figs. 1 and 2) demonstrated a large renal angiomyolipoma (AML) (15 × 14 × 13 cm) with a 2.3 cm intra-capsular false aneurysm and surrounding haematoma. No other hallmark signs were found to suggest Tuberous Sclerosis.

Multidisciplinary management was initiated involving Obstetricians, Urologists, Vascular Surgeons and Interventional Radiologists. As the patients' condition had stabilised and given the substantial risks associated with premature birth before 26 weeks, consensus was for conservative management until 28 weeks of pregnancy at which time an elective concurrent caesarean section and nephrectomy would be performed. The patient was managed as an inpatient to allow for continued monitoring. Antenatal corticosteroids were administered.

Ten days following the herald bleed, the patient developed new onset frank haematuria and crescendo flank pain. She became increasingly tachycardic and her haemoglobin decreased from 110 g/L to 90 g/L (reference range 115–150), prompting blood transfusion. Foetal observations were unchanged. The patients' haemodynamic instability, likely due to rupture of the intra-parenchymal false aneurysm, drove a decision to proceed with emergency surgery.

Due to the vascular nature of the tumour, an effort was made to minimise the risk of catastrophic bleeding through urgent pre-

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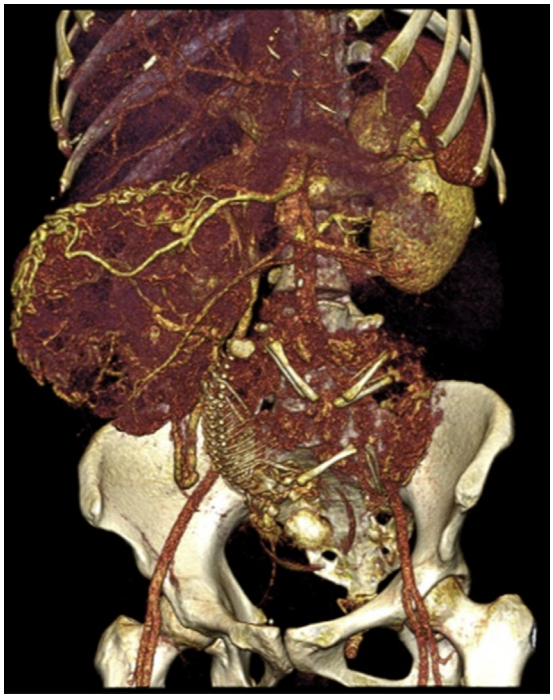

Fig. 1. 3D reconstruction showing relationship of AML and foetus.

operative angio-embolisation. To curtail radiation exposure no screening over the groin or pelvis was performed, the field of view was tightly coned to the right renal artery and injections minimised. Using endovascular plugs the interventional radiology team occluded the proximal right renal artery (Fig. 2) before direct transfer to the operating theatre. A right-sided radical nephrectomy was performed through a chevron incision. Severe haemorrhage remained a risk given the need to mobilise a large, friable vascular mass in a confined operating space restricted by a gravid uterus. Therefore the approach was to minimise mobilisation of the tumour until the inferior vena cava (IVC) was side clamped and the renal vein ligated and divided from the IVC. Gerota's fascia was left intact. Given a gestational age of only 25 weeks, the foetus was not delivered and no intra-operative foetal monitoring was performed. An obstetrician nonetheless remained on standby in case of an unanticipated emergency. Estimated intra-operative blood loss was 400 mL.

The procedure was uncomplicated and the patient made an uneventful recovery. Histopathology confirmed AML with a benign deposit of tumour in a local lymph node (Fig. 3). She was discharged on day nine post-procedure and a healthy girl was carried to term, delivered at 38 weeks by an elective caesarean section.

2. Literature review and discussion

AML is the most common benign mesenchymal tumour of the kidney, and consists of smooth muscle-, adipose- and vascular

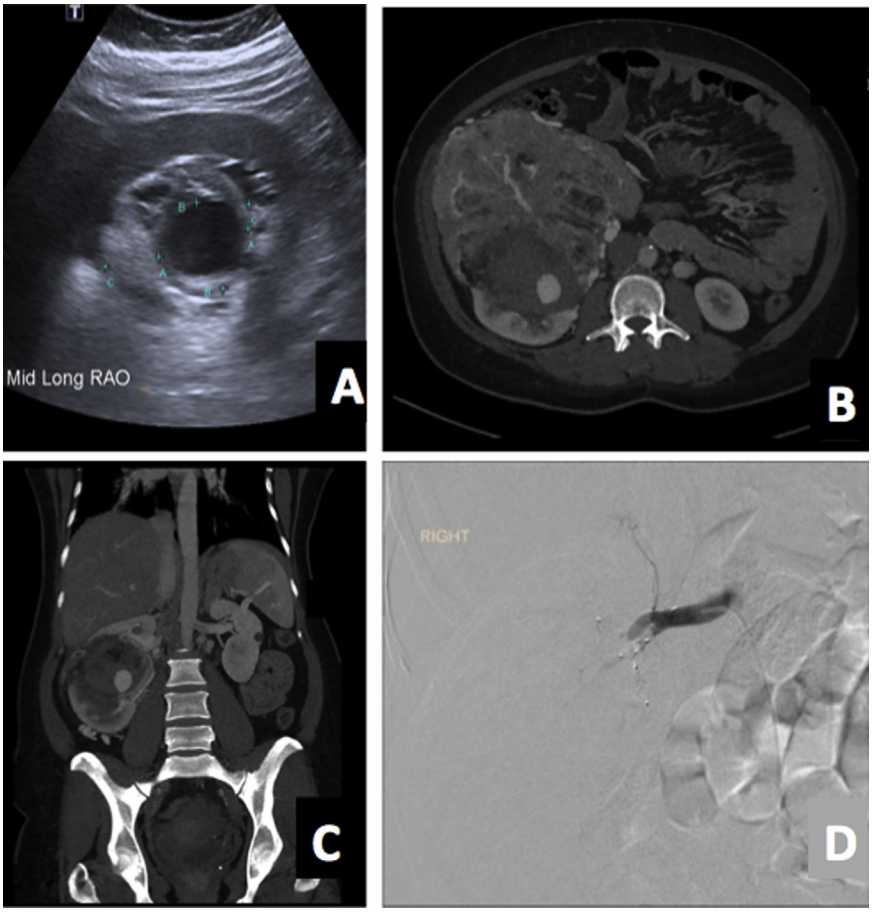

Fig. 2. (A) Ultrasound: 3.8 × 3.3cm false aneurysm in AML with surrounding hyperechoic haematoma. (B) Axial CTA: Classic CT findings of right AML, a heterogenous mass with apredominence of fat (−20 HU) and interspersed tissue density (muscular andvascular elements). 4.5% of AMLs are a 'minimal fat' subtype which impedes the reliability of CT diagnosis [7]. Large false aneurysm arrowed. (C) Coronal CTA: Haemorrhage and largefalse aneurysm within right AML, incidental left AML. (D) Digital Subtraction Angiogram: Embolised proximal right renal artery.