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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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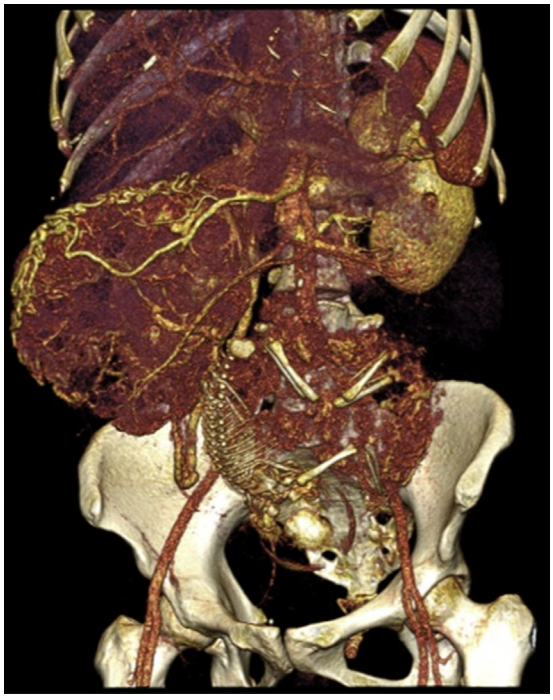

Fig. 1. 3D reconstruction showing relationship of AML and foetus.

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

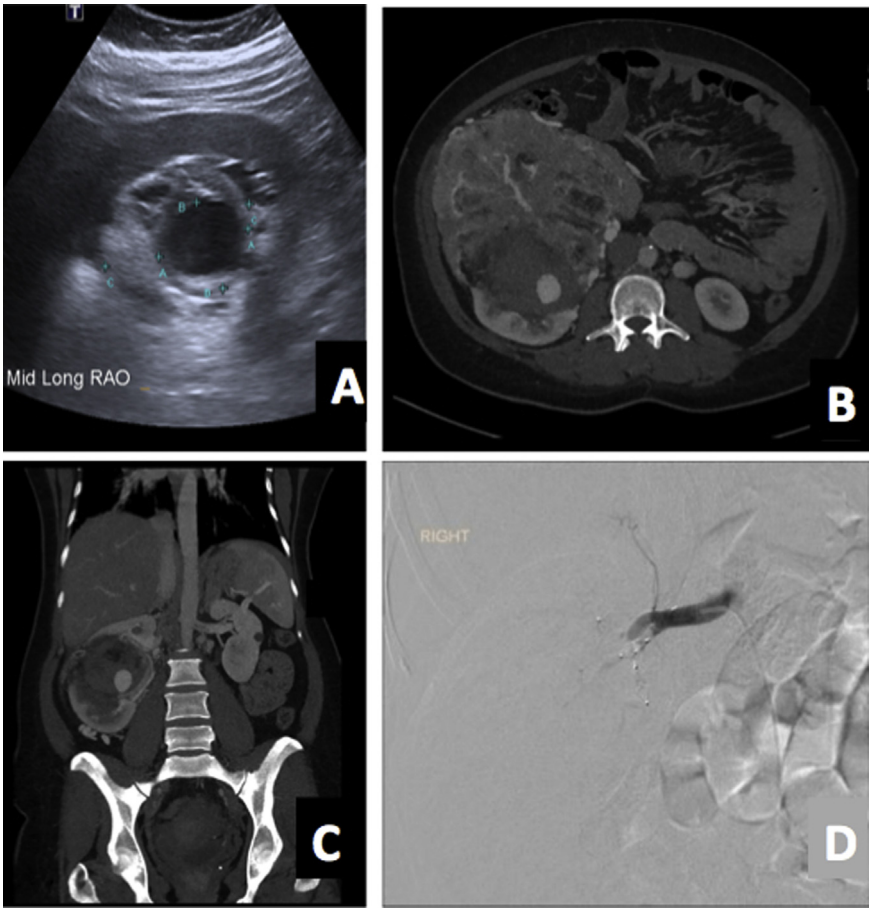

Fig. 2. (A) Ultrasound: 3.8 × 3.3cm false aneurysm in AML with surrounding hyperechoic haematoma. (B) Axial CTA: Classic CT findings of right AML, a heterogenous mass with apredominence of fat (−20 HU) and interspersed tissue density (muscular andvascular elements). 4.5% of AMLs are a 'minimal fat' subtype 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.

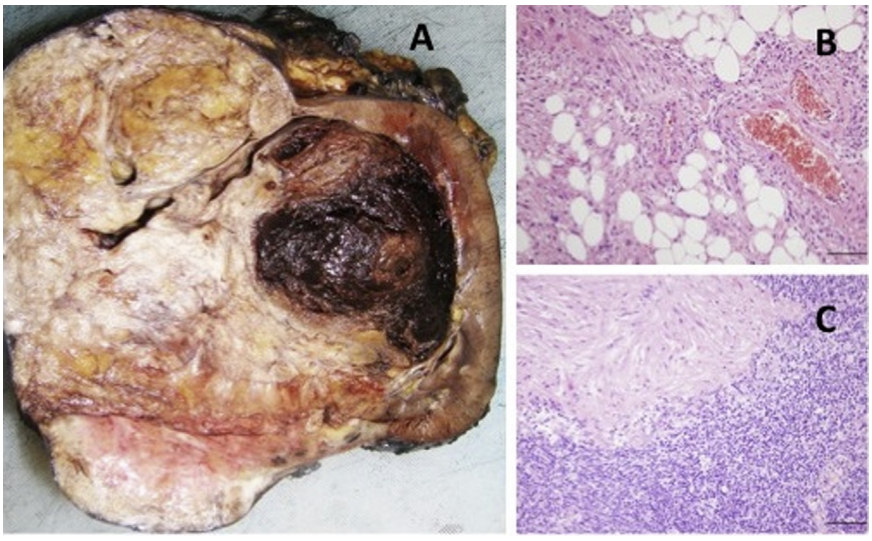

Fig. 3. Scale bars = 200 μm A. Macro: Circumscribed mass arising from kidney with focal haemorrhage. B. H and E stain: Blood vessels, spindle cells and mature adipocytes. C. AML deposit in a lymph node.

Table 1
Literature review of bleeding AML in pregnancy.

	First author	Journals	Years	Maternal age	Gestation	Foetal distress	Shock	Treatment	Pregnancy
1	Gallagher	*Obstet. Gynecol.*	1978	25	40	Yes	Yes	N	CS, FD
2	Atalla	*J. R. Army Med. Corps*	1987	39	40	No	Yes	N	VD
3	Lee	*J. Reprod. Med.*	1994	29	27	Yes	Yes	N	CS, FD
4	Yanai	*Urol. Int.*	1996	31	29	No	No	C	CT (CS)
5	Oka	*Hinyokika Kiyo*	1999	32	36	No	No	N	CS
6	Khaitan	*Urol. Int.*	2001	25	37	.	.	E	CT (VD)
7	Tanaka	*Obstet. Gynecol.*	2001	23	27	No	No	C	CT (VD)
8	Morales	*Cardiovasc. Intervent. Radiol.*	2005	28	10	No	No	E	CT (VD)
9	Storm	*Obstet. Gynecol.*	2006	32	39	No	No	C	VD
10	Gimeno Argente	*Actas Urol Esp*	2006	40	33	Yes	Yes	N	CS
11	Raft	*Gynecol. Obstet. Fertil.*	2006	40	34	Yes	Yes	L	CS
12	Al-Ateeqi	*Int. Urol. Nephrol.*	2007	31	32	No	Yes	C	CS
13	Koh	*J. Reprod. Med.*	2007	31	12	No	No	C	CS
14	Nicola	*Arch Ital Urol Androl*	2007	37	15	No	Yes	N	CT
15	Kontos	*Cases J.*	2008	28	33	Yes	Yes	N	CS
16	Binkowska	*Ginekol. Pol.*	2009	26	20	No	No	E	CT (CS)
17	Illescas Molina	*Ginecol. Obstet. Mex.*	2009	36	28	No	No	C	CT (CS)
18	Komeya	*Hinyokika Kiyo*	2010	39	38	.	.	E	CS
19	Gyimadu	*J. Obstet. Gynaecol. Res.*	2011	21	25	No	No	C	CT (CS)
20	Zapardiel	*Gynecol. Obstet. Invest.*	2011	30	35	Yes	No	E	CS
21	Iruloh	*J. Obstet. Gynaecol.*	2013	23	31	No	No	C	CT (CS)
22	Preece	*Int. J. Surg. Case Rep.*	2014	45	25	No	Yes	N	CT (CS)

AML treatment N = hrectomy, E = embolisation, C = conservative, L = exploratory laparotomy without nephrectomy pregnancy management: CS = caeserean section, VD = vaginal delivery, CT = Controlled to term, FD = associated foetal death.

tissue in varying amounts depending on the subtype [1]. The prevalence of AML in the general population approaches 0.13%, with a female to male gender ratio of 4.5:1 [2]. AMLs occur in two distinct ways, either sporadically or in association with Tuberous Sclerosis (TS). Approximately 50% of people with TS have AMLs [3], and in this cohort the tumours are usually small, multiple and bilateral. Although rare, AMLs can result in Wunderlichs syndrome [4]; the spontaneous rupture of renal parenchyma with retroperitoneal haemorrhage. The classically described triad associated with this is a palpable mass, flank pain and haemorrhagic shock.

AMLs have a greater tendency to increase in size and rupture during pregnancy. It is suggested that this higher risk is due to ubiquitous expression of oestrogen and progesterone receptors in AMLs [5], an increased maternal circulation and potentially the greater intra-abdominal pressures associated with pregnancy.

Using the Medline database, a review of the literature was conducted exploring management and outcomes of ruptured AML in pregnancy (Table 1) No limit was imposed on dates and keywords included 'angiomyolipoma; pregnancy; haemorrhage and rupture.' Articles lacking abstracts were excluded. 21 case-reports were identified over a 35-year period: 16 in English; two in Japanese; two in Spanish and one in French. The mean age of the mother was 30.8 years with a mean gestation of 29.6 weeks. Only 14.3% (3) of patients were known to have Tuberous Sclerosis and just one patient had a tumour size less than four centimetres in diameter. 76.2% (16) of patients had pain as a presenting symptom; 19% (4) had frank haematuria; a further 14.3% (3) had microscopic haematuria and 42.1% (8) were in shock. Just 9.5% (2) of patients presented with a palpable mass. Throughout the pregnancy; eight patients were treated conservatively; five were managed with angio-embolisation; one had an exploratory laparotomy with evacuation of haematoma only and seven underwent nephrectomy (one with pre-operative embolisation). In the nephrectomy subgroup; one woman underwent a vaginal delivery before puerperal rupture of an AML; whilst the majority of patients (five) underwent concurrent caesarean section with two associated foetal deaths.

Only one other case; (Nicola et al., 2007) [6] was identified in which the foetus was left undelivered and carried to term. This involved a 15-week pregnancy and an AML measuring 7.5 × 10 cm. In comparison; our case concerned a 25-week pregnant woman with a 13 × 13 × 12.5 cm AML. This case demonstrates that it is possible to successfully perform a radical nephrectomy for a large bleeding AML at a more advanced gestational age than previously known. This was potentially due to good haemorrhage control with pre-operative arterial embolisation and minimal handling of the tumour until the renal vein was ligated and divided from the IVC.

Bleeding AML in pregnancy is a rare and complex vascular surgical emergency and should be managed in a multidisciplinary team. Management is largely influenced by the haemodynamic stability of the mother, gestational age, presence or absence of foetal distress and the ready availability of facilities such as an angiography suite, operating theatre and neonatal intensive care. For haemodynamically stable patients, with no evidence of ongoing haemorrhage or signs of foetal distress, conservative management can be safely performed. However, the patient should remain in a monitored environment with a view to proceeding to more definitive management should circumstances require it. For patients in shock, or in those with ongoing haemorrhage, definitive treatment is required by embolisation and/or nephrectomy. Suitability of tumour embolisation, local expertise and resources should determine the treatment technique, however both treatments have shown favourable results. Foetal distress or a mature gestation may make nephrectomy more favourable as it allows for concurrent delivery.

In our case of a haemodynamically unstable patient with a premature foetus not in distress, a multidisciplinary approach resulted in successful nephrectomy without concurrent delivery, allowing the pregnancy to proceed to term.

Conflict of interest

All authors have no conflict of interest to declare.

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Ethical approval

The patient who is the subject of this case report gave consent to write up and publish her story. All personal information has been de-identified. The ethical principles of the Helinski Declaration were adhered to.

Author contribution

Preece – Primary author, literature review.
Mees – Vascular surgeon, second author, chief editor.
Norris – Urology trainee, literature review.
Christie – Pathologist, graphics + expert comment.
Wagner – Vascular surgeon, editor.
Dundee – Urologist, supervising author, editor.

Guarantor

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Consent

Written informed consent was obtained from the patient for publication of this case report.

Key learning points

- Ruptured AML is a rare occurrence, although the incidence is higher in pregnancy.
- Management of this vascular emergency is complex and involves consideration of mother, foetus, the available resources and local expertise.
- Previous case reports have shown that conservative management is safe in stable patients, whilst angio-embolisation or nephrectomy with concurrent caesarean section have been performed successfully in women with ongoing bleeding.
- This case report shows that it is also possible to perform a radical nephrectomy with pre-operative angio-embolisation in a 25-week pregnant woman with a haemorrhaging AML, leaving the foetus undelivered and allowing it to reach term in-utero.
- A multidisciplinary surgical team is appropriate to manage these patients.

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