

Renal angiomyolipoma in pregnancy: surgical management with fetal preservation - Approach in a developing setting

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Abstract

Renal angiomyolipomas (RAML) are uncommon benign renal tumours that are associated with a tendency to rupture resulting in sometimes-tormenting retroperitoneal hemorrhage as the Wunderlich syndrome or as severe potentially exsanguinating hematuria. When hemorrhage from RAML occurs in pregnancy it presents a unique challenge requiring timely and appropriately adapted intervention with the goal of preventing fatality, preserving renal function as well as preventing fetal loss if possible. We report the management of severe bleeding from RAML in pregnancy and highlight the need to adopt a management strategy that suits the practice environment and offers the patient standard and enduring care.

Introduction

Renal angiomyolipomas (RAML) are uncommon benign renal tumours,¹ composed of fat cells, smooth muscle cells and blood vessels.² They are associated with a tendency to rupture resulting in sometimes-tormenting retroperitoneal hemorrhage as the Wunderlich syndrome³ or as severe potentially exsanguinating hematuria.⁴ These lesions may be sporadic in about 80% of cases or be associated with genetic disorders such as the tuberous sclerosis complex.^{5,6} When hemorrhage from RAML occurs in pregnancy it presents a unique challenge requiring timely and systematic holistic definitive intervention with the goal of preventing fatality, preserving renal function as well as preventing fetal loss if possible.

Currently a gamut of management options are available including arterial embolization, radiofrequency ablation, cryoablation, mammalian target of rapamycin (mTOR) inhibitors; everolimus and partial or total nephrectomy.¹ We report the management of severe bleeding from RAML in pregnancy and highlight the need to adopt a management strategy that suits the practice environment and offers the patient standard and enduring care.

Ethics

The patient gave her written informed consent for publication of this report.

Case Report

A 26-year old trader, in her 16th week of pregnancy (Gravida 2 Para 1), presented on account of right flank pain and swelling with associated haematuria (often in clots), weakness, vomiting and anorexia.

Physical examination revealed severe pallor, pulse; 118 beats per min, blood pressure; 90/60 mmHg respiratory rate; 28/min, and a firm, tender right lumbar mass extending to the right upper quadrant. Blood clots were noted at the urethral meatus.

Investigations revealed a PCV of 18%. She was admitted and resuscitated with blood transfusions, analgesics and bladder washouts via a 24G 3 way Foley catheter. When normotensive, tocolysis was commenced with oral Nifedipine 20 mg with hourly monitoring of blood pressure. Subsequent doses were omitted if pre-dose readings were \leq 120/80 mmHg.

Abdominal ultrasound findings showed a pulsatile well delineable oblong hypoechoic fluid mass about 77.8x62 mm in size with its medial compartment in the mid pole. The fluid within the mass had a swirling character and on Doppler insonation, this fluid had characteristics consistent with AV fistulous aneurysmatic lesion or AV malformation. Urinary bladder harboured large blood clots. Liver harboured two hypoechoic solid focal lesions in the right lobe with features of haemangioma. These lesions measure about 1.9 cm to 2.5 cm uterus. There is a viable 14.5 weeks old intrauterine singleton cyesis.

Magnetic resonance imaging (MRI) showed a large well defined complex mixed intensity mass involving the anterior and inferior aspects of the right kidney, 15x12 cm in size and causes caliectasis of the supero-posterior remnant part of the kidney. Overall features suggested renal mass with areas of haemorrhage of varying ages. The liver harboured poorly circumscribed multiple masses, which are hyperintense on t1 and t2 and post contrast. They may represent hemangioma. Left

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kidney and spleen appear normal. No retroperitoneal adenopathy. No ascites (Figure 1). Large complex right renal mass with haematoma of varying ages. Hyperintense hepatic masses (in t1 and t2) suggestive of hemangioma.

Counseling was done, where the diagnosis and available treatment options (surgical extirpation after stabilization) were explained. She was also advised on the risk of perioperative fetal loss as well as the need for long term follow up for the clinically silent hepatic lesions. A right radical nephrectomy was done 11 days after admission. Findings were those of a perirenal haematoma and large hemorrhagic renal mass involving the lower and mid poles and compressing the upper pole; histopathology revealed renal angiomyolipoma (Figures 2 and 3).

Patient did well post operatively and was discharged home 1 week later. She has remained well on follow up 1 year after.

Discussion

Angiomyolipoma (AML) is a benign mesenchymal tumor composed of variable proportions of adipose tissue, spindle and epithelioid smooth muscle cells and abnormal thick-walled blood vessels; with the two organs most commonly involved being the kidney (77%)

and liver (14%).⁷ AMLs occur in two distinct ways, either sporadically or in association with tuberous Sclerosis (TS), and in this cohort the tumours are usually small, multiple and bilateral. Although rare, AMLs can result in Wunderlich's syndrome and or frank hematuria.^{3,4} The prevalence of sporadic renal angiomyolipoma is 0.44%, with 0.60% in the female and 0.28% in the male subpopulations.⁸ AMLs have a greater tendency to increase in size and rupture during pregnancy.^{4,9} Though the causal mechanism is not clearly defined,⁹ it is thought that this is due to ubiquitous expression of oestrogen and progesterone receptors in AMLs.¹⁰ Haemorrhage occurring in RAML in pregnancy has been shown to be rare; with only 22 cases reported in the last 35 years.⁴ The mean age of the mothers was 30.8 years with a mean gestation of 29.6 weeks.⁴ Our patient was 26 years old and had a 14.5-week gestation. Fetal survival till term in the patients treated by nephrectomy is also rare, to our knowledge, only reported in a 15 week pregnant patient with a RAML of 7.5x10 cm.^{4,10}

Our patient who was 14.5 weeks pregnant, though with a larger tumor 15x12 cm (MRI) had a similar outcome; she had caesarian section at 40 weeks with a healthy baby.

Concerning treatment options ranging from minimally invasive options such as embolization, radiofrequency ablation, cryoablation, novel therapies like everolimus to partial or total nephrectomy.¹ The choice of these are influenced by availability and expertise and surgeons' judgement. Earlier workers have shown that prevalence of major bleeding is high in sporadic AMLs with a diameter of >6 cm.⁶ and our patient demonstrated several episodes of haemodynamic instability. Bleeding AML in pregnancy is a rare and complex vascular surgical emergency and should be managed in a multidisciplinary team.⁴ 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.⁴ Challenges experienced in her management include the absence of facilities for angiography and embolization and absence of in-house facilities for MRI. This required careful consideration as to when patient could be moved to get the imaging done.

Conclusions

We demonstrate that it is possible to manage severe bleeding from RAML by resuscitation and well-timed radical nephrectomy at 14 weeks gestation with excellent maternal and fetal outcome.

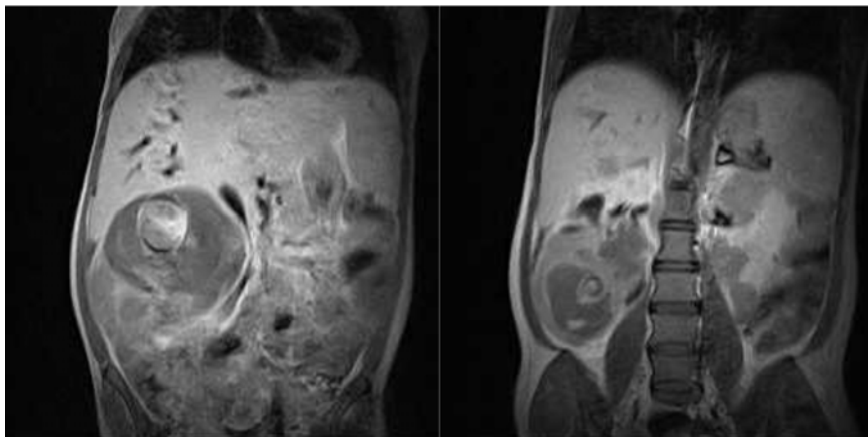


Figure 1. Magnetic resonance imaging abdomen showing the complex right renal mass and superior caliectasis.



Figure 2. Gross specimen showed a renal mass with areas of circumscribed hemorrhage compressing the remnant kidney upwards.

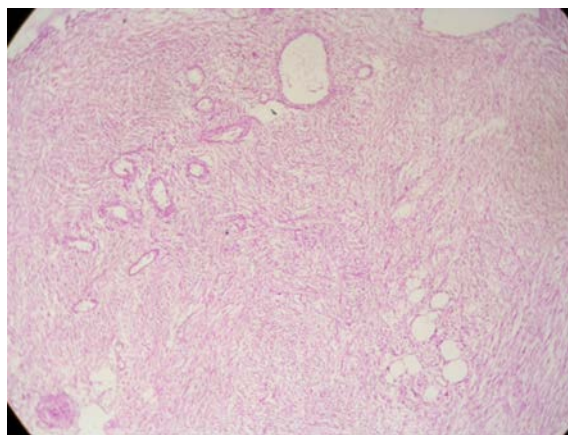


Figure 3. Photomicrograph of the lesion (hematoxylin and eosin) showing thick walled blood vessels and smooth muscles.

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