CASE REPORT

Spontaneous rupture of renal angiomyolipoma during pregnancy

Li Ao, Eriko Ogasahara, Yasuhiko Okuda, Shuji Hirata

Department of Obstetrics and Gynecology, University of Yamanashi, Chuo, Yamanashi, Japan

Correspondence to

Dr Li Ao,
aoli@yamanashi.ac.jp

Accepted 18 December 2016

SUMMARY

A renal angiomyolipoma (AML) is a rare benign tumour of kidney origin. Pregnancy is known to be associated with an increased risk of tumour rupture causing hypovolaemic shock, which is usually managed surgically or through an embolisation procedure. However, having surgery during pregnancy predisposes the mother to a preterm delivery, and the unknown influences of radiation exposure to the fetus make the management of such cases very challenging. A 30-year-old pregnant woman had a sudden onset of gross haematuria at the 20th week of her pregnancy. The MRI showed a 10 cm mass suggestive of AML in the left kidney, with evidence of an intrarenal haematoma. To avoid an iatrogenic preterm delivery and unnecessary fetal exposure to radiation, conservative management was conducted until 34 weeks of gestation, when she came to our hospital reporting of flank pain. An endovascular treatment was performed immediately after an emergency caesarean delivery.

BACKGROUND

The incidence of angiomyolipomas (AMLs) in the general population is 0.4%.¹ AMLs occur in two distinct ways, either sporadically or in association with tuberous sclerosis complex (TSC).² Rupture of a renal AML during pregnancy may jeopardise the mother and fetus. Management is largely influenced by the haemodynamic stability of the mother, gestational age, presence or absence of fetal distress and the immediate availability of facilities.

CASE PRESENTATION

A 30-year-old primipara presented to the emergency department of a secondary care centre at 20 weeks of gestation and reported haematuria, without abdominal/flank pain. There was no history of trauma, surgical intervention or any other medical comorbidity. She did not have per vaginal bleeding, and her pregnancy had been uncomplicated until her presentation. On presentation, she was afebrile and haemodynamically stable.

INVESTIGATIONS

Routine blood investigations showed a haemoglobin (Hb) level of 9.7 g/dL, a white cell count (WCC) of 10930/L and a C reactive protein (CRP) level of 0.3 mg/L. Her serum urea and creatinine levels were within normal limits. Transvaginal ultrasonography and pelvic ultrasonography were also normal. The abdominal MRI scan revealed a giant mass suggestive of AML originating from the left

kidney with evidence of intrarenal haematoma (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included urolithiasis, placenta percreta and uterine rupture. In addition, renal cell carcinomas and oncocytomas were also included in the differential diagnoses based on the MRI findings.

TREATMENT

Given the size of the tumour and presence of symptoms, she was admitted into our hospital. Multidisciplinary management was initiated involving obstetricians, urologists and interventional radiologists. As the patient's condition had stabilised, the consensus elected was for conservative management. The patient was managed as an outpatient with follow-up duplex renal ultrasonography every 6 weeks. The pregnancy proceeded with no other associated symptoms until 34 weeks of the gestation, at which time the patient was admitted to our hospital for severe left flank pain. Physical examination revealed a good general condition, an axillary temperature of 36.9°C, a blood pressure of 127/85 mm Hg and a heart rate of 92 bpm. Owing to the regularity of the pain, which was frequent and progressively worsening, and a breech presentation, endovascular treatment after emergency caesarean delivery was performed. Abdominal aortography demonstrated contrast extravasation originating from the left segmental renal artery. Subsequent selective left renal artery injections confirmed the diagnosis of two pseudoaneurysms arising from the feeding artery of the AML. Using a microcatheter, the feeding artery was embolised with diluted

Figure 1 An axial T2-weighted fat-saturated MR image showing a bulky heterogeneous lesion in the left kidney and completely enclosed by the renal capsule.

CrossMark

To cite: Ao L, Ogasahara E, Okuda Y, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-217284

Figure 2 A sagittal MR T2 image showing a 10 mm AML above the main lesion. AML, angiomyolipoma.

Figure 3 Before transarterial embolisation of aneurysms.

n-butyl cyanoacrylate (nBCA). Following this procedure, a control angiography documented no further filling of the pseudoaneurysm sac and showed the renal parenchyma was preserved (figures 3 and 4).

OUTCOME AND FOLLOW-UP

A healthy girl child weighing 1990 g was delivered. She had Apgar scores of 8 and 9 at 1 and 5 min, respectively. The patient was discharged in good condition 5 days later and underwent follow-up duplex renal ultrasonography at 6 weeks, which demonstrated that the size of the left renal AML had decreased from 10 cm to 9 cm. She will be evaluated by MRI in 6 months to determine the success of embolisation.

DISCUSSION

AML with spontaneous bleeding during pregnancy is a rare condition. Treatment options for AML include total or partial nephrectomy and trans-catheter selective arterial embolisation (TAE). Twenty-five reports of haemorrhaging AML in pregnancy

Figure 4 After the transarterial embolisation.

have been published in the last 30 years. Eight were treated conservatively to term, nine were managed with angioembolisation and six underwent nephrectomy.^{3–8} In this case, due to the haemodynamic stability and spontaneous cessation of haematuria at the 20th week of the pregnancy, it was decided that an expectant procedure would be performed. Approximately 3 months following the herald bleed, the patient developed a new onset of crescendo flank pain. In contrast to surgery, TAE is minimally invasive without the need for general anaesthesia or open surgery.⁹

To avoid fetal exposure to radiation, endovascular treatment was conducted immediately following the emergency caesarean delivery.

In conclusion, it appears that AML shows a greater growth index in pregnant women, and the issue in question is when is the appropriate time for treatment. Since a consensus has not yet been determined on this issue, individualisation for each case is necessary, even over time for the same patient.

Learning points

- ▶ Renal angiomyolipoma (AML) is a rare benign tumour that can expand rapidly during pregnancy due to increased hormone levels.
- ▶ When a pregnant woman presents with haematuria, obstetric causes such as placenta praevia, abruption placenta and uterine rupture should be ruled out. In addition, urolithiasis, oncocytomas, renal AML and cell carcinomas should also be included in the differential diagnoses.
- ▶ A multidisciplinary team approach has shown successful treatment for renal AML during pregnancy.
- ▶ Since a consensus has not been reached on when is the best time for treatment and what is the optimal treatment method for haemorrhaging AML during pregnancy, the individualisation of each case is very important.

Contributors LA contributed to conception and design, acquisition of the data and analysis; drafted the article and provided final approval. EO and YO contributed to analysis and interpretation of the data; revised it critically for important intellectual content and provided final approval. SH contributed to concept and design and analysis and interpretation of the data; drafted the manuscript; revised it critically for important intellectual content and provided final approval.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Fittschen A, Wendlik I, Oeztuerk S, *et al.* Prevalence of sporadic renal angiomyolipoma: a retrospective analysis of 61,389 in- and out-patients. *Abdom Imaging* 2014;39:1009–13.
- 2 Cook JA, Oliver K, Mueller RF, *et al.* A cross sectional study of renal involvement in tuberous sclerosis. *J Med Genet* 1996;33:480–4.
- 3 Preece P, Mees B, Norris B, *et al.* Surgical management of haemorrhaging renal angiomyolipoma in pregnancy. *Int J Surg Case Rep* 2015;7c:89–92.
- 4 Myoen S, Mitsuzuka K, Saito H, *et al.* Spontaneous rupture of a renal angiomyolipoma at 25 weeks of pregnancy treated with transarterial embolization: a case report and review of the literature. *Int J Urol* 2015;22:710–12.
- 5 dos Santos MM, Proenca SM, Reis MI, *et al.* Spontaneous rupture of renal angiomyolipoma during pregnancy. *Rev Bras Ginecol Obstet* 2014;36:377–80.
- 6 Pontis A, Piras B, Meloni A, *et al.* Rupture of renal angiomyolipoma in pregnancy. *J Obstet Gynaecol* 2013;33:628–9.
- 7 Ferianec V, Gabor M, Cano M, *et al.* Severe retroperitoneal haemorrhage in the first trimester of a multiple pregnancy after spontaneous rupture of renal angiomyolipoma. *Arch Gynecol Obstet* 2013;288:1193–4.
- 8 Shah J, Jones J, Miller MA, *et al.* Selective embolization of bleeding renal angiomyolipoma in pregnancy. *J R Soc Med* 1999;92:414–15.
- 9 Ewalt DH, Diamond N, Rees C, *et al.* Long-term outcome of transcatheter embolization of renal angiomyolipomas due to tuberous sclerosis complex. *J Urol* 2005;174:1764–6.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow