Case Report

Renal Angiomyolipoma During Pregnancy: What Can We Offer?

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Abstract

Renal angiomyolipoma is a rare disease seen during pregnancy. Rupture of renal angiomyolipoma could be catastrophic and might result in maternal and even perinatal mortality. Management includes conservative vs surgical approach. A 29-year-old woman Para 2 with history of bleeding renal angiomyolipoma in her first pregnancy at 11 weeks treated with selective arterial embolisation. The pregnancy was terminated. Even though having small residual tumour, her subsequent pregnancy progressed well with conservative management. Intervention is advisable in the presence of large or symptomatic renal angiomyolipoma prior to pregnancy in order to minimise potential life-threatening haemorrhage during pregnancy. For those with small tumour of less than 4cm, perhaps conservative approaches i.e. frequent follow-up and close monitoring would assist in early identification of any rupture or bleeding.

Keywords: Renal angiomyolipoma, embolisation, haemorrhage, nephrectomy, pregnancy

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Introduction

Renal angiomyolipoma (AML) is a relatively rare clonal neoplasm. It composes of mature adipose tissue, smooth muscle and thicken-wall blood vessels with chromosomal imbalances (1). The reported incidence was around 0.13% and was even less commonly seen during pregnancy (2). Most renal AML is slow growing tumour but pregnancy could increase its size leading to a higher risk of rupture and haemorrhage (3). Majorities were asymptomatic (75.2%), but some patients may present with flank pain, gross haematuria or fatigue (4). Nearly one quarter of cases were diagnosed before pregnancy (3). To date, not many cases have been reported (3). We report a case of bleeding renal AML in early pregnancy treated with embolisation. The pregnancy was terminated. She had a successful pregnancy despite presence of small residual tumour.

Case Report

A 29-year-old lady presented in her first pregnancy at 11 weeks gestation to a private centre complaining of left loin pain. There was no significant medical history or family history. She was diagnosed to have bilateral renal AML. A few days later, her loin pain worsened and she had tachycardia. Bleeding renal AML was suspected as her haemoglobin dropped from 13.3g/dl to 10.5g/dl. Magnetic Resonance Imaging (MRI) was done which confirmed haemorrhage from left renal AML. A total of three tumours were seen at her left kidney (11.5cm, 5.5cm and 6.0cm) and two tumours were seen at her right kidney (5.5cm and 4.0cm).

She underwent an emergency selective arterial embolisation, which was complicated by post-embolisation syndrome. Her pregnancy was terminated. Unfortunately, the embolisation was not

successful hence she was referred to our centre. A repeat embolisation was performed using bead blocks (100-300 micron and 300-500 micron). Approximately, 90% of the tumour on the right kidney and 80% of the tumour on the left kidney were successfully embolised. The size of residual tumours ranged between 2-3cm (Fig. 1,2).

She embarked on her second pregnancy four months later. She was counseled regarding the possibility of tumour increasing in size during pregnancy and risk of haemorrhage which could be life threatening. She had regular ultrasound surveillance on the tumour size throughout her pregnancy. Though she had several episodes of admission due to loin pain, there was no evidence of haemorrhage or change of tumour size.

She had an elective caesarean section at 38 weeks for breech presentation. The caesarean section was uneventful. She remained asymptomatic and the tumour remained similar size 4 months post delivery.

Discussion

Renal AML may present as two distinct clinical entities i.e. isolated or associated with tuberous sclerosis complex (TSC). Those associated with TSC tends to present at younger age, having multiple lesions, larger in size and commonly bilateral involvement (4). The mean age of presentation is 50 years in the general population but lower in pregnancy (26 - 40 years old) (5).

The average size of rupture was 11.7cm (3). Koo et al. (4) subanalysed their data found that, patients with tumour size ≥ 4cm were younger, more commonly symptomatic, multiple lesions, bilateral, associated with TSC and more likely required intervention.

Rupture of renal AML occurred commonly during second and third trimester but it had been reported to occur as early as 10th weeks gestation (5). During pregnancy, the risk of rupture and haemorrhage is higher as the tumour size increases. This is probably due to the effect of hormonal stimulation. L'Hostis et

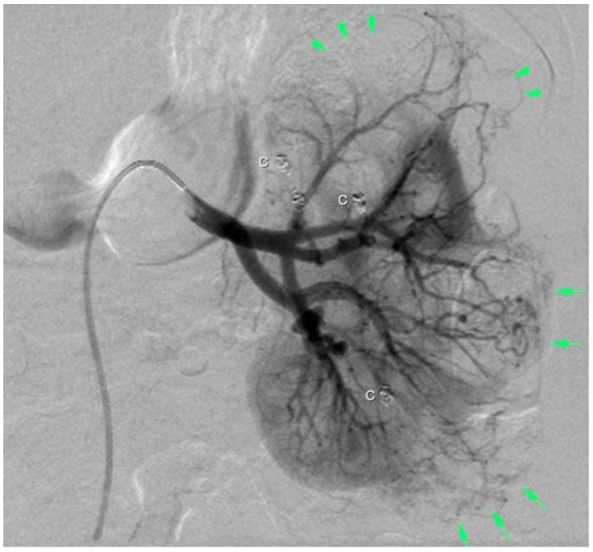

Figure 1: Pre-intervention left renal DSA image showing partial coil embolization (C) (which was performed at the former center) of the upper pole AMLs which are shown here as areas devoid of normal nephrogram (arrowheads). Residual AML lesions are still present in the lower pole (arrows).

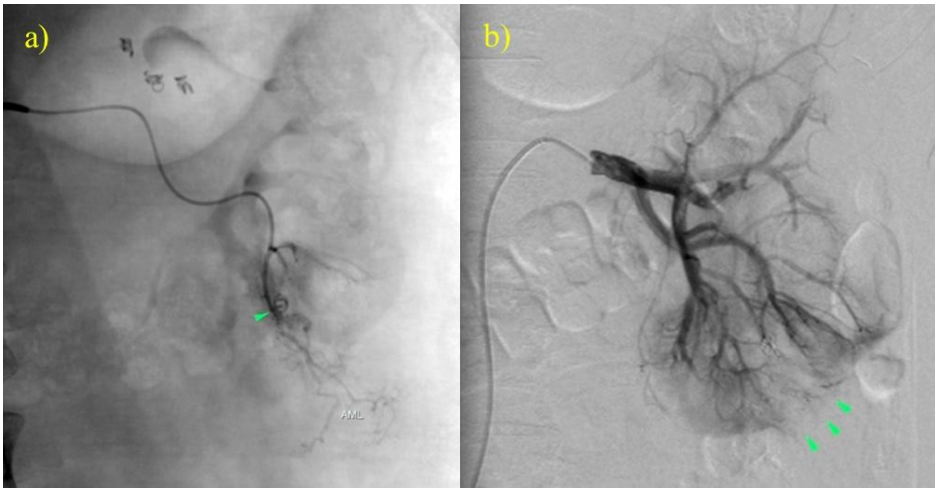

Figure 2: a) Selective embolization was performed via a microcatheter, with its tip placed distally in the tumour arterial feeders (arrowhead) b) Post-intervention angiogram image showing markedly reduced lower pole AML vascular beds. More than 90 percent of tumour vascular supply was occluded (arrowheads).

al. (6) reported that 25% of tumours showed positivity towards oestrogen and/or progesterone receptor. Others suggested that pregnancy-induced changes in urinary tract exaggerate weakness of the parenchyma or collecting system that contributed to spontaneous rupture and haemorrhage (7).

Our patient presented with haemorrhage as early as 11 weeks gestation with initial tumour size bigger than 4 cm i.e. left kidney (11cm, 5.5cm and 6.0cm) and right kidney (5.0cm and 4.0cm). Sadly, her pregnancy was terminated with the fear of radiation risk in early pregnancy. Following successful embolisation at second attempt, she had a successful pregnancy and the tumour size remained static at 2 - 3cm.

With the advancement and use of newer imaging modalities such as ultrasound, contrast enhanced Computed Tomography scan and MRI, the diagnosis of renal AML is made easier and with certainty. The detection of existing fat in a renal lesion helps to establish the diagnosis of renal AML and it is the only radiological feature that distinct it from renal cell carcinoma (8).

Fetuses usually not affected except in massive haemorrhage. Some author reported successful conservative management after rupture of renal AML (9) but others reported death of fetus (10). Treatment options for renal AML include open or laparoscopic, total or partial nephrectomy (11), radiofrequency ablation (12) or selective arterial embolisation (13,14). Conservative management should be considered as an option of treatment if the patient is asymptomatic and haemodynamically stable. There were numerous case reports showed good maternal and fetal outcome with this approach. Both successful vaginal delivery and caesarean section had been reported (5,9).

Coskuner et al. (11) reported a 26-year-old woman with giant AML being successfully treated with open partial nephrectomy. The blood loss was only 200cc. They concluded that this procedure is the main choice of treatment in young patients. Faddegon et al. (15) also reported high success rate in surgical approach where forty-two cases were successfully treated surgically. The decision for surgery was mainly due to diagnostic uncertainty or complex vascular anatomy that did not amenable to embolisation.

Selective arterial embolisation (SAE) allows rapid patient stabilisation in cases of acute massive haemorrhage and provides good renal preservation (15). Chan et al. (13) reviewed outcome of 28 cases of AML, where 15 patients had emergency SAE done for bleeding AML and 13 had it done prophylactically for asymptomatic AML sized more than 4.1cm. Ninety three percent of patients were successfully embolised in the first SAE. Renal surgery was required only in one patient. Of all variables that were being analysed, only tumour size >10cm was significantly associated with the need of subsequent renal surgery. Another retrospective study by Stoica et al. (14) reported 83% success rate at 18 months in 11 patients with retroperitoneal haemorrhage treated with emergency embolisation. One out of 9 patients in the prophylactic embolisation group required re-embolisation at 20 months due to arterial duplicity. Otherwise, there was no intra-operative complication. The author concluded that programmed embolisation effectively prevents the risk of bleeding without compromising renal function.

Conclusion

This case clearly illustrated the importance of intervention before embarking on pregnancy particularly in patients with large or symptomatic renal AML in order to avoid potential life-threatening haemorrhage during pregnancy. For those with small tumour of less than 4cm, perhaps conservative approach i.e. frequent follow up and close monitoring would help in early identification of any rupture or bleeding. Early referral to tertiary centre that equip with multi-disciplinary team management composed of interventional radiologist, nephrologist, obstetrician and neonatologist, is essential to minimize maternal and perinatal morbidity and mortality.

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