

Renal Angiomyolipoma In A Solitary Functioning Kidney Discovered Post Partum.

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Citation

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Abstract

Background:

Though rare, angiomyolipoma (AML), is the most common mesenchymal tumour of the kidney. AML is a benign tumour composed of varying amounts of mature adipose tissue, smooth muscle and thick walled blood vessels. The incidence in the general population is between 0.07% and 0.3%. . The association between renal AML and pregnancy is a rare one. The first case of renal AML associated with bleeding in pregnancy was reported in 1964. Since then, only 15 cases have been published in the literature. This is the first report in the literature of a large asymptomatic angiomyolipoma in a solitary functioning kidney found immediately post partum in a 28 year old female.

Case Presentation:

A 28 year old female was referred to the general surgeons from the labour ward 24hrs post partum with a non tender mass palpable in the right flank detected on routine examination. An ultrasound scan of the abdomen revealed a mass of mixed echogenicity involving and expanding the upper pole of the right kidney. In addition the left kidney was atrophic and non functioning (subsequent radionuclide scan). Consequently a decision to biopsy under ultrasound guidance was made. Histology confirmed a mixed mesenchymal tumour of mature fat, smooth muscle and vasculature, confirming the diagnosis of AML. The patient remained asymptomatic with no deterioration in renal function. She was then referred to a tertiary referral centre for selective embolization of the feeding vessels following which the patient made a complete recovery.

Conclusions:

The specific risk of renal AML and pregnancy is that of spontaneous haemorrhage and rupture. Although most AML are benign and asymptomatic, the literature suggests that symptoms develop in up to 80% of patients when tumour size reaches 4cm or greater. The most severe and life threatening symptom is that due to haemorrhage. Treatment options for AML include conservative and interventional (total/partial nephrectomy, cryotherapy, embolization). The current belief is that tumour size should determine whether prophylactic treatment is offered to women of child bearing age due to the possible dangerous (predominantly haemorrhagic) events that may occur in pregnancy. This case provides evidence that it may be possible to continue pregnancy and achieve a normal vaginal delivery when treated conservatively. Patients with angiomyolipoma who intend on becoming pregnant should be counselled about the risks and the option of treating these tumours prophylactically also discussed.

BACKGROUND

Though rare, angiomyolipoma (AML), is the most common mesenchymal tumour of the kidney. Initially described by Grawitz in 1900, AML is a benign tumour composed of varying amounts of mature adipose tissue, smooth muscle and thick walled blood vessels.¹ The incidence in the general population is between 0.07% and 0.3%. Clinically, two distinct presentations of renal AML are usually seen. Between 40 and 80% of patients with tuberous sclerosis have renal AML, which tend to be multiple, bilateral, asymptomatic and often less than 2cm in diameter.² However, approximately 50% of symptomatic AMLs occur

in patients who do not have tuberous sclerosis. These tumours usually are single and unilateral. The association between renal AML and pregnancy is a rare one. The specific risk of spontaneous haemorrhage and rupture poses a difficult diagnostic and therapeutic challenge. The first case of renal AML associated with bleeding in pregnancy was reported in 1964. Since then, only 15 cases have been published in the literature.^{3,4,5} There are however no cases in the literature described in the immediate post partum period. This is the first report in the literature of a large asymptomatic angiomyolipoma in a solitary functioning kidney found immediately post partum in a 28 year old

female. We review and discuss renal AML in pregnancy, along with its diagnostic and therapeutic challenges.

CASE HISTORY

A 28 year old female was referred to the general surgeons from the labour ward 24hrs post partum. The patient had successfully delivered a healthy infant following an uncomplicated spontaneous vaginal delivery at full term. She had no significant medical history and in particular no history of tuberous sclerosis. Prior to discharge, she underwent a routine clinical examination by the midwife. Palpation of the abdomen revealed a non tender fullness on the right. All other parameters including blood pressure and heart rate were within normal limits. A complete blood count revealed haemoglobin of 11.6g/dl, white blood cell count (WBC) of 12×10^9 and platelets 228×10^9 . Renal function was normal (creatinine $47 \mu \text{mol/l}$).

An ultrasound scan of the abdomen revealed a mass of mixed echogenicity involving and expanding the upper pole of the right kidney. In addition the left kidney was small measuring 5.3cm. Follow up computerised tomography of the abdomen confirmed a small atrophic left kidney and a 9cm x 9cm x 14cm mass of mixed attenuation in the upper pole of the right kidney. Subsequent DMSA scan showed only 1% uptake in the left kidney confirming suspicions of a non functioning kidney. In view of the solitary functioning kidney, a decision was made to biopsy the mass under ultrasound guidance. Histology confirmed a mixed mesenchymal tumour of mature fat, smooth muscle and vasculature, confirming the diagnosis of AML. The patient remained asymptomatic with no deterioration in renal function. She was then referred to a tertiary referral centre for selective embolization of the feeding vessels following which the patient made a complete recovery.

Figure 1

Figure 1: Computerised tomography of the abdomen demonstrating a large heterogeneous mass arising from the upper pole of the right kidney. Note small atrophic contralateral kidney.



CONCLUSION

Angiomyolipoma (AML) of the kidney is a benign, heterogeneous neoplasm composed of variable amounts of mature adipose tissue, smooth muscle and thick-walled blood vessels. They are generally regarded as choriostomas, a disordered arrangement of mature tissue occurring at a normal location.

AML is more common in women and in the sporadic form usually presents as a single tumour. Patients with tuberous sclerosis generally have multiple lesions bilaterally with a greater chance of spontaneous haemorrhage.

AML is a relatively slow growing neoplasm. However, there have been suggestions that AML may be hormonally dependant, and angiomyolipoma in pregnancy has been reported in the literature. In recent years, it has been shown that in more than 25% of cases of renal AML there is positivity for hormonal receptors (estrogen and/or progesterone). This is probably the reason why AML has a tendency to increase in size during pregnancy resulting in bleeding. In some of these cases, rapid growth during pregnancy has been documented, stimulated by the hormonal environment – in particular progesterone.

Although most AMLs are benign and asymptomatic, the literature suggests that symptoms develop in up to 80% of patients when tumour size reaches 4cm or greater. Flank pain (>50%), haematuria (>20%), and palpable mass (>10%) are the commonest symptoms. The most severe and life

threatening symptom is that due to haemorrhage which typically presents as acute pain. Indeed up to 20% are in shock on initial presentation¹¹. Thus, any pregnant woman presenting with abdominal pain and intra-abdominal bleeding, AML must be included in the differential diagnosis along with the more common obstetric causes such as placenta previa, abruption placentae and ruptured ectopic.

The diagnosis of AML is established with ultrasound (US), CT and angiography. US has a high degree of specificity, as the mass typically presents hyperechoic. The demonstration of fat on CT or magnetic resonance imaging (MRI) within a solid renal lesion is usually confirmatory of the diagnosis¹².

Treatment options for AML include conservative and interventional. The latter include open or laparoscopic, total or partial nephrectomy, cryoablation, radiofrequency ablation and transcatheter selective arterial embolization. In this particular case, the basis of treatment was to treat the AML with minimal loss of normal renal parenchyma – vitally important in the context of a single functioning kidney.

The current belief is that tumour size should determine whether prophylactic treatment is offered to women of child bearing age due to the possible dangerous (predominantly haemorrhagic) events that may occur in pregnancy¹³. There is however, as in this case, evidence that it may be possible to continue pregnancy and achieve a normal vaginal delivery when treated conservatively. Patients with angiomyolipoma who intend on becoming pregnant should be counselled about the risks, and the option of treating these tumours prophylactically discussed. If treated conservatively these tumours should be closely monitored during pregnancy.

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