A case of haemorrhagic angiomyolipoma after miscarriage

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Abstract

A case of a 27 year-old patient who presented after medical management of a missed miscarriage with an acute abdomen is described. She was found to have a haemorrhagic angiomyolipoma, which was successfully treated radiologically. This case highlights the need to consider this differential in the presentation of acute abdomen in the context of a recent or concurrent pregnancy. It is the first case report to describe this phenomenon in the context of a miscarriage.

Keywords

Acute abdomen; angiomyolipoma; pregnancy; miscarriage.

Case history

A 27-year-old woman with no previous medical history presented to the acute surgical unit with abdominal pain 5 days after being discharged following medical management of a missed miscarriage at 11 weeks gestation. She reported right upper quadrant pain and malaise. On examination, she had a palpable mass in her right upper quadrant but her abdomen was otherwise soft with no evidence of peritonism. Observations showed a septic clinical picture with swinging pyrexia, mild tachycardia and tachypnoea, while bloods on admission demonstrated raised inflammatory markers and anaemia (admission C-reactive protein 76 mg/l, rising to 200 mg/l within 6 h; white cell count 7.4 × 10⁹/L, haemoglobin 85 g/l).

The patient was started on empirical antibiotics, supportive intravenous fluids and had an abdominal ultrasound scan to investigate the right upper quadrant mass. This showed multiple, highly reflective, well-defined cortical lesions within both kidneys in keeping with multiple angiomyolipomas (AML). There was an incidental cyst at the inferior pole of the left kidney. In addition, within the right flank was a well-defined, vascular mass measuring 7.2 cm × 5.6 cm × 11.6 cm. This was seen to contain cystic areas and an anterior rim of fluid closely related to the right kidney and the lower lobe of the liver. The impression was of complex, potentially haemorrhagic AML (Fig. 1).

A computed tomography (CT) scan of her abdomen and pelvis showed bilateral renal AMLs. The vascular right upper quadrant mass was characterized as a haematoma containing a pseudoaneurysm extending from the right renal cortex to the pelvic brim. This venous phase section

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showed a dominant right-sided AML measuring 62.96 mm × 90.61 mm with a large haematoma extending from the right renal cortex to the pelvic brim. Within this, there was a focal area of vascular enhancement in keeping with a pseudoaneurysm (Fig. 2).

A diagnosis of a haemorrhagic AML was made and the patient had an emergency renal artery embolization (Fig. 3). Three days after the procedure, the patient developed pyrexia, nausea and abdominal pain. A repeat CT angiogram was subsequently performed, which showed no active bleeding and stable AMLs (Fig. 4). It was thought that the patient’s symptoms after the procedure were due to post-embolization syndrome. She responded well to paracetamol. Following complete resolution of her symptoms she was discharged home with a plan to follow up as an outpatient.

Given the patient’s child-bearing age, magnetic resonance imaging (MRI) was used to assess progression of AMLs as well as to confirm resolution of the right-sided haemorrhagic lesion. These follow-up investigations confirmed a gradual decrease in the size of the lesions with no further bleeding (Fig. 5). We are pleased to report that the patient has had no further
complications. She continues to have 6 monthly surveillance MRI scans. Moreover, she is due to be followed up by the clinical genetics team shortly for tuberous sclerosis testing.

**Discussion**

AML is a rare, benign tumour composed of smooth muscle, blood vessels and adipose tissue. The incidence of AML is thought to be around 0.3–3%. They most commonly affect females with some population studies showing a male/female ratio of 1:9\(^1\) with a median age at presentation of 50 years\(^2\).

There is a long-established association between renal AML and tuberous sclerosis, most notably in cases of bilateral or multiple AMLs. Studies show up to 25% of patients with AML may have
tuberous sclerosis\(^2\). In these patients, tumours are often larger, present at a younger age and are symptomatic requiring intervention.

Common presentations of AMLs are abdominal flank pain, gross haematuria and/or a tender abdominal mass\(^3\). This triad of symptoms has been described as Lenk’s triad. However, with the increasing use of abdominal imaging, large numbers of AMLs are now identified incidentally. As a general rule, AMLs larger than 4 cm warrant further investigation and close monitoring as they are associated with a greater risk (>50%) of becoming symptomatic or bleeding\(^4\). Larger AMLs (>8 cm) are prone to haemorrhage causing a presentation not dissimilar to an acute abdomen with features of shock.

A literature search performed on Medline on 27 July 2012 showed 78 results for “angiomyolipoma AND pregnancy”. Among these, 27 were case reports describing ruptured AMLs in the context of an ongoing pregnancy (5 of these were treated with embolization). Only one case report described haemorrhagic AML occurring in the postpartum period after a term delivery. This case report is the first to describe haemorrhagic AML in the context of a miscarriage.

Evidence suggests a possible relationship between pregnancy and the incidence of rupture of AMLs. It is thought that increased abdominal pressure, increased renal blood flow and hormonal influences may be responsible for causing accelerated growth of AMLs during pregnancy, contributing to the increased risk of haemorrhage\(^5\). There are few data available, however, on the quantitative relationship between gestational age and the risk of haemorrhage associated with AML.

Treatment options for AMLs can include monitoring, total or partial nephrectomy or, increasingly, transarterial embolization. The choice of treatment is dictated by the severity of the symptoms and the size of the AML\(^6\), as well as the localization of tumour in relation to the renal vessels and the suspected malignant potential\(^7\). In view of the benign nature of AMLs, renal-preserving treatments such as nephron-sparing surgery and transarterial embolization (TAE) are preferred\(^7\); TAE is the first choice treatment in most centres. Large retrospective studies have demonstrated that minimally invasive TAE results in a significant reduction in the size of symptomatic AMLs (mean percentage reduction in size of 33% at 6 months follow-up and 43% at long-term follow-up) with a very low incidence of complications such as post-embolization syndrome (characterized by fever, nausea, vomiting and pain), renal failure, relapse and abscess formation\(^8\).

Prognosis is generally very good for AMLs with exceptionally good results reported in the literature with both major forms of treatment, with no significant reported complications including renal impairment in an extended period of follow-up\(^7\).
Routine medical follow-up is recommended for symptomatic patients presenting with spontaneous haemorrhage of previously unknown AML. In patients with bilateral or multiple AML, testing for tuberous sclerosis is recommended. AMLs measuring between 4 cm and 8 cm are thought to have the most variable natural history and so regular surveillance is particularly warranted[9].

There is no general consensus on the management of larger asymptomatic AMLs (>8 cm). Although this has traditionally been managed conservatively with regular surveillance scanning, there is an emerging trend towards elective embolization in patients before the development of symptoms[10].

Teaching points

- AMLs are benign renal tumours and may be an incidental finding. In cases where they are >4 cm, they require regular surveillance as they carry a high risk of bleeding.
- As many as 25% of patients with AMLs may have an underlying diagnosis of tuberous sclerosis. Genetic testing is appropriate in patients with multiple or bilateral disease.
- AMLs are often asymptomatic, but Lenk's triad of abdominal pain, gross haematuria and a palpable, tender abdominal mass describes a common presentation.
- The diagnosis of AML with haemorrhagic complications should be considered in the differential of acute abdomen in the context of a recent pregnancy or miscarriage.
- Prognosis for AMLs is very good; nephron-sparing surgery and TAE are the mainstays of treatment in symptomatic patients.

Conflicts of interest: None

References