

Ruptured angiomyolipoma, a novel entity for emergency physicians in the differential diagnosis of haemorrhagic shock in a female patient of reproductive age

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SUMMARY

A woman in her 30s presented to the emergency department with acute onset, progressively worsening left-sided abdominal pain after exercise. She was found to be hypotensive and diaphoretic, with free intraperitoneal fluid detected on bedside point-of-care ultrasound. Resuscitation was initiated, a presumptive diagnosis of ruptured ectopic pregnancy was made, and obstetrics and gynaecology were consulted. Point-of-care urine pregnancy testing, however, was negative, and subsequent CT angiography of the patient's abdomen revealed an angiomyolipoma (AML) with active haemorrhage. Ultimately, embolisation was performed in the interventional radiology suite, with improvement of patient haemodynamics. Ruptured AML is a rare, life-threatening condition that needs to be included in the differential diagnosis of haemorrhagic shock in female patients of reproductive age presenting to the emergency department.

department via ambulance with abdominal pain approximately 1 year ago. She reported progressively worsening left lower quadrant abdominal pain radiating to her left flank that began abruptly after exercise 1 day prior. She also complained of numbness to her left lower extremity, as well as feeling a 'bulge' in the left lower quadrant of her abdomen.

On arrival to the emergency department, the patient had an initial blood pressure of 90/66 mm Hg, heart rate 84 bpm. She was tachypneic with a respiratory rate of 24, and an oxygen saturation of 100% on room air. Her initial oral temperature was 34.4°C. Generally, the patient appeared diaphoretic and pale, with cool skin and diminished capillary refill. Her abdomen was distended and exquisitely tender to palpation of the left lower quadrant with involuntary guarding. Throughout the exam, the patient periodically stated 'I feel like I am falling asleep'.

BACKGROUND

Renal angiomyolipomas (AMLs) are common, benign renal tumours. Their primary life-threatening complication is retroperitoneal haemorrhage. Acutely bleeding AMLs may manifest as Lenk's triad, with acute abdominal tenderness, flank pain and signs of internal bleeding.¹

While tumour size has historically been used to determine risk of rupture, a 2018 literature review revealed that multiple factors ultimately influence rupture risk.² These include low velocity force, exercise, female sex, pregnancy, intratumoral aneurysm formation and genetic association, primarily with tuberous sclerosis complex (TSC).^{1,2}

We report a case of ruptured AML requiring massive transfusion of blood products for resuscitation and haemorrhage control by embolisation with coiling in the interventional radiology (IR) suite. This case demonstrates the importance of considering vascular abnormalities such as AML in the differential diagnosis of haemorrhagic shock in young female patients.

CASE PRESENTATION

A woman in her 30s with a medical history of hypertension, chronic kidney disease (CKD) not yet on dialysis prior to transplantation, status post living unrelated kidney transplant one decade prior and IgA nephropathy, presented to the emergency

INVESTIGATIONS

A point-of-care ultrasound performed by the emergency physicians revealed free intraperitoneal fluid and a large complex-appearing left-sided intra-abdominal mass. Initial labs were significant for venous blood gas (VBG) showing pH 7.31, pCO₂ 41 and bicarbonate 20.6 mmol/L. Her haemoglobin was 82 g/L, decreased from baseline of 123 g/L. The lactate was elevated to 2.6. Her blood urea nitrogen (BUN) was 30 mg/dL and serum creatinine was 2.59 mg/dL, increased from baseline 21 mg/dL and 2.01 mg/dL, respectively, 6 months prior. Obtaining a urine pregnancy initially was difficult due to minimal urine output; however, once obtained, this test was negative. A CT scan of the abdomen and pelvis with intravenous contrast demonstrated large volume hemoperitoneum and left retroperitoneal haematoma with contrast extravasation from the left renal artery. A ruptured AML in the region of the patient's atrophic native left kidney was the suspected cause of haemorrhage. There were no changes noted on CT to the patient's transplanted kidney in the right lower quadrant.

DIFFERENTIAL DIAGNOSIS

Given the patient's age, free intraperitoneal fluid on bedside ultrasound, and vital sign derangements, the initial differential diagnosis was most suspicious for ruptured ectopic pregnancy. Once the



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Figure 1 Coronal view of the patient's large intraperitoneal haematoma on CT scan when she presented in the emergency department.

pregnancy test was found to be negative, the differential had to be expanded to include ruptured splenic artery aneurysm, haemorrhage from a neoplastic process, haemorrhagic ovarian cyst, rupture of an arteriovenous malformation or other vascular catastrophe.

TREATMENT

Resuscitation began with placement of two large bore peripheral intravenous lines. Simultaneously, the patient was started on 2 L of supplemental oxygen via nasal cannula. Crystalloid infusion



Figure 2 Axial CT images of the patient's intraperitoneal and retroperitoneal haematoma, from the level of L2 vertebra.



Figure 3 Axial CT images of the patient's intraperitoneal and retroperitoneal haematoma, from the level of L4 vertebra.

was initiated with 1 L of lactated ringers solution. Two units of uncrossmatched O+packed red blood cells were given due to presumed intra-abdominal bleeding causing haemorrhagic shock.

A multidisciplinary discussion was then conducted with general surgery and urology services. The general surgery service recommended IR consultation for embolisation, and acknowledged the patient would require operative intervention if IR intervention failed. General surgery had recommended against initial operative intervention for multiple reasons. These included the possibility of the patient improving with a less invasive approach by IR, and because immediate surgery might remove any possible tamponade effect being achieved with active bleeding on closed retroperitoneal and intraperitoneal spaces, potentially acutely worsening the patient's already tenuous clinical picture. The urology service agreed the patient would be best managed initially by IR or general surgery at this stage but remained on consult and followed the patient's clinical course in the event that nephrectomy of the atrophic, native, left kidney with AML should become necessary. The patient was deemed a candidate for IR embolisation procedure given that active haemorrhage was noted on CT imaging (figures 1–4).

The patient was managed by the emergency medicine physicians for a number of hours, during which time she became oliguric and continued to have a distended, tense abdomen. She received a 1:1:1 transfusion of red cells, plasma and platelets. Tranexamic acid with adjunctive aliquots of calcium gluconate was administered while awaiting IR suite availability. The patient was resuscitated with a total of 6 units of packed red blood cells. Ultimately, the patient was taken to the IR suite where a left renal angiogram demonstrated an actively haemorrhaging left renal artery (figures 5 and 6). The patient's left renal artery was embolised successfully with 5 cm³ of ethanol and ×6 coils. The patient's haemodynamics stabilised while in the IR suite, and she was admitted to the surgical-trauma intensive care unit (ICU) for further management.

OUTCOME AND FOLLOW-UP

While in the surgical ICU, the patient developed worsening acute allograft dysfunction, over her CKD with hyperkalaemia and oliguria. General nephrology and transplant nephrology services were consulted. Acute allograft dysfunction was thought



Figure 4 Axial CT images of the patient's intraperitoneal and retroperitoneal haematoma, from the level of the sacrum.

to be secondary to acute tubular necrosis due to the massive retroperitoneal bleed, hypotension and possibly a component of contrast nephropathy. A renal ultrasound of the patient's transplanted kidney demonstrated normal flow during her hospitalisation. Due to a potassium of 7.7 mmol/L, haemodialysis was initiated via temporary internal jugular venous dialysis catheter. She underwent three consecutive days of haemodialysis. Her creatinine peaked at 4.18 mg/dL, with an estimated glomerular filtration rate (GFR) of 13 mL/min. Fortunately, her kidney function and urine output improved and she did not need any

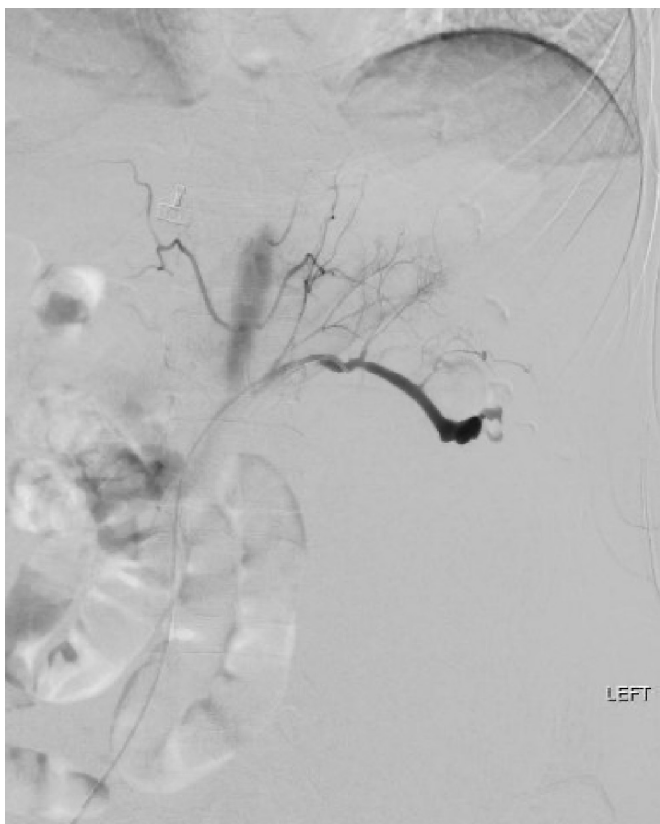


Figure 5 Angiogram still image demonstrating active extravasation involving the left renal artery.



Figure 6 Angiogram still image showing successful embolisation of the patient's left renal artery after coils placed.

further dialysis. Multidisciplinary discussions with urology, IR, transplant surgery and transplant nephrology continued during her hospitalisation. As she stabilised and clinically defervesced, the decision was made not to proceed with native nephrectomy during the current admission.

The patient's creatinine eventually returned to her baseline of approximately 2 mg/dL. She was discharged home from the hospital in stable condition 10 days following presentation to the emergency department. One month following hospitalisation, the patient has followed up as an outpatient with transplant nephrology and is doing well, without requiring any surgical intervention. Her renal function remains stable. Follow-up imaging included a non-contrast CT and renal ultrasound, which revealed diminishing haematoma size and no concerning features.

DISCUSSION

AMLs are a heterogeneous group of benign renal neoplasms composed of varied proportions of blood vessels, smooth muscle and adipose.¹ They are the most common benign renal tumour, with prevalence estimated between 0.2% and 0.6% of the general population.¹ Approximately, 80% of cases of AML are small and sporadic, with predominance in females of child-bearing age.³ AMLs associated with genetic conditions such as TSC or pulmonary lymphangioleiomyomatosis (LAM) occur mostly in younger patients, and are more likely to be large and bilateral.³

Tumour-specific risk factors for AML rupture include size >4 cm, development of intratumour aneurysms >5 mm, and association with TSC or LAM.^{2,4} Pregnancy poses increased risk of rupture due to physiologic changes that predispose to tumour and aneurysm growth.⁴ One literature review determined that mean gestational age for AML rupture in pregnant patients is approximately 27.7 weeks.⁵ Non-tumour-related risk factors include coagulopathic states, and the COVID-19, as noted in a recent case report.⁶ On literature review, there does not appear to be an increased risk of AML in patients with transplanted kidneys.

AMLs may be categorised as classic or epithelioid. Classic renal AMLs typically contain higher proportions of adipose tissue, while epithelioid AMLs have the potential to become malignant and subsequently metastasise.¹ AMLs are most commonly diagnosed using CT imaging. Most are found incidentally on imaging in patients presenting for unrelated complaints, and most do not progress to any significant morbidity or mortality. MRI is useful in distinguishing AML from other renal tumours.

Tumour size is generally used to determine treatment approaches for AMLs. While there is some debate in the scientific community, asymptomatic AMLs <4 cm are accepted as appropriate for observation. Some patients may benefit from immunosuppressive therapies such as mTOR inhibitors, which slow vascular epithelial growth and aid in reducing tumour size.⁷ Patients with incidentally detected AMLs should be referred to nephrology for further management.^{8,9} Renal artery embolisation is generally recommended as first-line treatment for bleeding AMLs, and may also be employed preventatively to treat AMLs between 4–10 cm.^{9,10} Patients may also undergo nephron-sparing surgery or full nephrectomy, radio frequency ablation and other therapies for symptomatic or large AMLs.^{11,12} For patients in haemorrhagic shock who are too unstable to be transferred to the IR suite, resuscitative endovascular balloon occlusion of the aorta may be considered as a life-saving bridge to embolisation therapy or emergent nephrectomy.¹³

Our patient was managed by a multidisciplinary team who after careful discussion opted to proceed with selective arterial embolisation of the renal artery to control life-threatening haemorrhage. Arterial embolisation therapy is considered the first line of therapy for patients with AML presenting with acute haemorrhage. Selective arterial embolisation has been shown to be safe and effective therapy for actively haemorrhaging AMLs.^{14–16} Arterial embolisation therapy has success rates emergently controlling haemorrhage reported over 95%.¹⁷ It can reach most AMLs including some involving the renal hilum. It limits blood loss, and has been shown to decrease the need for surgery, as well as improve outcomes of subsequent nephron-sparing surgery should it be necessary.^{14–16} There are concerns that patients who are treated with embolisation therapy may need reintervention for further bleeding or growth of the AML. However, some studies have demonstrated a low risk of reintervention for further bleeding or tumour growth following successful embolic therapy of patients presenting with haemorrhage from AML.^{16,18} Following arterial embolisation therapy, a significant number of patients develop a complication known as postembolisation syndrome with fever, flank pain and leucocytosis.^{15,18} If surgery is required following selective arterial embolisation, nephron-sparing surgical techniques are preferred over nephrectomy unless the AML has replaced most of the renal parenchyma or there is concern for malignancy. Nephron sparing surgery preserves renal function and improves overall survival compared with nephrectomy.¹⁴

Regarding the element of tamponade effect from bleeding in both the peritoneal and retroperitoneal spaces, this is well described in the literature regarding bleeding confined to the retroperitoneum. In a retrospective review of retroperitoneal bleeding sustained during cardiac cath, bleeding stopped spontaneously and some element of tamponade effect was achieved in 84% (approximately 39) of the 45 total patients who sustained this complication of catheterisation, so long as adequate blood product replacement was continued.¹⁹ There are limited descriptions in scholarly literature of pressure from fluid in the peritoneum tamponading retroperitoneal bleeding. A case report from Gutowski *et al* for a patient undergoing peritoneal dialysis with

spontaneous retroperitoneal bleeding demonstrated improvement of hypotension when the patient had peritoneal dialysate infused, thus showing that pressure from intraperitoneal fluid improved haemodynamics in the setting of retroperitoneal bleed.²⁰ While it cannot be shown definitively that tamponade effect from both intraperitoneal and retroperitoneal bleeding partially stabilised our patient, her surgical teams felt that the possible benefit of this effect outweighed the risk of immediate surgery.

The patient in this case was a woman of reproductive age presenting in haemorrhagic shock, with point-of-care ultrasound demonstrating free fluid in the abdomen. This clinical scenario was most concerning for ruptured ectopic pregnancy, and rapid point-of-care pregnancy testing is critical in confirming this diagnosis. In this case, the patient's oliguria delayed urine pregnancy testing. An important alternative not considered in this case but of importance for future practice would be placing a sample of whole blood on a point-of-care urine human chorionic gonadotropin (hCG) test. A recent study demonstrated that whole blood on a point-of-care urine pregnancy test is 95.8% sensitive down to 5 mIU/mL serum hCG.²¹ This method of identifying whether or not a patient is pregnant may be very useful in the management of shock in a woman of reproductive age.

AMLs are more common in reproductive age women and have higher risk of rupture in pregnant patients. This is the same demographic and risk profile for ectopic pregnancy. Providers caring for reproductive age women need to keep AML as a rare but important differential diagnosis after ruptured ectopic pregnancy in patients with haemorrhagic shock.

Patient's perspective

I am thankful for all the medical staff that have helped me. I'm glad to be alive and doing well!

Learning points

- ▶ AMLs are benign renal tumours that may rupture and result in life-threatening retroperitoneal and intraperitoneal haemorrhage.
- ▶ AMLs may rupture with minimal trauma such as exercise or even abdominal palpation.
- ▶ AMLs are more common in women of reproductive age, and are an important consideration in the differential diagnosis of haemorrhagic shock in these patients.
- ▶ Whole blood testing on a point of urine pregnancy test can rapidly determine pregnancy status in oliguric or anuric female patients, thus guiding differential diagnosis.
- ▶ Renal artery embolisation is generally recommended as first-line treatment for bleeding AMLs.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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