



## Spontaneous Rupture of a Renal Angiomyolipoma in a 20-Week Pregnant Woman: Case Report and Literature Review

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### Abstract

**Background:** Lymphangiomyomatosis is a rare disease, occurring predominantly in women. Its manifestations include renal angiomyolipomas which carry a higher risk of rupture during pregnancy.

**Case Presentation:** We report a case of a 20-week pregnant woman who presented with severe flank pain and haemorrhagic shock due to an angiomyolipoma rupture and was successfully treated with arterial embolization.

**Conclusions:** Endovascular embolization proved to be a safe and effective treatment option for managing the ruptured angiomyolipoma, stabilizing the patient during pregnancy. Complementary mTOR therapy further contributed to significant tumor shrinkage, highlighting a combined approach for optimal outcomes.

### OPEN ACCESS

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**Keywords:** Lymphangiomyomatosis; Renal angiomyolipoma; Pregnancy; Arterial embolization

### Introduction

Lymphangiomyomatosis (LAM) is a rare progressive disease, occurring predominantly in women, which affects mainly the lungs, and the kidneys and the lymphatic system.

It can be sporadic (S-LAM) or associated with tuberous sclerosis complex (TSC-LAM) [1,2].

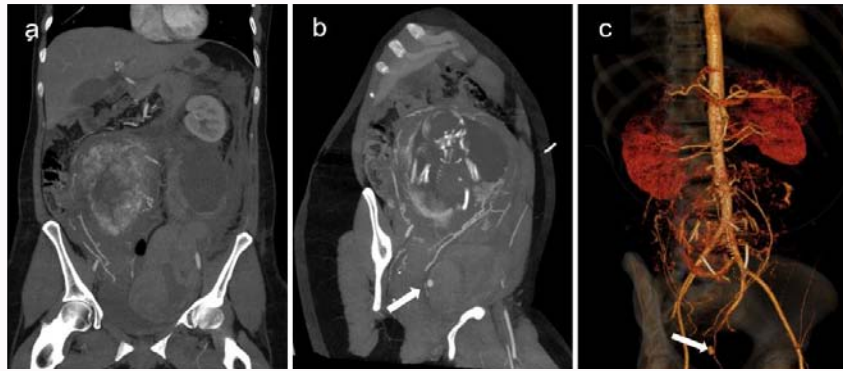
The cause is loss of function mutations in the tumour suppressor genes TSC1 or TSC2 that encode hamartin and tuberin proteins, respectively, leading to inappropriate signalling through the mTOR pathway, resulting in multifocal nodular proliferation of immature smooth muscle and perivascular epithelioid cells (LAM cells) and formation of cysts. Female sex hormones also appear to be implicated in the pathogenesis of LAM, despite the mechanism not being well understood, histopathology exams showed positivity for hormonal receptors [1,2].

Clinical manifestations include dyspnoea on exertion, cough, haemoptysis, chyloptysis, pneumothoraxes, chylous pleural effusions, abdominal masses including renal angiomyolipomas (AML) and lymphangiomyomas, oedema of lower extremities, pelvic or retroperitoneal lymphadenopathy, chyloperitoneum, chyluria and chylopericardium [1,3].

The angiomyolipomas are benign tumours, containing smooth muscle, blood vessels and fat, which occur in 30% of patients with S-LAM and up to 80% of those with TSC-LAM. They are mostly asymptomatic and they can present as a palpable mass, abdominal pain, recurrent haematuria, renal insufficiency or sudden flank pain and signs of haemorrhagic shock provoked by its rupture [4].

For AMLs, the presence of fat gives it a characteristic appearance on noncontrast computed tomography (CT), and biopsy is rarely needed [4].

Intervention is recommended for symptomatic AMLs and should be considered in lesions with intralesional aneurysms  $\geq 5$  mm, rapid growth ( $>5$  mm/year), large size (especially  $>6$  cm), or in women of childbearing age, particularly during pregnancy or hormonal therapy. Selective embolization



**Figure 1:** 1A. Coronal CTA image showing the pregnant uterus on the right side and a giant AML on the left. 1B. Sagittal CTA image presenting the foetus in the uterus and below a blush of contrast indicating active bleeding of the AML (arrow). 1C. Reconstruction CTA image: the bleeding arises from a branch of the left internal iliac artery (arrow).

is the first-line treatment, while nephron-sparing surgery is considered if a malignant tumour cannot be ruled out [5]. Thermal ablation is a promising but less established option [6]. AMLs can shrink with mTOR inhibitors but may regrow after treatment discontinuation [7].

This report aims to highlight the challenges encountered in treating rare cases of ruptured AMLs, particularly in pregnant women. By sharing knowledge and exploring potential treatment options, we aim to foster discussion and improve patient outcomes.

### Case Presentation

A 39-year-old female, 20 weeks pregnant after in vitro fertilization, with a 1-year LAM diagnose (multiple lung cysts, a left perirenal and lumboaortic AML with 220 × 110 mm, and multiple small uterine leiomyomas), went to the emergency department 2 times with left iliac fossa pain irradiating to the lumbar area and left lower limb and was discharged with pain medication, after no relevant analytic or imaging findings were found.

At the third visit, she presented with increased left iliac fossa pain irradiating to the lumbar area and left lower limb, no rebound or guarding at palpation, and also hypotension and tachycardia. Laboratory examination showed a drop on the haemoglobin value (4 g/dL over 72 hours). After ponderation of the risks and benefits of radiation exposure to the mother exposure to the mother and foetus, a computed tomography was performed and showed an active arterial a was performed an active arterial phase punctiform haemorrhage from the known AML (Figure 1).

After resuscitation, and multidisciplinary discussion, considering the non-viability of the foetus if delivered at that time, maternal life risk and a difficult surgical approach, it was decided to proceed with embolization, with the informed consent of the patient.

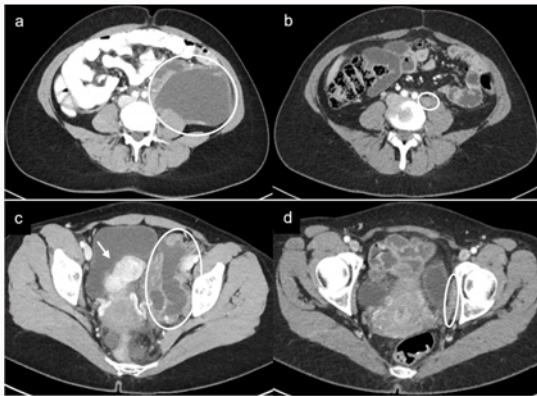
The procedure was performed under local anaesthesia through a right femoral puncture, using a 5F introducer. The aortic bifurcation was crossed with a RIM catheter. Selective arteriography was conducted using a 4F MP catheter, followed by super-selective catheterization with a Maestro® 2.8 F microcatheter (Merit Medical). Embolization of the tumour feeding arteries, branches of the left internal iliac artery, was achieved by deploying three coils: two Interlock™ 2D coils (3 mm × 60 mm) and one Interlock™ VortX™ coil (2 mm × 5 mm × 58 mm), all by Boston Scientific. The success of the procedure was confirmed via control angiography. Haemostasis at the puncture site was secured using an EXOSEAL® 5F vascular closure device (Cordis) (Figure 2).



**Figure 2:** 2A. Selective angiogram displaying the origin of the haemorrhage in a distal branch of the left internal iliac artery (arrow); 2B and 2C. Final angiographic control demonstrating coil placement and exclusion of AML vasculature (arrow).

Radiation safety was further optimized through the use of an iso-osmolar iodinated contrast agent (iodixanol 320 mg I/mL), diluted to 50% with normal saline (total volume 100 mL), to ensure adequate angiographic visualization while limiting total contrast load and avoiding repeated imaging runs. Two oblique projections (right and left anterior at 45°) were performed at 3 fps for target arterial branch catheterization, followed by postero-anterior projections (0°) at 3 fps and 2 fps. Pulsed fluoroscopy at 15 fps and 14 radiographic acquisitions (8 at 3 fps and 6 at 2 fps) were utilized. Exposure parameters included 80 kV and 17–51 mA.s. Copper (0.1 mm) and aluminum (1 mm) filters reduced secondary radiation. The source-to-patient distance was increased (working height of 81cm), while source-to-detector distances were minimized (117 cm for LAO, 112 cm for RAO, and 97–98 cm for PA). Larger fields of view (48, 37, and 31 cm) and maximum collimation were applied. Total exposure time was 46.7 minutes, with a dose-area product of 77.490 Gy·cm<sup>2</sup> and an estimated skin entrance dose (Air Kerma) of 607 mGy.

No complications occurred intra-operatively or during immediate follow up, with the wellbeing of the foetus assured by an obstetrician evaluation.



**Figure 3:** 3A, 3C – Before pregnancy: the patient presents a giant AML (white circles) - quistic mass anterior to left psoas muscle measuring 220 x 110 mm - and uterine leiomyomas (blue arrow); 3B, 3C – AML 2 years after coil embolization plus 14 months of Sirolimus: significant reduction in size, with maximum axial diameters of 26 x 7 mm, and also a significant shrinkage of the uterus leiomyomas.

**Table 1:** Measures to reduce radiation dose.

Measures to reduce radiation dose
To use higher tube voltage (kVp) with lower current (mA)
To reduce frame rates
To prioritize pulsed fluoroscopy
To perform digital subtraction angiography only when strictly necessary
To use preferentially postero-anterior projections
To limit magnification
To use X-ray field collimation
To use X-ray field filters
To place the image intensifier as close to the patient as possible while maintaining maximum distance from the X-ray tube

The patient gave birth to a term healthy baby, and after 6 months of lactation, started treatment with Sirolimus.

At the 1-year follow-up appointment, the patient reported no abdominal symptoms. Control CT imaging revealed a left retroperitoneal AML measuring 65 x 26 x 37 mm, located anterior to the psoas-iliac muscle, with residual fluid-filled loc

At the 2-year follow-up, control CT imaging demonstrated a further reduction in AML size, with maximum axial diameters of 26 x 7 mm, and also a significant shrinkage of the uterus leiomyomas (Figure 3).

## Discussion

LAM is not a contraindication *per se* to pregnancy; however women must be informed of the higher risk for disease progression and complications [5,8].

AML may grow during pregnancy, increasing the risk of its rupture and bleeding, haemorrhagic shock and foetal death. A number of factors are thought to be associated with this, namely increased circulating plasma volume, increased abdominal pressure, elevated blood pressure and oestrogen-induced smooth muscle proliferation [9].

Consequently, women with a known AML should have a preconception multidisciplinary evaluation and a close follow up during pregnancy.

In some cases, prophylactic pre-pregnancy transcatheter arterial embolization may be considered, owing to resulting in decreased growth and risk of bleeding during pregnancy, as shown by Toei H et al. [9]

The patient was counselled regarding the risks of disease progression and informed that, although emerging data suggest potential safety, definitive evidence on sirolimus use during pregnancy and lactation remains limited. After considering this information, the patient opted to postpone treatment.

When rupture and haemorrhage occurs during pregnancy, hemodynamic stability is a key factor in the treatment choice. For hemodynamically unstable patients, arterial embolization or emergency surgery are the main options of treatment. For asymptomatic patients, a conservative approach may be chosen, and definitive treatment may be postponed until the second trimester, after organogenesis, or the postpartum period [10]

Arterial embolization can be done under local anaesthesia, has a high technical success rate (>90%) and a quicker recovery [11].

Given the patient's instability, we proceeded with embolization.

Although a trans-radial approach offers greater flexibility in patient positioning during and after the procedure—an advantage in pregnant patients by mitigating the risk of vena cava compression—we opted for the standard femoral approach in this case [12].

Although particles are commonly preferred for AML treatment, alternative embolic agents include coils, vascular plugs, absolute ethanol, and glue. Available evidence does not demonstrate the superiority of any of these agents [13]. However, coils may be less effective due to their predominantly proximal occlusion, which can allow collateral revascularization and increase the risk of recurrence [14–17].

In this case, coils demonstrated successful and complication-free use for two years of follow-up.

The main complications of embolization are post-embolization syndrome (POS) (inflammation, fever, leucocytosis, and flank pain) and liquefactive necrosis. POS generally resolves spontaneously, whereas percutaneous drainage might be needed for liquefactive necrosis [10,18]. Renal function deterioration is also a possible complication, nonetheless it can take place either with surveillance, embolization, or surgery [19].

However, the biggest disadvantage of arterial embolization is the reintervention rate up to 20% [20].

Another concern is the use of radiation, especially in pregnant women. Fetal risks are considered negligible below 50 mGy, whereas higher doses may be associated with increased risk depending on gestational age, with the highest risk during organogenesis in the first trimester [12,21]. The major adverse effects on the fetus include abortion, teratogenicity, intellectual disability, intrauterine growth retardation, and cancer induction [21].

To optimize radiation protection during this endovascular procedure, we took as many measures as possible, addressing four key aspects: software, geometry, mechanical components, and the iodinated contrast medium, as enunciated in Table 1 [2].

## Conclusion

AML haemorrhage in a pregnant woman is a rare event but may jeopardize both the life of the pregnant woman and the foetus, being a challenging situation to manage

Endovascular embolization, with the utmost care taken to protect the foetus from radiation, swiftly addressed the cause of the haemorrhagic shock, resulting in the patient's survival and allowing the delivery of a term healthy baby. Combined with the initiation of mTOR therapy after puerperium, lead to a significant shrinkage in the tumour size.

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