

Spontaneous Fatal Rupture of a Large Pseudoaneurysm within a Renal Angiomyolipoma following Incomplete Embolization in a Patient with Bilateral Renal Angiomyolipomas

Aman Yusuf Khan¹ Donboklang Lynser¹ Akash Handique¹

¹Department of Radiology and Imaging, NEIGRIHMS, Shillong, Meghalaya, India

Address for correspondence Aman Yusuf Khan, MBBS, Department of Radiology and Imaging, NEIGRIHMS, Shillong, Meghalaya, 793018, India (e-mail: archs7007@gmail.com).

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Angiomyolipoma (AML)—a frequently seen solid renal tumor—is the most common benign tumor of the kidney. It was considered to be hamartomatous in origin and is now considered to be a member of the family of perivascular epithelioid cell (PEComa).¹ Classically thought to be associated with tuberous sclerosis that is an autosomal dominant disease with *TSC1* or *TSC2* gene mutation, around 80% AMLs may present sporadically.¹ They are also seen in patients with lymphangiomyomatosis and included in its diagnostic criteria.² Clinically, the tumor has myriad of presentations ranging from being completely asymptomatic at the time of diagnosis to abdominal pain, hematuria, recurrent urinary tract infection, and intratumoral bleed that can be occasionally massive requiring urgent embolization or nephrectomy.³ On ultrasound AMLs appear as markedly hyperechoic to hypoechoic depending on the fat content. Though they are more frequently echogenic compared with renal cell carcinoma, overlap in imaging features can occur.⁴ On color Doppler, pseudoaneurysms are seen as anechoic structure showing internal turbulent to-and-fro motion.⁵ On computed tomographic (CT) scan, the diagnosis of renal AML depends on the detection of intralesional fat.⁶

Case Presentation

A 24-year-old woman was referred to our interventional radiology unit from another hospital for possible embolization of a large left renal pseudoaneurysm that was considered as high risk for surgery. The patient had history of diffuse abdominal pain with enlarging abdominal girth. On general examination, she was of thin built and emaciated with significant pallor. On examining, there were no characteristic skin lesions. She was having tachycardia, and a lump was palpable, which extended from left hypochondrium to left lumbar region approximately 10 cm in length craniocaudally and 9 cm across extending up to the umbilicus. She had low hemoglobin at 6.4 g/dL and a slightly raised total leucocyte count of 12,000/mL.

Abdominal ultrasonography using Logiq P5 ultrasound system (General Electric Healthcare) with 3.5 to 5 MHz showed bilateral enlarged kidneys. Right kidney shows well-defined three hyperechoic lesions of varying sizes suggestive of fat as main content and left kidney shows well-defined heteroechoic lesion with large pseudoaneurysm occupying upper two-thirds of parenchyma (►Fig. 1A, B). Digital subtraction angiography (DSA) using BV Endura C-arm X-ray machine (Philips) confirmed the large left renal pseudoaneurysm (►Fig. 2C, E) along with significant tumor vascularity. Selective embolization of the tumor mass with polyvinyl alcohol (PVA) particles (PVA 300, Cook Medical) was done. Following this closure of the artery feeding, the pseudoaneurysm was attempted using 4-mm, 5 cm-fibered stainless steel coils (Cook) and 50% *N*-butyl cyanoacrylate (Endocryl, Samarth Pharmaceutical): Lipiodol (Gurbet Laboratories) mixture Endocryl glue. Attempts to occlude the feeding artery failed due to high flow, and the coils were dislodged into the pseudoaneurysm (►Fig. 2F). Next day ultrasound and contrast-enhanced CT (CECT) (128 multislice SOMATOM Definition AS; Siemens AG) showed the formation of a large intracavitary

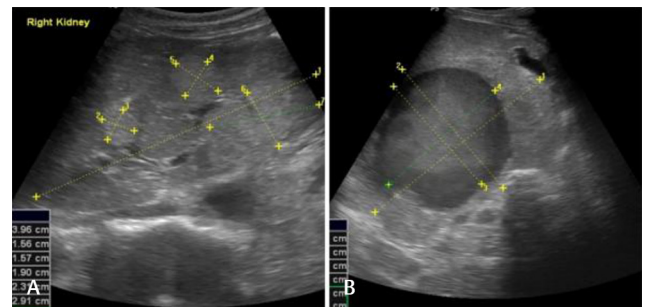


Fig. 1 Ultrasound of a 24-year-old woman with bilateral renal angiomyolipoma and large left renal pseudoaneurysm. (A) Enlarged right kidney with well-defined three hyperechoic lesions of varying sizes. (B) Enlarged left kidney with well-defined round heteroechoic lesion with posterior acoustic enhancement occupying upper two-thirds of parenchyma indicative of pseudoaneurysm.

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Fig. 2 Digital subtraction angiography (DSA) of a 24-year-old woman with bilateral renal angiomyolipoma and large left renal pseudoaneurysm. (A, B) DSA of right kidney shows supernumerary renal arteries supplying right kidney without evidence of any aneurysm. (C–E) DSA of left kidney shows multiple large and small intrasplenic pseudoaneurysms. (F) Dislodged coils post attempted coiling due to large aneurysmal neck.

thrombus (► Fig. 3A, D). The patient also developed post embolization fever and pain that was managed with antibiotics and nonsteroidal anti-inflammatory drugs (NSAIDs). The patient was planned for repeat embolization of the pseudoaneurysm along with either a vascular plug or using a balloon percutaneous transluminal angioplasty (PTA) catheter-assisted flow arrest during embolization coil deployment. However, before we could proceed, the patient started complaining worsening of the pain on the day 5, followed by cardiac arrest possibly from sudden severe internal hemorrhage due to rupture of the pseudoaneurysm.

Discussion

Angiomyolipoma is known to be more prevalent in females. Some studies have suggested that AML can present in a wide age range from 19 to 93 years with overall prevalence being to as high as 0.44%.⁷ The tumor consists of smooth muscles, adipose tissue, and malformed blood vessels as main components making it a triphasic tumor as described pathologically.¹ Up to 80% of AMLs occur sporadically, and whereas most of them present as classic AML with abundant detectable fat on imaging, the rest few may show very small amount of fat.¹ AML is commonly associated with aneurysm formation, seen in as many as 76% of the patients.⁸

The predictors of rupture include tumor and aneurysm sizes. When tumors sized 4 cm or larger and 6 cm or larger were used as predictors of rupture, sensitivity and specificity, were 100% and 38%, respectively, for the former criterion and 100% and 67% for the latter criterion. Mean aneurysm size was significantly larger in the group with ruptured tumor (13.3 ± 6.2 mm; range: 5–22 mm) than in the group with unruptured tumor (2.4 ± 2.9 mm; range: 2–11 mm). In their study of the eight ruptured aneurysms, all were at least 5 mm in sizes, and if aneurysms sized 5 mm or larger were used as criteria, rupture can be predicted with a sensitivity and specificity of 100% and 86%, respectively.⁸ Our case

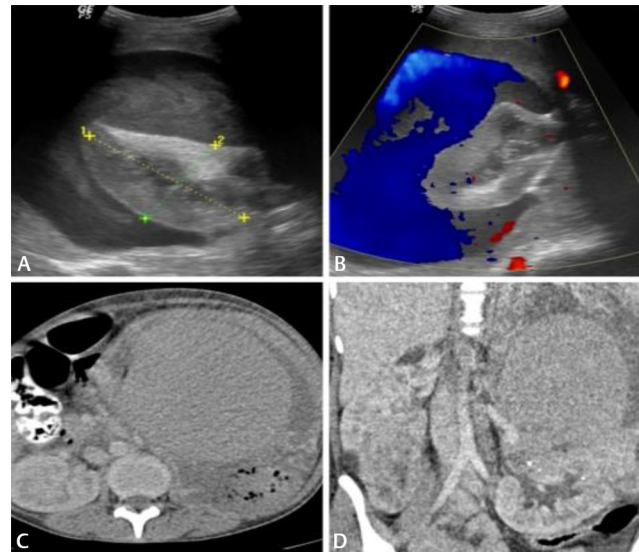


Fig. 3 Abdominal imaging of a 24-year-old woman with bilateral renal angiomyolipoma and large left renal pseudoaneurysm 1 day after attempted coiling. (A, B) Ultrasound shows an eccentric heterochoic content in pseudoaneurysm without vascularity suggestive of thrombus. (C, D) Contrast-enhanced CT scan in coronal and axial view shows the large left intrarenal pseudoaneurysm; note that the clot cannot be readily appreciated.

records an aneurysmal size of 10.1 cm × 10.8 cm, which to our knowledge is one of the largest aneurysms seen in a renal AML. Though the cause of possible aneurysmal rupture in our case is not determined, we hypothesized that the intra-aneurysmal thrombosis post attempted coiling might have led to a pathophysiologic cascade leading to rupture of the aneurysm.

Conclusion

Angiomyolipoma is a common tumor involving the kidneys and presents with varied clinical symptoms that may be associated with pseudoaneurysms. As discussed before, size of the pseudoaneurysm determines the probability of rupturing. As happened in this case in which the patient suddenly went into hypovolemic shock due to rupture of aneurysm, these cases need immediate intervention for decreasing the mortality.

Conflict of Interest

None.

Grant

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