

CASE REPORT

A Case of Life-threatening Rupture of Small Renal Angiomyolipoma with an Unidentified Intratumoral Aneurysm during Follow-up

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Abstract:

We report a case of a life-threatening ruptured renal angiomyolipoma (AML) that did not meet the criteria for prophylactic treatment (tumor >4 cm or intratumoral aneurysm >5 mm) during follow-up. A woman in her 70s was followed up for a 2.5-cm AML with a rich vascular component. An intratumoral aneurysm >5 mm was not identified for 2 years. She complained of a sudden abdominal pain with hypotension, and contrast-enhanced computed tomography revealed a retroperitoneal hematoma with contrast media extravasation from an intratumoral aneurysm. Emergency transcatheter arterial embolization was successfully performed using N-butyl cyanoacrylate glue. Rupture can occur in small AMLs or in AMLs not identified with intratumoral aneurysms during follow-up. AMLs with a rich vascular component at the kidney surface are more likely to rupture.

Keywords:

renal angiomyolipoma, retroperitoneal hemorrhage, treatment criteria, transcatheter arterial embolization, N-butyl cyanoacrylate

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Introduction

Renal angiomyolipomas (AMLs) are benign tumors containing varying fat, smooth muscle, and vascular component proportions. Spontaneous rupture is a severe complication of renal AMLs leading to a life-threatening retroperitoneal hemorrhage. Large tumors >4 cm or intratumoral aneurysms >5 mm have been advocated as prophylactic treatment criteria due to the known risk of rupture [1-3]. We report a case involving a life-threatening rupture of a 2.5-cm AML with a rich vascular component, wherein intratumoral aneurysm >5 mm was not identified during a 2-year follow-up period. Emergency transcatheter arterial embolization (TAE) using N-butyl cyanoacrylate (NBCA) glue was successfully performed.

Case Report

A woman in her 70s was diagnosed with an asymptomatic, sporadic, and classic AML 2 years ago. She had a

history of hypertension and was taking calcium channel blockers. Contrast-enhanced CT (CECT) at the time showed an 18 × 21 × 25-mm mixed-density mass comprising fat and vascular components at the posteroinferior portion of the right kidney. The rich vascular lesion protruded into the posterior perirenal space, although intratumoral aneurysm >5 mm was not identified (**Fig. 1**). No tumor growth or aneurysmal changes were observed by duplex ultrasonography conducted every 6 months on routine follow-up. However, 2 years into her follow-up period, she was brought to the emergency department with an acute right-sided abdominal pain complaint. Upon arrival, she was confused due to hemodynamic instability; her blood pressure and pulse were 80/50 mmHg and 120 beats per minute, respectively. Blood tests at the time of admission showed a hemoglobin level of 5.9 g/dL and a hematocrit level of 17%. CECT revealed a retroperitoneal hematoma (11 × 14 × 20 cm) with contrast media extravasation from an intratumoral aneurysm measuring 5 mm (**Fig. 2**). She was diagnosed with a ruptured AML and immediately transferred to the angiography room for

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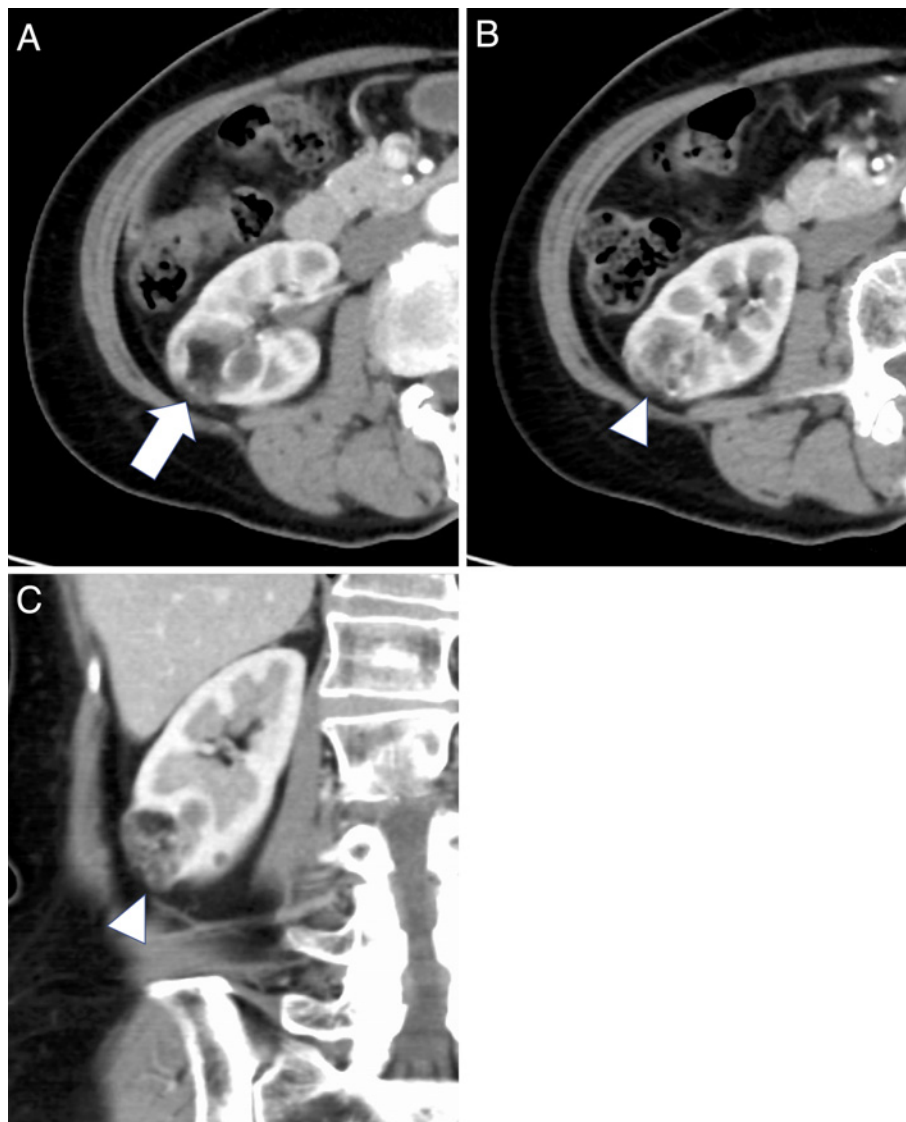


Figure 1. Contrast-enhanced computed tomography (CECT) 2 years before the renal angiomyolipoma rupture in the patient. CECT shows an 18 × 21 × 25-mm mixed-density mass comprising fat (A, arrow) and rich vascular components (1B and 1C, arrowhead) at the posteroinferior portion of the right kidney. CECT shows the hypervascular lesion protruding into the perirenal space, although intratumoral aneurysm >5 mm is not observed (1B and 1C, arrowhead).

TAE.

A 4-F angiographic catheter (RC-09; Medikit, Tokyo, Japan) was inserted into the right renal artery via the right femoral artery. Angiography revealed massive contrast media extravasation from a microaneurysm at the periphery of the tortuous parent vessel (**Fig. 3A**). A 1.9-F microcatheter (Carnelian SI; Tokai Medical, Kasugai, Japan) was advanced into the parent artery as far as possible over a 0.014-inch microguidewire (BEGIN; Asahi Inteccs, Nagoya, Japan) (**Fig. 3B**). NBCA (Histoacryl; B. Braun, Melsungen, Germany) and ethionic oil (Lipiodol; Laboratoire Andre Guerbet, Aulnay-sous-Bois, France) were mixed at a 1:2 ratio (33% NBCA) and slowly and continuously injected to fill the bleeding point of the microaneurysm and the proximal arterial branch (**Fig. 3C**). Angiography after the procedure revealed no further contrast media extravasation (**Fig. 3D**).

Follow-up CT confirmed shrinkage of the hematoma and showed that the bleeding site was filled with an NBCA-lipiodol cast (**Fig. 4**). Subsequent blood tests revealed no progression of anemia or worsening of renal function. The patient was discharged without complications 14 days after the TAE.

Discussion

Most AMLs can be followed up without rupture; however, some can lead to life-threatening ruptures. Therefore, a follow-up algorithm or active treatment criteria should be established. On imaging, tumor size >4 cm or >6 cm and/or the presence of intratumoral aneurysm >5 mm are often used as criteria for aggressive treatment. Asymptomatic patients with small sporadic AMLs or without intratumoral

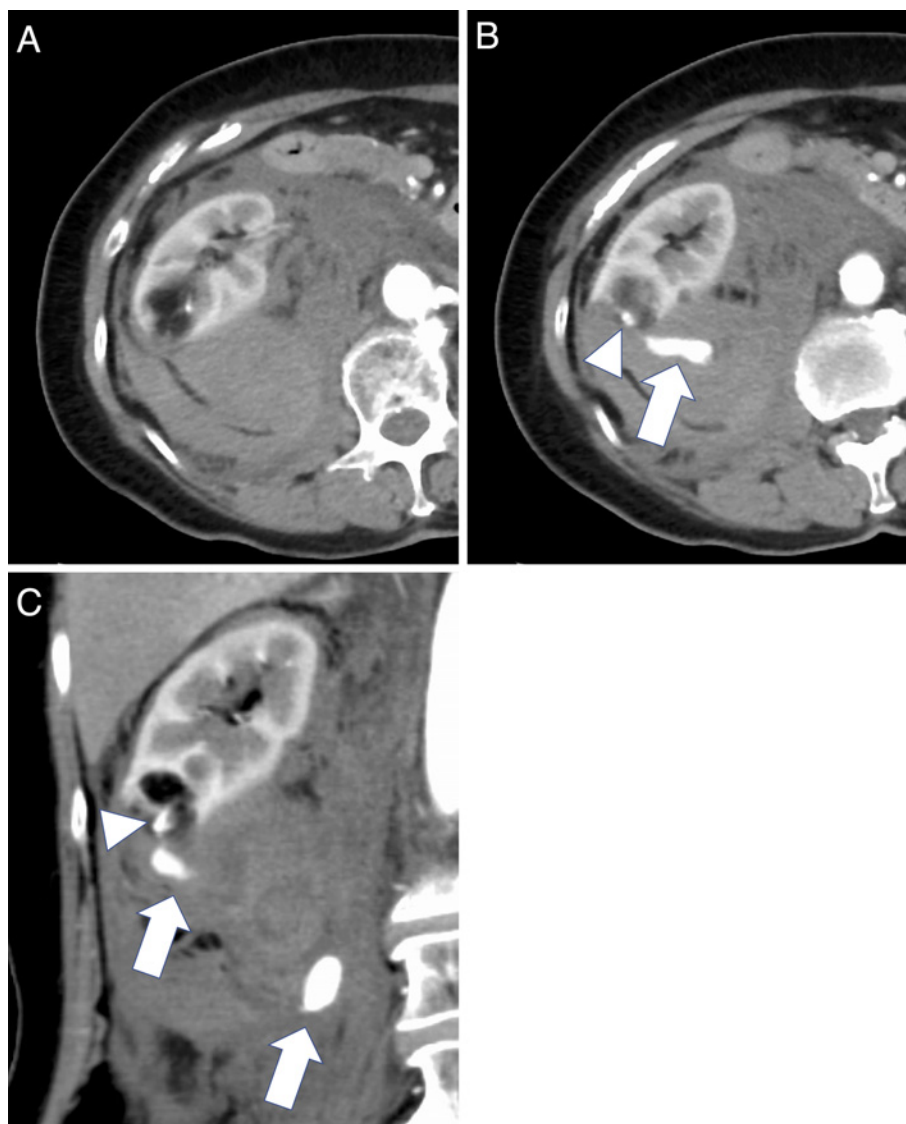


Figure 2. Contrast-enhanced computed tomography (CECT) at the time of admission. The axial cross-sectional images in Figures 2A and 2B, and the coronal cross-sectional images in Figure 3C correspond to the slices in Figures 1A, 1B, and 1C, respectively. CECT shows a retroperitoneal hematoma (11 × 14 × 20 cm) with contrast media extravasation (2B and 2C, arrow) from a microaneurysm (2B and 2C, arrowhead) in the renal angiomyolipoma.

aneurysms >5 mm are generally followed up with ultrasonography, CT, and MRI performed every 6 months or annually [4]. However, in the present case, we encountered a life-threatening rupture of a sporadic AML that did not meet the criteria for prophylactic treatment based on standard follow-up algorithms. Thus, the rupture of AML that does not meet the criteria for prophylactic treatment is rare but could occur, and appropriate follow-up protocols and identification of risk factors for rupture are necessary.

Risk factors for AML rupture have focused on tumor size [1, 5], with tumors >4 cm being traditionally accepted as a prophylactic treatment criterion based on a literature review from 1986 [1]. However, the 4-cm cut-off size has been reconsidered in recent years. A recent systematic review showed that no definitive correlation between AML size and rupture exists, although larger AMLs are likely associated

with a high risk of rupture [3]. Kuusk et al. reported that 9.4% patients with ruptured renal AMLs had a diameter of <4 cm, based on a pooled analysis of data from 441 patients [5]. In the present case, a small 2.5 cm AML caused a life-threatening hemorrhage. The present case and previous studies suggest that tumor size alone should not be used to predict the risk of rupture or as active treatment criteria. Even small AMLs can cause life-threatening rupture, and prophylactic treatment criteria other than tumor size must be established, including the patient's symptoms, tumor vascularity, growth rate, patient background, etc.

Recent studies have shown that intratumoral aneurysms or high vascularity associated with AMLs are stronger predictors of rupture than tumor size. Yamakado et al. reported that intratumoral aneurysm >5 mm was a more reliable predictor of rupture than tumor size >4 cm, suggesting that in-

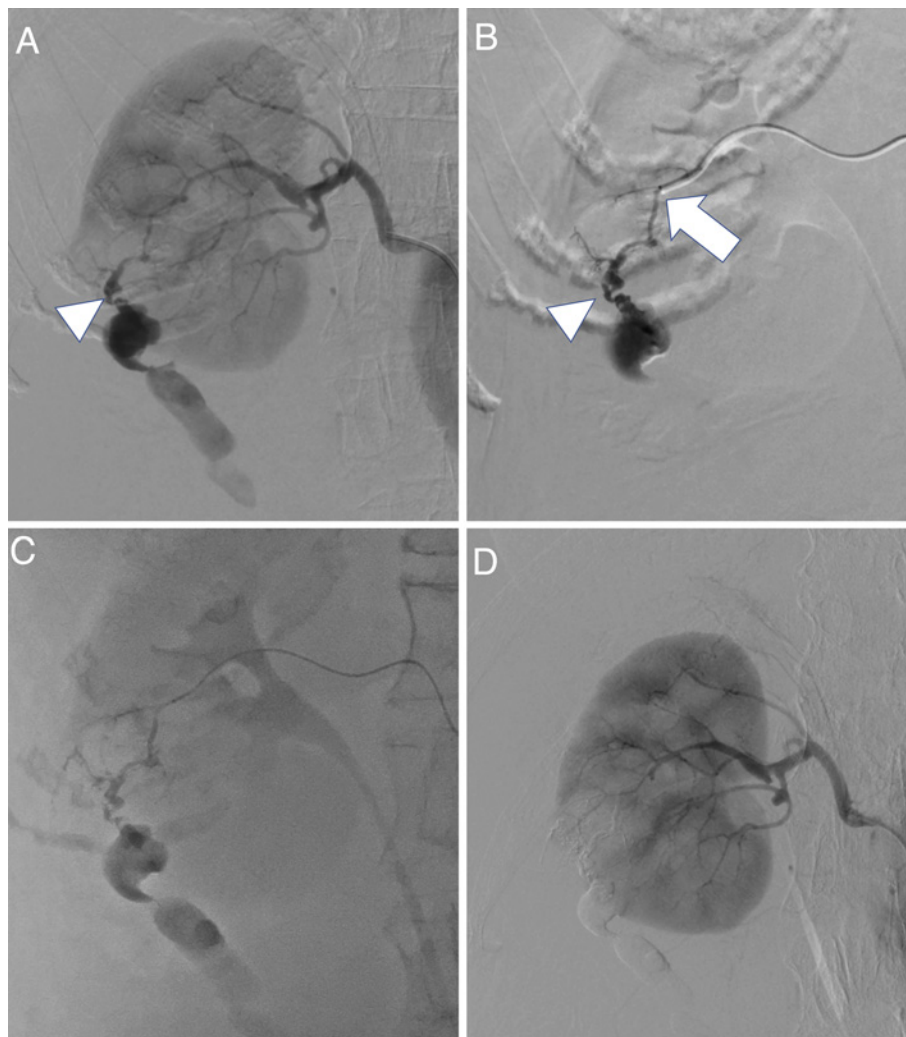


Figure 3. (A) Right renal arterial angiography reveals massive contrast media extravasation from the microaneurysm (arrowhead). (B) Selective angiography from the renal artery branch; the arrow points to the tip of the microcatheter. (C) The ruptured microaneurysm and the proximal arterial branch were embolized with N-butyl cyanoacrylate (NBCA)-lipiodol mixture in a 1:2 ratio (33 % NBCA). (D) Angiography after embolization reveals cessation of contrast media extravasation.

tratumoral aneurysms >5 mm should be recommended as prophylactic treatment criteria for AMLs [2]. Rimon et al. reported that AMLs with high vascularity on angiography are more likely to rupture than those with minimal vascularity [6]. However, changes in vascularity and development in microaneurysms in AML are unknown. In the present case, an AML <4 cm with a rich vascular component was detected, whereas intratumoral aneurysm >5 mm remained unidentified on CECT, and there was a life-threatening hemorrhage after 2 years of steady follow-up. The development of intratumoral aneurysm >5 mm was not observed on duplex ultrasonography performed every 6 months until rupture. Follow-up with ultrasonography is considered a reasonable option for small AMLs, with the advantages of lower cost and without radiation exposure. A case report also showed that color-flow Doppler sonography is useful for detecting intratumoral aneurysms in ruptured AMLs [7]. The intratumoral aneurysm's occurrence was unclear in the present case because ultrasonography's low accuracy may have unsuc-

cessfully detected its appearance. Gandi et al. showed that 64-slice CT angiography can help detect microaneurysms >3 mm in diameter [8]. Therefore, follow-up with CECT, which provides a more objective evaluation of vascularity, may be necessary for AMLs with a rich vascular component. In the present case, if CECT had been performed during follow-up, the development of a microaneurysm could have been detected and timely prophylactic treatment could have been provided.

In hepatocellular carcinoma (HCC), the tumor protrudes from the surface of the liver and is at risk of rupture, causing life-threatening intraperitoneal hemorrhage [9]. However, in renal AML, the relationship between tumor location and risk of rupture is still unknown. Lu et al (2012). reported that six out of seven cases of ruptured renal AMLs with massive retroperitoneal hematomas were located peripheral to the kidney and protruded into the perirenal space [10]. In addition, in two of their cases, AMLs <4 cm in size ruptured and caused a massive retroperitoneal hemorrhage.

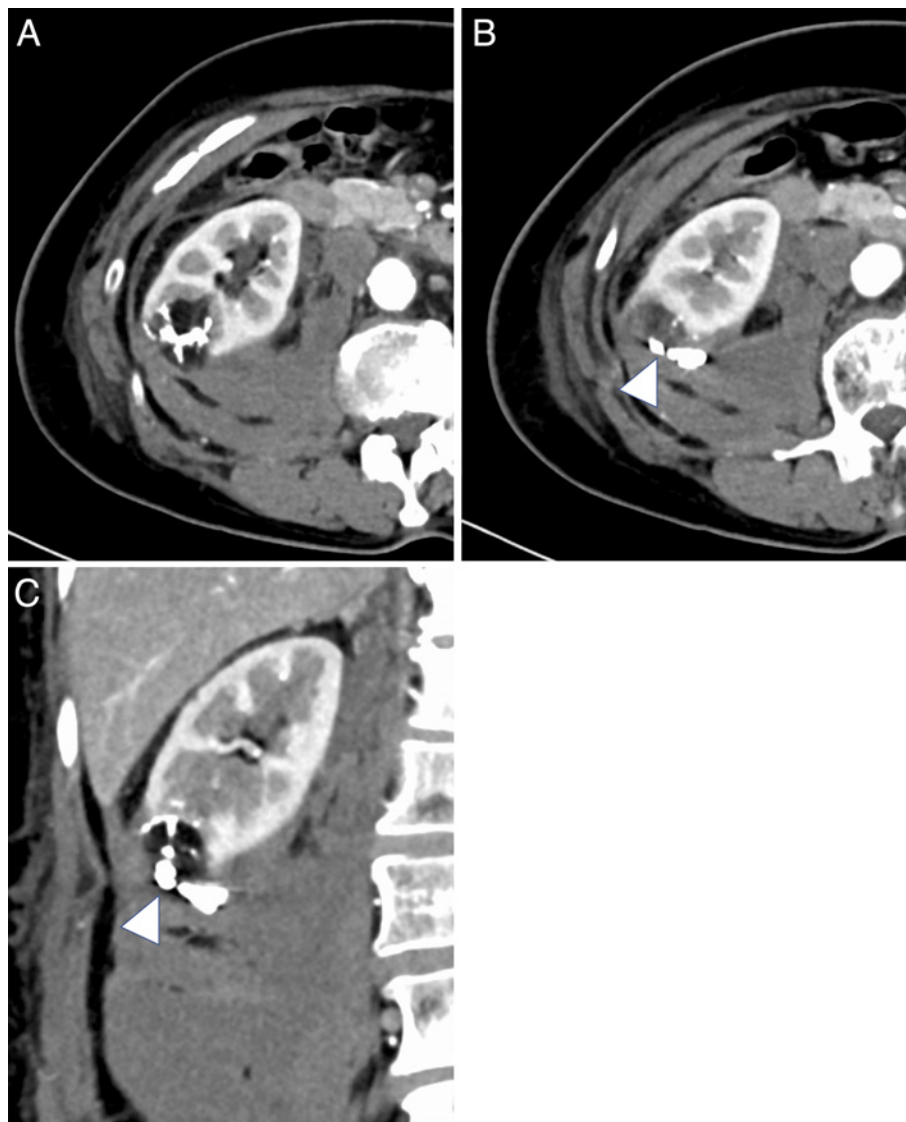


Figure 4. Contrast-enhanced computed tomography 10 days after the transcatheter arterial embolization revealed that the bleeding site was filled with N-butyl cyanoacrylate-lipiodol cast (arrowhead), showing shrinkage of the hematoma.

Similarly, in the present case, the 2.5 cm AML, in which a rich vascular component protruded into the perirenal space, caused a life-threatening retroperitoneal hemorrhage. Our literature review found no evidence that AML protruding from the kidney is prone to form aneurysms. However, like HCC, AMLs with a rich vascular component localized to the kidney surface may be prone to a life-threatening rupture, and such lesions should be targeted for careful follow-up and prophylactic treatment.

Emergency TAE was performed to address the ruptured aneurysm and surrounding vascular components for hemostasis rather than for the AML. Following emergency TAE, the patient underwent follow-up CT imaging at our hospital for 3 months. Subsequently, follow-up with ultrasonography and CT was continued at another hospital. After 5 years since embolization, the development of vascular components or new intratumoral aneurysms has not been observed. However, the clinicians monitoring the patient should be aware of the possibility of re-rupture if vascular lesions of the tu-

mor re-develop.

In conclusion, clinicians should be alert to tumor rupture when patients with AMLs complain of sudden abdominal pain, even if the tumor is small or an intramural aneurysm > 5 mm is unidentified during follow-up. AMLs with a rich vascular component at the surface of the kidney may require careful follow-up because these are prone to developing intratumoral aneurysms, leading to a life-threatening rupture. Further research is required to clarify the risk factors for rupture and vascular changes in renal AML.

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Author Contribution: M.T. was mainly involved in drafting

the manuscript and creating the figures. T.G. was involved in the intervention of the case and in drafting the manuscript. K.S., E.U., and T.O. were involved in drafting the manuscript. M.Y., K.S., and T.M. critically revised the entire manuscript. All authors reviewed the manuscript.

Informed Consent: Informed consent was obtained from the patient to submit this case report.

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