

Case Report

Postembolization Intratumoral Chronic Bleeding, without the Classic CT Feature of Active Extravasation, in Tuberous Sclerosis Complex-Related Renal Angiomyolipoma: Two Case Reports

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Keywords

Bleeding · Renal angiomyolipoma · Transcatheter arterial embolization · Tuberous sclerosis complex

Abstract

Two patients with tuberous sclerosis complex each had multiple bilateral renal angiomyolipomas. After undergoing embolization for a ruptured angiomyolipoma, patient 1 experienced long-lasting abdominal fullness; contrast-enhanced computed tomography (CECT) revealed a large chronic hematoma without contrast extravasation. Patient 2 underwent embolization for the largest right renal angiomyolipoma which contained a chronic hematoma. 2 weeks later, the symptom of abdominal fullness presented, and CECT revealed that the preexisting hematoma had enlarged without contrast extravasation. In both cases, a second embolization of the angiomyolipomas resulted in shrinking of the intratumoral hematomas and alleviation of the associated symptoms. Therefore, chronic postembolization intratumoral bleeding from renal

angiomyolipoma may present as a persistently large or growing hematoma with an associated symptom of abdominal fullness but without the typical CECT feature of active extravasation.

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Introduction

Renal angiomyolipomas (AMLs) typically comprise disorganized smooth muscle cells, adipose tissue, and abnormal vascular structures [1]. Approximately 95% of them can be specifically diagnosed based on fat tissue in the tumor detected by computed tomography (CT) or magnetic resonance imaging [2]. Tuberous sclerosis complex (TSC) is an autosomal dominant genetic syndrome with an estimated prevalence ranging from 1 in 6,000 to 1 in 12,000, and about 20% of patients with renal AMLs have TSC [3]. The blood vessels within AMLs are rigid, tortuous, aneurismal, and prone to rupture [1]. Renal AMLs with pseudoaneurysms larger than 5 mm are considered to be at high risk of bleeding [4]. Selective transcatheter arterial embolization (TAE) is the first-line treatment option in cases of acute bleeding, with the advantages of preserving normal renal parenchyma and avoiding surgery [5]. Extravasation of contrast agent visible on imaging can confirm the diagnosis and localization of active bleeding [6], enabling selective TAE to be performed in a timely fashion. This report presents two cases with TSC-related renal AMLs in which chronic bleeding after embolization continued without active extravasation.

Case Reports

Case 1

A 45-year-old woman with TSC had bilateral renal AMLs. She had undergone urgent TAE for a bleeding large left AML at another hospital. Although the ruptured pseudoaneurysm and its parent feeder were selectively occluded by coils, two other arteries feeding the same tumor were not included in the initial procedure. Mammalian target of rapamycin (mTOR) inhibitor was not used after the initial embolization.

Three months after the initial embolization, this patient continued to experience abdominal fullness and was referred to the authors' department. She had stable vital signs, a normal hemoglobin level of 12.3 g/dL, a normal serum creatinine level of 0.7 mg/dL, and an estimated glomerular filtration rate (eGFR) of 71 mL/min/1.73 m². Abdominal nonenhanced CT (NECT) and contrast-enhanced CT (CECT) showed that both kidneys had multiple fat-containing AMLs. The largest AML (11 cm in longest diameter) in the right kidney contained a pseudoaneurysm of 6 mm in diameter. In the left kidney, the largest AML contained a pseudoaneurysm (4 mm in diameter) and an intratumoral hematoma (volume, 503 mL) caused by the previous rupture, with the total size of the target AML and hematoma being 15 cm in longest diameter. On CECT, the hematoma was seen to be compressing the transverse and descending colon, thus explaining the patient's symptom of abdominal fullness; however, no active extravasation was detected (Fig. 1a). Hematoma volume was calculated with volume analyzer Synapse Vincent software (Fujifilm, Tokyo, Japan). Because neither CECT nor laboratory examinations indicated significant acute bleeding, an elective second TAE was scheduled for both kidneys.

The patient's symptom of abdominal fullness did not diminish over time. The second TAE was performed 6 months after the initial embolization. Selective left renal angiography revealed that the target tumor had two previously untreated feeding arteries and one pseudoaneurysm of 4 mm in diameter (Fig. 1b). Subsequent superselective angiography for those feeding arteries was performed with the use of a microcatheter (outer diameter, 1.98 Fr; Masters Parkway Soft; Asahi Intecc, Nagoya, Japan); no active extravasation of contrast agent was observed. Multiporous gelatin sponge particles (1 mm, Gelpart; Nippon Kayaku, Tokyo, Japan) followed by detachable coils (diameter, 1.5–2 mm; length, 3–4 cm; Target Detachable Coils; Stryker Corp., Kalamazoo, MI, USA) were injected to occlude the pseudoaneurysm and feeding arteries of the target AML. After embolization, confirmatory left renal angiography revealed no contrast-enhanced vessels within the target tumor (Fig. 1c). Subsequently, we performed a successful prophylactic embolization of the largest AML in the right kidney during the same procedure.

Within a few days after the second embolization, the patient's symptom of abdominal fullness disappeared. Follow-up CT examinations at 1, 6, and 26 months showed that the chronic hematoma within the left target AML had shrunk (volumes of 280, 90, and 10 mL, respectively), alleviating the compression of the transverse and descending colon. Thirty-one months after the second TAE, the serum creatinine was 0.63 mg/dL and the eGFR was 78 mL/min/1.73 m², indicating that renal function had not worsened.

Case 2

A 24-year-old woman with TSC had bilateral renal AMLs. About 8 years previously, she had experienced right renal AML bleeding, which resolved with conservative treatment. She came to our department for potential embolotherapy of renal AMLs. She had normal vital signs, a hemoglobin level of 10.0 g/dL, a normal serum creatinine level of 0.4 mg/dL, and an eGFR of 157 mL/min/1.73 m².

Abdominal NECT and CECT examination showed that both kidneys had numerous fat-containing AMLs with multiple pseudoaneurysms larger than 5 mm. The largest AML in the right kidney was about 23 cm in longest diameter and contained an intratumoral chronic hematoma (volume, 200 mL) without active extravasation (Fig. 2a). Compression of the transverse and ascending colon by the largest AML was detected on CT (Fig. 2a), but the patient did not have associated abdominal symptoms at that time.

Primary TAE was performed for the right kidney because of the chronic intratumoral hematoma. On selective right renal angiography, the multiple AMLs presented as diffuse abnormal tortuous vessels without identifiable tumor margins, and pseudoaneurysms larger than 5 mm in diameter (the largest one, 7 mm) and a predominant portion of tumoral vessels were located at the lower pole (Fig. 2b). Superselective angiography for the lower branches of the renal artery was performed, and the target tumoral vessels and aneurysms were occluded by 1-mm multiporous gelatin sponge particles (Gelpart; Nippon Kayaku) and detachable coils (diameter, 1–3 mm; length, 3–6 cm; Target Detachable Coils; Stryker). Confirmatory right renal angiography showed that most of the abnormal vessels and all of the pseudoaneurysms located in the lower pole had disappeared.

After the initial embolization, mTOR inhibitor therapy was not performed. Two weeks after the initial TAE, the patient complained of abdominal fullness, and laboratory examination showed a decreased hemoglobin level of 7.7 g/dL. Abdominal NECT and CECT showed that the preexisting intratumoral hematoma in the right kidney had grown (volume, 560 mL), leading to increased compression of the transverse and ascending colon (Fig. 2c). Although

CECT imaging revealed no active extravasation of contrast agent (Fig. 2c), the enlarged hematoma indicated significant chronic bleeding from the right renal AMLs. Therefore, a second embolization was performed for the right kidney on the same day. On right renal angiography, the absence of active contrast extravasation made the bleeding site undetectable. The tumoral vessels in the middle portion of the kidney, which had the largest pseudoaneurysm (4 mm in diameter) (Fig. 2d) and the largest feeding artery at that time, were highly suspected as the source of bleeding and were successfully embolized by using the same embolic agents as in the previous TAE (Fig. 2e).

The patient's symptom of abdominal fullness disappeared within a few days after the second embolization. Follow-up CT images at 17 days showed that the right intratumoral hematoma had decreased to 110 mL and that compression of the transverse and ascending colon had diminished. At 3 months, laboratory examination showed a hemoglobin level of 10.3 g/dL, the intratumoral hematoma had disappeared on CT, and a prophylactic TAE was performed on the left large AMLs that had pseudoaneurysm(s) larger than 5 mm. Renal function of the patient was normal throughout the postembolization period.

Both patients experienced postembolization syndrome (grade 2; CTCAE ver. 4.03) [7] including fever and pain in the back and flank, but no other complications occurred.

Discussion

In each of the current cases, after the initial TAE, the renal intratumoral hematoma remained large or grew, and the associated symptom of abdominal fullness was long-lasting or newly emergent; after the second TAE, the hematoma shrank dramatically, leading to quick resolution of the associated symptom and indicating that intratumoral chronic bleeding was present until the second procedure. In each case, because the second TAE was performed on previously untreated feeders, tumoral vessels not included in the initial embolization were causative for the postembolization chronic bleeding.

The typical CT feature of active contrast extravasation within hematomas is an important indicator of active bleeding [8]. However, because the presence of active extravasation is associated with several factors, including pressure difference (blood pressure, potential space), the small size of the disruption in the vessel wall (extent of injury), and the viscosity of the blood (anemia) [6], bleeding from renal AMLs does not always result in active extravasation appearing on CECT. One previous study demonstrated that CECT identified active extravasation in 36% of ruptured renal AML cases, and angiography identified active extravasation in 44% of cases [9]. In addition, CT featuring a rapid development of hematoma can suggest the presence of active bleeding [8], and it is easy to identify initial bleeding from renal AMLs by detecting new collections of blood on CT even without active extravasation. Nevertheless, when chronic bleeding from a renal AML occurs in the context of a preexisting hematoma and presents no active extravasation on CECT, it may be hidden by the previous hematoma and need more careful monitoring. In the two current cases, the only sign of chronic bleeding from postembolization renal AML was a persistently large or growing hematoma on CECT imaging, with no active extravasation of contrast agents.

Because the kidneys are located in the retroperitoneal space, it is uncommon for a renal hematoma to cause intestinal symptoms. In two previously reported cases, serious intestinal complications (duodenal obstruction) were caused by renal hematoma due to a bleeding renal AML or severe blunt renal trauma and were ameliorated after image-guided drainage of the hematoma [10, 11]. In the two cases reported here, the renal chronic hematomas were each

located within a large AML that allowed compression of the intestines; the persistent large hematoma (in case 1) or the growing hematoma (in case 2) compressed the intestines and caused the patients' long-lasting or newly emergent symptoms of abdominal fullness.

Renal AMLs associated with TSC are usually multiple and bilateral, and in these cases, preservation of the renal parenchyma is essential during the treatment of bleeding from AML. As a safe and minimally invasive procedure, superselective TAE of renal AML can preserve normal renal parenchyma as much as possible and can be performed multiple times [5, 12]. In fact, in each case reported here, despite multiple TAEs, chronic bleeding from renal AML was treated with consequent amelioration of the associated patient symptom, contralateral renal AML(s) with pseudoaneurysm(s) >5 mm were prophylactically embolized, and there was no significant deterioration of renal function in the postembolization period and no other complication except postembolization syndrome.

Conclusion

For cases with TSC-related renal AMLs that have tumoral vessels untreated by previous embolization, when considering reasons for the symptom of abdominal fullness, the possibility of postembolization chronic intratumoral bleeding must be taken into account, and it may present as a persistently large or growing hematoma but without the typical CECT feature of active extravasation.

Statement of Ethics

This report was approved by the institutional review board (JHS 17–0029) and the requirement for informed consent was waived.

Disclosure Statement

The authors declare no conflict of interest.

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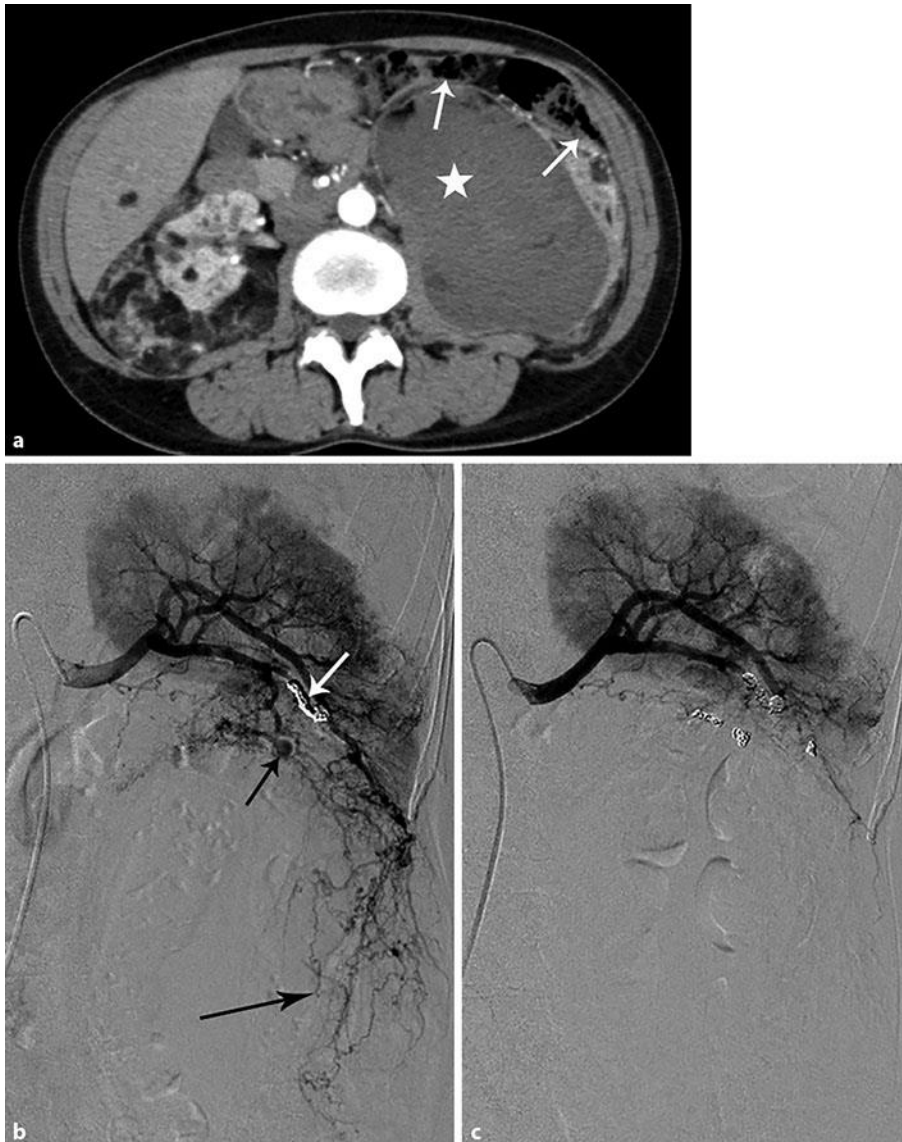


Fig. 1. **a** Axial CECT image obtained 3 months after emergency embolization of left renal AML. A large chronic intratumoral hematoma (star) without contrast extravasation is compressing the transverse and descending colon (arrows). **b** Left renal angiography before the second TAE shows abnormal tortuous vessels (long black arrow) and a pseudoaneurysm (short black arrow) which were not treated in the initial procedure. Coils (white arrow) used for the ruptured pseudoaneurysm and its parent artery during the initial TAE can also be seen. **c** Confirmatory left renal angiography after the second TAE. All abnormal vessels and the pseudoaneurysm of the target tumor have disappeared.

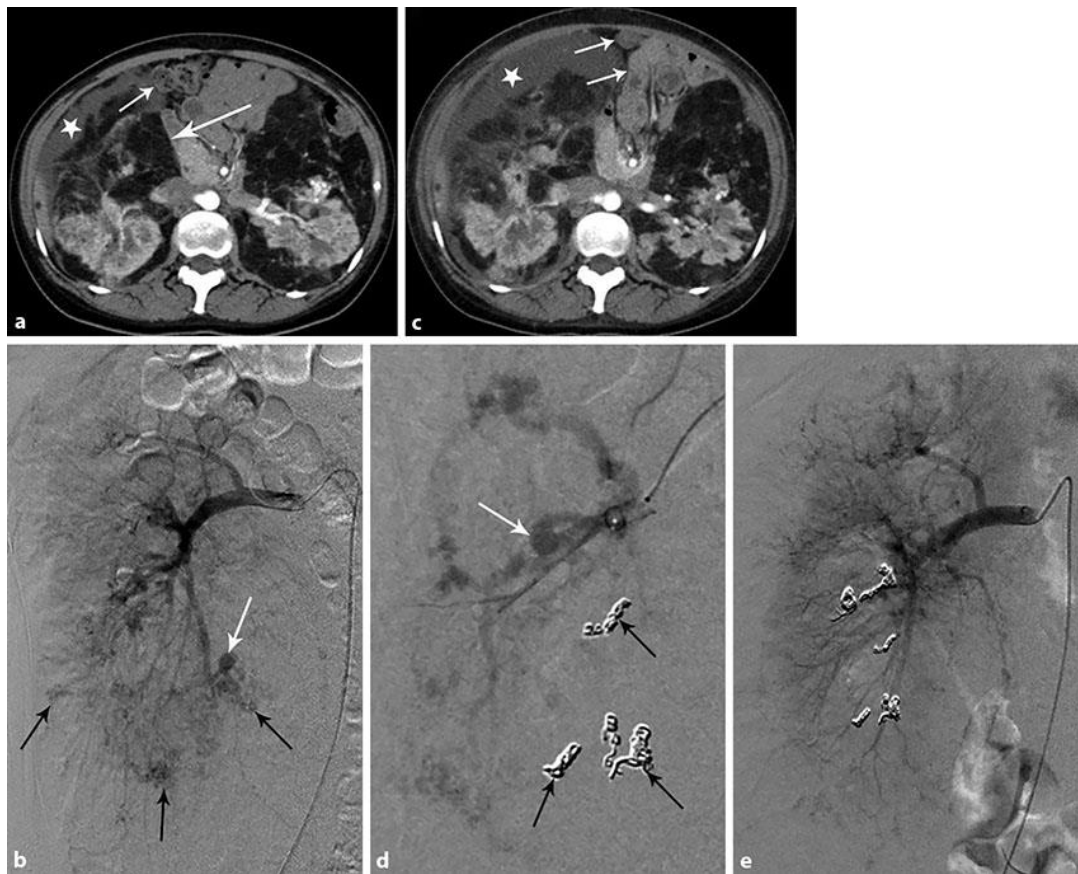


Fig. 2. **a** Axial CECT image obtained before the initial TAE. The largest AML (long white arrow) in the right kidney with an intratumoral chronic hematoma (star) is compressing the transverse and ascending colon (short white arrow). **b** Right renal angiography before the initial TAE. Right renal AMLs present as diffuse tortuous vessels without identifiable tumor margins. A predominant portion of abnormal vessels (black arrows) and the largest pseudoaneurysm (white arrow) are located at the lower pole. **c** Axial CECT image obtained 2 weeks after the initial TAE. The chronic hematoma (star) has enlarged, increasing the compression of the transverse and ascending colon (arrows). No contrast extravasation within the hematoma can be seen. **d** Superselective angiography of one major feeder located in the middle portion of the kidney, performed before the second TAE. It shows the largest pseudoaneurysm (4 mm in diameter, white arrow), from which there is no extravasation of contrast agent. Coils (black arrows) used during initial TAE for pseudoaneurysms and tumoral vessels located in the lower pole can be seen. **e** Confirmatory right renal angiography after the second TAE. Most tumoral vessels and all aneurysms located in the middle and lower portions of the right kidney have disappeared.