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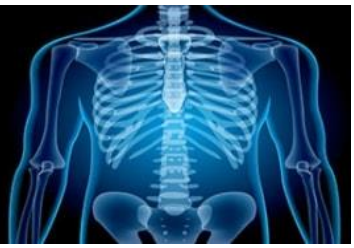

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A rare cause of retroperitoneal bleeding: A case report of a ruptured renal angiomyolipoma treated with Transarterial Embolization (TAE) in Tanzania

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Abstract
Renal angiomyolipoma is a rare, typically benign tumor with distinct radiological features and is often associated with Tuberous Sclerosis Complex. These tumors can develop both micro- and macro-aneurysms, which have the potential to rupture, leading to retroperitoneal bleeding. We herein present the case of a 39-year-old female with Tuberous Sclerosis Complex who presented with acute right-sided flank pain, bilateral tender flank masses, chest pain, a productive cough, and facial angiofibromas. Abdominal CT revealed retroperitoneal hemorrhage on the right side, along with bilateral angiomyolipomas. High-resolution CT of the chest showed features of lymphangioleiomyomatosis, while a brain CT revealed subependymal hamartomas of varying sizes. Multiple microaneurysms (<4 cm) were identified in both kidneys, with a single macroaneurysm (>4 cm) in the right kidney being the source of the retroperitoneal bleeding. The patient underwent transarterial embolization using Polyvinyl Alcohol particles and embolization coils. This case underscores the importance of considering prophylactic embolization in patients with giant angiomyolipomas and aneurysms larger than 4 cm to prevent potential rupture and its associated complications.

Keywords: Angiomyolipoma, tuberous sclerosis complex, retroperitoneal bleeding, aneurysm, Transarterial Embolization (TAE)

Introduction
Renal angiomyolipoma (AML) is rare tumor that, in most cases, follows a benign course and has clearly defined radiological characteristics. AML is associated with Tuberous sclerosis in 20% of cases. AMLs can develop micro and macro-aneurysms that can rupture and result in retroperitoneal bleeding. The frequency of symptoms and the risk of bleeding due to rupture increases with the size of AML. In cases of acute bleeding Transarterial embolization (TAE) can be considered as a first line option [1-3].
We herein present a case of a 39-year-old female patient with Tuberous sclerosis complex who presented with ruptured right angiomyolipoma and underwent Transarterial embolization (TAE) using PVA particles and pushable coils.

Case Report
A 39-year-old woman presented with a sudden onset of right-sided flank pain. She maintained normal urine output without any signs of hematuria or lower limb swelling. Additionally, she experienced chest tightness, which worsened in cold conditions and was accompanied by a cough producing whitish sputum. However, she did not report paroxysmal nocturnal dyspnea.
Her medical history was unremarkable for loss of consciousness, seizures, nausea, vomiting, fever, weight loss, or ocular issues. A musculoskeletal examination revealed no abnormalities.
On the general examination, the patient was stable. Notably, she had multiple small, erythematous papules symmetrically distributed across the central face, consistent with facial angiofibromas (Figure 1). Bilateral, palpable, tender flank masses were also observed.
Laboratory investigations revealed normocytic, normochromic anemia with a hemoglobin level of 10.49 g/dL. There was an increased white blood cell (WBC) count of 1.55,

accompanied by neutrophilia. Coagulation studies, including prothrombin time (PT) and partial thromboplastin time (PTT), were within normal limits. Renal function markers, including urea and creatinine levels, were also within the normal range.

A CT scan of the abdomen (Fig 2) showed a right-sided retroperitoneal hemorrhage along with bilaterally enlarged, heterogeneously enhancing, and distorted kidneys. The kidneys had a mixture of soft tissue and fat attenuation, with multiple cysts and mass effect on adjacent organs. However, no evidence of soft tissue infiltration was observed. The right kidney measured 13.43 × 11.50 × 9.36 cm, while the left kidney measured 10.0 × 10.9 × 8.3 cm. Based on these findings, a diagnosis of bilateral angiomyolipoma with right retroperitoneal hemorrhage was established.

High-resolution CT (HRCT) of the lungs (Fig 3) revealed findings consistent with lymphangioleiomyomatosis (LAM). The imaging showed diffuse bilateral thin-walled cystic lesions of varying sizes, with some coalescing into larger cysts, while the intervening lung parenchyma appeared normal.

A CT scan of the brain (Fig 4) demonstrated multiple bilateral subependymal and right basal ganglia calcific foci. These findings were consistent with subependymal hamartomas of varying sizes.

Echocardiography findings indicated normal cardiac function and structure.

The patient was referred to the Interventional Radiology (IR) department for Transarterial embolization (TAE).

A catheter was introduced via the right external iliac artery, advanced through the aorta, and navigated into the right renal artery. Once the catheter was in place, angiography was used to visualize the blood vessels feeding the angiomyolipomas (Fig 5). Multiple microaneurysms were identified, with a single macroaneurysm greater than 4 cm confirmed as the source of retroperitoneal bleeding. A mixture of polyvinyl alcohol (PVA) particles was injected to partially occlude the vessel, followed by the deployment of embolization coils to completely block blood flow to the affected area. The coils were strategically placed within the aneurysm, effectively halting the bleeding. The procedure was successfully completed, achieving the desired result of vessel occlusion and bleeding control.

A contrast-enhanced CT scan of the abdomen, performed one month after TAE, confirmed the presence of the embolization coil in the right kidney with no signs of retroperitoneal hemorrhage (Fig. 6). No biopsy was performed.

The patient is currently doing well, with no recurrence of right flank pain, and continues to attend follow-up clinics.

Discussion

Renal angiomyolipoma is a benign mesenchymal tumor that originates from perivascular epithelioid cells. It consists of a mix of fat, smooth muscle, and blood vessels. This type of tumor accounts for 1-3% of solid renal tumors, with a prevalence of approximately 13 per 10,000 adults [1, 4]. Most angiomyolipomas are sporadic, making up 80% of cases, while the remaining 20% are linked to phakomatoses, with the majority occurring in individuals with tuberous sclerosis [5].

Angiomyolipomas are typically asymptomatic and are often found alongside cysts, and sometimes renal cell carcinoma. They are rarely diagnosed in patients without tuberous sclerosis before puberty. Larger angiomyolipomas are more common in women than men and may grow rapidly during pregnancy, suggesting that hormones could influence their growth [4]. Bilateral renal angiomyolipoma in tuberous sclerosis, as seen in this case, is an uncommon occurrence, with a nearly equal prevalence between females and males [6].

According to documented case reports, the typical clinical presentation of angiomyolipomas includes a palpable, tender mass, flank pain, and gross hematuria, collectively referred to as Lenk's triad [7]. Less common symptoms associated with angiomyolipomas include nausea, vomiting, fever, anemia, and changes in blood pressure [8]. In this case, the patient did not present with heamaturia.

Renal angiomyolipomas (AMLs) develop in approximately 80% of individuals with Tuberous Sclerosis Complex (TSC), a genetic disorder inherited in an autosomal dominant manner. TSC is a multisystem condition characterized by the presence of benign tumors in various organs, including the skin, brain, kidneys, and lungs, with occasional malignant growths. Central nervous system (CNS) involvement is common, affecting about 85% of patients, typically manifesting in childhood. These neurological complications often lead to treatment-resistant epilepsy, intellectual disabilities, autism spectrum disorder, attention deficit hyperactivity disorder (ADHD), and behavioral challenges. In adults with TSC, kidney disease is the primary cause of mortality. Given the complexity and variability of TSC, comprehensive and well-coordinated medical management is essential [9].

Lymphangioleiomyomatosis (LAM) is a rare cystic lung disease that can develop due to mutations in the tuberous sclerosis genes or occur sporadically [10]. In the case of our patient, although brain subependymal harmatomas were present, there were no childhood CNS manifestations noted.

The most frequent serious complication of renal angiomyolipoma (AML) is hemorrhage. Due to the dense neovascularization within AMLs, there's a risk of spontaneous rupture, leading to bleeding. Large AMLs may develop micro- or macro-aneurysms, which can rupture unexpectedly, often causing sudden, severe pain and, in some cases, becoming life-threatening. Around 10% of cases present with hypovolemic shock caused by extensive retroperitoneal bleeding, a condition known as Wunderlich's syndrome [4, 11]. However, the patient in this report did not present with hypovolemic shock.

Rupture can happen spontaneously in patients taking anticoagulants or because of kidney trauma. It has also been documented during pregnancy or after childbirth [4].

Nephrectomy or nephron-sparing surgery is rarely the first-line treatment in urgent cases. Instead, urgent transarterial embolization (TAE) is a minimally invasive and technically viable procedure for managing severe hemorrhage while preserving renal function after a spontaneous rupture of renal AML. Embolization is performed using polyvinyl alcohol (PVA) particles and coils of various sizes [3]. Partial or radical nephrectomy is recommended when there is ongoing bleeding, suspected malignancy, or unsuccessful embolization. In cases involving bilateral lesions, such as in tuberous sclerosis (TS), nephron-sparing surgery, which can include selective embolization or partial nephrectomy (performed either open, laparoscopic, or robotic), should be carried out [2].

The likelihood of symptoms and the risk of bleeding

(rupture) rise with the size of an angiomyolipoma (AML). Treatment decisions for AMLs are based on the presence of symptoms. In most cases, patients with small tumors (less than 4 cm) that are typically asymptomatic are managed conservatively with regular follow-up through ultrasonography [12]. Tumors larger than 8 cm are usually symptomatic and have a higher risk of spontaneous or traumatic rupture, leading to hemorrhagic complications. As a result, these patients are typically treated with angiography and selective arterial embolization as the first-line approach [13].

Prophylactic embolization is recommended for asymptomatic lesions 4 cm or larger in certain high-risk patients, such as younger women planning future pregnancies or those for whom regular follow-up is challenging. Embolization helps reduce the risk of hemorrhage by cutting off the blood supply to the AML. The tumor size typically decreases by up to 40%, with reductions of about two-thirds. Tumor recurrence has been reported, and in some cases, repeated embolizations may be necessary [8].

AMLs can range in size from a few millimeters to over 20 cm, with those larger than 10 cm classified as "giant" AMLs. Since AMLs larger than 10 cm are uncommon, CT imaging plays a crucial role in distinguishing AMLs from other tumors, such as perinephric liposarcomas [14].

In digital subtraction angiography (DSA), angiomyolipomas are hypervascular lesions that typically exhibit distinct features. During the arterial phase, they appear as sharply defined, hypervascular masses with a dense early arterial network and tortuous vessels, creating a "sunburst" pattern, as shown in figure 6. In the venous phase, the peripheral vessels may display a whorled, "onion peel" appearance. Additionally, micro- or macro-aneurysms may be present, but there is no arteriovenous shunting observed [5].

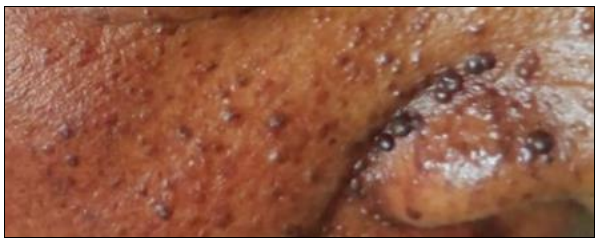

Fig 1: Facial angiofibroma as seen in the patient.

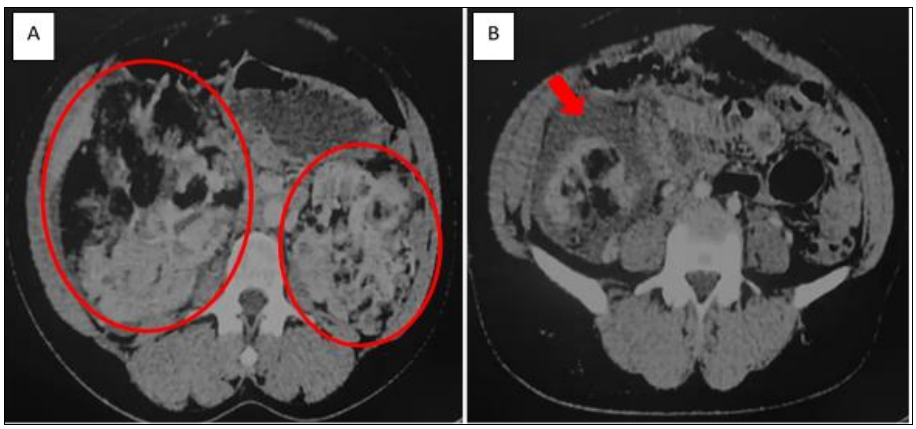

Fig 2: Contrasted Abdominal CT (Axial view) shows A. Bilateral Renal angiomyolipomas (red circles) B. Right retroperitoneal hemorrhage noted by the (red arrow).

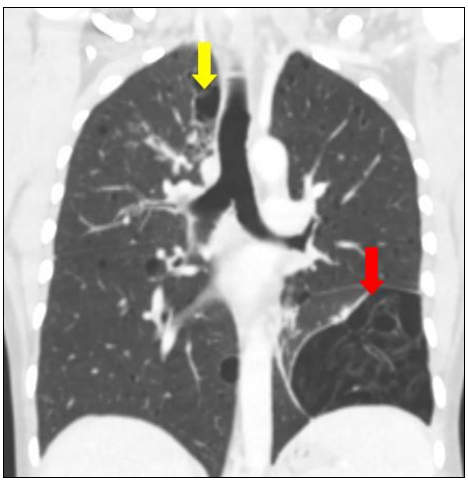

Fig 3: HRCT (coronal view) shows diffuse bilateral thin-walled cystic lesions of varying sizes (yellow arrow), some of which have coalesced into larger cyst at the left lower lobe (red arrow), with normal intervening lung parenchyma.

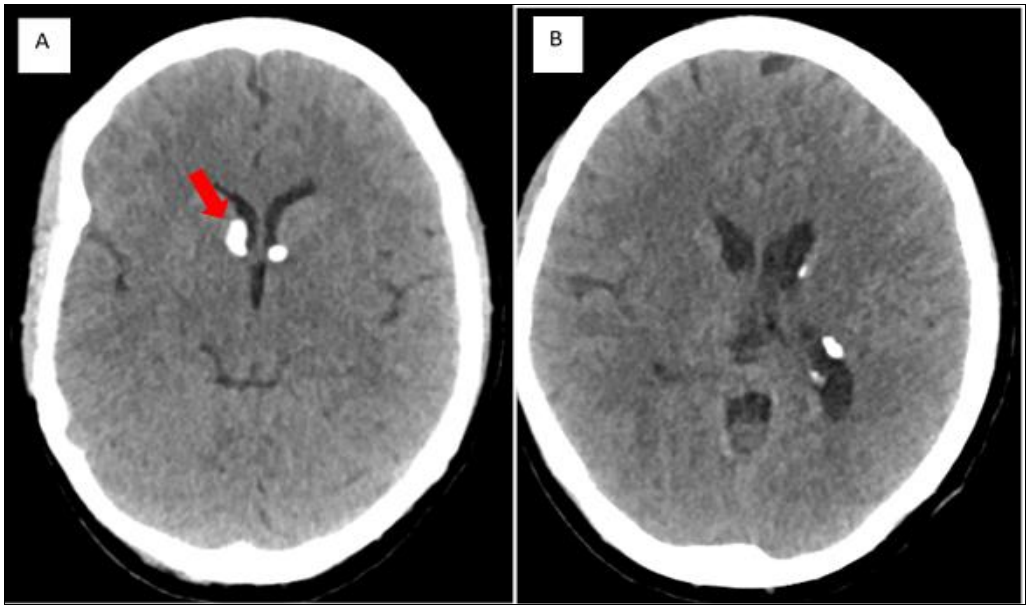

Fig 4: Non contrasted CT brain, axial view (A and B) shows multiple bilateral subependymal and right basal ganglia calcific foci (red arrow) in keeping with subependymal harmatomas of varying sizes.

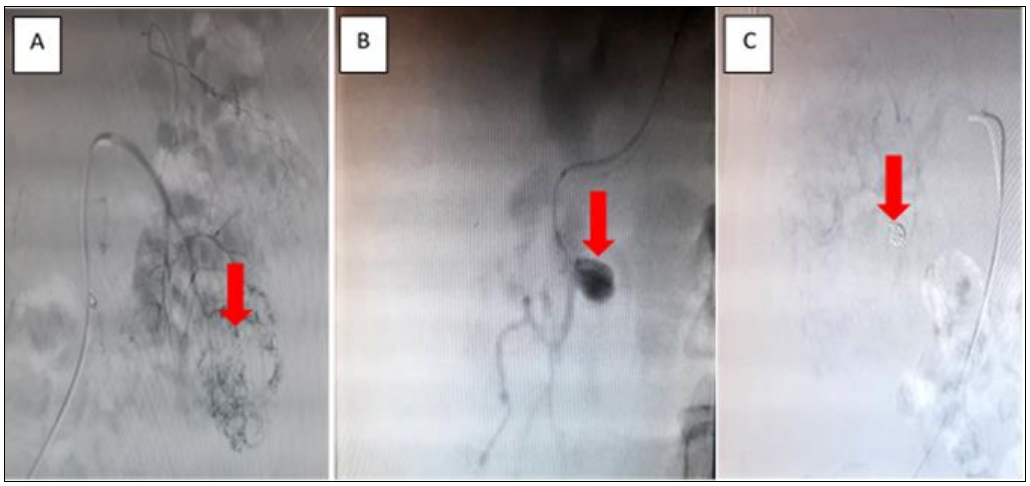

Fig 5: Digital subtraction angiography shows A- a hypervascular mass with a dense arterial network and tortuous vessels giving the "sunburst" appearance in the left kidney with multiple microaneurysm (<4cm) shown in red. B-Macroaneurysm(>4cm) noted in the right kidney (red arrow) C-Embolization coil in the microaneurysm noted in the right kidney (red arrow).

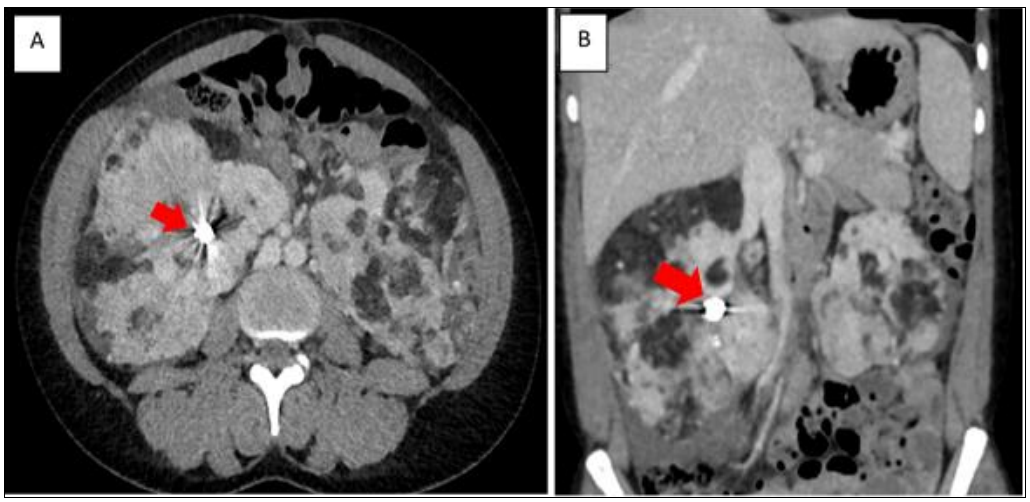

Fig 6: Contrasted abdominal CT A (Axial view) and B (coronal view) shows Bilateral Renal angiomyolipomas with embolization coil in the right kidney.

Conclusion

Prophylactic embolization should be considered in patients with giant angiomyolipomas (AML) and aneurysms greater than 4 cm to prevent the risk of rupture. Additionally, tuberous sclerosis should always be suspected in any patient presenting with angiomyolipomas, as it is a key condition associated with these tumors. A multidisciplinary approach in the diagnosis and management of AMLs is crucial, as it facilitates a timely and accurate diagnosis, ensuring that appropriate treatment strategies are implemented effectively to improve patient outcomes.

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References

1. Bouaziz H, Ghalleb M, Tounsi N, *et al*. A renal angiomyolipoma with a challenging presentation: a case report. J Med Case Rep. 2021;15:477. DOI:10.1186/s13256-021-03073-0.
2. Faddegon S, So A. Treatment of angiomyolipoma at a tertiary care centre: the decision between surgery and angioembolization. Can Urol Assoc J [Internet]. 2011 [cited 2022 Jul 9];5(6). Available from: https://pubmed.ncbi.nlm.nih.gov/21388589/.
3. Duan XH, Zhang MF, Ren JZ, Han XW, Chen PF, Zhang K, *et al*. Urgent transcatheter arterial embolization for the treatment of ruptured renal angiomyolipoma with spontaneous hemorrhage. Acta Radiol. 2016 Nov;57(11):1360-1365. DOI:10.1177/0284185115588125.
4. Eble JN. Angiomyolipoma of kidney. Semin Diagn Pathol. 1998 Feb;15(1):21-40. PMID: 9503504.
5. Silverstone L, Balasubramanian S, Salehzadeh H, *et al*. Renal angiomyolipoma. Radiopaedia.org [Internet]. (Accessed on 13 Feb 2025). Available from: https://doi.org/10.53347/rID-899.
6. Bansal C, Lakshmaiah V, Raavesha A, Waadhawan P, Priyanka MK. Tuberous sclerosis with bilateral renal angiomyolipoma. J Assoc Physicians India. 2013 Jun;61(6):420-423. PMID: 24640214.
7. Guttilla A, Crestani A, Cattaneo F, Zattoni F, Valotto C, Iafrate M, *et al*. Wunderlich's syndrome: Three cases of acute spontaneous renal bleeding, conservatively treated. Arch Ital Urol Androl. 2013;85:210-3. DOI:10.4081/aiua.2013.4.210.
8. Chronopoulos PN, Kaisidis GN, Vaiopoulos CK, Perits DM, Varvarousis MN, Malioris AV, *et al*. Spontaneous rupture of a giant renal angiomyolipoma—Wunderlich's syndrome: Report of a case. Int J Surg Case Rep. 2016;19:140-3. DOI:10.1016/j.ijscr.2015.12.017.
9. Annear NMP, Appleton RE, Bassi Z, Bhatt R, Bolton PF, Crawford P, *et al*. Tuberous sclerosis complex (TSC): Expert recommendations for provision of coordinated care. Front Neurol. 2019 Nov 6;10:1116. DOI:10.3389/fneur.2019.01116.
10. Cooper A, Baugh L, Kelley S, Huang H, Guileyardo J. Pulmonary lymphangioleiomyomatosis associated with aggressive renal angiomyolipoma. Proc (Bayl Univ Med Cent). 2018 Jan 3;31(1):81-83. DOI:10.1080/08998280.2017.1391038.
11. Chen YC, Lin YC. Wunderlich syndrome. QJM. 2013 Feb;106(2):187-188. DOI:10.1093/qjmed/hcr266.
12. De Luca S, Terrone C, Rossetti SR. Management of renal angiomyolipoma: a report of 53 cases. BJU Int. 1999 Feb;83(3):215-218. DOI:10.1046/j.1464-410x.1999.00932.
13. Bora A, Soni A, Sainani N, Patkar D. Emergency embolization of a bleeding renal angiomyolipoma using polyvinyl alcohol particles. Diagn Interv Radiol. 2007 Dec;13(4):213-216. PMID: 18092296.
14. Schieda N, Kielar AZ, Al Dandan O, McInnes MD, Flood TA. Ten uncommon and unusual variants of renal angiomyolipoma (AML): radiologic-pathologic correlation. Clin Radiol. 2015 Feb;70(2):206-220. DOI:10.1016/j.crad.2014.10.001.

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