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Case Report

Giant renal pseudoaneurysm complicating angiomyolipoma in a patient with tuberous sclerosis complex: An unusual case report and review of the literature

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ABSTRACT

Introduction: Angiomyolipomas (AMLs) are uncommon benign lesions, which are composed of dysmorphic blood vessels, adipose tissue, and smooth muscle components. They tend to bleed because of the hypervascularity and the presence of small aneurysms, leading to life-threatening complications.
Presentation of case: A 31-year-old female was presented to the emergency service of our hospital, complaining of left flank pain for 1 week followed by hematuria for one day. Radiologic imaging showed the features of a giant renal pseudoaneurysm. Superselective embolization was applied and she had an uneventful recovery.
Discussion: The blood vessels in AML are tortuous and thick-walled with the absence of supportive elastic tissue, which tend to the formation of the intralesional pseudoaneurysm. The risk of bleeding is higher with tumors larger than 4 cm, rapid tumor growth, and aneurysms larger than 0.5 cm. Early detection and treatment are essential for the prevention of bleeding and improving patient outcomes.
Conclusion: Giant pseudoaneurysm in a renal angiomyolipoma associated with tuberous sclerosis complex is a rare entity, often leading to potentially life-threatening bleeding. Selective angioembolization is recommended as firstline therapy for bleeding AML and is increasingly used as a preventive treatment for AML at risk of bleeding. However, a high incidence of the recurrence requires caution and a close longtime follow-up. Surgical intervention is indicated if the hemorrhage is not responsive to embolization or if there is suspicion of malignancy.

1. Introduction

Angiomyolipomas (AMLs) are uncommon benign lesions that are found in less than 0.3% of the general population and account for approximately 3% of all renal tumors. These lesions are composed of dysmorphic blood vessels, adipose tissue, and smooth muscle components [1]. While 80% of angiomyolipomas are sporadic and most of them inconsequential, approximately 20% are associated with tuberous sclerosis complex (TSC) [2].

AMLs tend to bleed because of the hypervascularity and the presence of small aneurysms, leading to life-threatening complications [3,4]. Clinically, the tumor has a 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 [5].

Giant pseudoaneurysm in a renal angiomyolipoma associated with tuberous sclerosis complex is rare. Endovascular embolization may at times be required to stop the bleed or prophylactically to prevent chances of intralesional pseudoaneurysm rupture. Surgery may be indicated when vascular interventional facilities are not available [6].

The authors present here a rare case of a giant renal pseudoaneurysm complicating angiomyolipoma in a 31-year-old patient with tuberous sclerosis complex, successfully managed with angioembolization.

This work has been reported in line with the SCARE 2020 criteria [7].

2. Case presentation

A 31-year-old female was presented to the emergency service of our

Abbreviations: AMLs, Angiomyolipomas; TSC, Tuberous sclerosis complex.

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hospital, complaining of left flank pain for 1 week followed by hematuria for one day. She was a known case of tuberous sclerosis and bilateral renal angiomyolipomas that had angioembolization for the left kidney pseudoaneurysm complicated renal angiomyolipoma with bleeding and retroperitoneal hematoma 4 years ago. She did not give any history of food or drug allergy and psychosocial problems.

The laboratory analysis yielded hemoglobin (Hb): 8.7 g/dl (12–16), white blood cells: 9420/mm3 (4500–11000) with neutrophils 81.1%, Urea: 33 mg/dl, creatinine: 1, 26 mg/dl, prothrombin time: 21.4 sec, INR: 1, and platelets: 292000/mm3. In contrast-enhanced abdominal CT images, almost all bilateral Kidney parenchyma was replaced by multiple masses of fat density (−59 HU), consistent with angiomyolipomas [Fig. 1a–b]. A focal dilatation reaching about 26 mm in diameter in the lower pole of the left kidney with homogenous contrast blush on arterial phase, compatible with a pseudoaneurysm and evidence for active extravasation in its neighboring region was noted[Fig. 2]. The fluid of blood density was observed in the bladder, but no hematoma in the retroperitoneal region was detected.

The patient was referred to the interventional radiology unite for the emergency endovascular procedure and urgent angioembolization. The catheter was inserted with right femoral artery intervention to the distal abdominal aorta by a selective renal insertion and an angiogram was obtained. A pseudoaneurysm with active extravasation was observed in the accessory renal artery. Embolization was performed by superselective catheterization, using a microcatheter. The control angiogram showed complete embolization of the active bleeding focus [Fig. 3]. No intraprocedural complications were noted and the patient was transferred to the intensive care unit (ICU) for further management. After 24 hrs she was started to be followed in the service room. On postprocedural day 5, the patient had an uneventful recovery and was discharged from the hospital with control recommendations of abdominal ultrasound and nephrology outpatient observation after 3 months.

3. Discussion

Renal angiomyolipoma was first defined by Fischer in 1911, which predominantly affects females rather than males [8]. Some studies have suggested that AMLs can present in a wide age range from 19 to 93 years with overall prevalence being as high as 0, 3% [1,5]. They occur as sporadic, isolated entities in 80% of cases. The remaining 20% of AMLs develop in association with tuberous sclerosis complex (TSC) or pulmonary lymphangioleiomyomatosis [9]. Tuberous sclerosis complex (TSC) is a rare hereditary disease caused by mutations of the TSC1 or TSC2 genes that are related to the PI3K/AKT/mTOR signaling pathway. These mutations induce overactivation of mammalian target of rapamycin protein (mTOR), which in turn induces uncontrolled cell growth [10].

TSC related angiomyolipomas are often multiple, bilateral, and rapidly growing. The blood vessels in AML are tortuous and thick-walled with the absence of supportive elastic tissue, which tend to the formation of the intralesional pseudoaneurysm. The risk of bleeding is higher with tumors larger than 4 cm, rapid tumor growth, and aneurysms larger than 0.5 cm [3,6]. Our patient was a known case of the TSC with bilateral renal angiomyolipomas and a huge (2.6 cm) left kidney

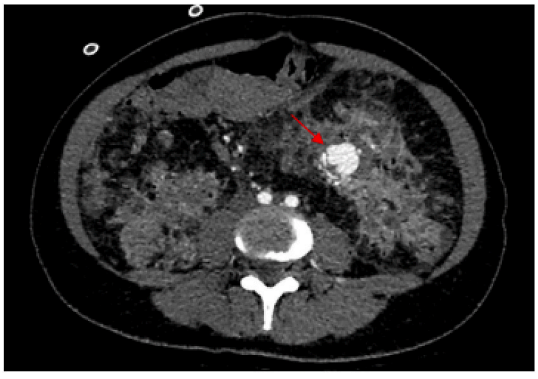

Fig. 1a. Axial contrast-enhanced CT angiogram showing bilateral renal multiple angiomyolipomas with left intra-renal pseudoaneurysm (arrow).

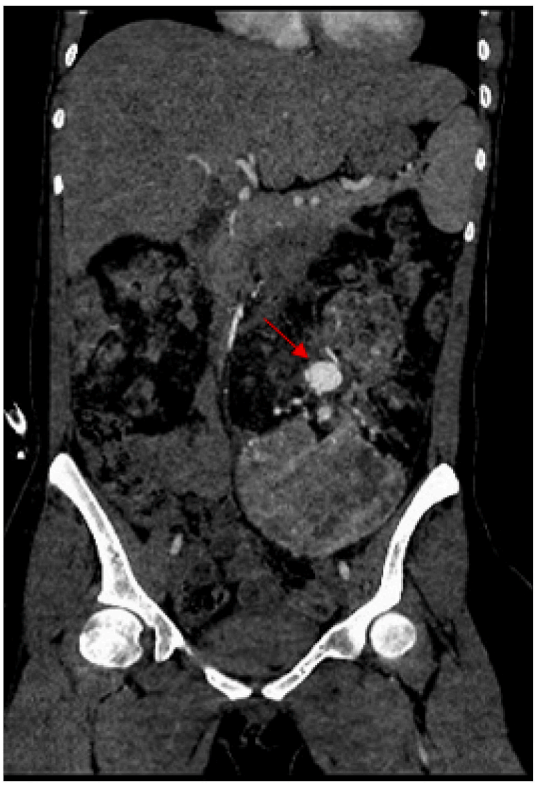

Fig. 1b. Coronal contrast-enhanced CT angiogram showing bilateral renal multiple angiomyolipomas with left intra-renal pseudoaneurysm (arrow).

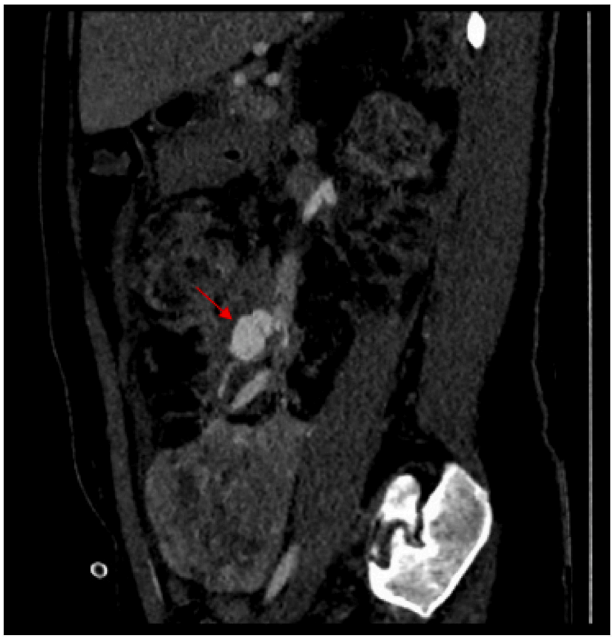

Fig. 2. Contrast-enhanced curved MPR CT angiogram, showing a giant left intra-renal pseudoaneurysm (arrow).

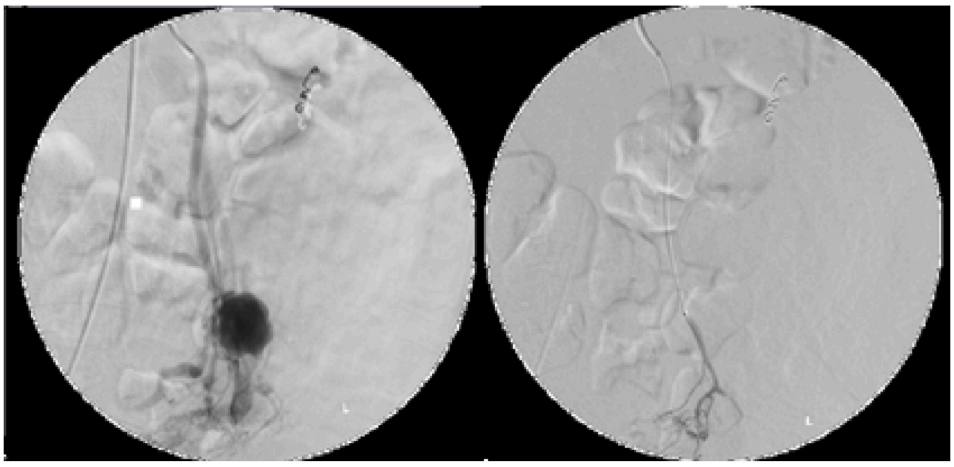

Fig. 3. Pre and post-embolization renal angiography images.

pseudoaneurysm.

The classic clinical triad of renal AML is flank pain, a palpable tender mass, and hematuria due to intracapsular or retroperitoneal bleeding [11].In our case, the patient had flank pain and hematuria without retroperitoneal bleeding, as documented by CT imaging before the endovascular treatment.

According to histopathology features, AML is classified into potentially malignant epithelioid AML and triphasic benign AML; the latter is further divided into classic AML and fat-poor AML. Classic AML is composed of 3 components: vessels, spindle cells, and adipose tissues. By contrast, fat-poor AML is defined by <25% fat per high power field under microscopy and does not contain enough fat to be detected with images [12].

Imaging plays a central role in the diagnosis and management of renal AMLs. The detection of adipose tissue is the fundamental diagnostic criterion of a classic AML. However, the fat-poor AML on imaging, making it harder to distinguish from renal cell carcinoma [9]. If the imaging techniques are not able to differentiate between benign and malignant masses, these atypical lesions should then be biopsied [10]. Thus, accurate preoperative diagnosis of renal AMLs is critical to prevent unnecessary nephrectomies and preserve renal functions [9]. Our patient had the classic AML as shown by CT imaging and there was no need for biopsy.

The most crucial aims of image checks are to identify and assess those signs associated with tumor ruptures such as an aneurysm or pseudoaneurysm formation. Therefore, radiologists and clinicians should always know what image features to expect before choosing the imaging techniques. Contrast-enhanced US can identify active bleeding or pseudoaneurysm formation and is a valuable real-time diagnostic workup for an abdominal emergency caused by AML. Besides that, using color-flow Doppler sonography, the blood flow of the tumor can be measured, and solid tumor, aneurysm, and pseudoaneurysm can be distinguished [12]. In contrast-enhanced CT images, renal AML appears as a circumscribed hypo-attenuating fatty mass (−30 HU to −70 HU). Associated perirenal hemorrhage is usually hyperdense with an attenuation of up to 60–70 HU [13]. Magnetic resonance imaging (MRI) has a high sensitivity for detecting fat tissues, so it is useful to differentiate AML from other renal masses. However, MRI has a high cost and is time-consuming when compared to ultrasonography and CT [8]. Angiography of the renal artery is a useful invasive method to provide obscure anatomical data and to treat the corresponding causes of bleeding [14].

Currently, renal angioembolization is recommended as first-line therapy for bleeding AML and is increasingly used as a preventive treatment for AML at risk of bleeding, but there are more recurrences and a need for secondary treatment than surgery [12,15]. Surgical intervention is indicated if the hemorrhage is not responsive to embolization or if there is suspicion of malignancy [16]. Microwave tumor ablation, radiofrequency, and cryoablation, are other possible treatments for small tumors. The only mTOR inhibitor approved for the treatment of renal angiomyolipoma is everolimus, which is indicated in the treatment of adult patients with TSC-associated angiomyolipoma who are at risk of complications but do not need immediate surgery [1,8,10]. In our case, the renal bleeding was successfully treated with superselective renal embolization with a favorable outcome.

Renal angiomyolipoma associated with TSC often results in substantial morbidity and mortality. Thus, proactive surveillance and monitoring of patients, beginning no later than the age of 10 years and continuing throughout the life with increasing frequency with advancing age, are recommended for all patients with TSC. Early detection and treatment are essential for the prevention of progressive damage to renal tissue and improving patient outcomes [17].

This is one of the documented cases, proved with imaging studies and managed by endovascular superselective angioembolization. However, lack of the longtime follow-up may be the only limitation for this case.

4. Conclusion

Giant pseudoaneurysm in a renal angiomyolipoma (AML) associated with tuberous sclerosis complex is a rare entity, often leading to potentially life-threatening bleeding. Early detection and treatment are essential for the prevention of bleeding and improving patient outcomes. Selective angioembolization is recommended as firstline therapy for bleeding AML and is increasingly used as a preventive treatment for AML at risk of bleeding. However, a high incidence of the recurrence requires caution and a close longtime follow-up. Surgical intervention is indicated if the hemorrhage is not responsive to embolization or if there is suspicion of malignancy.

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Ethics approval and consent to participate

The manuscript has got an ethical review exemption from the Ethical Review Committee (ERC) of our hospital as case reports are exempted from review according to the institutional ethical review committee's policy.

Consent for publication

Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Authors' contributions

Concept - HAE; Design – HAE; Supervision - MWN; Resources and data Collection- HAE; Literature Search - HAE; Writing Manuscript – HAE; Critical Review -.MWN. All authors have read and approved the final manuscript.

Research registration

Not applicable.

Guarantor

The corresponding author is the guarantor for the work and he has the responsibility of access to the data, and controlling the decision to publish.

Declaration of competing interest

The authors have no potential conflicts of interest to disclose.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.amsu.2021.01.014.

Provenance and peer review

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