CASE REPORT
XXX

Doi: 10.4274/jus.galenos.2023.2023-7-5
J Urol Surg

Treatment of Recurrent Giant Angiomyolipoma After Nephrectomy with Selective Arterial Embolization: A Case Report

Ali Nebioğlu[1], H. Erdal Doruk[1], Fadime Eda Gökalp Satıcı[2], Yasemin Yuyucu Karabulut[2], Caner Özer[3]

[1]Mersin University Faculty of Medicine, Department of Urology, Mersin, Turkiye
[2]Mersin University Faculty of Medicine, Department of Pathology, Mersin, Turkiye
[3]Mersin University Faculty of Medicine, Department of Radiology, Mersin, Turkiye

Abstract

Angiomyolipomas (AMLs) are mesenchymal tumors that typically originate from the kidneys and contain smooth muscle cells, fat cells, and blood vessels. They are usually benign in nature but can be fatal due to complications. AMLs almost always involve the kidneys, and only a few studies have reported the possibility of extrarenal involvement as a rare entity. In this study, we examined a patient who presented to our clinic with left-sided pain and was found to have a large AML measuring approximately 15 cm in size in the left kidney. The patient underwent a simple left nephrectomy, and AML was diagnosed after histopathological examination, with intact surgical margins. During the patient's routine 6-month follow-up visits, approximately four years after nephrectomy, a mass of approximately 13 cm was observed in the nephrectomy bed and radiologically interpreted as a recurrent AML. A tru-cut biopsy of the mass confirmed AML diagnosis. The patient was treated with selective arterial embolization, which was successful without any complications. We believe that our rare case of recurrent large AML will contribute to the diagnosis and treatment choices for patients with recurrent renal and extrarenal AML in the future and will add to the existing literature.

Keywords: Angiomyolipoma, nephrectomy, selective angioembolization

Introduction

Renal angiomyolipomas (AMLs) are benign mesenchymal tumors containing fat cells, smooth muscle cells, and blood vessels. AMLs constitute approximately 0.3-3% of all kidney tumors (1,2). Approximately 80% of AMLs are sporadic, while the remaining 20% are associated with tuberous sclerosis complex (3).

Although AMLs commonly exhibit renal localization, they can rarely present with extrarenal localization. Studies on extrarenal AMLs have shown their occurrence in various regions such as the liver, retroperitoneum, adrenal glands, colon, bladder, hilar lymph nodes, lungs, abdominal wall, fallopian tubes, hard palate, head, penis, spermatic cord, vagina, and uterus. The retroperitoneal space is the second most commonly observed site for extrarenal AMLs. Because of their ability to mimic other retroperitoneal tumors and the lower fat density of these lesions compared with renal AMLs, the diagnostic process through imaging can be more challenging for clinicians. Similar to renal AMLs, extrarenal retroperitoneal AMLs are more frequently observed in females (4).

AMLs were previously symptomatic (presenting with flank pain, palpable mass, hematuria, etc.) in approximately 64% of cases before the advent of radiological imaging, but nowadays they are mostly detected incidentally and asymptomatic (5,6).

Diagnosis of renal AMLs through radiological examinations such as computed tomography (CT), ultrasonography (USG), and magnetic resonance imaging is almost 100% sensitive. Lesions with negative attenuation values of 10 Hounsfield units or lower detected by CT, the presence of fat in the lesion, and the presence of a hyperechoic signal and acoustic shadowing on USG are diagnostic for AMLs (7).

The most important complication of AML is life-threatening retroperitoneal or collecting system bleeding, particularly in patients with large lesions (8). In a recent series, approximately 58% of patients required total or partial nephrectomy and

Correspondence: Ali Nebioğlu MD, Mersin University Faculty of Medicine, Department of Urology, Mersin, Turkiye
Phone: +90 507 973 68 04 **E-mail:** alinebioglu90@gmail.com **ORCID-ID:** orcid.org/0000-0001-6325-1534
Received: 22.07.2023 **Accepted:** 19.10.2023

Cite this article as: Nebioğlu A, Doruk HE, Gökalp Satıcı FE, Yuyucu Karabulut Y, Özer C. Treatment of Recurrent Giant Angiomyolipoma After Nephrectomy with Selective Arterial Embolization: A Case ReportJ Urol Surg

42% required embolization to control bleeding symptoms (2). One of the most significant complications of renal AMLs is the occurrence of massive retroperitoneal hemorrhage, often seen in pregnant individuals, following the rupture of an AML, a condition known as Wunderlich syndrome (9).

A wide range of treatment options are available for AML, ranging from active surveillance to total nephrectomy. Among these, the most commonly preferred treatments in recent years are selective arterial embolization and nephron-sparing surgical procedures (10-12). Although prophylactic treatments to prevent bleeding and interventions to treat active bleeding are effective and safe, preserving kidney function remains an important goal for new therapeutic approaches (13).

We present a rare case of AML that recurred in the nephrectomy bed after simple nephrectomy and was subsequently treated with selective arterial embolization.

Case Presentation

A 47-year-old female patient presented to our clinic with severe left flank pain. According to her medical history, four years ago, a large AML was detected in her left kidney, and a left simple nephrectomy was performed by our team. Pathological examination after surgery revealed AML, and the tumor was completely removed with intact surgical margins. The patient was invited to routine outpatient clinic follow-ups every 6 months because of having a solitary kidney in the postoperative period. During each visit, the patient underwent physical examination, renal function tests, and complete urinalysis. In addition, the aim was to detect any pathology in the solitary kidney, such as stones, masses, or pyelonephritis, to prevent renal failure due to preventable causes through early diagnosis. In routine follow-ups, the recurrent mass was detected by ultrasound in the second year of follow-up, with its size initially measured at 4 cm. Therefore, we decided to monitor the recurrent mass at regular intervals. Because of the absence of complications during the follow-up period and the patient's lack of request for intervention, despite an increase in the size of the mass, the patient was continued to be followed up for an additional 2 years. During the final physical examination conducted in the fourth year of follow-up, before embolization, the mass was palpable in the left costovertebral area, and imaging studies revealed that it had reached a length of 13 cm. Laboratory tests showed normal serum creatinine and hemogram values, but 26 erythrocytes were found in the complete urinalysis. Multiphasic renal CT was requested for the patient. In multiphasic renal CT, the left kidney was observed in an operated appearance, and a mass of 131x99x137 mm in size, macroscopic fat and soft tissue density, with prominent vascularity and compatible with AML was observed in the left renal fossa (Figure 1).

A true-cut biopsy was performed for definitive diagnosis of this recurrent lesion, and histopathological examination revealed a tumor lesion compatible with AML, containing a rich lipomatous component and thick-walled hyalinized vascular structures. The case was evaluated as recurrent AML (Figure 2a,2b).

After the patient was diagnosed, the interventional radiology department was consulted for treatment purposes, and the patient underwent selective arterial embolization (Figure 3a,3b). There were no complications during or after the procedure. The patient was discharged in a healthy condition on the first postoperative day after undergoing AML embolization. For follow-up purposes, the patient was invited to the urology

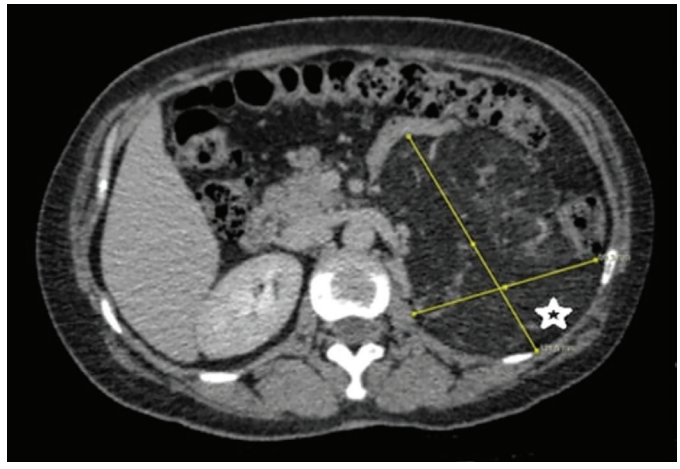

Figure 1. Late-phase images of multiphase renal computed tomography examination; a mass consistent with angiomyolipoma with heterogeneous density, located in the left retroperitoneal area, measuring 131x99x137 mm, well-defined, lobulated contours, and containing dense fatty content (indicated with a star)

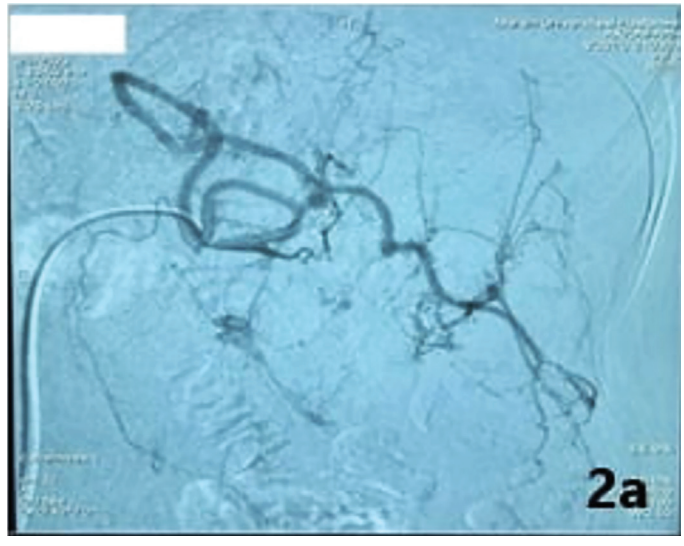

Figure 2a. Pre-embolization digital subtraction angiography image showing hypertrophic, tortuous, arterial structures supplying the angiomyolipoma consistent mass located in retroperitoneal localization on the left, and aneurysm formations within the mass

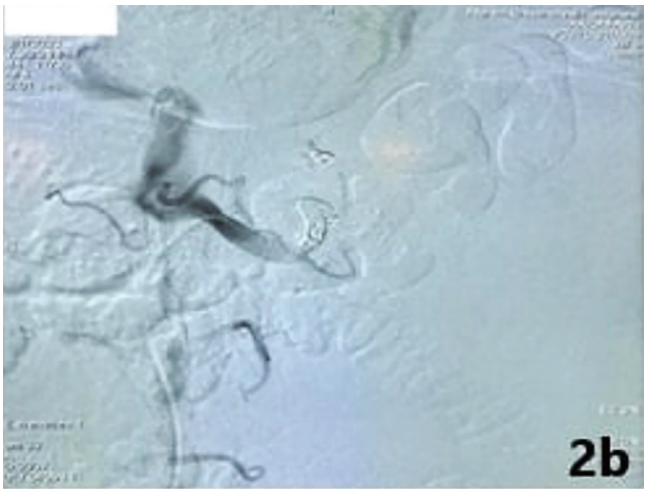

Figure 2b. Post-embolization angiographic examination shows complete devascularization of the mass after embolization of arterial structures supplying the mass with embolic particles and coils

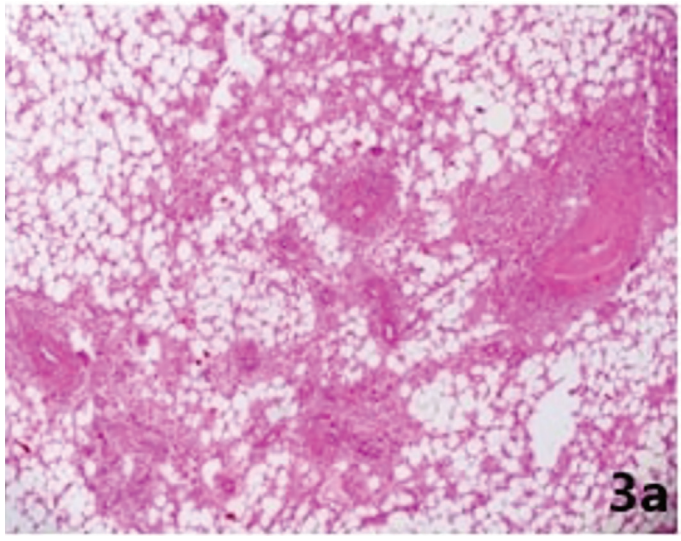

Figure 3a. Thick-walled vascular structures are observed in a background rich in adipocytes (H&E x40)

clinic at the 1-month and 6-month postoperative periods. At the first and sixth month follow-ups after the embolization procedure, serum creatinine levels, complete blood counts, and full urine analyses were within normal ranges. At the 6-month follow-up ultrasound imaging after the embolization procedure, it was observed that the recurrent mass had regressed to dimensions of 96x92x90 mm, and Doppler examination revealed no vascular coding within the mass. Verbal and written informed consent was obtained from the patient for the study.

Discussion

Renal AMLs are benign mesenchymal tumors composed of blood vessels, smooth muscle cells, and adipose tissue in the kidney.

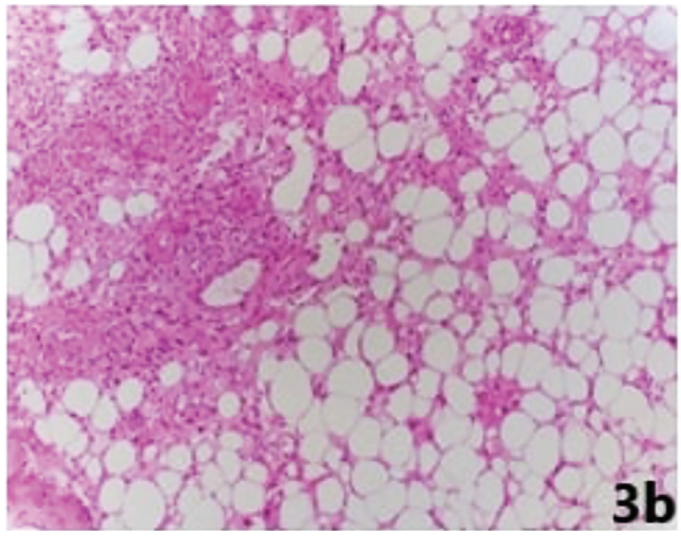

Figure 3b. Thick-walled vascular structures are observed in a background rich in adipocytes (H&E x100)

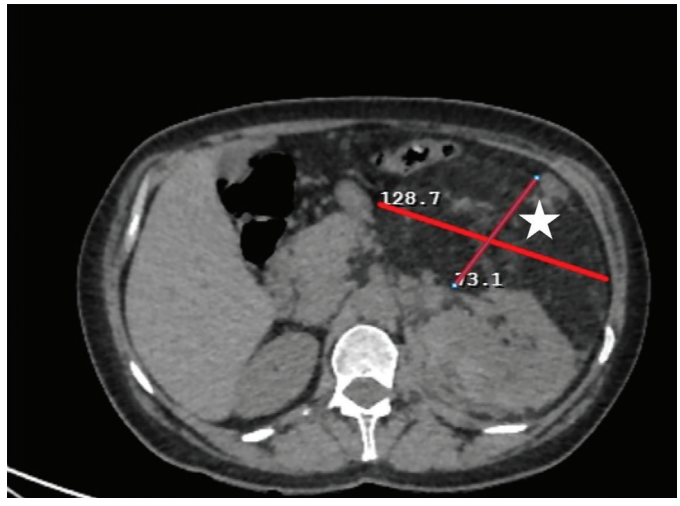

Figure 4. In the non-contrast abdominopelvic computed tomography examination; it demonstrates a mass consistent with angiomyolipoma, originating from the left kidney, with approximate dimensions of 15 cm in length, well-defined, lobulated contours, and containing dense fatty content (indicated with a star)

AMLs commonly exhibit renal localization, but they can rarely also present with extrarenal localization. AMLs with extrarenal localization are often incidentally found, and the retroperitoneal space is the second most common site of occurrence. Although most these extrarenal AMLs have a benign nature, excluding malignancy is challenging because of similarities in imaging techniques. Differential diagnosis includes lipoma, liposarcoma, papillary RCC, and adrenal myolipoma. Since the first case report by Friis and Hjortrup in 1982, sixty cases of extrarenal AML have been reported. Among them, only 16 have been reported as retroperitoneal extrarenal AML (RERAML) (4).

RERAMLs are usually larger than 10 cm in size and often have an asymptomatic course because of their retroperitoneal location.

However, they can manifest with non-specific symptoms such as abdominal pain, hematuria, epigastric fullness, and constipation. Imaging techniques play a crucial role in the diagnosis of RERAML, particularly in determining tumor size and localization, as well as guiding surgical planning. Additionally, they provide valuable guidance to clinicians in the follow-up of patients who have undergone nephrectomy due to AML (14).

In the follow-up and treatment selection of AML, bleeding risk should always be considered because of the unique structure of the mass. Generally, most symptomatic AMLs are relatively large, and most studies in the literature emphasize a cut-off value of 4 cm (3). In a comprehensive literature review, it was reported that 82% of patients with AMLs larger than 4 cm were symptomatic, with 9% experiencing hemorrhagic shock at admission, whereas only 23% of patients with small tumors were symptomatic (5). During the follow-up process of our case, the patient was called in for check-ups at the first and sixth months after the embolization procedure. Routine biochemical tests, complete blood count, full urinalysis, and abdominopelvic Doppler ultrasound examinations were performed during follow-up. It was observed that the mass had decreased in size and there was no vascularization.

In the treatment approach for these tumors, the most important goal is to avoid renal parenchymal damage as much as possible, especially in patients with hereditary AML or renal dysfunction, through nephron-sparing surgery or selective embolization. Preventive embolization can be applied in large asymptomatic tumors, in women of childbearing age, or in patients who may have limited access to follow-up or emergency care. Although some patients may experience post-embolization complications, selective transarterial embolization has been found to be effective in reducing tumor size in most renal AML cases with acceptable complication and recurrence rates. In our case, approximately 4 years ago, when we detected a large-sized AML in the left kidney (Figure 4), the reasons for choosing left simple nephrectomy as the treatment were as follows: At that time, our interventional radiology clinic did not have adequate physical equipment, and the tumor was quite large, significantly increasing the likelihood of spontaneous rupture.

In patients with sporadic AML, selective transarterial embolization can prevent recurrence for many years. However, patients with tuberous sclerosis and multiple AMLs continue to pose a therapeutic challenge with high recurrence rates of up to 60% for clinicians. Selective transarterial AML embolization has shown significant success in preventing recurrences and is considered safe (13). The question of whether selective transarterial AML embolization preserves normal renal parenchyma to the maximum extent compared with new percutaneous and laparoscopic ablative techniques such as cryoablation, radiofrequency ablation, and partial nephrectomy remains relevant.

In patients with acute or potentially life-threatening bleeding or those at risk of bleeding, total nephrectomy has been used as a treatment option. However, in cases such as the one presented here with recurrent AML after nephrectomy and a high risk of bleeding due to its size, selective transarterial AML embolization is a reliable treatment method that is much less invasive than surgery and can be applied with lower morbidity and mortality rates in experienced clinics to prevent recurrence and avoid the possibility of a high-risk, fatal bleeding.

Based on the literature search, in a case presentation published by Jawahar and Kazan-Tannus (14) in 2017, a recurrent mass, approximately 9 cm in size, consistent with renal AML, was identified in the nephrectomy site 8 years later. However, despite the clinical similarities with our case, a different treatment approach was chosen, which involved the administration of an mTOR inhibitor called everolimus, resulting in a minimal reduction in the size of the mass (13).

Conclusion

There are many treatment options for AML ranging from surveillance to nephrectomy. However, there are limited studies comparing nephron-sparing surgical procedures with total nephrectomy or selective arterial embolization among treatment options. As in our study, the application of selective arterial AML embolization, which is a minimally invasive treatment method, in the presence of extrarenal retroperitoneal AML with a high risk of bleeding and recurrence is both promising in terms of outcomes and more reliable in terms of complications when used in conjunction with surgical treatments. However, in post-embolization follow-ups, there may be a reduction in tumor size or AML revascularization, unchanged or increased tumor size, or unplanned re-embolization or re-operation due to acute retroperitoneal bleeding. Therefore, patient follow-up after AML embolization is also of great importance.

Ethics

Informed Consent: Verbal and written informed consent was obtained from the patient for the study.

Authorship Contributions

Surgical and Medical Practices: H.E.D., Concept: Y.Y.K., Design: H.E.D., C.Ö., Data Collection or Processing: F.E.G.S., Analysis or Interpretation: Y.Y.K., Literature Search: A.N., F.E.G.S., Writing: A.N.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declare that they have no relevant financial.

References

1. Soulen MC, Faykus MH Jr, Shlansky-Goldberg RD, Wein AJ, Cope C. Elective embolization for prevention of hemorrhage from renal angiomyolipomas. J Vasc Interv Radiol 1994;5:587-591.
2. Koo KC, Kim WT, Ham WS, Lee JS, Ju HJ, Choi YD. Trends of presentation and clinical outcome of treated renal angiomyolipoma. Yonsei Med J 2010;51:728-734.
3. Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. J Urol 2002;168:1315-1325.
4. Venyo AK. A Review of the Literature on Extrarenal Retroperitoneal Angiomyolipoma. Int J Surg Oncol 2016;2016:6347136.
5. Oesterling JE, Fishman EK, Goldman SM, Marshall FF. The management of renal angiomyolipoma. J Urol 1986;135:1121-1124.
6. Bissler JJ, Kingswood JC. Renal angiomyolipomata. Kidney Int 2004;66:924-934.
7. Lemaitre L, Claudon M, Dubrulle F, Mazeman E. Imaging of angiomyolipomas. Semin Ultrasound CT MR 1997;18:100-114.
8. Dickinson M, Ruckle H, Beaghler M, Hadley HR. Renal angiomyolipoma: optimal treatment based on size and symptoms. Clin Nephrol 1998;49:281-286.
9. Ulukaradag E, Memik Ö, Özkan TA. Renal Angiomyolipoma, Review. Bull Urooncol 2015;14:203-206.
10. Hocquelet A, Cornelis F, Le Bras Y, Meyer M, Tricaud E, Lasserre AS, Ferrière JM, Robert G, Grenier N. Long-term results of preventive embolization of renal angiomyolipomas: evaluation of predictive factors of volume decrease. Eur Radiol 2014;24:1785-1793.
11. Murray TE, Doyle F, Lee M. Transarterial Embolization of Angiomyolipoma: A Systematic Review. J Urol 2015;194:635-639.
12. Staehler M, Sauter M, Helck A, Linsenmaier U, Weber L, Mayer K, Fischereder M. Nephron-sparing resection of angiomyolipoma after sirolimus pretreatment in patients with tuberous sclerosis. Int Urol Nephrol 2012;44:1657-1661.
13. Kumar S, Jayant K, As S, Singh SK, Agrawal S. Page kidney secondary to large splenic artery aneurysm bleeding and its management by angioembolization. Nephrourol Mon 2014;6:e17144.
14. Jawahar A, Kazan-Tannus J. Retroperitoneal extrarenal angiomyolipoma at the surgical bed 8 years after a renal angiomyolipoma nephrectomy: A case report and review of literature. Urol Ann 2017;9:288-292.