

Case Report

When benign turns aggressive: renal angiomyolipoma with renal vein invasion

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ABSTRACT

Angiomyolipoma (AML) is the most common benign kidney tumor, made up of fat, smooth muscle, and abnormal blood vessels. It represents 0.3-3% of kidney tumors and is more common in women. AMLs are typically diagnosed via computed tomography (CT) or magnetic resonance imaging (MRI), with most being asymptomatic and requiring no treatment. However, larger or symptomatic AMLs can cause hematuria, flank pain, or bleeding, necessitating treatments like selective arterial embolization, partial nephrectomy, or active surveillance. Recent studies question the traditional 4 cm intervention threshold, suggesting that larger AMLs may be safely monitored based on individual risk factors. This case report discusses a 47-year-old woman with a 4.1×2.1 cm AML with renal vein invasion, treated successfully with laparoscopic nephrectomy. The findings highlight the need for personalized treatment strategies considering tumor characteristics and patient factors, and emphasize the importance of ongoing renal function monitoring.

Keywords: Angiomyolipoma, Thrombectomy, Laparoscopic nephrectomy and size threshold

INTRODUCTION

AML is the most common benign mesenchymal tumor of the kidney, composed of adipose tissue, smooth muscle, and dysmorphic blood vessels.^{1,3} It accounts for 0.3-3% of kidney tumors, with a higher prevalence in women (2:1 ratio). AMLs are mostly sporadic (80%), but 20% are associated with genetic conditions like tuberous sclerosis complex.

Sporadic AMLs typically occur later in life, are usually unilateral, and grow slowly, while TSC-associated AMLs are more aggressive and often bilateral.^{2,3} AMLs are diagnosed primarily through imaging techniques such as CT or MRI, which reveal their characteristic fat content. However, lipid-poor AMLs can be challenging to

distinguish from malignant renal masses, sometimes necessitating further imaging or biopsy. Most AMLs are asymptomatic and do not require intervention, but larger or symptomatic tumors may present with hematuria, flank pain, or hemorrhage, leading to treatments like selective arterial embolization, partial nephrectomy, or active surveillance.⁹

Recent studies have questioned the traditional 4 cm size threshold for intervention, suggesting that larger AMLs can also be safely monitored. Treatment decisions should consider individual risk factors, including tumor size and patient demographics. Given the limited comparative data on treatment outcomes, the best management strategy for AMLs remains unclear, highlighting the need for further research.⁸ 80% present as an incidental finding and 15%

as Wunderlich syndrome. Renal vein invasion is extremely rare with around 76 cases published in the literature.^{2,3,5}

CASE REPORT

47-year-old woman with a history of systemic hypertension and renal lithiasis. In January 2021, she presented with right renoureteral colic associated with gross hematuria with spontaneous remission, dysuria, straining and tenesmus. Physical examination was unremarkable, no signs of tuberous sclerosis complex. A contrast enhanced abdominal and pelvis CT scan showed right kidney with intraluminal lesion in the pyelocaliceal system and vascular tract of the right renal vein, with 79 Hounsfield Units (HU), without enhancement in the arterial phase, 4.1×2.1 cm in its long axes (Figure 1).

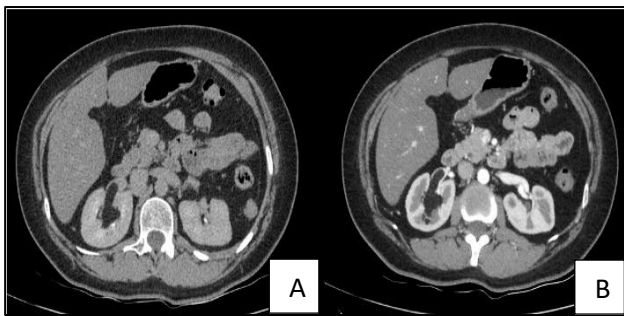


Figure 1 (A and B): CT image of renal lesion extending to the renal vein.

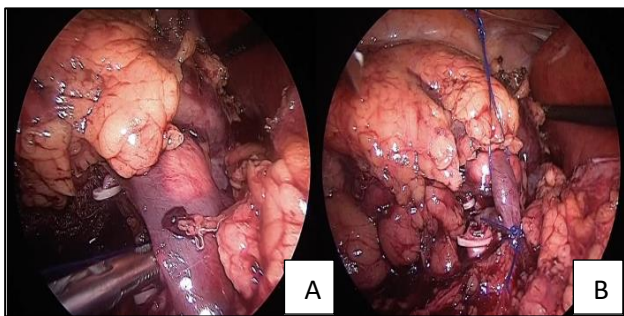


Figure 2 (A and B): Intraoperative scene, radical nephrectomy with renal vein thrombectomy.



Figure 3: Surgical outcome, radical nephrectomy with venous thrombectomy findings.

A right radical laparoscopic nephrectomy with renal vein thrombectomy was performed (Figure 2), with total operative time of 3 hours 15 minutes, blood loss of 400 ml with no transfusion needed. She was discharged on the second postoperative day and the drain was removed on the fifth postoperative day uneventfully.

Histopathological examination revealed an AML with predominance of adipose tissue in the renal sinus measuring 2.8×2.5 cm with extension to the renal vein (Figure 3).

DISCUSSION

AML represents approximately 1% of all renal tumors and is the most common mesenchymal neoplasm. It was first described in 1951 by Morgan and renal sinus invasion was first described in 1982 by Kutcher.^{5,6} The clinical presentation consists of flank pain, hematuria, and a palpable mass that can be observed in up to 41% of patients.

The diagnosis is made through a computed axial tomography, the presence of fat being characteristic, with the density-10 HU being almost diagnostic.² Despite its benign nature, chronic kidney disease (CKD) progression is common among AML patients, regardless of the treatment strategy, highlighting the need for ongoing renal function monitoring.^{4,7,9} The traditional 4-cm size threshold for active AML treatment is widely accepted but lacks strong evidence.

Treatment decisions should also consider tumor growth and patient preference. Kutcher et al. performed a right radical nephrectomy through a thoracoabdominal approach. The tumor extended into the right renal vein and inferior vena cava.⁶ Kheir et al, conducted a right radical nephrectomy and thrombectomy using cardiopulmonary bypass.⁵ Abreu et al, suggested an open right radical nephrectomy and renal thrombectomy, recommending a temporary inferior vena cava filter to prevent pulmonary thromboembolism.¹⁰ Laparoscopic surgery, a less invasive alternative to open surgery, has shown promise in treating large AMLs with renal invasion, suggesting it could become a preferred approach due to its effectiveness and lower morbidity.⁸⁻¹⁰

CONCLUSION

Surgical treatment for AML is justified in patients with tumors larger than 4 cm, symptomatic cases, or those with extension to the renal vein or inferior vena cava. Nephrectomy or selective embolization is recommended for cases with intratumoral or perinephric hemorrhage. Although laparotomy is traditionally recommended for venous involvement, laparoscopic surgery, in expert hands, offers comparable outcomes. The traditional 4-cm treatment threshold may need reevaluation, considering other factors such as patient age, tumor growth rate, and preference. While surgery remains the most common

active treatment, it is crucial to balance treatment decisions with the potential risk of CKD progression and the need for vigilant long-term follow-up.

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