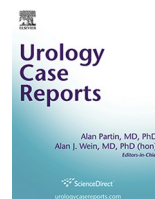




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Aggressive Renal Angiomyolipoma of the Lipomatous Variant With Inferior Vena Cava Thrombus: A Case Report and Review of the Literature

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ABSTRACT

Two variants of renal angiomyolipoma (AML)—classic and epithelioid—have been described. Although the epithelioid variant has been reported to demonstrate an aggressive clinical behavior, classic AML is usually benign. Herein, we report a case of a 42-year-old asymptomatic woman with a lipomatous variant of renal AML associated with an inferior vena cava thrombus managed with radical nephrectomy and caval thrombectomy.

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Introduction

Angiomyolipoma (AML) is a benign renal mesenchymal tumor affecting more than 10 million people worldwide, predominantly in women aged 40–50 years. It might be sporadic or occurs in association with tuberous sclerosis complex or lymphangiomyomatosis (LAM).¹ There are 2 variants of AML: classic (triphasic) and epithelioid.

Although AML is classically benign, the epithelioid variant can closely mimic renal cell carcinoma radiographically. Epithelioid AML has been reported to exhibit aggressive clinical course with metastases, recurrences, and high rate of mortality.^{2–4} Rarely, AML might invade the major renal vein and/or lymph nodes. However, involvement of regional lymph nodes is interpreted as multifocality of growth rather than true metastases or malignant behavior. Herein, we report a case of lipomatous AML that demonstrates an unusual aggressive behavior with inferior vena cava (IVC) tumor thrombus.

Case presentation

The patient is a 42-year-old asymptomatic woman with no past medical history referred to us on account of a hyperechoic right kidney mass and IVC thrombus found on routine abdominal ultrasound. Physical examination was unremarkable, and laboratory values were within normal limits, with hemoglobin of 13.2 g/dL and creatinine of 0.85 mg/dL. Computed tomographic (CT) scan of the abdomen confirmed a 3-cm right upper pole renal mass with central fat attenuation and a 5-cm level II IVC thrombus (extension into the right renal vein and IVC below the level of the hepatic veins; Fig. 1A and B). Shortly after imaging diagnosis, she presented with a 1-week history of pleuritic chest pain and shortness of breath in the recumbent position. Urgent chest CT angiogram showed a pulmonary tumor embolus (–65 HU) in the right anterior segmental branch of the pulmonary artery, with a corresponding infarct in the medial segment of the right lower lung lobe. The CT also revealed multiple bilateral lung cysts, suggesting a diagnosis of LAM.

She underwent a right radical nephrectomy and IVC thrombectomy through a modified Chevron incision. With full mobilization of the right kidney, control of the IVC proximal and distal to the thrombus, and the contralateral renal vein, a free-floating thrombus was easily delivered intact through an anterior ellipsoid cavotomy. Subsequent to IVC repair, a right radical

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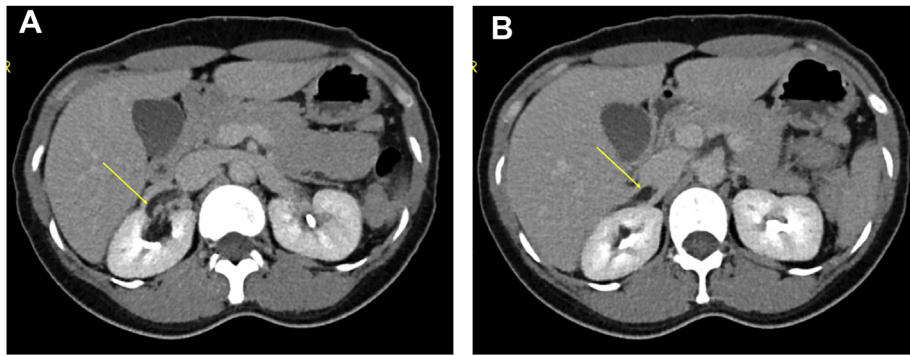


Figure 1. Axial abdominal computed tomographic scan showing (A) focal mass of fat attenuation in the right kidney and the right renal vein and (B) hypodense tumor thrombus in the inferior vena cava.

nephrectomy was performed without perioperative complications. The patient fared well postoperatively and was discharged home on postoperative day 4.

Gross specimen examination revealed a $2.5 \times 2.2 \times 2.0$ cm fatty tumor located in the upper pole of the right kidney, extending into the renal sinus. There was a 6.8×0.9 cm tumor thrombus protruding through the renal vein, without involvement of the vein wall (Fig. 2A). Microscopic examination revealed a tumor composed of adipose tissue predominantly, scattered thick-walled blood vessels, and minor smooth muscle cells surrounding abnormal vessels (Fig. 2B). Immunophenotypic expression includes positive staining for melanocytic markers (HMB-45) and smooth muscle markers (SMA, smooth muscle actin). S-100 immunostain showed positive cytoplasmic staining.

Discussion

AML is a benign triphasic renal tumor consisting of variable amount of adipose tissue (*-lipo-*), smooth muscle cells (*-myo-*), and abnormal thick-walled vessels (*-angio-*). AML most commonly are sporadic (80%) or are associated with tuberous sclerosis complex or LAM (20%), with the sporadic variety occurring with a 4:1 predominance in women. AML more commonly becomes symptomatic in lesions >4 cm, and include fever, gastrointestinal upset, flank pain, palpable renal mass, hematuria, hypertension, anemia, renal failure, and shock from retroperitoneal hemorrhage.

It is generally recommended that asymptomatic AML might be monitored annually or semiannually by CT or ultrasound if <4 cm in its largest diameter. However, persistently symptomatic lesions <4 cm or lesions ≥ 4 cm should be treated with selective arterial embolization, radiofrequency ablation, or nephron-sparing procedures.⁵ However, surgical extirpation might be used in cases of aggressive, epithelioid, or vessel-invasive AML.

The sequelae of vascular invasion and IVC tumor thrombus in an aggressive AML can be life threatening, with increased risks of vessel occlusion and spontaneous retroperitoneal hemorrhage (Wunderlich syndrome). AML with IVC thrombus, irrespective of size, must be managed urgently with radical nephrectomy and caval thrombectomy, as used in this case. Definitive treatment is essential to avoid threats of tumor embolism and subsequent respiratory compromise. Recently, a randomized trial of everolimus vs placebo in patients with >3 cm AML reported 42% objective response rate ($>50\%$ reduction in tumor volume) with treatment; however, there have been no studies in patients with locally advanced AML.¹

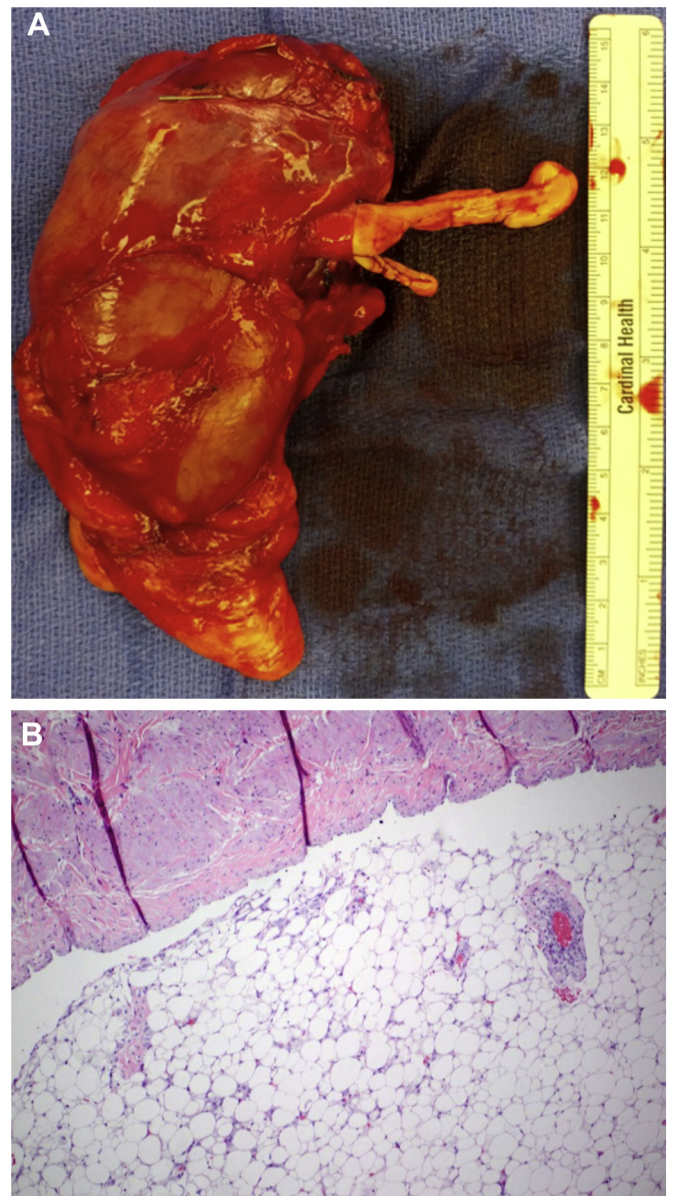


Figure 2. (A) Gross specimen of the right kidney showing lipomatous tumor thrombus in the renal vein. (B) Hematoxylin and eosin stain of angiomyolipoma showing 3 components: abnormal thick-walled blood vessel, spindle cells spinning of the vessel wall, and mature adipose tissue.

Conclusion

Rarely, classic renal AML can behave aggressively with tumor thrombus in the renal vein and IVC. AML with such characteristics should be managed aggressively with thorough preoperative radiographic characterization and prompt surgical intervention to avoid life-threatening complications.

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