

# Robotic-assisted radical nephrectomy for renal angiomyolipoma with inferior vena cava thrombus extension

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**Abstract** Renal angiomyolipoma with inferior vena caval venous extension is rare with only 40 cases reported in the literature. We report a case of a 35-year-old lady with angiomyolipoma with inferior vena caval thrombus that was managed surgically with robotic-assisted radical nephrectomy.

**Key Words:** Angiomyolipoma, inferior vena caval venous extension, robotic assisted radical nephrectomy

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## INTRODUCTION

Renal angiomyolipomas are benign lesions that account for 10% of all renal tumors.<sup>[1]</sup> They are usually single and occur predominantly in middle-aged females. Twenty-three percent of renal angiomyolipoma are associated with tuberous sclerosis.<sup>[1]</sup> These lesions are often diagnosed incidentally on routine imaging studies.<sup>[2]</sup> Despite their benign nature, rare cases have been reported with lymph node and vascular extension. Here we report the 40<sup>th</sup> case of a patient with AML with infrahepatic IVC thrombus.

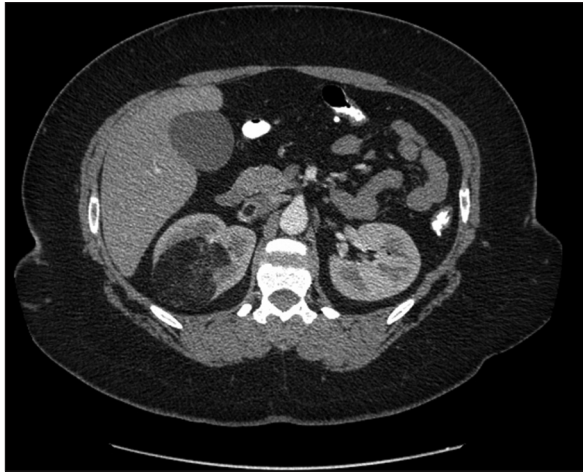
## CASE REPORT

A 35-year-old woman presented to our institution with non specific abdominal pain, her past medical history was significant for morbid obesity (body mass index of 50). She had no history of neurological disease, personal or family history of tuberous sclerosis (TS). On physical examination, there were no signs of

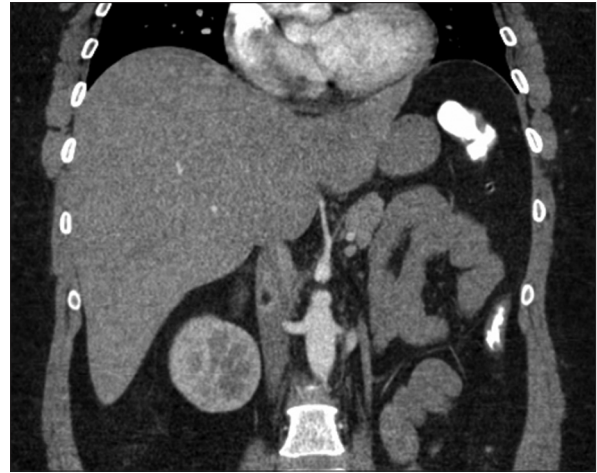
adenoma sebaceum, abdominal tenderness or palpable masses. Ultrasound examination of the abdomen showed a large lobulated hyperechoic lesion at the upper pole of the right kidney measuring 5.3 × 6.1 × 6.6 cm, showing no flow on Doppler interrogation that likely represent a large angiomyolipoma (AML). To better delineate the renal mass, a CT scan of the abdomen and pelvis with contrast was performed and demonstrated a well-defined round fat containing lesion in the mid upper pole of the right kidney measuring 4.8 × 5.6 × 4.6 cm. Moreover, there was a tumor thrombus extending from the right renal vein and inferior vena cava (IVC) 9 cm caudal to the hepatic veins [Figures 1 and 2]. The patient's hemoglobin was 139 g/dl and serum creatinine was 67 μmol/L.

Management options were discussed, with the understanding that malignancy cannot be excluded and the potential lethal nature of vena caval thrombus. The patient consented to undergo a robotic assisted radical nephrectomy with inferior vena cavalthrombectomy. Intraoperatively, we identified the renal vessels with a mobile venous thrombus just extending into the inferior vena cava. After clipping and dividing the renal artery, the venous thrombus completely retracted into the renal vein which was confirmed with intraoperative ultrasound and three large hem-o-lock clips were placed on the renal vein distal to the venous thrombus. The specimen was removed via a right Gibson incision [Figures 3 and 4]. There were no intraoperative or postoperative complications. The estimated blood loss was

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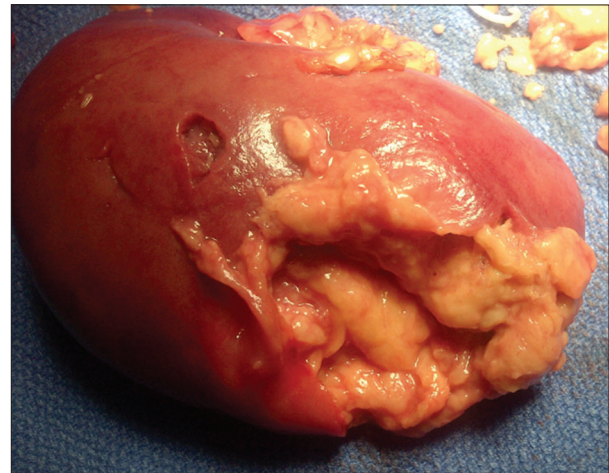
**Figure 1:** Cross-sectional CT scan of the abdomen. Shows right renal mass with the proximal extension of tumor thrombus into the right renal vein



**Figure 2:** Coronal view of the abdomen show the distal end of the inferior vena cava thrombus 9 cm caudal to the hepatic veins



**Figure 3:** Intraoperative picture of the right kidney shows the tumor thrombus protruding out from the right renal vein



**Figure 4:** Intraoperative picture of the right upper pole renal mass

200 ml. and total operative time was 245 minutes and the patient was discharged on the 4<sup>th</sup> postoperative day in a stable condition. The patient remained asymptomatic 3 months post operatively.

### Surgical pathology

The kidney measures 9.5 × 7.5 × 4.5 cm. The tumor was located in the mid upper pole measuring 5.5 × 5 × 4.5 cm. It extended into the sinus fat and grossly abutting the renal capsule. The venous thrombus was identified inside the renal vein with no signs of venous wall invasion. Both the renal mass and venous thrombus was rich in adipose tissue with no signs of cellular atypia, mitosis or necrosis confirming the diagnosis of benign angiomyolipoma.

### DISCUSSION

Angiomyolipoma is a benign mesenchymal neoplasm consisting aneurysmal vessels, smooth muscle and mature adipose tissue. The incidence is between 0.13 and 0.3% in

the general population.<sup>[1]</sup> These sporadic lesions are more common in females and occur in the age group between 40 and 50-year-old (median 45 years old).<sup>[2]</sup> Twenty percent of all renal angiomyolipoma are associated with tuberous sclerosis and they typically present at a younger age (mean age 30 years) with bilateral multifocal angiomyolipoma.<sup>[3]</sup> The diagnosis can be made with a CT scan and confirmed by the presence of fat within a renal lesion (a value of -20 Hounsfield Units of less).<sup>[4]</sup> Symptoms are usually pain, abdominal mass or hemorrhagic shock from retroperitoneal haemorrhage (Wunderlich syndrome) and commonly develop when the tumor size is more than 4 cm.

Management of angiomyolipomas are usually conservative unless large tumors are symptomatic. Oesterling<sup>[5]</sup> recommended a 4 cm size cut-off for more aggressive surveillance and intervention is required when symptomatic. On the other hand, Dickinson<sup>[6]</sup> recommended treatment in patients who had lesions with significant growth, at risk of trauma, symptomatic or tumors more than 8 cm.

Rarely, angiomyolipoma may present with venous thrombus extension raising the concern of malignant transformation. To date, there is no imaging that can differentiate between benign angiomyolipoma from liposarcoma or fat containing renal cell carcinoma.

Our literature review revealed 40 reported cases to date.<sup>[7-12]</sup> Upon presentation, two-thirds (27/40) of these patients were symptomatic with flank pain as the most common symptom. The mean age group affected is 45-year-old with a female predominance (male/female = 7/32). Moreover, there were three reported cases of angiomyolipoma that progressed into the inferior vena cava during pregnancy.<sup>[10,12,13]</sup> These findings would suggest that angiomyolipoma can be hormonally influenced. Furthermore, the average affected size was 9.2 cm suggesting that large tumors increase the risk of vascular invasion. In addition, bilateral and multifocal tumors were reported in 32% of cases (13/40). In keeping with Schade<sup>[7]</sup> literature review, we did not find any association between tuberous sclerosis and the risk of developing intravascular invasion.

As the field of Robotic surgery in the urology practice in gaining a wide spread. Robotic-assisted radical nephrectomy for renal masses with IVC thrombus seems to be a good and safe alternative to conventional open and laparoscopic approach. However, more studies are needed to assess further safety.

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