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Case Report

Surgical Case Report: Renal Angiomyolipoma with Inferior Vena Cava Involvement

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Abstract

Renal angiomyolipoma (AML) is a benign mesenchymal renal tumor composed of abnormal blood vessels, smooth muscle, and adipose tissue. Although most AMLs are incidentally detected and clinically indolent, large tumors may lead to severe complications. Extension into the Inferior Vena Cava (IVC) is extremely rare and significantly impacts clinical management.

We present the case of a 70-year-old woman admitted with acute right upper quadrant abdominal pain and abnormal liver function tests. Contrast-enhanced computed tomography revealed a large right renal mass with macroscopic fat, consistent with angiomyolipoma, associated with a level II tumor thrombus extension into the inferior vena cava and compression of adjacent organs. FDG positron emission tomography showed no pathological uptake, supporting the benign nature of the lesion and excluding the need for preoperative biopsy.

After multidisciplinary evaluation, the patient underwent open right nephrectomy with caval thrombectomy. The procedure was performed safely, and the postoperative course was uneventful. Histopathological examination confirmed a classic renal angiomyolipoma without malignant features.

Keywords: Renal Angiomyolipoma (AML); Inferior Vena Cava (IVC); Nephrectomy; Caval thrombectomy

Introduction

Renal Angiomyolipomas (AMLs) are benign mesenchymal tumors composed of smooth muscle and adipose tissue. While usually asymptomatic, larger AMLs (≥ 4 cm) may cause abdominal pain, hematuria, or mass-related symptoms [1]. A rare complication is invasion of the Inferior Vena Cava (IVC), which may lead to venous obstruction or embolic events [1,2]. Women are more commonly affected, and larger tumors are often observed in this cohort. This case describes a 70-year-old woman with a large right renal AML involving the vena cava, causing significant manifestations, who presented to our attention at ASST Papa Giovanni XXIII.

Case Presentation

The patient, a 70-year-old woman (72 kg, 155 cm), presented to the emergency department on July 1, 2024, with acute right hypochondrial pain, exacerbated by meals, and a single episode of bilious vomiting. The patient's medical history included hypertension, uterine prolapse, and a cholecystectomy performed approximately 40 years earlier. She denied recent cardiovascular events or relevant allergies, except for nickel sensitivity. On admission, vital signs were stable. Laboratory tests showed markedly elevated liver enzymes (AST 607 U/L, ALT 444 U/L), while total bilirubin (0.82 mg/dL) and coagulation parameters (INR 0.99) were within normal limits.

A contrast-enhanced abdominal CT scan revealed a large, dysmorphic, predominantly adipose mass arising from the right kidney (11 cm × 13 cm × 12 cm), consistent with a renal angiomyolipoma. The lesion displaced the kidney anteriorly and exerted a significant extrinsic compressive effect on adjacent structures. In particular, compression of the liver and biliary ducts resulted in dilation of the biliary tree and secondary alteration of liver enzyme levels, without evidence of intrinsic biliary obstruction. The mass also compressed the duodenum, although no complete gastrointestinal obstruction was observed. Therefore, the clinical and laboratory findings were considered as directly related to the mechanical compression from the renal mass. We also performed an FDG PET scan, and the right renal mass showed low FDG uptake. This lack of metabolic activity suggested that the lesion was benign, consistent with the typical behaviour of Renal Angiomyolipomas (AMLs), and, therefore, an invasive biopsy was not considered necessary.

A colangio-MRI excluded intrinsic obstruction of the biliary ducts and therefore the patient was considered a candidate for nephrectomy and caval thrombectomy (Figure 1 and 2).

Surgical intervention

An open right radical nephrectomy with caval thrombectomy was performed on November 29, 2024. Surgery was completed through

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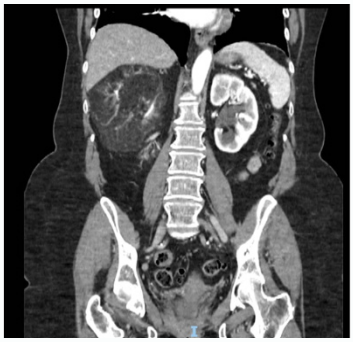

Figure 1: Computed tomography scan at time of admission showing a large right renal angiomyolipoma with heterogeneous appearance and macroscopic fat, extending into the inferior vena cava.

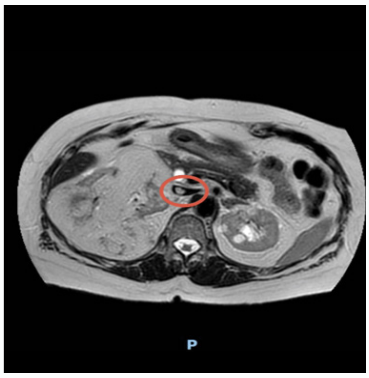

Figure 2: Axial abdominal MRI confirming the bulky right renal angiomyolipoma with inferior vena cava involvement and mass effect on adjacent structures.

an anterior Chevron abdominal approach. The kidney and tumor thrombus were removed end block. The operation proceeded without complications, and the patient experienced a smooth postoperative recovery.

Follow-up

After 12-month follow-up the patient is alive and abdominal CT scan showed no recurrence of the AML (Figure 3 and 4).

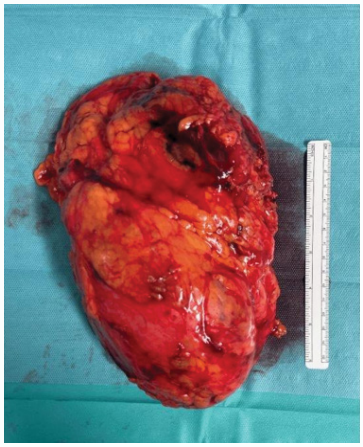

Figure 3: Gross specimen of a giant classic renal angiomyolipoma with well-defined, predominantly adipose appearance arising from the renal parenchyma.

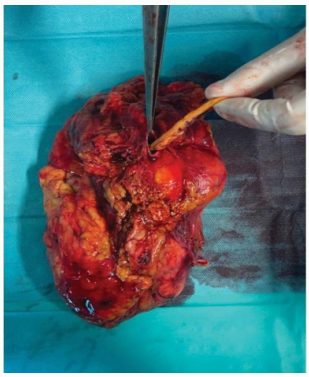

Figure 4: Intraoperative gross view demonstrating angiomyolipoma extension into the renal vein, forming an intraluminal tumour thrombus.

Pathology report

Histological examination revealed a classic variant of renal angiomyolipoma measuring 10.5 cm in diameter with extension into the perirenal fat and renal vein. Immunohistochemical staining was positive for SMA and HMB45 (focal), consistent with AML. No evidence of malignancy or metastasis was observed in the single lymph node analyzed (Figure 5).

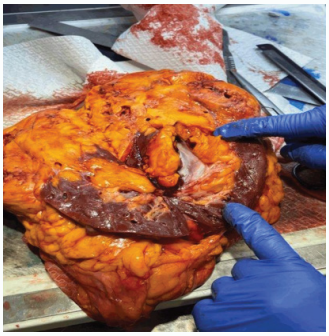

Figure 5: Gross section of the specimen showing the characteristic heterogeneous adipose, smooth muscle and vascular components of classic renal angiomyolipoma.

Discussion

We report the case of a large AML with direct invasion of the Inferior Vena Cava (IVC). Extension into the renal vein and Inferior Vena Cava (IVC) is exceedingly rare, with fewer than 100 cases reported in the literature, and represents a major diagnostic and therapeutic challenge [1]. In the present case, the large tumor size and extension into the IVC were associated with compression of adjacent structures, including the biliary tract and duodenum, resulting in acute abdominal symptoms and altered liver function tests. Such a presentation is uncommon and underscores the heterogeneous clinical behaviour of AMLs (Figure 6).

While typically benign, vascular involvement, particularly with the IVC, complicated diagnosis and treatment. IVC extension may lead to venous obstruction, thromboembolism, or life-threatening embolic events [2-4].

Imaging plays a central role in the diagnosis and management of renal AMLs. Computed tomography and magnetic resonance imaging typically allow differentiation from malignant renal tumors through

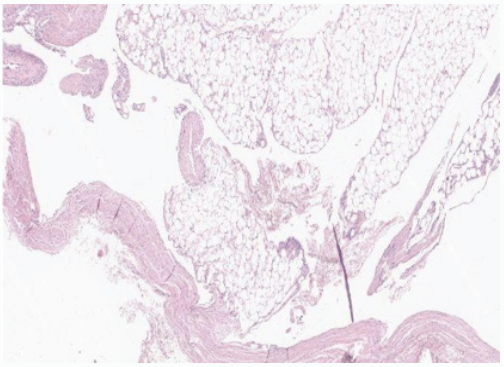

Figure 6: Histological section (H&E, ×4) showing a classic renal angiomyolipoma composed of mature adipose tissue, smooth muscle bundles, and thick-walled blood vessels, with extension into the renal vein.

the identification of macroscopic fat [4,5]. Classic AMLs are identified by macroscopic fat visible on CT, but minimal fat could complicate this distinction. MRI was effective for detecting both macroscopic and microscopic fat, with in-phase and opposed-phase sequences helping differentiate AML from RCC, especially when macroscopic fat was absent. Microscopic fat in minimal-fat AMLs causes a characteristic signal loss, known as the India ink artifact, which can aid in the differential diagnosis [6,7]. However, in cases with atypical features or extensive vascular involvement, differentiation from renal cell carcinoma or retroperitoneal liposarcoma may be challenging. Liu et al. [4] compared 6 pts affected by AML and 18 ccRCC with venous tumor thrombus: the presence of fatty components, distinct geometric growth patterns, and absence of venous wall invasion may serve as critical diagnostic clues for differentiating tumor venous thrombus of AML origin. An alternative diagnostic approach could involve the use of Positron Emission Tomography (PET). Renal Angiomyolipomas (AMLs) typically exhibited minimal to low Fluorodeoxyglucose (FDG) uptake on PET and PET/CT scans [8,9]. In our patient, FDG positron emission tomography demonstrated no pathological uptake, supporting the benign nature of the lesion and contributing to the decision to avoid preoperative biopsy. This approach is consistent with available evidence showing minimal FDG uptake in AMLs and limited diagnostic benefit of biopsy when imaging findings are concordant with a lack of malignant features [8-10].

The management of AMLs depends on tumor size, symptoms, and associated complications. Current recommendations support active surveillance or minimally invasive treatments for small, asymptomatic tumors, while surgical intervention is indicated for symptomatic lesions, rapid growth, or vascular extension [3]. In cases of IVC involvement, radical surgery with thrombectomy is required to achieve complete tumor removal and prevent potentially life-threatening complications such as venous obstruction or embolization.

In our case, open radical nephrectomy with caval thrombectomy allowed complete resection of the tumor and thrombus, with an uneventful postoperative course and AML was confirmed at final pathology. Due to the very rare incidence of this disease, only a few cases have been managed robotically [11]. A recent report described the first pure robotic-assisted level IV inferior vena cava thrombectomy for renal angiomyolipoma performed without cardiopulmonary bypass, demonstrating the technical feasibility of a minimally invasive approach even in cases of advanced venous involvement [11]. However, such an approach is currently limited to highly selected patients and specialized centers. In our case, considering the tumor size, extensive caval involvement, and compression of adjacent structures, an open surgical approach was deemed the safest and most appropriate option.

The patient is alive and free from recurrence at 12-month follow up. Data on long-term outcomes and recurrence after radical surgery for AML with IVC extension are limited, but available reports suggest a low risk of recurrence following complete resection in sporadic cases. Wang, et al. [12] reported 12 cases of AML with VTT, and no disease progression was observed during the follow-up period. The mean follow-up time in Liu, et al. [4], study was 36.6 months, with all patients surviving at the last follow-up, and no signs of tumor recurrence or metastasis were noted. Nevertheless, careful postoperative follow-up remains essential, particularly in patients with large tumors or complex presentations.

Conclusion

Renal angiomyolipoma with venous extension into the inferior vena cava represents a rare manifestation of a generally benign tumor that assumes a biologically aggressive behaviour [12]. The differential diagnosis mainly includes clear cell renal cell carcinoma, the most frequent cause of caval tumor thrombosis; the presence of a macroscopic adipose component within the thrombus, detectable by CT or MRI, represents the distinctive diagnostic element of angiomyolipoma [13,14]. Standard treatment is radical nephrectomy with caval thrombectomy, which prevents potentially fatal complications such as pulmonary embolism and intracardiac extension [12,13].

The prognosis is generally favorable after complete surgical resection, with the need for serial imaging surveillance to exclude recurrence [12].

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