

Sporadic Renal Angiomyolipoma with IVC Extension

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SUMMARY

Angiomyolipomas (AML) are the most common mesenchymal renal neoplasms arising in the cortex or medulla. Intra-renal and retroperitoneal hemorrhages have been frequently reported. AML can exceptionally involve the renal vein and inferior vena cava. We report a case with extension into the inferior vena cava.

CASE REPORT

A 50 year old previously well female patient had a pre-employment chest radiograph which showed straightening of the left heart border with elevated cardiac apex and pulmonary venous congestion. A trans-oesophageal echocardiogram shows dilated right heart chambers with 18 mm atrial septal defect and a tongue-like mass lesion projecting into the right atrium from the inferior vena cava (IVC).

Contrast enhanced abdominal CT was done for suspicion of caval thrombosis using Philips 256 slice scanner. Images were obtained in both nephrographic (90 seconds) and excretory (5 minutes) phases. The scans revealed a left renal sinus mass with fat attenuation measuring 1.7 x 2.4 cm (Fig 1). The mass extended into the renal vein and IVC to reach up to the right atrium (Fig 2). Other abdominal structures and lung bases were normal. Based on the clinical and imaging findings, diagnosis of renal angiomyolipoma (AML) with renal vein and IVC extension was made.

Patient then underwent a radical right nephrectomy and IVC thrombectomy followed by atrial septal patch closure. Intra-operatively the tumour was found to be too deep seated in the renal sinus and a partial nephrectomy could not be performed. The fat thrombus was found extending to the supradiaphragmatic segment of the IVC, just below the right atrium. The left kidney and renal vein were excised en-bloc post IVC cavotomy and thrombectomy. The histopathology analysis showed lobules of mature adipocytes haphazardly admixed with smooth muscle and blood vessels consistent with AML. No malignant cells or features were seen.

The post operative period was uneventful. Screening for tuberous sclerosis was not performed as unilateral single AML is not known to be associated. Follow up ultrasound was done for 18 months with no evidence of tumour recurrence noted.

DISCUSSION

AML are the most common mesenchymal renal neoplasms arising from the cortex or medulla and comprises of varying amounts of fat, smooth muscles and blood vessels¹. These are seen up to four times more commonly in the female population¹. Two types have been described: sporadic AML and AML associated with tuberous sclerosis complex (TSC). AML are found in 70-80% of patients with TSC and 20% of individuals with AML have TSC. Approximately 80-90% of renal AML occur sporadically².

Two distinct subtypes of AML have been described based on histological features: the classic triphasic type and monotypic epithelioid type. Classical AML are benign and composed of a proliferation of blood vessels, smooth muscle and adipose tissue in variable proportions. Epithelioid AML are composed of pure epithelioid cells with characteristic absence of adipose cells and abnormal blood vessels. These tumours are said to have malignant potential however the percentage of cases of renal malignancy due to epithelioid AML is not known³.

The diagnosis of AML is easily made based on characteristic imaging findings due to the presence of intratumoural fat. Ultrasound shows a lesion in the renal sinus, isoechoic with the sinus fat. These findings however can be difficult to demonstrate and are patient and operator dependent¹. On CT, it presents as a well-defined lesion with fat attenuation values. MRI shows a T1 hyperintense mass with loss of signal on fat suppression sequences. Small amount of fat can be detected using chemical shift imaging with loss of signal in the opposed phase MR sequence. Calcification is very rarely described in AML and presence of calcification should suggest an alternative diagnosis such as renal cell carcinoma.

The extension of AML into renal vein and IVC has been rarely described in the literature. The exact biological mechanism is debatable. Hypothesis have however been made such as tumour spread across vascular spaces into the vein particularly if it arose from the wall of a vein and then extends locally into the renal vein and IVC or due to rupture of AML directly into a renal vein⁴.

Blood vessels in AML frequently have an angiomatous arrangement with absence of elastic tissue in the wall thus predisposing the patient to small saccular aneurysm and spontaneous hemorrhage into the renal parenchyma, peritoneal cavity or retroperitoneum. TSC-associated AML tend to be larger, multiple and more likely to cause spontaneous hemorrhage than sporadic forms of AML.

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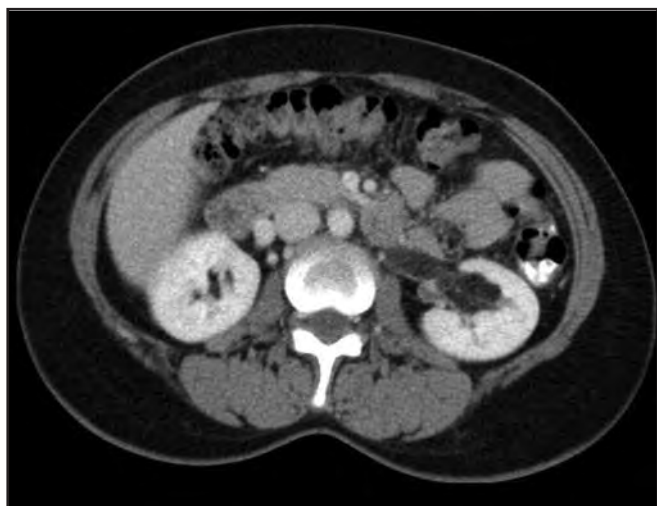


Fig. 1 : Contrast enhanced axial CT image showing a hypodense mass in the left renal sinus having similar attenuation to sinus fat. It is seen to extend into the left renal vein. The kidney parenchyma show symmetric enhancement.

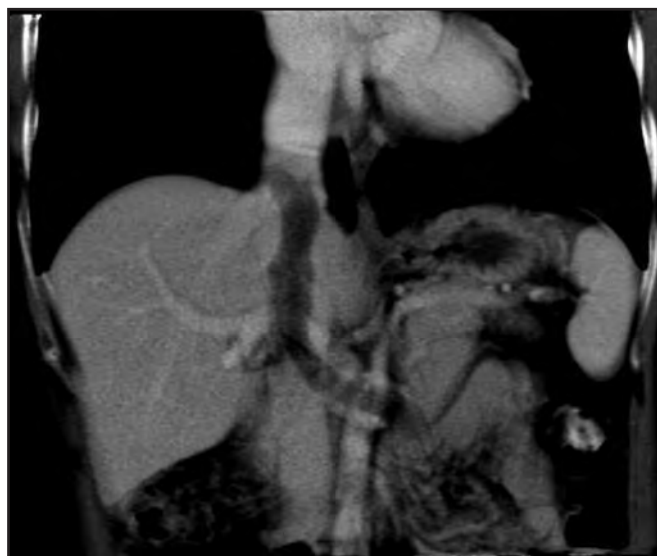


Fig. 2 : Coronal maximal intensity projection (MIP) CT scan shows fat density lesion in the left renal sinus extending into the left renal vein and supra-diaphragmatic inferior vena cava (IVC).

Oesterling et al have proposed a widely practice treatment protocol based on size and symptoms of AML. Annual US or CT is recommended for patients with isolated AML <4 cm in whilst 6-monthly US or CT is suggested for patients with lesions >4 cm for assessment of growth. Patients with TSC and AML <4 cm in diameter should be followed by a semi-annual US or CT⁵. CT kidneys in the portal venous phase is performed for patients requiring follow up in our local institution for radiation dose saving purposes.

AML often do not require intervention as these are mostly benign and asymptomatic. Indications for intervention however include suspicion of malignancy, spontaneous hemorrhage causing significant symptoms such as pain or hematuria and risk of rupture or other complication secondary to the formation of intrarenal aneurysm. Most

symptomatic AML can be managed by nephron-sparing approaches including angiographic embolization or partial nephrectomy¹. Some selected patients however may require complete nephrectomy as exemplified by the case presented.

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