

## Case Report: A Rare Case of A Renal Angiomyolipoma With Extension into the Renal Vein and IVC.

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**Abstract:** Renal angiomyolipomas are benign hamartomas composed of blood vessels, smooth muscle, and fatty tissue. AML is seen in two distinct clinical forms, a sporadic form and in association with Tuberous Sclerosis Complex (TSC). The sporadic form accounts for approximately 80% to 90% of cases of AML. Masses less than 4 cm in diameter are usually asymptomatic, but those larger than 4 cm often cause flank pain, hematuria, and anaemia and carry a significant risk of intratumoral or perinephric haemorrhage. We herein present a case of renal AML in a 60-year old female patient, who presented with left sided abdominal pain since last 6 months. Following abdominal ultrasound, computed tomography and MRI examination, the patient underwent abdominal laparotomy. The resected mass was sized 8×7×9 cm. Postoperative histopathological examination confirmed the lesion as a renal AML with extension into left renal vein and infra diaphragmatic segment of IVC. There are only few cases of renal AML reported in the literature to date which shown extension into the renal vein and IVC.

**Keywords:** Renal Angiomyolipoma, Renal Vein, IVC

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Date of Submission: 05-07-2019

Date of acceptance: 20-07-2019

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### I. Introduction

Renal angiomyolipomas are benign lesions composed of variable amounts of fat, smooth muscle and abnormal blood vessels. They occur spontaneously in the general population, mainly in women during their fifth decade; they occur at a much younger age and are frequently multiple in patients with tuberous sclerosis, with an incidence of 50–80%. They are rarely seen in neurofibromatosis and in autosomal dominant polycystic kidney disease. Angiomyolipomas are composed of thick-walled, inelastic blood vessels. The risk of haemorrhage is related to the size of the tumour, and is significantly higher in lesions greater than 4 cm in diameter. We herein present a case of renal AML in a 60-year old female patient, with extension into left renal vein and infra diaphragmatic segment of IVC. There are only few cases of renal AML reported in the literature to date which shown extension into left renal vein and infra diaphragmatic segment of IVC.

### Case report

A 60-year old female patient, presented with left sided abdominal pain since last 6 months. The patient underwent an ultrasound examination which revealed a well defined lobulated, eccentrically located hyperechoic mass sized ~8 cm arising from the inter and lower polar regions of left kidney also a hyperechoic content noted in the left renal vein and in the renal, suprarenal and hepatic segments of IVC. The IVC and left renal vein shown no flow on Colour Doppler examination. Furthermore, computed tomography (CT) examination revealed a large well defined eccentrically located mass of mixed density measuring ~7.6×6.8×8.2 cm with surrounding preserved fat planes. The lesion showed an attenuation value of about -80 to 120 HU. Normal excretion of contrast noted from both kidneys suggesting preserved renal function. Also the lesion is seen extending to the left renal vein and to the renal, suprarenal and hepatic segments of IVC. Few subcentimetric left paraaortic, central mesenteric and terminal ileocolic region lymphadenopathy noted. There were no symptoms or radiological findings suggesting tuberous sclerosis complex (TSC) in the patient. The adrenal glands bilaterally and the right kidney appeared to be normal. On MRI the lesion showed India ink artefact on chemical shift imaging, which is suggestive of fatty origin of the lesion, also shown fat suppression on fat saturation images. Following abdominal laparotomy mass was found to originate from the left kidney with extension into left renal vein and infra diaphragmatic segment of IVC.

The histopathological examination revealed that the lesion consisted of mature adipose tissue, thick-walled blood vessels and smooth muscle in different proportions, and originated from the left kidney. Immunohistochemical staining of the tumour revealed that the tumour cells were positive for smooth muscle actin, human melanoma black-45 (HMB-45), melan-A and S-100. Based on these findings, the patient was diagnosed with renal AML, with extension into left renal vein and to the renal, suprarenal and hepatic segments of IVC.

## II. Discussion

Renal angiomyolipomas are benign hamartomas composed of blood vessels, smooth muscle, and fatty tissue. AML is seen in two distinct clinical forms, a sporadic form and in association with TSC. The sporadic form accounts for approximately 80% to 90% of cases of AML.<sup>1</sup> The sporadic form is typically seen in middle-aged patients (mean age, 43 years) and is more common in women by at least a 4:1 ratio.

When found in children, AMLs are almost exclusively associated with TSC.<sup>2</sup> Masses less than 4 cm in diameter are usually asymptomatic, but those larger than 4 cm often cause flank pain, hematuria, and anaemia and carry a significant risk of intratumoral or perinephric haemorrhage.<sup>3,4</sup> CT usually demonstrates a fatty mass intermixed with areas of increased tissue density, although the amount of fat present is variable and it can even be absent.<sup>5</sup> The presence of intratumoral fat is almost diagnostic of AMLs,<sup>6</sup> although in rare cases, fat has been reported in Wilms' tumor in children<sup>7</sup> and in renal oncocytomas<sup>8</sup>. Retroperitoneal lymph node involvement may be discovered in patients with AML<sup>9</sup>. AMLs are always benign. However, the tumours may exhibit extrarenal extension. Large perinephric tumour components may be found and AMLs may extend into the renal vein and IVC<sup>2</sup>.

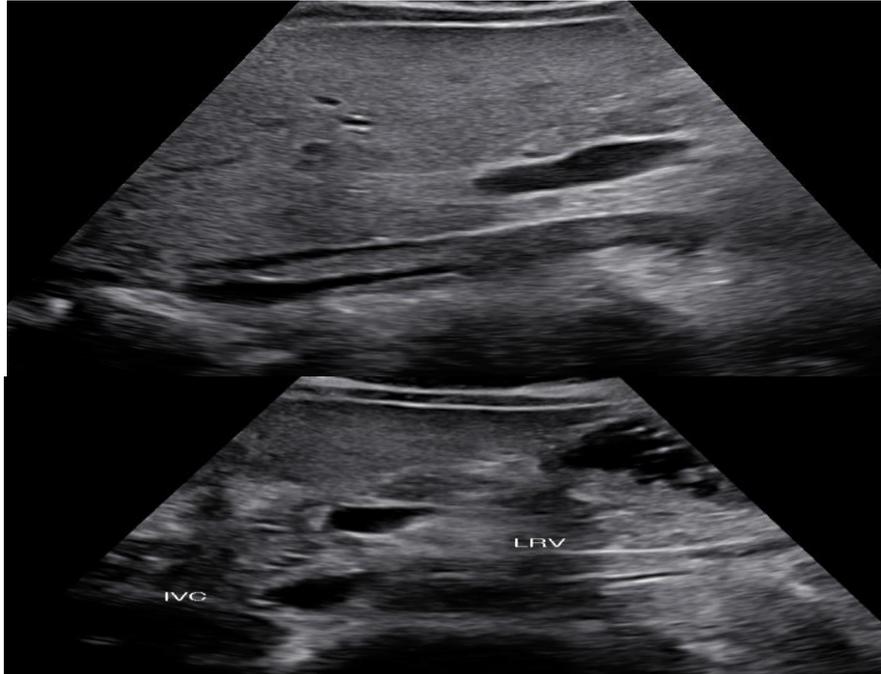
MRI may also detect the presence of fat in renal AMLs. The MRI characteristics of AML depend on the relative amounts of fat and other tissue components. The presence of macroscopic fat in a lesion is very specific for AML, although rarely RCC can also contain fat.<sup>10</sup> AML is identified on opposed-phase chemical-shift MRI by the presence of India-ink artefact (loss of signal at the interface of fat and non-fat-containing areas) at the interface of mass and renal parenchyma, or within the renal mass.<sup>11</sup>

Surgery and biopsy are rarely needed in asymptomatic patients with typical imaging findings of AML. Generally, asymptomatic patients with an AML approaching 4 cm in diameter are followed annually with CT or US, although in most of these patients the lesions show little change over time. Patients with lesions larger than 4 cm can be evaluated at shorter intervals.

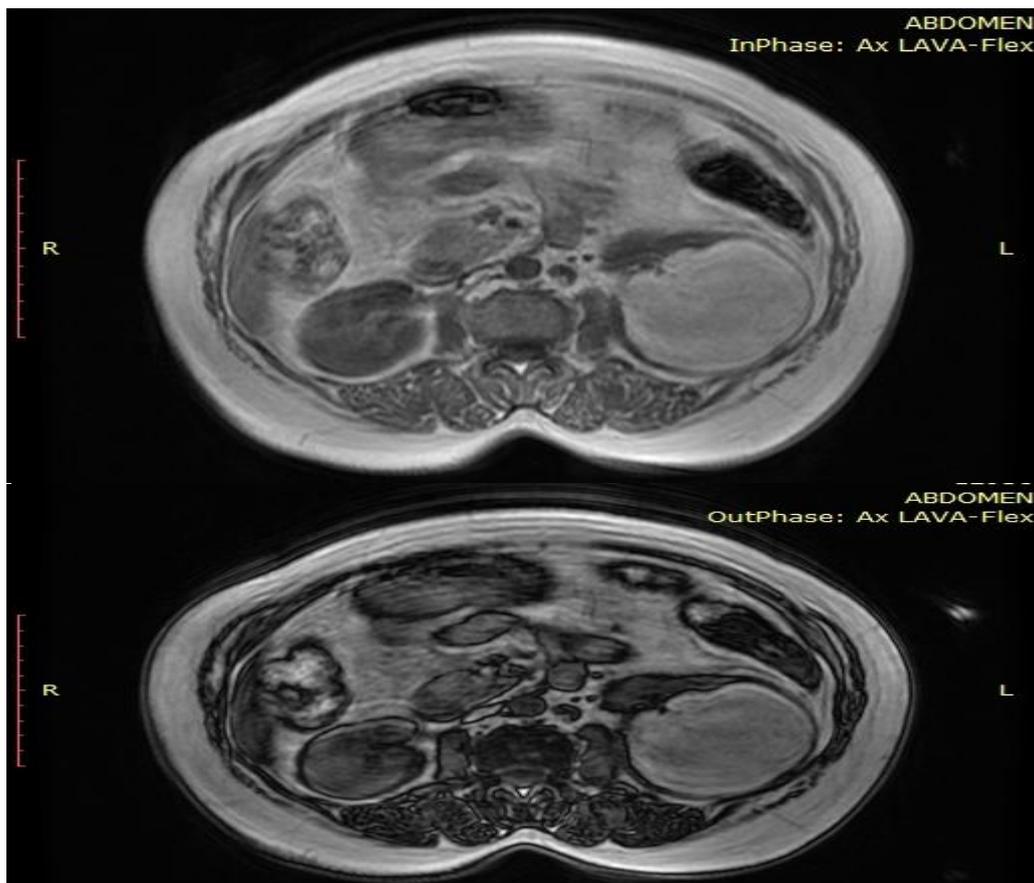
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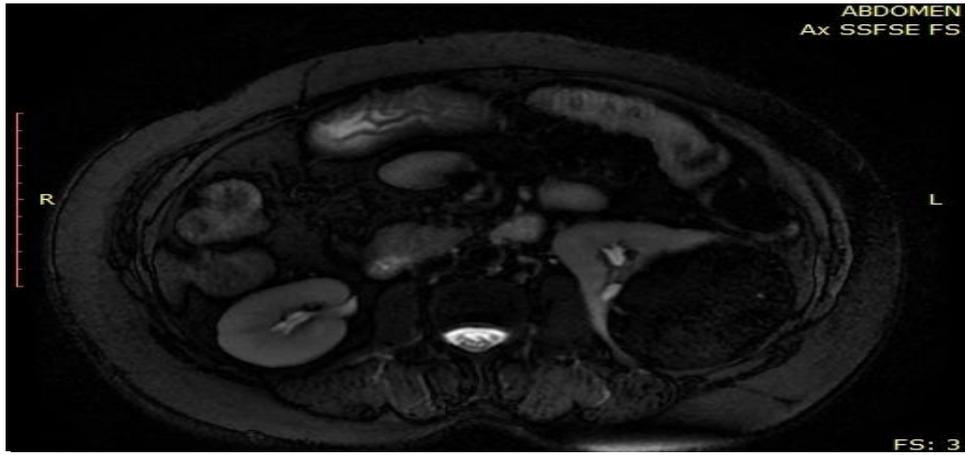




USG images showing a well defined hyperechoic lesion in the interpolar region of left kidney. Also the hyperechoic content is seen in the left renal vein and IVC. Suggestive of extension.



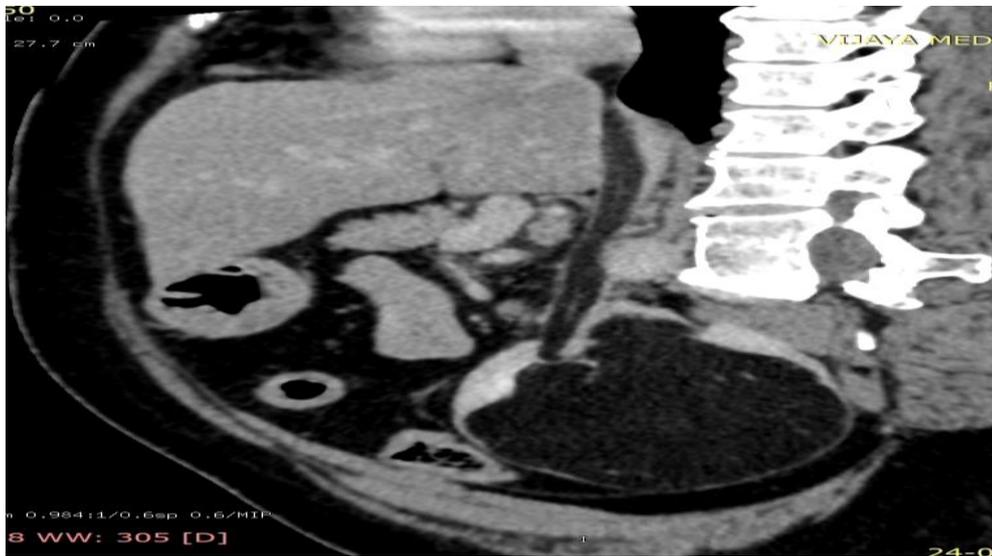
Chemical shift imaging of the lesion showing India ink artefact in opposed phase imaging. Suggestive of a fatty origin of lesion.



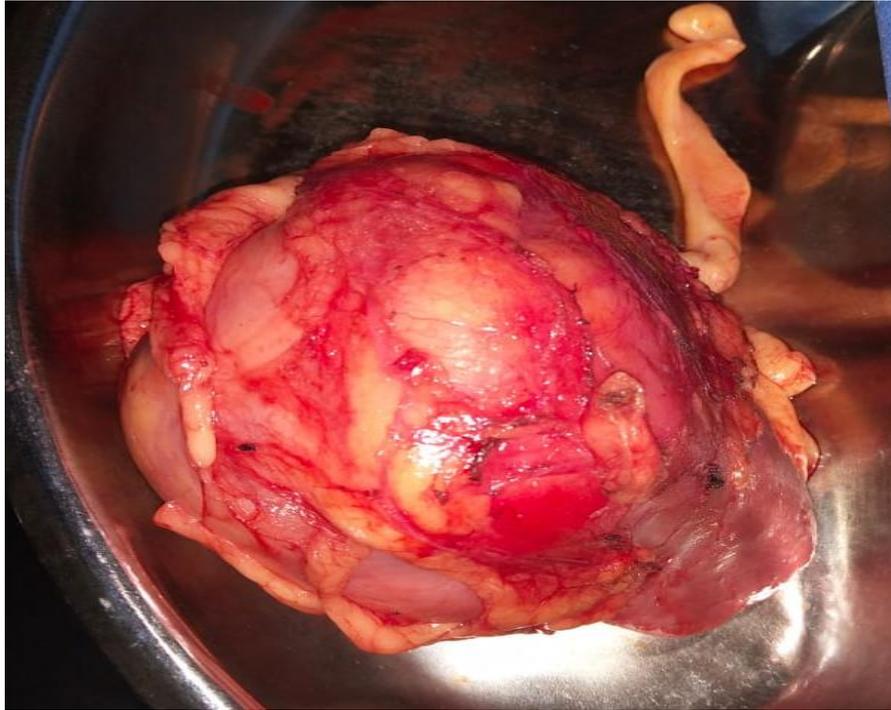
On Single Shot Fast Spin Echo Fat Saturation (SSFSE FS) imaging the lesion showed fat suppression.



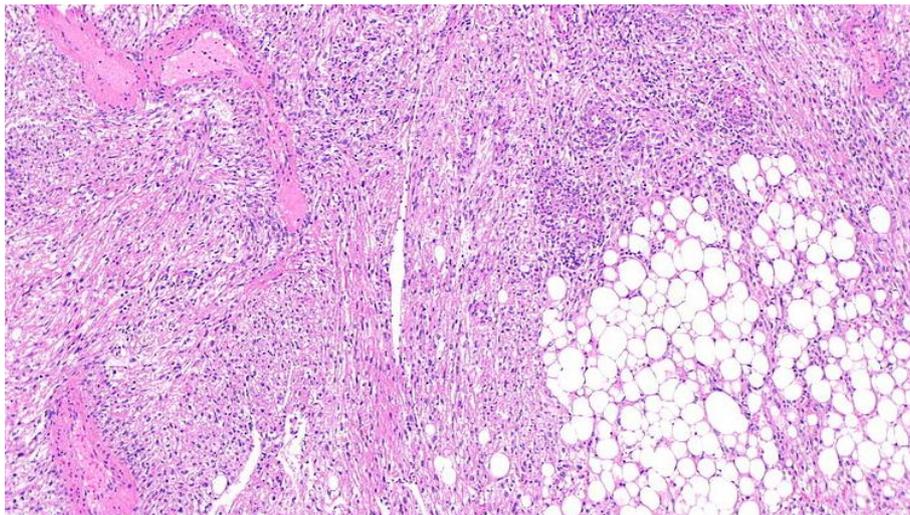
CECT abdomen showing a well defined fat attenuating lesion of the left kidney with extension into left renal vein.



Sagittal reformatted CT image showing well defined fat attenuating lesion of the left kidney with extension into IVC.



Surgical specimen after resection.



Micrograph showing fat, hyalinised vessels and spindle cells. Suggesting angiomyolipoma.

Dr.M.N.Jyothi" Case Report A Rare Case of A Renal Angiomyolipoma: With Extension into the Renal Vein and Ivc."IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 18, no. 7, 2019, pp 48-52.