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Giant Exophytic Renal Angiomyolipoma Mimicking as Retroperitoneal Sarcoma; A Case Report with Review of Literature

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A 30-year-old woman was admitted with a 7 month history of dull pain and palpable lump in left side of abdomen. No other significant history noted. Abdominal examination revealed mass in left side of abdomen measuring 25 x 20 cm, extending from left hypochondrium up to pubic symphysis, mass was smooth, firm, non-tender with well defined margins. Hemogram and renal function tests were within normal limits. Contrast-enhanced computed tomography (CECT) of abdomen revealed large well defined mass in left side abdomen with predominance of fat (-69 HU) measuring 23 x 20 x 12cm, extending from epigastrium to pelvis. It also showed multiple enhancing vessels within the lesion and an ill-defined interface with lower pole of left kidney [Table/Fig-1a-d]. Chest X-ray was normal.

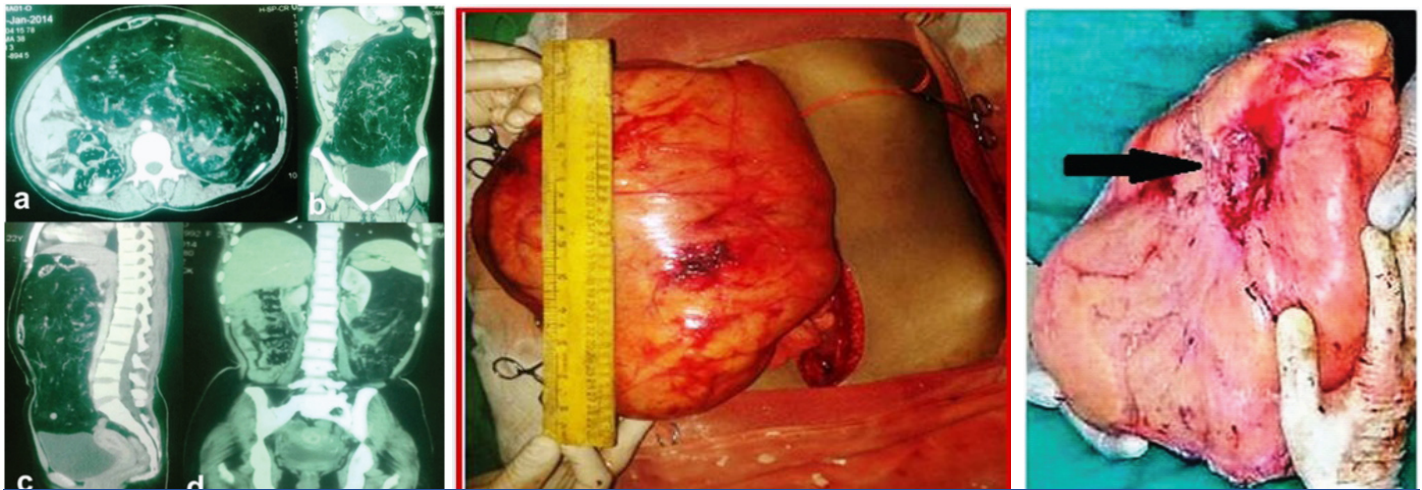
With a preoperative diagnosis of retroperitoneal liposarcoma, the patient was planned for exploratory laparotomy. At laparotomy, large bosselated mass was found arising mainly from lower pole of kidney [Table/Fig-2]. No regional or distant metastases were noted. Patient underwent partial nephrectomy with enbloc resection of the mass. Gross examination of the tumor showed a well encapsulated soft tissue mass measuring 30 x 25 x 12 cm and weighed around 8000 grams, with lower pole of kidney identified at its upper aspect [Table/Fig-3]. Histologically, the tumor was made of mature adipose tissue, numerous thick walled poorly organised blood vessels and smooth muscles [Table/Fig-4a&b]. Final diagnosis of angiomyolipoma was made. Patient is disease free at 14 months of follow up.

Angiomyolipoma (AML) is an unusual benign neoplasm of kidney composed of variable mixture of fully differentiated smooth muscles and mature adipose tissue. It accounts for 0.3% to 3% of all renal tumors [1]. The first case of “Angiomyolipoma” was originally described by Fischer in 1911 and term “Angiomyolipoma” was coined by Morgan in 1951. Initially, AML was considered to be a form of hamartoma or choristoma. Recent evidence suggests AML to be of monoclonal origin, possibly derived from perivascular epitheloid cells. Tumor shows strong positivity for estrogen and androgen receptor suggesting strong hormonal influence in its aetiology.

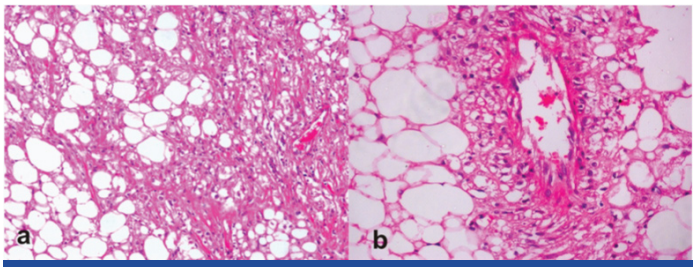
Majority of AMLs occur in females and are rare before puberty. AMLs can occur sporadically or in association with tuberous sclerosis, which accounts for 45 to 80% of cases. Typical sporadic AML is usually unilateral, asymptomatic and an incidental finding. Majority of sporadic AML occur in middle aged women (4th or 5th decade) [2]. Tuberous sclerosis associated AML is seen in younger age (mean age is 30 years), multiple, bilateral, less female to male predominance and are usually symptomatic.

Occasionally, AML have unusual extrarenal occurrence and is reported in hilar lymph nodes, retroperitoneum, liver and direct extension into venous system. Around 25% of AML’s have predominantly exophytic growth, extending extrarenally into perirenal spaces. Majority of patients were asymptomatic. Classical presentation includes flank pain, palpable mass and gross hematuria. Most important complication of AML is massive retroperitoneal haemorrhage (known as Wunderlich syndrome), noted in up to 10% of patients. The Wunderlich's syndrome is clinically characterized by the Lenk's triad which are acute flank pain, a flank mass and hypovolemic shock [3]. AML have a benign clinical course, with slow growth pattern and lack of distant metastasis.

Most AMLs are incidental finding on imaging and may resemble malignancy. Ultrasonography (USG) reveals a well circumscribed, hyperechogenic lesion with shadowing effect are classical findings that help to differentiate from renal cell carcinoma. AML appears to be diagnosed accurately on CECT scan by presence of fat within a renal lesion (confirmed by a value of –20 to -80 HU on imaging) [4]. About 5% of AMLs do not show the fat attenuation on imaging and hence cannot be differentiated from renal cell carcinoma [4].



[Table/Fig-1a-d]: CT Scan revealed large well defined mass in left side of abdomen, extending from epigastrium to pelvis with predominance of fat (-69 HU) measuring 23 x 20 x 12cm
[Table/Fig-2]: Laparotomy revealed large bosselated mass was found arising mainly from lower pole of kidney
[Table/Fig-3]: Gross examination of the tumor showed a well encapsulated soft tissue mass measuring 30 x 25 x 12 cm and weighed 8000 grams, with lower pole of kidney identified at its upper aspect (arrow mark)



[Table/Fig-4a&b]: Microscopy revealed tumor made of mature adipose tissue, numerous thick walled poorly organised blood vessels and smooth muscles

Angiography may show aneurysmal dilatation in 50% of AML and the size of aneurysm correlates with increased risk of rupture. Despite characteristic radiologic findings of AML, diagnosis can be confused in following three conditions: liposarcoma, fat poor AML resembling renal cell carcinoma and possibility of fat containing renal cell carcinoma. Giant exophytic AML may resemble a retroperitoneal sarcoma. It may be difficult to distinguish exophytic AML from a well differentiated liposarcoma, as both are fat containing tumours' and have similar appearances on imaging. Liposarcomas usually compress or extrinsically push the renal parenchyma whereas AML may cause a small indentation of the renal parenchyma and presence of enlarged vessels. Strong positivity for HMB-45, a monoclonal antibody is highly characteristic of AML and can be used to differentiate it from sarcoma. Surgical treatment includes tumour excision with or without nephrectomy, nephron sparing surgery or arterial embolisation based on tumor size, extent and its clinical presentation. Role of ablative therapies such as radiofrequency ablation and cryoablation is not well established and might be considered in multicenteric tumors or elderly patients with comorbidities.

AML is a rare benign neoplasm arising from perivascular epitheloid cells. Majority of AMLs are asymptomatic and can occur in association with or without tuberous sclerosis. These are detected incidentally on imaging and demonstration of fat within the tumour by computed tomographic imaging (-20 to -80 HU units) is thought to confirm the diagnosis of AML. Diagnosis of exophytic AML should always be considered before making a diagnosis of retroperitoneal liposarcoma, as they have similar radiologic features but differ in prognostic value and treatment approach.

ABBREVIATION

AML- Angiomyolipoma, USG- Ultrasonography, HU- Hounsfield units CT- Computed tomography, HMB- Human melanocyte B antigen.

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