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Renal angiomyolipoma: a case report

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CASE REPORT

ABSTRACT

Angiomyolipoma (AML) is a rare benign tumor composed of adipose tissue, smooth muscles and blood vessels with an incidence is 0.3-3% which is predominantly found in females. These tumors have a strong association with tuberous sclerosis but can occur sporadically. Clinical importance lies in their susceptibility to spontaneous haemorrhage. They can be unilateral or bilateral. Interestingly these tumors occur more in right kidney. We present a case of sporadic unilateral angiomyolipoma of left kidney in a 55 yrs old female who had complaints of off and on left lumbar pain. She was subjected to left nephrectomy and diagnosis was confirmed on histopathology.

Key Words

Angiomyolipoma(AML), kidney tumour, renal angiomyolipoma.

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INTRODUCTION

Angiomyolipomas (AML's), as per world health organization (WHO) classification of tumors are defined as tumors composed of mature adipose tissues, dysmorphic blood vessels and smooth muscle arising from the mesenchymal element of the kidney.[1] In 5% of tumors, fatty elements can be detected at microscopy. It is a rare benign tumor with an incidence of 0.3-3%.[2] Nowadays more cases are being detected due to advances in the imaging modalities. Predominantly found in females with a MALE: FEMALE (M:F) ratio of 1:4 and this shows a hormonal component to the growth of the tumor. Approximately in 80% cases, AML affect the right kidney.[3] In the past, AML lesions were considered as hamartomas due to its triphasic nature but recent clonality studies suggest them to be classified as neoplasms.[4] These lesions can lead to spontaneous haemorrhage which makes them clinically important.[5] Most common site of AML is kidney but they may occur at extra renal sites also including liver, lungs, lymph nodes and retroperitoneal soft tissues.[6]

CASE REPORT

A 55 yrs. old female presented with complaints of gradual abdominal distention since 8 yrs and off & on long standing pain in left flank region which rapidly increased during last 3 months. She had loss of appetite and was having general weakness. There was no history of Diabetes Mellitus, hypertension, pulmonary tuberculosis or any thyroid dysfunction which was confirmed by respective lab tests and imaging modalities. In the past she was managed by general practitioners but was not relieved. Physical examination revealed a healthy woman. No pallor or icterus. Abdominal examination elicited no bowel sounds and a large mass in the left hypochondrium extending to occupy left lumbar region with soft abdomen. Suspicion of massive splenomegaly or a renal mass, clinically the case was diagnosed as lump abdomen? spleen or renal mass. The case was then referred for the surgical evaluation. The patient was further investigated by radiological/biochemical investigations. Intra venous urography (IVU) was performed which showed a normal functioning right kidney. The pelvicalyceal system of the left kidney was poorly delineated by contrast. The poor function was interpreted by less excretion of the dye (fig.1).

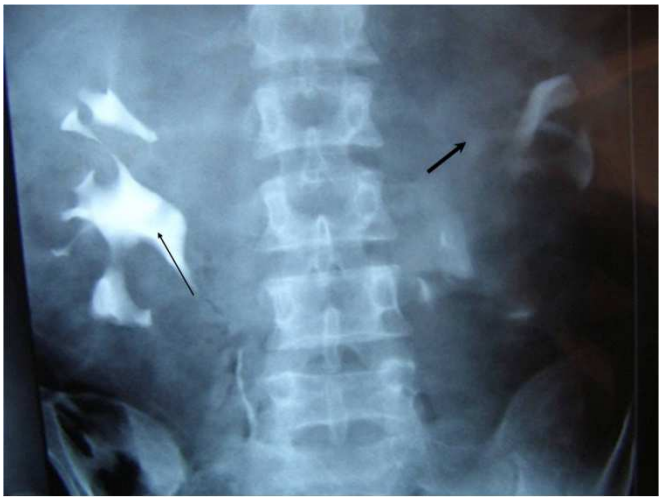

Fig. 1. IVU image with distorted pelvicalyceal system more on left side (Arrow head) as compared to right (Arrow) due to the mass effect of low density lesions

Ultrasonography (USG) abdomen confirmed a left renal mass with mostly echogenic areas of fat in the mass. Computed Tomography (CT) scan exhibited well defined regular fat density in a large retroperitoneal mass measuring 28 x 17 x 13 cms occupying the left renal fossa and almost half of the abdomen. There was a single renal artery on both sides (fig.2).

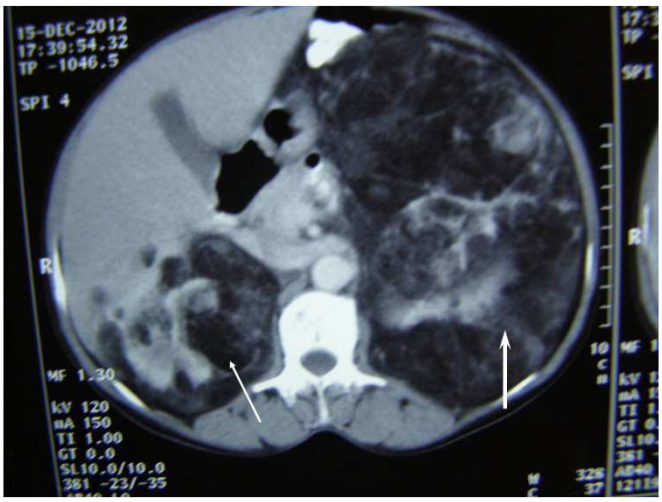

Fig. 2. CT scan image with large fat density retroperitoneal mass occupying left renal fossa (Arrow head) and similar small area in right kidney (Arrow)

There was another smaller fat density lesion seen in right kidney. The radiological findings were suggestive of renal angiomyolipoma. Patient was subjected to laprotomy. On exploration a huge retroperitoneal fatty mass was found occupying the left hypochondrium with extension into left iliac fossa. The left kidney was not visible and only part of ureter could be identified. A huge tortuous renal vein 3cms in diameter was seen. The left renal artery had many branches embedded in the lipomatous tumor. The spleen was normal and found pushed posteriorly. The tumor as a whole with left kidney along with part of ureter and renal vessels was resected out. The specimen was sent to histopathological examination. Post operatively the patient condition was uneventful. The specimen on gross examination appeared as a globoid well encapsulated lipomatous mass which was lobulated and surface was nodular. It weighed 1.5kgs and measured 26.0 x 18.0 x 15.0 cms. Sub capsular areas of haemorrhage seen at places. (fig.3 A & B).

Fig.3 A. Specimen of renal mass with ureter

Fig.3 B. Specimen of renal mass with ureter

The renal pelvis was identified on dissection deep in the tumor substance and was markedly dilated. A few solid tiny areas of renal tissue identified. Other structures such as ureter, renal artery were not seen in the hilum (fig.4).

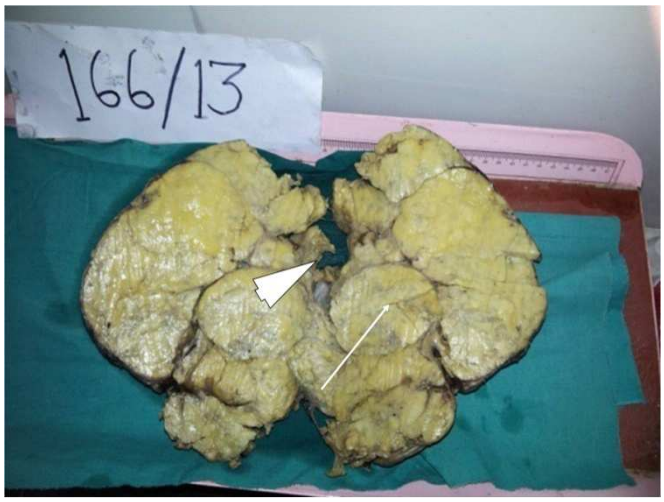

Fig. 4 Cut section of specimen showing renal cortex (Arrow head) & lobulated/lipomatous area (Arrow)

On microscopy, the entire renal parenchyma was found to be infiltrated by the lipomatous nodular mass leaving behind a few solid area showing normal renal structures of glomeruli, tubules, collecting ducts, interstitium and vessels. Few areas of thyroidisation of tubules are also seen. The lipomatous nodules showed triad of tissues comprising predominantly of mature fat cells, muscular arteries haphazardly arranged and proliferating smooth muscle bundles with abundant granular cytoplasm and large spindly vesicular hyperchromatic nuclei. No mitosis or malignancy noticed. The pathological findings were consistent with Renal Angiomyolipoma (fig.5 A, B & C).

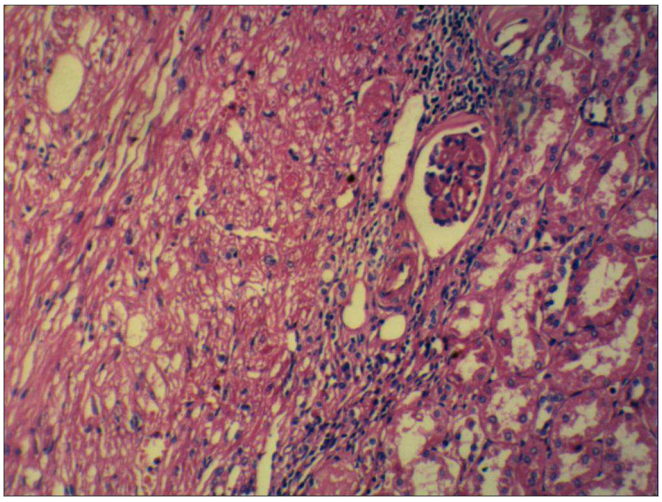

Fig. 5 A

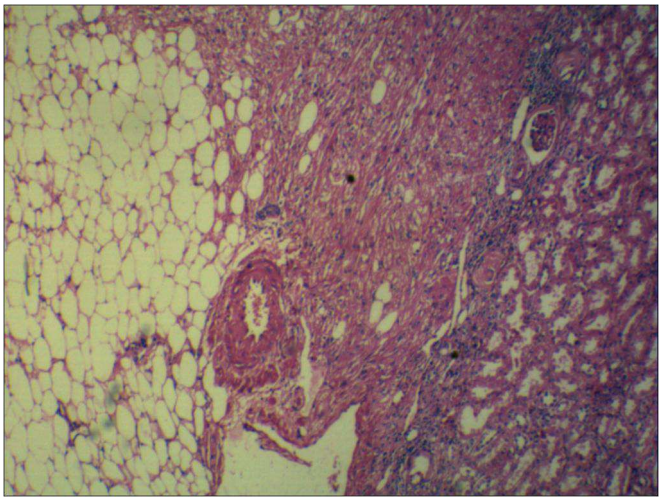

Fig. 5 B

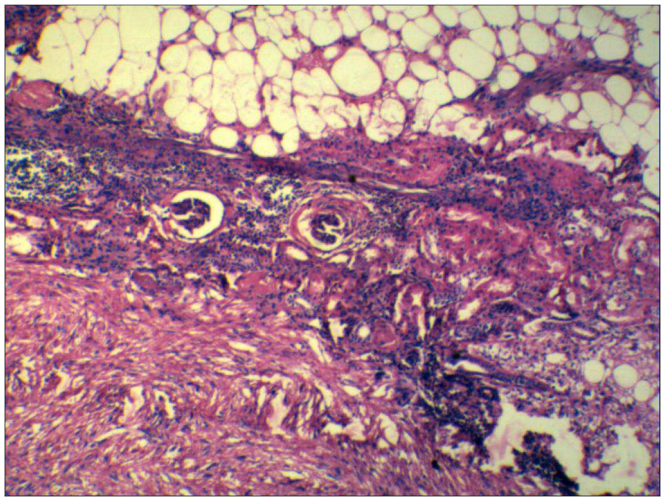

Fig. 5 C

Fig. 5 A, B & C. Photo micrographs of left renal mass microscopy showing mature fat cells, muscular arteries haphazardly arranged and proliferating smooth muscle bundles with abundant granular cytoplasm and large spindly vesicular hyperchromatic nuclei. (H & E Staining, 100X magnification)

DISCUSSION

Angiomyolipomas are rare benign tumors composed of fat, smooth muscles and varying number of thick walled tortuous blood vessels.[7] These tumors have an incidence of 0.3-3% and it is estimated that 10 million people worldwide have such lesions.[2] These lesions should be considered as a choriostoma, a disordered arrangement of mature tissue appearing at a site where that tissue should not be present normally.[8] AML is classified into 2 types: (a) Isolated AML (b) AML associated with tuberous sclerosis. Sporadic AML which accounts for 80% AML usually present in 4th-6th decade of life. Lesions are unifocal & larger with a M:F ratio of 1:2. AML associated with tuberous sclerosis accounts for 20% AML and present in a younger age before 3rd decade of life. These lesions are multifocal (87%) & bilateral (71%) with a M:F ratio of 1:1,[3,9] seen in 50-80% of patient with tuberous sclerosis.[8] Bourneville & Brissard in 1880 first described association of AML with tuberous sclerosis.[10] Tuberous sclerosis is an autosomal dominant neurocutaneous disorder. Clinical features are hamartomas, tumors of brain (subependymal giant cell tumor), lung (lymphangioleiomyomatosis), heart (rhabdomyomas), kidney (AML), retina, skin (cutaneous angiofibromas) and mental retardation, infantile/childhood seizures caused by alteration in tuberous sclerosis complex (TSC1 & TSC2 gene).[11] All patients with multiple AML should be evaluated for tuberous sclerosis as the association is very high. Beside tuberous sclerosis, AML also seen in other hereditary disorders like Von Hippel Lindau & Von Recklinghausen syndrome, autosomal polycystic disease.[8] Morphologic variants of AML are seen which are due to variations in the relative proportion of various components. Different variants of AML are reported as classical, epitheloid, oncocytoma like, AML with epithelial cyst, Leiomyoma like and

liposarcoma like AML.[4] AML with spindle cells of smooth muscle resemble leiomyosarcoma or gastrointestinal stromal tumor (GIST) which can be specifically diagnosed by immunopositive of CD117. If adipose tissue predominate it can resemble atypical lipomatous tumor and diagnosis can be achieved by low molecular weight keratin (CAM5.2, MAK6, CK7, CK18).[12] AML rich in epitheloid cells can resemble spindle cell renal cell carcinoma (RCC). Apitz described a variation in smooth muscle component a cell type with epitheloid features called Perivascular Epitheloid Component (PEC). AML is now considered to be a part of the PEComa family.[4]

AML is asymptomatic in majority of cases but in minority symptoms can be present as seen in our case where the symptom was pain in left lumbar region. Presence of symptoms depends on the tumor size which varies from few mm to 20 cms. Symptoms include flank or abdominal pain (53%), palpable tender mass (47%) & gross hematuria (23%) called "Lenk's Triad".[2,9] Flank pain is the most common complaint in symptomatic patient. AML grow rapidly during pregnancy due to the hormonal effect.[8] Anemia, fever, hypotension, renal failure can also be present in few patients.[7] In AML, blood vessels generally lack elastic tissue which predisposes the patient to aneurysm formation & spontaneous hemorrhage which is the most dangerous complication.[3] This retroperitoneal haemorrhage is called Wunderlich syndrome which mainly occurs during pregnancy and 20% of these patients present with haemorrhagic shock.[13] The 2nd most common cause of a spontaneous haemorrhage in kidney after RCC is spontaneous rupture of renal AML but it is rare & is seen in only 5-10% of cases. [14]

Till 1965, a review of literature revealed that only 150 cases of AML were detected which were not diagnosed preoperatively.[9] But now with availability of USG & CT scan, it is possible to accurately diagnose renal AML as due to fat content, these tumors have a characteristic appearance on CT. IVU is not diagnostic of AML although low density areas may suggest the possibility of this tumor. IVU findings may sometimes overlap with those of polycystic renal disease. CT is the modality of choice to diagnose AML. On angiography tumor may show high vascularity.[15] All AML show immunopositive for HMB-45, A-103 & actin,[6] which confirms the clinical diagnosis on histopathological examination. Our case was purely a histopathological diagnosis and no immunohistochemistry (IHC) was performed.

Management is aimed at preserving renal parenchyma & prevention of haemorrhage.[7] Renal AML can be classified into 4 categories for management protocol:-

(a) Asymptomatic lesion <4 cms. - Observation with CT scan follow up yearly.

(b) Asymptomatic lesion >4 cms. - Observation with CT scan at 6 months interval

(c) Symptomatic lesion <4 cms. - Conservative surgery is indicated

(d) Symptomatic lesion >4 cms. - Early partial nephrectomy is the treatment of choice[9]

First line of management in these cases is angiography & selective transarterial embolisation which is safe, reliable and well tolerated with few long term sequeal.[2,8] Anticoagulant therapy is contraindicated in a known case of AML. Nephrectomy in AML (partial or radical) is indicated in case of persistent haemorrhage, suspicion of malignancy, failed transarterial embolisation. Regional lymph nodes may get involved but it is regarded as an expression of multicentricity of lesion. If malignancy is suspected then exploration of retroperitoneum is suggested in case of AML.[16] A recent study shows that presence of ≥3 of the features are predictive of malignant behavior of AML: (a) ≥2mitotic figures/hpf, (b) atypical mitotic figures, (c) ≥70% atypical epithelial cells (d) necrosis.[4]

CONCLUSION

All AML, by enlarge are easy to diagnose histomorphologically and radiographically. Most of these are benign and asymptomatic, therefore needs no treatment. In the presence of haematuria, or evidence of extension in inferior vena cava or malignancy with lymphnode metastasis, needs surgical excision. The moto of treatment is to conserve the nephrons in the absence of indication for surgical resections.

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