ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 04 | April - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

GIANT RENAL ANGIOMYOLIPOMA WITH EPITHELIAL CYST IN YOUNG WOMAN CAUSING ABORTION: A RARE CASE REPORT

General Surgery

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ABSTRACT

Angiomyolipoma with epithelial cysts (AMLEC) is an uncommon subtype of kidney angiomyolipoma. AMLEC has a distinct histological and immunohistochemical staining pattern, which helps in the pathological diagnosis. We present a rare case of AMLEC in a 20-year-old female, associated with pregnancy lost, which was provisionally diagnosed as Retroperitoneal Liposarcoma on radiology and on histopathological examination diagnosed as AMLEC. She underwent surgical intervention to relieve her symptoms and successful possibility of upcoming pregnancy.

KEYWORDS

Angiomyolipoma, Neoplasms, Kidney, Retroperitoneal Liposarcoma, Diagnosis, Differential

Case Report

A 20-year-old female presented with pain in the left lumbar region. She aborted one month ago with her first 8 months old pregnancy. Abdominal swelling and pain after abortion continuously felt and day to day activities were painful. No history of family with given mass and no history of any substance abuse or other medical condition beside.

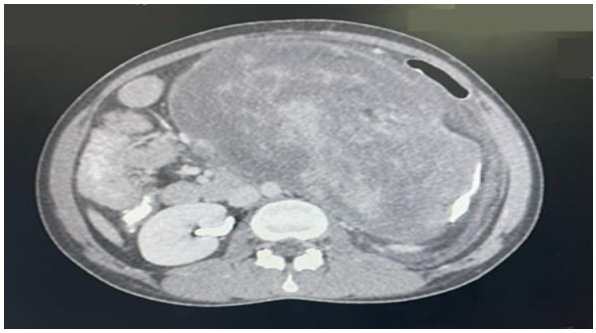
She had no history of haematuria, awareness of lump, tuberous sclerosis or intake of hormonal supplements. On examination, the abdomen was soft with fullness in the left renal angle and tenderness on deep palpation.

She underwent a contrast-enhanced computed tomography scan (CECT), which revealed a 14 x 16.5 x 21 cm soft tissue mass involving the upper two-thirds of the left kidney. The mass compressed the left renal vein, sub-hepatic inferior vena cava and part of the portal vein. Radiologically, it was provisionally diagnosed as large Retro peritoneal Liposarcoma.

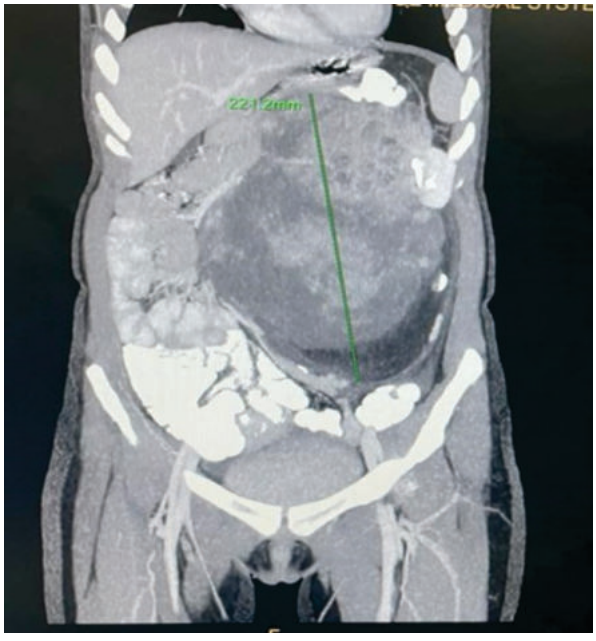
Her urinalysis showed only occasional red blood cells and urine culture was sterile and the routine laboratory investigations were within normal limits.

She underwent an Exploratory Laparotomy and on gross examination received distorted, large Retroperitoneal mass adherent to left kidney specimen with attached fat measuring 9x5 cm. A large soft multinodular partially capsulated globular soft tissue piece measuring 24x15x12 cm.

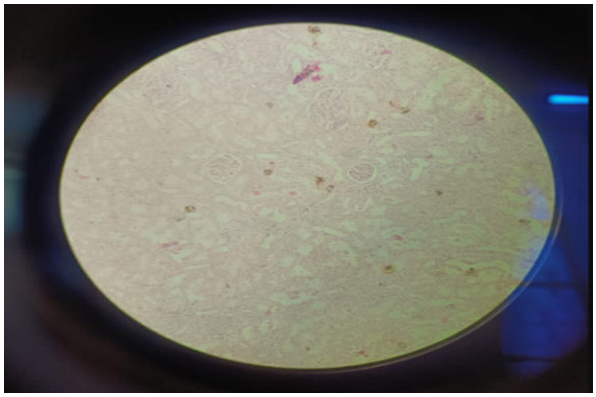
Thus, a histopathological diagnosis of renal angiomyolipoma with epithelial cysts (AMLEC) was made. At a 6-month follow-up, she is doing well with no complaints.



CT image showing axial view of the mass



CT image saggital view showing mass



histopathological slide of renal angiomyolipoma with epithelial cysts

DISCUSSION

Renal angiomyolipoma (AMLs) are the benign renal tumour. [1]. They are highly vascular, primarily composed of smooth muscle and adipose tissues, and are distinguished by perivascular epithelioid differentiation. Renal AMLs are usually solid neoplasms with multiple histological subtypes such as inflammatory, epithelioid, and some with a predominance of either muscle, adipose tissue or vessels. Apart from this, they may have a cystic change, particularly in large-sized tumour due to tumour necrosis or intra-tumoral haemorrhage. However, renal AMLs with true epithelial cysts have rarely been described in the literature. [2]. Although most of these tumours are found incidentally on radiological imaging, symptomatic presentations also exist. Typical symptoms would include flank pain, gross haematuria, and retroperitoneal haemorrhage. [3]. Imaging plays a central role in the diagnosis and management of renal angiomyolipoma. [4]. The tumours contain variable amounts of the same three tissue elements (vascular, muscular, and adipose). Variations will have different pathological, radiological, and clinical features. [5]. The fundamental diagnostic criterion of classic angiomyolipoma is detecting a large amount of adipose tissue on radiological imaging. [6]. Although considered benign, the tumours may extend into the surrounding perirenal fat or renal sinus and nearby organs and lymphatics. [7]. There are also rare, isolated reports of tumour thrombi extending through the renal vein into the vena cava. [8]. The management of angiomyolipoma is generally based on the clinical manifestations, tumour size, number, growth pattern, and malignant potential. [2].

Renal angiomyolipoma are rare kidney tumours that account for only 0.3 to 3% of all renal neoplasms. [9]. The overall prevalence rate of renal angiomyolipoma ranges from 0.13 to 2.2%, with over eighty percent isolated and sporadic cases. [10].

Angiomyolipoma are generally considered benign and will only very rarely become malignant. [11]. The majority of angiomyolipoma are asymptomatic and tend to remain stable in size. Only about 9% tend to enlarge over time. [12]. Larger tumours (>6 cm) are more likely to enlarge and grow, haemorrhage, and cause symptoms. [12]. Here her symptoms were not relieving and caused abortion. She underwent surgical intervention.

CONCLUSION

Though AMLEC is a rare entity, it should be considered in the differential diagnosis of Retroperitoneal Liposarcoma. Our study highlighted a rare case in an Indian woman who recently lost her first pregnancy and suffered abdominal pain and doubts about her upcoming pregnancy. Proper work up towards her clinical manifestations, aided with imaging, we considered her for surgery with explained risk. Patient managed surgically and later on diagnosed with histopathological, we impeded her upcoming pregnancy lost, preserved fertility and quality of life. At a 6-month follow-up, she is doing well with no complaints.

Footnotes

This study was carried out at the Mahatma Gandhi Hospital, Jodhpur, Rajasthan.

Ethics Statement: The authors retain informed consent signed by the patient herself for authorizing the data publication and the manuscript is as by the Institutional Ethics Committee rule.

Financial support: The authors declare that no financial support was received.

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