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Case report

ANGIOMYOLIPOMA OF A KIDNEY: A CASE REPORT

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ABSTRACT

Angiomyolipoma is a rare benign tumour of the kidney. They are composed of abnormal vasculature, smooth muscle and adipose elements. They may be associated with Tuberous sclerosis. The unilateral presentation is uncommon. Angiomyolipoma is presented as an enlarge lump in the abdomen may mimic malignant lesion such as renal cell carcinoma, yet malignancy has to be ruled out. The diagnosis of this condition may not be straight forward with imaging alone. Excision is recommended for definite histopathological diagnosis (in symptomatic patients) and to prevent the potential risk of haemorrhage and malignancy. A case of benign giant Angiomyolipoma is presented here because of its uncommon occurrence.

Keywords: Angiomyolipoma , Unilateral, Kidney

INTRODUCTION

Angiomyolipoma is a rare benign tumour of the kidney. It is found in approximately 45-80% of patients with tuberous sclerosis and are typically bilateral and asymptomatic. In patients without tuberous sclerosis, renal angiomyolipomas can be unilateral and tend to be larger.¹ Angiomyolipoma is a benign clonal neoplasm consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels. It is most likely derived from the perivascular epithelioid cells, and its growth may be hormone dependent. It shows female predominance and rarity before puberty.² It may mimic renal cell carcinoma. Diagnosis is done with the help of ultrasonography, CT scan and histology.

CASE REPORT

A 75 yr. an old woman from rural area presented with lump in right side of the abdomen that the patient had noticed 1 year previously. (Bharati Hospital). The lump had gradually increased in size over a period of time. She had complaints of pain since 1 day. On

examination, she was anaemic (8.3gm %) with large retroperitoneal lump that occupied right side of abdomen, the lump was firm in consistency and tender. She had no lymphedema or other palpable lymph node. Ultrasonography revealed a well defined echoic lesion noted of size 14.6 X 8.7 cm, associated with upper pole of the right kidney extending upto the epigastric region and midline on right side. CT scan revealed a large well defined mass lesion showing density with peripheral soft tissue component seen in right hypochondriac and right lumbar region measuring 13 X 11 X 10 cm. The mass is seen in medial to pancreas and (Inferior vena cava) displacing it superolaterally. Superiorly mass extending to liver and inferiorly to the right iliac fossa. The mass completely encasing right kidney and displacing it inferiorly, most likely liposarcoma. (figure 1) A malignant tumour was suspected based on the imaging and clinical features. In view of above finding and pain, malignancy needed to be ruled out. The patient underwent surgery and tumour was excised intact

(figure 2). Its weight was 1800 grams. A nephrectomy was done due to inability to differentiate it from renal cell carcinoma. 4 units of blood were transfused preoperatively and in addition 3 units were given (blood group AB positive) She made an uneventful recovery. An angiomyolipoma was confirmed by histopathological assessment. Post operatively, a single large well circumscribed an encapsulated firm mass measuring 20 X 15 X 9 cm weight 1800grams. The cut section appears yellowish brown with areas of hemorrhage (figure 3). Microscopy- tumour showed mature adipocytes, separated by fibrohyalinised septae showing vascular proliferation and area of smooth muscle cell in bundles (figure 4).



Fig 1: CT scan showing lesion



Fig 2 : Intra operative photograph

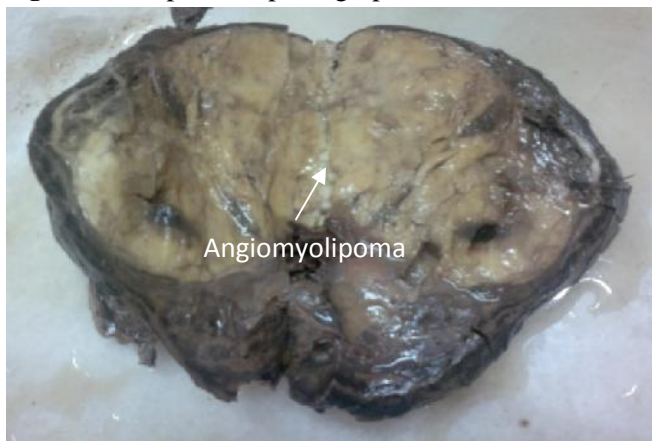


Fig 3 : Specimen showing renal Angiomyolipoma.



Fig 4: Histopathology - Angiomyolipoma

DISCUSSION

Angiomyolipoma is a rare benign tumour of the kidney. It is found in approximately 45- 80% of patients with tuberous sclerosis and are typically bilateral and asymptomatic with F: M predominance of 2:I. The mean age of presentation is 30 years. In contrast, of the 60 -70% of patient with AML who do not have tuberous sclerosis present later in life, during 5th or 6th decade and this tumour can be unilateral and tend to be larger than those associated with tuberous sclerosis. Tuberous sclerosis is an autosomal dominant disorder comprising adenoma sebaceum, mental retardation and epilepsy.¹

Angiomyolipoma consists of varying amount of mature adipose tissue, smooth muscle and thick walled vessels. It is mostly likely derived from perivascular epithelioid cell. Extrarenal occurrence have been reported in hilar lymphatics, retroperitoneum and liver and direct extension into the venous system.²

On diagnostic imaging, it may mimic a malignancy. On ultrasonography, it gives a well circumscribed, highly echogenic often associated with shadowing. On CT scan, well defined mass is seen (confined by a value of -20 to 80 Hounsfield units).²

Differential diagnosis for this are subtypes of sarcoma including fibrosarcoma, leiomyosarcoma, liposarcoma and renal cell carcinoma.³ Positive immunoreactivity for HMB-45 is characteristic for angiomyolipoma and can be used to differentiate it from sarcoma.^{4,5}

The patient with tumour with intermediate features or calcification should be managed proactively because the likely diagnosis in most such cases is renal cell carcinoma. Patients with isolated lesions less than 4 cm, can be followed up with a yearly CT scan or Ultrasonography to define the growth rate and clinical significance. Similarly, Patients with asymptomatic or mildly symptomatic lesions greater than 4 cm should

be followed up with semiannual ultrasonography. Patients with lesions greater than 4 cm with moderate or severe symptoms (bleeding or pain) should undergo surgical intervention either in the form of tumour excision with or without nephrectomy, renal-sparing surgery or renal arterial embolization.²

Complications are as follows, Retroperitoneal hemorrhage, hematuria, hypovolemic shock, hypertension, abscess formation.

CONCLUSION

Angiomyolipoma is a rare tumour of the kidney.⁵ They may be confused with malignant tumours especially when the presentation is unilateral. CT Scan and ultrasonography helps in diagnosis. Symptomatic patient requires surgical intervention.

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