

# Renal Angiomyolipoma – A Case Report

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## ABSTRACT

Angiomyolipomas are rare, benign tumours which are composed of an intimate admixture of blood vessels, smooth muscle cells and fat and hence the name. They occur at many sites, more commonly in the kidney. Usually they present as an incidental finding or with retroperitoneal haemorrhage in adults. They are seen in 25-50% of the patients with tuberous sclerosis.

A 29 year old female presented with a mass in the left loin with a dragging pain, which was incidentally found during a medical check up. She had no evidence of tuberous sclerosis. A nephrectomy was done. Microscopy revealed the classical features of angiomyolipoma, which comprised of mature adipose tissue, thick walled blood vessels and bundles of smooth muscle cells. No epithelioid component was seen.

**Key Words:** Angiomyolipoma, kidney tumours, tuberous sclerosis, epithelioid component

## KEY MESSAGE

- Conventional angiomyolipoma, the commonest variant among angiomyolipomas, has a benign behaviour, whereas the rare epithelioid variant has an aggressive behaviour.

## INTRODUCTION

Angiomyolipoma (AML) is an uncommon, benign, mesenchymal tumour (2-6.4% of all the kidney tumours) which arises from either the renal pelvis or the sinus, which is composed of an intimate admixture of blood vessels, smooth muscle cells and fat, hence the name. AML commonly occurs in women who are of the age group of 40- 70 yrs [1]. It may occur sporadically as isolated lesions (80-90%) or in association with an autosomal dominant disorder, tuberous sclerosis (TSC) (25-50%). TSC is characterized by seizures, mental retardation and hamartomatous tumours in multiple organs, which are associated with mutations in two genes – TSC1 and TSC2 [2]. Sporadic angiomyolipomas are usually unilateral and focal, whereas those which are associated with TSC are bilateral and multifocal and can occur at any age and in either sex. Usually it is asymptomatic and is often found incidentally in USG or CT. Sometimes it may present with retroperitoneal haemorrhage in adults [3].

## CASE SUMMARY

A 29 year old female presented with a mass in the left flank with a dragging pain of 1 year's duration. She had no history of tuberous sclerosis, haematuria and dysuria.

The haematological investigations and the renal function tests were within normal limits. CT scan showed a well defined hypodense mass lesion in the mid and lower pole of the left kidney, with a sharp renal parenchymal defect.

The nephrectomy specimen was received in 10% formalin and it was processed routinely.

## FNAC FINDINGS

A USG guided FNAC was done, which showed few benign spindle shaped cells and mature adipocytes, which was inconclusive and a histopathological examination was asked for.

## HISTOPATHOLOGICAL FINDINGS

The tumour weighed 400gms and measured 21 cms x 16 cms in diameter. The external surface of the tumour was yellowish to grey brown, with a lobulated surface [Table/Fig 1]. The capsule was intact and it could be easily stripped off. The cut section was greasy and yellowish to grey brown in colour, with areas of thickened blood vessels [Table/Fig 2]. The mass arose from the renal pelvis and it was soft to firm in consistency. Blood oozed out on cutting the fresh specimen.

The normal part of the kidney measured 9cms x 8cms x 6cms and the external surface and the cut section were unremarkable.

The microscopic examination revealed mature adipose tissue, thick walled blood vessels [Table/Fig 3] and bundles of smooth muscle cells, which are classical features of angiomyolipoma [Table/Fig 4]. A part of the normal kidney was seen adjacent to the tumour [Table/Fig 5]. No epithelioid component was seen.

There was no evidence of vascular invasion and it was not associated with epithelial cysts and renal cell carcinoma.

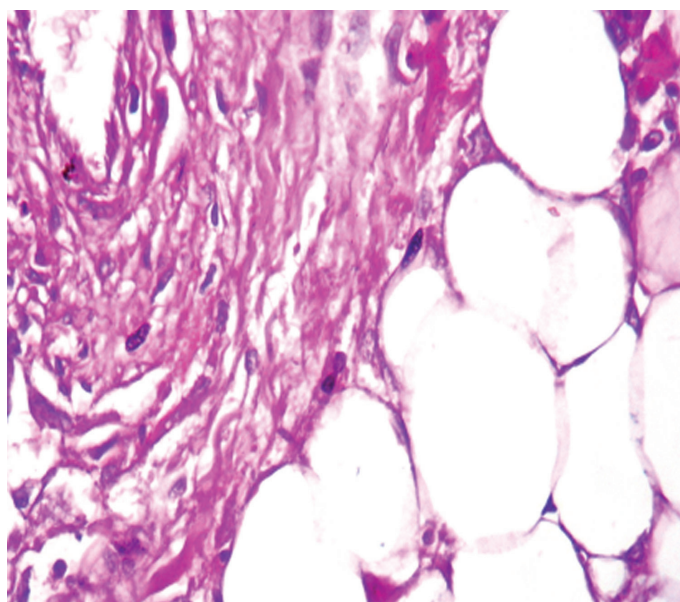
A final diagnosis of conventional renal angiomyolipoma was



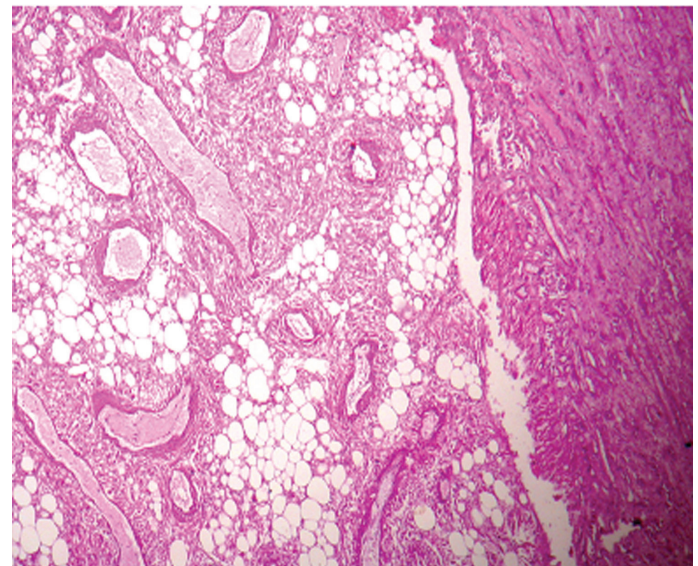
[Table/Fig-1]: External surface.



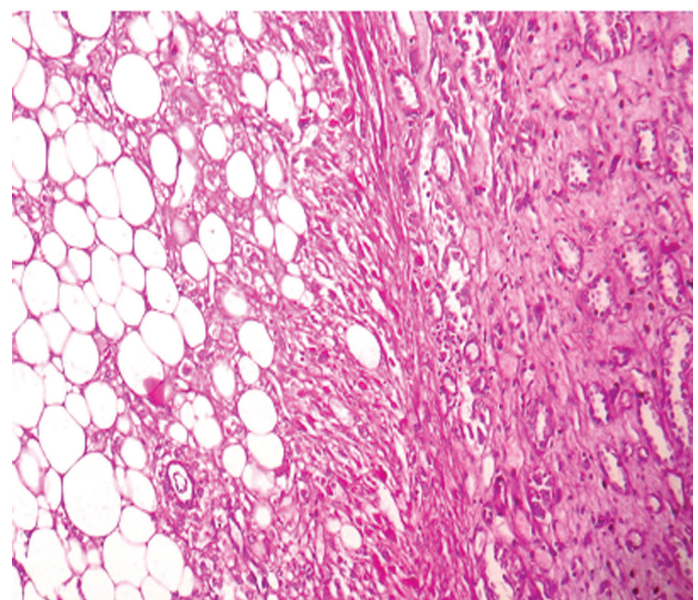
[Table/Fig-2]: Cut section



[Table/Fig-3]: Microphotograph - renal AML tumor showing fat & smooth muscle fibres in close proximity with blood vessel (40X).



[Table/Fig-4]: Microphotograph - renal AML showing all the 3 components i.e mature adipose tissue, thick walled blood vessels & bundles of smooth muscle cells (10x).



[Table/Fig-5]: Microphotograph - renal AML with adjacent normal kidney (10x).

made as it showed the classical text book picture.

### DISCUSSION

AML is a rare benign tumour which may occur as an isolated phenomenon or as a part of a syndrome which is associated with tuberous sclerosis. AML occurs not only as a rare tumour which is restricted to the kidney, but also as a biologically fascinating and morphologically heterogeneous entity. It is also seen at various other sites like the skin, appendix, colon, liver, lung and the smooth muscle fibres. Renal epithelioid angiomyolipoma is a recently recognized rare variant which originates from the perivascular epithelioid cell (PEC) and it has an aggressive clinical behaviour which includes local recurrence and metastasis [4]. Hence, one should extensively search for the epithelioid component.

Histologically, it can mimic renal cell carcinoma and it can be diagnosed accurately by immunostaining with HMB-45, Melan-A, CD68, CD117 and Ki-67 [5]. It mimics the same condition in imaging studies when it contains less fat. The main complication of AML is haemorrhage which is related

to the tumor size, increased vascularity and abnormal thickened blood vessels that are pre-disposed to the formation of microaneurysms and bleeding [3]. Very rarely, with vascular invasion into the inferior vena cava, it may present with a thrombus and secondary Budd – Chiari's syndrome. Pulmonary oedema can occur as a complication of the transcatheter embolization of renal AML in a patient with pulmonary lymphangioleiomyomatosis due to the tuberous sclerosis complex [6]. Conventional AML has got a very good prognosis as compared to the rare epithelioid variant of AML, which is potentially malignant [4]. Rarely, AML may present with epithelial cysts [7].

CT scan, USG, FNAC, histopathology and immunohistochemistry play a vital role in the diagnosis of renal angiomyolipoma [8].

As our case revealed a classical microscopic picture, a final diagnosis of AML was given after correlating it with the history and the radiographic findings.

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