

## Isolated Renal Angiomyolipoma – A Case Report

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**Abstract:** Angiomyolipoma is an uncommon benign tumour (hamartoma) composed of mature or immature fat, thick-walled blood vessels and smooth muscle elements in varying proportions. It is typically found in the young and inpatients with Tuberous Sclerosis Complex (TSC). We present a case of a 55-year old woman who has no TSC. A 55-year old post-menopausal house wife referred a peripheral hospital with subtle progressive right flank pain of 2years and right flank mass of 3 months duration. The pain remarkably worsened 3 months prior to presentation associated with poor appetite, weight loss and occasional vomiting. Other systematic symptoms and family history were unremarkable. She had right radical nephrectomy in view of a symptomatic huge right renal mass and clinical suspicion of malignancy. The pathology report revealed Angiomyolipoma. Clinicians should be wary of the presence of such an entity.

**Keywords:** angiomyolipoma, isolated, rare

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### INTRODUCTION

Angiomyolipoma (AML) is an uncommon benign tumour (hamartoma) composed of mature or immature fat, thick-walled blood vessels and smooth muscle elements in varying proportions [1]. There are two varieties of renal AML, one associated with tuberous sclerosis (TS or Bourneville disease) or other facomatosis (von Recklinghausen's neurofibromatosis and von Hippel Landau's disease) and another that appears as an isolated lesion [1–4].

In the first case, renal AMLs are most often multiple, bilateral, symptomatic and without female or male predominance [2]. Renal AMLs were found in 40–80% of patients with TS [1]. In the second case, renal AMLs are single, often asymptomatic, have a female predominance and are typical of the fifth and sixth decades of life [5,6].

Typically, angiomyolipomas are present in the young [7], in patients with Tuberous Sclerosis Complex [8] and continue to grow during adulthood [1,4,9]. These lesions distort renal architecture and can compromise function. Renal failure is a long-term complication of Tuberous Sclerosis Complex [7] and the leading cause of death in adults [10]. Although somewhat rare, angiomyolipomas also can undergo malignant degeneration [11]. Dysmorphic blood vessels in the angiomyolipoma can have micro aneurysms [12] or macro aneurysms [13]. These aneurysms may rupture and haemorrhage, resulting in significant morbidity and, possibly, death [8].

We report a case of isolated renal angiomyolipoma in a 55-year old female who does not have Tuberous Sclerosis complex or any other underlying co-morbidity

### PRESENTATION OF CASE

A 55-year old post-menopausal house wife referred a peripheral hospital with subtle progressive right flank pain of 2years and right flank mass of 3 months duration. The pain remarkably worsened 3 months prior to presentation associated with poor appetite, weight loss and occasional vomiting. Other systematic symptoms and family history were unremarkable. Examination revealed a stable but chronically ill-looking woman with a tender ballotable left renal mass.

Her haematocrit was 46%, urinalysis and renal function were normal but a huge heterogeneous intra-abdominal mass with sonoluculent periphery and measuring 11.9 x 11.4cm arising from upper and mid-pole of right renal parenchyma was noted. Abdominal CT scan revealed oval lobulated mixed density non-enhancing but predominantly hypodense right upper pole kidney mass (HU = -75 to -10), measuring 12.9 x 10.0 x 10.1cm in dimensions. No calcification was seen. There was an inferior pole simple cortical cyst measuring 10mm in diameter involving the left kidney. There were no enlarged lymph nodes and other intra-abdominal viscera were normal.

She had right radical nephrectomy in view of a symptomatic huge right renal mass and clinical suspicion of malignancy. The pathology report revealed

Renal Angiomyolipoma. Microscopically, the tumour showed mixture of mature adipose tissue, thick-walled poorly organised blood vessels and smooth muscles i.e.

classic triphasic pattern. Patient did well and was discharged home for follow-up.

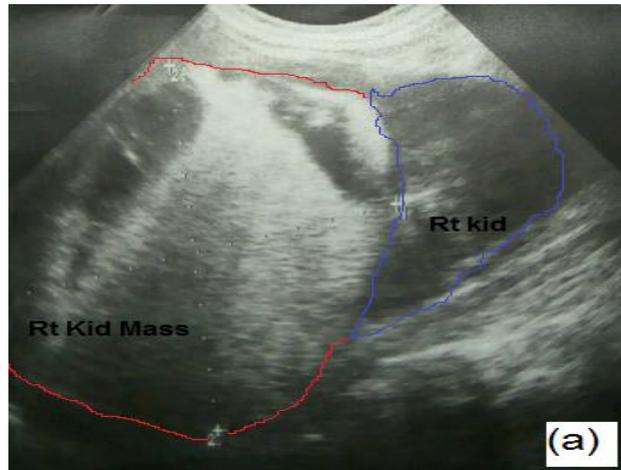


Fig-1: Renal ultrasound: heterogeneous mass in the right kidney.



Fig-2: Gross image of the tumour (left) showing cut surface (right)

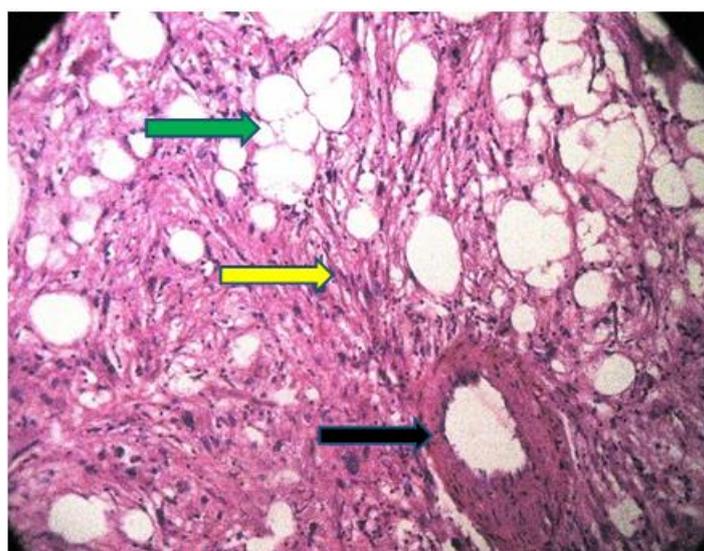


Fig-3: Photomicrograph showing admixture of mature adiposities (green arrow), thick-walled blood vessel (black arrow) and smooth muscle (yellow arrow) (H&E Mag. X100)

## DISCUSSION

Renal angiomyolipoma is an unusual, generally benign tumour that has been associated with tuberous sclerosis and is characterized by a histologic triad of tortuous, thick-walled blood vessels, phenotypic smooth muscle cells, and adipose cells in varying proportions [14].

Renal angiomyolipoma (AML) is a benign hamartomatous tumour that contains fat, smooth muscle, and abnormal blood vessels in varying proportions and accounts for up to 3% of kidney mass. The main complications of renal AML are retroperitoneal bleeding or bleeding into the urinary collection system, which can be life threatening [15, 16]. AML was recently described as the most common cause of perinephric haemorrhage [17].

The bleeding tendency is related to the angiogenic component of the tumour that includes irregular and aneurismal blood vessels [15, 17, 18]. The major risk factors for bleeding are the size of the tumour, the grade of angiogenic component of the tumour, and the presence of tuberous sclerosis [1, 16, 19, 20]. The main indications for treatment of patients with renal AML are a tumour with a threshold of greater than 4 cm and presence of symptoms for any tumour size [15, 18, 19].

The use of the grade of angiogenic component of the tumour as a predictor for bleeding is based on limited retrospective data and needs further confirmation before it is used as a criterion for treatment [16, 19, 20]. Treatment options for renal AML include surgery and selective arterial embolization (SAE), aiming toward avoidance of bleeding complications and preservation of the renal parenchyma [1, 9, 16, 18].

Several researchers have hypothesized that angiomyolipoma (AML) is a member of the perivascular epithelioid cell family of tumours that show a unique pattern of immune reactivity, with co-expression of muscle-specific markers (muscle-specific actin and smooth muscle actin) as well as melanocytic markers such as HMB-45 and melan-A [21, 22].

Expression of microphthalmia transcription factor (mitf), a nuclear regulatory protein critical for melanocytic differentiation, and expression of the tyrosinase gene, an enzyme involved in the synthesis of melanin [23], has been reported in melanocytic tumours. Recently, the use of these antibodies in the diagnosis of AML of the kidney has been evaluated [24].

Renal angiomyolipoma is an unusual tumour of uncertain histogenesis. Many authors have considered it to be a hamartomatous lesion. Because of a diversified phenotypic population, a cell of origin of

this tumour has been the subject of many studies. Recent detection of clonal genomic alterations in renal angiomyolipoma and rare case reports of malignant renal angiomyolipomas are features that favour a neoplasm [25-27].

Demonstration of both calponin and muscle-specific actin in (HHF-35) immune reactivity in renal angiomyolipoma further characterizes the smooth muscle cell nature of this unusual tumour; this is supported by ultrastructural study. The spindle-type cells in renal angiomyolipoma preferentially stained positive for the muscle markers calponin and muscle-specific actin in (HHF-35). This preferential staining pattern paralleled the ultrastructural characteristics of the 3 cell types in that, qualitatively, the spindle type cells contained more numerous microfilaments than either the epithelioid cells or the adipocytic cells. The lack of S100 staining makes it unlikely that these cells are of epithelial origin. The lack of either Z-bands or Weibel-Palade bodies, ultrastructurally, makes it unlikely that these cells are of either skeletal muscle or endothelial cell origin [14].

## CONCLUSION

Renal Angiomyolipoma is an unusual and rare tumour of children typically found in patients with Tuberous Sclerosis Complex. Even more rare, is its presentation in an elderly patient without Tuberous Sclerosis Complex. Clinicians should be wary of the presence of such an entity in patients who are otherwise not prone to this tumour.

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