



## Angiomyolipoma of Kidney- A Case Report

Authors

**Dr Gopinath Barui<sup>1</sup>, Dr Tripti Das<sup>2</sup>**

<sup>1</sup>Associate Professor, Department of Pathology, R.G. Kar Medical College & Hospital  
Email: [gnbarui@gmail.com](mailto:gnbarui@gmail.com), Mobile no.9831250951

<sup>2</sup>Post Graduate Trainee, Department of Pathology, R.G. Kar Medical College & Hospital  
Email: [triptidasrgk@gmail.com](mailto:triptidasrgk@gmail.com), Mobile no.9635781818

### Abstract

*Renal angiomyolipoma is an uncommon benign tumour characterized by a variable mixture of adipose tissue, smooth muscle and thickened blood vessels and hence the name. We here in, present a case of renal angiomyolipoma in a 60 years old female patient presenting with flank pain and swelling. Clinico-radiological evaluation suggested a mass lesion arising from kidney. Nephrectomy was carried out. The histological examination revealed admixture of spindle cells & adipose tissue along with thick walled blood vessels. The case was diagnosed as Angiomyolipoma of right kidney and the patient was followed up closely.*

**Keywords:** renal angiomyolipomas, nephrectomy, PEComas, mesenchymal, benign tumour.

### Introduction

Angiomyolipoma is a benign mesenchymal tumour composed of variable portions of adipose tissue, spindle cells, epithelioid smooth muscle cells and abnormal thick walled blood vessels. It belongs to a family of lesions called perivascular epithelioid cell tumours (PEComas), which are characterised by the proliferation of perivascular epithelioid cells<sup>1</sup> (WHO,2016).

Angiomyolipomas constitute approximately 1% of surgically removed renal tumours.

Angiomyolipoma has been considered an uncommon neoplasm, but its observed incidence is increasing as it is increasingly detected during imaging performed to evaluate other conditions<sup>2</sup>.

It can occur sporadically or in patients with tuberous sclerosis, an autosomal genetic disorder, characterized by mental retardation, seizures and the development of cellular proliferations such as angiomyolipoma, subependymal giant cell tumours, cutaneous angiofibromas, cardiac rhabdomyomas, lymphangiomyomatosis and pulmonary multifocal micronodular hyperplasia<sup>3</sup>. In patients with tuberous sclerosis, renal angiomyolipoma is found in the third or fourth decades of life; whereas sporadic angiomyolipomas tend to occur in older patients. Puberty may influence the development of angiomyolipoma<sup>4</sup>.

### Case Summary

This 60 yrs old female patient attended urology outdoor with complaints of pain and swelling in right flank region for last 6months. She had no history of tuberous sclerosis or haematuria or dysuria. Routine haematological and biochemical parameters were within normal limit.

USG of whole abdomen revealed two hyperechoic SOL at the upper part of right kidney. The nature of the lesion could not be ascertained.

CECT abdomen shows three hypodense SOLs (having fatty density) noted at upper pole of right kidney at lateral cortex, largest measuring 20x23 mm and suggested as possibility of angiomyolipoma or any mass lesion of right kidney (figure no.1).

FNAC of the renal SOL was done outside and reported as- suggestive of poorly differentiated carcinoma.

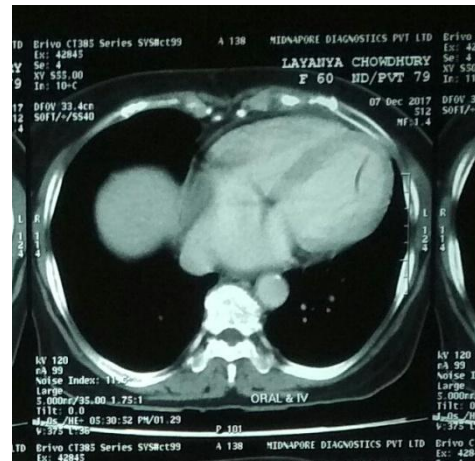
She was admitted to urosurgery ward and planned for right nephrectomy. The right nephrectomy was done and the specimen was sent to our Pathology department. The patient was on close follow up after the surgery.

The total specimen measured (10X6X4) cm. On cut section of right kidney a solid yellowish growth measured (2X2) cm in the cortical region (figure no.3). Another nodular growth noted on the surface of the kidney measuring 1.2 cm in maximum diameter.

The microscopic examination revealed variable mixture of mature adipose tissue, thick walled blood vessels and bundles of smooth muscles, which are classical features of angiomyolipomas (figure no.4). The normal kidney tissue was noted adjacent to angiomyolipoma which was clearly demarcated in histopathological examination (figure no.5).

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

A final diagnosis of renal angiomyolipoma was done.



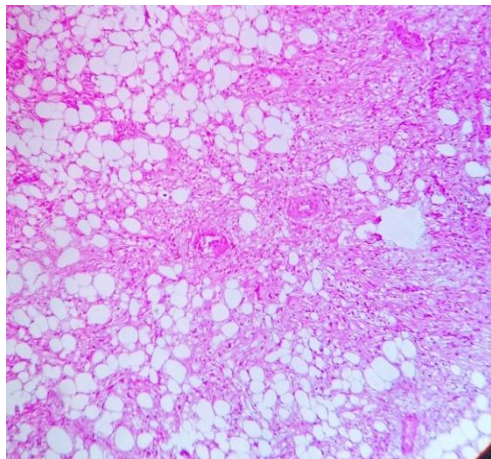
**Figure 1** CECT Abdomen show one large hypodense lesion at upper pole of right kidney at lateral cortex



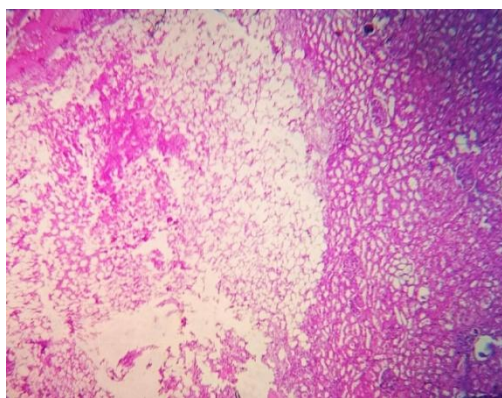
**Figure 2:** Photograph showing gross appearance of kidney- external surface



**Figure 3:** Photograph showing a yellowish solid growth in the cortical surface of kidney.



**Figure 4:** Photomicrograph showing renal Angiomyolipoma consists of all the three components i.e. mature adipose tissue, thick walled blood vessels and smooth muscle cells (H&E; 400X)



**Figure 5:** Photomicrograph showing renal angiomyolipoma with adjacent normal kidney (H&E; 100X)

### Discussion

Angiomyolipoma (AMLs) are distinctive neoplasms composed of variable combinations of smooth muscle, adipose tissue, and vasculature. Although most commonly found within the kidneys, they may occur at extrarenal sites including liver, lungs, lymph nodes and retroperitoneal soft tissues<sup>5</sup>.

Renal angiomyolipoma is a benign neoplasm arising from mesenchymal elements and was first reported in 1951<sup>6</sup>.

It was initially considered to be a hamartoma rather than a true neoplasm, but the clonal nature of the lesion is now well established<sup>7</sup>.

It may arise in the cortex or medulla of the kidney, as well as in retroperitoneal soft tissue, with or without renal attachment<sup>8</sup>.

Patients without tuberous sclerosis are more likely to have larger lesions, so are more likely to present with flank pain, haematuria, palpable mass or a combination of these signs and symptoms. Retroperitoneal haemorrhage may occur. Simultaneous occurrence of angiomyolipoma with renal cell carcinoma and oncocytoma in the same kidney has also been reported. CT and USG permit the preoperative diagnosis of angiomyolipoma in almost all cases<sup>9</sup>.

Uncommonly, it extends into the intra renal venous system, the renal vein or the vena cava. They are usually well demarcated from the adjacent kidney, but not encapsulated. Most of them are solitary, but multiple tumours may be present.

Most angiomyolipomas are composed of a variable mixture of mature fat, thick walled poorly organized blood vessels and smooth muscles (the classic triphasic etiology). The border between angiomyolipoma and the kidney is typically sharp. The smooth muscle cells appear to emanate from blood vessel walls in a radial pattern. They are most commonly spindle cells but may be rounded epithelioid cells. Some angiomyolipomas (so called capsulomas), which are often located subcapsularly and composed almost entirely of smooth muscle cells, resemble leiomyomas. A component composed of cells associated with thin walled, branching vessels with a pattern similar to lymphangioliomyoma is another variation of smooth muscle component.

The lipomatous component is typically consists of mature adipose tissue may contain vacuolated adipocytes suggesting lipoblast, resembling a liposarcoma when there is extensive adipocytic differentiation.

The blood vessels have thick walls and lack the normal elastic content of arteries.

Highly pleomorphic angiomyolipomas rich in epithelioid cells closely resemble sarcomatoid

renal cell carcinomas and so called malignant fibrous histiocyomas<sup>10</sup>.

Angiomyolipomas are characterized by coexpression of melanocytic markers (HMB45, melan A and microphthalmia transcription factor) and smooth muscle markers (smooth muscle actin and calponin). CD68, S100 protein, estrogen and progesterone receptors and desmin may also be positive, whereas epithelial markers are always negative.

Classic angiomyolipomas are benign. A very small minority are associated with complications and morbidity and mortality. Renal cyst and multiple angioliipomas in tuberous sclerosis patients can lead to renal failure<sup>11</sup>.

It is important to recognize the benign nature of these tumours and their characteristics radiological features. Most of these tumours are amenable to conservative management with frequent follow up. Even when surgical excision is indicated, the kidney itself can often be spared<sup>12</sup>.

Pulmonary oedema can occur as a complication of the transcatheter embolization of renal angiomyolipoma in a patient with pulmonary lymphangiomyomatosis due to the tuberous sclerosis complex. Conventional angiomyolipoma has got a very good prognosis as compared to the rare epithelioid variant of angiomyolipoma, which is potentially malignant. Rarely, angiomyolipoma can present with epithelial cyst<sup>13</sup>.

## References

1. Folpe AL, Kwiatkowski DJ(2010). Perivascular epithelioid cell neoplasms: pathology and pathogenesis. *Hum Pathol.* 9:198-203.PMID:11981115; Martignoni G, Pea M, Reghellin D, Zamboni G, Bonetti F(2008). PEComas: the past, the present and the future. *Virchows Arch.* 452:119-32.PMID:18080139 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
2. Fujji Y, Ajima J, Oka K, Tosaka A, Takehara Y (1995) Benign renal tumours detected among healthy adults by abdominal ultrasonography. *Eur Urol.* 27: 124-7.PMID:7744154 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
3. Steiner MS, Goldman SM, Fishman EK, Marshall FF(1993).The natural history of renal angiomyolipoma. *Urol.* 150:1782-6.PMID: 8230504 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
4. Cook JA, Oliver K, Mueller RF, Sampson J (1996). Across sectional study of renal involvement in tuberous sclerosis. *J Med Genet.* 33:480-4.PMID:8782048 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
5. Sternberg's Diagnostic Surgical Pathology, 6th Edition, Page no. 2003
6. Morgan GS, Straumford JV and Hall E: Angiomyolipoma of the kidney. *J Urol* 65:525-527, 1951.
7. Green AJ, Sepp T, Yates JR (1996). Clonality of tuberous sclerosis hamartomas shown by non-random X chromosome inactivation. *Hum Genet.* 97:240-3.PMID:8566961 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
8. Tallarigo C, Baldassarre R, Bianchi G, Comunale L, Olivo G, Pea M et al.(1992). Diagnostic and therapeutic problems in multicentric renal angiomyolipoma. *J Urol.* 148:1880-4.PMID:1433628 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
9. Steiner MS, Goldman SM, Fishman EK, Marshall FF (1993).The natural history of renal angiomyolipoma. *Urol.* 150:1782-6.PMID: 8230504 (WHO Classification of Tumours of the Urinary System and Male Genital Organs,2016)
10. Nese N, Martignoni G, Fletcher CD, Gupta R, Pan CC, Kim H, et al. Renal perivascular epithelioid cell tumors [(PEComa), so called epithelioidangio-

myolipoma (EAML): analysis of 61 cases including 44 with pure/predominant epithelioid (PPEComa) morphology and parameters associated with malignant outcome. *Lab Invest* 2009; 89 (Suppl 1):186A.

11. Schillinger F, Montagnac R (1996). Chronic renal failure and its treatment in tuberous sclerosis. *Nephrol Dial Transplant*. 11:481-5. PMID:8671819 (WHO Classification of Tumours of the Urinary System and Male Genital Organs, 2016).
12. Taylor R, Joseph DB, Koham EC, Wilson ER, Bueschen AI. Renal angiomyolipoma associated with lymph node involvement and renal cell carcinoma in patients with tuberous sclerosis. *J Urol* 1989; 141: 930-2.
13. Mikami S, Oya M, Mukai M. Angiomyolipoma with epithelial cysts of the kidney in a man. *Pathol Int*. 2008 Oct; 58(10): 664-7.