

# Renal Epithelioid Variant of Angiomyolipoma - A Rare Case Report

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## ABSTRACT

Epithelioid Angiomyolipoma (EAML) is a rare mesenchymal tumour with malignant potential. The epithelioid variant of angiomyolipoma was first described in 1995 by Martignoni et al as a distinct clinicopathologic variant of AML which is mainly characterized by a predominance of epithelioid cells. Histologically, EAML is characterized by sheets or nests of large polygonal epithelioid cells with abundant eosinophilic or occasionally clear cytoplasm, often with prominent nucleoli, including multinucleated and markedly pleomorphic forms. These tumours share a distinctive perivascular epithelioid cell phenotype; they belong to the PEComa tumour family. Nearly all EAMLs show immunoreactivity for both melanocytic and myoid markers. EAML can pose significant diagnostic challenge on H and E microscopy as it morphologically mimics a variety of neoplasms. An EAML diagnosed by the authors in an adult male patient is presented here.

**Keywords:** Angiomyolipoma, Epithelioid.

## INTRODUCTION

Angiomyolipoma (AML) is a common mesenchymal renal neoplasm and has been classified under the group of perivascular epithelioid cell tumours [PEComas]. Approximately 80% of AML are seen sporadically, the others being seen as part of Tuberous Sclerosis Complex. [1]

AML has various forms with its classic form having triphasic histopathologic characteristics, comprising of variable proportions of blood vessels, smooth muscle and adipose tissue. Other morphologic variants of AML that have been described in literature include epithelioid, oncocytic, fat-predominant, smooth muscle-predominant, angiomyolipoma with epithelial cysts and sclerosing types. [2-4]

The epithelioid variant of angiomyolipoma has also been referred to as

atypical angiomyolipoma and malignant angiomyolipoma.

The 2016 World Health Organization (WHO) Classification of Renal Neoplasms categorizes epithelioid-AML as a separate entity which has malignant potential. [5]

In 2011 Nese et al estimated that 120 cases of EAML had been reported in the literature. [6] The differentiating feature of EAML and classic form is that in EAML the classic triphasic histologic features are usually obscured and instead we see sheets of epithelioid cells with granular eosinophilic cytoplasm. However, the amount of epithelioid composition sufficient to warrant classification as an EAML has not been uniform, ranging from 5% to 100%. EAMLs can have a high degree of cytologic pleomorphism and atypia and can exhibit necrosis. [7] EAML can pose

significant diagnostic challenges as its morphology can mimic a variety of neoplasms, including renal cell carcinoma, renal oncocytoma, adrenal cortical neoplasm, epithelioid smooth muscle tumour, epithelioid gastrointestinal stromal tumor and epithelioid melanoma. [8]

### CASE REPORT

A 32 years old male patient was admitted in our hospital with complaints of pain in right side of abdomen for previous 10 days. There was no history of fever,

nausea, vomiting and haematuria. On examination: Mild tenderness was noticed in the right lumbar region and in right renal angle. Ultrasonography and CT Intra Venous Urography scan was done, they were suggestive of Angiomyolipoma of right kidney. Open right partial nephrectomy with removal of angiomyolipoma at upper pole was performed.

Operative findings: Approximately 8 x 4.2 cm mass was present at superior pole of the right kidney with hematoma in its centre, as depicted in figure 1.



Fig 1 – Intra-operative findings of resected mass over upper pole of kidney.

### HISTOPATHOLOGICAL EXAMINATION

Gross Examination – Received a single, grey – brown, soft to firm tissue mass measuring 6.5 x 7 x 2.5 cm in the histopathology section.

External surface was well circumscribed, partially encapsulated, homogenous with few cystic areas showing proliferative outgrowth with pushing borders as in figure 2.

Cut section showed variegated appearance with golden-yellow to tan-brown areas as shown in figure 3.



Fig 2 - External Surface

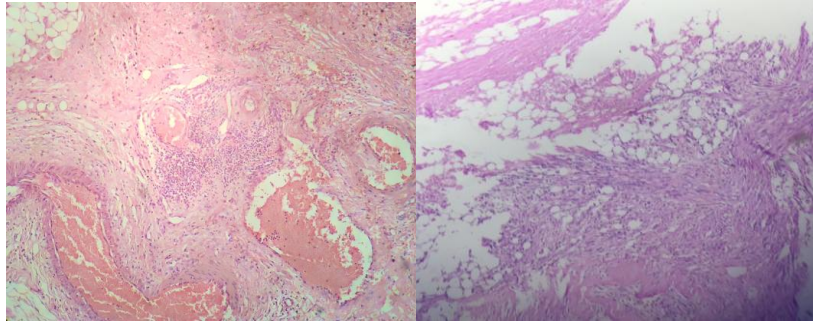


Fig 3 - Cut Section

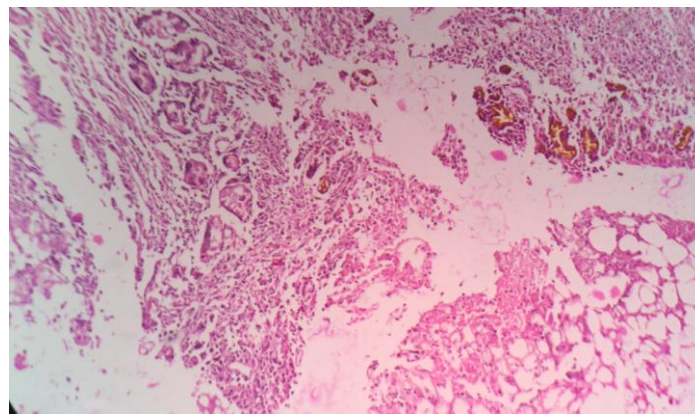
Microscopic Examination– Study of hematoxylin and eosin stained sections revealed an encapsulated lesion showing a polymorphous population of cells comprising epithelioid cells, spindle cells, mature adipocytes and few multinucleated giant cells. The spindle cells were arranged in short fascicles and whorls. Also seen were mononuclear polygonal cells having mild pleomorphic vesicular nuclei with eosinophilic granular cytoplasm along with thick walled blood vessels and few thin walled dilated blood vessels. Focal areas

showed perivascular cuffing by mononuclear cells as shown in figures 4 to 8. Adjacent renal tissue showed normal glomeruli and tubules.

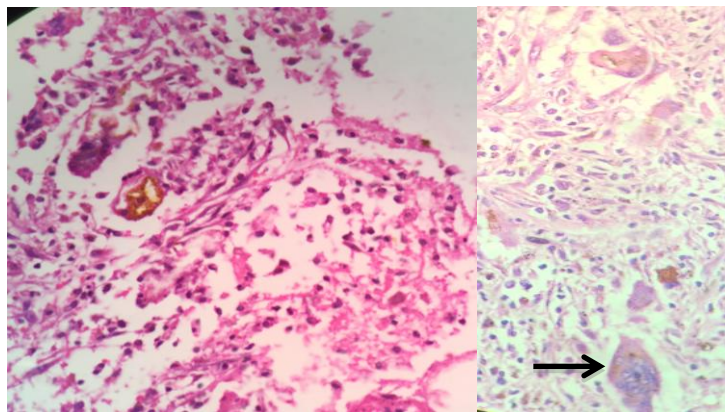
No evidence of mitoses / necrosis was noted in the sections studied.



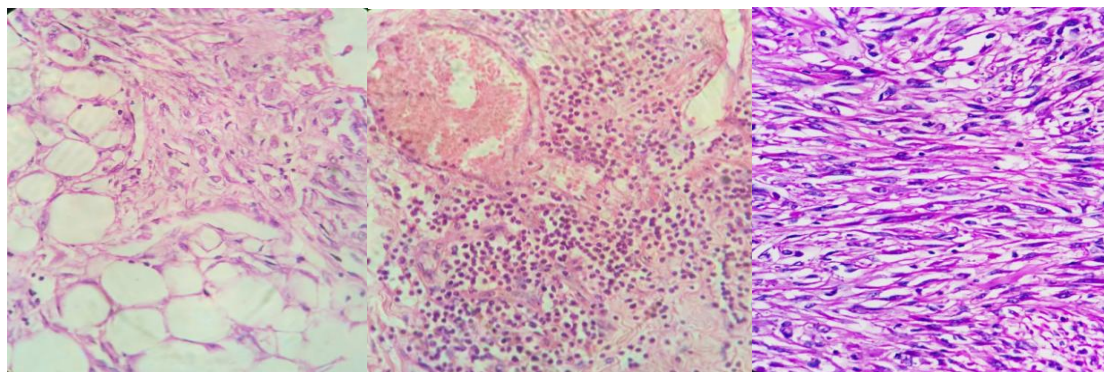
**Fig 4 – H and E 4X View (Scanner view) showing adipocytes, congested thickened blood vessels and spindle cells.**



**Fig 5 – H and E 10 X view - Pigments and Giant cells.**



**Fig 6 – H and E 40 X view – Pigments and Giant cells (Arrow)**



**Fig 7 – H and E 40 X view – Adipocytes and Spindle cells along with perivascular cuffing by mononuclear cells.**

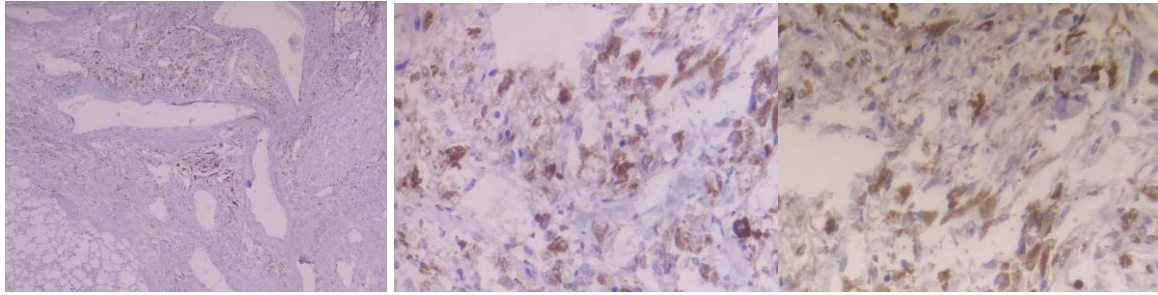


Fig 8 – ImmunoHistoChemical marker HMB 45 showing strong cytoplasmic positivity for epithelioid cells.

On H and E sections, the diagnosis given was of epithelioid variant of Angiomyolipoma.

## DISCUSSION

AML was originally believed to be a hamartomatous lesion. Most AMLs arise in kidney, though extrarenal AMLs are also described in various sites such as retroperitoneum, liver, lung, uterus, vagina, ovary, colon, lymph nodes, skin, bone, nasal cavity, oral cavity, and adrenal gland. [8] The kidney is the most common site of origin for classical and epithelioid AMLs. In the present case also, EAML was found in the kidney of the patient.

The epithelioid variant of AML (epithelioid angiomyolipoma [EAML]) was first described in 1995 by Martignoni et al as a distinct clinicopathologic variant of AML mainly characterized by a predominance of epithelioid cells. [9] Angiomyolipomas are slow-growing and usually exhibit expansile rather than infiltrative growth, with the possible exception of some epithelioid variants that may extend and bulge into the perirenal soft tissue. The present case showed a proliferative outgrowth with pushing borders as can be seen in figure 2 above.

As reported in literature the usual size of renal EAMLs is larger as compared to the classical AML. In the present case also it measured 6.5 cm. Intra-abdominal AMLs are usually asymptomatic, often incidentally found during radio-imaging, but some patients may present with abdominal discomfort (as seen in this case), flank pain, or hematuria.

The important prognostic factors for EAML include, (a) presence of tuberous sclerosis syndrome, (b) tumor necrosis, (c) extra-renal extension or renal vein invasion,

(d) carcinoma-like histology, and (e) tumor size greater than 7.7 cm. Brimoet al suggest another predictive model based on (a)  $\geq 70\%$  atypical epithelioid cells, (b)  $>2$  mitotic figures/10 HPF, (c) atypical mitoses, and (d) necrosis; presence of three or more of the features was highly associated with malignant behavior. [6] However, in the present case none of these factors were clearly noted which indicates benign nature of the tumour.

In the present case, IHC marker HMB 45 was used for confirmation of the diagnosis and it showed strong cytoplasmic reactivity and positivity, as depicted in figure 8.

However, other IHC markers can also be used for confirmation of diagnosis, as mentioned by Mete O and Kwast T.H. [8] AMLs are typically positive for HMB-45 antibody raised against melanosome-related antigen. They are also known to be positive for other melanocytic markers such as HMB-50, Mart-1/Melan-A, tyrosinase, and microphthalmia-associated transcription factor. Other markers for AML are CD63 and CD117. AMLs exhibit variable immunopositivity for myoid markers such as smooth muscle actin, muscle-specific actin, desmin, and calponin. Desmin is less often positive, and myoid markers are less frequently expressed in epithelioid and plump spindle cells. About 25% of AMLs express estrogen and progesterone receptors. Angiomyolipomas are typically negative for S100 protein and epithelial markers such as cytokeratin and epithelial membrane antigen.

## CONCLUSION

Epithelioid Angiomyolipoma is a rare tumour, as in the present case, having clinically aggressive behavior and malignant potential. On imaging studies EAML can mimic other tumours of renal origin and hence it should be considered in the evaluation of a fat poor and non-calcified renal mass, especially in a younger age group patient. In this case, initially the authors also suspected that the tumour may be malignant in nature. However, neither atypical epithelioid cells nor mitotic figures nor necrosis were found, which confirmed the benign nature of the tumour. Since there is paucity of available literature and the tumour aggressiveness in nature, it is advisable to diagnose such tumours and their variants which will aid in the post-operative treatment and prognosis of patients.

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## REFERENCES

1. Froemming A T, et al Renal Epithelioid Angiomyolipoma: Imaging Characteristics in Nine Cases With Radiologic-Pathologic Correlation and Review of the Literature. *AJR* 2013; 200:W178–W186
2. Fine SW, Reuter VE, Epstein JI et al. Angiomyolipoma with epithelial cysts (AMLEC): a distinct cystic variant of angiomyolipoma. *Am J SurgPathol* 2006; 30: 593 – 599.
3. Martignoni, G., Pea M, Bonetti F et al. Oncocytoma-like Angiomyolipoma. A clinicopathologic and immunohistochemical study of 2 cases. *Arch Pathol Lab Med* 2002; 126: 610-612.
4. Matsuyama A, hisaoka M, Ichikawa K et al. Sclerosing variant of epithelioid angiomyolipoma. *PatholInt* 2008; 58: 306 – 310.
5. <https://www.iarc.fr/wp-content/uploads/2018/07/BB7.pdf>
6. Nese N, Martigoni G, Fletcher CD, et al. Pure epithelioid PEComas (so-called epithelioid angiomyolipoma) of the kidney : a clinicopathologic study of 41 cases; detailed assessment of morphology and risk stratification. *Am J SurgPathol* 2011; 35: 161 – 176.
7. Faraji H, Nguyen BN, Mai KT. Renal epithelioid angiomyolipoma: A study of six cases and a meta-analytic study: development of criteria for screening the entity with prognostic significance. *Histopathology* 2009; 55: 525 – 534.
8. Mete O and Kwast T.H. (2011) Epithelioid Angiomyolipoma: A Morphologically Distinct Variant That Mimics a Variety of Intra-abdominal Neoplasms. *Archives of Pathology & Laboratory Medicine*: May 2011, Vol. 135, No. 5, pp. 665-670.
9. Aydin, H. C. Magi-Galluzzi , B. R. Lane , et al. Renal angiomyolipoma: clinicopathologic study of 194 cases with emphasis on the epithelioid histology and tuberous sclerosis association. *Am J SurgPathol* 2009. 33 (2):289–297

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