

**ANGIOMYOLIPOMA OF KIDNEY – A CASE REPORT**

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**Abstract**

Angiomyolipoma belongs to a family of neoplasms called perivascular epithelioid cell tumors (PEComas). The World Health Organization (WHO) defines PEComas as “mesenchymal tumors composed of histologically, ultra structurally, and immunohistochemically distinctive perivascular epithelioid cells. Renal angiomyolipoma is an uncommon benign tumor characterized by a variable mixture of adipose tissue, smooth muscle and thickened blood vessel and perivascular epithelioid cells also known as PEComa. Microscopically, the tumor shows mature adipose tissue, tortuous thick-walled blood vessels lacking elastic tissue lamina, and bundles of smooth muscle that seem to emanate from the vessel wall. A 55 year old male patient presented with a huge abdominal painful lump in left hypochondrium. Clinico-radiological evaluation suggests a mass lesion arising from kidney. Nephrectomy was carried out. Cut section of kidney showed multiple nodular yellow white and brown areas. Random sections from growth showed admixture of Spindle cells & adipose tissue along with thick walled blood vessel. Immunohistochemistry was carried out the result of which showed HMB 45 and Smooth Muscle Actin positive, but cytokeratin was negative. Considering the unilaterality, unifocality, larger size of the tumor and microscopic finding, and IHC result, the case was confirmed as sporadic angiomyolipoma.

**Key Words: Nephrectomy, Renal angiomyolipoma, perivascular epithelioid cell, PEComa, HMB45, SMA**

**Introduction**

Renal angiomyolipoma is an uncommon benign tumor characterized by a variable mixture of adipose tissue, smooth muscle and thickened blood vessel and perivascular epithelioid cells.<sup>[1]</sup> Microscopically, the

typical angiomyolipoma case shows mature adipose tissue, tortuous thick-walled blood vessels lacking elastic tissue lamina, and bundles of smooth muscle that seem to emanate from the vessel wall. A fourth component, which in reality is a variation of

the third, is represented by a cell type with epithelioid features originally described by Apitz and currently known as the *perivascular epithelioid cell (PEC)*.<sup>[2, 3]</sup> Predominance of this cell is responsible for the term *PEComa*. Often has an intimate relationship with blood vessel walls. Renal angiomyolipoma have an incidence of 0.3–3% and arise from the mesenchymal elements of the kidney.<sup>[4]</sup> Two distinct clinical pattern of this tumor exist. Those associated with tuberous sclerosis are usually bilateral; multifocal and vary in size.<sup>[1]</sup> The sporadic one is large in size, unilateral & unifocal.<sup>[1]</sup> Most small angiomyolipomata are asymptomatic and found incidentally on imaging studies, usually incidental from US or CT scans performed for unrelated clinical indications. In minority cases, they classically present with flank pain (53%), a palpable tender mass (47%) and gross haematuria (23%); this is known as 'Lenk's triad'.<sup>[4]</sup>

A case of sporadic angiomyolipoma was diagnosed in our institute on routine histology with special emphasis to its specific IHC marker.

#### **Case report:**

A 55 year old male patient attended urology OPD with pain and swelling in left hypochondrium. On palpation, a huge abdominal lump (size = 16x 10 cm) was felt in the loin along with the kidney. Routine hematological & Biochemical parameters are within normal limit. CT scan of abdomen showed a mass 15.51 x 10.26 cm size in left kidney showing hemorrhage and mass effect. He was admitted in urology wards and planned for left nephrectomy. The left nephrectomy was done and the specimen was sent to Pathology Department. The nephrectomy specimen measured 14cm x 11cm x 8 cm. Cut section of kidney shows a large growth (size=13cm x 10 cm x 8 cm) having multiple nodular yellow white and brown

areas replacing almost the entire kidney except in lower pole where a rim of grossly uninvolved renal parenchyma is noted. Random sections from different areas of growth showed admixture of Spindle cells & adipose tissue along with thick walled blood vessel. Mitosis was minimal. IHC marker for HMB 45 and SMA were positive, but cytokeratin was negative. Cytogenetic study could not be carried out. However, considering the unilaterality, unifocality and larger size of the tumor, the case was confirmed as sporadic angiomyolipoma on the basis of histomorphology and immunopositivity towards HMB45 & smooth muscle protein.

#### **Discussion**

Angiomyolipoma belongs to a family of neoplasms called perivascular epithelioid cell tumors (PEComas).<sup>[5]</sup> The World Health Organization (WHO) defines PEComas as "mesenchymal tumors composed of histologically, ultra structurally, and immunohistochemically distinctive perivascular epithelioid cells."<sup>[3]</sup> The perivascular epithelioid cell (PEC) is a "novel" cell type showing morphologic, immunohistochemical, ultrastructural, and genetical distinctive features, such as an epithelioid appearance with a clear to granular cytoplasm, a round to oval, centrally located nucleus and an inconspicuous nucleolus, and a typical perivascular location. Immunohistochemically, PEC expresses myogenic and melanocytic markers, such as HMB-45, Melan-A/Mart1, and smooth muscle actin and, less commonly, desmin.<sup>[6, 7]</sup> Epithelial markers were negative.<sup>[7]</sup>

Presence of admixture of all these components (mature adipose tissue, spindle cells and thick walled vasculature with presence of perivascular epithelioid cells) makes the diagnosis of angiomyolipoma easy. However predominance of any one of these component may lead to difficulty in

differentiating from other tumors of similar morphology. AMLs in which spindle cells of smooth muscle type predominate may look like leiomyomas, leiomyosarcomas, or gastrointestinal stromal tumors (GISTs).<sup>[8, 9]</sup> AMLs in which the adipose tissue is predominant and accompanied by atypical cells and lipoblasts can be easily confused with an atypical lipomatous tumor (well-differentiated liposarcoma). Highly pleomorphic AMLs rich in epithelioid cells closely resemble sarcomatoid renal cell carcinomas and so-called malignant fibrous histiocytomas.<sup>[10]</sup>

### Conclusion

Hence it is important for the pathologist to recognize that fatty tissue is part of lesion and not interpret as invasion. In all of these instances, clues to the recognition of this entity include: (1) presence of islands of mature fat among the other cell components; (2) collections of epithelioid clear cells in intimate relation to vessel walls; and (3) presence in the cytoplasm of the more pleomorphic cells of the grumous basophilic material above mentioned. Application of immunohistochemistry with specific marker like HMB45, smooth muscle actin and other melanocytic and myogenic marker should be carried out to confirm the suspicion of AML in any of these circumstances.

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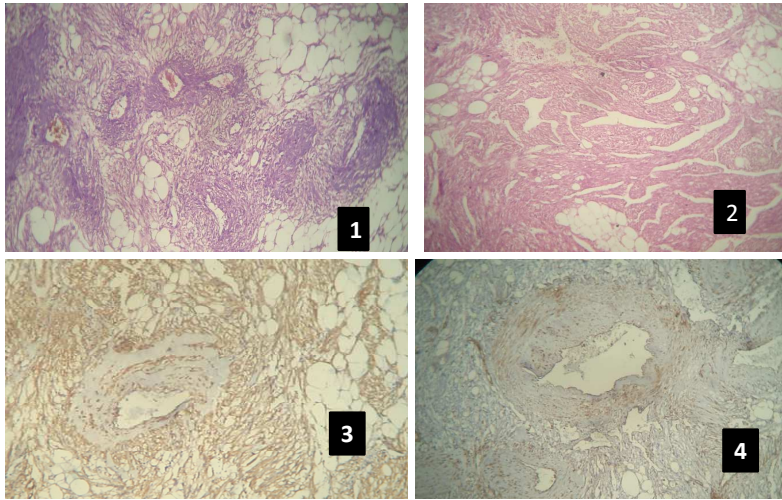


Figure 1 & 2: Admixture of spindle cell, adipose tissue & thick walled blood vessel;  
IHC showing smooth muscle actin positivity (3) & HMB 45 positivity (4)