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CASE REPORT

RENAL ANGIOMYOLIPOMA- A RARE PECOMA

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ABSTRACT:
Angiomyolipoma, though a relatively rare entity is yet the most common benign mesenchymal neoplasm of the kidney. This unusual lesion is composed of abnormal blood vessels, smooth muscles and mature adipose tissue elements. It has an incidence of 0.3-3% and arise from the mesenchymal elements of the kidney. It may occur as an isolated lesion or as a part of the syndrome associated with tuberous sclerosis. Angiomyolipoma belongs to the family of PEComa namely; perivascular epithelioid cell tumors which are mesenchymal tumors composed histologically of perivascular epithelioid cells. It has characteristic clinic-pathological features and its recognition is crucial for appropriate for patient management.
Key words: Angiomyolipoma, Benign neoplasm, Mesenchymal elements, PEComas, Renal.

INTRODUCTION

Renal angiomyolipoma are benign tumors with varying proportions of mature adipose tissues, smooth muscles and thick walled blood vessels. In about 5% of tumors adipose tissue is detected only by microscopy.[1]

Angiomyolipoma has been considered as a hamartoma and not a true neoplasm by many. But recently the clonal nature of this neoplasm has been demonstrated[2]

Angiomyolipoma is assigned to a family of neoplasm called perivascular epithelioid cell tumors (PEComas)[3-9]

The tumor has a distinctive cell type namely, the perivascular epithelioid cell or PEC which has no known normal tissue counterpart but shows morphological immunohistochemical, ultrastructural and genetically distinctive features. Morphologically the cells have an epithelioid appearance with clear to granular cytoplasm, round to oval centrally located nuclei and inconspicuous nucleoli in a typically perivascular location.[10-14]

CASE SUMMARY:

A 56 year old female patient presented with complaints of right sided flank pain and haematuria of 2months' duration. Laboratory investigations revealed decreased Haemoglobin and normocytic hypochromic blood picture. Ultrasonography showed a hyperechoic area measuring 7x3cms located in the cortical area of the right kidney associated with posterior acoustic shadowing. No evidence of vascularity was seen. A provisional clinic-radiological diagnosis of benign renal neoplasm in the upper pole of the right kidney was arrived at. The patient underwent laprotomy with mass resection and the mass was submitted for histopathological examination.

Histopathological findings:

Gross examination showed a single globular mass measuring 7x4x3 cms. Cut section showed an encapsulated grey-brown to grey-yellow mass with focal grey-white areas. (Figure 1)

Microscopy showed a well circumscribed benign mesenchymal tumor with an admixture of mature adipose tissue, smooth muscles and proliferated blood vessels. (Figure 2) The smooth muscles were also seen as individual fibres and were spindle shaped to epithelioid in appearance with round to oval bland nuclei. Occasional cells showed mild nuclear pleomorphism. Some of the blood vessels were large and congested with thickened walls. The normal renal parenchyma adjacent to the tumor was compressed.

Histopathological diagnosis - Renal Angiomyolipoma.

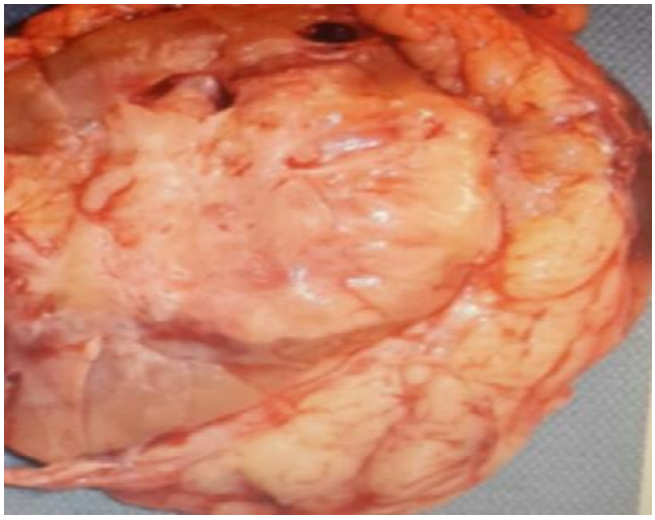

Figure 1: Globular mass measuring 7x4x3 cms. Cut section grey-brown to grey-yellow with focal grey-white areas

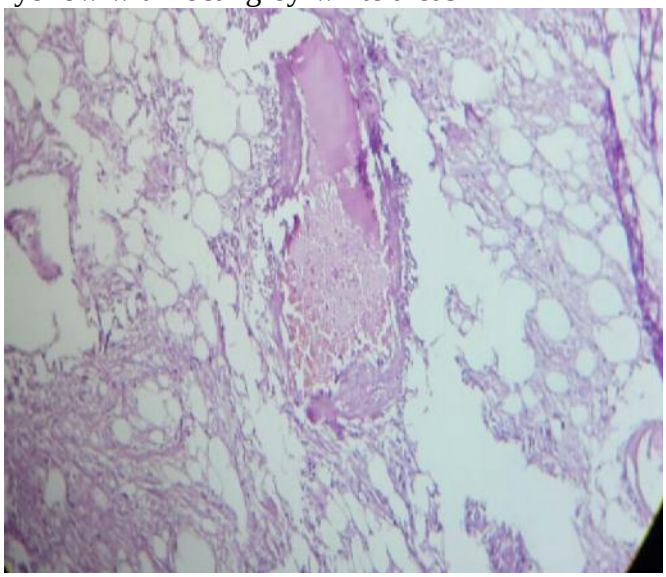

Figure 2 Microscopy showing benign mesenchymal tumor with an admixture of mature adipose tissue, smooth muscles and proliferated blood vessels. (H&E X45)

DISCUSSION:

Renal angiomyolipoma is an unusual benign renal lesion comprising 0.3-3% and about 1% of surgically removed renal tumors.[15]The tumor has an distinctive female predominance with a female to male ratio of 4:1.[7&15] as in our case. The mean age of occurrence is 53.9.[3,7&15]The common clinical presentation includes haematuria (33%), anemia (26.7%), hypertension (20%) and flank pain (13%) which was also seen in our case. Renal angiomyolipoma has clearly defined radiological characteristics in the form of a well-defined echogenic lesion which was also observed in our case. Classic renal angimyolipoma may have muticentric involvement but have always behave in a benign manner.[4] However, Bilaterality or multicentricity were ruled out in our case by radiology. The management of renal angiomyolipoma is based on clinical presentation, the size of the tumor, bilaterality and malignant potential.[16] Nephron sparing selective kidney or conservative surgery is the treatment of choice in cases without haemodynamic instability or when renal cell carcinoma is not suspected. Because of its known potential for malignancy a close follow up is required following surgery. Our patient is symptom free on follow-up 6 months after surgery.

CONCLUSION:

Renal angiomyolipoma is an uncommon benign tumor which may pose a diagnostic challenge to the clinician, the radiologist and the pathologist alike. The presence of adipose tissue in the tumor is highly suggestive.

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