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CASE REPORT

RENAL ANGIOMYOLIPOMA

M. Venu Gopal[1], C. Padmavathi[2], Karri Maruthi Devi[3], C. Aparna[4], Ramya Katta[5]

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ABSTRACT: A 60 years old female patient presented with flank pain. The radiological features were suggestive of angiomyolipoma of the kidney. The clinical features, gross and microscopic and immunohistochemical findings are presented.

KEYWORDS: Angiomyolipoma, Perivascular epithelioid cell tumor, HMB 45.

INTRODUCTION: Angiomyolipoma of the kidney is an uncommon benign mesenchymal tumor of the kidney and it belongs to the family of perivascular epithelioid cell tumors.[1] It is composed of blood vessels, adipose tissue and smooth muscle like cells. The smooth muscle cells are positive for HMB 45.[2]

Renal angiomyolipomas are usually associated with tuberous sclerosis and in these situations they present as bilateral small and asymptomatic nodules. In cases which are not associated with tuberous sclerosis they present as unilateral large and symptomatic tumors [3]. These tumors clinically mimic renal cell carcinoma.[3]

We report a case of angiomyolipoma of the kidney not associated with tuberous sclerosis.

CASE REPORT: A 60 year old female patient presented with intermittent flank pain for one year. Ultrasonography showed a large renal mass. She had no personal history or family history suggestive of tuberous sclerosis. Laboratory studies were normal.

CT scan showed a large heterogenous enhancing well defined mass with fat. Kidney was displaced and there was a mild displacement of IVC and aorta. A diagnosis of angiomyolipoma was made. In view of large size, nephrectomy was done.

Gross examination showed a single grey brown mass of size 18x14x11 Cm. Cut section showed a well circumscribed tumor with thin rim of normal kidney (Figure 1). The tumor grossly appeared yellow. Microscopic examination showed a tumor composed of thick blood vessels, smooth muscle cells and adipocytes. There were collections of epithelioid cells with abundant clear cytoplasm in some areas and pink cytoplasm in some areas. The epithelioid cells were seen around the blood vessels. Epithelioid cells constitute less than 20% of the tumor. There was no evidence of atypicality or necrosis or mitotic activity (Figure 2). IHC for HMB45 and SMA showed positivity (Figure 3). A diagnosis of Angiomyolipoma of kidney was made.

DISCUSSION: Angiomyolipoma is a benign mesenchymal tumor composed of blood vessels, smooth muscle cells and adipose tissue.[4] Angiomyolipomas belong to a family of PE Comas [Perivascular epitheloid cell tumors]. The incidence is 0.3% to 3%.[5] Females outnumber males.

The tumor may be multifocal, exhibit atypical histological features with involvement of regional lymphnodes.[6] Angiomyolipoma of kidney is a benign mesenchymal tumor constituting 0.1 to 2.1% of all renal masses.[3] Renal angiomyolipomas may or may not be associated with tuberous sclerosis.

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Those cases associated with tuberous sclerosis are bilateral, small multiple and asymptomatic. However, those associated with out tuberous sclerosis are unilateral, large and symptomatic tumors.[3] AML of the kidney is considered as benign lesion even though it shows nuclear pleomorphism and mitotic activity. AML showing extension in to inferior vena cava or in to regional lymph nodes is considered as multifocal origin rather than as metastasis. The commonest age group is between 40-60 years.

An Epithelioid variant of angiomyolipoma of the kidney is described which is capable of metastasis or recurrence.[6]

The patients usually present with flank pain, hematuria, and palpable mass or renal failure.

The preoperative diagnosis of the tumour is made by combined imaging of computed tomography or ultrasonography. CT scan finding include low attenuation areas of fat and ultrasonography shows hyper echogenecity from multiple fat and nonfat interfaces within the tumour.[3]

Macroscopically, the tumours are medium to large sized showing areas of hemorrhage and necrosis. Angiomyolipomas of the kidney are unencapsulated tumours which are well demarcated from the adjacent kidney. Histologically, the typical triphasic pattern of tumours are composed of mature fat, thick walled blood vessels and smooth muscle.[2] The smooth muscle cells appear to arise from the blood vessels in radial fashion. The smooth muscle cells may appear spindle shaped or rounded giving the appearance of epithelioid cells. The blood vessels are thick walled but the normal elastic content of the arteries is not seen. The lipomatous component appears as mature adipose tissue, but in few areas, cells appearing as lipoblasts are seen. Angiomyolipomas express melanocyte markers.[4]

The treatment of angiomyolipoma depends upon the size of the tumour and radiological features of malignancy. Tumours which are more than three centimeters and are symptomatic require surgical intervention. Follow up is needed for tumours which are asymptomatic and less than three centimeters in size. Basing on radiological features renal preserving surgery should be performed. In case of suspicion of malignancy radical nephrectomy is the treatment of choice.[6]

In our case there was no association with tuberous sclerosis, It was unilateral and histologically was not associated with a typicality.

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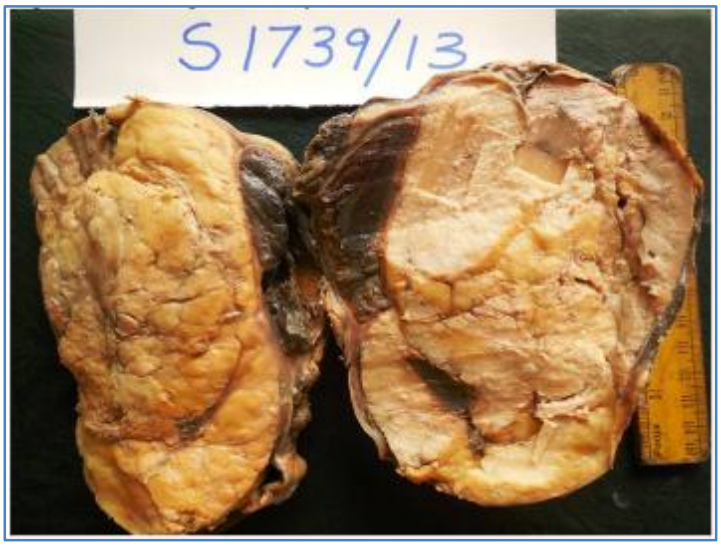

Fig. 1: Showing the kidney with Tumor

Figure 2a: low power view of the tumor with the normal kidney.
2b: low power view of the tumor.
2c: spindle cell and epithelioid cells.
2d: tumor cells around thick walled blood vessels.

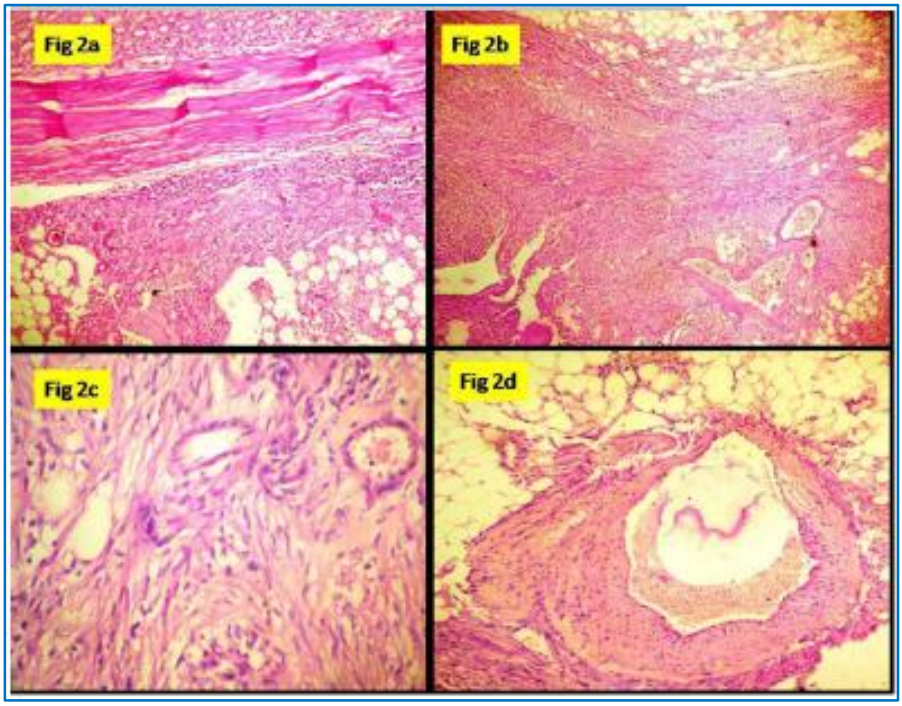

Fig. 2

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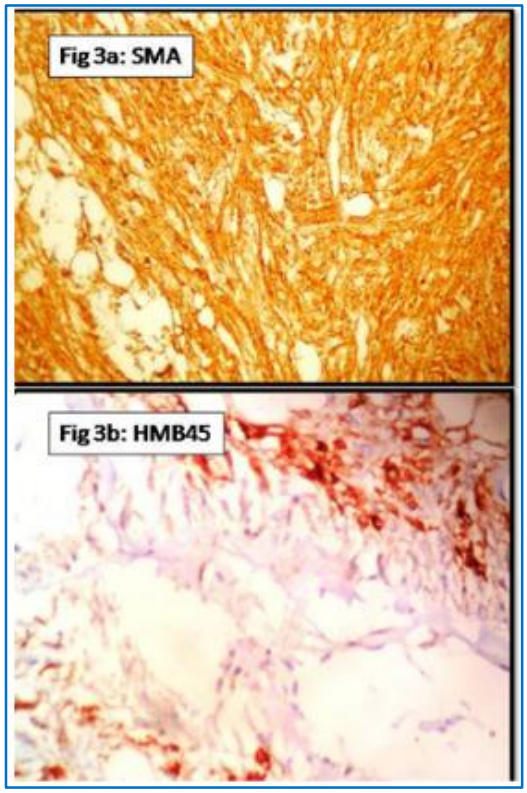

Fig. 3a and 3b: Showing SMA positivity and HMB positivity

AUTHORS:

1. M. Venu Gopal
2. C. Padmavathi
3. Karri Maruthi Devi
4. C. Aparna
5. Ramya Katta

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Pathology, Guntur Medical College, Guntur.
2. Professor, Department of Pathology, Guntur Medical College, Guntur.
3. Assistant Professor, Department of Pathology, Guntur Medical College, Guntur.
4. Associate Professor, Department of Pathology, Guntur Medical College, Guntur.
5. Post graduate, Department of Pathology, Guntur Medical College, Guntur.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. M. Venu Gopal,
Associate Professor,
Department of Pathology,
Guntur Medical College,
Guntur, Kannavanthota-522004,
Andhra Pradesh.
E-mail: drmvgopal@yahoo.com

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