Case Report

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HMB-45 Negative Adrenal Angiomyolipoma With A Synchronous Adrenal Adenoma – An Unusual Association

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ABSTRACT

Background: Angiomyolipoma (AML) is an uncommon mesenchymal tumour usually found in the kidney. The most common extrarenal site is usually the liver. We report a case of adrenal AML with a predominant vascular component co-existing with an adrenal adenoma.

Case Report: A fifty-year-old male presented with two months history of dysuria. Computed tomography showed a heterogeneous lesion involving the right adrenal gland abutting the right kidney and adjacent liver capsule. Urine noradrenaline levels were elevated. Adrenalectomy specimen showed an encapsulated mass measuring 8 x 5 x3.5 cm with a lobulated, grey to tan brown cut surface with areas of haemorrhage and tiny cystic spaces. Another yellowish nodule measuring 1.3 x 1.2 x 1 cm was also noted at one pole. Microscopy showed a neoplasm composed predominantly thick-walled vascular channels admixed with foci of smooth muscle bundles and adipocytes. Yellowish nodule showed features of adrenal adenoma. HMB-45 was negative and CD34 positivity highlighted the thick and thin walled vessels.

Conclusion: We present the very first case of HMB-45 negative adrenal AML with a predominant vascular component and a synchronous adrenal adenoma.

Keywords: *Adenoma, Adrenal Angiomyolipoma, HMB-45 negative*

Introduction

Angiomyolipoma (AML) is a class of soft tissue tumours of melanocytic differentiation included in the family of neoplasms derived from perivascular epithelioid cells. It is a very rare mesenchymal tumour most commonly found in the kidney. Extrarenal involvement is extremely rare with the most common site of involvement being the liver. Adrenal involvement is very rare and is usually an incidental finding during investigation for abdominal symptoms.

Here we report a rare case of adrenal AML with a predominant vascular component with a coexisting adrenal adenoma. Lack of expression of melanocytic markers (HMB-45) in our case makes it even unique.

Case Report

A fifty year old male, with no comorbidities, presented to the OPD with a two months history of dysuria. His general and systemic examinations were within normal limits. Urine routine and microscopy showed increased number of pus cells following which he was treated for urinary tract infection. A follow up USG abdomen detected an incidental mass in the right suprarenal region (Fig.1a).

Renal function tests were within normal limits. He was further evaluated with a CT scan of the abdomen and pelvis which showed a heterogeneous lesion involving the right adrenal gland with a washout of less than 60% on delayed images. The lesion was seen to abut the right kidney and adjacent liver capsule. Left adrenal appeared normal (Fig.1b).

Based on the CT findings, a possibility of phaeochromocytoma was suggested. Elevated urine noradrenaline levels were noted - 126.96 µg/24hrs (normal range - <90 µg/24hrs) whereas adrenaline levels were within normal limits.

Open right adrenalectomy was performed. Gross examination showed a nodular mass measuring 8x5x3.5cm. Cut section showed an encapsulated growth measuring 6.8x5x3.7 cm with a grey brown to tan appearance with areas of haemorrhage and tiny cystic spaces. Another well defined yellow nodule measuring 1.3x1.2x1 cm was also noted at one pole on serial sectioning (Fig.1c).

Microscopy from the largest nodule showed a neoplasm composed of predominantly thick walled blood vessels along with thin walled vascular channels admixed with foci of smooth muscle bundles and adipose tissue (Fig.2a-c).

Sections from well circumscribed yellow nodule showed a neoplasm composed of sheets and nests of tumour cells with clear to eosinophilic foamy cytoplasm and vesicular nuclei (Fig.2d). Compressed adrenal parenchyma noted in the periphery.

The possibility of angiomyolipoma with a predominant vascular component and a coexisting adrenal adenoma was considered.

IHC was done to confirm the same. Vimentin was diffusely positive. S-100 showed patchy positivity suggestive of normal adrenal tissue. HMB-45 and Melan A were negative. CD34 highlighted both thick and thin vascular channels. SMA was positive in the vascular and smooth muscle component of neoplasm. Inhibin was positive in adrenal adenoma nodule. Ki-67 was very low (≈1%) (Fig.3 a-f).

Considering morphology and IHC, a conclusive diagnosis of HMB-45 negative adrenal angiomyolipoma with a predominant vascular component and a synchronous adrenal adenoma was made.

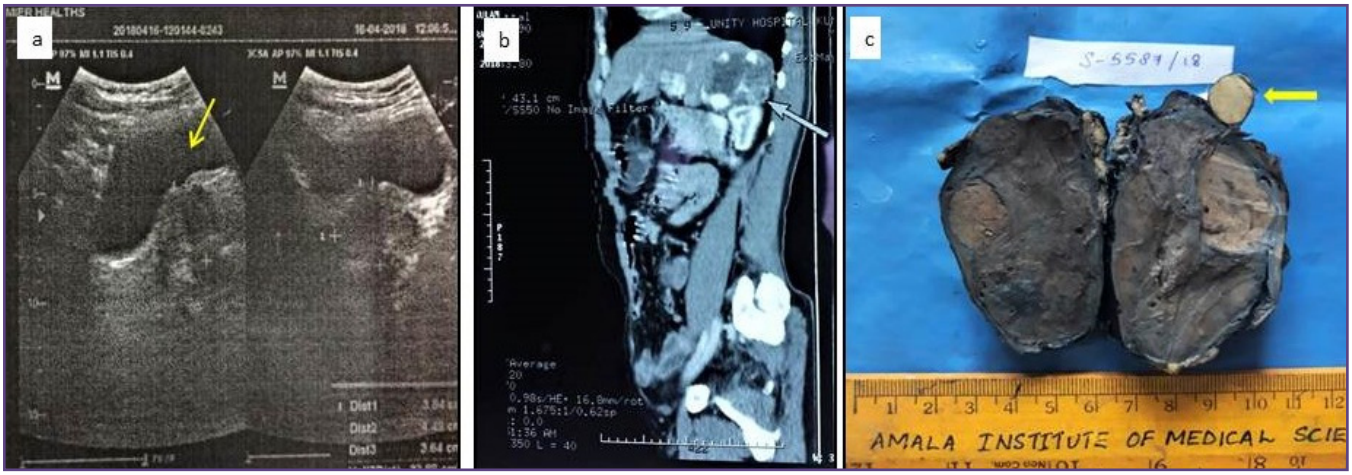

Fig. 1 (a): USG Abdomen showing a well defined round hypoechoic mass lesion (yellow arrow) in right suprarenal region. (b): CT scan showing a heterogeneous lesion in the right adrenal gland (blue arrow). (c): Nodular mass measuring 8x5x3.5cm. Cut section showed a capsulated growth with a grey brown to tan cut surface interspersed with areas of haemorrhage and tiny cystic spaces. Another well defined yellow nodule measuring 1.3x1.2x1 cm was also noted (yellow arrow).

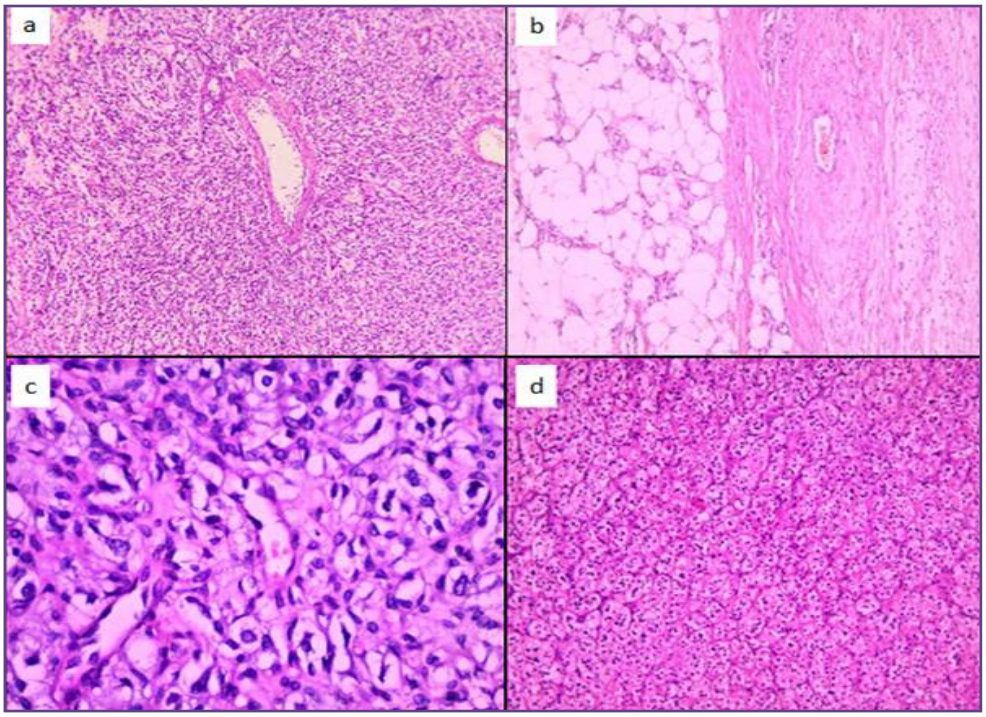

Fig. 2(a-c): Microscopy shows a neoplasm composed of predominantly thick and thin walled blood vessels admixed with foci of smooth muscle bundles and adipose tissue. (a & b : H&E – 40X; c: H&E – 400X). (d) : Microscopy of the yellowish nodule showing sheets and nests of tumour cells with clear to eosinophilic foamy cytoplasm (H&E – 100X).

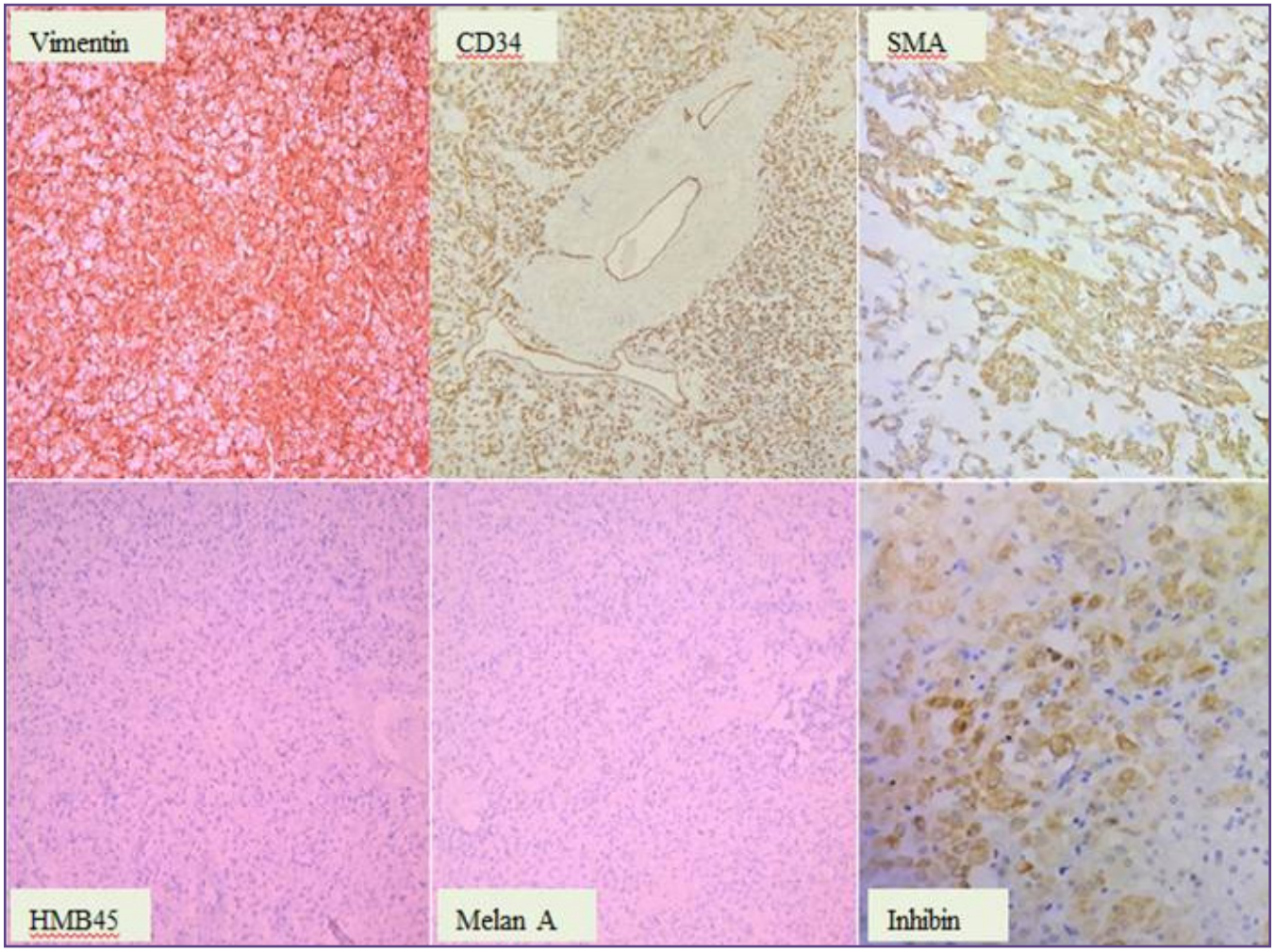

Fig. 3: (a – f) Immunohistochemistry staining: Vimentin , CD 34 , SMA , HMB -45 , Melan A. Inhibin is positive in adrenal adenoma.

Discussion

Angiomyolipomas are rare mesenchymal tumours. Though kidney is the most common site, extrarenal sites are also reported. Adrenal involvement is very rare with an incidence of 0.3-3% according to literature[1]. Sixteen cases have been reported till date. AML can occur in isolation or as part of a syndromic condition. Isolated neoplasms are usually asymptomatic. 20 to 50% have been reported to be associated with tuberous sclerosis in various literatures[1-3]. Though commonly benign, metastasis have been reported. CT and USG are found to be helpful in diagnosis owing to the high fat content in the tumour cells.

There are many theories regarding the pathogenesis of this condition. The most widely accepted theory states that this tumour is a metaplastic change in the reticuloendothelial cells of the adrenal blood capillaries in response to various stimuli such as necrosis, infection or stress.[1]

These tumours have a heterogeneous cell composition and consists of adipose tissue, vascular component and smooth muscle cells. It should be noted that any one of these components may predominate as in our case. Hence, a diligent search for all components is necessary to arrive at this diagnosis. Perivascular epithelioid cells are the characteristic feature and are usually positive for muscle markers and HMB-45. The latter is surprisingly negative in our case, which can be attributed to the rarity of epithelioid cells in the neoplasm. [4]

After extensive literature search, we believe that, this is the very first case of HMB-45 negative adrenal angiomyolipoma with a predominant vascular component and an adrenal adenoma to be reported.

Conclusion

Adrenal angiomyolipoma is a very rare, benign mesenchymal tumour. It is commonly an incidental

radiological finding and should be carefully differentiated from other retroperitoneal tumours. Imaging techniques are helpful owing to the adipose and vascular component. A careful histopathological approach can confirm this diagnosis. This entity is seen on the rising trend as a result of effective screening and extensive use of imaging modalities.

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Competing Interests
NIL

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