Case Report

Extrarenal retroperitoneal angiomyolipoma with oncocytoma

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ABSTRACT

The simultaneous presence of renal angiomyolipoma and oncocytoma is a rare occurrence. Extrarenal retroperitoneal angiomyolipoma is an even more rare neoplasm, and its simultaneous presence with renal oncocytoma has not been documented. We present herein the first case to be reported in English literature.

INTRODUCTION

Angiomyolipomas (AMLs) comprise about 1% of all renal masses. In contrast, extrarenal retroperitoneal AMLs (ERAMLs) represent extremely rare tumors with <60 reported cases.[1] Oncocytoma, originating from renal tubular cells, is a benign epithelial tumor that accounts for about 3%–5% of surgically resected renal neoplasms in adults. There have been only 16 reported cases of simultaneous renal angiomyolipoma and oncocytoma. Of these cases, all except one were female and three were associated with the tuberous sclerosis complex.[2]

While there have been case reports of oncocytic angiomyolipoma, there have been no reports of ERAMLs along with the simultaneous presence of renal oncocytoma, we present the first case.

CASE REPORT

A 60-year-old female presented with recurrent episodes of breathlessness and gradually increasing left flank pain for 2 years. Contrast-enhanced computed tomography (CECT) of the thorax and abdomen done a year ago showed bilateral pulmonary cysts with a right small pneumothorax, a 6.9 cm heterogeneously enhancing mass in the left kidney, with a nonenhancing central scar, along with multiple enhancing lymph nodes in the left hilar, para-aortic, interaortocaval, and precaval regions, largest measuring 4 cm [Figure 1]. Repeat CECT was done; there was resolution of the pneumothorax but an increase in size of left renal mass to 8.3 cm. She was not found to have any skin lesions. Our preoperative provisional diagnosis was oncocytoma/hybrid chromophobe-oncocytic renal carcinoma in a case of Birt–Hogg–Dubé syndrome (BHD). A laparoscopic left radical nephrectomy was performed along with lymphadenectomy of the hilar and para-aortic lymph nodes [Figure 1]. The histopathology of the renal mass showed round to polygonal cells arranged in nests and trabeculae, with abundant amount of granular eosinophilic cytoplasm and centrally placed hyperchromatic nuclei [Figure 2a]. IHC stain for HMB45 and vimentin was negative, consistent with oncocytoma. Sections from the lymph node showed proliferation of smooth muscle cells arranged in fascicular pattern. Few dilated vesicular channels lined by a single layer of endothelial cells were also present [Figure 2b]. IHC stain was positive for HMB45, vimentin, and SMA. It was negative for S-100, desmin, and CK7 consistent with ERAML. The location of the AML was from the extrarenal retroperitoneal hilar and para-aortic lymphnodes and was completely separate from the renal oncocytoma. We initially thought our case to be variant of BHD or tuberous sclerosis

Access this article online	
Quick Response Code:	Website: www.indianjurol.com
	DOI: 10.4103/iju.IJU_249_17

Received: 13.08.2017, **Accepted:** 08.10.2017

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

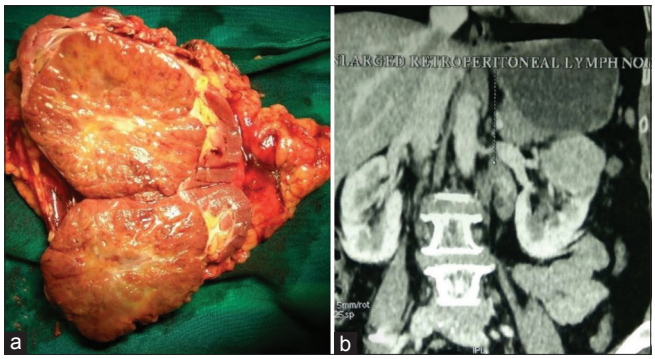

Figure 1: (a) Gross cut section - well-circumscribed solid tumor, mahogany brown, with a central scar, involving upper and mid pole of kidney. (b) Contrast-enhanced computed tomography scan image of the abdomen showing heterogeneously enhancing upper pole mass with enhancing enlarged lymph nodes in left hilar, para-aortic, and interaortocaval regions

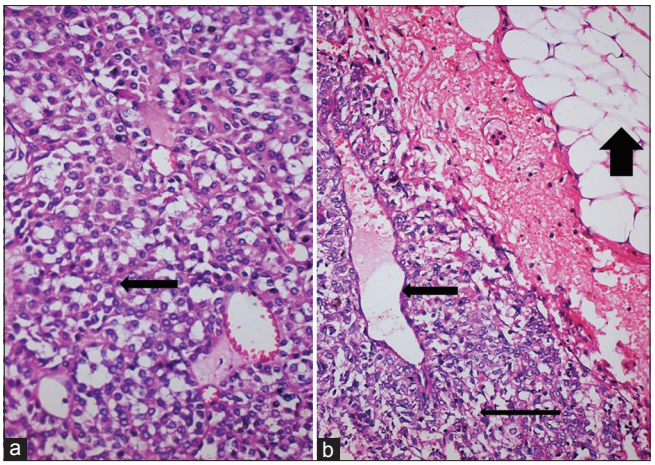

Figure 2: (a) Tumor cells having polygonal cell with clear or densely eosinophilic cytoplasm, large hyperchromatic nucleus (arrow). (b) Triphasic tumor comprising of mature adipose tissue (thick arrow) ill-formed thick-walled blood vessels (thin arrow) and smooth muscles (thinnest arrow)

complex, but to our surprise, genetic testing was negative for FLCN, TSC1, and TCS2. In addition, we also performed transmission electron microscopy on the tumor which showed multiple mitochondria and helped to rule out oncocytic angiomyolipoma. At 5-month follow-up, the patient was doing well.

DISCUSSION

Oncocytomas arise from intercalating cells of the cortical collecting ducts. They are considered benign, and the majority are single and unilateral. The characteristic histological features of oncocytomas are the dense eosinophilic cytoplasm, generally uniform nuclei, and abundant mitochondria.[2] On gross examination, there is little or no evidence of necrosis or hemorrhage, occasionally a central scar is observed.[3] As it is difficult to differentiate it from renal clear cell carcinoma on imaging, nephrectomy remains the treatment of choice.[2]

Most renal AMLs (approximately 70%–80%) were sporadic, but the remainders (approximately 20%–30%) were related to TSC. By contrast, 80% of patients with TSC have associated renal AMLs.[4] Hence, if AMLs are found incidentally in nephrectomy specimens together with other tumors, it is important to exclude TSC retrospectively. On the other hand, only 5 ERAMLs have been reported with tuberous sclerosis.[1]

AMLs are composed of three tissue elements: mature adipose tissue, thick-walled blood vessels, and smooth muscle cells. AMLs were originally classified as hamartomas but are now thought to belong to the family of perivascular epithelioid cell tumors.[1] Positive immunoreactivity for HMB45, a monoclonal antibody, raised against a melanoma-associated antigen, is characteristic of AMLs and can be used to differentiate them from other similar appearing lesions such as liposarcomas, lipomas, leiomyosarcomas, or leiomyomas. In the setting of oncocytoma and tuberous sclerosis, oncocytic angiomyolipoma, ,where the cells look like oncocytes having eosinophilic granular cytoplasm but have focal reactivity for HMB45 and are negative for epithelial markers, has to be ruled out.

In contrast to renal AMLs, extrarenal AMLs are very rare. Most common site is the liver, the retroperitoneum is the second most common location, and about 30 cases have been reported in literature. There is a wide age distribution in these cases (28–73 years); it is more common in females. The symptoms of retroperitoneal AMLs are nonspecific and associated with gastrointestinal symptoms.[1]

Retroperitoneal ERAMLs presents a unique diagnostic challenge since they can mimic other retroperitoneal benign and malignant tumors. Ultrasound and CT scan can correctly diagnose renal AMLs in 86% of cases. ERAMLs, however, are more difficult to diagnose on imaging as they often lack fat densities.[1] The definitive diagnoses of ERAMLs are dependent on histopathological examination.[4] As with AMLs, immunoreactivity to an HMB45 stain is also to differentiate ERAMLs from other retroperitoneal tumors. ERAMLs should be analyzed for mitotic index and the presence of epithelioid variant as these characteristics may be associated with distal metastasis and disease recurrence.

Although the majority of ERAMLs are benign, two cases of metastatic and recurrent ERAMLs have been reported. The definite mechanism of recurrence is not known.[1] Rare cases of AML malignant transformation with lymph node involvement have been documented in the literature; however, all cases involved patients with renal AMLs and tuberous sclerosis. It has been suggested that AMLs should be regarded as tumors that often exhibit benign biological behavior but a subset of such tumors would metastasize; therefore, perhaps, AMLs should be regarded as slow-growing malignant tumors with the potential to metastasize.[1]

Surgical excision is indicated in cases of symptomatic, complex appearing, radiologically enlarging ERAMLs, which tend to be associated with a higher potential to bleed spontaneously. Surgical excision has also been recommended for histological examination when it is difficult to differentiate suspected ERAMLs from other lesions involving the retroperitoneum; the rest can be managed conservatively.[1] Long-term follow-up is indicated, as recurrent and malignant cases have been reported, as well as to monitor the size.

Birt–Hogg–Dubé syndrome and tuberous sclerosis complex have to be considered in the differential diagnosis of multiple pulmonary cysts, AML, and oncocytoma kidney and were ruled out in our case by genetic analysis.

Acknowledgment

We would like to thank Dr Lovelesh Nigam and Dr Pranjal Modi.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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How to cite this article: Ali AM, Rizvi SJ, Kanodia KV. Extrarenal retroperitoneal angiomyolipoma with oncocytoma. Indian J Urol 2018;34:82-4.