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Renal Angiomyolipoma and Pulmonary Lymphangioleiomyomatosis: A Common Association Often Overlooked

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Abstract

Background: Renal angiomyolipoma is the most common benign tumor of kidney. It is often associated with pulmonary lymphangioleiomyomatosis specially when it is associated with tuberous sclerosis. This association is often missed and the patient bears the risk of hyperinflation lung injuries during or after the treatment of renal angiomyolipoma. *Case Report*: We hereby report a patient with pain and swelling in left side of the abdomen suggestive of a renal mass. She underwent left simple nephrectomy which eventually came out to be renal angiomyolipoma. She had other characteristics of tuberous sclerosis for which computed tomography of thorax and guided biopsy was done which revealed the presence of pulmonary lymphangioleiomyomatosis. *Conclusion*: We should be aware of the co-existence of both renal angiomyolipoma and pulmonary lymphangiomyomatosis specially when other features of tuberous sclerosis are present. We should ensure regular follow-up and treat both the diseases to ensure the complete recovery of the patient.

Keywords: Nephrectomy, Pain, Renal Carcinoma, Seizure, Tuberous Sclerosis.

Description of the Clinical Image

A 34-year-old woman presented in the outpatient Urology department of JIPMER in March, 2020 with pain and swelling in left side of the abdomen. She had history of seizures for last 15 years. On evaluation, she had facial adenoma sebaceum with a palpable left abdominal mass. CT urography showed bilateral renal angiomyolipomas with largest adenoma of 18×9 cm size in the left kidney. NCCT brain showed subependymal calcifications in lateral ventricles. CT thorax showed multiple thin-walled cysts and round nodules in both lungs. CT guided biopsy of lung lesions came out as pulmonary lymphangioleiomyomatosis. She underwent left simple nephrectomy. Histopathologically, tumor had spindle cells, mature adipose tissue and smooth muscle positive for HMB45 characteristic of angiomyolipoma. Oral sirolimus was given for lymphangioleiomyomatosis and anti-epileptic medications were given for seizures. Patient recovery was uneventful and she is on regular follow up for last 1 year.

Discussion

Renal angiomyolipoma (AML) is a mesenchymal tumor composed of adipose tissue, smooth muscle cells and thick-walled vessels. Tuberous sclerosis (TSC) has been present in approximately 10% of cases of renal AML along with pulmonary lymphangioleiomyomatosis (LAM) [1]

Pulmonary lymphangioleiomyomatosis is a rare progressive disease affecting women, primarily in their reproductive years with no effective cure [2]. It is caused by mutation in TSC2 gene encoding tuberin which regulates cell growth by inhibiting m-TOR complex. It is characterized by non-neoplastic proliferation of atypical smooth muscle cells within the lung parenchyma and elsewhere

leading to progressive loss of lung function. Both LAM and angiomyolipoma belong to a family of tumors termed as perivascular epithelial cell tumors (PEComas) [3]. Tuberous sclerosis is characterized by the inactivating mutations of either TSC1 (9q34.3) or TSC2 (16p13.3), which encode hamartin and tuberin, respectively. Renal angiomyolipomas have been reported in up to 100% of patients with TSC-LAM, and in up to 50% of those with sporadic LAM [4]. AML usually predates the onset of pulmonary disease.

Surgery or angioembolisation is the choice of intervention in patients with renal angiomyolipomas. Sirolimus, an mTOR inhibitor is effective in stabilizing the disease progression and improve quality of life in LAM along with reduction in AML volume [5]. Patients with angiomyolipoma having pulmonary symptoms and those associated with TSC should be screened for LAM. Physicians should be aware of the potential pulmonary complications and adopt precautionary strategies to avoid hyperinflation lung injuries. These patients should be counseled for regular pulmonary follow up and appropriate management.

Contributors: SK, RM: Clinical decision making, surgical management of the patient, designing of the manuscript, review of the drafted manuscript; AKP: Patient care in pre and post operative period, follow up of the patient, acquisition of data, literature review; LND: Clinical decision making, critical review and appraisal of the manuscript; PG: Assisting the surgery, follow up of the patient in outpatient department, review of literature, review of the drafted manuscript. AKP will act as a study guarantor. All authors approved the final version of this manuscript and are responsible for all aspects of this study.

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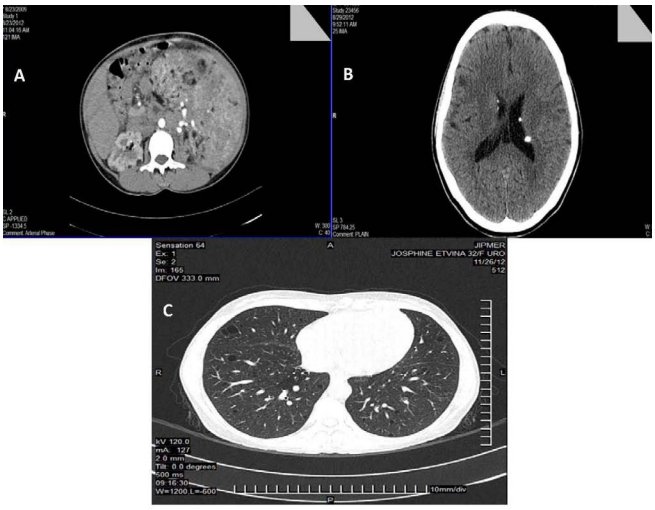

Fig.1 (a): *Abdominal computed tomography showing bilateral AML largest located in left kidney size 18×9 cm.* **(b):** *Brain tomography revealed multiple, small subependymal calcification.* **(c):** *Chest tomography describing multiple small air cysts in bilateral lungs suggestive of pulmonary lymphangioleiomyomatosis.*

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