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RESEARCH ARTICLE

CASE REPORT OF LYMPHANGIOLEIOMYOMATOSIS(LAM).

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Key words:-

Abstract

Background:-

Lymphangioleiomyomatosis(LAM) is a multisystem disorder predominantly affecting women of child bearing age. It is characterized by multiple cystic lung lesion, abdominal angiomyolipomas and lymphatic abnormalities. Pathologic features are caused by proliferation of smooth muscle cells. It may occur as Inherited form in association with tuberous sclerosis(TSC) and as sporadic form

Case report:-

A 21 year old non smoker female presented with progressive shortness of breath since last 4 yrs and currently on LTOT. She also complained of cough with scanty expectoration.she was prescribed ATT twice at periphery for same complain. There was no history of haemoptysis or abdominal pain. Clinical examination included clubbing (Gr 2), raised JVP, crackles in b/l interscapular,infra axillary and infrascapular area. Her family history includes one sister dying of similar complain undiagnosed 7 yrs back.

Investigations:-

Hb- 14g/dl, TLC – 7500 with normal DLC. Liver and renal profile was normal.HIV was negative and sputum was negative for AFB. CXR revealed reticular and cystic shadow in mid and lower zone with cardiomegaly. CT SCAN Chest showed diffuse cystic lesion of different size and diffuse ground glass opacity and significantly enlarged pulmonary trunk(40mm) and presence of PAH.ECHO revealed mildly dialated RV. Abdominal CT was normal

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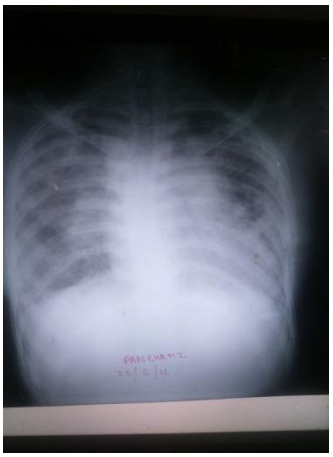

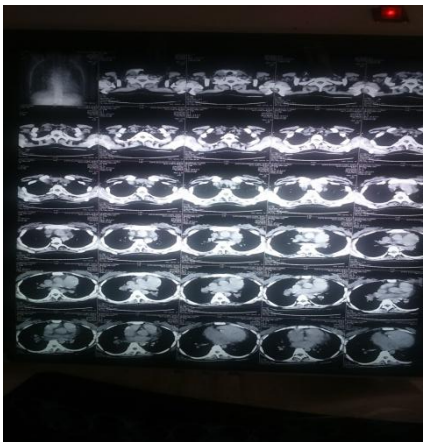

Result:-

patient was diagnosed as Lymphngioleiomyomatosis with PAH and is on LTOT since 1 year and symptomatic treatment**.**

Conclusions:-

LAM presents a diagnostic challenge to differentiate it from other cause of ILD. A high index of suspicion is required in females of reproductive age with no history of smoking.HRCT remains the diagnostic imaging to differentiate it from LCH and emphysema owing to limited accessibility to Surgical Lung Biopsy (SLB)