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LYMPHANGIOMYOMATOSIS - A RARE INTERSTITIAL LUNG DISEASE (ILD)

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Abstract:

We report a case of a 26 years old Female with Lymphangiomyomatosis , a rare multi system disorder. Clinical history was sudden onset of chest pain and was operated for Left Renal Angiomyolipoma 4 years back. Her Chest X-Ray showed Left sided Pneumothorax. Her CT Thorax was suggestive of bilateral diffuse well defined cystic shadows distributed all over lung fields surrounded by normal Lung Parenchyma along with Left Pneumothorax, distinguishing features for pulmonary Lymphangioleiomyomatosis. Lymphangioleiomyomatosis is under diagnosed by clinicians, so awareness of this disorder may be helpful to reduce morbidity and mortality.

Keywords: Lymphangioleiomyomatosis, Pneumothorax, Renal Angiomyolipoma CT-Thorax

INTRODUCTION:

Lymphangiomyomatosis **(LAM)** is a unusual multifocal origin disease which typically involves lung, kidney and lymph and may be associated with the tuberous sclerosis (TS).**(1)** Commonly it affects women of reproductive age group with incidence of 1:400,000 . **(2,3)** Proliferation of abnormal smooth muscle causing obstruction of venules and lymphatics which further carry doggedness of dilated lymphatics**. (4) There are two types of p**resentation of lymphangiomyomatosis in the chest. In initial phase, immature muscle cells are proliferating in such a way that they cover alveolar walls, bronchioles, pleura and vessels, including lymphatic routes. In the later stages cystic lesions appears in lung with more proliferation of muscle cells throughout the lung.(**5)**

HISTORY

A 26 yrs. old female came to the TB and Chest Diseases OPD with complaints of Chest pain Left side for last 2 days. It was sudden in onset and not associated with Palpitation / sweating / Shortness of Breathing (SOB). Her Chest X-Ray was suggestive of left sided Pneumothorax. Inter

Costal Tube Drainage (ICTD) was done and lung expanded. Chest tube was removed after 4 days and Patient remained asymptomatic for next 1 year. After 1 year, she again developed left sided chest pain. Chest X-Ray was suggestive of Pneumothorax left side. Again ICTD was done and lung expanded. This time, her past medical records were reviewed and it was found that she was operated for Left Renal Angiomyolipoma four years back. Her CT Thorax was suggestive of well defined cystic shadows distributed all over lung fields surrounded by normal Lung Parenchyma along with Left Pneumothorax. Hence a diagnosis of Lymphangi-omyomatosis was made as the patient was a young female in reproductive age group with past history left renal Angiomyolipoma.

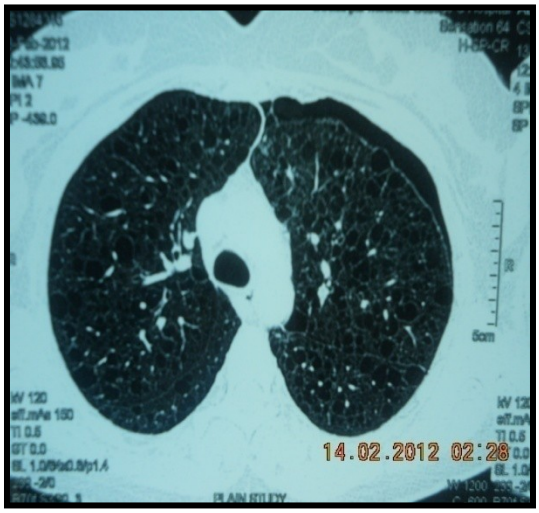

Figure No.1 CT scan Thorax

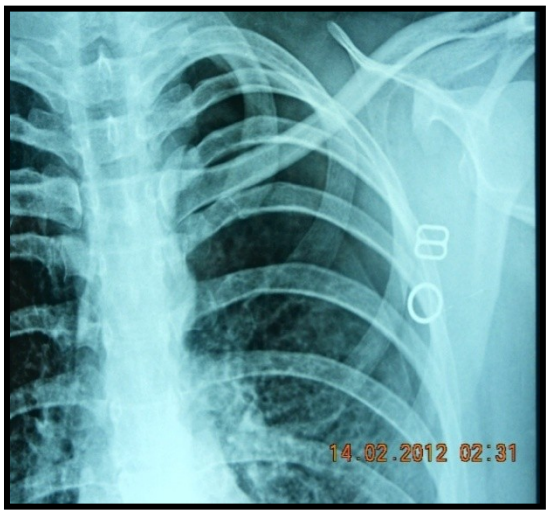

Figure No .2 Chest X-Ray

DISCUSSION

LAM is a rare disorder exclusively found in young females mainly between 30 and 49 years of age. It is characterized by abnormal proliferation of smooth muscle cells around pulmonary lymphatics, vessels and small bronchi.**(5)** Clinically patient presents with Chest pain, SOB, cough or Hemoptysis. Patient may develop Chylothorax or Pneumothorax. 1/3rd of patients may have Renal Angiomyolipoma.**(6)** lymphangio-leiomyomatosis is two types, one is sporadic and another is combined with tuberous sclerosis. Mutation in tumour suppressor genes on chromosome 9 (9q34) and on chromosome 16 (16p13.3) are root cause of this. **(7)**

Lymphangioleiomyomatosis commonly creates confusion with asthma, emphysema or pulmonary fibrosis. The diagnosis is made mainly on clinical findings and CT Thorax. A high-resolution CT scan can be very helpful in diagnosis of lymphangioleiomyomatosis correctly. **(8)** Rarely Lung Biopsy is required. The most common pulmonary function defects in

LAM are airflow obstruction and decreased lung diffusion capacity. With the disease progression, lung functions begin to decline with an average monthly rate of 7–9 mL of FEV1. Airflow obstruction was showed in about 60% of patients of LAM. This loss is caused by cystic destruction of lung parenchyma. **(8, 9)** Proliferating smooth muscle cell were from unknown origin and showed metastatic properties.Benign kidney tumours (angiomyolipoma) are also associated with 60% of cases of LAM.**(10)** The classic presentation of lymphangioleiomyomatosis is pneumothorax or chylothorax. Reccuent pneumothorax may suggest about the diagnosis of LAM as in this case and pneumothorax is managed by chemical or surgical pleurodesis **(10)**

Main Differential Diagnosis include Langer-han's cell Histiocytosis. Lymphangio-leiomyomatosis is managed by supportive treatment such as bronchodilator therapy, pulmonary rehabilitation, treatment of anxiety, oxygen therapy and eventually lung transplantation. Clinician should be careful about prescribing any medication which contain estrogen (An estrogen-MMP-driven process play a role in the destruction of lung parenchyma and may responsible for this condition among women).**(11)** Treatment options include Medroxy-progesterone acetate ,Tamoxifen , Gonadotropin releasing hormone agonists , Doxycycline and Sirolimus with varying results. In some cases Oophorectomy or Lung transplantation is indicated.**(12 ,13)**

CONCLUSION

Lymphangioleiomyomatosis sometimes under diagnosed by clinicians, awareness of this disorder may be helpful to reduce morbidity and mortality. Early and correct diagnosis of LAM through CT scan of women with TSC and who come with pneumothorax or nonspecific respiratory symptoms makes it possible to start proper treatment before permanent lung changes take place.

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Conflict of interest: None declared

Ethical approval: The study was approved by the institutional ethics committee

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