Pulmonary Lymphangiomyomatosis: A Rare Disease responsive to Progesterone

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Abstract

Lymphangiomyomatosis (LAM) is a rare multisystem disorder in women of child-bearing age. We present a case of a 28 year old lady who presented with cough and breathlessness. She had been diagnosed as a case of lymphangiomyomatosis by computer tomography of chest. She showed dramatic clinical improvement with hormonal therapy.

Keywords: **Lymphangiomyomatosis, Medroxyprogesterone.**

Introduction

Lymphangiomyomatosis (LAM) is a rare disease of women of child-bearing age. It is characterized by abnormal proliferation of spindle cells in lung parenchyma along the bronchioles leading to air trapping and formation of thin walled cysts. Rupture of these cysts leads to recurrent episodes of pneumothroax.[1]

Case Report

A 28 year old lady presented with history of cough and breathlessness for the last two years. She was taking bronchodilators but symptoms did not improve. On examination, it was found that she was having tachycardia with pulse rate of 104 beats/min and tachypnea with respiratory rate of 24/min. On chest examination, it was found that she had bilaeral rhonchi. Rest of the systemic examination was within normal limits. Her hemogram was normal with normal eosinophil count. Chest X-ray revealed prominent bronchovascular markings. Pulmonary function test revealed obstructive pattern. She was prescribed bronchodilators with steroids but no relief was observed. High resolution computer tomography (HRCT) of chest showed thin walled cysts in bilateral lung parenchyma predominantly in lower lobes with mosaic pattern suggestive of lymphangiomyomatosis (Fig. 1). USG abdomen and skeleton survey did not reveal any other abnormality. Bronchoscopic lung biopsy was planned but patient developed hypertension every time. The patient did not give consent for open lung biopsy. Atrial of medroxy progestrone was given. The patient showed dramatic improvement in symptoms. At present, she is taking only medroxyprogestrone for last three years (no broncho-dilators and steroids). The patient is showing clinical improvement but no radiological improvement. Fortunately, the patient did not have any episode of pneumothorax since the start of hormone therapy.

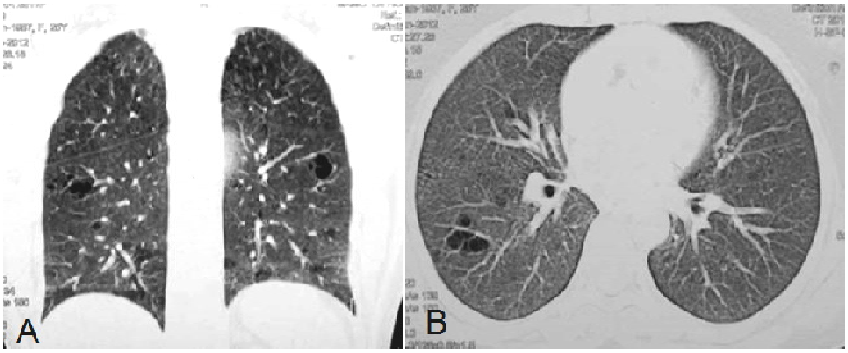

Figure 1.A. Coronal section of chest on CT Chest B. Transverse section of chest on CT Chest: Lung window showing well defined cystic lesions scattered in both lung fields predominantly in middle and lower lobes with ground glass haze

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Discussion

LAM is a rare multisystem disease which exclusively affects females in the reproductive age group. It typically manifests as a lung disease, although kidney and spleen can also be affected. Approximately one third of patients of LAM may have associated renal angiomyolipoma.[2] LAM is of two types: Sporadic LAM (S-LAM) and Tuberous Sclerosis Complex associated LAM (TSC-LAM) with later constituting approximately 15% of them.[3]

LAM is a rare cystic interstitial lung disease which is characterized by proliferation of abnormal smooth muscle cells in the lungs and cystic destruction of lungs. Patients of LAM can present with shortness of breath, chest pain, cough and hemoptysis. Breathlessness is the most common presenting symptom symptom seen in 75% of cases of LAM followed by spontaneous pneumothorax seen in almost half of the cases.[4] The classic presentation of LAM is with pneumothorax or chylothorax. Approximately 60-70% of patients of LAM have pneumothoraces with the highest rate of recurrence (70%) amongst all the chronic lung diseases.[5] It is one of the important causes of recurrent pneumothorax. Our present case is most probably of S-LAM presenting with breathlessness.

The diagnosis of LAM is based on clinical features and findings on computer tomography (CT) of chest. Although plain X-ray chest can reveal cysts and bulla, high resolution CT scan of chest is very helpful in making diagnosis of LAM as the type and pattern of distribution of these cysts can differentiate it among the other possibilities. Lung biopsy is rarely required. Pulmonary function declines rapidly with the progression of the disease because of cystic destruction of lung parenchyma. The most common pulmonary function defects found in LAM are airflow obstruction seen in 60% of cases and decreased lung diffusion capacity.[6] Other defects being mixed obstructive and restrictive.

The management of LAM includes supportive therapy including oxygen, bronchodilators, pulmonary rehabilitation and lung transplant in advanced cases. Immunohistochemical studies of resected specimens revealed LAM cells in the lung parenchyma with receptors for estrogen and progesterone.[7]An estrogen -MMP driven process plays a significant role in lung parenchymal destruction. Therapeutic options include medroxy -progesterone, tamoxifen, gonadotropin -releasing hormone (GnRH) agonists and oophorectomy.[8] In our case, we opted for the daily use of medroxyprogestrone and the patient showed dramatic improvement in her symptoms.

Conclusion

LAM is a rare disease exclusively of females of the childbearing age and is often under diagnosed. Early diagnosis of this condition thorough history and CT chest will enable the clinician to start therapy early before permanent damage to the lung has occurred.

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