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CASE REPORT

LYMPHANGIOMYOMATOSIS PRESENTING AS RECURRENT PNEUMOTHORAX IN A FEMALE CHILD: A CASE REPORT

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ABSTRACT: Lymphangiomyomatosis (LAM) is a rare, idiopathic, cystic, progressive disease seen almost exclusively in women of child bearing age which occurs sporadically or in association with tuberous sclerosis. It is clinically characterized by progressive dyspnea, recurrent pneumothorax, hemoptysis and chylous effusions. Due to its unusual and nonspecific symptoms, patients often receive a delayed diagnosis and despite a variety of treatments many patients require lung transplantation. Here we report the case of a 16 years old girl with lymphangiomyomatosis presenting as recurrent pneumothorax.

KEYWORDS: Pulmonary LAM (Lymphangiomyomatosis), Pneumothorax, Cystic lung disease.

INTRODUCTION: Lymphangiomyomatosis is a rare, diffuse and progressive lung disease that affects young women of child bearing age.[1] It is microscopically characterized by non-malignant smooth muscle cell proliferation in the lungs that causes progressive obstruction of small airways and lymphatic vessels resulting in progressive dyspnea, recurrent pneumothorax and hemoptysis.[2] Reported prevalence of LAM in Asians is 0.24 and 0.03 per 1 lakh in Singapore and Korea respectively, however prevalence in India is not known precisely.[3] The prevalence in children is very rare with only few case reports.[4] We are reporting a case of LAM in a 16 year old female child.

CASE REPORT: A 16 years old girl presented to our OPD with history of left sided chest pain and breathlessness since 15 days which was acute in onset and progressive in nature. There was no history of fever, cough, joint pains, paroxysmal nocturnal dyspnea, syncope, palpitations or hemoptysis. There was significant history of hospitalization 1 year back in private hospital when she had similar complaints and was diagnosed as miliary TB on the basis of chest X-ray findings (Fig. 1) and discharged on anti-tubercular drugs. One month later she was again admitted there with left sided pneumothorax with severe respiratory distress for which ICD was inserted and on improvement ATT was continued for 6 months.

This time on admission to our hospital, her vital parameters were stable with hyper-resonant note on percussion and decreased breath sounds on left side of chest. Cardiovascular and central nervous systems were unremarkable. Chest X-ray showed left sided pneumothorax for which needle aspiration was done and CAT II ATT was started considering relapse of TB. In view of persistent pneumothorax even after needle aspiration, HRCT Thorax was done which showed "Moderate left sided pneumothorax, thin wall cystic lesions in both lungs with emphysematous changes, marginally enlarged pretracheal lymph node and B/L axillary lymphnodes" [Fig. 3]. Lung biopsy showed marked thickening of intimal smooth muscle fibre suggestive of lymphangiomyomatosis and hence confirmed the diagnosis. ICD was inserted to drain the pneumothorax, ATT was stopped and she was started on oral progesterone therapy after which she improved.

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DISCUSSION: LAM is a rare, cystic interstitial lung disease that exclusively affects women of child bearing age and is associated with vascular proliferation of smooth muscle cells in the lung and cystic destruction of lungs.[1,5] LAM occurs in 2 main forms: tuberous sclerosis complex associated LAM (TSC-LAM) and sporadic LAM (S-LAM). LAM occurs in 30% cases of TSC, but TSC-LAM constitute only 15% of patients with LAM.[5]

Patients with LAM usually develop progressive dyspnea, recurrent pneumothorax, chylous collections and occasionally hemoptysis.[6] Pneumothoraces occur in approximately 60-70 % of LAM patients with a recurrence rate of 70%, the highest among all chronic lung diseases.[7] Extrapulmonary lymphadenopathy and lymphangioleiomyomas can result in abdominal and pelvic lymphatic obstruction.[8] LAM is often associated with angiomyolipoma in the kidneys.[9]

In LAM a plain X-ray chest usually shows a hyperinflated lung with reticular or reticulonodular pattern, with or without cysts and bullae.[7,9] However the recommended imaging technique is HRCT which characteristically shows multiple thin walled cysts with other features like adenopathy, pleural effusion, pneumothorax, ground glass opacities, pericardial effusion and dilatation of the thoracic duct.[10] Abdominal CT scan may be used to detect angiomyolipomas, lymphangioleiomyomas or lymphadenopathy which may be found in upto two-third of patients.[8]

Diagnosis is confirmed by lung biopsy (Open or thoracoscopic) which shows cysts and a multifocal nodular proliferation of immature smooth muscle and perivascular epitheloid cells.[11] Immunohistochemistry for HMB-45 and smooth muscle actin and desmin is an important adjunct to diagnosis.[12] However in presence of suitable clinical situation like recurrent pneumothorax, chylothorax or known tuberous sclerosis HRCT is usually enough to clinch the diagnosis.[5]

There is no standard treatment for LAM established so far. Thoracostomy is recommended for pneumothorax. According to latest evidence even the first episode of pneumothorax in a case of LAM should be treated with pleurodesis to avoid recurrence.[13] Since increased incidence of LAM is associated with oral contraceptives and pregnancy, estrogen is implicated in its pathogenesis. Hence progesterone, oophorectomy and tamoxifen have been tried in its treatment.[13,14,15] Other tried drugs include doxycycline, Rho inhibitors and tyrosine kinase inhibitors.[16] Sirolimus, a m-TOR inhibitor is being tried in LAM, however carries the risk of causing severe pulmonary toxicity.[17] Lung transplantation in patients of LAM is usually considered in case of recurrent refractory pneumothorax, severe hypoxemia or declining lung function (FEV1 less than 30% of predicted).[18]

LAM is a very rare disease and there are still many questions unanswered. However in a case of spontaneous recurrent pneumothorax which is confusing for the primary care physician, LAM should be considered, especially in young females. Also, early referral to a pulmonologist is important and essential.

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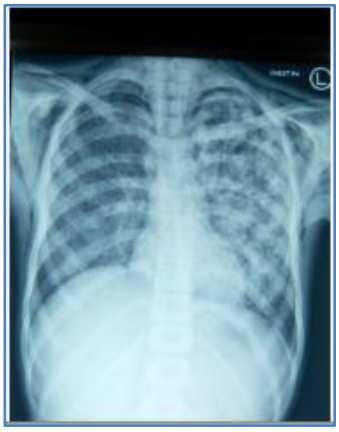

Fig. 1: CXR showing milliary TB like feature-min

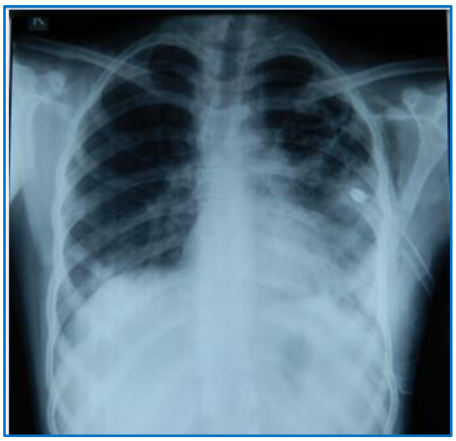

Fig. 2: Presenting with left sided pneumothorax with ICD-min

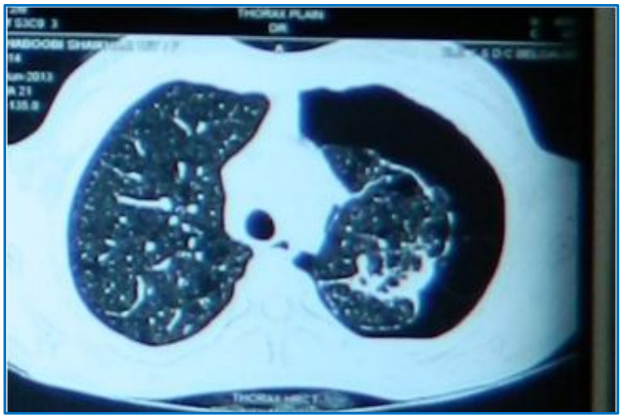

Fig. 3: HRCT showing Moderate left sided pneumothorax, thin wall cystic lesions in both lungs with emphysematous changes

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