Case Report

A rare case of lymphangioliomyomatosis with recurrent pneumothoracis

Ashish Deshmukh[1], Saumya Ramachandran[2*], Sunil Jadhav[3], Shivprasad Kasat[4], Hafiz Deshmukh[5], Chintan Yadav[6]

[1]Professor and HOD, [2,6]Resident, [3]Associate Professor, [4,5]Assistant Professor, Department of Pulmonary Medicine, MGM Medical College and Research Centre, N-6 CIDCO, Aurangabad, Maharashtra, INDIA.
Email: leosaumya21@gmail.com

Abstract

We are reporting a rare and an interesting case of lymphangioliomyomatosis in a middle aged woman with past history of bilateral pneumothacis, who presented with complaints of right sided chest pain and suden onset of breathlessness. Chest Xray was done which was suggestive of right sided pnemothorax. Her HRCT-chest showed multiple parenchymal cyst and was evaluated further with bronchoscopy. Transbronchial lung biopsy was taken which reported as lymphangioliomyomatosis. She was treated with Intercostal drain and oral progesterone supplements.
Key Word: lymphangioliomyomatosis, recurrent pneumothoracis

[*]**Address for Correspondence:**
Dr. Saumya Ramachandran, Resident, Department of Pulmonary Medicine, MGM Medical College and Research Centre, N-6 CIDCO, Aurangabad, Maharashtra, INDIA.
Email: leosaumya21@gmail.com
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INTRODUCTION

Lymphangioleiomyomatosis (LAM) is a rare, progressive, cystic lung disease that occurs almost exclusively in females, usually between menarche and menopause. The hallmarks of LAM are diffuse infiltration of the pulmonary parenchyma with atypical smooth muscle cells (LAM cells) which leads to airflow obstruction, pneumothorax, chylothorax, and progressive respiratory failure. LAM is often initially misdiagnosed as asthma or chronic obstructive pulmonary disease.

CASE REPORT

A 44yr old female before presenting to our department, admitted at a private centre with complaints of sudden onset right sided chest pain and breathlessness. She was diagnosed as having right sided pneumothorax on the basis of chest X ray frontal view for which intercostal drain was inserted and was referred to our hospital for further evaluation. She gives history of exertional dyspnoea since 5yrs, not on any medication and also underwent hysterectomy 4yrs back for menorrhagia, bilateral intercostals drains were placed 5 months back for bilateral pneumothoracis, she improved symptomatically and radiologically following which chest tubes were removed. Presently, her vitals were normal and oxygen saturation was 93% at room air. She was thouroughly investigated at our centre with haemogram, liver function tests, kidney function test which were all within normal limits. HRCT-chest (fig.3) was done which was reported as multiple variable sized lung parenchymal cyst diffusely in both lungs with thin and impeceptible wall at places. Calcified nodular opacity in superior ligular segment in view of multiple lung cysts possibility of lymphangioliomyomatosis needs consideration. Her bronchoscopy was performed which showed normal tracheobronchial tree and her transbronchial lung biopsy was taken. Histopathology report of biopsy showed cystic structures and infilteration of pulmonary parenchyma, airways, lymphatics with smooth muscle cells suggestive od lymphangiomyomatosis. With view of the above

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mentioned investigations patient was diagnosed to have lymphangioliomyomatosis. Oral Tab. Deviry 10mg OD (Methlprednisolone)was started along with oral and inhaled bronchodialators. Intercostal drain was removed following lung expansion. At time of discharge she was advised to continue oral, inhaled bronchodialators and oral medroxyprogestrone. She was also adviced regarding lung transplantation but she refused due to financial crisis. She was symptomatically much better on follow up visits.

CHEST X RAY

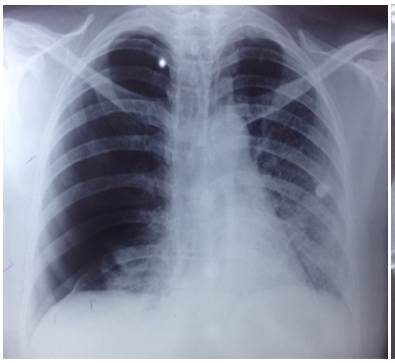

Figure 1: Pre ICD

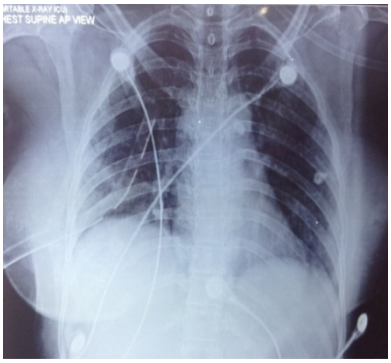

Figure 2: Post ICD

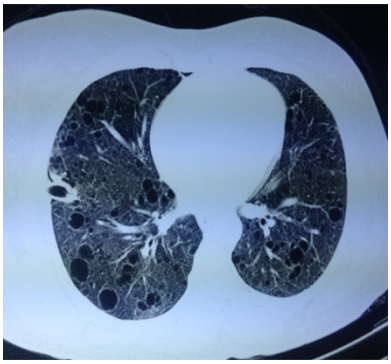

Figure 3: HRCT chest (3/1/2014)

DISCUSSION

Lymphangioleiomyomatosis (LAM) is a rare, progressive, cystic lung disease that occurs almost exclusively in females, usually between menarche and menopause. diffuse infiltration of the pulmonary parenchyma with atypical smooth muscle cells (LAM cells) leading to the formation of lung cysts and fluid-filled cystic structures (ie, lymphangioleiomyomas) in the axial lymphatics causing airflow obstruction, pneumothorax, chylothorax, and progressive respiratory failure.

Pathophysiology

Proliferation of lymphangioleiomyomatosis (LAM) cells obstruct the bronchioles leading to airflow obstruction[1], air trapping, formation of bullae, and pneumothoraces. Obstruction of lymphatics by abnormal LAM cells results in lymphangioleiomyomas, chylothorax, and chylous ascites. Obstruction of venules leads to hemosiderosis and hemoptysis. Excessive proteolytic activity, which relates to an imbalance of the elastase/alpha1-antitrypsin system or metalloprotease (MMPs) and their inhibitors (tissue inhibitors of metalloproteases TIMPs), may be important in lung destruction and formation of cysts[2]. Animal models suggest that estrogen may promote the metastasis of TSC2-deficient cells via lymphatics.[3]

Clinical feature

- Symptoms: chronic cough, progressive breathlessness
- The diagnosis of LAM can be delayed for 3 to 5 years after symptom onset because it is often confused with more common lung diseases, such as asthma or chronic obstructive pulmonary disease.[4-6]
- The most common presentations of LAM include progressive dyspnea on exertion, chylothorax, and pneumothorax in young and middle-aged women.[7]
- Pneumothorax and chylothorax are often the sentinel events that trigger the ordering of computed tomography (CT) of the chest, resulting in the diagnosis of LAM.
- About 70% of women with LAM will have pneumothorax, and an equal percentage will have recurrent ipsilateral pneumothorax or contralateral pneumothorax. Patients with LAM have an average of 2 pneumothoraces before the diagnosis is made.
- About 20% to 30% of patients with LAM develop chylothorax, which is usually unilateral.[8]
- Other, less common presentations of include chronic cough, atypical chest pain, chyloptysis, and hemoptysis.
- LAM may be present in approximately 30-40% of patients with tuberous sclerosis complex (TSC).

Diagnosis

1. *Chest xray*: may be normal. Fine reticular or reticulonodular interstitial infiltrate with preserved lung volumes is the most commonly observed abnormality. Pleural effusions or pneumothorax can be present.
2. *CT chest*: Shows diffuse thin-walled cysts - The defining appearance in LAM, adenopathy and

thoracic duct dilatation, rarely pleural effusion and most often pneumothorax. Ground-glass opacities - May be present, perhaps representing alveolar hemorrhage or interstitial disease. Multifocal multinodular pneumocyte hyperplasia (MMPH) - Can be seen in patients with tuberous sclerosus complex (TSC), but the pathology is distinct from LAM and at times pericardial effusion.

3. *Trans bronchial lung biopsy* : Histopathology report shows smooth muscle nodules in walls of cysts airways. Lymphangioleiomyomatosis(LAM) cells react with human melanoma black (HMB)–45, an antibody generated against an extract of melanoma.[9]HMB-45 staining is used for the identification of LAM cells and may help in confirming LAM on TBB.
4. *Pulmonary function testing*: On spirometry, airflow obstruction is the most frequent abnormality; restriction or mixed obstruction and restriction can also be seen.Lung volumes may show an increased ratio of residual volume to total lung capacity.
5. *DLCO* : decreased diffusing capacity for carbon monoxide is the most common abnormality seen, and it is often markedly reduced.[10,11]
6. *ABG*: Hypoxemia at rest, worsening with exercise, is a common finding.[12]
7. *USG / CT abdomen*: to detect angiomyolipoma.

Treatment

1. General advice and interventions like to refrain from smoking, maintain health and a quality life style(10-13)
2. Avoidance of oestrogen including contraceptive pill and hormone replacement therapy.
3. Information for patients concerning air travel and need for oxygen supplementation during flight to prevent hypoxia.(14)
4. Pulmonary rehabilitation.(15)
5. Influenza and pneumococcal vaccines.(16)
6. Assessment and management of osteoporosis.(17)
7. Inhaled bronchodilators provides clinical benefit.18)
8. Hormone replacement therapy with progesterone supplementation given for 12months with clinical evaluation and lung function at 3 monthly intervals. If lung function and symptoms continue to decline at the same rate on progesterone treatment after one year progesterone should be withdrawn.
9. Hormone theray other anti oestrogen interventions like oophorectomy(20-23), oral tamoxifen(24-17) and GnRH agonist.(28-30)
10. Lung transplantation surgery.

Prognosis

Predicting the prognosis of individual patients is different. Histological extent of the disease and some lung variables(31, 32,33) have been found to be predictive at diagnosis. Disease progression may be evaluated by repeating lung function tests at 3-6 monthly intervals during first year following diagnosis then at 3-12 monthly intervals depending on severity.

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