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A RARE CASE REPORT OF PULMONARY LANGERHANS HISTIOCYTOSIS IN A NONSMOKER FEMALE

Pulmonary Medicine

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ABSTRACT

Pulmonary Langerhans cell histiocytosis [PLCH] is a rare disease. From the insidious onset and non specific manifestation, it is difficult to diagnose PLCH. A report describes a case of 30 years old female with PLCH. She presented with left sided chest pain and sudden onset of breathlessness of few hours duration.

General examination revealed that the patient had tachypnea, but she was hemodynamically stable. Chest examination showed decreased breath sound on left side. Chest x ray revealed left sided pneumothorax. Oxygen was given to the patient and left sided chest tube was inserted.

HRCT of the chest revealed multiple bilateral pulmonary cysts. The final histopathological diagnosis was pulmonary Langerhans cell histiocytosis X after lung biopsy. Pleurodesis was performed. Follow up for more than 2 years revealed no recurrence.

KEYWORDS

Pulmonary Langherhans Cell Histiocytosis,pneumothorax,pleurodesis

INTRODUCTION

Cigarette smoking is not only the major cause of COPD and Ca Lung, but also implicated in smoking related ILD [SRILD], sudden PLCH, RBILD, DIP, IPF. LCH is a rare histiocytic disease and PLCH is likely undiagnosed in the general population.

PLCH is a rare interstitial lung disease affecting young adults and is closely related to smoking. Langerhans cells is derived from dendritic cell system and are normally found in lungs, pleura, skin and reticuloendothelial system. The disease is characterized by formation of multiple lung granulomatous nodules that destruct the terminal respiratory bronchioles producing multiple cystic spaces. Bones may be involved in less than 20% of cases with rare involvement of other organs.

The presentation of PLCH is variable and may be asymptomatic in 25% of cases. The most common presenting symptom is non productive cough followed by dyspnea. However, spontaneous pneumothorax may occur due to rupture of the pulmonary cysts. The recurrence rate for the patients with secondary spontaneous pneumothorax in PLCH, may be more than 60%, so interventions to prevent recurrences are recommended after first episode of pneumothorax.

In this case we are presenting a case of young female with left sided recurrent pneumothorax, which was eventually proved to be secondary to PLCH. PLCH in nonsmoker females is a relatively rare presentation.

CASE REPORT

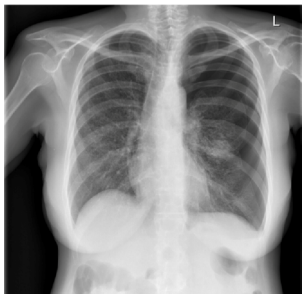
A 30 years old female, presented to ER of our hospital with sudden onset of breathlessness & chest tightness of few hours. She had persistant cough for 2 years, chest distress and fatigue for more than a year. She had breathlessness after activity, but no wheezing, joint pain, chest pain or rash in previous 2 years. She had history of left sided pneumothorax before 6 months, which was resolved after thoracic drainage. The doctor arranged an x ray but not thoracic computed tomography for her when the first pneumothorax occurred. After the first pneumothorax episode, the patient gradually felt the onset of chest distress, dyspnea and fatigue.

She had no past history of DM, hepatitis, tuberculosis, hypertension. She is housewife and not having any addiction. She has no special gas and toxic substance exposure injury. She does not have similar illness in family.

At the time of admission, her temperature was 36.9°C, blood pressure of 108/72 mm of Hg, pulse rate 136/min, RR 24/min, SpO_2 90 % on RA. There was no cyanosis, no distension of jugular vein and no clubbing.

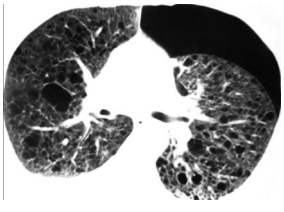
Trachea was shifted to right & there was increase in respiratory movement. On percussion of left side, no hyperresonant sound. On auscultation there was decreased air entry on left side. There was no rhonchi & crackles. There was no lower extremity edema. ABGA, PaO_2 96.7 mm of Hg, $PaCO_2$ 44.2 mm of Hg, SpO_2 95.7%, HCO_3 24.9. Liver and renal function tests were normal. Blood levels of glucose, calcium, total protein, albumin & globulin, WBC, Hb & PH were normal.

CXR PA s/o left sided pneumothorax.



O2 was delivered to the patient through a mask with a rate of 5 L/hrs. Intercostal tube was inserted on left side.

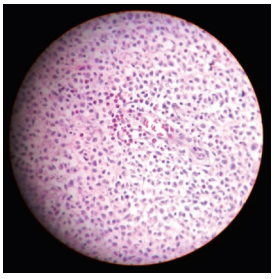
After confirming left sided pneumothorax, ICD was inserted on left side. HRCT thorax showed bilateral diffuse cyst especially upper lobe and middle lobe. ANA and ANCA were negative. To diagnose the patient definitively, we conducted bronchoscopy.



Bronchoscopy did not find neoplasms or any other abnormal lesion except bilateral mucus congestion. During bronchoscopy, we collected BAL and analysed the differencial count. BAL showed macrophages 23%, neutrophils 57%, lymphocytes 11% and eosinophils 9%.

The patient was consented for video – Assisted thoracoscopic surgery (VATS) for biopsy to assist in making a diagnosis.

Histopathology of the biopsy showed infected cell infiltrating in left UL & fiber vessel proliferation. The specimen showed section of lung tissue with honey comb appearance.



Microscopic pathology showed alveolar atrophy, island like fibroplasia and inflammatory cell infiltration. Immunohistochemistry (IHC) report was s-110(+) CD 1a(+) SMA(+) and definitive diagnosis of pulmonary Langerhans cells histiocytosis X was established. A week later pleurodesis on left side was done with povidone iodine. She was discharged after 7 days. She was on follow up for more than 2 years. She revealed no recurrence and both lungs were fully inflated with no difference between both sides.

DISCUSSION

Pulmonary Langerhans cell histiocytosis X (PLCH) is a rare interstitial lung disease characterized by formation of multiple lung cysts with rare involvement of other organs. A similar disease occurs in children, with multiple organ involvement known as peadiatric histiocytic disorder. The PLCH is also known as eosinophilic granuloma and it affects young adults with peak incidence around 20-40 years and males are more affected than females. In our case report the patient was 30 years old female which is rare presentation.

The actual prevalence of PLCH is 0.7-2.7 per million population.

The disease is related to tobacco smoking and starts as peribronchiolar granulomatous infiltrates especially in the upper and middle lung zones (sites of smoking-related lung disease) but in our case report smoking is not the cause. However genetic, environmental, radiation and chemotherapy are also considered as susceptible factors.

The PLCH granulomata are formed of activated Langerhans cells in addition to eosinophils, macrophages, lymphocytes, plasma cells and fibroblasts. The same findings were detected in current case.

The granulomata destructs the terminal respiratory bronchioles and produce stellate scars ending in formation of honeycombing and cystic spaces.

Presentation of PLCH is variable and the most common presenting symptom is non-productive cough followed by dyspnea, easy fatiguability, loss of weight, fever, chest pain, painful bone cysts which may lead to pathological fractures and about one quarter of patient are diagnosed incidentally during chest radiography.

Rupture of the pulmonary cysts will lead to spontaneous pneumothorax which is a recognizable presentation. In our case patient present with recurrent left sided pneumothorax.

There are no specific physical findings and are mostly related to the underlying presentation.

The PLCH must be differentiated from COPD, cystic fibrosis, idiopathic or interstitial pulmonary fibrosis, emphysema and sarcoidosis.

Laboratory findings are non specific and pulmonary functions may be normal but in advanced disease it will show restrictive, obstructive or mixed abnormalities.

The characteristic chest X ray findings include bilateral symmetrical ill-defined nodular lesion with ill defined outline which are mainly found in the upper or mid pulmonary zone with sparing of the costophrenic angles. The presence of cystic lesion of variable size and wall thickness on honeycomb appearance indicate advanced disease. HRCT may be diagnostic and the following findings are pathognomic: upper and mid zonal nodules which are variable in size and may be cavitatory in addition to presence of cysts of various wall thickness and diameter. Consistent findings were detected in the current patient, so pulmonary histiocytosis X is rare but the first differential diagnosis.

The standard diagnostic modality is bronchoscopy or open lung biopsy and its sensitivity and specificity are better than BAL and TBB. Langerhans cells are positive for s-100, CD 1a, moreover, electron microscopy examination of the suspected PLCH lesion would show characteristic increased number of intra cytoplasmic Birbeck granules. In our case the examination of the resected lung tissue taken by VATS confirmed the presence of typical Langerhans cells which was positive for s-100 in addiction to the presence of other granulomatous cells.

As PLCH is rare disease designed therapeutic protocols based on randomized prospective studies are lacking. However abstinence of smoking is the mainstay in the management of PLCH. Other therapeutic measures include control of infection, bronchodilators, if obstructive ventilatory defect is found, in addition to oxygen supplementation if there is significant hypoxemia.

Corticosteroid may be valuable in progressive disease and persistent symptoms. However lung transplantation may be an option in selected patient with advanced diseases.

If spontaneous pneumothorax is developed, insertion of chest tube is indicated and prevention of recurrence, which is common by talc insufflation, parietal pleurectomy and mechanical pleurodesis is recommended by most of the experts. In our case, pleurodesis is done with povidone iodine.

The course of the disease is unpredictable. Certain factors such as recurrent pneumothorax, severe pulmonary hypertension, multiorgan involvement other than bones, have poor prognosis. However most of the studies showed that the median survival is about 12-13 years after presentation.

In our case, patient followed up for more than 2 years without recurrence.

CONCLUSION:

PLCH is a rare disease and may present by spontaneous pneumothorax so high index of suspicion is required in smoker young male to diagnose the condition and to perform pleurodesis for prevention of recurrence of pneumothorax.

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