

PULMONARY LANGERHANS CELL HISTIOCYTOSIS PRESENTING AS SIMULTANEOUS BILATERAL SPONTANEOUS PNEUMOTHORAX IN A NON-SMOKER PATIENT

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Abstract- Pulmonary Langerhans Cell Histiocytosis (PLCH) is a rare idiopathic disorder that primarily affects young adult cigarette smokers. Affected patients often present with cough and dyspnea and about 20% of patients present with or later develop pneumothorax. It is striking that more than 90% of patients are smokers. We report a very unusual case of PLCH in a 20-year-old male patient with no smoking history in whom a life-threatening complication such as simultaneous bilateral pneumothorax was the presenting feature. The final diagnosis was made by open surgical biopsy and recurrent pneumothoraces necessitated surgical management with pleurodesis. We emphasize the early use of pleurodesis in managing patients with PLCH and spontaneous pneumothorax.

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INTRODUCTION

Pulmonary Langerhans cell histiocytosis is an uncommon but important cause of interstitial lung disease which occurs predominantly in adult cigarette smokers. Pulmonary Langerhans cell histiocytosis belongs to the spectrum of Langerhans cell histiocytosis diseases characterized by uncontrolled proliferation and infiltration of various organs by Langerhans' cells (1).

Spontaneous pneumothorax is a recognized feature of pulmonary Langerhans cell histiocytosis and can be the initial manifestation of the disease in about 15% of patients (2, 3). In addition, pneumothorax occurs in up to 25% of patients over the course of their disease and may be recurrent or bilateral (4).

Simultaneous bilateral spontaneous pneumothorax in a young non-smoker is quite unusual in pulmonary Langerhans cell histiocytosis and may even lead to the death of the patient. We strongly recommend the consideration of pulmonary Langerhans cell histiocytosis in the differential diagnosis of simultaneous bilateral spontaneous pneumothorax and an early effective pleurodesis for management of this presentation.

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CASE REPORT

A 20-year-old male patient with sudden onset of worsening dyspnea and previous history of slight cough, dyspnea and weight loss was admitted to our hospital. Vital signs at presentation include: PR=120/min, RR=30/min, BP=110/80 mmHg and T=37° C. The patient was a soldier with no underlying disease and physical examination revealed no significant finding except bilateral diminished breath sounds. Chest radiography showed bilateral pneumothorax and honeycombing of the lungs (Fig. 1). Laboratory tests were normal.

Following insertion of bilateral chest tubes and stabilization of the patient, fiberoptic bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial lung biopsy (TBLB) was performed which later proved to be non-diagnostic.

A chest CT scan (Fig. 2) showed bilateral cystic lung disease and honeycombing which following an open lung biopsy revealed pulmonary Langehans cell histiosytosis as the definitive diagnosis (Fig. 3).

The patient was evaluated for cystic lesions of the skull (Fig. 4) and pelvis and also for diabetes insipidus with negative results for the latter. Follow-up of the patient for whom corticosteroids had been administered, revealed recurrent pneumothorax which was treated with pleurodesis. Since one year ago, the patient has been stable and is advised to undergo lung transplantation.

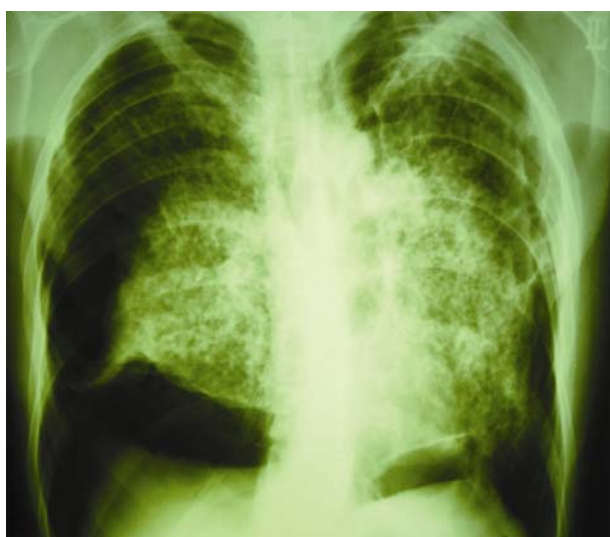


Fig. 1. Chest X-Ray shows spontaneous bilateral pneumothorax.

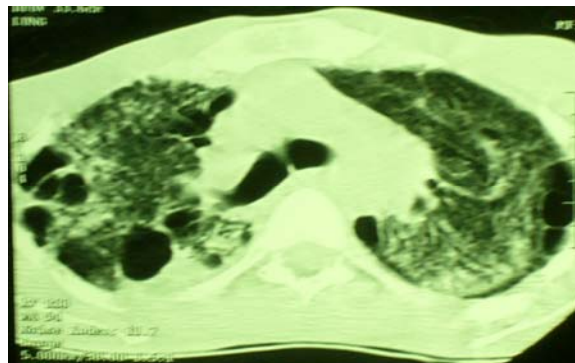


Fig. 2. Chest CT scan reveals bilateral cystic lung disease and honeycombing.

DISCUSSION

The most common conditions underlying secondary spontaneous pneumothorax, which can be the initial manifestation of pulmonary Langerhans cell histiocytosis, are reported to be COPD and *Pneumocystis Carinii* infection associated with HIV infection (5). However, simultaneous bilateral spontaneous pneumothorax is a very rare condition mainly seen in patients with underlying lung disease such as pulmonary metastases, histiocytosis X, tuberculosis, pneumonia and COPD (6) and has been reported to be the initial presenting feature of pulmonary Langerhans cell histiocytosis, even in very young children (7).

Pulmonary Langerhans cell histiocytosis is an isolated form of Langerhans cell histiocytosis that primarily affects cigarette smokers and its diagnosis is made in about 3.4% of individuals undergoing open lung biopsy for chronic diffuse infiltrative lung disease (8).

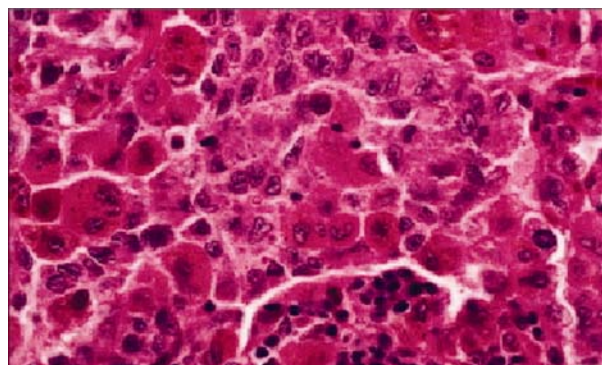


Fig. 3. Pathological diagnosis of pulmonary Langerhans cell histiocytosis.



Fig. 4. Skull X-ray shows cystic lesions.

Most patients with pulmonary Langerhans cell histiocytosis are symptomatic and the most frequent presenting complaints include non-productive cough and dyspnea. Fatigue, weight loss, chest pain that is frequently pleuritic, fever, hemoptysis and pneumothorax as the initial sign in 15% of cases comprise other less common manifestations (9).

The physical examination is notably unremarkable and surgical biopsy is often required for a correct diagnosis (10).

Between 4% and 20% of patients with pulmonary Langerhans cell histiocytosis display cystic bone lesions and a number of case reports demonstrate involvement of other sites including mediastinal lymph nodes, pituitary gland, skin, gut, heart and brain (11). Pulmonary Langerhans cell histiocytosis has been described in association with malignant neoplasia such as bronchogenic carcinoma, Hodgkin lymphoma (12) and lingual carcinoma (13). The course of pulmonary Langerhans cell histiocytosis in adults is variable and unpredictable, ranging from asymptomatic stability to progressive relentless disease leading to respiratory failure or death over a period of months (14) and even sudden death (15).

Regarding the life-threatening nature of bilateral spontaneous pneumothorax and a high recurrence rate of pneumothorax in patients with pulmonary Langerhans cell histiocytosis, we strongly support the early use of pleurodesis in managing patients with pulmonary Langerhans cell histiocytosis and spontaneous pneumothorax. Finally, pulmonary Langerhans cell histiocytosis should be considered in the differential diagnosis of bilateral spontaneous pneumothorax even in a non-smoker patient.

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Pulmonary langerhans cell histiocytosis

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