

Case Report

Pulmonary langerhans cell histiocytosis - Case report

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Abstract

Pulmonary Langerhans Cell Histiocytosis (PLCH) is an uncommon interstitial lung disease seen in smokers, it can often be misdiagnosed as other pulmonary disease. Here we describe a 27 year old male smoker who presented to us with history of fever, cough and breathlessness of 3 months duration. Patient was diagnosed as sputum negative pulmonary tuberculosis and he was put on antitubercular therapy 15 days prior hospitalization. Patient developed bilateral pneumothorax on day 2 of admission. Subsequent further evaluation with x-ray chest showed bilateral diffuse reticulocystic changes and pneumothorax. HRCT chest showed bilateral pneumothorax, bilateral nodular thin walled cysts of varying size in upper lobes classically suggestive of PLCH. Patient underwent intercostal tube insertion and the pneumothorax resolved in 15 days. Later on pleurodesis with tetracycline was done. Patient was discharged with advice of abstinence from smoking and supportive medication.

Keywords: Pulmonary Langerhans Cell Histiocytosis, Interstitial lung disease, Pneumothorax, Reticulonodular Infiltrates, Nodular Cysts

1. Introduction

Pulmonary Langerhans Cell Histiocytosis (PLCH) is an uncommon interstitial lung disease seen in adult smokers¹. PLCH belongs to the spectrum of Langerhans Cell Histiocytosis with varying severity of clinical presentation². Exact incidence and prevalence are not known. However, a large surgical lung biopsy series of patients with interstitial lung disease identified PLCH in 5% of specimens. This rare smoking related, diffuse lung disease affects men between the ages of 20 and 40 years. Clinical presentation varies from asymptomatic to rapidly progressive disease. Most common clinical features are cough, chest pain, weight loss and fever^{1, 2, 3}. Pneumothorax occurs in 25% of patients. The radiographic features include a combination of ill-defined reticular or nodular opacities, bizarre shaped upper zone cysts sparing the costophrenic angle⁵. HRCT that reveals combination of nodules and thin walled cysts is virtually diagnostic of PLCH obviating need for histopathological studies.

2. Case Report

A 27 years old male smoker presented with h/o fever, cough, breathlessness of 3 months duration to the casualty. He had been taking antitubercular treatment since the past 15 days but without any improvement.

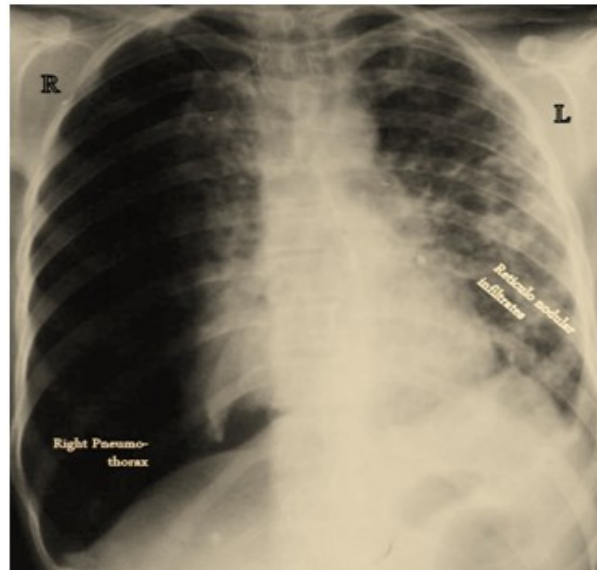
On General physical examination the patient was found to have asthenic build, pallor, Respiratory rate 32 cycles/min, pulse-110 per min, BP-100/70 mm Hg, No cyanosis, no lymphadenopathy. Systemic examination was normal.

2.1 Investigations and Course in Hospital

On further evaluation blood routine revealed Hb- 10gm/dl, TC-10,100/cu.mm, DC-N 70%,L 21%, M 6%, E 2%, B1%, ESR-20mm/hr, RBS- 100mg/dL, Blood urea 27mg/dl, creatinine 1.1mg/dl, LFT- Plasmaprotein-4.5gm/dl, Total albumin 2 mg/dL, AST 24IU/L, ALT-18IU/L. Electrocardiography revealed sinus tachycardia. 2D ECHO was normal. Chest X-ray at the time of admission showed bilateral reticulonodular infiltrates and cystic changes in both upper and lower zones.

On the second day of admission patient developed signs of pneumothorax (right side) and repeated chest x-rays revealed bilateral pneumothorax (more on right side) with mediastinal shift to left (Fig 1).

Fig 1:X-Ray taken on second day shows features of pneumothorax in the right side and reticulonodular infiltrates in left lung field.

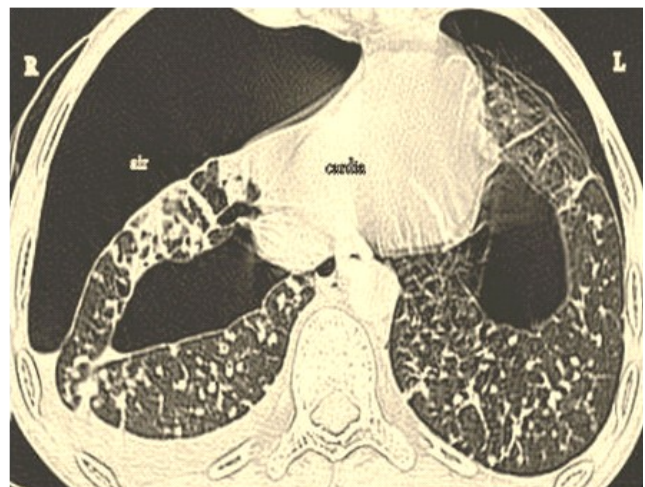


Patient underwent intercostal drainage on right side and he improved symptomatically. HRCT chest done subsequently showed bilateral nodules of varying size, thin walled cysts in both upper lobes with sparing of costophrenic angle, classical of PLCH (Fig:2,3).

Fig 2.HRCT of chest showing multiple thin walled cysts and nodules of varying size in both upper lobes, and bilateral pneumothorax with ICD *in situ*.

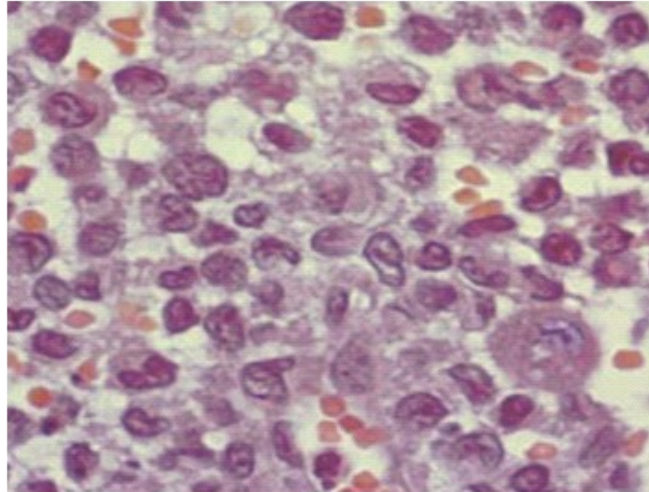


Fig 3.HRCT chest showing absence of cysts and nodules in basal lungs and costophrenic angle.



Histopathological staining of the surgical lung biopsy from the lesion showed the presence of histiocytes and few macrophages, thus confirming the diagnosis of Langerhans cell histiocytosis (Fig 4).

Fig 4. Biopsy of lung showing histiocytes at the centre with inflammatory infiltrate of macrophage at right lower position.



Antitubercular drugs were stopped after hospitalization. Patient was given inj. Ceftriaxone 1 gm bd for 10 days, deriphylline 100 mg tid, oxygen inhalation, haematinics and protein supplementation. Patient underwent pleurodesis with tetracycline after the pneumothorax resolved (Fig 5).

Fig 5. Patient after chest tube removal and pleurodesis pneumothorax did not recur later



3. Discussion

Pulmonary Langerhans Cell Histiocytosis (PLCH) is an uncommon but important cause of Interstitial Lung Disease (ILD) and it occurs predominantly in adult smokers. PLCH can often be misdiagnosed as other pulmonary disease¹.

The estimation of the prevalence and incidence of PLCH is difficult. A large surgical series of patient with ILD identified PLCH in 5% of specimens^{1, 4}. The principal epidemiological factor associated with PLCH is smoking². PLCH occurs principally in young adults between the ages of 20 to 40 years.

Pathogenesis of PLCH has been attributed to the accumulation of dendritic cells in lungs¹. It has been hypothesized that smoke-induced secretion of bombesin like peptides in neuroendocrine cells leads to induction of fibroblasts and

modulation of the cell responses. Other components of cigarette smoke such as tobacco glycoprotein and circulating immune complexes to tobacco antigens are also implicated in the pathogenesis of PLCH^{1,2}. Langerhans cell accumulate in tracheo bronchial tree. In PLCH increased production of tumour necrosis factor-alpha, granulocyte-macrophage colony stimulating factor and transforming growth factor beta occurs due to alveolar macrophages and fibroblasts^{2,3}. This production of the cytokines occurs in the vicinity of langerhans cells leading to their expansion s in peribronchiolar regions. Their persistence results in formation of inflammatory-granulomatous lesions composed of Langerhans cells and plasma cells².

Patients with PLCH commonly present with non-specific respiratory symptoms such as cough and exertional dyspnea. Cough (50-68%), dyspnea (30-50%), fever and weight loss(20-30%), chest pain (10%), constitutional symptoms of a varying severity can occur up to one third of patients^{1,3}. Approximately 25% of patients are asymptomatic at the time of presentation or have mild "smoker's cough". Spontaneous pneumothorax is the presenting diagnosis in approximately 10% to 15% patients. The physical examination including auscultation of lungs is frequently normal unless there are complications such as pneumothorax.

3.1 Diagnosis

Although the diagnosis of PLCH requires histological demonstration of typical lesions containing Langerhans' cells in lungs tissue, classical findings of PLCH in HRCT obviates the need for histopathological studies. The treatment for PLCH is only smoking cessation. Radiography is an important tool for diagnostic evaluation of PLCH. Chest x-ray often demonstrates micronodular or reticulonodular infiltrates in a symmetric and bilateral distribution with relative sparing of costopneumatic angles^{1,6}. HRCT of chest may reveal nodules and cystic of varying size that involves upper lobes with relative sparing of these lung bases pathognomonic of PLCH^{2,4,5,6}.

3.2 Treatment

There is no specific treatment for PLCH. Smoking cessation is advised for all individuals as it is known to stabilize the clinical course known to stabilize the clinical course of the disease. In patients who demonstrate progressive decline of lung function corticosteroid therapy is often used¹. Retrospective case series suggests that corticosteroid therapy in PLCH is associated with stabilization of disease and symptomatic improvement.

Various chemotherapeutic agents such as 2-chloro deoxyadenosine, vinblastine, methotrexate, cyclophosphamide, etoposide, and etanavcept have been tried in patients with progressive PLCH unresponsive to steroid therapy^{1,2}.

Pneumothorax, a well-recognized complication of PLCH is managed with chest tube and often with pleurodesis to prevent recurrence. Patients with progressive PLCH with severe respiratory impairment and limited life expectancy should be evaluated for lung transplantation¹.

References

1. R.Vassalo J.H Ryu. Pulmonary Langherhans Cell Histiocytosis: Clin Chest Medicine 2004 25, 561-571.
2. Krishna M. Sundar, Mar V.Gosselin et al. Pulmonary Langherhans Cell Histiocytosis Emerging Concepts In Pathobiology,Radiology and Clinical Evaluation Of Disease: Chest 2003 123 1673-1683.
3. Robert Vassalo, Day H.Ryu,Darrel R. Schroeder et al. Clinical Outcomes Of Pulmonary Langherhans Cell Histiocytosis In Adults : N Engl J Med 2002 vol 346, No 7.
4. Antonella Caminati, SergioHararl. Smoking Related Interstitial Pneumonias and Pulmonary Langherhans Cell Histiocytosis: Proceedingsofthe American Thoracic Society 2006 vol. 3.
5. Cristopher A. Meyer,Daniella M. Seaman et al. Diffuse Cystic Lung Disease at High-Resolution CT; AJR 2011 196.
6. DannyL.Leatherwood,DareleE.Heikamp,RobertE.Emerson. Pulmonary Langerhans Cell Histiocytosis: Radiographics January-February 2007, vol 27,266-268.