Nonproductive Cough as an Early Sign of Pulmonary Langerhans Cell Histiocytosis

Ereni Abuaita OMS III

Michael Habtemariam OMS IV

NEW YORK INSTITUTE OF TECHNOLOGY
College of Osteopathic Medicine
at Arkansas State University

NEA BAPTIST

BACKGROUND

PLCH is a rare interstitial lung disease primarily associated with young adult smokers.

It is characterized by focal collections of CD1a and S100 positive dendritic cells called Langerhans cells which induce activation of inflammatory response and subsequent tissue remodeling. In advanced disease, bronchial and alveolar destruction results in the appearance of scattered irregular cysts and nodularity.

PRESENTATION

A 31 year-old Caucasian female with an 8 pack-year smoking history presented with acute-onset left-side pleuritic chest pain and progressive dyspnea.

Chest x-ray revealed a spontaneous left pneumothorax. (Figure 1) Following chest tube placement, x-ray revealed decreased pneumothorax and visualization of bilateral airspace opacities.

CT without contrast revealed bilateral irregular cysts in the upper lobes with some areas of irregular septal thickening and nodularity. (Figure 2)

Bronchoalveolar lavage and biopsy from the RUL revealed eosinophilic and histiocytic inflammation. Immunohistochemical stains were positive for CD1a, CD86, and S100 confirming the diagnosis of PLCH.

Please scan to read the full paper and get our contact information.

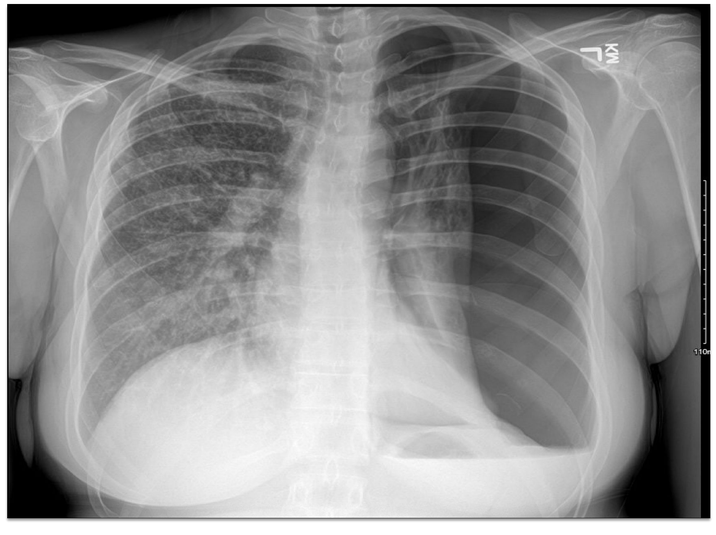


Figure 1. Large left pneumothorax with collapse of much of the left lung and some trace amount of left to right mediastinal shift

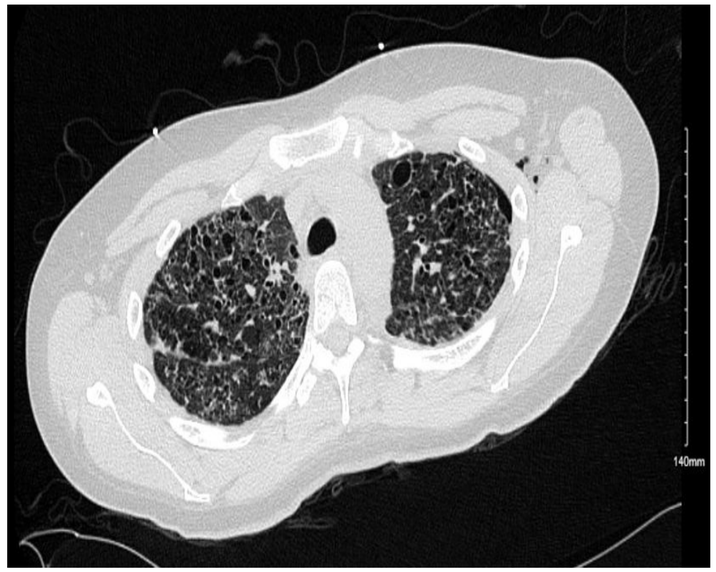


Figure 2. Primarily upper lobe cysts with irregular areas of septal thickening and nodularity bilaterally with atelectasis and small left pneumothorax.

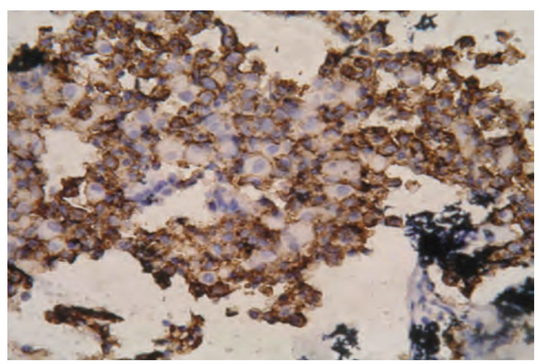


Figure 3. CD 86 positive immunohistochemistry stain

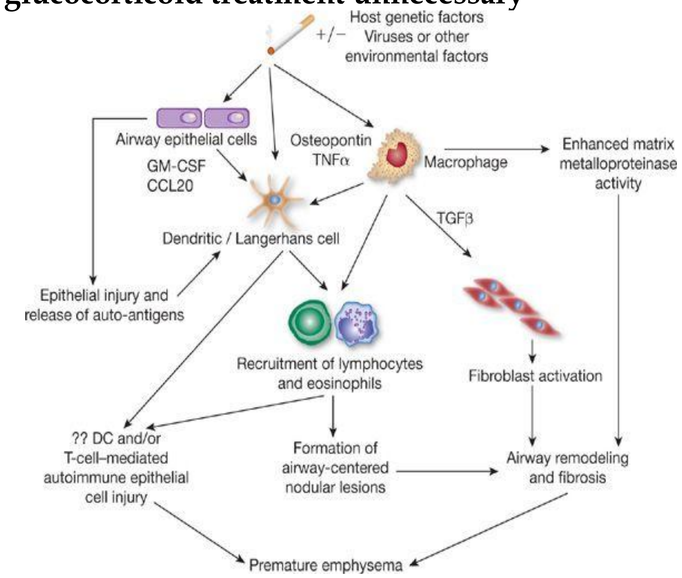
DISCUSSION

Clinical presentation of PLC is often unremarkable and its diagnosis is often delayed until development of acute symptoms such as pneumothorax

If the patient had not presented with pneumothorax, she most likely would have continued receiving inappropriate treatment without improvement

In Young adults with a smoking history, PLCH should be considered earlier in the differential diagnosis to improve patient outcomes

PLCH is associated with a good prognosis upon smoking cessation; often making high-dose glucocorticoid treatment unnecessary



Proposed pathogenesis of PLCH:

Cigarette smoke–induced recruitment and activation of Langerhans cells in small airways

Cigarette smoke triggers cytokine and chemokine production, promoting Langerhans cell recruitment and activation

Chronic inflammation, fibroblast activation, and tissue remodeling contribute to airway-centered fibrosis and cyst formation