



Rare Etiology of Cough and Chest Pain in a Young Male

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ABSTRACT

Chronic cough and chest pain should be dealt seriously in a young adult. Sometimes we may miss a diagnosis in such patients. We present a case of Pulmonary Langerhans Histiocytosis who presented with complaints of cough and chest pain

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Introduction:

Pulmonary langerhans cell histiocytosis is a rare interstitial lung disease commonly seen in young smokers. It may present with acute as well as chronic complaints. However this disease must always be kept as differentials when young adults come with primary spontaneous pneumothorax (1).

Case Report:

A 27-year male with a history of cigarette smoking started experiencing dull aching pain on the right side of the chest since 2 years ago, gradual in onset and intermittent.

The pain was radiating to the back on the right side with no aggravating or relieving factors. He also complained of gradually progressive shortness of breath since 1 year ago, which increased since the last 15-20 days, along with an increase in the intensity of the pain. The patient was taken to a local hospital where the chest X-ray was performed suggestive of bilateral multiple cystic lesions in both lung fields with right-sided pneumothorax (Figure 1).

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Figure 1: A chest-X-ray showing a bilateral multiple cystic lesions in both lung fields with right-sided pneumothorax

On arrival to our hospital, the case was conscious, oriented to time, place, and person with stable vitals. He was advised for tube thoracostomy regarding right-sided pneumothorax; however, he refused again as in the first visit to the outpatient department. He was given symptomatic management for the pain and breathlessness. On the follow-up after 1 week, he experienced an increase in chest pain and shortness of breath. He was admitted, and tube thoracostomy was performed in the right pleural cavity. The patient had symptomatic improvement.

Investigations

The patient's high-resolution computed tomography thorax was suggestive of bilateral cystic lesions (Figure 2). The results of a complete hemogram test showed leucocytosis with total leucocytes counts 12.5 thou/cumm with lymphocytosis. Liver function tests and kidney function tests were normal. Serum angiotensin convertase enzyme levels were normal. The results of the urine routine test and microscopy showed few white blood cells and were positive for bacteria. In addition, cultures were sterile. Sputum evaluation was performed which was negative for acid-fast bacilli (AFB), gram stain, and cultures. Viral serology was negative for hepatitis B surface antigen, HIV, and hepatitis C virus.

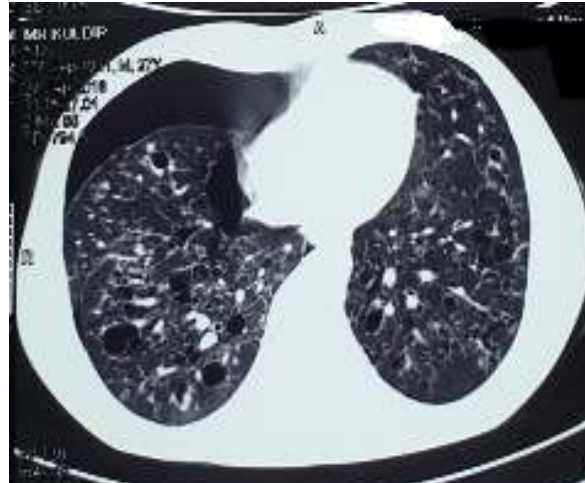


Figure 2: Computed tomography (CT) with a bilateral cystic lesions

Bronchoscopy was conducted, and bronchoalveolar lavage (BAL) was sent for analysis, which was negative for AFB, gram stain, cultures, and fungal culture. The BAL sent for *Pneumocystis carinii* was negative. Transbronchial lung biopsy was performed from bilateral lower lobes, and 4-5 biopsy samples were taken. Serial chest X-rays showed the expanded lung, and after clamping trial, ICD was removed. The patient's lung biopsy showed few hemosiderin-laden macrophages, broken alveolar septa, central nodule comprising sheets of macrophages, and occasional scattered eosinophils (Figure 3). The biopsy sample was sent for immunohistochemistry (IHC) for confirming S100 and CD1a. The IHC was positive for S100. To rule out multisystem involvement, skull and spinal X-rays showed no lesions.

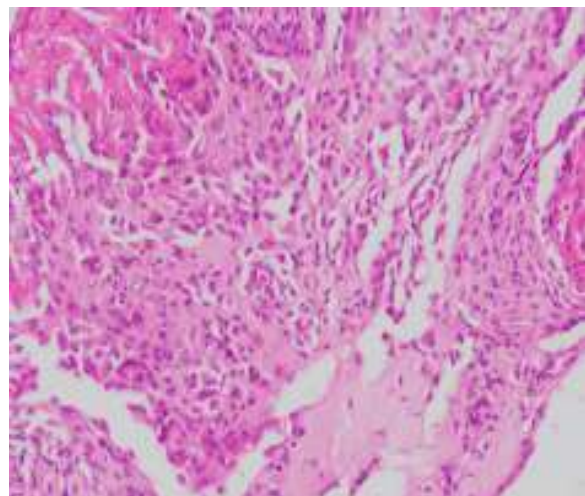


Figure 3: Lung biopsy showed few hemosiderin-laden macrophages, broken alveolar septa, central nodule

Discussion

Previously known as histiocytosis X and later named as Langerhans cell histiocytosis (LCH), this disorder involves multiple systems. Histopathological examination shows characteristic proliferation of histiocytes in various organs. These histiocytes have been identified as Langerhans cells. The Langerhans cells are dendritic cells observed in the epidermis, mucosa, lymph nodes, and bone marrow. According to the literature, two main hypotheses have been accepted one of which suggests the possibility of immune regulation disorder and the other suggests neoplastic proliferation causing the destruction of surrounding soft and hard tissue.

The clinical presentation and presentation age of LCH are used to categorize LCH into three groups, including (1) acute disseminated form with multiple system involvement occurring mainly in neonates known as Letterer-Siwe disease, (2) chronic localized form with solitary or multiple skeletal lesions and occasionally extraskeletal involvement mainly observed in adults known as an eosinophilic granuloma, and (3) chronic disseminated form with osseous lesions, which are frequently multiple with extraskeletal lesions known as Hand-Schuller-Christian disease. A congenital form of LCH presenting with deep subcutaneous lesions is known as Hashimoto-Pritzker syndrome (1).

The case of the present study was pulmonary LCH who presented with unilateral spontaneous pneumothorax, which has been reported to be an initial manifestation in the approximately 15% of patients and is observed more commonly in younger patients (2). Cases of spontaneous bilateral pneumothorax have also been reported (3). There was no skeletal involvement detected in the present case.

The patient was a smoker, and there is a well-established relationship between cigarette smoking and development of pulmonary Langerhans cell histiocytosis (PLCH). Adults developing PLCH have been observed to be active or passive smokers in about >90% of the cases

(4). Travis et al. also observed a significant association of cigarette smoking with PLCH (5). Smoking cessation may lead to the remission of lung lesions, which can be complete or partial (6). Peribronchial inflammatory changes are prominent in PLCH, which is due to the injury to the airways by cigarette smoking. Only a small number of smokers develop PLCH that suggests the possibility of the role of endogenous host factors and second hit by cigarette smoking (7).

The PLCH is categorized under interstitial lung diseases; however, the pathological features are predominantly suggestive of inflammatory and destructive bronchiolitis with granuloma-like nodules around small airways (5). The nodular inflammation and alveolar macrophages infiltration may be accompanied by extensive vascular involvement resulting in vasculopathy in the arteries and veins (8). The PLCH has characteristic cystic lesions due to the peribronchial involvement, which destroy the cellular and connective tissue components of the bronchial walls. This process leads to the formation of parenchymal cystic lesions, which are irregular. There is also associated reaction emphysema of alveoli and peribronchial fibrosis (9).

PLCH have characteristic langerhans cells (LCH cells) which have highly convoluted nuclear membranes, lack prominent nucleoli and have abundant cytoplasm, with poorly-defined border. These cells stain for CD1a and S100. Hence presence of these stains makes the diagnosis likely to be pulmonary langerhanscell histiocytosis.

The immunohistochemical staining for S100 and the CD1a antigen are present on the cell surface support are positive in PLCH. Our patient was negative for CD1a, while he was positive for S100. The combination of histopathological characteristic findings, along with positive immunohistochemical staining is crucial to finalize the diagnosis of PLCH. Furthermore, it is important to rule out other differential diagnoses for smoking-induced diffuse lung diseases, including respiratory bronchiolitis (a component of

which is almost always present in patients with PLCH), desquamative interstitial pneumonia, and eosinophilic pneumonia (7).

Conflicts of Interest

The authors declare that there is no conflict of interest.

References

1. Aruna DR, Pushpalatha G, Galgali S, Prashanthi R. Langerhans cell histiocytosis. *J Indian Soc Periodontol.* 2011; 15:276-9.
2. Mendez JL, Nadrous HF, Vassallo R, Decker PA, Ryu JH. Pneumothorax in pulmonary Langerhans cell histiocytosis. *Chest.* 2004; 125:1028-32.
3. Nakhla H, Jumbelic MI. Sudden death of a patient with pulmonary Langerhans cell histiocytosis. *Arch Pathol Lab Med.* 2005; 129:798-9.
4. Vassallo R, Ryu JH, Schroeder DR, Decker PA, Limper AH. Clinical outcomes of pulmonary langerhans'-cell histiocytosis in adults. *N Engl J Med.* 2002; 346:484-90.
5. Travis WD, Borok Z, Rous JH, Zhang J, Feuerstein I, Ferrans VJ, et al. Pulmonary Langerhans cell granulomatosis (histiocytosis X). A clinicopathologic study of 48 cases. *Am J Surg Pathol.* 1993; 17:971-86.
6. Mogulkoc N, Veral A, Bishop PW, Bayindir U, Pickering CA, Egan JJ. Pulmonary Langerhans' cell histiocytosis: radiologic resolution following smoking cessation. *Chest.* 1999; 115:1452-5.
7. Colby T V, Lombard C. Histiocytosis X in the lung. *Hum Pathol.* 1983; 14:847-56.
8. Fartoukh M, Humbert M, Capron F, Maître S, Parent F, Le Gall C, et al. Severe pulmonary hypertension in histiocytosis X. *Am J Respir Crit Care Med.* 2000; 161:216-23.
9. Kambouchner M, Basset F, Marchal J, Uhl JF, Hance AJ, Soler P. Three-dimensional characterization of pathologic lesions in pulmonary langerhans cell histiocytosis. *Am J Respir Crit Care Med.* 2002; 166:1483-90