Pulmonary Langerhans Cell Histiocytosis Presenting as Miliary Nodules in an HIV-Positive Man

Josebelo Chong, MD; Marvin Balaan, MD
Allegheny General Hospital, Pittsburgh, PA

Abstract

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INTRODUCTION: Pulmonary Langerhans Cell Histiocytosis (PLCH) is a rare interstitial lung disease of unknown etiology resulting from granulomatous infiltration of the bronchial walls by myeloid dendritic cells. There is no known association with HIV. We are presenting the second case of an HIV-positive patient who was diagnosed with PLCH.

CASE PRESENTATION: A 43-year old African-American male with human immunodeficiency virus (HIV) infection and rectal carcinoma was referred for abnormal radiographic findings. A recent CT scan of the thorax done for cancer staging revealed innumerable miliary nodules. He had no respiratory or constitutional symptoms, and was compliant with his medication regimen. He had not smoked cigarettes for over a year. A video-assisted thoracoscopic surgery (VATS) with biopsy was ultimately done. Histologic examination revealed a stellate, variably cellular and fibrotic process consisting of histiocytoid cells with admixed eosinophils and lymphoplasmacytic cells arranged in loose aggregates around the bronchioles and within the interstitium, diagnostic of PLCH. No immunosuppressive therapy was initiated.

DISCUSSION: Pulmonary involvement occurs in only 10% of Langerhans Cell Histiocytosis cases. The clinical presentation is variable, ranging from cough and exertional dyspnea to spontaneous pneumothorax. Up to 25% may be asymptomatic. Chest imaging usually shows reticulomicrocystic nodules separated by normal parenchyma predominating in the middle and upper lung fields. Diagnosis is made by biopsy, which demonstrates the accumulation of activated Langerhans cells into loose granulomas that develop in and destroy the distal bronchiole walls. Many patients with PLCH recover spontaneously or remain stable without specific treatment. Given the strong association between cigarette smoking and PLCH, smoking cessation remains the cornerstone of therapy. There is limited evidence regarding the efficacy of glucocorticoids and cytotoxic agents. Lung transplant may be considered for severe cases, although recurrence in the transplanted lung has been reported, especially if smoking is continued. There is no known association between PLCH and HIV. It has been suggested that HIV infection of immature dendritic cells or effects on cytokine expression may interfere with dendritic cell maturation but there is no evidence to support this.

CONCLUSIONS: To our knowledge, this is only the second report of PLCH in an HIV-positive patient. In contrast to the first, our patient was a current nonsmoker with an atypical appearance on chest imaging, and hence the diagnosis was not immediately suspected. The link between HIV and PLCH - if any - remains a mystery.


DISCLOSURE: The following authors have nothing to disclose: Josebelo Chong, Marvin Balaan
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