Adult Langerhans Cell Histiocytosis Masquerading as Hidradenitis Suppurativa

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Figure 1. Posterior–anterior chest X-ray revealing diffuse reticular and cystic lung changes.

A 32-year-old man with past medical history of central diabetes insipidus (DI) of previously unknown etiology and hidradenitis suppurativa (HS) presented with worsening dyspnea and bilateral chest tightness. He had no history of smoking, marijuana, or drug abuse. Physical examination was significant for decreased breath sounds of the right and left lower lung fields. A radiograph of the chest showed low lung volumes with diffuse reticulation and cystic changes of both lungs (Figure 1). Computerized tomography scan of the chest (Figure 2) revealed diffuse, bilateral, irregular cysts throughout the lungs. No other infiltrates or effusions were present. Pulmonary function testing showed a mixed obstructive and restrictive ventilatory defect with reduced diffusing capacity of the lung for carbon monoxide. The differential given the cystic findings on computerized tomography were: Langerhans cell histiocytosis (LCH), Birt-Hogg-Dubé syndrome, lymphocytic interstitial pneumonia, amyloidosis, and potential cystic metastases. Biopsy of the skin lesion within the right axilla (Figures 3 and 4) was performed, which revealed LCH and not HS. Given these findings, the patient’s DI was investigated with brain magnetic resonance imaging (Figure 5), which found a pituitary microadenoma. The patient’s constellation of symptoms and cystic imaging findings were attributed to multifocal adult LCH, which differs from pulmonary LCH in that it often occurs in nonsmokers, causes diffuse rather than upper and mid–lung field–predominant cystic disease, and involves visceral organs in addition to lymph nodes, bone, skin, and pituitary (1). He was started on cytarabine owing to multiorgan involvement that was unresponsive to corticosteroids. His DI and skin lesion improved in response to chemotherapy, but his lung function continued to deteriorate.

Multifocal LCH is an inflammatory neoplasm, and typical lesions are composed of approximately 8 to 30% BRAF or mitogen-activated protein kinase mutant tumor cells and the stromal cells they recruit (1–5).

Given the recent discovery of targetable driver mutations, some patients may be considered for therapy with BRAF or mitogen-activated protein kinase extracellular signal-regulated kinase inhibitors after appropriate genetic analysis. Lung involvement can occur in isolation or in conjunction with multiorgan disease. In those with multiorgan disease, DI is seen in 30% of adult patients,
whereas rashes and ulcerations occur in 24% of patients (2, 3). Our case is especially unique in that cutaneous LCH rarely mimics HS (6, 7). Like pulmonary LCH, smoking can lead to accelerated lung destruction in multifocal adult LCH, and smoking cessation should be strongly encouraged. Multiorgan disease refractory to corticosteroids can be treated with chemotherapeutic agents such as vinblastine, etoposide, cladribine, or cytarabine. Physicians should consider LCH in their differential diagnosis of young patients with cystic lung disease and be aware of its myriad skin manifestations. An abstract of this work was presented at the American Thoracic Society International Conference in San Francisco in 2016 (8).

**Author disclosures** are available with the text of this article at www.atsjournals.org.

**Figure 2.** Chest computed tomography scan revealing diffuse, bilateral, irregular cystic disease in the axial (A), coronal (B), and sagittal (C) images. Subcutaneous emphysema occurred after chest tube placement for bilateral pneumothoraces.

**Figure 3.** Ulcerating skin lesions with sinus tracts in the axilla.
References


