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CASE IMAGE

Spontaneous pneumothorax with isolated pulmonary Langerhans cell histiocytosis in an adult case: A common manifestation of rare disease

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Key Clinical Message

Pulmonary Langerhans cell histiocytosis should be evaluated even in adults with no previous medical history and no history of cigarette smoking who have spontaneous pneumothorax and evidence of multiple lung cystic lesions, and other organs should also be checked for multi organ Langerhans cell histiocytosis involvement.

Abstract

A 30-year-old man presented with sudden chest pain and evidence of multiple cystic lesions in both upper and lower lobes of lungs, as well as left-sided pneumothorax in high resolution computed tomography. In lung samples, hematoxylin and eosin- stained sections and IHC for CD1a, S100, and BRAF V600 were positive. The patient was diagnosed with isolated pulmonary Langerhans cell histiocytosis and was treated accordingly.

KEYWORDS

cavitary long lesion, pulmonary Langerhans cell histiocytosis, spontaneous pneumothorax

1 | CASE PRESENTATION

A 30-year-old man had sudden chest pain and shortness of breath for 2 h prior to admission. He denies previous medical or family history, and there was no history of medication usage, cigarette smoking, or alcohol consumption.

In the chest physical examination, there were evidences of asymmetrical chest expansion and reduced breath sounds in the left side. During the examination of the scalp, fingers, and nails, no lesions were discovered.

The high-resolution computed tomography (HRCT) scan of the chest revealed widespread broad spread bizarre-shaped cystic formation in both the upper and lower lobes of the lungs, as well as left-sided pneumothorax (Figure 1). A chest tube was placed on the left side of the chest for the patient. The patient underwent upper and lower lung lobes wedge biopsy. Hematoxylin and eosin (H&E)-stained sections and immunohistochemistry (IHC) for CD1a, S100, and BRAF V600E (VE1) were positive in many of the cyst wall cells, confirming pulmonary

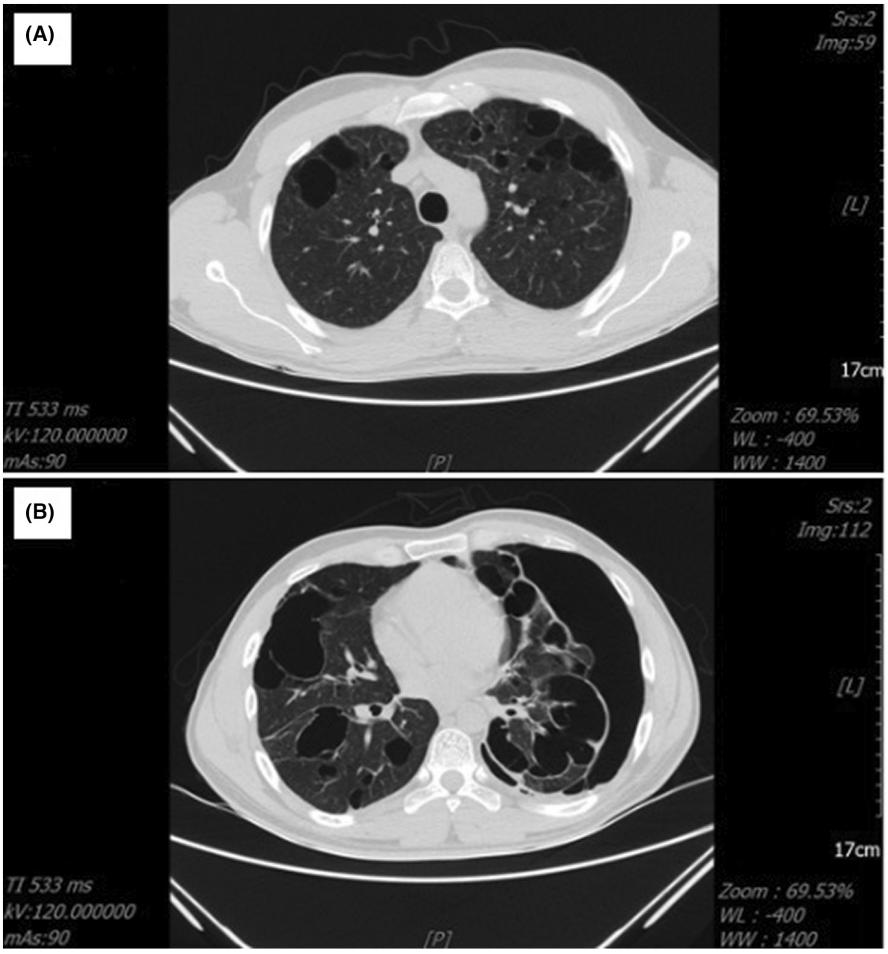

FIGURE 1 Diffuse wide spread bizarre-shaped cystic lesion in both (A) upper and (B) lower lobes of lungs, as well as left-sided pneumothorax.

Langerhans cell histiocytosis (PLCH). The patient was evaluated in terms of the involvement of other organs. The technetium-99m-methylenediphosphonate ([99m] Tc-MDP) whole–body bone scan revealed no abnormalities in bone structures. Dynamic pituitary magnetic resonance imaging (MRI) was performed, and there was no abnormal signal intensity in the pituitary. Ultimately, the patient received treatment for isolated PLCH through the administration of systemic corticosteroids, specifically 0.5 mg/kg/day dose of prednisone, along with supplemental oxygen.

Langerhans cell histiocytosis (LCH) is a clonal disorder of langerhans cells, which may affect various organs, such as bones, skin, pituitary gland, and lungs.[1,2] PLCH is a rare cystic pulmonary disease in adults that occurs almost (more than 95%) in cigarette smokers.[1] Isolated PLCH is seen in around 50% of PLCH patients.[2] PLCH has several manifestations, one of which is spontaneous pneumothorax, which occurs in about 30%–45% of patients and is the first presentation of PLCH in 10%–30% of patients.[2] As a result, PLCH disease should be evaluated even in adults with no previous medical history and no history of cigarette smoking, who have spontaneous pneumothorax and evidence of multiple lung cystic lesions, and other organs should also be checked for multi-organ LCH involvement.

AUTHOR CONTRIBUTIONS

Mitra Samareh Fekri: Supervision. **Faranak Salajegheh:** Data curation; project administration. **Mohsen Nakhaie:** Validation; writing – original draft. **Mohammad Rezaei Zadeh Rukerd:** Visualization; writing – original draft; writing – review and editing.

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FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors declare that there was conflict no of interest.

DATA AVAILABILITY STATEMENT

The data supporting this study's findings are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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