Pulmonary Langerhans cell histiocytosis

Glendy Maricela Orozco Cotí M.D.
Paul Reginaldo Villagrán Ovando M.D.
Brendy Marisol Velásquez Orozco M.D.
Iveth Sarahí Amador Rosa M.D.
José Carlos Echeverría Solis M.D.
Mary Geyovana Cotí Coyoy M.D.

Quetzaltenango, Guatemala

Case Report

Radiology

OPEN ACCESS

Background: Langerhans cell histiocytosis, a rare disease that occurs mainly in children, may present with a broad range of manifestations, from a single osseous lesion to multiple lesions involving more than one organ or system. The clinical course varies widely according to the patient's age. Multisystem disease may show particularly aggressive behavior in very young children, with the outcome largely depending on the disease stage and the degree of organ dysfunction at the time of diagnosis. Extraosseous manifestations are less common than osseous ones and may be more challenging to identify. To accurately detect extraosseous Langerhans cell histiocytosis at an early stage, radiologists must recognize the significance of specific clinical and laboratory findings, as well as the relevance of imaging features for the differential diagnosis. The pattern and severity of pulmonary, thymic, hepatobiliary, splenic, gastrointestinal, neurologic, mucocutaneous, soft-tissue (head and neck), and salivary gland involvement in Langerhans cell histiocytosis are generally well depicted with conventional radiography, ultrasonography, computed tomography, and magnetic resonance imaging. However, imaging features are not pathognomonic, and biopsy is usually required to establish a definitive diagnosis.

Keywords: Langerhans cell histiocytosis.

Pulmonary Langerhans cell histiocytosis is most often identified in children (aged 1–5 years). It is a rare disorder with no well-established gender predilection, appearing to be more common in the white population.

Langerhans cells proliferate in the bronchiolar and bronchial epithelium, forming granulomas. It is hypothesized that, as these cellular granulomas evolve, peripheral fibrosis develops, resulting in traction on the central bronchiole, which then acquires a cystic shape. An immune-mediated mechanism has been proposed, although no triggering agent has been identified.

The earliest change is a diffuse, bilateral, symmetrical reticulonodular pattern with a predilection for the mid and upper lung zones. Later, cyst formation can be observed, or a honeycomb-like appearance may develop due to the confluence of multiple air-filled cysts. Reduced lung volumes are uncommon and are seen only in end-stage fibrotic cases.

Distribution is key in differentiating pulmonary Langerhans cell histiocytosis from other cystic lung diseases, with a predilection for the mid and upper zones and regional preservation of the costophrenic recesses, the anterior segment of the right middle lobe, and the lingula of the left upper lobe.

Case report

A 2-year-old male patient from San Francisco El Alto, Totonicapán. The patient's mother reported that one year earlier, he developed scaly lesions on the scalp, chest, and back, for which he received dermatological treatments with no improvement. Three days prior to admission, he developed respiratory distress, which did not improve despite nebulization and antibiotic therapy. A private physician referred him to our center due to oxygen desaturation.

On admission, the patient was somnolent, febrile, normocephalic, with scaly lesions on the scalp, isochoric pupils with poor light reactivity, dry oral mucosa, mobile and symmetrical neck without lymphadenopathy, symmetrical and expandable thorax, palpable axillary lymphadenopathy, regular but tachycardic heart sounds without murmurs, decreased air entry in both lungs with crackles and wheezes, abdomen soft, depressible, non-tender with normal bowel sounds, no peritoneal irritation signs, no palpable inguinal lymphadenopathy, upper limbs mobile and symmetrical with capillary refill time >3 seconds and non-palpable distal pulses, lower limbs mobile and symmetrical with capillary refill time >3 seconds and non-palpable distal pulses.

From the Radiology Department at Hospital Regional de Occidente, Quetzaltenango, Guatemala. Received on August 21, 2025. Accepted on August 26, 2025. Published on August 28, 2025.

DOI 10.5281/zenodo.16990804

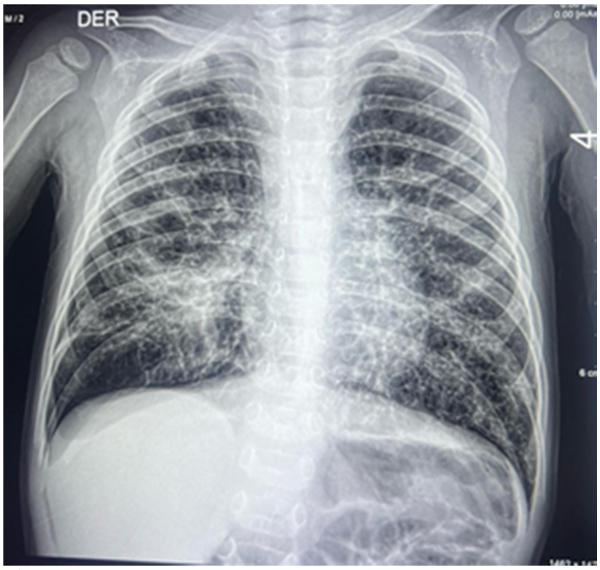

Figure 1. Chest x-rays shows severe generalized cystic lung disease with cysts of varying size and shape, a predominantly right-sided pneumothorax, and small solid nodules.

One month prior to consultation, a biopsy of the scaly lesions was performed, yielding a diagnosis of Langerhans cell histiocytosis. On admission, a chest CT scan revealed a characteristic pattern of pulmonary infiltration.

Discussion

Pulmonary Langerhans cell histiocytosis in adults is a rare disorder of unknown etiology that predominantly affects young smokers, with a peak incidence between 20 and 40 years of age. In adults, pulmonary involvement typically presents as a single-system disease characterized by focal Langerhans cell granulomas that infiltrate and destroy distal bronchioles.

High-resolution computed tomography (HRCT) of the chest is essential for diagnosis, typically showing a combination of nodules, cavitated nodules, and thin- or thick-walled cysts. An increased number of macrophages in bronchoalveolar lavage (BAL) fluid is common but nonspecific, merely reflecting tobacco smoke exposure. BAL is useful for ruling out infections and other infiltrative lung diseases seen in young adults. Langerhans cells can be identified in BAL fluid, but contrary to early expectations, this test has very low sensitivity and is rarely useful for diagnosis.

Definitive diagnosis requires identification of Langerhans cell granulomas, usually achieved through surgical lung biopsy from a site selected via HRCT. However, in clinical practice, lung biopsy is performed on a case-by-case basis.

To date, there is no effective treatment, and there is an urgent need to improve understanding of the mechanisms involved in the pathogenesis of pulmonary Langerhans cell histiocytosis, which should help in developing specific therapeutic strategies for patients with this orphan disease.

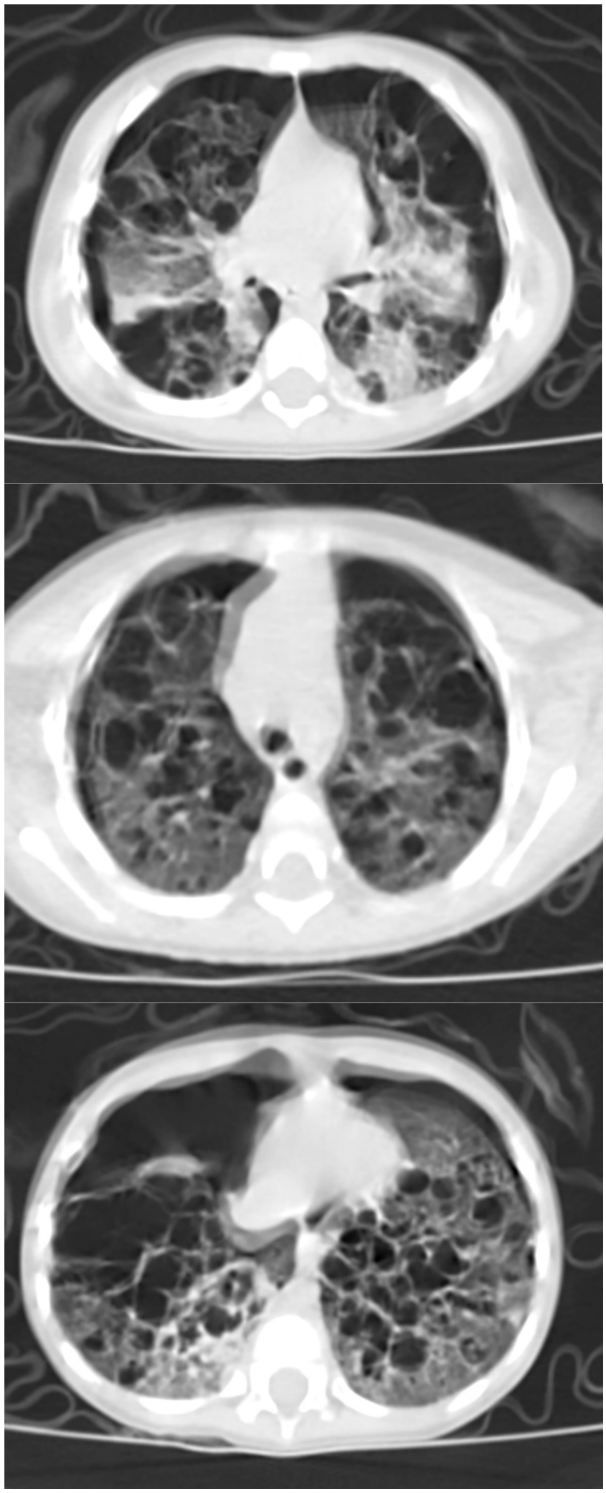

Figure 2. The presence of multiple thin-walled cystic lesions, some confluent, of varying size in both lung fields is observed. These lesions cause loss of lung parenchymal architecture, with bilateral spontaneous pneumothorax, predominantly on the right side.

DOI 10.5281/zenodo.16990804

Conclusion

Although some steps of the pathological process leading to pulmonary Langerhans cell histiocytosis are beginning to be elucidated, further research is needed to explain the various aspects of Langerhans cell granuloma formation. A key objective is to determine the mechanisms involved according to the disease's location and clinical expression. A better understanding of the pathogenesis of Langerhans cell histiocytosis, particularly the role of Langerhans cells, should ultimately lead to the development of rational treatments for this rare disease.

Conflicts of interests

The authors have no conflicts of interests.

Acknowledgements

We thank the Hospital Regional de Occidente for its support of the Radiology Department, providing the necessary supplies and equipment for the prompt diagnosis of patients. Special thanks to Dr. Jose Carlos Echeverria and Dr. Mary Cotí for their assistance in diagnosing this case.

References

1. Maller V, Sharma R, Weerakkody Y, et al. Pulmonary Langerhans cell histiocytosis. Reference article, Radiopaedia.org (Accessed on 07 Aug 2025) https://doi.org/10.53347/rID-5018.
2. Aburto M, Capelastegui A. Pulmonary Langerhans' cell histiocytosis in adults: an unknown old disease. Med Clin (Barc). 2014; 143 (10): 444-445.
3. Acta méd. Grupo Ángeles vol.19 no.3 México jul./sep. 2021 Epub 04-Abr-2022.
4. Eur Respir J 2006; 27: 1272–1285 DOI: 10.1183/09031936.06.00024004

Orozco Cotí Glendy Maricela
Department or Radiology

 DOI 10.5281/zenodo.16990804