# Diffuse cystic lung disease in a child

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A 3-year-old boy presented with fever and cough for 1 month and fast breathing for 20 days. He had no history of decreased appetite, weight loss or tuberculosis contact. There was no history of similar complaints in the past. Examination revealed a thin-built, tachypneic boy with no cyanosis or clubbing and bilateral, scattered, fine crepitations on auscultation. His arterial oxygen saturation was 92% on room air. Chest radiograph [Figure 1] showed bilateral reticular infiltrates with a few doubtful cystic lesions (arrow). Other investigations including complete blood count and tuberculosis work up were non-contributory. High-resolution computed tomography (HRCT) of the chest [Figure 2a and b] revealed bilateral diffuse cystic changes.

## QUESTION

What is the diagnosis?

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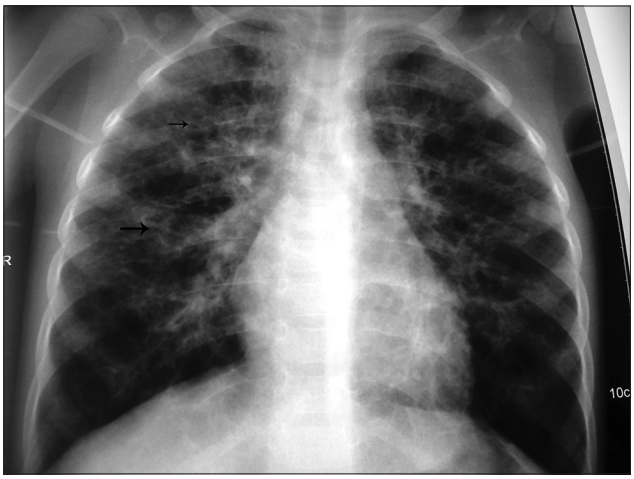

**Figure 1:** Chest X ray showing bilateral reticular infiltrates with few doubtful cystic lesions

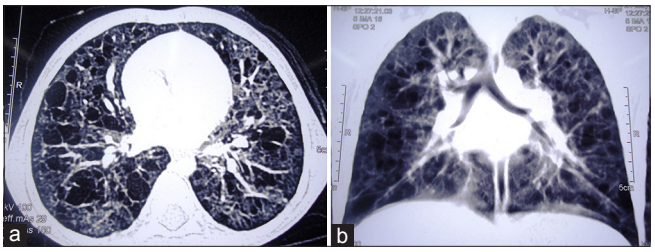

**Figure 2:** (a) HRCT chest showing bilateral numerous cysts of variable size, shape and wall thickness with normal intervening lung parenchyma. (b) HRCT showing bilateral diffuse involvement of lung parenchyma and normal bronchial tree

## ANSWER

Pulmonary Langerhans cell histiocytosis.

Diagnosis was confirmed by lung biopsy showing characteristic features of LCH with immunopositivity for CD1a. There was no evidence of histiocytic involvement of any other organ system.

Histiocytic involvement of lungs, as a part of multisystem disease, occurs in 23-50% of children with Langerhans cell histiocytosis (LCH), with isolated pulmonary LCH being rare in this population.[1] The presence of typical features on high-resolution computed tomography (HRCT) of chest allows the clinician to make a presumptive diagnosis of LCH. Most common HRCT findings of pulmonary LCH are bilateral cystic lesions of variable size (usually less than 20 mm), shape (round/ovoid/bilobed/cloverleaf) and wall thickness. The presence of nodules along with cystic lesions is nearly pathognomonic but is found early in disease course as lesions evolve sequentially from nodules, cavitated nodules and thick-walled cysts to thin-walled cyst and eventually confluent cysts.[2,3] Though, typically upper and middle lung zones are predominantly affected with sparing of lung bases, diffuse involvement has been described in pediatric population (as in the present case).[4-6] Main differential diagnoses of cystic lung disease on HRCT, apart from pulmonary LCH, include lymphangioleiomyomatosis with/without tuberous sclerosis, emphysema and bronchiectasis, which can be differentiated based on combination of clinical and imaging findings. Lymphangioleiomyomatosis and lung disease due to tuberous sclerosis occur in females of child bearing age and on HRCT cysts are small, thin walled, more regular in shape and uniformly distributed. Emphysematous cysts have no visible cyst wall and cysts follow bronchial pattern in cystic bronchiectasis.

The child has been put on treatment as per German-Austrian-Dutch group HX-83 chemotherapy protocol.[7] Follow-up is planned with serial HRCT chest to assess the response to treatment.

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