A 45-year-old female was hospitalized for evaluation of chronic cough and exercise intolerance of 2-months duration. Her past medical history was significant for smoking (20 pack years) and diverticulosis. Initial assessment revealed stable haemodynamic parameters with bilateral wheeze and crackles. Complete blood count and basic metabolic profile were unremarkable. A chest radiograph (Figure 1a) demonstrated bilateral reticular infiltrative pattern in middle lobes predominantly. Arterial blood gases showed pH 7.43, pO₂ 68 mmHg, pCO₂ 38.9 mmHg, HCO₃ 25.5 mEq/l. High-resolution computed tomography (HRCT) of chest (Figure 1b) exhibited bilateral nodular cystic lesions distributed in upper and middle lung zones. A restrictive pattern was noted on pulmonary function tests. An open lung biopsy was performed that showed infiltrates composed of Langerhans cells (indistinct cell borders with grooved nuclei) (Figure 1c, arrows), lymphocytes and eosinophils (arrow head). Immunostains were positive for cytoplasmic S-100 protein (Figure 1d). Diagnosis of pulmonary
Langerhans cell histiocytosis (PLCH) was established. She was commenced on steroids and advised to quit smoking. On subsequent follow-up after 6 months, she had improved symptomatically.

Isolated PLCH is a rare pulmonary disorder that primarily affects cigarette smokers. The pathogenesis of PLCH remains unknown. An uncontrolled immune response to an exogenous antigen in which Langerhans cell serve as accessory cells in the activation of T lymphocyte has been suggested.¹

The characteristic radiological (chest X-ray and CT) features are bilateral reticular and reticulonodular and cystic lesions that predominantly involve the upper and middle lung zones. There is no definite defined therapy for PLCH. Smoking cessation and corticosteroids have been advocated. Lung transplantation may be considered in patients with rapidly deteriorating lung functions. The prognosis is variable with frequent regression or stability of abnormalities.

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**Reference**