Case Report

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# Pulmonary Langerhans Cell Histiocytosis X Incidentally Diagnosed in a Non-Smoker Because Of Simultaneous Bilateral Spontaneous Pneumothorax

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## Abstract

**Introduction:** Pulmonary Langerhans cell histiocytosis X (PLCHx) is a rare interstitial lung disease of unknown aetiology and occurs mostly in young smokers. Lung involvement alone accounts for >85% of the cases; other organs may be involved too (5%–15%).

**Case report:** A 19-year-old boy was presented to the emergency service with sudden-onset, sharp, stabbing chest pain and dyspnoea. The patient had no history of smoking. His medical and family histories were negative for any disease. Thoracic radiography showed increased reticulonodular density with a near-total bilateral pneumothorax at the left and partially at the right. Hence, bilateral tube thoracostomy was performed. High-resolution computed tomography (HRCT) showed pronounced thin-walled cystic structures smaller than 10 mm and small millimetre-sized nodules in the upper and middle zones. Extrapulmonary Langerhans cell histiocytosis was excluded based on the radiological and laboratory findings. Based on the clinical and tomography findings, diagnosis of PLCHx was made.

**Conclusion:** Our case highlights that PLCHx should be suspected in non-smokers. Further studies elucidating the aetiology of PLCHx are needed.

**Keywords:** Histiocytosis X, spontaneous pneumothorax, interstitial lung disease.

## Introduction

Pulmonary Langerhans cell histiocytosis X (PLCHx) is a rare interstitial lung disease of unknown aetiology and occurs mostly in young smokers. It is characterised by abnormal accumulation of Langerhans-type histiocytes in the lung parenchyma[1,2]. An epidemiological hallmark of PLCHx is that it almost exclusively occurs in patients with a smoking history of >20 cigarettes per day. No other epidemiologic feature has been identified so far[3]. Herein, we report a case of PLCHx in a 19-year-old non-smoker. PLCHx was diagnosed incidentally during the treatment for simultaneous bilateral pneumothorax.

## Case Report

A 19-year-old boy was presented to the emergency service with sudden-onset, sharp, stabbing chest pain and dyspnoea. Physical examination revealed decreased respiratory sounds bilaterally, with louder sound on the left. The arterial blood pressure was 130/85 mm Hg, pulse was 115 bpm and fingertip oxygen saturation was 90%. The patient had no history of smoking. His medical and family histories were negative for any disease. Laboratory examination was negative for any pathology. Thoracic radiography showed increased reticulonodular density with a near-total bilateral pneumothorax at the left and partially at the right (Figure 1). Hence, bilateral tube thoracostomy was performed. Postoperative chest radiography showed improved lung expansion except for minimal pneumothorax detected on the left (Figure 2). High-resolution computed tomography (HRCT) showed pronounced thin-walled cystic structures smaller than 10 mm and small millimetre-sized nodules in the upper and middle zones (Figure 3). The right chest tube was removed on the 5th day and the left tube on the 12th day. Extrapulmonary Langerhans cell histiocytosis was excluded based on the radiological and laboratory findings. Because thoracotomy or thoracoscopy was not conducted, lung biopsy could not be performed. Based on the clinical and tomography findings, diagnosis of PLCHx was made. The patient was advised to avoid inhaling cigarette smoke. Steroid treatment was not started because he was clinically and medically stable following the pneumothorax treatment. He is on regular follow-up and is stable.

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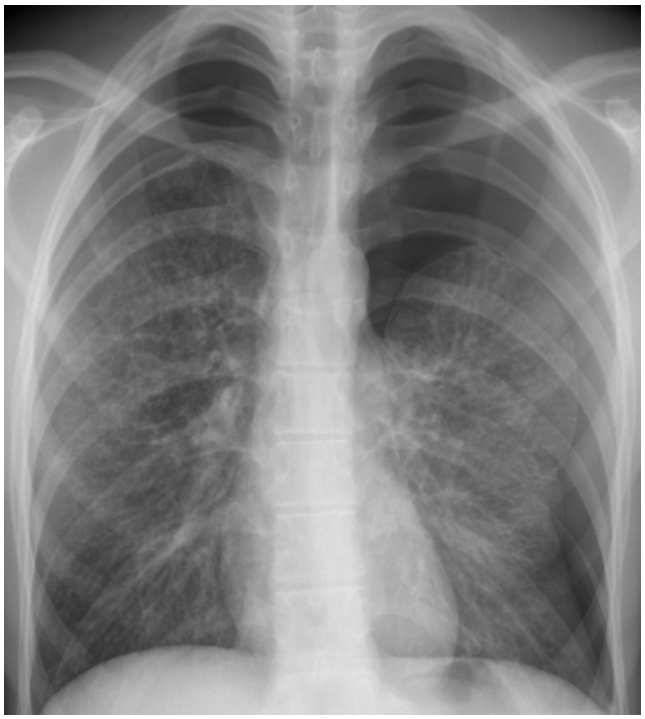

**Figure 1:** Chest radiograph with bilateral pneumothorax.

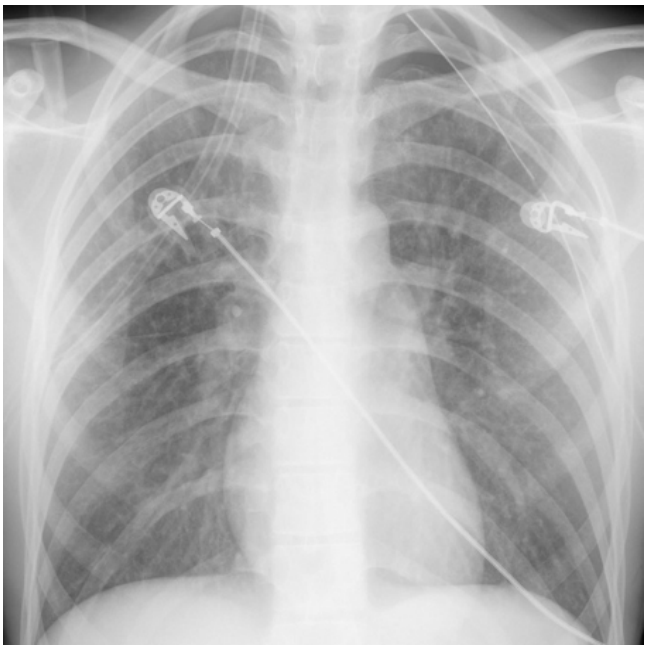

**Figure 2:** Bilateral lung re-expansion after bilateral drainage tube insertion

## Discussion

Langerhans cell histiocytosis is a group of disorders characterised by infiltration of a large number of Langerhans cells into tissues to form granulomas and is associated with a wide range of clinical features and outcomes. PLCHx is the pulmonary sub-form of this disorder; it is also known as pulmonary eosinophilic granuloma, pulmonary Langerhans

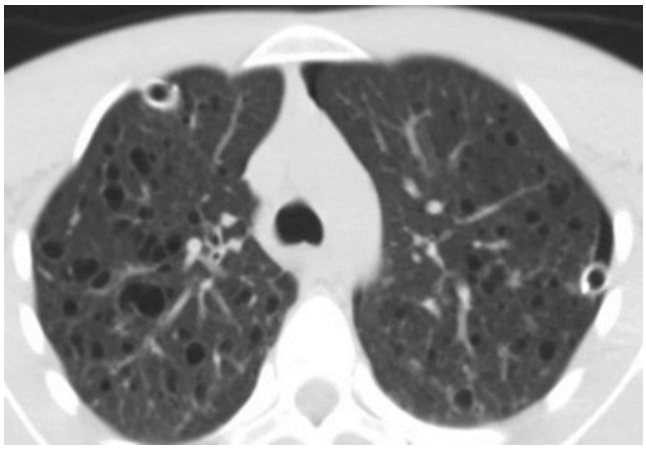

**Figure 3:** Chest computed tomography scan showing pronounced thin-walled cystic structures smaller than 10 mm in the upper and middle zones

cell granulomatosis and pulmonary histiocytosis[3,4]. Lung involvement alone accounts for >85% of the cases; other organs may be involved too (5%–15%)[2]. PLCHx occurs mostly in young adults; however, its actual incidence and prevalence are unknown. Almost all patients with PLCHx have a history of smoking; this indicates the critical role of smoking in its pathogenesis. However, no reliable epidemiological studies exist[2-4]. Our patient only presented with lung involvement, had never smoked and was not exposed to cigarette smoke. These findings imply that genetic and environmental factors could also be involved in the aetiopathogenesis of PLCHx. Therefore, further studies on this subject are required.

PLCHx is asymptomatic in approximately 25% of the cases and is mostly diagnosed incidentally during thorax radiography. Most patients present with cough, dyspnoea and laboured breathing. Recurrent unilateral or bilateral pneumothorax is seen in 10%–20% of the cases. Pneumothorax is resistant to air leak, and offering treatment options ranging from video thoracoscopy to pleurodesis might be required[5-7]. While our patient had no previous respiratory symptoms such as cough and dyspnoea, he presented with simultaneous bilateral pneumothorax. Pneumothorax was treated with tube thoracostomy, and no further surgical intervention was required.

PLCHx has characteristic findings on thorax radiography that help diagnosis, including irregularly limited or satellite nodules of 2–10 mm diameter, reticulonodular opacities, cysts and honeycomb appearance in the upper and middle zones and costophrenic angles. On HRCT imaging, thin-walled cysts sized <10 mm and nodules sized <5 mm are seen mostly in the upper zones. Cavitation can be observed in these nodules showing centrilobular and peribronchiolar distribution[1,2,8]. Thoracic radiography, HRCT and clinical findings are deemed sufficient for diagnosis when patients choose not to undergo surgical procedures and tissue samples cannot be obtained. The differential diagnoses basis tomography findings include lymphangioleiomyomatosis,

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tuberous sclerosis, hypersensitivity pneumonia, sarcoidosis and end-stage idiopathic pulmonary fibrosis.

The main treatment objective is cessation of smoking. Smoking cessation has been shown to stabilise the disease symptomatically, radiologically and physiologically[2-4]. However, reports of no progression up to 12 years in patients who continue smoking exist[9]. Similar to our case, PLCHx has been diagnosed in patients who never smoked and had no exposure to cigarette smoke. Reports have suggested that patients with predominant nodular lesions may respond to corticosteroid therapy. Cytotoxic drugs such as vinblastine, methotrexate, cyclophosphamide and etoposide can only be used in patients with multisystem involvement or steroid-resistant progressive disease[2-4].

## Conclusion

Our case highlights that PLCHx should be suspected in non-smokers. Further studies elucidating the aetiology of PLCHx are needed.

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