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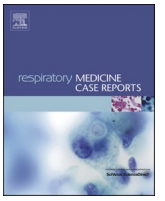

Case Report

# Pulmonary Langerhan's cell histocystosis presenting with bilateral simultaneous pneumothoraces – Case report

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ABSTRACT

We describe the case of a young male, with no significant medical history, who presented to the Emergency Department (ED) with severe respiratory compromise. He suffered a respiratory arrest shortly after presentation. An initial chest x-ray performed post intubation revealed bilateral pneumothoraces with evidence of abnormal underlying lungs. Through a series of investigations, he was diagnosed with Pulmonary Langerhans Cell Histocystosis. In this article, we outline the initial presentation, subsequent acute management and the clinical course pertaining to this man's presentation. We believe this is the first reported presentation of bilateral simultaneous pneumothoraces, with previously unknown Pulmonary Langerhans Cell histiocytosis (PLCH).

## 1. Introduction

Pulmonary Langerhan's Cell Histiocytosis (PLCH) is a rare condition characterised by infiltration of specialised myeloid cells that share morphological and surface receptor markers with epidermal Langerhans cells [1]. These cells carry mutations of the BRAF and/or NRAS, KRAS and MAPK1 genes [2]. The exact incidence of PLCH is unknown however the incidence of Langerhan's Cell Histocytosis is around 1-2 adults per million per year [2,3]. Spontaneous pneumothoraces can be a major issue for patients with PLCH [2,3]. Patients are vulnerable to pneumothoraces due to the cystic nature of the disease [2,3]. 30–45% of patients suffer from a pneumothorax at some stage of the disease while 15–20% of patients initially present with a unilateral spontaneous pneumothoraces [1]. In this article, the patient uniquely presented with bilateral spontaneous pneumothoraces. This patient was suffering from dyspnoea for roughly 1 year prior however no diagnosis was made in the community. Our case details the initial presentation of a patient with PLCH along with the investigations and management surrounding this presentation. This case demonstrates the complexities of managing a new diagnosis of PLCH as well as life-threatening bilateral pneumothoraces.

## 2. Clinical case

A young male was transferred from home by ambulance to the ED of a peripheral hospital in acute respiratory distress. He had an audible wheeze at bedside with a silent chest on auscultation. His oxygen saturations on pulse oximeter were 50% on room air.

Although he did not have any significant medical history, he had recently been referred by his General Practitioner for initial investigation of progressive dyspnoea. He also had a history of significant tobacco and cannabis use.

He was initially treated as a case of life-threatening asthma exacerbation. He received iv hydrocortisone, iv magnesium sulphate

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and nebulised bronchodilators. Arterial blood gas on presentation revealed a pH of 7.0, pCO2 of 15.7kPa, and a pO2 of 4.5kPa. He subsequently had a respiratory arrest and he was intubated. The first chest x-ray was performed post intubation (Fig. 1). This revealed bilateral pneumothoraces with significant underlying pulmonary parenchymal disease. Bilateral chest drains were placed following this x-ray.

He was transferred to a tertiary care centre. He was transitioned from invasive ventilation to successful extubation over the following days. He subsequently underwent a bilateral Video-Assisted Thoracic Surgery (VATS) with bilateral talc pleurodesis and a right upper and lower lobe wedge resection. He had bilateral drain insertions placed during the surgery. High resolution Computed Tomography (CT) thorax demonstrated extensive bilateral cystic lung disease with a large right sided persistent pneumothorax (Fig. 2).

He had a 58-day admission with complications, with lung re-expansion and persistent air leaks from the bilateral drains. He had his left drain removed after 2 weeks, however it was replaced during a CT guided procedure on week 4. He had a persistent right-sided air leak but his chest drain was eventually removed on week 4 with no subsequent re-insertions.

Wedge biopsy of the right lung confirmed Pulmonary Langerhans Cell Histiocytosis (PLCH). This correlated with the clinical presentation and history.

He had a persistent left sided pneumothorax on discharge. This was seen during his follow-up chest x-ray 2 weeks post discharge. However, he reported feeling much improved with no residual dyspnoea and agreed to smoking cessation. Pulmonary function testing (PFT) has not been performed to date, due to his pneumothoraces. Further follow up imaging has demonstrated complete resolution of pneumothoraces, with PFTs now planned for the future.

## 3. Discussion

PLCH most commonly affects young adult smokers [1]. It affects genders equally and greater than 90% of patients with PLCH are current or ex-smokers [1]. The quantity of cigarettes smoked or the duration of smoking does not influence the development of PLCH [2]. There have also been observations that 20–33% of patients with PLCH smoked both tobacco and cannabis [2]. The patient in the case fits the typical profile for PLCH as described above with both tobacco and cannabis use.

PLCH can present in a multitude of ways [1,2]. 60% of patients initially present with symptoms such as a cough and dyspnoea [1]. 15–20% of patients present with constitutional symptoms such as fever and night sweats [1,2]. PLCH presents with spontaneous unilateral pneumothorax in 15–20% of patients [1]. There is no previous data regarding presentation with bilateral pneumothoraces as in our case.

30–45% of patients have a spontaneous pneumothorax at some point during the course of their disease [2]. Bilateral spontaneous pneumothorax (SBSP) occur in 1.3% of all spontaneous pneumothorax events [4]. These figures are estimates based on case by case study reports due to the rarity of SBSP. SBSP occur in both primary and secondary disease [4]. For secondary pneumothoraces, there have been multiple conditions that are implicated as potential causes of bilateral pneumothoraces. These include chronic obstructive pulmonary disease, tuberculosis, silicosis, staphylococcus aureus pneumonia, pneumocystis carinii infection and PLCH [4–6]. On review of the literature, there was one case report of a patient with bilateral pneumothoraces with previously known PLCH [6].

If a patient initially presents with a pneumothorax, there is a 60% chance of pneumothorax recurrence during the disease course [2]. Pleurodesis is often recommended after the first episode of pneumothorax, however it has not been proven to reduce the number of pneumothoraces these patients suffer from overall [2]. The patient in this case had bilateral pleurodesis performed however suffered from right sided pneumothorax recurrence and issues with re-expansion post pleurodesis.

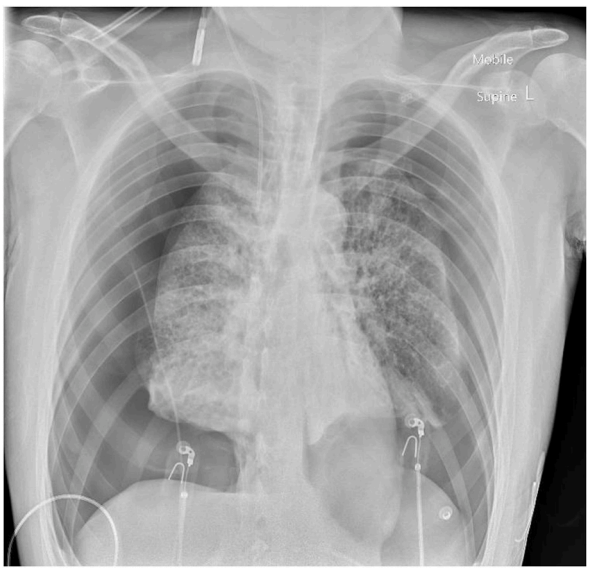


**Fig. 1.** Initial chest X-Ray.

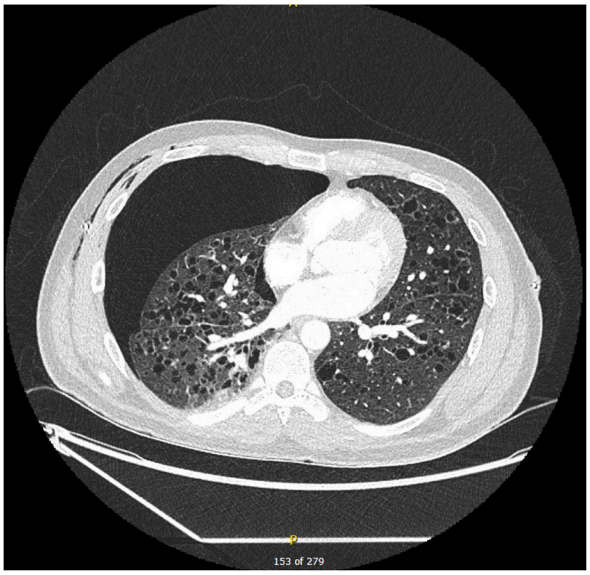


**Fig. 2.** High Resolution CT Thorax post bilateral VATS.

A study involving 102 adults with biopsy confirmed PLCH showed a median survival of 12.5 years [1]. A proportion of patients developed stable disease with little or no disease progression [1]. Predictors of worse outcomes include lower $FEV_1$, lower $FEV_1$/FVC ratio and higher RV/TLC ratio [1]. A significant proportion of patients develop obstructive lung disease [1]. This can contribute to the worsening of pulmonary function despite stable PLCH [1].

When managing patients with PLCH, a critical element of patient care is smoking cessation [2]. This can lead to partial regression and stabilisation of PLCH alone [2]. Glucocorticoids have also been recommended, however, it is not clear what the appropriate dose or duration of treatment is [2]. Chemotherapy agents are often used for paediatric cases, however, there is limited data available in adults with PLCH without systemic disease [2]. The discovery of BRAF mutation in PLCH has led to the use of targeted therapies [1]. Treatment with BRAF inhibitors such as Vemurafenib, have shown stabilisation of disease for some patients [1,2]. Lung transplantation can be offered to patients with respiratory failure or with significant pulmonary hypertension [2]. Patient with PLCH have similar outcomes post-transplant to patients with other interstitial lung diseases [2]. Less than 50% of patients survive for 5 years. Recurrence of PLCH has been found in 20% of patients post-transplant [2].

Regarding the management of the gentleman in this case, smoking cessation is essential in his ongoing management. He will have lifetime follow up with a respiratory specialist. There may be a role for BRAF therapies, or lung transplant in the future depending on disease progression.

## 4. Conclusion

In conclusion, this young gentleman had a history and presentation in keeping with PLCH. However, his initial presentation with bilateral pneumothoraces as opposed to unilateral pneumothorax is atypical. There is one other case reported in the literature, however in this case the patient had a known diagnosis of PLCH prior to their presentation with bilateral pneumothoraces.

## CRediT authorship contribution statement

**Sophie Buckley:** Writing – original draft. **Emily O'Reilly:** Writing – review & editing. **Deirdre Doyle:** Writing – review & editing. **Desmond Murphy:** Writing – review & editing.

## Declaration of competing interest

No conflict of interest.

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