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Tension Pneumothorax as the Presentation of Pulmonary Langerhans Histiocytosis

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Abstract A 36 year old male smoker presented to the emergency department with shortness of breath. A chest x-ray showed a tension pneumothorax. Chest tube was placed urgently. CT scan showed upper lobe predominant cystic lung lesions. Biopsy confirmed the diagnosis of pulmonary Langerhans Histiocytosis.

Keywords: *tension pneumothorax, pneumothorax, langerhans, histiocytosis, lagerhans histiocytosis*

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1. Introduction

Langerhaan's histiocytosis is rare disease affecting 1-2 cases per million [1]. Langerhans is characterized by a proliferation of bone marrow derived dendritic cells [2] that cause localized destruction at sites of proliferation. Gold standard for diagnosis is positive stain for CD1a cells [1,2,3,4]. The disease is classified based on organ involvement. Langerhans can be either multi organ system or single organ system. Pulmonary Langerhans histiocytosis can occur as part of a multisystem disease or isolated entity. Isolated pulmonary Langerhans accounts for 20% of adult disease [1].

2. Case

A 36 year old male smoker with no past medical history presented to the emergency department. He had first noted feeling ill five days prior with nasal drainage, night sweats and non-productive cough. Two days prior to admission he began experiencing right sided chest pain that was sharp, sudden in onset and unrelenting. When the chest pain wouldn't go away he came into the emergency department.

A chest X-ray in the emergency department showed a tension pneumothorax. A chest tube was immediately placed; repeat chest X-ray showed resolution of the tension pneumothorax and interstitial prominence and cystic changes. CT of the chest with contrast showed upper lobe predominance of cystic lesions.

The patient was admitted to the hospital for management of his pneumothorax and further work up of his underlying condition. A quantiferon gold, PPD and AFB smear x1 were all negative. Bronchoscopy was considered but thought to be relatively low yield. Cardiothoracic surgery took the patient to the OR for a video-assisted thoracoscopic surgery (VATS) biopsy.

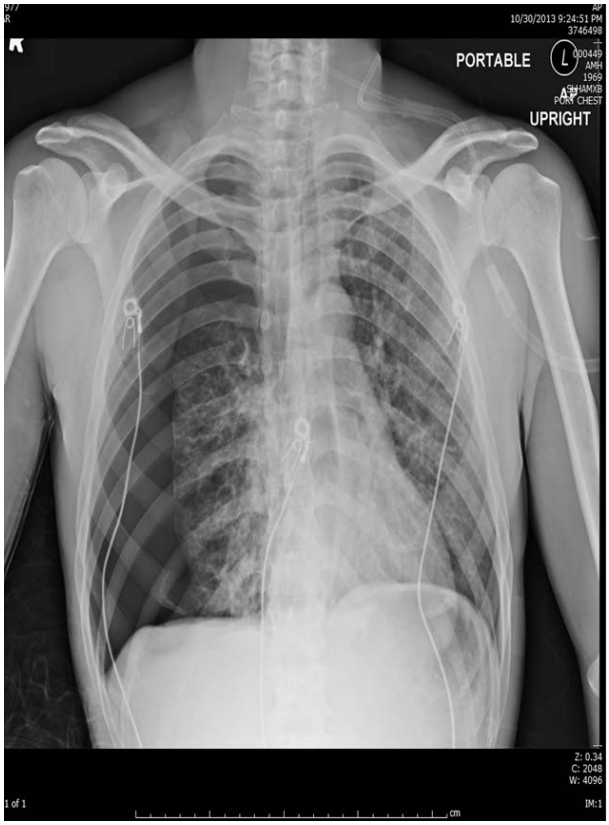

Figure 1. Portable Chest X-ray in the Emergency Department

There is a large right pneumothorax which causes compression of the right lung and leftward shift of the heart and mediastinum. There is no pleural effusion. Interstitial prominence and cystic changes are demonstrated in the lungs. No acute osseous injury is visible.

Lung biopsy showed on three of three samples the specimen was stained with S-100 that showed few scattered individual Langerhans histiocytes. Cd1a stain

was positive showing few scattered focal collections of Langerhans cells.

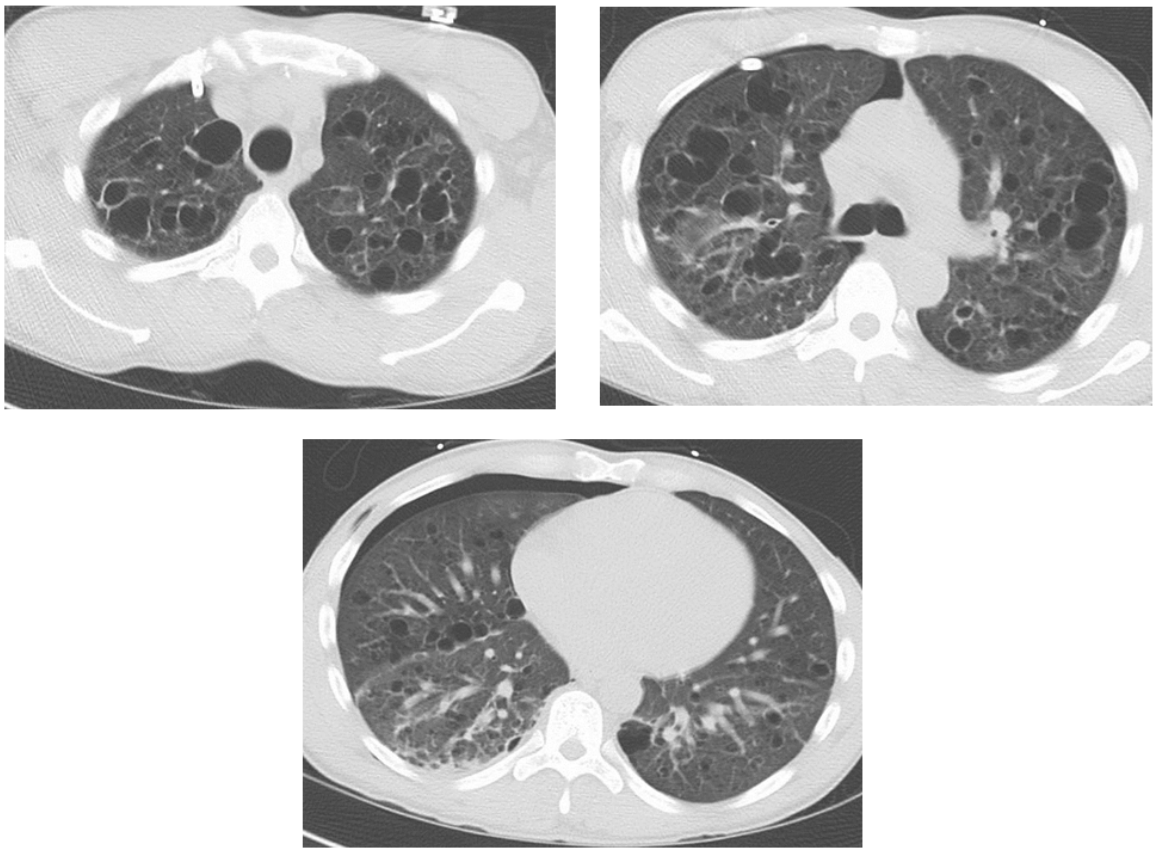

Figure 2. High Resolution CT scan done one day after admission shows an upper lobe predominant cystic disease

Multiple cysts are seen throughout the lungs bilaterally, some of which are bizarre shaped, in a predominantly upper lobe distribution. The lungs are otherwise free of focal consolidations. There is no evidence of bronchiectasis, reticulation, ground glass opacities, honeycombing or air trapping. There is no pleural effusion.

Further work up for extra pulmonary manifestations was planned as an outpatient. Unfortunately the patient was rehospitalized three times in the following 8 months for recurrent pneumothorax. He missed scheduled appointments and was briefly hospitalized for suicidal ideation. In total he only made one scheduled pulmonary appointment before being lost to follow up.

3. Importance

Pulmonary Langerhans is an upper lobe predominant nodular and cystic lung disease [3]. It is strongly associated with smoking [5] and smoking cessation is the critical [1,3] therapy for patients. Outcomes are variable and range from spontaneous resolution to progression to respiratory failure [3]. For those that progress to respiratory failure lung transplantation is a viable option [5].

The diagnosis of Pulmonary Langerhans Histiocytosis should be strongly considered in smokers presenting with pneumothorax and bilateral indeterminate infiltrates [3]. Definitive diagnosis is made by biopsy and stain with for CD1a cells [1].

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