Langerhans Cell Histiocytosis with Lungs and One Rib Involvement: a case report

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Pulmonary Langerhans cell histiocytosis (LCH) is a rare disease. The typical findings on chest computed tomography (CT) of this disease are multiple bizarre cystic spaces associated with small peribronchiolar nodules, predominantly in the upper third of both lungs. The peribronchiolar nodules are not always present due to the evolution sequence. For patients with histologically proven extrapulmonary LCH, the diagnosis of pulmonary LCH is usually established if the findings on high-resolution CT are consistent with pulmonary LCH. Recently we encountered one patient with histologically proven osseous LCH. The chest CT feature of this patient was multiple bizarre cystic spaces without obvious pulmonary parenchymal nodule.

Key words: Histiocytosis; Lung, CT; Lung, disease; Rib, neoplasms

Pulmonary Langerhans cell histiocytosis (LCH) is a rare disorder due to diffuse destruction of distal airways caused by granulomas containing Langerhans’ cells. The etiology is unknown and it typically affects young adult smokers [1]. The typical CT features of the lung are mixed irregularly shaped cystic spaces and small peribronchiolar nodular opacities, predominantly in the middle and upper lobes. The presence of typical CT features on high-resolution computed tomography (CT) frequently allows the clinician to make a diagnosis of pulmonary LCH without lung biopsy [2]. Here, we report a histologically proven case of LCH and the imaging findings.

CASE REPORT

A 27-year-old male was admitted to our hospital with chief complaint of left side chest pain. The patient’s medical history was unremarkable, but he had a habit of smoking about one pack per day for many years. He suffered from sudden onset of left side chest pain with mild dyspnea in early morning. The pain prompted him to seek emergent medical care later on the same day. At emergency room, physical examination showed decreased breathing sounds over his left lung. Chest radiograph showed pneumothorax at left lung with one well-defined, bubbly expansile lesion at the left seventh rib. Multiple bilateral ill-defined nodules and reticular opacities predominantly in the upper and middle lung zones were also identified. After left side chest tube insertion, the following chest radiograph showed that these multiple ill-defined nodules and reticular opacities at bilateral lungs were symmetrically distributed (Fig. 1a). The contrast-enhanced CT of the chest performed two days later revealed multiple cysts with thick wall but no obvious nodule (Fig. 1b). Confluent cysts led to the appearance of bizarre-shaped cystic spaces (Fig. 1c). These lesions were particularly predominant in the upper and middle lung fields, with sparing of costophrenic angles, bilaterally and symmetrically (Fig.1c, 1d). There was an expansile lesion on the left seventh rib without obvious

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sclerotic change (Fig. 2).

Thoracotomy with chest wall resection was performed. The pathology of the rib lesion showed that it was composed of many mononuclear polygonal cells, some eosinophils, some osteoclast-like giant cells, some lymphoid cells (Fig. 3) and occasional foreign body giant cells. After immunostaining, these mononuclear polygonal cells were positive for CD68, CD1a and S100. Thus, the pathological diagnosis of the bony lesion was LCH. Tissue biopsy of lung parenchyma wasn’t performed due to the risk of pneumothorax. However, the clinical diagnosis was pulmonary LCH.

**DISCUSSION**

LCH, formerly known as histiocytosis X or eosinophilic granuloma, is a disease of abnormal clonal proliferation of a unique type of cell in the monocyte-macrophage cell line known as the

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**Figure 1.** A 27-year-old male with LCH involving lungs and the rib. 

**a.** Chest radiograph shows symmetrical reticulonodular pattern at both lungs, more severe on both upper lung fields and one expansile lesion at the left seventh rib without obvious sclerotic rim. 

**b.** and **c.** Enhanced CT of chest shows multiple thick-walled bizarre-shaped cysts, particularly predominant in the upper and middle lung field. 

**d.** Enhanced CT shows that bilateral costophrenic angles are spared.
Langerhans’ cell [3]. The original description of this unique type of cell was made by Paul Langerhans in 1868 [4]. In 1985, the Histiocyte Society was founded to address the study of disorders of histiocytes. Currently, eosinophilic granuloma and histiocytosis X are referred to as LCH to reflect the cellular basis of the disease. The use of older terms is discouraged.

The combination of diffuse, irregularly shaped cystic spaces with small peribronchiolar nodular opacities, predominantly in the middle and upper lobes, is highly suggestive of pulmonary LCH [5]. However, parenchymal nodules are not always present in pulmonary LCH [5]. This may be due to the evolutionary sequence of pulmonary lesions of LCH, which is as follows: nodules, cavitated nodules, thick-walled cysts and then thin-walled cysts [6]. According to the literature reports, for pulmonary LCH in which parenchymal nodules were absent, the radiological images showing characteristic distribution of the cystic changes could aid in the diagnosis [5]. In our case, there were minimal peribronchiolar opacities, and the distribution of the cystic spaces did not involve costophrenic angle. These appearances are consistent with pulmonary LCH.

In addition, there was well-defined expansile and remodeled bony lesion on one rib without sclerotic rim, which was pathologically proved to be LCH. The radiological appearance of osseous LCH depends on the site of involvement and the phase of the disease [7]. Late lesions may show well-defined sclerotic margin and expansile remodeled appearance [7]. For patients with documented extrapulmonary LCH, such as in the skin or bone, the diagnosis of pulmonary LCH is usually established if high-resolution CT shows the specific features [5]. Thus, the final diagnosis of our case was pulmonary LCH.

In conclusion, the typical imaging findings of pulmonary LCH are mixed diffuse, irregularly shaped cystic spaces and small peribronchiolar nodular opacities, predominantly in the middle and upper lobes. But the peribronchiolar nodules are not always present due to the different stages of this disease. The characteristic distribution of pulmonary lesions is the most important discriminating factor [5]. Particular attention should be paid to the lowermost lung, especially costophrenic angles, where usually there is minimal or free of disease [5]. For patients with documented extrapulmonary LCH, the diagnosis of pulmonary LCH is usually established if high-resolution CT shows features of multiple bizarre cysts with or without small peribronchiolar nodules, predominantly in the middle and upper lobes of bilateral lungs.

**REFERENCES**

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同時出現肺部和肋骨之蘭格罕氏細胞組織細胞症：病例報告

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肺部Langerhans cell histiocytosis（LCH）是一種稀有的疾病。典型的胸部CT影像特徵是許多奇形怪狀的囊狀空腔與一些細支氣管旁的小結，主要是分佈在上部三分之一的肺部。但是由於疾病進展關係，細支氣管旁的小結並非總是會表現出來。在組織學下證明是肺外LCH的病患中，如果high-resolution CT的特徵與肺部LCH一致的話就可以診斷是肺部LCH。最近我們遇到一位在組織學下證明是骨的LCH的病患。這位病患的肺部影像表現出許多奇形怪狀的囊狀空腔但卻沒有明顯的肺內的小結。

關鍵詞：細胞組織細胞症；肺，電腦斷層攝影；肺部疾病；肋骨，腫瘤