Primary Pulmonary Langerhans Cell Histiocytosis: A Case Report

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Abstract: Pulmonary Langerhans cell histiocytosis (PLCH) is the clonal proliferation of Langerhans cell confined within the lung as an uncommon interstitial lung disease. The patients with PLCH are always asymptomatic while the spontaneous pneumothorax is its third common presentation. It has rarely been reported in Thai patients. Herein we report an additional case of PLCH. The patient is a 30-year-old Thai smoker who presents with sudden onset of left spontaneous pneumothorax without constitutional symptom. He is immediately treated with the intercostal drainage (ICD) and later with thoracotomy and lung repair because of the continuous leakage of the air from the lung. The HRCT reveals left pneumothorax, multifocal cystic lesions in bilateral upper lungs and superior segment of lower lungs, suggesting Langerhans cell histiocytosis. The microscopic pathology of the lung biopsy shows positive staining for S100 and CD1a, the typical findings of PLCH. He is finally diagnosed as PLCH. After lung repair, no pleurodesis, no corticosteroid, no systemic chemotherapy is offered to him. With only smoking cessation, he does not have any recurrent spontaneous pneumothorax within three months of follow-up although the recurrence of ipsilateral pneumothorax is common in case of PLCH who are treated with only ICD without pleurodesis.

Key words: Spontaneous Pneumothorax, Pulmonary Langerhans Cell Histiocytosis

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Pulmonary Langerhans cell histiocytosis (PLCH) is an uncommon interstitial lung disease that is almost exclusively found in the young cigarette smokers. Most cases of PLCH always manifest as an isolated disorder more than as a component of multisystem illness. The patients with PLCH are always asymptomatic while the second and third common presentations are insidious onset of nonproductive cough and spontaneous pneumothorax, respectively. Other symptoms may include pleuritic chest pain,
fatigue, weight loss and fever while the physical examination in most cases is always unremarkable\(^3\).

In Thailand, 15 cases of histiocytosis-X including 4 cases of eosinophilic granuloma were reported\(^4\) pulmonary involvement with massive pneumothorax in a child with Langerhans cell histiocytosis (LCH) was reported in 1992\(^5\), a fatal case of multisystem Langerhans cell histiocytosis with incidental pulmonary dirofilariasis was firstly publicized in 1998\(^6\) since then a case of congenital self-healing Langerhans cell histiocytosis\(^7\), 40 cases of Langerhans cell histiocytosis\(^8\) and two cases of PLCH are sequentially reported\(^9\,10\). Herein we report an additional case of PLCH from Thailand.

**Case Report**

A 30-year old Thai man presented with sudden onset of chest tightness and dyspnea without constitutional symptom for six days. He was examined to have spontaneous pneumothorax at the left lung. He smoked 25 pack-years. His body mass index was 21.23 kg/m\(^2\). He was immediately treated with the intercostal drainage (ICD). The chest film after the lung had expanded revealed the diffuse interstitial infiltrations.

Laboratory tests: Hb14.2 g%, WBC 9,500/mm\(^3\), platelet 314,000/mm\(^3\), MCV 70.8fL, MCH 22.1 pg, RDW 16.0 %, normal liver and kidney function tests, cholesterol 233 mg%, LDH 347 U/L, no HBsAg, no anti-HIV, Na 138.3, K 4.3, Cl 100.2, CO2 27.2 mEq/L.

After the immediate ICD, the air continued leaking from the lung for many days and he was finally operated with thoracotomy with lung repair and lung biopsy.

Lung biopsy: presence of small nodules and alveolar lining of mixed population of cells including Langerhans cells, eosinophils, histiocytes, the neoplastic cells revealed focally positive staining for S100 and CD1a; differential diagnosis included Langerhans cells histiocytosis.

The chest film at the initial presentation revealed left pneumothorax without the shift of the midline. The HRCT: left pneumothorax, multifocal cystic lesions in bilateral upper lungs and superior segment of lower lungs, differential diagnosis: Langerhans cell histiocytosis or lung cysts

He was finally diagnosed as PLCH and no chemotherapy was offered for him. He did not have recurrent spontaneous pneumothorax / lung nodule / lymphadenopathy / constitutional symptoms during three months of follow-up.

**Discussion**

Although PLCH is very rare disease, our case is pathologically proven to be PLCH because of the lung biopsy that shows the typical findings of the positive immunohistochemical staining for S100 protein and strong presence of CD1 antigen on the cell surface, the features of PLCH which will not be observed in other cells of histiocytic origin\(^11\). And it is good to finally treat him with thoracotomy and lung repair because the lung tissue can be yielded for pathological examination and the definite diagnosis can be established.

In the review of 102 adults with PLCH with 4-year median follow-up, their overall median survival is 12.5 years which is shorter than that expected for persons of the same sex and calendar year of birth. Half of deaths are attributable to respiratory failure.
(15 from 33 deaths from all 102). Variables predicting shorter survival are an older age, a lower forced expiratory volume in one second (FEV1), a higher residual volume, a lower ratio of FEV1 to forced vital capacity, and a reduced carbon monoxide diffusing capacity.\(^{(12)}\)

For treatment of PLCH, smoking cessation is strongly recommended and some patients respond well to it. So far, there is no definite guideline of treatment for PLCH. Only patients with prominent nodular opacities should be treated with corticosteroid because they will respond well to therapy.\(^{(3)}\) Corticosteroid is not offered to our case after operation because he has no lung nodule, no constitutional symptom, or no residual lung function abnormality after recovery from pneumothorax.

In general, spontaneous pneumothorax is the initial presentation among 16% of patients with PLCH, and 62.5% of these have recurrence. Fifty-eight percentage recurs to the ipsilateral side when pneumothorax is managed by observation or the chest tube without pleurodesis but 0% after surgical management with pleurodesis.\(^{(13)}\) However our case is still free from recurrence of pneumothorax within 3 months of follow-up with just only smoking cessation.

Besides of spontaneous pneumothorax, 15 of 17 patients (88.2%) with pathologically proven PLCH are demonstrated to have pulmonary hypertension (estimated pulmonary artery systolic pressure or PASP at rest \(>35\) mm Hg) on the echocardiography.\(^{(14)}\) It seems pulmonary hypertension is the far more common finding than any other symptoms or signs among patients with PLCH although they do not manifest any clinical clue. Therefore the echocardiography is also in the plan of further investigation in our case.

**Conclusion**

A 30-year-old Thai man suddenly develops left spontaneous pneumothorax. The intercostal drainage, thoracotomy with lung repair, and lung biopsy are performed. The microscopic pathology reveals positive S100 protein and CD1 antigen on the cell surface, the typical findings of pulmonary Langerhans cell histiocytosis (PLCH). No other lesion is recognized in anywhere else so he is definitely diagnosed as PLCH, the rare disease in Thailand. Without systemic chemotherapy, he has no recurrent pneumothorax during three-month follow-up.

**References**


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