Pulmonary Langerhans cell histiocytosis causing spontaneous bilateral pneumothorax in a child

Anupam Patra[a], Sujit K. Bhattacharyay[a], Arnab Maji[a], Abhijit Mandal[b]

[a]Department of Pulmonary Medicine, NRS Medical College and Hospital, Kolkata, [b]Department of Pulmonary Medicine, Bankura Sammilani Medical College, Bankura, West Bengal, India

Correspondence to Anupam Patra, MD, 2/15 A, Bijoygarh, Jadavpur, Kolkata - 700 032, West Bengal, India
Tel: 09836139511
e-mail: dranupam.patra@gmail.com

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Bilateral pneumothorax is very rare in childhood. Moreover, if it is due to pulmonary involvement of Langerhans cell histiocytosis, it is even rarer in childhood. In our case, a nonsmoker 12-year-old boy presented with bilateral pneumothorax, whose high-resolution computed tomography scan was highly suggestive of pulmonary Langerhans cell histiocytosis. Excision biopsy of a clinically palpable cervical lymph node and histopathological examination and immunohistochemistry positivity for CD1a indicated a diagnosis of Langerhans cell histiocytosis. Clinicians should consider pulmonary Langerhans cell histiocytosis in differential diagnoses in dealing such a case.

Keywords:
bilateral pneumothorax, childhood, pulmonary Langerhans cell histiocytosis

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Introduction

Bilateral spontaneous pneumothoraces may occur in children as a result of rupture of subpleural blebs into the pleural space, in children with underlying lung diseases such as cystic fibrosis, pulmonary Langerhans cell histiocytosis (PLCH) and those with connective tissue disorders such as Marfan syndromes, cutis lexa, Ehler–Danlos syndrome, and as a result of underlying genetic disorder [1,2]. PLCH has been attributed to the occurrence of recurrent pneumothorax mainly in adults in whom smoking is a key etiologic factor [3]. However, lung involvement in Langerhans cell histiocytosis (LCH) is very infrequent in children [4]. Here, we report a case of a 12-year-old child presented with bilateral pneumothorax along with a few extrapulmonary features, and PLCH was found to be the cause of his pneumothorax.

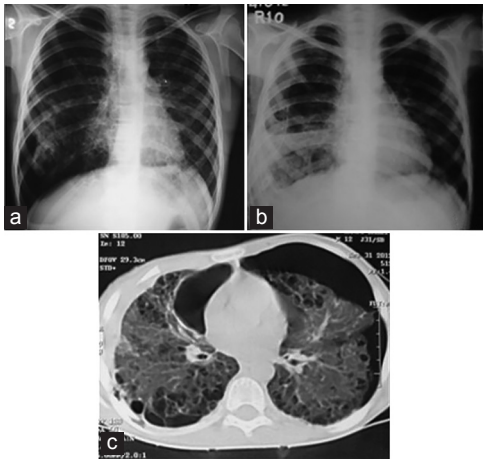
Case report

A 12-year-old boy was presented to us with sudden onset of dyspnea along with right-sided chest pain. He had this episode of dyspnea while playing football with his mates. On examination, the child was found to be malnourished, tachypneic (respiratory rate was 35/min), and hyper-resonant percussion note on most of the right hemithorax was observed. His peripheral blood showed hemoglobin of 10.7 g/dl, total leukocyte count of 11 000/mm^3, with neutrophils 50%, lymphocytes 47%, and eosinophil 3%, and normal liver function test. Chest radiography (posterior-anterior view) showed the presence of right-sided pneumothorax, with some reticular opacities in the mid-zones of both the right and the left lung (Fig. 1a). Intercostal chest tube was inserted in the right fourth intercostal space in the posterior axillary line and dyspnea decreased, but follow-up chest radiography even after the fifth day of tube thoracostomy did not reveal full resolution of pneumothorax. On the sixth day, dyspnea again increased and a repeat chest radiography revealed a left-sided pneumothorax for which another intercostal chest tube was inserted on the left side (Fig. 1b). High-resolution computed tomography (HRCT) scan of the thorax revealed bilateral pneumothorax along with multiple cystic spaces of unequal size involving all lobes of the lungs bilaterally (Fig. 1c). Meanwhile, the boy developed an enlarged right-sided cervical lymph node that was hard, discrete, and freely mobile. We did an excision biopsy of that lymph node, which on histopathological examination revealed characteristic reniform Langerhans cells with abundant eosinophils (Fig. 2). Immunohistochemistry was positive for CD1a. Red, papular, ulcerative skin lesions appeared on the anterior chest wall during the course of the disease. Oral mucosa and teeth were healthy. The radiography of skull was normal. Bone marrow aspiration and biopsy were normal. Ultrasonography of whole abdomen was normal. At 24 h, urinary volume was 1.3 l. MRI was also normal. Bronchoalveolar lavage (BAL) analysis revealed the presence of 1% Langerhans cells among the total BAL cells.

Discussion

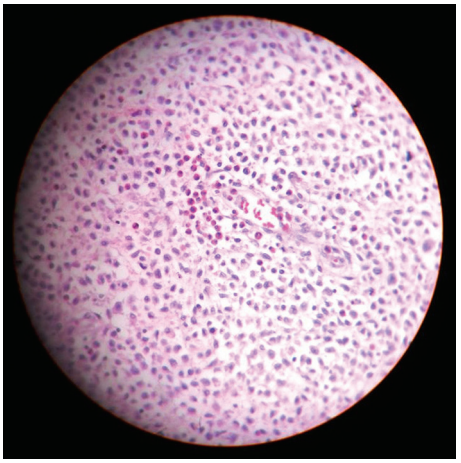
More than 90% of adult PLCH occurs in young adults who smoke, usually more than 20 cigarettes per day [5]. It is rare in children. The incidence of LCH

Figure 1



Chest radiography showing right-sided pneumothorax with nonhomogenous opacity in the mid-zone (a), appearance of left-sided pneumothorax with tube *in situ* on the right side (b), HRCT scan thorax showing bilateral pneumothorax along with multiple cystic spaces of unequal size bilaterally (c). HRCT, high-resolution computed tomography.

Figure 2



Characteristic reniform Langerhans cells with abundant eosinophils on H&E staining.

has been estimated to be two to 10 cases per million children aged 15 years or younger [6]. Pulmonary involvement is present in ~25% of children with multisystem low-risk and high-risk LCH. However, no data on isolated involvement of the lung in LCH in childhood are present. High-risk includes involvement of the liver, spleen, and bone marrow, and low-risk includes involvement of the skin, bone, lymph nodes, gastrointestinal tract, pituitary gland, and central nervous system. Pulmonary involvement carries no prognostic significance *per se* [7]. In our patient we found involvement of the lung predominantly, along with some lymph-nodal and skin involvement.

Radiologically, the classical description of the disease is bilaterally symmetrical involvement predominating the upper and middle lung fields, relatively sparing the costophrenic angle and giving a very characteristic picture on HRCT scan [4]. Initial nodular changes are followed by cystic changes or fibrosis in the late period. Confluence of cysts may lead to bullous formation, and bullae, especially in the lung apex, can rupture producing spontaneous pneumothorax, which often may be the first presentation of the disease as in our case.

If the HRCT scan of the thorax is typical of PLCH, then further evaluation with lung biopsy is usually not required. In our case, HRCT thorax features were suggestive of PLCH. A pathologic diagnosis is always required, and LCH cells should stain with antibodies to CD1a or anti-Langerin (CD207) to confirm the diagnosis of LCH [8]. BAL count of more than 4% of Langerhans cells is diagnostic of PLCH, although it is present in only 50% cases [9]. Histopathological diagnosis, as was already made by lymph node excision biopsy, and radiological picture were very highly suggestive of PLCH; we did not further proceed with lung biopsy because the child was too sick to tolerate it.

Although pulmonary involvement in LCH is rare in children, we have to consider it in differential diagnosis, especially when a child presents to clinicians with recurrent or bilateral pneumothorax.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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