Spontaneous pneumothorax with Isolated pulmonary langerhans cell histiocytosis in an adult case: A common manifestation of rare disease

Mitra Samareh Fekri[1], Faranak Salajegheh[1], Mohsen Nakhaie[1], and Mohammad Rezaei Zadeh Rukerd[1]

[1]Kerman University of Medical Sciences

February 6, 2023

Abstract

A 30-year-old man presented with sudden chest pain and evidence of multiple cystic lesions in both long and left-sided pneumothorax in HRCT. H&E stained section and IHC for CD1a and S100 were positive in lung samples. The patient was treated with the diagnosis of Isolated pulmonary langerhans cell histiocytosis.

Spontaneous pneumothorax with Isolated pulmonary langerhans cell histiocytosis in an adult case: A common manifestation of rare disease

Mitra Samareh Fekri[1], Faranak Salajegheh[2], Mohsen Nakhaie[3],

Mohammad Rezaei Zadeh Rukerd[3]

1. Cardiovascular Research Center, Institute of Basic and Clinical Physiology Sciences, Kerman University of Medical Sciences, Kerman, Iran, Kerman, Iran
2. Clinical Research Development Unit, School of Medicine, Afzalipour Hospital, Kerman University of Medical Sciences, Kerman, Iran
3. Gastroenterology and Hepatology Research Center, Institute of Basic and Clinical Physiology Sciences, Kerman University of Medical Sciences, Kerman, Iran

Corresponding Author: Mohammad Rezeai Zadeh Rukerd, MD

Gastroenterology and Hepatology Research Center, Institute of Basic and Clinical Physiology Sciences, Kerman University of Medical Sciences, Kerman, Iran

E-mail: Mohammadrezaei75@yahoo.com

ABSTRACT

A 30-year-old man presented with sudden chest pain and evidence of multiple cystic lesions in both long and left-sided pneumothorax in HRCT. H&E stained section and IHC for CD1a and S100 were positive in lung samples. The patient was treated with the diagnosis of Isolated pulmonary langerhans cell histiocytosis.

Keywords: pulmonary langerhans cell histiocytosis, spontaneous pneumothorax, cavitary long lesion

CASE PRESENTATION

A 30-year-old man had sudden chest pain and shortness of breath two hours before admission. He denies any past medical history and family history and there was no history of medication usage, cigarette smoking, or alcohol consumption.

In the physical examination of chest, there were evidences of asymmetrical chest expansion and reduced breath sounds in left side. There were no lesions found during the inspection of the scalp, fingers, and nails.

The high resolution computed tomography (HRCT) scan of the chest revealed widespread broad spread bizarre shape cystic formation in both lungs as well as left-sided pneumothorax (Figure 1). A chest tube was placed on the left side of the chest for the patient. Finally, the patient underwent upper and lower lung lobes wedge biopsy. Hematoxylin and eosin (H&E) stained section and immunohistochemistry (IHC) for CD1a and S100 on both blocks (positive in many of the cyst wall cells) confirmed pulmonary langerhans cell histiocytosis (PLCH). The patient is evaluated in terms of the involvement of the body's other organs. The technetium-99m-methylenediphosphonate ((99m) Tc-MDP) whole body bone scan showed no abnormalities in bone structures. Dynamic pituitary magnetic resonance imaging (MRI) was perform and there was no abnormal signal intensity in pituitary. Finally, the patient was treated with the diagnosis isolated PLCH.

Langerhans Cell Histiocytosis (LCH) is a clonal disorder of langerhans cells, which may affect various organs, such as bones, skin, pituitary gland, and lungs.[1,2]

PLCH is a rare cystic pulmonary disease in adults that occurs almost (more than 95%) in cigarette smokers.[1] Isolated PLCH is seen in around 50% of PLCH patients.[2]

PLCH have several manifestations, one of which is spontaneous pneumothorax, which occurs in about 30-45% of patients, and in 10-30% of patients, spontaneous pneumothorax is the first presentation of PLCH.[2]

As a result, PLHC disease should be evaluated even in adults with no previous medical history and no history of cigarette smoking who have spontaneous pneumothorax and evidence of multiple lung cystic lesions, and other organs should also be checked for LCH involvement.

AUTHOR CONTRIBUTIONS

MSF and FS were involved in patient clinical care. MN and MRZR were involve in literature review, initial manuscript writing, and revision of the manuscript.

FUNDING

None.

CONFLICT OF INTREST

The authors declare that there was conflict no of interest

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy

REFERENCES

1. CIUCHE A, NISTOR C, PANTILE D, MARIN D, TUDOSE A. Spontaneous Pneumothorax in a Case of Pulmonary Langerhans Cell Histiocytosis. Mædica. 2011 Jul;6(3):204–9.

2. Radzikowska E. Pulmonary Langerhans' cell histiocytosis in adults. Adv Respir Med. 2017;85(5):277–89.