

DOI: 10.5152/TurkThoracJ.2019.327

**[Abstract:0643] PP-195 [Accepted: Poster Presentation] [Clinical Problems - Diffuse Parenchymal Lung Diseases]**

## A Case of Pulmonary Langerhans Cell Histiocytosis

Çağla Koç Karaçar, Berat Uslu, Füsün Şahin

Clinic of Chest Diseases, Yedikule Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul, Turkey

**Introduction:** Pulmonary Langerhans Cell Histiocytosis (PLCH) is a rare interstitial lung disease. Most of the cases are young adults between 20 and 40 years of age and it is more common in men than women. PLCH should be considered especially in the differential diagnosis of young patients with a history of smoking and cysts in radiology. Our case is presented because it is a rare and less common disease in women.

**Case Presentation:** A 22-year-old female patient was admitted to our outpatient clinic with cough. The university student had no known additional disease and had a smoking history of 3 pack years. Bilateral small cysts and small nodules were observed on the chest X-ray. Thorax CT revealed thin-walled cystic lesions and millimetric nodules, which were more prominent in bilateral upper and middle lobes. The sputum specimens were negative for AFB smear and culture. PFT results were FVC 3.70 (%89), FEV1 3.25 (%89), FEV1/FVC%86, DLCO 6.88 (%69). On bronchoscopy the right and left lobar and segmental bronchi viewed open and normal, bronchoalveolar lavage was performed in lingula. Bronchoalveolar lavage total cell count was 1100/mm<sup>3</sup>, 15% lymphocyte, 10% neutrophil, 75% macrophage, 0% eosinophil, CD1A 9.92%. Bronchial lavage pathology results were positive for S-100 and CD1a. The patient was diagnosed as PLCH by clinical, radiological and laboratory findings. The patient was recommended to quit smoking and was followed up.

**Conclusion:** Although the etiopathogenesis of PLCH is not clear, it is a rare disease that is closely related to smoking. It is characterized by abnormal proliferation of monoclonal langerhans cells, which is the subtype of dendritic cells, located on epithelial surfaces. HRCT is the basic examination in diagnosis and typically shows a combination of nodules showing the upper and middle zone dominance, cavitated nodules and thick and thin-walled cysts. Although the definitive diagnosis of PLCH is established by pathological examination, it is known that 84-90% of patients are diagnosed correctly with HRCT without biopsy. Therefore, in a patient with smoker and appropriate symptoms, PLCH can be diagnosed in the presence of typical characteristic radiological findings in HRCT. If the radiological pattern is not typical for diagnosis, TBB and BAL are used to diagnose CD1a positive cell counts in both tissue and lavage. The focus of the PLCH treatment regimen is the cessation of smoking. With the cessation of smoking, the disease is symptomatic, radiologically and physiologically stabilized.

**Keywords:** Langerhans cell histiocytosis, smoking, bronchoalveolar lavage