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ABSTRACTS

A Case of Pulmonary Langerhans Cell Histiocytosis Presenting with Radiological Findings Suggestive of Atypical Pneumonia

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Introduction & Purpose: Pulmonary Langerhans Cell Histiocytosis (PLCH) is a rare interstitial lung disease that is typically associated with smoking, predominantly affecting young adults. The clinical and radiological presentation of PLCH can be highly variable, making the diagnostic process challenging. In this study, we aim to present a case of PLCH that initially presented as atypical pneumonia. Sharing the details of the diagnostic and therapeutic process in this case will contribute to a better understanding and increased awareness of this rare disease. In this context, we aim to emphasize the importance of evaluating clinical and radiological findings.

Case: A 22-year-old female patient presented to our outpatient clinic with complaints of cough, weight loss, and night sweats for approximately two months. She had no known medical history. Her family history revealed that her grandfather had died of lung cancer, and a cousin had passed away from an unspecified lung disease. The patient, a student, had a history of smoking 3 pack-years, with no known environmental exposures. She was not on any medication. Bilateral lung sounds were normal, and vital signs were stable. Chest radiography showed reticulations across all zones, more prominently in the upper zone of the right lung (Figure 1). A thoracic CT scan revealed reticulations, ground-glass opacities, scattered infiltrates, and cystic changes across all lung zones (Figure 2). The initial radiological report suggested atypical pneumonia. Blood tests, including a complete blood count and biochemistry, were normal; CRP was 24 mg/dL (Normal: 0-5), and anti-HIV was negative. Despite empirical antibiotic therapy, there was no clinical or radiological improvement Given the initial diagnosis of interstitial lung disease, particularly cystic lung diseases, collagen tissue and vasculitis markers were tested and found to be negative. Abdominal ultrasonography was within normal limits. Bronchoscopy was performed, and bronchial lavage and bronchoalveolar lavage (BAL) samples were collected. Tests for Mycobacterium culture, acid-fast bacillus staining, Mycobacterium PCR, non-specific culture, and fungal culture were all negative. The BAL analysis showed 10% neutrophils, 15% lymphocytes, a CD4/CD8 ratio of 1.4, and a CD1a level of 3%. Following a multidisciplinary evaluation, wedge resection of the upper and lower lobes of the right lung was performed. The pathology report revealed focal areas of scar tissue within the parenchyma, mixed inflammatory proliferation with eosinophils observed in these areas, infiltrates of eosinophilic cells with kidney-shaped nuclei, and cystic formations within the parenchyma. CD10 Clone GM003 Genemed and S-100 were positive. Based on these findings, a diagnosis of Langerhans Cell Histiocytosis was considered.

Discussion and Conclusion: This case demonstrates that Pulmonary Langerhans Cell Histiocytosis (PLCH) can present as atypical pneumonia. The fact that the patient was female and had a relatively short smoking history differs from the typical PLCH cases reported in the literature. The low CD1a level played a significant role in guiding the patient toward surgical intervention. In patients with a high suspicion of Langerhans Cell Histiocytosis, a second bronchoalveolar lavage (BAL) might be considered to confirm the diagnosis if CD1a levels are low. In this case, following multidisciplinary evaluation and discussion with the patient, surgery was decided as the final course of action.

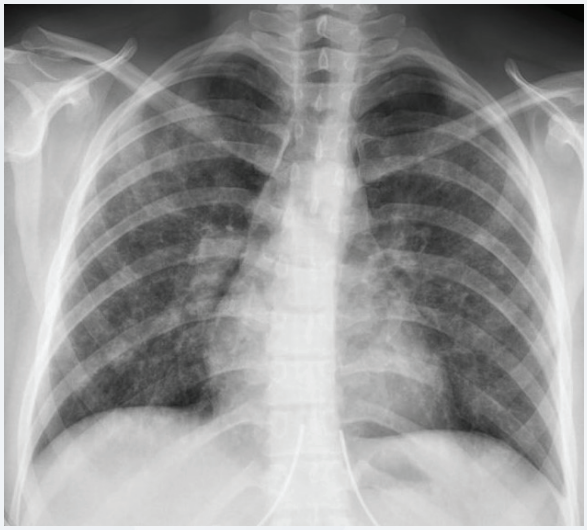

Figure 1. Chest radiograph showing bilateral reticulations, more pronounced in the upper zone of the right lung.

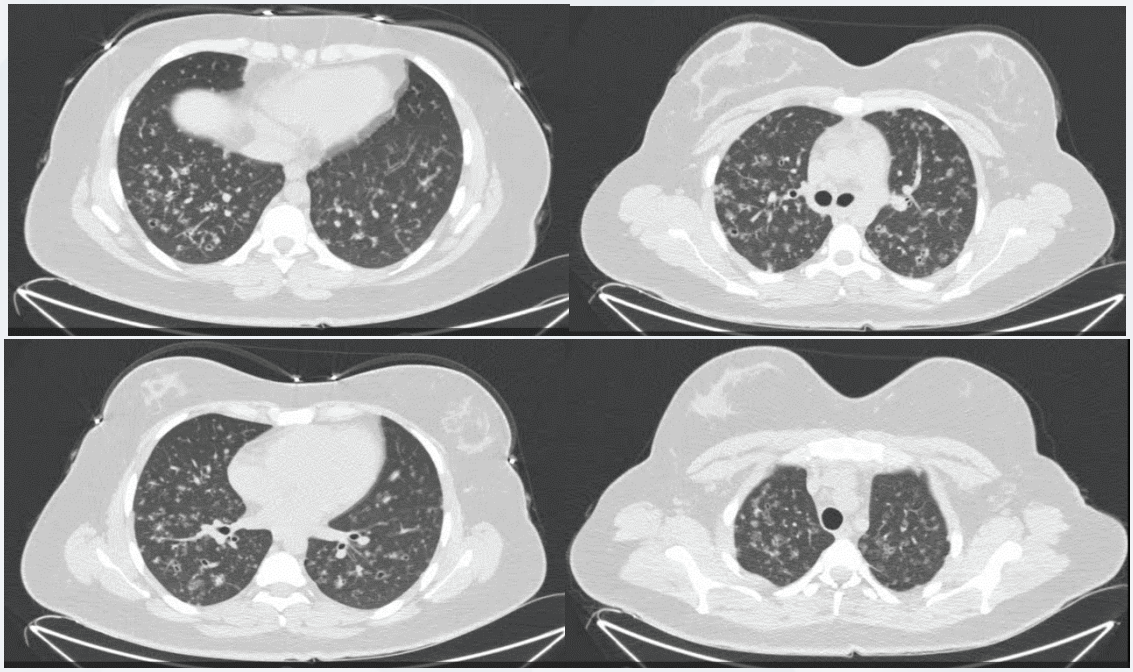

Figure 2. Thoracic CT scan showing reticulations, ground-glass opacities, scattered infiltrates, and cystic changes across all lung zones.

Frequency of Frailty in Interstitial Fibrosis Patients and its Relationship with Prognostic Factors

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Background: Interstitial Lung Diseases (ILD) are a group of diseases characterized by varying degrees of inflammation and fibrosis that cause damage to the lung parenchyma. ILD are a heterogeneous group of diseases. Predicting the risk of mortality or exacerbation in patients with chronic interstitial fibrosis is very difficult. The age, gender and functional physiological capacities of the patients and the extent of radiological fibrosis have been associated with prognosis. Frailty was originally defined as a state of increased vulnerability to stresses resulting from age-related decline in physical reserve and physiological functioning. However, recently it is characterized as "Fragility Syndrome" in other clinical conditions, including chronic diseases, independent of aging. Our aim in this study is to reveal the frailty score level in fibrotic interstitial lung patients and the relationship between these scores and functional parameters (Dyspnea score, FVC, DLCO value, 6-minute walk test result).

Methods: It was designed as a prospective observational study. Patient recruitment has started as of July 2023 and patient recruitment is still ongoing. Interstitial fibrosis patients over the age of 18 who were followed up in our outpatient clinic were included in the study. Demographic data of the patients, comorbidities, MMRc score, FVC and DLCO values, which are functional parameters evaluated during the course of the disease, and 6-minute walk test results were recorded. For frailty measurement, FRAIL frailty index was performed in all patients. In the FRAIL index evaluation, patients were scored according to each parameter (as 1 and 0) and categorized as non-frail (score= 0), pre-frail (score= 1-2) or frail (score= 3-5). Data were entered into the SPSS program and Spearman correlation analysis was performed.

Results: A total of 78 patients, 39 men and 39 women, were evaluated in the study. The average age of the patients was 67 ± 8.6 years. According to the FRAIL frailty scale, 9 patients (11.5%) were evaluated as non-frail, 50 patients (64.1%) as pre-frail, and 19 patients (24.4%) as frail. In respiratory function tests, the mean FVC value was 67.8 ± 19.63 lt, while the mean DLCO value was 60.67± 20.48 ml. No correlation was found between frailty and age ($p= 0.7$). Frailty levels were higher in women than in men ($p= 0.002$). As the frailty level increased, FVC and 6-minute walking distance decreased ($p= 0.04$ and $p< 0.0001$, respectively). As the MMRc dyspnea score increased, the frailty score also increased significantly ($p< 0.0001$). However, no significant correlation was found between DLCO values and frailty levels ($p= 0.19$).

Conclusions: This study showed indirect results showing that frailty assessment in fibrotic lung disease patients, which can be measured by a simple questionnaire, can also be used instead of FVC or 6MWT, which have proven to be effective as prognosis indicators in these patients. These data are preliminary results of the ongoing study. The study will continue to directly demonstrate the relationship between frailty level and prognosis (such as exacerbation and mortality). We think that its value in predicting prognosis will increase as the number of patients increases and follow-up periods increase.

Pulmonary Langerhans Cell Histiocytosis

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Langerhans cell histiyositosis (LHH) is a rare disease characterized by the proliferation of myeloid dentritic cells of the bone marrow. The clinical course is variable and the findings vary depending on the organ system held.

Our case had a history of MDR-TB in the past, and it was followed by continued respiratory distress. He was presented for LHH diagnosis with tests due to persistent shortness of breath. Our case is a 42-year-old man, working in the construction business, who was diagnosed with HRS-resistant tuberculosis five years ago and had been treated with multi-drug-resistant tuberculosis (MDR-TB) for about two years. Then, about five months ago, he was subjected to tube-toracostomy due to pneumothorax, and was discharged without any complication. In the radiology of the patient with recurrent shortness of breath, cysts of the lungs were observed in both lungs, especially in the upper lung areas, in conjunction with the common intra-subpleural air cyst, and cystic lung diseases in the foreground, along with thin centrilobular nodular infiltrations in both lower lobes. Left VATS Lung biopsy resulted in LHH compatibility. Patients with a history of smoking of 30 pack/year were advised to quit smoking first. The patient was under surveillance for 3 months after diagnosis, with no additional clinical complaints.

LHH is mostly one of the lung diseases that do not require any treatment after quitting smoking. Careful differential diagnosis and the proper management of other comorbid conditions are important in cases of comorbide conditions.

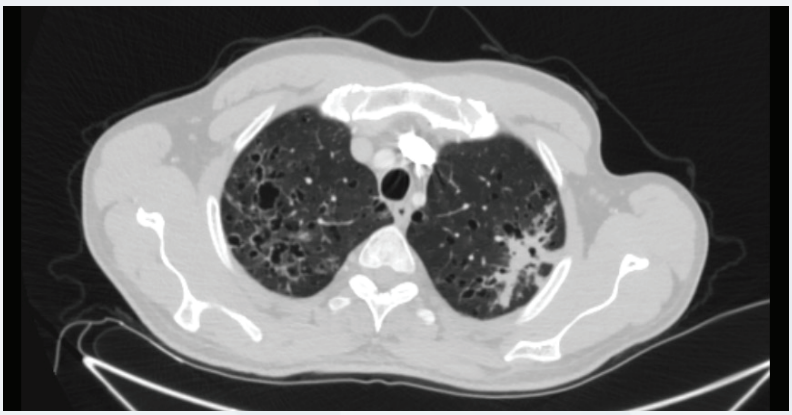

Figure 1. Thorax CT.

Retrospective Evaluation of Patients with Followed Interstitial Lung Disease at Our Hospital Since 2020

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Objective: Interstitial lung disease (ILD) is a chronic condition characterized by widespread inflammation and fibrosis of the lung parenchyma. ILD encompasses many diseases that are similar in clinical, radiological, and pathological features. The etiology of ILD is often idiopathic, but it can be associated with collagen tissue diseases (autoimmunity), granulomatous diseases (sarcoidosis), as well as drugs, infections, environmental factors, and occupational hazards. Patients typically present with symptoms of shortness of breath and persistent cough. The aim of this study is to evaluate the sociodemographic characteristics of 366 patients diagnosed with ILD who are followed and treated at our hospital.

Methods: In our study, we retrospectively evaluated the sociodemographic characteristics, presenting complaints, and radiological features of patients with ILD, whose diagnoses were confirmed pathologically or radiologically, at Afyonkarahisar Health Sciences University Faculty of Medicine Hospital.

Results: A total of 366 patients were included in the study, of whom 178 (48.6%) were female and 188 (51.4%) were male. The average age of the patients was 66.19 ± 13.12 years. Regarding presenting complaints, 345 patients (94.3%) had dyspnea, 238 patients (65%) had cough, 81 patients (22.1%) had sputum, 10 patients (2.5%) had hemoptysis, and 13 patients (3.2%) had chest pain. Evaluating posteroanterior chest X-rays, 157 patients (42.9%) had peripheral and lower zone opacities with coarse reticular, reticulonodular patterns, and areas of consolidation. According to the ILD subclassification, 162 patients (44.3%) had undifferentiated ILD, 94 patients (25.7%) had idiopathic pulmonary fibrosis, and 61 patients (16.7%) had sarcoidosis. Ethologically, 343 patients (93.7%) had negative immunological markers, while only 23 patients showed positive immunological markers. In the assessment of pulmonary function tests (PFT), 14 patients (3.8%) had obstructive-type respiratory failure, 67 patients (18.3%) had restrictive-type respiratory failure, and 119 patients (33.7%) had mixed-type respiratory failure. PFT was normal in 153 patients (41.8%), and 13 patients were unable to cooperate with the PFT.

Conclusion: ILD causes progressive dyspnea and reduces the quality of life. There is no medical treatment that can completely cure the disease. Therefore, early diagnosis and treatment are crucial. In patients presenting with dry cough and progressive dyspnea, if radiological findings show reticulonodular appearance and PFT indicates restrictive findings, ILD should be considered.

Keywords: Interstitial lung disease, progressive dyspnea, reticulonodular opacity.

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