DOI: 10.5152/TurkThoracJ.2019.319

[Abstract:0160] PP-166 [Accepted: Poster Presentation] [Clinical Problems - Diffuse Parenchymal Lung Diseases]

IPF or NSIP? An Interesting Case

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Introduction: Idiopathic pulmonary fibrosis (IPF) is a chronic and progressive interstitial lung disease. The presence of typical high resolution computed tomography (HRCT) findings for radiologically definite usual interstitial pneumonia (UIP) and the absence of risk factors leading to fibrosis are sufficient for the diagnosis of IPF in clinically compatible patients. However, sometimes HRCT findings cannot be consistent with UIP. In this case, histopathological evaluation is required.

Case Presentation: 50-year-old female patient. She has been suffering from shortness of breath and cough for 2 years. She was diagnosed with non-specific interstitial pneumonia (NSIP) and received steroid treatment for 1 year. She is housewife and feeding cows. She never smoked. Previously she was feeding chicken. In physical examination, there are velcro rales in bilateral lower areas and clubbing finger. She described reflux. One year ago (2017) thoracic CT revealed ground glass images, increased reticular density and traction bronchiectasies consistent with interstitial lung disease and was recommended investigate for NSIP. Thoracic CT of 2018 after steroid treatment showed reticular density, traction bronchiectasis, ground glass opacity and small honeycomb cysts in basal areas in both lungs with minimal progression compared to previous examinations (NSIP? possible IPF?). The cytological examination and direct cell counts of bronchoalveolar lavage were dominated by neutrophilia (65%) and procalcitonin levels were normal. Biopsy was performed by open lung method. Histopathologically, it was found to be compatible with UIP. The collagen tissue panel of the patient was negative. No collagen tissue disease (CTD) was detected by the rheumatology clinic. Antifibrotic therapy (pirfenidone) was started with the diagnosis of IPF. At the 6th month of the treatment, minimal improvement in respiratory function and regression of the patientıs symptoms were observed.

Conclusion: The diagnosis of IPF is based on the exclusion of other causes of UIP (CTD, chronic hypersensitivity pneumonia, drug use, etc.) as clinical and radiological appropriate patients. However, atypical manifestations can be observed radiologically. In this case, the work of clinicians who are confronted with the patient becomes difficult. We wanted to share an interesting case which was diagnosed with histopathological findings as definite UIP and received steroid therapy for 1 year.

Keywords: IPF, NSIP, interstitial lung disease