DOI: 10.5152/TurkThoracJ.2019.46

[Abstract:0379] MS-061 [Accepted: Oral Presentation] [Clinical Problems - Diffuse Parenchymal Lung Diseases]

Comparison of Serum SP-A, SP-D, KL-6 Levels, Echocardiographic, Functional and Radiological Features in CPFE and IPF Patients

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Objectives: Combined pulmonary fibrosis and emphysema (CPFE) has been recognised as an phenotype of pulmonary fibrosis that is characterised by upper lobe emphysema and lower lobe fibrosis. Surfactant protein-A (SP-A), surfactant protein-D (SP-D) and Krebs von den Lungen-6 (KL-6) KL-6 are reported to be sensitive markers for interstitial lung diseases. Our primary outcome were to compare serum SP-A, SP-D, KL-6 levels in patients with CPFE and idiopathic pulmonary fibrosis (IPF) without emphysema. Secondary outcomes were to determine and compare the functional parameters, transthoracic echocardiographic and radiological findings.

Methods: The patients who had been diagnosed by 'CPFE' and 'IPF without emphysema' are included in this study as two groups. The patients with connective tissue diseases are excluded. Demographic features, functional parameters, echocardiography (ECHO) findings and measurement of serum SP-A, SP-D and KL-6 compared in two groups.

Results: Forty-seven patients who had been diagnosed with CPFE and IPF without emphysema by their CT scan image, were prospectively enrolled in this study. The study population comprised 47 patients (41 male, 6 female) with a mean SD age of 70.12±8.75 years. CPFE patients were older, had lower diffusion capacity, force vital capacity (FVC, %), worse six minute walking distance (6MWD) than IPF group. Comparison of demographic, clinical and radiological features are summarized. When we compared CPFE and IPF group, although we found higher serum KL6 levels in CPFE group, there was no difference between serum SP-A and SP-D levels. Cardiovascular evaluation revealed patients with CPFE had significantly more comorbidities such as hypertension. BNP, a biomarker secreted in response to ventricular dysfunction and wall stretch indicating hearth failure, is significantly higher in CPFE patients. While left ventricular systolic (ejection fraction) and diastolic functions (mitral e/a ratio) showed no difference between groups, right ventricular diameter tended to be larger in CPFE group. Tricuspid annular plane systolic excursion (TAPSE), an important parameter for right ventricular function, is tended to be better in IPF patients. Although statistically non-significant, right ventricular tissue doppler ECHO revealed worse muscular function in systolic (s') phase in CPFE patients. Pulmonary artery pressure wasn't significantly different between the groups.

Conclusion: CPFE is an increasingly recognized condition and seems to be related worse pulmonary functional parameters than in IPF patients. Higher KL-6 levels is important to characterize the role of KL-6 in CPFE pathogenesis.

Keywords: Combined pulmonary fibrosis emphysema, idiopathic pulmonary fibrosis, SP-A, SP-D, KL6, echocardiography