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## Familial Idiopathic Pulmonary Fibrosis Case Series

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**Introduction:** Idiopathic pulmonary fibrosis (IPF) is a type of interstitial pneumonia with chronic, progressive fibrosis, and is usually seen in adults. Familial IPF is defined by the presence of at least two cases of pulmonary fibrosis in the same biological family. Familial IPF constitutes 2-20% of all IPF patients. In this case series, 3 brothers with IPF will be presented.

**Case 1:** A 58-year-old male patient was admitted with the complaints of shortness of breath and cough for five years in 2014. He had a smoking history of 40 pack/years. He had no occupational exposure. His physical examination revealed bilateral diffuse rales and clubbing on the fingers. In pulmonary function test (PFT) FEV1 was 1.35L (46%) FVC was 1.60L (44%) and FEV1/FVC was 84%. He was diagnosed with IPF clinically and radiologically as his high resolution computed tomography (HRCT) showed bilateral honeycombing, interseptal thickening. Pirfenidone treatment was started. There was significant progression in his clinical status, chest X-ray and PFTs (FEV1 1.19L 41% FVC: 1.44L 40% FEV1/FVC: 83%) in six months follow-up. The patient died of myocardial infarction ten months after diagnosis.

**Case 2:** The first patient's 60 years old brother was admitted to our outpatient clinic with complaints of cough and exertional dyspnea in March 2015. There were 35 pack/years smoking history. Physical examination revealed velcro rales in the bases of lungs. FEV1 in PFT was 2.21L 76%, FVC was 2.69L 74%, FEV1/FVC:82% and DLCO: 15.7 63%, DLCO/VA:4.07 97% and 6 min walking distance (6MWD) was 540 meters. HRCT showed bilateral interseptal thickening and fibrosis findings. The patient was diagnosed with IPF by lung biopsy in May 2015. The patient is still under follow-up and has been on pirfenidone treatment for about three years.

**Case 3:** A 65 years old third brother presented to our outpatient clinic with an exertional dyspnea on September 2018. The patient had a smoking history of 26 packs/years. PFT revealed FEV1: 2.79L 90% FVC: 3.01L 80%, FEV1/FVC: 87% DLCO: 5.85 65% DLCO/VA: 1.26 95% 6MWD 525 meters. HRCT showed bilateral subpleural early fibrosis, interlobular septal thickening and minimal honeycomb appearance. Rheumatological examination and immunological tests were negative. The patient was diagnosed as IPF clinically and radiologically and pirfenidone treatment was started. In the third month of the treatment, the patient is being followed up in our clinic.

**Conclusion:** Familial IPF is a rare condition. As in our cases, family history of all diagnosed patients should be questioned in terms of IPF.

**Keywords:** Familial idiopathic pulmonary fibrosis, IPF cases, IPF and genetics