*Abstracts of the Scientific Awards of XXXIX COMU 2020 - Research Classified - Panels Award - Case Report*

# Familial Pulmonary Fibrosis - a Case Report

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**Introduction**: Idiopathic pulmonary fibrosis (IPF), a chronic progressive idiopathic interstitial pneumonia (IIP), occasionally, IPF occurs in families. IPF is associated with an estimated survival of 20%–50% at 5 years. Familial pulmonary fibrosis (FPF) can be defined as the occurrence of at least two cases of some type or fibrous interstitial pneumonia in members of the same biological family. Is important to note that the forms of interstitial lung diseases that affect relatives not necessarily need to be the same. Adults with FPF are essentially indistinguishable from patientes with sporadic IPF in terms of clinical presentation, radiographic findings and histopathology, except that those with FPF tend to present it earlier.

**Objective(s):** To describe the clinical, functional, and radiological features of index cases of familial pulmonary fibrosis (FPF).

**Case overv**iew: Male, 72 years old, seeks pulmonology service in October 16th, 2019 with cough, sputum and sinus pain. Denied dyspnea or other respiratory symptoms. Non-smoker, no alcoholism history, related physical activity 3 times per week and no environmental exposures. Treatment for sinusitis was indicated with return in 2 weeks. On return, he was asymptomatic. Unfortunately, he said that his 74-year old middle brother was hospitalized with severe dyspnea, mMRC 4, SpO2 91% using oxygen 4L in cateter and was diagnosed with non-specific interstitial pneumonia (NSIP). The CT scan revealed a tomographic pattern of NSIP, with a fibrous evolution form and emphysema. Also, he has a familiar history of an older brother who died 5 years ago, at the age of 80, with pulmonary fibrosis, and a CT scan usual intersticial pneumonia (UIP) pattern. The pulmonary auscultation had: a preserved vesicular murmur, mild bilateral "velcro-like" crackles in pulmonary basis, heard during middle to late inspiration, no wheezes. The patient didn't accept to be submitted to invasive procedures, as bronchoscopy or surgical lung biopsy, because didn't have respiratory symptoms (although the importance of these exams has been explained). The diagnosis of familial pulmonary fibrosis can be established when, despite having different tomographic patterns, siblings have in common the fibrous feature, as in this case. Even if the younger brother doesn't have fibrosis yet, with the diagnosis confirmed by the other two brothers, the antifibrotic medication should be started.

**Keywords:** Familial pulmonary fibrosis; Interstitial lung diseases; Antifibrotic.

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