Combined Sarcoidosis and Idiopathic Pulmonary Fibrosis (CSIPF): Coincidence, Association or Mimicry?

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Clinical Image

A 70-year old patient with longstanding history of sarcoidosis (initially ATS- stage II), presented with progressive dyspnea and dry cough. Although corticosteroid therapy initially improved lung function, a decline (Figure 1) was observed. Thoracic CT-scan was compatible with definite unusual interstitial pneumonia potentially engrafted on the preexisting sarcoidosis. After multidisciplinary discussion antifibrotic therapy with Nintedanib was started. Lung function and fibrosis on the CT-scan improved (Figure 1, 2). Collins et al. [1] found a series of cases and coined the term “Combined Sarcoidosis and Idiopathic Pulmonary Fibrosis CSIPF”. IPF is defined as fibrosis without inflammation and sarcoidosis as a multisystem chronic inflammatory condition – these

Figure 1: Lung function revealing a progressive restrictive ventilatory defect and reduced diffusion capacity improving after antifibrotic therapy with nintedanib.

Figure 2: Calcified and enlarged hilomediastinal lymph nodes alongside with progressive basal and subpleural fibrosis between 2018 and 2019. Improvement of the progressive fibrosis after antifibrotic therapy in 2021.
supposedly being two distinct entities [2,3]. Did we experience coincidence, association or mimicry between these two entities? Lucky enough antifibrotic treatments is also licensed for progressive non-IPF fibrotic lung diseases.

References