A Curious Case of Disappearing Pulmonary Fibrosis?

I. S. Tora, A. Shambhu; Internal Medicine, Inova Fairfax Medical Center, Annandale, VA, United States.

Corresponding author's email: ibrahim24@gmail.com

CASE: A 73-year-old male presented for evaluation of an insidious onset progressive dyspnea and a dry cough starting 9-months prior. He had a CT scan of his chest with his local pulmonologist showing findings suggestive of interstitial lung disease. Notably, he had experienced some relief after being treated with steroids, but symptoms returned after steroids were stopped. He is a former smoker with a 60-pack year history having quit 9 years ago. He lived in a rural area but denied significant exposures aside from Agent Orange in Vietnam remotely. He does not have a personal or family history of connective tissue disease (CTD). His physical exam was notable for normal vitals and bibasilar crackles on lung auscultation, but he had no digital clubbing or stigmata of CTD. Serologies for CTD were negative. Pulmonary function tests revealed mild obstruction without restriction and a moderately reduced diffusion capacity. Six-minute walk test results showed normal distance without dyspnea. A high-resolution computed tomography (HRCT) demonstrated a pattern consistent with probable usual interstitial pneumonia (UIP) but with more ground glass opacities than expected. He had a surgical lung biopsy which showed fibrosing interstitial pneumonia with geographic and temporal heterogeneity consistent with UIP, although microscopic honeycombing was not identified; moreover, patchy organizing pneumonia and mild chronic pleuritis were noted. He was diagnosed with idiopathic pulmonary fibrosis (IPF) by a multi-disciplinary conference, and recommendation was to start antifibrotic while maintaining immunosuppression to address organizing pneumonia. He was started on mycophenolate and high dose prednisone while awaiting approval for nintedanib. Three months later, his cough had resolved and HRCT showed only upper lobe centrilobular emphysema with interval resolution of ground glass opacities as well as his interstitial infiltrates.

DISCUSSION: The patient in this case was found to have histologic and radiographic evidence of fibrotic lung disease meeting the diagnostic criteria for idiopathic pulmonary fibrosis. Ground glass opacities on HRCT and organizing pneumonia seen on biopsy prompted a trial of immunosuppressive treatment with significant improvement in not only the organizing pneumonia but also the interstitial infiltrates. This case shows that despite a multidisciplinary diagnosis of IPF, clinicians should consider judicious immunosuppression in select cases when significant inflammation is present. This is especially important with newer data from the SENCIS and INBUILD trials showing benefit of nintedanib in non-IPF ILDs with significant fibrosis prompting more widespread use of antifibrotics.