Cough Disguised as Airway Centered Interstitial Fibrosis

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Introduction: Airway centered interstitial fibrosis (ACIF) is a rare form of interstitial lung disease (ILD). ACIF has been seen in patients who have hypersensitivity pneumonitis, gastroesophageal reflux disease (GERD), and connective tissue diseases. Studies have also found it associated with inhalational exposures. It is characterized by small airway centered interstitial fibrosis and metaplastic bronchiolar epithelium encompassing fibrotic bronchioles. Case Description: A 44-year-old female with a past medical history of GERD presented to the pulmonary office with chronic productive cough for the past 4 years with worsening dyspnea on exertion. She previously worked in the textile industry and housekeeping. She indicated that she was an obsessive cleaner having chronic exposure to multiple household cleaning products. She also had two sisters who died at a young age from systemic lupus erythematosus. She was initially treated with a proton pump inhibitor for a cough secondary to GERD however her symptoms did not improve. She had a computed tomography (CT) of the chest which revealed bronchiectasis at the bases of the lungs bilaterally concerning for early ILD. Pulmonary function testing was remarkable for a decreased diffusion capacity for carbon dioxide. Bronchoscopy was unfortunately nondiagnostic. Rheumatologic workup was remarkable for a positive anti-nuclear antibody (ANA) however antibodies for all other connective tissue diseases were negative. She underwent an open lung biopsy which revealed fibrosis with prominent peribronchial metaplasia, lymphoid hyperplasia, and chronic pleuritis consistent with ACIF. She was started on prednisone and mycophenolate and reduced her exposure to household cleaning products with improvement in her symptoms. Discussion: Given our patient’s history of GERD, positive ANA, and inhalational exposures, she had many risk factors for developing ACIF. ACIF is typically seen in females with a mean age of 57 years old however our patient presented at a slightly younger age. The progression of ACIF is variable but it has been found to have a better prognosis than idiopathic pulmonary fibrosis. Overall, it is important to have a high index of suspicion for ACIF in patients who present with a history of GERD, inhalational exposures, or connective tissue diseases. Diagnosis often requires surgical lung biopsy but prompt treatment for these individuals could improve their long-term prognosis.

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