

Interstitial Lung Disease with Autoimmune Features Successfully Treated with Mycophenolate Mofetil: Case Report

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Keywords: Interstitial Lung Disease, Mycophenolate Mofetil, Autoimmune, Autoimmune Diseases

Published: 14 December 2023

Introduction: Interstitial lung disease (ILD) is a broad term used to describe a group of lung disorders characterized by fibrosis of the lungs. ILD is classified into known factors including occupational and environmental exposures, as well as idiopathic cases. ILDs can be associated with connective tissue disease including systemic sclerosis, rheumatoid arthritis, systemic lupus erythematosus, or limited autoimmune features¹. Patients will often present with dyspnea and non-productive cough. Mycophenolate mofetil (MMF) is an immunosuppressive drug that has shown to be well tolerated in ILD patients and restore pulmonary function². Efficacy is not established yet in ILD with autoimmune features.

Case Presentation: Patient is a 58-year-old male who presented with shortness of breath at rest. Medical history was significant for hypertension and chronic obstructive pulmonary disease (COPD). His pulmonary function testing shows diffusing capacity of the lungs for carbon monoxide (DLCO) of 30 (normal > 70%), forced vital capacity (FVC) of 67% of age predicted, and forced expiratory volume (FEV1) of 66% (normal between 70-80%). Initial CT scan performed demonstrated mediastinal lymph nodes with upper lobe fibrotic changes. His lung biopsy showed inflammation and evidence of non-specific ILD (NSIP). Laboratory results were positive for anti-nuclear antibodies (ANA) and elevated erythrocyte sedimentation rate (ESR) while negative for other immunological markers. The patient did not have any known occupational exposure. On physical exam, he had evidence of nail clubbing. The patient met criteria for diagnosis of interstitial lung disease with autoimmunity. He was started on treatment with 1000 mg of MMF twice daily as well as oral prednisone 2.5 mg daily. The patient was found at follow up appointments to be responding well to treatment.

Discussion: The case highlights a unique presentation of ILD in a patient with features of an autoimmune process including a positive ANA titer. Although there is not much data in the literature to delineate treatment of ILD with autoimmune features, we believe this case may support use of MMF in larger clinical trials.

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