The Undetected Myopathy: Anti-PL-12 Antisynthetase Syndrome

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Introduction

ANTISYNTHETASE SYNDROME

- A rare autoimmune condition that occurs when autoantibodies are formed in the body against aminoacyl-transfer ribonucleic acid (tRNA) synthetases
- Usually presents in the form of interstitial lung disease (ILD), myositis, or arthritis
- Anti-histidyl tRNA synthetase or anti-Jo-1 is the most commonly seen aminoacyl-tRNA synthetase
- Antisynthetase syndrome secondary to other autoantibodies are less common

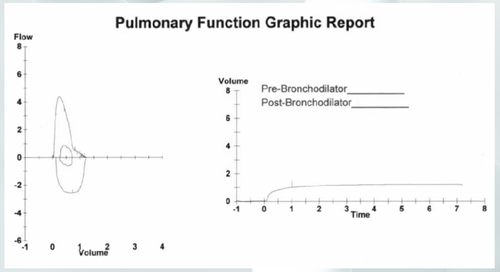

Figure 1

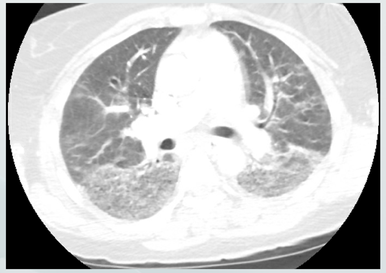

Figure 2

Case Description

- 47-year-old female with no significant past medical history found to have multiple admissions for recurrent pneumonia in the setting of joint pain in her hands
- Pulmonary function tests (PFTs) revealed severe restriction (Figure 1), and computed tomography (CT) of the chest demonstrated bilateral ground glass and reticular opacities (Figure 2)
- Diagnosed with ILD which was believed to be secondary to rheumatoid arthritis given her bilateral hand pain, and was started on immunosuppressants
- Later developed a recurring rash, Raynaud phenomenon, and diffuse myalgias
- Presented to the pulmonology office five years later where PFTs showed severe restriction and severely reduced diffusion capacity
- Repeat CT Chest revealed worsening ground glass and reticular opacities with traction bronchiectasis
- Upon further review of her prior ILD evaluation, her rheumatologic workup revealed a positive anti-nuclear antibody with a spindle and speckled pattern
- Myositis panel was negative for anti-Jo-1 antibody, but positive for anti-PL-12
- Hypersensitivity pneumonitis panel, angiotensin converting enzyme, vasculitis panel, and infectious workup were all unremarkable
- Subsequently diagnosed with anti-PL-12 induced antisynthetase syndrome and started on rituximab

Conclusion

- Our patient's initial diagnosis of antisynthetase syndrome may have been delayed due to her anti-Jo-1 autoantibody being negative and her initial lack of other symptoms
- Further myositis workup later revealed a positive anti-PL-12 autoantibody which is rare
- Anti-PL-12 autoantibody has a strong association with ILD predominant disease
- ILD associated with anti-PL-12 autoantibody tends to be more severe which places these patients at higher risk for mortality
- Ten-year survival rate for individuals who have antisynthetase syndrome due to an aminoacyl-tRNA synthetase other than anti-Jo-1 is much lower
- It is crucial to test for all of the aminoacyl-tRNA synthetases when you have a suspicion for antisynthetase syndrome given that anti-Jo-1 may not always be positive in these patients
- It is essential to have a high index of suspicion for antisynthetase syndrome when treating patients with a progressive ILD in the setting of myalgias, Raynaud phenomenon, and recurring rash.

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