Anti-Synthetase Syndrome Presenting as Rapidly Progressive Interstitial Lung Disease

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Anti-Synthetase Syndrome Presenting as Rapidly Progressive Interstitial Lung Disease

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Introduction

Anti-Synthetase Syndrome is a rare disease seen most commonly in patients with auto-immune inflammatory myopathies such as dermatomyositis. Typical characteristics of this exceedingly rare syndrome are arthropathy, interstitial lung disease (ILD), the presence of antibodies against amino-acyl tRNA synthetases, and rarely the typical myositis associated with these antibodies. We describe the unfortunate case of a patient with rapidly progressive ILD and acute hypoxic respiratory failure necessitating bilateral lung transplantation.

Case presentation

A 47-year-old female with past medical history significant for hypothyroidism and hyperlipidemia presented to the office with one month of progressively worsening dyspnea and productive cough; her initial chest radiography showed bilateral, multi-focal infiltrates. A bronchoalveolar lavage and transbronchial and endobronchial biopsies were performed via bronchoscopy without significant findings. Extensive infectious work-up was negative. Only a few days later, she was admitted to the hospital with severe hypoxemia. Computed tomography of the chest without contrast demonstrated worsening bilateral airspace disease and rapidly progressing traction bronchiectasis (Images 1 and 2). Video-assisted thorascopic surgery (VATS) was performed and wedge biopsy demonstrated alveolar tissue containing fibrin with surrounding cellular fibrosis. Extensive immunological work-up returned positive for the anti-histidyl-tRNA synthetase antibody, Anti-Jo-1, highly suggestive of a diagnosis of Anti-Synthetase Syndrome. She was initiated on pulse dose steroids and transferred to a transplant center where she received bilateral lung transplantation.

Imaging

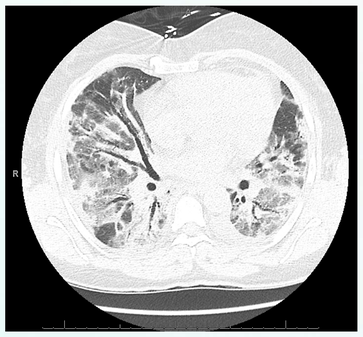

Image 1. Diffuse airspace disease with traction bronchiectasis.

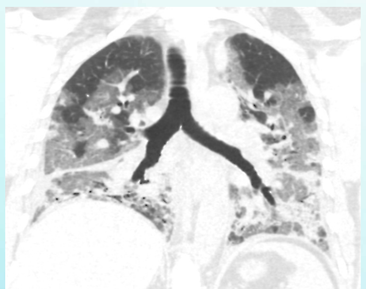

Image 2. Coronal view of patient's extensive lung disease.

Discussion

- Anti-Synthetase Syndrome is a rare auto-immune disease that has significant systemic manifestations.
- Commonly accepted diagnostic criteria for Anti-Synthetase Syndrome include the presence of an anti-aminoacyl tRNA synthetase antibody and evidence of ILD, Raynaud's phenomenon, arthritis, or mechanic's hands.
- While this disease falls within the clinical realm of the inflammatory myopathies, clinicians must be aware that this syndrome can and often does present primarily with rapidly progressing ILD, being the primary mediator of morbidity and mortality in these patients.
- Clinical suspicion for autoimmune disease should be high, particularly in patients with rapidly deteriorating respiratory function without evidence of active infection.

Conclusion

- This case demonstrates the importance of expeditious and accurate identification of autoimmune etiologies in patients presenting with respiratory decline.
- It is critical to eliminate treatable infectious etiologies to allow for initiation of immunosuppressive therapy.
- A low threshold for evaluation for transplant should also occur with rapidly progressing ILD associated with autoimmune features or serologies.

REFERENCE

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