A Case of Antisynthetase Syndrome Masquerading as Recurrent Pneumonia

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BACKGROUND

- Antisynthetase syndrome (ASS) is a rare autoimmune disorder characterized by the classic triad of myositis, arthritis, and interstitial lung disease (ILD).
- Diagnosis remains a challenge given its rarity and highly variable clinical presentation.

PRESENTATION

- A 56-year-old female with history significant for hypothyroidism and tobacco dependence presented with recurrent dyspnea, cough, and polyarthralgia.
- She has had multiple ED visits and hospitalizations, during each time she was diagnosed with multifocal pneumonia on imaging and treated with antibiotics. On her last hospitalization, she also presented with polyarthralgia, and autoimmune workup was nonrevealing. She was treated with steroids. Her symptoms initially improved however worsened at the end of the steroid taper, and she represented and hospitalized for the third time.

PHYSICAL EXAMINATION

Temp 97.4F, HR 92, BP 142/90, RR 21, SpO2 95% on 2L NC

- Unable to speak in full sentences, with bibasilar rales and wheezing on lung auscultation.
- Tenderness on palpation of bilateral DIP, PIP, and MCP, with swelling and limited ROM. Thickened, fissured, and scaly skin was noted on the digits of both hands.

DIAGNOSTIC STUDIES

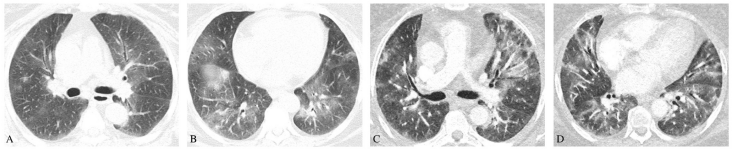
Initial workup:

Autoimmune workup from prior admission:

Creatine kinase	646 U/L
C-reactive protein	6.79 mg/dL
Sedimentation rate	71mm/hr
Antinuclear antibody	Negative
Cyclic citrullinated peptide	Negative
Rheumatoid factor	Negative

Infectious workup:

Blood culture	(-)
Sputum culture	(-)
Urine streptococcal antigen	(-)
Urine legionella antigen	(-)
Lyme IgG/IgM	(-)
COVID-19 PCR	(-)



CTPA: worsening scattered ground-glass opacities (C&D) compared to previous CT studies (A&B).

Further investigation:

Bronchoscopy with BAL: benign lung tissue, focal mild fibrosis, and chronic inflammation with intra-alveolar pigmented macrophages, mild anthracosis, and reactive pneumocytes, consistent with nonspecific interstitial pneumonia.

Further autoimmune workup:

Myositis panel: anti-Jo antibody	(+)
MDA-5 antibody	(-)
Hypersensitivity pneumonitis panel	(-)
Antineutrophil cytoplasmic antibody	(-)
Anti-scleroderma-70 antibody	(-)
RNA polymerase antibody	(-)
Anti-dsDNA antibody	(-)
Anti-smith antibody	(-)
Anti-SSA antibody	(-)
Anti-SSB antibody	(-)

CLINICAL COURSE

- During the early hospital course, antibiotics were started and discontinued after a negative infectious workup. She received IV steroids for presumed ILD while awaiting the results of most autoimmune workup.
- Patient was diagnosed with ASS-related ILD and the decision was made to initiate inpatient immunosuppressive therapy. She received a five-day course of IVIG at 2g/kg and methylprednisolone 1g for three days followed by a taper.
- She had significant clinical improvement and was eventually discharged with mycophenolate mofetil and steroids taper.

DISCUSSION

- ASS-related ILD presents with rapid onset and progression, which could often be confused with other more common acute processes such as pneumonia, especially when ILD can be the sole manifestation.
- Clinicians should meticulously assess for subtle signs and perform specific autoantibody testing when evaluating patients with a suspicion for undifferentiated autoimmune conditions involving the lungs, muscles, and joints in isolation or combination.
- A multidisciplinary approach involving pulmonology and rheumatology may lead to better recognition and management of this syndrome.

Connors et al., 2010[1]	Solomon et al., 2011[2]

Antisynthetase antibody plus one or more of the following:
• Myositis by Bohan and Peter criteria[3]
• ILD not explained by other causes
• Arthritis
• Unexplained, persistent fever
• Raynaud's phenomenon
• Mechanic's hands
	Antisynthetase antibody plus two major criteria or one major and two minor criteria.
	• Major criteria:
	  ○ Myositis by Bohan and Peter criteria[3]
	  ○ ILD not explained by other causes
	• Minor criteria:
	  ○ Arthritis
	  ○ Raynaud's phenomenon
	  ○ Mechanic's hands

Table 1: the two classification criteria for antisynthetase syndrome. ILD= interstitial lung disease

REFERENCES

1. Connors GR, Christopher-Stine L, Oddis CV, Danoff SK. Interstitial lung disease associated with the idiopathic inflammatory myopathies: what progress has been made in the past 35 years? Chest. 2010;138(6):1464-1474.
2. Solomon J, Swigris JJ, Brown KK. Myositis-related interstitial lung disease and antisynthetase syndrome. J Bras Pneumol. 2011;37(1):100-109.
3. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med. 1975;292(7):344-347.