A CHALLENGING DIAGNOSIS OF CELLULAR NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP) WITH ORGANIZING PNEUMONIA IN A PATIENT WITH ANTI-JO 1 NEGATIVE DERMATOMYOSITIS

Namratha Shripad, Nawal Ahmed, Taikchan Lildar, David M. Wisa
Flushing Hospital Medical Center, Flushing, NY, 11355 - USA

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Introduction :

- Nonspecific interstitial pneumonia (NSIP) is an interstitial lung disease (ILD) that can be idiopathic or associated with connective tissue disorders.
- The two subtypes of NSIP are cellular interstitial pneumonitis (CIP) and fibrotic, with CIP being less common.
- The subtypes can be distinguished by pathology and imaging. NSIP constitutes 14-36% of idiopathic interstitial pneumonia cases.
- Dermatomyositis associated with ILD has higher mortality, making diagnosis crucial. In specific, fibrotic NSIP has a high 10-year mortality rate, making differentiation relevant.
- We present a unique case of cellular NSIP with organizing pneumonia associated with dermatomyositis.

Case Description:

- A 36-year-old female presented with fatigue and erythematous papular lesions on her face, palms, shoulder, and neck for one month. She also developed a fever and dry cough a week before. She denied recent travel or sick contacts. COVID-19 was negative.
- On **exam**, she was tachypneic and tachycardic with a maculopapular rash. A pulmonary exam revealed bilateral fine crackles. CXR showed patchy dense consolidations on the left and mild right side.
- **Labs** revealed **(Table 1)** elevated inflammatory markers (ESR, CRP, LDH, and CPK). CBC and procalcitonin were normal. The chest CT **(Figure 1)** showed extensive patchy and confluent areas of opacification of the left lower lobe, including a mass-like area measuring 3.3 cm. Infectious workup and autoimmune testing (ANA, anti-CCP, anti-Jo-1, anti-SCL70, and anti-centromere Abs) were negative.
- **Bronchoscopic left lower lobe biopsy** showed cellular interstitial inflammation composed of lymphocytes, plasma cells, rare eosinophils, and foci of intra-alveolar fibrinous exudates, suggestive of CIP and organizing pneumonia.
- She was treated successfully with **corticosteroids** and was discharged on **prednisone**. Repeat autoimmune antibody workup was negative.
- The **skin biopsy** showed lichenoid lymphocytic infiltrates and necrotic keratinocytes consistent with dermatomyositis. **Mycophenolate** and **rituximab** were initiated; prednisone was tapered off. Follow-up chest CT **(Figure 2)** showed cleared infiltrates with symptomatic improvement.

Table 1: Laboratory Workup

Inflammatory Markers		
Erythrocyte Sedimentation Rate (ESR)	63 [0 - 25 mm/hr]	
C-Reactive Protein (CRP)	1.9 [<1.0 mg/dL]	
LDH	982 [313 - 618 U/L]	
Creatine Kinase Total (CPK)	517 [30 - 135 U/L]	
Auto-Antibody		
Antinuclear Antibodies (ANA)		Negative
Anti-CCP (Cyclic Citrullinated Peptide Ab, IgG)		<20 [<3.0 U/mL]
Sjogren's Ab	Anti-SSA	<0.2 [<1.0 AI]
	Anti-SSB	<0.2 [<1.0 AI]
Anti-Jo-1 Ab		<0.2 [<1.0 AI]
Anti-SCL70		<0.2 [<1.0 AI]
Centromere Ab, IgG		<0.2 [<1.0 AI]
Viral Panel		
HIV	Negative	
Hep A IgM	Negative	
Hep B Core IgM	Immune-Protected	Nonreactive
Hep B Surface Ag		Negative
Hep B Surface Ab		194 mIU/mL
Hep C Ab	Nonreactive	
Serology		
Lyme Ab	Negative	
*R. rickettsii*lg G, IgM	Not Detected	
Legionella Ag (Urine)	Not Detected	

Figures:

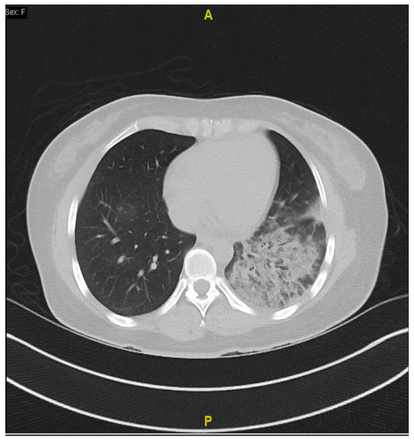

Figure 1: Computed tomography of the chest without contrast ***on admission*** showing extensive patchy and confluent areas of opacification.

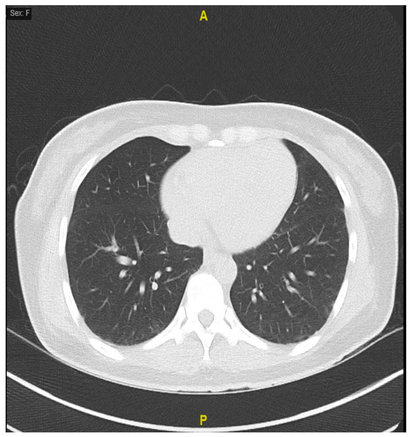

Figure 2: Repeated computed tomography of the chest without contrast done ***8-months later*** following treatment and resolution of symptoms.

Discussion:

- CIP is an uncommon form of NSIP. On chest CT, bilateral ground-glass opacities are the most common feature. CIP is characterized histologically by interstitial thickening due to the presence of inflammatory cells and type-II pneumocyte hyperplasia with preserved lung architecture.
- Treatment for ILD is corticosteroids. The prognosis is excellent. ILD associated with dermatomyositis is strongly associated with a positive Anti-Jo Ab, which was negative here, making diagnosis challenging.
- This patient was diagnosed with dermatomyositis using histological findings from a skin biopsy. She responded to steroids at acute presentation and treatment was tailored with the addition of mycophenolate and rituximab once DM was diagnosed, leading to complete recovery.
- Our case is unique as the patient had negative Anti-Jo-1 Ab, however, was found to have findings suggesting cellular NSIP on imaging and was diagnosed with dermatomyositis through a skin biopsy. Following diagnosis patient made a full recovery with treatment.

References:

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