ASS: Not Your Typical Myositis

Yejin Diana No, DO; Rina Yadav, DO; Lannie Cation, MD.
St. Vincent Health, Indianapolis, Indiana

Introduction

- Idiopathic inflammatory myopathies (IIM) are acquired autoimmune diseases classified by histopathological findings and include polymyositis, dermatomyositis, or inclusion body myositis.
- Antisynthetase syndrome (ASS), a subset of IIM, constitutes a constellation of symptoms - fevers, weight loss, myositis, Raynaud's phenomenon, mechanic's hands, arthritis, and interstitial lung disease (ILD).
- Patients with ASS demonstrate antibodies to aminoacyl-transfer ribonucleic acid (tRNA) synthetase enzymes.
- We present a case of new fever, shortness of breath, myalgia and rash that prompted autoimmune work-up revealing ASS.

CASE PRESENTATION

- A 50-year-old female with PMH of HTN, depression, and tobacco use presented to hospital with fever, chest pain, and shortness of breath.
- She reported fatigue, aching knuckles, and a "skin sloughing" body rash that was tender to palpation (Figure 2).
- Vital signs: temperature 103F, BP 87/54 mmHg, tachycardia, tachypnea, and oxygen saturation 87% on room air.
- Chest x-ray demonstrated left-sided atelectasis with significant hilar lymphadenopathy.
- Viral panel, blood, and fungal cultures were negative. ESR was 72, CRP was >27 and LDH was 2132.
- CT chest/abdomen/pelvis showed lymphadenopathy and an enlarged left groin lymph node (Figure 1). Lymph node biopsy revealed reactive lymphoid tissue.
- ANA positivity with titer 1:640 (reference range: 1:60) with cytoplasmic speckled appearance. Double stranded DNA, anti-Jo 1/Sm, sjogrens, and complement were within normal limits.
- Anti-histone antibody was elevated at 2.3 (reference range of <1). Anti-OJ antibody was elevated at 23 (reference range of <11). CPK was elevated at 397 indicating a mild myositis.

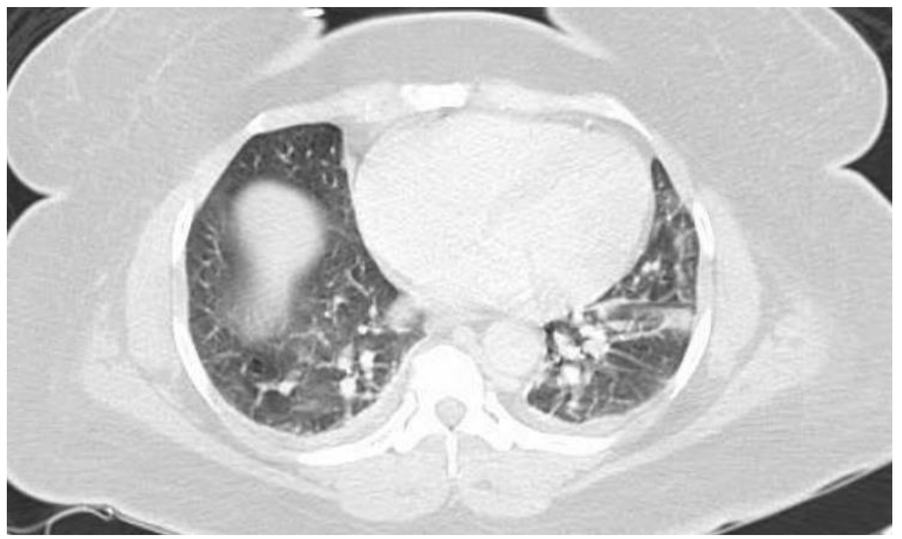

Figure 1. Mild septal thickening, diffusely enlarged lymph nodes and ground-glass opacities on patient's lung CT

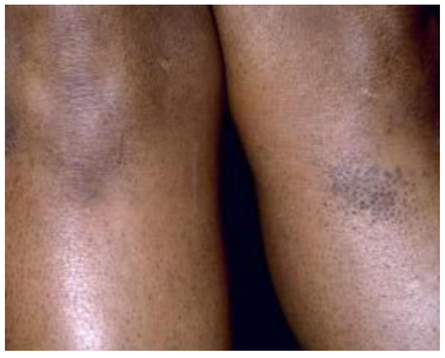

Figure 2. Demonstration of patient's "skin sloughing rash"

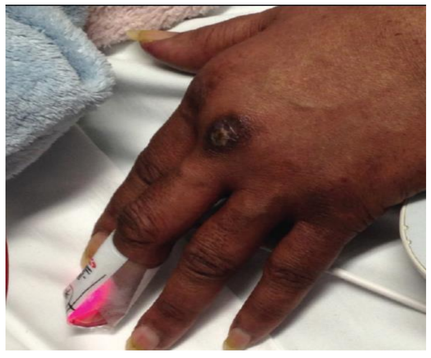

Figure 3. Demonstration of Gottron papules in our patient

PATIENT COURSE

- Patients symptoms persisted upon discharge and she developed new skin lesions on her hands (Figure 3).
- Oral steroid taper was started. Rheumatology confirmed diagnosis of ASS and hydroxychloroquine was started.
- On follow-up visit, patient endorsed improvement in muscle pain.
- Referral to pulmonology was made for PFT and management of ILD.

DISCUSSION

- ASS is a subset of IIM and constitutes a spectrum of symptoms that are both acute and debilitating.
- In a study with 110 patients with ASS, >70% of patients were women, 70% had arthralgias, 50% had muscle weakness, 50% had skin lesions and ILD.
- Reported lesions include ulceration at fingertips, Gottron papule, heliotrope rash, the shawl or V sign, periungual telangiectasia and calcinosis cutis.
- An anti-synthetase panel can help identify patients with that do not fall into the traditional IIM types.
- Once ASS is diagnosed through antibody workup, patients should follow closely with both rheumatology and pulmonology as ILD is major cause of morbidity and mortality.

REFERENCE

Fernandez C, Bardin N, De Paula AM, et al. Correlation of clinicoserologic and pathologic classifications of inflammatory myopathies: study of 178 cases and guidelines for diagnosis. *Medicine (Baltimore)*. 2013;92(1):15-24.

Masiak A, Marzec M, Kulczycka J, Zdrojewski Z. The clinical phenotype associated with antisynthetase autoantibodies. *Reumatologia*. 2020;58(1):4-8.

Oldroyd A, Lilleker J, Chinoy H. Idiopathic inflammatory myopathies - a guide to subtypes, diagnostic approach and treatment. *Clin Med (Lond)*. 2017;17(4):322-328. doi:10.7861/clinmedicine.17-4-322

Targoff IN. Autoantibodies and their significance in myositis. *Curr Rheumatol Rep*. 2008;10(4):333-340.