

CLINICAL VIGNETTE

Dermatomyositis: A Case Review

Delila Pouldar; Sharona Yashar, M.D.; and Veena Vanchinathan, M.D.

Case Presentation

A 53-year-old African American female presents with a one-year history of a “very itchy and painful rash”. The rash first began in a periorbital distribution, progressed to involve the jawline and then the chest, abdomen, back, bilateral upper extremities, and bilateral thighs. Months later, she developed concomitant fatigue and muscle weakness involving her shoulders, hips, and thighs. Due to her occupation as a caregiver with a patient with “a lot of dogs and fleas at his home,” Lyme disease was suspected. However, Lyme disease antibody was negative. The patient also started wearing a new wig about one year prior to onset of symptoms and rash. She was prescribed desonide 0.05% ointment, triamcinolone 0.1% ointment, and clobetasol 0.05% cream to apply to affected areas twice a day as well as prednisone, 60 mg daily. She was also started on methotrexate 25mg/week but showed no response. It was discontinued. IVIG infusions were started with a notable improvement in her erythema and pruritus, which decreased to intermittent and mild. She developed increased swelling in her distal legs despite improvement in myalgias and muscle weakness. Malignancy workup was unremarkable.

Skin exam revealed diffuse xerosis with mild periorbital edema and mild erythema and xerosis of forehead and cheeks, consistent with the “heliotrope sign” seen in dermatomyositis. The “shawl sign” was also apparent with diffuse xerotic coalescing severely erythematous to violaceous patches on neck, chest, abdomen, back, and upper and lower extremities. Other findings included erythematous scaly coalescing macules and patches primarily affecting the interjoint spaces on the bilateral dorsal hands. She also had cuticular fraying with dilated capillaries at proximal nail folds on many fingernails, Gottron’s papules, violaceous scaly papules overlying the metacarpophalangeal, and proximal interphalangeal and distal interphalangeal joints were also apparent.

Laboratory work revealed an elevated CK, elevated aldolase (13.4 U/l), elevated Anti-Nuclear Antibody titer (1:320), and abnormal CBC with elevated red blood cell distribution width, elevated platelet count, low lymphocyte count, and low MCH concentration. The comprehensive metabolic panel, TSH, Anti-Jo1 antibody, and Lyme disease antibody were unremarkable.

Punch biopsy from mid-back, upper left arm, and left thigh all revealed similar patchy interface dermatitis with vacuolar alteration of the dermal-epidermal junction. Both colloidal iron and alcian blue stains on all three specimens reveal a marked increase in dermal mucin deposition.

Discussion

Dermatomyositis is a multi-organ idiopathic inflammatory disorder characterized by proximal skeletal muscle weakness, muscle inflammation, and distinct skin manifestations. It is most commonly seen in females between 40-50 years old.¹ Patients usually report a subacute onset of muscle weakness that is symmetric and proximal. Affected muscles usually include the deltoids, hip flexors, and neck flexors.¹

Gottron’s papules and the heliotrope eruption are distinctive for dermatomyositis while photodistributed erythema, poikiloderma, nailfold changes, scalp involvement, and calcinosis cutis are also common manifestations,² though less specific. Gottron’s papules are described as erythematous to violaceous papules found symmetrically over the extensor aspects of the metacarpophalangeal (MCP) and interphalangeal (IP) joints. Eruptions between the MCP and IP joints may also be seen and be associated with scale and ulcerations.³ The heliotrope sign is an erythematous or violaceous eruption on the upper eyelids, usually accompanied by midfacial erythema similar to the malar rash seen in systemic lupus erythematosus.³ Dermatomyositis patients also demonstrate poikiloderma in sun exposed areas; skin that is both hyperpigmented and hypopigmented. This is most notably seen on the upper back (shawl sign) and lateral aspects of the thigh (holster sign).

Laboratory findings include elevated creatine kinase (CK), lactate dehydrogenase, and aldolase. Antinuclear antibodies (ANA) are present in up to 80% of patients with dermatomyositis and myositis specific autoantibodies, including anti-Jo1, anti-SRP, and anti-Mi2, are found in 30% of patients with dermatomyositis.³ Biopsy of skin lesions usually demonstrate interface dermatitis, described as “mild atrophy of the epidermis with vacuolar changes in the basal keratinocyte layer, as well as perivascular lymphocytic infiltrate in the dermis and increased dermal mucin.”⁴

The association between dermatomyositis and underlying malignancy has been widely accepted. A study by *Chen et al*⁵ confirmed that malignancy diagnosis, specifically nasopharyngeal carcinoma and ovarian cancer, was the highest in the first year after diagnosis of dermatomyositis. Extensive workup for malignancy is vital in patients presenting with dermatomyositis.⁵ Serologically, certain cancers have been associated with serological markers such as anti-p155, anti-155/140, anti-p155/140, anti-MJ, and anti-p140.^{6,7}

Conclusion

Our patient's clinical presentation, significant heliotrope sign, shawl sign, Gottron's papules, and muscle weakness in conjunction with the biopsy pathology are compatible with the clinical impression of connective tissue disease and suggestive of dermatomyositis.

Figures and Images

Figure 1: Patchy interface dermatitis with vacuolar alteration at the dermo-epidermal junction.

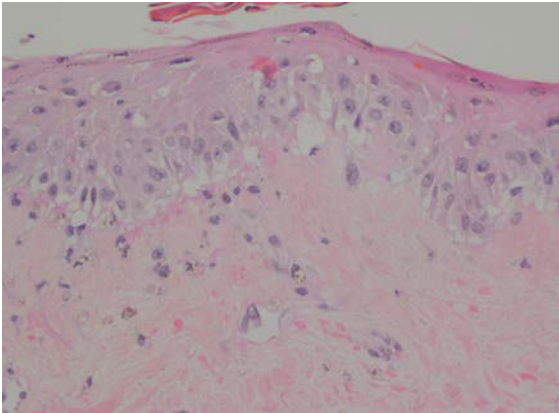


Figure 2: Increased dermal mucin deposition (blue stain).

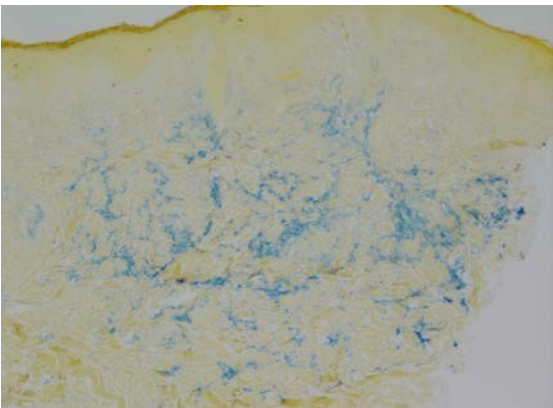


Image 1: Right Lateral Leg.



Image 2.



Image 3: Image of Back.



Image 4.



Image 5: Neck – Shawl Sign.



Image 6: Heliotrope Sign.



Image 7*: Left Lateral Leg.



Image 8*: Hands Revealing Gottron's Papules.



**Images 7 and 8 were taken following IVIG treatment.*

REFERENCES

1. **Tymms KE, Webb J.** Dermatopolymyositis and other connective tissue diseases: a review of 105 cases. *J Rheumatol.* 1985 Dec;12(6):1140-8. PubMed PMID: 4093921.
2. **Cao H, Parikh TN, Zheng J.** Amyopathic dermatomyositis or dermatomyositis-like skin disease: retrospective review of 16 cases with amyopathic dermatomyositis. *Clin Rheumatol.* 2009 Aug;28(8):979-84. doi: 10.1007/s10067-009-1152-9. Epub 2009 Mar 18. PubMed PMID: 19294477.
3. **Dugan EM, Huber AM, Miller FW, Rider LG; International Myositis Assessment and Clinical Studies Group.** Review of the classification and assessment of the cutaneous manifestations of the idiopathic inflammatory myopathies. *Dermatol Online J.* 2009 Feb 15;15(2):2. Review. PubMed PMID: 19336019.
4. **Smith ES, Hallman JR, DeLuca AM, Goldenberg G, Jorizzo JL, Sanguenza OP.** Dermatomyositis: a clinicopathological study of 40 patients. *Am J Dermatopathol.* 2009 Feb;31(1):61-7. doi: 10.1097/DAD.0b013e31818520e1. PubMed PMID: 19155727.
5. **Chen D, Yuan S, Wu X, Li H, Qiu Q, Zhan Z, Ye Y, Lian F, Liang L, Xu H, Yang X.** Incidence and predictive factors for malignancies with dermatomyositis: a cohort from southern China. *Clin*

Exp Rheumatol. 2014 Sep-Oct;32(5):615-21. Epub 2014 Jul 28. PubMed PMID: 25068842.

6. **Chinoy H, Fertig N, Oddis CV, Ollier WE, Cooper RG.** The diagnostic utility of myositis autoantibody testing for predicting the risk of cancer-associated myositis. *Ann Rheum Dis.* 2007 Oct;66(10):1345-9. Epub 2007 Mar 28. PubMed PMID:17392346; PubMed Central PMCID: PMC1994304.
7. **Fiorentino DF, Chung LS, Christopher-Stine L, Zaba L, Li S, Mammen AL, Rosen A, Casciola-Rosen L.** Most patients with cancer-associated dermatomyositis have antibodies to nuclear matrix protein NXP-2 or transcription intermediary factor 1 γ . *Arthritis Rheum.* 2013 Nov;65(11):2954-62. doi: 10.1002/art.38093. PubMed PMID: 24037894; PubMed Central PMCID: PMC4073292.

Submitted May 11, 2015