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Subtle periorbital edema and hyperkeratotic papules in a woman of color

Sara Asbeck, BS[a], Brittany Smirnov, DO[b,*]

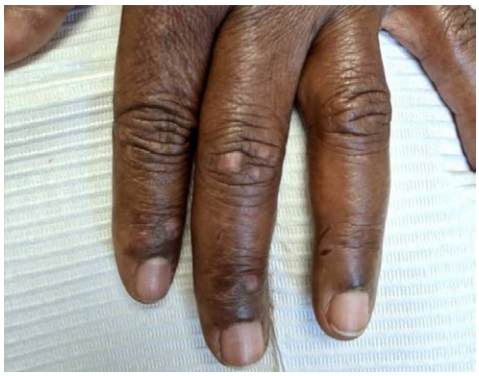

Fig. 1. Flat-topped, hyperkeratotic papules overlying the metacarpophalangeal and proximal interphalangeal joints.

Case summary

A 65-year-old African-American female presented to the dermatology clinic complaining of poorly controlled melasma over the past year. Physical examination incidentally revealed bilateral violaceous periorbital erythema and edema, accentuated on the left eye, patchy gray to violaceous pigmented patches on forehead and glabella, and flat-topped, hyperkeratotic papules overlying the metacarpophalangeal and proximal interphalangeal joints (Fig. 1). Upon further inspection with capillaroscopy, periungual, dilated, looped telangiectasias were evident (Fig. 2). The patient denied proximal muscle weakness, fatigue, weight loss, shortness of breath, chest pain, or dyspnea on exertion. A punch biopsy above the left metacarpophalangeal joint was taken revealing perivascular lymphocytic infiltrate with epidermal hyperplasia, acanthosis, slight increase of papillary dermal mucin, and mild vacuolar interface at the dermoepidermal junction. Direct immunofluorescence revealed C5b-9 and weak IgM and C3 deposition with cytoid bodies along the epidermal basement membrane.

Question 1

What is the correct diagnosis?

A. Psoriasis
B. Lichen planus pigmentosus
C. Dermatomyositis
D. Scleroderma
E. Systemic lupus erythematosus

Correct answer: C. Dermatomyositis

Discussion

Dermatomyositis is a rare idiopathic inflammatory myopathy with classic cutaneous characteristics. Dermatomyositis typically presents with a symmetric dusky erythematous to violaceous heliotrope rash with mild scaling and desquamation. The characteristic hand findings, termed Gottron's papules, consist of erythematous to violaceous papules and plaques, distributed over the bony prominences. Several other non-pathognomonic cutaneous findings may be present, including malar rash, poikiloderma in a photosensitive distribution, periungual and cuticular changes, and alopecia.[1] Most patients will also report proximal muscle weakness, but cutaneous findings without myopathy are possible - a process termed "amyopathic" dermatomyositis. In such a

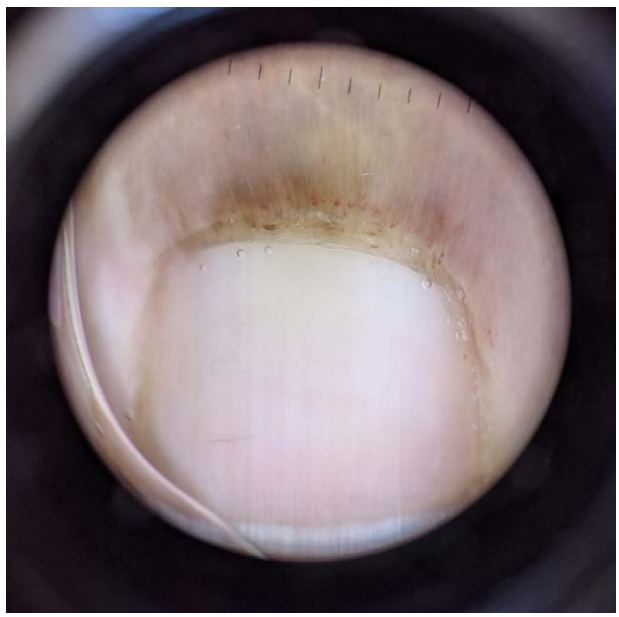

Fig. 2. Periungual, dilated, looped telangiectasias.

[a] *Fuchs Dermatology, Falls Church, VA,*
[b] *University of Miami Miller School of Medicine, Miami, FL*

Current address for Brittany Smirnov: 6565 Arlington Boulevard Suite 102, Falls Church, VA, 22042; Current address for Sara Asbeck: 5641 Gramercy Drive West Palm Beach, FL 33407.

** Corresponding author.*

E-mail address: smp268@med.miami.edu (S. Asbeck).

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case, repeat testing for myopathy may be warranted in the future, as cutaneous symptoms may precede muscle inflammation by up to 2 years.[1]

When assessing a patient with suspected dermatomyositis, providers should test for proximal muscle weakness and draw labs for markers of muscle inflammation. The diagnosis can be confirmed with a muscle biopsy demonstrating perivascular lymphocytic inflammation in the perimysium and electromyography (EMG), which may show membrane irritability, fibrillary potentials, and decreased action potential amplitude.[1] Skin biopsies are not routinely used in diagnosis, as findings resemble those seen in lupus erythematosus, but may be an important clue in the case of amyopathic dermatomyositis.[1] In our case, creatine phosphokinase, troponins, and muscle strength testing were all negative. Muscle biopsy was not performed due to lack of indication from laboratory and clinical testing; however, the classic cutaneous findings and skin biopsy are consistent with a diagnosis of amyopathic dermatomyositis.

The mainstay of treatment for dermatomyositis consists of immunosuppression, often with prednisone, either as monotherapy, or combined with another agent.[1] Muscle inflammation is typically more responsive to treatment than skin manifestations, which may take longer to resolve. Treatment for amyopathic dermatomyositis includes topical corticosteroids, topical tacrolimus, hydroxychloroquine, low-dose methotrexate, and strict sun protection and avoidance of UV light, most notably UVA.[1]

Our patient was started on systemic corticosteroids and underwent extensive screening to evaluate for the possibility of solid organ malignancy, with no malignancies identified. She will continue to receive age-appropriate screening, as the risk of malignancy is highest in the first year of diagnosis, but remains elevated for up to 3 years after symptom onset.[2] The patient is currently well-controlled on hydroxychloroquine 200 mg BID, prednisone 10 mg daily, and strict photoprotection.

This case was selected to increase awareness among medical practitioners, highlighting how dermatomyositis can subtly present in patients with skin of color. With a female-predominant condition such as dermatomyositis, where coexisting malignancy is identified in 20–25% of cases,[2] prompt identification, diagnosis, and referral for cancer screening is paramount, and practitioners should be confident in diagnosing this condition in patients of all skin types.

Question 2

What autoantibodies are most frequently associated with dermatomyositis?

A. Anti-Mi-2
B. Anti-SSA
C. Anti-Jo-1
D. Anti-Ro
E. ANA

Correct answer: A. Anti-Mi-2 antibodies are most frequently associated with the classic cutaneous features of DM and correlate to a lower risk of interstitial lung disease and malignancy compared to other autoantibodies in DM.[3] Anti-Jo-1 are most common in antisynthetase syndrome, now recognized as a distinct entity from other subgroups of myositis and are strongly associated with the development of interstitial lung disease.[3] ANA antibodies are found in 60-90% of dermatomyositis patients but are nonspecific and present in numerous autoimmune conditions.[1]

Conflicts of interest

None.

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None.

Author contributions

Sara Asbeck contributed to the performance of the research and writing of the manuscript. Brittany Smirnov contributed to the performance of the research and review of the manuscript.

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