CLINICAL VIGNETTE

Poikiloderma as a Presenting Sign of Amyopathic Dermatomyositis

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Case Presentation

A 71-year-old female presented to primary care with 4 months of a pruritic, erythematous rash on the right side of her face (Figure 1). She had no other symptoms with review of systems negative for myalgias, arthralgias, malar rash, photosensitivity, weakness, numbness, or tingling. Initial exam revealed a mottled, erythematous, blanching plaque on the right side of the face and normal strength throughout. Over a year the rash progressed to involve the left side of her face, eyelids, dorsal surface of the hands with violaceous papules over the extensor surfaces of the metacarpophalangeal and interphalangeal joints, forehead, upper arms, and thighs.

Laboratory testing showed normal AST, ALT, CK, aldolase, SSB, anti-histone Ab, Mi-2 Auto Ab. ANA was positive with a titer of >1:2560 with a speckled staining pattern. SSA and anti-small ubiquitin-like modifier activating enzyme (SAE) auto-antibodies were also positive.

Imaging studies included CT chest, abdomen and pelvis which was unremarkable. Age-appropriate cancer screening tests (mammogram and colonoscopy) did not show any malignancy.

The patient was referred to Dermatology and a punch biopsy revealed interface dermatitis with superficial-to-mid dermal perivascular lymphocyte-predominant inflammation consistent with connective tissue disease. There was no definite thickening of dermal collagen was not identified. She was treated empirically for localized scleroderma. However, the skin lesions did not respond to topical steroids or tacrolimus 1% cream.

Rheumatology was also consulted for assistance with further management and definitive treatment. At the time of evaluation by Rheumatology the rash had progressed, with erythema spreading to the chest, back, posterior neck and scalp. Due to the presence of poikiloderma, heliotrope rash (Figure 2), Gottron's papules (Figure 3), shawl sign (Figure 4), and holster signs (Figures 5a and 5b), there was concern for dermatomyositis – despite the absence of arthralgias or myalgias.

Given the pathognomonic skin findings above and absence of myopathy for more than 6 months our patient was diagnosed with amyopathic dermatomyositis and treated with intravenous immune globulin (IVIG), oral methotrexate, and a prednisone taper. The patient reported resolution of all skin lesions except for the heliotrope rash which persisted but significantly improved.

Discussion

Classical dermatomyositis (CDM) presents with both cutaneous involvement and symmetric muscle weakness as well as laboratory findings of muscle inflammation. There are several subtypes of CDM including amyopathic dermatomyositis (ADM), hypomyopathic dermatomyositis and postmyopathic dermatomyositis. Our patient's presentation of isolated skin findings in the absence of clinical and laboratory evidence of myopathy is highly suggestive of ADM. Amyopathic DM is historically known as dermatomyositis siné myositis, in which the characteristic skin findings of dermatomyositis are present for at least 6 months without the development of clinically significant myositis and with normal muscle enzymes.[1]

Dermatomyositis typically occurs in a bimodal age distribution affecting children as well as the middle-aged and elderly.[2] Females are predominately affected. The pathogenesis of these inflammatory myopathies is not well-understood. Amyopathic dermatomyositis is thought to represent 20% of all cases of dermatomyositis.[3]

The diagnosis of ADM can be made by identifying the pathognomonic cutaneous findings on physical exam in the setting of normal muscle enzymes. Skin biopsy is also helpful, as skin findings may mimic several other conditions including contact dermatitis, systemic lupus erythematosus, polymorphic light eruption or localized scleroderma. There is generally no role for EMG, MRI, and/or muscle biopsy, unless muscle weakness is present on exam. Treatment of cutaneous manifestations of all forms of DM includes photoprotection, antipruritic agents, topical corticosteroids, topical calcineurin inhibitors, antimalarials and/or methotrexate. Refractory cases require systemic therapy with medications such as IVIG or mycophenolate mofetil.[4]

The incidence of internal malignancy in ADM mirrors that of classic DM which is 5- to 7-fold that of the general population.[5] Thus a careful history and physical as well as age-appropriate cancer screenings are recommended in all dermatomyositis patients.[6] Furthermore, an association with interstitial lung disease (ILD) has also been documented but is not well-understood. More studies are needed to better understand the association of DM with increased risk of malignancy and ILD.

Conclusion

Our patient's clinical presentation of erythematous patches across the body, positive heliotrope rash, shawl and v-signs, Gottron's papules, holster sign, and poikiloderma of the face in the absence of myositis is consistent with amyopathic dermatomyositis. This diagnosis was further supported by the positive SSA and anti-SAE autoantibodies as well as the skin biopsy consistent with connective tissue disease. This was a unique case of ADM presenting as isolated poikiloderma that only over time progressed to the characteristic dermatomyositis skin findings, thus delaying diagnosis. ADM and other connective tissue diseases should be considered in the differential diagnosis of persistent skin lesions not responding to usual topical therapies and prompt referral for skin biopsy should be considered.

Figures

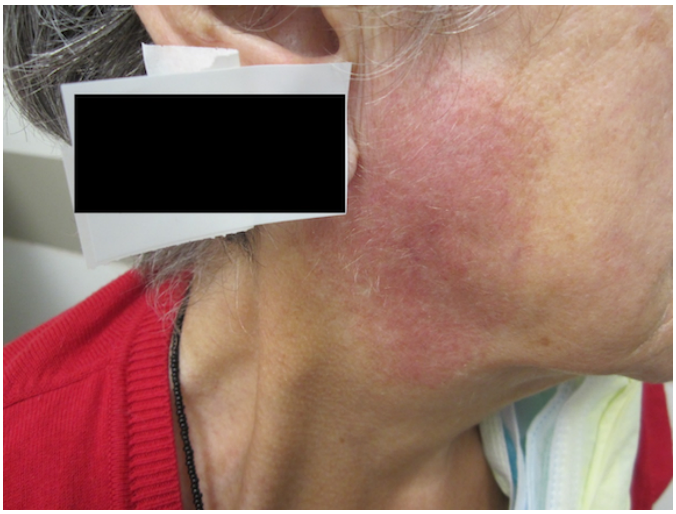

Figure 1. Poikiloderma: unilateral mottled erythematous blanching patch on the face

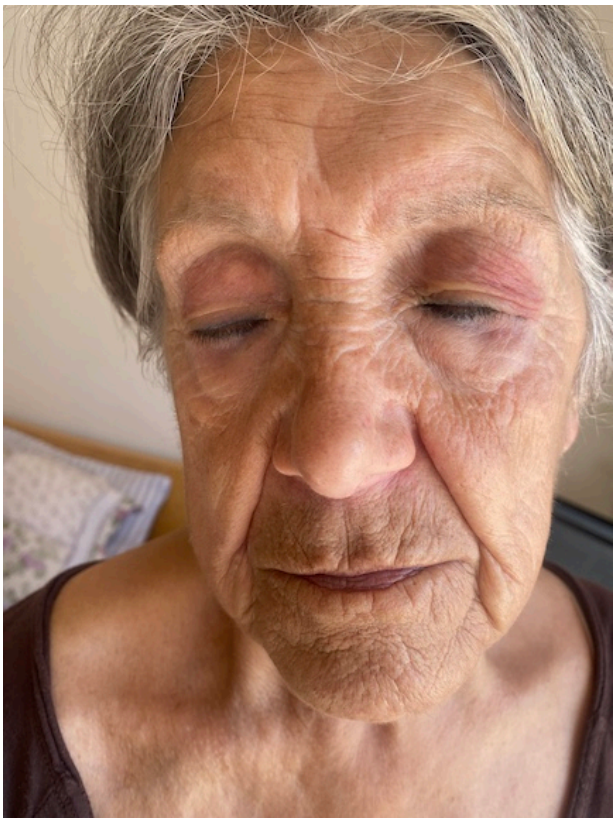

Figure 2. Heliotrope rash: bilateral violaceous rash on the eyelids

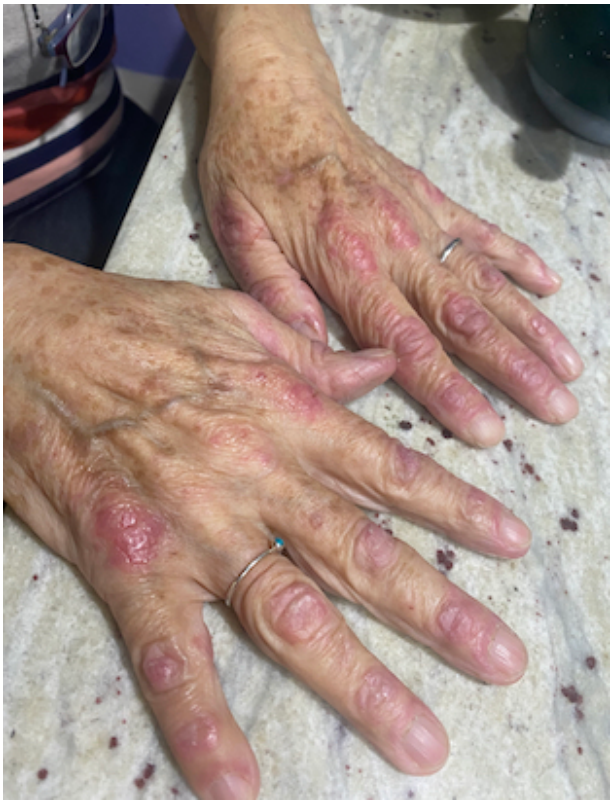

Figure 3. Gottron's papules: violaceous papules over the MCPs, PIPs and DIPs

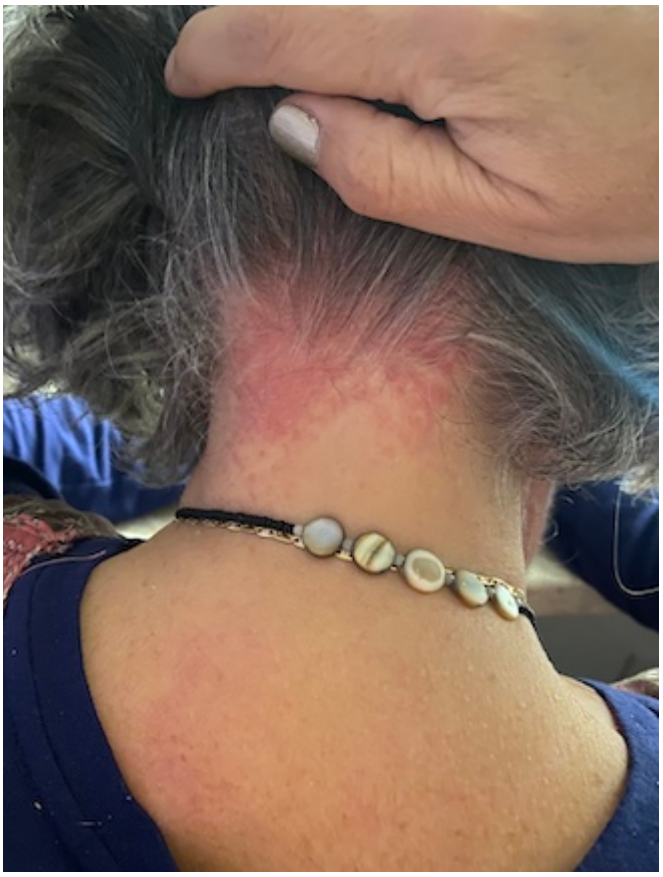

Figure 4. Shawl sign: erythema of the posterior neck

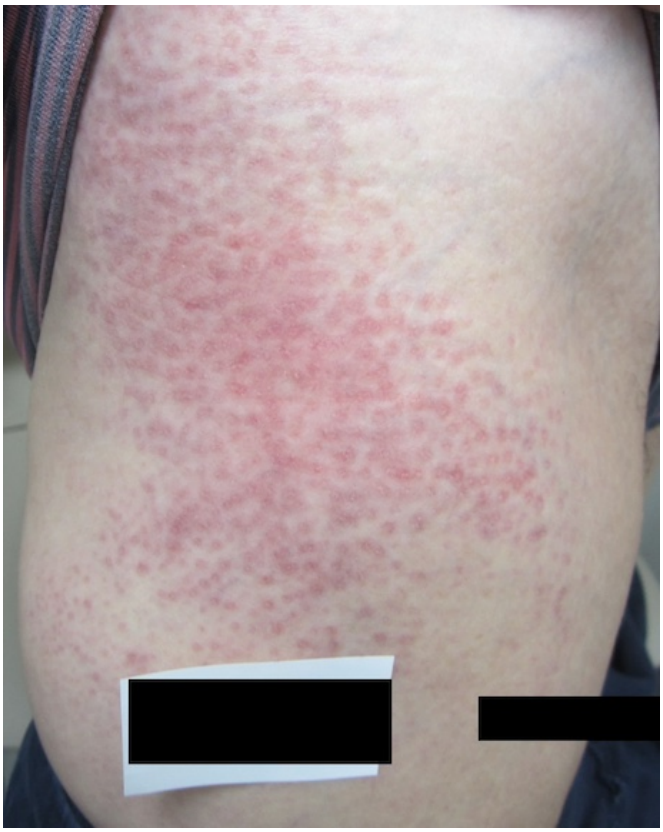

Figure 5a. Holster sign of the right thigh

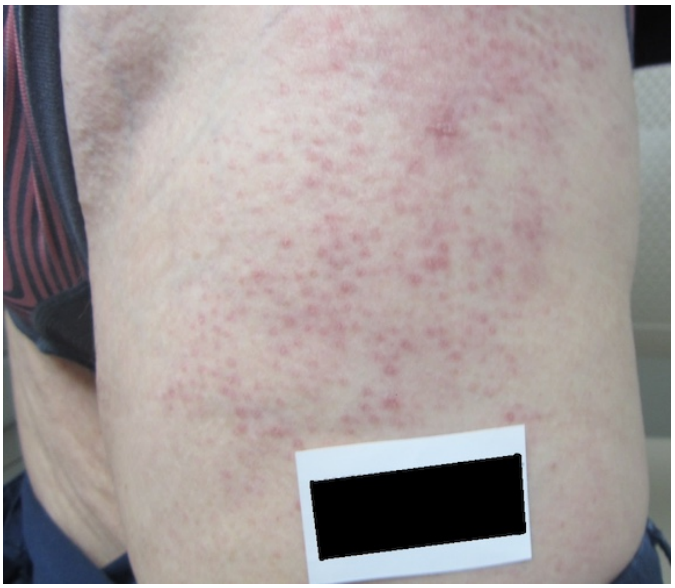

Figure 5b. Holster sign of the left thigh

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