Distinct purpuric lesions in patients with dermatomyositis

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Key words: dermatomyositis; purpura.

INTRODUCTION

Dermatomyositis (DM) is an autoimmune disease that can have significant skin, muscle, and pulmonary morbidity that has been estimated to affect 1 to 6 per 100,000 adults in the United States.[1] DM has diverse cutaneous and systemic manifestations that require a high level of suspicion for early diagnosis. Skin findings such as Gottron papules, Gottron sign, and heliotrope rash have been described as characteristic findings, although the diagnosis can still be difficult, especially in patients without myositis.[2]

We report a distinct cutaneous sign with purpuric morphology on the face and scalp that we believe to be the cutaneous manifestations of DM that have not been previously described.

CASE REPORT

Patient 1 is a 79-year-old White woman with an established diagnosis of clinically amyopathic DM with antitranscription intermediary factor 1-gamma (anti-TIF1-γ) antibodies and moderate skin disease for 4 years. She presented to the clinic with numerous purpuric macules on the scalp and frontal hairline with no scale and minimal erythema, most prominently on the frontal and bilateral parietal scalp (Fig 1). The patient had stopped taking methotrexate for 1 year prior to the appearance of the lesions. The lesions were mildly pruritic and fixed over time. These macules persisted for 3 years after resolving rapidly following the therapy with an investigational agent that was thought to interfere with interferon signaling.

Patient 2 is a 77-year-old White woman who was admitted for progressive dysphagia, oral ulcers, and cutaneous ulcers, despite being on azathioprine and prednisone treatment, with a recent diagnosis of DM. She presented with muscle weakness and elevated creatine kinase level with evidence of myositis on magnetic resonance imaging and was found to have antibodies against TIF1-γ. She was diagnosed with diffuse large B-cell lymphoma. She was noted to have macular ecchymotic and purpuric papules and plaques on the hairline and lateral aspect of the face (Fig 2) along with other classic cutaneous findings of DM. The lesions persisted for 12 weeks until the patient ultimately passed away because of the complications of her cancer.

Abbreviations used:

DM:	dermatomyositis
TIF1-γ:	transcription intermediary factor 1-gamma

Patient 3 is a 54-year-old White woman with an established diagnosis of classic DM for 4 years. She had a stable mild skin disease that was being maintained on mycophenolate mofetil treatment, and she was found to have anti-TIF1-γ antibodies. While continuing this medication regimen, she noted the new appearance of purpuric macules that were clustered into a well-demarcated patch at the left angle of the jaw (Fig 3) in the setting of her chronic Gottron papules, mechanic hands, and Holster sign. The lesions remained fixed and eventually resolved after 6 months.

DISCUSSION

We present the cases of 3 patients with DM with varying disease courses with purpuric lesions on the face and scalp. Lack of skin biopsy, unfortunately, precludes their further characterization, although a

From the Department of Dermatology, Stanford University School of Medicine.

Funding sources: None.
IRB approval status: Not applicable.

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JAAD Case Reports 2021;13:94-6.
2352-5126

https://doi.org/10.1016/j.jdcr.2021.05.006

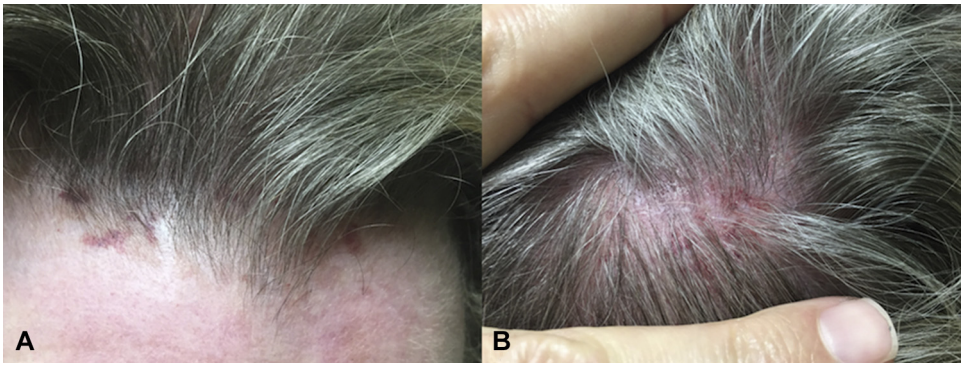

Fig 1. Patient 1 with dermatomyositis. **A,** Numerous purpuric macules on the frontal hairline and scalp. **B,** Close-up picture of purpuric macules on parietal scalp.

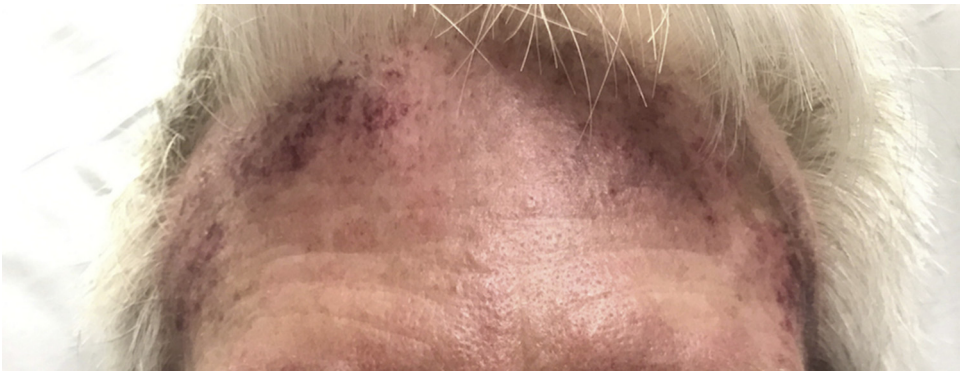

Fig 2. Patient 2 with dermatomyositis. Macular ecchymotic and purpuric papules on the hairline and lateral aspect of the face.

vasculitic or traumatic etiology is unlikely given their fixed and persistent nature over months to years. Although 2 patients were relatively stable in their disease course with an established diagnosis at the time the purpuric lesions appeared, 1 patient presented with the purpuric lesions in the context of severe proximal muscle weakness, dysphagia, and a recent DM diagnosis. Differential diagnosis of these lesions may include cutaneous manifestations, such as "red on white" and purpuric palatal patch, that are more commonly seen in patients with anti-TIF1-γ.[3,4] Although these are distinct findings from purpuric lesions, these mucocutaneous manifestations may be on the same spectrum. These are likely to be DM-specific findings of disease activity that are not necessarily representative of irreversible damage. In addition, leukocytoclastic vasculitis may also be a differential diagnosis; however, most small vessel vasculitic lesions are not fixed and do not persist over months to years.[5]

It may be of significance that all 3 patients were found to have antibodies against TIF1-γ. Although anti-TIF1-γ antibodies are associated with cancer, these purpuric lesions were not confined to patients with malignancy diagnosis. More data are required to determine whether they are a specific manifestation within the anti-TIF1-γ population.[6,7]

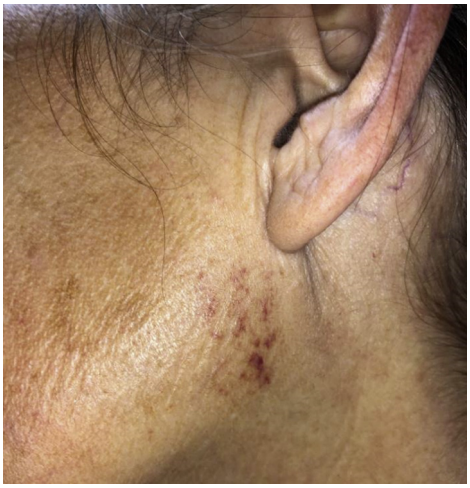

Fig 3. Patient 3 with dermatomyositis. Purpuric macules clustered into a well-demarcated patch at the left angle of the jaw.

We propose that these cutaneous findings might help support a diagnosis of DM in a patient with ambiguous findings and that they represent an unusual manifestation of active skin disease in DM.

Conflicts of interest

None disclosed.

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