CASE REPORT

Distinct purpuric lesions in patients with dermatomyositis

Sung Kyung Cho, BA, Elizabeth Messenger, MD, and David Franklin Fiorentino, MD, PhD
Stanford, California

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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.

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

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Correspondence to: David Franklin Fiorentino, MD, PhD, Department of Dermatology, Stanford University School of Medicine, 450 Broadway Street, Pavilion B, 4th Floor, Redwood City, CA 94063. E-mail: fiorentino@stanford.edu.

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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
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.

REFERENCES
1. Furst DE, Amato AA, Iorga ŞR, Gajria K, Fernandes AW. Epidemiology of adult idiopathic inflammatory myopathies in a US managed care plan. Muscle Nerve. 2012;45(5):676-683.
2. DeWane ME, Waldman R, Lu J. Dermatomyositis: clinical features and pathogenesis. J Am Acad Dermatol. 2020;82(2):267-281.
3. Bendewald MJ, Wetter DA, Li X, Davis MD. Incidence of dermatomyositis and clinically amyopathic dermatomyositis: a population-based study in Olmsted County, Minnesota. Arch Dermatol. 2010;146(1):26-30.
4. Bernet LL, Lewis MA, Rieger KE, Casciola-Rosen L, Fiorentino DF. Ovoid palatal patch in dermatomyositis: a novel finding associated with anti-TIF1γ (p155) antibodies. JAMA Dermatol. 2016;152(9):1049-1051.
5. Baigrie D, Bansal P, Goyal A, Crane JS. Leukocytoclastic vasculitis. In: StatPearls [Internet]. StatPearls Publishing; 2021.
6. Hamaguchi Y, Kuswana M, Hoshino K, et al. Clinical correlations with dermatomyositis-specific autoantibodies in adult Japanese patients with dermatomyositis: a multicenter cross-sectional study. Arch Dermatol. 2011;147(4):391-398.
7. Kaji K, Fujimoto M, Hasegawa M, et al. Identification of a novel autoantibody reactive with 155 and 140 kDa nuclear proteins in patients with dermatomyositis: an association with malignancy. Rheumatology (Oxford). 2007;46(1):25-28.