

## BRIEF ARTICLE

**Clue to Diagnosis: Flagellate Erythema in Anti-MDA5 Antibody-Positive Dermatomyositis in a Patient with Skin of Color**

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**ABSTRACT**

Anti-melanoma differentiation-associated gene 5 (MDA5) antibody-positive dermatomyositis (DM) is a rare subtype of clinically amyopathic DM characterized by minimal muscle involvement, variable skin manifestations, and a high risk of rapidly progressive interstitial lung disease (RP-ILD). Atypical skin findings are frequently misdiagnosed, delaying recognition and treatment. We present the case of a 46-year-old Black woman who developed flagellate erythema, facilitating diagnosis of anti-MDA5 DM complicated by RP-ILD. This case highlights the importance of recognizing uncommon skin findings like flagellate erythema, especially in patients with subtle or atypical presentations, and emphasizes the importance of early diagnosis and prompt immunosuppressive therapy to improve clinical outcomes.

**INTRODUCTION**

Anti-MDA5 DM is a subtype of clinically amyopathic DM characterized by absent inflammatory myopathy, diverse skin manifestations extending beyond hallmark signs such as Gottron's papules and heliotrope eruption, and a high risk of RP-ILD.<sup>1-4</sup>

We report a case of anti-MDA5 DM with RP-ILD, presenting with the rare cutaneous manifestation of flagellate erythema.

**CASE REPORT**

A 46-year-old woman from Zambia presented to the emergency department (ED) with a three-week history of progressive dyspnea, nonproductive cough, and fatigue. One month earlier, she had been treated with topical corticosteroids and a five-day course of oral prednisone for presumed allergic contact dermatitis from poison ivy, affecting the right dorsal hand (**Figure 1**), knees, and upper back. She refrained from scratching the rash, fearing it might worsen.

In the ED she was febrile with mild inspiratory crackles on auscultation and preserved muscle strength. Cutaneous findings included hyperpigmented patches on the forehead, knees, elbows, and right dorsal hand, along with linear hyperpigmented streaks forming a flagellate pattern on the



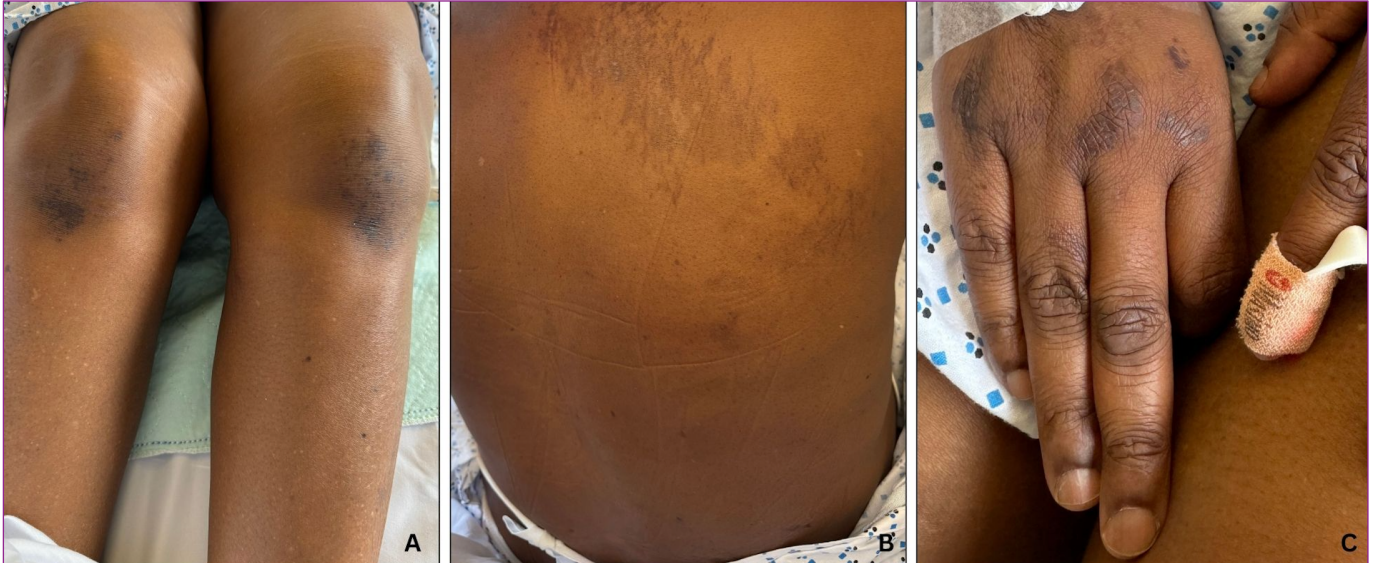
**Figure 1.** Gottron's papules on the right dorsal hand, one month prior to presentation.

right upper back (**Figure 2 - A, B, C**), raising concern for DM. Healing digital ulcers were also noted (**Figure 3**). Nailfold capillaroscopy revealed a tortuous capillary pattern and capillary dropout. A skin biopsy of the rash on the back showed findings consistent with post-inflammatory hyperpigmentation. Cutaneous direct immunofluorescence testing revealed discontinuous, weak granular deposition of C3 complement along the basement membrane, as well as nonspecific fibrinogen deposition within the connective tissue. Laboratory results revealed elevated inflammatory markers: C-reactive protein (CRP) (22.8 mg/L), lactate dehydrogenase (417 U/L), and ferritin (899 ng/mL), with normal creatine kinase and aldolase.

Initially diagnosed with pneumonia based on chest X-ray findings, she failed to improve with empiric antibiotics. High-resolution chest CT showed patchy consolidations and ground-glass opacities in the upper lobes and perihilar regions. Pulmonary function tests demonstrated moderate restrictive impairment and severely reduced diffusing capacity for carbon monoxide.

A myositis-specific panel returned positive for anti-MDA5 antibody. Antinuclear antibody testing was positive at a titer of 1:640 with a dense fine speckled pattern. The extractable nuclear antigen panel and myositis-associated antibodies were negative.

The patient was diagnosed with amyopathic anti-MDA5 DM with RP-ILD and treated with pulse-dose methylprednisolone, intravenous



**Figure 2.** (A) Hyperpigmented patches on the bilateral extensor knees. (B) Hyperpigmented streak-like eruption representing resolving flagellate erythema on the upper back. (C) Hyperpigmented patches on the right dorsal hand, representing healing Gottron's papules.



**Figure 3.** Healing periungual ulcers.

immunoglobulin (IVIG), mycophenolate mofetil, and tacrolimus. Despite initial clinical improvement and normalization of CRP levels to  $<3.0$  mg/L, she continued to experience exertional dyspnea. This prompted a switch from mycophenolate mofetil to combination therapy with rituximab and the Janus Kinase (JAK) inhibitor, tofacitinib, as well as a referral for lung transplantation. The case remains ongoing without a final outcome at this time.

## DISCUSSION

DM-specific antibodies identify distinct clinical subtypes and are crucial in predicting patient outcomes.<sup>2</sup> Anti-MDA5 DM is a rare systemic autoimmune disorder initially described in Japanese patients with clinically amyopathic DM.<sup>4</sup> This subtype is distinguished by a strong association with RP-ILD, a major contributor to early mortality.<sup>1,4</sup> Unlike classic DM, it often presents with minimal muscle involvement.<sup>1</sup>

Currently, no standardized treatment guidelines exist, and most recommendations are derived from observational studies and case reports.<sup>1,4</sup> Conventional therapy involves aggressive immunosuppression with high-dose glucocorticoids, calcineurin inhibitors, cyclophosphamide, mycophenolate mofetil, rituximab, and IVIG infusions.<sup>1,4</sup> Recently, JAK inhibitors have emerged as promising therapeutic agents.<sup>1,4</sup>

Prompt initiation of treatment relies on a timely diagnosis, often difficult in patients with anti-MDA5 DM as its cutaneous manifestations may mimic other conditions or differ from typical DM findings. The challenge is further exacerbated when patients present with secondary skin changes, such as post-inflammatory hyperpigmentation, due to the natural progression of primary lesions, as

occurred in this case. Diagnostic challenges are further amplified in the Black African American population due to limited data on DM-related skin presentations in this group.<sup>5</sup> Patients with anti-MDA5 DM can present with classic cutaneous findings such as heliotrope rash, Gottron's papules, and the "shawl sign" in 60-70% of cases.<sup>3,4</sup> However, these features may be absent, subtle, or misdiagnosed, as in our patient. Instead, individuals may exhibit unique cutaneous features, including palmar papules with hyperkeratosis and ulcerations, including periungual ulcers.<sup>1,4,6</sup> Other rare signs include auricular skin lesions, oral ulcers, panniculitis, alopecia, "Wong-type DM" (confluent reddish-orange plaques and follicular papules with "islands of sparing"), and flagellate erythema.<sup>1,3,4</sup>

Flagellate erythema is a rare, abrupt-onset eruption characterized by streak-like, linear, or curving erythematous macules, papules, or plaques, typically located on the trunk and resembling whiplash marks.<sup>2,3,7,8</sup> Initially reported with bleomycin toxicity or shiitake mushroom ingestion, these lesions may be pruritic or uncomfortable but often resolve spontaneously.<sup>3,8</sup> As demonstrated in our case, resolution may occur with residual hyperpigmentation.

Flagellate erythema may precede or accompany systemic manifestations, most notably RP-ILD, a major cause of morbidity and mortality in anti-MDA5 DM. It has been reported in approximately 5% of DM cases.<sup>3,7</sup> To our knowledge, only three cases of flagellate erythema associated with anti-MDA5 antibody positivity have been described.<sup>2,9,10</sup> Histopathology is nonspecific and may show vacuolar interface dermatitis, while direct immunofluorescence findings are variable and often inconclusive.<sup>1</sup>

In anti-MDA5 DM, recognizing uncommon cutaneous findings such as flagellate erythema is essential to avoid diagnostic and management delays. When accompanied by systemic features, such as fever or dyspnea, and supportive serologies, flagellate erythema should raise suspicion for DM, particularly in clinically amyopathic cases. Prompt recognition and workup are critical, as timely initiation of aggressive immunosuppressive therapy may improve outcomes.

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