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Clinical image: Heliotrope rash, Gottron's papules, and Mechanic's hands in a 29-year-old Woman with Dermatomyositis from Peru

Imagen clínica: Eritema en Heliotropo, Pápulas de Gottron y Manos de Mecánico en una Mujer Peruana de 29 años con Dermatomiositis.

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Conflicts of interest

The authors do not report having any conflicts of interest.

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Presentation

A 29-year-old Peruvian woman, previously healthy, presented with a 6-month history of fever, headache, facial rash, myalgias, weight loss, dysphagia, and progressive weakness. The weakness initially affected her lower extremities, progressively extending to her arms. Physical examination revealed dermal papules on both hands, and periorbital erythema (figure 1). Proximal muscle weakness, muscle edema, alopecia (figure 2), and limited proximal joint ranges were also observed. Laboratory tests revealed elevated levels of creatine phosphokinase (CPK) at 6187 U/L (reference range of 30-135 U/L) and lactate dehydrogenase (LDH) at 1771 U/L (reference range of 120-246 U/L). Additionally, a positive speckled ANA pattern was observed. Extractable nuclear antigen (ENA) panel (anti ds-DNA, Sm, RNP, Ro, 52kDa, SS-B, Scl-70, histone, and Jo-1) was negative and complement levels were normal. Electromyography indicated low-amplitude waves at small intervals, suggesting inflammatory myopathies. A gastroduodenoscopy and computed tomography scan were performed to evaluate dysphagia and rule out malignancies, with both yielding negative results. A skin biopsy from the forehead revealed perimysial and perivascular inflammation, confirming the diagnosis of dermatomyositis. Corticosteroid therapy was initiated at a dosage of 1 mg/kg/day, along with weekly methotrexate pulses. The patient showed partial clinical improvement before discharge, and CPK levels lowered down to 2959 U/l. At the 3-month follow-up, CPK levels were measured at 347 U/l. Clinical improvement remained partial, since the patient required assistance to stand and walk more than a few steps.



Figure 1. Displays multiple lesions, expanding the scope of the differential diagnosis. A) Facial erythema is visible, extending across the malar region while respecting nasal folds. The heliotrope rash, a pathognomonic manifestation of dermatomyositis, is evident, rarely occurring unilaterally such as this case. B) Gottron papules, another pathognomonic feature, are evident on the knuckle region and dorsum of the fingers. C) The inverse Gottron sign, represented by gottron papules extending from the dorsum to the ventral and lateral aspect of the fingers. Additionally, palm desquamation and thickening, cracked fingertips and cuticle erythema indicate mechanic's hands. D) Nail fold changes are visible, including irregular, thickened cuticles and ungueal telangiectasias.

Discussion

Dermatomyositis is a rare autoimmune disorder classified among idiopathic inflammatory myopathies.¹ It involves the activation of CD4+ T cells, leading to the destruction of capillaries in the peripheral muscle fascicles.¹ Gottron papules and heliotrope eruption, though typically pathognomonic, can rarely present inversely and unilaterally respectively. Both features may suggest anti-MDA5 antibody-positive dermatomyositis and interstitial lung disease.² Dermatomyositis affects muscles and skin, resembling other conditions. Skin manifestations can mimic alopecia and photodistributed poikiloderma in SLE³; mechanic's hands in antisynthetase syndrome; pruritus, skin thickening and unguis telangiectasias in systemic sclerosis⁴; and scaly patches and papulosquamous lesions in extensor areas in psoriasis.⁴ Fatigue, common in these entities, is often mistaken for true muscle weakness. Therefore, a comprehensive assessment is crucial for an accurate diagnosis. A negative ENA profile helped us rule out potential confounding diagnoses, including anti-ds-DNA and anti-histone for SLE, anti-Jo1 for antisynthetase syndrome, and anti-Scl-70 for systemic sclerosis. Aggressive surveillance for malignancy and follow-up in dermatomyositis is mandatory given its paraneoplastic nature.

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