Rev. Cuerpo Med. HNAAA, Vol 17 (3) - 2024

REVISTA DEL CUERPO MÉDICO HOSPITAL NACIONAL ALMANZOR AGUINAGA ASENJO, CHICLAYO, PERÚ

ISSN | impresa: 2225-5109; Electrónica: 2227-4731

Cross Ref. DOI: 10.35434/rcmhnaaa | OJS https://cmhnaaa.org.pe/ojs

Photo gallery

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

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

David Guevara-Lazo[1,a], Valeria Palacios-Tealdo[1,a], Natalia Nombera-Aznaran[1,a], Armando Calvo-Quiroz[1,2,b]

DOI

https://doi.org/10.35434/rcmhnaaa.2024.173.2452

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.

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 SLE[3]; mechanic's hands in antisynthetase syndrome; pruritus, skin thickening and ungual telangiectasias in systemic sclerosis[4]; 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.

FILIATION
1. Faculty of Medicine, Universidad Peruana Cayetano Heredia, Lima, Perú.
2. Department of Rheumatology, Hospital Cayetano Heredia, Lima, Perú.
a. Medical student.
b. Medical Doctor.

ORCID
1. David Guevara-Lazo / 0000-0001-7983-6553
2. Valeria Palacios-Tealdo / 0009-0000-7531-0642
3. Natalia Nombera-Aznaran / 0000-0003-1090-863X
4. Armando Calvo-Quiroz / 0009-0004-5625-8809

CORRESPONDENCE
David Guevara-Lazo
Jirón Huiracocha 2005, Jesús María, Lima, Perú.
+51 953522602.

EMAIL
david.guevara@upch.pe

CONFLICTS OF INTEREST
The authors do not report having any conflicts of interest

FINANCING
This study was self-funded by the authors.

AUTHOR CONTRIBUTIONS
D.G.L. and A.C.Q. contributed to the conception and design; D.G.L., N.N.A. and V.P.T. contributed to the review of literature and drafting the manuscript; D.G.L. contributed to the editing and the design of the figures; A.C.Q. contributed to the revision of key components of the manuscript. All authors approved the final version of the manuscript

PEER REVIEW
Received: 09/04/2024
Accepted: 15/09/2024

HOW TO CITE
Guevara-Lazo D, Palacios-Tealdo V, Nombera-Aznaran N, Calvo-Quiroz A. Imagen clínica: Eritema en Heliotropo, Pápulas de Gottron y Manos de Mecánico en una Mujer Peruana de 29 años con Dermatomiositis. Rev. Cuerpo Med. HNAAA [Internet]. 15 de noviembre de 2024 [citado 26 de marzo de 2025];17(3).
DOI: 10.35434/rcmhnaaa.2024.173.2452

Versión Impresa: ISSN: 2225-5109
Versión Electrónica: ISSN: 2227-4731
Cross Ref. DOI: 10.35434/rcmhnaaa
OJS: https://cmhnaaa.org.pe/ojs

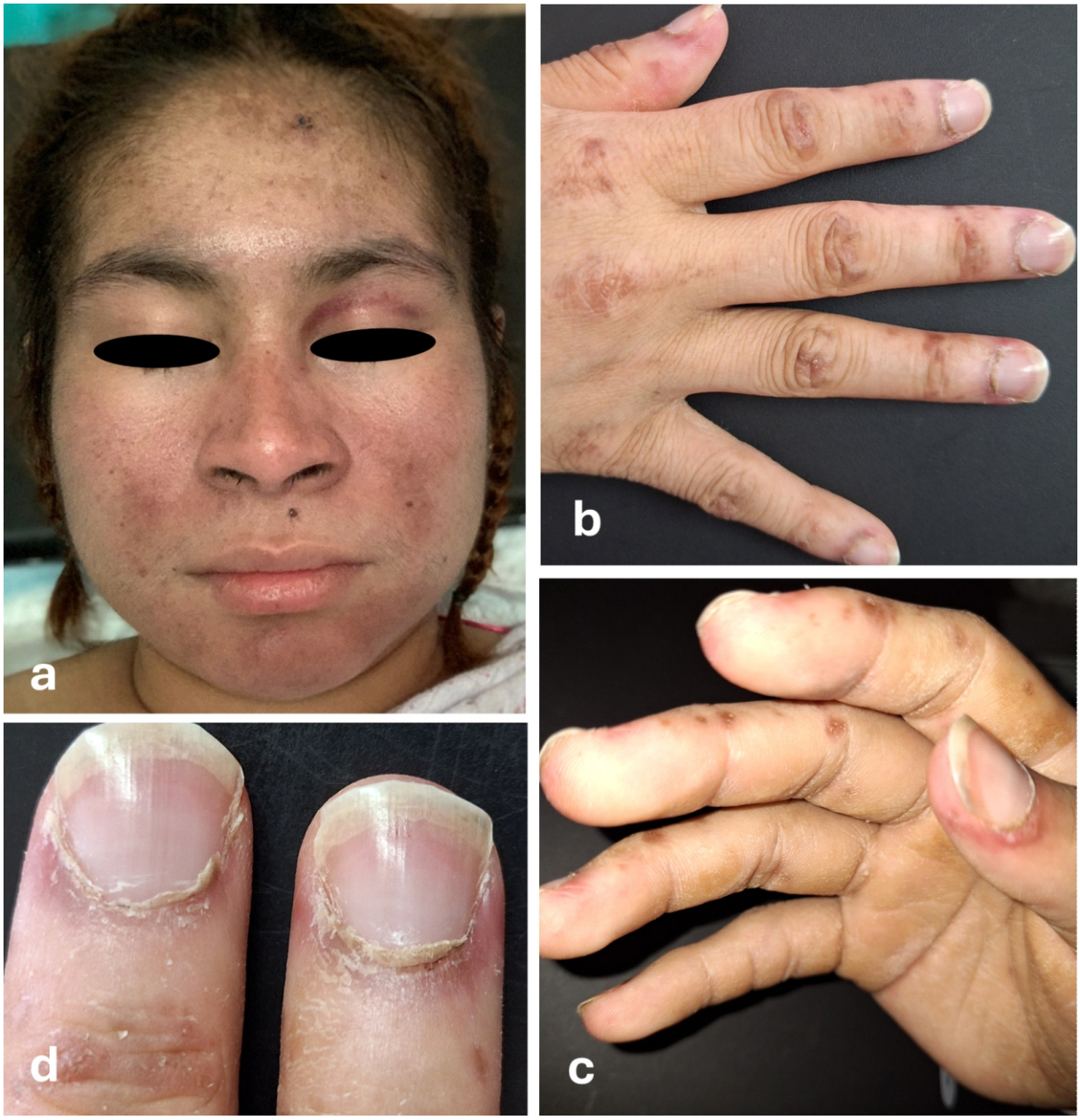

Figure 1.

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.

REFERENCES

1. Dalakas MC. Inflammatory muscle diseases. N Engl J Med. 2015;372(18):1734-1747. doi:10.1056/NEJMra1402225
2. Bitencourt N, Solow EB, Wright T, Bermas BL. Inflammatory myositis in systemic lupus erythematosus. Lupus. 2020;29(7):776-781. doi:10.1177/0961203320918021
3. Callen JP. Cutaneous manifestations of dermatomyositis and their management. Curr Rheumatol Rep. 2010;12(3):192-197. doi:10.1007/s11926-010-0100-7
4. Giannini M, Ellezam B, Leclair V, et al. Scleromyositis: A distinct novel entity within the systemic sclerosis and autoimmune myositis spectrum. Implications for care and pathogenesis. Front Immunol. 2023;13. Accessed January 18, 2024. https://www.frontiersin.org/articles/10.3389/fimmu.2022.974078