Reed D. Heliotrope Rash in Amyopathic Dermatomyositis. *DOM Clinical Insights*. 2026;1(2). doi:10.5070/V6.62998

Visual Diagnosis Pearl

Heliotrope Rash in Amyopathic Dermatomyositis

Diane Reed[1]

[1] Medicine, University of California, Los Angeles

https://doi.org/10.5070/V6.62998

UCLA Department of Medicine Clinical Insights

Vol. 1, Issue 2, 2026

An 82-year-old female with history of Parkinson's disease presented with a several-week history of a nonpruritic, nonpainful facial rash. Her physical examination revealed edematous, erythematous, scaly periorbital patches consistent with a heliotrope rash. No other rashes were noted on her physical examination. She reported subjective muscle weakness; however, strength testing in all extremities was noted to be 5/5. Her laboratory evaluation showed normal Creatine kinase (CK) and aldolase levels. A dermatomyositis panel was positive for anti–MDA5 antibodies (CADM-140), and skin biopsy findings were consistent with the diagnosis. The rash improved with topical tacrolimus. Rheumatology evaluation, in the absence of clinical or laboratory evidence of myopathy, confirmed a diagnosis of amyopathic dermatomyositis (AD). Patients who are positive for anti–MDA5 antibodies have a higher risk of developing interstitial lung disease (ILD). Given the antibody profile, evaluation for ILD with computed tomography (CT) imaging revealed no abnormalities. Age-appropriate malignancy screening was also unremarkable. Additional CT imaging of the chest, abdomen, and pelvis, performed for malignancy surveillance given the diagnosis of dermatomyositis, revealed no evidence of cancer.

DISCUSSION

Heliotrope rash results from a microangiopathy of the vascular endothelium and is initiated by a complement mediated cascade of inflammation eventually culminating in interface dermatitis and dermal edema that manifests as the erythematous or purple-violaceous rash seen in the above image (Image 1.) It is one of the hallmark clinical findings in patients with dermatomyositis. The latest evidence suggests that the process involves Janus kinase (JAK-1) mediated monocyte driven vasculopathy.[1]

Amyopathic dermatomyositis represents a small subset of dermatomyositis patients, estimated at 10–20%, who never develop muscle involvement.[2] The diagnosis can be made if a patient has only characteristic skin findings for six months or more, without evidence of muscle disease on laboratory or radiographic images. Of note, there are two subtypes of amyopathic dermatomyositis: true AD with no muscle weakness and normal laboratory findings and hypomyopathic dermatomyositis (HD), in which patients have no muscle weakness but have elevated CK and aldolase enzymes, abnormal electromyogram (EMG), abnormal magnetic resonance imaging (MRI), or muscle biopsy anomalies.[3]

Although these amyopathic dermatomyositis patients do not have the classical muscle involvement of dermatomyositis, they remain at elevated risk for malignancy and interstitial lung disease. Appropriate ongoing evaluation and monitoring are recommended.[2,4] Because AD patients who are positive for anti–MDA5 antibodies have a higher risk of developing not just ILD (13-44%) but a rapidly progressive form of the disease, regular screening is imperative. These recommendations are based on American College of Rheumatology (ACR) guidelines.[4] Strategies include ordering baseline high-resolution CT chest and pulmonary function tests (PFT) including spirometry, lung volumes, and diffusing capacity of the lung for carbon monoxide (DLCO) with ambulatory desaturation testing occurring at time of initial diagnosis. Follow-up screening with PFT's should occur every 3 to 6 months during the first year and every 6-12 months thereafter.

Even though AD patients positive for anti–MDA5 antibodies are at greater risk of ILD, they are also considered moderate risk for internal malignancy (14%). CT chest, abdomen, and pelvis, tumor markers CA-125, CA 19-9, carcinoembryonic antigen (CEA), mammography, pelvic ultrasound and cervical cancer screening at diagnosis then annually for 3 years are recommended. Surveillance extending to 5 years to capture 80% of cumulative cancer incidence is suggested by some experts. The most common malignancies associated with dermatomyositis include ovarian cancer, lung cancer, breast cancer, colorectal cancer, nasopharyngeal cancer (particularly in Asian populations), lymphomas, pancreatic and gastric cancers[3]

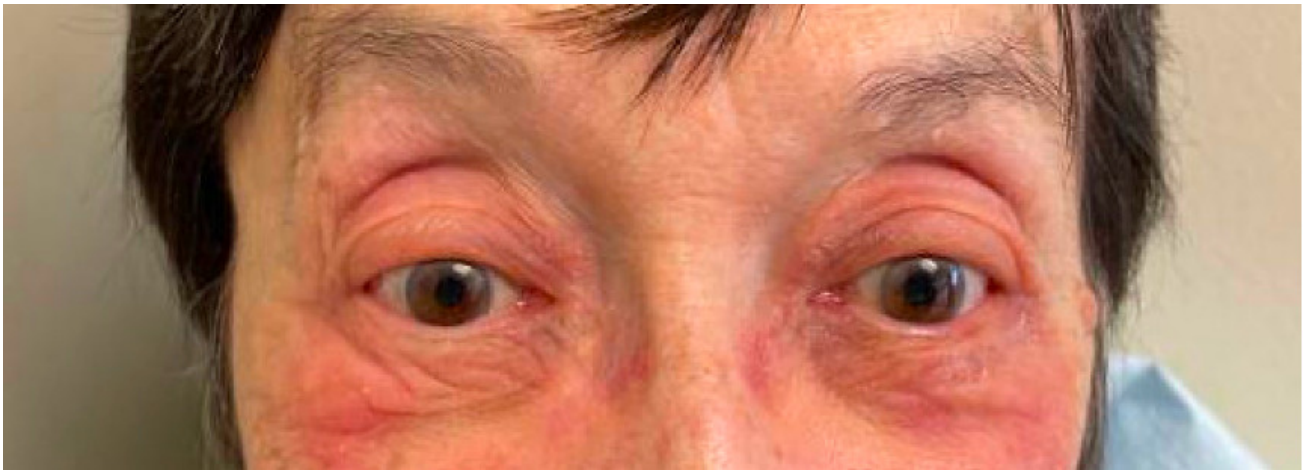

Image 1. interface dermatitis and dermal edema that manifests as an erythematous or purple-violaceous rash (Heliotrope) a patient with dermatomyositis.

REFERENCES

1. Osborne GA, Zhang L, Ma F, et al. Dermatomyositis is characterized by JAK1-mediated monocyte-driven vasculopathy and inflammation. *Sci Transl Med*. 2025;17(830):eaea9007. doi:10.1126/scitranslmed.aea9007. PMID:41442501

2. Caproni M, Cardinali C, Parodi A, et al. Amyopathic dermatomyositis: a review by the Italian Group of Immunodermatology. *Arch Dermatol (JAMA Dermatol)*. 2002;138(1):23-27. doi:10.1001/archderm.138.1.23

3. Fornaro M, Girolamo F, Giannini M, et al. Clinical, Histologic and Prognostic Features of Clinically Amyopathic Dermatomyositis. *Clinical and Experimental Rheumatology*. 2024;42(2):288-294. doi:10.55563/clinexprheumatol/kgpnbq

4. Selva-O'Callaghan A, Pinal-Fernandez I, Trallero-Araguás E, Milisenda JC, Grau-Junyent JM, Mammen AL. Classification and management of adult inflammatory myopathies. *Lancet Neurol*. 2018;17(9):816-828. doi:10.1016/S1474-4422(18)30254-0. PMID:30129477