Amyopathic Dermatomyositis With Plantar Keratoderma Responding to Methotrexate Therapy

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PRACTICE POINTS
- Dermatomyositis (DM) can present without muscular weakness as clinically amyopathic dermatomyositis (CADM).
- Clinically amyopathic dermatomyositis has cutaneous findings that can mimic other diseases including psoriasis.
- Clinically amyopathic dermatomyositis may have similar systemic associations as DM in general, such as an increased risk for malignancies.
- Treatments to consider for CADM should include systemic methotrexate.

Amyopathic dermatomyositis (ADM) represents a substantial subset of dermatomyositis (DM). Patients with this symptom of the disorder may present initially to a dermatologist. Amyopathic dermatomyositis shares cutaneous findings with DM and both overlap and differ with respect to other key disease characteristics, including autoantibody profile, associated lung disease, and malignancy risk. Plantar keratoderma is a rare skin finding in DM. We report a case of ADM uniquely marked by the finding of plantar keratoderma, which resolved with oral methotrexate therapy.


Case Report
A 54-year-old woman presented with a painful pruritic rash on the hands and feet of 7 years’ duration. She reported intermittent joint pain but denied muscle weakness. Physical examination revealed fissured fingertips and heavy scaling of the palms and lateral fingers (Figure 1). Violaceous scaly papules were seen on the distal and proximal interphalangeal joints (Figure 2). A severe plantar keratoderma also was noted (Figure 3). Pink scaly plaques were present on the bilateral elbows and postauricular skin. Diffuse mat telangiectases covered the malar skin. Extensive poikilodermatous skin changes covered approximately 20% of the total body surface area. Salt-and-pepper patches and papules were noted over the bilateral thighs. She reported an uncertain history of recent radiographs of one or both hands, which showed no joint degeneration characteristic of psoriatic arthritis. She previously had been given a diagnosis of psoriasis by an outside dermatologist but was not responding to topical therapy.

Several skin biopsies showed histologic evidence of dermatomyositis (DM)(Figure 4). Prominent basement thickening also was seen on periodic acid–Schiff staining (not shown). Laboratory workup showed negative antinuclear antibodies and anti–Jo-1, anti-Ku, and anti-Mi2 antibodies. Muscle enzymes including creatinine kinase and aldolase were within reference range. Pelvic ultrasonography and mammography were negative. Pulmonary function tests were unremarkable. High-resolution chest computed tomography (CT) was ordered because of a history of chronic cough; however, no evidence of malignancy or interstitial lung disease was seen. The patient was diagnosed with amyopathic dermatomyositis (ADM). Rheumatology was consulted and initiated oral hydroxychloroquine therapy. After 3 months, the patient’s cutaneous disease did not respond and she reported having headaches associated with this medication; therefore, methotrexate was started.

Within 2 months of treatment, full resolution of the plantar keratoderma (Figure 5) and clearance of the scaling/fissuring of the hands as well as the psoriatic-appearing plaques on the elbows was noted.

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The authors report no conflict of interest.

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Comment

Amyopathic DM is a subset of DM that accounts for 10% to 20% of DM cases. Sontheimer’s diagnostic criteria for ADM require histopathologic confirmation of the hallmark skin findings of classic DM and lack of muscle weakness or muscle enzyme (creatine kinase/aldolase) elevation for at least 2 years.

Similar to classic DM, ADM typically presents in the fifth decade of life and has a female predilection. The term hypomyopathic DM is used to describe patients who exhibit classic skin findings and evidence of muscle involvement on magnetic resonance imaging, electromyography, biopsy, or serum enzymes but have no clinical evidence of muscle weakness for at least 6 months. Together, hypomyopathic DM and ADM are referred to as clinically ADM (CADM). Patients who have met the criteria for hypomyopathic DM or ADM may later develop frank myopathy, progressing to a diagnosis of CADM, which may occur in as many as 10% to 13% of cases of CADM. Clinical evidence of muscle weakness typically is heralded by elevation of creatine kinase

FIGURE 1. Mechanic’s hands in amyopathic dermatomyositis with scaling of the lateral and volar surfaces of the digits as well as the palms.

FIGURE 2. Gottron papules in amyopathic dermatomyositis with scaling of the dorsal aspects of the interphalangeal joints with an underlying purplish erythema. Surrounding poikilodermatous changes were visible.

FIGURE 3. Plantar keratoderma with thick, white, hyperkeratotic plaques diffusely covering the sole.

FIGURE 4. A shave biopsy of the dorsal aspect of a proximal interphalangeal joint of the right hand with amyopathic dermatomyositis showed psoriasiform epidermal hyperplasia, a smudged dermoepidermal interface, and vacuolar alterations of basal layer (H&E, original magnification ×200).

FIGURE 5. Plantar keratoderma resolved after 2 months of treatment with oral methotrexate.
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and aldolase; therefore, patients with ADM should have muscle enzymes periodically checked.

Cutaneous findings of ADM are the same as the hallmark skin findings in CADM. Poikiloderma appears as thin telangiectatic skin in a background of mottled hyperpigmentation and hypopigmentation. It represents chronic inflammation and often occurs in sun-exposed areas. Poikiloderma located on the posterior neck and shoulders is known as the shawl sign and on the lateral thighs as the holster sign. The term mechanic’s hands is used to describe the clinical finding of palmar erythema with scaling and fissuring of the fingertips. Scalp findings include erythematous, atrophic, scaly plaques resembling psoriasis and nonscarring alopecia. Gottron papules are nearly pathognomonic for DM. These violaceous papules often are pruritic and found over the finger joints, in contrast to the hand rash of lupus erythematosus that involves the skin between finger joints. Psoriatic-appearing plaques overlying the elbows and knees are known as Gottron sign and can contribute to misdiagnosis as psoriasis. The classic heliotrope rash presents as a violaceous hue in the periorbital area and may be associated with periorbital edema. Calcinosis cutis is common in CADM but rarely is reported in ADM. Nail findings include periangual hypereremia, cuticular overgrowth, and nail bed changes due to avascular areas and dilated capillaries. The cutaneous histopathologic findings in ADM are the same as with CADM: a smudged dermoeipidermal interface, vacuolar alterations of the basal layer, and dermal mucin deposits.

Palmoplantar keratoderma rarely is reported as a cutaneous finding in DM. The finding of keratoderma has mainly been reported in association with Wong-type DM, a rare subtype of DM with features of pityriasis rubra pilaris. Palmoplantar keratoderma also has been reported in a case of an ADM-like hydroxyurea-induced eruption and as an early presenting feature in one patient with CADM and one with juvenile DM.

The autoantibody profile in patients with ADM varies from that of CADM and can be helpful in both diagnosis and prognosis. Similar to CADM, the majority of patients with ADM have positive antinuclear antibodies. Anti–Jo-1 (an anti–aminoacyl-transfer RNA synthetase) antibody frequently is found in CADM but rarely in ADM. Anti–Jo-1 is predictive of interstitial lung disease (ILD) in CADM. Positive anti–Jo-1 in combination with Raynaud phenomenon and mechanic’s hands is referred to as antisynthetase syndrome in patients with CADM. An antibody uniquely linked with CADM is the anti–CADM-140/MDA5 antibody and can be a marker of rapidly progressing ILD in these patients.

Several therapies have been found useful in ADM. Because lesions often are photoexacerbated, sun protection is essential. Antimalarials such as hydroxychloroquine are considered first-line therapy. Clinicians must be aware of 2 possible hydroxychloroquine side effects that can uniquely confuse the clinical picture in ADM. The first is a rash, most often morbilliform and pruritic, that occurs in DM more frequently than in other diseases. The second is a myopathy found in as many as 6.7% of patients using antimalarials for rheumatic disease, which can clinically mimic the progression of ADM to CADM.
Two small retrospective case series found that methotrexate was beneficial in ADM. Methotrexate also has been reported as an efficacious treatment of ILD in patients with connective tissue diseases. Intravenous immunoglobulin and other immunosuppressants are additional agents to be considered.

In summary, ADM is an important subset of DM and is more likely to present to dermatology practices than to other specialists. Amyopathic DM shares cutaneous findings with DM, and both overlap and differ with respect to key disease characteristics including autoantibody profile, associated lung disease, and malignancy risk. Palmoplantar keratoderma is a rarely reported skin finding in DM. We report a case of ADM with the unique finding of severe plantar keratoderma. The fact that our patient’s keratoderma and other skin findings resolved concomitantly during methotrexate therapy leads us to believe that the keratoderma was a unique skin manifestation of the ADM itself.

REFERENCES


