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Images in Rheumatology

Ulcerative Paraneoplastic Dermatomyositis in the Setting of Positive Transcriptional Intermediary Factor 1-γ Antibody

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Dermatomyositis (DM) is an autoimmune inflammatory disease of the skin with or without muscle involvement[1]. One-third of patients present with malignant disease within 3 years[2].

A 60-year-old woman with a remote history of ovarian cancer presented with a 1-month history of severe dysphagia, symmetric proximal muscle weakness, and a progressive painful ulcerative skin eruption associated with her second recurrence of metastatic ovarian cancer treated 4 years ago. Physical examination revealed 4/5 proximal muscle weakness, Gottron papules, heliotrope rash, and multiple irregular ulcers with overlying eschars (Figure 1A). Serum creatine kinase was normal, while liver function tests and aldolase were mildly elevated. Serology showed positive transcriptional intermediary factor 1-γ (TIF1-γ) and negative melanoma differentiation–associated protein 5 (MDA-5) antibodies. Electromyography confirmed proximal myopathy. Skin biopsy revealed interface dermatitis with underlying dermal mucinosis and pigment incontinence (Figure 2). High-dose pulse corticosteroids and intravenous Ig improved the myositis and arrested the spread of the ulcers (Figure 1B).

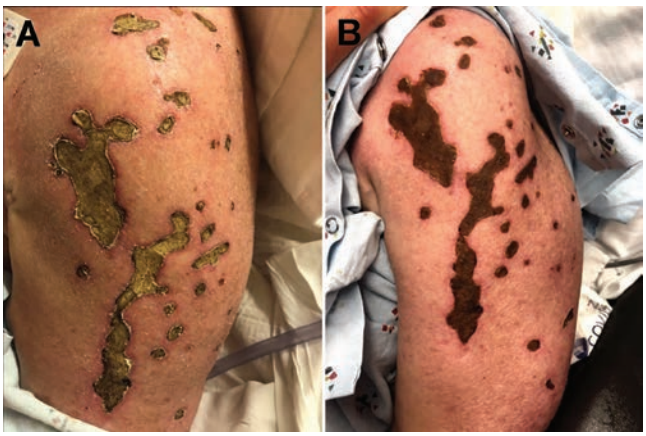

Figure 1. (A) Multiple well-defined geographic peripheral erythematous ulcers with adherent crust. (B) Ulcer expansion arrested after immunosuppressive therapy.

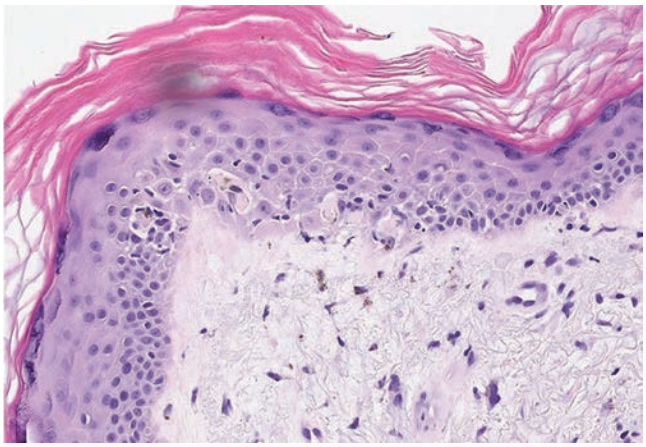

Figure 2. Extensive vacuolar interface dermatitis with focal epidermal dyskeratosis. H&E stain, 40× magnification.

TIF1-γ antibodies are associated with a DM phenotype of myositis and extensive skin involvement that may include classic heliotropic erythema, psoriasiform plaques, or near erythroderma. Ulcerated and eroded skin lesions can be seen with anti-MDA-5 DM, often in association with interstitial lung disease; they are also a well-described finding in paraneoplastic DM. Paradoxically, despite the high risk of malignancy in older patients with TIF1-γ autoantibodies, skin ulceration is phenotypically uncommon in comparison with classic adult-onset cancer-associated DM presentations[3,4,5].

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