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Images in Rheumatology

Symmetrical cutaneous ulcers: Are they associated with severe disease in children with juvenile dermatomyositis?

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A 4-year-old girl presented with difficulty in climbing stairs, getting up from supine position, and raising arms above the shoulder for 1.5 years. She also reported rash over face and dorsum of hands. There was worsening of symptoms in the last 1 month with fever and difficulty in holding neck. Physical examination revealed a heliotrope rash (Figure 1a); healed Gottron papules over bilateral metacarpo-phalangeal and proximal interphalangeal joints; weakness of the proximal limb, trunk, abdomen, and neck flexor muscles; and weak gag reflex. Symmetrical punched-out ulcers overlying bilateral scapula were also noted (Figure 1b). However, no calcinosis was noted. Laboratory investigations revealed the following: hemoglobin, 105 g/L; white blood cell count, 12.9 × 10^9/L; platelet count, 352 x 10^9/L; C-reactive protein, 3.4 mg/L; erythrocyte sedimentation rate, 68 mm in first hour; alanine aminotransferase, 70 IU/L; aspartate aminotransferase, 64 IU/L; lactate dehydrogenase, 561 U/L; and creatine kinase, 73.8 IU/L. Antinuclear antibody by indirect

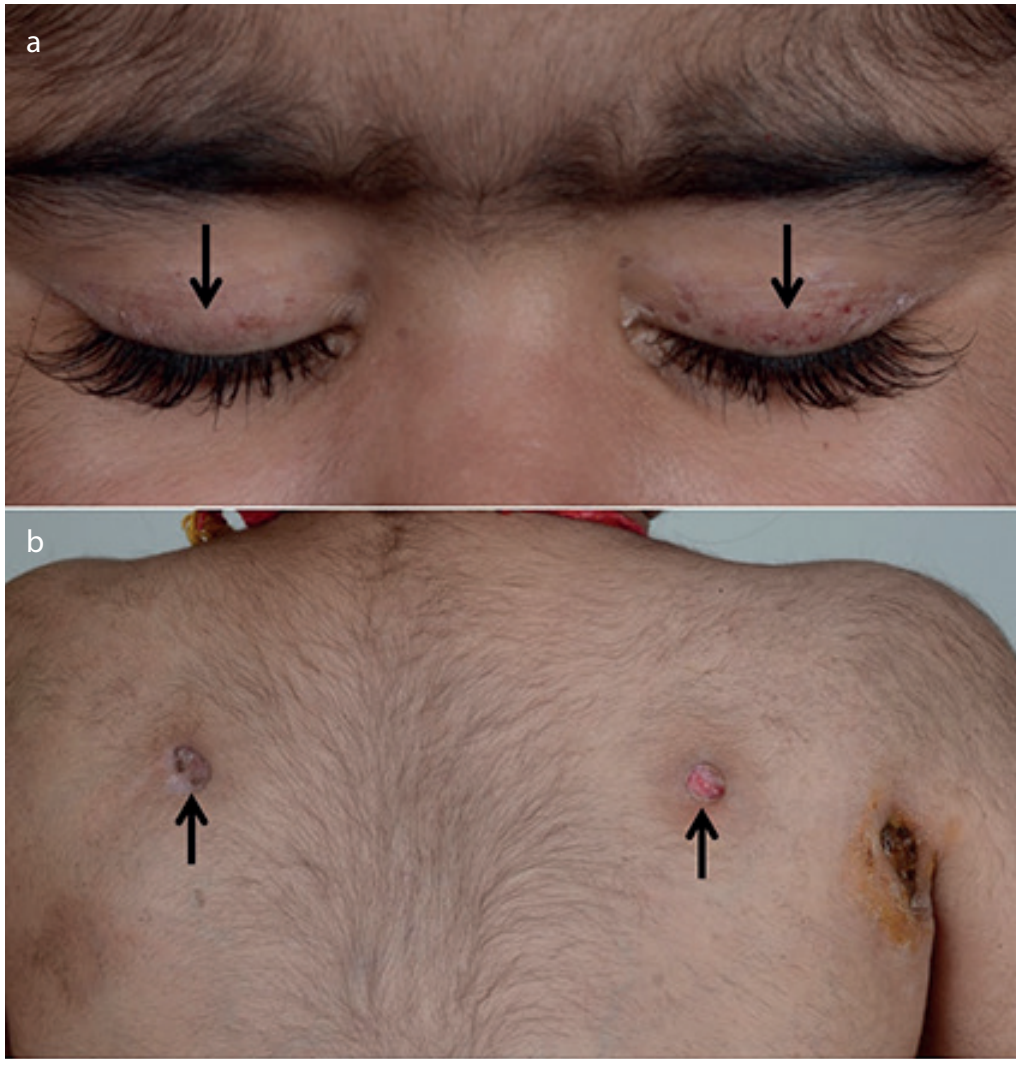

Figure 1. a, b. (a) Heliotrope rash and Symmetrical punched-out ulcers overlying bilateral scapula in a girl with juvenile dermatomyositis (b).

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immunofluorescence was negative. Magnetic resonance imaging revealed diffuse hyperintensities involving bilateral thigh muscles. She was diagnosed with juvenile dermatomyositis (JDM) and received five doses of intravenous methylprednisolone followed by tapering doses of oral prednisolone and subcutaneous methotrexate. Her symptoms persisted, and she received three doses of monthly intravenous immunoglobulin (1 g/kg body weight), intravenous cyclophosphamide (500 mg/m^2 body surface area/month) for six doses, and mycophenolate mofetil (1000 mg/m2 body surface area/day). Informed written consent was obtained from the caregiver.

JDM is the most common childhood inflammatory myopathy. Cutaneous ulcers occur in 5%-30% of children with JDM and are often associated with severe disease (1). These are associated with significant pain and can develop secondary bacterial infection. In children with JDM and cutaneous ulcers, the clinical course is marked by frequent muscle and skin relapses (2). These children show unresponsiveness to routine medical management and often require multiple immunosuppressants for disease control (1, 3). Development of cutaneous ulcers in patients with JDM should make the treating physician alert regarding a severe disease course in these children.

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