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Juvenile amyopathic dermatomyositis – Systemic lupus erythematosus overlap syndrome

Jüvenil amiyopatik dermatomiyozit – Sistemik lupus eritematozus örtüşme sendromu

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Dear Editor,

A 10-year-old child referred to our Rheumatology clinic with erythematous, desquamating skin lesions and arthralgia, mainly in the elbow and wrist joints. He did not complain of myalgias, muscle weakness or dyspnea. He had history of epilepsy for two years and nephrotic syndrome for five years. The patient denied photosensitivity, hair loss, oral ulcers, and Raynaud's phenomenon. There was no known family history of autoimmune disease. On examination, he had slight heliotropic periorbital rash, erythematous elbows, and desquamating papules (**Figure 1**). No muscle weakness was noted. The patient used the drug (Levetiracetam 500 mg 2×1 / Deltacortil 5 mg 2×1 posology) for epilepsy and nephrotic syndrome. Laboratory: Complete blood counts were normal. Erythrocyte sedimentation rate was 16 mm/h (normal range 0–20 mm/h), C-reactive protein was 0.78 mg/dl, aspartate aminotransferase was 16 IU/L, alanine aminotransferase was 22 IU/L, creatine kinase was 30 IU/L, myoglobin was 25 IU/L, lactate dehydrogenase was 65 IU/L, albumin was 2.5 g/dL, high antinuclear antibody positivity was (4+) and anti-double-stranded DNA (anti-ds DNA) antibodies positive and anticardiolipin antibodies IgG was 120 IU/L. Anti-Jo-1 antibody was negative. Serum complement levels were low with a C3 of 36 mg/dL (normal range: 101–186 mg/dL) and C4 of 6.7 mg/dL (normal range: 16–47 mg/dL). Urinalysis: Kreatinin; 0.6 mg/dl, showed 3+ proteinuria and 24-h urinary protein to creatinine ratio of 7.1 (>3 significant). So renal biopsy was done, which showed WHO stage IV histological-type diffuse glomerulonephritis with mesangial and subendothelial deposits. Our patient had no respiratory symptoms; a chest X-ray was performed but showed no pulmonary involvement. Electromyography was normal and was not indicative of a myopathic pattern. Hand cutaneous biopsy showed a vacuolar alteration of the basal cell layer of the epidermis, necrotic keratinocytes (apoptosis), vascular dilatation, and a sparse, superficial, perivascular lymphocytic infiltrate; immunofluorescence did not show IgG, IgA, IgM, C3 or C4 deposits. The diagnosis of juvenile amyopathic dermatomyositis (ADM) -SLE overlap syndrome was established based on the positive clinical and immunologic findings. The patient satisfied 4 of the 17 criteria of Systemic Lupus International Collaborating Clinics (SLICC) for classifying SLE namely epilepsy, nephrotic syndrome, low serum complement levels, and positive serum antibodies. ADM was

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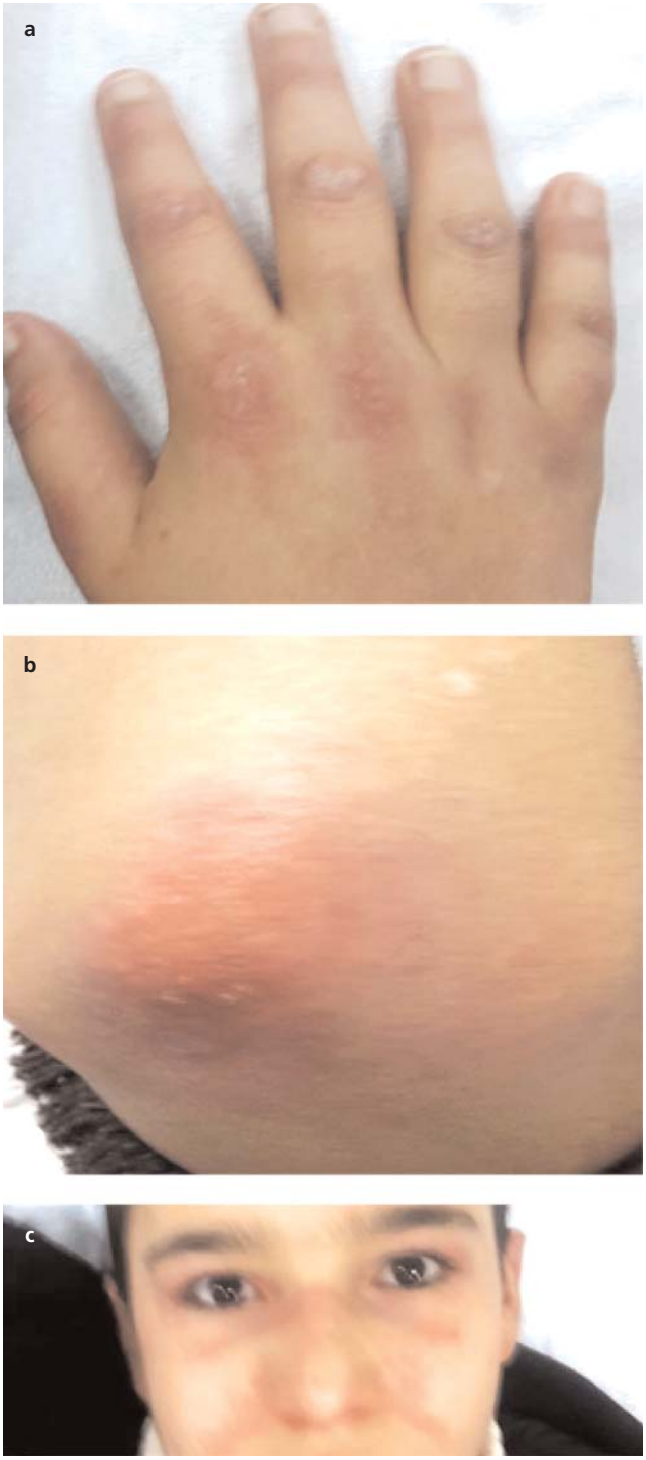

Figure 1. Pathognomonic manifestations of dermatomyositis. (**a**) Gottron's papules overlying the dorsal interphalangeal joints. (**b**) Gottron's papules on the elbow. (**c**) Heliotrope: a violaceous eruption with periorbital edema. [Used with permission of the patient].

diagnosed with patient's pathognomonic skin changes and skin biopsy specimen findings without clinical or laboratory evidence of muscle involvement with Euwer and Sontheimer published four diagnostic criteria. He was screened and investigated for associated malignancies. Thoracoabdominopelvic computed tomography and abdominal ultrasonography were normal. Systemic glucocorticoid therapy was instituted (0.5 mg/kg once daily) accompanied by weekly methotrexate (15-20 mg/m^2). At the follow up visit, he continued to do well and was enrolled in clinic for long-term management of ADM-SLE overlap syndrome.

ADM is a rare but distinct subtype of dermatomyositis. It presents clinically with the pathognomonic cutaneous manifestations of dermatomyositis (consisting of heliotrope rash, facial erythema and edema, Gottron's papules and periungal telangiectasia) but without associated skeletal muscle involvement. The three main factors predicting prognosis in ADM patients are the following: development of lung disease, development of malignancy, and development of clinical muscle weakness.[1]

A number of patients with dermatomyositis also meet the criteria for one of the connective tissue disorders. To be a true overlap syndrome, the patient must meet the diagnostic criteria for each separate disorder. Eleven to 40 percent of patients with dermatomyositis have been reported to have a concomitant diagnosis of a connective tissue disorder. These disorders include rheumatoid arthritis, scleroderma, systemic lupus erythematosus, Sjögren's syndrome, polyarteritis nodosa and mixed connective tissue disease, and many others.[2] Investigations for a malignancy should be done in all patients with dermatomyositis, including those with the amyopathic form. Ovarian and breast carcinomas in women, lung and prostate carcinomas in men, and lymphoma in both genders are highly associated with dermatomyositis.[3,4]

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