PAEDIATRIC DERMATOLOGY

WONG-TYPE DERMATOMYOSITIS IN 2 PACIENTS

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Background: Dermatomyositis is an inflammatory myopathy of proximal muscles with classic cutaneous findings usually consisting of heliotrope erythema, Gottron's papules over bony prominences, and periungual telangiectasias. Wong- Type dermatomyositis refers to a small subgroup of these patients who have additional clinical features of pityriasis rubra pilaris (PRP).
There are few reported cases in the literature.

Observation: Pacient 1: An 8-year-old girl presented erythematous-squamous lesions on the palms and soles associated with heliotrope erythema who referred a history of asthenia and muscular weakness of the last 2 months. A skin biopsy compatible with Pityriasis Rubra Pilaris was performed. The blood tests showed an increased lactate dehydrogenase (LDH), ASAT, aldolase, creatine phosphokinase (CPK) and a positive anti-ADN antibody (1:80). The muscle biopsy revealed myositis while the MRI reported diffuse and bilateral interfascial edema. Therefore, the diagnosis of Wong-type dermatomyositis was made. A treatment with systemic steroids and methotrexate (MTX) was started, with a favorable clinical evolution.
Pacient 2: A 15-year-old woman, with a history of Raynaud syndrome, consulted for erythematous-squamous lesions on palms and soles associated with heliotrope erythema. In addition, she referred marked asthenia denying muscular weakness while the blood tests revealed an increased LDH, aldolase and CPK. An MRI was performed, despite it was not conclusive, showed an inflammatory muscular involvement. We decided to perform a cutaneous biopsy which it was compatible with pityriasis rubra pilaris. Consequently, we started treatment with MTX and topical corticosteroids with good response.

Key message: We decided to present 2 patients with an extremely unusual clinical entity, Wong- type dermatomyositis, in which the overlap of clinical features between DM and PRP is manifested. We believe that an early diagnosis and rapid onset of treatment in the second patient may improve the prognosis of her disease