



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

WONG-TYPE DERMATOMYOSITIS, A CASE REPORT WITH GOOD CLINICAL RESPONSE

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Background: A very unusual finding has been reported in little more than 20 cases of Dermatomyositis (DM) in the world, corresponding to the presence of follicular hyperkeratotic papules that resemble pityriasis rubra pilaris, associated with clinical and histological findings compatible with DM. This entity has been called Wong-type DM. The aim of this report is to present a case of Wong variant and describe patient evolution with the treatment.

Observation: A 62-year-old man, without significant comorbidities, present with a history of 6 months of asymptomatic erythematous plaques. Denies myalgia, arthralgia and loss of proximal limb strength. Physical examination revealed erythematous-violaceous periocular and malar spots, as well as poikiloderma in the anterior and posterior cervical area. On both flanks, shoulders and lateral aspect of the thighs, hyperkeratotic erythematous follicular papules appear over the erythematous-violaceous surface. The diagnosis of amyopathic DM was suspected and laboratories was requested, obtaining ANA 1/512 mottled pattern, with all the rest of laboratory parameters normal. Paraneoplastic study was negative. Given the atypical presentation of pityriasis rubra pilaris type lesions, skin biopsy of the flanks and shoulders was obtained, resulting compatible with interface dermatitis associated with areas of hyperorthokeratosis and follicular plugs. The diagnosis of Wong-type DM was made and treatment was started with 60 mg per day of prednisone on decreasing doses and methotrexate 12.5 mg weekly, with clinical improvement at the month 2 after the start of treatment, and significant decrease of lesions at month 4. At 6 months of follow-up, the patient is only on methotrexate and there is an almost complete disappearance of the lesions, without recurrences.

Key message: Wong variant is a recently described variant of DM, with few cases reported in the world. This case describes the clinical presentation of a patient with this entity and the evolution with the treatment.

