

Efficacy of dapsona in two cases of amyopathic dermatomyositis*

*Eficácia da dapsona em dois casos de dermatomiosite amiopática**

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Abstract: Dapsone is a drug primarily used for the treatment of Hansen's disease, and it has also been employed in cases of bullous lupus erythematosus and some types of cutaneous vasculitis. Recently, its efficacy in the treatment of cutaneous lesions in dermatomyositis has been observed. We present two cases of dermatomyositis, amyopathic form, which were refractory to habitual treatment, but had an excellent response to dapsona therapy.

Keywords: Autoimmune diseases; Dapsone; Dermatomyositis

Resumo: A dapsona é uma droga utilizada no tratamento da hanseníase que vem sendo empregada em casos de lúpus eritematoso bolhoso e alguns tipos de vasculites cutâneas. Recentemente, foi observada sua eficácia no tratamento de lesões cutâneas da dermatomiosite. São apresentados dois casos de dermatomiosite, forma amiopática, refratários às medicações habituais, em que o uso de dapsona foi responsável pelo controle das lesões cutâneas.

Palavras-chave: Dapsona; Dermatomiosite; Doenças auto-imunes

Cutaneous manifestations of dermatomyositis (DM) often persist after therapy with steroids, immunosuppressing agents, antimalarian drugs or their combination.^{1,2} For half a century, dapsona has been used for its anti-inflammatory properties, particularly aimed at leukocytes, for the treatment of Hansen's Disease and autoimmune diseases, such as bullous lupus erythematosus and relapsing poly-chondritis. Literature has shown some reports of DM with good response of its cutaneous manifestations to the use of dapsona. We hereby present two patients with refractory amyopathic dermatomyositis, who responded to treatment with dapsona.

Case 1

Fifty five-year-old white male patient, who presented, four years ago, erythematous pruritic cutaneous eruptions, with the finding of photosensitivity in the face and upper trunk, shawl Sign and V-Sign on

physical examination, besides heliotrope and Gottron's Sign. Muscle strength was preserved (degree: 5+/5+). Histopathological study of the skin revealed an epidermis with discrete acanthosis, edematous dermis with mononuclear infiltrate surrounding vessels and annexes, besides decrease of pilous follicles. Antinuclear factor (ANF) was negative, and Creatinophosphokinase (CPK) values were within normal range. No muscle biopsies or electroneuromyography (ENMG) were carried out. The case was diagnosed as amyopathic dermatomyositis, and treatment with chloroquine diphosphate at 250 mg/day was begun, without satisfactory response. Dapsone at 50 mg/day was then introduced and after two months the dose was built up to 100 mg/day, with a significant improvement. Later on, patient had the onset of an insidious ventilatory-dependent thoracic pain associated with weight loss, with the finding, on tomographic investigation, of a pulmonary nodule, the histopatho-

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Conflict of interest: None

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logical examination of which revealed an adenocarcinoma.

Case 2

Forty three-year-old white female patient, with the onset, 7 years previously, of a dermatological picture characterized by an erythematous eruption, with findings on clinical examination of photosensitivity in the face, arms and cervical regions (Shawl Sign). Face eruption was diffuse, with heliotrope. Patient also presented Gottron's Sign in metacarpal-phalangeal and proximal interphalangeal joints, and slight alopecia. There was not, in any moment, any complaint of muscle weakness, with the observation of preserved muscle strength on physical examination (degree: 5+/5+). ANF was positive at 1:160 dilution, with a fine speckled pattern. Anti-SSA and anti-SSB antibodies were negative. Histopathological examination revealed normal muscles, and overlying skin likewise. Several CPK and aldolase assays were performed during follow-up, with all values within normal ranges. No histopathological examinations of the affected skin or ENMGs were carried out. The case was diagnosed as amyopathic dermatomyositis, and treatment was initiated with prednisone at 5 mg/day and hydroxychloroquine at 400 mg/day, which had no efficacy for the improvement of the cutaneous picture. Dapsone was then begun at 50 mg/day, and dose increased to 100 mg/day after two months, as no satisfactory results had been obtained with lower dosages. After the increase to 100mg, patient obtained a progressive improvement in the course of the following 12 months, and the medication was then withdrawn with no relapses.

Amyopathic dermatomyositis can make up to

10% of DM cases, and it is defined as an autoimmune disease with typical manifestations of DM, with no clinical evidences for proximal muscle weakness and with normal muscle enzymes.³ Treatment of DM cutaneous manifestations with antimalarian and immunosuppressing drugs is not always efficacious. Dapsone (4,4'-diaminodiphenylsulphone) has an anti-inflammatory action, for inhibiting polymorphonuclear leukocytes and the activation of the alternative pathway of the complement cascade.⁴ Having been first used in the treatment of Hansen's Disease, it has also been used as a second-line agent in the treatment of cutaneous manifestations of autoimmune diseases, such as cutaneous lupus,⁵ cutaneous vasculites, bullous dermatitis and herpes-like dermatitis.¹ In 1994, Konohana e Kawashima reported, in a patient with a classical refractory dermatomyositis, an improvement of cutaneous eruption and myositis, with decrease in CPK serum levels after two weeks of daily use of 75 mg of dapsone.⁶ Later on, Cohen described a satisfactory response to dapsone in two patients who had been diagnosed with oligomyopathic and amyopathic dermatomyosites, the treatment of which has been inefficacious with corticosteroids and immunosuppressing agents.² In both cases presented here of chloroquine-refractory amyopathic dermatomyositis, after excluding the possibility of G-6-PD deficiency, we were able to prove the efficacy of such drug for this condition, even in a patient who had DM probably secondarily to a neoplasia. These data allow us to conclude that dapsone can be used for the treatment of DM cutaneous manifestations which do not respond to therapy with chloroquine. □

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