



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

PANNICULITIS ASSOCIATED WITH AMYOPATHIC DERMATOMYOSITIS

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Background: Dermatomyositis is an autoimmune, chronic, idiopathic, inflammatory disease that affects the striated skeletal muscles, the skin and other organs. Amyopathic dermatomyositis constitutes 2-18% of patients with dermatomyositis and is characterized by pathognomonic cutaneous lesions of dermatomyositis, histopathology compatible with the disease and no evidence of muscle disease. Panniculitis is a rare skin manifestation of dermatomyositis and less than 30 cases of such association have been published since 1924, of which only one reported such association with the amyopathic subtype. In cases reported so far, the presence of panniculitis was associated with a better response to treatment and good prognosis in patients with dermatomyositis.

Observation: We report the case of a 49-year-old female patient with erythematous papules on metacarpophalangeal joints and violaceous erythema on the face, cervical region, upper back and upper limbs. The patient also showed progressive dyspnea, but did not show muscle weakness. After further investigation, the diagnosis of amyopathic dermatomyositis with pulmonary involvement was made. Screening for neoplasm was negative. One year after the patient's first symptoms, indurated, painful, violaceous, erythematous, subcutaneous nodules appeared on the upper and lower limbs, back, breasts and abdomen. Skin biopsy of the left inner thigh revealed lymphocytic panniculitis with plasma cells of a lobular pattern and vasculopathy. Treatment with prednisone 1mg/kg/day associated with chlorambucil was introduced but with poor responsive. The patient was also treated with pulse of methylprednisolone and then with methotrexate, but her skin manifestations of panniculitis remained unresponsive to treatment.

Key message: The association between dermatomyositis and lobular panniculitis is rare, with few cases reported in literature. Although, in other reports, panniculitis skin manifestations responded well to treatment, in some cases, like in our patient, lesions can be resistant.

