Severe adult dermatomyositis with unusual calcinosis

We present the case of a 49-year-old woman who presented with DM according to Bohan and Peter criteria: heliotrope rash, Gottron’s papules, muscle weakness, elevated creatine kinase, myopathic changes (electromyography) and perifascicular atrophy with perimysial cellular infiltrates (muscle biopsy). A 2-year remission was obtained after successive treatment, including steroids, CYC, IVIG and MTX. After tapering treatment, the patient relapsed with myalgia and muscle weakness. On examination there were large areas of painful inflammatory skin blotches (Fig. 1A) that were radio-opaque (Fig. 1B and C). Biopsy confirmed panniculitis (Fig. 1D) with adipocyte necrosis, septal inflammatory cells and calcinosis. There was no evidence of malignancy or interstitial lung disease. Anti-NXP2 autoantibodies were detected (linear dot). Steroids, monthly IVIG and MTX rapidly improved muscular symptoms, inflammation of the skin and, to a lesser extent, calcinosis.

This case stresses the usefulness of myositis-specific antibodies to classify myositis into subgroups. Anti-NXP2 antibodies are rare autoantibodies, mostly found in JDM [1]. They are strongly associated with skin extended calcifications. The treatment of calcinosis cuts, which may also occur in other autoimmune diseases, is not consensual, but is based on the treatment of the underlying disease [2]. In the current case, the immunosuppressive drugs improved both the panniculitis and the calcinosis.

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