Clinical vignette

Anti-NXP2 antibody-associated extensive subcutaneous calcinosis in adult-onset myositis

A 68-year-old woman was followed since 1988 for DM. A combined therapy of corticosteroids and MTX controlled muscle involvement. In 1993, multiple subcutaneous nodules of calcinosis appeared on the limbs, chest and abdomen. In 2013, extensive calcinosis severely impaired the patient's quality of life while myositis was under control (Fig 1). In 2016, a blood sample was collected in which circulating anti-NXP2 antibodies were detected.

Anti-NXP2 antibodies were first identified in 1997. Today, 25% of patients with juvenile DM demonstrate these autoantibodies in their blood samples, indicating a poor prognosis [1]. Anti-NXP2 antibodies are detected in only 1% of cases of adult patients with myositis, and are likely linked to the occurrence of neoplasms [2]. Therefore, although typical in paediatric patients, an extensive form of calcinosis in an adult case of anti-NXP2 antibody-associated myositis is extremely rare. In our case, the evolution of calcinosis is independent from muscle involvement. Furthermore, MTX, corticosteroids and IVIGs were inefficient in preventing the spreading of calcinosis. Currently, a topical sodium thiosulphate treatment is being trialled.

Thus, an early detection of anti-NXP2 antibodies could help identify a subset of adult patients at risk of developing calcinosis during the early stages of DM.

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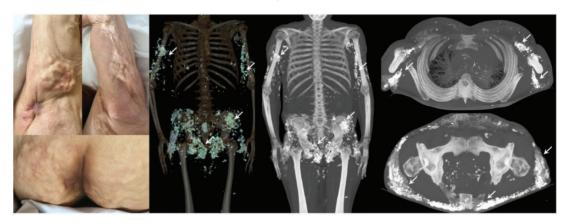
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Total body CT imaging reconstruction (bone-window settings).

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