EXTENSIVE CALCINOSIS IN JUVENILE DERMATOMYOSITIS

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Calcinosis is common in systemic autoimmune diseases such as juvenile dermatomyositis (JDM) or scleroderma[1]. The authors describe a 19-years-old white male with JDM diagnosed 3 years earlier because of a progressive appearance of arthralgias, proximal muscle weakness and tenderness, heliotrope rash and Gottron papules with electromyogram and muscle biopsy (deltoid) confirming the inflammatory myopathy. The patient was treated with prednisone (60 mg daily), azathioprine (150 mg daily) and hydroxychloroquine (200 mg daily), achieving clinical improvement. In the follow up, the patient presented multiple stony and tender subcutaneous nodules, localized in the extensor surfaces of both thighs, with progressive distal extension. Clinical examination revealed a large stony conglomerated mass that extended to the whole antero-external surface of the left thigh. In the right thigh, there were two isolated and non-fixed stony lumps (4 x 3 cm and 3 x 2 cm of diameter) at the external and internal surface, respectively. Serum creatinine, calcemia, urinary calcium, erythrocyte sedimentation rate, C-reactive protein, liver profile and creatine phosphokinase were within normal ranges.

Radiography and computed axial tomography scan of the lower limbs showed areas of homogeneous opacity with bony density compatible with calcium deposits in cutaneous and subcutaneous tissues, extended to the fascia in the left thigh (Figure 1). Bone scan with 99mTc demonstrated an extensive anomalous distribution of radiotracer in the soft tissues, affecting almost the entire left thigh and two circumscribed hyperintense areas in the right thigh (Figure 1). Once established the extension of calcinosis, medical treatment was started with aluminum hydroxide (30 ml 2 hours after meals), diltiazem (180 mg daily) and weekly alendronate (70 mg). After 4 months as there was no clinical improvement a low-dose warfarin (1 mg daily) was added to the treatment. After twelve months of warfarin treatment, extensive calcinosis of the left thigh decreased its surface extension, improving pain and tenderness. However, large accumulations of calcium tissue (tumoral calcinosis) showed no clinical significant changes. There were no new areas of calcinosis or local complications in pre-existing injuries.

Calcinosis cutis is a late complication of JDM, present in 20 – 70% of cases in the juvenile form[2]. It s a disorder characterized by hydroxyapatite crystals and amorphous calcium phosphates deposited in the skin and soft tissues[1,2]. In the reported case, we found ectopic calcium deposits on the skin (calcinosis cutis) and bulky subcutaneous accumulations (tumoral calcinosis) on the muscle fascia, all clinical presentations of a single pathogenic phenomenon. In the treatment of calcinosis, there is no

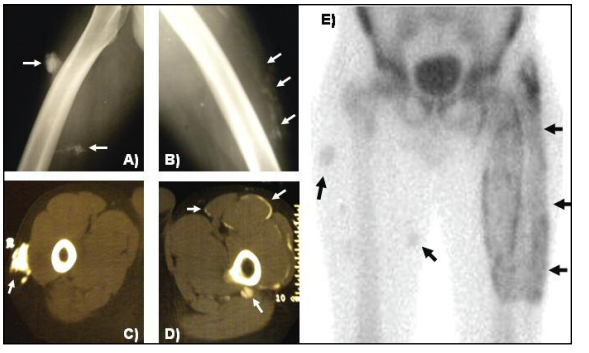

Figure 1. Identification of calcinosis in left and right thighs with different imaging methods. Lateral radiographs of the right (**A**) and left (**B**) thighs show deep and diffuse calcinosis respectively. Computed tomography of the thighs showing tumoral calcinosis in right leg (**C**) and diffuse calcifications along fascial planes of the muscles in left leg (**D**). Bone scan (**E**) shows increased soft-tissue uptake of the radiotracer with deep deposits of calcium in the right thigh and extensive subcutaneous calcinosis that spreads over almost the entire left thigh.

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evidence based on controlled studies that any of the pharmacological treatments would result in clinical benefits to establish therapeutic recommendations[1]. Different results were reported with the use of aluminium hydroxide antacids, bisphosphonates and diltiazem[1-5]. There seems to be some evidence in favour of using low-dose warfarin to treat calcinosis in systemic autoimmune diseases[5,6]. In this case, the long-term treatment with low doses of warfarin, seemed beneficial in the treatment of early and mild calcinosis but was ineffective on the injury previously established or in the large accumulations of calcinosis.

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