Extensive calcinosis cutis in a patient with systemic lupus erythematosus: An exceptional complication

Calcinosis cutis is a disorder defined by skin and subcutaneous calcifications. It has been described in association with autoimmune diseases, particularly with dermatomyositis and systemic sclerosis. The association with systemic lupus erythematosus (SLE) is rare, only a few cases have been reported [1,2]. We report a new case of SLE associated with extensive calcinosis cutis.

A 38-year-old Moroccan woman with a history of SLE presented with subcutaneous nodules. She had been treated with corticosteroids and Mycophenolate Mofetil for 4 years for her skin (disseminated discoid systemic lupus), articular and renal (Class IV nephritis) disease. Physical examination revealed cicatricial and atrophic discoid lupus lesions, with subcutaneous nodules on the head (figure 1C), the anterior, posterior and lateral side of both thighs, upper and lower limbs (figure 1D and E). The nodules were firm, yellow-to-white. There were no signs of disease and the patient was pain-free.

Plain radiographs showed an increase in density of the soft-tissues of the lower and upper limbs (figure 1A and B) consistent with diffuse calcinosis cutis. The calcinosis did not rely on the vascular frame and there were no signs of muscular or visceral involvement.

Laboratory tests showed no signs of SLE activity. White blood cell, red blood cell counts, erythrocyte sedimentation rate, were normal. The ANA by indirect immunofluorescence 1:160, anti-ribonucleoprotein antibodies, SS-A (Ro), SS-B (La), Sm, Scl-70 and Jo-1 were negative, anti-DNA double-chained antibodies by means of Crithidia luciliae negative, anti-cardiolipins (ACL) IgM and IgG, b 2-glycoprotein I, IgM and IgG and lupoid inhibitor negative, anti-cytoplasmic antibodies by immunofluorescence negative and total complement and C3 and C4 fractions were normal. There was no abnormal level of calcium, phosphorus, parathyroid hormone, 25-hydroxyvitamin D or muscle enzyme. The urine levels of calcium, phosphorus were also normal. There were no signs of a dermatomyositis skin pattern or muscular pain. We retained the diagnosis of diffuse calcinosis cutis associated with SLE. Pathology of the skin and subcutaneous tissue showed patches of black-colored staining, indicating tissue calcification under microscopy (figure 1F). No treatment was put in place, as there were no complaints from the patient.

Calcinosis cutis is the most common type and most notably occurs in autoimmune connective disease. It is primarily occurs in patients with dermatomyositis, systemic scleroderma and mixed connective tissue disease [1,2]. But Calcinoisis cutis in systemic lupus is rare [2–5], particularly in diffuse form. As in our case, it occurs with normal levels of calcium and phosphate and develops as a result of skin or subcutaneous damage/trauma or abnormalities.

The main pathogenic mechanism of calcinosis cutis in systemic lupus is not well known. Studies have shown the presence of activated macrophages and pro-inflammatory cytokines in the calcinosis of dermatomyositis. Raised tissue expression of advanced glycation end products and their receptor has been noted in patients with systemic sclerosis and systemic lupus erythematosus with calcinosis [6,7]. There are five types of calcinosis cutis: dystrophic, metastatic, iatrogenic, idiopathic and calciphylaxis. Systemic lupus is associated with the dystrophic type, defined by the deposition of calcium in previously damaged tissue, as in other inflammatory diseases [3]. Our case presented with the dystrophic type of calcinosis cutis without involvement of renal function or hyperparathyroidism. The
examination did not reveal skin thickening or Raynaud’s phe-
omenon, excluding an overlap syndrome with systemic scle-
rosis. Biologically, there were specific SLE antibodies, such as
anti-Sm, but no scleroderma-associated antibodies. There are
approximately 41 cases reported in the literature of dystrophic
calcinosis occurring in SLE. All of these cases have shown a
female predominance, and generally affected the patient in a
mean of 9.8 years following the diagnosis of SLE. Many of
these patients were on systemic steroids or had some type of
tissue injury, such as myopathy or skin ulcerations as a cofactor
of skin calcifications. Also, calcinosis lesions occurred most
frequently on the buttocks and extremities.

Although no treatment was uniformly effective, small and
symptomatic calcified deposits or larger localized lesions
can be treated with surgical excision. Medical treatment with
diltiazem, warfarin, biphosphonates and others, which are
primarily aimed at treating the process of calcinosis with
varying success provided benefit for some patients. Also, case
studies have shown that aggressive treatment of the underling
inflammatory condition with intravenous immunoglobulin,
anti-TNF agents, thalidomide and haematopoietic stem cell
transplantation has led to improvement of the calcinosis [8,9].
To date, however, there are still no best recommendations for
treatment due to the small number of cases and the short
period of follow-up.

This rare complication should be considered in differential
diagnosis, especially in those patients with systemic lupus in whom it was not possible to prove other causes such as vasculitis, antiphospholipid syndrome or infections and this suspicion should be greater with concomitant renal failure.

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Resolution of SLE-related soft-tissue calcification following haematopoietic

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