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

Calcifications cutanées extensives chez une patiente suivie pour un lupus érythémateux disséminé : une complication exceptionnelle

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 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 calcification 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 phenomenon, excluding an overlap syndrome with systemic sclerosis. 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 underlying 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.

Acknowledgements: Financial support: none.

Disclosure of interest: the authors declare that they have no competing interest.

References

W. Ammouri, H. Harmouche, O. Ahrikat, M. Maamar, M.Z. Tazi, M. Adanaoui
University Mohamed V of medicine, Ibn Sina Hospital, Internal medicine department, rue Lamfadel Cherkoua, BP 6527, Rabat, Morocco
Correspondence: W. Ammouri. Internal medicine department, Ibn Sina Hospital, Rue Lamfadel Cherkoua, BP 6527. University Mohamed V of medicine, Rabat, Morocco
wafaammouri@hotmail.com
Received 12 November 2017
Accepted 19 February 2018
Available online: https://doi.org/10.1016/j.lpm.2018.02.005
© 2018 Elsevier Masson SAS. All rights reserved.