LETTER TO THE EDITOR

Extensive calcinosis cutis universalis in a patient with systemic lupus erythematosus: 10-year treatment experience

To the Editor,

We report the case of a 37-year-old woman with systemic lupus erythematosus (SLE), who spontaneously developed several isolated, hard, erythematous, and painful plaques with ulceration in her right lower leg 4 years after the diagnosis. The plaques then gradually developed into nodules with a hard base. She suffered from the progression of purulent and chalky, milk-like discharge from the ulcerative wounds, even with oral prednisolone treatment (the dose varied from 10 mg to 30 mg per day).

The laboratory data showed normal serum calcium, phosphorus, parathyroid hormone, and muscle enzyme levels. Several episodes of cellulites were also noted over the lesion sites with various pathogens isolated. A plain film X-ray showed numerous small soft tissue calcifications extensively scattered over her pelvic region (Fig. 1A), thigh, knee, and lower leg (Fig. 1B). However, computed tomography revealed that the calcified lesions were only present in the skin and subcutaneous tissues, with no muscle, vessel, or visceral organ invasion (Fig. 1C). Pathology of the skin and subcutaneous tissue showed patches of black-colored staining, indicating tissue calcification under microscopy (Fig. 1D). The diagnosis was extensive calcinosis cutis universalis. During this period, she underwent surgery several times for the removal of the newly isolated calcified masses over her buttocks and four extremities. She also received bisphosphate 10 mg/kg/day for 6 months, ceftriaxone 2 g/day for 30 days, diltiazem 30 mg/day for more than 1 year, and intralesional corticosteroids. However, the general condition still progressed.

Calcinosis cutis is a rare disorder characterized by calcium deposition in the skin and subcutaneous tissues. There are five major types according to different etiologies: dystrophic, metastatic, iatrogenic, idiopathic, and calciphylaxis [1,2]. According to the involved area, it can be identified as calcinosis circumscripta if limited to a small area of the extremities and joints. Calcinosis universalis diffusely influences subcutaneous and fibrous structures of the muscles and tendons.

Calcinosis cutis is common in connective tissue diseases, and especially in systemic sclerosis with limited cutaneous scleroderma (approximately 25%), and it is also found in dermatomyositis and overlap syndromes. However, it has rarely been associated with SLE [3–4]. 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. The etiology of SLE is dystrophic in nature and almost always presents as calcinosis circumscripta, with the presentation of calcinosis cutis universalis being extremely rare [5].

Because of the rarity of this entity and an unclear pathophysiological mechanism, there are no standard treatment guidelines, although various treatments have been reported to be beneficial. Reiter et al. [5] reviewed the current treatment choices and proposed some suggestions: (1) either small calcified deposits or larger localized lesions can be treated with surgical excision; (2) small calcified deposits can be successfully treated with warfarin, surgical excision, carbon dioxide laser, ceftriaxone, or intravenous immunoglobulin; (3) larger calcified lesions may benefit from curettage, surgical excision, probenecid, diltiazem, aluminum hydroxide, minocycline, and bisphosphonates; and (4) extracorporeal shock wave lithotripsy can be used for pain control.

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.

Conflicts of interest: All authors declare no conflicts of interest.

http://dx.doi.org/10.1016/j.kjms.2014.03.009
1607-551X/Copyright © 2014, Kaohsiung Medical University. Published by Elsevier Taiwan LLC. All rights reserved.
Figure 1. Plain film X-ray shows (A) numerous small soft tissue calcifications extensively scattered over the patient’s pelvic region, and (B) extensive soft tissue calcifications over the thigh, knee, and lower leg. (C) Computed tomography shows that extensive calcified lesions only exist in the skin and subcutaneous tissues, with no involvement in muscles, vessels, or visceral organs. (D) Pathology of the skin and subcutaneous tissue reveal patches of black-colored staining, indicating tissue calcification under microscopy (hematoxylin-eosin stain, 40x).

References


