CASE REPORT

Discoid lupus erythematosus with dystrophic calcinosis cutis

Ayumi Korekawa, MD, Koji Nakajima, MD, Takahide Kaneko, MD, Hajime Nakano, MD, and Daisuke Sawamura, MD

Hirosaki, Japan

Key words: autoimmune connective tissue disease; lupus erythematosus; surgical therapy; calcification; cervical region; pain.

INTRODUCTION

Calcinosis cutis (CC) is a rare disorder known to occur commonly in association with underlying autoimmune connective tissue diseases. CC primarily occurs in patients with dermatomyositis, systemic scleroderma, and mixed connective tissue disease. It is only rarely associated with systemic lupus erythematosus (SLE) and even more rarely with discoid lupus erythematosus (DLE).

CASE REPORT

An otherwise healthy 33-year-old woman was referred to our hospital for erythematous atrophic lesions around the right side of her neck. Physical examination found dusky-red, well-demarcated erythematous atrophic, partially scarred, and telangiectatic plaques extending from the right side of her chest to the neck and back, and there were firm, yellow-to-white nodules on the right side of the clavicle (Fig 1). She had no systemic symptoms, but the epidermis of the right clavicle was painful to the touch, and the erythematous atrophic lesions were itchy. She indicated that the erythematous atrophic lesions had first appeared in her 20s, and years later the firm nodules had occurred.

Histopathologic examination of an erythematous atrophic lesion found slight hyperkeratosis, atrophy of the epidermis, slight liquefaction degeneration of the basal cell layer, and thickening of the Periodic acid–Schiff–positive basal membrane. Deposits of mucin could not be detected. Dilation and slight increasing of capillaries were seen in the papillary dermis. Lymphocytic infiltration was seen in the follicles and eccrine glands in the dermis (Fig 2, A)

There were calcified deposits, which were positive for Kossa stain, in the middle and reticular dermis (Fig 2, B). There were no abnormal findings in the subcutaneous fat. On direct immunofluorescence, granular deposits of immunoglobulin M were observed at the basal membrane.

The following laboratory studies found no abnormalities: white blood cell and red blood cell counts, erythrocyte sedimentation rate, antinuclear antibody, anti–double-stranded DNA, anti–SS-A antibody, SS-B antibody, C3, and CH50. Levels of
serum calcium, phosphorus, parathyroid hormone, and 25-hydroxyvitamin D were also normal. A chest radiograph showed calcification in the right clavicular area (Fig 3).

The clinical, histologic, and immunopathologic findings confirmed a diagnosis of disseminated DLE with dystrophic CC not accompanied by SLE. Association of lupus panniculitis was ruled out because of no histologic findings in the subcutaneous fat. Also, inflammation of DLE and irritation by the calcified nodules were suggested as a possible cause of dilation and slight increasing of capillaries in the papillary dermis. Steroid ointment therapy on the DLE was initiated, and the calcified nodules were surgically removed to provide pain relief.

**DISCUSSION**

CC is a condition in which large amounts of calcium are deposited. There are 4 subtypes of CC: dystrophic, metastatic, iatrogenic, and idiopathic. Dystrophic CC is the most common type and most notably occurs in autoimmune connective disease, but CC in SLE is rare. It occurs with normal levels of calcium and phosphate and develops as a result of skin or subcutaneous damage/trauma or abnormalities.

Although SLE with dystrophic CC is rare, there are 37 reported cases. On the other hand, case reports of DLE with dystrophic CC are very rare, with only 4 reports (5 cases) to date. Lesions occurred most frequently on the buttocks and extremities. The time between the diagnosis of DLE and the onset of CC was approximately 17.8 years (range, 4 years to more than 40 years). All of the cases occurred in women. Almost all patients (4) had SLE or other systemic diseases. Only one case, reported by Ueki et al in Japan in 1980, was of a woman with CC in DLE on her neck.

Our patient had neither SLE nor systemic disease; therefore, ours is the second report to our knowledge of DLE with dystrophic CC not accompanied by SLE. Interestingly, both cases were reported from Japan, and the DLE and CC were seen on the cervical region (neck or clavicle). Although the reasons that CC developed in the DLE lesion in this case remain unclear, she had scratched the DLE lesion for many years and frequently carried her shoulder bag on her neck and clavicle (the site of the DLE lesion), despite the fact that touching the lesion was painful. We hypothesize that repeated mechanical irritation of the DLE lesion and chronic inflammation by DLE induced a local tissue injury that resulted in calcification. Additionally, the naturally thin skin of the lesion may have encouraged local tissue injury.

Because the lesion of the DLE in this case was present below the neck, disseminated DLE was diagnosed. Because SLE may develop in the future, her case requires careful monitoring.

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