

SHORT COMMUNICATION

Solitary Plaque-type Morphea with Dystrophic Calcinosis Cutis

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Plaque-type morphea is the most frequent clinical form of morphea (localized scleroderma). It is characterized by thickened scar-like oval patches of skin, most frequently seen on the trunk and proximal extremities and typically confined to the dermis. Cutaneous calcification is a common finding in systemic sclerosis, but occurs rarely in morphea.

CASE REPORT

An otherwise healthy 54-year-old Caucasian man presented to our department in winter 2004 with a 3-month history of itchy skin lesion on his right shoulder. Physical examination showed an irregular and slightly hyperpigmented 7×5 cm plaque with central whitish areas, hypotrichosis and prominent induration (Fig. 1a, b). Histopathological examination revealed thickened collagen bundles in the upper and middle dermis, mild perivascular lymphocytic infiltrate and loss of skin appendages (Fig.

1c). Diagnosis of solitary plaque-type morphea was established, and treatment with high-potency topical corticosteroids was prescribed with marked improvement of the pruritus.

The patient reported that the lesion had remained unmodified for 10 years without treatment, but in recent months it had become itchy and more infiltrated. Topical corticosteroids were prescribed without improvement; therefore oral corticosteroids in decreasing doses plus oral methotrexate, 15 mg weekly, were initiated.

After a few months, a rough surface was detected in the middle of the lesion, accompanied by feelings of tightness according to the patient. There was no history of trauma, previous injection or infiltration around the lesion. An 18-MHz ultrasound image revealed an increased thickness of dermis compared with the contralateral side (0.73 vs. 0.42 cm, respectively) and multiple hyperechoic foci with distal acoustic shadowing, which suggested the presence of calcinosis in the dermis (Fig. 1d). Skin biopsy from the affected area revealed broad sclerotic collagen bundles in the dermis and calcium deposition at the center of the specimen (Fig. 1e). Multiple soft

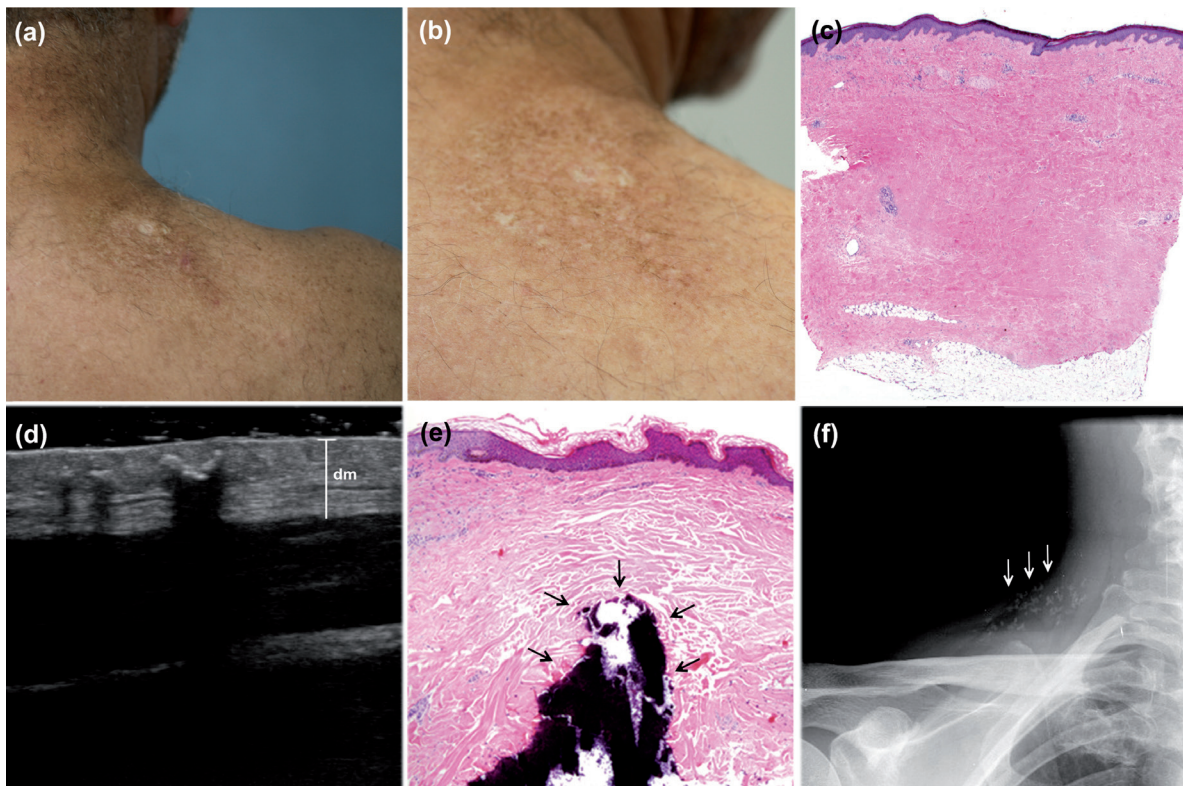


Fig. 1. (a) Indurated and slightly hyperpigmented plaque with central whitish areas located on the right shoulder. (b) Similar clinical appearance of the lesion at a different time of evolution. (c) Histopathological examination at the first visit revealed hypertrophic, sclerotic, closely packed collagen bundles in the dermis, and loss of skin appendages. No involvement of the subcutaneous tissue was detected (haematoxylin-eosin, original magnification ×20). (d) An 18-MHz ultrasound image of the lesion revealed an increased thickness of dermis (dm) and multiple hyperechoic foci with distal acoustic shadowing. (e) Biopsy specimen showing collagenous fibre accumulation and calcium deposition (arrows) in the dermis (haematoxylin-eosin, original magnification ×40). (f) X-ray showing multiple soft tissue calcifications (arrows).

tissue calcifications were also detected on the chest radiograph (Fig. 1f). At this time, blood cell counts, blood chemistry, renal function, alkaline phosphatase and erythrocyte sedimentation rate were normal. Calcitonin, parathyroid hormone and 25-OH-Vitamin D3 in serum and calcium and phosphate levels in serum and urine were also normal. Antinuclear antibody, anti-ribonucleoprotein (anti-RNP), anti-scl-70, anti-centromere, anti-SSA, anti-SSB antibodies and serology against *Borrelia burgdorferi* remain negative throughout evolution. The patient did not have sclerodactyly, Raynaud's phenomenon or any other symptoms of systemic involvement.

Clinical, histopathological and radiological findings confirmed the diagnosis of plaque-type morphea with dystrophic calcinosis. Because of the lack of major symptoms noted by the patient and the absence of a clearly defined successful treatment for this condition, we decided together with the patient to take an expectant clinical attitude. After 1 year of follow-up without treatment, the lesion has remained stable.

DISCUSSION

Calcinosis cutis is a rare syndrome characterized by deposition of insoluble calcium salts in the skin. It can be classified into 5 types: idiopathic, iatrogenic, dystrophic, metastatic and calciphylaxis (1). Dystrophic calcinosis (DC) occurs in damaged or devitalized tissues in the presence of normal calcium/phosphorus metabolism. It has been suggested that tissue structural damage and the consequent local inflammation may cause an imbalance between inhibitors and promoters of calcification, causing deposit of calcium salts in the affected area (2).

This type of skin calcification can be observed in autoimmune connective tissue diseases, such as dermatomyositis, lupus erythematosus or systemic sclerosis, and may involve a relatively localized area or be widespread (1, 2). DC in morphea has rarely been described. It has been reported in few cases of linear (3–5), generalized (6) and subcutaneous (7) forms of morphea, being exceptional in plaque-type. A large series of patients failed to detect this phenomenon: Christianson et al. (8) studied 83 patients with localized plaques of morphea and Muller et al. (9) 74 cases, and no instances of calcinosis cutis were found. As far as we know, only one case of calcinosis in plaque-type morphea has been reported in the English literature during the past 30 years: it was located on the upper back in a 69-year-old woman, and calcinosis was successfully

treated with surgical removal (10). There is no specific treatment for this condition, and various treatments have been reported with variable results: systemic or intralesional corticosteroids, colchicine, minocycline, bisphosphonates, warfarin, ceftriaxone, and surgical excision, among others (5, 7, 11).

In conclusion, DC is an infrequent finding in morphea. Reviewing the cases reported in the literature draws attention to the long evolution of the lesions of morphea with this phenomenon (up to 40 years of evolution). Because there are no satisfactory treatments, taking an expectant clinical management with periodic monitoring seems advisable if symptoms are not disabling, as in our case.

The authors declare no conflicts of interest.

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