

Calcinosis cutis: A rare feature of adult dermatomyositis

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Abstract

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestations. We describe a case of a 55-year-old woman with dermatomyositis who presented with dystrophic calcinosis resistant to medical treatment.

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestations, including heliotrope rash, Gottron papules, periungual telangiectasias, photodistributed erythema, poikiloderma, and alopecia. Although heliotrope rash and Gottron papules are specific cutaneous features, calcinosis of the skin or muscles is unusual in adults with dermatomyositis. However, it may occur in up to 40 percent of children or adolescents [1].

Calcinosis cutis is the deposition of insoluble calcium salts in the skin. Calcinosis cutis may be divided into four categories according to the pathogenesis as follows: dystrophic, metastatic, idiopathic, and iatrogenic.

In connective tissue diseases, calcinosis is mostly of the dystrophic type and it seems to be a localized process rather than an imbalance of calcium homeostasis. Calcium deposits may be intracutaneous, subcutaneous, fascial, or intramuscular.

Clinical synopsis

A 55-year-old woman was referred for evaluation because of multiple, firm nodules of the lateral hips since 1994. At that time, dermatomyositis was diagnosed based on cutaneous, muscular and pulmonary involvement. The nodules, gradually enlarging since 1999, have begun to cause incapacitation pain and many exude a yellowish material suggestive of calcium. She denied an inciting traumatic event. Combinations of oral prednisone, hydroxychloroquine, or chloroquine, have been able to control the heliotrope rash, Gottron papules, and myositis, but have not prevented progression of nodule formation.

Physical examination revealed multiple, firm, erythematous, whitish nodules, some of which exuded a chalky white material. They were located on the face, arms, and lateral hips; the largest were more than 6 cm in diameter (Fig. 1).

The laboratory studies showed normal serum calcium, inorganic phosphate, and alkaline phosphatase. The calcium-phosphate product was 42mg/dl (N <70mg/dl). Parathyroid hormone, vitamin D, and renal function were within normal range. Histological examination of one of the nodules showed epidermal hyperkeratosis, acanthosis, and massive dystrophic calcium deposits in the subcutaneous tissue (Figs. 2a, 2b).



Figure 1

Multiple firm erythematous-whitish nodules

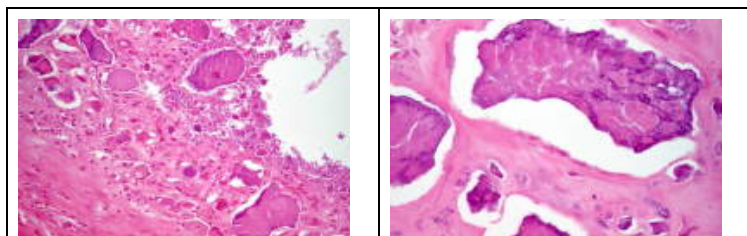


Figure 2A

Figure 2B

Epidermal hyperkeratotic and acanthosis and massive dystrophic calcium deposits in the subcutaneous tissue (HE, 20x; 40x)

Despite a trial of more than a year with oral diltiazem (120mg/day), aluminum antacids, diphosphonates (alendronate 10mg/day), and an elevation of prednisone dose (from 10mg/day to 40mg/day), her condition continued to deteriorate. The nodules became ulcerated and were complicated by repeated bacterial infections; some nodules required large surgical excisions (Fig. 3). The malar nodule was successfully excised without recurrence as were most of the other large nodules. They did not recur and the patient is still being treated with diltiazem and

diphosphonates.



Figure 3

**After large surgical excision
of the nodules**

Discussion

Calcinosis is an uncommon disorder characterized by hydroxyapatite crystals and amorphous calcium phosphates deposited in soft tissues, including the skin (calcinosis cutis) [2, 3].

Depending on the pathophysiologic mechanisms, calcinosis cutis has been classified as metastatic, dystrophic, idiopathic, or iatrogenic. Calcinosis in collagen vascular diseases is mostly of the dystrophic type, occurring in the setting of normal calcium and phosphate metabolism. Several theories have been suggested to explain dystrophic calcinosis, but the etiology remains unknown [4]. It is generally presumed to be associated with damaged, inflamed, or necrotic skin [5, 6]. One of the theories suggests that tissue necrosis caused by inflammation or injury may result in release of alkaline phosphatase by damaged lysosomes [7, 8]. Alkaline phosphatase acts on organic phosphate (which usually inhibits crystal formation), thus allowing calcium precipitation [4].

In dermatomyositis, calcification occurs three times more commonly in juvenile dermatomyositis than in the adult-onset form and may be observed in 40-70 percent of patients [6, 9]. The known risk factors for calcinosis in children include delayed treatment and severe disease [10].

In adults, calcification often presents as firm dermal or subcutaneous papules or nodules that are frequently most prominent around sites of repeated microtrauma, such as elbows, knees, buttocks, and hands. Large subcutaneous tumoral deposits can also occur on the trunk. Calcification of the muscles is generally asymptomatic and is only observed by means of radiological assessment [1]. Complications of calcinosis cutis include pain, cosmetic disfigurement, persistent ulceration with infection, and mechanical compromise [6].

Treatment of dystrophic calcification in dermatomyositis is challenging and there are no controlled studies of treatment of established calcinosis [6, 11]. Calcinosis in children can be prevented by aggressive early treatment [1, 12, 13] but is very difficult to treat when established [1]. Aluminum hydroxide antacids and diltiazem are currently used with less-than-ideal results [6]. Other agents can be tried such as probenecid, colchicine and warfarin [6].

Because patients with dermatomyositis show a high calcium turnover, and the ectopic calcium may, in part, originate from bone stores, bisphosphonates, such as alendronate, might be of value [10]. Alendronate may induce the inhibition of bone resorption, followed by reduction of calcium turnover, which may be a source of further calcium deposition [10].

The surgical removal of symptomatic cutaneous or subcutaneous calcium deposits can be considered as a last resort [14]. Because surgical trauma may stimulate calcification, it is necessary to treat a test site before proceeding further with a large excision. Calcification may recur locally on a smaller scale [6]. Smaller superficial lesions can be effectively treated with CO₂ laser [6].

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