Dystrophic Calcinosis a Rare Manifestation of Dermatomyositis

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ABSTRACT

Published on 30th December 2010

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestations. We describe a case of a 21-year- old woman with dermatomyositis who presented with dystrophic calcinosis as a late manifestation of the disease.

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

Keywords: Dermatomyositis, Calcinosis

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INTRODUCTION

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestations, including heliotrope rash, Gottron papules, periungual telangiectasias, photo distributed 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 maybe intracutaneous, subcutaneous, fascial, or intramuscular.

CASE HISTORY

An eighteen year old female presented with multiple subcutaneous swellings with discharging sinuses over the thighs buttocks, and arms of 2 months duration. It was of gradual onset and was not associated with fever or trauma.

She was diagnosed to have dermatomyositis 3 years back as she developed rash with proximal myopathy with raised Creatine phosphokinase (CPK) levels and confirmed with characteristic Electromyography (EMG) findings and muscle biopsy. She was on

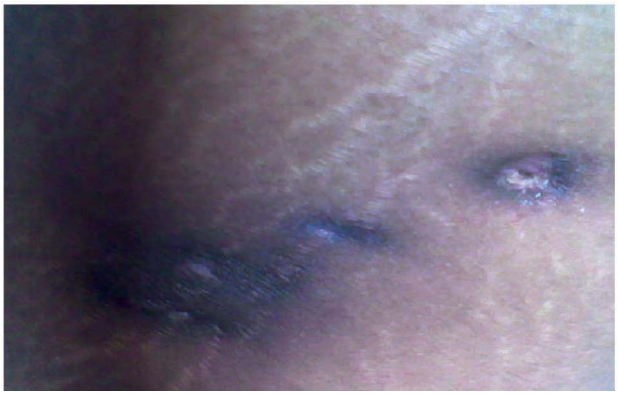

Figure1. **Discharging sinuses with chalky material**

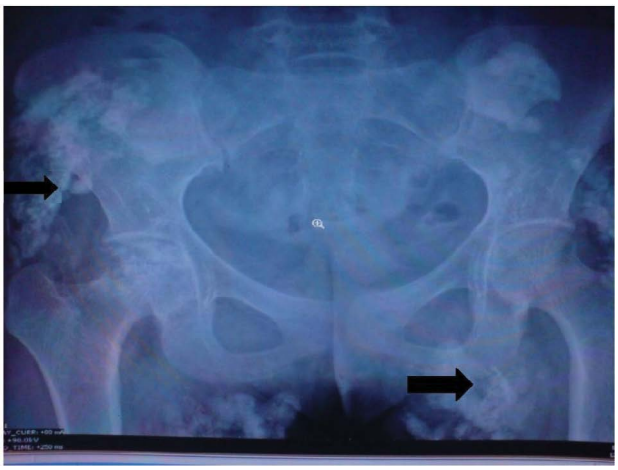

Figure 2. **X-ray pelvis showing calcified masses in subcutaneous planes**

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follow up with prednisolone, chloroquine for last six months. Her weakness and skin manifestations have improved markedly following therapy, but these swellings continued to increase. Physical examination revealed multiple hard subcutaneous swelling in thighs and buttocks. There were discharging sinuses (Figure 1) exuding chalky white material with gritty consistency suggestive of calcium crystals.

Hematology work up revealed low hemoglobin, renal and liver function tests were normal. Serum calcium and phosphorus were within normal limits, with ionic product of less than 50. Parathyroid hormone levels were normal. Radiograph of hip joint (figure 2) showed multiple sub cutaneous and intramuscular calcification.

With these clinical and laboratory features we made a diagnosis of calcinosis cutis [dystrophic calcification] as a late manifestation of dermatomyositis. Diltiazem and chloroquine was added to the patient's existing therapy which resulted in alleviation of pain and regression of the swellings.

DISCUSSION

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

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]

In adults, calcification often presents as firm dermal or subcutaneous papules or nodules that are frequently most prominent around sites of repeated micro-trauma, 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] We report a patient with late dermatomyositis who presented with distressing calcinosis cutis which improved remarkably with diltiazem and chloroquine therapy.

END NOTE

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Conflict of Interest: None declared

Cite this article as: Rojith K B, Suresh R Chandran, Nedumaran, Veera Kesari, Geetha, Ramkumar. Dystrophic Calcinosis a Rare Manifestation of Dermatomyositis. Kerala Medical Journal. 2010 Dec 30;3(4):122-124

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