Calcinosis Cutis in CREST Syndrome

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Abstract

Calcinosis cutis is a term used to describe a group of disorders in which calcium deposits form in the skin and classified as metastatic, dystrophic, idiopathic and iatrogenic.

The present case has calcinosis cutis with features of Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia (CREST syndrome) the patients with CREST syndrome often have better prognosis than diffuse systemic sclerosis. Hence pathologist should be aware that localized calcium deposition in dermis may be associated with systemic collagen vascular disease.

Introduction

Calcinosis cutis is rare disease characterized by the deposition of insoluble calcium salts in cutaneous tissue. From pathogenesis point of view, the calcific deposits can be classified as metastatic, dystrophic, idiopathic and iatrogenic.[1] Calcification seen in collagen disorders is dystrophic calcification and the calcium deposits are usually located in areas of tissue damage. Serum calcium and serum phosphorus levels are normal.[6] Calcinosis cutis with Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia is referred as CREST syndrome.[1,3,5]

Case Report

A forty year old male presented with painful discolouration of fingers on exposure to cold water since four years, swelling on right forearm and heart burn since two years. On examination, the hard, well defined nodule was seen on right forearm, with oozing of central whitish material. Multiple telangiectasia on face (Fig. 1), thickening and ulceration of finger tips were found (Fig. 2). Serum calcium and serum phosphorus levels were 10.2 mg% and 3.6 mg% respectively which were within normal limits.

Skin biopsy was performed from nodule on right forearm which showed calcium deposition in dermis (Fig. 3) and it was confirmed on Von Kossa stain. So this was an example of dystrophic calcification which was a component in CREST syndrome.

Discussion

The pathogenesis of calcinosis cutis is not entirely understood and number of metabolic, physical and other factors may be involved. This allows a variety of clinical scenario to occur. It is classified in four major types according to aetiology: dystrophic, metastatic, iatrogenic and idiopathic.[1]

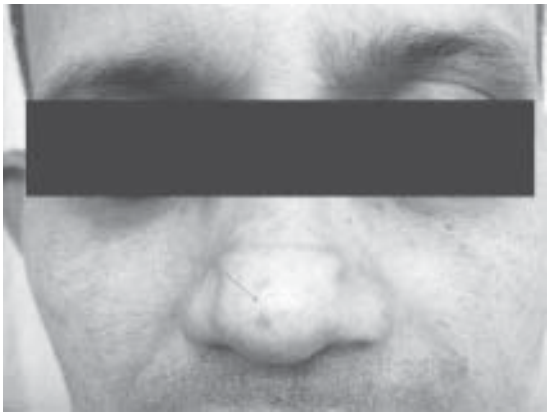

Fig. 1 : Multiple telangiectasias on face (arrow).

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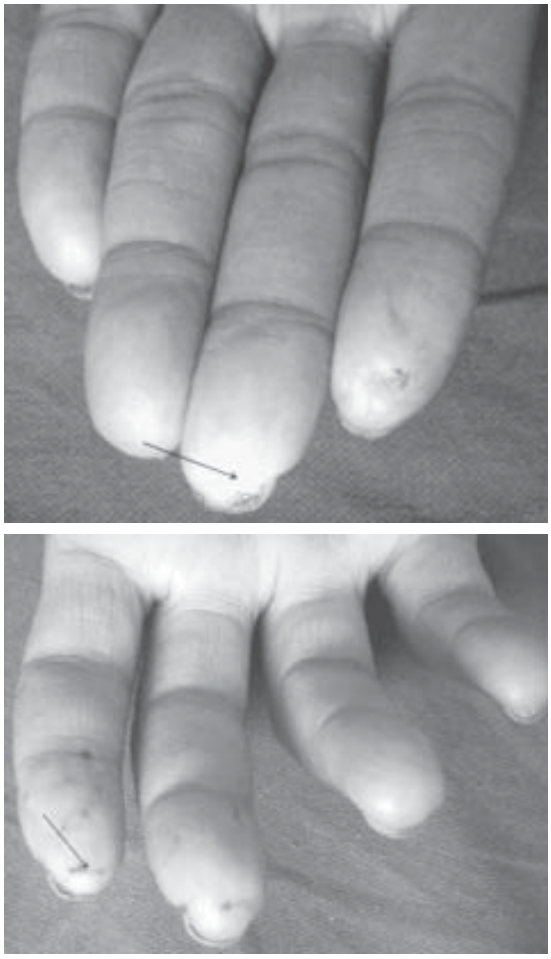

Fig. 2 : Thickening and ulceration of finger tips were found (arrow).

Patient may provide history of underlying disease such as connective tissue disorders or traumatic event in dystrophic calcification. This is most common type of calcinosis, occurs in previously damaged or diseased tissue. It occurs in localized or generalized forms without visceral involvement or serum calcium / phosphorus abnormalities.[2] Our case had localized deposits with normal serum levels of Calcium and Phosphorus.

Localized involvement occurs in many inflammatory lesions such as acne and granuloma, as well as benign or malignant

Fig. 3 : Calcium deposition in dermis (arrow) Hematoxylin & Eosin (100X).

neoplasm. Calcinosis circumscripta usually present a small deposits around fingers or elbows in patients with scleroderma or more commonly as CREST syndrome. It has been also reported in patients with systemic lupus erythematosus.[2] The deposits may ulcerate and extrude a thick, white granular calcified material as seen in our case.

Metastatic calcification occurs in a variety of condition in which the final metabolic defect results in elevated serum calcium or phosphate levels. It is generally a systemic disease with multi organ involvement, but can occasionally present as calcinosis cutis. It affects predominantly the kidneys, lungs, stomach and the media of arteries. Cutaneous lesions may present with chronic renal failure in whom poor renal clearance of phosphate results in hyperphosphataemia.[2]

Iatrogenic calcinosis has been reported after intravenous injections, if there is extravasation of calcium chloride and phosphate, as well as salt exposure from electroencephalography and electro-myographic electrode compounds. pentazosine and pitressin can also cause deep soft tissue calcifications. None of these factors were present on our case. Calcified nodules of the heels have been described after

numerous heel sticks in neonates. The mechanism likely involves both local elevation of calcium level and tissue damage.[2]

Idiopathic calcinosis cutis occurs in the absence of tissue injury or systemic metabolic effect. No causative factor is identifiable and calcification is most commonly localized to one general area. Idiopathic calcification of normal skin has been described in scrotum, penis, vulva and breast. The sub epidermal calcified nodule presents as a congenital or acquired hard, verrucuous nodule on the head or extremities of a child.[2]

When nodules or plaque are deposited, the surrounding tissue may become erythematous, infected, tender and painful, with ulceration of skin and discharge of chalky white material, which is seen in our case. Dystrophic calcifications are usually confined to areas of local connective tissue or lipid damage. Destruction and lyses of fat cells results in loss of cellular details which may combine with calcium ions to produce calcium soaps in affected area.[4] In both metastatic and dystrophic calcinosis cutis, calcium deposits stains dark blue with haematoxylin and eosin, black with Von Kossa stain. Calcium appears as fine granules in the sub cutis.

The pharmacological treatment of calcinosis cutis is difficult and a variety of drugs including bisphosphonates, intralesional corticosteroids, aluminium hydroxide, warfarin and diltiazem, have been tried with limited success. The local excision of painful or ulcerated nodule is the current existing therapeutic potion but local recurrence is common. The present case is of interest because it has features of Raynaud's phenomenon, heart burn, telangiectasia and sclerodactyly with microscopic finding of calcinosis cutis (CREST syndrome).[1] Patient with crest syndrome often have better prognosis than diffuse systemic sclerosis. Pathologist should be aware about various clinico-pathological categories associated with localized calcium deposition in dermis.

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