Calcinosis cutis – A study of six cases

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Abstract

Background: Calcinosis cutis is a very rare condition where in calcium deposits form in the skin. It occurs in four forms: metastatic, dystrophic, idiopathic and as a subepidermal nodule.

Aim: This study was done to analyze the clinical and histological features of calcinosis cutis which have an influence on patient management.

Material: A retrospective study of cases diagnosed in the Department of Pathology over a period of six years.

Results: Six cases were found during this period, which included two cases of idiopathic calcinosis cutis, two of scrotal calcinosis, one case of calcinosis cutis secondary to systemic sclerosis, and one subepidermal calcified nodule.

Conclusion: In the types in which there is an underlying systemic disease it is important to recognize this condition promptly for the proper management of the patient.

Key words: Calcinosis cutis, idiopathic, dystrophic; scrotal calcinosis; subepidermal calcified nodule.

Introduction

The deposition of insoluble calcium salts in the skin is known as calcinosis cutis. Metastatic, dystrophic, idiopathic and subepidermalnoduleare four subtypes of calcinosis cutis. Metastatic calcification results from elevated serum levels of calcium or phosphorus[1,2,3]. The latter three subtypes are associated with normal serum calcium levels. Dystrophic calcinosis cutis is the most common. Dystrophic calcinosis is calcification associated with infection, inflammatory processes, cutaneous neoplasms or connective tissue diseases and it is most frequently seen in association with underlying autoimmune connective tissue disease. Idiopathic calcinosis cutis is cutaneous calcification of unknown cause with normal serum calcium. Subepidermal calcified nodule, tumoral calcinosis and scrotal calcinosis (idiopathic calcinosis of the scrotum) are idiopathic forms of calcification[1,2,3]. Iatrogenic and traumatic calcinosis are also described, which are associated with medical procedures[2,3].

This study was done to analyse the clinical and histological features of calcinosis cutis which have an influence on the management of the patient.

Material

The cases of calcinosis cutis which were diagnosed in our institution from 2009 to 2013 were studied retrospectively. The clinical presentation, relevant investigations and morphological findings were recorded.

Results

There were two cases of idiopathic calcinosis cutis, two of scrotal calcinosis, one case of calcinosis cutis secondary to systemic sclerosis, and one subepidermal calcified nodule. The clinical and morphological findings are given in Table 1. Routine haematological, biochemical (including serum calcium and phosphous levels) and serological investigations were within normal limits in the first five cases. The sixth one was a known case of systemic sclerosis. On gross examination of the histopathological specimens the cut surfaces of the nodules had chalky white areas and felt gritty to cut (Figure 1). Histologically, with the routine haematoxylin and eosin stain, the calcified material

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had a basophilic, amorphous granular, and in some cases also a clumped appearance (Figure 2). This was confirmed by the Alizarin Red S stain which is specific for calcium.

Discussion

Table 1. The clinical and pathological findings of the patients in our study

Sl.No	Age/ gender	Clinical presentation and Diagnosis	Microscopic findings	Histolopatholgical Diagnosis
1.	24/M	Swelling on the trunk since 1 year - Calcified sebaceous cyst	Subepidermal basophilic granular clumps of calcification with formation of bone.	Calcinosis cutis
2.	65/F	Swelling since 20 years associated with pain since 3 years - Calcified bursa of the right iliac crest	Dermis and subcutaneous tissue showed a nodules of calcified material and thick fibrocollageneous tissue around.	Calcinosis cutis
3.	18/F	Lesion left arm since 2 years - Naevus	Epidermis had verrucous surface with hyperkeratosis and acanthosis. Upper dermis showed aggregates of calcium globules and masses.	Subepidermal calcified nodule
4.	38/M	Multiple small painless firm to hard nodules on the scrotal skin – 6 months	Dermis showed amorphous basophilic material.	Idiopathic Calcinosis of the Scrotum
5.	39/M	Multiple small firm to hard painless nodules on the scrotal skin – 1 year	Irregular large masses of calcified material in the dermis. Calcified material surrounded by a foreign-body reaction.	Idiopathic Calcinosis of the Scrotum
6.	35/F	Patient diagnosed as systemic sclerosis since 10 years Had features of CREST syndrome	Epidermis with hyperkeratosis and acanthosis. Dermis showed thick collageneous tissue with areas of dense calcification	Calcinosis cutis (in a case of systemic sclerosis)

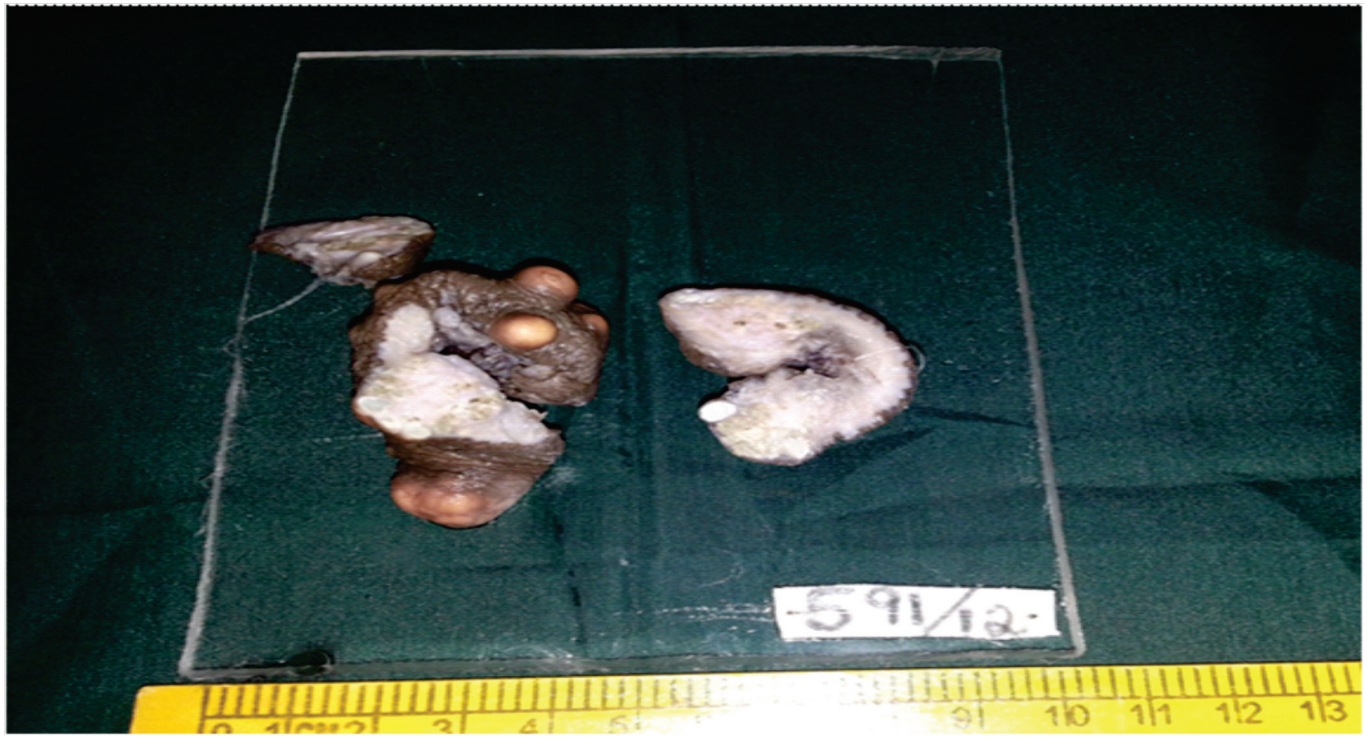

Figure 1. A case of scrotal calcinosis. Skin showing multiple nodules Cut surfaces of the nodules are chalky white and gritty

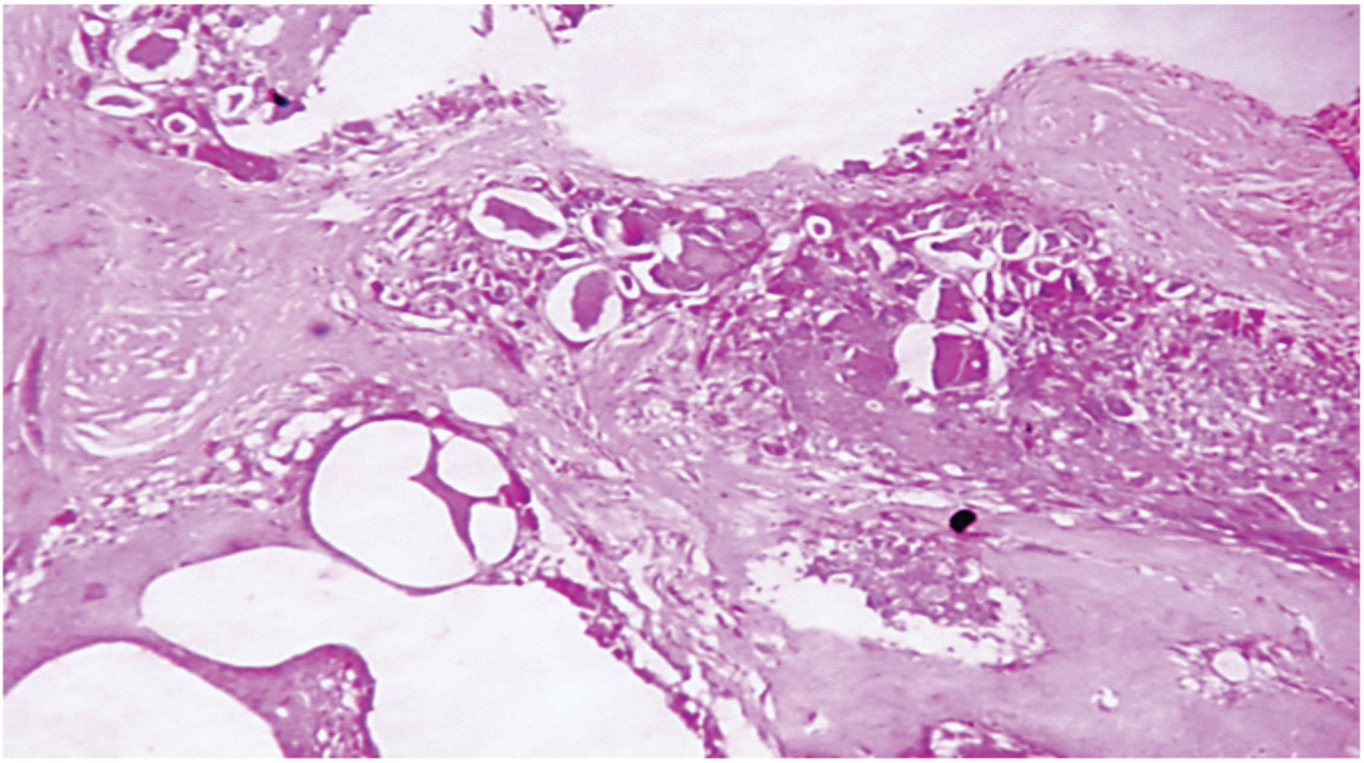

Figure 2. Dermis showing calcified material surrounded by a foreign-body reaction(H&Ex400)

Pathologic calcification is the abnormal tissue deposition of calcium salts. There are two forms of pathologic calcification. When the deposition occurs locally in dead or degenerating tissues it is known as dystrophic calcification; it occurs despite normal serum levels of calcium and in the absence of derangements in calcium metabolism. In contrast, the deposition of calcium salts in otherwise normal tissues is known as metastatic calcification, and it almost always results from hypercalcemia secondary to some disturbance in calcium metabolism[1,2,3].

Calcinosis cutis is a very rare condition where in calcium deposits form in the skin. There are four forms of calcinosis cutis: metastatic, dystrophic, idiopathic and subepidermal calcified nodule. The deposition can be in the dermis, subcutaneous tissue, or vascular endothelium when the local calcium concentration exceeds its solubility in the tissue[2].

Metastatic calcification develops as a result of hypercalcemia or hyperphosphatemia. Hypercalcemia may result from primary hyperparathyroidism, excessive intake of milk and alkali, excessive intake of vitamin D, or extensive destruction of bone through osteomyelitis or metastasis of a carcinoma. Hyperphosphatemia occurs in chronic renal failure. Metastatic calcification most commonly affects the media of the arteries and the kidneys. Instances of cutaneous metastatic calcinosis are rare[1,2,3].

Dystrophic calcification is the most common type and occurs as a result of local tissue injury. Although calcium and phosphate metabolism and their serum levels are normal, local tissue abnormalities, such as alterations in collagen, elastin, or subcutaneous fat may trigger calcification[2]. The internal organs usually remain unaffected. There may be numerous large deposits of calcium (calcinosis universalis) or only a few deposits (calcinosiscircumscripta).

Calcinosisuniversalis occurs as a rule in patients with dermatomyositis, but exceptionally it has also been observed in patients with systemic scleroderma[4].

Calcinosis circumpscrita occurs as a rule in patients with systemic scleroderma. Generally, in the presence of calcinosis, systemic scleroderma manifests itself as acrosclerosis (a combination of Raynaud's disease and scleroderma of the distal limbs, especially digits, the neck, and often the nose. This association is seen with the Thibierge-Weissenbach syndrome or the CREST (calcinosis cutis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, telangiectasia) syndrome[1,4]. One of the cases in our study was of a 35 year old woman who presented with features of CREST syndrome.

Dystrophiccalcinosiscutisalsooccurredincutaneous neoplasms such as pilar cyst, basal cell carcinoma, intradermal nevi, desmoplastic malignant melanoma, pyogenicgranuloma, hemangioma, trichoepithelioma, and seborrheic keratosis and mixed tumours (chondroidsyringomas)[2].

Idiopathic calcification occurs without any underlying tissue damage or metabolic disorder. The calcification is most commonly localized to one general area, but a case of unusually widespread calcinosis cutis has been reported[5]. Idiopathic calcinosis cutis comprises subepidermal calcified nodules, tumoralcalcinosis, and scrotal calcinosis[1,2,3]. Four of the cases in this study were idiopathic calcinosis cutis, in which no underlying cause could be found out. Subepidermal calcified nodules occur on the head and extremities, mainly as solitary, hard, white yellowish papules of 3 to 11 mm size. The disorder usually occurs in children and can even be present at birth[6,7]. Some investigators suggest that they represent calcified adnexal structures[8,9].

A special form of idiopathic calcinosis cutis is tumoral calcinosis which is characterized by large periarticular deposits of calcium resembling neoplasms and is found commonly around hip, shoulder, and elbow joints. It usually affects adolescents and young adults presenting as multiple lesions. Tumoral calcinosis as intramuscular or subcutaneous calcific masses around major joints in otherwise healthy adolescent in sporadic or familial cases had also been described[10].

Scrotal calcinosis is a rare benign local process in the absence of any systemic metabolic disorder that is characterized by multiple, painless, hard scrotal nodules. Histological examination reveals extensive deposition of calcium in the dermis, which may be surrounded by histiocytes and an inflammatory giant cell reaction, and in some cases, by a true cyst wall.

The exact pathogenesis is yet unknown and theories of origin include idiopathic calcification occurring within normal scrotal collagen, dystrophic calcification of inflamed scrotal epidermoid cysts, eccrine duct milia or dartoic muscle, and calcification secondary to minor trauma of the scrotum[11,12]. We found two cases of idiopathic calcinosis of the scrotum. The second case had evidence suggesting an origin from a pilar cyst. Despite the controversy about the origin of this entity, surgery is the treatment of choice and provides excellent results.

Conclusion

Calcinosis cutis is a rare condition and any underlying disease may be overlooked if the symptoms are mild. Hence it is important that they are specifically looked for as it is important for the proper management of the patient[11,12].

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