Idiopathic Tumoral Calcinosis Cutis- A rare clinical entity

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Idiopathic Tumoral Calcinosis Cutis- A rare clinical entity

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Abstract

The deposition of calcium in the skin, subcutaneous tissue, muscles and visceral organs is known as calcinosis. This condition commonly occurs in the skin, where it is known as calcinosis cutis or cutaneous calcification. Calcinosis cutis is a disorder caused by an abnormal deposition of calcium phosphate in the skin in various parts of the body.

We report one such case of idiopathic tumoral calcinosis cutis over forearm in a 16-year-old boy. Histopathological examination of the lesion revealed fibrocollagenous tissue with foci of calcification surrounded by foreign body giant cells, with no evidence of any underlying pathology. Idiopathic calcinosis cutis is a rare phenomenon and occurs in the absence of known tissue injury or systemic metabolic defect. Hence, it is important to delineate it from other causes of calcinosis cutis for further plan of management.

Introduction

The deposition of calcium in the skin, subcutaneous tissue, muscles and visceral organs is known as calcinosis. This condition commonly occurs in the skin, where it is known as calcinosis cutis or cutaneous calcification. Four types of this clinical condition are recognized, namely dystrophic, metastatic, idiopathic and iatrogenic. The signs and symptoms of calcinosis cutis vary depending on the underlying cause. In many cases the lesions develop gradually and are often symptomless. They usually appear as firm, whitish or yellowish papules, plaques or nodules on the surface of the skin. A solitary lesion may develop, although multiple lesions are more common. They may become tender and ulcerate, discharging chalk-like creamy material consisting mainly of calcium phosphate with a small amount of calcium carbonate.

We present a case of idiopathic calcinosis cutis over the forearm in a 16-year-old boy.

Case Report

16-year-old boy presented to the outpatients department with a painless swelling on left forearm of 2 months duration. The swelling which was initially pea sized, had progressed to about 5 × 4 × 3?cm at the time of presentation. There was no history of trauma or any such similar swellings elsewhere on the body. On examination, the swelling was located on posterior aspect of left forearm approximately 1 cm. from the elbow joint, irregular in shape and firm in consistency with no local rise of temperature. It was mobile in both horizontal and vertical axis. All movements at elbow joint were unrestricted and pain free. The skin over the swelling was normal, with no erythema, discoloration or discharge from the site. A provisional diagnosis of a lipoma was made.

Fig 1. The swelling located on the forearm
Fig 2. Close up view of the swelling

Routine hematological investigations were performed and the swelling was subjected to an ultrasonography and Fine Needle Aspiration cytology (FNAC). Sonography revealed a well-defined, lobular and densely calcified lesion in the subcutaneous plane measuring 5 × 4?cm, below the elbow along ulnar aspect of left forearm, with no involvement of the underlying muscles, blood vessels or nerves. Cytology was largely inconclusive revealing amorphous basophilic debris. An X-ray of the forearm was not performed as it was clearly a subcutaneous mobile swelling.

An excisional biopsy was then performed which revealed a single, irregular encapsulated greyish white mass measuring 5 × 4 × 3?cm.

Histopathological reporting was of fibrocollagenous tissue with foci of calcifications surrounded by foreign body giant cells (H & E stain 40 x) confirmatory of Tumoral Calcinosis Cutis.

The patient was then investigated further and serum calcium and alkaline phosphatase were found within the normal limits while Vitamin D levels were found to be insufficient (11.1ng/dl). Serum Phosphate (6.1mg/dl) and Parathyroid Hormone levels (60ng/dl) were marginally raised which could be attributed to low Vitamin D levels and were therefore not considered clinically significant. Based on this, and in the absence of factors which are associated with the other types of tumoral calcinosis being present, a final
diagnosis of Idiopathic Tumoral Calcinosis Cutis was made.

Fig. 3: Microscopic picture showing fibrocollagenous tissue with foci of calcifications surrounded by foreign body giant cells (H & E stain 40 x)

Discussion

Calcium plays a vital role in key physiologic events in many tissues, including the skin. In the epidermis, calcium plays an important role in control of major functions, including proliferation, differentiation and cell to cell adhesion. When the factors that regulate calcium in the skin are disrupted, either by local or systemic events, it results in cutaneous calcification. Calcinosis cutis is classified into 4 major types according to etiology: dystrophic, metastatic, iatrogenic, and idiopathic. (1)

Dystrophic calcinosis is calcification associated with infection, inflammatory processes, cutaneous neoplasm, or connective tissue diseases. There may be numerous large deposits of calcium (calcinosis universalis) or only a few deposits (calcinosis circumscripta). Calcinosis universalis occurs as a rule in patients with dermatomyositis whereas calcinosis circumscripta occurs as a rule in systemic scleroderma. (2)

Metastatic calcification results from and is associated with elevated serum levels of calcium or phosphorus. Hypercalcemia may result from (a) primary hyperparathyroidism (b) excessive intake of vitamin D (c) excessive intake of milk and alkali or (d) excessive destruction of bone through osteomyelitis or metastases. Hyperphosphatemia occurs in chronic renal failure as the result of a decrease in renal clearance of phosphorus and is associated with a compensatory drop in the serum calcium level.

Iatrogenic and traumatic calcinosis are those types which are associated with treatment or procedure, e.g. parenteral administration of calcium or phosphate.

Idiopathic calcinosis cutis however is cutaneous calcification of unknown cause with normal serum calcium levels. Subepidermal calcified nodule and tumoral calcinosis are idiopathic forms of calcification.

A careful evaluation of parameters of calcium metabolism combined with an assessment for associated systemic abnormalities is necessary for the correct classification of calcinosis. The various forms of calcification have already been discussed. (1, 2) “Calciphylaxis” meaning calcification of the small vessels of the dermis and subcutaneous fat has recently been added as a fifth variant of calcinosis cutis. (3) Calcinosis cutis with Raynaud’s phenomenon, oesophageal dysmotility, sclerodactyly, and telangiectasia is referred to as CREST syndrome. (3, 4) The term “idiopathic calcinosis” is used when neither local tissue injury nor systemic metabolic disorder can be demonstrated. (5) One entity is regarded as a special manifestation of idiopathic calcinosis cutis: tumoral calcinosis. It consists of numerous large, subcutaneous, calcified masses of that may be associated with papular and nodular skin lesions of calcinosis.

In the present case scenario, all the investigations to evaluate abnormal calcium metabolism i.e. serum calcium, serum phosphate, Parathormone and Vitamin D levels were within acceptable limits and there was no history of trauma, injury or constitutional symptoms. Histopathological report gave a diagnosis of tumoral calcinosis cutis hence; it was diagnosed as idiopathic tumoral calcinosis cutis.

Tumoral calcinosis may be either sporadic or familial. The familial form is associated with either hyperphosphatemia or a normophosphatemic state. The former is inherited as an autosomal recessive trait. In this present case it is sporadic due to lack of familial occurrence, solitary calcification, no history of antecedent trauma, and no evident biochemical abnormalities.

Idiopathic calcinosis cutis is a rare phenomenon and occurs in the absence of known tissue injury or systemic metabolic defect. It is important to delineate it from other causes of calcinosis cutis for further plan of management. Excision of a large calcified mass is the recommended treatment, although recurrence is not uncommon. Medical management is the recommended protocol in such cases. There has been use of intrallesional corticosteroids, probenicid and colchicine. (6) There is a report regarding the use of bisphosphonate therapy as an alternative to surgical treatment in patients with idiopathic sporadic tumoral calcinosis. (7)

Our patient is currently on follow up with no evidence of recurrence so far.

Conclusion

We recommend surgical excision of the lesions as it provides a successful resolution of the symptoms and also aids diagnosis of a rare clinical entity. Agents that modify calcium metabolism may be tried, but carefully controlled studies in support of many of these therapies is lacking.
References

Illustrations

Illustration 1

Fig 1: The swelling located on the forearm

Illustration 2

Fig 2: Close up view of the swelling
Illustration 3

Fig. 3: Microscopic picture showing fibrocollagenous tissue with foci of calcifications surrounded by foreign body giant cells