



Idiopathic Tumoral Calcinosis With Unusual Presentation Involving Small Joints: Presentation of 4 Cases With Review of Literature

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Abstract

Tumoral calcinosis is a rare familial disorder characterized by tumour like masses of calcification, usually in the soft tissues around large joints (para-articular). Masses of calcification sometimes cause gross deformity and disabilities requiring extensive surgical intervention. Involvement of distal joints like that of hand and feet is extremely rare. We present a series of 4 cases of tumoral calcinosis, two of them with unusual presentation involving great toe and thumb.

Key Words

Tumoral calcinosis, Calcinosis cutis, Calcifying lesions

Introduction

Tumoral calcinosis was first described by Duret in 1899 as endothelioma calcifie, and as lipocalcinogranulomatosis by Teutschlaeder in 1935 (1). The more widely accepted term, tumoral calcinosis was coined by Inclan in 1943 (2). Tumoral calcinosis is characterized by massive para-articular calcification in soft tissue usually involving large joints especially hip and shoulder and uncommonly knee. The usual presentation is firm, rubbery masses in soft tissues around joints. The swellings are painless and non tender (3). Tumoral calcinosis generally presents with multiple lesions and affects adolescents and young adults. Siblings are affected in half of the cases (4).

Case Summary :

Case 1: A 28 years old male presented with complaints of painful swelling of left big toe (MP joint). Swelling had been gradually increasing in size for past few weeks. There was no family history of similar complaints and the patient was otherwise healthy. On physical examination well defined mass 1.5×2 cm was present over antero-medial aspect of metatarso-phalangeal joint

of big toe, which was tender to touch. The laboratory variables were un-remarkable. X-ray (anteroposterior view) revealed multilobulated calcified mass 1.5 ×1.5 cm, around the antero-medial aspect of MP joint of big toe. However, there were no bony erosions. The patient underwent excision biopsy, which was subjected to histopathological examination.

Case 2: A 20 years old male presented with the complaints of pain & swelling right thumb (proximal interphalangeal joint) which was not responding to treatment with analgesics. Haematological & biochemical investigations were within normal limits. On radiological examination, extra-osseous calcification was seen on the radial aspect of proximal inter-phalangeal joint of thumb.

Case 3: A 15 years old male presented with painful bilateral gluteal swellings since 2 years. FNA done once had been reported as tuberculosis at community hospital & patient received anti-tubercular treatment for 9 months but without any relief. Patient was evaluated again at medical college hospital. X-ray (A/P view) revealed para-

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Fig. 1 Radiograph Showing Bilateral Para-Articular Calcified Masses in the Soft Tissue Around Hip Joint

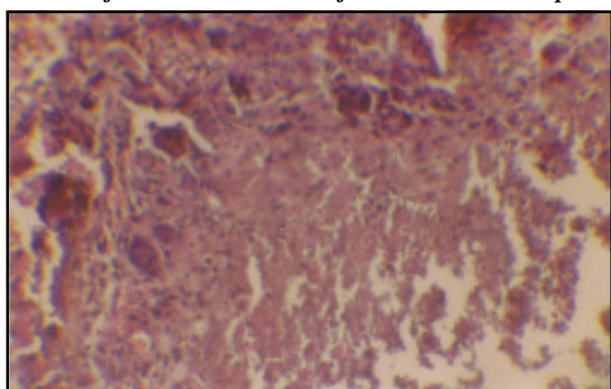


Fig. 2 Photomicrograph Showing Extensive Foreign Body Giant Cell Reaction Around the Calcified Necrotic Debris (H & E, 400 X)

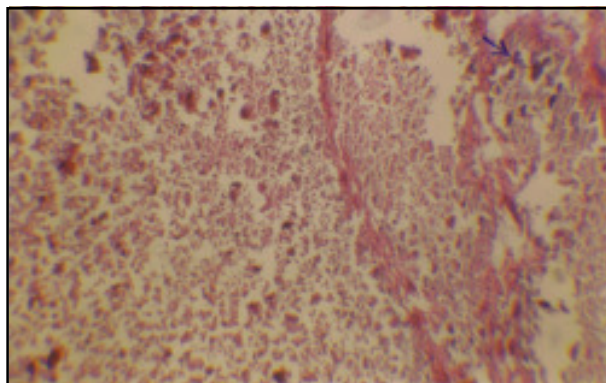


Fig. 3 Photomicrograph Showing Large Areas of Granular Calcified Debris (Arrow) [H & E, 400 X]

articular calcified masses involving both hips (*Fig 1*). Laboratory parameters were within normal limits. Excision biopsy was done & tissue subjected to histopathological examination.

Case 4: An 8 year female (sister of case 3) presented with swelling left shoulder. The symptoms were not

relieved with medical treatment. Laboratory examination including serum calcium, phosphate, and alkaline phosphatase were normal. Radiological evaluation revealed para-articular lobulated calcification. Excised tissue was subjected to histopathological examination.

Pathological Findings: Gross & microscopic examination of tissue masses received was similar in all the cases. On cut section, tissue masses were yellowish white in colour and released milky white toothpaste like material. Microscopic examination of multiple sections studied, revealed large pools of calcified material surrounded by a foreign body type of giant cell reaction admixed with chronic inflammatory cell infiltrate (*Fig 2 & 3*).

Discussion

Tumoral calcinosis is a rare disorder characterized by progressively growing tumour like masses of calcification, usually in the soft tissues around the large joints with a tendency to ulcerate the skin & encase the adjacent structures (5). The term was originally first coined by Inclan in 1943 (2). In India it was first reported by Reddy & Roa in 1964 (6). Tumoral calcinosis usually occurs in first and second decades of life with a slight male predominance (4, 5, 7). Involvement of small joints, especially of hands & feet is quite rare. In our series two cases had an unusual presentation involving small joints of thumb & great toe. Only few cases of tumoral calcinosis involving small joint have been reported till date (1, 8).

Juxtra-articular calcifications in tumoral calcinosis tend to occur predominantly on extensor surface, progressively grow over the period of months to years. Sometimes patients of tumoral calcinosis may experience successive appearance of multiple calcifications at different sites, thus clinically mimicking "tumour metastasis". However, there is no association with any actual malignancy (9). No consistent specific metabolic or biochemical abnormality has been identified in these cases (3).

The pathogenesis of tumoral calcinosis has been the subject of many theoretical postulations. Two main theories have been postulated by some authors. An aberrant tissue response to mechanical trauma or injury



leading to dystrophic calcification. The evidence favouring this theory, is the distribution of lesion in relation to pressure points. It is postulated that sleeping on the ground or wooden boards results in damage to collagen possibly by pressure / ischemia & this leads to calcification and development of tumoral calcinosis (1, 7, 10). Minor repeated trauma probably serves as a trigger mechanism that leads to chain of events, beginning with haemorrhage, fat necrosis, fibrosis and collagenization and finally collagenolysis and ultimately calcification. 1 Other theory relates tumoral calcinosis to an abnormality of calcium & phosphorus metabolism, however evidence for this theory is conflicting (10). We did not find abnormalities of calcium & phosphorus metabolism in any case of our series.

Smack *et al* (9) has formulated pathogenesis based classification, subdividing this entity into three types. (I) Primary normophosphatemic tumoral calcinosis: These cases lack familial occurrence and recurrences are rare. (II) Primary hyperphosphataemic tumoral calcinosis show strong familial patterns and increased levels of serum phosphate & calcitriol and normal serum calcium levels. (III) Secondary tumoral calcinosis: Characterized by presence of underlying disorders like secondary hyperparathyroidism, hypervitaminosis D, milk alkali syndrome, chronic renal failure. Two of our cases presented with unusual presentation involving small joints of hand & foot, could be grouped as primary normophosphataemic tumoral calcinosis. The other two cases were siblings (familial idiopathic tumoral calcinosis), however serum calcium and phosphate levels were normal in both the cases.

The lesions of the tumoral calcinosis have been histologically classified into 3 stages by Pakasa *et al.* (7) Stage 1 lesions have predominant cellular response and minimum calcification. Stage II lesions show extensive calcification in addition to inflammatory response. Stage III (inactive stage) is the end stage lesion showing cystic bland calcification without any associated cellular response. In our series histopathological features were consistent with stage II disease.

Complete surgical excision of lesion and capsule is considered the treatment of choice. In unresectable cases, partial surgical excision and concomitant oral aluminum hydroxide in combination with low calcium & low phosphate diet has been found to be effective (4, 9). Recurrences are related to incomplete removal. None of our cases recurred after surgical excision up to 2 years of follow-up.

Tumoral calcinosis generally occurs around large joints and involvement of small joints as seen in two of our cases is extremely rare. We conclude that tumoral calcinosis should be considered in the differential diagnosis of painful swellings developing in the vicinity of small joints of hand and feet.

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