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Case Series

Tumoral Calcinosis: A Case Series

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Abstract

Tumoral calcinosis is a rare condition of unknown etiology. It is a misnomer as they are not true neoplasms (as they do not have dividing cells) characterized by the deposition of inorganic calcium with serum exudates in soft tissues in periarticular location within dermis. Only few cases have been reported. Here, we are presenting a series of cases which we have encountered in our hospital with complaints of swelling without pain and no history of trauma. Initial assessment was done with the help of radiological investigations, serum calcium, and phosphate levels. Excision of the tumor was done and histopathological examination confirmed the diagnosis.

Key words: Calcium deposits, Idiopathic, Misnomer, Tumoral calcinosis

INTRODUCTION

Tumoral calcinosis is a rare condition of unknown etiology wherein there is calcium deposition in the soft tissue in periarticular location, i.e. around joints. It is a MISNOMER. The name indicates calcinosis (calcium deposition) which resembles tumor (like a new growth). Virchow initially described calcinosis cutis in 1855. They are not true neoplasms - they do not have dividing cells. They are just deposition of inorganic calcium with serum exudates. Children and adolescents (6–25 years) are the most commonly affected. They are more common around shoulders, hips, and elbows.[1] The name indicates calcinosis (calcium deposition) which resembles tumor (like a new growth).

CASE REPORT

1. A 15-year-old female patient presented to surgical outpatient department with complaints of swelling in the left gluteal region for 6 months. No history of pain and trauma associated with the swelling. On examination, a single, vertically oval, 15 cm × 10 cm, smooth, firm, non-tender, and mobile (both directions) swelling present in the left gluteal region extending into the lateral compartment of the thigh. Clinical diagnosis of soft tissue sarcoma was made; fine-needle aspiration cytology (FNAC) was inconclusive, core needle biopsy showed calcifications with fibrocollagenous stroma and giant cell reaction without signs of malignancy suggestive of tumoral calcinosis. Radiological investigations were done which supported our histological diagnosis [Figure 1].
2. A 55-year-old female patient has come with a complaint of swelling over lateral aspect of thigh right side for 8 months, gradually progressive in nature, no history of pain, and no history of trauma in the past. On examination, an irregular swelling of size 7 × 4 noted over the right anterior superior iliac spine which is non-tender, hard, and freely mobile. A provisional diagnosis of calcinosis cutis was made and the lesion was excised and sent for biopsy which on HPE showed large irregular deposits of calcium in a dense collagenous stroma identified by dense uniform basophilia in dermis [Figure 2].
3. A 60-year-old female patient with a complaint of swelling in the left and right iliac crest for 2 years, gradually progressive in nature and attained the current size. No history of trauma, pain in the past. On examination, the right side swelling is 5 cm × 3 cm over iliac crest and the left side swelling is 2.5 cm × 1 cm over the iliac crest. Both the swellings are freely mobile, non-tender, hard in consistency, and dark

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pigmentation present over the swelling. Provisional diagnosis was made as calcinosis cutis which was supported by X-ray, computed tomography (CT), and FNAC. Excision done and the tumor was sent for biopsy which confirmed our diagnosis [Figure 3].

4. A 40-year-old male patient came with a complaint of two swellings in the left gluteal region for 15 months with size of 5 cm × 3 cm and 4 cm × 2 cm. No history of pain, trauma, and sudden increase in size associated with the swelling. No history of similar swellings anywhere else on the body. Both the swellings are freely mobile in nature, non-tender, and hard in consistency. FNAC was inconclusive and core needle biopsy showed features of calcinosis cutis. Provisional diagnosis of calcinosis cutis was made which was later confirmed by HPE [Figures 4 and 5].

DISCUSSION

Calcinosis cutis is a term used to describe a group of disorders in which calcium deposits form in the skin. Virchow initially described calcinosis cutis in 1855. Calcinosis cutis is classified into four major types according to etiology: Dystrophic, metastatic, iatrogenic, and idiopathic. A few rare types have been variably classified as dystrophic or idiopathic. These include calcinosis cutis circumscripta, calcinosis cutis universalis, tumoral calcinosis, and transplant-associated calcinosis cutis.[2] The term tumoral calcinosis was originally described by INCLAN in 1943.[3]

In all cases of calcinosis cutis, insoluble compounds of calcium are deposited within the skin due to local and or systemic factors. These calcium salts consist primarily of hydroxyapatite crystals or amorphous calcium phosphate. The pathogenesis of tumoral calcinosis remains unclear and several theories have been proposed. Hyperphosphatemia has been described in some patients[4,5] while local trauma has been implicated in a few cases.[6] No metabolic abnormalities were found in our patient and she denied any history of local trauma.

Patients with dystrophic calcification may provide a history of an underlying disease, a preexisting dermal nodule (which

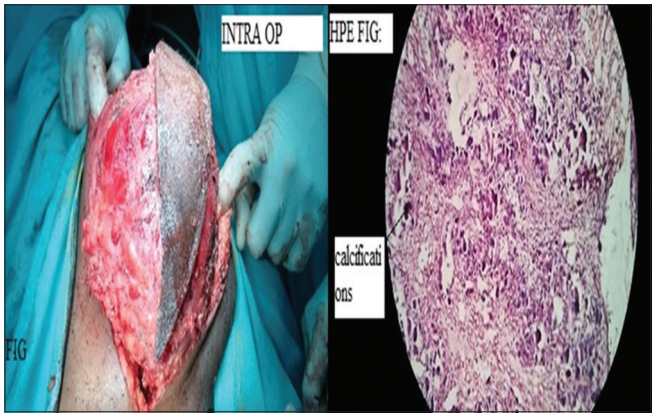

Figure 1: Intraoperative picture and histopathological slide

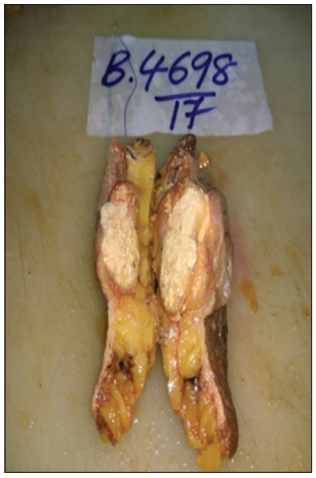

Figure 3: Cut section image

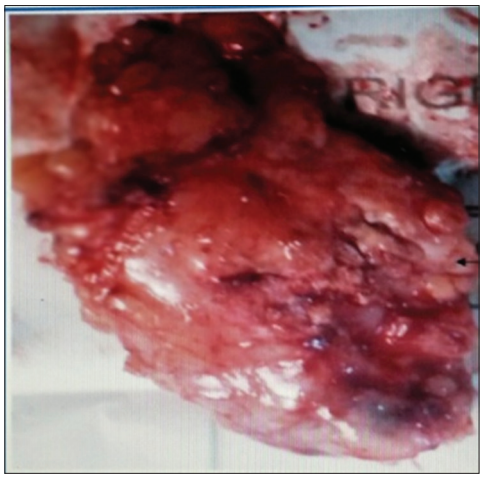

Figure 2: Specimen picture

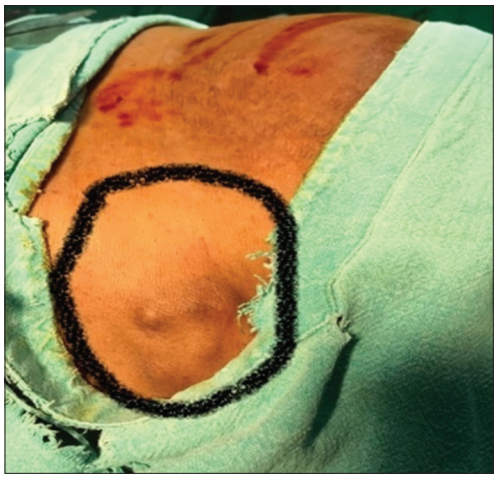

Figure 4: Pre-operative picture

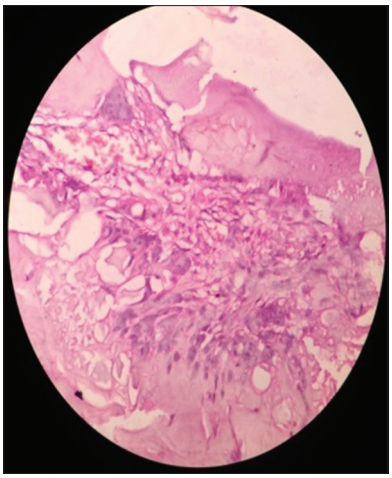

Figure 5: HPE image

represents a tumor), or an inciting traumatic event.[7,8] The patients with metastatic calcification most frequently have a history of chronic renal failure. Cases of idiopathic calcinosis cutis usually are not associated with previous trauma or disease. Those who develop iatrogenic calcinosis cutis generally have a history of recent hospitalization. The clinical presentation of calcinosis cutis can vary according to the diagnosis and underlying process. Tests of serum calcium, inorganic phosphate, alkaline phosphatase, and albumin levels may be helpful. Radiographic examination may demonstrate the extent of tissue calcification. Investigations such as CT scan and magnetic resonance imaging (MRI) are very useful in diagnosing this entity.[9,10] Bone scintigraphy with radiolabeled phosphate compounds (technetium Tc 99m methylene diphosphonate) is useful in evaluating non-visceral soft tissue calcification; this test is more sensitive than plain radiography.[11,12] CT allows for the identification of visceral and non-visceral calcification. CT is infrequently used in evaluating calcinosis cutis and primarily used in assessing tumoral calcinosis. MRI is of limited utility in evaluating calcified structures, but calcific deposits have characteristic patterns. The granulomatous foreign body reaction in tumoral calcinosis is evident.

On biopsy, granules and deposits of calcium are seen in the dermis, with or without a surrounding foreign-body giant cell reaction. Alternatively, massive calcium deposits may be located in the subcutaneous tissue. In areas of necrosis, calcium deposition is frequently found within the walls of small and medium-sized blood vessels. Calcium deposition may be confirmed on Von Kossa and alizarin red stains. Calcinosis cutis is characterized by a central mass of amorphous or granular calcified material surrounded by hyalinized fibrous tissue separating several cavities. The fibrous tissue is bordered by a granulomatous and chronic inflammatory infiltrate. There may be prominent small psammoma-like bodies or calcospherites.[13]

Medical therapy of calcinosis cutis is limited and of variable benefit. When identified, the underlying problem should be corrected.[14] Indications for surgical removal include pain, recurrent infection, ulceration, and functional impairment. A complete surgical excision along with the deposits is the mode of treatment although recurrences are common.

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