Tumoral Calcinosis in Distal Femur: First Documented Case in a Tertiary Medical Center in Philippines

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ARTICLE INFO
Article History
Submitted: 2 February, 2021
Accepted: 18 July, 2021
Published: 8 August, 2021

Source of support: None
Conflict of Interest: None

ABSTRACT

Tumoral calcinosis is a rare clinical and histopathologic syndrome characterized by calcium phosphate deposition in peri-articular soft tissue regions, most commonly in the large joints of the hips, shoulders, and elbows. We present the case of a 49-year old female with 8 months history of gradual increasing, painless swelling along her left distal thigh. Plain radiography demonstrated a dense lobulated cluster of calcific nodules within soft tissues consistent with a diagnosis of tumoral calcinosis, as confirmed by histopathological examination. Using this case as illustration, the first case of such nature in our institution, we reviewed pertinent literature related to the management of tumoral calcinosis. Although the pathogenesis in tumoral calcinosis is controversial, the ideal management is surgical excision with complete removal of abnormal tissue to prevent recurrence and to focus on the reduction of serum phosphate levels.

Key words: Distal femur; Tumoral calcinosis.

INTRODUCTION

Tumoral calcinosis is a rare condition in which there is calcium deposition in the soft tissue in peri-articular locations. The accumulations are outside the joint capsule. Calcinosis (calcium deposition) resembles tumor (like a new growth), however, it does not involve true neoplasms; it does not have dividing cells. Calcinosis is a deposition of inorganic calcium with serum exudates and is found commonly around hip, shoulder, and elbow joints.[1,2,3] Tumoral calcinosis usually presents with multiple lesions and affects adolescents and young adults. Men are affected more commonly than women. About two-thirds of the affected individuals are non-whites, mostly African American, and siblings are affected in half of the cases. Very few cases have been reported in literature. The disease is uncommon and current prevalence is not well known but a frequency range of 0.5-1.2% has been reported.[3] We report a middle-aged woman with multiple lesions of tumoral calcinosis for the rarity of this condition.

CASE REPORT

This is the case of a 49-year-old female with an 8-month history of gradual enlargement of mass in her left distal thigh with no other associated signs and symptoms and no history of trauma. Progressive enlargement of the mass was noted for over 2 months with limitation of range of motion of left knee with no pain or skin changes, prompting relatives to bring her for consult. Past medical history revealed no history of diabetes, hypertension, chronic kidney disease, asthma, cardiovascular disease, nor tuberculosis. Patient is a non-smoker and non-alcoholic beverage drinker, and has no history of any exposure to chemicals or radiation.

Figure 1: Xray of thigh AP-Lat and leg AP-Lat, showing partially calcified mass with numerous lobulations without any definite evidence of bone destruction

On physical examination, the patient was afebrile and not in cardiopulmonary distress. The rest of her systems were unremarkable and no adenopathies were noted. Examination of the left lower extremity revealed a firm, nontender mass (10x10cm- widest circumference- 62cm) at distal thigh. Range of motion was limited at this joint; hip flexion 90 degrees, knee flexion limited to 90 degrees, extension 0 degree; with full equal peripheral pulses. Examination of left and right arms revealed both having anteromedial aspect mass (1x2cm) firm, nontender; with full equal peripheral pulses.

Laboratory tests showed normal values for complete blood count, phosphorus, urea, magnesium, creatinine, and carcino-embryonic antigen. A slight elevation of calcium [T-calcium - 3.84 mmol/L(2.12-2.52); I-Calcium - 1.92 mmol/L (1.10-1.40)] was noted. Plain radiograph of the lower extremities showed heterogeneous calcification in soft tissue more in the distal femur and fewer in the proximal femur and proximal tibia (Fig. 1) Results of bone scan showed increased tracer activity in the bilateral humerus, left iliac crest, and left lower extremity (Fig. 2). Trochar biopsy of the mass on the left thigh revealed fragments of bone and connective tissue with atypical cell, hence, warranting an open biopsy for a definitive histopathologic diagnosis (Fig. 3a).The open biopsy revealed dystrophic calcification (Fig. 3b), hence the patient was referred for multidisciplinary approach and maximized medical management to decrease tumor size to allow an easier plane of dissection during surgery. On the 12th week, medical management was started with aluminium hydroxide, low calcium, and low magnesium diet, with follow up plan to start bisphosphate/ phosphate binders.

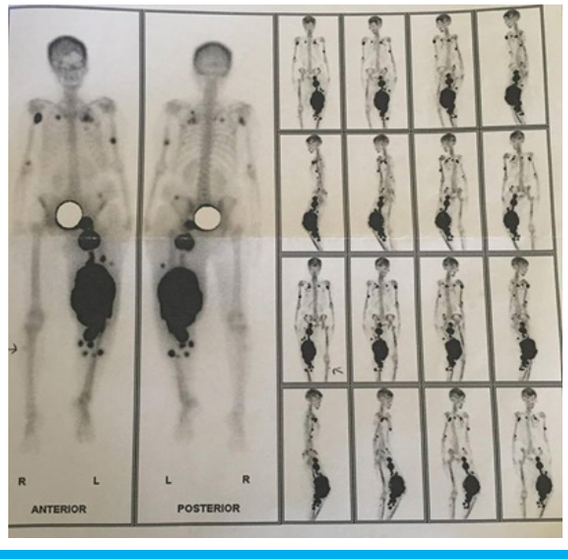

Figure 2: Bone scan showing increased tracer activity in bilateral humerus, left iliac crest, and left lower extremity.

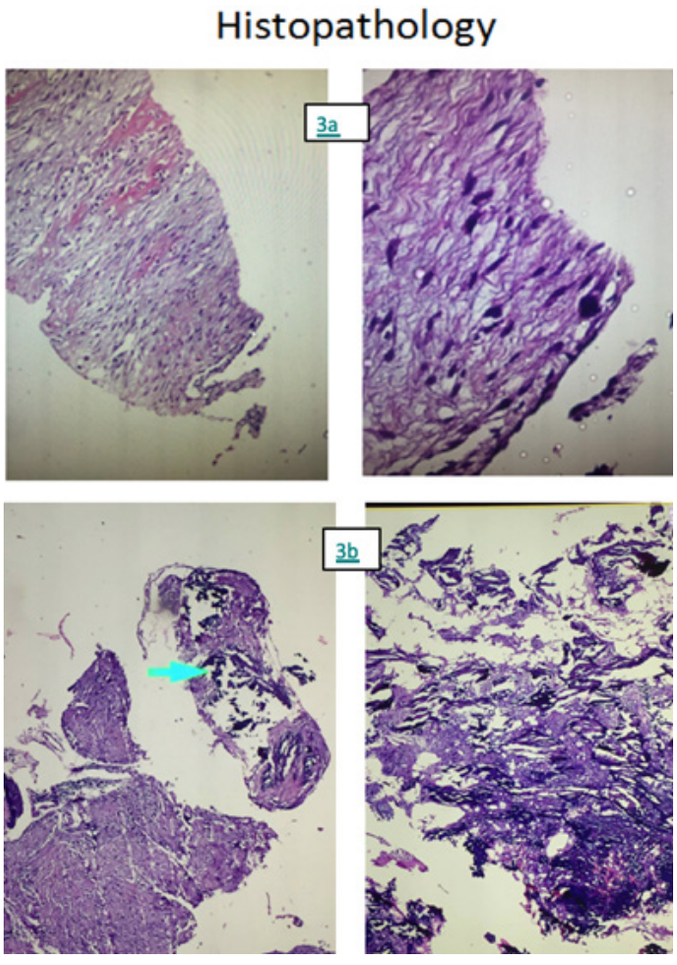

Figure 3a: Trochar Biopsy Histopathology result: fragments of bone and connective tissue with atypical cells; Fig 3b: Open Biopsy Histopathology result: dystrophic calcification

DISCUSSION

Tumoral calcinosis is characterized by the presence of lobulated soft tissue masses which are usually nontender and firmly attached to adjacent muscle and fascia. The common sites for these masses are hips, elbows, and shoulders.[1,2,3,4] Involvement of the hands and knees is uncommon.[5] There are no cases of gross bony destruction that have been reported with tumor calcinosis.

Tumoral calcinosis predominately involves healthy patients during childhood or adolescence, typically in the first or second decade of life with no sex predilection.[2,3,4] The disease is much more common in patients of African American descent.

The mechanism of calcinosis remains unknown; however, a pathogenesis-based classification of tumoral calcinosis describes three subtypes: 1) primary normophosphatemic, 2) primary hyperphosphatemic, and 3) secondary. Secondary tumoral calcinosis occurs in patients who have a concurrent disease capable of causing soft tissue

calcification.[4] Although autosomal dominant cases have been described, this condition may have an autosomal recessive pattern of inheritance as familial cases have occurred in siblings while the parents are unaffected. These familial forms are linked to mutations of various genes including GALNT3, FGF-23, and Klotho. As illustrated by the current case, the condition can also occur in patients without a positive family history. Usually, patients have hyperphosphatemia, normal serum calcium, and normal PTH. These findings, however, were absent in our patient. Tumoral lesions typically measure 5-15 cm in diameter and are composed of numerous small deposits of hydroxyapatite calcium salts. Margins are variable; some lesions have a well-defined pseudocapsule, whereas some infiltrate the surrounding tissues.

Microscopy in the active phase demonstrates a foreign body response with a rim of chronic inflammatory cells, giant cells, and macrophages surrounding the calcific deposits. A dense fibrous material is present around the central calcified material in the inactive phase. Typical X-ray findings are well-defined calcified para-articular masses that may be multiloculated or lobular. Involvement of the adjacent bone with erosions may be present but rarely.

Treatment for tumoral calcinosis focuses on the reduction of serum phosphate through the restriction of dietary intake and oral phosphate binders. Clinical and radiographic improvement has been reported after phosphorus deprivation therapy.[3] The recommended management of masses is surgical excision which requires a complete removal of the abnormal tissue to prevent recurrence.[4] Patients with multiple tumoral calcinosis affecting multiple joints have a high recurrence rate after surgical excision. In contrast with the primary idiopathic form, who have no family history of the condition or known disorders of phosphate or calcium metabolism, has normally a single event with low incidence of recurrence. Alternative treatment strategies using steroid and radiation therapy have also been proposed but do not consistently prevent lesion recurrence.[2,3,4,5]

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