Tumoral Calcinosis - A case report

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ABSTRACT

Tumoral calcinosis is a distinct clinical and histologic entity that is characterized by tumour like periarticular deposits of calcium that are found foremost in regions of hip, shoulder and elbow. The disorder occurs predominantly in otherwise healthy children, adolescents, young adults and is more often multiple than solitary. We report a case of Tumoral calcinosis in a 15 year old male and describe the histopathological features and main differential diagnosis to be considered.

KEYWORDS: Tumoral calcinosis, periarticular, multiple, elbow.

INTRODUCTION

Tumoral calcinosis is an unusual benign condition characterized by the presence of calcified soft tissue of varying size around joints. Tumoral calcinosis was originally termed endotheliomes calcifie in 1899 by Duret [1], and later as lipocalcinogranulomatosis in 1935 by Teutschlaender [2]. The term Tumoral calcinosis was proposed by Inclan et al [3] in 1945. The principal manifestation of the disease is the presence of a large, firm, subcutaneous calcified mass that is asymptomatic and slowly growing, often gradually enlarging over many years. The lesion is firmly attached to the underlying fascia, muscle, or tendon and may infiltrate these structures, but it is unrelated to bone. The underlying joints are unaffected, and with few exceptions patients with Tumoral calcinosis are in good health [4]. We report a case of Tumoral calcinosis in the elbow region (right & left) with abnormal findings such as normal calcium and phosphate levels.

CASE REPORT

A 15 year old male presented to the orthopaedic OPD with complaints of swelling and pain in right elbow since 5 years and in left elbow since 1 year. He had no history of trauma or any significant family history. On local examination of right elbow the swelling was diffuse with local rise in temperature and tenderness. Range of movements at right elbow was painful. On local examination of left elbow the swelling was diffuse, tenderness was present. Range of movements at left elbow was normal. No neurovascular deficits were present. Laboratory examination showed normal calcium, phosphorous, serum alkaline phosphatase and uric acid levels. X-ray revealed evidence of calcified mass around both elbow and appearing separate from bone.

Patient underwent an excision biopsy and sample was sent for histopathological examination. On gross examination specimen consisted of multiple grey white friable tissue largest of which measured 6x4x2 cm. Cut surface showed multiple spaces filled with chalky white material which was easily washed out. On microscopy multiple cysts surrounded by fibro collagenous tissue was seen. The cysts were filled with calcified material (basophilic) (Figure 1). The calcified material was surrounded by histiocytes, foreign body giant cells, lymphocytes, fibroblasts other chronic inflammatory cells (Figure 2).
DISCUSSION

Tumoral calcinosis is an uncommon form of extra osseous calcification characterized by large rubbery or cystic masses occurring mainly in relation to large joints. Prior studies have suggested that Tumoral calcinosis most commonly occurs around hip, shoulder, and elbow while it is less frequently seen in foot and ankle [5].

It has been suggested that Tumoral calcinosis should be divided into 3 subtypes on a pathogenetic basis: (a) primary normophosphatemic Tumoral calcinosis—in patients with normal serum phosphate and serum calcium levels and no evidence of disorders previously associated with soft tissue calcification; (b) primary hyperphosphatemic Tumoral calcinosis—in patients with an elevated serum phosphate level, a normal serum calcium level, and no evidence of disorders previously associated with soft tissue calcification; and (c) secondary Tumoral calcinosis—in patients with a concurrent disease capable of causing multiple soft tissue calcification and familial occurrence[6]. The present case has normal serum phosphate and serum calcium levels. These features indicate that our case could be classified as primary normophosphatemic Tumoral calcinosis.

Tumoral calcinosis should be differentiated from secondary calcinosis, calcinosis universalis, calcinosis circumscripta, soft tissue chondroma, pseudo gout, and calcareous tendinitis based on its clinical or pathologic characteristics.

Calcinosi universalis and calcinosis circumscripta likewise are located in the skin and sub cutis and are associated with normal serum calcium and phosphorus levels. Calcinosi universalis forms multiple nodules or plaques that occur mainly in children and are associated in about half of the cases with manifestations of scleroderma or dermatomyositis. It may ultimately lead to limited mobility, contractures, and ankylosis.

Calcinosi circumscripta, on the other hand, chiefly affects middle aged women and most commonly involves the hand and wrist, including tendon sheaths. It is associated in a large percentage of cases with Raynaud’s disease or scleroderma, sclerodactyly, or polymyositis[4].

Secondary Tumoral calcinosis is characterized by familial occurrence, multiple calcifications (including kidney, lungs, heart, and stomach), no history of antecedent trauma, and the presence of underlying disorders such as chronic renal failure, secondary hyperparathyroidism, hypervitaminosis D, milk-alkali syndrome, and metastatic bone tumour. Secondary Tumoral calcinosis can be differentiated from primary Tumoral calcinosis with careful patient history and laboratory evaluation [7].

CONCLUSION

The present case highlights the importance of considering Tumoral calcinosis in the differential diagnosis of a subcutaneous lump in the vicinity of a joint. In addition, if there is pressure of calcinosis for an extended period of time, especially for younger patients, it could induce underlying bone erosion or damage. Therefore, active treatment is indicated.

REFERENCES


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