

CASE REPORT

Tumoral Calcinosis an uncommon condition

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Abstract

Background: Tumoral calcinosis is an uncommon condition. Tumoral calcinosis can be classified into 3 types: primary normophosphatemic tumoral calcinosis (the most common), primary hyperphosphatemic tumoral calcinosis, and secondary tumoral calcinosis **Case description:** A 14 year old tribal boy presented with progressive swelling over right elbow. Radiographs revealed a multilobulated, calcified, progressive mass measuring 11x4x9 cm. Excision was performed, and Histological findings confirmed the diagnosis of tumoral calcinosis **Conclusion:** Tumoral calcinosis is a rare entity. Hence, the surgeon must be aware of this possibility.

Keywords: Tumoral calcinosis, Elbow, Large Joint.

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Introduction

Tumoral calcinosis is an uncommon condition resulting in deposition of calcium in soft tissue especially around large joints. It has been found in patients in Africa but rarely reported from other countries. ^[1] Its etiology remains uncertain. Tumoral calcinosis is attributed to an increased calcium phosphate product in the serum, leading to soft-tissue calcification ^[2]; the threshold value for precipitation is approximately 5.8 mmol/l. ^[3] Tumoral calcinosis can be classified into 3 types: primary normophosphatemic tumoral calcinosis (the most common), primary hyperphosphatemic tumoral calcinosis, and secondary tumoral calcinosis. ^[4] The first type affects young patients (without any familial history) and is usually a single lesion with low chance of recurrence after excision. The second type is hereditary and usually affects young black men living in the tropics. It is a metabolic disease with decreased fractional phosphate excretion and increased 1,25-dihydroxyl-vitamin D synthesis, whereas in proximal renal tubule the

response to parathyroid hormone is normal. It affects multiple sites including teeth, vessels, diaphysis and cranium, and recurrence is common. The third type refers to systemic diseases that promote ectopic calcification such as hyperparathyroidism and sarcoidosis.

Case Report

A 14-year-old tribal boy presented with a 7-month history of progressive swelling of the right elbow after falling from a swing. The mass was hardened not attached to the arm bones; there was no skin change. The patient had no fever and no pain, numbness, or weakness of the arm. The active range of movement was full, with flexion from 0° to 160° and hyperextension of 0° to 15°. Radiographs revealed a multilobulated, calcified, progressive mass measuring 11x4x9 cm (**Fig. 1**). There was no fracture or periosteal change, and the soft-tissue thickness was normal. MRI shows in T2 image inhomogeneous high signal intensity even though there is large amount of calcification. T1 images show inhomogeneous lesion with low signal intensity. The differential diagnoses

included tumoral calcinosis and other metabolic calcinosis (such as dystrophic calcinosis, collagen vascular disease, chronic renal disease, hyperparathyroidism). The serum calcium level was 2.43 (normal range, 2.20–2.70) mmol/l, the serum phosphate level was 1.83 (normal range, 0.81–1.94) mmol/l & the serum parathyroidhormone level was 2.8 (normal range, 1.6–6.9) μ mol/l. As the mass progressively increased in size and malignancy could not be excluded, excision was performed 7 months after presentation. Under general anesthesia, the patient was placed in a lateral

position with the right shoulder abducted to 90° and the right elbow flexed. A lobulated, yellowish mass with a pseudo-capsule measuring 9x7x4 cm was excised (Fig. 2). It was not attached to surrounding muscles, and some chalky, well-defined material emerged from the surface. Histological findings confirmed the diagnosis of tumoral calcinosis. The mass was transverse by fibrous septa with fibroblastic proliferation. Foreign body giant cells and histiocytic cells were found within the septa. There was no evidence of malignancy.



Fig 1: Preoperative AnterioPosterior & Lateral X-ray of 14 year old boy showing multilobulated calcified swelling over posterolateral aspect of right elbow

Fig 2: Intraoperative photograph, a lobulated swelling with yellowish thick liquid.

Discussion

Tumoral calcinosis is a distinct but rare entity in which there is deposition of calcium in peri-articular soft tissue. Our case falls in the idiopathic category since serum calcium and serum phosphorus levels were normal. Hence, it can be grouped under subtype primary normophosphatemic tumoral calcinosis. In a retrospective study in two institutions over a period of 10 years, amongst 9 patients, the most affected site was the elbow, with the hip coming at second position in our case elbow was involved [5] which is common site of involvement.

Other conditions such as secondary calcinosis, calcinosis universalis, calcinosis circumscripta, soft tissue chondroma, pseudogout, and

calcareous tendinitis need to be differentiated from tumoral calcinosis. Plain radiographs in tumoral calcinosis are often diagnostic, showing multiple areas of well-circumscribed, nodular masses with fibrous septae, giving a “cobblestone” or “chicken-wire” appearance. Films exposed with a horizontal beam may show the “sedimentation sign” due to mineral portion pooling dependently, creating a fluid calcium level. [6]

In our case radiological it was giving cobblestone like appearance posterior to elbow. Symptomatic treatment is the natural choice, as the cause of the disease is unknown. Only one case of spontaneous regression has been noted. [7] Medical treatment with use of calcitonin, bisphosphonates, steroids, phenylbutazone, and radiation therapy have

proved to be unsuccessful. [8] Complete surgical excision of tumoral calcinosis is considered to be the optimum treatment. The recommended management for tumoral calcinosis is surgical excision. Complete excision of mass is required to prevent recurrence. Medical treatment using agents that decrease serum phosphate levels have limited use in the management of tumoral calcinosis. Hence, surgical treatment is the only curative approach.

Conclusion

Tumoral calcinosis is a rare entity. Hence, the surgeon must be aware of this possibility. It needs thorough evaluation to exclude other causes of soft tissue calcification. Once the diagnosis is established, complete surgical excision is the only option for cure and prevention of recurrence.

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