Idiopathic tumoral calcinosis: The question of the cause is still not answered

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Abstract
Introduction
Idiopathic tumoural calcinosis is a rare disorder, characterized by tumour like periarticular deposit of calcium around the joints and the exact cause of this disease is still unknown. In this report we describe a case of primary idiopathic Tumoural calcinosis in a 17 years old female.

Case report
The patient was treated with surgical resection of the mass without recurrence at 5 years follow up, and this highlights the importance of complete surgical resection to prevent recurrence which is common following incomplete resections.

Conclusion
Complete surgical resection is effective in treatment of primary idiopathic Tumoural Calcinosis.

Introduction
Tumoural calcinosis is a rare disorder, in the literature it has been reported under various names such as calcifying bursitis, calcifying collagenolysis and kikuyu², it was first reported by Duret³ in 1899 as endotheliome calcifie then described by Teustschlaender¹ as lipocalcinogranulomatosis in 1935; the term tumoural calcinosis was introduced by Inclan⁵ in 1943 and it is the most common term.

It is characterized by tumour like periarticular deposits of calcium, most commonly around large joints such as the hip, shoulder and elbow⁶. The exact pathogenesis of Tumoural calcinosis (especially the idiopathic one) is still obscure and a matter of speculation²⁷, although factors like trauma and chronic pressure have been suggested as triggering factors⁹,¹⁰,¹¹.

Tumoral calcinosis is subdivided into:
- a) primary type with no associated disease.
- b) secondary type that follows other disorders such as renal failure, hyperparathyroidism, psedoxanthoma elasticum, sarcoidosis, sclerodema, hypervitaminosis D, milk-alkali syndrome and osteolysis¹⁰.

Then primary type has been sub classified into:
1) normophosphatemic (idiopathic) (serum concentrations of calcium and phosphate usually normal).
2) hyperphosphatemic type (serum calcium is normal but the phosphate concentration is raised).

The primary Tumoural calcinosis is the most common type². The primary hyperphosphatemic type is described as familial type and it has both autosomal dominant and recessive patterns of inheritance¹²; it has been found to result from mutations in the GALNT³ and KLOTHO gene that leads to inactivation of FGF23 which encodes a potent phosphaturic protein¹³,¹⁴.

Clinically patients usually present with asymptomatic, single or multiple, unilateral or bilateral, firm swellings over the hip, shoulder, elbow, and knee joints; rarely wrist, hands, ankle, scalp, eyelid and feet are involved¹⁵. Progressive growth and enlargement of a calcified mass can be associated with pain, functional impairment and nerve compression, long standing lesion may lead to skin ulceration, sinus tracks with chalky milk like drainage and chronic infection with secondary amyloidosis²; typically there is no bone involvement, although periostal reaction or erosion due to pressure may sometimes occur. The disorder has been reported in all age groups including infants¹⁶, it is more common in black Africans¹.

Tumoural calcinosis should come in the differential diagnosis of subcutaneous hard mass near a joint, Primary Tumoural calcinosis can be differentiated from secondary Tumoural calcinosis with careful patient’s history and laboratory evaluation.

Harkess et al.¹⁷ suggest the following criteria for diagnosing of primary (idiopathic) tumoural calcinosis:
- The presence of large, painless, calcified masses in juxta-articular sites, particularly around the elbows and hips.
- The absence of abnormal values of serum calcium or phosphorus.
- No associated renal disease, metabolic disorder, or collagen disease.
- The disease manifesting itself before the age of twenty.
- Evidence of familial or racial predisposition.
- Recurrence of the lesion particularly after incomplete excision.

The differential diagnosis of tumour-like lesions around the joint include osteosarcoma, chondrosarcoma and mycetoma¹⁵, an x-ray finding called (chicken wire) which is characteristic for Tumoural calcinosis¹⁸,¹⁹,²⁰ can be used to identify it, but histological examination is required for the definitive diagnosis and histological features as described in Inclan et al.⁵ are multiple cysts filled with calcified deposits lined by histiocytes, giant cells and xanthomatosus histiocytes.

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Case report

A 17 year old female presented with a hard swelling in the gluteal region around the right hip joint of one year duration. It started as a small swelling and gradually increased in size until it became a larger one. The patient experienced a pin prick sensation on the site of swelling which increased during walking. There was no history of similar conditions among the relatives of the patient and the patient could not recall episodes of trauma or injection over the affected area, excessive milk or antacid intake or any local or systemic illness prior to the development of the lesion. The patient had a history of excision of similar swelling around her left hip joint two years back which was also diagnosed as Tumoural calcinosis.

Physical examination revealed 15cm x10cm mass lateral to the right anterior superior iliac spine with a lot of striae on the skin over it; no sinuses or visible dilated veins. On palpation, the mass was bony hard, slightly tender, not hot and it was immobile with a nodular surface. The mobility of the hip joint was unaffected. The systemic examination was normal.

Her renal profile, serum calcium, phosphate and parathyroid hormones were normal, CBC revealed no abnormality. Plain X-Ray of the right hip showed a calcified lobulated subcutaneous mass around the right hip joint (Figure 1). Excision of the mass was done and the intra-operative finding showed a mass with multiple cysts containing chalky white material (Figure 2) and grossly the mass looks like trabeculated bone (Figure 3).

The wound was closed in a single layer without drain. The patient had an uneventful postoperative course. The mass was sent for histopathology which confirmed the diagnosis of Tumoural calcinosis. There was no recurrence at 5 years follow up.

Surgical excision is the general recommendation for treatment. Other methods of treatment have been described including medication with antacid, limitation of calcium and phosphate; partial parathyroidectomy is recommended as the therapeutic option for Tumoural calcinosis that associate renal osteodystrophy.

The aim of this article is to report a case of Tumoral Calcinosis which is a rare disease with obscure cause and to emphasis the surgical resection as good method of management.

Discussion

Primary (idiopathic) Tumoural calcinosis can be differentiated from secondary Tumoural calcinosis with careful patient’s history which is usually negative for conditions that associate secondary Tumoural calcinosis and with laboratory evaluation which shows normal serum calcium and phosphate levels, and in fact our case fulfils all criteria that are required for diagnosing Tumoural calcinosis, although currently any calcium deposit like tumour around a joint is considered Tumoural calcinosis, although currently any calcium deposit like tumour around a joint is considered Tumoural calcinosis regardless of patient’s age, sex or pre-existing disease. Surgical excision is an effective method of treatment in idiopathic Tumoural calcinosis.
importance of complete removal of the mass since incomplete resection is a risk for recurrence. Medical treatment with calcitonin, bisphosphonates, steroids and phenylbutazone are ineffective most of the time and recurrence often occurs.

Conclusion
Primary idiopathic Tumoral Calcinosis is still a disease of unknown cause but complete surgical resection is an effective method of management of this disease and it prevents recurrence.

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References