Acta Orthop. Belg., 2011, **77**, 410-413

CASE REPORT

Massive recurrent tumoral calcinosis : A rare presentation

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Tumoral calcinosis is an autosomal recessive, metabolic disorder, characterised by deposition of calcium salts in the subcutaneous tissues of the body. We present a case of massive, recurrent tumoral calcinosis in a 32-year-old male patient.

Key words : tumoral calcinosis.

INTRODUCTION

Tumoral calcinosis is an autosomal recessive, metabolic disorder, characterised by deposition of calcium salts in the subcutaneous tissues of the body. We present here a case of massive, recurrent tumoral calcinosis in a 32-year-old male.

CASE REPORT

A 32-year-old man presented with a painless huge swelling of the right hip for the last ten years. He also had similar swellings in both elbows. There was a history of excision of the both hip swelling in 1985 but gradually the swelling recurred with increasing stiffness of the hip. The elbow swellings showed no progression. There was no contributory parental or sibling history. There was no history of fever, head injury, local trauma or any other systemic complaints.

On examination the patient had a 40 cm × 35 cm bony hard, non-tender swelling over the right hip posteriorly (Fig. 1). The swelling was well defined and attached to the deeper structures with a fixed flexion deformity of the right hip joint. Movements of the right hip were grossly restricted. There was no distal neurovascular deficit. The swellings over the elbows were about 5cm × 5cm with fixed flexion deformities of both the elbow joints. General physical examination and systemic examination were non-contributory.

Routine blood investigations were normal. Serum calcium, phosphorus, alkaline phosphatase and uric acid were normal. Urinary calcium and renal function tests were normal. There was no evidence of parathyroid dysfunction. Radiographs of the pelvis (Fig. 2) showed a massive calcified soft tissue mass in and around the right hip joint with calcification of the surrounding soft tissues. Soft tissue calcification was also seen in both elbows. Since the calcified mass was obstructing the right hip joint it was excised through a posterior

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No benefits or funds were received in support of this study

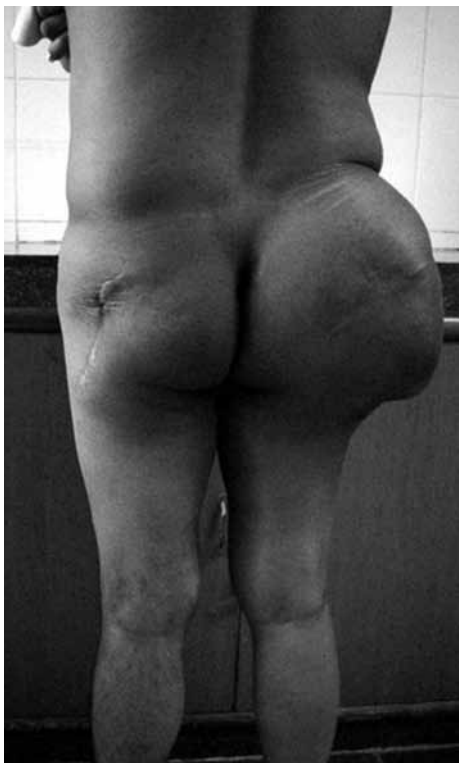

Fig. 1. — Clinical photograph of the same patient showing large swelling over right hip.

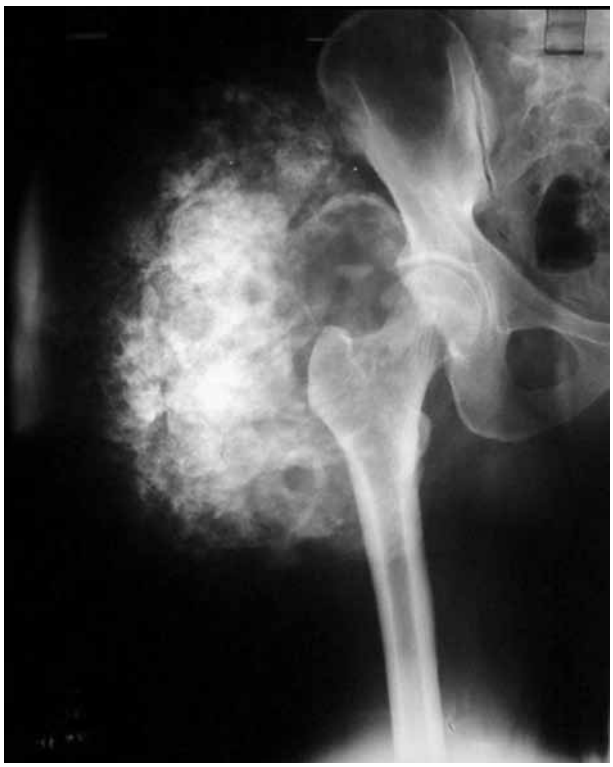

Fig. 2. — Anteroposterior radiograph of hip of same patient

approach (Fig. 3 & 4). The specimen was subjected to histopathology and on microscopy it revealed irregular round or stellate areas of granular calcium deposits in the soft tissues, surrounded by giant cells, granulation tissue and chronic inflammatory cells. No ossification was seen on any section. A clinico-pathological diagnosis of Tumoral Calcinosis was made. The postoperative period was uneventful with some restoration of useful hip joint movements.

DISCUSSION

Tumoral calcinosis is an autosomal recessive, metabolic disorder, characterised by deposition of calcium salts in the subcutaneous tissues of the body. Common sites affected are hip, shoulder, elbow and ankle joints with the hip joint being the most commonly affected (*3*). It occurs as a result of deranged calcium-phosphorus metabolism with consequent hyperphosphataemia. Cases with normal serum phosphorus concentrations have also been reported and recently a pathological classification based on serum phosphorus concentrations has been suggested. Patients can be classified as hyperphosphataemic or normophosphataemic (*5*). Pathologically, mutations in fibroblast growth factor 23 and GalNAc transferase 3 have been identified in the familiar forms of tumoral calcinosis (*6*). A foreign body giant cell reaction has been reported by some authors. The exact cause of such a reaction is not known ; no causative organisms have been isolated and calcification appears to be a primary phenomenon (*7*). Recently, the molecular pathogenesis of this group of disorders has been elucidated, leading to the identification of several proteins playing pivotal roles in the regulation of extraosseous calcification (*8*). The calcification pattern in tumoral calcinosis is very characteristic with large, juxta-articular lesions, progressive enlargement over time, and a

Fig. 3. — Exposure of calcinosis mass through posterior approach.

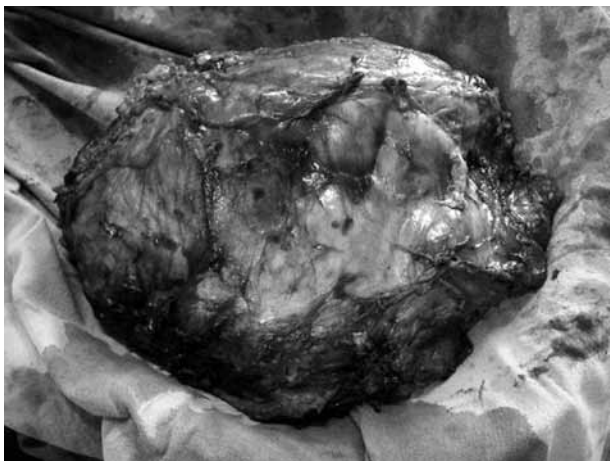

Fig. 4. — Excised specimen

tendency to recur after surgical removal (*4*). The differential diagnosis includes other conditions causing soft tissue calcification, i.e. myositis ossificans, calcified fracture haematoma, and systemic lupus erythematosus with secondary calcinosis. The condition occurs secondarily in systemic disorders like chronic renal failure, systemic lupus erythematosus, and hyperparathyroidism.

Chemical analysis of tumoral calcinosis mass shows the material to contain calcium phosphate and calcium carbonate. In one study employing the polarizing microscope, authors showed that the deposits of tumoral calcinosis are composed of calcium hydroxyapatite crystals, which are not birefringent. This is helpful in differentiating the condition from pseudogout where the calcium pyrophosphate dihydrate (CPPD) crystals show positive bifringence under polarized light (*9*). If the condition is remembered, differential diagnosis is not difficult (*6*). There are five main types of soft tissue calcification :

1) Dystrophic, where calcium is deposited in dead tissue.
2) Calcinosis, where the calcium lies in or under the skin as in calcinosis universalis or circumscripta.
3) In viscera, due to hypercalcaemia or hyperphostaemia.
4) Arterial calcification in atheroma or media degeneration.
5) Calcification within calculi.

Undoubtfully tumoral calcinosis is a separate and sixth entity. In myositis ossificans, there is actual bone infiltration into connective tissue and muscles, not calcium deposition.

The treatment includes phosphate depletion in the diet (in the hyperphosphataemic variant), topical steroid application, surgical excision of calcified deposits causing pain or limitation of joint movements, and curettage of sinuses discharging calcareous material (*2*). Parathyroidectomy has been tried in severe cases with varying success (*1*).

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