Tumoral Calcinosis

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ABSTRACT

We report a case in a 12-year-old girl with idiopathic tumoral calcinosis over the left elbow and right knee. Idiopathic tumoral calcinosis is a rare disorder affecting usually Africans and the tropical population.

CASE REPORT

A 12-year-old Chinese schoolgirl was referred to the orthopaedic clinic in May 1998 for a painful swelling over the left elbow of six months duration and a painless swelling over the right knee joint of one month duration.

She fell on her left elbow eight months ago but only noticed a small swelling about two months after the fall. It was painless and did not interfere with her elbow movement. It has gradually increased in size and has now restricted movement in her left elbow and caused her some discomfort.

There was no history of fever, loss of weight or appetite. There were no bleeding tendencies or similar swellings prior to this episode.

Examination of the left elbow showed a hard, subcutaneous swelling measuring 2 x 6 cm mainly over the posterior aspect with extensions medially and laterally. The swelling was not mobile. It was slightly warm with minimal tenderness. The range of elbow motion was from 0 to 110 degrees. Forearm rotation was normal.

There was another firm, subcutaneous swelling over the anterolateral aspect of the right knee measuring 2 x 2cm which was mobile and extraarticular. The range of motion of the right knee was normal.

Radiographs of the affected joints revealed a well demarcated mass of multi-lobulated, homogenous, calcifications in the peri-articular soft tissue (Figures 1 and 2).

The full blood picture and uric acid levels were normal. The serum calcium was normal at 2.25 mmol/l (normal range: 2.12 - 2.52 mmol/l) while the serum phosphate was slightly raised at 1.53 mmol/l (normal range: 0.29 - 1.26 mmol/l). She was referred to the paediatric endocrinologist but no other abnormalities were detected and she did not require any medical treatment.

Excision biopsy revealed the swelling over the left elbow extending deep to the triceps and occupying the posterior aspect of the elbow. The swelling was friable and consisted of chalky, yellowish material. It weighed 125 gm. The right knee lesion had ulcerated but it was not infected prior to admission.

The macroscopic histopathology examination of both tissue samples revealed a lobulated, yellow, well encapsulated mass of homogenous tissue measuring 3 x 2.5 x 5 cm from the elbow and 2 x 2.2 x 1.5 cm from the knee.

Microscopically, there were multinodular areas of calcification separated by fibromuscular septae. There was no cartilage tissue or evidence of malignancy within the tissue. The diagnosis was tumoral calcinosis.

In March 2000, almost two years after the first presentation, there was a recurrence of the swelling measuring 2.5 x 1 cm lateral to the previous surgical scar of her right knee. It was painless and did not interfere with her movement. There was no recurrence over her left elbow, which had regained full movement.

An excision biopsy of the recurrent right knee swelling (Figure 3) showed similar findings as her previous surgery. The histopathological examination confirmed the previous findings. She remains on yearly follow-up.

DISCUSSION

Tumoral calcinosis is a distinct clinicopathological entity characterised by a tumour-like deposition of calcium in soft tissues near large joints without an obvious associated metabolic calcium disorder. It is very rare and seems to affect Africans and African-Americans predominantly. It typically occurs in otherwise healthy children, adolescents and young adults with the peak incidence between the first and third decades.[1]

It is a disease of obscure aetiology with variable clinical expressivity that can demonstrate a familial trait in an autosomal recessive or autosomal dominant pattern of inheritance. The lesions are often multiple with bilateral symmetric involvement and are characteristically firm, nodular and non-tender located in the peri-articular soft tissue in the vicinity of major joints like the hip, shoulder, posterior elbow and knee.[1,2]

Sometimes the lesions cause ulceration of the overlying skin with the formation of a sinus that drains chalky material as shown in our patient. Other manifestations include anaemia, low-grade fever, splenomegaly and regional lymphadenopathy.[3]

Serum calcium levels, alkaline phosphatase and renal functions are not raised.[1] Hyperphosphataemia is present but not in all patients. Calcitriol levels may be elevated while serum parathormone levels may be normal or low. However, affected individuals are in positive calcium and phosphate balance.[3]

Plain radiographs reveal a well demarcated mass of

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multi-lobulated, homogenous, round or oval calcifications in the peri-articular soft tissue.[1,2]

Occasionally fluid levels, which are easily demonstrated by computer tomography and magnetic resonance imaging, may be seen in the larger long standing lesions and are the hallmarks of tumoral calcinosis on radiology.[1,3]

Interestingly, though dental abnormalities can be present, vascular or visceral calcifications do not occur.[3]

Macroscopically, the lesion consists of a well encapsulated firm mass of multiple calcified nodules that focally coalesce into larger, lobulated masses that are frequently filled with yellow-grey, pasty or milky fluid. Microscopically, there are large areas of amorphous or granular calcium masses with occasional interspersed, concentrically laminated calcospherites, some of which consist of hydroxyapatite.[1]

The diagnosis of idiopathic tumoral calcinosis is one of exclusion. Similar lesions are found in patients with underlying calcium disorders like secondary hyperparathyroidism or chronic renal failure. These patients are generally older and lesions are also present in visceral organs. Primary hyperparathyroidism, milk-alkali syndrome, hypervitaminosis D and scleroderma can also produce similar lesions. However they are associated with abnormal serum calcium levels.[1]

For treatment of lesions associated with hyperphosphataemia, a strict low phosphorus and low calcium diet with phosphorus binding antacids and dihydroxyaluminium sodium carbonate have proved successful. Treatment of choice is surgical removal of the lesion.[1,3]

Though recurrences are rare, they can occur in incompletely excised lesions as in our patient.[1,3] It is recommended that lesions be removed as early as possible when they are small and amenable to complete excision.[1,2]

Long standing lesions may develop secondary infection with abscess formation.

Other modes of treatment have included radiotherapy, corticosteroids and ACTH 2. Preliminary studies have shown the efficacy of calcitonin therapy since this hormone increases renal phosphate excretion.[3]

Figure 1. X-ray of the left elbow shows a homogenous densely calcified mass

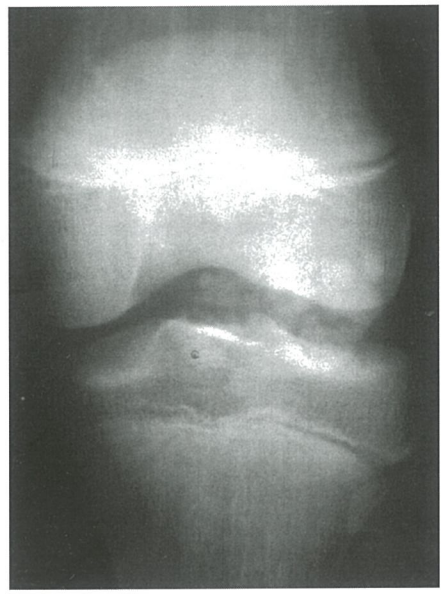

Figure 2. X-ray of the right knee shows the nodular calcified mass inferior to the patella.

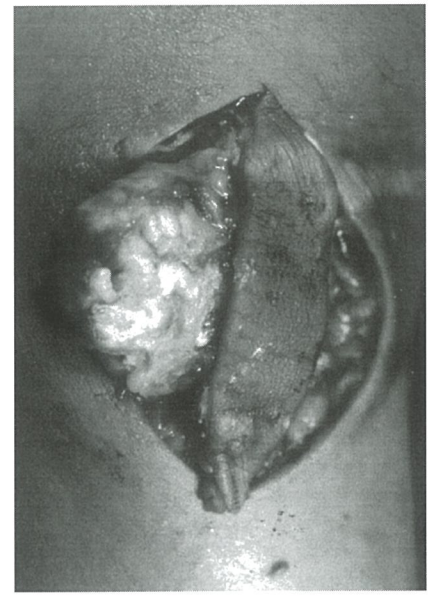

Figure 3. The intra-operative photograph shows the calcified nodular mass from the antero-lateral aspect of the right knee.

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