Tumoral calcinosis: a case report

Yuen Yee Leung, Raymond Lai
Department of Orthopaedics and Traumatology, United Christian Hospital, Hong Kong

ABSTRACT

An 11-year-old girl presented with a progressive swelling of the right elbow after a contusion. Radiography and magnetic resonance imaging showed a densely calcified lesion with no periosteal reaction or fracture. The underlying bone and muscles had normal signal intensity. A bone scan revealed increased uptake over the right elbow and the left buttock and acetabulum. Blood tests revealed no abnormality. As the mass progressively increased in size and malignancy could not be excluded, excision was performed 4 months after presentation. A lobulated, yellowish mass with a pseudo-capsule measuring 9x7x4 cm was excised. 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 transversed by fibrous septa with fibroblastic proliferation. Foreign body giant cells and histiocytic cells were found within the septa. There was no evidence of malignancy. Four months later, excision of the left elbow and buttock lesions was performed, and histology of both revealed the same diagnosis. At the 4-year follow-up, there was no recurrence.

Key words: arm; calcinosis; elbow

CASE REPORT

In July 2008, an 11-year-old girl presented with a 4-month history of progressive swelling of the right elbow after falling from a swing. The mass was hard and 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. Magnetic resonance imaging revealed the mass (measuring 6.2x2.4x5.9 cm) was hypointense in T1- and T2-weighted images, with a hyperintense rim in T2-weighted images and T1-weighted contrast-enhanced images, probably due to inflammatory changes (Fig. 2). The signal intensity of the humerus and muscles was normal; there was
no bone erosion, cortical breakage, periosteal reaction or joint effusion. Similar smaller masses were also noted over the left elbow and the left buttock (Fig. 3).

The differential diagnoses included tumoral calcinosis and other metabolic calcinosis (such as dystrophic calcinosis, collagen vascular disease, chronic renal disease, hyperparathyroidism).

A bone scan showed increased uptake over the right elbow and the left buttock and acetabulum, suggestive of myositis ossifican or heterotrophic ossification.

Blood tests revealed no abnormality; 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 parathyroid hormone level was 2.8 (normal range, 1.6–6.9) µmol/l.

As the mass progressively increased in size and malignancy could not be excluded, excision was performed 4 months after presentation. Under general anaesthesia, 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. 4). 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 transversed by fibrous septa with fibroblastic proliferation. Foreign body giant cells and histiocytic cells were found within the septa. There was no evidence of malignancy.

Four months later, excision of the left elbow and buttock lesions was performed. Histology of both also revealed the same diagnosis. At the 4-year follow-up, there was no recurrence.

**DISCUSSION**

Tumoral calcinosis (also known as Teutschlaender disease, calcifying bursitis, lipocalcino-granulomatosis, calcifying collagenolysis, and Kikuyu bursa) is a deposition of calcium phosphate and calcium hydroxyapatite within periarticular soft tissues.\(^1\) It is painless and asymptomatic and usually presents as a calcified soft-tissue mass around a large joint (usually extensor aspect) that affects the bursa and may cause pressure symptoms (e.g. nerve root compression). It commonly involves the hip, buttock, shoulder, and elbow; less commonly the knees, hands, and feet; and rarely the spine, larynx, or scalp.

Other differential diagnoses of soft-tissue calcification include mixed connective tissue diseases, dermatomyositis, calcinosis circumscripta, calcific tendinitis or calcific bursitis, and heterotrophic calcification. Neoplastic diseases such as parosteal osteosarcoma, chondrosarcoma, and synovial sarcoma should also be considered.

**Radiological features**

Tumoral calcinosis entails multilobulated cystic calcification with fluid levels, namely sedimentation signs on radiographs (Fig. 5). It may show amorphous, multilobulated calcification termed a ‘chicken-wire’ pattern of lucencies.\(^2\) Fluoroscopy may show a ‘cobblestone’ appearance (Fig. 5). Computer tomography can better delineate the calcified mass,
the cystic appearance and the sedimentation sign (Fig. 5). Homogenous lesions are suggestive of reduced metabolic activity and lower likelihood of future growth. In T1-weighted images, tumoral calcinosis shows non-homogenous, low signal intensity. In T2-weighted images, it may appear as a diffuse, low signal intensity, or a bright nodular pattern with alternating areas of high signal intensity and signal voids. Bone scans may reveal very intense tracer uptake in extraosseous lesions.

Tumoral calcinosis can be associated with diaphyseal bone marrow lesions (diaphysisis).\(^3\) Fluoroscopy may show circumferential endosteal and periosteal bone proliferation with patchy sclerosis of the medullary canal, and loss of sharpness on the inner cortical margin (Fig. 6). Bone marrow lesions (usually in children) are a definite component of tumoral calcinosis and usually affect long bones (occasionally the skull) and resolve spontaneously.\(^1\) They show patchy sclerosis of the medullary canal. Other associated lesions include calcific deposits of dental pulp with obliteration of the cavity (pulp stones) with bulbous and short roots (Fig. 7).\(^4\) Calcification over the skin, cornea, vessels and dura has also been reported.

**Histopathology**

Microscopic, crystallographic, ultrastructural and chemical properties of tumoral calcinosis have been reported.\(^5\) They are well-defined masses, circumscribed but not encapsulated, with an amorphous lobulated appearance. The contents are chalky-white fluid with yellow or white hard solid, separated by fibrous septa. Microscopically, the lobules are separated by fibrous septa, with a lining of mixed fibrous and inflammatory tissue with surrounding soft-tissue fibrosis and inflammatory changes (Fig. 8).

In contrast to osteosarcoma, chondrosarcoma, and synovial sacroma, tumoral calcinosis manifests as fibrous septa, lobular configuration, and calcium-fluid levels.\(^2\) Scleroderma has similar features to tumoral calcinosis but has no calcium-fluid levels, less fibroblasts, and thicker collagenous septa (about 40 nm).

**Pathophysiology**

Tumoral calcinosis is attributed to an increased calcium phosphate product in the serum, leading to soft-tissue calcification;\(^6\) the threshold value for precipitation is approximately 5.8 mmol/l.\(^7\) However, some authors consider it a hereditary metabolic dysfunction of phosphate regulation (with normal serum calcium levels) and distinguish it from calcification associated with renal osteodystrophy.\(^8\)

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**Figure 7** Calcific deposits of dental pulp with obliteration of the cavity (pulp stones) with bulbous and short roots (arrows).

**Figure 8** The lobules are separated by fibrous septa, with a lining of mixed fibrous and inflammatory tissue with surrounding soft-tissue fibrosis and inflammatory changes.
Tumoral calcinosis can be classified into 3 types: primary normophosphatemic tumoral calcinosis (the most common), primary hyperphosphatemic tumoral calcinosis, and secondary tumoral calcinosis. 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-dihydroxyvitamin 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.

Treatment
Treatments for tumoral calcinosis include excision, phosphate deprivation, and a combination of both. Complete excision is effective for early primary normophosphatemic tumoral calcinosis, and has a low recurrence rate. However, the recurrence rates are high for the other 2 types. In addition, recurrent masses are more likely to be progressive, particularly for incompletely excised lesions. Phosphate deprivation entails a low calcium and phosphate diet plus use of phosphate binding antacid e.g. aluminium/magnesium hydroxide, and complete resolution of the lesion has been reported. There is no evidence to infer that such treatment causes demineralisation of normal bone. The most effective treatment is a combination of surgical excision, phosphate deprivation, and use of acetazolamide.

REFERENCES