CASE REPORT

Tumoral calcinosis with unusual presentation A case report

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A case of multiple para-articular calcific masses involving the elbow, hand, back, knee and the feet was encountered. Pathology after surgical excision of all the masses revealed tumoral calcinosis. The unusual feature was the huge size of the lumbar and knee masses, and the distal location of the swellings in the hand and the foot ; there was no disturbance of calcium and phosphorus metabolism, which was an unusual feature of the condition in this patient.

INTRODUCTION

Tumoral calcinosis is an entity characterised by large periarticular deposition of calcium phosphate that resembles a neoplasm. It is usually multiple and occurs predominantly in adolescents and young adults. Approximately two-thirds of the cases involve blacks and about half affect siblings (1). The nomenclature in the literature is rather confusing, with different names being given to this condition like lipocalcinogranulomatosis, calcifying bursitis, calcifying collagenolysis, kikuyu bursa etc (11).

The lesion is usually asymptomatic and only rarely causes discomfort, pain or tenderness. The underlying joints are unaffected and as a rule the patients are in good health. For these reasons, the presentation of large-sized lesions is not uncommon. On examination, the mass is firmly attached to the underlying fascia, muscle or tendon. Complications are rare, but there may be ulceration of the overlying skin with secondary infection, fistula formation and discharge of yellow-white chalky fluid.

Most commonly the disease is diagnosed when the size of the swellings has not assumed large proportions ; the present case however presented to us with mechanical symptoms related to the size of the lesions, which interfered with normal life. There were additional unusual findings, such as normal calcium and phosphate levels, as well as presentation in the foot, which prompted us to present this case.

CASE REPORT

A 30-year-old male patient presented to the outpatient department of Rajindra Hospital, Patiala with swellings affecting the lumbar area, the right knee, the right hand, the right elbow and the right foot. The swelling had been present over a ten-year period (fig 1). The large swellings prevented him from wearing pajamas and shoes and the hand swelling had previously obstructed proper

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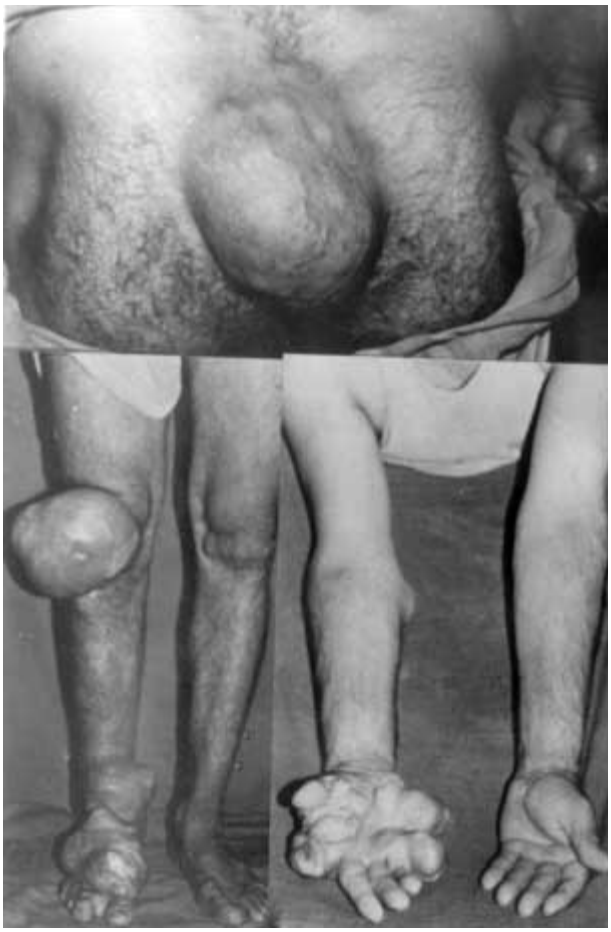

Fig. 1. — Photograph showing the large sacral lesion top), right sided large knee, ankle and foot lesion (bottom left) and the right elbow and hand lesions (bottom right).

gripping ; this led him to opt for amputation of the right little finger.

On examination all the lesions were noted to be on the right side, except for two nodular lesions on the left foot. The swellings were irregular, firm to hard in consistency and mobile over the underlying bone and joints. In the lumbar area, a large swelling 10×20 cm in dimension prevented him from lying supine ; there was a swelling in relation to the right knee (fig 1) which was discharging yellow fluid.

There was no family history of similar complaints, and the patient was otherwise healthy. Laboratory examination on multiple occasions showed normal calcium (8.7 mg/dl), phosphorus (4.3 mg/dl), serum alkaline phosphatase and uric

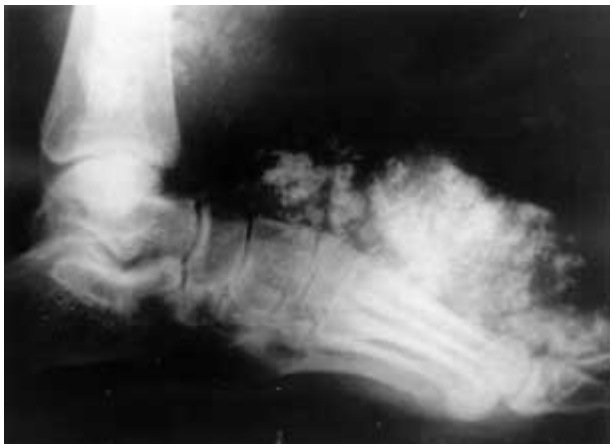

Fig. 2. — Right foot shows amorphous calcification and the involvement of the lower end of tibia.

acid levels. The serum 25-hydroxyvitamin D_3 level was 11.3 ng/ml, within normal limits . Radiographs of the hand showed multiple, round opacities separated by radiolucent lines, which had a predominant soft tissue element ; in the foot these masses were heavily calcified, and at both sites the underlying bones were normal (fig 2). Radiographs of the pelvis showed a large mass posterior to the sacrum, with radiological evidence of calcification. The right ulna was grossly increased in size and there was calcification in the marrow (calcific myelitis) (11) (fig 3). Lumbar spine radiographs were taken to see for disc involvement or chondrocalcinosis (11). There was no skin calcification (11).

CT-examination was done to assess the extent and anatomical position of the para-articular masses ; this showed irregular para-articular calcification, but the characteristic cystic spaces were not found. Fluid levels were not detected and clear-cut layering or sedimentation could not be demonstrated as has been reported (fig 4) (8). The lesions were solid with amorphous calcification. Some black areas were seen.

Per-operatively, the masses were multiple, rigid and rubbery ; they were hard when calcified and extended into adjacent muscles and tendons. Many vessels were present around the swelling and brisk haemorrhage was encountered when the swelling was not calcified ; where calcified, minimal bleeding was encountered. The masses were excised in

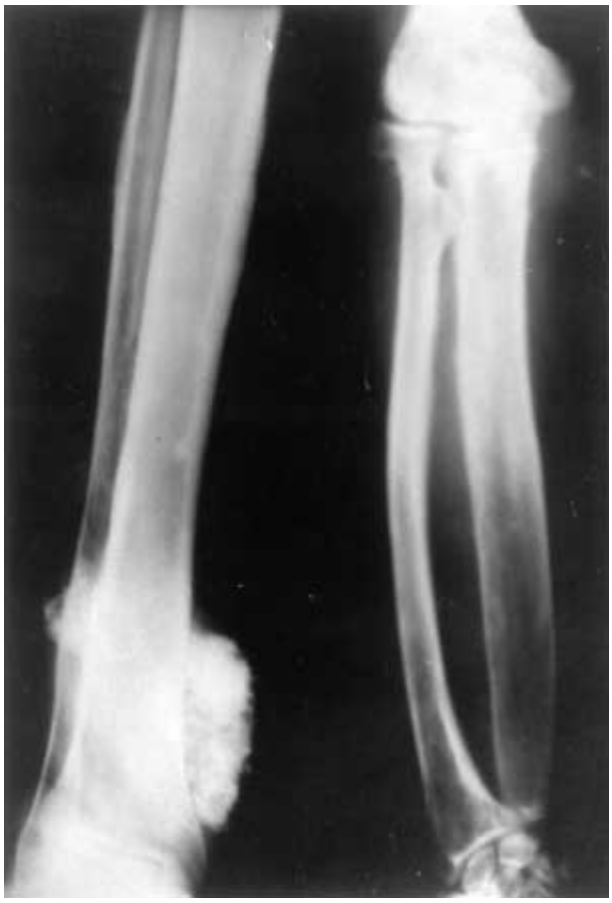

Fig. 3. — Radiograph of the right forearm and right leg shows the diaphysitis with some calcific areas in the marrow, the so-called 'calcific myelitis'.

different sittings, as they were quite large and infiltrating tendons and muscles, making dissection tedious. The tumours were polycystic within a thin fibrous capsule. On sectioning, the mass showed yellowish pasty calcareous material and there was a gritty sensation. Milky fluid was found in the elbow swelling.

Specimens of the excised tissues were reviewed independently by two different pathology centers, which gave concordant opinions. The sections showed uniform appearance of long slender, spindle-shaped fibroblasts closely arranged in bands and fascicles surrounded by varying amounts of collagen. The degree of cellularity varied at places with hypocellular hyalinised and calcified foci alternating with compact cellular areas. There was

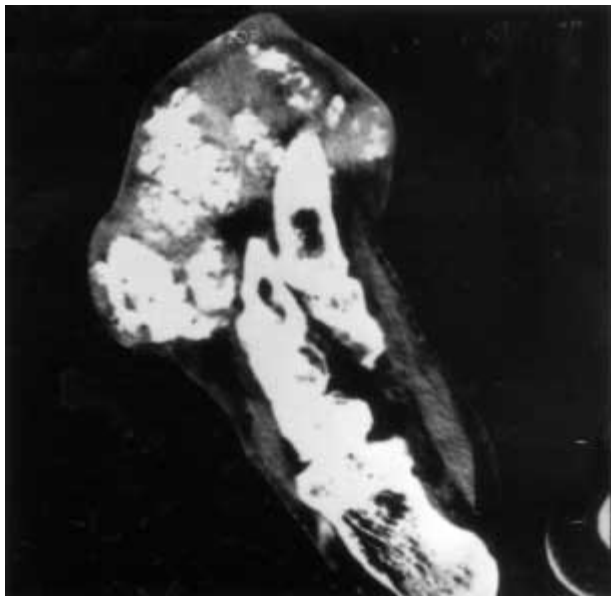

Fig. 4. — CT examination of the foot showing amorphous, calcium deposition, uncalcified soft tissue component. There is no involvement of the underlying bone.

dense focal infiltration of polymorphonuclear cells but few eosinophils and lymphocytes around the blood vessels and at other locations. There was no evidence of malignant change. The findings were suggestive of tumoral calcinosis.

At follow-up the skin wounds were healed except for the mild superficial infection in the foot and a sinus overlying the back that was excised. The patient was followed up for five years. There is recurrence of swelling in the hand after five years that needs reoperation.

DISCUSSION

The term tumoral calcinosis was first coined by Inclan *et al* (6) though the disease had been earlier reported by Duret (4) in 1899. The exact cause is not known, but the disease has been reported in siblings (1) and hyperphosphatemia was found in affected cases (1). Two theories have been put forward for its occurrence :

1. One theory suggests that an inborn error of phosphorus metabolism is the cause, probably inherited as an autosomal recessive gene (2).

2. The other theory states that mechanical trauma or repeated minor injuries are the essential underlying aetiology.

Whatever the causative factors, the primary lesion occurs from digestion of collagen near joints or bones by cells with the morphology and functions of osteoclasts, accompanied or followed by calcification in the digestion fluid. The process is associated with increased vascularity, and the calcification is a secondary event. The disease is basically the result of metaplasia of connective tissue cells and not of disturbed calcium or phosphorus metabolism (2). The disease has a genetic background and the primary defect is in the collagen, which undergoes calcification (4). Tumoral calcinosis may in some cases be the result of intrinsic proximal tubular defect allowing enhanced renal phosphate reabsorption (2) but we have not found elevated phosphorus levels in our patient.

Radiographs reveal a conglomerate of multiple, rounded opacities with distinct fluid levels in some cases (7). On CT scans, para-articular soft tissue masses with cystic components and septa are present. The dependent aspect of the cyst may show calcium layering resulting in what has been termed the 'sedimental sign' (11). This pattern is indicative of a metabolically active lesion. Another pattern consists of multiple globular components that are uniformly calcified and are metabolically more stable. There are no associated bony abnormalities and despite the large deposits of calcium in the lesions, there is no evidence of osteoporosis on the skeleton, as is often observed in patients with renal insufficiency and secondary hyperparathyroidism. Scintigraphy examination with 99^{m} Tc pyrophosphate has been shown to be useful in the identification of the lesion (11).

Morphologically, identical lesions may be encountered in patients with chronic renal disease, uraemia and secondary hyperparathyroidism, but most of these patients are older and show decreasing or abnormally low calcium levels. Tumoral calcinosis-like lesions associated with hyperparathyroidism may develop in patients with renal failure undergoing haemodialysis. Similar calcifying lesions in the soft tissue associated with hypercalcaemia occur in patients with hypervitaminosis D, milk-alkali syndrome and in patients with excessive osteolysis and abnormal resorption of bone. None of these biochemical abnormalities were detected in the present case.

Diaphyseal bone marrow lesions have been described (3). They consist of a periostitis with medullary calcification that could be misdiagnosed radiographically as bone marrow infarction or neoplasm or osteomyelitis. Awareness of this process may obviate biopsy or administration of antibiotics. In our patient we noted enlargement of the right ulna, which was clinically asymptomatic.

Various authors (11) have reported other associated findings like diffuse vascular calcification, dural calcification, wrist arthropathy, appearance of calcium pyrophosphate dihydrate deposition, calcification of articular cartilage and dental abnormalities like pulp stones, none of which were observed in the present case. Dark areas noted inside the lesions represent air inside the swelling that has gained entry from draining sinuses.

A syndrome similar to *pseudoxanthoma elasticum* is also probably a component of this disorder.

Review of the literature shows the following criteria for the diagnosis of tumoral calcinosis :

1. Presence of large, painless, calcified masses in juxta-articular sites.
2. Normal values of serum calcium and phosphorus.
3. No associated renal, metabolic or collagen disorder.
4. Disease manifesting itself before 20 years of age.
5. Evidence of familial/ racial predisposition.
6. Recurrence of lesion, particularly after incomplete excision.

At present an inborn error of phosphorus metabolism is accepted as the primary cause of this disease. The levels of 1, 25-dihydroxy vitamin D are also reported to be high (5). However, in our case the 1, 25-dihydroxy vitamin D levels were low to low/normal. The para-articular masses are thought to arise from bursal calcification, as they never involve the underlying bones.

Complete removal of the tumour cures the condition locally, but similar tumours may appear elsewhere (4). Recurrence after total excision is common and should be treated by early repeat excision (1). Phosphorus depletion therapy produces gross changes in the appearance of the lesions on bone scintiscans (10). A low calcium and phosphorus diet with large oral doses of aluminium hydroxide-containing antacids also improves the condition (8). This may be of value in patients where the lesions are due to hyperphosphataemia or renal disease. It was not tried in the present case, as the phosphorus levels were normal.

REFERENCES

1. **Balachandran S, Abbud Y, Prince MJ, Chausmer AB.** Tumoral calcinosis : Scintigraphic studies of an affected family. *Br J Radiology* 1980 ; 53 : 960-964.
2. **Baldursson H, Evans EB, Dodge WF, Jackson WT.** Tumoral calcinosis with hyperphosphatemia. *J Bone Joint Surg* 1969 ; 51-A : 913-925.
3. **Clarke E, Swischut LE, Hayden CK.** Tumoral calcinosis, diaphysitis and hyperphosphatemia. *Radiology* 1984 ; 151 : 643-646.
4. **Duret MH.** Tumeurs multiples et singulières des bourses séreuses (endothéliomes peut-être d'origine parasitaire). *Bull Mém Soc Anat (Paris)* 1899 ; 74 : 725-733.
5. **Hacihanefioglu U.** Tumoral calcinosis – A clinical and pathological study of eleven unreported cases in Turkey. *J Bone Joint Surg* 1978, 60-A : 1131-1135.
6. **Inclan A, Leon P, Gomez CM.** Tumoral calcinosis. *JAMA* 1943 ; 121 : 490.
7. **Kotwale TM, Bohrer SP.** Tumoral calcinosis with fluid levels in the tumour masses. *AJR* 1974 ; 120 : 461-64.
8. **Mozaffarian G, Lafferty WF, Pearson HO.** Treatment of tumoral calcinosis with phosphorus deprivation. *Ann Int Med* 1972 ; 77 : 741-745.
9. **Noyez JF, Murphree SM, Chen K.** Tumoral calcinosis : a clinical report of eleven cases. *Acta Orthop Belg* 1993 ; 59 : 249-254.
10. **Steinherz R, Chesney RW, Einstein B.** Elevated serum calcitriol concentrations do not fall in response to hyperphosphatemia in tumoral calcinosis. *Am J Dis Child* 1985 ; 139 : 816-819.
11. **Thompson JG.** Calcifying collagenolysis (Tumoral calcinosis). *Br J Radiology* 1966 ; 39 : 526-532.
12. **Veress B, Malik OA, Hassan AM.** Tumoral lipocalcinosis : A clinical study of 20 cases. *J Pathology* 1976 ; 119 : 113-118.