Case of multiple peri-articular masses on bone scintigraphy

Dr S Masikane[1], Dr LM Harry[1], Dr V Pilla[1], Dr L Gabela[1] , Dr TS Nxasana[1], Dr M Patel[1], Dr BP Hadebe[1] and Prof M Vorster[1]

1. University of Kwa-Zulu natal (UKZN)

Abstract

Tumoral calcinosis is a rare disorder that was initially described as an arthropathy of calcium pyrophosphate dihydrate deposition. It tends to occur more predominantly in the periarticular location of the extensor aspects of the large joints.

The index case is a patient with the history of multiple operations from 2019 to 2021, for tumoral calcinosis of the right knee. The patient was referred to our department with painless periarticular swellings of the right femur and right elbow. Biochemistry demonstrated elevated serum phosphate and normal PTH. Technetium-99m-Methylene Diphosphate (^{99m}Tc-MDP) bone scintigraphy was undertaken, which was consistent with tumoral calcinosis in the suspected sites, as well as identifying other, additional sites of involvement.

Bone scintigraphy has been proven to have good sensitivity, and its ability to scan the whole body in one acquisition gives the modality the added advantage of identifying tumoral calcinosis skip lesions.

Bone scintigraphy is therefore highly recommended in patients with tumoral calcinosis, as it is able to identify lesions both in the suspected and other (skip lesion) sites, which changes management.

Introduction

Tumoral calcinosis is a rare disorder that was initially described as arthropathy of calcium pyrophosphate dihydrate deposition, occurring more predominantly in the periarticular location on the extensor aspect of the hips, pelvis, shoulder, elbow joints, and in the knees commonly seen on the flexor surface[1,2,3].

We report a case of primary hyper-phosphataemic tumoral calcinosis in a young child and its features as seen on plain radiograph, Computed tomography, and the role of bone scintigraphy.

Case report

At initial presentation in 2019, our patient had a minor fall which resulted in swelling of the right knee. Clinically the patient had frontal bossing, microglia, and dental overgrowth. There was no family history of metabolic bone disease or related disorders. CT and X-rays demonstrated peri-articular calcium deposits, with no adjacent bone involvement and the right knee mass was excised.

The mass subsequently recurred along with a new painless peri-articular swelling of the right elbow. Biochemistry revealed hyperphosphatemia with normal Calcium and PTH levels.

Plain radiography (Figure 1) demonstrated features in keeping with tumoral calcinosis in both the right femur and elbow. Computed tomography (Figure 2) indicated periarticular multi-lobulated calcifications with sedimentation, which are in keeping with tumoral calcinosis.

^{99m}Tc-MDP bone scintigraphy (Figure 3), demonstrated multiple foci of increased uptake in the elbows, bilateral femurs, right knee medially, and right tibial shaft, consistent with tumoral calcinosis involving multiple joints and bone marrow.

Biopsies were performed that confirmed the clinical diagnosis, the lesions were excised and the patient was commenced on a low phosphate diet and oral aluminium hydroxide.

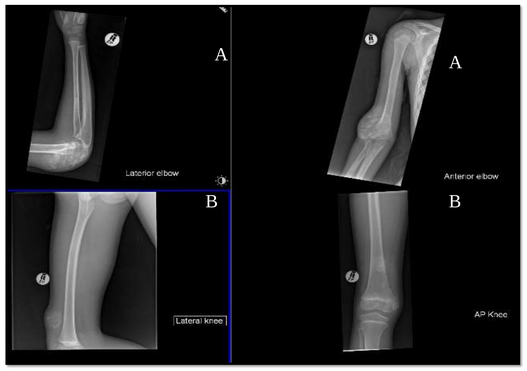

Figure 1: *A. Lobulated periarticular amorphous soft tissue calcification with some areas demonstrating a fluid level, noted in relation to the elbow joint. B. Amorphous lobulated soft tissue calcifications noted in relation to the right knee joint, within the patella-femoral joint and anterior to the distal femoral shaft. The appearance is compatible with tumoral calcinosis. No fluid levels identified.*

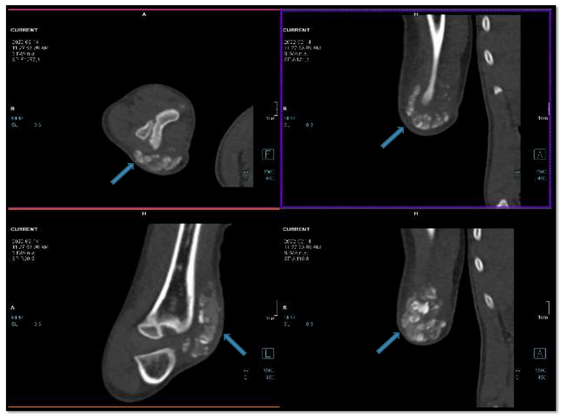

Figure 2*: Elbow. Periarticular multi-lobulated calcifications noted in relation to the knee joint (anteriorly and medially) and the elbow (posteriorly and laterally) demonstrating a cloud like appearance and sedimentation levels. Sclerotic foci are noted within the medulla of the proximal radius and ulna and distal humerus.*

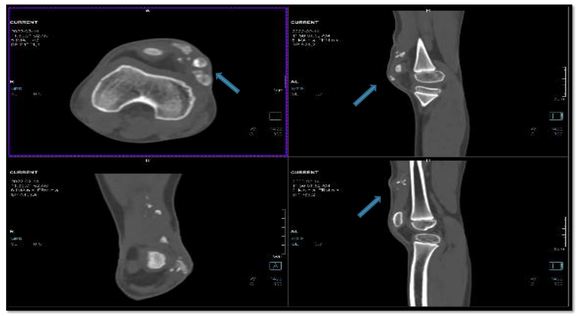

Figure 2: *Knee. Periarticular multi-lobulated calcifications noted in relation to the knee joint (anteriorly and medially) and the elbow (posteriorly and laterally) demonstrating a cloud like appearance and sedimentation levels (seen in relation to the elbow calcification).*

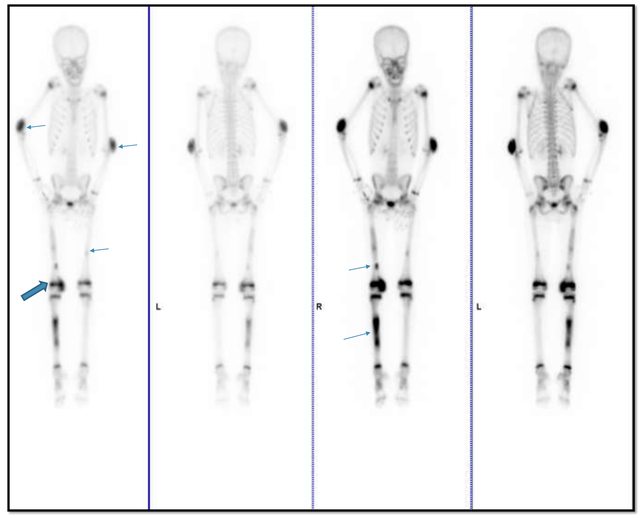

Figure 3: *Multiple foci of increased uptake is seen in the: Extensor aspect of bilateral elbows, Patchy multi-level uptake in bilateral distal femoral shafts, medial aspect Right knee joint, and right tibia shaft. Big arrow depict the know primary lesion, and small arrow indicating all lesions only seen on bone scintigraphy*

Imaging Features

Plain Radiography

Demonstrate multi-lobulated cystic calcification with fluid levels, phenomenon called sedimentation signs. Sedimentation results when films exposed with a horizontal beam shows the fluid calcium level. Also the so called 'chicken-wire' pattern of lucencies, which are well-circumscribed nodular masses, and multi-lobulated calcification.

Computer Tomography

Better outlines the disease, with clear lesion margins and sedimentation as seen on radiography. It further indicates the state of activity of the lesion and the likelihood of continued growth.

Bone Scintigraphy

^{99m}Tc-Methylene diphosphate (MDP) demonstrates focal areas of intensely increased uptake on delayed images.

Discussion

Tumoral calcinosis, is a benign familial metabolic dysfunction of phosphate regulation, which results into high levels of phosphate in the plasma leading calcific deposits of hydroxyapatite or amorphous calcium phosphate crystals in soft tissues. It is not only familial but can also results from renal failure, dialysis, vitaminosis D[123]. It is further classified based on the cause or underlying factors into 3 groups, Primary normophosphatemic tumoral calcinosis; primary hyperphosphatemia tumoral calcinosis; and secondary tumoral calcinosis by Smack D et al[4]. The first type has no genetic association with normal levels of phosphate and calcium, and usually seen in young patient with single lesion, it is more sporadic and low recurrence rate after post therapy.

Primary hyperphosphatemic tumoral calcinosis is hereditary, with elevated serum phosphorus and normal serum calcium, decreased fractional phosphate excretion and increased 1,25-dihydroxyl-vitamin D synthesis. This type has a higher recurrence rate, and is not confined to single lesion, but can affect multiple areas including periarticular regions, vessels, cranium, and the teeth. The incidence is higher in young black males. Secondary tumoral calcinosis, occurs secondary to systemic disease that results into ectopic calcium deposits, e.g. chronic renal failure with secondary hyperparathyroidism, hypervitaminosis D, sarcoidosis, and Milk-alkali syndrome. Tumoral calcinosis is noted to have a hyperphosphatemia threshold of about 5.7mmol/L to develop.

Treatment

Deprivation of phosphate is one of the important mechanism to manage the disease. The mass is surgically excised, but unfortunately has a high recurrence rate-especially the primary hyper-phosphatemic tumoral subtype. The combination of surgical excision, dietary and medical phosphate management has been shown to be most effective[5].

Conclusion

Our patient had no relevant family history, and had recurrent episodes. The onset in this case was sporadic, and trauma was incidental.

The patient was treated multiple times, with multiple surgical excisions over a period of more than 3 years. Management was directed by focal complaints and confirmed by localised conventional image investigations.

Bone scintigraphy demonstrated multiple skip lesions (which were asymptomatic), with bone marrow involvement, which necessitated a change in patient management. This highlights the importance of bone scintigraphy in suspected and confirmed tumor calcinosis.

References

1. Martinez S, Vogler JB 3rd, Harrelson JM, Lyles KW. Imaging of tumoral calcinosis: new observations. Radiology1990;174:215–22.
2. Kluger G, Kochs A, Holthausen H. Heterotopic ossification in childhood and adolescence. J Child Neurol 2000; 15: 406-13.
3. Yurdoglu C, Ozbaydar MU, Adas M, Ozger H. Familial tumoral calcinosis in three patients in the same family. Acta Orthop Traumatol Turc 2007;41:244-8
4. Smack D, Norton SA, Fitzpatrick JE. Proposal for a pathogenesis-based classification of tumoral calcinosis. Int J Dermatol 1996;35:265–71.
5. Kumaran MS, Bhadada S, Bhansali A, Shriram M, Kumar B. Young boy with multiple periarticular swellings and dischargingsinuses: Tumoral calcinosis. IJP 2004;71:1144.