TUMORAL CALCINOSIS: A CASE REPORT

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ABSTRACT

Enormous periarticular calcification of the soft tissues is a common radiological finding. However, tumoral calcinosis is a rare familial disorder characterized by extensive periarticular calcification which is lobulated and well-demarcated and is mostly seen along the extensor surfaces of the large joints. Since, there are many mimickers of this rare disorder; the radiologist plays a vital role in the diagnosis of tumoral calcinosis and in avoiding unneeded aggressive procedures.

Keywords: Tumoral calcinosis, hyperphosphatemia, calcium deposition, hip joints

Introduction

Tumoral calcinosis is an uncommon familial disorder characterized by massive painless periarticular calcifications secondary to metabolic dysfunction of the phosphate regulation.[1] The calcifications in tumoral calcinosis are typically well demarcated and lobulated which are usually distributed along the extensor surfaces of the large joints. The most frequently involved joints are hip, elbow, shoulder, foot and wrist in descending order.[2a] number of different conditions like tophaceous gout, calcific myonecrosis, calcinosis universalis and circumscripta, calcific tendinitis, chronic renal failure and some sinister diseases like osteosarcoma and synovial sarcoma can give similar appearances[2] which warrants better understanding and knowledge of this entity by the reporting radiologists who plays a critical role in making the correct diagnosis and to help in avoiding unnecessary invasive procedures.

Case Report

A 36-year-old female presented to our department with the history of bilateral hip joint pain for eight years. On general physical examination, there was generalized swelling of bilateral thighs. Subsequently, she was advised CT scan of the lower limbs which revealed extensive lobulated amorphous and heterogenous bilateral soft tissue calcifications involving the proximal thighs. The calcification was centered over the greater trochanter of bilateral femora causing their scalloping. No intra-articular extension of joint effusion was seen. (Fig.1a-c). Later on, she underwent MRI of lower limbs on the same day which showed extensive calcification in bilateral proximal thigh sencasing the bilateral proximal femoral shafts. The mass was also abutting the femoral vessels bilaterally without vascular encasement. Furthermore, the mass was also reaching upto bilateral sciatic foramina; potentially abutting the sciatic nerves bilaterally. On post contrast images heterogeneous enhancement of the mass was seen most likely secondary to the enhancement of fibrous septa and calcification (Fig.2a-d). On the basis of these radiological findings the diagnosis of tumoral calcinosis was made which was then confirmed on histopathology.

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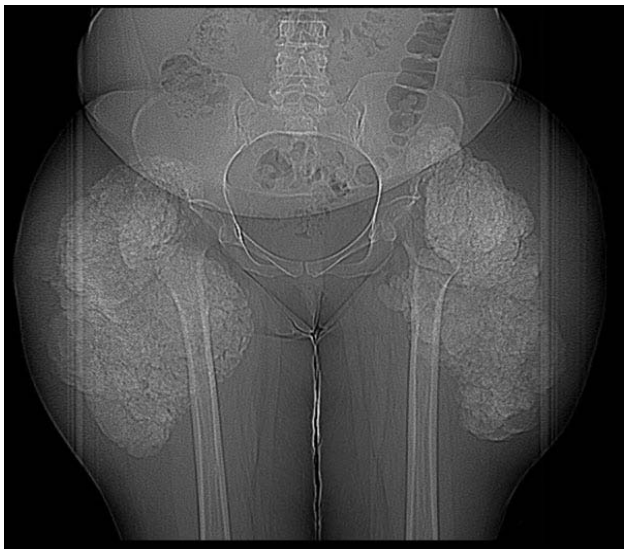

Figure 1a: Scout view from CT showing dense cloud like calcific masses around bilateral hip joints extending into the proximal thighs causing significant overlying soft tissue swelling.

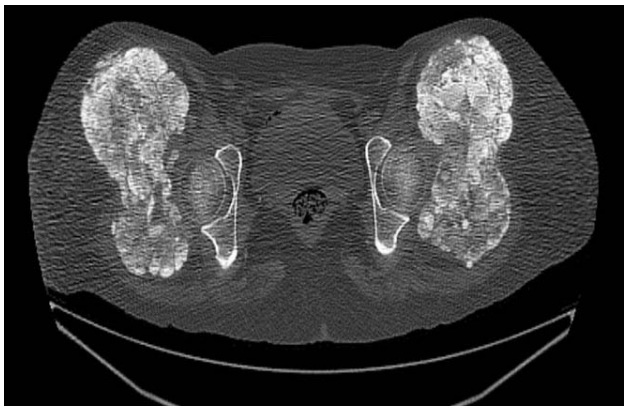

Figure 1b: CT scan, axial view (bone window) showing lobulated bilateral soft tissue calcification involving proximal thighs.

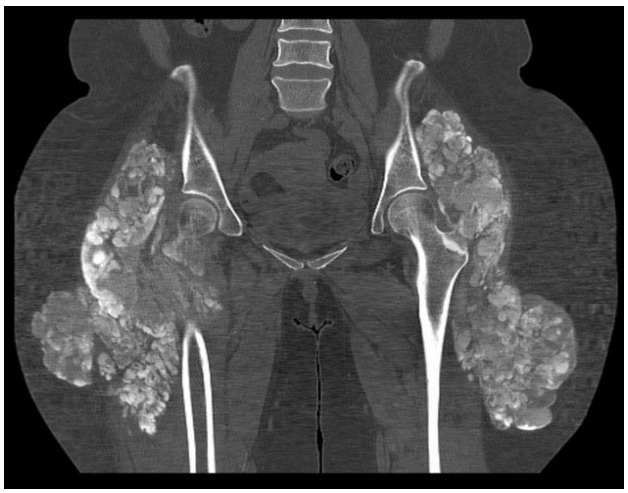

Figure 1c: CT scan, coronal view (bone window) showing bilateral soft tissue calcification involving proximal thighs and scalloping of bilateral greater trochanters.

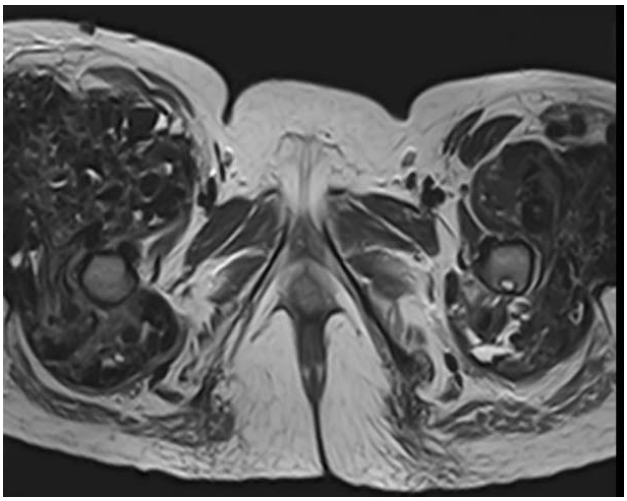

Figure 2a: MRI axial T2WI showing bilateral large areas of calcification returning low signals in proximal thighs along the lateral aspect of hip joints

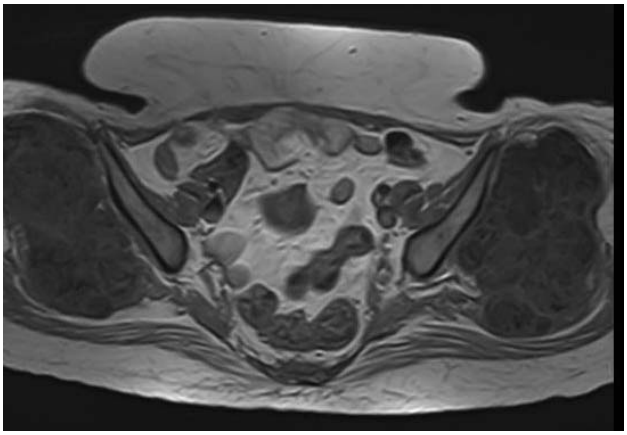

Figure 2b: MRI axial T1WI: showing bilateral large areas of calcification returning low signals in proximal thighs along the lateral aspect of hip joints

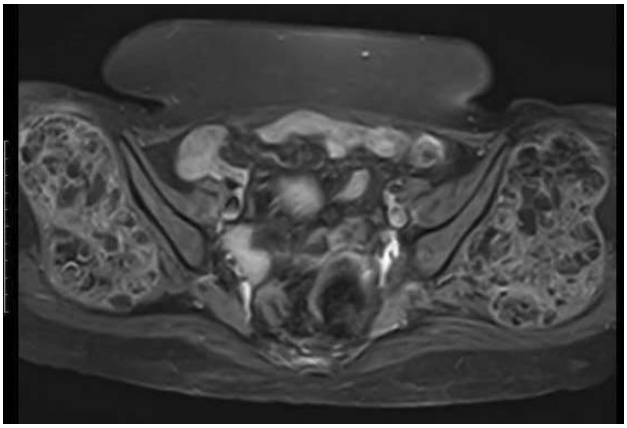

Figure 2c: MRI axial T1 post contrast images showing heterogeneous enhancement of the mass was most likely secondary to the enhancement of fibrous septa and calcification

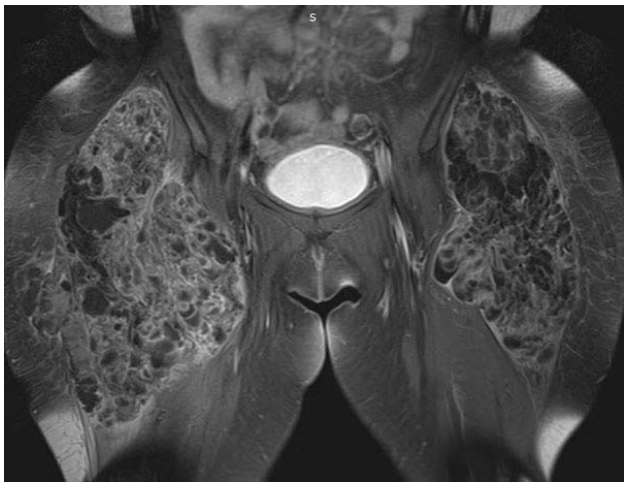

Figure 2d: MRI coronal T1 post contrast images showing heterogeneous enhancement of the mass was most likely secondary to the enhancement of fibrous septa and calcification

Discussion

Tumoral calcinosis was first described by Giard and Duret in 1898 and 1899 respectively.[2] Subsequently the disease process was further studied by Teutschlaender during 1930 to 1950.[3,4] In 1943, Inclan et al also published this disorder in the American literature describing its clinical and metabolic features and differentiating points from other causes of metabolic and dystrophic calcifications.[3] The tumoral calcification commonly presents in the childhood or early adolescence as lumps or swellings in the vicinity or around a joint and is usually seen in African-American females.[4]

Radiologically, on plain x-ray films these appear is cloud-like (amorphous and multi lobulated) calcified masses in periarticular regions. Sometimes, these masses show fluid-calcium levels which is referred to as sedimentation sign.[5] On CT scan, these masses can be seen as cystic collections with low-density centers and calcified walls showing fluid-calcium levels; indicative of metabolically active lesions and multilobulated masses with homogenous calcifications which are considered metabolically stable. Whereas, on MRI these calcified lesions show predominantly hypointense signals on T1 and T2 weighted images. Martinez et al reported two MRI patterns of tumoral calcinosis; i-e nodular pattern with areas of heterogeneous hyperintense signals and signal voids and diffuse hypointense signals.[6]

On aspiration of these lesions tooth paste like whitish color specimen is obtained which on histopathological evaluation consists of calcium phosphate, calcium hydroxyapatite and calcium carbonate. These lesions are usually treated with surgical excision; however, there is increased recurrence rate.[4]

Conclusion

Tumoral calcinosis is a hereditary disorder with a large number of mimickers making it difficult to diagnose. Knowledge of this uncommon disease is very essential as the radiologists play a vital role in the diagnosis and management and help in avoiding unneeded invasive procedures.

Conflict of interest: None

Ethical review board: Ethical approval for this case report was given exemption from the institutional ethical review board.

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