

Tumoral calcinosis with emphasis on Magnetic Resonance Imaging

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Section: Musculoskeletal system

Area of Interest: Musculoskeletal soft tissue

Procedure: Diagnostic procedure

Procedure: Education

Procedure: Imaging sequences

Imaging Technique: Digital radiography

Imaging Technique: Conventional radiography

Imaging Technique: MR

Special Focus: Calcifications / Calculi Metabolic disorders Case Type: Clinical Cases

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Patient: 10 years, male

Clinical History:

A 10-year-old male child presented with complaints of painless progressive swelling over the right hip since 2 months. He had similar swellings in the right index finger and elbow. The swelling over the right elbow had been operated before, but there was a recurrence. There was no history of trauma, fever or weight loss.

Imaging Findings:

Radiograph of right hip joint (Fig. 1), showing a large multiglobular calcific mass overlying the hip joint, with some globules showing fluid-calcium levels, termed "sedimentation sign". Radiographs of right elbow (Fig. 2) and right hand (Fig. 3) showing soft tissue calcifications along the extensor aspect of elbow and radial aspect of terminal phalanx of 1st digit. Underlying bones and joint were normal.

T1 weighted axial MR image (Fig. 4) showing heterogeneous isointense encapsulated soft tissue mass in the subcutaneous plane around the hip joint, displacing and insinuating between the gluteal muscles. T2 weighted (Fig. 5) and STIR images (Fig. 6), showing heterogeneous, multiloculated, predominantly hyperintense cystic mass with some cysts showing "shading" and "fluid levels". The septations between the cysts are iso to hypointense. On post contrast imaging (Fig. 7), these septae show marked heterogeneous enhancement with solid enhancing areas suggestive of "pseudo-tumoral" appearance. Gradient sequence (Fig. 8) shows areas of blooming in septae.

Discussion:

Tumoral calcinosis is strictly speaking a disease caused by a hereditary metabolic dysfunction of phosphate regulation associated with massive periarticular calcinosis [1]. It is characterised by the presence of large calcified soft tissue masses around the joints.

On histopathology, the masses are multiloculated cysts with walls composed of dense fibrous tissue and containing a viscous semifluid suspension of calcium triphosphate or carbonate salts in albumin [2].

The diagnosis of tumoral calcinosis is one of exclusion [3].

Radiographs show a typical imaging appearance of amorphous, multilobulated calcifications which can demonstrate

fluid-calcium levels, termed “sedimentation sign” [1]. Osseous destruction of adjacent bones is distinctly absent [1].

On MR imaging two distinct patterns have been observed (a) a diffuse, lower-signal-intensity pattern or (b) a bright, nodular pattern with alternating areas of high signal intensity and signal void [1].

In our case, “bright nodular pattern” was observed which can be explained by high signal intensity of the supernatant fluid on T2 weighted images while the calcium sediment produces a low signal intensity [3]. The isointensity to hypointensity of the septae on T1 and T2 weighted images with blooming on Gradient sequences could be explained by the calcium incrustations along the inner layers [3]. Marked enhancement of the septae and capsule is due to the connective tissue with varying degrees of vascularization and inflammatory reaction [3].

Pseudo-tumoral appearance of enhancing areas, which are hypointense on T1 weighted images and heterogeneous signal intensity on T2 weighted images with marked post-contrast enhancement could be due to collapsed cyst walls [3].

In conclusion, tumoral calcinosis is a rare benign entity occurring due to phosphate metabolic dysfunction, resulting in large soft tissue masses with areas of calcification predominantly along the extensor aspects of large joints.

MR imaging is useful to demonstrate the location, extent and relationship of the calcifications with adjacent structures. MR imaging may show unusual signal characters like heterogeneous hyperintensity of the cysts & intensely enhancing pseudo-tumoral areas and septae which can be mistaken for other conditions.

Correlating the radiographic findings and knowledge of these varied MR appearances, along with clinical history and blood parameters helps in narrowing down the differentials and arriving at the correct diagnosis.

Differential Diagnosis List: Primary hyperphosphatemic tumoral calcinosis of right hip, right elbow and finger., Metastatic calcification in Chronic Renal Failure or hyperparathyroidism, Myositis ossificans

Final Diagnosis: Primary hyperphosphatemic tumoral calcinosis of right hip, right elbow and finger.

References:

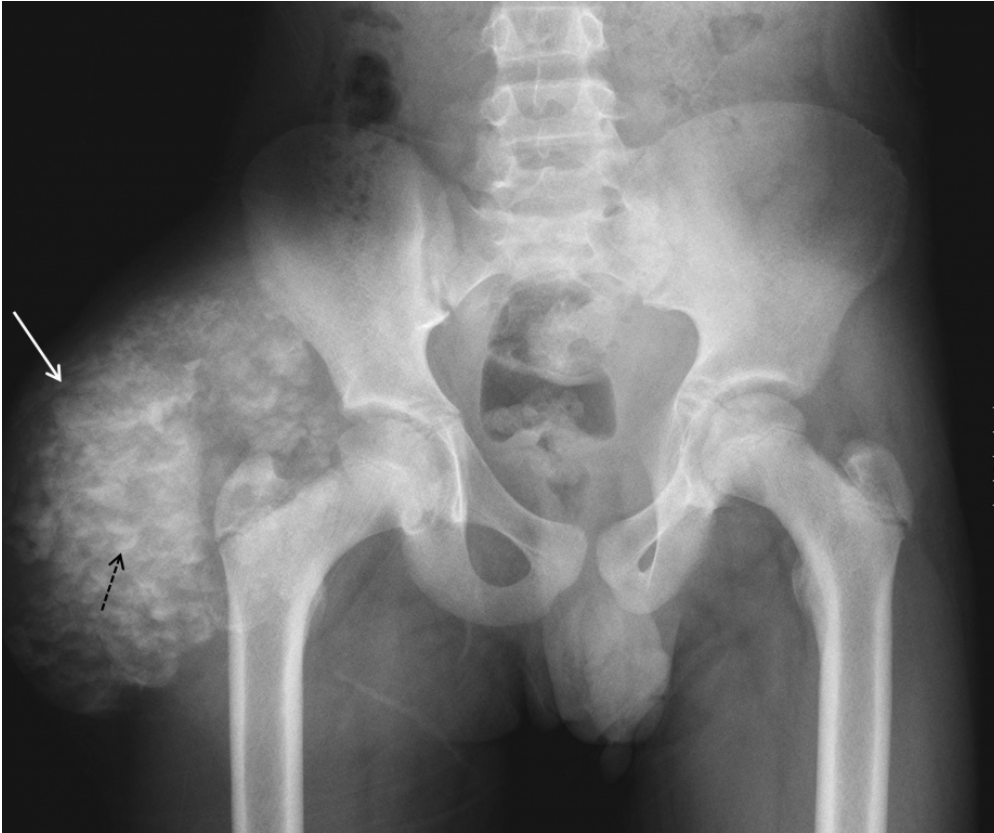
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Rowe LJ, Yochum TR (2005) Arthritic Disorders. Yochum TR, Rowe LJ eds. Essentials of Skeletal Radiology 3rd edition Lippincott Williams & Wilkins 2005: 1112

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Figure 1

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Description: Antero-posterior radiograph of the right hip joint (A) showing a large multiglobular calcific mass [arrow] overlying the hip joint, with areas of fluid-calcium levels [black dashed arrow]. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 2

a



Description: Soft tissue swelling with areas of globular calcification along the extensor aspect of the elbow joint [arrow]. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 3

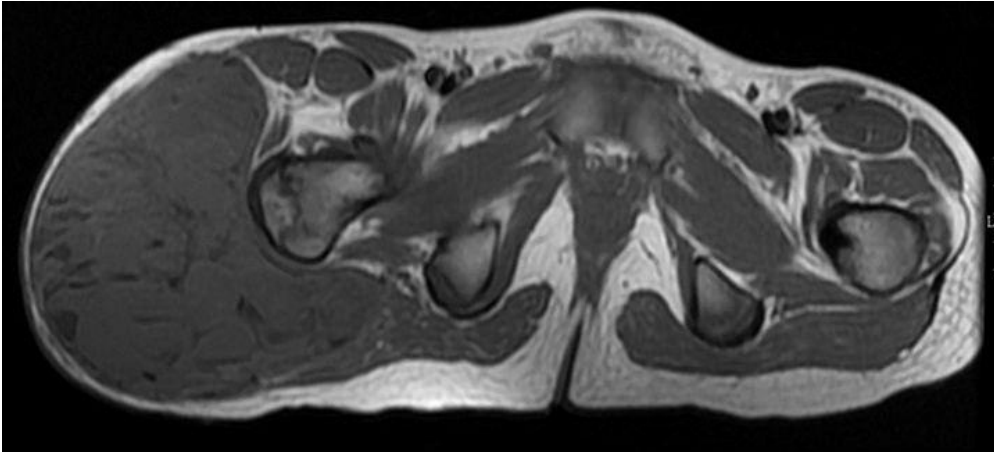
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Description: Soft tissue swelling with areas of calcification adjacent to the terminal phalanx of 1st digit on its radial aspect [arrow]. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 4

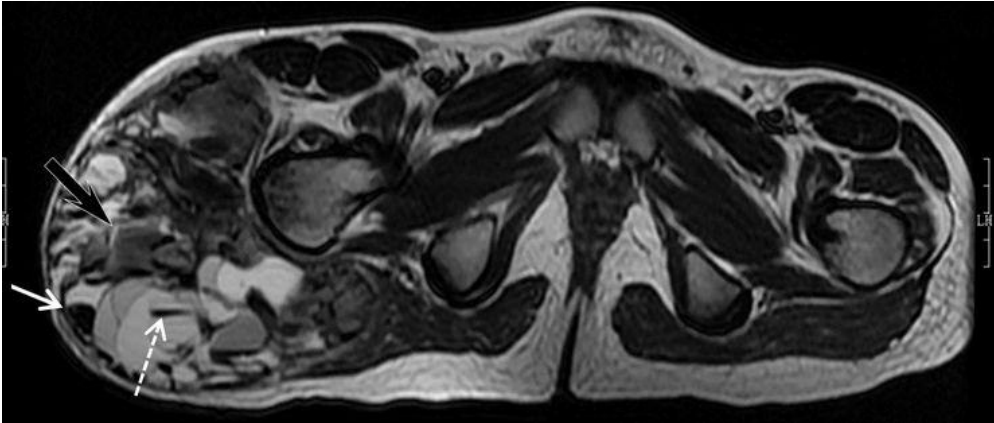
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Description: Heterogenous isointense well encapsulated soft tissue mass in the subcutaneous plane around the right hip joint, displacing and insinuating between the adjacent gluteal muscles. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 5

a

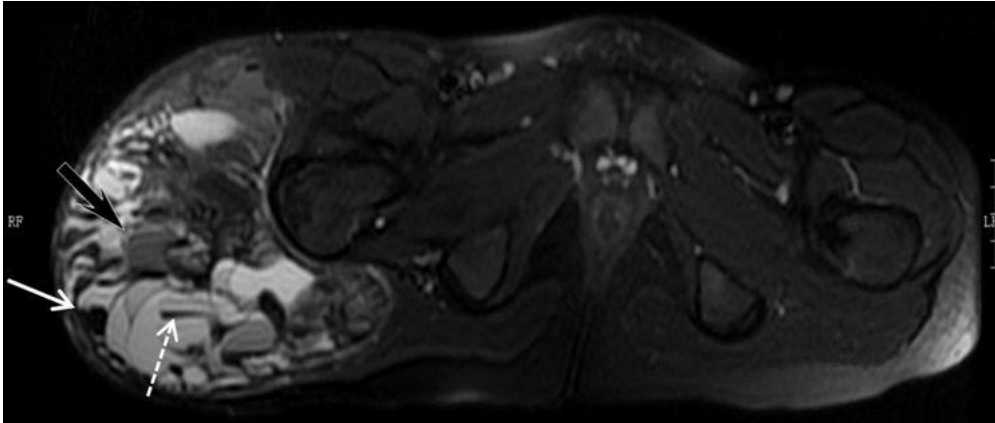


Description: Heterogenous, multiloculated cystic soft tissue mass , with foci of interspersed hypointensity [arrow]. Few cysts showing “shading” [black arrow] and “fluid levels” [white dashed arrow].

Origin: Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 6

a

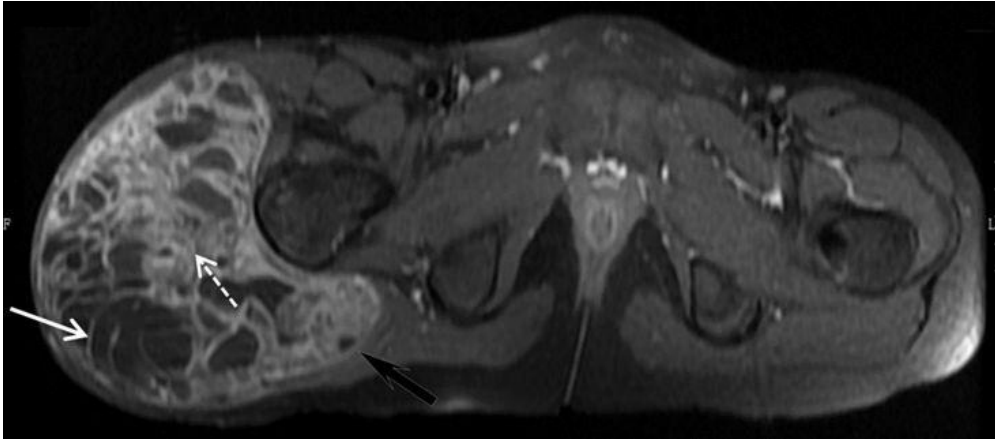


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Few cysts showing “shading” [black arrow] and “fluid levels” [white dashed arrow]. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 7

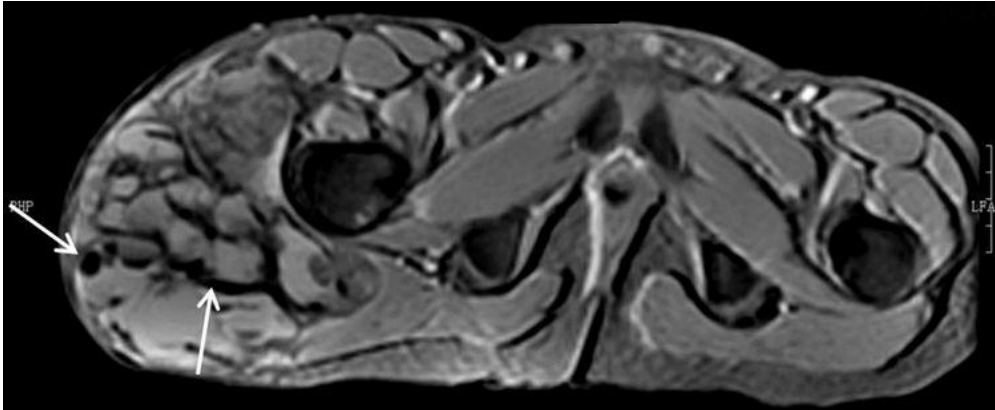
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Description: Mass displacing and insinuating between the adjacent gluteal muscles showing heterogenous enhancement of the septae [arrow], the capsule [dashed arrow], & solid enhancing pseudo-tumoral areas [black arrow]. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.

Figure 8

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Description: Areas of intense septal blooming [arrow] are seen within the mass. **Origin:** Ramakrishna N, Department of Radiology, Kasturba Medical College, Manipal, India.