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Case Report

A rare case of idiopathic tumoral calcinosis of bilateral hip joint: A case report

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ABSTRACT

Tumoral calcinosis or Teutschlaender disease is a relatively rare pseudotumor of unknown origin, characterized by the accumulation of calcium salts in peri-articular soft tissues despite normal plasma levels of calcium and parathyroid hormone. We report a case of tumoral calcinosis in a 26-year-old male patient presenting with a firm mass of soft tissue and limitation of bilateral hip joint. Laboratory investigations revealed a serum calcium level of 10.1mg/dl (Normal) and a serum phosphorous level of 4.2 mg/dl (Normal). PTH level was within normal limit (12.9 pg/dl) with vitamin D level of 40.2 nmol/L (Insufficient). Patient was taken up for surgery, en-bloc excision was done. A biopsy of the soft-tissue masses was sent for histopathology, which was suggestive of lobules of calcific material surrounded by histiocytic giant cells and was diagnosed as tumoral calcinosis. After 6 months of followup patients is doing well with good range of movement at left hip joint with no signs of recurrance. In coclusion, early recognition of Tumoral calcinosis and appropriate management are crucial to mitigate symptoms and improve patient outcomes.

1. Introduction

Idiopathic tumoral calcinosis (TC) in both the hip joint is a rare medical condition in which there is abnormal accumulation of calcium phosphate crystals on periarticular soft tissues. The tumour tends to present as insensitive, tough, small lumps or nodules which can undergo gradual increase in size over time and might cause a lot of discomfort and obstruct the range of motion of the affected joint. The etiology of TC is unknown, but it's suggested to be related to the disturbance in phosphate metabolism or the renal impairment which leads to hyperphosphatemia and the subsequent precipitation of the mineral in the soft tissues. It mostly affects young people, almost in their teen and twenties, and can be familial or spontaneous.[1,2] From a clinical point of view, patients can have different symptoms such as chronic mass enlargement, hip pain, restricted range of motion or pressure symptoms. The most common method of diagnosis is imaging studies such as X-rays or CT which shows amorphous and multilobulated cloud like calcification in periarticular tissue of hip joint. The differential diagnosis includes Myositis ossificans, Calcium pyrophosphate deposition diseases (CPPD), Calcinosis circumscripta, Milk alkali syndrome. Surgical excision is the primary treatment for familial and sporadic forms. On gross examination it shows firm, rubbery, unencapsulated, multinodular to multicystic mass and the cut surfaces shows cystic space filled with milky liquid and semisolid gritty material. Microscopically it shows lobular to irregular deposits of amorphous calcium within cystic space.

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2. Case Report

1. We report the case of a 26-year-old male came to outpatient department with the chief complaints of pain and limitation in range of motion over his left hip joint with a palpable mass in Bilateral hip region.
2. The patient was aware of a mass over Bilateral Hip (Left >Right) which was present for 6 years which was insidious in onset and had been progressively increasing in nature with dull aching pain after strenuous exertion additionally he gives history of limitation of range of movement in his left Left hip since 1 year.
3. A plain radiograph of the pelvis and bilateral hip joints AP along with lateral view was done which depicted a typical appearance of amorphous and multilobulated ("cloud-like") calcifications located in a periarticular distribution in both hips.
4. A CT scan, done for better delineation of the calcific masses showed no obvious erosion or osseous destruction by the adjacent soft-tissue masses.[3–7]
5. Laboratory investigations revealed a serum calcium level of 10.1mg/dl (Normal) and a serum phosphorous level of 4.2 mg/dl (Normal). PTH level was within normal limit (12.9 pg/dl) with vitamin D level of 40.2 nmol/L (Insufficient)
6. After routine pre-operative investigations and a pre-anaesthesia check-up, the patient was taken up for surgery, en-bloc excision was done. A biopsy of the soft-tissue masses was sent for histopathology, which was suggestive of lobules of calcific material surrounded by histiocytic giant cells and was confirmatory of tumoral calcinosis.

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After 6 months of followup patients is doing well with good range of movement at left hip joint with no signs of recurrance.

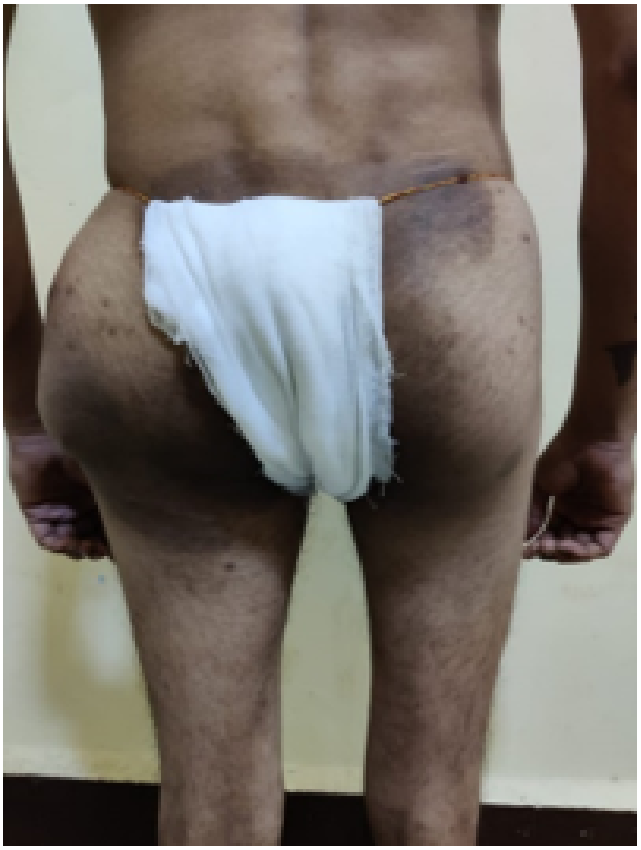

Figure 1: Preoperative clinical pictures

3. Discussion

The evolution of tumoral calcinosis is closely related to the progression of medicine and changing concepts on its pathophysiology. The construct of TC was a painstaking took through clinical experience and investigations which were carried out during the last century. At the outset of the 20th century, there were some case reports of such lesions, which were later described as TC and would show the growth of huge, calcified masses in the soft tissues that surrounded a joint. One of the works probably seminal to its definition was that of Inclan et al. in 1943, describing TC as familial cases characterized by these calcified masses at joints. In the mid-20th century, familiarity with both the familial and sporadic forms of TC disseminated; clinicians and researchers described cases that occurred in a wide range of ages and anatomic locations.[8]

Advances in imaging techniques, like X-rays, CT scans, as well as MRI, made critical technology available for lesion imaging and important characterizations of TC to aid diagnosis and planning for therapy. Genetic studies in the past two decades have uncovered mutations in some primary TC cases in genes concerned with phosphate metabolism, such as GALNT3, FGF23, and KL, thus indicating underlying genetic predispositions.

4. Conclusion

Idiopathic Tumoral calcinosis is a rare disorder in which there is abnormal deposition of calcium in periarticular soft tissue, which usually affects young people. Therefore, early diagnosis and appropriate treatment is essential to reduce symptoms and improve patient outcome.

5. Consent

Written informed consent was taken from the patient.

6. Conflict of Interest

The author declares no conflict of interest.

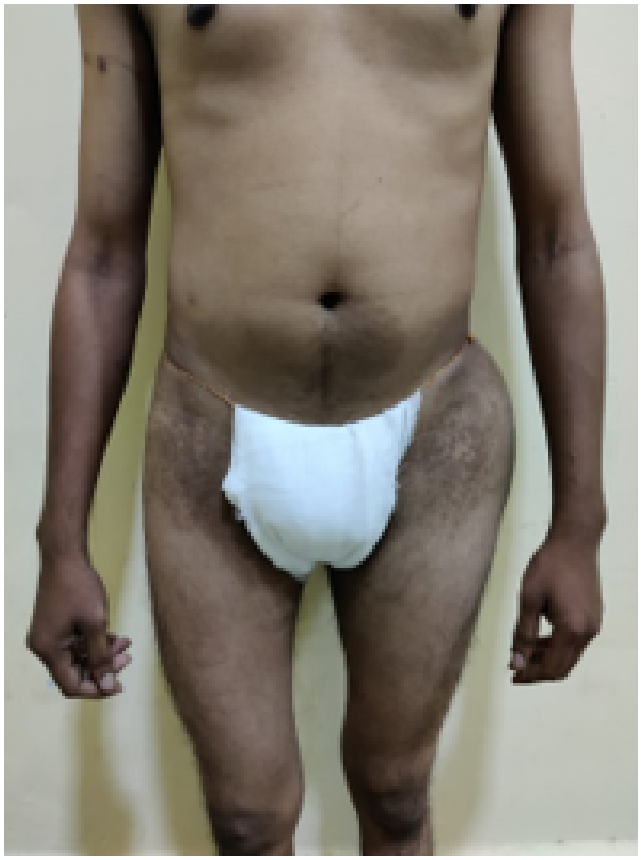

Figure 2: Preoperative clinical pictures

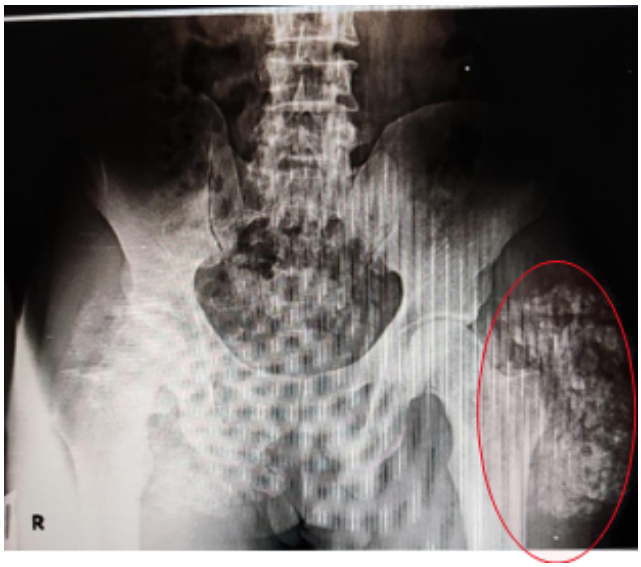

Figure 3: Radiograph pelvis with both hip shows evidence of amorphous multilobulated cloud like calcification in peri articular distribution of both hips.)

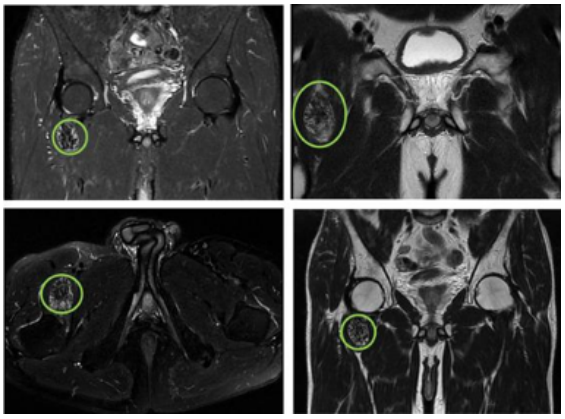

Figure 4: Computed tomography shows evidence of multilobulated calcific mass with no erosion of surrounding joint

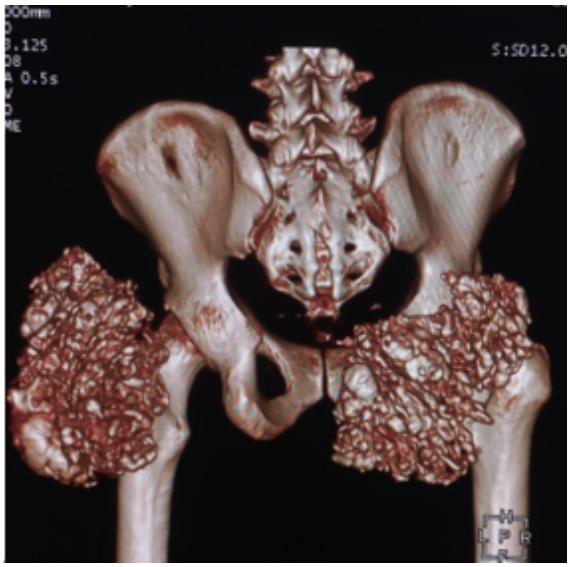

Figure 5: 3d reconstruction image)

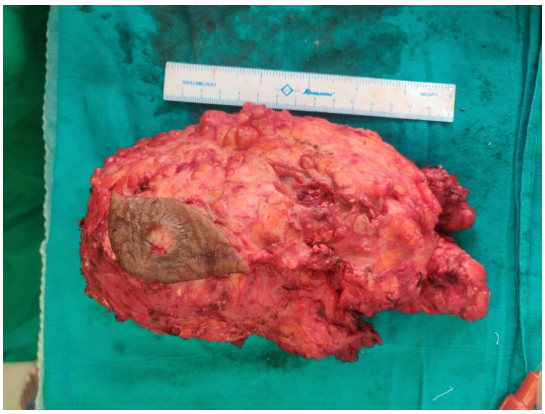

Figure 6: Excised specimen of tumoral calcinosis shows firm, rubbery, unencapsulated, multinodular to multicystic mass

Figure 9: After 6 month follow-up, adequate range of motion of bilateral hip joint with good satisfactory outcome

Figure 10: After 6 month follow-up, adequate range of motion of bilateral hip joint with good satisfactory outcome

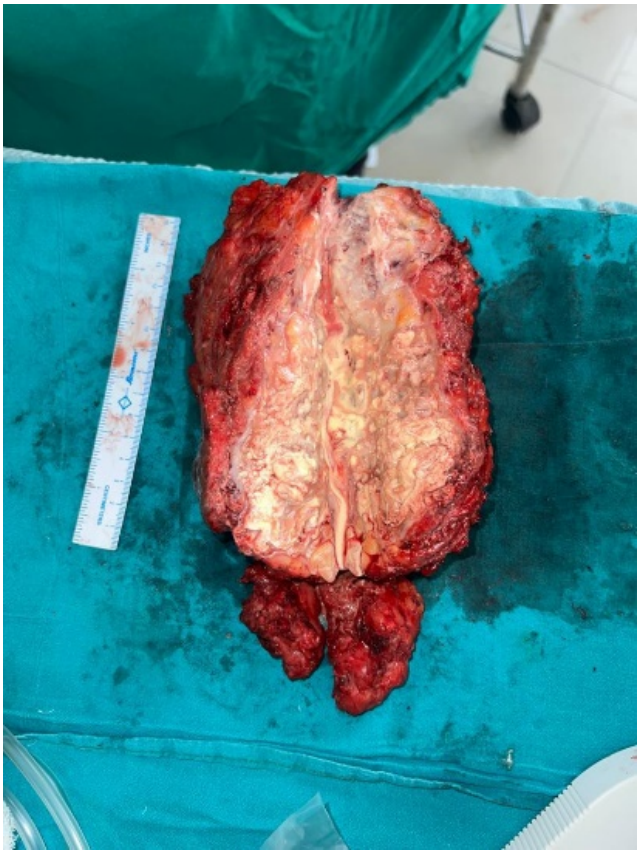

Figure 7: Cut surface shows cystic space filled with milky liquid and semisolid gritty material

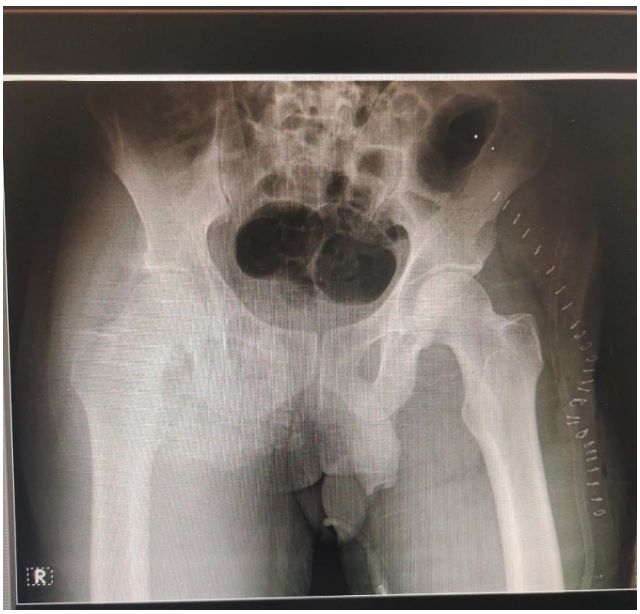

Figure 8: Post operative radiograph after surgical excision of left hip joint

7. Source of Funding

None.

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