Tumoral Calcinosis: A Rare Presentation In Sacrococcygeal Region

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Abstract

Tumoral calcinosis is a rare lobulated soft tissue mass in periarticular soft tissues, Also called as calcium hydroxyappetite deposition in soft tissue. Here is a case of a 35-year-old male, came with a large swelling in sacrococcygeal region since 2 years. The CT scan showed large lobulated mass showing heterogenous iso to hyperdense lesion with no obvious destruction of bones with multiple calcifications and fatty tissue within. Clinically the provisional diagnosis was made as sacrococcygeal teratoma. The mass was excised and the histopathology examination revealed multiple large foci of amorphous calcified material surrounded by foreign body giant cell reaction and fibrosis. Diagnosis of Tuomral calcinosis was given. Huge tumoral calcinosis requiring surgical treatment are rare and should be remembered in the differential diagnosis of recurrent calcified soft tissue lesions mimicking malignancy in sacrococcygeal region. Also, correlation with biochemical parameters helps in classifying the lesions thereby helping the correct management.

Keywords: Amorphous calcified material, Calcified soft tissue lesion, coccyx swelling, Benign

Introduction

Tumoral calcinosis is a rare benign lesion first named by Inclan et al. in 1943.[1] The exact etiology is unknown. However, different hypothesis have been postulated for its pathogenesis.[1] Three subtypes have been described: (1) Primary normophosphatemic tumoral calcinosis (sporadic). (2) Primary hyperphosphatemic tumoral calcinosis (familial). The pathogenesis in this variant is a defect in the phosphate resorption. (3) Secondary tumoral calcinosis which is secondary to diseases like chronic renal failure with concurrent hyperparathyroidism, hypervitaminosis D, and milk – alkali syndrome. Interestingly, tumoral calcinosis has been associated with vitamin D deficiency in an occasional case thereby supporting the hypothesis of a primary renal tubular defect.[2] Here is a case of 35 year old male patient with normal biochemical investigations and only Vitamin D deficiency, presented with a large sacrococcygeal mass since 2 years. The mass was excised and histopathological examination was done. Clinically provisional diagnosis was sacrococcygeal teratoma. Histopathologically, the diagnosis of tumoral calcinosis was given.

Case history:

A 35-year-old male, came with a large swelling in sacrococcygeal region since 2 years. It was associated with pain and discharge from the swelling since 1.5 months. There was no history of trauma.

The CT scan showed large lobulated mass showing heterogenous iso to hyperdense lesion involving posterior sacrococcygeal region with no obvious destruction of bones. The lesion showed multiple calcifications and fatty tissue within. On post contrast images, the lesion showed mild enhancement. Anteriorly the lesion extends upto coccyx and posteriorly upto skin. The lesion measures 22.5x19.5x8.5 cm.

Based on the radiological findings, USG guided tru cut biopsy was performed from the mass. The biopsy suggested fibrovascular tissue with patchy moderate chronic active inflammatory cells including aggregates of neutrophils, extensive calcification, foci of necrosis and myxoid change present. No evidence of high grade anaplasia / mitotically active hypercellular areas was present.

Excision was suggested for definite diagnosis.

The patient was posted for resection of the mass. Biochemical investigations revealed serum levels of calcium to be 9.3 mg/dL. Vitamin D- 13.5 ng/ml (Decreased). Serum phosphate- 3 mg/dL. The sacroccygeal mass was then surgically debulked by surgical gastroenterologist and sent for histopathology examination.

At histopathology department, we received a skin covered huge mass measuring 19x14x7 cm. Overlying skin measures 18x4 cm and was ulcerated. The cut surface was grey white solid necrotic and friable with yellowish white chalky areas. There was no rim of normal tissue at the periphery (Figure1). The histopathology examination revealed multiple large foci of amorphous calcified material surrounded by foreign body giant cell reaction and fibrosis. Foci of necrosis and active chronic inflammation were present. (figure 2,3).

There was ulceration of skin and infiltration of muscle. Inspite of extensive sampling there were no atypical cells or mitotic activity.

A histopathologic diagnosis of tumoral calcinosis was given. The patient is doing well 3 months post operatively without recurrence.

In our case, patient must be having primary normophosphatemic tumoral calcinosis. (Type 1) as all biochemical investigations were normal.

Figure 1: Gross image of Sacrococcygeal region mass

Figure 2: HPE image of Tumoral calcinosis (10x view)

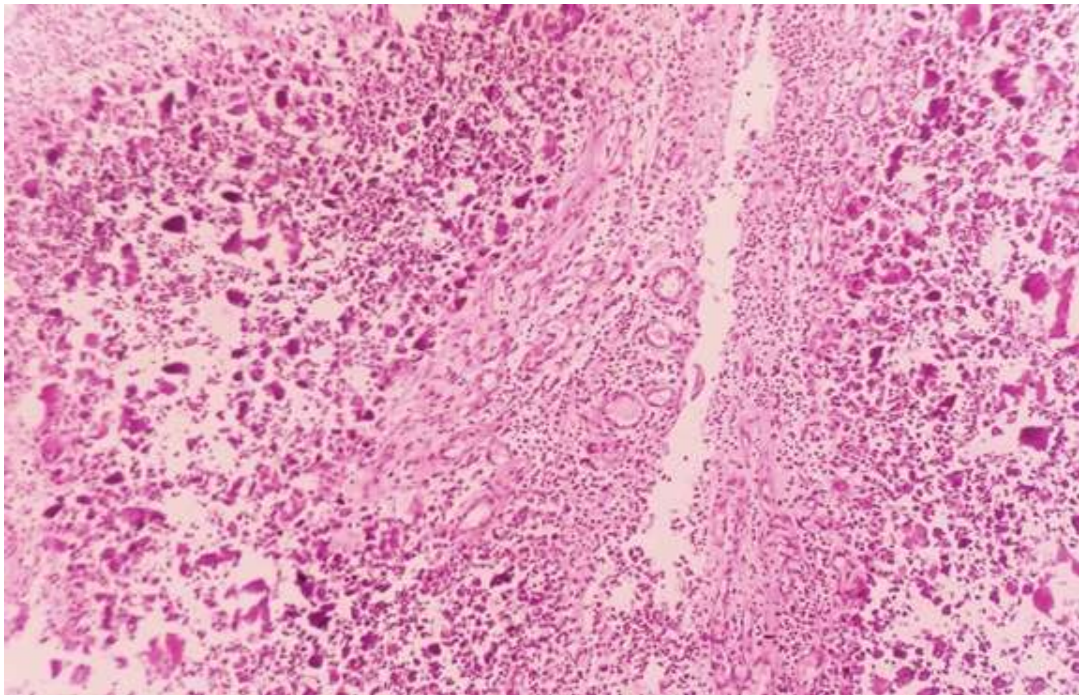
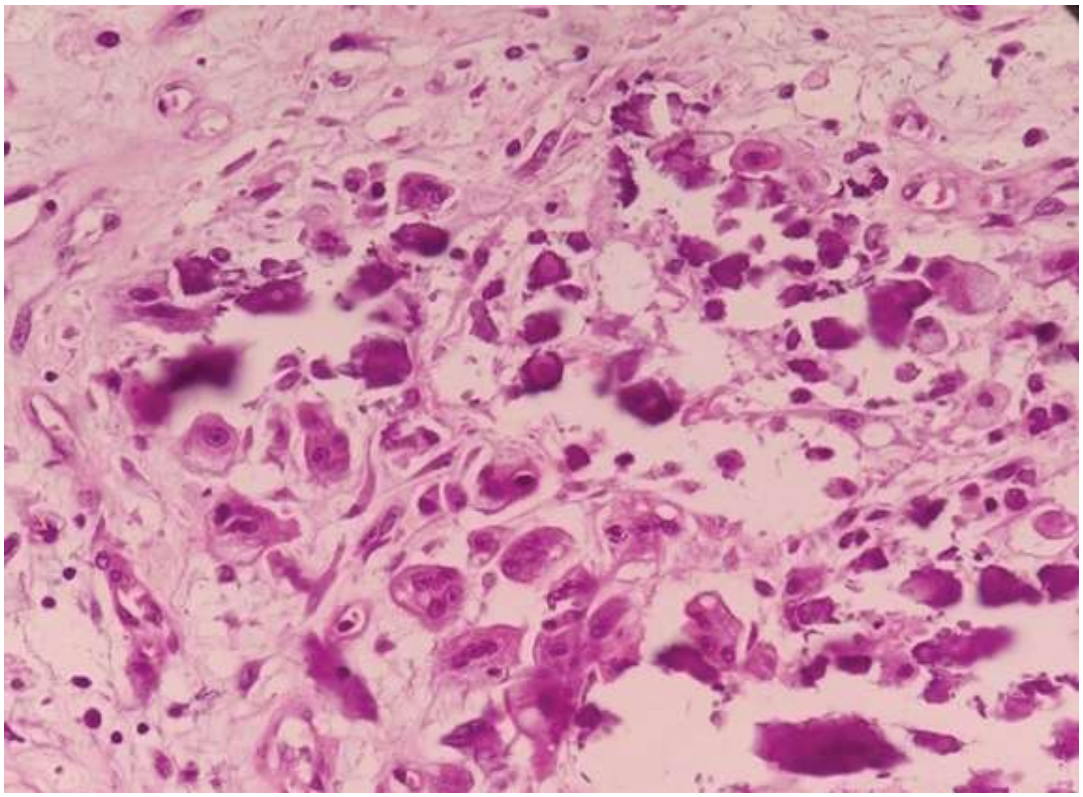


Figure 3: HPE image of Tumoral calcinosis (40x view)



Discussion:

Tumoral calcinosis is a rare benign lesion.The typical presentation of the disease is a small-sized firm to hard mass in the subcutaneous plane usually juxta-articular but rarely involving the joints and commonly seen around the hip, shoulder, and elbow.[3] Occasionally, however, large size can be attained leading to compressive symptoms, pressure necrosis of underlying bony surfaces, and skin ulceration as happened in our patient.[4],[5],[6] When the lesion is large, it may be mistaken clinically for a malignant neoplasm like synovial sarcoma, chondrosarcoma, or an osteosarcoma.[3] . In our case, the location of the mass suggested possibility of sacrococcygeal teratoma. The grey white necrotic areas and the large size seen on gross examination may further favor a neoplastic lesion even pathologically. However, the free flowing opaque milky white chalky material favors a diagnosis of tumoral calcinosis. A biopsy diagnosis may be achieved when a soft tissue neoplasm is suspected.[3] The characteristic histologic feature of amorphous calcified material surrounded by foreign body giant cell reaction and fibrosis distinguishes it from other causes of soft tissue calcification and neoplasms. The histopathologic features however, do not help to differentiate the three subtypes.[1]

The three clinical subtypes need to be distinguished as the management differs. Medical management with phosphate lowering drugs helps in types 2 and 3 and can result in complete resolution of small lesions. Surgical intervention in the form of excision and flap reconstruction is needed for large lesions causing functional impairment, pain, or skin ulceration.[1],[4],[5],[6] Since our patient had no hyperphosphatemia and a large size, a surgical management was done. To the best of our knowledge, sacrococcygeal region is the rarest site of tumoral calcinosis, So far it has not been reported in this region and this is the one of the largest tumoral calcinosis reported in literature.[1],[4],[5],[6]

Conclusion:

Huge tumoral calcinosis requiring surgical treatment are rare and should be remembered in the differential diagnosis of calcified soft tissue lesions mimicking malignancy in sacrococcygeal region. Also, correlation with biochemical parameters helps in classifying the lesions thereby helping the correct management.

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