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*RESEARCH ARTICLE*

# PRIMARY NORMO-PHOSPHATEMIC TUMORAL CALCINOSIS - A RARE ENTITY

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## *Abstract*

Tumoral calcinosis is a rare benign condition, characterized by massive deposition of calcium salts into peri-articular soft tissues. Majority are secondary to underlying chronic disorders like chronic renal failure. Primary Normo-phosphatemicTumoral Calcinosis is a rare entity. I hereby, report a case of primary normo-phosphatemictumoral calcinosis in a 11 years old female patient, managed by complete surgical excision.



## Introduction:-

Tumoral calcinosis is a rare entity. Exact incidence is not available in literatures [1]. The majority cases are secondary tumoral calcinosis [2]. It shows female preponderance; and is usually diagnosed in first two decades of life [3]. The term "Tumoral Calcinosis" was first proposed by Inclan et al in 1943[4].

Tumoral calcinosis is a benign condition, is characterized by massive deposition of calcium salts into peri-articular soft tissues [5]. Most commonly deposited salts are calcium hydroxy apatite crystals and amorphous calcium phosphate. It commonly involves extensor surface of peri-articular soft tissues of hip, elbow, shoulder, ankle, wrist and small joints of hand and foot [6].

**Case Report:**

A 11 years old female attended with chief complaints of non-traumatic, painless swelling over postero-lateral aspect of right elbow for last 3 years. The swelling is gradually increasing in size. On clinical examinations, there was a solitary globular firm mass of size 7cm × 3cm, which was non-tender, non-adherent to skin/underlying structures without neurovascular deficits. Range of motion at elbow joint was full and free. Her past history and family history were unremarkable. Biochemical markers including serum calcium, serum phosphate, alkaline phosphatase, parathyroid hormone wasfound within normal limit.

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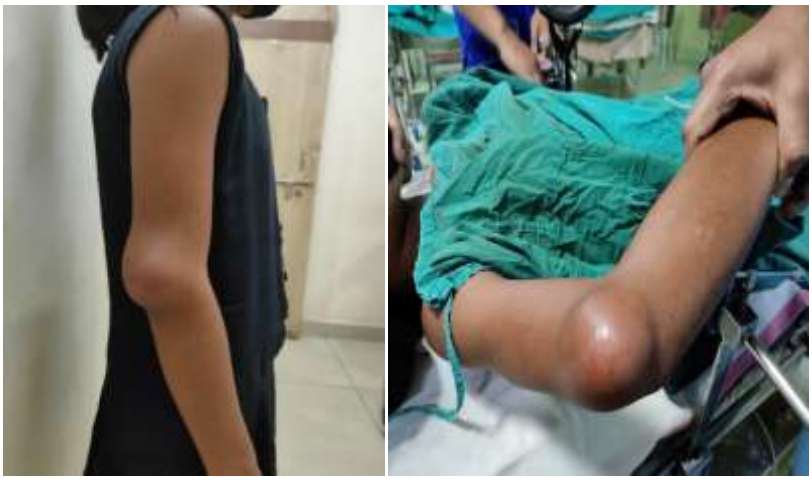

**Figure 1A/1B:-** Pre-operative clinical pictures showing tumoral calcinosis.

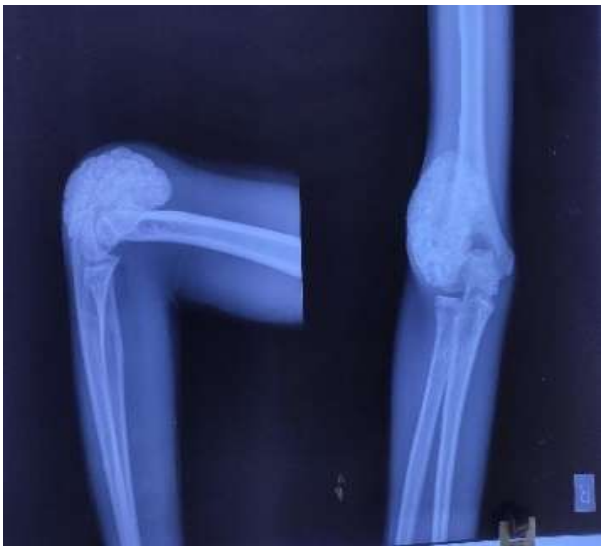

**Figure 2:-** Plain radiograph showing tumoral calcinosis.

**Figure 3A:-** Showing cut section of tumoral calcinosis ,**3B** showing histopathological slide.

**Figure 4A/4B: -** Showing clinical pictures at 18 months follow-up.

Plain radiograph of elbow joint was taken, which showed extra-articular lobulated clusters of calcific deposits. Clinico-radiological examinations including biochemical markers suggested diagnosis of "Primary Normo-phosphatemicTumoral Calcinosis". Pre-operative computed tomography (CT) scan was done. After diagnosis, patient was treated with excisional biopsy through posterior approach of elbow. Gross section of excised material revealed well defined capsulated mass with chalky white granular deposits. Histopathological examination revealed numerous irregular cyst like spaces of varying sizes with fibro-collagenous cystic wall. The cystic spaces were filled with abundant granular calcified materials. These histological features confirmed the diagnosis of tumoral calcinosis. At 18 months follow-up, there was no signs of recurrence/complications.

## Discussion:-

Giard reported first case of calcific deposits in peri-articular soft tissues in European Medical Literature in 1988[7]. Duret, in 1989 also reported similar condition [8].

Exact aetiology of tumoral calcinosis is unknown. In 1996, Smack et al proposed pathogenesis-basedclassification. They mentioned 2 types of tumoral calcinosis. Primary tumoral calcinosis, which is further classified into 2 groups; A) Primary normo-phosphatemictumoral calcinosis. Characteristic features are normal serum calcium and serum phosphate level. The majority cases present before second decade of life. Majority lives in tropical or subtropical regions. B) Primary hyper-phosphatemictumoral calcinosis. Characteristic features are raised serum phosphate but normal serum calcium level. Decreased excretion of serum phosphate leads to this condition due to inactivation of a phosphate excreting hormone. Secondary tumoral calcinosis; is most commonly occurred secondary to underlying disease like chronic renal failure (CRF) [3,9]. Here, in our case all biochemical markers specially serum calcium, serum phosphate; and epidemiological featureswere found similar to the features of primary normo-phosphatemictumoral calcinosis.

Diagnosis is usually made by plain radiographs. It shows characteristic amorphous, multi-loculated clusters of calcific deposits in peri-articular soft tissues. Computed tomography (CT) scan is required as pre-operative tools for delineation of extent of lesion. It shows "sedimentation sign" [10,11]. Microscopically in tumoral calcinosis, cystic spaces are filled with chalky white calcium salts. Cystic wall shows fibro-collagenous tissues [3]. Here, in our case radiologic and microscopic features were found similar to literatures.

Genetic studies can be done for further analysis of this condition [9].

Previous studies show that early surgical excision is the primary treatment modality. There is high rate of recurrence [12]. Here, in our case patient was treated with complete surgical excision. At 18 months follow-up, there was no signs of recurrence/complications.

Considering the dearth of published data on this topic from north-east region of Indian population, this report would serve as an important record for future analysis.

## Conclusion:-

Although primary normo-phosphatemictumoral calcinosis is a rare entity, complete surgical excision is the mainstay of treatment. Primary hyper-phosphatemictumoral calcinosis and secondary tumoral calcinosis require other medical interventions apart from complete surgical excision. Early and complete surgical excision is essential for good functional outcome and no evidences of recurrence/complications.

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