

Tumoral Calcinosis- A Rare Case Report

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ABSTRACT

Introduction: Tumoral calcinosis is a rare condition of unknown etiology. It's a misnomer as they are not true neoplasms characterized by deposition of calcium in soft tissues in periarticular location. Only few cases have been reported.

Case report: Herein we report a case of tumoral calcinosis in a fifteen year old girl with no metabolic abnormalities. Patient was successfully treated with wide local excision and the diagnosis was confirmed by histopathology.

Conclusion: Tumoral calcinosis is a hereditary disease of phosphate metabolic dysfunction but is commonly mistaken for a lesion. The most effective treatment is a combination of surgical excision, phosphate deprivation, and use of acetazolamide. In this case as phosphate levels were within normal limits wide local excision was done.

Keywords: Tumoral calcinosis, Misnomer, Calcium Deposition, Periarticular Unknown etiology.

INTRODUCTION

Tumoral calcinosis is a rare condition of unknown etiology wherein there is calcium deposition in the soft tissue in periarticular location i.e. around joints. It's a MISNOMER. The name indicates calcinosis (calcium deposition) which resembles tumor (like a new growth). They are not true neoplasms - they don't have dividing cells. They are just deposition of inorganic calcium with serum exudate. Children and adolescents (6 to 25 years) are the most commonly affected. They are more common around shoulders, hips and elbows.¹

CASE REPORT

A 15 year old female patient presented to surgical outpatient department with complaints of swelling in the left gluteal region since 6 months. No history of pain associated with the swelling, sudden increase in size of swelling, anorexia, significant weight loss, trauma, restriction of joint movements. No history of similar swellings in the family.

On examination a single, vertically oval, 15x10 centimeters, smooth, firm, non-tender, mobile(both directions)swelling present in the left gluteal region extending into the lateral compartment of the thigh. Skin over the swelling pinchable, no restriction of hip joint movements, neurovascular integrity distal to swelling maintained, no palpable regional lymph nodes. Clinical diagnosis of soft tissue sarcoma was made, fine needle aspiration cytology was inconclusive, core needle biopsy showed calcifications with fibro-collagenous stroma and giant cell reaction without signs of malignancy suggestive of tumoral calcinosis.

Radiological investigations were done which supported our histological diagnosis

X ray Pelvis: reveals evidence of large fairly well defined lobulated lesion with ring like calcifications seen in the soft tissue overlying the left upper end of the femur without periosteal reaction and no adjacent joint involvement.

CT scan: shows a large mass lesion with calcifications and hypo dense soft tissue areas with 20-30HU values. No obvious involvement of adjacent bone. (Fig 1)

MRI: Revealed multiple locules of hyper intensity interspersed with hypo intensity areas (Fig 1)

Laboratory investigations were found to be within normal limits (including serum calcium and phosphorus). Final diagnosis of tumoral calcinosis was made and surgery was planned.

SURGERY

Wide local excision was done and specimen was sent for biopsy which revealed lobular calcifications with giant cell reaction and psammoma formations supporting our diagnosis (Fig 2).

Post operative period was uneventful and there was no recurrence upto an year of follow up.

DISCUSSION

The term Tumoral calcinosis defines a condition in which either single or multiple tumor-like calcified masses are present without any associated calcium metabolism disorder. Calcium deposition in the soft tissue in periarticular location i.e. around joint. It's a MISNOMER. The name indicates calcinosis (calcium deposition) which resembles tumor (like a new growth). They are not true neoplasms - they don't have dividing cells. They are just deposition of inorganic calcium with serum exudates. Children and adolescents (6 to 25 years) are the most commonly affected.¹ They are more common around shoulders, hips and elbows. The term tumoral calcinosis was originally described by INCLAN in 1943.² The pathogenesis of Tumoral Calcinosis remains unclear and several theories have been proposed. Hyperphosphatemia has been described in some patients^{3,4}, while local trauma has been implicated in a few cases.⁵ No metabolic abnormalities were found in our patient and she denied any history of local trauma. The clinical presentation and radiological

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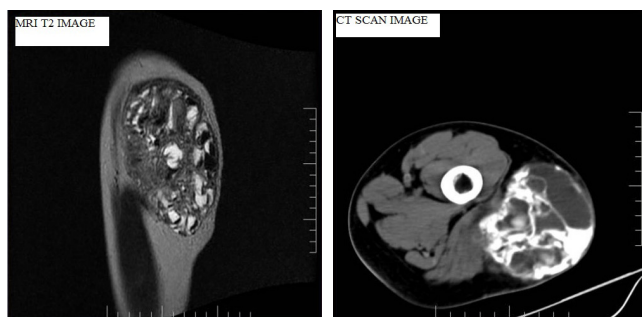


Figure-1: MRI and CT image

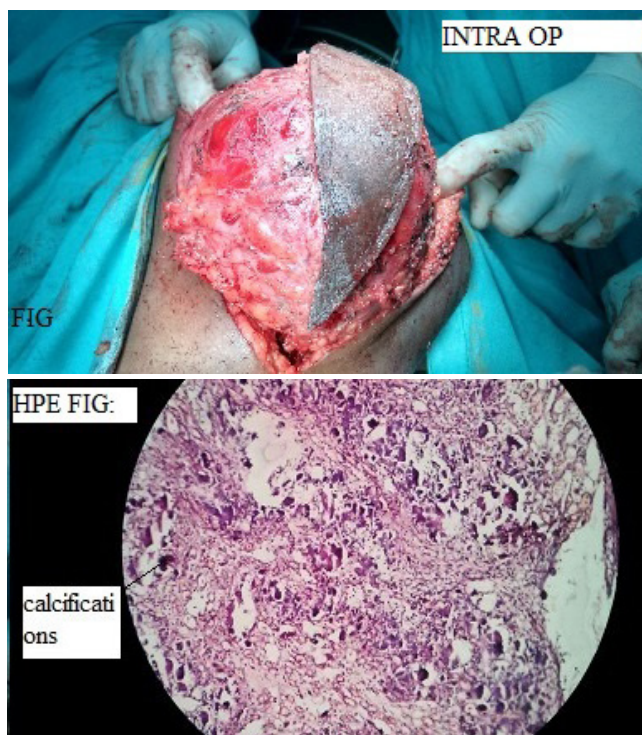


Figure-2: Intra-op and HPE pics

features are typical confirmed by pathological examination. Investigations such as CT scan and MRI are very useful in diagnosing this entity.^{6,7} Histopathological examination serves as an important diagnostic tool in diagnosing tumoral calcinosis and differentiating it from other lesions mimicking it. It is characterized by a central mass of amorphous or granular calcified material surrounded by hyalinized fibrous tissue separating several cavities. The fibrous tissue is bordered by a granulomatous and chronic inflammatory infiltrate. There may be prominent small psammoma-like bodies or calcospherites.⁸ A complete surgical excision along with the deposits is the mode of treatment although recurrences are common.

CONCLUSION

Tumoral calcinosis is a hereditary disease of phosphate metabolic dysfunction but is commonly mistaken for a lesion. The most effective treatment is a combination of surgical excision, phosphate deprivation, and use of acetazolamide. In this case as phosphate levels were within normal limits wide local excision was done.

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