Tumoral Calcinosis at Unusual Site: Diagnosis on Fine Needle Aspiration Cytology.

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ABSTRACT

Tumoral calcinosis (TC) is a rare benign clinical and histological entity. It presents as tumour-like periarticular deposits of calcium in hip, shoulder and elbow regions. Idiopathic tumoral calcinosis (TC) is different from the secondary calcium deposition associated with terminal renal diseases, hypervitaminosis D and milk alkali syndrome with disturbed calcium metabolism. There are no demonstrable abnormalities of calcium metabolism in Idiopathic TC. TC predominantly affects children and young adults and is rare in old age. When TC occurs in old patients, possibility of other differential diagnoses need to be considered and ruled out with the help of morphological examination of the tumorous swelling. Two cases of TC that occurred in 65 and 75 year old females respectively were initially diagnosed on the basis of fine needle aspiration cytology (FNAC) findings of the swellings and later confirmed by histopathological examination of the excised tissue. Diagnostic biopsies were avoided in these patients. Cytological study can prove to be a rapid and reliable method of preoperative morphological diagnosis of tumoral calcinosis.

Keywords: Tumoral calcinosis, morphological diagnosis, fine needle aspiration cytology

INTRODUCTION

Tumoral calcinosis (TC) is a type of idiopathic calcinocis cutis and is characterised by formation of large deposits of calcium in skin and subcutaneous tissues over bony prominences. TC is known to occur in African continent and its occurrence is uncommon in rest of the world[1]. The disorder is seen in healthy children, adolescents and young adults with no abnormality in calcium metabolism[2]. Soft tissue calcinosis that occurs in patients with end stage kidney disease at different body sites is known and accessible to FNAC diagnosis due to subcutaneous location of the tumorous masses.[3] However, when TC occurs in old persons without any specific clinical setting, diagnosis is often missed. Morphological diagnosis by FNAC can be of great help to suggest line of investigations before definitive surgical treatment is planned in these cases.

CASE REPORT

A 65 year old female presented to the surgical outpatient department with slowly enlarging right inguinal region swelling since one year. There was no complaint of pain or discharge from the swelling. There was no history of local trauma or any major illness. Her past and personal history was not significant. The patient was a healthy adult with good general condition and her vital signs were stable.

On local exam an irregular, nodular swelling of 5x5cm covered with normal skin was seen in the right inguinal region above inguinal ligament near right anterior superior iliac spine [Figure 1a]. The swelling was firm in consistency, mobile and nontender. No punctum or skin changes were noted. The patient did not have difficulty in right joint movements.

With clinical diagnosis of soft tissue tumour, the patient was referred for FNAC study. Whitish material was obtained on aspiration with 22-gauge needle. Smears made from the aspirate were fixed with Alcohol and stained with HE, Pap stains, while dry smears were stained with MGG stain. The smears revealed abundant amorphous crystalline refractile material. No cellularity was observed [Figure 2 a and b].
In view of the periarticular location of the swelling, the whitish granular nature of the aspirate and the cytological features, the diagnosis of tumoral calcinosis was conveyed. Estimation of serum calcium and phosphorus of the patient was advised, the levels were within normal limits. However not satisfied with the cytological diagnosis, the surgeon attempted incision and drainage of the swelling. Multiple tiny, whitish pieces that aggregated to size 4x3.5 cm were removed. The firm tissue pieces were embedded in paraffin blocks for histological study. Tissue sections showed extensive areas of calcification surrounded by fibrosis and collagenisation [Figure 2c]. The cytological diagnosis of tumoral calcinosis was confirmed on histopathological study.

The second case was of a 75 year old female who had reported with swelling on left foot above heel region of 10 months duration. The swelling was gradually increasing in size with mild pain. There was no other complaint or significant present or past history. Her General and systemic examination revealed no abnormality. Routine haematological investigations, liver and kidney function tests were normal. The 3x2 cm swelling on dorsum of foot near left lateral malleolus was firm and non-tender with restricted mobility and appeared calcified [Figure 1b]. Local radiograph revealed calcified soft tissue swelling without bone involvement.

FNAC carried out with a wide bore needle yielded white, chalky material. Wet and dry smears stained with regular stains showed amorphous, basophilic material and no cells. Cytological and histopathological findings were similar to those of first case. Cytological diagnosis of tumoral calcinosis was confirmed on histopathological examination of the excised swelling [Figure 2 and b] and normal serum calcium and phosphorus levels.

**DISCUSSION**

Calcinosis cutis is the deposition of calcium salts within the skin. It is divided into dystrophic, metastatic, idiopathic and iatrogenic types. Tumoral calcinosis (TC) is a variant of idiopathic calcinosis cutis. TC was first described in 1899 and the term tumoral calcinosis was coined by Inclan in 1943. It is characterised by formation of a single or multiple tumor like periarticular deposits of calcium that most frequently occur around hip, shoulder and elbow joints. Rare sites of occurrence are hands, feet, knees, neck, spine and temporomandibular joints.

Tumoral calcinosis manifests as slowly growing firm, irregular, painless subcutaneous masses in periarticular regions on extensor surfaces. TC commonly occurs in healthy children and young adults and is rare in persons older than 50 years. It shows equal sex predilection. The disorder has genetic basis and is inherited as an autosomal dominant trait with variable clinical expression. This distinct clinical and histopathological entity of TC shows geographical preponderance and larger series are reported from African countries where it is known as hip stone. There are no demonstrable abnormalities of calcium metabolism in Idiopathic TC.

TC may present with multiple lesions and some are bilateral and symmetric. Usually the subcutaneous swellings are large when noticed and may reach up to 20 cm diameter. TC usually manifests as slowly growing, irregular, painless subcutaneous masses located in the vicinity of large joints. The lesion is firmly attached to underlying facia, tendon or muscle but unrelated to the bone. TC may cause complications due to pressure on surrounding structures.

The calcium deposits are visualised radiologically. Radiograph may show lobulated radio opaque lesions of calcific density that surround the soft tissues of a large joint. The underlying joint is usually normal. MRI may show calcific foci infiltrating the surrounding muscles. Occurrence of TC is thought to be related to mechanical trauma or injury that triggers the calcifying process in genetically susceptible individuals. This assumption holds true when TC lesions occur at pressure points in persons belonging to low socio-economic condition and is
attributed to sleeping on ground or hard surfaces.\(^8\) However, the TC lesions of the cases described here were not exactly located at pressure points.

Subcutaneous calcium deposition can occur in clinical setting of primary normophosphatemia, primary hyperphosphatemia and secondary TC.\(^9\) Patients with idiopathic sporadic TC i.e. primary normophosphatemic type show normal Serum calcium and phosphorus levels. Hyperphosphatemia and elevated serum dihydroxy vitamin D level may occur with primary hyperphosphatemic type.\(^5\) Secondary TC is often seen in primary hyperparathyroidism, end stage kidney disease, hypervitaminosis D, myositis ossificans, milk alkali syndrome, sarcoidosis.\(^2\) Age at presentation in both cases described here was 65 and 75 years respectively that is a rare age of occurrence for idiopathic tumoral calcinosis. Both patients belonged to primary normophosphatemic type, revealed normal serum calcium and phosphorus levels and did not have any systemic abnormality.

On FNAC, yield of chalky white aspirate is a peculiar finding and should arouse the suspicion of a calcific lesion. Cytology smears reveal presence of abundant basophilic, amorphous granular material. Mononuclear histiocytic reaction may be seen. Calcium deposits may elicit inflammatory response, due to release of local or systemic pro inflammatory cytokines in patients undergoing haemodialysis.\(^10\) Typical locations of the lesion, its consistency, radiological findings, nature of aspirate are to be taken into consideration while interpreting the cytological findings.\(^11\)

On gross examination, the excised mass is irregular, firm and gritty to cut. Cut surface reveals chalky white areas of calcification. Biopsy of the tumorous mass on histological examination show normal Serum calcium and phosphorus levels. Hyperphosphatemia and elevated serum dihydroxy vitamin D level may occur with primary hyperphosphatemic type.\(^5\) Secondary TC is often seen in primary hyperparathyroidism, end stage kidney disease, hypervitaminosis D, myositis ossificans, milk alkali syndrome, sarcoidosis.\(^2\) Age at presentation in both cases described here was 65 and 75 years respectively that is a rare age of occurrence for idiopathic tumoral calcinosis. Both patients belonged to primary normophosphatemic type, revealed normal serum calcium and phosphorus levels and did not have any systemic abnormality.

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On gross examination, the excised mass is irregular, firm and gritty to cut. Cut surface reveals chalky white areas of calcification. Biopsy of the tumorous mass on histological examination show deeply basophilic granular material in cystic cavities surrounded by dense fibrous and collagenised tissue. Early lesions may show histiocytic and foreign body giant cell reaction.\(^11\) Presence of calcium in tissues can be confirmed by the refractile crystals within the calcified deposit. When TC occurs in older age group and at unusual sites, the possibility of the tumorous swelling being myxoma, skin adnexal tumour, soft tissue neoplasm or subcutaneous metastasis has to be ruled out before excision or debulking of the swelling is undertaken. Diagnosis of TC is rarely made on cytology or core biopsy.\(^12\) However the cytological diagnosis can help to decide about future investigations that will give the subtype of TC. Subtyping of TC is related to the response to treatment and the prognosis.

**CONCLUSION**

Tumoral Calcinosis is a morphological diagnosis and FNAC can proves to be very useful modality for the diagnosis. If the cytopathologist is aware of this uncommon but distinct entity of TC, the diagnosis becomes straight forward.

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