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Case report

Nonfamilial hyperphosphatemic tumoral calcinosis with ulnar neuropathy

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Abstract

We present a case of multiple large juxta-articular painless masses involving both the elbows and right hip in a 27-year old south Asian male who presented with ulnar neuropathy and constitutional symptoms. Radiology, blood investigations and biopsy confirmed it to be hyperphosphatemic tumoral calcinosis. Patient was also diagnosed with an extremely rare association, testicular microlithiasis. Complete surgical excision with low phosphate diet resulted in complete neurological recovery and no recurrence at 30 months. Tumoral calcinosis should be considered in the differential diagnosis of a case with multiple, symptomatic juxta-articular masses.
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Keywords: Tumoral calcinosis; Hyperphosphatemia; Ulnar neuropathy; Testicular microlithiasis

1. Introduction

Idiopathic tumoral calcinosis is a rare benign clinical condition of unknown etiology presenting as periarticular soft tissue calcifications. Symptoms and complications due to this condition are rarely seen. There are many conditions which present with a similar clinicoradiological picture like secondary hyperparathyroidism in chronic renal failure, myositis ossificans, tophaceous gout, calcific myonecrosis and extraosseous osteosarcoma [1,2]. Magnetic resonance imaging (MRI) and histopathology help to differentiate this condition. The typical laboratory parameters include hyperphosphatemia with normal serum calcium, alkaline phosphatase and PTH levels. Treatment involves total surgical excision despite reports of recurrence mainly due to incomplete removal [2].

This report describes a rare case of a 27-year old male with tumoral calcinosis affecting the hip and both elbows with ulnar neuropathy and the extremely rare association of testicular microlithiasis and elaborates the clinicoradiological features and management of this condition.

2. Case report

A 27-year old male presented in June 2005 with insidious onset, gradually enlarging painless swellings over few months, first noticed around the right hip followed by both the elbows. Numbness in his right medial 2 fingers appeared 3 weeks before examination. He also gave history of weight loss and anorexia with low-grade intermittent fever for the past few months. He did not recall any significant recent trauma in the area. Physical examination revealed nontender, bosselated, hard to soft swelling with free overlying skin and dilated surface veins (Fig. 1). Right elbow: 10×12 cm on the posterior aspect, fixed flexion deformity (FFD) of 45° with further flexion upto 90°, grip weakness in the right hand with 50% sensory loss on the ulnar side of forearm and medial 2 fingers; Left elbow: 8×10 cm on the posterior aspect, FFD of 30° and full further flexion; Right hip: 25×25 cm in the right gluteal area, FFD of 10° with full further flexion and restricted rotations. Plain radiographs showed amorphous, cystic, multilobular periarticular calcifications with no erosion of underlying bones (Fig. 2). MRI revealed multiple hypointense foci of calcification with interspersed cystic areas with fluid levels within the mass and infiltration into the surrounding muscles (Fig. 3). Ultrasonography of abdomen and pelvis revealed microlithiasis in both testes. Technetium bone scan showed

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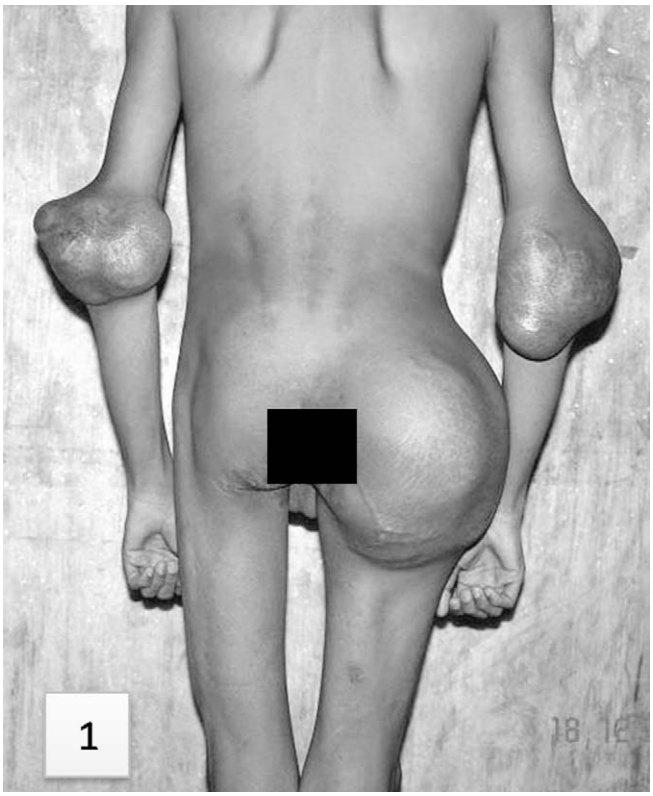

Fig. 1. Preoperative clinical photograph of patient showing bilateral elbow and hip masses.

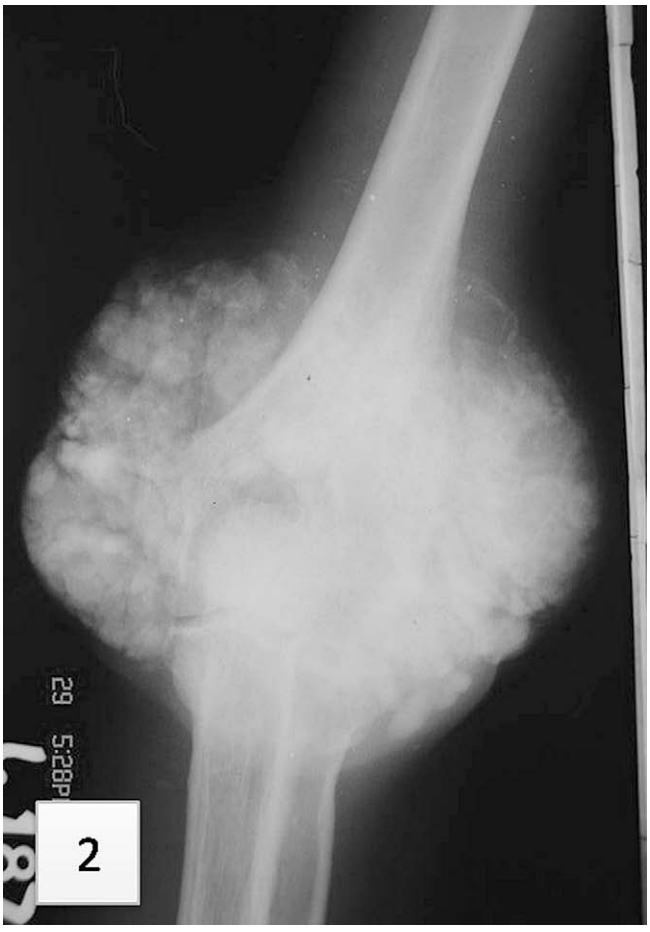

Fig. 2. Preoperative plain radiograph of elbow showing amorphous lobular appearance of mass.

increased uptake in all three masses. Blood investigations revealed hyperphosphatemia (6.4 mg %) with normal serum calcium, alkaline phosphatase, PTH and 1,25 (OH) Vit D levels. Urinary calcium and phosphorus were within normal limits. An open biopsy was performed from the hip mass confirming the diagnosis of tumoral calcinosis. Microscopy revealed fibrocollagenous tissue with extensive calcification, foreign body type giant cell reaction, xanthomatous cells and mononuclear infiltration.

Complete excision of all three masses was subsequently performed. Only the left elbow mass showed an intact capsule and could be removed en masse whereas the others showed extensions into the surrounding muscles and were removed piecemeal. The right ulnar nerve was found fully encased at the elbow and was carefully isolated. The cut surface of the mass was firm to gritty with many cystic areas containing milky fluid.

The patient had gradual complete recovery of ulnar neuropathy over 6 weeks postoperatively and regained his joint motion fully. The patient was put on low phosphate diet which was continued throughout the follow up period. No signs of local recurrence were observed 30 months post-operatively and the patient was asymptomatic with normal appetite and body weight.

3. Discussion

Inclan et al. in 1943 first described a condition of periarticular soft tissue calcification associated with hyperphosphatemia with normal serum calcium levels and coined the term tumoral calcinosis for it [2]. Tumoral calcinosis (TC) is broadly classified into 2 types – (i) Primary not associated with any systemic disease and (ii) Secondary associated with some underlying condition like chronic renal failure, hyperparathyroidism, hypervitaminosis D, sarcoidosis and milk alkali syndrome [3]. Smack et al. [4] further subdivided the primary type into normophosphatemic and hyperphosphatemic types. In the normophosphatemic type, serum concentrations of calcium and phosphate are normal whereas in the hyperphosphatemic type, the serum calcium concentration is normal but the phosphate concentration is slightly high. Hyperphosphatemia may be caused by a common intrinsic proximal tubular abnormality, leading to increased phosphate reabsorption via a pathway independent of PTH. It is associated with an inherited defect in the feedback mechanism regulating 1,25-dihydroxyvitamin D synthesis. This is shown by normal levels of 1,25-dihydroxyvitamin D in the presence of hyperphosphatemia and normal PTH levels and explains the normal calcium levels [5,6]. Approximately 30% of primary type are familial with an autosomal recessive inheritance [3,7]. Familial tumoral calcinosis has been associated with inactivating mutations in *GALNT3* gene resulting in decrease in 0-glycosylation of proteins that control circulating phosphate levels [8]. Recent studies have indicated

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