Hyperphosphatemic Tumoral Calcinosis: A Classical Clinico-Radio-Scintigraphic Presentation

Abstract

Tumoral calcinosis is a rare entity presenting with periarticular calcium deposits, leading to multiple swellings and biochemical hyperphosphatemia and normocalcemia. Skeletal scintigraphy in these cases is helpful by providing a whole-body survey and delineating the common multifocality of this entity. We present the case of a 16-year-old boy with multiple swellings around the knee and elbow joints, having developed over 4 years and diagnosed as tumor calcinosis.

Keywords: *Bone scan, hyperparathyroidism, methylene diphosphonate, metabolic bone disease*

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Interesting Image

A 16-year-old boy presented with gradually progressive swellings developing around the knees and right elbow over the past 4 years [Figure 1a and b]. Those over the knees had spontaneously ruptured, extruding chalky-white pasty discharge. In addition, the swellings were causing mechanical hindrance in the knee movements. Investigations revealed normal renal function, normocalcemia, hyperphosphatemia (serum phosphate 8.8 mg/dl), and increased renal tubular reabsorption of phosphate (96%). His fibroblast growth factor-23 (FGF-23) was inappropriately normal for the degree of hyperphosphatemia. Planar radiograph showed patchy areas of heterotopic calcification along the medial aspect of the thighs and the knee joints [Figure 1c and d]. ^{99m}Tc-methylene diphosphonate (^{99m}Tc-MDP) planar bone scintigraphy followed by hybrid single-photon emission computed tomography with computed tomography (SPECT/CT) showed areas of increased osteoblastic activity in multiple lesions around the right elbow, bilateral knees, and along the medial aspect of both the thighs [Figure 1e-g]. He was diagnosed with having primary hyperphosphatemic tumoral calcinosis. He was placed on a low phosphate diet and started on oral acetazolamide and sevelamer as a phosphate-binding drug. At follow-up, his serum phosphate has come down to 6.4 mg/dl and the lesions have not further increased in size. The surgical excision of the calcified masses surrounding the knees is being contemplated.

Hyperphosphatemic tumoral calcinosis presents as a single or multiple painless swellings in the peri-articular region. The biochemical profile is characterized by normal serum calcium and elevated levels of serum phosphate.[1] Plain radiographs of the affected region show cystic lesions with amorphous and multi-lobulated calcifications.[2] It is regarded as an idiopathic entity, in contrast to dystrophic calcification (inflammatory process with normal serum calcium and phosphate) and metastatic calcification (involving visceral organs, with hypercalcemia).[1] Genetic studies have shown the presence of mutations involving *GALNT3*, *FGF-23,* and α-*KLOTHO* genes leading to dysregulation of phosphate metabolism.[1]

Skeletal scintigraphy with ^{99m}Tc-MDP has a vital role in multiple oncologic and nononcologic, metabolic bone diseases.[3-5] In a patient with tumoral calcinosis, skeletal scintigraphy provides for a complete skeletal survey, thus delineating all the disease sites in a single study. This becomes important, especially as surgical excision of the lesions is curative. Increased uptake of ^{99m}Tc-MDP at the involved sites is due to increased vascularity and osteoblastic activity in the extra-skeletal tissues.[6,7] In

How to cite this article: Pal R, Parihar AS, Bhadada SK, Mittal BR. Hyperphosphatemic tumoral calcinosis: A classical clinico-radio-scintigraphic presentation. Indian J Nucl Med 2021;36:76-7.

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Received: 29-04-2020
Accepted: 22-05-2020
Published: 04-03-2021

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_86_20

Quick Response Code:

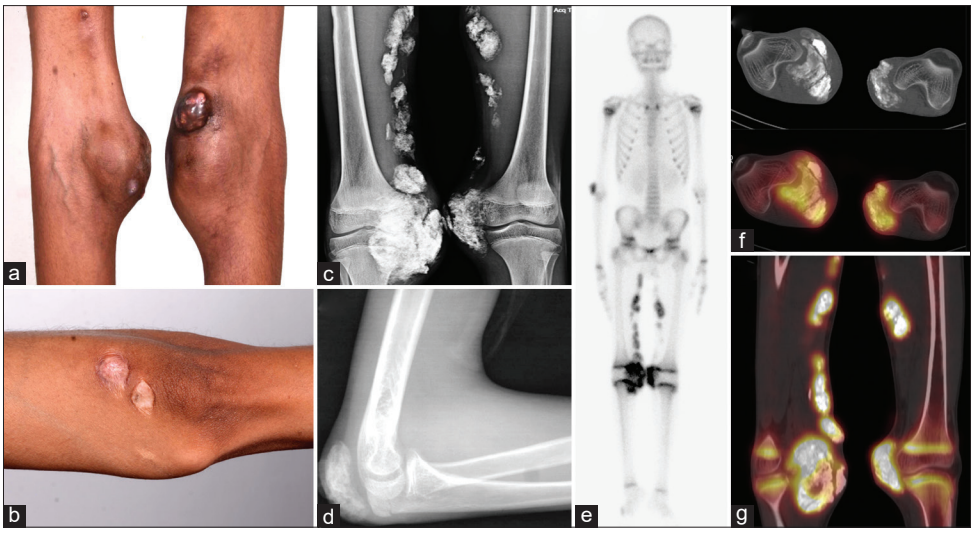

Figure 1: Clinical pictures of the patient showing multiple peri-articular discharging swellings in the bilateral knee (a) and elbow (b) joints. Planar radiographs in antero-posterior (c) and lateral (d) views showing patchy heterotopic calcification along medial aspect of bilateral thighs and knee joints. ^{99m}Tc-methylene diphosphonate whole-body planar (e) and single photon emission computed tomography with computed tomography (f and g) images showing increased tracer uptake in the areas of heterotopic ossification, corresponding to clinical and radiologic images

addition to planar scintigraphy, SPECT/CT can improve the lesion visualization, by eliminating overlap from surrounding structures and can also provide valuable anatomic correlation. Positron emission tomography with 18F-fluorodeoxyglucose and CT has also shown to be positive in patients with tumoral calcinosis, likely reflecting sites undergoing active mineralization, and thus an increased glucose metabolism.[8] In addition, the association of tumoral calcinosis with hyperparathyroidism makes imaging with ^{99m}Tc-MIBI useful, as it can detect the metabolically active sites as well as the presence of concurrent brown tumors.[9,10]

Tumoral calcinosis is a rare entity that needs accurate diagnosis, eliminating the closely mimicking differentials (such as calcinosis cutis and myositis ossificans) and adequate workup toward the disease extent. Scintigraphic studies play a vital role in this respect and thereby help plan surgical excision as a form of curative treatment.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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