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

Abstract
Tumoral calcinosis is a rare entity presenting with periartricular 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

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]. ⁹⁹mTc-methylene diphosphonate (⁹⁹mTc-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).[³]

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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 ⁹⁹mTc-MDP has a vital role in multiple oncologic and nononcologic, metabolic bone diseases.[¹-⁵] 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 ⁹⁹mTc-MDP at the involved sites is due to increased vascularity and osteoblastic activity in the extra-skeletal tissues.[⁶,⁷] In

Address for correspondence:
Prof. Sanjay Kumar Bhadada, Department of Endocrinology, Post Graduate Institute of Medical Education and Research, Chandigarh - 160 012, India.
E-mail: bhadadask@rediffmail.com

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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 99mTc-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.

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Conflicts of interest
There are no conflicts of interest.

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