Tumoralcalcinosis Cutis : A rare case report

Lavanya P[1], Bhagyashree Supekar[2], Vaishali Wankhade[3], Pritica Debnath[4], Rajesh P Singh[5]

ABSTRACT

Tumoralcalcinosis cutis is a rare clinical and histopathologic condition, characterized by calcium salt deposition in different peri-articular soft tissue region. It usually manifests in childhood or adolescence as painless, firm, tumour-like masses around the joints that may lead to joint function limitations specially when large in size. We report a case of tumoral calcinosis cutis in a 9 year old female child.

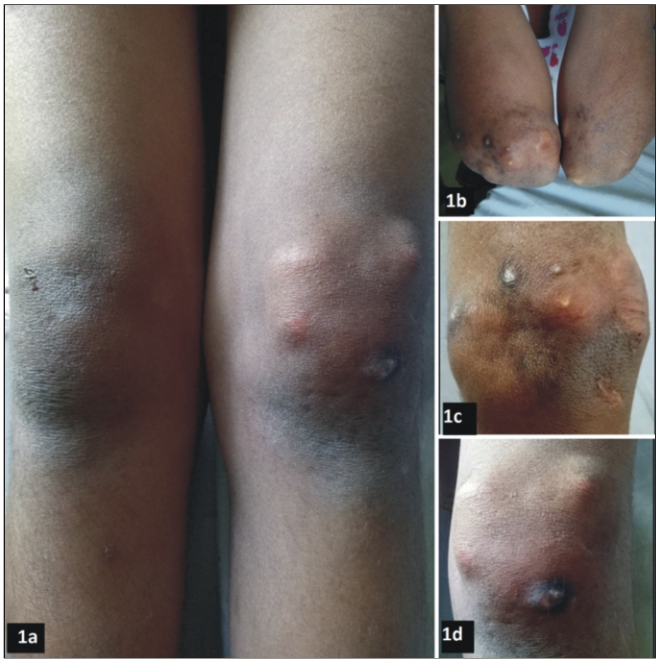

Fig. 1 a-d : **Multiple firm to hard ill defined nodules over extensor aspect of right elbow (a), left elbow (b), dorsum of left hand (c) right knee(d) and left knee**

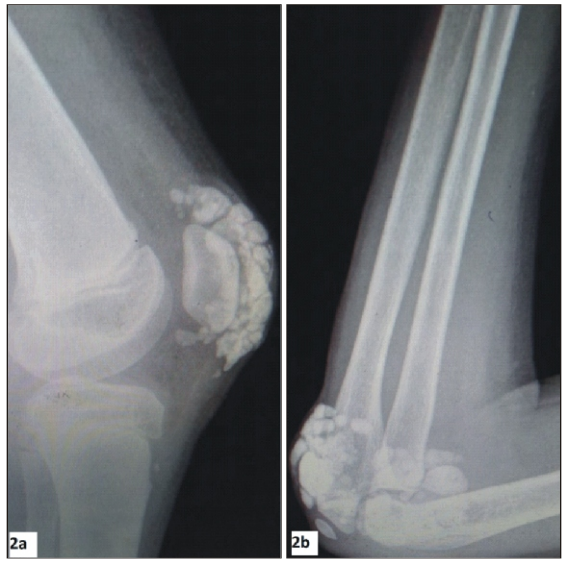

Fig. 2 a-b : **Local x-ray of right knee joint (a) and left elbow (b) and with asymmetrical, amorphous, lobulated calcific deposits in periarticular region.**

Introduction :

Calcinosis cutis is a rare disorder caused by an abnormal deposition of calcium phosphate in various parts of body.[1] Cutaneous calcification is divided into 4 main classes as dystrophic, metastatic, tumoral (idiopathic) and iatrogenic.[2] Tumoralcalcinosis cutis is a subtype of idiopathic

[1]Junior Resident, Senior Resident, Associate Professor, Assistant Professor, Professor and Head of the Department, Department of Dermatology, Govt. Medical College, Nagpur

Address for Correspondence -
Dr. Bhagyashree B. Supekar
E-mail : bhagyashreesupekar.23@gmail.com

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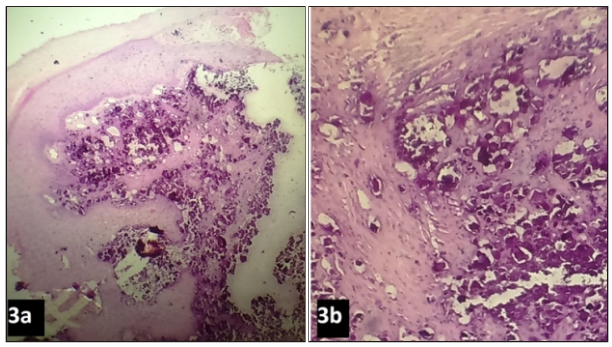

Fig. 3 a-b : **Histopathological examination from the lesion over knee revealed unremarkable epidermis and deeply basophilic amorphous granular material of varying sizes consistent with calcium deposits surrounded by dense fibrous tissue, suggestive of calcinosis cutis. (H&E 4x,10x)**

calcinosis cutis characterized by calcium deposits in subcutaneous or intramuscular tissues around major joints like hips, shoulders, elbows and knees.[2]

Case Report :

A 9-year-old female child, born out of non-consanguinous marriage, presented to dermatology department with multiple asymptomatic raised whitish lesions initially over both elbows and gradually progressed to involve both knee joints over a period of 1 year. There was no history of excessive milk intake or complaints suggestive of endocrine abnormalities and connective tissue disorders. There was no history of similar complaints in the family.

On examination, there were multiple, firm to hard, irregular nodules with ill defined margins over extensor aspect of bilateral elbows, dorsum of hands and knee joints with overlying normal skin. ***(Figure 1a-e)*** Routine hematological tests, liver function tests, kidney function tests and urine routine were normal. Specific investigations including serum calcium (8.3 mg/dl), serum phosphate (4.8 mg/dl), vitamin D3 (23.3 ng/ml), parathyroid hormones, alkaline phosphatase and LDH were in normal range. ANA by immunofloroscence was negative. Local x-ray of left elbow and right knee showed asymmetrical, amorphous, lobulated calcific deposits in periarticular deposits suggestive of calcinosis cutis. ***(Figure 2a, 2b)*** Histopathological examination from the lesion over knee revealed deeply basophilic amorphous granular material of varying sizes consistent with calcium deposits surrounded by dense fibrous tissue, suggestive of calcinosiscutis. ***(Figure 3a, 3b)*** Special stain for fat was not done due to lack of resources. Based on clinico-histopathological and radiological findings a diagnosis of tumoralcalcinosis cutis was reached.

Discussion :

Calcinosis cutis is a group of disorders characterized by abnormal deposition of calcium salts in the skin.[3] Virchow initially described the condition calcinosis cutis in 1855.[1] Datta *et al*[3] first reported a case of idiopathic calcinosis cutis in a 50 year old female with involvement around knee joint and Lanka *et al*[4] first reported case of idiopathic calcinosis cutis in children. Tumoral calcinosis can be widespread or localized. Pathogenesis of idiopathic calcinosis cutis is linked with high levels of Gla, a unique amino acid, which is found in calcified tissue and urine of patient of calcinosis cutis. Gla is normally found in bones and teeth hence ectopic soft tissue calcification can be triggered by production of Gla de novo at tissue sites.[4] Subtypes of idiopathic calcinosis cutis include : 1) Idiopathic calcinosis cutis of scrotum / penis / vulva, 2) Milia like idiopathic calcinosis cutis, 3) Subepidermal calcinosis cutis, 4) Tumoral calcinosis cutis, 5) Calcinosis cutis circumscripta and 6) Calcinosis universalis.[1]

Routine blood investigations, serum calcium level, serum phosphate levels and serum dihydroxy vitamin D levels should be done to rule out metabolic abnormality and local radiographs and histopathological examination for confirmation of diagnosis. Treatment is surgical removal of calcified mass in cases of functional impairment and unacceptable cosmetic appearances but has common recurrences.[5,6] Dietary modifications include restriction of phosphate. Medical therapy though of limited use probenecid, colchicine, magnesium or aluminum antacids, sodium etidronate, diphosphates, diltiazem, warfarin can be tried.

We present this case due to lack of published data of this uncommon variant of calcinosis cutis in pediatric age group.

References :

1. S. K. Jatana, Brig, V. Negi, Col and S. Das. A case of idiopathic calcinosis cutis. Med J Armed Forces India. 2012;68(4): 383-385.
2. Janet A. Fairley. Cutaneous Mineralization and Ossification. In: Wolff Klauss, Goldsmith L.A., Katz S.I., editors. Fitzpatrick's Dermatology in General medicine. 8th ed. 2012. p.1650
3. Datta C, Bandyopadhyay D, Bhattacharyya S, Ghosh S. Tumoral cacinosis. Indian J Dermatol VenereolLeprol2005;71:293-4.
4. Lanka P, Lanka LR, Ethirajan N, Krishnaswamy B, Manohar U. Idiopathic calcinosis cutis. Indian J Dermatol 2009; 54: 388-9
5. DenielaKroshinsky and Janet A. Fairley. Calcifying and ossifying disorders of the skin. In: Jean L. Bolognia, Julie V. Schaffer, Lorenzo Cerroni, editors. Dermatology. 4th ed. 2017. p. 790.
6. Andrew Morris. Inherited metabolic diseases. In: Christopher Griffiths, Jonathan Barker, Tanya Bleiker, Robert Chalmers, Daniel Creamer. Rook's textbook of dermatology. 9th ed. 2016. p. 81.19.