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Two females with multiple swellings in hand: Idiopathic calcinosis cutis

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Abstract
Calcinosis cutis is very rare disorder characterized by the deposition of calcium in the skin and subcutaneous tissue. Idiopathic calcinosis cutis has only rarely been reported in the literature. Here, we report cases of two healthy females 40 year old and 55 year old who presented with multiple asymptomatic hard nodules on the fingertips of their hands. Histopathological, radiological, and extensive blood investigations confirmed the diagnosis of idiopathic calcinosis cutis.

Keywords: Two females, idiopathic calcinosis cutis, idiopathic

Introduction
Calcinosis cutis is a condition where there is an abnormal deposition of calcium salt specifically calcium phosphate in the skin. Pathophysiology of this abnormal condition was first described by Virchow in 1855. There are five subtypes of calcinosis cutis depending on etiologies,

1. Dystrophic
2. Idiopathic
3. Metastatic
4. Iatrogenic
5. Calciphylaxis [1].

Idiopathic calcinosis cutis being the most uncommon of all, associated with normal calcium metabolism and without tissue injury. Idiopathic calcification of scrotum, subepidermal calcified nodule are the various forms of idiopathic calcinosis cutis [2, 4]. Our report highlights a 40 year old female suffering from a rare case of idiopathic calcinosis cut is presenting with yellowish white swelling involving multiple fingertips.

Case Presentation
A 40 year old female with no significant past medical history presented with chief complaints of multiple whitish yellow swelling over fingertips for the last two months. Her family history and social history was not significant. On Clinical examination she was found to have multiple, firm to hard whitish yellow looking papulo-nodular swellings over the pulp of the thumb, index, middle and little finger of left hand (Figure 1,2). On considering signs and symptoms and correlating clinically, differential diagnosis of gouty tophi and calcinosis cutis came forward. On further evaluation, her radiograph of bilateral hand revealed multiple, heterogeneous calcified soft tissue in finger pads suggestive of calcinosis cutis (Figure 4). We started further investigating and all her routine blood investigations were within normal limits which included liver function tests, renal function tests, serum electrolytes, C-reactive protein, and erythrocyte sedimentation rate. Serum calcium, phosphate, uric acid, vitamin D3, thyroid function and parathyroid hormone levels were also within normal limits. The workup for connective tissue disorders can't be done because of affordability issue of the patient. Our patient agreed for skin biopsy. Histopathology report helped us in making diagnosis (Figure 5, 6). After ruling out metabolic causes, autoimmune disorders and malignancy;

We made the final diagnosis of idiopathic calcinosis cutis.
Another patient 55 year old female with similar complaints of swelling over tip of finger (Figure 3).
In this patient sr. calcium was <8.0 ng/ml rest all investigation were normal. Only on the basis of proper history, clinical examination, basic hematological investigations and X-ray (Figure 5). We could conclude the diagnosis as calcinosis cutis. Now we are planning to start Diltiazem in one patient and steroid or warfarin in second patient to compare its efficacy.

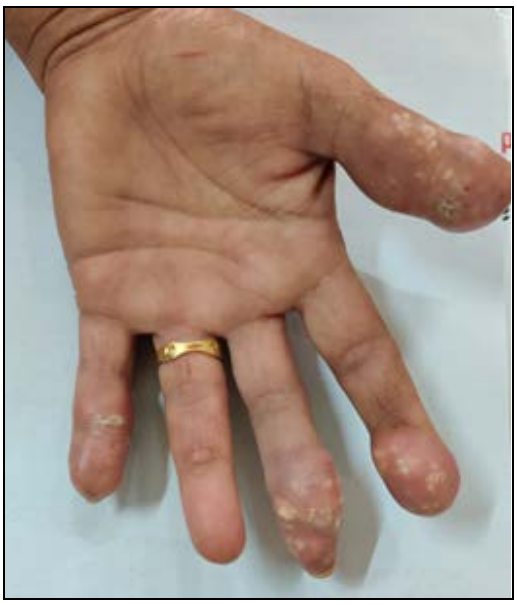

Fig 1: Clinical picture of case 1

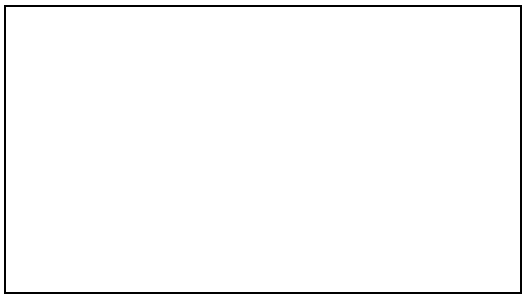

Fig 2: Whitish yellow papulo-nodular swelling on fingertip

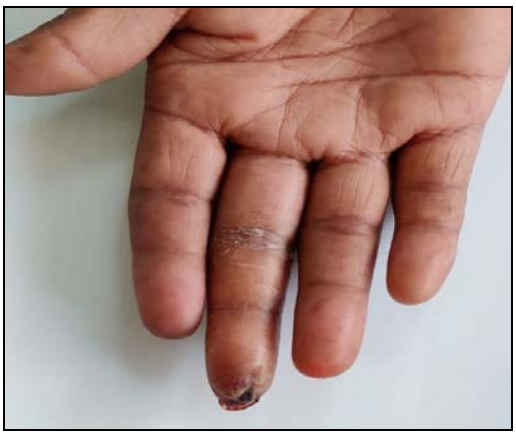

Fig 3: Clinical picture of case 2

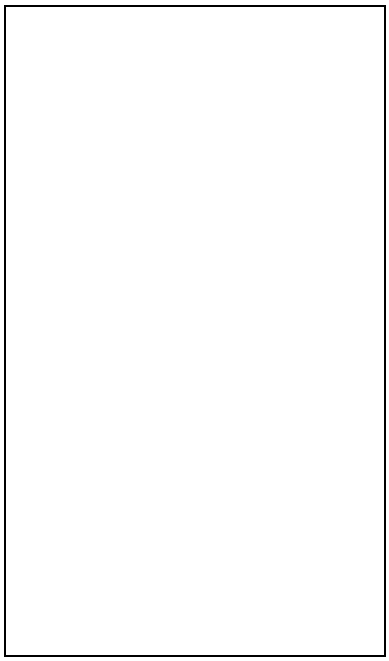

Fig 4: Xray PA view of fingers showing calcification over fingertips

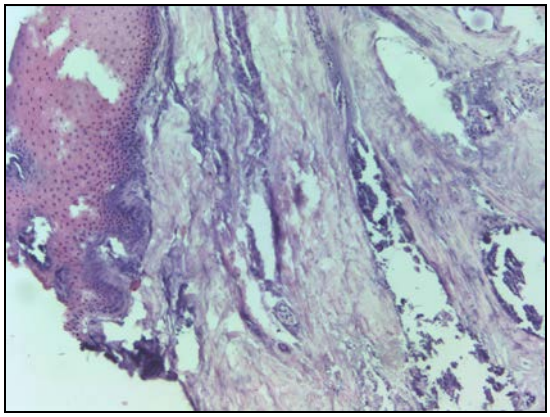

Fig 5: Histopathological slide photograph of the soft tissue obtained from patient

Discussion

Calcinosis cutis is characterized by deposition of insoluble calcium salts in the subcutaneous and cutaneous tissue.
Calcinosis may result from a variety of causes such as: Trauma to the region, Inflammation (bug bites, acne), Infections, Tumors (malignant or benign), Diseases of connective tissue, Varicose veins, Hypercalcemia, Hyperphosphatemia Calcinosis cutis is associated with systemic sclerosis.
Data on incidence of Calcinosis Cutis is not available on records. It is more common in blacks and middle aged with No sex differentiation [5].
Dystrophic calcification is the most common type of calcinosis cutis and associated with underlying tissue damage. Dystrophic calcification is found in many connective tissue disorders which include scleroderma, dermatomyositis, and systemic lupus erythematosus, mixed connective tissue disorder. Dystrophic calcification can be seen rarely with Werner syndrome, Ehler-Danlos syndrome, panniculitis, basal cell carcinoma and cysticercosis.

Metastatic calcification occurs in patients with abnormal calcium/phosphate metabolism (chronic kidney disease, hyperparathyroidism, milk-alkali syndrome, sarcoidosis and malignant neoplasm).

Calciphylaxis, iatrogenic and idiopathic are other presentation of calcinosis cutis. Calciphylaxis is a form of calcific vasculopathy usually associated with patients of end-stage renal disease, which involves small and medium-size vessels present in dermis.

Incidence of Idiopathic calcinosis cutis is very rare [6, 8]. It occurs without any tissue damage or abnormal calcium or phosphate metabolism. Idiopathic calcinosis has 3 subtypes, namely; scrotal calcinosis, familial tumoral calcinosis and subepidermal calcified nodules. The underlying pathophysiology behind this abnormal deposition of calcium salts in skin is unclear. Hypothesis says that abnormal metabolism of gamma carboxy glutamic acid (GIa) is responsible for abnormal calcium deposition in subcutaneous tissues, with increased production of GIa is attributed to soft tissue calcification [9]. A mutation in the gamma-glutamyl carboxylase gene is also reported to cause aberrant calcification in dermal fibroblast [10].

Treatment of calcinosis cutis is challenging. The limited role of warfarin, diltiazem, bisphosphonates, probenecid, colchicine, aluminium and magnesium antacids has been described in the literature. There are many views on using diltiazem in idiopathic calcinosis cutis with some report showing significant resolution with long-term therapy [11, 12]. It is believed that, Diltiazem, being a calcium channel blocker inhibits the calcium accumulation into the cells. The indications for surgical removal are ulceration, infection, pain and functional impairment. Our patient had none of these complications, so we decided to observe the patient with close follow-up.

Conclusions

In conclusion, our report shows the rare presentation of calcinosis cutis. Extensive evaluation to rule out potential abnormalities of calcium and phosphate metabolism, connective tissue disorders, renal dysfunction, and malignancy should be done. Though the role of medical management is limited in idiopathic calcinosis cutis, the possibility of correctable/secondary causes should be sought in patients presented with abnormal soft tissue calcification. Some reports showed promising results with diltiazem therapy. So we are planning to start it. Further study will be required to find best treatment of calcinosis cutis.

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