Calcinosis Cutis of the Thigh: A Surgeon's Perspective and Recent Advances

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Abstract

Calcinosis cutis is a clinical condition in which calcium salts are deposited in the skin and subcutaneous tissue. It is classified into four major types- dystrophic, metastatic, iatrogenic and idiopathic. Clinically, non-specific skin nodules to cutaneous swellings are seen. Diagnosis of calcinosis cutis is difficult alone and the need for ancillary techniques likes histological examination, laboratory techniques to determine metabolic abnormalities and radiological examination including plain X-ray, CT scanning to demonstrate the extent of tissue calcification. We report a 72 year old female who presented with a painless swelling over the right thigh since 2 years. Biopsy revealed calcinosis cutis. Radiological imaging was suggestive of a lesion confined to the skin. We report a case of calcinosis cutis of the thigh with relevant discussion on the same. Calcinosis cutis of the thigh is a rare clinical entity with only a handful of cases reported till date. In this case report, we discuss about calcinosis cutis and throw light on the various types, clinical presentation, diagnosis and management of the same.

Keywords: Calcinosis cutis; Cutaneous swellings; Thigh; Treatment

Introduction

Calcinosis cutis is a disorder wherein calcium salts are accumulated in the dermis and subcutaneous [1]. It is divided into five kinds: dystropic, metastatic, idiopathic, iatrogenic and calciphylaxis [2]. Dystrophic calcification, where Calcium levels are normal in the serum, is the most common cause. Tissue damage usually due to an underlying condition *viz.* systemic sclerosis, dermatomyositis, mixed connective tissue disease or lupus produce a nidus for calcification. Metastatic calcification has abnormal blood levels of calcium and phosphorus with deposition occurs [3]. Tumoral calcinosis, subepidermal calcified nodules and scrotal calcinosis are examples of Idiopathic calcification where there is no underlying tissue injury or aberrant laboratory findings. Iatrogenic calcification is produced by injection of calcium or phosphorus containing compound leading to precipitation of calcium salts [2,3]. Calcification related to chronic renal failure and dialysis, mainly of small and medium sized vessels is known as Calciphylaxis. Calcinosis cutis is categorised into:

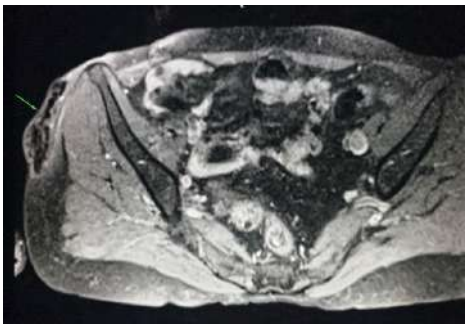
a. Calcinosis circumscripta: Limited to an extremity or joint [4,5].
b. Calcinosis universalis: Diffuse involvement of subcutaneous tissue and fibrous structures of muscles and tendons [6].

Case Report

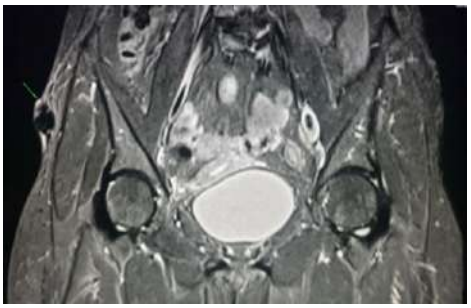
A 72 year old female presented with a history of a painless swelling over the right thigh for the last 2 years which had progressed in size over the last 6 months. There was no family history of similar lesions present. On examination, a single irregular swelling with lobulations fixed to skin, measuring 8 cm × 5cm, over upper outer aspect of right

thigh (Figure 1). Soft tissue ultrasound of right thigh revealed a hyperechoic lesion confined to the skin of right thigh and MRI was advised for further correlation. MRI thigh revealed subcutaneous calcification, revealing features consistent with calcinosis cutis (Figure 2). Biopsy was suggestive of calcinosis cutis. Dermatological opinion was taken and lesion diagnosed as calcinosis cutis. A surgical excision under local anaesthesia was done (Figure 3). Patient was followed up for 1 year with no recurrence.

Figure 1: Calcinosis cutis of the thigh.



2 (a)



2(b)

Figure 2a, 2b: MRI showing a lesion confined to the skin and subcutaneous tissue with dense calcification suggestive of Calcinosis cutis.

Figure 2: Postoperative picture.

Discussion

Etiology

Caused by trauma, inflammation, varicose veins, tumors, infections, connective tissue disorders, hyperphosphatemia and hypercalcemia. It is associated with systemic sclerosis.

Epidemiology

It is seen in mainly in patients suffering from systemic sclerosis. 25% to 40% of patients with limited systemic sclerosis. Seen in 30% of adults and 70% of children with dermatomyositis. Patients with SLE can present with periarticular calcification in 33% of the cases.

Pathophysiology

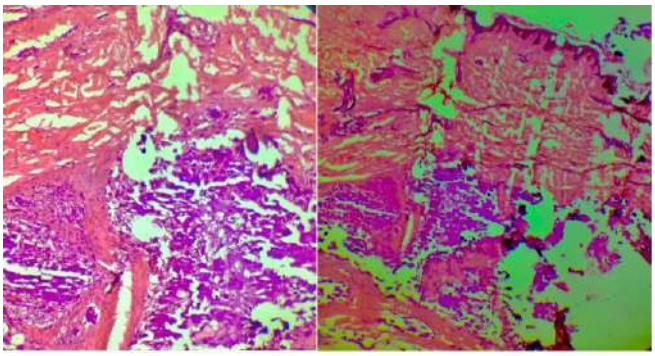
Dystrophic Calcification: It is the most prevalent form of calcinosis cutis. Serum calcium and phosphorus values are normal [2]. The proteins in the dying cells when tissue is damaged are bound by phosphate which is further bound by phosphate binding proteins and these complexes undergo calcification. Linked to systemic sclerosis, dermatomyositis and systemic lupus erythematosus [3]. It has also been observed in Rheumatoid arthritis, Scleroderma and Sjogren's syndrome [7]. Dystrophic calcification is seen in elastic pseudoxanthoma, Werner's syndrome, Ehler's Danlos syndrome, Pilomatrixoma, trichilemmal cyst, Basal cell carcimona, trichoepithelioma, pancreatic and lupus panniculits, onchocerciasis, cysticercosis, histoplasmosis, cryptococcosis, trauma and burns [6].

Metastatic Calcification: Abnormal serum calcium and phosphorus levels leads to deposition of calcium salts in the tissue (Figure 4). Mainly seen after calcium phosphate

exceeds 70 mg/dl. These aberrant levels precipitate the calcium salts and resolve with correction of calcium and phosphate levels. [8] These deposits are frequently situated in the periauricular area [2]. The most common cause of metastatic calcification is chronic renal failure. Other causes are - Hypervitaminosis D, Hyperparathyroidism, Sarcoidosis, Milk-alkali syndrome and malignant neoplasms. Milk-alkali syndrome arises with excessive ingestion of meals or antacids that include calcium [2,8].

Idiopathic calcification: Deposition of calcium salts without underlying tissue damage or abnormal calcium or phosphorus levels [6]. Three types are seen:

a. Familial tumoral calcinosis
b. Subepidermal calcifies nodules
c. Scrotal calcinosis



4 (a) 4(b)

Figure 4: Microscopy shows deposition of calcium salts and occasional mononuclear cells suggestive of calcinosis cutis.

Management

Though the treatment for calcinosis cutis is challenging, Steps can be taken to help facilitate treatment and increase blood flow to the extremities including avoiding trauma, smoking cessation, decreasing stress and exposure to cold [6]. Smaller lesions have been reported to respond to Diltiazem, Bisphosphonates, Probenecid, Aluminum hydroxide [4]. Patients with small and localized lesions are good candidates for surgical treatment whereas more generalized disease will require medical management [6,7]. Drugs used commonly are:

a. **Diltiazem:** Most commonly used. It acts by decreasing the amount of calcium that enters the cell and macrophages of damaged tissues. High dose of 2 mg/kg/day to 4 mg/kg/day is required for therapeutic response [2].
b. **Warfarin:** Vitamin K levels have been measured to be high in some patients with calcinosis cutis. Use of warfarin 1mg/day normalizes the levels and has some improvement for small lesions [2,8].
c. **Bisphosphonates:** Acts on macrophages which are active at the affected sites, thereby preventing the release of pro inflammatory cytokines. It also reduces calcium turnover and reabsorption. Shows good response mainly in dermatomyositis and systemic sclerosis [5,9].

   Etidronate: 800 mg/day.

   Alendronate: 70 mg/week.

   Pamidronate: 90 mg/week.

   Adverse effects include osteonecrosis of jaw, fever, infusion site reactions etc [4].
d. **Ceftriaxone:** Affects matrix metalloproteinases, chelates calcium and anti-inflammatory. Given in a dose of 2 mg/day [6,7].
e. **Aluminium Hydroxide:** Binds to phosphorus and reduces intestinal absorption of phosphorus. Given in dose of 2.24 g/day, 2.4 g/day and 1.8 g/day for calcinosis cutis, dermatomyosis and lupus respectively [3].

Conclusion

Calcinosis cutis is a rare clinical entity .To the best of our knowledge only a handful of cases are reported till date. This case report has been reported to shed some light on the management from a surgeon's perspective.

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