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Case Report *Volume 5 Issue 1*

Idiopathic Calcinosis Cutis Universalis: A Rare Case Presentation

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Abstract

Calcinosis cutis is a rare disorder marked by the gradual accumulation of hydroxyapatite crystals in the skin across different body regions. The condition is categorized into three types based on its causes: dystrophic, when there's tissue damage with normal calcium and phosphorus levels; metastatic, associated with hypercalcemia or hyperphosphatemia and idiopathic, when there's no tissue damage and normal calcium and phosphorus levels; While medical treatments have limited effectiveness, surgical excision has proven beneficial, offering symptomatic relief, however is out of question in Calcinosis Cutis Universalis. We encountered a rare case of Idiopathic calcinosis Cutis Universalis in a healthy 54-year-old female, devoid of connective tissue disorders or abnormal mineral metabolism. On treatment with Diltiazem it was noticed that the improvement of existing lesion was slow, but therapy was effective in preventing new lesions. This report aims to enhance medical awareness regarding the presentation, etiopathogenesis, and progression of the relatively uncommon Idiopathic Calcinosis Cutis Universalis.

Keywords: Calcinosis Cutis Universalis; Idiopathic; Diltiazem

Introduction

Calcinosis Cutis Universalis: This rare and perplexing dermatological disorder poses unique challenges to both clinicians and researchers, as its origins, manifestations, and optimal management strategies continue to elude our complete understanding [1].

Calcinosis cutis, characterized by the deposition of insoluble calcium salts within the skin and its appendages, presents itself in various forms. Among these, the Calcinosis Cutis Universalis stands out as a rare subset, where calcified deposits manifest extensively across the body [2]. However, there are instances where the underlying cause remains unidentified despite extensive work-up and termed Idiopathic Calcinosis Cutis Universalis. The diagnostic process for calcinosis cutis involves a thorough examination of the patient's history, looking for potential trauma or repeated exposure to calcium supplements. Additionally, a comprehensive assessment of signs and symptoms related to collagen vascular disorders is crucial during both history-taking and physical examination. Diagnostic tools such as X-ray or sonography of the extremities and trunk, aimed at identifying dense shadows indicative of calcium deposits, are essential. Biopsy plays a pivotal role in confirming the diagnosis [3]. In this report, we present a case of calcinosis

of a 54 year old female who displayed widespread calcific deposits, and notably, the etiology was found to be idiopathic.

Case Report

A 54-year-old female, presented with complaints of multiple subcutaneous nodules and plaques which had started about 3 years ago on her hip and subsequently spread progressively to her thighs, legs and arms. There was no family history of similar lesions. At the time of presentation, the patient was otherwise healthy and had no complaints of joint pain, skin rashes, Raynaud's phenomenon, dysphagia or dyspnea. She was on Anti-hypertensive since 10years and had no known drug allergies.

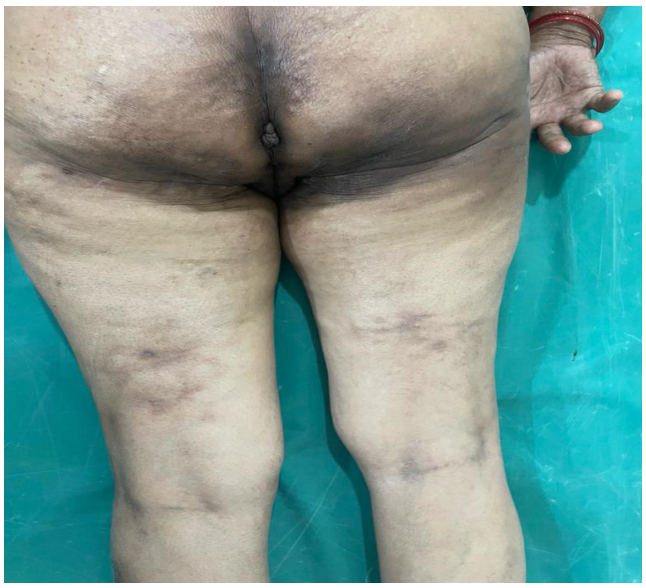

Figure 1: Indurated plaques in thigh region.

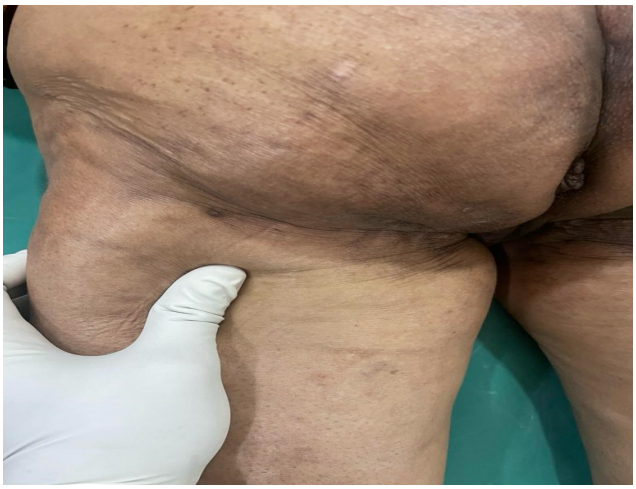

Figure 2: Indurated plaques in buttock region.

Physical examination revealed multiple hard indurated nodules and plaques of size varying from 1x1 cm to 10x7 cm over anterior and lateral aspect of both thighs, buttocks, posterior aspect of calf, axilla and arms (Figures 1 & 2). General examination and Systemic examination including neurological examination were normal.

Laboratory investigations revealed serum calcium (8.5mg/dl, normal: 8.5-10.3 mg/dl), phosphorus (4.2 mg/dl, normal: 3.0-6.0 mg/dl) as well as serum 25-hydroxy vitamin D (20 ng/dl, normal: 17-54 ng/dl), parathormone (PTH) (35 pg/dl, normal: 11-54 pg/dl). Biochemical examinations gave normal results for complete hemogram, erythrocyte sedimentation rate, blood sugar, uric acid, electrolyte, liver function and kidney function tests. Routine urine examination and 24-hour urinary calcium (122.6mg/24h, normal: 100-300mg) mg/dl)was within normal range. ANA profile and Myositis profile done and was found to be normal.

Radiological examination showed that there was no bone pathology and no joint involvement, but widespread extensive amorphous calcifications in the soft tissues. Ultrasonography showed that the calcifications were located in fat tissue under the skin, but not in deeper tissues or muscles. PET CT and Carotid Doppler revealed no significant anomalies. Histological examination of one of the nodules revealed massive calcium deposits as evident by Von kossa staining (Figure 3)

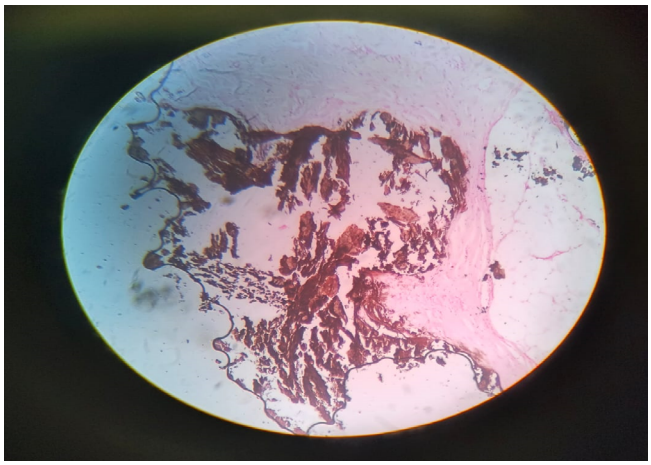

Figure 3: Histopathological image showing well-circumscribed calcium deposits stained black by Von kossa stain in dermis and the subcutaneous tissue.

At the time of presentation, patient was not on any treatment. Despite seeking input from multiple consultations for her concerns, the patient faced the challenge of misdiagnoses, initially being labelled with cellulitis and abscess. As a result, she underwent treatment that included antibiotics and

incision and drainage procedures.

A diagnosis of Idiopathic Calcinosis Cutis Universalis was made after thorough evaluation to rule out other causes of Calcinosis. Patient was initiated on Diltiazem 90mg/ day increased, under close cardiologic monitoring, to 180 mg/ day. It was observed that after 4 month period there was noticeable clinical improvement noted by reduction in stiffness and pain. Repeat skeletal survey also shows mild clearance of the calcification (Figures 4 & 5).

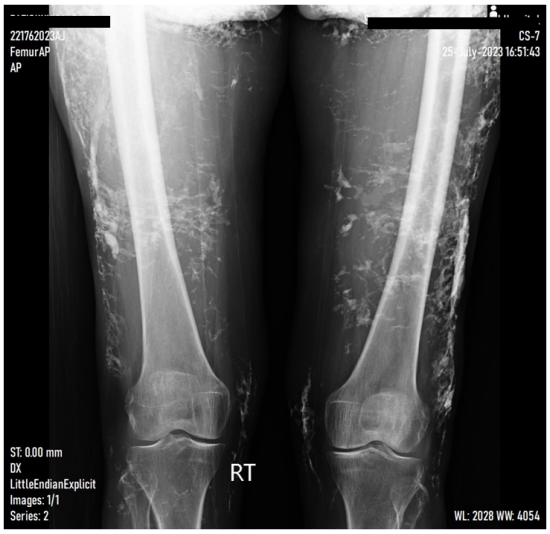

Figure 4: Radiograph of pelvis showing widespread extensive amorphous calcifications in the soft tissues in pelvis and both thighs (calcinosis cutis universalis).

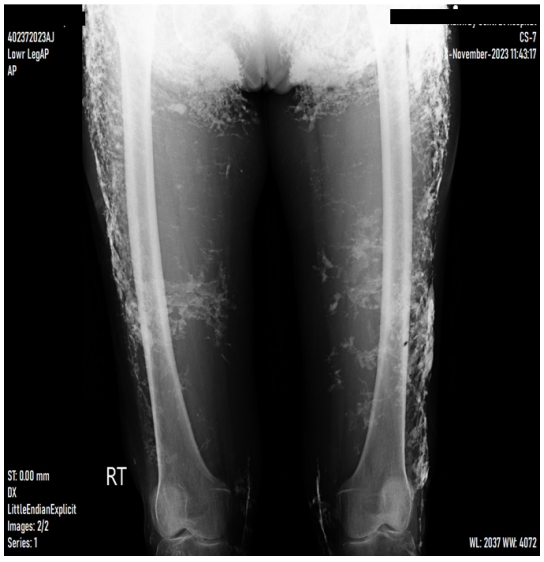

Figure 5: Radiograph of pelvis showing clearance of calcium deposits after 16 weeks of treatment with oral diltiazem.

Discussion

Calcinosis cutis is an uncommon condition, presenting as metastatic, dystrophic, or idiopathic types. Metastatic calcinosis results from spontaneous calcium and phosphate deposition in normal tissues due to elevated serum calcium and/or phosphate levels. Various factors, such as calcium supplementations, primary hyperparathyroidism, hypervitaminosis D, renal failure, and others, contribute to increased serum calcium in this type [4]. Dystrophic calcinosis is more prevalent and is linked to underlying diseases but with normal serum calcium and phosphate levels. Idiopathic calcinosis, a rare form, occurs without known causes or collagen vascular diseases [5].

The precise pathogenesis remains unknown and likely involves multiple factors. Idiopathic calcinosis cutis is particularly challenging to understand, occurring in apparently normal tissue without known causes. The ultra-structural morphology of localized skin calcifications in idiopathic cases with normal serum calcium and phosphate levels is not entirely clear [6].

Treatment approaches vary, with drugs such as bisphosphonates, corticosteroids, and dietary modifications having varying degrees of success. Surgical excision is considered effective in idiopathic calcinosis cutis but is reserved as a last resort due to potential risks [7].

The presented case involves a 54yr old female with Idiopathic Calcinosis Cutis Universalis, exhibiting widespread calcified deposits. Despite a lack of identified causative factors, the patient experienced noticeable improvement with Oral Diltiazem.

Due to its inhibitory effect on cell membrane calcium channels, Diltiazem has been proposed as a potential treatment for calcinosis. In an initial trial using an animal model of Duchenne's muscular dystrophy, a condition characterized by elevated muscle calcium content, Diltiazem demonstrated a reduction in calcium levels and an improvement in muscle function. The suggested mechanism of action involves the prevention of new calcium deposition, although this alone does not explain the additional re-absorption of existing calcium [8].

The process of re-absorption may be attributed to the scavenging actions of macrophages, which become more effective at clearing existing calcium deposits once the accumulation of new calcium has been halted. In summary, Diltiazem's potential therapeutic effects on calcinosis may not only involve preventing further calcium deposition but also facilitating the re-absorption of existing calcium through the enhanced activity of macrophages [1].

The management of Calcinosis Cutis poses a significant challenge, with no established consensus on the optimal treatment. Despite attempts to address the condition using substances like Aluminium Hydroxide and Diltiazem, as well as exploring alternative options such as Probenecid, Warfarin, Colchicine, and Bisphosphonates, the outcomes have been contentious. Surgical intervention to remove symptomatic lesions is typically considered a last resort due to concerns that local trauma might potentially trigger further calcification, leading to recurrence or worsening of the condition [6].

Source of Support

Nil.

Conflict of Interest

None declared.

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