

## Extensive Idiopathic Calcinosis in a Child – A Diagnostic and Therapeutic Imbroglia

### Abstract

Idiopathic calcinosis is a disorder characterized by diffuse calcium deposits at various sites of the body. Etiopathogenic associations are described with inherited disorders, connective tissue disorders, infections, tumors, trauma, and endocrine disturbances. No diagnostic tests or standard therapeutic guidelines are established for this entity. There is paucity of literature on idiopathic calcinosis. We describe a girl child with extensive calcinosis in the skin and around muscle bundles without any clinical and laboratory evidence for etiological associations. Aggressive treatment modalities resulted a notable improvement in lesions in index child. Growing evidence will help to establish the ground for understanding and developing standard therapy.

**Keywords:** *Calcinosis cutis, connective tissue disorders, pamidronate, rheumatological diseases*

### Introduction

Idiopathic calcinosis (IC) is a disorder of unclear etiology characterized by diffuse calcinotic deposits at various sites of the body. The etiology of IC remains a mystery, and it stands as a diagnostic and therapeutic challenge for clinicians.<sup>[1]</sup> There are few ongoing trials with different therapeutic modalities like sodium thiosulphate for treatment.<sup>[2]</sup> Growing evidence contribute to establishing the ground for better understanding and standard therapeutic options. We describe a girl child with IC with extensive calcinotic lesions on the skin as well as underlying tissues with encouraging treatment results.

### Case History

A 7-year-old girl born of non-consanguineous marriage had progressively enlarging large nodular oozing whitish calcinotic lesions at multiple sites for the past three years leading to occasional pain and limitation in joint movements. Past, family, nutritional, and immunization history were non-contributory. There was no history of trauma, recent infections, or constitutional symptoms. On examination, large calcinotic lesions were present around bilateral elbows, knees, abdomen, neck, axilla, and back ranging from 1 × 1 cm

to 5 × 5 cm in size [Figure 1]. It resulted in difficulty in flexion at the elbows and knees. There was no evidence of alopecia, Gottron's papules, heliotrope rash, shawl sign, or photosensitivity. Gag reflex, neck muscles, and proximal muscles were normal. She did not have gouty tophi, varicosities, osseous hyperplasia, warts, milia, sclerosis, or xanthomas.

Multiple hemograms showed normal counts and smear microscopy except for mild anemia (hemoglobin of 96 g/L). Erythrocyte sedimentation rate (30 mm in the first hour) and C-reactive protein (6 mg/L) were normal. Serum uric acid, calcium, and phosphorus were 3.4 mg/dl, 9.4 mg/dl, and 4.4 mg/dl, respectively. Liver, kidney, and thyroid function tests were normal. Levels of angiotensin-converting enzyme, parathormone, vitamin D, creatine phosphokinase, lactate dehydrogenase, lipase, aldolase, lipid profile, and complements were normal. Infectious diseases workup was non-contributory. Antinuclear antibodies, anti-double-stranded deoxyribonucleic acid (DNA) antibodies, Mantoux test, and karyotyping for trisomy were negative. Antibodies against extractable nuclear antigen and myositis-specific and myositis-associated antibody panels were negative. Nail-fold capillaroscopy (NFC) showed mild

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tortuosity, ectasia, and hemorrhages in the fingers. X-ray revealed multiple diffuse subcutaneous and deep tissue calcinosis [Figure 2]. There were no features of Ehler-Danlos syndrome, Werner syndrome, or Rothmund-Thomson syndrome. Biopsy from lesions showed predominant calcifications and mild inflammation (with plasma cells, histiocytes, and neutrophils) without evidence of panniculitis, granuloma, or malignancy. Opinions from international experts on calcinosis were also taken. Teutschlaender disease was unlikely because there was non-specific distribution, early age of presentation and progression, absence of family history, and no calcium/phosphate disturbances. A diagnosis of idiopathic calcinosis was offered after ruling out all other possibilities like inflammation, tumors, infections, connective tissue disorders (including juvenile dermatomyositis), gout, and xanthomas.

Since excision was not possible, she was commenced on intravenous immunoglobulin (IVIg - 1 gm/kg once), parenteral pamidronate (1 mg/kg/day for three days every three months), intravenous ceftriaxone (100 mg/kg/day for 20 days), and topical sodium thiosulphate application daily.<sup>[3]</sup> Various centers have tried different modalities for this distressing condition.<sup>[3]</sup> There were no adverse effects attributable to pamidronate or IVIg. Though there was no definite evidence of connective tissue disorders like dermatomyositis, we also added weekly subcutaneous methotrexate (15 mg/m<sup>2</sup>/week) for mild changes in NFC and calcification around muscle bundles.<sup>[4]</sup> Intravenous sodium thiosulphate could not be given due to unavailability and logistic issues. At follow-up after seven months, lesions had improved [Figure 3].

IC is a poorly understood disorder with calcium deposits in the skin, subcutaneous tissues, and various other sites.<sup>[1]</sup> In general, calcinosis cutis is categorized as dystrophic, metastatic, iatrogenic, calciphylaxis, and idiopathic.<sup>[5]</sup> In addition to the localized form, IC sometimes has a widespread distribution in sites other than skin and subcutaneous tissues. Though etiological associations are described with a wide range of disorders like trauma, connective tissue diseases (e.g., dermatomyositis, lupus, and scleroderma), panniculitis, infections, tumors, hyperparathyroidism, and inherited disorders (e.g., Ehler-Danlos syndrome, Werner syndrome, Rothmund-Thomson syndrome), investigations may not reveal any particular underlying cause.

No particular treatment modality is proven uniformly effective in IC.<sup>[6]</sup> Parenteral Pamidronate, IVIg, sodium thiosulphate, and excision are tried successfully in various centers for management.<sup>[2,4,7]</sup> There are reports of limited success in treating calcinosis with sodium thiosulphate either topical, intradermal, intralesional, or intravenous.<sup>[2,3,8]</sup>

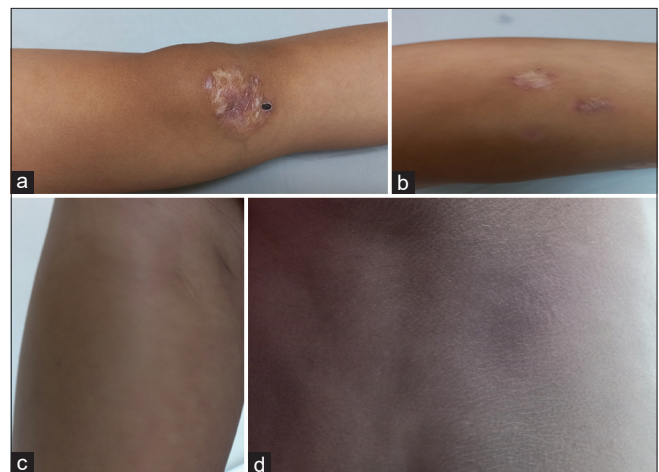
There is a paucity of literature and reports on IC from south Asian countries. Tawekji *et al.* reported diffuse idiopathic calcinosis cutis in a Syrian boy.<sup>[9]</sup> There are reports of idiopathic tumoral calcinosis in children also.<sup>[10]</sup>



**Figure 1:** Calcinosis lesions in left knee (a), right knee (b), axilla (c), abdomen (d), and back (e)



**Figure 2:** Radiographs showing diffuse calcinosis lesions over the axilla (a), abdomen (anteroposterior view) (b), iliac fossa, and back (lateral view) (c), and at bilateral thighs and knees (d)



**Figure 3:** Healed site of lesions with a scar in knee (a and b), inguinal region (c), and back (d)

Our case is noteworthy and unique as the lesions were not limited around the large joints but widespread on the skin, around soft tissues, muscle bundles, and deeper tissues. This report also represents the importance of excluding possible differentials and benefits of aggressive treatment modalities for IC. Extensive evaluation and aggressive treatment yield better outcomes for this distressing condition. Multicentric data and prospective studies

will contribute to establishing a consensus guideline for management.

### **Ethics approval**

This study was approved by the Institute Ethics Committee and the Departmental Review Board. All procedures performed in studies were per ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

### **Consent to participate and publish**

Informed written consent was taken from the parents of the patient. Assent was obtained from the child also.

### **Authors' contributions**

DB: Concept of the study, data analysis and interpretation, drafting, editing, critical revision, and final approval

AZB: Design of the study, acquisition of data and data analysis, editing, and final approval.

PKP: Data acquisition and analysis, and editing

UR: Acquisition of data and data analysis

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### **Conflicts of interest**

There are no conflicts of interest.

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