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Case Report

Calcinosis cutis associated with systemic sclerosis: a rare case report

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ABSTRACT

Calcinosis cutis is abnormal calcium deposition in the skin and subcutaneous tissues of the body. It is generally associated with autoimmune connective tissue disorders and in our case, it is systemic sclerosis. It most commonly occurs in the fingers presenting with pain and functional impairment. Here, we present a case of calcinosis cutis with systemic sclerosis in a teenage girl presented with bilateral gluteal pressure sores and multiple sites of calcium deposition like sacrum, upper limbs, knees and the face. We treated here with reconstructive surgery with Limberg flaps for the pressure ulcers with excision and primary closure of the other sites with calcium deposits.

Keywords: Calcinosis, Dystrophic, Systemic sclerosis, Surgery

INTRODUCTION

Calcinosis cutis is the deposition of calcium in the skin and subcutaneous tissues.[1] It is associated with autoimmune connective tissue diseases like systemic sclerosis, dermatomyositis, mixed connective tissue diseases, and more rarely, systemic lupus erythematosus (SLE).[2] Calcinosis in systemic sclerosis presents as subcutaneous nodules of different sizes and shapes, typically at sites of recurrent microtrauma. 25% of patients with systemic sclerosis have calcinosis at some time during their illness, on average more than 7.5 years after the onset of systemic sclerosis.[2,3]

The most frequent areas involved are the hands (palmar side of the terminal phalanges) and feet. Other common sites are the elbows and knees, arms and legs, trunk, and face.[2,4,5] Some studies have related calcinosis in systemic sclerosis with male gender, digital ulcers, digital tip pitting scars, acro-osteolysis, telangiectasias, anti-centromere antibody, and anti-PM/Scl (anti-exosome) antibody.[6-11]

CASE REPORT

A 15 years old girl presented along with her mother to the Department of Plastic and Reconstructive Surgery with bilateral gluteal ulcers for one-year duration in October 2017. She was a known case of juvenile systemic sclerosis on treatment under rheumatology. She had skin tightening, inability to sit cross legged, microstomia, Raynaud's phenomenon with digital ulcers in 2003. She also is a known case of bilateral interstitial lung disease confirmed by high resolution computed tomography of the chest and spirometry and with mitral valve prolapse and trivial mitral regurgitation on echocardiography. Due to worsening of oral ulcers and interstitial lung disease, she was started on mycophenolate mofetil and itopride. On examination, the girl was thin built and poorly nourished. Her vitals were stable. She had grade 3 bilateral gluteal pressure ulcers both measuring 3 x 3 x 2 cm with hard white chalky deposits in the ulcers (Figure 1). Her X-ray pelvis shows fluffy soft tissue calcifications adjacent to the bilateral inferior pubic rami, ischial tuberosities and obturator foramen (Figure 2) She was operated and the gluteal ulcers were debrided,

excised and reconstructed by Limberg flaps. Patient was comfortable in the post-operative period with well settled flaps and discharged on the 10th post-operative day after suture removal. Now, after 3 years, she presented with white chalky deposits emerging at the sacral region, right knee, left chin and nasal sill (Figure 3-6) These were excised and the wounds closed primarily. Now, her elbow joint has bony ankylosis (Figure 7).

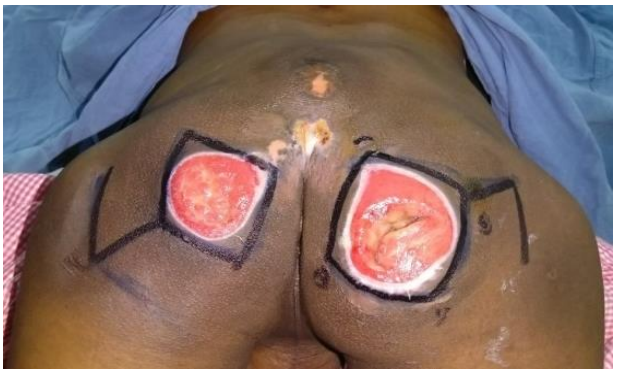

Figure 1: Bilateral gluteal pressure ulcers with Limberg flap markings.

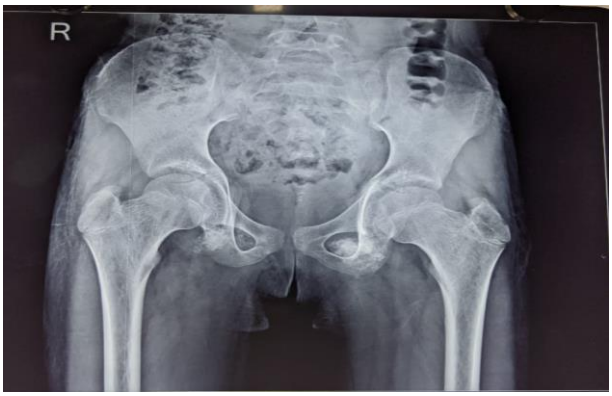

Figure 2: Plain radiograph of pelvis showing fluffy soft tissue calcifications adjacent to the bilateral inferior pubic rami, ischial tuberosities and obturator foramen.

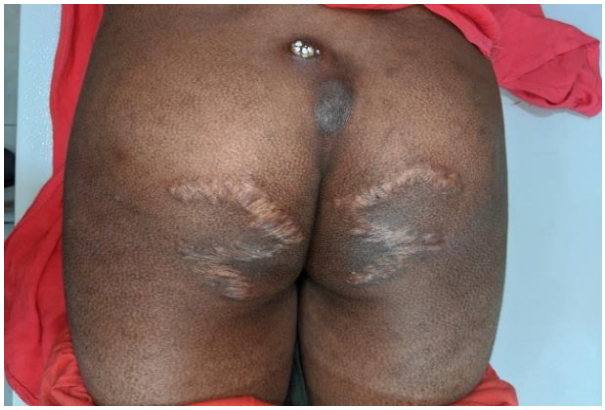

Figure 3: Healed Limberg flaps with white chalky deposits at the sacrum.

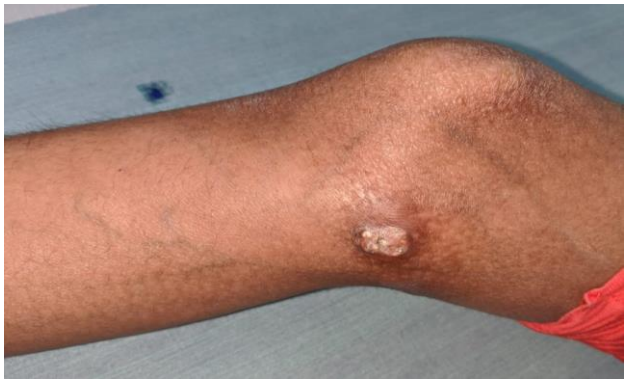

Figure 4: Calcinosis cutis of right knee.

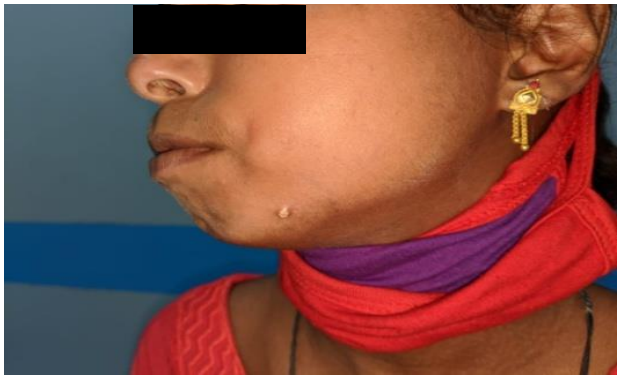

Figure 5: Calcinosis cutis of left chin.

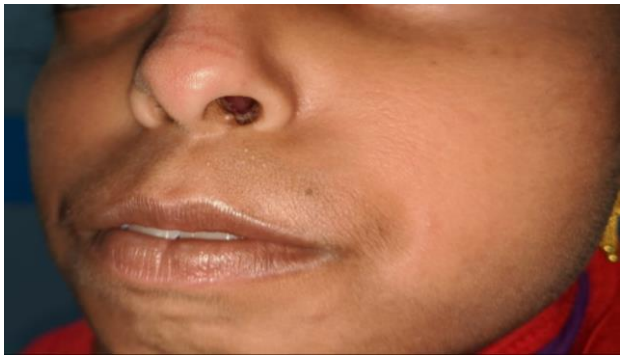

Figure 6: Calcinosis cutis of nasal sill.

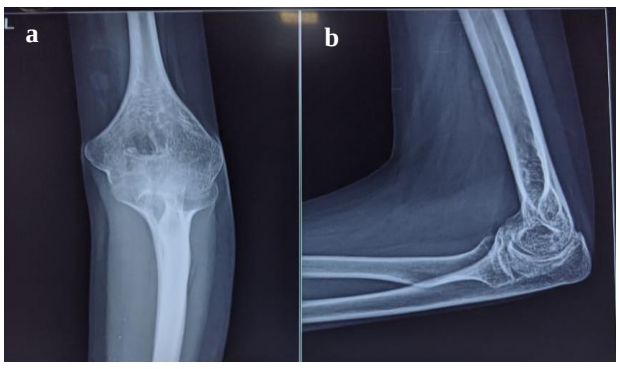

Figure 7 (a and b): Plain radiograph of right elbow joint showing bony ankylosis.

DISCUSSION

Calcinosis cutis is a rare and chronic condition characterized by deposition of insoluble calcium salts in the skin and subcutaneous tissues. There are 5 subtypes of calcinosis cutis namely dystrophic, metastatic, idiopathic, iatrogenic, and calciphylaxis.[12] Dystrophic calcinosis is the most common type of calcinosis cutis and is seen in association with autoimmune connective tissue diseases such as systemic sclerosis, dermatomyositis, lupus erythematosus, and lupus panniculitis. CC is thought to occur as a result of chronic local tissue injury and is a common complication of systemic sclerosis especially the limited form (CREST syndrome: calcinosis, Reynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia), affecting approximately 25% of these patients.[13] It usually presents as subcutaneous nodules in fingers or areas of pressure such as elbows, knees, or ischial tuberosities and might be associated with pain, soft tissue swelling, ulcers with toothpaste-like material discharging or deformities, which may lead to functional problems. [14] Treatment of calcinosis cutis is tough and challenging as there is no gold standard treatment.[15] Sodium thiosulphate is an inorganic salt, which increases calcium solubility and has been reported to be helpful in treating calcinosis.[13,14] The dystrophic type can frequently occur in the setting of scleroderma and CREST syndrome. The pathophysiology of dystrophic calcification may include chronic inflammation, vascular hypoxia, recurrent trauma, and abnormalities in bone matrix proteins.[14] General measures consist of improving blood circulation to the extremities, avoiding stress, cold exposure, and trauma, antibiotics and paracetamol, nonsteroidal anti-inflammatory agents, and opioids for pain relief.[14] The current treatment modalities include warfarin, bisphosphonates, minocycline, calcium channel blockers (diltiazem), ceftriaxone, aluminium hydroxide, probenecid, intravenous immunoglobulin, biologic agents such as infliximab and rituximab, intralesional corticosteroid, extracorporeal shock wave lithotripsy, curettage, surgical excision, and carbon dioxide laser. The type of treatment being used (systemic vs topical or medical vs surgical) depends on the severity and distribution of the lesions.[14,15] Sodium thiosulphate either as intravenous, intralesional, and topical have been tried.[14,16,17] Three mechanisms of action have been proposed for sodium thiosulphate which are increased calcium solubility (through its chelation effect for cations that produces soluble calcium thiosulfate complexes), vasodilatation, and antioxidant effect that restores endothelial cell function.[18] In 2008, Wolf et al reported a case of a leg ulcer with dystrophic calcification that was successfully treated with topical 10% sodium thiosulphate solution.[19] Bair and Fivenson reported 2 cases of ulcerative dystrophic calcinosis refractory to multiple topical treatments that had excellent responses to topical 25% sodium thiosulphate compounded in zinc oxide.[20] Here we report the successful treatment of superficial calcinosis cutis of gluteal region, left chin and nasal sill with surgery in a patient with systemic sclerosis.

CONCLUSION

Calcinosis is a common problem affecting about 25% of patients with systemic sclerosis. It most commonly affects the fingers of hands, and is related to inflammation and hypoxia, and can lead to complications like local inflammation, ulceration, and infection. Although there is no universally effective treatment for calcinosis in patients with SSc, several pharmacological therapies have been proposed as effective, as monotherapy or in combination. Surgical removal of calcinosis remains the mainstay for treatment. Newer methods with using novel outcomes are necessary to determine the efficacy of current and emerging treatments.

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