Idiopathic Linear Calcinosis Cutis - A Case Report

Dr. Jayanti Singh[1], Dr. Vijay Paliwal [2§], Dr. Puneet Bhargava[3], Dr. Deepak Kumar Mathur[4] and Dr. C.M. Kuldeep[5]

[1]Junior Resident, Department of Dermatology, SMS Medical College, Jaipur (Rajasthan) India
[2]Professor Associate Professor, Department of Dermatology, SMS Medical College, Jaipur (Rajasthan) India
[§]Email of Corresponding Author: : dr.vijaypaliwal@gmail.com
[3]Professor, Department of Dermatology, SMS Medical College, Jaipur (Rajasthan) India
[4]Senior Professor, Department of Dermatology, SMS Medical College, Jaipur (Rajasthan) India
[5]Senior Professor, Department of Dermatology, SMS Medical College, Jaipur (Rajasthan) India

Abstract—*Linear calcinosis cutis is a very rare disorder; reported in association with lichen sclerosus atrophicus, scleroderma, after intravenous infusion of calcium and acute monocytic leukemia. In available literature idiopathic Zosteriform Calcinosis Cutis is yet to be described. Eight year old child presented with spontaneous painful, slowly progressive, linear band like nodules over right half of the chest and back of trunk. It is a rare presentation in Skin Department. On the basis of clinical, laboratory investigation and histopathology a diagnosis of Linear Calcinosis Cutis was made. So it was decided to report this rare case of idiopathic Zosteriform Calcinosis Cutis because of unusual presentation and rarity of the disorder.*

Keywords: Linear Calcinosis, Zosteriform Calcinosis Cutis, LSA, Scleroderma.

I. INTRODUCTION

Calcinosis cutis is an uncommon disorder, characterized by tender, hard skin coloured dermal nodules. Four distinct categories described; dystrophic, metastatic, iatrogenic and idiopathic. Morphological variants include nodular, plaque, tumoral and linear; occurring localized or generalized in distribution.[1]

Linear calcinosis cutis is a very rare entity; reported in association with (lichen sclerosus atrphicus) LSA, scleroderma, after intravenous infusion of calcium and acute monocytic leukemia.[2,3,4,5,6]

This zosteriform calcinosis cutis is described in association with LSA. In best of our knowledge idiopathic Zosteriform Calcinosis Cutis is yet to be described. It is a rare presentation in Skin Department. So it was decided to report this rare case of idiopathic Zosteriform Calcinosis Cutis because of unusual presentation and rarity of the disorder.

II. METHODOLOGY

A 8 year old male presented in skin department with spontaneous painful, slowly progressive, linear band like nodules over right half of the chest and back of trunk. It was a rare presentation. Case was properly observed, evaluated, investigated and was found to be idiopathic zosteriform calcinosis cutis, which is a rare disorder. So it was decided to report this rare case of idiopathic zosteriform calcinosis cutis because of unusual presentation and rarity of the disorder.

III. CASE REPORT

Eight years old male child presented in skin OPD with complaint of linear painful nodules over right half of the chest for last 2-3 months. According to his parents these nodules seemed to have started as brownish discolored slightly raised lesions over right half of the chest. Over a period of 2-3 months, these lesions increased in size and progressed linearly from chest to back of trunk up to midline. Some

of these lesions discharged whitish chalk like material. Thereafter these lesions more or less remained unchanged. There was no history of trauma, any kind of injection over the area, muscle weakness, hardening or whitish discoloration of skin and any congenital anomaly over the affected area.

On examination there were multiple brown to skin colored, tender, hard to firm dermal nodules and hyperpigmented and atrophic scars distributed linearly over the right pectoral, infra axillary, adjoining scapular region in the distribution of 4^{th} spinal nerve. The surrounding and intervening skin was normal in color and consistency. Other general physical and systemic examinations were unremarkable.

Routine laboratory investigations including CBC, PBF, and ESR were within normal limits. Mantoux test was measured 3 mm after 72 hrs and considered as negative. Special investigation including ANA, DNA, Anti-Jo 1, Anti LA, Anti Scl 70 and anti-centromere antibody were negative. Both serum and 24 hour urinary calcium and phosphorus were within normal limits. Other biochemical investigations for kidney, liver and parathyroid were within the normal limits

Skiagram of chest was otherwise normal except multiple calcified linearly arranged opacities in the soft tissue of chest wall.

Histopathology revealed multiple dense area of calcification in entire dermis and subcutaneous tissue without any inflammatory reaction. In addition there was marked atrophy of overlying epidermis. The connective tissue and ground substances were unremarkable.

Figure 1:
Multiple linearly arranged brownish colored nodules and atrophic scars over right pectoral, infra axillary and infra scapular area

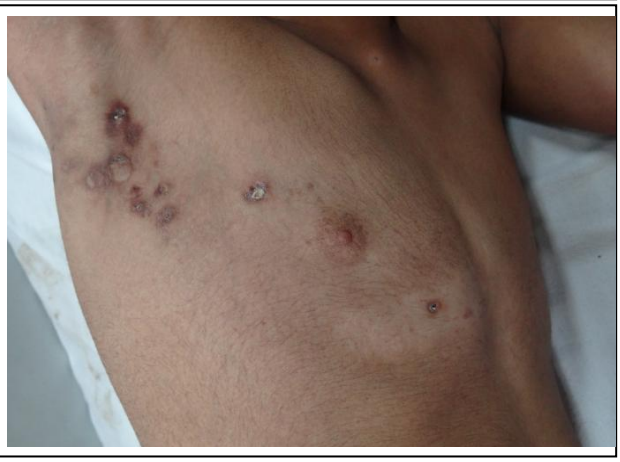
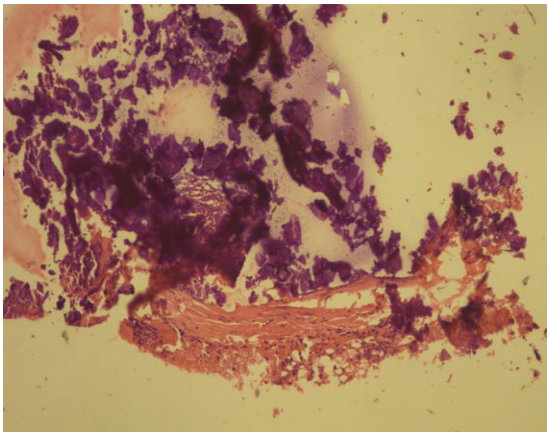


Figure 2:
Diffuse Calcium Deposits in Dermis



On the basis of clinical, laboratory investigation and histopathology a diagnosis of Linear Calcinosis Cutis / Zosteriform Calcinosis Cutis was made.

IV. DISCUSSION

Calcinosis cutis is classified in four categories; dystrophic, metastatic, iatrogenic and idiopathic. The linear calcinosis cutis usually occurs secondary to underlying disorder. Dystrophic linear calcinosis cutis is the commonest; it occur s in previously damaged tissues secondary to limited or diffuse scleroderma, panniculitis, cutaneous neoplasm, trauma and other inflammatory disorders. Linear calcinosis cutis is reported in association with morphea, intravenous calcium gluconate therapy and acute monocytic

leukemia. All above mentioned categories of calcinosis occurs with normal serum and urinary calcium level.

Metastatic calcification is associated with raised serum calcium level; however both serum and urinary calcium levels were normal in our case. A linear pattern of calcium deposition has previously been reported as a result of calcium gluconate infusions. These linear deposits occurred in the vein distal to the infusion site and presented 2 h to 24 days after the calcium infusion.[3,5] This is not true in our case as he never received calcium infusion(s).

Pathophysiology of calcinosis is not much understood, few studies indicate that it occurs secondary to trauma or tissue damage,[2,5] it is not true in this case as he never had any trauma or pathology over the affected area that can lead to tissue damage. The pathophysiology in our case remained an enigma, it could have developed due to some localized changes in the composition of dermal tissues particularly collagen which provided niche to calcium deposition.

In conclusion, zosteriform calcinosis cutis has been described with scleroderma, calcium infusion, acute monocytic leukemia and panniculitis. Idiopathic calcinosis cutis is yet to be described in literature. In best of our knowledge, probably this is the first case of idiopathic linear calcinosis cutis to be reported. The pathophysiology of calcinosis in this case remained an enigma, it requires further studies.

CONCLUSION

This case was idiopathic Zosteriform Calcinosis Cutis which is a rarely reported entity.

CONFLICT

None declared till date.

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