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Case Report

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A CASE OF IDIOPATHIC SCLEREDEMA: A RARE ENTITY

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ABSTRACT

Scleredema is a fibromucinous connective tissue disease characterized by symmetric, nonpitting edema and induration of the face, neck and trunk. Although the pathogenesis remains elusive, associations with infection, diabetes mellitus, and paraproteinemia have been established. We report here a case of scleredema in a 40 years old male patient with progressive thickening and induration of the skin over upper arms, face, anterior and posterior trunk since 6 months. On histopathology, markedly thickened reticular dermis with thickened collagen bundles and abundant mucin in between was found. We are reporting this case in view of its rarity and lack of effective treatment modalities.

KEYWORDS: Scleredema, non pitting, diabetes mellitus, paraproteinemia.

INTRODUCTION

Scleredema was described by Buschke in 1902 and is also designated as scleredema of Buschke and scleredema adultorum, although the latter is a misleading term because children or adolescents can develop the condition.[1] It is a rare disorder of unknown cause in which areas of induration appear in the skin, frequently after an infection but also in diabetes, and spontaneously clear in months or years. There is an excess of acid mucopolysaccharides in the dermis.[2] and thickened collagen bundles.

The condition has also been associated with monoclonal gammopathy.

It is characterized by painless, symmetric, edematous, non pitting induration of the skin of the face, scalp, neck, trunk and upper extremities. Hands and feet are often spared.

Numerous therapies have been tried, with mixed and inconsistent results. No standard treatment for this disease is currently known. Recently, UVA1[3, 4], bath PUVA[5], cream PUVA[6], low-dose broad-band UVA combined with colchicine[7], narrow-band UVB[8], prostaglandin E1[9] and tamoxifen.[10]

CASE REPORT

A 45 years old male patient presented in our outpatient department of Dermatology with gradual thickening of skin over the face, neck, proximal part of upper limbs, trunk and proximal part of lower limbs since 4 months. The patient was born out of a non- consanguineous marriage by a full term, normal vaginal delivery.

There was no history of any itching, preceding upper respiratory tract infection or raynaud's phenomenon, patient did not have any dyspnoea or dysphagia.

General physical and systemic examination did not reveal anything significant.

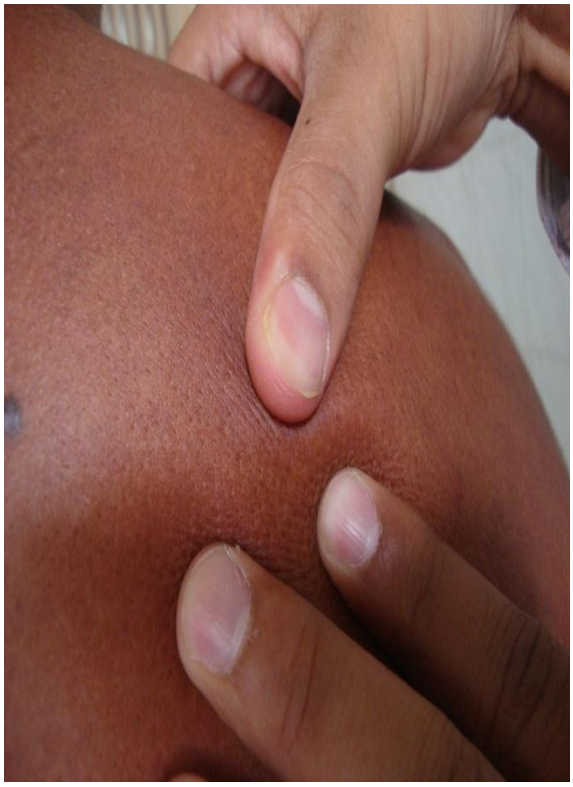
On cutaneous examination, patient had diffuse induration and thickening of the skin over the face, trunk and proximal extremities. There was mild erythema of the overlying skin with patulous follicles. {Figure1, Figure2} There was difficulty in pinching the skin. {figure3}.

Routine blood and urine investigation were normal. Blood sugar levels were normal. ASO titre was not raised.

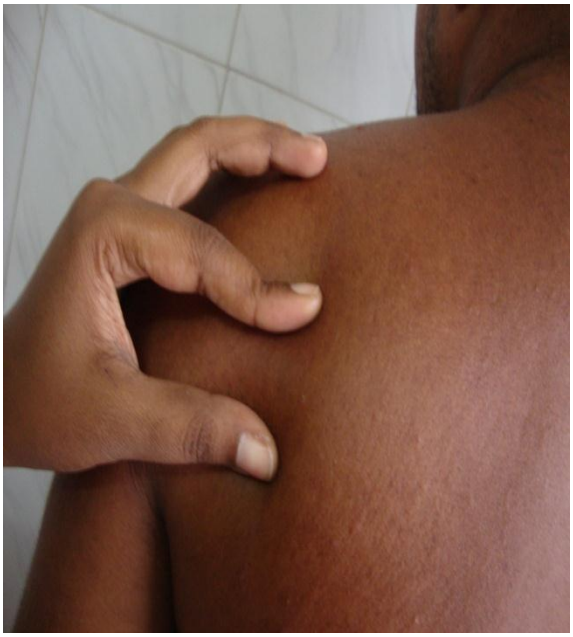
Histopathological examination from a representative site showed marked thickening of the reticular dermis with thickened collagen bundles. Abundant mucin was seen between the collagen bundles, especially in the lower reticular dermis and subcutis. Fibrocytes were not increased. The findings were consistent with Scleredema. {figure4}.

The patient was started with T. Methotrexate 10mg per week. Patient showed only mild clinical improvement in the beginning. The skin induration began to increase after 2 months of therapy. Now, the patient is on colchicine orally.

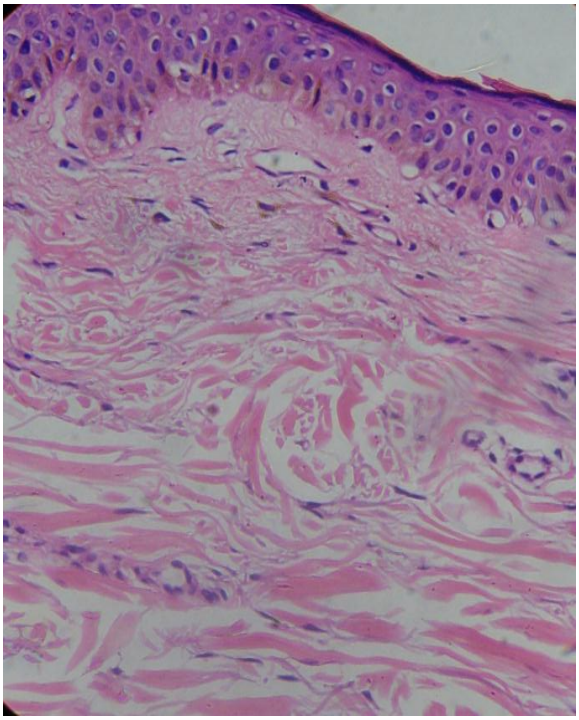
{FIGURE 1}- apparently normal skin, with mild erythema



{FIGURE 2}- patulous, dilated follicles



{FIGURE3}- difficulty in pinching the skin



{FIGURE4}- histopathological examination showing thickened reticular dermis with mucin in between thickened collagen bundles. (40X, H & E)

DISCUSSION

Scleredema is a fibromucinous connective tissue disease characterized by symmetric, nonpitting edema and induration of the face, neck and trunk. Although the pathogenesis remains elusive, associations with infection, diabetes mellitus, and paraproteinemia have been established. Scleredema tends to present in young than aged and in women cases against men. The lesions usually begin on the face and/or neck and spread to the arms, shoulders, back and chest giving a peau d orange appearance.

According to Graff et al., there are three recognized clinical groups of scleredema. In the first group, the disease starts abruptly after an acute upper respiratory tract infection and often resolves in a few months to years, while in the second group no respiratory tract infection is involved and has longer duration over a period of years. The third group consists of long standing scleredema associated with severe complicated diabetes mellitus.

In our case, the patient had non pitting induration of the skin of arms, face, trunk amd thighs. The investigations to rule out possible causes did not reveal any significant findings. But the histological examination revealed thickened collagen bundles and abundant mucin deposition in the lower reticular dermis and subcutis. These findings were consistent with scleredema.

Treatment of scleredema poses a challenge as a definitive modality of treatment is yet to be found. Patients have been treated with various drugs and phototherapy with variable outcomes in different studies.

Some known treatments include UVA1[1, 2], bath PUVA[3], cream PUVA[4], low-dose broad-band UVA combined with colchicine[5], narrow-band UVB[6], prostaglandin E1[7] and tamoxifen[8] and methotrexate.

A definitive treatment regimen is yet to be ascertained as evidence regarding the efficacy of these treatment modalities has not yet been found.

We hereby report this case where no definite cause could be elucidated. Further investigations are necessary to find out the exact cause of the disease in this patient.

Conflict of interest: none.

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