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Case Report

Scleromyxedema without internal malignancy – A case report and review

Shekhat PK*[1], Mahajan R[2], Pandya IA[1], Bilimoria FE[3]

[1]Resident, [2]Associate Professor, [3]Professor & HOD; Department of Skin & VD, Smt. B. K. Shah Medical Institute and Research Center, Sumandeep Vidyapeeth, Piparia, Waghodia, Vadodara, Gujarat, India

ABSTRACT

Scleromyxedema is a chronic, progressive condition characterized by confluent lichenoid eruption. It is usually associated with paraproteinemia. We present the case of 52 years old man diagnosed scleromyxedema without malignancy and paraproteinemia and treated with oral methotrexate and betamethasone.

Keyword :- Scleromyxedema, Malignancy

INTRODUCTION

Scleromxyedema is rare disease affecting men and women. It is chronic progressive condition characterized by dermal fibrosis and mucinosis. It is almost always associated with monoclonal paraproteinemia usually immunoglobulin G type. Herein we present the case of a 52 years old man with classical cutaneous features of scleromyxedema. This was confirmed by H&E (Hematoxylin and eosin stain) and special stain through skin biopsy. However no evidence of malignancy was obtained in spite of intensive investigations. This case is being presented due to the rare occurrence of scleromyxedema in absence of malignancy.

CASE REPORT

A 52 years old male patient presented with history of skin lesions all over body and progressive tightening of skin over face and body since 10-12 years. He gave history of difficulty in mouth opening and difficulty in swallowing. However there was no history of photosensitivity, Raynaud's phenomenon, muscle weakness or weight loss. The patient had received multiple treatments since the last 3-4 months to no avail. On examination, skin over face showed waxy translucent papules with loss of lateral one third of bilateral eyebrows. Waxy papules seen over upper back, bilateral retroauricular area and pinnae, (Figure 1a) resulting in stony hard feel of both pinnae. (Figure 1b) Skin was taut to touch with mouth opening reduced to two fingers. There was diffuse tightening of skin underlying the papules over the entire trunk. (Figure 2a) Loose folds of skin were seen over bilateral axilla, back, and groin. (Figure 2b) Patient was unable to flex the fingers of both hands due to tightening of skin over dorsum of both hands. There were no stellate ulcers, neither any evidence of calcinosis. Nail discoloration and nail dystrophy was seen over toe nails. Differential diagnosis considered was lepromatous leprosy, scleredema,

***Correspondence:**
E-mail: drparthshekhat@ymail.com

nephrogenic systemic fibrosis, systemic sclerosis, sarcoidosis, and granular annulare. Routine investigations including LFT were within normal limits (HB: - 10.2g/dl, TC 5200/cumm, RBS 154mg/dl, blood urea 42 mmol/L, creatinine 1.1 mg/dl). Chest x ray and USG (abdomen and pelvic) were normal. ELISA for HIV was negative. Serum protein electrophoresis (using agarose gel electrophoresis by Helena) showed normal levels of serum protein bands (gamma 1.28 g/dl, albumin 3.75 g/dl alpha-1 0.28 g/dl, alpha-2 0.82, beta 0.80g/dl). Histopathology revealed thinned out epidermis along with deposition of mucin in upper and mid reticular dermis accompanied by widespread proliferation of fibroblasts. This was confirmed by alcian blue staining.

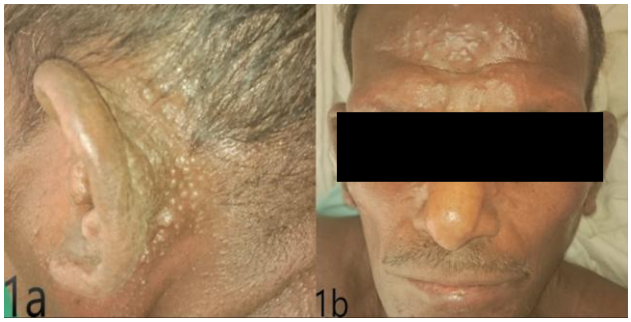

Figure 1: (a) waxy translucent papules in retroauricular region (b) taut shiny skin with waxy papules over the fore head

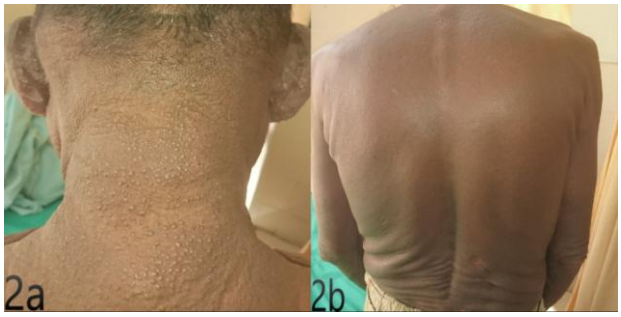

Figure 2: (a) waxy papules over nape of neck and upper back (b) increased folds of skin in upper lateral and lower trunk

DISCUSSION

Scleromyxedema is rare chronic and progressive disease. Middle age adults are usually affected. Its classical clinical features include generalized symmetric eruption of firm waxy papules on the upper trunk, extremities, neck, face and ears, occasionally resembling leonine facies of leprosy.[1]

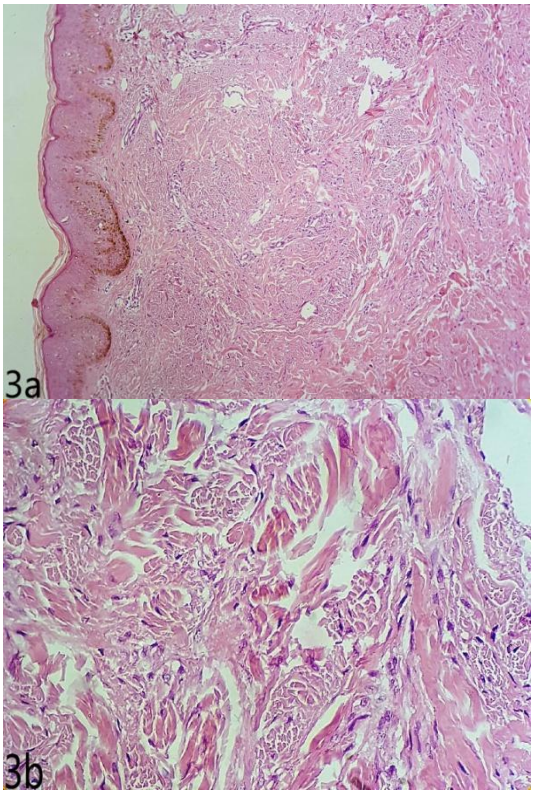

Figure 3: (a) Thinned out epidermis, deposition of mucin in upper and mid reticular dermis (b) Mucin deposition, proliferation of fibroblasts

Multiple systemic disorders such as neurological, cardiac, ophthalmological disorders, renal, muscular (proximal myopathy), gastrointestinal (esophageal dysmotility), and pulmonary are frequent associations. All of these together may result in severe disability and may be fatal in some cases.[2] It never affects mucous membranes and rarely affects the scalp. It is also known as the Arndt-Gottron syndrome or generalized lichen myxedematosus.[3]

Paraproteinemia is most common feature of Scleromyxedema. In 80% patients, monoclonal

gammopathy is IgG with lambda light chains type.[3] Atypical cases of lichen myxedematosus have been reported with absence of monoclonal gammopathy, albeit with the plethora of cutaneous and systemic clinical signs.[4]

Diagnostic criteria for scleromyxedema[4]

1. Generalized papular eruption and sclerodermoid features
2. Microscopic triad of mucin deposition, fibroblast proliferation and fibrosis
3. Monoclonal gammopathy
4. Absence of thyroid disorder

Histopathologic features [3]

- Diffuse dermal mucin deposition
- Increased collagen
- Numerous irregularly shaped fibroblasts

Table 1: Rongioletti's proposed classification for scleromyxoedema in 2006[4]

Generalised Form	Localized Forms	Atypical Forms
Scleromyxoedema	1) Discrete papular LM 2) Acral persistent papular mucinosis 3) Juvenile and adult variants of self‑healing popular mucinosis 4) Papular mucinosis of infancy 5) Nodular LM	1) Scleromyxedema without monoclonal gammopathy 2) Localized LM with monoclonal gammopathy 3) Localized LM with mixed features of different subtypes 4) Not otherwise specified

Table 2: Proposed staging of scleromxedema[3]

Stage I	limited cutaneous papular mucinosis
Stage II	Generalized cutaneous mucinosis and/or extracutaneous manifestation(s)
Stage III	Generalized cutaneous mucinosis and/or extracutaneous manifestation(s) and disease related Karnofsky PS <50%

Histopathology of scleromyxedema shows extensive proliferation of fibroblasts throughout the dermis with irregularly arranged collagen bundles. Collagen bundles are split by mucin. Amount of mucin is greater in the upper half than in the lower half of the dermis. Both scleromyxedema and nephrogenic fibrosing dermopathy are characterized by a dermal infiltrate of spindle cells with increased mucin and collagen, so it is difficult to distinguish scleromyxedema from nephrogenic fibrosis.[5]

Treatment of the scleromyxedema is disappointing inspite of significant advances in understanding disease pathogenesis. Many treatment options have been described in literature with limited efficacy.[2] Among the various treatments described are topical and systemic corticosteroids(daily or pulsed dose), psoralen combined with ultraviolet A (PUVA), oral retinoids, chloroquine, methotrexate,[6] thalidomide,[7] IVIG,[8] interferon-α and cyclosporine.[9] Therapeutic procedures such as plasmapheresis,[10] radiotherapy, autologous stem cell transplantation, extracorporeal photochemotherapy have also been tried. A combination of BCNU, etoposide, cytarabine and melphalan (BEAM regime) has been recommended prior to autologous stem cell transplantation.[3] Our patient was started on tab methotrexate 7.5 mg/week and betamethasone 0.5 mg (6tab/week). The patient reported mild

improvement in sensation of tightness over face and shoulders after 3 months of treatment, however cutaneous lesions were persistent.

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