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CASE REPORT Open Access

Atypical Scleromyxedema with Dramatic Response to Low Dose Prednisolone and Thalidomide

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Abstract:

Scleromyxedema is a rare condition which is clinically characterized by asymptomatic to itchy generalized papular and sclerodermoid eruption with histological findings which include mucin deposition and fibroblast proliferation. It can be associated with monoclonal gammopathy. Abnormalities in thyroid function must be ruled out in all cases where scleromyxedema is suspected.
There have been several treatment options tried which includes chemotherapy, glucocorticoids, thalidomide, intravenous immunoglobulin and extracorporeal phototherapy. However, no satisfactory conclusion has been drawn. The case described represents scleromyxedema with absence of monoclonal spike or M peak. In our case, the patient showed significant clinical improvement in symptoms like movement of jaw, neck movement and improved mobility of upper and lower limbs within one month of treatment with prednisolone and thalidomide. There was also visible improvement in appearance of patient and papular lesions associated with scleromyxedema.

Keywords: Prednisolone, Scleromyxedema, Thalidomide

INTRODUCTION

Scleromyxedema is a rare chronic cutaneous fibro mucinous disorder of unknown cause with normal thyroid function test, first described in 1953 by Montgomery and Underwood [1]. which was revised in 2001 by Rongioletti and Rebora, who classified it as having presence of generalized papular and sclerodermoid eruption along with fibrosis, mucin deposition and monoclonal gammopathy [2]. These features should be present in the absence of thyroid disease. Disease progression leads to extensive skin infiltration producing significant disability. Several systemic manifestations like neurological, in form of encephalopathy and seizures; gastrointestinal manifesting as malabsorption and abnormal motility of the gut; rheumatological and joint abnormalities; muscle weakness; respiratory complications, and cardiovascular manifestation may occur [3].

CASE REPORT

A 47 year male, presented in our outpatient department with chief complaints of generalized itching, difficulty in opening of the mouth, tightness during neck movement and gritty sensation while moving his fingers, for 6 months which progressed within a month.

On examination, generalized symmetrical waxy brownish yellow raised lesion was present, having predilection for sun-exposed areas. The face showed typical leonine appearance (Figure 1), doughnut sign attributable to thickening of the proximal interphalangeal joints with a central depression was present, and skin bound down sign was obvious (Figure 2).

There was negative history of other systemic features. Systemic examination was within normal limits. On investigation, his thyroid profile was within a normal range. His complete blood count, renal profile, liver profile, general blood picture, autoimmune profile, viral markers, ESR, CRP and chest X-ray was within normal limits, serum and urine protein electrophoresis revealed no M peak. Bone marrow biopsy revealed no abnormal plasma cells. Skin biopsy was done, and microscopy was performed revealing unremarkable epidermis with superficial dermis showing fibroblastic proliferation with collagen bundles and mucin deposition. Perivascular lymphocytes infiltrate present Alcain Blue stain highlighted increased dermal mucin with features favoring the clinical diagnosis of Scleromyxedema (Figure 3).

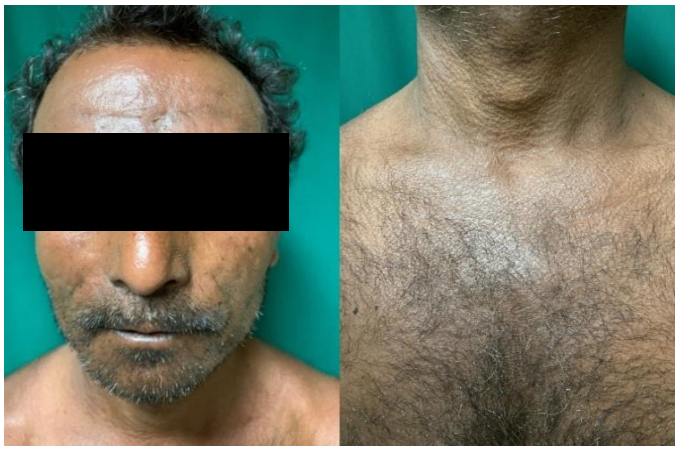

Figure 1| Presence of generalized papular eruptions showing predilection to photo exposed sites; with characteristic leonine face.

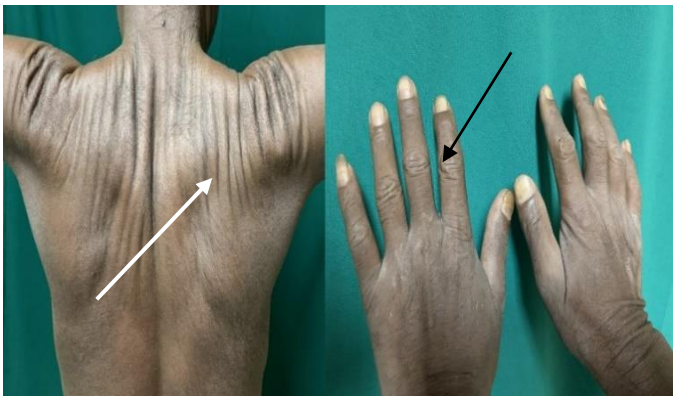

Figure 2| Characteristic doughnut sign attributable to thickening of the proximal interphalangeal joints with a central depression (indicated by black arrow). Presence of skin bound sign in Scleromyxedema (indicated by white arrow).

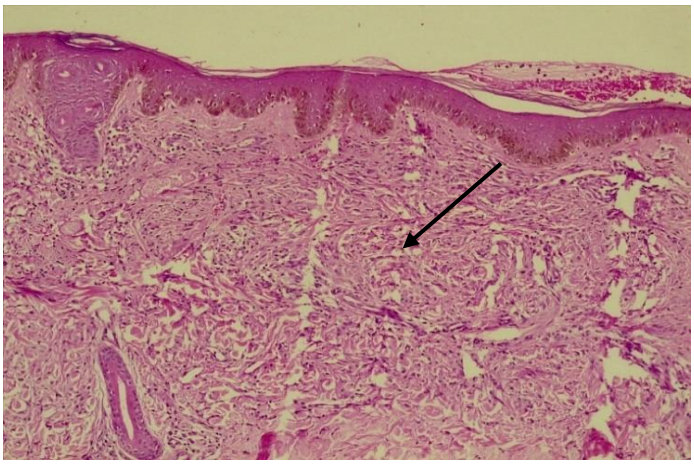

Figure 3 | Microphotograph of skin biopsy revealing unremarkable epidermis with superficial dermis showing fibroblastic proliferation with collagen bundles and mucin deposition (highlighted by black arrow). Perivascular lymphocytes infiltrate present Alcain Blue stain highlighted increased dermal mucin with features favoring the clinical diagnosis of Scleromyxedema. [H&E stain, 100x]

He was started on 20-milligram prednisolone equivalent and Thalidomide 200 mg, both once daily. Topical corticosteroids; cream mometasone furoate 0.1%(w/w) was given to be applied over the lesions of scalp, face, neck, axilla, nipple and groin area whereas cream clobetasol propionate 0.05%(w/v) was given for lesions over other parts of body. Adjunctive treatment with proton pump inhibitor, vitamin D_3 with calcium in tablet form and antihistamines tablet was prescribed. He was then followed up after one month of initial treatment during which marked improvement in symptoms was

present, especially while moving jaw, neck and limbs. There was visible improvement in papular eruptions and appearance of the patient (Figure 4).

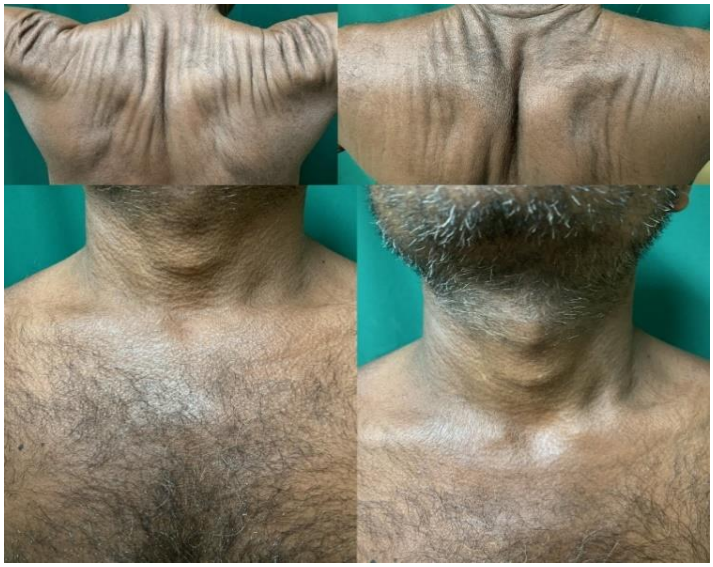

Figure 4| Pictures taken pre-treatment (left) and post-treatment at one month (right) - showing visible improvement in appearance with marked decrease in skin bound sign on extension of shoulder joint and decrease in papular eruptions in photo exposed area

DISCUSSION

Localized variants are confined to skin though it can progress into generalized form involving internal organ. So careful evaluation must be done during gradual follow up. Death generally results due to respiratory failure and infection. Treatment of paraproteinemia is the mainstay of treatment. Several treatment options have been tried like chemotherapy, glucocorticoids, thalidomide, intravenous immunoglobulin, extracorporeal phototherapy [4–8], with variable response. There have been satisfactory results in eight out of 10 patients treated with IVIg [9]. In recent literature, remission of scleromyxedema in number of patients with severe disease with autologous peripheral blood stem cell transplantation was observed [10]. Overall results have been unsatisfactory, also reflected by many treatment modalities available and tried. This case is proposed because of its atypical presentation and marked response in improvement of symptoms and skin lesions to treatment during follow up.

ADDITIONAL INFORMATION AND DECLARATIONS

Ethics approval and consent to participate: Not applicable

Consent for publication: Informed consent was obtained from the patient for publication.

Acknowledgment: None

Competing interest: None

Author Contributions: PKJ: Diagnosis, Conceptualization, Writing – Original Draft Preparation, Writing – Review & Editing; **SKS:** Conceptualization, Writing – Original Draft Preparation, Writing – Review & Editing; **RR:** Writing – Original Draft Preparation, Writing – Review & Editing; **AK:** Writing – Original Draft Preparation, Writing – Review & Editing; **SP, HSY and DP:** Writing – Review & Editing

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