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Case Report

Lichen Scleromyxoedema: Rare Disorder, Excellent Response to Steroid Pulse and PUVA Therapy: A Case Report

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ABSTRACT

Lichen is a rare, chronic, progressive, fibro-mucinous disorder of unknown etiology characterized by numerous waxy lichenoid papuloplaques with extensive thickening and hardening of skin in the absence of thyroid disease. It involves the skin of face, trunk, forearms and hands. Characteristic cutaneous involvement is associated with fibroblast proliferation and mucin deposition in upper dermis, monoclonal gammopathy and systemic multi-organ involvement and can be fatal. Despite anecdotal reports of success with various agents, no satisfactory treatment is currently available. We report a 41 year old male with Lichen scleromyxoedema who responded excellently to weekly dexamethasone pulse and PUVA therapy.

Key Words: Lichen, scleromyxoedema, mucinosis, monoclonal gammopathy, dexamethasone pulse, PUVA.

INTRODUCTION

Lichen scleromyxoedema is a rare disorder involving extensive mucin deposition, fibrosis and monoclonal gammopathy in middle aged adults, 30-80 years. [(1)] Four diagnostic criteria help to distinguish it from other sclerdermoid disorders,

1. Generalised popular and sclerdemoid eruptions.
2. Mucin deposition, fibroblast proliferation and fibrosis.
3. Monoclonal gammopathy
4. Absence of thyroid disease

Histopathology it is characterized by a diffuse deposition of mucin in papillary and mid reticular dermis, increased collagen deposition and proliferation of fibroblasts. [(2)] Gottron coined the term scleromyexoema in 1954 to describe this disease of unknown etiology. [(3)] Since then it became apparent that this disease have increased incidence of several systemic abnormalities, it can involve any organ of body. It has progressive dermal mucin deposition causing skin thickening and variety of gastrointestinal, neurological, pulmonary, cardiac and renal complications. It is notoriously difficult to treat. It has a chronic and many times a fatal course. [(4)]

CASE REPORT

We report a 41 year old male who presented in outdoor department of dermatology with asymptomatic swelling, tightening and thickening of skin over scalp, face, neck and upper chest for 3 weeks duration. He was primarily treated for

urticaria and angioedema at a secondary level health institution with no relief. Progressively he developed thickening and hardening of skin over extremities and trunk (back more than abdomen). He also developed shortness of breath, difficulty in diglutation and proximal muscle weakness with difficulty in standing from supine position. He was a febrile with no urinary and bowel complaints. No history suggestive of chronic disease like diabetes, hypertension and tuberculosis.

Examination revealed moderately built male with multiple, asymptomatic, skin colored small 2-5mm size waxy, firm, closely spaced papules over shiny, indurated and non-tender skin over scalp, face, neck, trunk back more than abdomen, forearms and hands symmetrically. Facial skin was indurated, swollen with deep creases showing leonine face (figure1). Over the proximal interphalangeal joints, a central depression surrounded by elevated rim, "doughtnut sign" (figure 6). Hair, nail and mucous membranes were normal on examination.

On general physical examination there was no lymphadenopathy and no organomegly on systemic examination.

Histopathology revealed normal epidermis with abundant alcian blue stained mucin in reticular dermis with increased fibrocytes, increased space between collagen bundles and sparse superficial peri-vascular lymphocytic infiltrate.

Hematological and biochemical parameters were in normal limits except low serum proteins. Thyroid functions and chest radiograph show no abnormality. Serum electrophoresis revealed IgG gammopathy. Muscle enzymes were in normal range. Urine analysis was normal with no proteins. Ultrasonography and echocardiography revealed mild plural effusion, ascitis and pericardial effusion (figure 8,9). Pulmonary functions showed mild obstructive defect.

He was started on high dose dexamethasone 40mg/day for 4days/week along with PUVA therapy three day/ week. Excellent response over a period of 8 weeks was observed with complete remission in 12 weeks. Patient reviewed after one year without any intervening relapse.

DISCUSSION

Dureuilh in 1906 and Reitmman in 1908 originally scripted scleromyexedema. Lichen myxedematosus was coined by Gotton in 1954. Rorgioletti and Rebora further classified lichen myxedematosus into two clinical subsets: Generalised (scleromyxedema) and localized (popular mucinosis). [(5)] Generalized variant have more cutaneous involvement along with systemic features and can be fatal some time. This necessitates prompt and aggressive therapy to minimize morbidity and mortality.

Exact patho-physiology of lichen myxedematosus remains unknown. Vitro studies reveals stimulates hyaluronic acid and prostaglandin E production by fibroblasts in patient's serum. It is also noted that fibroblasts in these patients produces more mucin then do the normal fibroblasts. [(5)] Monoclonal gammopathy is associated in 83% of times [(6)] but progression to multiple myeloma is rare. Systemic features can range from dysphagia, difficulty in chewing, horseness of voice, dsypnoea, aspiration pneumonia, ectropion, lagophthalmos, proximal muscle weakness, arthralgia, migratory arthritis, carpel tunnel syndrome, memory loss, vertigo, gait disturbances, halloucinations, seizures and coma. [(7)]

Paraprotein levels do not correlate with extent or progression of the disease. The role of M paraprotiens is questionable as this view is supported by the enhanced fibroblast proliferation in response to patient's serum where as purified paraprotein IgG from the same patient fails to stimulate a similar response. Thus the role of unknown serum factor is considered responsible for stimulating fibroblasts to deposit excessive mucin and collagen in skin and other tissues. [(8)]

Unknowm patho-physiology and lack of randomized trials, limited number of

case reports make it difficult to assign a therapeutic ladder to treat this rare entity. Multiple therapeutic agents have been tried with variable results including melphan, interferon alfa, autologous stem cell transplantation, thalidomide, cyclophosphamide, plasmapharisis and IV Ig and including systemic steroids.

IVIg 2g/kg and thalidomide 150 mg/day may be effective novel therapeutic combination in refractory disease [(4)] but thalidomide posses risk of teratogenicity and peripheral neuropathy

Therapeutic response to melphalan though equivocal yet long term usage can lead grave toxicity including hematological malignancies and septic complications. [(9)]

Treatment of this rare disorder remains challenge to physicians. Even increased morbidity and mortality have been reported with aggressive therapy.

Rayson et al reported complete remission to prednisone for two years after discontinuation of therapy. [(10)]

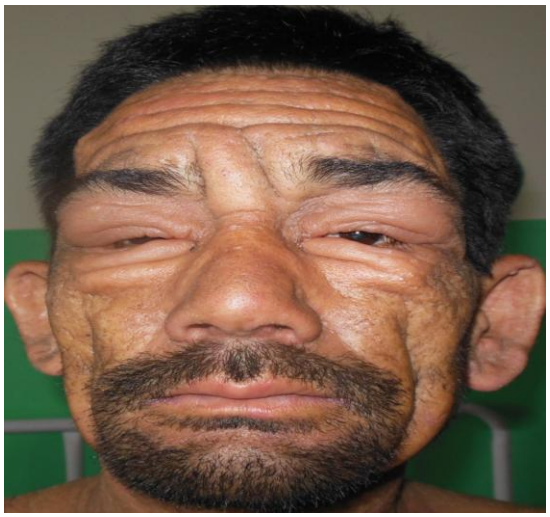


Figure1. Shows facial skin induration and edema with deep creases showing "leonine face".

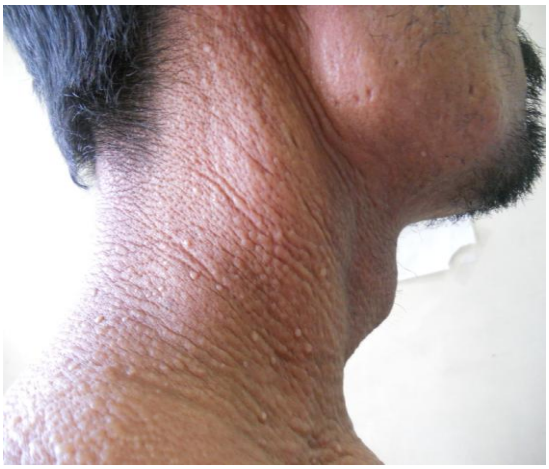
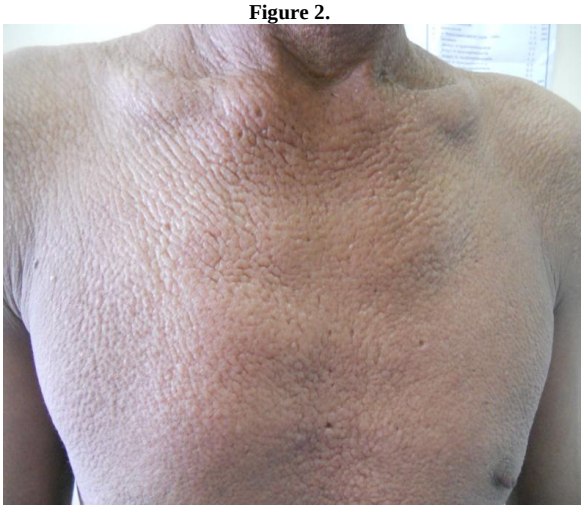


Figure 2.



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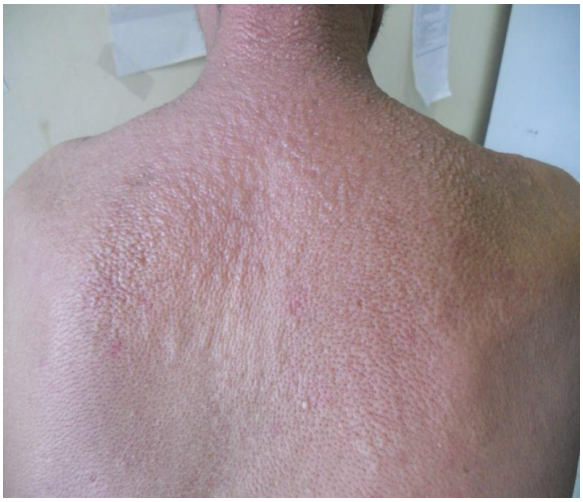


Figure4

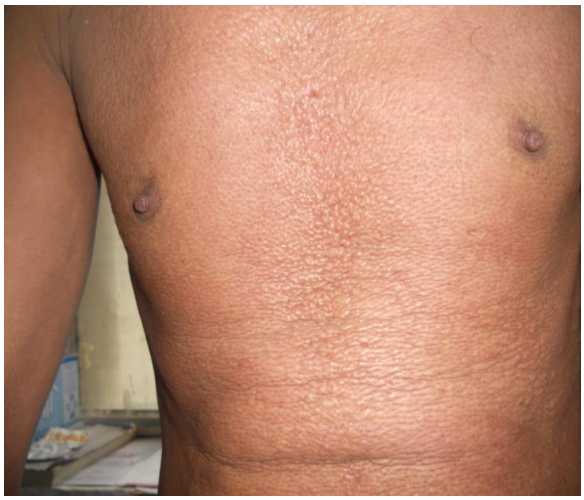


Figure 5

Figure 2, 3, 4, 5. Shows closely spaced, mildly erythematous, waxy, firm, papules over shiny, indurated and non-tender skin over neck, chest, lower abdomen and back.

In our patient use of high dose dexamethasone pulse in our patient was successful in inducing remission and patient remained in well till his last review after 1.5 years, dexamethasone is exceptionally long acting corticosteroid with potent glucocorticoid activity and preferably given in pulse therapy to alleviate risks associated with long term use of corticosteroids. Suppression of immune response and fibroblast proliferation is the mechanism leading to decreased paraprotein synthesis and decreased mucin production. Alkalating agents should be reserved for corticosteroid refractory disease. [(11)] PUVA had little beneficial effect on the skin thickening as reported by Schirren et al. [(12)]

CONCLUSION

We advocate pulse dexamethasone therapy as first line treatment option supplemented with PUVA therapy in lichen scleromyxoedema, a rare and difficult to treat disease.

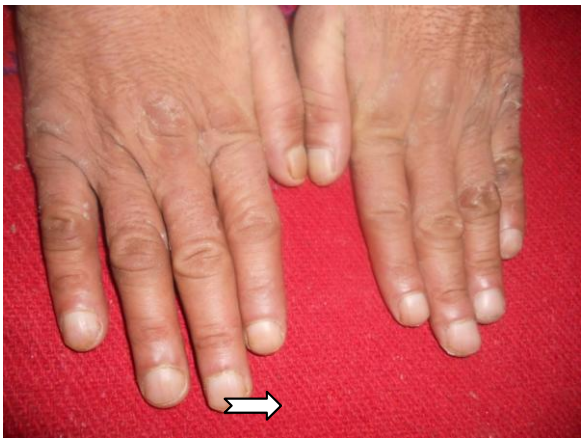
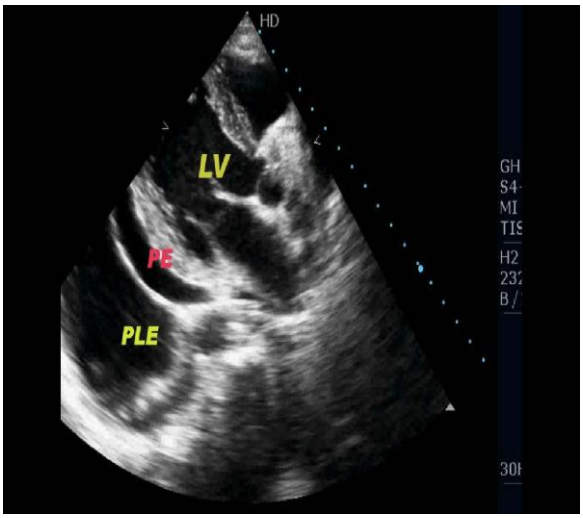


Figure 6. Over the proximal interphalangeal joints a central depression surrounded by elevated rim, "doughtnut sign".



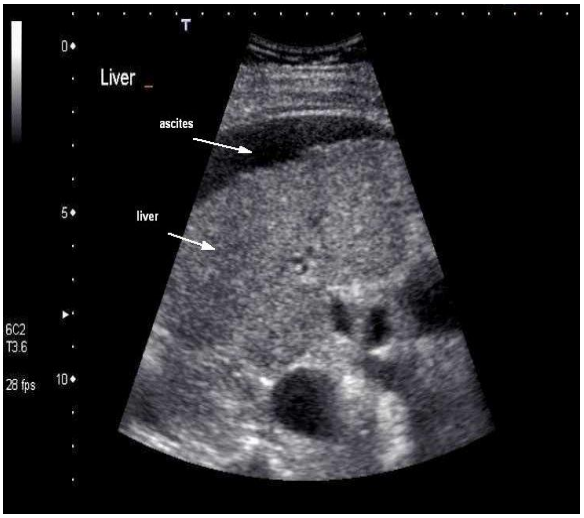


Figure 7,8. Echocardiography and Ultrasonography revealed mild plural effusion, ascitis and pericardial effusion.

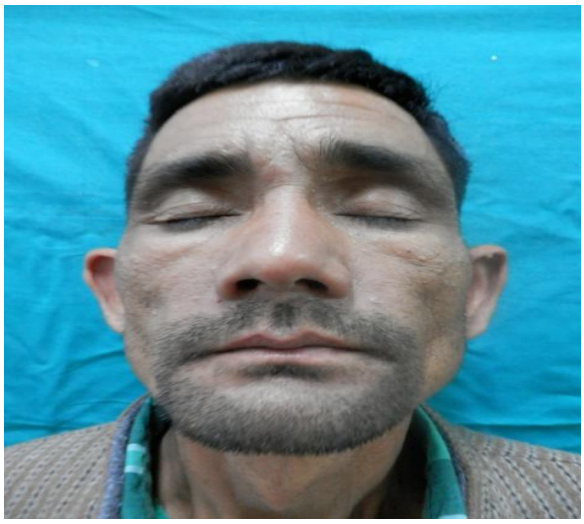
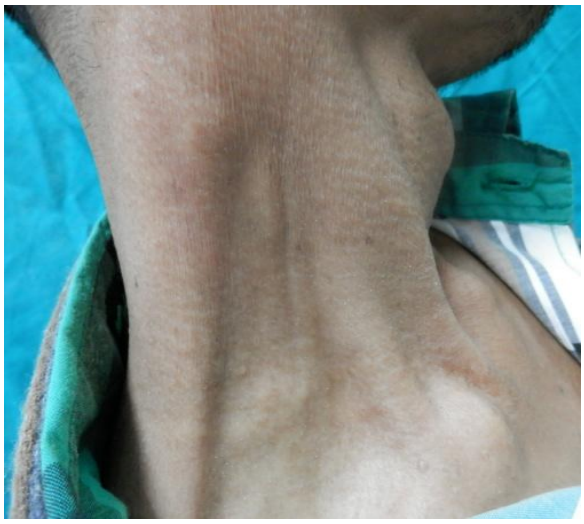



Figure 8,9. Post treatment marked improvement with no lesions and cutaneous induration.

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