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Case Report - Scleromyxedema: An Atypical Form in a 50 Years Old Male Patient.

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Abstract: *Scleromyxedema is a chronic, progressive condition characterised by dermal fibrosis and mucinosis. The clinical variants consist of generalised confluent lichenoid eruptions (scleromyxedema) with systemic manifestations and a localised variant with discrete papular eruptions. The cases which do not fit into either of the category are termed as atypical or intermediate form. We report such a case of a 50 years old male patient with numerous asymptomatic erythematous papules mainly over his face and diffuse hyperpigmentation with thickening of skin of his body with no systemic complaints. All of his investigations were within normal limits and histopathology showed large amount of mucin deposit in reticular dermis with increased fibrocytes and increased spaces between collagen fibres.*

Keywords: *scleromyxedema, lichenoid, papular, mucin.*

I. Introduction

Scleromyxedema (SM), is a chronic progressive disease characterized by generalized lichenoid papules and is a rare disorder of connective tissue. It is considered among the papular mucinosis group of cutaneous disorders and also referred to as "generalized lichen myxedematosus". [1],[2],[3] Although its etiology remains unknown, most theories focus on a pathogenic role by paraproteins; however, nonparaprotein factors have also been suggested to cause fibroblast proliferation and increased mucin production. [2]

LM was originally described by Dulbreuilh in 1906. [7],[10] Montgomery and Underwood [7] gave the first clinical classification in 1953. According to an updated classification by Rangioletti in 2001, LM includes includes 2 clinicopathologic subsets: a generalized papular and sclerodermoid form (scleromyxedema) with systemic, even lethal, manifestations and a localized form, which does not run a disabling course. A third group of atypical or intermediate forms, not meeting the criteria for either scleromyxedema or the localized form, includes cases of (a) scleromyxedema without monoclonal gammopathy, (b) localized forms with monoclonal gammopathy and/or systemic symptoms, (c) localized forms with mixed features of the subtypes, and (d) not well-specified cases. [1,4]

Ours case belongs to an atypical or intermediate form, not having either monoclonal gammopathy or any systemic symptoms.

II. Case Report

A 50 yrs old male patient came to the skin OPD of RIMS, Ranchi with the complaint of restricted movements in his fingers and thickening and darkening of skin over his face and body for the last 2 years. He was normal 3 years back when he started developing multiple well defined tiny papules over both his legs which gradually merged and thickened. Simultaneously diffuse hyperpigmentation and thickening of skin started developing all over his body. There was thickening of skin over his palms and difficulty in flexing his fingers associated with tingling sensation on his fingers especially on exposure to cold or on washing his hands.

On cutaneous examination, he had multiple asymptomatic, well defined, slightly erythematous papules all over his face more prominent on forehead, nose, ears, cheeks and chin.[Fig 1; Fig 2; Fig 3] A few of the papules coalesced to form plaques. Thickening and exaggregation of skin creases were present around eyes and nasolabial folds. Other areas of body had marked diffuse hyperpigmentation and thickening of skin with the texture like that of a “thick leather”.[Fig 5] Exaggregation of skin folds were present especially over his neck and back.[Fig. 4, Fig. 6,Fig.7]. The palms were thickened with flexion deformities of fingers and pallor was prominent. [Fig.8, Fig. 9]

He consulted many local doctors (including ayurvedic and homeopathic) for his condition but was not relieved. He also took MDT-MB (adult) for 3 months the previous year but no improvement occurred. He had no past history of diabetes mellitus, thyroid disorders, hypertension, tuberculosis, asthma.

His complete blood count, routine examination and culture sensitivity of urine, liver function test, renal function test, blood sugar and thyroid profile was within normal limits. Serum protein electrophoresis and bone marrow biopsy were also within normal limits.

Histopathology of skin lesions on hematoxylin-eosin stain showed abundant mucin within reticular dermis with sparse superficial perivascular lymphocytic infiltrate.[Fig.10] Reticular dermis also had increased number of fibrocytes and increased amount spaces between collagen bundles.[Fig.11] Any other stain like alcian blue or colloidal iron could not be done due to its unavailability in our centre.

The patient was initially counselled for the treatment with thalidomide; but as the patient belonged to a low socio-economic group, he showed his inability to afford the drug due to its high cost and long term use. The patient was then started on tablet hydroxychloroquine (200mg) twice daily. On a follow up visit about a month later, he did not showed any considerable improvement and the patient did not returned again for subsequent follow up.

III. Discussion

Scleromyxedema is a chronic, progressive and a rare skin disease of unknown etiology that affects men and women with equal frequency in their mid to later life.[6] Lichen myxedematosus, papular mucinosis and scleromyxedema are synonyms, and the condition is characterised by formation of numerous lichenoid papules which coalese to form generalised plaques causing extensive thickening and hardening of skin.[1],[4],[7]. Lichen myxoedematosus is uncommon. On review of literature approximately 115 cases have been reported in the world. Associated myopathy [11], seronegative polyarthritis, bizarre neurological findings including psychosis [12], accelerated coronary disease [13] , hepatomegaly and lymphadenopathy [14] have been reported in a few patients. It can also present with severe pruritus, scalp involvement, unrestricted mobility and associated eosinophilia without paraproteinemia. [14]

In our case the patient did not have any systemic involvement and had no paraproteins on electrophoresis. He however had raynauds phenomenon with no telangiectasias or calcinosis. The characteristic histopathologic triad which includes diffuse dermal mucin deposition, increased collagen and numerous irregularly shaped fibroblasts [8] was present in our patient. Thus, according to the clinical presentation and histopathological evidence, this was case of an atypical scleromyxedema.

The pathogenesis of scleromyxedema is not very clear, but serum from patients with scleromyxoedema had enhanced fibroblast proliferation with mucin deposition. Paraproteins or other unidentified factors in the blood may be the causative factors. [11, 15] However, nonparaprotein factors have also been suggested to cause fibroblast proliferation and increased mucin production. [2] In this case however no paraproteinemia was found.

There is no cure for scleromyxoedema. Treatments that have been used include intralesional hyaluronidase, topical oral or intralesional corticosteroids, psoralen ultraviolet A, Grenz irradiation, electron-beam therapy, retinoids, plasmapheresis, extracorporeal photochemotherapy and dermabrasion. Chemotherapeutic agents, particularly low-dose melphalan, have induced some improvement. A trial of steroid oral minipulse and methotrexate has also been tried with some success. [16]

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Scleromyxedema photographs.

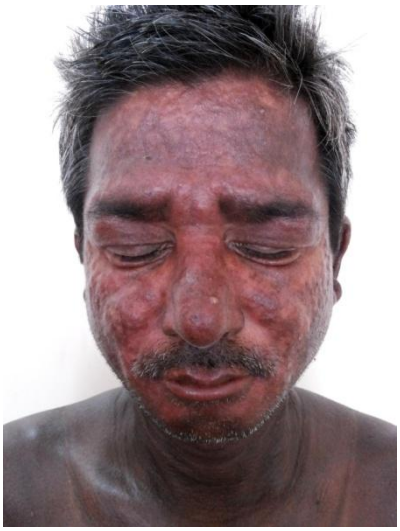

Fig 1: erythematous papules and plaques present over the forehead, cheek and nose.

Fig 2: multiple erythematous papules and plaques over the face. Ear lobe infiltration is also seen.

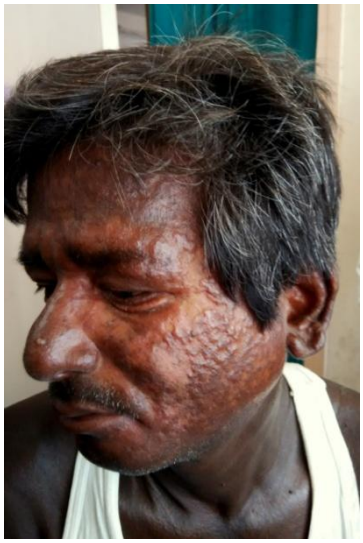

Fig 3: multiple erythematous papules and plaques over the face. Ear lobe infiltration is also seen with marked hyperpigmentation in neck region.

Fig 4: multiple erythematous papules and plaques over the face. Ear lobe infiltration is also seen with marked skin folding over the neck.

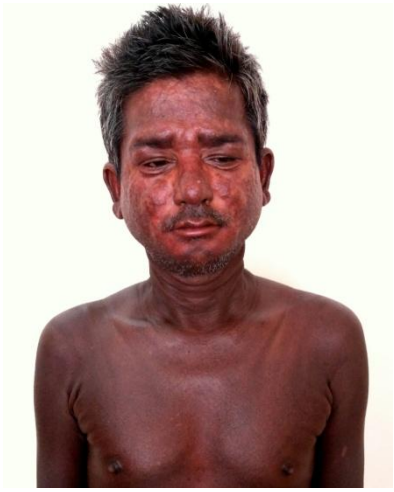

Fig 5: erythematous plaque and papules on face with marked hyperpigmentation of skin over upper truncal area.

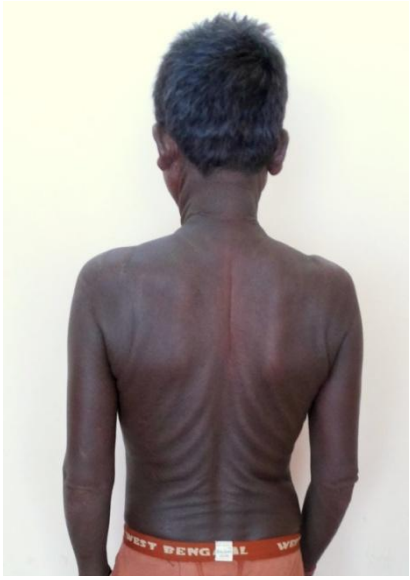

Fig 6: marked diffuse hyperpigmentation of the skin and increased folding over the back.

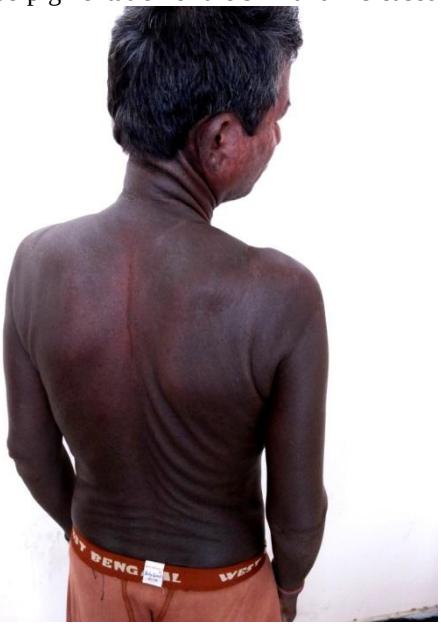

Fig 7: marked hyperpigmentation and thickening of the skin over back along with marked skin folding on neck and lower back.

Fig 8: marked pallor of the palms with flexion deformities of fingers on hand.

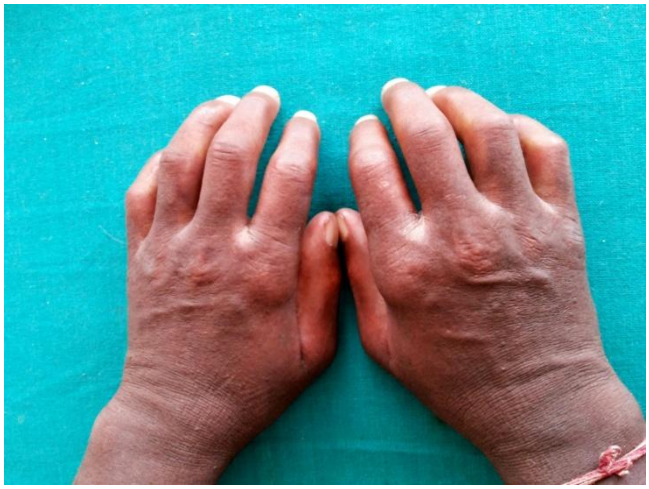

Fig 9: The hands could not be fully extended with a thickened and hyperpigmented skin over the dorsum of the hands.

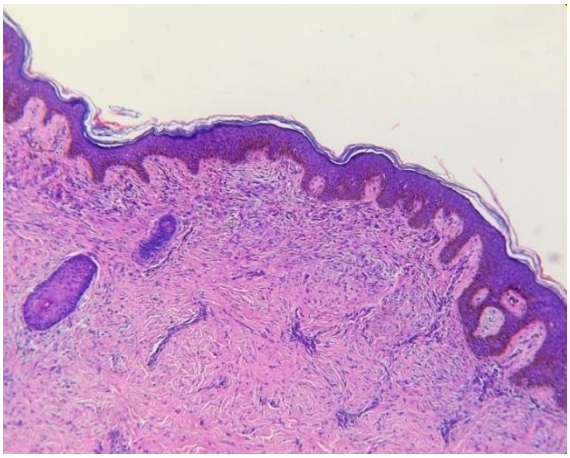

Fig 10: hematoxylin and eosin staining (10x) showing abundant mucin within reticular dermis with sparse superficial perivascular lymphocytic infiltrate. Reticular dermis also had increased number of fibrocytes and increased amount spaces between collagen bundles.

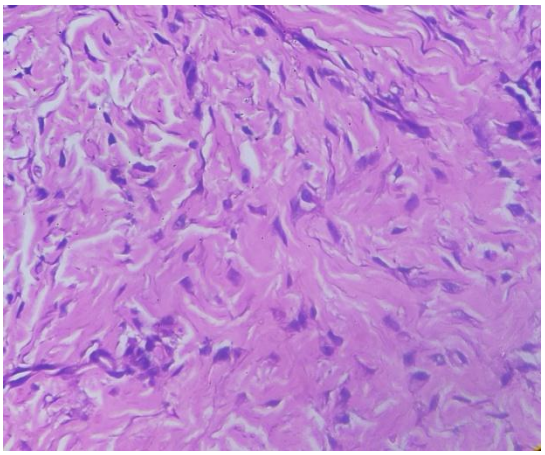

Fig 11: hematoxylin and eosin staining (40x) showing abundant mucin with increased fibrocytes and increased spacing between collagen bundles.