SCLEROMYXOEDEMA

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A 27-year-old male having scleromyxoedema is reported. The lesions were moderately pruritic, 2 to 3 mm papules situated over sheets of thickened, waxy skin on the face, neck, trunk and extremities. Paraproteinemia was absent, though the bone marrow showed a mild plasma cell hyperplasia.

Key words : Scleromyxoedema, Lichen myxoedematosus, Paraproteinemia, Plasma cell hyperplasia.

Scleromyxoedema is a rare disorder associated with paraproteinemia, proliferation of fibroblasts and dermal mucinous infiltration. Characteristic cutaneous changes include diffuse thickening of the skin along with confluent groups of lichenoid papules. Scleromyxoedema is a variant of lichen myxoedematosus. It has been given various names, such as lichen myxoedematosus, papular mucinosis and myxoedema.

Case Report

A 27-year-old wrestler had an episode of fever, which was followed by the appearance of moderately pruritic papular lesions over his trunk and face. A few days later, the eruption spread over the face, neck and trunk, each lesion measuring 3 to 5 mm in size and became symmetrical. This was accompanied by a gradually increasing swelling around the eyes, and the skin became thickened, firm and waxy. Sheets of non-pitting oedema were seen predominantly over the face, neck, axillae, back, lower abdomen, flexural aspects of the upper limbs and extensor surface of the thighs. The face was puffy, somewhat leonine, and the indurated skin was thrown into folds over the forehead, neck and axillae. The ear lobules were large. Follicular openings over the lesions were exaggerated (Figs. 1 and 2). There was no evidence of any systemic illness. The patient was well built and nourished. The thyroid was not enlarged. The tongue was moderately enlarged with impressions of teeth on its margins.

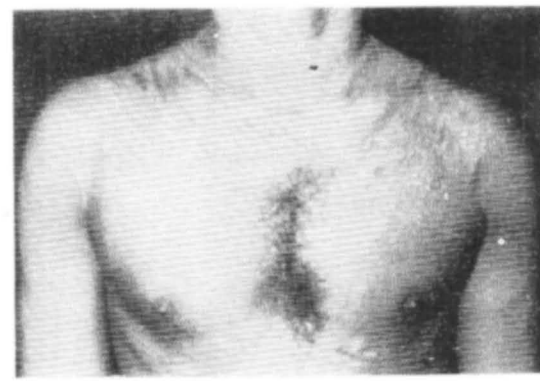

Fig. 1. Papulo-nodular lesions over the front of chest and exaggerated skin folds on the sides of neck.

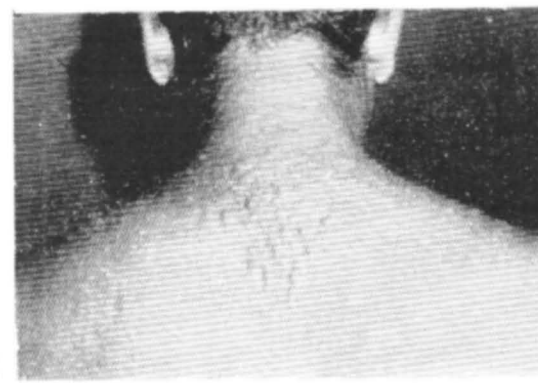

Fig. 2. Papulo-nodular lesions situated on thickened and indurated skin.

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Routine tests of urine, stools, blood electrolytes, urea, creatinine, SGOT and SGPT were within normal range. There was slight hypogammaglobulinaemia. Thyroid function tests were normal. Serum cholesterol was 310 mg percent and total phospholipids were 312 mg percent (both were elevated). Bone-marrow aspiration revealed a mild increase in plasma cells.

Hematoxylin-eosin stained section (Fig. 3)

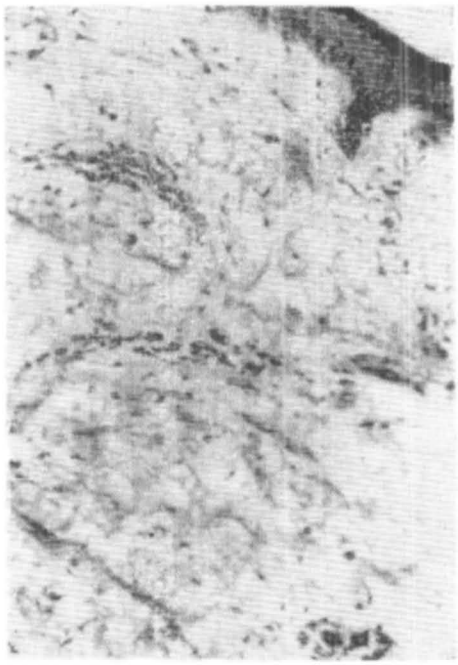

Fig. 3. Deposition of mucin in the dermis causing splitting of collagen bundles (H and E x 200).

of the skin lesion showed an increased amount of a lightly staining mucinous material in the upper and mid-dermis, intermingled with stellate, large and prominent fibroblastic reaction and associated with irregularly arranged bundles of collagen. The mucinous deposit caused dissociation and splitting of the collagen bundles. The material was PAS positive, hyaluronidase labile and could be stained with toluidine blue (Fig. 4).

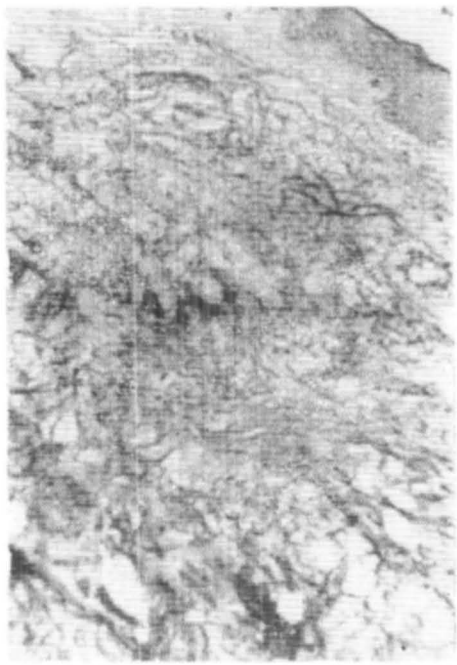

Fig. 4. Fibroplasia and mucin deposits in the dermis (Toluidine blue x 200).

Comments

Scleromyxoedema was first described by Gottron in 1954.[1] The peculiar and characteristic cutaneous lesions are a regular feature, while the paraproteinemia is variable. The abnormal protein may not be detected for years after the appearance of cutaneous lesions,[2] even though the fibroblastic proliferation and the dermal mucinous deposition continue to progress.

The basic etiology of scleromyxoedema is unknown. Though there are histopathological similarities with myxoedema, thyroid dysfuction has not been reported.

Perry et al[3] classified lichen myxoedematosus into four categories. (1) Generalised, uniformly distributed lichenoid papules 2 to 3 mm in size, having predilection for the hands, forearms, trunk, face and neck. The underlying infiltration of the skin may be so extensive as to lead to furrowing of the forehead,loss of facial expression and functional difficulties. (2) Discrete papular

lesions occurring on the trunk and extremities. The lesion are few, often less than fifty in number. (3) Generalised lichenoid plaques. (4) Nodular lesions or urticaria like plaques. When the disease is extensive and generalised, the prognosis is bad and systemic complications can occur. Runder et al[4] described weakness, weight loss, debility, vertigo, mental deterioration, peripheral nerve signs, dysphagia, myocardial infarction and even death. Unexplained neurological abnormalities have been the feature of some cases.[2,5] The symptoms vary in severity from headache and a transient disturbance in speech, to a rapidly progressive mental deterioration with death due to grand mal convulsions and coma. Cerebral haemorrhage and infarction have been demonstrated at postmortem, but mucopolysaccharide infiltration of the brain has not been described.[5] Myocardial infraction and cerebrovascular disease have also been reported.[2,6]

Our patient belonged to the first category. To the best of our knowledge no such case has so far been reported from India. Serum glucuronic acid and sulfated acid mucopoly saccharides have been reported to be elevated in these patients.[4] Fieldman et al[7] reported association of scleromyxoedema with abnormal serum proteins (gammaglobulin containing light chains of lambda type) and plasma cell hyperplasia in scleromyxoedema.

There is no specific treatment for scleromyxoedema. Corticosteroids, methotrexate, melphalan, superficial X-ray therapy, radiation therapy, topical hyaluronidase, vitamin A and E, growth hormones and gonadotrophins have been tried with varying eesults.[2,4,9-12] The association with a plasma cell disorder should be looked for and treated with cytotoxic drugs such as melphalan; otherwise, the use of potentially dangerous cytotoxic drugs and prednisolone is unjustified.[8] Some authorities however, regard the prognosis to be poor, death occurring at a relatively early age from myocardial infarction or cerebrovascular disease.[2,6] Lowe et al[2] reported therapeutic response to whole body electronbeam irradiation. Recently, Farr and Ive[12] reported significant improvement in a case on treatment with PUVA.

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