SCLEROMYXEDEMA

A K Bajaj, S C Gupta, A Tripathi and P A Singh

Scleromyxedema was observed in a 43-year-old male patient. The lesions were shiny waxy-looking, soft papules, 2 to 4 mm in diameter and were most marked on the neck and ear lobules. There was no evidence of paraproteinemia.

Key words : Scleromyxedema, Lichen myxedematosus.

Scleromyxedema, first described by Gottron[1] in 1954, is a rare disorder clinically characterised by a generalised extensive eruption of asymptomatic, soft papules, usually 2-3 mm in diameter, associated with diffusely thickened erythematous skin. The papules are densely grouped, but do not coalesce. Recently, Singh et al[2] have reported a single case of scleromyxedema from India. Because of the comparative rarity of case reports, the present communique was considered worthy of record.

Case Report

A 43-year-old male had developed thickening of the skin on the face, neck, upper extremities, upper trunk and proximal part of lower extremities for the past two years. The skin markings were prominent. There were shiny, waxy looking, soft papules, 2 to 4 mm in diameter, most marked on the neck and ear lobules (Fig. 1). The skin, though thickened, was freely mobile. General and systemic examination did not reveal any abnormal findings except for puffiness of the face. Thyroid was not enlarged.

Routine laboratory investigations were normal. Fasting blood sugar, total serum proteins, A:G ratio, serum urea and creatinine and SGOT and SGPT all were within normal limits. Rheumatoid factor test and LE cell phenomenon were negative. The value of IgG was slightly raised. It was 2334.0 mg/dl as against the normal range of 626 to 2304 mg/dl of serum. IgA and IgM were within normal limits. There was no laboratory evidence of paraproteinemia. Serum cholesterol and total phospholipid values were found to be increased to 317 and 330 mg percent respectively.

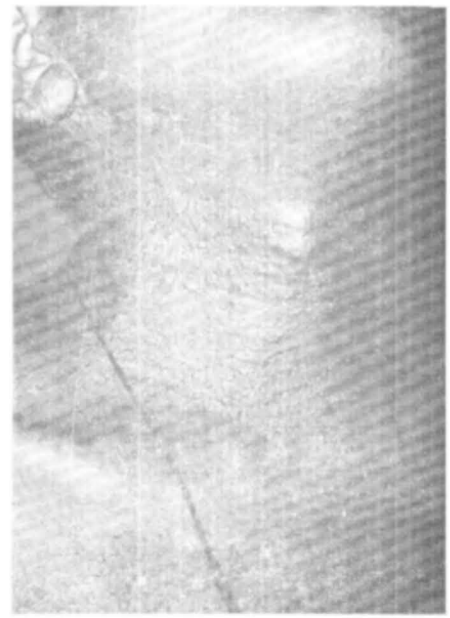

Fig. 1. Shiny, waxy-looking, soft papules most marke on the neck and ear lobule.

Histopathological examination of the papules revealed a slightly hyperplastic epidermis. Within the dermis, there were numerous pilosebaceous units. Scattered between these hair follicles were dense proliferations of fibroblasts surrounded by abundant mucinous material (Fig. 2). Histochemically, this material was found to be an acid mucopolysaccharide as

From the Departments of Dermatology and Pathology, M.L.N. Medical College, Allahabad, India.

Address correspondence to : Dr. S. C. Gupta.

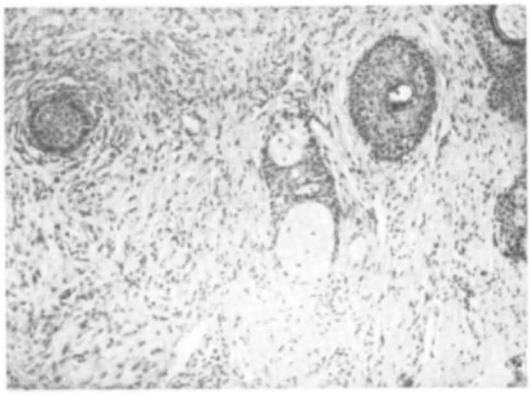

Fig. 2. Scattered between the hair follicles, dense proliferation of fibroblasts surrounded by abundant mucinous material (H & EX100).

supported by the metachromasia seen with the toluidine blue stain.

Comments

Scleromyxedema is a variant of lichen myxedematosus in which peculiar and characteristic cutaneous lesions are accompanied with extensive fibroblastic proliferation in the dermis. An upper dermal mucinous infiltrate is the hallmark of the lesion. The acid mucopolysaccharides present in the mucin contain hyaluronic acid which is hyaluronidase-labile and stains metachromatically with toluidine blue.[3] The present case also showed metachromasia in the dermis with toluidine blue, suggesting the presence of acid mucopolysaccharide.

An abnormal serum IgG which almost always possesses lambda type light chain has often been reported with scleromyxedema.[3] However, Lai et al[4] reported the association of paraproteinemia to be variable. It might not be detected for years after the appearance of cutaneous lesions. Except for a slightly raised level of serum IgG, there was no evidence of paraproteinemia in the present case even after 2 years of the cutaneous lesions. Even the bone marrow examination in this case was normal, although most of the cases showed hyperplasia of plasma cells in the bone marrow.[3]

The symptoms of the disease may vary from weakness, weight loss, vertigo and dysphagia to mental deterioration, myocardial infarction,[5] cerebro-vascular accidents,[6] grand-mal convulsions and coma.[4] This patient of ours did not have any systemic manifestation and has lived normally till date.

Histopathologically, scleromyxedema simulates myxedema and scleroderma. Myxedema is associated with hypothyroidism. In scleroderma, the thickened skin is not freely mobile. However, scleromyxedema is neither associated with hypothyroidism nor is the thickened skin fixed in this disease.

The basic aetiology of scleromyxedema is unknown. Direct immunofluorescence has demonstrated the presence of abnormal IgG in the dermal mucin.[7] Whether this IgG stimulates fibroblasts to increased activity[8] or it represents an auto-antibody which is a consequence rather than the cause of the disease[9] is controversial.

Acknowledgement

The authors are thankful to Dr. Daniel F. Richfield, Laboratory of Dermatopathology, Ohio, U.S.A. for histochemical studies and confirming the diagnosis.

References

1. Gottron HA : Scleromyxedema, Arch Dermatol Syphilol, 1954; 199 : 71-91.
2. Singh M, Kaur S, Kumar Bhushan et al : Scleromyxedema, Ind J Dermatol Venereol Leprol, 1985; 51 : 111-113.
3. Feldman P, Shapiro L, Pick AI et al : Scleromyxedema. A dramatic response to melphalan, Arch Dermatol, 1969; 99 : 51-56.
4. Lai A, Fat RFM, Suurmond D et al : Scleromyxedema (Lichen myxedematous) associated with a paraprotein IgG of the type Kappa, Brit J Dermatol, 1973; 88 : 107-116.
5. Rudner EJ, Mehregan A and Pinkus H : Scleromyxedema. A variant of lichen myxedematosus, Arch Dermatol, 1966; 93 : 3-12.

6. Bleehan SS : Metabolic and nutritional disorders,in : Text-book of Dermatology, 3rd ed, Editors, Rook A, Wilkinson DS and Ebling FJG : Blackwell Scientific Publications, Oxford, 1979; p 2059.

7. McCarthy JT, Osserman E, Lombardo PC et al : An abnormal serum globulin in lichen myxedematosus, Arch Dermatol, 1964; 89 : 446.

8. Piper W, Hardmeier T and Schafer E : Das Sklero-myxodem Arndt-Gottron : Eine paraproteinamische Erkrankung, Schweiz Med Wschr, 1967; 197 : 829.

9. Laurence DA, Tye MJ and Liss M : Isolation and characterisation of the basic diagnostic IgG globulin in the rare gammopathy, papular mucinosis, Prep Biochem, 1971; 1 : 1.