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SCLEROMYXOEDEMA

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A case of a 25-year-old male with Scleromyxedema is reported. The typical papular lesions with underlying woody, fibrous, sclerosis of skin were seen.

Key Words : Lichen myxoedematosus, Scleromyxoedema

Introduction

Scleromyxoedema, also known as the Arndt-Gottron Syndrome,[1] is a variant of lichen myxoedematosus, a rare disease characterised by fibrocytic proliferation, increased deposition of mucopolysaccharides in the skin, and the presence of a circulating paraprotein. There cases of scleromyoedema have been reported in India, until now,[2,3,4] Here, we peresent a case of classical scleromyxoedema.

Case Report

A 25-years-old male reported to the outpatient department of the SSG Hospital, Baroda, with densely grouped papular lesions over a thickened lichenified skin, mainly over the exposed parts, for 2 months. The lesions were pruritic. Glabellar furrowing and nodular infiltration of the ear were seen. There was thickening and loss of hair over the eyebrows. The underlying skin was markedly indurated on palpation.

Investigations done included complete haemogram, which was within normal limits. Urine analysis showed mild proteinuria, but Bence-Jones proteins were absent. Serum cholesterol, liver and renal function tests and serum electrophoresis were reported normal. Smear for Acid Fast Bacilli was done from the ear and eyebrows, and was reported negative. Bone marrow studies showed mild plasmacytosis. Biopsy from a papular lesion showed a normal looking epidermis, with clear spaces in the dermis. Staining with Alcian blue at pH 2.5 showed mucin in the dermis.

Treatment was initiated with prednisolone, in the dose of 30 mg daily, but the patient absconded after 10 days treatment, rendering follow-up impossible.

Comments

Lichen myxoedematosus occurs in adults in the third to fifth decades. Perry et al[5] have described 4 clinical presentations, viz., a generalised lichenoid eruption, or as a discrete papular eruption over the trunk and extremities (with, or without diffuse sclerodermatous changes), or as localized or generalised lichenoid plaques, or as urticarial plaques and nodules, especially over the trunk. The confluent papular and sclerotic form is called scleromyxoedema, wherein, diffuse thickening of the skin underlies the papules. Diffuse acral thickening without papulation, sclerodactly, Raynauds phenomenon, associated myopathy, seronegative inflammatory polyarthritis and a few cases of accelerated coronary or cerebrovascular disease have been reported.[6] Serum electrophoresis may show a cationic monoclonal-type band of IgG class,[7] which was not seen in our patient.

The diagnosis of scleromyxoedema in this patients was arrived at by clinical findings,

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the presence of plasmacytosis in the bone marrow, and by biopsy and staining with mucin specific stain viz., Alcian blue.

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