SCLEREDEMA ADULTORUM OF BUSCHKE ASSOCIATED WITH NON- SCARRING ALOPECIA OF SCALP

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A-3- year-old boy presenting with scleredema of Buschke confirmed histopathologically is being reported for its unusual association with reversible non- cicatricial alopecia of the scalp.

Key words : Scleredema adultorum of Buschke, Alopecia

Introduction

Scleredema is a rare connective tissue disorder, characterised by the development of skin induration which resolves spontaneously. It frequently follows an infection in children and is associated with diabetes in adults. It is occasionally associated with multiple myeloma, rheumatoid arthritis, Sjogren's syndrome and primary hyperparathyroidism. [1-3]

Case Report

A 3- year old boy presented with progressive thickening and hardening of upper half of the body of one month duration. There was history of frequent episodes of fever and cough since early childhood, the last episode being 3 weeks prior to the onset of present complaints. The child was otherwise asymptomatic and was accepting feeds normally. He had normal developmental milestones and had diffuse nontender, nonpitting edema and induration of skin affecting face, frontal region of scalp, neck, chest, upper back and proximal upper limbs. Hands and lower limbs were completely spared. Three weeks after patient's initial visit he developed diffuse loss of hair from the frontal region of scalp (Fig.1). Routine investigations including complete hemogram, ESR, urine and stool, renal and liver function tests were normal. Throat swab, ASO titre, ECG and X-ray chest revealed no abnormality. Histopathological examination of biopsy specimen from upper back showed an unremarkable epidermis with increased overall thickness of dermis. The appendages were " pulled up" with the secretory portion of eccrine glands being located in mid dermis. Gaps between collagen fibres were noted (Fig.2) which stained positive

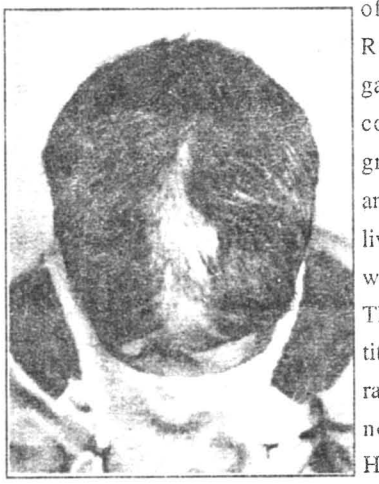

Fig. 1. Diffuse loss of hair in frontal region of scalp

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with alcian blue (pH2.5), thus confirming the presence of hyaluronic acid. Patient was given a course of amoxycillin for 10 days and vitamin E (200mg) daily for 3 months. There was complete resolution of induration and regrowth of frontal hair at the end of four months (Fig.4).

Fig. 2. Secretory portion of sweat gland located in mid dermis. Note the gaps between the collagen fibres which stained positive for hyaluronic acid (H& E x 100).

Discussion

Scleredema adultorum of Buschke is a misnomer as relatively high proportion of cases occur in children. Greenberg et al reported an incidence of 29% in children below 10 years of age and 22% between 10-12 years.[4] However, Venenice et al in a review of 33 cases have reported the age of onset of scleredema from 12-65 years.[5] All races are affected and occasionally the occurrence is familial. Amongst nondiabetic adults there appears to be female preponderance. The disease reaches maximum involvement in a few weeks and resolves spontaneously in months or years . There is an excess of hyaluronic acid in the dermis demonstrable as fenestration in the collagen bundles. On routine haematoxylin and eosin stained sections, the mucin present in scleredema lesions stains with colloidal iron. It is alcian blue positive at pH 2.5 but negative at pH 0.5 and shows metachromasia with toluidine blue at pH 7.0 and 4.0. Apart from cutaneous involvement, systemic manifestations like dysphagia, electrocardiographic abnormalities, hypertension, retinopathy, pleural and pericardial effusion have been encountered.[5] This patient had disease limited to skin following a respiratory infection which resolved relatively rapidly following treatment. Reversal of associated frontal alopecia along with the resolution of skin involvement suggests that the aetiopathogenesis of both these manifestations of the disease were identical. To the best of our knowledge, reversible non-scarring alopecia has not been earlier reported in association with scleredema.

References

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