SCLERODERMA (A CASE REPORT)

By

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Scleroderma is a disease of unknown aetiology characterised by diffuse indurative change throughout the connective tissue, due to increased deposition of collogen and obliterative lesions of small arteries (Price 1966). An unusual case presenting with autoamputation of the digit is presented.

Case Report

K. D. a 30 yr. old female was admitted to Sarojini Naidu Hospital, Agra on 26-7-68, the presenting complaints being recurrent ulceration of the finger tipes with partial loss of tip of right little finger.

The disease started 10 years back with stiffening and pigmentation of the skin all over the body, along with recurrent ulceration of the finger tips. The ulcer occured especially in winter. After exposure to cold she used to feel burning and tingling sensation in the peripheral parts of the limbs. Apart from the acroparesthesiae she was having difficulty in opening the mouth, chewing the food and dysphagia. For the past four years she was being treated as a case of leprosy by DDS without any relief.

On examination She was a young lady with a posture of generalised flexion and a pinched face (Fig. 1). There was dark black pigmentation over the face, upper limbs and both feet. The nails were small, semilunar and beaked with swollen nail folds. There was a partial loss of distal phalanges of the right little finger and both index fingers (Fig. 2). The local skin had a reddish violet hue. The skin of the face and the forearm could not be pinched up from underlying structures. The hair were sparse, short and dystrophic. The toes were normal but there were tender ulcers on both heels. There was no sensory loss nor any palpable thickening of the nerves. Clinical examination of the cardiovascular and respiratory systems did not show any abnormality.

On Investigations–She had microcytic hypochromic anaemia with Hb. 9 Gm.%, corrected ESR being 36 mm/1st hr. Serum proteins were 5.72 gms.% with reversal of A:G ratio. No L. E. cells could be demonstrated. Urine showed traces of albumen without any pus cells/casts. Urine culture was sterile. Barium swallow, Electrocardiogram, x-ray of the chest and kidney biopsy did not reveal any abnormality. Nasal smear did not show mycobacterium leprae. Lepromin test was negative.

Skin Biopsy showed atrophy of epithelium with sclerosis of subepithelial tissue and a histological diagnosis of scleroderma was made.

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The patient was put on haemetenics, butazolidine, and peripheral vasodilators (Duvadilan). She was discharged on 6-9-1968.

COMMENTS

Like all other collagen vascular diseases, scleroderma is a disease of unknown aetiology. The essential histological lesion in the skin is a quantitative increasse in the amount of collagen. Immunoflucrescent studies have failed to localize any gammaglobulin deposits. The histological changes in the kindeys, skin and other organs merge into those of DLE and dermatomyosites.

Raynaud's phenomenon has been reported in 75% of the cases with occasional anhydrosis, loss of hair and indolent ulcerations, but the occurence of autoamputation of digits is unusual.

The extent of involvement of the heart, lung and kidneys varies from case to case. In early cases of dysphagia, ordinary barium swallow may fail to demonstrate the lesion and cineradiography may be necessary (Price).

No specific treatment is known. Corticosteroids may induce transient euphoria but aggravate the skin condition and renal changes and sometime push the patient into a rapidly progressive form of the disease. Potassium para-aminobenzoate, chelating agent, relaxants and peripheral vasodilators have been used (Cecil), with unsatisfactory results. Sympathectomy has been reported to relieve Raynaud's phenomenon in 30% cases.

Summary

An unusual case of scleroderma who presented with autoamputation of digit, and had been earlier mistaken for leprosy is described.

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