GENERALISED MORPHOEA WITH LICHEN SCLEROSUS ET ATROPHICUS AND UNUSUAL BONE CHANGES

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A 26-year-old male patient presented with multiple plaques on the limbs and trunk suggestive of morphoea. He also exhibited multiple, small, atrophic, hypopigmented macules on the left side of the trunk, the histopathology of which was consistent with lichen sclerosus et atrophicus (LSA). The patient developed large ulcers on the left leg and foot, and contractures with flexion deformity of the left ring and little fingers. Radiological examination revealed osteolytic lesions in the phalanges of the left hand. This combination of generalised morphoea with LSA and unusual osteolytic bone changes is uncommon.

Key Words : Morphoea, Lichen sclerosus et atrophicus

Introduction

Generalised morphoea is characterised by widespread sclerosis of an idiopathic nature. Large plaques are commonly seen on the trunk and upper thighs, but other areas may also be involved.[1] Localised or generalised scleroderma may coexist with LSA, which presents as hypopigmented macules and atrophic areas.[2,3] Various osseous lesions have been described in morphoea. They include radiological abnormalities in the spine or ribs, shortened ulna, contracted pelvis or deformities of the feet and toes.[4]

There is no universally accepted or proven therapy for scleroderma. Favorable responses have been reported with azathioprine, and potassium para-amino benzoate. Topical or intralesional steroids, antimalarials and Vitamin E have also been reported to be effective.

Case Report

A 26-year-old male tailor was admitted with the history of generalised skin lesions and joint stiffness for 3 years, multiple leg ulcers for 2 years, and drooping of the fingers of the left hand for 6 months.

On examination, there were multiple, well-defined, large, dull, erythematous, indurated and sclerosed plaques over the limbs and the left side of the trunk. The right axilla, abdomen and groin showed plaques with central clearing and a well-defined periphery with scaling. The plaque on the left hand involved both the palmar and the dorsal aspects and was indurated. There were hypopigmented, atrophic macules exhibiting Koebner's phenomenon on the face, the left side of trunk and the limbs (Fig. 1). The anterior aspect of the left leg and dorsum of the left foot showed large nonhealing ulcers.

Investigations revealed a Hb of 8 gm%, TC of 10,300/mm^3 with a normal differential count, and a ESR of 140 mm/hour, The urinalysis was normal, The FBS was 63 mg%, PPBS 70 mg%, urea 21 mg%, serum calcium 10.2 I.U., serum phosphorus 5.3 I.U. and serum alkaline phosphatase 21 I. U. The LE cell phenomenon was negative on three occasions and the VDRL was nonreactive. Tests for rheumatoid factor, antinuclear antibodies and serum immunoglobulin levels could not be done due to lack of facilities. Pus

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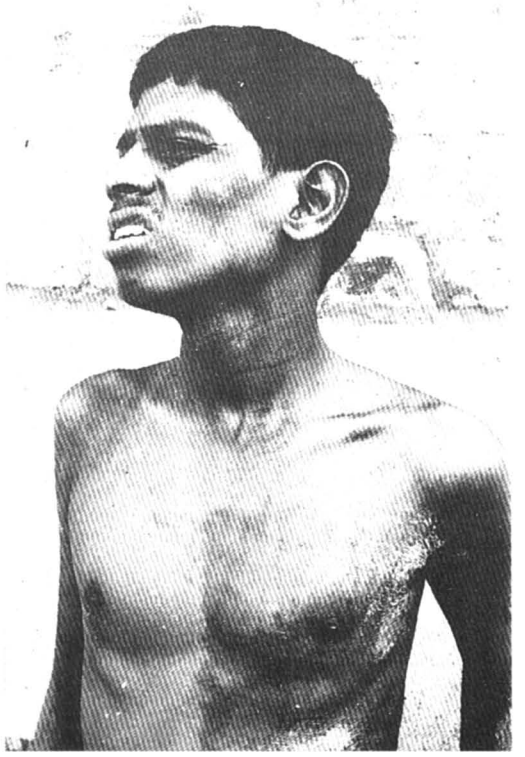

Fig. 1. Large atrophic patches on the left side of the trunk.

cultured from the ulcers revealed *Pseudomonas aeruginosa* sensitive to gentamycin and colistin. A skin biopsy from the hypopigmented macule revealed features suggestive of LSA (Fig. 2). The histopathology

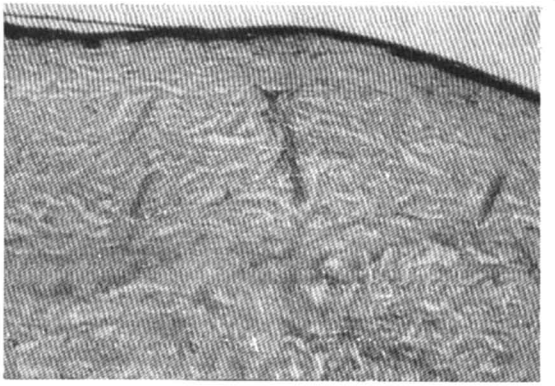

Fig. 2. Biopsy of a macule showing epidermal atrophy and homogenisation of the dermal collagen.

of one of the hyperpigmented plaques was consistent with the diagnosis of morphoea. Biopsy of the ulcer revealed on acute inflammatory cell infiltrate. An X-ray of the left hand showed osteolytic lesions on the proximal phalanx of the left ring and little fingers (Fig. 3). A skeletal survey was otherwise normal.

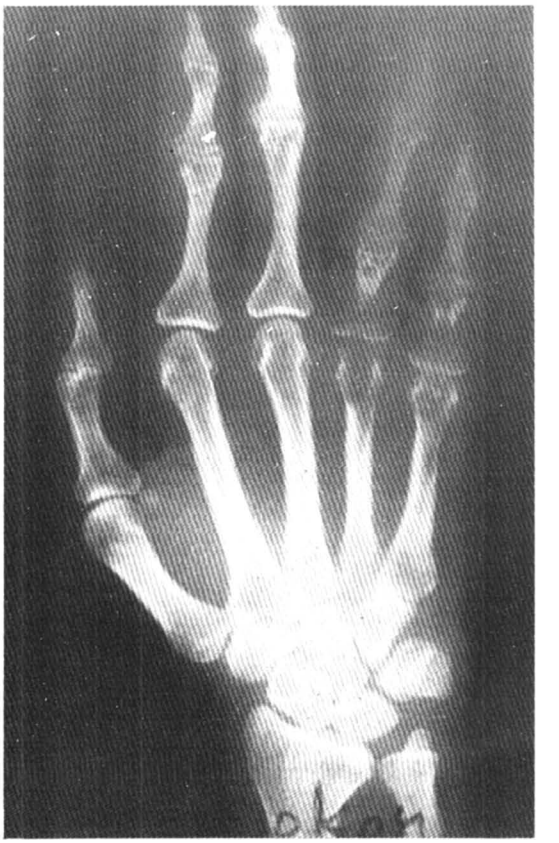

Fig. 3. Osteolytic lesions on the phalanges.

The patient was treated with chloroquine, 250 mg b.i.d., which was continued for 6 months at tapering doses. The ulcer healed with appropriate antibiotic therapy. Physiotherapy was given for the flexion deformity to prevent further contractures. After 6 months of treatment, the

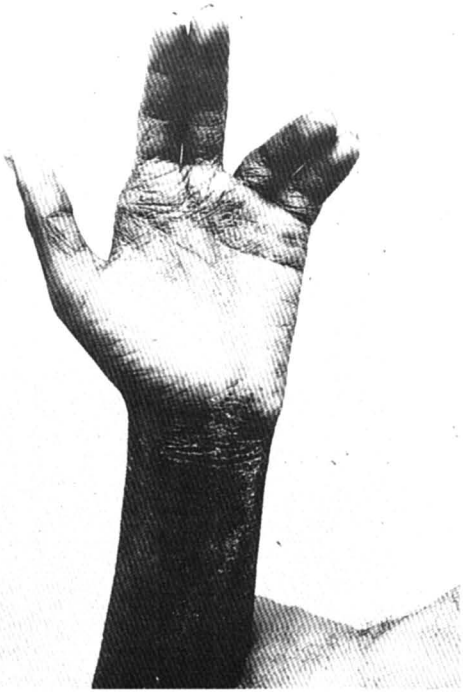

Fig .4. Flexion deformity of the left little and ring fingers (not numbered in the text).

patient was subjectively better. The colour of the plaques was returning to normal and their size and induration were less.

Discussion

Morphoea may be occasionally associated with LSA. The coexistence of LSA and morphoea has been reported in 10 patients by Uitto et al.[6] The histologic appearance of guttate lesions may vary from LSA to morphoea. This suggests that the aetiopathogenesis may be similar in both.[7] In our case the presence of LSA was confirmed by histopathological studies. This case presents a transitional form between plaque-like morphoea, generalised morphoea and LSA.

Many types of radiological abnormalities have been reported in localised or generalised scleroderma. Our patient exhibited osteolytic lesions on the proximal phalanges of the left ring and little fingers which led to the flexion deformity. These lesions, which are rare, are probably due to the presence of cutaneous changes. In addition, our patient also had chronic ulcers on the leg and foot which healed with appropriate antibiotic treatment. Other causes of leg ulceration, such as mycotic infection, malignancy, organic vascular diseases, necrobiosis lipoidica, were ruled out and the response of the ulcer to antibacterials points to a primary bacterial cause.

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