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Linear Scleroderma Occurring in a Patient with Systemic Lupus Erythematosus

– Short Report –

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A 38-year-old woman with systemic lupus erythematosus had developed a cutaneous lesion of linear scleroderma on the forehead 4 years after the diagnosis of lupus erythematosus. This case of coexistence of the two diseases can be regarded as a clinical variant of the connective tissue disease-overlap syndrome.

Key Words : *Linear scleroderma, Systemic lupus erythematosus.*

INTRODUCTION

Linear scleroderma is a type of localized scleroderma, generally considered to be a benign cutaneous form in the clinical spectrum of scleroderma. Although antinuclear antibodies can be found in patients with localized scleroderma, coexistence of this cutaneous lesions with systemic lupus erythematosus (SLE) is rare ; eleven cases have been documented in the English literature(Goldstein et al., 1990 ; Kleiner et al., 1989 ; Mackel et al., 1979 ; Shelkovitz et al., 1992).

In this report we describe a patient with SLE and her subsequent development of a linear scleroderma, which has not been documented in the Korean literature.

CASE REPORT

A 38-year-old woman had a facial lesion of brownish linear sclerosis on the forehead, which had first appeared 5 months prior to her visit. She was a known systemic lupus erythematosus (SLE) patient which was diagnosed at age 34 on the basis of the presence of nephritis, arthritis, hematologic disorders, antinuclear antibodies of high titer, and malar rashes.

In early 1993, when she recognized a cutaneous lesion developing on the forehead, the patient had been experiencing fatigue, arthralgia, and malar rashes. The activity of the SLE, at that time, seemed to be in a stage of aggravation, for which she was given prednisolone 40 mg/day over the next 6 weeks. The doses of the above remedy were slowly tapered, and subsequently the skin lesion became more apparent thus making a vertical linear groove (1.5×7cm) on the midforehead which spread down to the area of the nasal root (Fig. 1 A & B). Around the time of the dermatologic evaluation, she was taking prednisolone (15 mg/d) and hydoxychloroquine (200 mg/d). During this time the disease activity of the SLE was well controlled maintaining remission of nephritis and arthritis which she suffered previously. Besides the condition of the cutaneous lesion there were no other signs of systemic sclerosis/SLE-related cutaneous change. In her past and family histories there was nothing to be noted, and there had been no episodes of trauma

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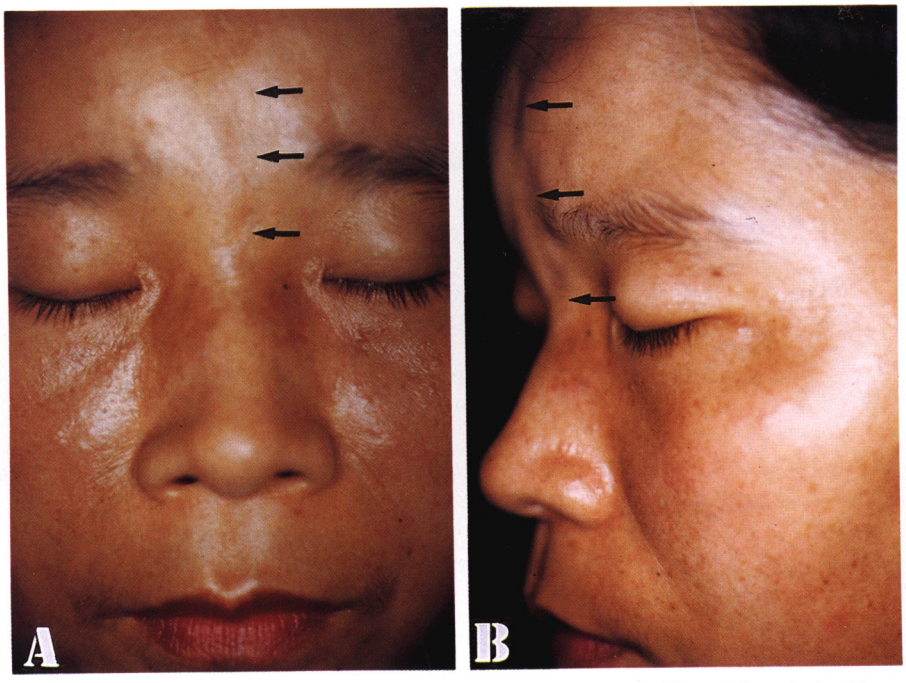

Fig. 1. A linear groove of sclerosis on the forehead ; frontal view(A) and lateral view(B).

or infection relevant to the skin lesion on the face.

A biopsy specimen from the forehead lesion showed thickened collagen bundles in the reticular dermis and moderate infiltrates of lymphohistiocytic cells between the collagen, and atrophic eccrine glands were also found in the deep dermis (Fig. 2).

Laboratory data of complete blood cell count, urinalysis, and stool guiac were within normal limits or negative. Erythrocyte sedimentation rate was 38 mm/hr. Chest roentgenogram was normal. Serum protein elctrophoresis revealed a polyclonal gammopathy. Quantitation of serum immunoglobulins showed an increased concenturation of IgG (2,200 mg/ml). Studies for complement profile (C3, C4, CH50), rheumatoid factor, VDRL, cryoglobulin, and results of kidney and liver function tests were in the normal ranges or negative. The assay for antinuclear antibodies using HEp-2 cell substrates showed positive at a titer of 1 : 160 (cytoplasmic pattern).

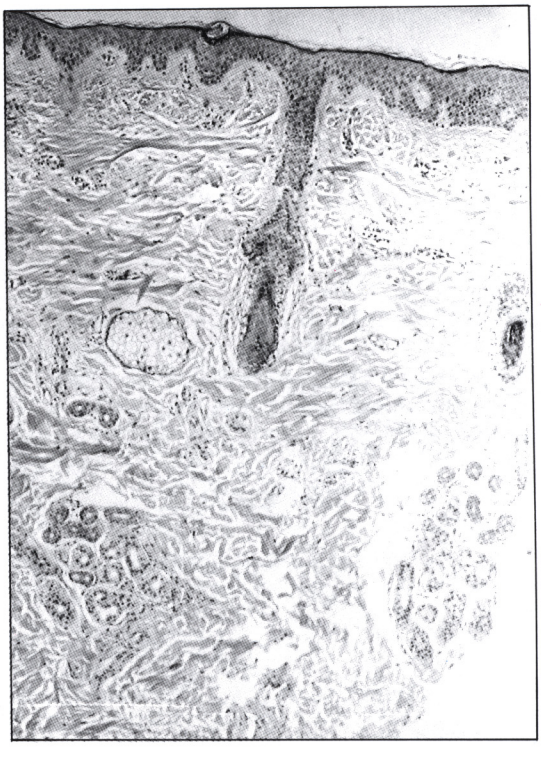

Fig. 2. Histologic study of a specimen taken from peripheral part of the lesion shows thickened/increased collagen bundles in the reticular dermis with some atrophic eccrine glands (H & E, ×100).

And antibodies against nDNA, Sm, Ro, La, Scl-70, Jo-1 were all negative in immunodiffusion test.

The patient was diagnosed to have linear scleroderma which occurred in the setting of SLE. Intralesional injections of triamcinolone suspension have been administered, in addition to the oral medicine being taken for the preexisting condition of SLE.

DISCUSSION

In the above case, features of linear scleroderma (en coup de sabre type) had developed in a patient sufferring from SLE. Combinations of scleroderma and other connective tissue diseases are not that rare and the coexistence of scleroderma and SLE have been previously reported (Goldsten et al., 1990 ; Hayakawa et al., 1993 ; Kleiner et al., 1989 ; Mackel et al., 1979 ; Shelkovitz et al., 1992). However, a case of localized scleroderma (linear scleroderma/morphea) which occurs around the time of a suspected clinical exacerbation of SLE, as in this patient, is a unique one. Although sporadic cases of concurrences of localized scleroderma and SLE have been described (Dubois et al., 1971 ; Goldstein et al., 1990 ; Kleiner et al., 1989 ; Mackel et al., 1979 ; Mitchel et al., 1980 ; Scarola et al., 1975 ; Shelkovitz et al., 1992 ; Tuffanelli et al., 1966), most of these patients had cutaneous lesions of scleroderma which preceded the onset of SLE by months or years ; in contrast to this woman patient and those cases described by Goldstein et al (1990) and Shelkovitz et al (1992) who showed SLE appearing 2 to 17 years before the onset of linear scleroderma or morphea.

Disorders of supposed autoimmune pathogenesis are occurring with increased frequency in patients with a previous history of another autoimmune disease. The polyglandular failure syndrome may be the best example of this (Eisenbarth et al., 1979). The relationship between SLE and linear scleroderma in this patient is not known, however there are possibilities that this case is a mere coincidence of the two diseases ; or there is an autoimmune association based on common pathogenic mechanisms causing the development of the two diseases presented as a clinical variant of connective tissue disease-overlap syndrome in this susceptible individual.

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