Coexistence of Recurrent Generalized Morphea and Systemic Sclerosis

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Sir,

Systemic sclerosis (SSc) is a generalized disorder of the connective tissue in which there is thickening of dermal collagen bundles, fibrosis and vascular abnormalities in the visceral organs (1). Generalized morphea (GM) is characterized by indurated plaques with widespread pigmentary change (1). Because sclerodactyly and visceral involvement are not observed in patients with GM, GM is thought to be a different entity from SSc. However, there have been several reports that suggest a relationship between the two disorders, e.g. coexistence of morphea and SSc, and the progression from morphea to SSc (2, 3). We describe here a case of a man with SSc who had recurrent episodes of worsening GM.

CASE REPORT

A 66-year-old Japanese man presented with erythemas on his chest and abdomen in September 1996 (Fig. 1A–B). Over the preceding 6 months, he had experienced erythema with itching on his chest. There were no other relevant medical, family or drug histories. He had been employed as an office worker for 40 years. There was no reported exposure to chemicals or toxins. A physical examination revealed erythematous sclerodermatous plaques on his trunk and slight sclerodactyly. The results of laboratory tests, including a full blood count, urinalysis and liver function test, were normal, except for an elevated antinuclear antibody (ANA) titre: ×640 (speckled) (normal values: <×40), C-reactive protein (CRP): 3.4 mg/dl (normal <0.2) and erythrocyte sedimentation rate (ESR): 35 mm/h (normal < 20). Tests for a range of antibodies were negative: anti-centromere, anti-Scl70, anti-dsDNA, and anti-RNP. A chest X-ray film and computerized tomography (CT) scan revealed lung fibrosis in the bilateral lower lobes. The patient reported experiencing heartburn and regurgitation due to reflux. An endoscopy revealed findings consistent with gastroesophageal reflux disease. There were no evident clinical features, such as Raynaud's phenomenon, pitting scars or contractures of the fingers. A skin biopsy from his trunk showed a thickening and homogenization of collagen bundles and perivascular lymphocytic infiltrates (Fig. 1C). His condition fulfilled two items from the 1980 American College of Rheumatology minor criteria for SSc. According to the classification proposed by LeRoy et al. (4), this case presented with limited cutaneous SSc.

We diagnosed the patient with a coexistence of morphea and SSc. He was admitted to our hospital and started on oral prednisolone, 30 mg/day. His symptoms responded well to this therapy and the dosage of prednisolone was tapered to 5 mg/day, taken for a period of 24 months. His erythema with induration had receded completely, leaving only residual pigmentation by November 1998. We continued to treat him with prednisolone 2.5 mg/day over the subsequent 3 years, without recurrence of noticeable lesions.

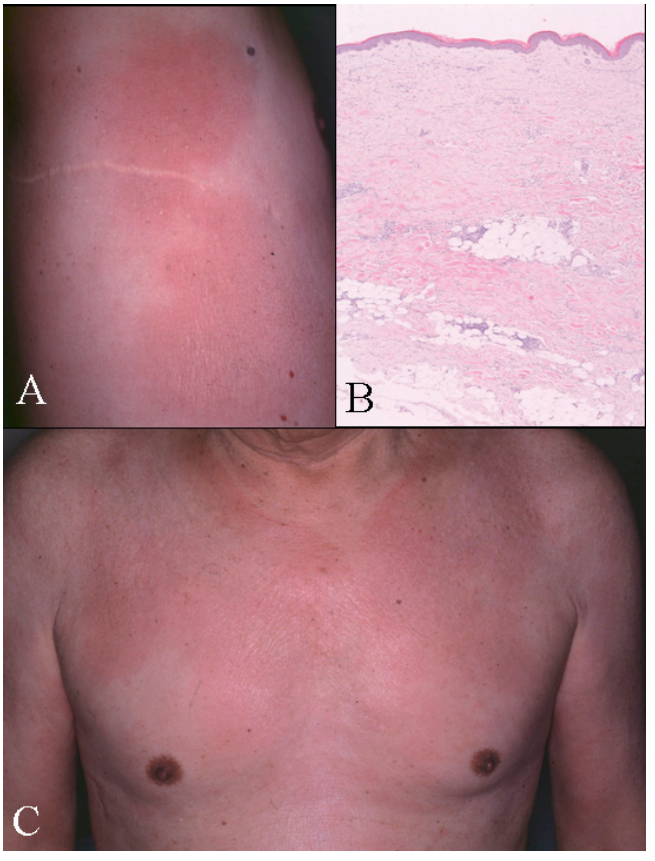

Fig. 1. (A) Erythematous sclerodermatous plaques localized on the chest of the patient in September 1996. (B) A skin biopsy from his trunk showed a thickening and homogenization of collagen bundles. (C) Sclerodermatous plaques recurred in January 2002.

In January 2002 he experienced sclerodermatous plaques on his chest and abdomen. The distribution was similar to that seen in 1996. A recurrence of morphea was diagnosed. Blood tests revealed the following abnormal values: ANA ×20 (speckled), KL6 552 U/ml, and CRP 0.69 mg/dl. A chest CT revealed lung fibrosis as had occurred before. Although inflammatory markers, such as CRP and ESR, were not extremely prevalent, the lesioned area of morphea was somewhat larger than before. The disease activity was evaluated as similar to

the previous episode, and he was subsequently treated with oral prednisolone 30 mg/day. He exhibited a positive response to this therapy for the sclerodermatous plaques. The prednisolone was decreased to 3 mg/day by the end of 2005.

Although no recurrence was found during a follow-up 24 months later, a recurrence of morphea on his trunk appeared suddenly in May 2008.

DISCUSSION

This case revealed sclerodactylia, lung fibrosis, reflux disease and morphea on the trunk. Morphea or localized scleroderma is categorized into the following major types; plaque morphea, GM and linear scleroderma (5). The morphea lesions in this present case seem to be consistent with GM. In GM, the plaques are commonly much larger than those seen in localized morphea, being many centimetres in diameter (1). They begin on the trunk and gradually increase in size, with the development of new plaques during the first year or two (1). Although Raynaud's phenomenon and the positivity of ANA were seen occasionally, there was no involvement of the internal organs such as the lungs or oesophagus (1). Because of this we diagnosed the present case as coexistence of SSc and morphea.

There have been several previous reports concerning the coexistence of SSc and morphea (2). Soma et al. (2) reported that nine (6.7%) of 135 cases of SSc had additional lesions of morphea, three of which exhibited multiple morphea lesions on their trunks. They concluded that morphea was one of the skin involvements of SSc, because a percentage of 6.7% was high enough to suggest such a relationship. Our case demonstrated that morphea activity was independent of the preceding SSc. Only the morphea lesion responded to systemic steroids and receded completely. When the morphea lesion recurred, other manifestations of SSc, such as lung fibrosis, sclerodactylia, and other abnormal serological findings, did not worsen. We thought that the morphea lesions presented in our case were not a skin manifestation of SSc.

The differential diagnosis of the present case included GM-like SSc. GM-like SSc has been described as revealing GM-like skin lesions and occasionally visceral changes of SSc (6). A correlation between its aetiology of SSc and exposure to organic solvent has been reported (6). In our case, the clinical course and occupational history excluded the possibility of this disorder.

An unusual manifestation of this case is the recurrence of morphea lesions. There have been only two reported cases of morphea recurrences similar to ours (7, 8). Mizutani et al. (7) reported a 45-year-old woman who had morphea lesions that recurred over 10 times in 6 years, and named this disorder "palindromic morphea". One or more morphea lesions appeared cyclically on various portions of her body and improved within 2 years. Oral corticoid therapy completely suppressed the recurrence. Fadilah et al. (8) reported another case, a 19-year-old male patient, who had experienced 30 recurrences of morphea lesions in 2 years, which were associated with extremely high eosinophil counts. The episode of morphea would last for one week and then resolve spontaneously. The recurrence of morphea in our case was less frequent than in the reported cases. This present case was similar in disease course to the former case (7).

In the case described here, prednisolone therapy for the early stages of palindromic morphea might suppress the recurrence, but decreasing the dosage to less than the minimum effective level may result in a recurrence. These findings suggest that the prevention of palindromic or recurrent morphea requires a maintenance dose of steroids for an extended period of time.

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