A Case of Atypical Localized Scleroderma Presenting with Pseudoainhum: Treatment with Tranilast, an Anti-fibrotic Agent

Sir,

Constricting bands of the extremities are usually classified into four groups: (1) true ainhum, (2) congenital bands, (3) ainhum-like bands associated with other diseases, and (4) bands secondary to trauma (1). True ainhum is characterized by a painful annular constriction of the fifth toe and occurs predominantly in Negro adults, with eventual spontaneous amputation (2, 3). Pseudoainhum is a term used by Neumann to distinguish the latter three groups from true ainhum and has numerous causes, including trauma (burns and frostbite), congenital anomalies, connective tissue diseases (scleroderma and Raynaud's disease), infections (syphilis and leprosy), diabetes mellitus, and keratoderma (keratoderma hereditarium mutilans, mal de Meleda, and pityriasis rubra pilaris) (2, 4, 5).

We report an unusual case of localized scleroderma of the bilateral forearms with clinical features resembling pseudoainhum. We have tried to treat the patient with tranilast, an anti-fibrotic agent.

CASE REPORT

A 63-year-old woman was referred to our dermatology clinic with skin lesions that had been present for 2 months. She had indurated erythematous and partly pigmented bands that symmetrically encircled the bilateral forearms and produced constriction. She complained of stiffness of the hands, but there were no manifestations suggestive of systemic scleroderma, including Raynaud's phenomenon, dysphagia, and dyspnea.

Laboratory tests revealed positivity for antinuclear antibody (× 160), but she was negative for anti-scl-70 and *Borrelia burgdorferi* antibodies. The complete blood count, liver function tests, and electrolytes were all normal, as were the findings on chest X-ray, CT scanning, and barium enema. Radiographs of the fingers revealed no bone absorption, and the radical artery pulse was normal. Histological examination of a biopsy taken from the left forearm revealed marked fibrotic changes in the entire dermis, which were consistent with the histological features of localized scleroderma. Direct immunofluorescence of the lesional skin was negative.

She was treated with tranilast at a dose of 300 mg daily. The lesions became soft after 8 months and the induration largely disappeared after 10 months. Rebiopsy of the left forearm after the 10 months of tranilast therapy revealed that the deep dermal fibrosis had disappeared and that slight fibrotic changes in the papillary layer remained.

DISCUSSION

Localized scleroderma is classified as linear, guttate, plaque-like, and generalized (6). However, the symmetrical lesions on the upper extremities observed in the present patient appear to be unique and do not belong to any of these groups. Although pseudoainhum associated with scleroderma is described in textbooks (2, 3, 7), the clinical details have never appeared in the literature.

Tranilast, N-(3,4-dimethoxycinnamoyl) anthranilic acid, has been used clinically as an anti-allergic drug. It inhibits passive cutaneous anaphylaxis in vivo and chemical mediator release from mast cells in vitro (8). Recently, it was found to selectively inhibit collagen accumulation in carrageenin-induced granulation tissue of rats (9) and to inhibit collagen synthesis as a pre-translational level in cultured fibroblasts from normal and diseased skin (e.g. keloid or scleroderma) (10). It has also been reported to improve the skin lesions of granulomatous diseases like granuloma annulare (11) or sarcoidosis (12). Evaluation of the response of localized scleroderma to tranilast is difficult, since this disease is sometimes self-limiting and shows spontaneous resolution. However, since all currently available therapies for localized scleroderma are only of limited value and since use as an anti-allergy drug has revealed no major side-effects of long-term tranilast administration, this agent may provide a possible therapeutic option for the treatment of localized scleroderma. It took a relatively long period (8 months) for remission of the lesions to occur, but this may not be unreasonable considering that the turnover of accumulated collagen is very slow.

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