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CLINICAL REPORT

Linear Scleroderma Along Blaschko's Lines in a Patient with Systematized Morphea

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We have previously shown that frontoparietal scleroderma en coup de sabre, a type of linear scleroderma that affects the face and scalp, follows the lines of Blaschko, but the question whether linear scleroderma that occurs in the limbs follows Blaschko's lines has not been answered. We describe the case of a 4-year-old girl with multiple morphea showing remarkable unilateral systematized distribution and whose linear lesions in the limbs appeared to follow Blaschko's lines. We suggest that linear scleroderma of the limbs, as well as frontoparietal scleroderma, may occur along the lines of Blaschko. Since both the unilateral distribution and the lesions along Blaschko's lines are the patterns created by genetic mosaicism, we suggest that a significant part of linear scleroderma and perhaps a smaller part of multiple morphea could be related to cutaneous mosaicism. ***Key words: Blaschko's lines; systematized morphea; linear scleroderma; mosaicism.***

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Localized scleroderma is a connective tissue disorder of unknown aetiology that is limited to the skin and the subcutaneous tissues underlying the skin lesions. Morphologically, it is classified within three variants: morphea, linear scleroderma and generalized morphea (1, 2). Linear scleroderma is a rare variant of localized scleroderma that is typically seen in the limbs as a paediatric disease (3). The linear distribution of the disease raises the question whether it follows the lines of Blaschko, but the question has never been answered. We describe a patient with multiple morphea showing predominant unilateral distribution and whose linear scleroderma in the limbs appeared to follow Blaschko's lines.

CASE REPORT

A 4-year-old girl presenting with a 1-year history of multiple sclerotic skin changes had sclerotic and hyperpigmented plaques on the left side of her chest, abdomen and back with remarkable unilateral distribution (Figs 1 and 2A). Severe skin sclerosis was also noted on her left buttock and her left leg, which was 1.5 cm shorter than her right leg. Linear sclerotic lesions extending down her left arm to the dorsal aspect of her left hand and her left index finger were causing flexion contracture (Fig. 2B). In addition, depigmented and slightly elevated sclerotic plaques were arranged in a linear distribution extending down her right posterior thigh to the calf (Fig. 2C). Routine blood test and urinalysis were normal. Autoantibody testing showed positive antinuclear antibody (1:320, speckled pattern) and negative rheumatoid factor, anti-single-stranded DNA antibody and anti-topoisomerase I antibody. A skin biopsy taken from her left buttock showed atrophy of epidermal appendages, thickened and hyalinized collagen bundles, and a mild perivascular lymphocytic infiltrate in the dermis. She was given a diagnosis of

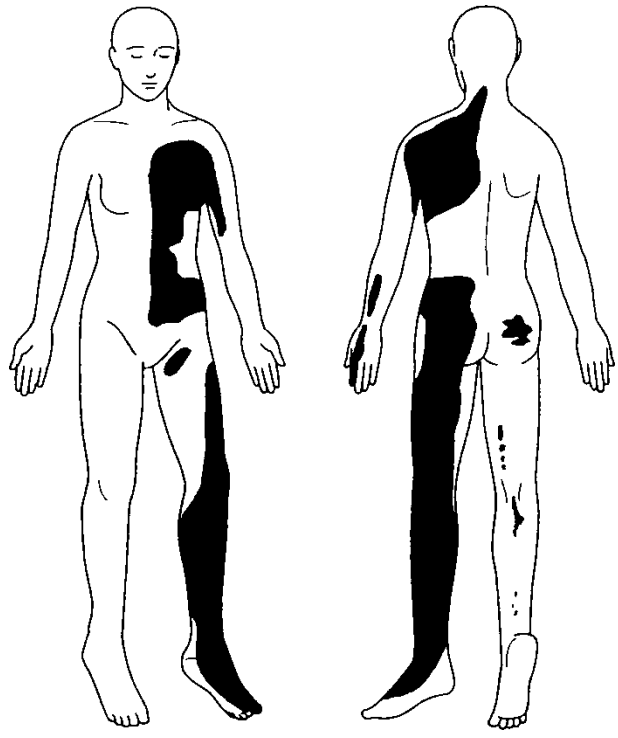

Fig. 1. Distribution of the sclerotic lesions.

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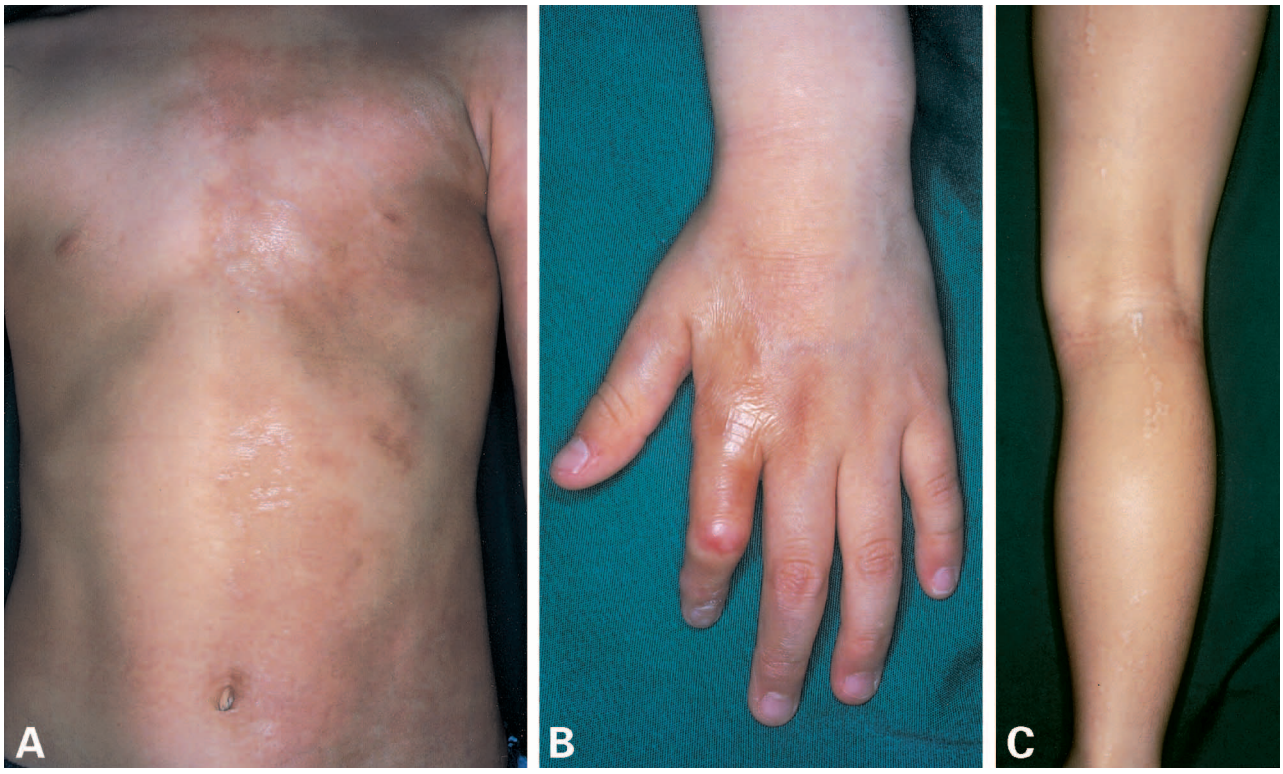

Fig. 2. A. Sclerotic and hyperpigmented plaques on the chest and abdomen (note the striking lateralization pattern). B. Severe skin sclerosis over the dorsal aspect of the left hand and left index finger with flexion contracture. C. Depigmented and slightly elevated sclerotic plaques arranged in a linear distribution extending down her right posterior thigh to the calf.

systematized morphea presenting with linear scleroderma of the limbs, and was treated with oral prednisolone, 15 mg per day, as well as topical corticosteroids. She responded satisfactorily to the treatment for the first month. Then the dose of prednisolone was gradually tapered and maintained at 2.5 mg per day without any signs of adverse effects.

DISCUSSION

Blaschko's lines determine the distribution pattern of many congenital and acquired skin diseases, such as epidermal naevi, sebaceous naevi, incontinentia pigmenti, linear lichen planus and lichen striatus (4, 5). They are sometimes mixed-up with the lines of dermatomes, the segments of skin that are defined by sensory innervation, but a close comparison reveals that Blaschko's lines cannot be related to sensory nerves (4). Although the exact nature of Blaschko's lines is unknown, there is a general consensus that they reflect the lines of embryonal development of the epidermis and epidermal derivatives, and may be produced by genetic mosaicism (6). Supporting this hypothesis, authentic genetic mosaicism has been proven in some congenital and acquired skin conditions that follow Blaschko's lines, including a type of epidermal naevus (7), unilateral palmoplantar verrucous naevus (8), segmental Darier's disease (9) and "Blaschkitis" (10).

The question whether linear scleroderma follows Blaschko's lines is controversial. An earlier study suggested a dermatomal distribution of the disease (11), and in a review published in 1994 it was noted that there have been no examples of patients with linear scleroderma along the lines of Blaschko (5). In 1996, however, Hauser et al. (12) described a patient with multiple morphea arranged along Blaschko's lines and discussed the close relation between morphea and linear atrophoderma of Moulin, which is also known to follow Blaschko's lines. In 1998, we first showed that frontoparietal scleroderma (en coup de sabre), a type of linear scleroderma that affects the face and scalp, follows the lines of Blaschko (13), and this idea has been supported by later observations (14–16). Our next interest was linear scleroderma of the limbs. It seemed difficult for us to determine whether linear scleroderma of the limbs follows Blaschko's lines, since the arrangement of Blaschko's lines in the extremities means that it is hard to make a distinction from a dermatomal distribution. In addition, lesions of linear scleroderma in the limbs are usually wide and discontinuous, making it hard to judge which lines are followed by linear scleroderma.

The patient presented here had widespread skin lesions on the trunk and extremities with typical clinical

and histological features of localized scleroderma. Then we diagnosed systematized morphea in the patient. Sclerotic plaques were linearly arranged in the left arm and the right leg. The unusually sharp line formed by the lesions in the right leg (Fig. 2C) reminds us of the lines of Blaschko, rather than the dermatomal distribution. We recently observed two children with lichen striatus on the leg (photographs not shown), whose lesions distributed in a pattern strikingly similar to the one drawn by the lesions shown in Fig. 2C. Since it has been repeatedly proven that lichen striatus follows the lines of Blaschko (4, 5), we concluded that linear scleroderma in the right leg of our patient occurred along Blaschko's lines. The lesions in her left upper limb may also be along Blaschko's lines, but because they are discontinuous and relatively wide it may be difficult to determine whether they follow Blaschko's lines or another pattern of mosaicism.

Another important feature of our patient is the remarkable unilateral distribution. Very recently, Nagai et al. reported a 6-year-old boy with generalized morphea showing remarkable unilateral distribution (17). A similar lateralization pattern is often observed in various naevoid conditions, which sometimes in part follow Blaschko's lines (4, 5). In CHILD syndrome, a striking lateralization pattern and lesions along the lines of Blaschko are considered the manifestation of functional X-chromosome mosaicism (6, 18). Thus the unilateral distribution seen in our patient may well be explained by hypothesizing a mosaic state.

Both linear scleroderma of the limbs and frontoparietal scleroderma en coup de sabre appear more commonly in children than in adults with various immunological abnormalities (3). Our observations shown in the present study suggest that a significant part of linear scleroderma of the limbs, as well as frontoparietal scleroderma, and maybe a smaller part of multiple morphea, could be related to cutaneous mosaicism. In patients with linear scleroderma, we speculate that genetically susceptible cells are arranged along Blaschko's lines, and subsequent exposure to an environmental trigger may result in the development of linear scleroderma.

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