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Case Presentation

Extraordinarily long linear cutaneous lupus erythematosus along the lines of blaschko

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Abstract

Linear cutaneous lupus erythematosus (LCLE) is a rare subtype of cutaneous lupus erythematosus. We describe a 22-year-old Japanese man who had an 11-year history of asymptomatic linear erythema from the right upper back to the dorsum of the right hand. The lesions followed the lines of Blaschko and spread over three large joints. Histological findings were compatible with discoid lupus erythematosus. Although the most common site for LCLE is the face, a few cases of LCLE on the extremities have been reported. To our knowledge, this is the first reported case of an extraordinarily long, continuous LCLE skin lesion.

Case synopsis

Linear cutaneous lupus erythematosus (LCLE) is a rare subtype of cutaneous lupus erythematosus. The term LCLE was proposed by Abe *et al* in 1998 [1]. LCLE develops linear lesion following the lines of Blaschko, although its etiology remains unknown. Histological findings are compatible with discoid LE (DLE) according to previous reports [1-4]. We describe a case of LCLE, which was distributed as a long constitutive line that crossed three large joints. This Japanese man noted the disease onset in his childhood. Such long skin lesions of linear cutaneous LE strongly connects this disorder with the lines of Blaschko.

A 22-year-old Japanese man presented with an 11-year history of asymptomatic linear erythema that had appeared on the right upper arm. The lesion expanded gradually to the right shoulder and the dorsum of the right hand. There was no history of trauma around the lesions, nor of intensive exposure to sunlight. He had neither photosensitivity nor family history of any skin disease. Scaly erythema with partly whitish atrophy were seen from the right upper back to the right hand (Fig. 1A-C). The lesions followed the lines of Blaschko [5] (Fig. 1D). Laboratory examination, including complete blood count, liver function test, blood urea nitrogen test, and urinalysis, showed no abnormalities. Anti-nuclear antibodies were positive with a homogeneous pattern (1:80). Other autoantibodies, including anti-SS-A, Sm, U1-RNP, ds-DNA, ss-DNA, and cardiolipin were all negative. Serum complement levels were normal.

Histological examination of the erythema reveals hyperkeratosis and liquefaction degeneration of the basal layer (Fig. 2A and B). In the dermis, dense lymphocytic infiltrates are observed in the perivascular and periadnexal areas. Thickening of the epidermis basement membrane is revealed by periodic acid-Schiff staining (Fig. 2C) and mucin deposition in the reticular dermais is confirmed by colloidal iron staining (Fig. 2D). These findings are compatible with discoid LE.

We started treatment with topical corticosteroid (Betamethasone butyrate propionate 0.05%) and continued follow up. There has not been improvement.

Discussion

LCLE is a rare subtype of cutaneous LE and more than a dozen cases have been reported [1-4]. The term LCLE was proposed according to the finding that the clinical morphology is linear, the lesions are limited to the skin, and there is no systemic involvement. LCLE develops as linear lesions following the lines of Blaschko, which was descrived by Alfred Blaschko in 1901 [5]. The lines represent pathways of epidermal-cell migration and proliferation during fetal development and reflect the existence of cutaneous mosaicism. The skin lesions of some nevoid skin diseases or inflammatory skin diseases have linear distributions following these lines [6]. In our case, the lesions seem to match the line patterns of Blaschko, which formed slightly curved lines from back to shoulder and almost straight lines or partly S-shape lines on the arm.

In most cases with LCLE, the age at onset is under 15 years. Predominance in females is recognized, without ethnic preference. It shows a predilection for the face and less frequently occurs on the limb and trunk. Most commonly, anti-nuclear antibodies are negative or slightly positive. Neither photosensitivity nor progression to systemic LE is observed. The histological findings in LCLE include hyperkeratosis, atrophy of the epidermis, hydropic degeneration of the basal cell layer in the epidermis, perivascular and periadnexal dense infiltrates of lymphocytes, and mucinous deposition in the dermis. These findings are compatible with DLE. Our patient was first diagnosed with lichen striatus owing to the clinical morphology. He was finally diagnosed as having LCLE on the basis of the histological findings. Diaminodiphenyl sulfone, topical steroids, or chloroquine are used for treating LCLE. Of these, however, Japanese medical insurance covers only topical steroids. Unfortunately, in our case, no significant improvement has been found to date.

Although the affected site of LCLE is most commonly face, to our knowledge, a few cases with LCLE that occurred on the extremities have been reported. Umbert and Winkelmann reported a patient with concurrent localized scleroderma and DLE at her forearm [2]. In the case reported by Heid E *et al* [7], the patient was a 35-year-old man who presented with a unilateral skin eruption from the forearm to the chest. In Daldon's case [8], the patient was a 15 year-old boy presenting with discoid erythematous lesions from the right hand to the right paravertebral area. Röckmann *et al* reported a 42-year-old woman who had erythemateous plaques on her right trunk and leg [4]. Our patient presented with linear erythematous lesions from the right upper back to the dorsum of hand, extending across three large joints. Such long skin lesions of LCLE has not been reported so far.

The linear lesions of LCLE may be caused by genetic mosaicism of keratinocytes in the lines of Blaschko combined with some external factors (sun exposure or viral infection). Recently, it is supposed that keratinocyte apoptosis is related to genetic alteration involving some apoptotic pathways and this may be an important factor for initiation of LCLE lesions [9].

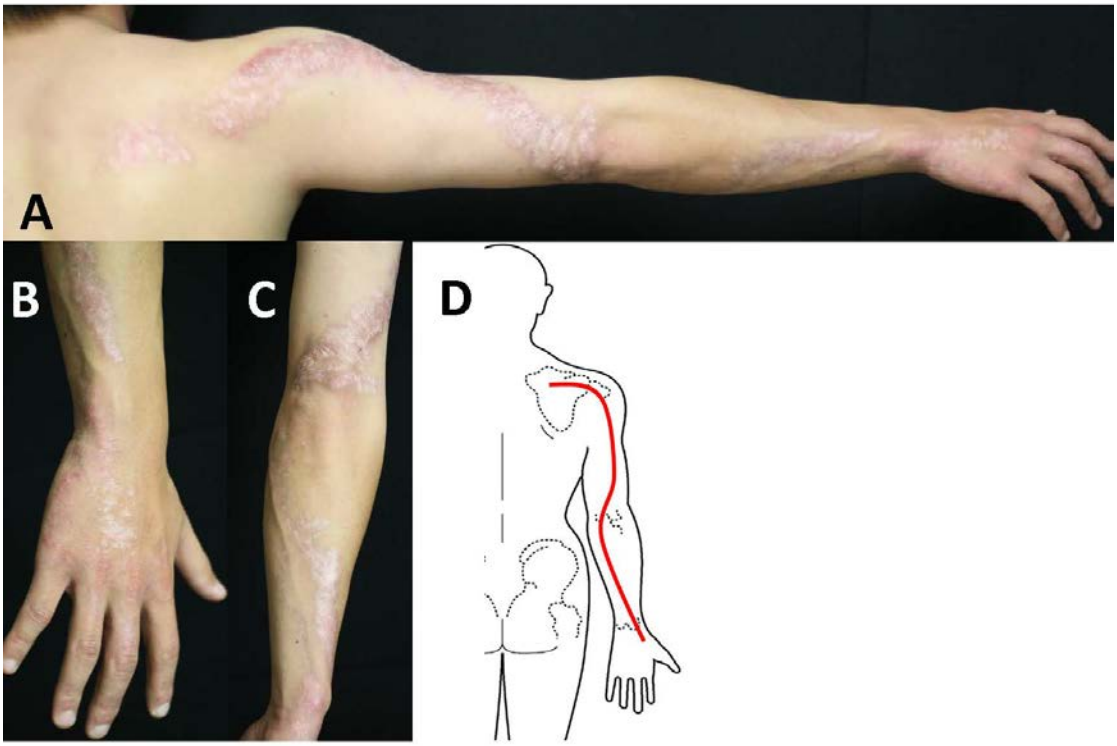

Figure 1. (A-C) Linear erythema with partly white atrophy and scales are evident from the right upper back to the right hand. (D) The lesions follow the lines of Blaschko.

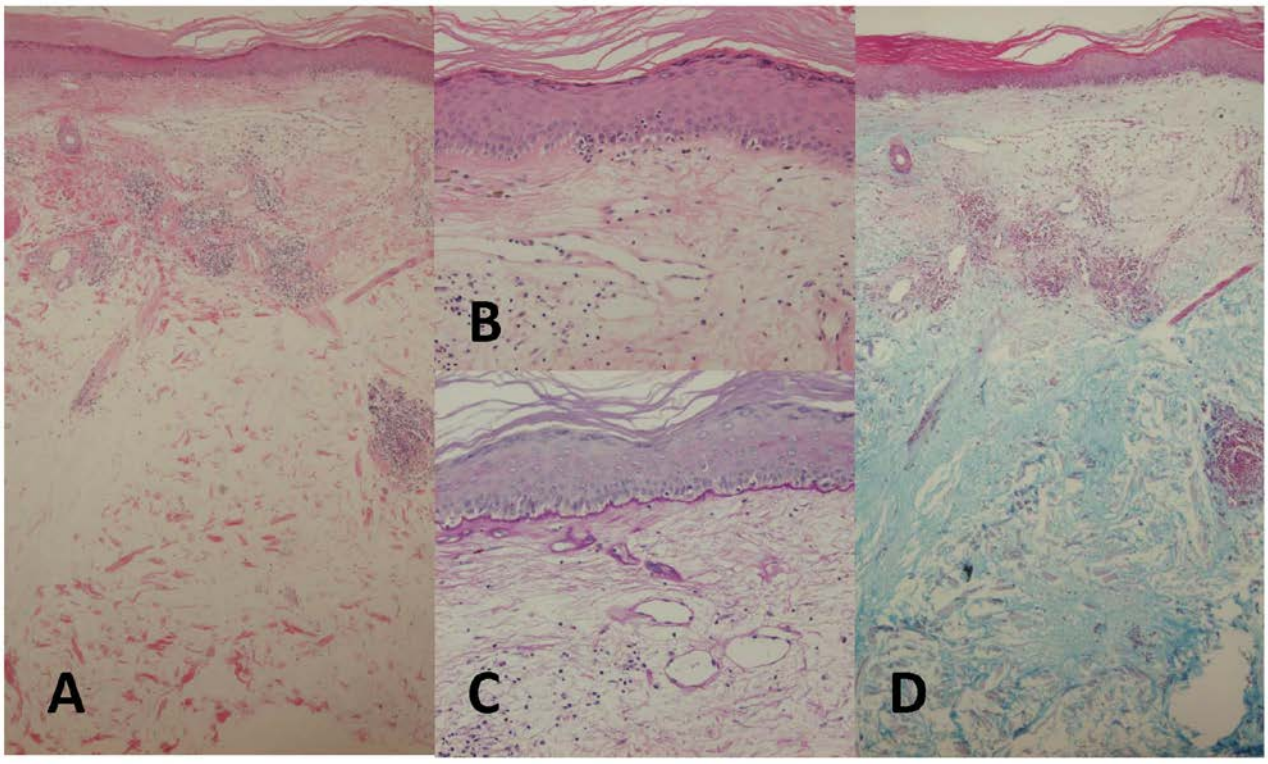

Figure 2. (A, B) Skin biopsy specimens of the erythematous area on the right upper arm. In the epidermis, hyperkeratosis and liquefaction degeneration of the basal layer. In the dermis, dense lymphocytic infiltrates were observed in the perivascular and periadnexal areas. (C)

Periodic acid-Schiff staining shows thickening of the epidermis basement membrane. (D) Colloidal iron staining reveals mucin deposition in the reticular dermis.

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