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CASE REPORT

A patient with subacute cutaneous lupus erythematosus along Blaschko lines: Implications for the role of keratinocytes in lupus erythematosus

Sebastian Yu [1, 2], Hsin-Su Yu [1, 3, *]

[1] Department of Dermatology, Kaohsiung Medical University Hospital, and Department of Dermatology, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan
[2] Graduate Institute of Clinical Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan
[3] National Institute of Environmental Health Sciences, National Health Research Institutes, Miaoli County, Taiwan

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ABSTRACT

Blaschko lines are manifestations of cutaneous mosaicism, with apparent contrast between normal cells and abnormal cells, which carry postzygotic mutations. Linear cutaneous lupus erythematosus is a rare subset of cutaneous lupus erythematosus (CLE), characterized by skin lesions along Blaschko lines. In this paper, we presented a 47-year-old female with subacute CLE lesions along Blaschko lines. Although the antinuclear antibody and anti-Ro titers were remarkably high, there were no systemic symptoms. The histopathology showed atrophic epidermis with vacuolization of basal keratinocytes and papillary edema. While no typical linear lupus band was found, the direct immunofluorescence revealed cytoid bodies with immunoglobulin G, immunoglobulin M, C3, and C4 depositions in the papillary dermis. The skin lesions completely resolved after topical tacrolimus treatment, and the clinical course was 10 months without recurrence. We reviewed the literature and summarized that CLE along Blaschko lines supports the functional role of Ro antigens and keratinocytes in the development of CLE.

Introduction

Linear cutaneous lupus erythematosus (LCLE) is a rare subset of cutaneous lupus erythematosus (CLE), characterized by skin lesions that follow Blaschko lines.[1] However, there are a variety of descriptions of lupus erythematosus (LE) lesions in these cases, including discoid LE and LE profundus among others. Although there is one case with other organ involvement that fulfilled the diagnostic criteria of systemic lupus erythematosus (SLE),[2] cutaneous lesions are the only manifestation in most of the LCLE cases. In this paper, we presented a female patient with LCLE. We discussed the origin of Blaschko lines and summarized that CLE along Blaschko lines supports the functional role of keratinocytes in the development of CLE.

Case report

A 47-year-old female presented with a 2-month history of scattered pigmented macules over her right waist. The lesions were neither pruritic nor painful. She had neither trauma history nor associated symptoms. She had no known underlying systemic disease except thalassemia. A complete blood count showed microcytic anemia, compatible with thalassemia. The antinuclear antibody (ANA) was weakly positive (1:40). One month later, however, the pigmented lesions became scaly and papular, progressing toward the right lower extremity. The lesions were arranged in arch shapes on the right waist (Figure 1A) and in linear configuration on the right lower extremity (Figure 1B). A skin biopsy was performed. The histopathology showed atrophic epidermis with vacuolization of basal keratinocytes and papillary edema. Scattered dyskeratotic cells in papillary dermis were also found. There was inflammatory lymphocytic infiltration around the vascular structure and

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* Corresponding author. National Health Research Institutes, 35, Keyan Road, Zhunan Town, Miaoli County 35053, Taiwan.
E-mail address: yup.kmu@gmail.com (H.-S. Yu).

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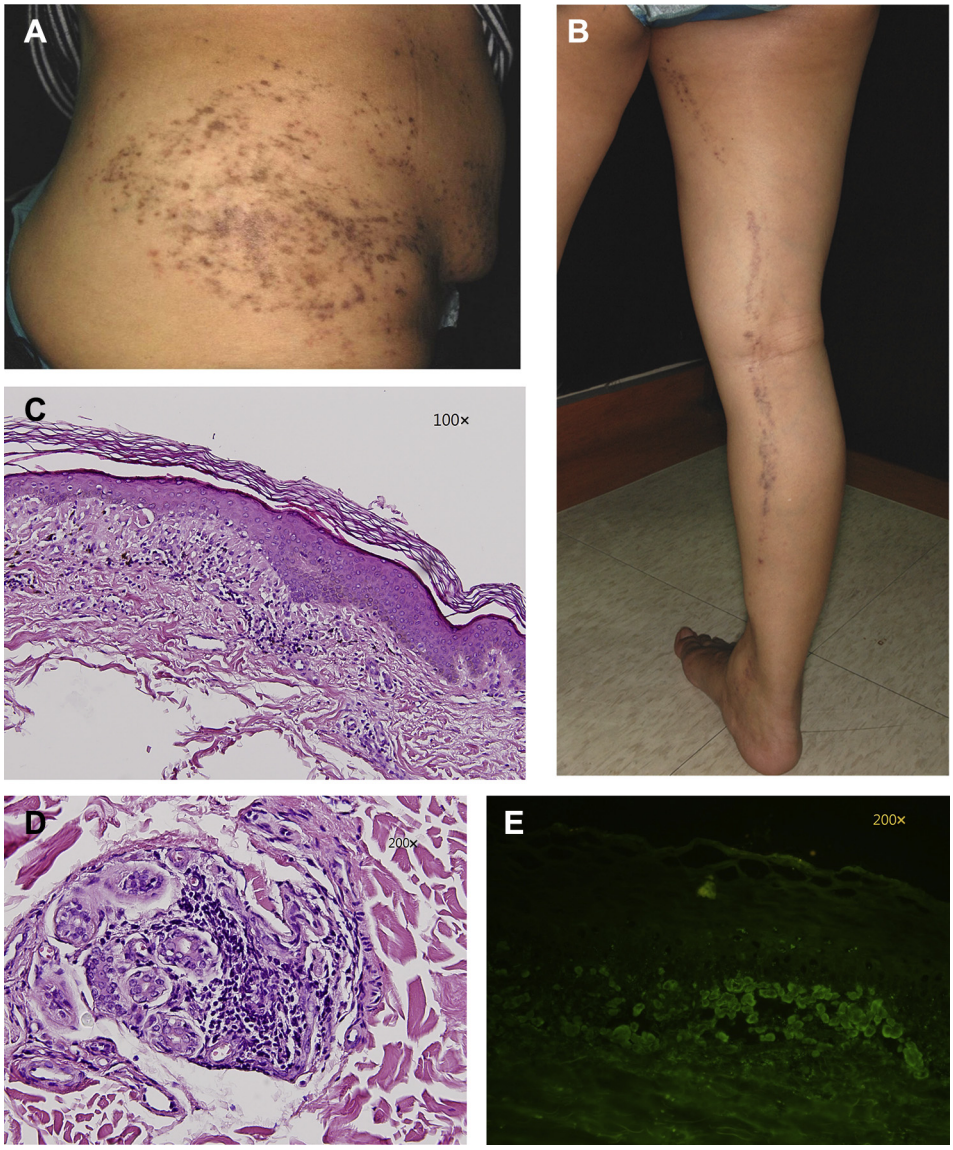

Figure 1 The pigmented papular scaly lesions on the right abdomen and right leg. (A) The figure shows lesions were arranged in arch shapes on the right side of the waist; (B) the figure shows lesions were in linear configuration on the right lower extremity; (C) the figure shows histopathology at the junction between the lesional (left side) and perilesional skin (right side). In the lesional skin, there was atrophic epidermis with vacuolization of basal keratinocytes and papillary edema. Scattered dyskeratotic cells were also found in papillary dermis; (D) the figure shows there was periadnexal lymphocytic infiltration; and (E) the figure shows cytoid bodies of immunoglobulin G in the papillary dermis in direct immunofluorescence examination.

melanophages in the upper dermis (Figure 1C). Periadnexal lymphocytic infiltration was also observed (Figure 1D). The histopathologic findings were compatible with LE.[3,4] Direct immunofluorescence revealed cytoid bodies with immunoglobulin G, immunoglobulin M, C3, and C4 depositions in the papillary dermis (Figure 1E). ANA was followed up and showed >1:1280 positive. Other autoantibody surveys showed anti-Ro positive (80.6 EliAU/mL, normal <7), anti-La negative, antiphospholipid immunoglobulin M and immunoglobulin G negative, and rheumatoid factor negative. The viral-infection survey showed negative for herpes simplex virus, varicella-zoster virus, and Epstein–Barr virus. Based on the clinical manifestation, histopathology, and autoantibody findings, subacute cutaneous lupus erythematosus (SCLE) along Blaschko lines was diagnosed. Because there were no systemic symptoms, topical treatment with 0.1% tacrolimus ointment was prescribed without oral medication. After a 5-month topical tacrolimus treatment, the skin lesions gradually resolved (Figure 2). The autoantibody survey was followed up again and showed ANA >1:1280 positive. The anti-Ro titer slightly decreased (51.3 EliAU/mL, normal <7) as compared to the previous anti-Ro titer. The topical treatment was continued, and the skin lesion resolved with complete clearance. No scaring or skin atrophy was noted. The total clinical course was 10 months.

Discussion

The first case of discoid LE with linear distribution along Blaschko lines was reported by Umbert and Winkelmann in 1978.[5] In 1998, the term LCLE was proposed by Abe et al[1] to describe discoid LE with linear configuration following Blaschko lines. From then on, several papers reported patients with LE lesions along Blaschko lines. In addition to linear discoid LE, some cases were diagnosed as linear LE profundus,[6–9] and one was diagnosed as linear tumid LE.[10] In 2006, Röckmann et al[11] reported a female with SCLE on Blaschko