Case Report

Nonisomorphic and Nonisotopic Multidermatomal Discoid Lupus Erythematosus – A Rare Subtype of Cutaneous Lupus Erythematosus

Abstract

Discoid lupus erythematosus (DLE) is the most common variant of chronic cutaneous lupus erythematosus. Several cases of Blaschko-linear lupus erythematosus, which is regarded as a distinct subtype of lupus erythematosus, have been reported across the globe. Dermatomal involvement in DLE is rare and that too has been reported in patients of systemic lupus erythematosus at site of healed herpes zoster either as an isomorphic or isotopic phenomenon. Here, we report a case of DLE on account of its unusual morphology, de novo multidermatomal configuration, without previous episode of herpes zoster or trauma.

Keywords: *De novo, discoid lupus erythematosus, isomorphic, isotopic, multidermatomal*

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Introduction

Discoid lupus erythematous (DLE) is the most common variant of chronic cutaneous lupus erythematosus.[1] Lesions of DLE can present in a linear configuration, which is often referred as linear cutaneous lupus erythematosus.[2] Cutaneous lesions of lupus erythematosus, distributed along the lines of Blaschko, i.e., Blaschko-linear lupus erythematosus, have been frequently reported.[3] Dermatomal involvement in DLE is rare and that too has been reported in patients of systemic lupus erythematosus (SLE) at site of healed herpes zoster either as an isomorphic or isotopic phenomenon.[4-6] Here, we report a case of DLE on account of its unusual morphology, de novo multidermatomal configuration, without previous episode of herpes zoster or trauma.

Case Report

A 37-year-old female presented with multiple hyperpigmented skin lesions over right side of face for 1 and half months. Lesions were associated with mild itching. There was no history of photosensitivity, trauma at the site of lesions, or any other dermatological disorder in the past. On examination, multiple, well-defined, unilateral, violaceous plaques were noticed in a dermatomal configuration involving right V3 (mandibular division of trigeminal nerve) and C3 dermatome [Figure 1a and b]. No contributory systemic findings were noted.

On the basis of these findings, zosteriform lichen planus was thought as the clinical diagnosis. A punch biopsy was taken from one of the representative lesions. On histopathological examination, epidermis showed orthokeratosis, follicular plugging, atrophy and basal vacuolar degeneration, whereas dermal findings included perivascular lymphomononuclear cell infiltrate and marked pigment incontinence. Periodic acid Schiff (PAS) stain showed faint thickening of the basement membrane and alcian blue stain showed mucin deposition in the dermis [Figure 2a-c].

On the basis of above histopathological findings, we revised our diagnosis as multidermatomal DLE. Anti-nuclear antibody and anti-ds DNA were negative. Baseline investigations, including complete blood count, erythrocyte sedimentation rate, urine examination, liver and renal function tests were within normal limits.

Patient was started on topical corticosteroids, sunscreen, and oral hydroxychloroquine. She improved after 1 month of therapy, with residual post-inflammatory hyperpigmentation [Figure 3].

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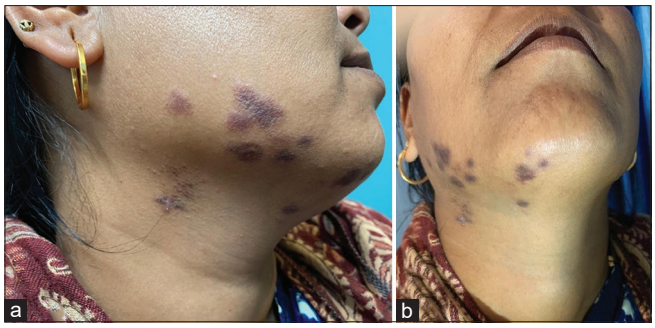

Figure 1: (a and b) Multiple, well-defined hyperpigmented plaques over right V3 (mandibular division of trigeminal nerve) and C3 dermatome

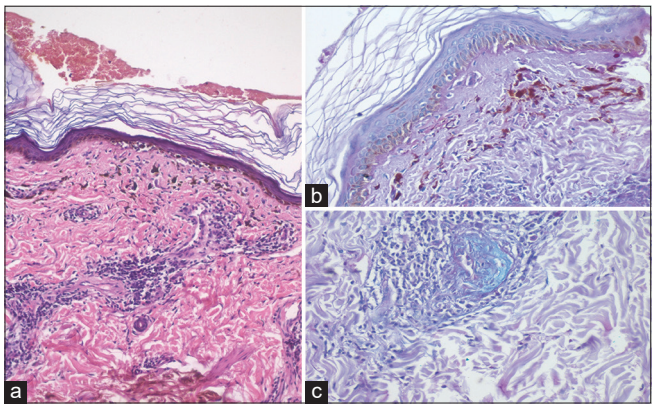

Figure 2: (a) Orthokeratotic hyperkeratosis, flattening of rete ridges, basal vacuolar degeneration, lymphomononuclear cell infiltrate, and marked pigment incontinence. (H and E, 10X). (b) Thickening of the basement membrane and marked pigment incontinence. (PAS, 40X). (c) Mucin deposition in the dermis (Alcian blue, 40X)

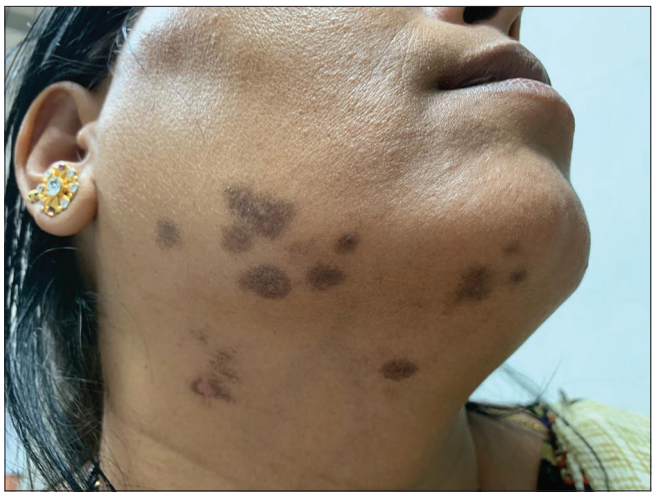

Figure 3: Postinflammatory hyperpigmentation after 1 month of therapy

Discussion

Cases of DLE with dermatomal involvement have been rarely reported in literature. To the best of our knowledge, only three cases have been reported so far. In all these cases, lesions of DLE were noted over a particular dermatome at the site of healed herpes zoster in patients who had already been suffering from SLE.[4-6] This phenomenon has been described as an isomorphic or isotopic response[4-6] and the proposed mechanisms include altered immunity, neuronal damage, or vulnerable connective tissue as a result of varicella zoster viral infection.[6] Our case presented with multiple violaceous plaques involving right mandibular and submandibular region suggestive of V3 (mandibular division of trigeminal nerve) and C3 dermatome. We report this unique case of DLE on account of its de novo, multi dermatomal presentation with no history of pre-existing dermatosis, or trauma at the site of lesions and without any evidence of systemic involvement. The exact pathogenesis of dermatomal lesions in the present case is not certain, but it could be due to some unknown neural factor in a background of genetic and immunological predisposition. The Blaschko-linear distribution follows the lines of Blaschko, but in dermatomal configuration, lesions follow cutaneous innervation. On the basis of these features, it is plausible that such cases of DLE with dermatomal involvement reflect a rare, distinct clinical subset of cutaneous lupus erythematosus.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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