Chronic Cutaneous Lupus Erythematosus in a Blaschkoid Pattern over Face
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Abstract
Linear lesions following the lines of Blaschko of chronic cutaneous lupus erythematosus is a highly unusual variant of cutaneous lupus, particularly in adult. The lesions are restricted to one anatomical site with facial predilection in most of the reported cases. We herein, report a case of chronic cutaneous lupus erythematosus due to rarity of presentation and later age of onset.

Key Words: Blaschkoid pattern, chronic cutaneous lupus erythematosus, face

Introduction
Chronic cutaneous lupus erythematosus is a well-known entity and is frequently encountered in dermatology outdoor; however, lesions distributed along the lines of Blaschko is a very rare phenomenon. Linear cutaneous lupus erythematosus is mostly reported in children and young adults, with facial predilection. Individual lesions exhibit characteristic clinical and histopathological findings of discoid lupus erythematosus.\(^1,2\) The first case of discoid lupus erythematosus with linear distribution along Blaschko lines was reported by Umbert and Winkelmann in 1978.\(^3\) In 1998, Abe et al. coined the term “linear cutaneous lupus erythematosus”.\(^1\) “Blaschkoid cutaneous lupus erythematosus” is the preferred terminology over “linear cutaneous lupus erythematosus” as linear configuration is seen in Blaschkoid as well as dermatomal lesions.\(^4\)

Herein, we report a case of chronic cutaneous lupus erythematosus in an adult male to highlight the unusual clinical presentation of this disorder.

Case Report
A 57-year-old male presented with multiple, mildly itchy, and depigmented elevated skin lesions arranged in a linear configuration affecting the right side of the face. The lesion started as barely appreciable skin problem over forehead and gradually progressed to the present dimensions over a 5-year duration. Opposite side of the face and rest of the body were unaffected. No history of preceding trauma or exposure to excessive sunlight was present. Cutaneous examination revealed two well-defined, scaly, depigmented, and atrophic plaques with hyperpigmented borders. One lesion with dimensions of 2 cm × 4 cm was located over the right temple and another of 3 cm × 5 cm extending medially from bridge of the nose to the right cheek [Figure 1a]. Our patient responded well to topical mometasone 0.1% cream twice daily application [Figure 1b]. Routine hematological and biochemical investigations were normal, and antinuclear antibody (ANA) was negative. Histopathologic examination revealed hyperkeratosis with follicular plugging, thinning and flattening of epithelium, and hydropic degeneration of the basal layer of epidermis. Scattered apoptotic keratinocytes in the basal layer and wavy thickening of basement membrane were noted [Figure 2a]. Dermis exhibited patchy lymphocytic infiltrate with accentuation of the pilosebaceous follicles, interstitial mucin deposition, and edema [Figure 2b]. Topical mometasone 0.1% cream was recommended twice daily.

Discussion
Linear involvement in cutaneous lupus erythematosus along the lines of Blaschko is extremely rare. Here the lupus erythematosus lesions follow the lines of Blaschko, which represents the pathways of epidermal cell migration...
and proliferation during fetal development and reflects the existence of genetic mosaicism of keratinocytes. 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 linear cutaneous lupus erythematosus lesions.[5]

Blaschkoid cutaneous lupus erythematosus, apart from discoid lupus erythematosus, might be the presenting feature of bullous lupus erythematosus, tumid lupus erythematosus, or subacute lupus erythematosus.[4]

Most of the reported cases were children with only few cases described in adults.[6,7] It has been seen that Blaschkoid cutaneous lupus erythematosus has predilection for face and less frequently can involve the limbs and trunk.[8] In majority of the cases, only a single anatomical site was affected. Our patient also had involvement of face. Neither photosensitivity nor progression to systemic lupus erythematosis was observed. Antinuclear antibodies are usually negative or slightly positive in such cases.[9] Our case was ANA negative.

Histopathological features of Blaschkoid cutaneous lupus erythematosus are compatible to that of discoid lupus erythematosus, such as hyperkeratosis, atrophy of the epidermis, hydropic degeneration of the stratum basale of epidermis, dense perivascular, and perineural lymphocytic infiltration, and mucin deposition in the dermis. Several treatments have been used including topical corticosteroids and systemic treatments such as hydroxychloroquine and dapsone. Improvement or complete resolution was obtained in most of the cases.[4]

**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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**Conflicts of interest**

There are no conflicts of interest.

**What is new?**

Linear involvement in cutaneous lupus erythematosus along the lines of Blaschko is extremely rare, especially in adult.

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