Facial Blaschkitis: case and review

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Abstract

We report on a 15-year-old female with a 3-month history of a pruritic, erythematous cutaneous eruption on the right cheek and perioral area. The lesion had a linear distribution following the lines of Blaschko. Histopathological findings and direct immunofluorescence were compatible with chronic cutaneous lupus erythematosus (LE). Treatment with topical steroids and systemic antimalarial agents over 2 months showed hardly any improvement contrary to similar cases reported in the literature in the past. Histological findings move this case close to LE. However, the unusual clinical presentation as well as the resistance to antimalarial drugs do not fully allow to confirm this suspicion. Therefore, we recommend to call this new entity LE-like facial Blaschkitis of the adult.
Key Words
Blaschkolinear dermatosis · Blaschkitis · Linear cutaneous lupus erythematosus

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Case Report
A 15-year-old female presented with a 3-month history of pruritic lesions on the right upper cheek and perioral area. Only cradle cap and dermatitis on the upper extremities were noted in her medical history. Topical steroids, antiviral and antibacterial agents had no effect. Papules, milia, and several small erythematous to livid plaques on the right cheek in a Blaschko linear pattern were found (fig. 1a, b). The differential diagnosis included hamartoma or an inflammatory dermatosis.

A skin biopsy showed thin epidermis with follicular hyperkeratosis and vacuolization of the dermoepidermal junction with apoptotic keratinocytes basally. Predominantly lymphocytic cells grouped densely around adnexal and vascular structures (fig. 1c, d). Direct immunofluorescence revealed granular deposits of IgM, IgG and C3 at the dermoepidermal junction. Laboratory workup including autoantibodies was unremarkable except for slightly elevated bilirubin levels judged irrelevant. A diagnosis of facial blaschkitis with histopathological findings of linear lupus erythematosus (LE) was made.

Worsening occurred on topical pimecrolimus q.d. with hydroxychloroquine 200 mg b.i.d. p.o. for 2 months. Isotretinoin 20 mg q.d. p.o. for 2 months had no effect. Intense pulsed light (Palomar LuxG 500–670 and 870–1,400 nm, 18–38 J/cm², 5–20 ms pulse length, 2 passes, 2 sessions) resulted in a partial improvement (fig. 2).

Discussion
At the beginning of the 20th century, Alfred Blaschko noticed a special linear disposition of certain congenital and hereditary dermatoses, and described cutaneous lines different from others such as dermatomes, Langer’s lines and Voigt’s lines.

On the face, Blaschko’s lines form an arch shape from the oral angle to the orbital region, and an L shape from the oral angle to the neck and from the preauricular area to the neck (fig. 3) [1].

Most linear dermatoses that follow the lines of Blaschko are either X-linked disorders or congenital and/or nevoid disorders [2]. Most of the acquired dermatoses that present in a blaschkolinear pattern are common dermatoses that are usually distributed in a random fashion. When lesions happen to be distributed along the lines of Blaschko, they exhibit the same clinical and histological picture as lesions of the more common distribution pattern. This occurs in psoriasis, lichen planus, or atopic dermatitis. A few acquired dermatoses always present in a linear pattern with their own clinical and histological hallmarks, such as lichen striatus, blaschkitis, or Moulin disease [3].

Blaschkitis is defined as a spongiotic dermatitis presenting with multiple re-
Fig. 1. a, b Linear erythematous, nonscaly, papular lesions along the line from the right lower eyelid to the cheek and the line parallel to the right upper lip. c, d Thin epidermis with vacuolization of the dermoepidermal junction and some apoptotic keratinocytes in the basal layer. Dense inflammatory infiltrates of predominantly lymphocytic cells around the adnexal and vascular structures in the underlying dermis. Hematoxylin and eosin; magnification ×100.

Fig. 2. Disappearance of the papules and brightening of the affected area after treatment with intense pulse light in two sessions (LuxG 500–670 and 870–1,400 nm, 18–38 J/cm², 5–20 ms pulse length, 2 passes).

Fig. 3. Blaschko lines on the face (reproduced from Bolognia et al. [1]).
lapsing itching papules and vesicles following the Blaschko lines especially on the trunk, with a preference for males in their forties [3], and only rarely occurring in children [4]. Lesions disappear spontaneously within days to weeks and may relapse several times for months and years [5]. Grosshans-Marot disease is distinct from blaschkolinear variants of known dermatoses, hamartoma, or X-linked dominant gene defects [6].

Blaschkolinear lesions probably reflect a mosaic condition from postzygomatic somatic mutation or lyonization (the inactivation of an X chromosome) [7]. Thus, cutaneous antigenic mosaicism, e.g. induced by a viral infection, could trigger a strictly localized T cell response [5]. This hypothesis relates blaschkitis to other cutaneous autoimmune diseases, as do antinuclear antibodies [6].

A few cases of linear configuration of LE have been reported in the literature. According to the analysis of 14 cases diagnosed as linear cutaneous LE (linear cutaneous LE, a term proposed for discoid LE with a linear configuration [8]), the mean age of onset of linear cutaneous LE was 15.8 years with a range of 3–42 years [9]. A case published by Mi-Woo et al. [10] in 2001 also showed linear lesions following the Blaschko lines on the cheek and peri-oral area. All cases presented in the literature except 1 with a systemic LE were well controlled with hydroxychloroquine, dapsone or topical corticosteroids.

The present case reports a blaschkolinear inflammation in early adulthood diagnosed as facial blaschkitis with histopathological findings of linear LE. On the basis of anatomical and clinical criteria this entity is distinct from dermatoses such as lichen planus or LE. In contrast to our observation, most published cases showed spongiform dermatitis histologically. In our case, the histological findings such as periadnexal and dermal dense lymphocytic infiltrates move it close to a blaschkolinear LE.

Although positive, direct immunofluorescence is not reliable for diagnosis since lesions in the face can provide positive results without clinical relevance and moreover, immune complexes and complement along the dermoeidermal junction can be detected in a number of other inflammatory skin disorders [11, 12].

The unusual clinical presentation and in particular the appearance of milia as well as resistance to antimalarial drugs do not fully conform to a diagnosis of linear LE. Further compilation of similar cases will help to shape the diagnostic entity of LE-like facial blaschkitis.

In summary, we report a case of facial blaschkitis with histopathological findings of linear LE in early adulthood unresponsive to standard lupus treatment modalities eventually improved by laser treatment.

### Disclosure Statement

The authors have no conflicting interests to declare.

### References