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Two cases of blaschkitis with prominent pigmentation

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Dear Editor,

In 1990, Grosshans and Marot 1 were the first to use the term “blaschkitis” to describe a relapsing linear inflammatory dermatosis in adults. However, it has been unclear whether blaschkitis and adult onset lichen striatus (LS) are the same or different disease entities 2,3. Here, I argue that they are the same disease entity. I also describe two cases whose features are consistent with those of blaschkitis, namely an adult onset and the presence of multiple lines/bands along Blaschko’s lines predominantly on the trunk. However, both cases differed from other reported cases of blaschkitis in that the lesions exhibited prominent ashy pigmentation after healing. They may be cases of a blaschkitis variant that is associated with pigmentation. Various diseases that closely resemble this variant have been reported 4-6 and I suggest here that these diseases should be united as blaschkitis variants.

Case 1: A 33-year-old Japanese male noticed a linear arrangement of dark blue-brown pigmented macules on the left side of his lumbar region 3 months before seeking medical attention (Fig. 1a). Linear lesions then gradually appeared on the left side of the patient’s back. The lesions were asymptomatic. A physical examination revealed multiple bands that consisted of pinhead-sized pale-pink papules and ashy macules that were restricted to the left side of the back (Fig. 1a). All of the lesions subsided during the 6-month observation period. Some of the lesions healed with ashy pigmentation. Topical 0.1% tacrolimus ointment applied once daily 7 effectively treated the fresh erythematous or papulous lesions. The pigmentation on the lumbar and hip regions did not change over time.

Case 2: A 27-year-old Japanese male presented with a 6-month history of linear lesions and dark-blue pigmentation on the left side of his back. Some of the lesions were mildly itchy. A physical examination revealed ashy pigmentation on the left lumbar and hip regions and brown macules on the posterior of the right axillae (Fig. 1b). Linear brown pigmentations with pinhead-sized red papules were observed in a crop on the left side of the upper back that ended sharply at the middle line (Fig. 1b) and continued to the anterior chest. Narrow linear lesions consisting of red-brown maculopapules were also found on the left upper limb (Fig. 1b). These maculopapules gradually subsided within a
couple of months. Some left ashy pigmentation. These lines disappeared within a few weeks without pigmentation. All red-brown maculopapules subsided within 10 months from onset without any treatment, but the dark-blue ashy pigmentation has not changed in the >12 months since onset.

Histopathology. Case 1: Two skin biopsies were taken from the pinhead-sized red papules and the ashy pigmentation on the back. Histopathological examination of the red papules revealed vacuolar degeneration with incontinentia pigmenti (Fig. 2a, b). Perivascular lymphocyte infiltration, but not band-like infiltration, was detected. The biopsy from the dark violaceous pigmentation failed to detect inflammation, but did show many melanophages in the upper dermis that suggested preceding interface dermatitis. Case 2: Histopathological examination of the red papules on the left arm revealed slight vacuolar changes in the basal layer and an epidermis with a saw-toothed appearance (Fig. 2c, d). Mild perivascular lymphocyte infiltration and melanophages were detected in the upper dermis, but band-like infiltration of lymphocytes was not observed. Sporadic pink-staining necrotic keratinocytes were observed.

Since Blaschko’s lines of the skin are thought to reflect pathways of ectodermal development, the distribution pattern of blaschkitis indicates cutaneous mosaicism, where a progenitor cell that is genetically different due to a somatic mutation acquired during embryogenesis passes on its mutation to its daughter cells, which migrate along Blaschko’s lines. While the abnormal keratinocyte clones are usually not apparent, a triggering event such as a viral infection can cause the immune system to recognize the clones, thereby leading to inflammation with the characteristics of blaschkitis.

There are several inflammatory acquired dermatoses that follow Blaschko’s line. These include blaschkitis, LS, linear lichen planus, linear lichen nitidus, lichenoid and linear fixed drug eruptions, lichenoid chronic graft-versus-host disease, linear chronic lupus erythematosus, and inflammatory linear verrucous epidermal nevus. The first two diseases are specifically characterized by linear lesions that follow Blaschko’s lines. An analysis of 340 cases of classic LS reveals it generally affects children, with a mean age at onset of 3 years. It is characterized by discrete, hypopigmented to erythematous, flat-topped papules that exhibit lichenoid spongiotic changes upon histopathological
analysis. In LS, a blaschkolinear lesion forms on a limb and the line is narrow, solitary, and unilateral. The lesions disappear spontaneously, usually within 1-2 years. It seems that adult onset LS has more in common with adult blaschkitis than with classic LS. More than a dozen cases of both adult onset LS and adult blaschkitis have been reported previously. The mean ages at onset for both adult onset LS and adult blaschkitis are 40-50 years and in both diseases, the lesions are arranged in multiple, relatively broad, lines that are distributed predominantly on the trunk in a Blaschkolinear pattern. The lesions disappear within a few months after onset, but often recur. Some authors have claimed that adult onset LS and adult blaschkitis differ because only the latter exhibits papulovesicular lesions and spongiosis. However, adult onset LS also exhibits spongiosis, exocytosis, and dyskeratosis upon histopathological analysis. I agree with T. Hofer’s opinion, namely that these two entities do not show any clinical or morphological differences. However, in my opinion, it may be better to use the term ‘blaschkitis’ rather than the term ‘adult LS’ for this linear lichenoid inflammatory disease as this would prevent it being confused with the classic LS disease entity.

Dark blue-brown pigmentation is a prominent characteristic of both of the cases reported here. Blaschkitis-related diseases that exhibit pigmentation have been given various names, namely ashy dermatosis or lichen planus pigmentosus, lichen planus pigmentosus presenting in a zosteriform pattern, and a rare linear variant of lichen planus pigmentosus. The cases of these diseases do not appear to differ critically from blaschkitis cases except that they always exhibit prominent pigmentation. Thus, I speculate that those cases and the two cases described here are a variant of blaschkitis, where prominent pigmentation occurs due to lichenoid changes that can be observed upon histopathological analysis.

In conclusion, blaschkitis is likely to be the same disease entity as adult onset LS, and there may be a subtype of this disease entity that exhibits prominent pigmentation. This clarification is likely to facilitate our understanding of the pathogenesis of this rare skin disease.
References

3. Hofer T. Lichen striatus in adults or 'adult blaschkitis'? There is no need for a new naming. Dermatology 2003; 207: 89-92.
Figure legends

Figure 1  Clinical features.
(a) Case 1. Dark violaceous brownish maculopapules and small red maculopapules are linearly distributed along the lines of Blaschko on the left back and buttock. Close-up view of the lesions on the left lumbar region (inset). (b) Case 2. Red and dark-brown maculopapules on the left side of the middle back are linearly distributed with a sharp demarcation at the middle line. They continue to the anterior chest along the lines of Blaschko. The arrows indicate the lesions on the upper arms.
Figure 2  Histopathology

Case 1: (a) A red maculopapule shows vacuolar changes in the basal layer and perivascular infiltration of lymphocytes. (b) A dark violaceous ashy macule contains dermal melanophages (arrows). Case 2: (c) The epidermis of a maculopapule on the arm has a saw-toothed appearance and the maculopapule exhibits perivascular infiltration of lymphocytes and (d) the presence of necrotic keratinocytes (arrows). (Hematoxylin and eosin staining. Original magnification, a, b: x400; c: x200; d: x1000).