Blaschkolinear Lupus Erythematosus: Strategies for Early Detection and Management

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PRACTICE **POINTS**

- Blaschkolinear lupus erythematosus (BLE), an exceedingly rare subtype of chronic cutaneous lupus erythematosus, usually presents during childhood as linear plaques along the lines of Blaschko.
- It is important to consider linear lichen planus in patients with a blaschkolinear eruption, as the clinical manifestations are similar but there are differences in histopathology, management, prognosis, and long-term follow-up.
- Serial monitoring is indicated in patients with BLE given the potential for progression to systemic lupus erythematosus, which may be delayed with early use of hydroxychloroquine.

To the Editor:

Chronic cutaneous lupus erythematosus (CCLE) is an inflammatory condition with myriad cutaneous manifestations. Most forms of CCLE have the potential to progress to systemic lupus erythematosus (SLE).[1]

Blaschkolinear lupus erythematosus (BLE) is an exceedingly rare subtype of cutaneous lupus erythematosus that usually manifests during childhood as linear plaques along the lines of Blaschko.[2,3] Under normal conditions, Blaschko lines are not noticeable; they correspond to the direction of ectodermal cell migration during cutaneous embryogenesis.[4,5] The embryonic cells travel ventrolaterally, forming a *V*-shaped pattern on the back, an *S*-shaped pattern on the trunk, and an hourglass-shaped pattern on the face with several perpendicular intersections near the mouth and nose.[6] During their migration, the cells are susceptible to somatic mutations and clonal expansion, resulting in a monoclonal population of genetically heterogenous cells. This phenomenon is known as somatic mosaicism and may lead to an increased susceptibility to an array of congenital and inflammatory dermatoses, such as cutaneous lupus erythematosus.[4] Blaschkolinear entities tend to manifest in a unilateral distribution following exposure to a certain environmental trigger, such as trauma, viral illness, or UV radiation, although a trigger is not always present.[7] We report a case of BLE manifesting on the head and neck in an adult patient.

A 46-year-old man presented with a pruritic rash of 3 months' duration on the right cheek that extended inferiorly to the right upper chest. He had a medical history of well-controlled psoriasis, and he denied any antecedent trauma, fevers, chills, arthralgia, or night sweats. There had been no improvement with mometasone ointment 0.1% applied daily for 2 months as prescribed by his primary care provider. Physical examination revealed indurated, red-brown, atrophic plaques in a blaschkolinear distribution around the nose, right upper jaw, right side of the neck, and right upper chest (Figure, A).

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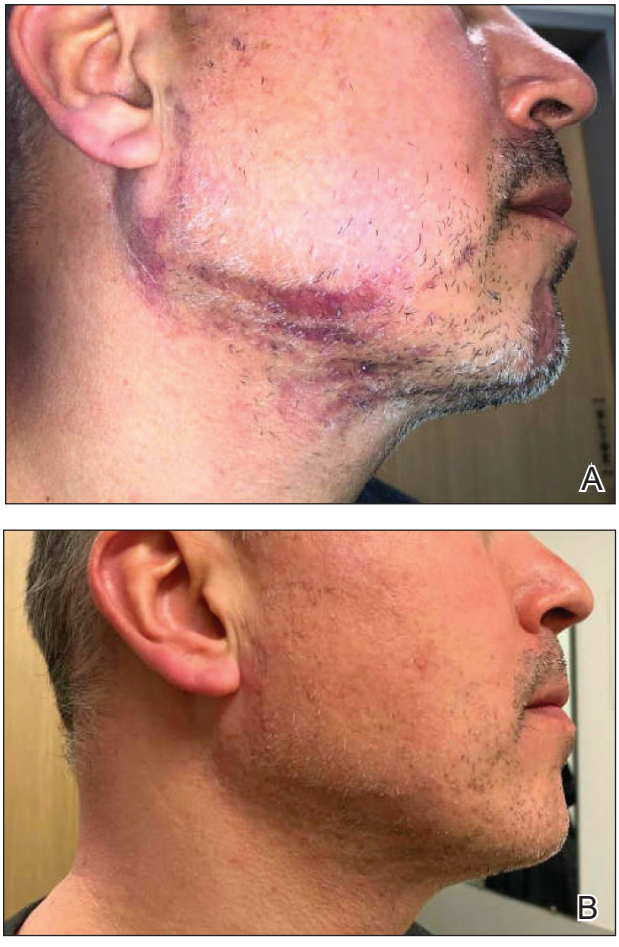
Histopathology of punch biopsies from the right jaw and right upper chest showed an atrophic epidermis with scattered dyskeratotic keratinocytes and vacuolar alteration of the basal cell layer. A superficial and deep perivascular and periadnexal lymphocytic infiltrate was observed in both biopsies. Staining with Verhoeff-van Gieson elastin and periodic acid–Schiff highlighted prominent basement membrane thickening and loss of elastic fibers in the superficial dermis. These findings favored a diagnosis of CCLE, and the clinical blaschkolinear distribution of the rash led to our specific diagnosis of BLE. Laboratory workup for SLE including a complete blood cell count; urine analysis; and testing for liver and kidney function, antinuclearantibodies, complement levels, and erythrocyte sedimentation rate revealed no abnormalities.

The patient started hydroxychloroquine 200 mg twice daily and methotrexate 25 mg weekly along with strict photoprotection measures, including wearing photoprotective clothing and avoiding sunlight during the most intense hours of the day. The patient followed up regularly, and by the 12-month visit, the pruritus had completely resolved and the rash showed considerable improvement (Figure, B). The patient demonstrated no signs of internal organ involvement that would point to progression to SLE, such as joint pain, oral ulcers, or neurologic signs; laboratory results indicating anemia, leukopenia, or thrombocytopenia; or positive antinuclear antibody testing.[8] After the 12-month visit, the patient stopped taking methotrexate, and the hydroxychloroquine was reduced to 200 mg/d.



A, Indurated, red-brown, atrophic plaques in a blaschkolinear distribution on the right upper jaw and right side of the neck, which was diagnosed as blaschkolinear lupus erythematosus following histopathology. B, After 12 months of treatment with methotrexate and hydroxychloroquine, the rash greatly improved.

Linear lichen planus is an important differential diagnosis to consider in patients with a blaschkolinear eruption.[7] Although the clinical manifestations of BLE and linear lichen planus are similar, they differ histopathologically. One study found that only 33.3% of patients (6/18) who clinically presented with blaschkolinear eruptions were correctly diagnosed before histologic examination.[7] Visualization of the adnexa as well as the superficial and deep vascular plexuses is paramount in distinguishing between linear lichen planus and BLE; linear lichen planus does not have perivascular and periadnexal infiltration, while BLE does. Thus, in our experience, a punch biopsy—rather than a shave biopsy—should be performed to access the deeper layers of the skin.

Because these 2 entities have noteworthy differences in their management, prognosis, and long-term follow-up, accurate diagnosis is critical. To start, BLE is treated with the use of photoprotection, whereas linear lichen planus is commonly treated with phototherapy. Given the potential for forms of CCLE to progress to SLE, serial monitoring is indicated in patients with BLE. As the risk for progression to SLE is highest in the first 3 years after diagnosis, a review of systems and laboratory testing should occur every 2 to 3 months in the first year after diagnosis (sooner if the disease presentation is more severe).[9] Also, treatment with hydroxychloroquine likely delays transformation to SLE and is important in the early management of BLE.[10] On the other hand, linear lichen planus tends to self-resolve without progression to systemic involvement, warranting limited follow-up.[9]

Blaschkolinear lupus erythematosus typically manifests in childhood, but it also can be seen in adults, such as in our patient. Adult-onset BLE is rare but may be underrecognized or underreported in the literature.[11] However, dermatologists should consider it in the differential diagnosis for any patient with a blaschkolinear eruption, as establishing the correct diagnosis is key to ensuring prompt and effective treatment for this rare inflammatory condition.

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