A 31-year-old man presented with a recurring pruritic rash on the right lateral flank and hip of 2 years’ duration. Physical examination revealed erythematous, verrucous, dry papules and plaques coalescing into larger plaques on the right flank and hip in dermatomal distributions involving the T10 and T11 dermatomes. A few papules were scattered in a linear eruption along the right flank and right upper thigh. Some postinflammatory changes were noted. No rash was noted over any other area of the body. Physical examination was otherwise unremarkable.

WHAT’S THE DIAGNOSIS?

a. blaschkitis
b. inflammatory linear verrucous epidermal nevus
c. lichen striatus
d. linear lichen sclerosus
e. unilateral lichen planus

PLEASE TURN TO PAGE E8 FOR THE DIAGNOSIS
A punch biopsy from the right lateral hip was performed. Histopathologic examination revealed orthokeratosis overlying mild psoriasiform epidermal hyperplasia associated with a lichenoid infiltrate composed almost entirely of lymphocytes (Figure). The infiltrate did not entirely obscure the dermoepidermal junction and spared the adnexal structures. The clinical presentation along with histopathologic analysis confirmed a diagnosis of blaschkitis. The lesions were treated with triamcinolone ointment twice daily as needed, and the patient reported symptomatic and clinical improvement with this intervention at 4-week follow-up.

Described by Grosshans and Marot in 1990, blaschkitis is an acquired inflammatory dermatosis following the lines of Blaschko. It predominantly is seen on the trunk and typically presents with pruritic papules and vesicles. It frequently has a relapsing course and is more commonly found in adults. Blaschkitis is considered a form of cutaneous mosaicism representing somatic mutations affecting epidermal cell growth and migration during embryogenesis. It has been proposed that these aberrant cells are not clinically apparent at birth; however, viral infection and drug or other environmental triggers can induce antigen presentation of the clone cells activating a T cell–mediated inflammatory response.2-4

The differential diagnosis includes other acquired Blaschko-linear dermatoses such as lichen striatus, inflammatory linear verrucous epidermal nevus, unilateral lichen planus, linear lichen sclerosus, linear psoriasis, linear fixed drug reaction, lichen nitidus, and others.1,4 Given the overlap in clinical and histopathologic presentations of the entities in the differential, it often is difficult to discern the etiology of the papular and vesicular eruption in question. Discrimination of one etiology from the others is further confounded by the fact that these lesions can all be pruritic and initially are treated with topical corticosteroids. A degree of clinical suspicion for blaschkitis coupled with prior understanding of lesional manifestations is helpful in this circumstance. Although classic lichen planus often affects the arms, legs, flexor surfaces, and occasionally the oral mucosa, blaschkitis generally is limited to the trunk. The lesions of lichen planus generally are violaceous, flattened, polygonal papules that tend to coalesce. They often have a thin, transparent, and adherent scale overlying the papular lesions, and they occasionally demonstrate Wickham striae, which are faint white reticulated networks typically seen in oral mucosal lesions. In the case of lichen nitidus, lesions often follow a geometric line due to the Köbner response, whereas physical trauma from scratching or injury causes lesions to form along the line of insult. Assessing patients for any newly initiated medications can help eliminate lichenoid drug eruptions. Lichen striatus perhaps has the most overlap with blaschkitis, having been described as also following the lines of Blaschko but occurring in children rather than adults. Inflammatory linear verrucous epidermal nevi also can be distinguished from blaschkitis on this premise, as these lesions arise during the first 5 years of life and generally affect the lower extremities.4,5

Histopathology is somewhat variable but generally includes spongiotic dermatitis with concomitant interface dermatitis characterized by T-cell infiltration. Spongiosis is a feature less commonly seen in lichen striatus. Lesions can progress over time from spongiotic dermatitis to spongiotic psoriasiform dermatitis and later to spongiotic psoriasiform lichenoid dermatitis.4 Treatment of blaschkitis should begin with reassurance of the benign nature of the dermatosis. Pruritic symptoms can be managed with a course of topical steroids.
Blaschkitis is a rare and self-limiting acquired dermatosis that should be incorporated into the differential diagnosis of Blaschko-linear dermatoses. Further investigation is needed to determine if blaschkitis and lichen striatus represent the ends of a disease spectrum or completely distinct entities.

REFERENCES


