Linear Systematized Porokeratosis—A Rare Case and Dermoscopic Clues to Diagnosis

Porokeratosis is a rare autosomal dominant keratinization disorder characterized clinically by well-defined scaly papules and plaques with hyperkeratotic ridges at the margin, and histopathologically by coronoid lamellae.[1] Linear subtype of porokeratosis normally presenting in a Blaschko-linear distribution can be confused clinically with many Blaschko-linear dermatoses.

A 43-year-old male presented with complaints of hyperpigmented papules which started over the distal aspect of upper and lower limb of left side around the age of 8 years. The papules spread centrifugally in a Blaschko pattern over a period of 15 years to involve the left side of the body without crossing the midline. On cutaneous examination there were multiple well-defined skin colored to hyperpigmented annular plaques of size varying from 0.5 × 0.5 cm to 2 × 2 cm, with central atrophy and peripheral raised thready hyperpigmented border with a collarette of whitish scales present unilaterally along the Blaschko lines on left upper limb, lower limb, neck and part of the trunk [Figure 1a-c]. Systemic examination and laboratory investigations did not reveal any abnormalities. Clinically differential diagnosis of Blaschko lichen planus and linear porokeratosis were considered.

Dermoscopy of the periphery of lesion (DermLite II hybrid m; 3 Gen; polarized mode, ×10 magnification) showed a greyish white track made by a collarette of scale with two free edges, broken at places, with a hyperpigmented blackish rim medial to it over a brown background [Figure 2a]. One of the lesion showed a perifollicular cast and exaggerated pigment network with grey globules [Figure 2b]. A 4-mm punch biopsy taken for histopathological evaluation from the thready border of the lesion revealed coronoid lamellae with few dyskeratotic cells, focally absent granular layer and dermal inflammatory infiltrate just below the coronoid lamellae [Figure 3a and b]. These features thus confirmed the diagnosis of linear systematized porokeratosis. The patient was started on tretinoin 0.05% cream once daily with no improvement on follow-up after 1 month.

Linear porokeratosis is an uncommon variant of porokeratosis which normally presents in infancy or childhood as hyperkeratotic flat papules arranged in a Blaschko-linear pattern.[1] It is thought to be a manifestation of post-zygotic mosaicism and carries the highest risk for malignant degeneration.[1] Linear porokeratosis can be further categorized into localized, zosteriform, systematized and generalized.[1,2] Localized form is unilateral, confined to an extremity and usually distal.[3] Systematized linear porokeratosis is a disseminated variant which may be unilateral or bilateral to become a generalized form.[1] Differential diagnoses for Blaschko distribution of lesions includes linear psoriasis, linear lichen planus, linear Darier’s disease, and inflammatory linear verrucous nevus.

Diagnosis of porokeratosis is confirmed by its characteristic histopathological feature of coronoid lamella consisting of a column of parakeratotic cornocytes in association with focal absence of stratum granulosum.[3] The dermoscopic findings of linear porokeratosis and its differential diagnoses are summarized in Table 1.[3-5]

This rare entity should always be considered in cases of Blaschko-linear dermatoses due to its risk of malignancy. Dermoscopy can

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be a useful auxiliary non-invasive tool in diagnosing this entity alleviating the need for biopsy.

**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.

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