## Letter to the Editor

# Linear Porokeratosis in an Adult Patient

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Dear Editor,

Porokeratosis is a rare, cutaneous disease characterized by abnormal keratinocyte differentiation. Linear porokeratosis lesions follow the Blashcko lines and usually begins during childhood or adolescence.[1] Here, we present a case of lineal porokeratosis successfully treated with a topical retinoid in an adult patient.

A healthy 31-year-old male presented with a 4-year history of multiple, hyperpigmented, annular plaques which started on the left arm and latter disseminated to the left side of the neck (Figure 1a,b). On histopathology, a tight column of parakeratotic cells (cornoid lamella) (Figure 2a), as well as a superficial mononuclear infiltrate (Figure 2b) were observed. Thus, the diagnosis of linear porokeratosis was established. Treatment was initiated with a 0.05% retinoic acid cream nightly for 6 months, showing complete lesions clearance.

Porokeratosis lesions typically show annular, hyperpigmented plaques with central atrophy and ridged borders.[2,3,4] The clinical variants of the disease include: porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis (LPK), porokeratosis palmaris et plantaris disseminata, punctate porokeratosis, and porokeratosis ptychotropica.[2]

Porokeratosis is a sporadic or autosomal-dominant inherited disorder of keratinization. The disease may be multifactorial and triggered by ultraviolet light exposure, trauma, immunosuppression, and/or infection by the human immunodeficiency virus.[3,5]

The linear variant is an infrequent form of porokeratosis that shows unilateral involvement along the Blaschko lines.[4,5] The disease often debuts during infancy, but cases in adulthood have also been reported.[3] LPK lesions are also anhidrotic and alopecic.[2,5] The disease may be localized or disseminated but it is generally confined to one side of the body. The upper extremities, trunk, and lower extremities are the most frequently involved sites, in decreasing order.[3]

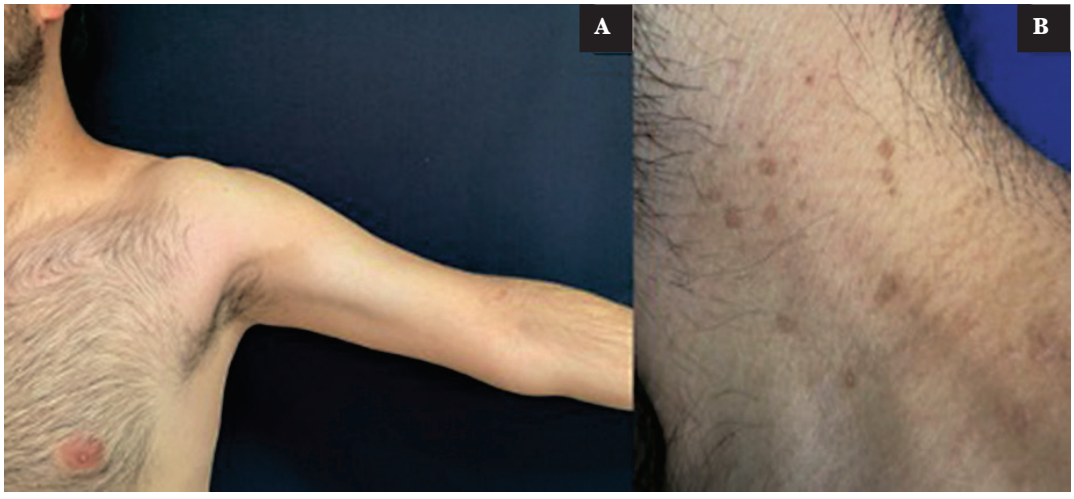


**Figure 1**
Linear Porokeratosis.
**A.** Clinical image showing the unilateral distribution of the lesions along the Blaschko lines.
**B.** Close-up view of the neck lesions.

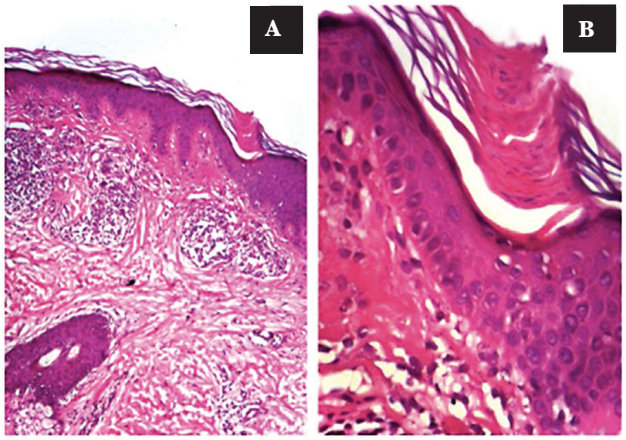


**Figure 2**
Hematoxylin and eosin-stained histopathology sections. A) Cornoid lamella and superficial mononuclear infiltrate (10X). B) At

Dermoscopy may increase diagnostic accuracy by observing central brown hyperpigmentation, raised keratin rim (white or keratin rim), peripheral double rim, peripheral black or brown dots and vascular structures such as linear vessels.[1]

The differential diagnoses include other linear dermatosis such as incontinentia pigmenti, Ito hypomelanosis, verrucous epidermal nevus, eccrine porokeratotic nevus, lichen striatus, lineal lichen planus, Moulin atrophoderma, segmental Darier's disease, etc.[3,4]

The definitive diagnosis is confirmed on histopathology. The cornoid lamella is a pathognomonic feature that consists of a dense column of parakeratotic cells over a dyskeratotic epidermis. Peripheral epidermal necrosis and amyloid in the papillary dermis can also be observed.[2,4]

Since treatment recommendations have not been standardized.[2] Topical therapies include keratolytics, vitamin D derivatives, 5-fluorouracil, imiquimod, diclofenac, retinoids, and tacrolimus. Other therapies such as ablative laser, cryotherapy, fractional photothermolysis, photodynamic therapy, and surgical excision have also been described.[4] Overall, cumulative evidence suggests that topical and systemic retinoids are the most effective agents.[3]

Porokeratosis is considered a premalignant lesion, with a risk of malignant transformation to non-melanoma skin cancer in 11-20% of the cases.[4] Therefore, sun protection measures and close medical surveillance are important to prevent new lesions, malignant degeneration, and in worst cases, metastatic disease.

Porokeratosis is a rare and potentially underdiagnosed condition, especially when the clinical presentations are not typical. Notably, delay in the diagnosis could result in malignant degeneration and worsen patient prognosis. Here, we describe a case of LPK in an adult patient with long-standing lesions that successfully remitted after a prolonged course of a topical retinoid. Undoubtedly, earlier recognition of the disease and prompt treatment establishment could significantly decrease the cost-of-care and improve patient outcomes, particularly in resource-limited scenarios such as ours.

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