Case No. 21

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Patient: A 41-year-old Thai female from Chonburi

Chief complaint: Pruritic erythematous papules and plaques on the left leg for 2 years

Present illness:
The 41-year-old female presented with multiple erythematous keratotic papules which gradually coalesced to keratotic plaques on left shin for 2 years. Various medications were previously given without clinical improvement.

Past history:
The patient denies any underlying diseases

Family history: No family members experienced similar condition to the patient’s.

Dermatological examination:
Multiple well-defined scaly erythematous papules coalescing to plaques with thread-like elevated border on the left leg.

Histopathology: Slide No. 58/3172 (left shin)
Sections display acanthotic epidermal hyperplasia with two columns of cornoid lamellae. There are several dyskeratotic cells underneath the columns.

Diagnosis: Linear porokeratosis

Treatment: She has been treated with topical corticosteroid and cryotherapy.

Discussion:
Porokeratosis are disorders of epidermal keratinization, manifesting clinically with annular or linear, sharply demarcated, hyperkeratotic plaques with distinct thread-like keratotic edge and histologically by the presence of the cornoid lamella, a column of parakeratotic corneocytes extending through the stratum corneum.1,2

Several clinical forms have been described including porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis, porokeratosis palmaris et plantaris disseminate, punctate porokeratosis, linear porokeratosis and other rarer clinical variants.1 The etiopathogenesis appears to be complex and multifactorial but is not well understood.

Linear porokeratosis is a rare variant regarded as an autosomal dominant disorder with variable penetrance that usually presents in childhood.2 Sporadic cases would be due to somatic mutation.1 Linear porokeratosis exists in two forms: in the more common localized form, lesions are confined to an extremity, frequently distal and unilaterally. In the rare generalized form, lesions are multiple, affect several extremities and also involve the trunk along Blaschko’s lines. The differential diagnosis includes linear psoriasis, lichen striatus or epidermal nevus.3

Malignant transformation incidence has been reported in 7-11% of all porokeratosis patients. Linear porokeratosis has the highest incidence of malignant transformation (11-19%).1,3-5 The most common malignancy is SCC, Bowen’s disease and rarely BCC, respectively. Risk factors are large lesion size, acral involvement and a long period of existence.3 It has been suggested that allelic loss in linear porokeratosis may explain the higher susceptibility to malignancy.6 UV exposure appears to be a trigger, causing p53 mutation.2,4

There is no known curative therapy for linear porokeratosis. Therefore, treatment may be warranted because of the risk of local malignant degeneration. The response to several therapeutic options is variable including topical keratolytics, topical and systemic retinoids, cryotherapy with liquid nitrogen and surgical approaches such as ablative laser surgery, electrodessication and curettage. Topical corticosteroids, topical tacrolimus, topical 5-FU, topical imiquimod, cryotherapy and photodynamic therapy have been successful in some cases.2,5-11

In our patient, she has been treated with topical corticosteroids combined with cryotherapy. The lesions show slight improvement. The biopsies were repeated several times and there were no malignant transformations.
References: