

Generalized Linear Porokeratosis Limited to One Side of the Body

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To the Editor:

Poroqueratosis (PQ) refers to a group of disorders of keratinization with the common histological feature of the presence of cornoid lamellae, narrow columns of parakeratosis on a focally disorganized epidermis.¹

We present 2 patients with lesions compatible with linear PQ, with extensive involvement of one side of the body.

The first patient was a 23-year-old man with no past history of interest. He was seen for the presence of multiple lesions that had been present for 20 years. The lesions had started to appear in the left inguinocrural region in the first year of life, affecting a progressively larger area, following a metameric pattern on the left lower limb and medial region of the left foot. At 4 years of age, when it appeared that the disorder had stabilized, he began to develop new lesions in the left scapular region and on the left upper limb, following a blaschkoid distribution. The

patient noticed that no new lesions had appeared after 18 years of age.

On physical examination the patient presented multiple annular lesions that became confluent, forming polycyclic and tortuous patterns. The basic lesion consisted of a central area of atrophy, occasionally verrucous, that was clearly demarcated by a peripheral hyperkeratotic border. The lesions were not infiltrated, were not adherent to the deep planes, and varied in size between 1 and 5 cm in their largest diameter. The lesions were present only on the left side of the patient's body, following a metameric or blaschkoid linear distribution, affecting the upper and lower limbs, trunk, and genital region (Figure 1), but were not found on the face.

A 5 mm biopsy that included the hyperkeratotic border and the central atrophic area was taken from a lesion on the left ankle. Central epidermal atrophy surrounded by a peripheral parakeratotic column was visible at low magnification (Figure 2). Higher magnification revealed



Figure 1. Linear porokeratosis. A. Right lower limb. B. Right scapular region. C. Right upper limb. D. Right side of the foreskin.

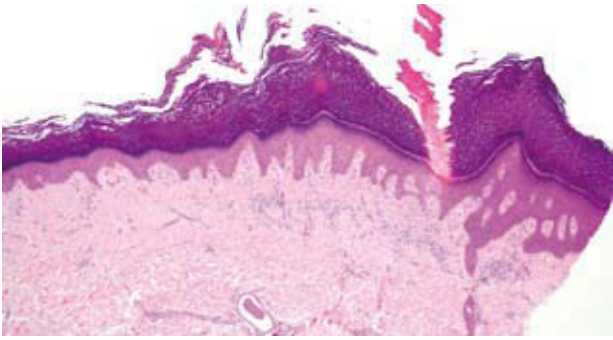


Figure 2. Histological image stained with hematoxylin and eosin. Presence of orthokeratotic hyperkeratosis surrounding an intensely eosinophilic area of parakeratosis (×40).

the presence of a column of parakeratotic hyperkeratosis, known as a cornoid lamella, associated with hypogranulosis and a discrete subjacent acanthosis.

The second case was of a 58-year-old woman, with no past history of interest. She was referred for evaluation and treatment of skin lesions situated in the right infraclavicular region and that had been present for more than 20 years.

Physical examination revealed the presence of multiple linear lesions of brownish color, with a well-defined, elevated border. The lesions were distributed over the upper third of the right side of the trunk, following a blaschkoid pattern (Figures 3A and 3B). There was a smaller number of similar lesions on the right upper and lower limbs.

With a suspicion of PQ, a 6 mm punch biopsy was performed, which showed findings compatible with this diagnosis.

The linear form of PQ, currently considered a mosaicism of classic PQ, is a rare variant that accounts for between 3.5% and 15% of all cases of PQ.^{2,3} It usually occurs



Figure 3. Linear porokeratosis affecting: A. Left scapular region. B. Left thoracic region. C and D, Left lower limb.

sporadically, and presents at birth or in early adolescence as a verrucous rash that can follow the Blaschko or metameris lines.⁴ The lesions are typically asymptomatic, and pruritus is very rare.⁵ There are currently 2 subtypes that have been described, one localized and the other generalized.^{4,6} The localized form is more common; the lesions are distributed unilaterally and are limited to a single limb, usually distally. The generalized form is rarer. It presents with very numerous lesions on more than 1 anatomic region, usually symmetrically, and can affect the trunk.^{4,6} The presence of erosions or ulcers, underlying bone abnormalities, and onychodystrophy have been reported very rarely in association with this variant of PQ.⁷

The patients we describe presented lesions that were clinically and histologically compatible with PQ, with a characteristically unilateral pattern of distribution, affecting the upper and lower limbs and the trunk, but not the face. This clinical pattern could therefore be considered to be a clinical variant within the group of generalized PQ.

The most important complication of PQ is malignant transformation of the lesions (mainly to squamous cell carcinoma and, to a lesser degree, to basal cell carcinoma),⁸ with frequencies that vary between 7.5% and 19%⁹ of cases on long-term follow-up. The lesions that present the highest risk of malignant transformation are large, long-standing lesions of the linear variant.²

It has been suggested that allelic loss or overexpression of tumor suppressor gene p53 detected in linear PQ could explain this higher risk of neoplastic transformation.^{7,10}

The therapeutic arsenal in PQ is extensive but lacking in efficacy, and the most important aspect in these patients is periodic follow-up for the early detection of malignant change in any of the lesions. Neither of our

patients developed a malignant tumor after long-term follow-up.

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Conflicts of Interest

The authors declare no conflicts of interest.

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Hair Transplantation in Temporal Triangular Alopecia

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To the Editor:

Temporal triangular alopecia (TTA) or congenital triangular alopecia is a rare form of alopecia circumscripta. First described by Saboraud,¹ its most common clinical presentation is that of a round or oval patch of noninflammatory and nonscarring alopecia, typically located in the anterior temporal area. The patch may

increase in size with the growth of the child, generally reaching a diameter of 2 to 4 cm. Inspection through a magnifying glass shows the presence of fine vellus hair and an absence of terminal hair.

TTA can be present at birth or can develop between the first few months of life and the age of 6 years. There are rare cases of adult onset,² and the current tendency is therefore to