Generalized Linear Porokeratosis: A Case Report and Discussion

Stephanie Blackburn, DO; Zaina Rashid, DO; Joan Moad, MD; Michelle Duff, DO; Jason Barr, DO
PGY-2, Affiliated Dermatology, Scottsdale, AZ, USA; Assistant Professor, Midwestern University, Glendale, AZ, USA; Laboratory Director Dermatopathologist, Dayton, OH, USA; Program Director, Affiliated Dermatology, Scottsdale, AZ, USA

Background
Linear porokeratosis is a clinical variant of porokeratosis that usually arises in infancy or childhood. It consists of one or more plaques that are similar in appearance to classic porokeratosis, but the plaques follow the lines of Blaschko and are most common on the extremities. Of all the different subtypes of porokeratosis, linear porokeratosis has the greatest chance of malignant transformation, with squamous cell carcinoma and basal cell carcinoma being the most common.

Methods
- Review of the literature on Linear porokeratosis was conducted
- Diagnosis of Linear porokeratosis based on:
  - Biopsy
  - Morphology
  - Clinical course of disease
  - Distribution of lesions

Case Presentation
We present a case of a 57-year-old man with a 45 year history of reddish-brown skin lesions showing central atrophy with surrounding scale, hyperpigmentation, and erythema on the right posterior back, right arm, right lateral leg and buttock (Fig 1). There was significant atrophic damage on his legs (Fig 2) that resolved with treatment (Fig 3).

Treatment Options & Considerations
- Generally disappointing
- Risk of malignancy
- Size & morphology of lesions
- Age of patient
- Cosmetic outcome
- Topical imiquimod
- Topical fluorouracil
- Topical steroids
- Topical retinoids & keratolytics
- Surgical options

Differential diagnosis
- Linear Darier’s
- Linear lichen planus
- Linear psoriasis
- Incontinence pigmenti

Histology
Two biopsies taken of the lower extremity showed definitive corneal lamellae with thin and flattened epidermises. Subtle interface change with few necrotic keratinocytes was also noted. There was mild superficial perivascular lymphocytic inflammation with melanophages. Focal parakeratosis with few superficial epidermal dyskeratotic keratinocytes was noted (Figure 4).

Table 1: Comparison of porokeratosis subtypes

<table>
<thead>
<tr>
<th>Variant</th>
<th>Location</th>
<th>Characteristics</th>
<th>Inheritance</th>
<th>Sequence</th>
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<tbody>
<tr>
<td>Classic porokeratosis of Mibelli</td>
<td>Extremities, anywhere</td>
<td>Prominent corneal lamellae, typically few lesions (&lt;20 cm)</td>
<td>Autosomal dominant</td>
<td>Increase in number and size, malignant degeneration reported</td>
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<tr>
<td>Disseminated superficial actinic porokeratosis</td>
<td>Anywhere (dissociated variant), sun-exposed areas (actinic variant)</td>
<td>Indistinct corneal lamellae development, uniform lesions (&lt;1 cm)</td>
<td>Autosomal dominant</td>
<td>Rapid dissemination, malignant degeneration reported</td>
</tr>
<tr>
<td>Porokeratosis palmaris et plantaris disseminata</td>
<td>Palm, soles, disseminated across body</td>
<td>Corneal lamellae, prominent keratotic ridge, disseminated, uniform lesions (&lt;1 cm)</td>
<td>Autosomal dominant</td>
<td>Malignant degeneration reported, bone and nail dystrophy</td>
</tr>
<tr>
<td>Linear porokeratosis</td>
<td>Deep dermis with unilateral linear distribution</td>
<td>Prominent corneal lamellae, characteristics, large plaques can develop</td>
<td>Mosaicism</td>
<td>Malignant degeneration reported, bone and nail dystrophy</td>
</tr>
<tr>
<td>Punctate porokeratosis</td>
<td>In association with other dermatoses, porokeratosis palmaris et plantaris disseminata variants, palm and soles</td>
<td>Discrete, punctate, hyperkeratotic, sebaceous hyperplasia, keratinocytic ridge, uniform lesions, corneal lamellae</td>
<td>Autosomal dominant</td>
<td>None reported</td>
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Discussion
Linear porokeratosis rarely affects adults and has two clinical variants. The most common variant is unilateral and confined to one extremity, while the rarer version affects multiple extremities and the trunk in a unique zosteriform pattern. Malignant transformation can occur in all porokeratosis. Linear porokeratosis has the greatest risk of developing into Bowen’s disease, SCC, and basal cell carcinoma. Risk factors include excessive sun exposure, radiation therapy, internal malignancies, and a family history of porokeratosis. It has been hypothesized that linear porokeratosis has increased malignant potential due to allelic loss and overexpression of the tumor suppressor gene p53.

Conclusion
Monitoring for suspicious lesions is key for patients with porokeratosis.

References