*** no patient handout*

Porokeratosis (linear, Mibelli, plantar)

Synopsis

Porokeratosis is a disorder of keratinization characterized by hyperkeratotic papules or plaques surrounded by a distinct peripheral, thin, ridge-like scale that expands centrifugally. Several variants of porokeratosis have been described, and a wide overlap between variants exists, suggesting that they are close in relation. Clinical variants, which are most commonly seen in the pediatric population, include linear porokeratosis, porokeratosis of Mibelli, and porokeratosis palmaris et plantaris disseminata (also known as plantar porokeratosis). All variants share a distinctive histological feature known as the cornoid lamella, which corresponds to the hyperkeratotic ridge surrounding these lesions.

Linear Porokeratosis
Linear porokeratosis is an uncommon variant that presents in infancy or early childhood as unilateral lesions contained within the lines of Blaschko. A less-common generalized form affects multiple extremities as well as the trunk. The linear variant of porokeratosis has the highest potential of malignant transformation of all porokeratoses, with squamous cell carcinoma being the most frequently associated malignancy.

Porokeratosis of Mibelli
The classic porokeratosis of Mibelli presents during infancy or early adulthood as an asymptomatic to slightly pruritic, brown to skin-colored, annular plaque with a peripheral hyperkeratotic, ridge-like scale. Lesions can range in size from 1 to 2 mm up to several centimeters in diameter. Larger lesions tend to form on the lower leg and foot, although giant facial and genital lesions have been described. The condition may be inherited as an autosomal dominant trait. Antecedent trauma or immunosuppression may be present. Lesions often persist indefinitely.

Porokeratosis Palmaris et Plantaris Disseminata (Plantar Porokeratosis)
Porokeratosis palmaris et plantaris disseminata is an autosomal dominant inherited genodermatosis presenting in adolescence or young adulthood. Initial lesions first appear on the palms and soles as small, uniform hyperkeratotic lesions with characteristic leading ridge with a longitudinal furrow. Subsequent lesions spread to involve other areas of the body, including non-sun-exposed sites as well as the mucous membranes. Males are affected twice as often as females.

Codes

ICD10CM:
Q82.8 – Other specified congenital malformations of skin

SNOMEDCT:
400080004 – Porokeratosis

Look For

All variants of porokeratosis are characterized by a distinct peripheral thin, hyperkeratotic ridge-like scale. Individual papules or plaques can range from flesh-colored to brown. Lesions range from 1 to 2 mm to several centimeters in diameter, depending on the variant.

Linear Porokeratosis

- Presents in early childhood (congenital presentations have been reported).
- Unilateral lesions confined to one extremity, often within lines of Blaschko.

Porokeratosis of Mibelli

- Presents in infancy to early adulthood.
- Some lesions may be large in diameter (up to several centimeters). Larger lesions tend to occur on the lower leg or foot.
- Multiple lesions may arise but are often unilateral and localized to one body area.

Porokeratosis Palmaris et Plantaris Disseminata (Plantar Porokeratosis)

- Presents in childhood through young adulthood.
- Initial lesions are located on the palms and soles; subsequent lesions may spread to involve other areas of the body, including non-sun-exposed skin and mucous membranes.

Diagnostic Pearls

The papules and plaques of porokeratosis are distinct, with an annular appearance and presence of a very fine outer scaly ridge.

Differential Diagnosis & Pitfalls

- **Tinea corporis** – KOH prep will demonstrate branching or curving fungal hyphae crossing cell borders.
- **Granuloma annulare**
- **Viral warts** (especially **flat warts**)
- **Actinic keratosis** – Scale present throughout the lesion.
- **Lichen planus**
- **Psoriasis**

Linear Variant

- **Lichen planus**
- **Incontinentia pigmenti** (present from birth)
- **Linear epidermal nevus**

Best Tests

Porokeratosis is often a clinical diagnosis. If unsure of the diagnosis, a biopsy of the peripheral scale will demonstrate the pathognomonic cornoid lamella with its column of parakeratotic cells extending throughout the entire thickness of the stratum corneum with underlying hypogranulosis and dyskeratotic keratinocytes.

Histopathology Findings:

- Cornoid lamella: column of parakeratosis with underlying hypogranulosis and either dyskeratotic or pale staining keratinocytes
- Epidermis may be normal, atrophic, or hyperplastic in central portion of lesion
- Localized inflammation beneath cornoid lamellae may be lichenoid or perivascular

Management Pearls

Porokeratosis is often asymptomatic, and malignant progression is rare. Therefore, medical intervention is often unnecessary, and simple observation is standard. Photoprotection with sunscreen and other sun-protection methods should be considered in all patients to decrease the risk of malignant transformation. Giant porokeratosis of Mibelli and linear porokeratosis have a slightly elevated risk of malignancy; thus, closer monitoring and biopsy of any suspicious lesions should be considered. Therapeutic intervention can be difficult, but a number of treatment options exist for lesions that are problematic or cosmetically unacceptable.

Therapy

All treatment options are poorly standardized, and outcomes are highly variable. To protect against potential malignant degeneration, discuss photoprotection with patients.

Topical Modalities
5-Fluorouracil
Topical steroids of high potency such as fluocinonide cream or ointment applied twice daily
Keratolytics
Topical retinoids
Imiquimod 5%

Systemic Modalities

Oral retinoids – isotretinoin 20 mg daily is effective, but recurrence rate is high upon discontinuation

Surgical Modalities
Cryosurgery – liquid nitrogen can be applied to individual lesions
Laser resurfacing – CO_2 laser, Nd:YAG, pulsed dye laser
Dermabrasion
Surgical excision

Drug Reaction Data

Below is a list of drugs with literature evidence indicating an adverse association with this diagnosis. The list is continually updated through ongoing research and new medication approvals. Click on Citations to sort by number of citations or click on Medication to sort the medications alphabetically.

Medication	Citations
amcinonide	1
betamethasone	1
clobetasol	2
Corticosteroid	2
desonide	1
desoximetasone	1
diflorasone	1
fluocinolone acetonide	1
fluocinonide	1

Medication	Citations
halobetasol propionate	1
hydrocortisone	1
mometasone furoate	1
Topical steroid	1
triamcinolone	1