Generalized Linear Porokeratosis: A Case Report and Discussion

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Abstract
Linear porokeratosis is a clinical variant of porokeratosis that usually arises in infancy or childhood, but may present in adulthood. There are two presentations, the first being more common and localized. It is unilateral and confined to one extremity. In the rarer version, the lesions affect multiple extremities and the trunk, appearing in a zosteriform pattern.1 Of all the variants of porokeratosis, linear porokeratosis has the greatest chance of malignant transformation, with squamous cell carcinoma and basal cell carcinoma being the most common. We present a case of a 57-year-old man with reddish-brown skin lesions showing central atrophy with surrounding scale, hyperpigmentation and erythema present on the right posterior back, right arm, right lateral leg and right buttock. Within the lesion on his leg there was noted actinic damage. There are numerous treatment options for porokeratosis, with varying benefits and risks. It is important to take into consideration the age of the patient and the morphology of the lesions being treated in order to leave the patient with the most cosmetically pleasing outcome. For our patient, we elected to treat with topical imiquimod 5% and fluorouracil 5% because of the large areas of involvement.

Introduction
The porokeratoses are a group of acquired or genetic disorders of epidermal keratinization characterized by singular or multiple, annular, atrophic lesions surrounded by a keratotic border.16 The peripheral keratinization of the demarcated lesions corresponds to a typical histopathologic feature, namely, the cornoid lamella. Various forms of porokeratosis have been established based on the clinical course of the disease, the morphology and the distribution of the lesions.1 Linear porokeratosis is a clinical variant of porokeratosis. It consists of one or more plaques that are similar in appearance to classic porokeratosis; however, the plaques follow the lines of Blaschko and are most commonly on the extremities. When linear porokeratotic lesions have a typical clinical appearance, it is easy to diagnose. However, in lesions that are smaller and have less elevation of borders, it may be confused with other linearly arranged lesions. Differential diagnosis includes inflammatory linear verrucous epidermal nevus, linear lichen planus, incontinentia pigmenti (stage II), linear psoriasis, linear Darier’s disease, and lichen striatus. We present a case of linear porokeratosis with arising SCC in situ in a 57-year-old male.

Case Report
A 57-year-old Caucasian male presented for evaluation of a lesion on the left lateral arm and was found to have extensive skin lesions showing central atrophy with surrounding scale, hyperpigmentation and erythema. The lesions were confined to the right side of his body and followed the lines of Blaschko. They were present on the right posterior back in a curved/whorled fashion, the right arm, and the right lateral leg and buttock (Figures 1-4). Extending down the lateral leg, it was evident that the inferior portion had actinic activity present (Figure 5). A shave biopsy was taken during his first visit, showing SCC in situ. Two punch biopsies taken
Biopsy of porokeratosis shows stacked, tightly packed parakeratotic cells that are well-differentiated from the rest of the corneocytes. The stratum granulosum is either absent or decreased, and the stratum spinosum may possess vacuolated or dyskeratotic cells. The defective desquamation of the corneocytes may be due to a decrease in the keratohyalin granules and lamellar bodies underneath the cornoid lamella.

During the 1980s, he visited a dermatologist with the Navy in Hawaii. He had a second biopsy performed and remembers being treated with a 5% “fading” cream that was applied to his right arm only and wrapped with cellophane. In approximately 2005, he was evaluated at an Air Force Base, where he saw a dermatologist. A biopsy was performed. He was told that it was not cancerous, and no further action was taken.

**Pathogenesis**

Porokeratosis is a premalignant disease of epidermal keratinization characterized by atrophic macules and patches with a surrounding border of hyperkeratinization. The cornoid lamella is the hyperkeratotic border of vertical mounds of parakeratotic corneocytes that lies in the periphery around the lesions. Clonal proliferation of atypical keratinocytes from the stratum corneum and superior epidermis, demonstrating abnormal terminal keratinocyte differentiation, leads to the formation of the cornoid lamella. The pathway that leads to the clonal proliferation of abnormal keratinocytes is not known; it has been thought that genetic susceptibility, UV-radiation exposure, viral infection, and immune status may be contributing factors. Immunodeficiency may be due to organ transplant, chemotherapy, chronic kidney disease, HIV, hepatitis C, or repeated trauma as well as other pathological processes. Mosaicism is a proposed genetic mechanism for two types of porokeratosis, porokeratosis of Mibelli and linear porokeratosis. Mosaicism occurs when cells within an individual have different genetic makeup. There is conflicting evidence as to the association between ultraviolet radiation and porokeratosis. Support for the relationship is due to the observation that disseminated superficial actinic porokeratosis (DSAP) occurs in individuals with extensive sun exposure, occurs on areas of sun-exposed skin, and occurs in experimental settings with the use of artificial ultraviolet radiation. However, the relative sparing of the face weakens the relationship between UV radiation and the development of porokeratosis. Also, treatment of DSAP with psoralen plus ultraviolet A (PUVA) has shown to improve lesions. Immunosuppression or immunodeficiency has been shown to increase the risk of porokeratosis. The evidence is due to reports of remission of porokeratosis after cessation of immunosuppressive therapy. Also, porokeratosis has developed in areas of long-term topical corticosteroid use.

**Discussion**

Since its first description by Mibelli and Respighi in 1893, many new variants of porokeratosis have been described. A patient may develop more than one type of porokeratosis simultaneously or consecutively. Each variant consists of its own properties regarding morphology, distribution and clinical course. The initial lesions present in a centrifugal manner as keratotic papules. These lesions then progress, showing central
atrophy with a collar of keratin. A biopsy of the lesion's border shows parakeratotic cells stacked tightly, sticking out from the rest of the stratum corneum. This cornoid lamella is the hallmark of porokeratosis. Further manifestations include thinning of the stratum granulosum, dyskeratotic cells in the stratum spinosum and subsequent thinning of the epithelium. Abnormalities in the maturation of keratinocyte clones has been implicated in the pathogenesis of porokeratosis.

The most common forms of porokeratosis are:

- **Classic porokeratosis of Mibelli (PM)**
- **Disseminated superficial actinic porokeratosis (DSAP) and its non-actinic variant, disseminated superficial porokeratosis (DSP)**
- **Linear porokeratosis**
- **Porokeratosis palmars et plantaris disseminata (PPPD)**
- **Punctate porokeratosis, which might represent a variant of PPPD**

Besides these, there are a few rare, atypical morphological forms such as facial porokeratosis, giant porokeratosis, punched-out porokeratosis, hypertrophic verrucous porokeratosis and reticulate porokeratosis. Porokeratosis ptychotropica is a recently described subtype of inflammatory perianal disease showing symmetrically distributed, reddish-brown papules and plaques involving the gluteal cleft and genital areas. Porokeratoma, otherwise known as porokeratotic acanthoma, is a tumor-like acanthoma showing cornoid lamellation characteristic of porokeratosis. These lesions have a keratotic or verrucous appearance and are commonly found on the limbs. Histologically, they have multiple and confluent cornoid lamellae. A rare congenital disorder of keratinization characterized by eczema and hair-follicle involvement is known as porokeratotic adnexal ostial nevus (POAN). This name was proposed to incorporate porokeratotic eccrine ostial and dermal duct nevus (PEODDN) and porokeratotic eccrine and hair-follicle nevus (PEHFN). Pruritic popular porokeratosis is a variant described in only about 10 previous reports in the English literature. This form of porokeratosis represents lesions that arise fairly abruptly in a patient with or without preexisting disseminated superficial porokeratosis and tend to resolve over months.

Less-commonly reported clinical entities that share the histopathologic characteristic of cornoid lamellation include viral warts, some ichthyoses, naevoid hyperkeratosis, seborrheic keratosis, squamous cell carcinoma, basal cell carcinoma, verruca vulgaris, scars, milia, and solar keratosis. A differential diagnosis includes psoriasis, actinic keratoses, Darier's disease, and lichen striatus, along with others.

Malignant transformation occurs in all of the five major forms of porokeratosis, with variable rates of transformation depending on the clinical variant. Lesions of linear porokeratosis have an increased risk of malignant transformation into squamous cell carcinoma, including Bowen's disease, and basal cell carcinoma. A few risk factors have been established, including excessive sun exposure, radiation therapy, internal malignancies, and a family history of porokeratosis. It has been hypothesized that the increased malignant potential for linear porokeratosis may be due to allelic loss in addition to overexpression of the tumor suppressor gene p53 within linear porokeratosis lesions. Monitoring for suspicious lesions is key in the care of patients with porokeratosis.

**Conclusion**

Linear porokeratosis is a rare variant of porokeratosis that has an increased risk of malignant transformation. Individuals with this type should have regular follow-up visits and yearly skin exams. There are multiple treatment options, and each patient case is different.

**References**


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