# Zosteriform porokeratosis – A case in a 7 year old girl of African descent

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**ABSTRACT**

Porokeratosis is an uncommon, inherited, autosomally dominant disorder of epidermal keratinization of uncertain cause. Five clinical variants of porokeratosis have been described. Zosteriform porokeratosis is an extremely rare presentation of linear porokeratosis with potential for malignant transformation. We report, for the first time, a 7 year old African girl with Zosteriform porokeratosis.

**Key word:** zosteriform, porokeratosis, cornoid lamella

## INTRODUCTION

Porokeratosis (PK) is an uncommon disorder of epidermal keratinization, characterized by annular plaques with an atrophic center bordered with a raised keratotic wall.[1] The disorders arise from a mutant clone of epidermal cells that expand peripherally leading to formation of the cornoid lamella at the boundary between the clonal population and normal epidermal cells.[2] The aetiology of PK remained unclear. Several clinical forms have been reported each with differing morphology, distribution and clinical course.[1,3]

## CASE REPORT

A 7-year-old girl presented to the dermatology clinic with a 4year history of progressive asymptomatic eruption on the right side of her chest. The eruption started as an asymptomatic tiny rash that was thought to be ringworm. Over the succeeding 4 year the rash had progressively expanded and enlarged over the left chest wall. The rash was unresponsive to antifungal creams and topical steroids. She denied history of trauma to the site of the lesion. There was no family history of similar rash.

Physical examination revealed a large, solitary plaque located on right chest wall in a zosteriform distribution along T7 dermatone. The lesions had a prominent raised peripheral hyperkeratotic ridge and central atrophic areas. Systemic examination revealed no abnormality. Results of routine blood tests and investigations were all within normal limits

Histopathology of Punch biopsy specimen revealed cornoid lamellae with loss of the granular layer with focal lymphocytic infiltrate (Figure 2).

The patient was treated with topical steroid and calcipotriene which led to moderate improvement of the plaque after 3 month of starting treatment.

## DISCUSSION

Porokeratosis is thought to be a genodermatosis inherited sporadically or in an autosomal dominant fashion with variable penetrance.[1] It may be associated with HIV infection,[4] diabetes mellitus[5]or liver disease.[6] Immunomodulating drugs used in patients with organ transplant may also trigger porokeratosis in genetically predisposed patients.[7]

Porokeratosis has been associated with an increased susceptibility to developing malignancies including Bowen disease, squamous cell carcinoma, and basal cell carcinoma.[8]

Clinically, porokeratosis starts as asymptomatic small brown to skin-colored keratotic papules that gradually enlarge over the years to form plaques that may measure several centimeters in diameter. Plaques are surrounded by a raised, sharply demarcated keratotic border with a longitudinal furrow. The center of the lesion may be hyperpigmented, hypopigmented, depressed, atrophic, or anhidrotic.[9] The extremities are most frequently involved, the lesions may occur anywhere on the body including mucous membranes. Multiple lesions may arise, but they are almost always

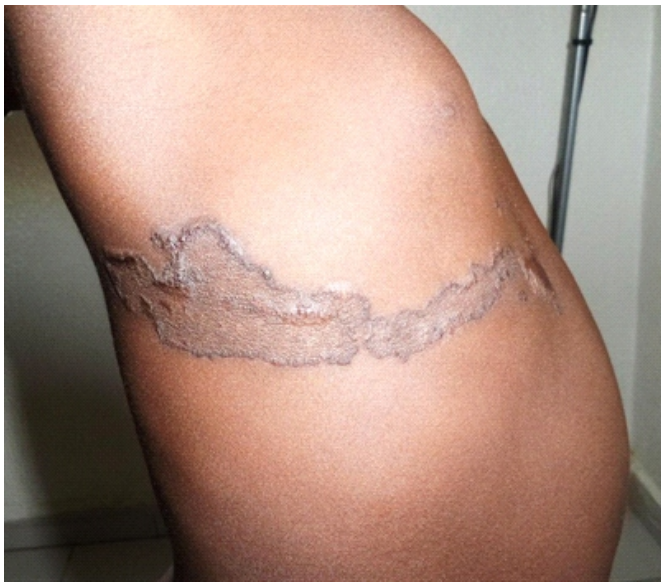

**Figure 1a:** A brown to skin-colored keratotic plaque with sharply marginated raised edge extending along T7 dermatone in a zosteriform distribution.

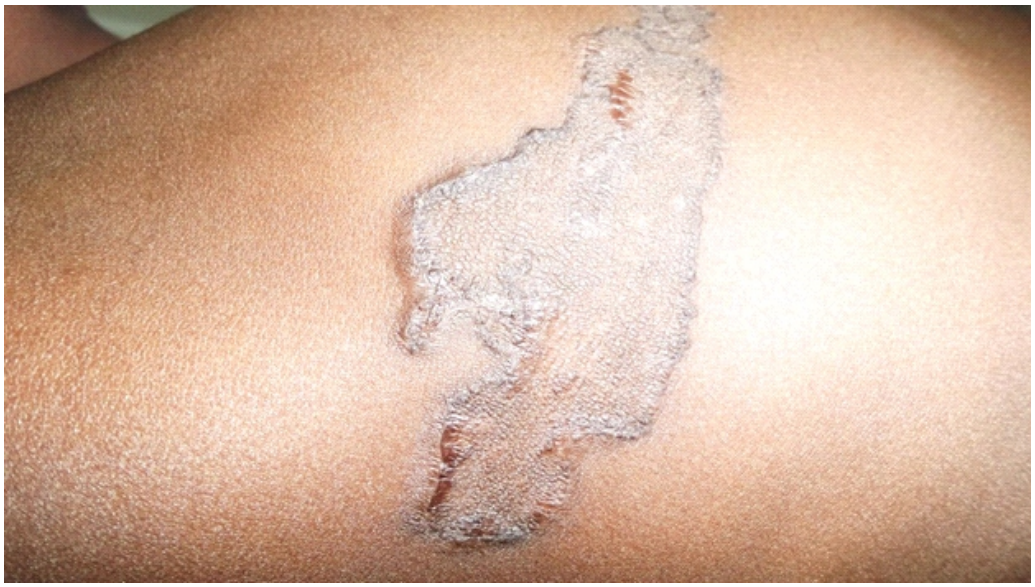

**Figure 1b:** Shows the thi thread-like keratotic edge and depressed atropic centre *(Close up)*

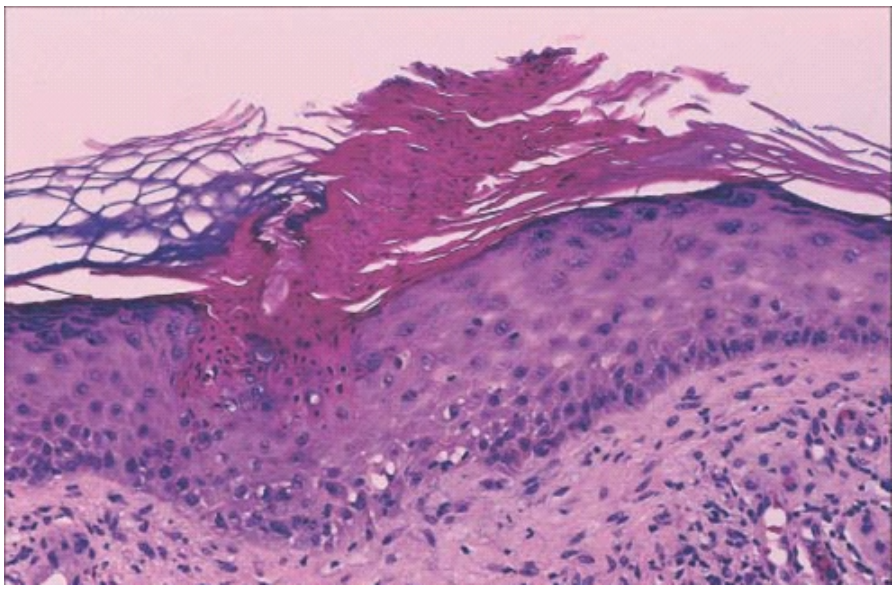

**Figure 2:** Shows a narrow column of parakeratotic cells (coronoid lamella) in the epidermis, granular layer was absent beneath the coronoid lamella

regionally localized and unilateral.[9,10]

Since the first description of classic porokeratosis by Mibelli in 1893,[11] at least 5 major clinical types have been identified. These include porokeratosis of mibelli, linear porokeratosis, disseminated superficial actinic porokeratosis, palmoplantar porokeratosis and punctate porokeratosis[9,12]

Linear Porokeratosis (LPK) is a rare variant that usually arises in childhood, although it may be seen at any age, from birth to adulthood, it is seen with equal incidence in men and women. It is composed of two forms: one common randomly distributed linear type or the other rare zosteriform variety. LPK is thought to occur through mosaicism, in which somatic mutations cause focal loss of heterozygosity.[13] All cases of LPK have a higher risk of malignant degeneration than other forms of porokeratosis, possibly because of the allelic loss hypothesized to be at fault for the formation of the lesions.[14]

Zosteriform porokeratosis (ZPK) represents a rare variant of LPK with fewer than 50 cases being reported worldwide. In contrast to other types of porokeratosis, familial cases of zosteriform porokeratosis are rare.[14] ZPK appears as a unilateral linear array of annular hyperkeratotic plaque, most commonly on the extremities.[15, 16, 17] Our patient, however , had the lesion on the trunk.

ZPK may be clinically confused with lesions from other dermatoses with striking zosteriform distribution such as nevus unius lateris, linear verrucous nevus, linear verruca vulgaris, keratosis follicularis, incontinentia pigmenti, lichen striatus, or other epidermal nevi, none of which have a cornoid lamella.[14,16,17]

All the clinical variants of porokeratosis exhibit similar histological features. They are characterized by the presence of a cornoid lamella, a tightly packed column of parakeratosis presenting within an indentation of the epidermis. The granular layer beneath the cornoid lamella either may be absent or reduced in thickness, and the keratinocytes may have a vacuolated appearance. A lymphohistiocytic infiltrate may be present in the subjacent papillary dermis. The epidermis in the central part of the lesion may appear healthy, thin, hyperkeratotic, or acanthotic.[9,12]

Several treatment modalities are available for the management of porokeratosis, however, the response usually is poor.[12] Topical therapies include 5-fluorouracil, imiquimod, salicylic acid, vitamin D analoques, glucocorticoids and retinoids.[9,12,18] Additional options include destruction of the lesion with cryotherapy, dermabrasion, electrodissection, laser ablation, photodynamic therapy[21] and/or surgical excision, all have shown varying degrees of success in treating this disorder.[9]

Systemic retinoids has been noted to be an effective treatment. However, recurrence is likely following cessation of therapy.[12]

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