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CASE REPORT

Zosteriform Porokeratosis:An Uncommon Presentation

Prathambigai S S, Neha Mariam Joseph, Samyuktha Sendhil, Sathya Narayanan R

Abstract

Porokeratosis refers to a heterogeneous keratinisation disorder. Linear porokeratosis is an uncommon type of porokeratosis with a mosaic manifestation. We present a case of childhood-onset zosteriform porokeratosis in a 14-year-old male. The zosteriform variant is a rare form of linear porokeratosis that has a propensity for malignant transformation.

Key Words

Porokeratosis, Porokeratosis of Mibelli, Linear Porokeratosis, Zosteriform Porokeratosis

Introduction

Porokeratosis is an autosomal dominant disorder.[1] The other synonymous terms for "porokeratosis" are "*parakeratosis centrifugata atrophicans*" and "*parakeratosis Mibelli*." The most common form of porokeratosis is disseminated superficial actinic porokeratosis (DSAP). While DSAP can exhibit a linear morphology, the linear type of porokeratosis follows the lines of Blaschko. It is of utmost importance to distinguish linear porokeratosis from other Blaschko-linear dermatoses.

Case Report

A 14-year-old male, hailing from Tirupati in the South Indian state of Andhra Pradesh, presented with asymptomatic raised lesions that initially started over the distal end of the right arm and spread centrifugally over the course of 10 years to involve the entirety of the right arm, the right shoulder and the right side of the neck, chest and back without crossing the midline. **(Fig 1)** Dermatological examination revealed multiple discrete as well as coalescing annular plaques, ranging in size from 1 x 1 cm to 10 x 4 cm, with raised, thread-like hyperpigmented scaly border and hypopigmented atrophic centre along the lines of Blaschko on the right arm, shoulder, neck, chest and back. Clinical differential diagnoses of linear porokeratosis, linear warts, lichen striatus, linear verrucous epidermal nevus (ILVEN) and porokeratotic eccrine ostial and dermal duct nevus (PEODDN) were made. Systemic examination was unremarkable. Routine blood investigations were normal and serology for hepatitis B, hepatitis C and HIV was non-reactive. Histopathological examination of a skin biopsy from the lesions revealed orthokeratosis of the stratified squamous epithelium with a striking cornoid lamella, focal hypogranulosis and focal lymphocytic infiltrates and the dermis showing a few dilated capillaries. **(Fig 2)** A diagnosis of zosteriform porokeratosis was made based on the clinical and histopathological features.

Treatment was initiated with oral isotretinoin 20 mg before bedtime and topical 5-fluorouracil (5-FU) cream applied twice daily for eight weeks. He also received four sessions of cryotherapy with liquid nitrogen at three-day intervals. The patient noticed mild improvement in the appearance of the lesions at the end of eight weeks.

Department of Dermatology, Saveetha Medical College and Hospital, Thandalam, Chennai 602105, Tamil Nadu, India

Correspondence to: Dr. Prathambigai SS, Postgraduate, Department of Dermatology, Saveetha Medical College and Hospital, Thandalam, Chennai 602105, Tamil Nadu, India

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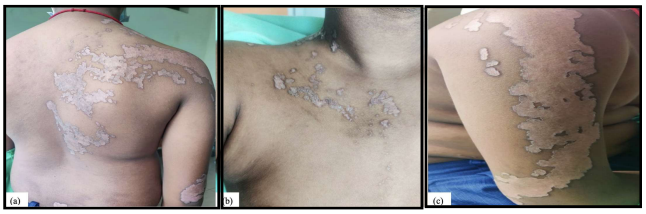

Figure 1: Hyperkeratotic annular plaques with thread-like border and atrophic centre noted over (a) right side of back and right shoulder (b) right side of neck and upper chest (c) right arm.

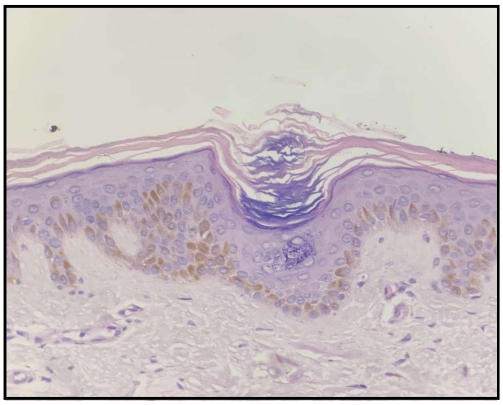

Figure 2: Section showing epidermis with focal keratotic plug, cornoid lamella and hypogranulosis. Dermis showing collagenous tissue and a few capillaries.

Discussion

Neumann first described porokeratosis in 1875.[2] However, the term 'porokeratosis' was coined by the Italian dermatologist Mibelli. He named it so because the lesion was found in association with the eccrine duct and was thus considered to be a pathology of the eccrine duct ostium.[3] Truffini described the linear form of porokeratosis in the year 1905.[4]

The pathology underlying porokeratosis includes mutations in the p53 gene, various genes on ch12q and those encoding enzymes of the mevalonate pathway.[5] Second-hit somatic mutations causing type 2 genetic mosaicism have been implicated in linear porokeratosis. Associated conditions include chronic liver disease, chronic renal disease, immunosuppression, use of steroids and biologicals, HIV infection, type 2 diabetes mellitus, photochemotherapy and radiotherapy.

Common forms of porokeratosis include disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis of immunosuppression, disseminated superficial porokeratosis of childhood, porokeratosis of Mibelli, giant porokeratosis, palmoplantar porokeratosis of Mantoux, linear porokeratosis and porokeratosis ptychotropica. Forms of linear porokeratosis maybe localised, systematised or generalised and zosteriform.[6] The zosteriform variant presents with a segmental distribution and is commonly seen over the extremities and the trunk.

The clinical hallmark of porokeratosis is the presence of a thread-like margin, which corresponds histologically to the cornoid lamella. Other histological features include focal hypogranulosis and vacuolar degeneration of keratinocytes. Cornoid lamellae are columns of parakeratotic keratinocytes derived from an abnormal clone of keratinocytes that fails to complete maturation. Although the cornoid lamella is classically described in porokeratosis, it is not uncommon in other inflammatory and inherited dermatoses like psoriasis, Grover disease, Fox-Fordyce disease, pachyonychia congenita etc.[7]

The risk of malignancy is highest in the linear type of porokeratosis with squamous cell carcinoma being the most commonly associated malignancy.[8] Other associated malignancies include Bowen's disease, basal cell carcinoma and rarely melanoma.

Available treatment options include potent topical corticosteroids, 0.1% tacrolimus cream, 5% imiquimod cream, 5% 5-fluorouracil cream, topical retinoids, topical diclofenac gel, topical vitamin D3 analogues, oral retinoids, cryotherapy, chemical peels, photodynamic therapy, radiotherapy, CO_2 laser and Q-switched lasers. Recently, treatments targeting the mevalonate pathway—such as topical lovastatin and topical cholesterol—have been explored, but the therapeutic response has been disappointing.[9] Despite the availability of various treatment options for porokeratosis, no single therapy has proven to be consistently effective.

Conclusion

The zosteriform presentation of porokeratosis is considered exceptionally rare. It's true prevalence is not established, and case reports in medical literature are limited.[10] It is more commonly seen in children and needs to be distinguished from other Blaschko-linear dermatoses that are prevalent in the paediatric population. A multimodal treatment approach, as demonstrated in our case, is crucial for achieving clinical improvement and potentially preventing malignant transformation.

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