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Facial Porokeratosis – An Uncommon Presentation of an Uncommon Disease

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Abstract

Porokeratosis is an uncommon keratinization disorder characterized by keratotic papules or annular plaques with elevated borders. It has distinctive histological findings. Porokeratosis with exclusive facial lesions is seldom reported. A 25-year-old unmarried female presented with multiple, asymptomatic, brownish plaques ranging in size from 3-6 mm over the face involving both cheeks, the dorsum of the nose, and the forehead. Examination of hair, mucosae, and nails was normal. Systemic examination was unremarkable. Histopathology revealed the presence of coronoid lamella atop a disrupted stratum granulosum and dyskeratotic keratinocytes. These features confirmed the diagnosis of porokeratosis. She was prescribed topical tazarotene gel for application at night for 6 weeks. The patient returned after 1 month with some improvement, and after that lost to follow-up.

Key words: Coronoid lamella; Face; Porokeratosis

Introduction

Porokeratosis is an uncommon keratinization disorder characterized by keratotic papules or annular plaques with elevated borders.[1] Despite being identified more than a century ago, the aetiology and pathology remain uncertain. Ultraviolet radiation, trauma, genetics, infectious agents, and immunosuppression are now considered possible causes.[2] It appears most frequently on sun-exposed skin. It is most commonly seen in the fifth decade of life but can occur at any age. It affects men and women equally. On histopathology, it shows the presence of coronoid lamella, which is a column of closely packed parakeratotic cells.[3]

Porokeratosis has several clinical variants. The most prevalent type is disseminated superficial actinic porokeratosis (DSAP). Only 15% of DSAP patients exhibit facial lesions, but in most cases, extra-facial locations, such as the extensor surfaces of the limbs, are involved. Only a few cases of porokeratosis with an exclusive facial presentation have been reported.[4]

Case report

A 25-year-old unmarried female visited the dermatology outpatient department (OPD) for several brownish lesions on her face for the last 4 months. These lesions were asymptomatic. There was no history of associated photosensitivity or recurrent infections. Her family members had no history of similar lesions. Dermatological examination revealed multiple brownish plaques with hyperkeratotic borders and atrophic centre, ranging in size from 3-6 mm on both cheeks, the dorsum of the nose, and the forehead (Figure 1). Examination of upper limbs, lower limbs, palms, soles, genitals, hair, mucosae, and nails revealed no abnormality. A clinical diagnosis of porokeratosis was made. A punch biopsy was done from the periphery of the lesion and subjected to histopathological examination, which revealed the presence of coronoid lamellae atop a disrupted stratum granulosum and dyskeratotic keratinocytes, which confirmed the diagnosis of porokeratosis (Figure 2).

Systemic examination was unremarkable. Her

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laboratory investigations were within normal limits, and the serological test for HIV was non-reactive. She was prescribed topical tazarotene gel for local application at night and sunscreen during the day for 4-6 weeks. At 1 month, there was some improvement in the pigmentation of lesions. However, the patient lost to follow-up after that.

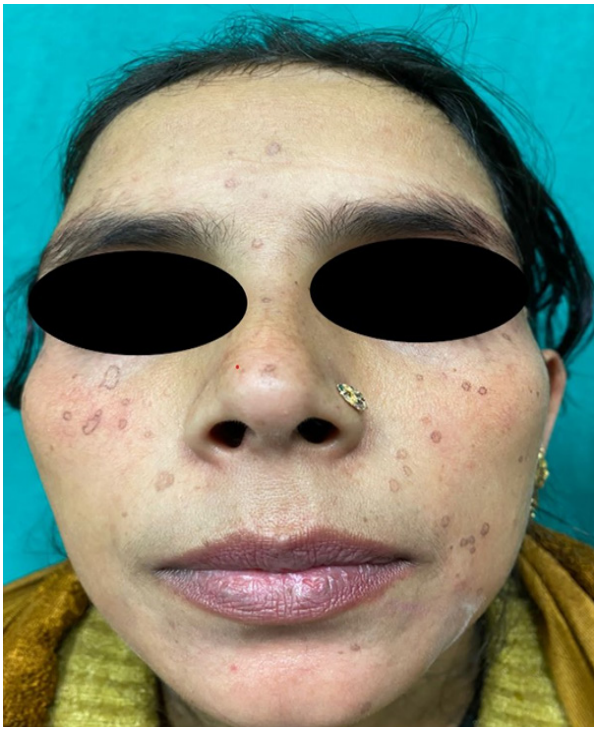

Figure 1: Multiple brownish plaques on both cheeks, the dorsum of nose and the forehead. (at first visit)

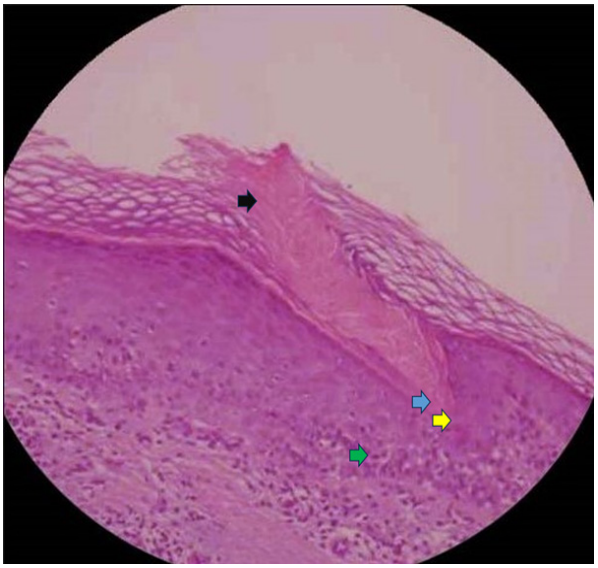

Figure 2: Histopathology showing presence of coronoid lamellae (black arrow), disrupted stratum granulosum (blue arrow), dyskeratotic keratinocytes (yellow arrow), and lymphocytic infiltrate (green arrow). (H&E stain, 400x)

Discussion

Porokeratosis is an uncommon disorder of epidermal keratinization. It encompasses a diverse group of disorders generally transmitted in an autosomal dominant fashion.[5] There are several clinical variants of porokeratosis, like disseminated superficial actinic porokeratosis (DSAP), classical porokeratosis of Mibelli, porokeratosis palmaris et plantaris disseminata, and linear porokeratosis. Rare variants include genitogluteal porokeratosis, facial porokeratosis, giant porokeratosis, porokeratosis ptychotropica, hypertrophic verrucous porokeratosis, eruptive pruritic popular porokeratosis, follicular porokeratosis, and reticulate porokeratosis.[6–9] The most prevalent variant is disseminated superficial actinic porokeratosis (DSAP). It is seen in middle-aged individuals and those with sun-sensitive skin and presents with multiple lesions, primarily in sun-exposed areas. Majority of the lesions occur in extrafacial locations, including limbs.[10] Facial lesions have been reported in 15% of the DSAP patients. Lesions of DSAP exclusive to face are uncommonly seen and hence reported.[4]

Porokeratosis has been linked to a variety of conditions, including psoriasis and phototherapy (UVA, NBUVB, and BB-UVB). Furthermore, an association with HIV infection, post-administration of immunomodulating drugs, solid malignancies, liver cirrhosis, diabetes mellitus, Crohn's disease, etc, have also been found.2 However, there were no systemic associations in our case.

Porokeratosis is histologically identified by a coronoid lamella, a thin column of densely packed parakeratotic cells. The epidermis will exhibit a column of parakeratosis with an absent granular layer and dyskeratotic cells in the upper spinous zone. This feature is found near the lesion's raised border. Below the coronoid lamella, the papillary dermis contains a dense lymphocytic infiltrate and dilated capillaries.[3] Our case had similar findings on histopathology.

The treatment of porokeratosis is challenging. General measures include strict sun protection by applying sunscreen and emollients. Several therapeutic approaches are available, with varying outcomes. These include topical 5-fluorouracil cream, topical tacalcitol, imiquimod cream, 3% diclofenac gel, oral etretinate, etc. Surgical treatment options include excision, dermabrasion with a diamond fraise, cryotherapy, erbium YAG, and CO2 lasers.[4,10] Our case was prescribed topical tazarotene 0.05% gel to be applied on the lesions at bedtime and topical sunscreen during the day.

Conclusion

Porokeratosis is a rare condition with several variants. It is unusual to observe an exclusive facial presentation. We present a case of a young woman who exhibited lesions localized to the face, accompanied by classical histopathological findings characteristic of porokeratosis.

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