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A Case Report of Keratinization Disorder-Porokeratosis of Mibelli

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Abstract

Porokeratosis is a diverse group of conditions acquired chiefly in an autosomal dominant form. It is a condition of keratinization of epidermis distinguished by annular plaques along with hyperkeratotic margins and an atrophic centre. We report a case of 45-year-old female who came with complaints of asymptomatic multiple lesions over the thighs with porokeratosis of mibelli proved histologically.

Keywords: ***Porokeratosis of mibelli; Keratinization; Cornoid lamella; Parakeratosis***

1. Introduction

Porokeratosis is a rare genodermatosis of unknown etiology, affecting epidermal keratinization due to an atypical clone of precursors of epidermis [1]. Classical form of this condition was described in 1893 by Vittorio Mibelli after which elucidation of disseminated palmoplantar porokeratosis, superficial and disseminated forms of porokeratosis, disseminated superficial actinic porokeratosis, punctate porokeratosis and linear porokeratosis was done by others [2]. Variants of porokeratosis can rarely occur together [3].

2. Case Report

A 45-year-old female presented to our dermatology outpatient department with complaints of asymptomatic multiple lesions over the thighs for the past 10 years. Similar lesions were presented, although with more elevated borders, on the medial aspect of left feet. The patient had tried numerous topical steroid creams and ointments, with no significant improvement in the appearance of lesions. The patient had no history of chronic exposure to radiation, heavy sun exposure or drug intake. The patient had a history of similar lesions in the family. Physical examination revealed multiple large, slightly scaly annular pigmented plaques measuring from 2 cm - 5 cm with elevated margins and atrophic center located over the medial aspect of both thighs and a single annular plaque with scaling and fissuring over the medial aspect of left feet as shown in the FIG. 1.

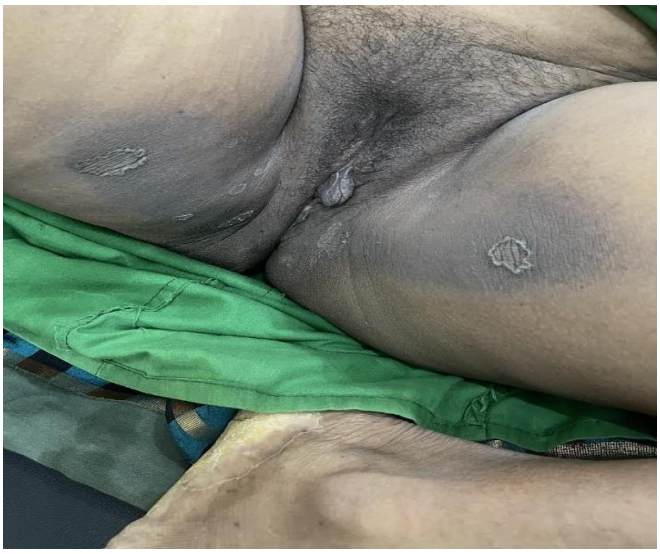

FIG. 1. **Annular pigmented plaques with elevated margins and atrophic center located over the medial aspect of both thighs and a single annular plaque with scaling and fissuring is seen over the medial aspect of left feet.**

Histopathological examination revealed a prominent cornoid lamella with invagination of the epidermis at the site of the cornoid lamella with the characteristic “smoke from chimney” appearance. Interruption of the granular layer below the cornoid lamella was noted. Based on the clinical and histopathological findings, definitive diagnosis of porokeratosis was made.

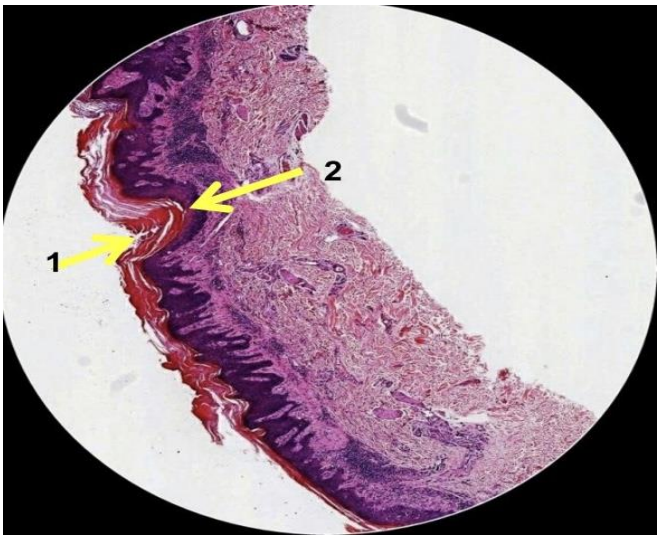

FIG. 2. **An epidermal invagination filled with keratin is seen (1). The apex of the invagination usually points away from the central portion of the lesion (2).**

3. Discussion

Porokeratosis is not a common diagnosis which occurs consistently in the fifth decade of life, although it can be seen in all groups with equal incidence in females and males. The classical form of porokeratosis and linear form of porokeratosis are commonly seen in infancy or childhood, whereas punctate porokeratosis during adolescence and disseminated superficial actinic porokeratosis during adult life [3]. It is usually an acquired condition, but familial component is also noted. Variants of

porokeratosis can rarely occur together. Later in the life, these lesions can undergo malignant transformation either into basal cell carcinoma less frequently or squamous cell carcinoma most frequently [4]. It is caused by growth disorganized epidermal cells or due to overexpression of p53 [5]. Eruptive form of this disease is related to immunosuppressive conditions such as malignancy, inflammatory states, and transplantation patients [6].

Porokeratosis is divided into localized and generalized forms. Involvement of genital region and its surrounding areas such as thighs, groin, buttocks and perineum can occur in localized or generalized forms. Localized involvement of thighs and perineal region is not a common presentation with around 24 cases detailed in history. These lesions can occur anywhere on the body but commonly involve trunk and limbs and less commonly face, scrotum and genitourinary areas [7]. They can be asymptomatic or associated with localised itching initially [8,9]. The skin lesions of porokeratosis are classically marked by single or multiple, well-marginated annular plaques and few papules which have an erythematous to brown colour with an atrophic and hypopigmented or pigmented center [10]. The disseminated form of porokeratosis is distinguished clinically by brown to pink macules and papules with elevated borders [6].

A rare variant reported in 1995, known as porokeratosis ptychotropica is distinguished by involvement of gluteal cleft and buttocks with pruritic annular plaques and having multiple cornoid lamellae on histopathological examination, which is a unique feature of this variant, making it a major differential diagnosis in porokeratosis involving the genitogluteal region [9,11].

Confirmation of diagnosis is made by doing a biopsy which reveals the characteristic cornoid lamella [12]. Along with biopsy, dermoscopic examination can also be done to confirm the diagnosis which reveals a central brownish pigmentation along with multiple dots that are blue-gray in colour with an adjacent hypopigmented band [13].

The treatment approach must be decided after analysing the anatomical location of the lesion, size of the lesion, patient's preference, risk of malignancy and cosmetic considerations. Main emphasis should be made upon sun protection. Treatment modalities comprise topical preparations like 5% imiquimod, 5-fluorouracil, vitamin-D3 analogs, corticosteroids and retinoids. Systemic therapy includes oral corticosteroids as well as retinoids. Other therapeutic options include surgery, photodynamic therapy, cryotherapy, CO2 laser and dermabrasion [14,15].

4. Conclusion

Porokeratosis is a rare skin disorder with a wide spectrum of clinical variants. The treatment of porokeratosis is often unsatisfactory and hence a watch-and-wait approach should be taken into consideration.

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