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Zosteriform Porokeratosis of Mibelli

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Porokeratosis is a distinct skin disorder thought to be caused by a clonal dyshomeostasis of epidermal keratinization. It is characterized by single to multiple flat or atrophic patches surrounded by a clinically and histologically distinctive hyperkeratotic border, so-called cornoid lamella. Several clinical variants have been identified. Herein, we reported a 27-year-old male showing a strikingly vivid zosteriform dermatosis that is histologically consistent with the diagnosis of porokeratosis. Zosteriform porokeratosis of Mibelli is an extremely rare presentation of porokeratosis and should be carefully differentiated from other linear dermatosis.

Key word: zosteriform, porokeratosis, cornoid lamella

INTRODUCTION

Porokeratosis (PK) is a specific disorder of epidermal keratinization with a characteristic histological presentation of the cornoid lamella, a thin column of stacked parakeratotic cells extending through the stratum corneum, corresponding to the clinical appearance of a hyperkeratotic ridge-like border.[1] The exact pathogenesis of PK is not fully understood, but it is presently believed to be resulted from an expanding mutant clone of keratinocytes which evolves into the so-called cornoid lamella.[2] The age of onset can range from birth to adulthood with variable sex predominance in different clinical types, among which the zosteriform PK of Mibelli is a rare subtype of linear PK and merits attention from clinicians due to a higher rate of malignant transformation in this variant.[3]

CASE REPORT

A 27-year-old male presented at our dermatology clinic with an asymptomatic progressive skin eruption which has been noted on the left thigh since he was three years old. Over the following years, the eruption extended both proximally and distally to assume a zosteriform arrangement. The past medical history was unremarkable. There was no family history of a similar skin condition. Physical examination revealed multiple annular plaques of various sizes arranged in a zosteriform pattern extending from the left buttock to the left dorsal foot (Fig. 1). The individual lesion appeared as a dark-brown hyperkeratotic plaque with a somewhat atrophic center surrounded by thread-like raised border (Fig. 2). In some areas, these annular lesions coalesced to form large polycyclic

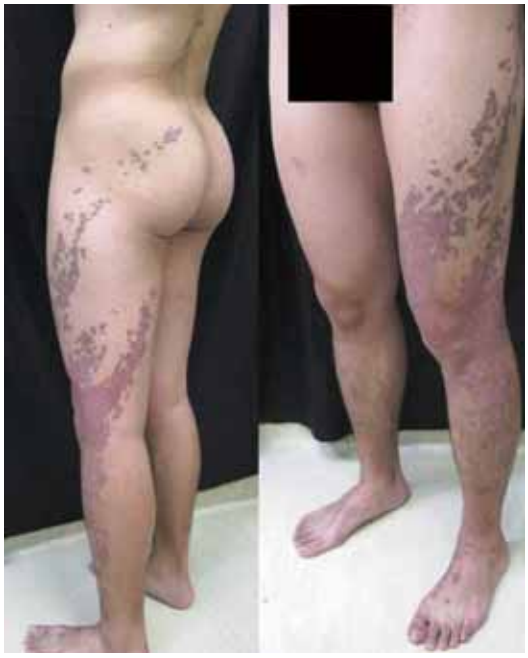

Fig. 1 Multiple discrete and confluent brownish hyperkeratotic plaques extended from left buttock to the left dorsal foot in a zosteriform distribution.

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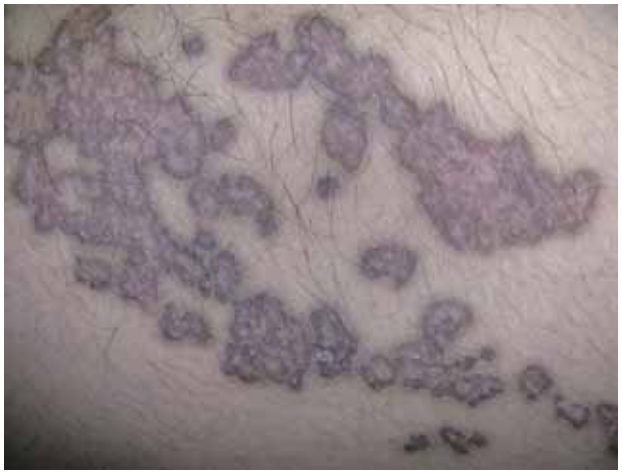

Fig. 2 The close-up picture revealed that the individual lesions are brownish hyperkeratotic circular plaques with atrophic centers and thread-like elevated borders. Some lesions are confluent into large polycyclic plaques.

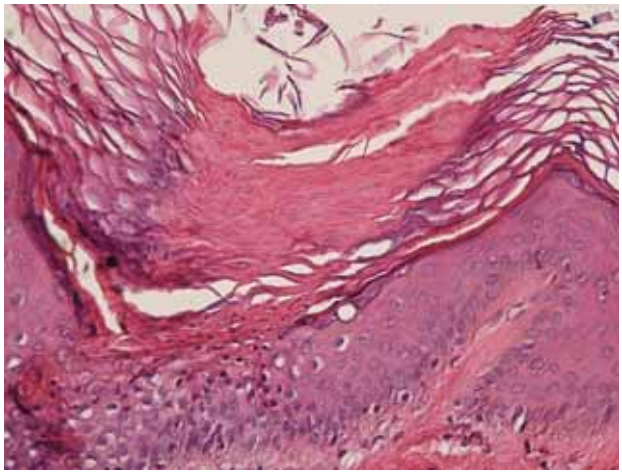

Fig. 3 Histopathology showed a thin column of tightly packed parakeratotic cells within a keratin-filled epidermal invagination. In the epidermis beneath the parakeratotic column, the keratinocytes are irregularly arranged and have pyknotic nuclei with perinuclear edema and no granular layer is seen under the parakeratotic column. (haematoxylin and eosin stain, original magnification ×400).

plaques. Histopathology of the specimen taken from the left thigh showed mild hyperkeratosis, acanthosis, and a thin column of tightly packed parakeratotic cells within a keratin-filled epidermal invagination. In the epidermis beneath the parakeratotic column, keratinocytes containing pyknotic nuclei with perinuclear edema were irregularly arranged and no granular layer was seen under the parakeratotic column. Infiltration of a few perivascular mononuclear cells and scattered melanophages in the papillary dermis were noted as well (Fig. 3). According to the clinico-pathologic correlation, a diagnosis of zosteriform PK of Mibelli was confirmed. He was then treated with 0.1% adapalene gel twice a day with moderate improvement of his skin lesions.

DISCUSSION

PK is an uncommon skin disorder of epidermal keratinization characterized by one or more annular hyperkeratotic plaques carrying an atrophic center with elevated thread-like ridge which expands centrifugally. The hallmark of PK is the formation of cornoid lamella, a distinct histopathologic feature, corresponding to the clinical manifestation of the elevated hyperkeratotic border. Under the microscope, the cornoid lamella represents a thin column of poorly stained parakeratotic cells within a keratin-filled epidermal invagination. In the epidermis beneath the parakeratotic column, the keratinocytes are irregularly arranged and have pyknotic nuclei with perinuclear edema. There is often an absent or markedly reduced granular layer under the cornoid lamella, indicating that the normal differentiation process has been altered. There are five clinical variants of PK being recognized, namely classic PK of Mibelli, disseminated superficial actinic PK, PK palmaris et plantaris disseminata, linear PK, and punctate PK.[3] Linear PK is a rare variant that usually arises in childhood and it is composed of two forms: one common randomly distributed linear type or the other rare zosteriform arrangement of multiple typical annular lesions.[4] Our patient represents the typical feature of the latter in both clinical and pathological aspects.

The genuine pathogenesis of PK remains elusive even after a century since the first description by Mibelli in 1893. Very rarely was taken as genodermatosis because of the many reported familial cases. In addition, evidence shown in cultured fibroblasts derived from porokeratotic lesions exhibited instability of the short arm of chromosome 3 as well as numerous re-arrangements and clone formation.[5] However, there are still many sporadic cases without family history like our patient. Therefore, genetic predisposition may only contribute partially to the pathogenesis of PK. Reed *et al.* suggested that the lesions of PK represented an expanding mutant clone of keratinocytes forming cornoid lamella.[2] This view was supported by the findings of abnormal DNA ploidy in the epidermis of PK.[6] On the other hand, linear PK was thought to represent a mosaic variant of the classic PK of Mibelli

resulting from a postzygotic mutation.[7]

Many factors may trigger the occurrence of PK in genetically predisposed patients such as ultraviolet exposure, immunosuppresion or organ transplantation.[8] Importantly, PK has the potential for malignant transformation into squamous cell carcinoma or basal cell carcinoma.[1] According to a large review of 281 cases with porokeratosis, 21 (7.5%) of them had an associated malignancy that evolved from the lesion.[9] Moreover, it was found that 8 out of the 21 (38%) malignancies arose in linear porokeratosis. Another case report represented appropriately an example of this predilection because the reported patient exhibited nine squamous cell carcinomas developing exclusively from the linear areas in a background of disseminated superficial porokeratosis.[10] Therefore, clinicians should advice patients to have adequate sun-protection and stay alert of any uncomfortable changes upon the PK lesions. PK usually runs a chronic and progressive course, whereas no single consistently effective therapy was available. Different treatment modalities with variable success including topical 5-fluorouracil, tretinoin, imiquimod, calcipotriol, cryotherapy, carbon dioxide laser and photodynamic therapy may be considered.[11-12] Our patient was treated with topical retinoid cream with moderate improvement of the skin lesions.

In conclusion, zosteriform PK is a rare variant of PK characterized by a clonal aberration of keratinization. The clinically striking zosteriform distribution shown in our case should be differentiated from other linear dermatoses such as nevus unius lateris, linear verruca vulgaris, keratosis follicularis, incontinentia pigmenti, lichen striatus, or other epidermal nevi.[13] Accurate diagnosis could be achieved by histopathologic examination. Owing to the higher rate of malignant transformation observed in this rare variant, long-term follow-up is necessary and highly recommended.

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