

Zosteriform Porokeratosis: An Unusual Presentation

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Abstract: Since the original description of porokeratosis in 1893 by Mibelli, various other forms of this disease have been described. Among these variants zosteriform porokeratosis is considered to be extremely rare with higher rate of malignant transformation. We are reporting an unusual presentation of zosteriform porokeratosis resembling nevoid psoriasis in a 19 year old boy over the left side of abdomen associated with intense pruritus.

Key words: zosteriform porokeratosis, nevoid psoriasis, pruritus

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I. Introduction

Porokeratosis is clinically characterized by sharply demarcated hyperkeratotic annular lesions with distinct keratotic edges corresponding histologically to the presence of cornoid lamella.^[1] 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.^[2]

II. Case report

A 19 year old boy presented to us with a pruritic well defined erythematous plaque approximately 35×3.5 cm in a linear and grouped array over the left side of abdomen along the line of blaschko extending from 12cm below the left nipple to 5cm below umbilicus since last 7 months[Fig1] Hair, nail and mucosal surfaces were normal. Other area and systemic examination were normal. There was no history of consanguinity, and no family history of similar disease.

The lesion, which initially started as grouped papules over the lateral side of the left part of abdomen 7 months back, gradually extended in linear fashion up to 5cm below umbilicus. We kept the differential diagnosis of inflammatory linear verrucous epidermal nevus and nevoid psoriasis. Histological examination of biopsy from the edge of the lesion showed parakeratotic column of cells (cornoid lamella) traversing the stratum corneum and a thin granular epidermis [Fig2]

III. Discussion

PK is an uncommon skin disorder of epidermal keratinisation characterized by one or more annular hyperkeratotic plaques carrying an atrophic centre 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] Sometime linear porokeratosis can resemble linear epidermal verrucous nevi.^[5] Rarely, verruciform lesions simulating hypertrophic lichen planus or warts can occur in porokeratosis.^[6] In our patient the lesion clinically mimicking to psoriasis [fig1]. So we kept the differential diagnosis of psoriasis vulgaris, nevoid psoriasis, and inflammatory linear verrucous epidermal nevus. Patient was treated with topical potent corticosteroid and antihistaminic. There was no change in lesions even after 20 days of

treatment. Histopathological features were consistent with porokeratosis. According to the clinico-pathologic correlation, a diagnosis of zosteriform PK of Mibelli was made and then he was treated with topical tretinoin 0.04% once a day with moderate improvement of his skin lesions [Fig3] but lesion recur after stopping treatment.

After that 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^[7] 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.^[8] This view was supported by the findings of abnormal DNA ploidy in the epidermis of PK.^[9] On the other hand, linear PK was thought to represent a mosaic variant of the classic PK of Mibelli resulting from a post zygotic mutation.^[10] Many factors may trigger the occurrence of PK in-genetically predisposed patients such as ultraviolet exposure, immune suppression or organ transplantation.^[11] Importantly, PK has the potential for malignant transformation into squamous cell carcinoma or basal cell carcinoma.^[12] Therefore, clinicians should advise 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.^[13-14] Our patient was treated with topical tretinoin cream with moderate improvement of the skin lesions [fig3]

In conclusion, zosteriform PK is a rare variant of PK. Zosteriform PK can be present as an unusual appearance mimicking nevoid psoriasis as in our case should be differentiated from other linear dermatoses. Accurate diagnosis could be achieved by histopathologic examination. Because of the higher rate of malignant transformation in this rare variant of PK, a long term follow up is necessary and highly recommended.

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Fig. 1: Skin biopsy shows marked hyperkeratosis, acanthosis and keratin filled epidermal invagination with an angulated parakeratotic tier (Cornoid lamella) with hypogranulosis deep to it. (H&E stain, 40 x)

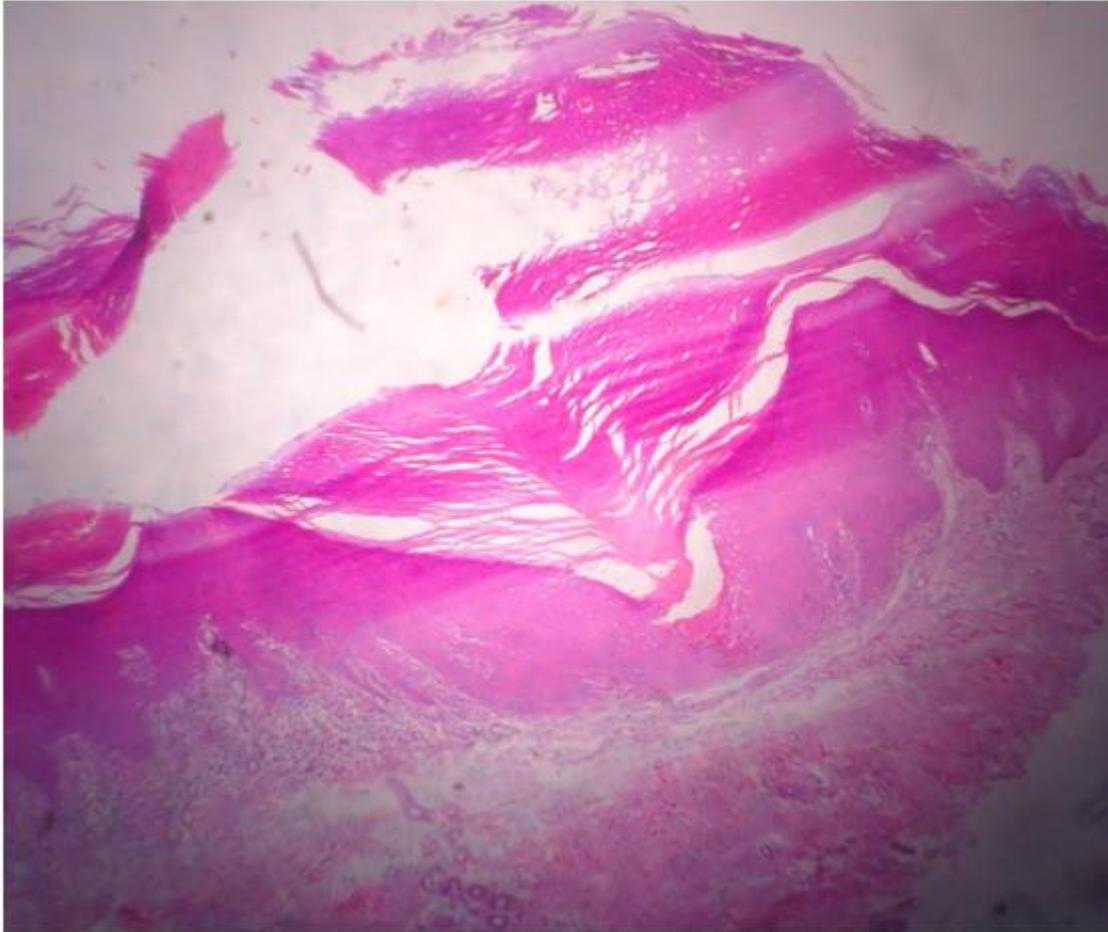


Fig. 2: Zosteriform porokeratosis after treatment with topical tretinoin 0.04%

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