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Case report

Porokeratosis ptychotropica with scalp involvement: A case report

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Porokeratosis, a spectrum of disorders characterized by abnormal keratinization, includes the rare variant porokeratosis ptychotropica, which typically manifests with hyperkeratotic plaques on the buttocks and genital area. Its clinical mimicry to various skin diseases and under-recognition frequently result in misdiagnosis. This challenge is compounded by the resistance of porokeratosis ptychotropica to treatment.

Herein, we report a 62-year-old Thai male with persistent pruritic hypertrophic plaques that affected his genitogluteal area, extremities, and scalp over 5 years, which was unresponsive to diverse topical agents. Histopathological findings revealed an acanthotic epidermis with a cornoid lamella, consistent with porokeratosis ptychotropica. Acitretin was administered orally at a dosage of 25 mg every other day to 25 mg daily for 7 months, resulting in significant improvement.

This report introduces the first documented case of porokeratosis ptychotropica with scalp involvement, expanding our understanding of disease manifestations. Although its treatment poses challenges because of resistance to many conventional treatments, this patient exhibited remarkable improvement after the oral intake of acitretin.

Keywords: Acitretin, genital disorder, porokeratosis ptychotropica, retinoid.

Porokeratosis is a disorder of abnormal keratinization with cornoid lamella formation upon histopathology.[1] Several clinical variations have been described, such as disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis, porokeratosis of Mibelli, linear porokeratosis, and eruptive disseminated porokeratosis.[2] Porokeratosis ptychotropica is a less-recognized subtype that primarily affects the genitogluteal area but occasionally extends regionally. Clinically, porokeratosis ptychotropica can mimic other lesions and is misdiagnosed frequently. This type is resistant to treatment, and no standard treatment guidelines have been established.[3] Existing knowledge recognizes oral retinoid therapy as a viable option.[4] In this report, the author highlights the noteworthy improvement in a patient after acitretin administration. Interestingly, this is the first report of a patient with porokeratosis ptychotropica involving the scalp apart from the anogenital area and extremities.

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Case report

A 62-year-old Thai male presented with a 5-year history of gradually enlarging pruritic verrucous and hypertrophic plaques involving the genitogluteal region. The lesions extended to the upper back, extremities, and scalp. Despite topical treatments, no improvement was observed. On physical examination, multiple hyperpigmented, hyperkeratotic plaques were observed on the penis, scrotum, intergluteal cleft, and buttocks. Similar plaques were observed on the upper back, arms, legs, and scalp **(Figure 1A - 1D)**. A dermoscopy of the upper back lesion revealed peripheral hyperkeratotic and elevated border plaque **(Figure 2A)**. A biopsy performed at the periphery of the lesion on the upper back showed an acanthotic epidermis with cornoid lamella **(Figure 2B),** confirming the diagnosis of porokeratosis ptychotropica. The patient received oral acitretin therapy at a dosage of 25 mg every other day to 25 mg daily. The lesions significantly improved after 7 months of treatment **(Figure 3A - B).**

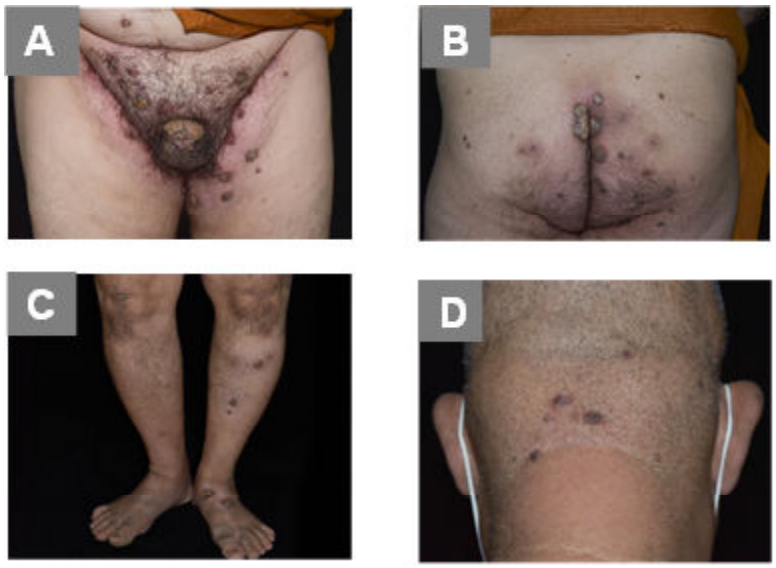

Figure 1. Multiple brownish to hyperkeratotic plaques on genital area **(A)**; buttock **(B)**; legs **(C)**; and scalp **(D)**.

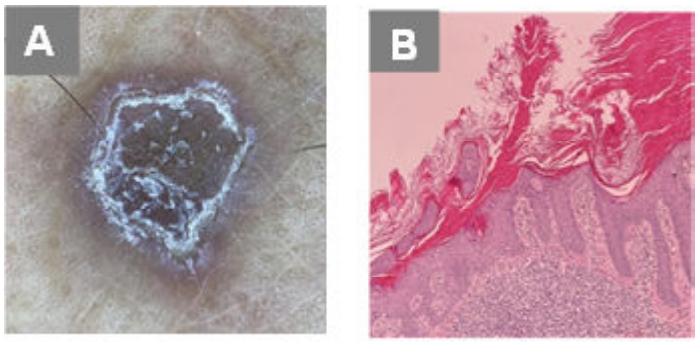

Figure 2. Dermoscopy reveals a hyperkeratotic thread-like elevated border plaque **(A)**; Hematoxylin-eosin shows dense lichenoid lymphohistiocytic infiltrate in the upper dermis with overlying acanthotic epidermis and cornoid lamella **(B)**.

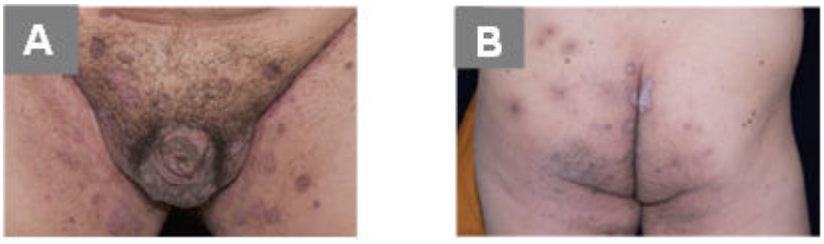

Figure 3. Clinical response after 7 months of systemic acitretin on genital area **(A)**; and buttock **(B)**.

Discussion

The exact etiology of porokeratosis ptychotropica remains unknown, and most cases are reported as sporadic and occurring more commonly in men.[5] Owing to its clinical resemblance to various conditions, porokeratosis ptychotropica is often misdiagnosed as viral warts, condyloma acuminata, psoriasis, chronic eczema, or lichen simplex chronicus.[6] Therefore, a skin biopsy ensures an accurate diagnosis and excludes other potential conditions. A shared histological finding among porokeratosis variants, including porokeratosis ptychotropica, is the presence of cornoid lamella, indicating abnormal keratinization. However, cornoid lamellation is not restricted to porokeratosis and can be detected in various inflammatory and neoplastic skin conditions, such as viral warts, seborrheic keratosis, and squamous cell carcinoma in situ.[7] Although only one case of porokeratosis ptychotropica progressing to squamous cell carcinoma was reported, it might be linked to a rare disease.[8] Therefore, an early diagnosis through history assessment and skin biopsy can confirm the diagnosis and exclude other conditions to avoid inappropriate treatment. Porokeratosis ptychotropica is challenging to treat, often resulting in dissatisfaction with variable responses.[3] Many therapeutic modalities have been described, such as topical steroids, topical retinoids, topical antifungals, 5.0% imiquimod, topical salicylic acid, cryotherapy, intralesional steroid, intralesional bleomycin, psoralen plus ultraviolet-A, and CO_2 laser. Despite these treatment options, none of them appear to accomplish total clearance. [9, 10] However, the potential efficacy of oral retinoid therapy, as well as dermatome and surgical excision, was reported.[10] Few studies have investigated the effect of oral retinoid therapy, specifically isotretinoin and acitretin, on porokeratosis ptychotropica. The outcomes show substantial improvement, typically noticeable from 1 month to a year. [3, 4, 11] Successful removal of the affected area on gluteal regions using a dermatome was reported, with no complications or recurrence.[12] Surgical excision is recognized as a highly effective modality, supported by several case reports that document complete remission without recurrence.[13 - 15]

Conclusion

This case report emphasizes the necessity for clinicians to consider porokeratosis ptychotropica in the differential diagnosis of pruritic hyperkeratotic plaque in the anogenital regions. Beyond its predilection for the genitogluteal area, porokeratosis ptychotropica can affect the extremities and scalp. A skin biopsy is crucial for diagnosis and exclusion of other possible conditions. Extended courses of oral acitretin therapy lead to substantial improvement and can be considered a promising treatment.

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Conflict of interest statement

Each author has completed and submitted an International Committee of Medical Journal Editors Uniform Disclosure Form for Potential Conflicts of Interest.

Data sharing statement

Data generated or analyzed for the present report are included in this published article. Further details are available from the corresponding author on reasonable request after deidentification of the patient whose data are included in the report.

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