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Rare Variant of Porokeratosis: A Case Report

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Awad, Nardin, "Rare Variant of Porokeratosis: A Case Report" (2021). *Stratford Campus Research Day.* 75. https://rdw.rowan.edu/stratford_research_day/2021/may6/75

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Rare Variant of Porokeratosis: A Case Report Nardin Awad, OMS-III

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Introduction

There are 5 clinically distinct variants of porokeratosis, including classic porokeratosis of Mibelli, disseminated superficial (actinic) porokeratosis, linear porokeratosis, punctate porokeratosis, and porokeratosis palmaris et plantaris disseminate. ¹ Porokeratosis ptychotropica (PP) is the sixth, lesser-known variant of the spectrum. ^{3,4} PP is characterized by verrucous plaques, often resembling psoriasis plaques, that affect the buttocks, most commonly the gluteal cleft, and can also involve the extremities, most commonly the lower legs and feet. ^{2,3} Due to its characteristic verrucous appearance, designations such as verrucous or hyperkeratotic porokeratosis are occasionally used. ¹ Besides its predilection for specific locations, PP also has a significant predilection for men versus women. ^{1,2,3}

Case Description

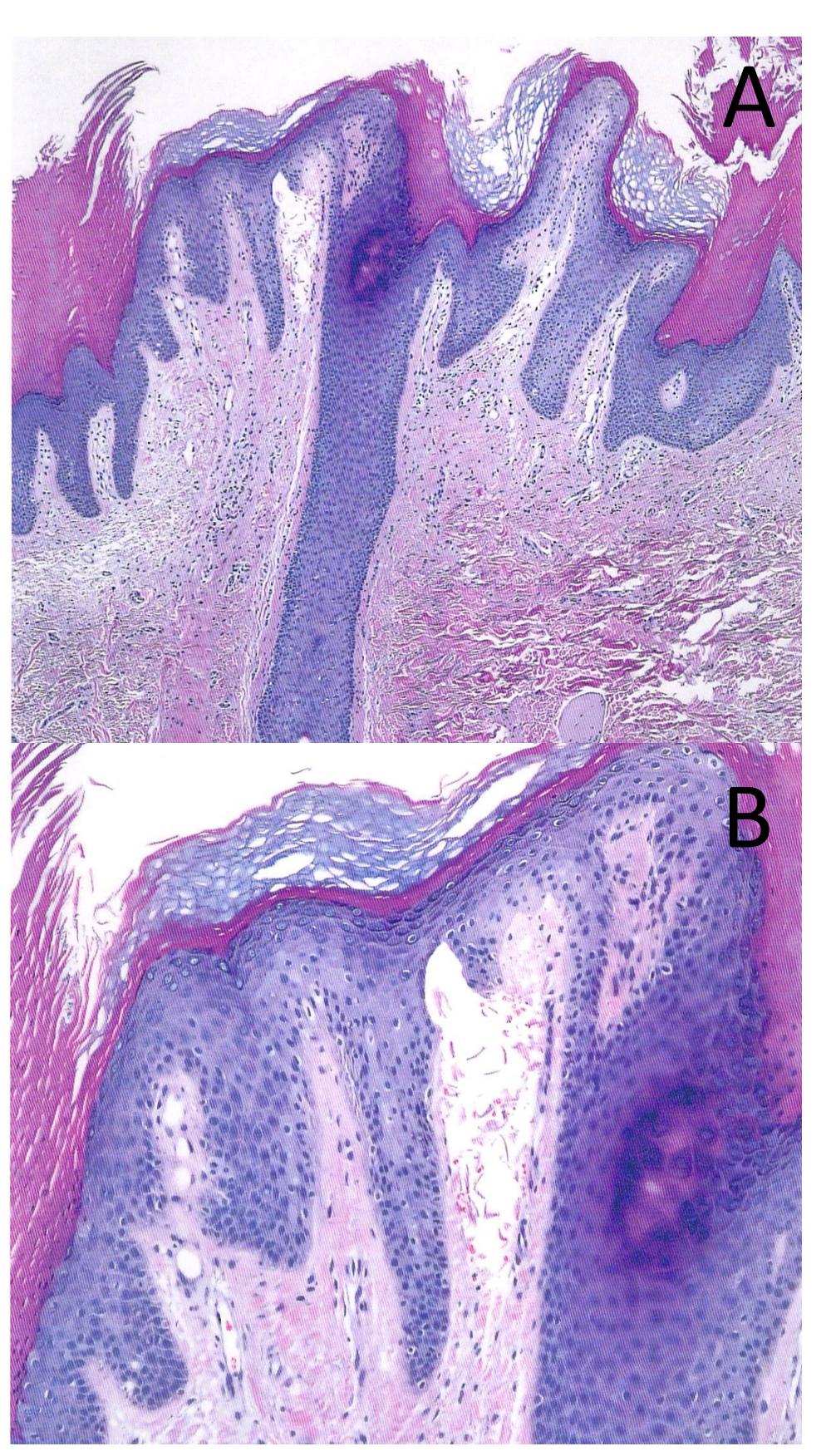
A 74-year-old male with a history of psoriasis, eczema, and basal cell carcinoma, presented with a pruritic rash affecting the gluteal cleft and groin which he noticed 3 months prior (Figure 1). Physical examination showed dark brown crusting located in the gluteal cleft and groin bilaterally. No similar lesions were found elsewhere on the body. He was initially misdiagnosed with tinea corporis at which time a fungal culture was taken, and the patient was prescribed topical econazole and hydrocortisone treatment. The culture returned negative, and the patient failed this treatment regimen. At his following visit, he was prescribed betamethasone ointment, and a punch biopsy was taken. His pathology results returned with a diagnosis of porokeratosis ptychotropica. Upon return for discussion of his biopsy results, the patient stated his pruritis was relieved and he reported the condition was no longer bothering him, so he declined treatment with topical 5fluorouracil.

Discussion

The etiology of the disease is not well established, although several risk factors have been named including UV radiation exposure, chemotherapy, renal or liver failure, and immunosuppressive diseases. ²



Figure 1. Patient presentation.



Figures 2A and 2B. H&E slides showing cornoid lamellae beneath which there is acanthosis, hypogranulosis, dyskeratosis and lymphohistiocytic infiltrate in the superficial dermis.

Discussion Continued

Because of its location, symptomology, and morphology, PP is often initially misdiagnosed as psoriasis, tinea corporis or condyloma acuminata, often subjecting patients to multiple failed therapeutic attempts prior to biopsy. ³ The prevalence of PP may be greatly underestimated secondary to its frequency of misdiagnosis. ³ Potential therapies that have been described in the literature with varying levels of success include cryotherapy, topical 5-fluorouracil, imiquimod, topical or systemic retinoids, dermabrasion, and CO₂ laser. ^{2,3} Attempts at therapy are often disappointing, as is the case with other porokeratosis variants. ^{1,4} Clinical signs that point to PP include the propensity for its lesions to form a butterfly shaped scaly plaque perianally with or without satellite lesions, and involvement of the lower extremities, often with slow growth over the course of several years. ^{3,4}

Histopathology reveals a digitate epidermis with numerous columns of parakeratosis overlying epidermal cells where there is a diminished granular layer and a number of dyskeratotic keratinocytes (Figures 2A and 2B). ³ Visualization of these cornoid lamellae predominates as the gold standard of definitive diagnosis of PP. ¹

Conclusion

This rare variant of porokeratosis can mimic the morphology and symptomology of other diseases at first glance. Education about the presentation of porokeratosis ptychotropica and its addition to the list of commonly described variants can help mitigate its frequent misdiagnosis, reducing treatment failure and unnecessary testing.

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