BRIEF ARTICLES

Bilateral Porokeratosis Ptychotropica on the Gluteal Cleft: A Case Report and Review of the Literature

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ABSTRACT

Porokeratosis ptychotropica (PP) is a rare variant of porokoretosis that is distinctive based on its clinical presentation of pruritic, verrucous papules and plaques that often form a “butterfly” shape, commonly located on the perinatal cleft with extension to the buttocks. Similar to other variants of porokeratosis, it is histologically distinguished by the presence of cornoid lamellae. Proper diagnosis is necessary as some studies suggest that PP may predispose to squamous cell carcinoma. Furthermore, there are limited evidence-based treatment options.

We report the case of a 47 year-old-male who presented with a rash on the buttocks and legs for 3 years. Physical exam revealed erythematous, annular, and verrucous plaques on the bilateral periglueteal area and bilateral distal lower extremities. The patient felt that lesions on the legs were disfiguring but otherwise asymptomatic. Biopsy results demonstrated hyperkeratosis and parakeratosis suggestive of cornoid lamellae. Clinical and histologic findings were suggestive of PP. Lesions on the legs were treated with cryotherapy, which resulted in resolution at a 3-month follow-up.

PP remains a diagnostic and therapeutic challenge due to its rarity. No standard of care has been established, though topical calcipotriol, topical imiquimod, topical tretonoin, and cryotherapy have been used with success in the literature. This case highlights unique characteristics of PP in order to aid in early detection and cancer prevention while also describing various treatment modalities.

INTRODUCTION

Porokeratosis comprises a heterogeneous group of disorders of epidermal keratinization. Though the exact etiology is unknown, it is likely due to aberrant terminal differentiation of keratinocytes.[1] Porokeratosis is histologically characterized by the presence of parakeratotic cells within central grooves of the epidermis, called cornoid lamellae. Ultraviolet exposure and immune suppression have been reported as triggers that worsen symptoms and can further promote subsequent development of malignant transformation in these lesions. Numerous subsets of porokeratosis have been described based on clinical and

histological features, including classic porokeratosis of Mibelli, disseminated superficial (actinic) porokeratosis, porokeratosis palmaris et plantaris disseminata, linear porokeratosis and punctate porokeratosis. However, newly described histologic subsets are emerging.[2,3]

Porokeratosis ptychotropica (PP) is a rare variant of porokeratosis, that is distinctive based on its clinical presentation of pruritic, verrucous papules and plaques located most commonly in the perinatal cleft with extension to the buttocks. It was first described in 1995 by Lucker et al, who emphasized this variant's unique predilection for flexural surfaces.[4] Since then, 20 cases have been individually reported. PP is often a diagnostic challenge, as it is a newly described entity and can be misdiagnosed as psoriasis, dermatophyte infection, condyloma acuminatum, or lichen sclerosis.[5] Controversy persists regarding both accurate diagnosis and the most effective therapeutic modalities. Here, we report a case of porokeratosis ptychotropica and a review of treatment options aimed at improving quality of life.

CASE REPORT

A 47-year-old-male presented with a rash on the buttocks and legs for 3 years. Physical exam revealed erythematous, annular, and verrucous plaques on the bilateral periglueteal area and bilateral distal lower extremities (Figure 1). Lesions on the legs were disfiguring and occasionally painful.

A skin biopsy was performed. Histopathological examination showed acanthotic epidermis with columns of parakeratosis. No lymphocytic infiltrate or amyloid deposition was noted (Figure 2A-2B). The clinical and pathological findings were most consistent with a diagnosis of PP. Disfiguring and painful lesions on the legs were initially treated with phototherapy without success and subsequently treated with cryotherapy, which resulted in resolution of pain and a satisfactory cosmetic outcome at a 3-month follow-up.

Figure 1. Original clinical presentation of erythematous plaque located on the bilateral gluteal region.

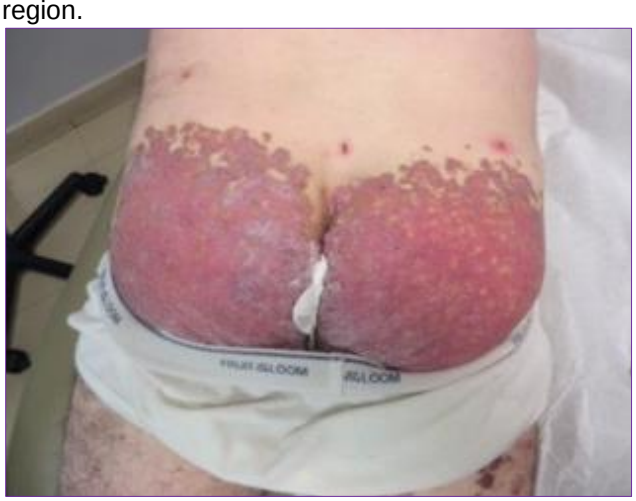


Figure 2A. Original magnification of specimen from gluteal cleft with haematoxylin/eosin stain.

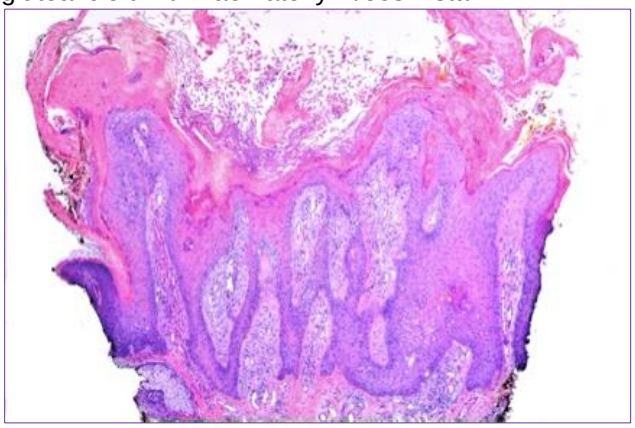
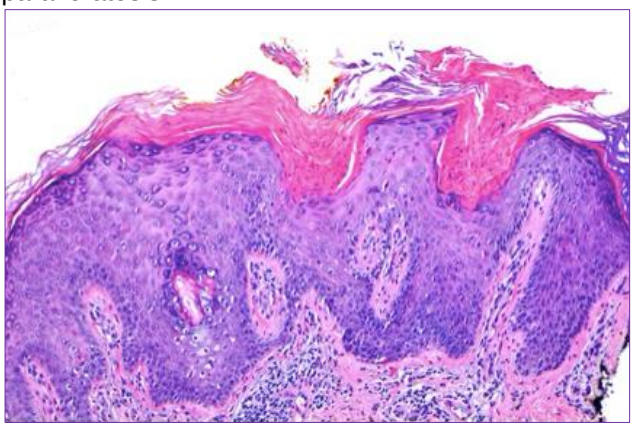


Figure 2B. On higher power, there is an acanthotic epidermis within the stratum corneum, with columns of parakeratosis.



DISCUSSION

The correct diagnosis of PP can be challenging. The differential diagnosis includes psoriasis, dermatophyte infection, and lichen sclerosis. Yet, PP can clinically be differentiated by the characteristic formation of a butterfly-shaped scaly plaque most often located on the buttocks surrounding the anus. Furthermore, the course of symptom development can aid in diagnosis, as this disease has an indolent course with slow growth generally over 5-10 years.[6] Epidemiologic evidence suggests that porokeratosis presents on average in the 5th decade of life with ~90% of cases occurring in males.[6]

Histological hallmarks of PP include cornoid lamella, dyskeratotic epidermal cells, and acantholysis of the basal layer. Lymphocytic infiltrates and amyloid deposition in the papillary dermis have also been reported.[7] While the genetics of all five primary forms of porokeratosis is proposed to be of an autosomal dominant mode of inheritance, the inheritance of PP has not been studied due to the small population affected by this newly recognized disease entity. However, a reported pair of brothers diagnosed with PP has been described and could suggest autosomal dominant inheritance.[6]

Early detection is crucial for proper therapeutic and prognostic purposes as malignant variants of porokeratosis have been noted, with squamous cell carcinoma being the most common cutaneous malignancy.[8] The most common indications for treatment of porokeratosis include pain, pruritis, and cosmetic concerns. Treatment can be challenging, as lesions often relapse, similar to the other five subtypes of porokeratosis. Treatment options include cryotherapy, topical corticosteroids, immunomodulators such as tacrolimus and 5-FU, calcipotriol, and finally retinoids.[7] Kawakami et al recently reported successful treatment of PP of the buttocks with topical 5% imiquimod treatment with resolution of lesions at the 12 month follow-up point.[5] Photodynamic therapy and methyl aminolevulinic acid were also reported to induce remission of pruritis and hyperkeratosis in two men with PP, with resolution of pruritis and hyperkeratosis for as long as 8 and 12 months respectively.[9] However photodynamic therapy was not successful in treatment of our patient. Additionally, successful induction of remission for 2 years was noted after treatment with an electric dermatome device.[10] Similarly, surgical excision was noted to result in remission for genitogluteal PP for 9 years.[11] Carbon dioxide lasers have also been used, however the few case reports that exist in the literature suggest that there is minimal long-term remission with recurrence of disease occurring shortly after treatment.[12]

CONCLUSION

In summary, PP remains a challenging diagnosis, as it is rare, newly described, and

bears semblance to several other dermatological conditions. Furthermore, PP lesions tend to recur and may be difficult to treat, with no established standard of care. We report the safe use of cryotherapy to relieve symptomatic PP lesions and highlight several other treatment modalities that can improve patient quality of life.

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