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Pink verrucous plaque in a man with systemic mastocytosis

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Abstract

Porokeratosis ptychotropica is a rare and commonly misdiagnosed subtype of porokeratosis involving the body folds. We present a 53-year-old man with systemic mastocytosis who presented with a pruritic, verrucous plaque in the gluteal fold that showed multiple cornoid lamellae on histopathologic evaluation, diagnostic of porokeratosis ptychotropica. Various treatments have been reported, including topical corticosteroids, retinoids, vitamin D analogs, calcineurin inhibitors, imiquimod, phototherapy, cryotherapy, or ablative laser therapy, but recurrences are common.

Keywords: porokeratosis, porokeratosis ptychotropica, verrucous plaque, cornoid lamellae

Introduction

Porokeratosis is a disease with abnormal keratinization characterized by the histologic finding of a cornoid lamella. Variants of porokeratosis include porokeratosis of Mibelli, porokeratosis plantaris, palmaris, et disseminata, linear porokeratosis, and disseminated superficial actinic porokeratosis [1]. Porokeratosis ptychotropica is a rare variant of porokeratosis that characteristically occurs in the body folds, most commonly gluteal, and shows multiple cornoid lamellae on histological examination.

Case Synopsis

A 53-year-old man with a history of systemic mastocytosis presented with a 1.5cm pink verrucous plaque in the left gluteal crease that developed five

months prior to presentation (**Figure 1**). His medications included imatinib, cetirizine, and ranitidine. The patient started on imatinib approximately 10 months prior to histologic diagnosis. The patient described the plaque as pruritic and irritated when rubbed. A shave biopsy was performed and histologically examined (**Figure 2**). Multiple angulated columns of parakeratosis with underlying hypogranulosis and dyskeratotic keratinocytes characteristic of cornoid lamellae were present. Additionally, papillated epidermal hyperplasia and a mild lymphocytic infiltrate were seen in the dermis.

Case Discussion

Unlike other variants of porokeratosis, porokeratosis ptychotropica is most commonly found within the



Figure 1: Verrucous, erythematous 1.5cm plaque within gluteal folds. The pen marks delineate the lesion from non-affected skin.

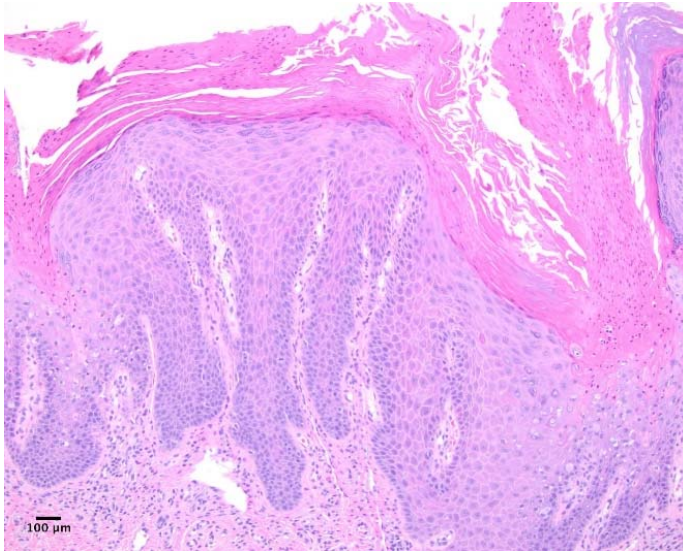


Figure 2: Multiple cornoid lamellae with papillated epidermal hyperplasia and a mild lymphocytic infiltrate. H&E, 20 \times .

gluteal folds, though rarely found in the genital region. It is typically a pruritic eruption of erythematous red-brown papules and verrucous plaques within the gluteal fold, but its presentation can be polymorphic [2-4]. Clinically, porokeratosis ptychotropica is often misdiagnosed as an inflammatory disease such as psoriasis, intertrigo, acrodermatitis enteropathica, or necrolytic migratory erythema [2]. Smaller or solitary lesions can be mistaken for condyloma acuminatum, seborrheic keratosis, or prurigo nodule [5]. Additional diagnoses to consider for genital lesions are squamous cell carcinoma in situ, extramammary Paget disease, granuloma annulare, lichen planus, and lichen sclerosus [4]. To differentiate between these lesions, it is helpful to recognize the predilection of porokeratosis ptychotropica for the gluteal folds, often forming a butterfly appearance. In addition, porokeratosis ptychotropica tends to show slow growth and male predominance in genital involvement [4, 5]. Common histological findings seen in all variants of porokeratosis include a cornoid lamella, but multiple cornoid lamellae are characteristic of porokeratosis ptychotropica.

The pathogenesis of porokeratosis remains poorly understood, but repeated trauma and immune dysregulation may play a role [4]. Both porokeratosis

and psoriasis share an increase in gene expression of keratin 16, keratin 6, and serine/protease inhibitors, among others. [1]. Immunosuppression may also play a role, as a subset of cases of porokeratosis is associated with immunosuppression [6-8]. These include a case of porokeratosis within the genitogluteal region associated with human immunodeficiency virus [4]. Although other variants of porokeratosis may show possible drug association, this has not yet been described in porokeratosis ptychotropica [9, 10]. Malignancy arising within porokeratosis is rare and careful monitoring of these lesions is prudent [4, 8].

Porokeratosis ptychotropica often responds to treatment poorly. Topical corticosteroids, retinoids, vitamin D analogs, calcineurin inhibitors, imiquimod, phototherapy, cryotherapy, or ablative laser therapy have all failed to demonstrate significant efficacy [3]. Topical corticosteroids and topical retinoids may provide symptomatic relief for some patients and topical 5-FU may reduce the thickness of the plaques [3]. Excision can be curative, but often not a feasible option given the size and location.

Conclusion

Porokeratosis ptychotropica is a rare subtype of porokeratosis most commonly occurring in the gluteal fold. It may resemble other common inflammatory or infectious entities such as psoriasis or condyloma acuminatum, but characteristic multiple cornoid lamellae on histological examination enable establishing a correct diagnosis. Unfortunately, treatment is often unsuccessful and relapses frequently occur.

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Potential conflicts of interest

The authors declare no conflicts of interests.

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