CASE REPORT

Genitogluteal porokeratosis - Case report*

Poroqueratose genitoglútea - Relato de caso

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Abstract: We report the case of a patient diagnosed with genitogluteal porokeratosis, a disorder of epidermal keratinization. The location described is extremely rare and very often late diagnosed or even misdiagnosed. Histopathology showed a typical cornoid lamella of great value to support this diagnosis. The importance of awareness of this entity by the specialist is emphasized as a differential diagnosis among genital diseases of chronic evolution and difficult treatment.
Keywords: Buttocks; Genitalia; Parakeratosis; Porokeratosis

Resumo: Relata-se o caso de um paciente com diagnóstico de poroqueratose genitoglútea, uma desordem da queratinização epidérmica, cuja localização exclusiva é extremamente rara, sendo muitas vezes tardia ou erroneamente diagnosticada. A histopatologia demonstra a clássica lamela cornóide, de grande valia para elucidação diagnóstica. Ressalta-se a importância do conhecimento desta entidade pelo especialista como diagnóstico diferencial entre as afecções genitais de evolução arrastada e de difícil tratamento.
Palavras-chave: Genitália; Nádegas; Paraceratose; Poroceratose

INTRODUCTION

Porokeratosis is a primary disorder of epidermal keratinization of unknown aetiology. It presents localized and generalized forms.[1-9] It is thought that the different forms of porokeratosis are phenotypic expressions of a common genetic disorder. Autossomal dominant inheritance of several clinical variants has been reported, as well as sporadic adult-onset cases associated with immunosuppression, AIDS, kidney or liver transplantation, drugs such as thiazide diuretics and immunodeficiency, hematologic malignancies, autoimmune diseases, and occupational exposure to benzene.[2-5] The involvement of the genital region and extension to adjacent areas occurs in both forms (localized or generalized), and localized presentation confined to these regions is extremely rare, with few cases reported in the literature.[2-7] The skin lesions are typically characterized by erythematous to brownish papules and plaques with elevated borders, single or multiple.[1] Histopathological examination demonstrates the classic cornoid lamella. The clinical exuberance, rarity and involvement of unusual locations led us to the presentation of this case.

CASE REPORT

Male patient, 37 years old, complained of the onset of skin lesions on the groin and buttocks for approximately two years accompanied by intense local itching. The dermatological examination showed erythematous papules and plaques with elevated and well-defined borders on the scrotum; brownish macules and papules on the groin and thighs; well-defined brownish erythematous keratotic papules and annular plaques on the buttocks and gluteal cleft (Figures 1, 2 and 3). The lesions started located on the scrotum and groin, with progression to the buttocks region (gluteal cleft). Currently stable. The patient

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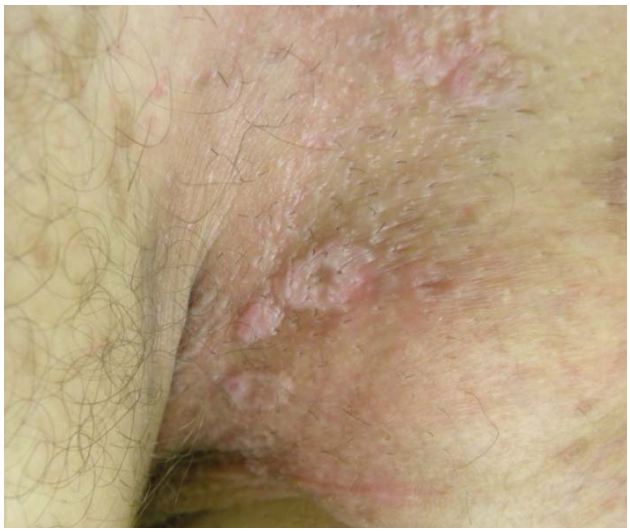

Figure 1: Scrotum - Papules and plaques with elevated and well-defined borders

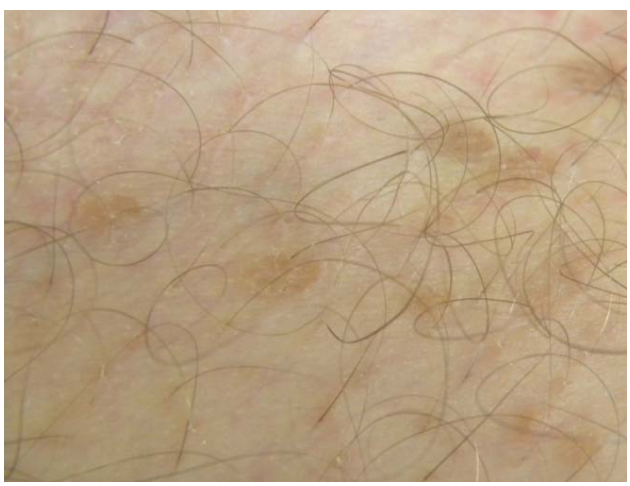

Figure 2: Groin - brownish macules and papules

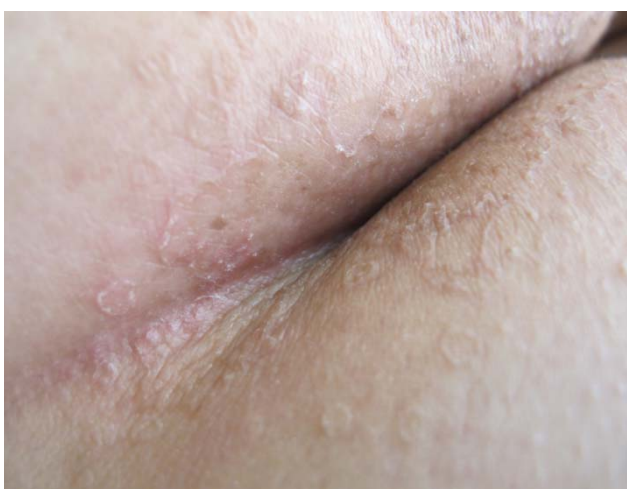

Figure 3: Buttocks and gluteal cleft - well-defined brownish erythematous keratotic papules and annular plaques

received previous treatment with systemic and topical antifungal drugs, isolated or combined topical corticosteroids, without clinical improvement. No significant personal or familiar pathological history. The previous topical therapy was discontinued, and an incisional biopsy of a papule of the left scrotum was performed. The histopathological examination showed superficial perivascular dermatitis with columns of parakeratosis, consistent with the diagnosis of porokeratosis (Figure 4).

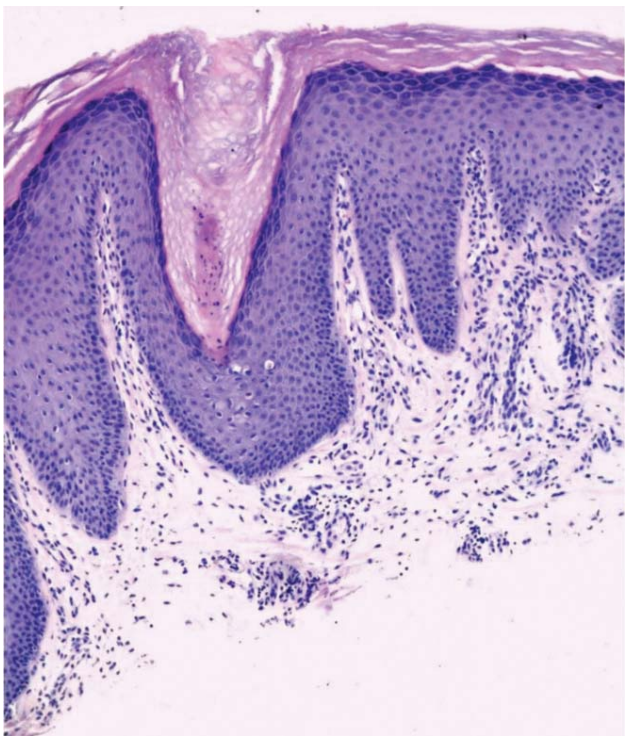

Figure 4: Underlying dyskeratotic cells and absence of the granular layer (HE, 100x)

DISCUSSION

Porokeratosis is a primary disorder of epidermal keratinization of unknown aetiology. It can be classified into localized and generalized forms.[1-9] The localized forms include porokeratosis of Mibelli, linear porokeratosis, and punctate porokeratosis, and the generalized variants include disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis, and disseminated palmoplantar porokeratosis.[1,2,7-9]

Porokeratosis involving the genital region occurs in generalized forms or localized confined to the genital area and adjacent sites (buttocks, perineum, groin and thighs), being the latter an extremely rare presentation, generally classified as a plaque-type porokeratosis of Mibelli, with 23 cases reported in the literature. Based on published reports, the skin

lesions are typically characterized by well-defined brownish erythematous keratotic papules and annular plaques single or multiple. The center of the lesion can be hyperpigmented or hypopigmented and atrophic. They can be asymptomatic or with severe local itching.[1,2,7] There is a predominance of males, and possible increased incidence may be observed in Asian and African American populations.[1-5, 7] The pathogenesis of this entity remains uncertain and both genetic and environmental components have been implicated. Autosomal dominant transmission and associated conditions such as local and systemic immunosuppression have been reported.[2,3,7] Despite presenting a typical cutaneous manifestation the diagnosis is delayed because of the rarity of location, and often misdiagnosed as sexually transmitted disease. The main differential diagnoses are condyloma acuminatum, syphilis (condiloma latum), granuloma annulare, lichen simplex chronicus, extramammary Paget disease and eczema. The skin biopsy is essential for the diagnosis.[4] Histologically, the characteristic alteration is called cornoid lamella, a column of parakeratotic cells in the epidermis.[2-4]

A variant called porokeratosis ptychotropica, that was initially described in 1995 as a pruritic eruption of the gluteal cleft, typically involves the buttocks and gluteal cleft mimicking an inflammatory disorder and is an important differential diagnosis in genitogluteal porokeratosis.[7] Although affecting similar locations, with clinical features that can be histologically superimposable, the ptychotropic form presents an unique feature that is the presence of multiple cornoid lamellae[7,8], not detected in this case. Therefore we preferred the term genitogluteal porokeratosis to encompass both the clinical and histopathological findings of this case.

The elective treatment is the surgical treatment, depending on the number and the size of the lesions. Other treatment options include cryotherapy (liquid nitrogen), CO2 laser therapy, oral and topical retinoids, topical vitamin D3 analogs, keratolytic agents, 5 - fluorouracil - occlusive and more recently with topical imiquimod 5% and topical diclofenac sodium 3% gel - with symptomatic and stabilizing effects on the lesions, with emphasis on its use in genital or genitogluteal porokeratosis. Photodynamic therapy is also described with different results. Regular monitoring is necessary because of the potential for malignant transformation, although there are no reports yet in the literature.[1-7]

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REFERENCES

1. Chen TJ, Chou YC, Chen CH, Kuo TT, Hong HS. Genital porokeratosis: a series of 10 patients and review of the literature. Br J Dermatol. 2006;155:325-9.
2. Valdivielso-Ramos M. Genital porokeratosis. Actas Dermosifiliogr. 2008;99:217-20.
3. Sengupta S, Das JK, Gangopadhyay A. Porokeratosis confined to the genital area: a report of three cases. Indian J Dermatol Venereol . 2008;74:80.
4. Liang C, Batra P, Patel R, Kamino H. Genital porokeratosis. Dermatol Online J. 2009;15:23.
5. Sertznig P, Felbert VV, Megahed M. Porokeratosis: present concepts. J Eur Dermatol Venereol. 2012;26:404-12.
6. Marks S, Varma R, Cantrell W, Chen SC, Gold M, Muellenhoff M, et al. Diclofenac sodium 3% gel as potential treatment for disseminated superficial actinic porokeratosis. J Eur Acad Dermatol Venereol. 2009;23:42-5.
7. Wanat KA, Gormley RH, Bennett DD, Kovarik CL. Genitogluteal porokeratosis involving the scrotum: an unusual presentation of an uncommon disease. J Cutan Pahtol. 2012;39:72-4.
8. Malek J, Chedraoui A, Kibbi AG, Ghosn S. Genitogluteal Porokeratosis: 10 years to make the diagnosis. Am J Dermathol. 2009;31:604-6.
9. Gon AS, Rocha SRT, Pozetti EMO,; Rodrigues VSM, Antonio JR. Porokeratosis: a clinical study of fifteen cases. An Bras Dermatol. 1993;68:271-5.

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