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CASE REPORTS

Genital Porokeratosis

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Abstract. *Introduction.* Porokeratosis is a primary disorder of epidermal keratinization. The term covers several clinical variants that have in common the presence of a cornoid lamella in histological studies. Although porokeratotic lesions may appear anywhere on the skin, genital lesions are uncommon and may occur in cases of generalized porokeratosis with genital involvement or be localized to the genital area.
Case description. We describe a 47-year-old man with a solitary porokeratotic plaque on the scrotum. He had no other lesions at other sites or relevant personal or familial history.
Discussion. Porokeratosis confined to the genitals is extremely uncommon. Only 23 cases have been reported in the literature. We undertook a clinical, epidemiological, and therapeutic review, compiling the distinctive characteristics of this rare entity.

Key words: porokeratosis, genital porokeratosis, review, distinctive characteristics.

POROQUERATOSIS GENITAL

Resumen. *Introducción.* La poroqueratosis es un trastorno primario de la queratinización epidérmica que engloba varias formas clínicas que comparten una histología común característica, definida por la aparición de la laminilla cornoide. Aunque las lesiones de poroqueratosis pueden encontrarse en cualquier parte de la superficie corporal la afectación del área genital ocurre raramente, y puede presentarse en casos de poroqueratosis generalizada que incluyan el área genital, o de forma exclusiva en esta localización.
Caso clínico. Presentamos el caso clínico de un varón de 47 años de edad con una placa única de poroqueratosis escrotal, sin lesiones en otras localizaciones, ni antecedentes médicos personales ni familiares de interés.
Discusión. La poroqueratosis de localización exclusiva genital es extremadamente infrecuente, con tan sólo 23 casos recogidos en la literatura. Realizamos una revisión clínica, epidemiológica y terapéutica, recogiendo las características distintivas de esta rara entidad.

Palabras clave: poroqueratosis, poroqueratosis genital, revisión, características distintivas.

Porokeratosis is a primary disorder of epidermal keratinization. The term covers several clinical variants that have in common the characteristic histologic finding of a cornoid lamella, which is defined by the presence of a thin column of parakeratotic cells extending through the stratum corneum, loss of the underlying stratum granulosum, and the presence of dyskeratotic or vacuolated cells in the stratum spinosum. The localized variants include classic porokeratosis or porokeratosis of Mibelli, linear porokeratosis, and punctate porokeratosis, and the extensive variants include disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis, and disseminated palmoplantar porokeratosis. It is thought that these are different phenotypic expressions of a common genetic disorder.[1] Autosomal 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, hematologic malignancies, autoimmune diseases, and occupational exposure to benzene, among others.[1-3] Some authors have suggested that porokeratosis results from the expansion of a clone of abnormal keratinocytes, while mosaicism may explain the localized or linear variants of porokeratosis.[4]

Although porokeratosis lesions may be located on any part of the body surface, their appearance in the genital area is considered extremely rare. They may occur in cases of generalized porokeratosis that includes the genital area or be present only in this location.[5] Here I report a new case of porokeratosis of the scrotum and review the cases of porokeratosis confined to the genital area that have been published to date.

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Table 1. Summary of the Published Cases of Genital Porokeratosis

Authors	*Sex, Age*	*Time Since Onset*	*Symptoms*	*Number*	*Location*
Levell et al[10]	Man, 27 y	2 y	Asymptomatic	3	Penis, scrotum, perineal raphe
Neri et al[5]	Man, 70 y	Years	Asymptomatic	1	Scrotum
	Man, 40 y	2 y	Asymptomatic	1	Penis
Tangoren et al[6]	Man, 75 y	2-3 y	No data	3	Penis
Porter et al[11]	Man, 56 y	2 y	Asymptomatic	1	Glans
Laino et al[4]	Man, 36 y	3 y	No data	Multiple	Scrotum
Huang et al[9]	5 men, 29-66 y, 1 woman, 43 y	1-9 y	No data	1-3 lesions	Scrotum, natal cleft (men), natal cleft (woman)
Perlis et al[12]	Man, 64 y	Years	Pruriginous	1	Penis
Chen et al[8]	10 men, 39-59 y	0.8-15 y	Pruriginous	1-several	Scrotum, penis, buttocks
Valdivielso-Ramos, 2007[a]	Man, 47 y	1.5 y	Pruriginous	1	Scrotum

[a]This study.

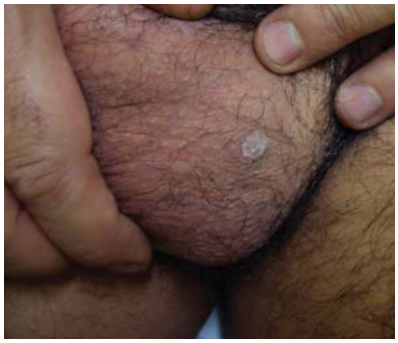

Figure 1. Well-defined uniform hyperkeratotic lesion on the left side of the scrotum

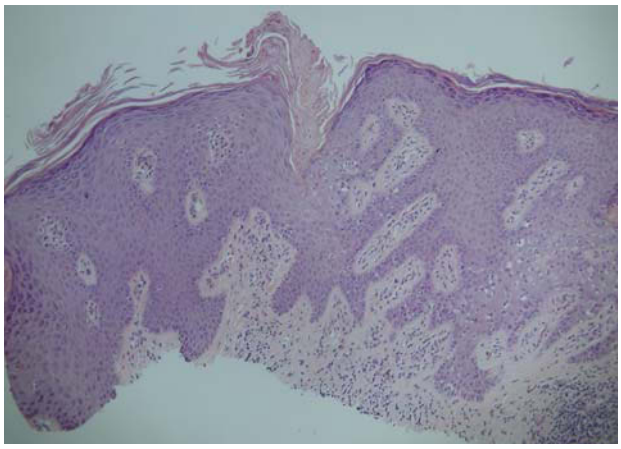

Figure 2. Cornoid lamella seen by histopathology (hematoxylin-eosin, ×20).

Case Description

A 47-year-old man was referred to our clinic with a pruriginous lesion on the scrotum that had appeared 18 months previously and that had not improved despite topical antifungal and corticosteroid treatment. He had no relevant medical history and was not receiving any medication; he worked as an administrative assistant.

Physical examination showed a very well-defined, uniform, hyperkeratotic plaque, 10 mm in diameter, located on the left side of the scrotum (Figure 1). Thorough exploration of the rest of the body surface revealed no other lesions. The patient reported no family history of porokeratosis.

A skin biopsy was performed showing several columns of parakeratotic cells in the epidermis and loss of the underlying stratum granulosum, with some keratinocytes displaying cytoplasmic vacuolation (Figure 2). This led to a diagnosis of porokeratosis of the scrotum.

As the lesion was small, we decided in agreement with the patient that the best course of action was surgical removal. No recurrence of the lesion was observed during a follow-up period of 1 year.

Discussion

The classic localized variant, porokeratosis of Mibelli, is rare and is found during the breastfeeding period or infancy, especially in male patients. Onset tends to be later in nonhereditary cases.[1] It can affect any part of the body, but has particular tendency to occur unilaterally on the extensor surface of the limbs. It has been described on the face, mucosa, and, very rarely, in the genital and paragenital region.[6]

As mentioned, typical symptoms include single or a few annular reddish-brown plaques, with a conspicuous

Family History	*Drugs*	*Treatment*	*Immunosuppression*	*Other*
No	No data	Cryotherapy	No data	Black race
No	No	No	No data	
No	No	No data	Ungual lichen planus	
No data	No data	Cryotherapy	No data	
No data	No	5-fluorouracil	No data	
No	No	No data	Decreased CD4+/CD8 + lymphocytes	Black race
	No data	Surgery, CO2 laser	No data	
No data	No	Triamcinolone	No data	Korean
No data	Oral hypoglycemic therapy in 3 patients	Surgery, CO2 laser, other	No data	3 patients diabetes mellitus, 1 genital warts, 1 syphilis
No	No	Surgery	No	

hyperkeratotic raised narrow margin, bordered by a characteristic longitudinal groove. The center of the lesion can be hypopigmented or hyperpigmented, hairless, atrophic, and anhydrotic. They steadily increase in size from the center outward and giant variants have been described. They are usually asymptomatic, although intense pruritus can occur.

Dermatoscopy has recently established the presence of a thin white or yellowish annular structure, sometimes double, that demarcates the central area of the lesion, where there is a pinkish-white scar-like area surrounded by slightly vascularized tissue.[7]

A review of the literature showed that only 23 cases of porokeratosis confined to the genital area have been described (Table 1). Chen et al[8] published the largest case series, which included 10 patients, of whom only 7 presented porokeratosis confined to the genital area. In that series, 30% had non-insulin-dependent diabetes mellitus and 2 patients had associated sexually transmitted diseases (one with genital warts and the other with syphilis). Huang et al[9] described 6 cases of genitogluteal porokeratosis in 5 men and 1 woman, without finding any associated disease. Neri et al[5] reported 2 new cases. The remainder consisted of isolated cases.[4,6,10-12] Helfman and Poulos[13] reported a case of reticulated lesions on the penis and scrotum that extended into the suprapubic area, groin, and thighs.

Genital porokeratosis has traditionally been included within porokeratosis of Mibelli or as a plaque form, although all the reported cases share some differential characteristics that suggest defining genital or genitogluteal porokeratosis as a distinct clinical variant.[8] Almost all reported cases have occurred in men. Only 1 case of vulvar disease has been reported, although the patient also presented other lesions on the perineum, a limb, and the sole of the foot.[14] Huang et al[9] described another woman with gluteal lesions. The lesions reported in the different studies appeared in middle age and were small. Except for the case published by Laino et al,[4] where there was an asymptomatic decrease in the percentage of $CD4^+/CD8^+$ lymphocytes, the individuals were not immunocompromised nor were they taking relevant medication. Although all the patients had typical lesions, it was difficult to establish a correct initial diagnosis, as due to its rarity the disease was not suspected during the differential diagnosis of the lesions in the genital region. There was no family history of porokeratosis in the cases described and there has been no report of malignancy during follow-up or of lesion progression after initial rapid growth (Table 2).

Table 2. Characteristics of Genital Porokeratosis

Almost exclusively found in men
Late onset
Single or few lesions, small size
No family history of porokeratosis
No immunosuppression (only 1 case of asymptomatic decrease in CD4+ lymphocytes)
No relevant associated drugs
No malignancy
Late diagnosis (due to rare location)

Lucker et al[15] described a clinical variant restricted to the natal cleft in a 34-year-old man. They attempted to clinically differentiate it as a porokeratosis confined to the cleft, designating it porokeratosis ptychotropica.

Subsequently, 3 more similar cases were described, although due to their distinctive characteristics they are not included in the definition of genital porokeratosis used here. Trcka et al[2] reported a case of a 70-year-old man with approximately 40 lesions on the scrotum, the natal cleft, and the upper region of both thighs, associating this with chronic occupational exposure to benzene. As these lesions extended beyond the genital area the case was not included in the series of genital porokeratosis.

Malignant porokeratotic lesions have been well documented in the literature, occurring in between 7% and 11% of cases. The risk of malignancy is greater in larger lesions that have been developing over a longer period on skin not exposed to sunlight, in elderly and immunocompromised patients, and especially in the clinical forms of linear porokeratosis and porokeratosis of Mibelli.[1,8] Intriguingly, some studies have shown that the localized variant that arises in adults has a higher potential (24%) to become malignant. However, no case of malignancy has been reported in porokeratotic lesions confined to the genital area.

Porokeratotic lesions must be treated due to their potential for malignant transformation, as outlined above. The treatment of choice is surgery, although this may at times be technically challenging due to the size, number, and location of the lesions. Several therapeutic options have been described, such as cryotherapy,[6,10] CO_2 laser therapy,[2,9] oral retinoids, and topical treatment with vitamin D3 analogs, keratolytic agents, 5-fluorouracil under occlusion,[11] and, more recently, imiquimod under occlusion,[7] and photodynamic therapy, with variable results. Patients should be warned to avoid exposure to UV light.

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Conflicts of Interest

The author declares no conflicts of interest.

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