Journal of Pakistan Association of Dermatologists. 2017;**27 (1)**:95-96.

Case Report

Genital porokeratosis: A rare entity

Kikkeri Narayanshetty Naveen, SB Athaniker, D Joshika Bhandary, Rahul R Shetty, MD Chandan, Anitha S*

Department of Dermatology, SDM College of Medical Sciences and Hospital, Sattur, Dharwad, Karnataka, India.
* Department of Pathology, SDM College of Medical Sciences and Hospital, Sattur, Dharwad, Karnataka, India.

Abstract Porokeratosis involving the genitalia can occur as part of generalized porokeratosis with genital involvement or as localized porokeratosis that is confined to the genital area. Porokeratosis confined to the genitalia (genital porokeratosis) is considered as an extremely rare entity and is classified as classic or plaque-type porokeratosis of Mibelli. Here, we report a 42-year-old male presenting with the itchy genital porokeratosis. This case has been presented for its classical presentation and rarity.

Key words
Genital, porokeratosis, itch.

Introduction

Porokeratoses (PK) are characterized by marginate scaling lesions, histologically showing a column of parakeratotic keratinocytes (the cornoid lamella). Five different forms of porokeratosis have been described i.e., classical plaque type porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis, palmoplantar porokeratosis and Giant porokeratosis.[1] Porokeratosis involving the genitalia can occur as part of generalized porokeratosis with genital involvement or as localized porokeratosis that is confined to the genital area. Porokeratosis confined to the genitalia (genital porokeratosis) is considered as an extremely rare entity and is classified as classic or plaque-type porokeratosis of mibelli.[2]

Here, we present a case of genital porokeratosis for its classical presentation and rarity.

Address for correspondence
Dr. Kikkeri Narayanasetty Naveen
Professor, Department of Dermatology, No 10, Skin OPD, Sri Dharmasthala Manjunatheshwara College Of Medical Sciences & Hospital (SDMCMS&H)
Sattur, Dharwad – 580009, India
Email: naveenkn80@yahoo.com

Case Report

A 42-year-old married male presented with lesions on the scrotum and inner aspect of thighs with itching since 1 month. There was no history of genital ulcer, inguinal swelling, or deviant sexual history. No similar complaints in the family or spouse.

Examination revealed well-defined hyperpigmented annular plaques with central atrophy and raised border on the scrotum and inner aspect of the thighs **(Figure 1)**. No such lesions were found on other areas of the body and systemic examination was normal.

Histopathology of skin specimen taken from the edge of lesion on inner thigh revealed epidermis with hyperkeratosis and parakeratosis along with deep invaginations of epidermis filled with keratin. These invaginations show parakeratotic column. Keratinocytes beneath parakeratotic column were irregular in arrangement. Dermis presented with sparse lymphoid infiltrate. **(Figure 2)**. The histopathology was consistent with the diagnosis of porokeratosis.

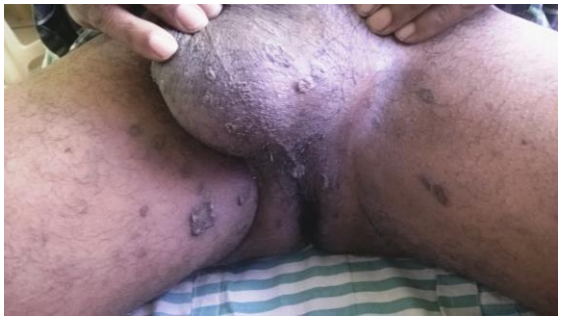

Figure 1 Hyperpigmented annular plaques with central atrophy and raised border on the scrotum and inner aspect of the thighs

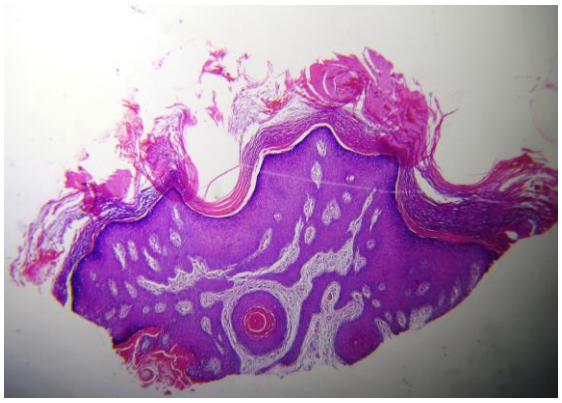

Figure 2 Histopathology shows epidermis with hyperkeratosis and parakeratosis along with deep invaginations of epidermis filled with keratin. These invaginations show parakeratotic column. Keratinocytes beneath parakeratotic column were irregular in arrangement (H&E 10x).

Discussion

Genital porokeratosis (GP) is a very rare skin condition and a very few reports have appeared in Indian dermatological literature. PK is two to three times more common in males than in females, but GP in females seems even rare. Robinson *et al.*[3] gave the first report of vulvar PK in a 39-year-old lady who had disfiguring lesions in the perineal area, medial thigh and sole. PK localized to the male genitalia has been reported most commonly in the scrotum, followed by penis, buttock, natal cleft, groins and adjacent thighs.[2] In the present case, PK was found on the scrotum and inner aspect of thigh. PK is an autosomal dominant disorder, but in our case none of the family members were affected by this disorder. Similarly, family history was noncontributory in all the 10 cases studied by Chen *et al*.[2]

Porokeratosis in general is not pruritic; however, pruritus was a presenting feature in most cases in the series of genital porokeratosis reported by Chen *et al*.[2] and was attributed to a hot and humid climate and repetitive trauma due to friction and scratching. In the present case, patient had intense itching and it was the primary symptom.

The differential diagnosis considered was annular lichen planus; however, the histopathological examination confirmed the diagnosis of PK.

Several therapeutic options have been described, such as cryotherapy, CO_2 laser therapy, oral retinoids, and topical treatment with vitamin D3 analogues, keratolytic agents, 5-fluorouracil under occlusion, and, more recently, imiquimod under occlusion, and photodynamic therapy, with variable results.[2]

References

1. Judge MR, McLean WHI, Munro CS. Disorders of keratinization. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's textbook of dermatology. Oxford: Wiley-Blackwell; 2010. P.19.90-19.91
2. Chen TJ, Chow 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.
3. Robinson JB, Im DD, Jockle G, Roshenshein NB. Vulvar porokeratosis: Case report and review of literature. *Int J Gynecol Pathol*. 1999;**18**:169-73.