

Genital Porokeratosis: A Case Report of Rarely Involved Sites

. Anup Kumar Tiwary¹, Dr. Firdous Jahan², Dr. Dharmendra Kumar Mishra³

1. Junior Resident; Department of Dermatology, Venereology and Leprosy, RIMS, Ranchi, India.

2. Senior Resident, Department of Dermatology, Venereology and Leprosy, RIMS, Ranchi, India.

3. Associate Professor; Department of Dermatology, Venereology and Leprosy, RIMS, Ranchi, India.

Abstract: Genital porokeratosis is a rare form of porokeratosis which can occur in two ways : in isolated form or as a part of disseminated porokeratosis. The former one is much rarer and less than 50 such cases of isolated involvement of genitalia in porokeratosis has been reported so far. Being a inherited disorder of keratinization, usual mode of heritance is autosomal dominant. The aim of present report is to document another case of genital porokeratosis in a 55 years old male with negative family history.

Keywords: autosomal dominant, disseminated, genital porokeratosis

I. Introduction

Porokeratosis (PK) is an inherited, heterogeneous group of clonal disorder of keratinization presenting as single or multiple characteristic annular lesions which are surrounded by raised, sharply marginated, keratotic borders with central atrophy and histopathologically characterized by 'cornoid lamella'. PK has several types: including classical plaque form, porokeratosis of mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis, porokeratosis palmaris et plantaris disseminata and punctate porokeratosis [1]. Apart from these forms, localized involvement of genital area may rarely occur as it has been reported earlier in literature. To the best of our knowledge, only less than 50 cases of genital porokeratosis are reported in the literature till now [2].

II. Case Report

A 55 yrs old male came to our outpatient department of dermatology presenting with multiple, mildly pruritic, skin coloured, brownish or hypopigmented lesions on the scrotum, penis and adjacent thighs for the last 2 years. These lesions started appearing as papule which slowly increased in number and size. No spontaneous regression was seen in any of the lesion. There was no history of extramarital sexual exposure, drug intake prior to the development of lesions and family history was also negative. Treatment with topical steroids and antifungals produced no results.

On local cutaneous examination, several hyperpigmented papules and well-defined hyperpigmented annular plaques of varying sizes and depigmented nodules were seen on the scrotum, penile shaft and adjacent thighs [Fig.1] [Fig.2]. Most of the plaques had mildly scaly atrophic center and a raised border and some showed thickened indurated surface with pigmentation without the typical annular ring . A total of around 20 lesions were present at these sites and maximum size was 2 cm x 1.5 cm. No similar skin lesions was present anywhere else on his body. There was no regional lymphadenopathy or any evidence of sexually transmitted disease. systemic examination was normal .Routine hematological and biochemical tests were within normal limits and VDRL and ELISA for HIV were also negative.

To come arrive at the final diagnosis, a tissue specimen was taken from the keratotic ridge of the scrotal lesion with 3 mm punch and sent for histopathological evaluation.



Figure 1: Hyperpigmented annular plaques of varying sizes on adjacent thigh and penile shaft



Figure 2: Hyperpigmented annular plaques and depigmented nodules on scrotum

Histopathological Examination

On histopathological examination of punch biopsy sample, hyperkeratosis and a keratin-filled angulated epidermal invagination was noted. In the center of this keratin-filled invagination, a poorly staining parakeratotic column of stratum corneum cells, 'cornoid lamella' is seen running through the surrounding normal staining cells. Within the parakeratotic column, cells appeared homogeneous with pyknotic nuclei. In the epidermis beneath the parakeratotic column, the keratinocytes are irregularly arranged, spongiotic having pyknotic nuclei with perinuclear edema. No granular layer was found at the site of parakeratotic column [Fig.3].

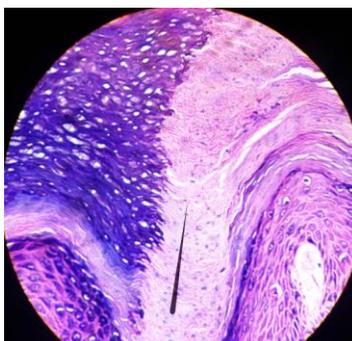


Figure 3: stacked, poorly stained parakeratotic column of stratum corneum cells (H & E, X40)

III. Discussion

Porokeratosis is an inherited, clonal disorder of keratinization presenting as single or multiple characteristic annular lesions having raised, sharply marginated, keratotic ridge with central atrophy and histopathologically showing a vertical column of parakeratotic stacked cells known as 'cornoid lamella'. [1] Its several forms have been recognized: classical plaque form, porokeratosis of mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis, porokeratosis palmaris et plantaris disseminata and palmoplantar porokeratosis of Montoux (punctate porokeratosis). An another type was also reported in Iraq, affecting only the face, called solar facial porokeratosis. [3]

Apart from these forms, genital PK has rarely been reported in literature, more among Asian population. [4] [5]. Genital PK is usually localized to the scrotum followed by penis in male and vulva is the most common genital site in females. [6] [7] The number of lesions are usually scanty. Other sites like buttock, natal cleft, groins and adjacent thighs may also be involved alongwith. [4] [8] In our case, PK was localized to the scrotum and penis and adjacent thighs also. Though the mode of inheritance of PK is known to be autosomal dominant, positive family history is not seen in each and every patient as in our case. [4] Overall, PK is twice more common in males.

Clinical distinct features of PK make it easy to diagnose but sometimes, absence of the thready, keratotic ridge usually leads to other possible differential diagnosis including annular lichen planus, annular syphilide, lichen simplex chronicus and others. [9] In such confusing cases, skin biopsy is done and specimen should be taken from peripheral hyperkeratotic ridge.

Histopathological examination of outer ridge shows hyperkeratosis and a keratin-filled angulated invagination of the epidermis of variable depth depending upon the forms of PK. In the center of this keratin-filled invagination, a poorly staining parakeratotic column of stratum corneum cells, the so-called cornoid lamella is seen running through the surrounding normal staining cells and representing the most characteristic feature of all forms of PK. Within the parakeratotic column, the horny cells appear homogeneous and possess pyknotic nuclei. In the epidermis beneath the parakeratotic column, the keratinocytes are irregularly arranged and have pyknotic nuclei with perinuclear edema. [10] Usually no granular layer is found at the site at which the parakeratotic column arises, but elsewhere the keratin-filled invagination of the epidermis has a well-developed granular layer. A moderate dermal lymphocytic infiltrate under the cornoid lamella, a lichenoid reaction pattern, and the presence of amyloid material may be detected. Many individual dyskeratotic cells were seen under the base of lamella reaching almost the basal layer of epidermis. While the dermis consisted of many dilated blood vessels with severe inflammatory reaction at the base of cornoid lamella, consisting of many lymphoid cells. The risk of malignant changes is very low in porokeratosis but rarely it can transform into squamous or basal cell carcinoma but fortunately, in genital porokeratosis, malignant transformation has not been reported till now. [5] [10] [11]

There are many treatment options but none of them have been proved to be effective so far. Topical steroid, 5%5-FU and imiquimod, cryotherapy, electrocautry, CO2 laser have been tried with variable

results.[12] Among these therapies, Co2 laser has been suggested as a most effective treatment in relieving the patient complaint and to halt the possibility of malignant transformation.

IV. Conclusion

Genital porokeratosis is an under diagnosed entity because it can be easily confused with annular lichen planus, STDs and other non-venereal diseases affecting genitalia. In addition to a thorough clinical evaluation, histopathological examination is mandatory. Moreover, all diagnosed cases should have regular follow-up to look for any malignant transformation.

References

- [1] Spencer LV. Porokeratosis. Available from: http://www.emedicine.com/derm/topic_343.htm. [Last accessed on 2006 Oct 9].
- [2] Valdivielso-Ramo M. Genital porokeratosis: Case Reports. *Actas Dermosifiliológicas*. 2008;99:217-20.
- [3] Sharquie KE, AL-Baghdady BA. Solar facial porokeratosis. *J Dermatol*. 2003;30:216-21.
- [3] 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] Sengupta S, Das JK, Gangopadhyay A. Multicentric squamous cell carcinoma over lesions of porokeratosis Palmaris et plantaris disseminata and giant porokeratosis. *Indian J Dermatol Venereol Leprol* 2005;71:414-6.
- [4] Porter WM, Du P MenagÈ H, Philip G, Bunker CB. Porokeratosis of the penis. *Br J Dermatol* 2001;144:643-4.
- [4] Robinson JB, Im DD, Jockle G, Roshenshein NB. Vulvar porokeratosis: Case report and review of literature. *Int J Gynecol Pathol* 1999;18:169-73.
- [5] Levell NJ, Bewly AP, Levene GM. Porokeratosis of Mibelli on the penis, scrotum and natal cleft. *Clin Exp Dermatol* 1994;19:77-8.
- [5] Lynch PJ. Lichen simplex chronicus (atopic/neurodermatitis) of the anogenital region. *Dermatol Ther*. 2004;17:8.
- [5] Judge MR, McLean WH, Munro CS. Disorders of keratinization. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook Of Dermatology*. 7th ed. Blackwell Science: Oxford; 2004. p. 34.1-34.111.
- [5] Neri I, Marzaduri S, Passarini B, Patrizi A. Genital porokeratosis of Mibelli. *Genitourin Med* 1995;71:410-1.
- [6] Huang SL, Liu YH, Chen W. Genitoglutealporokeratosis. *J Eur Acad Dermatol Venerol*. 2006;20:899-900.