International Journal of Research in Medical Sciences
Sangheetha K et al. Int J Res Med Sci. 2019 Jan;7(1):313-316
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20185400

Case Report

Porokeratosis ptychotropica: a rare case report with unusual presentation

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Received: 23 October 2018
Accepted: 30 November 2018

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ABSTRACT

Porokeratosis is a rare disorder of epidermal keratinization characterized clinically by annular plaque with thread like hyperkeratotic border with a central groove that expand centrifugally and this border corresponds to coronoid lamellae histologically which are the columns of parakeratosis that overlie an epidermal invagination with loss of granular layer and dyskeratosis of upper spinous keratinocytes. The disorder was erroneously named porokeratosis because the coronoid lamella was initially described as being present over a sweat pore, which is a fixed structure that cannot expand peripherally. Five primary clinical variants have been described: classic porokeratosis of mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis, punctate porokeratosis and porokeratosis palmaris et plantaris disseminate. Porokeratosis ptychotropica one of the rare variants of porokeratosis described by lucker et al which has been added recently in the classification. It is characterized clinically by symmetrical verrucous papules and plaques resembling psoriasis plaque in the gluteal cleft, buttocks and rarely extends to genitalia and histologically by multiple coronoid lamella. We report a case of 43year old female, presented with 10years duration of pruritic raised skin lesion over the left gluteal region. Dermatological examination revealed single well defined erythematous scaly plaque with central atrophy, hyperpigmentation and peripheral thread like elevated border. Histopathological examination revealed multiple coronoid lamella which is the hallmark for porokeratosis ptychotropica, confirmed the diagnosis. The patient was treated with 5-fluorouracil cream. we report this case due to its rarity and the unusual presentation of single plaque of porokeratosis ptychotropica.

Keywords: Gluteal cleft, Multiple coronoid lamella, Porokeratosis ptychotropica

INTRODUCTION

Porokeratosis is a morphologically distinct disorder of keratinization characterized clinically by hyperkeratotic papules or plaques surrounded by thread like border that expands centrifugally and has been likened to the great wall of china with a ridge and moat on its summit.[1,2] Mibelli first described the classical form which bears his name in 1893. All forms of porokeratosis have been reported to have familial clusters with autosomal pattern with variable penetrance. Somatic mutations may explain the sporadic occurrence of some cases. Porokeratosis seen predominantly in pale fair skinned ethnic groups.[3]

Though the etiology is not well understood, the various risk factors for the disease include exposure to ultraviolet radiation, infections like HIV, drugs, organ transplantation, mechanical trauma, liver failure and chronic renal failure. The distinctive histopathologic feature coronoid lamella, the hallmark of all clinical variants which are composed of parakeratotic keratinocytes which result from either faulty maturation

or an acceleration of epidermopoiesis. These coronoid lamellae correspond to the raised keratotic border evident clinically.[3]

Porokeratosis ptychotropica is one of the rare and lesser known variants of porokeratosis, with fewer than 50 cases described in the literature so far. It is usually confined to body folds and it starts as brownish to reddish macules or plaques that develop symmetrically over the perianal region. The presence of multiple coronoid lamella as seen histologically explains the keratotic or verrucous appearance and expansile papular growth. Involvement of the genital region and adjacent areas like buttocks, perineum, groin and proximal thigh may occur as a part of generalised porokeratosis, but porokeratosis localized to the genitogluteal region is rare.[2]

CASE REPORTS

A 43-year-old otherwise healthy female presented with 10 years duration of single verrucous asymptomatic lesion over the left gluteal region. The patient observed progressively increased size of the lesion with itching over the past 1year. There were no similar lesions among the family members. On examination, single well defined erythematous scaly pinkish plaque of size 6×4cms with central hyperpigmentation, atrophy (Figure 1) and characteristic thread like keratotic ridge (Figure 2) was seen in the left gluteal region near the natal cleft.

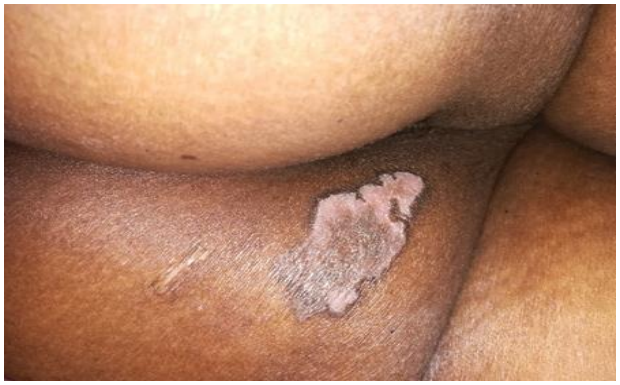

Figure 1: Well defined pinkish scaly plaque with central atrophy and hyperpigmentation present over the left gluteal region near the natal cleft.

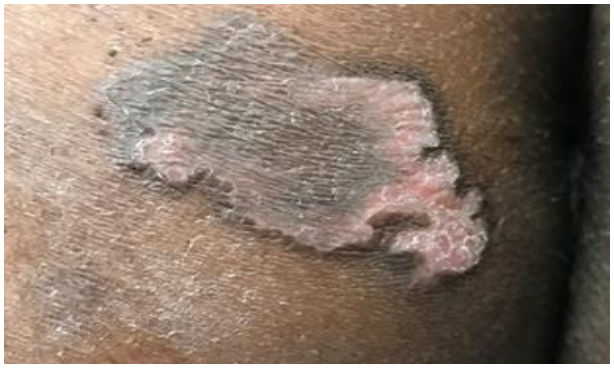

Figure 2: Characteristic thread like keratotic ridge.

Differential diagnosis considered was cutaneous tuberculosis and porokeratosis. Routine blood investigations, chest x ray was normal. Sputum for AFB and mantoux test was negative.

Dermascopy examination of the periphery of the lesion showed irregular double marginated white track border (Figure 3) and central groove on the keratotic ridge (Figure 4). Skin biopsy was taken from the peripheral border of the plaque. Histopathological examination showed multiple focal invagination of the epidermis by keratotic plug with focal reduction in the granular layers and adjacent parakeratosis featuring coronoid lamella (Figure 5 & 6). There was perivascular lymphocytic infiltrate and increased collagen in the dermis. Based on the clinical and histopathological findings, we made the diagnosis of porokeratosis ptychotropica.

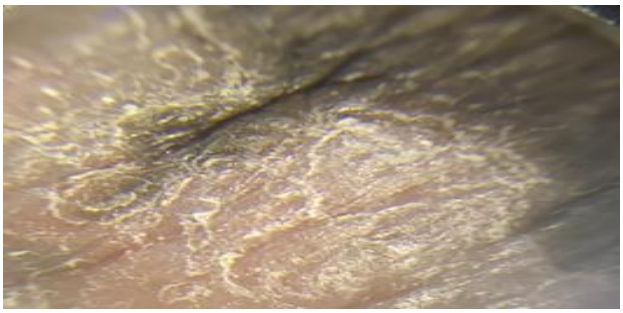

Figure 3: Irregular double marginated white track border on dermascopy.

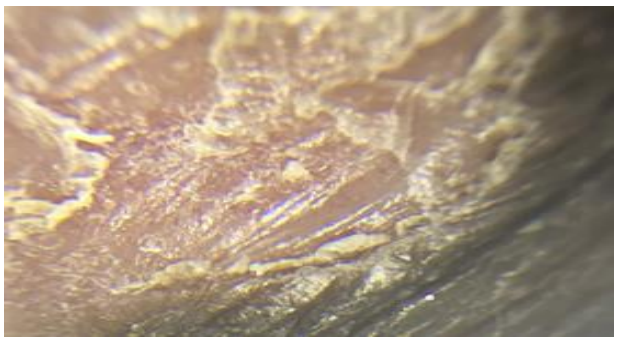

Figure 4: Groove on the keratotic ridge.

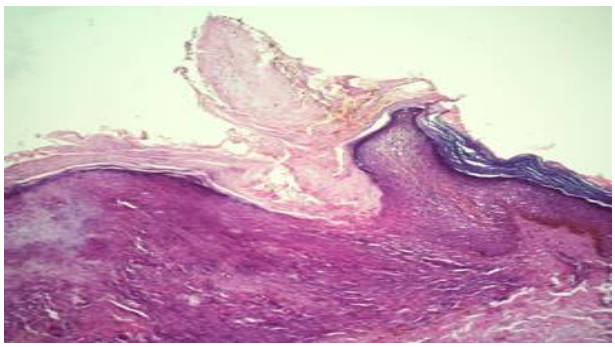

Figure 5: Histopathology revealed coronoid lamella composed of thin column of tightly packed parakeratotic cells within a keratin filled epidermal invagination with underlying hypogranulosis. (H&E stain, 40x).

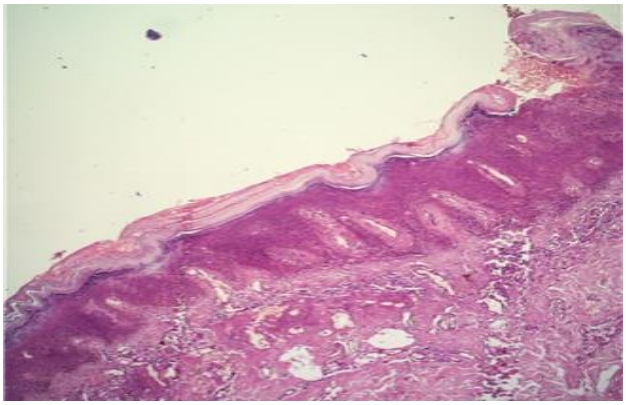

Figure 6: Histopathology showed multiple coronoid lamella. (H&E stain, 10x)

The patient was treated with 5-flurouracil cream and she is on regular follow up (Figure 7).

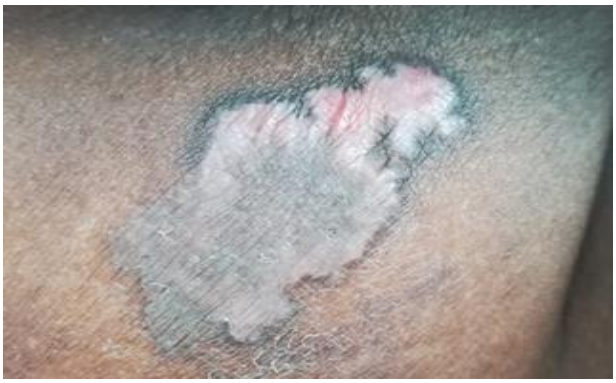

Figure 7: Slight flattening of the lesion after 10 days treatment of topical 5-flurouracil.

DISCUSSION

A porokeratosis is a clonal expansion of keratinocytes which differentiates abnormally but are not hyperproliferative.[3]

Clinical classification of porokeratosis:[3]

Localized Forms

- Porokeratosis of mibelli
- Linear porokeratosis
- Punctate palmoplantar porokeratosis
- Genital porokeratosis
- Perianal porokeratosis

Disseminated Forms

- Disseminated superficial actinic porokeratosis
- Disseminated superficial porokeratosis
- Systematized linear porokeratosis
- Disseminated palmoplantar porokeratosis

Perianal porokeratosis, also known as porokeratosis ptychotropica, verrucous porokeratosis. Porokeratosis ptychotropica is a quite rare and little-known disease with difficult diagnosis. In greek, the word "ptyche" means fold and "trope" means turning. It was first reported by Helfman and Poulos, who described it as reticular porokeratosis affecting the genital/pelvic region.[4] Lucker et al, described a 34-year-old man with a 9-year history of pruritic lesions confined to the natal cleft and he coined the term "porokeratosis ptychotropica".[5] Stone et al, described a similar case, in which a 32-year-old man with a 13-year history of pruritic lesions also confined to the natal cleft and he named it as “perianal inflammatory verrucous porokeratosis.[2]

The term genitogluteal porokeratosis refers to the porokeratosis restricted to male and female genitalia and those restricted to buttocks. It can be broadly divided into three groups namely classical porokeratosis restricted to the genital region, ptychotropic porokeratosis and penoscrotal porokeratosis (Table 1).[6]

Table 1: Comparison of different types of porokeratosis occurring on the genitogluteal region.

Types	Age	Gender	Sites	Histopathology
Porokeratosis of mibelli	All ages	Both male and female	Penis, scrotum, vulva	One coronoid lamella
Porokeratosis ptychotropica	All ages	Both male and female	Gluteal cleft	Multiple coronoid lamella
Penoscrotal porokeratosis	Third decade of life	Only males	Penile shaft and anterior scrotum	Multiple coronoid lamella

In a review of 22 patients by Takiguchi et al, porokeratosis ptychotropica appears to be more common in male and the main regions affected were buttocks, genito gluteal regions and buttocks with involvement of extremities.[7] Usually it is described as pruritic keratotic plaque and it presents as confluent perianal lesion that is symmetric on both sides of the buttocks with possible satellite lesions. This is sometimes described as butterfly shaped scaly plaque on the buttocks surrounding the anus unlike our case which is presented as single verrucous

plaque with peripheral thread like border and no satellite lesions because most reported cases of porokeratosis ptychotropica have resembled either psoriasis or verruca vulgaris. We considered plaque type of lupus vulgaris as the differential diagnosis, because cutaneous tuberculosis is also not uncommon in india and the buttocks are more commonly involved. The growth is often slow, occurring over the course of 5-10years. Pruritus is often present.[8] The etiology is not well established, but friction with clothing may be an aggravating factor and the patient may scratch the lesion due to itching which contributes to secondary amyloid deposition in the dermis. Though it is not seen in our case, it has been reported earlier.[9] Histologically multiple coronoid lamella is seen which is unique to this rare type. Malignant transformation of the porokeratotic lesions can occur in 7-11% of the cases and the risk factors includes nonexposed skin, long standing lesions, linear porokeratosis, large size lesions, older patients and previous radiation. Squamous cell carcinoma and basal cell carcinoma are the most common malignancies though it is not reported so far in porokeratosis ptychotropica.[9] Porokeratotic ptychotropica is often refractory to treatment and the various treatment modalities which produced no response/minimal benefit with symptomatic relief include topical corticosteroids, tacrolimus, pimecrolimus, calcipotriol, PUVA, retinoids, cryotherapy. Some improvement has been reported with topical 5 flurouracil, CO_2 laser and dermabrasion.[8,10]

We report this case due to its rarity, unusual presentation of single plaque in the gluteal cleft which is similar to the classical porokeratosis of mibelli and not like the symmetrical verrucous plaques which has been reported in the literatures so far.

DECLARATIONS

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Sangheetha K., Shareef R A., Jaffer A N. porokeratosis ptychotropica-a rare case report with unusual presentation. Int J Res Med Sci 2019;7:313-6.