A Case Report of Porokeratosis Ptychotropica without Genitogluteal Involvement

Ekkapap Puapermpoonsiri MD,

Nattaporn Sampattavanich MD.

ABSTRACT:

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DIVISION OF DERMATOLOGY, DEPARTMENT OF MEDICINE, PHRAMONGKUTKLAO HOSPITAL BANGKOK, THAILAND.

Porokeratosis ptychotropica is a rare hyperkeratotic verrucous variant of porokeratosis with preferable site at the genitogluteal area without typical keratotic rims as other types of porokeratosis. We report a very rare case of porokeratosis ptychotropica with no genitogluteal involvement. Our case was an elderly Thai man presented with multiple well defined hyperkeratotic verrucous plaques on the right knee, left leg and dorsum of both feet which was diagnosed as porokeratosis by the typical cornoid lamella in the histopathologic examination. Our patient had a favorable outcome from wide excision, oral acitretin, topical fluorouracil, and salicylic acid.

Key words: Verrucous porokeratosis, porokeratosis ptychotropica, genitogluteal, cornoid lamella

From: Division of Dermatology, Department of Medicine, PhramongkutklaoHospital, Ratchathewi Bangkok Thailand

Corresponding Author: Nattaporn Sampattavanich MD., email: nariyata@hotmail.com

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Introduction

Porokeratosis (PK) is a keratinocytes hyperproliferation disorder, typically presents with papules or plaques with keratotic rims and central atrophy[1]. PK is usually diagnosed by histopathology that shows characteristic features of cornoid lamella[2]. There are multiple variants of PK which may simultaneously occur in the same patient. Porokeratosis ptychotropica (PP) is a rare variant with manifestations of hyperkeratotic verrucous plaques without typical clinical features of PK and need pathologic examination to get the diagnosis. Most of the reported cases of PP had lesions at gluteal cleft and genital area and very rare case of PP had no involvement in this region. Herein, we report a case of PP without genitogluteal lesions, presented with large keratotic verrucous plaques and diagnosed PK by the histopathologic features.

Case presentation

A 59-year-old Thai man with hypertension was consulted from out-patient clinic of internal medicine due to the chronic rashes on both legs for 30 years. The rashes were slightly itchy and gradually enlarged. Twenty years ago, he had the history of skin biopsy but no documentation had been shown. The lesions were not improved with the previous treatment. Dermatological examinations showed multiple well defined hyperkeratotic verrucous plaques at right knee, left leg and both dorsum of feet (Figure 1).

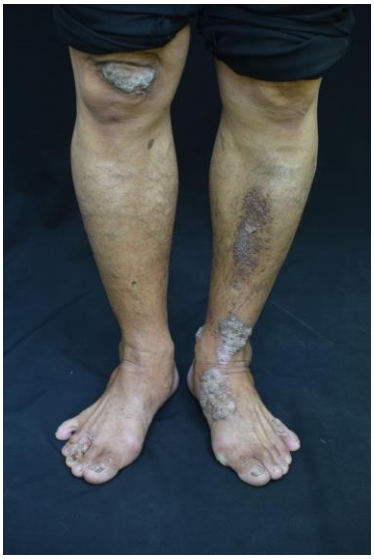

Figure 1 Multiple well-defined hyperkeratotic verrucous plaques at right knee, left leg and dorsum of both feet

Histopathology from the lesion on left dorsum of foot revealed marked hyperkeratosis, papillomatosis and irregular acanthosis epidermis without atypia. There are multiple columns of parakeratosis invaginating upper epidermis with necrotic keratinocytes under the column of parakeratosis (Figure 2). The histopathological result confirmed the diagnosis of porokeratosis. Tissue cultures for fungus, tuberculosis (TB) and non-TB mycobacterium were all negative. Anti-HIV test was done and showed negative result.

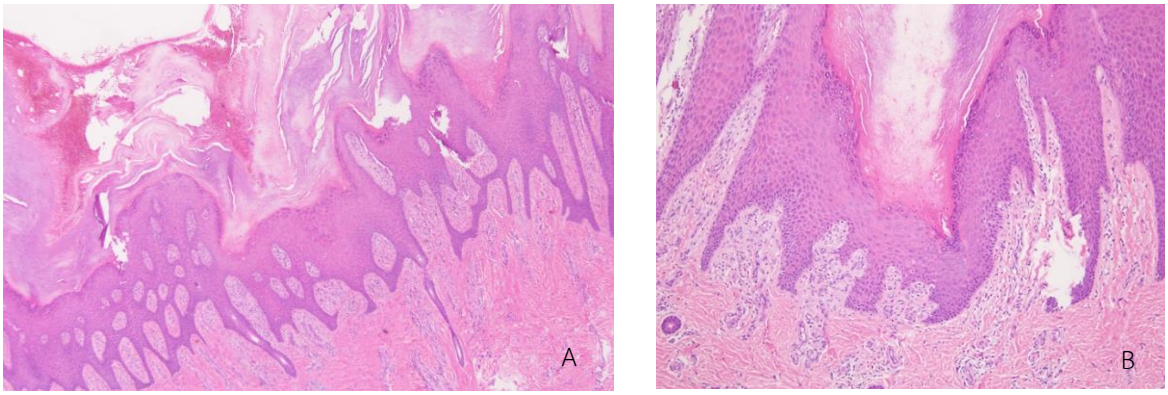

Figure 2 A) The skin biopsy from the patient's left foot revealed marked hyperkeratosis, papillomatosis and irregular acanthosis epidermis without atypia. Multiple columns of parakeratosis invaginating upper epidermis with necrotic keratinocytes are seen underneath the column of parakeratosis (H&E, X10).
B) Higher magnification (H&E, 40X) of section demonstrated a typical cornoid lamella.

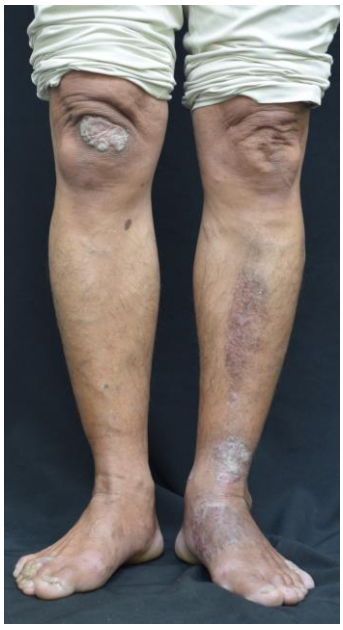

Figure 3 The lesion on the dorsum of left foot was excised with full-thickness skin graft. Concomitant treatments with oral acitretin and topical fluorouracil admixed with salicylic acid cream (Verrumal®) has improved the residual lesions.

The lesions on dorsum of left foot was excised with full-thickness skin graft and no malignancy was seen in the pathological report from the removed lesion. He also received oral acitretin 25 mg/day and topical fluorouracil admixed with salicylic acid (Verrumal®) for the treatment of other unresectable lesions which showed significant improvement.

Discussion

Porokeratosis (PK) is a disorder of keratinocytes hyperproliferation, first described by Mibelli in 1893[1]. The characteristic cutaneous features of PK are variable sized of papules or plaques with a keratotic ridge and central atrophy[1].

Etiology of PK is still undetermined. There were many cases reported association of PK with

immunodeficiency in organ transplantation, hematopoietic malignancies, HIV infection, use of immunosuppressive drugs, or chemotherapy[3]. Classical variants are disseminated superficial actinic PK (most common type), PK of Mibelli, punctate PK, PK palmaris et plantaris disseminate and linear PK. Variants of PK may also co-exist in the same patient[4], Porokeratosis ptychotropica has unique features of hyperkeratotic verrucous plaques without the typical keratotic rim, first described in 1995 by Lucker et al[5]. The predilection site of this variant is the genitogluteal region ("ptycho" means fold, "tropica" means turning)[6]. While clinical findings in our patient can be compatible with this variant, our patient showed no cutaneous involvement at the genitogluteal area. However, there was the report in India showing a patient with verrucous PK who had multiple lesions at other sites without genitogluteal region, the same as in our patient[7]. Usually, diagnosis cannot be done with cutaneous features alone, so the histopathology examination plays an important role for the definitive diagnosis. Although there are different clinical features in each variants of PK, all types of PK show the same histopathological pattern. The typical findings are thin column of parakeratotic cells and absence of a granular layer; the characteristic features of a cornoid lamella[2]. Incidence of malignant transformation is approximately 7%[8]. Malignant transformation has been reported in various types of PK, such as Bowen's disease have been reported in giant PK, and squamous cell carcinoma arising from giant PK of Mibelli in a Chinese man[8].Large lesion, long standing duration, linear type and previous ionizing radiation of PK are the high risk factors for malignancy[9].

There are many treatments of PK including surgical resection, cryosurgery, electrocautery, carbon dioxide laser ablation, oral retinoids, keratolytic agents, topical 5- fluorouracil, topical corticosteroids, topical imiquimod[3,10]. Choice of treatment depends on lesion location and size, functional and aesthetic requirements[10]. For verrucous PK, there were reports showing improvement in itching and flattening of lesions with oral isotretinoin, and topical 5-fluorouracil cream[6].

In our case, skin biopsy was done and demonstrated typical features of PK. So, our patient was diagnosed as verrucous PK. The lesion on the left dorsum of foot was excised with full-thickness skin graft. No malignancy was seen in the pathological report from the removed lesion. Moreover, concomitant treatments with oral acitretin at the dosage of 25 mg/day and topical fluorouracil admixed with salicylic acid cream (Verrumal®) improved other residual lesions in our patient (Figure 3). Even though, no malignant transformation was report, the surveillance is still

recommended especially in the case with long duration and large lesion.

Conclusion

We report a rare case of PP without genitogluteal involvement, which presented with hyperkeratotic verrucous plaques at both legs and feet. One of histopathological characteristics of porokeratosis ptychotropica is multiple columns of cornoid lamellae. Combination treatments composed of wide excision, oral acitretin, topical fluorouracil, and salicylic acid provided favorable outcome in our case.

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