

Hyperkeratotic Porokeratosis of Mibelli, Hyperkeratotic Disseminated Superficial Porokeratosis and Porokeratosis Ptychotropica with Dermal Amyloid Deposition

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ABSTRACT:

SARTITPITAKUL P, SUDTIKOONASETH P, PUANGPET P. HYPERKERATOTIC POROKERATOSIS OF MIBELLI, HYPERKERATOTIC DISSEMINATED SUPERFICIAL POROKERATOSIS AND POROKERATOSIS PTYCHOTROPICA WITH DERMAL AMYLOID DEPOSITION. THAI J DERMATOL 2013; 29: 178-182.

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Porokeratosis is a disorder of keratinization characterized by annular macules, papules or plaques with atrophic center and surrounded by distinctive hyperkeratotic thread-like border with longitudinal furrow. Histopathological diagnosis is cornoid lamellae. Multiple variants were subsequently described including the rare hyperkeratotic porokeratosis variant. Amyloidosis have been observed in many types of porokeratosis. The authors report a case of 58-years-old man who presented with co-existing three rare porokeratosis variants; hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica with dermal amyloid deposition. The skin condition was improved after applied 20% topical salicylic acid cream.

Key word: Hyperkeratotic porokeratosis of Mibelli, Hyperkeratotic disseminated superficial porokeratosis,

Porokeratosis ptychotropica, amyloid

บทคัดย่อ:

ภค สาธิตพิฐกุล, ปุณวิศ สุทธิกุลณเศรษฐ์, ไพลิน พวงเพชร รายงานการพบ **HYPERKERATOTIC POROKERATOSIS OF MIBELLI, HYPERKERATOTIC DISSEMINATED SUPERFICIAL POROKERATOSIS และ POROKERATOSIS PTYCHOTROPICA** ร่วมกับการสะสมของ **AMYLOID** ในผู้ป่วยรายเดียวกัน วารสารโรคผิวหนัง 2556; 29: 178-182.

สถาบัน โรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Porokeratosis เป็นโรคทางพันธุกรรมที่เกิดจากความผิดปกติของเคราตินซึ่งสามารถแบ่งตามลักษณะทางคลินิกได้หลายชนิด รวมทั้งชนิดที่พบบ่อยคือ hyperkeratotic porokeratosis มีรายงานไม่มากที่การตรวจทางจุลพยาธิวิทยาของโรคนี้พบการสะสมของ amyloid ร่วมด้วย รายงานนี้เป็นการรายงานการพบ Porokeratosis ชนิดที่พบบ่อย 3 ชนิดในผู้ป่วยรายเดียวกันคือ hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis และ porokeratosis ptychotropica ร่วมกับการตรวจทางจุลพยาธิวิทยาพบการสะสมของ amyloid โดยผู้ป่วยมาด้วยอาการผื่นคันนูนแข็งที่หน้าอก หลัง ก้น แขนและขาทั้งสองข้าง ให้การรักษาด้วยยาทากรดซาลิไซลิก 20% อาการของผู้ป่วยดีขึ้น

คำสำคัญ: Hyperkeratotic porokeratosis of Mibelli, Hyperkeratotic disseminated superficial porokeratosis,

Porokeratosis ptychotropica, amyloid

Porokeratosis is a hereditary disorder of keratinization clinically characterized by a distinctive ridge-like border and histologically by presence of cornoid lamella in the upper area of invaginated epidermis. Multiple variants have been described including hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica, three rare subtypes. We report a case of hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica with dermal amyloid deposition.

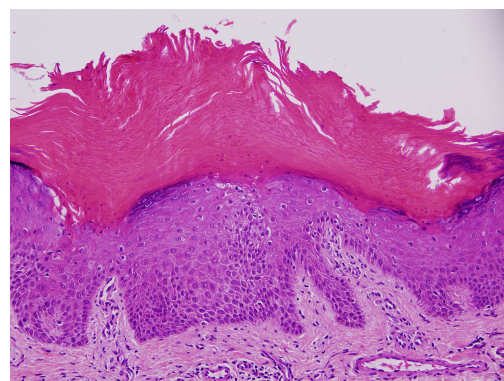


Figure 1-2 Hyperkeratotic porokeratosis of Mibelli

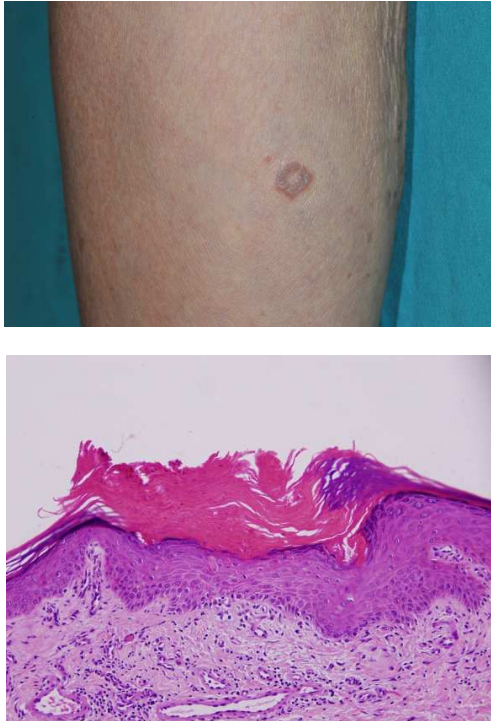


Figure 3-4 Hyperkeratotic disseminated superficial porokeratosis

Case report

A 58 years old male patient had visited our institute with history of pruritic papules and plaques which firstly appeared on his left knee and then extended to chest, back, both arms and legs 4 years ago. The lesions presented on both exposed any non-exposed areas. He was otherwise well, without any medication, and had no family history of and skin disease. Physical examination revealed two discrete large irregular-shaped hyperkeratotic plaques with atrophic center on left knee with slightly elevated border (figure 1), multiple brownish hyperkeratotic papules with raised hyperkeratotic borders on trunk and all extremities

(figure 3). Some papules tended to coalesce to form lichenified plaques. The palms, soles, mucous membranes, and nails were spared.

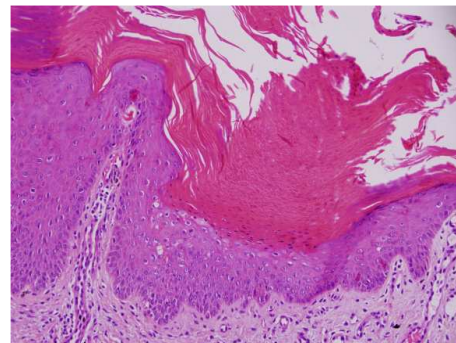


Figure 5-6 Porokeratosis ptychotropica

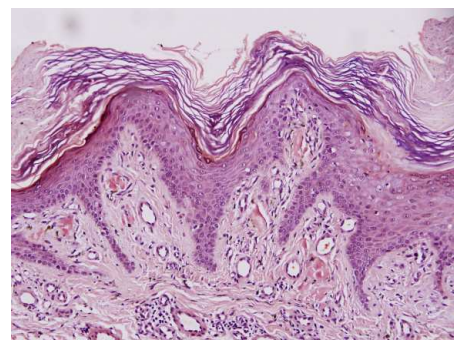


Figure 7 The congo red stained sections revealed dermal amyloid deposition.

Three years later, he developed several pruritic hyperkeratotic papules coalesce to form plaque on his perianal area. Physical examination showed bilateral brownish hyperkeratotic plaque with satellite papules on right gluteal area sparing anus (figure 5). The skin biopsy taken from lesions on right forearm (figure 2), left knee (figure 4) and right buttock (figure 6), revealed acanthotic hyperplastic epidermis with focal hyperkeratosis and multiple cornoid lamellae. Several necrotic keratinocytes were identified underneath. The dermis showed superficial perivascular infiltration of lymphocytes. Aggregated of amyloid material were noted in papillary dermis and were highlighted by congo-red stain (figure 7).

Based on these clinical and histological findings, a diagnosis of hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica with dermal amyloid deposition were made. Treatment with 20% topical salicylic acid was given and thickness as well as itchiness was decreased.

Discussion

Porokeratosis was firstly described by Mibelli and Respighi in 1893.^{1,2} Several variants of porokeratosis have been described including porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis, porokeratosis palmaris et plantaris disseminate, punctuate porokeratosis, linear porokeratosis and other rare

variants, such as porokeratotic ptyrotropica and the syndromic porokeratoses.³

Hyperkeratotic porokeratosis of Mibelli was initially reported by Degos in 1953.⁴ Hyperkeratotic porokeratosis of Mibelli is different from porokeratosis of Mibelli in that the former has late onset, more severe keratinization, more abundant cornoid lamellae and vascular enlargement in dermal papillae.⁵

Hyperkeratotic variant of disseminated superficial porokeratosis has been recently described by Inazawa et al in 2013.⁶ They reported a 80 year old man with 2 year history of multiple pruritic papules on both sun-exposed and non-sun-exposed areas. Histopathological examination demonstrated cornoid lamellae with dense lymphocytic infiltrate and amyloid deposition in the papillary dermis.

Porokeratosis ptychotropica was firstly described by Lucker et al in 1995.⁷ Other terms including hyperkeratotic porokeratosis, genitogluteal porokeratosis, porokeratoma and follicular porokeratosis have been used to describe this condition.⁸ Porokeratosis ptychotropica presents as an itchy expanding red-brown verrucous plaque in the perianal region with surrounding satellite lesions.⁹ The histology is characterized by acanthosis epidermal hyperplasia with multiple cornoid lamellae. In most reported cases, there is a subjacent dermal amyloid deposits.⁹

The presence of amyloid deposits in porokeratosis lesions was initially described in 1974.¹⁰ In 2008,

Ramírez-Santos studied secondary amyloid deposit in 34 biopsies of lesions clinically and histologically compatible with porokeratosis from 30 patients. They concluded that advanced age of the patients and the chronic nature of the lesions would have been predisposing factors for amyloid deposition.¹¹

Malignancy was developed in 7.5% of porokeratosis lesions.¹² Long-term clinical follow up may be prudent. To date, no case of malignancy transforming in hyperkeratotic porokeratosis lesions has been reported.

In our case, the patient presented with few annular plaques with central atrophy on the left knee, multiple small plaques symmetrically distributed on the sun-exposed and non-exposed skins and hyperkeratotic papules confluent into large plaque on right buttock. The biopsy was done and histopathologic findings of multiple cornoid lamellae and aggregated of amyloid material in tissue sections from right forearm, left knee and right buttock led to the diagnosis of hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica with dermal amyloid deposition. For treatment, topical keratolytics were prescribed. The symptoms were improved.

In conclusion, we report a case of co-existing three rare porokeratosis variants; hyperkeratotic porokeratosis of Mibelli, hyperkeratotic disseminated superficial porokeratosis and porokeratosis ptychotropica. The dermal amyloid deposition may suggest the advanced age of our patients and the chronic nature of the lesion.

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