

A Case of Porokeratosis Palmaris et Plantaris Disseminata

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Porokeratosis is a group of disorders characterized by epidermal keratinization associated with cornoid lamella. Porokeratosis has been described as having five distinct clinical subtypes: Mibelli or plaque type, disseminated superficial actinic porokeratosis, linear porokeratosis, punctate porokeratosis, porokeratosis palmaris et plantaris disseminata. We describe a 61-year-old Korean woman presented with porokeratosis palmaris et plantaris disseminata. (*Ann Dermatol* 16(2) 79~82, 2004)

Key Words: Porokeratosis palmaris et plantaris disseminata

INTRODUCTION

Porokeratosis is a group of disorders characterized by epidermal keratinization associated with a cornoid lamellae¹. In 1893, Mibelli coined the term porokeratosis for dermatosis in which typical lesions had their origin in the orifices of sweat gland ducts and/or hair follicles; this is termed classic porokeratosis of Mibelli. Respighi described a disseminated superficial porokeratosis. Subsequently, several other clinical variants of porokeratosis have been described: disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminata (PPPD), linear porokeratosis, and punctate porokeratosis. In addition, immunosuppression-induced porokeratosis has been found².

We present herein a rare case of PPPD found in Korean woman. To our knowledge, this is the third case of PPPD in Korea.

CASE REPORT

A 61-year-old Korean woman presented with generalized discrete annular brownish papules and palmoplantar keratotic plaques. The lesions developed first on both forearms 40 years ago. Over time, similar lesions developed on the palm, sole, lower extremities, trunk, and face. These lesions were not pruritic or painful and of cosmetic concern only. She complained of no exacerbation of the lesions during the summer. No mucosal lesions were found. She had been treated with urea and salicylic acid intermittently with slight improvement. The lesions on the palm were treated with carbon dioxide laser with some improvement. She had hypertension and osteoporosis with medication. On family history, her mother had similar lesions and diagnosed as having porokeratosis. Out of her six brothers and sisters, two brothers and one sister had the porokeratosis. On physical examination, there were multiple, superficial, small, round, uniform plaques demarcated by a distinct peripheral ridge of no more than 1 mm in height on the palm, sole, trunk, extremities, and face (Fig. 1. A, B). On histopathologic examination of the plantar porokeratotic lesion, the findings were as follows: thickened epidermis with parakeratotic column (cornoid lamellae) with diminished granular layer, few dyskeratotic cells, and focal vacuolar degeneration (Fig. 2. A, B). Also, sparse lymphocytic

Received February 23, 2004

Accepted for publication June 17, 2004

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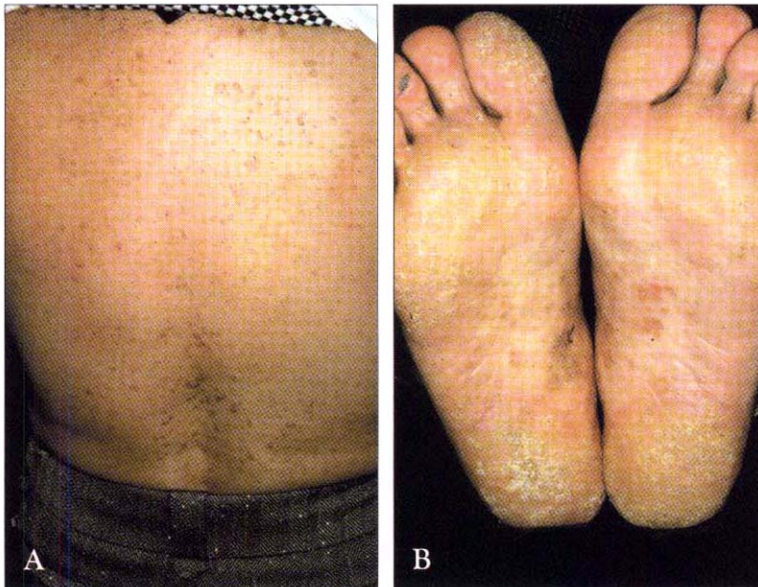


Fig. 1. A, B. multiple discrete pigmented papules with peripheral ridges on the back and both soles.

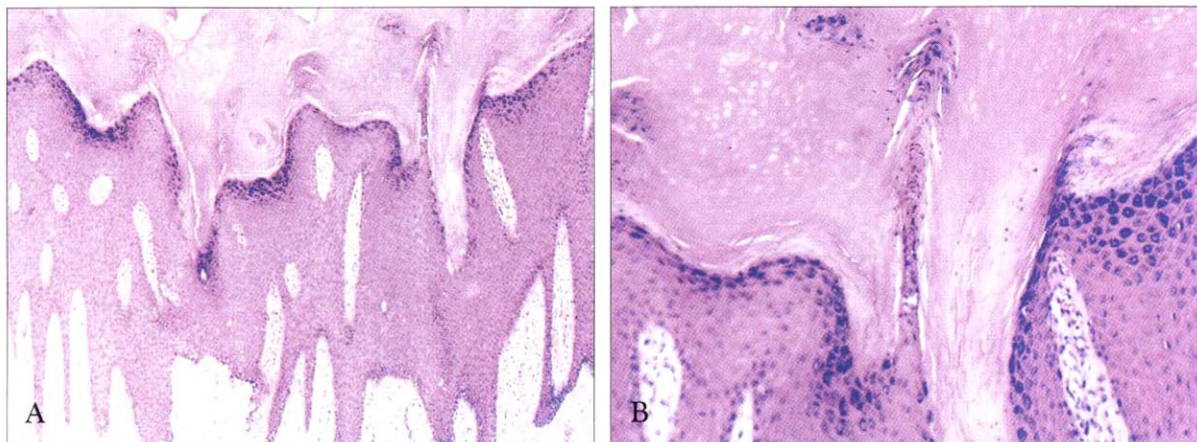


Fig. 2. A, B. pronounced hyperkeratosis with parakeratotic column(cornoid lamellae), focal diminished granular layer, irregular acanthosis in the epidermis and vacuolar degeneration, sparse inflammatory cell infiltrates in the papillary dermis (Fig. 2A: H&E \times 40, Fig. 2B: H&E \times 100).

infiltration and vascular dilatation in the papillary dermis were seen.

DISCUSSION

Porokeratosis palmaris et plantaris disseminata (PPPD) is a autosomal dominantly inherited genodermatosis characterized by multiple porokeratosis in both sun-exposed and non-exposed areas of the

body³. The lesions first appear typically on the palms and soles of patients in their early twenties and then gradually spread to the entire trunk and extremities³. The lesions are typically thick and confluent on the palms and soles, can cause disability, and may be associated with severe pruritus. Lesions on the body tend to be smaller and more discrete and superficial with the classic peripheral ridge that is characteristic of porokeratosis.

The fact that the lesions of our patients first

appeared on both forearms were not consistent with previous reports of PPPD and favored the diagnosis of disseminated superficial actinic porokeratosis (DSAP) rather than PPPD. But, the lesions of DSAP spare the palms, soles and the face. Also the lesions of DSAP usually appear in the third and fourth decades later than those of PPPD in which onset is during adolescence or early adulthood. Four previously described patients did not initially have involvement of the palms or of the soles⁴. We think our case is an addition to that category. There is an opinion that PPPD and DSAP are merely different expressions of the same clinical entity⁴. Wade and Ackerman classify PPPD and DSAP as a single clinical variant of porokeratosis and others suggest that the coexistence of DSAP with other variants of porokeratosis in the same family might request the simultaneous expression of two closely linked gene loci⁴. Recently, Wei et al.⁵ supported this hypothesis. According to their study, the PPPD locus was mapped by linkage analysis at chromosome 12q24.1-24.2 to a 6.9-cM interval between D12S1613 and D12S1341. Previously two loci for DSAP were located on 12q23.2-24.1 and 15q25.1-26.1 in two Chinese families by a genome-wide scan.^{6,7} The first locus for DSAP was located within the 9.6-cM interval between D12S1727 and D12S1605 at chromosome 12q23.2-24.1. The overlap region is about 0.6 cM from D12S1605 to D12S1613. PRDM4 is located centrally in this overlap region and seems to be an excellent candidate gene. PRDM4 has a role in cell differentiation and tumor suppression. So, there is a possibility that PRDM4 is not only associated with the development of DSAP and PPPD, but also involved in the pathogenesis of malignant transformation described in all varieties of porokeratosis.

Treatment is dependent on the type and extent of the lesion and should first involve sun protection, emollients, and observation for signs of malignant degeneration. Topical treatments such as salicylic acid and retinoic acid may be tried. Topical 5-fluorouracil has been shown to induce remission in all forms of porokeratosis.⁸ Other topical treatments include topical vitamin D3 analogues and topical imiquimod. In refractory cases, low dose oral retinoids can be utilized. It is possible that the use of oral retinoids in immunosuppressed patients, who are at higher risk for malignant degeneration, may reduce the risk of carcinoma developing in poroke-

ratotic lesions. Cryotherapy, electrodesiccation, and curettage can be used to treat small lesions. Although carbon dioxide laser ablation has been used with success, there is a high rate of recurrence.

After the first case report⁹ of typical lesions of PPPD in 37 year-old Korean woman in 1986, there was only another case report¹⁰ of PPPD. A 58-year old female is presented with basal cell epithelioma associated with PPPD, exhibiting the classical histopathologic criteria of the disease. Total excision for basal cell epithelioma and oral administration of etretinate for the treatment of porokeratosis plantaris, palmaris et disseminata and for the prevention of cancer development resulted in good response. To our knowledge, this is the third case of PPPD in Korea.

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