Coexistence of Porokeratosis of Mibelli and Disseminated Superficial Actinic Porokeratosis(DSAP)

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There have been several reports of more than one type of porokeratosis occurring in the same family or the same individual. We hope to support the view of different phenotypic expressions of a common genetic aberration by describing an additional case of porokeratosis of Mibelli on the perianal area and DSAP on the face, forearms occurring in a 45-year-old man.
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Key Words : Coexistence, Porokeratosis of mibelli, DSAP

Porokeratosis is a genodermatosis of autosomal dominant inheritance which has at least five different clinical variants: classic porokeratosis of Mibelli(PM); disseminated superficial actinic porokeratosis(DSAP); porokeratosis palmaris et plantaris disseminata(PPPD); linear porokeratosis; punctate porokeratosis[1]. All of these manifest the distinctive peripheral keratotic ridge which histologically corresponds to the cornoid lamella.

There have been several reports of more than one type of porokeratosis occurring in the same family or the same individual[2-10].

We hope to support the view of different phenotypic expressions of a common genetic aberration by describing an additional case of porokeratosis of Mibelli and DSAP occurring in an individual.

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CASE REPORT

A 45-year-old man had skin lesions on his face, extensor areas of both forearms, and perianal area for one year. He had no symptoms other than occasional slight itching. He has suffered from diabetes mellitus and hypertension for 8 years. He has been managed with insulin injections, oral hypoglycemics and antihypertensive drugs. Family history was unremarkable.

Physical examination revealed a 1.5 × 2 cm sized annular brownish atrophic plaque with a distinctive peripheral keratotic ridge and a few smaller lesions were present on his perianal area(Fig.1a). And multiple small brownish depressed macules with a faintly visible peripheral keratotic rim averaging approximately 8 mm in diameter were present on his face and extensor areas of both forearms(Fig.1b). A biopsy specimen taken from the edge of a plaque on the perianal area demonstrated a column of parakeratosis in an area of invaginated epidermis, focal absence of the underlying stratum granulosum, vacuolar changes in the stratum malpighii, and a minimal perivascular mononuclear infiltrates in the underlying papillary dermis(Fig. 2a). This confirmed the clinical diagnosis of porokeratosis of Mibelli. A biopsy specimen taken

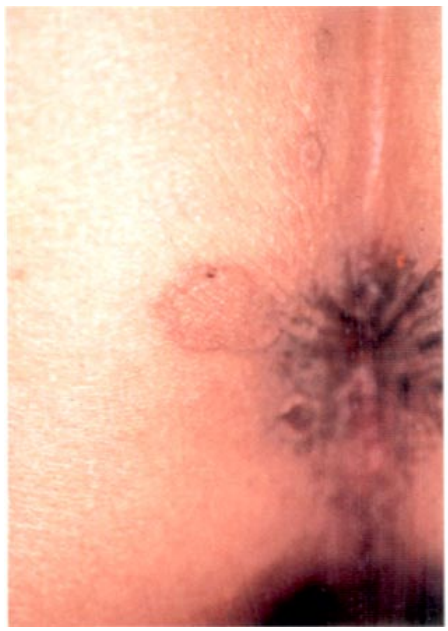

Fig.1a. Annular brownish atrophic plaques with a distinctive peripheral keratotic ridge on his perianal area.

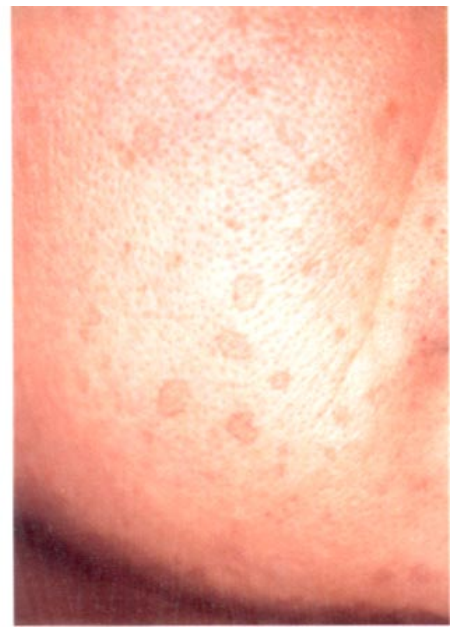

Fig.1b. Brownish depressed macules with a faintly visible peripheral keratotic rim on his face.

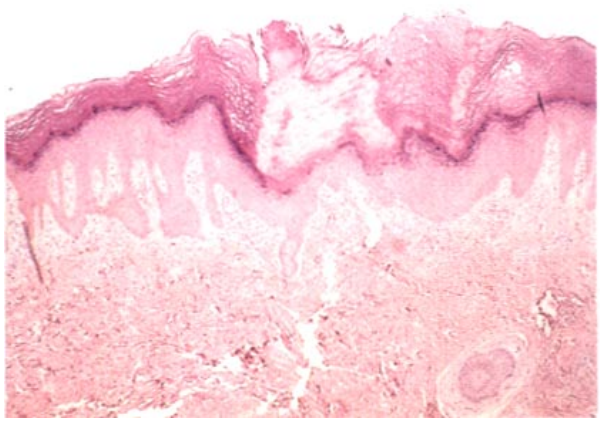

Fig.2a. Perianal area: A compact column of parakeratosis in an area of invaginated epidermis(H & E, × 100).

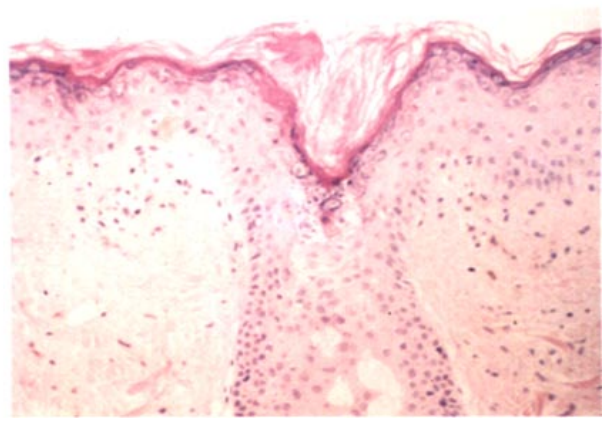

Fig.2b. Face: A less striking column of parakeratosis arising from a very superficially located furrow in the epidermis(H & E, ×200).

from the edge of a lesion on the face demonstrated a less striking cornoid lamellae arising from a very superficially located furrow in the epidermis confirming the clinical diagnosis of DSAP(Fig. 2b).

The patient was recommended to apply topical retinoic acid ointment onto the lesions twice daily and sunscreen whenever he went out.

DISCUSSION

Porokeratosis is a specific disorder of keratinization, which is characterized histologically by the presence of a cornoid lamella, a thin column of closely stacked parakeratotic cells extending throughout the stratum corneum.

The etiology of the various types of porokeratosis is unknown. Reed and Leone[11] feel that abnormal

clones of epithelial cells in the epidermis are responsible for the porokeratotic lesion. The tendency for abnormal clones to develop is probably inherited, probably an autosomal dominant condition with variable penetrance. The predisposition to porokeratosis is inherited but the predisposition to the type of lesion may not be. And additional triggering factors lead to clinical manifestation. Some triggering factors include irradiation with UV light from natural or artificial sources[12], photochemotherapy and phototherapy for psoriasis[13], and loss of immunocompetence[14]. An alternative hypothesis comes from the finding of an inflammatory mononuclear infiltrate composed of helper T cells, suppressor T cells, and Langerhans cells beneath the cornoid lamella[15].

In 1893, Mibelli first described annular and gyrate plaques with sharply elevated hyperkeratotic borders, occurring mainly on acral areas, which he called porokeratosis. Later, Chernosky and Freeman[16] described DSAP, a more common variant of porokeratosis, which differed from the classic Mibelli type, in that the lesions, while annular, were smaller with a less-pronounced peripheral ridge. They occurred on sun-exposed sites, were often accentuated by sunlight, and developed during the third and fourth decades, unlike the classic Mibelli lesions, which usually appear in childhood.

The coexistence of several porokeratotic variants has been described for porokeratosis of Mibelli with linear porokeratosis[2], for DSAP with porokeratosis of Mibelli or linear porokeratosis[3-8], for linear porokeratosis with punctate porokeratosis[9], and for DSAP, linear porokeratosis and punctate porokeratosis[10]. Porokeratosis of Mibelli and linear porokeratosis especially seem to be closely related. Many cases of the linear variant display distinctive annular lesions adjacent to the edge of the linear lesion, and many linear lesions appear to be made up of groups of annular lesions. So some investigators suggest the term, linear porokeratosis of Mibelli[3]. The coexistence of DSAP with other porokeratotic variants similar to our case is far less common.

Although different areas of the skin and different family members usually express the same morphological variant, the simultaneous expression of two closely linked genes could, perhaps, explain the coexistence of different porokeratotic variants and the similarities of clinical appearance and histopathology of porokeratosis.

Clinicians and investigators should therefore follow families of patients with porokeratosis to achieve a better understanding of the environmental modulation or other stimuli necessary for the expression of these clinical variants.

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