Porokeratotic Eccrine Ostial and Dermal Duct Nevus

Woo Chul Shim, M.D., Yoo Deuk Lee, M.D., Seung Hun Lee, M.D.

Department of Dermatology, Yonsei University Wonju College of Medicine
Wonju, Korea

A 6-year-old boy with porokeratotic eccrine ostial and dermal duct nevus on the left sole is reported. The patient was born with keratotic plugs, each measuring 1-2mm in diameter, on the left sole. Light-microscopic examination revealed epidermal invagination and cornoid lamella, which was found to be connected with hyperplastic intraepidermal and dermal eccrine duct. The present case can be histologically differentiated from nevus comedonicus and punctate porokeratosis. (Ann Dermatol 3:(1) 49–53, 1991)

Key Words: Porokeratotic eccrine ostial and dermal duct nevus

Porokeratotic eccrine ostial and dermal duct nevus is a rare congenital or adult-onset eccrine hamartoma characterized by linear distribution of groups of keratotic papules, especially on the palms and soles.

Histopathologically, it is characterized by the presence of cornoid lamella associated exclusively with the eccrine duct and ostia. Since Abell and Read first described this disorder in 1980; six cases have been reported in English literature[2-5]. Marsden et al[6] suggested that nevus comedonicus of the palm might be a sweat duct nevus. However, the case reported by Marsden et al as well as by Abell and Read are thought to be the same; therefore, nine cases of this desease have been reported including the cases written in other languages[7, 8].

We report another case of porokeratotic eccrine ostial and dermal duct nevus with electron microscopic findings.

Received October, 5, 1990
Accepted for publication December, 20, 1990
Reprint request to: Seung Hun Lee, M.D., Department of Dermatology Yonsei University Wonju College of Medicine 162, Ilsan-dong, Wonju, Korea
Presented in part at the meeting of the 5th International Congress of Pediatric Dermatology, Milano, Italy, July, 11, 1989.

REPORT OF A CASE

A 6-year-old boy visited our department because of linear skin lesion on the left sole which had existed from birth. Physical examination revealed multiple hyperkeratotic papules on the left sole in linear distribution measuring 1 to 2mm in diameter (Fig. 1). These keratotic papules were present on the heel and toe areas. There was no family history of similar skin disease. A heat-induced sweating test with starch-iodine technique showed a decrease in sweating at the lesional site. We took two biopsies from the lesion and examined them with a light microscope and electron microscope.

The histologic examination showed hyperkeratosis, acanthosis, focal parakeratosis and epidermal invagination (Fig. 2-A). Closer examination of the epidermis showed cornoid lamella-like parakeratotic column, and the base of the column showed an absence of granular layer and keratinocytes with vacuolated cytoplasm and pyknotic nuclei. We also observed small spaces considered to be acrosyringia in the cornoid lamella-like parakeratotic columns (Fig. 2-B). Also the dermal ductal portion of the eccrine sweat gland connected with the parakeratotic column was dilated and composed of several layers of ductal epithelium (Fig. 2-C). The intradermal ducts of basal coil were normal.

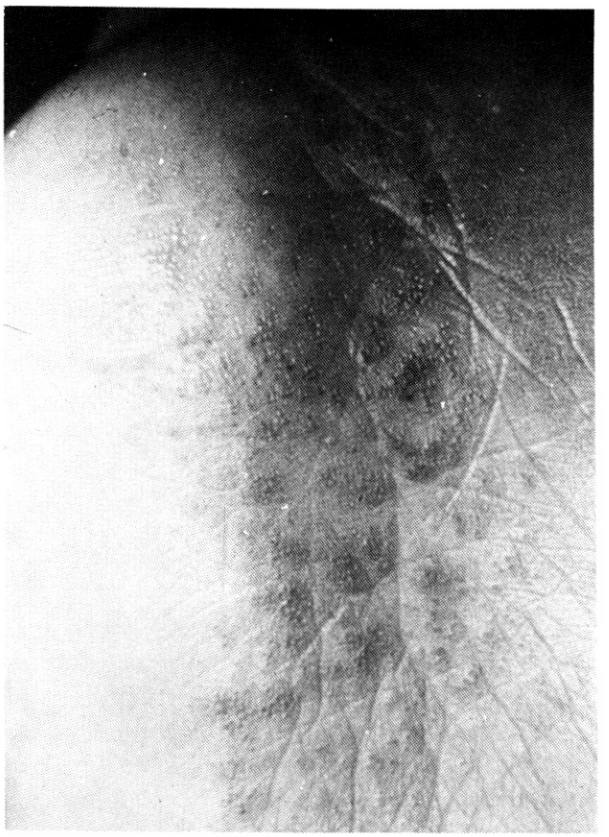

Fig. 1. Multiple small keratotic papules and comedo-like pits on the left sole in linear distribution.

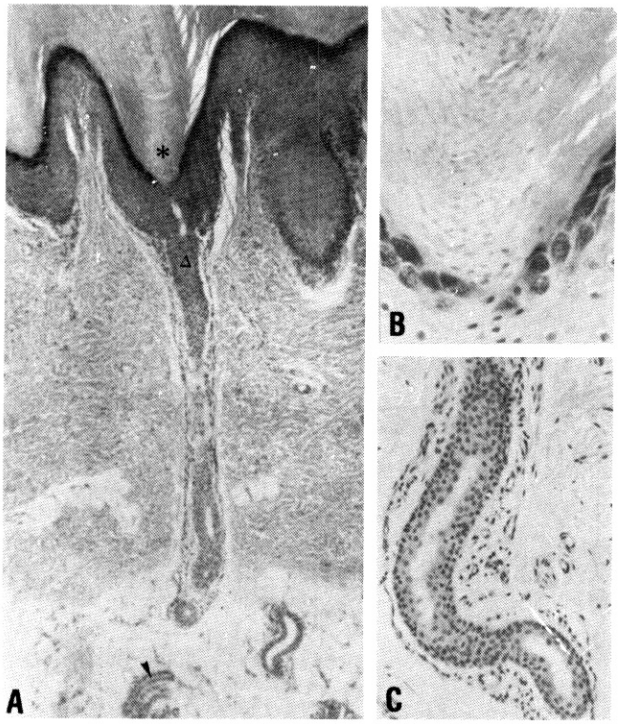

Fig. 2. A, Biopsy from sole shows a parakeratotic column over an eccrine sweat duct which is hyperplastic. The eccrine glands are normal (Hematoxylin-eosin stain, ×40).
B, High-power view of the lower portion of parakeratotic column shows focal absence of the granular layer and keratinocytes with vacuolated cytoplasm and pyknotic nuceli. Acrosyringia in the cornoid lamella-like parakeratotic column is also observed (×200).
C, The upper part of the dermal sweat duct is both hyperplastic and dilated (×200).

The scanning electron microscopic examination showed a distorted surface with numerous laminated keratotic plugs over the whole field (Fig. 3).

On transmission electron microscopic examination of the intradermal portion of eccrine duct, we observed an inner layer of luminal cells and at least four layers of outer cells. The hyperplastic outer cells were connected with each other by numerous, well-developed desmosomes and showed pronounced folding of their lateral borders (Fig. 4-A). The intradermal duct of basal coil showed an outer layer of basal cells and an inner layer of luminal cells. The cells of both layers showed pronounced folding of their lateral borders and numerous desmosomes (Fig. 4-B). Cells of the parakeratotic column contained pyknotic nuclei and numerous vacuoles and showed numerous desmosomes and pronounced folding of their lateral borders (Fig. 5).

DISCUSSION

Cornoid lamella is a histologic pattern characterized by a well-demarcated colum of compact hyperkeratosis containing parakeratotic cells situated within the stratum corneum. It is a characteristic feature of all forms of porokeratosis, but can be seen in other skin lesions as well[9]. Reed and Leone[10] suggested that cornoid lamella of porokeratosis does not originate from the epidermal portion of sweat duct but from the latent abnormal clones within the epidermis.

Porokeratotic eccrine ostial and dermal duct nevus, which was first reported by Abell and Read in 1980[1], is a eccrine hamartoma characterized clinically by congenital linear keratotic papules on

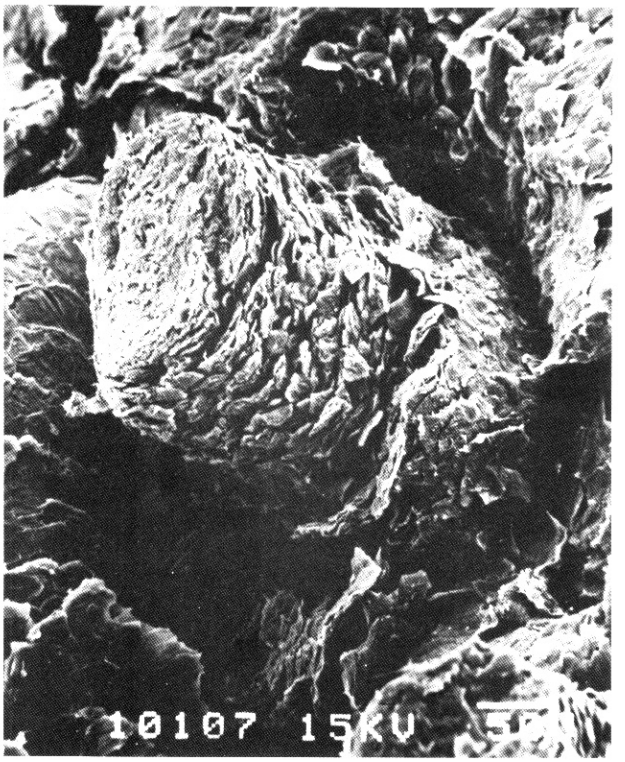

Fig. 3. A hyperplastic plug in the center of a pit. The hyperkeratotic plug is composed of laminated, horizontal corneocytes with surrounding keratin layer (SEM, ×200).

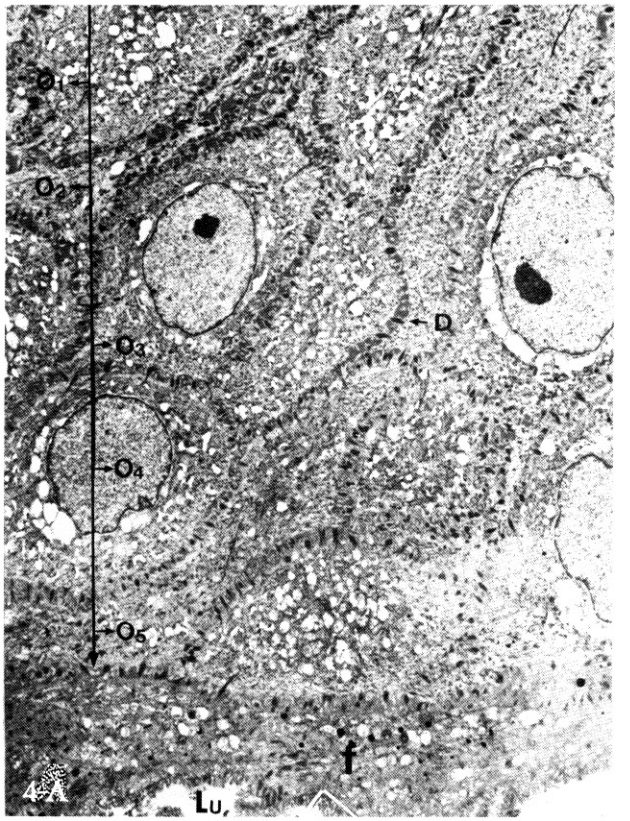

Fig. 4. A, Intradermal portion of hyperplatic eccrine duct (△ of Fig. 2) shows at least four layers of outer cells (01-5). The hyperplastic outer cells connect with each other by numerous, well-developed desmosomes(D) (TEM, ×3000). **B,** Intradermal duct of basal coil (△ of Fig. 2) shows an outer layer of basal cells(Bc) and an inner layer of luminal cells(Lc) (TEM, ×1500). Lu; lumen, mv; microvilli, f; periluminal filamentous zone

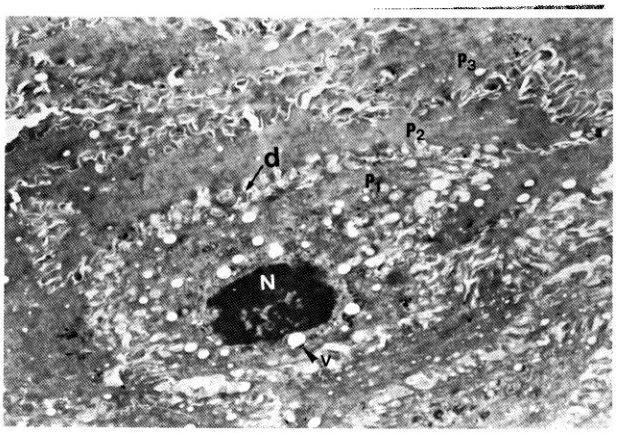

Fig. 5. Cells (P1-3) of a parakeratotic column (* of Fig. 2) contain a pyknotic nuclus(N), numerous vacuoles(v), desmosomes(d) and show pronounced folding of their lateral borders (TEM, ×4500).

the palms and soles. Histologically, it is characterized by cornoid lamella connected with hyperplastic intraepidermal and dermal eccrine duct.

Routine histologic examination of our case showed not only epidermal invaginations but also cornoid lamella-like parakeratotic columns associated exclusively with hyperplastic eccrine ducts.

We also observed acrosyringia in the cornoid lamella-like parakeratotic columns. Transmission electron microscopic examination revealed hyperplasia of cells of the intraepidermal duct, especially outer cells, as well as the intradermal duct. Regardless of levels of the eccrine duct, the ductal cells were connected with each other by numerous desmosomes and showed pronounced folding of their lateral borders, even in the cells of cornoid lamella-like parakeratotic columns. So the cornoid lamella-like parakeratotic column in this disease probably resulted from an increase of cohesion[11] between the ductal cells due to the numerous desmosomes and pronounced lateral borders. On the other hand, the parakeratotic column may be induced by an increased proliferation rate of eccrine ductal cells[12, 13].

This case should be differentiated from other dermatoses. Linear verrucous epidermal nevus, inflammatory linear verrucous epidermal nevus, and punctate palmoplantar keratoderma can easily be ruled out. Linear porokeratosis is a disorder which clinically resembles linear verrucous epidermal nevus with raised annular margin and atrophic center. Histopathologically, it has cornoid lamellation which is not associated with eccrine ductal hyperplasia[14].

Punctate porokeratosis is usually observed in adults, but is also observed in congenital forms. Clinically it shows numerous asymptomatic punctate pits and keratotic papules mainly on the palms and soles. Histopathologically, it shows cornoid lamella not only connected to the eccrine duct but also in hair follicles and in the epidermis proper. Additionally, no sweat gland abnormalities have been reported in this disorder[15-17].

The nevus comedonicus is considered to be hamartoma of the pilosebaceous apparatus. It rarely shows palmoplantar involvement, characterized by grouped papules with central keratotic plug resembling a comedo. It is characterized by comedo-like cystic dilatation and keratotic plugging. However, the features of parakeratotic column and association with eccrine duct have not been reported[18, 19].

The porokeratotic eccrine duct and hair follicle nevus described by Coskey et al[20] is a dermatosis in which cornoid lamellae involve hair follicles as well as a sweat gland duct and ostia.

A simple heat-induced sweating test with starch-iodine technique showed a decrease in sweating of the involved area. Therefore, as Aloi and Pippione have suggested[2], we think that this was probably due to obstruction of the sweat duct pore by horny material. On scanning electron microscopic examination, we confirmed the keratotic plugs firmly attached to the surface.

Stoof et al[5] recently reported another case of porokeratotic eccrine ostial and dermal duct nevus of adult onset which was thought to be a variant of the congenital form.

We believe from clinical, light microscopic and electron microscopic examinations that the case we are reporting is eccrine hamartoma having porkeratotic features, and this case should be considered porokeratotic eccrine ostial and dermal duct nevus.

REFERENCES

1. Abell E, Read SI: *Porokeratotic eccrine ostial and dermal duct naevus. Br J Dermatol 103:435-441, 1980.*
2. Aloi FG, Pippione M: *Porokeratotic eccrine ostial and dermal duct nevus. Arch Dermatol 122:892-895, 1986.*
3. Driban NE, Cavicchia JC: *Porokeratotic eccrine ostial and dermal duct nevus. J Cutan Pathol 14:118-121, 1987.*
4. Moreno A, Pudol RM, Salvatella N et al: *Porokeratotic eccrine ostial and dermal duct nevus. J Cutan Pathol 15:43-48, 1988*
5. Stoof TJ, Starink TM, Nieboer C: *Porokeratotic eccrine ostial and dermal duct nevus. Report of a case of adult onset. J Am Acad Dermatol 20:924-927, 1989.*
6. Marsden RA, Fleming K, Dawer RPR: *Comedo naevus of the palm-a sweat duct naevus? Br J Dermatol 101:717-722, 1979.*
7. Balato N, Cusano F, Lembo G, Ayala F: *Naevus sudoral eccrine porokeratosique pseudo-comedonien palmaire et plantaire. Ann Dermatol Venereol 113:921-922, 1986.*
8. Civatte J, Jeanmouging M, Denisart M et al: *Naevus sudoral eccrine palmaire pseudo-comedonien. Ann Dermatol Venereol 113:923-924, 1986.*

9. Wade T, Ackerman AB: *Cornoid lamellation: A histologic reaction pattern. Am J Dermatopathol 2:5-15, 1980.*
10. Reed RJ, Leon PL: *Porokeratosis-A mutant clonal keratosis of the epidermis. Arch Dermatol 101:340-347, 1970.*
11. Christopher E, Plewig G: *Formation of the acrosyringium. Arch Dermatol 107:378-382, 1973.*
12. Christopher E, Wolf HH, Laurence EB: *The formation of epidermal cell columns. J Invest Dermatol 62:555-559, 1974.*
13. Sato A, Anton-Lamprecht, Schnyder UW: *Ultrastructure of inborn error of keratinization. VII. Porokeratosis Mibeeli and disseminated superficial actinic porokeratosis. Arch Dermatol Res 255:271-284, 1976.*
14. Rahbari H. Cordero AA, Mehregan AH: *Linear porokeratosis: a distinctive clinical variant of porokeratosis of Mibelli. Arch Dermatol 109:526-528, 1974.*
15. Himmelstein R, Lynfield YL: *Puntate porokeratosis. Arch Dermatol 120: 263-264, 1984.*
16. Roberts LC, DeVillez RL: *Congenital unilateral punctate porokeratosis. Am J Dermatopathol 6:57-61, 1984.*
17. Sakas EL, Gentry RH: *Porokeratosis punctata palmaris et plantaris (punctate porokeratosis). Case report and literature review. J Am Acad Dermatol 13:908-912, 1985.*
18. Wood MG, Thew MA: *Nevus comedonicus. A case with palmar involvement and review of the literature. Arch Dermatol 98:111, 1968.*
19. Harper KE, Spielvogel RL: *Nevus comedonicus of the palm and wrist. Case report with review of five previously reported cases. J Am Acad Dermatol 12:185, 1985.*
20. Coskey RJ, Mehregan AH, Hashimoto K: *Porokeratotic eccrine duct and hair follicle nevus. J Am Acad Dermatol 6:940-943, 1982.*