

Epidermal Naevus with Eccrine Duct Changes: an Orthokeratotic Variant of Eccrine Ostial and Dermal Duct Naevus?

Elisabet Parera¹, Fernando Gallardo¹, Montserrat Gilaberte¹, Carles Barranco² and Ramon M. Pujol^{1*}

Departments of ¹Dermatology and ²Pathology, Hospital del Mar, IMAS, Passeig Maritim 25-29, ES-08003 Barcelona, Spain. *E-mail: Rpujol@imas.imim.es
Accepted July 27, 2006.

Sir,

Epidermal naevus is a congenital malformation of the epidermis, clinically manifested by verrucoid scaly plaques, often in a linear distribution following the Blaschko lines. It is considered a genetic cutaneous manifestation of mosaicism and may show different histological features. Hyperkeratosis, papillomatosis and acanthosis with elongation of rete ridges is the most common pattern in epidermal naevus. However, other different histopathological patterns, such as acrokeratosis verruciformis-like, epidermolytic hyperkeratosis, seborrhoeic keratosis-like, psoriasiform and acantholytic dyskeratosis have been reported occasionally (1).

In addition, some authors also include a few peculiar entities showing distinctive clinical and histological features, such as inflammatory linear verrucous epidermal naevus (ILVEN), porokeratosis-like epidermal naevus, and naevus comedonicus in the spectrum of epidermal naevus (1, 2).

We report here a congenital, solitary linear verrucous lesion, histologically showing epidermal hyperplasia and dilated acrosyringea filled with orthokeratotic plugs. This peculiar "epidermal naevus associated with acrosyringal changes" seems to represent an additional histopathological variant that has received little attention in the literature.

CASE REPORT

A 21-day-old girl was referred to our department for the evaluation of a solitary linear plaque on the right wrist that was present at birth. She was the full-term daughter of a healthy 38-year-old woman. The pregnancy had been complicated by diabetes during the last 3 months and had been treated with insulin. A caesarean section was performed because the labour was not progressing. No history of previous local trauma was recorded. There was no family history of similar dermatosis or other hereditary conditions.

Physical examination disclosed a brownish, flesh-coloured, slightly verrucoid, well-demarcated plaque, 20×10 mm in diameter, on the outer aspect of the right wrist (Fig. 1). No similar lesions were noted elsewhere. The rest of the physical examination was unremarkable.

Two different 4-mm punch biopsies were carried out. Histopathological examination disclosed a diffuse epidermal hyperplasia with basket-weave hyperkeratosis and discrete hypergranulosis. Several epidermal invagi-



Fig. 1. Brownish verrucous and linear plaque on the lateral aspect of the right wrist in 21-day-old girl.

nations corresponding to dilated deep acrosyringea with prominent hyperkeratosis and acanthosis were noted. Orthokeratotic keratin plugs extended from hyperplastic acrosyringea to the underlying dermal eccrine ducts (Fig. 2A). Dilated eccrine ducts in the dermis underneath epidermal invaginations were also observed (Fig. 2B). Neither loss of granular cell layer at the base of epidermal invagination or cornoid lamellae formation were observed. After multiple sections in both biopsy specimens, no evidence of parakeratotic columns within the epidermal invaginations could be demonstrated. Immunohistochemical staining for CEA, AE1-AE3, and α E β 7 antibodies confirmed that the epidermal invaginations corresponded to dilated acrosyringea connected with the underlying glandular sweat ducts (Fig. 2C).

DISCUSSION

We report here an unusual solitary linear congenital hamartomatous verrucous lesion, clinically identical to an epidermal naevus, characterized histologically by an epidermal hyperplasia, dilated acrosyringea filled with orthokeratotic plugs without cornoid lamella formation.

After reviewing the literature, we have found only one previous report showing a similar clinicopathological picture. In 1983, Imai & Nitto (3) reported a 22-year-old woman with several acquired papular lesions on the extensor aspect of her left leg under the term "eccrine nevus with epidermal changes". Histopathological examination disclosed acanthosis, hyperkeratosis, a serrate configuration and elongation of the rete ridges. A malformed eccrine sweat structure with ductal hyperplasia was noted.

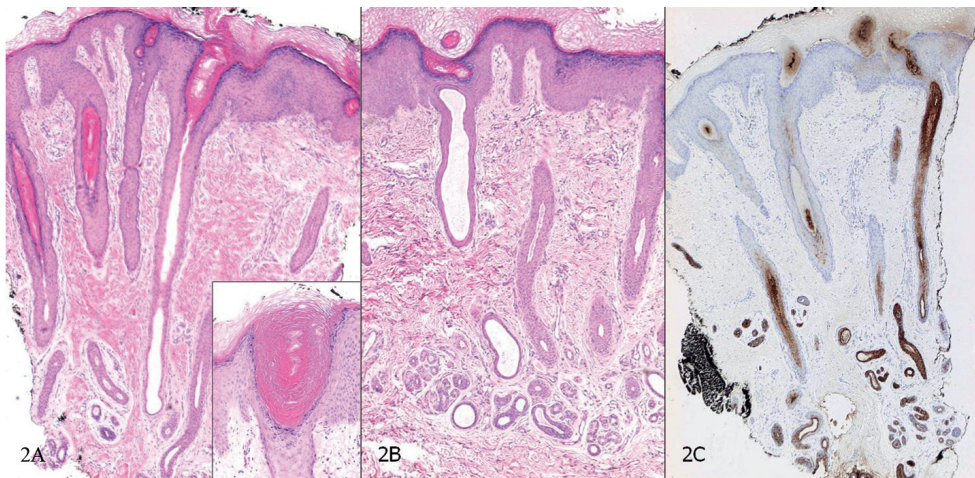


Fig. 2. Epidermal invaginations filled with keratotic plugs. (A) Hyperkeratosis, acanthosis and multiple dilated acrosyringia with infundibular keratinization (H&E $\times 40$). (B) Dilated eccrine ducts in the upper and mid dermis (H&E $\times 40$). (C) CEA staining in the dermal eccrine ducts and glands (H&E $\times 40$).

The differential diagnosis should be established with other non-hereditary genetic mosaic disorders, showing histologically comedo-like keratotic plugs corresponding either to dilated epidermal invaginations, dilated hair follicle infundibula (naevus comedonicus) or eccrine acrosyringia (porokeratotic eccrine ostial and dermal duct naevus (PEODDN)).

Naevus comedonicus is considered a naevoid disorder of the follicular apparatus (4). The observation of cases of naevus comedonicus involving the palms and soles is a rare phenomenon that can be observed in association with characteristic lesions present in hair-bearing skin. In such instances, palmar or plantar biopsy specimens showed comedo-like structures without eccrine connection (5, 6).

PEODDN is considered a non-hereditary genetic mosaic disorder of keratinization with eccrine involvement (7–10). PEODDN typically appears as a linear or band-like distribution of multiple grouped asymptomatic keratotic or verrucous papules with a comedo-like appearance. It is usually located on the acral portion of a limb (palms and/or soles) (10–11). Histologically, a comedo-like dilatation and hyperplasia of the acrosyringium with prominent parakeratosis and cornoid lamella-like plug formation within an epidermal invagination is usually observed. In PEODDN, cornoid lamellae occur exclusively in close association with or directly overlying the subjacent eccrine acrosyringial, which are frequently dilated and hyperplastic. Occasionally, associated follicular involvement has also been noted (10).

The precise cause of PEODDN remains elusive. The possibilities that the invagination is not derived from the acrosyringium and that the parakeratotic column could arise from the epidermal invagination have been postulated (8). Other authors have suggested that an abnormal clone of epidermal cells may produce the cornoid lamella-like column (9). The observed histo-

pathological features in our case were very close to those reported in PEODDN. However, the absence of an acrosyringial parakeratotic column within the epidermal invaginations permits us to rule out this diagnosis. The possibility that this lesion could represent a rare, orthokeratotic variant of eccrine ostial and dermal duct naevus cannot be ruled out completely.

REFERENCES

1. Su WPD. Histopathological varieties of epidermal nevus. *Am J Dermatopathol* 1982; 4: 161–170.
2. Altman J, Mehregan AH. Inflammatory linear verrucose epidermal nevus. *Arch Dermatol* 1971; 104: 385–389.
3. Imai S, Nitto H. Eccrine nevus with epidermal changes. *Dermatologica* 1983; 166: 84–88.
4. Weedon D, Strutton G. Epidermal and other nevi. In: Houston MJ, Black S, Naughton N, Gear S, eds. *Skin pathology*, 2nd edition. London: Elsevier Science, 2002; pp. 754–755.
5. Wood MG, Thew MA. Nevus comedonicus. *Arch Dermatol* 1968; 98: 111–116.
6. Harper KE, Spielvogel RL. Nevus comedonicus of the palm and wrist. Case report with review of five previously reported cases. *J Am Acad Dermatol* 1985; 12: 185–188.
7. Marsden RA, Fleming K, Dawber RP. Comedo naevus of the palm – a sweat duct naevus? *Br J Dermatol* 1979; 101: 717–722.
8. Bergman R, Lichtig C, Cohen A, Friedman-Birnbaum R. Porokeratotic eccrine ostial and dermal duct nevus. An abnormally keratinizing epidermal invagination or a dilated, porokeratotic plugged acrosyringium and dermal duct? *Am J Dermatopathol* 1992; 14: 319–322.
9. Jimenez J, Gomez I, Gonzalez C, Lopez J, Poblet E. Porokeratotic eccrine ostial and dermal duct naevus. *Br J Dermatol* 1995; 132: 490–492.
10. Kroumpouzou G, Stefanato CM, Wilkel CS, Bogaars H, Bhawan J. Systematized porokeratotic eccrine and hair follicle naevus: report of a case and review of the literature. *Br J Dermatol* 1999; 141: 1092–1096.
11. Moreno A, Pujol RM, Salvatella N, Alomar A, de Moragas JM. Porokeratotic eccrine ostial and dermal duct nevus. *J Cutan Pathol* 1988; 15: 43–48.