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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.

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Cornoid Lamella in Pachyonychia Congenita

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• **A 13-year-old white girl was admitted with pachyonychia congenita, Kumer and Loos type 1. In addition to all nails being distally elevated, plantar keratoses and bullae, and a scalloped tongue, the patient also had keratotic papules on the elbows and knees. A cornoid lamella was identified in the histologic examination of a keratotic papule.**

The cornoid lamella, widely regarded as pathognomonic for porokeratosis (Mibelli) and its variants, may not be specific. This histologic marker should be sought in other localized disorders of keratinization.

(*Arch Dermatol* 114:1795-1796, 1978)

The cornoid lamella, a parakeratotic column of cells below which the granular layer is absent, is widely held to be diagnostic of porokeratosis.[1-4] A case of pachyonychia congenita is presented in which the biopsy specimen of a keratotic papule showed a cornoid lamella.

REPORT OF A CASE

A 13-year-old white girl was first seen by us for bilateral painful plantar, keratotic lesions. She had been observed in other communities by other physicians and presented with a "known" diagnosis of pachyonychia congenita.

The mother of the patient noted that the first lesions were the "nails turning black several days after birth." Subsequently, as a crawling infant, papules became abundant on the knees. Since she began to walk, there have been recurrent bullae on the feet, progressing to keratoses. The patient has had no ocular, hair, or dental problems. The family history was normal.

The physical examination disclosed a scalloped, slightly coated tongue and follicular, keratotic papules on the elbows and knees. Keratoses and ruptured bullae were seen on the plantar surfaces. All nails were elevated distally over a large amount of keratotic debris (Fig 1).

A diagnosis of pachyonychia congenita, Kumer and Loos[5] type 1, was made, a biopsy specimen was taken from a keratotic papule on the left elbow, and a referral visit to a clinical geneticist was arranged. Upon review of the slide, a cornoid lamella was found in the hyperkeratosis (Fig 2).

COMMENT

The histology of pachyonychia congenita has been examined with special emphasis on the ventral nail plate,[6,7] oral lesions,[8,9] and laryngeal involvement.[10] The histopathologic description of pachyonychia congenita in two major dermatopathology texts[3,4] refers only to the nail and oral lesions. In several reports where the follicular keratotic papules are mentioned, they are regarded as areas of hyperkeratosis.[6,9,10] We could find no report of occurrence of cornoid lamella in pachyonychia congenita.

On the contrary, the cornoid lamella is widely regarded as "diagnostic" or "characteristic" of porokeratosis (Mibelli) and its variants.[1-4,11,12] Mikhail

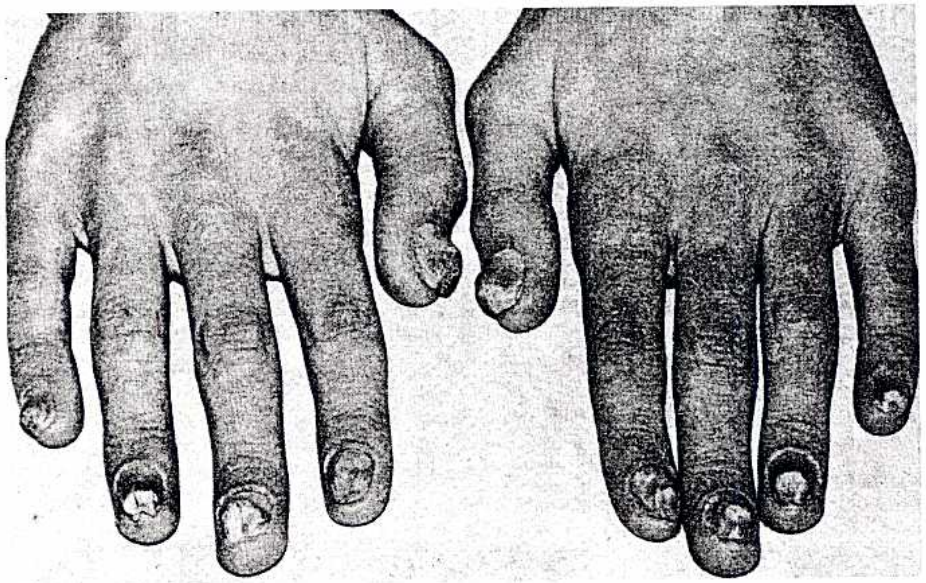

Fig 1.—Distal elevation of nails over a large amount of keratotic debris.

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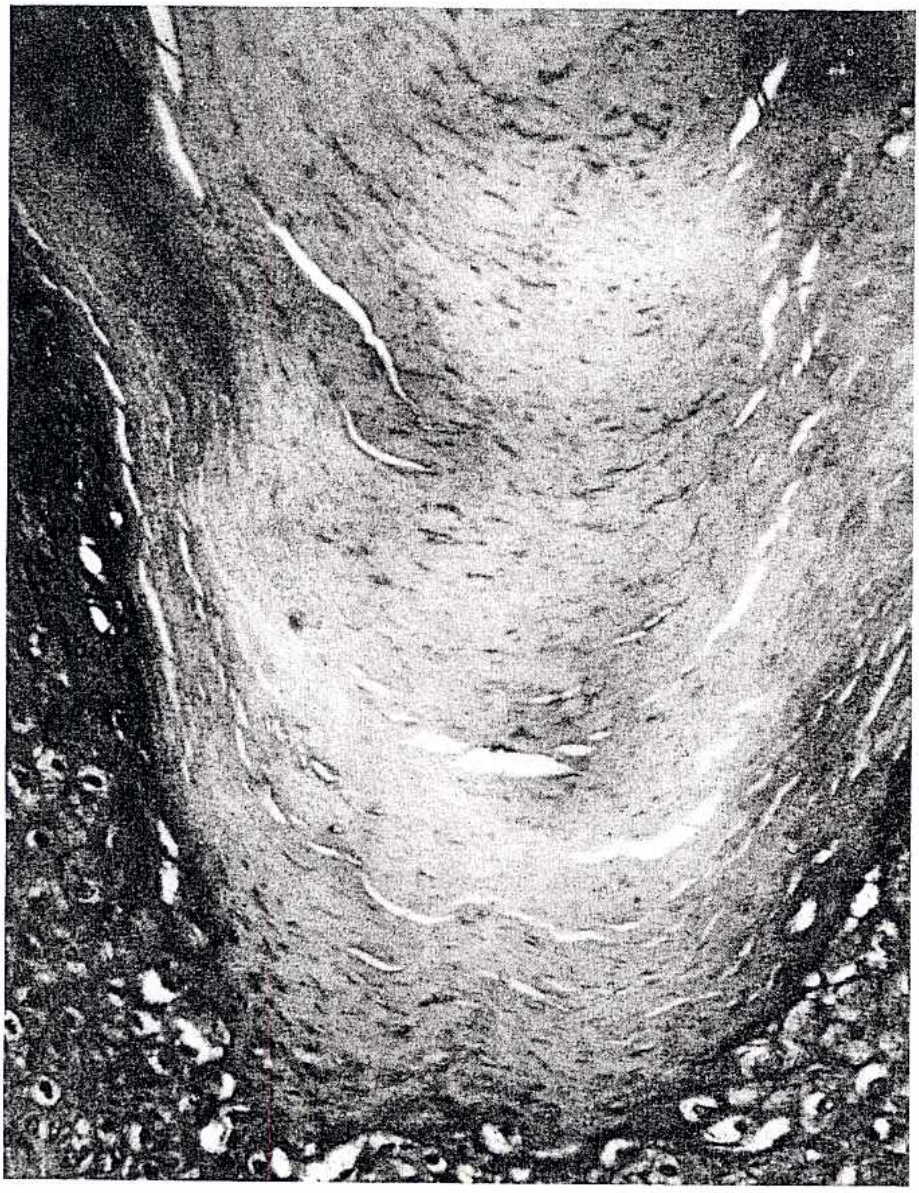

Fig 2.—Cornoid lamella in section of follicular, keratotic papule (hematoxylin-eosin, ×250).

and Wertheimer[1] accepted five types of porokeratosis: the classical plaque type; the superficial disseminated eruptive form; disseminated superficial actinic porokeratosis; a form with central thickening, hyperkeratosis, and erythema; and a hyperplastic inflammatory pattern. In addition, a zosteroid type has been proposed by Goldner.[11] Subsequently, Brown[13] and Herman[14] reported cases in which the cornoid lamella was observed in punctate keratoderma, or "punctate porokeratotic keratoderma." In none of these reports of porokeratosis did the clinical descriptions include nail changes or suggest pachyonychia congenita. Furthermore, the case reported here was quite typical of pachyonychia congenita, Kumer and Loos type 1.

Reed and Leone[12] have explained the histogenesis of porokeratosis as a stimulating influence on a clonal epithelium, with the cornoid lamella as a marker for the boundary between normal and abnormal epidermis. Two major dermatopathology texts[3,4] essentially agree with this hypothesis. The tendency for the development of the abnormal clone is held to be inherited, and actinic exposure may be an important operative factor.[12,15]

In summary, the cornoid lamella, characteristic of porokeratosis (Mibelli), has been found in a patient with clinically typical pachyonychia congenita, also a hereditary disorder with localized, disordered keratinization. Whether this is an isolated finding will be determined only by a careful search for the cornoid lamella in papules in other patients with pachyonychia congenita.

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