Cornoid Lamellation Occurring in Darier's Disease

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SUMMARY

Cornoid lamellation is a pathologic finding characteristic of porokeratosis. Nevertheless, it could be a part of histologic spectrum of few skin disorders. Histologically, "cornoid lamella" is a stack of closely packed parakeratotic cells on top of an area of hypogranulosis in the epidermis.

We report a case of Darier's Disease in a 13-year-old boy with cornoid lamellation. Review of literature as regards its occurrence in other diseases was carried out. We raise questions and inquiries about its pathogenesis and significance.



Fig. 1: Crusted papules on side of the neck.

Case Report

A 13-year-old boy had papular eruption on sides of his neck, chest and upper back for



Fig. 2: Punctate keratoses and minute pits on the palm.

about two years. Examination revealed firm crusted papules that are yellow-brown to brown in colour (Fig. 1). The eruption was confined mainly to the trunk. There was no lymphadenopathy.

Examination also revealed punctate keratoses and minute pits on the palms and soles (Fig. 2). The nails showed red and white longitudinal bands with V-shaped nick at their free margin, and slight splitting of some nail plates. No lesions could be detected on the mucous membranes. Routine urine and blood chemistry, endocrinologic, ophthalmologic and neurologic studies were all within normal limits.

A skin biopsy of trunk lesions showed features of Keratosis Follicularis (Darier's



Fig. 3: Low magnification shows "corps ronds", suprabasal acantholysis and cleft formation. Parakeratotic column is seen in the centre of epidermal component. H & E X100.

Disease); focal acantholytic dyskeratotic changes with the formation of "corps ronds", acantholysis and cleft formation (Fig. 3).

In the vicinity of these histologic findings, parakeratotic column shows at its base epidermal cells that are irregularly arranged and have pyknotic nuclei with perinuclear edema. No granular layer is found beneath the parakeratotic column (Fig. 4). A skin biopsy of palm lesions showed features of Darier's disease without cornoid lamellae.

Discussion

Cornoid lamella is a parakeratotic column of homogenous horny cells which posses deeply basophilic pyknotic nuclei. In the epidermis beneath it, the keratinocytes are irregularly arranged and have pyknotic nuclei with perinuclear edema.¹ In the upper stratum malpighii, some cells possess an eosinophilic cytoplasm as a result of premature keratinization.²

In porokeratotic lesions, cornoid lamella is usually not associated with a sweat pore, but crosses the fixed ostia of sweat ducts and hair follicles as it moves eccentrically.³ Cornoid lamella exists in the four clinical varieties of porokeratosis; porokeratosis of Mibelli, linear porokeratosis, porokeratosis plantaris et palmaris disseminata (PPPD), and disseminated superficial actinic porokeratosis (DSAP). However, porokeratosis plantaris

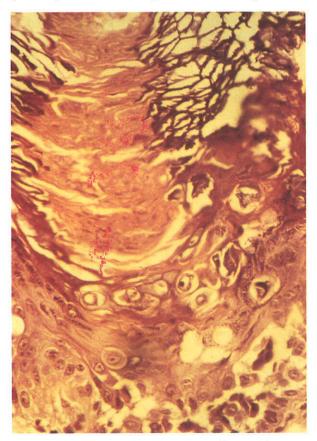


Fig. 4: Higher magnification shows "cornoid lamella" with epidermal cells at its base that have pyknotic nuclei with perinuclear edema and no granular cells. H & E X400.

discreta and porokeratotic eccrine duct nevus show cornoid lamella-like parakeratotic column.⁴

Cornoid lamellation is found in a variety of other conditions particularly in verruca vulgaris and solar keratosis.⁵ Also, it has been reported in cutaneous phaeohyphomycosis.⁶

Although parakeratotic column in Darier's Disease has been mentioned $1987)^{7}$ yet (Ackerman, in most of dermatopathology textbooks cornoid lamellation has not been described in Darier's Disease. 1,3

It has been suggested that the presence of a clone of abnormal epidermal cells at the base of the parakeratotic column explains the lesions of porokeratosis.8 Electron microscopic examination reveals that many keratinocytes beneath the parakeratotic column show signs of degeneration.9 The parakeratotic column is composed of cells with pyknotic nucleus and a containing cytoplasm many organelles. Few dyskeratotic cells could be seen. 10 Wade and Ackerman⁵ considered cornoid lamellation a histologic reaction pattern. In porokeratosis plantaris discreta, the cornoid lamella-like parakeratotic column is believed to be caused by the traumatically induced keratotic plugging of distal eccrine sweat duct.11

The presence of cornoid lamella in diseases with different etiologies is not fully understood. Why in our case cornoid lamella occurred in trunk lesions but not in palm lesions? We think that more investigations are needed to clarify the exact pathogenesis and significance of cornoid lamellation.

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