A Comparative Histopathological Examination of Biopsies from Patients with Either the Dominant or the Gamborg Nielsen Type of Hereditary Palmoplantar Keratoderma

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It has been previously reported that two different genetic forms of hereditary palmoplantar keratoderma (HPPK) were found in the northernmost county of Sweden (Norr-botten) [1]: a dominant form corresponding to the clinical and histopathological description of the Unna-Thost variety [2, 3] and a presumed recessive form, named the Gamborg Nielsen type, which clinically and ultra-structurally differed from the dominant form of HPPK and from mal de Meleda [4]. Since no significant histopathological differences between the two forms were found, it was considered of interest to examine a more comprehensive material to definitely decide whether or not any light-microscopical differences existed. The study was performed in a blind design. The correlation between a mononuclear cell infiltrate, which generally is found in the Unna-Thost variety, and dermatophytosis was also studied [3-5].

Twenty-eight biopsies obtained from 14 patients with the dominant form and 5 biopsies from patients with the Gamborg Nielsen type of HPPK were fixed and stained with HE and PAS. Biopsies from patients with the dominant form of HPPK were taken from the anterior part of the heel as well as from the medial aspect of the feet, 1 cm within the demarcation zone of the hyperkeratosis, corresponding to the metacarpophalangeal joint of the 1st toe. Only one biopsy from patients with the Gamborg Nielsen type of HPPK was taken from the anterior part of the heel. Due to detachment of the stratum corneum, one biopsy was excluded.

Significant morphological differences between the dominant form and the Gamborg Nielsen type of HPPK could not be demonstrated (fig. 1). In the dominant form, regional differences seemed to be more pronounced than differences between the two forms of HPPK. Cultures for pathogenic fungi were performed in the dominant form and in the Gamborg Nielsen type. Dermatophytes were cultured from soles of 7 patients with the dominant form and were distributed as follows: Trichophyton rubrum 6 and T. mentagrophytes 1. Candida albicans was found in 2 patients. T. rubrum (2), Epidermophyton floccosum (1) and Candida albicans (1) were isolated from soles of the Gamborg Nielsen type. In all cases a mononuclear cell infiltrate was found together with dermatophytosis. Hyphae and spores were simultaneously demonstrated in PAS-stained sections.

The comparative histopathological examination of biopsies taken from patients with either the dominant form or the Gamborg Nielsen type of HPPK allowed no differentiation between them. Differences in thickness of cell layers and acanthosis (length of rete pegs) could not be demonstrated, which agreed with previously performed studies [6]. It was concluded that these
two forms of HPPK could not be differentiated according to their micromorphology but only according to their clinical and ultra-structural characteristics. In both forms of HPPK a subepidermal mononuclear cell infiltrate was found together with hyphae and spores, which corresponds to previously performed studies [7]. The existence of a mononuclear cell infiltrate should most likely be interpreted as a consequence of dermatophytosis. Previously it has been shown that the prevalence of dermatophytosis in the dominant form of HPPK is 37.6% and that a mononuclear cell infiltrate, hyphae and spores occurred significantly more often in patients with dermatophytosis [3,8]. Inflammatory mononuclear cell infiltrate and presence of hyphae and spores without corresponding culture of dermatophytes was also demonstrated, and probably dermatophytosis occurs more often than expected.

References