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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.
Expression of Keratins (K10 and K17) in Steatocystoma Multiplex, Eruptive Vellus Hair Cysts, and Epidermoid and Trichilemmal Cysts

Hana Tomková, M.D., Wataru Fujimoto, M.D., and Jiró Arata, M.D.

We compared the patterns of keratin 10 (K10) and keratin 17 (K17) expression in epidermoid cysts, trichilemmal cysts, eruptive vellus hair cysts, and steatocystoma multiplex. Epidermoid cysts expressed K10 and eruptive vellus hair cysts expressed K17, whereas trichilemmal cysts and steatocystoma multiplex showed expression of both K10 and K17. Our findings support the opinion that eruptive vellus hair cysts, which stained negative for K10, and steatocystoma multiplex are distinct entities and not variants of one disorder.

Key Words: Keratin 10—Keratin 17—Steatocystoma multiplex—Eruptive vellus hair cyst—Trichilemmal cyst—Epidermoid cyst.

Keratins form one of six classes of intermediate filaments and are the major part of the cytoskeleton of epithelial cells. According to their molecular characteristics, keratins can be further classified as subfamily I (type I, acid keratins) and subfamily II (type II, neutral/basic keratins) (1,2). In tissues, they are expressed as pairs containing one member from each subfamily, and the expression of the pair is determined by the state of differentiation and proliferation of the tissues (2).

Both keratin 10 (K10) and keratin 17 (K17) belong to type I keratins, but they show distinct tissue distributions. K10 is formed in the epidermis and the follicular infundibulum by suprabasal keratinocytes (1). It is also formed by luminal cells of the eccrine duct (3) as well as by cells of the sebaceous gland and duct (1,4). K17 is normally expressed in the outer root sheath of the lower portion of the hair follicle, in the sebaceous duct and gland (1,4), and in the myoepithelial cells of the eccrine gland. Luminal cells of the eccrine duct showed only a weak and variable reaction to K17 (3). K17 has also been reported to be expressed in psoriasis (5) as another marker of keratinocyte hyperproliferation, besides K6 and K16 (6). We undertook an immunohistochemical study to determine the expression of K10 and K17 in epidermoid cysts, trichilemmal cysts, eruptive vellus hair cysts, and steatocystoma multiplex.

MATERIALS AND METHODS

We studied 5 biopsy specimens of epidermoid cysts, 5 specimens of trichilemmal cysts, 7 specimens of steatocystoma biopsied from 5 cases of steatocystoma multiplex or simplex, and 2 biopsy specimens from 2 patients with eruptive vellus hair cysts.

The studies were performed on formalin-fixed, paraffin-embedded sections, which were deparaffinized and rehydrated in xylene and graded ethanol solutions. Before staining for K17, the sections on slides were boiled...
in 10 mM citrate buffer. After inactivation of endogenous peroxidase activity and preincubation with 1% bovine serum albumin/10% skim milk, the sections were incubated overnight with the following monoclonal antibodies (MoAbs): monoclonal mouse anti-human cytokeratin 10 (Dako-CK10, DE-K10; 1:50, DAKO A/S, Denmark) and monoclonal mouse anti-human cytokeratin 17 (Dako-CK17, E3; 1:10, DAKO A/S, Denmark). The slides were then developed with biotinylated goat anti-mouse immunoglobulin (Histofine, Nichirei, Tokyo, Japan) and streptavidin conjugated with horseradish peroxidase (Histofine), followed by visualization of the reaction by diaminobenzidine (DAB) solution, counterstaining with 1% methylgreen, and mounting with Entellan Neu (Merck). We considered negative immunoreactivity of the epidermis and the outer root sheath as a negative control for anti-human cytokeratin 17 and 10, respectively.

RESULTS

The results of immunostaining for K10 and K17 in our study, aimed particularly at possibly differentiating steatocystoma multiplex from eruptive vellus hair cysts by immunohistochemistry, were as follows. In eruptive vellus hair cysts, in contrast to the overlying epidermis, cells lining the cyst did not express K10 (Fig. 1A), but showed strong immunoreactivity for K17 (Fig. 1B). In steatocystoma multiplex and simplex, unlike in eruptive vellus hair cysts, expression of both K10 and K17 was demonstrated in the suprabasal cells lining the cyst, as shown in Fig. 2A and B. In trichilemmal cysts, the presence of K10 and K17 was also observed in suprabasal lining cells (Fig. 3A and B). In epidermoid cysts, K10 was expressed in the suprabasal compartment of the cyst wall (Fig. 4), whereas K17 showed no immunostaining (data not shown).

DISCUSSION

An epidermoid cyst originates in the follicular infundibulum as a result of proliferation, retention, and hyperkeratosis (7). In regions devoid of pilosebaceous follicles, such as palms and soles, it can arise from implantation of epidermal cells into the dermis subsequent to trauma (8,9). Histologically, the wall of an epidermoid cyst is composed of the stratified squamous epithelium containing a granular layer. Horny material arranged in laminated layers is found in the cyst lumen (10). Our study demonstrated the presence of K10 in the epidermoid cyst wall in exactly the same fashion as in the normal epidermis and the follicular infundibulum.

A trichilemmal cyst is a keratin-containing cyst (10), the wall of which is derived from the outer root sheath of the lower portion of the hair follicle between the hair bulb and the orifice of the sebaceous duct (7). Its epithelial lining shows abrupt keratinization without the

FIG. 1. Eruptive vellus hair cyst. Negative keratin 10 (a) and positive keratin 17 (b) immunohistochemical staining.
presence of stratum granulosum (10). However, unlike the outer root sheath, the lining of a trichilemmal cyst stains positive with the serum raised against hair keratin extract (11). In our study, trichilemmal cysts showed reactivity not only for K17, normally expressed in the outer root sheath, but also for K10, which is not expressed in the lower hair follicle below the level of the sebaceous gland (1,4). Therefore, our findings further support those of Cotton et al. (11) indicating that pilar cysts show similarity but not antigenic identity with the outer root sheath of the hair follicle.

Steatocystoma multiplex (or simplex) is a cyst or, more precisely, a cystic hamartoma (12), lined by thin, wavy, sebaceous duct epithelium, containing sebaceous lobules of varying size as well as individual sebocytes.

There is no granular layer. Steatocystoma is associated with a vellus hair follicle, the hairs of which may be trapped in the cyst cavity (13). Electron microscopy showed that steatocystoma multiplex has features similar to the sebaceous duct and sebaceous gland (13). The expression of K10 and K17, also expressed in the sebaceous duct and gland (1,4), confirms the origin of the lining of steatocystoma.

Eruptive vellus hair cysts are small cysts arising from the structures of vellus hair follicles. The pathogenesis is unknown (7). A possible pathogenetic mechanism has been proposed by Esterly et al. (14): An initially abnormal vellus hair follicle, with keratinous plugs at the follicular infundibulum, deflects vellus hair shafts to the deeper part of the follicle, resulting in its cystic dilatation and mal cyst keratin small hair erup- catin hairs wall were tion, fundi part 1 We s in the junctu feren theliu press Tha atocy as we of bot cysts gators mas (.

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FIG. 2. Steatocystoma stained for keratin 10 (a) and keratin 17 (b).

FIG. 3. Trichilemmal cyst. Staining for keratin 10 (a) and keratin 17 (b).
tive vellus hair cysts and steatocystoma multiplex are distinct entities and not variants of one disorder, as suggested by some investigators (20,21).

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REFERENCES