Trichilemmal Keratosis Manifesting as a Cutaneous Horn

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CASE REPORT

A 45-year-old man presented with an asymptomatic, slowly growing, keratotic nodule on his left flank for one year. Physical examination revealed a pea-sized, horn-like plaque protruding from a brownish nodule (Fig. 1 A, B). The tumor had been present for years as a cyst-like lesion before, but the tumor ruptured 1 year before this consultation. Later, a horn-like structure developed on the center of it. The lesion was excised en bloc under the impression of keratoacanthoma. Sections showed a cup-shaped epidermal proliferation with overlying compact hyperkeratosis and focal parakeratosis (Fig 2). The epithelium invaginates into dermis and form connecting lobules. Palisading basal cells were seen at the peripheral of the lobules (Fig. 3). The epithelium were composed of pale keratinocytes that transform into a keratinized layer without a granular layer formation (Fig. 4). There is a clear distinction between trichilemmal keratinization and epidermoid keratinization of the perilesional skin. Vascular dilatation and chronic inflammatory cell infiltrations are noted in the dermis. The CD34 staining in this case shows positive in the peripheral cells of the lesion (Fig. 5).

DISCUSSION

Headington first used the term trichilemmal keratosis in 1976 to describe a rare keratinizing tumor resembling a cutaneous horn or hyperkeratotic actinic keratosis. The pattern of keratinization is similar to that seen in follicular isthmus and trichilemmal cyst. However, very few cases were studied at that time. In 1979, Brownstein proposed the name trichilemmal horn to describe the same condition. In his observation, only 19 trichilemmal horn were found after he had examined 75000 specimens (incidence: 0.021%). Among them, eleven patients were female and six patients were male. The age ranged from 16 to 72 years with median age of 50 years. DiMaio reviewed the literature and declared that only 28 cases of trichilemmal keratosis were reported so far. Clinically trichilemmal keratosis are solitary, keratotic, horn-like structures. The most common affected areas are the extremities, head and back in decreased frequency. Histologically, there is marked hyperkeratosis and verrucous epidermal hyperplasia. The epithelium showed trichilemmal keratinization in which the keratinocytes abruptly transform into dense lamellar keratin without formation of a granular layer. The keratinocytes were large polygonal cells with abundant eosinophilic cytoplasm. The pattern of keratinization is
similar with that of follicular isthmus and trichilemmal cyst. Contiguous lobule extended from epidermis into dermis. The dermal lobules were composed of central trichilemmal keratinization and peripheral palisading of basal cells, which simulated a miniature trichilemmal cyst. In some cases, the dermal lobules may connect with epidermis and extrude their contents into the surface.

The pathogenesis of trichilemmal keratosis is unclear. CD 34 is a specific marker for external root sheath epithelium of hair follicles and tumors derived from or differentiated toward this type of epithelium. Positive CD34 immunostaining has been demonstrated in cases of trichilemmal kera-
Therefore trichilemmal keratosis was considered to be originated from outer root sheath of hair follicles. However trichilemmal keratosis occurred on the palm had been reported, which opposed the theory of hair follicle origin and might represent a phenotypic change of epidermal keratinocytes.\(^4\)\(^,\)\(^7\) In our case, the CD34 immunostaining is positive which may imply that the trichilemmal keratosis origins from outer root sheath. Kimura observed intranuclear dense particles in trichilemmal horn and suggested that it’s occurrence may be related to viral infection.\(^8\) The relationship between the development of trichilemmal keratosis and trichilemmal cyst had been proposed. Poblet reported two cases of trichilemmal keratosis located in the area where a previous cyst ruptured into the surface.\(^4\) Both cysts located in the scalp and were highly suspected of trichilemmal cysts. Poblet suggested that the epithelium of ruptures trichilemmal cyst proliferated and produced a trichilemmal keratosis.\(^4\) In our patient there was a ruptured cyst prior to the development of trichilemmal keratosis. The patient’s father had a history of perforating pilomatricoma. But we didn’t know the nature of the cyst in this patient.

Cutaneous horn (cornu cutaneum) is a conical projection of keratinized material. It may arise from preexisting benign, premalignant and malignant conditions. The differential diagnosis of trichilemmal keratosis includes other cutaneous horn derived from various benign, premalignant and malignant disorders. The proportion of benign, premalignant and malignant changes of the base varied in different series. The most common lesions found on the base were seborrheic keratosis, verruca vulgaris, actinic keratosis and squamous cell carcinoma. Clinical diagnosis of a cutaneous horn is difficult. The diagnosis of cutaneous horn must depend on surgical excision of the base of the lesion.

It is concluded that we describe a cutaneous horn arising from the underlying epithelium showing trichilemmal keratinization. Trichilemmal keratosis or trichilemmal horn had been used to describe these rare and unique conditions. Surgical excision of the base is always needed to establish an accurate diagnosis.

REFERENCES