A Case of Trichilemmal Carcinoma Showing a Feature of Cutaneous Horn

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Trichilemmal carcinoma is a rare malignant neoplasm of the hair follicle, which is derived from or differentiates towards cells of the outer root sheath. We report a case of trichilemmal carcinoma in an 83-year-old female patient. She presented with a tender dome-shaped crusted papule showing a feature of cutaneous horn on the forehead, which was first detected 6 months before. Histopathologically, lobular patterned tumor cells with peripheral palisade of basaloid cells, nuclear atypia, and clear or pale, PAS-positive, diastase-sensitive cytoplasm were observed beneath the marked hyperkeratosis. After the diagnosis, total excision was done. The patient has been free of recurrence or metastasis till now.

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Key Words : Trichilemmal carcinoma, Cutaneous horn

Trichilemmal carcinoma is an uncommon cutaneous neoplasm, which occurs in sun-exposed areas of aged persons. It is thought to be originated from the external root sheath (trichilemma) of the hair follicle[1]. Despite the frequent presence of histologically malignant features, it has a relatively benign clinical behavior[2]. Cutaneous horn is the clinical term for a circumscribed, conical, markedly hyperkeratotic lesion in which the height of the keratotic mass amounts to at least half of its largest diameter[3]. We report a case of trichilemmal carcinoma showing a feature of cutaneous horn.

CASE REPORT

An 83-year-old female patient presented with the skin lesion on the forehead, which was about 1cm in diameter and first detected about 6 months before(Fig. 1). The lesion had increased in size gradually and had bled recurrently. We could observe the tender dome-shaped crusted papule showing a feature of cutaneous horn on the forehead.

Shave biopsy was done under the impression of squamous cell carcinoma. Histopathologically we could observe the multiple lobular or trabecular proliferation of tumor cells beneath the marked hyperkeratosis (Fig. 2A). At the periphery of each lobule, the cells were in palisading arrangement and became swollen toward the central area with a pale eosinophilic or clear cytoplasm. Most of the tumor cells showed nuclear atypia or pleomorphism and some had mitotic figures. The clear cytoplasm of tumor cells contained PAS-positive and diastase-sensitive material (Fig. 2B, 2C). On immunohistochemical staining, positivity in high molecular weight cytokeratin(Fig. 3) and EMA (Fig. 4), and negativity in CEA, S-100, and low molecular weight cytokeratin were observed.

Total excision was done under the diagnosis of trichilemmal carcinoma, and the patient has been free of recurrence or metastasis till now.

DISCUSSION

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Trichilemmal carcinoma usually appears in elderly individuals, primarily on sun-exposed areas. The tumor typically manifests as a single, superficial nodule, measuring less than 2cm that commonly undergoes ulceration, and often misdiagnosed as a basal cell carcinoma or squamous cell carcinoma. To date, the clinical behavior seems to be relatively benign, without the evidence of either metastasis or local recurrence[4]. Microscopically, trichilemmal carcinoma consists of relatively well circumscribed multiple lobules or trabeculae with a peripheral palisade of basaloid cells. The prominent tumoral cells are polygonal, with abundant clear or pale, PAS-positive, diastase-sensitive cytoplasm. Striking cytological atypia and a high mitotic index are usually present[4]. Trichilemmal keratinization, characterized by abrupt keratinization without a granular layer, is a characteristic finding[5]. Variable degrees of basilar or full-thickness interfollicular epidermal spread, with an abrupt or pagetoid interface with adjacent, cytologically unremarkable keratinocytes can be seen. Dermal invasion, typically extending into reticular dermis, accompanied by a plasma cell-rich lymphocytic infiltrate also can be found[6]. Immunohistochemically, trichilemmal carcinoma is positive in PAS and high molecular weight cytokeratin, but usually negative in CEA, EMA. In our case, positivity in EMA was observed. There are reports in which trichilemmal carcinoma showed positivity in EMA[4].

Histogenesis of trichilemmal carcinoma remains unclear. There have been reports of cases with a history of arsenic intake, in which both Bowen's disease and trichilemmal carcinoma occurred. Cases in which trichilemmal carcinoma occurred in a burn ulcer, a scar of discoid lupus erythematosus and a scar of tuberculosis verrucosa cutis have also been reported[7]. Chronic mechanical stimulation is a carcinogenic factor in trichilemmal carcinoma as well. From the fact that trichilemmal carcinoma occurs in sun-damaged skin of elderly people, Misago et al[7] insisted that considerable cases of trichilemmal carcinoma might develop de novo from actinically damaged skin like squamous cell carcinoma.

The term, cutaneous horn, has been used to describe either the morphologic or microscopic epithelial changes that may be present overlying certain benign, premalignant, and malignant lesions such as actinic keratoses, seborrheic keratoses,

Table 1. Previous reports of trichilemmal carcinoma in Korean literature

Case Report	Age/Sex	Site	PAS/Diastase	Immunohisto-chemistry	Treatment after diagnosis	Recurrence or metastasis
Park et al[10]	79/F	Right mandibular area	PAS(+) Diastase-labile	HMCK*(+) CK19(+) CEA(-) EMA(-) S-100(+)	Total excision	No
Ahn et al[11]	88/F	Right preauricular area	PAS(+)	HMCK*(+) CEA(-) EMA(-)	Total excision	No
Lee et al[12]	53/M	Left preauricular area	PAS(+) Diastase-labile		Total excision	No
Lim et al[13]	75/M	Lower lip Diastase-labile	PAS(+)		Total excision	No
Our case	83/F	forehead	PAS(+) Diastase-labile	S-100(-) HMCK*(+) EMA(+) CEA(-)	Total excision	No

*HMCK: High molecular weight cytokeratin

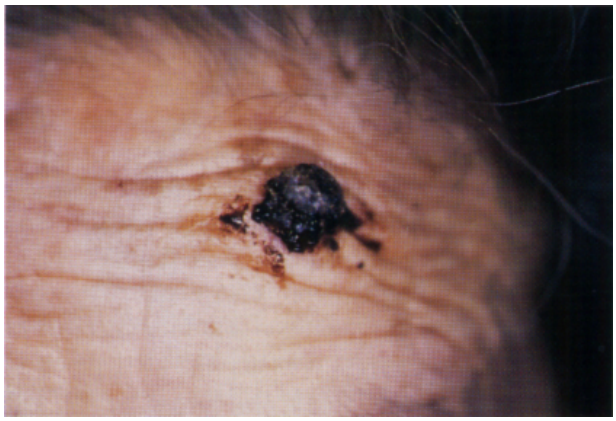

Fig. 1. Dome-shaped crusted papule showing a feature of cutaneous horn on the forehead.

squamous cell carcinomas, and verrucae vulgares. Trichilemmal keratosis(trichilemmal horn), a benign variant of trichilemmal carcinoma, is a keratinizing tumor which resembles a cutaneous horn or a hyperkeratotic actinic keratosis. Histologically, the lesion shows an epithelial lobules, composed of pale staining keratinocytes. A few germinal buds are noted to be proliferating from the periphery of the epithelial lobule, and there is margination of the basal cells at the periphery of the lobule. Also, there is verrucous hyperplasia and prominent orthokeratosis that is characterized by an abrupt transformation of the keratinocytes into the lamellar keratin without the formation of a granular layer[8]. In our case, hyperkeratosis overlying

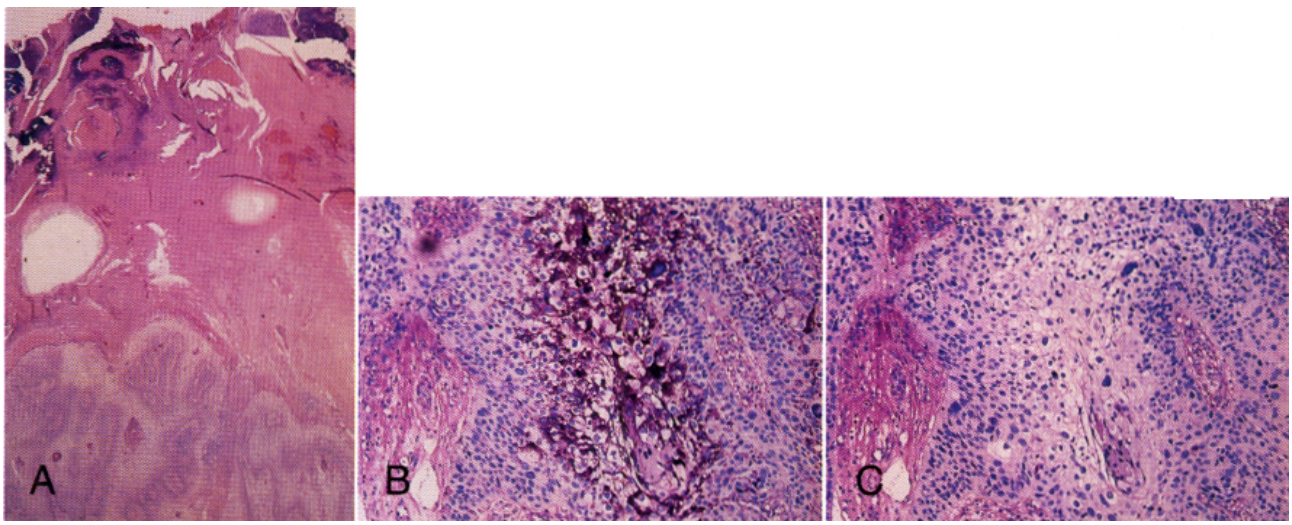

Fig. 2A. Multiple lobular or trabecular proliferation of tumor cells under the marked hyperkeratosis(H&E stain, ×10) B. The tumor shows PAS-positivity(PAS stain, ×100) C. The tumor shows diastase-sensitivity (d-PAS stain, ×100).

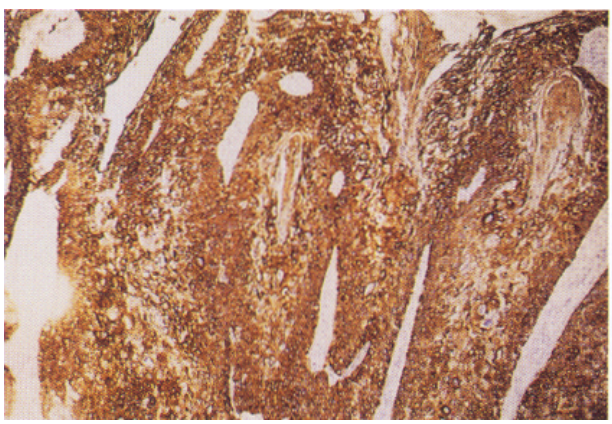

Fig. 3. The tumor shows positivity for high molecular weight cytokeratin(High molecular weight cytokeratin, ×40).

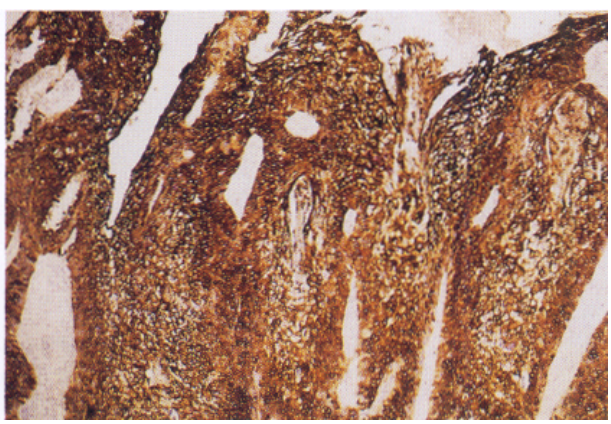

Fig. 4. The tumor shows positivity for EMA(EMA, ×40).

trichilemmal carcinoma showed a feature of cutaneous horn. The differential diagnosis of trichilemmal carcinoma with other malignant clear cell tumors of the skin is necessary. The distinction from clear cell squamous cell carcinoma is based primarily upon the architectural pattern rather than cytological findings. In clear cell squamous cell carcinoma, there is absence of trichilemmal keratinization, peripheral palisade of basaloid cells, and cytoplasmic glycogen[5,9]. Clear cell basal cell carcinoma may mimic trichilemmal carcinoma. But areas of clear cell change often only involve a minor portion of an otherwise conventional basal cell carcinoma. Sebaceous carcinoma show tumor lobules composed of eosinophilic cells at their peripheries and vacuolated or clear cells at their centers and the absence of PAS positivity. Demonstration of cytoplasmic lipid by special stain or electron microscopy supports the diagnosis[5,9]. Malignant proliferating trichilemmal tumor may be essentially identical with trichilemmal carcinoma. But the former is more keratinized and has a stronger tendency toward metastasis than the latter[2]. The most difficult differential diagnoses are those of actinic keratosis and Bowen's disease. Clinical and histological findings suggest that these lesions may be etiologically related. Actinic keratosis is thought to be involved in pilosebaceous units, but usually affects only the acrotrichium or infundibulum, and largely spares the outer root sheath. Bowen's disease may extend into pilosebaceous structures, and, unlike actinic keratosis, may appear to completely replace the infundibulum. In addition, Bowen's disease may have a pagetoid appearance. But Bowen's disease does not exhibit a dominant lobular pattern of growth, trichilemmal keratinization and outer root sheath involvement below the follicular infundibulum[6].

The recommended treatment for trichilemmal carcinoma is complete but conservative surgical excision due to the locally aggressive growth and recurrence or metastasis is not yet observed[2]. Table 1 summarizes the previously reported cases of trichilemmal carcinoma in Korean literature.

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