A Case of Trichilemmal Carcinoma

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We report a case of trichilemmal carcinoma in a 79-year-old woman who presented an atrophic scar remaining after excision of a recurrent nodule on the right mandibular area. Her past history revealed that there had been a painful pea-sized brownish nodule on the same site for two years. Eleven months before presentation, it had been excised but recurred 9 months later. Histopathologic findings showed a clear cell neoplasm with trichilemmal keratinization. The tumor cells showed PAS-positive cytoplasm and cytologic atypia with a few mitotic figures. Immunohistochemical staining for high molecular weight cytokeratin was positive but carcinoembryonic antigens and epithelial membrane antigens were all negative.

To our knowledge this is the first report of trichilemmal carcinoma in Korea.

(Ann Dermatol 8:(1)61~65, 1996).

Key Words : Trichilemmal carcinoma

Trichilemmal carcinoma (TLC) is a rare adnexal tumor of external hair sheath origin[1]. Headington[2] first proposed the term for a "histologically invasive, cytologically atypical clear cell neoplasm of adnexal keratinocytes which is in continuity with the epidermis and/or follicular epithelium". Since then, about 82 cases of this unusual tumor have been reported in the literature such as trichilemmal carcinoma, tricholemmal caricinoma, malignant trichilemmoma, tricholemmocarcinoma[1,3-13]. Despite the frequent presence of histologically malignant features, it has a relatively benign clinical behavior[3]. It is rarely, if ever, capable of regional or distant metastases[3,4]. Thus, the correct diagnosis of this tumor allows its distinction from other clear cell epithelial cutaneous neoplasms with a known aggressive behavior and prevents unnecessary radical procedures.

We report herein a case with the histopathologic and immunohistochemical findings of TLC and discuss the differential diagnosis from other clear cell tumors.

REPORT OF A CASE

A 79-year-old woman visited our department for the evaluation of an asymptomatic atrophic scar remaining after excision of a recurrent nodule on the right mandibular area (Fig. 1). Her past history showed that there had been a painful pea-sized brownish nodule on the same site for 2 years. Eleven months before presentation it had been excised at a private clinic without pathologic diagnosis but recurred 9 months later and was also excised at another local clinic 1 month prior to her visit. At that time it was diagnosed as a squamous cell carcinoma. When she came to us her general condition was relatively healthy and on physical examination, cervical lymph nodes were not palpated and there were no other similar skin tumors on the rest of her body. A routine laboratory tests including complete blood cell count, urinalysis,

Received July 12, 1995.
Accepted for publication November 15, 1995.
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The part of this article was presented at the 2nd meeting of Dermatopathology Workshop on Feb 25, 1995.

Annals of Dermatology
Vol. 8, No. 1, January 1996

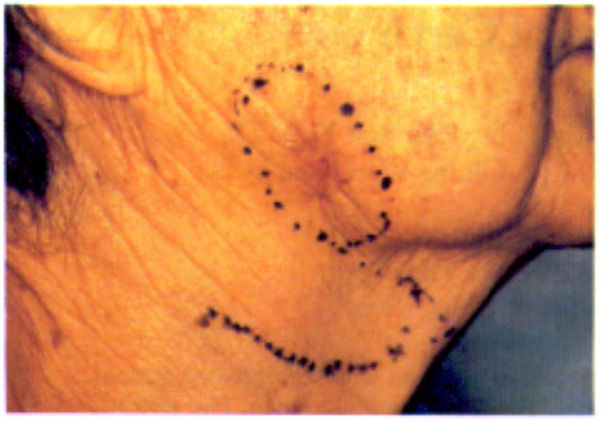

Fig. 1. Asymptomatic erythematous atrophic scar on the right mandibular area before the local flap excision.

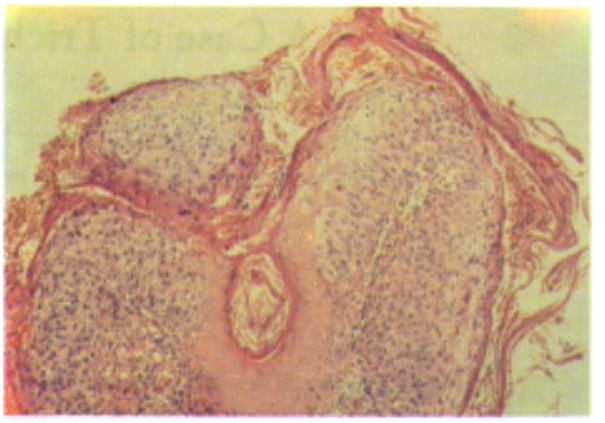

Fig. 3. Pagetoid intraepidermal involvement by the tumor cells largely efface the epidermis adjoining the follicular infundibulum (H & E, ×100).

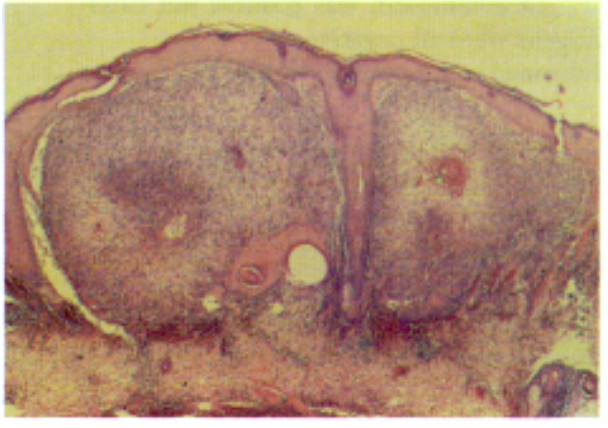

Fig. 2. Two lobules of clear cells are centered on a pilosebaceous structure and show central keratinization with palisading of the peripheral cells (H & E, × 40).

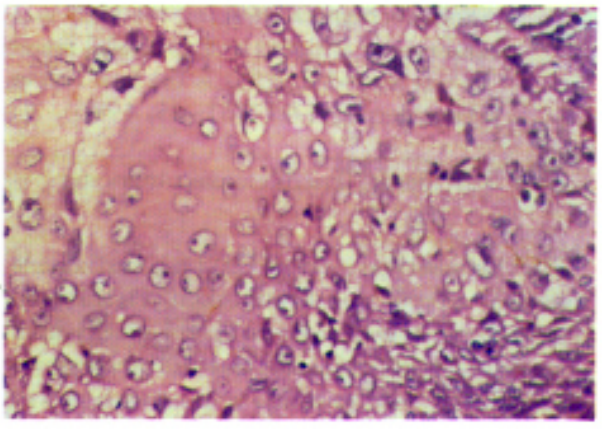

Fig. 4. Abrupt trichilemmal keratinization without keratohyaline granules is shown (H & E, ×200).

electrocardiogram and chest X-ray were negative or within normal limits. A head and neck computerized tomography revealed no metastatic evidences.

The atrophic scar was totally excised under local anesthesia and then covered with a pedicle flap. Specimen from the atrophic scar revealed mild perivascular mononuclear cell infiltration and scar formation but no malignant cells. We also reviewed the previous excision specimen taken from the local clinic 1 month previously. Histopathologic examination showed a tumor composed of multiple lobules or convoluted trabeculae extending from the epidermis to the dermis and it was in continuity with the normal epidermis at the edges. Many columnar cells in the peripheral layer of some of the lobules were arranged in a palisading pattern and they became swollen toward the central area with a pale eosinophilic or clear cytoplasm (Fig. 2). In some areas pagetoid tumor cells replaced the epidemis (Fig. 3) and trichilemmal keratinization without forming a granular layer was shown (Fig. 4). Most of the tumor cells showed a bowenoid pattern with atypical nuclei or pleomorphism and some had mitotic figures (Fig. 5). The clear cytoplasm of tumor cell contained PAS-positive, diastase-labile material (Fig. 6).

To evaluate the origin of tumor cells, we performed immunohistochemical staining with a series of monoclonal antibodies. Immunohistochemical staining revealed positive for high molecular weight cytokeratin (Fig. 7) but negative for cy-

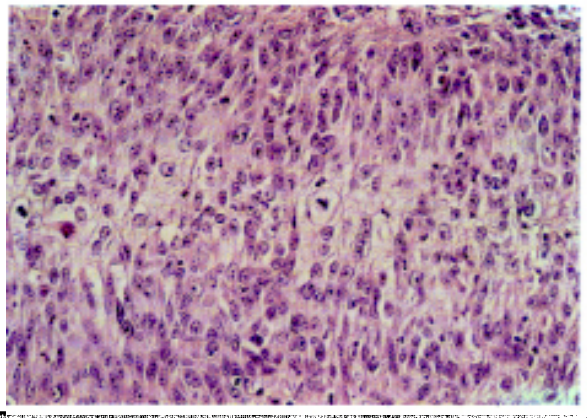

Fig. 5. The tumor cells contain pleomorphic large round to oval nuclei with irregular nuclear contour, increased nucleocytoplasmic ratio and show some mitotic figures (H & E, ×200).

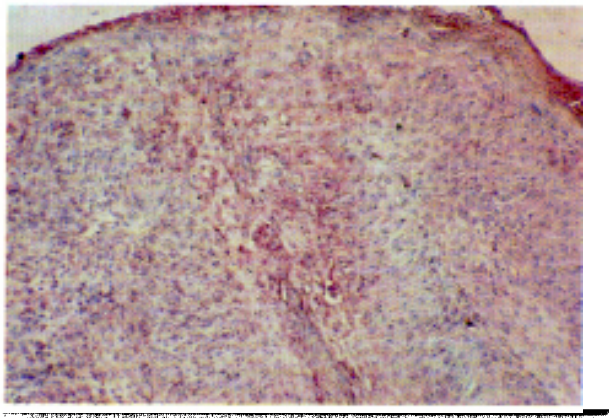

Fig. 7. Positive for high molecular weight cytokeratin (High molecular weight cytokeratin, ×100).

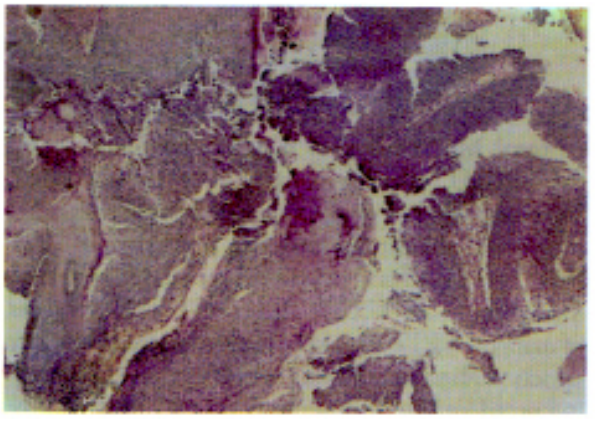

Fig. 6. Positive periodic acid-Schiff stain in tumor cells (PAS, ×40).

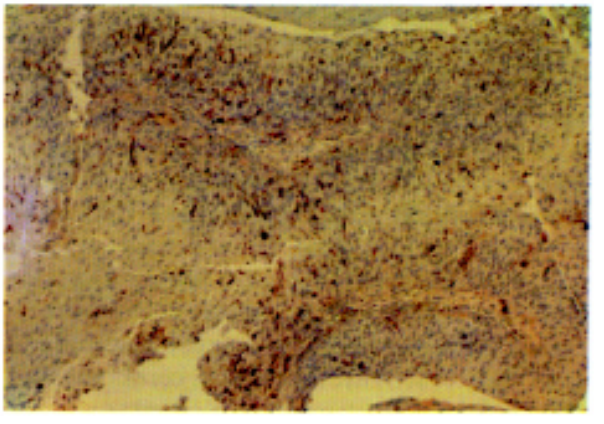

Fig. 8. Dendritic cells positive for S-100 protein (S-100, ×100).

tokeratin 19. Carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA) also showed negative immunostaining. S-100 staining was focally positive in dendritic cells (Fig. 8) but silver nitrate impregnation was negative.

No evidence of recurrence or metastasis has been observed 6 months postoperatively.

DISCUSSION

Trichilemmal carcinoma (TLC) is a rare malignant neoplasm of the hair follicle which is derived from or differentiates towards cells of the outer sheath. Clinically, TLC usually occurs as an asymptomatic solitary slow-growing red papule, keratotic nodule or indurated plaque with a smooth surface or crusted surface on sun-exposed, hair-bearing areas of the elderly. Although it is capable of multiple local recurrence, it rarely shows metastasis[3,4]. Generally the duration of TLC proir to diagnosis ranges from 2 months to 3 years and the female-male ratio is 3:1.6[4]. In our case, the tumor occurred as a recurrent painful brown nodule in a 79-year-old woman. It is unusual that she had painful symptom.

Histologically TLC is characterized by many features resembling the outer root sheath of the hair follicle such as lobules and trabeculae of clear cells rich in glycogen, palisading of the peripheral cells rimmed by thickened basement membranes and horn microcysts with trichilemmal keratinization[5]. The tumor is frequently connected to the follicular epithelium and the interfollicular epidermis and usually invades the dermis[4]. Our case

Table 1. Histopathologic differential diagnosis of trichilemmal carcinoma

	Trichilemmal carcinoma	Trichilemmoma	PTT	Malignant PTT	Sebaceous carcinoma	Clear cell BCC	Clear cell SCC	Our case
Major cell component	celar	clear	squamous	squamous	clear	basal	squamous	clear
Cell atypia & mitosis	+	−	−	+ +	+ +	+ +	+ +	+
Infiltrative pattern	+	−	−	+	+	+	+	+
Peripheral cell palisading	+	+	±	±	±	+	−	±
Lumen formation	−	−	+	+	+	+	−	−
Trichilemmal keratinization	+	−	+	+	−	−	−	+
PAS	+	+	+	+	−	−	−	+

* PTT:proliferating trichilemmal tumor * BCC:basal cell carcinoma * SCC:squamous cell carcinoma
* PAS:periodic acid-Schiff stain

also showed some features resembling the outer root sheath of the hair follicle, but peripheral palisading of nuclei was not frequent except in some areas. Cytologically malignant features of TLC can be confirmed by detecting foci of frank nuclear anaplasia and atypical mitotic figures. Our case exhibited similar striking nuclear atypia with irregular nuclear contour, prominent nucleoli and some mitotic activity. In addition, consistent with previously reported cases and the criteria suggested by Headington[14], we found no evidence of vascular or perineural invasion in our case. These histologic findings support our finding that our case represents a malignant adnexal neoplasm of outer root sheath origin.

The main importance of this tumor lies in their potential for errors in the interpretation on the histopathological diagnosis. The differential diagnosis for TLC includes other benign and malignant skin neoplasms (Table 1). Trichilemmoma is distinguished from TLC by the absence of infiltrative growth pattern, cellular atypia and frequent mitoses in this tumor[4]. Proliferating trichilemmal tumor (PTT) may be another source of confusion with TLC. But unlike TLC, areas showing clear cell changes and palisading of columnar epithelium often comprise only a minor portion of PTT[4]. Furthermore, PTT arises in association with a preexisting trichilemmal cyst typically occurring in the scalp of older women and it lacks an infiltrative growth pattern[5]. Malignant PTT may be essentially identical with TLC from the point of view that malignant transformation of epidermal cyst corresponds with squamous cell carcinoma[6]. But malignant PTT is more keratinized and has a stronger tendency toward metastasis than TLC[15,16]. Moreover, the diagnosis of malignant PTT should be applied to those cases in which malignant transformation of the preexisting proliferating trichilemmal cyst is histologically evident and to those cases with intradermal cystic lesions without connection to the epidermis, which suggests a malignant transformation of the trichilemmal cyst[17,18]. Sebaceous carcinoma can be also similar to TLC. However, this tumor cell has foamy, lipid-rich cytoplasm with centrally located nuclei and the features of outer root sheath differentiation are characteristically absent[3]. Basal cell carcinoma (BCC) may mimic TLC. But areas of clear cell change often only involve a minor portion of an otherwise conventional BCC and the peculiar fibromyxoid stromal reaction and perilesional stromal clefting that typify BCC are not encountered in TLC[7]. The differential diagnosis for TLC from squamous cell carcinoma (SCC) with clear cell change is very difficult. However the distinction from clear cell SCC can be based primarily upon the architectural pattern rather than cytologic findings[1]. The growth patterns of TLC are characterized

by well-circumscribed lobules of clear cells and peripheral cells of lobules tend to palisade. Moreover, in clear cell SCC, there is an absence of cytoplasmic glycogen and keratinization is of the epidermal rather than the trichilemmal type[1].

The present case showed a glycogen-rich, clear cell malignant neoplasm with trichilemmal keratinization. The immunohistochemical staining that was positive for high molecular weight cytokeratin and negative for cytokeratin 19, CEA and EMA suggest that the origin of the tumor cells is squamous epithelium rather than glandular epithelium. We could also exclude trichilemmoma and PTT which show a benign cytology and exclude sebaceous carcinoma, clear cell BCC and clear cell SCC which are negative for PAS stain. In the case of malignant PTT, we could exclude it from TLC because no evidence of preexisting proliferating trichilemmal cyst but continiuty to the epidermis was found in our case. Although a few dendritic cells which were positive for S-100 protein may be Langerhans cells, further studies for the recognition of these cells is required.

The recommended treatment for TLC is the complete but conservative surgical excision due to the locally aggressive growth and no recurrence or metastasis over a follow-up period of 8-10 years is expected after complete excision[3,4]. In our case, no evidence of recurrence or metastasis has been observed 6 months postoperatively.

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