case report

Recurrent trichilemmal carcinoma with a large cutaneous horn formation

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Ann Saudi Med 2012; 32(6): 642-645

DOI: 10.5144/0256-4947.2012.01.7.1530

We describe an unusual case of trichilemmal carcinoma accompanied by a large horn formation in a 79-year-old man who presented with a brown tumor mass including a 15-cm-long horn on the vertex for one and a half years. Two months after the surgical removal of the primary tumor, the tumor recurred with a dimension 10×8×8 cm. The tumor was then excised again and the wound was covered using skin from the patient's back. Microscopically, the tumor showed histology consistent with trichilem¬mal carcinoma. After 2 years, the patient had not shown any recurrence or metastases. We highlight the features that distinguish trichilemmal carcinoma from the other malignant tumors, such as squamous cell carcinoma, basal cell carcinoma, nodular melanoma, or keratoacanthoma.

Trichilemmal carcinoma is a rare cutaneous tumor that is usually solitary and occurs in the sun-exposed areas of elderly individuals.[1] Dozens of case reports of this malignancy have been published. We report an unusual case of trichilemmal carcinoma with a large horn formation. After the surgical removal of the primary tumor, the recurrent tumor was excised again, and the patient has not experienced further recurrence or metastases over the past 2 years.

CASE

A 79-year-old man presented with a brown tumor mass on the vertex of his head, which had been present for one and a half years. Initially, the tumor appeared as a small brown nodule with no obvious inducements, and one and a half years later, the nodule was 15 cm long and resembled a horn (**Figure 1A**). Soon after that, the tumor appeared to be ulcerated and accompanied by bleeding with the appearance of grayish-white mucous material. The patient felt pain and sought medical attention. The surface of the tumor horn was removed. Two months after the surgery, the tumor reappeared with a dimension 10×8×8 cm and was bleeding (**Figure 1B**). The patient's medical history showed that he had diabetes for 1 year and hypertension for 3 to 5 years. The patient did not have a significant head injury or trauma; however, he had evidence of alcohol and tobacco use, and a history of significant sun exposure throughout his life. Physical examinations revealed that the patient's heart, lung, and abdomen were functioning normally. Laboratory tests revealed high fasting blood glucose levels (12.11 mmol/L, normal range 3.9-6.1 mmol/L); however, other blood and urine tests were normal. A chest radiograph did not indicate any metastatic nodules. Skin examinations found the previously described tumor mass on the vertex, but no enlarged superficial lymph nodes.

The tumor was surgically excised and the wound was covered using the back trunk skin-grafting dermatome (**Figure 1C**). The tumor specimen was fixed in formalin and embedded in paraffin. Five-micron-thick sections were cut and stained with hematoxylin and eosin. Under the microscope, the tumor showed a lobular formation and plate-like growth of glycogen-rich clear cells with variable glycogen vacuolation, which were often surrounded by palisading cells and central keratinization (**Figure 2A,B**). Keratinaceous microcysts and small squamous eddies were also noted in the center of the lesion. The palisading basal cell layer resembled the external root sheath of hair follicles **Figure 1B**. A periodic acid-Schiff (PAS) stain-positive and diastase-resistant thickened basement membrane was partially

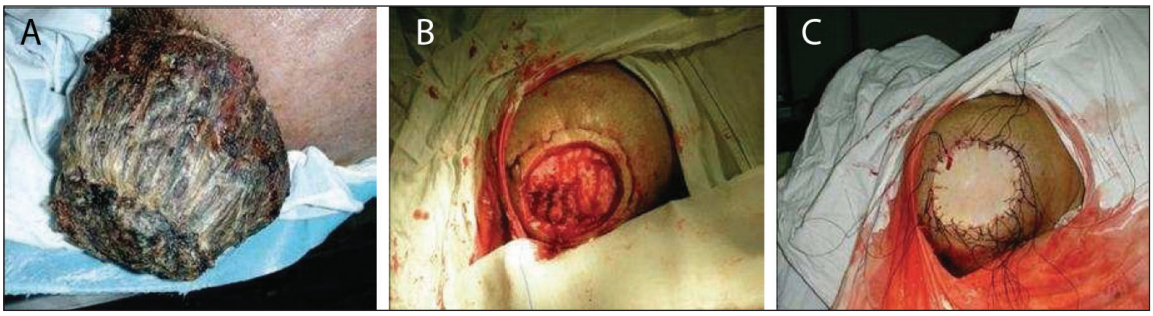

Figure 1. A. The primary tumor. The tumor mass with a 15-cm long horn shows bleeding and grayish-white mucous ulceration. B. The recurrent tumor. The tumor is 10 cm long. C. After excision, the wound was covered by using a back trunk skin-grafting dermatome.

surrounding the tumor cells (**Figure 3A**), and one or more hair follicles were always present. The tumor cells showed markedly atypical cellular and nuclear morphology **Figure 1B**. The immunostaining showed that pancytokeratins AE1/3 and cytoplasmic tyrosine kinase (HCK) (**Table 1**) were present in the tumor cell cytoplasm (**Figure 3B**), but CAM5.[2] and cytokeratin 7 (CK7) (**Table 1**) staining were both negative (**Figure 3C**). Therefore, a diagnosis of trichilemmal carcinoma with a large cutaneous horn formation was made. After the surgical removal of the primary tumor, the recurrent tumor was excised again, and the patient did not experience further recurrence or metastases over 2 years.

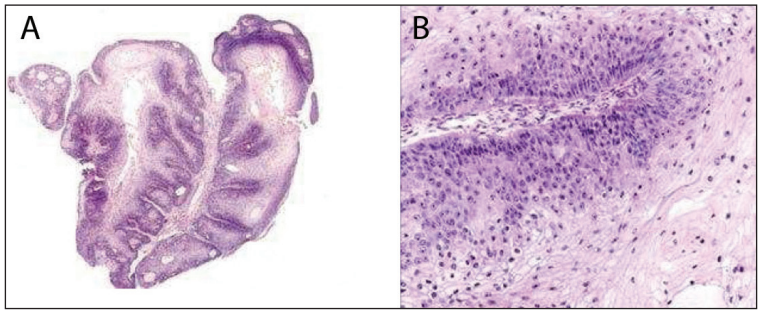

Figure 2. Tumor histology. HE stain shows lobular formation or plate-like growth of clear cells with keratinaceous microcysts and small squamous eddies. Marked cellular and atypical nuclei are present in tumor cells (A ×40, B ×100)

DISCUSSION

Trichilemmal carcinoma is a rare skin tumor that originates from the external root sheath of the hair follicle and is the malignant form of the usually benign trichilemmoma. The patient presented with a fast-growing tumor that reached 10 cm in 2 months after the primary tumor was excised. However, 2 years after excision for a second time, the tumor has not shown any metastasis. Therefore, a differential diagnosis is very important to distinguish it from the other malignant tumors, such as squamous cell carcinoma (SCC), basal cell carcinoma, nodular melanoma, or keratoacanthoma. These malignant tumors are more aggressive than trichilemmal carcinoma; they have a slower growth rate but a very distinct histology. In addition, a trichilemmal carcinoma is usually a solitary lesion, occurring separately from the proliferating trichilemmal tumor (PTT).[1]

Morgan et al[2] reported the results of immunohistochemical tests of 12 spindle cell SCC samples. The immunohistochemical battery consisted of CK7, pancytokeratins AE1/AE3, HCK (34 beta E12), and low-molecular weight keratins (CAM 5.2). The results were as follows: 34 beta E12 (12/12 samples, 100%), p63 (10/12, 80%), AE1/AE3 (8/12, 67%), and CAM 5.2 (7/12, 58%). The authors concluded that 34 beta E12 was the most promising cytokeratin for distinguishing spindle cell squamous carcinoma from the histological mimickers, such as, AFX (atypical fibroxanthoma), spindle cell/desmoplastic melanoma, scar, and cutaneous leiomyosarcoma. Du et al[3] reported a case of extramammary Paget disease mimicking acantholytic SCC in situ. The immunohistochemical staining showed a carcinoembryonic antigen (CEA), CK7, and cCK8 to be strongly expressed in the nests and singly arranged large tumor cells, and the surrounding epidermis was positive for CK5/6 and negative for CEA. In our case, we differentiated trichilemmal carcinoma from SCC by hematoxylin and eosin and PAS staining pattern.

Table 1. Antibodies used for immunohistochemistry during the study.

Antibody	Source and catalog number	Dilution
AE1/AE3	DAKO, M3515	1:200
CAM 5.2	Becton Dickinson, 349205	1:2
Cytokeratin 7	DAKO, M7018	1:100
HCK	DAKO, 34 beta E12	1:150

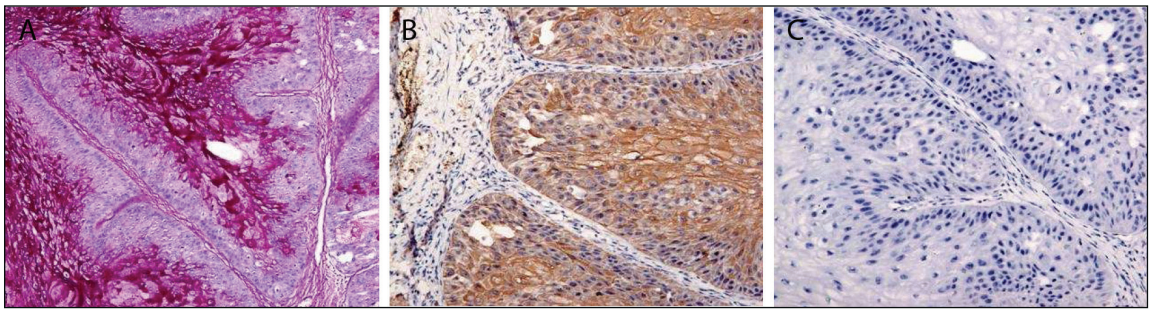

Figure 3. Chemical and immunohistochemical staining of the tumor. A. PAS positive staining; B. AE1/3 positive staining; C. CAM5.2 negative staining. Paraffin sections were prepared and subjected to chemical (for PAS) or immunohistochemical analyses. The antibodies used are listed in Table 1.

Immunohistochemistry revealed that AE1/3 and HCK (**Table 1**) were positive, but CAM5.[2] and CK7 (**Table 1**) were negative, strongly indicating trichilemmal carcinoma.

Malignant proliferating trichilemmal tumor (MPTT) is considered to be a malignant transformation of a PTT. Morphologically, MPTT has more extensive hyperkeratosis and a greater number of cysts than malignant trichilemmoma.[4] The incidence of recurrence or metastasis of MPTT is considered to be similar to that of SCC on burn scars. In terms of its general clinical features, MPTT mainly occurs on the scalp or face in elderly men. MPTT may be accompanied by ulceration, papillate formation, agglutination of slough, or rapid enlargement and is sometimes difficult to differentiate from SCC.[5] The MPTT histology is characterized by the presence of clear cells with atypical nuclei and trichilemmal keratinization of tumor cell nests,[4] and tumor cells are commonly PAS positive. There have been several reports of regional and distant metastases,[6] and fatal cases have also been recorded. Amaral et al[7] reported an inguinal case that, after surgical resection of the tumor, resulted in fatal metastasis. Hodl et al[8] reported a case with invasion and penetration of the cerebral sinuses, and the diagnosis of MPTT was made by the presence of trichilemmal keratinization and clear cells with atypical nuclei. Radiation or chemotherapy is used to treat MPTT, but the effectiveness of such treatments is unknown.[5] Local excision with removal of the regional lymph nodes and follow-up is usually the best treatment course option.

Schell et al[9] reported eleven cases of trichilemmal carcinoma and found that the tumor often occurred in the slightly damaged skin of elderly people. Its clinical appearance was nonspecific, but histologically, trichilemmal carcinoma is differentiated from trichilemmoma by the occurrence of markedly increased atypical nuclei and disordered tissue architecture. Although the histology of trichilemmal carcinoma indicates a high-grade malignancy, its biological behavior appears to be relatively benign. Furthermore, Swanson et al[10] analyzed the histological and clinical findings in 10 cases of trichilemmal carcinoma and found that, in every case, the tumor occurred in the hair-bearing and sun-exposed skin, and involved the scalp, face, trunk, or upper extremities. The lesions were usually slightly raised, pale, tan, or reddish, and keratotic. The tumor size was usually 0.4 to 2.0 cm with less than 1-year history. All patients were treated by a wide local excision and, neither recurrence nor metastasis was reported after 11 to 92 months of follow-up. Histologically, the tumors were composed of a lobular proliferation centered on the pilar apparatus. Tumor cells have a predominately glycogen-rich, mucin-negative, with clear or pale eosinophilic cytoplasm. The involvement of the interfollicular epidermis was invariably noted, with a superficial ulceration in 7 tumors. Transitional zones between trichilemmal carcinoma and the adjacent epidermis were not present, although pagetoid spread occurred in 2 examples. Invasion of the reticular dermis was present in 8 cases, with infiltration to the mid-dermis in 5. All tumors exhibited areas of trichilemmal-type keratinization, and dyskeratotic cells were present in 6 patients. Hyperkeratosis and parakeratosis were variably present as well. Actinic damage was a constant feature. Despite local invasion at diagnosis, the clinical course of trichilemmal carcinoma was indolent in all cases.

Wong et al[11] reported 13 cases of trichilemmal carcinoma with similar characteristics as the cases described by Swanson et al.[10] However, several reports indicate that trichilemmal carcinoma does occur in unexposed parts of the body, accompanied by metastasis. For example, Knoeller et al[12] reported trichilemmal carcinoma at the right distal thigh, and a metastatic tumor was found in the right inguinal lymph nodes. Five years after initial diagnosis and surgery, a

metastasis to the left tibia and fibula occurred. Allee et al[13] described a case of multiple recurrent trichilemmal carcinoma with perineural invasion. The outer root sheath differentiation of the neoplasm was confirmed using antibodies against cytokeratin 15 and 17 and c-erb-B2. The trichilemmal carcinoma demonstrated abundant cytoplasmic staining of cytokeratin 17 and c-erb-B2. The aggressiveness of the tumor was evident by tumor neurotropism and failure of multiple surgical excisions. Several more cases have been reported recently,[14-18] suggesting that there may be an increased incidence of trichilemmal carcinoma.

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