

Trichilemmal Carcinoma – A Rare Tumor: Case Report

Bernardo Cacciari Peryassú¹, Raphael Cacciari Peryassú², Marcius Achiamé Peryassú^{2,3}, Juan Piñeiro Maceira⁴, Marcia Ramos-e-Silva²

¹Sector of General Surgery, Miguel Couto Hospital; ²Sector of Dermatology, HUCFF/UFRJ and School of Medicine, Federal University of Rio de Janeiro; ³Sector of Dermatology, General Polyclinic of Rio de Janeiro; ⁴Sector of Pathology, HUCFF/UFRJ and School of Medicine, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil

Corresponding author:

Prof. Marcia Ramos-e-Silva, MD, PhD
Rua Dona Mariana 143/C-32
Rio de Janeiro
22080-020 Brazil
ramos.e.silva@dermato.med.br

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SUMMARY Trichilemmal carcinoma is a rare cutaneous cancer that usually occurs on photoexposed areas in elderly individuals. Most of the time, there is a unique lesion that presents a papulonodular aspect with possible keratosis or ulceration on the top of it. We report on a case of this rare tumor, discussing various aspects of this entity and possible therapy. Simple excision with adequate safety margin is a safe, low-cost and effective mode of treatment for this type of carcinoma. Although a rare form of neoplasia, trichilemmal carcinoma has good prognosis when treated correctly.

KEY WORDS: carcinoma, skin appendage, skin aging, photoaging of skin

INTRODUCTION

Trichilemmal carcinoma is a rare cutaneous malignancy that usually occurs on photoexposed areas of aged individuals. The number of reported cases has been increasing lately. The tumor originates on the outer root sheath and is a malignant form of trichilemmoma. Most of the time, lesions present a papulonodular aspect with possible keratosis or ulceration on the top of it. Prognosis is generally good (1).

CASE REPORT

A 53-year-old Caucasian male presented with a painless, papular, skin-colored lesion and a pearl-like aspect on the upper part of the nose persisting for the last 6 months. The lesion had an approximate diameter of 0.5 cm (Fig. 1). The patient reported rapid growth of the tumor and mentioned he had sought medical assistance due to the cosmetically awkward appearance of the lesion. Dermatoscopic examination revealed a few telangi-



Figure 1. Skin-colored, painless, papular lesion with a few telangiectasias and a pearl-like aspect on the nose, of 0.5 cm in diameter.

ectasias on its surface, thus basal cell carcinoma was suspected. Total excision was performed with a safety margin of 0.5 cm, through ellipse excision.

On histopathology, a circumscribed, although infiltrative epithelial neoplasm comprising squamous cell trabecules with wide and clear cytoplasm was observed at lower magnification (Fig. 2). At higher magnification, the infiltrative character and trichilemmal keratinization pattern of cell groups were evident (Fig. 3). In view of such histopathologic findings, the diagnosis of trichilemmal carcinoma was confirmed.

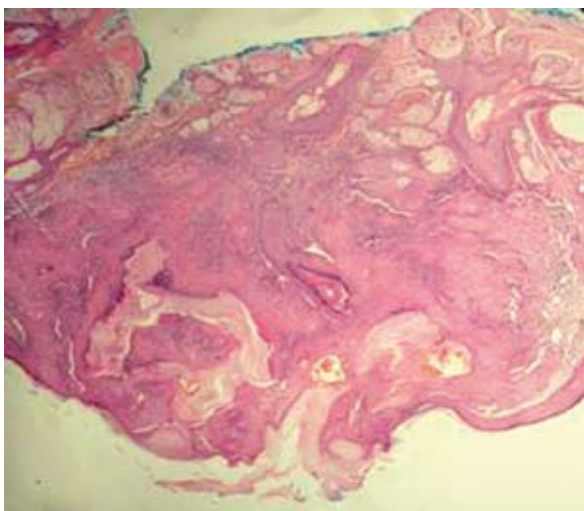


Figure 2. A circumscribed, although infiltrative epithelial neoplasm comprising squamous cell trabecules with wide and clear cytoplasm; hematoxylin-eosin stain; original magnification X40.

DISCUSSION

Trichilemmal carcinoma is a rare cutaneous malignancy, considered as a malignant variety of trichilemmoma. The term was proposed in 1976 by Headington, when observing, according to his description, a tumor composed of bright cells with cytoplasmic atypia of adnexal keratinocytes that were adjacent to the epidermis and follicular epithelium (2).

A higher incidence of this tumor has been observed on photoexposed areas of elderly individuals with fair skin. In published articles, age varies from 9 to 95 years, the youngest one being a xeroderma pigmentosum patient (3). It usually occurs as a single lesion, skin-colored, painless, nodular or pedunculated, and it may be ulcerated or keratotic on its top (4). There are no published reports worldwide of multiple lesions in a single patient. Rarely occurring on the limbs, the most affected areas are scalp, face and neck. There are descriptions of trichilemmal carcinoma arising from the pelvic region (5), on burn scars (6) and actinic keratosis (7). Men are more frequently affected than women, with a slight difference in proportion of 1.6:1 (1).

The exact pathogenesis of the lesions is not yet known. Their distribution suggests that solar radiation plays an important role in its genesis. Nevertheless, since trichilemmal carcinoma is an adnexal tumor, its most common location on the head and neck regions may be explained by the higher concentration of skin appendages in these areas (8). Trichilemmal carcinoma has been observed in elderly patients submitted to 50-60 chest x-rays for tuberculosis control (9). Takata *et al.* suggest,

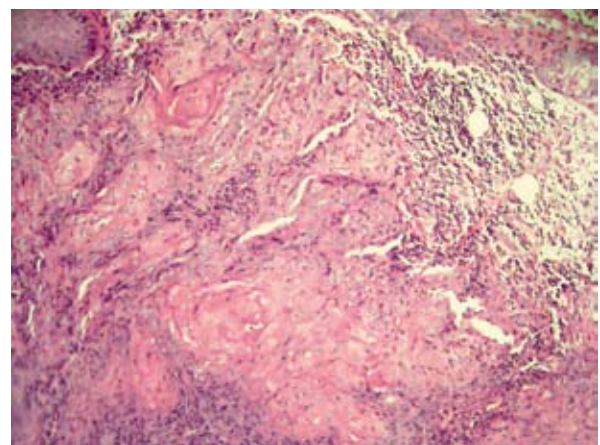


Figure 3. A detail showing the infiltrative character and trichilemmal keratinization pattern of cell groups; hematoxylin-eosin stain; original magnification X400.

in a case of malignant transformation of a trichilemmal cyst, that p53 deletion was responsible for the onset of the carcinomatous lesion (10).

Histologically, the tumor presents characteristics of its own. Optical microscopy features epithelial lobular proliferation centered in a pilosebaceous unit. The lobules contain big, polygonal and glycogen-rich bright cells, with eosinophilic cytoplasm, strong PAS positivity and mucin negativity. Trichilemmal keratinization is characterized by a decreased thickness or lack of the granular layer, abrupt keratinization of some of the cells, and, possibly, the formation of a dense, non-lamellar keratin. Inflammatory infiltrate with numerous lymphocytes and plasma cells around the lobules, in addition to hemorrhage and necrosis in major lesions, have also been observed. Differently from its benign variety, trichilemmoma, trichilemmal carcinoma exhibits cellular atypia with high mitotic rates, which is an indication of malignancy (8).

Despite its aggressive histologic aspect, the prognosis is good. Only a few recurrences, which had narrow surgical margins, have been reported. Lymph node metastases are rare with only three cases reported, all of them with primary lesions located in the pelvic region. Usually, simple excision with an adequate safety margin is enough as treatment. There is a case report of recurrent lesions with perineural invasion successfully treated with Mohs micrographic surgery. There was also a patient treated with 5% topical imiquimod cream (11).

On immunohistochemistry, there is positivity for keratin 17, which is an intermediary filament present in the outer sheath of hair follicles but not in the interfollicular epidermis. Positivity for *c-erbB-2* oncogenic receptors was demonstrated, and it was also present in a series of other cancers as squamous cell carcinomas and Paget's disease. This specific marker indicates the aggressive character of the lesion (1).

On differential diagnosis for trichilemmal carcinoma, we consider basal cell carcinoma, keratoacanthoma, squamous cell carcinoma, nodular malignant melanoma, epidermal cyst, and proliferative trichilemmal cysts/tumors (6).

Our patient responded well to simple excision of the lesion with a 0.5-cm safety margin, and did not present any signs of recurrence one year after the procedure.

In conclusion, trichilemmal carcinoma is a rare, cutaneous tumor of low-grade malignancy and simple excision with adequate safety margin is a safe, low-cost and efficient treatment.

References

1. Allee JE, Cotsarelis G, Solky B, Cook JL. Multiply recurrent trichilemmal carcinoma with perineural invasion and cytokeratin 17 positivity. *Dermatol Surg* 2003;29:886-9.
2. Headington JT. Tumors of the hair follicle. A review. *Am J Pathol* 1976;85:479-514.
3. Reis JP, Tellechea O, Cunha MF, Baptista AP. Trichilemmal carcinoma: review of 8 cases. *J Cutan Pathol* 1993;20:44-9.
4. Song MG, Min HG, Jung SY, Yang JM, Lee ES. Trichilemmal carcinoma with a cutaneous horn. *Br J Dermatol* 2000;143:646-7.
5. Dekio S, Funaki M, Jidoi J, Mihara M. Trichilemmal carcinoma on the thigh: report of a case. *J Dermatol* 1994;21:494-6.
6. Ko T, Tada H, Hatoko M, Muramatsu T, Shirai T. Trichilemmal carcinoma developing in a burn scar: a report of two cases. *J Dermatol* 1996;23:463-8.
7. Misago N, Tanaka T, Kohda H. Trichilemmal carcinoma occurring in a lesion of solar keratosis. *J Dermatol* 1993;20:358-64.
8. Laochumroonvorapong P, Kokta V, Quan MB. Trichilemmal carcinoma in an African American. *Dermatol Surg* 2002;28:284-6.
9. Chan KO, Lim IJ, Baladas HG, Tan WT. Multiple tumour presentation of trichilemmal carcinoma. *Br J Plast Surg* 1999;52:665-7.
10. Takata M, Rehman I, Rees JL. A trichilemmal carcinoma arising from a proliferating trichilemmal cyst: the loss of the wild-type p53 is a critical event in malignant transformation. *Hum Pathol* 1998;29:193-5.
11. Jo JH, Ko HC, Jang HS, Kim MB, Oh CK, Kwon KS. Infiltrative trichilemmal carcinoma treated with 5% imiquimod cream. *Dermatol Surg* 2005;31:973-6.