Review Article

Trichilemmal carcinoma, an uncommon tumor: update on its management and prognosis

Carcinoma triquilemal, um tumor incomum: atualização sobre manejo e prognóstico

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■ **ABSTRACT**

Introduction: Trichilemmal carcinoma (TC) is a rare neoplasm of skin appendages. It was first described in 1968 as tricoleptocarcinoma, and has an incidence of 0.05% in patients subjected to histopathological examination after excision of cutaneous lesions. TC has an indolent clinical course ; however, reports in the literature put in doubt this indolent behavior . **Objectives:** To provide an update on the management and prognosis of TC. **Methods:** A search of the PubMed and SciELO databases by using with the MeSH terms "trichilemmal carcinoma", "tricholemmal carcinoma", "adnexal skin tumor", and "carcinoma triquilemal" was performed. **Results:** Owing to the rarity of TC, most studies were case reports, which essentially corroborate the indolent nature of the disease. Surgical excision is the recommended treatment, and is associated with a low risk of recurrence and low morbidity and mortality. **Conclusion:** The behavior of the lesion and the procedure of treatment of TC are based on isolated cases or in a small series of cases. Because of its low prevalence, a multicenter collaboration of a greater number of cases can help define the best treatment recommendations, pathophysiology, and prognosis. Surgical excision remains the gold standard of treatment, and is associated with a low risk of recurrence.

Keywords: Scalp; Oncology; Skin neoplasms; Carcinoma of skin appendages; Neoplasms of adnexal and skin appendages; Prognosis.

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■ **RESUMO**

Introdução: O carcinoma triquilemal (CT) é uma neoplasia rara dos anexos cutâneos. Foi descrito pela primeira vez em 1968, como tricoleptocarcinoma, e tem incidência de 0,05% em pacientes submetidos a exame histopatológico após excisão de lesões cutâneas. Parece ser um tumor de baixa agressividade, porém, relatos na literatura colocam em dúvida tal comportamento indolente. **Objetivos:** Oferecer uma atualização sobre manejo e prognóstico do CT. **Métodos:** Pesquisa no PubMed e SciELO com os termos MeSH "*trichilemmal carcinoma*", "*tricholemmal carcinoma*", "*adnexal skin tumor*", "carcinoma triquilemal". **Resultados:** Devido a sua raridade, a maior parte da literatura se baseia em relatos de caso, os quais na sua maioria corroboram a natureza indolente da doença. Excisão cirúrgica é o tratamento preconizado, com baixo risco de recidiva e baixa morbimortalidade. **Conclusão:** O comportamento da lesão e as condutas no tratamento do CT baseiam-se em casos isolados ou de pequenas séries de casos. Devido a sua baixa prevalência, uma colaboração multicêntrica agrupando um maior número de casos pode ajudar a definir melhor recomendações de tratamento, fisiopatologia e prognóstico. Excisão cirúrgica continua a ser o padrão-ouro de tratamento, com baixo risco de recidiva.

Descritores: Couro cabeludo; Oncologia; Neoplasias cutâneas; Carcinoma de apêndice cutâneo; Neoplasias de anexos e de apêndices cutâneos; Prognóstico.

INTRODUCTION

Trichilemmal carcinoma (TC) is a rare neoplasm of the skin appendages, originating from the external root sheath of hair follicles. TC occurs primarily in sun-exposed skin areas such as the scalp, forehead, neck, torso and upper limbs, usually among elderly individuals, predominantly in women[1].

It was first described in 1968 as tricoleptocarcinoma, and occurred in 0.05% of patients that were subjected to histopathological examination of excised cutaneous lesions[2]. Headington was the first to propose the term TC in 1976 to describe a "histologically invasive, cytologically atypical clear cell neoplasm of adnexal keratinocytes which is in continuity with the epidermis and/or follicular epithelium"[3].

It seems to be an indolent tumor; however, reports in literature put in doubt this indolent behavior.

OBJECTIVE

The objective of this report was to provide an update on the management and prognosis of patients with TC through a literature review.

METHODS

A search was performed in the PubMed and SciELO online databases bhy using the MeSH terms "trichilemmal carcinoma", "tricholemmal carcinoma", "adnexal skin tumor", and "carcinoma triquilemal". Articles in Portuguese and English were assessed for eligibility.

DISCUSSION

TC occurs predominantly in the scalp, and shows a higher incidence in women and people of Caucasian ethnicity, generally in those between the fourth and sixth decade of life[4,5]. Skin areas with greater sun exposure, burns, or those subjected to previous irradiation have higher susceptibility to TC[6]. The lesions are usually smaller than 2 cm; however, some reported lesions reach up to 25 cm in diameter[7] (Figure 1).

A recent study has suggested that the description of TC in the literature as a well-characterized cutaneous neoplasia is exaggerated. This exaggeration may be due because the existence of other conditions that mimic TC are underestimated, including Bowen's disease[8].

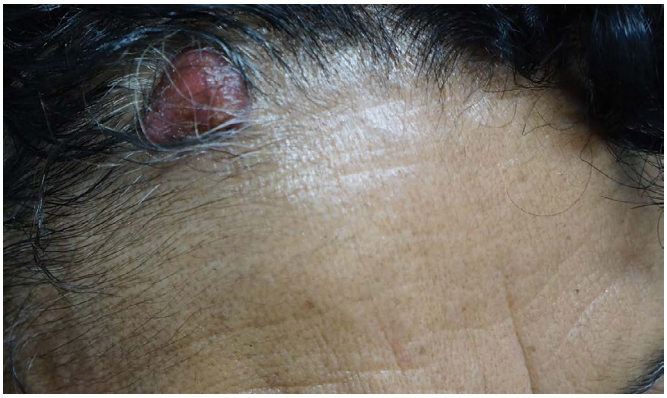

Figure 1. Clinical aspect of the lesion in the scalp of a 54-year-old woman, with one-year progression. Biopsy revealing a trichilemmal carcinoma.

TC usually occurs as a solitary lesion[9], predominantly in the head and neck regions[10]. Clinically, it appears approximately one year before diagnosis with a rapid growth rate[11].

Differential diagnosis includes benign tricholemmal cyst, squamous cell carcinoma (Figures 2 and 3), basal cell carcinoma, keratoacanthoma, verrucous cysts, proliferative trichilemmal cysts, and pseudocarcinomatous cysts[5,12].

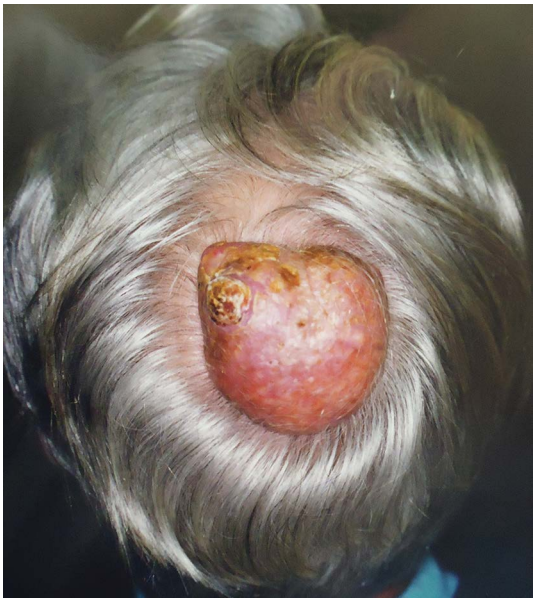

Figure 2. Friable, ulcerated lesion, of about 3 years of progression, in the scalp of a 71-year-old male patient. Histopathological analysis showing malignant proliferative trichilemmal tumor with squamous degeneration area (ulcerated area between 10 and 11 hours).

The malignant portion of trichilemmoma seems to originate from the outer layer of the hair follicle[13]. Histologically, TC seems to be an intermediate- to high-grade neoplasia, and represents a lobular proliferation centered on the pilosebaceous unit and composed of clear and pale cells with atypia,

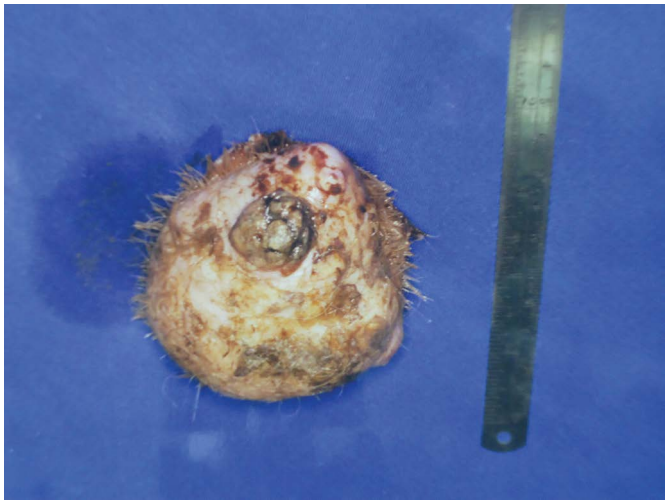

Figure 3. The surgical resection specimen of the lesion of Figure 2 (10 cm ruler for comparison).

containing glycogen with basilar invasion or total thickness of the interfollicular epidermis[14]. The growth of the tumor is lobular and infiltrative, with the lobules frequently centered and expanding the pilosebaceous unit. The initial intraepithelial impairment may extend to the reticular dermis. Actinic damage has been reported as a common feature[11]; nevertheless, there are reports of tumors in areas without sun exposure. No criterion for universal histopathological diagnosis has been established for TC[15].

The prognosis depends on the size of the tumor and the location, the degree of differentiation, and the histological subtype. In an analysis of 615 patients with squamous cell carcinomas (with behavior similar to TC), the key prognostic factors for metastasis were increase in tumor depth (> 2 mm), immunosuppression, lesion in the ear, and increased superficial diameter (> 6 mm)[16].

Despite its aggressive cytological appearance, TC represents an indolent tumor that usually evolves with a benign course and is associated with good clinical prognosis[3] and nonmetastatic potential[16,17].

However, there are several studies in literature reporting TCs with an aggressive course, with local lymph node invasion[9,18,19], recurrence[19], and even metastasis[20,21], and thus, the actual behavior of the lesion is questionable. In a recent series of 26 patients, survival at 5 years was 89.5%[5]. There is no standardized chemotherapy treatment for TC, but in reports of patients with recurrent tumors or metastases, regimes including cisplatin, cyclophosphamide, and adriamycin (similar to the one used for patients with advanced cases of squamous cell carcinoma) have shown control of tumor growth; however, cure was not attained[5,19].

Similar to information on the course of the lesion, there are discrepancies in the literature regarding the treatment of TC. Complete surgical excision with histological documentation of free margins is recommended as the standard treatment by some authors[4], which can be obtained using Mohs micrographic surgery[19]. Others, however, recommend a wide surgical excision[18].

Adjuvant radiotherapy is requested for high-risk cases, when complete resection is impossible, or in cases of recurrence or metastasis[22].

CONCLUSION

We can conclude that most of the information on the behavior and approach of the treatment of TC are based on isolated cases or small series of cases. Owing to the low prevalence of these tumors, larger and prospective studies are required to delineate the actual behavior of this lesion, since the behavior differs substantially from the indolent behavior, as described by a large number of authors.

Surgical excision with 1 cm margin is safe and simple, and is associated with a low risk of recurrence. It is important to discuss and warn the patient about the possibility of recurrence and, in cases of recurrent or aggressive tumors, multidisciplinary follow-up, including close liaison with the oncologist for adjuvant treatment, is essential.

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