Trichilemmal carcinoma in an Iranian patient: reporting a rare skin tumor

Ebrahim Esmati, MD [1]
Mojtaba Vand Rajabpour, MD [1,2]
Mohammad Shirkhoda, MD [2]
Vahid Soleimani, MD [3]
Maryam Garousi, MD [4*]

1. *Radiation Oncology Research Center (RORC), Cancer Institute of Iran, Tehran University of Medical Sciences, Tehran, Iran*
2. *Cancer Research Center, Cancer Institute of Iran, Tehran University of Medical Sciences, Tehran, Iran*
3. *Pathology Department, Cancer Institute of Iran, Tehran University of Medical Sciences, Tehran, Iran*
4. *Radiation Oncology Department, Iran University of Medical Sciences, Tehran, Iran*

**Corresponding author:*
Maryam Garousi, MD
Radiation Oncology Department,
Iran University of Medical Sciences,
Tehran, Iran
Postal code: 354-14665
Email: Garousi.m@iums.ac.ir

Received: 18 August 2020
Accepted: 12 November 2020

Trichilemmal carcinoma is a rare tumor of the outer sheath cells of hair follicles. These tumors occur on the sun-exposed skin of the elderly in the majority of cases. The gross tumor demonstrates exophytic, polypoid, ulcerative, or nodular lesions with or without bleeding. Histopathologically, it is characterized by an anastomosing trabecular, diffuse, or lobular growth pattern with clear keratinization. A critical differential diagnosis is squamous cell carcinoma (SCC), where the prognosis of trichilemmal carcinoma is far better than SCC. Surgery is a significant and effective treatment for trichilemmal carcinoma cases, and recurrence after complete surgical resection with a negative margin is uncommon. Trichilemmal carcinoma is considered a low-grade tumor with low metastatic potential. This paper reports a 48-year-old Iranian male with trichilemmal carcinoma of the scalp treated with surgery, having no recurrence after three years of follow-up. The role of other treatment modalities like radiation, local therapy, and systemic therapy are also discussed.

Keywords: carcinoma, skin, tumor, hair follicle, surgery

Iran J Dermatol 2021; 24: 147-149

DOI: 10.22034/ijd.2020.244451.1196

INTRODUCTION

Trichilemmal carcinoma is a rare tumor accounting for about 0.005% of all skin tumors [1]. This tumor is a neoplasm of the outer sheath cells of hair follicles [2]. Similar to squamous cell carcinoma (SCC), these tumors occur on the sun-exposed skin of the elderly's face in the majority of cases. Nevertheless, they can also emerge on other sites of the body [3]. In some cases, it is difficult to differentiate these two types of tumors through hematoxylin and eosin staining, while discrimination has an essential role in the prognosis and treatment. The prognosis of trichilemmal carcinoma is far better than that of SCC, and the risk of metastasis is rare [4].

Notably, ulceration may occur in the lesion, and the dimensions of the tumor are typically less than 10×10 cm [1]. The treatment of choice is surgery, and recurrence after a negative margin is not usual [5]. Herein, we report a case of trichilemmal carcinoma of the scalp resected after suspicion of lipoma, and the patient was followed for three years.

CASE PRESENTATION

A healthy 48-year-old Iranian male presented with an eight-month history of a 1.5-cm, firm, indurated lesion on the scalp without ulceration, pain, itching, or color change. The patient was visited by a general practitioner and surgeon, who suspected the lesion as a lipoma. Due to the patient's discomfort, the lesion was resected.

Microscopic examination of the lesion showed a squamoproliferative malignant neoplasm in continuity with the epidermis, composed of round lobules of atypical keratinocytes with eosinophilic to clear cytoplasm as well as palisading of peripheral cells in lobules, foci of keratinization, and prominent surrounding connective tissue (Figure 1). The tumor size was 1× 0.7 cm, and the surgical margins were negative. No lymphovascular or perineural invasion was identified. The patient was under follow-up for the first two years with MRI. At three years post-surgery, no recurrence or metastasis has occurred to date.

DISCUSSION

Most trichilemmal carcinomas present as long-standing, indolent lesions with recent rapid growth; in fact, a trichilemmal carcinoma is the malignant form of trichilemmoma and trichilemmal keratosis, which are benign proliferations of outer root sheath cells [3]. Sun-exposed skin regions including the scalp are the most common sites for trichilemmal carcinoma. However, other sites, such as the nasal dorsum and chest wall, have also been reported [1]. The macroscopic view demonstrates exophytic, polypoid, ulcerative, or nodular lesions with or without bleeding [6]. Histopathologically, it is characterized by an anastomosing trabecular, diffuse, or lobular growth pattern with clear keratinization [1]. A critical differential diagnosis is SCC, where the prognosis of trichilemmal carcinoma is far better than SCC and the risk of metastasis is rare. There is some evidence suggesting that UEA-I staining is strongly positive in trichilemmal carcinoma [4]. Other clinical differential diagnoses include basal cell carcinoma, nodular melanoma, and keratoacanthoma [5].

The treatment of choice is resection with a 1-cm safe margin. Lymph node metastasis and margin status are the prognostic factors for the outcome [7]. Mohs micrographic surgery (MMS) has been successfully used in non-melanotic skin cancers. A center reviewed seven head and neck trichilemmal carcinoma patients treated with MMS. After an average of 2.5 years follow-up, no recurrence or metastasis was detected, so MMS

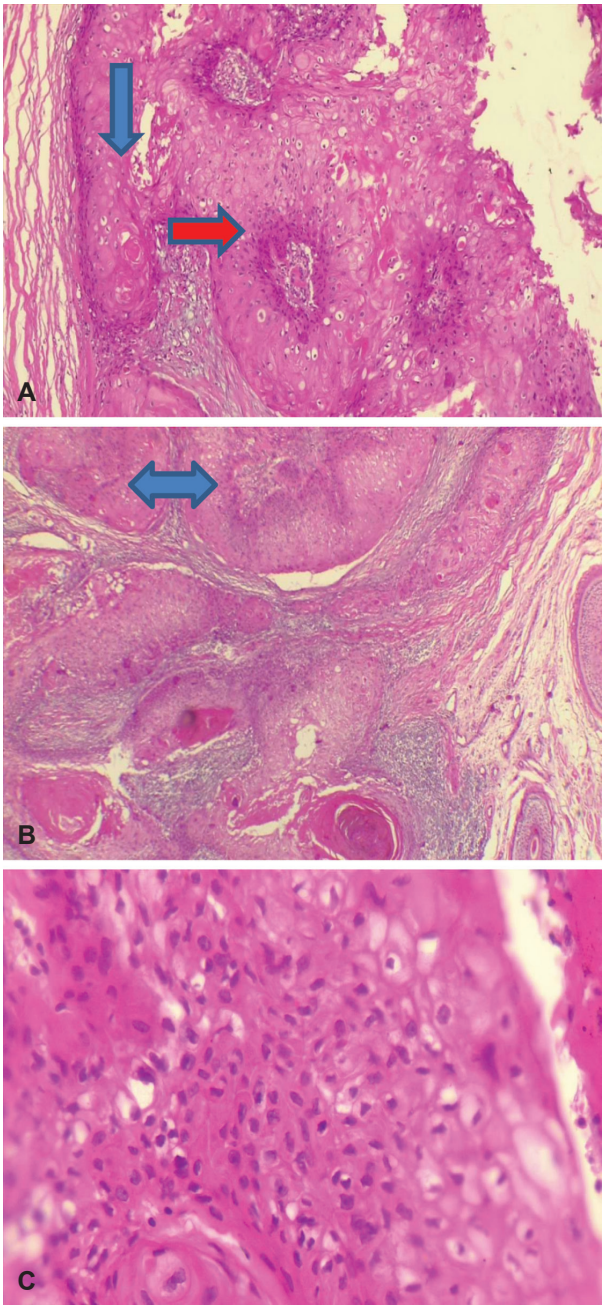

Figure 1. Pathologic findings in favor of trichilemmal carcinoma: A) keratinization foci (blue arrow) and atypical keratinocytes (red arrow) (H&E staining, ×10 magnification); B) lobules of atypical keratinocytes (left-right arrow); C) high-power view of atypical keratinocytes (H&E staining, ×40 magnification)

could be considered for the head and neck region where tissue preservation is required [7].

Recurrence after complete surgical resection is uncommon. On the other hand, Kulahci *et al.* reported multiple recurrences of trichilemmal carcinoma in the scalp of a young man who was ultimately treated by surgery [8]. The role of radiation treatment is not well-defined. Duncan *et al.* reported a 93-year-old man with a large temple trichilemmal carcinoma lesion who could not undergo surgery because of comorbidities and was treated with radiotherapy. He received 4500 cGy in 15 fractions with 9 MeV electron energy. A complete response was observed after five weeks [7].

Elsewhere, imiquimod 5% was used for eight weeks as treatment for an elderly patient with infiltrative trichilemmal carcinoma, and a complete histologic resolution was observed. After 16 months of follow-up, no recurrence was detected [9]. Hence, topical agents may be an alternative option in cases where surgery cannot be performed.

Trichilemmal carcinoma is considered to have low metastatic potential, though cases of distant metastasis have been reported in the literature. There is no standard treatment for metastatic disease. Cisplatin and cyclophosphamide have been reported to be effective with partial responses [10].

Our case was generally well-aligned with the findings described in the literature, and no recurrence of disease was detected after three years of follow-up. Thus, complete surgical resection seems to be sufficient.

CONCLUSION

Due to the rarity of trichilemmal carcinoma, evidence concerning its treatment is limited. According to our case, surgery seems to be an effective and sufficient option.

Conflict of interest: None declared.

REFERENCES

1. Evrenos MK, Kerem H, Temiz P, et al. Malignant tumor of outer root sheath epithelium, trichilemmal carcinoma: Clinical presentations, treatments and outcomes. Saudi Med J. 2018;39(2):213-216.
2. Headington J. Tumors of the hair follicle. A review. Am J Pathol. 1976;85(2):479-514.
3. Reis J, Tellechea O, Cunha M, et al. Trichilemmal carcinoma: review of 8 cases. J Cutan Pathol. 1993;20(1):44-9.
4. Ko T, Muramatsu T, Shirai T. Distribution of lectin UEA-I in trichilemmal carcinoma, squamous cell carcinoma, and other epithelial tumors of the skin. J Dermatol. 1996;23(6):389-93.
5. Xu DB, Wang T, Liao Z. Surgical treatment of trichilemmal carcinoma. World J Oncol. 2018;9(5-6):141.
6. Zhuang SM, Zhang GH, Chen WK, et al. Survival study and clinicopathological evaluation of trichilemmal carcinoma. Mol Clin Oncol. 2013;1(3):499-502.
7. Tolkachjov SN, Hocker TL, Camilleri MJ, et al. Mohs micrographic surgery in the treatment of trichilemmal carcinoma: the Mayo Clinic experience. J Am Acad Dermatol. 2015;72(1):195-6.
8. Kulahci Y, Oksuz S, Kucukodaci Z, et al. Multiple recurrence of trichilemmal carcinoma of the scalp in a young adult. Dermatol Surg. 2010;36(4):551-4.
9. Jo JH, Ko HC, Jang HS, et al. Infiltrative trichilemmal carcinoma treated with 5% imiquimod cream. Dermatol Surg. 2005;31(8):973-6.
10. Yi HS, Sym SJ, Park J, et al. Recurrent and metastatic trichilemmal carcinoma of the skin over the thigh: a case report. Cancer Res Treat. 2010;42(3):176.