CASE REPORT

INDONESIAN JOURNAL OF CANCER

Trichilemmal Carcinoma on the Head (Surgical Challenge to Achieve Free Margin): A Case Report

Daan Khambri

Department of Surgery, Oncology Division, Dr. M. Djamil Hospital – Medical Faculty, Universitas Andalas, Padang, Indonesia

ARTICLE INFO

Received : 14 June 2021
Reviewed : 21 June 2021
Accepted : 30 August 2021

Keywords:
head, sufficient free margin, trichilemmal carcinoma

Corresponding author:
Daan Khambri
Department of Surgery of Dr. M. Djamil Hospital/Medical Faculty of Universitas Andalas, Padang, West Sumatra, Indonesia
daan_khambri@yahoo.com

ABSTRACT

Introduction: Trichilemmal carcinoma is a rare tumor derived from the outer hair sheath. It usually appears on the face, ears, neck, scalp, and sun-exposed areas. It generally occurs in patients older than 50 years old, and there is no sex predilection to this disease.

Case Presentation: A 60-year-old woman came with a complaint of a lump in the head that often bled. The size of the lump was approximately like a bean seed and broke when she combed her hair. The lump was getting bigger within a year and easier to bleed. She often felt pain. Based on the history of surgery 2 months ago, the examination implies that the patient had trichilemmal carcinoma.

Conclusions: Trichilemmal carcinoma generally occurs in the elderly category. Although trichilemmal carcinoma has a benign clinical course and local recurrence cases are uncommon, sufficient free margin and immediate reconstruction are challenging if the predilection is in the head and neck. Surgical excision is the recommended treatment for this disease.

INTRODUCTION

Trichilemmal carcinoma is a malignant adnexal tumor that develops from the outer hair sheath, usually occurring in sun-exposed areas [1,3]. It is conventionally considered as a neoplasm derived from adnexal keratinocytes with glycogenotic clear cells and evidence of differentiation of outer root sheaths or trichilemmal and is a malignant partner of trichilemmoma [2,4,5]. Trichilemmal carcinoma was a term first introduced by Headington in 1976, but pathologists adopted it a few years later. This rare skin tumor can appear nodular or plaque-like, whitish, or hyperkeratotic with reddish and smooth surfaces, or ulcerated, and it is typically smaller than 3 cm in diameter. High incidence occurs in the elderly category with no gender dominance [1]. Trichilemmal carcinoma is considered to have low metastatic potential, meaning that it is a low-level carcinoma that rarely attacks other parts of the body [6]. This case is interesting to discuss because it is rare, and the number of cases is small.

CASE PRESENTATION

We report the case of a 60-year-old female patient who came in with a complaint of a lump in the head that often bled. At first, the lump felt like acne at the size of a bean seed. When the patient combed her hair, the lump was broken. It had been getting bigger since last year, measuring 9 x 8 cm. The lump was painful and bled easily. There are no lumps in the neck area and no complaints of breath. There is no full feeling and nausea or vomiting, no history of radiation exposure, and no history of continuous exposure to sunlight for a long time. There is no family history of having suffered from this disease before. Based on the history of surgery 2 months ago in the tumor area, it was found that the patient suffered from trichilemmal carcinoma. The actions performed for this patient are wide excision and full-thickness skin graft.

The patient was in the prone position under general anesthesia. We started the operation by making the surgical landmark approximately 3 cm outside from the induration area of the tumor. We prepared the patient with povidone-iodine 10% from all the head areas until the clavicle. We started making the incision on the landmark from the skin through connective tissue aponeurosis loose areolar until the surface of the periosteum. We performed wide excision from all the tissue inside the surgical landmark. There was no involvement of the tumor in the periosteum. The defect was approximately 12 x 12 cm. We closed the defect with full-thickness skin grafts taken from the periumbilical skin. The operation was conducted within 2 hours. We did the wound redressing 3 days after the skin graft had been 100% viable. The patient was discharged 3

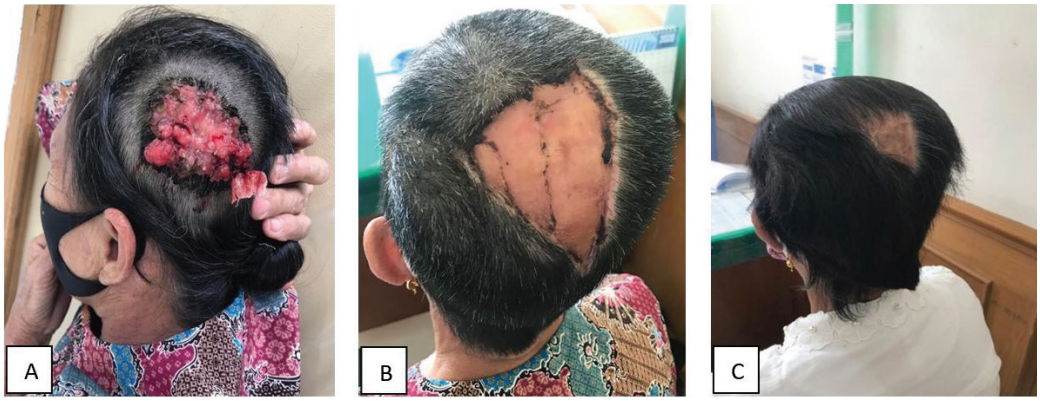

Figure 1. Clinical photos of patient. (A) Lession before surgical procedure; (B) The patient's after final condition after surgical; (C) The patient's after 3 months surgical procedure.

days upon admission. The procedure was well tolerated by the patient, and no adverse events were noted (**Figure 1.B**). Three months after the procedure, the patient had no recurrences, complaints, nor other complications (**Figure 1.C**).

DISCUSSION

Trichilemmal carcinoma is rare skin cancer and a malignant form of trichilemmoma. Trichilemmal carcinoma is a malignant adnexal tumor derived from the outer hair sheath that usually occurs in the elderly. It generally occurs in patients older than 50 years old, and there is no gender predisposition to this case [1,3,7]. Trichilemmal carcinoma usually appears on the face, ears, neck, scalp, and sun-exposed areas [1,8].

Based on the research by Hamman et al. [9], there were 103 cases of trichilemmal carcinoma with a median age of 72.5 years. The location of trichilemmal carcinoma on the face is on the cheeks (18%), forehead (10%), nose (9%), temples (6%), eyelids (4%), scalp (13%), ears (9%), and neck (9%). According to the figure above, the tumor is on the patient's scalp, and she is 60 years old. Tumors most often attack the elderly.

The pathogenesis of trichilemmal carcinoma is still unclear. Many risk factors are still under investigation, like high-dose irradiation therapy in previous lesions and repeated X-rays for diagnostic purposes associated with this type of carcinoma. In addition, with exposure to the sun, burn scars can affect the development of trichilemmal carcinoma [10–12].

Clinically, tumors can look like pale tan or reddish papules, ulceration or exophytic plaque or nodule, presenting keratosis or scabs. In general, it appears as a solitary lesion measuring 3 cm in a greater diameter [1,10]. The lesion usually appears less than 1 year and indicates a rapid growth phase, so medical attention is required to deal with it because trichilemmal carcinoma is rare, and its presentation is similar to other skin tumors [13]. The uncommon trichilemmal carcinoma lacks clinical features at inspection and usually appears in the ulcerative grayish to reddish-brown or flesh colors. Clinically, it is often misdiagnosed as basal cell carcinoma (BCC), actinic keratosis, or squamous cell carcinoma (SCC). There is no known association between trichilemmal carcinoma and Cowden syndrome, both of which appear as de novo proliferation and do not share common pathogenesis [1,14]. Therefore, the differential diagnosis is considered in this case.

Histopathologically, trichilemmal carcinoma has histological features like trichilemmoma with cellular atypia. Its growth is lobular, infiltrative, and often centers on pilosebaceous units. Tumor cells in the center are large, polygonal, and clear with periodic acid Schiff positive, sensitive diastase, and cytoplasm rich in glycogen. Its periphery cells are often palisaded, and the spread of adenoids can sometimes be seen. In some examples when hair follicles are found within the tumor, the outer root sheath is in continuity with the proliferative lobules. In contrast to trichilemmoma, trichilemmal carcinoma cells have atypical nuclei and high mitosis index. Intratumoral hemorrhage and necrosis can also be observed, especially at large masses. Immunohistochemical tests were carried out to confirm the diagnosis; the test showed negative results of CEA (carcinoembryonic antigen) and EMA (epithelial membrane antigen) although there were reports of late positive cases [3,7,9,13,15].

Trichilemmal carcinoma should be distinguished by other malignant skin lesions so that the selection in treatment is appropriate. The recommended treatment for trichilemmal carcinoma is surgical excision, wide excision with histologically documented clean margins; however, there may be a potential recurrence and local aggressiveness. Mohs micrographic surgery is useful for tracking pagetoid spread [2,7]. Surgery is considered to be the treatment of choice for trichilemmal carcinoma, and periodic surveillance without adjuvant therapy is generally sufficient [16]. Trichilemmal carcinoma has an indolent clinical course [7]. Based on the author's assumption, this is caused by the patient's hesitancy to see MDs in low middle-income countries, the patient's

lack of knowledge about trichilemmal carcinoma, and the patient's fear of surgery. If the patient is diagnosed with malignant carcinoma, they are afraid to have adjuvant therapy with chemotherapy. Also, in terms of cost, they need health insurance to pay for the treatment. Trichilemmal carcinoma generally has a good prognosis and reports of deep invasion, and the local recurrence case is uncommon [7,13,15]. Xu et al. [17] reported that a total of 26 cases of trichilemmal carcinoma were treated with surgical excision, and tumor recurrence occurred in only 2 cases. Zuang et al. [16] also found the same thing; of 26 patients who suffered from trichilemmal carcinoma studied, only 2 patients had a recurrence. If the distant tumor metastasis is confirmed, systemic chemotherapy should be considered. It can control or monitor the development of the disease [17]. In our case, the actions performed for this patient are wide excision and full-thickness skin grafts. After the surgery, the procedure was well tolerated by the patient, and no events were noted. A full-thickness skin graft was chosen because it offers several advantages, results in minimal contracture, and shows better mechanical and aesthetic outcomes [18]. Three months after the procedure, the patient came back with no recurrence, complication, or other complaints.

CONCLUSIONS

Trichilemmal carcinoma is a rare adnexal tumor. Surgical excision is a recommended treatment for the disease; however, there may be a potential recurrence and local aggressiveness. Trichilemmal carcinoma has an indolent clinical course and uncommon local recurrence. If the distant tumor metastasis is confirmed, systemic chemotherapy should be considered as it can control or observe the development of the disease.

DECLARATIONS

Competing of Interest

The authors declare no competing interest in this study.

Acknowledgment

Not applicable

REFERENCES

1. Barnhill RL, Crowson AN, Magro CM, Piepkorn MW. Dermatopathology. 3 ed. Europe: McGraw-Hill Education 2010.
2. Billingsley EM, Davidowski TA, Maloney ME. Trichilemmal carcinoma. J Am Acad Dermatol. 1997;36(1):107–9.
3. Headington JT. Tumors of the hair follicle. A review. Am J Pathol. 1976;85(2):479–514.
4. Dalton SR, LeBoit PE. Squamous cell carcinoma with clear cells: how often is there evidence of tricholemmal differentiation? Am J Dermatopathol. 2008;30(4):333–9.
5. Garrett AB, Azmi FH, Ogburia KS. Trichilemmal carcinoma: a rare cutaneous malignancy: a report of two cases. Dermatol Surg. 2004;30(1):113–5.
6. Dailey JR, Helm KF, Goldberg SH. Tricholemmal carcinoma of the eyelid. Am J Ophthalmol. 1993;115(1):118–9.
7. Reis JP, Tellechea O, Cunha MF, Baptista AP. Trichilemmal carcinoma: review of 8 cases. J Cutan Pathol. 1993;20(1):44–9.
8. Nemetz MA, da Cunha RM, Reeck P, et al. Trichilemmal carcinoma: a case study. Rev Bras Otorrinolaringol. 2004;70:832–5.
9. Hamman MS, Brian Jiang SI. Management of trichilemmal carcinoma: an update and comprehensive review of the literature. Dermatol Surg. 2014;40(7):711–7.
10. Chan KO, Lim IJ, Baladas HG, Tan WT. Multiple tumour presentation of trichilemmal carcinoma. Br J Plast Surg. 1999;52(8):665-7.
11. Ko T, Tada H, Hatoko M, et al. Trichilemmal carcinoma developing in a burn scar: a report of two cases. J Dermatol. 1996;23(7):463–8.
12. Misago N, Tanaka T, Kohda H. Trichilemmal carcinoma occurring in a lesion of solar keratosis. J Dermatol. 1993;20(6):358–64.
13. Swanson PE, Marrogi AJ, Williams DJ, et al. Tricholemmal carcinoma: clinicopathologic study of 10 cases. J Cutan Pathol. 1992;19(2):100–9.
14. Allee JE, Cotsarelis G, Solky B, Cook JL. Multiply recurrent trichilemmal carcinoma with perineural invasion and cytokeratin 17 positivity. Dermatol Surg. 2003;29(8):886-9.
15. Boscaino A, Terracciano LM, Donofrio V, et al. Tricholemmal carcinoma: a study of seven cases. J Cutan Pathol. 1992;19(2):94–9.
16. Zhuang SM, Zhang GH, Chen WK, et al. Survival study and clinicopathological evaluation of trichilemmal carcinoma. Mol Clin Oncol. 2013;1(3):499–502.
17. Xu B, Wang T, Liao Z. Surgical Treatment of Trichilemmal Carcinoma. World J Oncol. 2018;9(5-6):141–4.
18. Kim S, Chung SW, Cha IH. Full thickness skin grafts from the groin: donor site morbidity and graft survival rate from 50 cases. J Korean Assoc Oral Maxillofac Surg. 2013;39(1):21–6.