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Case Report

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A Rare Case of a Trichilemmal Carcinoma, Clinical Aspect, Management and Outcome

Asmae Akhana, Reda Fadel*, Dounia Kamal, Mohamed Noureddine Elamine Elalami

Service d'ORL et Chirurgie Maxillofaciale, CHU HASSAN II, FEZ, MOROCCO

***Corresponding author:** Reda Fadel, Service d'ORL et Chirurgie Maxillofaciale, CHU HASSAN II, FEZ, MOROCCO, E-mail: reda.fadel@usmba.ac.ma

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Abstract

The trichilemmal carcinoma is a rare hair follicle tumor that is thought to occur from a malignant transformation of a benign trichilemmoma. We report the case of a 69-year-old female patient with no specific history. Who seeks medical care for a gradually increasing in size scalp lesion evolving for more than two years. The patient underwent resection of the tumor with bilateral lymph node dissection. After repair by skin graft, the patient was referred for adjuvant radiotherapy. The 18-month follow-up did not show any recurrence. The diagnosis of proliferating trichilemmal cyst is suggested after a thorough examination and a precise dermatological examination of the suspected lesion. The latter would be confirmed by a histological examination. Despite respecting a correct margin of excision, the trichilemmal carcinoma has a considerable recurrence tendency.

Keywords: Trichilemmal, Carcinoma, Malignant, Cyst

Introduction

First described by WILSON JONES in 1966, the trichilemmal carcinoma is a rare hair follicle tumor that is thought to occur from a malignant transformation of a benign trichilemmoma. The tumor is believed to develop from the outer root sheath of the hair follicle. It is most often found on areas that are exposed to the sun. We report the case of 45 year old woman presenting this rare entity.

Case Report

We report the case of a 69-year-old female patient with no specific history. Who seeks medical care for a gradually increasing in size scalp lesion evolving for more than two years. On examination, we note the presence of an infected ten centimeters ulcerative mass of the vertex, bleeding upon contact with several permeation nodules, the largest of which is 5 cm. At the cervical level, we palpated several spinal lymph nodes. A biopsy was performed and was in favor of a trichelemmal carcinoma. A body CT scan revealed the presence of suspicious-looking cervical lymphadenopathies, without adjacent bone involvement or distant metastases.

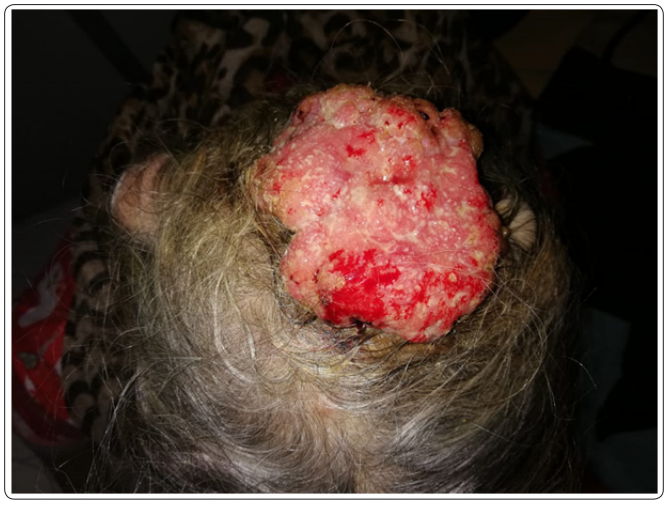

Figure 1: Preoperative Clinical Aspect of the Scalp Trichilemmal Carcinoma

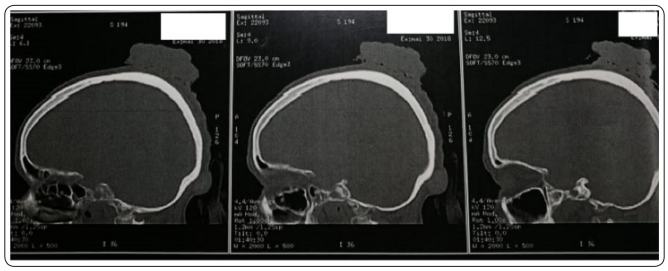

Figure 2: A sagittal CT scan showing the trichelemmal carcinoma with no bone involvement.

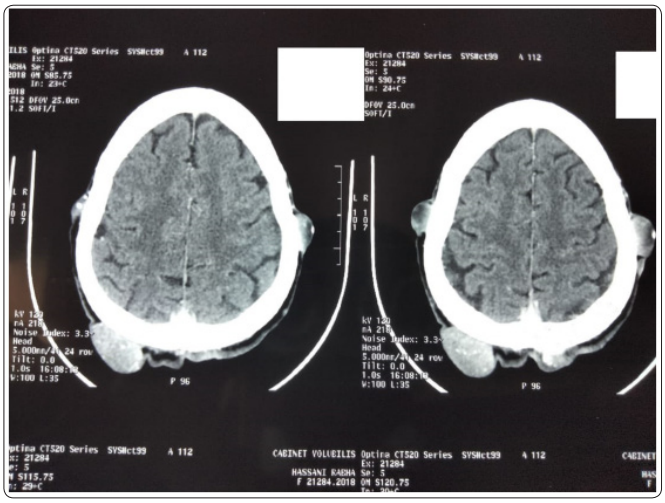

Figure 3: Axial contrast-enhanced CT scan showing the other benign trichelemmal cysts.

The patient underwent resection of the tumor with bilateral lymph node dissection. Histological study confirmed the biopsy, the margins were healthy, and the dissection negative. After repair by skin graft, the patient was referred for adjuvant radiotherapy. The 18-month follow-up did not show any recurrence and then the patient was lost to follow-up.

Discussion

The proliferating trichilemmal cyst remains rare, with a marked incidence in elderly women. Malignant forms are rare and only very few cases have been reported in the literature involving the vital prognosis [1].

Comparison of the results of the most significant surveys, suggests that the trichelemmal carcinoma is an increasingly frequent reason for consultation, especially in North American and European countries, this could be linked to the white racial predominance in these countries. In this context Sau et al, reported a rate of over 90% Caucasian in their series, while the other studies did not specify, which would highlight the need for more studies targeting the darker skinned population [2].

The diagnosis of proliferating trichilemmal cyst is suggested after a thorough examination and a precise dermatological examination of the suspected lesion. The latter would be confirmed by a histological examination which will specify on the one hand the cyst's type, and on the other hand would differentiate between benignity and malignancy; moreover it will help rule out other differential diagnoses. The contribution of immunohistochemistry in trichelemmal carcinoma is essential and thus resolves the problem of malignancy or benignity. According to Rangel and Gamboa, Ki67 is directly linked to the likelihood of the cyst proliferating. A mutation in the p53 gene would also cause carcinomatous proliferation. Other markers should be looked for and make it possible to retain the diagnosis, such as CK10, CK16, CD34 and the expression of involucrine.The management of a proliferating trichilemmal cyst is generally straightforward, based on excisional surgery with sufficient safety margins. We can sometimes, in malignant forms, resort to chemo-radiotherapy effective in addition to care [3-7].

Despite respecting a correct margin of excision, the trichilemmal carcinoma has a considerable recurrence tendency as expressed by a meta-analysis reporting a local recurrence rate of 3.7% and a remote location rate of 1.2% [8].

Conclusion

Its rarity, its aggressiveness in malignant forms, and its accessibility to prevention, encourage the screening and diagnosis of these lesions at an early stages in order to improve its prognosis and limit its aggressiveness in malignant forms.

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