Trichilemmal Carcinoma of the Scalp: A Clinical and Histopathological Diagnostic Challenge.

Abstract

Trichilemmal carcinoma is a rare malignant cutaneous adnexal tumor whose clinical presentation may mimic squamous cell carcinoma. We report the case of an 83-year-old woman presenting with a nodular and exophytic lesion of the midline parietal scalp that had progressively evolved over nine months. The clinical appearance was highly suggestive of a malignant skin tumor. An initial biopsy suggested a malignant proliferating trichilemmal tumor. Complete oncologic excision was performed with intraoperative assessment of the deep plane and immediate reconstruction using a rotation flap. Definitive histopathological examination confirmed trichilemmal carcinoma with clear surgical margins. The postoperative course was favorable, with no recurrence at one-year follow-up. This case highlights the clinical diagnostic difficulty associated with this rare tumor and the importance of histopathological examination in establishing the definitive diagnosis.

Keywords: trichilemmal carcinoma; scalp; adnexal tumor; histopathology; surgery.

1. Introduction

Trichilemmal carcinoma (TC) is a rare malignant cutaneous adnexal tumor characterized by trichilemmal differentiation, corresponding to differentiation toward the outer root sheath of the hair follicle. Initially described as a distinct entity among cutaneous carcinomas, it occurs predominantly in older individuals and is mainly located in the head and neck, particularly the scalp. Its clinical presentation is variable and often nonspecific, which can complicate clinical diagnosis. [1–4]

Clinically, TC may present as a nodule, an exophytic lesion, or an ulcerated lesion and may be mistaken for squamous cell carcinoma or other cutaneous tumors. Diagnosis relies on histopathological examination, which demonstrates trichilemmal differentiation associated with malignant features such as cytological atypia, mitotic activity, and tumor invasion. [2–4]

Because of its rarity, available data regarding its biological behavior and management are based mainly on limited series and individual case reports. Complete surgical excision with histological assessment of the margins is the most frequently reported treatment. [5]

We report a case of scalp TC in an elderly patient whose clinical appearance mimicked squamous cell carcinoma and whose definitive diagnosis was established by histopathological examination of the excised specimen.

2. Case Presentation

An 83-year-old woman was treated in Agadir for a scalp tumor. She had no known medical or surgical history and was not receiving any chronic medication. There was no history of tobacco or alcohol use.

The lesion had appeared approximately nine months before presentation and had progressively increased in size. The patient reported transient pain and pruritus. There was no contact bleeding and no frank ulceration of the tumor.

Clinical examination revealed a nodular, exophytic, polymorphous, and multilobulated lesion in the midline parietal scalp, with focal crusting. It measured approximately 2.5 cm in greatest diameter. The lesion was fixed to the deep planes and did not bleed on contact. The overlying skin was pale pink,

whereas the surrounding skin appeared healthy. No cervical lymphadenopathy was palpable. The clinical appearance was highly suggestive of squamous cell carcinoma.

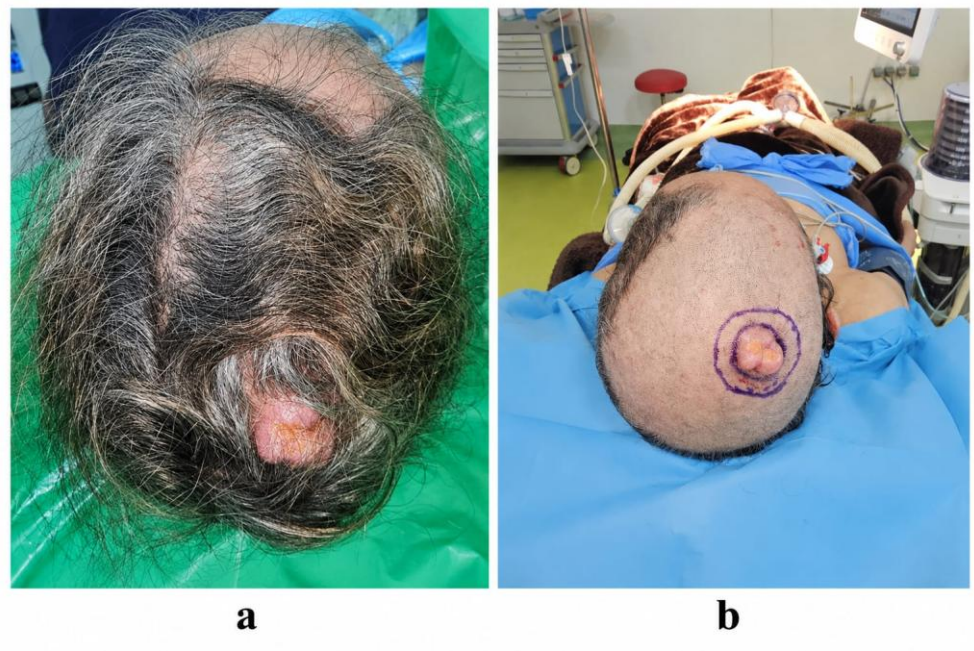
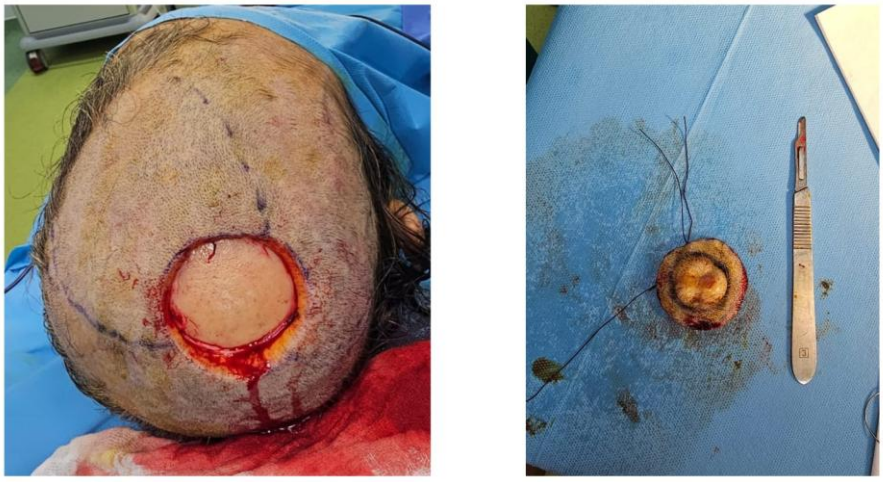


Figure 1. a: Preoperative clinical appearance: exophytic, polymorphous, multilobulated lesion of the midline parietal scalp. b: Preoperative view with 10-mm excision margins.

An outpatient biopsy performed before surgical management showed a cystic-appearing epithelial proliferation with invaginations and trichilemmal keratinization. The epithelial cells displayed moderate to marked cytonuclear atypia and abnormal mitotic figures. Invasive foci were present as lobules and cords with an epidermoid appearance, together with tumor necrosis. The diagnosis favored a malignant proliferating trichilemmal tumor.

Brain CT demonstrated an approximately 30-mm tissue proliferation involving the full thickness of the scalp, without involvement of the outer table of the skull. A thoraco-abdomino-pelvic CT scan performed for staging showed no definite secondary lesions. Pulmonary abnormalities were identified and led to additional phthisiological assessment, which concluded that the patient had pulmonary tuberculosis.



a b

Figure 2. Main surgical steps: a: oncologic excision with exposure of the outer table; b: excised specimen.

Based on the histological diagnosis and clinical appearance, oncologic excision was performed with a 10-mm clinical margin. The excision was completed in a single stage. Intraoperative frozen-section examination of the periosteum was performed and confirmed a clear deep margin, with no tumor infiltration. The resulting defect was immediately reconstructed using a helical rotation flap.

a b

Figure 3. Scalp reconstruction: a: elevation of the helical rotation flap; b: final result.

Definitive histopathological examination of the skin specimen, measuring 5 × 5 × 1.5 cm, identified a nodular lesion measuring 3.2 × 2.6 × 1.3 cm. Microscopic examination showed a malignant epithelial proliferation composed of cellular trabeculae and cords, with focal areas of whorled architecture. The cells

were large, with eosinophilic and focally clear cytoplasm, markedly nucleolated atypical nuclei, and mitotic figures. No neoplastic vascular emboli or perineural invasion were identified. Focal trichilemmal differentiation was present. The lateral surgical margins were free of tumor, with the closest margin located 7 mm from the tumor proliferation. The deep plane was also free of tumor, with a 2-mm margin. The final histopathological diagnosis was trichilemmal carcinoma measuring 3 cm in greatest diameter.

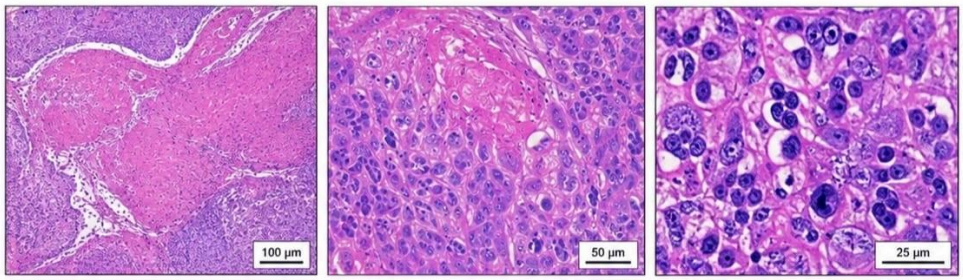


Figure 4a Prolifération tumorale faite de cordons et de lobules de cellules épithéliales (HE, ×100).

Figure 4b Cellules tumorales à cytoplasme éosinophile parfois clarifié, avec kératinisation trichilemmale (HE, ×200).

Figure 4c Atypies cytonucléaires marquées et figures de mitoses atypiques (HE, ×400).

Figure 4. Histopathological features: 4a, malignant epithelial proliferation composed of cellular trabeculae and cords (H&E ×100); 4b, focal trichilemmal differentiation (H&E ×200); 4c, marked cytonuclear atypia and atypical mitotic figures (H&E ×400).

The postoperative course was uncomplicated. The drain was removed after 48 hours and the sutures were removed on postoperative day 14. No postoperative complications occurred. No adjuvant treatment was administered. Local healing was favorable, with good wound healing and a satisfactory morphological outcome for the patient. At one-year clinical follow-up, there was no evidence of local recurrence, cervical lymphadenopathy, or metastatic disease.

3. Discussion

Trichilemmal carcinoma is a rare adnexal tumor defined by trichilemmal differentiation. Published data show a predilection for older individuals and a preferential location in the head and neck. A recent systematic analysis of 231 cases identified the cervicofacial region as the main site and highlighted the marked variability of its clinical presentation. [2]

In our case, the patient was 83 years old and the tumor was located on the scalp. The clinical appearance was that of an exophytic, nodular, and multilobulated lesion with progressive growth and fixation to the deep planes. This presentation could readily suggest squamous cell carcinoma. Several published cases have likewise described scalp TC initially considered clinically to be squamous cell carcinoma. [3,4]

Histopathological examination is therefore the decisive element in diagnosis. In our case, the biopsy had already identified a trichilemmal-type proliferation with cytonuclear atypia, abnormal mitotic figures, invasive foci, and tumor necrosis. Examination of the excision specimen confirmed the diagnosis of trichilemmal carcinoma and allowed more precise assessment of the tumor architecture and surgical margins.

Trichilemmal differentiation may be manifested by trichilemmal keratinization, lobular or trabecular architecture, and characteristic cytological features. The distinction from squamous cell carcinoma, particularly some clear-cell variants, may remain difficult on small biopsies. Immunohistochemical markers such as cytokeratin 7, EMA, CD34, or other differentiation markers may be considered in

diagnostically challenging situations, but their value varies among series and should be interpreted in conjunction with morphology. [6,7]

Malignant proliferating trichilemmal tumor is also an important differential diagnosis, particularly on the scalp. Although these entities share some histological features and may exhibit substantial growth, they should not be regarded as strictly synonymous. In our case, examination of the excision specimen ultimately established the diagnosis of trichilemmal carcinoma. [8]

Treatment is based primarily on complete surgical excision with histological assessment of the margins. Available evidence remains limited because of the rarity of the disease, but published series favor complete excision. Mohs micrographic surgery may also be considered in selected locations or when tissue preservation and precise margin control are particularly important. [5]

In our case, a 10-mm clinical margin was used, with intraoperative assessment of the periosteum. Definitive examination confirmed that the deep plane was free of tumor, with a 2-mm margin, and that the lateral margins were clear, the closest being 7 mm. Immediate reconstruction with a helical rotation flap provided satisfactory coverage of the defect and a favorable morphological result.

The prognosis of trichilemmal carcinoma is generally considered relatively favorable, although local recurrences and, more rarely, metastases have been reported. In available series, aggressive events appear to be mainly associated with locally advanced tumors or histological features associated with poorer prognosis. [2,5]

In our case, the absence of neoplastic vascular emboli and perineural invasion, complete excision with clear margins, and the absence of recurrence at one-year follow-up are favorable features. Nevertheless, the limited one-year follow-up does not allow definitive conclusions regarding the long-term risk of recurrence.

This observation has several limitations, including its isolated nature and the absence of an immunohistochemical study in the available documentation. Nevertheless, it is of interest because of the clinical diagnostic difficulty, the scalp location, and the correlation between the clinical, histopathological, and surgical findings.

4. Conclusion

Trichilemmal carcinoma of the scalp is a rare tumor whose clinical presentation may closely mimic squamous cell carcinoma. Definitive diagnosis relies on histopathological examination, which identifies trichilemmal differentiation together with malignant features. Complete oncologic excision with margin control constitutes the cornerstone of management. This case highlights the importance of considering this entity in the differential diagnosis of nodular scalp tumors in older individuals.

Declarations

Consent for publication: Written informed consent for publication of the case and clinical photographs should be confirmed in accordance with institutional and journal requirements before submission.

Conflicts of interest: None declared.

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