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Trichilemal Carcinoma: Report of Two Cases of a Rare Entity

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ABSTRACT

Trichilemmal carcinoma is a rare malignant neoplasm arising from the outer root sheath of the hair follicle, in the fourth and ninth decades of life, in sun-exposed skin. This tumor pressents as plaques, papules, or nodules, with or without ulceration, grow rapidly, and are locally aggressive.

Two cases of long-standing tumors are presented, one previously diagnosed as clear cell hidradenoma and the second with a history of multiple resections with continuous recurrence.

Histologically, tumors display polygonal clear cells (PAS positive, diastase sensitive), solid/lobular/trabecular infiltration, peripheral nuclear palisading, trichilemmal keratinization, high mitotic index and ulcerations. The cells are CK17, CD34and KRT15 positiv. The Immunohistochemistry is an important tool for differentiating it from other simulators.

Keywords

Trichilemmal carcinoma, Malignant trichilemmoma, Outer root sheath tumor, Hair follicle neoplasm, Cutaneous adnexal tumor, Clear cell neoplasm.

Introduction

Trichilemmal carcinoma, first described by Headington in 1976, is a malignant neoplasm arising from the outer root sheath of the hair follicle [1,2]. It is more common between the fourth and ninth decades of life and typically occurs in sun-exposed areas such as the face, scalp, and neck [3].

Clinically it may present as plaques, papules, or nodules, with or without ulceration; lesions are usually solitary, rarely multiple, grow rapidly, and are locally aggressive [4]. It can mimic other cutaneous tumors, including squamous cell carcinoma, basal cell carcinoma, keratoacanthoma, and melanoma [3].

Definitive diagnosis is made by histopathology, which shows a lobular, solid, or trabecular pattern composed of atypical polygonal cells with clear cytoplasm, peripheral nuclear palisading, and a high mitotic index [2,5,6]. Foci of trichilemmal keratinization without a granular layer—resembling the outer root sheath at the isthmus—may be observed [6].

Clinical Cases

Case 1: A 57-year-old woman, rural worker, with diabetes and hypertension, presented to the dermatology clinic at Hospital

General "Dr. Manuel Gea González" with a 4-year history of a progressive, pruritic scalp lesion. Examination revealed an exophytic, cup-shaped mass measuring 4 × 3 cm with well-defined borders, ulcerated surface and central necrosis. Initial incisional biopsy reported clear cell hidradenoma. Complete excision was performed for definitive classification.

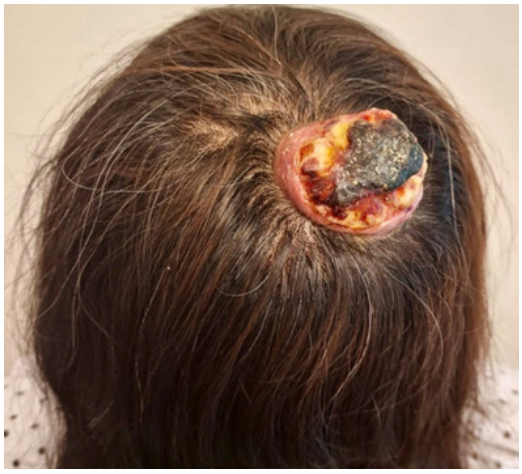

Figure 1: Clinical image of case 1.

Case 2: A 63-year-old man, hairstylist, presented with an exophytic lesion of 3.2 × 3.5 cm with well-defined borders and an ulcerated surface. The lesion had been excised twice previously and recurred with rapid growth. An excisional biopsy with negative surgical margins was performed.

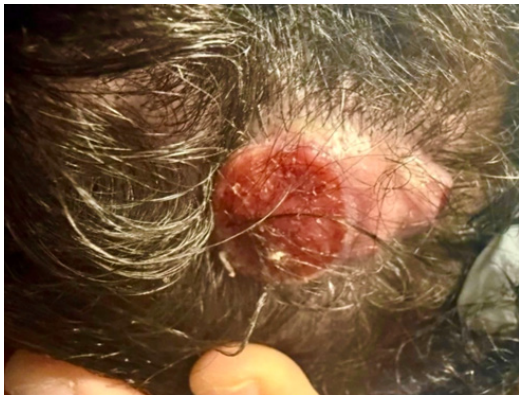

Figure 2: Clinical image of case 2.

Histopathological Findings

Both specimens showed an ulcerated malignant neoplasm contiguous with the epidermis and infiltrating in a lobular pattern into the deep reticular dermis. Tumor composed of epithelial cells with pleomorphic nuclei, open chromatin and clear cytoplasm, peripheral nuclear palisading, and numerous atypical mitoses (up to 25 mitoses per 10 high-power fields). Peripheral foci of trichilemmal keratinization and desmoplastic stroma were present.

Immunohistochemistry

Case 1: Membranous positivity for cytokeratin 17 (CK17), supporting origin from the outer root sheath.

Case 2: CD34 positivity, supporting follicular lineage versus other differentials.

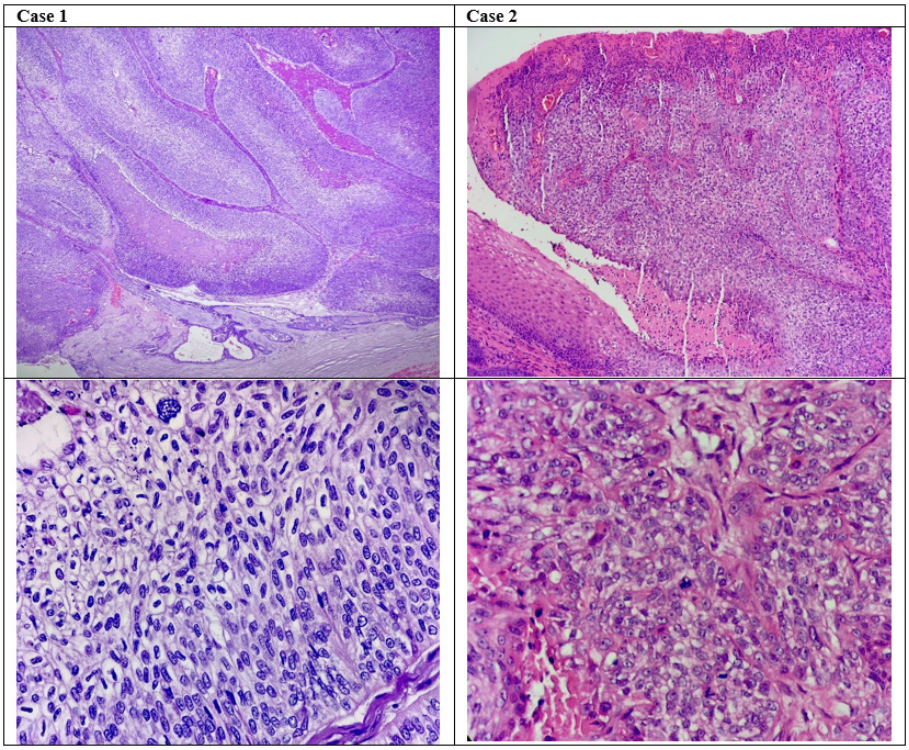

Figure 3-6: HYE photomicrographs: neoplasm with infiltrative pattern, cells with clear cytoplasm and peripheral palisade.

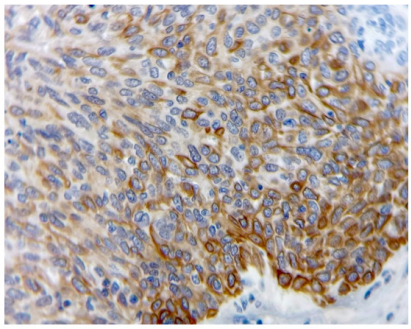

Figure 7: CK17 positivity

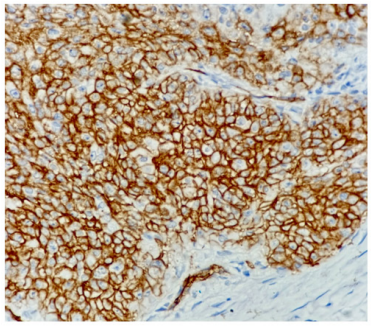

Figure 8: CD34 positivity.

Discussion

Proliferating trichilemmal carcinoma is a rare adnexal malignancy (~1% of adnexal carcinomas) originating from the outer root sheath of hair follicles [5]. It primarily affects sun-exposed head and neck skin in patients aged 50–90 years. Risk factors include ultraviolet radiation, solid-organ transplantation and immunosuppression, trauma, burns, arsenic exposure, bullous pemphigoid, and genetic conditions such as xeroderma pigmentosum and Cowden síndrome [4,5].

Pathogenesis is incompletely understood; molecular alterations described include abnormalities involving p53, loss of wild-type p53, PTEN alterations (associated and unassociated with Cowden), TOP1 amplification (linked to HPV), and some gene fusions [6-8].

Clinical and Pathological Features

Lesions are often <3 cm but can reach up to 20 cm; one lesion in our series measured 4 cm [2,4]. Macroscopic appearances include crateriform lesions, smooth translucent surfaces, telangiectasia, or verrucous changes [1]. Histologically, tumors display polygonal clear cells (PAS positive, diastase sensitive), solid/lobular/trabecular infiltration, peripheral nuclear palisading, trichilemmal keratinization, high mitotic index, and sometimes necrosis, hemorrhage or ulceration [1,4,6].

Differential diagnosis includes clear cell squamous cell carcinoma, basaloid tumors with clear cell change, sebaceous carcinoma, porocarcinoma, clear cell hidradenocarcinoma, balloon cell melanoma, and metastatic clear cell renal carcinoma. Some feautures like peripheral nuclear palisade, infiltration pattern, and trichilemmal keratinization support the diagnosis of trichilemmal carcinoma. Immunohistochemistry is often required for accurate distinction [1,6-10].

The Immunoprofile of Trichilemmal carcinoma commonly expresses CK17, a intermediate filament expressed in the outer root sheath of the follicles; CD34 and KRT15 are also useful [4,9-11]. CEA and EMA are typically negative (helps differentiate from hidradenocarcinoma and sebaceous carcinoma). Ber-EP4 negativity aids in distinguishing from basal cell carcinoma; SOX10 negativity helps exclude balloon cell melanoma [6-10]. UEA-1 lectin staining can assist in differentiating from squamous, basal cell or porocarcinomas [10].

Treatment

First-line treatment is surgical excision; recommended margins vary from 1–4 mm, with some advocating >1 cm to prevent recurrence or metastasis [12]. Mohs micrographic surgery is preferred in cosmetically sensitive areas (face, scalp) [13]. When surgery is not an option, alternatives include topical 5% imiquimod [14], excision with frozen sections, or local radiotherapy [15]. For metastatic disease—rare—platinum-based chemotherapy has been used; immune checkpoint inhibitors are emerging as options in refractory cases [16,17].

Prognosis

Although often indolent, trichilemmal carcinoma can be locally aggressive or metastatic, with rare intracranial involvement reported [18]. Reported 5-year overall survival is 89.2%. Prognostic factors include nodal metastasis, perineural invasion and surgical margins [12]. In a series of 212 surgically treated cases, local recurrence occurred in 16 (7.55%). Clinical follow-up every 6–12 months is recommended [10].

Conclusion

Proliferating trichilemmal carcinoma is a rare tumor that poses diagnostic challenges. Wide biopsy or complete excision is essential for definitive diagnosis, and immunohistochemistry is frequently necessary for accurate classification.

Differential Diagnoses

Neoplasia	Histological and Immunohistochemical Characteristics
Trichilemmal carcinoma	Origin: follicular Cells with clear PAS-positive cytoplasm in a lobular pattern with peripheral nuclear palisade Trichilemmal keratinization and high mitotic index. Positive for: CK17, CD34, and KRT15
Clear cell squamous cell carcinoma	Origin: epidermis Infiltrative pattern, clear cell changes with an area of keratinization. Positive: UEA-1 lectin
Clear cell hydra-denocarcinoma	Origin: eccrine Infiltrative pattern in the dermis, without connection to the epidermis Duct formation, PAS positive Positive: EMA and CEA (highlight ductal differentiation)

Sebaceous carcinoma	Origin: sebaceous glands Pattern: lobed, multivacuolated cells separated by fibrovascular stroma Connection to epidermis or follicular epithelium Positive: adipophilin (membrane), antigen receptor
Basal cell carcinoma with clear cell change	Origin: follicular-interfollicular Multiple growth patterns, peripheral nuclear palisade, and stromal retraction. Fibromyxoid stroma Positive: Ber-EP4, p63, CAM 5.2

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