Subtle Trichilemmal Carcinoma of Neck Initially Misdiagnosed and Ultimately Treated with Mohs Micrographic Surgery

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ABSTRACT

Trichilemmal carcinoma (TC) is an exceedingly rare malignant adnexal lesion, originating from hair follicle cells. Given its rarity, widely variable clinical presentation, and often non-specific appearance, TC is commonly misdiagnosed as a number of different lesions, some benign, which can lead to delays in treatment and suboptimal patient outcomes. In this report, we discuss a 65-year-old man with a neck lesion originally misdiagnosed as benign and later found to be TC all along. We also discuss several treatment options, and ultimately favor Mohs Micrographic Surgery (MMS) as the preferred treatment for TC in this case.

INTRODUCTION

Trichilemmal carcinoma (TC) is an uncommon malignant adnexal cutaneous neoplasm extending from the hair sheath that is typically present on hair-bearing, sun-exposed regions, most commonly on the scalp or face in an elderly population (mean diagnosis of 70 years old).¹ As of 2014, only 103 cases of TC had been reported, over half of which occurred on the face.² While TC rarely metastasizes, destructive features such as locally infiltrative ability, malignant histology, and high mitotic rate with rapid growth are observed.² Histologically, TC can be identified by the presence of invasive epithelial lobules composed of glycogen-rich transparent cells with peripheral palisading of basaloid cells.¹ The clinical presentation of TC is variable and non-specific, often leading to misdiagnosis and delays in treatment. Specifically, a lesion can appear as a singular papule, keratotic nodule, or a hardened erythematous plaque with either an ulcerated or smooth cover.²

Partially attributable to its rarity, there is sparse literature consensus on the ideal treatment for TC. We discuss several treatment options, and ultimately favor Mohs Micrographic Surgery (MMS) as the preferred treatment for this specific case report.

CASE REPORT

A 65-year-old Caucasian male with a history of squamous and basal cell carcinomas presented with a lesion on the left posterior neck that had persisted over the past year. On physical exam, a 1.0 x 1.1 cm firm erythematous plaque was observed with mild tenderness and no ulceration or drainage (Figure 1).

The lesion was originally confirmed histopathologically via shave biopsy to be a
benign trichilemmal cyst. Features at that time included polygonal cells with pale cytoplasm and uniform oval nuclei (Figure 2).

Several months following the shave biopsy, the lesion grew back larger at the same location and became more cyst-like and cumbersome. The area was re-biopsied 8 months later which showed a primary cutaneous malignant adnexal neoplasm of trichilemmal deviation, leading to the diagnosis of a trichilemmal carcinoma. The histology revealed significant atypia, mitosis, and necrosis at this time. The tumor extended into the dermis and was composed of expansive nodules of enlarged hyperchromatic epithelial cells, many containing clear cytoplasm with hyperchromatic and inconspicuous nuclei. In addition, there was stromal desmoplasia accompanying lymphocytic inflammatory infiltrate (Figure 3).

Immunohistochemical staining of hematoxylin and eosin showed epithelial cells to be strongly positive for p40 and negative for SOX-10. There was also sparse focal staining for epithelial membrane antigen (EMA) and CK7 while CD34 was mostly negative with some focal polygonal membranous staining, consistent with trichilemmal derivation of the tumor. Mohs micrographic surgery was completed with 5 mm margins, demonstrating clear margins with one stage and complex linear repair for closure without complication or recurrence at 6-month follow-up.
Figure 2. Original shave biopsy submitted for H&E showing polygonal cells with pale cytoplasm and uniform oval nuclei consistent with a trichilemmal cyst.

Figure 3. The re-biopsy slides submitted for H&E 8 months later revealed primary cutaneous malignant adnexal neoplasm of trichilemmal deviation with significant atypic, mitosis, and necrosis under 4x magnification.
Trichilemmal carcinoma is an extremely rare malignant lesion, originating from hair follicle cells. A comprehensive review of the literature identified 103 reported cases, with only 9% found on the neck, making this just the 8th reported case in the region. Given its rarity, variable clinical presentation, and often non-specific appearance, TC is commonly misdiagnosed as a number of different lesions. These include other malignant tumors such as basal cell carcinoma, clear cell squamous cell carcinoma, and keratoacanthoma or, more problematically, some benign tumors like pilar cyst or trichilemmoma. Even more troublesome is that the histological pattern of TC may mimic a benign trichilemmoma, thus requiring careful evaluation. In this case, the lesion was originally diagnosed as trichilemmoma with no treatment warranted, but later referred to be TC upon re-biopsy 8 months following. After review of the original biopsy slides, it was determined that the lesion was misdiagnosed originally.

TC should always be considered within the differential when plausible and histological evaluation with appropriate staining techniques can significantly aid in diagnosis. Relevant stains include markers such as CD34, EMA, p40, p53, p63 and CK. In terms of treatment for TC, surgical removal is the preferred choice. However, alternative approaches can be considered when determining the nature of the prognosis in relation to the patient and patient preferences. For instance, if the patient is subjected to pathergy, local management of intra-tumoral ethanol injections was noted to promote apoptosis of a proliferating trichilemmal tumor. In addition, infiltrative trichilemmal carcinoma subjected to imiquimod 5% cream for 8 weeks showed total resolution, though this was only reported in one patient. Recently, TC has been successfully treated with wide excision with 1-cm margins. However, MMS confers advantages over wide local excision, due to a higher cure rate, smaller defect size, low recurrence rate, and immediate closure. Although metastasis in TC is rare; in this scenario, MMS was recommended due to the carcinoma’s proximity to the cervical lymph nodes. In the event that TC does spread to the cervical lymph nodes, neck dissection and expanded resection followed by chemotherapy is recommended. Even though there is no official recommendation concerning the treatment of TC, recent literature has preferred using MMS over wide local excision on cosmetically-sensitive regions, like the head and neck. We strongly suggest using MMS to treat TC, because of its tissue-sparing technique that allows for immediate clearance of the peripheral and deep margins of the tumor with the highest cure rate.

Conflict of Interest Disclosures: None

Funding: None

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