A rare case of Trichilemmal Carcinoma: histology and management

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Introduction

Trichilemmal carcinoma (TC) is a rare, malignant, adnexal neoplasm that is derived from the outer root sheath (ORS) of the hair follicle. These tumors predominantly occur in elderly patients on sun-exposed areas, specifically on the head and neck with the face defined as the most common location. The mean age of diagnosis is 70 years old with a slight male predominance. These lesions are commonly identified as a papular, nodular, and sometimes, exophytic. They generally arise de-novo, but may also derive from an underlying proliferating trichilemmal cyst with a loss of p53, a seborrheic keratosis, a nevus sebaceus, or a scar. They can be locally aggressive and may exhibit telangiectasias and ulceration due to local destruction.

While the clinical differential diagnosis commonly includes basal cell carcinoma (BCC), squamous cell carcinoma (SCC), and keratoacanthoma, the histopathological differential also includes trichilemmoma, trichoepithelioma, clear cell SCC, clear cell porocarcinoma, and clear cell hidradenocarcinoma. The main differentiating histological feature in a TC is the evidence of trichilemmal keratinization, where the tumor exhibits an absence of granular layer between the stratum spinosum and stratum corneum. The standard of treatment for TC is wide local excision (WLE) with tumor-free margins, however, there has been increasing evidence for the efficacy of Mohs micrographic surgery (MMS) in recent years.

Case Report

Here we present a case of a 66-year-old Caucasian male who presented to the dermatology clinic for a routine full body skin exam and was diagnosed with a biopsy proven TC. The patient had a history of actinic keratoses treated with topical 5-fluorouracil (5-FU) and BCCs status-post excision and MMS. On physical exam, there was a pink to erythematous, pearly plaque with arborizing telangiectasias and fine scale located on the right anterior neck; no ulceration, discharge, or lipid deposits were noted (Figure 1). Tangential shave biopsy was performed of the lesion and histopathologic report revealed an adnexal neoplasm with trichilemmal differentiation, desmoplastic component and atypia with margins involved (Figures 2A & B). Immunohistochemical staining (IHC) was positive for CD34, and tumor cells displayed uptake of the Ki-67 proliferation marker; staining for p53 was negative (Figure 3A-B). The differential diagnoses, as described by the pathologist, included the favored diagnosis of a well-differentiated TC versus desmoplastic trichilemmoma with atypia. We discussed with the pathologist about differentiating these two entities, and it was decided that the cytologic atypia and pleomorphism outweighed only a small focus of desmoplastic change; therefore a diagnosis of TC was heavily favored. The pathologist advised for complete removal of the lesion with further evaluation. The patient deferred MMS in favor of WLE and was treated successfully with 3 mm standard margins.

Clinical and Histologic Findings

Figure 1. Pink to erythematous atrophic plaque with arborizing telangiectasias on the right anterior neck (labeled in the image as B). Figure 2. (A) Hyperkeratosis, variable parakeratosis and focal pagetoid spread in the epidermis. (B) The dermis contains a clear cell infiltrate, intracytoplasmic eosinophilic inclusions, peripheral palisading along a thickened basement membrane, surrounded by lymphocytic infiltrate (H&E, 4x). (C) Evidence of nuclear pleomorphism, abundant polygonal cells with clear cytoplasm and scattered inclusion bodies, small focus of desmoplastic change, (H&E, 10x). Figure 3. Immunohistochemical Staining (IHC) was positive for CD34 (A) and Ki-67 (B), but negative for p53 (not pictured).

Discussion

TC is a rare, adnexal tumor with evidence for follicular ORS or trichilemmal differentiation. It is considered the malignant analogue of trichilemmoma. Clinical presentation is variable; due to its ability to resemble different clinical entities, the diagnosis of TC relies on histological evaluation, accompanied by IHC. Microscopically, TC features a solid, lobular, or trabecular growth pattern often centered around a pilosebaceous unit. The tumor cells are clear, polygonal, and glycoprotein-rich (periodic acid-Schiff positive (PAS), diastase sensitive), reminiscent of clear cells of the ORS. It exhibits peripheral palisading of basoloid cells abutting a sometimes thickened hyalinized basement membrane. Upon closer inspection, cytologic atypia, nuclear pleomorphism, prominent nucleoli, and a high mitotic index can be seen. CD34 stains the external layer of the ORS without labeling the rest of the follicle, so it can be used to suggest outer sheath differentiation. Ki-67 and p53 labeling can be used to support the malignant nature of a suspected TC. Though it appears to be cytologically malignant under the microscope, TC typically demonstrates nonaggressive clinical behavior. Wide local excision with histological demonstration of clear margins has traditionally been the treatment of choice. Anciotal reports equate the risk of recurrence and lymph node metastasis to that of SCC, and therefore, TC is treated accordingly. Surgical margins generally emulate the 4-5 mm standard margins for SCC, though a few authors suggest utilizing safety margins greater than 1 cm. MMS has been increasingly employed for numerous rare adnexal tumors including TC, and was under consideration for this case as well.

Conclusion

TC is a rare adnexal tumor that grossly and microscopically mimics several entities. A biopsy is essential for diagnosis and often requires supplemental immunostaining. It should be distinguished from other tumors of follicular origin and other cutaneous malignancies. Complete excision with tumor-free margins is the typical treatment modality, though MMS has some advantages, including comprehensive visualization of the margins and a greater preservation of healthy tissue. As we struggle for uniformity in treatment recommendations, the therapeutic modality and surgical margins should be decided on an individual basis considering the patient’s preference and the lesion’s clinicopathologic features. Further characterization of the true nature and behavior of TC would contribute to a standard treatment recommendation.

References