INTRODUCTION

Trichilemmal carcinoma (TC) is a rare malignant adnexal neoplasm originating from the outer root sheath of the hair follicular epithelium characterized by an indolent clinical course, as described by Headington. This is usually encountered between the seventh and ninth decades of life in the sun-exposed areas with a predilection for face, neck, back, and extremities taking the form of plaques, papules, or solitary nodules with evidence of ulceration. In view of its low malignant potential, indolent course, and rare metastasizing potential, a wide local excision with follow-up is usually the treatment of choice.\[1,2\]

CASE REPORT

We report a case of a 60-year-old male presenting to the surgical outpatient department of our hospital with a chief complaint of nonhealing ulcer over the occipital region of the scalp for 2 months. There was no history of pain, discharge, or trauma to the localized area. Close clinical examination showed an ulceroproliferative growth of size 4 cm × 4 cm with areas of hemorrhage and necrosis over the floor [Figure 1]. There was no evidence of regional lymphadenopathy. A wide local excision was performed under general anesthesia, and the biopsy was sent for histopathological examination.

Gross examination showed an irregular, partly skin-covered soft tissue measuring 5 cm × 4.5 cm × 2 cm. The surface of the skin showed an ulceroproliferative lesion measuring 4 cm × 3.5 cm with necroinflammatory...

ABSTRACT

Trichilemmal carcinoma is a rare malignant adnexal neoplasm of follicular type usually seen over the sun-exposed areas of elderly people. We present a report of a 60-year-old male with ulceroproliferative lesion over the occipital region of the scalp for 2 months, clinically diagnosed as squamous cell carcinoma. A wide local excision was done, and histopathological examination showed atypical clear cells with abrupt keratinization. The histogenesis and other differential diagnoses are discussed.

Key words: Abrupt keratinization, adnexal neoplasm, trichilemmal carcinoma

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slough over the floor. Cut surface showed a solid, gray-white, and homogeneous area. The deep resected surgical margin appeared to be involved by the lesion. Microscopic examination showed a malignant adnexal tumor centered over the dermis composed of tumor cells arranged in nests and sheets separated by thin delicate fibrocollagen. Individual tumor cells were large and polygonal and showed vesicular nuclei with clear cytoplasm (periodic acid–Schiff [PAS] positive). Abrupt keratinization, atypical mitotic figures, and foci of necrosis were also seen [Figures 2-4]. Immunohistochemistry showed CK 1 positivity and CK 7 negativity. A histological diagnosis of TC was offered.

**DISCUSSION**

Tumors of the hair follicle were initially classified by Headington followed by Mehregan and Rosen, with currently no system being universally acceptable owing to considerable overlapping histological features. Trichilemmal tumors are a group of tumors characterized by cells differentiating toward the outer root sheath of hair follicles witnessed by a variable extent of clear change in the cytoplasm resulting from accumulation of glycogen. This entity comprises a number of benign and malignant tumors and has an association with Cowden's syndrome.[3] The concept of TC was introduced by Headington in 1976 which he described as a histologically invasive, cytologically atypical clear cell neoplasm of adnexal keratinocytes seen in continuity with the epidermis and/or follicular epithelium. He described the tumor cells as glycogen rich showing peripheral palisading with a prominent PAS-reactive, diastase-sensitive basement membrane and trichilemmal type of keratinization.[4]

The histogenesis of this neoplasm remains obscure till date, with many mechanisms being postulated. A significant number of cases arise de novo.[1] Factors such as actinic damage (sun exposure), prolonged
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radiation (more than 50–60 diagnostic chest radiographs), immunosuppression following renal transplantation, postsurgical radiation for other tumors, preexisting burn scars, and chronic mechanical stimulation play a key role.[1] Transformation from benign trichilemmoma, occurrence in slightly damaged skin of elderly people, association with arsenic intake, and xeroderma pigmentosa have been documented.[1]

Clinically, majority of the tumors are solitary, frequently seen in the elderly with no sex predilection. The tumors manifest as white to tan papules, plaques or nodules with ulceration, hyperkeratosis, or scabs usually <3 cm in the greatest dimension.[3] Head, neck, face, trunk, and extremities are commonly involved.[1] On clinical grounds, they are frequently misdiagnosed as squamous cell carcinoma, basal cell carcinoma, keratoacanthoma, or a proliferating trichilemmal tumor.[6]

For a histologic diagnosis of TC, Headington suggested six criteria:[1]

1. Continuity with a coexisting benign epithelial tumor, usually a trichilemmoma
2. Continuity with the outer sheath epithelium of a coexisting hair follicle
3. Light microscopy showing glycogen-rich epithelium, peripheral palisading, and prominent basement membrane zone
4. Trichilemmal keratinization (absent or minimal granular layer, abrupt single-cell keratinization, and formation of dense nonlamellar keratin)
5. Electron microscopic details similar to normal outer sheath epithelium or trichilemmoma
6. Immunocytochemical details similar to normal outer sheath epithelium or trichilemmoma.

On immunohistochemistry, TC shows positivity for cytokeratins, namely CK 1, 10, 14, 17, and 19. It is negative for CK 7, 8, 15, and 16. In accordance with the above-mentioned criteria, our case fulfilled criteria 3, 4, and 6.

Despite the histologic criteria by Headington, the existence of TC has been questioned by many authors. Ackerman believes that TC is an expression of basal cell carcinoma.[5] Dalton and LeBoit (2008) consider it to be a clear cell variant of squamous cell carcinoma. They described 40 cases of clear cell variant of squamous cell carcinoma, of which 38 cases demonstrated glycogen.[1] Roismann et al. reported a case of recurrent TC from a preexisting basal cell carcinoma.[6] Hence, diagnostic histopathological criteria of TC showed variation among several authors and series.

TC should be distinguished from other skin tumors with similar histomorphology because of the variation in management protocol. One of the most commonly entertained differential diagnosis is a malignant proliferating trichilemmal tumor. Grossly, these tumors are large, arising in a pilar cyst with a rapid clinical course and a favorable nonscalp region. Subtle histopathological differences such as presence of few squamous eddies are seen in malignant proliferating trichilemmal tumors. However, this finding is limited to only a few case reports. Immunohistochemically, loss of CD34 is seen in malignant proliferating trichilemmal tumor. In spite of ugly clinical appearance, increased nuclear pleomorphism, and presence of atypical mitoses, TC has a very indolent clinical course with unexceptional metastases, suggesting that cure is possible. The mainstay of treatment is surgery with wide local excision of the tumor and active follow-up. Histological clear margins must be documented because of the potential recurrences, as encountered in a study by Zhuang et al.[7] The other treatment modalities are Mohs micrographic surgery and immunomodulation with imiquimod cream. The main prognostic factors are surgical margins and lymph node status.[1] Local recurrences and metastases mandate chemotherapy and adjuvant radiotherapy.[2]

To conclude, TC till date remains a challenge to the histopathologist. Rarity, indolent clinical course, local aggressiveness, good survival rate, and occasional spontaneous regression make this a unique entity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.
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