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Case Report

Trichilemmal Carcinoma of the Upper Lip: Report of a Rare Case

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Abstract

Trichilemmal carcinoma is a rare unusual malignant tumour arising from the external root sheath of the hair follicle and predominantly involves the skin of the face in older individuals. Local recurrences and metastasis is rare, hence surgical excision without adjuvant therapy is the treatment of choice. It is imperative to consider the diagnosis of trichilemmal carcinoma along with other high grade malignant skin adnexal neoplasms for therapeutic and prognostic purposes. This article reports a rare and an uncommon case of Trichilemmal carcinoma involving the upper lip in a 55 year-old Indian male.

Key words: Skin Neoplasms;Hair Follicle;Adnexal;Skin Appendage Neoplasms;Trichilemmal Carcinoma;Root Sheath.

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Introduction

Trichilemmal carcinoma (TLC) is a rare skin tumour, classified under skin adnexal tumours which comprises of a group of benign and malignant neoplasms, exhibiting morphological differentiation towards one of the different types of adnexal epithelium present in normal skin.[1]

TLC most often affects individuals at the age of 40 years or older, at sites of sun exposure, especially the face, head and the neck, but may also affect the eyes, extremities and the trunk.[2,3] TLC has an indolent clinical course but shows high grade malignant features histologically.[3,4] It is usually found as a solitary lesion although occasional cases of multiple presentations have been reported.[5] The tumour has varied clinical appearances ranging from a flesh coloured spot to a papule, nodular or an exophytic growth that may even be keratotic.[3,6] Rate of local recurrence is low and it rarely metastasizes.[7,8] The treatment of this lesion is mainly complete surgical excision. TLC should be differentiated from other skin tumours. Since it is a rarely encountered tumour, it is very challenging even for an experienced pathologist to make a definitive diagnosis. We report here a rare case of trichilemmal carcinoma occurring on an uncommon site i.e the upper lip.

Case Report

A 55 year old male patient reported to the out patient department with an ulcerated growth over the vermillion border of the right upper lip. The patient gave a history of an ulcerated growth present for the past one year. The ulcerated nodular mass measured around 2 x 4 cm with irregular margins and mild discharge of blood (Fig 1a). Patient gave a history of squamous cell carcinoma 5 years back in the right auricular region for which he underwent high dose radiation therapy 5 days per week for 5 - 7 weeks. There was no history of any inherited or systemic illness. No other significant finding was found on head and neck examination. A clinical differential diagnosis of Basal cell carcinoma (BCC) or Squamous cell carcinoma (SCC) of the upper lip was made. An incisional biopsy was performed and the specimen sent for histopathological examination.

The haematoxylin and eosin stained specimen showed tumour cells proliferating in nests and islands into the underlying dense connective tissue stroma with a pushing margin. There was lobular epithelial proliferation with areas of trichilemmal keratinisation and necrosis. Cells with palisading hyperchromatic nuclei could be seen at periphery of the tumour nodules. Another population of cells with abundant clear cytoplasm with prominent nucleoli showing high mitotic activity and pleomorphism was also seen (Fig 1b & c). These features were consistent with the diagnosis of trichilemmal carcinoma. Surgical excision of the tumour with lip reconstruction was advised for the patient but the patient did not return for the surgery.

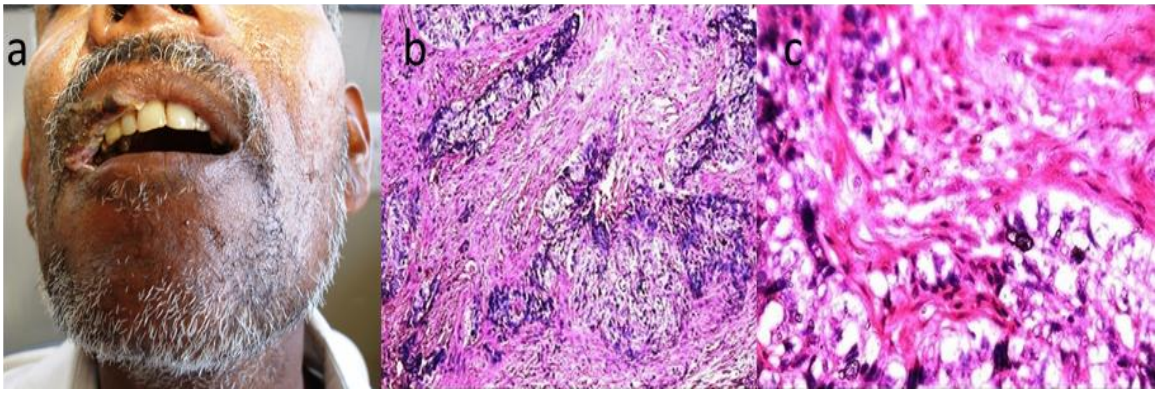

Figure 1: The Clinical photograph of patient showing ulcerative growth having irregular margins with brownish in colour on the vermillion border (a). The hematoxylin and eosin stained photomicrograph at low power view shows lobular proliferation of the tumor with palisading hyperchromatic cells (b) and at higher magnification, clear cells and pleomorphism is clearly evident (c).

Discussion

The term trichilemmal carcinoma was first given by Headington 1976, to describe a skin tumour which is a locally invasive, histologically malignant neoplasm of clear cells derived from the external root sheath epithelium of a hair follicle.[9] The pathogenesis of TLC remains unclear. It is the malignant counterpart of trichilemmoma and has also been reported to be associated with the burn scar.[10] Other factors like exposure to sunlight, repeated as well as long term doses of radiation have been implicated in the pathogenesis of trichilemmal carcinoma.[5] In the present case, the patient had a history of irradiation for squamous cell carcinoma in the left auricular region which may play a significant role in the etiopathogenesis of TLC.

Clinically the lesion presented as a solitary ulcerated mass on the upper lip which was brown in colour with a mild bloody discharge. The literature on TLC involving upper lip is insufficient, except for a recent case in which TLC was associated with xeroderma pigmentosa (XP) in a 25 year old man who presented with the history of photophobia and widespread brown pigmentation of skin. He had ulcerations on the face over the eyelid and upper lip. According to the authors the diagnosis of XP was entirely clinical but the TLC was confirmed only after the histopathological examination which revealed the characteristic lobular arrangement of the tumour along with clear cells.[11]

Histologically, TLC has a lobular, growth pattern, the tumour aggregates extend deep in the dermis up to the subcutaneous fat.[2] The tumour cells have a characteristic glycogen rich cytoplasm having multiple nucleoli. At the periphery of the lesion the epithelial cells show palisading and have a distinct thickened basement membrane.[12] Atypical mitotic figures and pleomorphism can be identified. The neoplastic cells inside the tumour aggregates exhibit trichilemmal keratinisation.[13] Some of these tumours show a pagetoid spread.[6,12] The present case exhibited the above findings suggestive of the diagnosis of TLC.

TLC must be distinguished histologically from other clear cell skin tumours particularly the clear cell variants of BCC and SCC. Lesions of SCC do not show lobular proliferation and trichilemmal keratinisation.[14] Keratinisation in SCC, when present, is infundibular. Presence of peripheral palisading alone does not confirm the diagnosis of basal cell carcinoma. The clear cells in BCC occupy only a small portion of the tumour aggregate.[13,14] According to Barr *et al* the electron microscopic studies have demonstrated that the clear cell change is the result of a degeneration phenomenon involving lysosomes.[15] Malignant proliferating tumours also have a similar histological picture but they usually arise in a pre existing cyst and have a predilection to occur in scalp. They also show high mitotic activity and sometimes abrupt keratinisation with frequent metastasis.[16] Establishing a diagnosis of malignancy is very important for therapeutic and prognostic purposes. Moreover some of these tumours may be associated with syndromes like multiple trichilemmoma seen in Cowden syndrome.[1]

TLC is an aggressive tumour but rare cases of metastasis have been reported. There is no unanimous consensus regarding the treatment of TLC, although surgical excision with tumour free margins is curative.[3,6] Recently Mohs' microsurgery technique has been successfully employed.[2] This technique is used for the treatment of contiguously spreading cutaneous cancers with precise microscopic control. Its primary advantages are conservation of normal surrounding tissues and structures, a high curative rate and low operative risk.[17] To establish a diagnosis of TLC, the specimen must be examined meticulously. The site, colour, (tan, brownish or telangiectatic), pattern of growth (lobular or plaque) should be carefully recorded. Since it is a rare malignant skin tumour, the diagnosis mainly relies on the histological evaluation.

TLC is a locally invasive and cytologically atypical tumour with very few cases of metastasis. Its treatment is less aggressive compared to the more common high grade malignant skin tumours like SCC, BCC and other clear cell tumours. Because of its clinical as well as histological similarity to these tumours it is essential to consider the diagnosis of TLC, so that an appropriate treatment can be rendered to the patient with minimum morbidity. Further, as the occurrence of TLC on upper lip is rare and the literature on the same is insufficient, the addition of this case report to the literature may prove helpful for reference in future cases.

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