Trichilemmal Carcinoma of Nasal Ala: A Case Report

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ABSTRACT

Trichilemmal carcinoma is a rare malignant adnexal tumour. It closely mimics the clinical presentation of squamous cell carcinoma. The management protocol is similar to non-melanotic skin cancers. Definitive diagnosis is made on the histopathology. We present a case of trichilemmal carcinoma of the right ala of the nose and the reconstruction of the defect. The patient underwent wide local excision and staged reconstruction. Aspects of diagnosis, management, and reconstruction are discussed.

Key Words: *Trichilemmal carcinoma, Nose, Reconstruction, Adnexal neoplasm.*

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INTRODUCTION

Trichilemmal carcinoma (TC) is a rare malignant adnexal tumour.[1] It originates from the outer sheath of the hair follicles. It commonly occurs over sun-exposed areas of the body. It usually presents as polypoidal growth with no specific symptoms; however, it may ulcerate or bleed in some patients. TC is usually mistaken as squamous cell carcinoma (SCC), basal cell carcinoma (BCC), or melanoma. Treatment options include Mohs micrographic surgery and wide local excision. Regular follow-ups and surveillance are needed in the postoperative period. Metastasis and recurrence are rare in these tumours.[2]

We herein present a case of TC of the right ala of the nose, which was managed with wide local excision and staged reconstruction of the defect.

CASE REPORT

A 42-year Asian female patient visited the clinic with the complaint of a pigmented lesion on the right ala of her nose for the past 2 years. The patient was a known hypertensive with no other comorbids. There was no history of any trauma. The lesion was initially increasing slowly in size but rapidly enlarged in the last 4 months. It was associated with itching and occasional bleeding.

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On clinical examination, it was a pigmented lesion of size 3×2 cm with surrounding telangiectasias (Figure 1). There were no cervical lymph nodes or presence of any other such lesions over the body. Incisional biopsy was performed previously from an outside facility which reported it to be a moderately differentiated SCC. Contrast-enhanced maxillo-facial CT scan was done preoperatively to rule out bony or intranasal involvement. The case was discussed in the tumour-board meeting and the management plan was made of wide local excision under frozen control, followed by reconstruction. After attaining negative margins, the defect was assessed. The defect consisted of part of the cheek unit, nasal wall, and ala of the right side (Figure 2). It was decided to reconstruct this defect by using a cheek advancement flap and staged forehead flap. Markings for both flaps are shown in Figure 2. Cheek advancement was used to cover the cheek defect and a forehead flap was used to reconstruct the ala and lateral nasal wall. No conchal graft was used to recreate the alar rim. The patient recovered uneventfully and the flap was divided and inset after 6 weeks under local anaesthesia. The final histopathology, which was performed from our own facility, reported no residual tumour and the patient was kept on surveillance. A review of the original biopsy showed the tumour to be TC rather than SCC (Figure 4). The patient follows up every 3 months and is disease-free till the last follow-up (Figure 3).

Trichilemmal keratinisation refers to a specific type of keratinisation observed in TC. Trichilemmal keratinisation is characterised by the formation of keratinocytes resembling those found in the outer root sheath of the hair follicles. These keratinocytes exhibit distinct morphological features, including pale eosinophilic to clear cytoplasm and the presence of trichilemmal-type keratinisation. This histopathological finding is essential for the diagnosis of TC and helps differentiate it from other skin tumours (Figure 4).

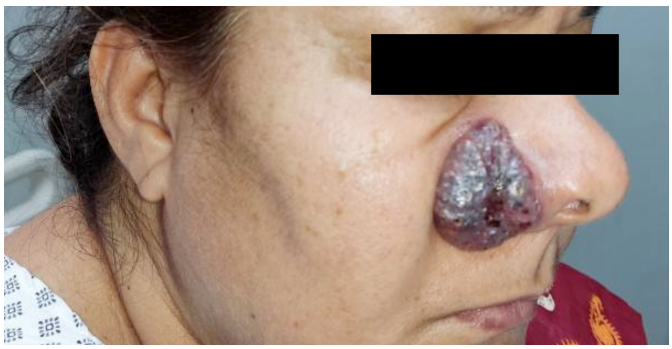

Figure 1: Preoperative photograph of the patient showing the lesion.

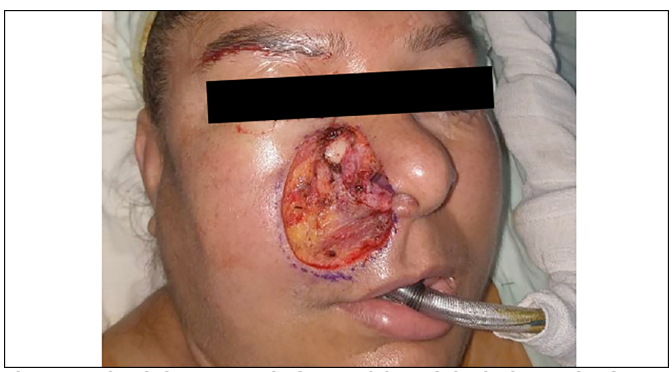

Figure 2: The defect created after excision of the lesion under frozen control.

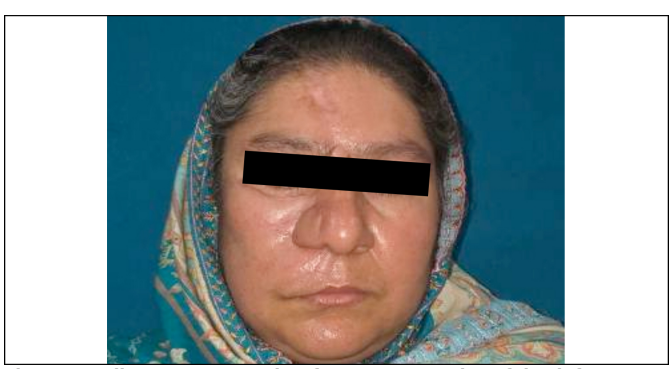

Figure 3: Follow-up at 3 months after reconstruction of the defect.

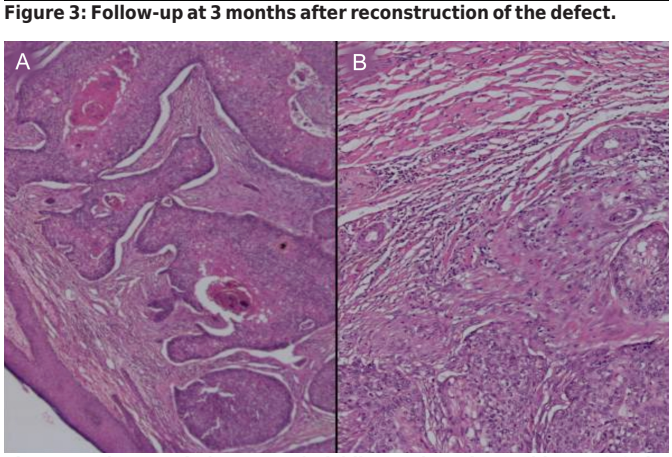

Figure 4: Microscopic features of trichilemmal carcinoma biopsy. (A) The section on the left shows tumour comprising of trabeculae and strands of basaloid cells attached with the overlying epidermis. The tumour cells exhibit prominent nuclear pleomorphism, hyperchromasia and brisk mitotic activity. Central keratinisation is also noted. (B) On higher magnification on the right: The cells show pale eosinophilic to clear cytoplasm. Foci of trichilemmal type of keratinisation is also seen. Tumour border is pushing type.

DISCUSSION

TC was first described by Headington in 1976 who described it as an epidermal-based tumour composed of atypical clear cells derived from adnexal keratinocytes.[3] It usually presents as a solitary lesion over sun-exposed areas of the body and has a clinical resemblance to SCC or BCC. In a review of 103 cases of TC, about 51% were found on the face, and out of these, only seven cases were of TC involving the nose.[4]

Risk factors for TC are similar to those of other skin tumours, ranging from sun exposure, immunosuppression, ionising radiation exposure, and genetic disorders (xeroderma pigmentosa, Cowden disease). However, this carcinoma has been reported in non-sun-exposed areas as well.[5] Apart from actinic damage, another striking risk factor is immunosuppression. A study from South Africa reported TC in a patient undergoing treatment of breast cancer.[6] Another report described the development of multiple TCs on the chest of a tuberculosis patient who had undergone multiple pneumonectomies and received multiple chest x-rays over the course of 30 years.[7] This patient, however, had none of these risk factors. TC has been reported mostly in older patients. However, it has been reported in children as low as 9 years of age in the literature.[1]

The usual presentation is of a solitary nodule with surrounding telangiectasia. Lesions are crusted or pigmented and can mimic BCC, SCC, or nodular melanoma. These lesions usually present over hair-bearing skin. To the best of our knowledge, there is one such exception, where a study reported TC of the nasal cavity.[5]

This tumour can be distinguished from other mimics upon histopathology, where it shows trichilemmal keratinisation. Other histological characteristics of TC include prominent nuclei, translucent cytoplasm, and atypical mitoses.[8] Ki-67 and *p53* can help distinguish the malignant nature of the lesion.[9] It is imperative to distinguish it from other cutaneous tumours, although such features may overlap. Immunohistochemistry can help in the differential diagnosis. TC is usually EMA, Ber-EP4, and *CD34* negative, whereas Ber-EP4 is positive in BCC. TC also exhibits a pushing-type growth pattern as opposed to an infiltrating one.

Wide local excision with histologically clear margins has been the treatment of choice. We took a 1 cm margin all around the tumour. Frozen section was done to attain clear margins and the defect was appropriately reconstructed. Mohs micrographic surgery is becoming increasingly popular for the treatment, but due to lack of services in our setup, we chose to excise the tumour. Owing to the small sample sizes, there is no clear evidence of one technique being superior to another in terms of recurrence. However, since Mohs surgery preserves healthier tissue, it is deemed to be superior in that aspect, as TC usually occurs in cosmetically challenging areas of the face.

TC is an indolent tumour. The recurrence rate is low but has been reported in literature.[4] It also has very low metastatic potential. Few metastatic examples have been reported.[10] The

consensus is for regular follow-up to detect recurrence or metastasis. This patient had no recurrence after 1 year of surgery, although the time elapsed is less to comment definitively.

TC is a rare cutaneous malignancy which closely mimics other skin tumours. Majority of cases have been managed with surgical excision. Mohs surgery and wide local excision compare favourably with each other, although the sample size is smaller. In the absence of established guidelines, regular follow-up similar to non-melanotic skin cancers should be employed.

PATIENT'S CONSENT:
Informed consent was taken from the patient.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
FS, SA: Drafting the manuscript.
MR, NM: Manuscript checking.
MG: Data collection.
All authors approved the final version of the manuscript to be published.

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