UNFOLDING THE LITERATURE OF A RARE CASE OF TRICHILEMMAL CARCINOMA

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ABSTRACT

A rare case of trichilemmal carcinoma in a 40-year-young female is reported. She presented with a horny ulcerated lesion over anterior aspect of left arm. Strong history of exposure to sun, which is the main contributory factor for its aetiopathogenesis is present. Patient underwent wide surgical excision and diagnosis was established on histopathological examination with positive surgical margins. Hence adjuvant radical radiotherapy was administered. She is now on disease free followup for last 4 years.

KEYWORDS
Trichilemmal Carcinoma; TLCA; Radiation-Induced; Skin Neoplasms.

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INTRODUCTION: Trichilemmal carcinoma (TLCA) is a rare skin neoplasm and only infinitesimal cases have been reported in the literature. The trichilemmal carcinoma is considered as a rarely metastasising low malignant lesion.[1] As a rule, it appears as a solitary lesion and occurs on sun-exposed skin, especially on the face, neck, scalp and hands commonly between the 4th and 9th decades of life.[2] The diagnosis is established by histopathological examination complemented with immunohistochemistry of the lesion. The present report is a rare case of trichilemmal carcinoma treated with wide surgical excision and adjuvant external beam radiotherapy and the outcome.

CASE REPORT: A 40-year-old married female presented with a history of nodular mass over anterior aspect of left arm for over 10 years. The mass was gradually progressing, localised brownish nodule of 9 cm size with horny projections. At presentation, the tumour was ulcerated, bleeding, exfoliative and with mild pain. There was no history of weakness, fever, loss of sensation or numbness in the affected limb. No other significant comorbidities were noted. Patient was a chronic beedi smoker for 15 years, non-alcoholic and directly exposed to sun for prolonged periods. Systemic examination was normal. Local examination of the lesion revealed tumour mass on the anterior aspect of left arm as described previously and remaining skin was normal. No enlarged lymph nodes were detected. Complete haemogram, routine blood biochemistry and chest radiograph were normal. Abdominopelvic ultrasonography of the patient was normal.

The patient underwent wide excision of the exfoliative growth and the wound was covered using the back trunk skin-grafting dermatome [Figure: 1].

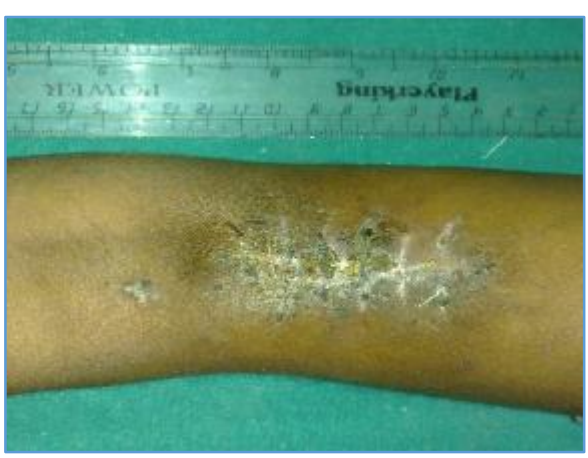

Fig. 1: Showing skin-grafting dermatome in a post excised area seen over anterior aspect of left arm

The pathological examination of 6.5×5.5×3.0 cm excised nodular mass revealed presence of infiltrative tumour with pilar type of keratinisation and pagetoid spread of lesion cells into epidermis with positive margins. Connective tissue sheath revealed peripheral palisading of atypical keratinocytes with pleomorphism and atypical mitotic figures and diagnosed as trichilemmal carcinoma, a malignant tumour of adnexal origin.

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In view of the positive margins in surgical resection, one month after surgical excision, the patient underwent adjuvant external beam radical radiotherapy 45Gy/25#/5 weeks. Four years after the radiation treatment, she is on disease free followup for last 4 years.

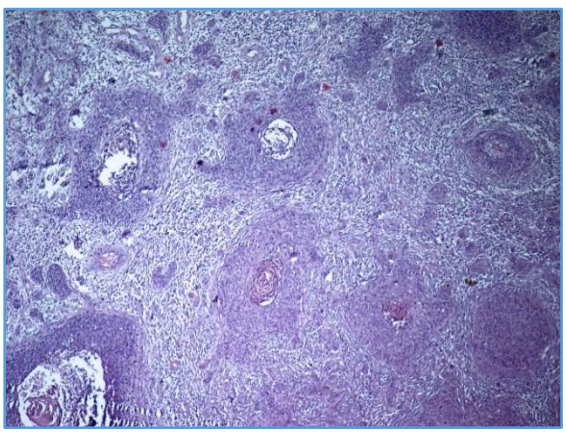

Fig. 2: Photomicrograph H&E stain; magnification×40 revealed multiple lobules of proliferating atypical keratinocytes.

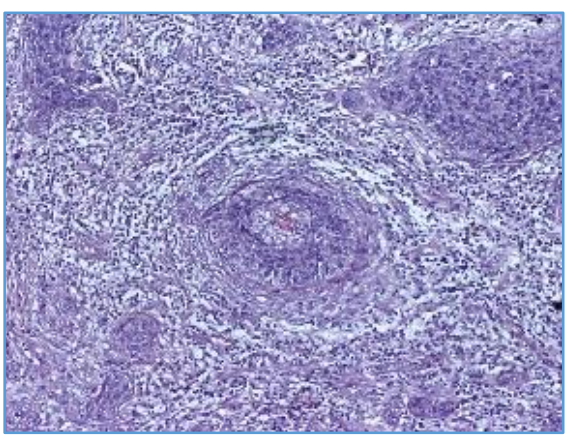

Fig. 3: Photomicrograph H&E stain; magnification×100 revealed a lobule surrounded by prominent connective tissue sheath with peripheral palisading of keratinocytes.

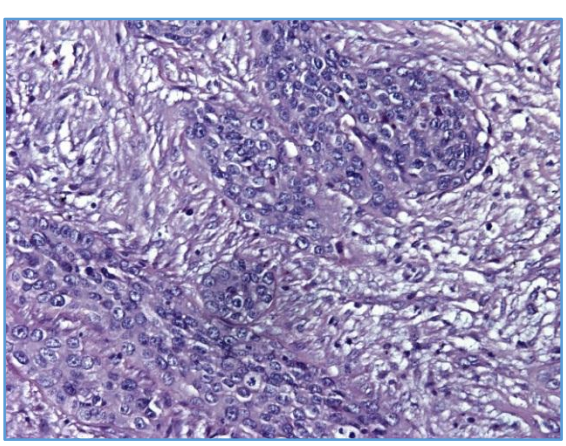

Fig. 4: Photomicrograph H&E stain; magnification×200 revealed atypical keratinocytes with clear cell change, pleomorphism and atypical mitotic figures.

DISCUSSION: The term trichilemmal carcinoma (TLCA) was originally described as a clinical entity by Headington in 1976.[1] Trichilemmal carcinoma is a rare cutaneous neoplasm that develops from the external root sheath of the hair follicle and is considered as a malignant counterpart of trichilemmoma.[2] So far only infinitesimal cases have been reported in the literature. It usually occurs on sun-exposed skin, especially on the face, neck, scalp and hands commonly between the 4th and 9th decades of life.[3] However, it may occur at non-sun-exposed site and may rarely metastasise.[4] There is no gender-based predisposition.[3] It has also been reported in patients exposed to radiotherapy treatment of benign head and neck diseases with long latency periods and accordingly as per the subjected radiation dose.

The histogenesis of TLCA remains unclear, although it indicates a high-grade malignancy, its biological behaviour appears to be relatively benign, and so it is considered to be a low-grade carcinoma with low metastatic potential.[2] The features of TLCA resemble the outer root sheath and their lesions are usually slightly raised, pale, tan, or reddish, frequently ulcerated or with crusts. Grossly, the tumours have been described as exophytic, ulcerated, polypoid or nodular lesions that may be keratotic and the histology demonstrates lobular proliferation centred on pilosebaceous structures in continuity with the epidermis or follicular epithelium as a pagetoid interface and is characterised by the presence of clear cells with atypical nuclei and trichilemmal keratinisation as present in our case also.[5,6]

Immunohistochemistry of the TLCA is usually negative for CEA (carcinoembryonic antigen) and EMA (epithelial membrane antigen), although late positive results have occasionally been documented.[6] Kurokawa et al. differentiated TLCA from follicular infundibulum by expression of cytokeratins CK 1, 10, 14 and 17 in TLCA.[2] CK 15 and 16, present in tumour nests in trichilemmoma, were absent in TLCA. Jih et al. reported the absence of CK15 in squamous cell carcinoma.[7] Absence of CK 15 might be responsible for malignant transformation. CK 16, hyperproliferative keratin was absent in TLCA.[2,7] These results suggest that the absence of CK 15 and 16 may be associated with malignant transformation from trichilemmoma to TLCA. In a comparison of CK expression between TLCA and trichilemmoma, the absence of CK 15 and 16 in TLCA may be related to transformation from trichilemmoma to trichilemmal carcinoma.[2]

In differential diagnosis, squamous cell carcinoma, basal cell carcinoma, malignant nodular melanoma, proliferating trichilemmal tumours or keratoacanthoma should be considered.[1-3] These malignant tumours are more aggressive than trichilemmal carcinoma. In addition, TLCA is usually a solitary lesion, occurring separately from the proliferating trichilemmal tumour.[1] Proliferating trichilemmal tumours (PTTs) usually arise in a pre-existing trichilemmal cyst and are frequently confined to the scalp and are larger than TLCA.[8] Making this diagnosis is imperative as the biological behaviour of TLCA is less aggressive than that of other epithelial malignancies, requiring a different treatment approach, mostly curative.[9]

Monoclonal antibodies against cytokeratin (CK) are crucial markers for evaluating the origin of epithelial tumours and the stage of differentiation.[2] The sensitivity of CK 5/6 marker in the malignant lesions (other than basal cell carcinoma) is 78%, while the specificity is 67%.[10] p53 tumour suppressor gene mutations are often found in patients with non-melanoma skin cancer. Complete loss of p53 on immunohistochemical staining represents the malignant transformation of PTT.[11] Also, focal staining for the presence of CD 34 is a marker of differentiation from the outer hair sheath.[8]

Many treatment options including radiotherapy and chemotherapy have been tried in the past, but mainstay of successful outcome remains surgical excision with wide margins and removal of regional lymph nodes. Moh's micrographic surgery has also been performed by many dermatologists to avoid wide excision.[6] Swanson et al analysed the histological and clinical findings in 10 cases of TLCA and all patients were treated by a wide local excision and neither recurrence nor metastasis was reported even after 8 years of followup.[12] In addition to Moh's micrographic surgery and surgical excision, imiquimod 5% cream is a promising therapeutic option for TLCA because of its nonsurgical approach and excellent cosmesis.[13] The trichilemmal carcinoma generally has good prognosis and reports of deep invasion and local recurrence cases are uncommon. However, there are cases of local recurrences and also metastatic disease which most commonly results after a gap of few years of incomplete surgery, subclinical metastatic or nodal disease at the time of surgery or inappropriate metastatic workup.[3] There is no established chemotherapy regimen for metastatic TLCA reported in the medical literature. However, Hayashi et al. administered chemotherapy regimen with cisplatin, Adriamycin, vindesine; usually known as CAV protocol.[6,14] Radiation or chemotherapy is used to treat recurrent or metastatic cases of TLCA, but the effectiveness of such treatments is unknown.[5] Local excision with removal of the regional lymph nodes and followup is usually the best treatment option. Adjuvant radiotherapy has shown promising results in surgical margin positive cases as in our case.[4,10] No chemotherapy was used. She is now on disease-free followup for last 4 years.

CONCLUSION: Trichilemmal carcinoma is a rare cutaneous neoplasm which usually occurs between the 4th and 9th decades of life. It develops from the external root sheath of the hair follicle of the sun-exposed skin areas. Histopathology with IHC plays important role for establishing the diagnosis and differentiating it from similar conditions like squamous cell carcinoma, basal cell carcinoma, malignant nodular melanoma, proliferating trichilemmal tumours or keratoacanthoma. Mainstay of treatment remains surgical excision with wide margins and removal of regional lymph nodes, which is mostly curative; however, in recurrent or metastatic cases, radiation or chemotherapy should be tried to explore the best results.

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