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Original Research Paper

Radiotherapy

A RARE CASE OF TRICHILEMMAL CARCINOMA WITH BRAIN AND LUNG METASTASIS

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ABSTRACT Trichilemmal carcinoma is rare cutaneous tumor affecting scalp, eyelids, neck and face. Generally seen in elderly women, it is large, often cystic and characterized by trichilemmal keratinization. However, it can have an aggressive clinical course propensity for distal metastasis. I report a case of 40 years old female who presented with scalp swelling in July 2019.She underwent wide local excision and histology revealed trichilemmal carcinoma. Patient lost to follow up and presented in 2022 with CT thorax showed lung metastasis and CT brain shows brain metastasis and she advised for palliative radiotherapy. Trichilemmal carcinoma with brain and lung metastasis is rare condition. The patient underwent palliative radiotherapy and expired 2 months after palliation.

KEYWORDS : Trichilemmal, Tumor, Metastasis, Palliative Radiotherapy

INTRODUCTION

Trichilemmal carcinoma is rare, malignant, adnexal neoplasm that is derived from outer root sheath of hair follicle. Their characteristic histological finding is the sudden compact amorphous keratinization of the epithelial cells that cover the cyst wall without a granular layer, and this phenomenon is called trichilemmal keratinization. These tumors predominantly occur in elderly patients on sun exposed areas. The lesion is often localized on the scalp, but it can also be observed more rarely on the neck, trunk, groin, lower and upper extremities. The mean age of diagnosis is 70 years with slight male predominance.

The lesions are commonly identified as a papular, nodular, sometimes exophytic. Because the tumor is rare, its biological behavior is unpredictable, and it is frequently confused with SCC histopathologically, standard guidelines for treatment could not be established. Most of the case reports in the literature have focused on the pathological features of the tumor rather than its clinical behavior and management. They generally arise de novo but may also derived from underlying proliferating trichilemmal cyst. They can be locally aggressive and metastasis rarely.

CASE REPORT

Here we present a case of 40 years old female who presented with scalp swelling in July 2019. Her CECT brain at that time suggestive of 6×5×9 cm soft tissue lesion in extra calvaria region of right parietal region, she underwent wide local excision and on histopathological examination suggestive of adnexal neoplasm, immunohistochemical staining (IHC) was positive for P63, ck7, CEA suggestive of trichilemmal carcinoma. Patient lost to follow up and presented in May 2022 with CT thorax showed lung metastasis and CT brain shows brain metastasis. Patient underwent palliative radiotherapy, we report this case because of its rarity.

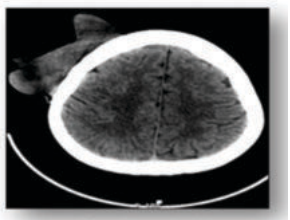

Fig.1 CT of head revealed large soft tissue swelling in right side of scalp

DISCUSSION

Proliferating trichilemmal tumor is the proliferating form of pillar cysts. This dermal neoplasm was first described by Wilson-Jones in 1996 and stated that it has a histological capacity that mimic SCC. Trichilemmal carcinoma is a rare, adnexal tumor with evidence for follicular outer root sheath. It is considered the malignant analogue of trichilemmoma. Clinical presentations are variable, due to its ability to resemble different clinic entities, the diagnosis of trichilemmal carcinoma relies on histological evaluation accompanied by IHC. Microscopically trichilemmal carcinoma features a solid, lobular, trabecular pattern. The tumor cells are clear, polygonal, glycogen rich. Trichilemmal carcinoma typically demonstrates non aggressive clinical behavior. Wide local excision with histological demonstration of clear margins has been treatment of choice. Mohs micrographic surgery also has been increasingly employed for numerous rare adnexal tumors including trichilemmal carcinoma. Adjuvant radiation therapy should be generally reserved for recurrent disease or residual macroscopic disease. Trichilemmal carcinoma with brain and lung metastasis carries a very poor prognosis. Palliative radiation therapy and Palliative chemotherapy is treatment for trichilemmal carcinoma with distal metastasis.

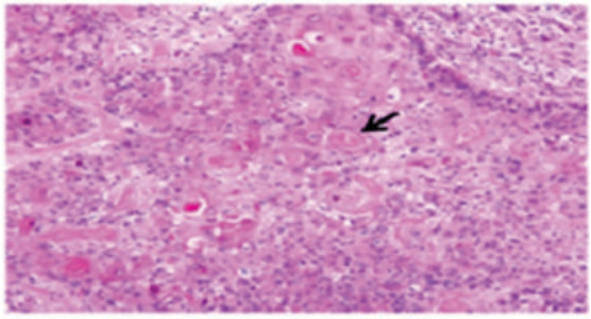

Fig. 2 Microscopy shows tumor cells arranged in sheets, cords and clusters with areas of keratinisation seen.

CONCLUSIONS

Trichilemmal tumors may exhibit aggressive local invasion across tissue plane. Multidisciplinary approach is required to diagnosis and treatment. It is not easy to correlate the histopathological features of tumor with its clinical presentation. It is essential to distinguish it from other similar

looking neoplasms. Trichilemmal tumor with intra cranial extension and lung metastasis is a rare entity and carries a poor prognosis. To create treatment algorithms with sufficient level of evidence there is need for increased case reports and comparative long-term results of different treatment options. Until these happen, close clinical follow up is most effective way to detect recurrence and metastasis.

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