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CASE REPORT

Pilomatrical Carcinoma of the Scalp in a Setting of Pilomatricoma and Trichilemmal Cysts—a Rare Entity

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Abstract

Pilomatrical carcinoma is one of the rare adnexal tumors with only 140 reported cases according to recent literature. The tumor has a propensity to arise in posterior neck, upper back and lower extremities and has 100% mortality if metastasizes. Hence, it is imperative to document the illusive nature of this deceptive tumor. The case under discussion is that of a 70-year-old male presenting with multiple small to large non-tender firm lesions over the scalp. The tumor, labeled as proliferative trichilemmal cyst, radiologically was excised, and the scalp was reconstructed by mobilization of the local area. Extensive sampling of the specimen received revealed morphology of pilomatricoma and trichilemmal cysts with a focus of stromal invasion in one of the lobules of pilomatricoma. Hence, a diagnosis of pilomatrical carcinoma was made, and the patient is on close follow-up since 1 year post-operatively with no signs of recurrence or cervical lymphadenopathy. Though of low malignant potential, the tumor is said to have recurrence potential. The prognosis is variable depending on the evidence of metastasis found if, at all, the mortality is 100%. Hence, extensive sampling and accurate diagnosis are essential to rule out the possibility of malignancy.

Keywords Pilomatrical carcinoma · Pilomatricoma · Trichilemmal cysts · Scalp

Introduction

Pilomatrical carcinoma is one of the rare adnexal tumors with only 140 reported cases according to recent literature [1]. It was first described in the year 1950 by Lopansri and Mihm [2]. The tumor has a propensity to arise in posterior neck, upper back and lower extremities and is common in males of older age groups [3–5]. The purpose of reporting this case is to emphasize the deceptive nature of the tumor as its histomorphological features may commonly be camouflaged by features of a co-existing proliferating trichilemmal tumor, though the management of the patient does not differ much. The case under discussion is a 70-year-old male presenting with multiple variably sized firm lesions over the scalp

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clinically suspected as sebaceous cysts. The histopathological examination revealed it to be a pilomatrical carcinoma in a background of multiple pilomatricomas and trichilemmal cysts.

Case Report

A 70-year-old male presented with multiple soft to firm swelling over the scalp since 3 years. The swellings were insidious in onset, and the patient did not complain of any loss of consciousness, diminished appetite, or significant weight loss. On examination, the swellings were firm to tense cystic and adhered to overlying skin. A non-contrast CT scan of the brain revealed multiple large exophytic scalp swelling of soft tissue attenuation with few hypodense cystic areas and areas of calcification noted arising from subcutaneous planes in bilateral occipital, right temporal, and bilateral parietal region, largest measuring 8.2 × 6.7 × 9.5 cm (AP × TR × CC) over left retro-auricular-parieto-occipital region. The features were suggestive of proliferative trichilemmal cyst (Fig. 1). Fine needle aspiration was performed from the multiple swellings which revealed blood mixed smears showing few clusters of squamoid epithelial cells with crystalline to amorphous debris, several foreign body multinucleated giant cells, and dispersed and few aggregated macrophages and inflammatory cells.

The largest swelling and all other smaller swellings were excised and sent for histopathological examination.

On gross examination, multiple swellings were received, largest swelling measuring 11.5 × 9 × 5.5 cm. On microscopical examination, sections from the largest tissue piece showed tissue lined by normal skin with the dermis showing solid sheets and nests of invasive tumor comprised of outer rim of basaloid cells with extensive keratinized material and shadow cells in the center with individual cells having nuclear atypia, vesicular nucleus, and punctate nucleoli. Brisk mitosis was noted. Focal nests and singly scattered cells were noted infiltrating into the underlying stroma. Areas of calcification and squamous differentiation were seen. Sections from the smaller tissue pieces also showed similar morphology along with occasional bizarre cells invading the stroma. Brisk mitotic activity was noted (Fig. 2). Focal areas of skin ulceration were noted. Few of the smaller skin-lined tissue fragments also showed the features of pilomatricoma and trichilemmal cyst. Lymphovascular invasion was not identified. Immunohistochemistry was performed; however, it proved to be non-contributory in differentiating the infiltrating component from the benign counterpart.

Lymph node dissection was not performed. However, the patient is on close follow-up and shows no evidence of local recurrence or metastasis.

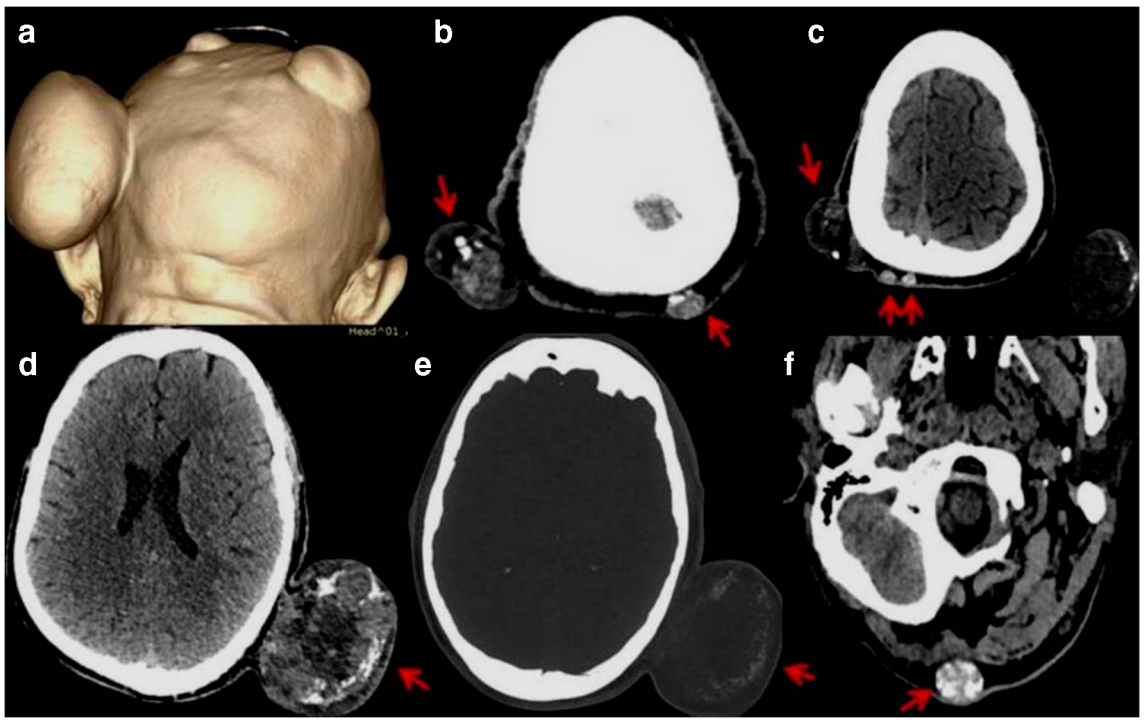

Fig. 1 A non-contrast CT scan of the brain from the base of the skull to the vertex showing multiple large exophytic scalp swelling of soft tissue attenuation with few hypodense cystic areas and areas of calcification arising from subcutaneous planes in bilateral occipital, right temporal, and bilateral parietal region

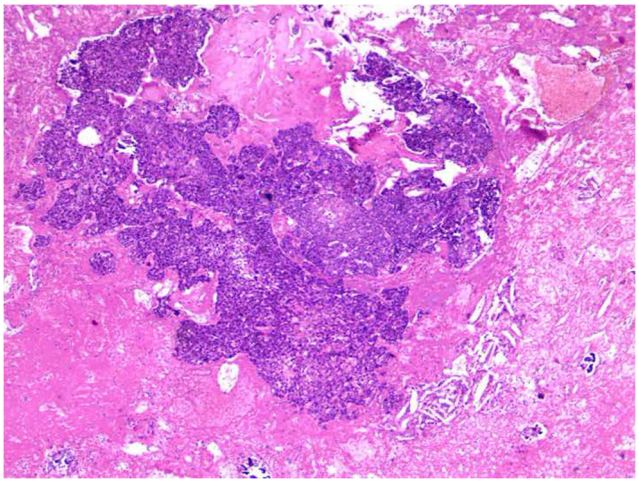

Fig. 2 Photomicrograph showing tumor cells with moderate pleomorphism and few bizarre forms with atypical mitosis. (× 100, H&E)

Discussion

Pilomatrical carcinoma, a tumor of the hair follicle, is hypothesized to develop de novo as opposed to another school of thought which believes it to arise from a pre-existing pilomatricoma. The debatable origin of the tumor is further supplemented by the contradicting epidemiology. Pilomatricoma is common in females and in a younger age group, whereas pilomatrical carcinomas are more common in males of middle to older age groups [3–7]. Our case concurred with respect to the age and the gender.

Furthermore, it has been reported that there are only 13 cases in the existing literature where the carcinoma has been reported on the scalp [8].

The categorization of the tumor into carcinoma depends on the histomorphological features. Tumor asymmetry with infiltrative growth pattern, marked pleomorphism, brisk atypical mitosis, mummified necrotic component vascular, and/or perineural invasion helps in making the diagnosis. The case under discussion had tumor asymmetry with infiltrative margins. Marked pleomorphism and brisk mitotical activity was also seen, though vascular and perineural invasion was not noted.

The immunohistochemical profile of the tumor is variable. Expression of p53 and beta catenin might be instrumental though not very useful.

It is important to diagnose this tumor as a close follow-up is highly warranted. Metastasis, though reported in very few cases, has been associated with mortality in almost 100% of the cases.

Surgical wide excision is the treatment of choice. The effects of radiotherapy are debatable. The treatment guidelines lack standardization due to the rarity of the tumor.

Conclusion

As pilomatrical carcinomas are notorious for being deceptive, it is essential to assess the tissue in its entirety in suspicious cases as the infiltrative component may not be evident on partial processing of the tissue. Moreover, in proven cases of pilomatrical carcinoma, close follow-up is mandatory as metastasis may lead to 100% mortality.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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