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Proliferating Trichilemmal Tumor: A Case Report and Literature Review

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Abstract

Proliferating Trichilemmal Tumor (PTT) is an uncommon append age al skin tumor that affects mainly elderly women. It originates from the external root sheath of the hair follicle and most commonly observes on the scalp with pathological abrupt keratinization without granular cell layer disturbance. Because of limited cases in the literature, the management of malignant PTT seems to be controversial. Although PTT mainly requires wide local excision, many other adjuvant modalities have been tried before. Here, we present a case of malignant PTT in a 42 years female patient who presented with scalp mass over the occipital region in the last four years.

Keywords: Trichilemmal cyst 1; Scalp; Epidermal cyst

Introduction

Proliferating Trichilemmal Tumors (PTTs) are neoplasms derived from the outer root sheath of the hair follicle. Most proliferating trichilemmal cysts arise from inside of a preexisting trichilemmal or pilar cyst. The proliferating trichilemmal cyst is more common in women compared to men and more than 80% of patients are elderly women. It is usually localized to the scalp, and about 90% of the cases occur on the scalp. The tumor shows abrupt keratinization without intervening granular cell layer (trichilemmal keratinization).

Case Report

A 42-years female presented with a history of swelling over the scalp occipital of four-year duration for the first time, gradually enlarging. The patient had a headache and vision loss one month before admission. She had no history of trauma or a similar lesion. On the examination, there was no evidence of regional lymph node enlargement.

On regional scalp malformation, there was a painless single swelling measuring 20 × 10 centimeters in diameter with a smooth surface and soft consistency was seen on the scalp. (Figure 1) Magnetic Resonance Imaging (MRI) showed a heterogenous scalp mass (Figure 2).

Figure 1: Morphological view of scalp.

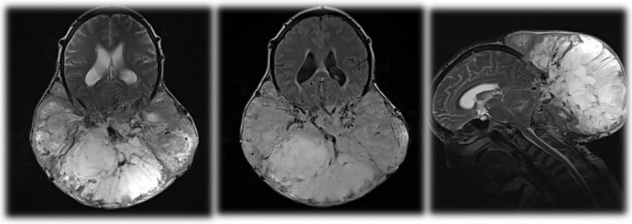

Figure 2: Heterogenous scalp mass.

Digital Subtraction Angiography (DSA) showed tumor-reperfusion. The tumor was supplied from the occipital artery of the external carotid (Figures 3A and 3B). Both arteries were carotid external clips and then biopsy samples were taken from the lesion (Figures 3C and 3D).

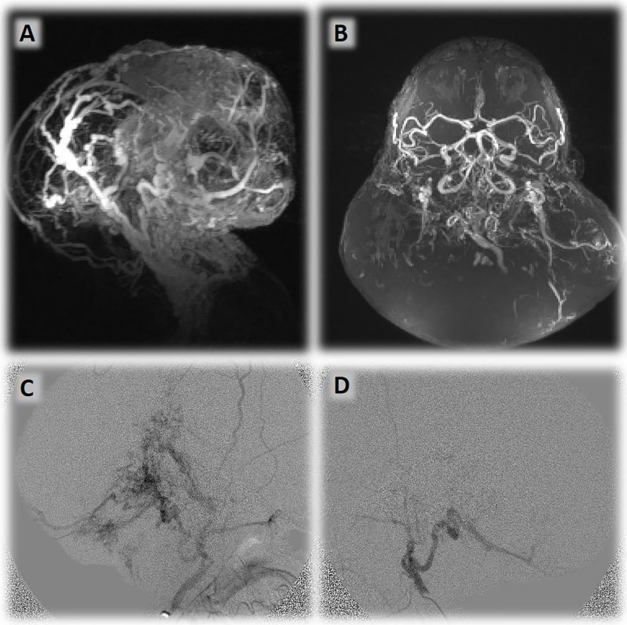

Figure 3: Occipital artery of the external carotid.

After surgery, the patient's lesion showed the progress of size reduction. She was suffering from a swallowing disorder that improved to the gradual.

Discussion

Cutaneous tumors derived from the outer root sheath of hair follicles, which show trichilemmal keratinization, are trichilemmal, proliferating trichilemmal, and malignant proliferating trichilemmal cysts. The trichilemmal cyst is by far the most common form of them. These cysts are mainly seen on the scalp and are relatively common. They well-circumscribed in the dermis with stratified peripheral cells, which rest on the thickened basement membrane. Centrally, the cells enlarge and accumulate abundant intracellular glycogen, with a resultant pale glossy appearance. There is the formation of a dense keratin layer without the interposition of a granular cell layer known as trichilemmal keratinization [1,2].

A PTC is often lobulated and can undergo ulceration [3,4]. It is regularly located on the scalp but can occur on other sites such as wrist [5], chest [5], elbow [6], vulva [7], the mons pubis [8], and buttock [8]. The lesion size can vary from 0.4 cm to 10 cm in greatest diameter [2] but can grow as large as 25 cm2 in the largest dimension [9]. Ulceration can occur over the lesions, mostly the following trauma, and discharge foul-smelling, doughy, whitish material. The lesion can be present from four months to 50 years [3]. About 84% of patients are older than 50 years of age [10] with females predominantly [8].

Lanugo hair follicles of bald scalp and follicles of other areas without nonterminal hair are unlikely to produce these tumors [11]. It has been reported under a variety of names including giant hair matrix tumor, invasive pilomatrixoma, proliferating epidermoid cyst, pilar tumor of the scalp, trichilemmal pilar tumor, trichochlamydocarcinoma, proliferating trichilemmal cyst, PTT, and proliferating follicular cystic neoplasm [12].

Malignant PTT can occurs *de novo*, but most often arises from a pre-existing benign proliferating trichilemmal cyst, the malignant transformation being in a stepwise manner from adenomatous to epitheliomatous and carcinomatous stages [11]. The diagnosis of malignancy in these tumors is predominantly based on histological features, though the biological behavior is unpredictable. In 1983, the first report which specifically identified malignant PTT was published [13]. Rapid enlargement of the lesion has been reported after trauma or spontaneously [3] and has indicated malignant transformation [14]. Malignant PTT, first described in 1983 by Saida et al. [11], is the rarest form of trichilemmal tumors. The biological behavior remains controversial. There is no immunohistochemical marker to detect this malignant transformation, unlike the loss of CD34 expression for PTT. Increased proliferation index and DNA aneuploidy in tumor cells may suggest premalignant nature [15].

Clinically, sudden enlargement of long standing scalp nodule, histologic evidence of abnormal mitoses marked nuclear atypia, and infiltrative margins may reflect malignant transformation [16]. Ye et al. studied 76 patients and categorized them into three groups [17]. Tumors of group-I were well circumscribed, showing trichilemmal keratinization and focal nuclear atypia with surrounding tissue showing infiltration by plasma cells, mononuclear cells, and giant cells. These tumors behaved benignly. Tumors of group-II were early invasive with moderate cytological atypia, foci of single-cell necrosis, and abrupt keratinization, with a desmoplastic stromal response. These were considered locally aggressive. Tumors of group-III were invasive with marked cytological atypia and desmoplastic stroma and considered malignant. Our case showed features of group-III of Ye et al. study.

The features favoring the diagnosis of malignant PTT over squamous cell carcinoma are the presence of trichilemmal keratinization, lobular growth pattern, and lack of precursor epidermal lesion like actinic keratosis. Since MPTT tends to metastasize and recur more frequently than Squamous Cell carcinoma, accurate diagnosis is essential [16]. Malignant PTT is a rare neoplasm with fewer than 100 cases reported in the 50 years since it was first described [18]. The patient should be followed closely after surgery. Patients presenting with large fungating masses along with regional nodal metastasis can show aggressive behavior and merit aggressive treatment.

Conclusion

In summary, malignant PTT is a rare malignant lesion and poses a diagnostic difficulty. Hence, wide surgical excision should be considered as the primary modality of treatment. Long-term follow up is recommended and alternative therapeutic methods need further evaluation.

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