

Giant aggressive forehead tumor: A 15-year follow-up

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Abstract

Proliferating trichilemmal tumors are rare tumors that originate from adnexal structures – specifically from the outer root sheaths of hair follicles. These tumors can be benign or malignant. We report a rare case of a giant aggressive forehead trichilemmal tumor that was paradoxically benign on pathology. It was surgically excised and followed-up for 15 years.

Introduction

Proliferating trichilemmal tumors (PTTs) are rare tumors that originate from adnexal structures specifically from the outer root sheaths of hair follicles. These tumors were first described in 1966 by Jones.¹ They usually arise in areas with high hair density. The scalp is the most common location in more than 90% of the patients but other locations were also described. They usually occur in patients in their sixth decade and are five times more common in women than in men.² There are some cases reported in the literature and the size ranged between 1 and 10 cm but a 25-cm mass was reported on the scalp.³ PTT generally has a benign course but can undergo malignant transformation.²

Here we report a rare case of a giant aggressive forehead trichilemmal tumor that was paradoxically benign on pathology. It was surgically excised and followed-up for 15 years to monitor for recurrence or malignant transformation. To the best of our knowledge, this is the largest tumor reported on the forehead region.

Case Report

A 65-year-old male patient with no significant medical history presented to our hospital for a huge mass on the frontal area that was bleeding easily. The mass has been slowly growing for 22 years, became huge and affected the vision. The lesion became

ulceronecrotic four months prior to presentation and enlarged rapidly after a trial of incision and drainage at another institution.

Physical examination showed a 17-cm dome shaped mass present on the forehead with central necrosis and bleeding (Figure 1).

No evidence of regional lymph node reaction was found in the head and neck region and no additional lesions were noted on physical examination. The rest of examination was normal.

Computed tomography scan showed a heterogeneous large mass containing multiple foci of calcifications and multiple concentric vessels which enhanced significantly. No intra-cerebral extension was seen. No lymphadenopathy was present. These findings were suggestive of either lymphangioma or cavernous hemangioma.

Under local anesthesia, the patient underwent incisional biopsy of the large ulcerating lesion. The histological diagnosis was consistent with proliferating trichilemmal cyst. The patient subsequently underwent, under general anesthesia, a complete resection of the tumor with 1cm margins from the necrotic skin. Absence of the anterior table of the left frontal sinus was noted after tumor excision and the defect was reconstructed with a corrugator muscle flap. Finally, multiple rotation flaps were used to close the defect (Figure 2).

Definitive histopathological study came out as complete excision of PTT (17×12×6 cm) with no evidence of malignancy (Figure 3).

After 3 years, patient presented with recurrent left supraorbital mass measuring 3 cm (Figure 4).

The mass was totally resected with 2 mm margins and sent directly for pathology as a frozen section due to the fear of malignancy. Pathology was consistent with proliferating trichilemmal cyst with no signs of

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malignancy and the margins were free of tumor. So, the defect was reconstructed with local flap. Definitive pathological result confirmed the complete excision of PTT with no signs of malignancy. To note that adjuvant treatment was not given.

Patient was followed biannually for the first year and then annually for 15 years. At each visit, an extensive clinical exam was done and patient showed no signs of local recurrence or distant metastases with long term follow-up.



Figure 1. Forehead ulcerated mass on initial presentation: (A) frontal view, (B) lateral view. The patient's consent has been obtained.



Figure 2. Frontal view image showing the reconstruction done by rotation flaps, post resection of the forehead tumor. The patient's consent has been obtained.

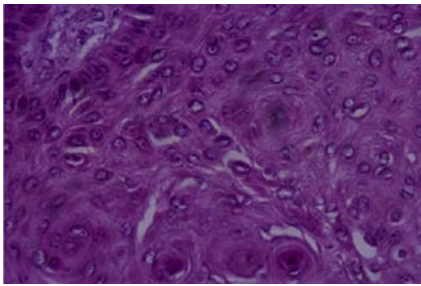


Figure 3. Histological exam showed a cellular tumor composed of variably sized lobules of squamous epithelium undergoing large abrupt change into eosinophilic amorphous keratin.

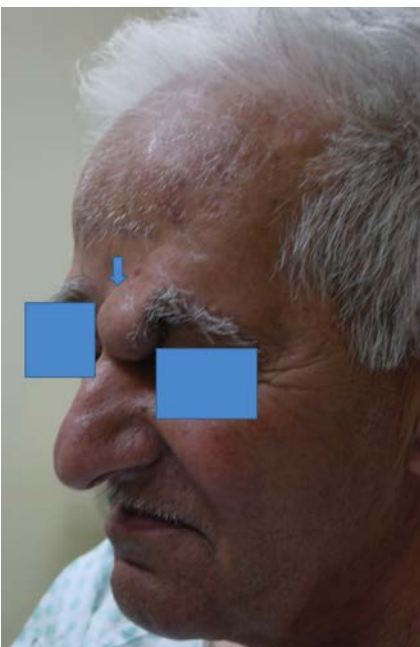


Figure 4. 3 years after resection of the forehead tumor. Arrow: left round supraorbital mass. The patient's consent has been obtained.

Discussion

PTT is a rare benign tumor usually arising from a pilar cyst derived from the outer root sheaths of hair follicles. Usually these tumors have a benign course but can undergo malignant transformation with necrosis, anaplasia, and invasion of neighboring tissues. In this case, they are referred to as malignant PTT.²

Many names are used to describe it: subepidermal acanthoma, invasive hair matrix tumor, invasive pilomatrixoma, trichochlamydocarcinoma, hydatidiform keratinous cyst, giant hair matrix tumor, and trichochlamydoacanthoma.⁴

It is commonly seen in elderly women in areas of excessive hair growth. It is unlikely to grow in non-hair bearing regions.⁵ These tumors are difficult to differentiate from squamous cell carcinoma⁶ and epidermoid cyst, keratoacanthoma, pilomatrixoma, sweat gland tumor, cylindroma, basal cell carcinoma, and angiosarcoma constitute the differential diagnosis.⁴ Macroscopically, they can be exophytic, polypoid, ulcerated, or nodular.⁷

Benign PTT can recur in 3.7% of the cases especially in the cases of limited surgical excision. Recurrence was reported in some cases 10 years post treatment. These lesions tend to be aggressive with local invasion.⁴

Moreover, benign PTT can undergo a malignant transformation and this can occur post infection or trauma.⁸

In our case, incision and drainage may be considered a traumatic event that lead to central necrosis and rapid enlargement. This gave us the impression of a possible malignant transformation and led us to do a total excision of the mass with 1 cm margins from the necrotic skin. To add that the anterior table of the left frontal sinus was absent due to the presence of erosions.

PTT can also be malignant although anaplasia could be absent. Therefore, malignant tumors can be identified based on functional criteria (invasion, rate of growth, and metastasis) or morphological criteria (cellular atypia and anaplasia). So, it is important to consider the clinical aspect of these tumors in their treatment.⁵ Our tumor was benign on pathology but had aggressive features in its clinical course.

Benign PTT is treated by simple excision.² But malignant PTT is very aggressive with high metastatic potential and increased morbidity and mortality and the treatment is controversial due to the limited number of cases. The treatment of choice for those without metastasis is wide local excision with 1 cm of normal tissue.⁹ Ethanol injec-

tion, lymph node dissection, radiotherapy, and chemotherapy can be used as adjuvant treatments in certain cases.¹⁰

Wide local excision, good reconstruction, and long-term follow-up are needed in aggressive tumors where communication between the surgeon and the pathologist is essential.¹¹

Back to our case, we presented a rare case of giant aggressive forehead PTT in a man with long term follow-up where wide excision and reconstruction with no adjuvant treatment seemed sufficient with no local or distant recurrence.

Conclusions

PTT are rare tumors arising from a pilar cyst. They are usually benign but can undergo malignant transformation. Multiple factors can encourage their transformation. Benign PTT on histology can be malignant clinically. Therefore, communication between the surgeon and pathologist is a must. As a result, wide excision should be considered in clinically aggressive types to prevent recurrence, malignant transformation, and metastasis, as evidenced by the long-term follow-up presented in the above case.

References

1. Jones EW. Proliferating epidermoid cysts. *Arch Dermatol* 1966;94:11-9.
2. Kim UG, Kook DB, Kim TH, Kim CH. Trichilemmal carcinoma from proliferating trichilemmal cyst on the posterior neck. *Arch Craniofac Surg* 2017;18:50-3.
3. Casas JG, Woscoff A. Giant pilar tumor of the scalp. *Arch Dermatol* 1980;116:1395.
4. Satyaprakash AK, Sheehan DJ, Sangueza OP. Proliferating trichilemmal tumors: a review of the literature. *Dermatol Surg* 2007;33:1102-8.
5. Sharma R, Sharma S, Verma P, Yadav P. Proliferating trichilemmal tumor of scalp: benign or malignant, a dilemma. *J Cutan Aesthet Surg* 2012;5:213-5.
6. Lindsey SF, Aickara D, Price A, et al. Giant proliferating trichilemmal cyst arising from a nevus sebaceus growing for 30 years. *J Cutan Pathol* 2017;44:639-42.
7. Karamese M, Akatekin A, Abaci M, et al. Unusual invasion of trichilemmal tumors: two case reports. *Modern Plastic Surg* 2012;2:54-7.
8. Ramos-e-Silva M, Peryassu MA,

- Peryassu BC, et al. Proliferating trichilemmal tumor: a case on the nose of a 70-year-old man. *Case Rep Dermatol* 2013;5:248-53.
9. Lobo L, Amonkar AD, Dontamsetty VK. Malignant proliferating trichilemmal tumour of the scalp with intracranial extension and lung metastasis – a case report. *Indian J Surg* 2016;78:493-5.
10. Hayashi I, Harada T, Muraoka M, Ishii M. Malignant proliferating trichilemmal tumour and CAV (cisplatin, adriamycin, vindesine) treatment. *Br J Dermatol* 2004;150:156-7.
11. Erdem H, Yildirim U, Uzunlar AK, et al. Posttraumatic giant proliferating trichilemmal cysts on the parietal region of the scalp. *Indian J Dermatol Venereol Leprol* 2011;77:707-9.

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