Malignant Proliferating Trichilemmal Tumor in the Right Post Auricular Region

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OBJECTIVE To report a case of a malignant proliferating trichilemmal tumor (PTT) in the right postauricular region, and to describe the clinical and histopathologic findings.
METHODS Interventional case report and literature review.
RESULTS A 46-year-old woman presented with a 15-year history of a nodule of 30×30×10 mm in diameter in the right postauricular region. It was diagnosed as a sebaceous cyst. A local mass excision was performed. Histopathologic examination revealed proliferation of the outer hair sheath epithelium with multiple central areas of trichilemmal keratinization. The presence of marked cellular atypia and frequent mitoses indicated a malignant transformation. A second operation employing an enlarged excision was conducted followed by a histopathologic examination showing that there was no malignant tumor remaining. Two weeks after the second operation, 50 cGy of regional prophylactic radiotherapy was applied. The patient was well after 26 months of follow-up and neither recurrences nor metastases were observed.
CONCLUSION *Malignant PTT is a rare skin neoplasm, with its diagnosis* depending on a histopathologic examination. An extend excision is the main treatment after diagnosis.

KEYWORDS: proliferating trichilemmal tumor, treatment, diagnosis.

Proliferating trichilemmal tumor (PTT), as a rare tumor, mostly emerging on the scalp of older women. Descriptions of this tumor are scarce in the literature. In this report we describe a case of malignant PTT, which was initially identified and diagnosed as a sebaceous cyst. A local excision was performed, with the subsequent pathological examination suggesting that it was a malignant PTT. Treatment as well as unique clinical findings of this case are described as follows.

MATERAILS AND METHODS

On October 17th, 2003, a 46-year-old woman came to our clinic with a nodule of 30×30×10 mm in diameter on the right post aural region. The tumor had developed over a period of 15 years. Initially, it was soft and moveable, 5×5×10 mm in diameter, but then grew to 30×30×10 mm within half a year and became fixed and solid. A month earlier, the hair on the skin of the tumor shed. It was diagnosed as a sebaceous cyst. A local excision was performed. Histopathologic examination established the diagnosis as malignant PTT. It showed proliferation of the outer hair sheath epithelium with multiple central areas of trichilemmal keratinization. The presence of marked cellular atypia and frequent mitoses indicated malignant transformation.

Microscopic examination demonstrated the presence of vari-

able-sized lobules composed of squamous epithelium that was well demarcated from the epidermis and surrounding tissue. The epithelium in the center of the lobules showed an abrupt change of the eosinophilic amorphous keratin (Fig.1). Under high magnification, part of the lesion showed a relatively benign area, as well as a malignant area. The presence of marked cellular atypia and mitoses, indicated malignancy within the PTT (Fig.2).

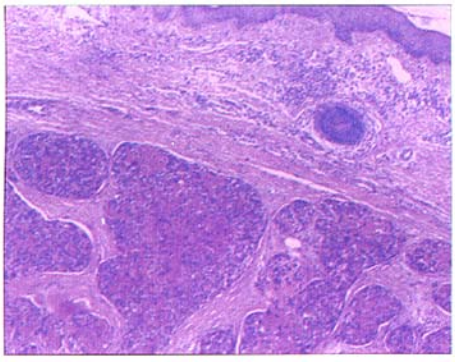

Fig.1. A lobulated tumor showing central, amorphous keratinization beneath the epidermis (H&E, × 40).

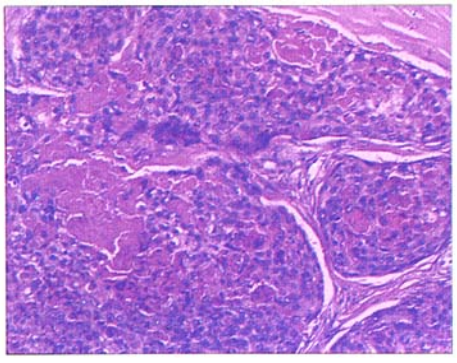

Fig.2. Part of the lesion showing relatively benign (upper) and malignant areas (lower). Note the abrupt trichilemmal keratinization, and also the dysplastic cells with marked cytologic atypia and frequent mitoses. (H&E, × 100).

An enlarged excision was taken after the patient was hospitalized followed by an examination showing that there was no remaining malignant transformation. Two weeks after the second operation, 50 cGy of regional prophylactic radiotherapy was applied.

RESULTS

After treatment with an enlarged excision and 50 cGy of regional prophylaxis radiotherapy, the patient was well within 26 months follow-up and neither recurrence nor metastases were observed.

DISCUSSION

Proliferating trichilemmal tumors (PTT) are a rare skin neoplasm derived from the outer sheath of an air follicle. They present as exophytic lesions or mutilobulated nodules, the skin may be atrophied or ulcerated and occur most commonly on the scalp. In many cases, they may cause protrusive areas of alopecia. These tumors have an incidence in women which is 2 to 5 times of that in men, and are most often noted beyond the age of 60.[1] These tumors may be found in people with multiple hereditary trichilemmal cysts, suggesting a common histogenesis of these two neoplasms. The proliferating variety is thought to develop initially as a focus of epithelial proliferation in trichilemmal cysts, perhaps as a consequence of trauma or chronic inflammation. While the age of affected individuals ranges form the 40 to 90, the mean age is about 65: The usual clinical manifestation of PTT is a long-standing, subcutaneous, cystic nodule that slowly progresses to a large (to 25 cm) nodular mass. This neoplasm is most commonly found on the scalp (90%) and the back of the neck, but it can occur on the forehead, wrist, chest, elbow, uvula,[2] nose, mons pubis, buttocks, eyelid and skin.[3]

True malignant PTT occurs in rare instances. Amaral et al.[3] hold that the diagnosis of carcinoma in PTT should be reserved for lesions showing poorly defined borders and clear-cut infiltrative properties with cytologic evidence of malignancy. Rapid growth or exophytic enlargement of long-standing nodules, and histologically extensive areas of severe dysplasia with surrounding dermal invasion are signs of malignant transformation. However, malignant transformation could be established only when there is evidence of metastasis.

Lopez-Ros, et al.[4] summarized 30 well documented cases of malignant PTT, which have been published in the English literature. There were 13 women and 7 men with ages ranging from 32 to 87 years (mean: 63.5 years). Malignant PTT arose in the scalp (18 patients) or groin (2 patients). Sizes ranged from 1.8 cm to 12 cm (mean: 5.1 cm). Lesions had been present for a period ranging from 1 year to 20 years (mean: 8.9 years). All patients underwent surgical excision. Combined radiotherapy and/or chemotherapy were also administered in eight cases. Nine tumors metastasized; five to regional lymph nodes, three to distant sites, and one to adjacent soft tissues. Four of the patients died from their disease. Follow-up ranged from 1 to 161 months (mean: 21.7 months).

Noto et al.[5] summarized 11 cases of mlignant PTT with metastases. Seven cases arose on the scalp, two on the head (ear and cheek, respectively) one on the arm, and one in the inguinal region. Among these cases two had metastesed to the local lymph nodes. The inguinal-metastasized case had lung, liver, and mediastinum metastases, which started in the cheek area and then underwent a generalized dissemination.

Histologically, PTT is made up of proliferating lobules of squamous epithelium, and some peripheral palisading is often observed. Nests of squamous cells may extend to the adjacent connective tissue, where they can simulate squamous cell carcinoma. There are multiple central areas of trichilemmal keratinization and homogenous keratin cysts. Areas of focal necrosis, calcifiration, and hyalinization may also be observed. There may be vacuolated cells, rare cellular atypia, squamaus vortices, individual cell keratinization, and scattered mitoses. Occasional cells with hyperchromatic nuclei and mitotic figures, and scattered dyskeratotic cells are not indicative of as malignant transformation. The histologic characteristics of malignant PTT are displayed as a high mitotic rate, atypical mitotic figures, nuclear pleomorphism, tumor invasion in adjacent structures, and the presence of metastatic lesions. Alternatively, some authors have proposed recently that PTT should be always considered as a low-grade carcinoma or a squamous cell carcinoma. In some cases, atypical areas are admixed with well-diferentiated areas. However, cases with little or no cytologic and architectural atypia may exhibit aggressive behavior and vice-versa. A morphometric analysis of PTT has no distinctive difference between benign and malignant PTT; instead the cell population is homogeneous in each sample, suggesting that the biological behavior of PTT is not related to its histologic appearance.

Park et al.[6]described a case of malignant PTT, with multiple distant metastases. Conventionally, the lesions were excised, and histologic specimens demonstrated a PTT with increasing nuclear atypia. After immunohistochemical staining for Ki-67, they measured the positivity in all of the six specimens to examine the degree of proliferation. They found that Ki-67 positivity related to a significantly increasing transformation tendency, which could serve to assess the extent of the tumor progression; and for each specimen, the positivity was increasing constantly.

The treatment for malignant cases is surgical excision with a clear margin. However, even with adequate resection, the tumor tends to locally recur. Because of the small number of published cases, the efficacy of alternative treatments for the malignant cases cannot be evaluated. Weiss et al. [7] reported that chemotherapy with cisplatin and fluorouracil led to a reduction in tumor size in one of their patients. Intratumoral ethanol injection in a patient exhibited a recurrent malignant PPT which was resistant to conventional treatments and surgical excision was not feasible. The biopsy specimen revealed edema, hemorrhage in the dermis, and degeneration of tumor cells. Amaral et al.[8] reported a case with incomplete excision, who underwent radiotherapy with 5,000 cGy. Autopsy findings showed no residual tumor at the primary site but there were generalized metastases. In our case, no recurrence occurred within 26 months of follow-up after surgical excision and radiotherapy.

REFERENCES

1 Mathis ED, Honningford JB, Rodriguez HE. Malignant proliferating trichilemmal tumor. Am J Clin Oncol. 2001; 24:351–353.

2 Ramesh V, Iyengar B. Proliferating trichilemmal cysts over the vulva. Cutis. 1990;45:187–189.

3 Lee, SJ, Choi, KH, Han, JH, Malignant proliferating trichilemmal tumor of the lower eyelid. The American Society of Opthalmic Plastic and Reconstructive Surgery, Inc. 2005;22:349–352.

4 Lopez–Rios F, Rodriguez–Peralto JL, Aguilar A, et al. Proliferating trichilemmal cyst with focal invasion: report of a case and a review of the literature. Am J Dermatopathol. 2000;22:183–187.

5 Noto G. 'Benign' proliferating trichilemmal tumour: does it really exist? Histopathology. 1999;35:386–387.

6 Park BS, Yang SG, Cho KH. Malignant proliferating trichilemmal tumor showing distant metastases. Am J Dermatopathol. 1997;19:536–539.

7 Weiss J, Heine M, Grimmel M, Malignant proliferating trichilemmal cyst. J Am Acad Dermatol. 1995;32:870 – 873.

8 Amaral AL, Nascimento AG, Goellner JR. Proliferating pilar (trichilemmal) cyst. Report of two cases, one with carcinomatous transformation and one with distant metastases. Arch Pathol Lab Med. 1984;108:808–810.