Case report

Proliferating Trichilemmal Tumor: Case Report

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Abstract

A proliferating pilar tumor (PPT) is a rare neoplasm arising from the isthmus region of the outer root sheath of the hair follicle. It is also commonly called a proliferating trichilemmal cyst. It was first described by Wilson-Jones as a proliferating epidermoid cyst in 1966. PPT was then distinguished from proliferating epidermoid cysts in 1995. It occurs most commonly on the scalp of middle age women. Most tumors arise within a preexisting pilar cyst. Even though they usually are benign in nature, malignant transformation with local invasion and metastasis has been described. They may be inherited in an autosomal-dominant mode, linked to chromosome 3.

Aim of study is to report case of proliferating trichilemmal tumor.

Key words proliferating trichilemmal tumor, Sequamous cell carcinoma, Proliferating trichilemmal cyst, epidermoid cyst.

Introduction

Proliferating trichilemmal tumors (PTT) also known as proliferating pilar tumor are rare neoplasm of external root of hair sheath that are largely benign , cystic in nature , and characterized as containing trichilemmal keratin.

These are exophytic tumors are mainly confined to the scalp (90%) and back of neck and most often reported in middle age female.(1)

PTT is thought to be complication of trauma, irritation, or inflammation and may be inherited in an autosomal-dominant mode linked to chromosome 3. (2, 3)

An asymptomatic nodule is often present for months to years before a rapid increase occurs in the size of the lesion, yielding lobulated and variably exophytic mass that occasionally might ulcerate. It is characterized by frequent local recurrence. (2,3,4,5,6)

Ye et al stratified these tumors into three groups:

Group 1 PPTs: Circumscribed silhouettes with "pushing" margins, mild nuclear atypia, and an absence of pathologic mitoses, necrosis, and invasion of nerves or vessels.

Group 2 PPTs: Similar to group 1 but manifested irregular, locally invasive silhouettes with involvement of the deep dermis and sub cutis.

Group 3 PTTs: Invasive growth patterns, prominent nuclear atypia, pathologic mitotic forms, and geographic necrosis, with or without involvement of nerves or vascular structures. (2)

PTT without atypia has a benign behavior, the complete removal of the lesion is recommended to prevent recurrences. Tumors with cytological atypia have unknown biological behavior and may recur locally or develop metastases.[7-14]

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30 years old female with no prior medical history had small scalp lesion 20 years ago after trauma, lesion was painless non

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tender. There is no similar case in the family.

Three years ago the lesion sometimes get worse and become bigger and painful and associated with purulent discharge. The lesion was removed surgically 2 years ago but one year after excision, the lesion recurred again. Skin examination revealed 8 x6 cm subcutaneous nodule on left post auricular region with two area of ulceration 1x1cmm. There was no lymphadenopathy.

MRI of head and neck region revealed large lobulated left post auricular complex mass with enhancing solid components, non-enhancing cystic components & amorphous calcification, no detectable underlying bony defect or intra cranial connection. Due to the high clinical suspicion for neoplasm, complete surgical excision of the mass was done with safety margin of one centimeter.

Macroscopically specimen examination is single rounded piece of tissue 7x5x3 cm with skin cover, nodular outer surface, serial section shows heterogonous area of multinodules of whitish to pink tissue of cheesy like material with greenish gelatinous material in cystic lesion, 4 pieces was taken.

Microscopically section examination showed diffuse proliferation of solid nests of mature sequamous epithelium with prominent pilar type keratinization, prominent cellular atypia and scattered mitosis, with pushing capsulated borders, excisional deep margin show peripheral thin capsulation with no extra capsular tumor component, resection margins were free from tumor, picture consistent with proliferating pilar tumor group 3 according Yet et al classification (figure 1).

Immunohistochemistry analysis revealed CD34 negative which indicate no lymphovascular invasion (figure 2). Ki67 immunostaining revealed immunore-activity with staining of about 10-15% of the nuclei, P53 immunostaining was negative (figure 3).

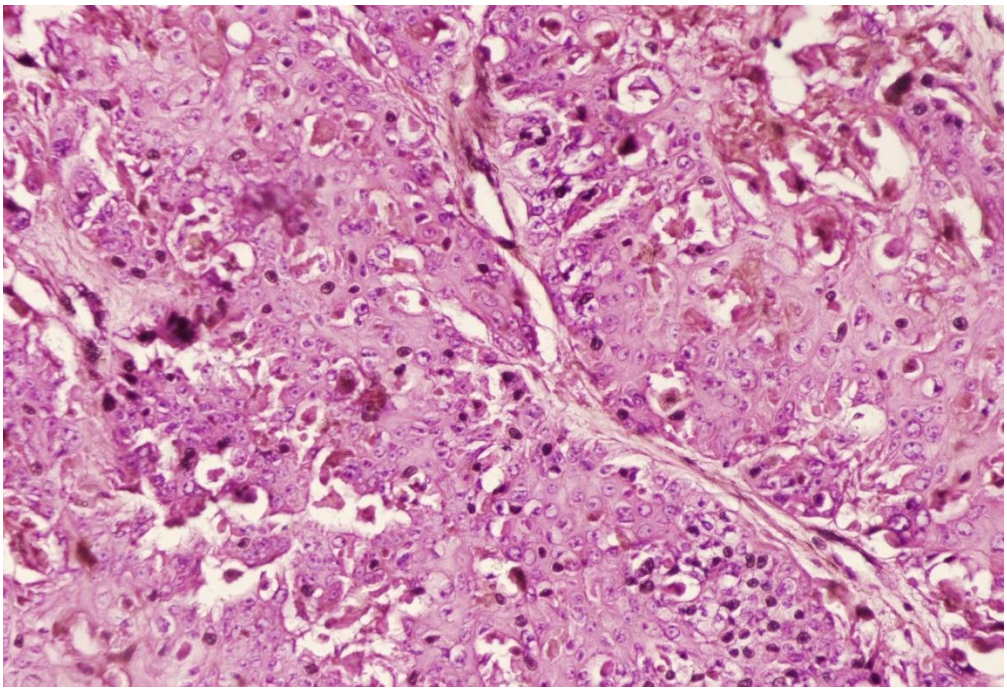

Figure1. Solid nests and bands of squamous epithelium with prominent atypia and trichilemmal type keratinization (H&E, 200X)

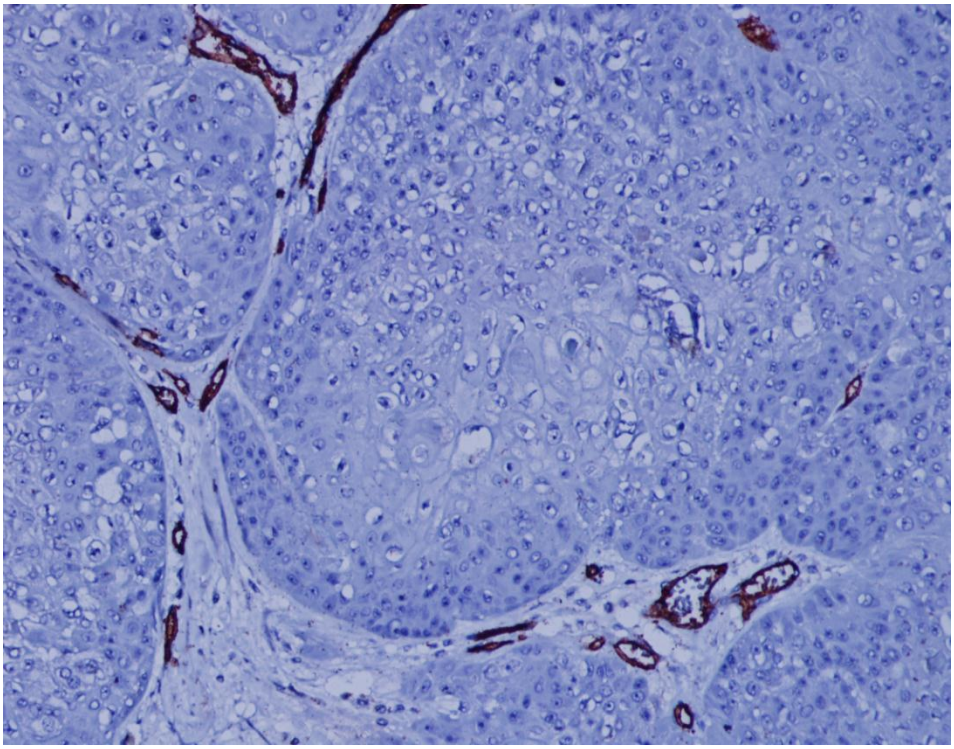

Figure 2. CD34 Immunhistochemical stain, shows no vascular invasion, 100X

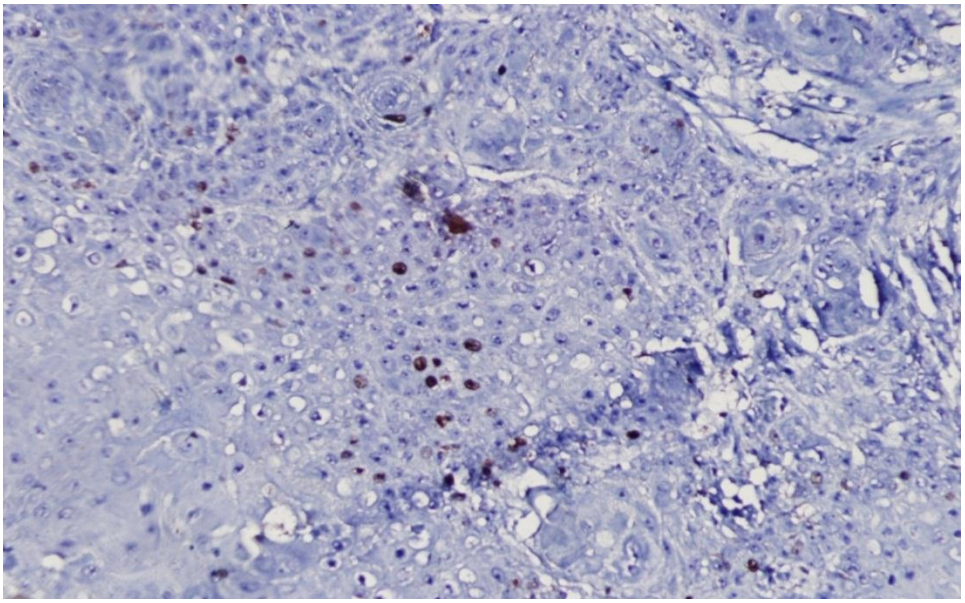

Figure 3. Ki67 Immunhistochemical stain shows significant proliferative index about 10 - 15 %, 200X

Discussion

PPT arises principally on the scalp and it has a clinical resemblance to keratinous or sebaceous cysts and have been commonly misinterpreted both clinically and histologically as squamous cell carcinomas. (15-19)

In most cases, PTT treated by resection with free surgical margins. (5-7) There are reports of PTT with aggressive clinical behavior with recurrence and/or metastasis. The more aggressive behavior is more common in tumors located out of the scalp, when it is fast-growing and infiltrative, tumors larger than 5 cm, presence of atypia and high mitotic activity, of note the recurrence does not mean malignant growth.(2,5,10-14)

The differentiation between a benign or malignant process in these lesions can sometimes be difficult. Careful clinical and histological assessment is necessary for an accurate diagnosis. (4)

Mohs micrographic surgery, which ensures precise lesion margin control, can be performed to reduce the recurrence rate after tumor resection. (3)

The role of adjuvant radiation therapy in pilar tumor, especially in malignant variant, is not very clear. This is mainly due to the rarity of the disease .Unlike head and neck squamous cell carcinomas which are locoregional pilar tumor are local only. (20)

For these reasons and because our patient had cytological atypia we chose complete surgical resection and strict follow up in every 3-12 months for two years, then every 6-12 months for three years and annually for life as the follow up plan of skin sequamous cell carcinoma. (21)

Conclusions

PPT is a rare neoplasm, it is more often benign and very rarely malignant. Since PPT has potential for malignant transformation, therefore every cystic mass of the scalp should be excised and must be subjected to histopathological examination.

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