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Case Report

Proliferating Trichelemmal Tumor - A Rare Case Report with Review of Literature

Archana Bidari[1], Khan Talat[2], Smita Mulay[3], Bhale CP[4]

[1]Lecturer, [2]Senior Resident, [3]Head of Department, [4]Professor,
Dept. of Pathology, MGM Medical College, Aurangabad, Maharashtra.

Corresponding Author: Khan Talat

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ABSTRACT

Proliferating trichelemmal tumors (PTT) are uncommon benign tumor originating from outer root sheet of the hair follicle. The histologic hallmark is the presence of trichelemmal keratinization/ abrupt keratinization. PTT is thought to originate from the Trichelemmal cyst (TC) and have the potential for malignant transformation at which point it is termed to be Malignant Proliferating Trichelemmal Tumor (MPTT). It is usually confused with Squamous Cell Carcinoma both sharing many common features. These lesions may cause considerable morbidity and even mortality and recurrence after simple local excision in common. We hereby present a rare case of a 50 old male patient with a rapidly growing swelling of the scalp on the occipital region which was ulcerating and foul smelling discharge was present. A wide excision was done and histopathological examination revealed proliferating trichelemmal tumor which on history and gross appearance was mimicking malignant tumor.

Keywords: Proliferating trichelemmal tumor, Squamous cell carcinoma, Trichelemmal cyst, Malignant proliferating trichelemmal tumor.

INTRODUCTION

Proliferating trichelemmal tumors are rare benign neoplasm of follicular lineage whose histological hallmark is presence of trichelemmal keratinization[(1)] It was first described by Wilson-Jones as a proliferating epidermoid cyst in 1966.[(2)] PTT was distinguished from proliferating epidermoid cyst in 1995. These lesions have considerable morbidity and mortality. PTT is thought to originate from trichelemmal cyst and has the potential for malignant transformation, known as malignant trichelemmal tumor (MPTT).[(1)] Recurrence after simple excision is common. The most common site for occurrence is the scalp in women older than 50 years.[(3,5)] The tendency for malignant transformation increases especially with the presence of infection, ulceration and rapid enlargement of long standing nodular scalp lesion together with histological evidence of significant abnormal mitosis, marked cellular pleomorphism, infiltrating margins and aneuploidy.[(4)] Diagnosis require proper histological examination, taking into consideration that such features may not correlate with clinical behavior. MPTT may

mimic more aggressive tumors with local recurrence and distant metastasis. Thus wide local excision of the lesion and long term follow-up is recommended.

CASE REPORT

Here we present a 65 yr old male with one year history of exophytic growth, painless mass in the occipital area. It was initially growing slowly and later on started to ulcerate with foul smelling discharge and was rapidly growing that interfered with the patient's daily life. That was the reason for surgical consultation. No past history of trauma and chronic irritation was noted. The patient was otherwise healthy with no other complains or symptoms. The non contrast-enhanced computed tomography scan of the head was performed to evaluate the underlying extent which revealed a soft tissue swelling within the subcutaneous scalp tissue with no bone involvement (Fig.1). FNA of the swelling was done. Pap stained smear showed clumps of squamoid cell cluster in a necrotic background (Fig.2). A diagnosis suggestive of benign squamous cell lesion was offered on cytology and excision was advised by the surgeon.

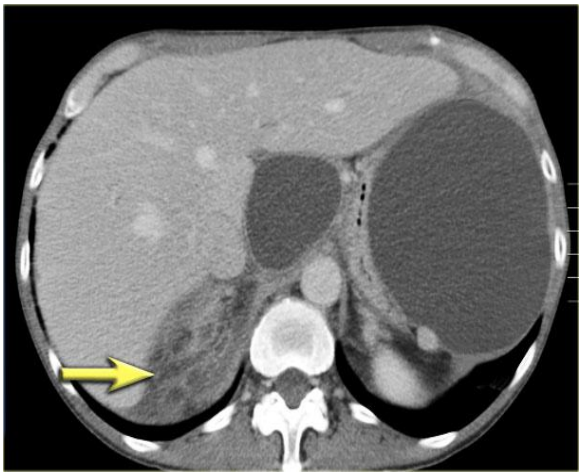

Figure 1: Non Contrast enhanced CT scan revealed a soft tissue swelling within the subcutaneous scalp.

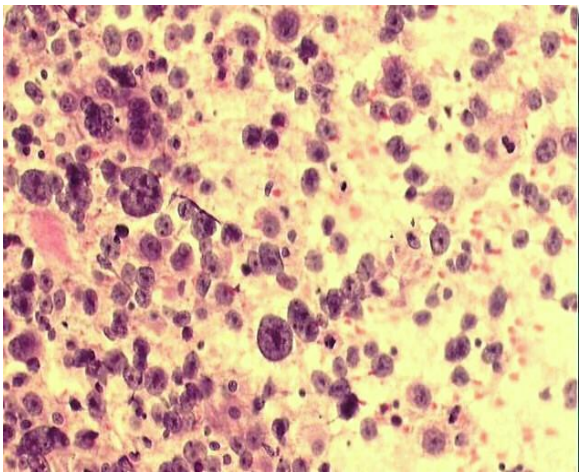

Figure 2: FNA smears studies show groups of squamous cells.(Pap x 100).

Figure 3(a): Gross; lobulated subcutaneous mass that measured 9x8x5cm.

Figure 3(b): Cut section; grayish white homogeneous solid areas with few cystic spaces.

On Gross examination it was skin covered, well demarcated, lobulated subcutaneous mass that measured 9x8x5cm (Fig.3a). Cut surface of the mass revealed grayish white homogeneous, solid with few cystic spaces (Fig 3b). On histopathological examination H & E stained sections showed the tumor was well demarcated from the surrounding tissue. The mid dermis, deeper dermis and subcutaneous tissue showed a cellular tumor comprised of lobules of squamous cells(Fig 4). The centre of lobules were filled with keratinous material derived from abrupt keratinization of large polygonal cells with abundant pale eosinophilic cytoplasm without any intervening granular cell layer (trichelemmal type) (Fig 5 a& b). The cells showed normal to mild pleomorphism, low mitotic activity without infiltrating margins (Fig 6). Hence the diagnosis of proliferating trichelemmal tumor was done. The clinical behavior was mimicking malignant tumor (ulceration and foul smelling discharge) probably due to its location in occipital region causing pressure effect.

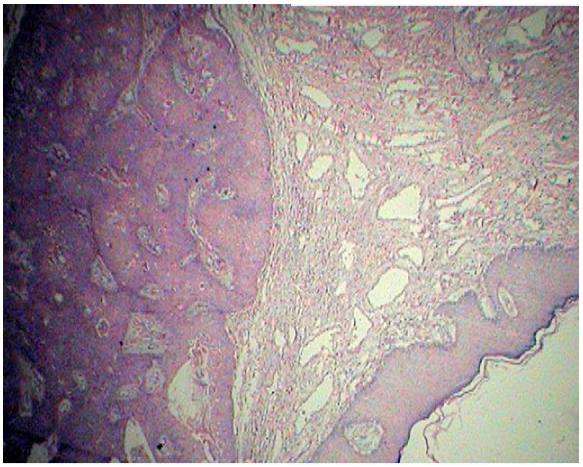

Figure 4: Dermis showing cellular tumor comprised of lobules of squamous cells.(H & E stain, X 50).

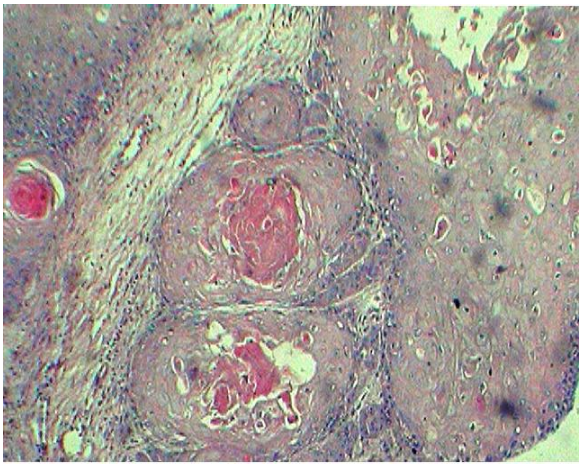

Figure 5(a): Centre of lobules filled with keratinous material without any intervening granular layer. H & E X 100.

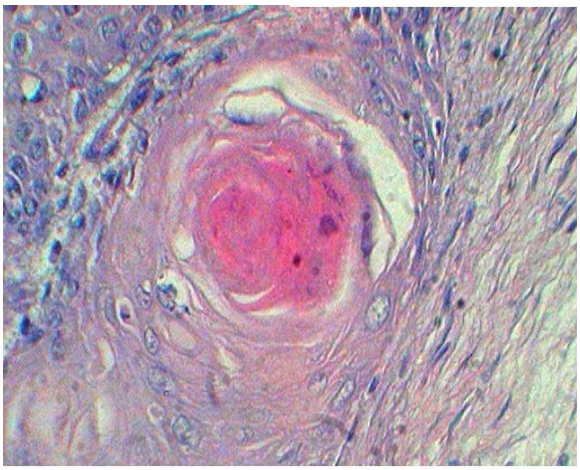

Figure 5(b): Centre of lobules filled with keratinous material without any intervening granular layer. H & E X 400.

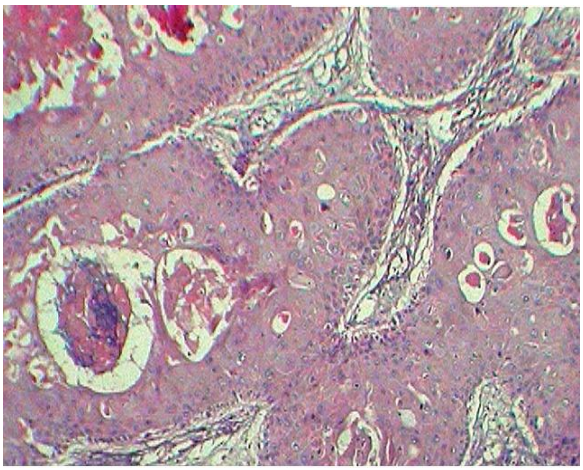

Fig. 6: Cells show normal to mild pleomorphism with low mitotic activity. H & E X 100.

DISCUSSION

Proliferating trichelemmal tumor is a solid cystic neoplasm that shows trichelemmal differentiation similar to that of the isthmus of the hair follicle.[6] These lesions include trichelemmal cyst, proliferating trichelemmal tumor and malignant proliferating trichelemmal tumor. PTT is an uncommon benign lesion which may be misdiagnosed as squamous cell carcinoma. It is also known as proliferating trichelemmal cyst and pilar cyst of the scalp.[7] Ninety percent of the PTT's occur on the scalp but they are also found on forehead , nose, back , chest, abdomen, buttocks, elbow, wrist, mons pubis and vulva.[8,9] Most patients are women (84%) who range in age from 27 to 83 years. Most cases occur in 6^{th} and 7^{th} decade of life.[7] In our case the patient was male, had a dormant swelling for one year, the swelling enhanced at a much rapid pace with ulceration and foul smelling discharge mimicking malignancy but histologicaly showed features of benign proliferating trichelemmal tumor.

Malignant PTT {MPTT} is an uncommon cutaneous neoplasm and this term MPTT was entered in the literature by Saida et al, because of a PTT that showed infiltrative growth marked cytological atypia, high mitotic activity including atypical forms, and lymph node metastasis.[10,11] Malignant transformation occurs occasionally which can be manifested by sudden rapid growth ulceration and foul smelling discharge. Histologicaly malignant PTT's show severe nuclear atypia marked cellular pleomorphism with atypical mitosis, dyskeratotic cells and infiltrating margins.[9,12] Heddington[13] proposed the term malignant proliferating trichelemmal cyst for the Proliferating trichelemmal cyst with malignant transformation. The real incidence of a malignant proliferating trichelemmal cyst is unknown due to its rarity and also due to inconsistency in nomenclature and misclassification as squamous cell carcinoma. Unfortunately distinctive histological and immunohistochemical markers of malignancy do not exist. Some recent reports have shown DNA aneuploidy and in some cases an increased proliferation index, suggesting that PTT may be a premalignant tumor.[14] Immunoprofile of PTT expresses fetal hair root cytokeratin as well as cytokeratin 7. The pathogenesis remains unknown. In some cases human papilloma virus has been implicated as the etiology.[6] The therapeutic approach in Proliferating trichelemmal tumor is same as other malignant skin lesions. Adequate surgical excision remains mainstay of the treatment. The patient should be followed closely after surgery. The efficacy for alternative treatment for these cases cannot be safely evaluated in view of a very small number of published cases.

SUMMARY

We present a rare case of PTT in a 50 year old male patient with complains of dormant swelling of one year, the swelling enhanced at a much rapid pace with ulceration and foul smelling discharge mimicking malignancy. Histologicaly on H&E section showed a cellular tumor composed of lobules of benign squamous cells. The centre of lobules filled with keratinous material derived from abrupt keratinization without any intervening granular cell layer. No features of malignancy were observed. A diagnosis of PTT was offered.

CONCLUSION

Proliferating trichelemmal tumors (PTT) are rare and pose a diagnostic dilemma for the pathologist. It has also been stated that the tumor has tendency to recur

and malignant transformation may occur. Wide surgical excision should be considered as a primary modality of treatment while alternative therapy is required for further evaluation.

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Conflicts of interest: None.

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