

HISTOLOGICAL MIMICKER OF CUTANEOUS SQUAMOUS CELL CARCINOMA – MALIGNANT PROLIFERATING TRICHILEMMAL TUMOR: CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Proliferating trichilemmal tumors (PTT) are benign solid-cystic neoplasms that show trichilemmal differentiation similar to that of isthmus of hair follicle. It comprises only 0.1% of skin biopsies. These tumors rarely exhibit malignant transformation. They develop from the wall of already existing pilar cyst in most cases. It is frequently seen in elderly women, more commonly on the scalp. Other locations could include face, trunk, back and forehead. These tumors are easily confused with squamous cell carcinomas. Malignant proliferating trichilemmal tumors carry significant morbidity and mortality, with possible recurrence after local excision. We present a case of a 65 year old female with rapidly growing swelling on the scalp, clinically suspected to be sebaceous cyst but later diagnosed with malignant proliferating trichilemmal tumor on histopathological examination. We also discuss the review of literature of this tumor considering the rarity of presentation.

KEYWORDS: trichilemmal, malignant, squamous cell carcinoma, pilar cyst.

BACKGROUND

In 1966, Wilson-Jones used the term “proliferating epidermoid cyst” to describe the benign “proliferating trichilemmal tumor (PTT)”^[1] In 1995, PTT and proliferating epidermoid cysts were distinguished as separate entities. They are uncommon cutaneous tumors which are derived from outer root sheath of the hair follicle.^[1] They occur most commonly in females after the sixth decade, more often on the scalp. These tumors have potential for malignant transformation, when they are called malignant proliferating trichilemmal tumors (MPTT) and even metastatic spread if not diagnosed and treated on time. As it is rare, they are misdiagnosed as squamous cell carcinomas or sebaceous cyst.^[2] Clinically, the classical presentation is a rapidly growing mass with necrosis of underlying tissue. On microscopic examination, MPTTs show abrupt keratinization without granular layer, peripheral palisading of small basaloid cells which differentiate into large pleomorphic keratinocytes.^[3] These tumors are treated with wide surgical excision, meticulous follow up and chemotherapy is indicated in case of metastases.^[3] Due to its aggressive biological nature, it is crucial for clinicians and histopathologists to be aware of this rare lesion to distinguish it from primary squamous cell carcinoma of the skin.^[4]

CASE REPORT

A 65 year old female presented to the surgical OPD with a swelling on the parietal region of scalp for the past 3 years. She observed a recent increase in size of the swelling. She was a diabetic for the past ten years on treatment for the same. She had no history of trauma or ulceration on the lesion. On examination, 4x3 cm nodular lesion was noted in the right parietal region, not fixed to underlying bone. No regional lymphadenopathy was noted. Clinical diagnosis of sebaceous cyst was done and simple excision was done. We received a well demarcated lobulated mass of size 4x4 cm (FIG-1). Outer surface was smooth with occasional brown areas. Cut section showed grey-tan homogenous appearance with some areas of hemorrhage (FIG-2). On histopathological examination, a well circumscribed cellular neoplasm involving the dermis and subcutaneous tissue with lobules of large polygonal pleomorphic squamous cells (FIG-3) with abundant eosinophilic cytoplasm and no intervening granular layer was noted. The centre of the lobules showed keratinous material (FIG-4). Focal invasion into surrounding tissue was noted. The histopathologic diagnosis of malignant proliferating trichilemmal tumor was made. Wide surgical excision was performed later which confirmed the diagnosis and resection margins were free of the tumor. The patient has been followed up for the past twelve months with no recurrence.



Fig. 1: Gross picture of the globular mass.

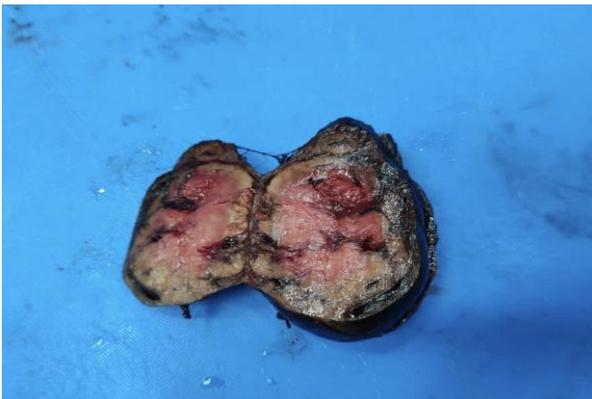


Fig. 2: Cut-Surface of The Mass.

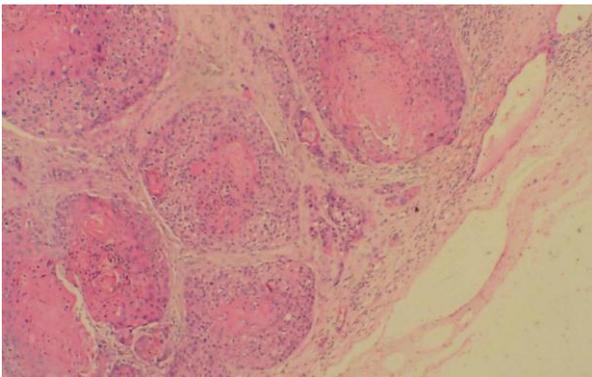


Fig. 3: Low Power View Showing Lobules of Squamous Cells.

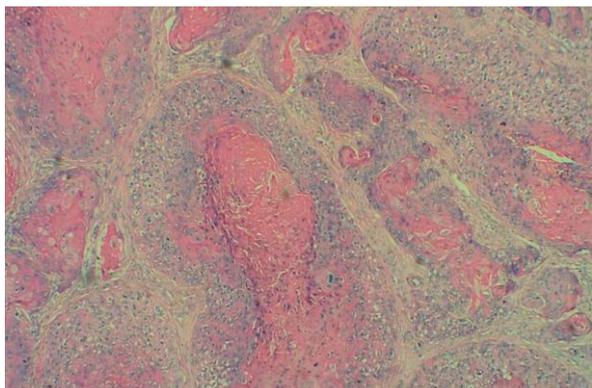


Fig. 4: Low Power View.

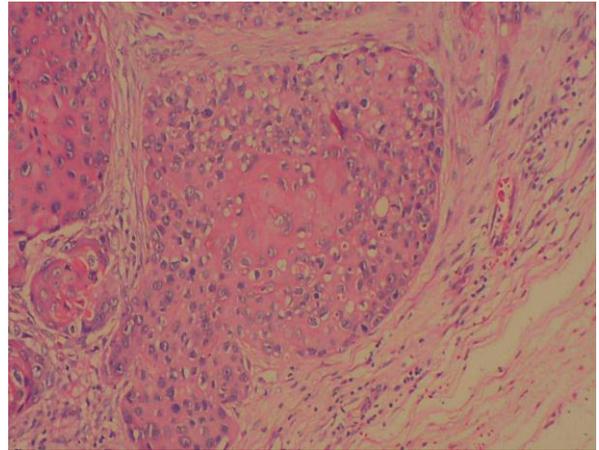


FIG. 5: Lobules of Squamous Cells In High Power.

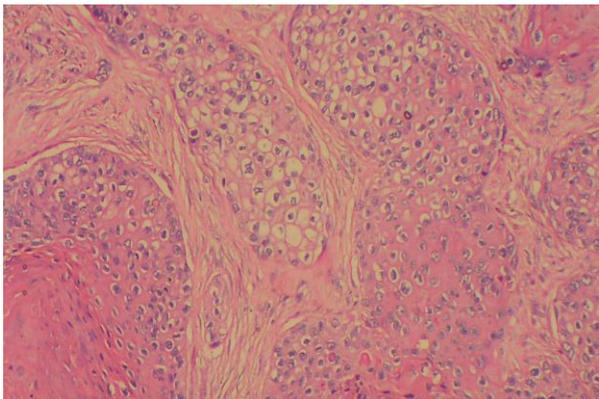


Fig. 6: Pleomorphic Squamous Cells.

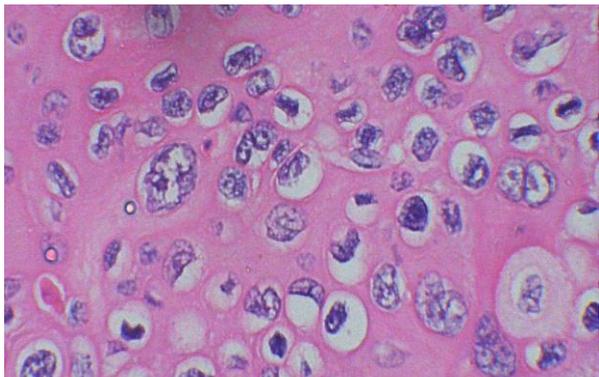


Fig. 7: High Power View of Pleomorphic Cells.

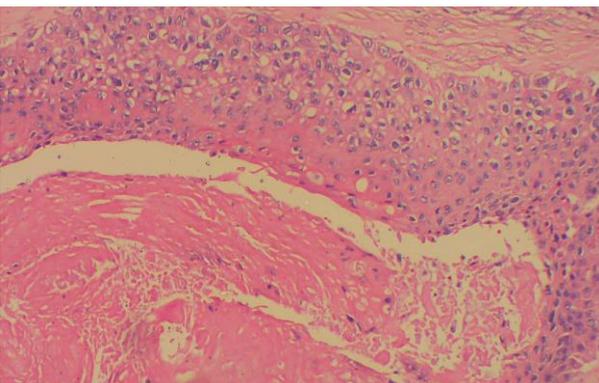


Fig. 8: Central Keratinous Material.

DISCUSSION

Malignant proliferating trichilemmal tumors (MPTT) are extremely rare cutaneous adnexal neoplasms with very few cases reported in literature. The term "malignant proliferating trichilemmal tumor" was suggested by Headington to distinguish it from trichilemmal carcinoma. In 1983, Saida et al reported a patient with multiple trichilemmal cysts and malignant transformation observed in one of them and reported the first case of MPTT.^[5] She also postulated that these tumors arise from lanugo hair follicles of bald scalp and follicles devoid of nonterminal hair.^[5] The biological nature of these tumors remain controversial. The malignant transformation from a benign proliferating trichilemmal cyst goes through adenomatous to epitheliomatous (PTT) to carcinomatous (MPTT) stage. Sudden increase in size of a long standing swelling, presence of nuclear atypia and invasion into surrounding tissues could represent malignant transformation.^[6] The real incidence of MPTT is not known due to different names that the tumor carries like giant hair matrix tumor, invasive pilomatrixoma, pilar tumor of the scalp and proliferating follicular cystic neoplasm.^[7]

Ye et al, did a study on 76 cases and classified PPTs into group 1 containing tumors with pushing margins and absence of mitosis, necrosis and invasion; group 2 PPTs with local invasion into deeper dermis and subcutis; group 3 PPTs with invasive growth, cytologic atypia, mitosis, necrosis and involvement of vascular structures. He concluded group 1 as benign, group 2 as tumors with local aggressive growth and group 3 with malignant potential.^[8]

In his studies on imaging findings in PTTs by Kim et al, it was given that PTT may be cystic or solid. When cystic, it may look benign on CT scans and when solid, the imaging intensities on MR may resemble other soft tissue tumors. Smooth soft tissue elevations from inner wall of the cystic mass may correspond histologically to proliferating lobules of epithelium, typical of MPTT.^[9]

Martinez et al, in his case study has mentioned that histological picture of MPTT may be similar to squamous cell carcinoma (SCC) and clinical differentiation is necessary due to different treatment options. SCC has a structured treatment format while MPTT still remains unpredictable. MPTT shows clear demarcation from surrounding stroma, is exophytic usually arising from a pilar cyst unlike SCC which arises from a precursor lesion such as actinic keratosis.^[10] There are no IHC markers for diagnosis of MPTT but CD34 and Ki-67 may show tumor behavior.

Agarwal et al postulated that IHC may play a role in differentiating between SCC and malignant PTT. Proliferation markers like Ki-67 and P53 are high in both the tumors. CD34 and calretinin are markers favouring trichilemmal differentiation. It was found that most PPT

showed positive staining for AE13 and AE14 but SCC showed no staining.^[11]

Gallant et al, did an extensive study on all possible cases of metastatic MPTT and identified 42 mutations with PIK3CA H1047R as the only known driver mutation. As there is less information about nonsurgical management of MPTT, it may be responsive to chemotherapy regimens and a small molecule inhibitor of PI3K, in the setting of a somatic PIK3CA activating mutation. The identification of a mutation may help learn the biology of the tumor and increase non-surgical options for the same.^[12]

There is no established treatment plan for MPTT but wide surgical excision with 1 cm margins is being followed. Radiotherapy may be needed for more localized tumors. Ethanol injections may be useful for local recurrence. Chemotherapy regimens including cisplatin, doxorubicin and vinca alkaloids may be used for metastatic disease.

CONCLUSION

MPTTs are rare skin adnexal neoplasms which can present as rapid enlargement of a already existent mass, most commonly seen on the scalp of elderly women. They can clinically resemble benign lesions and on microscopic examination be confused with squamous cell carcinomas. As these tumors are uncommon and not extensively studied, adequate clinical evaluation along with histopathological examination and use of IHC may be needed for adequate diagnosis. A regular follow up may be needed considering the local recurrent nature and metastatic potential of these tumors. Wide surgical resection, Mohs surgery, radiation therapy and chemotherapy are all reported in the management of MPTT.

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