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# The Follicular Propagation- Malignant Proliferating Trichilemmal Tumour

**Anubha Bajaj***

Consultant Histopathologist, AB Diagnostics, India

***Corresponding author:** Bajaj A, Consultant Histopathologist, AB Diagnostics, New Delhi, India, Tel: +919811693956; E-mail: anubha.bajaj@gmail.com

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## 1. Introduction

Cutaneous adnexal tumours derived from outer root sheath of hair follicles demonstrating a trichilemmal variety of keratinzation commonly manifest as trichilemmal cysts, proliferating trichilemmal cysts and malignant proliferating trichilemmal tumours. Proliferating trichilemmal tumour (PTT) is enunciated as an exceptional, benign, cutaneous adnexal neoplasm originating from outer root sheath of the hair follicle and is additionally nomenclated as proliferating trichilemmal cyst or pilar tumour of the scalp. Malignant proliferating trichilemmal tumour was initially described by Wilson Jones as a "proliferating epidermoid cyst" in 1966 and further segregated from proliferating epidermoid cyst in 1995 [1]. Although contemplated as a biologically benign lesion, proliferating trichilemmal tumour can emerge as an aggressive, locally infiltrative tumefaction, exceptionally delineating malignant transformation as evidenced with concurring regional or distant metastasis. Terminology of malignant proliferating trichilemmal cyst for describing a proliferating trichilemmal cyst exemplifying malignant transformation was proposed by Headington [1,2].

## 2. Disease Characteristics

Malignant proliferating trichilemmal tumour is an infrequently observed category of trichilemmal tumours. As an uncommon cutaneous neoplasm, subject to a controversial histogenesis, malignant proliferating trichilemmal tumour is nomenclated by Saida et al [3] on account definitive features delineated by proliferating trichilemmal tumour such as infiltrative growth pattern, significant cytological atypia, elevated mitotic activity, especially atypical mitosis and lymph node metastasis. Nevertheless, proliferating trichilemmal tumour usually exhibits a benign behaviour and a malignant course of disease is rarely evident [3].

Proliferating trichilemmal tumour as an uncommon, benign cutaneous adnexal lesion can be misdiagnosed as squamous cell carcinoma. Malignant metamorphoses of infrequently cogitated proliferating trichilemmal tumour is generally misinterpreted as squamous cell carcinoma on account of identical clinical and morphological features, thus, appropriate categorization is crucial and challenging. Proliferating trichilemmal tumour, as a diagnostic dilemma, frequently depicts a tendency for tumour reoccurrence and distant metastasis, in contrast to a squamous cell carcinoma [3,4].

Malignant proliferating trichilemmal tumour commonly appears within the elderly population exceeding 50 years and depicts a female preponderance. Majority (84%) of afflicted subjects are females betwixt 27 years to 83 years with a significant proportion of instances emerging betwixt sixth to seventh decade [3,4].

Incidence of malignant proliferating trichilemmal tumour is undetermined on account of the exceptional nature, enunciated misclassification as a squamous cell carcinoma and a lack of distinctive histological features or cogent immune markers exemplifying malignant metamorphoses [3].

## 3. Clinical Elucidation

Cogent discernment of malignant proliferating trichilemmal tumour is mandated from adjunctive, identical neoplasms on account of an aggressive clinical course. An estimated 90% of proliferating trichilemmal tumours occur upon the scalp although sites such as forehead, nose, neck, trunk, chest, abdomen, buttocks, elbow, wrist, mons pubis and vulva can be incriminated [4].

Malignant proliferating trichilemmal tumour usually emerges as a solitary, non-tender, painless, fixed, firm, nodular lesion of variable magnitude with an unremarkable cutaneous covering, situated upon the scalp. Nodules persist for a significant duration and are followed by brisk enhancement. Regional lymph nodes are usually non palpable. Malignant metamorphoses is occasional and exemplifies as an abrupt, rapid tumour evolution. Biologically aggressive, invasive and metastatic trichilemmal tumours depicting infiltration of circumscribing soft tissues in association with features of anaplasia and tumour necrosis are a manifestation of malignant proliferating trichilemmal tumour [4,5]. In contrast, proliferating trichilemmal tumour demonstrates an indolent clinical course with a systematic progression from an adenomatous stage towards an epitheliomatous stage with further evolution into a carcinomatous stage of malignant proliferating trichilemmal tumour. Although metamorphoses is exceptional, malignant proliferating trichilemmal tumour commonly engenders from a previously discerned trichilemmal cyst arising in young individuals [4,5].

## 4. Histological Elucidation

Gross examination exhibits a well-defined, lobulated, subcutaneous nodule of varying magnitude. Further dissection of the nodule demonstrates a well circumscribed lesion with a grey/ tan, homogenous appearance. Tumefaction can be exophytic, ulcerative, polypoid, nodular or the lesions can be keratotic [5]. On histological examination, the neoplasm is well- demarcated and distinct from adjoining soft tissues. A characteristic histological feature is an abrupt, compact, amorphous keratinization of epithelial cells enveloping the cyst wall in the absence of a granular cell layer, a phenomenon cogitated as trichilemmal type of keratinization.

Mid- dermis, deep-seated dermis or subcutaneous tissue exhibits a cellular tumour comprised of lobules of squamous epithelial cells [5]. Lobular centres are impacted with keratinous substance originating from abrupt keratinization of enlarged, polygonal epithelial cells impacted with abundant quantities of pale-staining or eosinophilic cytoplasm associated with an absence of intervening granular cell layer (trichilemmal subtype of keratinization). Tumour cells depict moderate to significant cellular pleomorphism, severe nuclear atypia, dyskeratotic cells, enhanced mitotic activity with the enunciation of atypical mitotic figures, configuration of tumour giant cells and definitive foci of tumour infiltration within encompassing soft tissue [5,6].

Rapid augmentation of a dormant nodule is indicative of malignant metamorphosis as described by an infiltrative tumour perimeter, nuclear atypia, mitosis and aneuploidy. Fine needle aspiration delineates clusters and aggregates of dysplastic squamoid cells disseminated within abundant amount of necrotic material. The features can be misinterpreted as a squamous cell carcinoma [6].

## 5. Differential Diagnosis

Malignant proliferating trichilemmal tumour requires a segregation from neoplasms such as basal cell carcinoma, sebaceous carcinoma, squamous cell carcinoma and clear cell hidradenocarcinoma. Clinical and histological differentiation of malignant proliferating trichilemmal tumour from a squamous cell carcinoma is necessitated on account of an identical disease representation [6,7].

## 6. Investigative Assay

DNA aneuploidy can be assessed in certain instances and an elevated proliferation index indicates a proliferating trichilemmal tumour to be a premalignant neoplasm. Magnetic resonance imaging (MRI) of a proliferating trichilemmal tumour displays an elliptical soft tissue mass with an inadequately defined tumour margin. In contrast to muscle tissue, the nodule depicts isointense signals on T1 weighted imaging and hyper-intense signals on T2 weighted imaging [7,8]. Contrast enhanced computerized tomography (CT) scan delineates an enlarged, well encapsulated, cystic nodule confined to the subcutaneous adipose tissue. Variably thickened, well defined perimeter of the tumefaction delineates multiple foci of calcification. Several foci of smooth, soft tissue elevations arising from inner wall of the nodule can be cogitated [7,8].

## 7. Therapeutic Options

Therapeutic approach of managing malignant proliferating trichilemmal tumour recapitulates the management of adjunctive malignant cutaneous lesions. A comprehensive and immediate surgical extermination of the neoplasm is the preferred treatment modality [7,8]. Comprehensive surgical excision with a broad perimeter of uninvolved tissue is recommended as a primary modality of treatment, even as alternative strategies of treatment require additional evaluation. Adjuvant chemotherapy or radiotherapy is unnecessary [7]. Extensive, periodic surveillance is necessitated following a surgical extermination, in order to exclude tumour reoccurrence. Nevertheless, therapeutic strategies for managing a malignant proliferating trichilemmal tumour are debatable on account of the exceptional nature of the condition and require further evaluation [8].

Malignant proliferating trichilemmal tumour with metastasis is appropriately treated by comprehensive surgical eradication with one centimeter broad margin of uninvolved tissue. Exceptional instances of malignant proliferating trichilemmal tumour with intracranial extension and/or pulmonary metastasis are associated with a significantly inferior prognosis. Palliative chemotherapy with administration of cisplatin and 5- fluorouracil with subsequent palliative radiotherapy is the recommended mode of therapy for managing a malignant proliferating trichilemmal tumour with distant metastasis [8,9].

As malignant proliferating trichilemmal tumour delineates an aggressive clinical and biological behaviour, the neoplasm is accompanied by a significant morbidity and potential mortality [9].

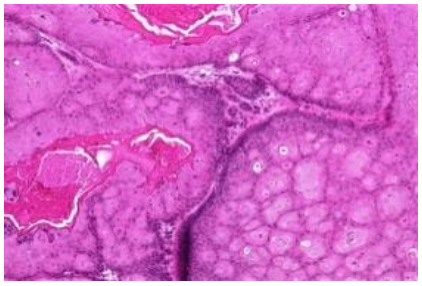

FIG. 1. **Malignant proliferating trichilemmal tumour with well circumscribed tumour lobules with abrupt, centric keratinization and aggregates of pale-staining squamous epithelial cells [10].**

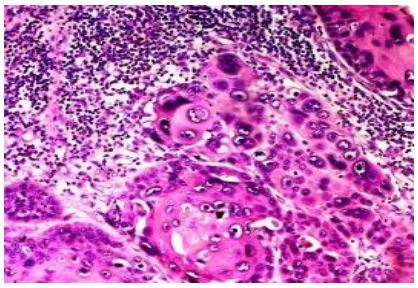

FIG. 2. **Malignant proliferating trichilemmal tumour delineating nests of atypical squamous epithelial cells with cellular pleomorphism, nuclear atypia, aberrant mitosis and abrupt keratinization [11].**

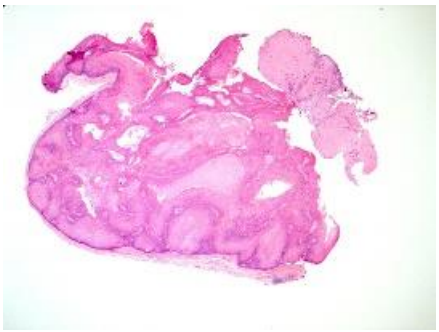

FIG. 3. **Malignant proliferating trichilemmal tumour exhibiting nodules of atypical squamous epithelial cells, centric, abrupt, trichilemmal type of keratinization and adequate circumscription [12].**

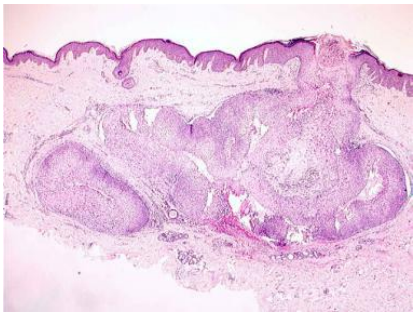

FIG.4. **Malignant proliferating trichilemmal tumour demonstrating a mid-dermal lesion with lobules and nests of aberrant squamous epithelial cells, centroidal keratinization and circumscription of cellular aggregates from adjacent tissue [13].**

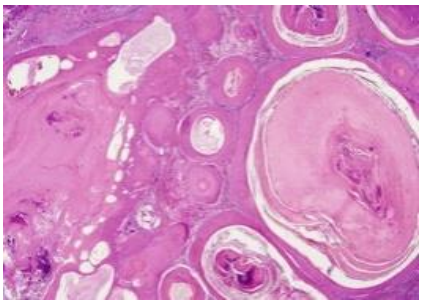

FIG. 5. **Malignant proliferating trichilemmal tumour exemplifying centric keratinization, aggregates of atypical squamous epithelial cells with abundant eosinophilic cytoplasm, cellular pleomorphism and nuclear atypia [14].**

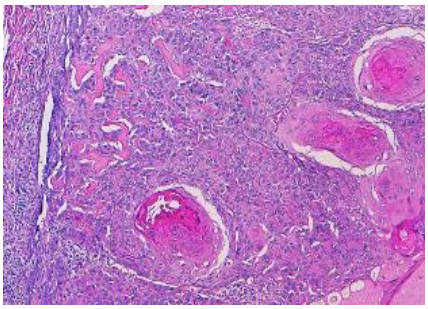

FIG. 6. **Malignant proliferating trichilemmal tumour depicting foci of centric keratinization, nests and clusters of atypical squamous epithelial cells with anisocytosis and aberrant mitosis [15].**

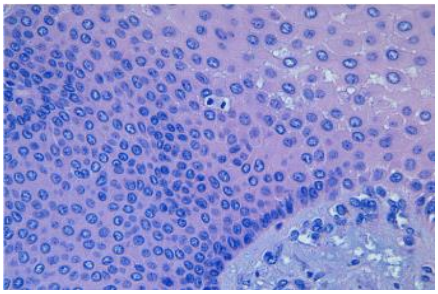

FIG. 7. **Malignant proliferating trichilemmal tumour exhibiting atypical squamous epithelial cells with nuclear and cellular pleomorphism, anisonucleosis, enhanced mitosis and aneuploidy [16].**

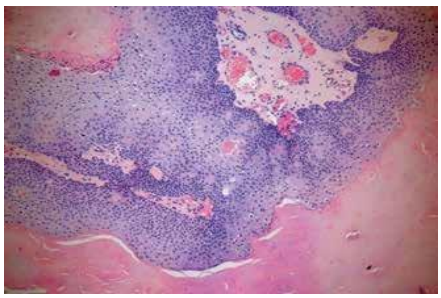

FIG. 7 **Malignant proliferating trichilemmal tumour with lobules of atypical squamous epithelial cells, central zones of keratinization and an encompassing fibrous tissue stroma**[17]**.**

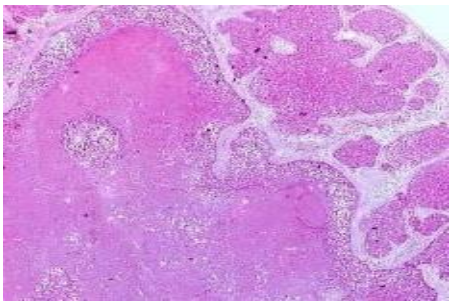

FIG.9 **Malignant proliferating trichilemmal tumour depicting accumulations of atypical squamous epithelial cells, centrally aggregated keratin and cellular pleomorphism[18].**

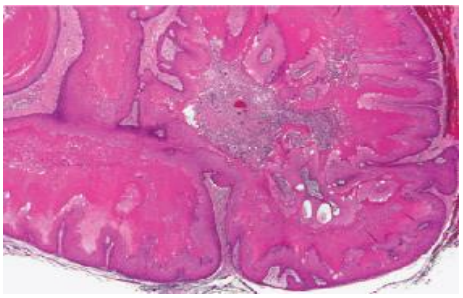

FIG. 8 **Malignant proliferating trichilemmal tumour enunciating aggregates of atypical squamous epithelial cells with severe nuclear atypia, mitosis, cellular pleomorphism and centrally accumulated keratin**[19]**.**

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