Multifocal Malignant Proliferating Trichilemmal Tumour: A Diagnostic Imitator Beyond the Scalp

Dr. Manika Alexander, Dr. Divyashree, Dr. B. P Bommanahalli

K H Patil Institute of medical sciences Gagag

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ABSTRACT

Malignant proliferating trichilemmal tumour (MPTT) is a rare cutaneous adnexal neoplasm arising from the outer root sheath of the hair follicle and represents malignant transformation of a proliferating trichilemmal tumour. It accounts for less than 0.1% of cutaneous malignancies and most commonly presents as a solitary scalp lesion in elderly women.

A 60-year-old male presented with progressively enlarging painful swellings over the scalp and left scapular region. Histopathological examination revealed a dermal epithelial tumour showing marked cytological atypia with abrupt trichilemmal keratinization. Periodic acid–Schiff staining demonstrated diastase-resistant cytoplasmic positivity. Immunohistochemistry showed focal CD34 expression with strong nuclear p53 and increased Ki-67 labeling index.

The findings were diagnostic of malignant proliferating trichilemmal tumour. Multifocal involvement occurring in a male patient is exceedingly rare. Recognition of trichilemmal keratinization and the use of histochemical and immunohistochemical markers are essential to distinguish MPTT from squamous cell carcinoma and guide appropriate management.

Keywords: Malignant proliferating trichilemmal tumour, scalp, scapular lesion, adnexal tumour.

INTRODUCTION

Malignant proliferating trichilemmal tumour (MPTT) is a rare follicular adnexal neoplasm showing differentiation toward the outer root sheath of the hair follicle[1,8]. It represents the malignant end of the proliferating trichilemmal tumour (PTT) spectrum arising from trichilemmal (pilar) cysts[2,6]. These tumours occur predominantly on the scalp and are most often reported in elderly women[7,8].

Although generally regarded as a low-grade malignancy, MPTT has the potential for local recurrence as well as regional and distant metastasis[5]. Clinically and histologically, it may closely mimic squamous cell carcinoma, making accurate histopathological evaluation essential for diagnosis.

Multifocal presentation involving anatomically distinct sites is exceedingly rare. We report a case of synchronous MPTT involving the scalp and scapular region in a male patient and highlight the characteristic histopathological, histochemical, and immunohistochemical findings important for appropriate diagnosis.

Case Details:

A 60-year-old moderately built and nourished male farmer presented with two progressively enlarging, painful swellings over the left scapular and scalp regions, present for 18 months and 3 months respectively. Each lesion measured approximately 3 × 3 cm in size.

Clinically, the swellings were immobile, globular, erythematous, with areas of serous discharge, non-pinchable overlying skin and firm to hard consistency. The patient complained of throbbing pain, which was aggravated on exposure to sunlight and relieved by analgesics. There were no other underlying comorbidities.

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Clinically, the lesions were provisionally diagnosed as soft tissue swellings (Figure1, figure 2). On dermatological evaluation, the scalp swelling was suspected to be a pilar cyst and the left scapular lesion was considered a probable epidermoid cyst.

Radiological evaluation may be useful in assessing tumour extent and regional lymph node involvement in suspected malignant proliferating trichilemmal tumours. Imaging modalities such as computed tomography (CT) or magnetic resonance imaging (MRI) can help evaluate deep tissue invasion and possible nodal metastasis, particularly in large or recurrent lesions. In the present case, ultrasonography revealed heterogeneously hypoechoic lesions suggestive of a neoplastic process; however, no clinical or radiological evidence of regional lymph node involvement was identified.

Fine-needle aspiration cytology (FNAC) suggested features of a malignant adnexal neoplasm, possibly of eccrine or trichilemmal origin.

The patient subsequently underwent wide local excision of both lesions, followed by skin grafting at the affected site. The resected specimens were then submitted for histopathological examination to confirm the diagnosis.

Gross Findings:

- **Left Scapular lesion:** A skin-covered soft tissue mass measuring 4 × 3 × 3 cm. The external surface showed erosion. The cut surface showed grey-white, firm mass measuring 3 × 1.8 × 2.8 cm, located 0.3 cm from the superior and inferior margins, 0.2 cm from the skin surface, and 0.5 cm from the deep resected margin. The overlying skin showed grey-brown discoloration (Figures 3 and 4).

- **Scalp lesion:** A partially skin-covered soft tissue mass measuring 3.5 × 3.3 × 2 cm, with surface erosion (3 × 3 cm) and rim of surrounding skin. On sectioning, an irregular grey-white solid mass measuring 3 × 2.5 cm with foci of haemorrhage was identified. The tumour was abutting the anterior and superior margins and located 0.1 cm from the inferior margin, 0.3 cm from the deep margin and 0.5 cm from both lateral and medial margins. (Figure 5, figure 6)

Microscopic Findings:

Sections studied show skin and subcutaneous tissue with a tumor in the dermis, composed of interlacing trabeculae, cords and lobules of tumor cells. These cells display moderate to marked pleomorphism, abundant eosinophilic cytoplasm and vesicular pleomorphic nuclei with prominent nucleoli (Figures 7, 8, and 9). Mitosis 5-6 per 10 high-power fields. The tumor is invaginated by the sclerotic collagen bundles. Margins are infiltrative, with focal extension into the overlying epidermis. Abrupt keratinization, areas of necrosis, stromal desmoplasia and a mixed inflammatory infiltrate comprising lymphocytes, plasma cells and eosinophils are noted. Evidence of vascular invasion is seen.

Diffuse cytoplasmic PAS positivity resistant to diastase digestion indicates trichilemmal keratinization (Figure 10). Immunohistochemistry showed focal cytoplasmic CD34 positivity supporting outer root sheath differentiation, diffuse strong nuclear p53 positivity (Figures 11, figure 12) and nuclear Ki-67 positivity indicating heightened proliferative activity (Figure 13). Based on histomorphology and immunohistochemistry, a final diagnosis of malignant proliferating trichilemmal tumour was rendered.

DISCUSSION

Malignant proliferating trichilemmal tumour (MPTT) is an uncommon malignant adnexal neoplasm arising from a pre-existing proliferating trichilemmal tumour within a pilar cyst[2,6,7,8]. The tumour predominantly affects elderly women with a marked female preponderance[2,7,8].

Histologically, MPTT is composed of atypical squamoid cells arranged in lobules, cords, and trabeculae showing nuclear pleomorphism, mitotic activity, necrosis, and infiltrative margins. A defining feature is trichilemmaltype keratinization, characterized by abrupt keratinization without formation of a granular layer, reflecting outer root sheath differentiation[2,7,8]. This feature is important in distinguishing MPTT from squamous cell carcinoma

(SCC), which demonstrates conventional keratinization with keratin pearl formation and a well-developed granular layer.

In the present case, diffuse PAS-positive and diastase-resistant cytoplasmic staining supported trichilemmal keratinization. Immunohistochemistry revealed focal CD34 positivity indicating outer root sheath differentiation, along with strong nuclear p53 expression and an increased Ki-67 labeling index, suggesting high proliferative activity. These findings correlated with previously reported profiles of MPTT and aided in differentiating it from SCC, which typically lacks CD34 expression[2,7,8].

Currently, there is no universally accepted staging system specific for malignant proliferating trichilemmal tumour. In cases demonstrating aggressive behaviour, staging principles similar to those used for cutaneous squamous cell carcinoma may be applied, taking into account tumour size, depth of invasion, and regional lymph node involvement.

Clinically, MPTT usually presents as a slowly enlarging scalp mass; however, rapid enlargement, ulceration, or pain may indicate malignant behaviour.

Complete surgical excision with histologically tumour-free margins remains the treatment of choice for malignant proliferating trichilemmal tumour. In the present case, wide local excision was performed and histopathological examination confirmed margin clearance. Adequate surgical margins are crucial to reduce the risk of local recurrence.

Although the prognosis is generally favourable, local recurrence (24.4%), regional lymph node metastasis (15.4%), and distant metastasis (9.2%) have been reported[5]. Given the multifocal nature of the lesions in this case, careful margin assessment and long-term follow-up are recommended.

The patient is currently under clinical follow-up, and no evidence of local recurrence or metastasis has been observed to date.

Feature	**Malignant Proliferating Trichilemmal Tumour (MPTT)**	**Squamous Cell Carcinoma (SCC)**
Origin	Outer root sheath of hair follicle	Epidermal keratinocytes
Keratinization	Abrupt trichilemmal keratinization without granular layer	Conventional keratinization with granular layer
Architecture	Lobulated tumour with pushing borders	Infiltrative nests and cords
Cytology	Atypical squamoid cells	Malignant squamous cells with keratin pearl formation
PAS stain	PAS positive and diastase resistant	Usually negative
CD34	Often positive	Typically negative
p53	Overexpression may be present	Frequently positive
Clinical behaviour	Usually localized with rare metastasis	Higher metastatic potential

Accurate recognition of trichilemmal keratinization and appropriate use of histochemical and immunohistochemical markers are essential to avoid misdiagnosis as squamous cell carcinoma, which may lead to differences in prognostic assessment and management strategies.

CONCLUSION

This case demonstrates the characteristic histopathological and immunohistochemical features of malignant proliferating trichilemmal tumour and emphasizes the rarity of multifocal lesions occurring in a male patient.

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Recognition of trichilemmal keratinization and appropriate use of histochemical and immunohistochemical studies are essential for distinguishing MPTT from squamous cell carcinoma.

MPTT is an uncommon cutaneous adnexal malignancy with potential for local recurrence and metastasis. Early diagnosis, complete surgical excision with tumour-free margins, and long-term follow-up are important to ensure optimal patient outcomes.

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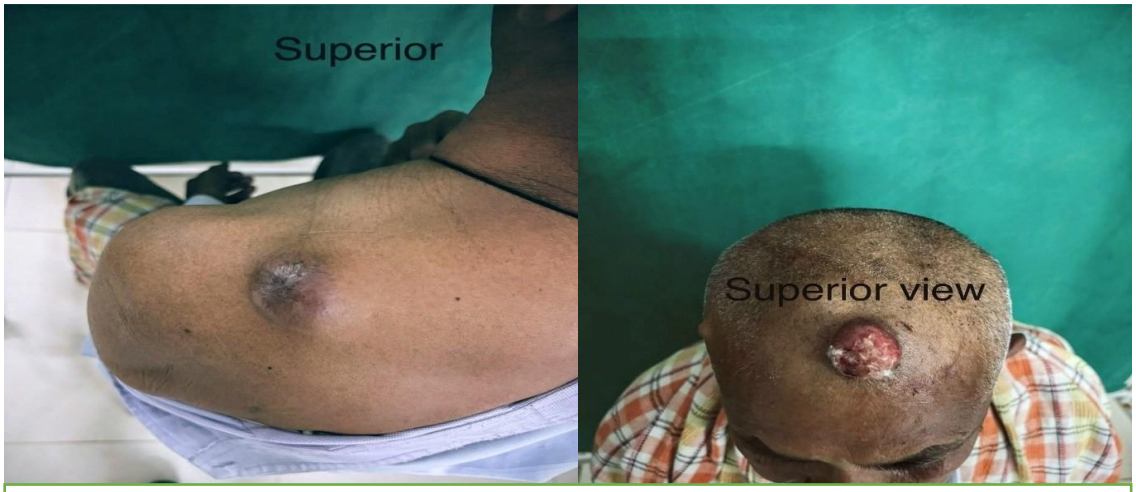

Figure 1 & 2: left scapular & scalp swelling- globular, firm to hard in consistency, painful swelling

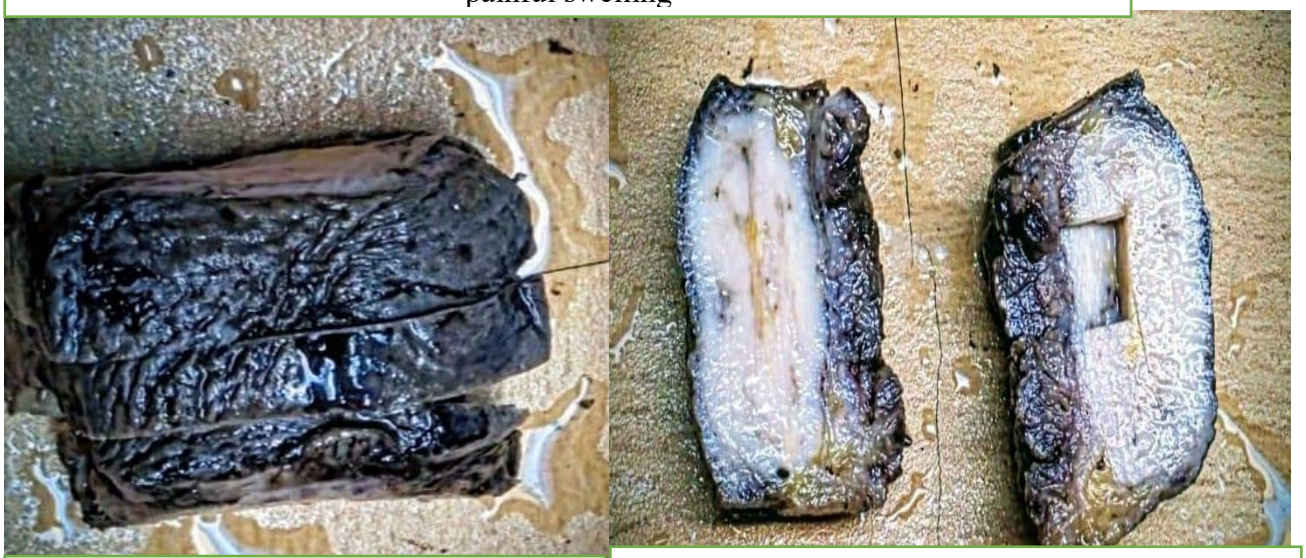

Figure 3: External surface of left scapular lesion showed grey-brown discoloration.

Figure 4: cut surface of from left scapular lesion showed grey-white, firm, and homogeneous, with interspersed grey-brown areas.

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Figure 5

Figure 5: scalp lesion partially skin-covered soft tissue mass measuring 3.5 × 3.3 × 2 cm. The external surface shows an ulcerration measuring 3 × 3 cm

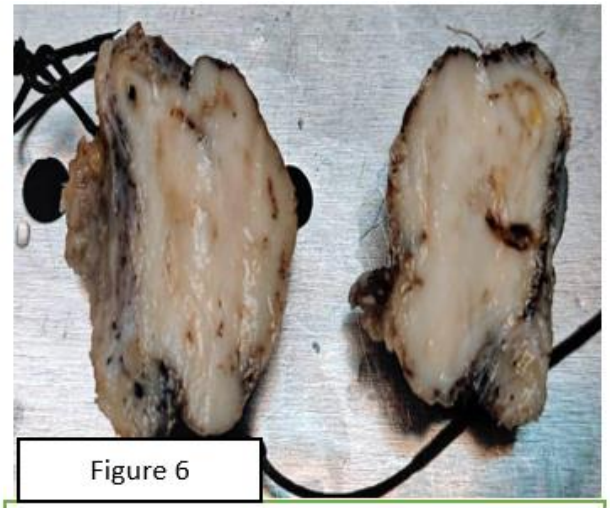

Figure 6

Figure 6: scalp lesion Sectioning showed irregular, solid, homogeneous grey-white tissue mass foci of haemorrhage

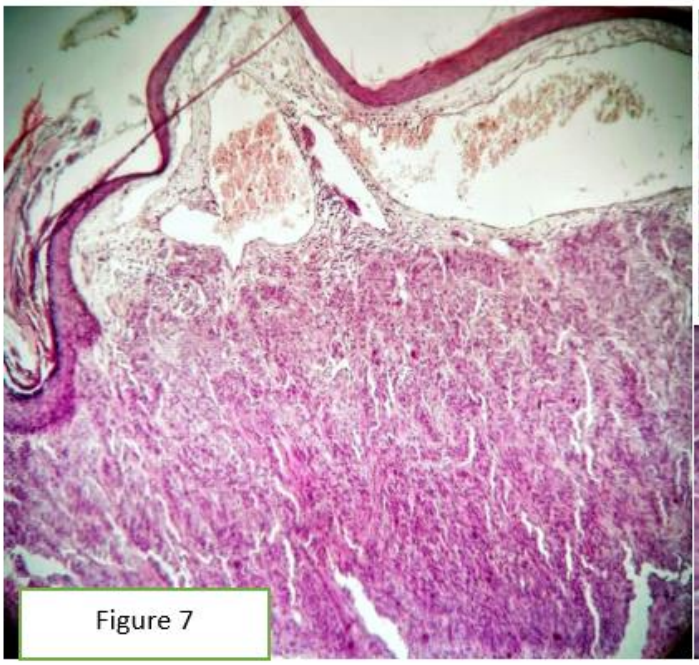

Figure 7

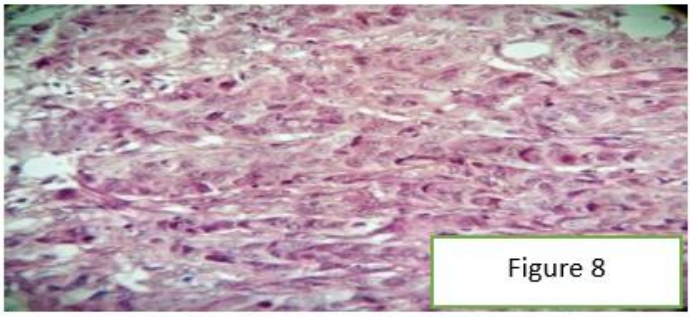

Figure 8

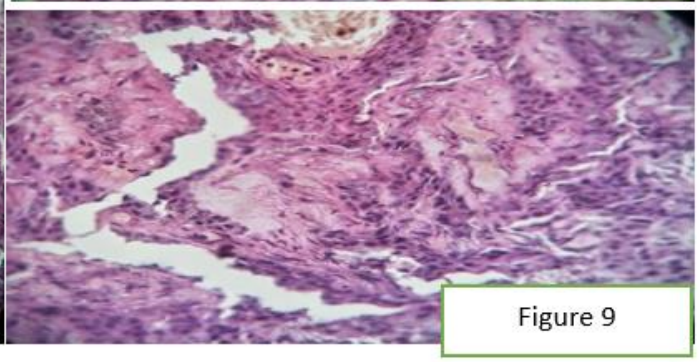

Figure 9

Figure 7: 4x H & E skin and subcutaneous tissue with a tumor in the dermis, composed of interlacing trabeculae, cords and lobules of tumor cells with intravascular tumour emboli.

Figure 8: 10x H & E Individual tumor cells are moderate to markedly pleomorphic with abundant eosinophilic cytoplasm, vesicular pleomorphic nuclei with visible nucleoli

Figure 9: 10x H & E Abrupt keratinization.

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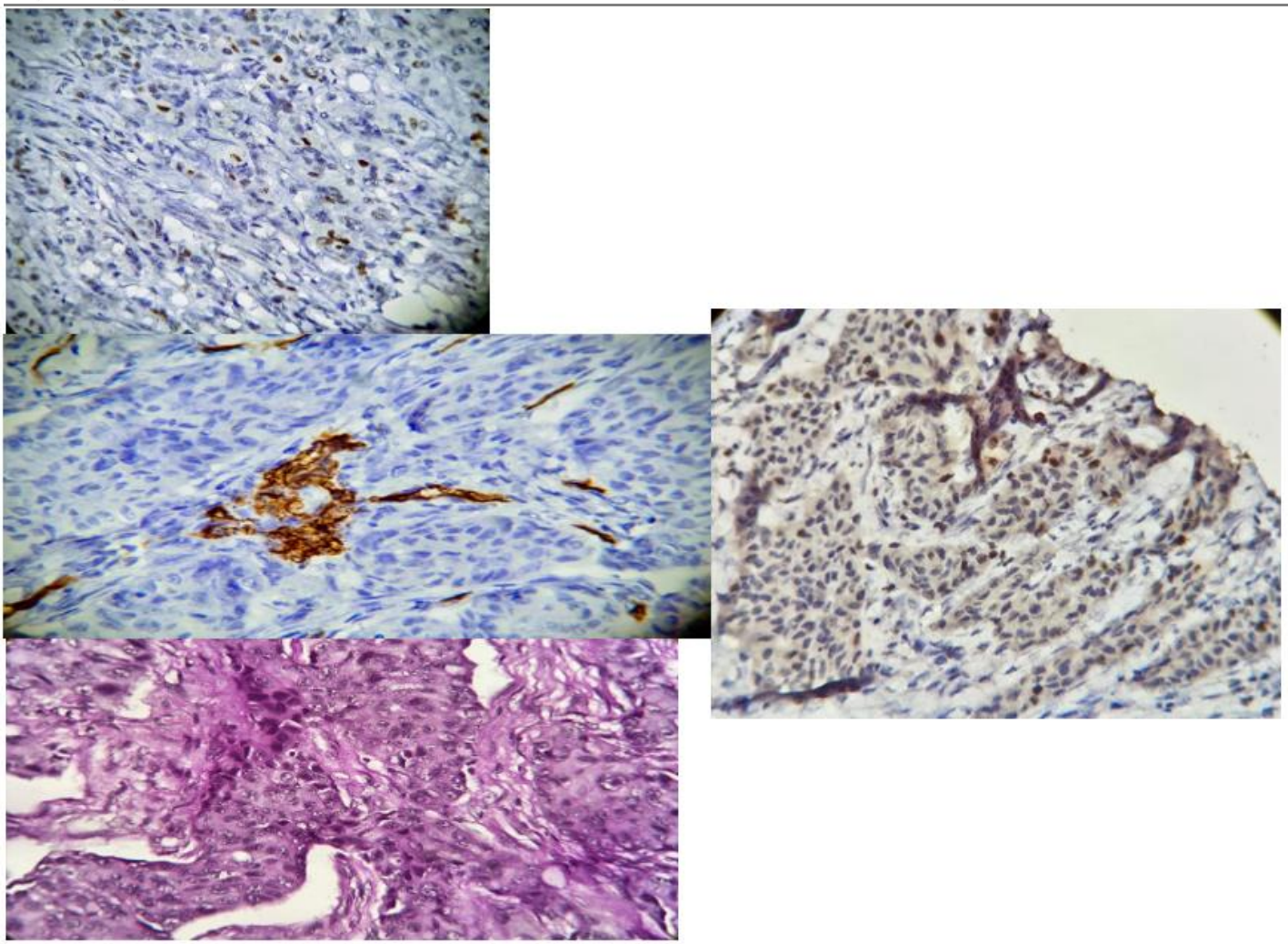

Figure 11