

Malignant proliferating trichilemmal tumour: a case report

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Abstract

An 86-year-old man presented with a painless, pea-sized growth over the left angle of his jaw, which had been gradually enlarging over two years. A clinical diagnosis of pilar cyst was made. Histopathological examination of the mass revealed a malignant proliferating trichilemmal tumour. During follow-up 4 months later, a palpable small upper jugular lymph node was noted at the left side of the neck. Biopsy revealed a metastatic malignant trichilemmal tumour. This case illustrates a rare malignant tumour which is a challenge to clinical diagnosis.

Key words: Proliferating trichilemmal tumour, lymph node

INTRODUCTION

Proliferating trichilemmal (pilar) tumour is an uncommon skin neoplasm. 39 cases have been reported from 1980 to 1998 in the medical literature,¹⁻¹⁰ of which 11 were **malignant**.^{1,2} The origin of these tumours appears to be the outer root sheath of the hair.⁸ More than 80% of reported patients were female and elderly. The tumour has a predilection for the scalp (90%) with the residual 10% occurring mainly on the back.¹¹ In rare instances, malignant transformation of proliferating trichilemmal cysts takes place. This is indicated clinically by rapid enlargement of the nodule.¹ Microscopically, extensive areas of severe dysplasia of the neoplastic epithelium and invasion into the surrounding tissue are seen.¹ Even though areas of trichilemmal keratinization are still in evidence, nuclear atypia and giant nuclei indicate **malignancy**.⁴ There are several reported instances of metastases, most of them **regional**.^{2,5,12} In one instance, however, the metastases were generalised and **fatal**.⁴ Penetration of the tumour into cerebral sinuses causing death has also been **reported**.¹³

CASE REPORT

An 86-year-old man presented with a pea-sized growth over the left angle of his jaw. He complained that it was gradually increasing in size over the past two years. There was occasional itchiness in the surrounding areas but the growth was painless. The clinical diagnosis was a pilar cyst and an excision biopsy was carried out.

Pathology

The histopathology laboratory received an elliptical piece of skin measuring 1.6 x 1.0 cm with an elevated nodule on the surface measuring 1.0 cm in maximum diameter. Microscopical examination revealed a skin covered tumour composed of irregularly-shaped lobules, nests and bands of squamous epithelium with abrupt central keratin formation. The tumour cells were pleomorphic with vesicular nuclei, prominent nucleoli and frequent aberrant mitoses (Figs. 1 & 2). Scattered giant nuclei were seen. Focal areas of stromal invasion were evident. The deep surgical margin was involved by the tumour. A histopathological diagnosis of malignant proliferating trichilemmal tumour (MPTT) was then made.

Clinical course

During follow-up 4 months later, he was noted to have a small palpable, mobile upper jugular lymph node at the left side of the neck just below the site of previous excision. The area of previous excision showed a healed scar with no evidence of residual tumour. The node was excised. Microscopical examination revealed lymph nodal tissue partly effaced by malignant tumour composed of islands and trabeculae of squamous epithelium, some with central necrosis (Figs. 3 & 4). A histopathological diagnosis of metastatic malignant trichilemmal tumour was made.

The patient was otherwise asymptomatic. Systemic examination and radiological investigations did not reveal further metastatic



FIG. 1: Malignant proliferating trichilemmal tumour composed of irregular lobules and islands of squamous epithelial cells beneath the skin. H&E X 40.

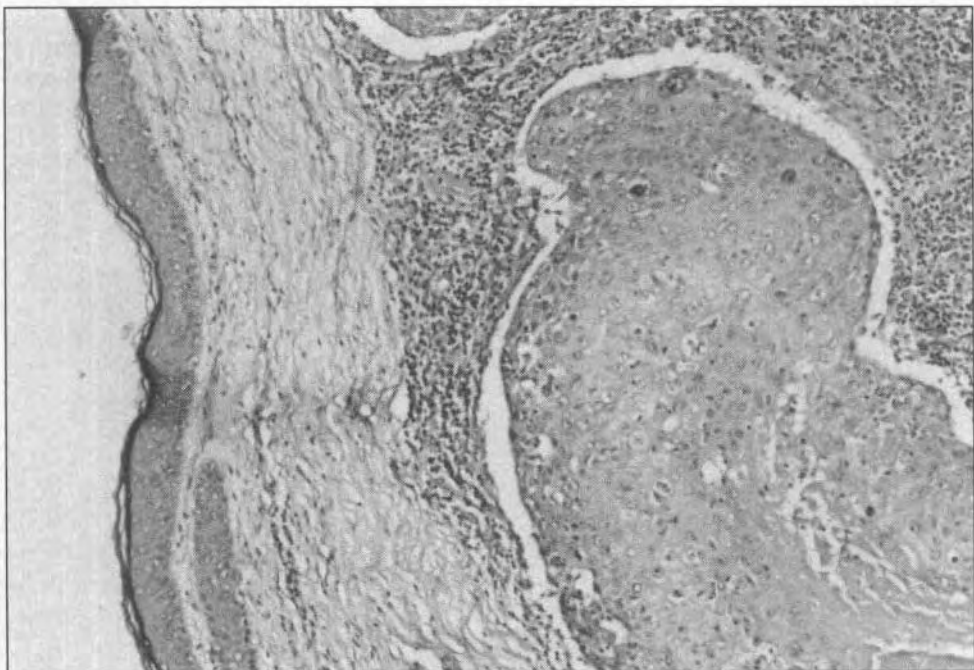


FIG. 2: A higher magnification view of the tumour epithelial cells showing the presence of nuclear atypia and abrupt central keratinisation. H&E X 100.

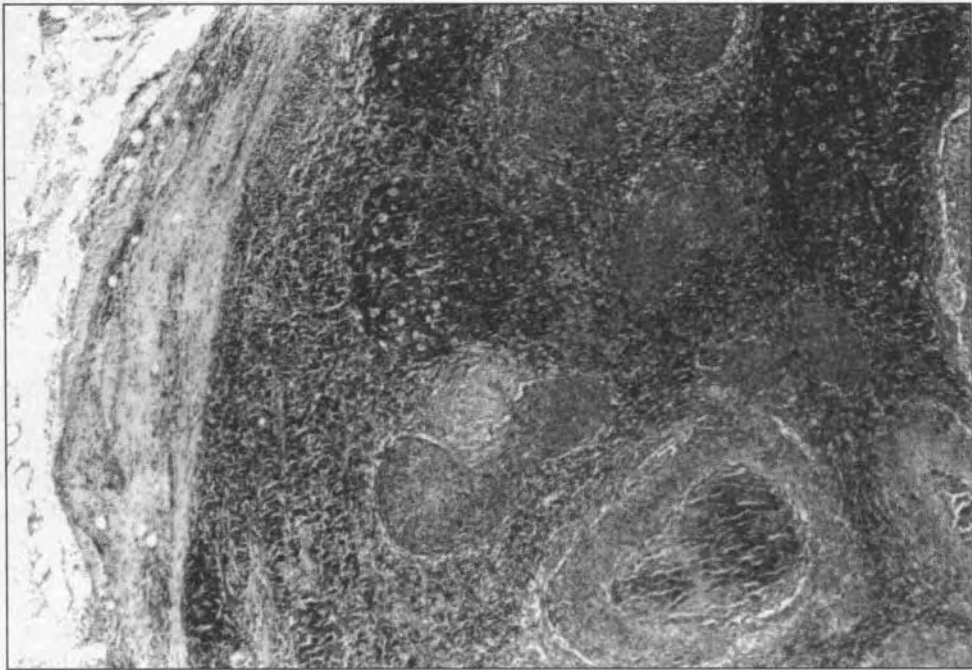


FIG. 3: Involvement of lymph node by metastatic tumour. H&E X 40.

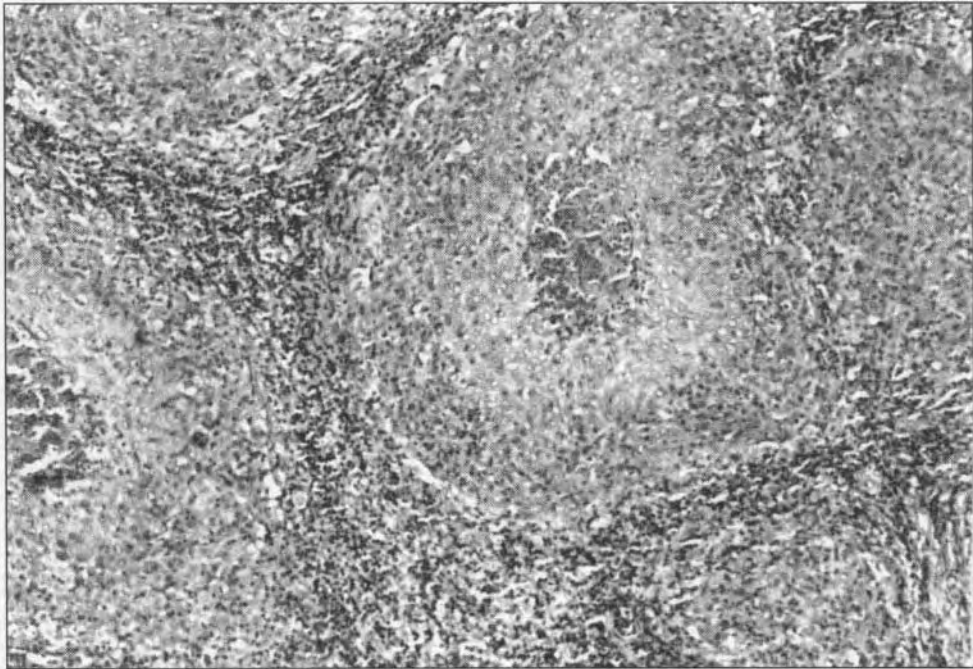
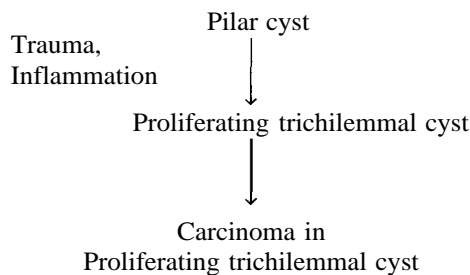


FIG. 4: A higher magnification of the metastatic tumour revealing trichilemmal epithelium with central necrosis. H&E X 100.

disease. However, he defaulted follow-up soon after and his present clinical status is unknown.

DISCUSSION

The proliferating trichilemmal (pilar) cyst occurs most commonly as a solitary tumour on the scalp of elderly women.¹⁴ They are thought to arise from pre-existing trichilemmal (pilar) cysts, possibly after inflammation or trauma.¹⁵ Frank malignant change is rare in trichilemmal (pilar) tumours, with only a small number of cases reported in the scientific literature.¹ It has been suggested that these tumours form a continuum from benign neoplasia to malignancy and that malignancy develops in a sequence as follows:¹⁶



Therefore, proliferating trichilemmal cyst can be regarded as a neoplastic entity rather than a hyperplastic one, a hypothesis which is supported by the finding of non-diploid DNA content in some of the cases.¹⁷ Malignant change in a proliferating trichilemmal cyst is a rare event and only 11 previous cases have been reported.¹ Most of these record a rapid increase in size in a long-standing tumour. Histologically, malignancy is characterised by poor circumscription with invasion of surrounding tissues. There is severe nuclear atypia, hyperchromasia and atypical mitotic activity.¹⁶ Several instances of local recurrences have been reported but metastases are exceptionally rare.¹ Metastases to lymph nodes and internal organs have been recorded in eight cases and was given as the cause of death in one.^{2,4,5,10,12,13} The few well-documented instances of metastases have occurred in tumours with clear-cut malignant microscopical features⁴ as in the case described here. Exceptionally, the malignant component has the features of a spindle cell carcinoma.¹⁸ Frankly malignant cells intimately associated with more typical areas of trichilemmal differentiation suggests the possibility that trichilemmal (pilar) tumours may

progress through a series of stages as discussed earlier. Ruty et al¹⁶ confirmed that morphological malignancy in trichilemmal tumours is accompanied by increased cell proliferation and DNA aneuploidy. The findings lend support to the idea of sequential progression to malignancy in proliferating trichilemmal tumours. Progression in trichilemmal tumours would be in keeping with the multistep theory of neoplasia.¹⁹ However, the stimuli which may induce this change are currently unclear although trauma and infection have previously been suggested.¹⁵

3 cases of malignant proliferating trichilemmal tumours were reported by Herrero et al⁶ in which, in addition, to the histological features, DNA ploidy, nuclear area and proliferative fraction were determined. CD34 immunoreactivity had also been tested. 2 cases were found to be aneuploid while 1 was diploid with increased proliferating index. PCNA immunostaining labelled 40% and 80% of tumour cells in the two aneuploid tumours respectively and 30% of the diploid neoplasm. In all cases, nuclear area was consistent with large pleomorphic tumour cells. No CD34 immunostaining was detected in tumour cells. The author concluded that aneuploidy was common in malignant proliferating trichilemmal tumour, particularly tumours with a high proliferative fraction. Therefore, evaluation of the DNA content, proliferation markers and CD34 immunostaining may be helpful in the diagnosis of MPTT.

A case of malignant proliferating trichilemmal tumour showing distant metastases was described by Park et al.⁷ For 10 years the patient had had a round mass in the occiput which recurred twice after wide excisions, and later metastasized to the cervical lymph nodes, periparotid area and chest. Each time the lesions were excised and histological examination demonstrated a proliferating trichilemmal tumour with increasing nuclear atypia. Serial specimens showed increasing Ki-67 positivity as the tumour advanced.

The above case was diagnosed based on the histopathological features of the tumour. No immunohistochemical stains were performed as the histopathological diagnosis was unequivocal. What was thought to be a benign pilar cyst on clinical examination turned out to be a malignant tumour with eventual lymph node metastases on histological examination. This case exemplifies the difficulties in clinical diagnosis of these rare tumours and their unpredictable course.

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