

Case Report

Proliferating trichilemmal tumour: a case report with review of literature

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ABSTRACT

Proliferating trichilemmal tumour is a solid-cystic neoplasm that shows trichilemmal differentiation similar to that of the isthmus of the hair follicle histologically characterised by the presence of trichilemmal keratinization. Proliferating Trichilemmal Tumour (PTT) appears mainly in elderly women and is in general a solitary lesion on the scalp. Proliferating trichilemmal tumours generally have a benign clinical course, and a clinical differentiation from squamous cell carcinoma is often difficult. We report a case of PTT in a 30 year old man presenting as a solitary 10x8 cm ulcerated nodule on the scalp since 3 months clinically resembled a malignant tumour. The therapeutic approach is surgical removal with a wide clear margin.

Keywords: Proliferating trichilemmal tumour, Trichilemmal keratinization, Scalp

INTRODUCTION

Proliferating Trichilemmal Tumour (PTT) is an uncommon neoplasm, first described in 1966 by Wilson and Jones as a "proliferating epidermoid cyst".¹

Proliferating trichilemmal tumour appears mainly in women during fourth to eighth decades of life and is in general a solitary lesion on the scalp. It presents as a proliferative cauliflower like growth mimicking squamous cell carcinoma. They typically undergo slow but progressive enlargement over several months to years, yielding lobulated and variably exophytic masses that occasionally might ulcerate.²

Here we report a case of Proliferating Trichilemmal Tumour (PTT) in a 30 year old male who presented with a large nodular ulcerated mass on the scalp clinically resembling a malignant tumour. However, histology confirmed the benign nature.

CASE REPORT

A 30 year-old male presented for the first time with a history of swelling over the right side of scalp since 3 months duration, increasing in size. A detailed clinical history was taken. On examination, a single swelling measuring 10x8 cm with a nodular and ulcerated surface and soft in consistency was seen on the scalp (Figure 1). It was painless and not adherent to the underlying bone. No lymph nodes were palpable in the neck. Initially an incisional biopsy was done which was diagnosed as clear cell hidradenoma (benign skin adnexal tumour) on histopathology. Subsequently the tumour was excised. Grossly, the tumour was predominantly solid, well circumscribed, grey tan in color with soft friable areas (Figure 2). It was covered with hairy skin and had a margin of normal tissue all around. The histopathology revealed tumour composed of sheets of well differentiated squamous cells exhibiting abrupt keratinization (Figure 3). Scattered foci of clear cells were also seen (Figure 4) and the tumour showed

minimal nuclear atypia (Figure 5). The stroma showed a mild mononuclear inflammatory infiltrate. All the surgical margins were free of the tumour. A diagnosis of benign proliferating trichilemmal tumour was made.

After six months of follow up, it is found out that the patient is free of any symptoms.



Figure 1: Photograph of gross morphology of proliferating trichilemmal tumour showing 10x8x6cms nodular ulcerated tumour arising from the scalp.



Figure 2: Photograph of cut section of proliferating trichilemmal tumour showing solid well circumscribed tumour grey tan in color with soft friable areas.

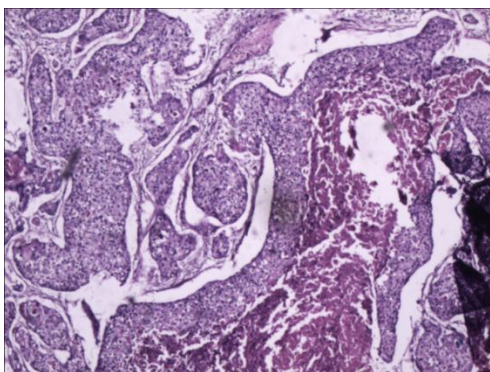


Figure 3: Photomicrograph showing sheets and lobules of squamous cells exhibiting trichilemmal keratinisation. (H&E; 40x).

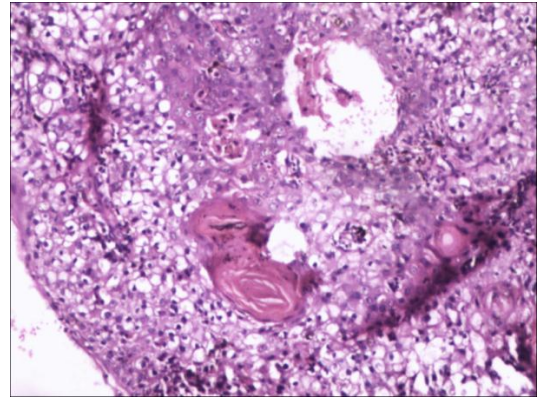


Figure 4: Photomicrograph showing sheets of squamous cells with clear cell change and abrupt keratinisation. (H&E; 100x).

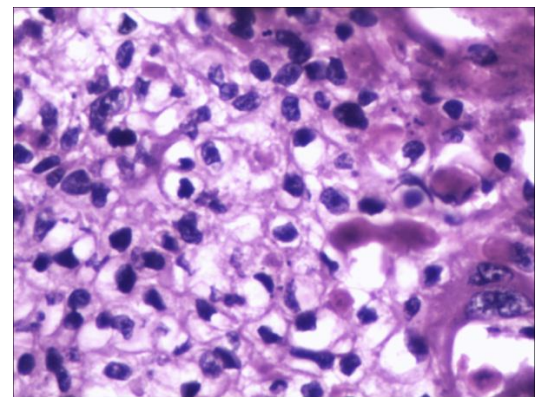


Figure 5: Photomicrograph showing tumour cells exhibiting mild nuclear pleomorphism. (H&E; 400x).

DISCUSSION

The proliferating trichilemmal cyst, also referred to as proliferating trichilemmal tumour or proliferating Pilar tumour, is nearly always a single lesion; rarely, there are two proliferating trichilemmal cysts.^{3,4} The neoplasm is more frequent in women than in men and most patients are elderly.⁵ More than 90% of the lesions are situated on the scalp. Other described locations, in decreasing order of frequency, include face, trunk, back and forehead.⁵ Multiple lesions are very rare. The size ranges from 2-10 cm in diameter, although lesions up to 25 cm in diameter have been described.⁶ Alopecia and ulceration can be found. Starting as a subcutaneous nodule suggestive of a wen, the tumour may grow into a large, elevated, lobulated mass that may undergo ulceration and thus greatly resemble a squamous cell carcinoma.⁷ In several instances, rapid enlargement of nodular scalp lesions has indicated malignant transformation.⁸

The proliferating trichilemmal cyst, or proliferating trichilemmal tumour, usually is well demarcated from the surrounding tissue.⁹ Proliferating trichilemmal tumour occurs on a morphologic continuum. On one end of the spectrum, it consists of a well circumscribed solid and

cystic neoplasm which involves the dermis and sometimes extends to the subcutaneous tissue. On the other end of the morphologic spectrum are neoplasms with malignant features such as invasive growth extending beyond the confines of the cyst wall coupled with nuclear pleomorphism and high mitotic activity. These areas may be indistinguishable from squamous cell carcinoma. It is composed of multiple, variably sized lobules composed of squamous epithelium. Some of the lobules are surrounded by a vitreous layer and show palisading of their peripheral cell layer.⁴ Characteristically, the epithelium in the centre of the lobules abruptly changes into eosinophilic amorphous keratin. The tumour cells in many areas show some degree of nuclear atypia, as well as individual cell keratinization, which at first glance suggests a squamous cell carcinoma.^{10,11} The tumour differs from a squamous cell carcinoma by a rather sharp demarcation from the surrounding stroma as well as an abrupt mode of keratinization.⁹ Features of malignant tumour include non-scalp locations, size >5 cm, recent rapid increase in size, infiltrative growth pattern on histology, palpable lymph nodes, fixity to deeper structures.

In our case though the tumour is more than 5 cm clinically, there were neither palpable lymph nodes nor fixity to deep structures and infiltrative growth pattern on histopathological examination. Some tumours show vacuolization or clear cell formation of some of the tumour cells as a result of glycogen storage.^{10,12} Up to 20% of the lesions may undergo malignant transformation into squamous cell carcinoma.⁵ Proliferating trichilemmal tumour expresses fetal hair root cytokeratin, as well as cytokeratin.^{7,13} The pathogenesis remains unknown. In some cases, human papilloma virus has been implicated in the etiology.¹⁴ It is unclear if proliferating trichilemmal tumours arise de novo or from pre-existing trichilemmal cysts.^{3,15} Proliferating trichilemmal tumours without atypical features generally behave in a benign fashion.¹⁶ Yet, complete excision is recommended to avoid recurrences, and to allow for complete histopathological evaluation. Tumours with an invasive growth pattern or cytological atypia have an unpredictable course. They may be locally aggressive, recur, or metastasize.

In view of the benign nature of proliferating trichilemmal tumour which sometimes clinically mimics malignancy, it has to be kept in the differential diagnosis of nodular or ulcerative lesions of the scalp.

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