CASE REPORT

A Rare Cutaneous Adnexal Tumour with a Rare Presentation

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Abstract:

Proliferating Trichilemmal Tumour (PTT) is a rapidly growing large cutaneous adnexal neoplasm. Although biologically considered as benign, it may be locally aggressive. Malignant transformation of these lesions, known as Malignant Proliferating Trichilemmal Tumour (MPTT) has rarely been reported. So far in the literature, only 39 well-documented cases of MPTT have been reported. MPTT has been stated to be a neoplasm of the older age group according to review of the literature. We present a case of MPTT in a young male. A 25 year old male presented with a scalp swelling of 2 years duration with a recent rapid enlargement. The swelling was excised and histopathological examination of the excised specimen revealed features of MPTT. The differential diagnosis of MPTT is squamous cell carcinoma as both share common features. Accurate diagnosis of MPTT is essential since it has a tendency to metastasize and recur more frequently than squamous cell carcinoma.

Keywords: Proliferating Trichilemmal Tumour, Malignant Proliferating Trichilemmal Tumour, Young Male

Introduction:

Proliferating Trichilemmal Tumour (PTT) is a rare, usually benign, cutaneous neoplasm originating from the external root sheath of a hair follicle [1]. Scalp is the most common site of these tumours and clinically they present as irregularly defined subcutaneous nodules which are often cystic [2]. Trichilemmal keratinization is the characteristic histological finding of PTT which consists of sudden compact amorphous keratinization of the epithelial cells that cover the cyst wall without a granular layer. PTTs are usually benign, but rarely can undergo a malignant transformation known as malignant proliferating trichilemmal tumour. MPTT is described as proliferating trichilemmal tumour that invades neighboring tissues accompanied with anaplasia and necrosis [3]. MPTT is usually confused with squamous cell carcinoma, its accurate diagnosis being essential, as it has a tendency to metastasize and recur more frequently than squamous cell carcinoma. MPTT is revealed to be a neoplasm of the elderly according to review of literature [4, 5]. We present a case of MPTT occurring in a 25 year male.

Case Report:

A 25 year old male presented with solitary painless swelling over scalp since 2 years which showed recent rapid enlargement. The patient was otherwise healthy with no significant past medical history. The mass was excised with a clinical diagnosis of dermatofibroma/sebaceous cyst. On gross examination, the mass was nodular, partly skin covered, measuring 5.5x4.3x3 cms. External surface showed foci of skin ulceration measuring 0.5x0.5 cms, areas of congestion and two tiny pale white nodules measuring 0.3 cms each. The cut surface of the mass revealed a solid, lobulated, grey white appearance.

Microscopic examination of the specimen showed a tumour in dermis and subcutaneous tissue arranged in lobules and diffuse sheets (Fig. 1). The major portion of the tumour was composed of oval to polygonal cells having abundant clear to eosinophilic cytoplasm (Fig. 2). The tumour lobules comprised of polygonal cells showed abrupt keratinisation. Tumour cells showed moderate to marked pleomorphism, high nuclear cytoplasmic ratio, nuclear hyperchromatism (Fig. 3) and high mitotic activity with abnormal mitotic figures. Few bizarre cells, spindle shaped cells, tumour giant cells, foci of invasion into surrounding tissue and extensive areas of tumour necrosis (Fig. 4) were also noted. Also notable was a distinct area, resembling a typical benign pilar tumour (Fig. 5), composed of interlacing lobules of smaller peripheral cells that were palisaded and matured into larger central cells with trichilemmal keratinization. Based on these findings, diagnosis of malignant proliferating trichilemmal tumour was made in this case.

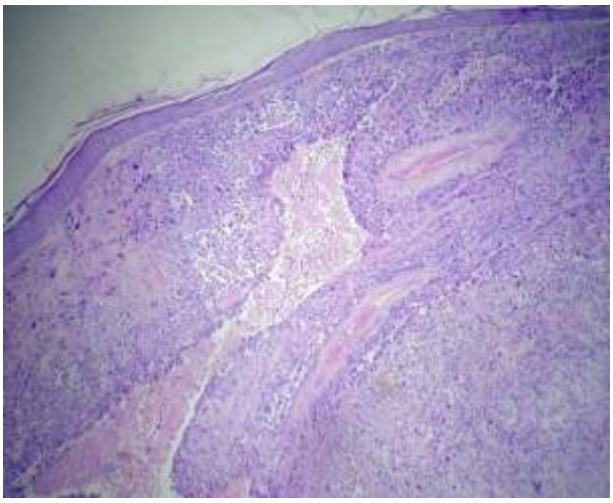

Fig 1: Photomicrograph showing Tumour Arranged in Lobules and Sheets (H&E,40x)

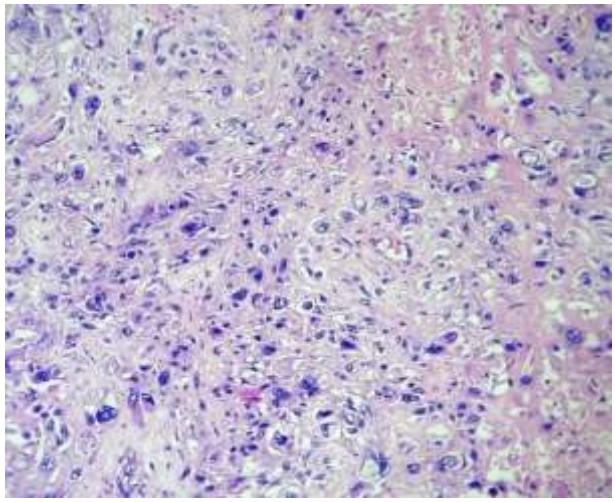

Fig 2: Photomicrograph Showing Cells with Clear to Eosinophilic Cytoplasm (H&E,100x)

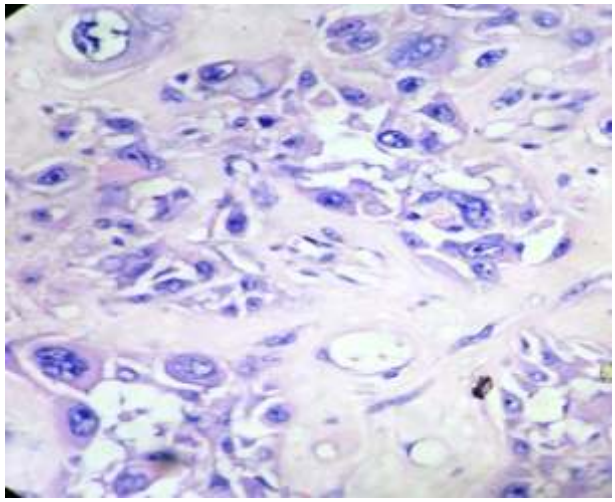

Fig 3: Photomicrograph showing Cells with Pleomorphic Hyperchromatic Nuclei (H&E, 400 x)

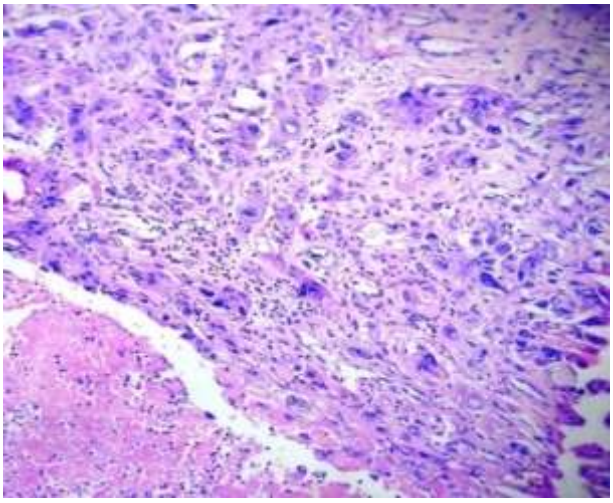

Fig 4: Photomicrograph Showing Areas of Necrosis and Cellular Atypia (H&E, 100x)

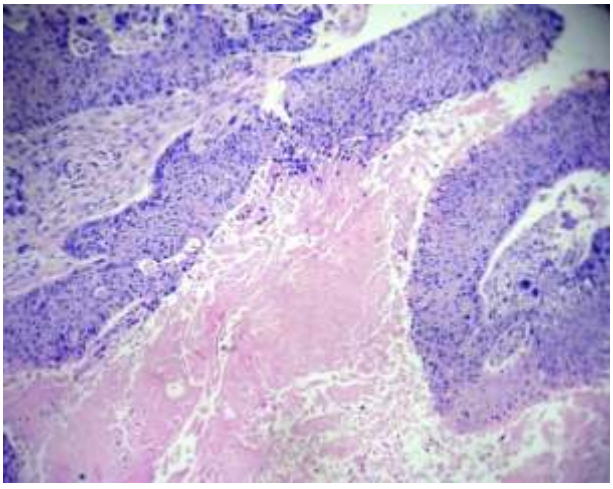

Fig 5: Photomicrograph Showing Features of Benign Pilar Tumour (H&E, 40x)

Discussion:

Cutaneous tumours derived from the outer root sheath of hair follicles, which show trichilemmal keratinisation, are trichilemmal cysts, proliferating trichilemmal cysts and malignant proliferating trichilemmal tumour [6]. Trichilemmal cyst is by far the most common among the trichilemmal tumours [5]. Proliferating trichilemmal tumour is a rare but morphologically distinctive tumour usually occuring in the scalp of elderly women [5]. Lanugo hair follicles of the bald scalp and follicles of other areas devoid of non-terminal hair are unlikely to produce these tumours. Therefore, pilar tumours are not seen in the bald scalp, being more common in areas with excess hair growth [2]. Proliferating trichilemmal tumour has been reported under a variety of terms including giant hair matrix tumour, invasive pilomatrixoma, proliferating epidermoid cyst, pilar tumour of the scalp, trichilemmal pilar tumour, trichochlamydocarcinoma, proliferating trichilemmal cyst, proliferating trichilemmal tumour, and proliferating follicular cystic neoplasm [7, 8].

Frank malignant change is rare in trichilemmal (pilar) tumours which occur in a step-wise manner with an adenomatous stage of the trichilemmal cyst to an epitheliomatous stage of the PTT evolving into the carcinomatous stage of the MPTT. Various factors have been implicated in this oncological transformation which include trauma and inflammation [9].

MPTT is the rarest of the trichilemmal tumours with only 39 well-documented cases of MPTT published till date in the English literature [10]. The term MPTT was originally described in 1983 by Saida *et al.,* and its biological behaviour still remains unpredictable [11]. MPTT can occur *de novo* but most often arises in a pre-existing benign proliferating trichilemmal cyst [10].

This tumour lacks a distinctive histological or immunohistochemical marker to suggest malignant transformation. Clinically sudden enlargement of long standing nodular scalp lesions and histological evidence of significant abnormal mitosis, marked cellular pleomorphism, infiltrating margins and aneuploidy reflect malignant transformation [11]. Our case possibly represents the event of malignant transformation in a proliferating trichilemmal tumour.

The differential diagnosis of MPTT is squamous cell carcinoma as both are known to occur at the same site [11, 12]. Histological examination of a malignant tumour in the scalp showing evidence of keratin production suggests squamous cell carcinoma, a more common tumour at this site [11]. Evidence of trichilemmal keratinization, lobular pattern and the lack of a precursor epidermal lesion such an actinic keratosis differentiate MPTT from squamous cell carcinoma. Since MPTT has a tendency to metastasize and recur more frequently than squamous cell carcinoma, an accurate diagnosis is essential [11]. The real incidence of a malignant proliferating trichilemmal cyst is unknown, due to its rarity and inconsistencies in nomenclature and misclassification as squamous cell carcinoma [10]. MPTTs are primarily local; hence, adequate wide surgical excision remains the mainstay of treatment. The patient should be followed closely after surgery [10].

Conclusion:

Malignant proliferating trichilemmal tumours can rarely occur in the young especially, in an individual with pre-existing proliferating trichilemmal tumour and pose a diagnostic dilemma for the pathologist. Since it follows an aggressive course, it is essential to distinguish it from other similar-looking neoplasms for an appropriate therapy.

References

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