

Giant Proliferating Trichilemmal Tumour: A Case Report

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ABSTRACT

A Trichilemmal Cyst (TC), which can also be referred to as a trichodermal cyst or trichodermal isthmus-degenerative cyst, is a noncancerous growth on the skin that arises from the outer sheath of hair follicles and is characterised by its rare occurrence and limited documentation. Herein, we report a 54-year-old female who presented with a swelling over her scalp for 15 years, which gradually progressed to a size of 8x5 cm. Computed Tomography (CT) scan revealed an extra-cranial well-defined lobulated heterogeneously enhancing isointense lesion with specks of calcification above the occipital bone in the midline with no intracranial or bony extension, likely benign in aetiology. She underwent surgical excision of the entire swelling, and histopathological analysis confirmed the diagnosis of Proliferating Trichilemmal Tumour (PTT) with squamous differentiation. The wound healed well without any recurrence after a follow-up of one year. Inspite of their enhanced propagation, TCs are usually benign and non-invasive. But in a few cases, these can transform to trichilemmal carcinoma, these carcinomas may cause local invasion, resulting in significant morbidity and mortality.

Keywords: Benign tumours, Epidermoid cyst, Hyperplastic hair follicle cyst, Pilar cyst, Pilar tumour, Scalp swelling

CASE REPORT

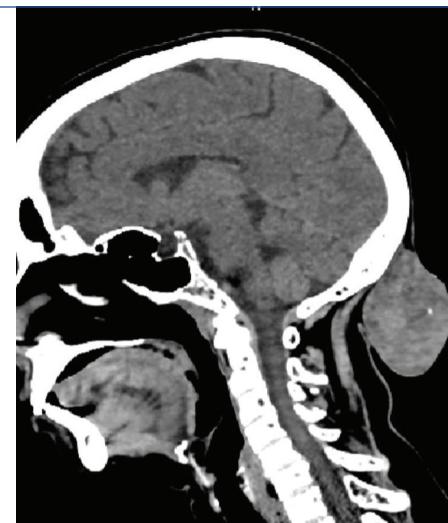
A 54-year-old female came to the outpatient department with the chief complaint of painless swelling over the scalp for 15 years, which gradually progressed to a size of 8x5 cm. She did not have any neurological complaints. Examination revealed around 8x5 cm solitary, partially ulcerated, firm mass in the occipital scalp region with no superficial skull or periosteal attachment [Table/Fig-1,2]. A Computed Tomography (CT) scan revealed an extra-cranial well-defined lobulated heterogeneously enhancing isointense lesion with specks of calcification seen above the occipital bone in the midline with no intracranial or bony extension, few vascular channels from the scalp seen supplying this lesion with the impression of well-defined heterogeneously enhancing soft-tissue lesion of the scalp as described- likely benign in aetiology [Table/Fig-3].



[Table/Fig-1]: Clinical photograph taken from left-side of the patient showing swelling over the scalp in the occipital region.

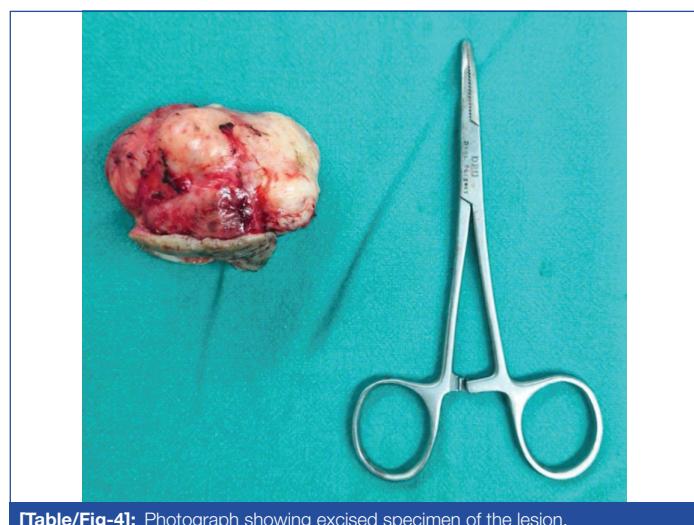


[Table/Fig-2]: Clinical photograph taken from behind the patient showing a small ulceration on swelling over the scalp.



[Table/Fig-3]: CT scan image showing well-defined heterogeneously enhancing soft-tissue lesion of the scalp.

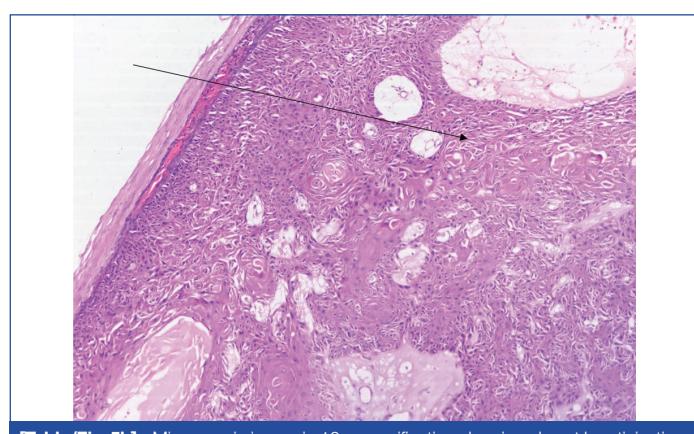
Based on clinical assessment and radiological findings, the differentials were proliferating TC, epidermoid cyst, dermoid cyst, and pilomatrixoma. Surgical excision of the swelling was performed and the excised specimen was sent for histopathological examination. During surgery, the lesion was excised [Table/Fig-4] with a 1 cm margin of normal skin in view of recurrence. On microscopy, Haematoxylin and Eosin (H&E) sections showed epidermis with subcutis and tumour, displayed a lobular pattern, tumour cells were polygonal with indistinct cell border and mild nuclear atypia, extensive squamous differentiation with abrupt keratinisation was visualised with focally noted mild increase in mitotic activity but there was no evidence of any invasion into the surrounding stroma or haemorrhage or necrosis in the sections studied. Histopathological findings were suggestive of PTT with squamous differentiation [Table/Fig-5a,b]. The patient was discharged on the third postoperative day after dressing. The wound healed well without any recurrence after a follow-up of one year.



[Table/Fig-4]: Photograph showing excised specimen of the lesion.



[Table/Fig-5a]: Microscopic image in 10x magnification, black arrows represent the dysplastic squamoid cells.



[Table/Fig-5b]: Microscopic image in 10x magnification showing abrupt keratinisation.

DISCUSSION

A TC, which can also be referred to as a trichodermal cyst or trichodermal isthmus-degenerative cyst, is a non-cancerous growth on the skin that arises from the outer sheath of hair follicles [1,2]. PTT is a well-defined neoplasm located in the dermis or subcutaneous tissue, noted for its trichilemmal keratinisation and squamoid cytological features [3]. In 1966, Wilson-Jones described this tumour as a histological entity that can resemble squamous cell carcinoma [4]. Description of this tumour has numerous terms such as proliferating TC, pilar tumour of the scalp, giant hair matrix tumour, proliferating epidermoid cyst, trichochlamydocarcinoma, hydatidiform keratinous cyst and invasive hair matrix tumour [5]. Classification of PTT is divided into three categories namely benign, locally aggressive, and malignant [6]. In 1969, Pinkus determined that the TCs develop from the sheath of the outer hair root located at the isthmus part of the hair follicle and also stated that the cells in this sheath experience growth due to changes in genes, although the exact types of gene changes are still unknown [7]. The PTCH1 gene could be associated with the development of this condition [8].

The World Health Organisation's Pathology and Genetics of Skin Tumours (WHO, 2006) listed them as lumps with hair follicle differentiation [9]. Rare instances of malignant transformation can be seen in TC [10], and if such transformation happens, it may result in distant metastases. TC lesions are frequently multiple, although some can be solitary [11-13]. These lesions typically occur in areas rich in hair follicles and where there is active hair growth. Over 90% of them are located on the scalp, with a small number found in the vulva and bulbar conjunctiva [14]. TC is more prevalent among middle-aged individuals, particularly women [15]. From a clinical standpoint, the disease progresses slowly, and most lesions appear as rounded, hard, and raised masses, without surface skin or hair involvement. CT scans or ultrasounds are useful for supplementary examination, while a postoperative pathological assessment is necessary for diagnosis, yielding a favourable prognosis [16]. Histopathologically, the lesions frequently show either sheet-like or arenaceous patterns of calcification [17]. The epithelial cells that line the cyst wall exhibit rapid keratosis and a granular layer deficient [18]. The cysts have well-defined boundaries with limited blood supply and are solid, exhibiting no cystic degeneration when observed under the naked eye. Zhu Z et al., from China, reported a case of scalp TC in a 41-year-old patient, in which there were multiple lumps (swellings) and smaller in size, whereas the swelling was solitary and larger in our case [19]. Jha AK et al., from India reported a case of painful multiple TC on the scalp in a young male, where a skin punch has been used as a therapeutic method, whereas the cyst was solitary, painless and excision of the whole cyst has been used as a therapeutic method in our case [20]. Jiang XL et al., reported a 27-year-old female patient with a palpable, painless mass at the scalp with pathologic findings suggestive of a hyperplastic external hair sheath scalp tumour (low-grade malignancy), whereas the pathological findings were suggestive of PTT with squamous differentiation (benign condition) in our case [21].

CONCLUSION(S)

A TC is an uncommon benign tumour of the skin, typically found in middle-aged women. Imaging techniques can be useful for confirming the cystic nature of the lesion and detecting other locations. The preferred treatment is complete surgical excision, and the diagnosis is primarily made through histopathological examination of the surgical specimen.

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