

Case Report

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Giant solitary trichoepithelioma masquerading as basal cell carcinoma

Surya Rao Rao Venkata Mahipathy^{1*}, Narayananamurthy Sundaramurthy¹,
Vimal Chander Rajamanohar², Alagar Raja Durairaj¹, Manimaran Ramachandran¹

¹Department of Plastic and Reconstructive Surgery, ²Department of Pathology, Saveetha Medical College and Hospital, Kanchipuram District, Tamil Nadu, India

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***Correspondence:**

Dr. Surya Rao Rao Venkata Mahipathy,
E-mail: surya_3@hotmail.com

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ABSTRACT

Trichoepithelioma is a rare benign adnexal tumour which can be of solitary non-familial type or multiple familial trichoepitheliomas. Here authors describe a middle-aged patient who presented with a swelling of the left nasolabial region diagnosed clinically as a basal cell carcinoma but proved to be a giant solitary trichoepithelioma (GST) following histopathological examination. This case is presented due to the rarity and the difficulty encountered in diagnosis of the case.

Keywords: Adnexal, Benign, Immunohistochemistry, Rare

INTRODUCTION

Trichoepitheliomas (TEs) are mostly hamartomas of hair germ cells that are poorly differentiated. Scalp, forehead, nasolabial folds and upper lip are common sites.^{1,2} TEs are of three types namely, solitary, multiple and desmoplastic. Multiple familial trichoepithelioma (MFT) are small papules found around nose and malar regions of face. The onset is usually before twenty years.¹ They are usually benign with a very rare potential to become malignant, TE may mimic basal cell carcinoma (BCC) both clinically and in sections.²⁻⁴ There are very few immunohistochemical stains differentiate between TE and BCC.⁵ The treatment for TE is excision with minimal margin whereas for BCC, it requires wide local excision with skin resurfacing.

CASE REPORT

A 54-year-old male patient presented to us with a blackish swelling of the left side of the face for the past three months. There was no history of trauma, bleeding,

ulceration or a previous mole at the site of lesion. The lesion was spontaneous in onset. There were no co-morbid illnesses. On examination, a 3 x 3cm dark brown lesion was present along the left nasolabial fold. It was irregular in shape with distinct borders and raised edges.



Figure 1: Pre- operative photograph showing the lesion.

The lesion was firm and non-tender (Figure 1). There was no cervical lymphadenopathy. A clinical diagnosis of basal cell carcinoma was made.



Figure 2: Intra- operative photograph showing the defect covered with an oblique forehead flap with primary closure of the secondary defect.



Figure 3: Photograph after flap division.

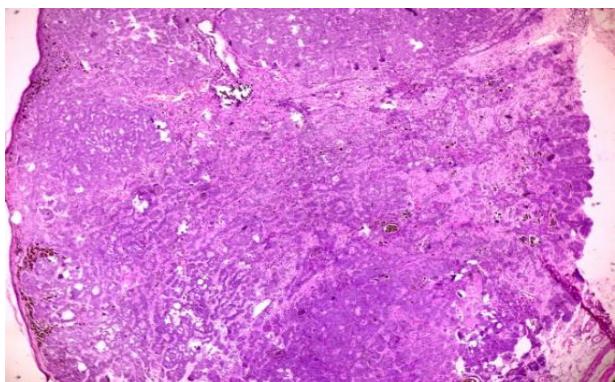


Figure 4: Section shows thinned out epidermis with underlying dermis showing a neoplasm composed of nests, cords and trabecular pattern of cells with focal areas forming tubules. H and E x40.

The plan was surgical excision with a flap cover. After obtaining anaesthetic fitness and consent, authors proceeded with wide local excision with a 5mm margin

and the result defect was covered with a pedicled peninsular oblique forehead flap with primary closure of the secondary defect (Figure 2). After 3 weeks and the flap being well settled, the pedicle was divided (Figure 3).

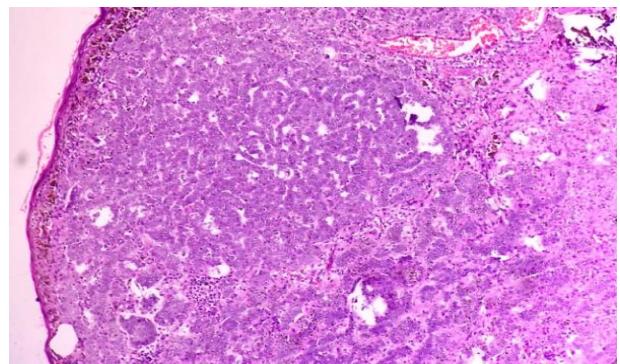


Figure 5: Section shows thinned out epidermis with underlying dermis showing a neoplasm composed of nests, cords and trabecular pattern of cells with focal areas forming tubules. H and E x100.

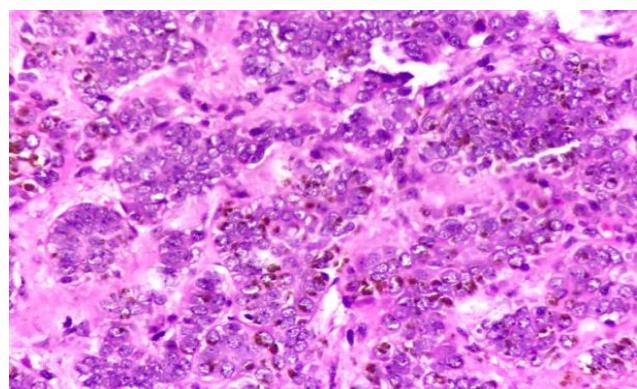


Figure 6: Nests, cords and trabecular pattern of cells with moderate eosinophilic cytoplasm and uniform round to oval vesicular nuclei. Some of the cells show melanin granules in the cytoplasm. H and E x400.

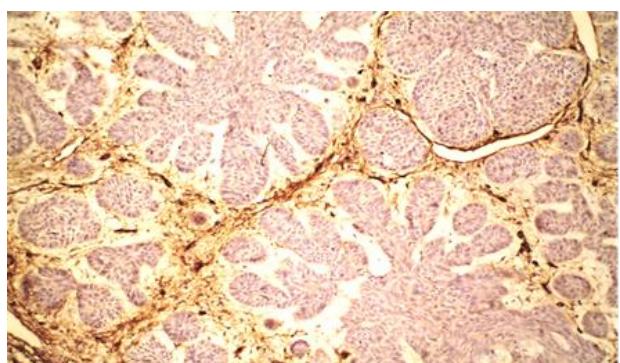


Figure 7: CD 10 positive for stromal cells and negative for tumour cells.

Histopathology revealed stratified squamous epithelium with adnexal structures and a lesion beneath composed of

lobules and nests of cells having scant to moderate eosinophilic cytoplasm and oval nuclei. There are a few ductal structures within and pseudo horn cysts scattered throughout. The stroma is fibrous with minimal inflammation. Mild increase in mitotic activity is noted with no evidence of deeper invasion (Figure 4,5,6). Immunohistochemistry showed Ki 67 positive (intense nuclear staining in 10-15% of tumour cells) and CD 10 negative in tumour cells with features suggestive of trichoepithelioma (Figure 7).

DISCUSSION

TE, when solitary usually is not familial.² MFTs are autosomal dominant affecting chromosome 9p21 with lesser expression and penetrance in a male.^{1,6} The lesions of TE are characterized by firm skin-colored papules or nodules 2 to 8 mm in size, mainly concentrated around the nasolabial folds and forehead. The lesions first appear in childhood and may grow larger and increase in number over time. In the present case the patient and her son had similar lesions all over the face since childhood, thereby suggesting familial disease. Clinically, the three forms of TE are a small solitary form, a small multiple form, (autosomal-dominant) and a rare giant solitary form.⁷ Multiple TE and GST can coexist.⁸ GST has been defined as a solitary trichoepithelioma with a diameter of 2 cm or more. TEs are superficial lesions within the dermis.¹ Small keratinous cysts lined by basaloid cells are the most characteristic histologic feature. Also, present are islands of uniform basaloid cells, showing peripheral palisading. The fibroblasts encircle these islands, which lack retraction artifact typical of BCC. These solid aggregates show invaginations which contain numerous fibroblasts and resemble follicular papillae also known as papillary mesenchymal bodies.

Table 1: Immunohistochemical markers to differentiate TE and BCC.

Markers	Tricho-epithelioma	Basal cell carcinoma
Bcl2 expression		
Epithelial	Positive in outermost layer	Diffusely positive
Stromal	Negative	Negative
CD 10 expression		
Epithelial	Negative	Positive
Stromal	Positive	Negative
CD 34 expression		
Epithelial	Negative	Negative
Stromal	Positive	Negative
EMA expression		
	Negative	Positive in areas of squamous differentiation with Squamo- BCC
PHLDA 1	Positive	Negative
CK 20	Positive	Negative

TE mostly causes cosmetic concerns only but rarely BCC can development can be associated with it.^{2,4} Histologically, the presence of horn cysts, papillary-mesenchymal bodies, lack of atypia and mitoses along with a cribriform pattern and stromal fibrosis favor a diagnosis of TE, whereas the presence of mucin, stromal edema and retraction artifact around the basaloid islands and mitoses suggest a diagnosis of BCC. The differences between TEs and BCCs in immunohistochemistry are shown in the Table 1.^{7,8}

The close relationship between TE and BCC is due to their common origin from from pluripotential cells like primary epithelial germ cells. Cylindromatosis tumor suppressor gene (CYLD) mutation is also seen in MFT.³ Brooke-Spiegler syndrome is associated with mutations of the CYLD gene causing multiple cylindromas and TEs.⁹

Possible malignant transformation in the form of ulcer formation or rapid growth warrants an excision biopsy of the lesion.¹⁰ TE is a benign lesion that may be excised with a small margin of healthy tissue, thereby facilitating surgical repair; however, BCC is the locally malignant tumor treated by excision with 3-4 mm margins. Hence, the differentiation between both the lesions is mandatory.

CONCLUSION

Solitary trichoepithelioma is a rare trichogenic tumor with potential for local recurrence. It may present at any age including at birth with predilection for old age. It has close resemblance to BCC and other skin adnexal tumors - clinically, cytologically, and histologically. CD10, CD34 and PHLDA1 are useful adjunct markers. Surgical excision is the standard treatment. Recurrence and possible transformation to BCC cautions follow up at regular intervals.

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Ethical approval: Not required

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