

Case Report

Multiple familial trichoepitheliomas: a case report

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ABSTRACT

Trichoepithelioma is a rare benign adnexal tumor that differentiates towards the folliculo-sebaceous-apocrine unit (trichoblast). It may present as solitary non-familial lesion or multiple lesions as a part of autosomal dominant inherited syndrome known as multiple familial Trichoepithelioma. Multiple familial trichoepithelioma is a relatively rare, disfiguring, benign adnexal neoplasm diagnosed by centropal distribution of papules and nodules, positive family history, related histopathological findings and can rarely undergo malignant transformation. Treatment is mainly for cosmetic concern. Here we report a case who presented with multiple skin coloured facial papules and nodules, with a history of similar lesions in other family members. Dermoscopy and histopathology confirmed the diagnosis of trichoepitheliomas.

Keywords: Brook-fordyce disease, Benign adnexal tumor, Basaloid cells, Dermoscopy

INTRODUCTION

Trichoepithelioma is a rare, benign tumor of the pilosebaceous unit, and it originates from the hair follicles.¹ It is divided into three subgroups Multiple Familial Trichoepithelioma (MFT), solitary non-hereditary trichoepithelioma and desmoplastic trichoepithelioma. MFT is also known as Brook-Fordyce disease.² Brook and Fordyce first described inherited multiple trichoepitheliomas in 1892 under the names 'multiple benign cystic epithelium' and 'epithelioma adenoids cysticum' respectively. MFT can also present in Brooke-Spiegler syndrome characterized by different combinations of inherited adnexal neoplasms including multiple trichoepitheliomas, cylindromas and occasional spiradenomas.

CASE REPORT

A 23 years old male presented with multiple, skin coloured, raised lesions over the nose, forehead and scalp.

Lesions started at the age of 10 years and continued to appear till date. Systemic examination was within normal limits. On dermatological examination, he had multiple, well defined, grouped and few discrete, skin coloured to yellowish, dome shaped papulo-nodular lesions with smooth surface, firm in consistency and measuring 2-5 mm in size, over the forehead, nose and nasolabial fold (Figure 1).

Few similar lesions were noted over the scalp. On dermoscopy multiple milium like cysts and arborizing pattern of vessels are noted (Figure 3). Skin biopsy was consistent with the clinical diagnosis of trichoepithelioma showing a benign neoplasm composed of basaloid epithelial cell nests with peripheral palisading, without retraction artefacts, surrounded by fibrous stroma with few horn cysts and papillary mesenchymal bodies (Figure 2). There was a history of similar lesions in the mother, maternal grandfather and maternal uncle (Figure 4).



Figure 1: Showing multiple centrofacial papulonodular lesions in the patient.

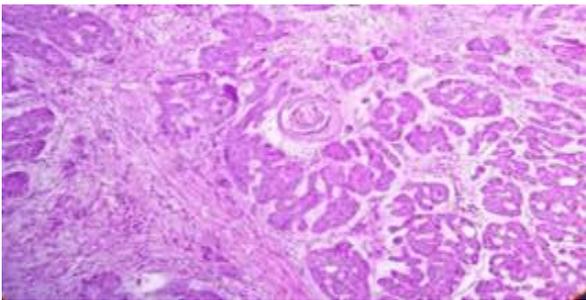


Figure 2: Histopathology showing lobules of small, dark basaloid cells, with peripheral palisading surrounding a central area of eosinophilic amorphous material.

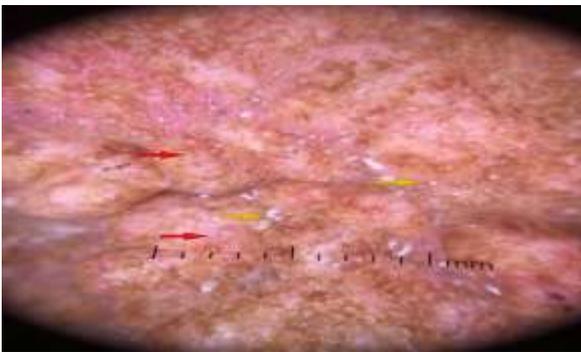


Figure 3: Dermoscopy with polarized×10 shows multiple arborizing vessels (red arrows), multiple milia like cysts (black arrows).



Figure 4: showing multiple trichoepitheliomas over the face in mother and maternal grandfather.

DISCUSSION

MFT is an autosomal dominant disorder caused by mutation in CYLD gene on chromosome 9p21 and 16q12-q13.² It presents with multiple skin coloured, pink or bluish, firm, rounded, translucent, shiny, well demarcated papules and nodules, center slightly depressed or umbilicated. It occurs predominantly on the face, particularly around the nasolabial folds, nose, forehead and eyelids, occasionally involves scalp, neck and upper trunk.³ Lesions are usually 2 -5 mm in diameter and may increase in size and number over years. Dilated telangiectatic blood vessels may be seen over the surface of large lesions.

Dermoscopy of the papules shows milia like cysts and arborizing vessels on a whitish background, which was consistent with our case.⁴ Histopathology typically shows horn cysts, tumour islands composed of basaloid cells arranged in peripheral palisading pattern². It is an asymptomatic condition. Transformation into malignant neoplasms can occur rarely – such as trichoblastic carcinoma or basal cell carcinoma.

Treatment is mainly for cosmetic concern. Various treatment modalities which have been tried include non-pharmacological methods such as excisional surgery, laser resurfacing (erbium:Yag and CO₂), chemical cauterization, electrosurgery, cryosurgery, dermabrasion and pharmacological methods such as topical 5% imiquimod cream (5 to 7 times/week).⁵ Aspirin orally 325 mg twice daily combined with Adalimumab (s.c injection for first 2 months 40 mg every other week and thereafter 40 mg every week for 8 months)⁶, 13 cis retinoic acid 1 mg/kg/day orally for 12 weeks and corticosteroids.

CONCLUSION

Trichoepithelioma can manifest as solitary non-familial type or multiple familial type. MFT is a relatively uncommon disease and high index of suspicion is needed to make correct diagnosis.

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