Brook–Fordyce disease in an Indian family: A case report

Sukhmani Brar¹, Meenal Makkar²

¹Consultant Dermatologist at Fortis Polyclinic, Chandigarh, India, ²Post graduation resident at Guru Gobind Singh Medical College & Hospital, Faridkot, Punjab, India

Corresponding author: Dr. Meenal Makkar, E-mail: meenal.makkar@gmail.com

ABSTRACT

Trichoepithelioma is an uncommon, benign, cutaneous tumor that originates from hair follicles. Trichoepitheliomas can be of solitary non familial or multiple familial type (Brook-Fordyce disease). They usually present on face as skin colored papules. We report here on a 27 year old female presented with 15 years history of multiple, asymptomatic, skin colored papular lesions mainly over central part of the face. History of similar presentation was there in 12 family members within 4 generations. Histopathological findings were consistent with the clinical diagnosis of trichoepithelioma. Patient considered her skin condition as a hurdle for her marriage which led to major psychological impact in patient and also in parents. Patient was successfully treated with staged Radiofrequency ablation followed by topical retinoid. Multiple familial trichoepitheliomas is a relatively uncommon disease and produces cosmetic disfigurement. Management is mainly for cosmetic concern. There is very rare risk of malignant transformation to Basal cell carcinoma.

Key words: Multiple trichoepitheliomas; Familial; Benign

INTRODUCTION

Trichoepithelioma is an uncommon, benign, cutaneous tumor that originates from hair follicles. It can be of solitary non familial or multiple familial type (Brook–Fordyce disease) [1]. They usually present on face as skin colored papules. Multiple trichoepitheliomas are also present in Rasmussen syndrome, Rombo syndrome and Brooke-Spiegler syndrome (BSS) [2-4]. Literature review showed that only 13 cases were reported up till 2015 with the first case in 1959 [5].

CASE REPORT

We report here on a 27 year old female presented with 15 years history of multiple, asymptomatic, skin colored raised lesions mainly over central part of the face (Fig. 1). Lesions started from 12 years of age and progressed till 19 years. Rest of the body parts is spared. History of similar presentation was there in 12 family members within 4 generations (Figs. 2 and 3).

On examination multiple skin colored, well defined papules and nodules of size ranging from 0.1 to 1cm were present over face. Histopathology showed presence of nest of basaloid cells with peripheral palisading surrounding a central area of eosinophilic amorphous material. They surrounded by stroma with increase no. of fibroblasts (Fig. 4). Few horn cysts are also present. These findings were consistent with the clinical diagnosis of trichoepithelioma. Patient considered her skin condition as a hurdle for her marriage which led to major psychological impact in patient and also in parents. Various treatment options were discussed with the patient. She opted for Radiofrequency ablation and resurfacing. Complications like scarring and recurrence of lesions post procedure were explained the patient prior to signing an informed consent. The patient underwent four sessions of Radiofrequency ablation, with two weeks interval to completely remove all lesions. Topical tretinoin 0.05% cream and sunscreen were used as maintenance treatment (Fig. 5). Patient was in our follow up till six months with no recurrence.

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Figure 1: Multiple centrofacial papulo-nodular lesions in the index case.

Figure 4: Histopathology showing nest of basaloid cells with peripheral palisading surrounding a central area of eosinophilic amorphous material (H and E X 10).

Figure 2: Multiple trichoepitheliomas over face in the brother.

Figure 5: Post treatment photograph of index case.

Figure 3: Multiple trichoepitheliomas in the paternal aunt of patient.

DISCUSSION

In 1892, Brooke originally described it as epithelioma adenoids cysticum (EAC) and Fordyce as a multiple

benign cystic epithelioma [6]. It is an autosomal dominant disease but more common in females due to lessened expressivity and penetration in males [7]. In our report also eight were females and four males which is correlating with female preponderance observed in literature. There is a genetic heterogeneity of multiple familial trichoepitheliomas (MFT). Initial reports linked MFT to chromosomes 9p21 but recent reports have found mutation in cylindromatosis tumor suppressor gene (CYLD), which maps to chromosome 16q12-q13 in most of cases. Treatment options are surgical excision, chemical cauterization, dermabrasion, laser resurfacing, topical retinoids and 5% imiquimod cream. Some reports suggest the use of adalimumab and aspirin as latest modalities. Surgical removal of multiple tumours is unfeasible. CO2 laser resurfacing can be employed as a destructive method, but it is associated with prolonged recovery time and

oozing. Radiofrequency devices can overcome the disadvantages of pre-existing laser devices by offering enhanced tissue penetration and a more accurate focus [8].

CONCLUSION

MFT is a relatively uncommon disease and produces cosmetic disfigurement. Management is mainly for cosmetic concern. There is very rare risk of malignant transformation to Basal cell carcinoma.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

REFERENCES

1. Karadag AS, Ozlu E, Zemheri E, Ozkanli S. Multiple papulonodular lesions located over the nasolabial sulcus. *Indian Dermatol Online J.* 2014;5(Suppl 1):S56-7.
2. Rasmussen JE. A syndrome of trichoepitheliomas, milia, and cylindromas. *Arch Dermatol.* 1975;111:610-4.
3. Michaelsson G, Olsson E, Westermark P. The Rombo syndrome: a familial disorder with vermiculate atrophoderma, milia, hypotrichosis, trichoepitheliomas, basal cell carcinomas and peripheral vasodilation with cyanosis. *Acta Derm Venereol.* 1981;61:497-503.
4. Kaur T, Puri KJ, Chahal KS, Budhwar J. Multiple Familial Trichoepitheliomas: A case report and. *Egypt Dermatol Online J.* 2012;8:11.
5. Snekszer M. Simultaneous occurrence of familial cystic adenoidal epithelioma (trichoepithelioma) and basalioma. *Borgyogy Venerol Surg.* 1959;35:282-5.
6. Brooke HG. Epithelioma adenoids cysticum. *Br J Dermatol.* 1892;4:269.
7. Malakar S, Mukherjee SS. Dermoscopic description of trichoepithelioma in the skin of colour. *Our Dermatol Online.* 2018;9:335-6.
8. Hong JY, Seok J, Kim JM, Jang YJ, Kim BJ. Successful treatment of trichoepithelioma with a novel insulated, monopolar, radiofrequency microneedle device. *Clin Exp Dermatol.* 2018;43:108-9.

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