Case Reports

Multiple Trichoepithelioma

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ABSTRACT

Multiple non-familial Trichoepithelioma is a rare benign adnexal tumour that originates from pilosebaceous follicle. The incidence is predominantly at puberty or early part of childhood. The common sites of occurance of trichoepithelioma are upper lip, forehead, scalp and nose. Trichoepithelioma in young women may lead to cosmetic disfigurement. We report a case of multiple non-familial trichoepithelioma who presented with complaints of multiple skin coloured raised lesions on her face since the age of 15 years.

Keywords: Benign adnexal tumour; horn cyst; multiple non-familial trichoepithelioma.

INTRODUCTION

Multiple trichoepithelioma is a tumour of skin that is both benign and rare originating from proliferation of epithelial-mesenchymal cells (1-3). The common sites include scalp, nose, upper lip and forehead. The autosomal dominant type manifests as multiple lesions while the sporadic type presents as solitary skin coloured papule (4). The tumour mostly appears in early childhood or at puberty. Multiple trichoepithelioma emerge as multiple skin coloured firm papulo-nodular lesions, which gradually increase in number producing significant cosmetic disfigurement. Malignant transformation to basal cell carcinoma is a rare complication (5).

Till date, only few cases of multiple non familial trichoepithelioma have been reported. We report here a rare case of multiple non-familial trichoepithelioma.

CASE REPORT

A 28 year old female patient presented with complaints of multiple skin colored raised lesions on her face since 15 years of age. The lesions initially started on her chin and then gradually progressed to involve the nose, forehead and cheeks. There was no history of topical application, no history of photosensitivity, no history of any drug intake. She did not give history of identical lesions elsewhere in the body. There was no significant family history.

Dermatological examination showed multiple well-defined, discrete, skin coloured papules 1-3mm in size with smooth surface involving her forehead, nose, nasolabial folds, upper lip and cheeks. On palpation, lesions were non-tender and firm in consistency. Scalp, oral mucosa, palms, soles, nails and genitalia were normal. Systemic examination was found to be normal.

Skin biopsy was done to confirm the clinical diagnosis. Section showed keratinized stratified squamous epithelium of skin with adnexal structure with an underlying circumscribed benign neoplasm composed of nest of cells with scant to moderate eosinophilic cytoplasm and uniform round to oval bland nuclei with focal areas showing horn cyst depicting abrupt type of keratinization. No necrosis, mitosis, atypia noted. Features suggestive of Trichoepithelioma.

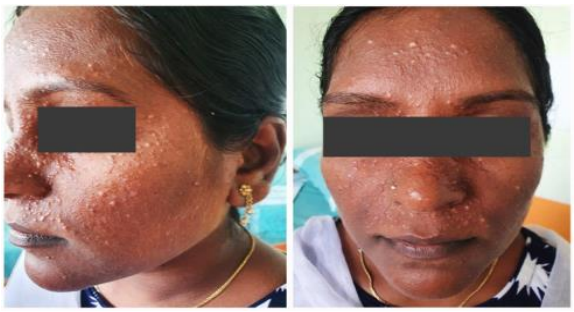

Fig. 1: Multiple well defined, discrete, skin coloured papules 1-3mm in size with smooth surface involving her forehead, nose, nasolabial folds, upper lip and cheeks.

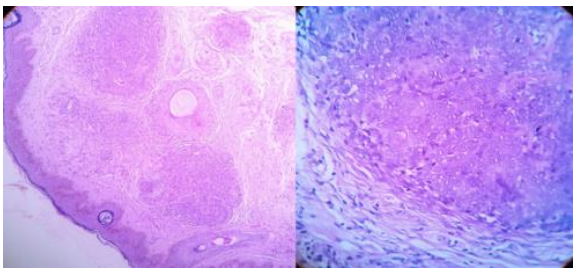

Fig. 2: Histopathology: Hematoxylin and Eosin stain- horn cyst and aggregation of basaloid cells surrounded by fibrous stroma.

DISCUSSION

Trichoepithelioma (Brooke Fordyce trichoepithelioma, epithelioma adenoides cysticum of Brooke, multiple benign cystic epithelioma, Trichoepithelioma papulosum multiplex) is a benign neoplasm originating from folliculosebaceous units.

Trichoepithelioma can present as a solitary tumour or as multiple lesions. Solitary trichoepithelioma is more common than multiple trichoepithelioma. It is not inherited. It presents clinically as a firm flesh colored nodule of size less than 2cm.

Trichoepithelioma has a variant termed as Giant solitary trichoepithelioma that is generally observed in the perianal region and thighs.

Multiple familial trichoepithelioma is an autosomal dominant condition associated with mutation of CYLD gene on chromosome 9p21 (6, 7). It predominantly affects the face involving the nose, nasolabial folds, periorbital area and scalp.

Multiple non familial trichoepithelioma is characterized by asymptomatic discrete well defined firm translucent papulonodular lesions with symmetrical distribution over the face, nose, upperlip, forehead, scalp, neck and trunk. It gradually increases in number and size causing cosmetic disfigurement. Rombo's syndrome and Basex syndrome amongst other rare syndromes encompasses Multiple trichoepithelioma. Rombo's syndrome also includes vermicular atrophoderma, peripheral vasodilation with cyanosis, hypotrichosis, milia, BCCs while Basex Syndrome includes hypotrichosis, hypohidrosis, BCCs and follicular atrophoderma (8).

Brooke Spiegler Syndrome is a rare autosomal dominant disorder characterized by multiple skin appendageal tumours mainly trichoepitheliomas and other benign skin appendageal tumours like cylindromas, spiroadenomas. Limited form of the disease includes familial cylindromatosis, multiple familial trichoepithelioma. This syndrome may be rarely associated with BCC, follicular cyst, nevus sebaceous (9-11).

Few other cases of MTE have been reported. 50y/F with multiple skin coloured papular and nodular lesions of varying sizes involving the face eyelids and ears since the age of 3 years with no family history has been reported in Haryana (5).

A case of unilateral multiple non-familial trichoepithelioma since the age of 5 years has also been reported in a 34y/F (12). A case of MTE without family history has been reported in a 9 year old child who presented with asymptomatic papules since the age of 5 months (13). A case of multiple non-familial trichoepithelioma has been reported in a 30y/F presented with multiple grouped skin coloured papules and nodules on her face including ear leading to facial disfigurement and hearing loss (14). A 6 year old girl child presented with 3 years history of hemifacial plaque of confluent nevoid trichoepithelioma with no family history (15). A case of unilateral nevoid non familial trichoepithelioma in a 14 year old boy presented with a plaque composed of confluence of multiple skin colored papules and nodules on the left side of the neck (16).

In our case, patient developed asymptomatic papules bilaterally distributed on the face around puberty and there are no nodular lesions. No history of similar complaints in the family.

Trichoepithelioma may be confused with milia, acne vulgaris, acneiform eruptions, syringoma, adenoma sebaceum, molluscum contagiosum, papular sarcoidosis, lupus miliaris disseminates faciei, microcystic adnexal carcinoma, basal cell carcinoma. Diagnosis is confirmed by histopathological examination in which horn cysts are the most characteristic histologic feature. It consists of fully keratinized centre surrounded by basophilic cells without atypia and mitosis. The keratinization is abrupt and complete. There may be few layers of cells with eosinophilic cytoplasm and large oval pale vesicular nuclei between the basophilic cells and horn cysts. Another major component is tumour islands composed of basophilic cells arranged in a lacelike or adenoid network with peripheral palisading of cells surrounded by stroma with increased fibroblasts. Fibroblasts are tightly associated with basaloid islands lacking retraction artefact. Adenoid network show invagination containing numerous fibroblasts and resemble follicular papillae, also known as papillary mesenchymal bodies. These basophilic cells lack mitosis, atypia and fibroblasts lack artifactual retraction which are characteristic features of BCC.

Although this condition is benign and asymptomatic, patients may seek treatment due to cosmetic disfigurement. Various modalities of treatment options have been proposed for the management of trichoepithelioma. These include surgical excision, dermabrasion, erbium: Yag and carbondioxide laser, chemical cauterization, cryosurgery, topical 5 % imiquimod cream and retinoic acid. Selection of proper choice of treatment depending on the depth of lesions is crucial to avoid over treatment. Scarring can occur after treatment. Partial removal may result in persistence or recurrence.

CONCLUSION

Trichoepithelioma is a benign hamartomatous tumour originating from folliculosebaceous unit. Multiple trichoepithelioma is not very common and diagnosis is usually associated with a strong family history but in this case it is sporadic which is a rare presentation.

CONFLICT OF INTEREST

Authors declare that there are no conflicts of interest.

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