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Multiple Non- Familial Trichoepitheliomas in a Nine Year Old Child

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Abstract

Trichoepitheliomas are rare benign tumours of poorly differentiated trichogenic origin. They present as translucent lesions most commonly on centrofacial regions. Solitary lesions are seen in sporadic cases while multiple lesions are inherited in autosomal dominant pattern. We present a nine year old child with multiple trichoepitheliomas at classical sites with none of the other family members involved.

Key words: Adalimumab; Genes, Hair Follicle; Suppressor

Introduction

Trichoepitheliomas (TE) are uncommon benign tumour arising from basal cells of pilosebaceous follicle. These were first described by Brooke in England in 1892 as “epithelioma adenoids cysticum” and in the United States by Fordyce as “multiple benign cystic epithelioma”.^{1,2} They may be solitary or multiple. Face is the commonest site involved. Multiple TE are usually inherited as an autosomal dominant disease. Sporadic cases usually have solitary lesions, there are only few cases of non-familial multiple TE reported.³

Case report

A nine-year-old male child presented to the dermatology outpatient department with the complaint of multiple asymptomatic lesions over face since 5 months of age, progressively increasing in size as well as in number. There was no history of seizures, any other neurological involvement or systemic complaint. There was no history of similar lesion in any of the two siblings or family. He was born by normal vaginal delivery to non-consanguineous parents and had no developmental, physical or intellectual impairment. On mucocutaneous examination he had multiple waxy, skin coloured, translucent, dome shaped papules and nodules ranging in size from 2 to 8 mm,

clustered over centrofacial region bilateral nasolabial folds and medial periorbital region with sparing of tip of nose. (Figure 1)

Few discrete papules were also present over chin, submental area and lateral neck. The lesions were soft to firm in consistency, non-tender and had no surface changes, umbilication, hair tuft or telangiectasia. There were no other significant cutaneous lesions. Systemic examination was within normal limits. Child was diagnosed to be having multiple non-familial trichoepithelioma. The differential diagnosis of adenomasebaceum, juvenile colloid milium and trichofolliculoma was kept. The histopathology from popular lesion revealed well circumscribed islands and solid aggregates of basaloid cells in the superficial dermis with presence of true horn cysts and papillary mesenchymal bodies (Figure 2,3).

After clinicopathological correlation diagnosis of multiple non-familial trichoepithelioma was made.

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Child was referred under school health scheme and was from poor family, lesions were asymptomatic so parents refused for any medical or surgical intervention. They were counselled about the malignant transformation of the disease and were advised regular follow up.

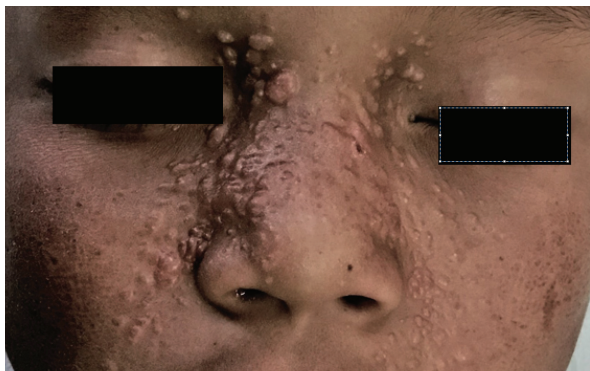


Figure 1: Skin coloured , waxy papulonodular lesions in centrofacial area.

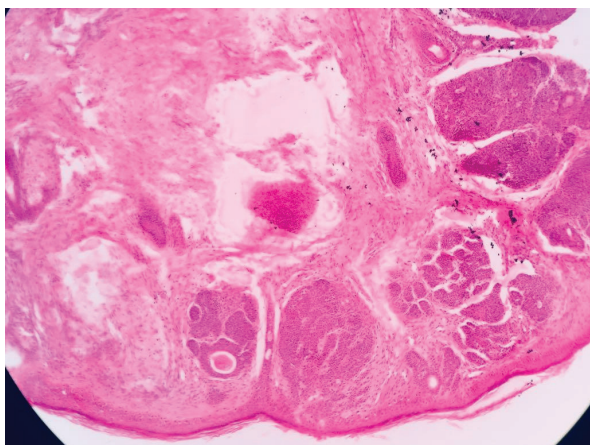


Figure 2: (H&E 40 X) Histopathology showing well circumscribed islands and solid aggregates of basaloid cells in the superficial dermis with presence of true horn cysts and papillary mesenchymal bodies.

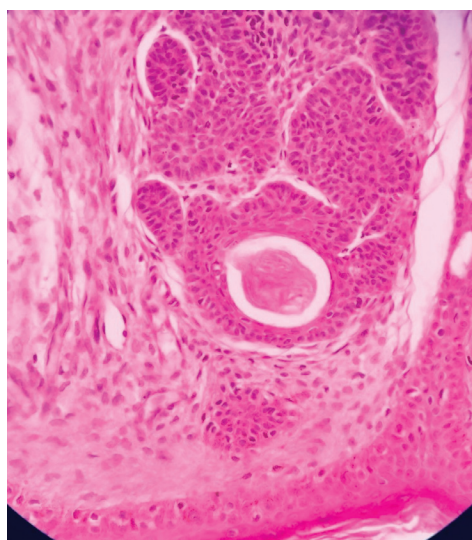


Figure 3: (H&E 400X) histopathology showing true horn cysts and papillary mesenchymal bodies.

Discussion

Trichoepitheliomas (TE) are rare benign hamartomas of the hair germ cells of unknown prevalence. Various synonymous terms e.g., trichoepithelioma papulosum multiplex (TPM), epithelioma adenoides cysticum, acanthoma adenoides cysticum, multiple benign cystic epithelioma, Brooke's tumour or Brooke-Fordyce trichoepithelioma are used for TE.⁴ They are classified as solitary sporadic, multiple familial (MFT) and desmoplastic.⁵ MFT is autosomal dominant genodermatosis. Multiple sporadic TE have been rarely described, with Sehrawat et al in 2016 reporting the first case.³ Our case had multiple TE with no family history of similar complaint.

Clinically, multiple TE is characterized by discrete, skin coloured to erythematous translucent well defined papules and nodules predominantly over face clustered over nose, nasolabial folds, periorbital area and scalp. Multiple lesions are symmetrically distributed all over the face. TE at other body sites have rarely been reported.⁶ The lesions are usually 2-5 mm in diameter. The disease usually starts in early childhood and may increase during puberty. They may have surface telangiectasia, central umbilication but there is no ulceration.

Clinically multiple TE have to be differentiated from molluscum contagiosum, multiple trichofolliculoma, adenoma sebaceum, basal cell nevus syndrome, juvenile colloid milium, trichoadenomas, histoid leprosy. Molluscum contagiosum lesions are pearly white papules with central umbilication and not symmetrical and show a tendency towards spontaneous regression with evidence of pseudo-koebnerization. Adenoma sebaceum are angiomatous and may show other findings of tuberous sclerosis. Trichofolliculomas are usually solitary but multiple lesions have also been described. A central tuft of vellus hair may be seen. Histoid leprosy will have associated neurological involvement and can be differentiated histopathologically. Basal cell nevus syndrome will also have associated stigmata of disease. Juvenile colloid milium may have similar presentation to TE but on puncturing the lesion gelatinous material is extruded.

Histopathology is same for solitary and multiple TE and is characterised by islands of tumour cells and horn cysts. Horn cysts consist of fully keratinized centre surrounded by basophilic cells lacking atypia and mitosis. Occasionally, primitive hair papillae, as well as hair shaft-like structures, can be observed in these central areas. The tumour islands are made up of basaloid cells with peripheral palisading and increase

insurrounding fibrous stroma. TE may resemble basal cell carcinoma (BCC), trichoadenoma and trichofolliculoma histopathologically. Features which help differentiate TE from BCC are fibrocystic rather than mucinous stroma of BCC, low mitotic activity, aggregations of cells with smooth borders, lack of stromal retraction, monomorphic nuclei and papillary mesenchymal bodies (resembling primitive follicular papilla).⁷ Immunohistochemistry in BCC shows positivity for BCL-2 and is negative for CD34 whereas TE are CD34 positive and BCL-2 negative. Horn cysts may be seen in trichoadenoma but surrounded by cells resembling infundibular portion of hair follicle. Trichofolliculoma shows central dilated hair follicle surrounded by hair follicles in various stages.

Multiple trichoepithelioma as stated earlier are autosomal dominant but may be occasionally sporadic as in our case. Reduced penetrance in inheritance may be responsible for sporadic disease in our case. Mutation has been identified in cyclin D1 tumor suppressor gene (CCND1) which leads to defective apoptosis and hence tumorigenesis.⁸

Multiple TE have been found associated with multiple renal and pulmonary cysts, basal cell adenoma of the parotid gland, ovarian cancer, breast cancer, steatocystomas, alopecia and myasthenia gravis.⁴ Multiple TE also form a part of other rare syndromes like Rombo syndrome (vermicular vermiculate atrophoderma, milia, hypotrichosis, basal cell carcinomas, TE and peripheral vasodilation

with cyanosis) and Bazex syndrome (follicular atrophoderma, hypotrichosis, basal cell carcinomas, occasional trichoepitheliomas, and localized or generalized hypohidrosis).³ Brooke-Spiegler syndrome is characterized by multiple skin appendage tumours (e.g. cylindromas, spiradenoma, TE, etc.).

TE commonly leads to disfigurement. The transformation of multiple TE to basal cell carcinoma is rare and late, so regular follow up of patients is advised.

Treatment modalities include electro-surgery, cryosurgery, dermabrasion, excisional surgery, laser resurfacing, topical 5% imiquimod cream and tretinoin 1% gel. Pharmacological agents with inhibitory effects on NF- κ B functions, such as corticosteroids and nonsteroidal anti-inflammatory drugs (NSAIDs), aspirin, adalimumab have been tried.⁹

But as the lesions are asymptomatic and have very low malignant potential; treatment is usually not necessary. Patients who are concerned about disfigurement need to be treated.

Conclusion

We are presenting this case because of presence of clinical lesions in a child with no family history. So if we suspect trichoepithelioma inheritance pattern should be observed.

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Conflict of interest to disclosure: None declared.

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