INTRODUCTION

Multiple trichoepithelioma was described by Brooke-Spiegler in 1892 as epithelioma adenoides cysticum (Brooke's tumor) and by Fordyce as multiple benign cystic epithelioma. It is an autosomal dominant condition, the gene of which has been mapped onto chromosome 9p21.[1]

We report two cases of multiple trichoepithelioma in a family affecting the mother and her son.

CASE REPORTS

A 35-year-old female presented with multiple asymptomatic nodules over her face. She had noticed the first lesion over her left cheek at the age of 9 years. Thereafter, the lesions gradually increased in number and size with the increase more evident in the last 10 years. Lesions also appeared over the scalp and external ear canals with consequent discomfort in the right ear due to blockage of the auditory canal. However, there was no hearing defect. A few scattered lesions also appeared over the neck in the course of the last 4 years.

Her 13-year-old son also developed similar lesions over his face and scalp for 2 years, but they were less in number and size. There was no similar illness in any other family member.

On examination, the mother had multiple skin-colored firm papules and nodules ranging in size from 4 × 4 mm to 2 × 3 cm on her face. The lesions were distributed predominantly over the central face in a symmetrical pattern. Multiple lesions were seen over the scalp, and both ear canals were almost plugged with the lesions. A few small lesions were present over the neck, and there was a single lesion on her left forearm. Examination of her son showed multiple, similar but smaller, and discrete lesions over the face, mainly the central area. Histopathological examination of the nodule from mother showed multiple...
keratinized horn cysts surrounded by basaloid cells and islands of basaloid cells lacking retraction artifact, which was consistent with trichoepithelioma [Figures 1-3]. There was no atypia or increased mitosis.

**DISCUSSION**

Multiple trichoepithelioma or epithelioma adenoides cysticum usually has its onset at puberty as multiple, small, flesh-colored firm papules, nodules, and cystic lesions with a translucent sheen. They are symmetrically arranged on the face, especially in nasolabial fold, but may be located on the scalp, neck, and trunk. Lesions have also been described in the external auditory meatus and eye lids. Large lesions may be yellow or pink or sometimes bluish from pigmentation, and there may be dilated vessels over the surface. Individual lesions reach a limiting size, but the number may increase over the years. At times, they may be found in unusual configurations such as a large hemifacial plaque. Continued growth or ulceration raises the suspicion of change into basal cell carcinoma, although it is a very rare event. Most of the cases reported as such in past are now regarded as the instances of nevoid basal cell carcinoma syndrome.

The simultaneous presence of multiple trichoepithelioma and cylindroma has been observed repeatedly. This may be due to the phenotypic dimorphism and variable penetrance of the causative gene on chromosome 9. Multiple trichoepitheliomas have also been rarely associated with a number of systemic conditions such as Rombo syndrome which includes vermiculate atrophoderma, milia, hypotrichosis, basal cell carcinoma, and peripheral vasodilation, and also with diseases such as systemic lupus erythematosus and myasthenia gravis.

On histological examination, multiple trichoepitheliomas are seen as small, well-circumscribed, symmetric, superficial dermal lesions. The presence of horn cysts is the most characteristic feature with fully keratinized center and surrounding basophilic cells similar to basalioma cells in basal cell carcinoma, except that they tend to lack high-grade atypia and mitoses. The second major component is the basaloid epithelial islands, tightly encircled by fibroblasts, lacking the retraction artifact of basal cell carcinoma.

Trichoepithelioma may present clinically in other forms as well. It may present as a solitary smooth nodule on the face of about 0.5 cm size with no risk of malignant transformation. Second, it may present later in life as a very large polypoid lesion in the lower trunk, frequently in the perianal area, and is called solitary giant trichoepithelioma. A third presentation mimicking basal...
cell carcinoma also occur with lesions having a depressed center and a raised rolled edge. It is usually seen on the face and is known as desmoplastic trichoepithelioma or sclerosing epithelial hamartoma.

No modality of treatment gives satisfactory results in multiple trichoepithelioma. Surgery, electrodessication and curettage, X-ray irradiation, dermabrasion, or cryotherapy may all be followed by recurrences or by new lesions. If there is any suspicion of malignant change, adequate excision and histological examination are mandatory.

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Conflicts of interest

There are no conflicts of interest.

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