A Rare Case Of Familial Multiple Trichoepithelioma

B Poornima, P Kanthilatha, P Sathish

Citation

B Poornima, P Kanthilatha, P Sathish. *A Rare Case Of Familial Multiple Trichoepithelioma* . The Internet Journal of Dermatology. 2013 Volume 11 Number 1.

Abstract

Trichoepithelioma (TE) represent benign hamartomas of the pilosebaceous apparatus that may occur as a non-hereditary solitary form or as multiple lesions that is dominantly inherited. Multiple TE may represent a syndrome in which tumors develop from undifferentiated germinative cells of the pilosebaceous –apocrine unit.1 We report a case of multiple trichepithelioma in a 35 year lady.

INTRODUCTION

Occurrence of multiple trichoepithelioma appears to be uncommon. They commonly appear in childhood and puberty, and are described more commonly in females. It can run in families and the gene for multiple TE has been mapped to a locus on 9p21 . 2

CASE REPORT

A 35-year-old woman presented for advice because of multiple facial tumors, which had been present since many years. She was deeply concerned about them for cosmetic reasons. Physical examination showed numerous small pearly white papules involving the nasal root, the medial part of the eyebrows, and the nasolabial folds.

Figure 1

Face of patient

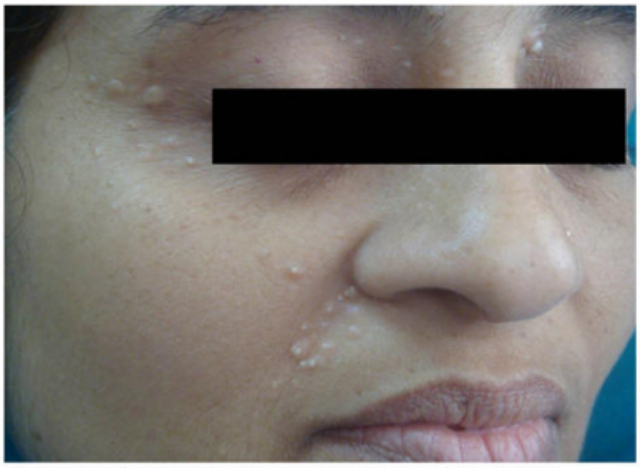

Fig 1: Picture shows multiple skin colored and pearly papules on the face

Histopathological examination of the lesion showed nests of bland appearing basaloid cells in the dermis with peripheral palisading, forming horn cysts. No mitotic figures were seen among tumor cells. Few of the nests showed follicular differentiation and an attempt to form abortive papillae.

Figure 2

Histology 100x

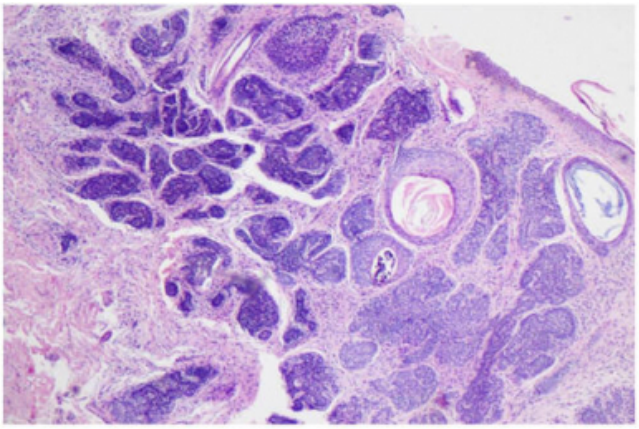

Fig 2: Photomicrograph shows nests of basaloid cells and horn cysts in dermis H&E,100X

Figure 3

Histology 200x

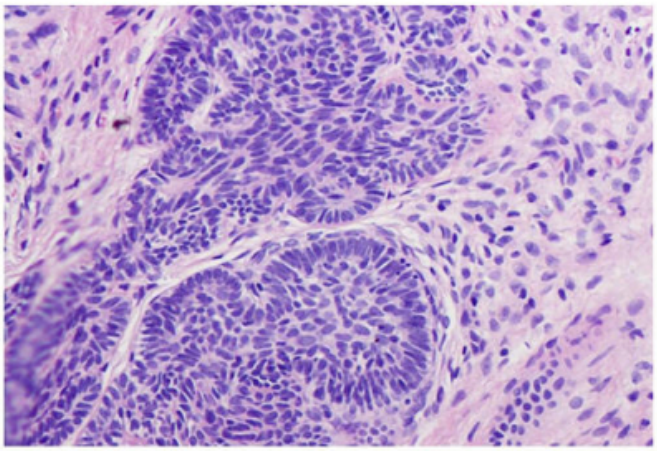

Fig 3 shows nests of bland appearing basaloid cells with peripheral palisading and follicular differentiation H&E, 200X

DISCUSSION

Multiple TE clinically present as multiple, firm, skin colored to pink papulo-nodular lesions commonly on the face, and sometimes on the scalp, neck and upper trunk. Patients present because of cosmetic disfigurement. Malignant transformation to basal cell carcinoma is rare, but can occur late in the course of the disease.3

The tumour cells form rudimentary hair follicles but do not form actual hair shafts and are characterized histopathologically by the presence of branching nests of basaloid cells, horn cysts, and abortive hair papillae. Histologic differentiation from keratotic basal cell carcinoma can be difficult.Absence of nuclear pleomorphism and infrequent mitosis help in differentiating trichoepithelioma from basal cell carcinoma.

Multiple TE needs to be differentiated clinically from milia, syringoma and molluscum contagiosum.

Family history revealed that the patient's daughter aged 10 years also had few small similar lesions on the face, hence possibility of a familial etiology is likely in our case.

Treatment is difficult and unrewarding. Surgical excision and other modalitilies like cryotherapy, dermabrasion, electrodissection and curettage have been tried with occasional good results.4

References

1. Clarke J, Ioffreda M, Hem KF .Multiple familial trichoepithelioma- a follicular sebaceous - apocrine genodermatosis. Am J Dermatopathol 2002; 23:402-5
2. Harada H, Hashimoto KY, Ko MSH.The gene for multiple trichoepithelioma to 9p21.
J Invest Dermatol 1996;107:41-43
3. Pariser RJ. Multiple hereditary trichoepithelioma and basal cell carcinomas. J Cut Pathology 1986;13:111-17
4. Sindu SK, Wakelin SH, Wilkinson JD. Multiple familial trichoepithelioma. Cutis 1999;63:239-40

Author Information

Baliga Poornima, Professor
Department of Pathology KMC International Center, Manipal University
Manipal, India

Pai Kanthilatha, Professor
Department of Pathology KMC International Center, Manipal University
Manipal, India

Pai Sathish, Professor
Department of Dermatology KMC, Manipal University
Manipal, India