

Case 6

A 36-year-old Thai woman from Bangkok

Chief complaint: Multiple skin color papules on her face for 30 years

Present illness: The patient presented with multiple asymptomatic small papules on her face for 30 years. The lesion gradually increased in number and slightly increased in size. Previous skin biopsy showed sebaceous gland hyperplasia and the lesions were treated with CO₂ laser, however new lesions still developed.

Past history: No underlying disease.

Family history: Her sister has similar lesions on her face

Skin examination:

Numerous 1 to 2 mm. slightly erythematous to light brownish papules on face.

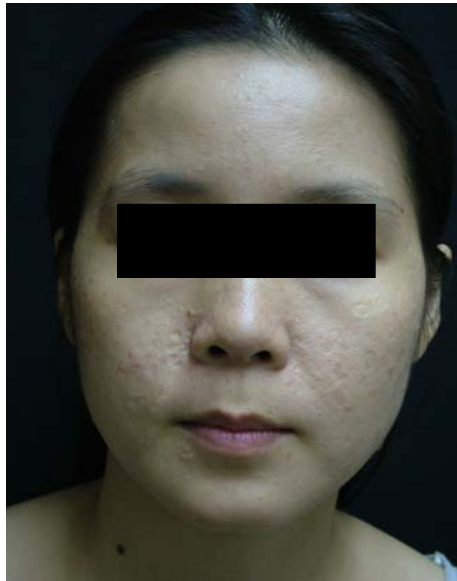


Fig. 6.1

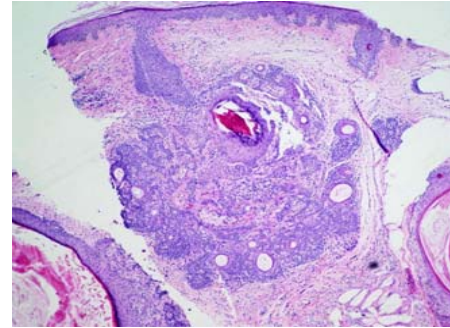


Fig. 6.2

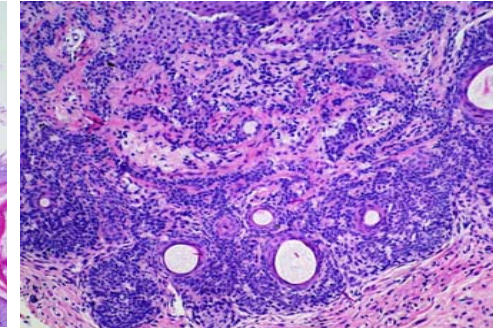


Fig. 6.3

Histopathology (S09-08836A, Fig 6.2, 6.3)

- Cribriform aggregates of basosquamous cells, some with follicular induction in the upper dermis.
- Many horn cysts, some ruptured with calcification.
- Dense fibrous stroma around the tumor.

Diagnosis: Multiple trichoepithelioma

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Discussion: Note

Trichoepithelioma are benign epidermal appendage tumor, mostly originating from follicular germinative differentiation which appear predominantly in childhood or in young adults. Three distinctive forms of trichoepithelioma are recognized, namely; solitary, multiple and desmoplastic.¹ Brook and Fordyce first described inherited multiple trichoepithelioma in 1982.²

Solitary trichoepithelioma occurs mainly as small (5-8 mm.) skin-colored papules on the face especially around the nose, upper lip and cheeks. Occasional lesions develop on the trunk, neck, scalp, extremities, buttocks and genital area. In multiple lesions (epithelioma

adenoids cysticum or Brooke's disease), the density of lesions are always greatest in the central face.

Trichoepithelioma most commonly appear as sporadic solitary lesion, while multiple trichoepithelioma are mostly transmitted as an autosomal dominant trait. Mutation in cylidomatosis (CYLD) gene³ on 16q12-13 have been identified as the cause of familial trichoepithelioma, familial cylidomatosis as well as Brook-Spiegler syndrome (familial cylidoma, spiradenoma, milia)⁴. The gene for multiple trichoepithelioma maps to a locus on chromosome 9p21⁵ but for sporadic trichoepithelioma demonstrates deletion at 9q 22.3⁶.

Multiple trichoepithelioma were reported to be associated with Rombo' syndrome, SLE, alopecia and myasthenia gravis. According to our patient, we did not find any associated conditions.

Trichoepithelioma usually persists and can be cosmetic concern to patients. Malignant transformation has only rarely been reported usually in elderly patients⁷. Treatment depends on number of the lesions. Solitary lesions can be excised. In case of multiple tumors, this surgical approach may not be feasible. Split-thickness skin grafting, dermabrasion, laser surgery (CO₂ laser vaporization and Erbium yag laser), electrocautery, cryotherapy, and retinoic acid application have been proposed, but the results of these procedures varies^{8,9}. There had been recent reported treatment of multiple familial trichoepithelioma with a combination of aspirin and neutralizing antibody to TNF α with promising result¹⁰.

References:

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