Multiple papulonodular lesions located over the nasolabial sulcus

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A 54-year-old woman presented with multiple, skin colored, papulonodular lesions located on nose and nasolabial sulcus since 30 years. There were no other cutaneous lesions. A systemic examination was normal. There was a history of similar lesions on the face of one sister and two brothers. There was no history of consanguinity.

Dermatologic examination showed multiple, skin colored, solid, 1-10 mm grouped papulonodular lesions, especially on the nose and nasolabial sulcus [Figure 1a and b]. Histopathologic examination showed a tumor consisting of multiple nodules within the dermis [Figure 2a], basaloid epithelial cells with focal follicular differentiation and small horn cysts [Figure 2b].

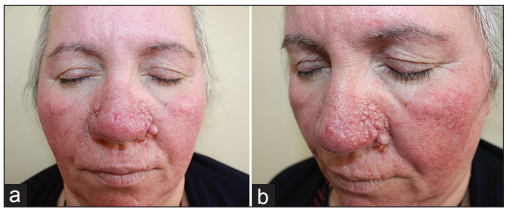

Figure 1: (a) and (b) Papulonodular lesions are around the nose

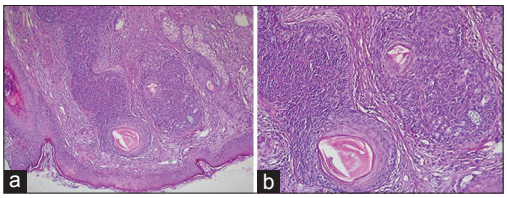

Figure 2: (a) Histopathologic photograph in low power (H and E, ×20); (b) better visualization in ×40

DISCUSSION

Trichoepithelioma is a benign trichogenic tumor that may differentiate into follicular pattern.[1,2] There are two clinical types of trichoepitheliomas: (1) Solitary non-familial type and (2) multiple familial type. The familial type is an autosomal dominant inherited syndrome known as multiple familial trichoepitheliomas or Brook–Fordyce disease. Brook–Fordyce disease is a rare disease and characterized by asymptomatic, multiple, skin colored papulonodular lesions located over the nose, forehead, and nasolabial sulcus.[3]

Multiple trichoepitheliomas may be associated with genetic diseases such as vermiculate atrophoderma, milia, basal cell carcinoma, peripheral cyanosis in Rombo syndrome, basal cell carcinoma, and follicular atrophy in Bazex syndrome.[3] Brook–Spiegler syndrome is an autosomal dominant inherited syndrome including cylindromas, trichoepitheliomas, and occasional eccrine spiradenomas.[3,4] Inherited multiple trichoepitheliomas can appear in this syndrome.

Multiple familial trichoepithelioma is a benign disease and malignant transformation is very rare. Cosmetic concern forms the main reason for treatment of multiple trichoepitheliomas.[3] There are various treatment modalities, including surgical excision, chemical cauterization, laser resurfacing, electrosurgery, dermabrasion, and topical 5% imiquimod cream. However, these treatments may not be effective.[3]

Brook–Fordyce disease is a very rare syndrome. Thus, patients with histologically proven Brook–Fordyce disease should be investigated for family history and systemic involvement. The accompanied syndromes should be investigated, and if detected, malignancy work-up should be performed.

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