Quiz

Leonine facies in an old man

A 68-year-old male farmer presented with multiple asymptomatic lesions over the face since childhood. These lesions gradually increased in size and number over a period of six decades to attend present size and shape. There was no history of bleeding from the lesions. On further probing, he mentioned similar kind of lesions involving face of his younger brother.

Examination revealed multiple, firm, skin colored, and few hyperpigmentd papules and nodules over the forehead, glabella, and nose alongwith obliteration of nasolabial fold [Figure 1].

A punch biopsy from one of the nodule over forehead was taken. Histopathological features of hematoxylin and eosin-stained section are shown in Figures 2-4.

WHAT IS YOUR DIAGNOSIS?

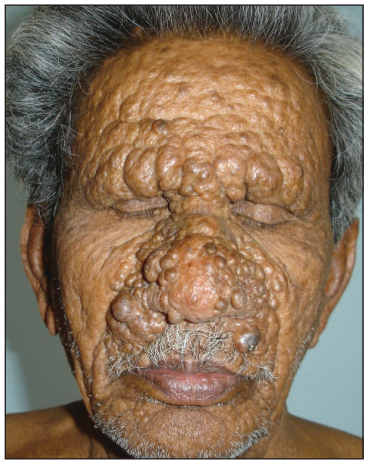

Figure 1: Multiple papules, plaques, and nodules on face

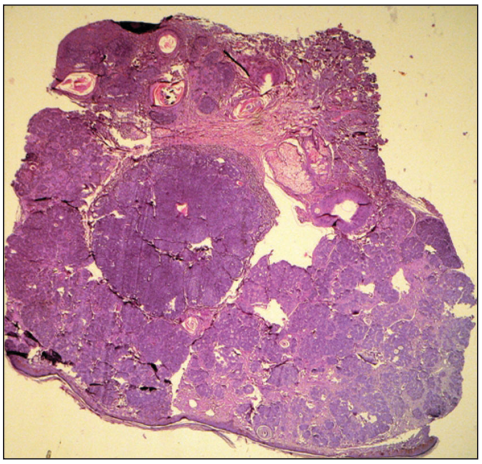

Figure 2: Skin biopsy low power view (H and E, 25×)

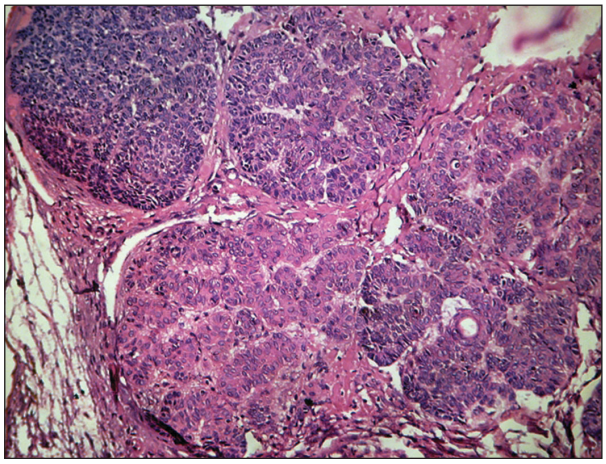

Figure 3: Skin biopsy higher magnification view (H and E, 200×).

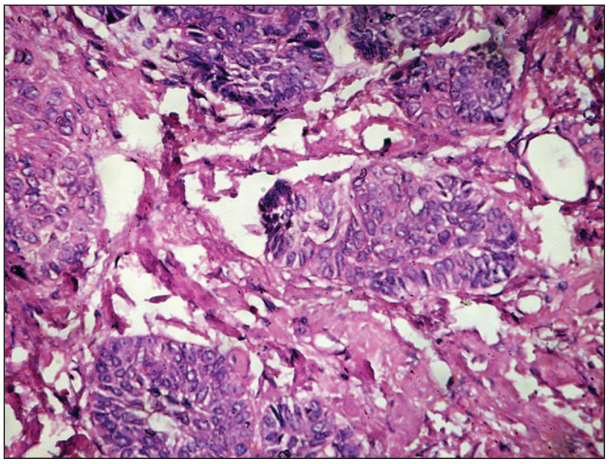

Figure 4: Skin biopsy higher magnification view (H and E, 400×).

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Answer: Multiple familial trichoepithelioma (Epithelioma adenoides cysticum)

DISCUSSION

Leonine facies is a face that resembles that of a lion. It can be seen in multiple conditions that include both infective and non-infective. It has been classically described for lepromatous leprosy. Apart from leprosy, leonine facies can be seen in actinic reticuloid, mycosis fungoides, leishmaniasis, scleromyxedema, and lymphoma. Though these entities may have a common clinical feature, i.e. leonine appearance of face, they have very distinctive histopathological findings which are diagnostic.

Trichoepithelioma also known as epithelioma adenoides cysticum was first described by Brooke and Fordyce in 1892. It is a benign tumor of folliculosebaceous origin.[1] Trichoepithelioma can occur in two forms: solitary and multiple familial trichoepithelioma.

Multiple familial trichoepithelioma is transmitted as an autosomal dominant trait. Onset is usually seen in the childhood as asymptomatic skin-colored papules over the face predominantly affecting nasolabial folds and central part of the face. Lesions may also appear on the neck and trunk. Lesions increase in size with the age, may coalesce to form large lesions, occasionally giving rise to a leonine appearance to the face. Trichoepitheliomas can be one of the multiple skin appendageal tumors that are seen in Brooke-Spiegler syndrome which commonly include cylindroma and spiradenoma. Malignant transformation of trichoepithelioma is very rare.[2]

Multiple familial trichoepithelioma-1 (MFT1) and multiple familial trichoepithelioma-2 (MFT2) have been reported to have a similar clinical picture but have mutations in the CYLD gene at different loci, on chromosome 16q12-q13 and 9p21, respectively.[3,4] Brooke-Spiegler syndrome, familial cylindromatosis and MFT1 have same genotypic mutation but different clinical manifestations and as they can occur in the same patient or in different patients within a single family; some authors consider them as different phenotypic expression of a single disease entity.[5,6]

On histopathology trichoepithelioma shows proliferation of basaloid cells in the dermis in various patterns of which cribriform or sieve like is the most common [Figures 2 and 3]. Infundibulocystic structures and germ and papillae structures are commonly seen [Figure 4]. Occasionally calcification can be seen. At times it may be difficult to distinguish trichoepithelioma from basal cell carcinoma.

Solitary trichoepithelioma can be excised surgically while multiple trichoepitheliomas are difficult to manage. Laser ablation, cryosurgery, and electrocautery have been found useful with variable success.[7-10]

Atul Dongre, Swati Gondse, Sumit Kar

Department of Skin and VD, MGIMS, Sevagram, Maharashtra, India

Address for correspondence:
Dr. Atul Dongre, Department of Skin and VD, MGIMS, Sevagram, Maharashtra. E-mail: atul507@yahoo.co.in

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REFERENCES

1. Clarke J, Ioffreda M, Helm KF. Multiple familial trichoepitheliomas: A folliculosebaceous- apocrine genodermatosis. Am J Dermatopathol 2002;24:402-5.
2. Lee KH, Kim JE, Cho BK, Kim YC, Park CJ. Malignant transformation of multiple familial trichoepithelioma: Case report and literature review. Acta Dermatol Venereol 2008;88:43-6.
3. Salhi A, Bornholdt D, Oeffner F, Malik S, Heid E, Happle R, et al. Multiple familial trichoepithelioma caused by mutations in the cylindromatosis tumor suppressor gene. Cancer Res 2004;64:5113-7.
4. Harada H, Hashimoto K, Ko MS. The gene for multiple familial trichoepithelioma maps to chromosome 9p21. J Invest Dermatol 1996;107:41-3.
5. Young AL, Kellermayer R, Szigeti R, Teszas A, Azmi S, Celebi JT. CYLD mutations underlie Brooke-Spiegler, familial cylindromatosis and multiple familial trichoepithelioma syndromes. Clin Genet 2006;70:246-9.
6. Biggs PJ, Wooster R, Ford D, Chapman P, Mangion J, Quirk Y, et al. Familial cylindromatosis gene localised to chromosome 16q12-q13: Evidence for its role as a tumour suppressor gene. Nat Gene 1995;11:441-3.
7. Shaffelburg M, Miller R. Treatment of multiple trichoepithelioma with electrosurgery. Dermatol Surg 1998;24:1154-6.
8. Duhra P, Paul JC. Cryotherapy for multiple trichoepithelioma. J Dermatol Surg Oncol 1988;14:1413-5.
9. Flores JT, Apfelberg DB, Maser MR, Lash H. Trichoepithelioma: Successful treatment with the argon laser. Plast Reconstr Surg 1984;74:694-8.
10. Sawchuk WS, Heald PW. CO2 laser treatment of trichoepithelioma with focused and defocused beam. J Dermatol Surg Oncol 1984;10:905-7.