

Multiple Familial Trichoepitheliomas Presenting as Leonine Facies

Abstract

Trichoepithelioma is a benign tumor of follicular unit. It has been rarely described as the cause of leonine facies. We are presenting a classical case of multiple familial trichoepitheliomas (MFTs) with characteristic histopathological features leading to leonine facies.

Keywords: Basaloid cells, familial, leonine facies, trichoepithelioma

Introduction

Trichoepithelioma is a benign follicular tumor that presents as small, skin-colored papules and nodules predominantly on the face. When more than one family member is affected, the disease is known as multiple familial trichoepitheliomas (MFT). It is usually inherited as an autosomal dominant disease. Leonine facies due to trichoepithelioma is uncommonly described. We hereby describe an adult male presenting with leonine face due to trichoepithelioma with a family history of similar illness. We have also summarized the causes of leonine facies described in the literature so far.

Case Report

A 20-year-old male, presented with a 10-year history of multiple skin-coloured, firm, asymptomatic papules, and nodules of size 2–10 mm over face and few similar discrete lesions over scalp, ears, and neck [Figure 1]. Lesions were very closely aggregated over central face leading to the appearance of leonine facies. There was no associated systemic abnormality and routine laboratory investigations were within normal limits. Similar lesions were also present in the patient's mother. A clinical differential diagnosis of nodular lepromatous leprosy, post-kala azar dermal leishmaniasis, non-Langerhans cell histiocytosis, familial cylindromatosis, and Brooke-Spiegler syndrome were kept. There were no hypopigmented or hypoesthetic lesions which virtually ruled out the differentials of leprosy and leishmaniasis.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Besides this, nerve trunks were not thickened. Lesions were not painful, and that helped us to some extent to rule out diagnosis of spiradenomas which are part of Brooke-Spiegler syndrome. A punch biopsy from face and scalp showed multiple nests of basaloid cells in the dermis with peripheral palisading in few nests. Some of the aggregates were containing horn cysts and showing follicular differentiation. Mitotic figures and atypical cells were not noticed in nests [Figure 2a and b]. Histopathological features further helped us to exclude the differential diagnosis of non-Langerhans cell histiocytosis, familial cylindromatosis, and Brooke-Spiegler syndrome. On clinicopathological correlation, a final diagnosis of MFTs was considered. Radiofrequency ablation (RFA) by loop cautery (Vesalius, cutting-coagulation mode) was done with moderately good cosmetic outcome after three sessions and he has been planned for further sessions [Figure 3]. In postoperative period, the patient was advised for strict photoprotection, but he did not follow the instructions and developed mild postinflammatory hyperpigmentation at the operated site. Interestingly, there was minimal surface scarring even at end of three sessions of RFA.

Discussion

Leonine facies is a face that resembles that of a lion. It is a rare clinical presentation and corresponds to the morphologic manifestation of diffuse dermal infiltration of the face. It has been classically described for lepromatous leprosy. Apart from

How to cite this article: Singh S, Sondhi P, Yadav D, Yadav S. Multiple familial trichoepitheliomas presenting as leonine facies. Indian Dermatol Online J 2017;8:358-60.

Received: March, 2017. **Accepted:** May, 2017.

Sanjay Singh,
Prateek Sondhi,
Deepika Yadav,
Savita Yadav

Department of Dermatology and Venereology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence:
Dr. Savita Yadav,
Department of Dermatology and Venereology, All India Institute of Medical Sciences,
New Delhi, India.
E-mail: savita2081@gmail.com

Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_67_17

Quick Response Code:





Figure 1: Multiple closely aggregated skin colored, firm, papules, and nodules over face and scalp

leprosy, leonine facies have been reported in cutaneous sarcoidosis, leishmaniasis, mastocytosis, actinic reticuloid, scleromyxedema, hematological malignancies, mycosis fungoides, leukemia cutis, and various other diseases [Table 1].^[1-12]

Trichoepithelioma is a benign tumor of folliculosebaceous origin.^[3] It was first described by Brooke and Fordyce in 1892.^[3] The name trichoepithelioma is preferable to other designations, such as epithelioma adenoides cysticum and multiple benign cystic epithelioma, because it is more suggestive of the differentiation of this tumor towards hair structures. Trichoepithelioma can occur in two forms: Solitary and MFTs. MFTs is transmitted as an autosomal dominant trait. Its exact prevalence is not known but appears to be an uncommon disease entity. 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 age, may coalesce to form large lesions, occasionally giving rise to a leonine appearance to the face.^[3] Telangiectasia can be seen on the surface of the larger lesions resembling basal cell carcinoma (BCC). In contrast to BCC, ulceration is very rarely present in trichoepithelioma. Rarely, trichoepithelioma can be seen in association with BCC.^[13] Recently, an interesting case

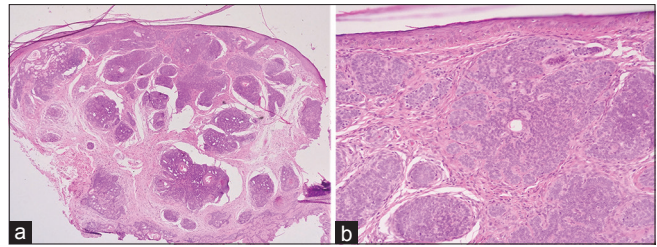


Figure 2: (a and b) Nests of basaloid cells containing few horn cysts in upper and mid dermis with follicular differentiation (Hematoxylin and eosin staining, ×40 and ×100)

Table 1: Causes of leonine facies

Leprosy
Chronic actinic dermatitis
Cutis verticis gyrata
Leishmaniasis
Lymphoma
Leukemia
Mycosis fungoides
Multicentric reticulohistiocytosis
Multiple keratoacanthoma syndrome (Grzybowski type)
Progressive nodular histiocytoma
Sarcoidosis
Scleromyxedema
Mastocytosis
Carcinoid syndrome
Pachydermoperiostosis (Touraine–Solente–Gole' syndrome)
Sebaceous hyperplasia and sebaceous adenomas
Trichoepithelioma
Indeterminate dendritic cell tumor
Amyloidosis
Trichodysplasia spinulosa
Scleredema diabeticorum
Malignant syphilis
Leontiasis ossea
Kaposi's sarcoma
Lipoid proteinosis
Setleis syndrome
Paraneoplastic Acanthosis Nigricans
Hyper eosinophilic syndrome

of sporadic multiple facial trichoepitheliomas presenting as diffuse waxy infiltration of the face and alopecia of eyebrows, and moustache leading to early leonine facies has been reported.^[14]

Diagnosis is mainly based on characteristic clinical and histopathological features. Typical trichoepithelioma is characterized by nests of basaloid cells containing numerous horn cysts with follicular differentiation. Basaloid cells in nests can present with peripheral palisading. A distinctive feature is the papillary-mesenchymal body (fibroblastic aggregate resembling abortive follicular papillae). Mitotic figures and peripheral palisading are uncommonly observed.



Figure 3: Moderately good improvement in glabellar region, nose, and adjacent cheek in form of significant flattening of lesions with minimal scarring and mild postinflammatory pigmentation

Occasionally, melanocytes can be seen within the nests as trichoepitheliomas recapitulate hair differentiation. Genetic analysis can be done in familial type which shows abnormalities in band 9p21.^[13]

Multiple trichoepitheliomas are seen in Brooke-Spiegler syndrome. It is a rare autosomal dominant disorder with *CYLD1* gene mutation and clinically characterized by a triad of trichoepithelioma, cylindroma, and spiradenoma. Other reported associations of multiple trichoepitheliomas include Rombo and Bazex syndrome.^[13]

Surgical excision, dermabrasion, RFA, and ablative lasers are the mainstay of therapy. Ablative laser and dermabrasion may improve the cosmetic appearance, but partial ablation of the tumour is usually followed by recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not

be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Kanathur S, Sarvajnyamurthy S, Somaiah SA. Characteristic facies: An index of the disease. *Indian J Dermatol Venereol Leprol* 2013;79:439-43.
2. Mutreja D, Purohit A, Singh PK, Pati HP. A 60-year-old lady with leonine facies: A rare diagnosis. *Indian J Pathol Microbiol* 2012;55:566-8.
3. Dongre A, Gondse S, Kar S. Leonine facies in an old man. *Indian J Dermatol Venereol Leprol* 2010;76:213-4.
4. Yuste M, Romo A, Nieto G, Unamuno P. Leonine facies in carcinoid syndrome. *Actas Dermosifiliogr* 2010;101:542-6.
5. Gupta V, Mridha AR, Sharma VK. Sebaceous hyperplasia and sebaceous adenomas presenting as leonine facies and improving with oral isotretinoin. *Clin Exp Dermatol* 2016;41:923-4.
6. Oh CW, Ivan D, Curry JL, Ellis R, Gerber H, Duvic M, *et al.* A case of indeterminate dendritic cell tumor presenting with leonine facies. *J Cutan Pathol* 2016;43:158-63.
7. Kirchhof MG, Shojania K, Hull MW, Crawford RI, Au S. Trichodysplasia spinulosa: Rare presentation of polyomavirus infection in immunocompromised patients. *J Cutan Med Surg* 2014;18:430-5.
8. Tran K, Boyd KP, Robinson MR, Whitlow M. Scleredema diabeticorum. *Dermatol Online J* 2013;19:207-18.
9. De Socio GVL, Simonetti S, Tomasini C, Ansidei V, Pasticci MB, Baldelli F. Malignant syphilis with ocular involvement in an HIV-infected patient. *Int J STD AIDS* 2011;22:298-300.
10. Mansilla-Lory J, Amezcua-Guerra LM, Vargas-Rodríguez A, Fernández-Tapia S, Leboreiro I, Resnick D, *et al.* Leontiasis ossea: A paleopathologic case report. *J Clin Rheumatol* 2007;13:269-72.
11. Tanabe A, Kusumoto K, Suzuki K, Ogawa Y. Treatment of Setleis syndrome. Case report. *Scand J Plast Reconstr Surg Hand Surg* 2001;35:107-11.
12. Kannenberg SMH, Koegelenberg CFN, Jordaan HF, Bolliger CT. A patient with leonine facies and occult lung disease. *Respiration* 2010;79:250-4.
13. Sehrawat M, Jairath V, Jain VK. Nonfamilial Multiple Trichoepithelioma: Few and Far Between. *Indian J Dermatol* 2016;61:78-80.
14. Bhari N, Mahajan R, Gupta S. Early leonine facies with alopecia in a young man. *Int J Dermatol* 2016;55:1299-300.