CASE REPORT

Leonine Facies in a case of Diffuse Sebaceous Hyperplasia and the response to Isotretinoin treatment

Verónica Úraga W.,* Annette Morán A.,* Enrique Úraga P.**

* *Dermatologist at the Dermatology Center Dr. Úraga*
** *Dermatologist and Director at the Dermatology Center Dr. Úraga*

Correspondence: veronica_uraga@hotmail.com

Keywords: Presenile diffuse familial sebaceous hyperplasia, isotretinoin, treatment, leonine facies.

ABSTRACT

Presenile diffuse familial sebaceous hyperplasia is an extremely rare condition, with less than 20 cases reported in the medical literature. The condition is more unusual when presented as leonine facies. This case report presents a patient with leonine facies and extrafacial involvement of which dermoscopy and histopathology were compatible with sebaceous hyperplasia. The patient received oral isotretinoin therapy with satisfactory results.

INTRODUCTION

Diffuse sebaceous hyperplasia is a rare variant which, contrary to classic sebaceous hyperplasia, manifests early in life with positive family history. Therefore, it is also known as presenile diffuse familial sebaceous hyperplasia. It shows diffuse expansion of the sebaceous glands and overproduction of sebum, which leads to the formation of plaques at the face, neck and upper thorax. This may progress into widespread skin thickening and the development of deep cutaneous grooves, giving rise to the leonine facies aspect. Leonine facies presentation cases are highly rare. Only six cases have been reported in the medical literature at reach.[1–5] A patient manifesting leonine facies with neck and upper thorax involvement due to diffuse sebaceous hyperplasia is described. Low doses of isotretinoin resulted in notable improvement.

CLINICAL CASE

A 49-year-old male patient presented with a clinical picture characterized by multiple small skin-colored papules located at the face which formed plaques and involved skin thickening and marked accentuation of facial folds, revealing the typical leonine facies appearance (Fig. 1a). No falling out of eyebrow tails was observed. The patient's complexion was oily. Additionally, he presented with numerous yellow papules in the neck and V area of the chest (Fig. 1b). The patient indicated his condition started more than twenty years ago and has continued to progress slowly. No other skin lesions were observed. Moreover, there was not any relevant pathological history nor regular medication intake. In addition, he referred his father and grandfather had suffered from similar but milder lesions.

Dermoscopy revealed multiple yellowish-white globules and non-arborizing telangiectasias (Fig. 1e).

A skin biopsy of the chin, which evidenced numerous mature sebaceous glands displayed in lobules throughout the dermis, was performed. (Fig. 2).

These findings lead to the diagnosis of presenile diffuse familial sebaceous hyperplasia. The patient initiated

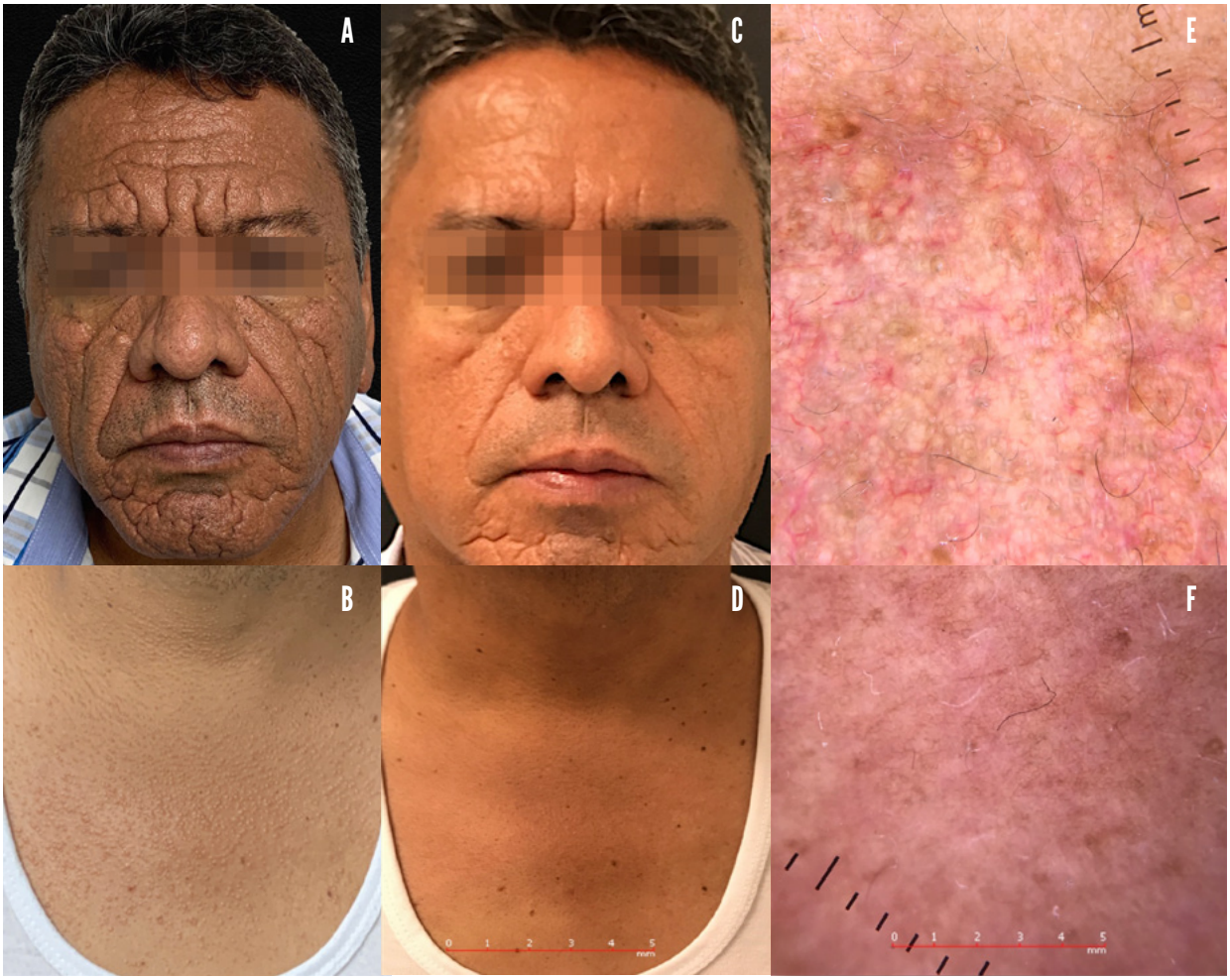

Fig. 1. (a) Skin-colored micropapules in the face forming plaques, with skin thickening and accentuation of facial folds. (b) Yellowish papules in the neck and neckline. (c) (d) Results of the face and chest after a 3-month treatment. (e) Dermoscopic evaluation previous to treatment, revealing multiple yellowish-white globules and non-arborizing telangiectasias. (f) Dermoscopic evaluation after a 3-month treatment.

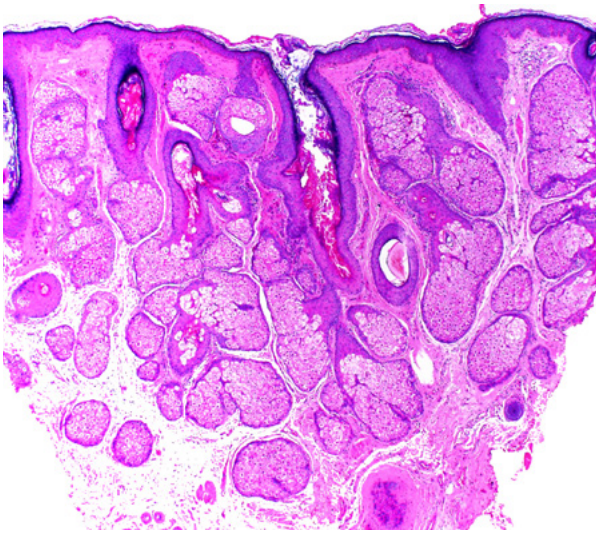

Fig. 2. Histopathology showing multiple mature sebaceous glands displayed in lobules throughout the dermis.

treatment on 20 mg daily doses of isotretinoin and presented with a dramatic response after three months. The face presented with a marked decrease of seborrhea and smaller lesional size, consequently reducing the skin thickening and grooves (Fig. 1c). Clinically, lesions in the neck and neckline virtually lightened (Fig. 1d) which was corroborated by dermoscopic findings (Fig. 1f). Presently, the patient continues with isotretinoin treatment. Additionally, apart from a very mild case of cheilitis, no notable secondary effects have been noted.

DISCUSSION

Conjointly with classical senile sebaceous hyperplasia, there are some rare variants: giant, linear, juxta-clavicular beaded lines,[6,7] and diffuse presenile (sporadic or familial).[8]

Diffuse sebaceous hyperplasia typically occurs during puberty or shortly after. It is thought to present autosomal dominant inheritance with incomplete penetrance.[9] It shows progressive expansion of overactive sebaceous glands, with facial skin thickening and irregularities, forming plaques while commonly respecting periorificial areas. It can spread over time, therefore compromising the neck and upper thorax.[10]

On rare occasions, diffuse hyperplasia progression of the face may lead to leonine facies,[2,5] as occurred in this case. Diagnosis requires a thorough clinical examination; dermoscopic evaluation may be a useful complementary tool and pathological study can confirm diagnosis and distinguish leonine facies from other pathologies.[3]

Despite the existence of several treatment options, full recovery is not likely to occur; there is risk of recurrence. Many destructive techniques with physical and chemical agents, as well as different types of laser, have been recounted. Nonetheless, depigmentation and atrophic scars are potential complications.[8,11]

Among patients, isotretinoin treatment has been described as a successful alternative therapeutic option.[4,5,12] Isotretinoin inhibits differentiation and proliferation of sebaceous glands. As a result, glandular size decreases and sebum production is suppressed. Effects are noted after the first week. They optimize between the third and sixth week, leading to perifollicular fibrosis after the twelfth week of treatment.[4,11]

Currently, there is debate about optimal loading and maintenance doses of isotretinoin to prevent recurrence.[11] In contrast to the use of isotretinoin in acne, there are no standardized schemes. Doses of 0,2 to 0,5 mg/kg and even 1 mg/kg per day have been administered for two to six-month periods, resulting in variable recurrence.[5,11,13] The patient in question has responded satisfactorily to a 3-month 20 mg dose treatment. The treatment plan consists in maintaining the dose for a period of four months before reducing it to 10 mg in combination with a topical retinoid. Treatment effectiveness ought to be assessed.

CONCLUSIONS

Leonine facies is an extremely rare presentation of sebaceous hyperplasia. The study of this variant, along with the use of dermoscopy and skin biopsy may contribute to distinguish this condition from other pathologies. In view of the patient's great response to low doses of isotretinoin, this was considered as a safe, reasonable and effective therapeutic option for the rest of subjects.

Lengthier studies are required to define proper dosage, as well as an adequate treatment plan and duration.

REFERENCES

1. Mohamed K. Facial Lesions Resembling Leprosy. Int J Lepr Other Mycobact Dis. 2001;69(1):35–7.
2. Abdel-halim MRE, El-nabarawy E, El-Tawdy A, Fawzy MM, Shalaby S, Ismail S, et al. Leonine facies and neck papules. Int J Dermatol. 2019;58(7):797–9.
3. Trakoolwannachai P, Kootiratrakarn TK. Familial sebaceous hyperplasia : A case report . Thai J Dermatol. 2016;32(4):255–61.
4. Wang W, Qiu Y, Zhou G, Pei Z, Zhang F. Premature sebaceous hyperplasia with satisfactory response to oral isotretinoin. Indian J Dermatology, Venereol Leprol. 2015;82(1):113.
5. Gupta V, Mridha AR, Sharma VK. Sebaceous hyperplasia and sebaceous adenomas presenting as leonine facies and improving with oral isotretinoin doi: Clin Exp Dermatol. 2016;41(8):923–4.
6. Donati P, Muscardin LM, Maini A. Juxtaclavicular Beaded Lines : A Malformative Condition Affecting Sebaceous Glands. Dermatology 2000;200:283. 2000;200:283.
7. Woldow AB, Houk LD, Samie FH. Juxtaclavicular beaded lines: A presentation of sebaceous gland hyperplasia. Dermatol Online J. 2009;15(4):1–6.
8. Eisen DB, Michael DJ. Sebaceous lesions and their associated syndromes: Part I. J Am Dermatology [Internet]. 2009;61(4):549–60. Available from: http://dx.doi.org/10.1016/j.jaad.2009.04.058
9. Boonchai W, Leenutaphong V. Familial presenile sebaceous gland hyperplasia. J Am Acad Dermatology. 1997;36(1):120–2.

10. Dupre A, Bonafe J, Lamon P. Functional familial sebaceous hyperplasia of the face and premature sebaceous gland hyperplasia: A new and unique entity? J Am Acad Dermatol. 1983;9(5):768–769.
11. Costa L, Consigli C, Consigli J. Hiperplasia sebácea. Tratamiento con isotretinoína. Dermatol Argent. 2016;22(3):146–50.
12. Tagliolatto S, Mota M, Alchorne DA. Sebaceous hyperplasia : systemic treatment with isotretinoin*. An Bras Dermatol. 2015;90(2):211–5.
13. Liu YS, Cheng Y, Liu C, Yang C, Yang C. Presenile diffuse familial sebaceous hyperplasia successfully treated with low-dose isotretinoin : A report of two cases and review of the published work. J Dermatol. 2016;43(10):1205–8.