Our Dermatology Online

Case Report

Unusual keloidal granuloma faciale recalcitrant to several treatments

Lina Al Soufi[1], Heba Fawal[1], Zuheir Al-Shehabi[2]

[1]Department of Dermatology, National Hospital, Lattakia, Syria, [2]Department of Pathology, Faculty of Medicine, Tishreen University, Lattakia, Syria

Corresponding author: Lina Al Soufi, MD, E-mail: fawalheba6@gmail.com

ABSTRACT

Granuloma faciale is a rare chronic skin disorder characterized by asymptomatic papules, nodules, and plaques on the face. Although its pathogenesis is unknown, researchers have suggested that one of the main causative factors may be sun exposure, as lesions are aggravated by sunlight and favor light-exposed areas. Herein, we are reporting the case of an adult female patient with keloidal granuloma faciale treated with a combination of several treatment methods with no improvement and a relapse. In this case, we attempted to highlight the difficulties that the doctor faces in treating the disease and to share the medical literature of our treatment experience.

Key words: Granuloma faciale; Keloidal; Chronic skin disorder; Grenz zone

INTRODUCTION

Granuloma faciale (GF) is a rare, idiopathic dermatosis of benign course and chronic progression characterized by single or multiple, reddish-brown or violaceous, cutaneous nodules or plaques, and their surface may show the exaggeration of follicular openings, telangiectasis, or scaling, most frequently on the face [1]. The term *granuloma faciale* was coined by Wigley in 1945, referring to the condition as eosinophilic granuloma of the skin [2,3]. The plaque is usually located on the face, yet may sometimes appear on the trunk, extremities, or in the nasal cavity (extrafacial GF) [4]. The disease may be seen at any age, yet is primarily a disease of middle age and is more common in males [5]. Granuloma faciale is confined to the skin and there is no systemic involvement, even in patients with disseminated lesions. The disease is usually difficult to treat and has a recurring character, with periods of exacerbation. Spontaneous resolution is rare.

CASE REPORT

A fifty-year-old Syrian female patient presented with asymptomatic, bilateral lesions on the face persistent for twenty years, which had become more pronounced with solar exposure. It began on the left side of the face as nodular eruptions, which had increased in size to form a large tumor. A general examination was within normal limits. A dermatological examination revealed the presence of multiple, brownish, erythematous papules of well-defined limits, located in the malar region. The surface of the lesions was smooth with follicular openings, telangiectasia, and palpable hardness (Fig. 1).

Several previous biopsies were performed and were consistent with the diagnosis of granuloma faciale. However, a histological examination revealed a rectified epidermis, with a subepidermal grenz zone separating the nodular dermal inflammatory infiltrate, composed predominantly of lymphocytes, histiocytes, neutrophils, and numerous eosinophils. There was no granuloma seen. No fungal elements were seen on Giemsa stain. Based on the clinical picture and histological examination, the diagnosis of granuloma faciale was confirmed (Fig. 2).

The condition had undergone numerous treatments, including topical steroids and cryotherapy, then

How to cite this article: Al Soufi L, Fawal H, Al-Shehabi Z. Unusual keloidal granuloma faciale recalcitrant to several treatments. Our Dermatol Online. 2022;13(3):302-304.

Submission: 18.02.2022; **Acceptance:** 05.04.2022

DOI: 10.7241/ourd.20223.15

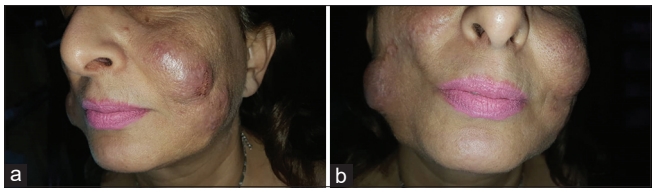

Figure 1: (a and b) Reddish-brown nodules on the left and right cheeks (notice the scar on the right side after the surgical removal of the lesion).

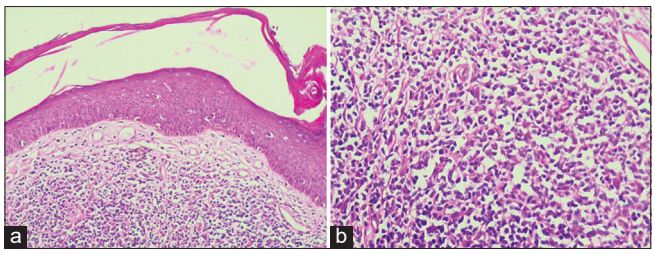

Figure 2: (a) Skin biopsy showing a subepidermal grenz zone separating the nodular dermal inflammatory infiltrate, composed predominantly of lymphocytes, histiocytes, neutrophils, and numerous eosinophils (H&E, 100×). (b) Perivascular infiltrates could penetrate the vascular wall and cause leukocytoclasis (H&E, 200×).

treatment with dapsone in combination with systemic corticosteroids yielded good results, yet the lesions relapsed after stopping the treatment.

Afterward, the patient was treated with antimalarials drugs in combination with systemic steroids, additionally to intralesional injection of triamcinolone and CO_2 fractional laser without improvement.

After the failure of the previous treatments, a surgical excision of the lesions was performed, which led to the healing of the nodular lesions, yet a recurrence occurred after one year on the wound edges of the lesion on the left cheek. A clinical response was evaluated by measuring the size and thickness of the GF.

DISCUSSION

Granuloma faciale (GF) is considered a localized form of small vessel vasculitis with frequent leukocytoclasis [1,6]. Granuloma faciale is a misnomer as granuloma formation is not a histopathological feature of this disease [2]. Clinically, it is characterized by single or multiple, asymptomatic, reddish-brown to violaceous papules, nodules, and plaques. The lesions are typically localized on the face, yet may uncommonly be extrafacial [6,7]. Researchers have suggested that one of the main causative factors for the disease may be sun exposure as lesions are aggravated by sunlight and favor light-exposed areas [8]. Although the clinical presentation of GF is usually characteristic, the diagnosis may be delayed by the relative rarity of the disease and the presence of numerous other pathologies with clinical similarities.

There are several main differential diagnoses, including lymphoma and pseudo-lymphoma, sarcoidosis, lupus erythematosus tumidus, polymorphous light eruption, fixed pigmented erythema, erythema elevatum diutinum, foreign body granuloma, and granulomatous rosacea [9]. The disease tends to be refractory to several therapeutic modalities, including topical and intralesional corticosteroids, antimalarial drugs, dapsone, surgical excision, phototherapy, cryosurgery, dermabrasion, electrosurgery, as in the case of our patient. The specificity of the disease in terms of its refraction to treatment constitutes a challenge for doctors and causes a psychological problem for the patient. However, more recent studies have shown good results after four months with topical tacrolimus 0.1% [10]. Meanwhile, other studies have shown good and promising results with intralesional rituximab [11]. In this case, we attempted to highlight the difficulties that the doctor faces in treating the disease and to share the medical literature of our treatment experiences.

CONCLUSION

Herein, we have reported a case of a refractory keloidal granuloma faciale in an adult female patient persistent for twenty years. GF is a highly persistent disease that resists treatment. However, dermatologists should be encouraged to share experiences regarding the management of granuloma faciale in future publications, due to the low incidence of GF, which would provide a greater opportunity to exchange these therapeutic experiences.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Ortonne N,Wechsler J, Bagot M, Grosshans E, Cribier B. Granuloma faciale: A clinicopathologic study of 66 patients. J Am

Acad Dermatol. 2005;53:1002-9.
2. Ziemer M, Koehler MJ, Weyers W. Erythema elevatum diutinum: A chronic leukocytoclastic vasculitis microscopically indistinguishable from granuloma faciale? J Cutan Pathol. 2011;38:876-83.
3. Wigley JE. Eosinophilic granuloma? Sarcoid of Boeck. Proc R Soc Med. 1945;38:125-6.
4. Konohana A. Extrafacial granuloma faciale. Dermatol. 1994;21:680-2.
5. Lindhaus C, Elsner P. Granuloma faciale treatment: A systematic review. Acta Derm Venereol. 2018;98:14-8.
6. Deen J, Moloney TP, Muir J. Extrafacial granuloma faciale: A case report and brief review. Case Rep Dermatol. 2017;9:79-85.
7. Nasiri S, Rahimi H, Farnaghi A, Asadi-Kani Z. Granuloma faciale with disseminated extra facial lesions. Dermatol Online J. 2010;16:5.
8. Burgdorf WHC. Granuloma faciale. In: Freedberg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Katz SI, Fitzpatrick TB, editors. Fitzpatrick's dermatology in general medicine. 5th ed. New York: McGraw-Hill; 1999. p. 1138-40.
9. Oliveira CC, Ianhez PE, Marques SA, Marques ME. Granuloma faciale: Clinical, morphological and immunohistochemical aspects in a series of 10 patients. An Bras Dermatol. 2016;91:803-7.
10. Lima RS, Maquiné GÁ, Silva Junior RC, Schettini AP, Santos M. Granuloma faciale: A good therapeutic response with the use of topical tacrolimus. An Bras Dermatol. 2015;90:735-7.
11. Morgado-Carrasco D, Giavedoni P, Mascaró JM Jr, Iranzo P. Assessment of treatment of refractory granuloma faciale with intralesional rituximab. JAMA Dermatol. 2018;154:1312-5.

Source of Support: Nil, **Conflict of Interest:** None declared.