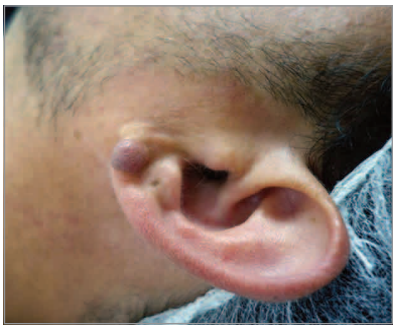
JAMA Dermatology Clinicopathological Challenge

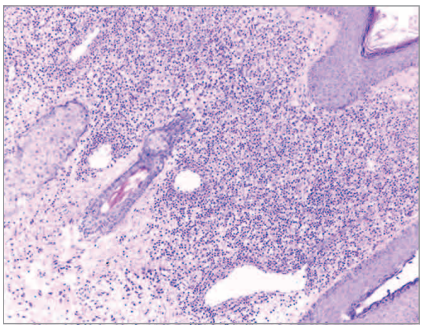
Unusual Tumor on the Earlobe

María Jesús Hernández San Martín, MD; Javier Arellano, MD; Yamile Corredoira Salum, MD

A Nodule on the left ear



B Original magnification ×10



C Original magnification ×40

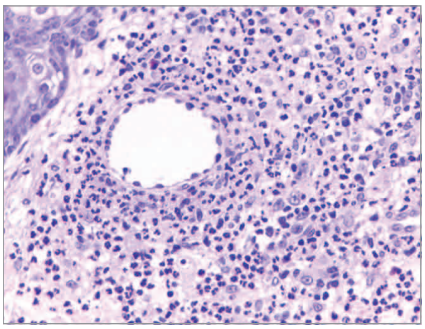


Figure. A, Clinical photograph of a pruriginous, reddish tumor on the earlobe. B and C, Histopathologic findings reveal mixed diffuse dermatitis involving the reticular dermis, with small-vessel vasculitis, characteristic sparing of a superficial dermal grenz zone as well as the dilated vessel luminal, and mixed leukocytic infiltrate with eosinophils, lymphocytes, and neutrophils involving the full thickness of the vessel wall (hematoxylin-eosin).

A man in his 30s presented with a 3-month history of gradual growth of a pruriginous, reddish nodule on the left ear. He had no relevant medical history. On medical examination, a reddish, smooth, superficial nodule with small dimples and a soft consistency that measured approximately 1.5 cm in diameter was seen on the left earlobe (**Figure**, A). It was not painful to the touch. The patient reported no history of lesions or foreign bodies at this location. A biopsy specimen was obtained for histopathologic analysis (Figure, B and C).

WHAT IS YOUR DIAGNOSIS?

A. Sweet syndrome

B. Extrafacial granuloma faciale

C. Angiolymphoid hyperplasia with eosinophilia

D. Leishmaniasis

Diagnosis

B. Extrafacial granuloma faciale

Microscopic Findings and Clinical Course

The biopsy results revealed a mixed dermal infiltrate with eosinophils, neutrophils, and plasma cells, and a nonleukocytoclastic vasculitic component without epidermal changes. The histologic appearance was compatible with extrafacial granuloma faciale (GF).

Discussion

Granuloma faciale is a chronic, benign, and infrequent inflammatory skin disease of unknown origin. It usually manifests as a solitary reddish-violet plaque on the face, and extrafacial involvement may occur; other potential locations include the ears, scalp, trunk, and limbs.[1,2] Lesion size varies from several millimeters to several centimeters in diameter. The lesions are usually smooth on the surface and can often present with superficial telangiectasias and prominent follicular orifices, which sometimes show an orange peel appearance.[3] Granuloma faciale can be divided into plaque or nodular types, with the latter being generally resistant to therapy.[4]

In a retrospective analysis of 66 patients with GF, i27 (41.5%) patients presented with a nodule, 25 (38%) had multiple lesions, 6 (9%) had extrafacial involvement alone, and 1 (1.5%) had facial and extrafacial involvement.[5] Some studies have included GF in the spectrum of IgG4-related diseases.[1-4] Although the precise pathogenesis remains unknown, interferon-γ and increased local production of interleukin 5 may be important mediators.[1]

Granuloma faciale is characterized by a diffuse polymorphous inflammatory infiltrate that mainly affects the upper half of the dermis.[5,6] The histologic characteristics of leukocytoclastic vasculitis are more prominent in the beginning, with older lesions tending to have fewer neutrophils and more eosinophils and plasma cells, as well as fibrosis.[1] The clinical appearance of GF is distinctive, but the differential diagnosis may include lymphoma, persistent arthropod bite reactions, angiolymphoid hyperplasia with eosinophilia, tumid lupus erythematosus, erythema elevatum diutinum, and various granulomatous disorders.

Treatment for this condition is limited. First-line therapies include corticosteroids (intralesional injection or topical), topical use of tacrolimus, or complete surgical removal.

Sweet syndrome is characterized by a constellation of clinical symptoms that include high fever and sensitive erythematous cutaneous lesions that are usually painful. Histopathologic results reveal a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis. In this case, the patient did not comply with the necessary criteria.[7] In contrast, angiolymphoid hyperplasia with eosinophilia is characterized by papules or nodules with an angiomatoid aspect and that are predominantly located on the head, especially around the ears. The histopathologic characteristics of angiolymphoid hyperplasia with eosinophilia include abnormal vascular proliferation formed by capillaries grouped around arterial or venous vessels, with lymphocytic infiltrates with eosinophils instead of a mixed dermal infiltrate.[8]

Localized cutaneous leishmaniasis tends to occur in exposed areas of the skin and begins as a pink papule which gets bigger and becomes a nodule or a lesion similar to a plaque. The distinctive differences between GF and leishmaniasis are its clinical presentation and the evolution of the lesion; treatment for leishmaniasis can take months or years. In addition, the histopathologic results include a granulomatous infiltrate in the dermis and the presence of amastigotes in the acute phase of infection.[9,10]

Herein, we present a rare case of nodular extrafacial GF. The patient underwent surgical excision of the lesion, with no recurrence reported. This condition should be included in the differential diagnosis of chronic plaques or nodules in the ears, scalp, trunk, and/or extremities, particularly in middle-aged men.

ARTICLE INFORMATION

Author Affiliations: Department of Dermatology, University of Chile, Santiago, Chile (Hernández San Martín, Arellano); Department of Dermatology, Clinical Hospital San Borja Arriarán, Santiago, Chile (Arellano); Department of Pathology, University of Chile, Santiago, Chile (Corredoira Salum).

Corresponding Author: María Jesús Hernández San Martín, MD, Department of Dermatology, University of Chile, Santos Dumont 999, Independencia, Santiago, Chile 8280456 (majesus.hsm@gmail.com).

Published Online: November 27, 2019. doi:10.1001/jamadermatol.2019.3440

Conflict of Interest Disclosures: None reported.

Additional Contributions: We thank the patient for granting permission to publish this information.

Self-assessment Credit: This article is eligible for journal-based self-assessment (1 credit) for Maintenance of Certification (MOC) from the American Board of Dermatology (ABD). After completion of an activity, please log on to the ABD website at www.abderm.org to register your credits. This may be done after each exercise or after accumulating many credits.

REFERENCES

1. Nasiri S, Rahimi H, Farnaghi A, Asadi-Kani Z. Granuloma faciale with disseminated extra facial lesions. *Dermatol Online J*. 2010;16(6):5.

2. Deen J, Moloney TP, Muir J. Extrafacial granuloma faciale: a case report and brief review. *Case Rep Dermatol*. 2017;9(2):79-85. doi:10.1159/000477960

3. Long H, Zhang G, Wang L, Lu Q. Eosinophilic skin diseases: a comprehensive review. *Clin Rev Allergy Immunol*. 2016;50(2):189-213. doi:10.1007/s12016-015-8485-8

4. Paradisi A, Ricci F, Sbano P. "Drug-resistant granuloma faciale": treatment with carbon dioxide-GaAs laser. *Dermatol Ther*. 2016;29(5):317-319. doi:10.1111/dth.12365

5. Ortonne N, Wechsler J, Bagot M, Grosshans E, Cribier B. Granuloma faciale: a clinicopathologic study of 66 patients. *J Am Acad Dermatol*. 2005;53(6):1002-1009. doi:10.1016/j.jaad.2005.08.021

6. Marcoval J, Moreno A, Peyr J. Granuloma faciale: a clinicopathological study of 11 cases. *J Am Acad Dermatol*. 2004;51(2):269-273. doi:10.1016/j.jaad.2003.11.071

7. Korkut M. A dermatologic emergency; Sweet's syndrome. *Am J Emerg Med*. 2019;37(9):1807.e1-1807.e3. doi:10.1016/j.ajem.2019.06.012

8. Bastos JT, Rocha CRMD, Silva PMCE, Freitas BMP, Cassia FF, Avelleira JCR. Angiolymphoid hyperplasia with eosinophilia versus Kimura's disease: a case report and a clinical and histopathological comparison. *An Bras Dermatol*. 2017;92(3):392-394. doi:10.1590/abd1806-4841.20175318

9. Adhikari RC, Shah M. Cutaneous leishmaniasis. *Journal of Pathology of Nepal*. 2017;7(2):1212. doi:10.3126/jpn.v7i2.18031

10. Aronson N. Cutaneous leishmaniasis: clinical manifestations and diagnosis. https://www.uptodate.com. Accessed on August 21, 2019.