

**Case Report****Idiopathic lethal granuloma of the midline facial tissues****Troncoso Alcides\*<sup>1</sup>, Maria Laura Rossi<sup>1</sup>, Bava Amadeo Javier<sup>2</sup>****<sup>1</sup>Department of Microbiology, Parasitology and Infectious Diseases, School of Medicine, Buenos Aires University, Argentina****<sup>2</sup>Department of Micology, La Plata University, Argentina****\*Corresponding author: microbiologiayparasitologia@yahoo.com.ar****Abstract**

Granuloma faciale (GF) is a rare chronic inflammatory dermatosis usually appearing only on the face, associated with high morbidity and mortality. Exact etiology is uncertain. We present an illustrative case of GF. A 32 year-old female patient was admitted at the infectious diseases service of the School of Medicine Buenos Aires University for presenting progressive destruction of face, nose, paranasal sinuses, palate, oral and para-oral structures. The intra-oral examination revealed the features of GF. A biopsy at that time showed GF. GF has an unknown etiology, but possible predisposing factors include actinic exposure, radiation, trauma, allergy, or an Arthus-like reaction.

**Keywords:** Granuloma faciale; Facial neoplasms; Oropharynx, Lethal midline**Introduction**

Cases of idiopathic lethal granulomas of the midline facial tissues are being reported in the general medical literature <sup>(1)</sup>. GF is a rare benign chronic inflammatory dermatosis usually appearing only on the face. Lethal midline granuloma (LMG), is a disease that develops in the mouth and nose and spreads rapidly to other parts of the face <sup>(2)</sup>. GS is a devastating disease which destroys the soft and hard tissues of the oral and para-oral structures <sup>(3)</sup>. This devastating lesion may involve the nose, palate, antrum and virtually any part of the face. The facial deformities of those who survive GF are generally serious. Survivors suffer the two-fold afflictions of oro-facial mutilation and functional impairment, which require a time-consuming, financially prohibitive surgical reconstruction <sup>(4)</sup>.

**Case report**

A 32 year-old female patient was admitted with history of progressive destroys the the soft and hard tissues of the mouth, nose, face, paranasal sinuses and neighbouring areas for the last 4 months duration. The peripheral oedema extended to the the upper labial, labial commissural, mandibular and pre-parotid regions. On examination, her general condition was fair. Physical examination showed a erythematous indurated plaque on the her nasal tip and mouth, friable tissue and intense fetidity and anatomical disappearance of the oropharynx, amputated uvula, and absence of both pillars (Fig 1-2). The adjacent palatal mucosa was severely oedematous. Submental, submandibular and cervical lymph nodes were mildly painful upon palpation. The patient was not pyretic. Halitosis was pronounced. The lesions were asymptomatic, and the patient did not report a history of similar lesions or any other lesions on the body. A biopsy of the lesion was performed and sent for

histologic evaluation. Histology confirmed the diagnosis of GF. On hematoxylin and eosin staining there was a dense, inflammatory exudate of predominantly neutrophils and eosinophils, and to a lesser extend lymphocytes and plasma cells in the upper dermis with sparing of the epidermis and pilosebaceous complexes. Ectatic vessels were seen in the dermis with prominent endothelial cells and neutrophils in the vessel walls. A characteristic band of subepidermal collagen or Grenz zone free of inflammatory exudate was noted. GF can be distinguished from erythema elevatum diutinum (EED) by the absence of eosinophils and a Grenz zone in EED. Based on these histologic findings, a diagnosis of GF was made. She dies in hypovolemic shock from severe epistaxis, in few days after her admission.

**Fig 1.** Granuloma Faciale

## Discussion

Granuloma faciale is an uncommon but well-defined cutaneous condition which is seen almost exclusively on the face<sup>(5)</sup>. A granulomatous and necrotic process takes place progressively in this disease that destroys the nose, the perinasal sinuses and other facial structures. The lesions of GF typically present as single, asymptomatic, erythematous, non-changing nodules or plaques. We present an illustrative case of GF<sup>(6)</sup>. Clinically, the lesions appear as well-circumscribed smooth erythematous papules, nodules, or plaques that may increase in size and number. Once fully developed, GF usually does not change<sup>(7)</sup>. LMG is a clinical term, that describes a rare clinical entity of cause unknown, characterized by a progressive and often fatal ulceration with destruction of the superior airways that involves the nose, the paranasal sinuses and the soft tissues of the face<sup>(8)</sup>.



**Fig 2.** Anatomical disappearance of the oropharynx

LMG is an unusual but devastating spectrum of lymphoproliferative disorders that is now thought to include four entities: idiopathic midline destructive disease, Wegener's granulomatosis, polymorphic reticulosis, and nasal lymphoma<sup>(9)</sup>. These disorders must be considered in the differential diagnosis of any patient with chronic, nonspecific complaints localized to the upper airway. An aggressive diagnostic and therapeutic approach may significantly decrease morbidity and death resulting from these lesions<sup>(10)</sup>. This is an uncommon disease and may be produced by different neoplastic, autoimmune and idiopathic diseases. Most of the patients carriers of this disease had a short-term evolution<sup>(11)</sup>. Survivors suffer the two-fold affliction of oro-facial disfigurement and functional impairment. Reconstructive surgery of the resulting deformity is time-consuming and financially prohibitive for the victims who are poor<sup>(12)</sup>.

## Acknowledgements

The authors are grateful to Prof. RG. Edmiston for his corrections and Charlotte McClelland for secretarial assistance.

## References

1. McBri de P. Photographs of a case of rapid destruction of the nose and face. *J Laryng and Otol* 1987; 12: 64.
2. Harold C, Lane M, Dodd K: Idiopathic lethal granuloma of the nose and face. *J Laryng and Otol* 1965; 51: 461-469.

3. Slewarl JP: Progressive lethal granulomatous ulceration of the nose. *J Laryng and Otol* 1933; 48: 657.
4. Trotoux J, Vilde F, Bassot V, Angelard B, Clavier A. Granulome malin centro-facial, problemes diagnostiques et therapeutiques. *Rev Laryngol Otol Rhinol* 1989; 110: 147-150.
5. LeBoit PE. Granuloma faciale: a diagnosis deserving of dignity. *Am J Dermatopathol* 2002; 24:440-443.
6. Requena C, Castejón P, Sanmartín O, Botella-Estrada R, Nagore E, Serra-Guillén C, Sorní G, Guillén C. Rhinophyma-like granuloma faciale. *J Eur Acad Dermatol Venereol* 2006; 20:881-882.
7. 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.
8. Galli M, Formenti A, Pettenati C, De Bartolo G, Mariscotti C, Ponzi S. Immunological features of nonhealing midline granuloma. *ORL J Otorhinolaryngol Relat Spec* 1986; 48: 256-260.
9. Saheeb BD, Ojo MA. Midline lethal granuloma complicating pregnancy: case report. *East Afr Med J.* 2003;80(7): 391-2.
10. Skitarelic N, Dominis M, Matulic Z, Dujella J, Dzelalija B. Lethal midline granuloma. *Lijec Vjesn* 2004;126(5-6): 129-32.
11. Vidal E, Dean A, Alamillos F, Salas J, Lopez R. Lethal midline granuloma in a human immunodeficiency virusinfected patient. *Am J Med* 2001;111(3):244-5.
12. Batra P, Shah N, Mathur S. Midline lethal granuloma—a clinical enigma. *Indian J Dent Res* 2003; 14(3):174-83.