



## Case Report

## A destructive centrofacial granuloma: Case report

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## ABSTRACT

**Introduction:** Centrofacial granulomas have several etiologies, which poses diagnostic difficulties and delays in management. Herein, we report a confusing case revealing granulomatosis with polyangiitis (GPA) in its localized form.

**Case presentation:** A 76-year-old man presented with a pruritic centrofacial placard that had been evolving for 3 years. On examination, there was a centrofacial infiltrated and erythematous papulonodular placard. Skin biopsies were not conclusive. The immunological assessment was negative. The evolution was marked by the extension of the placard, the destruction of the nasal pyramid and nasal mutilation. Only surgical biopsy revealed dermohypodermal, cartilaginous and endonasal non-necrotizing granulomatous tuberculoid inflammation with leukocytoclastic and necrotizing vasculitis. The diagnosis of GPA in its localized form was retained. Treatment with prednisone was initiated combined with monthly boluses of cyclophosphamide. The assessment for systemic involvement remained negative during the follow-up.

**Clinical discussion:** Localized forms represent up to 29% of GPA cases. There are clinical, but also biological differences, since ANCA are found in more than 90% of diffuse forms and only in 50–78% of localized forms. Our case may represent a rare distinctive subset of GPA limited to the facial region and upper airway mucosa but showing a locally aggressive behaviour leading to cartilage and bony destruction.

**Conclusion:** It is necessary to evoke GPA in its localized form and to perform multiple deep biopsies in front of any facial granulomatosis. Early diagnosis and appropriate treatment prevent mutilating and disfiguring sequelae.

## 1. Background

Centrofacial granulomas are characterized by a double definition: clinical by the presence of extensive ulcerations and necrotic lesions located at the level of the upper airways evolving towards the destruction of the mid-facial region; and histological by the presence of an epithelio-giganto-cellular granuloma. Etiologies are multiple: infectious, inflammatory and tumoral, which poses diagnostic difficulties and delays in management. We report a confusing case revealing granulomatosis with polyangiitis (GPA) limited to the facial region and upper airway mucosa, lacking systemic involvement after a prolonged follow-up but showing locally aggressive behaviour. The work has been reported in line with the SCARE 2020 criteria [1].

## 2. Case report

A 76-year-old man, with no significant past medical history, presented with a pruritic centrofacial placard that had been evolving for 3

years (Fig. 1), progressively extending to the cheeks and forehead, causing intermittent nasal obstruction initially, followed by hyposmia, hypoguesia, and hypoacusis.

On examination, there was a centrofacial infiltrated and erythematous papulonodular placard, taking the nose, both cheeks and the forehead, associated with fine telangiectasias and lupoid papules.

Multiple skin biopsies revealed large perifollicular lymphoplasmacytic infiltrates with epithelioid and gigantocellular granuloma outlines, without evidence of vasculitis. After biopsy, several diagnoses have been considered such as: granulomatosis with polyangiitis, extranodal NK/T-cell lymphoma, pyoderma gangrenosum, sarcoidosis, granulomatous rosacea, leprosy or tuberculous lupus.

Hemogram and chest X-ray were normal. The immunological assessment (including c-ANCA) and 24-h proteinuria were negative, as was the nasal smear for Hansen's bacillus. One month after his first visit, the evolution was marked by the extension of the placard, the destruction of the nasal pyramid and nasal mutilation (Figs. 2 and 3). In view of a phlyctenular tuberculin intradermal reaction and a positive

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Fig. 1. Centrofacial erythematous plaque made of lupoid papules and telangiectasias.



Fig. 3. Evolution over time of the centofacial granuloma.



Fig. 2. Evolution over time of the centofacial granuloma.

quantiferon, the diagnosis of tuberculous lupus was retained and the patient received 6 months of antibacillary drugs with worsening of the lesions. Subsequently, a surgical biopsy revealed dermo-hypodermal, cartilaginous and endonasal non-necrotizing granulomatous tuberculoid inflammation with leukocytoclastic and necrotizing vasculitis.

The diagnosis of GPA in its localized form was retained on the basis of ENT symptomatology, facial ulceration and the presence of granulomatous inflammation with leukocytoclastic and necrotizing vasculitis lesions on endonasal biopsy. Treatment with prednisone was initiated at a dose of 1 mg/kg/d combined with monthly boluses of cyclophosphamide. During the follow-up, the assessment for systemic involvement remained negative. Unfortunately, the patient died after 6 months.

### 3. Discussion

Centrofacial granulomas derive from multiple etiologies: infectious, inflammatory, and tumoral; which poses diagnostic difficulties that lead to delayed management [2].

One such etiology is GPA (formerly called Wegener's disease). This is a systemic necrotizing vasculitis of small and medium-sized vessels, according to the Chapel Hill nomenclature (2012), typically associated with the presence in the serum of approximately 90% of patients of autoantibodies directed against the cytoplasm of the neutrophils with cytoplasmic reinforcement in indirect immunofluorescence (cANCA), and antiproteinase 3 (PR3) specificity [3,4].

GPA presents a wide spectrum of manifestations and remains one of the most challenging diagnostic dilemmas in clinical medicine, especially when it presents with isolated symptoms such as ocular disease or oral ulcers [5-7]. Clinically, the most characteristic manifestations are ENT, pulmonary and/or renal involvement [5,6]. Mucocutaneous involvement has been reported in approximately 23% of patients, palpable purpura being the most common finding. Less commonly, skin

ulcers, cutaneous vasculitis, urticarial, and erythematous nodules may be present [8,9].

An important feature of our case is just that GPA presented as localized, albeit aggressive disease without systemic spreading. A distinction can be made between diffuse and localized forms, the first one is more related to vasculitic phenomena and the second one to granulomatous inflammation [10,11]. Limited and severe subsets of GPA have been described by The Wegener's Granulomatosis Etanercept Trial Research Group [12]. It represents up to 29% of Wegener's granulomatosis cases [10]. Patients with limited disease were observed to have an earlier onset, a longer disease duration, and an increased risk of disease exacerbation following remission. A higher frequency of non-organ-threatening upper airway manifestations (sinus involvement, nasal septal perforations, nasal collapse) was also observed in the limited disease phenotype. In comparison, patients with severe disease have features of significant alveolar hemorrhage, renal involvement, and nervous system manifestations.

Finally, c-ANCA, which are considered the marker for multisystem GPA but have been reported to be absent in up to 40% of GPA cases and particularly in those lacking renal involvement, resulted negative in our patient [13,14]. In contrast, other studies found a high frequency of ANCA-positive patients with localized disease, making unlikely ANCA negativity as the laboratory hallmark for this subset [15,16].

#### 4. Conclusion

Our case represents a rare distinctive subset of PGA limited to the facial region and upper airway mucosa but showing a locally aggressive behaviour leading to cartilage and bony destruction. Therefore, is necessary to evoke granulomatosis with polyangiitis in its localized form and to perform multiple deep biopsies in front of any facial granulomatosis, especially after a well conducted treatment with unfavorable evolution. Early diagnosis and appropriate treatment prevent mutilating and disfiguring sequelae.

#### Patient consent

Since the patient died, the consent was given by the patient's son.

#### Ethical approval

This study complies with internationally-accepted standards for research practice and reporting.

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#### Author contribution

Zineb Tazi Saoud: Drafting of the manuscript. Fatima Zahra Elfatoiki: Critical revision of the manuscript for important intellectual content. Soumiya Chiheb: Critical revision of the manuscript for important intellectual content.

#### Registration of research studies

1. Name of the registry:
2. Unique Identifying number or registration ID:
3. Hyperlink to your specific registration (must be publicly accessible and will be checked):

#### Guarantor

Zineb Tazi Saoud.

I, Zineb Tazi Saoud, corresponding author of the manuscript, accept full responsibility for the work and the conduct of the study, had access to the data, and controlled the decision to publish.

#### Consent

Written informed consent was obtained from patient's son for publication of this case report and accompanying images, since the patient died.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Declaration of competing interest

No conflicts of interest.

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