Midline destructive lesion. Case report

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Abstract

Introduction: Midline destructive syndrome is an entity characterized by a group of signs and symptoms, secondary to the condition, usually destructive and located in the middle of the face.

Objectives: Clinical case report and literature review.

Case Report: 38-year-old male patient, from Chaco, with mucocutaneous Leishmaniasis diagnosis, who was hospitalized due to a midline destructive lesion. After multiple diagnostic options and follow-up studies, the final diagnosis of granulomatosis with polyangiitis (Wegener's granulomatosis) is reached.

Discussion: These patients pose a diagnostic challenge in internal medicine due to the multiple etiology factors that can be responsible for its development, with similar clinical symptoms but with a different treatment and prognosis.

Key word: Midline destructive lesion. Wegener's granulomatosis.

Received: March, 14th, 2013. Accepted: July 20th, 2013.

Introduction

The midline destructive lesion is an entity characterized by a group of signs and symptoms secondary to pathologies that cause inflammation and necrosis of the upper respiratory tract both suprapalatal and infrapalatal, orbital floor, paranasal sinuses, ethmoid bones, vomer, maxillary, inferior and medium turbinates, nose and adjacent structures bones. It is described in 6 of every 10,000 inhabitants1 and, more frequently,
within the fifth and sixth decades of life. It prevails in women and has been observed in all races.

It manifests as fast-progression ulcerations and perforation, though, in some cases it may have chronic evolution and consequences in time. Patients show symptoms that suggest diseases of the superior airways such as rhinorrhea with or without nose deformity, as well as ulceration of nasal mucosa, mouth or gum. It is infrequent for patients to show ocular symptomatology due to orbital compromise.

These patients pose a diagnostic challenge in internal medicine due to the multiple infectious, oncologic-hematological and rheumatologic etiologies that may cause the disease, as well as to the aggressive behaviour many of them have.

**Objectives**

Clinical case report and literature review that may serve as a guideline to assess patients with diseases affecting the central structures of the face.

**Case Report**

38-year-old male patient, from Chaco, without relevant personal or family history. His symptoms had started one year before the consultation with photophobia, epiphora, reduction of visual acuity and influenza-like symptoms associated with pustular cutaneous lesions which evolve to punch ulcers. He was hospitalized in Chaco province where a biopsy of one of the cutaneous lesions was taken and cytoplasmic inclusions “compatible” with *Leishmania sp.* were evidenced. He was under treatment with meglumine antimoniate and, then, with liposomal amphotericin B; since a regular response was obtained, a second cutaneous biopsy was performed and reported pyoderma gangrenosum.

He started treatment with prednisone 1 mg/kg/day plus amphotericin B as a double dose and, as his answer was favorable, he was released from hospital. Soon afterwards, he presented frontal headache and severe compromise of bilateral vision with necrotizing scleritis, panuveitis, epistaxis and nasal septum destruction. A nuclear magnetic resonance showed severe ocular and sinonasal lesions.

Within that context, he was referred to our institution. On admission, physical examination showed saddle nose, secondary to nasal septum destruction (fig 1.), thorax pustular cutaneous lesions, as well as other lesions with cicatricial appearance (Fig 2), bilateral endophthalmitis with blurred vision in both eyes. (Fig 3). Routine tests, as well as other parameters that could guide the etiologic diagnosis, were required. The results were as follows: leukocytes 11,100 /uL. Conserved formula, microglobulin 2300 ng/mL. Proteinuria 0.18 g/24 hs. Rheumatoid factor <5UI, C3 18 mg%, C4 29 mg%, ANA (-), ASMA (-), ANCA C and P (-). Negative viral serologies and VDRL. VSG: 107mm/h. Ferritine 546 ng/dl.

![Figure 1 Photograph of the patient showing the typical saddle nose consequence of nasal septum destruction](image-url)
Lesions with irregular edges and cicatricial appearance and other pustular lesions the patient had.

Facial bones CT scan showed mucous and bone compromise of paranasal sinuses (fig 4). Cave biopsies were taken for pathologic anatomy (PA) and showed an intense inflammatory process and nonspecific lymphoplasmacytic infiltration; culture for bacteria, fungus and leishmania ssp. was negative. In order to rule out the possibility of an oncologic and hematological pathology, immunofenotipification was required in peripheral blood and in cave lesion biopsy, which did not show fenotipical findings compatible with clonal processes in progress. The cutaneous biopsy which had been obtained in Chaco was required with revision purposes, and the presence of the previously reported amastigotes was ruled out. Finally, with the histopathologic findings obtained from this sample, the diagnosis of granulomatosis with polyangiitis (Wegner’s granulomatosis) was reached (Fig 5 and 6).

He started treatment with meprednisone 40 mg/day and the answer was favorable but, as he had progressive ocular activity, he started treatment with pulsed-cyclophosphamide. Six months after the diagnosis had been made and once under treatment with oral cyclophosphamide and descending corticoids, the disease stopped its progress and improved clinical and laboratory parameters were observed.
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Discussion

Although the clinical symptoms of all midline destructive lesions are similar, it is during prognosis and treatment that the heterogeneity of the causes appear. Hence the importance of reaching an accurate etiologic diagnosis in order to start with the appropriate treatment as soon as possible.

A good way of approaching the etiologic diagnosis is grouping the wide range of diagnostic possibilities, according to their etiopathogenic mechanism, into infectious, immunologic, oncologic and other causes. (Refer to Table 1).

Each one of these entities will have risk factors and clinical symptoms which will be indicative of diagnosis. Therefore, according to interviews, history and epidemiologic data, these follow-up studies should be required:

- Serologies Ebstein Barr, HIV; VDRL.
- Collagen profile (ANCA C, ANCA P PR3)
- Electrophoresis proteinogram, Bence-Jones proteinuria, immunohistochemistry and cytometry of blood cells (with suspicion of oncologic and hematological diseases).
- Erythrocyte sedimentation rate and other acute phase reactants.
- Cultures for bacteria, micobacteria, fungus and parasites (according to epidemiology).
- Histopathological evaluation, for which more than a biopsy may be required due to the technical difficulty of obtaining significant samples without compromising the central structures of the face.

The following imaging follow-up studies are necessary to determine the extension of the lesion:
1) Conventional radiology of paranasal sinuses: where rarefaction and thickness reduction of sinus walls can be observed.
2) Computed tomography of skull, paranasal orbits and sinuses: where progressive volume increase of septum is observed as a manifestation of non-specific anti-inflammatory reaction. Diffuse inflammation of sinus mucosa with bone reaction of walls is an indication of a granulomatous lesion that may require a biopsy for anatomopathologic studies.
There are two possibilities that may hinder diagnosis: The first one is midline destructive idiopathic disease that is noticed in 5 to 15% of cases. It should be considered in patients with histopathologic evaluation who show active chronic nonspecific inflammation, but do not show systemic manifestations or follow-up studies that may refer to a definitive diagnosis. It is believed that this entity results from a fulminating inflammatory answer to an unknown antigen. It is fatal when treatment is not followed, which consists of a local radiation therapy. The second possibility is the existence of etiology combinations such as neoplasias, necrosis and added infection, as well as secondary vasculitis. Therefore, we may conclude that the etiologic diagnosis of any midline destructive lesion is fundamental to start a specific treatment as soon as possible due to the aggressiveness of many of said etiologies. To do so, high clinical suspicion, lab tests, cultures and mainly anatomopathologic studies are necessary and may be required considering the interview, personal and epidemiologic history.

**Literature**


**Table 1**: It groups all possible causes of midline destructive lesions in infectious (within them viral, bacterial, mycotic and parasitic causes), neoplastic, inflammatory, toxic and other causes.

<table>
<thead>
<tr>
<th>INFECTIOUS</th>
<th>VIRAL:</th>
<th>Epstein-Barr</th>
</tr>
</thead>
<tbody>
<tr>
<td>BACTERIAL</td>
<td>• Tularemia, • syphilis, • actinomycosis. • Tuberculosis, leprosy and other Mycobacteria</td>
<td></td>
</tr>
<tr>
<td>MYCOTIC</td>
<td>• Mucormycosis (in diabetic patients) • Candidiasis • Histoplasmosis • Blastomycosis • Coccidioidomycosis • Rhinosporidiosis • Aspergillosis</td>
<td></td>
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<tr>
<td>PARASITIC</td>
<td>Mucocutaneous Leishmaniasis Myasis</td>
<td></td>
</tr>
<tr>
<td>NEOPLASTIC</td>
<td>• Nasal lymphomas (express immunophenotypings CD4, CD20 andD45) 1 • Nasopharyngeal carcinoma • Melanoma of superior airways • Metastatic lesions (thyroids, kidney, lungs, intestinal and genitourinary tract) • Rhabdomyosarcoma and other such as the histiocytic sarcoma</td>
<td></td>
</tr>
<tr>
<td>INFLAMMATORY</td>
<td>• Wegener’s granulomatosis (granulomatosis with polyangiitis) • Midline destructive idiopathic disease. • Sarcoidosis</td>
<td></td>
</tr>
<tr>
<td>TOXIC</td>
<td>• Cocaine inhalation • Inhalation of other toxic substances.</td>
<td></td>
</tr>
<tr>
<td>OTHER</td>
<td>• Polymorphic reticulosis (CD 43+) or inflammatory granulomatosis. Prelymphomatous lesions associated with T cell nasal lymphoma 1 • Midline destructive idiopathic disease</td>
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