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Case report

Chronic rhino-orbito-cerebral mucormycosis: A case report and review of the literature

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HIGHLIGHTS

- We describe the case of a chronic rhino-orbito-cerebral mucormycosis.
- Chronic mucormycosis has low rate of frequency and is difficult to diagnose.
- A quarter of chronic rhino-orbito-cerebral mucormycosis cases are in immunocompetents.
- Surgical debridement is the corner stone of mucormycosis treatment.
- Survival rate in chronic mucormycosis cases (83%) is higher than acute cases (10–35%).

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ABSTRACT

Mucormycosis is a life-threatening disease, were rhinocerebral infection is most commonly seen in the clinical setting. Chronic mucormycosis is a rare presentation that exhibits a challenging diagnosis. We describe the case of a 47 year old diabetic man with complains of left zygomatic arch swelling of 3 months evolution. He had received previous antibiotic treatment without improvement. Biopsy of maxillary sinus revealed the presence of non-septated, 90° angle branched hyphae compatible with zygomicetes. The patient was treated with surgical debridement and amphotericin B until there was no evidence of fungi in the tissue by biopsy. We reviewed chronic rhino-orbito-cerebral mucormycosis from 1964–2014 and 22 cases were found, being this the second case of chronic mucormycosis reported in Mexico. A quarter of the cases were seen in immunocompetent hosts. As only 20% of the causal agent can be isolated by culture, the diagnosis is mainly made by biopsy. Besides treatment with amphotericin B, posaconazole as alternative, and control of the underlying comorbidities, surgical debridement represents the corner stone therapy. We recommend at least 36 month follow-up, due to the 13% risk of recurrence. A chronic presentation has a general survival rate of approximately 83%.

1. Introduction

Mucormycosis is an invasive infection caused by filamentous fungi belonging to the order Mucorales with rhinocerebral infection being the most common clinical presentation [1]. According to time of evolution the infection can be acute or chronic, with the latter having a low frequency (5.6% of rhinocerebral mucormycosis cases). [1,2].

We present a case of chronic mucormycosis rhinocerebral

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infection in a 47yo male and a review of literature. To our knowledge, this is the second case of chronic mucormycosis reported in Mexico [3].

2. Presentation of case

A 47-year-old male was evaluated because of a 3-month history of paresthesia, pain and swelling in the left zygomatic bone. He had previously attended the clinic on several occasions where he was diagnosed with type 2 diabetes mellitus (DM2) and sinusitis, receiving unspecified antibiotic treatment. Subsequently, he came to our institution with swelling on the left half of his face, and erythema and pain on palpation. A CT scan of his paranasal sinus (Fig. 1) revealed an erosion of the malar bone and the lateral orbit wall, edema of preseptal tissue without involvement of intraorbital structures. The patient was subjected to several biochemical tests and presented a white blood cell count of 10.3 Kc/mL, platelets 27 Kc/mL, BUN 10 mg/dL, serum creatinine 0.8 mg/dL, and a serum glucose level of 239 mg/dL. Biopsies were obtained during a Cadwell-Luc procedure. Microscopically the tissue of the maxillary sinus revealed the presence of non-septated 90° angle branching hyphae compatible with zygomycetes (Figs. 2 and 3). For this reason debridement with a modified midfacial degloving approach of the left maxillary mucosa, the anterior and lateral maxillary walls, and the adjacent area of the malar bone was performed; the floor of the orbit was preserved. Biopsy revealed presence of hyphae zygomycetes, with some areas of acute infiltrate, granulomatous chronic infiltration and regions with evident angioinvasion.

In addition, Ziel-Nielsen and periodic acid-Schiff stains were carried out and both were negative. Amphotericin B deoxycholate therapy was administered with the patient receiving a total dose of 2575 mg in 26 days. The patient presented tubulointerstitial nephritis (Serum Creat 2.5 mg/dl) and was assessed by a nephrologist. In consensus with the patient, it was decided to continue the same therapy (because of a lack of posaconazole in our setting) until evidence of mycotic tissue was absent, achieved by left type III antrostomy and biopsy of surgical bed tissue. During evaluation, a tissue culture was done but there was no growth of fungi or mycobacteria. At four months follow-up, there has been no evidence of recurrence and the patient is currently asymptomatic.

Fig. 1. Bone erosion of left malar bone is identified in the computed tomography scan with extension to the lateral orbital rim, zygomatic arch and ipsilateral palate, associated with edema of preseptal soft tissue, the presence of small subperiosteal collection upper outer margin, with occupation of the maxillary sinus, without enhancement after contrast administration.

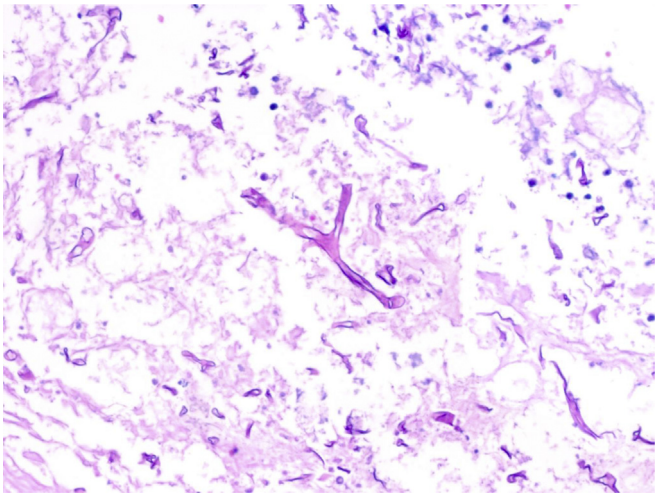

Fig. 2. Irregular, thick, non-septate and fragmented hyphae between the necrotic tissue. PAS 40x.

3. Discussion

We searched PubMed database from 1964 to 2014 for all available articles in the English language related to rhino-orbital-cerebral chronic infections caused by fungi of the order Mucorales and found 22 cases. In Table 1 we summarize the characteristics of the reported chronic rhino-orbito-cerebral cases.

Mucormycosis is an opportunistic infection that affects immunosuppressed patients with a decreased ability to phagocytize. [1,4] It also has been described in immunocompetent patients with an incidence of 4–19%. [5–8] The infection originates in the upper airway by inhalation of spores. [1,9] Its pathogenic form was recognized in 181510, with the first case of mucormycosis in humans being described by Palauf in 1855.11,12 The first documented case of chronic mucormycosis was described by Vignale in 1964.2.

The infection can present in locations such as the skin, lung, gastrointestinal tract, and as a rhino-orbito-cerebral infection, this being the most common. [4,11,12] In our review we found that 26% of chronic rhino-orbito-cerebral mucormycosis occurred in immunocompetent patients. Initial clinical manifestations include

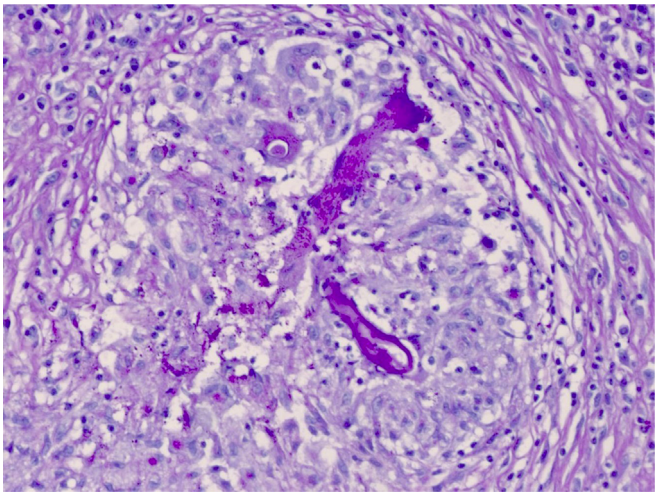

Fig. 3. Granuloma image consisting of lymphocytes and epithelioid cells, highlighting its central portion a non septate hyphae. PAS 40x.