Successful Management of Extensive Rhino-Orbito-Cerebral Mucormycosis: Prompt Neuroimaging-Supported Diagnosis and Aggressive Antifungal and Surgical Treatment

Abordagem Bem-Sucedida de Mucormicose Rino-Orbito-Cerebral Extensa: Diagnóstico Radiológico Célere, Terapêutica Antifúngica e Cirúrgica Agresssiva

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Mucormycosis is a life-threatening fungal infection caused by fungi from the zygomycetes class, often affecting immunocompromised individuals. It is a rare infection in Europe, with an estimated incidence of 0.04 – 0.12 cases/100 000 people annually.[1-3] Worldwide, rhino-orbital-cerebral mucormycosis is the most common presentation and has a poor prognosis.[1,3,4] Extensive disease is associated with high mortality rates (40% - 80%). In these cases, early aggressive antifungal and surgical treatment can be lifesaving.[5] Since the confirmation of diagnosis can be delayed (involving histopathological analysis and/or culture of samples), imaging studies can be key to reach a diagnosis.

We present a case of rhino-orbital-cerebral mucormycosis in a 61-year-old woman with well-controlled diabetes, sarcoidosis treated with prednisolone 40 mg/day in the last month, and a history of breast cancer (treated, without recurrence). The patient developed a fever, cough with purulent sputum, nasal obstruction, sinus pain, hematic rhinorrhea, periocular inflammation, and a persistent headache over the previous two weeks. She was found unconscious and admitted with a Glasgow Coma Scale score of 13, total right ophthalmoplegia, periocular edema, and purulent discharge. Imaging studies revealed severe sinus and orbital involvement, as well as signs of cerebral ischemia and a pseudoaneurysm. Given her immunosuppressed state, invasive fungal infection was suspected, and she was transferred to a tertiary center for a multidisciplinary surgical approach. Corticosteroid therapy was discontinued, and within 48 hours she was promptly started on liposomal amphotericin B. She underwent three extensive surgeries, including right orbital exenteration and neurosurgical debridement.

An ethmoidal sinus biopsy was performed, followed promptly by a polymerase chain reaction assay, which tested positive for order *Mucorales*. Histological analysis confirmed the presence of *Mucor* zygomycetes, confirming the diagnosis. Unfortunately, fungal cultures from this sample were non-viable. The patient was kept on liposomal

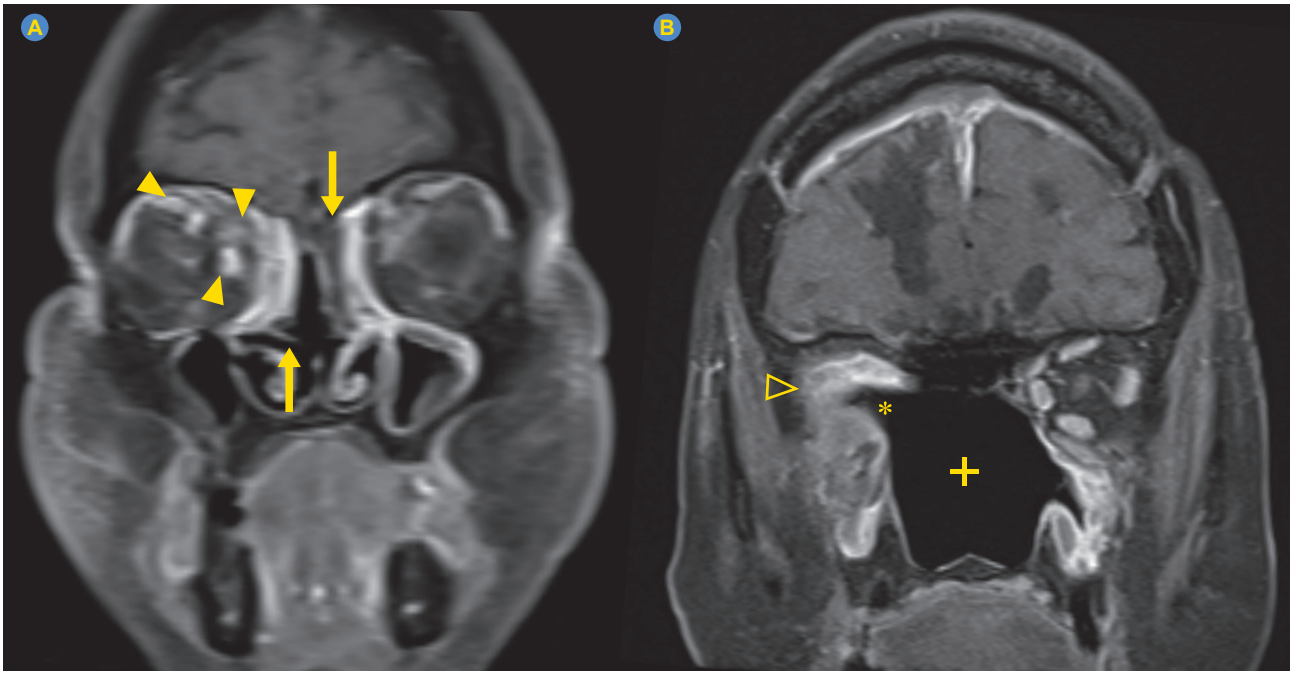

Figure 1 – Magnetic resonance imaging (MRI) at diagnosis, coronal post-contrast T1 Dixon Water-Only (A). Extensive inflammatory/infectious filling of the ethmoidal air cells and the superior parts of the nasal cavities, and maxillary sinus mucosal thickening. Notably, there is minimal or no contrast enhancement of the nasal mucosa, as well as the middle turbinates, known as the 'black turbinate sign' (indicated by arrows), in regions of low signal on T2 STIR (not shown) which is suggestive of acute invasive fungal rhinosinusitis. Right orbital involvement is seen reflected by signal abnormalities of the intraorbital fat alongside edema (on T2 STIR, not shown), thickening and increased contrast uptake of the medial and superior rectus and superior oblique muscles (indicated by arrowheads). Additionally, there is evidence of slow flow of the superior ophthalmic vein. Follow-up MRI (1 year later), coronal post-contrast T1 Dixon Water-Only (B), reveals the extent of surgical interventions: right orbital exenteration (indicated by asterisk), total and bilateral ethmoid-maxillo-sphenoidectomy, turbinate and nasal septum resection (indicated by plus sign), and bilateral resection of the gyrus rectus with bifrontal cranioplasty. Residual inflammation is noted in the right orbital compartment (indicated by open arrowhead) and fronto-basal region, but there are no signs of an active infectious process.

amphotericin 5 to 10 mg/kg for 52 days, followed by isavuconazole 200 mg. She was discharged on oral isavuconazole, maintained for two years and stopped due to sustained imagiological and clinical improvement. The patient has adapted well to the sequelae of her extensive surgery. She has preserved eyesight in her remaining eye and performs her daily activities autonomously with good support from her family.

This case emphasizes the importance of rapid recognition of mucormycosis from clinical and imagological findings and prompt surgical and medical treatment. Imaging plays a crucial role in evaluating disease extent, particularly with rhino-orbital-cerebral involvement, and follow-up evaluation during treatment.[5] Aggressive management, including antifungal therapy, surgical debridement, and control of underlying conditions, can lead to favorable outcomes, even in the presence of extensive disease.[4,5]

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AUTHOR CONTRIBUTIONS

ECS: Study design, writing and critical review of the manuscript.
ACM: Writing and critical review of the manuscript.
SC: Image collection, critical review of the manuscript.
All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

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