Rhinocerebral Mucormycosis of the Optic Nerve

Jameson K. Mattingly, MD and Vijay R. Ramakrishnan, MD

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

Keywords
rhinocerebral mucormycosis, optic nerve

Received May 4, 2016; revised June 13, 2016; accepted June 14, 2016.

A 55-year-old man initially presented with progressive right-sided vision loss during hospitalization following revision liver transplant. He underwent treatment with high-dose corticosteroids for presumed optic neuritis, with initial vision improvement that subsequently progressed to complete visual loss. He was then transferred to our facility. Repeat radiographic analysis demonstrated left maxillary sinus disease, enhancement of the right orbital apex, and enlargement of the optic nerve without associated sphenoid mucosal disease (Figure 1A, B). Due to his history of immunosuppression and the diagnostic dilemma, we elected to explore his left nasal cavity and paranasal sinuses, where necrotic tissue was discovered within the middle meatus and maxillary sinus. Given this finding, along with the presence of his right-sided vision loss and the findings on imaging, his right nasal cavity and sphenoid sinus were examined. This revealed an ulcerative lesion of the optic nerve (Figure 1C). Based on pathologic analysis of this lesion, along with the contents from his left-sided surgery, he was diagnosed with invasive fungal sinusitis (Figure 1D). Fungal polymerase chain reaction identified Rhizopus oryzae as the causative organism. His treatment was multimodal, consisting of antifungal therapy and limiting immunosuppression. At 4-month follow-up, he was alive without disease, but his vision did not return.

This study was approved by the Colorado Multi-institutional Review Board (13-1555).

Discussion

Acute invasive fungal sinusitis is an aggressive and often fatal angioinvasive infection commonly involving the paranasal sinuses. Risks factors for acquiring this infection include chronic immunosuppression, hematologic malignancies, and uncontrolled diabetes mellitus. Common causative organisms in the paranasal sinuses include Zygomycetes (Mucor, Rhizopus) and Aspergillus species. These organisms are usually associated with decomposing organic matter but can colonize the aerodigestive tracts in humans and become pathologic under ideal circumstances.

Presenting symptoms can be quite variable, which may lead to a delay in diagnosis. These can include nasal congestion, facial swelling and discomfort, fever, ophthalmoplegia, vision loss, headaches, and cranial neuropathies—all of which can progress rapidly. Early diagnosis is key to initiate aggressive surgical and medical therapy, the latter of

Figure 1. Localization of disease to the orbital apex and optic nerve is noted on coronal postcontrast magnetic resonance imaging (A) and noncontrast computed tomography (B), where enlargement of the nerve and enhancement of the surrounding soft tissue are seen (arrowheads) within a normal-appearing sphenoid sinus. Endoscopic visualization upon opening the sphenoid sinus demonstrated focal ulceration over an exposed optic nerve (arrow; note perineural blood vessel). Pathologic analysis confirmed invasive fungal hyphae consistent with Rhizopus (arrowheads, D).

1Department of Otolaryngology, University of Colorado School of Medicine, Aurora, Colorado, USA
2Department of Neurosurgery, University of Colorado School of Medicine, Aurora, Colorado, USA

Corresponding Author:
Jameson K. Mattingly, MD, Department of Otolaryngology, University of Colorado School of Medicine, 12631 E 17th Ave, B205, Aurora, CO 80045, USA.
Email: jameson.mattingly@ucdenver.edu
which includes providing intravenous antifungal antibiotics and controlling contributing underlying disease processes, if able. Despite aggressive treatment, the overall mortality rate remains high at approximately 50%, with a slightly better prognosis for diabetic patients and a poorer one for those with intracranial involvement and for those without surgery as part of their therapy.¹

**Author Contributions**

Jameson K. Mattingly, design of work, drafting and revising; Vijay R. Ramakrishnan, design of work, revising, final approval.

**Disclosures**

**Competing interests:** Vijay R. Ramakrishnan, Medtronic, Inc—consultant; NeilMed Pharmaceuticals—prior research grant funding.

**Sponsorships:** None.

**Funding source:** None.

**Reference**