Successful Right Orbital Exenteration for Mucormycosis in a Young Woman

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Abstract Purpose. We report successful treatment of a 32-year-old woman with a history of acute lymphoblastic leukemia who presented with rhino-orbital mucormycosis and successfully underwent right orbital exenteration and treatment of her mucormycosis. Mucormycosis is a severe fungal infection that is often fatal in immunosuppressed patients. The mainstay of therapy is intravenous amphotericin B and surgical debridement of affected tissue. Methods. Treatment included intravenous antifungal agents, debridements with local drug delivery, hyperbaric oxygen, deferasirox, and right orbital exenteration. Results. Integration of numerous therapeutic modalities led to a successful outcome. The patient is alive and well more than 3 years later. Conclusions. Surgical excision is traditionally considered the only treatment for mucormycosis. This case is a reminder of the adjunctive forms of treatment.

Keywords fungal eye infections; mucormycosis; orbital exenteration

1 Introduction

Mucormycosis is a severe fungal infection that is often fatal in immunosuppressed patients [5]. The mainstay of therapy is intravenous amphotericin B and surgical debridement of affected tissue [2]. We report a case of mucormycosis that shows the value of adjunctive treatments.

2 Case report

A 32-year-old woman with a history of acute lymphoblastic leukemia (ALL) initially presented to her primary physician with a 2-week history of jaw pain. With symptoms refractory to oral antibiotics, she was admitted to the hospital for a suspected jaw abscess.

On presentation, she exhibited notable black necrotic areas in the hard palate. She was afebrile, neutropenic, and hyperglycemic. Initial treatment included broad-spectrum intravenous antibiotics. On hospital day 2, biopsy of her hard palate revealed phycomycoses of the sinuses. Amphotericin lipid complexes, 5 mg/kg daily, were initiated. Magnetic resonance imaging (MRI) of the brain/sinus was consistent with sinusitis. The patient underwent excisional sinus debridesments by otolaryngology.

On day 4, the patient’s ocular examination was positive for right-sided periportial edema, proptosis, chemosis, and vision of 20/200 (previously 20/20) OD and 20/50 OS. Her right pupil was dilated with minimal reaction to light. Extraocular movements were restricted by 20% OD. Funduscopy was normal. MRI of the brain showed inflammation extending to the orbital apex effacing the right optic nerve (Figure 1).

By day 5 of therapy, vision in her right eye had improved to 20/50. The otolaryngology team continued bedside debridements and irrigation with amphotericin B. The patient also received granulocytes to strengthen her immunity.

On hospital days 6 and 7, the patient was able to open her eyes spontaneously. By day 9, however, her clinical status declined. Visual acuity deteriorated to no light perception.

Figure 1: Magnetic resonance imaging of the brain showed proptosis, inflammation extending to the orbital apex effacing the right optic nerve, and opacification of the right ethmoid and sphenoid air cells.
Pathologic analysis of the orbital exenteration specimens was positive for Zygomycetes (Figure 2). The cultures did not grow a specific species throughout the course of the case, but the Centers for Disease Control confirmed a 100% match by polymerase chain reaction and sequencing identification to Rhizopus oryzae. Three months later, the patient was discharged home and is doing well more than 3 years later. She is on maintenance chemotherapy and posaconazole without evidence of either disease relapse.

3 Discussion
Surgical excision is traditionally considered the only treatment for mucormycosis; however, this case is a reminder of the adjunctive forms of treatment. Unfortunately, even with these advances, we were unable to ultimately avoid orbital exenteration.

Diagnostically, selective involvement of extraocular muscles seen on neuroimaging is uncharacteristic of mucormycosis (Figure 1). Along with the conjunctiva specimen, which showed inflammation and fibrosis, the orbital biopsy showed reactive epithelium, inflammation, sloughing cells, and some polymorphonuclear cells. Inflammatory cells were also found in the skeletal muscle. Some of the cells were suggestive of reactive lymphocytes, indicating possible leukemia. Special stains of CD10, terminal deoxynucleotidyl transferase, CD3, CD20, CD5, and CD68 all yielded negative results. All these findings led us to try to save the orbit with more conservative treatment. Once it was apparent that the fungal infection reached the orbital border, it was obvious that exenteration was unavoidable. Our patient survived mucormycosis despite her immunosuppressed status. Her treatment required different modalities including intravenous antifungal agents, debridements with local drug delivery [1,4], hyperbaric oxygen, and deferasirox. Systemic control of blood sugar, coagulation, immunity, and hemoglobin were also important for the successful treatment.

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References