Mucormycosis is a rare fungal infection caused by one of the species in the Mucorales order that usually affects immunocompromised patients. These fungi are ubiquitous in nature and cases have been reported all over the world. Mucormycosis most commonly affects patients with uncontrolled diabetes mellitus, those with hematologic malignancy undergoing chemotherapy, and patients that have received allogeneic stem cell transplants. Additional risk factors include severe neutropenia, iron overload, major trauma, and prolonged steroid use (1). The most commonly affected sites are the sinuses, followed by the lungs and skin (1). These fungi are angioinvasive, with infection resulting in tissue necrosis and thrombosis. Presenting symptoms of rhinocerebral mucormycosis may include those of rhinosinusitis or orbital cellulitis. Progressive infection can cause cranial nerve deficits, facial pain, headache, and vision changes. Classic physical exam findings also include a black, necrotic middle turbinate or a necrotic eschar of the hard palate. Diagnosis is made by tissue biopsy showing angioinvasion by fungal hyphae. Surgical treatment of rhinocerebral mucormycosis consists of aggressive, endoscopic sinus surgery to remove necrotic tissue. Amphotericin B is considered first-line antifungal therapy, with posaconazole or isavuconazole used in patients with refractory disease, amphotericin B intolerance, or for de-escalation (2). Successful management of invasive rhinocerebral mucormycosis requires early diagnosis, surgical debridement, antifungal therapy, and control of underlying immunocompromise.

Case Presentation

A 55 year old man with type 2 diabetes mellitus presented with dental pain and left facial swelling. He was found to be in diabetic ketoacidosis. He developed diplopia with left gaze and an MRI was obtained which showed opacification of the left sphenoid sinus and mucosal thickening of the left ethmoid and maxillary sinuses. His acidosis was corrected and he developed diplopia with left lateral gaze as well as left sided facial numbness in the distribution of cranial nerves V2 and V3. Repeat CT head showed no intracranial findings. MRI brain showed left sphenoid and ethmoid opacification and myositis of the left nasopharyngeal and masticator space as well as narrowing of the cavernous left internal carotid artery. The patient underwent left maxillary antrostomy, total ethmoidectomy, sphenoid sinustomy, and fronto-sinus exploration. Intraoperative findings showed necrotic mucosa and debris in the lateral recess of the sphenoid and histopathology showed invasive mucormycosis. The patient was started on liposomal amphotericin B and two days later developed weakness of left cranial nerve VII. He was transferred to the intensive care unit for aggressive blood sugar management. His hyperglycemia was controlled and his facial pain and weakness worsened. Micafungin was added and serial MRIs showed worsening inflammation and infiltration of the skull base and intracranially. Eight days postoperatively, the patient was started on ivasconazole in addition to amphotericin B and other antibiotics were discontinued. The patient completed 6 weeks of amphotericin B and 11 months of ivasconazole. Over the following months, he developed facial paresthesia and burning as his facial sensation began to return. His face progressed from House-Brackmann 6/6 to 3/6 over the course of a year. In the eighth month after his diagnosis, he completed 30 treatment sessions of hyperbaric oxygen therapy (5). Hyperbaric oxygen therapy is thought to help by enhancing neutrophil killing of pathogens, inhibiting fungal growth, and improving wound healing (4). A case series showed a high survival rate of 86% in patients treated with adjunct hyperbaric oxygen therapy (5). Further research is needed to determine the optimal treatment for this uncommon disease.

Discussion

The overall mortality rate of mucormycosis is greater than 40% with even higher rates reported in those with hematologic malignancy (3). Prompt initiation of treatment is crucial to increase the chances of survival. Surgical debridement is important to reduce the fungal burden and remove devitalized tissue. Amphotericin B is considered first-line antifungal therapy, with posaconazole or ivasconazole used for salvage therapy or long-term treatment (4). Hyperbaric oxygen therapy is thought to help by enhancing neutrophil killing of pathogens, inhibiting fungal growth, and improving wound healing (4). A case series showed a high survival rate of 86% in patients treated with adjunct hyperbaric oxygen therapy (5). Further research is needed to determine the optimal treatment for this uncommon disease.

Conclusion

Successful management of invasive rhinocerebral mucormycosis requires early diagnosis, surgical debridement, antifungal therapy, and control of underlying immunodeficiency. Even with adequate treatment, this rare infection carries a high mortality rate.

References


Abstract

Objectives: Present a case of recovery in a patient with invasive rhinocerebral mucormycosis. Radiologic findings, diagnosis and treatment are discussed.

Methods: The clinical course of a patient is described based on clinical interaction and chart review. We also provide a review of the literature on rhinocerebral mucormycosis.

Results: A 55 year old man presented with oral pain and left sided facial swelling. He was found to be in diabetic ketoacidosis. He developed diplopia with left gaze and an MRI was obtained which showed opacification of the left ethmoid and sphenoid sinuses as well as cavernous sinus thrombosis. Endoscopic sinus surgery was performed and histopathology showed invasive mucormycosis. The patient was placed on amphotericin B and experienced progression of his disease including left facial weakness. Serial MRIs demonstrated occlusion of left internal carotid and invasion of the skull base including the left intracranial trigeminal nerve. The patient’s condition worsened over the following weeks and he was transitioned to long-term ivasconazole. Several months later he began to experience improvement in his cranial nerve deficits. Endoscopic sinus exams have shown no recurrence of invasive mucormycosis 13 months out from diagnosis.

Conclusions: Treatment of rhinocerebral mucormycosis includes surgical intervention as well as antifungal medications and aggressive management of underlying immunocompromised state.

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Table of Figures

Figure 1. 3 day pre-operative CT shows opacification of the left sphenoid and ethmoid sinuses.

Figure 2. Biopsy shows anastomotic hyphae with right-angle branching, characteristic of mucormycosis (40X).

Figure 3. 3 day post-operative T2 flair MRI shows myositis involving the left nasopharyngeal and masticator spaces with invasion of the left medial pterygoid muscle.

Figure 4. 6 week post-operative T1 MRI shows occlusion of left internal carotid artery and enhancement of left temporal pole.

Figure 5. 3 week post-operative T1 MRI shows continued left internal carotid artery occlusion and enhancement of left trigeminal nerve.

Figure 6. 2 month post-operative T1 MRI shows reduction in size of area of necrosis of the left skull base.

Figure 7. 12 month post-operative T1 MRI shows reduction in size of necrotic area of the left skull base with persistent occlusion of left internal carotid artery.