Rhinocerebral Mucormycosis in a Child with Autoimmune Hepatitis: A Case Report

Jeffrey Bayer, MD and Hassan Ramadan, MD, MSc, FACS
West Virginia University Hospitals, Department of Otolaryngology- Head and Neck Surgery

ABSTRACT

The primary objective of this case report is to review the natural history of mucormycosis in children and discuss the management of this rare entity. The patient was a 12-year-old female with autoimmune hepatitis who was diagnosed with rhinocerebral mucormycosis and managed with surgical debridement, medical therapy, and correction of the underlying condition. The patient managed to survive the condition and is doing well over two years later. This case report highlights the importance of early diagnosis and emergent treatment of mucormycosis to prevent irreversible damage to vital organs.

REFERENCES


INTRODUCTION

Rhinocerebral mucormycosis is a rare but life-threatening fungal infection that affects the paranasal sinuses and orbits. This condition is most commonly seen in immunocompromised patients, such as those with diabetes, hematopoietic stem cell transplantation, or solid organ transplantation. The disease is characterized by rapid spread and destruction of soft tissue and bone, leading to significant morbidity and mortality if not treated promptly. The mainstay of treatment is surgical debridement, medical therapy, and correction of the underlying condition. This case report describes a 12-year-old female with autoimmune hepatitis and rhinocerebral mucormycosis who was managed with multimodal therapy and survived the condition.

CASE REPORT

A 12-year-old female was admitted to West Virginia University Children's Hospital with right-sided frontal headache, facial numbness, periorbital pain, and nasal congestion. On examination, she had right sided facial swelling and edema of the right maxillary and ethmoid sinus region. MRI demonstrated hypointense signal in the right nasal cavity and increased signal in the maxillary and ethmoid sinuses. CT demonstrated right sided nasal cavity, maxillary, and ethmoid sinus involvement. She was started on amphotericin B deoxycholate and underwent transorbital frontal craniectomy for intracerebral abscess evacuation.

Further treatment included: completion of medical therapy, surgical debridement, and correction of the underlying condition. Over 2 yrs later, she is alive without any signs of rhinocerebral mucormycosis.

DISCUSSION

This individual presentation is unique for several reasons:

1. The patient was a 12-year-old female with autoimmune hepatitis, a rare condition in pediatric population; a hematologic malignancy or diabetes mellitus.
2. She did not present with either neutropenia or fever.
3. Her immunosuppressed state and metabolic derangement resulted in a hyperglycemic ketoacidotic state, and uncontrolled diabetes (8, 9, 14-19).

CONCLUSIONS

Although rare, invasive fungal sinusitis should always be considered in any immunocompromised child presenting with acute sinusitis or orbital symptoms. Diabetes and hematologic malignancy are the most frequently cited predisposing factors in the pediatric population.

The most consistent findings for presentation are fever and neutropenia, while orbital and sinusosal symptoms are common as well. In the pediatric population, early diagnosis and emergent treatment may be the key to survival. Management may include multimodality approach with surgical debridement, medical therapy, and correction of the underlying condition.

An aggressive surgical approach was taken initially without pathologic or culture confirmation so that there would be no further delay in treatment. In light of her pathologic examination, the approach was switched to multimodality approach. This allowed for rapid orbital and intracranial exploration.

Although recent literature has described increased cases of mucormycosis in immunocompromised patients, there is limited data on the outcomes of treatment in this population. This case report highlights the importance of early diagnosis and emergent treatment of mucormycosis to prevent irreversible damage to vital organs.