International Journal of Research in Medical Sciences
Rohit et al. Int J Res Med Sci. 2024 Sep;12(9):3472-3475
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20242634

Case Report

Acute rhino-orbito-cerebral mucormycosis in a patient with uncontrolled type I diabetes mellitus: a case report

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Received: 27 June 2024
Revised: 10 August 2024
Accepted: 13 August 2024

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ABSTRACT

Rhino-orbito-cerebral mucormycosis (ROCM) is a severe and often fatal fungal infection that primarily affects immunocompromised individuals, especially those with diabetes mellitus. The fungi, primarily Rhizopus oryzae, exhibit angio-invasive properties leading to extensive tissue necrosis and vascular occlusion, contributing to high morbidity and mortality. Early diagnosis and prompt intervention are crucial for improving patient outcomes. We present the case of a 32-year-old female with uncontrolled type 1 diabetes mellitus who experienced progressive blackening of the skin on the right side of her face, weakness in both upper and lower limbs, swelling in the right orbital region, proptosis, and diminished vision in her right eye over 10 days. Diagnosis was made through physical examination and MRI findings. The patient underwent extensive debridement surgery and was treated with intravenous amphotericin B. MRI indicated complete mucosal thickening and air-fluid levels in the right maxillary sinus extending to the ethmoid, frontal, and sphenoid sinuses, and the "Black turbinate sign" was observed in the ipsilateral turbinates. Additional findings included right orbital cellulitis with endophthalmitis, optic neuritis, and an acute infarct in the right temporal lobe and ipsilateral lentiform nucleus. Successful surgical debridement and antifungal therapy followed the ROCM diagnosis. This case highlights the need for a high index of suspicion for ROCM in diabetic patients with facial and orbital symptoms, and emphasizes the necessity of early imaging and aggressive treatment to reduce the associated high risks.

Keywords: Rhino-orbito-cerebral mucormycosis, Rhizopus oryzae, Diabetes mellitus, Fungal infection, Angio-invasive, Tissue necrosis, Optic neuritis, Acute infarct, Intravenous amphotericin B, Debridement surgery

INTRODUCTION

Mucormycosis arises from fungi within the order Mucorales, with Rhizopus oryzae being the most prevalent organism identified in mucormycosis patients, responsible for about 70% of cases.[1] Rhino-orbito-cerebral mucormycosis (ROCM) is a severe fungal infection commonly affecting individuals with diabetes and compromised immune systems. The necrotizing and angio-invasive nature of these fungi enables them to spread from sinuses to the orbit and brain, leading to serious repercussions such as cavernous sinus thrombosis. ROCM is known for its significant residual morbidity and mortality due to the fungus's angio-invasive properties, resulting in vascular occlusion and extensive tissue necrosis. Early detection and prompt intervention are crucial for successful treatment.[2]

India boasts the world's second-largest population of individuals with diabetes, totaling 65.1 million, with almost 70% of cases classified as uncontrolled diabetes.[3] In cases of diabetes complicated by rhino-orbital-cerebral mucormycosis (ROCM), it constitutes a medical and

surgical emergency. A high level of suspicion is imperative for diagnosis, and initiating treatment promptly, even before the diagnosis is confirmed, is crucial for improving outcomes. Patients in such situations often necessitate extensive debridement surgery while receiving intravenous amphotericin.

CASE REPORT

A 32-year-old female presented with blackening of skin over the right side of her face, weakness in bilateral upper and lower limbs and swelling of the right orbital region, proptosis and diminution of vision in the right eye began almost 10 days back and progressed gradually. She was a known case of uncontrolled type 1 diabetes mellitus. On examination at presentation, the patient was conscious and afebrile; pulse and blood pressure were normal.

MRI of face, orbits and brain findings reveals complete mucosal thickening with collection and air-fluid level noted in right maxillary sinus extending into ipsilateral ethmoid, frontal and sphenoid sinus with "Black turbinate sign" of ipsilateral middle and inferior turbinate. FLAIR hyperintensity noted in right temporalis medial and lateral pterygoid muscles- inflammatory changes.

Deformed right globe with FLAIR hyperintense scleral signal and orbital content with coning of the posterior part of the right eyeball with fat stranding in orbital fat with bulky and hyperintense intraocular muscles and FLAIR hyperintensity in the right premaxillary region likely orbital cellulitis with endophthalmitis. Diffusion restriction in right optic nerve- optic neuritis. FLAIR hyperintensities showing true diffusion restriction in the right temporal lobe and ipsilateral lentiform nucleus with few tiny blooming foci within with mild perilesional edema-Acute infarct.

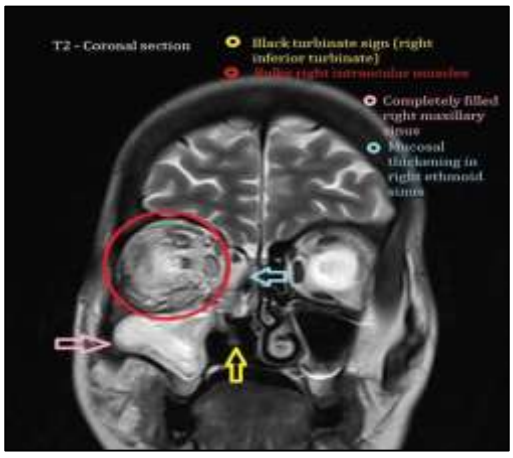

Figure 1: T2 coronal section showing black turbinate sign (yellow arrow). Bulky right intraocular muscles (red circle).

Filled right maxillary sinus (light pink arrow). Mucosal thickening in the right ethmoid sinus (blue arrow).

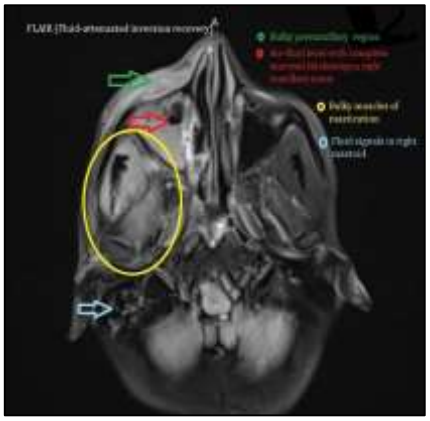

Figure 2: FLAIR axial section showing bulky muscles of mastication (yellow circle) and premaxillary region (green arrow).

Air-fluid level with complete mucosal thickening in the right maxillary sinus (red arrow). Fluid signal seen in right mastoid (blue arrow).

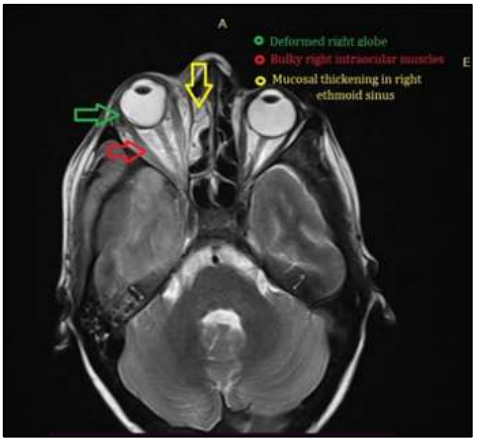

Figure 3: FLAIR axial section is showing deformed right globe (green arrow).

Bulky right intraocular muscles (red arrow). Mucosal thickening in right ethmoid sinus (yellow arrow).

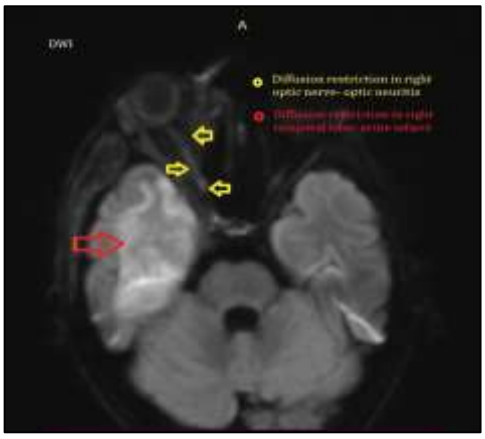

Figure 4: DWI axial section showing restriction in right optic nerve-optic neuritis (multiple small yellow arrows) and right temporal lobe-acute infarct (red arrow).

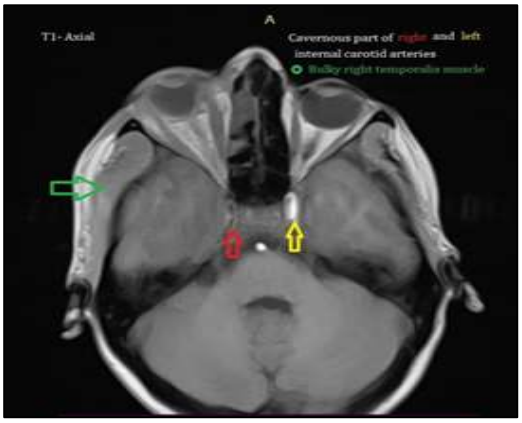

Figure 5: T1 Axial section showing cavernous part of right (red arrow) and left (yellow arrow) internal carotid arteries with a filling defect in right side-thrombus and bulky right temporalis muscle (green arrow).

DISCUSSION

These fungi are known to cause infections mainly in individuals with diabetes or deficiencies in phagocytic function (e.g., those linked with neutropenia or glucocorticoid therapy). Patients with elevated levels of free iron, which promotes fungal growth in the bloodstream and tissues, are also at a higher risk of developing mucormycosis.[3]

PCR techniques can aid in identifying the fungal species in histologic specimens, even if the culture results are negative. Additionally, MALDI-TOF mass spectrometry can be utilized for species identification in culture samples.[4-6] Severe complications may include cavernous sinus thrombosis, carotid artery blockage, CNS infarction stemming from mycotic intravascular thrombosis, presenting with symptoms like hemiparesis, hemiplegia, coma, and fatality.

CNS hemorrhage, abscesses, inflammation, and vision impairment have been extensively documented. Airway obstruction due to infections in the head and neck region is also a possibility. The antifungal medication IV amphotericin is linked with notable nephrotoxicity and electrolyte imbalances, particularly with the conventional amphotericin B deoxycholate formulation. Survivors often experience disfigurement post-surgical debridement, necessitating psychological or psychiatric assistance. Up to two-thirds of patients may face lingering consequences from the illness.

Differential diagnosis

Maintaining a high level of suspicion is crucial, as any delay in diagnosis followed by a delay in initiating IV amphotericin treatment can exacerbate morbidity and mortality.[7] The potential differential diagnosis can vary depending on the presenting symptoms and signs and may include the following.

Allergic fungal sinusitis

This condition, caused by an allergic response to dematiaceous fungi, can mimic sinusitis in immunocompetent individuals with symptoms such as proptosis and a rhinocerebral mass. It typically does not invade tissues and can persist for extended periods, often accompanied by underlying conditions such as asthma, allergies, mucosal polyps, and elevated IgE levels. In some cases, eosinophil-rich fungal mucin may erode into the adjacent orbit and intracranial space, leading to bony erosions due to the pressure exerted by the thick mucin.

Invasive aspergillosis

Rhinocerebral aspergillosis can present as a sinusitis-like condition involving the orbit and brain, particularly in immunocompromised patients, with an abysmal prognosis. Treatment typically involves IV amphotericin therapy combined with surgical debridement. Eye discomfort may result from inflamed eyelids, conjunctivitis, preseptal and orbital cellulitis, migraines, gingival or dental infections, or prolonged sinusitis. Orbital protrusion can also occur due to subperiosteal hematomas, inflammatory pseudotumor, cavernous sinus thrombosis, or orbital neoplasms

CONCLUSION

This case report highlights the critical importance of early recognition and intervention in rhino-orbito-cerebral mucormycosis (ROCM), particularly among patients with uncontrolled diabetes mellitus. The study emphasizes how imaging and rapid antifungal therapy, alongside surgical debridement, are essential in reducing the high morbidity and mortality associated with this aggressive fungal infection. By presenting a comprehensive analysis of clinical presentation and management strategies, this report adds to the understanding of ROCM, underscoring the need for heightened clinical awareness and prompt treatment initiation to improve patient outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Rohit, Khanduri S, Khan Z, Imam TA, Ashok Gupta A, Alam N, Mahmood N, et al. Acute rhino-orbito-cerebral mucormycosis in a patient with uncontrolled type I diabetes mellitus: a case report. Int J Res Med Sci 2024;12:3472-5.