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CASE REPORT

A Rare Battle: Multidisciplinary Care for a Child with Rhino-Orbital Cerebral Mucormycosis in Somali Region of Ethiopia

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Background: Rhino-orbital-cerebral mucormycosis (ROCM) is a rare, aggressive fungal infection with high mortality, primarily affecting immunocompromised individuals, especially those with uncontrolled diabetes mellitus, though it can occur in immunocompetent hosts. Cerebral involvement carries a near 100% fatality rate if untreated, and overall mortality remains high (>30%) even with therapy. We present a pediatric ROCM case from the resource-limited Somali region of Ethiopia.

Case Presentation: A 12-year-old female with type 1 diabetes mellitus presented with a 10-day history rapidly progressing from headache and fever to right-sided periorbital swelling, proptosis, vision loss, and a necrotic eschar. She had uncontrolled hyperglycemia and diabetic ketoacidosis (DKA) upon arrival. Imaging via CT and MRI revealed findings highly suggestive of ROCM, demonstrating right orbital involvement, cavernous sinus thrombosis, and further intracranial extension including cerebral abscesses/cerebritis and meningeal enhancement. Microbiological confirmation was unavailable due to resource limitations. Treatment included intravenous liposomal amphotericin B (requiring dose adjustment for transient nephrotoxicity), extensive endoscopic nasal debridement, right eye exenteration, and intensive glycemic control.

Discussion and Conclusion: This case highlights the diagnostic and management challenges of ROCM in resource-limited settings. Despite the high mortality associated with ROCM, particularly in children, the patient improved significantly and was discharged in good health, albeit with persistent visual impairment. Successful management involved prompt recognition, aggressive multimodal therapy (medical and surgical), and close multidisciplinary collaboration (Pediatrics, Ophthalmology, ENT). This outcome demonstrates the feasibility of successful treatment even in resource-constrained environments and underscores the critical importance of managing underlying conditions like diabetes to prevent opportunistic infections.

Keywords: mucormycosis, rhino-orbital-cerebral mucormycosis, pediatric, diabetic ketoacidosis, Ethiopia, resource-limited setting, fungal infection

Introduction

Mucormycosis is a serious fungal infection primarily affecting immunosuppressed individuals, although it can also occur in immunocompetent hosts. This infection is caused by fungi belonging to the subphylum Mucoromycotina and the order Mucorales, with eleven genera and 27 species documented as pathogenic.[1] Notably, genera such as Rhizopus, Mucor, and Lichtheimia account for approximately 75% of reported cases.[1] Key risk factors associated with mucormycosis include poorly controlled diabetes mellitus, hematological malignancies, severe or prolonged neutropenia, trauma, iron overload, and deferoxamine therapy, among others. Recent studies have highlighted prematurity as a significant risk factor for mucormycosis in infants under 12 months of age.[2] A comprehensive epidemiological study involving 63 cases across 15 countries revealed that hematological malignancies constituted the primary underlying risk factor, accounting for 46% of cases.[2] Conversely, a review of

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179 cases of rhino-orbital-cerebral mucormycosis (ROCM) indicated that nearly 70% of patients—126 individuals—had diabetes mellitus, with many presenting in diabetic ketoacidosis upon hospital admission.[3] Fungal spores are ubiquitous in nature. Infection typically occurs when these spores are inhaled by susceptible individuals, gaining an opportunity to cause disease primarily when host immunity is compromised. This often leads to local invasion of the paranasal sinuses, resulting in sinusitis. The infection can progressively disseminate to other organs, including the lungs, orbits, brain, and gastrointestinal tract. Alternative routes of transmission have also been documented.[1,2,4] Patients with ROCM typically present with symptoms akin to sinusitis, such as fever, headache, nasal congestion, and discharge. As the infection advances to the orbital region, patients may experience unilateral facial pain, periorbital cellulitis, eyelid swelling, proptosis, ophthalmoplegia, and potentially acute vision loss. A defining characteristic of this condition is the presence of a black eschar on necrotic lesions.[4] We present a typical case of ROCM, wherein the patient's symptoms progressively worsened, culminating in vision loss within ten days of illness onset.

Case Presentation

A 12-year-old female adolescent, diagnosed with type 1 DM for four years and receiving insulin therapy at a dose of 0.6 IU/kg/day, presented to the emergency department with right-sided peri-orbital swelling and right eyelid drooping of 10 days duration. According to her mother, the initial symptoms included severe headache, high-grade fever, and right retro-orbital pain for three days. On the fourth day, the patient exhibited worsening symptoms characterized by right peri-orbital swelling, right ocular protrusion, extreme ipsilateral facial pain, and loss of vision in the right eye. Subsequently, the swelling ruptured, leading to darkish pus discharge from the eye and nose (Figure 1). There was no history of facial trauma or animal bites, and the mother denied any abnormal body movements or weakness. Initially treated at a nearby hospital, the patient remained hospitalized for four days, receiving unspecified intravenous medications without improvement. Consequently, her family opted to self-refer her to our facility. Two days prior to her arrival, the patient experienced excessive thirst and urination, accompanied by altered mental status and uncontrolled blood glucose levels. Upon arrival at our pediatric emergency department, she appeared acutely ill and stuporous. Vital signs revealed tachycardia (PR: 134 bpm), Kussmaul breathing (RR: 33 bpm), a normal temperature (T: 36.9°C), and hyperglycemia (RBS: 559 mg/dL). Examination revealed ulcerated peri-orbital lesions with necrotic tissue affecting both the right eye and right nose (Figures 2 and 3).

Figure 1 This photograph was taken on day seven of the child's illness, three days prior to hospital admission. It illustrates significant right eye proptosis, extensive erythema, and the presence of necrotic lesions on the medial aspect of the right eye and adjacent nasal area.

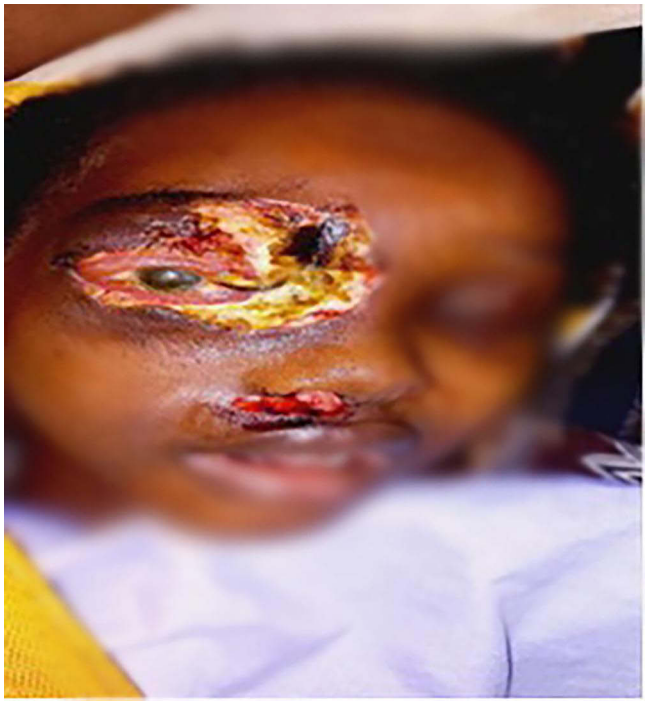

Figure 2 This image was captured on the day of hospital admission. It reveals a necrotic purulent ulcer accompanied by a black eschar, which has extensively covered the entire eye and peri-orbital cavity.

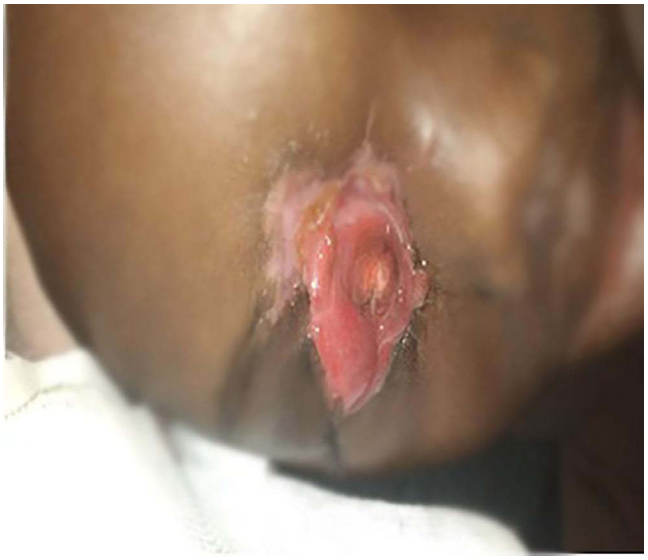

Figure 3 This photograph was taken during the first follow-up appointment, two weeks post-discharge. It provides a visual record of the patient's progress and healing status following the treatment received during hospitalization.

Clinical Findings

Neurological examination indicated a Glasgow Coma Scale (GCS) score of 12/15 (E: 3, V: 4, M: 5). The left pupil was mid-sized and reactive to light; the right pupil could not be adequately assessed due to extensive periorbital swelling, proptosis, and necrotic tissue. Plantar reflexes were bilateral flexor. Further neurological assessment was limited due to the patient's altered level of consciousness. However, examination of other systems including the cardiovascular, abdominal and musculoskeletal systems, were unremarkable. The patient was diagnosed with type 1 DM and moderate diabetic ketoacidosis (DKA) secondary to orbital cellulitis. Laboratory investigations included complete blood count

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(CBC), urine analysis, renal function tests (RFT), liver function tests (LFT), hemoglobin A1c (HbA1c), serum electrolytes and HIV test. CBC results indicated leukocytosis (WBC: 13,700/mm³) with neutrophil predominance (74%), normocytic normochromic anemia (Hgb: 8 g/dL), and a mean corpuscular volume (MCV) of 92.3 fL. Urine analysis demonstrated glucosuria (+2) and ketonuria (+2). The HbA1c level was elevated (10.4%). RFT showed normal creatinine and urea (Cr: 0.6 mg/dL, Urea: 22.8 mg/dL). The HIV test result was negative. Fluid management with 0.9% sodium chloride and regular insulin was initiated for DKA, alongside empirical antibiotics including vancomycin (45 mg/kg/day) and ceftriaxone (75 mg/kg/day). Consultations with ophthalmology and ENT were requested.

Therapeutic Intervention

The patient stabilized from DKA after 36 hours of management, with blood glucose levels dropping to 242 mg/dL and urine ketones becoming negative. A standard insulin regimen was resumed. Upon re-evaluation, the patient remained conscious, although proptosis of the right eye and ptosis of the eyelid persisted, along with cranial nerve palsies affecting CN III, IV, V, VI, and VII. Otherwise, the motor and sensory examinations were normal, with no signs of weakness. Imaging via orbital brain CT scan and MRI suggested rhino-orbital cerebral mucormycosis, revealing not only orbital disease and cavernous sinus thrombosis but also direct cerebral involvement including multifocal meningeal enhancement and intra-axial lesions in the right cerebral hemisphere consistent with abscess formation or cerebritis (Figures 4–7) Despite initial plans for microbiological evaluation of orbital tissue samples obtained during debridement, this could not be performed due to the unavailability of appropriate mycology laboratory facilities/reagents for fungal culture and microscopy at our center. Consequently, treatment with Liposomal Amphotericin B (5 mg/kg IV daily) was initiated based on strong clinical and radiological evidence.

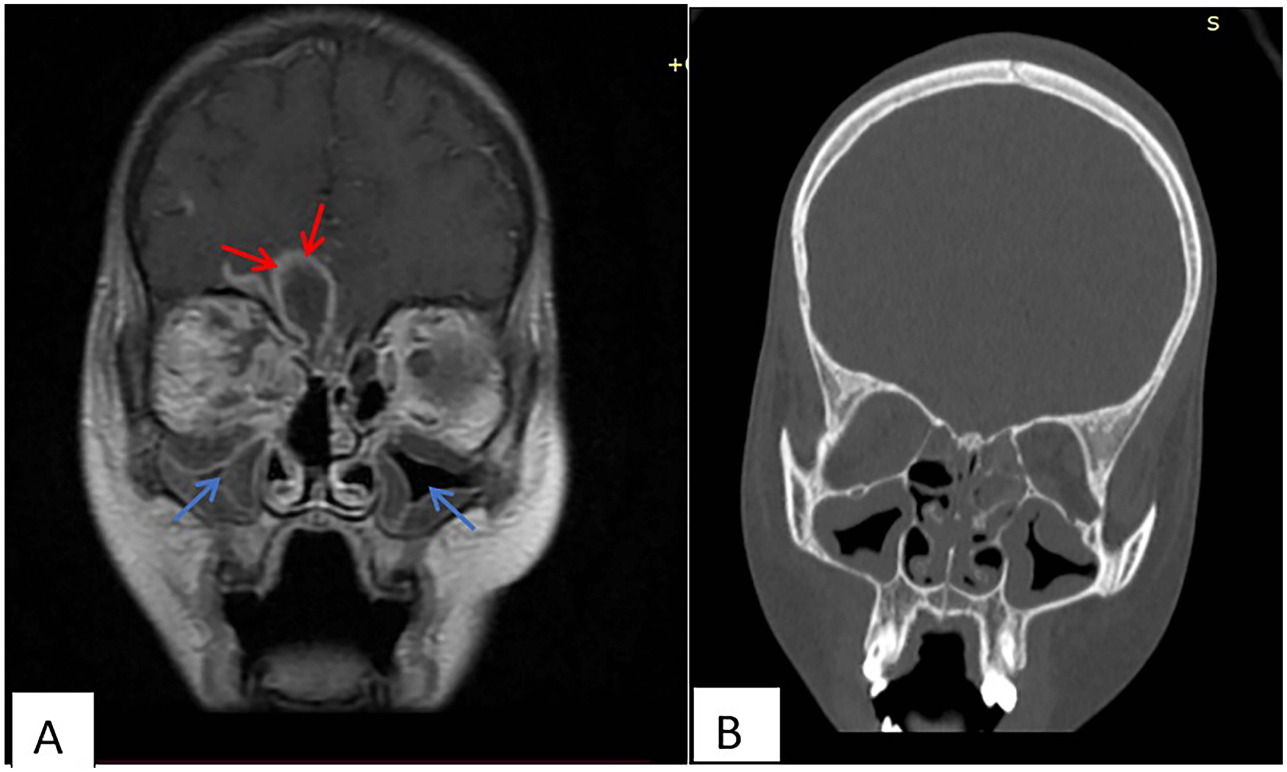

Figure 4 Imaging of Sinuses (**A**) Coronal post-contrast T1-weighted (T1W) MRI and (**B**) Coronal T2-weighted (T2W) MRI showing marked thickening and enhancement of the bilateral maxillary blue arrow in (**A**), ethmoid, and sphenoid, frontal sinus mucosa red arrows in (**A**) with meatal obstruction.

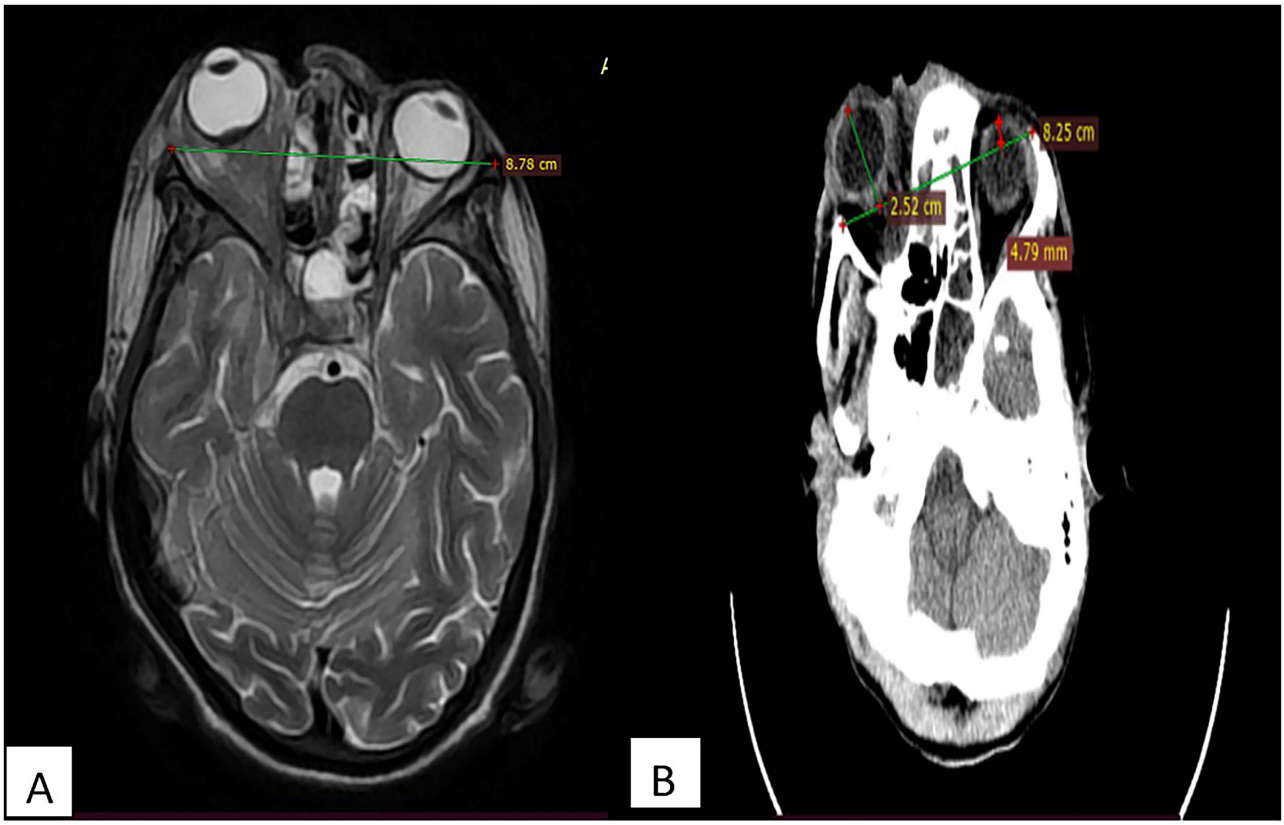

Figure 5 Axial Imaging of the Orbit. (**A**) Axial T2-weighted (T2W) MRI and (**B**) Axial CT soft tissue window image demonstrating proptosis of the right globe. The green line is The reference line for measurement of proptosis green line indicates the interzygomatic line, the upper limit of normal distance from this line to the anterior surface of the globe is 23 mm, above which indicates proptosis here it measures 25 mm.

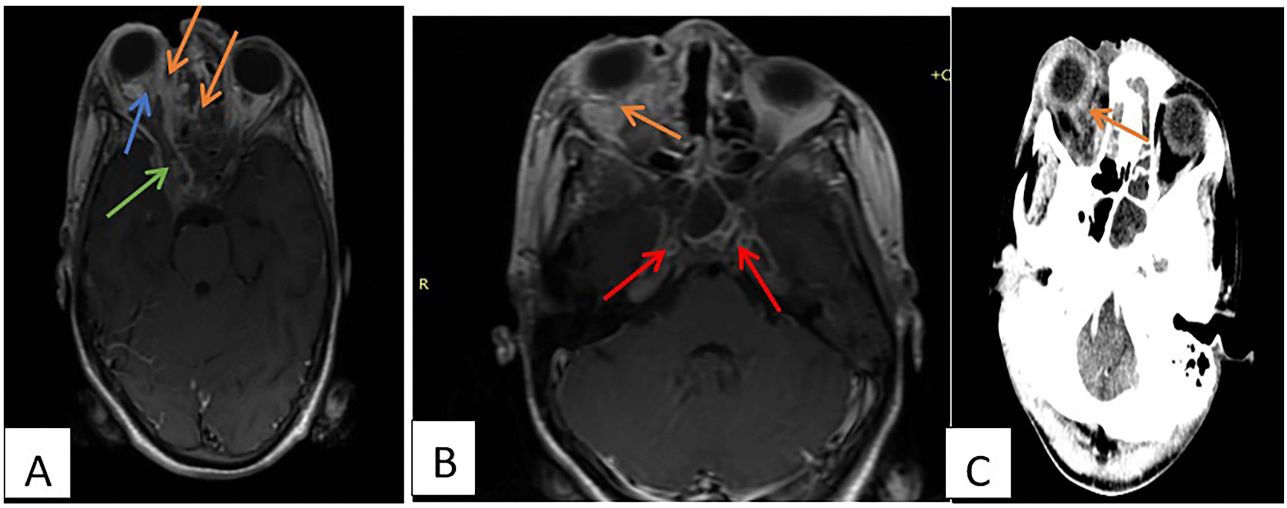

Figure 6 Axial Imaging Demonstrating Disease Extension. (**A**) Post-contrast axial T1-weighted (T1W) MRI, (**B**) Post-contrast axial CT, and (**C**) Axial CT bone window. These images show heterogeneously enhancing soft tissue and extensive fat stranding in the right retro-antral and right inferior periorbital regions orange arrows in (**A**–**C**). There is extension into the right pterygopalatine fossa green arrow in (**A**) and right orbital cavity blue arrows in (**A**) (intra- and extra-conal compartments), causing proptosis. Posterior extension involves the orbital apex, optic nerve, and optic chiasm. Extension into the right cavernous sinus red arrows in (**B**) with an intraluminal filling defect suggests thrombosis red arrows in (**B**) visible in (**A** and **B**).

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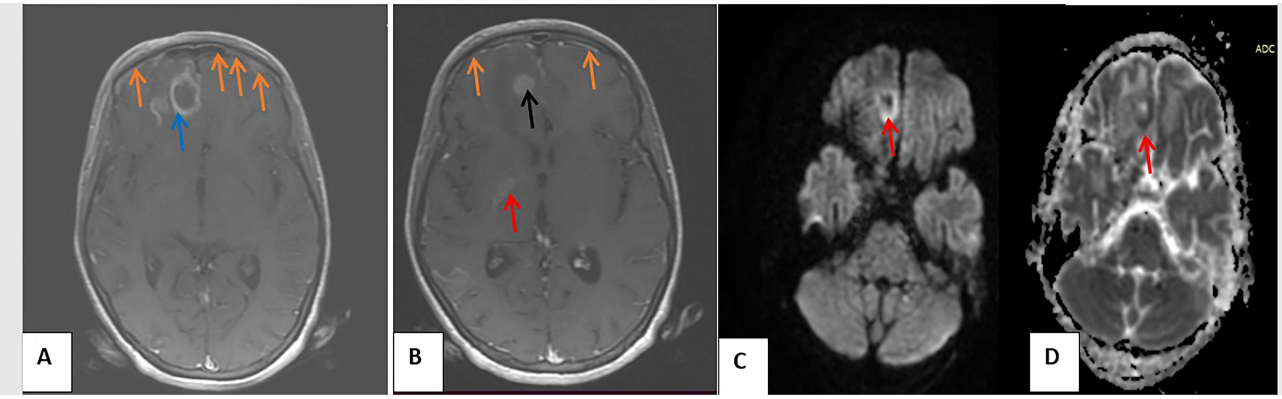

Figure 7 (**A** and **B**) post contrast axial, (**C**) DWI and (**D**) ADC brain MRI. Brain MRI Findings. (**A**) Post-contrast axial T1-weighted image showing multifocal patchy meningeal enhancement Orange arrows in (**A** and **B**), along with nodular (black arrow in (**B**) and ring-enhancing (blue arrows in (**A**) intra-axial lesions right frontal, temporal, parietal lobes, right basal ganglia red arrow in (**B**) with peripheral vasogenic edema. (**B**) Diffusion-weighted imaging (DWI) and (**C**) Apparent Diffusion Coefficient (ADC) map showing peripheral ring diffusion restriction in the right frontal lesion red arrow in (**C** and **D**), characteristic of abscess formation or cerebritis.

Clinical Course and Follow Up

After four days, the patient underwent extensive nasal endoscopic surgical debridement, with irrigations of amphotericin B. Post-operatively, she showed significant improvement, prompting the discontinuation of antibiotics after two weeks. However, nephrotoxicity from the amphotericin B necessitated a temporary cessation of the medication, which was reintroduced at a lower dosage upon stabilization of renal function, and it was continued for three weeks until a satisfactory clinical response was achieved. In the fourth week, the patient underwent right eye exenteration, after which she was discharged with a revised insulin regimen of 1 IU/kg/day. A follow-up appointment was scheduled, during which marked improvement was noted, including well-controlled blood sugar levels and no discharge from wound sites.

Discussion

The present case details the successful management of rhino-orbital-cerebral mucormycosis (ROCM) in a 12-year-old female with type 1 diabetes mellitus in a resource-limited setting. The significance of this case is multi-faceted: it highlights the rarity of pediatric ROCM survival, particularly in environments with constrained resources, and underscores the potential for positive outcomes driven by astute clinical judgment and aggressive, coordinated care even when definitive microbiological confirmation is unobtainable. This case is particularly significant given the rarity of ROCM, especially in pediatric populations, and the high mortality rates associated with this infection.[5] The patient's presentation with diabetic ketoacidosis (DKA) aligns with prior findings suggesting that uncontrolled diabetes, often complicated by DKA, is a major risk factor for mucormycosis in low- and middle-income countries.[5] This is in contrast to studies from developed nations, where hematological malignancies are more commonly reported as the primary predisposing factor.[2]

The initial symptoms of headache, fever, and retro-orbital pain, progressing rapidly to periorbital swelling, proptosis, vision loss, and finally, necrotic eschar formation, are consistent with the classic presentation of ROCM.[4] The progression to cranial nerve palsies, cavernous sinus thrombosis, and direct cerebral parenchymal involvement (including intra-axial lesions suggestive of abscess/cerebritis and meningeal enhancement as detailed in Figure 7), as revealed by imaging (see Figures 4–7), highlights the aggressive nature of the infection. Intracranial extension, particularly the development of cerebral abscesses or widespread cerebritis as observed in our patient, is associated with a grim prognosis in ROCM, with historical mortality rates approaching 80–100% even with treatment in some series, especially if surgical debridement of intracranial foci is incomplete or not feasible.[4,5] The survival of our patient despite such extensive cerebral involvement underscores the efficacy of the aggressive, early multimodal therapy implemented. The patient's clinical findings also emphasize that the presence of a black eschar is a hallmark sign of mucormycosis and should prompt urgent medical attention. While our initial diagnosis was made based on clinical presentation and imaging studies, the absence of microbiological confirmation, due to resource limitations, remains a significant drawback. However, this limitation itself underscores a critical aspect of managing ROCM in many parts of the world: clinicians must often rely on a high

index of suspicion based on characteristic clinical features and radiological evidence to initiate life-saving therapy without delay. This highlights the challenges faced in resource-constrained settings where rapid and accurate diagnostic tests are not always accessible.[1]

The management of our patient involved a multidisciplinary approach encompassing aggressive glycemic control, intravenous liposomal amphotericin B, and surgical debridement, including right eye exenteration. The initial escalation of amphotericin B dosage, followed by dose reduction due to nephrotoxicity, exemplifies the delicate balance clinicians must achieve in the management of mucormycosis. The use of endoscopic surgical debridement, performed by the otolaryngologist, was crucial in removing necrotic tissue and controlling the local spread of infection. This combined medical and surgical approach is consistent with the current recommendations for mucormycosis treatment.[6]

This case report contributes significantly to the limited body of literature on mucormycosis in Ethiopia and in pediatric populations globally. Notably, this represents the second reported case of ROCM from Ethiopia.[7] The previously published case by Alemayehu et al[7] described an adult male with COVID-19 and newly diagnosed, uncontrolled Diabetes Mellitus who developed ROCM and unfortunately did not survive, despite treatment that included amphotericin B and surgical debridement. Several factors may differentiate our case and its outcome. Firstly, our patient was pediatric, whereas the previous case was an adult. While both had diabetes, the acute inflammatory milieu of COVID-19 in the adult patient, coupled with newly diagnosed and likely severely uncontrolled diabetes, may have presented a more complex immunological challenge and potentially more rapid or aggressive disease progression. Secondly, while specific details of the extent of surgical debridement or intracranial involvement in the previous case were not exhaustively detailed for direct comparison, the aggressive and repeated debridement combined with early eye exenteration in our pediatric patient likely played a crucial role in source control. Our patient also had confirmed extensive cerebral involvement (abscesses/cerebritis), and her survival highlights that positive outcomes are achievable even in such advanced stages with comprehensive care. In contrast to the fatal outcome of the previous report, our patient recovered well and was ultimately discharged in good health. In contrast, our patient recovered well and was ultimately discharged in good health. This difference highlights the crucial role of comprehensive multidisciplinary care and its significant impact on survival in such complex cases, further distinguishing our findings from the previous local report and reinforcing the positive outcome despite the infection's high case fatality rate up to 62%,[8] and even higher with extensive cerebral disease. The primary importance of this case, therefore, lies in its illustration of successful ROCM management and patient survival in a pediatric patient within a severely resource-constrained environment. The successful management of this patient underscores the importance of early diagnosis, multidisciplinary care, and resourcefulness in the face of limited diagnostic capabilities. This report serves as an important demonstration that a high index of clinical suspicion, supported by characteristic imaging findings, coupled with aggressive multimodal therapy, can lead to survival in pediatric ROCM even when definitive mycological diagnosis is not feasible. Our report emphasizes the feasibility of achieving successful treatment outcomes, even in low-resource settings, when early intervention and diligent, coordinated care are provided.

Conclusion

This case report presents a rare and challenging case of ROCM in a pediatric patient from Ethiopia. The significance of this case report stems from documenting the survival of a pediatric patient with clinically and radiologically diagnosed ROCM in a region with limited diagnostic resources. It emphasizes the importance of early recognition, multidisciplinary management, and highlights the challenges and successes of care in a resource-limited setting. The experience also serves as a reminder of the crucial role of prompt management of DKA, and the necessity of accessible, high-quality, and timely diagnostic and treatment facilities, especially given that this case demonstrates that survival from ROCM is achievable even when ideal diagnostic pathways are unavailable. It underscores the need for more investment in strengthening health systems and improving healthcare accessibility to achieve universal health coverage, in line with the Sustainable Development Goals.

Data Sharing Statement

Data supporting the conclusions of this report are contained within the report. Additional non-relevant patient data are protected under patient privacy regulations and policies.

Ethical Approval

Written informed consent was obtained from the child's parent for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Ethical approval for the study and the publication of these case details was granted by the Jigjiga University, Institute of Health Science, Research and Health Research Ethics Review Board, Jigjiga (Reference number: IHRRC004/2025; approved on February 10th, 2025). The study was conducted in accordance with the Declaration of Helsinki.

Author Contributions

Each author contributed significantly to the work reported, whether it was through conception, study design, execution, data acquisition, analysis, and interpretation, or in all of these areas; they all contributed to draft, revise, or critically review the article; they approved the final version that was published; they all agreed on the journal to which the article was submitted; and they all agreed to take responsibility for the work in its entirety.

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Disclosure

The authors report no declarations of interest in this work.

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