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Case Series Of Exenteration In Postcovid Rhino-Orbito- Cerebral Mucormycosis In A Tertiary Care Centre

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Abstract

Rhino-orbito-cerebral mucormycosis (ROCM) is a dreaded opportunistic infection seen in immunocompromised individuals. COVID 19 infection causes significant immunosuppression in otherwise normal individuals. This, along with concomitant steroid use & pre-existing diabetes mellitus provide a fertile ground for mucormycosis, leading in a few patients to exenteration of the eye. We studied the demographic details of postcovid ROCM patients who underwent exenteration & also its impact on survival of rhino-orbito-cerebral mucormycosis patients.

Keywords: COVID 19, Rhino-orbito-cerebral mucormycosis, Diabetes Mellitus, Steroid use, Exenteration.

Introduction

Rhino-orbito-cerebral mucormycosis is a dreaded opportunistic infection caused by the Mucorales group of fungi in immunosuppressed individuals[1] which invades the vascular endothelium[2] causing endarteritis obliterans and angioinvasion, inducing a prothrombotic state and resulting in ischaemia and necrosis of tissues [3,4] It begins in the nose and progresses rapidly through the paranasal sinuses, invading the orbit and eventually the intracranial structures *via* the orbital apex or through blood vessels.[5–7] The classical hallmark is necrotic black eschar or crust in the nasal cavity, nasal dorsum, palate or face. The risk factors for rhino-orbital-cerebral mucormycosis include: uncontrolled diabetes mellitus, corticosteroid therapy, immunosuppressant drugs, haematological malignancies and organ transplantation.[8] Expanding lesions of the orbital apex result in a predictable neurological deficit known as orbital apex syndrome (OAS). OAS involves damage to the oculomotor nerve (III), trochlear nerve (IV), abducens nerve (VI), and ophthalmic branch of the trigeminal nerve (V1) in association with optic nerve (II) dysfunction.[9] Visual loss from optic neuropathy, ptosis, ophthalmoplegia, loss of corneal sensitivity, and facial numbness are the hallmarks of an OAS. Coronavirus disease 2019 (Covid-19) infection and its management have caused significant immunosuppression in otherwise immunocompetent individuals, inviting opportunistic infections. Diabetes mellitus type II is associated with a two-fold increase in mortality as well as severity of Covid-19 relative to non-diabetic patients. Covid-19 patients admitted to the intensive care unit, on mechanical ventilation, and staying in hospital for longer than 50 days, are more prone to fungal co-infections.[10] The majority of patients develop symptoms of rhino-orbital-cerebral mucormycosis after recovering from Covid-19, which indicates the importance of follow up in these high-risk Covid-19 patient subset.

Amphotericin B is the first-line drug for systemic therapy. [11] Cure with medical therapy alone is exceptional and should not be relied on as an alternative to adequate combination therapy. [12] Concomitant surgical debridement remains a key feature of management because of the difficulty in eradicating mucor with amphotericin B alone from the necrotic tissues. Until recently, the standard therapy for rhino-orbital mucormycosis has been IV amphotericin B, extensive sinus debridement, and orbital exenteration to prevent intracranial extension of this infection.

The most critical decision in the management of rhino-orbital mucormycosis is whether the orbit should be exenterated. The literature fails to provide a broad base of information of how clinicians determine the need for exenteration in daily practice. The decision for exenteration often depends on the judgment of the treating otolaryngologist and the ophthalmologist. Hence this study was chosen.

Aims & Objectives:

1. To study demographic details of postcovid rhino-orbito-cerebral mucormycosis patients who underwent exenteration.
2. To study impact of exenteration on survival of rhino-orbito-cerebral mucormycosis patients.

Materials & Methods:

It was a retrospective study. It was conducted from May 2021 to September 2021. Out of 192 cases of mucormycosis, orbital exenteration was performed in 10 cases.

A detailed history of patient diagnosed with covid was taken. History of diabetes and hypertension with its duration and other associated co-morbidities was taken. Visual acuity , anterior and posterior segment examination were done. RT PCR , labrotary investigations and contrast MRI of brain and orbit was done.

A multidisciplinary approach consisting of opinion of ophthalmologist, otolaryngologist, radiologist and neurologist was followed regarding decision of exenteration.

Current indications for exenteration in ROCM includes

1. Stage 3c or worse as proposed in staging by Honavar et al
2. Non salvageable globe
3. Significant proptosis with fulminant orbital involvement and rapidly progressive disease
4. Necrosis and associated thrombosing vasculitis
5. Unilateral cases and failure to respond to conservative therapy (worsening or no improvement within 72 hours)

Inclusion & Exclusion Criteria:

Inclusion Criteria:

1. Clinically diagnosed patients with Rhino-orbito-cerebral mucormycosis.
2. Patients with no PL or with involvement of cavernous sinus.

Exclusion Criteria:

1. Patients who refused to give consent.
2. Patients who aborted the treatment.

Observations & Results:

- Out of 10 patients 9 were males and 1 was female.

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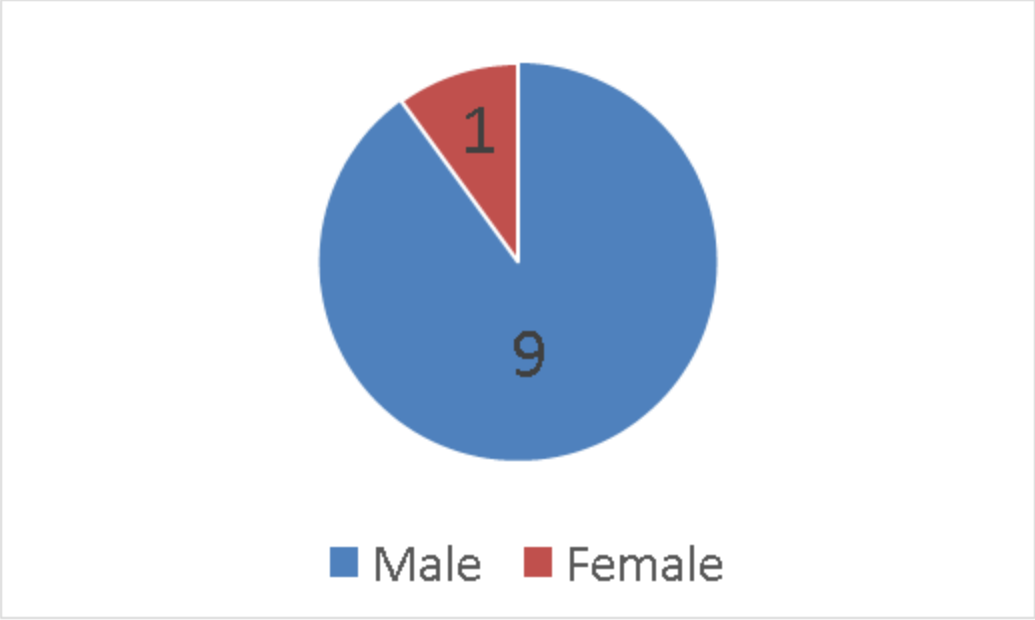
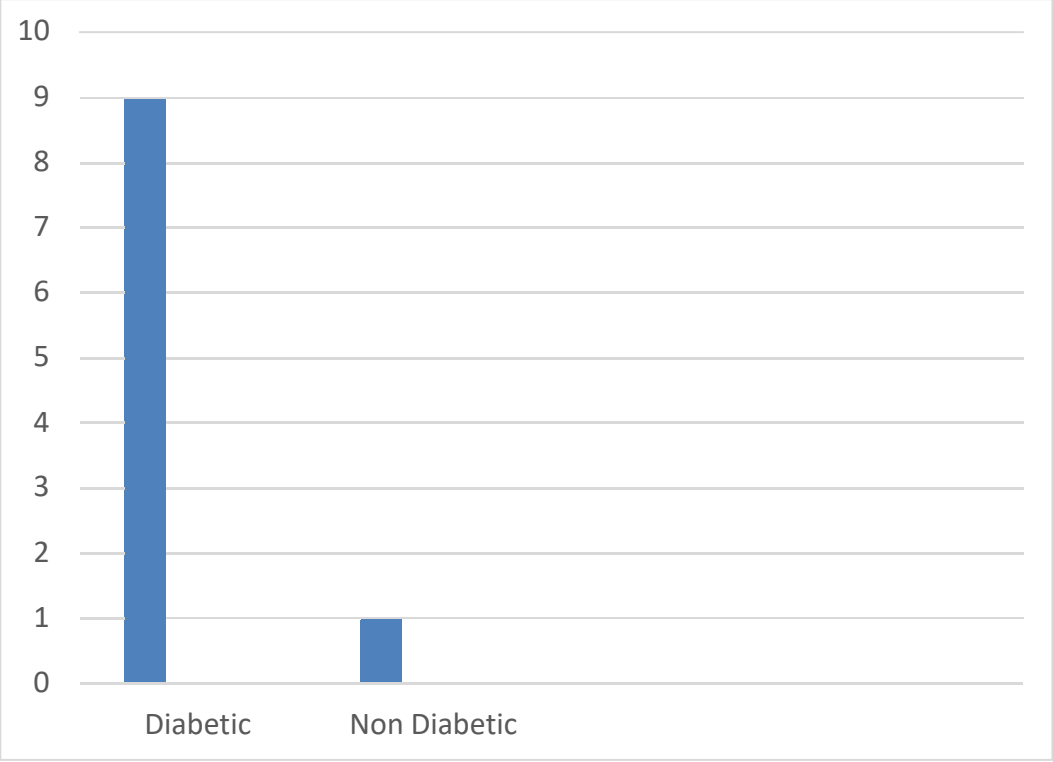


Mean age was 51.2 years (range of 32- 70 years)

9 patients were diabetic.



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In 8 patients RT - PCR was positive and in 2 patients diagnosis was confirmed by nasal endoscopy.

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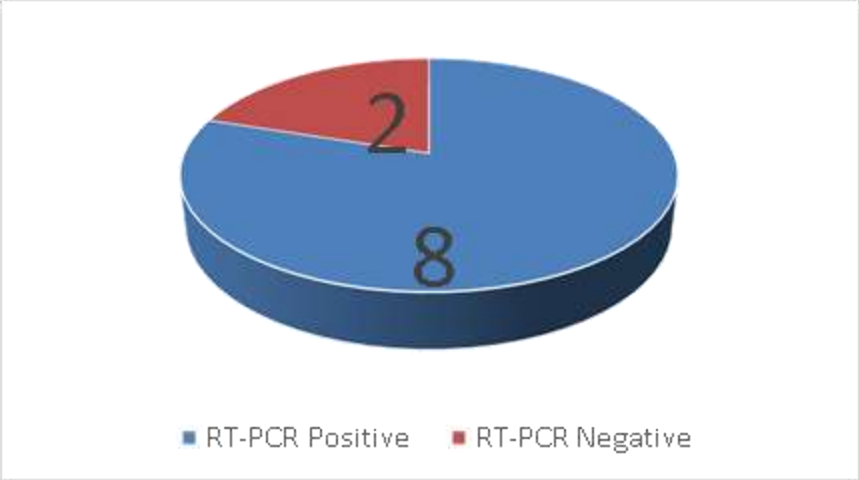
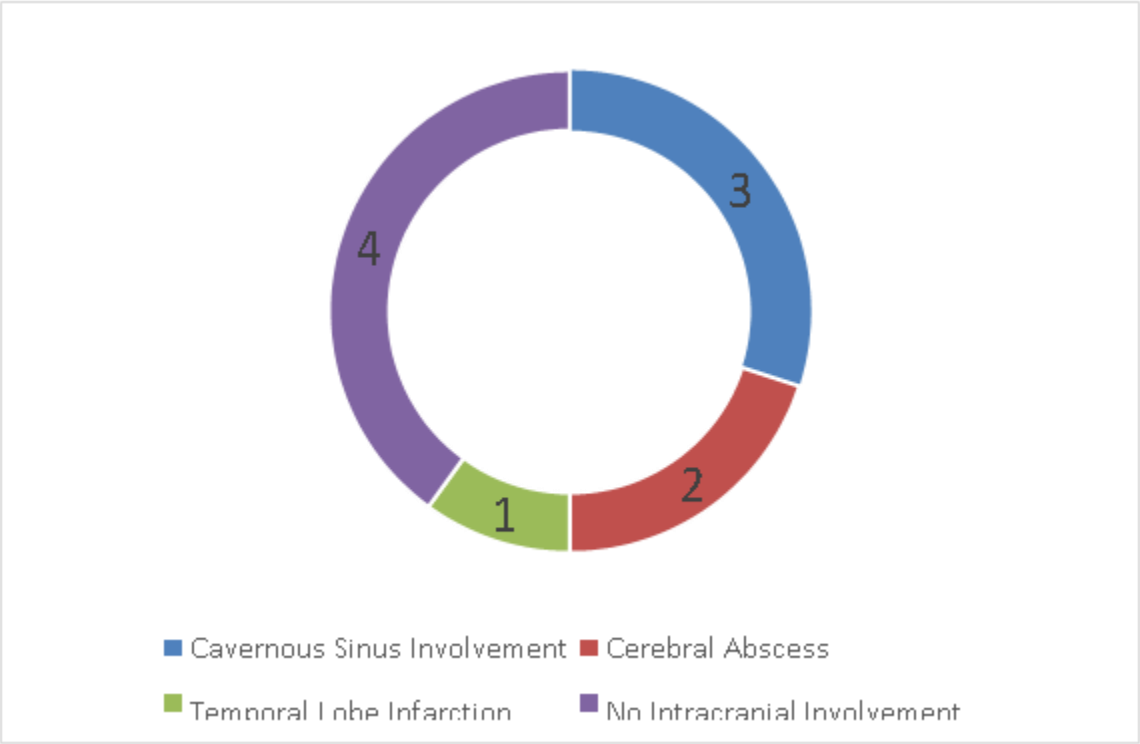


1. O2 and IV steroids were received by 8 patients.
2. In all patients vision in exenterated eye was no PL.
3. Proptosis, partial or total ophthalmoplegia and ptosis were presenting symptoms in exenterated eye.
4. Out of 10 patients 9 patients had unilateral ocular findings. MRI contrast of brain and eye documented intra and extraconal involvement along with orbital apex involvement.
5. Cavernous sinus involvement and cerebral abscess were noted in 3 and 2 patients respectively.
6. Infarction of temporal lobe was noted in one case. Four patients did not have intracranial involvement.



Death occurred in 3 patients.

Discussion:

Piromchai et al[8] observed that more than 70% of the mortalities occurred in the patients who exhibited symptoms of the disease within 14 days before admission. Hence, ROCM management requires an urgent, individualized multidisciplinary, approach including combination of appropriate systemic antifungal treatment, surgical debridement of necrotic

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tissues and correction of underlying conditions like hyperglycemia etc. COVID-19 infection, its treatment, resultant immunosuppression, and associated comorbidities had made patients vulnerable to opportunistic infections including mucormycosis[9] and rapid progression due to worsening of the disease process requiring exenteration. 9 patients were diabetic, 8 patients were positive for COVID 19 and 8 patients received IV steroids suggesting the role of steroid administration in contributing to the disease etiology due to immunosuppression.

Several guidelines have been proposed by different scientific bodies to aid in decision-making for exenteration. S G Honavar[10] proposed a management algorithm for ROCM in the setting of COVID-19, wherein orbital exenteration is recommended for extensive orbital involvement (central retinal artery or ophthalmic artery occlusion, superior ophthalmic vein thrombosis, orbital apex involvement, loss of vision and bilateral involvement), limited CNS involvement (focal or diffuse cavernous sinus involvement or thrombosis) and in cases of extensive CNS involvement (involvement beyond cavernous sinus, skull base involvement, brain infarction or diffuse CNS involvement) if general systemic condition of patient permits surgery. In our study all patients had extensive orbital involvement with 6 patients having varying CNS involvement & were eligible for exenteration.

There are multiple ways of approaching the procedure which include lid sparing exenteration, extended exenteration and endoscopic orbital exenteration. In our study, lid sparing exenteration was preferred due to its better cosmetic outcomes with rapid healing.[11] Post- operative course of all 7 patients who survived in our study was uneventful, while 3 patients succumbed in the postoperative period.

Conclusion:

Mucormycosis involves the orbit and other ocular structures. Ophthalmologists are one the first to see the patients with this morbid condition. Delay in establishing diagnosis and initiating early treatment could prove fatal. Aggressive medical and surgical management is most essential and requires multi-disciplinary approach. Management should be coordinated and planned among the various multidisciplinary services and in-depth discussion with patient and family regarding prognosis are essential to have a better outcome & overall good patient care.

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