The Clinical Pattern of Orbital Mucormycosis in a Tertiary Eye Care Hospital

M. V. S. Prakash, P. Ashok Kumar, T. G. Umamaheswari, V. Harivanzan
Department of Orbit and Oculoplasty, Regional Institute of Ophthalmology and Government Ophthalmic Institute, Chennai, Tamil Nadu, India

Abstract

This study aimed to report the clinical profile of patients with rhino-orbital mucormycosis at a tertiary eye care center in Tamil Nadu. This was a single-center, retrospective case series of patients who were diagnosed as rhino-orbital mucormycosis between January and December 2018. The study included nine patients of rhino-orbital mucormycosis with a mean age of 53.33 ± 8.3 years (range: 35–61 years), comprising five males and four females. All the cases were unilateral in presentation. Uncontrolled diabetes mellitus was the most common underlying predisposing factor. Four patients presented with proptosis, two patients complained of headache, and five patients had defective vision. Computed tomography scan showed pansinusitis in six patients. Total ophthalmoplegia was noted in four patients. All patients received parenteral therapy with amphotericin B. Rhino-orbital mucormycosis is a rare, life-threatening, and insidious fungal infection. Uncontrolled diabetes mellitus and immunosuppression are the most important risk factors. The orbital findings may range from orbital pain to ophthalmoplegia and blindness. We present this case series to emphasize the importance of early diagnosis and treatment of rhino-orbital mucormycosis.

Keywords: Diabetes mellitus, orbital, pansinusitis, rhino-orbital mucormycosis

Introduction

Rhino-orbital mucormycosis is an aggressive opportunistic infection that occurs in debilitated patients, particularly individuals with uncontrolled diabetes mellitus associated with ketoacidosis, immunocompromised, and renal disease. Healthy persons are rarely affected due to normal containment by phagocytes. The orbit is a site for mucormycosis in about 10% of patients with hematological malignancies.[1] The sinus and nasopharynx are inoculated by fungal spores (which are ubiquitous in soil, air, skin, body orifices, manure, and food), which grow and spread into the tissue by means of hyphae. This organism has a propensity to invade and occlude vascular lumina, leading to infarction which forms a characteristic black eschar. Spread to intracranial cavity may be rapid and devastating, leading to a fatal outcome. Early diagnosis allows for containment and successful therapy. The earliest orbital symptom consists of apical boring pain, and progression is associated with increased cellulitis, proptosis, and defective vision. There may be increased density of soft tissue and enlargement of optic nerve.

Materials and Methods

This was a hospital-based, retrospective case series which analyzed patients with rhino-orbital mucormycosis, referred from the ENT Department, Rajiv Gandhi Government Hospital, Chennai, to our institute from January 2018 to December 2018. All patients with a clinical diagnosis of rhino-orbital mucormycosis were included in this study. After taking a detailed ocular and systemic history, each patient underwent a complete eye examination including assessment of best-corrected visual acuity, slit-lamp biomicroscopy, and fundus examination with +90 D and indirect ophthalmoscopy. Laboratory investigations performed were complete blood count, fasting blood sugar, urine sugar, ketone bodies, blood urea, and serum creatinine. Diagnosis was made based on clinical presentation and supported with radiological and laboratory investigations.

Address for correspondence: Dr. T. G. Umamaheswari, Regional Institute of Ophthalmology and Government Ophthalmic Institute, No. 132, Rukmani Lakshmipathy Salai, Egmore, Chennai - 600 008, Tamil Nadu, India.
E-mail: umaa_01@yahoo.com

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RESULTS

Table 1 shows case series of nine cases of rhino-orbital mucormycosis were referred to our institute from January 2018 to December 2018. The mean age at presentation was 53.33 ± 8.3 years (range 35–61 years). Five patients were male (55%) and four were female (45%). All the cases were unilateral in presentation. Uncontrolled diabetes mellitus was the most common underlying predisposing factor as shown in Table 1. Four patients presented with proptosis, two patients complained of headache, and five patients had defective vision. Computed tomography scan showed pansinusitis in six patients. Total ophthalmoplegia was noted in four patients. All patients received parenteral therapy with amphotericin B.

All the patients under our study had a chronic history of diabetes mellitus, except one patient who was newly diagnosed. Chronic renal failure was noted in one patient; diabetes was associated with hypertension in two patients. Five patients presented with diabetic ketoacidosis. All patients had undergone functional endoscopic sinus surgery. Pansinusitis was noted in eight patients. Three patients had a magnetic resonance imaging finding of optic nerve involvement and were prescribed tablet prednisolone. Injection amphotericin was given for all patients, except for one patient in whom it was changed to posaconazole due to associated renal failure. Optic nerve sheath decompression was done for one patient. Clinical improvement in terms of visual acuity was observed in one patient, four patients had a decline in vision to less than 6/60, vision remained the same in one patient, and three patients had total loss of vision. One patient’s condition deteriorated requiring orbital exenteration.

DISCUSSION

Rhino-orbital mucormycosis is an acute and often lethal opportunistic fungal infection, typically affecting patients with uncontrolled diabetes mellitus or immunosuppression caused by fungi of the class Zygomycetes. Other factors predisposing to invasive mucormycosis include hematological malignancies, severe burns, chronic kidney disease, malignancies, severe burns, chronic kidney disease, and pregnancy.

Table 1: Clinical Profile, treatment and outcomes in orbital mucormycosis

| Age/sex | Predisposing factors | BCVA | Clinical presentation | Diagnosis | Treatment | Outcome |
|---------|----------------------|------|-----------------------|-----------|-----------|---------|
| 55/male | Diabetes*6 years/ hypertension/CVA | 6/18p | 6/18p | RE proptosis, chemosis, restriction of movements | CT PNS - right acute ethmoid sinusitis | Injection amphotericin | Visual acuity declined |
| 65/male | Diabetes*8 years Chronic renal failure | 6/12p | HM+, PL+, PR intact | Headache, RE blurring of vision | CT PNS-right-sided pansinusitis | Injection amphotericin | Visual acuity declined |
| 35/female | Diabetes mellitus*4 years | 6/18 | 6/9 | LE proptosis | CT PNS-left-sided pansinusitis | Injection amphotericin | Visual acuity declined |
| 51/female | Diabetes mellitus*6 years | 6/36p | 6/24p | LE proptosis | CT PNS-left-sided pansinusitis | Injection amphotericin | Visual acuity remained the same |
| 52/Male | Diabetes mellitus*5 years | 6/18 | 6/36 | LE defective vision | MRI brain optic nerve involvement | Injection amphotericin + tablet prednisolone | Visual acuity declined |
| 55/female | Newly diagnosed diabetes mellitus | 6/36 | CFCF PL + PR intact | LE axial proptosis Multiple cranial nerve palsies | CT PNS, B/L pansinusitis | Injection amphotericin | Patient deteriorated |
| 55/male | Diabetes mellitus*3 years | 6/18 | 6/9 | Headache, RE blurring of vision | CT PNS-Right-sided pansinusitis | Injection amphotericin | Improvement in visual acuity noted |
| 61/female | Diabetes mellitus*10 years Hypertension | CFCF | No PL | LE total loss of vision Total ophthalmoplegia | MRI brain optic nerve involvement | Injection amphotericin | No improvement was noted |
| 55/male | Diabetes | 6/6 | No PL | LE total loss of vision Drooping of left upper eyelid | MRI brain optic nerve thickening | Injection amphotericin | No improvement was noted |

*History of Diabetes Mellitus in Years. BCVA: Best-corrected visual acuity, RE: Right eye, LE: Left eye, MRI: Magnetic resonance imaging, CT PNS: Computerized tomography of the paranasal sinuses, B/L: Bilateral, RAPD: Relative afferent pupil defect, FESS: Functional endoscopic sinus surgery, PL: Perception of light, PR: Projection of rays, HM: Hand movements, CFCF: Counting fingers close to face.
acquired immunodeficiency syndrome, immunosuppressant use following solid organ transplant.\textsuperscript{(2)} It is characterized by sinusitis and a painless necrotic black palatal or nasal septum eschar.\textsuperscript{(3)} Clinically, it can present as fever, sinusitis, headache, periorbital or facial swelling, ptosis, visual loss, and ophthalmoplegia.

In all cases, sinus and palatal disease debridement should be done. This would help in reducing the load of infection in the sinuses and also provide tissue for histopathological diagnosis. Exenteration should be done in advanced involvement of the orbit. None of our cases showed invasion to the vessels of the brain. The diagnosis was confirmed histopathologically by tissue invasion and subsequent tissue reaction to fungi rather than just the presence of organism.\textsuperscript{(4)} Ulceration of the hard palate is interpreted as a sign of disease extension from the sinus. The infection has a high incidence in diabetic patients due to the greater availability of glucose to the pathogen, lower response of the T-cells, reduced serum inhibitory activity against the \textit{Rhizopus} in lower pH, and increased expression of some host receptors that mediate the invasion of human epithelial cells by microorganisms.\textsuperscript{(5)}

The management includes reversal of underlying predisposing conditions, prompt initiation of antifungal therapy, and early and aggressive surgical treatment. A rapid correction of the underlying metabolic derangement is the most important criterion for controlling the disease. Amphotericin B must be promptly initiated to bring the disease under control.

Despite advances in diagnosis and treatment, a high mortality rate of 30\%–70\% exists for the disease. Mucormycosis may be the first manifestation in patients with undiagnosed diabetes mellitus. Diabetes must be promptly controlled with soluble insulin, and the patient must be put on broad spectrum antibiotics.\textsuperscript{(6)} In extreme cases, aggressive surgical debridement must be done to reduce mortality.

**CONCLUSION**

In patients with mucormycosis, early detection, suitable antifungal therapy, appropriate surgical debridement, and control of risk factors such as diabetes mellitus should be implemented. Patients often have a history of immunosuppression and the onset is insidious. There must be a high risk of suspicion for mucormycosis to avoid delay in treatment.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Rootman J, editor. Inflammatory diseases. In: Diseases of the Orbit. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2003.
2. Petrikkos G, Skiada A, Lortholary O, Roilides E, Walsh TJ, Kontoyiannis DP. Epidemiology and clinical manifestations of mucormycosis. Clin Infect Dis 2012;54 Suppl 1:S23-34.
3. Hadzri MH, Azarisman SM, Fauzi AR, Kahairi A. Invasive rhinocerebral mucormycosis with orbital extension in poorly-controlled diabetes mellitus. Singapore Med J 2009;50:e107-9.
4. Parfrey NA. Improved diagnosis and prognosis of mucormycosis. A clinicopathologic study of 33 cases. Medicine (Baltimore) 1986;65:113-23.
5. Mohammadi R, Meidani M, Mostafavizadeh K, Jari B, Hamedani P, Sayedain SM, et al. Case series of rhinocerebral mucormycosis occurring in diabetic patients. Caspian J Intern Med 2015;6:243-6.
6. Oladeji S, Amusa Y, Olabanji J, Adisa A. Rhinocerebral mucormycosis in a diabetic case report. J West Afr Coll Surg 2013;3:93-102.