Section: ENT

Original Research Article

AN EVOLVING TREND OF ISOLATED POSTERIOR GROUP OF PARANASAL SINUSES INVOLVEMENT IN RHINOORBITAL MUCORMYCOSIS

Maranganti Deepthi[1], Meesala Tejaswi[2], Neelap Uma[3], Jatoh Bhupender Singh Rathod[4]

[1]*Assistant Professor, Department of ENT, Gandhi Medical College and Hospital, Secunderabad, India.*
[2]*Assistant Professor, Department of ENT, Gandhi Medical College and Hospital, Secunderabad, India.*
[3]*Assistant Professor, Department of ENT, Maheshwara Medical College and Hospital Sangareddy, India.*
[4]*Professor, Department of ENT, Gandhi Medical College and Hospital, Secunderabad, India.*

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Corresponding Author:
Dr. Neelap Uma,
Assistant Professor, Department of ENT, Maheshwara Medical College and Hospital Sangareddy, India.
Email: uma.neelap@gmail.com

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ABSTRACT

Background: Rhino-orbito- cerebral Mucormycosis is an invasive fungal infection occurring in immunocompromised patients. Isolated involvement of posterior ethmoid and sphenoid sinus with almost normal looking anterior ethmoids is rare. High index of suspicion is required to identify such patients. Patients present with vague eye complaints like blurring of vision, diplopia, vague headache, retro orbital pain with no nasal complaints. If not diagnosed early it becomes rapidly fatal causing loss of vision and intra cranial involvement. This change in trend of presentation and orbital apex syndrome poses challenges to ENT surgeons. Prompt interventionand aggressive treatment are required to reduce morbidity and mortality associated with the disease. **Objectives:** The study is aimed to identify the presentation of rhino-orbital Mucor mycosis as isolated involvement of posterior ethmoid and sphenoid sinus.To bring attention towards this changing trend and highlightthe challenges associated with diagnosis of orbital apex syndrome andanalyse various treatment methods used and post-operative sequela

Materials and Methods: We studied207patients rhino orbitalmucormycosis admitted in a tertiary care hospital in Telangana during period of 12 months fromdecember 2021 to november2022. A detailed case history including predisposing factors and comorbidities were taken.Diagnostic nasal endoscopy, CECT of nose and paranasal sinuses and MRI orbit scan of orbits and brain was done. Treatment started with injectionliposomal Amphotericin B on admission and surgical debridement of involved structures in nose, paranasal sinuses and orbit was done and tissue sent for KOH mount, fungal culture and histopathology for confirmation and followed up for 18 months.

Results: Total 207patients of mucor mycosis with male predilection (2:1). Most common age group is 40-60 years. Patients are being categorised into 4 groups. Group I include 16 patients and treated with liposomal amphotericin B only. Group II includes 74 patients and treated with amphotericin B and FESS.Group III includes 63 patients and treated with intravenous and retrobulbar injection of amphotericin B and underwent FESS. GROUP IV includes 54 patients who underwent FESS and orbital decompression.

Conclusion: RCOM is a fatal infection requiring multidisciplinary management. There is a change in presentation of mucor from frank nasal and palatal symptoms to presentingwith eye complaintspredominantly with relative less nasal involvement. Such cases should be investigated thoroughly on keen suspicion , and prompt medical and surgical management should be started early to reduce morbidity and mortality associated with disease.

Keywords: Orbital apex syndrome, Rhino orbital Mucormycosis,Liposomal amphotericin B, Diabetes mellitus, Ocular manifestations.

INTRODUCTION

Mucormycosis is a fungal infection belongs to the order Mucoraceae,[1] causesvascular invasion and host tissue necrosis by fungal hyphae which presents as characteristic black eschar over nasal turbinates or hard palate(1).Paranasal sinus mucosa when invaded by fungus may directly involve orbital apex and spread intracranially.[1] Mucor mycosis is difficult to diagnose early due to its vague symptoms ,but once it involves the orbit, it becomes too late to save the vision as well as patients life as it spreads very fast like wild fire.[1] Mucormycosisis seen in immunocompromised individuals with uncontrolled diabetes mellitus,solid organ transplant, iron overload, chronic use of corticosteroids, neutropenia, haematological malignancies.[2]

Fulminant infection of nose and paranasal sinus by mucormycosis is well known but spectrum of the disease is changing. We found a changing trend in presentation of mucor mycosis from typical presentation to atypical in immunocompetent individuals with very vague complaints at presentation. The infection of posteriorethmoid sinuses without involvement of middle turbinate or any anterior group of sinuses, spreads to orbit and its apex through lamina papyracea leading to orbital apex syndrome and optic nerve when affected leads to loss of vision.

Chances of isolated sphenoid sinus involvement is very rare.[3] It is because of anatomical location of sphenoidand decreased nasal airflow in that area. The symptoms in these patients are different such as headache, retro orbital pain, diplopia, exophthalmos, and blindness. This is due to rapidly progressing infection causing early destruction of bony walls of sphenoid sinus and surrounding vital structures such as optic nerve, internal carotid artery, cavernous sinus and pituitary gland.[4] fungus may invade the cavernous sinus and the brain parenchyma which may produce vascular thrombosis and infraction.[6]

Orbital apex syndrome as presentation is very rare ,characterised by retro orbital pain, vision loss and complete ophthalmoplegia with involvement of the II, III,IV, VI, V1 nerves.[7] Other syndromes like superior orbital fissure and cavernous sinus syndromes may show similar symptoms except for loss of vision 7. Rhino orbito cerebral mucor shows 3 stages during its natural course, sinus involvement which may be unnoticed , symptomatic orbital involvement and a cerebral involvement.[8] Superior orbital fissure contents such as cranial nerves III, IV, VI and branches of V1, V2 when involved causes diplopia , ophthalmoplegia,[8] and loss of corneal and face sensations.[1]

Early Diagnosis of mucormysis in its atypical presentation is challenging and requires a keen eye towards it .Due to this insidious onset and no nasal complaints in patients, cases are missed, and diagnosis is delayed. They spread like wild fire and present as orbital apex syndrome with loss of vision or intra cranialextension. This study is aimed at bringing this atypical presention to notice.

MATERIAL AND METHODS

A Total of 207 patients of post covid mucormycosis were admitted in our hospital from December 2021 to November 2022 .

Patients with minimal nasal complaints like nasal discharge ,pain at root of nose, headache and orbital symptoms like diplopia , blurring of vision, minimal proptosis, epiphora ,upper and lower eyelid swelling, were included in our study.

A detailed history including predisposing factors and comorbidities gave a clinical suspicion of mucor. Diagnostic nasal endoscopy, nasal smear for KOH mountand biopsyand CECT PNS scan was done . MRI scan of Nose , paranasal sinuses and orbit was done wherever required . Patients with Disseminated mucormycosis, intracranial extension, and extensive orbital involvement with loss of vision were excluded from the study.

Treatment started empirically with Injection Liposomal amphotericin B at 5mg/kg body weight on admission along with other symptomatic medications .We worked as a team formed by ENT surgeon, general physician, ophthalmologist, neurosurgeon and anaesthesiologists. Surgical debridement of involved areas in nose, sinuses, and orbit was done. Tissue sent for fungal culture and histopathology for confirmation .

All patients were divided into 4 groups based on symptoms at presentation, DNE findings, Radiology and treatment followed .Nasal smear for KOH mount and histopathology is positive for mucor in all the groups.

All patients were given injection Liposomal amphotericin B 5 mg /kg body weight/day added to 100 ml of 5% dextrose slow iv for 15 to 20 days followed by Tab.Posoconazole 300 mg/day in divided doses at discharge from hospital. Surgical debridement was done in group II,III and IV along with medical management.In group III and IV patients, Intraorbitalretrobulbar injection liposomal amphotericin B was given as 5 doses on alternate days along with intravenous amphotericin B.Group IV patients underwent FESS and orbital decompression along with 5 doses of retro bulbar amphotericin injections . Patients are followed up for 1 year in ENT opd.In each follow-up patient is accessed for control of disease and recurrence using tools like DNE and Radiology on suspicion of recurrence.

Group	Symptoms	DNE	Radiology	Treatment
I	vague nasal symptoms, no orbital symptoms	congested and edematous mucosa	Normal	Injection Liposomal AmphotericinB ,and other symptomatic treatment

II	Nasal complaints and headache , no orbital complaints	oedema and mucin in middle meatus and spheno ethmoid recess	CECT PNS showing Anterior and posterior group of sinuses involvement	Injection liposomal amphotericin B + FESS
III	Nasal and minimal orbital symptoms like blurring of vision and ,periorbital oedema	Fungal mucin ,minimal eschar	CECT showing nose, sinusesand periorbita involved but no orbital involvement	Injection liposomal amphotericin B + Surgical debridement including FESS + Intra orbital retrobulbar injection of liposomal amphotericin B
IV	Nasal and orbital symptoms ,ophthalmoplegia without loss of vision	Eschar in nasal cavity	CECT and MRI showing involment of nose, sinuses and Orbit	Injection liposomal amphotericin B + Intra orbital retrobulbar injection of liposomal amphotericin B +Surgical debridement including FESS +Orbital decompression

RESULTS

In our study done in 207 patients of mucor mycosis , male predilection (2:1)was observed and most common age group is 40-60years.patients are being categorised into4groups based on their symptoms at presentation , DNE findings ,CT findings and orbital symptoms .Among 207patients

- Group I includes 16 patients who had minimal nasal symptoms ,
- Group II includes74patients,who had nasal and sinus involvement
- Group III includes 63patients ,who had nose ,sinus and peri orbita involvement
- Group IV includes 54 patients ,with nose sinuses and orbit.

Group II and III are the most common presentation to ENT opd with nasal and minimal orbital symptoms.

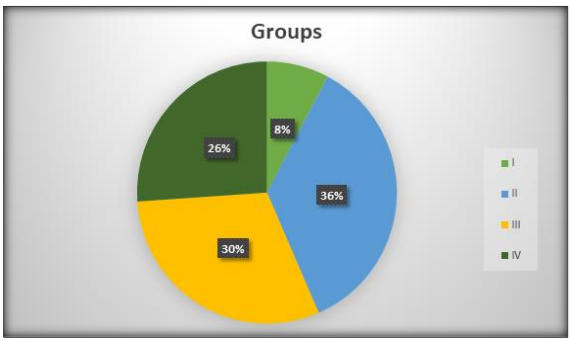

Figure 1: Paranasal sinus involvement

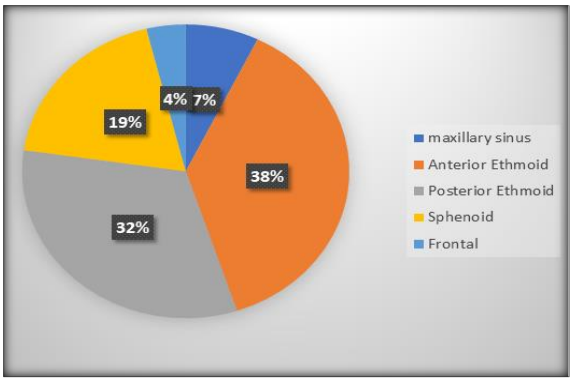

Figure 2

Anterior ethmoids(78) and posterior ethmoids(67) ,sphenoids(39) ,maxillary(15) and frontal sinuses(8) were involved in the same order indicating the change in the trend of involvement of posterior ethmoids and sphenoids more commonly than maxillary and frontal sinus .

Medical treatment was given to all patients with injection liposomal amphotericin B ,FESS was done in 191 patients ,Intra orbital retrobulbar injection liposomal amphotericin was given in 63 patients and orbital decompression was done in 54 patients.

All patients were followed up for a period of 1year .Group I patients required Tab.posaconazole 300 mg /day per orally in divided doses ,recovered 95 % in 3-4 months .Group II patients required treatment for 4-6 months and recovered by 92 % .Group III patients also required treatment for 4-6 months but recovered by 78%.Group IV patients required treatment for 9 months to 1 year and recovery rate was 63 %.18%of cases developed recurrence who were readmitted and required surgical debridement and injection liposomal amphotericin B.

DISCUSSION

We had treated maximum number of mucor mycosis patients presenting to our institute as it was declared as the nodal centre for covid and mucor mycosis for our state .There was a mucor epidemic following second wave of covid which presented with nasal symptoms, middle turbinate and maxillary sinus involvement, loose tooth, palate involvement, rarely orbit, frontal sinus ,and brain .Those patients with orbit and cerebral involvement had very bad prognosis and was fatal.

There is a change in trend of presentation of mucor mycosis following third wave of covid when compared to those following second wave. Most of the times, clinical presentation is atypical with vague headaches, showing unnoticed nasal discharge or obstruction , ethmoids and sphenoid sinus involvement more commonly than maxillary and frontal sinuses as seen in our study and orbital involvement more common than palate and maxilla.[5] Orbital involvement drives the patient for consultation.[5] Our study showed similar atypical presentations like nasal discharge, occipital headache, periorbital oedema, diplopia, blurring of vision. Diagnostic nasal endoscopy of all suspected cases should be done which may show stippled mucosa, thick fungal mucin, oedematous mucosa or a frank black eschar which represents

devascularised areas. Nasal swab for KOH mount and biopsy of representative sample of nasal mucosa for histopathology clinches diagnosis. Contrast enhanced CT scan initially may be misleading as it may show simple thickening of sinus mucosa indicating inflammation,[5] instead of irregular heterogenous soft tissue enhancement with focal areas of non enhancement indicating tissue necrosis and bone destructions.[6] Normal CT scan or MRI initially does not exclude the diagnosis, imaging may have to be repeated with contrast enhancement and dedicated orbital imaging,[7] on strong clinical suspicion during the course of treatment.

Treatment of mucor to be successful involves three principles.[6] Firstly early diagnosis with keen eye of suspicion to diagnose even atypical presentations. Secondly surgical debridement which forms main stay of therapy as it allows the tissue penetration of anti fungals, and reduces the fungal load.Third is medical therapy with broad spectrum antifungal liposomal amphotericin B along with correction of risk factors and comorbidities.

There is no fixed protocol to say how much to resect in surgical debridement, removal of all devitalised tissue aggressively till fresh bleeding is encountered is important. This can be done by open or endoscopic approach .In our study we have done functional endoscopic sinus surgery (FESS)and orbital decompression also in few cases where orbits were also involved. Orbital involvement with diplopia, blurring of vision, endoscopic orbital decompression along with FESS plays a major role in improvement of symptoms.[2,9]

Liposomal amphotericin B 3.5 to 5 mg /kg body weight mixed in 100ml of 5% dextrose infused slowly is the main stay of medical therapy .Tab posaconazole300mg /day in divided doses is the alternate therapy. In mucor patients presenting with minimal orbital complaints without vision loss retrobulbar injection of liposomal amphotericin B helps in disease control. Drug concentration reaches the orbit in high concentration without systemic side effects,this reduces orbital complaints like proptosis ,improves extraocular muscle movements and ptosis.[10] In progressive rhino orbital disease ,retrobulbar injection 5 doses daily for 5 days is an effective alternative to orbital exenteration.[10] In our study patients presenting with minimal orbital complaints in group III and IV were given 5 doses of retro bulbar injection of liposomal amphotericin B as a measure to restrict progression of orbital disease .Drug delivered at a dose of 1 ml (3.5 mg/ml),[11] on alternate days improved the compliance of the patient.

Mortality rate was as high as 60% in patients who were treated with medical therapy alone when compared with patients treated with both surgical and medical therapy which was 11% only.[6,8] This signifies the importance of surgical debridement.

CONCLUSION

Mucor mycosis after the third wave of Covid -19 pandemic, there is a significant change in presentation probably due to better immune status of individual. The presentation is vague nasal complaints and involvement of posterior group of sinuses more commonly with almost normal looking anterior group and maxilla. Identification of mucor in early stage is important as it involves orbit early in the course of disease, and if not recognised it may lead to fulminant orbital apex syndrome. Have a keen eye in picking up mucor mycosis patients early, diagnosis is confirmed by KOH mount and biopsy of suspected nasal mucosa for histopathology Normal imaging with CT and MRI does not exclude mucormycosis. Repeat orbital imaging should be considered if there is strong clinical suspicion Treatment plan may differ and tailored according to disease severity, extension and patient's medical condition. Medical Therapy with antifungals and adequate surgical debridement is required to prevent morbidity and gives better prognosis.

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