CASE REPORT

Rhinocerebral Mucormycosis with Isolated Sixth Nerve Palsy in An Immunocompetent Patient

Shraddha Jain, MBBS, MS(ENT), MD, Sunil Kumar, MBBS, MD, Amit Kaushal, MBBS

Department of Otolaryngology and HNS and Department of Medicine, Jawaharlal Nehru Medical College, DMIMSU, Sawangi (Meghe), Wardha, Maharashtra 442005, India

SUMMARY

We report a case of rhinocerebral mucormycosis in a 31 year old immunocompetent female presenting initially like acute rhinosinusitis with nasal stuffiness, severe headache, vomiting who soon developed isolated right lateral rectus palsy. Computed tomography(CT) scan of the Post-Nasal Spaces(PNS) showed an ill defined expansile heterogenous density mass in the sphenoid with extension into the ethmoids, nasal cavity, optic canal, superior orbital fissure, clivus and right temporal lobe with signal void in Magnetic Resonance Imaging(MRI). The debris and polypoid mucosa obtained on nasal endoscopy revealed mucormycosis on histopathologic examination. The patient was managed with urgent surgical debridement and medical management.

KEY WORDS:
Rhinocerebral, Mucormycosis; Immunocompetent; Debridement; Itraconazole

INTRODUCTION

Rhinocerebral mucormycosis is a rapidly progressive acute fungal infection which if not recognized early and treated promptly may be rapidly fatal. It almost always occurs in individuals who are immunologically or metabolically compromised, like those with diabetes mellitus, chronic renal disease and malignancies. There are very few reports in immunocompetent individuals.

CASE REPORT

A 31 year old female presented to the Otorhinolaryngology Out-Patient Department with complaints of nasal stuffiness and frontal headache, initially treated as acute rhinosinusitis for 5 days. On the day of admission, the patient developed a severe right retro-orbital pain and right lateral rectus palsy (Fig.1). Suspecting intracranial complication due to worsening of headache with development of vomiting, we admitted the patient and put her on broad spectrum injectable antibiotics including piperacillin-tazobactam, amikacin and metronidazole. There was no past history of nasal disease, allergies, tuberculosis, diabetes or chronic kidney disease. The patient did not have any addictions.

Anterior rhinoscopy showed deviation of nasal septum to left with oedematous and congested nasal mucosa. Besides an isolated right lateral rectus palsy, eye examination revealed normal vision and fundus examination. There was no lid or periorbital swelling. Other cranial nerves were normal.

Nasoendoscopic examination done on the same day revealed edematous right middle meatus with polyp in left middle meatus. Nasal mucosa was slightly congested. Thick greenish mucinous secretions were present in right middle meatus which were sent for aerobic, anaerobic and fungal culture and sensitivity. Bilateral uncinate processes were polypoidal. The culture reports came negative for all organisms.

Her haematological investigations were normal with haemoglobin of 12g%, normal blood counts and peripheral smear. Her blood sugar and kidney function tests were normal. Enzyme-linked Immuno-Sorbent Assay for HIV was negative.

CT scan orbit and PNS showed ill an defined expansile heterogenous density mass lesion with minimal post contrast enhancement arising from sphenoid sinus eroding all its walls involving on the right side, the optic canal, superior orbital fissure, cavernous sinus, with extension to right temporal lobe; anteriorly into bilateral ethmoid air cells, posteriorly up to the clivus; and superiorly eroding the floor of sella(Fig-2). The mass was seen to cause erosion of the infundibulum and uncinate process of maxilla on both sides. The diagnosis of fungal granulomatous disease or malignancy was considered. MRI to confirm this revealed an expansile hypointense lesion on T1 weighted image and hyperintense onT2 weighted image with signal void in the sphenoid sinus. (Fig-3). This signal void gave the strong possibility of fungal infection. So oral itraconazole in the dose of 100mg twice daily was started empirically even before taking the patient for surgery on the basis of radiological suspicion.

Functional endoscopic sinus surgery and bilateral uncinectomy with middle meatal antrostomy was done 24 hours after admission. Both anterior ethmoids, posterior ethmoids and sphenoid sinuses were opened. Black fungal debris was found in both posterior ethmoids and sphenoid sinus which was removed by curettage and suction irrigation. The mucosa of posterior ethmoids appeared to be normal with no polyp formation. Histopathology revealed mucormycosis with features of angioinvasion and acute inflammatory cells. The patient was taken to the operation theatre on multiple occasions for repeated endoscopic clearance of the fungal debris from sphenoid sinus as

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Corresponding Author: Shraddha Jain, Department of Otolaryngology and HNS and Department of Medicine, Jawaharlal Nehru Medical College, DMIMSU, Sawangi (Meghe),Wardha, Maharashtra 442005, India Email: sjain_med@yahoo.co.in

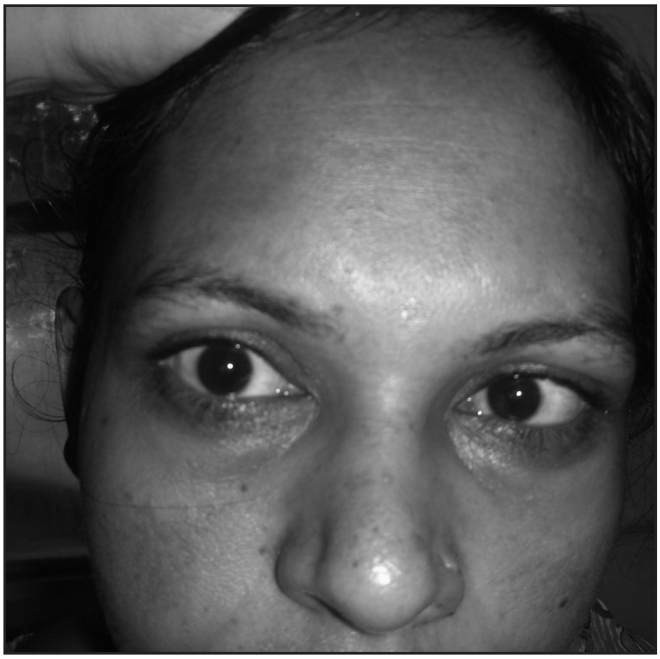

Fig. 1: patient photograph showing right lateral rectus palsy.

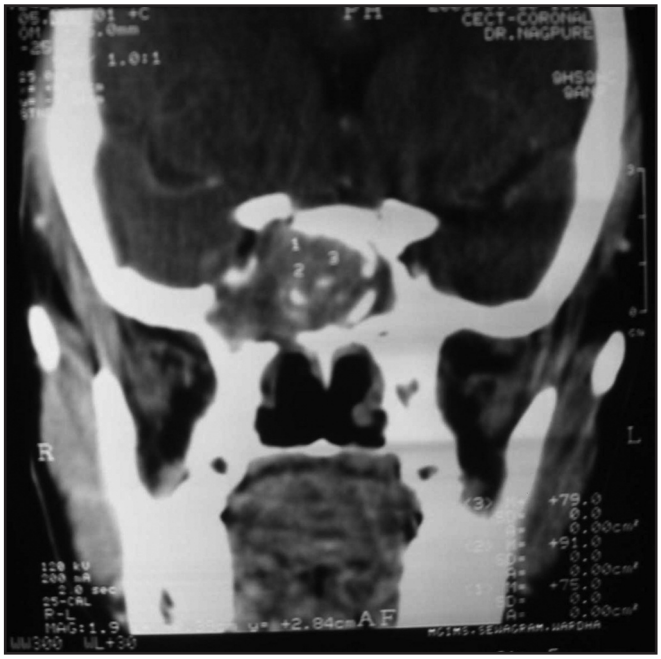

Fig. 2: CT scan (coronal cuts) showing expansile heterogenous density mass lesion arising from sphenoid sinus with erosion of its wall.

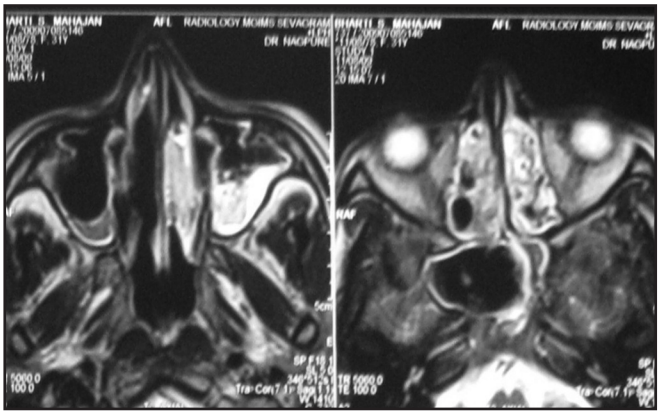

Fig. 3: MRI showing hyperintense onT2 weighted image with signal void in sphenoid sinus.

clearance was difficult to achieve from the posterior wall where erosion was present.

The dose of oral itraconazole was increased to 100 mg thrice daily. The lateral rectus palsy of the patient improved. Headache and vomiting resolved completely. Patient was discharged stable on oral antibiotics and antifungals for three weeks, and is doing well on follow up.

DISCUSSION

Rhinocerebral mucormycosis is the commonest form of systemic mucormycosis[1,2]. It is common in diabetics and other immunocompromised patients but the disease is reported even in normal individuals, especially zygomycetes fungi[3,4]. In rhinocerbral mucormycosis, the infected nasal mucosa may appear normal in earliest stage, then progresses through an erythematous phase, with or without edema, followed by violaceous appearance, and finally the development of black, necrotic nasal or palatal eschar as blood vessels become thrombosed with ensuing tissue necrosis. So, both the absence of facial or periorbital swelling and normal looking nasal mucosa in an immunocompetent patient does not always mean early disease or less likelihood of mucormycosis as the disease may predominantly involve the posterior sinuses with orbital apex and intracranial extension with limited disease in nasal cavity or anterior sinuses. Early and repeat endoscopic examinations with biopsies are required for confirmation of diagnosis as the disease spreads rapidly.

The histopathological hallmark of the disease is angioinvasion by irregular broad, nonseptate hyphae that branch at right angles with a predilection for the internal elastic lamina of blood vessels. Culturing the organisms from a potentially infected site is not the investigation of choice for diagnosis of mucormycosis as the fungus is ubiquitous, may even be present as a commensal in normal individuals or may be a laboratory contaminant. Moreover, the organism may be killed during processing of tissue specimen for culture[1].

Rhinocerebral mucormycosis requires urgent extensive and repeated debridements to remove of all fungal debris and involved tissues along with antifungal therapy on suspicion of the diagnosis. There is doubt regarding antifungal treatment as there have been no prospective randomized trials to define optimum antifungal therapy for mucormycosis. Amphotericin B deoxycholate and its lipid derivatives have been used extensively. Itraconazole is the only marketed azole drug that has in vitro activity against Mucorales. It has been found to have in vivo activity against a strain of Absidia and variable in vitro activity against Mucor

and Rhizopus. It has been found to have in vitro activity against Apophysomyces elegans. There are case reports of successful therapy with itraconazole alone[5].

The predictive value of in vitro susceptibility tests on the therapeutic outcome is not yet proven. Moreover the low interlaboratory reproducibility of *in vitro* susceptibility tests against fungi and difficulty in culturing the fungal strains makes it difficult to decide the optimum and correct antifungal drug. Since our patient showed good initial clinical response with empirical itraconazole therapy even before confirmation of diagnosis, we continued the drug with repeated surgical debridements and patient recovered well and is normal on follow up.

CONCLUSION

This report highlights the possibility of occurrence of this rapidly fatal condition even in an immunocompetent patient with normal looking nasal mucosa. High index of suspicion is required to prevent complications as the course of the disease is very rapid. In our patient with a very short history of five days of symptoms, the diagnosis was difficult with disease involving predominantly the posterior group of sinuses. Repeated surgical debridement of fungal debris and prompt medical management with itraconazole showed good clinical response.

REFERENCES

1. Spellberg B, Edwards J, Ibrahim A. Novel perspectives on mucormycosis: pathophysiology, presentation, and management. Clin Microbiol Rev 2005; 18: 556-69.
2. Munir N, Jones NS. Rhinocerebral mucormycosis with orbital and intracranial extension: a case report and review of optimum management. J Laryngol Otol 2007; 121: 192-5.
3. Roden MM, Zaoutis TE, Buchanan WL, Knudsen TA, Sarkisova TA, Schaufele RL, Sein M, Sein T, Chiou CC, Chu JH, Kontoyiannis DP, Walsh TJ. Epidemiology and outcome of zygomycosis: a review of 929 reported cases. Clin Infect Dis 2005; 41: 634-53.
4. Elinav H, Zimhony O, Cohen MJ, Marcovich AL, Benenson S. Rhinocerebral mucormycosis in patients without predisposing medical conditions: a review of the literature. Clin Microbiol Infect 2009; 15(7): 693-7. Epub 2009 Jul 16.
5. Eisen, D. P., and J. Robson. Complete resolution of pulmonary Rhizopus oryzae infection with itraconazole treatment: more evidence of the utility of azoles for zygomycosis. Mycoses 2004; 47: 159-62.