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MUCOR MYCOSIS: A FULMINANT CASE WITH EXTENSIVE INVOLVEMENT AND REMARKABLE RECOVERY

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ABSTRACT

Mucor mycosis is a rare but fatal illness. It is seen in diabetics and immune compromised individuals. A case is presented with fulminant course, rapid progression and extensive involvement. He was treated with Fungizone and Itraconazole alongwith surgical debridement of infected nasal mucosa. Recommendation is made to increase the awareness of this fatal illness among local practitioners.

Keywords: Mucor mycosis, Fungizone, Diabetics.

INTRODUCTION

Mucor mycosis is a fatal fungal infection of nose that often invades brain. Intracranial spread can occur through the ophthalmic artery, superior fissure, or cribriform plate. The fungi invade the blood vessel lumina and cause thrombosis through inflammatory occlusion. Strong association with diabetes has been described. Mortality rates of 30-70% are quoted in literature. Survival rate declines when interval from diagnosis to treatment is more than 6 days.

A case is presented with widespread fulminant involvement. He was given dual antifungals and showed remarkable recovery with some residual deficit due to complication.

CASE REPORT

A 55yrs old male came to our outpatient for one day history of difficulty in use of right hand. Eight days earlier he had developed urticarial rashes all over the body. He was given some injectables by local general practitioner that relieved the symptoms. Three days later, he developed pain in left temple. Gradually left face became hard and swollen. Next morning he was not able to open left eye. Pain improved by some injectables given by local GP. There was no vomiting and he had remained afebrile throughout. Evening before admission, he developed weakness of right hand that brought him to hospital. He was diabetic and hypertensive for 5-6 years which was not very well controlled.

On examination he had a large swelling of left cheek, involving conjunctiva and eyelid.

His speech and gait was normal. Vision on left was limited to hand movement only. There was afferent pupillay defect on left side. Right pupil was 6mm round and reacting briskly to direct and very sluggishly to indirect stimulus. Left pupil was nonreacting at all. Left Massaters and Temporalis were normally contracting but there was decreased pain sensation on left V1 and V2 distribution. There was severe LMN type facial weakness on left. There was no hearing loss. Palate and Tongue were also normal. Shoulder abduction was 3+/5 on right but there was no movement at fingers. Power in left hand and both legs was normal. There was no gross difference in DTRs on the two sides. Plantars were both flexor. There was no loss of pain over the body and joint position sense was normal. On investigation Total leukocyte count was 18.7. ESR was 90mm/1st hr. Random blood sugar was 426mg%. Urea, creatinine and electrolytes were normal. Blood culture was negative for bacterial growth. CAT scan head and brain was unremarkable. MRI showed a small infarct in left parietal cortex. A large soft tissue swelling was also seen in left cheek extending upto the temple and into left orbit.

CAT scan cervical spine was normal. Endoscopic nasal examination showed black discoloration of left middle turbinate. Biopsy was taken. Fungal smear showed few aseptate fungal hyphae.

Diagnosis of Mucor Mycosis was made and Injection

Fungizone (Amphotericin B) 20mg was started that was later gradually increased to 80mg IV OD. Three days later, Cap ITRACONAZOLE 200mg BD was added. Over next one week swelling was seen to have regressed a little. After about 10 days of treatment he was sent for Endoscopic nasal debridement (FESS). There he became unconscious. When examined he was barely arousable. Two days later he opened eyes but was mute and his right side was moderately weak. Repeat MRI scan head showed occlusion of Left ICA with a big Left MCA territory infarct.

Antifungals were continued and Anti platelets were added. Renal and Hepatic functions were carefully monitored. Serum K+ used to be on lower side and was constantly replaced orally.
After 3 weeks of treatment he developed fever (102oF) and passed small amount of blood from right (opposite) nostril. Blood cultures and 5 sets of MPs were sent that were negative. A five day course of Cap. Ofloxocin 500mg was given. Fever subsided in three days. He never bled later on.
Amphotericin was held for two days during fever.

After one month of treatment nose was reexamined with rigid endoscope. Necrotic material and blood clots were seen that were removed and healthy mucosa came out.

Amphotericin was stopped after 5 weeks (total of 3.5Gm). Patient was discharged after 1½ months of stay. Itraconazole was continued for next 3 months.

He made a follow up visit in 5th month. Swelling was no more there. He had started to speak a few words. He could understand most of the things. He could follow complex commands and could express emotions. He could walk with unilateral support. There was total ophthlmoplegia in left eye and there was no vision.

DISCUSSION

First case of Mucor Mycosis was described by Paltauf in 18851. Since then little has changed in diagnosis and outcome of the disease2. Mucor mycosis is a rare, often fatal fungal infection. It is caused by an aerobic saprophytic fungus belonging to the order mucorales and class zygomyecetes. It is a common inhabitant of soil, decaying vegetation and can also be cultured from human nose, throat, oral cavity and stools. It typically originates in the nasal or oral mucosa, spreads to the paranasal sinuses and enters the orbit via the ethmoid and maxillary sinuses or via the nasolacrimal duct3. It may also spread into the neck4. We had CT scan neck and cervical spread was ruled out.

Mucor mycosis is the third most common mycosis after candidiasis and aspergillosis. It is also now increasingly recognized in India, especially in patients with diabetes or trauma. Based on anatomic localization six forms have been described (1) Rhinocerebral, (2) Pulmonary, (3) Cutaneous, (4) Gastrointestinal, (5) Disseminated and (6) Uncommon presentations5. Isolated involvement of kidneys have also been reported6.

Our case was a diabetic and was not compliant of medicines. His illness started with some urticarial rash followed by headache alongwith swelling of left side of face. Perhaps the rashes were not associated with the Mucor infection because it subsided by local treatment most likely anti allergic or steroids, never recurred and when was examined after admission there was no skin involvement. He continued to go to local practitioner who could not realize the gravity of situation. It was finally the weakness of right hand that brought him to us. He had involvement of multiple cranial nerves of one side. The suspicion was high about that been mucor7. MRI scan showed a soft tissue mass in the left facial area. It was seen involving the orbit and the nasal cavity too. In addition a small infarct in the superior parietal area was seen describing the weakness of hand. Association of mucor with stroke has been described8.

The overall survival rate of patients with mucormycosis is approximately 50%. Rhinocerebral mucormycosis has a higher survival rate than does pulmonary or disseminated mucormycosis9.

Rhinocerebral mucormycosis requires urgent extensive and repeated debridements to remove all the fungal debris and involved tissue along with antifungal therapy on suspicion of the diagnosis10. Current data supports the first-line use of high-dose liposomal amphotericin B for mucormycosis, particularly for cases of central nervous system disease, with amphotericin B lipid complex serving as a reasonable second-line agent. A cumulative dose of at least 3 g of amphotericin B is recommended to treat rhinocerebral phycomycosis11. Currently, novel regimens for the treatment of mucormycosis include combination of lipid-based amphotericin plus either an echinocandin or itraconazole or both12.

Our patient underwent surgical debridement of the nasal infection. It was associated with the combination

Figure 1: Huge swelling Left cheek (with permission)

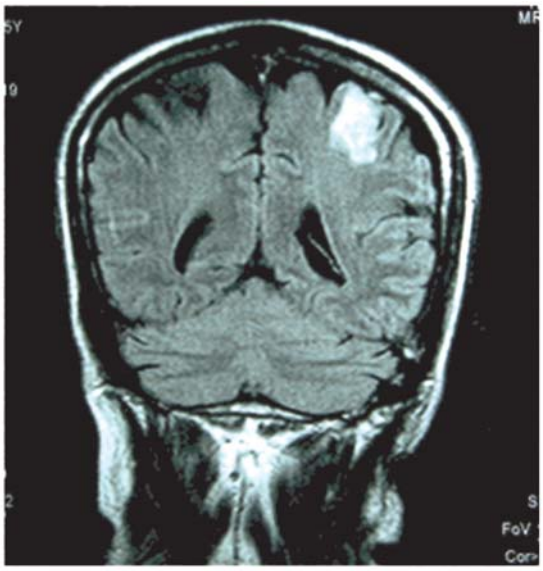

Figure 2: Small Left superior parietal cortical infarct.

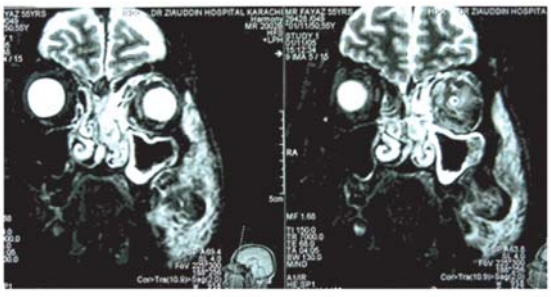

Figure 3: Huge swelling of soft tissue in left check eye and nose

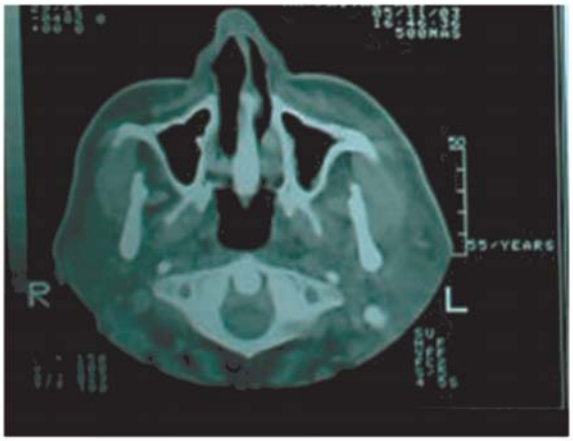

Figure 4: CT scan cervical region did not show any extension of the mass into the neck.

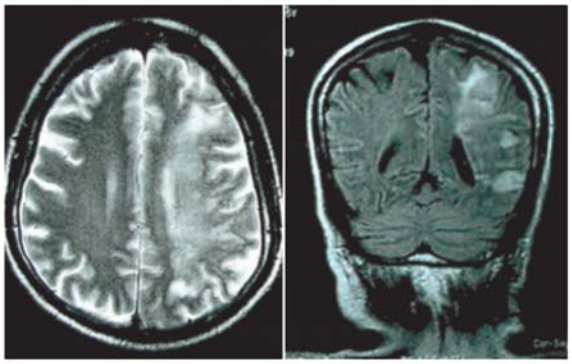

Figure 5: Repeat MRI scan showing major left MCA infarct.

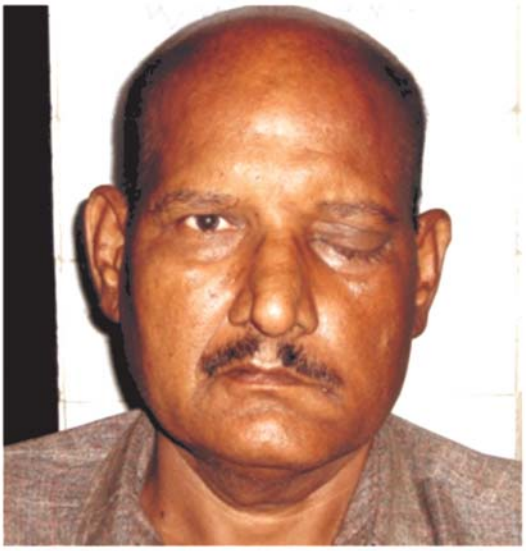

Figure 6: Follow up visit after five months showing remarkable recovery. (with permission)

of Fungizone (total of 3.2 Gm was given) and Itracocazole (continued for 3 months). He had started to show signs of improvement during hospital stay. The size of the facial swelling started regressing. Unfortunately he developed a large MCA infarct on left side. Treatment, however, was continued. In the follow up visit there was complete subsidence of swelling although he had lost the eye and facial weakness was permanent.

CONCLUSION

High index of suspicion is required to prevent complications as the course of the disease is very rapid. Awareness among the local practitioners is recommended as in our case the local practitioner could not recognize the illness and kept on treating symptomatically.

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