Isolated Cerebral Mucormycosis in an Immunocompetent Patient—A Case Report

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Introduction

Rhino-orbitocerebral mucormycosis is the most common form of central nervous system (CNS) infection with mucormycosis. It is a rare but life-threatening infection. Isolated cerebral infection with mucormycosis without any rhino-orbital focus is extremely rare. Isolated cerebral mucormycosis is common in intravenous (IV) drug abusers and immunocompromised patients. Diabetes, long-term steroid usage, and immunosuppression are the important risk factors for the infection. Usually the disease presents as an acute illness with rapid course and is fatal if untreated. In a case review of 30 cases by Verma et al in 2006, 11 patients had associated systemic infection, 17 had history of IV drug abuse, and 2 were diagnosed on the basis of surgical and histopathological features.1 Our patient is a healthy immunocompetent, nondiabetic woman presenting with gradual onset of symptoms similar to a slow-growing tumor in comparison to a classical rapid course in patients with mucormycosis.

Case History

A 54-year-old woman presented with complaints of headache and forgetfulness for 3 months. Her headache was gradual in onset, progressive in nature, more on the right side, and associated with retro-orbital pain, which got aggravated after work and walking and relieved by rest and medication. History of forgetfulness had been noticed by husband since past 3 months. There was no history of neck pain, fever, facial swelling, trauma, vomiting, or seizures. There is no history suggestive of chronic cough, nasal discharge, breathlessness, blood stained sputum, and loss of weight and appetite. The patient is not a known hypertensive or diabetic. The general examination of the patient was normal. CNS examination did not reveal any abnormal findings or any neurologic deficit.

Radiological Features

Computed tomography (CT) scan of the brain showed right temporal hypodense area (►Fig. 1). Further evaluation with magnetic resonance imaging (MRI) scan of the brain (plain and contrast) showed an isointense lesion on T1 WI, hyperintense/heterogeneous on T2 WI with heterogeneous contrast enhancement and disproportionate edema in the right temporal lobe (►Figs. 2–4).

Evaluation, Surgery, and Postoperative Care

A preliminary workup for metastasis was performed, including chest X-ray and ultrasound abdomen, which were normal. After a thorough preoperative evaluation, the patient was taken up for a right temporal craniotomy and...
excision of the lesion. Intraoperatively, the lesion was firm, grayish, and nonsuckable with multiple pus points. There were dense adhesions with the medial temporal dura, which were separated and a complete excision of the tumor was achieved. The postoperative period was uneventful. The pus sent for evaluation was negative for Gram stain, fungal stain, and acid-fast Bacillus (AFB) stain, and no growth was seen in culture and sensitivity. Her histopathology was suggestive of intense inflammation characterized by fibrosis, dense collection of neutrophils, lymphocytes, plasma cells, and scattered eosinophils. Occasional small, irregular broad septate fungal filaments with surrounding fibrocartilaginous wall suggestive of mycotic infection and staining was positive for silver methenamine and periodic acid–Schiff (PAS) stain, which was suggestive of mucormycosis (►Figs. 5 and 6). The patient was evaluated for the possible focus of rhino-ocular infection, but no focus was detected and was started on antifungal therapy with amphotericin B. She was planned for total dosage of 2 g of amphotericin B. In view of toxicity and side effects, it was administered in daily dosage of 1 mg/kg/day to attain a cumulative dose of 2 g. Her renal parameters, electrolytes, and platelet counts were regularly monitored during the course of the therapy. She developed

Fig. 1 Selected axial images of plain CT scan. Brain showing right temporal, predominantly white matter, hypodense area.

Fig. 2 Selected T1 axial images of brain MRI, showing ill-defined mixed intensity lesion with perifocal edema.

Fig. 3 Selected T2 axial images of brain MRI showing ill-defined mixed intensity lesion with perifocal edema.
mild side effects that were managed accordingly. The follow-up brain CT scan (plain and contrast) (►Fig. 7) at discharge showed no residual or recurrent lesion. The follow-up MRI scan of the brain (plain and contrast) done after 3 months of completion of antifungal treatment did not reveal any residual lesion (►Fig. 8).

**Discussion**

Mucormycosis is a systemic fungal infection caused by members of class Zygomycetes, order Mucorales. It is a rare but life-threatening infection.\(^1,3\)

Rhino-oculo-cerebral form is the most common form of mucormycosis\(^3\) followed by pulmonary and cutaneous disease.\(^3\) Usually infection with Mucor will present as an acute, life-threatening condition and is often fatal.\(^1,4\) Major predisposing factors include diabetes mellitus and immunocompromised status.\(^1,4,5\) The most important predisposing factor is uncontrolled diabetes with acidosis because the organism is believed to have an active ketone reductase system, which enables it to thrive in an acid pH “rich
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medium. Acidosis is also believed to permit invasion of the blood vessel walls by the fungus. Other predisposing factors include hepatic coma, uremia, leukemia, aplastic anemia, and transplant recipients on artificial immunosupression. Our patient is an immunocompetent woman without any of the predisposing factors mentioned.

Isolated infection of the CNS without rhino-orbital focus is rare. In our review of literature, a very few cases were noted with isolated CNS infection. Isolated CNS infection with Mucor is common in IV drug abusers, immunocompromised patients, and long-term uncontrolled diabetics. In a review of literature done by Verma et al on isolated cerebral mucormycosis, only two patients were without any systemic predisposition or IV drug abuse.

Chronic presentation in mucormycosis is not uncommon. Rumboldt and Castillo published a case of preputont Mucor infection with indolent course of over several months. A case series of four patients has been published, with radiologic features similar to lung carcinoma and a chronic clinical course similar to lung carcinoma but biopsy showing mucormycosis. Our case presented with nonspecific symptoms and clinical course of over 3 months. Indolent course in our patient may be due to the intense inflammation and fibrosis around the lesion on histopathological examination, suggesting a host reaction in an immunocompetent person.

Infection in an immunocompetent patient with extracerebral mucormycosis is in a rise during the present days. Sridhara et al in 2005 published a report of eight cases of mucormycosis in an otherwise healthy persons from 1999 to 2003; seven out of the eight cases were diagnosed in histopathological and microbiological examination after surgery. Sethi et al in 2012 have done a large observational study of cerebral fungal infections over a period of 2 years and an infection was found in 28% of immunocompetent persons.

Imaging characteristics definitive for mucormycosis are difficult to define. Because of the lack of inflammatory response, radiologic findings are often nonspecific and are frequently mistaken for tuberculous meningitis, pyogenic abscess, or brain tumor.

Cerebrospinal fluid (CSF) analysis with proper microbiological examination may help diagnose CNS mucormycosis. In 2012, Ramadevi et al published isolated cerebral mucormycosis case report based on CSF culture of mucormycosis.

Treatment includes early histological diagnosis and institution of antifungal therapy. Amphotericin B is presently the only available antifungal drug for systemic mycoses. Because penetration of the CNS by amphotericin B is poor, the application of high-dose therapy may be useful in cases of cerebral zygomycosis. Successful management of mucormycosis depends on early diagnosis, correction of predisposing risk factors, radical surgical debridement, and administration of active antifungal agents.

Conclusion

Isolated cerebral mucormycosis in an immunocompetent patient is rare. Early surgical excision, histopathologic diagnosis, early institution of therapy (antifungals), and regular follow-up may help in better prognosis for the patient.

References