**Rhinocerebral Mucormycosis**

—Case Report—

Hidenobu OCHIAI, Tsutomu ISEDA, Satoshi MIYAHARA, Tomokazu GOYA and Shinichiro WAKISAKA

Department of Neurosurgery, Miyazaki Medical College, Miyazaki

**Abstract**

A rare case of rhinocerebral mucormycosis occurred in a 74-year-old diabetic male with gradually progressive right visual loss and total ophthalmoplegia. Computed tomography and magnetic resonance imaging revealed an invasive right orbital apex mass, destroying the medial wall of the orbit and extending into the right cavernous sinus and right middle fossa. Laboratory data demonstrated no signs of inflammation. A carcinomatous lesion originating in the paranasal sinuses and extending into the intracranial space was diagnosed. The mass was totally removed through a subfrontal approach to confirm the histological diagnosis and decompress the optic nerve. The histological diagnosis was mucormycosis. Despite aggressive medical therapy, dissemination resulted in mucor pleuritis and mucor encephalitis or meningitis. He died of septic shock and acute renal failure.

**Key words:** mucormycosis, computed tomography, magnetic resonance imaging

**Introduction**

Mucormycosis is a quite rare fungal infection caused by ubiquitous opportunistic organisms of the genera, *Rhizopus, Mucor, and Absidia*. The rhinocerebral type is the most common and most important type for neurosurgeons. The most common predisposing factor of this disease is diabetes mellitus, because the presence of ketone, low pH, and hyperglycemia enhance fungal proliferation by hampering host phagocytosis and mobilization of polymorphonuclear leukocytes. The other predisposing factors are leukemia, bone marrow transplantation, and other immunosuppressive states.

As this disease is frequently fatal, early diagnosis and immediate treatment are indispensable. However, premortem or preoperative diagnosis of this disease is quite difficult. We report a patient with rhinocerebral mucormycosis and discuss neuroradiological findings and treatment for mucormycosis.

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**Case Report**

A 74-year-old male had had a right frontotemporal pain since April, 1990. His visual acuity gradually decreased and he became completely blind in the right eye in November, 1990. Right blepharoptosis also developed. He was admitted to our department in March, 1991. He had no history of fever, malignant neoplasms, or drug abuse.

Neurological examination on admission revealed blindness, blepharoptosis, and total ophthalmoplegia on the right side with absence of right corneal reflex and hypesthesia of the right face. No right orbital cellulitis was present. Laboratory data revealed hyperglycemia but he denied any history of diabetes mellitus, which suggested the existence of uncontrolled diabetic mellitus. No signs of inflammation were found.

Plain skull x-ray films showed that the right anterior clinoid process and ipsilateral sphenoidal plane were destroyed and the right superior orbital fissure was dilated. Computed tomographic (CT) scans disclosed an invasive mass extending into the right orbital apex, destroying the medial wall of the orbit and extending into the right cavernous sinus and right middle fossa (Fig. 1 left). Mucosal thickening in the ethmoid and sphenoid sinuses...
was also seen (Fig. 1 right). Magnetic resonance (MR) imaging demonstrated a hyperintense rim along the ethmoid sinus wall on the T2-weighted image (Fig. 2 left), and an orbital apex mass extending into the right cavernous sinus and right rectal gyrus, which was hyperintense on T1- and proton density-weighted images and homogeneously enhanced after gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA) administration (Fig. 2 center, right). A carcinomatous lesion arising from the mucous membrane of ethmoid sinus was considered most likely.

This mass was removed through a subfrontal approach to decompress the optic nerve and confirm the diagnosis histologically. The dura mater in the sphenoidal plane was thickened and adhered tightly to the right rectal gyrus. The mass had destroyed the ipsilateral anterior clinoid process and sphenoidal plane, partially penetrated the dura mater at this location and invaded the ipsilateral rectal gyrus. The mass was dark grayish-colored, elastic soft, and hypervascular. After total removal of the main mass, the orbital apex was opened and the optic nerve was decompressed. The optic nerve was tightly en- cased by the mass. Finally, the ethmoid sinus was opened. A yellowish hard mass had filled this sinus. Histological examination revealed an aggregation of fungal hyphae with broad and short branches at an obtuse angle (Fig. 3). The diagnosis was mucormycosis.

Systemic administration of amphotericin B was started intravenously and the blood sugar level controlled strictly. On the 3rd postoperative day, he began to complain of dyspnea. A great quantity of right pleural effusion was noted on chest x-ray films. Mucor pleuritis was considered and the pleural effusion drained continuously. Dyspnea improved after this procedure but about 2 liter of yellowish clear fluid was drained daily. On the 5th postoperative day, he developed status epilepticus. Postcontrast CT scans demonstrated brain swelling and diffuse gyral enhancement. Mucor encephalitis or meningitis was considered. On the 8th postoperative day, he fell into septic shock with concurrent acute renal failure and died on the 10th postoperative day in spite of aggressive treatment. Autopsy was not permitted.

**Discussion**

Rhinocerebral mucormycosis usually begins in the nose and paranasal sinuses. Plain skull x-ray examination usually demonstrates three typical signs, nodular thickening of the sinus linings, absence of air-fluid level, and spotty multicentric bone destruction. Multicentric bone destruction is an important finding to distinguish mucormycosis from a carcinomatous lesion arising from the mucosa of paranasal sinuses.

![Fig. 1](image1.png)

**Fig. 1** left: CT scan, demonstrating an invasive mass of the right orbital apex extending into the ipsilateral cavernous sinus and middle fossa. right: CT scan, showing mucosal thickening of the ethmoid sinus.

![Fig. 2](image2.png)

**Fig. 2** left: T2-weighted axial MR image, disclosing hyperintense rim along the ethmoid sinus wall. No air-fluid level was present in this sinus. center, right: T1-weighted coronal (center) and sagittal (right) MR images after Gd-DTPA administration, showing a hyperintense mass at the right orbital apex involving the ipsilateral internal carotid artery and invading the right rectal gyrus.
sinuses, and the absence of air-fluid level distinguishes it from bacterial sinusitis.

CT demonstrates various findings in proportion to the development of mucor invasion. In the period of sinus infection, mucosal thickening and absence of air-fluid level are noted. In the next stage, the mucor usually destroys the medial wall of the orbit and invades the medial rectal muscle, orbital apex, and ipsilateral cavernous sinus, resulting in swelling of the medial rectal muscle and of the optic nerve. The mucor often invades vascular vessel walls, especially the arterial wall, resulting in ipsilateral brain infarction, especially watershed infarction. This finding distinguishes mucormycosis from other diseases. An unenhanced superior ophthalmic vein is also a specific finding of this disease.2)

MR imaging of rhinocerebral mucormycosis has seldom been reported.5,7,8) There are three specific findings caused by mucor invasion. First, sinus involvement appears as a hyperintense rim along the sinus wall. Second, a hyperintense lesion on the T2-weighted images extends from paranasal sinuses to the intracranial cavity via the orbital apex.7) Third, narrowing or slow flow occurs in the ipsilateral internal carotid artery.7)

In our case, CT and MR imaging revealed mucosal thickening without an air-fluid level, and the mass arising from a paranasal sinus, destroying the medial orbital wall and extending into the cranial cavity. Therefore, we suspected mucormycosis preoperatively, although we diagnosed a carcinomatous lesion. The differential diagnosis from carcinoma in this region is very important. A carcinomatous lesion contains few clear sinus cells among opacified cells, and usually extends widely with bone destruction over a large area.14) Invasion of vascular vessels in the early stages is rare. Based on these points, the differential diagnosis of mucormycosis from carcinoma is not so difficult.

A paranasal mass extending into the intracranial space through the orbital apex in a patient with diabetes mellitus, metabolic acidosis, or immunosuppressive state indicates mucormycosis and the diagnosis should be confirmed by sinus biopsy as soon as possible. Once the diagnosis is made, immediate aggressive treatment consisting of surgical debridement, control of underlying disease, and systemic administration of amphotericin B should be started.1,8,10,12) In our case, there were two problems with the treatment. We debrided the mass as much as possible but the mucor were disseminated, resulting in mucor encephalitis or meningitis. Furthermore, in addition to general anesthesia and surgical suppression of immunity, the mucor was disseminated systemically regardless of aggressive postoperative nutritional support. Second, after confirming the diagnosis, we administered amphotericin B systemically. Amphotericin B causes renal toxicity, and when given to a patient in a dehydrated state, renal dysfunction occurs. In our case, a large amount of pleural effusion occurred, and despite fluid management, coupled with sepsis and disseminated intravascular coagulation, acute renal failure developed. Liposomal amphotericin B may prevent this side effect.9) Once the mucor was disseminated systemically, treatment becomes quite difficult and strict nutritional support, fluid management, and other systemic management are necessary in addition to administration of amphotericin B.

References


Address reprint requests to: H. Ochiai, M.D., Department of Neurosurgery, Miyazaki Medical College, 5200 Kihara, Kiyotake-cho, Miyazaki-gun, Miyazaki 889-16, Japan.