

CLINICAL PHOTOGRAPHS

Mucormycosis is an aggressive opportunistic infection caused by fungi of the class Zygomycetes, first described in 1885, the species most commonly involved being *Rhizopus* and *Mucor*. The disease develops in people with severe underlying diseases and appears extremely rare in immunocompetent persons. The most common clinical form is rhinocerebral mucormycosis (Figure 1).

Conditions most commonly associated with mucormycosis include diabetes (usually poorly controlled diabetes), chronic steroid use, metabolic acidosis, organ transplantation, leukemia, lymphoma, treatment with deferoxamine, and AIDS.

Syndromes associated with mucormycosis include:

- Rhinocerebral infection (infection of sinuses and brain): may start as a sinus infection, may progress to involve inflammation of cranial nerves, may cause blood clots that block vessels to the brain (thrombosis)
- Pulmonary mucormycosis – pneumonia that gets worse quickly and may spread to the chest cavity, heart and brain
- Mucormycosis of the gastrointestinal tract, skin and kidneys.

Symptoms of rhinocerebral mucormycosis include: eye swelling and protrusion (proptosis), dark nasal eschar (scabbing), fever, headache, redness of skin overlying sinuses, sinus pain or congestion.

Possible complications: blindness (if the optic nerve is involved), clotting or blockage of brain or lung blood vessels (thrombosis), death, nerve damage (Figure 2, 4a, b).

Mucormycosis, an extremely rare disease, is a surgical emergency, so early diagnosis and treatment are essential.

The diagnosis requires a high degree of clinical suspicion, in patients with suppressed immune status.

Treatment should include in the same time correction of risk factors, Amphotericin B in high doses for a long period of time (4-6 weeks), and aggressive surgical therapy, with removal of all necrotic lesions. Surgery should be done immediately to remove all dead and infected tissue. Surgery can lead to disfiguration because it may involve removal of the palate, parts of the nose, or parts of the eye. Without such aggressive surgery, however, chances of survival are greatly decreased.

Histological findings: Pathognomonic changes of broad, irregular, nonseptate, right-angled, branching hyphae are demonstrated by H&E and by specialized fungal stains. Vascular invasion and necrosis are the characteristic consequences of the infective process. A neutrophil infiltrate is typical, and a granulomatous reaction may be observed (Figure 3).

Mortality rate: 50-85% despite early treatment.



Figure 1 Complete necrosis of the inferior right turbinate (endoscopic view)



Figure 2 Necrosis of the hard palate

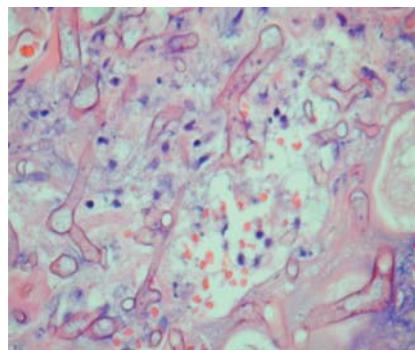


Figure 3 Histopathological examination

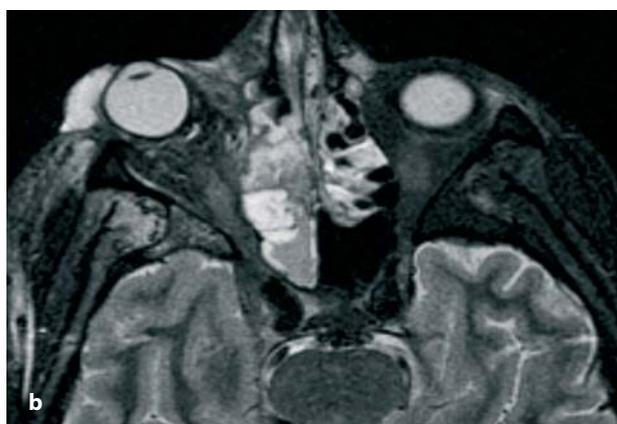
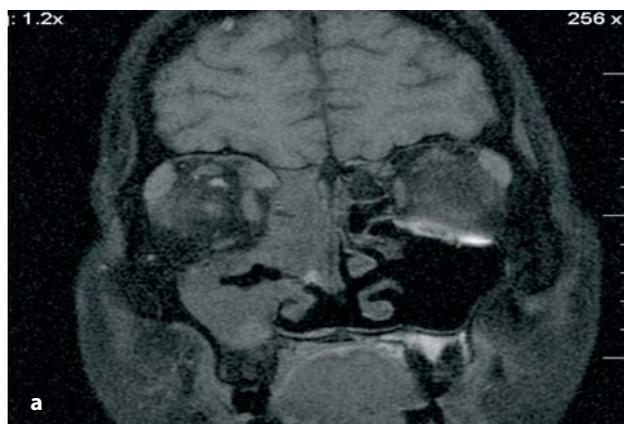


Figure 4a, b MRI showing invasive fungal pan sinusitis with invasion of the fungal right orbit