

# Rhinomaxillary mucormycosis: A palatal ulcer

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## Abstract

Rhinocerebral mucormycosis (zygomycosis) is an opportunistic fungal infection caused by saprophytic fungus. It involves several areas of the body, but the rhinocerebral form is most relevant to health care providers. Zygomycosis is associated with medically compromised patients. Our case reports an unhealed ulcer present over the palate of 15 days duration associated with swelling over the maxillary sinus region. This case is a blend of clinical, radiological, and histological manifestations of mucormycosis in a patient.

**Keywords:** CT scan, palatal ulcer, rhinocerebral mucormycosis, waters' view

## Introduction

Rhinocerebral mucormycosis is a fatal opportunistic infection caused by a saprophytic fungus belonging to the group of Phycomycete. Paltauf in 1885 described the first case of this uncommon infection in human beings. Six clinical variants such as "rhinocerebral, pulmonary, cutaneous, gastrointestinal, central nervous system, disseminated type" have been recognized so far. Here, we present a histopathologically proven case of rhinomaxillary mucormycosis presented as a deep palatal ulcer in diabetic patients.

## Case Report

A 32-year-old man reported to the department with the chief complaint of pain and swelling below the left eye obstructing the vision from the same eye from past 1 month and an ulcer on the roof of mouth from the past 15 days. Pain was gradual in onset, severe, throbbing, continuous, and radiating to the left temporal region associated with swelling over the left maxillary sinus region that was insidious in onset and gradually progressed to the present size causing nasal obstruction and oedema around the left eye. There was a history of recurrent

nasal discharge from the left nostril since 20 days. Around 15 days back, an ulcer developed over the palate which was rapidly increasing in size and was interfering with speech.

Patient's medical history revealed that he was uncontrolled type 1 diabetic patient from past 2 years and was on treatment (Inj. Mixtard 14 units per day). There was a family history of diabetes as patient's father had type 1 diabetes. The patient was tobacco chewer from past 15 years.

On general physical examination, the patient appeared febrile and the BP was recorded to be 160/90 mmHg.

On extraoral examination, a diffuse swelling was appreciated over the left maxillary sinus region extending superoinferiorly from the left supraorbital margin to a line joining the left corner of mouth to the left ear and anteroposteriorly from nasal bridge to left ear. Overlying skin appeared normal with no secondary changes and on palpation was soft to firm in consistency with diffuse borders and tender with no local rise in temperature [Figure 1].

On intraoral examination, an infiltrating ulcer approximately

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**Figure 1:** Extraoral picture depicting ptosis of the left eye and swelling over the left maxillary sinus region

3 × 4 cm<sup>2</sup> with irregular borders was appreciated over the hard palate. Ulcer was covered over by the necrotic slough and on the anterior aspect of the ulcer, part of the underlying bone was also exposed. Ulcer was nontender with few areas of erythema over the margins [Figure 2].

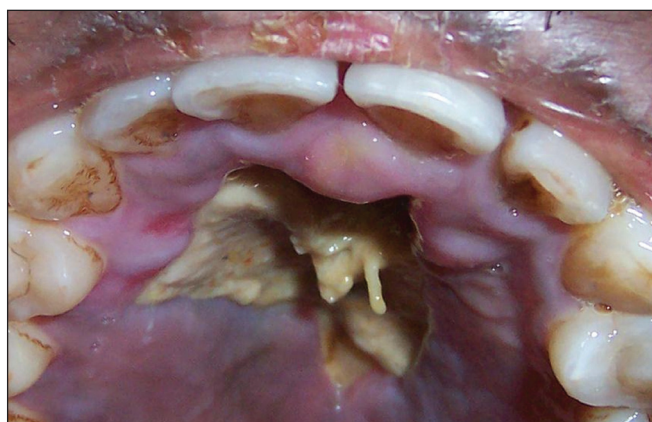
Considering the patient's medical history and a rapidly proliferating ulcer denuding the underlying bone, a provisional diagnosis of deep mycotic infection of palate was made. The differential diagnosis of midline lethal granuloma and malignant ulcer was thought.

Later the patient was subjected to blood investigations which revealed raised ESR (34 mm/1st hour) and random blood sugar around 220 mg%. Tridot and HBs test were negative.

### Radiological investigations

Water's view revealed diffused radiopacity over the left maxillary antrum extending into left nasal cavity [Figure 3].

CT scan in both axial [Figures 4a and b] and coronal sections [Figures 5a and b] revealed radiopacities along the walls of the



**Figure 2:** Intraoral picture showing a proliferating palatal ulcer

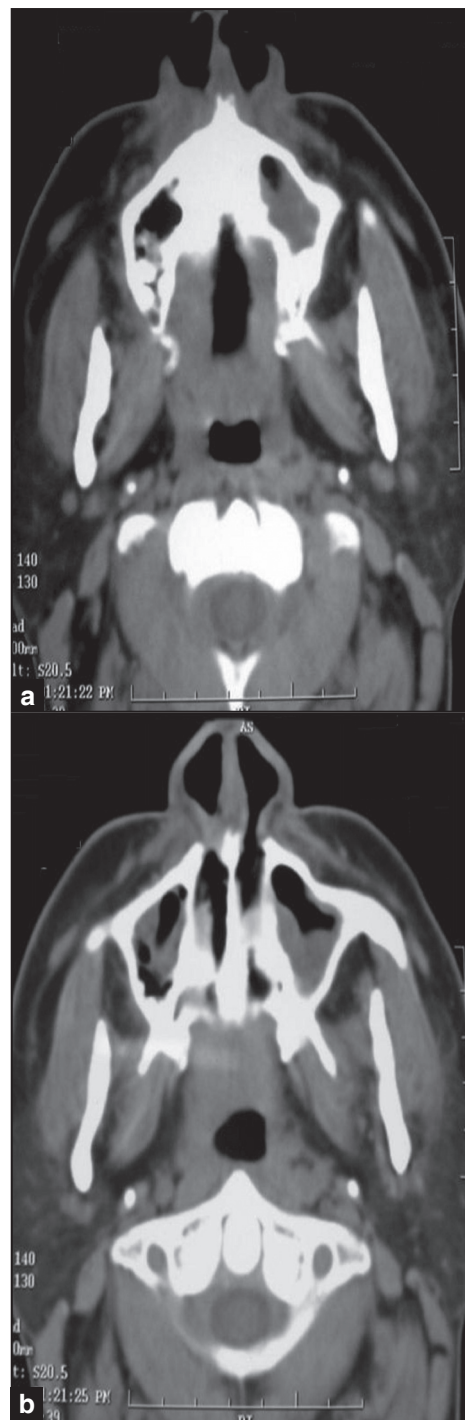


**Figure 3:** Waters' view showing opacification of the left maxillary sinus region

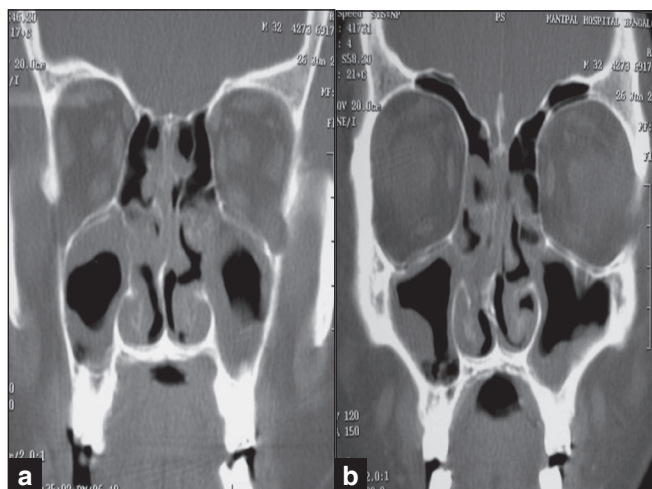
left maxillary antrum with focal destruction of the bony walls extending into the left nasal cavity and floor of the left orbit.

The histopathological report suggests the presence of numerous aseptate fungal hyphae branching at 90° with neutrophilic infiltrate invading the smaller blood vessels (PAS staining) [Figure 6].

Based on the above findings, final diagnosis of "Rhinomaxillary



**Figure 4 (a and b):** Axial sections revealing patchy opacifications along the walls of left maxillary sinus



**Figure 5 (a and b):** Coronal sections revealing opacification along the superior, medial, and lateral wall of left maxillary sinus extending into the orbit and left nasal cavity

mucormycosis—A subdivision of rhinocerebral mucormycosis” was made.

### Treatment

The following treatments were carried out:

- Surgical debridement of the necrotic tissue.
- Amphotericin B 1 mg/kg/day for 1 month.
- Nebulisation

### Discussion

#### Synonyms

- Zygomycosis
- Phycomycosis

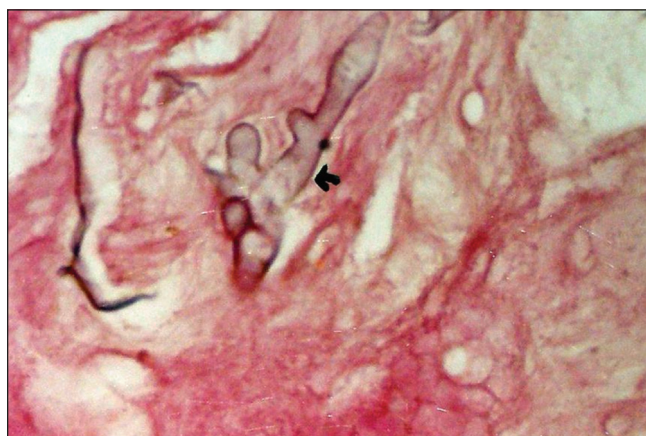
#### Definition

- It is a fatal opportunistic infection caused by a saprophytic fungus belonging to the group of Phycomycete.<sup>[1]</sup>
- Paltauf in 1885 described the first case of this uncommon infection in human beings. He coined the term mycosis mucorina which subsequently became mucormycosis.
- Disease received less recognition until 1942, when Gregory *et al.* reported three fatal cases of central nervous mucormycosis.<sup>[2]</sup>
- The disease may present with various manifestations, but there is a predilection for the paranasal sinuses.<sup>[3]</sup>

#### Organism

Organism is aerobic, but it can survive *in vitro* for 2–5 days. It is usually found in soil, bread moulds, decayed fruits, and vegetables. This organism can also be cultured from nasal cavity, throat, oral cavity, and stools of healthy patients. Organism belongs to:

- CLASS – Phycomycetes
- ORDER – Mucorales
- FAMILY – Mucoraceae
- GENERA – Rhizopus, Rhizomucor, Mucor, Absida



**Figure 6:** Histopathological picture suggests aspartate fungal hyphae branching at 90° with neutrophilic infiltrate

**Table 1: Risk factors**

Systemic factors	Local factors
Uncontrolled diabetes	Burns
Blood dyscrasias, leukemia, and neutropenia	Knife wounds
Malignancies, i.e. lymphoma.	Use of needles
Renal failure	Insect/spider bites
Protein-calorie malnutrition.	Trauma
Cirrhosis	
Organ transplants	
Patients on corticosteroid and immunosuppressive therapy	

#### Clinical variants<sup>[4]</sup>

Eisenberg *et al.* described six clinical variants:

- Rhinocerebral (Rhinomaxillary)
- Pulmonary
- Cutaneous
- Gastrointestinal
- Central nervous system
- Disseminated type

Adam in 1995 analyzed 116 cases and found:

- 39% – Rhinocerebral
- 22% – Pulmonary type
- 16% – Disseminated and cutaneous type
- 4% – GIT.
- 3% – Other locations.

Mucormycosis is the third most common invasive fungal infection, following aspergillosis and candidiasis, and accounts for 8.3–13.0% of all fungal infections found in autopsies of hematologic patients.<sup>[5]</sup>

#### Pathogenesis

Everyone is exposed to and inhales the spores; the nasal ciliary system transports these spores down in the pharynx to be cleared by the gastrointestinal tract. The spores inhaled by the lungs are cleared by the phagocytes. In the susceptible individuals [Table 1], the infection begins in the middle and inferior nasal turbinates. From the nasal cavity, the infection spreads to the paranasal sinus region and then

to the retro-orbital region *via* direct extension or either through ethmoidal, lachrymal, and angular vessels. Infection can also spread to brain through the cribriform plate and the ophthalmic vessels. Following spread, function of the cranial nerves II, III, IV, and VI may be impaired and can lead to ptosis, proptosis, pupillary dilatation, visual loss, periorbital cellulitis, and numbness.<sup>[6]</sup>

#### Pathogenic relationship between diabetes and mucormycosis

Uncontrolled diabetes associated with ketoacidosis.

↓

Abnormal polymorphonuclear chemotaxis.

↓

Impair immunity.

↓

Decreased polymorphonuclear phagocytic activity.

↓

Oelaert and colleagues found that the patients with renal dialysis and iron overload patients treated with Deferoxamine B (Fe and Al chelator) are more prone to mucormycosis.

Rhizopus is more commonly associated with diabetes because of the active ketone reductase system it can thrive well in high glucose and acidotic conditions.

#### Clinical manifestations<sup>[7]</sup>

- Age – any age group.
- Sex – both the sexes are equally affected.
- Site of invasion – Paranasal sinuses, lungs, kidneys
- Mode of spread – either through direct extension or through blood vessels/lymph vessels.
- Sites involved – Palate being the most common site
  - Buccal mucosa
  - Upper and lower lip
  - Mandible

In 1977, Eisenber *et al.* reported the first case of mandible mucormycosis.

In 1986, Brown and Finn reported the second case of mandible mucormycosis. The rhinocerebral form of mucormycosis is more common and is associated with homolateral facial palsy, anesthesia of the areas supplied by first and second division of trigeminal nerve.

#### Rhinocerebral mucormycosis is often associated with triad of symptoms

- Uncontrolled diabetes mellitus
- Periorbital infection
- Meningoencephalitis

The 25% cases of the rhinocerebral mucormycosis form shows facial gangrene (Limongelli *et al.* 1975), the disease is known to present oral manifestations.

Intraorally, ulcer with raised erythematous borders with surface of the ulcer appearing black and necrotic with areas of denudation is seen.

Other associated signs and symptoms are:

- Nasal obstruction
- Bloody nasal discharge
- Facial pain and headache
- Visual disturbances
- Facial paralysis

If there is cranial vault involvement, it can lead to blindness, lethargy, seizures or can even lead to death.

According to Moye and Cardill in 1980, black eschar formation is due to the thrombosis of blood vessels. Fungal hyphae have the affinity for blood vessels and invade the tissues. Direct penetration into blood vessels and growth of the fungus leads to thrombosis and necrosis of the adjacent tissues.<sup>[2]</sup>

#### Radiological features<sup>[8]</sup>

Nodular thickening of sinus mucosa, cloudy sinusitis without fluid levels, spotty destruction of bony walls of Paranasal sinuses were noted.

CT findings show opacification of paranasal sinuses, thickening of sinus mucosa, and erosions of the bony walls.

#### Investigations

Surface swabs

Tissue biopsy

Culture and histological examination are imperative for the correct diagnosis of mucormycosis. Culture examination from the swab specimen can be negative for mucormycosis as it tends to invade deeply in the tissues. Therefore, it is mandatory to obtain the histological specimen as well culture specimen from the necrotic wound surface.

#### Stains used

- Hematoxylin eosin stain
- Periodic acid Schiff stain
- Gridley's modification
- Gomori's methenamine silver nitrate

Culture media used are Sabour's glucose agar.<sup>[9]</sup> Sporulation of fungal hyphae species for mucormycosis. Within 24–48 h of plating spores come to the surface of petridish.

#### Histological examination

Extensive areas of tissue necrosis and the presence of numerous fungal hyphae were noted. Hyphae are irregular, ribbon shape, aseptate with non-dichotomous branching at an obtuse angle and with areas of focal dilatations.

Width of hyphae – 15–20  $\mu\text{m}$   
Length – 200  $\mu\text{m}$

Large necrotic areas with foci of suppuration, invasion of blood vessels, and their walls leading to thrombosis and tissue necrosis, and infiltration with PMN neutrophils and eosinophils.

### Differential diagnosis

- Wegener's granulomatosis
- Midline lethal granuloma
- Necrotizing sialometaplasia
- Squamous cell carcinoma
- Other deep seated mycotic infections – Aspergillosis

In the later stage if palatal perforation occurs, differential diagnosis is [Figure 7]:<sup>[10,11]</sup>

### Treatment<sup>[2]</sup>

In 1955, first cure of mucormycosis was reported by Harris.

1. Symptomatic treatment
2. Specific treatment
  - a. Medical t/t
    - Topical – Amphotericin B 50 mg of vial of AMB suspended in 10 ml of sterile water and diluted to 500 ml of glucose solution.
    - Nebulisation – 4 ml in each nostril 4–6 times/day.
    - Systemic – Hyperbaric oxygen 2 atm. For 1 h for 30 days.
    - Amphotericin B 1.0–1.5 mg/kg/day.
    - Liposomal AMB are the lipid formulations of AMB and can be prescribed safely to the patients with renal failure.
  - b. Surgical t/t
    - Caldwell Luc operation
    - External ethmoidectomy
    - Local debridement

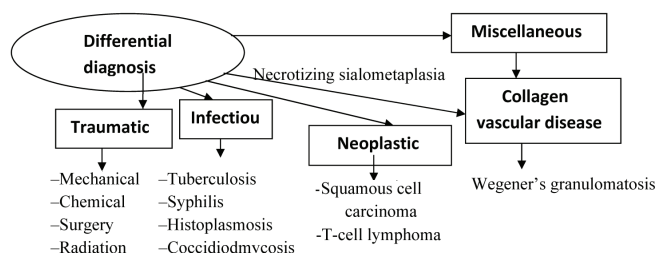
### Mortality rate<sup>[12]</sup>

Acc. To Adam *et al.*:

- 16% – Cutaneous mucormycosis.
- 67% – Rhinocerebral.
- 83% – Pulmonary.
- 100% – GIT and disseminated.

### Conclusions

A non-specific palatal ulcer could well be the presenting



**Figure 7:** Differential diagnosis of palatal perforation

sign of mucormycosis and it is therefore essential for dental practitioner be alert to early signs and symptoms of this disease, especially when evaluating the patients with the high-risk group.

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