

# Fronto-Orbito-Maxillary and Cutaneous Mucormycosis: A Case Study

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**Abstract:** *Mucormycosis is a rare infection caused by fungi of the opportunistic type. It is severe, of an acute and fast course, with a high mortality rate. We present the case of a patient who was admitted for mucormycosis of the frontal, ethmoidal, and superior maxillary sinuses superior and the orbit. The patient was unaware of suffering from diabetes mellitus, a comorbidity diagnosed in the context of mycosis. The port of entry was a leg injury caused by being rural worker. We present surgical therapeutics and established medical treatment.*

**Keywords:** Mucormycosis. Mucorales. Diabetes

## 1. Introduction

Mucormycosis is a rare fungal infection frequently associated with immunosuppression disorders, especially diabetes, chemotherapy treatments, chronic use of steroids or neutropenia, among others. <sup>[1]</sup>

It is caused by an aerobic, opportunistic saprophyte fungus of the class Zygomycetes, of the order Mucorales (Mucor, Rhizopus, Absidia and Rhizomucor), which produces a condition of acute course, with tissue destruction and vascular invasion. Spores enter the body through continuity solutions, especially cutaneously, being able to produce local necrotic lesions, through respiratory tract, implanting in the oral mucosa and other routes, such as the conjunctival or catheters. <sup>[1][2][3][4]</sup>

Spores are commonly found in the soil and moist soils with high nitrogen content, decomposing matter, vegetable waste, manure, etc. <sup>[3]</sup>

Once the fungus enters the body, it invades the elastic lamina of the arteries, veins and lymphatics, causing thrombosis, with consequent ischemia of the tissues. This impairs immunity by altering the macrophage effectiveness. <sup>[3][4]</sup>

Different clinical forms are described, the rhino-cerebral being the most common, followed by pulmonary, cutaneous, gastrointestinal presentations, and systemic by hematogenous dissemination. <sup>[3][5]</sup>

The prognosis is ominous with high, short term mortality. The mortality rate ranges from 20 to 70%. <sup>[1][3][4][6][7]</sup> Generally not pathogenic for a immunocompetent host, this being the determinant in evolution. <sup>[6]</sup>

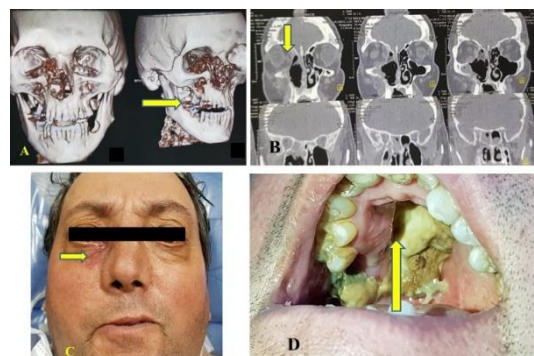
## 2. Clinical Case

43-year-old rural worker from the province of La Pampa. Consulted for upper airways symptoms, asthenia and

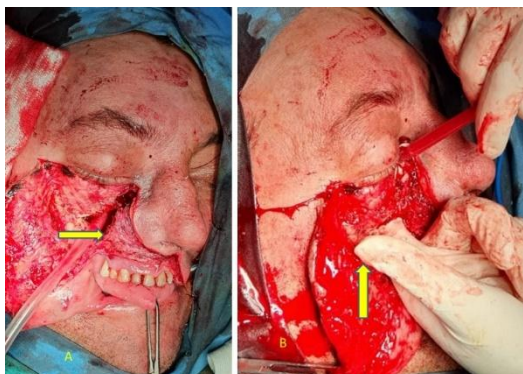
general malaise; a CT scan was requested, which revealed soft tissue formation that compromised the right maxillary sinus, projecting to the nostril. During hospitalization, he was diagnosed with diabetes mellitus, with blood glucose levels greater than 300mg/dl. A culture is made that reflects Mucormycosis so it is referred to our medical center. The port of entry had been a cutaneous lesion on the right ankle that developed a 2 cm necrotic eschar, that was circumscribed without local progression, healing in 2 weeks, that also was positive for mucor.

The patient was admitted with lagophthalmos and compromised motility of right eye, with normal vision. CT scan demonstrated cortical involvement of the right maxillary sinus, destruction of the turbinates, compromise of orbital floor and retro ocular fat, extension to the pterygoid fossa and frontal and sphenoid sinus occupation (Photo 1).

The pre-surgery evaluation was requested and the patient was operated under general anesthesia through a Weber Fergusson incision. Complete destruction of the superior maxillary sinus, hard palate, orbital floor, and lateral wall of the nostril was found. (Photo 1). We proceeded to complete resection down to the pterygopalatine fossa. Material was extracted and again, grew mucormycosis. (Photo 2)



**Photo 1:** A. Osteolysis of the upper jaw, external wall of nose. B. Osteolysis of the orbital floor and ethmoids. Occupation of sinuses. C. Compromise of the orbital floor and skin. D. Destruction of the hard palate



**Photo 2:** A. Weber Ferguson incision. B. Lesion of superior maxillary sinus and orbital floor

A catheter was left for local instillation of Amphotericin B (50 mg every 8 hours), also a nasojejunal tube for enteral nutrition and central line for parenteral Amphotericin B application (1.5 mg / kg / day). (Photo 3)

After 72 hours, he suffered a seizure episode with a subsequent cardiorespiratory arrest.

### 3. Discussion

Kurchenmeister presented the first case of mucormycosis in a patient with lung cancer, then, in 1885, Paltauf created the term Mucormycosis and describes the first Rhinosinusal case.<sup>[3] [8]</sup> Usually 2 forms are recognized, a primary cutaneous form, and a secondary to metastatic locations (rhinocerebral, pulmonary or intestinal).<sup>[6]</sup> Our patient presented with an initial cutaneous lesion in the ankle area. Mucormycosis is usually unilateral, rapidly spreading to contiguous tissues, and a differential diagnosis of Midline Granuloma, Syphilis, Tuberculosis, Rhinosinusal tumors should be considered.<sup>[9] [10]</sup> Compromise of the ethmoidal sinus is observed between 60 and 100% of cases.<sup>[11]</sup>



**Photo 3:** Immediate post-operative with catheter for local instillation

Diabetes is the most common predisposing factor and has a preponderant role in rhino-orbit-cerebral mucormycosis. Diabetes patients show a decreased phagocytic function of neutrophils and of its adherence to the endothelial wall.<sup>[10]</sup> Due to the high prevalence of diabetes, the number of patients at risk for this infection is high. Approximately half of the patients present with diabetic ketoacidosis.<sup>[7] [12]</sup>

Several authors cite the pterygopalatine fossa as the main reservoir of mucor and from there it spreads to other places.<sup>[8] [13]</sup> Cavernous sinus thrombosis is a frequent complication.<sup>[14]</sup>

In the immunocompromised patient, macrophages prevent infection through phagocytosis and oxidative processes that allow it to eliminate spores; when this function is altered, the spores that are not eliminated transform into hyphae and produce the tissue invasion. Hyperglycemia and acidosis decrease macrophage chemotaxis.<sup>[1]</sup>

Necrosis would have its explanation in thrombosis and venous arteries leading to tissue injury.<sup>[4]</sup> The ischemic necrosis and thrombosis generate low flow blood, which favors fungal proliferation. For this reason, some authors suggest the use of therapy local antifungal as an adjunct to systemic therapy.<sup>[14]</sup>

The bibliography consulted agrees that the gold standard of treatment is early surgery for an extensive debridement, associated with intravenous antifungal therapy.<sup>[6]</sup> Orbital exenteration is disputed; most authors recommend it in cases of great ocular involvement, blindness and ophthalmoplegia. Other therapeutic alternatives presented consist of the use hyperbaric chamber, which reduces tissue hypoxia and acidosis that accompanies tissue invasion by the fungus.<sup>[15]</sup> The diagnosis of certainty is provided by pathology and mycological study.<sup>[6]</sup>

### 4. Conclusion

Mucormycosis, although rare, is considered an emergency due to rapid and aggressive progression of this microorganism and its high mortality. It must be suspected in every immunocompromised patient with skin lesions, especially necrotic or ophthalmologic conditions of rapid appearance and progression. The culture for the diagnosis, and initiation of immediate therapy is the only way to improve survival rate.

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