Rhinomaxillary Mucormycosis: An Updated Overview with Emphasis on the Histological Findings

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Abstract
Rhinomaxillary Mucormycosis has been overlooked, for years, in the differential diagnoses of rapidly devastating lesions. This fungal infection, if not early diagnosed, is fatal. Therefore, rapt attention should be paid towards comprehending Rhinomaxillary mucormycosis.

Keywords: Mucormycosis; Zygomycosis; Rhinomaxillary; Aspergillosis; Fungal; Rhinocerebral

Introduction
In 1885, Paltauf documented the first histologic description of generalized mucormycosis in a 52-year-old patient. Since then, opportunistic infections caused by fungi of the order Mucorales have been recognized in association with diabetes, hematologic malignant diseases, immunosuppressive therapy, thermal burns and surgery [1]. Mucorales of the Class Zygomycetes, fungal organisms, cause rhino-cerebral, pulmonary, gastrointestinal, cutaneous or disseminated infection in immune compromised individuals with various clinical manifestations and morbidities. The rhino cerebral clinical form is the commonest and is subdivided into (i) highly fatal rhino-orbito–cerebral form; and (ii) Rhinomaxillary form. The former form is invasive and may involve ophthalmic and internal carotid arteries, while the rhino maxillary form is seen most commonly in individuals with uncontrolled diabetes mellitus. It is imperative to note that rhino maxillary mucormycosis can progress to the rhino-orbito–cerebral form [2,3]. Ubiquitous etiologies of RM exist in the environment. Possible infections occur following inhalation, ingestion or implantation of spores. However, no scomial outbreaks of mucormycosis are not as common as hospital-related Aspergillus contagions, but have sometimes been linked to construction or renovation work, as well as to contaminated ventilation systems [4,5].

Overview
Rhino maxillary mucormycosis (RM) involves sphenopalatine and greater palatine arteries resulting in thrombosis of turbinates and necrosis of the palate [1,3]. Oral manifestations of mucormycosis are frequently the first clinical signs to arise especially after dental extractions. Intra orally, the hard palate is usually affected because of its proximity to the infection of the nasal fossa and paranasal sinuses. Radio graphically, opacification of sinuses may be observed in conjunction with patchy effacement of bony walls of sinuses [6].

Culture examination, on sabouraud's dextrose or glucose agar, from the swab specimen and histological examination from the necrotic wound surface are imperative for the correct diagnosis of RM. Even if they came falsely negative, histological examination can rule out malignancies. Other than the conventional Hematoxylin eosin stain, special stains are also used including Periodic acid Schiff stain (PAS), Gridley's modification and Gomori's methenamine silver nitrate [7].
Common risk factors for mucormycosis include patients with uncontrolled diabetes mellitus, especially with ketoacidosis, are at high risk. Patients with cancer especially those who are neutropenic and receiving broad-spectrum antibiotics as well as individuals receiving immunosuppressive agents including high-dose oral or intravenous steroids and tumor necrosis factor (TNF) alpha blockers are at risk. Blood dyscrasias, lymphoma, prolonged neutropenia, and leukemia [2,6].

Other than the effacement of the normal architecture of the palatal mucosa, characteristic to the histologic picture of fungal and algal infections is the presence of extensive angioinvasion associated with mycotic thrombosis, and angiotropic ischemic necrosis. This necrosis is usually centered on endothelial vessels harboring fungal organisms. There, numerous non-septate, broad, branching hyphae, positive for PAS, are seen within the necrotic marrow tissue. Within this picturesque, mucormycosis and aspergillosis are similar. However, aspergilliosis has septa branching hyphae that can be distinguished from mucormycotic hyphae by a smaller width and prominent acute angulations of branching hyphae. Infiltration with PMN neutrophils and eosinophils are also evident [6,11].

It is also important to differentiate fungal infections from ulcerative granulomatous lesions. The latter group reveals giant cells in small granulomatas as well as a caseous necrosis. The granulomatous tissue appears cheesy. The only confusion between the above mentioned groups is observed in cases of actinomycosis. To differentiate, other than the characteristic "sulfur granules around pus areas", the actinomycotic giant cell is very irregular in shape and size and has varying numbers of scattered nuclei. Unlike tuberculosis and sarcoidosis, the actinomycotic giant cell does not recapitulate a specific appearance. In TB, giant cells always show a horse shoe appearance while sarcoidosis reveals Schumann bodies along with Langhans-type multinucleated giant cells [12].

Differential diagnosis of RM, as a fulminating palatal lesion, should include squamous cell carcinoma, nasopharyngeal carcinoma, peripheral T-cell lymphoma, chronic granulomatous diseases, other deep fungal infection and necrotizing sialometaplasia. Treatment should combine surgery and antifungal therapy [13]. Recently, molecular biological markers, including semi nested PCR targeting the 18S rDNA of Mucorales and Phenotypic methods based on carbohydrate assimilation profiles are claimed to be of prime importance. However, further studies are required to validate these new approaches [14].

**Conclusion**

Cataclysmic complications could endanger immunocompromised patients if fungal infections were not diagnosed early. Dental professionals and otolaryngologist must be mindful to investigate the medical history and to follow up susceptible patients adequately.

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