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Case Report *Medical research*

Rhinocerebral mucormycosis: Report of a rare case

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ABSTRACT

Rhinocerebral mucormycosis is a life threatening fungal infection occurring in humans, which is caused by the ubiquitous saprophytic fungi of order Mucorales. A timely diagnosis in patients with predisposing factors leading to immunosuppression is of great importance in reducing mortality and morbidity. We describe a patient presenting with typical clinical manifestations of rhinocerebral mucormycosis involving the paranasal sinuses and the orbit.

Keywords: *Mucormycosis, Fungal Infection, Rhizopus, Amphotericin B, Surgical Debridement*

INTRODUCTION

Mucormycosis is an opportunistic and frequently fulminating fungal infection caused by members of the family Mucoraceae, order Mucorales and class Zygomycetes [1]. These are ubiquitous fungi surviving on decaying these are ubiquitous fungi surviving on decaying vegetation and diverse organic matter [2]. Depending on the immunological status of the patient and the site of the body that is affected, the disease may manifest in six different ways as rhinocerebral, pulmonary, cutaneous, gastrointestinal, central nervous system or disseminated forms [3]. Rhinocerebral mucormycosis is the most common type accounting for 30% to 50% of the cases and its extension to the orbit and brain is quite usual making it potentially life-threatening disease [3]. It is commonly reported in immunocompromised patients such as

poorly controlled diabetes mellitus, blood dyscrasias, malnutrition, neutropenia, iron overload, organ transplant, and immunosuppressive therapy [4]. Diagnosis is confirmed by histopathological demonstration of the organism in the affected tissue [4]. Early diagnosis and treatment of mucormycosis is extremely important due to the aggressive course of the disease3. Control of underlying disease need to be established, metabolic abnormalities corrected and antifungal therapy should be combined with surgical debridement of all necrotic tissues [1-4].

The aim of this case report is to present a patient with rhinocerebral mucormycosis in order to draw attention to its existence in our environment and to emphasize the need for high index of suspicion.

CASE REPORT

A 40 year old female with poor glycaemic control presented to a medical ward with a history of swelling of left eye associated with diminution of vision on ipsilateral side for last 1 week, associated history of swelling, weakness and numbness of left side of the face for 10 days with drooping and difficulty in moving the left eye for 4 days. She was a non-compliant type II diabetes mellitus patient for 10 years. Examination revealed echar on apex of nose with absent extraocular movement on left eye (image 1). A diagnosis orbital apex syndrome was made and contast CT planned which revealed medial wall orbital perforation with no evidence of cavernous sinus thrombosis, MRI Brain revealed inflammation of left extraocular muscles, left optic neuritis and focal cerebritis of left frontal lobe (Image 2, 3), A rigid nasal endoscopy (RNE) revealed fungal debris and pus in the left maxillary region and polypoidal lesions lateral to the maxillary turbinate. A biopsy of the latter was sent for histology. Histology report suggested a fungal infection, possibly mucormycosis (Fig 1, 2).

Based on the clinical presentation and histopathology, intra-venous amphotericin-B (conventional type) was started, and she responded to antifungal and discharged uneventfully. A final diagnosis of Rhinocerebral Mucormycosis with poor glycaemic control was made.

Image 1: Patient with black eschar on nose with left orbital cellulitis with left sided fix pupillary movement.

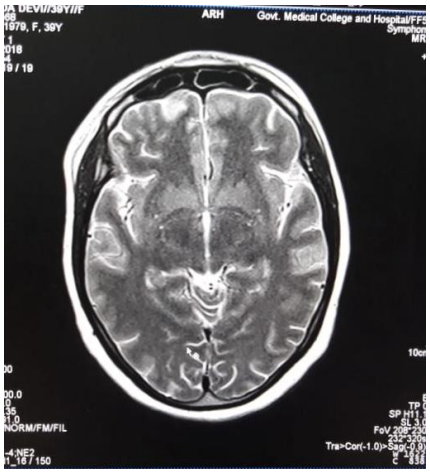

Image 2 : MRI Brain : left frontal lobe focal cerebritis.

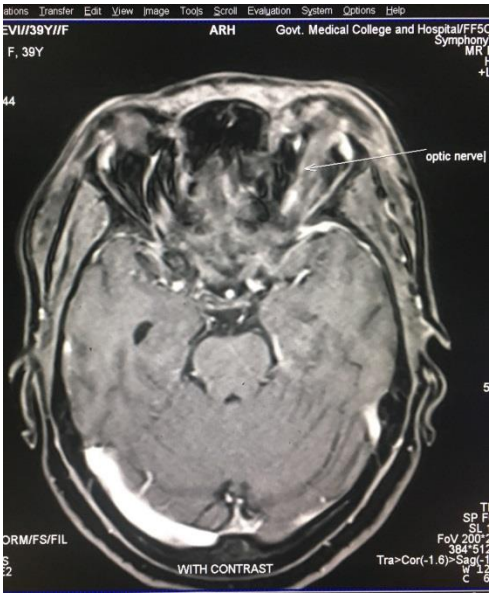

Image 3: Post contrast enhancement seen in left extraocular muscles with patchy enhancement of left optic nerve suggestive of left optic neuritis.

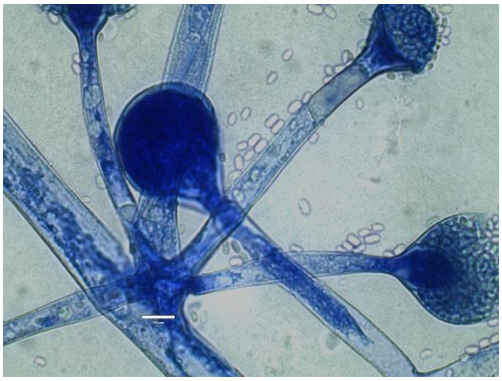

FIG :2 Microscopic features leading to identification of *Rhizopus* spp. (x10magnification)

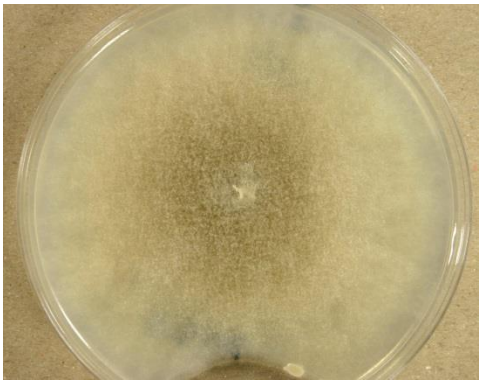

FIG: 2 Macroscopic appearance as seen in a SDA plate.

DISCUSSION

Mucormycosis is a serious, relatively uncommon invasive fungal infection and one of the most aggressive and lethal invasive mycoses [5]. The predisposing factors for mucormycosis are uncontrolled diabetes (particularly in patients having ketoacidosis), malignancies such as lymphomas and leukemias, renal failure, organ transplant, immunosuppressive therapy, cirrhosis, and AIDS [6]. Mucorales species are vasotropic,

causing tissue infarctions, and the mucormycosis spectrum ranges from cutaneous, rhinocerebral, and sinopulmonary to disseminated and frequently fatal infections, especially in immune-compromised hosts [7]. Reports have suggested that the ability of serum of immuno-compromised patients to inhibit Rhizopus invitro is reduced, which makes them suitable hosts to opportunistic fungal infections [8]. In diabetic patients, especially with elevated blood sugar levels, the spores germinate, hypae develop, fungi begin an inexorable march through the tissues as blood vessels become involved, thrombosis occurs, resulting in tissue necrosis and fungi continue to grow in this devitalised tissue causing further damage to surrounding tissues [9]. The presence of chronic diabetes in our patient too led to necrosis of palate. Rhinocerebral mucormycosis (ROCM) is the most common form of mucormycosis in patients with diabetes mellitus. The infection develops after inhalation of fungal sporangiospores into the paranasal sinuses. Upon germination, the invading fungus may spread inferiorly to invade the palate, posteriorly to invade the sphenoid sinus, laterally into the cavernous sinus to involve the orbits or cranially to invade the brain which can prove fatal [10]. Our patient had uncontrolled diabetes which is a well-known predisposing factor for mucormycosis along with spread of lesion from the palate into adjacent tissues. The initial symptoms of ROCM are consistent with those of sinusitis and periorbital cellulitis and include eye and/or facial pain and facial numbness followed by blurry vision [5, 10]. Signs and symptoms that suggest mucormycosis in susceptible individuals include, unilateral periorbital facial pain, facial cellulitis, orbital inflammation, eyelid edema, proptosis, acute ocular motility changes, nasal discharge, nasal stuffiness headacheand acute vision loss [5]. If blood supply to eye is affected by invasion of retinal artery, blindness develops. A black necrotic eschar is the hallmark of mucormycosis [12, 13].

Among the differential diagnosis, rhinocerebral mucormycosis may initially resemble bacterial sinusitis and may mimic malignancy. Rhinocerebral mucormycosis may be confused with allergic fungal sinusitis, which is caused by phaeohyphomycoses in individuals with histories of allergic rhinitis, elevated immunoglobulin E levels, nasal polyps and recurrent or chronic sinusitis. Allergic fungal sinusitis slowly progresses over months to years, although it causes proptosis and a large rhinocerebral mass, it does not invade tissue or meninges. Aspergillosis can cause a similar disease, with CNS invasion, and carries a poor prognosis. An important difference is that itraconazole may play a role in treatment. Histologic staining can differentiate between the fungi [15]. The pleiotropic clinical manifestations with other lesions were ruled out with radiographic and histopathology report. A definitive diagnosis of mucormycosis can be made by tissue biopsy that identifies the characteristic hyphae. In order to differentiate histopathologically from other types of lesions such as aspergillosis, which shows presence of narrow, septae hypae with narrow angle branching was done. In this case, the fungus was identified by hematoxylin and eosin stain.

Treatments of mucormycosis need to be fast and aggressive because by the time even the presumptive diagnosis is made, often the patient has suffered significant tissue damage that cannot be reversed. Most patients require both surgical and medical treatments. The line of treatment for such cases requires aggressive surgical debridement of the infected area; otherwise, the patient is likely to die. Medications also plays an important role. Two main goals are sought at the same time: antifungal medications to slow or halt fungal spread and medications to treat any debilitating underlying diseases. Amphotericin B (initially intravenous) is the usual drug of choice for antifungal treatment. Patients with underlying diseases like diabetes need their diabetes optimally controlled. Patients may need additional surgeries and usually need antifungal therapy for an extended time period (weeks to months) depending on the severity of the disease [15]. The prognosis of mucormycosis is usually fair to poor; the prognosis depends on the overall health of the patient, the speed of diagnosis and treatment, the patient's ability to respond to treatments and the complete debridement of the infected body area. In this patient, the line of treatment advised was aggressive surgical debridement of the infected area along with administration of amphotericin B as it is the drug of choice in treatment of mucormycotic infection [2, 6, 11]. The patient was under observation for his ability to respond to treatment due to old age and overall health of patient.

Mucormycosis is a serious, relatively uncommon invasive fungal infection. Early

diagnosis is essential because by the time even the presumptive diagnosis is made, often the patient has suffered significant tissue damage that cannot be reversed. The pleiotropic clinical manifestations and elusive presentation of mucormycosis often delay diagnosis, with resultant poor outcomes. Timely diagnosis is critical to survival and minimization of morbidity. Institution of surgical and medical therapy is critical in maximizing the likelihood of good outcome.

Conflict of Interests

The authors declare that they have no conflict of interests.

Authors' Contribution

The final paper has been seen and approved by all authors. The authors accept full responsibility for the design and conduct of the study, had access to the data, and controlled the decision to publish.

Disclosure

The authors also report the absence of any significant financial support in any organization. The paper had not been published elsewhere previously.

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