Rhino-orbito-cerebral Mucormycosis in an Immunocompetent Patient: Case Report and Review of Literature

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ABSTRACT

Rhino-orbito-cerebral mucormycosis is a fungal infection that can be fatal especially in immunocompromised patients. It is extremely rare in immunocompetent individuals. We describe here an immunocompetent patient who survived rhino-orbito-cerebral mucormycosis due to *Saksenaea vasiformis*, and provide a literature review of this rare entity.

Key words: Immunocompetent, Saksenaea vasiformis, Rhino-Orbito-Cerebral Mucormycosis

INTRODUCTION

Mucormycosis is a rare but aggressive opportunistic fungal infection that is commonly caused by members of the family *Mucoraceae* that include *Rhizopus*, *Rhizomucor*, *Mucor* and *Absidia*. Mucormycosis can cause severe, sometimes fatal disease in susceptible individuals with uncontrolled diabetic ketoacidosis, neutropenia, chronic glucocorticoid use, hematological malignancy, chronic malnutrition and burn patients.

Rhino-orbito-cerebral mucormycosis (ROCM) is an uncommon infection in immunocompetent hosts. ROCM invades vessels, soft tissue, nerves, bone and cartilage producing tissue infarction and ultimately leading to tissue necrosis and vessel thrombosis. We present a case of an immunocompetent patient who survived ROCM due to *Saksenaea vasiformis*, and review the literature of this fatal condition.

CASE REPORT

A 40-year-old healthy male presented with painful swelling in the left eye for two weeks. Other symptoms included blurring of vision and diplopia. There was no history of eye trauma, dental carries, recent surgery, sinusitis or skin infection. The visual acuity was 6/60 in the left eye with 2-mm axial proptosis, with conjunctival congestion and limited ocular motility. The pupils were reactive bilaterally, with no evidence of a relative afferent pupillary defect in either eye.

The patient was diagnosed with orbital cellulitis in the left eye. However, the primary cause was undetermined. Broad spectrum antibiotic therapy was initiated that included intravenous cloxacillin 500 mg every 6 h, cefuroxime 750 mg every 8 h and metronidazole 500 mg every 8 h. There was minimal clinical improvement with the above regime after one week of hospitalization. Thus, the patient elected to discharge himself against the medical advice.
The patient presented two weeks later to a tertiary hospital with severe pain and persistent purulent discharge and no light perception in the left eye. The left globe perforated with extensive regions of necrotic tissue. Both upper and lower lids were swollen with discharging fistula proximal to the lateral canthus [Figure 1]. The right eye was normal with 6/6 visual acuity. General examination revealed an alert, afebrile and well-oriented patient with no neurologic deficit.

Plasma glucose, blood urea, full blood count and liver function test were normal. HIV serology was non-reactive. Computed tomography (CT) of the brain and orbit showed destruction of the left eye, with an ill-defined mass and ring enhancement involving left pterygopalatine, inferotemporal fossa and adjacent bony destruction. It extended posteriorly to the orbital apex, causing a widening of the optic nerve canal and the superior orbital fissure. The cavernous sinus showed signs of thrombosis and presence of areas of infiltrate at the left maxillary, frontal and sphenoidal sinuses [Figure 2].

A presumptive clinical diagnosis of ROCM was made. Intravenous amphotericin B 1.0 mg/kg/day was initiated after a test dose. The patient had no significant side effects due to intravenous Amphotericin B. The patient consented to exenteration of left eye and extensive sinus debridement surgery. Histopathology revealed broad, aseptate, long and right angled branching hyphae consistent with mucormycosis [Figure 3]. A tissue culture grew *Sakasandra vasiformis*. The intravenous Amphotericin B stopped after 4 weeks of therapy and he was discharged with instructions to instill topical Amphotericin B for another four weeks.

Postoperatively, the patient refused to repeat imaging and implantation of eye prosthesis due to poor family support and financial constraints. To date, the patient has been followed up for two and a half years with recurrence of ROCM.

**DISCUSSION**

ROCM is usually associated with a fatal outcome especially in immunocompromised patients. For the last decade, ROCM has been increasingly reported in healthy individuals. We reviewed 11 cases of ROCM in immunocompetent hosts published in PubMed from 2000 to 2011. Table 1 summarizes age, gender, presentations, risk factor, causative organism and final outcome.

The presenting age ranged between 16 and 59 years and both genders were equally affected. Nine out of 11 published cases were due to *Apophysomyces elegans*. There was only one reported case of ROCM caused by *Sakasandra vasiformis*. *Apophysomyces elegans* is an emerging pathogen that causes ROCM via inhalation. Whereas, *Sakasandra vasiformis* is a soil saprophytic fungus and the only species in the genus *Sakasandra*.

In the previously reported cases, 7 immunocompetent patients who survived were infected by *Apophysomyces elegans* and one with *Sakasandra vasiformis*. Two of these three
Table 1: Summary of previous literature on Rhino-orbito-cerebral mucormycosis in immunocompetent patients published in PubMed from 2000 to 2011

<table>
<thead>
<tr>
<th>Case reports</th>
<th>Age/gender</th>
<th>Presentations</th>
<th>Risk factor</th>
<th>Organism isolated</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fairley C et al. (2000)¹</td>
<td>59/Male</td>
<td>Deep and sharp retro-orbital pain and fever following high-pressure water jet injury to right inner canthus while cleaning air conditioner filter</td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, radical debridement and intravenous liposomal Amphotericin B</td>
</tr>
<tr>
<td>García-Covarrubias et al. (2000)²</td>
<td>24/Male</td>
<td>Closed-head trauma with Fort II complex facial fracture after motorcycle collision</td>
<td></td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, multiple debridement, hyperbaric oxygen treatment, intravenous liposomal Amphotericin B, Tobramycin, Penicillin</td>
</tr>
<tr>
<td>Chakrabarti et al. (2003)³</td>
<td>20/Female</td>
<td>Pain, redness, swelling, and protrusion of right eye, rhinitis, headache, fever</td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after local debridement, orbital exenteration and intravenous Amphotericin B</td>
</tr>
<tr>
<td>Rao SS et al. (2005)⁴</td>
<td>45/Male</td>
<td>Unilateral facial pain, proptosis, nasal discharge, nasal obstruction, and vision loss</td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, debridement and intravenous Amphotericin B</td>
</tr>
<tr>
<td></td>
<td>26/Female</td>
<td></td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, debridement and intravenous Amphotericin B</td>
</tr>
<tr>
<td></td>
<td>26/Male</td>
<td></td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, debridement and intravenous Amphotericin B</td>
</tr>
<tr>
<td></td>
<td>16/Male</td>
<td></td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after orbital exenteration, debridement and intravenous Amphotericin B</td>
</tr>
<tr>
<td></td>
<td>55/Male</td>
<td></td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Died</td>
</tr>
<tr>
<td>Schütz P et al. (2006)⁵</td>
<td>31/Male</td>
<td>Right painful proptosis eye with progressive visual loss</td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Died</td>
</tr>
<tr>
<td></td>
<td>18/Female</td>
<td>Proptosis of left eye</td>
<td>None</td>
<td>Apophysomyces elegans</td>
<td>Survived after extensive surgical debridement with removal of left eye was done</td>
</tr>
<tr>
<td></td>
<td>56/Female</td>
<td>Nasal blockage, nasal bleeding and headache for one month</td>
<td>None</td>
<td>Saksenaea vasiformis</td>
<td>Died due to cerebral mucormycosis</td>
</tr>
</tbody>
</table>

patients were above 50 years of age and died due to extensive brain involvement and severe cerebral vascular insult.⁶,¹¹ This finding is similar to the findings of Hargrove et al.¹² that patients older than 46 years with concurrent frontal sinus involvement and fever were less likely to survive orbital mucormycosis.

Our patient was very fortunate to survive this fatal condition despite delayed diagnosis and treatment. This is likely due to his relatively young age and the lack of a pre-existing chronic medical illness. To the best of our knowledge, this is the first reported case of an immunocompetent patient with ROCM due to Saksenaea vasiformis who survived.

Our patient and other immunocompetent patients who survived ROCM were treated with a combination of systemic Amphotericin B, orbital exenteration and adjacent tissue debridement.¹ ³,⁶ This is consistent with Roden et al.,¹ who reported a survival rate of 70% for cases treated with antifungal and surgery. In contrast, only 61% of cases treated with amphotericin B deoxycholate only, 57% of cases that underwent only surgery and only 3% of untreated patients survived.¹

In conclusion, diagnosis of ROCM in immunocompetent patients is always misleading and possibly causes delay in treatment. It is essential to alert the managing ophthalmologist of the emergence of this rare disease among healthy individuals.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.