Case Report

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Rhino-orbital Mucormycosis in an Immunocompetent Pediatric Patient, Resembling an Orbital Mass- A Case Report

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Running Title Rhino-orbital Mucormycosis, Resembling an Orbital Mass

ABSTRACT

Rhino-orbital mucormycosis in an immunocompetent pediatric patient can present as an orbital mass. We report a 9-year-old male that presented with periorbital swelling and limitation of left eye movement from one month ago. The patient was treated at another center with a diagnosis of mucormycosis but was referred due to worsening symptoms. Orbital and paranasal sinus CT scan revealed opacities in the left paranasal sinus and soft tissue density in the medial and inferior orbital wall. The patient underwent orbitotomy and mass debulking surgery on suspicion of a possible neoplastic mass. Pathologic evaluations revealed mucormycosis. After receiving intravenous liposomal amphotericin-B that was followed by oral posaconazole syrup for two months and sinus debridement, the symptoms regressed. In immunocompromised pediatric patients, mucormycosis should be considered in the differential diagnosis of an orbital mass.

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Introduction

Infections caused by the omnipresent fungi from the order of Mucorales are generally called Mucormycosis. Most of these infections are caused by the genus Rhizopus and Mucor.1, 2 Mucorales invade arteries that results in damage to the endothelium and intravascular thrombosis which, in turn, threatens blood supply to the involved tissues.3 These infections are likely to have different presentations. One of these conditions which is the most prevalent one is rhino-orbital mucormycosis.4 These rapid and progressive infections are expected to happen in individuals with underlying diseases. These comorbid illnesses include uncontrolled diabetes mellitus, diabetic ketoacidosis, hematologic malignancies, organ transplantation, and immunosuppression due to certain medications.5 Although rare, there are reports of mucormycosis in immunocompetent patients.6 Here we report a case of rhino-orbital mucormycosis in an immunocompetent patient. To our knowledge, this is the first case of rhino-orbital mucormycosis in an immunocompetent host being reported in Iran.

Case Presentation

A 9-year-old male was referred to "……." with swelling and limitation of movement of the left eye from one month ago. The previous history of any external periorbital trauma or a medical condition such as sinusitis was negative. No history of receiving oral or parenteral antibiotics and other medication over the past 6 months was mentioned before the onset of symptoms. There was no history of any chronic inflammatory and infectious diseases in the family. On examinations, the left eye was proptotic. The pupillary response was normal and relative afferent pupillary defect (RAPD) was negative. Visual acuity of the left eyes was minimally decreased, possibly due to ocular surface problems. Ocular movements of the left eye were limited mostly in adduction and elevation (Figure 1).

This patient had been admitted to another center after onset of symptoms. Primary evaluation with orbital and paranasal sinuses (PNS) CT scan had revealed the possible diagnosis of orbital cellulitis in combination with paranasal sinusitis. Based on this diagnosis, after unresponsiveness to primary medication, the patient took intravenous antibiotics therapy with sufficient doses of clindamycin and ceftriaxone. After a poor response to the mentioned therapeutics, the patient underwent excisional biopsy of the lesion and also functional endoscopic sinus surgery (ethmoidectomy and sphenoidectomy). According to biopsy results that had confirmed granulomatous reaction with fungal infection (non-septate hyphae) the patient had taken sufficient doses of intravenous liposomal amphotericin-B in addition to vancomycin and ceftazidime in that medical center.

After the patient was referred to our clinic, orbital and paranasal sinuses (PNS) CT scan displayed opacities in the left paranasal sinuses (maxillary, ethmoid, and sphenoid) and soft tissue density in the medial and inferior orbital wall (Figure 2). We assumed that the lesion could be possibly a neoplastic mass. Therefore, the patient was planned to undergo orbitotomy and mass debulking surgery. Surprisingly, the primary diagnosis was confirmed as the microbiological and pathologic evaluations revealed mucormycosis (Figure 3). After the diagnosis was confirmed, complete investigations had been launched to discover any possible underlying medical condition. Complete blood count (CBC), PPD (confirmed with interferon-gamma release assay), HIV tests, HbA1C, Dihydrorhodamine (DHR) Flow cytometric Test, Immunoglobulin and complement levels, and other molecular and genetic laboratory tests were within the normal limits and any possible congenital and acquired immunodeficiency conditions were ruled out. Finally, after debridement of the lesion with the patients receiving intravenous liposomal amphotericin-B followed by oral posaconazole syrup for two months, the symptoms regressed (Figure 1). Then, the patient received a prophylactic dose of itraconazole for one year.

Parental consent was obtained for the preparation of this work.

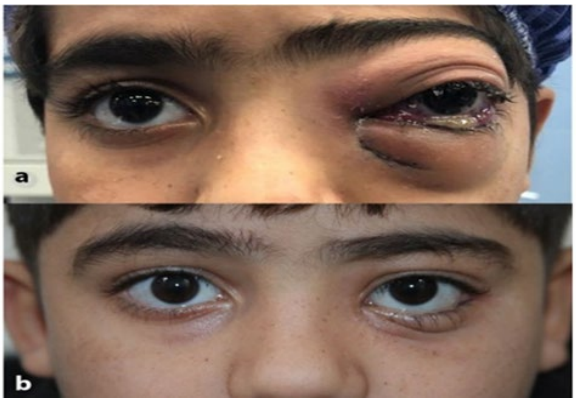

Figure 1. Periorbital swelling and erythema and dystopia with chemosis and conjunctival discharge. Note the sutured incision from previous biopsy on the lower eyelid (a). 3 weeks after surgery with patient still on the antifungal medication (b).

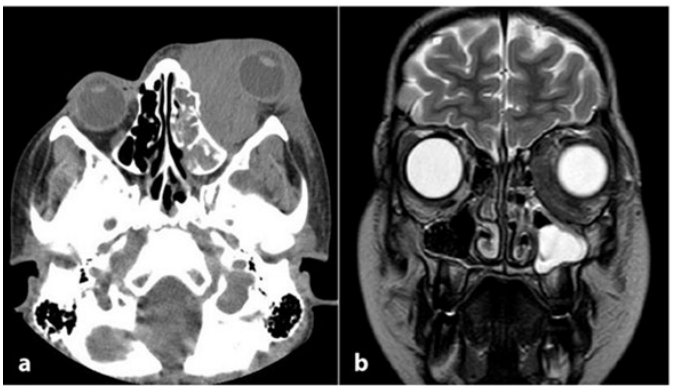

Figure 2. Axial orbital and paranasal sinus CT scan. Note involvement of ethmoidal and sphenoidal sinuses and orbital mass-like lesion (a). Coronal head MRI (T2). Note mucosal thickening and air-fluid level in the maxillary sinus (b). Images were not taken synchronously.

Discussion

Mucormycosis is a rapid life-threatening infection caused by fungi from the order of Mucorales. Ubiquitous spores of these fungi are found commonly in bread, air, soil, dust, plants, and animal excreta.2 These spores when inhaled are expected to be phagocytosed by neutrophils and macrophages.7 However, the inhalation of these spores puts the paranasal sinuses and adjacent tissues in danger of infections by these fungi when fungicidal activity is impaired by acidosis and immunosuppression.8, 9 These conditions are often observed in association with other comorbidities. The most common predisposing diseases are diabetes mellitus, hematological malignancies, and solid organ transplantation.10 There are reports of different types of these infections in immunocompetent patients. Lee et al. reported that 12.6% (11 out of 87 cases) of their study subjects had no underlying disease.11 Ayoadea et al. reported a similar case of rhino-orbital mucormycosis in a 67-year-old immunocompetent host following Hurricane Irma.12 Although our patient lived in one of the south-eastern states of our country which is highly prone to disastrous floods, as Ayoadea et al. mentioned in their report, it's difficult to prove the correlation. As the mucosal epithelium functions as a barrier against tissue invasion of this mold, trauma, prior infections, and cytotoxic damage to the nasal epithelium seem to be related to rhino-orbital mucormycosis in healthy hosts.13 The diagnosis of mucormycosis in an immunocompetent patient is challenging because in absence of any predisposing disease, clinical suspicion is low. We encountered the same challenge during management of our case. Although primary histopathologic evaluations reported fungal elements compatible with mucormycosis, clinical suspicion of a possible neoplastic scenario was still strong and this in addition to poor response to liposomal amphotericin B made us decide to plan for orbital decompression of necrotic tissues.

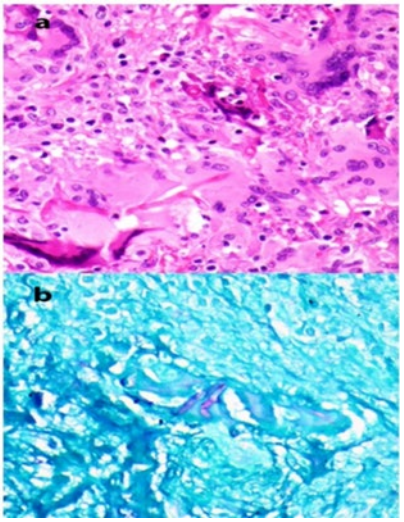

Figure 3. Hematoxylin and Eosin stains of a histologic section showing diffuse mixed acute and chronic necrotizing granulomatous inflammation with marked eosinophilic infiltration and a few fungal elements (non-septate mycelium). Mag × 400 (a). Periodic Acid–Schiff (PAS) stain (light green) of a histologic section showing fungal non-septate mycelium. Mag ×400 (b).

Our patient was treated primarily with liposomal amphotericin B. As the patient situation worsened under this treatment, the patient underwent surgical debridement. After surgery and under treatment with liposomal amphotericin B, patient symptoms started to regress. Although immunocompetent patients with mucormycosis can be successfully managed with medical treatments,9 our patient didn't respond to medical treatment alone. After significant regression, we continued the treatment with posaconazole which has been verified as an alternative therapeutic option for such patients.14

Our case is consistent with other pediatric case reports of rhino-orbital mucormycosis in immunocompetent hosts,15, 16 and this reminded us to include mucormycosis on our differential diagnostic lists in patients with similar presentations. This seems to be even more important if we notice the fatal course of this condition

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of interest

The authors declared no conflict of interest.

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