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Chronic Rhinocerebral Mucormycosis and Bilateral Optic Neuropathy: A Rare Case Report

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Abstract

Introduction: Mucormycosis is an emerging disease commonly seen in diabetic and immune-compromised patients. The common presentation consists of a rapidly progressive infection with a high mortality rate while the other rare presentation is a challenging diagnosis with a chronic progression and high morbidity as was seen in this case. Rhino-orbito-cerebral mucormycosis is one of the common forms of the disease. The most common presenting feature of Chronic Rhinocerebral Mucormycosis (CRM) is ophthalmologic. The incidence of internal carotid artery and cavernous sinus thrombosis is higher in CRM patients than in those with the acute disease.

Case report: We report a 53-year-old male who was presented with episodes of severe headache and symptoms of rhinosinusitis for six months. He was diagnosed and treated as an allergic fungal rhinosinusitis initially. The final histopathologic examination confirmed the diagnosis of chronic invasive mucormycosis and treated accordingly.

Conclusion: Chronic rhinocerebral mucormycosis is a rare form of mucormycosis which may present with atypical symptoms. It will have catastrophic consequences if it is suspected late or misdiagnosed with other diseases with similar symptoms such as allergic fungal rhino sinusitis or a simple chronic rhino sinusitis.

Keywords: Fungal rhinosinusitis; mucormycosis; Optic neuritis

Introduction

In the Chronic Rhinocerebral Mucormycosis (CRM), the disease course is indolent and slowly progressive, often occurring over weeks to months. CRM occurs predominantly in patients with diabetes and ketoacidosis. The most common presenting features of CRM are ophthalmologic and include ptosis, proptosis, visual loss, and ophthalmoplegia. We visited a case of CRM presented with severe headache and bilateral progressive visual loss leading to blindness of his left eye due to CRM.

Accurate diagnosis and prompt therapeutic interventions is the critical point in the face of these cases to prevent catastrophic consequences if CRM diagnosed and treated as an allergic fungal or simple chronic rhinosinusitis by mistake.

Case presentation

A 53-year-old male with history of diabetes mellitus was evaluated for a non- pulsatile, gradually increasing vertex headache with six-month duration. He had previously attended several neurology clinics where he was diagnosed with chronic headache and given analgesics. Subsequently, he developed nasal blockage, more on the left side and hyposmia. He referred to our clinic. Endoscopic nasal examination revealed absence of crust, ulcer, necrotic tissue, or any mass. A CT scan of his paranasal sinuses showed generalized mucosal thickening, soft tissue density mass filling ethmoid sinuses, and skull base erosion (Figure 1).

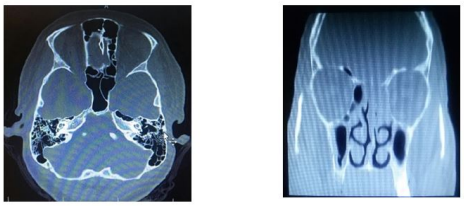
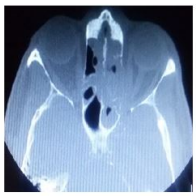


Figure 1 (Left): An axial bone window non-contrast CT scan of PNS depicting a soft tissue density mass filling the ethmoid sinuses.

Figure 1 (Right): Coronal bone window non-contrast CT scan in the same patient revealing skull base erosion.

He underwent endoscopic surgery with the working diagnosis of allergic fungal rhino sinusitis. Operative finding was fungal secretion, dehiscence of posterosuperior portion of left lamina papyracea, and anterior skull base epidural abscess which was drained with fungal secretion sent for histopathological examination. The patient's headache and nasal obstruction relieved and he was discharged from the hospital. On day 14 after discharge, his symptoms relapsed, and he developed impairment of left eye vision as well. The right eye was normal. Nasal endoscopic examination was performed in clinic and the suspected area was sent for frozen section which was negative for necrosis, indicative of mucoral infection. MR imaging of PNS disclosed a diffuse enhancement of ethmoid sinuses with a pressure effect on left optic nerve through a dehiscent lamina papyracea (Figure 2). Medical treatment with amphotericin-B, voriconazole, meropenem, and vancomycin was initiated for him empirically suspicious of invasive fungal rhinosinusitis. The microscopic description of previous surgical specimen revealed islands of 45 degrees branching hyphae with frequent septation suggestive of Aspergillus organisms. Following this result, amphotericin-B was removed from his drug regimen while voriconazole and antibiotics were continued. The patient was followed up with serial imaging as well as endoscopic nasal examination and debridement.



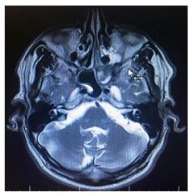


Figure 2 (left): The axial PNS CT scan demonstrating dehiscent left lamina papyracea posteriorly as noticed during operation.

Figure 2 (Right): The axial T2-weighted MR image showing a pressure effect of secretion over the left optic nerve via a dehiscent lamina papyracea.

Despite aggressive medical treatment, the patient's visual loss progressed on the left side leading to complete blindness with visual acuity on the right side also beginning to diminish. Given the worsening of his condition, a second time endoscopic surgery was planned. Wide surgical debridement was performed and left orbit was opened at posterior part to release the pressure effect of pus secretion over the optic nerve. Permanent sections confirmed broad, nonseptate hyphae consistent with mucormycosis. The bacterial culture was positive for Staph aureus. In addition, PCR testing for TB was negative. Treatment with intravenous liposomal amphotericin B was added to previous antibiotics and voriconazole. During the follow-up as in patient, he was subjected to several endoscopic surveillance as well as biochemical tests which presented a white blood cell count of 2000/mcl, platelets 21000/mcl, and Hb level of 11.2 gm/dl. Bone marrow aspiration showed hypocellularity. After haematologic consultation, voriconazole was eliminated from patient's antifungal regimen due to pancytopenia as a side effect of this medication. Oral prednisolone with a dose of 7.5mg/day was started. The blood glucose was regulated with insulin while he was on the corticosteroid. After 3 days, his right visual acuity fully recovered but left eye blindness sustained. He was discharged from the hospital with a maintenance dose of 7.5 mg of prednisolone for another two weeks. Recently (after three months) he seems stable, his left eye has no vision, and right eye is normal.

Discussion

We presented a 53–year-old diabetic male who was affected by chronic rhinocerebral mucormycosis. Mucormycosis is a devastating opportunistic infection of the paranasal sinuses and nasal cavity with progressive involvement of the orbit and brain [1]. It is an invasive infection caused by filamentous fungi of the Mucoraceae family. Most of the cases occur in patients with uncontrolled diabetes mellitus as seen in our patient and compromised immune status with a decreased ability to phagocytise [2]. It also has been described in immunocompetent patients with an incidence of 4-19% [3].

According to time of evolution, the infection can be acute or chronic, with the latter having a low frequency (5.6% of rhinocerebral mucormycosis cases). There is no exact definition of chronicity, which can vary from weeks to months. Classically, it is defined by symptoms that last for more than 4 weeks [1]. In the chronic infection, the disease course is indolent and slowly progressive [4].

The most common presenting features of CRM are ophthalmologic and include ptosis, proptosis, visual loss, and ophthalmoplegia. The incidence of internal carotid artery and cavernous sinus thrombosis is higher in CRM patients than in those with the acute disease [4]. There were no clinical and imaging findings indicative of orbital apex syndrome or cavernous sinus thrombosis in our patient.

The mechanism of orbital involvement is in two ways: either through direct invasion of lamina papyracea and involvement of orbital contents and further extension posteriorly into the orbital apex and even cavernous sinus, or via central retinal artery occlusion due to angioinvasiveness nature of mucoral infection which causes necrotizing vasculitis and resulting thrombosis of the vessel lumen [5].

Our patient diagnosed and treated for allergic fungal rhinosinusitis initially because he presented with atypical symptoms of a chronic form of mucormycosis infection.

Diagnosis of CRM is challenging and a high degree of clinical suspicion is required for correct diagnosis and prompt initiation of appropriate treatment due to atypical presentation, as was in this case. Macroscopically, it is characterized by tissue necrosis. Definitive diagnosis requires microscopic identification of the fungus in tissue specimens characterized by non-septated, 90° angle branching hypha, though the causative agent is isolated in less than 11% of chronic cases [6].

Differential diagnoses include chronic bacterial sinusitis, allergic fungal rhinosinusitis, sinonasal and orbital neoplasms, and granulomatous disease [7]. This reported case had been diagnosed and treated as allergic fungal rhinosinusitis initially.

Treatment is similar in both presentations, including wide surgical debridement of the involved tissue and systemic amphotericin B therapy while controlling the underlying comorbid factors. We also did the same after confirmation of mucor infection [8].

Conclusion

Mucormycosis is an opportunistic infection that affects mainly immunocompromised patients. Chronic rhino cerebral mucormycosis is a rare presentation and may present with atypical symptoms or coinfection with another agent. For this reason, it requires a high index of suspicion for correct diagnosis. In cases of persistent nasal congestion or ocular/facial pain in an immunocompromised patient with no clear cause of the symptoms, mucormycosis should be considered as causative agent.

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