

Uncommon Common Cold: Intracranial Fungal Ball

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ABSTRACT

Aim: To present an unusual clinical opportunity with invasive fungal sinusitis.

Background: Mucormycosis is a rare opportunistic zygomycotic fungal infection occurring mainly in immunocompromised patients. Rhinocerebral mucormycosis is the most common form of the disease.

Case description: Here we present a rare case of intracranial mucormycosis in an immunocompetent patient. The patient presented with left nasal block and recurrent nasal discharge. Nasal endoscopy showed multiple pale polyps. Magnetic resonance imaging findings showed left frontal, ethmoidal, and sphenoid sinusitis with leptomeningeal enhancement.

Management: A dual endonasal–transcranial approach was used by a neurosurgeon and ENT surgeon for the complete clearance of the disease. The histopathological examination was reported as mucormycosis, and the patient was started on inj liposomal amphotericin B. Patient was followed up postoperatively by serial nasal endoscopy and imaging as required.

Conclusion: Mucormycosis in an immunocompetent patient with polyps and dural erosion is a rare entity and hence requires complete removal of the disease process and reconstruction of the dura followed by the inj liposomal Amphotericin B. The patient needs regular clinicradiological follow-up to detect recurrence.

Clinical significance: Awareness of unusual manifestations and complications of paranasal sinus fungal disease is essential.

Keywords: Anterior skullbase, Fungal ball, Mucor, Polyposis.

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BACKGROUND

Rhinocerebral mucormycosis, also known as phacomycosis or zygomycosis, is a rare fatal opportunistic infection caused by the family of mucoraceae (Mucor, Rhizomucor, Rhizopus, and Absidia species) that are commonly found in soil and among decaying vegetation.¹ Several clinical forms have been seen, such as pulmonary, cerebral, gastrointestinal, rhino cerebral mucormycosis, or disseminated form. In patients with malignancy, the lungs are the more likely to be affected, while diabetic patients are more likely to have sino-orbital disease. Cutaneous infection is most likely associated with history of trauma. Rhino-orbital-cerebral is the most common form in diabetic patients. There is not enough data about mucormycosis in immunocompetent patients; some studies showed that cutaneous infection is the most affected organ in apparently healthy patients (42.5%) followed by rhino-orbital-cerebral mucormycosis (38.2%).² Management of mucormycosis still represents a big challenge and is based on different strategies that envisage a rapid diagnosis, elimination of risk factors (drugs or underlying diseases), rapid and aggressive antifungal infusion (polyenes) with or without surgical debridement,^{3,4} and lastly with adjuvant therapies in patients refractory or intolerant to polyene-based therapy (posaconazole, deferasirox plus lipid polyenes, recombinant cytokines granulocyte colony-stimulating factor, granulocyte macrophage colony-stimulating factor, or interferon- γ , hyperbaric oxygen).⁵

CASE DESCRIPTION

A 36-year-old male office worker presented to outpatient department (OPD) with complaints of recurrent left side nasal discharge and nasal block since 2 years and global dull aching heading ache since 6 months with no history of any comorbidities and vision disturbance or neck stiffness. Nasal endoscopy showed

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multiple pale polyps in the left side nasal cavity, and the right nasal cavity was normal. Computerized tomography (CT) of the paranasal sinuses showed irregularly enhancing mass in the left maxillary and frontal sinus with eroded posterior table of left frontal sinus (Fig. 1). Magnetic resonance imaging (MRI) with contrast was done which showed features suggestive of left side maxillary, ethmoid, and sphenoid sinusitis with leptomeningeal enhancement (Fig. 2).

Hence, a provisional differential diagnosis of invasive fungal sinusitis/fungal ball was made, and the patient underwent surgery under general anesthesia. Surgery involved a team of a neurosurgeon and an ENT surgeon. He was planned for a combined single-sitting transcranial microsurgical and transnasal endoscopic approach to achieve maximum clearance.

Procedure

A pericranial flap was elevated (Fig. 3), hemicraniectomy was performed, frontal sinus was opened and multiple fungal balls with

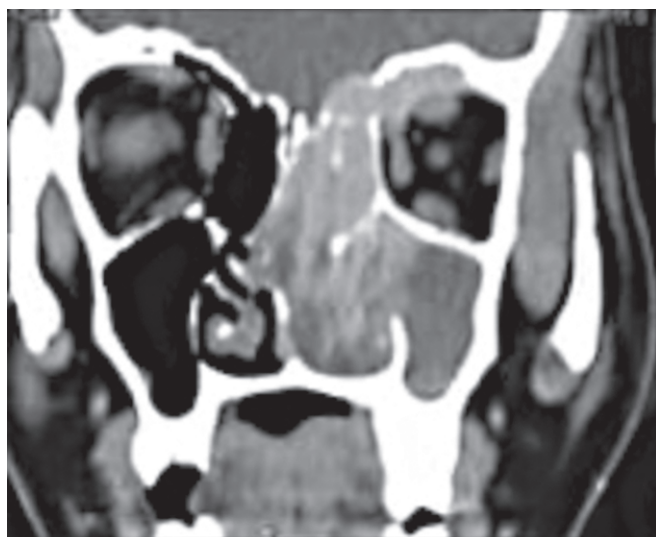


Fig. 1: Coronal computed tomography (CT) slice

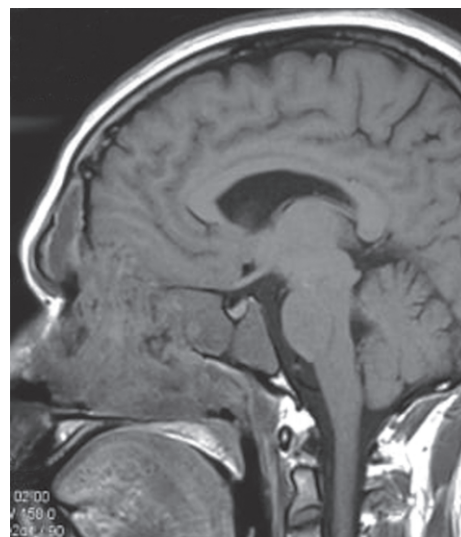


Fig. 2: Sagittal magnetic resonance imaging (MRI)

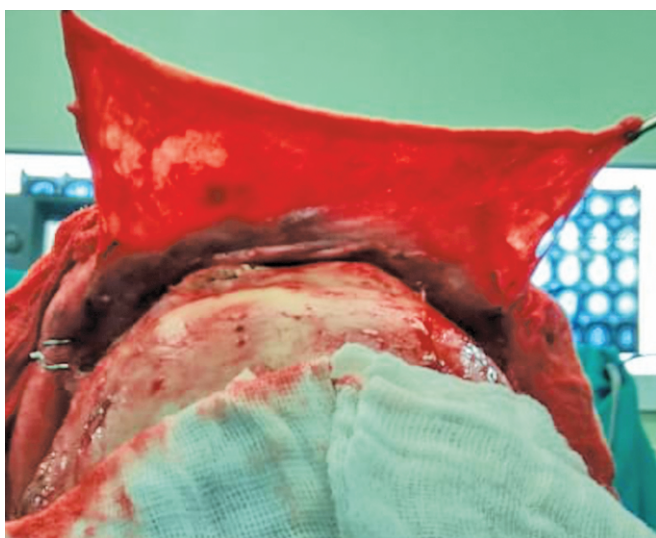


Fig. 3: Pericranial lap



Fig. 4: Frontal sinus muck

thick tarry mucus was seen in the frontal sinus (Fig. 4), with erosion of the posterior table of frontal sinus and minimal erosion in anterior table. The dura was exposed, erosive changes were seen, and the eroded dura was excised and reconstructed using autologous fat and fibrin glue. The eroded frontal table was debrided and reconstructed using titanium mesh.

Functional endoscopic sinus surgery was performed on the left side, and the maxillary, ethmoid, and sphenoid sinuses were cleared of polyps and fungal debris with fungal balls. The sphenoid sinus was relatively free of fungal balls and could be evacuated with a small sphenoidotomy.

Skullbase disease clearance was confirmed transnasally and intracranially by both surgical teams. A wide cribriform and posterior frontal defect was formed after disease clearance.

Reconstruction of the skull base was done using a pericranial flap⁶ and fat from the thigh intracranially and a Haddad-Bassagasteguy flap⁷ intranasally.

The fungal ball was sent for histopathology suspecting Aspergillosis but was reported as having aseptate hyphae suggestive of mucormycosis. Patient was administered a total dose

of 1.5 g iv of liposomal Amphotericin B over a period of 2 weeks. He tolerated the treatment well. He was regularly followed weekly for 3 weeks following which a repeat MRI was done which showed absolute clearance of the disease with no signs of recurrence. He was put on prophylactic itraconazole 100mg orally for 3 weeks following discharge.

Postoperatively nasal endoscopy showed well taken up nasal septal flap with no signs of cerebrospinal fluid leak. There was no postoperative complication like infection or flap give way.

The patient was on OPD follow-up for a period of 1 year following the surgery; after a quiet period, he presented with sinonasal polyposis with ethmoiditis on the right side (non operated side) about 10 months postoperatively with a relatively disease free maxillary and right frontal sinus on CT scan. This responded well to a course of systemic steroid followed by intranasal corticosteroid. He continues to be on outpatient follow-up.

DISCUSSION

Fungal infections of the paranasal sinuses have been well documented and form a significant portion of the rhinologist's

workload. While invasive fungal paranasal sinus disease usually presents in immunocompromised individuals with characteristic clinical features of spreading insensate ischemic patches of mucosa followed by necrosis, sinonasal polyps are a well-recognized clinical feature in fungal ball/allergic fungal sinusitis of the paranasal sinuses.⁸ This patient had no features of immunocompromise, and clinical evaluation of this patient was leading to a probable diagnosis of fungal ball with characteristic thick mucus and polyps. The erosion of the posterior table of the frontal sinus was a warning to the clinician that this was not a routine case of fungal ball. Although erosion of the paranasal sinus bones is seen in chronic noninvasive fungal disease of the PNS,⁹ MRI gave an additional surprise with loss of meningeal planes and leptomeningeal enhancement. Preoperative imaging enabled successful coordination of neurosurgeon and rhinologist to operate upon the patient with adequate patient information and preparation. Intraoperative findings again were in favor of fungal ball with characteristic ball-like fungal debris and thick muck with the exception of inflamed and friable dura mater over the affected area. Reconstruction of large anterior skullbase defect was successfully accomplished by use of vascular pericranial and nasal septal flaps on either side of the defect coupled with fat obliteration of the defect. Postoperative surprise was the histopathology report of aseptate hyphae leading to a provisional diagnosis of *Mucor* species infection. *Mucor* species have also been frequently identified in fungal balls in patients with noninvasive fungal disease of the paranasal sinuses.^{10,11} There are several hypotheses for the erosion of paranasal sinus bone in noninvasive fungal infection, including chronicity of the associated inflammation, bacterial coinfection, and limited local invasion by fungus.⁵ The successful treatment of mucormycosis requires 4 steps: (1) early diagnosis; (2) reversal of underlying predisposing risk factors, if possible; (3) surgical debridement where applicable; and (4) prompt antifungal therapy.³ Among patients with rhino-orbital-cerebral disease, CT typically reveals only sinusitis, so CT that indicates the absence of deeper infection does not rule out mucormycosis. MRI is more sensitive than CT for detecting orbital and central nervous system involvement.¹²

While fungal ball does not warrant treatment with Amphotericin B, in this patient, we had to initiate amphotericin B treatment due to grave clinical features, such as meningeal involvement and large skullbase erosions.

Use of vascular pedicled flaps on both sides of the defect gave the surgeons confidence in the postoperative result. Fat obliteration of the defect and frontal sinus with cranialization of the frontal sinus eliminated the need for wide transnasal exposure of the frontal sinus and the probable subsequent reconstruction of the posterior table.

Recurrence of nasal polyps on the unoperated side with absence of signs of fungal disease and clinical improvement with medical management indicates a possible pathology of nasal polyposis including a component of allergy to fungal antigens of *Mucor* species—testing of which is technically challenging.

CONCLUSION

An immunocompetent patient presented with clinical features of sinonasal polyposis with possible fungal ball—unusual extensive skullbase erosions and dura involvement were noted.

Surgical management involved extensive skullbase reconstruction by transcranial and transnasal approaches. Identification of *Mucor* necessitated postoperative amphotericin-B therapy. *Mucor* infection may present in fungal balls, invasive fungal sinusitis, and also has a possible role in immune dysfunction in patients with polyps.

CLINICAL SIGNIFICANCE

Any patient with sinonasal polyposis should be carefully evaluated with appropriate radiological investigations to rule out skullbase complications. In the presence of such complications, a team approach with neurosurgeon and otolaryngologist is important for a clean surgical plan and reconstruction. Fungus identification is important for postoperative decision-making and prognosis.

ETHICAL AND HUMANE CONSIDERATIONS

Relevant informed consents were obtained from the patient, the patient's identity has not been disclosed; no clinical trial was conducted.

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