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Case Report

Sphenoid sinus fungus ball presenting with bilateral visual disturbance

Bo-Nien Chen[1,3], Kuo-Ming Chang[2]

Departments of [1]Otolaryngology-Head and Neck Surgery, [2]Pathology, Hsinchu MacKay Memorial Hospital, Hsinchu, Taiwan; [3]Mackay Junior College of Medicine, Nursing and Management, New Taipei, Taiwan

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Abstract: Objectives: Sinonasal fungus ball is an extramucosal and noninvasive mycotic proliferation that fills one or more paranasal sinuses. Patients with sphenoid sinus fungus ball commonly present with nasal symptoms and headache. Visual disturbance is rarely reported and the recovery rate is very low. Methods: We report a 61-year-old man presenting with a 6-month history of progressively decreased visual acuity in both eyes. Computed tomography revealed opacified sphenoid sinuses with intralesional hyperdensity and bony wall defects at the optic canals on both sides and the lateral wall of the left sphenoid sinus. Magnetic resonance imaging revealed leptomeningitis involving the cavernous sinus and cerebral falx. Results: The sphenoid sinus fungus ball was removed through transethmoidal sphenoidotomy. Postoperative intravenous antifungal therapy was administered. His visual disturbance was markedly alleviated and no recurrence of a sphenoid sinus fungus ball was observed at the most recent follow-up, 3 months after surgery. Conclusion: A high index of suspicion is crucial for preventing complications involving visual disturbance in patients with sphenoid sinus fungus ball, particularly when bony defects of the sphenoid sinus are present. Timely surgical interventions are required for these patients who have visual disturbance.

Keywords: Sphenoid sinus, fungus ball, aspergilloma, vision loss, leptomeningitis

Introduction

Sinonasal fungus ball is an extramucosal and noninvasive mycotic proliferation that fills one or more paranasal sinuses [1, 2]. Patients with sphenoid sinus fungus ball commonly present with nasal symptoms (purulent rhinorrhea, post-nasal drip, nasal obstruction, cacosmia, and blood-streaked nasal discharge) and periorbital or retro-orbitalheadache [1-5]. However, visual disturbance is rarely reported in these patients (incidence 17%) and the recovery rate is very low [1]. Here, we report a case of sphenoid sinus fungus ball presenting with bilateral visual disturbance that was treated successfully through surgical intervention and antifungal therapy.

Case report

A 61-year-old man visited our hospital, presenting with a 6-month history of progressively decreased visual acuity in both eyes. He had type 2 diabetes mellitus and hypertension. However, he denied nasal symptoms, headache, and a history of nasal trauma or intranasal surgery.

Initial laboratory studies on admission revealed the following abnormal findings: C-reactive protein, 0.97 mg/dL; erythrocyte sedimentation rate, 27 mm/hour; ac sugar, 111 mg/dL. However, his white blood cell count was within the normal limit (7800/uL).

On ophthalmic examination, the patient's bilateral visual acuity consisted of only light perception. His extraocular movements were full and free. Fundoscopy revealed that the optic discs and retinal vessels were normal, and no retinal hemorrhage or exudates were observed. Visual field examination revealed an advanced visual field defect in both eyes.

On endoscopic nasal examination, a nasal polyp obliterating the left sphenoid sinus ostium was observed (**Figure 1A**).

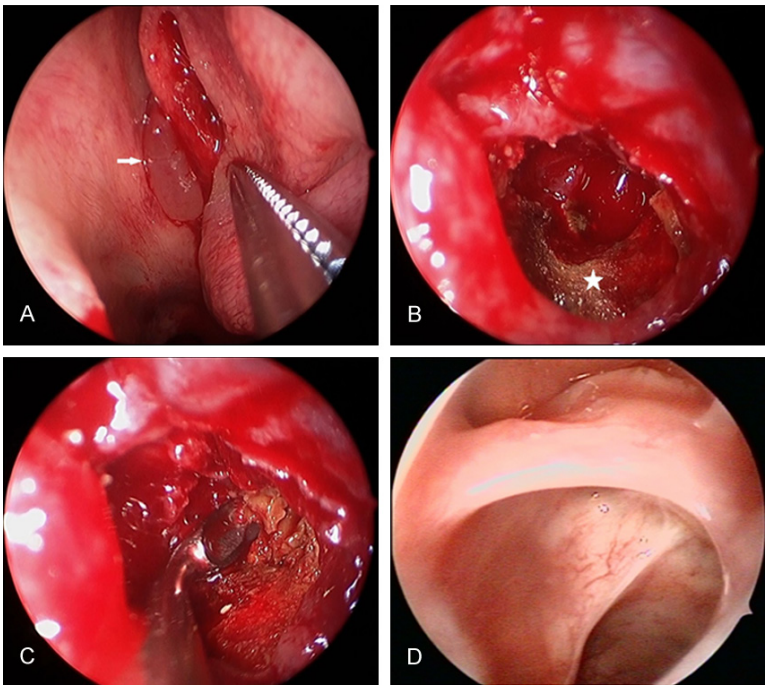

Figure 1. A. Endoscopic nasal examination revealed that a polyp (arrow) had obliterated the left sphenoid sinus ostium. B. Transethmoidal sphenoidotomy was performed and a soil-like fungus ball (asterisk) was noted in the left sphenoid sinus. C. The fungus ball was removed by curette. D. Follow-up endoscopy performed 3 months after surgery revealed no recurrence of a sphenoid sinus fungus ball.

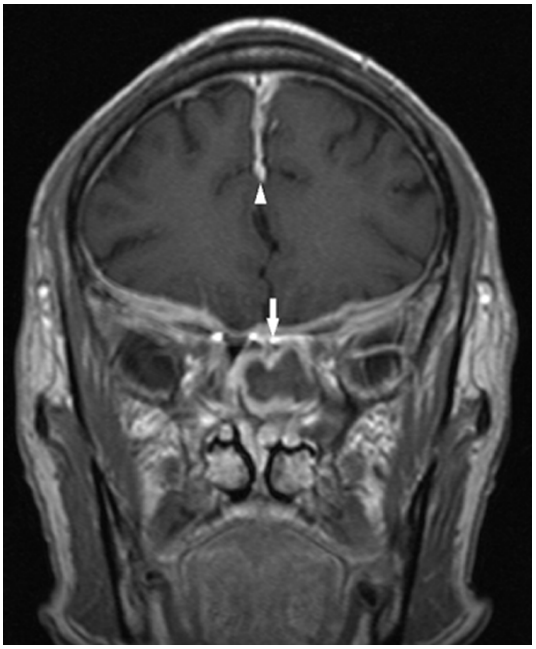

Figure 2. Preoperatively, contrast-enhanced, coronal T1-weighted brain magnetic resonance imaging revealed leptomeningitis involving the cavernous sinus (arrow) and cerebral falx (arrow head).

Brain magnetic resonance imaging (MRI) with gadolinium contrast revealed bilateral sphenoid sinusitis with leptomeningitis involving the cavernous sinus and cerebral falx (**Figure 2**). Computed tomography (CT) of the paranasal sinuses revealed opacified sphenoid sinuses with intralesional hyperdensity (microcalcification) and bony wall defects at the optic canals on both sides and the lateral wall of the left sphenoid sinus (**Figure 3**). The imaging studies favored a diagnosis of sphenoid fungal sinusitis with leptomeningitis.

A lumbar puncture was performed, and normal opening pressure was observed. Cerebrospinal fluid (CSF) was clear in appearance. Routine CSF examination revealed acellular CSF. Glucose (79 mg/dL), protein (51 mg/dL), and lactic dehydrogenase (16 mg/dL) levels in the CSF were slightly increased. Gram staining, acid-fast bacillus staining, potassium hydroxide mount, India ink staining, and CSF cultures were negative.

The patient subsequently underwent a surgical intervention. Transethmoidal sphenoidotomy was performed bilaterally under endotracheal general anesthesia. After sphenoidotomy, an intrasinus soil-like fungus ball was noted (**Figure 1B**). The fungus ball was removed by curette through a large sphenoidotomy (**Figure 1C**), and the sinus was irrigated profusely to remove all fungal debris. The time to surgical intervention following onset of visual disturbance was 6 months. The patient's postoperative course was uneventful. His visual disturbance was markedly improved on the first postoperative day.

Postoperatively, histopathological sections revealed sinonasal mucosa with mild to marked chronic inflammation. Many colonies of *Aspergillus* were found (some degenerating), which are clearly shown in PAS-D and GMS

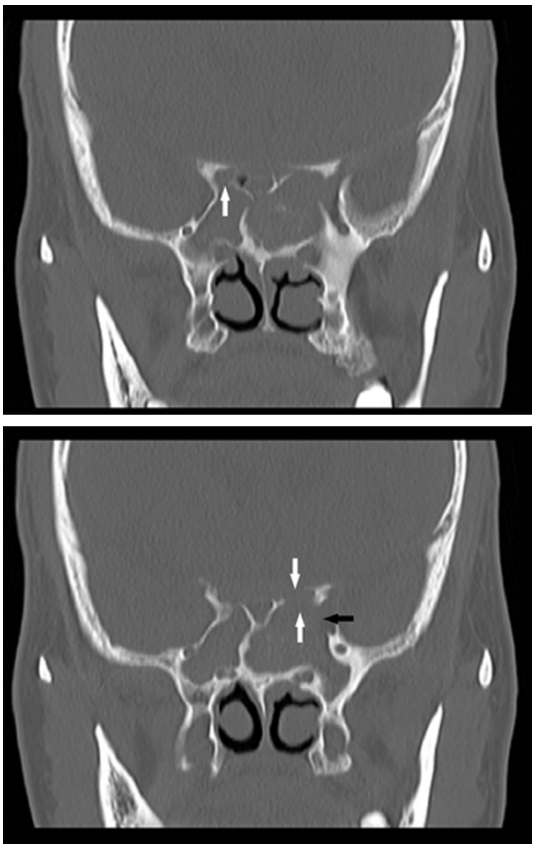

Figure 3. Preoperatively, coronal computed tomography of the paranasal sinuses revealed opacified sphenoid sinuses with intralesional hyperdensity and bony wall defects at the optic canals on both sides (white arrows) and the lateral wall of the left sphenoid sinus (black arrow).

stains; acute inflammatory cells are present at the periphery of many of them. Histopathological results demonstrated chronic paranasal sinusitis with many colonies of *Aspergillus* (fungus ball). Although the histopathological results were inconsistent with invasive fungal sinusitis, we decided to use a systemic antifungal agent under consultation with the infectious disease department because of the leptomeningitis revealed on the MRI. Antifungal therapy with intravenous voriconazole was administered for 12 days (loading dose: 6 mg/kg q12h for the first 24 hours, maintenance dose: 4 mg/kg q12h).

Follow-up endoscopy (**Figure 1D**) performed 3 months after surgery revealed no recurrence of a sphenoid sinus fungus ball.

Discussion

The prevalence of sinonasal fungus ball is increasing rapidly. The wide spread use of imaging and the increasingly frequent use of CT for the assessment of headache or chronic nasal discharge have considerably increased the rate of identification [5]. Between 4.5% and 26.8% of cases of sinonasal fungus ball have sphenoid sinus involvement [1, 3]. Patients are typically over 50 years old at the time of diagnosis and are predominately female (approximate 2:1 female: male ratio) [1, 2, 4, 5].

The etiopathogenesis is poorly elucidated, but decreased aeration of the sinus appears to play a major role in the development of the main pathogen, *Aspergillus fumigates* [6]. However, there was no significant correlation between the presence of sinonasal fungus ball and structural variations such as nasal septal deviation, concha bullosa, and Haller cells [2].

Most sphenoid sinus fungus balls remain in an indolent state, and endoscopic nasal examination is normal in approximately half of patients [5]. One study found that in 16% of patients, sphenoid sinus fungus ball was asymptomatic [7]. Thus, diagnosis is often delayed [1, 4, 5]. Because symptoms are nonspecific, the possibility of the disease is often overlooked, and most patients are not diagnosed with sphenoid sinus fungus ball until complications occur or by chance at a regular health checkup. The final diagnosis can be made only after an operation [1].

Sphenoid sinus fungus ball is a relatively uncommon entity, and although it is not invasive, if left untreated, it can lead to severe long-term complications because many crucial structures lie adjacent to the sphenoid sinus [3, 4].

Among patients with sphenoid sinus fungus ball, old age, underlying diabetes mellitus, and a sphenoid sinus wall defect visible in CT scans are factors significantly related to the occurrence of visual disturbances. But only sphenoid sinus wall defect which was found in 40% of patients with sphenoid sinus fungus ball showed significance in multivariate analysis [1]. The appearance of a bony wall erosion on a CT scan and the acute onset of sphenoid sinus aspergilloma symptoms were found to be significantly associated with the onset of orbital complications [4]. The walls of the optic nerve canal may be extremely thin, and defects have been described. A study pertaining to the danger areas of the sphenoid sinus suggested that

the thickness of the bony wall over the maximum bulging of the optic nerve canal averaged 0.28 mm, and bony defects were evident in 12% of cases [8]. Thus, it is important to have a high index of suspicion of visual disturbance in patients with sphenoid sinus fungus ball, especially when bony defects of the sphenoid sinus are present.

Vision and eye movements can be disturbed by neuritis, compression, or ischemic infarctions resulting from thrombophlebitis of the optic nerves in isolated cases of sphenoid sinus disease. Extraocular muscle restriction was observed in 63% of patients with sphenoid sinus fungus ball and visual disturbance [1].

Laterally, the sphenoid sinus is bordered by the cavernous sinus, containing the internal carotid artery and the cranial nerve (CN) VI (abducens). CN III (oculomotor), IV (trochlear), V1 (ophthalmic), and V2 (maxillary) are located in the cavernous sinus wall. The bone overlying the internal carotid artery is dehiscent in up to 25% of the population. Bone resorption also occurs with age, and bone thinning occurs in these regions in 80% of people older than 85 years [9]. Thus, cavernous sinus thrombosis and fungal internal carotid artery aneurysm are rare and fatal complications of sphenoid aspergillosis [10-12].

Endoscopic sinus surgery is the treatment of choice for sinonasal fungus ball, with a low morbidity and recurrence rate. Recurrence or residual diseases are observed in only 1.1% of cases [2, 3]. However, optic neuritis and visual disturbances lasting for more than 6 months were regarded as poor prognostic factors [13]. Patients with complete or near-complete visual loss did not show improved visual functioning [1]. Because of this low recovery rate, sphenoid sinus fungus ball with visual disturbance may be considered an emergent condition. A timely operation may prevent permanent sequelae.

Voriconazole is recommended as the first choice of antifungal agents for aspergillosis. *Aspergillus* infection is strongly invasive into arterial vessels. It is crucial to consider the possible occurrence of cerebrovascular disease when treating aspergillosis invasion of the central nervous system [14].

Conclusion

This study describes the importance of having a high index of suspicion to prevent complications involving visual disturbance in patients with sphenoid sinus fungus ball, particularly when bony defects of the sphenoid sinus are present. Moreover, timely surgical interventions are required for such patients with visual disturbance.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Bo-Nien Chen, Department of Otolaryngology-Head and Neck Surgery, Hsinchu MacKay Memorial Hospital, No. 690, Sec. 2, Guangfu Road, East Dist, Hsinchu 30071, Taiwan. Tel: +886 36119595; Fax: +886 36110900; E-mail: chenbonien@yahoo.com.tw

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