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CASE REPORT

Retrobulbar optic neuropathy associated with sphenoid sinus mucormycosis

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Key words
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Abstract

Because fungi usually spread from the paranasal sinuses to the orbital apex in invasive fungal sinusitis (IFS), IFS often presents as an orbital apex syndrome (OAS) characterized by dysfunction of cranial nerves II, III, IV, V1, and VI. We report a case of sphenoid sinus mucormycosis that presented as isolated retrobulbar optic neuropathy. A 94-year-old woman presented with acute blindness in the right eye. Examination revealed the absence of light perception and pupillary reflex in the right eye. Head MRI showed a mass in the right sphenoid sinus, which was contiguous with the right optic nerve. She underwent endoscopic surgery, and a histopathological diagnosis of mucormycosis was established. Treatment with intravenous liposomal amphotericin B reduced the size of the mass. She has survived for more than 1 year without recurrence. Clinicians should consider that IFS can present as isolated retrobulbar optic neuropathy.

Introduction

Invasive fungal sinusitis (IFS) is the infiltration of fungi from the sinus to the surrounding tissue.[1] It can cause unilateral or bilateral vision loss, which often presents as orbital apex syndrome (OAS).[2] Thus far, isolated optic nerve dysfunction associated with compression of the optic nerve by a mucor mass has not been reported. We experienced a case of sphenoid sinus mucormycosis that presented as retrobulbar optic neuropathy, which was promptly diagnosed using magnetic resonance imaging (MRI) and surgical biopsy.

Case Report

A 94-year-old woman presented with acute blindness in the right eye, which was noted 1 day prior to admission. Her past medical history included chronic heart failure, for which she had been taking digoxin. In addition, she had cataract and had undergone intraocular lens implantation surgery for both the eyes. Her best-corrected visual acuity (BCVA) was 20/40 in both eyes 14 days before admission during a regular visit to an ophthalmologist. On admission, neurological examination revealed the absence of pupillary reflex in the right eye and mild cognitive impairment. There were no other neurological abnormalities. Her BCVA in the right eye changed to no light perception, while that in the left eye was unchanged. Eye pressure, anterior segments, and fundus findings in both eyes were normal. Blood tests demonstrated no elevation of the level of β-D-glucan or aspergillus antigen. Head MRI revealed a mass with peripheral contrast enhancement in the sphenoid sinus that was contiguous with the right optic nerve (Fig. 1a,b). She underwent endoscopic endonasal transsphenoidal surgery, and wide nonbranching aseptate hyphae established a histopathological diagnosis of mucormycosis (Fig. 1c). Treatment with intravenous liposomal amphotericin B 100 mg daily for 1 month reduced the size of the mass, but her visual acuity remained unchanged. Fundus examination showed the right optic disk atrophy on day 45 of hospitalization. She has survived for more than 1 year without recurrence.

Discussion

We noted two clinically important findings in this patient. First, IFS can present as retrobulbar optic neuropathy. According to a review of 14 patients with IFS having ophthalmological symptoms, most cases were correctly diagnosed after presenting with complete OAS, probably because initial symptoms such as pain, headache, and fever are nonspecific.[2] Only one case of unilateral optic nerve infarction possibly caused by vascular invasion of *Mucorales* has been reported.[3] Our report describes a rare case of IFS caused by *Mucorales* presenting as retrobulbar optic neuropathy, which was a precursor to orbital infiltration.

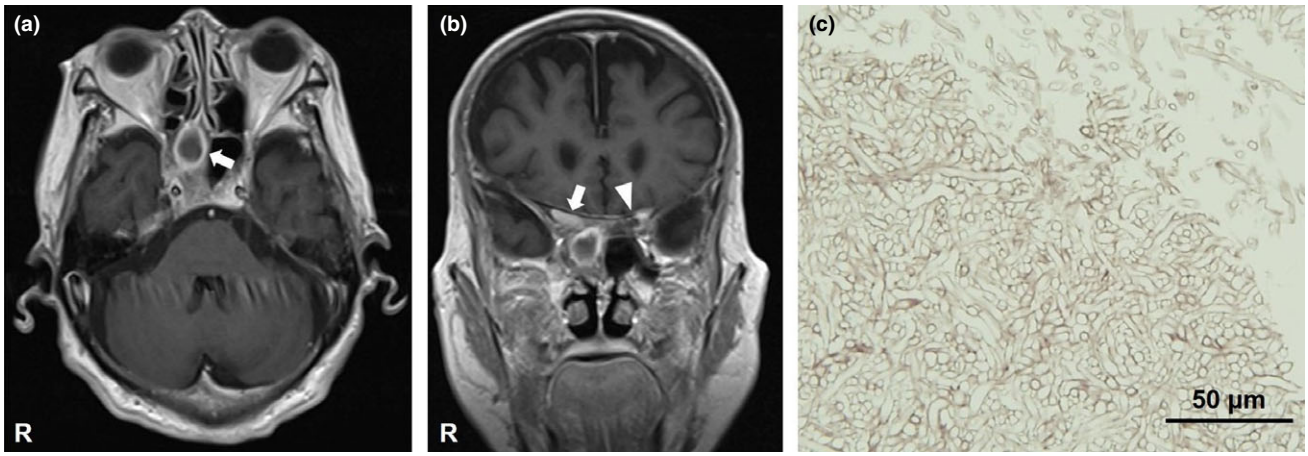

Figure 1 (a) Axial and (b) coronal brain magnetic resonance imaging (MRI) reveals a mass (a, arrow) in the sphenoid sinus and a contiguous hyperintense signal mass involving the right optic nerve (b, arrow) on gadolinium-enhanced T1-weighted imaging. The left optic nerve appears intact (b, arrowhead). (c) Photomicrograph of the surgical specimen shows wide nonbranching aseptate hyphae of *Mucorales* (Grocott staining, ×400).

Second, the combination of MRI and surgical biopsy was useful for the correct diagnosis of her condition. MRI is more sensitive than CT for identifying the involvement of extrasinus lesions in IFS.[2,4] Surgical biopsy and histopathological examinations are also important because cultures of specimens obtained from patients with mucormycosis are usually negative, even when the specimen is taken from the site of infection.[2,5] Furthermore, a histopathological analysis is useful for distinguishing *Mucorales* from other fungi such as *Aspergilli* whose hyphae are branching. Because IFS is a lethal condition, the combination of MRI and surgical biopsy is recommended to ensure correct diagnosis and selection of an appropriate treatment approach.

In this case, IFS mimicked the course of isolated optic neuritis, but other diseases can cause similar conditions.[6,7] A recent study found that 10% of cases diagnosed as optic neuritis were caused by other factors, including tumors, ischemic or hypertensive neuropathies, retinal or choroid disorders, infection, autoimmune diseases, hereditary diseases, and toxins.[6] MRI is also useful to differentiate these rare but serious conditions.

The prognosis of acute IFS is very poor, perhaps because IFS is considerably misdiagnosed at onset because of its unclear symptoms. MRI and surgical biopsy should be promptly performed to confirm this rare but fatal condition in light of the differential diagnosis of optic neuritis.

In conclusion, we reported a rare case of sphenoid sinus mucormycosis that presented as retrobulbar optic neuropathy.

Acknowledgment

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References

1 deShazo RD, Chapin K, Swain RE. Fungal sinusitis. *N. Engl. J. Med.* 1997; **337**: 254–9.
2 Lee DH, Yoon TM, Lee JK, Joo YE, Park KH, Lim SC. Invasive fungal sinusitis of the sphenoid sinus. *Clin. Exp. Otorhinolaryngol.* 2014; **7**: 181–7.
3 Alsuhaibani AH, Al-Thubaiti G, Al Badr FB. Optic nerve thickening and infarction as the first evidence of orbital involvement with mucormycosis. *Middle East Afr. J. Ophthalmol.* 2012; **19**: 340–2.
4 Thurtell MJ, Chiu AL, Goold LA *et al.* Neuro-ophthalmology of invasive fungal sinusitis: 14 consecutive patients and a review of the literature. *Clin. Exp. Ophthalmol.* 2013; **41**: 567–76.
5 Roden MM, Zaoutis TE, Buchanan WL *et al.* Epidemiology and outcome of zygomycosis: a review of 929 reported cases. *Clin. Infect. Dis.* 2005; **41**: 634–53.
6 Horwitz H, Friis T, Modvig S *et al.* Differential diagnoses to MS: experiences from an optic neuritis clinic. *J. Neurol.* 2014; **261**: 98–105.
7 Weerasinghe D, Lueck C. Mimics and chameleons of optic neuritis. *Pract. Neurol.* 2016; **16**: 96–110.