Ophthalmology Practice

Sinonasal carcinoma presenting as chronic sinusitis and sequential bilateral visual loss

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Sinonasal undifferentiated carcinoma-related rhinogenic optic neuropathy is rare and may lead to visual loss. To the best of our knowledge, this is the first report of bilateral sequential visual loss induced by this etiology. It is important to differentiate between chronic sinusitis and malignancy on the basis of specific findings on magnetic resonance images. Surgical decompression with multidisciplinary therapy, including steroids, chemotherapy, and radiotherapy, is indicated. However, no visual improvement was noted in this case, emphasizing the rapid disease progression and importance of early diagnosis and treatment.

Key words: Encasement, infiltration, optic neuropathy, rhinogenic, sinonasal undifferentiated carcinoma

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Optic neuropathy is characterized by damage to the optic nerve caused by compression, inflammation, infiltration, ischemic change, and glaucoma, which may lead to the different severity of visual impairment, even visual loss.[1] Optic neuropathy caused by lesions from the nasal or paranasal sinus is called rhinogenic optic neuropathy. Herein, we report a case of rhinogenic optic neuropathy resulting from sinonasal carcinoma, presenting with the sequential bilateral visual loss. Furthermore, a literature review was performed to discuss the possible mechanism, clinical presentation, imaging features, treatment choice, and prognosis of this rare case.

Case Report

A 75-year-old Asian female without systemic disease initially presented with nasal congestion, mucous rhinorrhea and progressively decreasing sense of smell. She was treated as allergic rhinitis with oral antihistamine and nasal steroid spray, but no obvious improvement. One month after the symptoms, paranasal sinus computed tomography (CT) showed heterogeneous soft tissue densities in the left maxillary, frontal, bilateral ethmoid, and sphenoid sinuses, with extensive bony destruction [Fig. 1]. Sinusitis was impressed and additionally given oral antibiotics.

However, 6 months later, the patient was referred to our clinic with a 2 weeks history of declining vision in one eye (oculus sinister [OS]). Examination disclosed no light perception and revealed a relative afferent pupillary defect, but normal fundus appearance [OS; Fig. 2a]. Visual field testing (oculus dexter) showed a temporal defect with moderate constriction [Fig. 3]. The diagnosis was not confirmed with suspicion of central retinal artery occlusion or optic neuropathy. Orbital magnetic resonance imaging (MRI) was suggested and indicated multiple sinusitis with bone erosion and intracranial extension. Furthermore, encasement of the right optic nerve and infiltration of the left optic nerve near the optic chiasm were noted without abnormal optic nerve enhancement after contrast [Fig. 4]. Due to the finding of the intracranial lesion, the cerebrospinal fluid examination was suggested and yielded normal results. Surgical debridement for sinusitis with suspected optic neuropathy was suggested by the ophthalmologist but not performed immediately.

Moreover, 3 weeks after the first episode of vision loss (OS), vision loss suddenly occurred in the other eye.

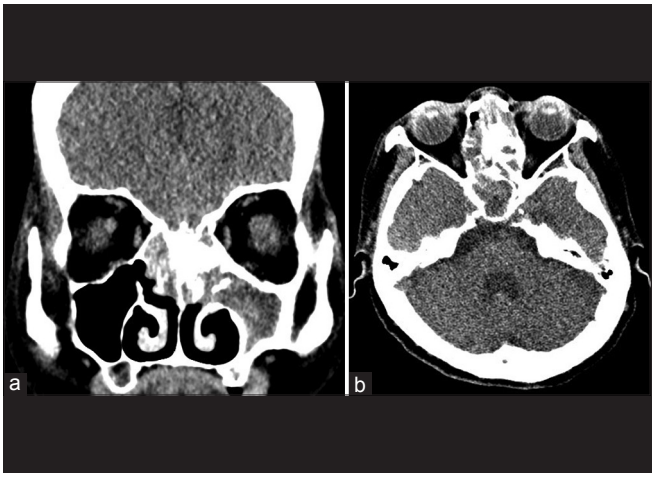

Figure 1: Computed tomography (CT). Paranasal sinus CT (a: Coronal view, b: Axial view) shows sinusitis with bony erosion over left maxillary, frontal, bilateral ethmoid and sphenoid

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Under the impression of optic neuropathy associated with presumed sinusitis (oculus uterque [OU]), the patient received mega-dose steroids (methylprednisolone 250 mg q6 h for 5 days) and emergent bilateral sinusectomy. Because of unusual clinical course, specimen was collected for pathology, which surprisingly indicated undifferentiated carcinoma. The findings were highly suggestive of sinonasal undifferentiated carcinoma (SNUC)-related retrobulbar infiltrative optic neuropathy (OU); therefore, the patient underwent concurrent radiochemotherapy as malignancy therapy.

Three months after the first episode of visual loss, ocular examination still showed no light perception (OU). The pattern visual evoked potential (VEP) failed to perform due to poor vision and flash VEP [Fig. 5] revealed delayed latency and decreased amplitude. The electroretinography results were no obvious abnormality [Fig. 6]. At the final visit (6 months after the first visual loss episode), no light perception (OU) persisted even though the disc appeared pinkish [Fig. 2b].

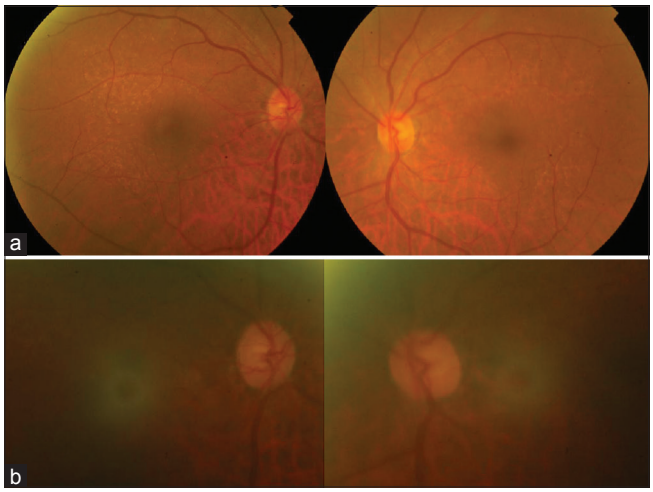

Figure 2: Fundus appearance. Fundus appearance reveals no obvious abnormality, including disc, macula and vessels initially 2 weeks after left visual loss (a). Six months later with bilateral visual loss, disc does not reveal pale appearance (b) (poor quality due to loss of fixation)

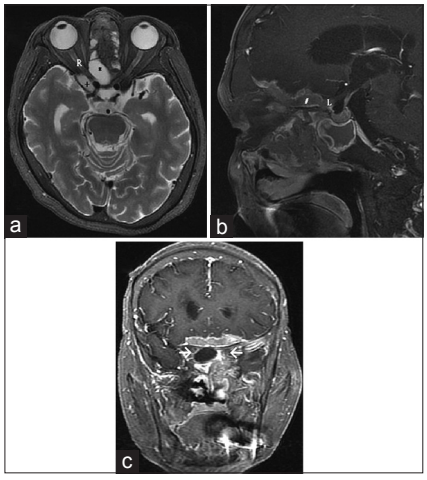

Figure 4: Optic nerve in orbital magnetic resonance imaging. Encasement of the right optic nerve (marked as "R") by the fluid-filled Onodi cell (*) and the fluid-filled anterior clinoid process (+) is suspected on T2-weighted image (a); left optic nerve (L) is infiltrated by the anterior cranial fossa lesion (#) (b, T1 with enhancement and fat suppression), which are indicated in coronal view (c, T1 with enhancement and fat suppression, right optic nerve marked as "→," left optic nerve marked as "←")

Discussion

Rhinogenic optic neuropathy is characterized by optic nerve damage secondary to the sinonasal lesion, and includes various sinonasal diseases such as sinusitis, mucocele, and sinus surgery,[1] with the main mechanism being compression and inflammatory changes.[2] In addition to optic neuropathy, orbital invasion is a common manifestation with ocular findings such as lid edema, blepharoptosis, conjunctival chemosis, conjunctival injection, diplopia, limitation of ocular motility, globe displacement, epiphora, anisocoria, eye pain, and photophobia.[3] The main treatment for rhinogenic optic neuropathy includes surgical decompression and steroid use. The efficacy of steroid therapy for rhinogenic optic neuropathy has not been elucidated, possibly due to the limited number of cases.[1] Surgical intervention in the form of orbital or optic nerve

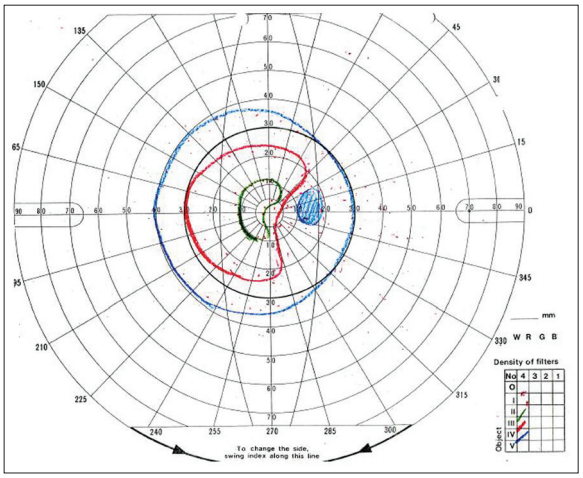

Figure 3: Visual field (VF). VF (oculus dexter) shows temporal defect with moderate constriction while visual loss

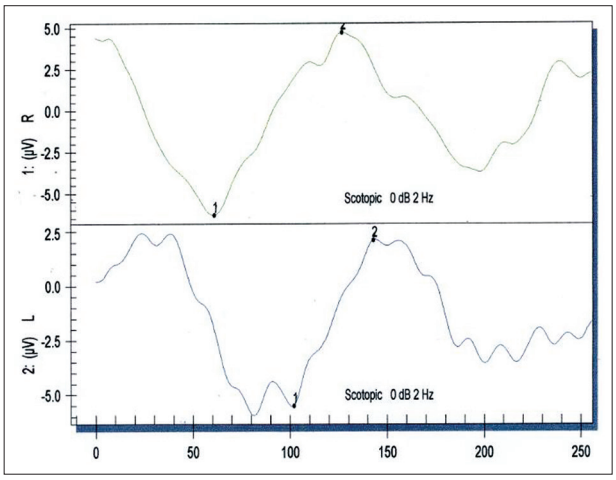

Figure 5: Flash visual evoked potential (VEP). Two months after visual loss (oculus dexter [OD]) and 3 months after visual loss (oculus sinister [OS]), pattern VEP fails due to tracing inability with no light perception; flash VEP reveals delayed latency (OD: 120 ms, OS: 140 ms) and decreased amplitude (OD: 11 uV, OS: 7.5 uV)

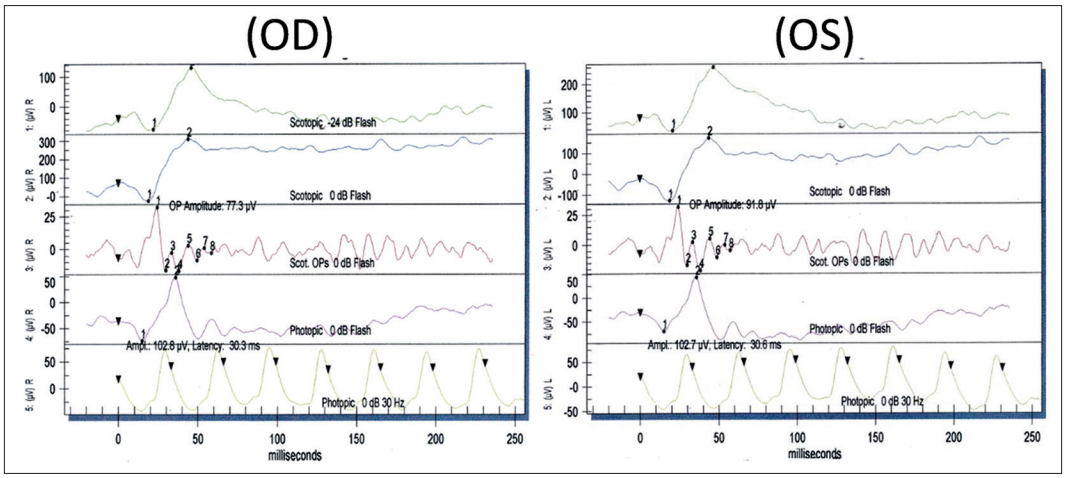

Figure 6: Electroretinography (ERG), ERG reveals no obvious abnormal amplitude both in rod and cone function

decompression accompanied by systemic steroids resulted in favorable visual outcome;[1,4] however, poor prognosis was still mentioned in case reports of sphenoid sinus mucocele and eosinophilic chronic rhinosinusitis.[2] Preoperative visual acuity tended to correlate with prognosis; therefore, patients with severely impaired vision should be given more attention and treated aggressively.[5]

Malignancy is also a possible reason for rhinogenic optic neuropathy. SNUC, a rare and aggressive malignancy, was first described in 1986 as a highly aggressive neoplasm arising from the nasal cavity and paranasal sinuses.[5,6] The most common locations of SNUC are the nasal cavity and ethmoid sinus; in addition, orbital and intracranial invasion and distant metastasis are frequent findings.[7] Bone destruction and invasion of adjacent structures such as the orbit, cranial vault, and skull base is frequently seen.[8] In addition to retrobulbar optic neuropathy, other ophthalmic symptoms of the oculomotor nerve palsy, optic atrophy, and proptosis are possible manifestations.[9] Due to the frequent cranial invasion, there are several possible mechanisms to explain the sudden visual loss with intracranial mass lesions, including ischemia due to vascular compression of the visual pathways or demyelination secondary to compression.[10]

In all previous cases of SNUC related optic neuropathy, one eye is affected more than the other eye,[5] or just only one eye affected,[9] while this case is totally blindness in both eyes. The possible mechanisms included infiltration or anatomical contact, such as encasement and decompression. The cause of bilateral visual loss was possibly ischemia, originating from a compromised visual pathway vascular system caused by the SNUC, which could not be excluded even though no optic disc pale was noted 3 months after first visual loss (OS).

Distinguishing mucosal inflammatory and neoplastic lesions of the paranasal sinuses by medical imaging modalities may be appropriate but difficult, as seen in our patient. SNUCs have typical but nonspecific imaging characters, such as a large, expansile, and aggressive lesion with bone destruction and invasion of adjacent structures, including the anterior cranial fossa, adjacent paranasal sinuses, and orbits.[8] On CT scans, the tumors are noncalcified, with variable contrast enhancement. On MRI scans, the lesions

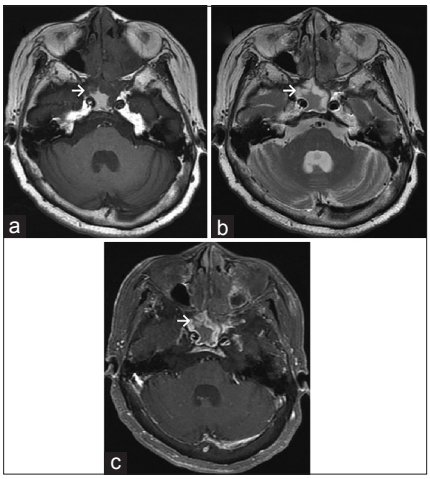

Figure 7: Sinonasal undifferentiated carcinoma (SNUC) in different magnetic resonance imaging modalities. To differentiate SNUC by different modalities, the lesion reveals isointense to muscle on T1-weighted image (a, marked as "→"), hyperintense to muscle on T2-weighted image (b, marked as "→") and heterogeneous appearance after enhancement (c, marked as "→")

are typically isointense to muscle on T1-weighted scans, iso-to hyperintense to muscle on T2-weighted scans, and heterogeneous on enhancement with gadolinium [Fig. 7a-c].[8] These imaging findings could not provide much advantage in differentiating SNUC from other pathological entities in this region.

Overall survival of patients with SNUC is poor in most reported series,[6,11] especially for those presenting with brain parenchymal invasion.[11] The visual outcome is not available in the literature owing to the limited number of cases; however, surgical decompression with steroid therapy did not results in any improvement in our patient. No clear consensus exists regarding treatment for SNUC, but a combination of operation, chemotherapy, and radiotherapy may be suggested.[1,2,5,11]

To the best of our knowledge, this is the first report of bilateral sequential total blindness induced by SNUC-related rhinogenic optic neuropathy. Because of aggressive optic

neuropathy and poor survivor in SNUC, it is important to differentiate between chronic sinusitis and SNUC, or other sinonasal malignancies, based on clinical presentation and image study of MRI. Atypical clinical presentation includes poor response to medication, such as antihistamine and antibiotics, ocular and visual symptoms; furthermore, imaging suggests sinus wall bony erosion, multiple invasion, and obvious intralesional necrosis. More attention and aggressive surveillance, such as early biopsy, should be given to the patients mentioned above.

Early surgical decompression with multidisciplinary therapy, including chemotherapy and radiotherapy, is suggested. Furthermore, steroid use in optic neuropathy may be beneficial. However, in our experience, surgical decompression by sinusectomy with systemic steroids and antibiotics did not improve the visual outcome, which emphasizes the rapid disease progression and importance of early diagnosis and treatment.

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