• Letter to the Editor •

A brain stem tumor with visual discomfort

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Dear Editor,

Primary intracranial tumor is a leading cause of morbidity and mortality in patients with neurological diseases[1]. Due to variations in size, location, and pathology, patients with brain tumors exhibit a wide range of symptoms. In the early stage, patients may present visual symptoms in particular conditions. It may include typical visual symptoms due to intracranial hypertension or atypical symptoms[2]. Furthermore, optic disc abnormalities such as papilledema, optic atrophy, or disc pallor can also be observed in conditions affecting the visual pathway at various levels[3-4]. Notably, the mean duration from the onset of visual symptoms to diagnosis of intracranial tumor can be as long as several years, underscoring the need for timely recognition of ocular signs[5]. Thus, it is important to suspect brain tumors in patients presenting with visual symptoms as their initial complaints and promptly conduct appropriate evaluation and management. We present a case of a young man whose only complaint was difficulty maintaining visual fixation during head movements—despite normal central vision—and was unexpectedly found to have a brain tumor.

Ethical Approval This study was approved by the Institutional Review Board (IRB) of Soonchunhyang University Seoul Hospital (IRB No. 2024-07-008). This study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent was obtained from the patient for publication of the images and data included in this article.

Case Presentation An 18-year-old male patient visited our clinic for blurring and difficulty focusing on targets while riding in a car or walking from 2mo ago. He had no history of any significant trauma, systemic diseases, ophthalmic diseases, or surgical history. His best-corrected visual acuity (BCVA) was 20/20 in both eyes. The intraocular pressure (IOP) was 21 mm Hg in his right eye and 22 mm Hg in his left eye. Physical examination showed jerky type gaze-evoked nystagmus (GEN) on right gaze, whereas no nystagmus was present in the primary position or on left gaze. There was a right hypertropia of 2 prism diopters (PD) in the primary position, which increased to 4 PD with right head tilt and to 3 PD with left head tilt. There was no abnormal head posture. In the fundus examination, an incyclotorsion was observed in the right eye and an excyclotorsion was observed in the left eye, along with mild papilledema (Figure 1A). We conducted a Hess screen test and observed right hypertropia (Figure 2). Since the patient had no drug history, the possibility of drug-induced GEN was ruled out. However, considering the clinical findings of hypertropia and mild papilledema, a central brain lesion was strongly suspected and brain imaging was performed immediately.

On brain magnetic resonance imaging (MRI), a 3.9 cm×3.3 cm sized lobulated solid and cystic mass was identified in the fourth ventricle (Figure 3). Because of hydrocephalus, emergency endoscopic third ventriculostomy was performed. Over a two-week period, the patient developed new symptoms including headache, oscillopsia, diplopia, and dizziness. His BCVA declined to 20/32 in both eyes. Right hypertropia increased to 6 PD in the primary position, and GEN was observed in all directions. These symptoms were likely associated with the progression of the mass effect and secondary hydrocephalus prior to tumor resection. Subsequently, the neurosurgery department performed tumor resection surgery. The brain tumor was histologically confirmed to be a pilocytic astrocytoma (WHO grade I). There was no unanticipated event due to surgery. Two weeks after the surgery, the patient presented with 15 PD of right hypotropia, 20 PD of esotropia, and persistent diplopia. Fundus examination revealed ongoing papilledema (Figure 1B). At 6mo postoperatively, his BCVA was 20/80 in the right eye and 20/40 in the left eye, with 10 PD of residual esotropia. Papilledema had improved on fundus examination, although temporal disc pallor persisted (Figure 1C). Four years after surgery, his BCVA improved to 20/25 in the right eye and 20/20 in the left eye. Despite management, nystagmus persisted.

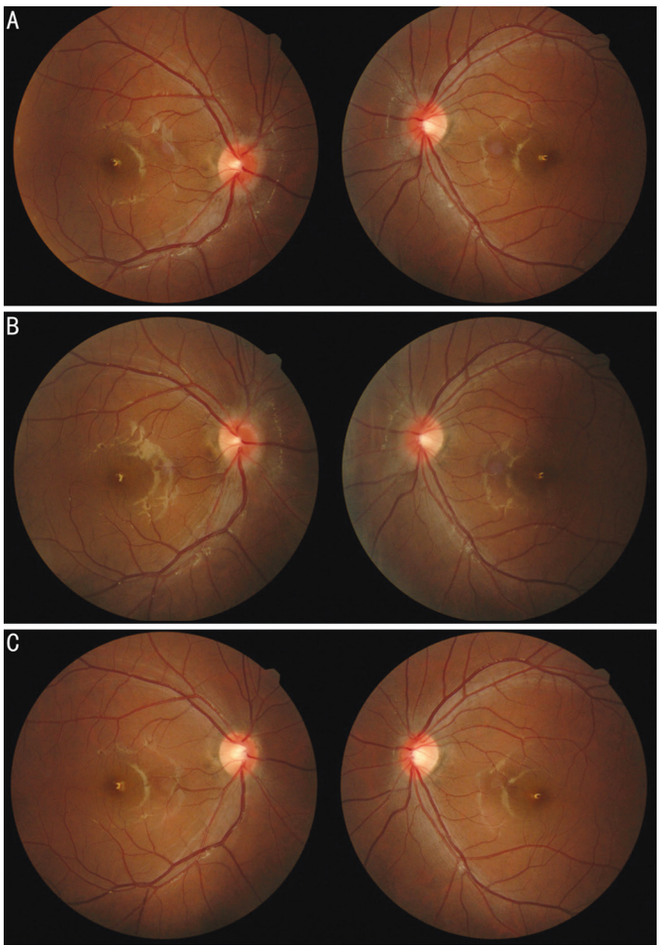


Figure 1 Progression of fundus photographs A: Fundus photograph at initial visit showing incyclotorsion of right eye and excyclotorsion of left eye with mild papilledema; B: Fundus photograph at 2wk after tumor resection showing persisted papilledema; C: After 6mo, the papilledema improved.

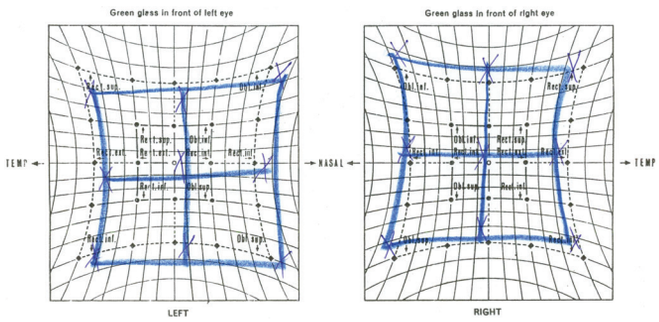


Figure 2 Hess screen test showing right hypertropia.

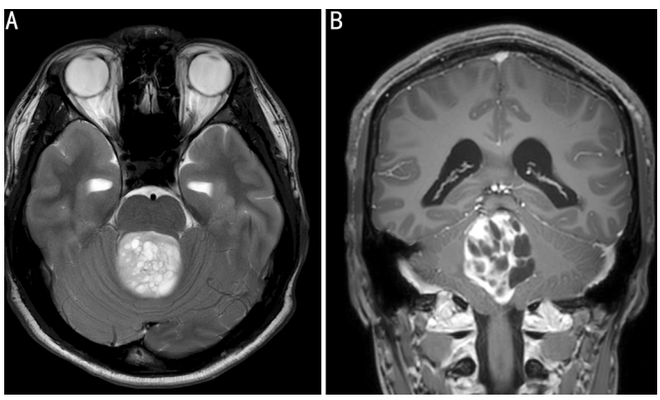


Figure 3 The first brain magnetic resonance imaging The axial T2-weighted image (A) and the coronal T1-weighted image with gadolinium (B) showing a 3.9 cm×3.3 cm sized lobulated solid and cystic mass in the fourth ventricle.

DISCUSSION

This case presents a young patient diagnosed with a fourth ventricle tumor after visiting the ophthalmology clinic with only subtle visual symptoms, specifically difficulty focusing during motion. At the first presentation, GEN, hypertropia, and mild papilledema were noted—findings that ultimately led to the suspicion of a central lesion and subsequent diagnosis of pilocytic astrocytoma.

Clinicians should be concern about structural brain abnormalities or drug/metabolic disorders when vertical, torsional, or GEN is detected. While drug-induced toxicity is the most common cause of GEN[6], the absence of such a history in this patient raised suspicion of a central lesion. The nucleus prepositus hypoglossi (NPH) and the medial vestibular nucleus (MVN), which serve as neural integrators, are located near the floor of the fourth ventricle[7]. When their function is compromised, the eyes fail to stay at the desired position in one direction and drift back to primary gaze, prompting GEN as a compensatory mechanism[8]. In this case, the large fourth ventricle mass likely impaired the MVN, NPH, and flocculus through mass effect, leading to the observed nystagmus. Distinguishing GEN from physiologic end-point nystagmus (EPN) is also clinically important. GEN is typically sustained with a larger amplitude (4 degrees or more) and is often asymmetrical[8].

Following treatment, papilledema, hypertropia, and cyclotorsion improved, but the nystagmus persisted. The improvement of nystagmus after removal of a brain tumor varies case by case. Adams *et al*[9] reported complete disappearance of nystagmus following resection of a 4-cm giant pituitary adenoma in patient with seesaw nystagmus. Kuwano *et al*[10] have reported improvement of nystagmus after tumor removal in a patient with spontaneous downbeat nystagmus caused by a 3 cm sized posterior fossa meningioma. However, there is also a report of new-onset vertical nystagmus appearing after surgical resection of a brain stem cavernoma, confirmed on follow-up videonystagmography 6mo postoperatively, suggesting that irreversible damage to oculomotor structures may occur regardless of tumor removal[11]. In the present case, following the diagnosis of a brain tumor, endoscopic 3rd ventriculostomy was performed to treat hydrocephalus, followed by tumor resection surgery 2wk later. Subsequently, there was improvement in papilledema, although nystagmus persisted. This outcome might be due to the tumor's large

preoperative size, which had already caused irreversible damage to the brainstem and cerebellum, resulting in persistent nystagmus even after surgery.

Brain tumors can initially manifest with mild or atypical visual disturbance, even when central vision remains intact. Therefore, clinicians should not underestimate these signs, and must thoroughly examine eye movements and the optic nerve for abnormalities such as nystagmus, vertical strabismus, fundus torsion, or papilledema. Early neuroimaging is essential for timely diagnosis.

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