CASE REPORT

Alternating Skew Deviation from Traumatic Intracranial Hypotension

Stephen J. Moster and Mark L. Moster

*Department of Neuro-Ophthalmology, Wills Eye Institute, Philadelphia, Pennsylvania, USA; and
Departments of Neurology and Ophthalmology, Thomas Jefferson University School of Medicine,
Philadelphia, Pennsylvania, USA*

ABSTRACT

A 56-year-old woman developed progressive headache, mental status changes, and diplopia after trauma. She was diagnosed with alternating skew deviation caused by intracranial hypotension. This is the first case of alternating skew deviation reported from intracranial hypotension and perhaps a differential pressure between intracranial and intraspinal spaces plays a role in the development of these findings.

Keywords: Alternating skew, intracranial hypotension, trauma

INTRODUCTION

Alternating skew deviation refers to a supranuclear vertical ocular deviation that reverses in different directions of gaze.¹ For example, one might have a left hypertropia in left gaze and a right hypertropia in right gaze. Prior reports have shown that this occurs most commonly in lesions of the cerebellum, cervicomedullary junction or pretectal region.^{1,2} We report a patient with alternating skew deviation associated with intracranial hypotension.

CASE REPORT

A 56-year-old woman was seen as an inpatient neuro-ophthalmic consultation for diplopia while hospitalised for mental status change.

One year prior, she had fallen and hit the back of her head on concrete steps with subsequent loss of consciousness. For the ensuing year, she had progressive symptoms including postural headache, nausea, vomiting, and dulling of her mental status.

Magnetic resonance imaging (MRI) a few weeks prior to neuro-ophthalmologic consultation revealed

diffuse meningeal enhancement, 8 mm of cerebellar tonsillar herniation, and inferior displacement of the diencephalon, consistent with intracranial hypotension (Figure 1). Lumbar puncture at a different institution was reportedly normal, but opening pressure was not measured.

Evaluation for a source of the cerebrospinal fluid (CSF) leak was unrevealing. She received a lumbar blood patch with transient improvement for a few weeks. She then developed further mental status changes, difficulty walking, postural headache, nausea, vomiting, and vertical diplopia.

Neurologic examination revealed mild mental status changes—recalling 1 of 3 items at 5 minutes, improving to 2 items with prompting. Neuro-ophthalmic examination revealed a 5-prism-dioptre (PD) right hypertropia in right gaze and a 2-PD left hypertropia in left gaze consistent with alternating skew deviation. Bed rest while hospitalised yielded much relief in her mental status and diplopia. Treatment with large-volume (25-cc) blood patch resulted in further improvement in her headache, gait, and diplopia.

On short-term and 6-month follow-ups, she had a persistent right hypertropia, with resolution of her left

FIGURE 1 Sagittal T1-weighted MRI with gadolinium showing meningeal enhancement and downward shift of brainstem and cerebellar tonsils.

FIGURE 2 Sagittal T1-weighted MRI with gadolinium at 8-month follow-up showing improvement in downward shift of brainstem and cerebellar tonsils.

hypertropia. Diplopia was mild and her mental status had normalised. Sequential MRIs over 8 months showed moderate improvement as well (Figure 2).

DISCUSSION

Diplopia is a frequent finding in patients with intracranial hypotension, occurring in 30–35% of

patients. Most often the abnormality is a unilateral or bilateral 6th nerve palsy and less commonly a 3rd or 4th nerve palsy occurs. Potential causes may include traction on the cranial nerves from downward displacement, cranial nerve compression, or vascular congestion of the nerves.³ Rare cases of a supranuclear vertical gaze palsy have been reported, such as a patient with a dorsal midbrain syndrome that was attributed to stretching of the midbrain structures controlling vertical gaze.⁴

Alternating skew deviation refers to a supranuclear hypertropia that reverses direction in different positions of horizontal gaze. One form—with a right hypertropia in left gaze and left hypertropia in right gaze—may be confused with a bilateral 4th nerve paresis. More commonly, the pattern that was seen in our patient occurs—a right hypertropia in right gaze and left hypertropia in left gaze. This can be seen in patients with brainstem or cerebellar disease and often occurs in cervicomedullary junction lesions along with downbeat nystagmus.²

This appears to be the first case reported of alternating skew from intracranial hypotension. Other reported causes have included cervicomedullary junction disease (e.g., Arnold-Chiari, basilar invagination, or platybasia), cerebellar or spinocerebellar degeneration, posterior fossa involvement with infarct, haemorrhage, tumour, encephalitis, trauma or demyelination, Wernicke encephalopathy, lithium toxicity, Joubert syndrome, a rare motor neuronopathy, anti-glutamic acid decarboxylase (GAD) antibody syndrome, paraneoplastic syndrome, syringomyelia/syringobulbia, tentorial herniation, or idiopathic intracranial hypertension.^{1,2,5–11}

One might argue that the diplopia in our patient is coincidental to the intracranial hypotension, since it did not disappear after treatment. However, the timing of the onset simultaneous to the rest of the clinical presentation, and the partial improvement along with the partial improvement on MRI, favours the intracranial hypotension as the cause of the alternating skew deviation.

The mechanism for alternating skew deviation is not clear. Zee has proposed that in patients with cerebellar disease, there is an imbalance in otolith-ocular pathways and a loss of the normal correction for the differences of pulling direction and strength of the extraocular muscles as the eyes change position in the orbit.⁵

Our patient, along with some other reported associations, particularly Arnold-Chiari malformation,² cerebellar vermis haemorrhage¹⁰ and idiopathic intracranial hypertension,⁶ might suggest that some of this dysfunction is related to the anatomic distortion that occurs with a pressure differential between intracranial and intraspinal spaces with downward shifting of brainstem and/or cerebellar structures.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

REFERENCES

- [1] Keane JR. Alternating skew deviation: 47 patients. *Neurology* 1985;35:725–728.
- [2] Moster ML, Schatz NJ, Savino PJ, Benes S, Bosley TM, Sergott RC. Alternating skew on lateral gaze (bilateral abducting hypertropia). *Ann Neurol* 1988;23:190–192.
- [3] Zada G, Solomon TC, Giannotta SL. A review of ocular manifestations in intracranial hypotension. *Neurosurg Focus* 2007;23:E8.
- [4] Fedi M, Cantello R, Shuey NH, et al. Spontaneous intracranial hypotension presenting as a reversible dorsal midbrain syndrome. *J Neuroophthalmol* 2008;28:289–292.
- [5] Zee DS. Considerations on the mechanisms of alternating skew deviation in patients with cerebellar lesions. *J Vestib Res* 1996;6:395–401.
- [6] Bruce BB, Newman NJ, Biouesse V. Ophthalmoparesis in idiopathic intracranial hypertension. *Am J Ophthalmol* 2006;142:878–880.
- [7] Couch SM, Brodie SE, Leavitt JA, Brodsky MC. Something to sink your teeth into. *Surv Ophthalmol* 2011;56:544–549.
- [8] Hamed LM, Maria BL, Quisling RG, Mickle JP. Alternating skew on lateral gaze. Neuroanatomic pathway and relationship to superior oblique overaction. *Ophthalmology* 1993;100:281–286.
- [9] Thakore NJ, Pioro EP, Rucker JC, Leigh RJ. Motor neuronopathy with dropped hands and downbeat nystagmus: a distinctive disorder? A case report. *BMC Neurol* 2006;6:3.
- [10] Tsuda H, Nagamata M, Tanaka K. Alternating skew deviation due to hemorrhage in the cerebellar vermis. *Intern Med* 2012;51:2793–2796.
- [11] Ances BM, Dalmau JO, Tsai J, Hasbani MJ, Galetta SL. Downbeating nystagmus and muscle spasms in a patient with glutamic-acid decarboxylase antibodies. *Am J Ophthalmol* 2005;140:142–144.