Ocular Tilt Reaction Caused by a Polycystic Astrocytoma: A Case Report

Thong Le, BOrth&OphthSc(Hons)

Department of Clinical Vision Sciences, La Trobe University, Melbourne, Australia

ABSTRACT

An ocular tilt reaction consists of a vertical misalignment of the eyes (skew deviation), a head tilt and conjugate ocular torsion in the direction of the hypotropic eye. Ocular tilt reaction, however, may be mistaken for a superior oblique palsy due to the similarities in clinical findings of the two conditions. They both display similar vertical deviations and head tilts on presentation. In order to differentiate between the two conditions torsion needs to be assessed and measured. An ocular tilt reaction will display paradoxical conjugate torsion unlike a superior oblique palsy. This paper describes a case of a patient with an ocular tilt reaction that was initially thought to be a superior oblique palsy. An MRI scan revealed a polycystic astrocytoma in the midbrain region consistent with reports in the literature associating midbrain lesions and an ocular tilt reaction. Although rare, an ocular tilt reaction should never be ruled out until torsion is measured in patients presenting with a vertical deviation and head tilt.

Keywords: ocular tilt reaction, polycytic asrocytoma, torsion

INTRODUCTION

The vestibular system, along with the optokinetic and smooth pursuit systems, contributes to eye movements by maintaining clear vision during head movements[1, 2]. The vestibular system consists of a central and peripheral component, where the otolith organs and semicircular canals comprise the peripheral component[2]. There are also connections to the vertical gaze centres in the upper midbrain region[1-3]. The otoliths are responsible for the otolith-ocular reflex that counter-rolls the eyes during a head tilt[2]. Physiological counter-rolling of the eyes produced by the otolith-ocular reflex is characterised by conjugate ocular torsion in the opposite direction of the head tilt (Figure 1). This ocular counter rolling reflex occurs because of the central projections from the semi-circular canals and otoliths to the ocular motor sub-nuclei[1,4]. All of these structures are important in maintaining a stable retinal image. Lesions, such as haemorrhages, abscesses or tumours, that can cause a disturbance in the otoliths or their vestibular connections may interfere with the otolith-ocular reflex which in turn may cause a condition known as an ocular tilt reaction (OTR)[3, 5].

Correspondence: **Thong Le**
Department of Clinical Vision Sciences, La Trobe University, VIC 3086, Australia
Email: tn5le@students.latrobe.edu.au

An OTR consists of a triad of clinical findings including a head tilt, conjugate ocular torsion and a skew deviation[3-6] (a skew deviation being a vertical strabismus caused by disruption of the prenuclear inputs into the oculomotor and trochlear nuclei[1, 2]). However, in OTR it is typical to find the direction of head tilt and ocular torsion are all towards the same side of the hypotropic eye[7] (Figure 2). The ocular torsion is also inconsistent with superior oblique palsy where the hypertropic eye demonstrates extorsion as opposed to the intorsion seen in OTR[3].

This paper presents a 9-year old child found to have a polycystic astrocytoma in the midbrain area causing an OTR. The causes of OTR and the importance of differential diagnosis are discussed.

CASE REPORT

A 9-year old female presented to the clinic displaying a head tilt to the right and complaining of intermittent vertical diplopia. Her mother had noticed the head tilt the day before but could not rule out the possibility of it occurring earlier. The patient's medical history was unremarkable along with her past ocular history. However, there were earlier reports of headaches and nausea but at the time of presentation, the patient was no longer experiencing either of these symptoms.

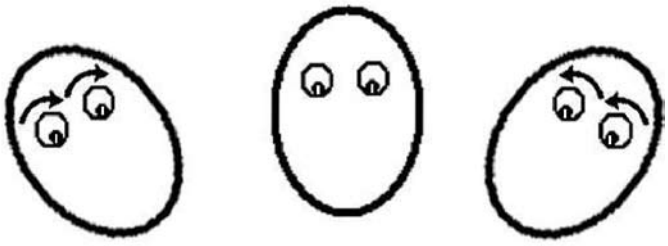

Figure 1. Physiological counter-rolling of the eyes in head tilt. A head tilt to the right causes conjugate torsion towards the opposite direction, that is, intorsion of the right eye and extorsion of the left eye. A head tilt to the left causes the opposite effect.

Figure 2. In OTR, conjugate torsion occurs towards the side of the head tilt and lower eye. In this figure, there is intorsion of the hypertropic eye and extorsion of the hypotropic eye. In a superior oblique palsy there is extorsion of the hypertropic eye instead. This conjugate torsion is also in contrast with the physiological counter roll reflex where there is torsion in the opposite direction of the head tilt.

On examination, uncorrected visual acuities of 6/7.5 OU were recorded. On cover testing, a small right hypotropia was detected. It remained fairly consistent in the 9 positions of gaze. Pupillary reactions were normal. The patient had signs and symptoms suggestive of a left superior oblique palsy. Fundus examination, however, revealed intorsion of the left eye and extorsion of the right eye. This was inconsistent with a left superior oblique palsy where one would expect to find extorsion of the hypertropic eye[3]. The presence of conjugate ocular torsion represents a skew torsion and together with a right head tilt and right hypotropia, led to a diagnosis of OTR. Due to these clinical findings magnetic resonance imaging (MRI) scans were performed and a polycystic astrocytoma was identified in the midbrain region (Figure 3). This was consistent with the diagnosis of an OTR.

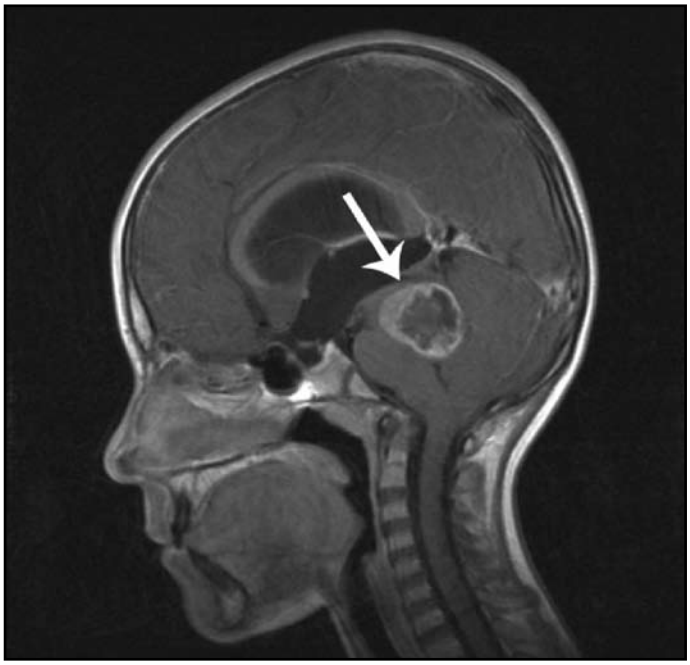

Figure 3. An MRI scan, T1 weighted image, shows the astrocytoma located in the region of the midbrain extending towards the cerebellum (arrow).

DISCUSSION

Westheimer and Blair[7] first described OTR in 1975 in a monkey by stimulating the midbrain regions lateral and dorsolateral to the III and IV nerve nuclei, with the prominent stimulation site being the interstitial nucleus of Cajal (INC). Leuck et al[8] came upon a similar finding, however this time in a human, where a patient underwent insertion of an electrode into the periaqueductal grey area for the relief of chronic pain but instead developed OTR. It was found that the electrode was situated in the INC. Since then, several reports regarding lesions involving the midbrain have been reported to cause an OTR[3, 9-11]. Cases have been described with more localised lesions involving areas outside the midbrain that have been reported to cause an OTR. They include the vestibular nerve[12, 13], cerebellum[5, 14] and utricular nerve[6].

By considering the pathways and structures in the midbrain that are involved in vertical eye movements pathways and how the vestibular system integrates into it, a better understanding of why an OTR occurs can be made. Vertical and torsional saccades are thought to be generated in the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF)[1, 15]. Projecting axons that reach the ocular motor neurons for smooth pursuit originate in the vestibular nuclei. The medial longitudinal fasciculus (MLF) is the important ascending pathway for these projections which carry the neural signals, from medulla to midbrain, for vertical smooth pursuit and vestibular eye movements[1, 15]. Other contributing pathways include the brachium conjunctivum and the ventral tegmental tract, but the MLF is the most important pathway[1, 15]. The vestibular nuclei also combine inputs from the semicircular canals and the otolith organs[1]. These peripheral components of the vestibular system also converge at the ocular motor nuclei to perform their vestibular function in the vertical and roll plane[16].

The INC is the neural integrator for the vertical gaze system[1, 15]. The INC receives input from the riMLF and the vestibular nuclei. It also has many projections which include the oculomotor, trochlear nuclei, contralateral INC, mesencephalic reticular formation (MRF) and also bilaterally to the riMLF. The major burst neurons of the INC involve the mechanisms for gaze-holding and eye coordination in the roll plane[1, 15]. The numerous inputs and projections to and from the INC make it an important neural integrator for vertical gaze and vestibular function. It may be for this reason, disruption to this structure may cause OTR[7, 8].

Table 1. Clinical similarities and differences between OTR and isolated superior oblique palsy.

Clinical Characteristic	OTR Cause	Superior Oblique Palsy
Hypertropia	Yes	Yes
Head tilt	Towards lower eye to correct subjective visual vertical	Towards lower eye to minimise vertical deviation and excylotorsion
Torsion	Intorsion of higher eye and extorsion of lower eye	Extorsion of higher eye

However, lesions including the vestibular nerve[12, 13], cerebellum[5, 14] and utricular nerve[6] have also been reported to cause OTR. This suggests that lesions anywhere along the graviceptive pathways, which are the pathways subserving the vestibulo-ocular reflex involving the semicircular canals and the otolith organs, can cause OTR[3, 16, 17]. The graviceptive pathways begin at the vestibular organs, travel through the brainstem and reach the midbrain to contribute to the vertical gaze structures[16, 18]. The triad of clinical findings in OTR correlates with known function of these pathways. The vertical skew deviation occurs because of the disruption of the vestibular nuclei which is responsible for vertical smooth pursuit and has inputs to the INC for gaze holding[1]. Conjugate torsion occurs because the otolith-ocular reflex is disrupted leaving it unbalanced in the roll plane. This may also explain the head tilt which occurs due to a tilt in the subjective vertical visual and not in order to minimise the vertical deviation[3]. The position of the eyes and head are adjusted to what the vestibular system has mistakenly judged to be the vertical[19].

The triad of clinical findings in OTR may also present a challenge to the examiner because of its similarities to an isolated superior oblique palsy[3, 20]. A recently acquired superior oblique palsy may present with a vertical deviation and a compensatory head tilt to the side of the lower eye, similar to OTR. However, unlike OTR, a superior oblique palsy will display extorsion of the hypertropic eye whereas an OTR will display intorsion of the hypertropic eye. This makes measuring torsion of the hypertropic eye using ophthalmoscopy, fundus photography or the double Maddox rod an important step in differentiating the two conditions [3, 20]. Although isolated inferior oblique palsies are rare, they too may be mistaken with OTR due to the similarity in vertical deviation and head tilt[21]. Measuring ocular torsion will also differentiate the two conditions[21]. The Bielschowsky head tilt test was not used in the assessment of this patient. Donahue, Patrick, Lavin and Hamed[20] reported that the Bielschowsky head tilt test does not definitively differentiate the two conditions and suggested that measuring torsion should be the fourth step added to the '3 step test'. Associated neurological signs like ataxia may also lead the examiner to suspect more widespread brainstem disease rather than isolated cyclovertical muscle palsy[20]. Table 1 summarises these findings.

Furthermore, it is important to differentiate an OTR from an isolated superior palsy because of the difference in treatment implication each condition brings. OTR is often thought to be transient which implies that treatment should remain conservative until spontaneous recovery occurs[3]. Prisms may provide relief from diplopia, but it must be reiterated that these will not alleviate the head tilt. The head tilt in OTR occurs in response to altered subjective visual vertical unlike in superior oblique palsy where a head tilt occurs to minimise the vertical deviation and unilateral excyclotorsion[1, 3, 20, 22]. Furthermore, surgically weakening or strengthening an oblique muscle in a persistent OTR to correct the skew deviation may be contraindicated because of its negative effect on ocular torsion[20, 21]. However, Donahue, Patrick, Lavin and Hamed[20] have reported success in one patient who underwent surgical weakening of an overacting cyclovertical muscle.

Our patient presented with subtle signs of headache and nausea but no other neurological signs or symptoms of poor coordination or balance. The OTR occurring in our patient was most likely due to midbrain pathology involving the INC and its connections with the vestibular nucleus.

REFERENCES

1. Leigh RJ, Zee DS. The neurology of eye movements. 4th ed. New York: Oxford University Press, 2006.
2. Ansons A, Davis H. Diagnosis and Management of Ocular Motility Disorders, 3rd ed. London: Blackwell Publishing 2001.
3. Vaphiades MS. The ocular tilt reaction. Am Orthopt J 2003;53:127-33.
4. Zee DS, Hain TC. Otolith-Ocular reflexes. In: Sharpe JA, Barber HO, eds. The Vestibulo-ocular reflex and vertigo. New York: Raven Press, 1993.
5. Min W, Kim S, J. P, C S. Ocular tilt reaction due to unilateral cerebellar lesion. Neuro-ophthalmol 1999;22:81-85.
6. Ranalli PJ, Sharpe JA. The Vertical vestibulo-ocular reflex. In: Barber HO, Sharpe JA, eds. Vestibular disorders. Chicago: Year Book Medical Publishers, 1988.
7. Westheimer G, Blair SM. The ocular tilt reaction--a brainstem oculomotor routine. Invest Ophthalmol 1975;14:833-839.
8. Lueck CJ, Hamlyn P, Crawford TJ, et al. A case of ocular tilt reaction and torsional nystagmus due to direct stimulation of the midbrain in man. Brain 1991;114:2069-2079.
9. Ohashi T, Fukushima K, Chin S, et al. Ocular tilt reaction with vertical eye movement palsy caused by localised unilateral midbrain lesion. J Neuro-ophthalmol 1998;18:40-42.
10. Nokura K, Ozeki T, Yamamoto H, et al. Posterior canal-type ocular tilt reaction caused by unilateral rostral midbrain hemorrhage Neuro-ophthalmol 2004;28:231-236.

11. Ragge NK, Harris CM, Dillon MJ, et al. Ocular tilt reaction due to a mesencephalic lesion in juvenile polyarteritis nodosa. Am J Ophthalmol 2003;135:249-251.
12. Arbusow V, Dieterich M, Strupp M, et al. Herpes zoster neuritis involving superior and inferior parts of the vestibular nerve causes ocular tilt reaction. Neuro-ophthalmol 1998;19:17-22.
13. Lee H, Lee S-Y, Lee S-R, et al. Ocular tilt reaction and anterior inferior cerebellar artery syndrome. J Neurol Neurosurg Psychiatry 2005;76:1742-3.
14. Mossman S, Halmagyi GM. Partial ocular tilt reaction due to unilateral cerebellar lesion. Neurology 1997;49:491-493.
15. Bhidayasiri R, Gordon GT, Leigh RJ. A hypothetical scheme for the brainstem control of vertical gaze Neurology 2000;54:1985-1993.
16. Brandt T, Dieterich M. Vestibular syndromes in the roll plane: topographic diagnosis from brainstem to cortex. Ann Neurol 1994;36:337-347.
17. Brandt T, Dieterich M. Skew deviation with ocular torsion: a vestibular brainstem sign of topographic diagnostic value Ann Neurol 1993;33:528-34.
18. Brandt TH, Dieterich M. Different types of skew deviation. J Neurol Neurosurg Psychiatry 1991;54:549-550.
19. Brandt T, Dieterich M. Two types of ocular tilt reaction: the 'ascending pontomedullary VOR-OTR and the 'descending' mesencephalic integrator-OTR. Neuro-ophthalmol 1998;19:83-92.
20. Donahue SP, Lavin PJ, Hamed LM. Tonic ocular tilt reaction simulating a superior oblique palsy: diagnostic confusion with the 3-step test. Arch Ophthalmol 1999;117:347-352.
21. Donahue SP, Lavin PJ, Mohney B, Hamed L. Skew deviation and inferior oblique palsy. Am J Ophthalmol 2001;132:751-756.
22. Dieterich M, Brandt T. Ocular torsion and tilt of subjective visual vertical are sensitive brainstem signs. Ann Neurol 1993;33:292-299.