

Paroxysmal Ocular Tilt Reaction Post Gunshot Injury: A Case Report and Review of Literature

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BACKGROUND

Several peripheral and central utricular–ocular pathways play a role in order to maintain a normal ocular motor activity. An imbalance of any of the underlying pathways can cause misalignment of the eyes and possible diplopia and nystagmus (1).

Skew deviation is defined as a vertical misalignment of the eyes that does not map to any cyclovertical muscle, associated with posterior fossa lesion(s) (2). It can be caused by both central and peripheral conditions, the most common cause being brainstem stroke (1). When skew deviation is accompanied with cyclotorsion and abnormal head tilt, then it is referred to as ocular tilt reaction (OTR) (3). Three different types of OTR are described by Brandt and Dietrich (4): Type 1 is Tullio phenomenon that is a disconjugate upward deviation of the eyes caused by a lesion in the utricle, for example, stapes foot subluxation, superior canal dehiscence, etc. Type 2 is hypertropia of one eye (lesion in the dorsolateral medulla oblongata) with predominant excyclotorsion of the ipsilateral side. Type 3 is a simultaneous hypertropia of one eye and hypotropia of the other eye (lesion is in the midbrain tegmentum), which is reflected in cases of paroxysmal OTR in patients with upper brainstem injury. Ocular torsion consists of incyclotorsion of the hypertropic eye with excyclotorsion of the hypotropic eye (3). This article describes a case of paroxysmal ocular tilt reaction occurring years after a sustained gunshot brain injury.

CASE REPORT

We describe a case of a 25-year-old man with a history of right-sided gunshot brain injury in 2014 and residual left-sided hemiparesis and left facial palsy, presented for sudden-onset right eye ptosis and headache. He also reported intermittent untriggered attacks of blurry vision and image jump lasting for few minutes, which improved on right head tilt, happening several times a day.

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Computed tomography (CT) of brain in 2014 revealed a right occipital burr hole, a cortical/subcortical cavity measuring 4 cm containing some bony structures, the biggest measures 9 mm, and a metallic foreign body at the right thalamus reaching the right aspect of the ventral midbrain. Medial structures are in place. Bony fragments and foreign bodies are present at the level of the temporal lobe. Further imaging with MRI was not possible due to the presence of metallic foreign bodies (Fig. 1).

Attacks started 5 years after gunshot injury. Witnessed during the examination process, they were of no predictable pattern and could not be elicited by any mean. They could last anywhere from 30 seconds up to 5 minutes. During the attack, the patient exhibited a left hypertropia of 40–45 prism diopters (see Video, **Supplemental Digital Content 1**, <http://links.lww.com/WNO/A513>). In addition to a fast

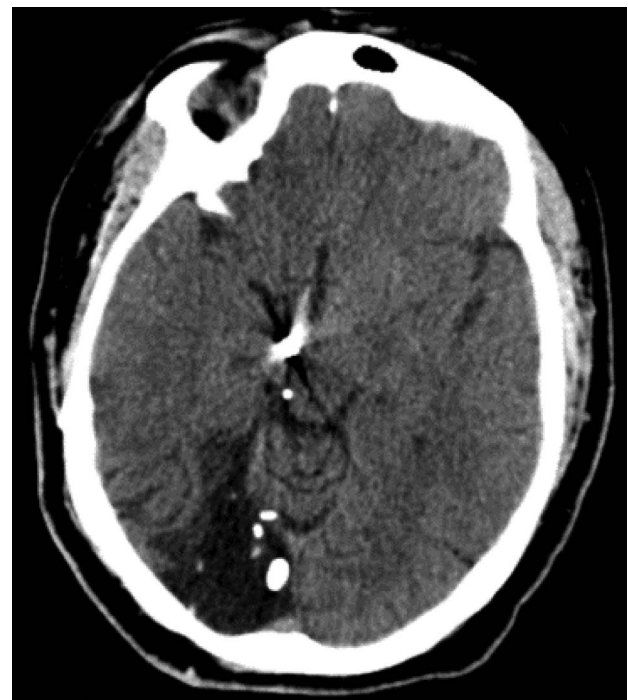


FIG. 1. Computed tomography of brain (infratentorial region) showing right occipital burr hole, a cortical/subcortical cavity measuring 4 cm containing some bony structures, the biggest measures 9 mm, and a metallic foreign body at the right thalamus reaching the right aspect of the ventral midbrain.

conjugate torsional nystagmus with the upper poles beating to the right, there was a 5° conjugate ocular torsion (right excyclotorsion and left incyclotorsion) and a right head tilt (Figs. 2, 3). Enophthalmos of the right eye was confirmed by Hertel exophthalmometry. Right eye ptosis was more pronounced during the episode with a margin reflex distance (MRD1) of 0 mm. During the attack, visual acuity decreased from 20/20 to 20/40 in both eyes. *Worth four dot test* reflected alternate suppression during attack, as the patient saw 3 green lights or 2 red lights, depending on the fixing eye. Outside the attack, the test showed fusion into one image (normal), and the patient saw 4 dots.

The patient had a normal slit-lamp examination, equal pupil sizes with no afferent pupillary defect, normal intraocular pressure, unremarkable dilated fundus examination, and full ductions. Optic nerve and macula were noted to be within normal limits on optical Coherence tomography. Repeat CT brain did not display any significant changes that could explain the new onset of symptoms. Electroencephalogram was negative for abnormal electrical activities. In addition, blood test for lead level was taken at a later follow-up, and it turned out to be 1.54 µg/dL.

A trial of beta-blockers for 4 weeks did not show any improvement. Carbamazepine was started, and patient improved clinically after 6 weeks of treatment.

DISCUSSION

We present a patient with sudden-onset intermittent OTR: skew deviation, head tilt to the right, and ocular torsion. Unlike trochlear nerve injury, which is the most common posttraumatic cause of vertical misalignment, our patient displayed excyclotorsion of the hypotropic eye and conjugate torsion of both eyes. The pathway for the otolithic-ocular response projects from the vestibular end organ to the vestibular nuclei in the medulla and on to the interstitial nucleus of Cajal in the midbrain. This pathway decussates in the pons: hence, static OTRs from hypofunction are *ipsiversive* with peripheral vestibular and pontomedullary lesions and *contraversive* with pontomesencephalic lesions (3). In addition, ptosis seen in our patient is hypothetically a pseudoptosis related to the intermittent hypotropia of the



FIG. 2. Image of the patient showing hypotropia of the right eye as part of the episodic attacks related to his paroxysmal skew deviation.



FIG. 3. Image of the patient showing eye movement back to baseline after few minutes.

right eye rather than a true ptosis, which is unlikely to manifest in skew deviation.

Only a handful cases of paroxysmal skew deviations have been reported in the literature. Radtke et al reported the case of a young woman presenting with paroxysmal alternating skew deviation accompanied by left beating, horizontal, torsional nystagmus, after biopsy of the inferior cerebellar vermis for suspected brainstem glioma, resulting in destruction of the uvula (5). Rabinowitz et al described paroxysmal skew deviation with concurrent attacks of elliptical pendular oscillations of variable duration in a patient with multiple sclerosis (6). Lawden et al described a patient with an arteriovenous malformation causing damage to the left vestibular nuclei, in whom attacks of paroxysmal torsional horizontal and vertical nystagmus to the left occurred regularly at 2-minute intervals lasting 15 seconds and causing oscillopsia (7). Hedges et al reported a patient with a focal brainstem abscess in the region of the zona incerta who developed a chronic oculocephalic dyskinesia in which paroxysmal tonic skew deviation and torsion of the eyes were coupled with ocular oscillations and head tilt (8).

All the cases described above are similar in one way or another to our presented case, being paroxysmal skew deviation associated with nystagmus. However, our case is the first to report a traumatic paroxysmal, nonalternating, nonperiodic skew deviation with unpredictable trigger associated with unilateral nystagmus. This presentation correlates with CT brain findings of a midbrain foreign body likely responsible for disturbance of underlying supranuclear pathways and hence the clinical picture, as depicted by Greenberg et al who presented the case of an elderly male with nonalternating skew deviation, rotatory nystagmus, head tilt, and lid retraction with CT scan showing a right midbrain lacunar infarct in the area of interstitial nucleus of Cajal (9). Nonetheless, that patient exhibited periodic intermittent diplopia, lasting 2–3 minutes followed by 1–2 minutes of quiet period, unlike our patient who had nonperiodic attacks.

Among the differential in our case is ocular neuro-myotonia (ONM), a rare clinical entity, triggered by prolonged eccentric gaze and characterized by episodic diplopia, as a result of tonic contractions or spasms of the extraocular muscles (10). ONM usually affects cranial nerve VI and is most commonly related to radiation therapy to the

brainstem, but ONM involving CNIII has been described. ONM is less likely to be the case here due to the bilaterality of the presentation, whereas most cases of ONM are unilateral (11). Moreover, we expect with ONM to have lid retraction rather than ptosis as the levator muscle is stimulated (12). In addition, while treatment of ONM by carbamazepine might cause nystagmus as a side effect, ONM by itself is not known to be associated with nystagmus or oscillopsia. Other differential diagnoses of acquired episodic diplopia include superior oblique myokymia, convergence spasm, and ocular myasthenia gravis (11), which do not fit the clinical picture.

It is reasonable to check lead levels as a contributor for OTR. Blood lead level in our patient was $1.54 \mu\text{g/dL}$ (normal range is defined as less than $10 \mu\text{g/dL}$ (13)). Therefore, lead level in our patient is within normal limits, and thus, it does not contribute to the delayed paroxysmal OTR.

Skew deviation in OTR can be managed with prisms, pharmacotherapy, or surgery. Binocular vertical diplopia in skew deviation can be treated with temporary patching or prisms if the deviation is small and comitant. Other OTRs were treated effectively with carbamazepine (6). Patient's objective symptoms improved after carbamazepine trial reinforcing our diagnosis of OTR. Nevertheless, it is uncertain whether this improvement is due to the clinical effect of the drug or an acquired suppression as detected on *Worth four dot test*. On examination, the frequency of the attacks were less but with little if any symptoms when they occur. For larger deviations or ones associated with ocular torsion, vertical rectus recession or resection is recommended (2); however, its role in this case is questionable given the intermittency of symptoms.

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