

Ipsiversive Partial Ocular Tilt Reaction in a Patient with Acute Paramedian Thalamic Infarctions

Chien-Ming Chen¹ and Sung-Hsiung Lin²

Abstract- The ocular tilt reaction (OTR) consists of skew deviation, ocular torsion and head tilt. A 54-year-old woman developed sudden onset of vertical diplopia. On primary gaze, there was skew deviation with the left eye higher than the right eye. The photography of fundus disclosed 15 degrees of excyclotropia of the right eye and 20 degrees of incyclotropia of the left eye. There was no motor deficit, sensory impairment, ataxia or changes in consciousness. Brain MRI, including T2WI, FLAIR and DWI, revealed two lesions of high signal intensities in bilateral paramedian thalamus, with the much larger and brighter one on the right side. These findings constituted an ipsiversive partial OTR, i.e. skew and torsion toward the side of the lesion. OTR as the only manifestation of paramedian thalamic stroke is rare. A previous report by Dieterich and Brandt indicated that if an OTR occurred in a paramedian thalamic infarct, there should be concurrent ischemia of the interstitial nucleus of Cajal, and it was always contraversive. In contrast, the lesions in our case were quite localized in the paramedian thalamus, not extending into the midbrain. In addition, this report demonstrated an OTR could be ipsiversive under such conditions, opposite to the direction mentioned in previous reports.

Key Words: Ocular tilt reaction, Skew deviation, Ocular torsion, Head tilt, Paramedian thalamic infarction

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INTRODUCTION

Skew deviation is a vertical malalignment of the eyes that cannot be explained on the basis of an ocular muscle palsy. It results from an imbalance in tonic signals within the otolith-ocular reflex pathways. Patient with a skew deviation complains of vertical diplopia,

and usually has a cyclorotation of both eyes associated with an illusion of tilt of the visual world (subjective visual vertical tilt, SVV tilt). The head may also be tilted, toward the side of the lower eye. The triad skew deviation, ocular torsion (OT) and head tilt together form the ocular tilt reaction (OTR)⁽¹⁾.

The OTR can occur with lesions anywhere along the

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otolith-ocular pathway, including the labyrinth, vestibular nerve, medial longitudinal fasciculus (MLF), pons, and midbrain. Either complete OTR or skew torsion without head tilt indicates a unilateral deficit of otolith input or a unilateral lesion in the graviceptive pathways from the vestibular nuclei to the interstitial nucleus of Cajal (INC). The pathway crosses at the lower pontine level where the vestibular nuclei are localized. The OTR is ipsiversive (ipsilateral eye lowermost, ocular torsion and head tilt toward the side of lesion) with unilateral peripheral or pontomedullary lesions below the decussation, indicating the involvement of the medial and/or superior vestibular nuclei. The OTR is contraversive (contralateral eye lowermost, ocular torsion and head tilt away from the side of lesion) in unilateral pontomesencephalic brainstem lesions above the decussation, indicating the involvement of the MLF or INC and riMLF (rostral interstitial nucleus of MLF).

Unilateral lesions of the vestibular structures rostral to the INC typically are presented with only SVV tilt without concurrent eye-head tilt (OTR or skew torsion)²⁻⁴. According to the study by Dieterich and Brandt et al., ipsiversive or contraversive SVV tilt may occur in posterolateral thalamic lesions. Contraversive OTR may occur in paramedian thalamic lesions but it is the result of concurrent involvement of the INC². However we report a patient with paramedian thalamic infarctions exhibiting a partial OTR that is ipsiversive, the opposite direction to the previous report.

CASE REPORT

A 54-year-old woman with a 5-year history of hypertension was admitted for sudden onset of vertical diplopia on April 1, 2004. She was clear and alert in mind, with one eye closed intentionally to avoid the discomfort of binocular diplopia. Except the ocular abnormality, general physical and neurological examinations were unremarkable, without motor deficit, sensory impairment or ataxia. Skew deviation with the left eye higher was noted on primary gaze straight ahead (Fig. 1a). The ranges of vertical and horizontal eye movements appeared full and ptosis was absent. Pupillary size was symmetric and reactivity to light was normal. In fundus photographs, taken on the 6th day of the illness, there were 15 degrees of excyclotropia of the right eye and 20 degrees of incyclotropia of the left eye (Fig. 1b), determined by the angle between the papilla-fovea meridian and the horizontal line². The brain CT performed at the onset day was unremarkable. The brain MRI five days later revealed two lesions of high signal intensities, as seen on T2WI, FLAIR and DWI, in bilateral paramedian thalamus, with the much larger and brighter one on the right side (Fig. 2). In addition to hypertension, the evaluation of vascular risk factors indicated overweight and hypercholesterolemia (249 mg/dl). Carotid duplex study was normal. She was treated as ischemic stroke, with aspirin 100 mg once daily and pentoxifylline 400 mg twice a day orally. Hypertension and

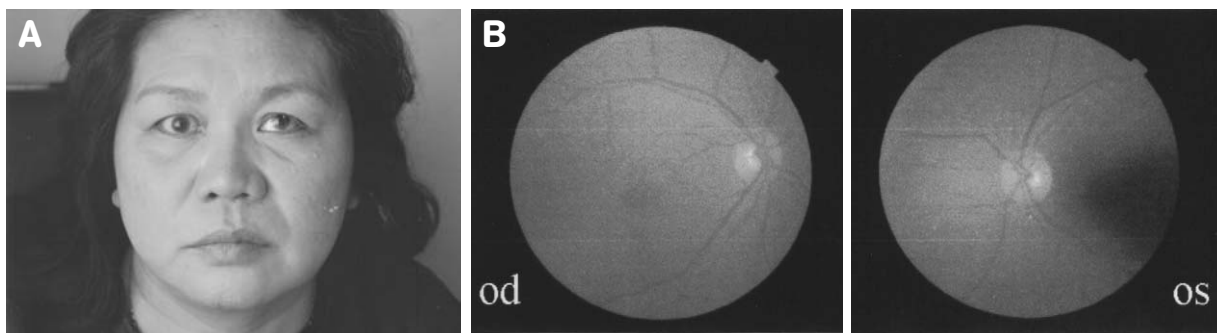


Figure 1. (A) Picture taken on the second day after the onset: skew deviation with the left eye at a higher position than the right one on primary gaze. (B) In fundus-photographs, taken on the 6th day of the illness, there were 15 degrees of excyclotropia of the right eye and 20 degrees of incyclotropia of the left eye, determined by the angle between the papilla-fovea meridian and the horizontal line.

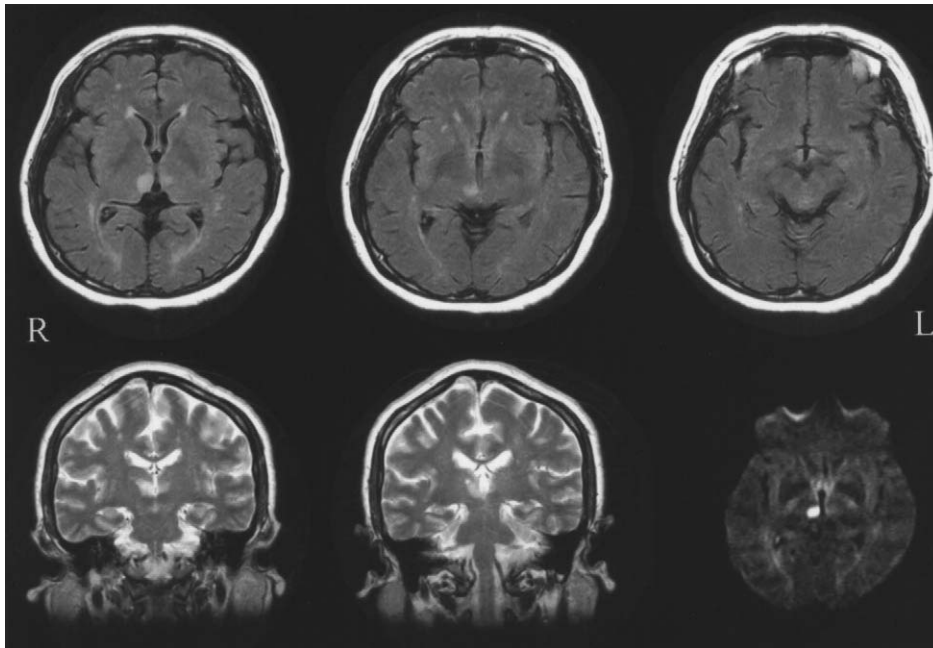


Figure 2. Brain MRI on the 6th day (April 6, 2004): axial FLAIR and DWI, and coronal T2WI images revealed lesions of high signal intensity in bilateral paramedian thalamus, with the much larger and brighter one on the right side, not extending into the midbrain.

hypercholesterolemia were managed with accupril 10 mg and atorvastatin 10 mg per day respectively.

The severity of vertical diplopia lessened two weeks later. The ocular malalignment also abated gradually, which disappeared within four months.

DISCUSSION

The most comprehensive studies about the correlation between the direction of the OTR and the human lesions along the otolith-ocular pathway came from the works by Dieterich and Brandt et al. It is always ipsiversive in lateral medullary infarctions, and always contraversive in mesencephalic lesions. Pontine lesions can cause either ipsiversive or contraversive OTR, depending on whether the location is below or above the decussation^(2,3,7,8). An unilateral cerebellar lesion may give rise to a partial contraversive OTR, because the otolith-ocular reflex is under the inhibitory control of the ipsilateral caudal cerebellum, possibly the nodulus⁽⁶⁾. All skew deviations were associated with concomitant ocular tor-

sion and tilts of subjective visual vertical toward the undermost eye. Skew deviation or more correctly, ocular skew torsion is a sensitive brainstem sign of localizing and lateralizing value⁽⁵⁾. The hypothetical explanation of the OTR by the lesions of the vertical semicircular canal pathways in the brainstem is illustrated in Fig. 3^(3,4).

For the lesions rostral to the brainstem, Dieterich and Brandt analyzed 35 patients with acute thalamic infarctions (17 posterolateral, 14 paramedian, 4 polar) and their effects on vestibular function in the roll plane. None of 17 patients with posterolateral infarctions had complete OTR, but 11 exhibited SVV tilt (7 ipsilateral, 4 contralateral) and 5 exhibited slight OT (4 ipsilateral, 1 contralateral), none had head tilt. In the 11 cases with SVV tilt, vestibular thalamic nuclei (nucleus ventro-oralis intermedius, nucleus ventro-caudalis externus, and nucleus dorso-caudalis) were involved. Eight of 14 patients with paramedian infarctions had complete OTR, all contraversive. Those with polar infarctions had no effect on SVV, OT, skew deviation or head tilt. By the projections of the infarcted areas in CT scans or MR

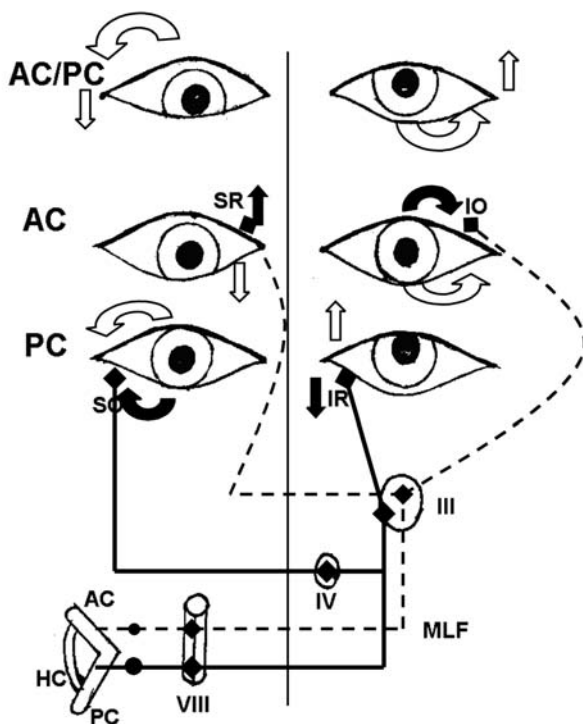


Figure 3. Hypothetical explanation of the OTR by the lesions of the vertical semicircular canal pathways in the brainstem. The excitatory actions by the pathways on the extraocular muscles are expressed as solid arrows and the counteractions by the lesions in the pathways are expressed as open arrows. The actions by the rectus muscles are depicted as straight arrows because of their primary actions are elevation (SR) and depression (IR), those by the oblique muscles are depicted as curved arrows because of their primary actions are intorsion (SO) and extorsion (IO) respectively. An excitatory ascending pathway from the PC activates the ipsilateral SO and the contralateral IR (solid line). A destructive lesion in this pathway causes extorsion of the ipsilateral eye and hypertropia of the contralateral eye. Another excitatory pathway from the AC activates the ipsilateral SR and contralateral IO (dotted line). A destructive lesion in this pathway causes hypotropia of the ipsilateral eye and intorsion of the contralateral eye. A combined defect of both PC and AC pathways gives rise to a complete OTR, with skew deviation, ocular torsion and head tilt in the same direction, the direction ipsilateral to the semicircular canals. III: oculomotor nucleus; IV: trochlear nucleus; VIII: vestibular nucleus; MLF: medial longitudinal fasciculus; SO: superior oblique; IR: inferior rectus; SR: superior rectus; IO: inferior oblique; AC: anterior semicircular canal; HC: horizontal semicircular canal. (modified from references: 3 & 4)

images onto the appropriate transverse sections of stereotaxic atlases, they concluded that the posterolateral infarctions exhibiting SVV tilt should involve the vestibular thalamic nuclei, and that those OTRs by paramedian infarctions were not thalamic actually, but due to concurrent ischemia of the rostral midbrain tegmentum including the INC, which constitutes the most rostral structure to cause the eye-head synkinesis. An INC lesion with OTR is contraversive, but a posterolateral thalamic lesion with SVV tilt can be ipsiversive or contraversive presumably related to bilateral vestibular input to the thalamus^(1,9). Several studies of electrical stimulation or surgical destruction, from animal or human, also supported the role of the INC, whereas stimulation of it induced ipsiversive OTR and destruction of it induced contraversive OTR⁽¹⁰⁻¹³⁾. OTR was paroxysmal when it was ipsiversive in the reported meso-diencephalic lesions, caused by multiple sclerosis, meso-diencephalic abscess or infarct^(14,15). The only difference between

ipsiversive and contraversive OTRs appeared to be the temporal pattern: an ipsiversive OTR was always paroxysmal (phasic), and a contraversive OTR was always persistent (tonic)⁽⁸⁾. Tonic contraversive OTR could be due to disfacilitation or reduced spontaneous activity of INC neurons whereas paroxysmal ipsiversive OTR could be due to disinhibition or increased spontaneous activity of INC neurons⁽⁸⁾.

The paramedian thalamus is supplied by the paramedian thalamic arteries, which also supply the rMLF and INC, arising from the proximal P1 peduncular segment of the posterior cerebral artery. In about one-third of brains, they arise from one side or from a common pedicle⁽⁹⁾, and bilateral paramedian infarcts may occur in a single vascular event. In our case, the thalamic infarctions were quite limited to the paramedian portions, and did not extend into the midbrain. The much larger right one brought on a tonic ipsiversive partial OTR (skew deviation and OT). Even from the possibility of the indi-

rect effect (compression or edema) to affect the INC or the posterolateral thalamus, our case did not obey the Dieterich and Brandt's theory, which claimed an INC lesion caused tonic contraversive or phasic ipsiversive OTR and a posterolateral thalamic lesion caused only SVV tilt. An even lower possibility was the left smaller one induced a contraversive OTR, but it was much far away to affect the INC. Vestibular input to the thalamus was thought to be the posterolateral portion. In a rat study using stereotaxic tracer injection, there is a projection from the INC to the intralaminar nucleus⁽¹⁶⁾, which has connection to the medial nucleus of the thalamus⁽⁴⁾. This might explain the presentation of the OTR in our patient. Possibly the projection is bilateral, like that to the posterolateral thalamus, and the ipsiversive one is an alternative. Neither an INC involvement is necessary for the paramedian thalamic lesion with an OTR, nor the OTR in a paramedian thalamic lesion is necessarily contraversive.

OTR as the only manifestation of paramedian thalamic stroke is rare. Our case demonstrates the opposite direction to that of the previous reports. OTR often goes unrecognized because (1) severely ill bedridden patients may have an unnoticed head tilt, especially while supine; (2) OT can be diagnosed objectively only with fundus photography; and (3) the most obvious sign, skew deviation, is a subtle ocular abnormality that requires a high index of suspicion and an experienced examination for diagnosis⁽¹⁷⁾.

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