CASE REPORT

Ocular Tilt Reaction due to a Cerebellar Hemorrhage

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Abstract

A 78-year-old man with essential hypertension abruptly developed complete ocular tilt reaction (OTR) which consisted of concomitant skew deviation with left hypertropia, extorsion of the right eye and intorsion of the left, and rightward head tilt. Cranial computed tomography demonstrated a localized cerebellar hemorrhage involving the left nodulus. The patient became asymptomatic within two weeks. This is a first reported case of complete OTR due to a cerebellar hemorrhage. Concomitant skew deviation is a common symptom of cerebellar lesions. Moreover, unilateral damage to the utricular pathway due to involvement of the left nodulus might cause rightward conjugate ocular torsion and rightward head tilt.

Key words: nodulus, ocular torsion, skew deviation, utricular pathway

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Introduction

Skew deviation is a term that refers to an acquired vertical divergence of the eyes due to supranuclear dysfunction (1, 2). Ocular tilt reaction (OTR) is defined as a triad of skew deviation, conjugate ocular torsion, and head tilt, commonly caused by brainstem lesions (2). In particular, concomitant skew deviation is a poorly localizing sign of posterior fossa dysfunction and commonly induced by cerebellar lesion (1, 3-6). However, there have so far been only a few reported cases of conjugate ocular torsion with or without head tilt due to a cerebellar lesion (7-14), and its precise pathogenesis remains unclear. We herein describe the first reported case of complete OTR due to a cerebellar hemorrhage.

Case Report

A 78-year-old Japanese man with essential hypertension abruptly presented with nausea, dizziness, and vertical diplopia in October 2012. The following day, the patient was admitted to our hospital. Corrected visual acuities were 0.8 in each eye. Funduscopic examination demonstrated cataracts in both eyes. The pupil diameter was 3 mm bilaterally in the lightened room. The pupil responses to light and near were prompt in either eye. Palpebral fissure measured 8.5 mm bilaterally. Two prism diopter of exotropia with 6 of left hypertropia was detected in the primary position. Rightward conjugate ocular torsion, which consisted of extorsion of the right eye and intorsion of the left eye, was detected by the Maddox rod test. Concomitant skew deviation with left hypertropia was observed (Fig. 1) and thereafter was confirmed by the red glass test. Rightward head tilt was observed. From these results, the patient was diagnosed to have complete OTR. In addition, gaze-evoked horizontal nystagmus during leftward gaze was observed. There were no other neurologic abnormalities. The complete blood cell count and blood chemistry findings were within the reference ranges. Electrocardiogram and chest radiograph examinations demonstrated normal findings. Cranial computed tomography demonstrated a localized cerebellar hemorrhage involving the nodulus, tonsil, and anterior and posterior paravermis on the left side (Fig. 2). Under blood pressure management and rehabilitation, his neurological symptoms gradually resolved within two weeks.

Discussion

In 1964, Smith et al. (3) reported a case of concomitant skew deviation after surgery for a primary non-malignant tumor in the right cerebellum. Keane (1) described 3 cases of

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concomitant skew deviation due to cerebellar tumor without brainstem involvement, though the detailed localization of the lesions in the cerebellum was not noted. Brennan et al. (4) reported that, of 12 cases of cerebellar hemorrhage, concomitant skew deviation was observed in 5 cases. However, in these previous articles (1, 3, 4), the neuroimaging findings were not shown. Based on the analysis of cranial magnetic resonance imaging findings in 5 cases of skew deviation, Wong et al. (5) and Chandrakumar et al. (6) stated that concomitant or incomitant skew deviation could be caused by focal cerebellar lesion, and no correlation between the laterality of the hypertropic eye and the side of unilateral cerebellar lesions was found.

In 1977, Rabinovitch et al. (15) reported the first case presenting with a triad of skew deviation, conjugate ocular torsion, and head tilt secondary to multiple sclerosis, and
proposed the term ‘ocular tilt reaction (OTR)’. Regarding its etiology, paroxysmal activation of brainstem utricular projections were speculated (15). Brodsky et al. (2) and Brandt et al. (16, 17) stated that OTR might be caused by asymmetrical utricular neuronal input owing to interruption of the peripheral or central utricular pathways; a unilateral utricular imbalance therefore produced OTR in the roll plane. The utricular signals are relayed from the vestibular nuclei, medullary reticular formation, inferior olive, and lateral reticular nucleus to the nodulus and uvula in the cerebellum, and influence the deep cerebellar nucleus (18). Consequently, OTR can be induced by involvement of the cerebellum, though its precise responsible region remains controversial (2, 7-14).

Lee et al. (7) reported 2 cases of incomplete OTR secondary to unilateral infarction of the anterior inferior cerebellar artery, which consisted of skew deviation and ipsiversive conjugate ocular torsion. The authors speculated that damage to the inner ear of the root entry zone of the cranial eighth nerve might be responsible for incomplete OTR (7). Baier et al. (8, 9) noted that, among 43 cases of purely unilateral cerebellar infarction, 9 cases showed complete OTR. Therefore, the authors proposed that OTR may be a common symptom in patients with unilateral cerebellar lesions, and lesions of the cerebellum induced a dysfunction in utricular pathways that mediated vestibular information in the roll plane. Moreover, based on an analysis of 56 cases of cerebellar infarction, an affection of the dentate nucleus in particular was associated with the contralateral sign of OTR. In contrast, when the dentate nucleus was spared and the lesions were located in the middle cerebellar peduncle, tonsil, biventer, and inferior semilunar lobules, OTR was observed ipsilaterally (8, 9).

There have only been 3 reported cases of isolated bilateral nodular infarction (10, 11). In one case, conjugate ocular tilt with head tilt was observed, despite no skew deviation (11). The authors speculated that leftward conjugate ocular tilt as well as leftward head tilt might be caused by asymmetrical damage to the nodulus, specifically to the right than to the left side (11).

Mossmann et al. (12) described 2 cases of partial OTR. In one case, skew deviation with right hypertropia and rightward conjugate ocular torsion were caused by left caudal cerebellar hemorrhage involving the left nodulus. In another case, rightward conjugate ocular torsion and leftward body lateropulsion were induced by infarction of the left medial branch of posterior inferior cerebellar artery involving the left tonsil, biventer lobules, and left inferior vermis including the left nodulus. Moreover, Min et al. (13) reported 2 cases of incomplete OTR, which consisted of ocular torsion and head tilt due to a cerebellar lesion. Skew deviation was not observed in either case. In one case, there was a left caudal cerebellar hemorrhage involving the nodulus and dentate nucleus on the left side. In another case, neuroimaging demonstrated infarction of the left inferior cerebellar artery involving the nodulus and uvula as well as the vestibular nucleus on the left side (13). Consequently, Mossmann et al. (12) and Min et al. (13) speculated that the mechanism of contraversive conjugate ocular torsion secondary to unilateral cerebellar lesion might be an increased tonic resting activity in the ipsilesional vestibular nucleus due to a loss of inhibition from the lesioned nodulus. Kim et al. (10) noted that 3 cases of unilateral posterior inferior cerebellar artery infarction with ipsilateral nodulus involvement presented with skew deviation with hypertropia of the contralateral eye as well as contraversive conjugate ocular torsion. In contrast, in 10 cases of unilateral posterior inferior cerebellar artery infarction without ipsilateral nodulus involvement, none of them presented with either skew deviation or abnormal ocular torsion (10). Moon et al. (11) reported 6 cases of isolated unilateral nodular infarction. All of these cases developed body lateropulsion toward the opposite side of the lesion. One case presented with conjugate ocular torsion as well as head tilt toward the opposite side of a nodular infarction. The authors speculated that conjugate ocular torsion and head tilt might thus be caused by asymmetry in the tonic resting activity of secondary otolithic neurons attributable to asymmetrical nodulo-vestibular inhibition (11). As a result, these reported cases (10-14) noted that unilateral nodular involvement might cause contraversive conjugate ocular torsion with or without leftward head tilt.

In our patient, complete OTR was observed, which consisted of concomitant skew deviation with left hypertropia, rightward conjugate ocular torsion and rightward head tilt. We speculated that rightward conjugate ocular torsion with rightward head tilt might therefore result from damage to the left nodulus. However, to date there has been no reported case of skew deviation secondary to isolated nodular lesion. Concomitant skew deviation is a common symptom in patients with cerebellar lesions (1, 3-6), though its responsible region remain unclear. Therefore, in our patient, we speculated that concomitant skew deviation might have been induced by a localized cerebellar damage in the vicinity of the left nodulus. In conclusion, we emphasize that this is the first reported case of complete OTR due to a cerebellar hemorrhage.

The authors state that they have no Conflict of Interest (COI).

References

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