

Alternating Skew Deviation due to Hemorrhage in the Cerebellar Vermis

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Abstract

A 76-year-old Japanese woman with essential hypertension and diabetes mellitus abruptly presented with nausea, dizziness, an occipital headache, truncal ataxia, gaze-evoked nystagmus and alternating skew deviation (ASD) with abducting eye hypertropia. Cranial computed tomography demonstrated hemorrhage in the cerebellar vermis and its vicinity. These symptoms gradually resolved within three weeks. This is the first reported case of ASD secondary to cerebellar hemorrhage without hydrocephalus. The vertical misalignment of the eyes during the right-sided gaze was consistently larger than during the left-sided gaze. We speculated that bilateral and asymmetrical damage to the utricular pathway due to the bilateral involvement of the nodulus and uvula might have caused the ASD.

Key words: cerebellar hemorrhage, nodulus, ocular tilt reaction, otolith, utricular pathway, uvula

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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). Alternating skew deviation (ASD) is a subtype of skew deviation with a frequency of 12% (3), in which the side of the higher eye changes depending upon whether the gaze is directed to the left or to the right side (2-6). In ASD, the abducting eye is commonly hypertropic (2-6), which may be produced by a relative inferior rectus muscle weakness (4) or overactive superior oblique muscle (5). Regarding the possible cerebellar causes of ASD, a tumor (5), Arnold-Chiari malformation (4, 6), various degenerative diseases (4, 6), lithium overdose with permanent cerebellar dysfunction (3) and hemorrhage in the superior vermis with hydrocephalus (3) have been reported. However, its pathogenesis remains uncertain. We herein describe a case of ASD secondary to a hemorrhage in the cerebellar vermis and its vicinity without hydrocephalus.

Case Report

A 76-year-old Japanese woman with essential hyperten-

sion and diabetes mellitus abruptly presented with nausea, dizziness, a persistent occipital headache, and unsteadiness in December 2011. Three days later, the patient was admitted to our hospital. Horizontal gaze-evoked nystagmus bilaterally on lateral gaze and marked truncal ataxia with mainly backward falling were observed. She complained of slight bilateral vertical diplopia upon viewing the lateral sides. Her corrected visual acuities were 1.2 in both eyes. A fundoscopic examination demonstrated diabetic retinopathy in both eyes. Her pupil diameter was 3 mm bilaterally in a lightened room. The pupil responses to light and the near response of the pupil were prompt in both eyes. The palpebral fissure measured 8.5 mm bilaterally. In the primary position, despite 2 prism diopters of exotropia, vertical misalignment of the eyes was not detected. The Maddox rod test demonstrated normal ocular torsion in both eyes. Spontaneous head tilt was not observed. However, vertical misalignment of eyes was the most apparent in lateral gaze down. In addition, vertical misalignment of eyes on the right lateral gaze was the most impressive in the down gaze, which was confirmed by the red glass test for vertical diplopia. Based on these results, the patient was diagnosed to have ASD with abducting eye hypertropia (Fig. 1). There were no other neurologic abnormalities.

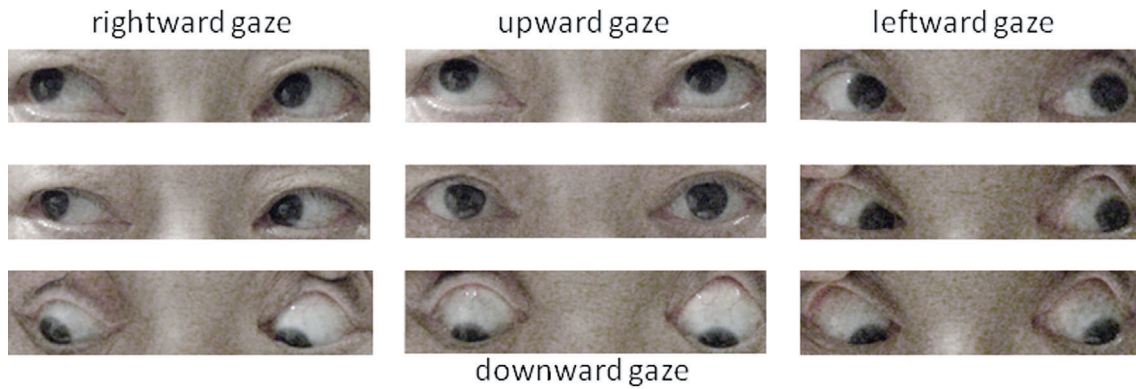


Figure 1. Alternating skew deviation with abducting eye hypertropia was observed.

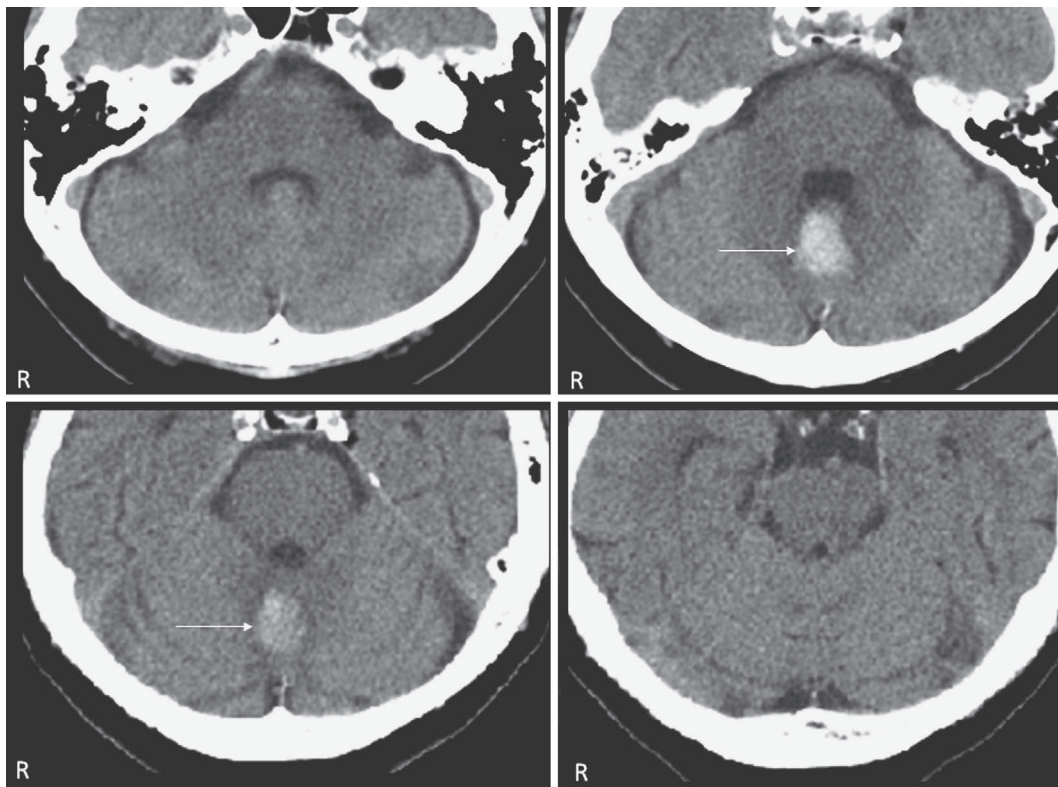


Figure 2. Cranial computed tomography demonstrated a hemorrhage in the cerebellar vermis and its vicinity (arrow).

The patient's complete blood cell count was within the reference ranges. A blood chemistry analysis demonstrated that the HbA1c was 8.8%. An electrocardiogram and chest radiography examination demonstrated normal findings. Cranial computed tomography demonstrated a hemorrhage in the cerebellar vermis and its vicinity without hydrocephalus or a brainstem lesion (Fig. 2). Under blood pressure management and rehabilitation, her neurological symptoms were gradually resolved within three weeks. During her clinical course, the vertical misalignment of her eyes during the right-sided gaze was consistently larger than during the left-sided gaze.

Discussion

Smith et al. (7) reported a case of comitant skew deviation after surgery for a primary non-malignant tumor in the right cerebellum. Keane (1) described three cases of comitant skew deviation due to cerebellar tumor without brainstem involvement, although the detailed localization of the lesions in the cerebellum was not described. Brennan et al. (8) reported that, of 12 cases of cerebellar hemorrhage, comitant skew deviation was observed in five cases. However, in these previous articles (1, 7, 8), the neuroimaging findings were not shown. Based on the analysis of the cranial magnetic resonance imaging findings in five cases of

skew deviation, Wong et al. (9) and Chandrakumar et al. (10) stated that comitant or incomitant skew deviation could be caused by focal cerebellar lesions, and no correlation between the laterality of the hypertropic eye and the side of unilateral cerebellar lesions was found.

In 1977, Rabinovitch et al. (11) reported a case presenting with a triad of conjugate ocular torsion, head tilt and skew deviation secondary to multiple sclerosis, and proposed the term 'ocular tilt reaction' (OTR). Regarding its etiology, paroxysmal activation of brainstem utricular projections was proposed (11). 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 (12). Consequently, OTR can be induced by involvement of the cerebellum, although the precise region responsible still remains controversial (13-20). Lee et al. (13) reported two cases of OTR secondary to unilateral infarction of the anterior inferior cerebellar artery, and indicated that damage to the inner ear of the root entry zone of the cranial eighth nerve might be responsible for OTR. Baier et al. (14, 15) stated that, based on an analysis of 56 cases of cerebellar infarction, an effect on the dentate nucleus was associated with the contralateral sign of OTR. In contrast, when the dentate nucleus was spared and lesions were located in the middle cerebellar peduncle, tonsil, biventer, and inferior semilunar lobules, OTR was observed ipsilaterally (14, 15). Moreover, unilateral involvement of the nodulus may be a cause of OTR (16-20). There have been only three reported cases of isolated bilateral nodular infarction (19, 20). Among these, OTR was observed in one case, probably because of asymmetrical damage to the nodulus (20).

Because ASD due to a cerebellar lesion has been rarely reported (3-6, 21), its pathogenesis remains unclear. Moster et al. (4) reported 10 cases of ASD due to cerebellar diseases, in which there was an Arnold-Chiari malformation in six cases, spinocerebellar degeneration in three cases, and alcoholic cerebellar degeneration in one case. However, they did not discuss the region responsible for ASD in detail (4). Hamed et al. (5) described two cases of ASD secondary to tumors involving the cerebellar vermis and tonsils. Versino et al. (6) and Zee (21) wrote that damage to the dorsal vermis and underlying posterior fastigial nucleus might cause ASD, based on cases of Arnold-Chiari malformation and cerebellar degenerative diseases. Keane (3) reported two cases of ASD due to cerebellar lesions, which were caused by a lithium overdose with permanent cerebellar dysfunction and superior cerebellar vermis hemorrhage, respectively, although neuroimaging findings were not shown. In cases of superior cerebellar vermis hemorrhage, secondary hydrocephalus might induce ASD (3). In contrast, Brodsky et al. (2) and Brandt et al. (22, 23) stated that OTR might be caused by asymmetrical utricular neuronal input owing to interruption of the peripheral or central utricular pathways, and a unilateral utricular imbalance therefore produces an

OTR in the roll plane. Furthermore, they speculated that a bilateral utricular imbalance might cause ASD, because the vertical components combine to produce the slow phase vertical drift of both eyes, while the torsional components cancel each other (2, 22, 23).

In our present patient, ASD with abducting eye hypertropia developed abruptly. Based on the cranial computed tomography findings, the nodulus, uvula, pyramid, tuber, folium, declive, culmen, lingula, anterior paravermis, and posterior paravermis might have been bilaterally involved by a cerebellar hemorrhage. However, neither hydrocephalus nor a brainstem lesion was observed. From these results and based on the proposed hypothesis of the etiology of ASD (2, 22, 23), we considered that the ASD might have been caused by marked bilateral damage to the utricular pathway, which resulted from bilateral involvement of the nodulus and uvula. During our patient's clinical course, the vertical misalignment of the eyes during the right-sided gaze was consistently larger than that during the left-sided gaze. Based on this, we speculated that the damage to the nodulus and uvula might have been asymmetrical. In conclusion, we herein reported the first known case of ASD secondary to cerebellar hemorrhage without hydrocephalus.

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

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