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Viewing condition dependence of the gaze-evoked nystagmus in Arnold Chiari type 1 malformation

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ABSTRACT

Saccadic eye movements rapidly shift gaze to the target of interest. Once the eyes reach a given target, the brainstem ocular motor integrator utilizes feedback from various sources to assure steady gaze. One of such sources is cerebellum whose lesion can impair neural integration leading to gaze-evoked nystagmus. The gaze evoked nystagmus is characterized by drifts moving the eyes away from the target and a null position where the drifts are absent. The extent of impairment in the neural integration for two opposite eccentricities might determine the location of the null position. Eye in the orbit position might also determine the location of the null. We report this phenomenon in a patient with Arnold Chiari type 1 malformation who had intermittent esotropia and horizontal gaze-evoked nystagmus with a shift in the null position. During binocular viewing, the null was shifted to the right. During monocular viewing, when the eye under cover drifted nasally (secondary to the esotropia), the null of the gaze-evoked nystagmus reorganized toward the center. We speculate that the output of the neural integrator is altered from the bilateral conflicting eye in the orbit position secondary to the strabismus. This could possibly explain the reorganization of the location of the null position.

1. Introduction

Saccadic eye movements rapidly shift gaze from one target to the other. For horizontal saccades, pontine burst neurons generate phasic velocity commands to move the eyes swiftly to a new position [1]. Once the eyes reach the new position, they have to counteract the orbital elastic forces in order to maintain that position. Thus, to achieve a steady gaze, the velocity commands are mathematically integrated into a tonic position signal in the rostral medulla [2–5]. These neural integrators, however, are inherently leaky. Their output characterizes drift of the eyes away from the desired position, and a null position where the drifts are absent. Feedback through the cerebellum minimizes these drifts and improves the fidelity of the neural integrator in assuring a steady eccentric gaze. Cerebellar lesions cause gaze-evoked nystagmus featuring drifts toward the null [6–8].

We report a patient with Arnold Chiari type 1 malformation with intermittent esotropia and horizontal gaze-evoked nystagmus who had a shift in the null position depending on the viewing condition. In this patient, we show that the output of the leaky neural integrator and the location of the null position can be altered by misaligned eyes.

2. Methods

2.1. Clinical description

A 28 year-old woman had progressively worsening horizontal diplopia and vertigo for 11 months. The diplopia was worse while viewing objects at distance. The patient would often close her right eye to prevent diplopia. The vertigo was described as the sensation of spinning in the horizontal plane which was improved with eye closure. The vertigo was prominent in right-ward gaze. Clinical examination revealed small angle esotropia of about 10–12 prism diopters during bed-side cover–uncover test. During binocular viewing condition, a left-beating horizontal nystagmus was present in straight-ahead gaze. The slow-phase velocity of the nystagmus worsened during leftward gaze and the nystagmus resolved when the eyes were turned about 15° to the right. The direction of nystagmus reversed when the eyes moved farther away, beyond 15° to the right. These findings suggested a shift in the null position to approximately 15° on the right. There was no evidence of vertical nystagmus in any gaze orientation. Head repositioning with respect to gravity did not affect the amplitude or the direction of the nystagmus. These deficits are illustrated in the video clip recorded during clinical examination (see supplementary material). Saccades had normal velocity as depicted in main-sequence (see supplementary material, Fig. S1). We could not comment on saccadic accuracy in the presence of gaze-evoked nystagmus. Smooth pursuit was interrupted by the quick-phase of the nystagmus.

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Vestibulo-ocular reflexes during vertical and horizontal head impulses were normal. Patient's uncorrected visual acuity was 20/20 in both eyes.

Magnetic resonance imaging (MRI) of the brain revealed the extension of the cerebellar tonsils into the foramen magnum at the level of the mid-body of the second cervical vertebra (Fig. 1A). Such findings were consistent with Arnold Chiari type 1 malformation. The compression of the brainstem due to the herniation of the cerebellar tonsils was evident on the axial MRI sequence (Fig. 1B). It appeared that a brainstem compression was worse on the left side as compared to the right (Fig. 1B).

2.2. Eye movement recordings

The experiment protocol adhered with the Declaration of Helsinki, and it was approved by the Cleveland Clinic institutional review board. The subject gave written informed consent. Horizontal and vertical eye positions were measured non-invasively with a fast video-based eye tracker (EyeLink 1000®, SR Research, Ontario, Canada). Head was supported on a chin-rest, 55 cm away from the LCD monitor. The subject was instructed to follow a visual target that was projected on the LCD screen at straight-ahead, 5°, 10°, and 15° to the right and left, 5° and 10° up and down, and in oblique (10° vertical and 15° horizontal) orientations. The visual target was circular, and its diameter comprised a 0.5° visual angle. The eye positions were measured as the subject attempted to fixate the gaze on the visual target. Binocular eye positions were measured at a temporal resolution of 500 Hz. The digital output from EyeLink® and the distance between the LCD screen and the subject's nasion was used to compute the visual angle. Further analysis was done off line on calibrated visual angle vectors. In monocular viewing condition, one eye was covered with the infrared permissive filter. This filter allowed infrared waves but blocked visible light waves, hence, preventing vision through the covered eye. The infrared permissive filter allowed measurement of the position of the covered eye. Technique and experimental protocol used to measure the positions of both eyes were otherwise identical for binocular and monocular viewing conditions.

2.3. Data analysis

The eye position at the beginning of the drift was used to determine the eye in the orbit position. Horizontal and vertical positions of each eye were analyzed separately. Quick-phases and saccadic eye movements were identified with the Engbert algorithm [9,10]. Saccades and quick-phases were excluded from further analysis. Epochs of eye positions between two adjacent quick-phases were considered as one drift. All eye position epochs comprising drifts were differentiated, and smoothed with a Savitzky–Golay filter (frame length: 11) to compute drift velocity. The median value of drift, velocity, i.e. the slow-phase eye velocity, determined the severity of nystagmus. Statistics and curve fitting toolboxes available through Matlab® (Nattick, MA) were used for further statistical analyses.

3. Results

3.1. Binocular viewing condition

Fig. 2A illustrates one second epochs of horizontal and vertical eye positions during attempted gaze holding. Rightward drifts (slow-phase) followed by leftward corrections (quick-phase) were seen in both eyes when the subject attempted to hold the gaze in a straight-ahead position. The slope of the eye position (the velocity) during drift increased when the subject attempted to hold the gaze on the target that was 15° to the left. In contrast, stable gaze holding was achieved when the eyes were turned 15° to the right. The severity of horizontal nystagmus was independent of the vertical gaze eccentricity. Vertical gaze was stable during straight-ahead, upward, downward, rightward or leftward gaze. Fig. 2B depicts the dependence of the slow-phase velocity of horizontal nystagmus on horizontal and vertical eyes in the orbit positions. The slow-phase eye velocity of horizontal nystagmus was measured during gaze holding in 0°, 5°, 10°, and 15° to the right and left; 5°, 10° up and down, and oblique (i.e. combinations of 5°, 10° vertical and 5°, 10°, and 15° horizontal) orientations. The horizontal gaze velocity was maximal during left gaze as depicted by a surface plot in Fig. 2B. The gaze was stable in a rightward position (Fig. 2B). Vertical drifts were minimal, and they lacked the influence of the eye in the orbit position (Fig. 2C).

We fitted a linear function to quantify the relationship between drift velocity and the corresponding eye in the orbit position. The slope of the linear fit suggests the eye in the orbit position dependence of the drift velocity, while the intercept of the linear fit with the *x*-axis depicted the null position. The slope and 95% confidence interval representing horizontal eye position dependence of the horizontal drift velocity

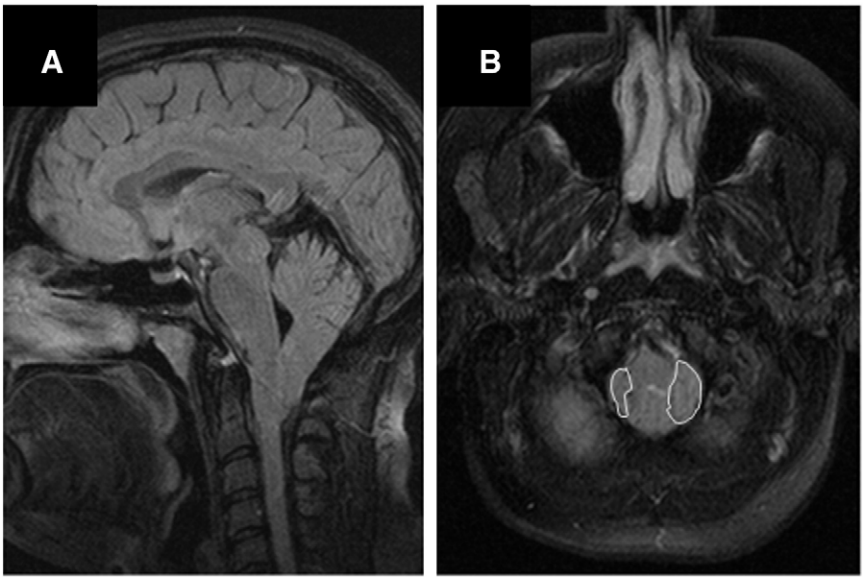

Fig. 1. Brain MRI in fluid attenuated inverse recovery sequence suggesting Arnold Chiari type 1 malformation. Descent of cerebellar tonsils to the level of second cervical vertebra is depicted in panel A. Area of axial MRI in panel B marked with a white border shows the extent of the herniated cerebellum on each side. It appears that the compression due to descended cerebellum was worse on the left side.