Monocular torsional oscillopsia in dentato-olivary disconnection

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Case Report

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Abstract

Monocular torsional eye oscillations are a rare form of disconjugate nystagmus and the underlying pathophysiology is not well understood. Here, we present and discuss a case with disabling torsional oscillopsia in one eye only. The patient exhibited spontaneous pendular torsional nystagmus of the left eye and rhythmic involuntary movements of the soft palate and uvula, consistent with the syndrome of oculopalatal tremor with monocular nystagmus. Brain MRI revealed an infarct of the left dentate nucleus in the cerebellum and, more caudally, a secondary hypertrophic degeneration of the right inferior olivary nucleus. To account for the presence of torsional nystagmus on the eye contralateral to the side of inferior olivary hypetrophy and ipsilateral to the lesioned dentate nucleus, we discuss the hypothesis of a (inferior olivary nucleus-mediated) malfunctioning adaptation of the anterior canal vestibulo-ocular reflex.

Background

Monocular spontaneous nystagmus is a rare manifestation of eye oscillations confined to one eye only. Only a few syndromes have been recognized as causative of this extreme form of disconjugate nystagmus. The Heimann-Bielschowsky phenomenon is a benign syndrome with monocular vertical nystagmus reflecting the coexistence of unilaterally impaired vision due to optic neuropathy and a strategic disruption in brainstem gaze holding centres [1]. Torsional, intermittent, monocular nystagmus can also occur in superior oblique myokymia, usually due to a neurovascular compression of the trochlear nerve [2, 3]. Here, we describe a case with monocular torsional nystagmus after a strategic infarct in the cerebellum.

Case

A 42-year-old man had multiple brain infarctions due to septic embolism from cardiac vegetations in the course of infective endocarditis two years before presentation in our outpatient clinic. He complained of continuous disabling oscillopsia in his left eye only. The objects appeared to oscillate clockwise and counterclockwise with a small vertical component (“bouncing up and down”). The visual scene was stable when looking with the right eye only. On examination a spontaneous pendular torsional nystagmus of the left eye was seen (video and Fig. 1). Pupils were equal and reactive to light, eye movements were full range in all directions and alternating cover testing did not elicit any horizontal or vertical misalignment. Moreover, rhythmic involuntary movement of the soft palate and uvula was seen upon examination of the oral cavity (video).

Besides multiple small infarcts in both hemispheres, brain MRI revealed an infarct of the left dentate nucleus in the cerebellum (Fig. 2). More caudally, a signal hyperintensity was seen in the right inferior olivary nucleus (ION) indicative of hypertrophic olivary degeneration,

Discussion

It is well established that lesions interrupting the connection between the red nucleus, the ipsilateral ION and the contralateral dentate nucleus (triangle of Guillain and Mollaret) lead to transneuronal degeneration causing hypertrophy rather than atrophy of the ION. This manifests as oculopalatal tremor characterized by continuous rhythmic oscillations of the pharynx and oscillopsia due to pendular nystagmus [4, 5]. Ocular oscillations are mostly vertical with additional torsional components and are mostly conjugate in bilateral ION hypertrophy. Unilateral ION signal changes, however, are associated with dissociated nystagmus in over half of the cases, which is more pronounced on the eye contralateral to the side of ION hypetrophy [6, 7].

Although ION hypertrophy appears to be a necessary factor in the emergence of pendular oculopalatal oscillations, it is in itself not a sufficient factor. Instead, the vestibulo-ocular reflex (VOR) pathway may be critically involved in the genesis of pendular nystagmus in oculopalatal tremor. ION neurons send climbing fiber axons to Purkinje cells in the contralateral cerebellar cortex via the inferior cerebellar peduncle. The ION is actively involved in error based learning and timing of eye movements by modifying the synaptic weights of parallel fibers projections onto the floccular Purkinje neurons. Experimental lesions of the ION in animals disrupts adaptation in the gain of the VOR [8, 9]. Accordingly, pendular nystagmus in oculopalatal tremor may result from a dysfunctional VOR gain adaptation (leading to an oscillatory state) due to hypertrophic olivary degeneration [10]. However, the mechanism of discojugancy of eye oscillations is less well understood in this context. Nakada and Kwee [10] argue that the malfunctioning ION-mediated VOR adaptation is an inherently unilateral syndrome, while bilateral manifestations (i.e. conjugate nystagmus) represent the summation of the unilateral (i.e. monocular nystagmus) disorder. Their hypothesis is based mainly on experimental data provided by Masao Ito and collegues, who demonstrated that monocular torsional oscillations may ultimately result from a disinhibition of the anterior canal VOR pathway due to a dysfunctional floccular control, given the fact that the flocculus indirectly regulates the tone of the superior and inferior oblique muscle of the contralateral eye [11]. Hence, spontaneous monocular nystagmus with a dominant torsional component as observed in the present case, may reflect the faulty activity of a maladapted anterior canal VOR.

In conclusion, we presented a unique case of monocular torsional oscillopsia due to unilateral degenerative ION hypertrophy as a consequence of a circumscribed infarct of the contralateral dentate nucleus. In view of the challenging differential diagnosis of monocular nystagmus, oculopalatal tremor due to disruption of the dentate-olivary pathway should be kept in mind as a rare cause.

Declarations

Ethical Approval

The study was performed in accordance with the ethical standards laid down in the Declaration of Helsinki (1964).

Informed Consent

The informed and written consent for publication of the data and videos was obtained by the patient.

Competing interests

The authors declare that they have no conflict of interest.

Authors' contributions

E.A.: study conception and design, manuscript preparation.

S.G: study conception and design, manuscript preparation

G.A.: study conception and design, manuscript preparation.

A.Z.: study conception and design, final approval of manuscript.

G.V..: study conception and design, manuscript preparation.

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Availability of data and materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Disclosure of Conflicts of Interest

None of the authors has any conflict of interest to disclose.

Ethical Publication Statement

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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Figures

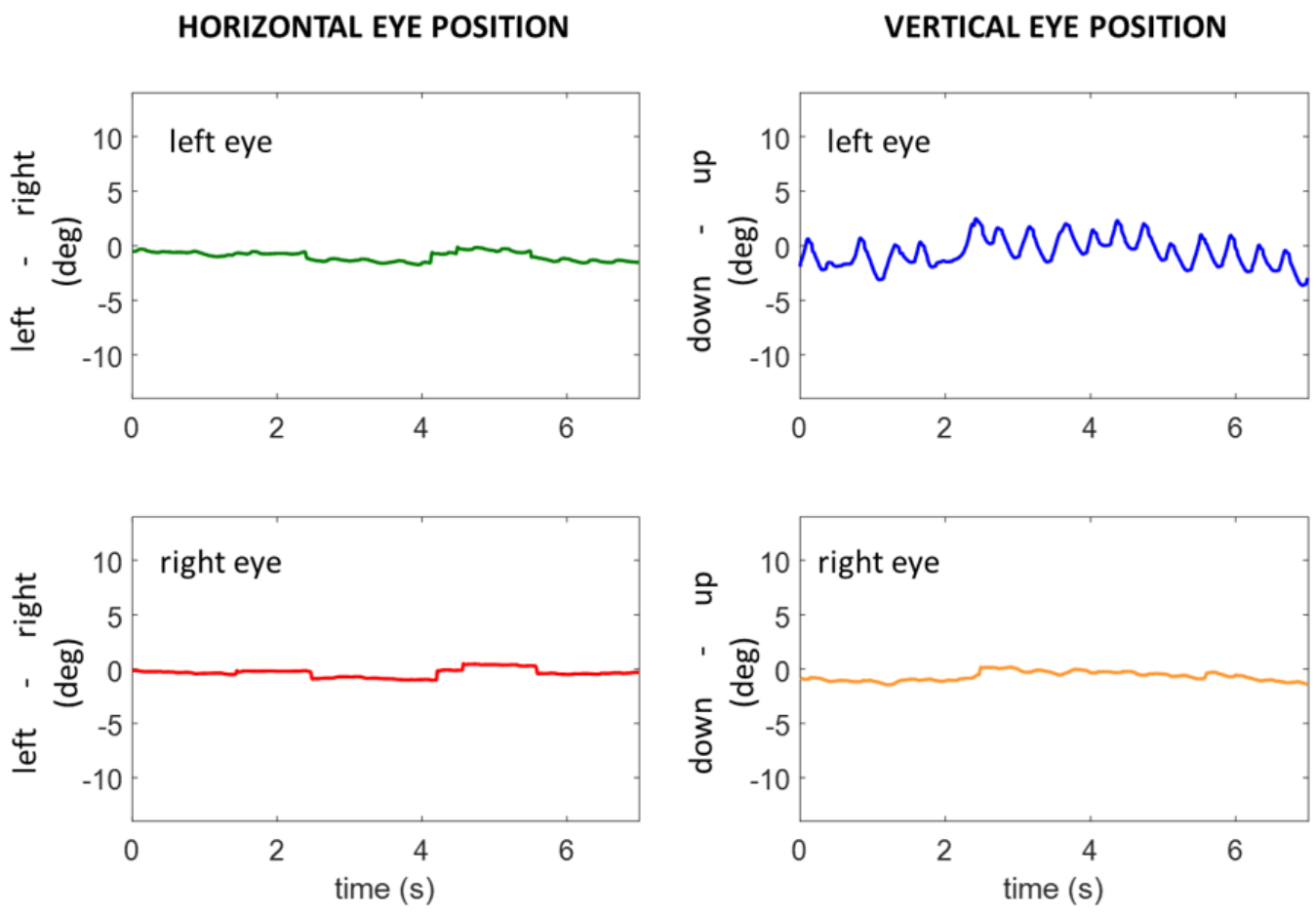

Figure 1

Binocular video-oculographic during straight-ahead fixation. Pendular oscillations are clearly seen in the vertical trace of the left eye. Torsional eye movements could not be measured by this recording technique.

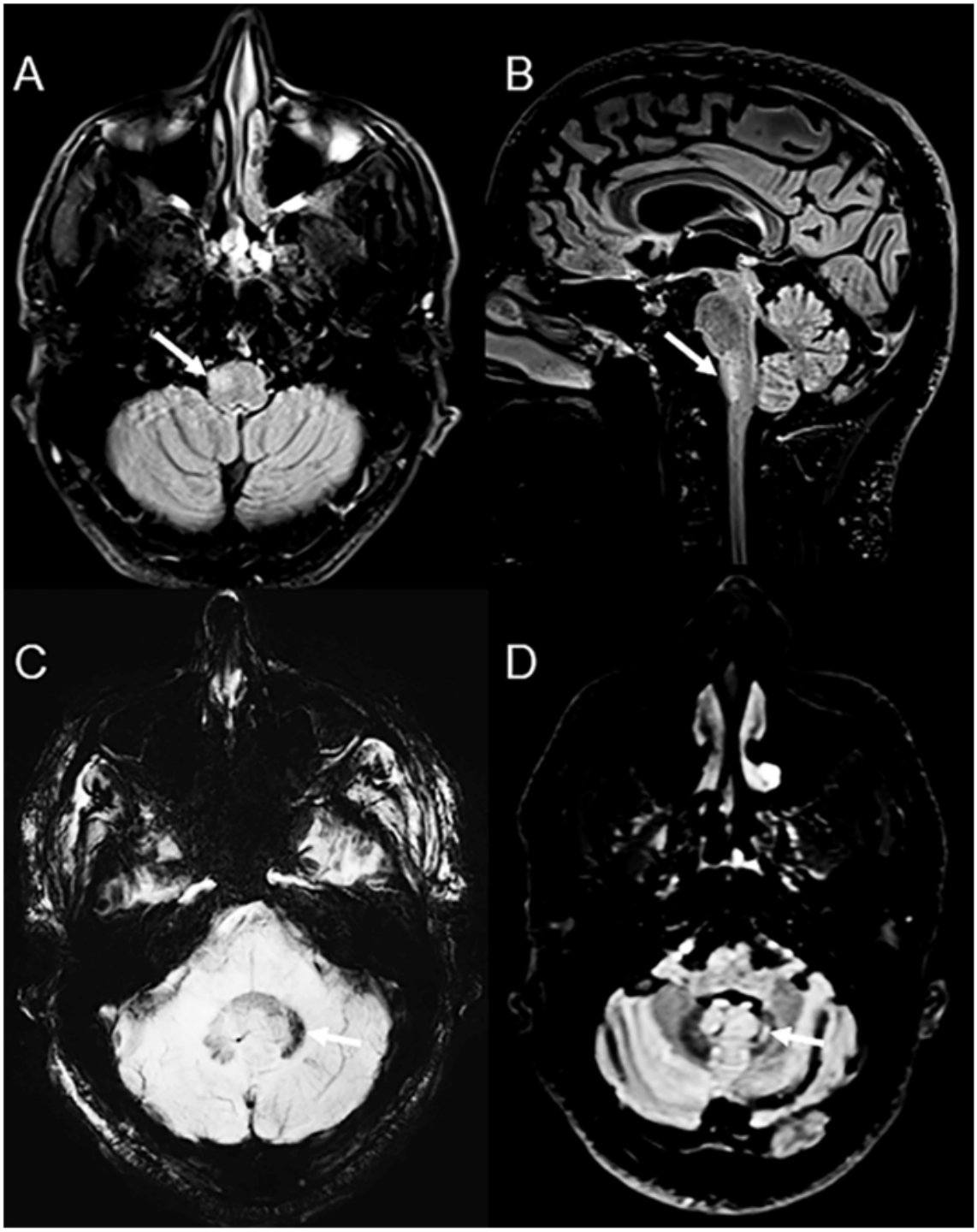

Figure 2

Axial T2- Fluid Attenuated Inversion Recovery (FLAIR) image (A) reveals hyperintense enlargement of the right inferior olivary nucleus (arrow A). Olivary hyperintensity and enlargement are also seen on T2-FLAIR sagittal image (arrow B). Hemosiderin deposition on Susceptibility Weighed Imaging (SWI) (arrow C) and hyperintensity on T2-FLAIR image (arrow D) are noticed in the left dentate nucleus of the cerebellum, due

to a chronic infarct. Hypertrophic olivary degeneration is attributed to the primary lesion in the contralateral dentate nucleus, which is part of the Guillain-Mollaret triangle.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- video.mp4