CASE STUDY

Periodic Alternating Nystagmus – A Case Report

Nistagmus alternante periódico: caso clínico

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Case Report

A 71-year-old man complained of 5 years of progressive postural instability, more intense when standing up from bed, especially in darkness and during the day when walking on street. It lasted a few seconds and resolved spontaneously. No reference to vegetative symptoms, hearing loss or rotary movements.

Relevant personal clinical antecedents were slowly progressive visual loss that began at age 20, followed by the diagnosis of pigmentary glaucoma at age 40, and three episodes of retinal detachment repair (twice in the right eye and once on left eye, during the last two decades), and 6 months ago he underwent a combined surgery directed to right glaucoma and cataract.

Other antecedents were lymph node tuberculosis and radical prostatectomy three years ago and benign prostatic positional vertigo of horizontal and posterior right semicircular canals treated with repositioning maneuvers two years ago.

The latest otoneurologist examination detected a spontaneous horizontal nystagmus in the primary position that periodically reverses its direction with a transition period of several seconds, with visual suppression.

An approximately 155 s left-side nystagmus (slow phase velocity (SPV): 4.9/s), an approximately 15 s pause with low amplitude square-wave jerks, and an approximately 105 right-side nystagmus were recorded (video 1†, Fig. 1).

With straight head-hanging position an upbeat nystagmus appeared with a small torsional component. The smooth pursuit and saccadic pursuit were normal.

His ophthalmological status on the latest otoneurological examination was 10/10 visual acuity on right eye, and 50/100 visual acuity on the left eye.

A mild bilateral hearing loss was found.

We performed the head impulse test to evaluate the vestibulo-ocular reflex evoked by the stimulation of the 6 semicircular canals with a video-based system (v-HIT, GN Otometrics, Denmark) and the results were normal for both ears and each semicircular canal (normal gain, no refixation saccades). Sinusoidal harmonic accelerations (frequency range 0.01–0.64 Hz, peak velocity: 50/s) showed a phase advance and normal gains and symmetry,

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† Video—The patient shows a 9 min sequence of periodic alternating nystagmus (PAN) that consists of left beating nystagmus (more durable), a null phase, and right beating nystagmus, and then a repetition of all phases. Link: https://www.dropbox.com/s/e5egzk6n1hvebuf/PAN%20final.m4v.

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without per-rotatory visual suppression with ocular fixation, generating a sinusoidal pattern. On visual-vestibular interaction, a sinusoidal harmonic acceleration with optokinetic stimulus (frequency range 0.01–0.64 Hz, peak velocity: 50°/s) phase, gain and symmetry were normal. Magnetic resonance imaging revealed periventricular leukoencephalopathy on oval centers bilaterally.

The patient underwent treatment with baclofen (GABAergic drug), 30 mg/day without recovery; however after that, the patient treatment a new surgical treatment on the left eye, recovered the visual acuity and the PAN disappeared.

Discussion

Acquired periodic alternating nystagmus (PAN) is an uncommon disorder in which there is a continuous spontaneous horizontal nystagmus, present in central gaze, that periodically reverses direction (approximately every 90–120 s), with a null period of several seconds on which downbeat nystagmus and square wave jerks.1-4

PAN can be congenital or acquired.1-7 Acquired PAN has been reported in various neurologic disorders, such as Arnold–Chiari malformation and other hindbrain anomalies, multiple sclerosis, cerebellar degenerations, cerebellar tumor, abscess, cyst, and other mass lesion, Creutzfeldt–Jakob disease, ataxia telangiectasia, brainstem infarction, lithium, anticonvulsant medications and seizures, hepatic encephalopathy, trauma, following visual loss (vitreous hemorrhage or cataract) and aperiodic form as a component of congenital nystagmus, especially in albinos.

In most patients with acquired PAN, the nystagmus has the same characteristics in light or in darkness.1 Because the period of oscillation is about 4 min, the diagnosis may be missed unless the examiner observes the nystagmus for several minutes.1

The per-rotatory nystagmus is normally suppressed by visual fixation; however, this suppressive effect is lost after damage of the uvulonodulus.1

The pathomechanisms thought to be responsible for PAN came from experimental studies of ablation of the cerebellar nodulus and uvula (NU) in monkeys, causing PAN in darkness.1-5 The cerebellar NU control the time course of rotationally induced nystagmus ("velocity storage" system). Thus, dysfunction of NU causes PAN by prolongation of velocity storage and the instability of VOR. It was postulated that normal vestibular "repair mechanisms" act to reverse the direction of nystagmus, producing the oscillations. And the latter can be blocked by visual stabilization mechanisms. However, cerebellar disease that causes PAN also impairs these "compensatory mechanisms".

Normal results in the vHIT precluded more examinations as there is convincing evidence of normal peripheral
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vestibular function. Although the caloric test is possible, some caution must be taken into account at the time of evaluating results as it is recommended to observe nystagmus during the whole test to disclose reversal of the expected caloric nystagmus due to PAN.

The baclofen (GABAb-ergic drug) abolishes successfully acquired PAN due to its inhibitory control on the vestibular rotational responses on Purkinje cell activity of the nodulus and uvula.1,4

In our patient there was no evidence of any underlying neurologic disorder even on MRI, and we presume that PAN emerged due to an impairment of central visual stabilization mechanisms and deficient visual input and fixation, caused by his ophthalmological antecedents.

Another point of interest in our case was the asymmetry of the nystagmus, probably due to the asymmetry of visual input (deficient visual acuity on left eye), suggesting that the left VOR may be relatively more unstable than the right.

It was postulated that unilateral visual loss itself does not cause PAN1 and in our case it was not demonstrated. However, PAN was abolished after the latest successful ophthalmologic surgery.

Conflict of Interest

No conflict of interests.

References