Aperiodic alternating nystagmus: report of two cases and treatment by baclofen

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We report two cases of non periodic alternating nystagmus, one of vascular origin (ischemia of the vertebrobasilar territory) and a second of traumatic origin (whiplash injury) with otoneurological signs, typical of lesions in posterior cranial fossa and in particular of vestibulum-cerebellum and brainstem: gaze paretic nystagmus, rebound nystagmus, saccadic dysmetria, vestibular hyperreflexia and impaired visual suppression test.

In one case it was possible to give baclofen therapy, which yielded positive results. Suspension of drug administration resulted in the worsening of clinical signs. The mechanism of action of the drug will be discussed.

Key-Words: Nystagmus alternans — baclofen

Introduction

Alternating nystagmus is a rare form of spontaneous nystagmus first described by Borries in 1920, since which time no more than 100 cases have been reported. It is characterized by alternation of direction, as a rule periodic, the cycles being unaffected by head movements. The changes of direction are preceded by an interval of variable length during which the eyeballs are immobile. The nystagmus is present in the primary position of gaze, horizontal or horizontal-rotatory; it is often associated with vestibular hyperreflexia, rebound nystagmus and down-beating vertical nystagmus. A prevalence of left-beating nystagmus has sometimes been described [19].

Alternating nystagmus may be congenital or acquired. Congenital nystagmus is generally an isolated phenomenon and can often be reduced or abolished in relation to certain head postures. Acquired alternating nystagmus is accompanied by other neurological signs. In 5 cases that reached the dissecting table, an anatomical substrate was found: arachnoidal cyst of the cerebellum [24], Arnold-Chiari malformation [24], multiple sclerosis [13], cerebellar astrocytoma [12] and cryptococcal cyst of the floor of the fourth ventricle [8, 15].

In clinical studies alternating nystagmus has been reported in cases of chronic otitis media with labyrinthine fistula, head injury, vertebrobasilar insufficiency, encephalitis, nervous system syphilis, Friedreich ataxia, tumors of the cerebellum and corpus callosum [2, 6, 16, 17]. It has also been reported in cases of congenital nystagmus with or without albinism and nystagmus associated with total blindness [6].

We report here two clinical cases of aperiodic alternating nystagmus, one secondary to vertebrobasilar insufficiency and the other to head injury with cervical whiplash. The first patient was treated with baclofen with satisfactory results.

Case reports

Case 1
This 68 year old man was admitted to our hospital because of a sudden inability to maintain the erect position due to left lateropulsion and oscillopsia. He had a history of hypertension, with a systolic pressure of 180mm Hg, dating back to the age of
The patient, aged 55, was admitted to the neurological department with a history of dyslipidemia, a myocardial infarct dating back to the age of 55 and a duodenal ulcer at 56. At the time of admission, the patient appeared to be in fairly good general condition with BP 180/100. Neurological examination revealed mild anisocoria (OD > OS), a coarse spontaneous left-beating nystagmus unaccompanied by vertigo or cochlear or vagal signs, truncal ataxia with severe left lateropulsion, some uncertainty in the heel-knee test on the left but no dysmetria. The blood chemistry tests showed high plasma values of nitrogen (0.58 mg/100 ml), creatinine (1.3 mg/100 ml), uric acid (8.1 mg/100 ml) and triglycerides (331 mg/100 ml). An X-ray of the cervical spine showed diffuse uncovertebral arthrosis and spondyloarthrosis; ECG showed signs of a healed myocardial infarct; EEG and CT scan of the skull with contrast medium were normal whereas Doppler ultrasonography of the supra-aortic vessels revealed a stenosis of the left vertebral artery and signs of diffuse vascular sclerosis.

He underwent otoneurological assessment at the ENT department of Siena University. Tonal and vocal audiometry and impedance tests were normal. Auditory brainstem response (ABR) showed a normal tracing in morphology and latencies with left ipsilateral stimulus but altered on right ipsilateral stimulation with the first 3 peaks missing and increased latency of wave V (Fig. 1). Electronystagmography showed severe ocular dysmetria due to overshoot of the leftward saccadic movement and inability to perform pursuit movements. Gaze paretic left-beating nystagmus on fixation was present (Fig. 2); with Frenzel glasses onset in the primary position of gaze of rare jerks of left-beating horizontal nystagmus with short bursts of right-beating nystagmus (mostly lasting 2-15 sec) at totally nonperiodic intervals (Fig. 3). This nystagmus was not affected by position of gaze or head. A vestibular hyperreflexia with caloric stimulation by Fitzgerald Hallpike method and deficit of visual inhibition in 3 out of 4 stimulations were present (Fig. 4). A 15-minute recording confirmed the complete nonperiodicity of the alternation of the phases of nystagmus, with mainly left-beating phases and short free pauses between cycles, not exceeding 10% of the entire recording. As reported by Halmagyi (1980) the patient was treated with baclofen (30 mg daily in three divided doses). Control recordings after 5 and 10 days showed considerable changes in the pattern: the alternating nystagmus was still present but the two phases were balanced in duration and the free intervals took up about 70% of the recording (Fig. 5). The left-beating gaze paretic nystagmus, ocular dysmetria and chaotic pursuit persisted. The vestibular hyperreflexia was present only with right caloric stimulation, in which a deficit of visual inhibition appeared. In the recording after 10 days treatment, it was possible to detect the presence of rebound nystagmus, not previously found. The patient showed distinct clinical improvement, managing to walk without support.

Fig. 1. Patient 1. Horizontal bitemporal lead.
Upper tracing: ocular calibration showing left overshoot.
Central tracing: impaired smooth pursuit.
Lower tracing: left gaze paretic ny (eyes open with Frenzel glasses).

![Patient 1. Horizontal bitemporal lead.](image)