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Caso clínico

Downbeat nistagmus – 2 cases reports

Downbeat nystagmus – 2 casos clínicos

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Abstract

Downbeat nystagmus (DBN) as a clinical sign of central nervous system (CNS) abnormality, mainly occurs in structural lesions of vestibulocerebellum (floccular and nodular lesions). However it could also have a peripheral cause or be idiophatic. The authors describe two cases reports of DBN and make a revision of the literature.

Key Words: Downbeat nystagmus, central nervous system, etiology

Resumo

O Downbeat nistagmus (DBN) como sinal clinico de alteração do Sistema Nervoso Central ocorre principalmente em lesões de estruturas do vestibulocerebelo (lesões a nível do nódulo e do flóculus). Contudo, pode também ter uma causa periférica ou idiopática. Os autores descrevem 2 casos clínicos de DBN e fazem uma revisão da literatura acerca do tema.

Palavras-chave: Downbeat nystagmus, Sistema Nervoso Central, etiologia

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Introduction

Downbeat nystagmus (DBN) in primary gaze is the most common form of acquired persisting fixation nystagmus[1-6]. It is characterised by slow upward drifts and fast downward phases. Slow-phase velocity (SPV) increases on lateral, downward gaze and convergence (Alexander's law is obeyed). However atypical presentations may occur, with enhancement of the DBN on upward gaze or suppression on convergence [1,2] .

The most common presenting symptoms of DBN are illusory motion of their visual environment (oscillopsia) that increases on lateral gaze, which leads to postural instability and clumsiness [1,2].

Case Reports

Patient 1

A 76 years old man, complained of 3 years of progressive postural instability and vegetative symptoms when walking, that diminish in the sitting position and remiss in the supine position. Complained of one episode of positional vertigo and nausea, one month ago that lasted 2 hours and resolved with sulpiride.

Also referred a light bilateral progressive hearing loss, not concomitant with any of the described events.

The otoneurologist examination (Figure 1, video 1) found a vertical downbeat nystagmus, without torsional component, on primary gaze, with visual fixation, increasing velocity on lateral gaze, bilaterally. Without visual fixation the nystagmus became greater (in amplitude), with a torsional component (right-horizontal).

A suppression of the horizontal nystagmus component is detected on per-rotatory nystagmus with visual fixation, maintaining the vertical one on both (with and without visual fixation).

On straight head-hanging position, vertical nystagmus became greater (either on velocity and amplitude), and maintained on supine and on sitting up position. No modification of DBN was detected on head-shake test.

The remainder neurological examination was normal, with exception for a left retropulsion on Romberg.

A mild mixed bilateral hearing loss was found.

The subjective postural vertigo was normal. The video head impulse test, revealed an abnormal result on left ear (low gain of left posterior canal).

Computerized dynamic posturography detected a pathological result, due to a vestibular deficit, with a composite score of 54.

Brain computed tomography (CT) was normal.

Magnetic resonance imaging (MRI) could not be obtained because of patient's pacemaker.

The patient underwent with 3-4 diaminopyridine (30mg/day), and improved significantly his complaints.

Patient 2

A 66 years old man, complained of four years of progressive unsteady gait with blurred vision, that usually started during the breakfast, increased with head movements, reading and walking, and submitted when sitting or on supine position.

The otoneurological examination detected a vertical downbeat nystagmus, without torsional component, on primary gaze with visual fixation, increasing velocity on lateral gaze, bilaterally, and without visual fixation.

DBN increased with head-shake test (Figure 2, video 2), infraversion, and on straight head-hanging position.

The smooth pursuit testing was a pathological saccadic pursuit, due to the addition of a DBN that increased intensity during all horizontal pursuit.

A deficient visual suppression of per-rotatory nystagmus was detected.

The video head impulse test, revealed an abnormal result on right and left ear (low gain of anterior and

Figure 1: Spontaneous and gaze-evoked nystagmus (with and without visual fixation, patient 1).

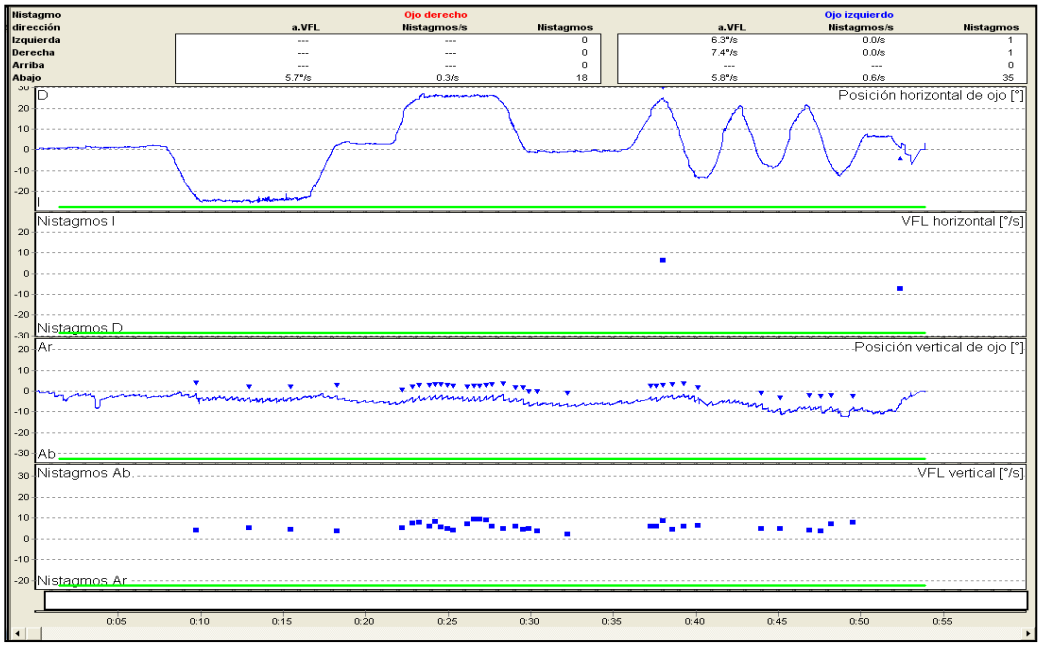
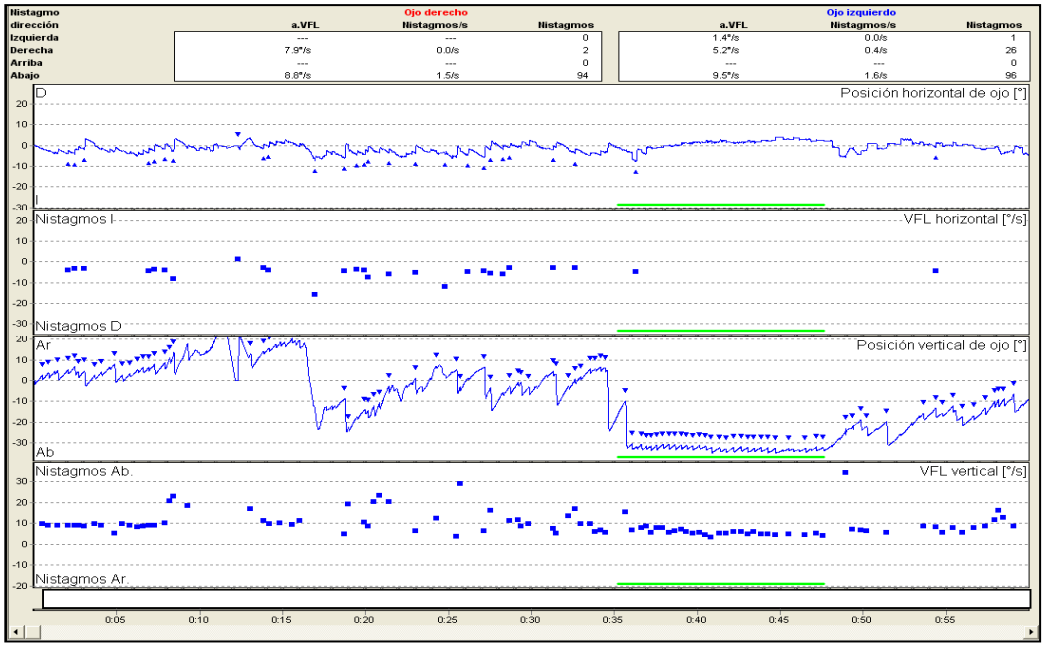


Figure 2: Head-shake test (patient 2).



posterior canals bilaterally, due to DBN).

Computerized dynamic posturography was normal with a composite score of 73.

The remainder neurological examination was normal, with exception for a right lateropulsion walking and a Babinski-Weil with deviation to the right.

MRI revealed a residual ischemic lesion on frontal lobe. Cervical MRI and lumbar puncture were normal.

The patient underwent with 3-4 diaminopyridine (30mg/day), and improved significantly his complaints.

Discussion

The aetiology of DBN is diverse. It could have a central cause, peripheral cause or be idiopathic.

DBN as a clinical sign of central nervous system (CNS) abnormality, mainly occurs in structural lesions of vestibulocerebellum (floccular and nodular lesions) and in conditions such as multiple system atrophy, multiple sclerosis, cerebellar degeneration, lesions near the craniocervical junction and hydrocephalus[2,3]. Drug intoxication, especially with lithium was already been described[2].

Three pathomechanisms are thought to cause the spontaneous upward drift: first, a tone imbalance of the central vestibular pathways of the vertical eye movements, including otolith pathways as suggested by the findings that DBN is gravity-dependent; second, an imbalance of the smooth pursuit tone in which the imbalance of upward visual velocity commands results in spontaneous upward drift and third a mismatch in the three-dimensional neural coordinate system for vertical saccade generation due to a defect of the neural velocity-to-position integrator for gaze holding[1].

It has been also proposed that DBN is caused by a reduced function on the inhibitory, vertical gaze-velocity Purkinge cells (PCs) in the cerebellar flocculus[5], showing a physiological asymmetry, having a preponderance of cells with downward on-directions (fast phase) [1,5]. This, conducted to the investigation of several agents that act on GABAergic PCs receptors increasing the resting activity and excitability of the them, such as $GABA_A$ agonist (baclofen), clonazepam, 4-aminopirydinine, 3,4 – diaminopyridine and chlozoxazone[1,5,6]. They significantly decrease the SPV of DBN, improving visual acuity and postural imbalance, despite their particularly secondary effects.

Relating with peripheral cause, the uncommon an-

terior canal (AC) variant of benign paroxysmal positional vertigo (BPPV) and the atypical forms of posterior canal (PC) BPPV should be considered on differential etiologic diagnostic of DBN[3] and so, Dix-Hallpike and straight head-hanging positional (SHHP) maneuvers should be tested, before assuming that DBN has a central cause[3,4].

In both of the presented cases, patients had a diagnostic of DBN of central cause. However it is important to note that its syndrome has a huge differential diagnostic, and the clinicians should take aware for all the presentations and variations that could occur.

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