SHORT REPORT

Upbeat and downbeat nystagmus occurring successively in a patient with posterior medullary haemorrhage

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Abstract

In a patient with posterior medullary haemorrhage, first upbeat and later downbeat nystagmus occurred in the primary position. The lesion was limited to the posterior and medial part of the medulla. Clinical and electro-oculographic examination first showed upbeat nystagmus in the primary position and upgaze, with downbeat nystagmus in downgaze. Two and a half months later, there was downbeat nystagmus in the primary position and downgaze and upbeat nystagmus in upgaze.

Vertical nystagmus is often associated with degenerative diseases of the brain stem or malformations of the cervico-occipital junction. Localised lesions rarely cause this abnormality.¹⁻⁴ Most authors have clearly distinguished downbeat nystagmus, which often accompanies lower cerebellar lesions or the Chiari malformation,⁵⁻⁷ from upbeat nystagmus.

The following case report shows that upbeat nystagmus followed by downbeat nystagmus may be observed in succession with a postero-medial lesion of the medulla.

Case report

A 21 year old woman, with no past neurological history, complained of sudden occipital headache. Two hours after the event, neurological examination only showed somnolence. A few hours later, severe tachycardia (200/min) and pulmonary oedema developed. A CT scan showed a non-enhancing hyperdense lesion, probably a haematoma of the fourth ventricle. During the following days, upbeat vertical nystagmus developed. The pupils were meiotic and reactive. Operation on the eighth day, revealed a haematoma in the medial medullary sulcus which continued under the obex. The lateral floor of the fourth ventricle was spared. Post operatively, bilateral palsies of the pharynx and tongue and nystagmus were present, with gait imbalance. The latencies of the auditory brain-stem potentials and the somatosensory evoked potentials were normal.

Two and a half months after the haemorrhage, the direction of the primary position nystagmus had reversed. The patient was followed regularly for three years. Bilateral lingual atrophy persisted with downbeat nystagmus in the primary position and downgaze and upbeat nystagmus on upgaze. An MRI showed a circumscribed scar lesion in the postero-medial medulla (fig 1).

Oculomotor examination

An electro-oculographic examination was performed six weeks after the haemorrhage. The patient complained of oscillopsia. Saccades to command and in response to visual targets presented on a television screen were measured. Sinusoidal smooth pursuit was also elicited on a television screen (5, 10, 15, 20°/sec) and oculocephalic movements by passive rotation of the head. In the primary position, with and without fixation, an upbeat nystagmus (4°; 2 Hz) was noted (fig 2, A1). It was temporarily reversed by convergence (fig 2, A2), and its amplitude increased on upgaze (fig 2, A3). It was replaced by a downbeat nystagmus on downgaze (fig 2, A4) and a horizontal-

Figure 1 MRI 10 months after haemorrhage: postero-medial medullary hyperecho in the floor of the fourth ventricle.

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Figure 2 Nystagmus at the early (A: 1 and a half month) and late (B: 10 months) stages after medullary haemorrhage: primary position (1), convergence (2), upgaze (3) and downgaze (4).

Figure 3 Vertical smooth pursuit at the early (A) and late (B) stages, at 5°/s (1) and 15°/s (2). Vertical oculo-cephalic movements (3).

rotatory nystagmus on lateral gaze. Saccadic eye movements were normal. Smooth pursuit was affected, especially on upward movement (fig 3, A1 and A2). Vertical vestibulo-ocular reflexes were reduced (fig 3, A3). Vertical optokinetic nystagmus was unsustained. Convergence was brisk, but the return to primary position was slow.

Eye movement measurements were repeated nine months later. Downbeat nystagmus (2°; 1.9 Hz) was observed in the primary position (fig 2, B1) and downgaze (fig 2, B4), with upbeat nystagmus in upgaze (fig 2, B3). The downbeat nystagmus became upbeat nystagmus with convergence (fig 2, B2). Convergence was unchanged.

Discussion

In this patient, the major effect of the haemorrhage in the medulla were oculomotor problems and partial bilateral lingual atrophy. Clinical examination, pre-operative findings, MRI and evoked potentials suggested a medial lesion in the area⁸ of the medial longitudinal fasciculi, hypoglossal nuclei, intercalated nuclei and dorsal vagal nuclei. The dorsal longitudinal tracts of Schutz were probably damaged, as well as the posterior-medial reticular formation. The nuclei prepositi hypoglossi may also have been affected.

Initially, an upbeat nystagmus was present in the primary position and increased in upgaze. Convergence temporarily reversed it. This phenomenon was similar to that described by Cox *et al*⁹ in a case of Wernicke-Korsakoff syndrome. Upbeat nystagmus associated with discrete focal lesions of the brain stem is rare. It was described in a case of arterial thrombosis of the anterior spinal artery¹⁰ and in cases with bulbar lesions.^{13,4,11} Keane, *et al*⁴ submitted an anatomo-clinical case in which bilateral lesions affected the postero-median bulbar structures, especially the medial longitudinal fasciculi. In another case,² lesions of the floor of the fourth ventricle extended from under the sixth cranial nerve to the lower part of the medulla.

Upbeat nystagmus could be due to an imbalance of vestibular impulses to the elevator and depressor muscles of the eyes. A lesion in the neural circuitry originating from the anterior semicircular canals, going through the superior vestibular nucleus, brachium conjunctivum and ending in the contra-lateral superior rectus subnucleus, may be responsible. In our case, a lesion of the superior vestibular nucleus or part of the brachium conjunctivum may have occurred in the initial phase. Other authors, however, have assumed that upbeat nystagmus may be caused by lesions in the nuclei surrounding the hypoglossal nucleus, especially the nucleus prepositus hypoglossi.^{2,4} With such a lesion, the vestibulo-ocular reflex would be expected to remain normal. In our patient, the deficit of the vertical vestibulo-ocular reflex suggested a dysfunction of the system linking the vestibular and oculo-motor nuclei.

Two and a half months after the haemorrhage, the patient displayed downbeat nystagmus in the primary position. We again found an inversion of this nystagmus during convergence.^{9,12} The Chiari malformation, cerebellar ectopia¹³ and flocculus lesions¹⁴ have generally been incriminated as the cause of downbeat nystagmus. This nystagmus is very rare in brain stem lesions.⁶ In animals, medial medullary transection may give rise to downbeat nystagmus.¹⁵ The lesions responsible would affect a circuit linking the posterior vertical semicircular canals, the medial vestibular nucleus, the medial longitudinal fasciculus and the part of oculomotor complex innervating the inferior rectus. This system is more medial than that previously considered in upbeat nystagmus.^{16,17}

How can one interpret the successive occurrence of upbeat and downbeat nystagmus? According to the findings of Ito *et al*,¹⁶ it is likely that in the initial phase, oedema induced dysfunction of the superior vestibular nuclei or part of the brachium conjunctivum is the cause. When the oedema subsided, the underlying damage to the medial longitudinal fasciculi was responsible for the persistent downbeat nystagmus.

We also observed that, after convergence, the eye movements back to primary position were slow. This resembled convergence spasm. We suggest that lesions of the vestibular oculo-motor circuitry may be responsible for such a phenomenon. There could be an effect of the impulses of vestibular origin on the return to the primary position of the eyes.

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