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Monocular downbeat nystagmus

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Summary. A patient with sporadic pontocerebellar degeneration and downbeat nystagmus limited to the left eye is described. The nystagmus was not modified by head movements, but was associated with a purely tonic upgaze paresis in the same eye. Absence of internuclear ophthalmoplegia indicated sparing of the medial longitudinal fasciculus. It is suggested that the vertical oculomotor abnormalities are due to dysfunction of the ipsilateral brachium conjunctivum.

Key words: Downbeat nystagmus – Cerebellar degeneration – Tonic upgaze paresis – Brachium conjunctivum

Downbeat nystagmus (DBN) is a primary position binocular nystagmus, which is not modified by fixation, but characteristically increases during lateral gaze [4, 8, 10] and sometimes with head movements in the vertical plane [7, 10]. The exact site of the lesion causing DBN has not been found, but most authors think that dysfunction of the vestibulocerebellum, especially the nodulus, is responsible [4, 7, 9, 12].

As all previously published cases have had binocular nystagmus, we now report a patient with strictly monocular DBN, in whom the features associated with cerebellar involvement suggested upper pontine dysfunction, possibly of the ipsilateral brachium conjunctivum.

Case report

A 26-year-old housewife was admitted for investigation of progressive ataxia. Since 18 years of age, she had developed ataxia with awkward movements of the limbs, dysarthria, and dystonia of the neck and limbs. She had no family history of neurological disease, and her birth and infancy were described as normal. During adolescence, she suffered episodes of anorexia nervosa, which cleared when she was aged 21. She was married and had one normal child.

Examination showed her to be well-developed and alert. She had normal olfaction, vision, gustation and hearing. Optic fundi and visual fields were normal, as were pupillary reactions. She had monocular DBN in the left eye with the associated oculomotor features described in the following. Her speech was dysarthric, and she showed occasional dystonic rotation and extension of the neck. Muscle strength was normal. Ataxia was observed in all the limbs and was more prominent on the right side, with action tremor, hypermetria, and dysdiadochokinesia. During active mobilization of the limbs, dystonic movements predominating distally also appeared. She walked stiff-legged, slowly, on a 30-cm base, with severe instability, and staggered on turning. She was not able to stand with the feet together. The tendon reflexes were decreased in the upper limbs and absent in the lower limbs. Abdominal reflexes were absent and plantar responses were flexor. Sensation was normal in the upper limbs, but position sense was decreased in both feet. Neuropsychological examination was normal. General examination did not show any abnormality, except cervico-dorsal kyphoscoliosis.

Oculomotor findings. Eye movements were repeatedly examined at the bedside, filmed, and recorded with AC ENG (time constant: 5 s). Horizontal and vertical saccadic and pursuit eye movements were tested as the patient followed a spot of light projected onto a circular screen placed 1 m in front of her. Optokinetic nystagmus was induced by projecting alternating black and white stripes on the screen (20–40°/s). The vestibulo-ocular reflex (VOR) was tested with the patient sitting in a motorized chair, by rotating the chair in total darkness (20°/s^2, 200°/s). In the primary position, DBN (8–15°/s) was present in the left eye, without associated movements in the right eye (Fig. 1). This DBN increased during lateral gaze, and decreased during upgaze. It was not modified by bringing the patient from a sitting to a supine position with hyperextension of the neck or by other movements of the head. Optokinetic stimuli directed upward increased the monocular DBN, which disappeared with upward optokinetic stimuli and was replaced by a weak upward beating response. In the left eye, upward voluntary saccades were normal but were followed, after 0.5–1 s, by a downward drift (tonic paresis) superimposed with persisting DBN (Fig. 1). Downward pursuit in the left eye was interrupted by DBN. In the right eye, vertical saccades and pursuit were normal. Horizontal voluntary saccades were normal, but smooth pursuit was moderately saccadic. Horizontal optokinetic responses were bilaterally slowed. Convergence was preserved. Pre- and postrotatory responses were symmetrical and normal.

A computed tomographic scan (Fig. 2) showed dilatation of the prepontine cisterns, suggesting pontine atrophy. The size of the ventricles was normal, and no evidence for cerebral or cerebellar atrophy could be found. CSF (obtained by spinal tap) was clear and showed no leucocytes, with protein of 355 mg/l. Protein electrophoresis and immunoglobulin levels were normal. A Venereal Disease Research Laboratory Test was negative. Visual and brain-stem auditory evoked potentials were normal. On neurographic examination, motor con-

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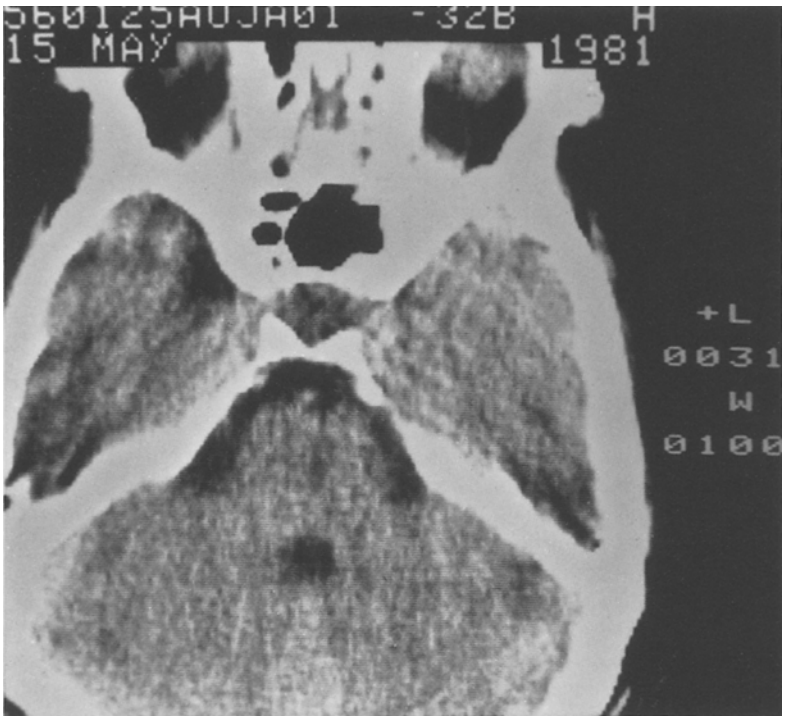

Fig. 1. CT scan. Dilatation of prepontine cistern suggesting atrophy of the pons

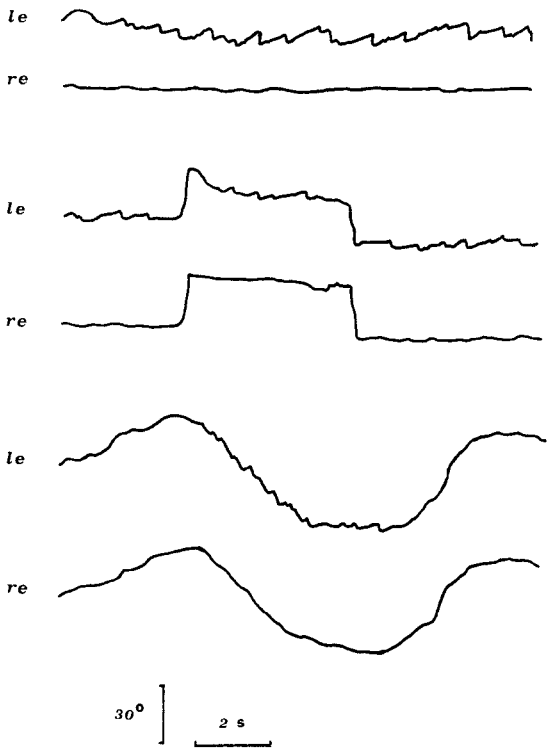

Fig. 2. Vertical electro-oculogram. Lines 1 and 2: primary position downbeat nystagmus (DBN) in the left eye. Lines 3 and 4: vertical saccades are normal in both eyes, but the left eye shows a drift towards the midline after an upward saccade, with superimposed DBN. Lines 5 and 6: in the left eye, downward pursuit is interrupted by DBN (*le:* left eye; *re:* right eye)

duction velocities were normal, but distal sensory potentials (sural nerve) could not be obtained, and H responses (soleus) were absent. EMG studies gave normal results. Standard blood and urine tests and ECG were normal. Copper, ceruloplasmin, folic acid, vitamin B_{12} blood levels, and phytanic acid (blood and urine) were also all normal. Enzymatic studies from a fibroblast culture (obtained by skin biopsy) showed no abnormality (hexosaminidase A/B, α/β galactosidase, glucosidase).

A diagnosis was made of sporadic ponto-cerebellar degeneration, possibly olivo-ponto-cerebellar atrophy.

Discussion

Our patient with probable sporadic (olivo-)ponto-cerebellar atrophy showed downbeat nystagmus limited to the left eye. Purely monocular vertical pendular nystagmus has been reported in 2 of 23 cases with multiple sclerosis and pendular nystagmus [1], but monocular vertical jerk nystagmus has not been reported in the literature, to our knowledge. Monocular DBN induced by lateral gaze has been reported in Wallenberg's syndrome [9], but primary position DBN is not present in this condition. In cerebellar degenerations, primary vertical jerk nystagmus, either up- or downbeating, has been described [6, 11, 13, 15, 17], but both eyes have always been affected (although sometimes with some degree of dissociation), as in other cases with acquired DBN [1, 4, 8].

Different physiopathological hypotheses have been suggested to explain the mechanism of DBN. According to Zee et al. [16], DBN may be due to defective downward pursuit, which is suggested by the usual finding of saccadic downward pursuit movements. However, it has been argued that the nystagmus itself may be at the origin of this pursuit defect [2, 10], and Baloh and Spooner [2] suggested that DBN corresponds to a central vestibular dysfunction, with imbalance of the vertical semicircular ocular reflexes by loss of flocculus inhibition. This imbalance was not confirmed by Halmagyi et al. [10], who explained the dependence of nystagmus slow-phase velocity on vertical head position in some cases by an imbalance in otolith-ocular reflexes, as previously suggested by

Chambers et al. [7]. In fact, the differences between the clinical features of DBN among the reported cases strongly suggest that there may be several types of DBN.

We must emphasize that in our case, the affected eye also showed a monocular tonic upward gaze paresis, with a downward drift following a normal upward voluntary saccade. Such a tonic paresis may be related to dysfunction of vestibulo-oculomotor fibres involved in vertical gaze [3, 5]. The medial longitudinal fasciculus has such fibres, but its involvement in our patient seems improbable because no internuclear ophthalmoplegia was present. Moreover, it has been suggested that this pathway is intact in DBN and transmits the inputs involved in vertical gaze, which generate the nystagmus [10]. However, another pathway may have been involved in our case, i.e. the brachium conjunctivum, which carries inputs (mostly excitatory) from the superior vestibular nucleus to the contralateral oculomotor nucleus, especially to the superior rectus subnucleus [5, 14]. As this subnucleus innervates the contralateral superior rectus, involvement of the left brachium conjunctivum could give rise to tonic upgaze paresis in the left eye, associated with ipsi- or contralateral cerebellar dysfunction (according to the caudal or rostral level of lesion). The role of cerebellar and brachium conjunctivum involvement has already been emphasized in the genesis of acquired pendular nystagmus, which may sometimes be monocular [1]. The fact that DBN in our case was limited to the left eye suggested that the vertical gaze system dysfunction was located rather close to the nuclear motoneurons involved in vertical gaze, where their afferent pathways become mainly unilateral. The presence of tonic paresis of upgaze in the eye with DBN may thus indicate dysfunction in the ipsilateral brachium conjunctivum, and suggests that alteration of tonic functions involved in vertical gaze may be an important factor for the genesis of primary position nystagmus beating in the opposite direction.

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