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Wall-eyed bilateral internuclear ophtalmoplegia presenting as monocular alternating nystagmus: a non-epileptic phenomenon in a case of diffuse axonal injury

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ABSTRACT – A 15-year-old male in a persistent vegetative state due to diffuse axonal injury presented with seizures and spontaneous alternating monocular nystagmus. The cranial MRI revealed diffuse axonal injury involving supratentorial and infratentorial structures, and the splenium of the corpus callosum. The monocular alternating nystagmus was thought to be independent of seizures and occurred as a result of diffuse axonal injury affecting the medial longitudinal fasciculus bilaterally. [Published with videosequences]

Key words: monocular alternating nystagmus, WEBINO, diffuse axonal injury, epilepsy

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Monocular nystagmus (MN) is a rare phenomenon. Unilateral disruption of the anterior visual pathway can produce MN, which usually occurs in a horizontal or vertical direction and can be jerky or pendular (Yee *et al.* 1979). This type of nystagmus is observed in different pathologies such as monocular blindness, spasmus nutans, brainstem infarction, or multiple sclerosis (Yee *et al.* 1979, Baram and Tang 1986, Donin 1967). Monocular abduction nystagmus can also be associated with lesions of medial longitudinal fasciculus (MLF). Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) is a variation of internuclear ophthalmoplegia (INO) associated with exotropia as a result of a bilateral, MLF lesion. Clinically, the patient is profoundly exotropic and has abducting nystagmus. The syndrome mimics bilateral medial rectus palsies, but the abducting nystagmus is the clinical clue (Mc Gettrick and Eustace 1985).

Epileptic nystagmus can be seen as an ictal phenomenon and it can be monocular or binocular (Kaplan and Leser 1989, Kaplan and Tusa 1993, Thurston *et al.* 1985, Jacome and Fitzgerald 1982). The aim of this article is to present a case of persistent vegetative state, seizures, and monocular alternating nystagmus (MAN) that developed as a result of diffuse axonal injury involving the brainstem. Because the MN appeared alternatively in both eyes, and occurred independently of epileptic seizures, we believe that the MAN was due to widespread lesions affecting the MLF bilaterally.

Case report

A 15-year-old male was involved in a car accident in November 2001. A ventriculo- peritoneal shunt was inserted after draining the temporo-parietal epidural hematoma on the right side. He remained in a persistent vegetative state until June 2003 when he was hospitalized again because of uncontrolled, generalized motor seizures.

On examination, he was not responsive to verbal stimuli. There was a reaction to painful stimuli only with flexion in the extremities. The eyes were closed. Both pupils were large and not reactive to light. The near response could not be evaluated. Bilateral exotropia, more prominent on the left, was observed in primary position.

The right eye exhibited frequent, horizontal, right beating nystagmus 2-16 beats, at intervals of 2-8 seconds. The left eye had a left beating nystagmus lasting for 1-8 beats, with irregular intervals and sometimes accompanying the right beating nystagmus (see video sequence). Adductions and vertical eye movements were not elicited with the doll's head maneuver. Ophthalmoscopic examination revealed no abnormality.

The past history of the patient indicated no eye problems prior to the head trauma. The observed eye movements were best described as monocular, alternating nystagmus. Lid movements accompanied the nystagmus from time to time in the right eye. During the hospitalization, the patient frequently presented focal seizures characterized by eyelid jerks on the right side, with clonic manifestation involving the hemiface, and sometimes resulting in a secondary generalized tonic-clonic seizure. On the EEG recording, high amplitude sharp waves, originating in the fronto-temporal regions, were more prominent on the right side, spreading to central parts of both hemispheres and lasting for 16 seconds. This wave was associated with the seizure, which originated from the right eyelid and became generalized. The interictal EEG showed diffuse background abnormalities consisting of theta and delta activity, more prominent in both fronto-temporal regions. Isolated or repeated sharp wave discharges of high amplitude, lasting 1-4 sec, were observed in both frontal regions, spreading to temporal areas as well. These high amplitude, sharp waves in the frontal regions of both hemispheres did not seem to be correlated with the abnormal eye movements.

Initially the patient was given carbamazepine (200 mg/day), increased later to 800 mg/day. The routine laboratory tests gave the following values: glucose level, 106 mg/dL; BUN, 7 mg/dL; Na, 138 mEq/L; K, 3.3 mEq/L; Ca, 7.8 mg/dL; WBC, 10.560; Hgb, 12.1; Htc, 34.2; Plt, 202000.

The cranial MRI revealed a central hyperintense, peripheral hypointense signal that depicted a central encephalomalacic area and hemosiderine rim in the pons and midbrain on FSE T2-weighted images *(figure 1)*. Gradient echo MR images showed a hypointense signal that depicted hemosiderine and chronic hemorrhage in bilateral lentiform nuclei, head of the right caudate nucleus, right frontal, bilateral temporal regions and third ventricular wall (*figure 2a)*, and brainstem *(figure 2b)*. The lesion in the tegmentum encompassed the medial longitudinal fasciculus, the oculomotor and trochlear nerve nuclei and the left medial lemniscus. It partially extended to the left posterior tectum, as well as to the pons at the boundary of the 4th ventricle. There were artefacts caused by the metallic clips in the right frontal region, and the metallic implant of the shunt catheter under the skin in the right temporo-occipital areas.

On the last examination in May 2004, he was seizure-free, but the level of consciousness and the nystagmus pattern was not changed.

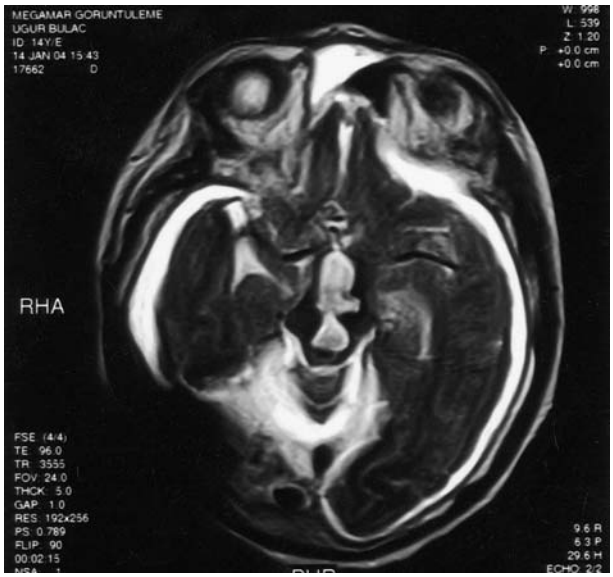

Figure 1. FSE T2 weighted images revealed central hyperintense, peripheral hypointense signal in the brainstem. There were artifacts caused by the metallic clips in the right frontal region and the metallic implant of the shunt catheter under the skin in the right temporo-occipital areas. And also bilateral subdural effusion is noted.

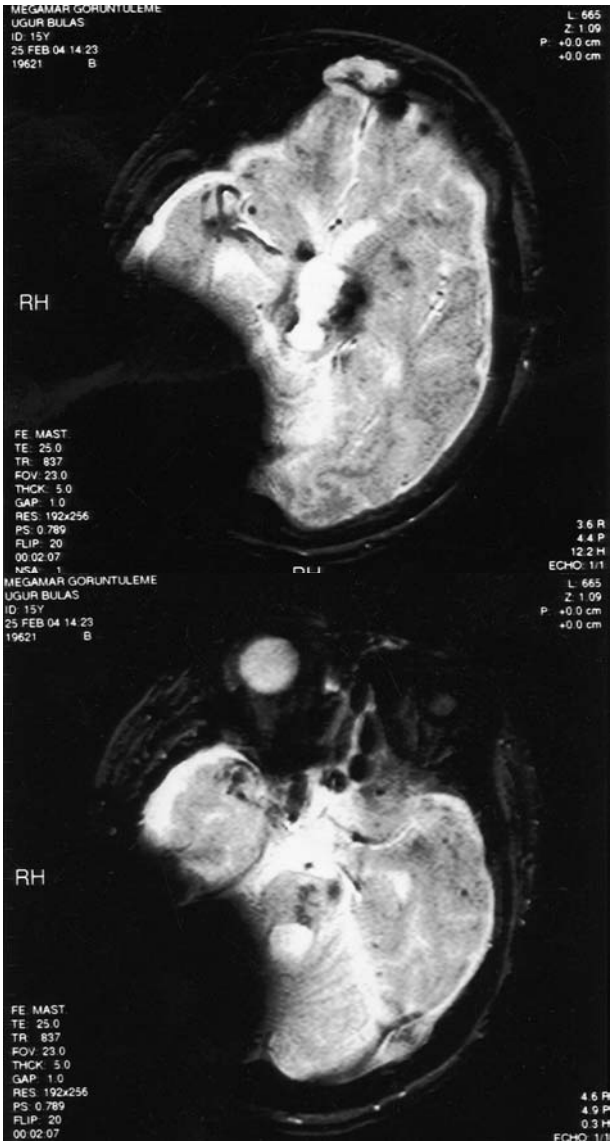

Figure 2. Gradient echo MR images showed hypointense signal that depicts hemosiderine and chronic hemorrhage in bilateral lentiform nuclei, head of the right caudate nucleus, right frontal, bilateral temporal regions and third ventricular wall (a) and brainstem (b).

Discussion

Ocular findings in this patient were compatible with nuclear third nerve palsy and wall-eyed bilateral internuclear ophthalmoplegia (WEBINO). The unusual feature was the presence of a non-periodic, monocular nystagmus with different frequency and duration in both eyes, being more evident in the right eye. This occurred intermittently, probably when he had spontaneous, nonpurposeful, roving eye movements which gave the erroneous impression of spontaneous, alternating nystagmus.

Since the patient has widespread MR lesions and partial onset secondary generalized epileptic seizures, the monocular alternating nystagmus could be assumed to be an epileptic seizure. Epileptic nystagmus can be binocular and monocular as a rare manifestation of seizure activity (Kaplan and Leser 1989, Kaplan and Tusa 1993, Thurston *et al.* 1985, Jacome and Fitzgerald 1982). The EEG accompaniment of horizontal binocular epileptic nystagmus varies for frontal, temporal, parietal or occipital lesions on one side (White 1971). The eye and its movements may be ipsilateral or contralateral to the EEG focus (Horita *et al.* 1977). Monocular nystagmus has been described in a patient with epilepsy originating from a contralateral lesion (Grant *et al.* 2002). Because the eye activity alternated between both eyes in the horizontal plane as monocular activity, and the activity in both eyes was not correlated with the EEG, we have reached the conclusion that, in our case, this phenomenon is independent of epileptic seizures. Even though it is known that epileptic binocular nystagmus appears with contralateral and rarely with ipsilateral lesions, it is difficult to explain ipsilateral and alternating contralateral monocular nystagmus with the same epileptic focus. Another type of nystagmus that has been associated with seizures in a single case after hypoxic ischemic encephalopathy, is periodic, alternating nystagmus (PAN) (Moster and Schnayder 1998). However, the pattern of nystagmus in our patient, which varies in direction, duration and frequency in two eyes, was not compatible with PAN in which both eyes exhibit primary position nystagmus that stops for a few seconds after 60 to 120 seconds and then, starts beating in the opposite direction.

Brainstem injuries (BSI) can be classified as primary or secondary. Primary lesions are those that result from the initial traumatic force, whereas secondary ones are those that develop subsequent to the initial trauma (Gentry *et al.* 1989, Adams *et al.* 1977, Adams *et al.* 1980, Adams *et al.* 1982). The most common type is associated with widespread diffuse axonal injury (DAI). Brainstem DAI rarely occurs without the presence of histologically similar lesions in the corpus callosum and deep cerebral white matter, and the lesions are characteristically located in the dorsolateral quadrants of the rostral brainstem (midbrain and upper pons) (Adams *et al.* 1977, Adams *et al.* 1980, Adams *et al.* 1982). Secondary BSI occur at a later stage as a result of descending tentorial herniation. These are termed Duret hemorrhages and consist of centrally placed collections of blood in the tegmentum of the rostral pons and midbrain (Adams *et al.* 1977, Adams *et al.* 1980, Adams *et al.* 1982, Parizel *et al.* 2002). We associated our case with DAI based on the infra and supratentorial locations of the lesions, particularly in the cerebral hemispheres and the splenium of corpus callosum. In our case, the brainstem lesion in the tegmentum encompassed the medial longitudinal fasciculus, the oculomotor and trochlear nerve nuclei and the left medial lemniscus.

The fact that the eyes exhibit an alternating monocular nystagmus and that the two eyes do not assume the same central position, has led us to believe that the MN is due to bilateral mesencephalic MLF lesions and the nystagmus was caused by the adaptational attempt of the brain to correct hypometric saccades of the weak medial rectus. □

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