Epileptic nystagmus in infancy

C.M. Harris*, S. Boyd, K. Chong, W. Harkness, B.G.R. Neville

Great Ormond Street Hospital for Children, NHS Trust and Institute of Child Health, University College London, London WC1N 3JH, UK

Received 20 March 1997; accepted 9 April 1997

Abstract

Epileptic nystagmus (EN) is a rare form of nystagmus that occurs only during epileptic seizures. We report an infantile case in which EN was first noted at 10 days of age. Electronystagmography showed a right-beating nystagmus with predominantly linear slow phases that traversed the midline. Neuro-imaging revealed dysplasia of the left middle temporal gyrus extending posteriorly into the parieto-occipital cortex. The right hemisphere and subcortical structures appeared normal. Perfusion studies demonstrated interictal hypoperfusion with ictal hyperperfusion in the left temporal lobe. Electroencephalography demonstrated spiking over the left temporal-parieto-occipital region. Following extensive surgical resection of this area and weaning of anti-convulsants, the child has remained seizure-free without nystagmus. This case demonstrates the cortical origin of EN, and shows that infant cortex has functioning efferent connections to brainstem oculomotor centres from 10 days of age. © 1997 Elsevier Science B.V.

Keywords: Visual development; Cortex; Infant vision; Epilepsy; Optokinetic nystagmus; Smooth pursuit

1. Introduction

Epileptic nystagmus (EN) is a rare form of rhythmic eye oscillations that occur only during epileptic seizures. In the majority of reported cases the nystagmus is jerk, horizontal, conjugate, and usually accompanied by tonic horizontal eye and head deviations. Typically the seizure focus is in the temporal-occipital-parietal region (T-O-P) of the cerebral hemisphere contralateral to the direction of the nystagmus beats, and in some cases, the nystagmus may result from excitation of the cortical centres for smooth pursuit or optokinetic eye movements (Kaplan and Tusa, 1993). In view of the uncertain role of cerebral cortex in the control of infant eye movements, the presence of EN in infancy is of special interest. We present a child in whom EN began at 10 days of age, which was subsequently shown to have a cortical origin.

2. Patient and investigations

The patient was a female child of non-consanguineous parents born at 42 weeks gestation by breech delivery with a weight of 3.9 kg following a pregnancy complicated by a small bleed at 9 weeks gestation. Apgar scores were 2, 8, 10 at 1, 5, 10 min and oxygen was required by mask. There were no neonatal problems although parents reported that the infant seemed placid. The first recognised seizure occurred at 10 days, and lasted about 1 min with the eyes rolling up and flicking from side to side, and with lip-smacking and possible altered conscious level. At 13 days the seizure pattern changed, so that following the abnormal eye movements there was jerking of the leg followed by jerking of the arm on the same side (usually right), deviation of the head to the right and secondary generalisation. These seizures lasted 1–2 min and occurred up to 50 times per day. The following investigations at the age of 17 days were normal: full blood count, urea, glucose, sodium, calcium, magnesium, C-reactive protein, serum ammonia, skull ultrasound and X-ray; chest X-ray, organic acids, amino acids, bile acid analysis, and lumbar puncture.

*Corresponding author.
By 2 months developmental progress was slow, and there was a decrease in movements of the right side with a reluctance to respond to visual stimuli presented to the right visual field, but orientation to the right for sounds was present.

Magnetic resonance imaging (MRI) (Fig. 1) revealed a region of cortical dysplasia in the left middle temporal gyrus extending into the left angular gyrus (Brodmann areas: 21, 37, and 39) with abnormal underlying white matter. Adjacent white matter was also abnormal indicating immaturity of myelination in the middle left temporal and occipital lobes. Some calcification was shown in the posterior temporal cortex on CT. The left parieto-insular cortex appeared to be within normal limits. There was no enhancement with gadolinium on MRI. The right cerebral hemisphere and subcortical structures appeared normal. The seizures proved intractable to anti-convulsant therapy, and the patient was referred to this tertiary hospital for evaluation for possible surgery.

Time-locked video/digital EEG recording was carried out at the age of 8 months. The interictal record showed almost continuous spiking at around 2 / s (150 μv) over the left posterior temporal regions (overlying Brodmann areas 39). Rhythmic 6 / s activity appeared over the left posterior region (overlying areas 37 and 39), spreading rapidly to the mid-temporal and Sylvian electrodes (overlying areas 21 and 22), followed within 2 s by a tonic deviation of the eyes to the right. 10 s later a marked nystagmus was seen as the predominant motor manifestation of the seizure (Fig. 2A). The right hemisphere became involved in the seizure, but only after the onset of the nystagmus.

At 8 months, ictal and inter-ictal SPECT cerebral blood perfusion studies were carried out with 99mTc-HMPAO. Interictally, there was hypo-perfusion in the left temporal lobe, which became clearly hyper-perfused during a typical seizure (radio-label injected at onset of nystagmus).

Eye movements were recorded at 9 months using dc-electrooculography and video. Seizures typically began with a right tonic deviation of the eyes followed by right-beating nystagmus with approximately linear slow phases traversing the midline and almost the full horizontal oculomotor range at times. The nystagmus beat frequency was erratic, but was approximately 4 cycles per second at it most intense (Fig. 2B). Nystagmus beats were frequently accompanied with blinking, which caused a concomitant upward movement of the eyes, which was thought to be a normal Bell’s phenomenon. Similar ictal episodes occurred in complete darkness. Tonic deviations to the limit of gaze were observed ictally in either direction, but not interictally. During eye movement recording, seizures were frequent and usually followed by ‘absent’ periods during which saccadic eye movements could not be elicited. Interictally, full-field binocular optokinetic nystagmus was normal for stimulus motion to the right but completely absent to the left (Fig. 2C). Vestibular nystagmus induced by constant rotation in the dark showed asymmetry with lower gain for slow phases to the left.

At 11 months, a surgical resection involving the left temporal, occipital and parietal cortices was performed, which was seen on post-operative CT. Intra-operative electrocorticography showed extensive spiking over the left posterior T-O-P. At the time of writing the patient has been without medication for 12 months. Throughout the post-operative period there have been no seizures and no nystagmus.

3. Discussion

Tonic deviations of the eyes and head are common in epilepsy, but nystagmoid eye movements are an unusual ictal manifestation, and have been observed in less than 10% of cases with occipital lobe epilepsy (Salanova et al., 1992). In the majority of reported cases the EN has been horizontal and conjugate with a focus in the temporo-occipital-parietal cortex contralateral to the direction of the nystagmus beats (Kaplan and Tusa, 1993). EN with an onset in the first year of life is rare, and to our knowledge has only been reported by Giove (1960) in a 5-month-old with unspecified beat direction and a focus over the temporo-occipital region.

Two distinct types of conjugate horizontal EN have been described. In EN-1, the nystagmus is confined to the opposite side of the focus with contraversive quick-phases and decelerating ipsiversive slow phases that do not cross the midline (Thurston et al., 1985). It has been proposed that cortical saccade centres are stimulated by the epileptic discharges, and the slow phases reflect poor gaze-holding, possibly secondary to medication or coma, although this has been questioned (Morrow, 1994). In EN-2, the nystagmus has a large amplitude and the slow phases cross the midline, which have been shown in two cases to have roughly constant velocity (linear slow phases) (Furman et
Fig. 2. (A) Selected channels from Video-EEG telemetric recording. (Electrodes over the vertex and the right parietal region were technically unsatisfactory). Channel 1 is an electro-oculogram. Time lines are 2 s apart. Note the frequent spikes seen over the posterior half of the left hemisphere. Approximately 2 s before the clinical attack (C) is noted there is a build up of 6 to 7 Hz activity over the left parieto-occipital region (E), rapidly increasing in amplitude, and followed in turn by the appearance of nystagmus after about 10 s. (B) Ictal dc-electrooculographic recording of (upper trace) horizontal eye movements showing right beating nystagmus with rapid roughly linear slow phases and (lower trace) vertical eye movements showing frequent blink artefacts coinciding with horizontal quick-phases. (C) Interictal horizontal recording showing normal full-field OKN with curtain rotating to patient’s right (upper trace) and absent OKN with curtain rotating to the left.

al., 1990; Tusa et al., 1990). It was quite clear from both video and EOG that our patient exhibited EN-2.

In EN-2 it has been proposed that the ipsiversive slow phases result from excitation of the cortical smooth pursuit or optokinetic centres (Furman et al., 1990; Tusa et al., 1990; Kaplan and Tusa, 1993). In our patient, the region of cortical dysplasia extended from the middle temporal gyrus (Brodmann area 21) posteriorly encompassing parts of areas 37 and 39, which overlap the putative cortical region of visual motion sensitivity (V5) and the presumed homologue of monkey MT/MST (junction of areas; 19, 37, 39), which is important for the generation of ipsiversive smooth pursuit (Lekwuwa and Barnes, 1996). Thus it seems plausible that the EN originates from excitation of this oculomotor centre. The absence of ipsiversive OKN slow phases is also consistent with a lesion in this region of T-O-P. This implies a lack of afferent visual motion information from ipsilateral striate and extrastriate cortex relaying motion from the contralateral visual field, and from the callosal connection from the contralateral cortex that relays motion signals from the ipsiversive visual field.

Typically the nystagmus commenced some seconds after a tonic deviation contraversive to the focus. This sequence suggests that the contraversive tonic deviation, which is a common ictal manifestation, has a low triggering threshold and occurs early in the seizure. Whereas, the slow phase oculomotor centre effectively has a much higher threshold (Tusa et al., 1990), or equivalently, can only be activated after disinhibition from more remote centres, such as frontal fixation circuits.

Although the pathway that mediates EN-2 slow phases cannot be precisely located, there is little doubt that it has a cortical origin. It has been claimed that infantile smooth pursuit does not develop until 2 months (Aslin, 1981). It has also been postulated that human neonatal OKN may be initially mediated subcortically via a direct retino-pretectal pathway, which is eventually superseded by the maturation of parallel cortical pathways (Hoffman, 1982), although this has been disputed (Harris et al., 1996). Thus, this rare case demonstrates that by 10 days, the infant cerebral cortex has at least functioning efferent connections to brainstem oculomotor centres.
References