Case Report

Epileptic monocular nystagmus and ictal diplopia as cortical and subcortical dysfunction☆

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Abstract

We present the case of a patient with ictal monocular nystagmus and ictal diplopia who became seizure-free after resection of a right frontal focal cortical dysplasia (FCD), type 2B. Interictal neuroophthalmological examination showed several beats of a monocular nystagmus and a spasm of the contralateral eye. An exclusively ictal monocular epileptic nystagmus could be an argument for an exclusively cortical involvement in monocular eye movement control. The interictal findings in our patient, however, argue for an irregular ictal activation of both the cortical frontal eye field and the brainstem.

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1. Introduction

In the context of a historical debate of cortical versus subcortical control of eye movements (Helmholtz, published in 1867, versus Hering, published in 1868) [1,2], the existence of epileptic monocular nystagmus has been disputed [3–5]. Diplopia and strabismus as epileptic phenomena are rarely reported but could be an argument for an exclusively cortical involvement in monocular eye movement control [6,7]. We present the case of a patient with monocular nystagmus and ictal diplopia who became seizure-free after resection of a right frontal focal cortical dysplasia (FCD 2B).

2. Case report

A right-handed 36-year-old man had video-EEG monitoring in our epilepsy surgery program. Since the age of 16, this patient had seizures 30 times a day, with turning of the head and asymmetric gaze shift of both eyes to the left, more prominent of the adducting right than of the abducting left eye, with resulting strabismus convergens and complaint of diplopia, accompanied by a 4- to 8-Hz prominent leftward nystagmus of the left eye. Noninvasive and invasive video-EEG monitoring documented seizure origin from a right frontal lesion (focal cortical dysplasia (FCD) type 2B by MRI criteria; Fig. 1). The patient was seizure-free subsequent to extended resection of the right frontal lesion with the histopathological diagnosis of FCD 2B, with a follow-up of 1 1/2 years so far.

Interictal neuroophthalmological examination showed several subtle beats of a monocular nystagmus of the right eye in an extreme lateral gaze to the right and a spasm of the right eyelid, with other findings normal (Supplementary video 1; Table 1). A second neuroophthalmological examination replicated the findings. Five months after successful surgery, only a minimal residual horizontal gaze nystagmus was found.

Ictal videos (Supplementary videos 2 and 3) show, with a tonic leftward gaze deviation, the evolution of a convergent strabismus accompanied by a nystagmus of the left eye. Other videos recorded while monitoring with subdural grid electrodes show seizures out of sleep (Supplementary videos 4 and 5). Note that the head and eyes turn to the left; the eyes are convergent; the right eye has maximal adduction to the nose. The left eye shows a prominent monocular nystagmus to the left (EEG artifact in Fig. 1). The left palpebral fissure opens earlier and wider than the right.

3. Discussion

We documented epileptic monocular nystagmus and strabismus in videos and EEG, with a right frontocentral EEG seizure pattern and unilateral left-sided eye artifacts (Fig. 1). The MRI shows a lesion.

Abbreviations: FCD, focal cortical dysplasia.

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near the right frontal eye field (Fig. 1). Interictal nystagmus of the contralateral eye, interictal contralateral facial spasm before surgery, and normalization of neuroophthalmological findings after surgery support the hypothesis of irregular brainstem imbalance that overrides the regular binocular cortical and subcortical movement control mechanisms in this patient. Interictal ipsilateral, right-sided monocular nystagmus and right facial spasm of the eyelids point to a dysfunction of the right brainstem (nuclei of cranial nerves VI and VII), possibly
with “Todd’s paralysis” of the analogous left brainstem nuclei because of the high seizure frequency. Our hypothesis for regular and, in our case, irreglar ictal activation of the cortical frontal eye field and the brainstem is illustrated in Fig. 1.

One previous case study of monocular nystagmus showed an EEG sample with supposed unilateral eye artifacts [3]. The artifacts, however, turned out to display "pops" [4]. Another study presented a patient with generalized epilepsy with severe mental retardation suggestive of diffuse encephalopathy, with a marked monocular visual deficit of the eye affected by the nystagmus so that a nonepileptic origin of the monocular nystagmus must be suspected [5].

To our knowledge, there are only two case reports of ictal strabismus. In one case, the authors supposed a loss of fusional control during the seizures [6]. A recent study of ictal strabismus describes two patients with disconjugate contraversive horizontal eye movements in invasive monitoring with subdural grid electrodes, one during electrical cortical stimulation of the frontal eye field and the other during focal seizures with spread from the supero-posterior Sylvian bank to the adjacent regions, including the frontal lobe [7]. The interictal ophthalmological findings were normal, so that the authors concluded that cortical function in the frontal eye field comprises both contralateral version and vergence, without evidence of brainstem involvement.

4. Conclusion

With regard to the historical debate by Helmholtz and Hering [1,2], ictal monocular nystagmus and strabismus in videos and EEG in our patient, together with interictal monocular nystagmus of the contralateral eye and postoperative normalization of ophthalmological findings, argue for Hering who claimed both eyes to be, as a rule, directed together according to cortical intention and subcortical coordination (“Doppelaue”) as opposed to Helmholtz who proposed cortical binocular coordination as a result of learning. Hering’s rule of simultaneous cortical and subcortical control of eye movements was, according to our observation, transiently overridden by epileptic activity with coactivation of the right frontal cortical focus and the subcortical nuclei of cranial nerves III, VI, and VII in the ictal state with lateralization to the left eye (activation) and in the interictal state with lateralization to the right eye (Todd’s paralysis).

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ebcr.2013.05.002.

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