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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.

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

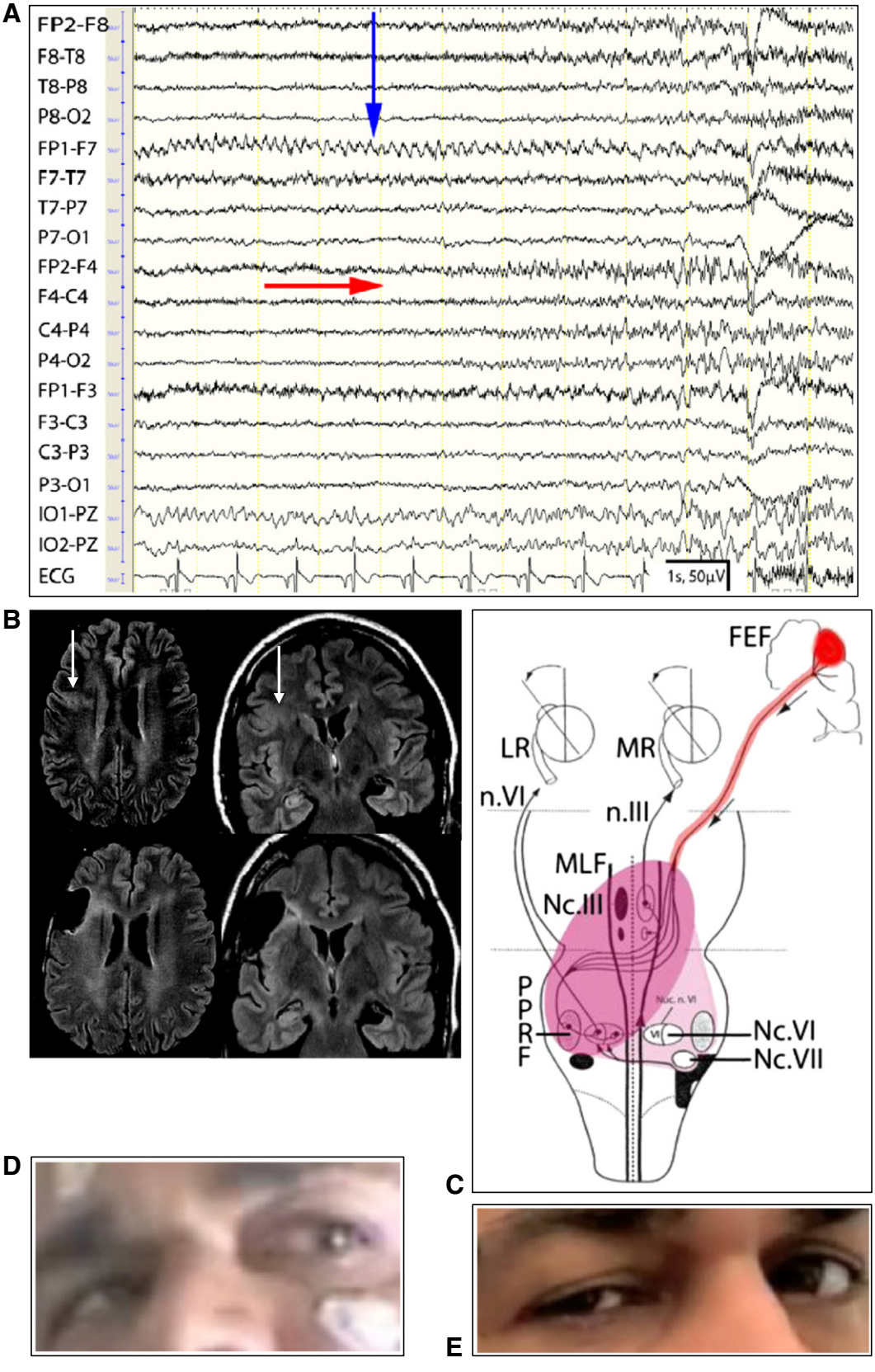

Table 1
Neuroophthalmologic examinations, seizures, and medications.

	Neuroophthalmology (interictal)	Seizures	AEDs
Preoperative (6 months)	Monocular lateral gaze nystagmus, right eye[a]	Left eye monocular nystagmus and strabismus, 10 per day	OXC 1200 mg, LEV 3000 mg (10 days after exchange VPA → OXC)
Preoperative (6 weeks)	Dissociated lateral gaze nystagmus, right > left eye[a]	The same, 10 per day	The same (10-OH OXC 13.4 mg/L)
Postoperative (5 months)	Lateral gaze nystagmus (minimal)[a]	Seizure-free	OXC 1800 mg, LEV 3000 mg
Postoperative (6 months)	Lateral gaze nystagmus (to right more than to left)[b]	Seizure-free	OXC 1800 mg, LEV 3000 mg (10-OH OXC 26.8 mg/L)
Postoperative (12 months)	Slight lateral gaze nystagmus (to right more than to left)[b]	Seizure-free	OXC 1800 mg, LEV 1000 mg (10-OH OXC 25.2 mg/L)

Abbreviations: AEDs = antiepileptic drugs, OXC = 10-OH-Oxcarbazepine, LEV = levetiracetam, VPA = valproic acid.
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with "Todd's paralysis" of the analogous left brainstem nuclei because of the high seizure frequency. Our hypothesis for regular and, in our case, irregular 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 ("Doppelauge") 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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Fig. 1. Hypothesis of brain stem involvement in epileptic monocular nystagmus. A: EEG: channels FP1-F7, F7-T7, and IO1-PZ (left infraorbital) show an eye artifact by monocular nystagmus of the left eye, with evolution of frequency (initially 7 Hz, later 4 Hz); vertical arrow; the EEG with reference PZ shows most prominent deflections in F7 and IO1 which indicate an additional vertical component of the rapid monocular eye movements (Supplemental material). From second 5 on, a rapid EEG seizure pattern evolves over the right frontocentral region (phase reversal at channel F4-C4; horizontal arrow). B: Pre- and postoperative MRI (coronal und transversal planes); white arrows indicate the right frontal focal cortical dysplasia. C: Hypothesis for brainstem involvement: a) *Usual ictal activation*: right frontal cortical seizure activity (FEF = frontal eye field; dark red) propagates to the contralateral left paramedian pontine reticular formation (PPRF), nucleus nerve VI (nc.VI), and via the right medial longitudinal fasciculus (MLF) to the contralateral right nucleus nerve III (nc.III), with subsequent activation of the lateral rectus muscle of the left eye (LR) and the medial rectus muscle of the right eye (MR); effect: gaze of both eyes to the left. b) *Irregular ictal activation*: in addition, activation of the left nc.III; effect: coactivation of the medial and lateral rectus muscles of the left eye, with left monocular nystagmus, strabismus, and diplopia (D). The comparatively wide left palpebral fissure could result from innervation of the levator palpebrae muscle of the left eye (innervation by the 3rd nerve, n.III). Usual plus irregular ictal brainstem activation is marked in dark magenta. c) *Irregular interictal state* (bright magenta): ictal dysfunction with predominance of the left brainstem is followed by residual dysfunction of the right nc.VI and nc.VII. This results in a net hyperfunction of the right n.VI with monocular lateral nystagmus of the right eye and involves the right n.VII with right periocular spasm (E). For further illustrations, see also the videos in the Supplemental material. The dysfunction of the brainstem ceased after successful right frontal cortical epilepsy surgery and neuroophthalmological findings normalized.