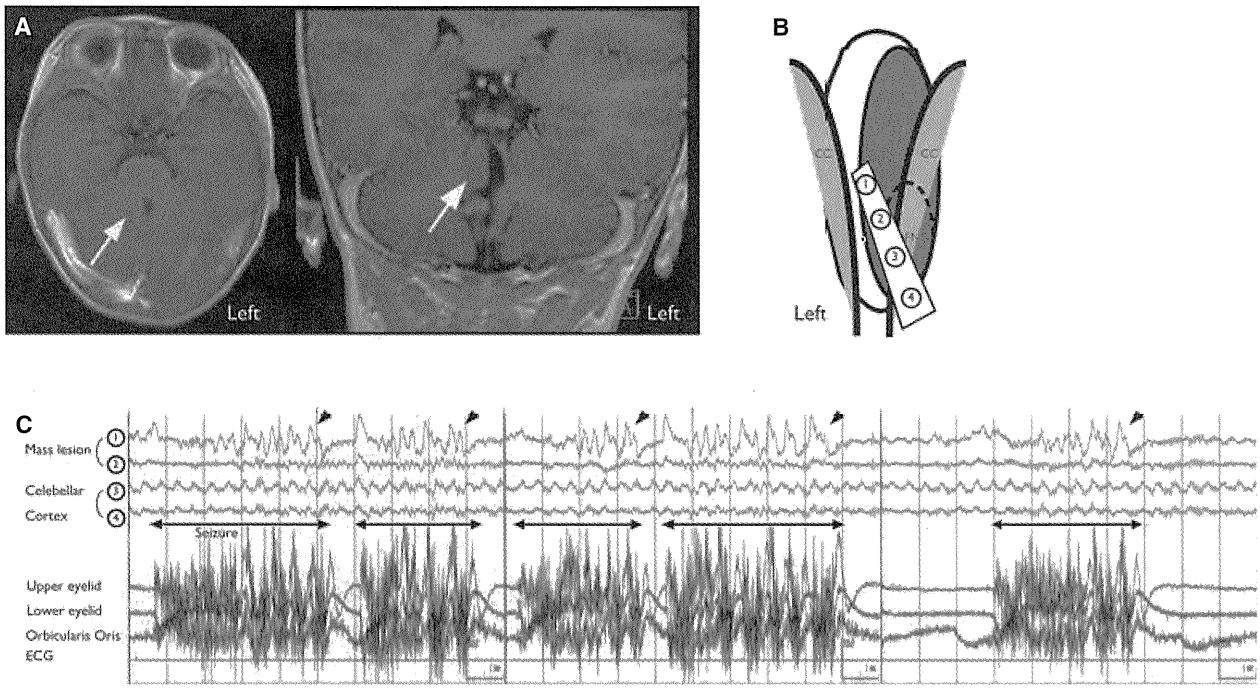

Figure 1.

Brain magnetic resonance imaging (MRI) and electrical activity of the tumor and cerebellar cortices at the time of hemifacial seizures. (**A**) Preoperative gadolinium-enhanced T_1-weighted MRI: The right side of the fourth ventricle was occupied by an iso-intensity tumor (arrow). (**B**) The schema of intraoperative electroencephalography (EEG) recording in the fourth ventricle: (Tumor) The gray oval indicates the mass lesion; (CC) The light gray semicircle indicates bilateral cerebellar cortices; and (FNN) The broken line indicates the facial nerve nucleus (not directly shown). A strip electrode with four contacts (2.3-mm diameter, 10-mm interelectrode spacing) was used. No. 1 and No. 2 electrodes were attached on the tumor and No. 3 and No. 4 were attached on the right cerebellar hemisphere. The numbers on the strip correspond to the numbered records of electrical activity shown in panel C. (**C**) Electrical activity of the gangliocytoma and clinical seizures. The upper two records (No. 1 and No. 2) show electrical activity detected by the strip electrode on the tumor. The EMG indicates hemifacial seizures. Theta waves on the mass lesion (No. 1, indicated by arrow heads) correspond to the seizures (indicated by arrows). The recordings of electrical activity in the right cerebellar cortex (No. 3 and No. 4) were contaminated by artifact from the arterial pulse.

The patient has been seizure-free since the operation without any neurologic deficit, and he has developed to be age-appropriate at the age of 2 years.

Discussion

The present study confirms the correlation between DNFFV and HFS directly using intraoperative electroencephalography (EEG) and EMG monitoring for the first time. Recently, several case reports have described paroxysmal episodes induced by DNFFV, but no report has described a direct correlation between abnormal current in the DNFFV and seizure manifestation (Al-Shahwan et al., 1994; Harvey et al., 1996; McLone et al., 1998; Arzimanoglou et al., 1999; Chae et al., 2001; Delalande et al., 2001; Mesiwala et al., 2002; Pontes-Neto et al., 2006; Dagcinar et al., 2007; Park et al., 2009).

There are two hypotheses for the causal relationship between DNFFV and HFS; one involves the cerebellum (Harvey et al., 1996; Chae et al., 2001), and the other involves the facial nerve nucleus (FNN) (Delalande et al., 2001). Some previous reports showed abnormal electrical activity in the cerebellar cortex and hypothesized a cerebellar seizure (Harvey et al., 1996; Mesiwala et al., 2002). However, one case showed abnormal cerebellar activity following the intralesional polyspikes of depth electrodes (Mesiwala et al., 2002); therefore, the abnormal cerebellar activity might represent secondary activity spread from the lesion. Recently, Delalande et al. (2001) suggested that the seizures arose from inside the lesion because of the anatomic configuration, the presence of intralesional beta and theta waves, and the lack of seizures after tumor resection. In our current case, intracranial EEG indicated that the theta waves of the tumor corresponded directly to HFS. Furthermore, no activity congruent with HFS was found in either cerebellar cortex. Therefore, these results strongly support the FNN hypothesis.

Based on our survey, there are 17 published papers containing 20 cases with DNFFV and 85% (17 of 20 cases including our case) of children with DNFFV showed hemifacial spasms (Supporting Information). In those three cases without HFS, MRI showed tumor involving the cerebellar vermis and seizures with shaking of the both upper extremities, jerking of the lower and upper extremities and trunk and myoclonic movements of the proximal lower extremities (McLone et al., 1998; Mink et al., 2003; Koh et al., 2010). We speculate that most cases with DNFFV cause HFS, whereas DNFFV involving only the cerebellar vermis may manifest differently. Interestingly, except for one case, HFS has always been reported as occurring on the side of the face ipsilateral to the tumor, suggesting the infranuclear effect of the tumor. These findings also support the FNN hypothesis. This hypothesis of cranial nerve origin, however, cannot explain the secondary generalization (Mesiwala et al., 2002). To confirm more positive pathophysiology, simultaneous monitoring of DNFFV, cranial nerve nuclei including the FNN, cerebellar cortex, and scalp-EEG are warranted.

Sometimes the tumor in question is located in a hazardous area, and the border between the tumor and normal tissue is not necessarily obvious macroscopically; therefore, in surgical procedures, careful functional monitoring is needed. In fact, some patients with DNFFV have been left with severe deficits after surgical treatment (Chae et al., 2001; Delalande et al., 2001; Mesiwala et al., 2002). Our current study also shows the utility of simultaneous intraoperative monitoring by both intracranial EEG and facial EMG monitoring. By means of this monitoring, we could confirm the disappearance of seizures directly and avoid a hazardous resection procedure.

In conclusion, we suggest that HFS associated with DNFFV is a specific clinical entity, and that the HFS are caused by a direct effect from the DNFFV to the FNN.

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Disclosure

None of the authors has any conflict of interest to disclose. We have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Table S1. Characteristics of seizure induced by dysplastic neurolonal lesion in the fourth ventricle.

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