

Cerebellar seizures

Report of 2 cases

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Epilepsy, especially with refractory seizures, is thought to arise only from cortical lesions or substrate. The authors report on 2 patients with refractory epilepsy and cerebellar lesions. Depth electrodes were placed within the cerebellar lesions in both patients, and intracranial electroencephalographic recordings showed seizure origin from the cerebellar lesions. One patient eventually attained seizure control with antiepileptic drugs. The other case involved a child with generalized myoclonic epilepsy associated with a pilocytic astrocytoma of the cerebellum. This patient obtained seizure control following gross-total resection of the tumor.
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KEY WORDS • cerebellar seizure • pilocytic astrocytoma • low-grade glioma •
depth electrode • epilepsy surgery

CLASSIC epilepsy teachings dictate that epilepsy arises only from the cerebral cortex, not subcortical structures like the cerebellum.^{10,12} As detailed by McCrory et al.,¹² John Hughlings Jackson, who has been credited with developing the modern concept of epilepsy, observed and described cerebellar seizures in the late 1800s and early 1900s. His clinical concept of cerebellar seizures is thought to have been overshadowed by studies of decerebrate rigidity. Later, Penfield and Erickson postulated that “it is conceivable that some seizures characterized by stereotypic movements, or slow postural changes, are due to a cerebellar discharge ... but we have never found clinical confirmation of such a conclusion.”¹⁵ Since then there have been a number of cases demonstrating that cerebellar seizures can occur.^{1–11,13,14,16,19}

In this article we document 2 such patients. The first is a child with a hypodense lesion on MRI deep in the cerebellum resulting in complex partial seizures that were eventually medically controlled. The second case is a child who had generalized epilepsy associated with a pilocytic astrocytoma of the cerebellum. In both cases we used depth electrodes to monitor seizure activity from the lesions, verifying that these abnormalities were, in fact, the origin of the seizures.

Case Reports

Case 1

History and Examination. This 8-year-old boy pre-

sented with intractable, symptomatic partial-onset seizures. He had no family history of epilepsy. His seizures started at 3 years of age and had previously been treated with oxcarbazepine, topiramate, valproic acid, clonazepam, phenytoin, levetiracetam, carbamazepine, pregabalin, and felbamate. At the time of examination he was experiencing approximately 20 complex partial seizures per month.

Magnetic resonance imaging demonstrated a hypodense, nonenhancing abnormality deep in the right cerebellum (Fig. 1). In 2 previous biopsies it was concluded that the abnormality was consistent with either gliosis or an LGG. His video-EEG monitoring with scalp electrodes showed no interictal epileptiform activity. Multiple simple and complex partial seizures were recorded in this patient. Electrographically, there were no changes seen with the simple partial seizures. His complex partial seizures lateralized to the left hemisphere and his clinical features had a consistent semiology with right body motor activity. Magnetoencephalography and magnetic source imaging were ordered to aid in the localization of spontaneous epileptiform discharges, receptive language areas, and somatosensory cortex prior to surgery. During the magnetoencephalography the patient had frequent epileptiform discharges that localized to the left temporal and frontal lobes. Language localization showed bilateral activation.

Procedure. Based on these data, the decision was made to perform a repeat biopsy and insert a 4-contact depth electrode within the lesion for ictal clarification.

Abbreviations used in this paper: EEG = electroencephalography; LGG = low-grade glioma.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.

Two cases of cerebellar seizures

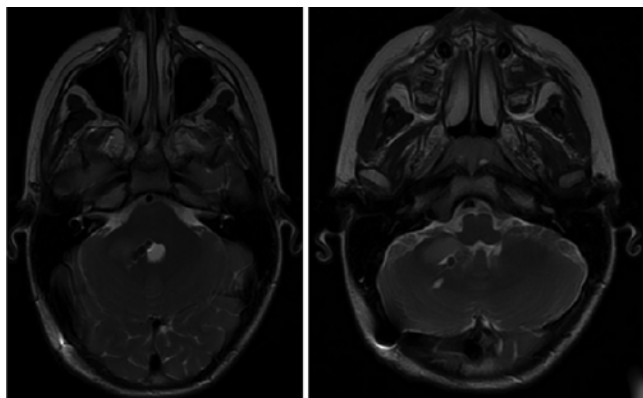


Fig. 1. Case 1. Axial T1-weighted noncontrasted images demonstrating the cerebellar expansile lesion with a 4-contact depth electrode placed into the stereotactic biopsy tract for ictal recording. Note the hyperintense signal from the previous biopsy tract as well.

The biopsy and electrode placement were performed using MRI-guided frameless stereotaxy, without complication (Fig. 1 right).

Outcome. Monitoring from the depth electrode indicated rare spikes arising from the right cerebellar lesion. The patient had 2 secondarily generalized tonic-clonic seizures characterized by turning from his side to a supine position with flexing of both legs and the right arm. He had subtle clonic jerks of the right arm followed by postictal Todd paresis. Onset of these events was clearly recorded by the cerebellar depth electrode. The EEG recording showed onset with rhythmic theta frequencies from the right cerebellar depth electrode in contacts 1–4 (Fig. 2). After the event was over, he had attenuation in all 4 contacts.

Although the seizures were found to be arising from the cerebellar lesion, the family decided to continue with medication management because of their concern for possible postoperative deficits. Postoperatively, the patient's seizures have been fairly well controlled on lamotrigine and carbamazepine. He has 1 brief complex partial seizure (approximately 20–30 seconds in duration) every 3–4 months, and has gone as long as 1 year free of seizures. Follow-up MRI studies have shown that the lesion has remained stable for 4 years. At the moment no further surgery has been entertained. His third biopsy result was consistent with gliosis, but LGG remains a concern.

Case 2

History and Examination. This patient was a 3-year-old boy with no family history of seizures or brain tumors who presented with right hand tremor at the age of 4 years. Neuroimaging showed a right cerebellar mass. He underwent partial resection at another institution, which revealed a Grade I cerebellar pilocytic astrocytoma. He did well for 1 year, and then had onset of generalized seizures. The initial event was a generalized tonic-clonic seizure, but he then developed refractory generalized myoclonic seizures, accompanied by vocalization; he had more than 20 events per hour. These episodes were initially not thought to be related to the cerebellar tumor and did not

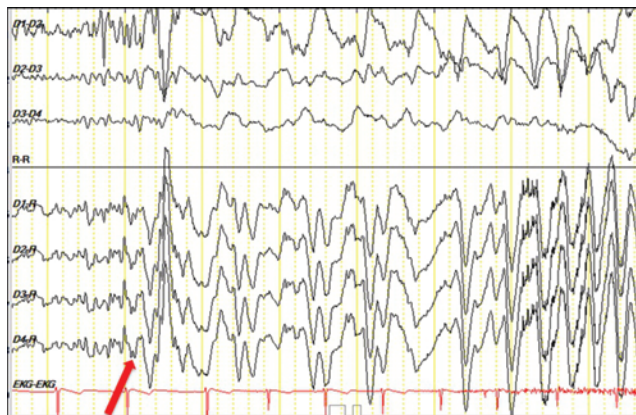


Fig. 2. Case 1. This EEG reading demonstrates the onset of the cerebellar seizure as recorded from the 4-contact depth electrode. The first 3 channels are bipolar recordings linking electrodes 1 to 2, 2 to 3, and 3 to 4. The second 4 channels are referential recordings linked to a surface electrode. Note the change in background rhythm to a rhythmic high-voltage theta at the onset of the seizure.

respond to medical treatment with zonisamide, lamotrigine, phenobarbital, diazepam, valproic acid, levetiracetam, phenytoin, clonazepam, lacosamide, oxcarbazepine, or topiramate.

Phase I evaluation was performed, and MRI showed a residual enhancing mass in the right cerebellum near the midline on the superior aspect of his prior surgery site (Fig. 3A and B). The patient underwent prolonged video-EEG monitoring with scalp electrodes. The interictal EEG study showed generalized spike and wave complexes, and he had multiple generalized myoclonic and myoclonic-tonic seizures with generalized epileptiform activity seen on EEG recordings. The decision was made to proceed to a Phase II evaluation with placement of depth electrodes into the cerebellar tumor.

Procedure. Frameless stereotactic MRI-guided placement of 2 posterior fossa depth electrodes was performed (Fig. 3C). A 6-contact depth electrode was placed in the superior right cerebellum in the lesion, and a 4-contact inferior cerebellar electrode was placed just inferior to the lesion.

Outcome. After placement of the depth electrodes, the patient underwent prolonged video-EEG monitoring performed with his intracranial electrodes and scalp electrodes. Interictal EEG recordings showed essentially continuous spikes seen from the superior cerebellar depth electrode. Occasionally these spikes were associated with a generalized burst of spike and slow wave complex or bifrontal epileptiform discharges seen on the simultaneous scalp EEG studies (Fig. 4A). Many of the cerebellar discharges were not associated with any activity on the scalp EEG recording. During ictal events, the EEG study showed bilateral spike and slow wave complexes recorded by the depth electrode. The changes were recorded by the depth electrode 20–50 msec before changes were recorded on the scalp EEG study (Fig. 4B).

After monitoring, the patient was taken back to surgery for tumor resection and depth electrode removal. Following resection, an intraoperative MRI study was

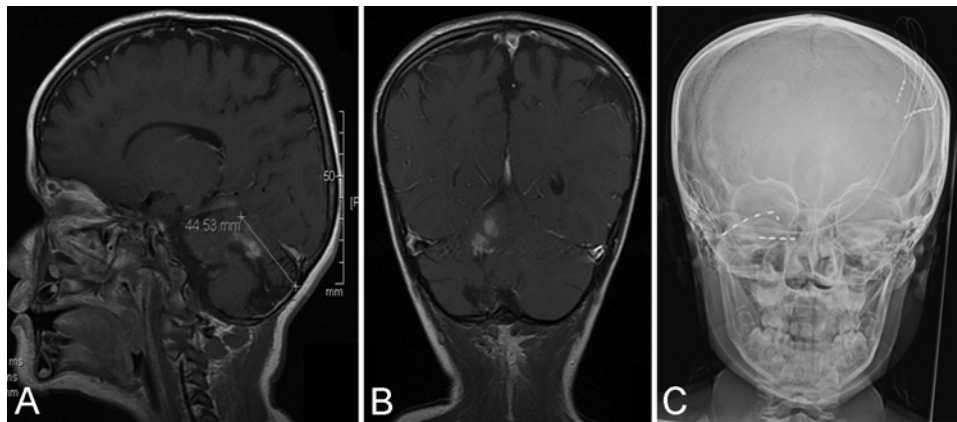


FIG. 3. Case 2. Sagittal (A) and coronal (B) T1-weighted Gd-enhanced images demonstrating residual partially enhancing tumor within the right cerebellar hemisphere and extending to the tentorial incisura. A postoperative anteroposterior skull x-ray (C) was obtained following Stage I surgery, demonstrating 2 depth electrodes in place, a 6-contact depth electrode at the superior aspect of the tumor, and a 4-contact depth electrode at the inferior aspect of the tumor.

performed, showing a gross-total resection. Postoperatively, the patient experienced 3 typical seizures the night of surgery, but was then seizure free. He was discharged home on lamotrigine. As an outpatient, he later required addition of ethosuximide. At this time, he has been seizure free for the 1.5 years since surgery, and his interictal scalp EEG findings are now normal. Follow-up MRI studies have shown no evidence of residual or recurrent tumor (Fig. 5).

Discussion

Subcortical structures are classically thought to have only ancillary effects on cerebral seizure activity. However, a number of cases found in the literature indicate otherwise (Table 1). Epileptic activity of cerebellar origin is typically associated with cerebellar dysplasia or glio-

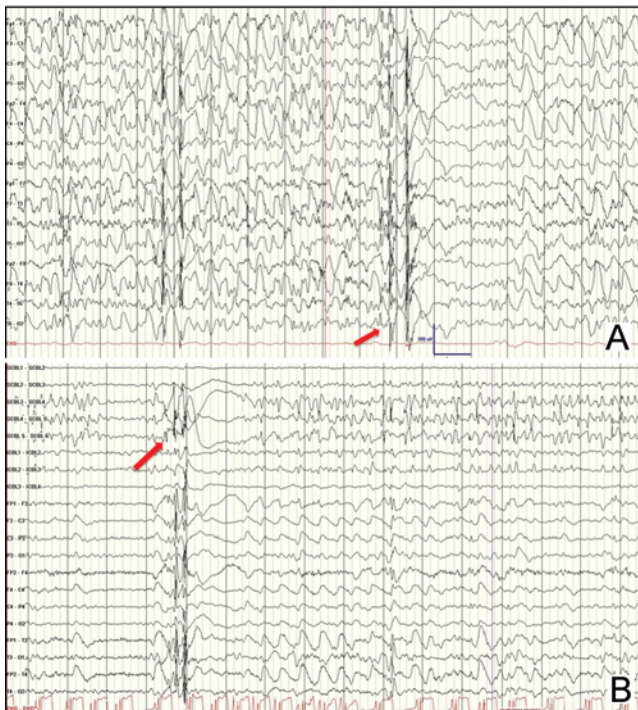


FIG. 4. Case 2. A: Scalp-recorded EEG study showing generalized epileptiform bursts and a generalized burst followed by generalized attenuation associated clinically with a myoclonic seizure (arrow) characterized by a brief nod of the head and vocalization. B: An EEG recording from depth and scalp electrodes capturing a similar myoclonic seizure. The burst clearly begins near the depth electrode (arrow) and spreads over 20 msec to the rest of the electrodes.

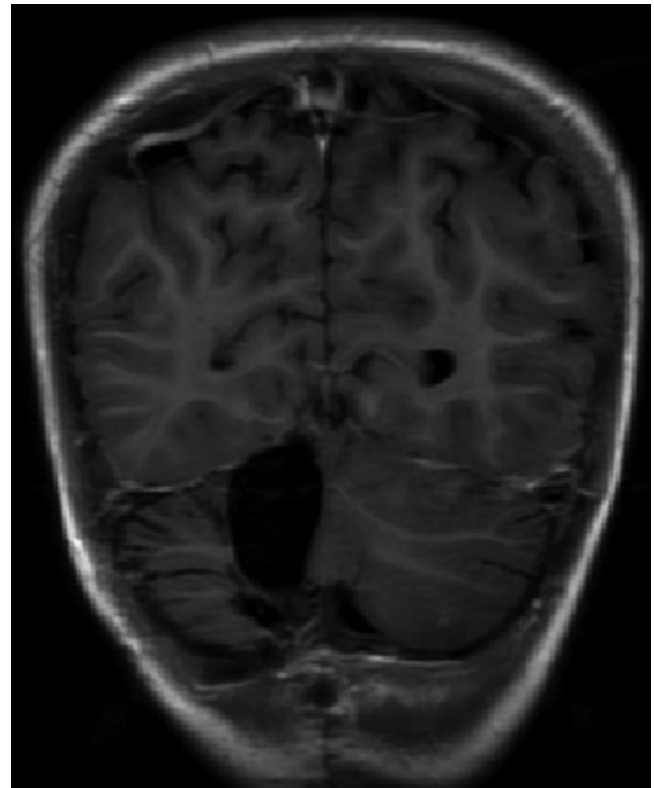


FIG. 5. Case 2. A T1-weighted nonenhanced coronal MRI study obtained 1 year after resection of the pilocytic astrocytoma, at a point in time when the child had remained seizure free—but while he still needed treatment with anticonvulsant medications (lamotrigine and ethosuximide)—demonstrating postsurgical changes within the resection cavity with no residual tumor.

Two cases of cerebellar seizures

TABLE 1: Literature review of cases published between 1976 and the present*

Authors & Year	Patient Age, Sex	Scalp EEG Results	Neuroimaging Findings	Pathology	Invasive Monitoring	Sz Control
Langston & Tharp, 1976	1.5 mos, M	abnormal	mass bulging into lt superior 4th vent	ganglioglioma	no	PR; szs resumed at lower frequency
Jayakar & Seshia, 1987	12 mos, M	normal	CT: mass at lt cerebellar hemisphere	low-grade astrocytoma	no	PR
Flüeler et al., 1990	1 wk, F	normal	MRI: mass at rt MCP & SCP	none	no	not controlled
	10 mos, F	ND	MRI: mass extending into rt MCP & ICP & compressing 4th vent	none	no	not controlled
Bills & Hanieh, 1991	3 wks, M	ND	CT/MRI: mass at lt SCP compressing 4th vent	ganglioglioma	no	PR, no szs 3 mos out
al-Shahwan et al., 1994	1 day, F	normal	MRI: mass at rt MCP & SCP	ganglioglioma	no	PR; szs resumed at lower frequency
Harvey et al., 1996	1 day, M	normal	MRI: mass at lt MCP & SCP effacing 4th vent	ganglioglioma	yes	CR, no szs w/o meds
Chae et al., 2001	4 mos, M	normal	lesion, lt SCP	ganglioglioma	yes	PR, then CR
Delande et al., 2001	36 mos, F	abnormal	lesion, floor 4th vent	hamartoma	yes	resection
	36 mos, F	normal	lesion, floor 4th vent	hamartoma	no	resection
Mesiwala et al., 2002	1 day, M	abnormal	MRI: mass at lt MCP & SCP displacing 4th vent	ganglioglioma	yes	PR, then CR
Strazzer et al., 2006	32 mos, F	abnormal	lesion, rt deep median-paramedian cerebellar peduncle	pilocytic astrocytoma	no	PR + AEDs, but szs persist
Kulkarni et al., 2007	48 mos, M	abnormal	lesion, rt cerebellum/4th vent	ganglioglioma	no	AEDs
Park et al., 2009	3 mos, M	ND	lesion, floor 4th vent w/ extended cerebellar peduncle & hemisphere	hamartoma	yes	PR, then CR
Hanai et al., 2010	7 mos, M	normal	MRI: lesion, lt MCP	ganglioglioma	no	CR
Yagyu et al., 2011	20 mos, M	abnormal	MRI: dysplastic neuronal lesion, floor 4th vent	gangliocytoma	yes	PR
present study	96 mos, M	abnormal	lesion, rt deep cerebellum	gliosis or LGG	yes	AEDs
	36 mos, M	abnormal	lesion, rt medial cerebellar hemisphere	pilocytic astrocytoma	yes	PR, then CR

* AEDs = antiepileptic drugs; CR = complete resection; ICP = inferior cerebellar peduncle; MCP = middle cerebellar peduncle; ND = not done; PR = partial resection; SCP = superior cerebellar peduncle; Sz = seizure; vent = ventricle.

neuronal tumors. It is usually seen in young children and is rarely controlled with antiepileptic drugs.

Our first case report presents a child with either gliosis or an LGG. The EEG recordings from the depth electrodes placed within the lesion showed that the tumor was the origin of his seizure activity. Concerns by the family that surgery presented the possibility of new neurological deficit made us pursue medical therapy rather than further surgery. This case is unique because it is the only case in the literature describing cerebellar seizures originating from a suspected LGG that is controlled by medical therapy without resection.

Most of the literature suggests that complete resection of cerebellar glioneuronal tumors is necessary for seizure control. There is 1 case of cerebellar seizures stemming from a low-grade astrocytoma that was reported in 1987 by Jayakar and Seshia.⁸ This case reported that seizures were controlled after only partial resection. In 2006, Strazzer et al.¹⁶ reported a case of a child who underwent resection of a pilocytic astrocytoma at 32 months of age for cerebellar

seizures. This resulted in reduction of seizures with periods of seizure control, although initiation of anticonvulsant therapy was necessary. Follow-up MRI studies in this patient revealed a small area of residual tumor.

Our second patient with pilocytic astrocytoma is an interesting demonstration of a focal lesion causing generalized epilepsy. During monitoring, the patient's scalp EEG recordings demonstrated generalized epileptiform discharges interictally, and also in association with the patient's typical seizures, which were generalized myoclonic seizures. Although scalp EEG changes were noted, the EEG recording from the depth electrodes clearly demonstrated that the lesion was the origin of the epileptic discharges. That, coupled with the fact that the seizure activity resolved after complete resection of the tumor, suggests that the lesion itself was the cause of the epileptic activity. Focal lesions causing generalized epilepsy syndromes and generalized EEG changes have been described previously. The original cases noted were infants with congenital focal lesions and infantile spasms associated with a hypsar-

rhythmic EEG background.^{9,17} More recently, older children and young adults with congenital or early acquired cortical lesions and generalized EEG findings have been shown to be surgical candidates in some instances.¹⁸

Our report is unusual in demonstrating that focal cerebellar lesions can also lead to generalized EEG abnormalities and even a seizure semiology, which appears to be generalized in nature. A similar case of myoclonic cerebellar seizures associated with a pilocytic astrocytoma has been reported, but depth electrode monitoring was not used to prove that the lesion was the origin of the seizure activity.¹⁶ The patient reported on by Strazzer et al. did have a reduction of seizures following subtotal tumor resection.

Conclusions

These case reports, in conjunction with the reviewed literature, lead to the conclusion that in patients with cerebellar structural lesions and seizures a causative relationship should be considered. We have demonstrated, as have others, that surface EEG recordings may not be diagnostic, and invasive monitoring is necessary to confirm that a cerebellar lesion is the origin of the epilepsy. Findings on PET or SPECT studies may also be supportive. Medications and partial resections are often ineffective in controlling the seizures associated with cerebellar lesions, whereas complete resection of the lesion is generally curative. As shown in our first case, medical therapy may be effective when surgical options could lead to further neurological deficit; however, this is distinctly uncommon.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: FA Boop, Wheless. Acquisition of data: S Boop. Drafting the article: S Boop, Van Poppel. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: FA Boop.

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