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Chronic isolated hemifacial spasm as a manifestation of epilepsy partialis continua

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Abstract

The objective of this case study was to describe the clinical and EEG/fMRI data of a case of isolated hemifacial spasms (HFS) due to epilepsy partialis continua in a 59-year-old man with abnormal hemifacial movements that disappeared during voluntary tasks, were absent during sleep, and responded to carbamazepine. His neurological examination was normal; EEG showed right inferior frontal epileptiform discharges. EEG-fMRI showed increased blood oxygenation level-dependent contrast in the right inferior and middle frontal gyri corresponding to the contralateral motor and premotor cortex responsible for facial movements (BA 44, 45, 45, 9) with widespread BOLD signal deactivations suggestive of epileptic network involvement despite a very focal epileptogenic process. We hypothesize that the response of some cases of HFS to carbamazepine, a first-line treatment in the pre-botulinum toxin era, may have been due to its anti-epileptic effects rather than to modulation of facial nerve hyperexcitability.

Keywords

hemifacial spasm; epilepsy partialis continua; EEG/fMRI

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Competing interests

JPS is a paid consultant for Compumedics USA, Ltd.

Note added in proof: A patient with almost continuous right hemifacial spasm, right hemiparesis, and dysarthria caused by a subacute hemorrhage in the left frontal cortex leading to epilepsy partialis continua has been recently reported. This example of secondary HFS further emphasizes the epileptic etiology of HFS (Mov Disord. 2007 Jul 26; [Epub ahead of print])

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Introduction

Epilepsia partialis continua (EPC) represents a cortical rather than brainstem or cranial nerve epiphenomenon that may present as continuous and irregular muscle jerks. In adults, the most common identifiable cause of EPC is cerebrovascular disease.(1) When it affects face, EPC may be mistaken for hemifacial spasm (HFS) which is a peripheral movement disorder. Isolated hemifacial spasms are characterized by involuntary, irregular, clonic, or tonic movements of muscles innervated by the facial nerve. HFS is believed to result from hyperexcitability of the facial nerve nucleus or ephaptic transmission within the proximal facial nerve segment of the facial nerve nucleus. Compression of the proximal part of the facial nerve by a vascular structure, such as an ectatic vessel or aneurysm, is the most commonly recognized cause.(2) As in the case of EPC, the diagnosis of HFS is clinical but its etiology is often elusive, even after multiple investigations. Similarly, despite focal EEG abnormalities, the etiology of EPC remains uncertain in about 20% of patients.

Few reports have addressed the epileptic phenotype of HFS. Common findings include a paroxysmal presentation, more generalized distribution of abnormal movements, and associated neurological deficits. These have been recognized in a patient with simple partial seizures of temporal lobe origin (3) and in three children with associated neurological deficits and structural lesions in the ponto-medullary junction or superior cerebellar peduncle.(4–6)

The objective of this report is to describe an adult case of chronic HFS without any associated neurological abnormalities, clinically similar to the segmental facial myoclonic movements of idiopathic HFS, but demonstrating an abnormal electrodiagnostic evaluation suggestive of EPC and to outline the anatomical and functional underpinnings of the irritative zone using combined EEG/fMRI.

Report of a case

A 59-year-old was evaluated for a six-month history of left eye blinking followed by upper face twitching and continuous facial discomfort without pain. He experienced a similar bout of left facial twitching 3 years prior to presentation, which spontaneously resolved after 3 months and for which he sought no medical attention. He had no prior illnesses except for mild depression. He denied any history of head trauma, meningitis/encephalitis or other precipitating events. The movements had tonic and fine myoclonic components that partially attenuated or disappeared during volitional tasks (Figure 1). The movements were absent during sleep. There were no other abnormalities on the neurological or general examination. Repeated brain MRIs with and without gadolinium (including a high-resolution 3-Tesla study) performed several months apart were normal. Standard outpatient EEG while the patient was experiencing clinical facial twitching identified frequent epileptiform discharges with right inferior frontal maximum. Carbamazepine provided almost complete resolution of the facial twitching although he temporarily discontinued it due to sleepiness with subsequent recurrence of the abnormal movements. The diagnosis of EPC was confirmed by the finding of irregular epileptiform discharges or rhythmic/semi-rhythmic focal slowing over the right inferior frontal head region during prolonged video/EEG monitoring while the patient was experiencing clinical symptoms (Figure 2A).

EEG/fMRI

The patient underwent 4 Tesla EEG/fMRI (61.5 cm bore Varian Unity INOVA system; Varian, Inc., Palo Alto, CA equipped with a standard head coil) after signing informed consent approved by the Institutional Review Board of the University of Cincinnati. The patient was fitted with MRI-compatible 64-channel EEG cap with braided carbon-fiber cabling and current-limiting in-line resistors (Compumedics USA, Ltd., El Paso, TX). Once electrode

placement according to the standard 10/20 system and low electrode impedance were confirmed (<10 kOhm), a fast localizer scan for head positioning and a T1-weighted MDEFT structural image ($T_{MD}=1.1$ s, $TR=13$ ms, $TE=6$ ms, $FOV=25.6 \times 19.2 \times 19.2$ cm, matrix $256 \times 192 \times 96$ pixels, flip angle=20 degrees) were obtained. This was followed by continuous acquisition of a 20 minutes long EEG sample (sampling rate 10 kHz) with simultaneous T2*-weighted spin-echo Echo Planar Imaging (EPI) pulse sequence ($TR/TE = 3000/45$ ms, $FOV = 25.6 \times 25.6$ cm, matrix 64×64 pixels, slice thickness = 4mm, flip angle = 90° ; NR = 400).

EEG data collection and processing was performed with the Scan 4.3.5 software (Compumedics USA, Ltd., El Paso, TX). After low-pass filtering with a cutoff frequency of 60 Hz, the EPI gradient artifacts induced by imaging were averaged over the first 3 TR periods precisely aligned using the embedded time marks. Average gradient signals were then subtracted from each epoch of the raw data following a method described by Allen et al.(7) In the Scan software, this subtraction procedure was further improved by optimizing temporal alignment between the average gradient waveform and the raw data based on the position of the peak of their cross-correlation. Ballistocardiographic artifact was removed in the same fashion using ECG events as time marks. The cleaned data were subsequently decimated to 200 Hz and derived in a standard, 16 channel bipolar montage.

For fMRI data analysis, two reference time course files were constructed. The first file contained designation whether particular brain volumes should be included or excluded from analysis due to excessive electrode artifact in the EEG data or due to movement artifact. Based on the above restriction, 92.5% (370/400) EPI whole brain volumes were included in the analysis. Second reference time course file contained the exact timing of the epileptiform discharges ($N = 40$). Cincinnati Children's Hospital Image Processing Software (CCHIPS[®]) was used for fMRI data analysis. The fMRI images were motion corrected and smoothed with a 4 mm Gaussian filter. Baseline drift correction was performed using a quadratic baseline correlation on a pixel-by-pixel basis. Maps of the t -statistic were created using the timing of each epileptiform discharge as an event in the fMRI analysis. At each voxel, event-related analysis was achieved by computing t -statistic values on a pixel-by-pixel basis from series data correlated with reference waveform with peak at 5 seconds.(8)

Results

Ictal EEGs obtained during prolonged video/EEG monitoring (Figure 2) and during the EEG/fMRI procedure (Figure 2B) showed rhythmic/semi-rhythmic intermittent or continuous focal right fronto-temporal slowing and frequent epileptiform discharges ($F8>T2$ maximum). Analysis of the EEG/fMRI data showed increased blood oxygenation level-dependent (BOLD) contrast in the right frontal cortex (Figure 2C; centroid's Talairach coordinates 25, 23, 11; gray and white matter of the inferior frontal gyrus; BA 44 and 45). The area of activation associated with the epileptiform discharges in the right frontal cortex was relatively large and also encompassed prefrontal cortex of the middle frontal gyrus (BA 46 and 9). Additional small areas of increased BOLD signal were also noted in the ipsilateral (centroid Talairach coordinates 47, -26, 7; superior temporal gyrus) and contralateral (centroid's Talairach coordinates -47, 5, 3; left insular cortex) hemispheres. Global deactivations (negative BOLD signal changes) were observed (Figure 2D).

Discussion

In this case report, we present the clinical, electrographic, and EEG/fMRI correlates of EPC originally interpreted as idiopathic isolated hemifacial spasms. This report emphasizes that isolated HFS may not only represent a peripheral movement disorder but also an epileptic phenomenon – EPC. The clinical features and investigations presented herein suggest that the

phenomenology of EPC-induced facial movements may not be easily distinguished from the focal and jerky spasms of isolated HFS. Furthermore, it raises the possibility that the success in the treatment with carbamazepine in a proportion of presumed idiopathic HFS patients, common before the advent of botulinum toxin, may be related to its antiepileptic properties rather than to the inhibition of posttetanic potentiation at the facial nerve nucleus.

The clinical phenotype in prior reports on HFS of epileptic origin had pointed to a *paroxysmal* phenomenon associated with ictal crying or oculomotor dysfunction.(3–6) The former resulted from simple partial seizures of left temporal lobe origin, propagating to the left cingulate region, and resolving after surgical treatment.(3) The latter was described in children with structural lesions in the pontomedullary junction,(4) superior cerebellar peduncle,(5,6) and cerebellar hemisphere.(6) Lacking other ictal deficits and imaging abnormalities, the epileptic nature of the HFS case reported here was initially elusive given their clinical disappearance during voluntary tasks and sleep, unlike what is reported to occur in focal seizures, where motor phenomena are typically not suppressed or substantially modified by volitional tasks. However, the persistence of the facial movements throughout the day ultimately raised the consideration of an epileptic phenomenon, which was suggested with a standard outpatient EEG. Appropriately suspected to have HFS, this patient had been referred to a movement disorders clinic for chemodenervation with botulinum toxin. However, such therapeutic strategy would not have addressed the underlying pathophysiologic mechanism, elucidated with EEG and further characterized with EEG/fMRI.

Concurrent EEG and fMRI has recently been used for non-invasive evaluation of lesional and non-lesional focal epilepsies.(9,10) In many patients, it identifies widespread areas of activation/deactivation with maximally increased BOLD signal in the putative area of seizure onset. The EEG/fMRI technique can evaluate widespread epileptogenic processes and networks involved in the generation and propagation of SWD and accurately localize the zone of ictal onset. Using this technique, we were able to visualize the area of EPC onset by correlating frequent epileptiform discharges with the BOLD signal changes in areas involved in the control of left facial movements. We also found widespread BOLD activations and deactivations: BOLD signal increases in left insula and right temporal lobe and widespread signal decreases (Figure 2D). This is suggestive of a possible widespread network involvement in the genesis of epileptiform discharges in EPC and in the epileptogenic process overall as has previously been noted in a study of network connectivity in patients with focal onset epilepsy.(11) Therefore, addition of EEG/fMRI to the evaluation of this patient not only established that the source of epileptiform discharges was co-localized with the area responsible for facial movements (motor and premotor cortex) but also confirmed the ability of this technique to evaluate the networks involved in seizure generation and propagation.

In summary, this report expands the clinical presentation of EPC and provides the first anatomical and functional imaging correlates for isolated ictal hemifacial movements. Although IHFS is overwhelmingly idiopathic or (cranial) neuropathic, our case should serve to alert clinicians regarding the potential cortical etiology. EEG should be considered in patients with isolated facial movements of unknown or unclear etiology.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

Right hemifacial involuntary contraction, beginning in the left orbicularis oculi (A) and spreading to the ipsilateral hemiface (C), was attenuated with voluntary tasks such as smiling (D) and tongue protrusion (E). Intermittent presence of voluntary right eyelid closure throughout, particularly preceding full hemifacial spread (B), was the result of discomfort caused by the abnormal movements. Complete resolution was documented 2 months after the initiation of carbamazepine (F), once a target dose of 1,400 mg/day had been achieved.

Figure 2.

EEG/fMRI findings. A. Ictal EEG showed semirhythmic or rhythmic right fronto-temporal epileptiform discharges (F8/T2 maximum) and/or semirhythmic or rhythmic right fronto-temporal theta/delta slowing. B. EEG (bipolar montage) after removal of EPI and ballistocardiographic artifacts. Findings similar to A were noted. C. Increases in BOLD signal associated with epileptiform discharges superimposed on T1 anatomical image (images are in radiological convention – right on the picture is left in the brain). Only activations with $t \geq 3.1$ are shown. D. Similar as in C but both, fMRI activations and deactivations are shown.