Epilepsia, 51(10):2108–2115, 2010
doi: 10.1111/j.1528-1167.2010.02662.x

FULL-LENGTH ORIGINAL RESEARCH

Bilateral akinetic seizures: A clinical and electroencephalographic description

***Rafael Toledano, *†Irene García-Morales, ‡Mónica M. Kurtis, §Ángel Pérez-Sempere, ¶Roberto Ciordia, and *Antonio Gil-Nagel**

***Epilepsy Unit, Department of Neurology, Hospital Ruber Internacional, Madrid, Spain; †Epilepsy Unit, Department of Neurology, Hospital Clínico San Carlos, Madrid, Spain; ‡Movement Disorders Unit, Department of Neurology, Hospital Ruber Internacional, Madrid, Spain; §Department of Neurology, Hospital General de Alicante, Alicante, Spain; and ¶Department of Neurology, Hospital de Cruces, Bilbao, Spain**

Summary

Purpose: The main feature of akinetic seizures is the inhibition of voluntary movements without impairment of awareness. Most clinical information about akinetic seizures has been obtained from cortical electrical stimulation studies, whereas clinical and video–electroencephalography (EEG) features have not been described thoroughly. We aimed to analyze clinical and EEG characteristics of bilateral akinetic seizures (BAS).

Methods: Patients with BAS were retrospectively identified from 1,858 consecutive video-EEG studies. All patients had ictal video-EEG, comprehensive clinical evaluation, neuropsychological testing, and brain magnetic resonance imaging (MRI).

Results: Ten patients (nine men) were identified; mean age was 22.5 years (range 0.3–71 years) at the time of epilepsy onset and 34.9 years (range 5–73 years) at the time of evaluation. BAS was the only seizure type in four patients. BAS consisted of sudden speech and motor arrest in eight patients, whereas in two patients seizures were characterized by abrupt freezing precipitated by gait initiation. Startle precipitated BAS in four patients. Magnetic resonance imaging (MRI) showed mesial frontal lobe lesions in six patients. Epileptiform activity was restricted to the frontal midline electrodes in all patients, with variable extension to frontal regions. In five patients, BAS were initially misdiagnosed as generalized seizures or nonepileptic events.

Discussion: BAS should be considered in the differential diagnosis of patients reporting paroxysmal inability to move with preservation of awareness, bearing in mind that these seizures can occur spontaneously or be precipitated by startle. The diagnosis can be achieved with video-EEG monitoring, showing stereotyped semiology and distinctive EEG abnormalities, and is often supported by the presence of lesions involving the frontal lobes.

KEY WORDS: Akinetic seizures, Epilepsy, Seizure semiology, Akinesia, EEG.

Human brain electrocortical mapping has shown negative motor areas, where stimulation results in inhibition of voluntary movements with preserved postural tone and consciousness (Penfield & Welch, 1949, 1951; Lim et al., 1994; Lüders et al., 1995). With this technique, once cortical stimulation has ceased, patients typically report being aware of having experienced focal or bilateral inhibition of voluntary movements. Based on these observations, the name ''akinetic seizures'' has been suggested for seizures characterized by inhibition of voluntary movements without impairment of awareness (Lüders et al., 1998). Although muscle tone can be lost during these seizures, akinesia is the most important feature. However, clinical and electroencephalographic descriptions of this seizure type are scarce, since most data come from cortical electrical stimulation studies. The aim of this study was to describe the electrographic and clinical features of seizures characterized by inhibition of bilateral voluntary motor activity with preservation of awareness and muscle strength and tone.

Accepted May 13, 2010; Early View publication July 16, 2010.
Address correspondence to Antonio Gil-Nagel, Department of Neurology, Hospital Ruber Internacional, La Masó 38, 28034 Madrid, Spain.
E-mail: agnagel@ruberinternacional.es

Methods

Patient selection

In line with previous reports, we chose the term ''akinetic seizures'' (Lim et al., 1994; Lüders et al., 1998) as the name that best describes ictal events resulting in akinesia, defined as the impossibility to initiate a movement or maintain a movement that has already started without loss of consciousness. When selecting this name, we wanted to distinguish these types of seizures from those defined by the terms ''ictal paralysis'' or ''focal akinetic seizures,'' referring to seizures where movement impairment is produced

by weakness, most often affecting distal muscles (Noachtar & Lüders, 1999; Iriarte et al., 2002; Matsumoto et al., 2005). To avoid confusion with the aforementioned conditions, we added the term ''bilateral'' and defined ''bilateral akinetic seizures'' (BAS) as having the following clinical characteristics: (1) bilateral and axial motor activity inhibition; (2) preservation of awareness and recollection of the event; (3) absence of abnormal posturing, paresis, loss of tone, or emotional reactions such as fear or terror, that could explain motor arrest; (4) abrupt onset and ending. We retrospectively identified 15 patients with BAS meeting these strict clinical criteria in our database of 1,858 consecutive patients evaluated with video–electroencephalography (EEG) from 1998 to 2008. Ten of them had ictal video-EEG recordings and were thus included in this study. Video-EEG was performed for differential diagnosis or seizure characterization in all patients. Patients with more than one seizure type were included as long as they were able to differentiate BAS from the others. To complete video-EEG information with reliable descriptions from patients (i.e., to verify that awareness was not impaired, and ask the patients to explain why they could not move), we excluded those in whom BAS always evolved into complex partial (CP) or secondary generalized tonic–clonic seizures (SGTC), as well as those with severe psychomotor delay. Clinical information was obtained from past medical records and a systematic interview with patients and seizure witnesses. To complete or confirm clinical information after the study was started, when necessary, patients and witnesses were interviewed again by phone or during follow-up clinic visits.

EEG and video recordings

Seizure semiology

Semiology of seizures was assessed by interrogating the patient and witnesses, and by postprocessed video analysis. Trained nurses or an epileptologist assessed patients' responses to commands and orientation, both during and after their seizures, in order to evaluate ictal awareness and the postictal state. The presence and aura characteristics were determined by history and patient's description during video-EEG monitoring.

EEG recordings

Long-term video and surface EEG recordings were performed with either a Nicolet-BMSI 5000 (Nicolet-BMSI Inc., Madison, WI, U.S.A.) or a XLTEK NeuroWorks system (Natus medical Inc., San Carlos, CA, U.S.A.). EEG was recorded from 23 scalp sites, placed according to the International 10–20 System, and always including electrodes at the midline positions (Fpz, Fz, Cz, Pz, and Oz). The bandwidth during EEG acquisition was 0.3–70 Hz. Ictal EEGs were evaluated for onset, lateralization, distribution, progression, and duration, according to patterns and labels described previously (Foldvary et al., 2001). Midline spikes were defined as focal epileptiform discharges localized to, or of highest amplitude at, one of the midline electrodes (Fpz, Fz, Cz, Pz or Oz). Semiologic phenomena captured on video were simultaneously recorded with EEG in a split-screen.

Brain imaging

High-resolution brain magnetic resonance imaging (MRI) was performed in all patients. Imaging studies always included volume acquisition, T_1- and T_2-weighted, and fluid-attenuated inversion recovery (FLAIR) sequences. In addition, positron emission tomography (PET) or ictal/interictal single-photon emission computed tomography (SPECT) was done in four patients.

Neuropsychological testing

Neuropsychological testing included full-scale intelligence quotient (FSIQ) and standard cognitive tests for evaluation of speech (Boston Naming Test), verbal memory [Hopkins verbal learning test and Wechsler Adult Intelligence Scale (WAIS) and Wechsler Intelligent Scale for Children (WISC)], nonverbal memory (Rey figure and Benton test to shape visual screening), visual-perceptive function (Hooper and Benton test), and executive function (Trail Making A and B, and Stroop test).

Results

Study population

Nine of the 10 patients were male; mean age was 34.9 years (range 5–73 years) at the time of evaluation and mean age at epilepsy onset was 22.5 years (range 0.3–71 years). Patients were followed for a mean of 3.3 years (range 1.3–8 years). BAS were first documented at mean age 29.2 years (range 3–71 years). In four patients, BAS were the only seizure type; the remaining patients also had CP and SGTC seizures. During video-EEG monitoring, one patient had two isolated BAS and BAS evolving to a generalized tonic–clonic (GTC) seizure. The remaining patients only had BAS during video-EEG. Antiepileptic drugs controlled CP and SGTC seizures when present. BAS were completely controlled with antiepileptic drugs in four patients; the remaining six also benefited from treatment, showing marked reduction in seizure duration, frequency, and severity. In five patients, ictal manifestations were initially misdiagnosed as anxiety, psychogenic nonepileptic events, clumsiness, or idiopathic generalized epilepsy, with a mean diagnosis delay of 4.2 years (range 2–5 years). Misdiagnosis was more common in patients without previous history of CP and SGTC seizures. None had a family history of epilepsy in first- or second-degree relatives. Neurologic examination was normal in all but one patient (Patient 9), who had moderate cognitive delay and mild ataxia. Two patients (Patients 5 and 6) showed slight executive dysfunction characterized by slowness of information processing

Table 1. Patient characteristics

Pt/sex/age[a] (years)	No. seizures recorded	Age at epilepsy onset	Age at diagnosis of BAS	Precipitating factors	Ictal semiology	Seizure duration[b]	BAS response to medication
1/M/21	8	7	19	None	Speech and motor arrest. Bilateral arm myoclonus.	<20 s	SF
2/M/5	7	1.6	4	None	Speech and motor arrest.	<20 s	PR
3/M/18	1	9	9	None	Initial motor hesitation. Speech and motor arrest.	<15 s	SF
4/M/39	2	29	34	Auditory startle	Speech and motor arrest.	<15 s	PR
5/M/23	8	10	10	Performing multiple tasks simultaneously.	Initial motor hesitation. Speech and motor arrest. Bilateral arm myoclonus.	<15 s	PR
6/M/36	1	28	29	Auditory startle. Performing multiple tasks simultaneously.	Speech and motor arrest.	70 s	SF
7/M/50	3	46	46	None	Speech and motor arrest. Bilateral arm myoclonus.	4 m	PR
8/M/73	7	71	71	None	Speech and motor arrest. Bilateral arm myoclonus.	20 m	SF
9/F/55	28	0.3	47	Walking. Auditory or somatosensory startle while walking, but not while sitting.	Sudden gait freezing with: Initiation of gait, turning, avoiding obstacles on terrain.	<10 s	PR
10/M/29	18	24	29	Walking. Auditory or somatosensory startle while walking, but not while sitting.	Sudden gait freezing with: Initiation of gait, turning, avoiding obstacles on terrain.	<10 s	PR

[a]Age patients were first evaluated in our video-EEG monitoring unit.
[b]Seizure duration was classified according to the longest seizure recorded.
s, seconds, m, minutes; BAS, bilateral akinetic seizures; SF, seizure free; PR, partial response (>50% reduction in frequency and marked decrease in seizure duration).

and motor execution speed (identified by WAIS subtests, and Trail Making and Stroop tests) (Table 1).

Clinical characteristics

Based on ictal semiology, two groups of patients could be differentiated. In the first group (Patients 1–8) seizures consisted of sudden arrest of speech and motor activity. During the event, they were unable to move, speak, or follow commands. Patients appeared completely motionless or frozen. At the end of their seizures, they returned to their previous activity without postictal motor, verbal, or cognitive deficits. Motor hesitation at seizure onset was observed in two patients (Patients 3 and 5). Subtle symmetric or asymmetric myoclonus of the upper limbs was an additional ictal feature in four patients (Patients 1, 5, 7, and 8), only evident with arms extended up front (i.e., captured on video-EEG when ictal activity interrupted reading a newspaper or a book). Startle or tasks requiring a higher level of attention, such as sequential actions or simultaneous motor tasks (i.e., writing a note while talking on the telephone or using a fork and a knife during a meal), were seizure-precipitating factors in three patients (Patients 4, 5, and 6). None of the patients in this group had seizures induced by gait or standing (Table 1).

In the second group (Patients 9 and 10), BAS consisted of abrupt gait interruption, precipitated by standing up, gait initiation, turning, and stumbling. Unexpected auditory or tactile startle occurring while walking could also precipitate their seizures. Onset was abrupt, not preceded by hesitation or a shortened stride. Impairment of stance was accompanied by clumsiness and efforts to maintain balance, with restricted voluntary arm swing and trunk swaying. During baseline examination in the supine position, leg motor function and ability to perform stepping and pedaling movements were preserved and did not trigger seizures. Both patients could talk throughout their seizures and described that their feet ''were glued to the ground.''

In both groups, most seizures were short, lasting 20 s or less. In addition to frequent brief episodes, two patients (Patients 7 and 8) had prolonged seizures and recurrent episodes of status epilepticus. Throughout these seizures, both patients had their eyes open and were unable to speak or move, with fluctuating severity, showing intermittent myoclonus. In four patients, seizures aggregated in flurries. Three patients reported nonlocalizing vague auras. Most patients described their seizures as ''freezing'' or ''inability to move.'' We did not observe other clinical features of mesial frontal lobe epilepsy, such as bilateral tonic posturing, screaming, emotional behavior, or bizarre movements.

Neuroimaging characteristics

MRI disclosed epileptogenic lesions in six patients (Fig. 1). Three patients had focal lesions (two cortical

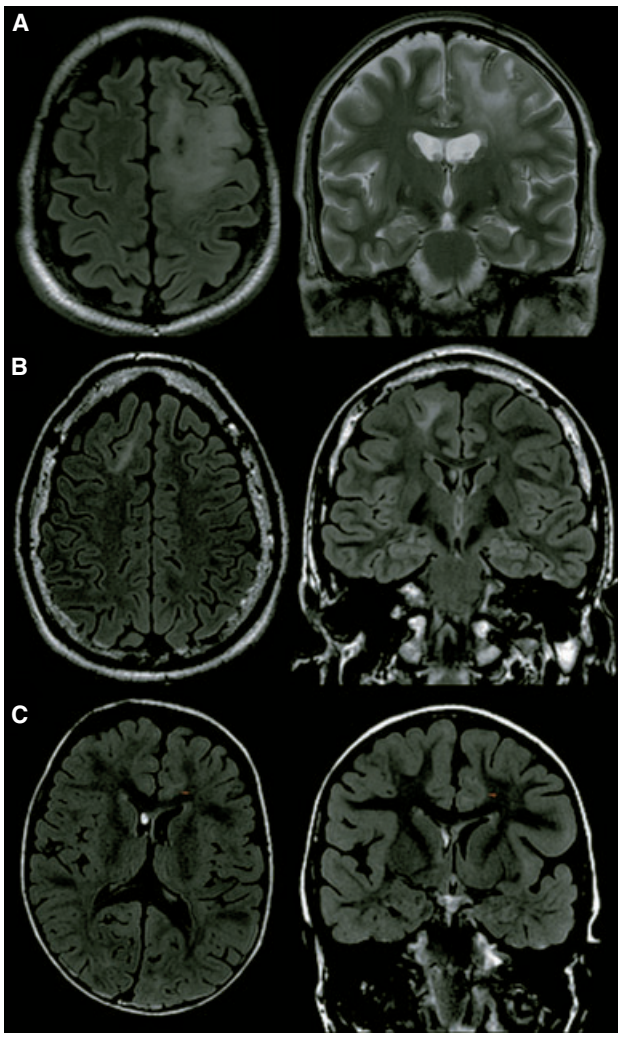

Figure 1.
Representative MR images of different lesions involving the frontal superior gyrus and mesial aspect of the frontal lobe. Axial FLAIR and coronal T_2-weighted sections in Patient 6 show a low grade astrocytoma (**A**). Axial and coronal FLAIR sections in Patient 3 show a hyperintense circumscribed area consistent with focal dysplasia (**B**). Axial and coronal FLAIR sections in Patient 2 show an area of hyperintensity that extends from the corpus callosum to the left mesial frontal lobe. The blurring of the gray–white matter interface and increased gyrus thickness are consistent with focal cortical dysplasia (**C**).

dysplasia, Patients 2 and 3; one low grade astrocytoma, Patient 1) involving the mesial aspect of the frontal lobe. Two patients had bilateral frontal ischemic lesions involving white matter and the gray–white matter junction (Patients 7 and 8). One patient with a history of perinatal asphyxia had bilateral leukoencephalopathy affecting the mesial aspect of the frontal and parietal lobes (Patient 9). In two patients, functional neuroimaging (ictal SPECT and PET) revealed abnormalities in the frontal lobe (Patients 1 and 2) (Table 2).

Electroencephalography

Interictal EEG was abnormal in seven patients, showing epileptiform activity over frontal midline electrodes (Fz, Cz, or both), with variable extension to frontal regions (Fp1, Fp2, F3, or F4). In one case (Patient 4), interictal spikes were also present beyond frontal regions. Eighty-three seizures were recorded in the 10 patients available for analysis (mean 8.3 seizures per patient; range 1–28). Ictal activity was recorded in all patients and always involved frontal midline electrodes (Fz, Cz or both) at onset, remaining localized in four patients (Patients 4, 8, 9, and 10), and propagating to adjacent frontal electrodes in the other six. Ictal patterns included repetitive spikes or spike–wave discharges, and low-amplitude high-frequency activity (alpha or beta) (Fig. 2). In patients with isolated involvement of midline electrodes, ictal patterns could be overlooked if Fz and Cz were not included in a bipolar transverse montage (Fig. 2E) (Table 2).

Discussion

This study contributes to the existing data about akinetic seizures, a seizure type that was not included in the classification of the International League Against Epilepsy (ILAE) but described in the semiologic seizure classification by Lüders et al. (Commission on Classification and Terminology of the International League Against Epilepsy, 1989; Lüders et al., 1998; Engel, 2001). Our patients comply with the proposed definition of akinetic seizures, since they show epileptic events characterized by inhibition of voluntary movements with preservation of consciousness. However, their seizure semiology poses a nomenclature challenge, since motor arrest in akinetic seizures, as originally described by Lüders et al. (1998), could be possibly accompanied with paresis or loss of muscle tone. We believe that akinetic seizures involving weakness and tone deficits are better described by the term ''ictal paralysis'' (Abou-Khalil et al., 1995; Iriarte et al., 2002; Matsumoto et al., 2005), although others have also used the term ''focal akinetic seizures'' (Noachtar & Lüders, 1999). We excluded these features in our study in order to focus on akinetic seizures whose main characteristics were (1) inability to initiate or maintain a movement, (2) axial and bilateral involvement, (3) preservation of consciousness, and (4) preservation of muscle strength and tone, and have called them ''bilateral akinetic seizures.''

This seizure type may be included in the group of ''inhibitory motor seizures'' within focal seizures, as proposed by the ILAE Commission Report of 2001 (Engel, 2001), as well as in the Report of the ILAE Classification Core Group (Engel, 2006). On the other hand, seizure semiology in our patients is very similar to that of those patients recently

Table 2. Neuroimaging and EEG findings

				Ictal EEG	
Pt/MRI description[a]	Extension	Functional imaging	Interictal EEG	Onset pattern	Evolution
1/Normal	–	Frontal mesial hyperperfusion (SPECT)	BFM SW FM spikes	BFM fast activity	BFM regular SW
2/Left frontal dysplasia	Corpus callosum, cingulate gyrus and frontal superior gyrus	Left frontal hypometabolism (PET)	BFM pSW/SW	BFM fast activity	BFM fast activity
3/Right frontal dysplasia	Mesial and polar region	Not done	Normal	FM fast activity	Right FM irregular theta activity
4/Normal	–	Not done	LT spikes LC spikes FM spikes	FM repetitive spikes	FM repetitive spikes
5/Normal	–	Not done	BFM pSW/SW	BFM fast activity	(1) Generalized fast activity (2) BFM fast activity
6/Left frontal astrocytoma	Mesial and lateral region	Not done	FM spikes	FM repetitive spikes	Left FM repetitive spikes
7/Multiple ischemic lesions	Subcortical lesions in the frontal lobe at the gray–white matter junction	Normal (PET)	BFM SW	BFM repetitive spikes	Left FM repetitive spikes
8/Multiple ischemic lesions	Subcortical lesions in the frontal lobe at the gray–white matter junction	Not done	Normal	FM repetitive spikes	FM repetitive spikes
9/Extensive leukoencephalopathy	Mesial aspect of both frontal and parietal lobes	Not done	FM spikes	FM repetitive spikes	FM repetitive spikes
10/Normal	–	Normal (SPECT)	Normal	FM fast activity	FM irregular theta activity

[a]Nature of lesions was presumed according to radiologic features and medical history.

Pathologic confirmation was achieved in Patient 6. SW, spike and slow wave; pSW, polyspike and slow wave; FM, frontal mesial (maximal electronegativity at electrodes Fz, Cz or Fz/CZ); BFM, bilateral frontal mesial (maximal electronegativity at frontal mesial electrodes and Fp1/Fp2, F3/F4 or both); LT, left temporal; LC, left central.

reported by Ikeda et al. (2009). These authors argued that the term ''negative motor seizures'' might be more appropriate to refer to this seizure type, since ictal semiology in their patients resembles responses elicited by supplementary negative motor area (SNMA) cortical stimulation (Lim et al., 1994; Ikeda et al., 2009). Being aware of their arguments, we have used the term ''bilateral akinetic seizures'' because: (1) this term indicates akinesia as the main clinical feature; (2) lack of movement is not caused by weakness; (3) movement impairment affects trunk and extremities bilaterally; (4) the term akinesia is more descriptive than negative motor phenomena; and (5) the term akinetic seizures has already been coined and defined in the literature (Lüders et al., 1998).

The small number of subjects we identified and the paucity of descriptions in the literature suggest that BAS are either uncommon or difficult to recognize. Regarding the latter, ictal motor arrest is frequently reported in frontal lobe epilepsy (Manford et al., 1996; Fogarasi et al., 2001), suggesting that it is not a rare phenomenon. However, it is unclear whether patients from previous reports had impairment of consciousness during their seizures. Similarly, isolated ictal speech arrest has been described previously in lesions involving the premotor area of the superior frontal gyrus (Chee et al., 1997; Wieshmann et al., 1997), but it is unclear whether motor tasks were examined during seizures and, therefore, the possibility of widespread motor inhibition remains questionable.

Our study suggests that a diagnosis of BAS can be overlooked (epileptic events were not considered for several years in some patients), probably because of little knowledge about this seizure type among physicians. The importance of EEG recording, including midline electrodes, is highlighted, as ictal EEG activity can be missed if these electrodes are not included or not linked in appropriate transverse montages. Workup should pay particular attention to the differential diagnosis with paroxysmal kinesogenic dyskinesia. This entity, considered to be at the boundaries between epilepsy and movement disorders, is characterized by brief episodes of dystonia, chorea, or ballism triggered by sudden movement (Fahn, 1994). In some of our patients, a history of intermittent and paroxysmal episodes, along with precipitating factors and a negative family history, may suggest sporadic kinesogenic dyskinesia. However, other clinical features are more consistent with

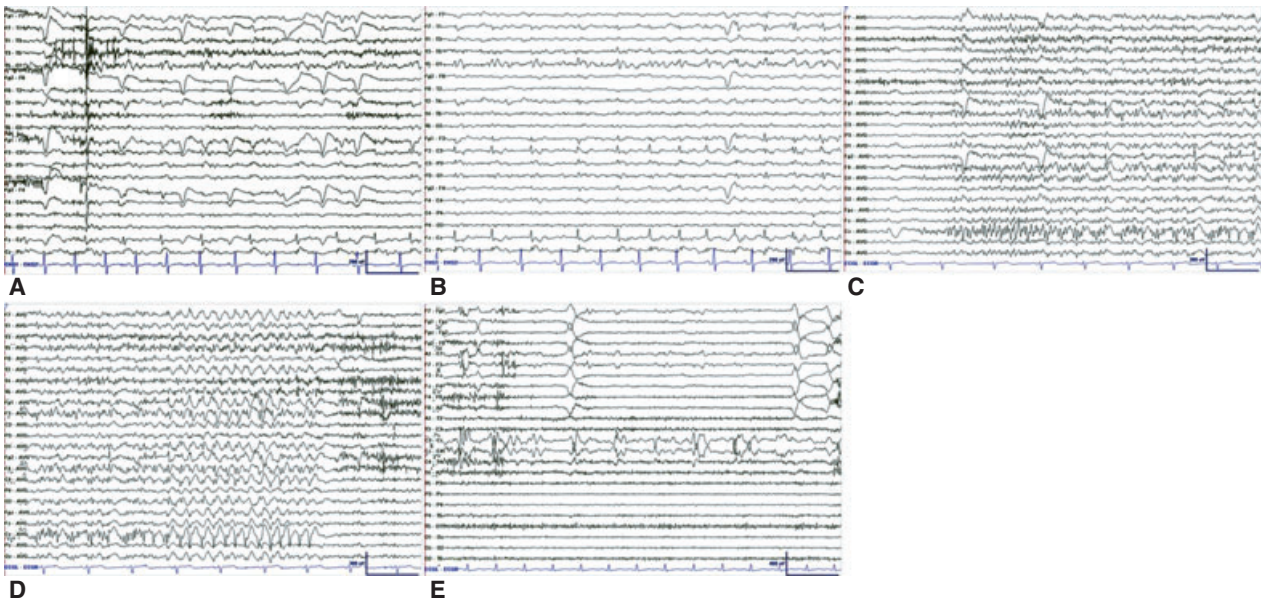

Figure 2.
Representative examples of ictal onset patterns. In Patient 6, ictal onset showed focal repetitive spikes localized in Fz–Cz (**A**), propagating to left frontal central electrodes (F3–C3) (**B**). Patient 1 had paroxysmal fast activity localized in Cz (**C**), evolving into spikes and slow waves over both frontal regions (F3 and F4) (**D**). Patient 9 showed localized Cz spikes, with minimal involvement of F7 (**E**), a pattern that could be easily missed if midline electrodes are not linked in a coronal montage.

focal epilepsy, especially the absence of hyperkinetic movements, its frequent association with CP and SGTC seizures, the presence of ictal epileptiform activity on EEG, and abnormalities involving the cortex while sparing the basal ganglia on brain imaging.

Surface EEG, brain imaging, and electrostimulation studies as well as seizure semiology have localized the symptomatogenic area of BAS to the frontal anterior mesial cortex. In line with the four previously reported cases (Meletti et al., 2003; Ikeda et al., 2009), surface EEG recordings provided excellent localization to the central and anterior midline in all our cases; six patients had structural lesions and one had abnormal functional neuroimaging involving the mesial aspect of the frontal lobe. However, in contrast with other mesial frontal epilepsies, characterized by nonlocalizing slowing or attenuation ictal patterns, we found well-localized anterior mesial discharges (Morris et al., 1988; Salanova et al., 1995; Bautista et al., 1998; Foldvary et al., 2001). These electrical differences may be due to a more superficial ictal dipole, easily detected by midline electrodes. Clinically, BAS also differs from other mesial frontal epilepsies, since the predominant feature is akinesia, occasionally accompanied by minor motor manifestations (subtle bilateral myoclonus and motor hesitation) versus hypermotor behaviors or bilateral asymmetric posturing typically associated with other mesial frontal epilepsies (Morris et al., 1988; Salanova et al., 1995).

The role of the rostral mesial frontal lobe in movement planning, programming, and organization has been demonstrated by lesion studies (Della Sala et al., 2002), human functional brain-imaging and electrical stimulation studies (Lim et al.,1994; Cunnington et al., 2002; Rushworth et al., 2002; Sahyoun et al., 2004), as well as direct neuron recording in primates (Tanji & Shima, 1994; Matsuzaka & Tanji, 1996; Nakamura et al., 1998; Shima & Tanji, 1998; Fujii et al., 2002; Isoda & Hikosaka, 2007). The SNMA may play a crucial role in complex motor movements such us action ignition, action plan changes, or execution of motor sequences (Matsuzaka & Tanji, 1996; Nakamura et al., 1998; Shima & Tanji, 1998; Fujii et al., 2002). Activation may enable a switch in motor tasks, by suppressing an automatic, inappropriate action and facilitating a controlled, desirable action (Isoda & Hikosaka, 2007). In addition, SNMA electrical stimulation can result in bilateral inhibition of movement, sudden arrest of ongoing motor activity, and interruption of speech, thus reproducing ictal behavior characteristic of BAS (Lim et al., 1994; Lüders et al., 1995). In some of our patients, seizures were triggered by sequential or simultaneous motor tasks, gait initiation, or gait program changes and startle, further supporting this localization as the area of seizure onset. Furthermore, electrical stimulation of the supplementary motor cortex, an area adjacent and posterior to the SNMA, can elicit negative myoclonus, as seen in four of our

patients (Rubboli et al., 2006). Finally, frontal mesial lesions can produce gait initiation and execution deficits, similar to those observed in two of our patients (Della Sala et al., 2002).

Our clinical and pathophysiologic findings are limited by the small patient sample as well as the lack of confirming intracranial recordings and response to epilepsy surgery. Nevertheless, accurate and prolonged video-EEG monitoring concordant with neuroimaging is considered to reduce the risk of biases in cases not undergoing surgery (Kellinghaus & Lüders, 2004). In addition, our localization hypothesis is supported by striking clinical similarities between ictal behavior in our series and data obtained from neurophysiologic and functional imaging studies, as well as animal studies with direct neuronal recordings of the rostral mesial area.

In conclusion, our study not only improves our understanding of the pathophysiologic mechanisms and localization underlying BAS, but also stimulates physician awareness, accurate diagnosis, and thus appropriate treatment of this entity. BAS should be considered in the differential diagnosis of patients reporting paroxysmal inability to move or episodic motor arrest. Diagnostic workup should include video-EEG monitoring with midline scalp electrodes, and high-resolution neuroimaging, as lesions involving the mesial frontal lobes may be found. Ictal semiology and EEG activity, as well as structural abnormalities when present, localize the symptomatogenic area to the mesial frontal lobes, an area that plays an important role in movement programming as demonstrated by human and animal studies.

Disclosure

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

References

Abou-Khalil B, Fakhoury T, Jennings M, Moots P, Warner J, Kessler RM. (1995) Inhibitory motor seizures: correlation with centroparietal structural and functional abnormalities. *Acta Neurol Scand* 91:103–108.

Bautista R, Spencer D, Spencer S. (1998) EEG findings in frontal lobe epilepsies. *Neurology* 50:1765–1771.

Chee MWL, So NK, Dinner DS. (1997) Speech and the dominant superior frontal gyrus: correlation of ictal symptoms, EEG, and results of surgical resection. *J Clin Neurophysiol* 14:226–229.

Commission on Classification and Terminology of the International League Against Epilepsy. (1989) Proposal for revised classification of epilepsies and epileptic syndromes. *Epilepsia* 30:389–399.

Cunnington R, Windischberger C, Deecke L, Moser E. (2002) The preparation and execution of selfinitiated and externally-triggered movement: a study of event-related fMRI. *Neuroimage* 15:373–385.

Della Sala S, Francescani A, Spinnler H. (2002) Gait apraxia after bilateral supplementary motor area lesion. *J Neurol Neurosurg Psychiatry* 72:77–85.

Engel JR. (2001) A proposed diagnostic scheme for people with epileptic seizures and with epilepsy: report of the ILAE Task Force on Classification and Terminology. *Epilepsia* 42:796–803.

Engel J Jr. (2006) Report of the ILAE Classification Core Group. *Epilepsia* 47:1558–1568.

Fahn S. (1994) Paroxysmal dyskinesias. In Marsden CD, Fahn S (Eds) *Movement disorders*, vol 3. Butterworth–Heinemann, Oxford, UK, pp. 310–345.

Fogarasi A, Janszky J, Faveret E, Pieper T, Tuxhorn I. (2001) A detailed analysis of frontal lobe seizure semiology in children younger than 7 years. *Epilepsia* 42:80–85.

Foldvary N, Klem G, Hammel J, Bingaman W, Najm I, Lüders H. (2001) The localizing value of ictal EEG in focal epilepsy. *Neurology* 57:2022–2028.

Fujii N, Mushiake H, Tangi J. (2002) Distribution of eye- and arm-movement-related neuronal activity in the SEF and in the SMA and pre-SMA of monkeys. *J Neurophysiol* 87:2158–2166.

Ikeda A, Hirasawa KI, Kinoshita M, Hitomi T, Matsumoto R, Mitsueda T, Taki J, Inouch M, Mikuni N, Hori T, Fukuyama H, Hashimoto N, Shibasaki H, Takahashi R. (2009) Negative motor seizure arising from the negative motor area: is it ictal apraxia? *Epilepsia* 50:2072–2084.

Iriarte J, Urrestarazu E, Artieda J, Alegre M, Schlumberger E, Lázaro D, Viteri C. (2002) Ictal paralysis mimicking Todd's phenomenon. *Neurology* 59:464–465.

Isoda M, Hikosaka O. (2007) Switching from automatic to controlled action by monkey medial frontal cortex. *Nat Neurosci* 10:240–248.

Kellinghaus C, Lüders HO. (2004) Frontal lobe epilepsy. *Epileptic Disord* 6:223–239.

Lim SH, Dinner DS, Pillay PK, Lüders H, Morris HH, Klem G, Wyllie E, Awad IA. (1994) Functional anatomy of the human supplementary sensorimotor area: results of extraoperative electrical stimulation. *Electroencephalogr Clin Neurophysiol* 91:179–193.

Lüders HO, Dinner DS, Morris HH, Wyllie E, Comair YG. (1995) Cortical electric stimulation in humans: the negative motor areas. In Fahn S, Hallett M, Lüders HO (Eds) *Negative motor phenomena. Advances in Neurology*, vol 67. Lippincott-Raven, New York, pp. 115–1129.

Lüders HO, Acharya J, Baumgartner C, Benbadis S, Bleasel A, Burgess R, Dinner DS, Ebner A, Foldvary N, Geller E, Hamer H, Holthausen H, Kotagal P, Morris H, Meencke HJ, Noachtar S, Rosenow F, Sakamoto A, Steinhoff BJ, Tuxhorn I, Wyllie E. (1998) Semiological seizure classification. *Epilepsia* 39:1006–1013.

Manford M, Fish DR, Shorvon SD. (1996) An analysis of clinical seizure patterns and their localizing value in frontal and temporal lobe epilepsies. *Brain* 119:17–40.

Matsumoto R, Ikeda A, Hitomi T, Aoki T, Hanakawa T, Miki Y, Tomimoto H, Shimohama S, Shibasaki H. (2005) Ictal monoparesis associated with lesions in the primary somatosensory area. *Neurology* 65:1476–1478.

Matsuzaka Y, Tanji J. (1996) Changing directions of forthcoming arm movements: neuronal activity in the presupplementary and supplementary motor area of monkey cerebral cortex. *J Neurophysiol* 76:2327–2342.

Meletti S, Rubboli G, Testoni S, Michelucci R, Cantalupo G, Stanzani-Maserati M, Calbucci F, Tassinari CA. (2003) Early ictal speech and motor inhibition in fronto-mesial epileptic seizures: a polygraphic study in one patient. *Clin Neurophysiol* 114:56–62.

Morris HH III, Dinner DS, Lüders H, Wyllie E, Kramer R. (1988) Supplementary motor seizures: clinical and electroencephalographic findings. *Neurology* 38:1075–1082.

Nakamura K, Sakai K, Hikosaka O. (1998) Neuronal activity in medial frontal cortex during learning of sequential procedures. *J Neurophysiol* 80:2671–2687.

Noachtar S, Lüders HO. (1999) Focal akinetic seizures as documented by electroencephalography and video recordings. *Neurology* 53:427–429.

Penfield W, Welch K. (1949) The supplementary motor area in the cerebral cortex of man. *Trans Am Neurol Assoc* 74:179–184.

Penfield W, Welch K. (1951) The supplementary motor area of the cerebral cortex; a clinical and experimental study. *Arch Neurol Psychiatry* 66:289–317.

Rubboli G, Mai R, Meletti S, Francione S, Cardinale F, Tassi L, Lo Russo G, Stanzani-Maserati M, Cantalupo G, Tassinari CA. (2006) Negative myoclonus induced by cortical electrical stimulation in epileptic patients. *Brain* 129:65–81.

Rushworth MF, Hadland KA, Paus T, Sipila PK. (2002) Role of the human medial frontal cortex in task switching: a combined fMRI and TMS study. *J Neurophysiol* 87:2577–2592.

Sahyoun C, Floyer-Lea A, Johansen-Berg H, Matthews PM. (2004) Towards an understanding of gait control: brain activation during the anticipation, preparation and execution of foot movements. *Neuroimage* 21:568–575.

Salanova V, Morris HH, Van Ness P, Kotagal P, Wyllie E, Lüders H. (1995) Frontal lobe seizures: electroclinical syndromes. *Epilepsia* 36:16–24.

Shima K, Tanji J. (1998) Both supplementary and presupplementary motor areas are crucial for the temporal organization of multiple movements. *J Neurophysiol* 80:3247–3260.

Tanji J, Shima K. (1994) Role for supplementary motor area cells in planning several movements ahead. *Nature* 371:413–416.

Wieshmann UC, Niehaus L, Meierkord H. (1997) Ictal speech arrest and parasagittal lesions. *Eur Neurol* 38:123–127.