

Gelastic seizures involving the right parietal lobe

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ABSTRACT – Gelastic seizures have been described in various epilepsies arising from the temporal or frontal lobes, although the most commonly encountered form is related to the presence of an hypothalamic hamartoma. We report a patient with gelastic seizures involving the right parietal lobe. Our patient, a 32-year-old man, underwent video-EEG monitoring, interictal and ictal brain SPECTs during gelastic seizures. Subtraction ictal SPECT co-registered to MRI (SISCOM), was performed to localize any ictal hyperperfusion during these gelastic seizures. The seizures consisted of brief staring followed by smiling and laughing. Electroencephalography during the gelastic seizures showed rhythmic sharp waves in the right parietal lobe. SISCOM showed ictal hyperperfusion in the right parietal lobe and medial portions of right cerebellum. Our findings suggest that the right parietal lobe may actively participate in the particular epileptogenic network generating gelastic seizures.

Key words: gelastic seizure, parietal lobe, cerebellum, SPECT

Gelastic seizures (GS) are characterized by bursts of laughter, often associated with other types of seizures, such as, generalized tonic-clonic and atonic seizures, and are poorly responsive to antiepileptic drugs (Chen and Forster, 1973). GS or laughter epilepsy has been described in various epilepsies as arising most commonly from hypothalamic hamartoma, and rarely from temporal (Tassinari *et al.* 1997) or frontal regions (Sartori *et al.* 1999).

However, GS have not been described in parietal lobe epilepsy. We report on an adult man with GS arising, on the basis of video-EEG monitoring and SISCOM (subtraction ictal SPECT co-

registered with MRI), from the right parietal lobe.

Patient and method

A 32-year-old, right-handed man had been attending our hospital for the evaluation of frequent complex partial seizures, since the age of 29 years. His seizures consisted of brief staring followed by smiling and then giggling. He could not recall any sensory symptoms prior to the seizures. No postictal confusion was observed. Family and personal past medical history were unremarkable. Seizures occurred five to 10 times per day despite treatment associating 40 mg of diphenylhydantoin

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toin, 60 mg of phenobarbital, 700 mg of carbamazepine and 200 mg/d of topiramate. Video-EEG monitoring captured fourteen seizures. During the ictal period, the patient stared for a few seconds and then suddenly started smiling and laughing for 20-40 seconds senselessly, inappropriately, and involuntarily. He could not remember the event afterwards. Interictal EEGs showed frequent spike waves on the right parietal region, and pre-ictal repetitive sharp waves on the right parietal region (*figure 1A*) followed by ictal rhythmic spikes intermixed with fast activities in the same region (*figure 1B*). Brain magnetic resonance imaging showed no definite abnormality. The detailed cortical morphology of the right parietal lobe was inspected by a gyroscore function of Analyze 6.0 (Biomedical Imaging Resource), which revealed no abnormalities. However, the cortex of the right parietal lobe was significantly thinner than that of left parietal lobe (left parietal lobe: 3.22 ± 0.49 mm, right parietal lobe: 3.13 ± 0.39 mm, $p < 0.0001$), which suggested mild cortical gray matter atrophy of the right parietal lobe region. However, interictal ^{18}F -fluorodeoxyglucose positron emission tomography (FDG-PET) showed no focal hypometabolism. Ictal brain single photon emission computed tomography (SPECT) with $^{99\text{m}}\text{Tc}$ -ethylcysteinate dimer (ECD) was performed during his GS (injection time: 20 sec for a 37 sec seizure duration) and the interictal brain SPECT was performed by injection of $^{99\text{m}}\text{Tc}$ -ECD following a 24 hour or more, seizure-free period. Following epilepsy monitoring and adjustment of treatment, the seizure frequency decreased to one per month on 900 mg of oxcarbazepine and 20 mg/d of clobazam. SISCOM was performed using a commercial software package ANALYZE 6.0 (Lee, 2000).

Result

Ictal SPECT showed slightly increased perfusion in the right parietal lobe (*figure 1B*) and SISCOM showed definite ictal hyperperfusion in the right parietal region, right cerebellar vermis and right medially located cerebellar nuclei (fastigial nuclei, emboliform nuclei, dentate nuclei) (*figure 1C*).

Discussion

We describe a patient with adult-onset gelastic seizures (GS) with ictal EEG and ictal hyperperfusion localized to the right parietal lobe. GS are characterized by sudden attacks of laughter, which are most commonly associated with hypothalamic hamartomas. The later onset of gelastic epilepsy in the presence of a hypothalamic hamartoma appears to be associated with a milder epilepsy syndrome, less severe learning difficulties and behavior problems, and better occupational and social status (Mullatti, 2003). Our patient too showed no learning difficulties or behavior

problems and his seizures were relatively well controlled by antiepileptic medications.

A previous study reported a patient in whom electrical cortical stimulation applied to the inferior temporal gyrus produced mirth alone or laughter preceded by mirth (Sattow *et al.* 2003). A case report including a SPECT study, showed ictal hyperperfusion in the right cingulate gyrus, but not in the hamartoma, suggesting that the cingulate gyrus could be either the origin or part of the seizure pathway (Gordon *et al.* 1996). Electrical stimulation in the right cingulate gyrus elicited smiling and laughter, but no mirth in another study (Sperli *et al.* 2006). Arroyo *et al.* (1993) suggested that the motor act of laughing and the processing of its emotional content were represented separately in, respectively, the anterior cingulate area and the basal temporal area (the fusiform gyrus or parahippocampal gyrus, or both). Fried *et al.* (1998) suggested not only that laughter and mirth were represented in the pre-supplementary motor area, but also that there was close linkage between the motor, affective, and cognitive components of laughter.

In our case, SISCOM revealed ictal hyperperfusion in the right parietal lobe, while ictal SPECT showed slightly increased perfusion in the right parietal region. The higher sensitivity of SISCOM compared to conventional ictal SPECT in this context, has been previously reported (Lee *et al.* 2000).

The interictal scalp EEG findings may show focal abnormalities in temporal, frontal, or parasagittal regions (Berkovic *et al.* 1988; Kuzniecky *et al.* 1997; Gordon *et al.* 1996). Focal discharges have also been reported in these areas during seizures (Kuzniecky *et al.* 1997; Gordon *et al.* 1996), but the most common EEG features are diffuse changes, such as, voltage attenuation, slowing, and even generalized spike-wave paroxysms (Munari and Francione, 1997; Berkovic *et al.* 1988). In symptomatic forms, EEG studies with deep electrodes have shown that the onset of ictal paroxysmal activity corresponds to the lesion site, although surface EEG show simultaneous diffuse changes (Munari and Francione, 1997; Kahane *et al.* 2003). In the case reported here, the EEG abnormalities were always focal, interictal on the right parietal region, and ictal on the right parietal lobe.

A previous study showed that in two patients, an ictal smile arose from the parietal lobe during other complex partial seizures (Berkovic *et al.* 1988). They observed smiling during parietal (two patients) and temporal lobe (three patients) seizures, and the right hemisphere appeared to be involved with a greater frequency. Neuroanatomic models of emotional processing, especially laughter, suggest the temporal lobe as the main site of initiation for emotional processing. The cerebellum is another important structure in emotional processing through the cerebro-ponto-cerebellar pathway (Parvizi *et al.* 2001). Under normal circumstances, a perceived external stimulus (e.g. being told a funny joke) or a recalled one (e.g.

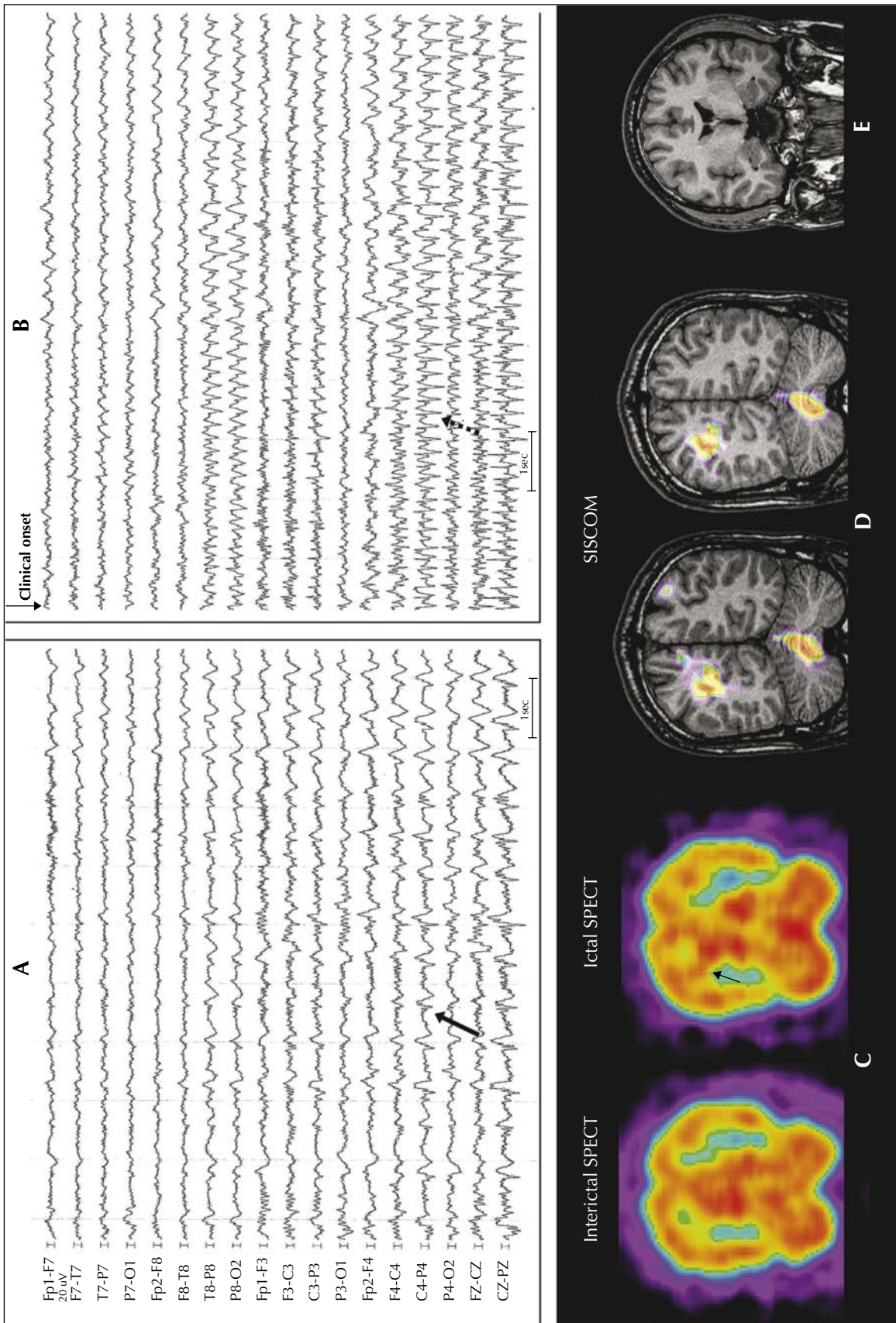


Figure 1. Ictal EEG and SISCOM. The patient's gelastic seizure (arrow) was preceded by pre-ictal repetitive sharp waves (solid arrow) in the parasagittal region with a right parietal maximum (A), and then ictal discharges progressed to repetitive spikes intermixed with fast activities (dotted arrow) on right parietal region (B). Ictal SPECT (arrow) and SISCOM showed ictal hyperperfusion in the right parietal region and right cerebellar vermis (C). Hypothalamus, frontal and temporal lobe show no abnormality (D).

remembering the death of a friend) will trigger an emotional response, but only if the cognitive/social context is appropriate. Although the laughter and crying components of the emotional responses are largely pre-programmed and stereotyped, their intensity, duration and certain aspects of the overall pattern depend on the cognitive/social context in which the triggering stimulus appeared (Provine, 1996). It is suggested that the cerebellum plays an important role in the modulatory control described above (Parvizi et al. 2001). In a group of 14 subjects who re-enacted a previous sad event, eight of whom cried during the acquisition of images, the left basis pontis and the right cerebellum showed increased activity (Fiez and Raichle, 1997). In our case, SISCOM revealed ictal hyperperfusion in the medial portions of the right cerebellum including the vermis, fastigial and dentate nuclei (figure 1), which suggests that the right cerebellum is activated during GS.

The most common cause of symptomatic gelastic epilepsies is hypothalamic hamartoma which, like ectopic grey matter, has an intrinsic paroxysmal activity (Munari and Francione, 1997). Thus, on spreading to the temporal, frontal, and diencephalic structures this epileptogenic activity would trigger other clinical signs (Kuzniecky et al. 1997). Although brain MRI and FDG-PET of our case showed no abnormalities on visual inspection, the measurement of cortical thickness revealed decreased cortical thickness in the right parietal lobe. Moreover, ictal EEG abnormalities in our case were focal, unlike most diencephalic gelastic seizures in which ictal EEG abnormalities are diffuse (Berkovic et al. 1988).

In summary, we describe a patient with GS who showed ictal EEG discharges on the right parietal region and ictal hyperperfusion in the right parietal lobe and right cerebellum. □

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