

Mirthful gelastic seizures with ictal involvement of temporobasal regions

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Received January 15, 2008; Accepted January 26, 2009

ABSTRACT – Ictal laughter is the cardinal clinical sign of gelastic seizures in hypothalamic hamartomas and may also occur in extrahypothalamic epilepsies. Laughing consists of an affective and a motor component. It has been suggested that the affective component may result from an involvement of temporobasal structures, whereas the motor part is related to an involvement of the mesial frontal cortex. So far, evidence is based on a limited number of cases with spontaneously recorded seizures or in whom electrical stimulation of invasive intracranial EEG recordings has been performed. We report a patient who suffered from epigastric psychic auras, complex partial seizures with a gelastic component and secondarily generalized seizures. To evaluate a possible epileptogenic role of the hippocampus and dysplastic region in the right mid-temporal gyrus, intracranial monitoring with subdural electrodes over the temporobasal and temporolateral regions, as well as a deep brain electrode in the hippocampus, were performed. During the initial part of the seizure, consisting of an intense retrosternal ascending feeling with sexual connotation, rhythmic spikes in temporolateral contacts were detected. Concomitant with the development of smiling and laughter, a rhythmic activity over the temporobasal regions evolved. The patient became seizure-free following right temporal lobe resection. This case supports the assumption that ictal involvement of temporobasal structures is crucial for gelastic seizure components in patients with temporal lobe epilepsy. Progression to temporobasal regions was associated with the feeling of happiness whereas motor components of laughter occurred later. These findings are in accordance with the interpretation of surface recordings by Dericiglu and co-workers in a similar previous case. [*Published with video sequences*].

Key words: gelastic seizures, right temporal origin, invasive recordings, intracranial EEG, temporal lobe, ictal laughter



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Gelastic seizures are usually regarded as an almost pathognomonic sign of an epileptogenic region in the hypothalamus, typically due to a hypothalamic hamartoma (Munari *et al.* 1995, Palmioli *et al.* 2005). However, cases with extrahypothalamic epileptogenic zones, such as the cingulate gyrus and basal temporal regions, have also

been reported (Arroyo *et al.* 1993, Dericiglu *et al.* 2005, Stefan *et al.* 2004). Electrophysiological data from patients with extrahypothalamic seizure onset zones may provide a better understanding of the symptomatogenic regions. Laughing during gelastic seizures may consist of an affective and a motor component (Gascon and

doi: 10.1684/epd.2009.0245

Lombroso 1971). It has been suggested that the affective component may result from an involvement of temporobasal structures, whereas the motor part is related to an involvement of the mesial frontal cortex. So far, evidence for the localisation of the affective component of laughter is based on a limited number of cases in whom electrical stimulation of invasive intracranial EEG recordings has been performed (Arroyo *et al.* 1993, Stefan *et al.* 2004).

Case report

A 42-year-old right-handed, otherwise healthy, male patient had suffered from epileptic seizures since the age of 16 years. Initially these were characterized by an isolated feeling of extreme pleasure localized to the epigastric region, which could be triggered by emotional stimuli (e.g. watching emotive films) and were associated with smiling or laughing (type I). In the course of the disease, he developed complex partial seizures with early gelastic components consisting of grinning, beating of the thighs with pleasure and snorting with laughter, followed by hypermotor limb movements, fixed gaze, loss of responsiveness and rarely clonic movements of the head (type II). This often evolved into head version to his left side, vocalization and secondary generalization of seizures (type III). Postictal amnesia for seizure events was common.

The epilepsy was refractory to antiepileptic treatment with phenytoin, lamotrigine, oxcarbazepine and levetiracetam. The patient thus underwent presurgical evaluation at the Freiburg Epilepsy center.

High resolution MR scanning showed a slight T2 hyperintensity of the deep right mid-temporal gyrus and possible signal changes in the right hippocampus. On video-EEG-monitoring with surface electrodes, interictal spikes over the right temporal lobe were shown. In the same area ictal patterns consisting of rhythmic activity were also detected; however, they only occurred well after clinical seizure onset. Neuropsychological testing gave evidence of slight deficits in verbal memory, suggesting a dysfunction of the temporal lobe of the dominant hemisphere. Functional MRI revealed left hemispheric language representation based on left opercular and temporolateral activations during tasks of semantic decision and word generation (Spreer *et al.* 2002).

In order to carry out a tailored resection in the absence of an unequivocal lesion, invasive recordings were performed using right subdural electrode strips, three strips located basally and three laterally over the temporal lobe, and a depth electrode placed longitudinally into the right hippocampus (*figure 1*). Furthermore, two days later standard surface electrodes were added.

Interictal spikes occurred mostly temporolaterally, but also temporobasally and within the hippocampus. Seven

seizures were recorded: four auras, very short and without the full feeling of mirth, that initially resembled type I, one complex partial seizure with preceding gelastic phenomena (type II), and two secondarily generalized seizures with an initial gelastic component (type III).

In all seizures, the ictal patterns, consisting of rhythmic spikes, started over the right lateral temporal lobe (*figure 1A*). The initial gelastic semiology was a slight elevation of the right hand in an attentive manner indicating the patient's subjective experience of mirth, two seconds after electroencephalographic seizure onset in the right temporolateral cortex, mid-temporal gyrus, with rapid spread to temporobasal contacts (*figure 1B*). Following transition to low amplitude fast activity discharges and spread to temporobasal regions, clinical manifestations of smiling and subsequent laughter developed. Later, hypermotor movements of the legs and clonic movements of the head followed with further spread of epileptic activity (*see video sequence*).

During isolated auras, consisting of retrosternal feeling typically preceding the experience of mirth, the ictal activity remained localized to the temporolateral cortex. According to the results of invasive monitoring, a tailored resection restricted to the temporomedial and inferior gyrus was performed (*figure 2*). Histology showed a focal cortical dysplasia Type 2a according to Palmini *et al.* (2004). The patient has been seizure-free so far for more than 12 months under monotherapy with lamotrigine.

Discussion

This case provides evidence for the origin of seizures in the temporolateral cortex in a patient with gelastic seizures comprising both emotional and pronounced motor phenomena. The onset of gelastic signs coincided with the rapid spread of ictal epileptic activity to temporobasal regions, which is compatible with the role of temporobasal limbic regions as the symptomatogenic region also in this patient. Thus, our results are in agreement with data from patients suffering from gelastic seizures with a temporobasal origin (Dericioglu *et al.* 2005). The role of this region in the generation of mirth has been shown by electric stimulation in earlier studies (Arroyo *et al.* 1993, Satow *et al.* 2003), whereas the patients reported in these studies did not suffer from gelastic seizures. As the patient described here had no electrodes implanted in frontal or cingular regions, spread of the epileptic activity to these regions and their exact role in the generation of motor phenomena of laughter could not be evaluated.

Whereas the semiology of the gelastic seizure component does not allow us to distinguish this patient from patients with seizure origin in the hypothalamus (Oehl *et al.* 2008), the late onset of the disease is very uncommon in those with

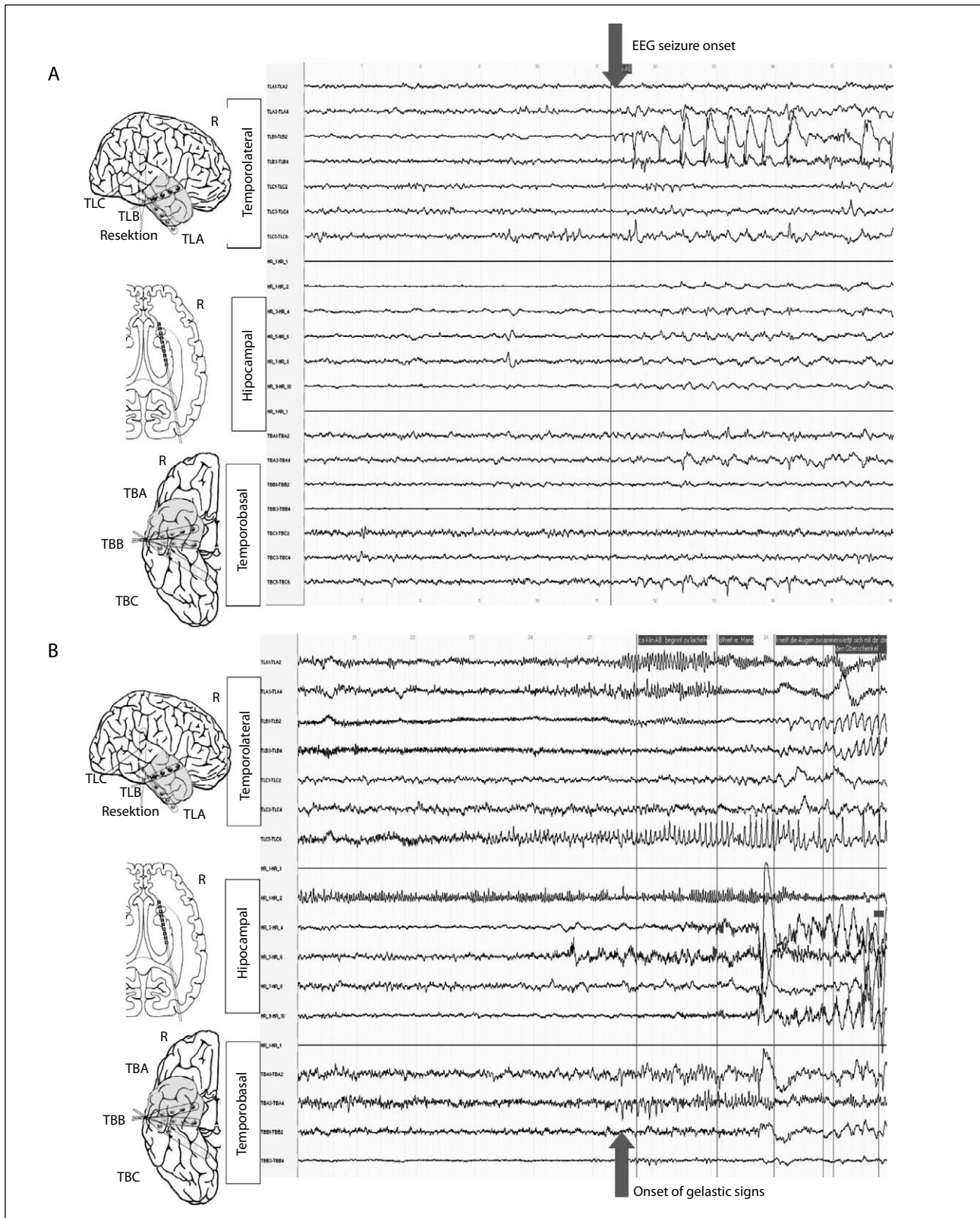


Figure 1. A) EEG recording showing temporolateral electroencephalographic seizure onset. B) EEG recording at clinical seizure onset with gelastic component: spread of rhythmic epileptic activity to the temporobasal region.

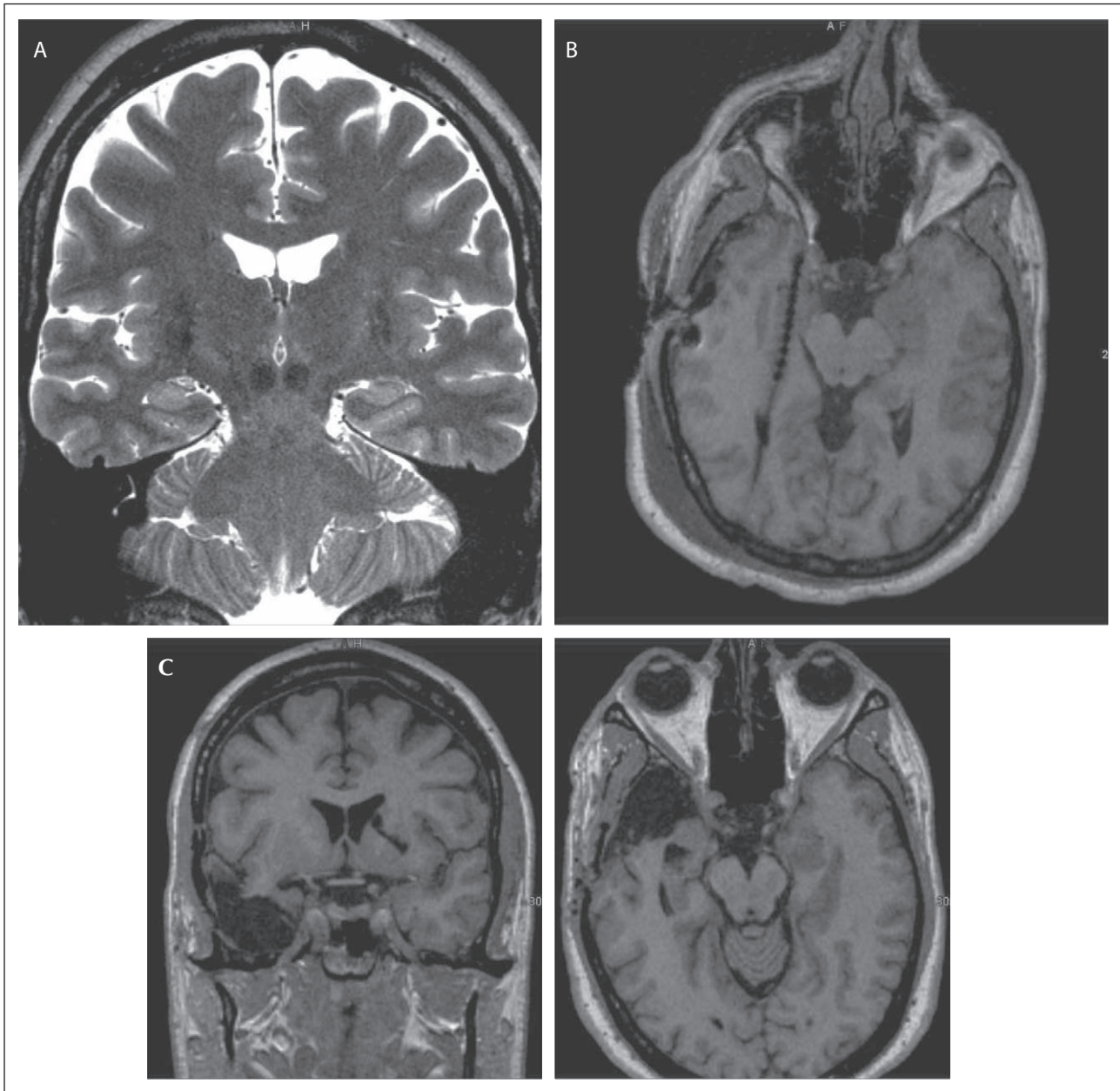


Figure 2. A) MRI, T2, showing the temporolateral cortex and the hippocampi. B) MRI, T1, showing the right hippocampal depth electrode. C) MRI, T1-mp-rage, three months after tailored resection.

hypothalamic hamartomas and may thus point towards a less common extrahypothalamic lesion. Resection was successful as is often the case in patients with focal cortical dysplasia (Fauser *et al.* 2004, Fauser *et al.* 2008) thus offering a better outcome compared to patients with hypothalamic hamartomas (Schulze-Bonhage *et al.* 2007).

In conclusion, this case report is the first to demonstrate the coincidence of the beginning of the affective component of epileptic laughter and the evolution of electrophysiological seizure activity in temporobasomesial structures. □

Legend for video sequence

As initial gelastic semiology the video shows a slight elevation of the right hand indicating the patient's subjective experience of mirth two seconds after EEG seizure onset in the right temporo-lateral cortex – medial temporal gyrus with rapid spread to temporo-basal contacts. Following transition to low amplitude fast activity discharges and spread to temporobasal regions, smiling and subsequent laughter are to be seen succeeded by hypermotor movements of the legs and clonic movements of the head.

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