CLINICAL CASE REPORT

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Epileptic Laughter: 2 Case Reports

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Key words: *gelastic seizures; epileptic laughter; urinary incontinence.*

Summary. *Two cases of gelastic epilepsy in a 6-year-old girl with attacks of mirthful laughter and a 38-year-old male patient with episodes of laughter without any positive emotions are presented. Temporal lobe epilepsy was diagnosed in the first case and possible frontal lobe epilepsy in the second case. It is concluded that that this rare form of epilepsy can be difficult to diagnose and treat, and can clinically be accompanied by urinary incontinence.*

Introduction

Gelastic epilepsy is a relatively rare form of epilepsy characterized by laughing seizures and was first described by Armand Trousseau in 1877. It has been thought to account for less than 0.5%–1% of all types of epilepsy (1). Gascon and Lambroso gave the criteria to define gelastic epilepsy. According to these criteria, it is characterized by stereotyped seizures of prevailing laughter, inappropriate to the context, concomitant with other manifestations generally compatible with seizures, and associated with interictal or ictal discharges on electroencephalography (EEG) (2).

In most cases, gelastic epilepsy is associated with hypothalamic hamartomas originating from the tuber cinereum or the mammillary bodies. However, in less frequent cases, gelastic seizures may originate from the frontal or temporal regions (3). Depending on the location of the lesion, gelastic seizures may or may not be accompanied by a subjective feeling of mirth (3). It has been supposed that gelastic seizures with the sense of mirth involve structures crucial in emotional processing such as medial temporal regions (amygdala, fusiform gyrus, parahippocampal gyrus) and seizures without the sense of mirth are more likely to be related to anterior cingulate, premotor frontal, and opercular areas (4). Other phenomena may be present as well in addition to gelastic features of the seizure reflecting the origin and spread of epileptic activity in the brain.

Case Reports

Case 1

A 6-year-old girl had suffered from the bouts of laughter since early childhood, usually beginning with a mirth situation. She could not get out of the laughter cycle, which lasted up to 15 minutes. The attack always ended with wetting her pants. It occurred only during daytime. Such attacks occurred several times a day (up to 10–12 attacks per day), usually in emotional situations, for example when a large group of people were together (in kindergarten or family meetings).

Her parents were worried about wetting her pants; the laughter did not seem to be forced or unreasonable to them. However, they had been consulted by a pediatric nephrologist who diagnosed stress urinary incontinence and recommended special physiotherapy and an every-day regimen, which had no effect on the problem.

She is the first child in the family, born in term, and had a normal development. She has been free of diapers since the age of 2 years and has had the bouts of laughter resulting in wetting the pants ever since.

At the age of 6 years, she was evaluated by a psychologist who advised to consult with a child neurologist. During the first interview with the pediatric neurologist, gelastic epilepsy was suspected because the mother said that when starting to laugh, her daughter could not stop laughing, and at the end of attack, she was not responding to speech. The attacks always ended with wetting her pants, and the child could not avoid it. EEG revealed epileptiform spike-slow wave complexes and sharp waves in the right T4F8 areas accompanied by focal slowing (Fig. 1). There were normal features of sleep with persisting epileptiform changes over the right hemisphere. The child stayed at the hospital for 4 days (with her mother); no episodes of epileptic laughter occurred, and thus, it was not possible to record the ictal EEG.

MRI revealed no structural changes (Fig. 2). Psychological examination showed that the child had a completely normal development and cognitive functions according to her age.

Treatment with oxcarbazepine with a final dose of up to 900 mg daily was administered. After 2 weeks of treatment, the laughing bouts diminished by half during the first 3 months of treatment, but EEG findings after 1 month were the same. The decision was made to add valproic acid, and the mother

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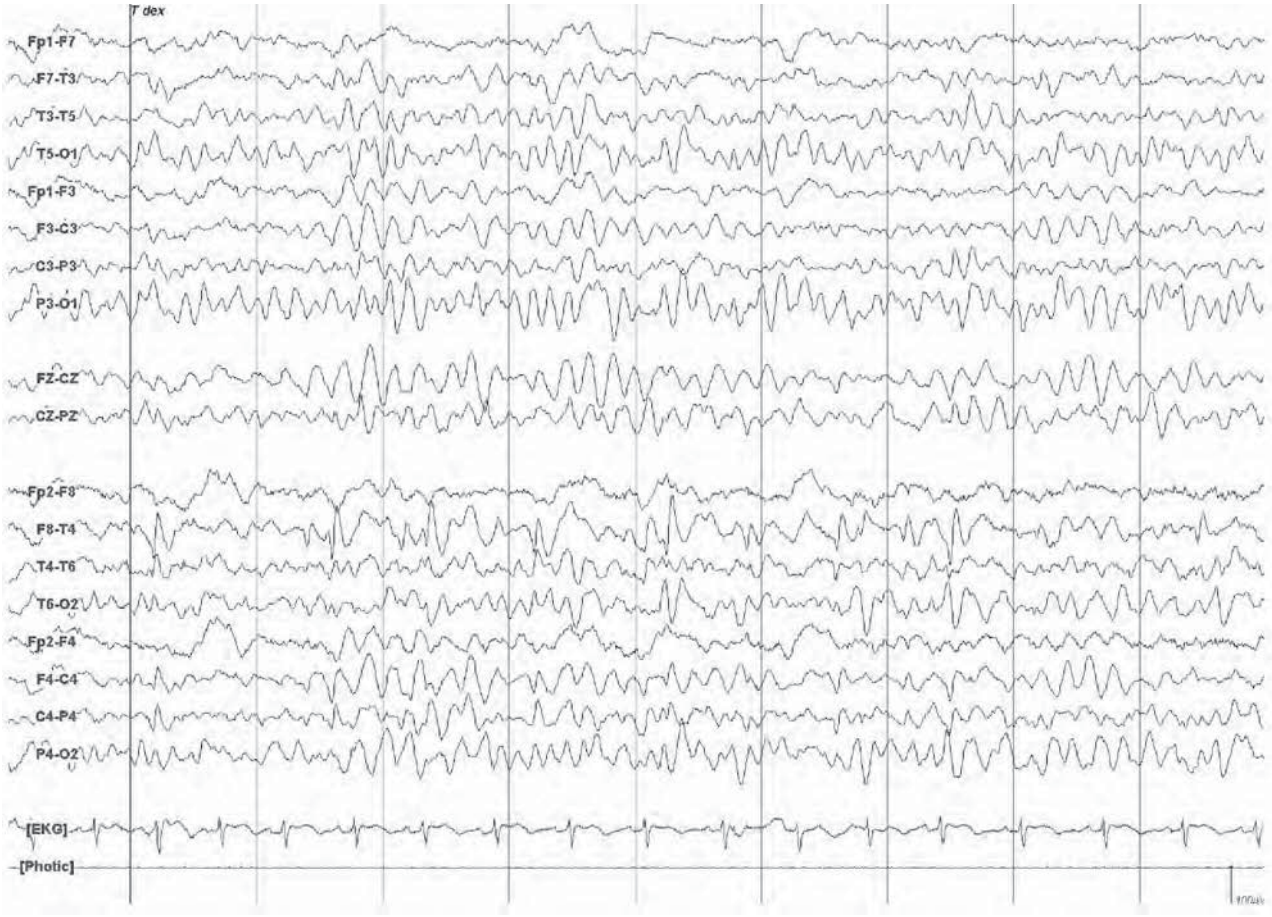


Fig. 1. EEG of patient 1 showing a normal background activity with interictal epileptic sharp waves and focal slowing in the right temporal region (before treatment)

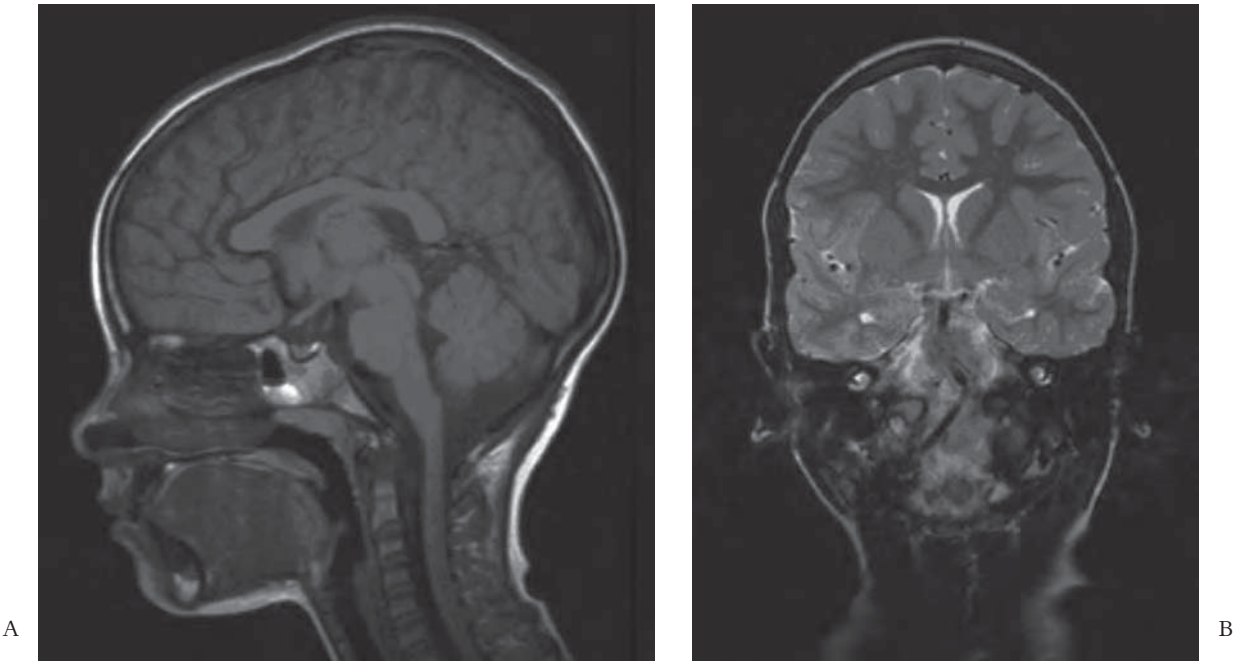


Fig. 2. Magnetic resonance imaging
A, fluid-attenuated inversion recovery sagittal plain; B, T2-weighted coronal plain.

was informed about possible adverse events. After 3 weeks of treatment, the child was free of seizures, and EEG findings were normal 3 months later. During the follow-up period, the child stayed free of seizures with medications.

Case 2

A 38-year-old male patient had treatment-resistant epilepsy with the frequent seizures of laughter. He described short episodes (some seconds in duration) with a vague unpleasant hollow feeling in the stomach and head. Moreover, the patient compared the subjective feelings with shivering with cold, or having a wave in the abdomen and the chest, or fear of heights. Usually he had many episodes (up to 50) a day; occasionally, several times every day. These subjective feelings increased in intensity, and within some seconds, laughter for the next 5–10 seconds followed. The laughter was never accompanied by the subjective feelings of mirth. The patient as a rule forcefully hid his face with his hands or any available object (book). Sometimes while trying to escape from this situation (shameful as he felt at that stage), he fell. Spontaneous micturition was typical of the episodes whilst he was awake. The patient claimed to remember events during the seizure. He said that he had never completely lost control over his actions. The patient had similar, but not so frequent episodes during sleep (1–2 times every night). Usually, there was no micturition during the nocturnal seizures.

The first seizures appeared at the age of 14 years. The frequency of seizures was 7–10 times a day, most often at night.

Treatment with carbamazepine was started, and the patient achieved remission. After being free of seizures for 3 years, the treatment was discontinued. The patient reported that the nocturnal seizures persisted all the time. The seizures during daytime reoccurred at the age of 32 years, and the treatment with carbamazepine and later lamotrigine was administered. Although the patient had frequent daily seizures, he experienced that the seizure frequency could be extremely high after the withdrawal of his medication. He noticed that strong alcohol had a preventive effect on seizures for some following days.

Neurological examination did not show any cognitive impairment and motor or sensory deficiency. Cranial MRI showed a small nonspecific lesion in white matter of the left frontal lobe (Fig. 4). No hypothalamic hamartoma or any other relevant pathology was detected. Repeated EEG studies did not show any interictal epileptic activity.

Long-term EEG monitoring was performed for 70 hours. During the monitoring, 20 habitual seizures were recorded. The majority of the seizures occurred during sleep and 4 seizures whilst awake. Video recording of the seizures showed the patient turning his head to mask his face by the pillow and doing some automatic movements with hands and legs (nocturnal seizure), or along with some automatisms in hands and legs hid his head and face with hands or the book and started to laugh (Video). The seizure lasted about 5–10 seconds. At the end of the seizure, the patient immediately acted adequately, answered the questions, and claimed to remember everything. Still according to the video material, he seemed to respond with latency during this short seizure. Quite often, the patient had urinary incontinence during the seizure. Electroencephalographically only desynchronization of the background rhythm could be seen at the beginning of the seizure, which was then replaced by mild slowing and movement artefacts (Fig. 3). After the seizure, a normal rhythm of wakefulness appeared.

The seizures were refractory after the trials of several anticonvulsants as mono- or polytherapy regimens (carbamazepine, lamotrigine, topiramate, and levetiracetam). Clonazepam at a dose of 8 mg per day was relatively effective against short-lasting subjective "auras" for some weeks after introducing the drug. Currently the patient is using carbamazepine at a dose of 1800 mg per day and has many seizures every day, but the situation in general is somehow better than previously.

Discussion

The first case presents a rare form of temporal lobe epilepsy presenting with gelastic seizures. Despite the fact that there are some features not typical of gelastic seizures, mainly the duration of episodes, which may last as long as 15 or even 20 minutes, there were continuous epileptiform discharges in the EEG, and the patient responded very well to the treatment with

Video. Two typical seizures of patient 2 are presented

Seizures during the daytime show the episodes of giggling as the patient is trying to hide his face with his hands. The nurse is trying to communicate with the patient, as he appears to be fully cooperative and conscious.

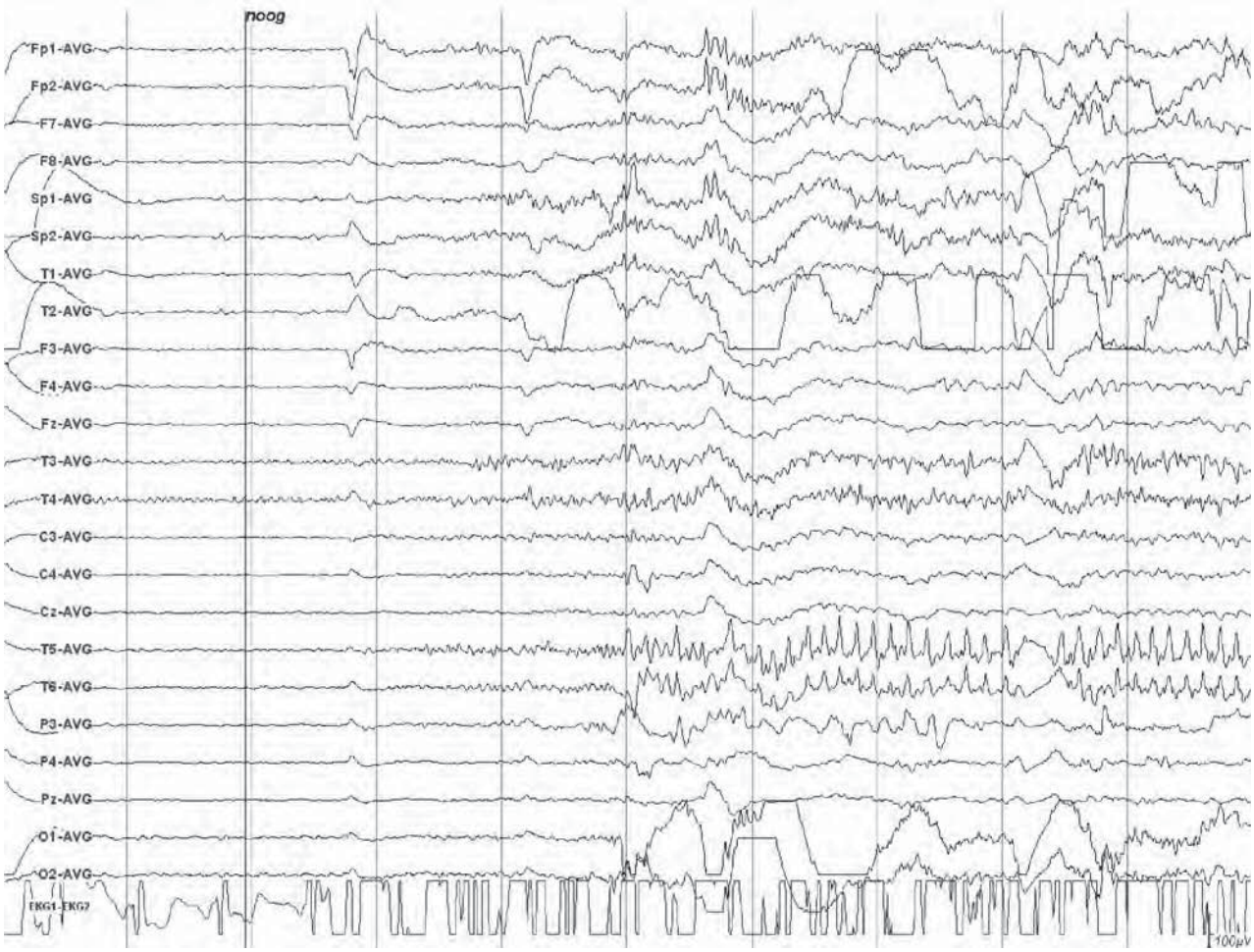
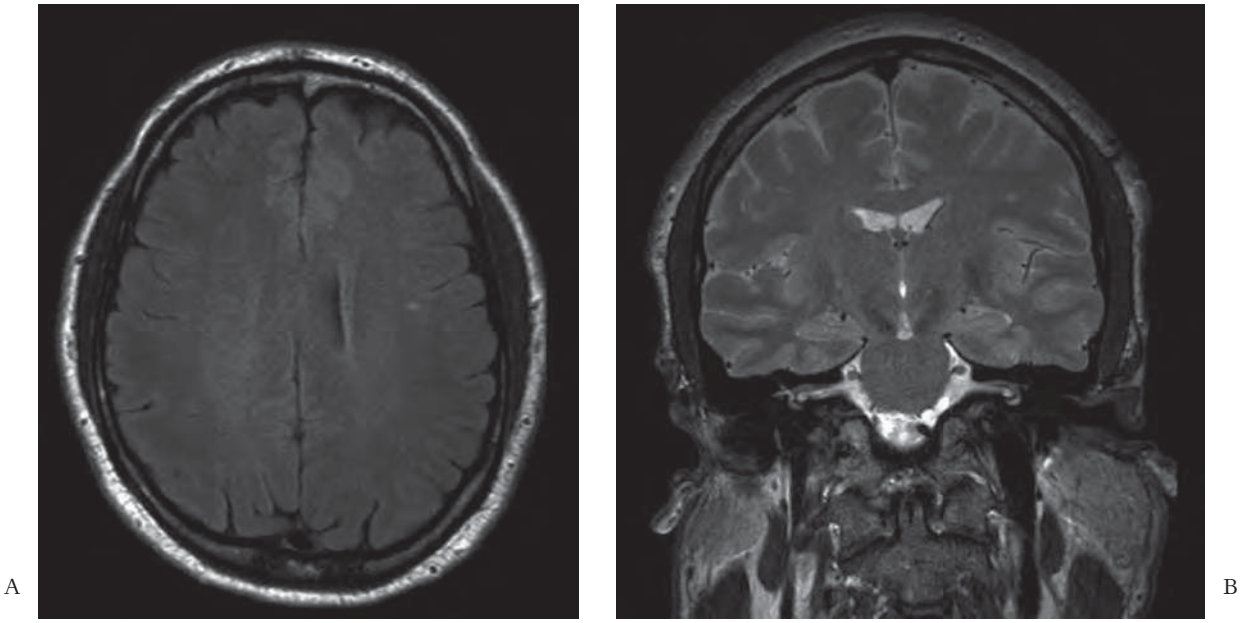


Fig. 3. Ictal EEG of patient 2 showing the beginning of the seizure
Desynchronization of the background rhythm is replaced by mild slowing and movement artefacts.



A B

Fig. 4. MRI images of patient 2
A, fluid-attenuated inversion recovery axial image; B, T2-weighted coronal plain.

an antiepileptic drug. Probably, the usual appearance and long duration of these laughing episodes were the main reasons for relatively late consultations with a neurologist and the diagnosis of gelastic epilepsy. The diagnosis was delayed also because the focus was on the urinary incontinence, which was the main problem and interfering symptom for the family.

In the second case, the brief inappropriate bursts of laughter were soon recognized as epileptic seizures, and treatment with an antiepileptic medication was initiated, but after an initial period of possible remission, the seizures recurred and were proven refractory thereafter. Although, clinical manifestations of the seizures and the lack of sense of mirth would fit to the seizure onset in the frontal lobe, imaging studies have not revealed compatible pathologic changes in the brain.

The pathophysiology of mirth and laughter is complicated, and various regions of the brain are involved both in the emotional and motor components of the process. Gelastic seizures may originate from different lobes, and evidently, the expressive symptoms depend on the site of the lesion (3, 5). In the first case, the accompanying subjective feeling was not known, but to some degree, the presence of mirth could be suspected. This and the concordant epileptic discharges in the right temporal region indicate that the seizures originate from the temporal lobe. In the second case, only the motor expression of laughter was present (without any epileptic changes on the EEG), and therefore, the origin of seizures from the frontal lobe can be suspected.

Although various motor symptoms and also micturition have been described in previous cases of gelastic epilepsy (6), it may be speculated that the prominent symptom – urinary incontinence at the end of each attack – indicates a decreased level of consciousness and/or spread of the epileptic discharges to other areas of the brain. However, urinary incontinence occurs more often with generalized convulsions and is thought to be the result of suddenly increased abdominal pressure or accompanying seizures from the parasagittal frontal lobe regions (7). In the second case of our case report, the involvement of consciousness and its relationship to micturition are difficult to determine. The seizures were short lasting, and the patient claimed to be conscious during the whole seizure, but his behavior seen on the video recording refers to certain latency in his behavior, which could be related to the disturbances of consciousness.

It has been described that in certain cases, gelastic seizures can be triggered by several factors including manual activation (8) or even emotional excitation (9). The parents of the child described that the girl's seizures often started with normal laughter in an emotional or humorous situation. Therefore, in the first case, it could be hypothesized that the seizures could have been triggered by normal laughter and emotional state.

Cryptogenic or symptomatic gelastic epilepsy with etiologies other than a hypothalamic hamartoma can be medically difficult to treat. In one study of 10 children with gelastic epilepsy of various etiologies, 4 became free of seizures with drug therapy (10). The first patient achieved remission with antiepileptic drugs, and the whole family improved its quality of life. The initial results of drug therapy were excellent for the second patient as well, but became disappointing later.

Conclusions

Two cases of gelastic epilepsy are presented, which indicated that the etiology and origin of gelastic seizures can be associated with several brain regions outside hypothalamic hamartomas, and this rare form of epilepsy can be difficult to diagnose and treat and can clinically be accompanied by urinary incontinence.

Statement of Conflict of Interest

The authors state no conflict of interest.

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