Epileptic Disord 2012; 14 (2): 187-9

Psychogenic gelastic seizures in a patient with hypothalamic hamartoma

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Received December 16, 2011; Accepted April 6, 2012

ABSTRACT – Gelastic seizures are classically associated with hypothalamic hamartoma. The most effective treatment for gelastic epilepsy is surgery, although confirming that a hypothalamic hamartoma is an epileptic lesion prior to surgical intervention is challenging. Here, we report the case of a patient with a hypothalamic hamartoma who was diagnosed with psychogenic non-epileptic gelastic seizures using video-EEG monitoring. [*Published with video sequences*]

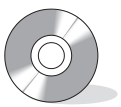
Key words: hypothalamic hamartoma, gelastic seizure, psychogenic non-epileptic seizure, video-EEG monitoring

Hypothalamic hamartomas (HH) are classically associated with gelastic seizures. Epilepsy associated with HH is characterised by early onset of gelastic seizures and later development of multiple seizure types, medical intractability, and encephalopathy. Surgical or radiosurgical lesion ablation offers the best chance of seizure control (Tellez-Zenteno *et al*., 2008). Surface electroencephalographic (EEG) recordings are of limited utility in patients with HH and gelastic seizures (Troester *et al*., 2011). Indeed, the decision to proceed with surgery may rely heavily on clinical evidence of an epilepsy syndrome consistent with HH in a patient with a known lesion.

We report the case of a patient with HH who had non-epileptic gelastic seizures based on observed semiology during video-EEG monitoring. This case underscores the need for video and EEG monitoring in the verification of intractable clinical epilepsy syndromes.

Case Report

A 50-year-old man presented to our epilepsy clinic with a complaint of gelastic seizures. Imaging conducted seven years prior had incidentally found a hypothalamic hamartoma. He reported the subsequent development of gelastic seizures, which involved spontaneous paroxysms of laughing or crying that lasted seconds to minutes and occurred up to 12 times daily. He reported maintained consciousness during events, which sometimes occurred out of sleep. In retrospect, he attributed frequent outbursts of laughter and anger in childhood to his epilepsy.



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doi:10.1684/epd.2012.0509

He also reported generalised tonic-clonic seizures five to six times per month, staring spells six times per day, and drop attacks one to two times per month. A final seizure type involved tingling across the back of his head, followed by right arm and leg jerking and repetition of the word "mom". He was conscious during the events but unable to interact with his environment.

He was taking 400 mg gabapentin, three times daily. Seizure control had previously been attempted with multiple agents, none of which controlled his episodes, and all of which were discontinued due to rash. After previous evaluation at other centres, he had been told his seizures were non-epileptic.

Magnetic resonance imaging revealed a suprasellar mass consistent with hypothalamic hamartoma, as well as extensive bilateral cerebral band heterotopia, accessory sulcation, and focal pachygyria of the right fronto-temporal region (*figure 1*). The patient was admitted to our in-patient epilepsy monitoring unit for further characterisation of his seizures as part of a work-up for possible epilepsy surgery. Continuous video-EEG monitoring was conducted for four days and no antiepileptic drugs were given. He was noted to bring a large stuffed toy with him to the hospital.

Five events were captured during video monitoring, including two events involving laughter-like vocalisations. The most dramatic occurred during a hyperventilation procedure *(see video sequence*). A second event involved tingling over his head for six minutes, after which he said "ha-ha" repeatedly, but more slowly and with longer breaks between vocalisations than the above-described event. He was non-responsive and his eyes were closed. He stuttered and whispered when asked to name objects afterwards. Three other non-stereotyped events were recorded which involved eye closure, stuttering, whispering, grunting, arm raising, and apparent arm paralysis. There were no surface EEG changes with any of these clinical events. The interictal EEG was also unremarkable. All of these events were determined to be psychogenic non-epileptic seizures.

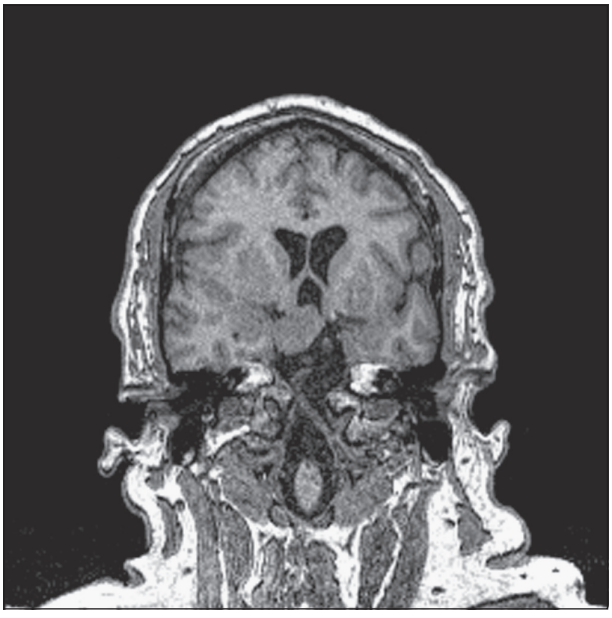

Figure 1. Coronal cut of T1 weighted magnetic resonance imaging revealing a suprasellar mass, iso-signal to grey matter, extending into the hypothalamic region posterior to the pituitary infundibulum.
This is consistent with a hypothalamic hamartoma. Note also accessory sulcation of the right fronto-temporal cortex.

Discussion

Hypothalamic hamartomas are rare in the general population, but the incidence of gelastic seizures in patients with these lesions has not been reported. In this case, the association of a known HH and the patient's stated history, including a supposed early onset of gelastic seizures with later development of different seizure types and medical intractability, biased clinical reasoning towards surgical work-up (Brandberg *et al*., 2004; Tellez-Zenteno *et al*., 2008; Troester *et al*., 2011). However, intractable events are common in psychogenic non-epileptic seizures (PNES), a diagnosis consistent with his late-life presentation following discovery of an incidental lesion and previous diagnoses of anxiety and somatoform disorders (LaFrance and Benbadis, 2011).

A diagnosis of PNES was made in this patient based on an observed semiology that was inconsistent with epileptic gelastic seizures. The laughter during epileptic gelastic seizures has been described as explosive, inappropriate, and stereotyped. Though the clinical semiologies of gelastic seizures are variable, some have described them as mirthful and similar to spontaneous normal laughter (Berkovic *et al*., 1988; Oehl, 2010). In contrast, our patient showed slow and rhythmic laughter that would be difficult to mistake for true mirthful laughter and differed in quality between the two laughing spells that were captured. While ictal laughing may reasonably be without surface EEG findings, his other ictal movements are unlikely to be electrographically silent (Troester *et al*., 2011).

The presence of a stuffed toy in the monitoring room, ictal stuttering, ictal eye closure, and a fluctuating course of motor activity were all observed in this case and are highly specific for PNES (Hoerth *et al*., 2008; Avbersek and Sisodiya, 2010). Additionally, we were able to provoke an event with hyperventilation, a test with high sensitivity for inducing events in patients with PNES (Benbadis *et al*., 2000).

It is important to exclude the possibility that a single patient may have both epileptic and non-epileptic

events. Here, each observed clinical event involved some or all of the characteristics noted above to be specific for non-epileptic semiology. Despite his report of multiple daily occurrences of more classic epileptic seizure types at home, none of these were observed in the hospital. Also, his interictal EEG was without abnormalities. For these reasons, we were confident that epileptic seizures were not present in this patient. This case illustrates that, for some patients, observation of events and the use of clinical judgment is the most accurate method of making the proper diagnosis. Use of his stated history and imaging results as justification for surgery would have exposed him to substantial operative risk from a procedure not directed towards his true illness. □

Disclosures.
Michael Macken has the following disclosures: Advisory Board, Speaker Bureau - UCB Pharma; Advisory Board, Speaker Bureau - Lundbeck Inc. All other authors have no financial disclosures to report and have declared that no conflict of interest exists.

Legend for videosequence

A psychogenic non-epileptic seizure in a patient with a hypothalamic hamartoma. The patient's eyes flutter and he raises both arms above his head. His eyes then close, and he repeats "ha-ha" in a continuous, rhythmic manner while keeping his arms raised. His left arm begins to move side-to-side, and continues to intermittently do so. He repeats "ha-ha" for 50 seconds and then begins to repeat the word "mama". He lowers his arms at this point and flaccid paralysis is noted. About 90 seconds after laughing started, he opens his eyes and is asked to name a pen. He responds by whispering and stuttering a "p" sound. He continues not to obey commands for about 9 minutes, after which he begins to cry. The episode lasts about 11 minutes in total. The observed quality of ictal laughter differs from published descriptions of epileptic gelastic seizures. Additionally, note the presence of eye closure, fluctuating motor movements, and stuttering, all of which are specific signs of a non-epileptic event.

Key words for video research on www.epilepticdisorders.com
Syndrome: gelastic seizures with hypothalamic hamartoma
Etiology: hamartoma (hypothalamic)
Phenomenology: gelastic seizure; nonepileptic paroxysmal event
Localization: midline lesion

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