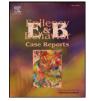
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Ictal laughter and crying: Should they be classified as automatisms?

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ABSTRACT

Gelastic seizures (GS) describe ictal laughter and are associated with hypothalamic lesions, as well as other cortical areas. Dacrystic seizures (DS), characterized by ictal crying, also have been reported in hypothalamic lesions and focal epilepsy. We describe a young girl with drug resistant focal dyscognitive seizures associated with gelastic and dacrystic features. However, neither laughter nor crying was correlated with a stereotyped electroencephalographic (EEG) pattern or involvement of a particular brain region. Additionally, based on the variety of epileptogenic foci associated with GS and DS in the literature, laughter and crying appear to represent ictal or peri-ictal automatisms.

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1. Introduction

Gelastic seizures (GS) are ictal events that are characterized by episodes of stereotyped, unprovoked laughter. They have been most classically associated with hypothalamic lesions, particularly hamartomas [1]. GS have also been described in patients with epileptogenic zones in frontal, parietal, and temporal lobes [2]. Inappropriate laughter, either secondary to epilepsy or other neurologic diseases, has been implicated in multiple cortical, subcortical and brainstem structures [3]. Dacrystic seizures (DS) are characterized by paroxysmal ictal or peri-ictal crying or weeping [4]. They have been reported in patients with hypothalamic hamartomas, frontotemporal and orbitofrontal seizures [5].

GS and DS have rarely been found to co-exist in patients with hypothalamic hamartomas, both during the same seizures and within the same patients [6]. Here, we describe child with drug-resistant parietalopercular epilepsy who presented with a both GS and DS. While the patient became seizure free after the combination of parietal and opercular resection with a subsequent posterior insular laser ablation, neither scalp nor intracranial EEG localized these behaviors to a well-defined symptomatogenic zone suggesting that laughter and weeping represent automatisms, which may be due to activation of cortical–subcortical networks and/or disinhibition of brainstem centers [7].

1.1. Case presentation

A 5 year-old girl with pharmacoresistant epilepsy was referred for presurgical evaluation. Her caretaker described her seizures as episodes of inappropriate laughter, stiffening of right arm and increased movement in left arm. She had no perinatal complications. She exhibited global developmental delay and mild right hemiparesis. She underwent MRI, scalp-video-EEG, ictal SPECT, as well as intracranial EEG monitoring and intraoperative electrocorticography. MRI revealed cortical thickening and dysplasia involving the left parietal operculum, which extended to the posterior insula (Fig. 1A). Ictal SPECT showed increased uptake in the left anterior temporal/posterior parietal region and right cerebellar hemisphere. During the ictal SPECT, the patient only exhibited focal seizures with motor symptomatology, and did not exhibit gelastic features.

Scalp video-EEG monitoring was performed on two occasions (once two years earlier) and 23 seizures were available for review from two evaluations. The seizures were focal dyscognitive, associated with ictal laughter or crying, nonversive head-turning to the right, and dystonic posturing of right arm with increased repetitive stereotyped movements of the left arm. During many seizures, the patient also reached for her caretaker and held her in a prolonged embrace. Autonomic changes were also noted during seizures, including ictal tachycardia and respiratory changes (hyperventilation). The ictal discharge began with left centroparietal 1–2 Hz spike-and-wave complexes lasting 10– 20 s, intermittently attenuating and alternating with paroxysmal fast activity (10–15 Hz). Laughter was not correlated with a stereotyped ictal discharge. When GS occurred, they were observed early in the

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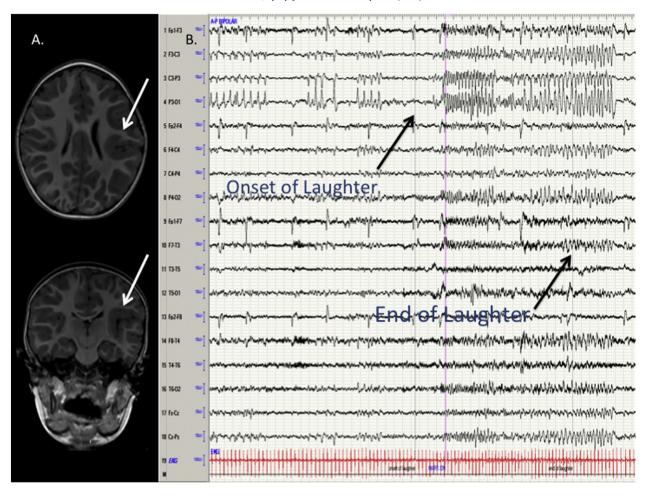


Fig. 1. Neuroimaging and ictal scalp EEG sample. MRI images on the left (top axial, lower coronal), red arrows indicating dysplastic cortex, and ictal scalp EEG sample on the right. The patient was laughing during and between recurring paroxysms of spiking or paroxysmal fast activity (her photo and scalp EEG sample is on the right, recorded on an anterior-posterior bipolar montage).

seizures with an average latency of 12 s from the EEG onset (Video 1). It was the initial clinical symptom in 10 (45%) seizures. Dacrystic features usually occurred near the end of the seizure, or postictally (Video 2).

The patient was implanted with subdural grids and depth electrodes. Her antiseizure drugs (ASDs) were continued (at half her home dose) throughout most of the monitoring, and only held for one day prior to the end of the recording. No electrodes sampled the cingulate gyrus to document mesial propagation. Thirteen clinical seizures were recorded, but only once did she exhibit gelastic features. Ictal smiling and crying occurred 5 and 3 times, respectively. Most seizures were associated with an ictal discharge in the parietal opercular dysplastic cortex, as well as the insular depth electrode. The seizures involving gelastic and dacrystic behavior demonstrated no difference in ictal correlate from other seizures. Dacrystic features did however occur mostly during the latter aspect of the seizure or immediately postictally.

2. Results

2.1. Semiological analysis

Two investigators reviewed all recorded seizures, marking the onset and offset of gelastic and dacrystic semiology with respect to the ictal EEG onset and termination on both scalp and intracranial EEG recordings. The presentation of gelastic and dacrystic behaviors was also correlated with other ictal symptoms, such as smiling or grimacing, embracing, and other motor features, mainly dystonic posturing of the right upper extremity. Clustering of semiological symptoms from scalp and intracranial video-EEG recordings are listed in Table 1. Laughter (35%) occurred more often than crying or weeping (24%) during her seizures. Laughter occurred as an isolated symptom in five out of 12 seizures (41.7%), and crying occurred as an isolated symptom in only one out of 8 seizures (12.5%). They only coincided in two seizures, and in both cases, laughter preceded crying or weeping. When other behaviors were present, laughter and crying clustered with motor features, in 33% and 37.5%, respectively and embracing in 16% and 37.5%, respectively. Motor symptoms and embracing were clustered in 53% of seizures (Table 1).

2.2. Surgical procedures and post-operative outcome

She underwent a parietal opercular resection with partial reduction of her seizures at age 5 years and 8 months, followed by laser ablation of the parietal operculum and posterior insula at age 72 months (6 years old) rendering her seizure free for over a year. Postoperatively, she developed transient right arm weakness.

3. Discussion

This case report describes a patient with medically refractory focal dyscognitive seizures with gelastic and dacrystic behaviors. In contrast to other such reports in the literature she does not have a hypothalamic hamartoma, but rather a parietal opercular cortical dysplasia extending into the posterior insula. While she was rendered seizure free by resection and laser ablation of the dysplastic cortices, neither scalp nor intracranial EEG recordings identified a well-defined symptomatogenic zone or stereotyped ictal EEG to correlate with gelastic or dacrystic behaviors.

| Table 1 | |
|-----------------------------|------------------------------|
| Scalp and intracranial vide | o-EEG semiological findings. |

| Video-EEG evaluation | Clinical seizures | Smiling | Laughter | Motor symptoms | Embrace | Crying |
|----------------------|-------------------|------------|-----------------|----------------|------------|-----------------|
| Scalp #1 | 13 | 4 | 6 (1 postictal) | 8 | 7 | 4 (3 postictal) |
| Scalp #2 | 8 | 3 | 5 | 1 | 2 | 1 (postictal) |
| Intracranial | 13 | 5 | 1 | 6 | 6 | 3 |
| Total | 34 | 12 (35.2%) | 12 (35.2%) | 15 (44.1%) | 15 (44.1%) | 8 (23.5%) |
| Associated symptoms | | | | | | |
| Smiling | 2 only | 4 | | 6 | 6 | 1 |
| Laughter | 4 | 5 01 | nly | 4 | 2 | 2 |
| Motor symptoms | 6 | 4 | - | 2 only | 8 | 3 |
| Embrace | 6 | 2 | | 8 | 1 only | 3 |
| Crying | 1 | 2 | | 3 | 3 | 1 only |

Legend: Total number of seizures demonstrating gelastic or dacrystic behaviors, and their clustering with other ictal motor symptoms are presented. Scalp #1 and #2 indicate her scalp video-EEG evaluation.

This, in combination with previous literature describing the association between GS and DS with various cortical epilepsies [6,8,9], suggest that GS and DS are not associated with a particular epileptogenic zone or brain region. Rather, gelastic and dacrystic semiologies represent ictal automatisms and their expression results from the interaction of cortical and subcortical networks. These automatisms may be provoked by cortical–subcortical loops of involving the limbic pathways; or, as with oral automatisms, they may be a result of deactivation of cortical volitional pathways modulating emotional expression. More specifically, ictal cortical deactivation could lead to disinhibition of the hypothalamic-periaqueductal gray-pontine pathway [3], thus leading to inappropriate emotional expression.

3.1. Conclusion

In summary, we suggest that GS and DS are likely to represent ictal or peri-ictal automatisms. These behaviors are thought to be manifestations of activation of cortical-subcortical limbic networks, associated with cortical disinhibition of subcortically or brainstem-mediated motor behaviors [7]. In conjunction with other types of automatisms, gelastic or dacrystic semiologies in our case were not as useful for localizing the epileptogenic zone as previously reported.

Conflict of interest statement

We have no conflicts of interest to declare.

Study ethics and patient consent

This study was performed in accordance with "The Code of Ethics of the World Medical Association" and consent for publication of this case report was obtained from the patient's mother. The following are the supplementary data related to this article. Supplementary data to this article can be found online at http://dx. doi.org/10.1016/j.ebcr.2016.11.004.

References

- Téllez-Zenteno JF, Serrano-Almeida C, Moien-Afshari F. Gelastic seizures associated with hypothalamic hamartomas. An update in the clinical presentation, diagnosis and treatment. Neuropsychiatr Dis Treat 2008;4:1021–31.
- [2] Kovac S, Diehl B, Wehner T, Fois TD, Walker MC, et al. Gelastic seizures: incidence, clinical and EEG features in adult patients undergoing video-EEG telemetry. Epilepsia 2015;56:e1–5.
- [3] Lauterbach EC, Cummings JL, Kuppuswamy PS. Toward a more precise, clinicallyinformed pathophysiology of pathological laughing and crying. Neurosci Biobehav Rev 2013;37:1893–916.
- [4] Offen ML, Davidoff RA, Troost BT, Richey ET. Dacrystic epilepsy. J Neurol Neurosurg Psychiatry 1976;39:829–34.
- [5] Burghardt T, Basha MM, Fuerst D, Mittal S. Crying with sorrow evoked by electrocortical stimulation. Epileptic Disord 2013;15:72–5.
- [6] Munari C, Quarato P, Kahane P, Tassi L, Minotti L, Hoffman D, et al. Gelastic and dacrystic seizures. In: Lüders HO, Noachtar S, editors. Epileptic seizures: pathophysiology and clinical semiology, 1st ed.Philadelphia: Churchill Livingston; 2000. p. 458–71.
- [7] Szabó CÀ, Lüders HO. Postictal automatisms. In: Lüders HO, Noachtar S, editors. Epileptic seizures: pathophysiology and clinical semiology, 1st ed.Philadelphia: Churchill Livingston; 2000. p. 658–62.
- [8] Tran TP, Truong VT, Wilk M, Tayah T, Bouthillier A, Mohamed I, et al. Different localizations underlying cortical gelastic epilepsy: case series and review of literature. Epilepsy Behav 2014;35:34–41.
- [9] Blumberg J, Fernández IS, Vendrame M, Oehl B, Tatum WO, Schuele S, et al. Dacrystic seizures: demographic, semiologic, and etiologic insights from a multicenter study in long-term video-EEG monitoring units. Epilepsia 2012;53:1810–9.