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Case Report

Refractory Temporal Gelastic Seizure: A Case Report

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Abstract

Gelastic seizures are rare epileptic events characterized by sudden, unprovoked bursts of laughter that are typically associated with hypothalamic hamartomas but may also arise from cortical epileptogenic foci. We report the case of an eight-year-old boy born prematurely at 28 weeks' gestation who presented with a two-year history of recurrent hypermotor seizures, loss of consciousness, behavioral arrest, falls, and frequent episodes of inappropriate laughter. Video electroencephalography demonstrated epileptiform activity consistent with left temporal lobe epilepsy with mild diffuse encephalopathy, while a 1.5 Tesla brain magnetic resonance imaging (MRI) showed no structural abnormality. Clinical evaluation also revealed mild microcephaly and learning difficulties. Initial treatment with carbamazepine followed by combination therapy with levetiracetam failed to adequately control seizures. However, seizure frequency improved after substitution of levetiracetam with lamotrigine. This case highlights that gelastic seizures may originate from the temporal lobe even in the absence of hypothalamic hamartoma and may be resistant to first-line therapy. Awareness of this rare presentation may facilitate earlier recognition and optimization of treatment strategies in affected patients.

Keywords: Gelastic, Temporal, Seizure, Refractory

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Introduction

Gelastic seizures are rare epileptic events causing sudden, unprovoked bursts of laughter or smiling, often brief (seconds) and inappropriate to the situation.[1]They are classically associated with a benign glial tumor called hypothalamic hamartomas and are slightly more common in males.[2] Laughing seizures can also arise from a frontal or temporal lobe focus. Inappropriate laughter with Ictal or interictal discharges correlates are diagnostic.[2],[3] The clinic-radiologic syndrome of hypothalamic hamartoma (HH) manifests with a variety of symptoms, including pharmaco-resistant epilepsy with multiple seizure types, precocious puberty, behavioral disturbances, and cognitive impairment.[3][4] The scalp electroencephalogram (EEG) is usually normal in children with gelastic seizures, causing a delay in the diagnosis of epilepsy and the finding of a hypothalamic hamartoma.[5]However, in a proportion of children with HH, there is an epileptic progression, in which complex partial seizures with frontal, temporal, and lateralized clinical features appear, usually with the appearance of focal slowing and epileptiform activity on the interictal EEG.[5] Gelastic seizures are usually resistant to treatment but can be achieved through surgical excision of causative intracranial lesions or use of anti-seizure medications for focal seizures, such as carbamazepine.[6][7] Interstitial radiosurgery was efficacious in significantly improving gelastic epilepsy, though weight gain may occur as a side effect.[8] This report, therefore, aims at creating an awareness of this rare gelastic seizure of temporal origin and how its refractory seizures were successfully managed.

Case Report

We describe the case of D.S., an eight-year-old male born preterm at 28 wks G.A with a birth weight of 1.9kg. He presented to our facility with a history of recurrent hypermotor seizures with loss of consciousness, behavioral arrest and falls, bursts of laughter that occurred every day, of two years duration. Each episode lasted for about 3 minutes. There was an associated pre-ictal aura as the boy could tell when the seizures were about to occur, but could not describe how he felt. The seizures were not associated with post-ictal coma and occurred several times a day. The first episode of seizure in the child's life occurred with fever; it was generalized-clonic in nature but was poorly managed in a peripheral hospital. There is no family history of seizures. Gross motor milestone was delayed till two years of age, speech was not delayed, and social milestone was attained within normal limits. On presentation to our facility, a routine video electroencephalogram (vEEG) revealed an abnormal study in keeping with left temporal lobe epilepsy (T5) with mild diffuse encephalopathy. Brain MRI (1.5T) done showed a normal study. However, a further epilepsy protocol MRI was not done due to its unavailability in our facility.

Physical examination showed a hyperactive child with bruises and a non- tender mass measuring 2 by 2 cm on the forehead due to repeated falls, and had a normal gait. He was conscious and alert, well oriented in time, person and place. He had mild microcephaly, learning difficulties (dysgraphia and poor cognition) with suboptimal Draw-A-Person Test (DAPT), and he could not copy a circle. He had normal tone in the limbs with brisk deep tendon reflexes. Ankle clonus was not sustained, and there were no signs of meningeal irritation. Other systemic examination findings were essentially normal. Hence, a diagnosis of Gelastic seizures with left temporal complex partial seizures, Microcephaly and Learning difficulty secondary to Prematurity was made.

Initial treatment with carbamazepine followed by combination therapy with levetiracetam failed to adequately control seizures. However, seizure frequency improved after substitution of levetiracetam with lamotrigine.

Ethical approval was obtained from the Human Research Ethics Committee (HREC) of Federal Medical Centre, Umuahia and written parental consent was obtained, with the patient's anonymity adhered to in the report of this case. The CARE (Case Report) guideline[9] was adopted in the writing of this report.

Discussion

Investigations into the neurological correlates of laughter remain fragmentary and exist mainly as case reports. By employing the classical methods of neurology, brain regions associated with symptomatic (pathological) laughter have been determined and catalogued under other diagnostic signs and symptoms of such conditions as epilepsy, strokes and circumspect brain lesions.[8] Gelastic seizures are usually associated with

hypothalamic hamartomas, but can also be seen in frontal lobe and temporal lobe epilepsies.[2] The index patient in this report had a vEEG with evidence of temporal lobe involvement. This is in keeping with the five-year cohort study by Kovac et al. [3]. It was documented that out of 19 patients with gelastic seizures, one-third had gelastic seizures of temporal lobe origin. Another one-third had seizures originating from the hypothalamus, while the others were either from the parietal lobe, frontal, multifocal or undetermined. However, a case report by Costa et al [7] showed a gelastic seizure of frontal origin, while Mirandola et al documented that out of 16 patients who had gelastic seizures, the hypothalamic hamartoma was more frequent.[1] Scholly et al [2], on the other hand, documented that Intracerebral stereotactic EEG (SEEG) explorations demonstrated more complex, both reciprocal and hierarchical, relationships within the hypothalamo-cortical epileptogenic networks. And suggested that network formation may be due to either secondary epileptogenesis or widespread epileptogenicity present at the outset.[2] It was also demonstrated that gelastic and dacrystic seizures were correlated with discharges within the HH, whereas other seizure types were related to discharges affecting cortical regions, which sometimes seemed to be triggered by HH. The concept of kindling-like secondary epileptogenesis was suggested as a possible cause of epileptic encephalopathy.[2] Another report contrary to that of our index patient is that of a gelastic seizure in a 29-year-old woman caused by a focal cortical dysplasia, which was difficult to localize with EEG. Localization for the patient was done through MRI and SPECT imaging.[10] Our index patient had a vEEG with evidence of temporal lobe involvement, with a 1.5 Tesla brain MRI that showed a normal study. However, further investigations, such as epilepsy-protocol MRI or SPECT imaging, could not be done for the patient because of the paucity of the needed gadgets in our centre.

Concerning the epidemiology of gelastic seizures, the prevalence of those without a hypothalamic hamartoma, like in our index patient, is not known. This is because gelastic seizures are rare.[11] However, the childhood prevalence of gelastic seizures caused by hypothalamic hamartomas was estimated at 0.5 per 100,000 [11], and the number of cases reported in the literature is considerably smaller.

The pathophysiology of gelastic seizure is yet undefined.[12]

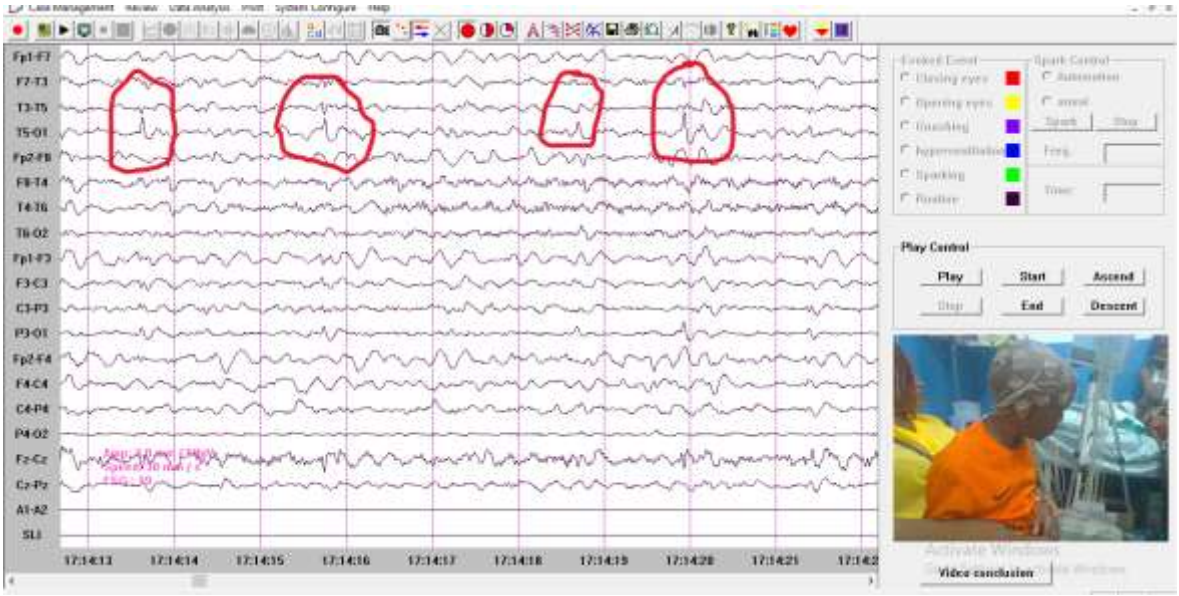
The index patient in this study had seizure semiology that manifested as concurrent outbursts of laughter and hypermotor complex partial seizures, which were refractory, while Kovac et al documented automotor seizures, which are mainly seen in temporal lobe epilepsies but can be seen in frontal lobe epilepsies too.[3] Mirandola *et al*[1] documented that the majority of the patients studied had outbursts of laughter while the rest smiled. The patients with Hypothalamic hamartoma had emotional laughter, while other epileptogenic zones showed no emotional laughter. Costa *et al* [7] documented a case report of a 76-year-old woman with spontaneous intracranial hemorrhage presented with right lower limb focal motor seizures with myoclonic jerks of her right lower limb and also intermittent bursts of emotionless, non-provoked laughter, alternating with less frequent episodes of uncontrolled crying (dacrystic). Investigations excluded autoimmune, paraneoplastic, metabolic, infectious, and endocrine causes. Demir et al [13] in a multi-centre study observed that gelastic seizures with smiling were a more homogenous group with regard to location in the temporal lobe. This is, however, contrary to the finding on the index patient, who instead had emotional bursts of laughter despite having temporal lobe epilepsy. These differences in the clinical manifestations of gelastic seizures may not be clearly understood, as extra-hypothalamic gelastic seizures are rarer with poorly defined pathophysiology.[12] Other clinical features of gelastic seizures noted by Scholly *et al* [2] were that pharmaco-resistant epilepsy with multiple seizure types caused precocious puberty, behavioural disturbances and cognitive impairment. Half of the patients studied had epileptic syndrome due to involvement of distal cortical regions and persistent seizures.[2] The precocious puberty, behavioural disturbances and cognitive impairment were also noted by Striano et al.[4] These clinical features could be due to involvement of the hypothalamus, which, however, were not observed in the index patient.

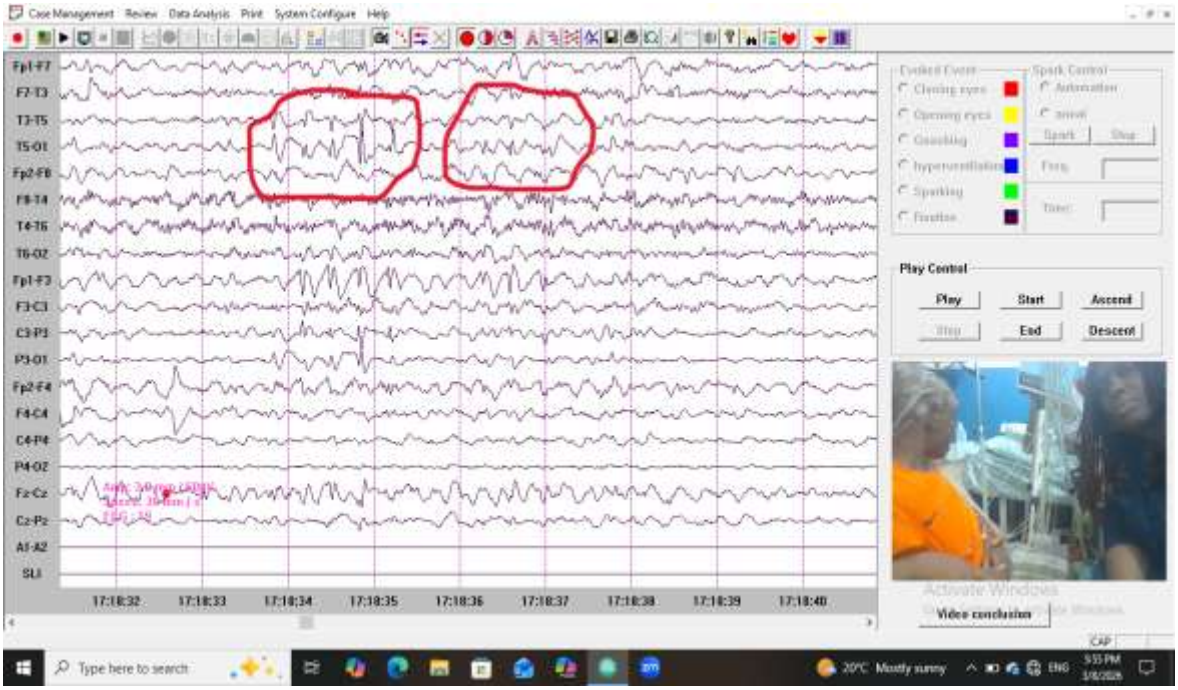
A thorough workup for gelastic seizures should include neuroimaging with attention to the suprasellar region and EEG.[14] This is similar to the investigations carried out for the index patient, even though the 1.5 Tesla brain MRI showed a normal study. More advanced investigations, like SPECT or epilepsy-protocol MRI, would have been able to reveal if there was actually a structural lesion, but they were not done due to the unavailability of the needed gadgets to run such imaging. Accurate, early diagnosis and patient education have been documented to be critical in averting excessive and unnecessary treatments.

For the treatment outcome of gelastic seizure, the index patient did not respond to a combination of carbamazepine and levetiracetam. However, he later responded when Levetiracetam was replaced with Lamotrigine. On the other hand, the case report by Costa et al [7] documented a good response to phenytoin and levetiracetam combination. This discrepancy could be because of the effect of drug-drug interaction, which could differ in different drug combinations and in different individuals. There was no need for surgical intervention in the index patient since the 1.5 Tesla brain MRI done showed a normal study, and the patient responded well to medical therapy. The most effective treatment for gelastic epilepsy is surgery. However, it is challenging to confirm that a hypothalamic hamartoma is an epileptic lesion prior to surgical intervention.[15] The patients studied by Kovac et al [3] underwent hippocampal sclerosis and gamma knife radiosurgery, with good control of the seizures in one-quarter having a good outcome post-surgery. Hypothalamic hamartoma ablation followed by temporal lobectomy was recorded by Scholly et al [2] to have led to seizure freedom in two patients. While the same procedures led to partial improvement in patients with distal cortical lesions. They also documented inefficient therapeutic procedures in half of the patients. A short time window from epilepsy onset to surgery was noted to be crucial to cure epilepsy by direct surgery in hamartomas.[2] Savasta *et al* [6] documented good response to carbamazepine monotherapy in three patients with complete seizure control in addition to a benign clinical and cognitive outcome. This confirms that gelastic epilepsy without hypothalamic hamartoma, both in cryptogenic or symptomatic patients (one child showed a dysplastic right parietotemporal lesion), usually has a more benign natural history. It was noted that carbamazepine seems to be the most efficacious therapy to obtain both immediate and long-term seizure control in gelastic seizures. [6] This is partly similar to our index patient, who responded well to a carbamazepine- lamotrigene combination therapy rather than carbamazepine monotherapy.

Conclusion

Gelastic seizures can be of temporal lobe origin and may be difficult to treat, necessitating ASM polytherapy or surgery.





References

1. Mirandola L, Cantalupo G, d'Orsi G, Meletti S, Vaudano AE, Di Vito L, Vignoli A, Tassi L, Pelliccia V. Ictal semiology of gelastic seizures. *Epilepsy Behav.* 2023;140:109025. doi: 10.1016/j.yebeh.2022.109025. Epub 2023 Feb 11. PMID: 36780776.
2. Scholly J, Bartolomei F. Gelastic seizures and the hypothalamic hamartoma syndrome: Epileptogenesis beyond the lesion? *Handb Clin Neurol.* 2021;182:143-154. doi: 10.1016/B978-0-12-819973-2.00010-1. PMID: 34266589.
3. Kovac S, Diehl B, Wehner T, Fois C, Toms N, Walker MC, Duncan JS. Gelastic seizures: incidence, clinical and EEG features in adult patients undergoing video-EEG telemetry. *Epilepsia.* 2015;56(1):e1-5. doi: 10.1111/epi. 12868. Epub 2014 Dec 17. PMID: 25516460.
4. Striano S, Santulli L, Ianniciello M, Ferretti M, Romanelli P, Striano P. The gelastic seizures-hypothalamic hamartoma syndrome: facts, hypotheses, and perspectives. *Epilepsy Behav.* 2012;24(1):7-13. doi: 10.1016/j.yebeh.2012.02.013. PMID: 22503469.
5. Harvey AS, Freeman JL. Epilepsy in hypothalamic hamartoma: clinical and EEG features. Semin *Pediatr Neurol*. 2007 Jun;14(2):60-4. doi: 10.1016/j.spen.2007.03.003. PMID: 17544948.
6. Savasta S, Budetta M, Spartà MV, Carpentieri ML, Trasimeni G, Zavras N, Villa MP, Parisi P, Gelastic epilepsy without hypothalamic hamartoma: Three additional cases, *Epilepsy & Behavior*, 2014: (37); 87-90, https://doi.org/10.1016/j.yebeh.2014.06.012.(https://www.sciencedirect.com/science/article/pii/S1525505014002182)
7. Costa A, Supermanian S, Matar M, Scott J. Adult New Onset Gelastic Seizures of Cortical Origin. *Neurohospitalist.* 2020 Apr;10(2):148-149. doi: 10.1177/1941874419884216. Epub 2019 Nov 4. PMID: 32373282; PMCID: PMC7191665.
8. Schulze-Bonhage A, Trippel M, Wagner K, Bast T, Deimling FV, Ebner A, Elger C, Mayer T, Keimer R, Steinhoff BJ, Spreer J, Fauser S, Ostertag C. Outcome and predictors of interstitial radiosurgery in the treatment of gelastic epilepsy. *Neurology.* 2008 Jul 22;71(4):277-82. doi: 10.1212/01.wnl.0000318279.92233.82. PMID: 18645166.
9. Riley DS, Barber MS, Kienle GS, Aronson JK, von Schoen-Angerer T, Tugwell P, Kiene H, Helfand M, Altman DG, Sox H, Werthmann PG, Moher D, Rison RA, Shamseer L, Koch CA, Sun GH,

Hanaway P, Sudak NL, Kaszkin-Bettag M, Carpenter JE, Gagnier JJ. CARE guidelines for case reports: explanation and elaboration document. *J Clin Epidemiol.* 2017;(89):218-235. doi: 10.1016/j.jclinepi.2017.04.026. Epub 2017 May 18. PMID: 28529185.

10. Cheung CS, Parrent AG, Burneo JG. Gelastic seizures: not always hypothalamic hamartoma. Epileptic Disord. 2007; 9(4):453-8. doi: 10.1684/epd. 2007.0139. PMID: 18077234.
11. Jeremy LF, Eeg-Olofsson O. Gelastic Seizure. *Neupsykey internet*. www.https//neupsykey.com/gelastic seizure/ (accessed 22/03/2026)
12. Chaouki S, Boujraf S, Atmani S, Elaqam L, Messouak W. Gelastic seizures and fever originating from a parietal cortical dysplasia. J Pediatr Neurosci. 2013;8(1):70-2. doi: 10.4103/1817-1745.111433
13. Demir AB, Öz BY, Yıldız MO, Türk BG, Tanrıverdi T, Bekar A, Yeni N, Bora İ. A clinical evaluation of gelastic and dacrystic seizures: a multicenter study. Arq Neuropsiquiatr. 2022 Dec;80(12):1204-1212. doi: 10.1055/s-0042-1758755. Epub 2022 Dec 29. PMID: 36580957; PMCID: PMC9800159.
14. Beckwith NL, Khil JC, Teng J, Liow KK, Smith A, Luna J. Inappropriate Laughter and Behaviours: How, What, and Why? Case of an Adult with Undiagnosed Gelastic Seizure with Hypothalamic Hamartoma. *Hawaii J Med Public Health*. 2018 ;77(12):319-324. PMID: 30533284; PMCID: PMC6277842.
15. Scarella T, Macken MP, Gerard E, Schuele SU. Psychogenic gelastic seizures in a patient with hypothalamic hamartoma. *Epileptic Disord.* 2012 Jun;14(2):187-9. doi: 10.1684/epd. 2012.0509. PMID: 22595332.