Case Report

DOI: 10.7860/JCDR/2014/7036.4146

Cryptogenic Gelastic Epilepsy: A Pediatric Case Vignette

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ABSTRACT

Gelastic seizures, characterized by epileptic laughter, are rare and the majority is associated with hypothalamic hamartomas. We report a case with cryptogenic Gelastic seizure (without hypothalamic hamartoma), as the MRI was normal and, EEG and clinical data suggested a focal origin of the seizures.

Keywords: Child, Hypothalamic hamartoma, Laughing episodes

CASE REPORT

A 4-year-old child, growing well, started having seizures, which were atonic in nature. After three such seizures over a period of one month, the child started having two distinct type of seizures: (i) abnormal tonic-clonic movements initiated on the right half of body and then becoming generalized and followed by post-ictal drowsiness (right simple partial seizures with secondary generalization), and (ii) sudden episodes of inappropriate laughter lasting for 15-20 seconds (gelastic seizures). A CT scan of the head was normal. The initial frequency was 10-15/day (gelastic seizures) and 3-4/week for the partial seizures. The seizures remained controlled on sodium valproate 50mg/kg/day for 3½ months (May 2009 onwards), but restarted. He was referred to us for the management of the epilepsy.

He had a normal birth and perinatal history. No family history of epilepsy reported. There was no evidence of precocious puberty. Development quotient was within normal range. EEG showed abnormal awake record consistent with left hemispheric temporal origin of discharges. Video-EEG showed similar findings. MRI head with epilepsy protocol was normal. Interictal PET scan was normal.

The seizures were refractory to valproate monotherapy (maximum dose 62mg/kg/day, pre-dose serum valproate level 96 mg/dL). Sequential addition of carbamazepine (maximum 30mg/kg/d), clobazam (maximum dose of 1.5 mg/kg/day), and levetiracetam (68 mg/kg/day) did not achieve satisfactory control of gelastic seizures (3-4 seizures/day), although the partial seizures responded. Discontinuing levetiracetam and carbamazepine, followed by addition of phenobarbitone (6 mg/kg/day) led to control of seizures. At last follow-up, 17 months after complete seizure control, child was seizure-free and developmentally normal.

Interestingly, at the last follow-up, the younger male sibling (2½-year-old) also started having complex partial seizures with left temporal focus, but is well-controlled on carbamazepine monotherapy.

DISCUSSION

Gelastic seizures (GS), epileptic seizures characterized by inappropriate, stereotyped ictall aughter, are quite rare and are usually mixed with other seizure types [1]. The most frequently reported association with these seizures is the presence of hypothalamic hamartoma (HH), and is associated with a poor prognosis [2].These seizures also arise from temporal and frontal lobe tumors, and atrophic lesions; and are rarely cryptogenic [1,3-6].

Patients with GS and HH have been reported previously from India both in adults and children [7-9]; however, the other varieties have rarely been reported [10]. In most cases, the laughter lacks any sensation but occasionally a feeling of mirth may be associated with the laughter [3]. Gascon and Lombrosos [11] suggested the following criteria: stereotyped recurrence, absence of external precipitants, concomitance of other manifestations generally accepted as epileptic, presence of ictal or interictal EEG epileptiform discharges, and absence of conditions in which pathological laughter may occur. These criteria were fulfilled in our patient.

Hypothalamic hamartoma causes precocious puberty and in a few cases, it is accompanied by gelastic seizures [8]. Hypothalamic hamartoma usually presents in childhood or adolescence and without surgical intervention, the gelastic seizures may progress to other seizure types, including generalized epilepsy, and are generally refractory to antiepileptic drugs. Surgical treatment, including endoscopy and radiosurgery are the accepted treatment options [12].

Although association of GS with hypothalamic hamartoma is most common and consequently the best known, association with a temporal focus and other sites has also been reported [3]. Cases of gelastic seizures originating in the frontal lobe are uncommon and may or may not be associated with a structural lesion [5]. Temporal focus, as was the case in this child, has more frequently been reported [1,4].

Striano et al., reported a case-series of GS with nine patients seen between 1986 and 1997 [1]. Five of these patients had symptomatic localization-related epilepsy (four hypothalamic hamartoma, one tuberous sclerosis). The remaining four were labeled as 'cryptogenic' cases as the MRI was normal and, EEG and clinical data suggested a focal origin of gelastic seizures. The ictal, clinical and EEG features of the symptomatic and cryptogenic cases were comparable, with cryptogenic patients having transient drug resistance or partial response to treatment. All the cryptogenic cases had onset before 12 years (one in infancy), had a high seizure frequency and no precocious puberty or cognitive deficit. All had periodic spontaneous remissions or transient drug resistance on follow-up [1]. Some similarities with our patient were presence of a temporal focus (all four); and family history of epilepsy and response to phenobarbitone in one patient each [1]. They hypothesized that improvement in neuroimaging techniques may lead to identification of structural abnormalities in these so-called cryptogenic cases also [1].

CONCLUSION

The purpose of reporting this case is to make the pediatrician aware that gelastic seizures may also be observed in patients without a MRI lesion and with normal neurological status. Such cases are likely to be treatment-resistant. However, the majority of gelastic seizures are symptomatic in nature and a detailed neuroimaging workup of all patients is advisable.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jul 24, 2013**
Date of Peer Review: **Nov 30, 2013**
Date of Acceptance: **Dec 09, 2013**
Date of Publishing: **Mar 15, 2014**