

Gelastic Seizures In An Infant Without Hypothalamic Hamartoma: A Video Case Report

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Citation

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Abstract

Gelastic epilepsy or "laughing" seizures are most frequently associated with hypothalamic hamartomas in children and adults. We report what appears to be gelastic seizures in a 5 month old boy without hypothalamic tumor. The patient had focal hydrocephalus of the right lateral ventricle, migrational brain abnormality, septo-optic and hypothalamic dysplasia, and pan hypopituitarism. This case underscores that midline brain abnormalities other than hypothalamic tumors are associated with gelastic seizures.

INTRODUCTION

Gelastic or "laughing" seizures involve repetitive vocalization, often associated with smiling. These seizures are seen in infants, children, and adults, and are most often associated with hypothalamic hamartomas (1). There are rare reports, however, of gelastic epilepsy associated with other brain anomalies (2,3,4) and in patients with an electrographic focus but normal brain MRI (5,6). We report gelastic seizures in an infant with multiple brain anomalies other than hypothalamic hamartoma including hypothalamic dysplasia, agenesis of the corpus callosum, focal hydrocephalus, and migrational abnormalities.

CASE REPORT

The patient is a five-month-old male infant with recurrent vocalizations during wakefulness and sleep lasting 15 minutes to 2 hours. He was referred for Video EEG monitoring at Riley Hospital for Children after an exhaustive workup for these episodes including a normal 2-day video EEG performed locally, and a flexible bronchoscopy which showed normal vocal cord function at the time of the event.

Past medical history showed that the baby was born at term with a birth weight of 3.4 kg. Pregnancy was significant for prolonged rupture of membranes 35 hours prior to delivery. Persistent hypoglycemia within the first week of life prompted an endocrinology investigation which determined the baby to have panhypopituitarism due to low thyroid, cortisol, and growth hormone levels. Brain MRI showed agenesis of the corpus callosum, septo-optic and

hypothalamic dysplasia (see Figure 1). Multiple nodular gray matter intensity foci were seen in the periventricular and ventricular lining regions representing nodular heterotopias. Band heterotopias were seen in the bilateral occipital, temporal, and frontal lobes. There was focal hydrocephalus of the right lateral ventricle (see Figure 2). The baby required a ventriculo-peritoneal shunt at one month of age. He was started on phenytoin by a local pediatrician who thought the vocalizations could be seizures. Other medications included levothyroxin, hydrocortisone, ranitidine and desmopressin.

Figure 1

Figure 1: Sagittal, FLAIR brain MRI image with contrast showing agenesis of the corpus callosum and hypothalamic dysplasia.

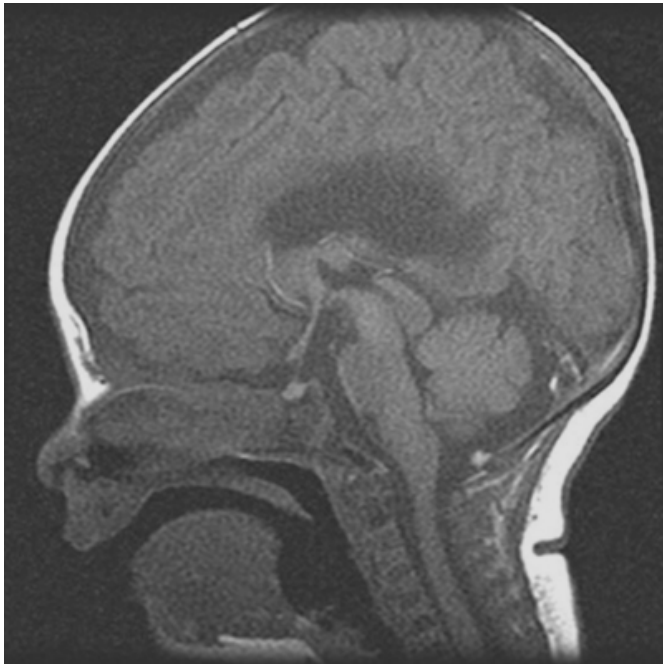
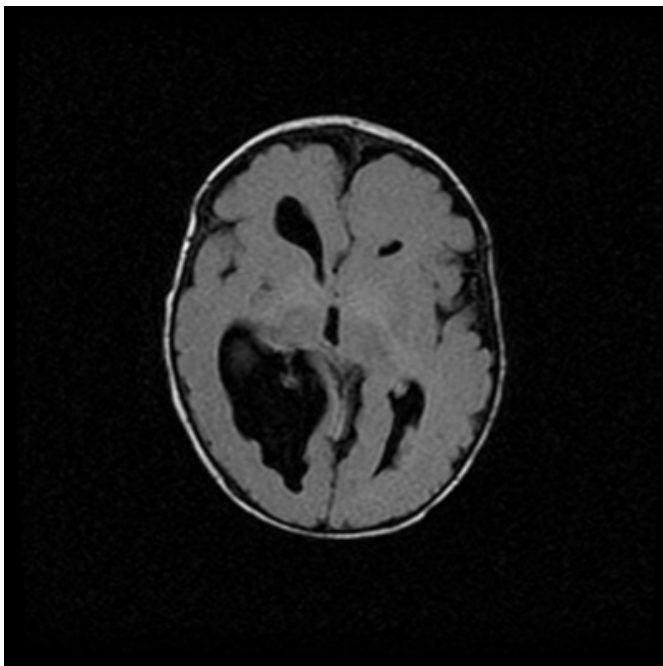


Figure 2

Figure 2: Axial, FLAIR brain MRI image with contrast showing focal hydrocephalus of the right lateral ventricle, band and nodular heterotopias.



Examination at 5 months was significant for blindness with continuous nystagmus, micropenis, and truncal hypotonia with increased tone in the extremities. The child was

significantly delayed, showing poor head control and inability to roll over.

During Video EEG monitoring, the patient engaged in repetitive vocalizations which lasted up to 8 seconds followed by 4 seconds of silence. During these episodes he bent forward at the waist and flexed his knees. These events occurred during wakefulness and sleep, lasting from 30 seconds to 30 minutes (see Video tape during drowsiness). No electrographic ictal activity was associated with these events. Otherwise, the EEG was normal during wakefulness and sleep.

Demonstration of the gelastic seizure obtained during drowsiness. The patient awakes, bends at the waist, flexes his legs and engages in vocalizations lasting approximately 8 seconds followed by 4 seconds of silence.

DISCUSSION

This case demonstrates how gelastic epilepsy can occur without the presence of hypothalamic hamartoma. We feel that the events in question represent gelastic seizures due to their stereotypic nature and because of their occurrence during wakefulness and sleep. The fact that the EEG was normal during the ictus and that there were no interictal abnormalities is not surprising. Normal electrographic patterns have been reported in young children with gelastic epilepsy whose EEGs may show progressive abnormality as the child matures (7). Also, the fact that phenytoin did not successfully treat these seizures is not surprising as they are often refractory to medication (1).

MRI showed multiple brain abnormalities in our patient: hypothalamic dysplasia, right focal hydrocephalus, agenesis of the corpus callosum, and band and nodular heterotopias. Similar anomalies have been reported to be associated with gelastic seizures such as focal obstructive hydrocephalus (2,3), focal cortical dysplasia of the cingulate gyrus (4), and multiple subependymal nodules (8). Also, our patient showed panhypopituitarism and septo-optic dysplasia, suggestive of midline dysfunction close to the hypothalamus. Phenytoin was maximized with the plan to also add Phenobarbital if the seizures persisted.

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