Late-Onset Cryptogenic Gelastic Epilepsy

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Summary

Gelolepsy or gelastic epilepsy (GE) refers to a type of epilepsy in which the epileptic seizures (ES) are „gelastic" („gelastic" being the Greek word for „laughter"). Laughing ES are characterized by stereotyped recurrence, absence of external precipitants, concomitance of other manifestations generally accepted as epilepsy, presence of interictal EEG epileptiform discharges, absence of condition in which pathological laughter might occur and response to antiepileptic drugs. Although inappropriate epileptic laughter can be so similar to the patients normal laugher that it can go without diagnosis for a long time period. GE is a rare type of epilepsy. GE can be symptomatic (typically manifesting in infants and children, more rarely in adults) or cryptogenic. There are very rare reports of late-onset cryptogenic GE. A case of 46 years-old men with gelastic ES is described. Its laughing attacks started two years ago. They have been spontaneous, unprovoked and uncontrollable. Laughing ES lasted less than one minute. Gelastic ES have been followed by coughing, flushing of the face, repeated head moving to the back and associated with altered consciousness. Family history and past history were negative. His physical, neurological and mental examinations were normal. EEG showed generalized abnormalities (sharp waves, spikes and spike-wave discharges). The CT brain scan and MRT brain scan revealed no focal abnormalities. Patient remained seizures free with treatment with Carbamazepine (Tegretol) at a dose of 1000 mg/day. Our case shows that the epileptic laughter may be only a part of a more complex seizure disorder that includes different neurovegetative and motor manifestations.

Key words: gelastic epilepsy, epileptic laughter.

Introduction

Gelastic („Gelastic" from the Greek word „gelasticos" means laughter) epilepsy (gelolepsy; epilepsie gelastique) (13) refers to a type of epilepsy in which sudden, unprovoked and unvoluntary laughter is the defining and starting epileptic manifestation. Gelastiv epileptic seizures (ES) are rare (19) and account for less than 1% of all epilepsies (3).

Epileptic laughter is stereotyped and unmotivated and is usually of short duration (less than 1 minute). Epileptic laughter may be an isolated manifestation or associated with other ES and neurological symptomatology. Consciousness can be also altered. Gelastic ES may occur at any age but they are typically manifesting in early childhood (4, 5). In young children epileptic laughter can be so similar to the normal laughter that it can be confused with behavioral or emotional disorders and this may delay the diagnosis for a long period of time. Gelastic epilepsy can be symptomatic and cryptogenic (8, 11, 24). The cases of late-onset cryptogenic gelastic epilepsy in adults are extremely rare (7, 11).

Case Report

A 46 year-old man P.S.D. was examined as out-patient at the Medical Center for Neurology and Neurosurgery of Military Medical Academy in Sofia in May 2008 (Ambul. Number 663/15.05.2008; Case history number 175/15.05.2008), because of laughter attacks. These attacks started two years ago. Laughting seizures have been stereotyped, unmotivated and involuntary. The laughter lasted less than one minute. Gelastic seizures have been followed by coughing, flushing of the face, repeated head moving to the back and have been associated with altered consciousness. Family history and past history were negative for epilepsy and other neurological disorders. His physical, neurological and mental examinations were normal. EEG showed generalized abnormalities (sharp waves, spikes and spike-wave discharges). The CT brain scan and the MRT brain scan revealed no focal abnormalities. Patients remained seizures free with treatment with Tegretol (Carbamazepine) at a dose of 1000 mg/day.

Discussion

The anatomical and physiological basis of the normal laughter is quite complex and debated. The normal laughter is probably represented in a large neuronal network where stimulation of any of its constituent units may activate the network entirely or in part (8). It is believed that normal laughter can be a result of an interaction of different brain structures such as the temporal and frontal neocortex, the limbic structures, the visual, auditorial and olfactorial areas, the hypothalamus and some structures of the brain stem. Gelastic ES of neocortical origin are confirmed by resective surgery (16). Focal brain lesions associated with gelastic ES are located in mesio-basal temporal regions (8), in fronto-mesial areas (6), the anterior part of the supplementary motor area (6, 10) as well as in the fronto-parietal regions (12). Involving of the anterior cingulate region also can elicit laughter (8). As laughing ES can be followed by loss of postural tone (atonic ES) (15) the participation of the brain stem structures can be suspected. The most common area of the brain which gives rise to gelastic ES is the hypothalamus. The most common cause is hypotalamic hamartoma (1, 4, 5, 21, 25, 26). Hamartoma is a benign growth made up of abnormal mixture of cells and tissue as a developmental abnormality of the brain (neuronal migration disorder). Gelastic ES of hypothalamic hemartomas etiology usually occur in neonatal period or early childhood (peak at 2 to 3 years). Of every 1000 children with epilepsy only one will have gelastic ES. In child with gelastic ES associated with precocious puberty (under 10 years of age) hypothalamic hamartoma may be suspected. During school-age years other epileptic seizures

type may develop and behavior problems can occur (4). Gelastic ES associated with other type tumors of the temporal or frontal lobe, papillomas of the third ventricle and neuronal migration anomalies are rare. Gelastic ES may be very rare symptoms of other brain diseases such as encephalitis, meningitis, tuberous sclerosis, disseminated sclerosis, pseudobulbar polsy, schizophrenia, lipid storage diseases etc. Very rarely no cerebral pathology can be identified in the etiology (cryptogenic gelolepsy). Even though much experiments and clinical observations have been put into understanding how gelastic ES are being generated, the relationship between an epileptic focus and gelastic ES still remains unclear.

The epileptic laughter may be associated or not with mirth (the subjective feeling of amusement). It is believed that mirth and laughter are separate functions (8). Taking care not to confuse epileptogenic lesions and the propagation of epileptic discharges which is responsible for the electro-clinical symptoms, it seems possible to distinguish broadly: (i) epileptic laughter in seizures originated in the cortex, particularly the temporal cortex; in this case the laughter expresses an emotion and (ii) epileptic laughter confined to the motor manifestations of laughter with no emotional contex originating either from hypothalamic circuits or involving lower levels (18). The anterior cingulate region is involved in the motor aspect of laughter, while the basal temporal cortex is involved in the processing of mirth (2, 7, 8, 14).

Interictal electroencephalograme (EEG) of patients experiencing laughter attacks may be normal or more usually shows focal or generated epileptic abnormalities (depending on the associated pathology). Computed tomography (CT) brain scan may be not able to reveals very small tumors. A magnetic resonance imaging (MRI) brain scan is more powerful than the CT scan (4) and should be done in every case of gelastic ES (20).

Although gelastic ES and associated other types of ES are usually pharmacoresistant (20), some gelastic ES can be well-controlled with antiepileptic medication (7, 11). The antiepileptic drugs usually used to treat gelolepsy include Carbamazepine, Oxcarbazepine, Lamotrigine, Topiramate etc. (19). It should be stressed that the effectiveness of antiepileptic treatment depends on associated brain pathology. Indications for surgical treatment of gelastic ES and their prognosis depend also on associated brain lesions (9, 17, 22).

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