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CASE REPORT

Cryptogenic gelastic epilepsy of frontal lobe origin: A paediatric case report

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Gelastic (laughing) seizures are an uncommon seizure type which in most cases has an organic cerebral pathology and specifically a hypothalamic hamartoma. The interictal EEG frequently shows focal activity. This report describes a $3\frac{1}{2}$-year-old boy who presented with episodes of unmotivated laughter associated with other epileptic symptomatology before the age of 3 years. Prolonged ambulatory EEG monitoring recorded electroclinical seizures starting in the right frontal area and spreading to the adjacent frontotemporal region. Neurological examination and brain magnetic resonance imaging were normal. Vigabatrin resulted in immediate remission of the seizures and normalization of the EEG.

Key words: ambulatory EEG; child; cryptogenic epilepsy; frontal lobe; gelastic seizure.

INTRODUCTION

Epileptic seizures with laughter (gelastic epilepsy) are rare, accounting for less than 1% of all epilepsies[1,2]. The most common cause is hypothalamic hamartoma[3,4], although it has also been seen in other diseases of the temporal and frontal lobes[1,5,6]. In particular, when the lesion substrate remains unknown, then these cases are considered cryptogenic forms[7].

Focal interictal abnormalities have been reported in surface electroencephalographic (EEG) studies, whereas during a seizure the most common sign is diffuse desynchronization of the recording; nevertheless, this examination can be normal[1,7].

The clinical, ictal and interictal EEGs (standard and ambulatory monitoring), and neuroimaging studies from a child diagnosed with cryptogenic gelastic epilepsy of a probable frontal origin are reported.

CASE REPORT

A $3\frac{1}{2}$-year-old boy was admitted with suspected epileptic seizure. There was no family history of epilepsy. The psychomotor development of the child was normal and he had no previous febrile seizures. Three days before admission, the parents noticed that the child had episodes of laughter (even roaring with laughter) without apparent reason, twisting the trunk with flexion or extension of the limbs, and saying words like 'horse', 'it hurts', 'come on'. These episodes, which were repeated frequently during the day, lasted for several seconds, after that the child resumed his normal activity. Nine months before, for 1 or 2 weeks he had suffered very similar episodes consisting of intermittent laughter followed by 'bicycling' movements; physical and neurological evaluation, and a routine EEG were normal.

On admission, examination revealed no abnormal findings; however, the EEG study (drowsiness and light sleep) showed bursts of spikes and sharp waves ranging in amplitude from 30 to 100 μV on the right temporal region (Fig. 1). On the following day, a 24-hour ambulatory EEG monitoring (A-EEG, Oxford System) was performed in which 15 electroclinical seizures were recorded, all of them showing unmotivated laughter with no apparent emotional state of mirth; the limbs adopted different tonic posturing in each seizure, with either flexion or extension in upper or lower limbs and even sometimes unilaterally mainly affecting the right half of the body. Simultaneously, there were bursts of spikes, sharp, and slow

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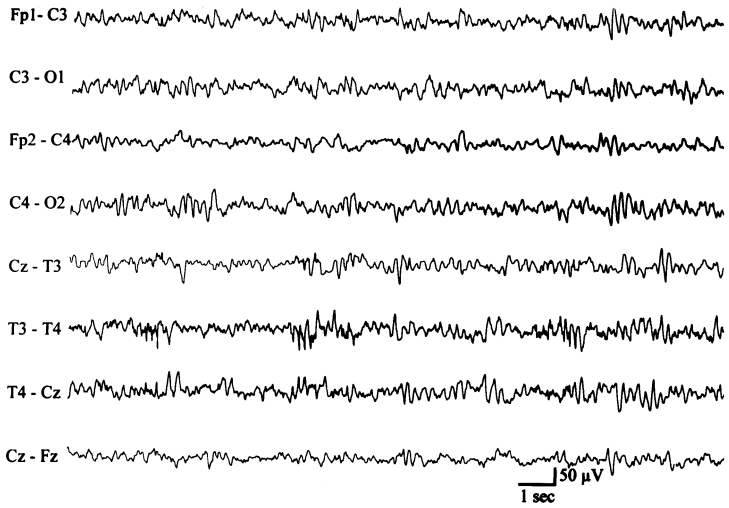

Fig. 1: Interictal EEG showing focal spikes and sharp wave discharges on the right temporal area (T4). Normal background activity during drowsiness.

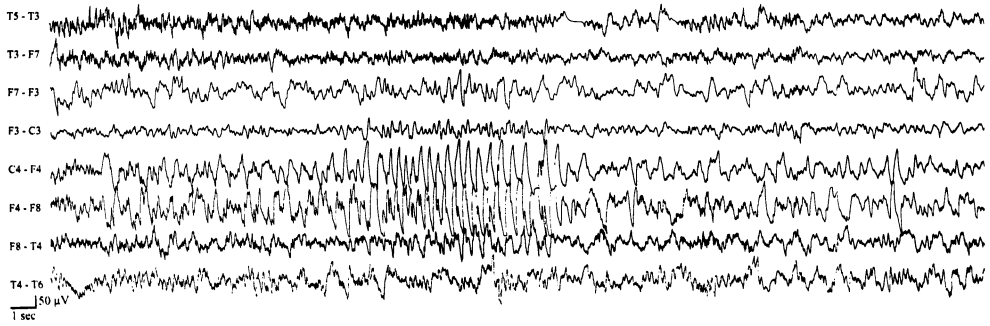

Fig. 2: Ambulatory EEG during a gelastic seizure. This figure shows repetitive sharp and slow waves of increased amplitude initially prominent on the right frontal area (F4) spreading to adjacent frontotemporal leads followed by slowing in these regions.

waves of increasing voltage in the right frontal area spreading to the adjacent frontotemporal region lasting from 10 to 30 seconds; the episodes ended with a transient slowing down in these leads (Fig. 2). Between seizures there were abnormalities such as those described in the standard EEG and the bioelectrical background activity was normal. A magnetic resonance imaging (MRI) did not detect brain abnormalities (sequences SE-T_1, EG-T_2 and FSE-T_2 weighted imaging, by using 4-mm-thick sections in the axial, coronal, and sagittal planes). The seizures were completely controlled with vigabatrin (500 mg/day). Regular EEG recordings (awake and asleep) during 3 years of follow-up invariably showed normal results.

DISCUSSION

Laughter is usually a physiological act in which the limbic system and the brain stem are involved. However, it can be an abnormal sign, particularly in the following conditions: pseudobulbar palsy, emotional lability, behavioural disinhibition and gelastic seizures[1, 5]. Our patient presented the diagnostic criteria for gelastic epilepsy; that is, recurrent attacks of laughter with no apparent reason, interictal and/or ictal EEG abnormalities, and no neurological signs suggesting any other type of pathological laughter[5]. In gelastic epilepsy, attacks of laughter may be the only sign, or they can be associated with other types of partial or generalized seizures; a syndrome consisting of gelastic seizures; precocious puberty and behavioural problems whose anatomical substrate is hypothalamic hamartoma have even been suggested[3].

The interictal EEG findings in the scalp consist of focal abnormalities in temporal[1–4, 8–11], frontal[6, 11], or parasagittal regions[5]. Focal discharges have also been reported in these areas during a seizure[1, 5, 6, 11] but the most common signs are diffuse changes such as volt-

age attenuations, slowing and even generalized spike-wave paroxysms[3, 8, 10–12]. In the symptomatic forms, EEG studies with deep electrodes have shown that the onset of the ictal paroxysmal activity corresponds to the site of the lesion, although in the surface EEG a simultaneous diffuse change is seen[10]. In the case reported here, the EEG abnormality was always focal, interictal in the temporal area, and ictal in the frontal region spreading to the frontotemporal area. Although focal EEG abnormalities in gelastic epilepsy have been reported more commonly in the left than in the right hemisphere[1, 2], in our case the EEG changes were always located in the right hemisphere.

The clinical signs in our patient consisted not only of unmotivated laughter, but also of other motor actions and brief vocalizations. It should be noted that language automatisms, although they are common in temporal and frontal epilepsies, have no lateralizing value[8, 13]. The motor component of laughter is also part of the possible clinical manifestations of frontal lobe epilepsies[13–15]. We cannot tell for sure whether the pathological laughter or our patient was associated with a feeling of mirth, which would imply the involvement of the basal temporal cortex[5]. However, the predominance of motor events over those with an affective or psychic appearance, the shortness of the episodes, their high frequency, the low or absent postictal confusion, and the lack of a history of febrile seizures point to a frontal rather than a temporal origin of seizures[5, 13, 15–18]. On the other hand, interictal EEG changes in frontal lobe epilepsies may be noticed on temporal areas of the scalp[15, 17–19]. The vocalization, bicycling movements and postures of our patient seem to suggest involvement of mesial areas of the frontal lobe[14, 15, 17], but we were unable to confirm this hypothesis because the EEG recordings were obtained on the scalp.

The most common cause of symptomatic gelastic epilepsies is hypothalamic hamartoma, which as ectopic grey matter has an intrinsic paroxysmal activity[10]; on spreading to temporal, frontal, and diencephalic structures this epileptogenic activity would trigger other clinical signs[11, 20]. In our case this condition is ruled out by the normal brain MRI, which is also consistent with the absence of other clinical signs usually associated with hypothalamic hamartoma and with good pharmacological control of seizures[3–5, 7, 9, 11]. Moreover, the ictal EEG abnormalities in our case were focal, unlike most diencephalic gelastic seizures in which ictal EEG abnormalities are diffuse[3, 5]. Some other cases of gelastic epilepsies of a frontal origin have been reported, but unlike our case, their brain MRI showed some type of lesion[5, 6].

In our opinion, the existence of gelastic seizures should be considered even though brain MRI shows no evidence of structural lesion. A-EEG monitoring is highly useful for the diagnosis. The case reported here is a paediatric cryptogenic gelastic epilepsy whose clinical and EEG findings were suggestive of epilepsy of a frontal origin. Invasive EEG studies would be required for accurate recording of the onset of seizures[21], but the good clinical response to anticonvulsant treatment and the disappearance of EEG abnormalities advised against such tests.

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