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Case Report

Gelastic epilepsy: Beyond hypothalamic hamartomas

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ABSTRACT

Gelastic epilepsy or laughing seizures have been historically related to children with hypothalamic hamartomas. We report three adult patients who had gelastic epilepsy, defined as the presence of seizures with a prominent laugh component, including brain imaging, surface/invasive electroencephalography, positron emission tomography, and medical/surgical outcomes. None of the patients had hamartoma or other hypothalamic lesion. Two patients were classified as having refractory epilepsy (one had biopsy-proven neurocysticercosis and the other one hippocampal sclerosis and temporal cortical dysplasia). The third patient had no lesion on MRI and had complete control with carbamazepine. Both lesional patients underwent resective surgery, one with complete seizure control and the other one with poor outcome. Although hypothalamic hamartomas should always be ruled out in patients with gelastic epilepsy, laughing seizures can also arise from frontal and temporal lobe foci, which can be surgically removed. In addition, we present the first case of gelastic epilepsy due to neurocysticercosis.

1. Introduction

The term "gelastic epilepsy" derives from the Greek word *gelos* (laughter) and was introduced in 1957 by Daly and Mulder. It is used to name seizures characterized by sudden laughter attacks, out of social context, without any particular emotion like joy or happiness [1]. In 1971, Gascon and Lombroso suggested as diagnostic criteria the presence of stereotyped laughter episodes in the absence of external triggers that can be associated with other epileptic manifestations, ictal or interictal discharges on the electroencephalogram (EEG), and absence of other conditions that could explain the pathologic laughter [2]. The vast majority of gelastic epilepsy series reported include only children, and they have found an association with hypothalamic hamartomas, a lesion that therefore must be always ruled out in these patients. However, other lesions and localizations have also been reported, mainly in other adult patients [3,4,5]. Here, we describe three patients with gelastic seizures (GS) with no evidence of hypothalamic hamartomas.

1.1. Case 1

A 35-year-old man with a history of neurocysticercosis was treated with antiparasitic drugs as a teenager. Since the age of 29 years, he had almost daily spells characterized by sudden unmotivated laughter attacks which were described by his relatives since the patient was not aware. In addition, he had occasional seizures which started with forced head version towards the right, followed by right upper and lower limb jerks, and finally secondary generalization. He received carbamazepine, topiramate, and levetiracetam with poor response. Magnetic resonance imaging (MRI) showed upper right and inferior left mesial frontal lobe nodular lesions, consistent with calcified neurocysticercosis (Fig. 1A). Five GS were recorded on scalp video-EEG monitoring, all of them showing ictal patterns arising from the left anterior temporal lobe (Fig. 1B). Positron emission tomography (PET) was performed, showing left mesial frontal and anterior temporal hypometabolism (Fig. 1C). Neuropsychological evaluation showed bilateral frontotemporal alterations with left hemisphere predominance. Invasive recordings were performed with left frontal deep electrodes and foramen ovale electrodes, and ictal and interictal activities arising from the left mesial frontal region was demonstrated (Fig. 1D). Resection of the left frontal pole, which included the lesion, was performed (Fig. 1E), and the biopsy showed cysticercosis. The patient has been seizure-free since the surgery (5 years of follow-up) and under no medication for 3 years (ILAE class I).

1.2. Case 2

A 45-year-old man with epilepsy since the age of 14 years had seizures characterized by sudden laughter attacks, associated with feelings of joy and happiness, which lasted about 30 s, occasionally

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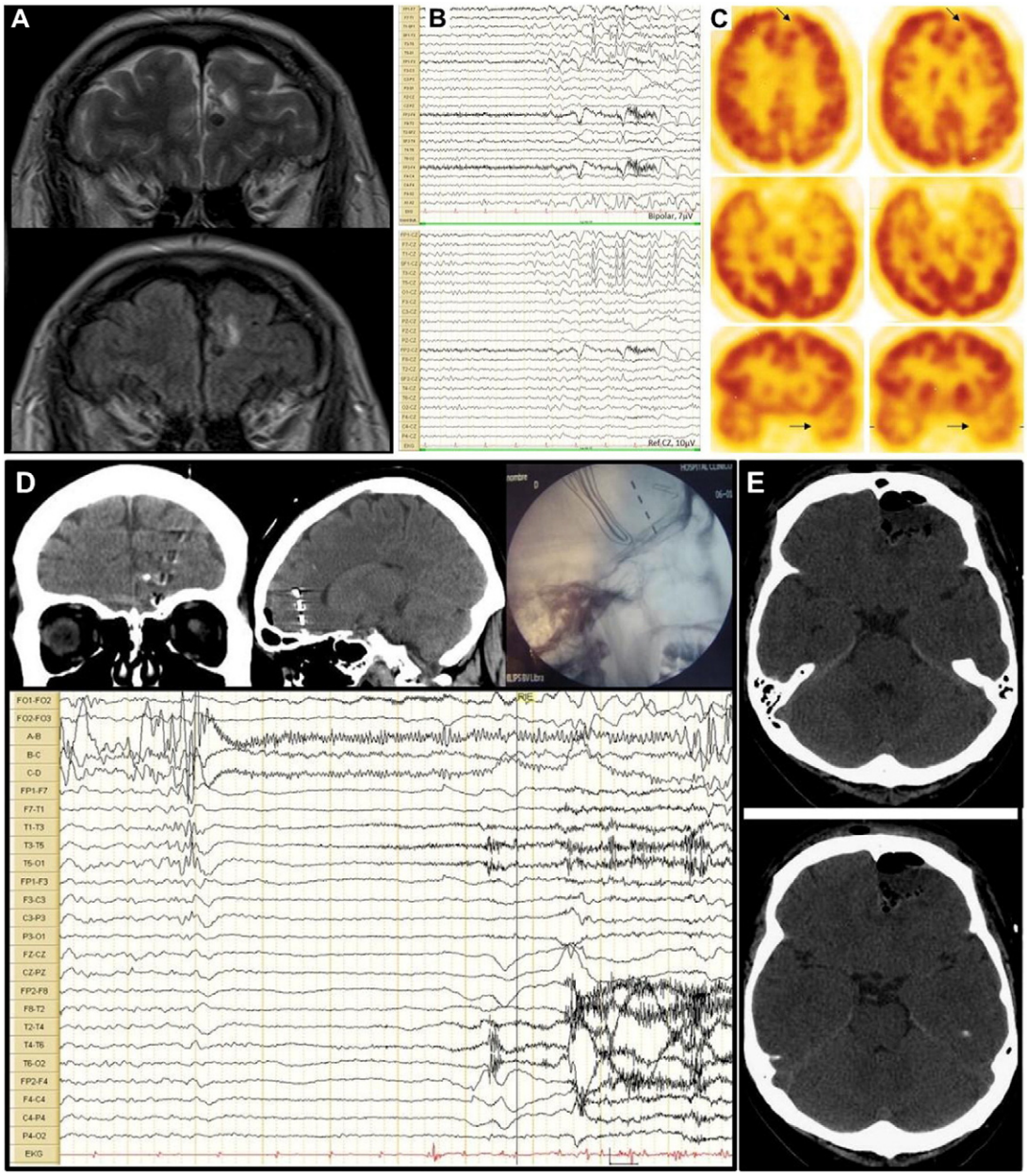

Fig. 1. MRI, scalp EEG, PET and invasive EEG monitoring of case 1. A. MRI T2-weighted and FLAIR showing neurocysticercosis lesions in both frontal lobes. B. Surface EEG recording showing left anterior temporal onset of gelastic seizure on bipolar montage and referenced to Cz. C. FDG-PET showing left mesial frontal and temporal hypometabolism (arrows). D. Depth electrode EEG recording showing left mesial frontal onset of the gelastic seizure (electrodes A–D vs electrodes on both foramen ovale FO1–2). E. Head CT showing left frontal pole resection.

associated with disconnection followed by one to two minutes of postictal confusion. He had up to 5 episodes per day in spite of multiple antiepileptic treatments (carbamazepine, phenobarbital, levetiracetam, and topiramate). His brain MRI showed left mesial temporal sclerosis (Fig. 2A); surface EEG recordings showed left frontotemporal interictal epileptiform discharges (Fig. 2B); two gelastic seizures with left frontotemporal ictal onset were recorded. Fluorodeoxyglucose Positron Emission Tomography showed left anteromesial temporal hypometabolism (Fig. 2C). Neuropsychological evaluation showed bilateral mesial and anterior temporal alterations with predominance of the left hemisphere. Left amygdalohippocampectomy was performed (Fig. 2D), and the biopsy showed hippocampal sclerosis associated with cortical dysplasia. The patient remained seizure-free for a year, and seizures relapsed afterwards, with a 50% reduction compared to presurgical average (ILAE class IV).

1.3. Case 3

A 42-year-old man had no relevant medical history. At 15 years old, he experienced brief laughter attacks, apparently unmotivated, that was perceived as strange behavior by his family. Gelastic epilepsy was diagnosed, and the patient was placed on carbamazepine; the treatment was stopped after a seizure-free period of 6 years. Five years after the medication withdrawal, he began again with episodes of unmotivated laughter, with no emotional correlate, associated with disconnection. Routine EEGs and brain MRI were normal. Carbamazepine was restarted