Epilepsy & Behavior Case Reports 8 (2017) 27–30

Contents lists available at ScienceDirect

Epilepsy & Behavior Case Reports

journal homepage: www.elsevier.com/locate/ebcr

Case Report

Localization of ictal pouting in frontal lobe epilepsy: A case report

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1. Introduction

Changes in facial expression in frontal lobe epilepsy (FLE) have been described in the literature [1,2]. However, this discrete feature of ictal semiology is often missed because of more striking events such as hypermotor or bizarre behaviors. One such facial feature, characterized by a turned-down mouth, is traditionally described in French as the "chapeau de gendarme," referring to the shape of the French policeman's hat during the reign of Napoleon. Ictal pouting (IP), which mimics expressions of fear, displeasure, or disgust, appears frequently during focal seizures. IP is described as an inverted smile. It consists of a turned-down mouth with puckering of the lips and symmetrical sustained (>5 s) lowering of the labial commissures, commonly accompanied by contraction of the chin. This very interesting sign of FLE is related to the involvement of the mesial frontal areas, particularly the anterior cingulate cortex (ACC) [3]. We report a case whose ictal semiology consisted only of pouting.

Brain magnetic resonance imaging (MRI) studies revealed that focal cortical dysplasia (FCD) in the right mesial frontal lobe and ACC and tissue histopathology confirmed FCD type IIB.

2. Case report

A 24-year-old woman was admitted to our video-EEG monitoring (VEM) unit for evaluation of seizures that began at the age of 5 years and occurred 15–20 times a day. According to her description, she had preserved conscious and remembered what happened during seizures. Her seizures began with blurred vision, followed by tearful appearance, and ended with a minor contraction of the chin. If these events occurred while she was walking, she was able to continue, although her motion was slower. She also had palpitations, a flushed face, and hot flashes during seizures. Her interictal neurological examination and neurocognitive functions were normal. There was no history of behavioral or psychotic disorders and no family history of epilepsy. She had been on 200 mg lamotrigine daily after her previous antiseizure drugs, valproate and primidone, were discontinued due to inefficacy.

During VEM, she had 11 stereotypical seizures. She stared at the beginning of her seizure for 2–4 s to mimic an expression of sadness. It appeared like an inverted smile, the mouth was turned down with puckering of her lips that was symmetrical and sustained for 10–20 s. Lowering of the labial corners at the very end accompanied by contraction of the chin (Fig. 1, Video 1). She was able to communicate during seizures. The EEG background activity suddenly changed at the onset of seizures, with diffuse attenuation and subsequent rhythmic delta activity bilaterally before changing to spike-and-waves that were dominant over the right hemisphere during the last 3–4 s (Figs. 2, 3). Before and during seizures, heart rate on the ECG increased from 72 beats per minute to 96 beats per minute respectively. The seizures lasted for 15–25 s. All the seizures were similar, with the only ictal semiology being IP. Brain MRI revealed focal cortical dysplasia on the right mesial frontal lobe and ACC (Fig. 4).

After VEM, the patient was considered a favorable candidate for epilepsy surgery and was referred for further evaluation; however, she declined surgery at that time. Topiramate was initiated as an add-on therapy at 200 mg/day and subsequently discontinued due to cognitive side effects. Carbamazepine and levetiracetam were initiated as add-on therapy at 600 and 1000 mg/day, respectively. Despite treatment with 200 mg/day lamotrigine, 600 mg/day carbamazepine, and 1000 mg/day levetiracetam, the patient continued to have seizures 5–10 times per day. After participating in multiple antiseizure drug trials that did not affect her seizures, she decided to undergo epilepsy surgery. Pathological examination revealed type II focal cortical dysplasia with balloon cell. After surgery without any complications, she has remained seizure free for 9 months (Engel Class I) and remains under antiseizure drug treatment [4].

3. Discussion

The ictal semiology of this case consisted of pouting with contraction of the chin at the end. There were no other semiological features that showed seizure propagation. Souirti et al. investigated 11 patients with IP; the epileptogenic zone was localized to ACC (n = 4), the orbito-frontal region (n = 2), the mesial prefrontal, premotor cortex (n = 3), the supplementary motor area (n = 1), and the inferior frontal gyrus (n = 1). All the patients had neurovegetative symptoms with pouting, consistent with the symptoms observed in our patient. In addition to IP, the ictal semiology of these patients included vocalization, head and eye deviation, tonic posturing, agitation, and hyperkinetic movements, which were not observed in our patient [3]. Tan et al. presented a patient with ictal pouting with an epileptogenic zone localized to the left frontal operculum extending to the insula. In this case, the

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http://dx.doi.org/10.1016/j.ebcr.2017.04.003

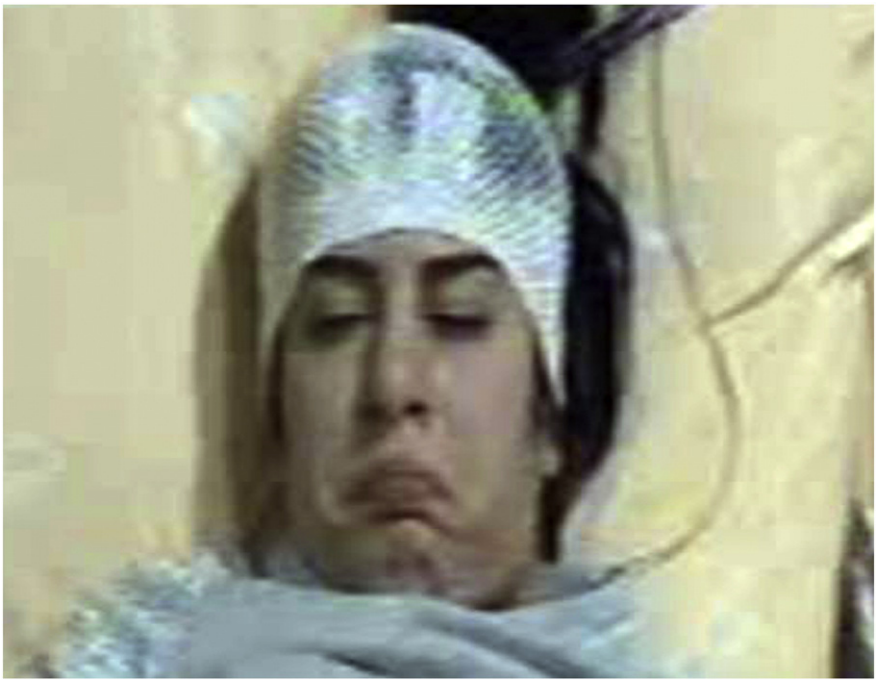

Fig. 1. Facial expression of the patient with ictal pouting and distress. This seizure pattern ends with contraction of the chin.

semiology consisted of right gaze deviation and tonic extension of the right arm, followed by the left arm. Despite remaining conscious, our patient could not speak during seizures. This may be explained because her lesion was in the right hemisphere, while the other patient's was on the left side [5]. Leitinger et al. reported a patient with ictal pouting as an early component of seizure without ictal EEG changes in the intracranial frontomesial electrodes. However, Chassoux commented that these results may be due to an insufficient sampling area [6,7]. In our patient, IP was not an early sign of semiology and it presumed no spreading to other areas of the frontal lobe. After resection of the focal cortical

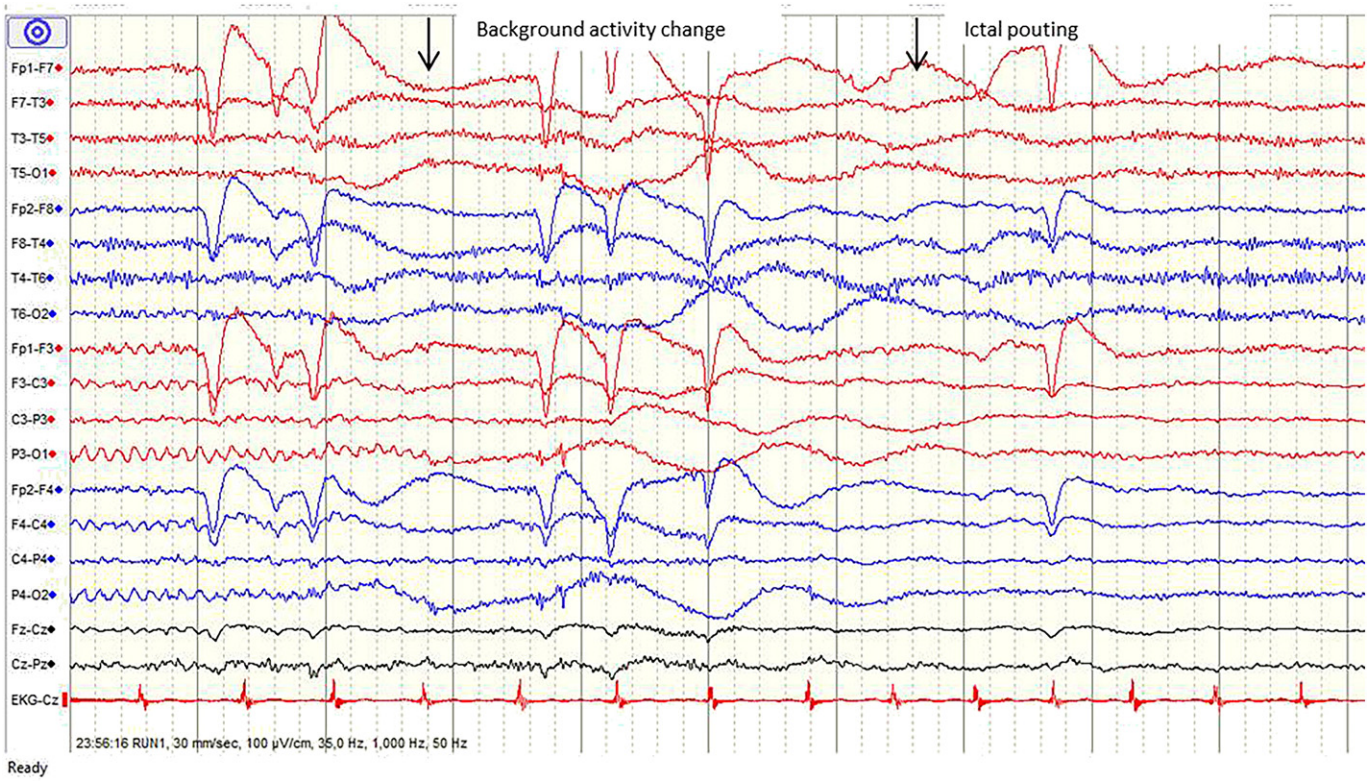

Fig. 2. Fifth seizure; the first arrow marks an abrupt attenuation change of background activity with ictal EEG onset, and the second arrow indicates the beginning of ictal pouting and clinical seizure onset.