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Case Report

A Dancing and Rhythmic Epilepsy. A SEEG Case Report

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ABSTRACT

Background: Recognizing ictal semiology is essential to identify the epileptogenic zone, particularly in refractory epilepsy where its correct identification is what determines the surgical prognosis. Dancing is a very unusual ictal semiology and its underlying neural networks remain somehow unclear since both the temporal and the frontal lobe have been implicated in its genesis.

Methods: We present a 21-year-old with refractory epilepsy characterized by dancing and rhythmic seizures. Noninvasive presurgical evaluation localized the epileptogenic zone within a gross lesion localized in the left frontal lobe. A SEEG allowed us to localize the ictal onset zone at mesial middle, inferior and orbito-frontal cortex with rapid propagation of ictal activity backward and laterally to the precentral regions. A complete surgical resection was proposed as surgical treatment.

Conclusions: Ictal dancing is an atypical presentation of seizures arising from the frontal lobe. Its correct identification allows us to plan invasive studies and to localize the seizure onset zone. Homemade videos are very useful in the evaluation of patients particularly those with atypical presentations. More intracranial recordings are needed to fully comprehend the underlying networks and interactions of cerebral areas during dancing seizures.

INTRODUCTION: CASE PRESENTATION

It is broadly accepted that the semiology of frontal lobe seizures is difficult to characterize, especially those arising from anterior frontal regions. The connectivity of frontal lobe supramodal associative areas supports spread through distant cortico-cortical efferent pathways, which can be both multilobar and multidirectional, typically resulting in rapid, widespread propagation of seizure discharges originating in frontal regions. Here we presented a case of frontal refractory epilepsy with an unusual semiology. AL, is a 21 years-old male, left-handed with no significant past medical or family history. He had his epilepsy onset at 14 years old. Initially, seizures presented during sleep or at awakening and were generalized tonic-clonic seizures. The seizures started to be different at AL´s 17 years old, described by his family and friends as a dance-like routine with rhythmic movements of the pelvis, the trunk and the upper limbs. Homemade recordings requested to the family showed an organized, naturalistic movement strongly reminiscent of a complex dance routine even including a jump spinning and vocal production of a rhythmic beat box-like melody during the

seizures (Figure 1). It is interesting to note that the patient never received formal dance or music training, nor did he show any particular interest in it.

Figure 1: Sequential screen captures of homemade video of a seizure.

Seizure's frequency was 1-2 per day and progressively increasing in frequency over time until reaching 9-10 per day at 19 years old. A Video Electroencephalogram (VEEG) showed awareness impairment followed by an asymmetrical tonic posture of the right arm, a dance-like integrated gestural motor behavior with distal hand stereotypies, a fixed facial expression with rhythmic sound production with the mouth and lips. Seizures usually lasted less than 15 seconds and he had no postictal symptoms. Since the VEEG recordings were obtained with the patient in bed, semiology was less florid as the home recordings in which the patient was standing. Electrical findings showed a low amplitude, fast frequency rhythmic activity on the left frontocentral area. Routine laboratory tests were negative.

MRI evidenced a widespread area of grey-white matter junction blurring in the anterior left frontal lobe, involving the left frontal pole, middle and inferior frontal gyrus and pre-motor cortex. FLAIR hyperintensity was present at the posterior third of the superior frontal sulcus (Figure 2). Functional MRI of the motor function of both hands showed activation in the precentral motor primary cortex bilaterally. On the left side, it appears to be separated from the lesion by the precentral sulcus. The motor activity of the mouth showed the usual codominance with activations in the caudal precentral cortex bilaterally. On the left side, it appears to be in contact with the upper limit of the lesion. As for the language, fMRI shows dominance exclusively on the right hemisphere for both comprehension and lexical tasks.

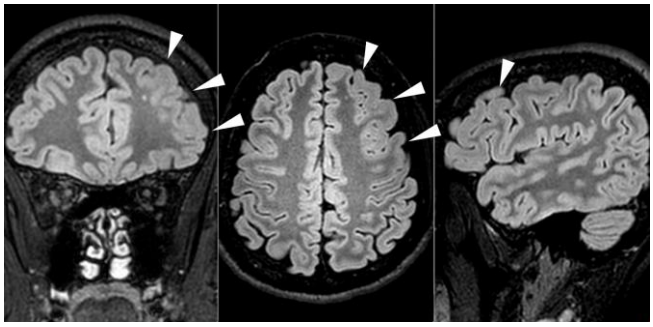

Figure 2: Magnetic Resonance Images (MRI) acquired on a 3 T scanner, FLAIR sequence, of the brain viewed from the side coronal (left), axial (central) and sagittal (right). White arrows indicated an area of hyperintensity localized at the posterior third of the superior frontal sulcus. FLAIR=fluid-attenuated inversion recovery.

A Stereo-Electroencephalogram (SEEG) was decided to evaluate the extension of the epileptogenic zone within the lesion and to delineate its limits in order to establish a surgical strategy. A left sided SEEG was performed with 9 electrodes exploring the rostral prefrontal ventrolateral regions and the rostral cingulate gyrus. The patient presented 38 short lasting seizures similar to the ones seen on the VEEG with a clear predominance during sleep. Interictal electrical recordings showed diffuse high amplitude spikes and slow waves with the involvement almost all of the implanted electrodes. The ictal onset was located at the mesial middle, inferior and orbito-frontal cortex with rapid propagation of ictal activity backward and laterally to the precentral regions (Figure 3). In light of the SEEG findings, it was decided to perform a complete resection of the lesion. The surgery will be performed in the next few months.

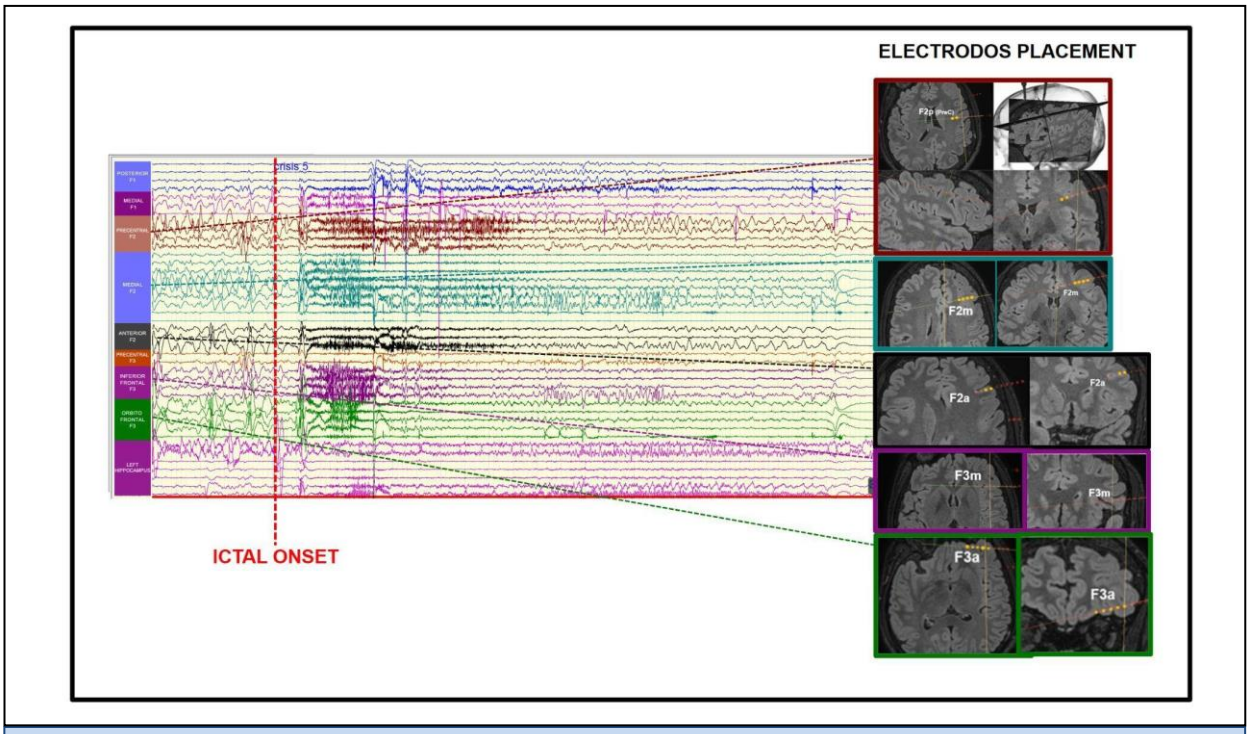

Figure 3: SEEG. The vertical red line indicates the ictal onsetlocalized at the mesial middle, inferior and orbito-frontal cortex with rapid propagation to the precentral regions. Each tracing colorcorresponds to a different electrode. The localization of each electrode is indicated on the left side of the image. (F1: first frontal gyrus, F2: second frontal gyrus, F3: third frontal gyrus). Onthe right side are the images obtained by fusion of images from pre implantation MRI and post implantation tomography, showing electrodes placement.

DISCUSSION

Ictal dancing is a very unique and striking seizure semiology. Its integrated and naturalistic nature challenges classical conceptions regarding seizure semiology particularly in hiperkinetic seizures. A few cases have been previously reported in humans [1-4], and none in animals 12. In humans, only two cases had intracranial ictal electroencephalography available, one electrocorticography and one SEEG [1,5]. The underlying neural networks of this semiology remains somehow unclear since both the temporal and the frontal lobe have been implicated in its genesis. The dorsolateral prefrontal cortex seems to be the most likely area involved particularly in patients with ictal dancing and singing such as our patient [5]. The frontal lobe is the largest lobe in the brain accounting for about 40% of the cerebral cortex. The organization of the frontal cortex, which is predominantly made up of heteromodal association areas, is extremely complicated and incompletely understood, particularly in the prefrontal region. Seizures arising from the frontal lobe are particularly prone to misdiagnosis due to their sometimes bizarre or atypical appearance, as well as to the fact that surface EEG does not necessarily show interictal or ictal abnormalities especially those arising from anterior frontal regions [6]. It is widely accepted that semiology and electrical patterns of frontal lobe seizures are difficult to characterize, and can be misleading in predicting the seizure onset localization[7] and even be confused as Psychogenic Nonepileptic Seizures (PNES). Nevertheless, in a given patient with frontal lobe epilepsy, seizures are generally similar and, if present, with stable and reproducible electroencephalographic findings [7,8]. The localizing value of specific semiological features is, in general, less well-understood in Frontal Lobe Epilepsy (FLE), compared with other localizations such as Temporal Lobe Epilepsy (TLE).

General characteristics of frontal seizures are briefness with sudden onset and termination, often arising during sleep, may occur in clusters, have a tendency to rapid secondary

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generalization and produce minimal postictal confusion [9,10]. A classification of semiology of FLE has not yet been reached. While the recent anatomical and functional categorization of frontal seizure subtypes proposed by Chauvel and colleagues [11] remains preliminary, it provides a useful way to consider the localizing value of certain patterns of the seizure semiology. Seizures arising from the anterior lateral and medial prefrontal regions are usually complex, such as semirhythmic tapping of the hands or feet, or grasping motions. They are often associated with asymmetrical tonic or dystonic posturing of upper and/or lower limbs. Vocalization may also occur. In our patient, the complex motor behavior is characterized by a fixed facial expression with sound production, integrated gestural motor behavior characterized by rocking, jumping and turning in on itself with distal stereotypes and rhythmic movements of the pelvis and trunk; a dancing seizure certainly. Early spread networks underlying these clinical manifestations according to Chauvel and colleagues [11] involved rostral prefrontal ventrolateral regions (Brodmann areas 47/12, 10, 11 and 46) and the rostral cingulate gyrus (Brodmann areas 32 and rostral 24). This semiology is described in patients who presented systematic co-involvement of ventrolateral prefrontal cortex and the anterior cingulate area, either simultaneously at seizure onset or by propagation in a lateromedial direction. In our case, the decision about where to place the electrodes in the SEEG was based on the hypotheses regarding the likely brain structures involved in the Epileptic Zone (EZ). These hypotheses were formed by the epilepsy team and were based on the ensemble of all the available non-invasive data. With the information obtained in the SEEG, we were able to identify the epileptogenic zone on the 1st and 2nd circumvolutions of the dorso lateral frontal lobe cortex in accordance with what has been published so far [12].

CONCLUSION

Bizarre or atypical seizures such as the one presented by our patient offer an interesting starting point for the analysis of the areas involved in seizure`s semiology. In patients with atypical clinical presentations, focusing the initial study on the atypical manifestation rather than on the possible affected lobe may be useful for defining the possible epileptogenic zone. It is also important to highlight the importance homemade videos can have, providing vital information of the semiology that we could not obtain during the realization of a VEEG due to the fact that the patient usually stays lying in bed during the study and this can limit the range of movements patients can have during a seizure. Frontal lobe is the biggest lobe of the brain and its complexity allows the occurrence of diverse seizure`s semiology. It is very important to closely describe the different semiological aspects of the seizure in order to extract the most information as possible. Although there is not a universally accepted classification for frontal seizures, Chauvel and colleagues [11] offered a useful description and localization value of different clinical manifestations of frontal lobe epilepsy. It was very helpful for us when we had to plan the SEEG. The underlying networks and interactions of cerebral areas during dancing seizures remains to be completely elucidated, more intracranial recordings are needed to fully comprehend it.

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