Seizure: European Journal of Epilepsy 67 (2019) 71–72

Contents lists available at ScienceDirect

Seizure: European Journal of Epilepsy

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

Clinical letter

Disappearance of symptomatic generalized 3-Hz discharges after focal surgery in a patient with tuberous sclerosis

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1. Introduction

Patients with tuberous sclerosis complex (TSC) usually present with epileptic spams and/or focal seizures and can be good candidates for epilepsy surgery [1]. Typical absence seizures (TAS) are generalized with sudden onset and termination, lasting a few seconds, and electrographically defined by a unique EEG signature: diffuse, regular, > 2.5-Hz spike-and-wave discharges (SWD). Some data suggest a focal origin for these generalized SWD, in particular, from themesial or polar aspects of the frontal lobe.

We report the case of a TSC patient who presented with epileptic spasms (ES) and generalized 3-Hz SWD accompanied by slow reading, in whom a frontal tuber resection resulted in the disappearance of both seizure types.

2. Case report

This 13-year-old, right-handed girl with TSC and *TSC2* mutation suffered with infantile spasms at six months, which responded to vigabatrin. Drug-resistant seizures started at the age of eight, characterized by daily clusters of tonic spasms on awakening and frequent "absence-like" seizures with loss of contact, lasting a few seconds. Interictal electroencephalogram (EEG) revealed right frontal slow waves and spikes, maximum at F4, as well as an independent spike focus at P4. The ictal phase of the two seizure types was recorded by video-EEG. The first seizure type was characterized by 3-Hz generalized SWD lasting from 4 to 10 s and accompanied by slow reading (Fig. 1A). The second seizure type was a cluster of bilateral tonic contractions of the upper arms and slight head deviation to the right, with an arrest of the interictal slow waves and spikes, followed by a diffuse rapid activity and the appearance of a pattern of spasms with periodic diffuse high-amplitude slow waves. MRI demonstrated bilateral tubers in the frontal, posterior temporal, and parietal regions. The biggest tubers were right-sided with a clump of tubers in the right fronto-polar region, extending to the lateral prefrontal region, and a large tuber in the right parietal region, consistent with interictal EEG findings. A stereo-EEG (SEEG) investigation was performed with 12 electrodes implanted orthogonally, eight in the right hemisphere (Fig. 1C) and four in the left hemisphere. Interictal activity revealed a significant spike and wave activity in the right fronto-polar tuber, sometimes extending to the lateral prefrontal part of the clump of tubers. An independent permanent spiking activity was observed in the right parietal tuber. Both seizure types were recorded. TAS consisted of 3-Hz SWD observed at all recording leads (right and left), except those used to explore the right parietal tuber which continued to display an independent permanent spiking activity (Fig. 1B). Clusters of spasms corresponded to a series of fast periodic activity beginning within the clump of tubers in the fronto-polar and fronto-lateral regions, followed by progressively more widespread discharges among the right and then bilateral contacts as the clinical spasms occurred.

The large clump of tubers in the right fronto-polar and lateral prefrontal cortex were resected (Fig. 1C). The patient has been seizure-free since then with Engel Class Ia at 24 months of follow up. Antiepileptic medication has been reduced from three to one drug. A 24-hour scalp EEG performed three and 18 months after surgery confirmed the disappearance of spasms and diffuse 3-Hz SWD. Only slow waves and spikes were recorded at P4.

3. Discussion

We report a TSC patient who presented with clusters of ES and independent generalized 3-Hz SWD accompanied by slow reading, in whom the resection of a mass of right fronto-polar and lateral prefrontal tubers resulted in the disappearance of both types of seizure.

TSC patients presenting with epileptic spams and/or focal seizures can be good candidates for epilepsy surgery [1]. In some patients, Invasive EEG investigation may be necessary to determine the epileptic tuber to be respected. Based on the presence of bifocal interictal abnormalities in the right hemisphere (frontal and parietal) and bilateral frontal, parietal, and temporal tubers, bilateral SEEG implantation was performed, which demonstrated that ES originated within the mass of right frontal tubers. 3-Hz SWD was recorded at all leads (right and left) with slow reading, except those in the right parietal tuber (electrode G; contacts 10–15), possibly due to the fact that the latter was

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https://doi.org/10.1016/j.seizure.2019.02.003
Received 16 December 2018; Received in revised form 4 February 2019; Accepted 5 February 2019

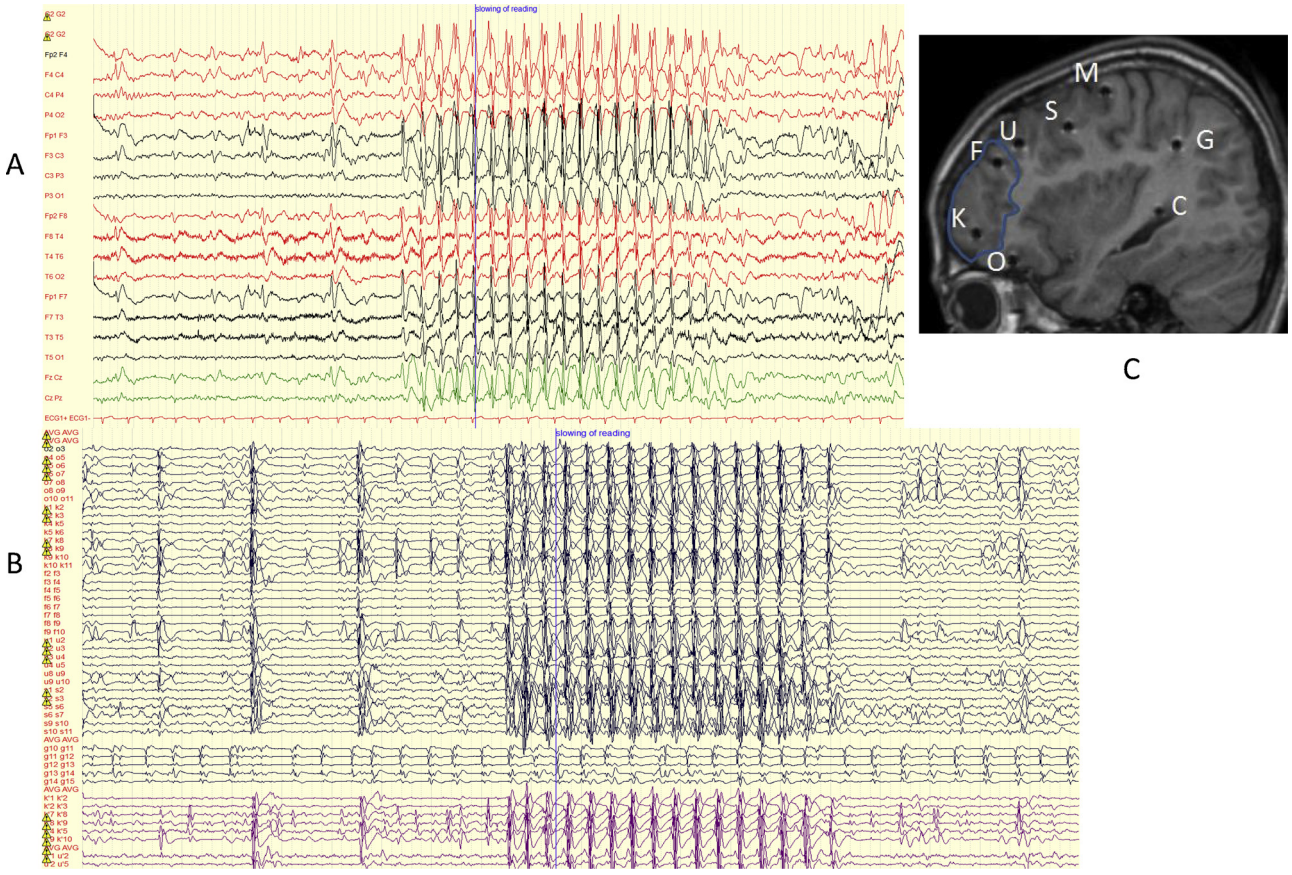

Fig. 1. A: 3Hz diffuse spike and wave discharge (SWD) on scalp EEG, clinically accompanied by slowing of reading. B: 3Hz spike and wave discharge (SWD) on stereo EEG, clinically accompanied by slowing of reading. Black right, purple left C: Right electrodes and scheme of the frontal area resection (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article).

permanently affected by an independent spiking focus. Right frontal surgery was proposed mainly in the hope of curing the ES, but the first type of seizure with 3-Hz SWD also disappeared.

Diffuse, regular, > 2.5-Hz SWD, such as those recorded in our patient, can be found in typical absence seizures seen in generalized idiopathic epilepsies. Loss of consciousness in these latter cases are nonetheless typically more profound and the symptomatology is not limited to slow reading. The same regular SWD have also been described in phantom absences with no clear clinical symptomatology which does not apply to our patient. Finally, the closest phenomenology described in the literature would be that of transient cognitive impairment which is now considered as a subtle seizure.

Generalized EEG abnormalities that disappear after surgery have been reported in children with focal lesions, but mostly in cases with very large surgical resections such as hemispherotomy.

Research data suggest that thalamo-cortical circuits are involved in the generation of SWD observed in absence seizures with involvement of frontal as well as posterior cortices [2]. The fronto-polar region has been suggested as one of the subregions of the frontal lobe involved in absence seizures [3]. A second hypothesis could be secondary bilateral synchrony that has been described in focal lesional frontal epilepsies [4].

Our case report reinforces the theory that generalized 3-Hz SWD are not truly “generalized” but rather involve selective cortical networks among which the fronto-polar region is a good candidate [3]. From a clinical point of view, our data illustrate the fact that the presence of generalized SWD associated with focal frontal lobe seizures should not exclude patients from epilepsy surgery, especially with a fronto-polar lesion.

Conflict of interest

Authors declare that there is no conflict of interest.

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