Epileptic Disord 2019; 21 (5): 463-5

Self-stimulation in an adult misdiagnosed with focal epilepsy

Pedro Correia [a], Elizabeth Hartl, Soheyl Noachtar

Epilepsy Center, Department of Neurology, University of Munich, Munich, Germany
[a] Current address: Department of Neurology, Unidade Local de Saúde do Alto Minho, Portugal

Received May 23, 2019; Accepted August 18, 2019

ABSTRACT – Self-stimulation is a normal part of development and a common behaviour in children before puberty, but very rare in adults. The stereotyped semiology can sometimes raise the suspicion of epilepsy. We present a 30-year-old patient who came to our epilepsy monitoring unit for differential diagnosis of nocturnal episodes, interpreted elsewhere as hypermotor status epilepticus associated with a known diagnosis of focal epilepsy and septo-optic dysplasia. The recorded events during video-EEG were consistent with psychogenic self-stimulating behaviour, which improved with psychotherapy. Disturbed sexual development with hypopituitarism and poor eyesight, androgen replacement therapy, alongside a protective environment provided by her parents, were the identified predisposing factors for this uncommon entity in an adult. [*Published with video sequence*].

Key words: self-stimulation, epilepsy, psychogenic

VIDEO ONLINE

Correspondence:
Soheyl Noachtar
Epilepsy Center,
Dept. of Neurology,
University of Munich,
Marchioninistr. 15,
D*81377 Munich, Germany
<noa@med.uni-muenchen.de>

doi:10.1684/epd.2019.1099

Self-exploration and self-stimlation are a normal part of development and are common behaviour in children before puberty, even in early childhood (Strachan and Staples, 2012; Rodoo and Hellberg, 2013). The stereotyped nature of the paroxysmal behaviour sometimes raises a suspicion of epilepsy and occasionally only video-EEG monitoring can establish a diagnosis (Wulff *et al.*, 1992). In adults, this type of behaviour seems to be extremely rare.

Case study

A 30-year-old female presented at our epilepsy monitoring unit for differential diagnosis of nocturnal episodes diagnosed as hypermotor status epilepticus associated with focal epilepsy by outside institutions. Her past medical history included septo-optic dysplasia (also known as De Morsier syndrome), a congenital heterogeneous malformation that includes variant degrees of optic nerve hypoplasia, pituitary hormone abnormalities,

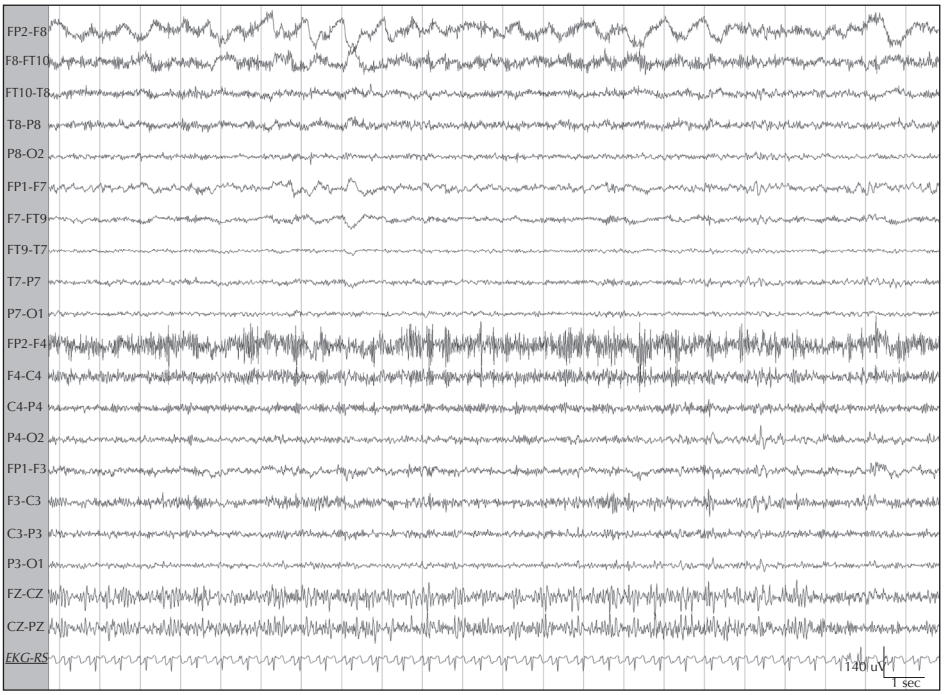

Figure 1. During the event (see *video sequence*) EEG shows a normal wake pattern, along with a movement artifact.

and midline brain defects (Webb and Dattani, 2010). The patient had a history of focal epilepsy since one year old. Her left arm clonic seizures were well controlled with carbamazepine. Although her daily living was limited by very poor vision, she learned Braille language and worked in a workshop for the blind. The parents reported stereotyped episodes in bed at night, where the patient had body-rocking movements and altered breathing patterns. The attacks were sometimes initiated by manipulation in the lower back when the parents tried to calm her. This observation led to the diagnosis of a reflex epilepsy by an outside institution. The parents alternately slept in the same bed with the patient because they were afraid of SUDEP.

During video-EEG monitoring (over a one-week period of admittance), we recorded nine habitual attacks (*see video sequence*), each up to five minutes in duration. The patient rocked her trunk and pelvis repetitively in a prone position while the EEG showed a normal awake pattern (*figure 1*). During these recordings, her medication was fully withdrawn. Two left arm clonic seizures (one of them progressing to a generalized tonic-clonic seizure) were also recorded which were associated with right central seizure patterns.

Discussion

We report an adult case of self-stimulation which was misdiagnosed as focal epilepsy for years. The semiology of the events was somewhat similar to hypermotor seizures as pelvic movements predominated, and in the context of a patient with a confirmed diagnosis of frontal lobe epilepsy, it was certainly an option to consider an epileptic nature for these events. However, there are some clues in the history of the patient that strongly argue against an epileptic cause for the events. First, it would be unusual for frequent uncontrolled hypermotor seizures and status to never propagate to other brain regions, resulting in other motor seizures, or progress to bilateral tonic-clonic seizures. Moreover, it is strange that the same seizure onset zone that produced left arm clonic seizures would also lead to complex motor semiology (Bonini *et al*., 2014). Second, it would be unusual for a patient to have nightly hypermotor seizures while being completely free of the habitual left arm clonic seizures. Third, the fact that the events could be precipitated by her parents touching her lower back is a very unlikely trigger for epileptic seizures. Reflex epilepsies are very uncommon and even more so for hypermotor seizures. Nevertheless, video-EEG monitoring is required in patients, such as the one described, to diagnose the condition.

Other differential diagnoses include parasomnias, although the EEG awake pattern at onset of the events easily ruled these out. The EEG awake pattern must be distinguished from REM-sleep patterns which are found in REM-sleep behaviour disorder (RBD), although the semiology of the case would be very uncommon for RBD.

This patient with septo-optic dysplasia was receiving hormone substitution and androgen replacement for pituitary insufficiency. There is evidence that

administration of dehydroepiandrosterone (DHEA) in women with hypopituitarism enhances libido, alongside other qualities of life, with benefits (Johannsson *et al*., 2002). Disturbed sexual development in a patient with hypopituitarism, alongside a protective environment provided by her parents based on a misdiagnosis of epilepsy, exacerbated the condition. The patient and her parents were very understanding of the psychogenic mechanisms of the condition. The attacks improved with psychotherapy. Whenever they rarely occurred thereafter, she had a strategy to limit them and rapidly get back to sleep without disturbing her parents. There was no need to stop androgen replacement therapy. □

Legend for video sequence

During video-EEG monitoring, nine habitual attacks were recorded. The patient rocks her trunk and pelvis repetitively in a prone position while the EEG shows a normal awake pattern. The attacks were sometimes initiated by manipulation of the lower back when the parents tried to calm her.

Key words for video research on www.epilepticdisorders.com

Phenomenology: hypermotor
Localisation: not applicable
Syndrome: non epileptic paroxysmal disorder
Aetiology: non epileptic paroxysmal disorder

Supplementary data.
Summary didactic slides are available on the www. epilepticdisorders.com website.

Disclosures.
None of the authors have any conflict of interest to declare.

References

Bonini F, McGonigal A, Trébuchon A, *et al*. Frontal lobe seizures: from clinical semiology to localization. *Epilepsia* 2014; 55(2): 264-77.

Johannsson G, Burman P, Wirén L, *et al*. Low dose dehydroepiandrosterone affects behavior in hypopituitary androgen-deficient women: a placebo-controlled trial. *J Clin Endocrinol Metab* 2002; 87(5): 2046-52.

Rodoo P, Hellberg D. Girls who masturbate in early infancy: diagnostics, natural course and a long-term follow-up. *Acta Paediatr* 2013; 120(7): 762-6.

Strachan E, Staples B. Masturbation. *Pediatr Rev* 2012; 33: 190.

Webb E, Dattani M. Septo-optic dysplasia. *Eur J Hum Genet* 2010; 18(4): 393-7.

Wulff C, Ostergaard J, Storm K. *Epileptic fits or infantile masturbation? Seizure* 1992; 1: 199-201.