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Video-Clinical Corners

"I feel my arm shaking": partial cataplexy mistaken for drug-resistant focal epilepsy

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1. Introduction

Diagnostic delay of narcolepsy regularly spans several years [1,2]. This is partly due to the insidious nature of disease onset and incomplete presence of characteristic symptoms [2]. Moreover, narcolepsy may mimic other neurological diseases (in particular, epilepsy) due to the presence of paroxysmal events [3]. Epilepsy, which is much more prevalent, might thus appear as a straightforward diagnosis, especially if the symptoms are focal and associated with abnormal movements or jerks (see Table 1).

Here we report a case of a 24-year-old man, who complained of recurrent episodes of "shaking" of his right arm and was erroneously treated for epilepsy for two years. Video-electroencephalogram (EEG) monitoring during cataplectic attacks helped to unravel the misleading features of segmental cataplexy.

2. Case description

A 24-year-old man without remarkable medical history started to have transient spells in the course of stressful life events. These spells, which he himself described as "shaking of his right arm," were associated with the inability to speak and rarely affected the legs, causing a fall. He was unsuccessfully treated with four antiepileptic drugs for suspected focal seizures. Among these, Gabapentin was tapered after an episode of daytime visual hallucinations: the patient saw the socket of his room on fire [4]. Valproic acid caused/contributed to a weight gain of 35 kg/70lb and significant tiredness. The pharmaco-resistance and psychological context incited his epileptologist to refer him to video-EEG monitoring, which allowed the correct classification of these shaking events as partial cataplexy (Fig. 1, recordings performed according to local ethics guidelines).

The sleep study confirmed the diagnosis with five out of five sleep-onset rapid-eye-movement sleep (REM) periods. A mean sleep latency of 1.4 min and an Epworth score of 16 revealed an excessive daytime somnolence, despite there being no spontaneous clinical complaint. Furthermore, polysomnography evidenced major sleep fragmentation and REM sleep without atonia. HLA DQB1*0602 was positive. Tapering of valproic acid allowed for a significant weight loss (30 kg/60lb). The patient became symptom free with modafinil and venlafaxine. He has not complained of somnolence or cataplectic attacks for two years.

3. Video analysis

Video-EEG-monitoring documented two episodes of several minutes duration without specific prodromes. The patient presented with head drop, weakness of upper limbs and facial muscles resulting in eyelid closure and slurred speech with preserved consciousness (Video 1); the latter was confirmed by clinical examination (supplementary videos). When he attempted to move, instability of muscle tone generated shaking movements of the right upper limb, similar to negative myoclonus (Video 2). Subjectively, episodes started with a feeling of "tremor," not necessarily visible, of parts of his muscles and limbs, which otherwise felt "heavy." EEG recordings did not show any epileptic discharge, but captured significant changes in electromyogram (EMG) artifacts of

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Table 1
Characteristics of atonic attacks in narcolepsy type 1 (NT1) and different forms of epilepsy. While drop attacks in epilepsy can be of multiples origins (myoclonic or tonic seizures, asystole, etc.), genuine atonic seizures are very rare in focal epilepsy (bilateral synchrony) and generally limited to severe childhood epilepsy in generalized forms of the disease (Doose and Lennox-Gastaut syndrome, rare forms of childhood epilepsy). See supplementary table for complete reference[1–11].

	Cataplexy in NT1	Atonic seizures in focal epilepsy	Atonic seizures in symptomatic generalized epilepsy	Myoclonic seizures in idiopathic generalized epilepsy
Age of onset	Variable, Generally 2nd or 3rd decade[1]	Variable, more frequent in adulthood	Severe childhood epilepsies, onset often 2–6 years[2]	Variable, onset peaks in adolescence[3]
Associated symptoms	Somnolence Sleep paralysis Hallucinations Sleep fragmentation	Focal seizures of location related semiology, +/− secondarily generalized seizures	Mental retardation Variety of seizures (+myoclonic in Doose's, + tonic in Lennox-Gastaut)	Generalized seizures (tonic-clonic), absences seizures
Immediate triggers of the attack	Frequent: laughing, joking, surprise, tiredness	Usually absent (exceptionally reflex epilepsies)	Usually absent (rare reflex epilepsies)	Infrequent (photic stimulation, awakening)
Prodrome to the onset of the attack	Absent	Exceptional in atonic seizures, in focal seizures aura indicative of onset zone	Usually absent	Usually absent
Attack onset	Abrupt	Abrupt, may occur during a seizure with focal onset	Abrupt	Abrupt
Consciousness	Present	Most often absent in atonic seizures	Most often absent (unresponsiveness)	Most often present
Language	Absent or slurred	Absent, speech arrest in prolonged focal seizures	Absent	Can be preserved
Eyes	Eyelid weakness	Variable, fixed gaze if prolonged partial seizure	Variable, half closed	Variable, possible eyelid myoclonia
Facial muscle tonus	Atonic, cataplectic facies, jaw tremor	Atonic	Atonic	Possible facial myoclonus
Body muscle tonus	Partially or globally affected, atonic falls	Atonic predominant on the limbs	Atonic predominant on the trunk	Present, positive myoclonus predominant on the upper limbs
Myoclonus	Absent during daytime (except for “negative myoclonus” during attacks)	Exceptionally present (epilepsia partialis continua)	Can be associated (myoclonic-astatic seizures)	Present, often affecting upper limbs or eye lids
Duration	Seconds, rarely minutes	Seconds, associated focal seizures often last minutes	Seconds	Seconds
Postictal confusion	Absent	Can be present	Can be present	Absent
Ictal/postictal amnesia	Absent	Usually present	Usually present	Usually absent
Deep tendon reflexes	Absent	?	?	(rather enhanced)[4]
EEG findings	Unremarkable (wake)	Bilateral synchrony	Irregular spike and wave discharge < 3Hz	Polyspike waves
EMG findings	Abolition of postural muscle tone	Abolition of muscle tone	Abolition of muscle tone	Myoclonus, burst of EMG activity < 500 ms (cortical often < 100 ms)[5]
PSG findings	SOREMPs, REM sleep without atonia, Frequent dyssomnia	Possible activation of spike waves during sleep	Possible activation of spike waves or tonic seizures during sleep	Possible activation of spike and polyspike waves during sleep

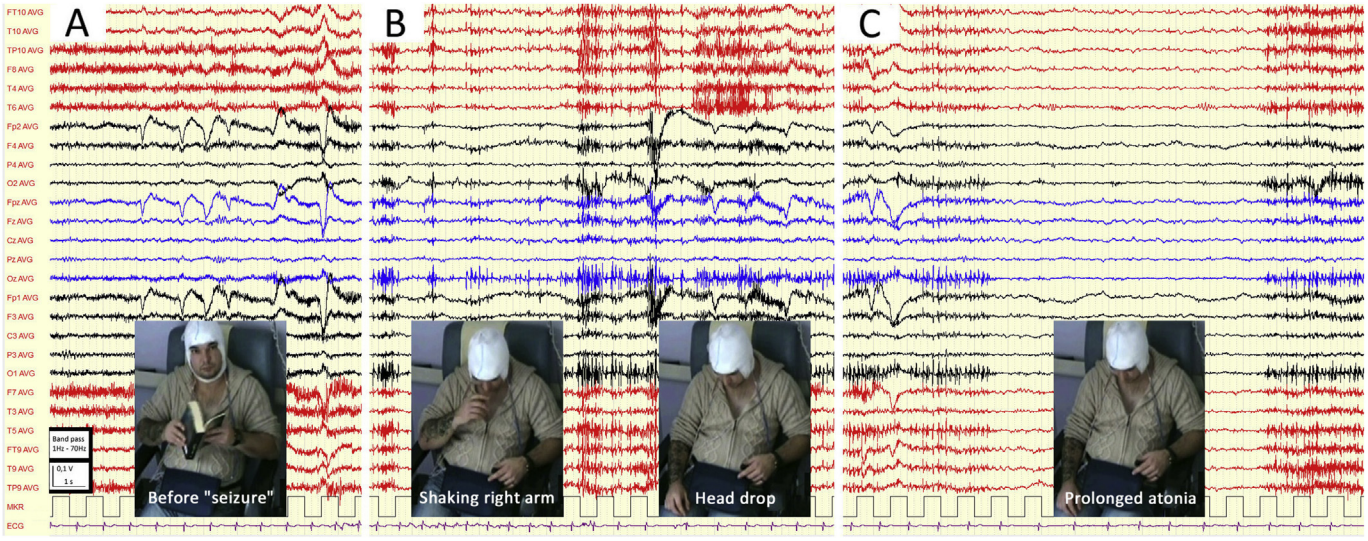

Fig. 1. “Shaking” without epileptic activity on EEG. (A) Muscle artifacts before the “seizure” (supplementary Video 1), soon followed by onset of cataplexy (Video 1). (B) “Shaking” of the right arm equivalent to negative myoclonus of upper limbs and head during partial cataplexy, with intermittently abolished muscle activity (Video 2). (C) Prolonged atonia with abolished muscle artifacts (suppl. Video 3), without epileptic activity.

facial muscles. EMG-amplitude increased, showed a ragged appearance and was intermittently interrupted for several seconds during prolonged atonia. Facial EMG thus demonstrated instability of muscle tone on a short and longer time scale.

Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.sleep.2017.05.003

The episodes lasted 3–4 min without loss of consciousness, confusion or amnesia.

4. Discussion

This case graphically illustrates the diagnostic pitfalls of narcolepsy, which some authors claim to be a master of disguise [2]. Beyond the misleading psychological context [5], the present case was particularly challenging due to the manifest segmental symptoms of partial cataplexy. Similarities between narcolepsy and epilepsy have been discussed since Gelineau in 1880 [3]. With the exception of one case description in children [6], the medical literature largely represents the differential diagnosis between narcolepsy and generalized epilepsy [3,7] (see Table 1). In our patient, though, it is specifically the focal nature of symptoms which entailed two years of ineffective treatment. Some further elements puzzled the physicians: slurred speech emanating from facial weakness, mistaken for epileptic speech arrest, the prolonged duration of the episodes without immediate falls whereas emerging sleepiness and weight gain were considered as an adverse effect of valproic acid.

Video-EEG monitoring documented intermittent segmental atonia resulting in visible “shaking,” equivalent to negative myoclonus [3,8], and in prolonged atonic spells with head drop [9]. In congruence with the study by Vetrugno and colleagues [10], the EMG signature of narcolepsy spanned very brief events, in the millisecond range, to several seconds of atonia. We propose that the subjective feeling of “tremor” corresponds to sustained episodes of intermittent milliseconds interruptions of muscle tone.

Our report on misleading focal motor symptoms completes the current literature, which so far focuses on the differential diagnosis with suspected generalized epilepsy. [3,7], indicating that partial cataplexy, as a prevalent hallmark of narcolepsy type 1 [9,10], may still be less well-known in the neurological community [2]. Thorough characterization of segmental atonia with video and neurophysiological monitoring may therefore help to overcome the diagnostic challenges of partial cataplexy.

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Conflict of interest

The ICMJE Uniform Disclosure Form for Potential Conflicts of Interest associated with this article can be viewed by clicking on the following link: http://dx.doi.org/10.1016/j.sleep.2017.03.014.

Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.sleep.2017.04.006.

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