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Video-Clinical Corners

Persistence of limb dystonia and myoclonus during sleep in corticobasal syndrome: a case series

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1. Introduction

Corticobasal syndrome (CBS) encompasses a clinical phenotype caused by various neurodegenerative pathologies, mainly tauopathies, characterized by a combination of basal ganglia and cortical dysfunction, with a typical asymmetric presentation [1]. Thus far, few studies have focused on sleep in CBS, documenting rapid eye movement (REM) sleep behavior disorder (RBD) or periodic limb movements during sleep (PLMS) [2]. No study has evaluated the possible sleep modulation of the motor abnormalities, such as dystonia or myoclonus. Here, we present our findings from three CBS patients who underwent nocturnal video-polysomnography (PSG) through an extensive electromyography (EMG) montage, showing the persistence of dystonia/myoclonus during sleep.

2. Case description

Three patients (mean age, 70 years) with a history of asymmetric akinetic-rigid syndrome, L-dopa unresponsive, were prospectively evaluated. Patients described the mainly affected limbs as feeling foreign, with cramps, tingling, and abnormal posturing. Neurological evaluation revealed the presence of dystonia ± myoclonus, involving the left hemibody in cases 1 and 2, and the right side in case 3 (Table 1). Ideomotor apraxia and cortical sensory impairment were observed in all of them. Diagnoses of CBS were consequently made, fulfilling current clinical criteria [3].

All patients underwent nocturnal PSG, with an extensive EMG montage comprehensive of the arm muscles affected with dystonia/myoclonus and the contralateral ones. Sleep scoring and analysis of EMG signal coupled with video analysis were performed off-line by two sleep expert (E.A. and F.P). Atonia index analysis was performed off-line by means of Hypnolab 1.2 sleep software analysis (SWS Soft, Italy) on the submentalis muscle [4].

3. Video analysis

3.1. Case 1 (Video 1, part 1)

Video-PSG revealed the occurrence of myoclonic jerks and tonic activity, involving the left biceps brachii and the extensor carpi ulnaris muscles, through all the sleep stages, sometimes recurring in a periodic fashion (every 20 s). In addition, prolonged painful contractions of left arm, arising both during wakefulness and sleep, were observed.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.sleep.2018.12.025.

3.2. Case 2 (Video 1, part 2)

Video-PSG showed a tonic activation on the left extensor carpi ulnaris muscle, during NREM sleep (included slow waves sleep), associated with further phasic increment of muscular tone over the same muscle.

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Table 1
Clinical features and laboratory findings of corticobasal syndrome (CBS) patients.

	Case 1	Case 2	Case 3
Age (y)	64	78	68
Disease duration (y)	3	2	3
Cortical clinical features	Limb apraxia, cortical sensory dysfunction, alien limb phenomenon	Alien limb phenomenon, limb apraxia	Limb apraxia, cortical sensory dysfunction, alien limb phenomenon
Motor abnormalities	Limb rigidity, limb dystonia, limb myoclonus	Limb rigidity, limb dystonia	Limb rigidity, limb dystonia, limb myoclonus
[123I]FP-CIT brain SPECT	Reduced uptake in the right putamen	Slightly reduced uptake in the right putamen	Reduced uptake in the left putamen
Cerebral MRI	Asymmetric parietal cortical atrophy (predominant on the right side)	Right fronto-parietal cortical atrophy	Asymmetric fronto-parietal cortical atrophy (predominant on the left side)
Cerebral 18F-FDG PET/CT	Glucose metabolic reduction in the right parietal lobe	Not performed	Glucose metabolic reduction in the left parietal lobe
PSG findings			
Sleep efficiency	55%	63%	50%
PLMS index	9.6/h	35.5/h	1.3/h
Sleep stages (%)	N1 19.5%	N1 10.3%	N1 71.6%
	N2 45.5%	N2 43.4%	N2 15.4%
	N3 25.1%	N3 37.5%	N3 0%
	REM 10%	REM 8.9%	REM 13%
SL	4 min	17 min	25 min
REML	232 min	178 min	134 min
ODI	5/h	1.2/h	16.9/h
REM sleep atonia index	0.59	0.89	0.37

[123I]FP-CIT SPECT, iodine-123-N-ω-fluoropropyl-2β-carboxymethoxy-3β(4-iodophenyl)nortropanesingle photon emission computed tomography; 18F-FDG PET/CT, 18F-fluorodeoxyglucose positron emission tomography/computed tomography; MRI, magnetic resonance imaging; ODI, oxygen desaturation index; PLMS index, periodic limb movements per hour of sleep; REM, rapid eye movement; REML, REM sleep latency; SL, sleep latency.

3.3. Case 3 (Video 1, part 3)

Video-PSG disclosed the occurrence of myoclonic jerks and co-contraction/tonic activity, especially involving the right arm (biceps, flexor and extensor carpi ulnaris muscles) in all sleep stages, mainly during light sleep, even if with low-voltage amplitude compared to those recorded during wakefulness.

In addition, all patients showed severe impairment of sleep architecture and markedly reduced sleep efficiency (mean of 56%) (Table 1). PLMS occurred with an index of 35.5/h in case 2 but was normal in the cases 1 and 3. Atonia index analysis showed a reduced mean value (0.62), although no episode of RBD was observed or clinically reported.

4. Discussion

Our study documented in CBS patients the persistence during sleep of dystonia and myoclonus, involving the same arm affected by the alien limb phenomenon. Sleep usually leads to disappearance of almost all abnormal movements, putatively due to basal ganglia dysfunctions, including dystonic posturing in idiopathic isolated dystonia [5]. Accordingly, the persistence of dystonic posture and myoclonus during sleep in our CBS patients could indicate a cortical rather than subcortical generator, in line with previous neurophysiological studies, suggesting abnormal motor cortical excitability [6,7]. In addition a recent functional magnetic resonance imaging study demonstrated the isolated activation of the primary motor cortex, instead of the whole sensory-motor network, associated with levitation and tentacular movements in CBS [8].

To our knowledge, there is only one study in which a PSG montage included arms muscles (exclusively the extensor digitorum brevis muscles) in a CBS patient, detecting the presence of periodic arm and leg movements, without side prevalence [9].

Furthermore, the mean atonia index of the patients was reduced, suggesting abnormal control of muscle tone during REM sleep. However, REM sleep behavior disorder was not clinically suspected or documented in the video-PSG study. Of note, two patients were on selective serotonin reuptake inhibitor (SSRI) treatment, although this was suspended one week before PSG, and therefore we cannot exclude a contribution of medications on computation of the atonia index. However, our finding are in agreement with previous reports of REM sleep without atonia in CBS [2].

To conclude, our sleep study showed, in contrast to what has been reported in idiopathic dystonia, the persistence of the movement disorders during sleep; thus leading us to infer a likely cortical origin of the latter. This observation should be confirmed by a larger sample, and compared with findings in a control group.

Informed consent

The patients provided written informed consent for video publication.

Conflict of interest

The authors declare no 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: https://doi.org/10.1016/j.sleep.2018.12.025.

References

[1] Bezerra Parmera J, Dieh Rodriguez R, Studart Neto A. Corticobasal syndrome: a diagnostic conundrum. Dement Neuropsycol 2016;10:267–75.
[2] Abbott SM, Videnovic A. Sleep disorders in atypical parkinsonism. Mov Disord Clin Pract (Hoboken) 2014;1:89–96.
[3] Armstrong MJ, Litvan I, Lang AE, et al. Criteria for the diagnosis of corticobasal degeneration. Neurology 2013;80:496–503.
[4] Ferri R, Rundo F, Manconi M, et al. Improved computation of the atonia index in normal controls and patients with REM sleep behavior disorder. Sleep Med 2010;11:947–9.
[5] Antelmi E, Ferri R, Provini F, et al. Modulation of the muscle activity during sleep in cervical dystonia. Sleep 2017;40(7).

[6] Valls-Sole J, Tolosa E, Marti MJ, et al. Examination of motor output pathways in patients with corticobasal ganglionic degeneration using transcranial magnetic stimulation. Brain 2001;124:1131–7.
[7] Frasson E, Bertolasi L, Bertasi V, et al. Paired transcranial magnetic stimulation for the early diagnosis of corticobasal degeneration. Clin Neurophysiol 2003;114:272–8.
[8] Onofrj M, Bonanni L, Delli Pizzi S, et al. Cortical activation during levitation and tentacular movements of corticobasal syndrome. Medicine (Baltimore) 2015;94:e1977.
[9] Wetter TC, Brunner H, Collado-Seidel V. Sleep and periodic limb movements in corticobasal degeneration. Sleep Med 2002;3:33–6.