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Brief Communication

Status dissociatus evolving from REM sleep behaviour disorder in multiple system atrophy

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Abstract

Objective: We present two patients, a 52-year-old man and a 56-year-old woman, with rapid eye movement sleep behaviour disorder (RBD) since the age of 50.
Method: In both the patients RBD was videopolysomnographically documented.
Results: Both patients developed, with time, autonomic and motor symptoms consistent with the diagnosis of multiple system atrophy. During the course of the disease RBD episodes diminished in frequency but the patients' sleep became even more abnormal, with nearly continuous motor and verbal abnormal behaviours and ambiguous and rapid oscillations of state-determining polysomnographic variables, now consistent with status dissociatus (SD).
Conclusion: If SD represents the evolution and most extreme form of RBD it should be investigated longitudinally in neurodegenerative diseases.

Keywords: REM sleep behaviour disorder; Status dissociatus; Sleep; Wake; Polysomnography; Multiple system atrophy

1. Introduction

A breakdown of state-determining boundaries during which elements of one state of being (Wake, NREM and REM sleep) persist or are pathologically recruited into another state identifies a condition named status dissociatus (SD) [1,2]. According to the prevailing present state, Wake, NREM and REM sleep variations of SD exist. SD has been reported in several clinical and experimental situations such as hypothalamic, thalamic and brainstem lesions, pharmacological interventions, or after sleep deprivation [1]. We describe here two patients suffering from probable multiple system atrophy (MSA) and affected with REM sleep behaviour disorder (RBD), who later developed abnormal behavioural and neurophysiological patterns during sleep consistent with SD, with nearly continuous motor and verbal behaviour in the absence of polygraphically defined conventional REM and NREM sleep stages. SD, in our two patients, appeared to follow upon and to represent the evolution of RBD.

2. Case report 1

A 52-year-old man was referred because of violent behaviour during sleep he had experienced for two years. In particular, the patient had noted that his dreams had become more vivid and intense, their content characterized by threats or attacks by unfamiliar people or animals and by the patient fighting the attacker to protect himself. The wife reported that the patient talked, yelled, gesticulated, grabbed and

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punched during sleep almost every night. At age 51, erectile failure, a harsh high-pitched nocturnal breathing noise and gait impairment began. A neurological examination showed gait ataxia and dysarthria plus bradykinesia and brain MRI cerebellar and brainstem atrophy. Cardiovascular reflex responses to standing and to the Valsalva maneuver were defective and consistent with the diagnosis of central autonomic failure in probable MSA [3].

The patient underwent night videopolysomnographic recordings (VPSG) including EEG (C3-A2, O2-A1, Cz-A1); right and left electro-oculography; surface EMG of the submental, wrist extensor, tibialis anterior and intercostalis muscles; ECG; microphone; oro-nasal, thoracic and abdominal respirograms; endoesophageal pressure; systemic blood pressure; and oxygen saturation.

During Wake, the patient was quiet with a 10–11 Hz posterior EEG alpha rhythm. Sleep time was 181 min (NREM sleep stages 1–2: 33.6%; NREM sleep stages 3–4: 51.5%; REM sleep: 17.9%), interrupted by several awakenings with wakefulness after sleep onset (WASO) of 117 min (Fig. 1A). Sleep latency was 36.5 min and REM latency 235 min. Arousal index (AI, number of arousals per hour of sleep) was 4 (n.v. $\leqslant$ 10), PLMS index (PLMS-I, number of periodic limb movements per hour of sleep) 29 (n.v. $\leqslant$ 5) and the cyclic alternating pattern (CAP) rate (percentage of NREM sleep occupied by CAP sequences) 21.2% (n.v. $35.9 \pm 8.9\%$ [4]). Paradoxical breathing (PB, a phase-out of thoracic as opposed to abdominal respiratory traces), recurrent gasps and stridor were documented, but despite a respiratory disturbance index (RDI, number of obstructive apnoea–hypopnoea per hour of sleep) of 14 (n.v. $\leqslant$ 5), oxyhaemoglobin saturation (SaO_2) never fell below 90% (range 92–98). During REM sleep, chin muscle tone persisted and two episodes with recurring upper and lower limb muscle twitching and jerking were recorded, with the patient outstretching and/or flinging

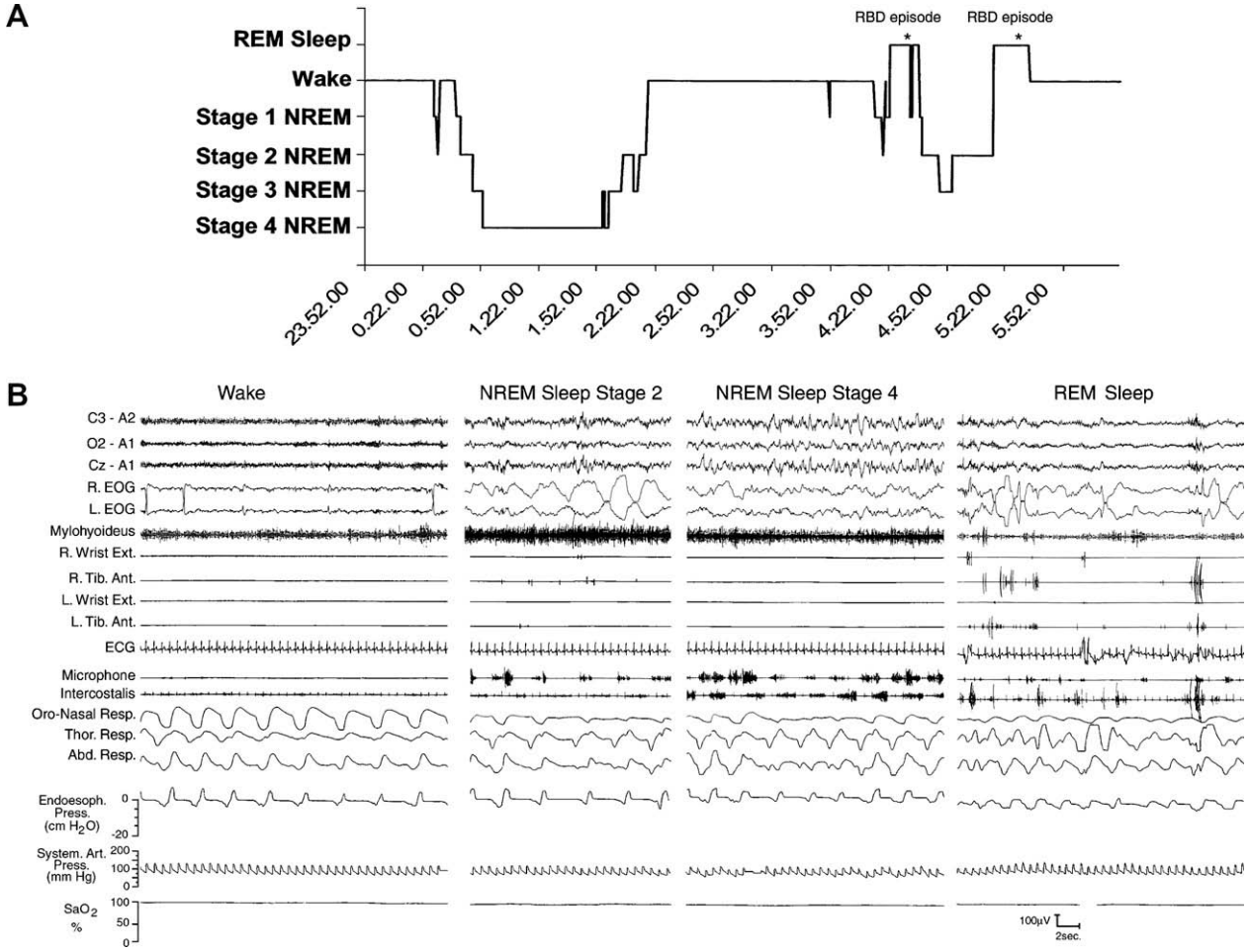

Fig. 1. (A) Upper panel. Patient 1 first VPSG histogram. Despite a short sleep time, a cyclic structure of sleep is still present. REM sleep was without atonia, and two RBD episodes occurred (marked by *), (B) lower panel. PSG recordings showing wake state (Wake), light (NREM sleep Stage 2) and slow (NREM sleep Stage 4) NREM sleep, and REM sleep during RBD (REM Sleep). EOG, electro-oculogram; Wrist Ext., wrist extensor; Tib. Ant., tibialis anterior; ECG, electrocardiogram; Oro-Nasal Resp., oro-nasal respirogram; Thor. Resp., thoracic respirogram; Abd. Resp., abdominal respirogram; Endoesoph. Press., endoesophageal pressure; System. Art. Press., systemic arterial pressure; SaO_2, oxygen saturation; R, right; L, left.