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Case report

Longitudinal change in REM sleep components in a patient with multiple system atrophy associated with REM sleep behavior disorder: paradoxical improvement of nocturnal behaviors in a progressive neurodegenerative disease

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Abstract

A 60-year-old patient with multiple system atrophy (MSA) who presented with rapid eye movement (REM) sleep behavior disorder was investigated longitudinally by all-night polysomnography. REM sleep components, i.e. rapid eye movements and chin muscle activity, were analyzed together with the frequency of behaviors/movements on the videorecording. Decreased frequency of elaborate motor activity during REM sleep with time in this patient was compatible with the observation by his wife, and this change seemed to correlate with predominant tonic chin electromyogram with relatively suppressed phasic chin muscle activity, but the reduction of the REM sleep behavior disorder (RBD) episodes could be interpreted as being due to the increased rigidity along with MSA progression. The chronological change in REM sleep components in RBD with neurological disorders is worth studying in large follow-up series to enlarge our knowledge about the mechanism of behavioral manifestation of RBD in humans.

Keywords: Rapid eye movement sleep; Rapid eye movement sleep behavior disorder; Rapid eye movement sleep without atonia; Polysomnography; Multiple system atrophy; Rapid eye movements; Chin muscle activity

1. Introduction

Although abnormality of rapid eye movement (REM)-mediated muscle atonia has been considered to be a pathophysiological basis for causing REM sleep behavior disorder (RBD) [1], it is not a sufficient condition to generate behaviors/movements during REM sleep [2]. The natural history of RBD itself varies, as some cases are progressive and some remain stable in the frequency of events. Polysomnographical (PSG) studies investigating detailed REM sleep parameters have been scarce, and much fewer repeated PSGs have been performed to confirm whether any alteration in components of REM sleep occurred along with the change in the frequency and intensity of abnormal behaviors/movements. We report longitudinal change in various components of REM sleep in an RBD patient with multiple system atrophy (MSA).

2. Subject and methods

A 60-year-old patient was referred with a complaint of a 4.5-year history of gait disturbance and gradual worsening of dysarthria. His wife became aware of his vigorous sleep talk associated with limb and body jerking and snoring almost every night 1 year prior to referral. Some of his behaviors seemed to be enacted by dream contents. No present or past history was noted of excessive alcohol intake, drug abuse, or psychotropic medication. Physical examination was unremarkable and his neurological findings on the first exam were mainly cerebellar. A tentative diagnosis of MSA with cerebellar features was made taking into account the characteristic brain MRI finding (cerebellar and brainstem atrophy associated with high signal intensity

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in the cruciform shape in T2 weighted images) [3], and this diagnosis was confirmed 1 year later with development of urinary dysfunction, mild Parkinsonian features (rigidity and postural instability), and extensor planter responses with hyperreflexia [4]. A repeat brain MRI scan revealed enhanced cerebellar and brainstem atrophy. With the progression of MSA, his wife reported his sleep talk and behaviors became less frequent and intense. The patient had no medication throughout the course.

Three all-night polysomnographies (PSG) with video recording were performed at 4.7, 6.2, and 7.0 years after the onset of MSA, following standard procedure [5] with additional surface electromyogram (EMG) recording from wrist flexors, extensors and tibialis anterior muscles.

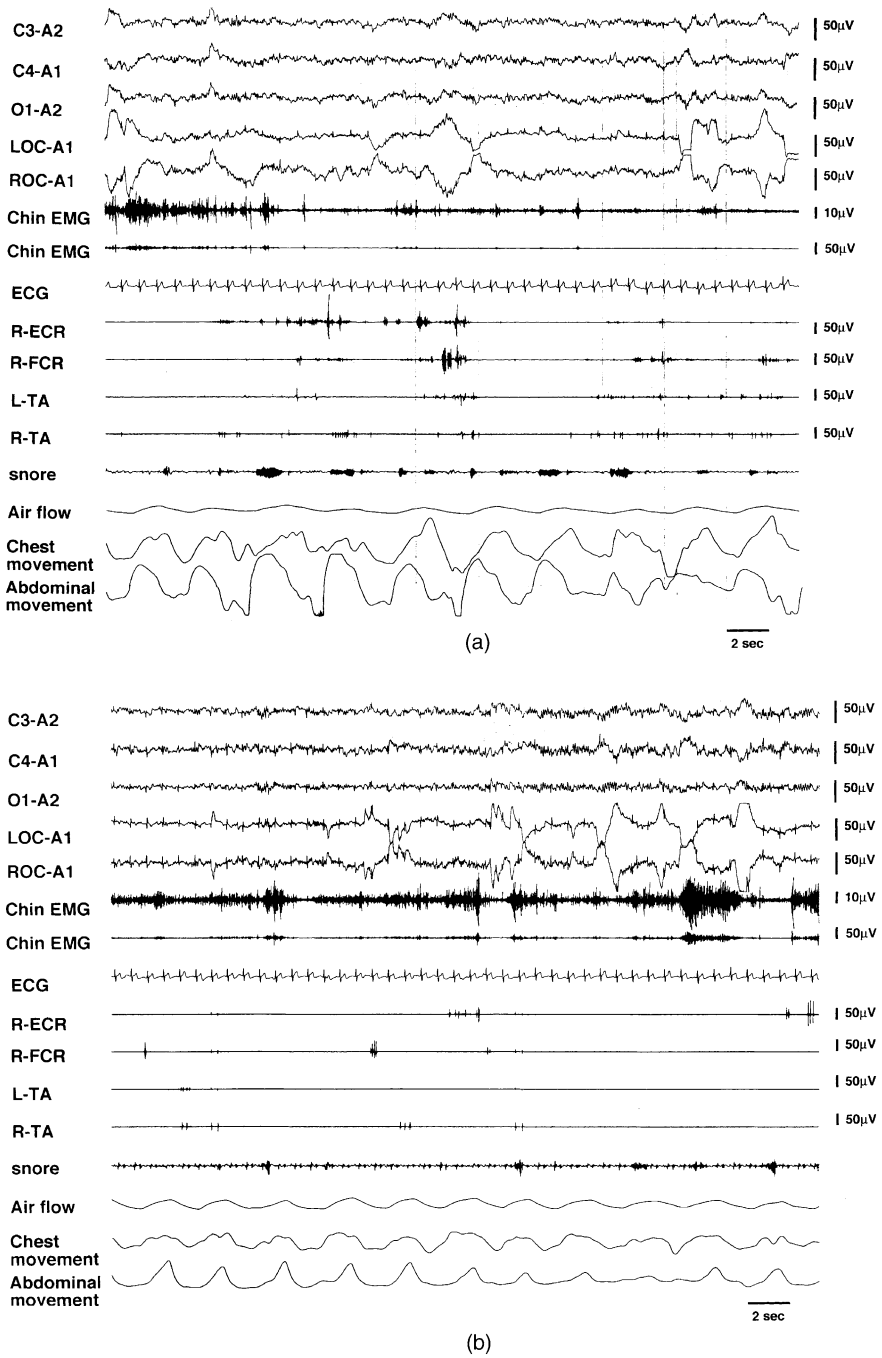
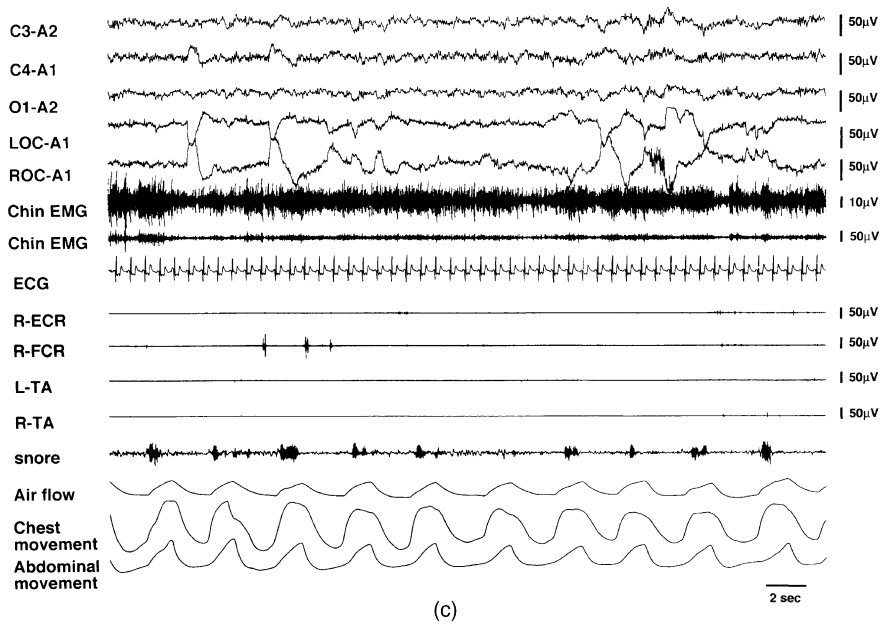


Fig. 1. Polygraphic records on three occasions showing REM sleep without atonia. In the first PSG, chin and limb EMGs are predominantly phasic (A). The second PSG shows low-amplitude tonic chin EMG overlaid by phasic muscle (B). In the third PSG, there is continuous tonic chin EMG and scarce EMG activity in the limb muscles (C). LOC, left outer canthus; ROC, right outer canthus; ECR, extensor carpi radialis; FCR, flexor carpi radialis; TA, tibialis anterior.



(c)

Fig. 1 (*continued*)

Staging of wake and non-REM (NREM) sleep was performed according to the standard criteria [5], and the modified version was used for staging REM sleep and REM sleep without atonia (RWA) [6]. REMs were defined as deflection of the electro-oculogram (EOG) with a peak-to-peak amplitude of 75 μV or larger, and 50° or greater angle of rise when recorded with calibration at 50 μV/cm. REM density was calculated as the percentage of the total number of 3-s mini-epochs containing at least one REM to the total number of 3-s mini-epochs of REM sleep. Phasic mentalis muscle activity (PMMA) was defined as the EMG discharge from the chin muscle lasting less than 2 s and having a peak amplitude of at least 50% above the baseline amplitude during REM sleep [7]. Tonic mentalis muscle activity (TMMA) was also defined as increase in chin EMG activity with the same amplitude criteria, but the duration should be 2 s and more [8]. PMMA and TMMA densities were calculated as the percentage of the total number of 3-s mini-epochs containing at least one PMMA or episode of TMMA to the total number of 3-s mini-epochs of REM sleep. When one episode of muscle activity began in a certain mini-epoch and terminated in the following mini-epoch, two consecutive mini-epochs were counted as positive.

On all three PSGs, abnormal behaviors/movements associated with sleep talking were observed during RWA, but their frequency decreased with time. The patient sometimes snored, but no stridor was observed. Sleep architecture including NREM/REM cycle was intact, although sleep spindles became scarcer in the third PSG. No EEG slowing in REM/RWA periods was detected visually. The percentage of REM sleep (including RWA) in total sleep time remained almost the same (11.9, 21.2, 16.3%), but the ratio of RWA to the whole of REM sleep increased (75.8, 84.7, 100%). TMMA density increased with time (23.0, 58.4, 91.8%), but PMMA density in the second and third PSGs was less than that in the first one (58.0, 30.8, 32.4%). In the first PSG, EMG in the limbs also showed twitchy activity (Fig. 1A), but in the second (Fig. 1B) and third PSGs (Fig. 1C), it became much less frequent. REM density was more than 30% on all the three PSGs (50.4, 40.1, 55.0%).

3. Discussion

Decreased behaviors/movements with time was confirmed on PSG/video recording in this patient, which was consistent with the report by his wife that these symptoms associated with sleep talking became less frequent and intense. It seems paradoxical, but one study has shown that the frequency of nocturnal events decreased with time in 26% of untreated patients with neurodegenerative disease [9].

At a more advanced stage, the ratio of RWA to the whole of REM sleep increased in our patient, while the sleep architecture as well as the percentage of REM sleep was maintained. This suggests that this PSG change is attributed to extensive brainstem pathology in the domain of neuronal systems responsible for REM-related atonia. In addition, as MSA progressed, the PSG showed decreased phasic muscle activity and increased tonic chin muscle activity. Increased phasic EMG activity has been observed in clinical RBD patients with various background diseases [6], and it may play a more crucial role in behavioral manifestation than increased tonic EMG activity, reflecting overacting excitatory motor systems generating phasic muscle activity during REM sleep [10].

Another explanation is that progression of the extent and severity of brainstem lesions in MSA may alter

the manifestation of RBD, regardless of the feature of RWA. Or more simply, it could be interpreted as due to the increased rigidity as MSA progressed.

REM density remained high on all three occasions; it is unclear whether high REM density is a necessary condition for generating behaviors/movements during RWA. Neuronal substrates executing REMs are complicated, and it is also unknown whether increases in REM density are induced by MSA itself.

Further longitudinal studies in a large series are required to determine whether this paradoxical course of RBD and change in RWA is common in MSA patients with RBD.

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