Sleep-Related Head Jerk Presenting With Dream Enactment Behavior: A Case Report

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Sleep-related head jerk (SRHJ) is conceived as a physiological motor phenomenon, which is mostly seen during rapid eye movement (REM) sleep. It should be distinguished from movements during REM sleep, mainly REM sleep behavior disorder (RBD). A 25-year-old male complained of daytime sleepiness with snoring and dream enactment behavior (DEB). Video polysomnography (PSG) excluded RBD and obstructive sleep apnea; frequent SRHJs during REM sleep with or without respiratory effort-related arousals, and SRHJs followed by one episode of DEB was observed. On PSG with continuous positive airway pressure titration, SRHJ with arousal events still remained but DEB symptom was resolved. REM without atonia unrelated to head jerks was not observed on all PSG data. Excessive SRHJs with arousal related to a sleep disturbance could indicate an SRHJ disorder, which might be a novel sleep-related movement disorder. In addition, clinicians should be careful in diagnosing young patients with RBD.

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INTRODUCTION

Sleep-related head jerks (SRHJs), also named as neck myoclonus, appear as brief flexions or versions of the head during sleep and are seen mostly during rapid eye movement (REM) sleep.1 Neck myoclonus during REM sleep was described and investigated by the Innsbruck group in 2010. On polysomnography (PSG), it is characterized by typical vertical striped movement-induced artifact over the electroencephalography (EEG) leads lasting up to 2 s.2 Recent articles have adopted the term SRHJ instead of neck myoclonus; given the duration, the term ‘jerk’ is preferable to myoclonus; thus, the term SRHJ is more appropriate and aligned with the third version of the International Classification of Sleep Disorders (ICSD-3), in which movement disorders are named with reference to their relationship with sleep (e.g., sleep-related rhythmic movement disorder).3 SRHJs are neither a disease category nor a normal variant in the ICSD-3, likely because it was described only recently and also due to unclear characterization and uncertain clinical relevance. SRHJ events are often related to microarousals, respiratory efforts, leg movements and sleep fragmentation or sleep stage shifting; these suggest that respiratory- or motor-related arousals trigger sternocleidomastoid muscle contraction and induce head jerks.4 REM sleep behavior disorder (RBD) is an REM sleep parasomnia characterized by the loss of muscle atonia during REM sleep. Dysfunction in the key neuronal structures that generate REM sleep muscle atonia leads to abnormal behaviors such as vocalization and dream enactment movements. RBD may either occur in isolated or in associated with other conditions such as neurodegenerative disorders and narcolepsy. Autoimmune, paraneoplastic diseases, brainstem lesions, and medications can cause symptoms of RBD, less frequently.5 Prodromal RBD is a stage in which covert but quantifiably observable features of evolving RBD are present during video-polysomnography (v-PSG) but do not conform to established diagnostic criteria for RBD.6 In a few studies, the frequency of head jerk was higher in patients with RBD or REM without atonia (RWA) than in patients without RBD or RWA.1,4 SRHJs could also be conceived as a variant of the excessive phasic muscle activity in REM sleep with prodromal RBD or parasomnia overlap disorder.

Here, we describe a 25-year-old male who presents with head...
jerks, dream enactment behavior (DEB), and sleep talking during REM sleep.

CASE REPORT

A 25-year-old male presented with daytime sleepiness and non-restorative sleep about one year ago. His Epworth Sleepiness Scale score was 10, in addition to the reported snoring. He reported a habitual sleep period from midnight to 7:00 am. He was not taking any medications and had no significant medical history. He added that his friends have noted that he talked and flailed his arms during sleep several times. When he drank alcohol and slept, he felt his body move a lot; however, he did not complain specifically of head movements and was unaware of SRHJs. He denied sleep paralysis and cataplexy; he had no history of sleep walking as a child. He had a body mass index of 21.6 kg/m² and was not obese. The patient underwent diagnostic v-PSG to evaluate abnormal movements and apnea/hypopnea during sleep. On his PSG, his sleep onset latency and REM sleep latency were 36 min and 97.5 min, respectively; his sleep architecture was notable for an increased REM sleep percentage at 27.0% and within normal limits for other sleep stages (N1, 6.2%; N2, 56.6%; N3, 10.2%). He had an apnea-hypopnea index (AHI) of 1.6/h and respiratory disturbance index of 12.0/h, and an oxyhemoglobin saturation nadir of 94%. Video recording and PSG revealed jerking movements of the head, which were sudden rotations of the head from left to right or right to left. Head jerks were recognized visually throughout the recording and manually labeled according to previous publication. The entire recording was closely investigated, with each 30-s epoch inspected for the characteristic “stripe-shaped” movement-induced vertical visible artifacts over the EEG traces (Fig. 1A and Supplementary Video 1 in the online-only Data Supplement). During total sleep period, 23 head jerks were observed. The head jerk index (HJI), defined as the total head jerks per hour of total sleep time, was 4.0/h. The HJI during REM sleep (REM-HJI) was 13.8 and that during non-REM sleep was 0.2/h. One episode of DEB was observed and followed by SRHJs. During DEB, tonic chin electromyography (EMG) activity and phasic legs EMG activities were seen (Fig. 1B and Supplementary Video 2 in the online-only Data Supplement). However, REM sleep without

Figure 1. Four polysomnographic data of 30 s each in the patient with sleep-related head jerks (SRHJs). A: SRHJs occurring during rapid eye movement (REM) sleep that followed respiratory effort-related arousal (RERA). B: SRHJs with RERA followed by sleep talking with dream-enacting behavior. C: SRHJs occurring during REM sleep with spontaneous arousal on continuous positive airway pressure titration. D: SRHJs followed by a complete awakening.
ataxia was not observed in other REM periods where there is no neck movement. Of the 23 head jerks, 11 (47.8%) followed respiratory effort-related arousal (RERA), three and five followed hypopnea and spontaneous arousal, respectively.

Although AHI itself was not an indication for continuous positive airway pressure (CPAP) therapy, we observed SRHJs associated with RERA and the patient complained of daytime sleepiness. Thus, we tried CPAP titration and to determine whether SRHJ and DEB would resolve. On the second PSG for CPAP titration at 5 cm of water, the apnea, hypopnea, and RERA resolved; however, SRHJs remained. During REM, 18 head jerks were observed, 13 events were associated with spontaneous arousal (Fig. 1C), and two were followed by a complete awakening (Fig. 1D). However, DEB, sleep talking, and RWA were not observed on this second PSG. We did not prescribe CPAP use because SRHJ and arousal events remained unresolved. Also, considering frequent arousal with abnormal movement and daytime sleepiness with non-restorative sleep, we recommended clonazepam before bedtime, which he refused; he decided to follow-up the symptoms while receiving education of routine sleep hygiene at our outpatient clinic.

**DISCUSSION**

We reported the clinical and PSG findings of a young patient with SRHJs, who complained of daytime sleepiness with DEB. In literature, SRHJs are associated with leg movements, respiratory effort, and micro-arousal. This is the first case report of SRHJs associated with DEB during REM sleep.

SRHJs are non-periodic and occur mainly during REM sleep. Several studies have investigated SRHJs using PSG data in various sleep disorders. They found that SRHJs are common during REM sleep in about 54.6%–79.7% of the study population and are more frequent in individuals less than 45 years of age. A study with healthy controls showed that REM-HJI was 0.79±1.59/h. Our patient’s REM-HJI of 13.8/h was high, with nearly all head jerks occurring during REM sleep (92%).

SRHJs were associated with arousal in about 64.5%–80%, leg movements in 38.1%–52.2%, and respiratory events in 9.6% of the cases. In our patient, SRHJs were associated with arousal in about 70% of the events; in his first PSG, 47.8% were associated with RERA. Although previous studies have shown that SRHJs are associated with arousal or a complete awakening with body movements, it is unknown whether SRHJs are a physiological phenomenon or pathological disorder. SRHJ occurs in various sleep disorders, as does periodic limb movements of sleep (PLMS). However, SRHJ events were almost associated with cortical arousals that appeared simultaneously or immediately after the head jerk, this phenomenon is different from that of PLMS seen in RLS. A recent study analyzed the clinical features and PSG data of 30 adult patients with REM-HJI >30/h. Among the 30 patients, 25 had main sleep disorders with unaware of head movements during sleep and only five complained of frequent head movements during sleep with sleep disturbance and daytime sleepiness. Likewise, our patient was also unaware of head movements but complained of daytime sleepiness with non-restorative sleep. In addition, frequent SRHJs with arousal still remained on CPAP titration in our case, in contrast to a previous case report for SRHJs, which disappeared on CPAP titration. The authors suggested diagnostic criteria for SRHJ disorder; the criteria are: 1) head jerks confirmed by v-PSG, 2) frequency of SRHJ ≥30/h in REM sleep, 3) SRHJs associated with significant sleep disturbances, and 4) SRHJs and its associated symptoms not explained by another sleep or medical disorder. However, the authors reported a cutoff value with reference to limitations. Therefore, validation studies for cutoff that discriminates normal from pathological conditions are needed, and this value needs to be adjusted for gender and age similar to that with the diagnostic criteria of PLMS.

In our case, the patient complained of repetitive episodes of sleep-related vocalization and DEB; one episode of DEB after head jerks was noted on the first PSG. However, RWA did not fit the EMG criteria for RBD. In addition, the DEB of our patient resolved during CPAP titration. In patients with obstructive sleep apnea, respiratory efforts and/or breathing restart are related to vocalization and motor events, particularly during REM sleep, which can be misdiagnosed as RBD. In our case, the video analysis showed that abnormal motor behavior only occurred during arousal at the end of head jerk event with RERA; head jerks associated with RERA may induce cortical microarousal and provoke DEB. However, in some conditions with sleep-related movements, true RBD may overlap with disorders that may mimic RBD. The differential diagnosis can be difficult if based only on the history. Therefore, in diagnosing RBD, especially in young patients, a step-by-step approach is necessary.

The international RBD study group suggests that prodromal RBD could be defined as the presence of clinical features of evolving RBD with behavioral, neurophysiological aspects or both, which do not conform to the established diagnostic criteria for RBD. Stefani and Högl suggested that SRHJ may be a variant of the excessive phasic muscle activity in REM sleep, and this might be REM parasomnia with a potential overlap with RBD, or SRHJs itself might be a phenomenon of prodromal RBD. Two studies have shown that the frequency of SRHJs is higher in patients with RBD or RWA than in those without RBD or RWA. However, one sampled too few RBD patients; the other study compared clinical and PSG features between patients with an REM-HJI >30/h and patients with isolated
RBD and found no correlation with tonic or phasic EMG activity of REM sleep in patients with SRHJs and those with iRBD. As the duration of the SRHJ (approximately 0.5 s) was longer than that of a cortical myoclonus, SRHJ is more likely to be a movement of subcortical origin. A study suggested that SRHJs involving muscle groups with axial function are a type of subcortical myoclonus; the origin may be located in the medullary reticular formation. In a recent case report, two siblings with a mutation in the alpha-1 subunit of the glycine receptor gene showed frequent SRHJs with hyperekplexia. Glycine is an important inhibitory neurotransmitter in the brainstem, which mediates the inhibitory action of spinal cord interneuron during REM sleep. Further research on the physiological mechanisms of SRHJs based on the glycinergic dysfunction hypothesis are needed. SRHJs and RBD may be due to distinct REM sleep-related disorders or they may have a common underlying pathological mechanism.

SRHJs may belong to the RBD spectrum or represent a distinct sleep-related motor phenomenon. These issues require further investigation. Sleep movements, such as SRHJs, that are associated with REM sleep should be approached carefully and distinguished from RBD, especially in young patients. In addition, symptomatic SRHJ should be categorized as a new sleep-related movement disorder.

Supplementary Video Legend
Video1. Jerking movement of the head during sleep.
Video2. Dream enactment behavior followed by sleep-related head jerk.

Supplementary Materials
The online-only Data Supplement is available with this article at https://doi.org/10.13078/jsm.220023.

Ethics Statement
The study was approval and the need to obtain informed patient consent was waived by the CHA Medical Center Institutional Review Board (IRB approval No.: 2022-11-019).

Conflicts of Interest
The author has no potential conflicts of interest to disclose.

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