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SLEEP MEDICINE PEARLS

A Patient with Rhythmic Movements during REM Sleep

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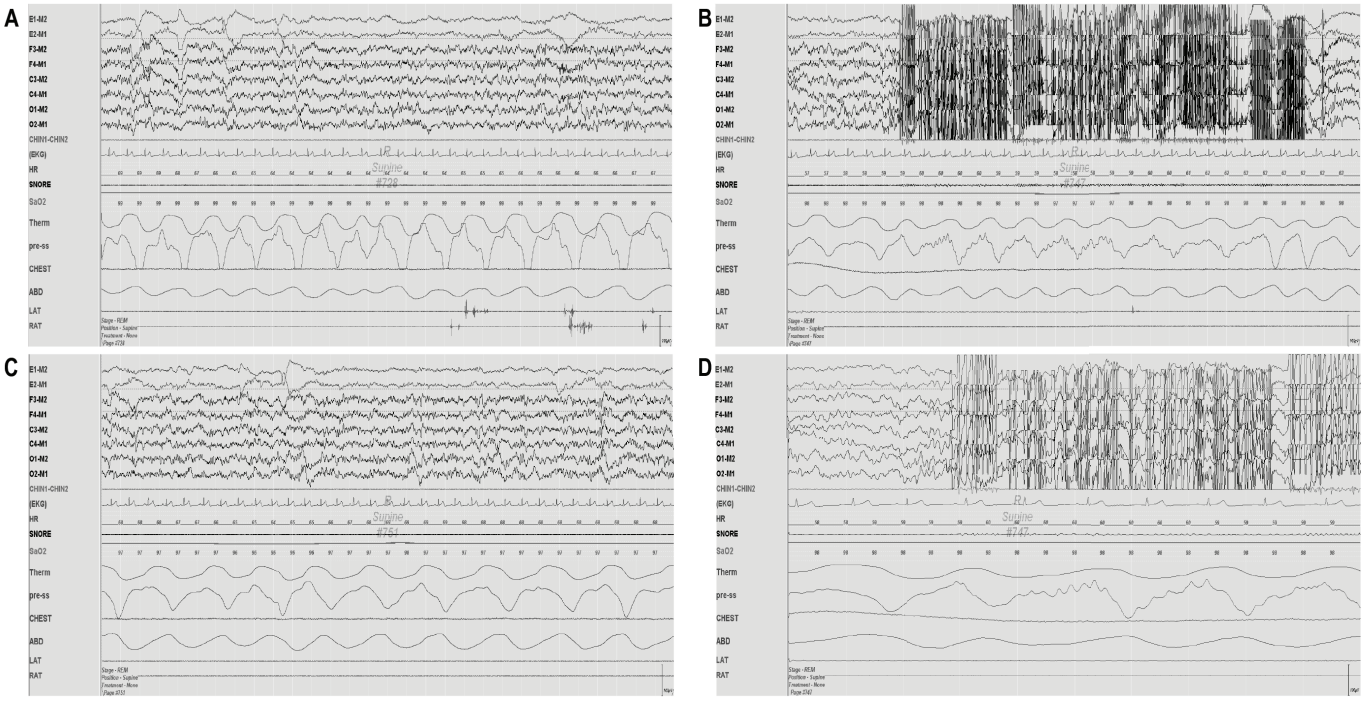
A 29-year-old male with no medical or family history presented with excessive daytime somnolence (Epworth Sleepiness Scale score 22/24) for more than a year, including falling asleep while driving. He endorsed taking frequent naps during the day with sleep paralysis (SP) upon awakening. He reported seeing shadows in his room when falling asleep consistent with hypnagogic hallucinations. The patient also endorsed brief episodes of cataplexy characterized by muscle weakness, such as dropping objects even if held in both hands, and bilateral leg weakness at the knees, triggered by laughter and lasting several seconds. Cataplexy and sleep paralysis were his most distressing symptoms due to recurrent daytime episodes. Furthermore, his wife complained of the patient shaking or twitching while he slept. Physical examination was normal. Cataplexy could not be induced during the clinic visit.

Diagnostic polysomnography (PSG) preceding a multiple sleep latency test (MSLT) showed 80% sleep efficiency and sleep onset of 13.5 min. REM latency was prolonged at 185.5 min, and the patient spent 20% of the night in stage R. Apnea-hypopnea index was unremarkable. On EEG, apparent spikes occurring singly and in semi-rhythmic runs up to 30 s were seen over the left hemisphere during 4 different 10- to 15-minute periods (**Figure 1**). One episode was present in stage N2 and the remainder in REM sleep. Review of the video showed intermittent rhythmic rolling and side-to-side head shaking while the patient lay supine with his head tilted to the left. After awakening, the patient was unaware these movements occurred. On MSLT, the mean sleep latency (MSL) was 15.8 min with no sleep onset REM periods (SOREMPs).

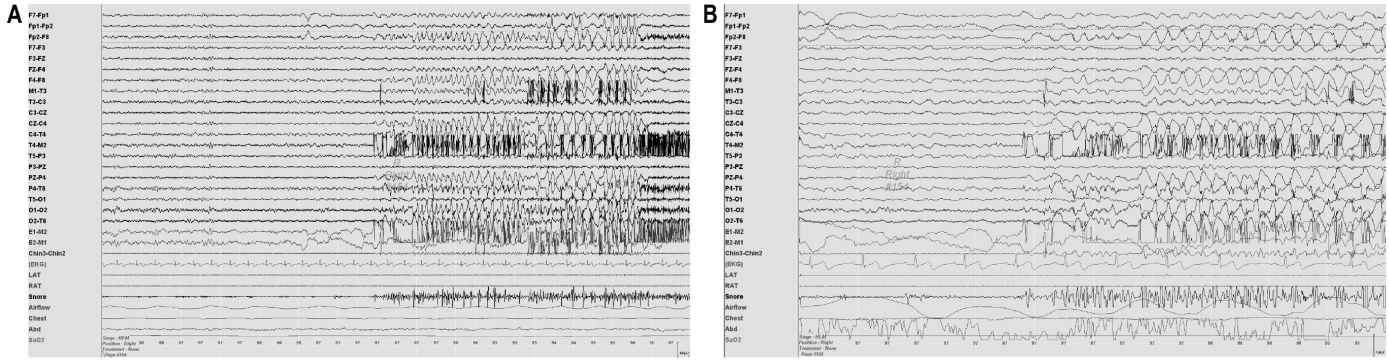
QUESTION: What is your diagnosis?

Figure 1—Initial polysomnography



(A) 30-s epoch with the patient lying supine but with head tilted to his left side showing stage R prior to the start of rhythmic movements. **(B)** 30-s epoch showing stage R during rhythmic movements associated with apparent spikes and lasting approximately 20 sec. **(C)** 30-s epoch showing stage R approximately 2 min after the rhythmic movements in B. Note the absence of arousal or increased slow activity. **(D)** 10-s epoch of rhythmic movements shown in B. Note the apparent spikes are located in the left-sided electrodes (F3, C3, O1). EOG: E1-M2, E2-M1. EEG: F3-M2, F4-M1, C3-M2, C4-M1, O1-M2, O2-M1. EMG: Chin1-Chin2, LAT (left anterior tibialis), RAT (right anterior tibialis). EKG, electrocardiogram; HR, heart rate; SaO2, oxygen saturation; Therm, thermistor; Pres-ss, nasal air pressure sensor; Abd, abdomen.

Figure 2—Full EEG polysomnography



(A) 30-s epoch showing stage R during rhythmic head movements. **(B)** 10-s epoch showing stage R during rhythmic movements. In both A and B, the EEG shows 90-130 μV theta activity with a sinusoidal morphology in right-sided electrodes only (maximal Fp2-F8, F4-F8, Cz-C4, C4-T4, Pz-P4, P4-T6, O1-O2, O2-T6). There is myogenic artifact, primarily in T4-M2. EOG: E1-M2, E2-M1. EEG (transverse montage): F7-Fp1, Fp1-Fp2, Fp2-F8, F7-F3, F3-Fz, Fz-F4, F4-F8, M1-T3, T3-C3, C3-Cz, Cz-C4, C4-T4, T4-M2, T5-P3, P3-Pz, Pz-P4, P4-T6, T5-O1, O1-O2, O2-T6. EMG: Chin3-Chin2, LAT (left anterior tibialis), RAT (right anterior tibialis). EKG, electrocardiogram; SaO2, oxygen saturation; Airflow, thermistor; Abd, abdomen.

ANSWER:
Narcolepsy with cataplexy associated with sleep-related rhythmic movements.

Despite the MSLT findings, narcolepsy with cataplexy (NC) was strongly suspected by history. A repeat MSLT was recommended to the patient, but was not pursued at his request. He was distressed over daily episodes of sleep paralysis and/or cataplexy and did not want to delay treatment despite attempts to provide reassurance. The International Classification of Sleep Disorders second edition (ICSD-2) does not require an MSLT to show MSL < 8 min and 2 SOREMPs or a cerebrospinal fluid (CSF) hypocretin level < 110 pg/mL to diagnose NC if there is a definite history of cataplexy.[1] Both MSLT and CSF hypocretin level are recommended but not required for the diagnosis of NC because neither test provides additional diagnostic value over the clinical history of definite cataplexy.

In a large retrospective study of 2,472 MSLTs,[2] 170 subjects were diagnosed with narcolepsy based on either of the following diagnostic criteria: (1) a complaint of, or history compatible with, excessive sleepiness and definite cataplexy, or (2) MSL < 8 min, ≥ 2 SOREMPs, and no other medical, psychiatric, or other sleep disorder sufficiently severe to account for sleepiness, associated symptoms, and SOREMPs. Of the patients with NC studied with a MSLT, 13% did not have any SOREMPs and 29% did not have MSL < 8 min and ≥ 2 SOREMPs. Further, 6/15 (40%) patients with NC who underwent 2 MSLTs did not have MSL < 5 min or ≥ 2 SOREMPs on either study. The authors concluded that overemphasizing MSLT results "may prevent the diagnosis of narcolepsy in some patients who actually have the diagnosis."[2] CSF hypocretin levels are also recommended but not required by ICSD-2 for the diagnosis of NC, but this test was validated against the diagnostic criteria of a history of cataplexy, the same standard as used in our patient.[3] Based on this evidence, no further evaluation was justified clinically for our patient, and the diagnosis of NC was based on the clinical history of excessive sleepiness and definite cataplexy. He was started on sodium oxybate.

Although the diagnosis of NC was not in doubt, the head rolling movements raised the possibility of a second diagnosis. Classifying nocturnal movements as simple or complex, and if present in either NREM or REM sleep, assists with narrowing the differential diagnosis.[4] Motor activity in REM sleep is not frequent but has been described in REM sleep behavior disorder (RBD) and epilepsy. Furthermore, NC has been associated with RBD, periodic limb movements during sleep, and sleep-related rhythmic movements (SRMs).[5] RBD was not suspected in this patient due to lack of complex movements. While the assumed cause of the EEG abnormalities was movement artifact, the waveform morphology appeared epileptiform-like on the limited EEG montage of the PSG. Further, simple movements associated with interictal spike-wave discharges have been reported during both NREM and REM sleep.[6]

Repeat PSG with full montage EEG using the international 10-20 system showed a normal waking background with good organization and reactivity. Stereotypic head rolling occurred only in stage R with the patient lying on his right side. SRMs were associated with high amplitude 4-5 Hz theta activity throughout the right hemisphere (**Figure 2**) without increased slowing or arousal on EEG. The full montage EEG PSG demonstrated that there was no epileptiform-like activity and that the apparently abnormal waveforms represented movement artifact.

SRMs observed in this case were similar to those seen in sleep-related rhythmic movement disorder (SRMD). SRMD is rarely seen in adults or REM sleep, and must result in sleep impairment and not be explained by another diagnosis.[1] The patient did not meet criteria for SRMD because daytime sleepiness, cataplexy, and SRMs improved after starting sodium oxybate for NC, suggesting the SRMs were related to NC. In a recent report, three patients with NC had intermittent SRMs of the head, legs, and/or body coinciding with stage R and SP on PSG.[7] SRMs in stage N2 were not seen. The authors found that two of three patients recalled initiating the movements to

"shake out" of SP. Treatment for NC led to improvement in the SRMs. A similar phenomenon may account for the findings in this case.

Treatment for NC with sodium oxybate led to improvement in the SRMs for this patient. At 8 months follow-up, the patient was tolerating 3 grams of sodium oxybate twice per night without side effects and endorsed no episodes of cataplexy, rare sleep paralysis, and an estimated 90% reduction in daytime sleepiness (Epworth Sleepiness Scale score 7/24). His wife stated that she no longer observed any nocturnal movements.

CLINICAL PEARLS

1. The diagnosis of narcolepsy with cataplexy may be made based on clinical criteria if there is a definite history of cataplexy. An MSLT and/or CSF hypocretin level are recommended but not required for diagnosis.
2. Motor findings in REM sleep occur infrequently and may be caused by REM sleep behavior disorder, epilepsy, or sleep-related rhythmic movements.
3. Defining abnormal movements during sleep as simple or complex can assist in narrowing the differential diagnosis.
4. Narcolepsy with cataplexy is associated with abnormal movements during REM sleep that can be either simple (e.g., periodic limb movements during sleep or sleep-related rhythmic movements) or complex (e.g., REM sleep behavior disorder).
5. Treatment of narcolepsy with cataplexy may help relieve symptoms of sleep-related rhythmic movements when they coexist.
6. Differentiating seizures from primary sleep disorders may require a full montage EEG PSG.

CITATION

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