A 78-year-old man with a longstanding history of obstructive sleep apnea (OSA) presented to the sleep medicine clinic requesting a new continuous positive airway pressure (CPAP) machine. He had been diagnosed with OSA approximately 20 years ago, after a polysomnogram showed moderate OSA. He had been using the CPAP machine nightly for the past 20 years and felt that it had worked well; however, in the past year he noticed that his sleep had become less restorative and he was experiencing some abnormal sleep-related behaviors.

With regard to his sleep history, the patient reported controlled snoring, choking arousals, and documented apneas on his CPAP device. In the past year, he had experienced a few instances of falling out of bed that appeared to be related to dream enactment. His wife noted that he would yell, kick, and scream while sleeping. He remembered an episode of doing a “karate chop” in a dream that caused him to flail his arm. On occasion, he had swung his arm onto his wife. Gradually, these behaviors became more frequent and disruptive, which led to his spouse sleeping in a separate bedroom.

The patient had also experienced gradual progressive difficulty with cognitive functioning, leading to frustration and occasional angry outbursts. This caused significant discord within his family. Recently, he had begun having episodes of confusion, lasting anywhere from a few minutes to an entire day. During one episode of confusion, he was brought to the emergency department where he was given haloperidol to treat psychosis after preliminary testing. He subsequently became stuporous, leading to hospitalization for further management. After monitoring and detailed diagnostic testing that did not reveal any acute pathology, he was discharged after his mental status improved.

After that discharge home, he continued to have frequent angry outbursts, mood fluctuation, and visual hallucinations. He also experienced constipation, loss of sense of smell, resting limb tremors, and difficulty with initiation of gait.

His physical examination was concerning for turning “en bloc,” postural instability, retropulsion, and decreased arm swing. He was also noted to have a 3- to 5-Hz resting tremor in his right hand and bilateral cogwheel rigidity.

Neuropsychological testing was performed that showed early signs consistent with dementia with Lewy bodies (DLB). A polysomnogram was performed that showed adequately treated OSA and an increase in electromyographic tone during rapid eye movement (REM) sleep associated with jerking of the arms and legs.
The patient was diagnosed with REM sleep behavior disorder (RBD) with concurrent OSA. The RBD predated the development of cognitive symptoms, which is suggestive of a neurodegenerative disorder (in this case DLB), and hence can be considered a prodromal feature.

For the safety of the patient, modifications were made to his bedroom at home. It was recommended that his spouse sleep in a separate bed to avoid injury. Given the safety concerns and after discussion with the patient and his family, he was eager to start pharmacotherapy. He was initially prescribed 3 mg/day of melatonin with a gradual titration up to a maximum dose of 15 mg/day to be taken at bedtime.

The patient returned for follow-up several weeks later and described an improvement in his dream enactment behavior with decreased frequency of episodes.

He was then prescribed donepezil, a cholinesterase inhibitor, which improved his behavioral symptoms. Carbidopa-levodopa was also given, which improved his motor symptoms of tremor and bradykinesia. During longitudinal follow-up, the patient’s depression was addressed with bupropion, which improved his quality of life significantly and allowed him to engage in social activities.

The patient’s family was involved in the discussion about the diagnostic implication of DLB, and the patient’s electronic health record was updated so clinicians would avoid prescribing typical antipsychotics in the future.

**DISCUSSION**

RBD affects approximately 0.38% to 0.5% of people in the United States. RBD is a parasomnia characterized by disinhibition of motor control, facilitating dream enactment behaviors during sleep.

The International Classification of Sleep Disorders, third edition requires all of the criteria listed in Table 1 for a diagnosis of RBD.

The hallmark features of RBD include behaviors during REM sleep that may cause sleep disruption and even injury to the patient or bed partner because these dreams are often action-filled, violent, and unpleasant. An electromyographic recording during an overnight video PSG will demonstrate abnormal increase in muscle tone in the patient during REM sleep.

A thorough clinical sleep history is important in any patient presenting with sleep-related complaints, as RBD may be seen in the setting of an underlying neurodegenerative disorder in older people, in people with narcolepsy, and in younger patients who take antidepressants such as selective serotonin reuptake inhibitors. Because REM sleep typically occurs approximately 90 minutes after sleep onset, the clinical history usually suggests abnormal behaviors in the second half of the night.

RBD is more common in men, and patients typically are older than age 50 years, although cases in younger men have been reported. In patients presenting after the age of 50 years, RBD can represent one of the premotor symptoms predictive of eventual progression to a neurodegenerative condition, notably alpha-synucleinopathies including Parkinson’s disease, DLB, and multiple system atrophy. Conversion rates of 81% have been reported.

DLB is the second most common cause of degenerative dementia. In addition to cognitive impairment, it is characterized by core clinical features of fluctuation of alertness, visual hallucinations, and parkinsonism. Suggestive features include RBD and neuroleptic sensitivity.

The differential diagnosis of RBD includes OSA, sleepwalking, high-magnitude periodic limb movements, nocturnal seizures, dissociative states, and malingering. In some cases, these can be differentiated clinically with the assistance of a reliable collateral history (such as from a spouse), based on temporal occurrence in sleep, the ability of the patient to arouse during the episode, and the association with vivid dreaming. Patients with RBD typically can be easily awakened from sleep during an episode and will often recall the details of the dream; this is not seen with other parasomnias or sleep-related movement disorders. Early detection and treatment should be instituted to prevent injury to the patient and bedpartner. Counseling about risk
of development of a neurodegenerative disorder and yearly surveillance is imperative. This enables the clinician to manage behavioral and psychiatric symptoms in a timely manner, improves quality of life for the patient, and decreases caregiver burden.

CONCLUSION

RBD has a high association with future development of alpha-synuclein neurodegenerative disorder. A meticulous patient history, corroborated by polysomnography, can help distinguish it from other diagnoses by the presence of increase in muscle tone in REM sleep. Safety measures for the patient at home should be instituted promptly along with pharmacotherapy, either melatonin or clonazepam, for symptomatic relief. A detailed history of psychiatric, neuropsychological, and cognitive symptoms along with neurological examination and neuropsychological evaluation should be obtained and the patient observed longitudinally with yearly surveillance for evolution of a neurodegenerative disorder. This allows for early detection and management of symptoms that may have a significant impact on the quality of life for patients and their families.

REFERENCES


TABLE 1.

Diagnostic Criteria for Rapid Eye Movement Behavior Disorder

- Repeated episodes of sleep-related vocalization and complex motor behaviors
- Behaviors are shown by PSG to occur during REM sleep, or based on clinical dream enactment, presumed to occur during REM sleep
- PSG demonstrates REM sleep without atonia
- Disturbance is not explained by another sleep disorder, mental disorder, medication use, or substance use

Abbreviations: PSG, polysomnogram; REM, rapid eye movement. Adapted from the American Academy of Sleep Medicine.