Case Report

Idiopathic REM Sleep Behavior Disorder: A Report on Two Cases with Contrasting Features

Sriniwas Gupta, M. S. V. K. Raju¹, Alka Pawar²

ABSTRACT

REM sleep behavior disorder (RBD) is a rare parasomnia in which persons exhibit uncharacteristic violent behavior, while dreaming. Secondary RBD occurs due to some neurological conditions, psychoactive substance or psychotropic drug use. There are no case reports on idiopathic RBD in India. We report here two cases to underscore the importance of identifying the disease as behavior associated with RBD may be quite serious in nature and might lead to catastrophic consequences.

Key words: Dream enactment behavior, narcolepsy, REM sleep without atonia, sleep related injury

INTRODUCTION

REM sleep behavior disorder (RBD) is fundamentally characterized by REM sleep without atonia, dream enactment behavior (DEB), and sleep-related injury to self or others. As the atonia, which occurs in normal REM sleep is absent in RBD the patient exhibits complex motor behavior in response to dream mentation. The condition was first described by Schenck et al. in 1986.¹ Its essential clinical features include uncharacteristic abnormal vocalizations (grunting, shouting, screaming, swearing etc.); abnormal motor behavior in the form of running, jumping from bed, punching, flailing and kicking; and altered dream mentation, which generally consist of insects or animals or bellicose strangers chasing the dreaming man and he defending against them. Unlike in normal dreams the patient retains memory of the vivid dreams for months or years. There have been instances of catastrophes like international flights getting redirected for emergency landing and brides getting seriously injured on wedding nights because of RBD.²

Secondary RBD occurs in conjunction with neurological conditions (synucleinopathies and narcolepsy), drugs such as selective serotonin reuptake inhibitors, tricyclics and serotonin-norepinephrine reuptake inhibitors, and psychoactive substances.³ Even idiopathic RBD may lead to emergence of synucleinopathies after a lapse of years in 10-81% of cases.¹,⁴,⁵ A prevalence of 0.38-0.5% has been reported in the west,⁶ while in the Asian population the same is reported as 0.04%.⁷ Prevalence of idiopathic RBD in India is not known. In a recent report from India 19.4% of patients with Parkinson’s disease (PD) were found to be having RBD,⁸ which is much less than the figures of 42.6% quoted in the western literature.⁹ To the best of our knowledge, there are no reports from India on idiopathic RBD cases.¹⁰,¹¹

CASE REPORTS

Case 1
A doctor aged 68 presented with history of violent dreams of more than 50 years duration and some strange happenings over the years.

Departments of Psychiatry, Bharati Vidyapeeth University Medical College, Dhankawadi, ¹Peoples College of Medical Sciences, Bhopal, Madhya Pradesh, ²Department of Psychiatry, Maharashtra Institute of Mental Health, Pune, Maharashtra, India

Address for correspondence: Dr. M. S. V. K. Raju
Department of Psychiatry, Peoples College of Medical Sciences, Bhopal, Madhya Pradesh, India. E-mail: msvkraju@rediffmail.com
Anamnesis brought out that he started having vivid dreams of wild animals attacking him at night at 9 years of age. On one night he attacked a person sleeping beside him with a stout stick when he dreamt of an animal attacking him. No further violent episodes occurred until much latter.

When he was 11 he witnessed a murderous assault on his father and developed symptoms of posttraumatic stress disorder (PTSD). Vivid dreams continued along with trauma related dreams and memories. He would shout and scream at night once in a month or so, but exhibited no violence. At about 17, he developed sleep attacks and after a year had a single episode of sleep paralysis associated with visual and haptic hypnogogic hallucinations. Sleep attacks resolved spontaneously after his marriage at 22.

After many years, he consulted a psychiatrist for PTSD symptoms and recovered with treatment but overly aggressive vivid dreams continued. He would now make elaborate plans of defense while actively dreaming but did never act on his plans until he was 55. On one night he clobbered his brother when he saw a wild animal attacking him in dream. His wild screams woke up others and further damage was averted. He continues to get vivid violent dreams, but while dreaming he knew he was dreaming and learned to dismiss them. He continues to lead an active life running his busy practice.

Physical examination and mental status examination revealed no abnormality. He could recall all his dreams in great detail. He was satisfied with the knowledge that he had a sleep disorder and declined to get investigated or to take treatment.

Case 2
A 64-year-old male sought psychiatric consultation for excessive dreams and violent behavior during dreams of 2 years duration.

Onset was sudden while on a train journey. He dreamt of thieves entering into the compartment. Screaming loudly he attacked other passengers. Well-meaning fellow travellers intervened to deescalate the situation. Since then he experienced vivid dreams in profusion at night nearly every day. In the dreams, he would be invariably under attack from strangers. In an effort to drive them away, he would let out wild screams and throw punches, which invariably would land on his wife. Family members thought he was under the influence of an evil spirit at night as he would be his usual docile self during the day. Exorcism brought him no relief. Some of his friends slept by his bed side for some time but retreated after repeated hits. His wife bore his incessant assaults stoically until one day when he delivered powerful blows to her face dislodging four of her front teeth. He felt ashamed and left the house and came for consultation on a friend’s suggestion.

He worked well as a mechanic until his retirement 2 years before. After retirement he remained active doing social work in his housing society. He was a teetotaler and had no medical illness or fits. Physical examination revealed no abnormality clinically. Mental status examination revealed a well groomed person who talked fluently. Cognitive functions were intact. He could recall all his dreams in great detail with appropriate expression of feelings. Hematological parameters, biochemical parameters were within normal range. Awake electroencephalography showed no seizure activity. He declined to undergo polysomnography as he thought it was not necessary. Magnetic resonance imaging of the brain revealed no abnormality.

Response to treatment with clonazepam 1 mg and melatonin 3 mg at night was quick: by the end of 2nd week, he was free from all violent dreams but reported “normal dreams” which he could not recall. At 1 year follow-up, he continues to remain asymptomatic.

DISCUSSION

Though RBD is a disorder of the elderly male and generally associated with frequent DEB, widely variable frequency of DEB and child hood onset with narcolepsy is not unknown. The first case had onset in child hood and violent dreams continued for over 50 years with sparse DEB. The patient suffered narcolepsy, which resolved spontaneously. The disease appears to have made him vulnerable to PTSD as two of his siblings exposed to the same traumatic scene did not suffer from PTSD. The longest period of conversion to any synucleinopathy after the onset of RBD that is reported in literature is 29 years. Narcolepsy in this case occurred after a lapse of nearly 10 years and resolved spontaneously; the man himself is 68 now, endured RBD for well over 50 years and enjoying robust health. We therefore would like to consider this case as idiopathic RBD. The long period of dreaming appears to have enabled the individual to adapt to RBD.

The second case presented with classical features and responded well to pharmacotherapy. As conversion to any of the synucleinopathies (PD, corticobasal degeneration, Lewy body dementia, and multiple system atrophy) can occur even very late (mean age 71.9 ± 6.6) in 81% of cases the diagnosis of idiopathic RBD in the second case may well have to be revised later. The only advantage of the large gap between onset of RBD and onset of synucleinopathies is that it offers
Neurobiology of RBD is not yet elucidated well. In normal sleep motor tone and elaborate motor activity is known to be inhibited by the activity of the cholinergic pedunculo pontine nucleus (PPN), which through the medullary magnocellular reticular formation and ventrolateral reticulospinal tract influence the anterior horn cells of spinal cord.[13] Some as yet unidentified degenerative condition of the brain stem probably causes RBD. Recently, Albin et al. demonstrated, by DTBZ-PET, reduced dopamine binding in substantia nigra (SN) in idiopathic RBD patients.[14] SN has reciprocal connections with PPN.[14] This lends further support to the view that RBD is a marker for emerging synucleinopathies. Clonazepam up to 4 mg at bed time or melatonin up to 12 mg 30 min before bed time is recommended along with protective security measures and psychoeducation to treat this condition.[13] Carbamazepine, quetiapine, pramipexole, and levodopa also reported to be useful. Sodium oxybate 3G at bed time and 3G after 6 h was reported to be effective.[2]

Going by the lack of reports, RBD in general and idiopathic RBD in particular appear to be extremely rare in India, but considering the recent report from India[8] RBD may not be as uncommon as it appears to be but apparently escaping notice because of infrequent DEB. However, DEB when it does occur can be extremely dangerous as brought out in the cases illustrated here. It is also possible that DEB is more common in idiopathic RBD than secondary RBD. Due to lack of awareness some cases may even be getting treated as psychoses. Screening instruments are now available to identify the disease.[13] The natural history of various forms of RBD, its clinical features and epidemiology merit serious study.

REFERENCES

1. Schenck CH, Bundlie SR, Ettinger MG, Mahowald MW. Chronic behavioral disorders of human REM sleep: A new category of parasomnia. Sleep 1986;9:293-308.
2. Boeve BF. REM sleep behavior disorder: Updated review of the core features, the REM sleep behavior disorder-neurodegenerative disease association, evolving concepts, controversies, and future directions. Ann N Y Acad Sci 2010;1184:15-64.
3. Schenck CH, Mahowald MW. REM sleep behavior disorder: Clinical, developmental, and neuroscience perspectives 16 years after its formal identification in SLEEP. Sleep 2002;25:120-38.
4. Postuma RB, Gagnon JF, Montplaisir JY. REM sleep behavior disorder: From dreams to neurodegeneration. Neurobiol Dis 2012;46:553-8.
5. Ohayon MM, Caulet M, Priest RG. Violent behavior during sleep. J Clin Psychiatry 1997;58:369-76.
6. Lam SF, Zhang J, Tsao J, Ho CKW, Mok V, et al. RBD in psychiatric population. J Clin Psychiatry 2010;71:1101-3.
7. Vibha D, Shukla G, Goyal V, Singh S, Srivastava AK, Behari M. RBD in Parkinson’s disease: A clinical case control study from North India. Clin Neurol Neurosurg. 2011;113:472-6.
8. Poryazova R, Oberholzer M, Baumann CR, Bassetti CL. REM sleep behavior disorder in Parkinson’s disease: A questionnaire-based survey. J Clin Sleep Med 2013;9:55-9A.
9. Chokroverty S. Overview of sleep & sleep disorders. Indian J Med Res 2010;131:126-40.
10. Shah N, Bang A, Bhagat A. Indian research on sleep disorders. Indian J Psychiatry 2010;52:S255-9.
11. Nevsimalova S, Prihodova I, Kemlink D, Lin L, Mignot E. REM behavior disorder (RBD) can be one of the first symptoms of childhood narcolepsy. Sleep Med 2007;8:784-6.
12. Schenck CH, Boeve BF, Mahowald MW. Delayed emergence of a Parkinsonian disorder or dementia in 81% of older men initially diagnosed with idiopathic rapid eye movement sleep behavior disorder: A 16-year update on a previously reported series. Sleep Med 2013;14:744-8.
13. Rye DB. Contributions of the pedunculopontine region to normal and altered REM sleep. Sleep 1997;20:757-88.
14. Albin RL, Koenne RA, Chervin RD, Consens FB, Wernette K, Prey KA, et al. Decreased striatal dopaminergic innervation in REM sleep behavior disorder. Neurology 2000;55:1410-2.
15. Trotti LM. REM sleep behaviour disorder in older individuals: Epidemiology, pathophysiology and management. Drugs Aging 2010;27:457-70.