CASE REPORT

Pediatric Narcolepsy Masquerading as Acute Insomnia

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Abstract

Narcolepsy, a disorder of the borderline between wakefulness and sleep is rarely diagnosed in the pediatric population. This is despite the fact that majority of narcoleptic signs and symptoms begin in the second decade. Misdiagnosis and delayed diagnosis inadvertently leads to increased social and economic burden on the children and their families with interference in normal mental and physical well being and academic performance. The commonest cardinal symptom of narcolepsy is excessive daytime sleepiness. However, patients with narcolepsy have significantly disturbed sleep patterns and may have associated mood disorders. This report describes a child presenting with acute insomnia who was found to have narcolepsy.

Keywords: Narcolepsy, insomnia, paradoxical insomnia, early morning awakening, sleep perception

Introduction

Narcolepsy is a chronic neurological disorder associated with hypofunctioning of the hypocretin neuropeptide system leading to rapid eye movement (REM) sleep intrusion into wakefulness[1]. It effects 1 in 2000 population without significant differences between males and females[1]. Onset of majority of narcolepsy is between 15-30 years of age, 30% prior to 15 years, 16% before 10 years and 4% before 5 years of age[2]. however, only 4% are diagnosed prior to 15 years. Considering the protean manifestations of childhood narcolepsy, the inadvertent delays in establishing early diagnosis cannot be undermined. We report a child presenting with acute paradoxical insomnia as the only symptom of narcolepsy.

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Report of Case

A 15-year-old student of presented with sleep disturbance for 2 months with short sleep latency (5-10 minutes), early morning awakening and profuse dreaming. The sleep diary showed sleep duration of 3-5 hours. He had difficulty concentrating on studies, heaviness of head and considerable daytime fatigue though he denied daytime sleepiness. He was irritable, had anger outbursts and complained of poor memory causing him to stop attending school. There were no symptoms of restless legs syndrome, snoring, choking, apneic spells, sleep paralysis, hallucinations or cataplexy. There was no tremor, palpitations, heat or cold intolerance, change in appetite or weight over this period. The boy denied any cause for anxiety. Apart from his elder brother suffering

from a personality disorder with increased sleep latency and aggressiveness, family history was noncontributory. The child had not been on any medications for sleep or other illness. He had body mass index of 19.1, pulse rate 108/minute and blood pressure 120/70 mm Hg and was quiet and withdrawn with decreased affect. Epworth Sleepiness Scale (ESS) score was 9/24, Hospital Anxiety and Depression Scale score 27 out of 42, suggesting severe anxiety and depression and DS14 questionnaire 4 revealed Type D personality with high negative affectivity and social inhibition. Biochemical tests showed TSH- 3.47 (0.2-5.1 µIU/mL), Vitamin B12-153 (180-914 pg/mL), Vitamin D- 7.38 (30-150ng/mL) and ferritin- 29 (22-322 ng/mL). Polysomnography showed short sleep latency (0.5 minutes), good sleep efficiency (97%), early onset REM sleep (latency 18.0 minutes) with relatively increased REM sleep (24.5%) (Figure 1A). Increased arousals, mainly spontaneous (arousal index 17.9) were noted without obstructive sleep apnea. Patient conveyed he had not slept throughout the polysomnography suggesting possibility of paradoxical insomnia. Multiple Sleep Latency Test (MSLT) was able to clinch the diagnosis of narcolepsy with short sleep latency (average 2 minutes 22 seconds)in all 4 naps and sleep onset REM (SOREM) in 3 of 4 naps with average REM latency of 5 minutes 20 seconds (Figure 1B). The major marker HLA-DQ B1*06 allele was negative, and other HLA markers were not tested for due to poor affordability. Also, testing for hypocretin levels in cerebrospinal fluid is not currently available in India. With a diagnosis of narcolepsy Type 2, he was treated with amitryptiline and armodafinil as associated features of anxiety and depression were present. Sodium oxybate is not available in India. After initial improvement for a month there was re-occurrence with inability to tolerate armodafinil. After shifting to escitalopram with sleep scheduling, day naps and behavioural therapy, overall improvement was noted after 7 months.

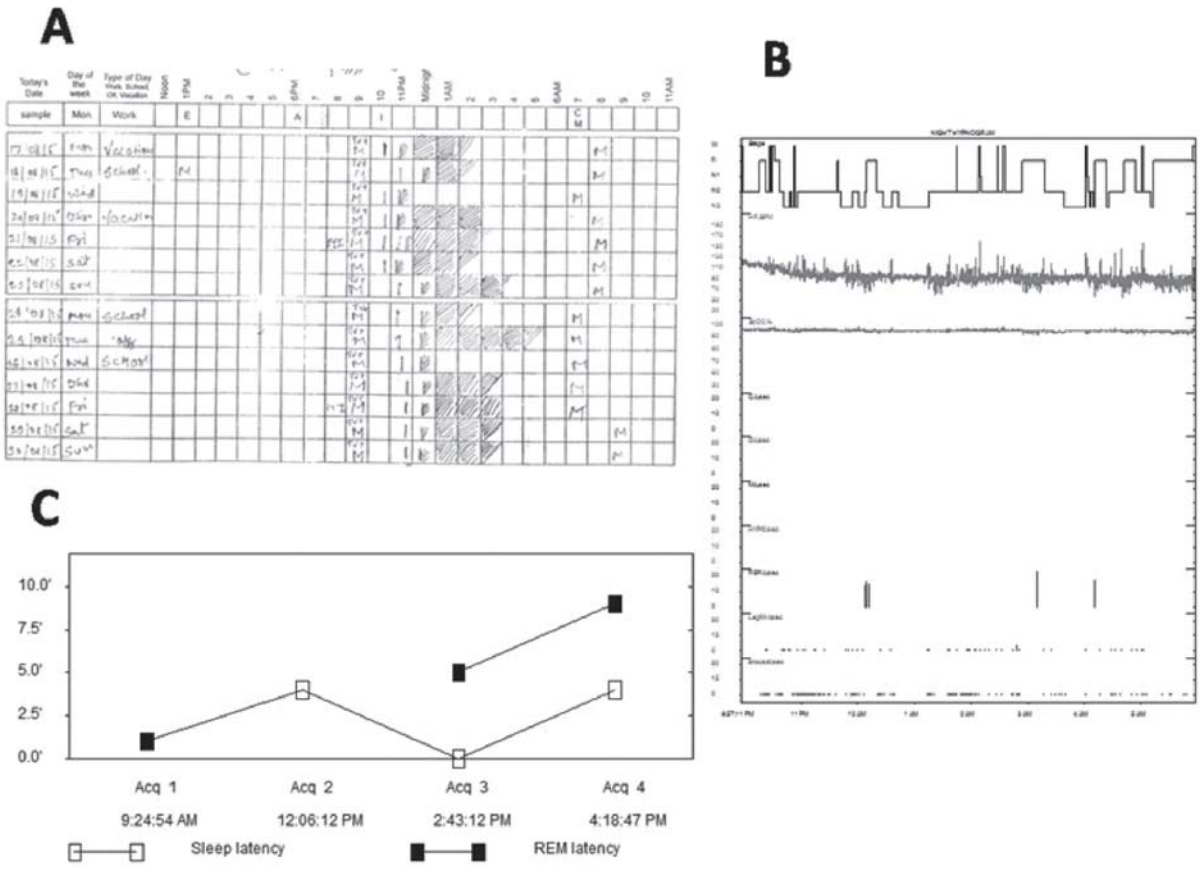

Figure 1: A. Sleep diary demonstrating poor perception of nocturnal sleep; B. Hypnogram showing good sleep efficiency contrary to patient's perception, short REM latency and increased REM duration; C. MSLT showing sleep onset REM in 3 out of 4 naps

Discussion

The diagnosis of childhood narcolepsy is often fraught with delayed diagnosis or mis-diagnosis with patients visiting multiple centers for their complaints. Though it is known to have a bimodal distribution, it is rarely diagnosed before adulthood[1,2,5]. Experience from various sleep centers have shown that symptoms occur earlier than generally expected[5,7]. The classical tetrad of excessive daytime sleepiness, sleep paralysis, hypnagogic hallucinations and cataplexy is rarely seen in childhood narcolepsy. On the other hand, these children present with behavioural and emotional disturbances and end up being misdiagnosed as psychiatric disorder[6,8]. Patients may also remain undiagnosed and be improperly treated during the formative years.

The frequency of principal symptoms of narcolepsy are excessive daytime sleepiness (100%), cataplexy (70%), hypnagogic hallucinations (25%) and sleep paralysis (5%)[5]. While patients usually do not manifest all 4 cardinal symptoms, hypersomnolence is the commonest[6]. Our index case represents insomnia as a rare presenting feature of narcolepsy. This is contrary to the accepted concept that patients with narcolepsy sleep a lot. This case exemplifies that their sleep is actually of poor quality resulting in inappropriate sleepiness during the daytime. Early morning awakening, disturbed sleep with vivid dreaming and unrefreshing sleep is common. This disconcertingly unrefreshing sleep was actually perceived as sleep maintenance insomnia by our patient. However, polysomnography revealed short sleep latency, good sleep efficiency with increased arousals and REM sleep suggesting possibility of sleep state misperception in addition. The sleep disturbance can therefore easily be confused by the patient and family members as insomnia specially in the acute stage. It is not unusual that insomnia be confused with narcolepsy. Both patients have disrupted night sleep with disturbing dreams. However, mechanisms of dream recall may be different in both.

The secondary fallout of insomnia, though paradoxical was anxiety and depression, fatigue and impact on school attendance and academic performance. Patients may use the term 'fatigue' interchangeably with excessive daytime sleepiness. While excessive daytime sleepiness is defined as "the inability to maintain wakefulness and alertness during the day with sleep occurring unintentionally or at inappropriate times almost daily for at least 3 months", fatigue may also lead to reduced waking function.1,5 Also, a sleepy narcoleptic brain may lead to mood disturbances, cognitive dysfunction, difficulty focusing and attention deficit disorder.

This spectrum of symptoms specially in adolescents may easily lead to diagnosis of depression and personality disorder as seen in our patient. He also scored poorly on HADS and had Type D personality suggesting that the sleep disturbance also secondarily influences the psychiatric profile of an individual. In fact, one third of narcoleptics may have mood disorders[9]. Hypocretin, that effects the balance between monoamines and acetylcholine, may influence both narcolepsy and affective disorder. Insomnia and sleep fragmentation is also a common association of 90% of psychiatric disorders. Short REM sleep latency and increase in REM sleep are also markers of depression as well as narcolepsy. The additional relief of symptoms in our patient with use of antidepressants which are REM suppressants should not be interpreted as confirmation of mood disorders in a patient with true narcolepsy. Therefore, it was understandable that our patient presented with primary concerns regarding lack of sleep (and not hypersomnia)as well as disorder of affect. Only the MSLT was able to clinch the diagnosis of narcolepsy. The major marker HLA-DQ B1*06 allele was negative, and other HLA markers were not tested for due to poor affordability. Also, hypocretin levels in cerebrospinal fluid is not currently available in India.

Conclusion

Narcolepsy should be evaluated for in patients, especially children and adolescents, presenting with complaints of nonrestorative sleep or overwhelming daytime fatigue. Insomnia, specially sleep maintenance insomnia, early morning awakening and paradoxical insomnia, may be a rare symptom of narcolepsy.

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