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# Monosymptomatic Presentation of Narcolepsy in Adolescents and Young Adults

R. Namitha, DNB,<sup>a</sup> and N. A. Uvais, MBBS, DPM<sup>b,\*</sup>

Excessive sleepiness can be found in up to 20% of school-aged children and adolescents.<sup>1</sup> A significant number of adolescents with excessive sleepiness might be suffering from narcolepsy, a chronic sleep disorder that causes a dysregulation of the sleep-wake cycle that affects approximately 0.05% of the population and is often characterized by daytime somnolence, cataplexy, sleep paralysis, and hypnagogic hallucinations.<sup>2</sup> The literature suggests that most of these patients either do not consult a physician due to poor awareness or are misdiagnosed with another neurologic or psychiatric disorder due to failure of the physician to specifically request an investigation of sleep disorders and the lack of awareness of the physician regarding the presentation of narcolepsy in this age group.<sup>3</sup> Moreover, the phenotypic presentation of narcolepsy in children and adolescents is highly variable, and the disorder often presents with only one cardinal symptom, making the diagnosis difficult in this age group.<sup>1</sup> The diagnosis of narcolepsy was determined, on average, 10–15 years after the first symptoms appeared.<sup>4</sup> There are very few narcolepsy cases reported from India, especially among adolescents and young adults. Here, we report the case of a male patient who presented with excessive sleepiness who was diagnosed with narcolepsy based on polysomnography (PSG) findings.

## Case Report

An 18-year-old man presented to the psychiatry outpatient services of our hospital with complaints of excessive daytime sleepiness of approximately 1 year duration. He reported that despite a good night's sleep, he started sleeping for 4 to 5 hours during the daytime, with total sleep duration of 12 to 14 hours/day. If not woken, he would continue to sleep further. Because of continued excessive daytime sleepiness, the patient was unable to study, which led to academic decline. He could barely concentrate on activities. Gradually,

he started becoming irritable with family members about trivial issues. There was no history suggestive of sleep attacks, sleep paralysis, hallucinations, or cataplexy. There was no past or family history of any psychiatric illness. There was no history suggestive of hypothyroidism, substance abuse, or seizure disorder and no family history of any sleep disorders. His routine investigations including thyroid function tests were within normal limits. However, he gave a history of occasional snoring while sleeping. Considering his history of snoring and obesity (body mass index = 30.9 kg/m<sup>2</sup>), an overnight PSG was done. The total duration of sleep evaluated was 391 minutes. The report ruled out obstructive sleep apnea as a cause for excessive daytime sleepiness (Apnea Hypopnea Index = 1.4). Sleep latency was 0.9 minutes. The Multiple Sleep Latency Test (MSLT)<sup>5</sup> was performed the next day, 3 hours after waking up in the morning with a 2-hour interval until the patient was given 4 opportunities to nap and monitored for at least 15–20 minutes after the onset of sleep. The MSLT revealed the onset of rapid eye movement (REM) sleep within 9 minutes and 3 sleep onset REM periods (SOREMPs). Based on the PSG findings, he was diagnosed with narcolepsy, and oral armodafinil 250 mg/d was started.

## Discussion

The diagnosis of narcolepsy was established in our case based on excessive daytime sleepiness, short REM latency, short mean sleep onset latency (0.9 min), and 4 SOREMPs during an MSLT. Diagnosis of narcolepsy in children and adolescents is a challenge. The variability of clinical presentation, low prevalence rate, and comorbid psychiatric symptoms often make the diagnosis difficult. Literature suggests that most of the children and adolescents who presents to physicians with narcolepsy symptoms are often misdiagnosed and exposed to various anticonvulsant, antipsychotic, and antidepressant medications.<sup>3</sup> Monosymptomatic presentation of narcolepsy among this age group also leads to misdiagnosis and inappropriate treatment. Our case highlights the need to properly investigate adolescents and young adults who present with excessive sleepiness with overnight PSG to diagnose narcolepsy early in its course so that appropriate early treatment, both pharmaceutical and supportive, can be initiated, which can have meaningful effects on quality of life, socialization, and school performance.<sup>6</sup>

There are very few cases of narcolepsy reported from India. After an extensive literature search, we found 1 observational study with a sample size of 20, 1 case series of 4

<sup>a</sup>Department of Pulmonology, Iqraa International Hospital and Research Centre, Calicut, Kerala, India

<sup>b</sup>Department of Psychiatry, Iqraa International Hospital and Research Centre, Calicut, Kerala, India

\*Corresponding author: N. A. Uvais, MBBS, DPM, Department of Psychiatry, Iqraa International Hospital and Research Centre, Malaparamba, Calicut, Kerala 673009, India (druvaisna@gmail.com).

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Table 1. Cases of Narcolepsy Reported From India

Authors	Type of Study	Age at Onset (y)	BMI (kg/m <sup>2</sup> )	Excessive Daytime Sleepiness	Cataplexy	Hypnagogic Hallucinations	Sleep Paralysis	Sleep Attacks	Arousal Index (events/h)	Sleep Latency (min)	REM Latency (min)
Gupta et al <sup>2</sup> (n=20)	Observational	Mean ± SD: 25 ± 10	Mean ± SD: 26 ± 4	All	12/20 (60%)	8/20 (40%)	12/20 (60%)	All	Mean ± SD: 18.7 ± 10.3	Mean ± SD: 1.3 ± 0.3	Mean ± SD: 2.5 ± 0.6
Bhatia and Arif <sup>9</sup> (n=1)	Case report	NA	26.5	Yes	Yes	No	No	Yes	NA	0.46	NA (short)
Shehna et al <sup>10</sup> (n=1)	Case report	30	NA	Yes	Yes	Yes	Yes	Yes	NA	0.25	5
Gupta et al <sup>8</sup> (n=1)	Case report	35	26	Yes	Yes	Yes	Yes	Yes	NA	0.7	8
Panda <sup>11</sup> (n=1)	Case report	56	28.6	Yes	Yes	Yes	No	Yes	NA	1.4	NA (short)
Ray et al <sup>12</sup> (n=4)	Case series	Mean ± SD: 26.2 ± 6.4	Mean ± SD: 24.2 ± 4.7	All	1/4 (25%)	1/4 (25%)	1/4 (25%)	All	38.5 (range, 14.5–61.9)	Mean ± SD: 2.7 ± 1.3	Mean ± SD: 5.7 ± 2.9
Gupta et al <sup>3</sup> (n=1)	Case report	16.5	NA	Yes	Yes	Yes	Yes	Yes	NA	NA	NA
Swarnakar and Deotare <sup>7</sup> (n=1)	Case report	12	17.5	Yes	Yes	Yes	Yes	Yes	NA	1.5	7
Present study	Case report	18	30.9	Yes	No	No	No	No	29	0.9	9

Abbreviations: BMI = body mass index, NA = not available.

patients, and 6 reports of individual cases (Table 1).<sup>2,5,7–12</sup> Among the case reports, only 2 were regarding adolescent patients with narcolepsy. Gupta et al<sup>3</sup> described the case of an 18-year-old man with narcolepsy with illness onset at 16.5 years of age, who presented with all the cardinal features of narcolepsy. However, no PSG was done in this case. Swarnakar and Deotare<sup>7</sup> described the case of a 15-year-old girl with narcolepsy with illness onset at 12 years of age, who presented with all the cardinal features of narcolepsy. Her PSG findings showed sleep latency of 1.5 minutes and REM latency of 7 minutes.<sup>6</sup> Our case differs from the above 2 cases in the monosymptomatic presentation.

The incidence of narcolepsy usually peaks in adolescence. However, we found only 2 adolescent patients with narcolepsy reported from India. Based on the lack of literature, it can be argued that the prevalence of narcolepsy in India is lower than that in the Western world. However, it is also important to consider the possibility that a significant number of adolescent patients who present with a single symptom at the onset, like in our case, may go undiagnosed due to lack of awareness among patients, lack of awareness among physicians regarding such variable presentations, and lack of facilities for overnight PSG evaluation in most of the centers.

In conclusion, monosymptomatic presentation of narcolepsy is common in children and adolescents. Clinical suspicion and overnight PSG along with MSLT can help in the early diagnosis of narcolepsy in this age group.

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