Case Report

Narcolepsy type 1

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ABSTRACT

Although being the second most common cause of disabling daytime sleepiness in the world, narcolepsy in India is seldom diagnosed. So far, only two cases had been reported. This is the case of narcolepsy in a 15-year-old female student struggling in her scholastic pursuit because of the disease.

KEY WORDS: Cataplexy, Epworth Sleepiness Scale, multiple sleep latency test, narcolepsy

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INTRODUCTION

Narcolepsy is a clinical syndrome of daytime sleepiness with cataplexy, hypnagogic hallucinations, and sleep paralysis. Narcolepsy type 1 is narcolepsy with cataplexy and type 2 is without cataplexy. Narcolepsy type 1 has a prevalence of 25–50/100,000 people according to the Western Data.[1] Report of such cases from India are scarce in the literature.[2] Here, we have described a 15-year-old slim, tall, girl student who presented with narcolepsy type 1 and her improvement with the use of armodafinil.

CASE REPORT

A teenage school girl was referred for evaluation of excessive daytime sleepiness for the past 3 years. She was mainly concerned about the uncontrollable urges of sleep during lectures. She is an eleventh-grade meritorious student. She had to attend 5 h of lectures, 6 days a week. Apart from lectures, she studies on her own 5 h/day, as she is preparing for medical entrance examination. She had been in this curriculum for the past 3 months. Due to her sudden urges of sleep, she could not attend her class with 100% concentration. Sometimes, she even attended the class while standing so as not to get bout of sleep, but standing also did not improve her problem. She even fell asleep during standing, leading to fall on the bench.

Sleepiness was first noted during her ninth grade, which gradually worsened over a time span of 3 years. Daily, she has refreshing sleep of 7–8 h at night without snoring or awakening. Before sleep, she studies book for 3 h. The patient had no history of coffee consumption or excessive television watching before sleep. She could not wake up on her own, and daily somebody had to wake her up. Although she felt refreshed after waking up in the morning, by about 3 h later, she felt excessive sleepiness for a brief time in the lecture. On an average, she used to get 2–3 urges of excessive sleep per day. She generally takes 1-h nap in the afternoon. Sometimes, she had noticed a shadow or an image in the room during falling sleep. After waking up, she had a transient weakness and tingling sensation in her arms and legs. Her movements became slow and clumsy just after recovering from sleep. The patient could not lie down for 5 min without falling sleep. The patient had also experienced several episodes of forgetfulness, which was affecting her academic performance. The patient did not complain of any snoring problem. She had not experienced any weakness or tingling sensation with emotional outbursts. She does not drink coffee or tea.

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Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/lungindia.lungindia_389_15

How to cite this article: Swarnakar R, Deotare A. Narcolepsy type 1. Lung India 2019;36:337-9.

She denied the use of illicit drugs, smoking, and alcohol. She was otherwise healthy and was taking no medication. There was no history of flu vaccination. There was no history of seizures or any psychotic disorder. There was no family history of excessive sleepiness.

The patient was evaluated by the Epworth Sleepiness Scale (ESS)[3] Questionnaire. Her ESS score was 19. The physical examination was unremarkable. She weighed 48 kg with a body mass index of 17.5. Full night polysomnography (PSG) was done by Philips Alice 6 LDE 16 channel polysomnogram using Sleepware G3 diagnostic software. The total duration of sleep evaluated was of 437 min. The report ruled out obstructive sleep apnea as a cause for excessive daytime sleepiness. Reduced sleep efficiency of 65% was seen with the presence of spontaneous awakenings and the onset of rapid eye movement (REM) sleep of 1.5 min.

Multiple sleep latency test (MSLT) was performed on the very next day, 4 h after waking up in the morning with 2-h interval until the patient was given four opportunities to nap and monitored for at least 15–20 min after the onset of sleep. The MSLT revealed the onset of sleep within 2 min. All the four monitored naps revealed onset of REM sleep within 7 minutes of going to sleep. Magnetic resonance imaging of the brain was normal.

The patient was started on treatment with armodafinil 150 mg OD at morning. We counseled the patient on sleep hygiene and told her to get at least 8 h of sleep at night and take a short afternoon nap of 15–30 min. After 2 months of treatment and improving her sleep hygiene, the patient had significantly improved wakefulness, memory, and attention. ESS was again administered, and the score came down to 7.

DISCUSSION

Narcolepsy is more commonly diagnosed in adolescence as in our case of 15-year-old girl student. She was struggling with the symptoms of narcolepsy while studying for competitive exams during her most important year which decides the future of her career. When she sought consultation from various doctors about her predicament and till she presented to us, she was labeled as a student disinterested in studies to malingerer, signifying lack of awareness about narcolepsy among general practitioners and consultants alike. She had to bear scolding from her teacher for frequent dozing off during lecture while being a subject of ridicule among the peers to the extent that she at one time decided to end life because of frequent embarrassment and guilt of not able to cope up with the study because of the symptoms she suffered of narcolepsy. She was also erroneously referred for a psychiatric evaluation. Her first symptom as is seen in all narcolepsy cases was an excessive unpredictable urge to sleep also called as "sleep attack." Later she started having hypnogogic hallucination in class and awakened with weakness in limbs, signifying sleep paralysis, which justified our diagnosis of narcolepsy.

Obstructive sleep apnea was ruled out by a normal Respiratory Disturbance Index of <5/h;[4] AHI of 4 with no episodes of snoring or oxygen desaturation was recorded in a sleep study. MSLT done after PSG in the preceding night for a minimum of 6 h of sleep clinched the diagnosis, recording a sleep latency of <8 min and >2 sleep onset REM periods.[5] No evidence of any parasomnia was witnessed in our patient. Nocturnal sleep disturbance as was witnessed in our patient is usually present in a patient of narcolepsy with cataplexy and not seen in a subject without it. Two other tests with very limited applicability are human leukocyte antigen (HLA) typing[6] (HLA DQB1*0602 And HLA DQA1*0102) and measurement of cerebrospinal fluid hypocretin level,[7] which were not done in our patients.

Managing narcolepsy is a combination of nonpharmacologic and pharmacologic therapy. Maintaining good sleep hygiene, avoiding sedatives, and psychological support constitute nonpharmacologic approach. Modafinil and its active R-enantiomer armodafinil can be considered as the first line of pharmacologic treatment.[8] Apart from children, these drugs have a safe, side effect profile. In children, modafinil can rarely cause Stevens–Johnson Syndrome, so it should be avoided in them.[8] In the past, stimulants and antidepressants had played a major role in management, but due to their serious cardiovascular side effects and the emergence of newer, safer drugs, these classes now constitute a second line of treatment. Sodium oxybate is very effective in treating symptoms of typical cataplexy.[8]

We conclude that more awareness about narcolepsy among the doctors in India is needed to diagnose narcolepsy early and prevent miseries suffered by undiagnosed patients, as was in our case. While OSA is still the most common cause of excessive daytime sleepiness, narcolepsy should always be kept in mind, especially in young nonobese patients. With increased awareness about narcolepsy, self-help or support group to help patients will go a long way in managing narcolepsy.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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