

Case Report

Kleine-Levin Syndrome and Idiopathic Hypersomnia: Spectrum Disorders

Ravi Gupta, Vivekananda Lahan, Malini Srivastava¹

ABSTRACT

Kleine–Levin syndrome (KLS) and idiopathic hypersomnia (IH) are primary sleep disorders of unknown etiologies, which often run a chronic course. The common core symptoms of these syndromes are hypersomnolence and sleep drunkenness, with periodic hypersomnolence and hyperphagia being the prominent symptoms of KLS. Psychiatric manifestations are common to both and include irritability, depression, apathy, inattention and poor concentration. Both disorders are diagnosed clinically and no specific laboratory investigation is available to confirm the diagnosis. We present a case highlighting the overlapping of the symptoms of KLS and IH, producing a complex clinical picture.

Key words: *Hyperphagia, hypersomnolence, idiopathic hypersomnia, Kleine–Levin syndrome, periodic hypersomnolence*

INTRODUCTION

Kleine–Levin Syndrome (KLS) is characterized by recurrent hypersomnia and often accompanied by additional symptoms like megaphagia, hypersexuality and cognitive disturbances.^[1] This disorder of uncertain etiology has a male preponderance and often manifests during adolescence. KLS usually runs a benign course with complete recovery without any long-term consequence.^[2] On the other hand, idiopathic hypersomnia (IH) is characterized by excessive sleepiness, increased nighttime sleep, long and non-refreshing naps, difficulty in awakening from sleep, sleep drunkenness with psychiatric and neuro-vegetative symptoms and a positive family history.^[3] Like KLS, IH

also manifests during adolescence or early adulthood.^[4] Unlike KLS, IH usually runs a chronic course, although complete remission of symptoms has been reported in a few patients.^[4] Diagnosis of IH is difficult due to lack of specific diagnostic marker, and hence made by excluding other causes of excessive daytime sleepiness.^[5] The diagnostic challenges posed due to overlapping of symptoms of KLS and IH are highlighted in the following case.

CASE REPORT

A 9-year-old boy presented with complaints of excessive duration of sleep, increased appetite, weight gain, excessive daytime sleepiness, loss of interest in sports activities, irritability and snoring since 1.5 years after he was shifted to a residential school.

Before attending residential school, he used to follow a regular sleep schedule, with bedtime at 10 p.m. He did not have any issue with sleep onset and used to sleep alone in the bed. According to his father, he used to sleep in supine position and did not show any sign of sleep-related breathing disorder. His mother used to

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Departments of Psychiatry, ¹Psychology and Holistic Medicine, Himalayan Institute of Medical Sciences, Jolly Grant, Dehradun, Uttarakhand, India

Address for correspondence: Dr. Vivekananda Lahan

Department of Psychiatry, Himalayan Institute of Medical Sciences, Swami Ram Nagar, Doiwala, Dehradun - 248 140, India.

E-mail: drlahanviv@yahoo.com

wake him up at 5 a.m. and he usually left the bed within 10 minutes, feeling fresh. He was good in studies till the age of 7 years and had many friends.

After 6 months of shifting to residential school, his father received complaints from the school regarding deterioration in studies and sleeping in the class. His teachers had also noticed that he was gaining weight and losing interest in sports activities. Then, his father took him back home before 1 year.

After bringing him home, since the past 1 year, his parents noticed a gross change in his appetite with an increase in frequency and amount of food. A delay in meeting his demand of food used to result in irritability shown by the child. He continued to put on weight at home. His parents also noticed an increase in the time spent in sleep. For the past 1 year, he started feeling sleepy by 9 p.m. During the night, he would snore and spent most of his sleep in prone position. On some occasions, his father had found saliva on the pillow in the morning. His parents were not able to wake him up till 7 a.m., that too, with a lot of difficulty. After waking up, he took nearly an hour to become active. In addition, he started taking 3–4 hour nap after lunch each day. If he was not allowed to take nap any day, he would fall asleep by 5 p.m to wake up at 7 a.m. in the morning. Any force to avoid nap resulted in irritability. He lost interest in sports activities since then.

The child's father also noticed frequent memory lapses resulting in misplacing his belongings. There was no history to suggest childhood depression, frequent rhinitis, tonsillitis, attention deficit hyperactivity disorder, restless leg syndrome, cataplexy, hypnogogic or hypnopompic hallucinations, sleep paralysis or any other parasomnia. There was no evidence of any neurological disorder, epilepsy, head trauma or substance abuse. His birth history and developmental history were noncontributory. Family history was also nonremarkable.

His craniofacial examination showed presence of central obesity, dental overjet, Mallampatti grade IV upper airway, submental fat and high arched palate. Epworth Sleepiness Scale score was 24. His weight was 56 kg and height was 132 cm, leading to body mass index (BMI) of 32.18. His neck circumference was 34 cm.

Mental status examination showed normal psychomotor activity. Child was irritable on occasions following trivial issues and was reluctant to comply with examination procedure. He failed to comply when higher mental functions were being tested.

His lateral neck skiagram showed adenoid hypertrophy [Figure 1] and magnetic resonance imaging (MRI)

brain was noncontributory. IQ assessment was also ordered. Since the child did not comply on the first day, the tests were performed after 2 days. At the time of administering tests, child was in better mood and performed all the tests. On developmental screening test, he attained a score of 90; on Vineland Social Maturity scale, his score was 70; on Malin's Intelligence Scale (Indian Adaptation) – Wechsler Intelligence Scale for Children (WICS) (performance test and verbal test), he attained a score of 70 each. Thus, the comprehensive score was 75.

Considering the clinical picture, diagnosis of obstructive sleep apnea was made and narcolepsy without cataplexy and KLS were kept as differential diagnoses. Consequently, a level-I video polysomnography followed by multiple sleep latency test (MSLT) was ordered.

Level I polysomnography was done with a total recording time of 452 minutes. Total sleep time was 362 minutes. Objectively, the boy had sleep efficiency of 90% ($100 \times \text{Total Sleep Time (TST)} / \text{Sleep Period Time (SPT)}$); sleep onset latency of 44 minutes; Rapid Eye Movement (REM) latency of 187 minutes and Wake after Sleep Onset (WASO) of 47 minutes. During the study night, N1 was 12%; N2 was 48%; N3 was 18% and REM was 22%. Hypnogram suggested frequent arousals [Figure 2]. He spent most of time in right lateral position [Figure 3] and Respiratory Disturbance Index (RDI) was 2 (REM=4; Non Rapid Eye Movement (NREM)=1). Respiratory events were position dependent. Average saturation during REM was 97% and during NREM was 98%. Oxygen saturation dropped to 87% during REM and 94% during NREM. Snoring was also observed with an index of 8.9 without any effect of sleep stage.



Figure 1: Neck skiagram showing adenoid hypertrophy in the child (lateral view)

MSLT was done the following day. Four naps were recorded as per the standard protocol [Figure 4]. Results of this test are depicted in Table 1.

DISCUSSION

KLS is commonly seen in adolescent boys.^[1] This may present with prominent psychiatric features that may delay the diagnosis until a clear hypersomnia pattern is established.^[6] Amongst the psychiatric symptoms, bizarre behavior and mania like symptoms have been described.^[6,7]

Periodic hypersomnia with an increase in total sleep time is an essential feature in the diagnosis of KLS.^[1] Each

episode may last from a few days to years.^[1] During the episode of hypersomnolence, patients may be awakened but they become irritable when prevented from falling asleep.^[1] This was seen in the present case also. One important feature that we have found in the present case is compensatory advancing of time to bed that has never been reported previously, to the best of our knowledge.

KLS may be precipitated after flu-like illness, overseas journey, anxiety, alcohol consumption or sleep deprivation.^[2,8] However, most cases are idiopathic.^[4] In the present case, we could not find any factor that could lead to this type of clinical picture except the possibility of stress on the child owing to his shifting to a residential school. But the child clearly denied

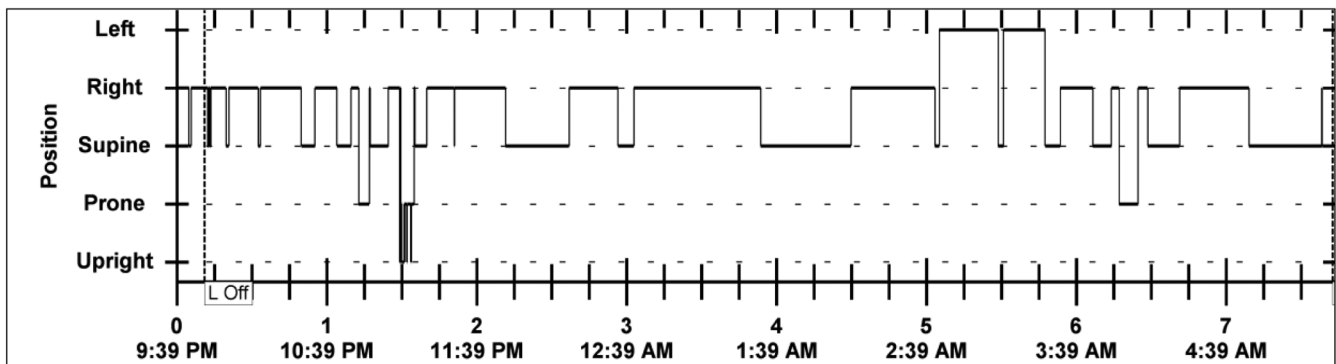


Figure 2: Hypnogram showing poor maintenance of sleep

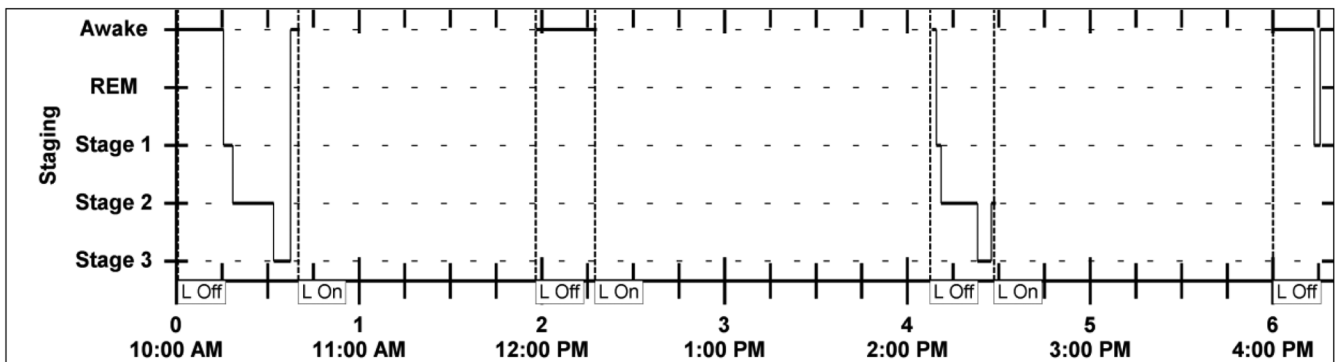


Figure 3: Hypnogram showing frequent shifting of body position during sleep

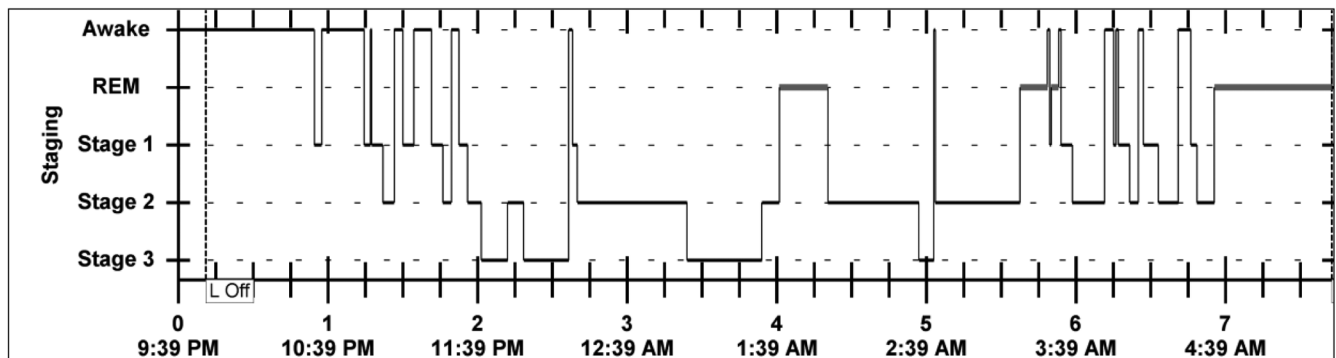


Figure 4: Hypnogram showing daytime naps

Table 1: Multiple sleep latency test report

	Nap 1	Nap 2	Nap 3	Nap 4	Mean values
Start time	10:00:48 a.m.	11:58:18 a.m	2:07:48 p.m	4:00:18 p.m	-
End time	10:40:18 a.m	12:17:48 p.m	2:28:48 p.m	None	-
Time in bed (minutes)	39.50	19.50	21.00	20.50	25.13
Total sleep time (minutes)	22.00	0.00	19.00	2.00	10.75
Sleep latency* (minutes)	15.00	None	2.00	13.50	12.63
REM latency (minutes)	None	None	None	None	Invalid

*Time taken to fall asleep; This report will display a default sleep latency of 20 minutes if the patient fails to sleep during a nap

any stress before the onset of symptoms. Moreover, the symptoms kept persisting even after the child was brought back home, for nearly 1 year. Other illnesses that can be precipitated during stress (e.g. depressive disorder) were ruled out on the basis of history and mental status examination. It must also be considered that depression may also present with megaphagia and hypersomnia.^[9]

Cognitive dysfunction is also common in KLS which may disappear with the termination of episode.^[2,7] In addition, loss of spontaneous speech, mutism, communication with monosyllabic or short sentences with restricted vocabulary have also been reported.^[1] Short-term memory dysfunction is usually present in these cases that may persist after recovery.^[10] We found poor verbal attention and execution with impaired short-term memory. Similarly, visual attention and executive abilities were also affected. IQ assessment revealed an IQ score of 75 which was in normal range for age.

Polysomnography in KLS suggests normal sleep cycle with normal or reduced N3 sleep.^[6,7] Sleep onset latency and REM latency may be reduced.^[2,6,7] Frequent arousals from N2 sleep with poor sleep efficiency is also known in addition to the frequent stage shifts.^[2,8] In KLS, sleep changes depend upon the phase of the illness – the slow wave sleep (SWS) is low in the first of the symptomatic period to return to normal during the asymptomatic period of the illness.^[8] An improvement in sleep efficiency, N1 and N3 stages may be expected at the end of the episode.^[2] In addition, it must be remembered that polysomnographic evidence of sleep architecture abnormalities is not found in all subjects, but only a small number of patients.^[2,6] The child in the present case showed normal distribution of sleep stages except for an increase in N1. However, like many previous reports, frequent stage shifting was noticed in this case. Possibly, the child was in second phase of illness and this could be one reason why we did not find classical polysomnographic abnormalities reported earlier.

Sleep onset REM on MSLT has been reported only in a small number of patients (21%).^[1] In the present

case, the child was still in the symptomatic stage, yet MSLT did not add to the diagnosis, probably because of the apprehension borne by the child. Even then, the third post-lunch nap at the usual nap time for the child showed reduced sleep latency. This is still a debatable issue and requires further research.

Sexual disinhibition is also seen in KLS cases; however, we could not find any evidence for the same in our case.^[6] One reason for this may be pre-pubertal age of the boy.

The differential diagnosis in the present case included obstructive sleep apnea that was ruled out based upon the overnight polysomnography. Hypothalamic structural abnormality was ruled out by normal MRI brain. However, we did not assess the functional activity of hypothalamus owing to contradictory literature.^[8] In addition to excessive daytime sleepiness, narcolepsy is characterized by cataplexy, hypnagogic or hypnopompic hallucinations, sleep paralysis with reduced REM latency and at least two SOREMPs on MSLT. In most patients with narcolepsy, brief naps and prolonged sleep periods give them a refreshing feeling.^[4] The absence of aforementioned features in our case ruled out narcolepsy. IH is another condition characterized by onset during adolescence, with increased daytime sleepiness, sleep drunkenness and polysomnographic features of shortened sleep latency, increased total sleep time and a normal sleep architecture.^[4] The sufferer is often difficult to wake up in the morning and has normal MSLT and without nighttime sleep fragmentation.^[5] The presence of hyperphagia, mood disturbances outside morning time and fragmented sleep during night in our case ruled out IH. This explains the withdrawn behavior and irritability present in our case, thereby ruling out Major Depressive Disorder.

KLS is a clinical diagnosis. Except for the presence of hyperphagia in our case, the overlap of symptoms with IH cannot be denied. IH has many commonalities with narcolepsy and atypical depression.^[3] This suggests that these disorders, viz. KLS, IH and narcolepsy, may lie in a continuum. Further research is required to get a clearer picture.

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