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# Kleine Levin Syndrome

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**ABSTRACT**

Kleine-levin syndrome is characterized as a periodic hypersomnia crisis as well as changes in appetite and variable behavioural symptoms. The exact aetiology is unknown and it is diagnosed based on clinical manifestations. This report describes the finding of a 15- year- old boy with KLS. He presented with hypersomnia, auditory hallucinations and persecutory delusions and all the episodes started with skin lesions over the leg. He had amnesia to his episodes. Kleine-levin syndrome should be considered in patients with periodic hypersomnia and skin lesions. The objective of this case report is that clinicians should have a high index of suspicion of Kleine Levin syndrome in case of periodic hypersomnia.

**Key words:** Psychosis–Adolescence–Skin Lesions–Hypersomnia

## 1 INTRODUCTION

**Kleine-Levin syndrome,** also known as **Sleeping Beauty Syndrome (**Saha A,2008**)**, is a rare disorder primarily seen in adolescent males, characterised by the need for excessive amounts of sleep (up to 20 hours a day) and excessive food intake, as well as behavioural changes such as an abnormally uninhibited sexual drive (Ramdurg S.2010). At the onset of an episode the patient becomes progressively drowsy and sleeps for most of the day and night (hypersomnolence), waking only to eat or go to the bathroom. When awake, the patient's whole demeanor is changed, often appearing "spacey" or childlike. When awake he experiences confusion, disorientation, complete lack of energy (lethargy), and lack of emotions (apathy). Most patients report that everything seems out of focus, and that they are hypersensitive to noise and light. In some cases, food cravings (compulsive hyperphagia) are exhibited. Instances of uninhibited hypersexuality during an episode have also been reported. Kleine-Levin Syndrome episodes are cyclical (Justo.LP et al., 2007). When present, KLS symptoms persist for days, weeks or even months, during which time all normal daily activities stop. Individuals are not able to attend school, work or care for themselves. Most are bedridden, tired and uncommunicative even when awake. Not everyone affected by KLS exhibits all of the symptoms described above. Affected individuals may go for a period of weeks, months or even years without experiencing any symptoms, and then they reappear with little warning. In between episodes those diagnosed with KLS appear to be in perfect health with no evidence of behavioral or physical dysfunction. However they function daily with the frightful reality that they could become sick again at any moment. KLS episodes may continue to reoccur for a decade or longer with devastating effects on the adolescent's life and family. KLS robs children and young adults of big pieces of their lives, one agonizing episode at a time.

The cause of Kleine-Levin Syndrome is not known. This disorder should be differentiated from encephalopathy, recurrent depression, or psychosis.

**Diagnostic Criteria: Recurrent Hypersomnia (ICSD)**

A. The patient has a complaint of excessive sleepiness.

B. The episodes of somnolence last for at least 18 hours a day.

C. The excessive sleepiness recurs at least once or twice a year, lasting a minimum of 3 days and up to 3 weeks.

D. The disorder occurs predominantly in males, with an age of onset typically in adolescence.

E. Associated features during the episodes include at least one of the following:

1. Voracious eating
2. Hyper sexuality
3. Disinhibited behaviour, such as irritability, aggression, disorientation, confusion, and hallucinations

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4. Absence of urinary incontinence and presence of verbal responses on strong stimulation

F. Polysomnographic monitoring during an episode demonstrates all of the following:

1. A high sleep efficiency
2. Reduced stage 3 and stage 4 sleep
3. Reduced sleep latency and REM latency
4. An MSLT with a mean sleep latency of less than 10 minutes.

G. The hypersomnia is not associated with other medical or mental disorders, such as epilepsy or depression.

H. The symptom is not associated with other sleep disorders, such as narcolepsy, sleep apnea syndromes, or periodic limb movement disorder.

Note: If the disorder is solely one of recurrent episodes of hypersomnia, state and code as recurrent hypersomnia monosymptomatic type. If the disorder is associated with voracious eating or hypersexuality, state and code as recurrent hypersomnia Kleine-Levin type.

**Minimal Criteria:** A plus B plus C plus G plus H.

## 2 CASE REPORT

15 years old boy studying in $8^{th}$ grade, belonging to rural background and of low socio economic status presented to child and adolescent psychiatric OPD of NIMHANS. He was temperamentally easy child with no significant family history and past history of any psychiatric or medical illness. His developmental milestones were normal. He presented with symptoms of 2 years duration, episodic in nature and acute in onset, and each episode lasted for about 7-10 days duration. Each episode was characterised by skin lesions with severe itching all over the legs. Following which he was noted to have excessive sleep, which would last for about 16-18 hours per day. He had persecutory delusions and would refuse to eat and report that poison has been mixed in his food. It was associated with 2nd person and 3rd person auditory hallucinations. These symptoms would last for about 7-10 days and would spontaneously remit. These episodes used to occur once in 2-3 months. Initial 5-6 episodes were not treated. Later he was treated with multiple medications like risperidone, paliperidone and atomoxetine with minimal improvement. In spite of treating with these drugs symptoms used to recur once in 2-3 months with similar symptomatology. Inter-episodic period patient used to reach premorbid levels. In these two years the boy also had significant decline in academic performance when compared to his premorbid levels. It was not associated with any comorbid medical or psychiatric illness. On mental status examination, child was reluctant to talk with decreased speech output, came very slowly for the interview, dull affect, also reported of 2nd and 3rd person auditory hallucinations with persecutory delusions. Detailed medical evaluation including neurological examination was done which revealed no significant findings. Various investigations were done. It included routine hemogram and biochemistry which was within normal limits. Even thyroid function tests were normal. CT brain and EEG also showed normal study. All the medications which the patient was previously taking were stopped and lithium was started. It was started with a dose of 300mg and gradually increased to 750mg/day. Serum lithium was in therapeutic range. The patient started to show favourable response to therapy within 2 days of starting lithium. Patient followed up in psychiatric OPD for 2 years with no relapse of episodes. Patients academic performance has also improved comparitively.

## 3 DISCUSSION

To summarise the symptoms in this boy, he had hypersomnia for about 18 hours, excessive sleepiness with verbal responses on strong stimulation, which used to recur once in 3months, and associated features are 2nd and 3rd person auditory hallucinations and persecutory delusions. It was not associated with any medical or psychiatric disorders. This symptom profile clearly fits into the diagnostic criterion of Kleine Levin syndrome (ICSD). Child also had recurrent skin lesions associated with itching which occurs prior to each episode. Some case reports have reported of temporal association of onset of kleine Levin syndrome and application of isotretinoin ointment. (Smedje H et al.,2010). These episodes occur once in 3 months with acute onset and remit spontaneously within a weeks time and the child reaches pre morbid level with normal functioning. This symptom profile is commonly not seen in recurrent depressive disorder or episodic psychosis. Child also didn't improve with antipsychotics. After the diagnosis of Kleine Levin syndrome, patient was started on lithium, with which he showed significant improvement and the child is symptom free till date. This diagnosis was delayed due to failure to understand the pathology, of the disorder thereby leading patients to wander.

It is tempting to hypothesize that periodic hypersomnia may be harbingers of KLS in some patients and clinicians should have high index of suspicion to ask for further evaluation of KLS.

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