KLEINE-LEVIN SYNDROME – A CASE REPORT

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ABSTRACT:

A rare neuropsychiatric disorder presenting as episodes of hyper-somnolence, behavioral and cognitive disturbance, hyperphagia and hyper-sexuality. Until date, no definite underlying cause has been established nor have there been any definitive management guidelines. This article aims to focus on this disorder by means of a case report. A young adolescent boy presented to our outpatient department with an abrupt onset of excessive sleep for about 18 to 20 hours per day lasting 1 to 2 weeks associated with confusion, hyperphagia and hypersexuality. Family members revealed that the patient had three similar episodes in the past but the patient himself denies remembering these
episodes. Inter-episodal periods showed complete recovery. No positive family history of similar illness. He denied having any hallucinations or delusions, or history of any substance abuse. Neuroimaging was normal. He was diagnosed as Non-organic hypersomnia-Kleine Levin Syndrome. He was started on psychostimulants for excessive sleepiness and on lithium therapy to reduce the episode frequency. Family members were psychoeducated about the disorder. Patient maintained well on the same treatment and is asymptomatic for past one year. A rare disorder often misdiagnosed or unrecognized and should be considered in any adolescent presenting with other symptoms. More long term studies are needed for better understanding of this rare disorder.

Keywords: Hypersomnia, Kleine Levin syndrome, excessive sleep


**INTRODUCTION:**

Kleine Levin Syndrome, a rare sleep disorder which is characterized by periods of excessive sleep, sexual disinhibition, voracious eating (megaphagia), and mental disturbance\(^1\). The duration of attacks ranges from several days to several weeks. Young adolescent boys are usually affected. Mental abnormalities may precede each episode and may persist for weeks thereafter. The psychiatric symptoms included delusions and hallucinations, unreality feelings, Irritability and mental confusion. Klein-Levin syndrome is rare, affecting an estimated population of 1-5 per million individuals\(^2\).

The diagnosis of Klein Levin Syndrome must have the following criteria:

A. At least 2 recurrent episodes of excessive sleepiness and sleep duration, each persisting for 2 days to 5 weeks.

B. Episodes recur usually more than once a year and at least once every 18 months.

C. The patient must demonstrate at least one of the following during episodes:
   1. Cognitive dysfunction
   2. Altered perception
   3. Eating disorder (anorexia or hyperphagia)
   4. Disinhibited behavior

D. The patient has normal alertness, cognitive function, behavior, and mood between episodes

E. These symptoms are not better explained by another sleep disorder, other psychiatric disorder, neurologic, or use of other drugs.
CASE REPORT:

A 17 year old adolescent boy was brought to our Psychiatric outpatient department by his parents. According to his parents, ‘patient was feeling drawn to his bed and was unwilling to wake up in the morning.’ Even though patient was arousable, patient had irritability and aggressive behavior whenever he was prevented from his sleep and slept for about 16-18 hours/day. When patient was awake, he would be confused, disoriented lasting for few minutes. Patient had increased appetite, asking for flavoured rice and chips. Patient reported increased frequency of masturbation and had disinhibited behavior towards women. The total duration of illness was 10-14 days with similar 3 episodes as mentioned above in the past. Duration of each episode was for about 7-14days with complete remission in between the episodes. There was no past history of psychiatric illness or neurological disorder or positive family history. His complete physical examination inclusive of general, systemic and neurological examination was normal. Biochemical investigations were within normal range. His cardiac status measured by Electrocardiogram, his electroencephalogram, and Magnetic Resonance Imaging (MRI) brain was found to be normal. Polysomnography was tried for which patient was not co-operative. As per International Classification of Sleep Disorders, the diagnosis of Kleine Levin Syndrome was made ruling out other possible causes of hypersomnia. Patient was started on Tab. Lithium 400mg BD and Tab. Modafinil 100mg BD for day time sleepiness. The patient showed improvement in his symptoms and frequency of hypersomnolence episodes had reduced. Tab. Modafinil was stopped when his hypersomnia was reduced and patient is currently on Tab. Lithium 400mg BD and is asymptomatic for 1 year.

DISCUSSION:

In 1942, Critchley first wrote about this disorder, which had the characteristic hallmark of voracious appetite and periodic somnolence. He assigned it to the nomenclature of the Kleine Levine Syndrome in 1962. Kleine Levin Syndrome was prevalent in Europe and Asia, more common among males with a male female ratio of 3:12 . There are about 48 cases of KLS reported from India. The usual onset with febrile illness, and inflammatory lesions which were detected in thalamus and hypothalamus by Carpenter et al, suggested a possible viral etiology. In this case, patient’s past three episodes were preceded by a brief febrile illness, and current episode by RTA. Educating patients parents is considered main stay of treatment. The patient should be advised to take rest within a comfortable environment. Lithium is considered to be preferred drug for treatment as well as prophylaxis and to reduce the frequency of future episodes. Modafinil can be considered for excessive daytime sleepiness. Antidepressant, anxiolytic mood stabilizer or antipsychotics can be considered according to symptoms. The patient’s serum lithium levels were monitored regularly. The patient showed improvement on regular medications and is asymptomatic for 1 year.

CONCLUSION:

Kleine Levin Syndrome, a neurological disorder which is inclusive of a lot of psychiatric symptoms. Such cases may be presented to a physician, or to a neurologist, or to a psychiatrist depending on patient’s insight about the
illness, feasibility and symptomatology. Hence clinicians must be aware with such atypical presentation and be suspicious on such cases. Any case which has a presentation of hyper somnolence which is episodic in nature should have a detailed evaluation for timely diagnosis and appropriate management must be given.

REFERENCES:


