Case Report

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Kleine–Levin Syndrome: Sleeping Beauty: A Rare Case Report on Sleep Disorder

Ajaykumar Dhage, Rahul Mandaknalli

Assistant Professor, Department of Psychiatry, M.R. Medical College, Gulbarga, Karnataka, India

Abstract

Kleine–Levin syndrome (KLS) also called sleeping beauty syndrome, is a rare sleep disorder with onset in early adolescence. Common symptoms are episodes of hypersomnolence, behavioral and cognitive disturbances (including specific feelings of derealization), hyperphagia and hypersexuality. The cause is not known and neither there are any definitive management guidelines. It remains a diagnosis of exclusion after ruling out other psychiatric and neurological disorder. Disturbed behavior may dominate the picture, suggesting that the essential problem is a personality disorder or even schizophrenia, and it is important for a psychiatrist to be well informed about the condition to avoid the erroneous diagnosis. KLS is an illness with devastating course, which disturbs socio-occupational activities. An early diagnosis and effective management can help patient escape from the morbidity caused by this disorder. Modafinil and lithium were found to be effective, in this case. The priority of this report is to add the existing clinical knowledge of psychiatrists, neurologists and physicians. In the near future, research is needed on neurobiology, genetic etiology and management of this disorder.

Key words: Hyperphagia, Hypersomnolence, Kleine–Levin syndrome, Lithium, Modafinil

INTRODUCTION

Kleine–Levin syndrome (KLS) is a rare disorder with periodic hypersomnia, eating disturbance, cognitive and behavioral disturbances. The disease was named, "KLS" by Critchley in 1962 after Willi Kleine and Max Levin who studied multiple cases of hyper somnolence and emphasized the association of periodic somnolence with megaphagia to morbid hunger from 1925 to 1936.[1]

International Classification of Sleep Disorders-3 criteria (2013) states following five key points for diagnosis:

A. At least two recurrent episodes of excessive sleepiness of 2 days to several weeks
B. Episodes recur at least 1 per 18 months
C. Normal alertness, cognitive function, behavior and mood between episodes
D. At least one of these during an episode:
   - Cognitive dysfunction
   - Altered perception, derealization
   - Eating disorder (anorexia or hyperphagia)
   - Disinhibited behavior (such as hypersexuality).
E. Symptoms not better explained by other disorders.[2]

This disorder because of its sporadic presentation has unknown prevalence. A systematic review of 186 cases showed incidence usually in adolescence with a course lasting for 8 years or more.[3]

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CASE REPORT

Mr. X 21-year-old male, Hindu nuclear family, studied up to 10th std., belonging to low socio economic group presented with complaints of excessive sleep episodes of abrupt onset lasting for 10-12 days 2-3 times a year for the past 3 years. He was found to be sleeping more than the usual with average sleeping time of 18-19 h a day. It was difficult to arouse him while he was sleeping, on waking up he was generally irritable extending at times to severe aggression, confused, disinhibited behavior, decreased speech output, marked decline in self-care. It was also associated with hyperphagia. He would eat unusually

Corresponding Author: Dr. Ajaykumar Dhage, Plot no. - 162, Chinde Layout, Near Laxmi Temple, Swastik Nagar, Sedam Road, Gulbarga - 585 105, Karnataka, India. Phone: +91-9901082778/9538404111. E-mail: jayd11@gmail.com

more quantities of food. Inter-episodic periods showed complete recovery. There was no positive past or family history of neurological or psychiatric disorder. History of meningitis, head injury, and substance abuse were ruled out. All biochemical, endocrine, radiological parameters and electroencephalography were found within normal range. A clinical diagnosis of KLS was made, and patient was put on modafinil 100 mg oral dose for 30 days and was switched to lithium 450 mg and he is maintaining well with no recurrence.

DISCUSSION

Patient fulfilled the criteria for KLS, but due to the rarity of the disorder he was previously diagnosed as psychosis not otherwise specified. This disorder is commonly considered as a neuro-psychiatric disorder and psychotic symptom can be part of this disorder.[4] The disorder is rare in females.[5] The disease mimics and shares psychiatric conditions in many ways, and hence it is easily misdiagnosed as personality disorder or major mental illness. Furthermore, therefore, it is important to have a high index of suspicion in any case presenting with complaints of episodic hypersomnolence and after ruling out differentials like Kluver–Bucy syndrome, atypical depression, substance abuse and other differentials for hypersolomnelence[2] and finally diagnosis of KLS should be made. Lithium, valproate, carbamazepine, amphetamine, L-dopa, modafinil, armodafinil have been tried for symptomatic treatment and for prevention of relapse with variable results as there are no definitive guidelines for treatment.[3,6] KLS is a neuro-psychiatric disorder therefore a case can present to neurologist or psychiatrist depending upon patients underlying symptoms, so it is important for clinicians to have high index of suspicion mainly for atypical presentations to avoid delay in diagnosis or making erroneous diagnosis. Though most cases have an abrupt onset but precipitating events like neurological infection should not stop clinician to suspect KLS.[7,8]

In the above case report after a short course of modafinil, started on lithium and patient is maintaining well with no recurrence. Thus, modafinil and lithium was found to be effective in this case.[9]

CONCLUSION

This case report aim to highlight that KLS though considered a rare disorder but not uncommon and lack of enough available research data is likely to be responsible for missed or erroneous diagnosis; thus we require more systematic studies regarding etiologies and treatment.

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