CASE REPORT

Kleine - Levine Syndrome (A Rare Case Report)

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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 hyper somnolence, 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 disorders. Our case,a 15 year old boy with duration of illness of 3 years, episodic presented with hypersomnia,increased appetite, irritability, hyper sexuality and behavior abnormality mainly fearfulness and suspiciousness. Patient responded well to lithium therapy. The priority of this report is to add the existing clinical knowledge of psychiatrists, neurologists and physicians.

Keywords : Hyper phagia, Kleine–Levin syndrome, hyper somnolence, Lithium

INTRODUCTION

Kleine–Levin syndrome (KLS) is a rare disorder with periodic hypersomnia, 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 morbid hunger from 1925 to 1936.[1]

International Classification of Sleep Disorders-3ncriteria (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

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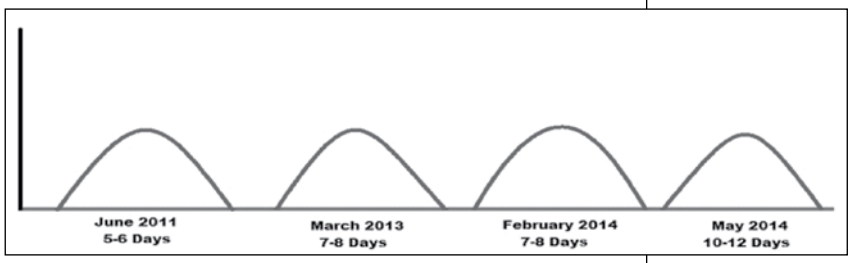
186 cases showed incidence usually in adolescence with a course lasting for 8 years or more.[3]

CASE REPORT

A 15 years old male student reported in May 2014 in emergency department with complaints of high grade fever for one day followed by excessive sleepiness almost 16 to 18 hours per day and difficult to arouse from sleep. He was admitted in Department of medicine. His routine biochemical and specific



investigations (CT & MRI brain, CSF analysis and EEG) were within normal limits. Patient became afebrile on next day after parenteral antibiotics and antipyretics. But the patient was still excessively sleepy and difficult to arouse. He was attended by psychiatrist.On examination, he was sleepy, difficult to arouse and was not answering to any question. He was confused,irritable, and showed marked decline in self-care. Probable diagnosis of catatonia was made and patient was shifted to psychiatry ward for further management. Inj. Lorazepam (4mg) was given stat but his condition neither improved nor deteriorated. On observation it was found that he would eat unusually more quantities of food. Patient denied for any delusion or hallucination. In next 5 days he recovered completely with normal sleep, appetite and behavior. However, he was completely amnestic about the event. There were three episodes in past three years having excessive sleepiness,hyper-sexuality and irritability with self remission in 7-8 days. Inter episodic periods showed complete recovery for that he never consulted to any psychiatrist.

There was no positive past or family history of anypsychiatric illness or neurological disorder. Pre morbid traits were well adjusted. Finally he was diagnosed as a case of Kleine – Levine syndrome and he was put on lithium 600mg in divided doses. He is maintaining well with no recurrence.

DISCUSSION

Kleine-Levin syndrome (KLS) is a peculiar disorder that primarily affects males. This disorder is conventionally considered as a neuro-psychiatric disorder and has been classified under the category of sleep disorder- recurrent hypersomnia or disorder of excessive somnolecence.[4]

The International Classification (ICD-10) has included Kleine-Levin as a disorder of organic origin and has therefore, classified in chapter VI in Diseases of Nervous System (G47.8).[5]

The diagnosis of Kleine-Levin syndrome is based on clinical features alone, as there are no specific laboratory tests that can help in establishing the diagnosis of Kleine-Levin syndrome.[6] Impairment of cognitive functions and a wide variety of behavioral abnormalities are commonly associated as elicited in the present case report.[7]

Kleine-Levin syndrome generally has sudden onset though sometimes a flu-like illness or a period of physical stress including the head trauma has ante-dated the first attack.[8]Febrile illness prior to first episode as seen in our case is not a common presentation.[9, 10]

The case fulfilled the criteria for Kleine–Levine syndrome because of the episodic cluster of behaviors mainly hypersomnia and related psychiatric symptomatology, but due to rarity of the disorder he remained undiagnosed for three years. As disease mimics and shares psychiatric conditions in many ways, so it is important to have a high index of suspicion in any case presenting with complaints of episodic hyper somnolence and after ruling out differentials like Kluver–Buccy syndrome, atypical depression, substance abuse and other differentials

for hyper solomnelence[2], diagnosis of KLS should be made. Lithium, sodium valproate, carbamazepine, amphetamine, Ldopa, modafinil, armodafinil have been tried for symptomatic treatment and for prevention of relapse with variable results in the absence of definitive treatment.[3,13] In the present case the patient was put on Tablet Lithium for the first time and his condition is stable since the treatment has started.[14] Hence, the question arises whether it is advisable to give maintenance treatment to Case or wait for another episode to start a maintenance drug.[11,12]

Though most cases develop spontaneously but presence of precipitating events like neurological infection should not stop clinician to suspect KLS.[8,15]

CONCLUSION

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

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