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CASE

"Sleeping beauty syndrome": two cases of Kleine-Levin syndrome

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Abstract

Two cases of Kleine-Levin syndrome (KLS), manifested in fifth and first decade of life respectively, are presented. The first case presented with episodic pattern of hyper somnolence and mood changes whereas the second case presented with hyper somnolence, behavioural and cognitive symptoms. Both of them had responded well with adequate trial of lithium. In both the cases, onset is in the extremes of their ages, which is rarely found.

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Introduction

'Sleeping beauty syndrome', also known as Kleine-Levin syndrome (KLS), is a rare disorder of uncertain aetiology. Presentations range from the monosymptomatic, to the classical presentations consisting of hypersomnolence, hyperphagia, hypersexuality or behavioural disturbances. Typical syndrome was first described by Kleine in 1925. The second edition of the International Classification of Sleep Disorders (ICSD-2) has described KLS as 'Recurrent episodes of excessive daytime sleepiness lasting for two days to four weeks, episodes recur at least once per year, in addition to hypersomnia, there should be cognitive disturbance or behavioural symptoms or hyperphagia or hypersexuality'.[1] Critchley and Hoffman[2] emphasised that it is not excessive hunger which is characteristic of KLS but sight of food induces an impulse to eat voraciously and so termed this behaviour as megaphagia. According to the systematic review of 186 cases by Arnulf et al.,[3] 68% were males and common symptoms were hypersomnia (100%), cognitive changes (96%, including a specific feeling of derealisation), eating disturbances (80%), hypersexuality (43%), compulsions (29%), and depressed mood (48%). Of the 186, only ten cases were from India.[3] We are reporting two cases presented to the Lokopriya Gopinath Bordoloi Regional Institute of Mental Health (LGBRIMH), Tezpur, Assam, India.

Case 1

A 46-year-old married businessman with well-adjusted pre-morbid personality presented with acute onset episode of excessive sleepiness of one week duration. He developed this episode after his daughter, who was close to him, had opted for studying away from home. He predominantly complained of sleepiness whole day and night, feeling irri-

table and fatigued, and did not talk to others during the episode. There were two similar episodes six months and one year back, which lasted for 15 and 20 days respectively, without any precipitating cause. He continued his business as usual in between attacks. No history of substance or any medication use was reported. There was no history of any medical illness or any other significant past history. Mental status findings on the day of presentation were not contributory. Provisional diagnosis of KLS was made after exclusion of other similar disorders and was started on lithium 900 mg in divided doses, patient was on regular follow up on a monthly basis. For the last one year there has been no recurrence.

Case 2

A 11-year-old girl studying in class VI presented with episodic attacks of sleepiness of abrupt onset which lasted for an average of 15 days. The child was completely alright in inter-episodic interval. The episode presented with excessive sleepiness, headache, singing songs inappropriately, decreased interest in daily activities, decreased self-care and hygiene, avoiding company, and remaining irritable. She was forgetful of her behaviour during attacks. For the last three years she had ten to 12 attacks, with average of two to three months of asymptomatic inter-episode duration. Positive findings during mental status examination revealed depersonalisation. The child reported that it was like a dream. She complained of headache and was irritable, also reported that she felt bad sometimes. Initially provisional diagnosis of childhood psychosis was made. Electroencephalography (EEG) and computed tomography (CT) scan of the brain were normal. She was also assessed in medical college, magnetic resonance imaging (MRI) brain was done with other necessary blood work-up, but without discovering any specific cause to which symptoms could be attributed. Then the child was brought back to our institute, she was started on oral olanzapine 2.5 mg and clonazepam 0.25 mg BD. On subsequent follow up, olanzapine was increased to 7.5 mg and then clonazepam was gradually tapered and stopped. After five months of asymptomatic period the child again presented with similar attack, even though she was on regular medication. Then, diagnosis as a case of KLS was considered after exclusion of other possible differentials, and started on lithium 450 mg per day. In follow up visits the child had less frequent episodes, but was not fully asymptomatic. Then the dose of lithium was increased to 600 mg, and thereafter she has been maintaining well without any recurrence for the last eight months.

Discussion

We have reported two cases of KLS from northeastern part of India, with important implication in differential diagnosis and management.

Critchley[4] described the predominance of KLS in adolescent males. The median age of onset was 15 years (range four to 82 years, 81% during the second decade).[4] In our report, case 1, presented with episodic pattern of hyper somnolence and mood changes in fifth decade, which is very rare. Unusual stress has been seen to precipitate KLS in 20% of patients.[3] In our case, the third episode was precipitated by stress but that was not of much severity.

Our second case, described in a, 11 years old female child, which is typical with behavioural and mood dysregulation, and depersonalisation. In 81% of all the studied cases, KLS onset occurred during the second decade.[3] The age of onset in this patient was in the first decade which is much earlier. During episodes she remained irritable and despondent most of the times, and had compulsions to sing songs which are similar to the finding by Arnulf et al.[3] Arnulf et al.,[3] in his systemic review found to have irritability in 92% of patients and depression in 48% of patients, and compulsion to sing songs in 29% of patients.[3]

Both of our cases initially masqueraded as other conditions like psychosis, mood disorder, seizure disorder. Both cases were started on lithium and recovery was very satisfactory. The complexity may be due to polygenic influence or associated features that are non specific or less in consideration by clinicians as a differential. Although the syndrome is clearly defined but no clear underlying biological correlates have been found till now. Substantial proportion of patients remits spontaneously, again creating doubts of effective treatments used for cases in the past. Orlosky[5] reviewed 33 well-documented cases of KLS and was of the opinion that KLS may be a mirror-image of anorexia rervosa. Others have hinted to either mood disorder or psychosis.[6] Currently it is classified into hypersomnia of central origin, according to ICSD-2.[1] Future change in the nosological status is more likely as biological correlates may be uncov-

ered and as our understanding is improving. Overall, five per cent of cases have been reported within family members, suggesting some degree of increased genetic susceptibility; however, no genetic abnormality has been yet identified.[3] The DQB1-0201 haplotype was found to be twice as frequent in a controlled study of 30 European patients.[7] Abnormalities of hypothalamic pituitary axis (HPA) axis, neurotransmitter abnormalities, inflammatory and infectious aetiologies have also been proposed but are inconclusive.[3] Several treatments have been used, including stimulant, anti-epileptic, anti-depressant and anti-psychotic drugs, with some benefit reported, but because the condition is not so common, long-term follow up is difficult.[8] Despite multitude of agents that are tried, amphetamine had a response rate of 71% which mainly targeted drowsiness during episodes without much effect on cognition dysfunction, and lithium had a response rate of 41% in preventing relapse.[3] However, no evidence indicates that pharmacological treatment for KLS is effective and safe.[8]

Conclusions

KLS can have varied presentation, sometimes mimic psychosis or mood disorders. Correct identification and better understanding of the condition can improve lives of many patients who are still undiagnosed or wrongly diagnosed. The eventual, spontaneous resolution of the symptoms and the mechanisms determining its periodicity is a secret that needs to be unraveled and should prompt future investigation and development of therapeutics.

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