KLEINE-LEVIN SYNDROME: A REPORT OF TWO CASES

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ABSTRACT

Two cases of Kleine-Levin Syndrome with illness duration of three and four months respectively are presented. Both cases in their adolescence presented with typical features of the syndrome—onset after high grade fever, episodic course and spontaneous remission of each episode and normalcy in between the episodes. Characteristic features of each episode in both cases were hypersomnia, eating excessively, disinhibited behaviour, affective features like irritability, social withdrawal and lack of personal care and cognitive disturbance. The second case had sexual disinhibition which is another important feature seen in Kleine-Levin Syndrome. Both patients responded well to lithium therapy.

Keywords: Kleine-Levin syndrome

The Kleine-Levin syndrome is a rare episodic disorder of uncertain etiology, typically with the onset in adolescence and with a tendency to spontaneous remission over years (Critchely, 1962; Lishman, 1998). First described by Kleine in 1925, the syndrome is characterised by episodes of hypersomnia with marked increase in total sleeping time and hyperphagia with impulsive or compulsive eating, greedy manner of eating and at times a preference for sweets (Orlosky, 1982; Lishman, 1998). Critchely in 1962 included disturbance of mood as an important feature of Kleine-Levin syndrome. Most frequent psychological symptoms are irritability, excitation, uneasiness and social and sexual dis-inhibition (Gillberg, 1987; Sagar et al., 1990). These patients also present with withdrawal from social contact with acute onset of stupor, irritability on awakening and hyperphagia. Impairment of cognitive functions is commonly associated.

From India very few cases of Kleine-Levin syndrome have been reported (Prabhakaran et al., 1970; Shukla et al., 1982; Sagar et al., 1990; Malhotra et al., 1997). Here we present two cases of Kleine-Levin syndrome which were referred to the Child and Adolescent Psychiatry Clinic of G.B. Pant Hospital, New Delhi for management during last two years.

Case 1: A 11 years old boy studying in 5th standard, hailing from a Hindu nuclear family, with uneventful birth and developmental history, without past and family history of neurological and psychiatric illness presented with an episodic illness of 3 months duration with each episode lasting for 7-8 days at the interval of 1-2 weeks. Onset of the first episode was associated with high grade fever lasting for two days prior to the symptom manifestation. During each episode, the patient complained of mild continuous headache, constriction in nature, not associated with nausea, vomiting, photophobia or weakness. He was found to be sleeping more than the usual with average sleeping time of 17-18 hours a day. It was difficult to arouse him while he was sleeping. On waking up he was generally irritable, talked less, made excuses, and cried excessively without provocation. At times he looked confused, spitted excessively, chewed pieces of paper and showed no concern for his personal care. During these episodes his eating pattern also changed. He would eat unusually more quantities of food and insist on having the food items of his choice and eating food
KLEINE-LEVIN SYNDROME: A REPORT OF TWO CASES

from his sister's plate after finishing his own. He also did not go to his school during these episodes. The patient had complete spontaneous recovery after 7-8 days each time with partial amnesia. These episodes occurred at the interval of 1-2 weeks and this period between two episodes was completely normal with his usual interest in his studies and attending to school. He was subjected to routine laboratory investigations for blood count, thyroid function tests, electroencephalogram (EEG), and CT scan head. All these investigations were found to be within normal limits. He was diagnosed to be suffering from Kleine-Levin syndrome. He responded very well to lithium therapy and during 6 months of follow up there was no re-appearance of these episodes.

Case 2: A 13 years old boy studying in 8th standard from a Hindu joint family, with normal birth and developmental milestones without past and family history of neurological and psychiatric illness presented in the clinic with an episodic illness of 4 months duration, with each episode lasting for 8-10 days. First episode was preceded by high grade fever lasting for two days. During each episode, the patient had increased sleep with duration of 16-18 hours per day. On waking up, he was irritable appeared dull and confused, cried excessively without any apparent reason and expressed death wish. At times he was seen lying prone in the bed and rubbing his genitalia against the bed. He frequently got angry and lost temper with his parents when they prohibited him from doing so. He also complained of throbbing frontal headache, repeatedly demanded sweets, chocolates, soft drinks and ate other family members' share of food unlike when he was well. He also remained absent from the school during these episodes. He had complete and spontaneous recovery from each episode with complete amnesia. These episodes occurred at a variable interval of 2-4 weeks and during these intervals he was completely normal and attended his school regularly. Routine laboratory investigations including blood count, thyroid function test, electroencephalogram (EEG) and CT head were found to be normal. He was diagnosed to be a case of Kleine-Levin syndrome and was treated with lithium. He responded very well to the therapy and remained under regular follow-up. At 6 months of follow-up he had no recurrence of these episodes.

DISCUSSION

Kleine-Levin syndrome is a peculiar disorder that primarily affects males but up to a quarter cases are now reported in females (Duffy et al., 1968). 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 somnolence (Moore et al., 2000). The International Classification (ICD-10) has not included Kleine-Levin syndrome under the heading of behavioural syndromes associated with physiological disturbances and physical factors (F50-F59) or non-organic hypersomnia (WHO, 1992). It has been considered as a disorder of organic origin and has therefore, classified in chapter VI in Diseases of Nervous System (G47.8). 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 (Gallink, 1954). Impairment of cognitive functions and a wide variety of behavioural abnormalities are commonly associated as elicited in both the cases reported (Pike, 1994). The two cases reported here are the considered examples of the Kleine-Levin syndrome because of the episodic cluster of behaviours mainly hypersomnia and related psychiatric symptomatology.

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 (Will et al., 1988). Fabriile illness prior to first episode as seen in both of our cases is not an uncommon presentation (Critchley, 1962; Lishman, 1998).

The first patient's symptoms aggregated around a general disinhibition hyperphagia and hypersomnia, which is episodic in nature. His young age may be particularly responsible for the incomplete presentation, especially with regard
to hypersexuality which some authors argue for (Billard 1988). The second patient is a typical case of Kleine-Levin syndrome.

There are very few published reports from India on Kleine-Levin syndrome. Malhotra et al. (1997) have documented it with evidence for hypothalamic-pituitary axis dysfunction while Sagar et al. (1990) have studied inter-episodic morbidity and found significant maladjustment. Prabhakaran et al. (1970) and Shukla et al. (1982) have reported cases as we have done. Both cases we reported have classical symptom manifestation of Kleine-Levin syndrome and have responded to lithium therapy which is consistent with earlier reports on treatment of this disorder (Ogura et al., 1976).

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