Kleine-Levin Syndrome

A 4 year-old boy presented with excessive sleepiness for 3 days with a history of having sustained a head injury a week ago. The CT scan was normal. On examination, the child was drowsy and irritable when aroused from sleep. Vitals signs were stable. No tonsillar or adenoid hypertrophy was present on examination. There was no focal neurological or signs of meningeal irritation abnormality. Blood counts, renal and liver functions, cerebrospinal fluid analysis, EEG and Magnetic resonance imaging did not give any clues. After hospitalization he was noticed to be asleep most of the time in excess of 18 hours in a day. He was hardly arousable and would abuse if disturbed while sleeping. The only stimulus which would arouse the boy from slumber was the sight or smell of food. He was noticed to be excessively hungry and the parents noticed that he had an abnormal voracious appetite and craving for food. A 24-hour dietary recall revealed an astonishing intake of 3450 calories and 50 grams of proteins. He was also noticed to have abnormal sexual behavior for his age in the form of attempts at masturbation. Based on this unusual constellation of hypersomnia and hyperphagia a diagnosis of Kleine-Levin syndrome was made. He was subjected to a 24-hour polysomnography. The sleep architecture consisted of predominantly slow-wave sleep pattern throughout the recording with intermittent short bursts of REM. He was awake for only 3 hours during this period and when awake had excessive intake of food. SPECT scan was normal and no perfusion abnormality was detected. He was treated with fluoxetine and he improved gradually over a period of 10 weeks. He is now asymptomatic and is on regular follow-up.

Kleine-Levin Syndrome (KLS) belongs to the category of recurrent hypersomnia, and not better explained by a sleep disorder, neurologic disorder, a mental disorder, or the use of drugs(1). The essential clinical criterion of KLS is recurrent episodes of hypersomnia. Moreover, patients have to experience at least one of these symptoms only during the episodes: (*i*) cognitive or mood disturbances, (*ii*) megaphagia with compulsive eating; (*iii*) hypersexuality with inappropriate or odd behaviour; and (*iv*) abnormal behavior. Thus our patient satisfied the essential criteria for diagnosis, i.e., hypersomnia along with megaphagia and behavioral disturbances.

A total of 186 cases have been reported so far in literature till 2004 including 10 from India. Age at onset was 16.9±8.5 years with a median of 15 years and a range of 4-82 years(2). The youngest case of KLS reported so far in literature was a 4 year old boy from China(3). A history of head injury as a precipitating cause has been reported in 9% cases.

A multitude of drugs have been tried in the treatment of this condition which include amphetamines, modanafil, methylphenidate, fluoxetine and lithium. Of these, only lithium had a reported response rate higher than medical abstension(5).

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