CASE REPORT

“Sleeping Beauty” – A Case of Kleine-Levin Syndrome

Amitabh Saha

Level III Hospital, UN Mission, Democratic Republic of Congo

Corresponding author: Amitabh Saha, M.D., Level III Hospital, UN Mission, Democratic Republic of Congo; E-mail: amipal321@yahoo.co.in

Abstract

This is a case of Kleine-Levin Syndrome, which is described in a teenage girl who presented with features of hypersomnia, hyperphagia, and hypersexuality. The patient’s initial presentation was with academic decline due to excessive somnolence, which was recurrent in nature. She was started on stimulants and lithium medications, to which she showed favorable response (German J Psychiatry 2008; 11: 73-75).

Keywords: hypersexuality; hypersomnia; Kleine-Levin syndrome; megaphagia; periodic

Introduction

Kleine-Levin Syndrome is a rare disorder that causes recurring periods of excessive drowsiness and sleep (up to 20 hours per day). Symptoms, which may last for days to weeks, include excessive food intake, irritability, disorientation, lack of energy, and an abnormally uninhibited sex drive. Affected persons are normal between episodes, although depression and amnesia may be noted temporarily after an attack. It may be weeks or more before symptoms reappear. Onset is typically around adolescence to the late teens. Symptoms may be related to malfunction of the hypothalamus, the part of the brain that governs appetite and sleep (American Academy of Sleep Medicine, 2005).

Multiple cases of recurrent hypersomnia were first reported in Frankfurt by Willi Kleine in 1925 (Kleine, 1925). Max Levin (1929, 1936) emphasized the association of periodic somnolence with morbid hunger in 1929 and 1936. Prodromal symptoms included sudden overwhelming tiredness, i.e. ‘feeling drawn towards his bed’, or ‘reluctant to get up in the morning’ (Prabhakaran et al., 1970). At the end of an episode, a short-lasting insomnia was noted in three cases (Galinek, 1967; Frosher et al., 1991; Russell and Grunstein, 1992). Sleep symptoms changed from frank hypersomnia during the first episodes to a sense of “heavy fatigue”

Mood disturbances described are irritability, depression, euphoria and lability. Confusion, amnesia, delusions, thought blocking, vivid dreams, auditory and visual hallucinations have also been described during the episodes. The onset of the illness is usually spontaneous and appears to be more commonly seen in males. The duration of the episodes may range, from a few days to several weeks with normal inter-episodic periods lasting a few days to several months. The mean frequency of episodes may be two per year and may vary from one to twelve per year. The syndrome seems to ‘burn itself out’ spontaneously in due course without any therapeutic intervention. However they may recur in adulthood and eventually cease altogether but no associated mortality is reported.

Case Report

A 16 year old girl, belonging to poor socioeconomic background, with no past or family h/o psychiatric illness, presented with complaints of excessive sleep, academic decline, increased appetite and excessive irritability of 02 years duration. The onset was insidious and gradual in progression and was noted by parents initially. She would sleep at times for 14 to 16 hours in a day much to the surprise of her mother. The patient would find it difficult to stay awake during her school classes much to the annoyance of her teacher who would keep complaining the matter to her parents and school authorities. At these times she would feel lazy, lethargic, would seem as if to “search for her bed”. During her waking hours she was noted to have a voracious appetite. These symptoms would continue for about three week’s
before finally settling down. However it would recur after approximately 3 to 4 months in a year with similar symptomatology but in between this period she would behave appropriately. In these two years the girl had to change her school twice due to constant failure and poor performance in her examinations as she was unable to concentrate properly. The patient also developed an excessive sexual urge as noted by her mother who would see many boys frequenting her house and on direct questioning the patient later accepted having sexual intercourse with her boy friends. Mother noted the patient to be very irritable whenever she would be scolded and would go in phases of low mood wherein she would appear aloof and withdrawn. The parents even had to shift houses and move into new neighborhoods on account of the patients increasing sexual disinhibition and resultant sexual partnerships which was much to the annoyance of the people living in the same community.

As the symptoms persisted with almost a periodic pattern consisting of the triad of hypersexuality, hypersomnia and megaphagia with associated academic decline the parents sought psychiatric consultation for her. Detailed psychiatric history helped in revealing no h/o seizures, psychosis, mood disorder, substance use etc. She was noted to have gained weight of about 16 Kilograms in this two year period. Her previous weight before start of symptoms as per parents was about 41 kilograms. At time of admission she weighed 57 kilograms. The typical symptoms of cataplexy, sleep paralysis, hypnagogic hallucinations and automatic behavior characteristic of narcolepsy was however not noted.

Mental state examination revealed a tense and anxious girl who appeared preoccupied with her problems. She appeared groggy and at times had to strain to concentrate on the questions being posed to her. Psychomotor retardation activity was reduced, no prominent depressive cognitions were noted, no evidence of psychosis was seen. Biodrives were deranged in that sleep was increased, she had increased libidinal urge and her appetite was increased with reduced energy levels.

Detailed medical evaluation including neurological examination was done which revealed no significant findings. Radiological investigations were normal, EEG was normal study and all other detailed reports too were not significant.

She was managed with methylphenidate which was slowly increased from 5 mg/day to 20 mg/day. Lithium was introduced gradually from 600 mg daily in divided doses to 1200 mg per day and psychotherapy was combined after adequate rapport could be established. The patient started to show favorable response to therapy and after one year follow up has maintained remission with no episodes of excessive somnolence She showed improvement in academic spheres and with parental support has been compliant with medications. Her weight also reduced, by about 06 kilograms over this period of follow up and weighed about 51 kilograms at time last reviewed.

Discussion

As seen in this case, the patient had symptoms of hyper-somnia which is the most prominent symptom reported during each of the episodes. Other behavioral disturbances as noted in this syndrome like suicidal tendencies, motor retardation, pathological guilt. vivid imagery, visual and auditory hallucinations, features reported elsewhere, were not reported during or between attacks of hypersomnia in the patient described. The prominent symptoms of Narcolepsy which is seen commonly with this disorder was not observed in this patient. It has been seen from various studies done earlier that Narcolepsy, at times, need not be the presenting or main features in this otherwise uncommon disorder. Precipitating factors that have received consideration are hot weather, stress, head trauma, post flu and encephalitis among others but none were clearly recognized in this case (Dale et al., 2004).

The benefit of stimulants and mood stabilizers needs to be emphasized in the management of these cases. Lithium therapy has been of benefit in patients with Klein-Levin syndrome as can be ascertained from case reports (Kellett, 1977; Smolik and Roth, 1988; Poppe et al., 2003). Valproic acid and carbamazepine have also been claimed to be of therapeutic benefit (Mukaddes et al., 1999b). However it is important to exclude mood disorders and sleepiness during the premenstrual period in teenaged girls. Newer drugs like modafinil have also been introduced to treat specific symptoms of sleepiness. In conclusion early diagnosis, treatment with patient education remain vital for alleviating suffering, loss of schooling or work and allaying anxieties of family members, teachers and school or work colleagues.

References

