

An Adolescent with Kleine-levin Syndrome: A Case Report

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Kleine-levin syndrome is characterized as a periodic hypersomnia crisis as well as dietary and variable significant psychic symptoms. This report describes the finding on an evaluation of a 15-year-old girl with KLS. She had amnesia to her episodes. All episodes were correlated with menstrual period. Polysomnographic finding of this patient was normal with 5 sleep cycles in sleep architecture and sleep efficiency of 99%. Decreasing in stage I, increasing of stage III and IV and decreasing in REM sleep was notable. Dextroamphetamine was given to the patient in order to decrease sleepiness.

Kleine-levin syndrome should be considered in patients with periodic hypersomnia.

Key Words: Adolescent, Hypersomnia, Sleep disorders

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Kleine-levin syndrome is a rare disorder which affects mainly young men 10 to 25 years of age. Kleine-levin syndrome is characterized as a periodic hypersomnia crisis as well as dietary and variable significant psychic symptoms. Individuals are normal between the attacks (1). The cause and pathogenesis of KLS remains unknown. Many etiologies have been considered, especially regarding a hypothalamic dysfunction. Physiologic, radiologic, anatomical and polysomnographic studies have not shown clearly conclusive results, but the syndrome appears to be related to affective disorders (2). This report describes the finding on an evaluation of a 15-year-old girl with KLS.

Interestingly, of the 186 reported cases, 53 were from Asia and only 3 cases were from Iran (2). This case has a positive family history that is not often observed in these cases. Correlation of all the episodes with menstrual period is interesting. She also experienced functional impairment which is unusual in Kleine-levin syndrome.

Case Report

This adolescent girl was referred to the child and adolescent psychiatric clinic of Iran hospital for her hypersomnic and hyperphagic episodes. She had experienced such episodes since 5 years ago. Each episode had lasted 3 to 7 days. In the last year, duration of her episode had increased to 10 days. In each episode, she had irresistible urge to sleep for more than 16 hours and after long hours of sleep she ate a large amount of food, sweets and chocolate. After finishing the episode, she could not exactly remember the past event.

In the last year, she experienced one episode every 2.5 months. She showed functional decline in academic performance due to several absence from school. It should be noted that after her menarche at 13, all the episodes were correlated with menstrual period. Her episodes had been started 3 to 7 days before beginning of her menses. The patient stated that with starting menstrual period she felt that 4 hours of sleep was enough. She experienced elated mood and talkativeness. She had elevated energy, sang with a loud voice and liked to relate with boys. These symptoms disappeared within 7 days. Her mother and one of her brothers suffered from similar episodes. This disorder should be differentiated from narcolepsy and sleep apnea hypersomnia syndrome. Each of these disorders has certain symptoms that help clinicians in the diagnosis.

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Diagnostic evaluation

Computerized tomography and EEG of the brain was normal. The patient underwent standard overnight polysomnography. Raw data was manually scored in 30 seconds for sleep stages using standard criteria (3). Respiratory events and limb movements were defined according to AASM standard criteria. During the standard PSG, total sleep time was measured 454 minutes with sleep efficiency of 99%. Sleep latency was 3.5 minutes and REM latency was 128.5 minutes. Distribution of sleep stages was:

Stage I 1.9%, stage II 51.3%, stage III 14.3%, stage IV 21.6% and REM stage 10%.

ECG study showed normal sinus rhythm with mean heart rate of 81. RDI was 2.4/hour sleep. Mean Sao₂ was 95.1% and 0.8 minutes < 90%. PLMS Index was 3.8. These findings excluded sleep-related breathing disorders and PLMS as a cause of the patient's sleepiness.

Treatment

To decrease sleepiness, Dextroamphetamin was given to the patient. At the four months follow up, she did not experience new episodes. Among stimulants, only amphetamines significantly reduced sleepiness in these patients.

Discussion

Primary KLS cases were found mostly in men (68%), but the patient in this case was a female. In the systematic review of the 186 cases, it has been observed that KLS lasted longer in women and in patients with less frequent episodes during the first year. The age of onset for this patient was 10, but the age of recognition for her was 15. The age of onset in this patient was lower than usual. In 81% of all the studied cases, KLS onset occurred during the second decade.

In this case, after her menarche at 13, all episodes were correlated with menstrual period. In some other patients, lactation and menses(4), but not at menarche, were factors reported occasionally at KLS onset.

Mood change was observed in patients with KLS. A few cases (8%) reported to be hypomanic for a couple of days at the end of a KLS episode (5, 6). Half of the patients had a depressive mood during the episodes. Fifteen percent of the patients reported suicidal thoughts and two patients attempted suicide (7, 8).

Nearly half of the patients had symptoms consistent with hypersexuality during their episodes.

She could not remember the past event exactly after the episode. Many of such patients reported amnesia of the events that occurred during an attack.

Despite a few cases in systematic review (8/168) that reported academic decline, our patient showed functional decline in academic performance due to several absence from school(2).

Interestingly, the mother and brother of this patient had experienced similar episodes; this could be accounted for the long interval between the onset and referral of the patient. The majority (98%) of cases in the systematic review of patients were sporadic (2).

Most of these patients showed relatively normal sleep structure with decreased sleep efficiency due to numerous awakenings from the sleep stage 2 (9). However, the polysomnographic finding of this patient was normal sleep architecture with 5 sleep cycles and sleep efficiency of 99%. Decreasing in stage I, increasing of stage III and IV and decreasing in REM sleep was notable.

Kleine-Levin syndrome is a clinical diagnosis which should be considered in patients with periodic hypersomnia.

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