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EC PSYCHOLOGY AND PSYCHIATRY
Case Report

A Case of Behavioral Change Associated with Kleine-Levin Syndrome

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Received: June 17, 2021; **Published:** August 29, 2022

Abstract

Kleine-Levin syndromeis a cognitive and behavioral disorder. It is a rare and unique recurrent disease with episodes of excessive sleeping, overeating, and excessive sexual desire. The first excessive sleep attack is usually triggered by an infection, although the exact cause is not known yet, other hypotheses include catching a chill, a blow to the head after anesthesia, alcohol, and a severe emotional trauma. Boys are affected more often than girls. More than half of the patients have binge eating, excessive sexual desire (mainly men) or depressed mood (mostly women) and 30% of patients experience anxiiety, delusions and hallucinations. Between attacks, patients usually have normal sleep patterns, cognition, mood and eating habits. In this study, a 27-year-old, who has hyper-sleeping attacks, having hallucinatory dreams with sexual contents, being in a state of confusion between sleep, being only able to meet the need for toilet and food, eating foods that he normally despise in an impolite way and overeating, excessive irritability. In the light of the literature, this case applied to the sleep polyclinic with complaints of sexual desire and excessive sexuality, excessively obscene and vulgar speech during sexual intercourse and diagnosed with Kleine-Levin syndrome due to increased daytime sleepiness and recurrent attacks is presented in the light of the literature.

Keywords: *Kleine-Levin Syndrome; Behavioral Disorders; Sleep Disorders; Polysomnography*

Introduction

KLS, first described by Kleine in 1925, is a rare neuropsychiatric disorder characterized by recurrent episodes of hypersomnia, varying degrees of behavioral and cognitive disorders, compulsive eatin, behavior (hyperphagia) and excessive sexual desire (hypersexuality) [1]. Attack in the syndrome progressing in episodes; starts abruptly, lasts from a few days to a few weeks, and ends abruptly. There is no sleep disorder in the periods between attacks, the person is physically and mentally healthy. Cases with an attack frequency of two to twelve attacks per year have been reported. In the symptomatic period, the patient spends most of the day asleep and gets out of bed for toilet and food needs. Behavioral disorders such as hyperphagia and hypersexuality are compulsive [1].

American Academy of Sleep Medicine, in its International Classification of Sleep Disorders Third Classification (ICSD3) (American Academy of Sleep Medicine) among the central disorders of the episode of hypersomnolence; There are two types of narcolepsy type 1 (NT1) and narcolepsy type 2 (NT2). Other hypersomnolent disorders include idiopathic hypersomnia and Kleine-Levin syndrome [2]. Diagnostic criteria for periodic hypersomnia, including Kleine-Levin Syndrome (KLS), are hypersomnia episodes lasting from two days to four weeks, episodes occurring at least once a year, normal arousal between attacks, normal physical functions and behaviors, It was defined as hypersomnia that could not be explained by another sleep disorder, medical or neurological disorder, mental disorder, drug or substance use. When defining the KLS subtype, it is stated that these symptoms should be accompanied by behavioral and cognitive symptoms (AASM 2005).

KLS is a very rare disease with unknown prevalence and etiopathogenesis. It is 9 times more common in men than women.It has been reported that it usually occurs after head trauma, following psychogenic stress or viral infections [3,4]. It is suggested that disorders such as hypothalamic dysfunction involving abnormal growth hormone and prolactin secretion, dysfunction in serotonergic and dopaminergic pathways play a role [5]. In this article, we discuss our 27-year-old male case with hypersomnia, hyperphagia and hypersexuality, apathy, slowing in thought and memory, prominent behavioral disorders and a good prognosis in line with the literature, and lithium treatment is effective.

Case Report

Our patient, a 27-year-old male who works as a technical staff in a media organization; He was brought to our outpatient clinic by his colleague because of his history of excessive sleeping in the form of attacks at work and his inability to do his work.

He had a severe flu infection about 4 months before his complaints, and his complaints started 2 - 3 months ago, again in the form of an attack; It lasted for about 15 - 17 days, The first attack was mild and spontaneously resolved, the second recurrent attack was more severe, there was a change in behavior during the attacks, he behaved extremely nervously, had sexual dreams, often confused with reality because his dreams were hallucinatory. It was learned that he could not go to work because he was sleeping all day, he woke up in a confused state only for his physical needs such as the need for toilet and food and when he woke up he could not understand whether what was happening around him was real.

When the relatives of the patient were interviewed, it was learned that the patient had another importent condition. The patient is too hasty; It was learned that he ate quickly and in fact too much that he could not eat normally, and things that his stomach could not normally take, without following the rules of etiquette.

The patient stated that he had excessive sexuality between waking up with his girlfriend, but even in this case, he felt like he was in a dream, had dreams with excessive sexual content that he confused with reality and desired excessive sexuality, used obscene words during sexual intercourse that his girlfriend said but he did not remember, and spoke extremely rude. According to his colleague and other teammates; They called the patient because he had not come to work for about 10 days and they got a response that the patient was sleeping all the time, they told him that he had to go to the doctor, but they learned that he preferred to sleep all the time instead of going to the doctor. They said that they forced the patient to come to the workplace due to excessive disruption of the work, they could not believe the changes they saw in the patient at the beginning, they thought that the patient did not want to come to work, they thought he was pretending. They said that they forced the patient to come to the workplace due to excessive disruption of the work, they could not believe the changes they saw in the patient at the beginning, they thought that the patient did not want to come to work, they thought he was pretending. Then, when the patient came to the workplace by force, they saw that he was sleeping all the time at the workplace and they understood the situation; They said that the previous personality of the patient was a hardworking person, and they reported that they had to bring the patient to the doctor themselves because their work had come to a standstill. Mayer G., *et al*. (1988), with 5 subjects

in symptomatic and asymptomatic periods; In the study they conducted on 4 cases aged 17 - 28 and 1 female case aged 30 years, diagnosed with Kleine Levin Syndrome, they found that all patients had normal neuroimaging.

Laboratory examinations show that it is generally within normal limits during and between attacks, basal levels of pituitary and other hormones are normal, CSF protein is found to be within normal limits, although it is found to be slightly increased, on EEG the background activity generally slows down, Sometimes there are 4 - 6 Hz paroxysmal waves [6]. Because of the largely negative findings, Kleine Levin Syndrome is generally considered a functional disorder of limbic and/or hypothalamic structures [7]. General physical examination and systemic examination of our case were within normal limits. No abnormality was observed in the brain in EEG and MRI scans, and no characteristic was detected in laboratory values.

In the literature, a psychogenic mechanism has been implicated in the etiopathogenesis of Kleine Levin Syndrome, but no basis has been demonstrated [8,9]. In our case, perceived psychological stress in the home, work, and family areas was not detected, and the symptoms due to relapses were observed to improve in approximately 14 - 18 days. One night polysomnography in the sleep laboratory and snoring, sleep apnea syndrome and periodic leg movement disorder that could cause hypersomnia were not detected in PSG that continued with MSLT for four times the next day. In both examinations, it was determined that the patient always preferred to stay in bed and sleep, except for his physiological needs, and that he was in a continuous sleep, albeit in fragments, with short-term awakenings in which his sleep was interrupted by frequent waking reactions during the night and daytime hours. It was determined that he was always sleepy and irritable in the confusional state even during the waking periods, and his deep sleep was relatively reduced. In MSLT, it was determined that sleep begins with REM (SOREM) in 3 out of 4 recordings. In the light of the information received from the patient and his relatives, he was diagnosed with Kleıne Levın syndrome due to increased daytime sleepiness and recurrent attacks. Growth hormone (GH) and cortisol levels were checked and they were found to be normal.

It was learned that there was no one in the family of the patient who was clinically diagnosed with Kleine Levin Syndrome and had similar complaints before. The patients and his relatives were informed about the disease, and suggestions were made during the attack periods.

Treatment was started with Lithium and modafinil Lithium. It was gradually increased to 3x 300 mg. While under lithium and modafinil treatment, it was learned that the patient still had attacks at the 6th month follow-up, but hypersomnia attacks lasted shorter, and behavioral symptoms were absent, and it was at a level that would affect work performance slightly. At the end of the 9th month, the patient had his 5th attack, albeit mild, and in addition to the treatment, carbamazepine was started, it was increased to 2 x 400 mg, and the patient's attacks ended.

Discussion

Kleine-Levin syndrome is a rare sleep disorder that mostly affects males, with intermittent hypersomnolence, behavioral and cognitive changes, hyperphagia and, in some cases, hypersexuality.

In the study in which Critcheley collected a number of cases, and he found that the age range extended from adolescence to the age of 45, there were relapses ranging from two to twelve in one year [10]. Each attack may last one or two weeks, and affected people are completely asymptomatic between their attacks.

A definite cause has not been determined, but hypothalamus dysfunction is emphasized. Attacks can occur every few weeks or once a month. Kleine-Levin syndrome may resolve spontaneously or may persist for ten years or more. Although there is no effective treatment, the prognosis of this disorder is good in general, in our case, the prognosis was good, supporting the literature. Our case was a 27-year-old

male with each episode lasting approximately 14 to 21 days and relapsed within four months to six months. In addition to stimulants such as methylphenidate and modafinil that we use for treatment, mood stabilizer lithium carbonate has also been successfully used in the treatment of many patients in the literature.

Conclusion

Kleine-Levin syndrome (KLS) is a rare neuropsychiatric disorder that mainly affects young men, the main features of which are intermittent hypersomnolence, behavioral and cognitive disorders, hyperphagia, and in some cases, hypersexuality. It occurs in episodes of sudden onset following a viral infection, head injury, or psychogenic stress. Between attacks, patients are physically and mentally normal. There are no objective diagnostic criteria for the diagnosis of KLS. Electroencephalography and polysomnography studies of the patients do not include disease-specific changes. Polysomnographic findings of this syndrome include decreased sleep period, increased number of awakenings during the night, decreased deep sleep and shortened REM latency.

KLS should be considered in the differential diagnosis of hypersomnia attacks, especially in young male patients. In this article, the clinical picture of a 27-year-old male patient who was followed up in our clinic and accompanied by behavioral disorders was examined. The polysomnographic findings of our patient have the typical features seen in KLS. In polysomnography examination; It was determined that his deep sleep was relatively reduced. It was determined that sleep started with REM (rem latency shortened) in 3 of 4 recordings made in MSLT.

Although KLS is a very rare syndrome, its response to treatment is quite good. In the treatment of KLS, modafinil, methylphenidate, valproic acid, carbamazepine and melatonin are the most preferred treatment options [11]. In the treatment of KLS; stated that lithium treatment reduced the frequency of attacks or stopped the attacks [11]. In resistant cases, carbamazepine and lithium treatment can be used together [12]. In our case, lithium, modafinil and carbamazepine were used and response to the treatment was obtained. Response to treatment can also be accepted as a supporting finding for the diagnosis of KLS. In the future, we need more research on the genetic etiology and treatment management of this disorder.

Bibliography

1. Karadağ Hasan., *et al*. "Kleine-Levin Syndrome: a case report". *Clinical Psychiatry* 13 (2010): 92-96.
2. Thorpy Michael. "International classification of sleep disorders". Sleep Disorders Medicine. Springer, New York, NY (2017): 475-484.
3. Arnulf Isabelle., *et al*. "Kleine-Levin syndrome: a systematic review of 186 cases in the literature". *Brain* 128.12 (2005): 2763-2776.
4. Jeffries J Joel and Arlette Lefebvre. "Depression and mania associated with Kleine-Levin-Critchley syndrome". *Canadian Psychiatric Association Journal* 18.5 (1973): 439-444.
5. Karadağ H., *et al*. "Kleine-Levin Syndrome: a case report". *Clinical Psychiatry* 13 (2010): 92-96.
6. Mayer G., *et al*. "Kleine Levin syndrome". *Sleep* 21.3 (1998): 278-284.
7. Parkes JD. "Disorders of sleep". In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD, editors. Neurology in Clinical practice: Principles of Diagnosis and Management. 2nd edition. Gold Worth Heinemann (1996): 1666-1667.
8. Chakraborty PK., *et al*. "Kleine levin syndrome". *Medical Journal Armed Forces India* 57.4 (2001): 335-336.
9. Adams RD., *et al*. "Principles of Neurology". 6th edition. McGraw Mill Inc. (1997): 394.

10. Critcheley M. "Periodic hypersomnia and megaphagia in adolescent males". *Brain* 85 (1962): 627-656.

11. Muratori F., *et al*. "Efficacy of lithium treatment in Kleine-Levin syndrome". *European Psychiatry* 17.4 (2002): 232-233.

12. Karadeniz D., *et al*. "Monosymptomatic Kleine-Levin syndrome: A Case Report". *Cerrahpaşa Medical Journal* 31 (2000): 243-248.

Volume 11 Issue 9 September 2022