Kleine-Levin syndrome: A Case Report from Iran

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Kleine-Levin syndrome (KLS) is a rare syndrome characterized by periodic hypersomnia, hyperphagia, behavioral disturbances, and in some instances hypersexuality. This report depicts a17-year-old boy with KLS. He has experienced long periods of hypersomnia, sometimes up to 40 days. During these periods he was impatient, aggressive, depersonalized, suffering from hyperphagia, and amnesia. He did not have abnormal neurologic sings and his brain CT, EEG, and routine laboratory tests were normal. Fluoxetine, lithium and carbamazepine were given to him to reduce the signs of his disease. In conclusion, Kleine-Levin syndrome should be considered in patients with periodic hypersomnia.

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Introduction

Kleine-Levin syndrome, sometimes known as Rip van Winkle disease, is a rare sleep disorder mainly affecting adolescent males. The main features of this disorder are intermittent hypersomnolence, behavioral and cognitive disturbances, dysautonomia, hyperphagia and in some cases hypersexuality (1). Unexplained fever has been reported to occur in a few patients (2). Episodes are separated by weeks or months of normal sleep and behavior (1). The syndrome appears to be almost invariably self-limited, and enduring remission occurs spontaneously before the age of 40 in early-onset cases (2). The etiology and pathophysiology of it remain unknown but hypothalamic dysfunction, genetic predisposition combined with environmental factors and autoimmune etiologies have been suggested (3). Diagnosis is usually based on clinical manifestations. Physical examination including neurological evaluation is usually normal. EEG, brain imaging and CSF examination are normal (3). Some medications like fluoxetine, lithium, carbamazepine, valproate, and moclobemide have been reported to provide some benefits for the treatment of patients with Kleine-Levin syndrome. Among stimulants, only amphetamines significantly reduce sleepiness in these patients (3). The presence of diffuse paroxysmal slowing in EEG of some patients might indicate a convulsive disorder (5). Interestingly, of the 186 reported cases, 53 were from Asia and only 3 cases were from Iran (2). This case report depicts a 17-year-old male with typical manifestations of KLS.

Case Report

This 17-year-old boy was referred to the psychiatric clinic of Rasoul Akram Hospital for his hypersomnic periods. Following a common cold and taking anti-histamine tablets, he had unexpectedly experienced hypersomnic periods lasted about 40 days. The duration of his episodes was approximately 20 hours. He has experienced these episodes since 2 years ago. In each episode, he had irresistible urge to sleep for more than 20 hours and after a long period of sleeping he ate large

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amount of food rapidly without mastication. His family mentioned that he was laconic, uncommunicative, aggressive, impatient, taciturn, downhearted and disappointed during these periods. He also suffered from loss of energy and interests and had 6 attempted suicides.

He had seizure-like movements accompanied by snoring and limb stiffness that lasted 30 minutes during his second period of hypersomnia which lasted 3 months. He had not experienced defecation and urinary incontinence and mentioned that he was conscious during the attack and enabled to remember next events.

Despite the increasing number of attacks, duration of them has reduced from 40 days to 18 days.

In his past medical history, he had a motor vehicle accident without head trauma and also varicocele surgery. His social history showed that he was nicotine smoker of 2-3 cigarettes per day for the past year. According to his family history, his father, uncle and grandmother might have suffered from depressive disorder.

He did not have any problem in his prenatal and perinatal periods and also early childhood. Nonetheless, in his middle to early young adulthood (11 to 20 years of age), he had functional decline in academic performances and had to leave the school at grade 10. It should be taken into account that he quitted school due to his indifference for education, not for his disorder. He did not mention hypersexuality. In his mental status examination, his speech was hypotonic, monotonic and hypovolemic. Moreover, he showed depressed mood and affect, hopelessness, impaired concentration and problems in recent memory.

His EEG and brain Computerized tomography (CT) were normal. Routine laboratory tests such as thyroid and liver function tests, blood sugar and CSF analysis were normal as well. Polysomnography was not performed for him because it is an expensive test and his family was in a low socio-economic condition and did not have satisfactory cooperation.

Treatment started when he was in the active phase of disease with fluoxetine (40 mg/daily) followed by lithium (900 mg/daily in three separate doses), and carbamazepine (400 mg/daily). Two different mood stabilizers were used because he did not response to the first drug appropriately. After 13 days, his signs and symptoms decreased, hyperphagia and aggression were controlled, the episodes of his sleeps reduced and he was discharged.

Although he showed good compliance with treatment, his problems relapsed after three months.

Discussion

The goal of this paper was to present a KLS patient with typical patterns. Herein, we have compared some characteristics of KLS with our patient's signs and symptoms. Disorder of the present case has been differentiated from Major depressive disorder (MDD), other mood disorders, narcolepsy and primary hypersomnia, each of which has certain symptoms helping clinicians with diagnosis. MDD involves relatively normal sleep onset, repeated awakenings during the second half of the night and early morning awakening. Sleep attacks of narcolepsy represent episodes of irresistible sleepiness often associated with cataplexy, after which the patient feels refreshed. Sleep paralysis is an uncommon symptom observed in these periods (2). Primary hypersomnia is a normal variation without complaints about difficulties with the awakening motivation and performance (4).

This patient confirms the fact that KLS occurs more frequently in males (9). In 81% of all studied cases, KLS onset was during the second decade of life. The age of the disease onset was 17 in this patient as well. Mood changes are significant problems occur during the syndrome (7). In this regard, fifteen percent of the patients reported suicidal thoughts (8,11). As it has been mentioned earlier this patient was completely hopeless and had 6 attempted suicides.

Some KLS predisposing factors include viral infections like influenza, Ebstein-barr virus, varicella zoster virus (VZV), tonsillitis, and urinary tract infection (UTI). It is interesting that primary signs of KLS in our patient began with a common cold and taking

anti-histamine.

Instances of hypersexuality have also been reported in KLS (14) but we could not detect it in our patient.

Despite the fact that only a few KLS cases with academic decline have been reported in a previous systematic review, this patient had a considerable decline in his academic performances which finally led to leaving the school (6).

It should be taken into account that focal neurologic defects are rare in KLS (2,13), and not every patient affected by KLS exhibits all symptoms and signs described in the literature (12).

There were not any soft signs in this patient. Patients with KLS might have a complete remission and good prognosis. Although the duration of the syndrome might be 4 to 8 years, manifestations might be more prolonged and severe in females.

Mood stabilizers were prescribed for him according to some previous successful experiences of treating KLS patients with these medications (7). While somnolence decreased in 40% of cases using stimulants in another study, neuroleptics and antidepressants were of poor benefit. Only lithium (but not carbamazepine or other antiepileptics) had a higher reported response rate (41%) of stopping relapses in comparison with medical abstention (19%) (6).

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