CASE REPORT

A boy with episodic hypersomnolence- a case report

S Bihari, N Ramakrishnan
Nithra Institute of Sleep Sciences, Chennai

Indian J Sleep Med 2007; 2.3, 109-111

Abstract

A 15 year old boy presented with complaints of episodic hypersomnolence, sleeping for 16 to 18 hours a day which would happen for continuously 7 to 8 days, seven to eight times a year for the past 10 years. There was also history of voracious eating during such episodes. There was no history of trauma, seizure -like activity and no significant family history or psychosocial history. His blood sugar, growth hormone, prolactin levels were all normal. MRI brain with contrast done was also normal. Sixteen channel overnight polysomnography and Multiple Sleep Latency Test (MSLT) were done and found to be normal. Clinical features and significant negative tests led to a diagnosis of Kleine-Levin syndrome. He was started on Lithium and being monitored for reduction in frequency of symptoms.

Keywords: Hypersomnolence, Polysomnography, Multiple Sleep Latency Test, Klein-Levin Syndrome, Lithium

Abbreviations

KLS: Kleine-Levin syndrome; **REM:** Rapid eye movement**; SPECT:** Single photon emission computed tomography

Key Messages : Kleine-Levin syndrome is usually found in adolescent males associated with episodic hypersomnolence compulsive overeating, lack of sexual inhibition and personality change, most often it represents a benign and self-limited entity and does not warrant extensive investigations or treatment

Introduction

Kleine-Levin syndrome is usually a self limiting benign disorder usually affecting adolescent males. It is important to recognize this disorder early as delay in diagnosis leads to unnecessary extensive investigations and empiric treatment

Case history

A 15-year-old boy was brought by his parents with complaints of episodic hypersomnolence. His parents were concerned that his episodic hypersomnolence may affect his board examinations. On further enquiry he had a history of sleeping for 16-18 hours a day which would happen for continuously 7-8 days, seven to eight times a year for the past 10 years. Apparently he had no 'warning symptoms' and could not predict when he would have such episodes. In addition to sleeping 16 to 18 hours per day, he was also reported to eat voraciously while awake. If woken up from the sleep he was rather irritable and went to sleep quickly. Barring these episodes he was a normal child with normal intelligence and behavior. He ranked in the top 10th percentile in his class and had no other significant medical history. In particular, there was no history of trauma, seizure like activity, snoring or hypersomnolence at other times. There was no significant family history or psychosocial history.

On examination his height and weight were normal for age. Investigations done prior to referral to our Sleep Center including liver functions, renal functions, thyroid functions, blood sugar, growth hormone, prolactin levels

Address for correspondence:
Dr. N. Ramakrishnan
Nithra Institute of Sleep Sciences
Chennai

were all normal. MRI brain with contrast was normal. A 16 channel over night polysomnography was done and found to be normal. In particular, the sleep architecture was normal and there was no evidence of sleep disordered breathing or periodic limb movement disorder. Multiple Sleep Latency Test done the following day did not suggest hypersomnolence and there were no Sleep onset REM periods. Of note, the study was done during patient's reported normal days and not during one of his 'episodic' abnormality as the frequency of these was unpredictable. A diagnosis of Kleine-Levin syndrome (KLS) was made. It was decided to start him on Lithium ant to monitor the frequency of his symptoms.

Discussion

Kleine-Levin syndrome is a rare disorder characterized by the need for excessive amounts of sleep (hypersomnolence), (i.e., up to 20 hours a day); excessive food intake (compulsive hyperphagia); and an abnormally uninhibited sexual drive occurring almost exclusively in adolescent males though there have been few reports of its occurrence in females as well[1]. Mean age of onset is 15.8 $\pm$ 2.8 years and usually patients experience three to four episodes per year with mean duration of single hyper somnolent attack of 11.5 $\pm$ 6.6 days[2]. It has been noted that the disease usually lasts 8-10 years and is self-limiting before adulthood.

Though the cause is usually not known it is believed that hereditary factors may cause some individuals to have a genetic predisposition to developing the disorder. There have been reports of two siblings who shared uncharacteristically prolonged episodes of hypersomnolence, and the HLA-DR2 haplotype[3]. Secondary Kleine-Levin syndrome have been described after episodes of encephalitis[4] and trauma [5,6]

KLS is usually associated with compulsive overeating, lack of sexual inhibition and personality change. Sexual responses though not always present include inappropriate sexual advances and overt masturbation, especially in males. Compulsive overeating with rapid weight gain may occur. Personality changes may include irritability, depersonalization, depression, confusion, occasional hallucinations and impulsive behavior. Symptoms of KLS are typically cyclical. On recovery, total or partial loss of memory (amnesia) for what has happened is usual, although disgust at overeating is common. Between episodes, physical and mental health is usually normal.

Diagnosis and Treatment

It is not uncommon to note that these patients are seen by a variety of specialists including endocrinologists, psychiatrist and neurologist before being evaluated by a sleep specialist. The mean delay in diagnosing Kleine-Levin syndrome is reportedly 3.8 $\pm$ 4.2 years[2].

All imaging tests except SPECT are usually normal. The subtracted SPECT studies may show hypoperfusion in the left hypothalamus, bilateral thalami, basal ganglia, bilateral medial and dorsolateral frontal regions, and left temporal lobe during the symptomatic period[7]. CSF hypocretin-1 measurements were performed during a period of hypersomnia has also been found to be abnormal [8].

Currently there is no formal treatment for KLS due to the lack of knowledge regarding its underlying cause. Only lithium[9] had a higher reported response rate (41%) for stopping relapses when compared to medical abstention (19%). The goal of treatment is to counsel the patient and family and also keep these episodes to a minimum level through out the year.

Most of these patients are either labeled as lazy or classified as narcolepsy or idiopathic hypersomnia. But absence of associated clinical features such as cataplexy and characteristic polysomnographic features such as sleep-onset REM episodes and presence of polyphagia render positive diagnosis

It is important that the diagnosis is suspected early, especially in adolescent males who present with recurrent episodes of somnolence, increased appetite, and abnormal behavior, since it most often represents a benign and self-limited entity and does not warrant extensive investigations or treatment. It is also important to distinguish this syndrome from more serious organic and psychiatric diseases with more serious prognosis.

References

1. **Kesler A**, Gadoth N, Vainstein G, Peled R, Lavie P. Kleine Levin syndrome (KLS) in young females. *Sleep*. 2000 Jun 15; 23(4): 563-7.

2. **Gadoth N**, Kesler A, Vainstein G, Peled R, Lavie P . Clinical and polysomnographic characteristics of 34 patients with Kleine-Levin syndrome. *J Sleep Res*. 2001 Dec; 10(4): 337-41.

3. **Katz JD**, Ropper AH. Familial Kleine-Levin syndrome: two siblings with unusually long hypersomnic spells. *Arch Neurol*. 2002 Dec;59(12):1959-61

4. **Sethi S**, Bhargava Subhash C Kleine-Levin syndrome and encephalitis. *Indian journal of pediatrics* 2002. Volume : 69,11: 999-1000

5. **Cheung G**. Posttraumatic Kleine-Levin syndrome. *Gen Hosp Psychiatry*. 2006 Sep-Oct;28(5):443-5

6. **Pelin Z**, Ozturk L, Bozluolcay M. Posttraumatic Kleine-Levin syndrome: a case report. *Eur Psychiatry*. 2004 Dec;19(8):521-2.

7. **Huang YS**, Guilleminault C, Kao PF, Liu FY. SPECT findings in the Kleine- Levin syndrome. *Sleep*. 2005 Aug 1; 28(8): 955-60

8. **Podesta C**, Ferreras M, Mozzi M, Bassetti C, Dauvilliers Y, Billiard M. Kleine-Levin syndrome in a 14-year-old girl: CSF hypocretin-1 measurements. *Sleep Med*. 2006 Nov 10;

9. **Muratori F**, Bertini N, Masi G. Efficacy of lithium treatment in Kleine-Levin syndrome. *Eur Psychiatry*. 2002 Jul; 17(4): 232-3.