ELSEVIER

Sleep Medicine 9 (2008) 172–176

SLEEP MEDICINE

www.elsevier.com/locate/sleep

Original article

# Atypical Kleine–Levin syndrome: Can insomnia and anorexia be features too?

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Received 31 August 2006; received in revised form 23 February 2007; accepted 23 March 2007
Available online 20 July 2007

**Abstract**

*Background:* Kleine–Levin syndrome is an uncommon disorder with recurrent episodes of hypersomnia, clearly associated with behavioral abnormalities like binge eating, hypersexuality and abnormal behavior. Many patients may not necessarily fulfill minimum criteria described for diagnosis. We aim to report such patients with atypical presentation resembling the Kleine–Levin syndrome.

*Method:* We evaluated all patients at our clinic who had episodic disturbance in sleep and/or appetite lasting a few days to weeks, not necessarily fulfilling the International Classification of Sleep Disorders (ICSD) criteria for a diagnosis of Kleine–Levin syndrome, over 4 years. All clinical details, especially regarding sleep, appetite and behaviour during episodes, about prior and co-existing illnesses were noted. All patients were investigated with brain magnetic resonance imaging (MRI), electroencephalogram (EEG) and some with polysomnography.

*Results:* Eighteen patients (5 females, 13 males) ranging in age from 12 to 55 years (median 18 years) were included in the study. The median duration of symptoms was 1.5 years, and the median number of episodes in each patient was six. The range of episode length was 18–300 h with a mean of 91.2 h. Fourteen patients had a history of hypersomnia, 3 had only insomnia and 3 had both during their episodes, while 5 patients reported hyperphagia, 11 reduced appetite and 2 no change in appetite. Ictal EEG revealed evidence of sleep, while polysomnography showed reduced rapid eye movement (REM) latency and normal sleep architecture during the episode. MRI was normal in all patients, except one who showed non-specific abnormalities. All patients showed improvement with carbamazepine.

*Conclusion:* There are many patients with episodic alteration in sleep, appetite and behaviour with a course and treatment response similar to the classical Kleine–Levin syndrome, who otherwise do not fit the classical description for diagnosis of this condition.


*Keywords:* Kleine–Levin syndrome; Atypical; Anorexia; Insomnia

## 1. Introduction

First described in 1925 and formally named by Critchley and Hoffman in 1942, the Kleine–Levin syndrome has been believed to be an uncommon disorder, mainly involving circadian regulation of sleep and appetite [1–3]. According to the modified definition of the International Classification of Sleep Disorders (ICSD), the disorder is characterized by recurrent episodes of hypersomnia and binge eating (rapid consumption of a large amount of food), usually with onset in early adolescence in males but occasionally later in life and in women [4]. The ICSD-2 includes criteria for diagnosis of this syndrome under the entry for recurrent hypersomnia [5], many features of which may not be

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doi:10.1016/j.sleep.2007.03.016

fulfilled by several patients. Kleine–Levin syndrome has been described as an uncommon disorder with recurrent episodes of hypersomnia, clearly associated with behavioral abnormalities like binge eating and hypersexuality.

There is a detailed description of the demographic features, course, and associated features required for the diagnosis of this condition. However, many patients may have features different from the essential features described and may not necessarily fulfill all these criteria yet present with a similar syndrome of unknown etiology. Since no definite etiopathophysiologic basis has yet been identified for the occurrence of this disorder, no diagnostic tests are available. Hence, it might be important to include patients with a wider spectrum of clinical manifestations essentially including episodicity and disturbance of sleep and/or appetite in order to avoid underdiagnosis. In this study, we aim to report some atypical patients presenting with episodic disturbance of sleep and/or appetite that resembles Kleine–Levin syndrome as described in the ICSD but does not necessarily fulfill the essential features and criteria proposed.

## 2. Material and methods

We included and prospectively followed up all patients presenting to the Sleep Disorders Clinic, Department of Neurology, All India Institute of Medical Sciences, New Delhi, India between January 2001 and March 2005 with episodic disturbance in sleep and/or appetite with or without behavioral abnormalities, but not fulfilling all minimal criteria proposed by the ICSD. Emphasis was laid on the exclusion of patients with psychiatric disorders such as depression or other sleep disorders such as narcolepsy or sleep-disordered breathing. Patients with a history of epilepsy and those in altered levels of consciousness in previous or current episodes (based on history of arousability and urinary and fecal continence) were excluded. Patients with current history of any medical or neurological co-morbid conditions were also excluded.

All patients received thorough clinical evaluation and then underwent routine hemogram and serum biochemical tests, including fasting and postprandial blood glucose estimation, liver and renal function tests and thyroid profile to rule out any metabolic abnormalities which may contribute to fluctuations in alertness. Magnetic resonance imaging (MRI), electroencephalogram (EEG) and single photon emission computerized tomography (SPECT) were obtained to look for any structural or functional, focal or diffuse abnormalities. Other investigations were obtained based on differences in clinical presentations of different patients. Polysomnographic studies were performed whenever possible during the episodes, and during the interepisode periods.

## 3. Results

Eighteen patients (5 females, 13 males) ranging in age from 12 to 55 years (median 18 years) were included in the study. The median duration of symptoms was 1.5 years with the median number of episodes in each patient being six. The range of episode length was 18–300 h with a mean of 91.2 h.

### 3.1. Characteristics of episodes

Hypersomnia was a feature in 14 among the 18 patients, while insomnia was a feature in 6 patients (remaining 4 patients as well as 2 patients who would have either hypersomnia or complete lack of sleep during different episodes). Hyperphagia was reported by 5 patients, reduced appetite by 11 patients, and 2 patients had no alteration in appetite. Abnormal behaviour was observed in 13 patients; however, none of the patients had hypersexuality during these episodes. This included severe anxiety in one patient with insomnia, while paranoid delusions and childish behaviour were present in another patient, and the other 9 patients had confusion and violent behaviour with use of abusive language. One patient had prominent obsessive-compulsive symptoms with compulsive handwashing and other activities during episodes, and another patient experienced hallucinations and spoke to deceased grandparents during episodes (Table 1).

All patients except patients 5 and 8 would be absolutely normal in between the episodes, with no sleep, appetite or behaviour-related abnormalities. All patients had been assessed in detail for any underlying psychiatric illness and during interictal periods, they had no morbidity at all. None of the patients with insomnia and/or anorexia during episodes had excessive sleep or appetite upon cessation of the episode or during another part of the episode. Patient 5 developed an increase in appetite and coarsening of facial features, but investigation did not reveal any cause for it; all hormonal and imaging studies were normal. Examination revealed no abnormality except mild proximal muscle weakness in patient 8 who had congenital myopathy. No patients had any abnormality in alertness, cognitive functioning or behaviour between the attacks.

#### 3.1.1. Past history

Seven of the 18 patients reported had past history of major febrile illness; viral encephalitis, Dengue fever, malaria, Kala-azar and an undiagnosed prolonged febrile illness were reported in one patient each, while two patients had history of tubercular meningitis. All patients had these febrile illnesses more than at least 1 year prior to onset of current symptoms.