

# Atypical Kleine–Levin syndrome: An elusive entity?

Swarndeeep Singh<sup>1</sup>, Saurabh Kumar<sup>1</sup>, Rohit Verma<sup>1</sup>, Nand Kumar<sup>1</sup>

<sup>1</sup>Department of Psychiatry, All India Institute of Medical Sciences, New Delhi, India

### ABSTRACT

Kleine–Levin syndrome (KLS) is a rare disorder (around 1.5 cases per million population), often presenting with triad of recurrent episodes of hypersomnia, hyperphagia, and hypersexuality. However, cases of atypical KLS with features opposite to that being commonly reported are often misdiagnosed as psychosis and present as a diagnostic challenge for the physicians, psychiatrists, and neurologists. We describe a case of atypical KLS which was misdiagnosed as unspecified nonorganic psychosis previously, highlighting the various points which would be helpful in identifying and diagnosing cases of atypical KLS in future.

**Keywords:** Atypical, India, International Classification of Sleep Disorders, Kleine–Levin syndrome, psychosis

### Introduction

Kleine–Levin syndrome (KLS) is a rare disorder (around 1.5 cases per million population), named after Willi Kleine and Max Levin who studied and reported the association of periodic somnolence with morbid hunger from 1925 to 1936.<sup>[1]</sup> This was followed by multiple case reports which erroneously lead to the popular triad of hypersomnolence, hyperphagia, and hypersexuality being suggested as the core symptoms of KLS.<sup>[1]</sup> Sleep problems are found in up to 50% of children presenting to primary care physicians and about 4% of children have formal sleep disorder.<sup>[2,3]</sup> Hence, it is important for primary care physicians to be familiar with sleep disorders seen in children and adolescents. We describe a case of atypical KLS which was misdiagnosed as unspecified nonorganic psychosis, highlighting the importance of recognizing atypical features of KLS in making an early and accurate diagnosis.

### Case Report

Mr. X, a 18-year-old right-handed single male educated up to 12<sup>th</sup> standard presented to our psychiatric outpatient clinic in

September 2015 with complaints of excessive sleep episodes of abrupt onset lasting for 7–14 days occurring 1–2 times every year from the last 5 years (onset at 13 years of age) associated with feeling of depersonalization and derealization with abnormal behavior. He would experience sudden-onset extreme fatigability with an irresistible need to rest, sleeping for 22–23 h/day with spontaneous waking for 2–3 times to pass urine or stools. He would have to be coaxed to eat once or twice in the day and would report loss of appetite when awakened by family members. He also reported feeling of depersonalization and derealization when awake during this period as if in a dream-like state and felt as if he was dead or in a movie with things around him being unreal. He would be apathetic and stopped doing his routine activities. During the episodes, there was no history of hyperphagia, hypersexuality, and prominent mood or anxiety symptoms. There was no positive past or family history of any psychiatric or neurological disorder.

In the first episode, the patient also had, in addition to above-described symptoms, sudden-onset persecutory delusion and disorganized behavior lasting for 2–3 days. He was diagnosed as a case of unspecified nonorganic psychosis (according to the International Classification of Diseases [ICD-10]) and had received multiple antipsychotics (olanzapine, aripiprazole,

**Address for correspondence:** Dr. Saurabh Kumar, Department of Psychiatry, Fourth Floor, Teaching Block, All India Institute of Medical Sciences, Ansari Nagar, New Delhi - 110 029, NCT-Delhi, India.

E-mail: saurabhksingh.singh@gmail.com

#### Access this article online

##### Quick Response Code:



**Website:**  
www.jfmpc.com

**DOI:**  
10.4103/2249-4863.214963

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

**For reprints contact:** reprints@medknow.com

**How to cite this article:** Singh S, Kumar S, Verma R, Kumar N. Atypical Kleine–Levin syndrome: An elusive entity? J Family Med Prim Care 2017;6:161-3.

trifluoperazine) with episodes occurring despite of patient being compliant to antipsychotic treatment.

The patient was admitted in psychiatry ward for detailed evaluation to rule out any neurological or sleep disorder causing the present condition. Routine (liver functions, renal functions, hemogram, fasting blood sugar, urine routine and microscopy, chest-X-ray, electrocardiogram) and other laboratory investigations (serum Vitamin B-12 and folic acid, serum adenosine deaminase, serum ferritin, blood ammonia) did not detect any abnormality. Endocrine evaluation (serum thyroid-stimulating hormone, T3, T4, thyroid peroxidase antibody, serum testosterone, serum adrenocorticotrophic hormone, serum prolactin) was within normal limit. Electroencephalography was normal with no epileptiform discharges and magnetic resonance imaging of the brain did not reveal any structural abnormality. Polysomnography was also done to rule out any sleep-related disorder and did not detect any abnormality. Technetium-99m ethyl cysteinate dimer brain perfusion study (single-photon emission computed tomography) revealed hypoperfusion in left frontal (precentral gyrus), left temporoparietal, left basal ganglia, and left thalamus consistent with findings reported in KLS patients.<sup>[4]</sup>

Our case fulfills the criteria for KLS given in the ICD-3 (2013) guideline.<sup>[5]</sup> Antipsychotics were stopped and the patient continues to maintain symptom-free on conservative management and is in active follow-up.

## Discussion

This was an atypical presentation of KLS with no history of hyperphagia or hypersexuality (in any of the episodes), but rather a loss of appetite and decreased interest in sexual activity associated with hypersomnolence episodes. Smolik and Roth have defined “atypical KLS” as a form of recurrent disorder, in which hypersomnia, hyperphagia, and/or hypersexuality are replaced by its or their opposite(s) of insomnia, anorexia, and hyposexuality.<sup>[6]</sup>

In our review of literature, we found two large systematic studies comprising 108 and 120 KLS patients, mostly from western countries. The first study reported the frequency of decreased appetite and decreased sexuality during at least one episode of KLS in 108 patients as 36% and 6%, respectively.<sup>[7]</sup> Further, a continuously altered dream-like perception with or without feeling of derealization was found to be highly sensitive (100%) KLS symptom,<sup>[8]</sup> possibly missed in many previous case reports,<sup>[9]</sup> was suggested to be used in further studies to increase the sensitivity and specificity of diagnosing KLS cases, especially with atypical presentations (like in our case). The second study describing the frequency of symptoms reported among 120 KLS patients supported the findings of the previous study with decreased appetite and decreased sexuality reported in around 39% and

9% of cases, respectively.<sup>[8]</sup> This suggests that atypical features are seen in around one-third of KLS cases, making atypical KLS a fairly common entity. A case series of 18 consecutive patients presenting at a tertiary care center from India, with episodic sleep disturbances, appetite, and behavioral changes, reported that four had both insomnia and hypersomnia while two had only insomnia, 11 reported reduced appetite, and none reported hypersexuality during any of the episodes.<sup>[10]</sup> This study concluded that many patients with episodic alteration of sleep, appetite, and behavior, with a course and treatment response similar to classical KLS, do not fit with the classical description and should be classified as atypical KLS. Further, patients of KLS with hypersomnolence and myriad of possible associated symptoms as discussed above may present to primary care physicians as the first point of contact for the medical treatment of sleep disorder. Increased awareness among primary care physicians will possibly lead to an early identification and appropriate management of KLS.

To the best of our knowledge, this is the first case report of atypical KLS from India, which describes features of decreased appetite and sexual activity in the absence of commonly reported symptoms of hyperphagia or hypersexuality. The present report aims to increase the awareness among clinicians (psychiatrists, neurologists, and physicians) about the atypical presentations of KLS and highlights the various points which would be helpful in identifying and diagnosing cases of KLS in future.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

## References

1. Arnulf I, Rico TJ, Mignot E. Diagnosis, disease course, and management of patients with Kleine-Levin syndrome. *Lancet Neurol* 2012;11:918-28.
2. Liu X, Liu L, Owens JA, Kaplan DL. Sleep patterns and sleep problems among schoolchildren in the United States and China. *Pediatrics* 2005;115 1 Suppl: 241-9.
3. Meltzer LJ, Johnson C, Crosette J, Ramos M, Mindell JA. Prevalence of diagnosed sleep disorders in pediatric primary care practices. *Pediatrics* 2010;125:e1410-8.
4. American Academy of Sleep Medicine. Diagnostic and Coding Manual, International Classification of Sleep Disorders. 3<sup>rd</sup> ed. Westchester, Illinois: American Academy of Sleep Medicine; 2013.
5. Huang YS, Guilleminault C, Kao PF, Liu FY. SPECT findings in the Kleine-Levin syndrome. *Sleep* 2005;28:955-60.
6. Smolik P, Roth B. Kleine-Levin syndrome: Etiopathogenesis and treatment. *Acta Univ Carol Med Monogr* 1988;128:5-94.
7. Arnulf I, Lin L, Gadoth N, File J, Lecendreux M, Franco P, *et al.* Kleine-Levin syndrome: A systematic study of 108 patients. *Ann Neurol* 2008;63:482-93.

8. Lavault S, Golmard JL, Groos E, Brion A, Dauvilliers Y, Lecendreux M, *et al.* Kleine-Levin syndrome in 120 patients: Differential diagnosis and long episodes. *Ann Neurol* 2015;77:529-40.
9. Arnulf I, Zeitzer JM, File J, Farber N, Mignot E. Kleine-Levin syndrome: A systematic review of 186 cases in the literature. *Brain* 2005;128(Pt 12):2763-76.
10. Shukla G, Bhatia M, Singh S, Goyal V, Srivastava T, Behari M. Atypical Kleine-Levin syndrome: Can insomnia and anorexia be features too? *Sleep Med* 2008;9:172-6.