Case Report

The Annals of Family Medicine

Unpredictable Kleine-Levin syndrome: A mysterious phenomenon?

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Received Date : June 04, 2024
Accepted Date : June 06, 2024
Published Date : July 04, 2024

ABSTRACT

With only 1.5 instances per million people, Kleine-Levin syndrome (KLS) is an uncommon condition that frequently manifests as a triad of repeated bouts of hypersomnia, hyperphagia, and hypersexuality. Nonetheless, instances of atypical KLS with characteristics dissimilar to those frequently documented are frequently mislabeled as psychosis and provide a diagnostic difficulty for medical professionals, psychiatrists, and neurologists. We present a case of atypical KLS that was previously misdiagnosed as undefined nonorganic psychosis, emphasizing the several aspects that could be useful in recognizing and classifying atypical KLS cases in the future.

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

INTRODUCTION

Kleine-Levin syndrome (KLS) is an uncommon condition with approximately 1.5 instances per million people. It is named for Willi Kleine and Max Levin, who conducted research and reported on the connection between morbid hunger and periodic somnolence between 1925 and 1936.[1]

Subsequently, there were several case reports that mistakenly suggested hypersomnolence, hyperphagia, and hypersexuality as the primary symptoms of KLS.[1]

Up to 50% of children who visit primary care doctors have sleep issues, and 4% of them have a documented sleep disorder.[2,3] Therefore, it is critical that primary care physicians understand the various types of sleep problems that affect children and adolescents. We present a case of atypical KLS that was mistakenly identified as nonorganic psychosis without a clear cause, emphasizing how crucial it is to unusual KLS characteristics in achieving a prompt and precise diagnosis.

CASE STUDY

Mr. X, an 18-year-old single male who is right-handed and educated up to the 12th standard, came to our psychiatric outpatient clinic in September 2015 complaining of abruptly starting excessive sleep episodes that lasted 7–14 days and happened 1-2 times a year for the previous 5 years (beginning at age 13). These episodes were accompanied by feelings of depersonalization and derealization as well as abnormal behavior. He would become extremely fatigued suddenly, needing to sleep for 22–23 hours a day and waking up 2-3 times on his own to go to the bathroom or urinate. He would complain of being hungry when family members woke him up and would need to be persuaded to eat once or twice a day. He also mentioned experiencing depersonalization and derealization while conscious throughout During this time, he appeared to be in a dreamlike condition and felt as though he was dead or in a movie where nothing was real. He would become indifferent and cease performing his daily tasks.

There was no prior history of hyperphagia, hypersexuality, or severe depressive or anxious symptoms at the time of the episodes. There was no positive family or personal history of any neurological or psychiatric illness.

Along with the symptoms listed above, the patient experienced a sudden onset persecutory hallucination and chaotic conduct that persisted for two to three days during the first episode. The International Classification of Diseases [ICD-10] classified his diagnosis as unexplained nonorganic psychosis, and he was prescribed many antipsychotics, including olanzapine and aripiprazole.

After being admitted to the psychiatry ward, the patient underwent a thorough evaluation to rule out any neurological or sleep disorders that might be causing their current symptoms. Laboratory tests (serum vitamin B-12 and folic acid, serum adenosine deaminase, serum ferritin, blood ammonia) and routine tests (liver functions, renal functions, hemogram, fasting blood sugar, urine routine and microscopy, chest X-ray, electrocardiogram) did not find any abnormalities.

Serum thyroid-stimulating hormone, T3, T4, thyroid peroxidase antibody, serum testosterone, serum adrenocorticotropic hormone, and serum prolactin were all within normal ranges according to the endocrine evaluation.

Neurological abnormalities were not detected by magnetic resonance imaging, and electroencephalography showed no epileptiform discharges. Additionally, polysomnography was performed to rule out any disorders connected to sleep, and it revealed no abnormalities.

Single-photon emission tomography brain perfusion research using technetium-99m ethyl cysteinate In line with findings from KLS patients, a technetium-99m ethyl cysteinate dimer brain perfusion study using single-photon emission computed tomography indicated hypoperfusion in the left frontal (precentral gyrus), left temporoparietal, left basal ganglia, and left thalamus.[4] Our case satisfies the KLS requirements listed in the ICD-3 (2013) guideline.[5] Antipsychotics were discontinued, and the patient is actively being monitored and maintaining symptom-free status with conservative treatment.

DISCUSSION

This was an uncommon presentation of KLS, with no history of hypersexuality or hyperphagia (in any of the episodes), but rather a decline in interest in sexual activity and lack of appetite linked to periods of hypersomnolence. According to Smolik and Roth, "atypical KLS" is a type of recurrent illness in which anorexia, hyposexuality, and/or sleeplessness are substituted for hypersomnia, hyperphagia, and/or hypersexuality.[6] We discovered two sizable systematic investigations in our literature analysis, with 108 and 120 KLS patients, the most of whom were from western nations. According to the first study, 36% and 6%, respectively, of the 108 patients experienced decreased appetite and decreased sexuality during at least one episode of KLS.[7] Moreover, a persistently changed dream-like impression, with or without emotion derealization was discovered to be a very sensitive (100%) KLS symptom,[8] maybe overlooked in numerous prior case reports,[9] and was recommended to be employed in future research to improve the sensitivity and specificity of detecting KLS cases, particularly those with unusual presentations (like our case). Reduced appetite and decreased sexuality were reported in about 39% of the 120 KLS patients in the second research, which detailed the frequency of symptoms among them. These findings were consistent with the prior study's findings.

To the best of our knowledge, this is the first case report of atypical KLS from India that details characteristics such as reduced sexual activity and hunger while hyperphagia or hypersexuality are not usually reported symptoms.

The goal of this paper is to raise clinicians' (physicians, neurologists, and psychiatrists) knowledge of the atypical presentations of KLS. It also identifies and highlights some key factors that may be useful in the future when diagnosing and treating KLS cases.

Conflicts of interest

There are no conflicts of interest.

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