

Kleine-Levin Syndrome: The Importance of Accurate Diagnosis and Treatment

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ABSTRACT

Kleine-Levin Syndrome (KLS) is a rare sleep disorder of unknown etiology that most commonly affects adolescent males and is characterized by relapsing-remitting episodes of hypersomnia with associated symptoms of hyperphagia, irritability, hypersexuality, lethargy, disorientation, and/or hallucinations that are often cyclical in nature and may persist for days to weeks. KLS often goes unnoticed and is therefore frequently misdiagnosed as another psychiatric condition or medical disorder. This highlights the importance of reporting KLS cases including detailed information regarding diagnostic workup, symptomatology, and treatment. The following is a case report of a 14-year-old male who presented with a two-year history of hypersomniac episodes occurring every 4 weeks to 6 weeks and lasting for about 7 days to 10 days. His family reported that he would be disoriented, lethargic, and confused during these episodes and had no recollection of what occurred. The patient was started on 300 mg of lithium twice daily, and he responded very well with no adverse effects. The patient's family reported that he has not experienced any hypersomniac episodes for over six months and has exhibited marked improvement in his mood.

This case underscores the importance of conducting a meticulous diagnostic workup to accurately identify KLS and provide appropriate therapeutic treatment to restore the patient's quality of life.

KEYWORDS

Kleine-Levin Syndrome; Hypersomnia; Case report

INTRODUCTION

Kleine-Levin Syndrome (KLS) is a rare sleep disorder of unknown etiology that most commonly affects adolescent males and is characterized by relapsing-remitting episodes of hypersomnia associated with psychiatric, cognitive, and behavioral changes [1,2]. Symptoms may include hyperphagia, hypersexuality, irritability, lack of

energy, lack of emotions, disorientation, and/or hallucinations [1,2]. These symptoms are often cyclical in nature and may persist for days to weeks. The precise prevalence of KLS is unknown because it often goes unnoticed and therefore remains undiagnosed [1]. An important corollary of this is that KLS is frequently misdiagnosed as another psychiatric condition or medical disorder. This highlights the importance of reporting

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KLS cases including detailed information regarding diagnostic workup, symptomatology, and treatment. The following case describes a patient who was diagnosed with KLS and responded positively to medical therapy with lithium.

CASE REPORT

A 14-years old male presented with a two-year history of hypersomniac episodes occurring every 4 weeks to 6 weeks and lasting for about 7 days to 10 days. His family reported that he had to be woken up to eat, shower, and use the restroom. During this time, he would be disoriented, lethargic, and confused. The patient stated that he has no recollection of what transpires during these episodes. In between episodes, he is responsive to directions and questions. He is also able to complete his academic work and activities of daily living with reminders. The patient also has poor awareness of interpersonal boundaries, frequently makes inappropriate comments, and behaves impulsively which have all negatively impacted his academic progress in school and his relationships with peers.

DISCUSSION

The patient underwent a thorough diagnostic workup including brain imaging, neuropsychological testing, and neurological evaluation. CT and MRI of the brain yielded normal results. Neuropsychological testing showed multiple abnormalities. The results revealed an IQ in the borderline impaired range, low average working memory, borderline verbal comprehension, and borderline visual perceptual reasoning. Additionally, the testing demonstrated impairment in attention, executive functioning, and processing speed. Results of the pediatric polysomnogram demonstrated mild obstructive sleep apnea and sleep-related hypoventilation with considerably elevated end-tidal CO₂ levels. While EEG did not reveal any abnormal patterns, consultation with a pediatric neurologist was conducted to rule out complex partial seizures. Ultimately, the patient was diagnosed

with KLS based on his history, presenting symptoms, and exclusion of other possible psychiatric or medical conditions. His recurrent episodes of hypersomnia with associated symptoms of cognitive impairment, mood instability, and lack of recollection of events that occurred during these episodes all support a diagnosis of KLS. Consultation with the pediatric neurologist further confirmed this diagnosis.

Prior to being diagnosed with KLS, the patient's treating physician attributed his symptoms to bipolar disorder and ADHD. Subsequently, he was placed on a combination of Adderall, Effexor, and Lamictal with no improvement in his symptoms. After weaning him off of these medications over a period of three months, we started the patient on 300 mg of lithium twice daily, and he responded very well with no adverse effects. In the medical literature, lithium therapy for KLS has been reported to reduce the duration and frequency of hypersomniac episodes as well as abnormal behavior during these episodes [3]. The patient's family reported that he has not experienced any hypersomniac episodes for over six months and has exhibited a significant decrease in impulsive behavior. Additionally, he has shown marked improvement in his academic work and interpersonal relationships with peers. Thyroid function tests conducted two months following initiation of lithium therapy showed an increase in TSH levels with normal free T3 and free T4, supporting a diagnosis of lithium-induced subclinical hypothyroidism. An endocrinologist was consulted, and a recommendation was made to closely monitor TSH, free T3, and free T4 levels every three months.

CONCLUSION

This case clearly underscores the criticality of properly diagnosing, evaluating, and treating KLS particularly in the setting of comorbid conditions that can complicate the clinical presentation of symptoms [4]. It is our hope that this case report elucidates the significance of

conducting a meticulous diagnostic workup to accurately identify KLS and subsequently providing appropriate therapeutic treatment to restore the patient's quality of life.

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