Manipal Journal of Medical Sciences

Volume 7 | Issue 1 Article 8

9-20-2022

A rare cause of recurrent hypersomnia- Klein-Levin syndrome

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CASE REPORT

A rare cause of recurrent hypersomnia- Klein-Levin syndrome

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Abstract

Recurrent episodes of severe hypersomnia coupled with cognitive and behavioral abnormalities like disorientation, apathy, derealization, compulsive eating and hypersexuality are hallmarks of unusual sleep disease known as Klein-Levin syndrome. We hereby discuss a 46-year-old gentleman who came with recurrent episode of loss of muscle tone, was diagnosed with Klein-Levin syndrome and responded well to medications.

Keywords: Klein-Levin syndrome, Hypersomnia

Introduction

Recurrent episodes of severe hypersomnia coupled with cognitive and behavioral abnormalities like disorientation, apathy, derealization, compulsive eating and hypersexuality are hallmarks of unusual sleep disease known as Klein-Levin syndrome (KLS). The symptoms last for few weeks followed by a gap of few weeks or months [1]. We hereby report a patient who presented with recurrent episodes of loss of tone of body, which turned out to be KLS.

Case Report

A 46-year-old gentleman presented to OPD with history of recurrent episodes of sudden loss of tone in his muscle over past one week. The episodes happen multiple times in a day. This happens even while driving, when he noticed that he is unaware of when he changed the lane. During his weekend holidays he would often sleep up to 18 hours in a day. Patient says he doesn't have control over his dietary habits and frequently goes for binge eating. He has recurrent and intense sexual urges. He has headaches on and off, however no weakness of arms or legs. He denies having hallucination (including hypnagogic and hypnopompic) or slurring of speech. He had similar episode before for which he was evaluated by a neurologist. His EEG and polysomnography tests were normal except for increased total sleep time. The patient has irritable bowel syndrome and he is on probiotics for the same. He has no history of diabetes, hypertension, hyperlipidemia or hypothyroidism. He is not on any chronic medications and denies any psychiatric disorder. On examination, he is obese with Body Mass Index of 40 kg/m2. The blood pressure, pulse and saturation were normal. His neurological examination was normal. The cognitive function assessed using mini-mental status examination and was normal. Examination of cardiovascular, respiratory and abdomen was unremarkable. His blood tests including complete blood count, renal function test, liver function tests, electrolytes, ammonia levels and thyroid function tests were normal and urine toxicology screen was negative. Based on history, clinical examination and analysis of laboratory, a diagnosis of KLS was made and he was started on modafinil 100 mg once daily. He was advised to sleep in safe environment and advised to refrain from driving. He had a good response to modafinil and he noticed a significant improvement in his symptoms. During follow up

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Manuscript received: 22 January 2022
Revision accepted: 20 September 2022

How to cite this article: Khalid M, Karim T, Anwar W. A rare cause of recurrent hypersomnia- Klein-Levin syndrome. *MJMS.* 2022; 7(1): 13-14

he has shown remarkable improvement and plan is to slowly reduce and stop modafinil as most patient show spontaneous resolution.

Discussion

KLS is a rare condition whose estimated prevalence ranges from one to five cases per million population [1]. Although psychological basis was considered most likely when disorder was first described, other hypothesis like birth difficulties, genetic and immune mediated causes were considered [2]. The median age of onset of KLS is 16 years, with age range between 4-82 years. Milder cases have one to three short episodes per year, but moderate cases have monthly episodes of 7 to 10 days each [3, 4]. Hypersomnia is one of the cardinal symptoms and is mandatory for diagnosis [5]. The second most common presentation is cognitive and memory disturbance [4]. Sensation of unreality is common during the episodes and are probably most specific symptom of the syndrome [4]. Other symptoms include apathy and disinhibition like hyperphagia and hyper sexuality [4]. CSF analysis is normal during and between episodes [5, 6]. EEG, during an episode may be useful if aspects of history are suggestive of seizures and is supportive of KLS if there is focal or global EEG slowing [7]. Polysomnography when performed during episodes shows increased total sleep time, otherwise nonspecific. Structural brain computed tomography (CT) and magnetic resonance imaging (MRI) studies are normal in patients with KLS [7]. Treatment includes supportive care during episodes like educating family members and caregivers, to prevent the patient getting injured during periods of hypersomnia [8]. Severe anxiety and psychosis should be treated symptomatically [8]. Stimulants like methylphenidate and non-amphetamine wakefulness promoting agents like modafinil and amantadine have modest benefits [2]. Prolonged episodes of disabling symptoms can be considered high dose glucocorticoids as per experience by Lavault et al. [9].

Conclusion

Our patient had clinical features, which fit into the diagnostic criteria as per the International classification of sleep disorders (ICSD-3) [10]. He responded well to modafinil. The differential diagnosis of KLS include a variety of psychiatric, neurologic and sleep disorders, most of which are much more common than KLS.

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