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Kleine Levine syndrome (Sleeping beauty): A case report from Saudi Arabia

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Background

Kleine – Levin syndrome (KLS), also called "Sleeping beauty syndrome" is a rare sleep disorder characterized by recurrent episodes of hypersomnia, hyperphagia, cognitive disturbances, and hypersexuality. The underlying pathophysiology remains unknown. Kleine-Levin syndrome is often misdiagnosed as depression, bipolar disorder, psychosis, seizures, or intoxication. The disorder is at the interface of neurology and psychiatry. Attacks decrease in frequency and eventually cease in most cases of adolescent onset. we are reporting the first case of KLS from Saudi Arabia, a 14-year-old boy, with delay in diagnosing KLS.

CASE REPORT

14 – year – old boy was referred from other hospital as a case of recurrent hypersomnia with abnormal behavior of 2-year duration, for further management. His history started 2 years prior to presentation with fever which was treated empirically, after that started to have excessive sleepiness up to 20 hours per day, with difficulty to awake him from sleeping, initially there was no medical intervention until he became unable to communicate or concentrate. He lost his interest and started to have inappropriate words, hypersexual desire, in addition to throbbing headache over the left temporal area. During awake state, he would appear apathetic with minimal interaction with others. His speech would remain slurred with low tone and volume or sometimes even not understandable. The patient was irritable and aggressive whenever prevented from sleep. No other neurological symptoms. In between the attacks, he was completely normal. The patient was admitted 2 times to other hospital and was diagnosed as epilepsy. He received carbamazepine and valproate for 2 years without significant improvement. At physical examination, the patient was looking well, normal growth parameters and no dysmorphic features or any other systemic abnormality.

Hospital course: The patient was admitted initially to epilepsy and monitoring unit. EEG monitoring showed no epileptiform discharge. His Neuroimagins were unremarkable. The pediatric pulmonary team were consulted and sleep study with sleep multiple latencies (MSLT) were requested. The result of the sleep study showed total recording time of 522.2 minute, total sleep time 513.5 minute, sleep efficiency 98.3%. No evidence of epileptiform activity was noted. No significant sleep pathology noted.

The MSLT reported severely reduced sleep latency confirming pathogenic sleepiness. This MSLT result contributed to polysmnography diagnosis of narcolepsy, yet the patient clinical presentation of Kleine Levine syndrome maybe responsible for the diagnosis. The diagnosis of KLS was made as per diagnostic criteria of International Classification of Sleep Disorders (1) after ruling out other possible causes of hypersomnia. His HLA typing of DQB1*1 came positive for DQB1*02, DQB1*03 which has mentioned in the literature as possible association with the disease

Discussion

Brierre de Bosmont in 1862 was perhaps the first reported case of KLS [(1)]. The condition took the name from Willi Kleine who, in 1925, explained a series of cases of periodic hypersomnia and also from Max Levin who in 1930, described a case of periodic hypersomnia and hyperphagia [(1)]. Kleine -Levin syndrome has mild, moderate and severe form. Hyperphagia and hypersexuality are often cited as characteristic symptoms of Kleine -levin syndrome and were viewed as mandatory diagnostic criteria before the 2005 international guideline were published [(1)]. Isabelle et al reported hyperphagia in only 71 (66%) of 108 patients and hypersexuality in 57 (53%). This symptom of hyper sexuality affects boys more than girls and frequently manifests as substantially increased masturbation or demands on sexual partners. In our case, he presented with hypersexuality and no hyperphagia, actually he was having poor appetite. Apathy affects all patients including our case. Adolescent often stop normal activities such as washing and styling their hair [(2)]. Naresh et al, in a recent comprehensive review mentioned 36 cases with KLS reported from India, 8 of them were females [(3)]. The first episode was preceded with fever in 50% of cases, as in our index case, while in others with postpartum psychosis, familial conflict and stress. The triad of hypersomnia, hyperphagia and hypersexuality is not mandatory for the diagnosis of KLS as it is present in only 45% patients. Hypersomnia is kept as an obligatory criterion. Our patient presented with hypersomnia. Many studies of physiopathology for KLS showed hypothalamic dysfunction, which would justify the alterations of the sleep regulatory system, sexual behavior and increased appetite. In favor of post- infectious autoimmunity as a cause of this disorder, a European group recently identified the human leucocyte antigen (HLA) subtype DQB1*02 as possibly being associated with the disease [(4,5)]. In our case we were able to detect this (HLA) subtype DQB1*02.

Conclusion

KLS is a rare sleep disorder with a lot of psychiatric symptoms that can be wrongly diagnosed as neurological or psychiatric illness. The framework of core symptoms of hypersomnia, slowed cognitive functions, apathy and derealization should supersede the previously recognized hypersomnia-hyperphagia- hypersexuality triad. Hence, it is important for clinicians to have high index of suspicion on such atypical presentations. There is no definitive treatment for KLS, however some cases showed clinical improvement with some medications. The prognosis of KLS is considered to be good with spontaneous remission in most of the cases in later years.

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