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Case Report

Kleine-Levin Syndrome and Its Successful Treatment with Armodafinil

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Abstract

Kleine-Levin syndrome is a rare disease of uncertain etiology characterized by recurrent episodes of hypersomnia and to various degrees, behavioral or cognitive disturbances, compulsive eating behavior and hyper sexuality. There is no definitive treatment for Kleine-Levin syndrome. We are reporting a case of Kleine-Levin syndrome from India and its successful treatment with armodafanil (German J Psychiatry 2014; 17 (1): 25-26).

Keywords: Kleine-Levin syndrome, hypersomnia, hypersexuality, armodafanil

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Introduction

Kleine–Levin syndrome (KLS) or "sleeping beauty syndrome" is a rare disease of uncertain etiology, characterized by recurrent episodes of hypersomnia and to various degrees, behavioral or cognitive disturbances, compulsive eating behavior and hypersexuality (Hauri, 2005). The disease predominantly affects adolescent males. The diagnosis of KLS is very difficult since there are no symptoms that allow for a positive diagnosis; KLS is instead a diagnosis of exclusion. The diagnosis is entirely clinical. According to the International Classification of Sleep Disorders (Hauri, 2005), it belongs to the category of recurrent hypersomnia, defined as episodes of excessive sleepiness lasting more than 2 days and less than 4 weeks, intermixed with long intervals of normal alertness lasting usually months to years, recurring at least every year, and not better explained by a sleep disorder, a neurologic disorder, a mental disorder or the use of drugs. The essential clinical criterion of KLS is recurrent episodes of hypersomnia. Moreover, patients have to experience at least one of these symptoms only during the episodes: (1) cognitive or mood disturbances (confusion, irritability, mutism, aggressiveness, derealization, hallucinations, and delusion) (2) megaphagia with compulsive eating; (3) hypersexuality with inappropriate or odd behavior; and (4) abnormal behavior such as irritability, aggression, and odd behavior. This syndrome has a benign clinical course, with spontaneous disappearance of symptoms.

Patients with KLS are often mistakenly diagnosed with a psychiatric disorder. The periods of somnolence, hyperphagia, and withdrawal can mimic severe depression, and some people experience a brief period of high energy following these episodes which looks like a manic episode, so that some patients are incorrectly diagnosed with bipolar disorder. Narcolepsy, Klüver-Bucy syndrome, and temporal lobe epilepsy (which was ruled out here by EEG) can also produce similar symptom profiles.

From India, few cases of Kleine-Levin syndrome have been reported (Prabhakaran et al., 1970; Shukla et al., 1980; Sagar et al., 1990; Malhotra et al., 1997; Saha, 2008). We are reporting a case of Kleine-Levin syndrome and its successful treatment with armodafanil.

Case History

Mr. A, a 15-year-old adolescent Hindu male with uneventful birth and developmental history without past and family history of neurological and psychiatric illness presented an episodic illness of 2 years duration with complaints of excessive sleep, increased appetite and excessive irritability. Each episode of illness lasting for 10–15 days at the interval of 3–4 months. During each episode, he started sleeping more than the usual with average sleeping time of 17–18 hours a day. It was difficult to arouse him while he was sleeping. On waking up, he was generally irritable, would prefer to go right back to sleep, and would not like to talk to anyone. During these episodes his eating pattern also changed. During his waking hours, he was noted to have a voracious appetite. In addition, he would be seen rubbing his penis against the pillow, ignoring the presence of others. He frequently got angry and lost temper when his parents prohibited him from doing so. His self-care also decreased and he was forced to take baths and change clothes. He also did not go to his school during these episodes. These features resolved of their own after 10- 15 days. However, it would recur after approximately 3 to 4 months in a year with similar symptomatology but in between this period, he would behave appropriately with his usual interest in his studies and attending to school. Detailed medical evaluation including neurological examination was done, which revealed no significant findings. Radiological investigations were normal, EEG was normal study and all other detailed reports too were not significant. He was diagnosed to be suffering from Kleine-Levin syndrome as per diagnostic criteria of the International Classification of Sleep Disorders (Hauri, 2005). The patient was started on armodafinil 50mg per day, which was increased to 150mg from day two on. With this continuing treatment, the patient's severity and duration of each episode gradually decreased. After 8 months of treatment, there was no recurrence of these episodes, and currently the patient is under regular follow-up.

Discussion

Our patient satisfied the essential criteria for diagnosis of Kleine-Levin syndrome, i.e., hypersomnia along with megaphagia, hypersexuality and behavioral disturbances. Other behavioral disturbances as noted in this syndrome, like visual and auditory hallucinations, suicidal tendencies, motor retardation, pathological guilt, vivid imagery features reported elsewhere, were not reported during or between attacks of hypersomnolence in our patient. Precipitating factors such as hot weather, stress, head trauma, post flu and encephalitis associated with this syndrome but none were clearly recognized in our case.

There is no definitive treatment for Kleine-Levin syndrome during episodes as well as interepisodic periods. Lithium had been reported to be useful but is associated with long-term problems of regular serum monitoring (Oliveira et al., 2009). Various stimulants including methylphenidate, D-amphetamine, methamphetamine, amphetamine, modafinil, armodafinil can be useful (Arnulf et al., 2005).

Our cases responded very well to armodafinil. As compared to lithium, which has a narrow therapeutic index and requires a regular monitoring of serum levels, armodafanil is relatively safer and does not require close monitoring. As compared to other stimulants, armodafanil was preferred in view of its low addiction potential. Armodafinil has been reported to successfully treat a patient of KLS in a previous case report from India (Mittal & Mittal, 2008), but more systemic research is warranted to study the long-term effects of this drug on the course of Kleine-Levin syndrome. In conclusion, early diagnosis, treatment with patient education remain vital for alleviating suffering, loss of schooling or work and allaying anxieties of family members, teachers and school or work colleagues.

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