Case Report

Psychosis in a Case of Kleine-Levin Syndrome: A Diagnostic Challenge

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ABSTRACT

Kleine-Levin syndrome (KLS) is a rare disorder of sleep diagnosed mainly on clinical grounds. It presents a unique diagnostic dilemma for neurologists and psychiatrists; especially due to a high risk of being diagnosed as a psychiatric condition like a mood disorder. However, there is literature available documenting the cooccurrence of psychiatric illnesses in patients diagnosed with KLS. The following case highlights the above points.

Key words: *Bipolar disorder, diagnosis, Kleine-Levin syndrome, psychosis*

INTRODUCTION

Kleine-Levin syndrome (KLS) named by Critchley and Hoffman after Klein and Levin who described the disorder first, is a sleep-related disorder, the exact cause of which is not established. However, an immunological pathology is believed to be likely leading to hypothalamic dysfunction.[1,2] This is because of the observation that symptoms are often preceded by infectious diseases like influenza.[3] There is also the association between KLS and HLA (Human Leukocyte Antigen)-DQB1*0201 which supports the autoimmune etiology.[1] Factors believed to act as precipitators of illness include infections, sleep-deprivation, trauma, alcohol consumption, among others.[3] It typically affects adolescent males and though it has a recurring-remitting course, it is generally believed to be a benign condition that has a tendency to resolve spontaneously.[3] There is also a proposed role for neurotransmitters like serotonin, dopamine, and orexin (hypocretin).[4,5] From available research, it is widely believed that a hypothalamic dysfunction is the core pathology in KLS, in view of the characteristic alteration in sleep, appetite, and sexual behavior.

KLS is an intriguing disorder which appears to have several features overlapping with psychiatric disorders, especially bipolar disorder. There are a few cases reported from India, some highlighting the occurrence of psychiatric symptoms during illness.[6,7]

CASE REPORT

A 20-year-old, right-handed, XIth standard educated male was admitted at the psychiatry ward with complaints of abnormal behaviour which had started approximately 20 days prior to admission. The complaints of the father were extreme irritability, abusive and aggressive behavior without provocation; talking irrelevantly; decreased sleep; decreased self-care; spitting excessively. On examination, he was found to be grandiose and disinhibited, and had delusions of reference and persecution predominantly against family members. He was found to be oriented to his surroundings at all times during ward stay.

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On detailed evaluation, he was found to have a history of periodic episodes of intense drowsiness and sleeping throughout the day. He would be difficult to arouse and irritable and withdrawn if aroused. These episodes had been occurring since 2 years and lasted for variable periods of 10 to 20 days and subsided spontaneously. His family sought faith-healing most of the times. The episodes were separated by symptom-free periods of approximately 2-5 months. The father could not recall several key facts like any precipitating illness prior to onset of symptoms or when the latest such episode had occurred. During these episodes, there was no evidence of mood/behavioral changes suggesting depression/anxiety or any other major mental illness. His father also did not report hyperphagia/hypersexuality in these periods. There was history of nicotine dependence, but no other significant personal and family history was obtained.

When he exhibited the presenting disturbed behavior, he was seen by a private psychiatrist who, after obtaining the above information diagnosed him as suffering from rapid-cycling bipolar disorder, and prescribed a combination of lithium and typical antipsychotic medication and advised admission. However, patient was uncooperative and noncompliant because of which he was brought to our outpatient department and then admitted in the ward.

A provisional diagnosis of brief psychotic disorder in a case of KLS was made, and patient was started on tablet olanzapine 10 mg in two divided doses which was gradually increased to 30 mg in three divided doses and later supplemented with haloperidol (20 mg in divided doses) and trihexyphenidyl (8 mg in divided doses) to achieve better behavior control.

He was subjected to several investigations to rule out an organic basis of the presenting disturbances. Routine investigations (complete blood count, liver and renal function tests, fasting blood sugar, chest X-ray and electrocardiogram) revealed only an elevated aspartate amino-transferase-138 IU/mL. Ophthalmology evaluation did not reveal any abnormality including Kayser-Fleischer ring. Cerebrospinal fluid (chemistry, microscopy, and culture), Thyroid function tests and antinuclear antibodies did not reveal any significant abnormality. Magnetic resonance imaging of the brain and electroencephalogram (EEG) were normal. Neuromedicine evaluation finally produced a clinical diagnosis of KLS with brief psychotic disorder. The patient was discharged on 23rd day after admission at which time he showed 100% improvement.

After 3 months of onset of psychotic symptoms, patient is well-maintained and symptom-free. He continues to follow-up on outpatient basis.

DISCUSSION

This case highlights the difficulties faced by psychiatrists in establishing diagnosis in some patients. Our patient was initially thought to be suffering from rapid-cycling bipolar disorder based upon the cyclical sleep disturbance. However, data from father and later from the patient himself did not reveal presence of any mood disturbances during the hypersomnolent periods. The facts that support our diagnosis include patient's gender, age of onset of symptoms (approximately 18 years), hypersomnia of cyclical nature with spontaneous remissions, and recurrences and lack of any symptoms in between episodes. The lack of hyperphagia and/or hypersexuality indicates that this was most likely an atypical form of KLS.

The patient could not be subjected to polysomnography evaluation as he was not experiencing the hypersomnolent episode at presentation. However, it might have revealed EEG changes like slowing of rhythm and reduced Rapid Eye Movement (REM) sleep latency, among others during the periods of symptoms. These changes, however, are not found in all patients with KLS and thus, cannot be diagnostic.[3,8]

The occurrence of psychotic symptoms with KLS has been recorded previously in case reports.[9] However, whether there is any association between the two is still not determined as not all patients with KLS go on to develop psychosis. The point of this report is to highlight the difficulty it presents in diagnosis. The psychiatrist, who first evaluated our patient, diagnosed him as having rapid-cycling bipolar disorder. The hypersomnolent episodes may have be taken for recurrent atypical depressive episodes, several of which were followed by what was taken as a manic episode (presenting symptoms). The issue is complicated by the well-known fact that depression which is recurrent and atypical is highly likely to be of bipolar nature. To add to this is the fact that treatment strategy for both conditions overlap significantly-lithium, stimulants, fluoxetine among others. This just adds to the confusion. Psychotic symptoms have been described during the somnolent phases, though the patient's history did not reveal so. Our patient's overt psychotic symptoms warranted a diagnosis of brief psychotic disorder.

It remains to be seen if the diagnosis of the patient holds up in the future, mainly as there is no definitive method available to us to establish it irrefutably. The development of newer techniques for diagnosis of KLS would be a boon to these rare set of patients and definitely improve their management.

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