KLEINE LEVIN SYNDROME

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Introduction

PPrimary wake disorders encompass various conditions of excessive daytime sleepiness and / or increased nightmare sleep, of unknown origin beginning most often in adolescence and of recurrent natural history. Kleine in 1925 [1] and Levin in 1936 [2] described an episodic disorder characterized by periodic hypersomnolence, excessive eating and behavioural changes. For days and weeks, the patients, mostly adolescent boys, sleep for eighteen or more hours from which they can be aroused. This awakening is long enough to eat, attend to toilet activities and is usually marked by withdrawal from social contacts and they return to bed at the first opportunity. Emotional stress and febrile illness occasionally precipitate episodes of hypersomnolence. The patient appears dull, often confused, restless and may display apathy, irritability, preference for sweets, incoherent speech, loss of sexual inhibitions, delusions, hallucinations, excitation or depression, slowness in thinking and subsequent poor memory of the episode. We report one such case.

Case Report

14 year old son of a serving soldier was referred to psychiatric OPD of a large Military Hospital by medical specialist on 15 Feb 2000 with history of fever for two days, one week ago and since then the boy was withdrawn, did not talk or play and felt like sleeping all the time. The boy complained of feeling sleepy all the time. Detailed history given by the mother elucidated that the patient had a low grade fever on 8 Feb 2000, for which he was given treatment from the MI Room and the fever subsided within two days. Following fever, the patient was noticed to be sleeping excessively, lethargic, was not studying, while earlier, he was quite studious. He used to get up to eat food that too on coaxing, hurriedly did toilet activities and again used to sleep. When he was sent to school forcibly with his elder brother, the report came that he keeps on sleeping in the class, does not read, write or play with other children. The mother also reported that although her son can be woken up from sleep, he remains drowsy, irritable, looks as if lost in thoughts and immediately goes back to sleep at the first

opportunity and starts snoring (goes to deep sleep). She also noticed that he eats a lot of sweets during this period. She also reported that her son had similar episode of hypersomnolence following a bout of febrile illness for two days in Nov 99. He had recovered completely from this episode within a period of two weeks or so. When this happened for the second time she got worried and brought the child to hospital.

General physical examination and systemic examination of the child was within normal limits. Mental status examination revealed a drowsy child who spoke in monosyllables and told that he was feeling sleepy. He was irritable, apathetic and did not cooperate during formal tests of cognitive functions. His sleep and appetite were increased. He did not have any delusions, hallucinations or depressive cognitions.

The patient was investigated and haematological, urinalysis, biochemical parameters, ECG, EEG, CECT brain and MRI brain were within normal limits. A referral was made to the neurophysician to look out for any organic basis for this condition. A repeat EEG done was also normal. Clinically, the patient was diagnosed as a case of Kleine Levin Syndrome and he was serially reviewed in the psychiatric OPD. The mother of the child was reassured, psychoeducation was given about the nature of the illness and its course and prognosis, to allay her anxiety. The child started improving gradually, he was asymptomatic by 24 Feb 2000, and he had poor memory of the episode. The patient is being reviewed periodically and he is maintaining improvement.

Discussion

Critchley [3] collected a series of cases and found the age range was from puberty to 45 years, episode varied in duration from 2 to 12 in a year. Our case was also of a 14 year old boy with two episodes within four months of 14 to 18 days duration. The hyperphagia suggested a hypothalamic disorder, but anatomical verification is lacking. Mayer G et al (1988) studied 5 subjects, 4 men aged 17-28 years and 1 woman aged 30 with Kleine Levin Syndrome during symptomatic and asymptomatic periods. MRIs of the brain were normal in all patients [4]. Laboratory investigations are usually within normal limits both during and between the attacks. Basal levels of pituitary and other hormones are usually normal. CSF may

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show mild increase in protein content but is usually within normal limits. EEG usually shows general slowing of background activity sometimes with paroxysmal bursts of generalized high voltage 4-6 Hz waves. Neuroimaging is usually normal. Because of the largely negative findings, Kleine Levin Syndrome is generally considered to be a functional disorder of limbic and/or hypothalamic structures-perhaps with internal desynchronization of different aspects of circadian rhythmicity, resulting in a combination of appetite and sleep disturbances [5]. Our case did not reveal any abnormality on EEG, CT scan and MRI brain.

A psychogenic mechanism has also been proposed, but is without any foundation. In most of the instances the disease is self-limiting and disappears in early adult life [6]. There were no perceived psychological stressors in the spheres of home, school, and family in our case and the symptoms remitted within 14-18 days on both the occasions.

Prophylactic lithium or carbamazepine has shown some promising results in decreasing the frequency of the episodes [7]. We have not put this patient on prophylactic medication, as it was only the second episode. The slight consciousness disturbance that our patient had, seemed to cause the hypersomnic state.

Several neurophysiological findings also support this hypothesis [8].

Generally, the prognosis of this disorder is good. It is yet to be seen in our case how it progresses further. Hence, the child is being followed up in psychiatric OPD so that timely psychopharmaco-therapeutic intervention can be carried out when required.

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