CASE REPORT

Electrophysiological Findings in Kleine-Levine Syndrome in a Female

Basaran Demir, M.D., Constance Moore, M.D., Marietta Belamy, M.D., Ismet Karacan, M.D.

INTRODUCTION

Recurrent hypersomnia is a periodic sleep disorder which includes the Kleine-Levine Syndrome and its incomplete variants. The Kleine-Levin syndrome is characterized by recurrent episodes of hypersomnia associated with binge eating and hypersexuality. It usually occurs in adolescent males, but may occur in middle-age females. The episodes generally last from several days to several weeks and usually occur twice a year (1). No epidemiological study of the syndrome has been reported, therefore its true incidence is unknown. Although the pathogenesis of the syndrome remains unknown, a dysfunction at the hypothalamic level has been suggested (2). Incomplete forms of the recurrent hypersomnia present with recurrent hypersomnia as the only symptom (3,4). According to The International Classification of Sleep Disorders this last group of patients are coded as "monosymptomatic type" (5).

Little data exist from all night polysomnographic and multiple sleep latency testing on Kleine-Levine patients, particularly in females. In this report we describe nocturnal polysomnographic evaluation, multiple sleep latency test results, clinical EEG, brain magnetic resonance imaging (MRI) and immunogenetic findings on a female patient who was diagnosed with recurrent hypersomnia (Kleine-Levine type).

From the Sleep Disorders Center, Baylor College of Medicine, Houston, Texas, USA.
Address reprint requests to: Basaran Demir, Department of Psychiatry, University of Hacettepe, Ankara, Turkey.
Accepted May 21, 1999.

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A. 32 year-old African-American female was initially referred to our sleep disorders center for complaint of sudden onset of excessive sleepiness. Her symptoms began about nine years prior to evaluation and three weeks following the uncomplicated vaginal delivery of her first child. The patient reported that her problems with excessive sleepiness occur every six months, generally last 2-3 weeks and disappear without any intervention. She appeared somnolent and exhausted and was unable to hold a conversation with others. She answered questions with a short "yes" or "no". The family reported that during hyporsomnolent attacks, her desire for food was unusually high. No sexual disinhibition was reported. They also reported that during the interim period of remission, she had no trouble with excessive sleepiness or food binges and was able to function normally.

The patient's family health and her general physical and neurological examinations were otherwise unremarkable. Detailed questioning about the beginning of her sleep problems gave no other clue that may signify postpartum depression.

The patient received polysomnographic recording for two nights. A multiple sleep latency test consisting of five nap opportunities was administered after night one. Additionally a clinical EEG was performed prior to the second night. Two weeks following this evaluation, her symptoms disappeared completely.

Five months later, the patient was admitted to our Sleep Center with the same complaints. She was sleepy and unable to function again. She was re-evaluated in the sleep laboratory with the same parameters recorded. We again
performed a clinical EEG prior to sleep study. This hypersomnolent episode lasted about three weeks, and recovery was spontaneous. The patient was later re-evaluated in the sleep laboratory during the asymptomatic period.

**Symptomatic Periods:**

**All Night Polysomnography Findings:** During the symptomatic periods the following findings were recorded (Table 1 and Figure 1); sleep efficiency index was between 74-85 % (normal range 91-97 %). Her sleep was disturbed by frequent arousals and mid-cycle awakenings during the night. Alpha intrusions into her EEG were noted in all sleep stages. She had no delta sleep on two out of three nights. She had a very low percentage of delta sleep (2 %) on the other night (normal range 4-14 %). Sleep onset latency was prolonged in both symptomatic periods (32-74 minutes, normal range 2-12). REM latency was also prolonged (113-400 minutes, normal range 62-96) on two out of three nights, and REM sleep percentage was low (8-14 %, normal range 17-23 %) on two nights. During evaluation nights she had abnormal EEG activity in the form of spikes and sharp waves, which were temporocentral predominant and unrelated to any abnormal behavior or activity during sleep. They were temporocentral predominant and unrelated to any abnormal behavior or activity during sleep. She did not have clinically significant sleep-related breathing disorder or any evidence for periodic limb movement disorder.

**Multiple Sleep Latency Test Findings:** During daytime naps the patient fell asleep only during the nap two. REM sleep also occurred during this nap. Sleep latency was six minutes, with a REM sleep latency of eight minutes (Table 2).

**Clinical EEG Findings:** Spikes and sharp waves were noted during one of the clinical EEG runs as well as during the night. Again the spikes were temporocentral and occurred most often on the left side of the brain. Hyperventilation and photic stimulation did not induce any important changes.

**Asymptomatic Period:**

**All Night Polysomnography Findings:** During the asymptomatic period, sleep efficiency was normal (97 %) for her age. Sleep onset latency was also within normal range (6 min). Sleep architecture measures showed normal REM latency and total REM time, slightly increased stage 2 (69 %) and decreased delta sleep (2 %).

**Other Test Results:** Complete blood count and thyroid function tests results were normal. HLA-DR typing was performed using the lymphocytotoxicity test. Results indicated positive results for HLA -DR 7-8 (DQ2-4;DR53). MRI of the brain revealed no pathological finding.
DISCUSSION

The patient described in this report had results consistent with the diagnostic criteria for periodic hypersomnia (Kleine-Levine type) (5). This patient is one of the few females who has this diagnosis in the literature. Although there are some reported cases with onset of symptoms temporally related to head injury (6) or febrile illnesses (4,7), this is the first case beginning during a postpartum period.

Polysomnographic studies revealed decreased sleep efficiency, prolonged sleep latency, and fragmentation of sleep by frequent arousals during the symptomatic period. In addition to these findings slow wave sleep (stages 3 and 4) was decreased. Similar findings were reported previously by some investigators (8,9) and seem to be the most significant established nocturnal findings in Kleine-Levine Syndrome. According to the polysomnographic results during the asymptomatic period, the amount of slow wave sleep was still low. The meaning and importance of this is unclear.

We did not observe REM sleep onset during the three nocturnal polysomnographic evaluations, but did on one of the MSLT naps. This finding supports the results of some researchers (7,8) and opposes the findings of others (1,9,10).

The occurrence of alpha intrusions throughout sleep is also a replicated finding in this patient group(7), the importance of which is also not clear .

Finally, the analysis of HLA-DR2 antigens on lymphocytes in this patient revealed that HLA-DR type is different from the one seen in the large majority of narcoleptic patients. Vischer and associates had already reported two Kleine-Levine Syndrome cases whose HLA-DR types were HLA-DR1 and HLA-DR1 and DR3 (11). Our findings implies that there may not be HLA-DR homogeneity in Kleine-Levine patients.
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REFERENCES


