Indian Journal of Pediatrics
https://doi.org/10.1007/s12098-024-05132-y

SCIENTIFIC LETTER

Response to Carbamazepine in a Case of Kleine-Levin Syndrome

Esra Ulgen Temel[1] · Esra Serdaroglu[2] · Habibe Koc Ucar[3] · Ayse Serdaroglu[2] · Ebru Arhan[2]

Received: 21 January 2024 / Accepted: 4 April 2024

To the Editor: A three-year-old boy was presented with 36–72-h attacks of unresponsiveness with normal vital signs and absence of specific neurological symptoms. Cerebrospinal fluid (CSF) studies, metabolic screening tests, anti-GAD antibodies, and thyroid tests were normal. In CSF, autoimmune encephalitis autoantibodies were not detected. He had ALT and AST elevations. Cytomegalovirus IgM and PCR were positive. EEG and brain MRI were normal. He recovered in 10 d. Within 7 mo, the patient experienced two more similar episodes. Since he had recurrent attacks, intravenous immunoglobulin and steroid treatments were administered. During the episodes, he became violent and disoriented.

He had his fourth attack in our clinic. The EEG was consistent with non-REM 1–2 sleep without any encephalopathic findings. The patient had food aversions and crying attacks. Video polysomnography-electroencephalography (PSG-EEG) monitoring showed very short sleep latency (150 s) and consisted of 73% non-REM-2, 16% non-REM-3, and 1.3% REM episodes. In the multiple sleep latency test, there was no sleep-onset REM period (Supplementary Figs. S1 and S2). PSG findings suggested the diagnosis of Kleine-Levin syndrome (KLS) with increased total sleep time, short sleep latency, increased non-REM-2 sleep, and absence or decrease of REM sleep [1]. The patient woke up spontaneously after 72 h.

The patient was diagnosed with Kleine-Levin syndrome based on clinical observations, which included recurring attacks of excessive sleepiness, eating disorder, disinhibited behavior, cognitive dysfunction, and PSG findings [1, 2]. Carbamazepine therapy was initiated. He has been attack-free for 4 y. The reported median disease duration was 8 y. Therapeutic duration is unclear. Attacks may recur after drug withdrawal [3].

Infectious conditions are the most frequent triggers. Our patient's first episode started with a cytomegalovirus infection, and a long journey probably triggered the last [4].

Carbamazepine is suitable for treating KLS in young children because of its wider dosing range, higher tolerance, and fewer side effects [3].

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s12098-024-05132-y.

Declarations

Conflict of Interest None.

References

1. Gadoth N, Kesler A, Vainstein G, Peled R, Lavie P. Clinical and polysomnographic characteristics of 34 patients with Kleine-Levin syndrome. J Sleep Res. 2001;10:337–41.
2. Perez-Carbonell L, Leschziner G. Clinical update on central hypersomnias. J Thorac Dis. 2018;10:S112–23.
3. Arnulf I, Zeitzer JM, File J, Farber N, Mignot E. Kleine-Levin syndrome: a systematic review of 186 cases in the literature. Brain. 2005;128:2763–76.
4. Billiard M, Jaussent I, Dauvilliers Y, Besset A. Recurrent hypersomnia: a review of 339 cases. Sleep Med Rev. 2011;15:247–57.

✉ Ebru Arhan
petekarhan@yahoo.com.tr

[1] Division of Child Neurology, Cengiz Gökçek Maternity and Children's Hospital, Gaziantep, Turkey

[2] Department of Child Neurology, Faculty of Medicine, Gazi University, Ankara, Turkey

[3] Division of Child Neurology, Adana City Training and Research Hospital, University of Health Sciences, Adana, Turkey

Published online: 19 April 2024