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Clinical Observations

Kleine-Levin Syndrome: A Case Report and Review of Literature

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ABSTRACT

BACKGROUND: Kleine-Levin syndrome presents with recurrent hypersomnia along with a number of other neuropsychiatric features, of which hyperorality has not been described frequently. **METHOD:** We report a male adolescent who presented with recurrent hypersomnia, hypersexuality, and hyperorality. Magnetic resonance imaging of the brain and overnight polysomnography followed by a multiple sleep latency tests were ordered. Excessive daytime sleepiness was assessed with the Epworth Sleepiness Scale. **RESULTS:** Magnetic resonance imaging of the brain did not reveal any abnormality. Overnight video-synchronized polysomnography and multiple sleep latency tests ruled out narcolepsy. Epworth Sleepiness Scale score at baseline was 22. He was prescribed lithium carbonate 300 mg twice a day. The symptoms improved within a week after starting lithium carbonate therapy. **CONCLUSION:** Kleine-Levin syndrome may present with hyperorality, and our patient responded well to lithium carbonate therapy.

Keywords: Kleine-Levin syndrome, recurrent hypersomnia, adolescent, lithium

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Introduction

Kleine-Levine syndrome (KLS) is characterized by recurrent episodes of excessive sleepiness that last from 2 days to 4 weeks, with at least annual recurrence.[1] The alertness, cognitive functioning, and behavior remain normal between attacks. The diagnosis of KLS is clinical; however, other causes of hypersomnia must be ruled out.[1] Around 60% of the cases have one precipitating factors, the most frequent of which is a trivial flu-like symptom or nonspecific fever.[2] It is an exceptionally rare disease, having only 200 reported cases through 2005.[1,2] Since then, just five more cases have been reported from India.[3-7]

KLS can be divided into primary and secondary forms (depending on presence of neurological symptoms before appearance of KLS symptoms that persisted between episodes).[1,2] The secondary forms are characterized by a later age at onset, longer episodes, higher number of episodes, and more incapacitation.[3] Secondary forms have been reported to be associated with stroke, posttraumatic brain hematoma, genetic or developmental diseases, multiple sclerosis, hydrocephalus, autoimmune encephalitis, and severe infectious encephalitis.[1-3]

We are here presenting a case of primary KLS that showed dramatic response to lithium carbonate.

Case report

A 16-year-old boy presented with an episodic hypersomnia for the past 3.5 years. The symptoms were first noticed at age 14 after an upper-respiratory tract infection. After this, his sleep extended beyond his usual nighttime sleep of 8 hours (10 pm to 6 am) for the next 20 days. He remained asleep even during the day and was difficult to wake during this period. Forceful awakenings made him irritable. On spontaneous and forceful awakenings, he appeared to be in an oneiroid state. According to him, everything appeared unreal at that time. On one occasion, he shouted and verbally abused at an imaginary hawker claiming that he belonged to a local underworld gang and would kill the hawker if he ever dared to come to his house again. During this period, he had to be stimulated repeatedly to gain his attention. Also, at times he began to chew on unusual items such as paper, nails, and plastic. On other

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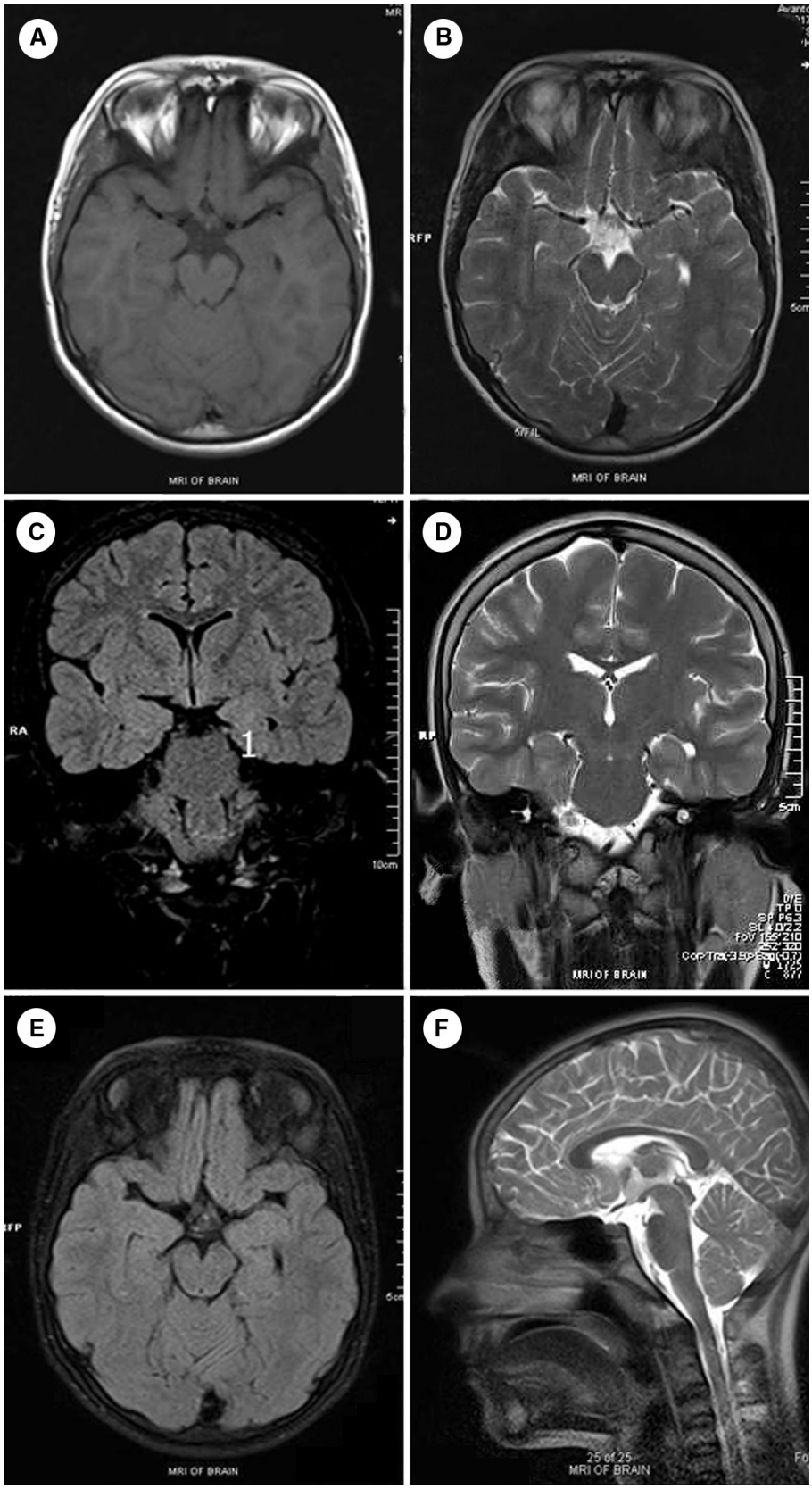

FIGURE.
(A) T1-weighted image showing a normal medial temporal lobe. (B) T2-weighted image showing a normal medial temporal lobe. (C) Fluid-attenuated inversion recovery showing no abnormality in the amygdala or hippocampus. (D) T2-weighted image showing normal medical temporal lobe. (E) fluid-attenuated inversion recovery image showing normal medial temporal lobe. (F) T2-weighted image showing a normal hypothalamus.