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Case Report

Kleine–Levin Syndrome: A case report ☆

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ABSTRACT

The Kleine–Levin Syndrome is a differential diagnosis for patients with diurnal excessive sleepiness and a suspicion of narcolepsy. It is characterized by paroxysmal attacks of diurnal excessive sleepiness, associated with one or more symptoms of hyperphagia, hypersexuality, coprolalia and copropraxia. During crisis intervals, there are no symptoms. This pathology predominantly manifests itself in teenagers, being more frequent among males. The course of this disease is unpredictable, with variable duration and frequency. The most accepted physiopathology is that of a hypothalamic dysfunction, although and recently, there has appeared a hypothesis of a post-infectious autoimmune disorder. These patients show an elevated body mass index, which can predispose to association with comorbidities such as the sleep obstructive apnea syndrome. Treatment involves medications with different effects, but there is no specific and effective therapy. Our article shows a classic case of Kleine–Levin Syndrome associated with sleep obstructive apnea syndrome, a rare association in the literature.

1. Introduction

The Kleine–Levin Syndrome (KLS) is a disease characterized by recurrent periods of hyper-sleepiness, with absence of urinary incontinence and the presence of verbal responses to intense stimuli. Hyperphagia, hypersexuality and behavioral or cognitive alterations (irritability, aggressiveness, mental confusion, and disorientation) are signs and symptoms found in isolation or in conjunction among these patients. A KLS patient might sleep for 18 hours a day and the crisis usually manifests at least once a year, ranging from 3 days to 3 weeks [1,2]. At crisis intervals, behavior, cognition and sleep are normal.

The disorder is more frequent among teenagers of the male gender [1–5] and has an unpredictable course with

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Table 1 – General characteristics.

	Basal	After 5 years
Age (years)	19	24
Height (m)	1.78	1.78
Weight (kg)	101.8	113.0
IMC (kg/m^2)	32.1	35.7
Cervical circumference (cm)	43	44
Abdominal circumference (cm)	100	110
Arterial blood pressure (mmHg)	130 × 80	140 × 80

Body mass index (BMI).

remissions and recurrences which can last for years. This disease can disappear as subtly as it appears (an average of 8–13 years duration) [2,5].

The first KLS episode is generally preceded by an event associated with the beginning of the symptoms, more commonly an infection, alcohol ingestion, sleep deprivation, stress, physical exhaustion, travels, brain trauma, surgeries with local or general anesthesia, lactation, menstruation and the use of drugs [2,3,5–7]. The precipitating factors are found more easily at the first episode of the disorder, being much less frequent in subsequent episodes [2,4].

Patients may also refer intense dreams, hallucinations, persecution nightmares and compulsions. They may also show difficulty of speech and reading, disorientation and perception alterations [2,5]. Sleep paralysis and cataplexy are not common. Patients during a crisis are able to wake up spontaneously to go to the bathroom and eat, but may become aggressive and irritated if awoken or impeded of sleeping [2]. Hypersexuality is more frequent among men and appears to be predictive of a more prolonged disease [2,4,5]. Dis-autonomic complaints may be identified in up to 25% of the patients (heavy sudoresis, blushing, congested or edematous face, hypotension, bradycardia and nausea) [4]. After the crisis, the patient might show insomnia, sometimes associated with euphoria and to partial amnesia with a duration of up to 24 h [2,4,5].

2. Clinical case

FSM, 19 years old, born in the state of São Paulo, began to show the symptoms in March, 2008, at 14 years of age, with "excessive sleepiness periods", referred by his parents.

The patient was born through a C section, normally and without any problems. The patient had normal neuropsychomotor development, vaccination according to the Brazilian schedule and without history of previous diseases. The patient's mother referred that the patient ate too much, although the food was not of a high nutritional content. His sleep pattern involved a 30 min nap after lunch, waking up well and sleepless. He usually went to bed between 22 and 23:00 h, taking about 30 min to sleep and waking up at 5:30 h in the morning. He slept about 6 h during the week and between 9 and 10 h on weekends. In addition, he showed eventual somniloquy and high and constant snoring, without episodes of observed apnea. There was no familiar history of excessive sleepiness. His parents demonstrated eventual snoring and the mother was hypertense and obese. The diurnal excessive sleepiness (DES) attacks began around 13 years of age and had a duration of 8–17 days and were only interrupted by familiar interference. The attacks were accompanied by a lethargic behavior with intense apathy as a triggering factor. Concomitantly, there was a decrease in verbal communication – "answered only when asked something" – demonstrating increased sudoresis as the crisis intensified.

At physical examination, the patient was of 1.78 m height, 101.8 kg weight, with cervical and abdominal circumference of 43 cm and 100 cm, respectively; regular 2-beat cardiac rhythm, heart rate of 80 bpm and arterial pressure of 130/80 mmHg. Table 1 shows the main characteristics of the patient. Oral examination demonstrated elongated uvula, web palate, tonsils occupying less than 50% of the oral-pharynx and a Mallampati index of grade II.

At first consultation, the patient was already being followed up by a psychiatrist and was taking venlafaxine 75 mg/day and oxycarbamezepine 300 mg/day. His performance at school had not been decreasing and his grades followed the pattern before the onset of the disease. The psychiatrist described that the teenager had shown persecutory delirium at the onset of the disorder and that the crisis were triggered by stress. There were no complaints of hallucinations or cataplexy. The patient referred "laziness", "sleepiness", and "slow thinking" at the beginning of the crisis, isolated himself during the crisis and only awoke to eat or go to the bathroom and stated that he did not have recollection of what was going on during the crisis. The family described hypersexuality during the crisis. The interval between crises was of 3 and 4 months' duration. The patient also referred that his anxiety triggered his crisis. When awake during the crisis, the patient stayed quiet, did not laugh or talk to the family, demonstrating sudoresis and normal thinking process.

The first polysonogram, in May 2009, demonstrated increase in the percentage of slow waves, without other alterations. The electroencephalogram demonstrated diffuse slowing waves during crisis. The diagnosis was that of Kleine–Levin Syndrome and we initiated a reduction of venlafaxine to 37.5 mg/day until posterior suspension. The dose for oxycarbamazepine was increased to 450 mg/twice daily.

His humor progressively improved along the days of treatment. He finished high school and is now working. However, since he has been taking oxycarbamazepine 600 mg/day, his family describes that his humor only improved ("he is less