A Patient with Intractable Attacks of Cataplexy

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A 29-year-old white man presents for evaluation of refractory daily generalized convulsions and drop attacks previously diagnosed as cataplexy. The patient's typical convulsive events started at 8 years of age, occur daily, and consist of jerking in all extremities with preserved consciousness. He has been on multiple anti-epileptic drugs (AEDs) including phenobarbital, valproic acid, and levetiracetam without decrease in event frequency. At age 17 he started having daily drop attacks consisting of full body weakness followed by collapse. The drop attacks, referred to as "cataplexy" by the patient, always occurred in public, have no clear trigger and have never resulted in injury to the patient. A typical drop attack is shown in **Video 1**.

Magnetic resonance imaging of the brain, and 3 routine electroencephalograms (EEGs) without video monitoring were normal. A polysomnogram (PSG) followed by multiple sleep latency testing (MSLT) performed at an outside institution while on AEDs was interpreted as showing 5 out of 5 sleep onset REM sleep periods (SOREMPs), with an average sleep latency of 1.2 minutes (data from PSG was not available). With complaints of daytime sleepiness, the patient was diagnosed with narcolepsy with cataplexy and started on sodium γ -hydroxybutyrate (GHB) with no improvement in drop attack frequency. Clomipramine and venlafaxine were added with no improvement noted.

The patient started hearing voices at age 19 while not on any stimulant medications. Auditory hallucinations occurred in the context of paranoid ideation

that others were going to harm him. The patient fulfilled criteria of the Diagnostic and Statistical Manual of Mental Disorders IV for paranoid-type schizophrenia.

His current medications are sodium γ -hydroxybutyrate (GHB) 6 grams at bedtime and 6 grams at 2 AM; levetiracetam 500 mg twice per day; and venlafaxine 150 mg once per day.

The patient reported snoring and waking up gasping for air. Physical exam revealed a body mass index (BMI) of 42 with Mallampati IV and large tongue. Obstructive sleep apnea (OSA) was highly suspected. A repeat PSG showed an apnea hypopnea index of 28. Continuous positive airway pressure (CPAP) PSG titration revealed a pressure requirement of 9 cm H₂O. The patient reported nightly CPAP use, but downloadable data was not available and patient did not complete sleep diaries as requested.

A repeat PSG-MSLT on 9 cm of CPAP was performed. The MSLT was interpreted as having no SOREMPs and was terminated after 4 naps. After sleep physician review, a single epoch of REM sleep was noted in the first nap and the average sleep latency was 1 minute. The patient was on levetiracetam during the PSG-MSLT. During a 30-min video-EEG recording, multiple typical generalized convulsive episodes with eyes closed and maintained ability to communicate were captured (**Video 2**). Neither epileptiform activity nor changes in the normal waking background EEG were noted during the event (**Figure 1**). The video-EEG recording was consistent with a diagnosis of psychogenic non-epileptic seizures.

Figure 1

Typical generalized convulsive event capture on video-electroencephalogram (EEG) recording. EEG showed neither epileptiform activity nor changes in the normal waking background activity during the convulsive event. This finding was consistent with a diagnosis of psychogenic non-epileptic attacks.

QUESTION: What is the diagnosis of the patient's drop attacks seen in Video 1?

ANSWER: Pseudo-cataplexy**DISCUSSION**

Video recording of a typical drop attack showed backward collapse from a seated position, without emotional precipitant (**Video 1**). During the event deep tendon reflexes were 2+ at the biceps tendons bilaterally, 2+ at the patellar tendons bilaterally, and 1+ at the Achilles' tendons bilaterally. The patient regarded this episode as his typical cataplexy attack.

A very high frequency of "seizures" completely unaffected by AEDs should suggest the possibility of a psychogenic etiology. Eye closure during an ictal event, as seen in **Video 2**, is unusual during an epileptic seizure and also suggestive of a psychogenic etiology.

Cataplexy, a pathognomonic finding of narcolepsy, is defined as an emotionally triggered sudden loss of focal or generalized muscle tone with absence of deep tendon reflexes.¹ It is imperative to accurately evaluate the patient with suspected cataplexy due to the diagnostic and treatment implications.

Episodes of muscle weakness have frequently been reported in non-narcoleptic subjects, especially in the presence of sleep-disordered breathing.² The differential diagnosis of cataplexy includes: gelastic atonic seizures, drop attacks due to brainstem ischemia, and pseudo-cataplexy. Gelastic seizures have been associated with hypothalamic hamartomas; gelastic atonic seizures with Neiman-Pick Disease; and atonic seizures with Lennox-Gastaut Syndrome. Gelastic atonic seizures, characterized by laughter and loss of muscle tone, may be difficult to differentiate from cataplexy, as both ictal and inter-ictal EEG may be normal.³ Though gelastic seizures may resemble true laughter, the patient does not experience mirth and often feels a building internal pressure to laugh.

Pseudo-cataplexy has been described in narcolepsy with cataplexy.⁴ Pseudo-cataplexy events are characterized by the absence of emotional triggers or in some cases negative emotional triggers such as crying, generalized weakness, abrupt onset, preserved ability to communicate, and preserved deep tendon reflexes. Pseudo-cataplexy events as well as pseudo-seizures have a higher occurrence in public places.⁵ Our patient fits many of the previously described features of pseudo-cataplexy, such as generalized weakness, abrupt onset, preserved consciousness, and public occurrence. In our patient emotional triggers did not precede his events.

Pseudo-cataplexy has been associated with mood disorders, while psychogenic non-epileptic seizures have been associated with both mood and psychotic disorders.⁶

There is clinical and electrophysiologic overlap between narcolepsy, cataplexy, and depression. Narcolepsy and depression both result in alterations to REM sleep; cataplexy and depression both respond to antidepressant medications; and cataplexy has features resembling catatonia/psychomotor retardation.⁷

In our patient, the presence of a SOREMP in the MSLT may be secondary to sleep deprivation, mood disorder, medication withdrawal, schizophrenia,⁸ or narcolepsy. The International Classifications of Sleep Disorders precludes the diagnosis

of narcolepsy in those with psychiatric conditions, or those on medications that may lead to excessive daytime sleepiness.¹ The MSLT as a diagnostic tool in hypersomnias of central origin has been challenged by the presence of SOREMPs in many other conditions (e.g., obstructive sleep apnea); and the poor test-retest reliability of the MSLT.⁹ Cataplexy, however, is considered pathognomonic of narcolepsy and should be carefully investigated in each patient. The concomitant diagnosis of OSA, narcolepsy, and pseudo-cataplexy in our patient cannot be definitively excluded.

The patient was slowly and sequentially titrated off levetiracetam, GHB, and venlafaxine, and started extensive psychotherapy. The patient is currently on haloperidol 5 mg per day, paliperidone 9 mg per day, and benztropine 2 mg per day. Schizophrenia paranoid type has a better prognosis than the other schizophrenia subtypes, and the patient is showing gradual reduction in convulsion and drop attack frequency.

SLEEP MEDICINE PEARLS

1. The differential diagnosis of cataplexy includes: gelastic seizures, gelastic atonic seizures, atonic seizures, drop attacks due to brainstem ischemia, and pseudo-cataplexy.
2. Pseudo-cataplexy events are characterized by negative emotional triggers such as crying, generalized weakness, abrupt onset, preserved ability to communicate, and preserved deep tendon reflexes.
3. Pseudo-cataplexy has been associated with mood disorders.
4. Pseudo-cataplexy and psychogenic non-epileptic attacks do not preclude the diagnosis of concurrent cataplexy or epilepsy. Careful evaluation is needed to exclude these conditions.

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