NEW RESEARCH

JCSM
Journal of Clinical Sleep Medicine

CASE REPORTS

A Case of Narcolepsy with Strictly Unilateral Cataplexy

David E. McCarty, M.D.

Louisiana State University Health Sciences Center, Shreveport, LA

I report a case of a 50-year-old African American woman who presented for a sleep medicine evaluation complaining of excessive daytime sleepiness of greater than 10 years' duration. She reported falling asleep while driving, talking with friends, and eating. Her Epworth Sleepiness Scale score was 21/24. Her history was positive for the presence of sleep paralysis. She denied visual or auditory hallucinations in the peri-sleep period.

Keywords: Cataplexy, narcolepsy, atonia, unilateral

Citation: McCarty DE. A case of narcolepsy with strictly unilateral cataplexy. *J Clin Sleep Med* 2010;6(1):75-76.

Cataplexy is a transient decrease in voluntary muscle tone triggered by characteristic emotional stimuli and, along with excessive daytime sleepiness, sleep paralysis, and hypnogogic hallucinations, is part of the diagnostic tetrad for narcolepsy. The legs, arms, muscles of the jaw, and postural muscles are typically involved, and, in some patients, cataplexy spells can be sufficiently frequent and severe as to impair work performance or social activities. There can be considerable variation in cataplexy severity both between patients and within the same patient, though episodes of weakness are most often symmetric in distribution.[1] We have recently encountered an interesting patient with strictly unilateral cataplexy associated with narcolepsy.

REPORT OF CASE

A 50-year-old African American woman presented for a sleep medicine evaluation complaining of excessive daytime sleepiness of greater than 10 years' duration. She reported falling asleep while driving, talking with friends, and eating. Her Epworth Sleepiness Scale score was 21/24. Her history was positive for the presence of sleep paralysis. She denied visual or auditory hallucinations in the peri-sleep period.

When questioned about possible cataplexy, the patient described episodes of right-sided weakness that had begun approximately 10 years prior. Episodes were sporadic, initially occurring approximately once monthly, but, over the preceding 3 years, they had increased to occur up to several times weekly. The spells were typically brief, lasting anywhere from several seconds to 2 minutes, and the muscles involved were unswervingly right sided, including those of the right side of her face, the right arm, and the right leg. The events were precipitated by episodes of laughter or feelings of hostility or anger. They never occurred spontaneously. Events were unassociated with any headaches, sensory disturbances, cardiopulmonary symptoms, visual auras or disturbances, loss or alteration in consciousness, or seizure activity. Left-sided muscles were never involved. Though the events varied in their severity, all 3 areas (face, arm, and leg) were always affected similarly. When at their most severe, the events would produce a facial droop and slurred speech, which the patient reported was noticeable to others, as well as an inability to walk or use the right hand until the event would resolve. Upon resolution of the event, the patient would have full return of function.

Her past medical history was unremarkable. She had no history of migraines, head injury, seizures, cardiac disease, hypertension, or cerebrovascular accidents. There was no history of head trauma. She denied symptoms of depression. She denied any history of inflammatory arthritis symptoms. Laboratory tests that had been performed in the preceding year included a complete blood count, chemistry panel, liver function tests, and renal profile: the results were all within normal limits. The patient took no medications regularly and had never smoked cigarettes.

On examination, she was well appearing, with a normal blood pressure. Findings from her general medical examination, including her cardiopulmonary examination, were unremarkable, with normal heart sounds and no carotid bruits. Neurologic examination showed normal motor strength throughout, with normal reflexes. The results of magnetic resonance imaging of the brain with and without the use of intravenously administered contrast dye showed no abnormalities. Data from polysomnography and a Multiple Sleep Latency Test were diagnostic for narcolepsy (**Figure 1**). Standard-polysomnography montage electroencephalographic data showed no evidence of unusual slowing or epileptiform spikes. Unfortunately, attempts made during the MSLT (between naps) to provoke an event were unsuccessful.

The patient was diagnosed with narcolepsy, with the question of an unusual cataplexy variant. Because the patient was uninsured, the treatment plan required tailoring to allow for financial limitations. Over the ensuing months, she continued to have several episodes per week of transient right-sided weakness, precipitated by laughter or anger. Because these were not limiting her activities, specific anticataplectic therapy was not felt to be a justified expense. To assist with her daytime sleepiness, the patient was begun on methylphenidate ER 20 mg every morning, with a modest degree of success, though she continued to have problems with uncontrollable sleepi-

Figure 1—Sleep laboratory data

Polysomnographic Data

Latency to sleep onset:	0 min
Sleep efficiency:	88%
Sleep Staging:	78% N1/N2, 6% N3, 16% REM
Arousal Index:	17/hour
REM latency:	6 min
Leg movements:	n/a
Apnea-Hypopnea Index:	0.3/hour
Apnea Index:	0/hour

Multiple Sleep Latency Test Data

Nap #	Sleep Onset Latency (min)	REM Latency (min)
1	0	3
2	1.5	n/a
3	1.5	1.5
4	1	n/a
	Mean SOL= 1 min	2 sleep-onset REM periods

ness at midday. The patient did not tolerate immediate-release methylphenidate, which was tried in an attempt to address her breakthrough sleepiness. Strategic midday naps were added to her regimen with some limited success.

Due to continued symptoms of daytime sleepiness, as well as some symptoms of mild depressed mood, venlafaxine 75 mg was added as a single morning dose, with the hope that this would improve her daytime alertness. This resulted not only in the patient's improved ability to maintain wakefulness during the midday lull, but also in a near-complete resolution of her episodic right-sided weakness. Over a year has elapsed since the patient first began taking venlafaxine. She reports that her symptoms of unilateral weakness continue to respond to venlafaxine treatment, with breakthrough events occurring only if she runs out of her medication.

DISCUSSION

We describe here a case of recurring strictly right-sided weakness that we believe represents an unusual variant of cataplexy in a patient with both clinical and polysomnographic features of narcolepsy. Other than the strictly unilateral distribution, the episodes were typical of cataplexy in other ways: they were brief and consistently provoked by laughter or anger; they resolved completely between attacks; and they were dramatically improved with the introduction of venlafaxine, an agent that has a trusted record of efficacy for the treatment of cataplexy symptoms.[2] Other possible etiologies of episodic weakness include transient ischemic attacks, hemiplegic migraines, akinetic seizures, and conversion disorder, all of which seem relatively unsupported by the history: there was no other evidence of vascular disease either on examination or sensitive central nervous system imaging, events were unaccompanied by other features of either epilepsy or migraines, the patient had no other evidence of cardiac disease to suggest transient dysrhythmias, and there was no history or clinical evidence of other significant psychiatric disease. Lastly, the patient never sought any medical intervention for any degree of "secondary gain" to suggest malingering.

The intensity of weakness associated with cataplexy can be quite variable, producing a clinical syndrome that spans the distance between a trifling annoyance to a full-blown disability. This, coupled with the fleeting nature of the events and the rarity with which they can be objectively observed by physicians, has historically flummoxed the systematic study of cataplexy. One of the best investigations of the clinical characteristics of cataplexy came from Anic-Labat and colleagues,[1] who demonstrated that 68% to 81% of cataplexy sufferers report "always bilateral" symptoms, implying that as many as 32% may have unilateral symptoms, at least some of the time. What is not at all clear, however, is how many patients may manifest symptoms solely constrained to the same side of the body across all events. A case of strictly right-sided cataplexy has been reported in a patient with systemic lupus erythematosus and daytime sleepiness, but polysomnographic data were not described and, being 1976, only rudimentary brain imaging was obtainable.[3]

The fundamental mechanisms of rapid eye movement sleep-related atonia (and, by extension, cataplexy) are complex and incompletely understood.[4,5] It is our hope that this description of this unusual strictly unilateral cataplexy phenotype may add to the evolving understanding of this fascinating problem.

REFERENCES

1. Anic-Labat S, Guilleminault C, Kraemer HC, Meehan J, Arrigoni J, Mignot E. Validation of a cataplexy questionnaire in 983 sleep-disorders patients. *Sleep* 1999;22:77-87.
2. Guilleminault C, Fromherz S. Narcolepsy: diagnosis and management. In: Principles and Practice of Sleep Medicine, 4th ed. Kryger M, Roth T, Dement W, eds. Philadelphia, PA: Elsevier Saunders; 2005:780-90.
3. Lascelles R, Mohr P, Peart I. Unilateral cataplexy associated with systemic lupus erythematosus. *J Neurol Neurosurg Psych* 1976;39:1023-6
4. Brooks PL, Peever JH. Glycinergic and $GABA_A$-mediated inhibition of somatic motoneurons does not mediate rapid eye movement sleep motor atonia. *J Neurosci* 2008;28:3535-45.
5. Lydic R. The motor atonia of REM sleep: a critical topics forum. *Sleep* 2008;31:1471-72.

SUBMISSION & CORRESPONDENCE INFORMATION

Submitted for publication November, 2008
Submitted in final revised form July, 2009
Accepted for publication August, 2009

Address correspondence to: David E. McCarty, M.D., Sleep Medicine Program, LSUHSC-Shreveport, 1501 Kings Highway, Shreveport, LA 71130; E-mail: dmcca1@lsuhsc.edu

DISCLOSURE STATEMENT

The author has indicated no financial conflicts of interest.