NEW RESEARCH

JCSM
Journal of Clinical Sleep Medicine

http://dx.doi.org/10.5664/jcsm.3456

Status Cataplecticus as Initial Presentation of Late Onset Narcolepsy

Samhita Panda, DM, Neurology

Department of Sleep Medicine and Neurophysiology, Sir Ganga Ram Hospital, New Delhi, India

CASE REPORTS

Narcolepsy, one of the important causes of hypersomnia, is an under diagnosed sleep disorder. It has a bimodal age of onset around 15 and 35 years. It is characterized by the tetrad of excessive daytime sleepiness, cataplexy, hypnagogic/hypnopompic hallucinations, and sleep paralysis. Cataplexy is by far the most predictive feature of narcolepsy. Status cataplecticus is the occurrence of cataplexy repeatedly for hours or days, a rare presentation of narcolepsy. This report describes an elderly gentleman with late onset narcolepsy in the sixth decade of life presenting with initial and chief symptom of status cataplecticus.

Keywords: Narcolepsy, cataplexy, status cataplecticus, head drop, late-onset

Citation: Panda S. Status cataplecticus as initial presentation of late onset narcolepsy. *J Clin Sleep Med* 2014;10(2):207-209.

Narcolepsy is a disorder of the normal boundaries between the sleeping and waking states.[1] Although not rare, it is an under recognized and under diagnosed entity. Its prevalence in the US is 1 in 2000 affecting males and females equally.[1] Though narcolepsy can present at any age, majority does so between 15-30 years of age and only 6% do so prior to 10 years of age. Rarely, narcolepsy is reported after age of forty.[2] Also cataplectic symptoms generally decrease with age so much so that the patient may stop taking anti-cataplectic medications. We report a patient who first presented with narcolepsy in the sixth decade of life with status cataplecticus.

REPORT OF CASE

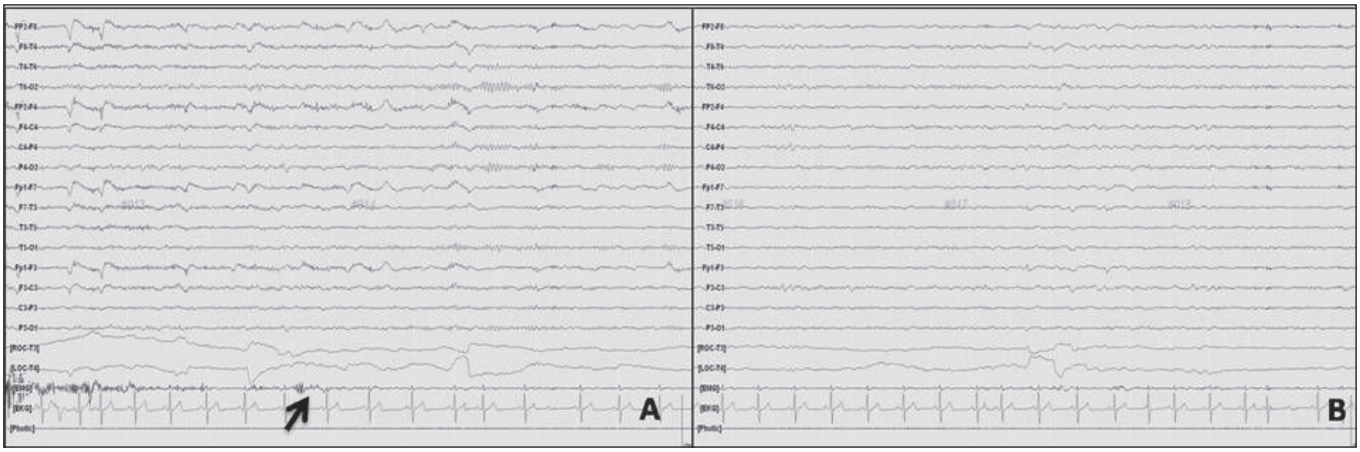
A 57-year-old gentleman presented with recurrent episodes of head jerking resulting in repetitive head nodding for 6 months. At times this was associated with closure of eyes and transient change in character of voice. The subject would also inadvertently drop objects or stumble. Though briefly unresponsive, he was aware and arousable both during and after the attacks. The events would occur while standing, walking, talking, conversing over telephone, watching TV, pressing a doorbell, or even eating. There was no correlation with change in mood or emotional situations. The initial frequency (1-2/week) had recently increased to multiple daily episodes and were almost continuous for preceding one month. Increased daytime somnolence was also noted during the preceding 2 years. Snoring was occasionally noted without choking or apneic spells, restless legs, limb movements, or other abnormal behavior in sleep. Excessive dreaming was observed during the preceding 4-5 months, to the extent that at times he would hallucinate in wakefulness and would feel himself talking to long-deceased friends and family members.

There was no aura, presyncopal symptoms, generalized tonic clonic seizures, tongue biting, frothing, incontinence, or major injuries. There was no significant illness in the past. The subject regularly used alcohol and smoked cigarettes for the past 25 years. There was no history of similar illness, epilepsy, or excessive daytime sleepiness in the family.

The subject was a middle-aged gentleman of medium build with weight of 78 kg, height 165 cm, and body mass index of 28.6. Blood pressure was 130/90 mm Hg. There was no abnormality on general or systemic examination. Epworth Sleepiness Scale (ESS) score was 14/24. Intermittent head nodding, drooping and closure of eyes, and thickening of voice were noted during examination. He was quiet and unresponsive for a brief period with diminished deep tendon jerks.

Magnetic resonance imaging of brain showed few chronic lacunar infarcts in left frontal subcortical region and pons. Short-term video-electroencephalography (EEG) monitoring was done using 16-channel scalp EEG (International 10:20 electrode configuration), bilateral electrooculographic (EOG) tracings, surface electromyography (EMG) of submental musculature, and electrocardiography (EKG). It demonstrated normal awake EEG between the events. Innumerable events were noted, almost in tandem. There was uncontrollable nodding of head and asynchronous, irregular jerks of upper limb, at times amounting to dropping of food and drinks (see **Videos 1** and **2**). **Video 1** shows brief cataplectic attacks resulting in head nodding and excessive daytime sleepiness, and **Video 2** shows status cataplecticus with preserved consciousness while eating. Jerks were of varying intensity and lasted from few seconds to 5 minutes. EEG showed change in EMG tone to relative atonia with intermittent rapid eye movements (REM) and mixed frequencies (**Figure 1A** and **1B**). Polysomnography was done subsequently using EEG tracings (modified 10-20 system with bilateral central and occipital channels), EOG tracings, surface EMG from submental and tibialis anterior muscles, EKG, arterial oxygen hemoglobin saturation via finger pulse oximetry, pressure transducer for airflow,

Figure 1—EEG during a cataplectic attack showing **(A)** transition from wakefulness to sudden atonia in submental EMG; **(B)** onset of cataplectic attack with REM intrusion



piezoelectric belts for chest and abdominal movements, and snoring intensity via decibel meter recording. Polysomnography showed sleep efficiency of 94.9% and short sleep latency (0.5 minutes). There was early onset REM sleep with REM predominance (27.9%) with no evidence of obstructive sleep apnea. Multiple sleep latency test (MSLT) showed average sleep latency of 84 seconds. MSLT was not extended further beyond 3 naps, as there were sleep onset REM (SOREMs) in all 3 naps. A diagnosis of narcolepsy with status cataplecticus was finally considered. HLA DQ typing by DNA-SSP method was positive and homozygous for HLA-DQ B1*06 allele. CSF evaluation for hypocretin was not done as the diagnosis was clear. Detailed evaluation of the family members for narcolepsy and HLA typing was not done due to economic constraints.

The patient was treated with clomipramine (20 mg per day) and armodafinil (150 mg per day). There was significant improvement in clinical symptoms. After 6 months follow-up, the head dropping episodes have gradually reduced to once in 2-3 weeks, daytime somnolence has reduced, night sleep has improved, and there are no hypnagogic hallucinations. No new neurological symptoms or deficits have been noted, and the patient has resumed his job which involved travelling and supervision.

DISCUSSION

Cataplexy is the second most common symptom and most predictive feature of narcolepsy.[1] It is an isolated REM intrusion into wakefulness associated with sudden loss of muscle tone with preserved consciousness as seen in the index case. Narcolepsy has a bimodal age of onset around 15 and 35 years, rarely occurring after age of forty.[2,3] The case described here exemplifies narcolepsy with later onset in the sixth decade and status cataplecticus as the initial and primary symptom.

Cataplexy is usually localized to specific muscles or parts of the body causing sagging of face, eyelid or jaw, dysarthria, dropping of head, weakness of arm or leg, and buckling of knees as seen in our patient.[1] Face and neck were more specifically involved, resulting in characteristic head drops, which resulted in initial diagnostic confusion with myoclonic or atonic seizures, movement disorders, hysterical disorders,[4] and fatigue. This was clarified by video EEG monitoring. Unlike classical cataplexy that is usually triggered by sudden excitement, emotions such as laughing, anger, fear, frustration, nervousness, sadness, embarrassment, and extreme exertions, this patient had essentially spontaneous attacks. More specifically, status cataplecticus has been noted after abrupt cessation of medications such as clomipramine,[5] addition of prazosin for hypertension, childbirth, sexual excitement, and in patients with symptomatic cataplexy, especially children with Niemann Picks disease. Interestingly, no definite precipitating factors were noted in our patient. Cataplexy differs from typical REM sleep in many aspects, as waking and a low-voltage EEG are maintained during short cataplectic attacks. This was also observed in our patient. Consciousness is always maintained at the onset of cataplexy, but patients may experience sleepiness, dreamlike hallucinations, or sleep-onset REM periods as the attack continues.

Idiopathic narcolepsy with cataplexy usually does not have radiological correlate. Symptomatic cases have been seen mainly with hypothalamic lesions, and rarely with pathology involving the midbrain, diencephalon, and medial pontine tegmentum.[6] Also, focal lesions are associated with borderline narcolepsy symptoms. No significant abnormality was found on neuroimaging which could explain these symptoms. A few subtle areas of chronic ischemic insult in the pons with no associated symptoms at onset or after 6 months ruled out symptomatic narcolepsy. HLA DQB1*0602, a sensitive marker for narcolepsy across all ethnic groups, was positive in our patient. HLA DQB1*0602 positivity has been found to have a tighter association in patients of narcolepsy with cataplexy (76.1%) than without cataplexy (40.9%).[7] Deficiency of neurotransmitter hypocretin in the hypothalamus was not documented here, as evaluation of CSF hypocretin was not considered to provide additional information. Low CSF hypocretin is seen in 99% of patients with cataplexy positive for HLA DQB1*0602.[8] However, it is possible that coexisting anatomical lesions may facilitate the appearance of narcoleptic symptoms in genetically predisposed subjects. Stressors and sudden lifestyle changes may trigger onset of late onset narcolepsy with potential genetic-environmental interactions.[3]

The Indian subcontinent does not have prevalence data on narcolepsy. Only 22 cases of narcolepsy have been reported to date; 10 had narcolepsy with cataplexy.[9] Also, no case of status cataplecticus has been reported in the subcontinent. It is important for physicians, neurologists, sleep specialists, and psychiatrists to be aware of this condition, as it may be often confused as seizure or hysteria.

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SUBMISSION & CORRESPONDENCE INFORMATION

Submitted for publication May, 2013
Submitted in final revised form October, 2013
Accepted for publication October, 2013

Address correspondence to: Dr. Samhita Panda, Consultant Neurologist, Department of Sleep Medicine and Neurophysiology, Sir Ganga Ram Hospital, Rajender Nagar, New Delhi-110060; Tel: 91-11-42251714 (Office); Tel: 91-09810838268 (Mobile); Fax: 91-11-25861002; E-mail: samhitapanda@yahoo.com

DISCLOSURE STATEMENT

This was not an industry supported study. The author has indicated no financial conflicts of interest.