REM behavior disorder in myotonic dystrophy type 2

Figure A 60-second epoch from the patient's polysomnography

The top 6 channels represent EEG recording with electrodes placed according to the international 10-20 electrode placement system. E1-M1 and E2-M1 represent left and right electrooculogram (EOG) channels with reference to the left mastoid (M1). The chin channel represents EMG activity from the mentalis muscle. Multiple muscle recording includes EMG activity recorded from the following muscles: right sternocleidomastoid (RtSterno1), bilateral biceps and triceps, lumbar paraspinals (Paraspinals) and bilateral quadriceps femoris referenced to the respective iliac crests (Rt-Quad), bilateral gastrocnemius (Gast1) and tibialis anterior (Tib) muscles. Chest and abdominal effort were measured using respiratory inductive plethysmography belts. EKG (electrocardiography), SaO₂ (oxygen saturation by finger oximetry), HR (heart rate), and snore channels are also displayed. The C-flow channel demonstrates obstructive apneas with arousals. The epoch represents REM sleep, as evidenced by rapid eye movements in the EOG channels and a desynchronized, low voltage EEG; however, there are excessive phasic bursts in the chin and limb EMG channels. This activity was present in more than 50% of REM epochs, meeting the criteria for REM without atonia.

A 63-year-old woman with myotonic dystrophy type 2¹ and obstructive sleep apnea by polysomnography, not on medications traditionally known to cause REM behavior disorder (RBD), had dream-enacting behavior (DEB). Overnight continuous positive airway pressure titration captured an episode of DEB with clear recall (video on the *Neurology*[®] Web site at www.neurology.org). There was absence of muscle atonia and excessive phasic muscle bursts during REM, confirming RBD (figure).

RBD, commonly seen with neurodegenerative diseases, particularly synucleinopathies,² has not been described in myotonic disorders, making this a novel presentation. We speculate that RBD may have resulted from brainstem involvement of her multisystem generalized membrane disorder.

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Supplemental data at
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The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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