CASE REPORT

A restless abdomen and propriospinal myoclonus like at sleep onset: an unusual overlap syndrome

Simone Baiardi, Chiara La Morgia, Susanna Mondini, Fabio Cirignotta

Department of Biomedical and Neuromotor Sciences, University of Bologna, Neurology Unit, S. Orsola-Malpighi Hospital, AOU Bologna, Italy

Correspondence to
Professor Fabio Cirignotta,
fabio.cirignotta@unibo.it

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SUMMARY

We report for the first time the association between restless abdomen, a phenotypic variant of restless legs syndrome in which symptoms are limited to the abdomen, and propriospinal myoclonus at sleep onset causing severe insomnia. The treatment with a low-dosage of dopaminergic drug (pramipexole) induced the immediate disappearance of both symptoms, which was documented by video-polysomnography.

BACKGROUND

Restless legs syndrome (RLS), also known as Willis-Ekbom disease (WED), is a motor and circadian sleep disorder that commonly causes insomnia due to difficulty falling asleep.¹ The main features of RLS/WED are the urge to move legs at night time, which often occurs with a strange discomfort in lower limbs, and sudden relief after starting the activity.² In most cases, patients also experience involuntary and periodic jerks affecting muscles of the legs during relaxed wakefulness or while they are sleeping (periodic limb movements, PLMs). The symptoms are usually confined to the lower limbs, nevertheless, other body parts may also be affected; particularly, a phenotypic variant of RLS/WED has been recently described in which symptoms were limited to the abdomen muscles.³

Propriospinal myoclonus (PSM) is a type of spinal myoclonus in which myoclonic activity arises in the spinal cord but does not remain restricted to segmentally innervated muscles, spreading up and down the length of the spinal cord along propriospinal (intersegmental) pathways.^{4–6}

'PSM at sleep onset' is a distinct subtype of propriospinal myoclonus, which occurs during relaxed wakefulness when the patient attempts to fall asleep, making it difficult to initiate sleep and consequently causing insomnia;^{7–9} in some patients it was reported in association with typical RLS/WED.¹⁰

We report a case of a patient with a long history of insomnia in which clinical features suggest an atypical RLS associated with myoclonic jerks involving the abdominal wall, in which video-polysomnography demonstrates the presence of a spinal myoclonus with characteristics of a PSM-like at sleep onset.

CASE PRESENTATION

An 81-year-old man had a family history positive for RLS in the mother. Medical history was relevant for hypertension, hypercholesterolaemia, abdominal aortic aneurism, coronary artery disease, mild renal insufficiency and left renal artery

stenosis. The patient's concomitant medications were amlodipine (5 mg/day), bisoprolol (3 mg/day), irbesartan (150 mg/day), acetylsalicylic acid (160 mg/day), ticlopidine (250 mg/day) and simvastatin (20 mg/day).

Since he was 47 years old he reported of abdominal 'cramps' associated with unpleasant restless sensation of discomfort with the urge to move occurring in the evening when lying in relaxed wakefulness and when trying to fall asleep. These symptoms disappeared completely if the patient stood up and walked around. Initially, the symptoms occurred rarely, but their frequency increased over time, and at the time of our observation they recurred every evening, making falling asleep impossible.

INVESTIGATIONS

Neurological examination was unremarkable. Routine blood analysis, including iron metabolism examination, B₁₂ vitamin and folate were normal. Brain and spinal MRIs were not performed because of the presence of coronary medicated stents, but a spinal CT scan was unremarkable.

The patient underwent video-polysomnography recording (baseline VPSG₀) including EEG (F₄-M₁, C₄-M₁, O₂-M₁), electro-oculography, surface electromyography of mental and appropriate limb and trunk muscles (deltoid, rectus abdominis, thoracolumbar paraspinalis, rectus femoris and tibialis anterior), airflow parameters (nasal pressure transducer and thoracoabdominal respiration), electrocardiography and blood oxygen saturation. The VPSG₀ recording demonstrated, when the patient lay down in relaxed wakefulness, the occurrence of involuntary jerks involving at first the axial muscles (recti abdominis muscles and thoracolumbar paraspinalis muscles) followed by, at varying delays (range 50–300 ms), most of the caudal and rostral muscles, with a propriospinal pattern, and recurring at pseudoperiodic intervals (mean 18±10.5 s, range 6–57 s; [figures 1](#) and [2](#) and [videos 1](#) and [2](#)). The total number of jerks was 24 and the event rate was 24.4/h calculated on total bedtime (8.4/h considering the total recording time). The motor events were prevalent during the first part of the night (15 vs 9 events observed in the second part of the night); heart rate did not change in relation to the motor jerks. These repetitive and pseudoperiodic movements were associated with a subjective sensation of discomfort and restlessness involving the abdomen, which made falling asleep impossible. The symptoms disappeared completely when the patient stood up and walked around the room,

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Figure 1 Polysomnographic recordings of a single propriospinal myoclonus jerk: the onset of the electromyography activity (solid line) is in the right rectus abdominis muscle with later involvement of right thoracolumbar (T-L) paraspinalis (Parasp), right rectus femoris (Fem), right deltoid and right tibialis anterior (Tib Ant) muscles. The EEG discloses diffuse alpha activity.

reappearing with the same characteristics whenever he went back to bed. This pattern of myoclonic jerks recurred overnight, forcing the patient to spend 65% of the total registration time out of bed. The patient was conscious of the abdominal jerks and claimed he could not suppress them, but when he was distracted (ie, by interviewer's questions, or if he was reading a newspaper), they immediately disappeared. The total sleep time was significantly reduced (18 min 30 s) and no myoclonic jerk was recorded during sleep (figure 3). Interestingly, VPSG₀ did not document PLMs during wakefulness or during sleep.

TREATMENT

A therapy with pramipexole 0.18 mg/day was started with complete and immediate disappearance of symptoms.

OUTCOME AND FOLLOW-UP

The follow-up VPSG₁, performed after 44 days of therapy, showed the disappearance of myoclonic jerks and normalisation of sleep pattern (sleep efficiency was increased by more than 5 times compared to the baseline recording, 67.5% vs 10.9%; figure 2). The clinical benefit was confirmed after 2 years of treatment at the same dosage.

DISCUSSION

We report a patient with the occurrence of restless abdomen and spinal jerks with a propriospinal-like appearance occurring during relaxed wakefulness preceding sleep onset, which caused him severe insomnia.

This case suggests that PSM-like jerks at sleep onset could represent a variant of periodic limb movement in wakefulness (LM/PLMW) in our patient, associated with an atypical RLS/WED with isolated abdominal distribution. Jerk duration, distribution of intervals between motor events and the higher prevalence in the first part of the night as well as excellent response to low dosage of dopamine agonist pramipexole support this hypothesis.

Nevertheless, given that the motor phenomenon disappeared when the patient was distracted, we cannot exclude a functional (or psychogenic) origin of the movement disorder.^{11–13}

The close temporal association between propriospinal myoclonus appearance/disappearance and the sleep–wake transition suggests that, during relaxed wakefulness preceding sleep onset, firing patterns of brainstem and forebrain ‘modulatory’ neuron populations possibly involved in controlling the motor activity of an abdomen motor generator can change. In addition, the effectiveness of dopaminergic therapy for RLS/WED (including

Figure 2 From top to bottom: a brief sequence of events separated by approximately 10–20 s intervals. Heart rate did not change in relation to motor jerks (A). The histogram of the distribution of intervals showed that events are mostly separated by intervals lasting 10–20 s (B).

this patient's abdominal phenotypic variant), PLMs and PSM indicates that a dysregulation of the complex spinal/sovraspinal dopaminergic networks could play a significant role in the pathogenesis of these motor phenomena. Therefore, we speculate that all the pathological conditions described above are primarily expressions of a dysregulation of a complex spinal/

sovraspinal dopaminergic network that could be modulated by several factors including the sleep–wake transition.

Finally, in our opinion, an organic cause should always be considered in patients with chronic severe insomnia, and video-polysomnographic investigation is very useful when a sleep motor disorder has been suspected.

Video 1 The video shows a single propriospinal myoclonus event, characterised by sudden contraction of the abdominal muscles and subsequent irradiation to adjacent muscles (spreading up and down the spinal cord).

Video 2 Immediately after propriospinal myoclonus (PSM) the patient, who is awake, seems disturbed and touches his abdomen with the left hand.

Figure 3 From top to bottom: the baseline video-polysomnography (VPSG₀) hypnogram shows an unequivocal relationship between propriospinal myoclonus (PSMs) and sleep–wake cycle (all myoclonic jerks were recorded during wakefulness), and between PSMs and body position (the myoclonic jerks were triggered by the lying-down position); the follow-up video-polysomnography (VPSG₁) hypnogram displays the complete disappearance of myoclonic jerks and the consequent normalisation of sleep pattern.

Learning points

- ▶ Restless legs syndrome Willis-Ekbom disease (RLS/WED) symptoms that are usually confined to the lower limbs can involve other body regions such as the abdomen and upper limbs.
- ▶ Dopaminergic drugs, the first-line treatment in RLS/WED, are also effective in treating abdomen RLS and propriospinal myoclonus at sleep onset.
- ▶ Changes in brain neuronal network activity at the sleep–wake transition can facilitate the appearance of motor disorder when patients attempt to fall asleep, causing insomnia.

Competing interests None.

Patient consent Obtained.

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