Propriospinal Myoclonus: Report of A Case

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Abstract- Propriospinal myoclonus is an unusual movement disorder. Here we reported a patient with electrophysiological data. The 32-year-old man had spontaneous, intermittent and brief jerks over the trunk, hip and lower limbs muscles for 4 years. The jerks were not sensitive to external stimuli and usually exacerbated whenever the patient felt relaxed, particularly preceding the onset of sleep. Electromyographic (EMG) recordings showed a long duration in each EMG burst (over 200 milliseconds) with a slow propagating pattern initiating from the rectus abdominis muscles. There was no time-locked cortical potential before the spontaneous jerks; however, we found movement-related cortical potential, while the patient was requested to mimic the jerks voluntarily. These results provided evidence to demonstrate that the jerks were not cortical origin and supported the diagnosis of propriospinal myoclonus. A comprehensive clinical evaluation including distinctive electrophysiological findings is important for the diagnosis.

Key Words: Propriospinal myoclonus, Cortical potential, Myoclonic jerk, EMG, EEG

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INTRODUCTION

The term propriospinal myoclonus was first proposed by Brown et al. in 1991[1]. It is an unusual symptom characterized by spontaneous, sometimes stimulus-sensitive muscle jerks extending over many segments with propagation in the spinal cord via slow-conducting propriospinal pathways. The underlying etiologies include disease entities of a wide range and may be idiopathic in some of the patients[2-4]. Electrophysiological investigation is important to make a correct diagnosis. Reports of propriospinal myoclonus in oriental people were scarce. Therefore we presented a case with emphasis on the electrophysiological basis.

CASE REPORT

A 32-year-old man suffered from tight and sore sensation on his right thigh about 5 years ago. Such discomfort sensation always aggravated before sleep at night. One year later, he began to notice intermittent, irregular, spontaneous and brief jerks over right lower limb. The jerks sometimes produced a violent hip flexion. The jerks progressively spreaded to both legs and the trunk during the following years; however, they were never found over the head, neck, or upper extremi-

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ties. Since the trunk flexion movement became more violent and unpredictable, he decided to quit his job. Actually the jerks rarely occurred in daytime while the patient was engaged in doing something. The frequency of the jerks significantly increased whenever he lay down in a relaxation state preceding the sleep. But no jerks developed once he was in sleep. He visited our hospital because of exacerbated trunk flexion jerks after an attack of acute gastroenteritis several days ago. Past medical history was not contributory except that he had been a carrier of hepatitis B. There was no family history of neurologic diseases. Physical examination revealed no abnormalities except the involuntary jerks over the trunk. The jerks mainly involved the abdominal wall, lower back muscles and hip joints. They were not inducible by touching or tapping stimuli. Functions of mentality, cranial nerve, muscle power, muscle tone, deep tendon reflex, sensory system, cerebellar system and extrapyramidal system were normal. Laboratory blood tests including basic biochemistry study, thyroid function and tumor markers were unremarkable.

Electrophysiological study

Multi-channel surface electromyography (SEMG) were recorded over the muscle bellies of bilateral rectus abdominis muscles, paraspinal muscles at the L4 level, and the quadriceps femoris and tibialis anterior muscles (Fig 1). The signals were amplified (Digitimer 360, Welwyn Garden City, Hertfordshire, England) with a bandpass filter of 100 Hz to 1 kHz and analyzed by Spike 2 software. Spontaneous EMG bursts were first noted in the rectus abdominis and then propagated to the back muscles and the lower extremities. Some minor attacks were only recordable over the rectus abdominis. The duration of EMG bursts always lasted for longer than 200 milliseconds.

Electroencephalographic (EEG) recordings with jerk-locked back average were conducted. We selected the EMG onset of the most active muscle, i.e. left rectus abdominis, as a trigger point to process EEG data. Data from three Ag/AgCl scalp electrodes (FCz, C3 and C4) were obtained for analysis. We selected the EEG epoch between 800 milliseconds (ms) before to 200 ms after the onset of EMG activities. The EEG and EMG signals were sampled by a rate of 1 kHz and band filtered (EEG: 0.05-70 Hz, EMG: 30-200 Hz. NeuroScan SynAmps, Neurosoft, Inc. Sterling, Va, USA). We did not find any time-locked cortical potential before each spontaneous jerk (Fig. 2). To investigate whether the jerks were truly involuntary, we also requested the patient to mimic the

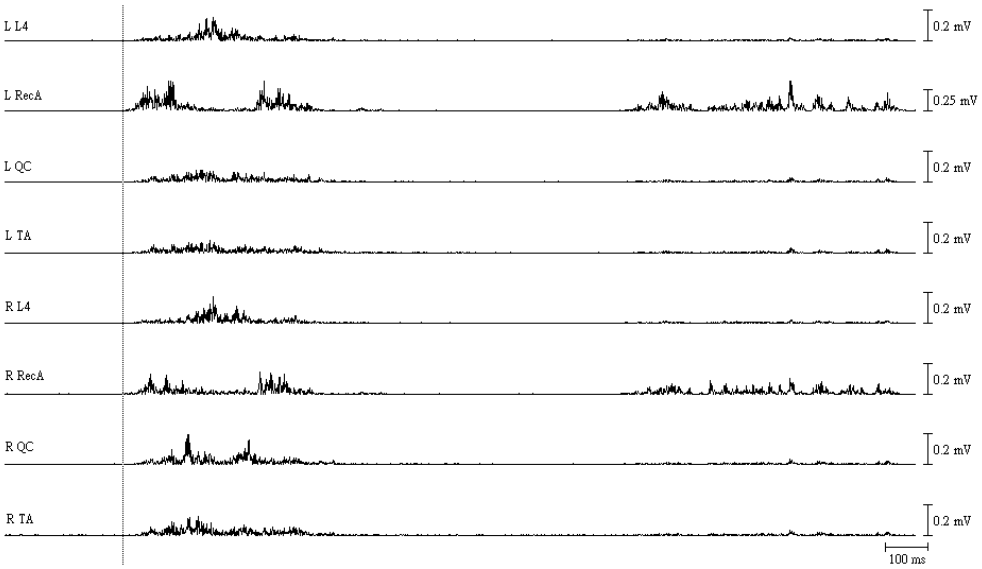

Figure 1. Multi-channel surface electromyographic (SEMG) recordings in the patient. EMG signals were first noted over the rectus abdominis muscles
The duration of EMG was longer than 200 milliseconds (ms). The intensity was not homogenous in each burst, but the most obvious changes were on the rectus abdominis muscles. (L: left; L4: paraspinal muscle at the 4th lumbar spine level; QC: quadriceps femoris; R: right; RecA: rectus abdominis; TA: tibialis anterior)

jerks as possible and recorded the movement-related cortical potential by the same trigger muscle. A rising negative potential developed at about 500 ms before the EMG onset in the electrodes of FCz and C3 (Fig. 2).

DISCUSSION

The current case is presented with an unusual and distinct form of jerks, clinically compatible with the manifestations of propriospinal myoclonus[1,5,6]. The EMG signals spreaded from the rectus abdominis muscle caudally to involve the quadriceps femoris and tibialis anterior muscles. Since there was no involvement of cranially innervated muscles and upper limbs muscles in this patient, it is not likely that the source of generators for the myoclonus resided in the brain stem or cervical spinal cord. No jerk-locked cortical potential was noted in the current case and this supported the notion that the myoclonus is not of cortical origin. Furthermore, the longer duration of the EMG bursts provided another line of evidence to eliminate the possibility of cortical origin. We suggested that the origin of myoclonus in the current patient was in the lower thoracic spinal cord. The velocity of propagation could be calculated by the relative latencies of muscles innervated by different spinal segments and the distance between the two recording locations[2]. The conduction velocity in this case was about 10 m/sec, which is similar to the previous reports[2,5,7]. The result indicated that the myoclonus was propagating along the slow-conducting pathways. The burst on the EMG record was not always simple in the patients with propriospinal myoclonus. Sometimes it consisted of a complex pattern[6]. In the current case, there were several components during a burst on the EMG tracings. Brown et al. proposed that propriospinal myoclonus may involve the release of complex spinal pattern generators[6]. Concerning the mechanism of propriospinal myoclonus, they suggested that damage to the more rostral spinal cord, particular the cervical cord, may be an important factor to release the myoclonic generators in the thoracic level[6].

The significant exacerbation of the myoclonic symptom before the sleep in the current case was noticeable. In 1997, Montagna et al. first described that the propriospinal myoclonic jerks can arise whenever patients relaxed mentally and physically, particularly preceding the onset of sleep[8]. They furtherdiscovered that the jerks occurred when alpha activities on the EEG recordings spreaded over the scalp[9]. The same group also presented three patients with propriospinal myoclonus showing restless legs syndrome and periodic limb movements during sleep[10]. Their symptoms of restless leg and peri-

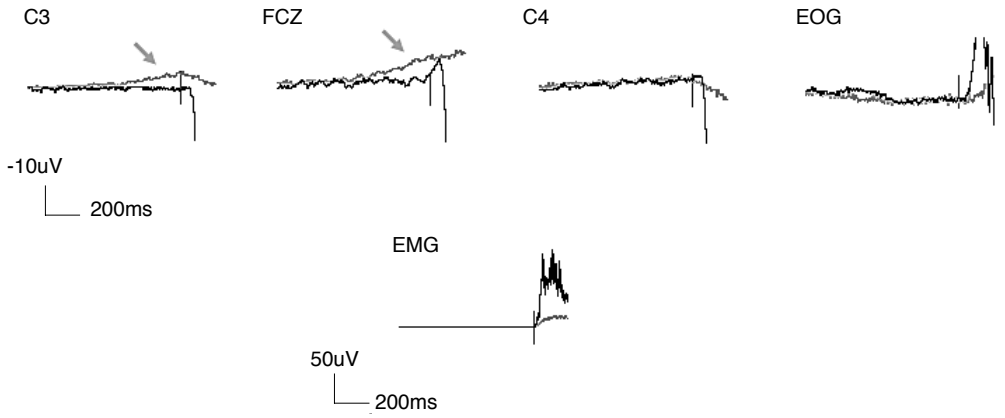

Figure 2. Recording of cortical potentials
Jerk-locked cortical potential (black line) and cortical potential related to voluntarily mimic movement (gray line) from three scalp positions (FCZ, C3 and C4). There was no pre-movement cortical activity before spontaneous jerks but negative shifting potential was noted on C3 and FCZ before intended movement (gray arrows). The short vertical line marked the movement onset. Electro-oculography (EOG) did not show notable signal before movement.

odic limb movement were noted prior to the appearance of propriospinal myoclonus. The sensation of discomfort before the development of the myoclonus in our patient seemed to share a similar course. Whether propriospinal myoclonus carries a close relationship to restless leg syndrome or they were coexistent by chance in some of the patients needs further evaluation. However, it is possible that reduction of the sleep-related spinal inhibition in restless leg syndrome may cause the activation of various motor generators, including those associated with propriospinal myoclonus[10].

The myoclonic jerks observed in the current case were spontaneous and not sensitive to external stimuli. This feature basically excluded the possibilities of reticular reflex myoclonus and hyperekplexia, which were usually stimulus-sensitive. Another major concern is to differentiate the jerks from psychogenic or voluntary jerks. Based on the comparison of the jerk-locked cortical potentials and the mimic movement related cortical potentials in this patient, we suggested his jerks were involuntary rather than voluntary because the negative shifting only occurred in the later condition.

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