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Electroencephalographic activity related to palatal myoclonus in REM sleep

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Summary. Polysomnography, including electroencephalography, electromyography and electro-oculography was performed in three patients with palatal myoclonus (PM). The amplitude of the myoclonus decreased during sleep. The frequency did not change during non-REM sleep, but increased during REM sleep in two patients. Ocular myoclonus synchronized with PM disappeared during deep sleep stages in two patients and reappeared during REM sleep in one of them. In the other patient, ocular myoclonus was noted only in REM sleep, being absent even when the patient was awake. All patients showed episodic EEG activities synchronous with myoclonic jerks only in REM sleep. These episodes were noted 5–15 times throughout the night, and each episode lasting for 1–7 s. They were negative or positive waves of saw-tooth appearance which were distributed predominantly in the central region. During the episodes, the frequency of myoclonic jerks increased in two patients. Although it is known that REM sleep influences PM and ocular myoclonus, this is the first report demonstrating the electroencephalographic activity associated with PM.

Key words: Palatal myoclonus – EEG activity – REM sleep

Introduction

Palatal myoclonus (PM) is an uncommon involuntary movement characterized by rhythmical twitching of the soft palate, often associated with synchronous movements of the pharynx, larynx and other structures derived from the embryonal branchial arches. This disorder has been associated with either destruction of the cerebellar dentate nucleus, the red nucleus, the central tegmental tract or the inferior olivary nucleus (the Guillain-Mollaret triangle), and has been postulated to have a rhythm generator in the brain stem [1, 10, 11]. Occasionally, beyond this territory, the rhythm of PM spreads to eyes, facial muscles or extremities. However, no PM-related EEG phenomena have been described in awake patients – not even with the jerk-locked averaging method. In this article, the polysomnographic findings in three patients with PM are reported, which include the characteristic EEG activities related to PM during REM sleep.

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Case reports

Case 1

The patient was a 44-year-old woman with PM and progressive ataxia. There was no family history of neurological disease. At age 39 she suffered from titubation and dysarthria, which progressed slowly. She had ataxia, bilateral pyramidal signs, urinary incontinence and bilateral oculo-palato-pharyngolaryngo-diaphragmatic myoclonus. Brain computed tomography (CT) and magnetic resonance imaging (MRI) showed an atrophic brain stem and cerebellum. Spinocerebellar degeneration with palatal myoclonus was diagnosed.

Case 2

A 41-year-old man was admitted to our hospital because of slowly progressive emotional and gait disturbances. On examination, dementia, emotional disturbance, dysarthria, left facial nerve palsy, bilateral pyramidal signs, cerebellar ataxia and urinary incontinence were observed in addition to bilateral palato-facial-pharyngo-laryngeal myoclonus. MRI disclosed atrophy of the brain stem and cerebellum. Since uveitis and oral and genital ulcers were noted, neuro-Behçet disease was diagnosed.

Case 3

A 50-year-old woman lost consciousness for several hours following the development of tetraparesis and dysarthria. CT revealed bleeding in the tegmentum pontis. Twelve months after the attack she showed bilateral abduction paralysis, dysarthria, left hemiparesis, left hemi-hypaesthesia, ataxia in the left limbs and oculo-palato-pharyngo-laryngo-diaphragmatic myoclonus.

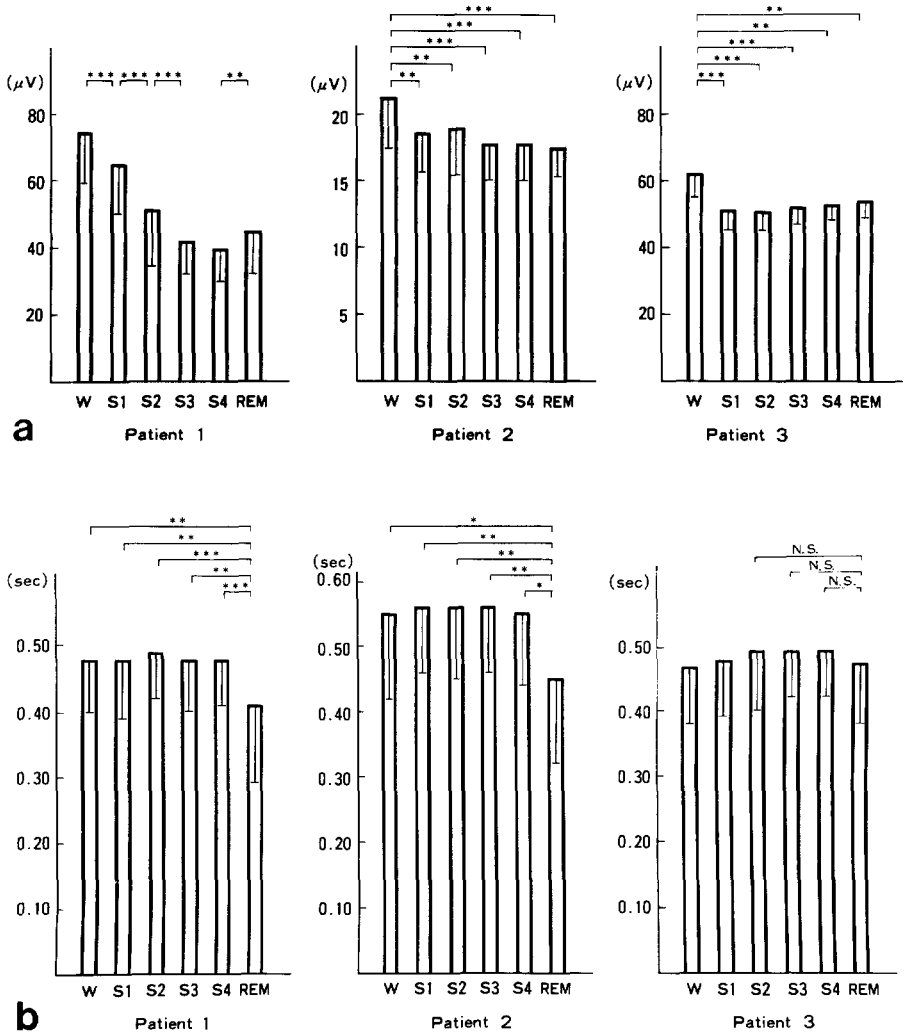

Fig. 1. Mean and standard deviation of the amplitude (**a**) and interval (**b**) of myoclonus in various stages of sleep. *, $P<0.05$; **, $P<0.01$; ***, $P<0.001$, Duncan's multiple range test

Methods

Standard polysomnographic sleep recordings were carried out using electroencephalography (EEG), electro-oculography (EOG), and submental electromyography (EMG). EEG was recorded from monopolar and bipolar leads of electrodes situated in the frontal, central and occipital regions. In an attempt to record PM, surface EMG of suprahyoidal laryngeal muscle was recorded in cases 1 and 3, and that of facial muscle (right levator angular oral muscle) in case 2. By other polygraphy examinations it was confirmed that the EMG of laryngeal or facial muscle was completely synchronized with the surface EMG recorded directly from the palate when the patients were awake. Sleep stages were scored by standard criteria [7]. Sixty random samples of myoclonus, each of which lasted for 10 s, were analysed as to amplitude and interval when the patients were awake and in each sleep stage.

Results

Polysomnographic recordings showed that PM continued during sleep in all patients. The amplitudes and intervals of myoclonic jerks when awake and in each sleep stage are shown in Fig. 1a and b. The amplitude in sleep for each patient was reduced compared with that in wakefulness. During sleep, the amplitudes in stages 3 and 4 were decreased from those of sleep stages 1 and 2 in case 1, but not in cases 2 or 3. On the other hand, there were no significant differences in the intervals of PM among sleep stages except for REM sleep. A decrease in interval was recognized between REM and other sleep stages in cases 1 and 2. In case 1, myoclonic jerks became slightly irregular in REM sleep, which was demonstrated by the finding that the standard deviation of their intervals in REM sleep was larger than those in other sleep stages.

Ocular myoclonus synchronized with PM in cases 1 and 3, decreased in amplitude during sleep stages 1 and 2, and disappeared in deep sleep stages. In case 1, it reappeared during REM sleep. In Case 2, it was observed paroxysmally only in REM sleep. In case 1, ocular myoclonus disappeared on treatment with 12 mg trihexyphenidyl, with no change in PM.

All patients showed paroxysmal EEG activities related to myoclonic jerks only in REM sleep (Figs. 2, 3). They were negative or positive waves with a saw-tooth form, which were distributed predominantly in the central region. On bipolar leads there was phase-reversal of these waves in the central region (Fig. 3). The frequency of these waves was 2–3 Hz and their amplitude was 20–100 μV. These episodes were noted 5–15 times through-

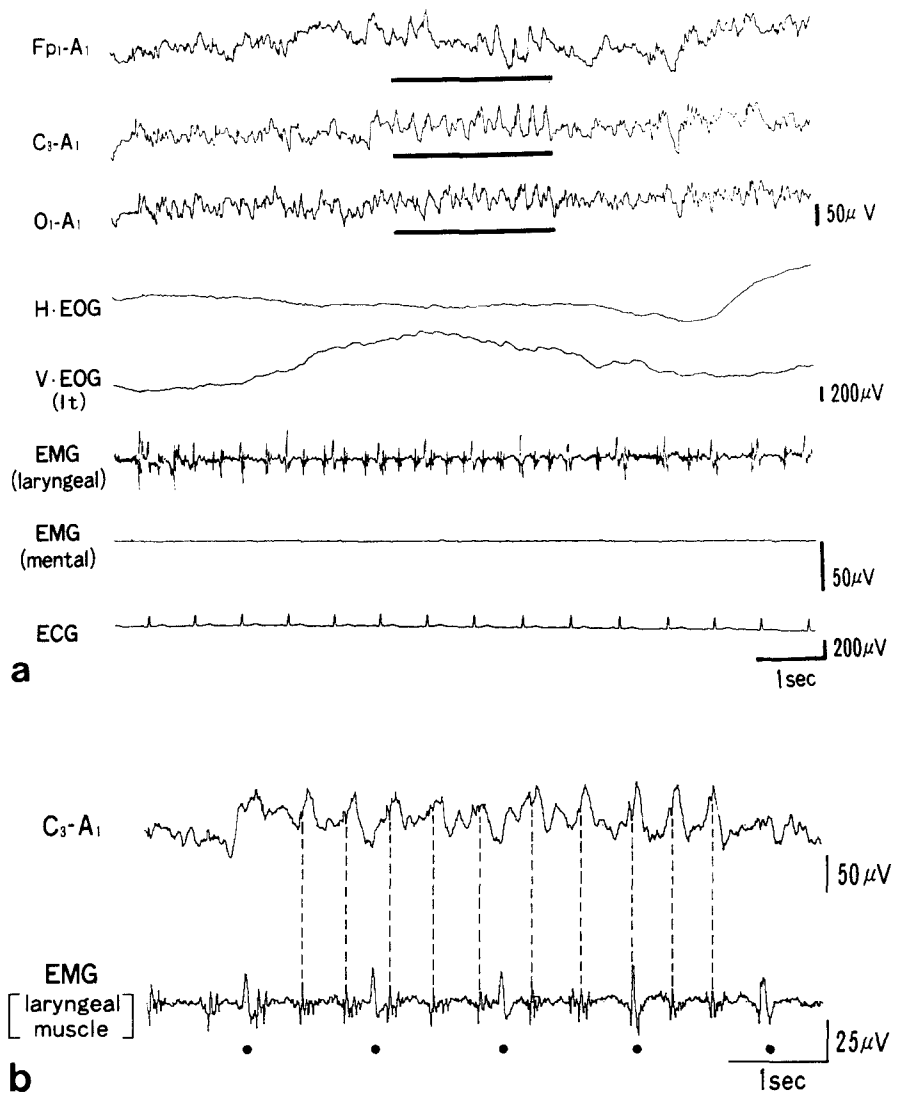

Fig. 2. a Polygraphy in case 1 during REM sleep. Palatal myoclonus (PM)-related EEG was observed *(underlined)*. Ocular myoclonus disappeared after treatment with trihexyphenidyl. **b** Magnification of EEG(C_3–A_1) and EMG of laryngeal muscle. The negative waves were synchronized with myoclonic jerks. *Closed circels* indicate electrocardiac activity

Table 1. Intervals of myoclonus during REM sleep and paroxysmal EEG activity related to myoclonus

	During REM sleep	During paroxysmal EEG activity related to myoclonus
Case 1	0.42 ± 0.14 s	0.32 ± 0.10* s
Case 2	0.46 ± 0.16 s	0.32 ± 0.12* s
Case 3	0.47 ± 0.09 s	0.46 ± 0.10 s
		(mean ± standard deviation)

* $P < 0.001$; Student's test

out the night, each episode lasting for 1–7 s. They were almost completely synchronized with myoclonic jerks. During the episodes, the frequency of myoclonus increased in cases 1 and 2 (Table 1). In case 1, the baselines of EOG records shifted during the episodes. The characteristics of these waves in case 1 did not change after the ocular myoclonus had disappeared following the treatment.

Discussion

Since Nathanson [6] first confirmed the persistence of PM during sleep by visual observation and palpation of the throat, many authors have reported that PM continued during both natural and induced sleep except in a very few cases [3, 13]. This is considered to be one of the important and characteristic features of PM, which is different from tremor, athetosis and many other involuntary movements. However, there have been few reports of polygraphic studies of PM throughout an entire night [4, 10, 12]. Yap et al. [12] first demonstrated myoclonus in all stages of sleep by polysomnography. They observed that myoclonus became irregular in amplitude and frequency during deep sleep stages, and that ocular myoclonus disappeared in all stages of sleep. Kayed et al. [4] observed, in three patients, that PM occurred in the form of clusters of two or four high-amplitude movements at rather variable intervals during REM sleep. Tahmoush et al. [10] quantitatively analysed the amplitude and interval of PM in two patients and found that the ampli-

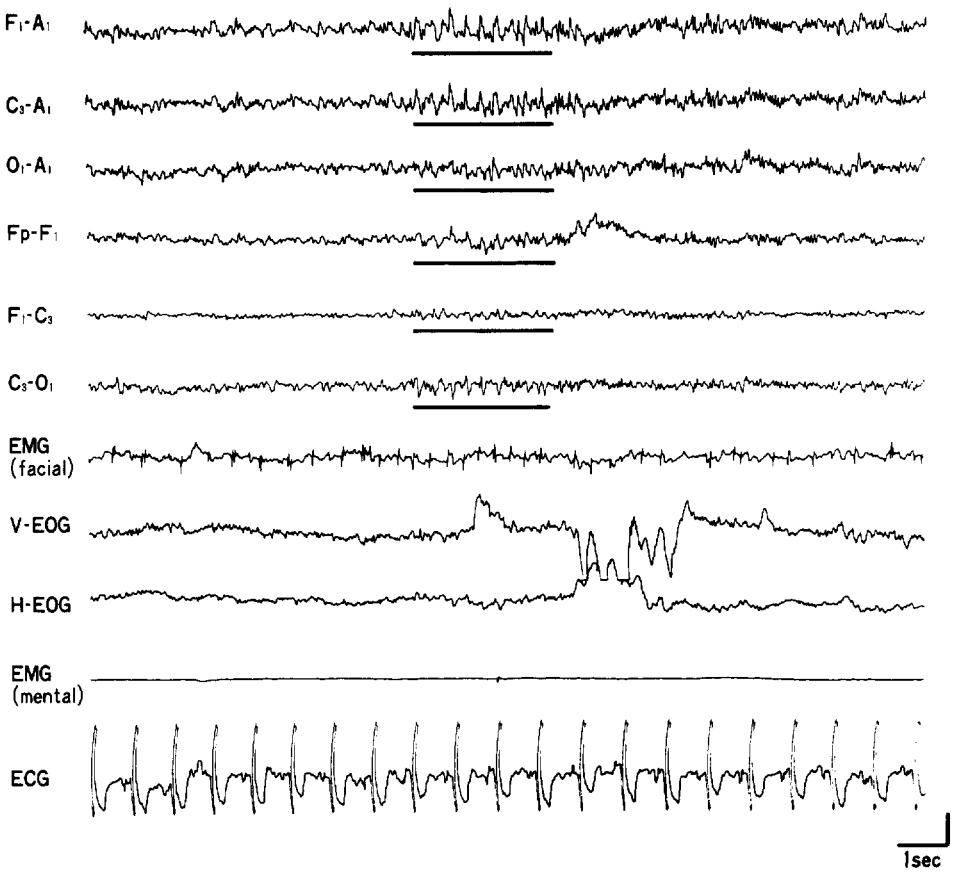

Fig. 3. Polygraphy in case 2 during REM sleep. PM-related EEG was observed *(underlined)*

tude of PM was reduced with fall of amplitude of background EMG, while the interval did not change between sleep stages. Fluctuation of the amplitude and frequency of PM during deep or REM sleep was not noted. Ocular myoclonus was absent during non-REM sleep in two patients, but reappeared in REM sleep in one of them.

Also in our patients the amplitude decreased when the patients fell asleep. The amplitude decreased further as the sleep became deep in case 1, but this was not apparent in cases 2 or 3. Although there was no significant difference in the frequency of myoclonus among non-REM sleep stages, an increase in frequency was noted in REM sleep in cases 1 and 2. Also, slight fluctuation of the interval of myoclonus during REM sleep was observed in case 1, but not in the other cases. Our study and other reports have indicated that PM principally decreases in amplitude during sleep, but does not change in frequency. During REM sleep, however, the frequency of PM increased and the interval fluctuated in some patients.

Ocular myoclonus in our cases 1 and 3 disappeared during deep sleep stages but was present in sleep stages 1 and 2. In case 1, ocular myoclonus reappeared during REM sleep, as seen in the patient of Tahmoush et al. [10]. Furthermore, in case 3, ocular myoclonus was observed only in REM sleep, although it was absent even when the patient was awake. These facts suggest that, in some patients with PM, REM sleep influences the palato-ocular synchrony in a way different from non-REM sleep.

We found in all patients that the EEG activity was related to myoclonic jerks only in REM sleep. It was differentiated from artefacts due to ocular myoclonus in that it was noted even when ocular myoclonus disappeared. It was not EMG activity because it was predominant in the central region, and the wave form was different from artefacts of EMG. Therefore, we considered that these waves indicated EEG activity and call it here PM-related EEG. Saw-tooth waves, ponto-geniculo-occipital waves and occipital spikes are known as characteristic EEGs in REM sleep. PM-related EEG was similar to saw-tooth waves in frequency, amplitude, duration, waveform and distribution [2, 8, 9].

It is well known that the causative lesion of PM lies in the brain stem or cerebellum. Since myoclonus appeared mainly in the oropharyngeal muscles, Trelles [11] postulated that PM was a result of the release of the nucleus ambiguus from the normally inhibitory action of the dentato-olivary system. Bender et al. [1] demonstrated that PM was elicited by electrical stimulation of the reticular formation just dorsal and medial to the inferior olivary nucleus. In addition, the fact that rhythmic movements of eyes, facial muscle, tongue and extremities associated with PM are synchronous with PM might suggest that there is a rhythm generator of PM in the brain stem. Nagaoka and Narabayashi [5] found that the rhythm of PM caudally influenced the motoneurons in lumbar segments on studying the H-reflex of soleus muscle. On the other hand, rostrally, the oculomotor nucleus is often influenced; ocular myoclonus is a common finding, as seen

in our patients. However, EEG activity associated with PM has not been reported.

The frequency of PM increased during the PM-related EEG in two patients. In case 1, the baseline of EOG records shifted a few minutes before PM-related EEG, which might be due to a galvanic skin reflex of the forehead. These phenomena suggested some peculiar involvement the autonomic nervous system during PM-related EEG. The mechanism underlying PM-related EEG is unclear. One possibility is that the activity of the rhythm generator of PM in the brain stem influenced the EEG either directly or through an effect on the sleep phenomena. Another possibility is that during the REM period in which saw-tooth activity was seen, there was particular release of the myoclonic jerks in a time-locked fashion.

Previous reports demonstrated that, in some patients with PM, the rhythm or amplitude of PM is influenced by REM sleep. In addition, our study demonstrates that there is EEG activity associated with PM in REM sleep.

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