Short report

Somatosensory and acoustic brain stem reflex myoclonus

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SUMMARY A patient with brain stem reflex myoclonus due to a massive midbrain infarct was studied electrophysiologically. Myoclonic jerks were elicited at variable latencies by tapping anywhere on the body or by acoustic stimuli, and mainly involved flexor muscles of upper extremities. The existence of convergence of somatosensory and acoustic inputs in the brain stem was suggested. This myoclonus seemed to be mediated by a mechanism similar to the spino-bulbo-spinal reflex.

Myoclonus is frequently sensitive to stimulus of single or multiple modalities. This reflex myoclonus is broadly classified into two types depending on the possible reflex centre. Pyramidal myoclonus¹ or cortical reflex myoclonus² is electrophysiologically characterised by an enhanced cortical response to somatosensory stimulation and by the presence of cortical spike preceding the myoclonic jerk.²⁻⁴ This type of myoclonus is commonly encountered in patients with progressive myoclonic epilepsy including lipidosis.^{3,4} The second type is the so-called reticular reflex myoclonus, characterised by involvement of all muscles including proximal ones and by the upward activation of cranial nerves starting from the medulla.⁵ There are EEG spikes but they are not time-locked to myoclonus, and cortical responses to stimuli are not enhanced. The latter type of myoclonus has been commonly reported in patients with post-hypoxic myoclonus,^{5,6} but its pathophysiological mechanism has not been fully understood.

We report a patient with massive midbrain infarct

manifesting myoclonic jerks which were easily elicited by tapping any part of the body or acoustic stimulation; the relationship with reticular reflex myoclonus as well as with spino-bulbo-spinal reflex⁷ will be discussed.

Case history

A 65 year old man suddenly developed nausea and vomiting, vertigo, left facial numbness, speech disturbance and headache. He became unconscious on the following day, and showed bilateral miosis, loss of the left corneal reflex, left peripheral facial palsy and quadriplegia. Cranial CT scan revealed a low density area involving almost the whole aspect of the midbrain and extending down to the pons on the left. Electroencephalogram (EEG) showed a poorly organised background activity with theta waves as a dominant rhythm and occasional bifrontal delta waves. There were no spikes. Flash or auditory stimuli did not cause any alteration of the EEG.

At the time of investigation a month and a half after the onset, he manifested akinetic mutism, decerebrate posture, reactive small pupils, bilateral partial oculomotor palsy, left trigeminal and facial nerve palsy, torticollis to the right, and quadriplegia more on the left with increased deep reflexes and positive Babinski reflex bilaterally. Myoclonic jerks were elicited mainly in the flexor muscles of upper extremities by either tapping any part of the body or by acoustic stimuli such as clapping hands. There was no spontaneous myoclonus, and other stimuli such as soft touch, pin prick, sudden passive flexion or extension of joints, or flash failed to induce any myoclonic jerk.

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Methods

Myoclonus was elicited by tapping the body with a tendon hammer which was equipped with a micro-switch in order to obtain a trigger pulse. There was a time lag of 8 to 10 ms from the onset of hammer contact with a metal plate to the time when the pulse was actually generated. For acoustic stimulation, tone bursts or clicks of 1,000 Hz were generated by an auditory stimulator and presented binaurally through earphones at variable stimulus rates. In order to study a possible interaction of somatosensory and acoustic stimuli, tapping on the chin and tone burst were alternately given at an overall stimulus rate of twice a second.

Electromyograms (EMGs) associated with myoclonic jerks were recorded by placing a pair of disc electrodes 1 to 3 cm apart over each muscle. The filter setting of amplifiers was 75 to 1,000 Hz. EEGs and EMGs were simultaneously recorded, but the technique of jerk-locked back averaging⁸ could not be employed owing to the absence of spontaneous myoclonus.

Somatosensory evoked potentials (SEPs) were recorded by stimulating each median nerve at the wrist by electric shocks of 0.2 ms duration and strength 10% above the motor threshold at variable stimulus rates. EEGs were recorded from disc electrodes placed at C3, Cz and C4 (International 10–20 system) referred to the left ear lobe for the first two electrodes and to the right earlobe for the C4 electrode. Electrodes were also placed over the 7th cervical spinous process and at each Erb's point, both being referred to Fz electrode. The filter setting of the EEG amplifiers was 1 to 1,000 Hz. EMGs were simultaneously recorded from biceps brachii muscles bilaterally and were averaged along with the EEGs by using the stimulus pulse as a trigger.

Auditory evoked potentials (AEPs) were recorded by giving tone bursts of 1,000 Hz through earphones and by recording EEGs from electrodes placed at T3 and T4 both referred to Fz. Photic evoked potentials (PEPs) were recorded by giving flash stimuli from strobe lamp at a stimulus rate of once every second. Electrodes were placed at O1 and O2 both referred to Cz electrode. For both acoustic and photic stimulation, EMGs were simultaneously recorded from multiple muscles.

Results

SEPs following electrical stimulation of the right as well as left median nerve were normal at the electrodes placed at each Erb's point (N9) and over the 7th cervical spinous process (N13), but cortical potentials were markedly abnormal. Following the right median nerve stimulation, there was a small potential of very long duration at the C3 electrode, its peak occurring 31.8 ms after the stimulus (fig. a). No other cortical potentials were identified. Following the left median nerve stimulation, there was a positive peak at all scalp electrodes at a latency of 14.8 ms, but no other potentials were recognised. Electrical stimulation did not cause myoclonus. PEPs of normal wave form and normal latency were recorded from the

occipital electrodes, but no myoclonic jerks were elicited.

Gentle tapping anywhere on the face, trunk or extremities elicited myoclonic jerks predominantly in the flexor muscles of the right upper extremity. Myoclonic jerks induced by strong tapping were more vigorous and involved more muscles throughout the body, including extensors of extremities. The onset latency of the reflex myoclonus recorded from the right biceps brachii muscle was 22.4 to 36.8 ms (median value 29.6 ms) following the chin tap (actually following the pulse generated by the hammer micro-switch), 27.2 to 46.8 ms (median value 37.0 ms) following the left wrist tap and 36.0 to 62.4 ms

Fig (a) Cortical and short latency SEPs following electrical stimulation of the right median nerve at wrist. A1 and A2; left and right ear lobe, respectively. Average of 200 sweeps. A negative peak at the right clavicle (C1, Erb's point) relative to Fz (N9) and another negative peak at Cv7 (7th cervical vertebra) relative to Fz (N13) are normally recorded, but cortical response consists only of a small negative potential of long duration at C3, its peak latency being 31.8 ms (arrow). No myoclonic response was elicited in EMG (bottom 2 channels). A response starting at a latency of 5.0 ms in the right biceps lead reflects a propagating action potential elicited in the median nerve. (b) Click-induced myoclonic discharge in the right biceps brachii muscle. Single sweep. Comparison of 2 different rates of stimulus presentation. There are recruiting responses with the rate of once every 500 ms (top), while responses are only occasionally seen with the rate of once every second (bottom).

(median value 49.2 ms) following the right ankle tap. The latency was variable from stimulus to stimulus even within the same stimulus-recording setting. The duration of the EMG discharge associated with a reflex myoclonus was 50 to 60 ms.

Tone burst elicited myoclonic jerks in facial and upper extremity muscles synchronously on both sides. The latency of myoclonic jerks elicited in the biceps brachii muscles by tone burst was variable from stimulus to stimulus, ranging from 25.2 to 39.2 ms (median value 32.2 ms), but the latency difference of each myoclonus between the left and right biceps muscles was small (0.8 to 4.8 ms). The duration of the evoked myoclonic EMG discharge was 50 to 70 ms. There was no recognisable EEG response to tone burst. Auditory stimulation by clicks elicited myoclonus mainly in the right biceps brachii muscle, and its latency ranged from 32.6 to 75.8 ms (median value 54.2 ms).

Occurrence of the reflex myoclonus was significantly influenced by the rate of stimulus presentation both for somatosensory and acoustic stimuli. When click stimuli were given once every 250 to 500 ms, myoclonic jerks of increasing amplitude as well as of increasing duration were elicited (fig. b). Myoclonic jerks were only occasionally elicited when the same stimuli were given at a lower stimulus rate (fig. b), and myoclonic jerks were seldom elicited at a stimulus rate higher than once every 250 ms. The same phenomenon was observed for tap stimulation. When tone burst and the chin tap were alternately presented at an overall stimulus rate of once every 500 ms, myoclonic jerks were recruited as if stimulus of a single modality was presented at the same stimulus rate.

Discussion

Myoclonic jerks observed in the present case were elicited only by somatosensory and acoustic stimuli, and not by photic stimuli. Cortical SEPs were nearly absent while subcortical responses were present. Photic stimulation evoked normal cortical responses but no myoclonic jerks. Taking into account the clinical evidence of decerebration and the radiological evidence of massive infarct involving nearly the whole aspect of the midbrain, this myoclonus is best explained as a reflex mediated by structures caudal to the midbrain.

Moreover, alternate presentation of somatosensory and acoustic stimuli at an overall stimulus rate of once every 500 ms produced recruiting myoclonic responses as if either one of the two stimuli was given at the same rate. Since the stimulus presentation of a single modality, whether somatosensory or acoustic, at a rate of once every second did not produce

recruitment of myoclonic responses, this finding suggests the existence of some convergence of inputs from the two stimuli of different modalities. It is reasonable to postulate that the convergence has taken place in the brain stem.

One of the characteristics of this reflex myoclonus seems to be a significant variability of the latency. The variable latencies of tap-induced myoclonic jerks might be due to variabilities in the strength and speed of hammer tapping itself or in the time jitter between actual onset of tapping and pulse generation. Tone burst- or click-induced myoclonic jerks, however, also were variable in their latency from stimulus to stimulus in spite of the constant stimulus condition. This feature has not been reported in cortical reflex myoclonus.

Myoclonic jerks in the present case appear to be similar to the reticular reflex myoclonus described by Hallett *et al*⁵ in that the jerks mainly involved proximal muscles bilaterally. The latency jitter of evoked myoclonus has also been described in reticular reflex myoclonus.⁵ There are, however, clear differences between the two. Patients with reticular reflex myoclonus have been reported to have EEG spikes although those spikes are not time-locked to spontaneous myoclonic jerks.⁵ The present case did not show any spontaneous myoclonus, and nor were there EEG spikes.

Shimamura and Livingston⁷ in 1963 recorded responses from ventral roots at various levels in decerebrated cats after electric stimulation of dorsal roots or peripheral nerves. The reflex was shown to involve a relay in the bulbar reticular formation and re-entry into the spinal cord from above downward, and was called the spino-bulbo-spinal (SBS) reflex. The pathway was shown to conduct impulses rapidly during ascent and relatively more slowly during descent. Shimamura and Yamauchi⁹ further showed that, in cats under chloralose anaesthesia, a single tactile stimulation to the skin elicited a late response corresponding to the SBS reflex, and that jerky muscle reaction seen in those cats may be related to the SBS reflex. On weak stimulation, the late response or the muscle jerks mainly involved flexor motor nerves. While the SBS reflex is usually difficult to be recorded in normal man,¹⁰ Shimamura¹¹ proposed a similar mechanism to explain startle response seen in children with cerebral palsy. Those children showed a generalised jerky flexion following click and tactile stimuli. Recording of the jerky response by the surface EMGs from various muscles demonstrated sequential involvement from the rostral muscles to the caudal ones. The latency was about 25 ms longer for the EMG response following tactile stimulation to foot than for the click-induced response. Myoclonic responses observed in the present case involved pre-

dominantly flexor muscles of upper extremities, and weak stimulation elicited myoclonic responses almost exclusively in the flexor muscles. The latency of the evoked myoclonic jerk was 17 ms on average longer for tapping the ankle than for the tone burst stimulus.

Conduction velocity of ascending and descending impulses of the reflex pathway in the present case could not accurately be obtained owing to a considerable variability of latencies. On average, however, the difference of 12.2 ms between the latency of the response recorded from an arm muscle following the wrist tap and that recorded from the same muscle following the ankle tap suggests that the ascending pathway is rapidly conducting. Among a variety of somatosensory stimuli, only tapping was effective for eliciting myoclonic responses; pin prick, soft touch, sudden joint displacement or electric stimulation of the peripheral nerves was not effective. These observations seem to be in conformity to the SBS reflex which is predominantly influenced by impulses arising from cutaneous sources.^{7 9 11}

The efferent conduction velocity was even more difficult to estimate, because the myoclonus was not elicited in leg muscles. However, the relatively long latency of the myoclonus elicited in the arm muscle following the chin tap (29.6 ms) as well as following the tone burst (32.2 ms) seems to suggest the slowly conducting efferent pathway found in the SBS reflex.

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