Spinal myoclonus

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SYNOPSIS A case of rhythmic myoclonus affecting only the lower part of the body is described. This occurred as an acute self-limiting illness. The changes in the cerebrospinal fluid (CSF) suggested a viral infection. Clinical and electrophysiological findings indicated that the involuntary movements were arising at spinal level and were independent of suprasegmental influences. There are few previously reported cases of spinal myoclonus, all different in various respects from the present one, which is reminiscent of the results of experimental inoculation of virus into feline spinal cord.

Myoclonus may be defined as a sudden jerk which occurs in a muscle or in part of a muscle, and which is not under voluntary control. Myoclonic jerks may be caused by lesions at various levels of the nervous system, but evidence that clearly shows that myoclonus arises at the spinal level is sparse. We present a case of acute spinal myoclonus with virtually complete recovery. Electrophysiological studies helped confirm that these movements were of spinal origin.

CASE REPORT

(St.B.H. 510289) A previously healthy barrister aged 36 years returned to England from a holiday in Greece. A few hours after his return he noticed a feeling of heaviness of the legs and difficulty in walking. Shortly after, his legs began to twitch, and he noticed some hesitancy of micturition. He attended a casualty department, where a diagnosis of hysteria was made and he was sent home. The twitching of his legs became continuous and more violent, and he was unable to sleep at all that night, although he did not feel ill. He was admitted next morning to St Bartholomew's Hospital.

On examination, he was a fit-looking man, without fever or neck stiffness. The cranial nerves and arms were normal. There were continuous rhythmic thrusting movements of the pelvis and repetitive jerks of the legs. These sometimes were bilaterally synchronous. Their frequency was about 100 to 150 times per minute, and their amplitude sufficiently violent to shake the bed. Because of the transmitted movement it was difficult to be sure of the upper limit of muscles involved by these movements, but it appeared that all muscles above segmental level T12 were spared. Full voluntary power of the legs was retained. The jerks were not modified by voluntary movement, and they continued unabated during sleep. Examination of other systems was normal.

He was treated with diazepam 30 mg a day. After four days the jerks were less violent and no longer continuous. He was able to walk without obvious

FIG. 1. Muscle action potentials recorded from left gastrocnemius evoked by stimulating the left medial popliteal nerve (a). The evoked responses from left gastrocnemius on constant stimulation of the right medial popliteal nerve are shown in b, c, and d.
myoclonus and he could lie in bed without jerks for increasingly long periods. He could, however, still bring on bouts of myoclonus by adopting certain postures, particularly by turning on to his side with legs and knees flexed. Myoclonus was also precipitated by vibration induced by tapping the clavicle, or even by tapping the bed. Loud sounds had no effect. Tapping the tendon of a muscle of the lower limb elicited a contraction not only in that muscle, but in other muscles of both lower limbs. Tapping the Achilles tendon resulted in a simultaneous contraction of both tibialis anterior and triceps surae, but the net result was always dorsiflexion of the foot.

INVESTIGATIONS The haemoglobin concentration, white cell count, and differential count were normal. The ESR was 23 mm in the first hour, and radiographs of the chest were negative. Four days after the first symptoms the cerebrospinal fluid (CSF), under normal pressure, contained 57 lymphocytes and 5 polymorphs per mm$^3$; protein 107 mg/100 ml, IgG 7.0 mg/100 ml; sugar 69 mg/100 ml. Bacterial culture was sterile. The CSF was examined again on the ninth day of the illness. On this occasion there were 58 red cells, 40 lymphocytes, and 3 polymorphs per mm$^3$. The protein had fallen to 56 mg/100 ml, IgG 6.6 mg. On the sixth day, complement fixing antibodies to Herpes simplex (1:256), H. zoster (1:1024), and measles (1:64) were found in the blood, but no antibodies to these viruses were present in the CSF. Enteroviruses were not isolated from monkey kidney tissue culture inoculated with stools and CSF. Stools were inoculated into suckling mice in a search for Coxsackie virus, with negative results.

Electrophysiological studies were performed five days after the onset of the illness, at a time when the jerks were no longer continuous. Stimulating cathodes were placed over both medial popliteal nerves, the anodes being over the patellae. Surface recording electrodes were placed over the left gastrocnemius muscle. At a stimulus intensity insufficient to cause a direct (M) response in the left gastrocnemius muscle, widespread myoclonic jerks in both lower limbs appeared in response to stimulating the left medial popliteal nerve. These jerks continued for 1–2 s. The initial negative deflection shown in Fig. 1a had a latency of about 43 ms. When the right popliteal nerve was stimulated at an intensity insufficient to cause a twitch of the right gastrocnemius at intervals of about 10 s, the initial negative response from left gastrocnemius also had a latency of about 40 ms. The shape and amplitude of the potential evoked in the left gastrocnemius varied in shape and amplitude (Fig. 1b, c, d), even though the stimulus was con-

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**FIG. 2.** Repetitive firing in left gastrocnemius in response to a single shock to the right medial popliteal nerve.

**FIG. 3.** (a) A normal Hoffmann response in left gastrocnemius after clinical recovery. In b, c, and d note absence of response on stimulation of contralateral medial popliteal nerve.
stant. On a different sweep speed (Fig. 2) repetitive
synchronous firing of many motor units can be seen
at intervals of 40–50 ms. Examination of the left
gastrocnemius and left tibialis anterior at rest with a
concentric needle electrode showed occasional
fasciculation potentials, but was otherwise normal.
When seen three weeks after the onset of the illness,
no myoclonus could be induced, and both knee and
ankle tendon jerks were absent, although the arm
tendon jerks remained normal. The left ankle jerk
had returned two weeks after this. At this time an
apparently normal H reflex with a latency of 30 ms
could be obtained from the left gastrocnemius
muscle on stimulating the left medial popliteal nerve
(Fig. 3a) but there was no response in the left
gastrocnemius to stimulation of the right medial
popliteal nerve (Fig. 3b). Occasional fasciculation
potentials were found in the left gastrocnemius.

DISCUSSION

An early demonstration that involuntary move-
ments might arise in the spinal cord was that of
Turtschaninow (1894), who produced jerks in the
legs of dogs by intravenous injection of 5% phenol.
Since the jerks were not abolished by transection of the
brain-stem or spinal cord although eliminated by section of the peripheral
nerves, they were clearly arising in the spinal
cord. Other relevant animal experiments are those of Luttrell et al. (1959), who injected the
virus of Newcastle disease into the spinal cord of
cats. Bilateral synchronous jerks developed in the
muscles supplied by the injected segments, and remained unaffected by surgical isolation
of these segments.

The possibility of a viral infection was con-
sidered by Campbell and Garland (1956) who
reported three patients who suffered a rapidly
fatal illness, characterized by myoclonic jerks of
the lower limbs. Histological changes strongly
suggestive of a viral infection were found, almost
exclusively confined to the spinal cord. The main
changes were lymphocytic cuffing and microglial
nodules. These authors also mention that
damage to the anterior horn cells was incon-
spicuous. Their cases differ from the present one
in that the patients were all extremely ill, with
fever, sweating, and severe pain from the muscu-
lar spasms. The general condition of our patient
never gave cause for alarm, and pain was not a
feature. Two of their patients had received
recent radiotherapy to the spine, and they
speculated on the role this might have played in
predisposing to viral infections. Activation of a
latent virus by irradiation would now also be
considered a possibility.

Other cases of spinal myoclonus described in
the literature also bear little resemblance to the
present case. Silfverskiöld (1962) collected three
cases all of which began in the autumn of 1958 in
the area of Stockholm, suggesting infection as
the causal agent, but no such agent could be
demonstrated. All were children who had rhyth-
mic jerks of some muscles of arms and legs which
did not interfere with voluntary action. These
movements persisted for up to 18 months.
Electromyography in one case demonstrated that
brief jerks affected different muscles in the same
limb synchronously, though they were seldom
simultaneous in two extremities. However, it
appears that the involuntary movements were
not entirely independent of supraspinal control
as they were increased by emotion and decreased
both during voluntary use of the limb and during
mental arithmetic; they also disappeared during
sleep. Swanson et al. (1962) reported two
patients with localized involuntary movements
considered to be myoclonic, of a benign nature
in both; one had lifelong jerks of the left arm and
diaphragm, the other had a 10 month history of twitching of the muscles of the left leg.
In both cases, these movements were no more
than a nuisance. Electromyography showed
brief asynchronous discharges, sometimes only
in parts of individual muscles.

A convincing case of myoclonus of purely
spinal origin was described by Garcin et al.
(1968). Myoclonus of the right upper arm was
produced by an astrocytoma of the spinal cord,
extending between the C3 and C5 vertebral
bodies. The frequency of jerks was 43 per
minute, and they were synchronous in all the
muscles affected. The amplitude was only
slightly modified by passive movement of the
limb, but voluntary contraction of any of the
affected muscles temporarily abolished its myo-
clonus. Passive stretch of the muscle had no
effect. The myoclonus was continuous during
sleep. After surgical removal of the tumour, the
jerks changed in character and were replaced by
irregular asynchronous contractions of biceps
and supinator longus.

In the present case, there was good evidence
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for purely spinal disease. The jerks were confined to the lower part of the trunk and legs. Many muscles were affected synchronously and fairly symmetrically between the two sides. The movements continued during sleep, and they were not exacerbated by loud noises or voluntary movements. When spontaneous myoclonus had ceased, manoeuvres which directly or indirectly stretched the muscle spindles, such as tapping tendons or the skeleton, or knocking the bed, were effective in eliciting myoclonic jerks. Furthermore, it was demonstrated that stimulation of the medial popliteal nerve induced myoclonic jerks in both ipsilateral and contralateral legs with almost identical latencies of about 40 ms. This is a little longer than the 37 ms found by Marsden et al. (1973) for the latency of the ankle jerk in a normal subject, but considerably shorter than the 75 ms latency they found for the tonic stretch reflex in the long flexor of the big toe, which they suggest involves a loop via fast conducting fibres through the cerebral cortex. Mayer and Mawdsley (1965) reported a mean latency of 29 ms for the H reflex recorded from the mid-calf in young adults (range 26–32 ms). The latency of the myoclonic jerks in our patient induced by stimulation of the medial popliteal nerve thus suggests conduction through a spinal pathway with more than one synapse, the figure of 40 ms being too long for a monosynaptic route, and probably too short for any pathway extending up to supraspinal levels. The absence of clinical weakness and of denervation potentials in the present case, and the subsequent disappearance of the tendon reflexes in the affected segments suggests that the predominant lesion lay in the intercalated neurones of the dorsal horn, the anterior horn cells remaining largely intact.

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