Segmental Spinal Myoclonus and Syringomyelia :
A Case Report

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Abstract

We report the case of a 75-year-old woman who developed involuntary jerks of the abdominal musculature. They occurred spontaneously or triggered by a forced inspiration or attempts to rise from the supine position. Electromyography (EMG) recorded abnormal bursts of muscle activity in the abdominal, thoracic paraspinal, and intercostal muscles up to the 3rd intercostal space. The bursts were bilateral, arrythmic and synchronous in all muscles. Magnetic resonance imaging (MRI) of the spine revealed a syringomyelic cavity between the T3 and T10 levels. The topological correlation between the EMG muscle activities and the MRI findings was consistent with spinal myoclonus arising from the thoracic spinal cord. The synchronous bursts in muscles depending from few adjacent spinal segments suggested the diagnosis of segmental spinal myoclonus (SSM). There are few reports of SSM related to syringomyelia in the literature.

Key words : Syringomyelia ; spinal myoclonus ; segmental spinal myoclonus.

About forty years after the first description of spinal myoclonus by Babinski in 1913 (1), the concept has been proposed and defined by Campbell and Garland (5). Spinal myoclonus has been classified further into segmental spinal (SSM) and propriospinal (PSM) myoclonus (Brown *et al*., 1991). SSM is characterized by synchronous and unilateral jerks involving localized segments of the body, whereas PSM is defined as the presence of flexion or extension jerks of axial musculature with limb involvement. Many cases are idiopathic (Brown *et al*. 1991, Chokroverty *et al*. 1992, Pisano *et al*. 1995, Montagna *et al*. 1997, Vetrugno *et al*. 2000), but infectious (Lubetzki *et al*. 1994, De la Sayette *et al*. 1996), iatrogenic (Jankovic and Pardo 1986, Ford *et al*. 1997), tumoral (Garcin *et al*. 1968, Renault *et al*. 1995), traumatic (Bussel *et al*. 1988, Fouillet *et al*. 1995), ischaemic (Davis *et al*. 1981), demyelinating (Jankovic and Pardo 1986, Kapoor *et al*. 1992) and malformation (Levy *et al*. 1983, Nogues *et al*. 2000) lesions have been reported for each type. Four cases of PSM and three of SSM

related to syringomyelia have been reported previously (Nogues *et al*., 1999). We report on a new case of SSM related to this condition.

Case report

A 75-year-old woman developed involuntary jerks of the abdominal musculature in August 2002. She had been operated on for lumbar stenosis several years earlier. She had no other relevant past medical history. She was admitted to the department of neurology in October 2002. She did not report any infectious or traumatic event. The first episode had not been elicited by a Valsalva maneuver. The jerks were clinically confined to the abdominal muscles without involving the limbs or the cranial nerves and without flexion or extension jerks of the trunk. They were bilateral and most often arrhythmic. They occurred spontaneously, up to twice a second, but could also be triggered by a forced inspiration or by attempting to rise from the supine position. Mental exercise or acoustic, sensitive, or painful stimuli failed to provoke the jerks. They could not be controlled by will. Jerks disappeared during rest, after about 30 minutes in the supine position, and when asleep. Despite the contractions, the patient was able to remain standing, but walking was very uncomfortable, and she was forced to lie in bed until the jerks disappeared. Talking was also disturbed by involuntary noises due to the forced expiration produced by the vigorous abdominal contractions. Simultaneous movements of the limbs were also observed, but their weak amplitude suggested transmission from the abdominal jerks. Neurological examination was otherwise normal, except for a mild motor and sensory deficit with a decreased patellar reflex in the right leg related to the lumbar stenosis for which she had previously had surgery. No dorsal dissociated sensory loss was noted. Extensive blood testing and assays for antibodies (Epstein-Barr virus, cytomegalovirus, human immunodeficiency virus, mycoplasma pneumoniae, borrelia burgdorferi, herpes zoster virus) were normal. Anti-GAD anti-
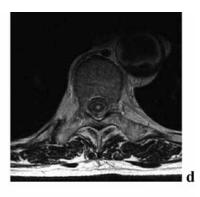
Fig. 1 a-d. — Magnetic resonance imaging of the spine

a. Reconstituted whole spine sagittal spin echo T2-weighted MRI shows a syringomyelic cavity (black arrows) revealed by a high-signal in the thoracic spinal cord between the T3 and T10 levels without a Chiari malformation. The cavity is the largest narrowing the T9-T10 disk. b. Sagittal spin echo T1-weighted MRI without Gadolinium focused on T9-T10 level shows a syringomyelic cavity with an intramedullary hypo-signal. c. Sagittal spin echo T1-weighted MRI focused on T9-T10 level shows the hypointense cavity without gadolinium enhancement. No tumour is detected along the vertebral canal. d. Axial turbo spin echo T2-weighted MRI at T9-T10 level shows that the intramedullary cavity is central.

bodies were absent. MRI of the spine showed a syringomyelic cavity between the T3 and T10 levels without a Chiari malformation (Fig. 1). The lumbar canal was large. The MRI was enhanced to look for a tumor or an arterio-venous malformation underlying the syrinx and revealed no enhancement. Examination of the cerebrospinal fluid was not performed.

Fractionated doses of clonazepam (0.5 mg 4 times daily) rapidly controlled the jerks, which systematically reappeared after drug cessation. Two years later, neurological examination was similar. The patient was still dependent on clonazepam at same doses to control the involuntary jerks.

Electrophysiological investigations

Electroencephalography (EEG) was normal. A 24-hour video-EEG did not show any cortical epileptic activity during the abnormal movements, which were observed in a semi-lying position during daytime only. Somatosensory evoked potentials from stimulation of the median nerves were normal. Stimulation of the tibialis nerves disclosed normal lumbar activities but prolongation of the initial positive cortical SEP, which excluded lumbar stenosis as the cause of the abnormal movements (central conduction time : 38.5 ms and 23.5 ms for the right and left tibialis nerves, respec-

tively). Magnetic cortical motor stimulation to the tibialis anterior muscle was normal (central conduction time : 16 ms and 12.8 ms for the right and left tibialis anterior muscles, respectively).

EMG activity was recorded with a four channel device (*Nicolet Viking*) using concentric bipolar needle electrodes to eliminate the possibility of volume conduction and amplifier filter band widths of 20 Hz and 10 kHz. About forty successive recordings were picked up in order to assess their reliability. Activity was recorded in the following muscles of her right side : biceps, sternocleidomastoid, T7 paraspinal, external intercostal muscles of the 3rd and 8th spaces, upper and lower rectus abdominis and obliquus externus abdominis muscles. The external intercostal muscles of the 3rd and 8th spaces were recorded bilaterally. EMG showed muscle activity bursts of variable duration (256 to 400 ms ; mean, 328 ms) in all muscles, except for the lower rectus abdominis, sternocleidomastoid muscles and biceps where no abnormal activities were recorded (Fig. 2). The EMG discharges were synchronous in all muscles and corresponded to segmental spinal activation at the T3-T10 levels. The time interval between each discharge was 604 to 1340 ms (mean, 792 ms). The rate of the jerks was 45 to 100 cycles per minute. Bilateral EMG recording on thoracic level showed synchronous activity between both sides. A variable time
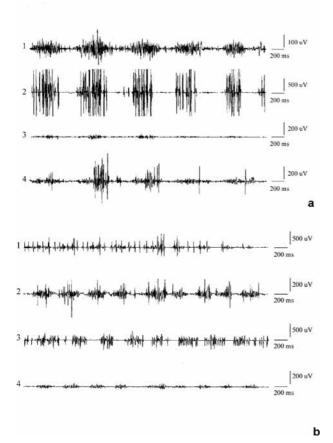
FIG. 2 a-b. — Spinal myoclonus on electromyography
a. Four-channel needle electromyography shows rhythmic bursts of 256 to 400 ms of muscle activity in the intercostal muscles of the 3rd space (channel 2) and of the 8th space (channel 3) and in the upper rectus abdominis muscle (channel 4). Channel 1 records activity of the upper rectus abdominis at the level of the costal insertion. These electrophysiological findings are consistent with spinal myoclonus originating from the spinal T3 to T10 segments. b. Four-channel surface electromyography records synchronous muscle activity between right (channel 1) and left (channel 3) external intercostal muscles of the third spaces, and between right (channel 2) and left (channel 4) external intercostal muscles of the 8th spaces.

interval ranging from 40 to 92 ms was observed in about 50% of the recordings between the upper rectus abdominis muscle and the obliquus externus abdominis muscle.

Discussion

The involuntary movements of our patient fulfilled the clinical criteria for myoclonus limited to the thoracoabdominal muscles (Marsden *et al.*, 1982). Electroencephalographic recordings established that the myoclonus was not of cortical origin. The EMG muscle activity in muscles innervated by the thoracic spinal cord and the topological correlation with the location of the syringomyelic cavity were compatible with myoclonus of spinal origin. Two types of spinal myoclonus may be considered, the segmental spinal (SSM) and pro-

priospinal (PSM) myoclonus. These are two distinct types of myoclonus with different clinical presentations and EMG features in relation to the neurophysiologic organization of the spinal cord, which includes local segmental circuits and propriospinal pathways that link activity in many segments (Brown *et al.* 1991, Chokroverty *et al.* 1992). SSM produces focal or segmental repetitive jerks, confined to one or several adjacent spinal segments (Halliday 1967, Ford *et al.* 1997, Jankovic and Pardo 1986). Most reported cases of SSM have unilateral rhythmic myoclonus involving the limbs at rest, in the recumbent posture, and often persisting during sleep. Voluntary activation of the involved musculature may reduce or attenuate the myoclonus by a normal inhibitory reflex (Garcin 1968, Ford *et al.* 1997). In contrast, PSM is characterized by repetitive, predominantly axial, and often arrhythmic, flexor or extensor jerks involving many spinal segments (Brown 1991). Both PSM and SSM may be stimulus-sensitive (Brown 1991, Davis *et al.* 1981).

The distribution of spinal myoclonus has long been recognized as having great clinical localizing value (Garcin 1968). In our patient, the absence of significant delay between the muscles involved in the movements is in favour of SSM arising from the thoracic spinal cord. The delay recorded between the upper rectus abdominis and the obliquus externus muscles was small and intermittent, by contrast to another case with a constant delay of 40 ms between the EMG activity of these muscles, and diagnosed by the authors as PSM (Pisano, 1995). Moreover, according to Nakazato *et al.* (19), the presence of an intermittent delay is not an exclusion criteria for SSM. On a clinical point of view, another argument that excludes PSM is that our patient showed no flexion jerks of the trunk or the limbs as described in PSM. EMG did not record abnormal activity in limb muscles nor signs of rostro-caudal propagation at low velocity along the spinal cord segments. Based on EMG recordings and after review of the literature, we propose the diagnosis of SSM, despite its unusual bilateral presentation. We did not look for an activity of the diaphragm muscle because of the risks related to the procedure. Such activity would have been a sign of a propriospinal spread beyond the syringomyelia.

Both SSM and PSM are usually idiopathic, although numerous types of spinal cord lesions have been reported. Syringomyelia is one of the spinal cord diseases that may cause spinal myoclonus (Nogues *et al.* 1999, Bagnato *et al.* 2001). The former reports four cases of PSM and three of SSM due to this condition. In all cases of SSM, spinal myoclonus was associated with an extended cervical and thoracic syrinx, and with a type-1 Chiari malformation. By contrast, our patient presented a syrinx limited to the thoracic

spinal cord, without Chiari malformation. In syringomyelia, a particular type of SSM is called minipolymyoclonus, and is characterized by small movements with amplitudes just sufficient to produce movements of the joints of the fingers or toes. It has been reported in 10 patients so far (Nogues *et al.* 1999, Bagnato *et al.* 2001).

Both SSM and PSM are the result of spontaneous discharges from groups of anterior horn cells (Brown *et al.* 1991, Bagnato *et al.* 2001). The reason why syringomyelia induces spinal myoclonus remains unknown. Possible explanations include direct damage of the motoneurons by the syrinx or inflammation around the cavity, lesions of spinal interneurons, and/or damage of the descending motor pathways by the syrinx or the associated Chiari malformation (Brown *et al.* 1991, Bagnato *et al.* 2001, Nogues *et al.* 1999).

In conclusion, we report a new case of segmental spinal myoclonus related to syringomyelia with limited thoraco-abdominal movements. The original findings in this case are the axial location of the myoclonus, restricted to the abdominal and thoracic muscles, and its bilateral presentation.

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