

Short report

Jerking stiff-man syndrome

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SUMMARY A female patient had permanent axial muscular rigidity similar to the "stiff-man syndrome", together with axial myoclonus triggered by stretch reflexes and by supramaximal stimulation of the supraorbital nerve. The disorder responded to treatment with diazepam and baclofen. This disorder bore a marked similarity to the so-called "jerking stiff-man syndrome".

The "stiff-man syndrome" is a disease of unknown aetiology¹ in which no structural alterations of the central nervous system have been reported.² The disorder is characterised clinically by permanent axial muscular contraction and spasms, which respond to treatment with diazepam³ and baclofen.⁴⁻⁵ There may occasionally be nocturnal myoclonus,² but no other type of myoclonus has been described in this disorder. Recently,⁶ there has been a report of a patient who had persistent axial muscle rigidity together with a peculiar myoclonus and clinical and radiological signs suggestive of a structural lesion of the central nervous system. This disorder has been termed "jerking stiff-man syndrome" and is considered to be different from the "stiff-man syndrome".⁶ We report another case in which there was permanent axial rigidity associated with myoclonus clinically similar to that described by Leigh *et al.*⁶

Case report

At 40 years of age, this patient noticed that the toes of her right foot became fixed in plantar flexion, at first only when walking, and later permanently. Six years later she suffered stiffness and sharp jerks in the legs when walking quickly, which interrupted her gait. These jerks also appeared during sleep. The stiffness in the legs progressed to the point where she was unable to walk, and at age 52 the patient was diagnosed in another hospital as having spastic paraparesis. A decompressive cervical laminectomy was done, which did not improve her condition and she was then treated with diazepam for two years, the patient being able to walk with assistance. Clinical examination at 54 years of age showed a marked increase in muscle tone, which produced a wooden consistency, in both the abdominal and dorso-lumbar muscles. Only the left arm was hypertonic; a clasp-knife response was sometimes obtained and it predomin-

ated in the proximal and flexor musculature. The legs were set in almost immovable extension, with the toes in plantar flexion. Muscle power was normal in the arms and could not be tested in the legs. Stretch reflexes were exaggerated in the arms, particularly on the left, and there was bilateral knee clonus. Achilles tendon and abdominal reflexes were unobtainable and the plantar reflexes were equivocal. Long-lasting muscle spasms were easily provoked. The rest of the examination was normal.

The patient was treated with 20 mg of diazepam and 30 mg of baclofen a day and seven days later only the abdominal and dorso-lumbar muscles were still extremely rigid. In the electromyographical study, muscle relaxation was easily achieved in the left leg, while in the right leg it was very difficult and in the abdominal musculature impossible, since there was permanent muscular activity with low voltage intermediate to interferential tracings in these muscles. Other routine EMG studies gave normal results. The following studies were normal: blood and urine analysis, glucose tolerance curve, serum electrolytes including calcium and magnesium, liver function tests, protein electrophoresis, muscle enzyme determinations, urine catecholamine studies, routine CSF analysis, ECG, EEG, radiography of the skull, spinal column and chest, myelography and CT scan. Sleep EEG studies were normal, but showed that the muscular rigidity disappeared during sleep.

Medication was suspended and after 24 hours the severe muscular rigidity had returned. However, the most striking feature was the appearance of violent jerks, which affected the axial and leg muscles symmetrically and synchronously. These jerks caused adduction in the arms, extension of the trunk and hyperextension of the legs, together with a noisy exhalation which interrupted speech. The jerks were apparently spontaneous, but could be triggered by any stimulus. The frequency of the myoclonus increased to as much as twice a second, producing severe respiratory distress and profuse sweating. The patient was given 5 mg of diazepam intravenously and two minutes later she was deeply relaxed and able to walk needing a support because of instability. The spontaneous myoclonus had disappeared. The effect of the injection lasted for about three hours and during this time it was found possible to provoke the myoclonic jerks by testing the jaw-jerk and other stretch reflexes. If the mouth was kept closed and the head braced, a tap to the jaw did not produce a myoclonic jerk; neither did one appear with tactile perioral stimulation. Supramaximal electrical stimulation of the supraorbital nerve also produced a myoclonic jerk, which

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was recorded in the trapezius with a latency of 75 ms with respect to the stimulus. The jerks were not accompanied by changes in the EEG.

The patient was treated with 60 mg of diazepam a day orally, giving blood levels of 6.69 $\mu\text{g/ml}$. Slight hypertonia persisted in the left arm and the abdominal muscles were only a little contracted. The knee jerks were exaggerated and the plantar responses downgoing. However, somnolence, dysarthria and ataxia appeared, so that the patient needed support in walking. She was finally treated with 30 mg of diazepam and 60 mg of baclofen a day, the blood level of diazepam being 1 $\mu\text{g/ml}$. The patient, who rarely suffered muscle spasms, was able to walk almost without help over short distances. Sleepiness had disappeared and ataxia and dysarthria decreased.

Discussion

Our patient's syndrome was characterised by the association of a permanent muscular rigidity with a peculiar myoclonus. The muscular rigidity was predominantly axial, disappeared during sleep, was accompanied by painful muscle spasms and responded to low intravenous doses of diazepam. These characteristics are the same as those described for the permanent muscular rigidity in the "stiff-man syndrome".¹ The response to diazepam is of great diagnostic value⁷ and makes it unnecessary to carry out other studies⁸ to determine whether the muscle rigidity is central or peripheral in origin. Moreover, our patient's clinical picture differed clearly from other permanent muscular contractions already reviewed.^{9 10}

The myoclonus was characterised by the following: it appeared on withdrawing medication, was apparently spontaneous and had a high frequency, affecting the axial and leg muscles bilaterally and synchronously was severe and unpleasant, and responded dramatically to diazepam. Under treatment, the spontaneous myoclonus disappeared and gave way to a reflex myoclonus produced by stretch reflexes and electrical stimulation of the supraorbital nerves. To our knowledge, this type of myoclonus has been described on only one occasion⁶ in association with permanent muscular rigidity similar to that of our patient. The only difference is that in that study the myoclonus was also triggered by tactile perioral stimulation. According to Leigh *et al*⁶ the origin of this myoclonus is in the brainstem, and its production mechanism is different from those hitherto described in other reflex myoclonus.^{11 12} It is difficult to know what is the relationship between the myoclonus, muscular spasms and permanent muscular rigidity. The three signs respond to the same medication and this may support the idea that all three have a common pathophysiological mechanism.

The myoclonus separates our case from typical cases of "stiff-man syndrome". On the other hand, although our

case shares important features with that of Leigh *et al*⁶ they do not entirely overlap. In these authors' case there were clinical and radiological signs which suggested a structural lesion in the brainstem and cerebellum and these signs were not found in our study. Our patient's dysarthria and ataxia probably was related to the high doses of medication administered.

Although it is difficult to determine accurately the nosological status of our case, the name "jerking stiff-man syndrome" defines it adequately. It is necessary to point out that the myoclonus only became apparent in the later stages of the evolution of the disorder and medication had to be suspended in order to show it clearly. The number of cases found similar to ours may increase if in those with slowly evolving stiff-man syndrome medication is suspended and the manoeuvres proposed by Leigh *et al*⁶ to trigger this type of reflex myoclonus are carried out.

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