pathologically to be related to a plaque in the lower medulla.

Although a diagnosis of multiple sclerosis in life was felt to be likely in our patient, the clinical presentation, the stepwise progression with no previous history of neurological dysfunction to a final, irreversible, locked-in state, was unusual. Furthermore, oligoclonal bands were absent from the CSF on two occasions, a finding usually seen in only 30% of patients with multiple sclerosis. Subsequent transverse myelitis of the spinal cord showed dissemination of the disease in time and place. Furthermore, the midbrain/brain stem demyelinating lesion showed the pathological features of multiple sclerosis. The asymmetry and extent of this lesion are features against a diagnosis of central pontine myelinolysis. Necrotising myelopathy of the vertebra seen in this patient may occur in association with carcinoma, but is probably more often associated with plaques of multiple sclerosis elsewhere in the nervous system. Overall, the diagnosis of multiple sclerosis best fits the clinical and pathological features of this most unusual case.

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Spontaneous ejaculation secondary to spinal cord disease

Spinal cord lesions in humans are associated with impaired ejaculation,1-3 suggesting that the effects of descending pathways on the spinal ejaculatory mechanism are predetermined by corticospinal input. In rodents, however, there may be an additional and important descending inhibitory pathway.4 In keeping with the presence of a similar inhibitory pathway in humans, we report the case of a patient with a spinal cord lesion who was unusually associated for a period of time with spontaneous or easily provoked ejaculation.

The patient, a 70-year-old man, had noted an episode of spontaneous ejaculation when his glans touched the side of a catheter bottle during admission to hospital for a myocardial infarction five years previously. About every three weeks thereafter, in response to this report, he ejaculated at the onset of micturition, in association with symptoms of urinary urgency and the sensation of bladder fullness. Spontaneous ejaculation also occurred occasionally during midstream micturition. These episodes occurred with a flaccid penis and with a buildup of pelvic floor tension followed by minor, rhythmic contractions of the pelvic floor which he described as "mini-orgasm".

He had not had any spontaneous erections since the onset of these symptoms, though sexual function had previously been normal.

Two years before presentation he developed constant lower limb paresthesiae sometimes affecting the buttocks. These sensory symptoms were unpleasantly resolved within five to 10 minutes of lying flat, and resumed as he placed his feet on the floor. For two years he had had worsening stiffness of his legs, which was aggravated by prolonged walking and sometimes associated with discomfort in his buttocks. In recent months he had noted spontaneous jerks of his legs when lying flat. He had been in atrial fibrillation since his myocar dial infarction and he described a number of carotid transient ischaemic attacks without any abnormalities on the cranial CT scan or digital subtraction angiography of his neck vessels during subsequent investigation.

On examination his blood pressure was 150/90 and his pulse irregularly irregular. The dorsalis pedis pulses were absent but the rest of the peripheral vascular examination was normal. His cranial nerves and arms were also normal, but in his legs there was a moderate spastic paraparesis with hyperreflexia and extensor plantar responses. His abdominal reflexes were preserved and there was no sensory loss. Bulbo cavernous and anal reflexes were present, as well as the left cremasteric reflex.

Routine haematological and biochemical tests were normal and spinal serology negative. The electrocardiogram confirmed atrial fibrillation. Urodynamical studies indicated the presence of detrusor hyperreflexia and marked detrusor-sphincter dyssynergia, with a residual bladder volume of 250 ml. Cerebral MRI, analysis of the cerebrospinal fluid, and a number of electrophysiological studies were normal, including lower limb nerve conduction studies, spinal and pudendal somatosensory evoked potentials, brainstem auditory and visual evoked potentials, and finally electromyography of the anal sphincter. A full myelogram and computed myelography a L4-5 and L5-S 1 were also normal. MRI of the thoracolumbar region (figure) showed a high signal lesion extending over the whole mid-thoracic cord. The lesion was situated in the anterior part of the cord and was bilateral. The cord was not expanded on the myelogram or in the MRI study. Overall, the results of the imaging studies suggested that the cord lesion was ischaemic in origin.

Although the patient's ejaculation was not tested for the presence of semen, the description of his symptoms favors the occurrence of true ejaculation rather than seminal emission or the passage of urinary gravel, as rhythmic contractions of the pelvic floor accompanied the expulsion of fluid. The most unusual feature in this patient was the spontaneous or easily provoked ejaculation in the presence of a spinal cord lesion. Earlier reports of this association have been anecdotal,4 and cord pathology is generally found to lead instead to impairment of ejaculation. It is known, however, that a reflex mechanism exists in the T9-L2 (sympathetically driven emission of semen) and S2-3 cord segments (somatically driven propulsion of semen) from which involuntary ejaculation may be produced.5 7 Thus in patients with spinal injuries above T9, emission can be induced using electrocutaneous or sympathetic stimulation below the level of the lesion.8 9 In rodents this spinal sexual mechanism receives descending inhibition to pudendal motor neurons and lumbar sacral interneurons, and in male animals with a spinal cord transection ejaculation can be induced by fluid distention of the urethra in a manner reminiscent of our patient's symptomatology. In humans the overriding supraspinal influence on sexual function is usually excitatory, but it is possible that spinal lesions could result in a situation resembling that in rodents by selectively interfering with an additional, smaller descending inhibitory pathway projecting to the spinal ejaculatory mechanism. Support for this argument may also be found in human spinal injury where a variety of stimuli, including bladder distension, can induce exaggerated autonomic responses such as sweating and hypertension.1 As emission is primarily under sympathetic control2 10 it is at least theoretically possible that our patient's symptoms could be viewed as a form of autonomic disinhibition, in this instance affecting the whole reflex ejaculatory mechanism.

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Abnormal evoked potentials in Miller-Fisher syndrome: further evidence of combined central and peripheral demyelination

We read with interest the short report by Ferrer et al.1 about a patient with Miller-Fisher syndrome in whom CNS demyelination was documented by MRI a few years later.2 We would like to direct attention to an additional case supporting the possibility of combined central and peripheral demyelination in this syndrome.

A 66-year-old previously healthy woman was referred for diplopia of 24 hours duration, an unsteady gait, and bilateral ptosis of the upper limbs. Examination showed bilateral ptosis, complete external ophthalmoplegia, and severe axial and limb ataxia. On the second day of admission to hospital, progressive proximal muscle weakness developed accompanied by generalised areflexia. The plantar responses were flexor. All sensory modalities were intact. Routine laboratory examinations, CT of her brain, and EEG were normal. The sterile cerebrospinal fluid obtained on the fifth day of admission to hospital was clear and under normal opening pressure, with a protein concentration of 0·75 g/l and no oligoclonal bands.

Electrodiagnostic studies disclosed prolonged distal motor latencies, slow conduction, and prolonged F-responses in the median, ulnar, common peroneal, and posterior tibial nerves. Needle electromyography indicated active symmetrical generalised denervation. The visual evoked potentials (VEPs) were markedly abnormal for prolonged latency of the P100 peaks (right 126 ms; left 128 ms; normal 100–110 ms), which retained their normal amplitude. Brain stem auditory evoked potentials (BAEPs) showed an impaired wave pattern with prolongation of N1, N2, and brain stem conduction time.

Within one week and with only conservative treatment, spontaneous improvement was noted of the axial and horizontal eye movements, but there was still partial limitation of vertical gaze.

Neurological examination after three months showed a full range of eye movements, normal muscle strength, and deep tendon reflexes. The VEPs and BAEPs had returned to normal.

The aetiology and pathogenesis of the Miller-Fisher syndrome are not well outlined, particularly the location of the pathological changes. Most workers favour a peripheral origin, although others suggest brain stem inflammatory lesions or a combination of central and peripheral demyelination.4 Abnormal BAEPs have been described in only a few cases.5 Results from MRI have usually been normal, except in the reports of Giroud et al.1 and Ferrer et al.3 Abnormal BAEPs and VEPs, together with evidence of demyelination on brain MRI, support the hypothesis that in the Miller-Fisher syndrome there is a combination of central and peripheral central demyelination. Considering the dissimilarities between central and peripheral myelin, however, this conclusion leaves the immunological mechanism by which this process is mediated unclear.

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