MRI showed a marked decrease in the bulk of the tumour from the original excision site, but extensive lateral extension along the floor of the skull base with encasement of the intrapetrous portion of the left internal carotid artery. Digital subtraction angiography with intravenous contrast showed a normal venous phase and the absence of vessel displacement or tumour circulation.

Using a postero-lateral approach a further resection achieved a good macroscopic clearance apart from leaving some residual tumour in the cavernous sinus. The histology was identical to that of the sample taken during the first operation. Post-operatively there were left sided sixth and seventh cranial nerve palsies together with marked speech and swallowing difficulties. A feeding gastrostomy was required.

In spite of radiotherapy this neurological deficit has persisted and the left side of the patient's face has become numb. MRI one year after the second operation has shown extensive tumour recurrence affecting the clivus, extending anteriorly into the nasopharynx and posteriorly to the posterior fossa with compression of the medulla and cerebellum.

**DISCUSSION**

Skull base chordomas are rare tumours producing infiltrative destruction of bone. The tumours are locally and systemically aggressive with the potential to cause a variety of cranial nerve palsies at anatomically unrelated sites. The cranial nerve most often affected is the sixth, followed by the ninth and tenth.1

Isolated lesions are uncommon from any cause. Chondroid chordomas may be the cause and exceptionally may arise within the hypoglossal canal.2 Inability of the patient to speak and to move the tongue on either side is an unusual symptom which, as far as we are aware, has not been described previously in relation to intracranial chordomas. It is reminiscent of the neck-tongue syndrome in which afferent fibres from the lingual nerve travelling via the hypoglossal nerve to the second (and possibly the third) cervical root are compressed at the craniovertebral junction on the side of head movement.3 The neck-tongue syndrome is usually a benign disorder not associated with serious intracranial pathology. It causes unilateral numbness of the tongue on head movement and patients may have difficulty in that their tongue is being twisted sideways in their mouth. Dysarthria may accompany the hemiglossal sensory disturbance.4 Our patient had restricted tongue movement and an inability to speak without altered sensation, perhaps suggesting that on neck flexion the tumour compressed the hypoglossal nerve itself rather than the second and third cervical roots.

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**Locked-in syndrome in fulminant demyelinating disease**

The locked-in syndrome is a state of paralysis of all limbs and the lower cranial nerves, with preserved consciousness, vertical eye movements, and blinking. The pathological damage is usually located in the basis pontis of the ventral pons, the most common cause being infarction. Rarer causes include primary tumours, central pontine myelinolysis, pontine haemorrhage, and brain stem encephalitis. This report describes a fulminant first presentation of demyelinating disease in lesions of multiple sclerosis, which evolved within weeks into a fatal locked-in state. Pathological examination showed extensive demyelination of the ventral pons.

**CASE HISTORY**

A previously fit 50-year-old Spanish man developed slurred speech followed two days later by a mild weakness of his right arm and leg. Four days later he developed unsteadiness of his legs and two days later presented with normal neurological examination. On examination he was dysarthric. Fundi, visual acuity, and fields were normal; his pupils were mid-pupil and briskly reactive, with preserved spontaneous and primary prosopid and diplopia in all directions of gaze, except the right lateral. There was mild pyramidal weakness of his right leg. His reflexes were brisk with extensor plantars and he had marked ataxia of the trunk and limbs.

Computed tomography showed a low density, non-enhancing area in the left rostral midbrain, with normal opacification of the basilar artery. His CSF was acellular, with normal protein and glucose; oligoclonal bands were absent. Routine haematological and biochemical tests, clotting studies, erythrocyte sedimentation rate, C reactive protein, and fasting lipids were normal. An antoantibody screen was negative. An ECG and chest radiograph were normal. A preliminary diagnosis of brainstem stroke-in-evolution was made, and he was instigated intravenously with heparin. Subsequent vertebral angiography, however, was normal, and the heparin treatment was stopped. Visual evoked responses were slightly delayed. Despite the absence of oligoclonal bands in the CSF, the possibility of multiple sclerosis was considered, and high dose methylprednisolone was given for three days.

His initial response to steroids was good. The diplopia and ataxia improved. A right internuclear ophthalmoplegia was now apparent. Ten days after the end of the steroid treatment he developed complete transverse myelopathy with a sudden onset of flaccid paraplegia with areflexia and a sensory level to all modalities at T10. Myelography was normal and CSF protein, glucose, and cell count were also normal. Oligoclonal bands were again absent.

High dose steroids were restarted. MRI showed high signal lesions on T2-weighted images in the midbrain, cervical region, and parietal area, consistent with infarction or demyelination. One week later he developed weakness and sensory loss in the upper limbs, with acute aponia and ventilatory failure requiring ventilation. He had a flaccid paresis of the arms with sensory disturbance to C5. Within 24 hours he had developed the locked-in syndrome with full consciousness, the only movements remaining being blinking and vertical eye movement. He had a cardiac arrest and died 11 days after developing the locked-in syndrome.

**PATHOLOGICAL EXAMINATION**

Acute threcobronchitis was found at necropsy. The brain was externally normal. No hemispheric or periventricular demyelinating lesions were seen. The midbrain and brain stem down to the lower medullary level were macroscopically abnormal. Externally, the pons and medullas were expanded and on dissection there was extensive discolouration and blurring of normal structures, with central granularity of texture. Sections stained with haematoxylin-eosin and luxol fast blue-cresyl violet showed sharply defined, irregular loss of myelin staining from the ventral half of the midbrain at the inferior collicular level (figure) to the ventral half of the pontine isthmus. Microscopic examination showed selective demyelination in the affected zone, which occupied the greater part of the pons and extended down into the medulla, the ventral two-thirds of which were affected. Axons were relatively spared and neurons and vessels were intact in the affected area; there was a marked cellular response affecting lymphocytes, macrophages, and astrocytes.

The spinal cord was almost entirely necrotic, with a loss of normal structure. Necrotising myelopathy affected all tissue elements. Ghost neurones were visible at some levels. Peripheral cord myelin was focally spared, as was myelin in the spinal nerve roots.

**DISCUSSION**

The case has been described of a patient with fulminant demyelination resulting in a fatal locked-in state, and with pathological evidence of extensive demyelination in the ventral pons.

Although the locked-in syndrome has been described previously in patients with multiple sclerosis,2,3 demyelination of the ventral portion of the brain stem was not present. Instead, demyelination was found in the internal capsule, cerebral peduncles, and tegmentum. Boor et al4 reported a patient with stepwise deterioration of lower brain stem dysfunction, similar to that described here, but without development of the locked-in state. The main clinical features of Boor et al's patient were bulbar weakness together with the failure of automatic respiration, which was shown...
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/brainstem demyelinating lesion showed the typical 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,1,2 suggesting that the effects of descending pathways on the spinal ejaculatory mechanism are predominant rather than excitatory. In rodents, however, there may be an additional and important descending inhibitory pathway.3 In keeping with the presence of a similar inhibitory pathway in humans, we report the case of a patient who developed a spinal lesion 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, unless he observed 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 a "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 usually resolved within five to 10 minutes of lying flat, and resolved 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 myelopathy, carotid infarction and he described a number of carotid transient ischaemic attacks with- out 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 present 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 fluid serology negative. The electrocardiogram confirmed atrial fibrillation. Urodynamic studies indicated the presence of detrusor hyperreflexia and marked detrusor-urinary dyssymmetry, 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 myelography and computed myelography a L-4–5 and L-5–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 ejaculate was not tested for the presence of semen, the description of his symptoms favours 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 presence of such 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 (symptomatically driven emission of semen) and S2–3 cord segments (somatically driven propulsion of semen) from which involuntary ejaculation may be produced.5,6 Thus in patients with spinal injuries above T9, emission can be induced using electrical or sympathetic stimulation below the level of the lesion.6,7 In rodents this spinal sexual mechanism receives descending inhibition to pudendal motor neurons and lumbosacral inter-neurons, and in male animals with a spinal cord transection ejaculation can be induced by fluid distension of the urethra8 in a manner reminiscent of our patient’s symp-tomatology. 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 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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