brain stem strokes the lesions were exclusively located in the pontine tegmentum. Therefore it is likely that the efferent pathways originating from the pontine micturition centre were affected at the level of the medulla in our patient. This case shows that transient urinary retention with minimal neurological symptoms can be associated with medullary lesions, and is probably caused by direct viral infection or by parainfectious demyelination.

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Long term continuous bilateral pallidal stimulation produces stimulation independent relief of cervical dystonia

Idiopathic cervical dystonia is the most common form of focal dystonia in adults.[1 2] It is characterised by involuntary, sustained contractions of the cervical muscles and produces abnormal head movements or postures. Although deep brain stimulation (DBS) of the globus pallidus internus (GPi) is now accepted in the treatment of a wide spectrum of dystonias including cervical dystonia (for a review, see Krauss[3]), the mechanisms underlying its beneficial effects on dystonia remain unclear. We report a patient in whom continuous long term (22 month) bilateral pallidal stimulation eventually produced stimulation independent relief of cervical dystonia.

This 54 year old right handed man had a one year history of involuntary head rotation toward the left. He had no previous exposure to neuroleptics and no family history of dystonia. On admission in June 2000, he manifested severe cervical dystonia with neck pain characterised by left and posterior head turn and tilt (fig 1A). The right sternocleidomastoid muscle was contracted and hypertrophied. His cervical dystonia decreased in the supine position or when performing a sensory trick which consisted of putting his right hand on his chin or neck. On the Toronto western spasmodic torticollis rating scale (TWSTRS), his total severity score (TSS) was 25 (maximum = 35), and his total disability score (TDS) was 21 (maximum = 30).

Brain magnetic resonance imaging showed no obvious abnormalities. Sequential pharmacological trials including diazepam (4×2 mg/day), trihexyphenidyl (4×2 mg/day), tiapride (3×25 mg/day), and sulpiride (3×50 mg/day) produced unsatisfactory results. Botulinum toxin (BTX) treatment is now established as a treatment of choice in patients with cervical dystonia, while bilateral pallidal stimulation is still an investigational treatment. However, at the time of these trials, the use of BTX injections was not approved in Japan for the treatment of cervical dystonia. The patient was referred for surgery after informed consent had been obtained from both him and his family.

Quadripolar DBS electrodes (model 3387, Medtronic Inc, Minnesota, USA) were implanted as previously described.[4 5] The target points were determined to be 2 mm anterior and 20 mm lateral to the midpoint of the anterior to posterior commissure line, and 1 mm dorsal to the third ventricle floor. The most ventral contacts were placed exactly on the target points (fig 1B, C). As six day stimulation tests confirmed the beneficial effects of DBS, a receiver for the external transmitter was implanted (Mattrix transmitter, model 3272, Medtronic). The cervical dystonia and neck pain were markedly alleviated within minutes of initiating DBS.[4] Extensive trials showed that optimal results were produced at a frequency of 60 Hz, pulse width 500 μs, and amplitude 6.0 V.[4 5] As the external stimulator that we first employed permitted bipolar but not monopolar stimulation, contact 0 was used as the cathode and contact 1 as the anode. Upon stimulation, the cervical dystonia was markedly alleviated (fig 1D), with TWSTRS TSS and TDS of 6 and 5, respectively. By contrast, when stimulation was discontinued, the dystonic symptoms reappeared immediately and resumed at the preoperative level (fig 1E).

With stimulation, the patient was able to return to his job and resume his normal life. However, he began to complain that using the external transmitter was inconvenient. Moreover, he was afraid of the immediate return of his cervical dystonia when he discontinued stimulation—for example, when replacing the battery or taking a bath. Therefore, in June 2001 the external transmitter receiver was replaced with internal pulse generators (IPGs) (Itrel III, Medtronic Inc). Extensive trials showed that optimal results were produced at 60 Hz, 450 μs, and 2.6 V. Monopolar stimulation was applied using contacts 0 and 1 as the cathode and the pulse generator as the anode. At this optimal setting, his TWSTRS TSS and TDS were 5 and 5, respectively. Continuous bilateral pallidal stimulation allowed him to continue a normal life. With two active electrodes stimulating (at 60 Hz, 450 μs, and 2.6 V) for 24 hours a day, the battery life of the Itrel III is about 24 months. Thus, in April 2003 (22 months after placement of the DBS electrodes), revision of the IPGs was carried out before depletion of the batteries. This was the first time of discontinuation of the DBS since the implantation of the IPGs. Expecting that his cervical dystonia would reappear as usual when stimulation using the external transmitter was discontinued, we planned to switch on the generators immediately after surgery. However, without stimulation, the cervical dystonia did not worsen and at the time of writing (fig 1F) his TWSTRS TSS and TDS continue to be 6 and 5, respectively. Compared with the values obtained before the implantation of the DBS electrodes, there was approximately 76% improvement in the cervical dystonia. Pharmacotherapy had been continued unchanged from before the surgery: diazepam (4×2 mg/day), trihexyphenidyl (4×2 mg/day), tiapride (3×25 mg/day), and sulpiride (3×50 mg/day).

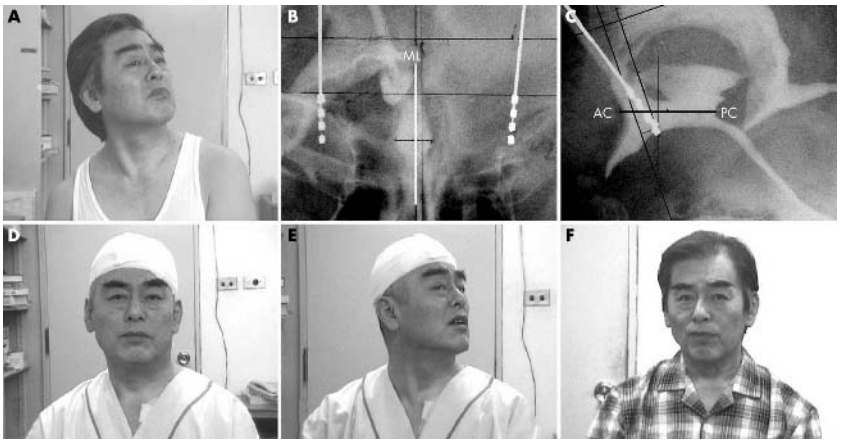

Figure 1 (A) Photograph taken before implantation of the deep brain stimulation (DBS) electrodes. Note severe cervical dystonia characterised by left and posterior head turn and tilt. The right sternocleidomastoid muscle was contracted and hypertrophied. (B, C) Location of the electrodes superimposed on the frontal (B) and lateral (C) view in selective third ventriculography. The target points are indicated by asterisks. AC, anterior commissure; ML, midline; PC, posterior commissure. (D, E) Photographs taken 10 days after implantation of the DBS electrodes. On stimulation with the external stimulator, cervical dystonia was markedly alleviated (D). In contrast, upon discontinuation of stimulation, the dystonic symptoms reappeared immediately and returned to the preoperative level (E). (F) Photograph taken six months after discontinuation of the GPi DBS with internal stimulators. Without stimulation, there was marked relief of the cervical dystonia.

Comment

Interestingly, long term (about 22 months) continuous bilateral GPi DBS with IPGs produced stimulation independent relief of this patient's cervical dystonia, although short term (less than 24 hours because of limited battery life) and discontinuous DBS with the external transmitter had not yielded such beneficial effects. We cannot exclude the possibility that spontaneous remission of the cervical dystonia occurred[1] during the use of IPGs. However, our experience with this patient suggests that chronic GPi DBS may result in normalisation of the altered basal ganglia related motor circuits that are implicated in the occurrence of dystonia,[6–8] and that over time the normalised state may persist independently of DBS. Alternatively, chronic GPi-DBS may lead to the reorganisation of the functional anatomy of the motor circuits at unknown levels, thus resulting in suppression of dystonia. Long term remission of idiopathic cervical dystonia after BTX treatment has also been reported.[9] It is presently unknown whether peripheral BTX injection and GPi-DBS share a common mechanism in producing remission of cervical dystonia. From a practical standpoint, in patients undergoing GPi-DBS for cervical dystonia treatment, it may be necessary to apply DBS as continuously as possible and to avoid any unnecessary discontinuation of the IPGs. In addition, when battery depletion makes it necessary to revise the IPGs, it may be advisable to determine whether further stimulation is actually needed in these patients.

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Severe vasculitic neuropathy following influenza vaccination

Current Department of Health (UK) guidelines suggest that all people aged over 65 years should receive annual influenza vaccination. There is a range of adverse medical sequelae associated with this. Two serious complications are Guillain-Barré syndrome[1] and systemic vasculitis.[2 3] The strength of association between influenza vaccination and Guillain-Barré syndrome has been questioned, and little is known of the pathogenesis.[3] Several investigators have reported a close temporal association between influenza vaccination and systemic vasculitis of various classifications.[2 3] Vasculitic peripheral neuropathy was not a feature in any of the reported cases.[2 3]

We report a case of biopsy proven vasculitis, presenting as mononeuritis multiplex, following influenza vaccination. The clinical picture evolved rapidly into a syndrome indistinguishable from axonal Guillain-Barré syndrome. This suggests a differential diagnosis for post-vaccination neuropathy, with implications for management. We believe this is the first report in which there was an associated peripheral neuropathy at presentation. It raises issues about the aetiology and pathogenesis of vaccination associated neuropathy.

Case report

A previously fit and active 72 year old woman was admitted after routine influenza vaccination. She gave a history of progressive distal arm and leg symptoms, tiredness, and anorexia. One week after vaccination she experienced pain in her left buttock which radiated down her left leg. In the following 24 hours she developed weakness in her right hand in a median nerve distribution. This was followed by similar symptoms in the left hand. In the 24 hours before admission her left foot became numb and weak and she complained of general exhaustion and anorexia.

There was no previous history or relevant family history of neurological disorder, and the only drug she used regularly was pravastatin, which she had been taking for three years. She had received similar influenza vaccination on three previous occasions without complications. There was no relevant past medical history.

Examination revealed bilateral median nerve palsies and left lower limb sensory loss consistent with abnormality of the distal sciatic nerve. Cranial nerve examination, including fundoscopy, was normal. General examination revealed a purpuric rash on both ankles. There was no evidence of involvement of other organs.

On admission the erythrocyte sedimentation rate (ESR) and C reactive protein were raised at 105 mm/h and 35 mg/l respectively. Full blood count, urea and electrolytes, uric acid, thyroid function, liver function (apart from reduced albumin (34 g/l)), total protein and electrophoresis, calcium, creatine kinase, glycosylated haemoglobin (HbA1C), hepatitis B surface antigen, vitamin B-12, and folate were all normal. A vasculitic screen—including antinuclear antibody, perinuclear and cytoplasmic antineutrophil cytoplasmic antibody, and rheumatoid factor—was negative. Influenza A and B complement fixation tests were negative. Urinanalysis was normal. The patient declined lumbar puncture.

Nerve conduction studies carried out on admission confirmed a peripheral neuropathy with features of a mononeuritis multiplex syndrome. Motor nerve studies are shown in table 1. Sensory conduction was preserved in both upper and lower limbs.

A skin biopsy of one of the purpuric ankle lesions showed prominent vasculitis involving the small arterioles without granuloma formation. On radial nerve biopsy there was severe axonal loss but no definite vasculitis. No evidence of myelin debris was found.

She was treated with high dose oral steroids, azathioprine, and monthly pulses of intravenous cyclophosphamide. Six weeks later there was evidence of further clinical deterioration, although inflammatory markers were low (ESR 21 mm/h). She had severe distal weakness, sensory loss, and hyporeflexia of all four limbs, suggestive of involvement of all peripheral nerves. She was very disabled, unable to walk or use either hand.

The electrophysiological findings at this stage were indistinguishable from a severe acute motor and sensory axonal neuropathy of Guillain-Barré syndrome type (table 1, study 3). Electromyography demonstrated complete denervation in the small hand muscles bilaterally and severe partial denervation in lower limb groups (the gastrocnemius and tibialis anterior were tested). All sensory action potentials were absent (radial, median, ulnar, and sural groups; tested antidromically). F waves were normal at presentation; however, they were absent in

Table 1 Motor nerve conduction studies

Nerve	Study	DML (ms)	CV (m/s)	Amplitude (distal/proximal) (mV)
Right median	1	4.0	52	1.5/1.5
	2	6.8	–	0.2/0.0
	3	Absent	–	Absent
Left median	1	3.8	37	1.7/0.9
	2	3.8	–	0.5/0.0
	3	Absent	–	Absent
Right ulnar	1	3.0	61	7.0/7.0
	2	3.2	62	2.5/2.5
	3	Absent	–	Absent
Right medial popliteal	1	5.2	40	6.0/6.0
	2	4.6	41	0.5/0.5
	3	Absent	–	Absent

Amplitude, peak to peak amplitude of compound muscle action potential; study 1, at presentation; study 2, two weeks later; study 3, six weeks later.
CV, conduction velocity; DML, distal motor latency.