

SHORT REPORT

Unilateral globus pallidus internus stimulation improves delayed onset post-traumatic cervical dystonia with an ipsilateral focal basal ganglia lesion

J W Chang, J Y Choi, B W Lee, U J Kang, S S Chung

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Treatment with unilateral left globus pallidus internus (GPi) deep brain stimulation is reported in a patient with severe delayed onset post-traumatic cervical dystonia. He had sustained severe head trauma at the age of 17 and had developed a mild right hemiparesis. Three years after the head injury, cervical dystonia with head turning to the left side developed. Magnetic resonance imaging (MRI) showed a discrete GPi lesion on the left side. At the age of 23, he underwent unilateral left GPi deep brain stimulation. He experienced immediate but short lasting benefit from the microlesioning effect of the electrode. With activation of deep brain stimulation, there was significant improvement of the cervical dystonia, persisting for 12 months of follow up. This case underlines the importance of the globus pallidus internus in the generation and amelioration of cervical dystonia.

The pathophysiology of cervical dystonia is not well understood, and the mechanism of improvement associated with stereotactic surgical procedures is not clear either.^{1–3} The rationale for globus pallidus internus (GPi) stimulation in dystonia is partly based on the observation of an improvement in L-DOPA induced involuntary movements by pallidotomy or pallidal stimulation in patients with Parkinson's disease. Although a role of the GPi in the pathogenesis of dystonia is strongly suspected, presumably acting through alteration of thalamic inhibition,⁴ focal traumatic lesions of the GPi have only rarely been reported to produce secondary dystonia.

In this paper, we describe a patient who sustained a motor vehicle accident that eventually led to a focally restricted lesion of the GPi with cervical dystonia, which was successfully treated by unilateral GPi stimulation.

CASE REPORT

A 23 year old man sustained a severe closed head injury in a car accident at the age of 17. He had a right hemiparesis and was obtunded for several weeks before recovering slowly over the next few months. Three years after the initial head injury, cervical dystonia developed, and this progressed in severity for six months. The cervical dystonia did not improve with medical treatment, which included clonazepam, L-DOPA, baclofen, and trihexyphenidyl. Two years later, he was treated with botulinum toxin injections to the neck, but there was only a transient ameliorating effect. There was no family history of movement disorders. The patient was referred to our institution for further evaluation and management.

Examination

On admission at the age of 23, the patient's general condition was unremarkable. He was fully oriented, without memory

deficits, but had a mild right sided hemiparesis. His gait was hemiparetic, and he had mild dysarthria. His head was turned towards the left side. When lying on his back, he was able to turn his head to the neutral position by using trick manoeuvres: he could follow the movement of his right finger visually and turn his head toward the midline; he was also able to turn his head by lightly contacting his face with his palm.

Neuroimaging

Initial computed tomography (CT) of the brain obtained after the car accident showed a traumatic subarachnoid haemorrhage in the quadrigeminal cistern and contusional haemorrhage in the left basal ganglia (fig 1A). Brain magnetic resonance imaging (MRI) done five days after the initial injury showed diffuse brain oedema around the left basal ganglia and right thalamus (fig 1B). Six years after the initial injury, brain MRI revealed a confined and small lesion in the left GPi (fig 1C). Brain CT obtained after electrode implantation for deep brain stimulation showed that the electrode was positioned in the left posteroventral GPi just medial and anterior to the lesion (fig 1D).

Operation

A Leksell stereotactic frame was used to implant an electrode into the GPi. A tentative target was determined using customised software, which automatically calculated the target from the magnetic resonance image. The tentative target was set at 21 mm lateral to and 3 mm below the intercommissural line and 2 mm anterior to the midcommissural point. Microelectrode confirmation at the GPi target was undertaken as previously described.⁵ Conventional single and multiunit extracellular recordings were made with a gold and platinum plated tungsten electrode insulated with Parylene-C. The mean discharge rate of neurones in both segments of the globus pallidus was considerably lower than that reported in idiopathic Parkinson's disease in previous studies^{6,7} (data not shown).

The final target was just medial and anterior to the lesion in the left GPi. A local anaesthetic was given and a precoronal burr hole was made on the right side 2.5 cm lateral to the midline. A deep brain stimulation electrode (Medtronic, Minneapolis, Minnesota, USA) was then inserted into the target using a guiding cannula. Electrical stimulations were performed to assess the proximity of the internal capsule and the optic tract. These stimulations resulted in a mild decrease in EMG activity in the right sternocleidomastoid muscle (fig 2). The electrode was fixed to the burr hole, connected to the IPG (Itrel II, model 7424; Medtronic), and implanted in a subcutaneous pouch in the left subclavicular region under general anaesthesia.

Postoperative course

Immediately after the operation, the patient was able to turn his head to the midline without any sensory tricks while lying

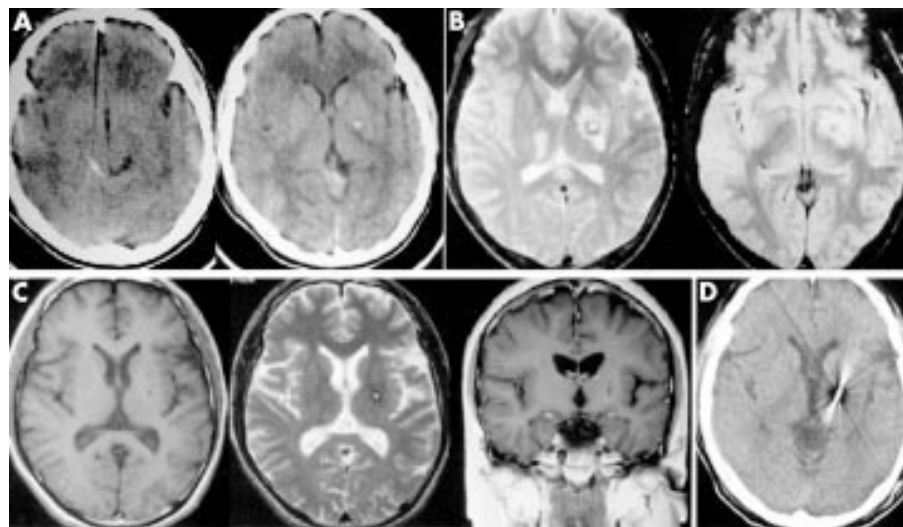


Figure 1 Brain imaging studies throughout the patient's clinical course. (A) Initial brain computed tomography (CT), showing a traumatic subarachnoid hemorrhage in the quadrigeminal cistern and contusional hemorrhage in the left basal ganglia. (B) Magnetic resonance imaging (MRI) obtained five days after the initial injury, showing diffuse brain oedema around the left basal ganglia and right thalamus. (C) MRI images obtained six years after the initial injury showing a small confined lesion in the left sided GPI. (D) Postoperative CT showing appropriate positioning of the electrode.

on his back. However, his ability to turn his head to the midline while standing or sitting did not improve. Seven days after implantation of the stimulating electrode, the initial improvement of head turning ceased. One month after implantation, deep brain stimulation was begun and the patient again showed improvement in head turning. His head turning ability then improved gradually up to six months after the operation.

The initial setting of the stimulation parameters was as follows: amplitude 2 V, pulse width 180 μ s, frequency 160 Hz. The patient was able to turn his head to the midline voluntarily without sensory tricks while lying on his back. In addition, he could now turn his head to the midline while standing or sitting, although with considerable effort and slowness.

Three months after electrode implantation, the cervical dystonia was stable. We explored various IPG parameters, but no further improvement in symptoms was observed over 12 months of postoperative follow up. Functionally, the patient could enjoy watching television and eat without too much effort, which he could not do before the surgery. There were no adverse effects associated with the operation.

DISCUSSION

Post-traumatic movement disorders secondary to severe head injuries have been reported in 22.6% of surviving patients.⁸ Movement disorders were described as being transient in 10.4% and persistent in 12.2% during a mean follow up period of 3.9 years. Dystonia was observed in 4.1% of these patients. Hemidystonia is the most common presentation of dystonia caused by brain trauma, and is typically manifested by a delayed onset. In most cases of dystonia, contralateral lesions are found in the caudate, putamen, or thalamus.^{8,9} The clinical features of delayed onset post-traumatic cervical dystonia are indistinguishable from those of idiopathic cervical dystonia in terms of the gradual progression of symptoms and signs and the development of sensory tricks and activation manoeuvres. Both conditions are distinct from acute onset post-traumatic cervical dystonia.³

Most cases of delayed dystonia secondary to various acquired conditions have been associated with pathological lesions in the putamen, and cases that have been attributed to lesions of the globus pallidus have been extremely rare.

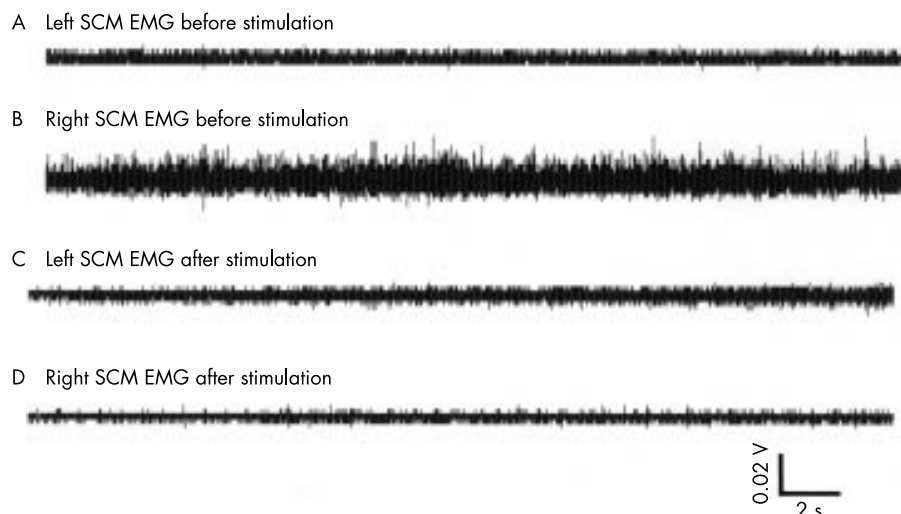


Figure 2 Electromyographic activity in the sternocleidomastoid muscle (SCM) on both sides while the patient was sitting with his neck in its natural position of almost 90° turned to the left side (before and after deep brain stimulation).

Involvement of the GPi, particularly unilaterally, was most unusual in reviews of reports from around the world.^{4 10 11} King *et al* described the delayed onset of hemidystonia and hemiballismus following an isolated lesion of the GPi and presumed that the cause of the symptom was partial preservation of the Gpi.⁴ They suggested that the partial involvement of the GPi may have provided the pathological basis for the overall progression of the symptoms.

It is generally believed that dystonia results from the relay of erroneous information from the thalamus to the premotor, supplementary motor, and probably the primary motor cortices. This could be attributed to pathological changes at several sites, including the putamen, the globus pallidus, the thalamus itself, and rarely the caudate nucleus.⁴ It is not clear why a lesion of the putamen is the most common anatomical site producing dystonia.^{12 13} Experience with isolated lesions of the globus pallidus has not only demonstrated that they may cause dystonia, but has also shown that they may cause parkinsonism, behavioural disorders, or no clinical dysfunction.^{3 4 10}

Lesioning or deep brain stimulation of the GPi has been used successfully to treat both parkinsonian dyskinesia and dystonia.^{6 7 14-18} The rationale for pallidotomy or pallidal stimulation in cases of Parkinson's disease may be the removal of excessive output from the GPi, which has been shown to occur in this condition. On the other hand, a slower than normal firing rate in the GPi and a lower than normal thalamic metabolic rate are noted in dystonia. Thus the mechanism of the postoperative amelioration of dystonia after pallidal stimulation has not been established, but may involve abolition of the abnormal firing pattern rather than a simple decrease of the increased firing rates.^{4 15}

Experience with pallidal stimulation for focal dystonias including cervical dystonia is very limited. Islekel *et al* reported that a unilateral pallidal lesion or stimulation was an effective method of treatment in focal dystonia.¹⁶ They emphasised that the target must be the pallidum contralateral to the contracted sternocleidomastoid muscle. Parkin and colleague presented data to show that bilateral GPi stimulation led to a progressive improvement over several months in the pain, voluntary head movement, and abnormal posture caused by spasmodic torticollis.¹⁸ Kulisevsky *et al* reported two patients with idiopathic cervical dystonia with only mild motor improvement but marked amelioration of pain symptoms following bilateral deep brain stimulation of the posteroventral GPi. The wide dissociation between pain and motor improvement in their patients suggested that dysfunction of motor and somatosensory circuits in dystonia did not always proceed in parallel.¹⁹

Our patient improved after unilateral deep brain stimulation on the same side of the initial abnormal lesion, suggesting that abnormal neuronal activity resulted from the initial lesion and was ameliorated by subsequent deep brain stimulation in the adjacent area of the same structure.

Conclusions

We report the beneficial effect of deep brain stimulation in the GPi in a patient with delayed onset post-traumatic cervical dystonia. Consistent with the clinical features, which are identical to those of idiopathic cervical dystonia, the response to GPi stimulation suggests a common central pathogenic mechanism and shared anatomical features between delayed onset post-traumatic cervical dystonia and idiopathic cervical dystonia. In addition, the unilateral nature of the lesion and

the unilateral placement of deep brain stimulation in the GPi underlines the central importance of that structure in the generation and amelioration of cervical dystonia.

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