Case Report

Spondylotic myelopathy in patients with cervical dystonia

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Abstract

The treatment and outcome of cervical dystonia (CD) with myelopathy (CDM) varies among studies. We retrospectively reviewed the clinical features, neuroimages, methods of treatment, and modified Rankin scale (mRS) score of patients with CDM in our hospital. There were seven male and three female patients, with a mean age of 53 years. The mean age at onset of CD was 24.9 years, with a mean interval from dystonia to the diagnosis with myelopathy of 28.1 years. The dominant level of cord compression was at C3/C4 in seven patients, C4/C5 in one patient, and C5/C6 in two patients. Four of five patients with an initial mRS ≥ 3 remained moderately to severely disabled after surgery, with a follow-up periods of 3–5 years, and one of them experienced recurrent cage dislocation and neurological deterioration due to an unstable spine. Peri-operative stabilization is important to minimize neurological sequelae. Whether surgical intervention improves the functional outcome of patients with moderate-to-severe disability demands further randomized controlled studies.

Keywords: botulinum toxin; dystonia; neurosurgery; spinal cord compression; treatment outcome

1. Introduction

Cervical dystonia (CD) may precipitate disc degeneration, cause cervical cord compression, and result in neurological deterioration. 1–3 However, the treatment and outcome of CD with myelopathy (CDM) vary among studies. 1,4–6 We retrospectively reviewed the medical records of all patients with CDM who visited our hospital between January 1, 2000 and June 30, 2009, and evaluated the treatments and outcomes.

2. Case reports

Patients who had prior use of botulinum toxin injections (BTX), dystonia affecting more body parts than the head and neck, a duration from dystonia to the diagnosis of myelopathy of less than 2 years, or a history of cervical trauma before the diagnosis of cervical spondylosis were excluded.

Myelopathy was defined as magnetic resonance imaging (MRI)-proven spondylotic cervical cord compression with increased deep tendon reflexes of the lower limbs and an bilateral extensor response in the plantar reflexes with or without a sensory level, limb weakness, spastic gait, or incontinence. Radicular pain was defined by pain radiating along the dermatome. Sensory impairment was defined as impaired sensation either with a sensory level or by radicular distributions.

In total, the medical records of 10 patients with CDM were systematically assessed and recorded, including clinical features, neuroimages, methods of treatment, and functional outcome assessments using the modified Rankin scale (mRS).

2.1. Case 1

A 36-year-old man with athetoid cerebral palsy had had involuntary movement of the neck and grimacing since the age...
of 1 year. At the age of 17, he experienced an electrical sensation that ran down his back on bending his neck forward. The neck pain gradually improved after rehabilitation therapy. He remained independent until the age of 36, when weakness of his legs occurred, with frequent, unexpected falls. Cervical cord compression at the C3/C4 level was diagnosed at another hospital, and he received anterior discectomy and interbody fusion cage without perioperative stabilization. Nevertheless, his already unsteady gait deteriorated in the following 3 months, and urinary incontinence developed.

The patient visited our hospital, where neurological examination disclosed a spastic gait and a sensory level at T5. A cervical MRI scan disclosed cage dislocation and cord compression at C3/C4. The cage was removed, and the patient received an autogenous bone graft and internal fixation with a plate. A hard neck collar and BTX (Dysport 600 U at 3-monthly intervals) was also prescribed for perioperative stabilization.

However, the patient’s spine remained unstable, and he experienced two more episodes of plate dislocation in the first and third years after the first surgery at our hospital, later undergoing two more fixation operations. Five years after the first surgery at our hospital, he became wheelchair-bound.

2.2. Case 2

A 63-year-old man with primary CD had had involuntary movement of his neck since the age of 61. He was well except for painful contractions in his neck. Two years later, he experienced intermittent, lancinating pain in his right forearm triggered by neck movement. He was at this point living independently and walking without a cane.

Increased deep tendon reflexes of the lower limbs and bilateral extensor responses of the plantar reflexes were noted. Other neurological examinations were unremarkable. Cervical MRI showed posterior disc herniation with cord compression at C3/C4. Because the CD was responding poorly to anticholinergics and muscle relaxants, BTX (Dysport 600 U at 3-monthly intervals) was prescribed. The involuntary forceful rotation of his neck improved. The patient remained stable without any neurological disability in the follow-up period of 6 years.

3. Patient summary

In total, there were seven male and three female patients with a mean age of 53 years, a mean age at onset of CD of 24.9 years, and a mean interval from dystonia to the diagnosis with myelopathy of 28.1 years. The dominant level of cord compression was at C3/C4 in seven patients, C4/C5 in one patient, and C5/C6 in two patients. The underlying causes of CD were primary CD in five patients and athetoid cerebral palsy in four. One patient, who had tardive dystonia caused by taking sulpiride 200 mg per day, stopped sulpiride after the diagnosis with myelopathy was made at our hospital.

Six patients received surgical intervention with a hard neck collar after surgery, and three of these also received perioperative BTX. Three of five patients with an initial mRS ≥ 3 showed an improved mRS score after surgery, but two of them remained moderately to severely disabled. The other two patients with an initial mRS ≥ 3 showed a worsened score after surgery and became severely disabled. One of five patients with an initial mRS of 1 had an improved score and remained stable after surgery. The other four patients with an initial mRS of 1 received only BTX and were able to live independently (over follow-up periods of 4–9 years).

4. Discussion

Perioperative stabilization is important for patients with CDM in order to achieve solid fusion and minimize neurological sequelae. Without a good control of involuntary movement, conventional surgical treatment for CDM is not beneficial and can even be harmful, owing to additional cervical instability after surgery. Postoperative stabilization is largely achieved by a neck collar or BTX. However, to our knowledge, no controlled study has been conducted on the efficacy of BTX in the treatment of CDM with mild neurological deficit. Chronic pallidal stimulation, which relieves dystonia, is an useful adjunct to provide adequate postoperative stabilization for patients who have failed medical management.

Four of five patients with an initial mRS ≥ 3 remained moderately to severely disabled after their operation. Previous studies also showed that patients who required a wheelchair still ambulated with help after surgery. In contrast, patients who were not wheelchair-bound showed an improvement in ambulation after surgery. The benefit of surgery for patients with CDM with moderate-to-severe disability needs to be subjected to further randomized controlled studies.

The most commonly involved level of cervical cord compression was C3/C4, which is compatible with previous studies. In the human cervical spine, flexion and extension, which are common in normal persons, are greatest at the C5/6 level, while lateral bending and axial rotation, which are common in patients with CD, are greatest at the C3/C4 and C4/C5 levels. Cervical myelopathy may rarely occur above the C2 level in patients with congenital atlas stenosis.

Cervical stabilization is important for all patients with CDM and can be achieved by BTX, or in some patients by chronic pallidal stimulation. Whether surgical intervention improves the functional outcome of patients with moderate-to-severe disability should be studied further in randomized controlled trials.

References


