Unexpected outcome (positive or negative) including adverse drug reactions

Reversal of lifelong mutism after anterior cervical discectomy and fusion for myelopathy

Ranjith Babu, Betsy H Grunch, Carlos A Bagley, Oren N Gottfried

Division of Neurosurgery, Duke University Medical Center, Durham, North Carolina, United States

Correspondence to Mr Ranjith Babu, ranjith.babu@duke.edu**Summary**

A 34-year-old man with a history of spina bifida occulta, fetal alcohol syndrome and mutism presented with an acute on chronic decline of unsteady gait and right arm and leg weakness over the period of a few months. The patient was non-verbal and communicated using hand gestures. MRI of the cervical spine showed severe stenosis at C4-5 with T2 signal abnormalities. Brain MRI demonstrated mild ventriculomegaly. The patient underwent an anterior cervical discectomy and fusion for severe cervical spine stenosis. Postoperatively the patient's myelopathic symptoms improved. He also became verbal and engaged in conversation. Ten months after surgery, the patient who had previously been non-verbal had developed a vocabulary of more than 50 words.

BACKGROUND

Cervical spinal stenosis is associated with a variety of neurological disorders including neurapraxia and myelopathy.¹⁻³ However, the neuropsychological impairment from disorders of the cervical spine is less recognised and poorly understood. Several studies have shown a decrease in cognitive performance after cervical spine injury including deficits in problem-solving ability and attention particularly in spinal cord injury.⁴⁻⁷

Interestingly, neuropsychological deficits as a result of spinal cord compression may improve after posterior cervical decompression.⁸⁻⁹ We present a unique case of a 34-year-old previously non-verbal male with severe signs of myelopathy and cervical stenosis that had significant neuropsychological improvement and new ability to speak and verbally communicate after anterior cervical decompression surgery. We discuss several hypotheses to explain this improvement.

Figure 1 (A) Sagittal T2 weighted MRI of the cervical spine revealing cervical stenosis, most severe at C4-5 with T2 hyperintensities. (B) Axial T1 weighted MRI of the brain demonstrating mild ventriculomegaly without mass lesion. (C) Postoperative sagittal T2 weighted MRI of the cervical spine revealing interval decompression of the spinal cord and canal at C4-5 with expected increased signs of myelomalacia. (D) Postoperative axial T1 weighted MRI of the brain revealing unchanged ventricular size.

CASE PRESENTATION

A 34-year-old male with a history of fetal alcohol syndrome and mutism presented to the emergency department due to worsening of unsteady gait and right arm and leg weakness over the period of a few months. Since birth, the patient communicated using few hand gestures and did not speak. His neurological examination included right lower extremity hip flexion to be 2/5 and the quadriceps, dorsiflexion and plantar flexion were 4-/5. He had 5/5 strength in the left lower extremity. He had sustained clonus in his right lower extremity and was hyperreflexive at 3+ in the upper and lower extremities.

INVESTIGATIONS

MRI of the brain and cervical spine were performed. MRI of the cervical spine showed severe stenosis at C4-5 with T2 hyperintensities (figure 1A). MRI of the brain revealed mild ventriculomegaly without mass lesion (figure 1B).

TREATMENT

The patient underwent a C4-5 anterior cervical discectomy and fusion.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient began to speak and form words a few days after surgery, and over several weeks, began to engage in conversation. He started with simple words and gradually developed the capability to form full sentences. At 10 months after surgery, he has a spoken vocabulary of more than 50 words. Also, he has the ability to make jokes and express happiness. Moreover, his gait also improved. His muscle strength improved on the right to 4/5 in hip flexion, 4+/5 in the quadriceps, dorsiflexion and plantar flexion. In the right upper extremity, he was 4+/5 throughout, except 4/5 in the intrinsic hand muscles, and left upper extremity improved to full strength. Recently, he started cognitive, physical and occupation therapy to further promote his recovery. Postoperative MRI of the cervical spine at 8 months revealed interval decompression of the spinal cord and canal at C4-5 with expected increased signs of myelomalacia (figure 1C), and MRI of the brain was unchanged (figure 1D).

DISCUSSION

To date there are only two studies specifically investigating improvement of cognitive function after surgical intervention for cervical spine disorders. The first study by Rosner *et al* involved 16 patients with Chiari syndrome and/or congenital cervical stenosis who had been offered decompressive craniocervical surgery.⁸ The patients in the study underwent pre- and postoperative regional cerebral blood flow (rCBF) studies using single photon emission CT and answered questions regarding neuropsychological complaints. Postoperatively, the patients demonstrated a generalised bi-hemispheric increase in cerebral blood flow. Also, left frontal lobe blood flow increased by approximately 10%, with 4% and 12% increases in the right occipital and

parietal lobes, respectively. Patients showed an increase in concentration and memory.

Another study included 79 patients with cervical myelopathy who underwent cervical laminoplasty for decompression of the cervical cord and/or cervical nerves.⁹ Ten of these patients with memory impairment underwent neuropsychological and neuroimaging testing. Preoperatively, it was noted that 8 out of 10 patients had decreased cerebral blood flow in the parietal and/or occipital lobes. However postoperatively, regional cerebral blood flow was shown to have increased in 50% of patients. Also, the authors showed that there was a statistically significant improvement in performance on the Kohs Block Design Test (which tests visuospatial organisation) and the Benton Visual Retention Test (which tests memory and visuo-construction skills) after surgery.

Currently, the precise mechanism of impaired cerebral blood flow due to spinal cord compression is unclear. One explanation is that cord compression disrupts the sympathetic nerves from the superior cervical ganglion, and somatosensory input, which both control cerebral blood flow.^{10 11} Another explanation for the decreased blood flow is cerebrospinal fluid (CSF) flow obstruction as a result of spinal canal compression. The Monro-Kellie hypothesis states that the intracranial volume is fixed, and as the sum of volumes of brain, CSF, and intracranial blood is constant, an increase in one results in the decrease in one or two of the remaining components.¹² This has been shown as arterial pulsations (which result in changes in intracranial blood volume) are compensated by the bi-directional flow of CSF through the foramen magnum.¹³ When a region of the brain is activated, it is accompanied by an increase in regional cerebral blood volume (CBV) due to the increased metabolic activity. Therefore when there is cerebral activation, the increase in CBV should be compensated by transient CSF flow into the spinal canal. This process may not occur if there is spinal canal obstruction, preventing CSF flow across the foramen magnum and would prevent the required increases in cerebral blood flow volume, thereby impairing cognitive function. Reversal of the spinal canal obstruction by decompressive surgery allows for this transient CSF flow into the canal, allowing for appropriate increases in CBV during activation.

Additionally, the neuropsychological improvement seen after decompressive surgery may also be due to cortical reorganisation. A study by Holly *et al* demonstrated that areas of motor and somatosensory cortex become enlarged and less focused in patients with cervical spondylotic myelopathy.¹⁴ Functional MRI (fMRI) was used to identify areas of cortical activation in four patients with cervical spondylotic myelopathy. It was shown that decompressive surgery resulted in neurological improvements and more focused patterns of cortical activation resembling those of healthy volunteers. This cortical reorganisation is thought to be due to synaptic modification of pre-existing connections and/or development of new connections. These changes may also occur in other cortical areas and contribute to an improvement in neuropsychological functioning.

Learning points

- ▶ Cervical spine disorders may result in decreased neuropsychological functioning.
- ▶ Cervical decompression may reverse neuropsychological impairments caused by spinal cord compression.
- ▶ The mechanism for this improvement may be due to increases in cerebral blood flow or cortical reorganisation as a result of decompressive surgery.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Torg JS**, Corcoran TA, Thibault LE, *et al*. Cervical cord neurapraxia: classification, pathomechanics, morbidity, and management guidelines. *J Neurosurg* 1997;**87**:843–50.
2. **Hashimoto I**, Tak YK. The true sagittal diameter of the cervical spinal canal and its diagnostic significance in cervical myelopathy. *J Neurosurg* 1977;**47**:912–6.
3. **Grüniger W**, Gruss P. Stenosis and movement of the cervical spine in cervical myelopathy. *Paraplegia* 1982;**20**:121–30.
4. **Radanov BP**, Bicik I, Dvorak J, *et al*. Relation between neuropsychological and neuroimaging findings in patients with late whiplash syndrome. *J Neurol Neurosurg Psychiatr* 1999;**66**:485–9.
5. **Radanov BP**, Dvorák J, Valach L. Cognitive deficits in patients after soft tissue injury of the cervical spine. *Spine* 1992;**17**:127–31.
6. **Hohmann GW**. Some effects of spinal cord lesions on experienced emotional feelings. *Psychophysiology* 1966;**3**:143–56.
7. **Roth E**, Davidoff G, Thomas P, *et al*. A controlled study of neuropsychological deficits in acute spinal cord injury patients. *Paraplegia* 1989;**27**:480–9.
8. **Rosner MJ**, Johnson A, Rosner SE. Craniocervical Decompression, Cerebral Blood Flow and Neuropsychological Dysfunction in FMS and CFS. National Fibromyalgia Research Association's Subgroups in Fibromyalgia Symposium, 26–27 September 1999, Portland, Oregon.
9. **Hoshimaru M**. Neuropsychological improvement in patients with cervical spondylotic myelopathy after posterior decompression surgery. *Neurol Med Chir (Tokyo)* 2010;**50**:554–9.
10. **Adachi T**, Meguro K, Sato A, *et al*. Cutaneous stimulation regulates blood flow in cerebral cortex in anesthetized rats. *Neuroreport* 1990;**1**:41–4.
11. **Van Lieshout JJ**, Wieling W, Karemaker JM, *et al*. Syncope, cerebral perfusion, and oxygenation. *J Appl Physiol* 2003;**94**:833–48.
12. **Mokri B**. The Monroe-Kellie hypothesis: applications in CSF volume depletion. *Neurology* 2001;**56**:1746–8.
13. **Shibuya R**, Yonenobu K, Koizumi T, *et al*. Pulsatile cerebrospinal fluid flow measurement using phase-contrast magnetic resonance imaging in patients with cervical myelopathy. *Spine* 2002;**27**:1087–93.
14. **Holly LT**, Dong Y, Albistegui-DuBois R, *et al*. Cortical reorganization in patients with cervical spondylotic myelopathy. *J Neurosurg Spine* 2007;**6**:544–51.

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