Spinal Cord (2005) 43, 249–251

www.nature.com/sc

Case Report

Foramen magnum decompression for the treatment of Arnold Chiari malformation type I with associated syringomyelia in an elderly patient

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Study design: Report of a rare case of an elderly patient with late onset of Arnold Chiari malformation type I with associated syringomyelia that was successfully treated with foramen magnum decompression.
Objective: To report this rare case along with a literature review.
Setting: Gifu, Japan.
Methods: A 69-year-old woman with a 4-year history of dull pain in her right arm was referred to the clinic. After physical and radiographical examinations, she was diagnosed with Arnold Chiari malformation type I with associated syringomyelia. A foramen magnum decompression by the removal of the outer layer of the dura mater was performed.
Results: At 2 years postoperatively, MRI revealed a decrease in the size of the syringomyelia. Her symptoms had also remarkably improved.
Conclusions: A rare case of symptomatic Arnold Chiari malformation type I with associated syringomyelia in an elderly woman was successfully treated with foramen magnum decompression by the removal of the outer layer of the dura mater.
Spinal Cord (2005) **43,** 249–251. doi:10.1038/sj.sc.3101675; Published online 2 November 2004

Keywords: Arnold Chiari malformation type I; syringomyelia; foramen magnum decompression; elderly; surgical treatment

Introduction

In Arnold Chiari malformation type I, abnormal cerebrospinal fluid flow occurs and frequently results in syringomyelia.[1,2] Various surgical methods for treating neurologically symptomatic cases of syringomyelia associated with Arnold Chiari malformation type I have been reported.[1,3–7] Posterior fossa craniectomy (foramen magnum decompression), with[1] or without plugging of the obex,[3,8,9] has been one of the most commonly performed surgical procedures in the belief that it would ameliorate the abnormal cerebrospinal fluid flow. However, this surgical procedure has been primarily indicated for treating relatively young and middle-aged patients.[1,3,4,6,10] Few reports are available on the late onset of symptoms associated with Arnold Chiari malformation type I in elderly patients and possible surgical treatments.[8,11] This report is of a very rare case of an elderly woman with late-onset Arnold Chiari malformation type I and syringomyelia that was successfully treated with foramen magnum decompression.

Case report

In May 2000, a 69-year-old woman was referred to the clinic. She had experienced a history of dull pain in her right arm for 4 years. Additionally, dorsalgia and bilateral leg pain radiating from her gluteal region appeared 1 year prior to her evaluation. Owing to the dorsalgia and leg pain, her gait became gradually disturbed. On her first visit to the clinic, the neurological examination revealed sensory disturbances in her right arm, trunk and thigh. Deep tendon reflexes were depressed in the right arm and exaggerated in both legs. Plain X-ray of the cervical spine showed moderate degenerative changes. MRI showed cerebellar tonsillar herniation into the foramen magnum and syringomyelia from the medulla oblongata to the T11 level (Figure 1). However, compression of the spinal cord due to spondylotic change or disc herniation at the cervical spine level was not observed. Therefore, she was

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Figure 1 Preoperative T1-weighted MRI scan of the occipitocervical junction and the cervical spine. Sagittal section just at the midline of the spinal cord demonstrates syringomyelia from medulla oblongata to upper thoracic levels

Figure 2 Postoperative T1-weighted MRI scan of the occipitocervical junction and cervical spine. Sufficient decompression at the foramen magnum, upper deviation of cerebeller tonsil and remarkable reduction in size of the syringomyelia are shown

diagnosed with symptomatic Arnold Chiari malformation type I with associated syringomyelia.

Surgical procedure

Foramen magnum decompression by the removal of the outer layer of the dura mater was performed, as reported by Isu *et al*[6] in 1993. First, a suboccipital craniectomy and removal of the arch of C1 was performed. It is worth noting that in a suboccipital craniectomy, the occipital bone is removed far laterally.[6] A cruciate incision was made in the outer layer of the dura mater. The outer layer was dissected from the inner layer and removed. Immediately after this removal, the inner layer of the dura mater began bulging and the cerebellar tonsil was pulsating. Therefore, the effectiveness of decompression at the foramen magnum was immediately ascertained.

At 1 month postoperatively, the sensory disturbances in her right arm and both legs improved and her difficulty in walking disappeared. The dorsalgia and bilateral leg pain radiating from her gluteal region were remarkably reduced. No perioperative complications occurred. In the postoperative MRI, both the upper deviation of the cerebellar tonsil and a reduction in size of the syrinx were recognized (Figure 2). After a 2-year follow-up, her postoperative course has been excellent.

Discussion

According to the literature, the clinical condition of this patient is doubly rare and worth reporting. First, because the Arnold Chiari malformation type I with associated syringomyelia initially manifested its symptoms at an advanced age, and second, because a satisfactory surgical outcome following foramen magnum decompression was achieved in an elderly patient.

There are several reports on symptomatic Arnold Chiari malformation type I associated with syringomyelia.[1,3,8–10,12,13] Among these, a few authors presented cases with late onset of symptoms due to Arnold Chiari malformation type I-associated syringomyelia.[8,12] Moriwaka *et al*[14] reported that the average age of onset of the first symptoms due to syringomyelia was 28 -years old and that syringomyelia due to an Arnold Chiari malformation showed a tendency to have a younger age of onset.

In the case reported here, both MRI and CT-myelogram revealed that there was no compression of the spinal cord due to degenerative changes in the cervical spine. Therefore, foramen magnum decompression was chosen to treat the patient, with the expectation

that this procedure would improve the cerebrospinal fluid flow and alleviate her symptoms.

There is apparently only one other report, by Geroldi *et al*,[11] of the successful surgical treatment of an elderly patient using foramen magnum decompression for Arnold Chiari malformation type I with associated syringomyelia. That case differs from the present one in that a cervico-spinal decompression was performed, although the surgical procedure was not precisely described. In the current case, because there was no observable compression of the cervical spinal cord, foramen magnum decompression by the removal of the outer layer of the dura mater[6] was performed. Among the various methods for foramen magnum decompression, this method has several advantages. First, it is less invasive in comparison with other previously reported procedures. Second, the reported incident rate of readhesion is low, and third, the decompression effect is maintained for a long period because the inner layer of the dura mater is preserved.[15] In addition, because this foramen magnum decompression procedure is extradural, there are no demonstrated risks of such postoperative complications[6] as cerebrospinal fluid collection in the operative wound,[4] pseudomeningocele[10] or meningitis.[9,16] Therefore, the authors are convinced that this method is advantageous when performing a foramen decompression in an elderly patient.

In a report by Isu *et al*,[6] the oldest patient who underwent this procedure was 54 years old. This patient presented with symptoms induced by a cervical lesion at 65 years of age. In elderly patients, cervical myelopathy is encountered frequently in clinical practice and is difficult to differentiate from other conditions by a physical examination and plain X-ray alone. The symptoms of Arnold Chiari malformation type I with associated syringomyelia vary and this clinical condition has no pathognomonic clinical sign.[8,17] In the current patient, the MRI scan from the occipitocervical junction down to the cervical spine was very useful in making a correct diagnosis. Moreover, the MRI scan revealed the postoperative reduction of the syringomyelia, which corresponded well with the alleviation of her symptoms. Awareness of symptomatic Arnold Chiari malformation type I-associated syringomyelia in elderly patients would help clinicians make correct diagnoses.

Conclusion

A rare case of Arnold Chiari malformation type I associated with syringomyelia was reported in an elderly woman, who presented with sensory disturbances and pain in her right arm, trunk and both legs. She was successfully treated by foramen magnum decompression by the removal of the outer layer of the dura mater.

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