Chiari I malformation associated with syringomyelia: can foramen magnum decompression lead to restore cervical alignment?

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Abstract

Purpose There are few researches that demonstrate the relationship between the extent of syringomyelia and sagittal alignment of the cervical spine. The purpose of this study is to investigate the correlation between the change of syrinx size and cervical alignment.

Methods From January 2001 to June 2008, we operated on 207 patients who had syringomyelia. The associated diseases were categorized by Chiari I malformation, tumor, trauma, spinal stenotic lesion, inflammatory disease and idiopathic causes. Thirty patients who had Chiari I malformation associated with syringomyelia and who underwent foramen magnum decompression (FMD), participated in this study. We excluded patients with scoliosis, cervical instrumentation, tumor, trauma, myelomeningocele, hydrocephalus, tethered cord and congenital vertebral

anomalies. Lateral radiographs in neutral and magnetic resonance imaging were taken pre- and postoperatively.

Results Mean follow-up was 6.5 ± 1.5 years (ranged from 4.0 to 9.5 years). The mean pre- and postoperative lordosis angles at C2–C7 were $-5.9^\circ \pm 1.0^\circ$ and $-10.4^\circ \pm 1.0^\circ$, respectively ($P = 0.001$). There was significant correlation between the differences of syrinx width and the cervical lordotic angles before and after surgery ($P = 0.016$). After FMD, syringomyelia and cervical alignment improved in 28 (93.3 %) and 25 (85.18 %) of 30 patients, respectively. There was significant correlation between recovery rate by Japanese Orthopaedic Association scores and the difference of the cervical lordotic angles before and after surgery ($P = 0.022$).

Conclusions The present results demonstrate that the decrease of syrinx size by FMD may restore the cervical lordosis. We suggest that the postoperative cervical alignment might be a predictive factor for neurological outcome.

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Keywords Cervical spinal alignment · Chiari malformation · Foramen magnum decompression · Syringomyelia

Abbreviations

FMD Foramen magnum decompression
MRI Magnetic resonance imaging
JOA Japanese Orthopaedic Association

Introduction

The syringomyelia is associated with various etiologies including Chiari I malformation and shows diverse clinical characteristics according to their associated

diseases [1, 10, 12, 17, 20]. Several studies have revealed that Chiari I malformation and syringomyelia are associated with scoliosis [1, 5, 9, 21]. For syringomyelia associated with Chiari I malformation, surgical procedure such as the syringoperitoneal shunt, syringosubarachnoid shunt, or duroplasty is performed. In recent years, foramen magnum decompression (FMD) has been conducted more frequently for the disease [2, 15, 21]. Foramen magnum decompression could improve neurological status and scoliosis often in those patients [3, 11, 15]. It means that syringomyelia may be an aggravating factor of spinal alignment. However, there are few researches that demonstrate the relationship between the extent of syringomyelia and sagittal alignment of the cervical spine. We investigated the relationship between the change of syrinx size and cervical alignment.

Materials and methods

From January 2001 to June 2008, we operated on 207 patients who had syringomyelia. All of their syringomyelias were confirmed by magnetic resonance (MR) imaging and we reviewed their medical records and radiological data, retrospectively. The associated diseases were categorized by Chiari I malformation, tumor, trauma, spinal stenotic lesion, inflammatory disease and idiopathic etiologies. Thirty patients who had Chiari I malformation associated with syringomyelia and who underwent FMD, participated in this study. We excluded patients with scoliosis, cervical instrumentation, tumor, trauma, myelomeningocele, hydrocephalus, tethered cord and congenital vertebral anomalies. The definition of Chiari I malformation in the present study was that the cerebellar tonsil descended more than 5 mm below the foramen magnum.

All patients underwent FMD, C1 laminectomy and expansile duroplasty using the artificial dura mater (Fig. 1). The neck extensor muscles arising from the spinous process of C2, such as the rectus capitis posterior major and semispinalis cervicis were not dissected. All bone and dural works were performed under a microscope. Postoperative fixation with a cervical collar was omitted. An adjuvant syringosubarachnoidal shunt was placed in three patients.

Lateral radiographs in neutral and MR imaging were taken pre- and postoperatively. The angles created by a line parallel to the inferior aspect of the C2 body and a line parallel to that of the C7 body were measured on the lateral radiographs in neutral position before surgery and at the final follow-up evaluation (Fig. 2d). The negative value of the angle indicated lordosis and the positive meant kyphosis. The pre- and postoperative size of syrinx was determined in T2-weighted sagittal view of MRI at their widest level before surgery and at the final follow-up evaluation. Difference

ratio according to the width of syrinx, which indicates the degree of normalization after surgery, was calculated using the following formula: (preoperative width – postoperative width)/(preoperative width). Difference ratio “0” means no improvement of the syrinx size, whereas the value “1” means complete disappearance of the syrinx.

The preoperative stage of cerebellar tonsillar descent was classified into the following three grades by MR imaging: Grade 1, the tonsil descends over the foramen magnum but does not reach the C1 arch; Grade 2, the tonsil reaches the C1 arch; Grade 3, the tonsil descends over the C1 arch [13]. The relation between tonsillar descent, changes of the cervical alignment, and neurologic recovery rate was investigated.

To measure the intra- and interobserver reliability of radiographic parameter, a spine surgeon and a radiologist independently performed the measurements three times. There was at least a 2-week interval between each measurement. All measurements were made using a picture-archiving and communication system, DICOM (version 3.0; CompuMed, Inc., Tucson, AZ, USA). The Japanese Orthopaedic Association (JOA) scoring system was used to ascertain the neurological status [19]. Furthermore, the neurologic recovery rate at last follow-up was calculated using the following formula: (postoperative score – preoperative score) $\times$ 100/(full score [17 points] – preoperative score).

For statistical analysis, SPSS software (version 12.0, 2003; SPSS, Inc., Chicago, IL, USA) was used. The differences between the pre- and postoperative values were tested using Student's *t* test. The relationships between clinical and radiographic parameters and the difference of the cervical lordotic angles before and after surgery were assessed using Spearman rank correlation coefficients. The Mann–Whitney *U* test was used to analyze the differences of cervical sagittal alignment and JOA recovery rate between groups according to the extent of tonsillar descent. Intra- and interobserver reliability of the measurements was estimated using intraclass correlation coefficient (ICC), which provides a parameter-specific index of variability comparing error due to repeated trials to the true error expected due to other sources. The coefficient was calculated using the first and third trials for each examiner. According to Fleiss's guidelines [4], ICCs less than ± 0.40 indicate poor; ± 0.40 – 0.75 , fair or good; and ± 0.75 – 1.00 , excellent reliability. A *P* value <0.05 was considered statistically significant. When applicable, the mean values are presented with standard deviations.

Results

There were 10 men and 20 women. The mean age at surgery was 33.4 years (range 13–61 years). Mean duration of

Fig. 1 Intraoperative photographs showing structures exposed when performing foramen magnum decompression. All patients underwent suboccipital craniectomy, C1 laminectomy and expansile duroplasty

using the artificial dura mater **a** structures after duroplasty **b** photograph after expansile duroplasty using the artificial dura mater

follow-up was 6.5 ± 1.5 years (ranged from 4.0 to 9.5 years). The means of the JOA scores before surgery and at the final follow-up evaluation were 10.3 ± 3.1 points (range 4–15 points) and 13.4 ± 2.8 points (range 8–17 points), respectively, and the mean of the recovery rate was 46.2 ± 21.2 %. There were three surgery-related complications. Two patients had pseudomeningocele and one experienced a superficial wound infection, all of which were cured after conservative managements.

Changes of syrinx size and cervical sagittal alignment

On radiographic study, the assessment of intra- and inter-observer reliability showed excellent agreement between the three measurements for syrinx width and the cervical lordosis angle (ICC 0.88 and 0.82, respectively). The mean value of the syrinx width was 6.1 ± 2.7 and 2.4 ± 1.8 mm before and after FMD surgery, respectively. There was significant difference between the pre- and postoperative values ($P < 0.000$). The mean pre- and postoperative lordosis angles at C2–C7 were $-5.9^\circ \pm 1.0^\circ$ and $-10.4^\circ \pm 1.0^\circ$, respectively. The difference between the values is significant ($P = 0.001$, Fig. 2).

The shrinkage of the syrinx was confirmed in 90.0 % (27 patients) on postoperative MR images. No shrinkage of the syrinx occurred in the other 10.0 % (three patients), their clinical symptoms had not improved (two: clinically

worsening and one: no change of symptoms). There was no correlation between decrease in syrinx size after surgery and recovery rate by JOA scores from the neurological symptoms ($P > 0.05$; Spearman rank correlation test was used).

There was no significant correlation between baseline characteristics (age at surgery, gender, preoperative syrinx width, preoperative stage of tonsillar descent and preoperative cervical lordotic angle) and difference of cervical lordotic angles before and after surgery (Table 1). There was significant correlation between the difference ratio of syrinx width and the difference of the cervical lordotic angles before and after surgery ($P = 0.016$ by Spearman rank correlation test, Fig. 3). There were no significant differences of the cervical alignment, and neurologic recovery rate between groups according to the extent of cerebellar tonsillar descent ($P > 0.05$; Mann–Whitney *U* test was used). After FMD, syringomyelia and cervical alignment improved in 28 (93.3 %) and 25 (83.3 %) of 30 patients, respectively. No improvement of the cervical alignment occurred in the other 16.6 % (five patients), their clinical symptoms had not improved (three: clinically worsening and two: no change of symptoms). There was significant correlation between recovery rate by JOA scores and the difference of the cervical lordotic angles before and after surgery ($P = 0.022$; Spearman rank correlation test was used).

Fig. 2 A representative case of a 23-year-old female. Preoperative (a, b), postoperative (c, d) magnetic resonance imaging and lateral radiograph reveal that size of syrinx width decreased (from 8.6 to

3.1 mm) and the cervical lordotic angle restored (1.8° kyphosis to −11.3° lordosis) after foramen magnum decompression

Discussion

The association between Chiari malformation (exclusive of myelodysplasia) and scoliosis has been recognized [1, 3, 6, 14, 16]. Previous researchers suggested that scoliosis can improve after decompression of syringomyelia [11, 15]. It means that syringomyelia may be an aggravating factor of

spinal alignment. However, there are few researches that demonstrate relationship between the extent of syringomyelia and sagittal alignment of the cervical spine. This study has documented differences in the cervical sagittal alignment in Chiari I malformation with syringomyelia after FMD. In the present study, after FMD, syringomyelia and cervical alignment improved in 28 (93.3 %) and

Table 1 Relationships between baseline characteristics and changes of the cervical alignment

	Difference of cervical angle (<i>P</i> value)
Age at surgery	0.506
Gender	0.068
Preoperative syrinx width	0.192
Preoperative stage of tonsillar descent	0.097
Preoperative cervical lordotic angle	0.051
Difference ratio of syrinx width	0.016
Recovery rate	0.022

Difference ratio = (preoperative width – postoperative width)/preoperative width

Recovery rate by Japanese Orthopaedic Association scoring system = (postoperative score – preoperative score) × 100/(17 points – preoperative score). Relationships were assessed using Spearman rank correlation coefficients. The Mann–Whitney *U* test was used to analyze the differences between groups according to the extent of cerebellar tonsillar descent

Fig. 3 There was significant correlation between the differences ratio of syrinx width and the cervical lordotic angles before and after surgery. Spearman rank correlation test was used

25 (83.3 %) of 30 patients, respectively. Moreover, there was significant correlation between the differences ratio of syrinx width and the cervical lordotic angles before and after surgery. Thus, we suggest that FMD could help to restore cervical alignment.

The association between Chiari I malformation and scoliosis has been not completely understood. Huebert and MacKinnon theorized that the syringomyelia compromises cells in the cord that are responsible for muscle balance of the trunk, leading to the scoliosis [8]. They

insisted that early decompression appears to have more favorable results on scoliosis. In the current study, cervical spinal alignment significantly improved after decompression of the syrinx in adult patients with Chiari I malformation. Mean value of the cervical lordotic angle in the present study was less lordotic than that in a previous research with asymptomatic subjects [7] (-5.9° vs. -34°). We suggest that space occupying syrinx in the spinal cord might make the cervical alignment kyphotic as a physiologic response. Accordingly, decompression of the spinal canal by FMD may restore the cervical sagittal alignment.

In the current study, the mean of the recovery rate was 46.2 ± 21.2 % according to the JOA scores before surgery and at the final follow-up, although there was no correlation between the difference of syrinx size and recovery rate. These findings are consistent with previous reports [2, 13, 21]. Thus, we suggest that foramen magnum decompression is considered to be an effective surgical technique for Chiari I malformation occurring with syringomyelia. Furthermore, there was significant correlation between recovery rate by JOA scores and the difference of the cervical lordotic angles before and after surgery. Considering the relationship, we suggest that the postoperative cervical alignment might be a predictive factor for neurological outcome. In the context, surgeons should observe the cervical alignment carefully. A postoperative plain lateral radiograph of the cervical spine could be helpful in the clinical evaluation of a patient with Chiari I malformation associated with syringomyelia. Previous researcher reported thoracic hyperkyphosis as an indicator of syringomyelia in idiopathic scoliosis [18]. They suggested that musculoskeletal abnormalities constitute the initial manifestation of intraspinal lesions in a large percentage of cases.

A limitation of this study involves the age of enrolled patients. The mean age at surgery was 33.4 years (range 13–61 years). Thus, further investigation for pediatric patients should be needed. Moreover, there were a small number of patients in the study. Larger number of patients would be necessary for more detailed determination of the change of cervical alignment after FMD.

Conclusion

There is significant relation between the decrease in size of syringomyelia and cervical alignment. The present results demonstrate that the decrease of syrinx size by FMD may restore the cervical lordosis. There was significant correlation between recovery rate by JOA scores and the difference of the cervical lordotic angles before and after surgery.

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Conflict of interest None of the authors has any conflict of interest in the subject under discussion in this paper.

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