CLINICAL ARTICLE

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Observation between Clinical Outcomes and the Size of the Syrinx with Magnetic Resonance Image

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Objective : This study was conducted to examine the correlation between clinical outcomes and the size of the syrinx in post-operative magnetic resonance imaging(MRI) and symptom duration.

Methods : The authors investigated twelve patients who underwent various operations for syringomyelia from January 1995 to December 2003. The authors retrospectively analyzed medical records, pre- and post-operative MRI findings, features and durations of symptoms, and the method of surgical treatments. The clinical outcomes were assessed on Prolo scale at 6 months of post-operative period.

Results : Neurologic symptoms did not promptly disappear after the shrinkage of syrinx, but post-operative MRI demonstrated most patients showed reductions in the size of the syrinx. There is no statistical relationship between clinical improvements and decrements of the syrinx size. However, patients who underwent surgical treatment within 2 years from the symptom onset had more favorable outcome than those who had operations after 2 years from the onset of symptoms.

Conclusion : Change in the size of the syrinx in post-operative MRI is not directly proportional to favorable clinical outcomes. However, symptom duration before surgical treatment has considerable impact on the clinical outcomes.

KEY WORDS : Syringomyelia · Syringosubarachnoid shunt · Posterior fossa decompression · Chiari malformation.

Introduction

The increasing awareness of the importance of early detection of malformation has led to increased use of magnetic resonance imaging(MRI). Thus, with the widespread use of MRI in recent years, syringomyelia has been diagnosed in an increasing number of patients. Many authors of recent reports have suggested that early diagnosis and proper application of surgical techniques, such as posterior fossa decompression and syringosubarachnoid shunt(SSA) have made it possible to reduce the size of the syrinx[12]. However, improvements in syringomyelia-related symptoms do not always correspond to reductions in the size of the syrinx, and this may pose treatment difficulties[8,11,30].

The purpose of our study is to estimate the correlation between the severity and duration and range of clinical symptoms attributable to syringomyelia with MRI appearance.

Materials and Methods

Between January 1st 1995 and December 31st 2003, we examined 12 operated patients with MRI-verified syringomyelia. Among 12 patients, the age range was 11 to 63 years old (median 32.5 years old). Post-operative follow up period was 10 to 62 months (median 35 months). All patients performed pre- and post-operative MRI to compare the size of the syrinx.

The authors retrospectively evaluated operative methods, post-operative symptom improvements, features and durations of symptoms and change in the size of the syrinx based on the medical records and MRI findings. Anteroposterior diameter of the syrinx was measured pre- and post-operatively at the maximum syrinx distention on T2 weighted MR image in the mid-sagittal plane. 6 months after operation, Prolo's scale was used to estimate the clinical outcomes. The relationship

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Table 1. Summary of clinical features of patients with syringomyelia

Case No.	Age(years) sex	Chief complaint	Cause	Location of syrinx	Surgical procedures
1	57/M	Sensory impairment in rt arm	Infection	C7-T12	SSA shunt
2	42/F	Truncal pain, motor weakness in lt hand, Muscle atrophy	Trauma	C2-C7	SSA shunt, PHL C5,C6
3	16/M	Truncal pain, gait disturbance	Truma	C2-C7	SSA shunt, TL C4,C5
4	32/M	Slight lt hemiparesis	Trauma	C3-T1	SSA shunt, STL C7
5	11/F	Gait disturbance, Headache	Chiari I malformation	C1-C3	PFD, Duraplasty
6	42/F	Rt hemiparesis, Headache	Trauma	T11-L1	SSA shunt
7	63/M	Rt hemiparesis, truncal ataxia	Infection	T10-T12	SSA shunt, TL T11
8	30/F	Motor weakness, clumsiness in both hands	Trauma	C4-C7	LP shunt
9	38/F	Quadriparesis, muscle atrophy	Trauma	C1-C4	SSA shunt, PL C1-C3
10	20/M	Pain in both hands and feet	Chiari I malformation	C2-C4	PFD, TL C4 Duraplasty
11	34/M	Sensory impairment in lt arm	Trauma	C3-C6	SSA shunt
12	60/M	Sensory impairment in lt trunk and lt upper extremity	Trauma	C3-C7	AIF C4-C6 with amslu cage, LP shunt

Abbreviations : M : male, F : female, Rt : right, Lt : left, C : cervical spine, T : Thoracic spine, SSA shunt : syringosubarachnoid shunt, LP shunt : lumboperitoneal shunt, TL : total laminectomy, STL : subtotal laminectomy, PHL : partial hemilaminectomy, PL : partial laminectomy, AIF : anterior interbody fusion, PFD : posterior fossa decompression

Table 2. Number of patients with postoperative size reduction of the syrinx according to the operative method

Type of surgery	No. of cases No. of reduced size of the syrinx
Syringosubarachnoid shunt	6/8
Posterior fossa decompression	1/2
Lumboperitoneal shunt	1/2

between the reduction in the size of the syrinx and symptom durations and symptom improvements were evaluated by using McNemar test, and the reliability index was 95%($p<0.05$).

The authors tried to select operative methods according to the patient's clinical characteristics and the cause of syringomyelia. Syringosubarachnoid(SSA) shunt, lumboperitoneal (LP) shunt, posterior fossa decompression were used as operative options. Sometimes, posterior fossa decompression and duraplasty with artificial dura were undertaken at the same time. In case of Chiari I malformation, cervical laminectomy was undergone with posterior fossa decompression and duraplasty. LP shunt was used when the syrinx size was small and the fourth ventricle had no connection with the syrinx. In one post-traumatic case with syrinx cavity in the C4-C6 area, anterior interbody fusion (AIF) with cages and LP shunt were applied.

Results

The initial symptoms were motor weakness in 6 cases, segmental sensory impairment in 3 cases, pain on the trunk and limbs in 3 cases, and walking difficulty in 3 cases. Neurologic symptoms were bilateral in 4 cases, unilateral in 10 cases (Table 1). The causes of syringomyelia were trauma in 8 cases, infection in 2 cases, and Chiari malformation in 2 cases (Table 1). 8 patients underwent SSA shunt, 2 patients LP shunt and 2 patients posterior fossa decompression with duraplasty (Table 1). The interval between the onset of the initial symptoms to the surgery ranged from 0.6 to 8 years (mean 1.7 years). The preoperative anteroposterior diameter of the syrinx ranged from 3.5 to 11.2mm (mean 8.3mm) and the post-operative diameter of the syrinx ranged from 2.0 to 11.7mm (mean 5.1mm). There was no significant relationship between the operative methods and the post-operative syrinx shrinkage (Table 2). Post-operative desirable clinical outcomes included 3 excellent grade (Prolo's scale 10~9) cases and 5 good grade (Prolo's scale 8~7) cases. Fair grade (Prolo's scale 6~5) was 4 cases, and poor grade (Prolo's scale below 5) did not exist. A relationship between the reduction of size of the syrinx and symptom improvements was analyzed. 8 in 12 patients showed decreased size of the syrinx post-operatively, but 5 among those 8 patients demonstrated symptom improvements (Fig. 1, 2). Moreover, 3 out of 4 patients who had not had reductions in the size of the syrinx had symptom improvements (Fig. 3). Although this study has limitations in terms of statistics because of the small number of cases, the authors drew a conclusion that the change of the syrinx size has nothing to do with clinical symptom improvements, statistically (Table 3, $p>0.05$). A correlation between symptom improvement and time to surgical treatment was

analyzed. 9 patients had surgical corrections within 2 years after the symptom onset, while the other 3 patients underwent operation more than 2 years after the symptom onset. Among the former 9 patients, 7 had symptom improvements, while the latter 3 patients had 1 case of symptom improvement. A significant relationship between symptom improvement and time to surgical correction was detected (Table 4, $p<0.05$). Patients who had less than 2 years to operation from the symptom onset had more favorable outcomes, whereas patients who had more than 2 years to operation from the symptom onset had less favorable outcomes. Post-operative CSF leakage appeared in 2 cases but were treated successfully with conservative management. Neither central nervous system (CNS) infection nor catheter obstruction happened.

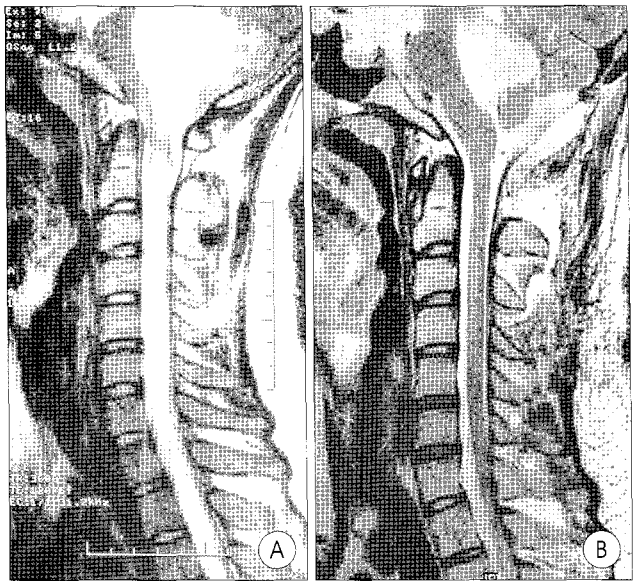

Fig. 1. A 20-year-old man with Chiari I malformation had symptom improvement after the operation and the size of the syrinx was reduced. A : T2 weighted magnetic resonance image shows ventriculomegaly and syrinx that extends caudally to the level of T1 vertebral body. B : Marked deflation of previous syrinx was noted in the postoperative T2 weighted magnetic resonance image after 6 months of surgery.

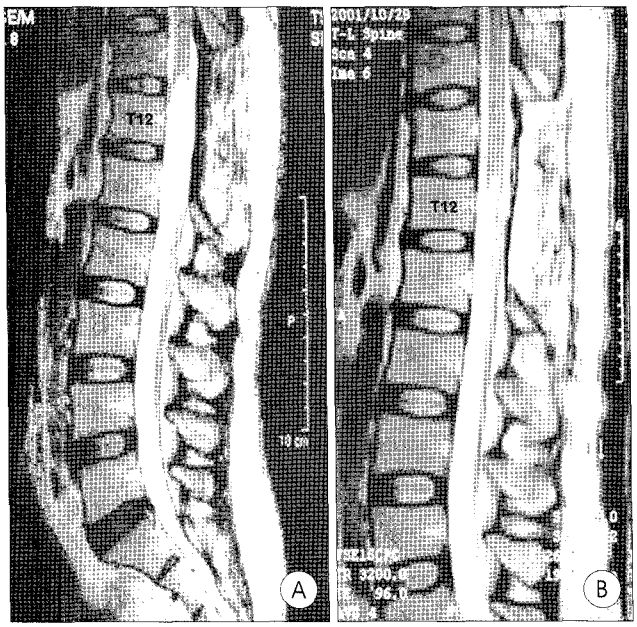

Fig. 2. A 42-year-old woman presenting with sensory impairment had symptom improvement after the operation with subsequent reduced size of the syrinx. A : Syrinx cavity is observed at the previously operated site due to arachnoid cyst. B : Postoperative T2 weighted magnetic resonance image reveals marked reduction in the size of the syrinx cavity.

Discussion

The pathophysiology of syringomyelia is still controversial but two different mechanisms have been suggested.

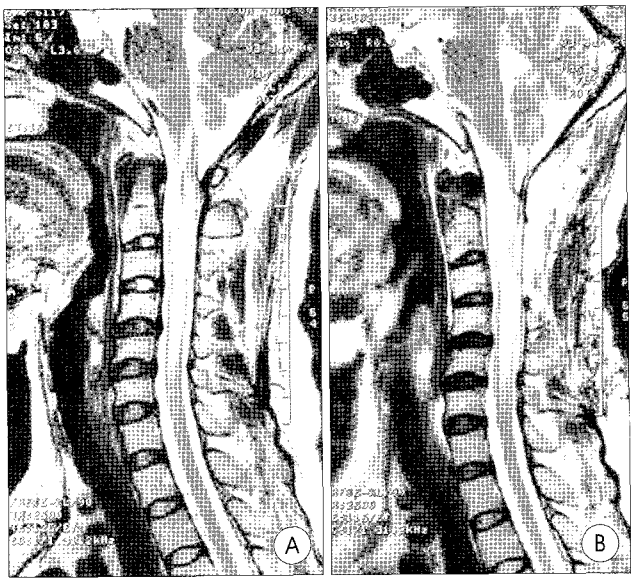

Fig. 3. A 38-year-old woman complaining of quadriparesis(motor grade IV+) had symptom improvement after the operation despite postoperative enlargement of the size of the syrinx. A : Syrinx extends from C1 to C4 in the preoperative T2 weighted magnetic resonance image with elongated shape. B : T2 weighted magnetic resonance image obtained 6 months after the surgery demonstrating increased syrinx size with severe obliteration of the subarachnoid space.

Table 3. Correlation between reduction of the size of the syrinx and desirable postoperative outcomes

Reduction of the size of the syrinx	Desirable outcome	
	No	Yes
No	1/12	3/12
Yes	3/12	5/12

L>0.05 by McNemar test, Desirable outcome includes excellent and good outcome according to Prolo's scale

Table 4. Correlation between symptom improvement and time to surgical treatment after the symptom onset

Symptom improvement	Time to surgical treatment	
	<2 years	>2 years
No	2/12	2/12
Yes	7/12	1/12

P<0.05 by McNemar test

Gardner[10] proposed impaired outflow of spinal fluid to the CSF space at the level of fourth ventricle and the role of systolic pulsation that transmitted to the syrinx cavity as a generation factor for syringomyelia(water hammer effect). Williams[31] proposed that increased intracranial venous pressure, as during coughing or other valsalva-like maneuvers, cause cranio-spinal dissociation of pressure. In the end, syringomyelia develops with partial obstruction at the level of foramen magnum(sloshing mechanism). Moreover, under these circumstances, hindbrain anomalies encourage the movement of CSF into the spinal cord, which is another driving force to generating syringomyelia.

Different surgical treatments have been recommended for spinal cord cavities in response to varying pathogenic and etiologic conditions. In cases of cavities associated with foramen magnum lesion, especially Chiari malformations, various surgical techniques have been advocated; drainage procedures and craniocervical decompression are, however, most frequently and widely adopted[2]. Ventricular shunting has been recommended as the first procedure for syrinx occurring in association with hydrocephalus[20,32,33], and syrinx shunting procedures have been widely accepted for the treatment of any kind of cavities except those that are tumor-related. In spinal cord cavitation associated with foramen magnum lesions, obstruction of the subarachnoid space at the foramen magnum and/or the CSF outlet from the fourth ventricle is responsible for the cavitation[16]. Therefore, craniocervical decompression, in which various intradural procedures are performed to resolve the CSF blockage and to restore normal CSF dynamics seems to be the most rational surgical procedure for the treatment of spinal cord cavities associated with foramen magnum lesions, especially Chiari malformations. In our two cases of Chiari malformation, we also adopted this surgical procedure. Idiopathic cavities have been treated by syrinx shunting such as SSA shunt[27,29] and syringoperitoneal shunt[5,9,19,22,26], but rarely by craniocervical decompression[15,23]. When considering craniocervical decompression, caution should be given not to excessively open up the posterior fossa. A wide opening of the foramen magnum will be enough. Holly and Batzdorf[14] have reported cases with symptomatic cerebellar ptosis following craniocervical decompression for Chiari I malformations.

The pre-operative duration of the disease has an effect on the post-operative results because irreversible changes in the spinal cord, such as neuronal loss, glial scar formation and myelomalacia, may have occurred before surgical correction[3,6,21]. Bogdanov and Mendelevich[6] reported that spontaneous drainage forms between syrinxes and the subarachnoid space because of fissuring through the thinned cavity walls and this spontaneous shunt in chronic syringomyelia works as a compensatory mechanism to lessen the effect of surgical treatment.

MRI has been the most effective diagnostic tool in patients with syringomyelia because of its non-invasiveness and prominent ability to delineate soft tissue structures. Imaging findings were well documented through various literatures[18,24]. With development of advanced MRI techniques, more detailed analysis of CSF flow became realized, including pulsatile movement of the CSF in the spinal canal[25]. Spatial modulation of magnetization(SPAMM) technique was developed for the evaluation of cardiac wall motion, moving blood or cerebrospinal fluid[4,7,17]. Terae et al.[28] discussed pulsatile movement of the hindbrain and the motion of the intra-syrinx fluid in syringomyelia patients associated with Chiari malformation. They found downward displacement of the band stripe on the cine-MRI of the spinal canal with presaturation bolus tracking. They postulated that increased pulsatile movements of the spinal cord, together with the one-way valve mechanism at the level of CSF outflow obstruction, act as a "vacuum-pump" to extend the syrinx. Recently, many techniques including phase contrast velocity imaging and bolus tracking method were used in the evaluation of syringomyelia[1,13]. SPAMM, in contrast to other techniques, can provide more detailed and apparent visual data of CSF motion and direction of the flow within the spinal canal. In our study, these advanced MR techniques were not used. If they are adopted in future studies, it will be helpful in clinical correlation.

Conclusion

The reduction of the syrinx size and clinical symptom improvement have no statistical significance. Most of surgically treated patients have shown the reduction of the syrinx size in the follow up MRI, but all of them haven't showed symptom improvements. However, time to surgical treatment after the symptom onset and symptom improvement have statistical significance. Patients who are surgically treated within 2 years after the symptom onset have more favorable outcome. Because this study has a small number of cases, clinical analysis with a larger number of cases and a long-term follow up will be needed.

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Commentary

This paper addressed the issue whether we could expect a good outcome if we can reduce the size of the syrinx. Several papers have already addressed this issue, and the consensus was that we could not expect a good outcome always. The previous literatures were cited in the discussion of this paper.

I do not need to reiterate this issue, so I like to mention several weak points of this paper, which were not discussed in the paper. I also hope to have a better paper as the authors.

Firstly, their study population was too diverse to draw a convincing data. Even though the authors admitted that their study has a small number of cases and a shorter follow up, they did not mention diversity of their study materials with various surgery types would be a much more serious problem. They included syringomyelia from the trauma, infection or Chiari malformation. The majority were from the trauma, not from Chiari malformation. I do not expect similar postoperative outcome from these two different causes.

Secondly, they used Prolo scale 6 months after the surgery. Prolo scale is about pain and working, so is popular for evaluating lumbar disc surgery. The study population presented with pain in only 3 patients. The majority presented with either motor weakness or sensory impairment. Myelopathy scales such as Japanese orthopedics scale or Nurick grade would be a better measurement tool.

Thirdly, they did not mention when they took the MR images postoperatively. They compared the syrinx size and Prolo scale at 6 months postoperatively. I do not understand why they provided the timing of Prolo scale only without mentioning the timing of MR image study.

Fourthly, the importance of CSF flow study could not be too emphasized as they mentioned in the discussion. However, the CSF flow abnormality has not been studied well in syringomyelia without Chiari malformation. Lee et al.[1] published an interesting paper about this issue in Spinal Cord, which was not included in their reference.

Considering the limitation posed by the rarity of syringomyelia and diversity of its etiology, this paper was written

reasonably well. I read this paper with a greatest interest, not because their result gave me a new one, but because they provided a balanced view about the syringomyelia.

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