Intradural Pathology Causing Cerebrospinal Fluid Obstruction in Syringomyelia and Effectiveness of Foramen Magnum and Foramen of Magendie Dredging Treatment

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■ OBJECTIVE: **This article discusses the procedure of foramina magnum and Magendie dredging, summarizing the pathologic changes in the intradural region of the craniocervical junction in patients with syringomyelia and the pathophysiologic mechanism of cerebrospinal fluid (CSF) circulation obstruction.**

■ METHODS: **Clinical data from 50 adult patients with syringomyelia treated at Xuanwu Hospital from July 2018 to January 2019 were collected and retrospectively analyzed. All operations were performed with foramina magnum and Magendie dredging, and all intradural factors that may have induced the obstruction of CSF circulation were recorded.**

■ RESULTS: **Intradural pathology was found in all patients. The pathologic changes that may have caused obstruction of the CSF circulation include tonsil occupying the foramen magnum and overlying foramen of Magendie in 88% (44/50), intertonsillar arachnoid adhesions in 36% (18/50), tonsil to medulla arachnoid adhesions in 18% (9/50), medialized tonsils in 70% (35/50), vermian branch of posterior inferior cerebellar artery in 22% (11/50), arachnoid veil in 16% (8/50), cisterna magna cyst in 4% (2/50), and tonsil to dura mater arachnoid adhesions in 8% (4/50). Mean duration of follow-up was 13.3 months. The long-term effective rate was 96.0%. Postoperative magnetic resonance imaging revealed that the size of the syringomyelia was reduced or completely resolved in 88% of patients. The mean preoperative Japanese Orthopaedic Association score was 12.9 ± 3.1, which improved to 14.7 ± 3.2 ($P < 0.05$) at last clinical follow-up.**

■ CONCLUSIONS: **Intradural pathology that causes CSF circulation obstruction exists in many forms. Relieving the obstruction of the foramen magnum and foramen of Magendie is key to surgical treatment.**

INTRODUCTION

The natural history of syringomyelia (SM) is unpredictable.[1] The etiology and pathogenesis of SM are diverse and not yet fully understood. SM is often associated with Chiari malformation (CM) and various other craniocervical junction malformations, which may occur with trauma, infection, and tumors.[2-4] Regarding the mechanism of SM, there are 3 generally recognized theories: the water-hammer theory proposed by Gardner et al.,[5] the cranial-spinal pressure dissociation theory proposed by Williams and Fahy,[6] and the piston-like effect theory proposed by Oldfield et al.[7] The work by Stoodley's group[8] is also helpful to understand the pathophysiology of SM. However, none of these theories can explain the mechanism of SM formation alone.[9]

Few systematic clinical data exist on the incidence of intradural pathology in patients with SM. The pathophysiologic significance of these changes in relation to the pathogenesis and outcome of SM has not been reported in great detail. There is no uniform surgical protocol for treatment of SM. Surgical methods include posterior fossa decompression (PFD) and posterior fossa

Key words
- Cerebrospinal fluid
- Chiari malformation type I
- Craniovertebral junction
- Pathology
- Syringomyelia

Abbreviations and Acronyms
CM: Chiari malformation
CSF: Cerebrospinal fluid
FMMD: Foramina magnum and Magendie dredging
MRI: Magnetic resonance imaging
PFD: Posterior fossa decompression
PFDD: Posterior fossa decompression and duraplasty
PICA: Posterior inferior cerebellar artery
SM: Syringomyelia

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decompression and duraplasty (PFDD).[10,11] Although different surgical methods have certain effects, they are only partially beneficial in patients with SM.

In this study, we collected imaging, intraoperative and follow-up data from 50 adult patients with SM who were admitted to our department in a 6-month period to analyze the pathophysiologic significance of intradural pathology in relation to the pathogenesis and outcome of SM. Foramina magnum and Magendie dredging (FMMD) was used to relieve the factors causing obstruction, providing a theoretical basis for further improvements in surgical procedures to improve efficacy. In addition, we used FMMD to treat patients with syrinx who had persistent or worsening of symptoms and achieved good results.[12]

MATERIALS AND METHODS

General Data

This study was approved by the Institutional Research Ethics Committee of Xuanwu Hospital. The cases consisted of 59 consecutive adult patients in whom SM was diagnosed by magnetic resonance imaging (MRI) at Xuanwu Hospital from July 2018 to January 2019. However, 9 of 59 patients did not meet the inclusion criteria owing to a lack of long-term MRI or loss to follow-up. Thus, there were 22 men and 28 women 18–69 years of age (mean age of 43.1 years) with a disease course of 1–11 years (mean disease course of 4.1 years); the patients were followed for 12–15 months (mean follow-up of 13.3 months).

Inclusion criteria included the following: 1) patients with SM and CM type I, with head and neck MRI showing cerebellar tonsils >5 mm below the foramen magnum; 2) patients with SM without CM, but with imaging suggesting abnormalities in the subarachnoid space of the craniocervical junction; 3) patients with SM without CM, but SM up to the medulla oblongata was detected with a channel between the fourth ventricle and the syrinx. Exclusion criteria included the following: 1) patients with complex craniocervical junction deformities, such as atlantoaxial dislocation and basilar invagination, in which the anterior border of the foramen magnum or the odontoid process caused compression of the brainstem; 2) patients with asymptomatic syrinx/cavities found incidentally; 3) patients with localized cavities caused by intraspinal canal tumors, trauma, or central nervous system infection; 4) patients with a history of craniocervical junction surgery.

Preoperative Assessment

Clinical Evaluation. Clinical manifestations including the following: 1) SM symptoms—16 patients with dissociative sensory disorder, 12 with weakness, and 7 with muscle atrophy; 2) brainstem and cranial nerve dysfunction symptoms—23 patients with neck and shoulder pain and 7 with facial numbness; 3) cerebellar symptoms—11 patients with gait instability; 4) increased intracranial pressure symptoms—15 patients with headaches and 1 patient with hydrocephalus. Neurological functions were evaluated using the 17-point scoring system of the modified Japanese Orthopaedic Association scale.[13]

Preoperative Imaging Examination. All patients underwent MRI (3.0 TSOMATOM Definition superconducting MRI scanner; Siemens Healthineers, Erlangen, Germany) to reveal the presence and degree of CM and whether there was an abnormality in the subarachnoid space at the craniocervical junction. In the sagittal view, the inferior edge of the cerebellar tonsil was observed to be 5 mm beyond the McRae line in 44 patients (range, 6–17 mm; average, 10.5 mm); in 32, 10, and 2 patients, the tonsil reached the level of the C1 posterior arch, C2 vertebral arch, and C3 vertebral arch, respectively. There were 4 patients with abnormal signals or membranous structures in the subarachnoid space at the craniocervical junction. Two patients without CM or abnormal signals in the craniocervical junction demonstrated SM up to the medulla oblongata with a detectable channel between the fourth ventricle and the syrinx. MRI was also performed to understand the SM segments and morphology. In this group, 7 patients had cervical SM, 38 patients had cervicothoracic SM, 5 patients had holocord SM, and 7 patients had SM that reached the medulla oblongata (syringobulbia). Syrinx size was measured via MRI, and the maximum diameter in axial images was recorded in millimeters with accuracy to 0.1 mm.

Surgical Procedures

All operations were performed via PFD with intradural exploration using FMMD to relieve the factors inducing obstruction, and all intradural pathology and factors potentially causing cerebrospinal fluid (CSF) circulation obstruction were recorded via the operating microscope. The specific surgical procedures are described next.

Exposure. After administration of combined general anesthesia, patients were placed in the prone position, and the head was fixed in a head frame. Routine disinfection and draping procedures were conducted. A straight incision was made approximately 4 cm from the posterior midline of the external occipital protuberance; then cuts were made along the midline to the occipital bone layer by layer. The posterior edge of the foramen magnum and part of the occipital squama were exposed, and the posterior edge of the foramen magnum was opened. Part of the occipital squama was removed for the migration of the cerebellar vermis and cerebellar tonsil. The upper edge of the posterior arch of the atlas was removed (if the cerebellar tonsillar herniation exceeded the inferior edge of the atlas, the entirety of the posterior arch of the atlas was removed), and the posterior atlanto-occipital membrane was peeled away.

Microscopic Intradural Exploration. The dura mater and arachnoid membrane were longitudinally incised and suspended on both sides to fully expose the visual field. Intradural exploration was performed under the microscope. First, the arachnoid membrane in the cisterna magna was opened using microscissors and lysed. Second, sharp separation of the adhesions and thickened arachnoid was performed between the bilateral cerebellar tonsils, between the cerebellar tonsils and the dura mater, and between the tonsils and brainstem, thus freeing the bilateral cerebellar tonsils. Third, after the brainstem and posterior inferior cerebellar artery (PICA) were well protected with a cotton pad, for herniated and medialized cerebellar tonsil hypertrophy and cerebellar tonsils with a slight degree of herniation, low-power bipolar subpial electric coagulation of the cerebellar tonsils was performed such

that the tonsils ascended upward and outward; for cerebellar tonsils with obvious herniation, subpial resection was performed, and electrocoagulation was used to seal the pial opening after resection to confirm the absence of active bleeding. The goals of resection were to ensure that there was no significant obstruction of CSF circulation in the foramen magnum under natural conditions and to ensure that there was no significant occlusion of the foramen of Magendie by the herniated or medialized cerebellar tonsils under natural conditions. The integrity of the pia mater was maintained to avoid potential adhesion, scarring, and recurrence. Fourth, exploration was employed to determine if there was a PICA or a branch located around the foramen of Magendie, and the vessels were released; the obstruction of the foramen of Magendie by the vessels was relieved, thus facilitating CSF flow. Finally, the foramen of Magendie was explored, and the membranous structure was cut. After satisfactory release, CSF was observed to flow out of the foramen of Magendie.

Suture. The dura mater was sutured in situ. If the tension was too high such that the dura mater could not be sutured, a small amount of fascia or muscle was used for the repair. The muscle and skin tissues were tightly sutured layer by layer without placing a drainage tube.

Postoperative Assessment

Evaluation of MRI. Patients underwent MRI of the posterior fossa and the cervical and thoracic spine 3 days, 3 months, and 1 year after surgery. If the size of the SM was unchanged or the syrinx decreased in size <20%, we deemed it stable; if the size of the SM was smaller (syrinx decreased in size ≥20%) or the SM had resolved, we considered it improved; and if the size of the SM was increased, we considered it worsened.

Evaluation of Clinical Efficacy. Patients underwent clinical examination 3 days, 3 months, and 1 year after surgery. The effectiveness of surgery in regard to the main symptoms was judged before and after surgery and during follow-up according to the Tator criteria.[14] Patients with symptoms that were significantly improved and signs that had basically resolved were assessed as improved. Patients with symptoms that remained basically the same and the presence of no new neurological impairment were assessed as stable. Patients with symptoms that were significantly worsened or the presence of new pathologic signs were assessed as worse.

Statistical Analysis

All study data were statistically analyzed using PASW Statistics 18 (SPSS Hong Kong Ltd., Quarry Bay, Hong Kong), and enumeration data were expressed as percentages or constituent ratios and statistically analyzed using the Pearson χ^2 test and Fisher exact test. Differences were considered statistically significant at $P < 0.05$.

RESULTS

Clinical Outcome

The short-term curative effect was evaluated by observing improvements in symptoms and signs 3 days after surgery, and the long-term curative effect was evaluated 1 year after surgery. The follow-up period was 12–15 months (mean 13.3 months).

Short-Term Curative Effect. The short-term curative effect for 50 patients after surgery was as follows: 42 (84%) patients were improved, 6 (12%) were stable, and 2 (4%) had deteriorated.

Long-Term Efficacy. The long-term curative effect for 50 patients after surgery was as follows: 44 patients (88%) were improved, 4 (8%) were stable, and 2 (4%) had deteriorated. The long-term improvements in symptoms of patients are shown in Table 1. The mean preoperative Japanese Orthopaedic Association score was 12.9 ± 3.1, which improved to 14.7 ± 3.2 ($P < 0.05$) at the last clinical follow-up.

Postoperative Complications and Treatment. One patient developed a postoperative fever, and infection was diagnosed by lumbar puncture. The infection resolved after 1 week of antibiotic treatment. The remaining patients did not present with any significant postoperative complications.

Radiologic Follow-Up: SM Changes

Patients underwent imaging 1 year after surgery. The syrinx diameter was significantly decreased (before surgery 7.1 ± 3.7 mm, after surgery 2.9 ± 2.8 mm; $P < 0.05$) in 44 of 50 patients (88%), and the cavity size was unchanged in 6 patients (12.0%).

Representative Cases

Case 15: Obstruction of the Foramen of Magendie (Figure 1). A 50-year-old man presented with acute episodes of dysphagia and dyspnea and reported right shoulder pain for 4 months. Preoperative MRI showed SM with extension into the medulla. Intradural surgery demonstrated a nearly impermeable arachnoid veil overlying the foramen of Magendie, which was removed. Postoperative MRI showed a marked reduction in the size of the SM. After the surgery, the dysphagia and dyspnea were almost resolved, but the pain and superficial sensory abnormalities persisted.

Case 21: Obstruction of the Foramen Magnum (Figure 2). An 18-year-old man presented with a 5-year history of occipital pain and dizziness. He also reported partial numbness in his limbs and right-hand inflexibility. Preoperative MRI showed cervicothoracic SM and suspicious abnormal signals in the cisterna magna. Microsurgical dissection and resection of the adhesion were performed. Postoperative MRI showed a marked reduction in the size of the SM. Postoperative symptoms were relieved except for hypoesthesia and right-hand inflexibility.

Case 29: Obstruction of the Foramen Magnum and Foramen of Magendie (Figure 3). A 29-year-old woman presented with a 10-year history of right-sided headache. A ventriculoperitoneal shunt had been placed 5 years previously and was removed 3 years ago because of fever. Two weeks ago, the patient suddenly lost consciousness for a few minutes and experienced headache, nausea, and vomiting. Preoperative MRI showed cervicothoracic SM without CM type I and a cyst in the cisterna magna. Microsurgical separation and resection of the cystic wall were

Table 1. Postoperative Long-Term Improvements in Different Symptoms

Symptom or Sign	Postoperative Long-Term Efficacy of Different Symptoms	
	Patients (*n*)	Improvement Rate
Dissociated sensory disturbance	16	37.5% (6/16)
Weakness	12	41.7% (5/12)
Atrophy	7	28.6% (2/7)
Headache	15	80% (12/15)
Neck and shoulder pain	23	87% (20/23)
Facial numbness	7	71.4% (5/7)
Impaired ambulation	11	81.8% (9/11)

performed. Postoperative MRI showed a significant reduction in the size of the SM. Postoperative symptoms were partially relieved.

Case 32: Tonsillar Obstruction of the Foramen Magnum and Foramen of Magendie (Figure 4). A 52-year-old woman presented with a 2-year history of finger numbness and reported posterior neck stiffness for the past month. Preoperative MRI demonstrated cervicothoracic SM with CM type I. The vermian branch of the PICA was lysed, and the arachnoid veil was removed. Postoperative MRI showed a significant reduction in the size of the SM. Symptoms were relieved after surgery.

Surgical and Intradural Findings

The pathologic entities that appeared to block CSF flow through the foramen of Magendie and the foramen magnum in all patients were noted and recorded (Table 2). The pathologic entities that prevented CSF from passing through the foramen magnum were cisterna magna cysts (Figure 3D) in 4% (2 of 50) of patients and tonsil to dura mater arachnoid adhesions (Figure 3F) in 8% (4 of 50) of patients. Obstruction of the foramen magnum and foramen of Magendie owing to the tonsils occupying the foramen magnum and the overlying foramen of Magendie (Figure 4D) was the most common finding in 88% (44 of 50) patients. The other pathologic entities that prevented CSF from passing through the foramen of Magendie were intertonsillar arachnoid adhesions (Figure 5A) in 36% (18 of 50) of patients; tonsil to medulla arachnoid adhesions (Figure 5B) in 18% (9 of 50) of patients; medialized tonsils (Figure 4D), which refers to significant occlusion of the foramen of Magendie by the herniated or medialized cerebellar tonsils under natural conditions, in 70% (35 of 50) of patients; obstruction of the foramen of Magendie by the vermian branch of the PICA (Figure 4F) in 22% (11 of 50) of patients; and blockage of the foramen of Magendie by the arachnoid membrane or scar tissue (Figure 4F) in 16% (8 of 50) of patients. Cerebellar tonsillar reduction was performed, arachnoid adhesions were lysed (Figure 5C), and the foramen of Magendie (opening of the fourth ventricle) was explored.

DISCUSSION

SM is a chronic progressive degenerative disease of the spinal cord characterized by somatosensory and motor dysfunction and autonomic nerve dysfunction. Previous studies have shown that both epidural and intradural factors are involved in the development of SM.[15-19] Epidural factors, including embryonic mesodermal somite occipital dysplasia, encroach on the posterior fossa, resulting in cerebellum and brainstem herniation below the level of the foramen magnum. This causes obstruction to the flow of CSF in the regions of the foramen magnum and foramen of Magendie, resulting in the development of SM.[20] However, in some patients with SM, the SM is not relieved after these epidural factors have been removed. Additionally, some patients have a normal posterior fossa volume, including patients without CM, and SM can be produced by intradural factors that cause obstruction to the flow of CSF.[21] All of these findings indicate that intradural factors play a crucial role in the generation of SM.

Surgery is currently the most commonly used and most effective treatment. However, controversies remain in terms of the surgical methods and procedures. The obstruction to the flow of CSF in the foramen of Magendie and foramen magnum is an important mechanism of SM development and progression. These subdural pathologic changes, discussed above, may be the pathogenic factors contributing to the obstruction of the CSF circulation. Surgical treatment of these pathogenic factors is the key to successful operation.

Pathogenesis of SM

CSF Obstruction in the Foramen of Magendie. The water-hammer theory was proposed by Gardner et al.,[5] who believed that "partial obstruction of the outflow of CSF from the fourth ventricle directs the systolic pulsations of CSF from the fourth ventricle through" a patent "central canal and transmits" a systolic "water-hammer" pressure wave "into the syrinx causing development and progression of syringomyelia." In the present study, we found that the tonsils overlying the foramen of Magendie, interstitial arachnoid adhesions, tonsil to medulla arachnoid adhesions, medialized tonsils, the vermian branch of the PICA, and an arachnoid veil or scarring were involved in the pathogenesis of SM, but only a few patients had SM communicating with the fourth ventricle. In most cases, the foramen of Magendie was partially obstructed and was often associated with other factors that contributed to SM. In case 15, we observed a complete impermeable veil obstructing the foramen of Magendie. SM caused by complete obstruction was typically characterized by the extent of the syrinx reaching the medulla oblongata, and the degree of syrinx was severe. The communication between the SM and the fourth ventricle could be observed at a specific level.

CSF Obstruction in the Foramen Magnum. The cranial-spinal pressure dissociation theory was proposed by Williams[14,22] and emphasizes the ball-valve effect of foramen magnum obstruction. The movement of intracranial CSF into the spinal canal is impeded and redirected into the central canal. Another representative of this theory is the "piston-like effect" by Oldfield et al.,[7,23] which suggests that as the piston-like cerebellar tonsils

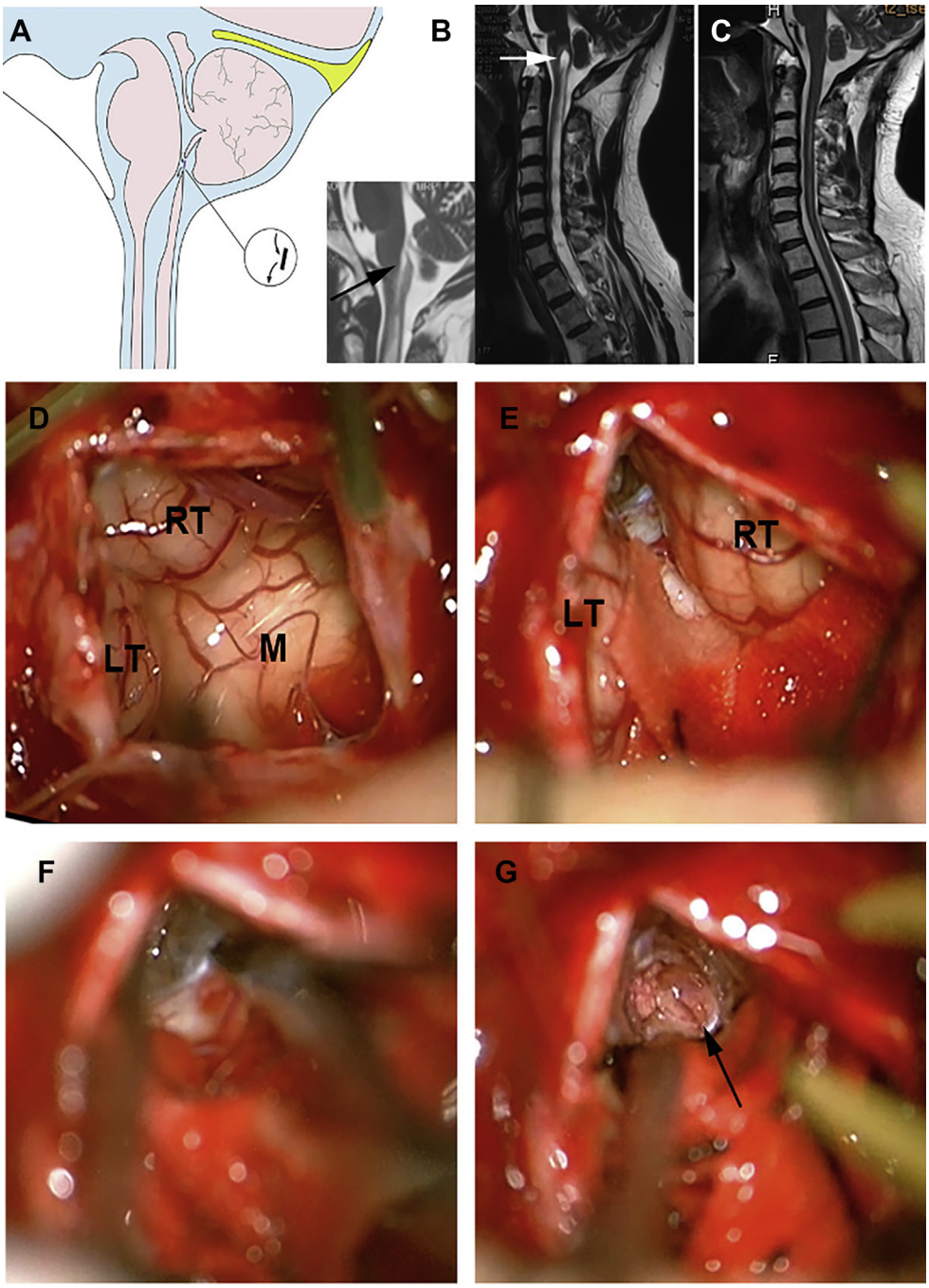

Figure 1. Case 15. (**A**) Schematic drawing of the foramen magnum region, sagittal view, illustrating the proposed mechanism of syringomyelia (SM) with obstruction of the foramen of Magendie. (**B**) Preoperative midsagittal T2-weighted magnetic resonance imaging suggestive of cervicothoracic SM without Chiari malformation type I, SM with syringobulbia (*white arrow*), and a detectable channel between the fourth ventricle and the syrinx (*black arrow*). (**C**) Postoperative magnetic resonance imaging displaying a marked reduction in size of SM. (**D**) After incision of the arachnoid, the tonsils are observed; no arachnoid opaque membranes or adhesions are found. (**E**) Nearly complete impermeable arachnoid veil overlying the foramen of Magendie. (**F**) Opening of the arachnoid veil. (**G**) Completely opened foramen of Magendie (*black arrow*). LT, left tonsil; M, medulla; RT, right tonsil.

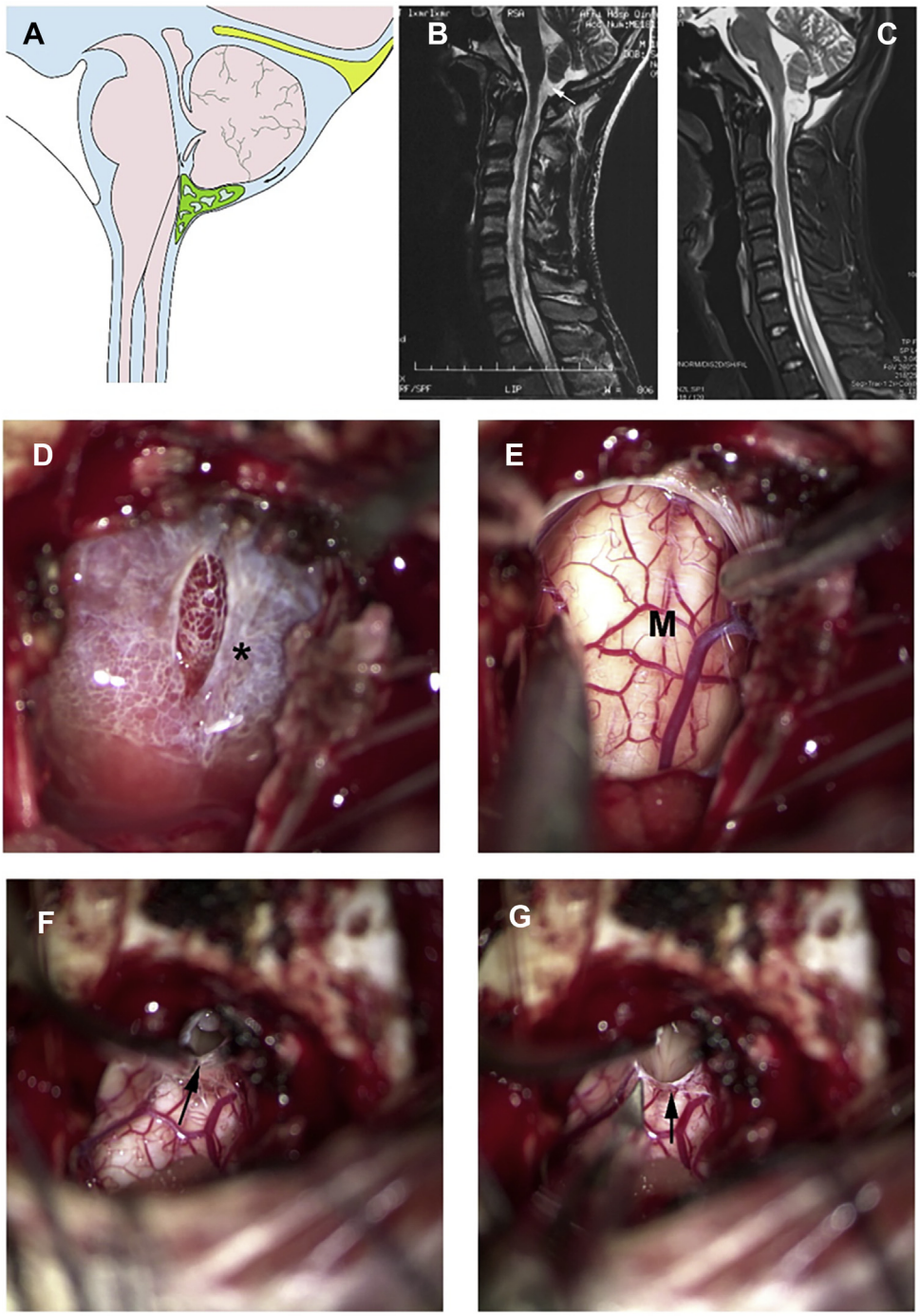

Figure 2. Case 21. (**A**) Schematic drawing of the foramen magnum, sagittal view, illustrating the projected mechanism of syringomyelia (SM) combined with obstruction of the foramen magnum. (**B**) Preoperative midsagittal T2-weighted magnetic resonance imaging suggestive of cervicothoracic SM without Chiari malformation type I and suspicious abnormal signals in the cisterna magna (*white arrow*). (**C**) Postoperative magnetic resonance imaging demonstrating a significant reduction in size of SM. (**D**) After incising the dura, a translucent arachnoid adhesion is observed extending from the retrocerebellar space to the level of the C1 perimedullary space (*asterisk*). (**E**) Microsurgical dissection and resection of the adhesion. (**F**) The foramen of Magendie is uncovered, in which the cerebrospinal fluid appears to flow freely (*arrow*). (**G**) Enlargement of the outlet of the foramen of Magendie (*arrow*). M, medulla.

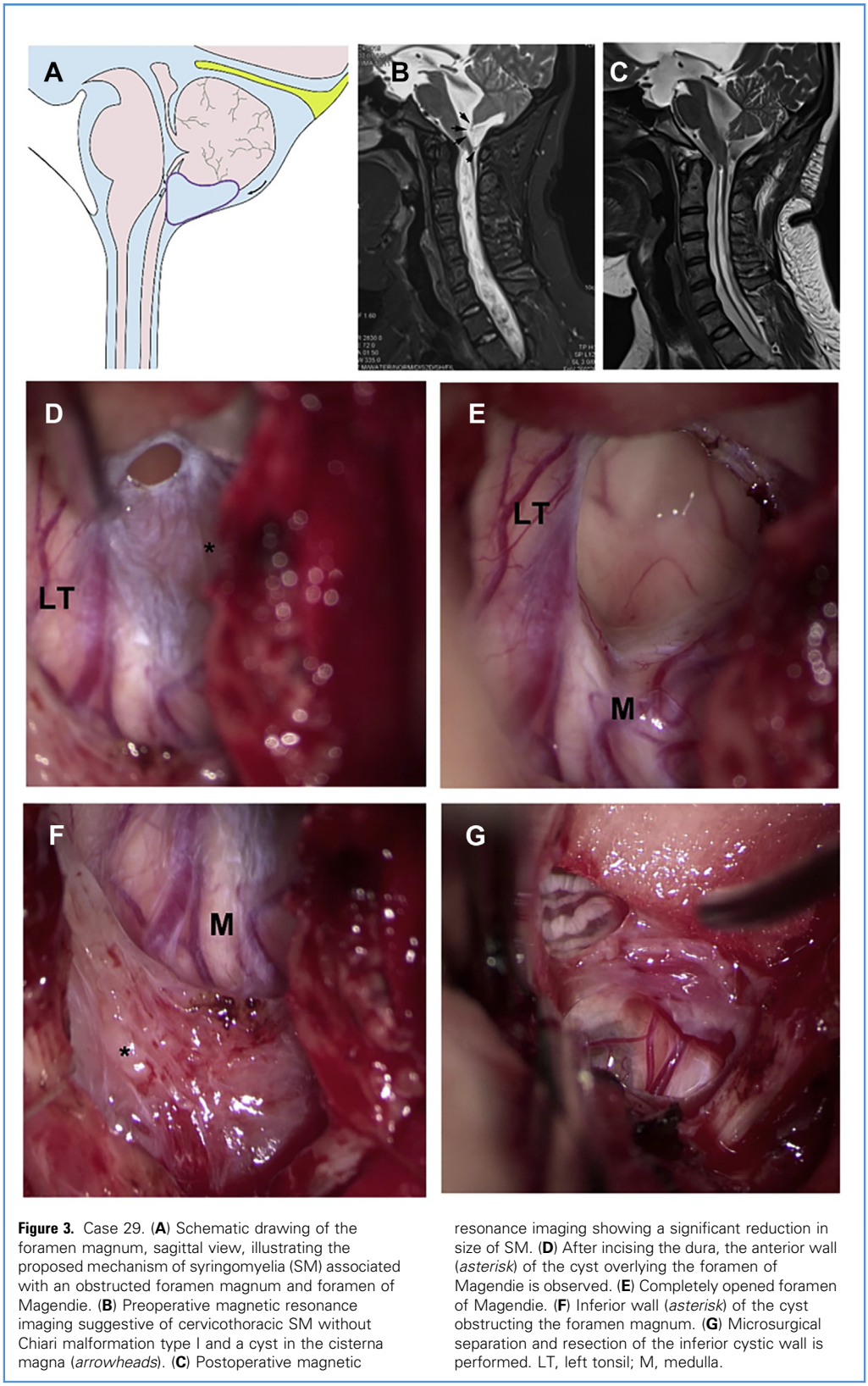

Figure 3. Case 29. (**A**) Schematic drawing of the foramen magnum, sagittal view, illustrating the proposed mechanism of syringomyelia (SM) associated with an obstructed foramen magnum and foramen of Magendie. (**B**) Preoperative magnetic resonance imaging suggestive of cervicothoracic SM without Chiari malformation type I and a cyst in the cisterna magna (*arrowheads*). (**C**) Postoperative magnetic resonance imaging showing a significant reduction in size of SM. (**D**) After incising the dura, the anterior wall (*asterisk*) of the cyst overlying the foramen of Magendie is observed. (**E**) Completely opened foramen of Magendie. (**F**) Inferior wall (*asterisk*) of the cyst obstructing the foramen magnum. (**G**) Microsurgical separation and resection of the inferior cystic wall is performed. LT, left tonsil; M, medulla.

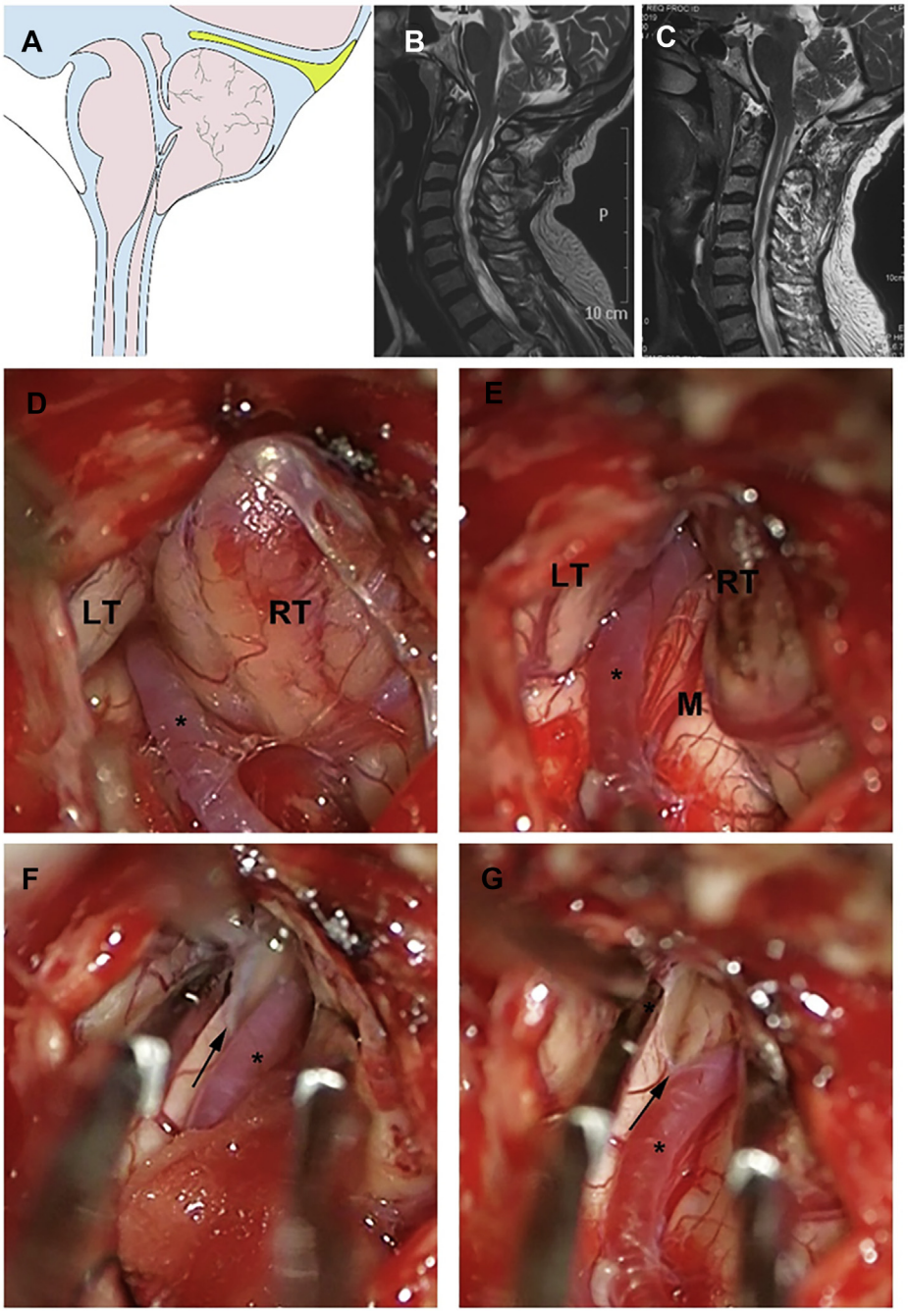

Figure 4. Case 32. (**A**) Schematic drawing of the foramen magnum, sagittal view, illustrating the proposed mechanism of syringomyelia (SM) associated with tonsils obstructing the foramen magnum and foramen of Magendie. (**B**) Preoperative magnetic resonance imaging demonstrates cervicothoracic SM with Chiari malformation type I. (**C**) Postoperative magnetic resonance imaging reveals a significant reduction in size of SM. (**D**) Incision of the dura shows the medialized tonsils, tonsils occupying the foramen magnum, and overlying foramen of Magendie. *Asterisk* indicates vermian branch of the posterior inferior cerebellar artery (PICA). (**E**) Retraction and low-power bipolar reduction of the right tonsil shows that the vermian branch of the PICA (*asterisk*) is obstructing the foramen of Magendie. (**F**) The foramen of Magendie is visible, and a complete veil (*arrow*) is observed. No cerebrospinal fluid is observed flowing through the foramen of Magendie. *Asterisk* indicates vermian branch of the PICA. (**G**) The vermian branch of the PICA (*asterisk*) is lysed, and the arachnoid veil (*arrow*) is removed. LT, left tonsil; M, medulla; RT, right tonsil.

Table 2. Intradural Pathology Associated with Cerebrospinal Fluid Obstruction*

	Intradural Pathology Associated with Cerebrospinal Fluid Obstruction	Patients (*n*)
Obstructing foramen magnum	Cisterna magna cyst	2/50
	Tonsil to dura mater arachnoid adhesions	4/50
Obstructing foramen magnum and foramen of Magendie	Tonsil occupying foramen magnum and overlying foramen of Magendie	44/50
Obstructing foramen of Magendie	Intertonsillar arachnoid adhesions	18/50
	Tonsil to medulla arachnoid adhesions	9/50
	Medialized tonsils	35/50
	Vermian branch of PICA obstructing foramen of Magendie	11/50
	Arachnoid veil obstructing foramen of Magendie	8/50

PICA, posterior inferior cerebellar artery.
*Upper: intradural pathology obstructing foramen magnum; middle: intradural pathology obstructing foramen magnum and foramen of Magendie; lower: intradural pathology obstructing foramen of Magendie.

move back and forth during cardiac cycles, increased systolic CSF power causes subarachnoid CSF to enter the spinal cord through the perivascular and interstitial spaces to form SM. Although the 2 theories differ in terms of the origin of CSF entering the central canal, they both suggest that CSF obstruction at the foramen magnum is the cause of SM. In this study, we found that tonsils occupying the foramen magnum, tonsil to dura mater arachnoid adhesions, and cisterna magna cysts could block the foramen magnum and participate in the development of SM. In case 21, we observed that CSF was obstructed only in the foramen magnum, while the foramen of Magendie was not obstructed, and SM did not communicate with the fourth ventricle. It was confirmed that CSF may enter the spinal cord via the perivascular space and the interstitial space to form SM.

Surgical Treatment of SM: FMMD

Because the pathogenesis of SM remains unclear, there is no uniform surgical procedure. Traditionally, there have been 2 common surgical methods for the treatment of SM: PFD and PFDD.[24,25] PFD is a simple procedure in which bone removal is performed, and the dura mater is opened; this has a certain effect in some cases, but there are complications, such as incomplete decompression, CSF leakage, intracranial infection, and postoperative fever.[26,27] PFDD is based on PFD, keeping the subarachnoid space intact, using an artificial dura (or autologous fascia) to repair the dura, and suspending the dura to maintain CSF patency.[24] This procedure does not involve opening the arachnoid membrane, and it does not address intradural factors that may cause obstruction to the CSF circulation; therefore, it does not relieve SM in some cases. In some studies, intraoperative ultrasound or MRI has been used during surgery to determine whether bone removal or duraplasty has provided sufficient space at the foramen magnum for normal pulsatile movement of CSF in the subarachnoid space at the level of the foramen magnum. However, all these studies were limited by the fact that CSF flow dynamics across the foramen magnum changed significantly when the patient was positioned for surgery; therefore, the results are unreliable.

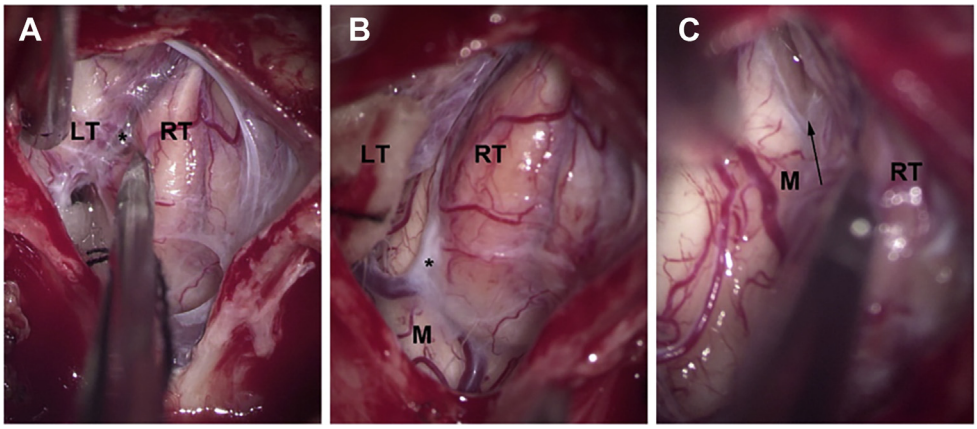

Figure 5. (**A**) Removal of the medialized tonsils shows intertonsillar arachnoid adhesions (*asterisk*). (**B**) Tonsil to medulla arachnoid adhesions (*asterisks*). (**C**) Lysed arachnoid adhesions and exposed foramen of Magendie (*arrow*). LT, left tonsil; M, medulla; RT, right tonsil.

Thus, with increased understanding of SM, we proposed that relieving the obstruction of CSF in the foramen magnum and foramen of Magendie is key to treatment. The treatment concept was changed from decompression to dredging. FMMD is the most fundamental method for the surgical treatment of SM. The key points of this technique are small bone window PFD, intradural exploration, and dural suture in situ.

Necessity of Intradural Exploration. For management of the dura, the choice is to remove only the posterior atlanto-occipital membrane or to remove only the outer layer of the dura, leaving an intact inner layer.[27] Conventional PFD and PFDD both involve incising the dura, but subarachnoid exploration is not performed. Both PFD and PFDD improve CSF circulation in the foramen magnum by resecting bony compression and augmenting the posterior cranial volume with extended repair of the dura mater, failing to identify intradural factors that may lead to higher reoperation rates.[28] If the disorder of CSF circulation is caused solely by blockage of the foramen magnum by the tonsils, the symptoms of patients with improved CSF circulation will be relieved after the expansion of the posterior fossa. However, in the present study, we found that in addition to tonsils occupying the foramen magnum, tonsil to dura mater arachnoid adhesions and cisterna magna cysts can also cause CSF circulation obstruction in the foramen magnum region. More importantly, CSF obstruction in the foramen of Magendie is an important factor contributing to SM formation, and tonsils overlying the foramen of Magendie, medialized tonsils, and intertonsillar arachnoid adhesions may achieve remission with the expansion of the posterior fossa. Tonsil to medulla arachnoid adhesions, the vermian branch of the PICA, and an arachnoid veil or scarring represent pathologic changes that hinder CSF circulation of the foramen of Magendie and must be removed by subdural exploration for relaxation. The statistics of this study demonstrated that the incidence of these intradural pathologies was significantly higher than expected. These pathologic changes often coexist, and the incidence may be as high as 20%. In this study, 50 patients were treated by opening of the dura mater to explore the subarachnoid space and the foramen of Magendie, a method that was effective for restoring CSF circulation. Postoperative MRI showed significant improvements in SM.

Dura Suturing In Situ. We did not advocate duraplasty during surgery. Although duraplasty expands the posterior fossa, the operation is time-consuming and provides space for the cerebellum to continue to herniate. Because the cerebellar tonsils of the lower hernia were fulgurated or removed for internal decompression to reconstruct the CSF circulation, the in situ suturing of the dura mater can play a role in supporting the contents of the posterior fossa; that is, secondary herniation of the cerebellum and medulla oblongata can be avoided if the dural tension is too high to be sutured in situ. Additionally, a small amount of fascia or muscle is required for repair. No related complications, such as CSF leakage, occurred in this group of patients.

In the literature, a large number of studies have been conducted on the effectiveness of dural opening,[9,29,30] and surgeons must compare the risk of severe complications caused by dural opening with the risk of missing pathology at the foramen of Magendie and foramen magnum, which cannot be relieved by bone decompression alone. Many surgeons do not open the dura for such cases, even with syrinx. In other centers, opening the dura does not seem to matter so much. It has been shown in the literature that SM associated with Chiari malformation can be resolved by using other surgical techniques.[31] Moreover, some authors have suggested that arachnoid scarring is associated with an unfavorable long-term outcome.[32,33] In our experience, intradural pathology is present in all patients with persistent or worsening syrinx.[12] Dlouhy et al.[15] and Klekamp[33] provided data on foramen of Magendie obstructions. Furthermore, Klekamp[33] described arachnoid adhesions in his article, providing a classification for it and results suggesting that these arachnoid features do have an impact on postoperative outcomes. These observations support the data given here. The aim of surgery was to precisely relieve the cause of obstruction rather than blind decompression. By using the FMMD procedure, factors that caused obstruction of the CSF circulation of the foramen magnum and the foramen of Magendie were relieved, and the normal circulation of the CSF was restored, thus eliminating the cause of disease, preventing the development and deterioration of lesions, reducing the size of the SM after surgery, and resulting in the rapid recovery of symptoms and the possibility of low rates of recurrence. In addition, the posterior occipital incision was 3–4 cm, and blood loss during the entire operation was approximately 20–30 mL. The incidence of postoperative reactions and complications was significantly reduced, and the mean hospital stay was nearly 1 week.

Limitations

Limitations of this research include its retrospective design, selection of a cohort of patients with no control subjects, and short follow-up time.

CONCLUSIONS

There are many pathologic changes in the intradural space of patients with SM, and these changes are the cause of CSF circulation obstruction. Intradural pathology was found during surgery in all patients in this study. FMMD may be the fundamental surgical treatment for SM.

CRediT AUTHORSHIP CONTRIBUTION STATEMENT

Jian Guan: Writing - original draft, Data curation. **Chenghua Yuan:** Writing - review & editing, Visualization. **Can Zhang:** Software. **Longbing Ma:** Methodology. **Qingyu Yao:** Software, Writing - original draft, Data curation. **Lei Cheng:** Software. **Zhenlei Liu:** Methodology, Resources. **Kai Wang:** Resources. **Wanru Duan:** Resources. **Xingwen Wang:** Resources, Visualization. **Hao Wu:** Resources. **Zan Chen:** Resources. **Fengzeng Jian:** Writing - review & editing, Project administration.

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