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ORIGINAL PAPER

Posterior Fossa Decompression Combined with Resection of the Cerebellomedullary Fissure Membrane and Expansile Duraplasty: A Radical and Rational Surgical Treatment for Arnold–Chiari Type I Malformation

Chen Jian Liang · Qiu Jian Dong · You Heng Xing · Ma Shan · Lian Xiao Wen · Zhong Yuan Qiang · Zhang Qing Ping · Peng Zhi Tao · Huang Xiao Ping

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Abstract This study aims to introduce a new surgical procedure for the treatment of Arnold–Chiari type I malformation (ACM-1) and to compare its effectiveness with the techniques described in previous studies. We performed the following procedures: foramen magnum decompression combined with the removal of both the atlanto-occipital membrane, extended resection of the cerebellomedullary fissure arachnoid membrane, and artificial duraplasty to enlarge the membranic posterior fossa without resecting the cerebellar tonsils and syringosubarachnoid shunting. There were 21 ACM-1 patients: 12 cases had osteo-compression on the cerebellar hemisphere, 18 cases had thickened adhered fabric ring that stretched from arachnoid membrane to cerebellar hemisphere, and 15 cases with syringomyelia. The patients were followed up for 6 months to 3 years after the surgery. All patients showed a remarkable recovery of syringomyelia. There were no morbidity or death related to the surgery. Most of ACM-1 patients, the osteo- and membrane compression on cerebellar hemisphere and tonsil were observed during the operation. Therefore, decompression of foramen magnum and posterior craniocervical combined with the removal of cerebellomedullary fissure arachnoid membrane and placement of an artificial dural graft should be considered as a comprehensive option of minimally invasive surgery and rational and radical treatment of ACM-1. Our experience showed that, by using our procedure, shunting becomes no longer necessary in the treatment of ACM-1-associated syringomyelia.

Keywords Arnold–Chiari type I malformation · Foramen magnum decompression · Cerebellomedullary fissure · Arachnoid membrane · Treatment · Efficacy comparison

C. J. Liang (✉) · Q. J. Dong · Y. H. Xing · M. Shan · L. X. Wen · Z. Y. Qiang · Z. Q. Ping · P. Z. Tao · H. X. Ping
Department of Neurosurgery, The Affiliated Futain Hospital of Guangdong Medical College, The Fourth Hospital of Shenzhen, Shenzhen P.O. 0086 518033, Guangdong, People's Republic of China
e-mail: szftsjwk@163.com

H. X. Ping (✉)
Neurosurgical Department, Traditional Chinese Medical hospital of Shenzhen, Shenzhen P.O. 0086 518033, Guangdong, People's Republic of China
e-mail: hwangxiao000123@163.com

Introduction

Posterior craniocervical decompression is the most commonly used surgical technique to treat Arnold–Chiari type I malformation (ACM-I) with or without syringomyelia and in the absence of hydrocephalus. We reviewed various surgical treatments reported in the literature, and present our series of 21 patients diagnosed with ACM-1 who underwent suboccipital craniectomy and a C_1 (or C_1/C_2) laminectomy, as well as foramen magnum opening, Y-shaped dural incision with extended removal of cerebellomedullary fissure arachnoid membrane, and expansile duraplasty with the use of synthetic grafting material. The outcomes were analyzed during a follow-up period of 6 months to 3 years. W compared our technique with other surgical treatments reported in the literature. The comparison shows that this type of posterior fossa decompression leads to better outcomes with minimal complications and side effects. It also shows that shunting is no longer an appropriate method for the treatment of syringomyelia.

Materials and Methods

Clinical Materials

From June 2003 to March 2011, 21 ACM-1 patients underwent posterior fossa decompression combined with extended removal of cerebellomedullary fissure arachnoid membrane and C_1 (or C_1/C_2) laminectomy. Fifteen of the patients were male and six were female, and their ages ranged from 17 to 47 years (mean 34.4 years). The patients have been with ACM-1 for an average of 2.4 years (range, 1–5 years).

Clinical Presentations

Three patients reported headache on cervical occipital region; eight complained of dump of both the upper limbs; seven suffered from inability to feel pain and increased sensitivity to high temperature, and three reported dizziness.

Imaging

All 21 patients underwent plain radiography of their cervical spine. Basilar invagination was diagnosed if the odontoid was above palatine-occipital line. All patients underwent magnetic resonance imaging (MRI), and 15 were diagnosed of syringomyelia.

Surgical Procedures

Under general anesthesia, the patients were laid in a flexion prone position, and an incision in occipital-neck midline skin was made. The procedures included a bony decompression of enlarged foramen magnum and removal of the occipital squama (with the diameter of 4–6 cm of bone window). The posterior arch of the atlas was also removed. Patients diagnosed with syringomyelia underwent laminectomy of the upper portion of C_2. During operation, 12 patients were found to have compressed cerebellum due to proliferated occipital squama, six patients with thick and compressed atlanto-occipital fascia. Eighteen patients were also found to have arachnoid membrane adhesion, and nine with cerebellar topsils downward to C_2.

The thicken atlanto-occipital fascia was incised and the dura opened with Y-shaped incision under a surgical microscope. The cerebellomedullary fissure arachnoid membrane was opened along topsils upward to the cerebellum lateral, and arachnoid membrane of cerebellum and topsils was separated and removed. The arachnoid of the upper spinal cord was then removed, and the topsils, medulla oblongata, and upper spinal cord were separated and released. When topsils were pushed upward 2–3 cm, the cerebellomedullary cistern can be reestablished. The bone window with diameter of 4–6 cm was left open to enlarge the posterior fossa. Then, expansile duraplasty with artificial membrane was performed to incorporate the dura. No shunting and puncturing of spinal cavity was performed for ACM-1-associated syringomyelia.

Results

Based on the criteria of Tator et al. [1], 13 patients experienced improvement and symptoms disappeared completely. Conditions of six patients also improved, and two of them did not experience any progression of their symptoms. The patients were followed up for 6 months to 3 years, and MRI performed during this time frame showed that the compression at the craniocervical junction eased and that syringomyelia abated. Two patients experienced postoperative fever, which was treated with intravenous antibiotics. One patient experienced subcutaneous hydroma and failed medical treatment for more than 20 days. This patient, however, completely recovered after the removal of the dura graft. No patient died.

Representative Case Report

A 25-year-old male patient was admitted because of dizziness, loss of temperature sensation and thigmesthesia in his left hand. He reported experiencing dizziness 2 months prior to admission. One month later, he reported that his left hand could not distinguish the difference between steel and wood and that he lost temperature sensation. An MRI revealed ACM-1 with syringomyelia. He underwent surgery, which involved a Y-shaped incision in the dura. However, the cerebellum and medulla oblongata cistern could not be detected due to cerebellum herniation. The topsils herniated downward to the upper portion of C_2. After opening the cerebellomedullary fissure arachnoid membrane and dissecting the lateral side of the cerebellum, fibronectin was found beneath the arachnoid. Many indentations on the cerebellum were also found. With the incision of arachnoid membrane, a free space was obtained. Pushing upward topsils, it can retract 3 cm, the dura were closed with a 4 cm $\times$ 6 cm^2 graft to incorporate it. In the following days, the patient reported that his dizziness disappeared and the dump of his left hand and return of temperature sensation. One month later, an MRI revealed that the cerebellar topsils retract to its position and syringomyelia almost disappeared (Fig. 1).

Discussion

ACM-1 is a congenital disease characterized with a downward herniation of the cerebellar topsils, most of

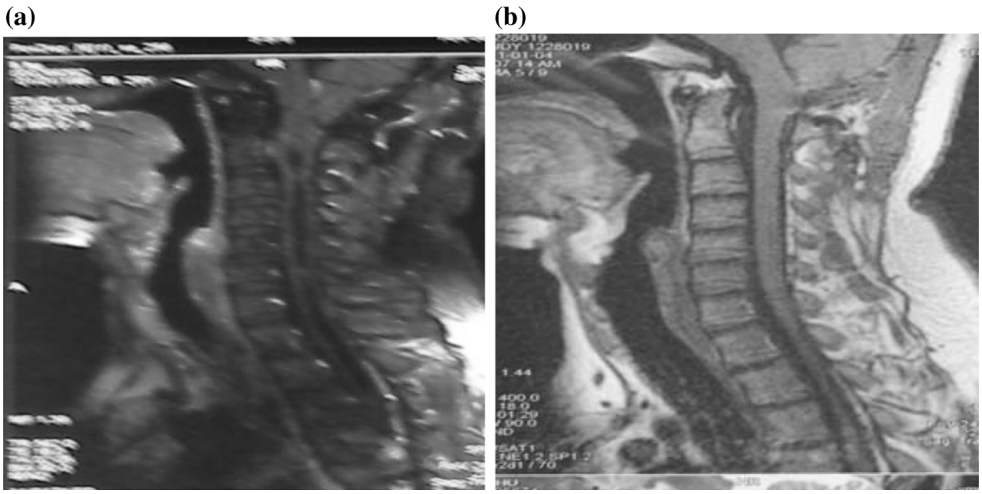

Fig. 1 MRI ACM-1 with syringomyelia. **a** Preoperation; **b** 3 months postoperation

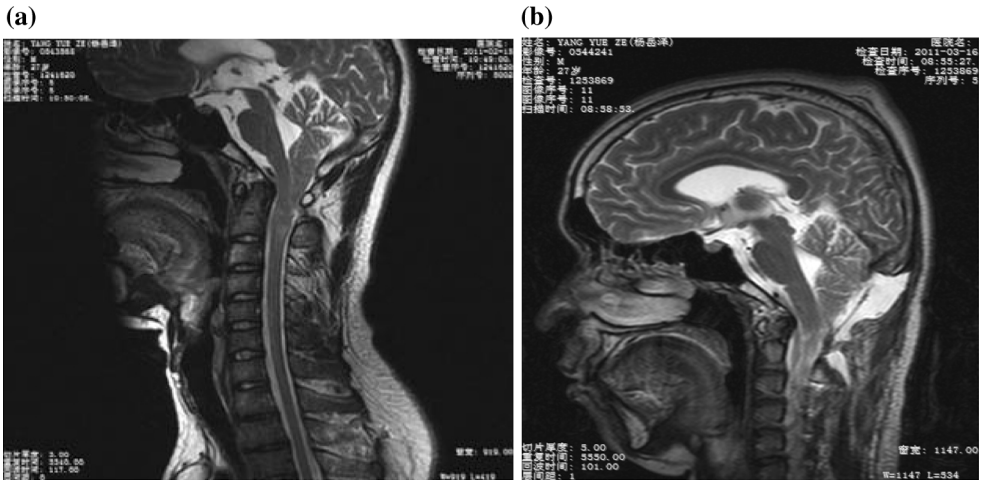

Fig. 2 ACM-1 without syringomyelia. **a** Preoperation; **b** 3 months postoperation

them with syringomyelia [2–5]. Its mechanism is still unknown, although it is thought to result from the failure of the normal occipital bone structure to form during embryonic development, resulting in a small posterior fossa and forcing part of the cerebellum into foramen magnum. Most cases of this disease are accompanied with abnormal cerebrospinal fluid (CSF) dynamics. The purpose of surgery is to restore the impeded flow of CSF through the subarachnoid space at the cranio-cervical junction and simultaneously to enlarge the posterior fossa to decompress the brain stem from the downward cerebellum topsils and recover the function of spinal cord (Fig. 2).

Most ACM-1 patients also have basilar invagination. Some of them have bony hyperplasia and thicken atlanto-occipital fascia resulting in compression of the cerebellum and occasionally with thickened arachnoid.

Once patients present symptoms of ACM-1, surgery should be considered. There are many surgical techniques available, but decompression of the foreman magnum and the posterior fossa is the basic method, which can be used particularly in patients with basilar invagination. This can resolve bony compression and can enlarge the cavity of the posterior fossa [4]. There is still no consensus regarding the removal of the occipital bone. Some experts advocate an extensive removal of bone overlying the posterior fossa 6 cm $\times$ 6 cm to increase the volume of the posterior fossa. However, this view is not shared by all surgeons. Some think that extensive bone removal of the posterior fossa may cause the hindbrain to migrate downward after decompression for hindbrain hernia [5, 6].

Duraplasty is also controversial. Some experts insist that after suboccipital decompression and resection of the posterior arch of C_1 or C_2 (depending on the extent of the

caudal displacement of the tonsils), duraplasty is necessary and can either use autologous (pericranium) or synthetic grafting. Complication of duraplasty (e.g., CSF leakage) should also be prevented. A previous study involving 30 patients who underwent duraplasty (18 with artificial graft and 12 with autologous pericranium graft) reported that six patients experienced CSF leakage regardless of the type of duraplasty used [7]. Other experts insist that opening the outer dura layer and leaving the inner one intact will prevent complication by allowing the remaining layer to stretch sufficiently after surgery [8]. However, other experts prefer to open the dura to allow a more complete and immediate enlargement of the subarachnoid space [9]. Therefore, we consider that the dura must be opened by which adhesive bands could be discovered and excised to reset the herniated cerebellum or its topsils. In this study, only the last 8 patients underwent duraplasty with artificial graft membrane. We left the dura open in the first 13 patients due to limit of our experience.

Syringo-subarachnoid shutting is most commonly used treatment for associated syringomyelia. Hida et al. [10] reported that clinical symptoms and radiological findings improved much more quickly in patient that underwent syringe-subarachnoid shunting than those who underwent foramen magnum decompression. However, other experts have different opinion. Sindou and Gimbert reported [11] that syringomyelia associated with ACM-I must be treated by craniocervical decompression alone. Shunting also no longer appears to be an appropriate method for the treatment of syringomyelia. We believe that the main cause of syringomyelia is the obstruction of CSF due to foramen magnum malformation and cerebellar topsils herniation. In our experience, CSF circulation can be restored by decompressing posterior fossa. Hence, shunting and puncturing the spinal cavity are unnecessary in ACM-1-associated syringomyelia.

Nevertheless, for a comprehensive decompression of the posterior fossa and to restore CSF circulation, more attention should be paid to the influence of the arachnoid membrane. Arachnoid membrane adhesion at the major cistern and surperficial of cerebellum is very common. Some even form rough strings or brands compressing cerebellum. If the arachnoid membrane is not opened, the adhesion would not be seen, and this was not mentioned in previous studies. We suggest that the intra-arachnoid procedures be performed in every ACM-1 patient, so that complete decompression of the hindbrain and restoration of CSF flow can be achieved. In this study, patients underwent opening of the arachnoid membrane and resection with extended cerebellomedullary of fissure arachnoid membrane approaching, and we achieved exciting results.

It is still a matter of debate whether the cerebellum topsils be removed [12]. However, we believe that doing so is not helpful. None of our patients underwent resection of the cerebellum topsils, and after follow-up 18 patients were observed to have experienced retraction of the herniated cerebellar topsils and improvement of the syringomyelic cavity.

After studying the currently available surgical procedures, we found that it is necessary to employ a more rational and radical procedure. Therefore, we explore a new surgical technique, which consists of suboccipital craniectomy and a C_1 (or C_1/C_2) laminectomy, foramen magnum opening, an atlanto-occipital fascia release accompanied by incision and removal of the cerebellomedullary fissure arachnoid membrane and duraplasty, an expansile duraplasty using artificial material, and separate and free cerebellum topsil without syringe-subarachnoid shunting.

All 21 ACM-1 patients with basilar invagination experienced good surgical outcomes. Based on our findings, the followings points should be emphasized: (1) The size of the bone to be removed is about 4 cm × 6 cm to achieve enough decompression and to prevent displacement of cerebellum and brain stem. (2) This effective decompression can restore the dynamics of CSF flow at the craniocervical junction; thus, the compression could be released totally. (3) In order to reset the herniated cerebellar topsils, 1.5–2 cm of the posterior arch should be resected. Partial laminectomy of the C_2 is sometimes necessary to restore CSF circulation. (4) Further decompression could be achieved by a thorough excision of the atlanto-occipital fascia and of the proliferated tissue of epidural and by opening the upper dura of C_2. (5) When the sharp angle resulting from basilar invagination becomes a relatively obtuse angle and the bony compression of foramen magnum is released, the operation is done. (6) Arachnoid adhesion and upper bilateral arachnoid of cerebellum should be completely removed with the opening of cerebellomedullary fissure arachnoid membrane. (7) Adhesion between the bilateral cerebellar topsils and the brain stem and upper spinal cord should be separated so that the cerebellar topsils can be fully freed. (8) The dura graft incorporated into the dura should be sutured without any tension. (9) For the associated syringomyelia, syringe-subarachnoid shunting is unnecessary.

In patients with Chiari I malformation with basilar invagination, treatment should aim at restoring CSF flow to decompress the cerebellum spinal cord and cranial nerves. Bone decompression of the posterior fossa and foramen magnum and atlanto-occipital fascia release is insufficient for arachnoid adhesive bands of cerebellum, making it necessary to perform bony and arachnoid membrane decompression. Resetting the cerebellar topsils 2 cm upward will achieve better result. Therefore, we conclude that combined foramen magnum and posterior

craniocervical decompression, removal of cerebellomedullary fissure arachnoid membrane, and placement of an artificial dural graft are a comprehensive, minimally invasive, and rational and radical surgical technique to treat ACM-1.

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