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Cerebellar Tonsillectomy with Suboccipital Decompression and Duraplasty by Small Incision for Chiari I Malformation (with Syringomyelia): Long Term Follow-up of 76 Surgically Treated Cases

Siringomyeli ile Birlikte Chiari I Malformasyonu için Küçük İnsizyonla Suboksipital Dekompresyon ve Duraplasti ile Serebellar Tonsillektomi: Cerrahi Olarak Tedavi Edilen 76 Olgunun Uzun Dönemli Takibi

Junpeng MA[1], Chao YOU[1], Haifeng CHEN[1], Siqing HUANG[1], Chengcheng IEONG[2]

[1]West China Hospital, Department of Neurosurgery, P. R. China
[2]Sichuan University, Westchina Medical School, P. R. China

Correspondence address: Siqing HUANG / E-mail: huangsiqing28@163.com

ABSTRACT

AIM: To explore the surgical effect of cerebellar tonsillectomy with suboccipital decompression and duraplasty by small surgical incision (3~4cm around the foramina magnum) on treating Chiari I Malformation (CM I) patients.

MATERIAL and METHODS: A retrospective study was undertaken on 76 CM I patients treated by this surgery. The surgical efficacy on clinical symptoms and syringomyelia were overall evaluated and analyzed.

RESULTS: The study included 76 cases (36 men and 40 women; age range, 5–58 years; mean age at surgery, 38.8 years). Preoperative MRI confirmed 56 cases associated with syringomyelia. The follow-up period ranged from 25 to 58 months (median, 46 months). At the end of follow-up, 61 patients (80.26%) had improved, 12 patients (15.79%) were stabilized, and 3 patients (3.95%) had worsened. In MRI scan, syrinx of 55 patients (98.21%) had improved or stabilized compare with syrinx growth in only one patient (1.79%). Statistical analysis reveal the surgical effect tend to be much better in patients with short duration of preoperative symptoms (P=0.001).

CONCLUSION: Cerebellar tonsillectomy with suboccipital decompression and duraplasty can provide long-time cure for most CM I cases. Early diagnosis and surgery is necessary to improve the surgical effect. A 3~4cm incision around foramen magnum is enough for these operations and may be conducive to reduce postoperative complications.

KEYWORDS: Chiari malformation, Syringomyelia, Small incision, Surgery, Tonsillectomy, Suboccipital decompression

ÖZ

AMAÇ: Chiari I Malformasyonu (CM I) hastalarında küçük cerrahi insizyonla (foramina magnum etrafında 3~4 cm) suboksipital dekompresyon ve duraplasti ile serebellar tonsillektominin cerrahi etkisini inceleme.

YÖNTEM ve GEREÇLER: Bu cerrahiyle tedavi edilen 76 CM I hastasının retrospektif bir çalışması yapıldı. Cerrahinin klinik belirtiler ve siringomyeli üzerindeki etkinliği genel olarak değerlendirildi ve analiz edildi.

BULGULAR: Bu çalışmaya 76 olgu (36 erkek ve 40 kadın; yaş aralığı 5-58 yaş; cerrahi sırasında ortalama yaş 38,8 yıl) alındı. Preoperatif MRG 56 hastada ilişkili siringomyeli gösterdi. Takip dönemi 25 - 58 ay arasındaydı (medyan 46 ay). Takip sonunda 61 hasta (%80,26) daha iyiydi, 12 hasta (%15,79) stabilize olmuştu ve 3 hasta (%3,95) daha kötüydü. MRG taramasında, 55 hastanın (%98,21) sirinksi daha iyi veya stabilize durumdayken sadece bir hastada sirinkste büyüme olmuştu (%1,79). İstatistiki analiz cerrahi etkinin preoperatif belirtilerin süresinin daha kısa olduğu hastalarda çok daha iyi olma eğiliminde olduğunu gösterdi (P=0,001).

SONUÇ: Suboksipital dekompresyon ve duraplasti ile serebellar tonsillektomi çoğu CM I olgusu için uzun dönemli düzelme sağlayabilir. Cerrahinin etkisini arttırmak üzere erken tanı ve erken cerrahi gereklidir. Bu ameliyatlar için foramen magnum etrafında 3~4 cm insizyon yeterlidir ve postoperatif komplikasyonları azaltmak konusunda etkili olabilir.

ANAHTAR SÖZCÜKLER: Chiari malformasyonu, Siringomyeli, Küçük insizyon, Cerrahi, Tonsillektomi, Suboksipital dekompresyon

INTRODUCTION

Chiari I malformation (CM I) is a congenital disease characterized by downward decent of the cerebellar tonsil and crowding in the craniocervical junction area, which was first described by Hans Chiari over one century ago (5). Generally, about 50~70% CM I cases are associated with syringomyelia (SM), which will slowly lead to chronic and sometimes irreversible myelopathy (4, 9, 12, 20). Surgery is the only way to cure this disease. There are still obvious controversies in current surgical strategies although much operative progress has been made in the last few decades and the expansion of posterior fossa volume has been widely accepted as the surgical goal (1-3, 9, 12, 14, 16).

Aiming to explore the surgical effect of cerebellar tonsillectomy with suboccipital decompression and duraplasty together to reduce the risks and postoperative complications of this disorder, we have performed these operations on patients with CM I using a small incision (3~4cm) since 2005. A total of 76 CM I cases underwent this small incision surgery at our department from January 2006 to October 2007. The postoperative complications and its efficacy regarding the clinical symptoms and SM in CM I patients were retrospectively evaluated overall by long-term follow-up and analyzed in this report.

PATIENTS and METHODS

A total of 76 symptomatic patients with CM I (36 men and 40 women; age range, 5–58 years; mean age at surgery, 38.8 years) were treated by surgery at the Department of Neurosurgery of our hospital between January 2006 and October 2007. Preoperative MRI scan revealed that 56 cases were associated with SM. We retrospectively analyzed the surgical results with a minimum of 2 years of postoperative follow-up (mean 46 months). All included patients have received at least 1 preoperative and 1 postoperative MRI scan in this study.

Preoperative Clinical Symptoms

Preoperative neurologic examinations were routinely performed. Mean duration of symptoms from origin to surgery amounted to 3.5 years (2 months to 12 years). Sensory disturbance (30.36%), pain (26.79%) and motor weakness and muscular atrophy (21.43%) were the three main symptoms in CM I patients with SM. Pain (35%), cerebellar dysfunctions (25%) and cranial nerve dysfunctions (20%) were the three main symptoms in patients without SM. Further details are presented in Table I.

Preoperative Imaging

Patients generally underwent magnetic resonance imaging (MRI) preoperatively and the diagnosis of CM I was defined as tonsillar herniation extending at least 5mm below the foramen magnum without meningocele and a descent of the cerebellum and the fourth ventricle (15). The degree of the tonsils' downward displacement was classified as three types: I°**:** 0.5~1cm below the foramen magnum; II°**:** 1~2cm below the foramen magnum; and III°**:** beyond 2cm below the foramen magnum (Table II). SM was found by preoperative MRI in 56 patients. Three-dimensional reconstruction scan of the cervical vertebrae was performed in 13 cases to exclude suspected atlanto axial dislocation.

Surgical Technique

All patients in this series were operated on under general anesthesia, in the left lateral position. We used a 3~4cm surgical incision in the midline around foramen magnum (Figure 1). Following a clean incision for each layer of the skin and muscles, suboccipital decompression and a laminectomy of the atlas were performed to form a bone window with the size about 2×3 cm. This limited surgical incision and relevant suboccipital decompression is sufficient to expose the cerebellar tonsil (Figure 2). After bone removal, we opened the dura in "Y" format and dissected the arachnoid adhesion among the cerebellar tonsils, medulla oblongata and spinal

Table I: Preoperative and Postoperative Symptoms/signs in 76 CM I Patients

Symptom/Sign	Preoperative Status (%) (No. of patients)		Postoperative result (No. of patients)	
	CM+SM	CM	CM+SM	CM
Pain	15 (26.79%)	7 (35%)	1/2*	1
Sensory disturbance	17 (30.36%)	1 (5%)	4	---
Weakness & muscular atrophy	12 (21.43%)	1 (5%)	4/1*	---
Gait problems or Gait ataxia	9 (16.07%)	5 (25%)	1	---
Cranial never dysfunction	2 (3. 58%)	4 (20%)	---	1
Nystagmus	1 (1. 79%)	---	---	---
Vomiting	---	1 (5%)	---	---
Dizziness	---	1 (5%)	---	---
Total	**56**	**20**	**10/3**	**2/0**

The figure behind "/" represent the case load of symptomatic deterioration. Only the major complaint or problem is listed.* ***CM:* *Chiari malformation,* ***SM:*** *syringomyelia. Patients with symptoms caused by limited posterior fossa volume (brainstem or cerebellar symptoms) may improve (91.30%) more than those with myelopathy (70.97%) and radiculopathy (81.82%), but there is no statistical significance (P=0.169).*

Table II: Summary of Preoperative Status and Postoperative Grade in 76 CMI Patients Grouped by the Presence of Preoperative SM, Preoperative Degree of the Tonsils' Downward Displacement

Postoperative grade	Preoperative Status CM+SM (No. of patients)				Preoperative Status CM (No. of patients)			
	I	II	III	Total	I	II	III	Total
A	28	13	2	43	14	4	0	18
B	7	3	0	10	1	1	0	2
C	2	1	0	3	0	0	0	0
Total	37	17	2	56	15	5	0	20

***A:** good result (cure and obvious improvement); **B:** stabilization of symptom; **C:** bad result with symptom deteriorated. Degree of the tonsils' downward displacement: **I°:** 0.5~1cm below the foramen magnum; **II°:**1~2cm below the foramen magnum; and **III°:** beyond 2cm below the foramen magnum. There is no significant difference in the surgical effect among patients grouped by preoperative degree of the tonsils' downward displacement (P=0. 960) and the presence of SM (P=0.374).*

Figure 1: 3~4cm surgical incision in the midline around the foramen magnum. The broken line represents conventional surgical incision.

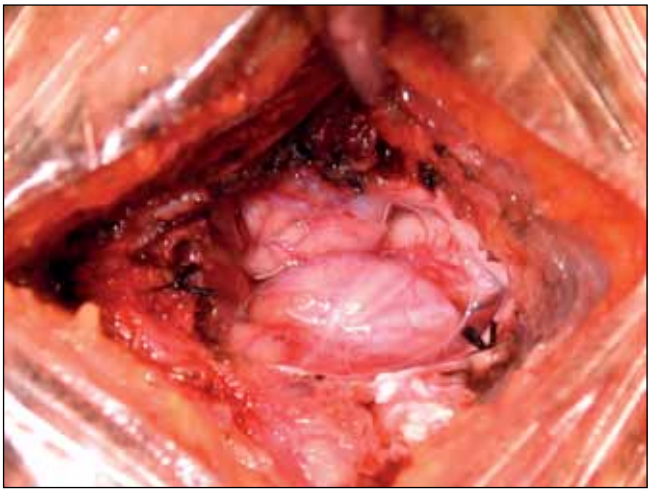

Figure 2: Intraoperative view after the dura is opened in "Y" format through median suboccipital craniotomy. The cerebellar tonsil is clearly visualized.

cord. Cerebellar tonsillectomy was then performed by subpial resection. Subpial operation is necessary for this step in order to protect the surrounding structures (such as arteriae cerebelli inferior posterior). Owing to very small incision, it was improper to use autogenous fascia for duroplasty. Therefore, we used a dural onlay graft (product of Johnson& Johnson Company) to enlarge the posterior fossa and reconstruct a spacious Cisterna Magna. Finally, the incision was meticulously closed in anatomical layers and no drain was used. All of the surgeries were conducted by Prof. Huang and Prof. Chen at our department with the same standard surgical technique.

Statistical analysis

The statistical analyses were conducted to assess the different surgical effect on specific variables. We used the chi-square test to establish the presence of significant differences in clinical outcome (cure or obvious improvement, stabilization, deterioration) among patients grouped by preoperative degree of the tonsils' downward displacement, patients with or without syrinx and the duration of preoperative symptoms. Statistical significance was established at a *P* value of less than 0.05.

RESULTS

Early postoperative result and complications

Six patients (7.89 %) suffered from progressive headache after surgery. The blood contained cerebrospinal fluid (CSF) was considered and continued lumbar CSF drainage successfully resolved the symptoms. Three patients (3.95%) had definite intracranial infection (positive CSF germiculture). Seven patients (9.21%) had isolated fever with no infection (normal lumbar puncture and biological test results). Only one patient (1.32%) had postoperative CSF leakage. All these postoperative transient complications were cured during the duration of hospital stay. There was no case with deterioration of neural function before discharge. The average postoperative hospital stay was 4.7 days.

Progression of postoperative symptoms

Patients were followed up by letters, telephones and outpatient follow-up visits. We classified the progression of postoperative symptoms into three categories: grade A: good result (cure or obvious improvement); grade B: stabilization of symptoms; grade C: bad result with deteriorated symptoms**.** The mean postoperative follow-up period was 46 months

(range, 25–58 months). All the patients were available to be asked to judge personally the effect of surgery and postoperative symptoms. At the end of follow up, 61 patients (80.26%) had improved, 12 patients (15.79%) were stabilized, and 3 patients (3.95%) had worsened. The details of both preoperative and postoperative clinical symptoms are summarized in Table I, II and Table III. In the three patients with worsening postoperative symptoms during the follow-up period, one patient was associated with enlarged syringomyelia in postoperative MRI scan and reoperation was required. One patient complained of additional paresthesia, pain in the limbs and insomnia with the improvement of syrinx on the MRI scan and a negative physical examination. These subjective symptoms were relieved after intermittent sedative treatment and psychotherapy. The other patient thought his condition had worsened with progressive anaesthesia and the pain in the limbs, together with the disappearance of syrinx on the MRI. The exact reason for symptomatic deterioration is still unknown but we considered that syrinx shrunk too fast and the translocated spinal cord could drag the nerve root and cause increased symptoms.

On statistical analysis, there was no significant difference in the surgical effect among patients grouped by preoperative degree of the tonsils' downward displacement (P=0.960) and patients with or without syrinx (P=0.374). Patients with symptoms caused by limited posterior fossa volume (brainstem or cerebellar symptoms) improved (91.30%) more than those with myelopathy (70.97%) and nerve root symptoms (81.82%), but there was no statistical significance (P=0.169). The surgical effect tended to be much better in patients who had a short duration of preoperative symptoms (P=0.001).

Reoperation

One patient underwent syringo-subarachnoidal shunting surgery and a shunt tube was placed in the dorsolateral subarachnoid space at 1 year after surgery because of persistence of symptoms and exacerbated syringomyelia. He had a good prognosis (obvious remission of initial symptoms and shrunken syrinx on MRI) at the 2.5-year follow-up after the shunt surgery.

MRI Follow-up

MRI was routinely performed at three months and 1 year postoperatively. At the one year postoperative follow-up, MRI was available for all patients with SM. The syrinx almost entirely disappeared in 32 cases (57.1%). Syringomyelia of 55 patients (98.21%) had improved or stabilized compare with syrinx growth in only one patient (1.79%). The details of postoperative syrinx size on MRI are presented in Figure 3.

DISCUSSION

Although many theories continue to improve our knowledge on CM I associated with SM, the exact pathogenesis of this disease is still unknown and no animal models successfully reproduced this disorder. A relatively narrow posterior fossa is widely considered as the leading cause for this malformation, but it cannot completely explain the etiology and mechanism (10, 11, 17, 18). Correspondingly, surgical techniques for the treatment of CM I with SM at present entertain many controversies. Various surgical approaches have been described in the literature and there are two main surgical options: 1) expansion of the posterior fossa volume by posterior fossa decompression (PFD) with or without duraplasty; 2) reduction of the syrinx cavity by shunt surgery (syringoperitoneal shunting, syringopleural shunt, syringosubarachnoidal shunting or syringostomy). The effect of these surgical options for CM I have been reported with varying success in the literature. A series of surgically treated CM I patients were overall summarized by *Nozar Aghakhani* (2). The data reveal that syrinx shunting seems to have a higher risk of worsening (20.7% versus 15.6%), lower rate of improvement and a higher probability of reoperation than PFD. However, there are several difficulties in accurately

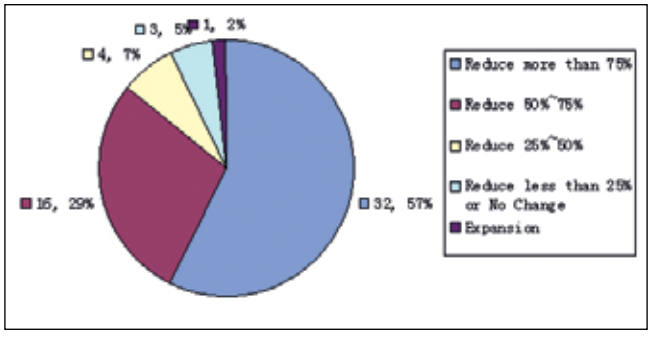

Figure 3: Effect of surgery on the cyst size at one year postoperative MRI follow-up.

Table III: Summary of the Duration of Preoperative Symptoms and Postoperative Results in 76 CM I Patients

Postoperative grade	Preoperative Status CM+SM (No. of patients)					Preoperative Status CM (No. of patients)			
	I	II	III	Total		I	II	III	Total
A	23	15	5	43		13	5	0	18
B	2	4	4	10		0	0	2	2
C	0	1	2	3		0	0	0	0
Total	25	20	11	56		13	5	2	20

A: *good result (cure and obvious improvement);* ***B:*** *stabilization of symptom;* ***C:*** *bad result with deteriorating symptoms. The duration of preoperative symptoms:* ***I:*** *less than two years;* ***II:*** *two to four years;* ***III:*** *more than four years. There is significant difference in the surgical effect among patients grouped by duration of preoperative symptoms (P=0.001).*

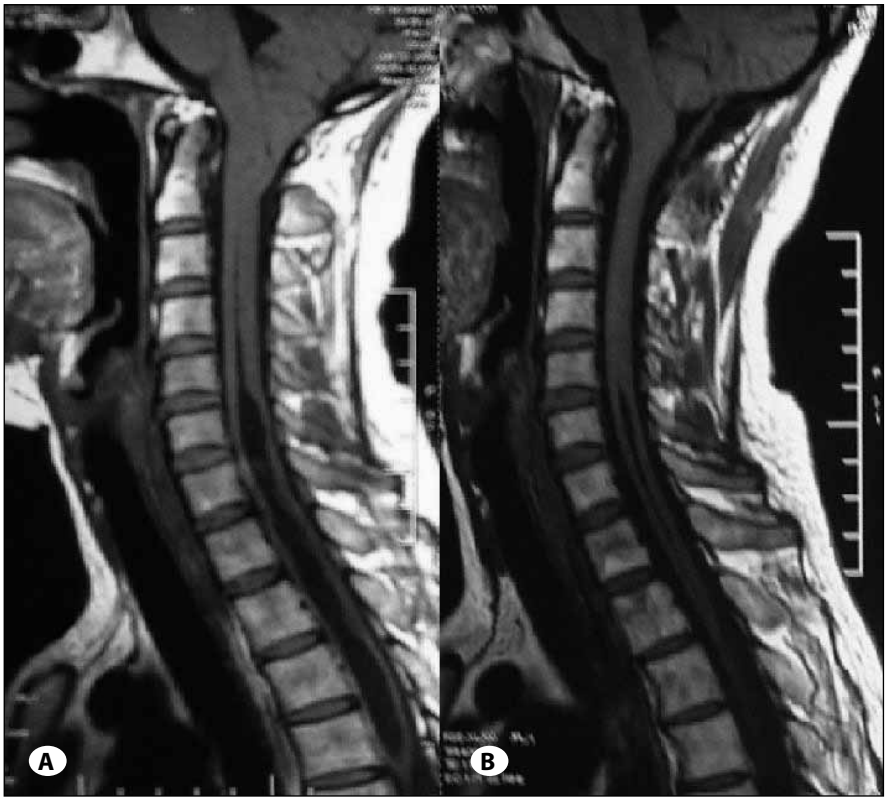

Figure 4: A) Preoperative MRI scan reveals tonsillar herniation extending below the foramen magnum more than 2.5cm in addition to obvious syringomyelia. **B)** Postoperative MRI scan reveals the absence of cerebellar tonsil with obvious remission of syringomyelia.

analyzing the effect of these different surgical strategies. The simple use of the same surgical effect may be improper because there are no clear or reproducible clinical scores to analyze postoperative outcome of CM I patients. Even in the same report, the authors performed different procedures, and their results cannot be easily distinguished. Considering that shunting surgery may cause direct spinal cord injury, PFD is primarily recommended to treat patients with CM I and SM in recent articles and shunt surgeries mainly act as supplementary methods to resolve SM in patients who have unfavorable results after PFD (1, 2, 6, 8, 21). In this series, one patient underwent syringo-subarachnoidal shunting surgery by the hemilaminectomy approach and a shunt tube was placed in the dorsolateral subarachnoid space. We routinely perform this type of shunting surgery for the treatment of postoperative CM I patients with exacerbated SM and symptoms following failure of PFD. The reason is as follows: 1. This procedure is easiest to perform compared with other shunting procedures. 2. Syringo-dorsolateral subarachnoid shunting may avoid dorsal column injury resulting from the posterior midline myelotomy and shunt tube insertion. 3. Pneumothorax and hydrothorax are not uncommon complications of the syringo-pleural shunt. The syringo-subarachnoid shunt can avoid these complications. 4. The hemilaminectomy approach is used to avoid postoperative spinal deformity.

Craniectomy is generally recognized as the mainstay for PFD. Compared with osseous decompression, removing the herniated cerebellar tonsil may be a way to improve the posterior fossa volume mismatch and to increase communication between the fourth ventricle and the spinal compartment. There is no direct neurological deficit that has been demonstrated as a result of tonsillar resection (13). However, the surgical effect of cerebellar tonsillectomy is questioned as tonsillar manipulation may lead to further arachnoidal adhesions later on at the foramen magnum with aggravation of syringomyelia and symptoms (19).

Our surgeries use a small incision to perform suboccipital decompression, cerebellar tonsilectomy and duraplasty. As the cerebellar tonsil will retract after subpial resection and coagulation, the neurosurgeon can perform subpial cerebellar tonsillectomy as long as the upper pole of the cerebellar tonsil is exposed. Even with cases where the cerebellar tonsil has reached the C2, the 3~4cm length incision is enough for operation (Figure 4A,B). In this series, the rare postoperative complications (1.32% CSF leakage and 3.95% intracranial infection) and the result of long-term follow-up (80.26% improved, 15.79% stabilized, and 3.95% worsened with 3.5 years of mean follow-up) are similar to those of the best prognosis report series of the literature summarized by *Nozar Aghakhani* (2). It demonstrates that removal of herniated cerebellar tonsils by subpial manipulation is safe and curative for CM I patients. At the same time, the limited incision is conducive to wound healing and reduces the risk of CSF leakage. However, it should be noted that cerebellar tonsillectomy must be under subpial manipulation, or it may do more harm than good regarding the relative complications.

To achieve successful surgical treatment with a favorable long-term postoperative outcome for CM I patients requires a thorough understanding of the underlying pathophysiology of CM I with syringomyelia and management of this malformation may be different in particular cases. However, there is still controversy on the necessity of cerebellar tonsillectomy. As some authors reported, tonsillectomy without craniectomy (13) can provide long-term cure for CM I patients, whether the hypertrophic and herniated cerebellar tonsil plays a role for limited volume of the posterior fossa and the symptoms is worthwhile to take into account in future research. Recently, some authors have reported endoscopic approaches for the management of Chiari I malformation (7). The use of an endoscope may be helpful to assist micro-invasive surgical strategy for CM I patients. By this approach, an endoscopic access to the cisterna magna might be feasible to visually resect herniated cerebellar tonsil without craniotomy for reducing postoperative complaints which will be our focus in further clinical research.

CONCLUSION

Cerebellar tonsillectomy with suboccipital decompression and duraplasty can provide long-time cure for most CM I cases. Early diagnosis and surgery are necessary to improve the surgical effect. A small incision about 3~4cm around the foramen magnum is sufficient to perform these operations and may be conducive to reduced postoperative complications.

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