Original article

The clinical efficacy study of treatment to Chiari malformation type I with syringomyelia under the minimally invasive surgery of resection of Submeningeal cerebellar Tonsillar Herniation and reconstruction of Cisterna magna

Yongli Loua, Jichao Yanga, Liuxiang Wanga, Xi Chen a, Xin Xina, Yong Liub,⇑

a Department of Neurospinal Surgery, The Zhengzhou Central Hospital Affiliated to Zhengzhou University, Zhengzhou 450000, China
b Department of Neurospinal Surgery, Yuquan Hospital of Tsinghua University, Beijing 10000, China

ABSTRACT

Purpose: Discuss the clinical efficacy of treatment to Chiari malformation type I with syringomyelia under the minimally invasive surgery of resection of Submeningeal Cerebellar Tonsillar Herniation and reconstruction of Cisterna magna.

Methods: 130 Chiari malformation type I with syringomyelia patients, divided into treatment group, literature group and control group, were collected to be treated under the monitoring of ultrasound in the surgery.

Results: 6 months after operation, the lesions were decreased or disappeared, the symptoms were relieved obviously. According to MRI and Mimics 17.0 software, the volumes of Cisterna magna increased distinctly (P < 0.001), the proportions of brain in foramen magnum region were decreased (P < 0.001). Assessed by CCOS scale and Tator methods, the improvement rates of treatment group were 97.7% and 94.6%, the literature group and control group were 82.2% and 77.8%, respectively.

Conclusion: The efficacy of Chiari malformation type I with syringomyelia under the minimally invasive surgery of resection of Submeningeal Cerebellar Tonsillar Herniation and reconstruction of Cisterna magna is remarkable, and the complications are fewer. This surgery emphasizes recovery of tonsil of cerebellum and reconstruction of Cisterna magna and the circulation path of cerebrospinal fluid, which is a safe and efficient treatment.

Keywords: Minimally invasive surgery
Chiari malformation type I
Syringomyelia
Reconstruction of Cisterna magna

0. Introduction

Syringomyelia is a chronic disease, resulted by many causes, and its annual incidence is 8.4/100 thousand (Wen et al., 2014; Ollivier, 1827). The main characteristic is deficiency of algesia and thalposis, mostly one side initially (Cheng et al., 2002; Zhu et al., 2007; Brugieres et al., 2000). Once the anterior commissure is involved, the algesia and thalposis will lack while the tactile sensation and bathestesia occur, which is sensory separation.

Dyskinesia presents amyotrophy and myo-asthenia with the deficiency of deep reflexes. The symptom of hyperkinesia may be caused by pressure or injury of downward motion path. Spastic paralysis can be below the plane of the lesion, occurring the increasing muscle tension and negative abdominal wall and positive Babinski, which has higher disability rates. What's more, it can also occur the dysfunction of the respiratory muscles, affecting the quality of patients seriously (Killeen et al., 2016; Liu et al., 2014). The database of syringomyelia shows about 75% patients suffer from this disease because of the Chiari malformation, and the type I, caused by heredity generally, is most common. There aren't standard statements in pathogenic factors and nosogenesis and treatment regimen. But, the surgery we recommend can eliminate the aetiological agent fundamentally and prevent from deterioration and makes satisfied outcomes (Loukas et al., 2011; Durham and Fjeldolenc, 2008; Milhorat et al., 2010).
1. Material and methods

1.1. Object

130 Chiari malformation type I with syringomyelia patients were elected by MRI, among, 50 males and 80 females, age was between 18 and 70 years old, the average was 42.5 ± 6.9 years old, and the average pathography was 65.6 months, 70 cases of headache (53.8%), 45 cases of numbness of limbs (34.6%), 32 cases of pain in neck and back (24.6%), 15 cases of disability (11.5%), 12 cases of dizziness (9.2%), 10 cases of ataxia (7.7%), 5 cases of sleep-apnea (3.8%), 3 cases of hydrocephalus (2.3%), 2 cases of dysphagia (1.5%) (see Table 1).

1.2. Imaging manifestations

All patients were checked with 3.0T MRI of head and neck and chest, measured the Chiari. And the criteria was its value is more than 5 mm than the foramen magnum’s, which showed all patients’ Cisterna magna disappeared without the atlantoaxial dislocation, but with syringomyelia in varying degrees.

1.3. Measuring the volume of Cisterna magna

60 normal persons and 65 patients elected randomly checked 3.0T MRI of head and neck plus T2-weight-3D-SPC test of posterior fossa. Set 324 as threshold value according to Mimics 17.0 software and the range of Cisterna magna in advance to get the almost real spatial three-dimensional model. Compare the volume of Cisterna magna and c and the proportions of brain in foramen magnum region with the normal’s.

1.4. Method

Patients were in left lateral position and fixed the head under general anesthesia. After location of 4 cm below occipital tuberosity or 1 cm beyond median hairline, cut the skin 2 cm vertically to expose the atlas occipital fascia and peel off then to expose the spinal dural mater adequately. Assess the range of motion and shape of Chiari and observe direction and speed of cerebrospinal fluid with ultrasound before cutting the spinal dural mater to presume the operative range. Then cut the spinal dural mater and arachnoidea, suspend to both sides to avoid blood vessels, and then incise pia mater in the middle of cerebellar tonsil. Gasify and remove Chiari with side hole aspirator and bipolar coagulation and ensure not to destroy the wholeness of pia mater. Separate the adhesion nerves and choroid tissue between cerebellar tonsil and spinal cord to expose lateral cistern of cerebellomedullary and cerebrospinal fluid flew from central foramen of fourth ventricle and spinal canal. After removing Chiari, the bottom of fourth ventricle and oblongata could be seen and ensure no active bleeding, then close the incision with bipolar coagulation. In order to expand subarachnoid space, sew up arachnoidea and its edge to rebuild Cisterna magna, then observe the direction and speed of cerebrospinal fluid and the shape of Cisterna magna with ultrasound. After sewing was completed, spinal dural mater can undulate up and down with respiration without effusion of cerebrospinal fluid.

1.5. Efficacy evaluation

After operation, antibiotics and nutriments were used according to the wound. 2 weeks later, assess the recovery of patients with CCOS scale and Tator methods, and the all would check MRI regularly for evaluation.

1.6. Statistical analysis

Statistical analyses were performed with SPSS version 21, and all dates examined by homogeneity of variance test met the conditions of t-test. Measurement data was indicated with average ± standard deviation, two arms were compared with t-test or paired t-test. Enumeration data was indicated with frequency and percentage, chi-square test and Fisher exact probability were used between arms. P < 0.05 was considered to be statistically significant.

2. Results

2.1. Contrast changes

Patients’ cerebellar tonsil almost turned normal after removing the Chiari as showed in picture 1 and 2. Compared to traditional cutting (about 8 cm), the surgical incision was much shorter (only 2–3 cm). Checking MRI 6 months after surgery, the reconstruction of cerebellomedullary cistern was very satisfying. (as indicated by arrow in picture 3).

2.2. Volume change of Cisterna magna

Compared with the volume of Cisterna magna after surgery between treatment and control group, the former’s volume was increased obviously (P < 0.001), but there was no change in the volume of foramen magnum smaller than normal’s, and the proportions of brain in foramen magnum region was markedly decreased (P < 0.001). As showed as MRI and Mimics 17.0 software (picture 4), the volume of Cisterna magna improved signally and was close to the normal’s (see Table 2).

2.3. Comparison of two evaluation

Compared to 167 control group patients, 127 treatment group patients were better and 3 worse, examined by Fisher exact probability, P < 0.05, which showed the efficacy in treatment group was

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### Table 1

| Symptom of Patients (Cases) | Frequency | Percentage |
|-----------------------------|-----------|------------|
| Numbness of limbs           | 45        | 34.6%      |
| Pain in neck and back       | 32        | 24.6%      |
| Disability                  | 15        | 11.5%      |
| Dizziness                   | 12        | 9.2%       |
| Ataxia                      | 10        | 7.7%       |
| Sleep-apnea                 | 5         | 3.8%       |
| Hydrocephalus               | 3         | 2.3%       |

**Note:** The table above represents the frequency and percentage of symptoms reported among the patients.
better, and the difference was statistically significant (Table 3, see Figs. 1–4).

Compared to 378 literature group patients, 123 treatment group patients were better and 3 stable, examined by Fisher exact probability, \( P < 0.05 \), which showed the efficacy in treatment group was better, and the difference was statistically significant (Table 4).

### 3. Discussion

Chiari malformation, a congenital malformation, has complex mechanism and clinical manifestation, its character is the Chiari enters the foramen magnum. The diagnosis depends on the under-part of Chiar is 5 mm beyond of the lower edge of foramen magnum (Urbizu et al., 2013; Strahle et al., 2011; Smith et al., 2013; Meadows et al., 2000). Syringomyelia, caused by many reasons, with Chiari malformation type I is most common, the incidence is 40.0–80.5% (Strahle et al., 2015). If there isn't apparent symptom, the frequency of Chiari malformation type I with syringomyelia is small. However, there isn't uniform theory of mechanism, most think the Chiari malformation is caused by congenital abnormality of opisthion (such as hypoevolutism, atlas occipital fusion, sag of the skull base, flat base of skul, etc.) resulted the volume of posterior fossa is small, the cranial nerves and nerve tissues are overcrowding, which leads to the obstruction of foramen magnum. While the cerebrospinal fluid circulation is abnormal and the pressure of cranial cavity and vertebral canal lose balance, the central substance of spinal cord occurs degeneration, liquefy and shrinkage, caused syringomyelia (Sahuquillo et al., 1994; Besachio et al., 2015; Houston et al., 2018).

### Table 2

| Group       | Values (cases) | Age (years old) | Volume of Cisterna magna (cm\(^3\)) | Volume of foramen magnum (cm\(^3\)) | Proportions of brain (%) |
|-------------|----------------|-----------------|------------------------------------|-------------------------------------|--------------------------|
| Normal      | 61             | 38.66 ± 6.89    | 9.70 ± 3.50                        | 95.01 ± 11.66                      | 89.85 ± 3.44             |
| Pre-operation | 65          | 45.39 ± 3.37    | 2.46 ± 1.89                        | 80.08 ± 14.87                      | 110.98 ± 7.04            |
| Post-operation | 65          | 45.39 ± 3.37    | 5.50 ± 1.99                        | 80.08 ± 14.87                      | 93.13 ± 2.01             |

### Table 3

Comparison of two surgeries (CCOS scale).

| Group                | Better (eleven-sixteen) | Worse (four-ten) | Total | Improvement rate |
|----------------------|-------------------------|-----------------|-------|------------------|
| Treatment group      | 127                      | 3               | 130   | 97.70%           |
| Control group        | 137                      | 30              | 167   | 82.20%           |

![Fig. 1. Comparison of MRI pre-and post-operation.](image1)

![Fig. 2. Comparison between pre- and post-operation.](image2)
Many methods can be selected to measure the volume of posterior fossa, for example, ultrasound, CT and MRI (Yanase et al., 2006; Hayashi, 1987). Mimics software based on medical imaging is used in clinical study extensively to process the objects, it can also set parameters automatically and has the function of modification (Tierney et al., 2002). Based on the modelling technique of positron emission tomography, the data after processed is used properly, the remaining will be deprecated as the useless. The three-dimensional modeling can present the shape of cisterna magna thoroughly in different angle and can perform virtual operation freely to measure relative data, which makes doctors have a more deep understanding of disease and make surgical plans in advance and the communication is more intuitive. The tools can measure the shape of samples and presume the side effect, which provides the support in our study.

PFD, also referred to cranial cervical decompression (CVD), is a traditional method used in 1932 firstly (Ellertsson et al., 1970). It usually contains the removing of partial cisterna and 1 or 2 posterior arch of neck, cerebellar tonsil remaining lower hernia and the opening of spinal dural mater, which has certain effect. But with the more deeper understanding of Chiari malformation type I and the development of technology, there are some deficiencies of this method, it’s reported 17% patients need secondary surgery, such as the surgical trauma is severe, recovery slower, meningeal irritation symptom because of the opening of spinal dural mater resulted the haemorrhage flowing into subarachnoid space, and if the haemorrhage is not absorbed well, fever and pain in head and neck will be occurred. In addition, there are also many side effect, such as the leak of cerebrospinal fluid, intracranial infection, cephalomeningitis, bulbar paralysis, the dysfunction of respiration and refractory of haematoma and so on. What is more, scar hyperplasia and muscle contracture compresses the posterior fossa influencing the effect, and bone decompression extensively results the cerebellum lacks support. Caused cerebellar ptosis, which triggers refractory and aggravation of symptoms and the range of decompression is not sure.

PFR, also referred to posterior fossa decompression with dura-plasty (PFDD), is an improved method aimed at the deficiencies of PFD. It was put forward by Sahuquillo (Williams, 1986) in 1994 when treating Chiari malformation type I with syringomyelia. Based on PFD, it keeps the fullness of subarachnoid space and rebuilt the cisterna magna used artificial or autologous dura mater to repair the cutting to make the cerebrospinal fluid flow smoothly. But, it does not open the arachnoidea and change the circulation of cerebrospinal fluid thoroughly. Vanacloca (Heiss et al., 1999) thinks this method is likely to occur the leak of cerebrospinal fluid, subcutaneous exudate, pseudomeningoecele and intracranial infection.

In recent years, most scholars treat the symptom and prevent the progression of chiari malformation type I with syringomyelia.
and its relative diseases by microtechnic (Shrestha et al., 2015; Aydin et al. 2005). But there are some standard required to the technology, the range examining is wide and the side effects are also a lot. So, detail tests should be examined before surgery, and ultrasound should be used in surgery to measure the volume of cisterna magna compared with the normal’s and to judge the degree of plugging to decrease the side effects possibly. It has made great progression in treatment of this disease with microtechnic. It’s popular shaving the head locally not fully before surgery and the cutting can be covered by hair. The microsurgery, processing the rhino scope automatically and separating the arachnoidea sharply by the 2.0–3.0 cm cutting (8–15 cm traditionally) to keep the fullness of subarachnoid space and repair the chiari, can meet the standard of anatomical reduction, stable of internal milieu and the improvement of structure-function to correct the malformation in foramen magnum. Assessed scientifically and reasonably, this acute and careful surgery can recover and restore the cerebellar tonsil, and eliminate deformity of posterior fossa, and resect dissemination and valve of obex, and remove the internal oppression, rebuilt the cisterna magna, and restore the circulation of cerebrospinal fluid, with the result that eliminating etiology, preventing the progression and relieving the symptom, which has satisfactory effects. In addition, due to the 2.0–3.0 cm cutting, the amount of bleeding is about 20–30 ml in the surgery, equal to the bleeding of laboratory examination, which is unbelievable if the amount of bleeding is about 20–30 ml in the surgery, equal to the bleeding of laboratory examination, which is unbelievable if the amount of bleeding is about 20–30 ml in the surgery, equal to the bleeding of laboratory examination, which is unbelievable in the amount of bleeding is about 20–30 ml in the surgery, equal to the bleeding of laboratory examination, which is unbelievable.

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Y. Lou et al. / Saudi Journal of Biological Sciences 26 (2019) 1927–1931

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