



Controversies related to pediatric Chiari I malformation

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Abstract

Purpose Foramen magnum decompression is a common surgical treatment for Chiari type 1 malformation (CM-1). There are several methods for the intervention to the dura matter such as dural plasty sectioning the dura, also removing the outer membrane of the dura without intradural manipulation. We compared retrospectively our cases with a presentation of illustrative cases. Finally, we add our comments to the worldwide questionnaire which was offered.

Methods Twenty-six patients (9 males and 17 females) who underwent surgery from 2010 to 2019 were included. The age distribution was 1 to 55 years old (mean: 16.4 ± 11.3). They were divided into two groups, group A which underwent outer membranectomy and group B which underwent dural plasty.

Results There were 15 cases in group A and 11 cases in group B, and tonsillectomy was performed in 5 cases out of the 11. Symptoms improved in 7 cases (47%) and 5 cases (45%), respectively. Improvement of the syrinx was seen in 14 cases (93%) and 10 cases (90.9%), respectively. No complications were seen in group A; however, one CSF leak and 2 pseudomeningoceles were seen in group B.

Conclusion The number of our cohort is not large enough to analyze statistics, but it seems that outer membranectomy is similar to dural plasty in terms of the results of surgery with a lower risk of complications for the treatment in non-complicated CM-1.

Keywords Chari type 1 malformation · Outermenbrenectomy · Surgical indication

Introduction

Since it is not completely understood, Chari type 1 malformation (CM-1) is a complicated disease. The unclear mechanism of the development of syringomyelia, cases with disassociation between the symptoms and images, and the existence of asymptomatic cases lead to the difficulty in treatment. In this article, we will report our cases of CM-1 which underwent foramen magnum decompression (FMD) with dural plasty and cases which underwent FMD + outer membranectomy. In this series, we excluded the so-called complex cases as Bollow et al. described [1].

Typical symptoms

According to Tubbs et al. [2], the most frequent symptom which is seen in CM-1 is headache (40%) followed by scoliosis (18%). The headache which the patients complain is occipital headache or posterior cervical pain. It occurs or worsens when they cough or sneeze. Also, symptoms caused by brain stem or cerebellar compression may be seen such as nystagmus, swallowing disturbance, apnea, and syncope. Lee et al. [3] described that in their series, a clinical presentation in all cases under 3 years old showed brain stem compression signs and 75% showed hydrocephalus, and craniosynostosis was seen in 25% in this group. There was syringomyelia in 80–100% of the other older group. Therefore, they suggested that there might be a possibility that cases under 3 years old may have a different pathology which mainly presents stem compression signs. In Japan, there is a screening of scoliosis a couple of times during elementary (6–12 years old) so CM-1 cases with scoliosis are frequently found through this screening. Also, other important symptoms are symptoms due to

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Fig. 1 Chiari type 1 malformation associate with syringomyelia extending caudally beyond the thoracic level

syringomyelia. The common symptoms from syringomyelia are dissociated sensory disturbance which is sometimes associated with muscle weakness and atrophy, spasticity, and loss of bowel and bladder control. According to these reports, our

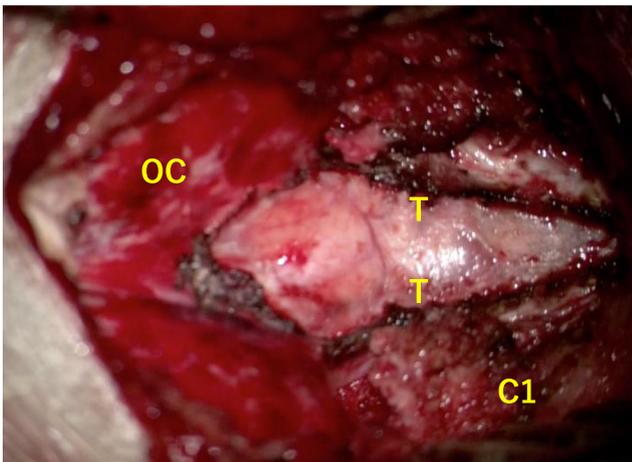


Fig. 2 Intraoperative view of FMD + outer membranectomy. Note the transparent tonsils (T) (which was pulsation after thinning the dura mater). OC: occipital bone, C1

institute's definition of "common" symptoms in CM-1 are listed below.

- a) Pressure like headache at the occipital area which worsens with coughing and sneezing
- b) Scoliosis
- c) Nystagmus, snoring/sleep apnea, and syncope which are considered symptoms due to brain stem compression
- d) Dissociated sensory disturbance which is sometimes associated with muscle weakness and atrophy which are suspected symptoms due to the syrinx
- e) Hydrocephalus caused by CSF flow disturbance adjacent to the craniocervical junction due to CM-1

Criteria for surgical indication

In our institute, if the patient is symptomatic, we consider the patient to have an indication for surgery. However, it is still in debate in asymptomatic cases. As we described above, there are a couple of cases that CM-1 is found in scoliosis patients. In these cases, the patients will be under close observation and if the scoliosis worsens, or before the treatment of scoliosis itself, the patient will undergo FMD. If the patient was asymptomatic at the initial visit but revealed a syrinx in the MR image, we will consider that there is a surgical indication, since a syrinx is a destructive lesion.

Type of surgery commonly used

The commonly accepted surgical treatment for CM-1 is foramen magnum decompression (FMD). There are several reports seen in the literature of adding dural plasty to this procedure. In our institute, in non-complicated CM-1, three surgical methods are applied.

1. FMD with dural plasty: the removing area of the occipital bone is decided by considering the width of the foramen magnum, and the posterior arch of the atlas is also removed. Dural excision is done in a Y shape by cutting downwards to the removed C1 posterior arch level. Artificial dura (Gortex[®]) is applied to create a pouch to extend the intradural volume. If the patients had symptoms which were due to compression of the brainstem, dural plasty is applied in our institute. If intraoperative findings such as severe arachnoid adhesion or the bulging of the tonsils are seen, an additional tonsillectomy was considered.
2. FMD without dural plasty: the removal of the occipital bone and C1 posterior arch is the same as FMD with dural plasty. To enlarge the intradural space, an outer

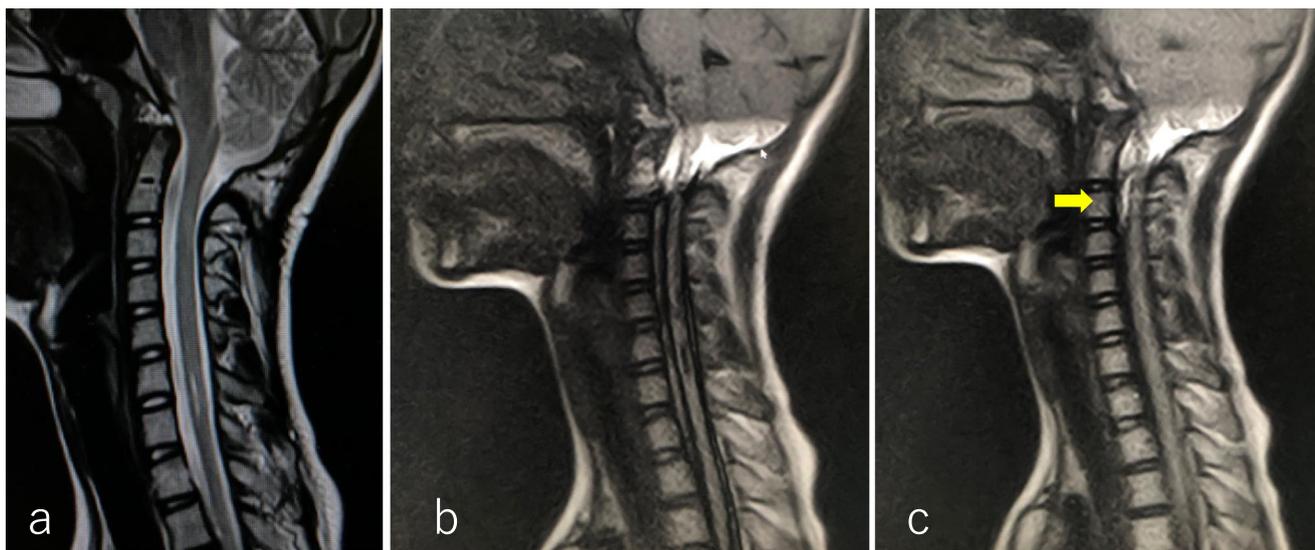


Fig. 3 a The decreased size of the syrinx. b Image of time-SLIP imaging. c A highlighted CSF by the movement of the highlighted CSF (yellow arrow)

membranectomy is conducted until the pulsating tonsils are visible through the transparent thinned dura.

3. If the patient suffers from symptoms due to hydrocephalus, endoscopic third ventriculostomy (ETV) will be selected as a first choice.

When are the results considered good?

Basically, if the patient’s symptoms improve, it is considered as a good outcome. As an objective measurement, if the syrinx

size decreases in the MR images after surgery, it could be defined as a good result. The symptoms may not improve because syrinx is a destructive lesion. Also, we think that an useful findings to predict the outcome during surgery is the pulsating tonsils. Additionally, an ultrasound device is used to see the pulsation of the CSF ventrally to the medulla. We have applied a sequence of MR imaging which is called the time-spatial labeling inversion pulse (time-SLIP) technique. This sequence uses CSF as an endogenous tracer which can detect CSF dynamics without contrast enhancement. It can detect a longer measurement range compared to cine MRI up to 1.5 to 6 s [4]. Finally, in hydrocephalus patients improvement of

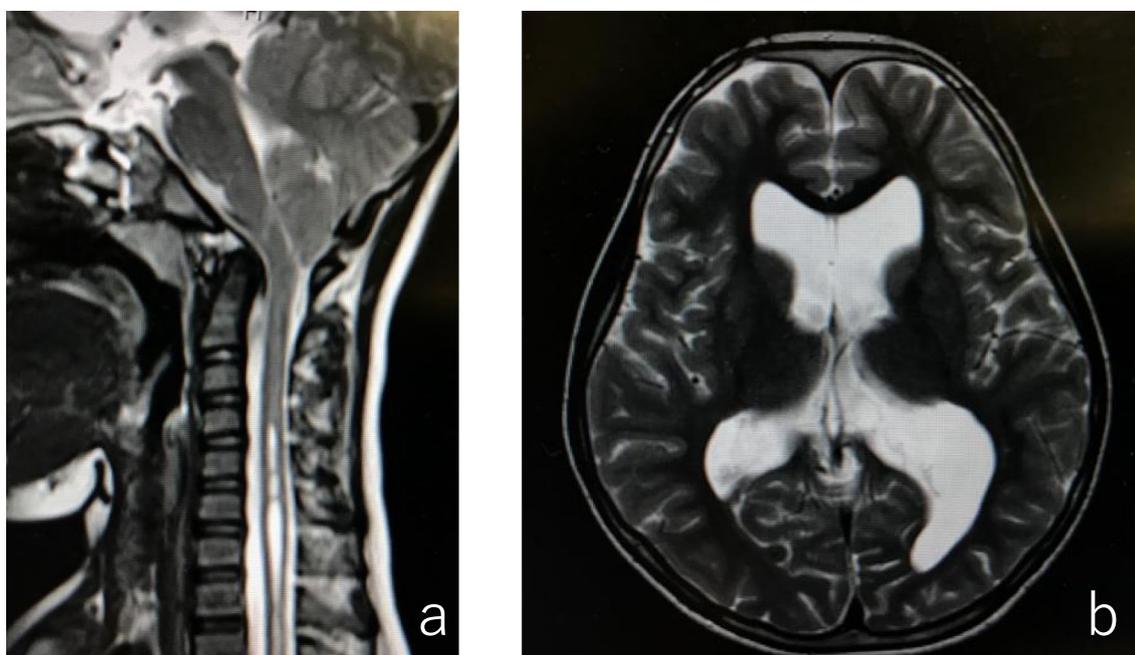


Fig. 4 a Chiari type 1 malformation associated with syringomyelia. b Hydrocephalus was also recognized by MR images of the brain

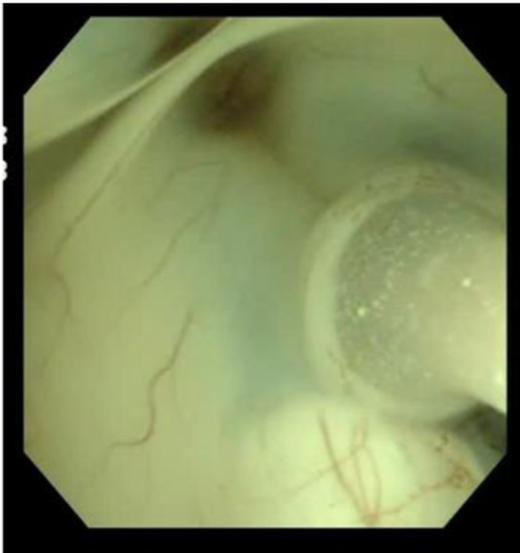
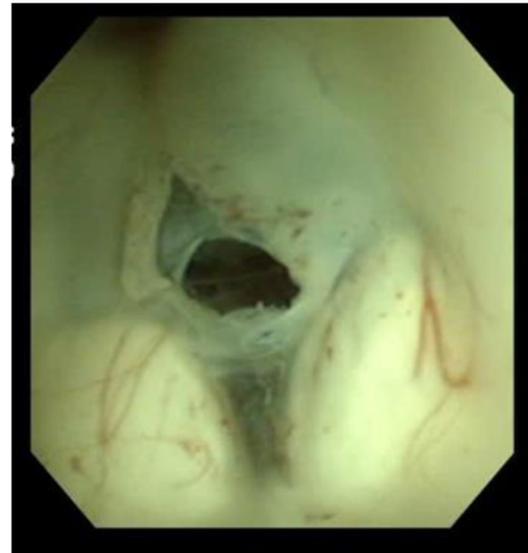


Fig. 5 Standard ETV was performed



hydrocephalus will be considered as a good outcome in CM-1 patients who underwent ETV.

Short presentation of the series (results and complications)

As described, FMD with dural plasty or FMD with outer membranectomy is applied in non-complicated CM-1 in our institute. Twenty-six patients (9 males and 17 females) underwent surgery from 2010 to 2019. The age distribution was 1 to 55 years old (mean 16.4 ± 11.3). The patients were divided into two groups: group A which underwent FMD + outer membranectomy and group B which underwent FMD + dural plasty. The determination of outcome was divided into 3 groups: improved, stable, and worsened groups. There were 15 cases in group A and 11 cases in group B, and in group B, tonsillectomy was performed in 5 cases out of the 11 cases. Symptoms improved in 7 cases (47%) in group A and 5 cases (45%) in group B. Improvement of the syrinx was seen in 14 cases (93%) of group A and 10 cases (90.9%) of group B. In group A, the symptoms were stable in 8 cases and there were no cases which showed worsening. In group B, on the contrary, the symptoms were stable in 5 cases and one case showed a deterioration with no change in the size of the syrinx. Therefore, s-s shunting was conducted. None of the complication cases were seen in group A; however, 3 complication (2 CSF leak and one pseudomeningocele) cases were seen in group B. The number of our cohort was not large enough to analyze the statistics, but it seems that outer membranectomy does not have a huge difference in terms of the results of surgery compared to cases with dural plasty. On the other hand, it seems that outer membranectomy has a lower risk of complications.

Illustrative cases

1. A 11-year-old girl who was diagnosed with scoliosis was referred to our institute. The MR image revealed CM-1 and syringomyelia (Fig. 1). There were no additional



Fig. 6 The post operative MR image showed a decrease of the syrinx. Also, the stoma is intact as the flow void was seen at the floor of the 3rd ventricle

anomalies in the craniocervical junction so she was defined as non-complicated CM-1. She did not have any neurological deficits however, there might be a risk of worsening of scoliosis and the existence of the syringomyelia could be a risk to develop symptoms. Also, this condition could be a risk of complications when she needs fixation of the scoliosis, so that she underwent FMD with outermembranectomy. C1 laminectomy and suboccipital craniectomy were performed followed by removal of the dural band and outer membranectomy. The pulsation of the tonsils was recognized. So that it was defined as sufficient decompression (Fig. 2). Postoperative MR images revealed the decreased size in the syrinx (Fig. 3a), and there was enough movement of the ventral CSF pulsation in the time-SLIP method compared to the preoperative images (Fig. 3b, c).

2. An 8-year-old girl with slight mental retardation was also diagnosed as scoliosis. She was referred to our institute, and the MR images revealed CM-1 and syringomyelia (Fig. 4a). Also, hydrocephalus was recognized (Fig. 4b). ETV was performed (Fig. 5). Postoperative images showed improvement in syringomyelia and hydrocephalus (Fig. 6).

Conclusion

The number of our cohort is not large enough to analyze the statistics, but it seems that outer membranectomy is similar to

dural plasty in terms of the results of surgery with a lower risk of complications for the treatment of non-complicated CM-1.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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