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Clinical Study

Surgical results of arachnoid-preserving posterior fossa decompression for Chiari I malformation with associated syringomyelia

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ABSTRACT

We analyzed the outcome of posterior fossa decompression accompanied by widening of the cisterna magna, without disturbing the arachnoid, in patients with Chiari I malformation (CMI) associated with syringomyelia. Twenty-five adult patients with CMI and syringomyelia, who underwent surgery between October 2000 and December 2008, were enrolled in this study. All patients underwent foramen magnum decompression with C1 decompression, with or without C2 decompression. Three surgeons performed a dura opening with duraplasty in 20 patients, and another surgeon excised the outer layer of the dura without duraplasty in five patients. Clinical and radiological assessments were performed preoperatively and during the follow-up period. After surgery, 20 (80%) patients achieved a significant improvement in their clinical symptoms. However, four patients (16%) achieved only a stable state, and one patient's symptoms worsened. Radiological analysis showed that 17 patients (68%) had a favorable result; that is, a total collapse, or a marked reduction, of the syrinx. Seven patients (28%) were stable in terms of syrinx size. However, the syrinx enlarged in one patient who had undergone excision of the outer dura. Twenty-four patients achieved a widened cisterna magna with ascent of the cerebellar tonsils into the posterior fossa and acquisition of a more rounded shape. Postoperative complications included a transient headache and vomiting in three patients and transient motor weakness in one patient. Two patients developed a superficial wound infection. This study shows that arachnoid-preserving posterior fossa decompression is a safe and effective treatment for patients with CMI with associated syringomyelia.

1. Introduction

Chiari I malformation (CMI) is a cerebellar tonsillar herniation through the foramen magnum associated with an abnormal posterior fossa constitution.[1–4] This condition may result in syringomyelia due to an obstruction of the flow of cerebrospinal fluid (CSF) at the craniocervical junction.[2,5,6] Although a number of surgical techniques have been described to restore the CSF circulation and decompress the neuraxis, no consensus has been reached as to the optimal technique.[5,6] Usually, surgery consists of a posterior decompressive craniectomy of the cervico-occipital junction associated with duraplasty, arachnoid opening and, sometimes, tonsillectomy. However, a number of surgical adjuvants to standard bony decompression remain controversial. Furthermore, some disadvantages, such as a high rate of CSF fistulas, aseptic meningitis, and long hospitalization periods, have been reported in patients who have undergone tonsillectomy and arachnoid dissection.[7–9]

To minimise the risks related to handling of the arachnoid membrane and tonsillectomy, we performed decompressive craniectomy and arachnoid-preserving duraplasty consecutively in adult patients with CMI and syringomyelia. This procedure resulted in decompression and shrinkage of the syringomyelia and ascension of the cerebellar tonsils. We report the functional results achieved by adult patients with symptomatic CMI and syringomyelia using arachnoid-preserving posterior fossa decompression and show its efficiency using radiography.

2. Materials and methods

2.1. Patient characteristics

Forty-three consecutive patients with CMI with associated syringomyelia underwent arachnoid-preserving foramen magnum decompression with excision of the outer dura or duraplasty at our institution between October 2000 and December 2008. Medical charts, radiographical features and outcomes determined by patient interviews were retrospectively assessed. Inclusion criteria were: (i) a diagnosis of CMI with syringomyelia; (ii) an age >18 years at the time of surgery; (iii) a minimum of two years of postoperative follow-up; and (iv) at least one preoperative and postoperative MRI. Patients with a tumor, a craniocervical bony

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anomaly, hydrocephalus, a history of meningitis, or history of previous posterior fossa decompression or shunt surgery were excluded. Twenty-five patients who met the inclusion criteria were enrolled in this study.

2.2. Surgical technique

Patients underwent surgery in the prone position with the head fixed in a Mayfield head holder. A midline linear skin incision was made from the inion to C2 or C3, depending on the extent of cerebellar herniation. The suboccipital region below the inferior nuchal line was carefully decompressed by removing the posterior lip of the foramen magnum. After laminectomy of C1 and C2 (if necessary), the dural surface of the craniocervical junction was examined under an operative microscope and bands of thickened tissue on the outer dura were carefully removed. A constricting band of thickened tissue on the outer surface of the dura was removed at the craniospinal level. Three surgeons (ESK, JSK, HJS) opened the dura and performed duraplasty (Supplementary video). Duraplasty was performed using an autologous periosteum patch or a Lyoplant (B. Braun, Melsungen AG, Melsungen, Germany). Care was taken to ensure that the underlying arachnoid was not compromised. Osteodural decompression was considered appropriate when we could observe, under a transparent arachnoid, good pulsation of the cerebellar tonsils and a free flow of CSF at the craniospinal level. Another surgeon (JIL) opened the outer layer of the dura, without duraplasty, to widen the cisterna magna.

2.3. Clinical and radiological assessment

Clinical and radiological assessments were performed preoperatively and during follow-up by two neurosurgeons not involved with the surgery. Preoperative assessments considered symptom duration, neurological features, and syrinx levels and diameters. Patients underwent a cerebrospinal MRI and a tonsilar herniation evaluation. After surgery, neurological findings and complications were assessed. Direct examinations were performed one, three, and six months after surgery, and subsequently at six-monthly intervals. Patients underwent craniospinal MRI three months after surgery, and this was repeated depending on the clinical evolution and MRI findings. MRI were used to determine the postoperative size of the syrinx, widened cisterna magna volumes in the midsagittal plane, and the size of the CSF space surrounding the medulla. Syrinx improvement was defined as a decrease in the maximal syrinx diameter on MRI.

3. Results

3.1. Clinical and radiological findings

The cohort was composed of 17 female and eight male patients (mean age = 36.6 years, range = 18–67 years). Follow-ups were conducted over 24 to 76 months, and the duration of symptoms ranged from two to 120 months. Sensory disturbances were the presenting symptom in 23 patients (92%); eight patients (32%) exhibited motor weakness, and eight patients (32%) had a suboccipital headache. Suboccipital headaches were exacerbated by valsalva maneuvers and sudden changes in posture. Lower cranial nerve deficits were observed in four patients (16%). One patient had a basilar impression and Klippel–Feil syndrome, which did not interfere with the CMI assessment. The cerebellar tonsils were positioned just below the foramen magnum in two patients, at the C1 level in six, and at the C2 level in three. Preoperative clinical and radiological features are summarized in Table 1.

3.2. Surgery and complications

Twenty patients (80%) underwent posterior cranial fossa decompression, C1 laminectomy, and duraplasty. An additional C2 laminectomy was required in five patients (21%) who had incomplete tonsillar decompression after C1 laminectomy. No severe adhesive arachnoiditis or subarachnoid scarring associated with CMI was observed. Accidental pinholing of the arachnoid membrane occurred during dural opening in three patients who underwent duraplasty. In addition, a large accidental arachnoidal rent occurred in one patient during a difficult opening of the dura mater. Postoperative complications included a transient headache and vomiting in three patients who underwent dural opening and transient motor weakness occurred in one patient. Two patients experienced a superficial wound infection, but responded to antibiotics. No complications occurred in the five patients who did not undergo dura opening.

3.3. Follow-up outcomes

After surgery, 20 of 25 patients (80%) showed significant improvement of their clinical symptoms. However, four patients (16%) achieved only a stabilized state of clinical symptoms and one patient's symptoms worsened. Improvements were observed in 18 (78.3%) of 23 patients with sensory disturbances, after surgery (Table 2). A suboccipital headache, when present, improved in eight (88.9%) of nine patients, motor weakness improved in four (44%) of nine patients and lower cranial nerve deficits in two of four patients (Fig. 1). Postoperative MRI showed that 17 of 25 patients (68%) achieved a favorable outcome; that is, syrinxes were totally collapsed, or syrinxes or segments were remarkably reduced in diameter. Syrinx size stabilized in seven patients (28%). Of the 20 patients who underwent duraplasty, 14 showed a decrease in syrinx volume and clinical improvement. Six patients showed symptomatic improvement without syringomyelia improvement. Of the five patients who did not undergo duraplasty, three had clinical improvement with a decrease in cavity size. One patient had symptomatic improvement with no syringomyelia improvement, and in one patient, who had no clinical improvement, the cavity size increased. A wider cisterna magna was detected in 24 patients and ascent of the cerebellar tonsils into the posterior fossa with acquisition of a more rounded shape occurred in 24 of 25 patients (Fig. 2).

Table 1
Demographic data of 25 patients who underwent extra-arachnoidal cranio-cervical decompression for Chiari I malformation associated with syringomyelia

	No. patients
Gender	
Male	8
Female	17
Mean age (years) ± SD (range)	36.6 ± 14.2 (18–67)
Clinical symptoms	
Sensory disturbance	23
Motor weakness	9
Neck pain	9
Cranial nerve symptoms	4
Syrinx level	
Cervical	7
Cervicothoracic	16
Entire cord	2
Type of operation	
FMD + DO	5
FMD + DO + DP	20

DO = opening of the outer layer of the dura, DP = duroplasty without arachnoid opening, FMD = foramen magnum decompression, SD = standard deviation.