Surgical Management of Hemifacial Spasm Associated with Chiari I Malformation: Analysis of 28 Cases

Jian Cheng[1], *Jinli Meng*[2], *Ding Lei*[1], *Xuhui Hui*[1], *Heng Zhang*[1]

■ OBJECTIVE: **Hemifacial spasm (HFS) associated with Chiari I malformation (CIM) is rare. This study aimed to analyze the frequency of HFS associated with CIM in our department and further to investigate the clinical characteristics, treatment strategies, and outcomes of these cases.**

■ METHODS: **Twenty-eight of 831 patients with HFS who fulfilled the criteria for CIM were analyzed retrospectively. In this series, microvascular decompression (MVD) was performed in 23 patients (82.1%). The remaining 5 patients (17.9%) with obvious symptoms attributable to CIM were treated only with foramen magnum decompression. The mean follow-up period was 41 ± 21.7 months.**

■ RESULTS: **The frequency of HFS associated with CIM was 3.4%. There were 19 women (67.9%) and 9 men (32.1%) with a mean age of 36.4 ± 7.5 years. The most common symptoms were headache, paraesthesias, and muscular weakness with the exception of typical HFS. Seventeen patients (73.9%) experienced immediate postoperative spasm relief, and 21 patients (91.3%) were spasm relief at discharge after MVD. However, 3 patients (14.3%) experienced delayed recurrence of HFS after successful MVD in the follow-up. After foramen magnum decompression, 3 of 5 patients experienced complete relief of the spasm, and 4 patients showed improvement in the CIM-related symptoms.**

■ CONCLUSIONS: **The results suggest that MVD can still be an effective treatment for HFS when it coexists with CIM. Furthermore, posterior fossa crowdedness may be a common risk factor for the 2 diseases, and foramen magnum decompression should be considered as the primary procedure in patients with HFS and symptomatic CIM.**

INTRODUCTION

The etiology of hemifacial spasm (HFS) generally has been accepted as the result of vascular compression of the facial nerve root exit zone (REZ).[1-3] Microvascular decompression (MVD) leads to the elimination of neurovascular conflict and successful relief from spasm, strongly supporting this theory, which is regarded widely as an effective method for curing the disease.[3,4] However, the specific pathophysiology underlying the development of neurovascular conflict is still unclear, and relevant studies suggest that it may be associated with posterior fossa crowdedness.[5,6]

Chiari I malformations (CIM) is a congenital disease characterized by caudal displacement of the cerebellar tonsils into the spinal canal.[7] It is suggested that an underdeveloped occipital bone, possibly due to underdevelopment of the para-axial mesoderm, causes overcrowding in the posterior cranial fossa, cerebrospinal fluid (CSF) flow disturbances at the craniovertebral junction, and herniation of the cerebellar tonsils.[8-10] Foramen magnum decompression is accepted widely as the surgical treatment of choice for symptomatic CIM.[11]

HFS associated with CIM rarely is reported in the literature.[12-15] Interestingly, decompression of the foramen magnum or shunting

Key words
- Chiari I malformation
- Foramen magnum decompression
- Hemifacial spasm
- Microvascular decompression
- Outcome
- Posterior fossa crowdedness

Abbreviations and Acronyms
CIM: Chiari I malformation
CSF: Cerebrospinal fluid
HFS: Hemifacial spasm
MRI: Magnetic resonance imaging
MVD: Microvascular decompression
REZ: Root exit zone
VPS: Ventriculoperitoneal shunt

From the [1]*Department of Neurosurgery, West China Hospital, Sichuan University, Chengdu; and* [2]*Department of Radiology, Hospital of Chengdu office of People's Government of Tibetan Autonomous Region (Hospital C.T.), Chengdu, China*

To whom correspondence should be addressed: Heng Zhang, M.D. [E-mail: cjbs2012@163.com]

Jian Cheng and Jinli Meng contributed equally to the study and should be considered as co–first authors.

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procedures for associated hydrocephalus have led to relief from symptoms of HFS.[12-14] These results suggest that there may be some underlying connections between CIM and the genesis of HFS. However, little has been published regarding this association. For this reason, we conducted a retrospective study to analyze the frequency of HFS associated to CIM in our department and further to investigate the clinical characteristics, treatment strategies, and clinical outcomes of these cases. In addition, we also reviewed the relevant literatures and discussed the mechanisms of causation of HFS associated with CIM and optimal treatment strategy in this situation.

METHODS

Patient Population

Between October 2008 and December 2016, a total of 831 patients with HFS underwent surgical treatment at the Neurosurgery Department of West China Hospital. In this series, patients who met the criteria for diagnosis of CIM were included in this study.[7,16] Patients with other facial dyskinesias, such as blepharospasm, facial tics, hemimasticatory spasm, myokymia, or psychogenic conditions were excluded. Data pertaining to clinical features, therapeutic management, and clinical outcomes were collected.

Imaging Evaluation

Magnetic resonance imaging (MRI) was performed in each patient preoperatively. This included a standard protocol of conventional axial T1-weighted, axial T2-weighted, and a special 3-dimensional sequence focused over the posterior fossa: 3D-T2-driven equilibrium (DRIVE) sequence, which aimed to exclude the space-occupying lesions and identify the possible neurovascular conflict at REZ. Twenty-eight patients who fulfilled brain MRI criteria for CIM were included in this study.[7,16] The diagnosis of CIM was based mainly on the anatomical demonstration of the cerebellar tonsils extending 5 mm or more below the foramen magnum.[7]

Surgical Technique

The surgical treatment of the patients with HFS and associated CIM was tailored to the individual. Patients with no significant symptoms attributable to the CIM were treated with MVD. The surgery was performed via a standard suboccipital retrosigmoid approach, which has been described in detail in our previous study.[5] In addition, for patients who presented with HFS and CIM-related symptoms simultaneously, foramen magnum decompression was performed first, which consisted of suboccipital craniectomy, C1 laminectomy, arachnoid dissection, and duraplasty. If the spasm was not cured after foramen magnum decompression, then MVD was recommended. Hydrocephalus was treated, if necessary, with ventriculoperitoneal shunt (VPS) placement.

Follow-Up and Outcome Assessment

All patients were followed at the neurosurgery outpatient department or by telephone. Detailed clinical outcome assessment was performed immediately after surgery, at discharge, and at 6- to 12-month follow-up intervals. The surgical outcome of HFS was categorized as success (spasm free) or failure (persistent spasm). Postoperative success was defined as complete spasm resolution with no residual twitching. The clinical outcome of CIM was defined as improvement, unchanged, or aggravated, as compared with the preoperative symptoms. Operative complications also were recorded immediately after surgery and at follow-up.

Statistical Analysis

SPSS software was used for statistical analyses (version 24.0; IBM Corp., Armonk, New York, USA). The mean was expressed ± standard deviation throughout. Descriptive statistics were used to summarize patient characteristics and clinical features. Kaplan-Meier analysis was performed to evaluate the long-term outcome of patients treated with MVD. Associations were considered statistically significant when $P < 0.05$.

RESULTS

Patient Characteristics

Twenty-eight of 831 patients with HFS fulfilled the criteria for CIM; the frequency of HFS associated with CIM in our sample was 3.4%. There were 19 women (67.9%) and 9 men (32.1%) with a mean age of 36.4 ± 7.5 years (range, 17–54 years). The left side was affected in 16 patients (57.1%) and the right in 12 patients (42.9%). The mean HFS duration before surgery was 43 ± 28 months (range 6–120 months) (Table 1).

As to clinical symptoms, with exception of typical HFS, there were 5 patients (17.9%) who showed CIM-related symptoms. Among them, 5 patients (17.9%) had headache that radiated to the neck and shoulders. Three patients (10.7%) showed various degrees of paraesthesias on the extremities. Two patients (7.1%) showed muscular weakness. Cervical syringomyelia associated with CIM was found in 3 patients (10.7%), and preoperative hydrocephalus was noted 1 patient (3.6%) (Table 1).

Surgical Characteristics

In this series, MVD was performed in 23 patients (82.1%). In all cases, the vessels compressing the facial nerve REZ were identified. The predominant offending vessel types were the anterior inferior cerebellar artery in 13 patients (56.5%), posterior inferior cerebellar artery in 7 patients (30.4%), vertebral artery in 2 patients (8.7%), and vein in 1 patients (4.3%). Five patients (17.9%) with obvious symptoms attributable to CIM were treated with foramen magnum decompression (Figure 1). In addition, VPS placement was performed in 1 patient who presented with HFS, CIM, and hydrocephalus simultaneously.

Clinical Outcomes

The mean follow-up period was 41 ± 21.7 months (range 6–84 months). In 23 patients treated with MVD, 73.9% of patients experienced immediate postoperative relief of spasm, 4 patients (17.4%) experienced delayed resolution within a mean time of 6.3 ± 2.5 days (range 3–9 days), and 91.3% had relief at discharge. However, 3 of 21 patients (14.3%) experienced delayed recurrence of HFS after successful MVD in the long-term follow-up. Although repeat MVD was recommended for the recurrent spasm, all 3 patients selected the treatment of botulinum toxin injections.