Hemifacial Spasm: A Neurosurgical Perspective

Hemifacial spasm (HFS) is characterized by tonic clonic contractions of the muscles innervated by the ipsilateral facial nerve. Compression of the facial nerve by an ectatic vessel is widely recognized as the most common underlying etiology. HFS needs to be differentiated from other causes of facial spasms, such as facial tic, ocular myokymia, and blepharospasm. To understand the overall craniofacial abnormalities and to perform the optimal surgical procedures for HFS, we are to review the prevalence, pathophysiology, differential diagnosis, details of each treatment modality, usefulness of brainstem auditory evoked potentials monitoring, debates on the facial EMG, clinical course, and complications from the literature published from 1995 to the present time.

KEY WORDS: Hemifacial spasm · Microvascular decompression · Craniofacial abnormalities.

INTRODUCTION

Hemifacial spasm (HFS) is characterized by unilateral, intermittent contractions of the muscles of facial expression, which typically begins in the orbicularis oculi and spreads to the other muscles of expression over several years. At early period, it usually affects the periorbital muscles unilaterally. With time, other ipsilateral facial muscles become affected and brief clonic switching may gradually lead to more sustained tonic contractions. There is considerable evidence that primary HFS is in almost all cases related to a vascular compression of the facial nerve at its Root Exit Zone (REZ) from brainstem, and that microvascular decompression (MVD) constitutes its curative treatment.

Here, in an attempt to understand the overall craniofacial abnormalities and to perform the optimal surgical procedures for HFS, published articles from 1995 to the present time were reviewed along with our accumulated experiences.

EPIDEMIOLOGY

Valid prevalence estimates of HFS are elusive because of underdiagnosis, misdiagnosis, and the absence of population-based data. To our knowledge, there have been few studies documenting the prevalence of hemifacial spasm. These studies demonstrated the total prevalence of HFS in range of 9.8 to 11 per 100,000 in the total population. Unfortunately, little is known about the accurate prevalence in Asia, although HFS is known to be more prevalent than trigeminal neuralgia in northeastern Asia.

PATHOPHYSIOLOGY

Since Dandy proposed first that cranial nerves by an ectatic vessel can cause clinical syndromes in 1934, Jannetta further explored and popularized MVD as an effective treatment for HFS and trigeminal neuralgia. He assumed that the mechanical effect of a pulsating blood vessel was the cause of the disease and that the compression must be at the root entry zone (REZ) of the cranial nerve to cause symptoms.

However, several authors suggested that the compression can occur at any point along the CNS segment of cranial nerve. The length of the CNS segment of the trigeminal nerve is longer than that of the facial nerve. Assuming that the CNS segment of cranial nerves is more vulnerable to microtrauma than the PNS segment, more people would be expected to have trigeminal neuralgia than hemifacial spasm. This study was inconsistent with Jannetta's
hypothesis that symptoms are caused by vascular compression of the REZ, the length of which is similar in all cranial nerves. De Ridder et al. supposed that the incidence of trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia is clearly related to the length of their CNS segments. Consequently, the authors hypothesized that the site for a vascular compression to become symptomatic should be at the CNS segment and not only at the REZ. As an evidence of this suggestion, Sindou et al. described that in his 579 patients with trigeminal neuralgia, the neurovascular compression was at the root entry/exit zone in only 52.3%, while compression was in the mid portion of the root in 54.3%. Although etiology of HFS is not well-known yet, a possible association between HFS and arterial hypertension has been suggested although not clearly evident. Three recent case-control studies suggested that primary HFS may be associated with arterial hypertension. Compression of the ventrolateral medulla (VLM) at the root entry zone of cranial nerves IX and X may induce hypertension. Because the rostral VLM is close to the origin of the facial nerve, vessel loops originating from the vertebral/basilar arteries or their branches might be responsible for multiple neurovascular compressions, inducing both HFS and hypertension. Alternatively, hypertension could be a risk factor for HFS because it may produce vascular anomalies resulting in compression of the seventh cranial nerve. Under this hypothesis, one would expect that hypertension would precede hemifacial spasm.

DIFFERENTIAL DIAGNOSIS

Craniofacial dyskinesias encompass a variety of abnormal spontaneous craniofacial movements that often appear similar in morphology. Physicians including neurosurgeons, neurologists, and other medical practitioners (including those involved in oriental medicine) should be aware of a variety of abnormal spontaneous craniofacial movements (Table I).

Blepharospasm

Blepharospasm is a bilateral condition characterized by focal, excessive involuntary closure of the eyelids generally caused by spasm of the orbicularis oculi muscles and it may sometimes lead to functional blindness. Despite having different pathophysiology, both HFS and blepharospasm produce involuntary eyelid closure due to contractions of the orbicularis oculi muscle. Blepharospasm is known to be due to hyperexcitability of brainstem interneurons, as a result of organic dysfunction of the basal ganglia. Patients may complain of photophobia, ocular irritation, or dry eyes. Hallett et al. demonstrated in their case-control study that blepharospasm was more associated with generalized anxiety disorder than hemifacial spasm.

<table>
<thead>
<tr>
<th>Table 1. Differential diagnosis of craniofacial dyskinesias</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hemifacial spasm</strong></td>
</tr>
<tr>
<td>Involvement Characteristics</td>
</tr>
<tr>
<td>Unilateral</td>
</tr>
<tr>
<td>Intermittent contractions of facial muscles</td>
</tr>
<tr>
<td>Typically begins in the orbicularis oculi</td>
</tr>
<tr>
<td>It may persist during sleep</td>
</tr>
<tr>
<td>On EUG, with later spread response (LSR) and variable synkinesis on blink reflex</td>
</tr>
<tr>
<td>Aggravation</td>
</tr>
</tbody>
</table>
Craniofacial tics

Tics are brief, repetitive, stereotyped, and involuntary movements or sounds. The most severe type of tic disorder is Gilles de la Tourette’s syndrome. It usually begins in the face, spreads caudally, and fluctuates in frequency, intensity, and distribution. Treatment includes benzodiazepines, calcium channel blockers, and dopamine depletors.

Ocular myokymia

Myokymia is characterized by undulating, vermicular, rippling, and wavelike movements across the muscle surface. Most cases are idiopathic and may constitute a generalized benign or essential myokymia syndrome. Almost all of idiopathic cases resolve spontaneously in a few days or weeks. Eyelid myokymia occurs in many people who are tired and fatigued. Unfortunately, this disease entity was not clearly defined and not widely accepted as a unique disease entity despite high incidence.

TREATMENT

As is already well known, widely-accepted treatment modalities of HFS include injections of Botulinum toxin as less invasive procedure and microvascular decompression (MVD) as the definitive treatment.

Botulinum toxin A (BTX-A)

Botulinum toxin type A (BTX-A) injections have been widely accepted as a safe and efficacious modality for the treatment of both blepharospasm and HFS, which has been approved by Food and Drug Administration for HFS in U.S., can be injected subcutaneously or intramuscularly into the involved facial muscles. At the early state of HFS, BTX-A injection is very attractive method because it is less invasive and easily approached by physicians. The onset of action is about 3 to 5 days and the mean duration of benefit is 3 to 6 months. Thus, patients require repetitive injections two to four times per year. Complications of BTX-A injection include ptosis, blurred vision, and diplopia that usually improve in days to weeks. Most common injection sites are pretarsal and preseptal portions. Cakmur et al. showed that pretarsal injections of BTX-A in patients with involuntary eyelid closure due to contractions of the orbicularis oculi muscle are associated with higher efficacy and lower frequency of complications than preseptal injections. Although BTX-A injection shows high success rate of treatment of HFS and blepharospasm, its substantial limitation is that it required repeat injection, which eventually lead to charge of high economic cost because of uncoveryage under current national health insurance policy.

Microvascular decompression (MVD)

Whereas Botulinum toxin injection does not provide long-term relief because of its temporary benefit, surgical treatment using MVD results in long-term relief of the symptoms in the majority of patients. In our series, all of the surgical procedures were performed via a lateral retrosigmoid suboccipital approach. The patient is placed in a lateral park bench position with the head rotated approximately 10 degrees away from the affected site and the vertex dropped 15 degrees toward the floor. A 20 × 25 mm sized craniotomy is performed with extension to the flat occipital squama inferiorly and the sigmoid sinus laterally, below the posterior end of the digastric notch on the mastoid process.

Opening

The operation technique for MVD is variable according to the surgeon’s preference. For example, some authors favor supine position and others lateral decubitus position (it is so called “park-bench position”). In our series, all of the surgical procedures were performed via a lateral retrosigmoid suboccipital approach. The patient is placed in a lateral park bench position with the head rotated approximately 10 degrees away from the affected site and the vertex dropped 15 degrees toward the floor. A 20 × 25 mm sized craniotomy is performed with extension to the flat occipital squama inferiorly and the sigmoid sinus laterally, below the posterior end of the digastric notch on the mastoid process.

Intradural procedure

After the cisterna magna or lateral cerebellomedullary cistern is opened, the cerebellum gives way so that the VIIth to VIIIth cranial nerve complex can be approached with minimal use of brain retractors. Some drainage of

---

Fig. 1. Variable patterns on microanatomical findings of neurovascular conflicts. (Park et al. in processing). (A: thick arachnoid bands, B: branching vessels, C: sandwich vessels, D: small perforating vessels.)
CSF is necessary to minimize potential injury by placing the retractor on the cerebellum. After CSF is drained, a tapered retractor blade is placed over the previously placed rubber dam and cottonoid. After careful dissection of the arachnoid membrane and gentle retraction of the floculus, the root exit zone (REZ) of the facial nerve can be visualized. A good access from below is of most importance, because most conflicts are located ventro-caudally to the facial exit zone at the brainstem[61]. Janetta et al. and Sindou et al. indicated that reaching the facial nerve along an infra-flocular route was important for two reasons: 1) Neurovascular conflicts are usually located ventro-caudally at the REZ, 2) A lateral-to-medial retraction of the cerebellar hemisphere would exert stretching of the VIIIth nerve and lead to hearing loss[62,63].

To reduce the cerebellar retraction, according to Nakanishi’s approach, the operator’s head and the patient’s head are at the same height, and the operator can look at the surgical field through the microscope in a horizontal direction[67]. The compressing vessel, so called offender, can be identified near to the REZ. Several pieces of Teflon sponge are placed between the compressing vessel and the REZ.

Microanatomical observations
In our accumulated experience of about 900 cases, we found that the patterns of neurovascular conflicts by offending vessels are so variable in each case. As anatomical variation of neurovascular conflicts could be observed in trigeminal neuralgia[67], we found that a variety of anatomical compressive patterns contributed to the neurovascular conflicts. In some instances, focal thick arachnoid band was often a major contributing factor to compression. In others, two or even three compressing vessels were often observed with a type of sandwich, branching vessels, or multiple perforating vessels (unpublished data, Fig. 1). These findings were already mentioned by Sindou et al.[67] and it could be the source of insufficient decompressive surgery and consequently surgical failure[69]. However, no statistical significance was shown between variable neurovascular conflicts and clinical outcome.

Closure
After the neurovascular decompression, the dura mater can be closed in the following fashion which was newly designed[67]. Four to five interrupted stitches are made at intervals of 6-7 mm. Several pieces of muscle that have been obtained from the adjacent muscles can be interposed into the space between the stitches. The muscle piece should be small enough to enter the slit but large enough to fill the defect between the stitches[77]. In cases where the mastoid cavity is opened, bone wax should be applied to completely seal the opened surface of the cavity. Cranioplasty is performed using polymethyl methacrylate (PMMA) bone cement according to the surgeon’s preference. No fixation device is necessary because of the bulky muscle groups placed over the lesion. Irrigation with droplets of papaverine solution (10% concentration) is used in all cases as routine during surgical dissection of the neurovascular conflict and before closing of the dura mater.

BRAINSTEM AUDITORY EVOKED POTENTIALS (BAEP) MONITORING; ITS IMPORTANCE FOR HEARING PRESERVATION

Microsurgical maneuvers involved in relieving the neurovascular conflict may endanger the cranial nerve VIII. The use of intraoperative monitoring of brainstem auditory evoked potentials (BAEPs) is generally acknowledged to reduce the risk of hearing impairment. It can be estimated that Peaks I and II are generated in the distal and proximal parts of the cochlear nerve, respectively, and Peaks III to V along the brainstem auditory pathways from the cochlear nucleus up to the inferior colliculus. Damage to the VIIIth nerve results in a delay in latency and reduction in amplitude of Peak V. When this delay becomes significant, the neurophysiologist must inform the surgeon to discontinue the procedure until the cause is identified, and the dangerous maneuvers corrected. The main critical situations are the following: stretching of the VIIIth nerve when retracting the cerebellum; manipulation of the labyrinthine artery and/or the antero-inferior cerebellar artery; direct trauma to the nerve by instruments or a nearby coagulation; and at the end of surgery, neo-compression of the nerve by the prosthesis interposed between the conflicting vessel and the VIIIth-VIIIth nerve complex[55,56]. For most clinical neurophysiologists, latency of Peak V is considered the best electrophysiological indicator for signaling cochlear nerve damage by operative manipulations[75,160]. Grundy et al.[80] used the value of 1.5 milliseconds delay in latency of Peak V to warn the surgeon. Hatayama and Möller[46] paired latency delay of Peak V greater than 1 millisecond with amplitude modifications of this wave. Polo et al. demonstrated that latency of Peak V was the most frequently observed and the most significant phenomenon, especially during cerebellar retraction and the decompression step of the microvascular decompression procedure[61]. The main practical contribution of this study consists in neurophysiological parameters identified by the authors as “warning values” to avoid an irreversible Injury to the hearing function. Conversely, in our series, decrease of amplitude of Peak V wave was independently predicting value for hearing loss during MVD rather than the delayed latency of Peak V (unpublished). In clinical

358
practice, we are using these amplitude changes of Peak V wave as the treatment guideline. In conclusion, we strongly believe that close collaboration between neurosurgeon and neurophysiologist is very important to guarantee the safety of MVD.

**INTRA-OPERATIVE MONITORING OF THE FACIAL EMG RESPONSES**

In the 1980s, Møller and Jannetta showed that lateral spread responses (LSR) could be recorded from one muscle innervated by the superior branch of the facial nerve when the inferior branch was stimulated or conversely. These phenomena were presumed to be related to clefted transmission at the lesion site alone or in combination with motor nucleus hyperactivity. They advocated routine intraoperative EMG recording of LSR during MVD, to ensure that adequate decompression was achieved. Mooij et al. demonstrated that a guiding role of Intraoperative monitoring was apparent in 33.8% of patients, and a confirming role was demonstrated in 52.7% of patients, which showed that facial EMG can be useful as an aid in the “learning curve” as well as in guiding even the experienced neurosurgeon to obtain the best results. However, Hatem and Sindou et al. documented that the persistence of LSR did not necessarily indicate postoperative residual spasm. In their study of 30 cases, the authors suggested that the practical value of their intraoperative disappearance as a test of an effective decompression remains controversial. Based on our series of 300 cases, facial EMG monitoring of the LSR was proved to be useful in predicting the outcomes and might be an effective assistant tool to accomplish complete decompression if exploration is incomplete. Patients with persistent LSR after MVD showed poor one-year outcome, as compared with patients with absence of LSR after MVD. In the future, a well-organized and controlled study will be required for the evaluation of efficacy of LSR on the facial EMG monitoring.

**CLINICAL OUTCOME: IMMEDIATE OR GRADUAL IMPROVEMENT**

MVD of the facial nerve for idiopathic HFS has a long-term success rate ranging from 83 to 97%\(^{4,12,18,26,32,39}\). On the statistical analysis, no prognostic factor such as gender, age, side, and nature of neurovascular conflict showed any significant influence on outcome\(^{39}\). In our series of 455 cases, cure rate after one year was established at 95.7%.

It is of note that resolution of HFS is often gradual, whereas dramatic resolution is expected by patients and surgeons in most of cases\(^{3,27-39}\). Understanding of precise course of postoperative HFS is very crucial to make decision when the second reoperation should be done\(^{39}\). Ishikawa et al. observed that approximately one-half of their patients (50.3%) who underwent MVD for HFS continued to experience the condition to some degrees after the operation. Goto et al. demonstrated that the longer the symptom duration of HFS, the longer the time interval until complete resolution\(^{19}\). The immediate resolution of HFS could result from the disappearance of the spontaneous or ectopic excitation by the pulsatile compressive force of the offending vessel\(^{130}\). Saito et al. interpreted that delayed resolution could be attributed to the complete regeneration of the micro-injury of the facial nerve or the gradual stabilization of the facial motor nucleus\(^{30}\). According to their suggestion, reappearance of HFS within 4 days after microvascular decompression (MVD) does not necessarily suggest a failed procedure, and patients should be observed for at least a few months before another procedure is considered. On the analysis of our 236 series during recent two years, we found that 76 of a total patients (32.2%) who were eventually cured had residual symptoms for a variable period of time (from several months to a year), while 144 patients (61%) improved immediately after MVD (unpublished). Based on these results, we believe that we could predict the result after at least postoperative 3 months of MVD and the postoperative one-year is the optimal time to judge the final outcome.

For the evaluation of success rate of MVD, Chang et al. investigated the role of postoperative 3D-TOF MRA in predicting clinical outcomes and complications after MVD treatment of HFS\(^{35}\). Their study revealed that postoperative 3D-TOF MRA for patients with HFS might provide surgeons with anatomic information regarding neurovascular changes of the facial nerve REZ, although its correlation with clinical outcome was not proven yet.

**COMPLICATIONS OF MVD**

In the numerous reported series\(^{4,11,30}\), deafness accounted in a range of 1.6 to 2.6% and moderate hearing loss 0.6 to 0.7%. Permanent facial weakness was seen 3.4 to 4.8% following operations. Among other complications, CSF leakage affected 2.7% of the patients. In our series, hearing loss after MVD was in 1.8% of patients and rate of facial palsy was 0.8%. On an account of above mentioned muscle-plugging method\(^{39}\), CSF leakage rate was only 2 cases (0.2%) in 678 patients. Other than acute facial weakness, postoperative delayed facial palsy was seen during the follow-up period. Delayed facial palsy can be found in 2.8-8.3% of patients following vestibular schwannoma surgery. In 21 of 410 patients (5.4%), we could find the delayed facial palsy

359
at the postoperative 7-23 days (average 12.1 days). Our study revealed that the incidence of delayed facial palsy after MVD was not low as was previously reported in the literature but almost all patients achieved complete recovery without any further special treatment.

REOPERATION OF MVD

With recent advances in the diagnostic technology using high resolution MRI, offending vessels leading to neurovascular conflicts can be easily detected. Unlike trigeminal neuralgia, for which many medical and surgical options besides a second MVD are available if pain recurs, for patients with recurrent hemifacial spasm, botulinum toxin is the only other treatment option. Accordingly, Park et al. documented that repeated MVD could be applied for residual or recurrent HFS. However, time to re-operate MVD for recurrent disease should be carefully determined. Because spasm is often gradually improved after MVD (e.g. several months to even a few years), Sindou et al. did not recommend early re-operation in patients with initial failure or early poor results until one year at least has passed. Based on our limited experience of recurrent HFS, careful decision making on reoperation should be appreciated, considering high risk of morbidity on second MVD.

CONCLUSION

By Tan et al. in 2004, an interesting finding was reported that among 203 family physicians who participated in a “video” test on HFS, only 9.4% were able to diagnose HFS and 46.3% of them did not know how to manage the condition. It implies that specialists need to play a greater role in implementing educational programs to increase the awareness of the common movement disorders in the primary care setting. This will reduce misdiagnosis and inappropriate treatment for disorders such as HFS, which can be effectively treated.

Recently, Lunsford et al. mentioned that many patients who are eligible for MVD have a tendency of favoring a less invasive procedure for trigeminal neuralgia. This current trend toward less invasive technique may be similar in the treatment of HFS, because less invasive technique such as BTX-A injection clearly has less impact on their quality of life and allows rapid return to work. Accordingly, we should be aware of this trend and overcome this limitation of invasive technique such as microsurgical procedures. By experts’ hands under the guidance of intraoperative electrophysiological monitoring of BAEP, MVD procedure can be done with minimal morbidity and long-benefit. At the same time, more enthusiastic efforts and ongoing endeavor should be made to improve surgical technique toward the best outcome.

References


360


41. Lang J, Reiter U: Infratentorial length of the brain path and nerve tracts of the 1 to 4 cranial nerves. Neurochirurgia (Stuttgart) 27: 125-128, 1984

42. Lang J, Reiter U: Infratentorial length of the trigeminal nerve. Neurochirurgia (Stuttgart) 27: 159-161, 1984


69. Sindou MP: Microvascular decompression for primary hemifacial spasm. Importance of intraoperative neurophysiological monitoring.
Acta Neurochir (Wien) 147: 1019-1026; discussion 1026, 2005