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CLINICAL ARTICLE

Surgical treatment of pediatric hemifacial spasm patients

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Abstract
Purpose To study the clinical features and outcomes of pediatric primary hemifacial spasm patients who underwent microvascular decompression procedures.
Methods In this retrospective study, five pediatric (<18 years old) primary hemifacial spasm patients underwent microvascular decompression. After surgery, resolution of spasms and surgical complications were observed. Their social adaptability was evaluated using a social adaptation scale, which was designed specially for Chinese middle-school students.
Results Four typical hemifacial spasm patients had immediate excellent or good relief. However, the microvascular decompression procedure did not help the atypical patient much (50% relief of spasm). The score of social adaptation of the pediatric hemifacial spasm patients was 111.6±8.2. Compared with the ordinary healthy Chinese middle-school students, whose score is 170.8±25.4, the patients experienced great difficulty in social adaptation (P<0.01). After surgery, the scores of two patients increased to a normal level; however, the other three patients remained unchanged.
Conclusions Microvascular decompression is effective and safe to typical primary hemifacial spasm patients younger than 18 years old. Hemifacial spasm is harmful to the children's social adaptation. However, only some of the patients recovered to the normal social adaptation level even when the spasms were cured.

Keywords Pediatric · Hemifacial spasm · Microvascular decompression · Social adaptation

Introduction

Hemifacial spasm (HFS) is a neuromuscular disorder characterized by frequent, involuntary facial muscle contractions. Most patients complain of difficulty in their social lives caused by the involuntary spasms, which are worsened under stressful situations. It is widely recognized [1, 2, 11] that the cause of HFS is neurovascular compression at the root exit zone (REZ) of the facial nerve. HFS generally affects middle-aged and elderly people, and its occurrence in children is extremely rare [3]. In the elderly patients, the offending arteries are usually elongated, redundant and atherosclerotic, resulting from the aging process and hypertension. In children, such age-related changes are absent; therefore, this might account for the low incidence of HFS in children[3, 10].

In some reports in the literature, young-onset HFS (age of onset≤30 or 25 years) has an estimated prevalence rate of 0.9-6.5% [10, 17]. Some explanations have been proposed for the etiology of early development of HFS in youth. Levy et al. [12] indicated venous offending vessels at the REZ might be responsible. Kobata et al. [10] reported

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thickening arachnoid surrounding the offending vessels may cause HFS in youth by trapping and encasing the artery to compress the root exit zone. Besides, some diseases [4–7] may cause symptomatic HFS, such as anatomic variations, cerebellopontine angle arachnoid cyst, Chiari type I malformation, venous sinus thrombosis.

In these reports, the "young-onset" HFS was defined as cases in which the patient was less than 25 or 30 years old at the time of onset and less than 30 years old at time of surgical treatment [3, 10, 17]. Milani et al. [13] reported on a 10-year-old with HFS, and found arterial compression at the REZ of the facial nerve in the microvascular decompression (MVD) surgery. Singer et al. [16] reported a "childhood-onset" HFS patient (12 years old) who was treated successfully with periorbital botulinum toxin injections. To our knowledge, there are very few articles that specifically discuss the clinical features of pediatric primary HS patients (<18 years old) who underwent MVD procedures. Here we report our experience of treating five pediatric HFS patients in recent years.

Patients and methods

Patient population

Between July 2008 and July 2010, 1,304 patients underwent MVD for HFS at our institution (Table 1). Among them, five patients (0.38%) were under 18 years old at the time of surgery (mean 14.8, range 9-18), and the onset of HFS ranged from 5 to 14 years old (mean 9.8). The mean duration of the symptom was 5 years (range 2-11 years). Two boys and three girls were included in this study, with right-side symptoms in three of five (60%). Four patients were typical HFS with the initial site of symptom on the lower eyelid, and gradually extending over the entire ipsilateral hemi-face. Neurological examination of these cases revealed intermittent contractures of the orbicularis oculi and the orbicularis oris muscle. No other neurological sign was found. The other patient (case no. 5) was atypical, beginning with a customary wink on both sides, and 1 year later facial spasm emerging which mainly affected the left angulus oris. All these five patients had tried acupuncture and medicine (carbamazepine, clonazepam and so on) several times without any therapeutic effect. One patient (case no. 1) had received botulinum toxin therapy twice before admission, but it failed. The other four cases had not received botulinum toxin injection. There was no positive family medical history. Their birth had been uneventful. Preoperative magnetic resonance image (MRI) was performed in all these patients to exclude anatomical mass lesions, and three-dimensional time of flight magnetic resonance angiography (3D-TOF MRA) was performed to help the surgeons analyze the anatomic relationship between the facial nerve and arteries.

Operative procedures

MVD was performed in the same way as for adults, under general anesthesia with the patient in the contralateral decubitus position, via a lateral retrosigmoid suboccipital approach. A straight incision is made behind the hairline. Craniectomy is made below the transverse sinus and medial to the sigmoid sinus, to expose just the borders of both sinuses. Afterwards, the dura mater is opened. The cisterna magna is opened to drain cerebrospinal fluid (CSF), to provide enough space for gentle cerebellar retraction. After adequate CSF drainage, the cerebellum is gently elevated, and the caudal cranial nerves (IX-XI) are first identified. The arachnoid mater over the rootlets of cranial nerves IX–XI is sharply dissected, and then the arachnoid over the vestibulocochlear nerve and the facial nerve. The entire tract of the facial nerve was thoroughly explored, following the way as Campos-Benitez and Kaufmann reported [2], so as to avoid missing multiple vessel compressions. Following identification of the offending vessel, Teflon felt was placed between the vessel and the brainstem so as to transpose the course of the offending vessel. Sometimes the transposition is impossible because the offending vessel has some perforators running directly into the brain stem, and

Table 1 Information of five pediatric HFS patients underwent MVD

ID	Age of onset (years)	Age of surgery (years)	Sex	Vessels/compression site	Spasm relief	Operative complications	Social adaptation before/after MVD
1	12	14	M	AICA/REZ	Excellent/ immediate	None	114/181
2	5	9	M	AICA+vein/REZ+CP	Good/immediate	None	102/105
3	14	16	F	AICA/REZ	Good/immediate	None	108/172
4	7	18	F	AICA/AS *thickened arachnoid	Excellent/immediate	None	110/118
5	11	17	F	AICA/REZ+CP	Fair/immediate	None	124/121

AICA anterior inferior cerebellar artery, *MVD* microvascular decompression, *HFS* hemifacial spasm, *REZ* root exit zone, *CP* cisternal portion, *AS* attached segment

then thin pieces of Teflon felt is interposed between the vessel and the brainstem or the facial nerve.

Efficiency evaluation and follow-up

We evaluated the therapeutic efficiency of MVD procedures using the grades of Park et al. [15], as listed below: (1) "excellent," if an HFS was absent; (2) "good," if the HFS was more than 90% resolved; (3) "fair," if the HFS was more than 50% resolved; (4) "poor," if the HFS was less than 50% resolved; (5) "failure," for all remaining results, i.e., no resolution or even deterioration. The "immediate" result was assigned to the complete abolition of spasm at the next day of operation. The remainding cases were defined as "delayed." We also recorded any operative complications. After discharge, postoperative follow-up was done by telephone interview. The mean duration of the postoperative follow-up period was 16.8 months, ranging from 3 to 27 months.

Evaluation of social adaptability

Since the psycho-social life of HFS patients is usually affected by the involuntary spasms [14, 18], and the youngsters are very self-conscious and sensitive in public situations [14], we supposed that surgical treatment of HFS might help these pediatric patients to restore their social adaptability. The social adaptability of these five patients was evaluated using a social adaptation scale [9], which was designed specially for Chinese middle-school students. This scale covers six aspects of the youngsters' social lives: self-image and self-organization, adaptation to study, tolerance of frustration, relationship with parents and the family, relationships in school, and relationships outside of school and family. The total score of this scale is 236 points, and the least is 59 points. The original Chinese text can be accessed online (http://acad.cnki.net/kns55/detail/detail.aspx?dbcode=CMFD&QueryID=0&CurRec=1&dbname=CMFD0911&filename=2008189511.nh). And the English version of this inventory is provided in the complementary material.

Results

Preoperative MRI revealed that in each case there definitely was an artery in close relation to the facial nerve (Fig.1). The offending vessels were not elongated or shifted. In all these cases, there was no obvious elongated, redundant vertebral artery, which is very common in adult HFS patients. According to MRI, there was no intracranial tumor or Chiari's malformation.

Intraoperatively, all patients were found to haave definite offending vessels (Table 1, Fig. 2). The anterior inferior cerebellar artery (AICA) turned out to be the culprit vessel in all patients. In four cases, the AICA was the only offending vessel. The other patient (case no. 2) had an additional small vein in the REZ of the facial nerve. A thickening of the arachnoid was seen around the facial nerve in only one patient (case no. 4). After the thickened arachnoid was dissected, we found that the AICA was surrounded and immobilized at the REZ by the arachnoid (Fig. 2). We did not find vascular compression of the posterior inferior cerebellar artery (PICA), vertebral artery (VA) or the superior cerebrellar artery (SCA). We did not observe anatomical variations of the vessels at the base of the brain. In this group of pediatric patients, we did not find focally atherosclerotic change of the offending artery, which is very common in adult cases because of the hemodynamic effects brought about by the aging process or hypertension. In addition, we were impressed by the plump cerebellum of these pediatric patients, which made it

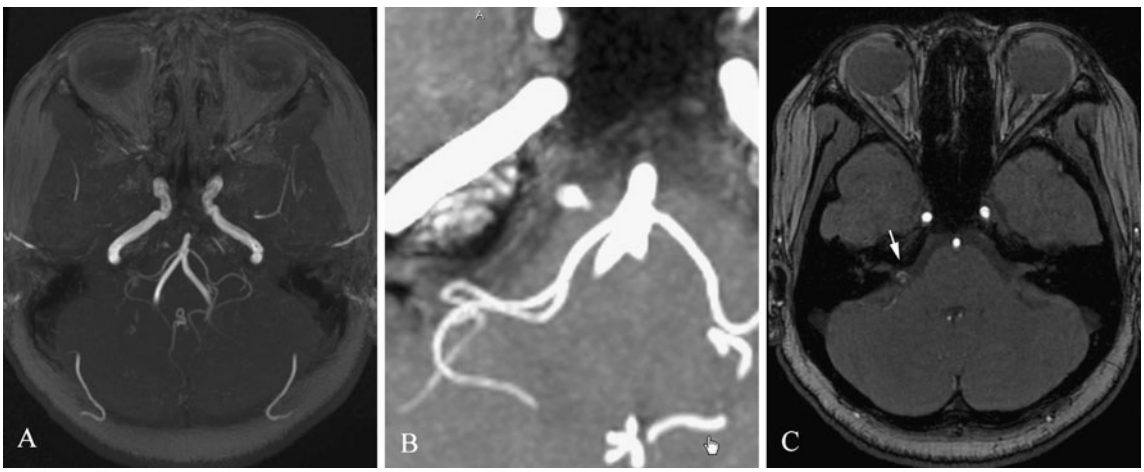

Fig. 1 Preoperative MRI of pediatric HFS patient. There definitely was an artery in close relation to the facial nerve. The offending vessels were not elongated or shifted. There was no obvious elongated, redundant vertebral artery

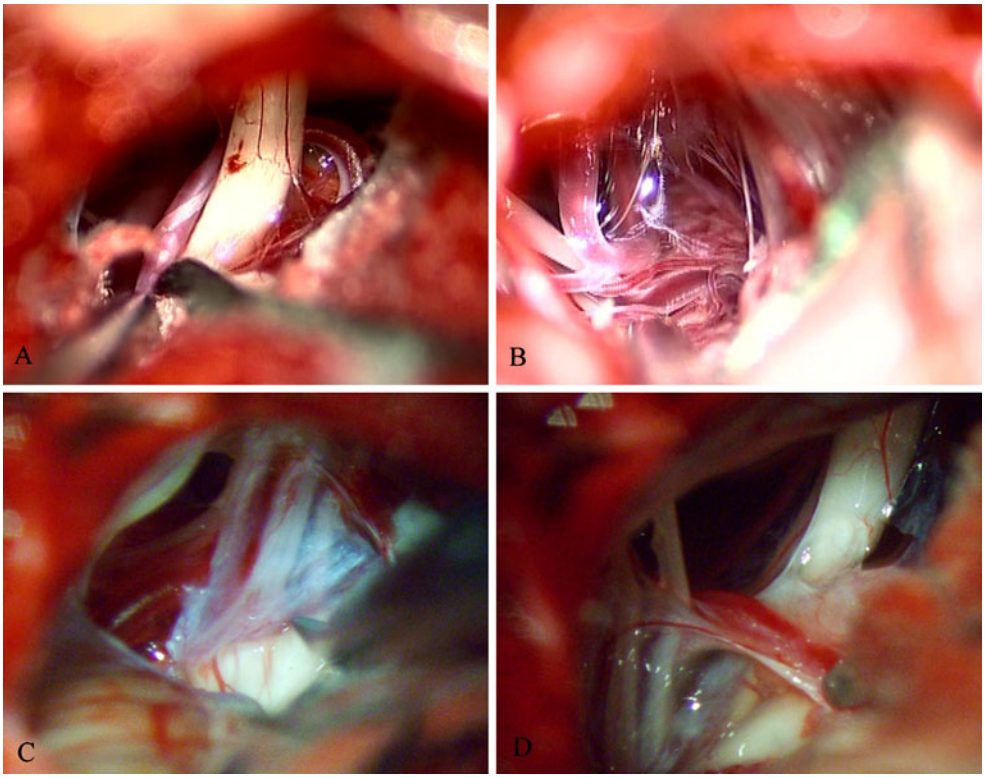

Fig. 2 **a**–**d** Intraoperative findings of pediatric HFS patients. **a** AICA compresses the REZ of facial nerve. **b** Besides the vascular compression of AICA, there is a vein in the REZ. **c** Arachnoid thickness was found around the roots of cranial nerves VII and VIII. **d** After the thickened arachnoid was dissected, we found the AICA was surrounded and immobilized at the REZ by the arachnoid

difficult to elevate the cerebellum and expose the cranial nerves. Therefore, more patience should be paid to draining the cerebrospinal fluid so as to provide enough space for cerebellar retraction.

Four typical HFS patients got immediate "excellent" or "good" resolution of spasms (Table 1). There was no recurrence during the follow-up. The atypical patient (case no. 5) had spasm relief to a degree of about 50%, and within the 27-month follow-up period the spasm did not resolve any more. For all cases, there were no surgical complications.

The score of social adaptation of the pediatric HFS patients was 111.6±8.2. Compared with the ordinary healthy Chinese middle-school students, whose score was 170.8±25.4 [9], this group seemed to experience great difficulty in social adaptation ($P<0.01$). After surgery, the scores of two patients (case nos. 1 and 3) increased to the normal level (Table 1); however, the other three patients remained unchanged.

Discussion

HFS frequently affects middle-aged and older individuals, especially in their 50s and 60s. Primary pediatric patients are extremely rare. Some pediatric HFS cases reported in the literature are symptomatic [6]. In this study, no patient had an anatomic malformation or cerebellopontine angle tumor which may cause symptomatic spasm.

The major offending vessel of all the five pediatric HFS patients was the AICA. It has been reported [3] that the PICA was the most common offending vessel among young HFS patients. However, we did not find the PICA, VA or SCA as culprit vessels.

The pathogenesis of primary HFS may differ in children since atherosclerotic changes do not occur in this population. MRI revealed that the shape and course of vertebral arteries are smooth and natural. However, offending vessels were definite, according to both MRI and intraoperative findings. As

Table 2 Summary of pediatric primary HFS patients (<18 years old at treatment) treated with MVD in the literature

ID	Author/ref.	Age of onset	Age of treatment	Spasm relief	Operative complications	Possible cause
1	Kobata et al. [10]	15	17	Immediate relief	None	Arachnoid thickness
2	Jho and Jannetta [8]	6	13	Delayed relief	NA	Vein complex
3		12	15	Delayed relief	NA	NA
4	Milani et al. [13]	7	10	Immediate relief	NA	NA

NA not available

mentioned above, it has been reported that venous offending vessels [12] and thickening arachnoid [10] may account for the etiology of HFS in youth. In this study, a thickening of the arachnoid turned out to be the cause of one case (no. 4). An offending vein was found in another case (no. 2), but the concomitant AICA seemed to be the real culprit vessel. Jho et al. [8] suggested that anatomical variations at the REZ might contribute to the development of HFS in childhood; however, we did not find any anatomical variations in these patients. We do not know the real reason why the AICA comes in close contact with the facial nerve, but we would suppose that it might just be due to variation and polymorphism in population.

In older patients, MVD is the most effective treatment with high curation and low morbidity rate. Some authors [3, 10] also reported that young HFS patients have good surgical outcomes as well (Table 2). In this study, four typical HFS patients had immediate excellent or good relief. However, MVD procedure did not help the atypical patient much. Therefore, we suppose that the decision for an MVD procedure for an atypical pediatric HFS patient should be very cautious, even if MRI indicates a close relation between an artery and the facial nerve.

Our findings also indicated that HFS did considerable harm to the children's social lives. Surprisingly, only two patients recovered to the normal social adaptation level after surgery. Cases no. 2 and 4 continued to give significantly lower scores even when their spasms were cured, maybe because of their relatively longer duration of spasm symptoms. We suppose that such patients need systemic psychological aid after surgery.

Conclusions

MVD is effective and safe for typical primary HFS patients younger than 18 years old. HFS is harmful to the children's social adaptation. However, only some patients recovered to the normal social adaptation level even when the spasms were cured.

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Conflicts of interest None.

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Comment

Owing to the huge number of hemifacial spasms (HFSs) treated in a very short period of time (2 years), Feng et al. were able to isolate five paediatric cases (<18 years old) out of a total of 1,304 patients who underwent a microvascular decompression (MVD) of the seventh nerve. As in the adult population, the typical cases greatly surpassed the atypical cases, and a general overview on this series shows that there are few clinical, radiological and surgical differences between children and adults, except their rarity at a young age, which was already known from previous individual case reports, and other particularities described in this paper, such as the AICA as the sole culprit for the compression and the "plump" aspect of the cerebellar hemisphere due to the younger age.

One of the interesting points is the appreciation of the result using a Chinese score of social adaptation for middle-school children, showing that, even if the surgery was uneventful and cured the HFS, only two patients improved to a normal score, the other three (two typical and one atypical HFS) had still some problem of social adaptation.

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