

Original Investigation

Stereotactic and Functional



Received: 06.02.2024 Accepted: 29.11.2024

Published Online: 25.06.2025

Non-Root Exit Zone Exploration during Facial Nerve MVD: A Discussion of the Pathogenesis in Atypical Cases of **Hemifacial Spasm**

Gaochao SONG^{1,2*}, Yuanyang WU^{1*}, Qi YAO¹, Guiping NI¹, Jianhong SHEN¹

¹Affiliated Hospital of Nantong University, Department of Neurosurgery, Nantong, Jiangsu Province, China ²Second Affiliated Hospital of Nanjing Medical University, Department of Neurosurgery, Nanjing, Jiangsu Province, China

ABSTRACT

AIM: To summarise atypical cases of hemifacial spasm (HFS) found during microvascular decompression (MVD) and to investigate its pathogenesis and range of exploration.

MATERIAL and METHODS: We retrospectively analysed cases of HFS performed in our department in recent years and summarised the intraoperative electrophysiological monitoring findings, vascular and nerve exploration, and postoperative symptoms. We then discussed the pathogenesis of and treatment for atypical HFS.

RESULTS: In total, 85 cases of facial nerve MVD were performed in the past 3 years, of which 77 (90.6%) were responsible factors in the root exit zone (REZ) and eight (9.4%) in the non-REZ. For patients without vascular compression of the REZ, the compression factors outside the REZ were separated, and the arachnoid band around the facial nerve was released; subsequently, the amplitude of the abnormal muscle response of the facial nerve diminished or disappeared. Facial twitch symptoms disappeared or improved significantly after surgery. Most symptoms disappeared after 3 months of postoperative follow-up.

CONCLUSION: Factors responsible for non-REZ observed during MVD of the facial nerve are not rare. It is suggested that fulllength exploration should be performed during facial nerve MVD under electrophysiological monitoring.

KEYWORDS: Hemifacial spasm, Microvascular decompression, Intraoperative electrophysiology, Abnormal muscle response

ABBREVIATIONS: HFS: Hemifacial spasm, MVD: Microvascular decompression, REZ: Root exit zone, AMR: Abnormal muscle response, MRTA: Magnetic resonance tomographic angiography, AICA: Anterior inferior cerebellar artery, PICA: Posterior inferior cerebellar artery, VA: Vertebral artery, BA: Basilar artery, SCA: Superior cerebellar artery

■ INTRODUCTION

emifacial spasm (HFS) is a common neurosurgical condition. The pathogenesis of HFS is generally believed to be vascular compression of the facial nerve root exit zone (REZ), resulting in changes in the local neural structure and involuntary twitching of the orbicularis oculi and orbicularis oris muscles due to similar abnormal discharges

(2). This is known as the microvascular compression theory, and based on this theory, microvascular decompression (MVD) has become the standard surgery for treating HFS; however, symptoms commonly fail to improve after MVD. In some cases, this failure may be related to incomplete surgical decompression, whereas in others, it may involve understanding the pathogenesis.

Gaochao SONG (D : 0000-0003-0092-8776

: 0000-0001-9445-0384

: 0000-0003-2231-0897 Guipina NI Jianhong SHEN (0): 0000-0003-2270-8555



This work is licensed by "Creative Commons Attribution-NonCommercial-4.0 International (CC)".

^{*}Gaochao Song and Yuanyang Wu contributed equally to this work

Our department has performed 85 HFS procedures in the past 3 years, including some atypical cases. In this paper, we analyse the surgical effects and particular characteristics observed intraoperatively in these patients and discuss the pathogenesis and surgical exploration methods of atypical cases.

MATERIAL and METHODS

This study has been approved by the Ethics Committee of the Affiliated Hospital of Nantong University (NO. 2024-K075).

Patient Data

The study included 85 patients aged 24 to 80 years (median, 55 years) with a disease course ranging from 1 month to 20 years (median, 3 years). Thirty-three were male (18 affected on the left, 15 on the right), and 52 were female (27 affected on the left, 25 on the right). Before the operation, neurotrophic medications were administered to 17 patients, carbamazepine to 28 patients, acupuncture to 13 patients (which proved beneficial in five cases), botulinum toxin injections to eight patients, and no treatment to 19 patients. All patients underwent facial nerve magnetic resonance tomography (MRTA) angiography before surgery.

Surgical Indications

The surgical indications were as follows: 1) a clear diagnosis of primary HFS; 2) MRTA revealing closely related blood vessels surrounding the facial nerve; 3) exclusion of history of facial nerve injury; 4) obvious symptoms that significantly impacted the quality of life and did not improve with conservative treatment; and 5) tolerance to general anaesthesia.

Surgical Material

Recently, China has been regulating the management of human implants. Teflon cannot be used for MVD surgery because it is not registered or accessible in China. Polyester is permitted for MVD surgery, and a medical gasket resembling Teflon is made entirely of polyester fibres. The safety and efficacy of MVD have been thoroughly confirmed after several years of clinical use. Follow-up findings were satisfactory.

Surgical Strategy

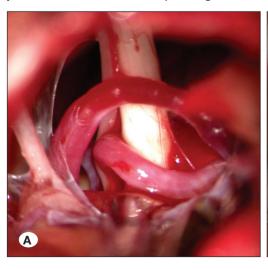
In the lateral recumbent position under general anaesthesia, a straight incision was created approximately 6 cm behind the affected side. A small bone window was opened behind the transverse sinus and sigmoid sinus. The blood vessels close to it were explored from the facial nerve REZ and separated with polyester. The operation was performed under a microscope or neuroendoscope. Furthermore, 80 patients were operated on under electrophysiological monitoring.

Atypical Cases

Case 1: Female, 51 years old, with paroxysmal convulsions of the left cheek and corner of the eye for 10 years. During the operation, the anterior inferior cerebellar artery (AICA) was shown to pass through the facial and auditory nerves, wherea the posterior inferior cerebellar artery (PICA) was ventral to the facial nerve immediately adjacent to the REZ; thus, the PICA and AICA formed a clamp in the REZ (Figure 1A). The facial nerve was separated from the two blood vessels using polyester; however, the AMR did not disappear. We continued to explore the facial nerve until the AMR disappeared before entering the inner ear foramen; however, no blood vessels were observed. Here, the AMR disappeared following the separation of the facial and auditory nerves using polyester (Figure 1B); it reappeared after withdrawal and disappeared after re-insertion.

Case 2: Female, 55 years old, with paroxysmal convulsions on the left side of the face for 3 months. During surgery, the PICA from the vertebral artery was found to oppress the initial segment of the facial nerve instead of the normal REZ. The AMR disappeared after the PICA was separated from the facial nerve, revealing that the AICA passed through the REZ of the facial and auditory nerve (Figure 2A). The PICA was carefully pushed away from the facial nerve with polyester, and the AICA was separated from the facial nerve (Figure 2B).

Case 3: Female, 53 years old, had paroxysmal convulsions on the left face for 9 years. During the operation, the facial and auditory nerves were found to be in close proximity, without gaps, seemingly merging into one branch. Electrophysiological



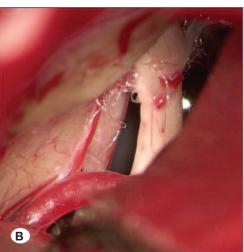
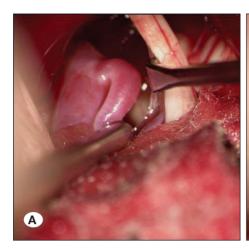


Figure 1: Vascular compression in the facial nerve root exit zone (REZ); however, the abnormal muscle response (AMR) did not disappear after treatment. A) The anterior inferior cerebellar artery (AICA) passed between the facial and auditory nerve, and the posterior inferior cerebellar artery (PICA) was ventral to the facial nerve immediately adjacent to the REZ. B) Near the inner ear foramen, the AMR disappeared after separating the facial nerve and auditory nerve.



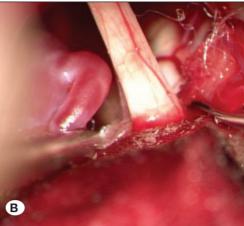
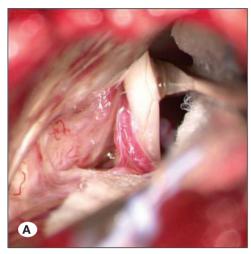


Figure 2: The posterior inferior cerebellar artery (PICA) from the vertebral artery oppressed the initial seament of the facial nerve and superimposed the anterior inferior cerebellar artery (AICA), forming a compression series. A) The PICA from the vertebral artery oppressed the initial segment of the facial nerve: the abnormal muscle response (AMR) disappeared after the PICA was pushed away. B) Separating the AICA passing between the root exit zone (REZ) of the facial nerve and the auditory nerve.



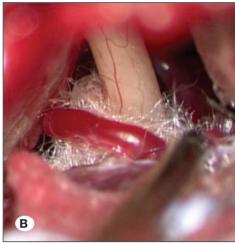


Figure 3: The intracranial segment of the facial nerve and auditory nerve are in close proximity and seem to merge into one branch. A) The anterior inferior cerebellar artery (AICA) hovered from the auditory nerve root exit zone (REZ) to the deep place. B) The abnormal muscle response (AMR) disappeared after separating the AICA from the facial nerve.

monitoring confirmed that the deep surface was the facial nerve fibre. The AICA hovered from the REZ of the auditory nerve to the middle of the facial nerve (Figure 3A). The AICA was separated from the facial nerve fibres with polyester (Figure 3B), and the AMR disappeared.

Case 4: Male, 56 years old, with paroxysmal convulsions of the left face for 2 years. During the operation, the AICA compressed the distal segment of the facial nerve. The AMR in the orbicularis oris muscle area disappeared after separating the AICA from the facial nerve with polyester (Figure 4A). There was no change in the AMR in the orbicularis oculi muscle area. After further exploration, a pulsatile great blood vessel was observed dorsal to the facial nerve REZ, which was confirmed as the vertebral artery (VA). Electrophysiological observations showed that the amplitude of the AMR in the orbicularis oculi muscle area decreased after the facial nerve was separated from the VA using polyester (Figure 4B).

■ RESULTS

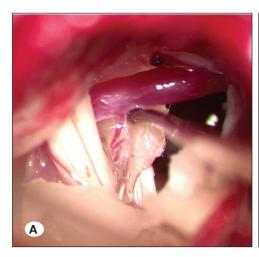
In most cases, one or two close vessels were found near the facial nerve REZ, including the basilar artery (BA) (n = 2), VA (n = 21), superior cerebellar artery (SCA) (n = 2), AICA (n = 41), PICA (n = 24), and the petrosal vein and its branches (n = 4). The remaining vessels were branched arterioles.

Of the 85 patients, 63 resolved after surgery, 13 showed clear relief, four had mild facial paralysis, and five had mild hearing loss. Twelve of the 13 patients who improved postoperatively no longer experienced convulsions at the 3-month follow-up. The four patients with mild facial paralysis fully recovered, and the five with mild hearing loss improved.

Four atypical cases were observed under the microscope, and all postoperative symptoms of facial convulsions vanished; however, in Case 4, convulsions recurred 4 days after the operation, although less than those before the operation, and vanished once more 9 days later. No postoperative effects on hearing were observed in any of the four cases.

DISCUSSION

HFS is a common functional disorder in neurosurgery, and understanding its pathogenesis directly affects the treatment. Since Jannetta proposed that vascular compression in the REZ is the main pathogenic factor for HFS, MVD has become the standard modality for the aetiology of HFS, with definite curative effects (3,7,8).



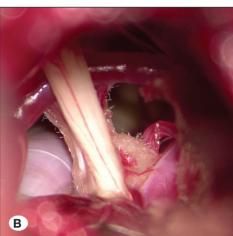


Figure 4: Vascular compression of the distal segment of the facial nerve. After treatment, there was no change in the abnormal muscle response (AMR) of the orbicularis oculi muscle area. A) The anterior inferior cerebellar artery (AICA) compressed the distal segment of the facial nerve. After separation, the AMR of the orbicularis oris muscle area disappeared. B) Vertebral artery compression was seen dorsal to the facial nerve root exit zone (REZ), and the AMR amplitude of the orbicularis oculi muscle area decreased after separation.

In the clinic, there are some instances where the conventional REZ has no clear accountable vessel during MVD, or electrophysiological alterations in the REZ are not immediately apparent following vascular separation. By summarising these cases, we propose novel ideas regarding the aetiology of therapeutic approaches for HFS.

The traditional theory of peripheral vascular compression suggests that local demyelination caused by vascular compression of the facial nerve REZ is the pathological basis for developing HFS, explaining the postoperative symptom relief and delayed remission in some cases where the intraoperative AMR did not disappear. However, the demyelination explanation could not account for symptoms that disappeared immediately after surgery in a significant percentage of patients, including 63 of the 85 patients included in our study. Zheng et al. proposed the sympathetic bridge hypothesis (12), which suggests that the facial nerve REZ is compressed and abraded by blood vessels over an extended period, thereby breaking the vascular wall. Consequently, the exposed sympathetic fibres of the vessel wall come in contact with the exposed facial nerve fibres after demyelination, and sympathetic excitation releases neurotransmitters that act on the damaged facial nerve and cause it to produce impulses, leading to clinical symptoms. Although the sympathetic bridge hypothesis explains the immediate postoperative disappearance of symptoms, it is still considered a peripheral theory.

Treatment strategies for the blood vessels in the REZ have always been the focus of research on facial nerve MVD. Park et al. categorised the vessels responsible for vascular loop compression into arachnoid, perforating arterial, vascular clamping, and series compression types; however, the actual situation of MRTA examination is that even on the healthy side, there is a high probability of the presence of closely related small blood vessels around the facial nerve, which leads to confusion regarding how to accurately identify the responsible vessels (9). Can completion of the treatment of vessels that compress the REZ be used as a criterion for the end of the operation? AMR is a delayed response to stimulation of one branch of the facial nerve motor branch, recorded in muscles innervated by other branches. It was first described in 1986. and its disappearance has been positively correlated with the

clinical results of MVD (6). Although the exact mechanism of AMR is unclear, similar to that of HFS, there are peripheral (demyelination of the facial nerve) and central hypotheses (4) (increased excitability of the facial nucleus). Microvascular decompression for HFS is now frequently accompanied by changes in AMR, and the dependability and efficacy of decompression can be assessed (13). Several studies have analysed such cases and concluded that AMR could be used as a "biomarker" for HFS, which is helpful for diagnosis and monitoring during MVD (10,13).

It was not unusual for AMR to disappear following non-REZ treatment in several instances, including Cases 1-4. In Case 1, the AICA and PICA clamped the facial nerve REZ, but the AMR persisted even after decompression. If decompression of the REZ was taken as the standard, the operation could be terminated. However, recent literature has also explained that the pathogenic factors causing HFS may include vascular and non-vascular factors outside the REZ (11), such as bony stenosis of the internal auditory meatus (5) and strangulation of the arachnoid band (1). In this case, the AMR disappeared after separating the facial and auditory nerves at the internal auditory meatus, which proved that the appearance of AMR may also be related to compression outside the REZ and illustrated that the disappearance of an AMR is an important indicator of surgical efficacy. The disappearance of AMR following the separation of the facial and auditory nerves may be due to the displacement of the facial nerve at the internal auditory meatus to avoid bony entrapment.

Intraoperative observations in Case 2 showed the PICA compressing the distal segment of the facial nerve REZ. The AMR disappeared after pushing the PICA away. Further, exploration showed that only the AICA passed through the facial nerve REZ. This may be due to cascade compression of the REZ by the PICA superimposed on the AICA. The pathogenesis of this case may be explained by the theory of peripheral vascular compression in the REZ; however, the compression of the distal segment of the facial nerve REZ by the PICA and the immediate disappearance of the AMR after separation of the two are inconsistent with the theory of compression in the REZ.

In Case 3, the AICA was seen hovering closely to the middle segment of the facial nerve, and the disappearance of the AMR after treatment was not consistent with the REZ compression theory. Similarly, the intraoperative disappearance of AMR in the orbicularis oris region in Case 4 did not result from treatment of the REZ; however treatment of the AICA in the non-REZ did not attenuate AMR in the orbicularis oculi region. Therefore, it is necessary to explore the blood vessels outside the REZ. The above cases show that the persistence of AMR after treatment of the REZ does not mean it does not disappear at the end of MVD. Many surgeons have also proposed the necessity for a full-length exploration of facial nerve root areas I–IV.

In summarising our data and combining our findings with those in the literature, we believe that vascular factors, bone compression, fibre adhesion, and nerve demyelination caused by peripheral factors involving the entire length of the facial nerve form an important pathological basis for HFS. The central factors (excitatory "igniting" of the facial nucleus) are indispensable for typical clinical symptoms. Full-length exploration of facial nerve MVD is necessary to improve the intraoperative disappearance rate of AMR and the postoperative negative rate of HFS. Of course, if the compression and traction factors have been removed, there is no need to worry about whether the AMR disappears, avoiding excessive perturbation of the facial and auditory nerves which may lead to increased complications.

CONCLUSION

The theory of vascular compression in the REZ cannot explain all the incidences of HFS. There is great uncertainty in the intraoperative identification of the responsible factors and the prediction of surgical effects. A full-length exploration of the facial nerve should be performed during MVD. For atypical HFS, electrophysiological monitoring of AMR is of great help in identifying the causative factors.

Declarations

Funding: This study was supported by the following grants: Jiangsu Provincial Medical Innovation Center (CXZX202212); Jiangsu Provincial Research Hospital (YJXYY202204); Medical Innovation Team Project of the "14th Five-Year Plan" Science Education and Health Project (NTCXTD48).

Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

AUTHORSHIP CONTRIBUTION

Study conception and design: GS, JS Data collection: GS, YW, QY, GN Analysis and interpretation of results: GS, JS

Draft manuscript preparation: GS, JS Critical revision of the article: GS, YW, JS

All authors (GS, YW, QY, GN, JS) reviewed the results and approved the final version of the manuscript.

■ REFERENCES

- El Refaee E, Marx S, Rosenstengel C, Baldauf J, Schroeder HWS: Arachnoid bands and venous compression as rare causes of hemifacial spasm: Analysis of etiology in 353 patients. Acta Neurochirurgica 162:211-219, 2019. https:// doi.org/10.1007/s00701-019-04119-5.
- Han IB, Chang JH, Chang JW, Huh R, Chung SS: Unusual causes and presentations of hemifacial spasm. Neurosurgery 65:130-137; discussion 137, 2009. https://doi.org/10.1227/01. NEU.0000348548.62440.42.
- Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS: Etiology and definitive microsurgical treatment of hemifacial spasm. J Neurosurgery 47:321-328, 1977. https://doi. org/10.3171/jns.1977.47.3.0321.
- Kameyama S, Masuda H, Shirozu H, Ito Y, Sonoda M, Kimura J: Ephaptic transmission is the origin of the abnormal muscle response seen in hemifacial spasm. Clin Neurophysiol 127:2240-2245, 2016. https://doi.org/10.1016/j. clinph.2016.02.004.
- Ligas B, Khatri D, Higbie C, Wagner K, Langer D: Hemifacial spasm due to bony stenosis of the internal auditory meatus: Look beyond the loop. World Neurosurgery 137:179-182, 2020. https://doi.org/10.1016/j.wneu.2020.01.196.
- Liu MX, Zhong J, Xia L, Dou NN, Sun H, Li B, Visocchi M, Li ST: The significance of abnormal muscle response monitoring during microvascular decompression for hemifacial spasm. Trends in reconstructive neurosurgery. Acta Neurochir Suppl 124:297-301, 2017. https://doi.org/10.1007/978-3-319-39546-3 43.
- Ma Q, Zhang W, Li G, Zhong W, Yang M, Zheng X, Yang X, Li S: Analysis of therapeutic effect of microvascular decompression surgery on idiopathic hemifacial spasm. J Craniofac Surg 25:1810-1813, 2014. https://doi.org/10.1097/SCS.000000000000000990.
- Park CK, Lee SH, Park BJ: Surgical outcomes of revision microvascular decompression for persistent or recurrent hemifacial spasm after surgery: Analysis of radiologic and intraoperative findings. World Neurosurg 131:e454-e459, 2019. https://doi.org/10.1016/j.wneu.2019.07.191.
- Park JS, Kong DS, Lee JA, Park K: Hemifacial spasm: Neurovascular compressive patterns and surgical significance. Acta Neurochir 150:235-241, 2008. https://doi.org/10.1007/s00701-007-1457-x.
- Sen CN, Møller AR: Signs of hemifacial spasm created by chronic periodic stimulation of the facial nerve in the rat. Exp Neurol 98:336-349, 1987. https://doi.org/10.1016/0014-4886(87)90246-9.
- 11. Son BC, Ko HC, Choi JG: Hemifacial spasm caused by vascular compression in the cisternal portion of the facial nerve: Report of two cases with review of the literature. Case Rep Neurol Med 2019:8526157, 2019. https://doi. org/10.1155/2019/8526157.
- 12. Zheng X, Hong W, Tang Y, Wu Z, Shang M, Zhang W, Zhong J, Li S: Sympathetic nerves bridge the cross-transmission in hemifacial spasm. Neurosci Lett 517:52-55, 2012. https://doi.org/10.1016/j.neulet.2012.04.023.
- Zhu W, Sun C, Zhang Y, Xu J, Wu S: AMR monitoring in microvascular decompression for hemifacial spasm: 115 cases report. J Clin Neurosci 73:187-194, 2020. https://doi. org/10.1016/j.jocn.2019.10.008.