Microvascular Decompression for Patient With Coexistent of Trigeminal Neuralgia, Hemifacial Spasm and Glossopharyngeal Neuralgia- a Case Report

Tao Sun  
Sun Yat-sen University First Affiliated Hospital

Wentao Wang  
First Affiliated Hospital of Guangdong Pharmaceutical College

Longshuang He  
First Affiliated Hospital of Guangdong Pharmaceutical College

Yu Su  
Sun Yat-sen University First Affiliated Hospital

Ning Li  
Sun Yat-sen University First Affiliated Hospital

Jinlong Liu  
Sun Yat-sen University First Affiliated Hospital

Chao Yang (✉ st1649496818@163.com)  
Sun Yat-sen University First Affiliated Hospital  https://orcid.org/0000-0002-9599-7194

Case report

Keywords: Microvascular Decompression, Trigeminal Neuralgia, Hemifacial Spasm, Glossopharyngeal Neuralgia

Posted Date: November 15th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-1064619/v1

License: © This work is licensed under a Creative Commons Attribution 4.0 International License.  Read Full License
Abstract

Background: Primary trigeminal neuralgia (TN), hemifacial spasm (HFS) and glossopharyngeal neuralgia (GN) are common diseases of nervous system, with similar pathogenesis and treatment strategies. Coexistent of such disease, especially coexistent of TN-HFS-GN simultaneously, is very rare. To date, only nine cases have been reported.

Case Presentation: A 70-year-old male with a history of hypertension and diabetes complained of severe involuntary contraction for about 10 years, knife-like and lighting-like pain, which was restricted to the distribution of the second and third branches of trigeminal nerve and pharynx and root of tongue, for about 2 years. Coexistent of TN HFS and GN was diagnosed and MVD was carried out. After MVD, the patient completely free from symptoms and no recurrence and hypoesthesia were recorded in 18 months follow up.

Conclusion: Here we report the tenth and oldest male patient with coexistent of TN-HFS-GN. Despite limited reports, MVD is the preferred choice for such diseases which can free patients from spasm and neuralgia.

Introductions

Incidence of primary TN, HFS and GN is quite different, TN and GN are characterized by extremely severe and electric shock-like pain restricted in the distributions of trigeminal and glossopharyngeal nerve while HFS is characterized by involuntary, usually unilateral and intermittent, contractions of muscles innervated by facial nerve[1, 2]. At present, it is widely accepted that compressions of adjacent vessels are the main causes of such diseases[3]. Compressions lead to local demyelination and hyperexcitability, then the nerve is prone to response to external stimuli[4]. For such disease, medication is generally preferred, and patients can usually get various degrees of symptoms relief, but long-term effect is relatively far from satisfactory[5, 6]. MVD can completely release nerves from neurovascular conflicts (NVCs) with no damage to the structural and functional integrities of nerves and it is currently the only widely accepted treatment that can fundamentally solve symptoms, maintain long-term efficacy with low complications rate[7]. In general, most patients suffer from one of the three mentioned cranial nerve diseases and can seldom suffer from two or more, which is called multiple hyperactive dysfunction syndrome (HDS) with a proportion of less than 3%, and only nine coexistent of TN-HFS-GN cases have been reported so far[8–10]. We share a case of coexistent of TN-HFS-GN caused by common arterial NVCs, to the best of our knowledge, it is also the oldest reported male case.

Case Presentation

A 70-year-old male with a history of hypertension and diabetes complained of severe involuntary contraction for about 10 years with no obvious triggers. It mainly involved left lower eyelid initially and gradually aggravated to involved left upper lip, which badly impacted his life quality. HFS was diagnosed in local hospital and treated with carbamazepine 200mg per day, which showed initial effect, but gradually wear off. Then knife-like and lighting-like pain occurred eight years later, which was restricted to the distribution of the second and third branches of trigeminal nerve and pharynx and root of tongue. Pain and muscle contraction usually attacked simultaneously, so the patient began to increase the dosage to 1200 mg per day gradually but did not respond well. Physical examinations showed as above mentioned with a Cohen score 4 and BNI pain intensity score 4, brain MRA indicated that left trigeminal nerve, hemifacial nerve and glossopharyngeal nerve were compressed by adjacent vessels, no other intracranial lesions were observed. (Figure 1-A-C)

Under general anesthesia, MVD was performed with a bone window modest wider than usual to fully expose the three nerves. Trigeminal, facial, and glossopharyngeal nerve were explored orderly, and we found extraordinarily complicated NVCs that trigeminal nerve was compressed by anterior inferior cerebellar artery, facial nerve and glossopharyngeal nerve were compressed by posterior inferior cerebellar artery simultaneously. (Figure 1-D-E) After dissected NVCs totally, Teflon patches were firmly inserted between the nerves and vessels. The patient fed back pain free but mild spasm in the first postoperative day, but gradually disappeared in a week. No complications were recorded, the patient complained of no pain, no involuntary contraction, no hypoesthesia during the 15-month follow-up.
Discussion

Primary TN, HFS and GN are mainly caused by compressions of adjacent vessels, REZ of each nerve, where oligodendrocyte of sheath myelin transits to Schwann cell, is the most common compressed area and it is vulnerable to external pressure[11]. Furthermore, for the similar pathogenic mechanisms and clinical manifestations of TN and GN, GN may manifest as pain in trigeminal nerve distribution, for example lower jaw[12]. It may lead to misdiagnose, so strictly identify is necessary. TN can also involve forehead, upper jaw and the anterior two-thirds of tongue beside lower jaw, GN mainly involves the posterior third of the tongue, pharynx or radiates to auricular area. A small number of GN patients may be accompanied by vagal stimulation symptoms for their close anatomical relationship[13]. Local anesthetic spraying is helpful to differentiate them, but attentions should be paid to coexistent of TN-GN, as TN were accompanied by GN in some conditions.

Some researcher held that looped VBA might played a part in the occurrence of coexistence of TN-HFS-GN[8, 10], a lower posterior fossa volume might involve in its occurrence, for that such disease many occurred in female, while study demonstrated that female has a posterior fossa volume than male[14], but the truly mechanism remains inclusive. Although there have been many previous reports about coexistent of TN and HFS or coexistent of TN and GN, only 9 cases have been reported about coexistent of TN-HFS-GN simultaneously[8–10], our case is the 10th case and the oldest male patient (Table 1). At present, treatments of mentioned diseases are similar, for example antiepileptic drugs, but Botulinum Toxin injection can also control the symptom of HFS, and partial sensory rhizotomy (PSR) could be used for TN and GN. Many patients can get remission, but these treatments are greatly limited for their impermanent and side effects. MVD can complete separate the nerves from NVCs to achieve long-term relief and it is the only method that can surely cure these diseases permanently[7].
Table 1

| Author               | Age(Y)/Sex/Side | Duration(Y) | Culprit Vessels (TGN;HFS; GPN) | Surgery               | Outcome                                                                 |
|----------------------|-----------------|-------------|--------------------------------|-----------------------|------------------------------------------------------------------------|
| Yong-Nan Wang et al. | 61/M/R          | HFS:14;TGN-GPN,4 | PV;VA+AICA;PICA              | MVD                  | Symptoms Resolved Immediately                                         |
|                      | 56/M/L          | TGN,5;GPN,3;HFS,2 | SCA;PICA;PICA               | MVD                  | Symptoms Resolved Immediately                                         |
|                      | 45 /F/R         | HFS,5;TGN-GPN,2 | SCA;AICA;PICA               | MVD                  | Symptoms Resolved (TGN within 1 week, HFS immediately,GPN improved)   |
|                      | 53/F/           | HFS,7;TGN,3;GPN,1 | SCA;VA;PICA                 | MVD                  | Pain resolved immediately and HFS within 3 month                      |
|                      | 69/F/L          | HFS,3;TGN,1;GPN,0.25 | AICA;PICA;PICA             | MVD                  | Symptoms Resolved (TGN within 2 week, HFS within 1 week,GPN improved) |
|                      | 77/F/L          | HFS,12;TGN,0.5;GPN,0.25 | SCA;VA+AICA;PICA          | MVD                  | Symptoms Resolved Immediately                                         |
| Jingwei Cao et al.   | 60/F/no mentioned | No mentioned | AICA+SCA;PICA;PICA          | MVD                  | Symptoms Resolved                                                      |
|                      | 65/F/no mentioned | no mentioned   | SCA;AICA+PICA;AICA         | MVD                  | Symptoms Resolved                                                      |
| Perez-Roman et al.   | 66 /M/R         | HFS:1;TGN-GPN,3  | VA                          | MVD+PSR(PGN)         | Symptoms Resolved                                                      |
| Tao SUN et al (current) | 70/M/L         | HFS,10;TGN-GPN,2 | AICA;PICA;PICA              | MVD+ sling           | Pain resolved immediately and HFS within 1 week                        |

We should clearly know that MVD for the three nerves at a time is extremely difficult, not only for the large number of NVCs, but also vertebrobasilar dolichoectasia or massive arteriovenous malformations are often occurred in such conditions[15, 16]. Therefore, preoperative imaging evaluation, bone window design and responsible vessels judgment are extremely important. In such patient, the bone window and dural incision should be mild wider than usual to fully expose NVCs, and it should be noted that we should first dissected all the compressions and then inserted Teflon patches. Once trigeminal decompression is totally accomplished, MVDs of facial nerve and glossopharyngeal nerve could be greatly restricted for limited operating space in such
surgery and even result in patches shift and incomplete decompression or omission responsible vessels: two common recurrent factors of such diseases[17, 18].

**Conclusion**

Here we report the tenth and the oldest male patient with coexistent of TN-HFS-GN. Despite limited reports, MVD is the preferred choice for such diseases which can greatly free patients from symptoms.

**Abbreviations**

TN: trigeminal neuralgia; HFS: hemifacial spasm; GN: glossopharyngeal neuralgia; MVD: microvascular decompression; NVCs: neurovascular conflicts; HDS: hyperactive dysfunction syndrome; PSR: partial sensory rhizotomy; PV: petrosal vein; SCA: superior cerebellar artery; AICA: inferior anterior cerebellar artery; PICA: posterior inferior cerebellar artery; VA: vertebral artery.

**Declarations**

**Acknowledgements:**

We would like to thank Chuangfeng Li M.D. for useful guideline on MRA.

**Author Contributions:**

_Yang:_ supervision, overall idea, and design of the study. _Sun:_ data collection, conception, writing and modification. _Li, Su and Liu:_ data collection, graphics production and literatures review.

**Funding:**

This study did not receive any funding or financial support.

**Submission statement:**

This manuscript is original and has not been submitted elsewhere in part or in whole.

**Ethics approval and consent to participate:**

This case report was approved by Ethics Committee of the First Affiliated Hospital of Sun Yat-sen University.

**Consent for publication:**

The authors have obtained the patient's written informed consent for print and electronic publication of this case report.

**Conflict of interests:**

no conflict of interest of all the authors.

**Informed consent:**

Not applicable for informed consent waiver has been approved.

**References**

1. Jiang C, Liang W, Wang J, Dai Y, Jin W, Sun X, et al. Microvascular decompression for hemifacial spasm associated with distinct offending vessels: A retrospective clinical study. Clin Neurol Neurosurg 2020;194:105876. 10.1016/j.clineuro.2020.105876.
2. Headache Classification Committee of the International Headache Society (IHS) the International Classification of Headache Disorders, 3rd edition. Cephalalgia 2018;38(1):1-211. 10.1177/0333102417738202.

3. Zhong J, Li ST, Zhu J, Guan HX, Zhou QM, Jiao W, et al. A clinical analysis on microvascular decompression surgery in a series of 3000 cases. Clin Neurol Neurosurg 2012;114(7):846-51. 10.1016/j.clineuro.2012.01.021.

4. Fujiwara S, Sasaki M, Wada T, Kudo K, Hirooka R, Ishigaki D, et al. High-resolution diffusion tensor imaging for the detection of diffusion abnormalities in the trigeminal nerves of patients with trigeminal neuralgia caused by neurovascular compression. J Neuroimaging 2011;21(2):e102-08. 10.1111/j.1552-6569.2010.00508.x.

5. Duarte GS, Rodrigues FB, Castelao M, Marques RE, Ferreira J, Sampaio C, et al. Botulinum toxin type a therapy for hemifacial spasm. Cochrane Database Syst Rev 2020;11:D4899. 10.1002/14651858.CD004899.pub3.

6. Jost WH, Laskawi R, Palmowski-Wolfe A, Urban PP. [therapy of hemifacial spasm with botulinum toxin]. Laryngorhinootologie 2019;98(4):247-51. 10.1055/a-0834-4188.

7. Zhang YQ, Yu F, Zhao ZY, Men XZ. Combined hyperactive dysfunction syndrome of the cranial nerves: Analysis of 37 cases and literature review. World Neurosurg 2019;129:e650-56. 10.1016/j.wneu.2019.05.237.

8. Perez-Roman RJ, Chen SH, Sur S, Leon-Correa R, Morcos JJ. A unique case of microvascular triple decompression for combined simultaneous trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia because of the dolichoectatic vertebrobasilar system. Oper Neurosurg (Hagerstown) 2020;18(6):692-97. 10.1093/ons/opz205.

9. Cao J, Jiao J, Du Z, Xu W, Sun B, Li F, et al. Combined hyperactive dysfunction syndrome of the cranial nerves: A retrospective systematic study of clinical characteristics in 44 patients. World Neurosurg 2017;104:390-97. 10.1016/j.wneu.2017.05.020.

10. Wang YN, Zhong J, Zhu J, Dou NN, Xia L, Visocchi M, et al. Microvascular decompression in patients with coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Acta Neurochir (Wien) 2014;156(6):1167-71. 10.1007/s00701-014-2034-8.

11. Araya E, Claudino RF, Piovesan EJ, Chichorro JG. Trigeminal neuralgia: Basic and clinical aspects. Curr Neuropharmacol 2020;18(2):109-19. 10.2174/1570159X17666191010094350.

12. Wang X, Meng D, Wang L, Chen G. The clinical characteristics and surgical treatment of glossopharyngeal neuralgia with pain radiating to the innervated area of the trigeminal nerve. J Oral Maxillofac Surg 2021;79(4):781-86. 10.1016/j.jmsj.2020.11.034.

13. Shah RJ, Padalia D. Glossopharyngeal neuralgia. 2021.

14. Chan LL, Ng KM, Fook-Chong S, Lo YL, Tan EK. Three-dimensional MR volumetric analysis of the posterior fossa CSF space in hemifacial spasm. Neurology 2009;73(13):1054-57. 10.1212/WNL.0b013e3181b9c8ce.

15. Dou NN, Hua XM, Zhong J, Li ST. A successful treatment of coexistent hemifacial spasm and trigeminal neuralgia caused by a huge cerebral arteriovenous malformation: A case report. J Craniofac Surg 2014;25(3):907-10. 10.1097/SCS.0000000000000567.

16. Lakhan SE. Teaching NeuroImages: Concurrent hemifacial spasm and trigeminal neuralgia due to vertebrobasilar dolichoectasia. Neurology 2013;81(8):e52. 10.1212/WNL.0b013e3182a1aeb.

17. Liu J, Wu G, Xiang H, Liu R, Li F, Hei B, et al. Long-Term retrospective analysis of microvascular decompression in patients with recurrent trigeminal neuralgia. Front Neurol 2020;11:584224. 10.3389/fneur.2020.584224.

18. Feng BH, Wang XH, Li ST. Posterior fossa Re-Exploration for recurrent trigeminal neuralgia: Operative findings and surgical techniques. J Craniofac Surg 2018;29(5):1284-86. 10.1097/SCS.0000000000004576.

Figures
Figure 1

A-C. MRA indicated that trigeminal nerve was compressed by anterior inferior cerebellar artery (red arrow) originates from distal part of basilar artery, facial nerve and glossopharyngeal nerve were compressed by posterior inferior cerebellar artery (yellow arrow green artery) originates from distal part of vertebral artery. D-E. Trigeminal nerve was compressed by anterior inferior cerebellar artery, facial nerve and glossopharyngeal nerve were compressed by posterior inferior cerebellar artery simultaneously.