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TECHNICAL NOTE - NEUROSURGICAL TECHNIQUES

Microvascular decompression in patients with coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia

Yong-Nan Wang · Jun Zhong · Jin Zhu · Ning-Ning Dou · Lei Xia · Massimiliano Visocchi · Shi-Ting Li

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Abstract
Background Trigeminal neuralgia(TN), hemifacial spasm (HFS) and glossopharyngeal neuralgia (GPN) were referred to hyperactive dysfunction syndromes (HDSs) of the cranial nerves. These symptoms may occur synchronously or metachronously, but the combination of three diseases is extremely rare.
Methods From 2007 through 2013, six patients with coexistent GPN-HFS-TN were treated in our department. The combined symptoms occurred on the same side in three and on both sides in three. These patients underwent nine microvascular decompression (MVD) procedures in total. The clinical data including operative findings were respectively analyzed, and the etiological factors as well as treatment strategies were discussed.
Results Intraoperatively, in all the cases a small posterior fossa was found, which was crowded with cranial nerve roots and cerbellar vesels. Postoperatively, spasm was stopped immediately in four and within 3 months in two; the symptom of TN disappeared immediately in four and within 2 weeks in two; the symptom of GPN was relieved immediately in four and improved with medication in two. During the up to 77 months' follow-up, no changes, recurrence or any dysfunctions of cranial nerves were observed in any of the patients.
Conclusions The combination of HFS-TN-GPN is extremely rare and is often associated with a looped VBA and a smaller posterior fossa. However, MVD is still a good choice for treatment. To achieve a safe and effective outcome, dissection of the caudal cranial nerves and proximal transposition of the vertebral artery before decompression of the affected nerve roots are strongly recommended.

Keywords Combined hyperactive dysfunction syndrome · Posterior fossa · Looped vertebrobasilar artery · Microvascular decompression · Trigeminal neuralgia · Hemifacial spasm · Glossopharyngeal neuralgia

Introduction

Trigeminal neuralgia(TN), hemifacial spasm (HFS) and glossopharyngeal neuralgia (GPN) are referred to hyperactive dysfunction syndromes (HDSs) of the cranial nerves, which are believed to be caused by vascular compression at the root entry or exit zone of the cranial nerves [10, 11, 13, 21]. These clinical syndromes may appear alone or together [18]. TN and HFS are the most common presentations of the HDSs, with a prevalence rate of 5 to 10 per 100,000 [1, 20, 33, 37, 40], whereas GPN is rare, with an incidence of 0.7 per 100,000 (0.9 and 0.5 in men and in women, respectively) [2, 17, 26]. These symptoms may occur synchronously or metachronously, but the combination of three diseases is extremely rare [19]. Combined HDS is defined as when more than one disease is involved; this may occur on one or both sides. Various treatment modalities have been proposed so far, and the most effective and worldwide accepted management is microvascular decompression (MVD) of the nerve roots, which is capable of providing complete resolution of symptoms in the majority of cases [15, 39]. The present work was carried out to respectively analyze those HDS cases treated by MVD in our department and to discuss etiological factors as well as treatment strategies.

Y.-N. Wang · J. Zhong (✉) · J. Zhu · N.-N. Dou · L. Xia · S.-T. Li
Department of Neurosrugery, XinHua Hospital (The Cranial Nerve Disease Center of Shanghai), Shanghai JiaoTong University School of Medicine, Shanghai 200092, China
e-mail: zhongmdphd@sjtu.edu.cn

Y.-N. Wang
e-mail: hnwang_2010@126.com

M. Visocchi
Institute of Neurosurgery, Catholic University of Rome, Rome, Italy

Table 1 Summary of the patients' data

Patient	Sex/age (years)	Affected side	Duration of symptoms	Offenders	Outcomes
1	M/61	R.TN-GPN-HFS	TN-GPN, 4 years; HFS,14 years	PICA (IX), PV (V), VA & AICA (VII)	Symptoms disappeared immediately
2	M/56	L.TN-GPN-HFS	TN, 5 years; GPN, 3 years; HFS, 2 years	PICA (IX & VII), SCA (V)	Symptoms disappeared immediately
3	F/45	L.TN-GPN R.HFS	TN-GPN, 2 years; HFS,5 years	AICA (VII), SCA (V) PICA (IX)	Spasm ceased immediately Symptom of TN relieved within 1 week Symptom of GPN improved
4	F/53	L.GPN-HFS R.TN	TN,3 years; GPN, 1 years; HFS, 7 years	VA (VII), SCA (V), PICA (IX)	Pain free immediately Spasm relieved within 3 months
5	F/69	L.TN-HFS R.GPN	TN,1 years; GPN, 3 months; HFS, 3 years	PICA (IX & VII), AICA (V)	Symptom of TN relieved within 2 weeks Symptom of GPN improved Spasm relieved within 1 week
6	F/77	L.TN-GPN-HFS	TN, 6 months; GPN, 3 months; HFS, 12 years	PICA (IX), SCA (V), VA & AICA (VII)	Symptoms disappeared immediately

AICA anterior inferior cerebellar artery; *PICA* posterior inferior cerebellar artery; *SCA* superior cerebellar artery; *VA* vertebral artery; *V* trigeminal nerve; *VII* facial nerve affected; *IX* glossopharyngeal nerve

Clinical materials and methods

From 2007 through 2013, six patients with coexistent HFS-TN-GPN were treated in our department. Those patients, two men and four women with an average age of 61 years old, presented with facial spasm and/or pain for 3 months to 14 years. Except for one, all patients were started with HFS followed by TN and/or GPN after 2 to 11 years. The combined symptoms occurred on the same side in three of the patients, and the other three had the symptoms on both sides (Table 1). They had been treated by medications or acupuncture, but in vain. All patients underwent a three-dimensional time-of-flight magnetic resonance imaging (3D-TOP MRI) scan pre-operatively, and no neoplasms were found in the cerebellopontine area, but a tortuous vertebrobasilar artery (VBA) loop (Fig. 1).

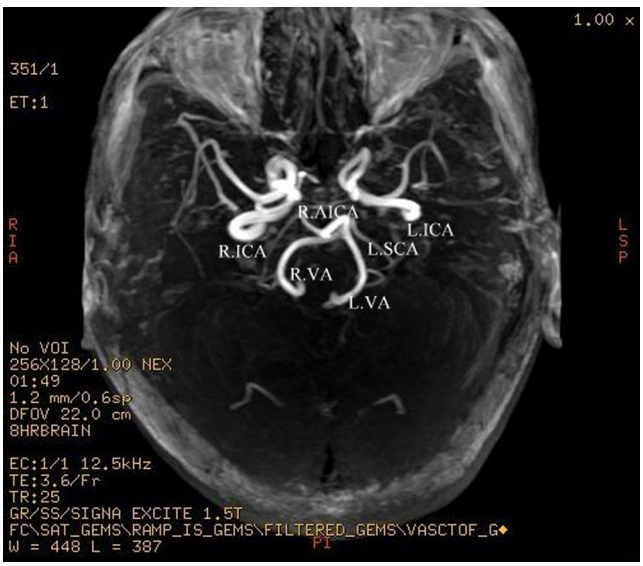

Fig. 1 Magnetic resonance imaging findings. Three-dimensional time-of-flight magnetic resonance imagings delineated a tortuous vertebrobasilar artery (VBA) loop in the cerebellopontine area in all the cases. V: trigeminal nerve; VII: facial nerve; IX: glossopharyngeus nerve. R.AICA: right anterior inferior cerebellar artery; L.ICA: left internal carotid artery; R.ICA: left internal carotid artery; L.SCA: superior cerebellar artery; L.VA: left vertebral artery; R.VA: right vertebral artery

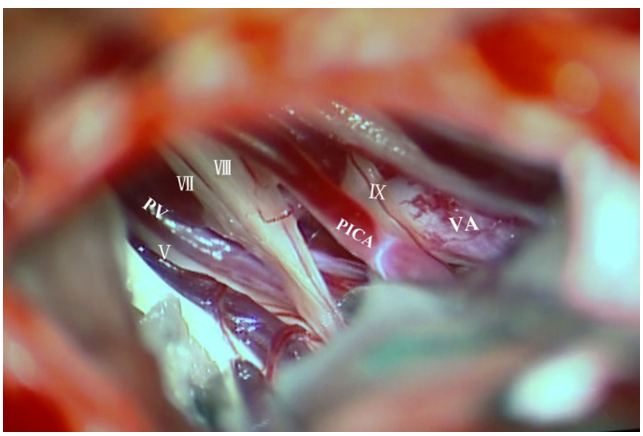

Fig. 2 Intraporative findings. With microscopy, a crowded cerebellopontine angle (CPA) was visualized. V: trigeminal nerve; VII: facial nerve; VII: vestibulocochlear nerve; IX: glossopharyngeus nerve; VA: vertebral artery. PICA: posterior inferior cerebellar artery; PV: petrosal vein

These patients underwent nine MVD procedures in total. When bilateral cranial nerves were affected, the second MVD was performed 2 weeks later.

Surgery

The operation was performed with a routine retrosigmoid approach [12, 36, 38]. The dissection started from the caudal cranial nerves. Once the arachnoid membrane around the nerves had been opened, the cerebellum was gradually raised until the pontomedullary sulcus was visualized. The vascular relationship was carefully studied to identify the vessel in contact with glossopharyngeal or facial nerves. After the offending vessel had been moved away from the nerve, small pieces of shredded Teflon sponge were gently placed between the vessel and the nerve. Next, the microscopic view was transferred rostrally to show the corner between the tentorium and os petrosum. Once all the arachnoids surrounding the trigeminal nerve had been opened sharply, the whole intracranial root of the fifth nerve, including the lateral pons and root entry zone, was thoroughly exposed. Any artery discovered contacting the nerve was transposed, and more pieces of Teflon were then put between the nerve and vessel. Meanwhile, any venule attaching to the fifth nerve was coagulated and cut. Finally, the dura mater was sutured in a watertight pattern.

Results

Operative findings

During surgery, the cerebellopontine angle (CPA) cisterns of all patients were found narrower than in controls. In such a small area, cranial nerves (CN-V, VII and IX) and vessels were crowded. As shown in Fig. 2a, a large looped VBA was identified in all the cases. This VBA raised the PICA and affected both CN-VII and -IX in cases 2 and 5, while it raised the AICA and affected CN-VII in cases 1, 3 and 6. The looped VBA directly compressed CN-VII in patient 4 (Table 1).

Postoperative outcomes

Postoperatively, as soon as the six patients awoke from the anesthesia, the spasm stopped immediately in four and within 3 months in two; the symptom of TN disappeared immediately in four and within 2 weeks in two; the symptom of GPN was relieved immediately in four and improved with medication in two (Table 1). They were followed up for 3–77 months, and no recurrence or any dysfunction of cranial nerves was found.

Discussion

TN, HFS and GPN were referred to hyperactive dysfunction syndromes by Jannetta in 1990 [13]. The combination of these dysfunctions has been described as multiple cranial nerve dysfunction [14], multiple cranial neuropathy [8], multiple brain stem and REZ compression syndrome [27], and painful tic convulsif (especially in the combination of TN and HFS) [24]. Although there have been a considerable number of reports describing bilateral TN [3, 6, 25, 27, 29–32], an ipsilateral combination of TN–HFS [9, 23, 28, 39] or TN-GPN [4, 5, 16, 22, 35] individually, very few have been reported on the combination of HFS-TN-GPN so far.

Chan et al. [7] conducted a case-control MRI volumetric study in 82 subjects (41 patients and 41 controls). It was found that the CSF volume in the posterior fossa was lower in patients with HFS compared to matched controls, which was more predominant in females. The condition of posterior fossa crowding may explain the etiological characteristics of vascular compression at the REZ of the respective nerve causing HDS. Our previous study regarding the painful tic convulsif also found a large looped VBA in most of the concurrent HFS-TN patients [39]. Kyung-Hoon Yang et al. [34] retrospectively studied 51 patients with combined HDSs. Comparing combined and single HDS groups, they found that old age was closely associated with the prevalence of combined HDS. The aging process and arteriosclerotic changes of the vessel might facilitate elongation and redundancy of vessels, inclining them to compress the root entry/exit zone of the cranial nerves.

The outcomes of MVD for combined syndromes are satisfactory and comparable to those achieved in the single syndrome series. Still, greater care must be taken to prevent operative complications, because this subgroup consists of relatively older patients, and the posterior fossa is usually narrower and smaller, and also a large looped VBA is always encountered. Based on our experience, to achieve a more effective outcome and a safer operation process, dissection beginning from the caudal cranial nerves and moving the VBA proximally before decompression of the affected cranial nerve roots is strongly recommended.

Conclusions

The combination of HFS-TN-GPN is extremely rare and is often associated with a looped VBA and a smaller posterior fossa. However, MVD is still the treatment of choice. It was suggested to move the VBA proximally before decompression of those with affected cranial nerve roots.

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Conflict of interest None.

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