INTRODUCTION

Neurovascular compression syndrome (NVCS) is defined as compression of a cranial nerve by a redundant or aberrant vascular structure at the root entry zone or root exit zone (REZ) of the corresponding cranial nerve. Trigeminal neuralgia (TN) is the most frequent syndrome, followed by hemifacial spasm. Despite ongoing debate in the literature, pulsatile tinnitus is also known to be a neurovascular compression syndrome resulting from vascular compression of the vestibulocochlear nerve. Combined manifestations of neurovascular compression symptoms are very rare. We report a case with a simultaneous manifestation of trigeminal neuralgia, hemifacial spasm, and pulsatile tinnitus as neurovascular compression syndrome with separate offending vessels. A 53-year-old female presented with lancinating pain in the left face, left hemifacial spasm, and ipsilateral pulsatile tinnitus for 2 years. Trigeminal neuralgia and hemifacial spasm were diagnosed after a neurological examination, imaging study, and electromyography. Microvascular decompression via a retrosigmoid approach for separate offenders, including the superior cerebellar artery, anterior inferior cerebellar artery, and posterior inferior cerebellar artery was performed using Teflon sponges. The patient’s hemifacial spasm, pulsatile tinnitus, and hemifacial pain improved immediately after microvascular decompression.

KEY WORDS: Trigeminal neuralgia, Hemifacial spasm, Tinnitus, Microvascular decompression surgery

Microvascular triple decompression for combined simultaneous trigeminal neuralgia, hemifacial spasm, and pulsatile tinnitus due to separate offending vessels

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Neurovascular compression syndrome is generally caused by vascular compression at the root entry or exit zone of the corresponding cranial nerve. Trigeminal neuralgia is the most frequent syndrome, followed by hemifacial spasm. Despite ongoing debate in the literature, pulsatile tinnitus is also known to be a neurovascular compression syndrome resulting from vascular compression of the vestibulocochlear nerve. Combined manifestations of neurovascular compression symptoms are very rare. We report a case with a simultaneous manifestation of trigeminal neuralgia, hemifacial spasm, and pulsatile tinnitus as neurovascular compression syndrome with separate offending vessels. A 53-year-old female presented with lancinating pain in the left face, left hemifacial spasm, and ipsilateral pulsatile tinnitus for 2 years. Trigeminal neuralgia and hemifacial spasm were diagnosed after a neurological examination, imaging study, and electromyography. Microvascular decompression via a retrosigmoid approach for separate offenders, including the superior cerebellar artery, anterior inferior cerebellar artery, and posterior inferior cerebellar artery was performed using Teflon sponges. The patient’s hemifacial spasm, pulsatile tinnitus, and hemifacial pain improved immediately after microvascular decompression.

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Neurovascular compression syndrome (NVCS) is defined as compression of a cranial nerve by a redundant or aberrant vascular structure at the root entry zone or root exit zone (REZ) of the relevant cranial nerve from the brainstem [1-5]. Trigeminal neuralgia (TN) is the most frequent syndrome with an incidence of 4–20/100,000 population [6,7], followed by hemifacial spasm (HFS), glossopharyngeal neuralgia (GN), and vestibular paroxysmia [8]. Pulsatile tinnitus is also known NVCS by vascular compression of the caudal surface (cochlear nerve) of the vestibulocochlear nerve [9]. However, this condition has not been clearly defined.

The combined manifestation of NVCS is very rare with the prevalence of ap-
Microvascular triple decompression

approximately 3% \[10,11\]. Previous studies indicated the combination of TN-HFS-GN by multiple offending vessels or dolichoectatic single offender \[10\]. Pulsatile tinnitus is sometimes accompanied with HFS due to the anatomically close relationship between the cochlear nerve and the facial nerve.

Here, we report a unique case of simultaneous coexistence of left TN, HFS, and pulsatile tinnitus because of the separate offending vessels including right superior cerebellar artery (SCA), posterior inferior cerebellar artery (PICA), and anterior inferior cerebellar artery (AICA), respectively, in which the symptoms were successfully treated with microvascular decompression (MVD).

CASE REPORT

A 53-year-old female with no past medical history presented paroxysmal lancinating pain in left face, left HFS and ipsilateral pulsatile tinnitus for 2 years. On physical examination, neurological examination revealed intermittent paroxysmal lancinating pain in left V1, V2, and V3 dermatome. She was treated with medication using tegretol and botulinum toxin injections. Although, after three times botulinum toxin injections for 1 year, she was suffered from progressive left facial pain, especially V1 dermatome, which was often triggered by minimal stimulation of the affected area and worsened ipsilateral pulsatile tinnitus with HFS.

Magnetic resonance imaging angiography revealed a neurovascular compression of the left trigeminal nerve and facial nerve by the SCA and the PICA, respectively (Fig. 1, 2). The patient was taken to MVD surgery through a retrosigmoid approach. Intraoperative neurophysiological monitoring (facial nerve electromyography and brainstem auditory evoked potential [BAEP]) monitoring was conducted. Neurovascular compression was confirmed with intraoperative finding caused by SCA at root entry zone for trigeminal nerve, PICA at REZ for facial nerve, and AICA at cisternal portion for vestibulocochlear nerve (Fig. 3–5). The offending vessels were moved away from the site and the decompression was secured by inserting Teflon sponges, placed between the offending vessels and the cranial nerves, respectively. The patient reported complete resolution of all her previous symptoms including lancinating facial pain, HFS and pulsatile tinnitus in the immediate postoperative period. She remained neurologically intact and asymptomatic at 36 months follow-up visits.

Ethical statements

Informed consent was waived by the institutional review board.

DISCUSSION

NVCS is characterized by functional disturbance of a cranial nerve by vascular compression at the REZ of the relevant cranial nerve from the brainstem. The accepted underlying pathophysio-

Fig. 1. Preoperative 3-T time-of-flight magnetic resonance angiography sequence shows the left superior cerebellar artery compressing the trigeminal nerve.

Fig. 2. Preoperative 3-T time-of-flight magnetic resonance angiography sequence shows the left posterior inferior cerebellar artery loop site causing neurovascular compression of the facial nerve.
logical mechanism of these compression syndromes suggested that the nerve encounters a blood vessel in the region of the REZ of the relevant cranial nerve at the brainstem. The nerve is especially sensitive to mechanical stimulation at the site of transition between central and peripheral myelin, causing clinical symptoms of nerve compression [2,8].

TN is the most common NVCS with overall prevalence is estimated to be in the range of 4–20/100,000 population [6,7], followed by HFS, GN, and vestibular paroxysmia [8]. TN is usually induced by the SCA alone or in association with other vessels was the compression artery in 88% of patients, AICA in 25%, and a vein in 5.5% [8]. For HFS, Barker et al. [12] reported compression of the facial nerve was by the PICA in 68%. The AICA and the vertebral artery accounted for most of the remaining cases.

Combined NVCS is rare. Combined NVCS with TN, HFS, and GPN has only been reported nine times in the literature [10]. Most cases of combined NVCS were associated with a tortuous vertebobasilar artery and a narrow posterior fossa [13]. When it comes to tinnitus, it has various etiologies. Therefore, it is difficult to determine if a patient’s tinnitus is related to NVCS of the auditory nerve. MVD for pulsatile tinnitus provide benefit to patients with accurate diagnosis of NVCS.

In this case, TN, ipsilateral HFS, and pulsatile tinnitus on the ipsilateral side were present simultaneously. In most of the previous literature, it was often caused by a vertebobasilar artery and a narrow occipital fossa. However, in this case, it is noteworthy that combined NVCS was caused by separate offenders.

What was disappointing in this case was that the BAEP test was not performed before the operation, because pulsatile tinnitus was initially considered as an indirect auditory nerve compression symptom of facial nerve compression. Usually, in the case of pulsatile tinnitus, there are characteristic BAEP findings, but it is a pity that it was not confirmed before surgery.

The selection criteria for MVD operations for tinnitus have varied. All the patients for the above discussed study [14] had severe tinnitus and were selected for the operation on the basis of history and to some extent on audiological test results. Narrow dips in the pure tone audiogram, poorer speech discrimination than normal based on hearing loss are signs of involvement of the auditory
nerve. Abnormalities in the BAEP, in the form of prolonged interpeak latencies I–III and delayed or absent peak II were strong indications of vascular contact with the auditory nerve.

Unlike the trigeminal nerve and facial nerve, the characteristic of auditory nerve vascular compression is not only found mainly in the neuromuscular origin, but also in the junction between the neuromuscular origin and the periphery in many cases. In our case, it was confirmed that the trigeminal nerve and facial nerve were being compressed at the root of the nerve root, but in the case of the auditory nerve, they were being compressed further away from the origin.

MVD is non ablative procedure without association with facial numbness, therefore, it is the most attractive option for NVCS such as TN and HFS, where surgical success rates are approximately 80–88% and 80%, respectively [15,16]. MVD for tinnitus has lower success rate. Only a few studies of the results of MVD operations for tinnitus have been published involving much fewer patients than studies of MVD operations for TN and HFS. The reported results of MVD operations for tinnitus are not nearly as encouraging as they are for TN, HFS or pulsatile tinnitus. Therefore, it is even more complex and demanding to select tinnitus patients for MVD operations. In this case, preoperative BAEP was not performed for pulsatile tinnitus. During the operation monitoring for tinnitus, improved. If pulsatile tinnitus caused by vascular compression, it’s better to perform preoperative BAEP as baseline study.

CONCLUSION

The combined NVCS composed of TN, HFS, and pulsatile tinnitus is rare and may be manifested with separate offenders. In such cases, MVD using the Teflon via retrosigmoid approach should be considered as an effective and generally safe option for the transposition of the offending artery and decompression of the affected nerve roots and brainstem.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES