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Hemifacial Spasm Caused by Vascular Compression of the Distal Portion of the Facial Nerve Associated with Configuration Variation of the Facial and Vestibulocochlear Nerve Complex

Fasial ve Vestibülokoklear Sinir Kompleksinin Konfigürasyon Varyasyonuyla İlişkili Olarak Fasial Sinir Distal Kısmının Vasküler Kompresyonuna Bağlı Hemifasial Spazm

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ABSTRACT

It is generally accepted that hemifacial spasm (HFS) is caused by vascular compression at the root exit zone (REZ) of the facial nerve. We saw an HFS patient caused by vascular compression of the distal portion of the facial nerve associated with configuration variation of the facial-vestibulocochlear nerve complex. A 50-year-old female with left HFS was admitted to our hospital. Preoperative magnetic resonance image demonstrated no offending artery around the facial nerve at the nerve's REZ. Microvascular decompression of the left seventh cranial nerve was performed via a lateral suboccipital infrafloccular approach. The facial nerve arose more than 5 mm away from the vestibulocochlear nerve in the brain stem and both traveled apart toward the internal acoustic meatus in the cerebello-pontine cistern. No offending vessel was observed near the REZ of the facial nerve. The abnormal muscle responses of the mentalis muscle disappeared when the AICA was separated from the distal portion of the facial nerve. The patient was completely free of the HFS following surgery. The facial nerve arising away from the vestibulocochlear nerve in the brain stem is rare. It might influence the cause of HFS with compression of the distal portions of the seventh cranial nerve.

KEY WORDS: Configuration variation, Distal portion of the facial nerve, Hemifacial spasm, Root exit zone, Vascular compression, Vestibulocochlear nerve

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ÖZ

Genellikle kabul edilen hemifasial spazmın (HFS) fasial sinirin sinir çıkış bölgesinde (REZ) vasküler basısı nedeniyle oluşmasıdır. Fasial-vestibülokoklear sinir kompleksinin konfigürasyon varyasyonuyla ilişkili olarak fasial sinir distal kısmının vasküler kompresyonuna bağlı bir HFS hastası deneyimimiz oldu. 50 yaşında bir sol HFS hastası bayan hastanemize başvurdu. Preoperatif manyetik rezonans görüntüleme fasial sinirin REZ bölgesi çevresinde bası oluşturan herhangi bir arter göstermedi. Lateral suboksipital infraflokular yaklaşımla sol yedinci kranyal sinirin mikrovasküler dekompresyonu yapıldı. Beyin sapında fasial sinir vestibülokoklear sinirden 5 mm den fazla uzaklaştı ve cerebello-pontin sistern içinde her ikisi internal akustik meatusa doğru ayrı ayrı gidiyorlardı. Fasial sinirin REZ bölgesinde bası oluştural bir damar görülmedi. AİCA fasial sinirin distal kısmından ayrıldığında mentalis kasındaki anormal cevaplar kayboldu. Cerrahi sonrasında hastanın HFS'si tamamen düzelmişti. Beyin sapında vestibülokoklear sinirden ayrı çıkan fasial sinir nadirdir. Yedinci kranyal sinirin distal kısımlarının kompresyonu ile birlikte olan HFS'nin sebebini etkileyebilir.

ANAHTAR SOZCÜKLER: Konfigürasyon varyasyonu, Fasial sinir distal kısmı, Hemifasial spazm, Kök çıkış bölgesi, Vasküler bası, Vestibülokoklear sinir

INTRODUCTION

It is generally accepted that hemifacial spasm (HFS) is caused by vascular compression at the root exit zone (REZ) of the facial nerve. However, patients with HFS caused by compression of distal portions of the seventh cranial nerve have recently been reported (1,12,14,17). In this study, we report a rare case of HFS caused by vascular compression of the distal portion of the facial nerve associated with configuration variation of the facial-vestibulocochlear nerve complex. Anatomic variation of the facial-vestibulocochlear nerve complex and magnetic resonance (MR) image techniques to understand the relationship between vascular structures and cranial nerves in the cerebello-pontine cistern are also discussed.

CASE REPORT

A 50-year-old female with a 5-year history of left HFS was admitted to the Department of Neurosurgery, Kitasato University Hospital, on May 15, 2007. The spasm was initially confined to the superior orbicularis oculi muscle, but gradually affected the entire left side of her face. She had been treated with botulinum toxin three times before admission. The effectiveness of the treatment was temporary. Her clinical and familial histories were non-contributory.

General physical examination revealed no abnormalities. Preoperative neurological examination revealed no abnormal findings other than tapping noise heard in her left ear. Abnormal synkinesis between the orbicularis oculi and the orbicularis oris muscles was recorded by electromyographic examination of the blink reflex. Preoperative magnetic resonance (MR) image demonstrated no offending artery around the facial nerve at the nerve's root exit zone (REZ). MR angiography revered a tortuous anterior inferior cerebellar artery (AICA) suspected to be an offending artery. Finally, basiparallel anatomic scanning (BPAS) MR image (9, 10) revealed detailed anatomic relationship between the facial-vestibulocochlear nerve complex and AICA. The facial nerve seemed to arise from the brain stem near the lateral end of the pontomedullary sulcus away from the point at which the vestibulocochlear nerve joined the brain stem at the lateral end of the sulcus. The basilar artery gave rise to the left AICA whose premeatal segment was adhered to the distal portion of the REZ of the seventh nerve (Figures. 1A,B,C,D).

Microvascular decompression (MVD) of the left seventh cranial nerve was performed via a lateral suboccipital infrafloccular approach (3) under general anesthesia on May 21, 2007. Auditory brain-stem evoked responses (ABRs) were monitored throughout the surgery. In addition, abnormal muscle responses (AMRs), which were evoked by the innervations of other branches of the facial nerve after stimulating of one branch of the facial nerve in the patient, were also monitored. Electrical stimulation of a branch of the facial nerve caused AMRs of the facial muscles. The facial-vestibulocochlear nerves were supposed to be making a complex formation as they usual arise together in the superior olivary fossete. However, the facial nerve arose more than 5 mm away from the vestibular nerve in the brain stem, as shown on the BPAS MR image, and both traveled apart toward the internal acoustic meatus in the cerebello-pontine cistern (Figures. 2A,B,C). The REZ of the facial nerve was clearly visualized. The posterior cerebellar

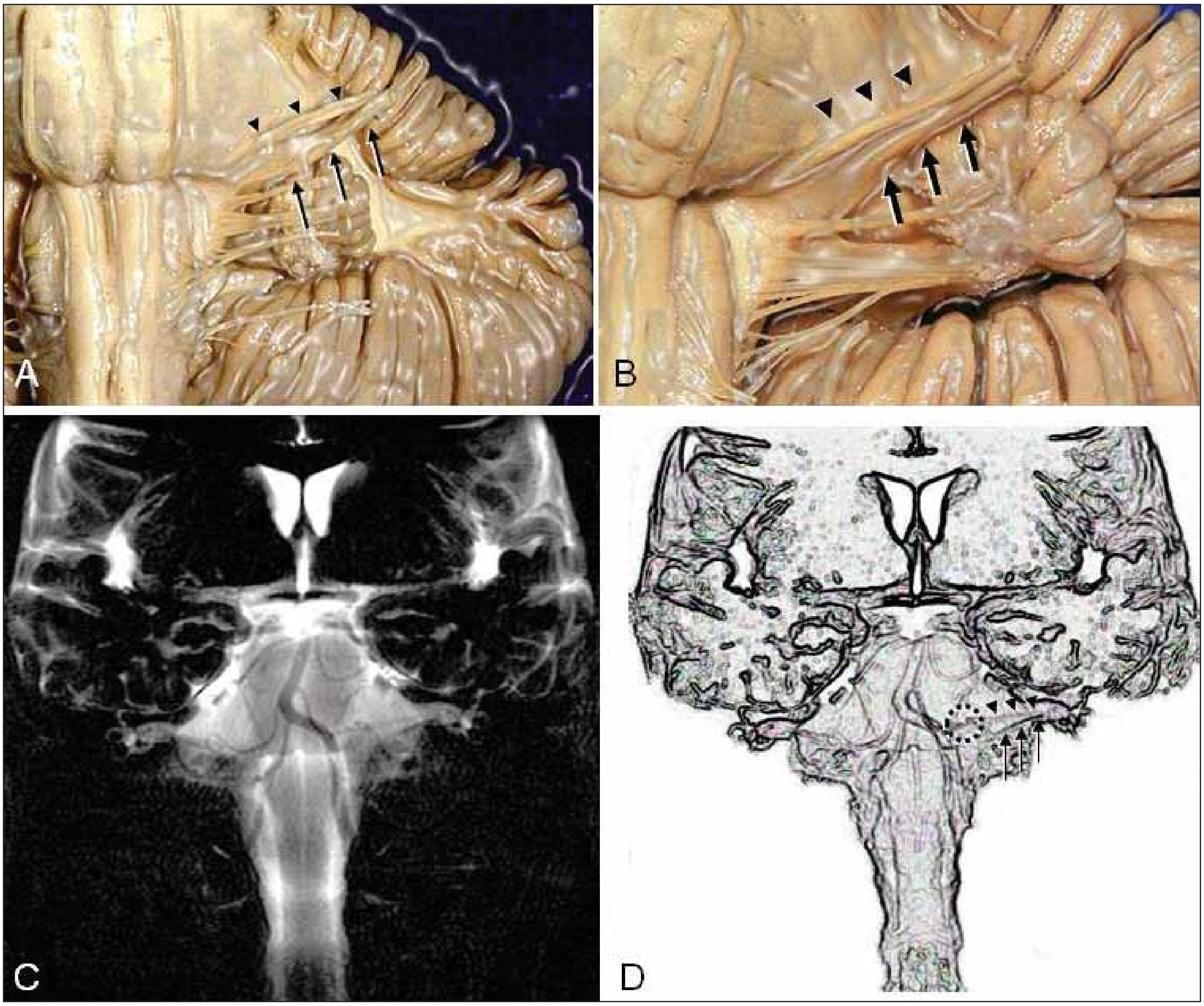

***Figure 1:** Cadaveric specimen of the petrous surface of the left cerebellum (**A** and **B**), basiparallel anatomic scanning (BPAS) MR image of the anterior aspect of the brain stem in this case (**C**) and outline of the BPAS MR image (**D**). (**A** and **B**) The facial (arrow heads) and vestibulocochlear (arrows) nerves arise just rostral to the foramen of Luschka near the flocculus at the lateral end of the pontomedullary sulcus 1 to 2 mm anterior to the point at which the vestibulocochlear nerve joins the brain stem at the lateral end of the sulcus. The interval between the facial and vestibulocochlear nerves is the greatest at the level of the pontomedullary sulcus and decreases as these nerves approach the meatus. (**C** and **D**) The BPAS MR image reveals detailed anatomic relationship between facial-vestibular nerves complex and AICA. The facial nerve (arrow heads) arises from the brainstem more than 4 mm away from the point at which the vestibulocochlear nerve (arrows) joined the brainstem. The basilar artery gave rise to the left AICA whose premeatal segment was adhered to the distal portion of the REZ of the seventh nerve (dotted circle).*
CN: cranial nerve

artery (PICA) coursed adjacent to the REZ of the facial nerve, but was not in contact with the caudal aspect of the REZ. No other offending vessels were observed near the REZ (Figure 2D). Although the PICA was not suspected to be the offending artery, the PICA was separated from the REZ of the facial nerve without manipulation of the nerve just in case. Small pieces of sponge were placed between the PICA and the pons. When the PICA was separated from the REZ of the facial nerve, the AMRs of the mentalis muscle did not disappear. The surgeons noted that the AICA penetrated between the facial and vestibulocochlear nerves, and the meatal segment of the AICA compressed and bended the cisternal segment of the facial nerve. The AICA was carefully detached from the facial and vestibular nerves. A small piece of sponge was placed between the premeatal segment of the AICA and brain stem

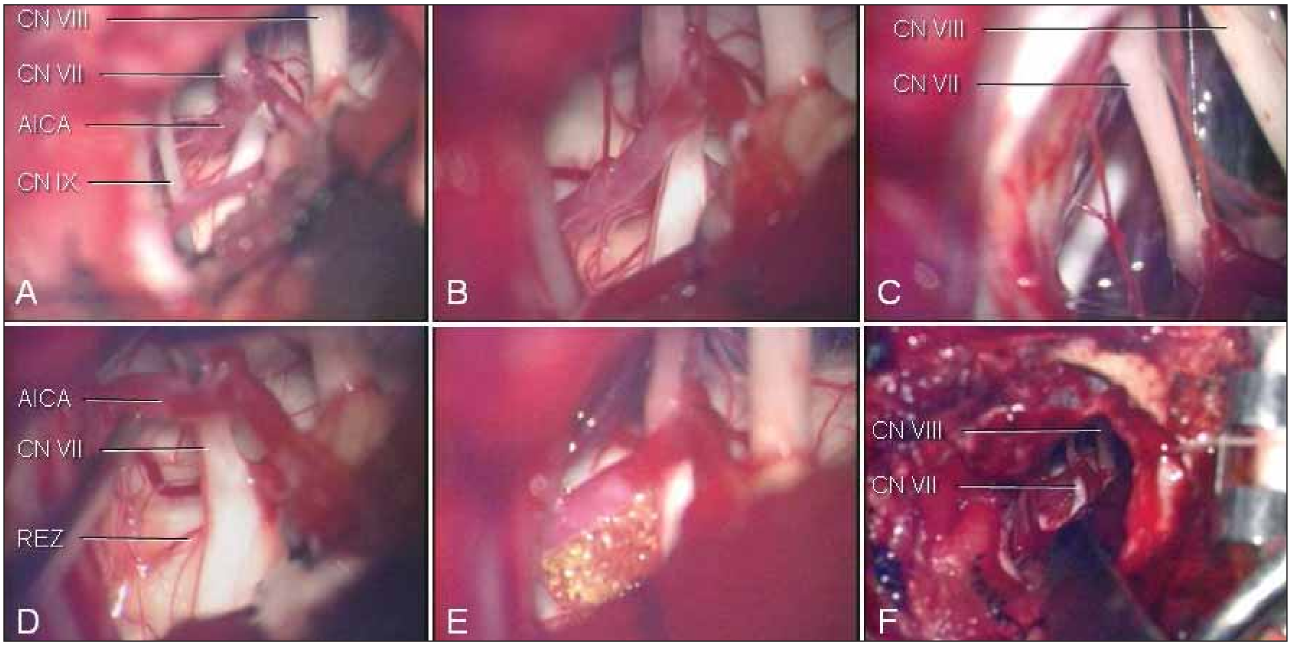

Figure 2: *Intraoperative photographs of the left microvascular decompression. (**A** and **B**) The facial nerve arose more than 5 mm away from the vestibulocochlear nerve in the brain stem. (**C**) Both nerves traveled apart toward the internal acoustic meatus in the cerebello-pontine cistern. (**D**) No offending vessel was observed near the REZ of the facial nerve. (**E**) The AICA penetrated between the facial and the vestibular nerves, and the meatal segment of the AICA compressed and bended the cisternal segment of the facial nerve. A small piece of sponge was placed between the premeatal segment of the AICA and brain stem without manipulation of the facial nerve. (**F**) Finally, the sponge helped to straighten the bent facial nerve.*
CN: cranial nerve

without manipulation of the facial nerve (Figure 2E). Finally, the sponge helped to straighten the bended facial nerve (Figure 2F). During dissection of the AICA, the onset latency of the AMRs became variable (Figure 3-1). When the AICA was partly separated from the facial nerve, the AMRs of the mentalis muscle temporarily disappeared (Figure 3-2). Finally, the AMRs disappeared completely after the sponge was placed between the premeatal segment of the AICA and brain stem (Figure 3-3). The ABRs were normally recorded during the surgery.

The HFS was completely resolved immediately following the surgery. The postoperative course was uneventful with no signs of facial paresis or any new deficits. Postoperative MR angiography showed a small piece of sponge placed between the premeatal segment of the left AICA and brain stem (Fig. 4A). Postoperative axial MR image of the left cerebello-pontine angle demonstrated the changed course of the facial nerve that traveled more closely to the vestibulocochlear nerves postoperatively (Figs. 4B and C). There has been no recurrence of HFS in an 11-month follow-up period.

DISCUSSION

Jannetta et al. in 1977 clearly referred to vascular compression at the REZ of the facial nerve as the cause of HFS for the first time (4). The REZ of the facial nerve is the location at which the central oligodendroglia that comprise the myelin sheath of each axon of the facial nerve in the brain stem are replaced by peripheral Schwann cells and is most easily irritated by mechanical stimulation such as vascular compression (15). Nowadays, neurosurgeons perform MVD at the REZ of the facial nerve to treat HFS, often ignoring vascular compression of this nerve at other sites. Several authors, however, reported patients with HFS caused by vascular compression of the distal portion of the facial nerve (1,12,14,17). One of our authors and his co-workers previously examined the cisternal portion of the facial nerve and its contact arteries in 20 autopsied adult patients (6). They report that more than two thirds of the facial nerves were attached at two points: the root exit zone and the distal cisternal portion.

The mechanism of HFS caused by distal vascular compression is unknown. Partial demyelination and

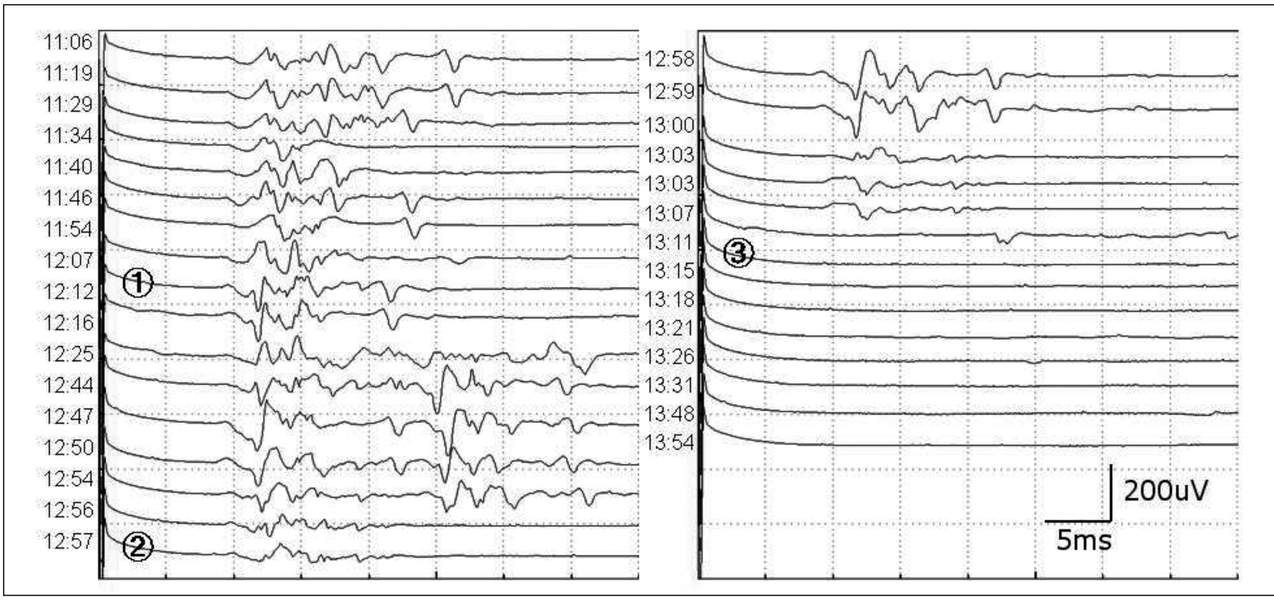

Figure 3: Monitored abnormal muscle response (AMR). During dissection of the AICA, the onset latency of the AMRs became variable (1). When the AICA was partly separated from the facial nerve, the AMRs of the mentalis muscle temporarily disappeared (2). Finally, a small piece of sponge was placed between the premeatal segment of the AICA and the brain stem, and the AMRs disappeared (3).

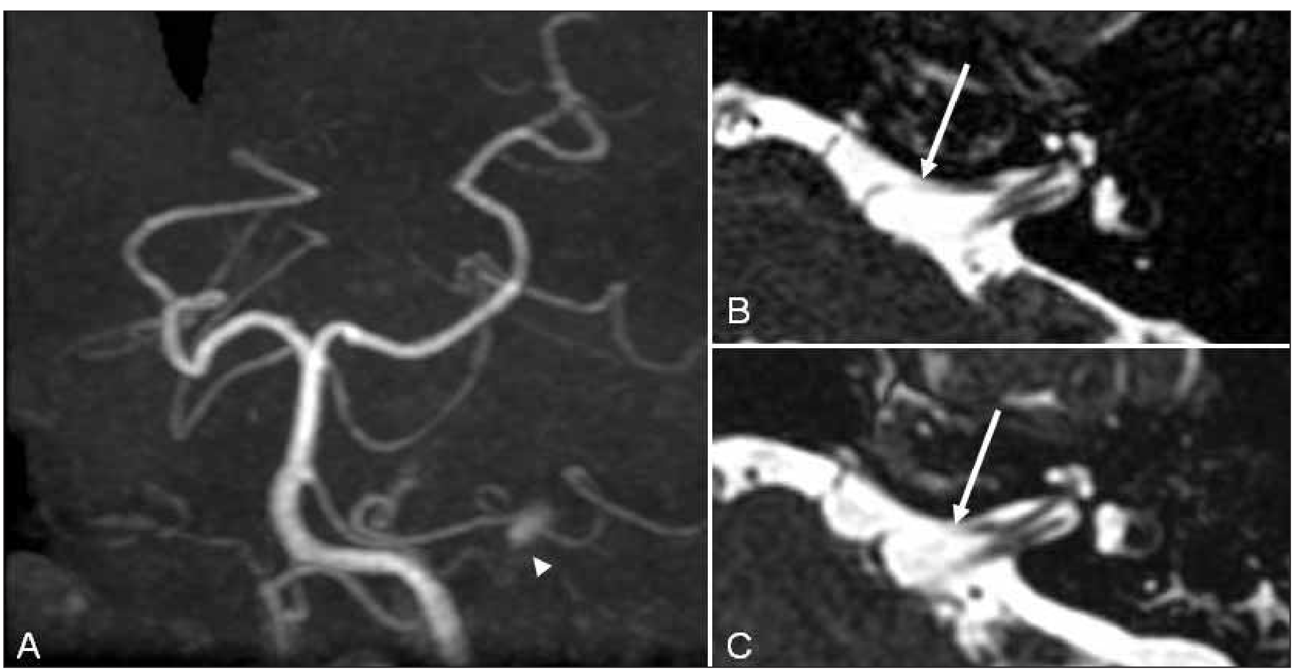

Figure 4: (A) Postoperative MR angiography showing a small piece of sponge placed between the premeatal segment of the left AICA and the brain stem (arrow head). Preoperative (B) and postoperative (C) axial MR images of the left cerebello-pontine angle demonstrating the changed course of the facial nerve (arrows). The facial nerve travels more closely to the vestibulocochlear nerve postoperatively.

axonal degeneration, which are associated with the hyperactivity of the facial motor nucleus that causes HFS, may occur due to compression of the distal portion of the facial nerve (14). On the other hand, Nagahiro et al. (11) reported that the results of surgery in patients with the meatal branch of the AICA coursing between the seventh and eighth cranial nerves and causing compression of the dorsal aspect of the seventh cranial nerve (sandwich-type compression) were poor with operative failure and recurrence of HFS. According to their report, there were seven patients in whom the meatal branch of the AICA coursed the seventh and eighth cranial nerves and compressed the dorsal aspect of the seventh cranial nerve; this was usually associated with another artery compressing the ventral aspect of the nerve. Of these patients, five (71%) had poor results including operative failure in one and recurrence of spasm in four. In our case, the meatal branch of the AICA coursed the seventh and eighth cranial nerves and compressed the dorsal aspect of the seventh cranial nerve, but ventral arterial compression at the REZ was not observed. It is important, however, to ascertain that the AICA is the offending vessel in most cases of compression of the distal portion of the facial nerve (70-100%), whether it penetrates between the facial and vestibular nerves or not (1,12,14).

The facial nerve arises from the brainstem near the lateral end of the pontomedullary sulcus 1 to 2 mm anterior to the point at which the vestibulocochlear nerve joins the brain stem at the lateral end of the sulcus (13). The interval between the facial and vestibulocochlear nerves is the greatest at the level of the pontomedullary sulcus and decreases as these nerves approach the meatus. The lateral meatus can be considered to be divided into four portions forming transverse and vertical crests, with the facial nerve being anterosuperior, the cochlear nerve anteroinferior, the superior vestibular nerve posterosuperior, and the inferior vestibular nerve posteroinferior. The interval between the facial and vestibulocochlear nerves in the present case was more than 5 mm at the brain-stem and more than 4 mm in the cistern. Although detailed information related to the interval between the facial and vestibulocochlear nerves at the level of the brain stem and the cistern has been rarely mentioned in the literature, this is probably one of the greatest ever reported.

The connection between the vascular and neural structures must be evaluated prior to HFS surgery. New magnetic resonance sequences and three-dimensional (3D) imaging methods, such as the 3D constructive interference of steady state and 3D fast imaging with steady state acquisition (3D FIESTA), have recently been developed (2,16,18). In the present study, the relationship between the facial and vestibulocochlear nerves and vascular structures including the vertebral artery, AICA, and PICA were evaluated using FIESTA sequences prior to the surgery, but the understanding of their precise 3D relationship was difficult. The basiparallel anatomic scanning (BPAS)-MR imaging technique is the modification of the surface anatomic scanning-MR imaging technique to reveal the surface appearance of the intracranial vertebrobasilar artery (9,10). It requires only a 2-cm-thick, heavily T2-weighted coronal image, parallel to the clivus. Moreover, BPAS-MR imaging can also clearly show the relationship between the facial-vestibulocochlear nerve complex and vertebrobasilar arteries including its branches. The three dimensional relationship of the facial-vestibulocochlear nerve complex, vertebrobasilar artery, AICA, and the PICA were precisely evaluated preoperatively and applied to the surgery in the present case.

Recording electromyographic responses during MVD for the treatment of HFS is useful for identifying the offending vessel causing the spasm. Absence of AMRs at the end of the MVD apparently guarantees a high probability of relief from HFS (5, 7, 8). In our case, monitoring of AMRs during the surgery was helpful in identifying the meatal segment of the AICA as the cause of the HFS. AMRs disappeared immediately after decompression of the meatal segment of AICA.

In conclusion, we saw a patient with HFS caused by vascular compression of the distal portion of the facial nerve associated with configuration variation of the facial-vestibulocochlear nerve complex. An HFS patient with not only with the AICA compressing the distal portion of the facial nerve without arterial compression of the REZ, but also a configuration variation of the facial-vestibulocochlear nerve complex has not been reported previously. The facial nerve was strongly bended posteriorly, without forming a nerve complex with the vestibulocochlear nerve, which might be associated with partial demyelination and axonal degeneration in its cisternal segment

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