

Case Report

A Case of Hemifacial Spasm Caused by an Artery Passing Through the Facial Nerve

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Hemifacial spasm (HFS) is a clinical syndrome characterized by unilateral facial nerve dysfunction. The usual cause involves vascular compression of the seventh cranial nerve, but compression by an artery passing through the facial nerve is very unusual. A 20-year-old man presented with left facial spasm that had persisted for 4 years. Compression of the left facial nerve root exit zone by the anterior inferior cerebellar artery (AICA) was revealed on magnetic resonance angiography. During microvascular decompression surgery, penetration of the distal portion of the facial nerve root exit zone by the AICA was observed. At the penetrating site, the artery was found to have compressed the facial nerve and to be immobilized. The penetrated seventh cranial nerve was longitudinally split about 2 mm. The compressing artery was moved away from the penetrating site and the decompression was secured by inserting Teflon at the operative site. Although the facial spasm disappeared in the immediate postoperative period, the patient continued to show moderate facial weakness. At postoperative 12 months, the facial weakness had improved to a mild degree. Prior to performing microvascular decompression of HFS, surgeons should be aware of a possibility for rare complex anatomy, such as compression by an artery passing through the facial nerve, which cannot be observed by modern imaging techniques.

Key Words : Hemifacial spasm · Penetrating artery · Facial nerve · Facial weakness.

INTRODUCTION

Hemifacial spasm (HFS) is a clinical syndrome characterized by unilateral facial nerve dysfunction, such as intermittent painless, involuntary and spasmodic contractions of muscles. HFS typically begins as spontaneous paroxysms of eye twitching, and progresses to involve other muscles innervated by the facial nerve²⁷). In 1970, Jannetta et al. popularized the concept of compression of the facial nerve at its exit from the brain stem by one or more arteries or veins as the most common etiology of HFS, and favored microvascular decompression as an effective treatment^{19,20}). The most common arteries involved in HFS, in order of most frequent observation, include the anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA) and vertebral arteries. Other less frequent etiologies have been described to cause HFS including aneurysm, arteriovenous malformation, tumor, ectatic vertebrobasilar arteries and Paget's disease of the skull. However, in review of the literature, we were unable to find any report of HFS associated with an artery

that had passed through the facial nerve. Therefore, we report a case of HFS associated with penetration of the facial nerve by the AICA.

CASE REPORT

A 20-year-old male patient presented with a 4-year history of progressive HFS beginning with left orbicularis oris spasms, often triggered by eating or drinking. He described the spasms as progressing from the mouth to the eye and eventually involving the platysma, which is unusual in typical HFS. The patient's hearing was normal and he exhibited no cranial nerve palsy or neurological deficits.

Concentric needle electromyography examination of the left orbicularis oculi and orbicularis oris revealed irregular bursting discharge at rest and normal motor unit potential on volition. Also, evidence of synkinesis in the orbicularis oculi and mentalis muscle was observed during supraorbital nerve stimulation. Magnetic resonance imaging (MRI) and magnetic reso-

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nance angiography (MRA) revealed a compression of the left facial nerve root exit zone by the AICA (Fig. 1).

Based on the above, microvascular decompression was selected for treatment. During the operation, penetration of the distal portion of the facial nerve root exit zone by the AICA was observed. At the penetrating site, the artery was found to have compressed the facial nerve and to be immobilized. After discussing the risk of facial paresis with the patient's family, the penetrated seventh cranial nerve was longitudinally split about 2 mm. The compressing artery was moved away from the penetrating site and the decompression was secured by inserting Teflon at the operative site (Fig. 2).

Although the facial spasm disappeared in the immediate postoperative period, the patient continued to show moderate facial weakness. At postoperative 4 months, the facial weakness had improved to a mild degree (Fig. 3).

DISCUSSION

HFS is a benign condition, characterized by unilateral paroxysmal chaotic and clonic spasms of the orbicularis oculi, paranasal, perioral and platysma muscles^{41,45}. Most cases of HFS are due to a vascular pathology that compresses the exiting nerve root of nerve VII^{7,35}. In a previous study, Han et al.¹⁵ reviewed 1642 cases of HFS to evaluate unusual possible causes and clinical presentations, and reported that 1633 cases (99.4%) involved neurovascular compression. The other unusual causes were a secondary causative structural lesion (0.5%), a tumor (0.4%) and a vascular malformation (0.1%)¹⁵.

Upon literature review, the most common causes of HFS, besides neurovascular compression, were cerebellopontine angle tumors (epidermoid, arachnoid cyst, lipoma, and vestibular

schwannoma)^{2,3,9,15,26,38}, and other unusual causes including cerebellopontine angle medullary venous malformations^{15,24}, Paget's disease^{16,25}, occipital falcine meningioma⁵, cerebellopontine angle meningioma¹¹, acoustic schwannoma¹¹, pontine glioma⁴⁵, fourth ventricle ganglioglioma⁶, pontine infarction⁴³, syringobulbia⁴, multiple sclerosis⁴², trauma²⁹, hypothyroidism¹³, idiopathic intracranial hypertension³⁷, vertebrobasilar ectasia²¹, craniovertebral anomalies²⁸, glomus jugular tumor²³, parotid gland^{8,11,12,32}, and arterial hypertension³³. A few cases of HFS alone or in combination with trigeminal neuralgia have been reported as a false localizing sign in patients who had a contralateral posterior fossa mass or acoustic neuroma^{30,31}. This present case involved an unusual cause of HFS, and there have been no reports of HFS caused by penetration of the nerve by an artery.

From an embryological point of view, the beginning of the facial nerve, the facioacoustic primordium, appears during the third week of gestation. The facial nerve then splits into two parts at the end of the fourth week and is complete by the fifth to sixth gestational week³⁶. The AICA appears later, in the fourth to fifth month of gestation, when all definitive communications of the facial nerve are established^{22,34}. Accordingly, we surmised that the penetration of the facial nerve may have happened as the AICA attempted to pass through the split in the facial nerve that appears at the fifth to sixth gestational week.

The introduction of MRI and MRA has improved the detection of neurovascular conflicts (vascular contact and/or compression of the root exit zone of the facial nerve)^{14,39}. High-resolution MRI and MRA techniques highly sensitive for neurovascular conflict are frequently needed^{14,39}. These imaging studies generally show great sensitivity in detecting unusual causes of HFS; however, in the present case, such imaging studies failed to reveal the penetration of the nerve by the AICA, as shown in Fig. 1.

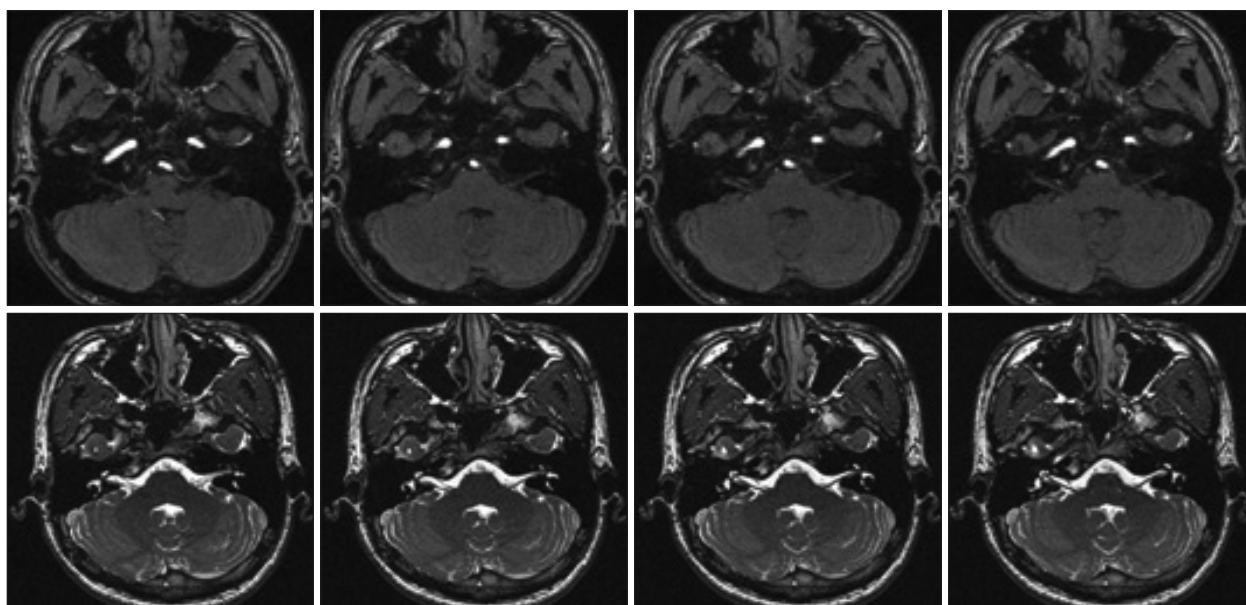


Fig. 1. Preoperative magnetic resonance angiography. Compression of the left facial nerve root exit zone by the anterior inferior cerebellar artery is observed.



Fig. 2. Intraoperative findings. Penetration of the distal portion of the facial nerve root exit zone by the anterior inferior cerebellar artery is revealed (arrow). After longitudinally splitting the penetrated nerve, the compressing artery was moved away from the penetrating site and the decompression was secured by insertion of Teflon.

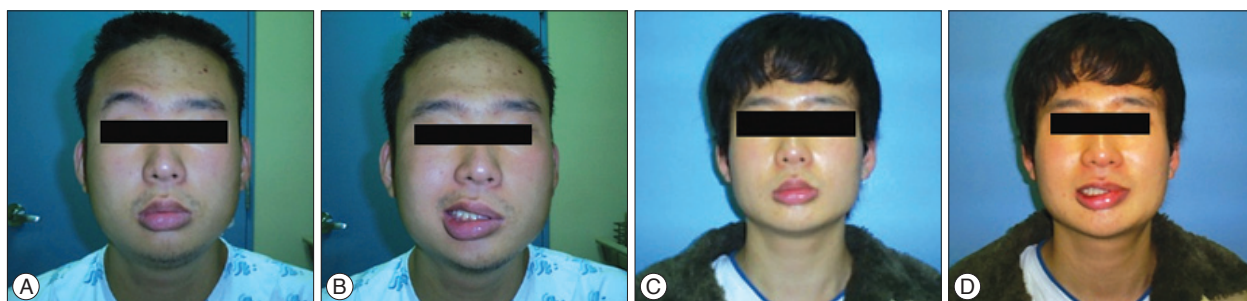


Fig. 3. Postoperative photographs. The patient showed moderate facial weakness although the facial spasm disappeared in the immediate postoperative period (A and B). After 4 months of follow-up (C and D), the facial weakness had improved to a mild degree, from House and Brackmann grade 4 to 2.

Patients with HFS usually present between 40 and 50 years of age^{10,40,44}. In most cases of HFS, the initial site of onset is the orbicularis oculi muscle, although a few cases have reported the cheek and the perioral region as the initial site of onset^{10,18,40}. Over months to years, the spasms spread gradually to other muscles innervated by the ipsilateral facial nerve in a synchronous manner. However, as we described above, this case was unusual in that the onset and clinical presentation of HFS did not follow the normal progression of symptoms, as the spasms progressed from the mouth to the eye, and eventually involved the platysma. Such unusual clinical presentations could indicate unusual causes of HFS^{2,11,17}.

In this case, although the penetration of the distal portion of the facial nerve root exit zone by the AICA was longitudinally incised after discussing the risk of facial paresis with the patient's family, it is important to consider which is more beneficial to leave without facial spasm or leave without facial weakness. The authors considered that the importance for each person could be different. The compressing artery was moved away from the penetrating site and the decompression was secured by inserting Teflon at the operative site. Although the facial spasm disappeared in the immediate postoperative period, the patient continued to show moderate facial weakness after 4 months of follow-up. In retrospect, transposition of the vessel structure to alleviate compression of the nerve may have been an alternative option, instead of the longitudinal incision of the facial nerve we performed. However, this would not be an option through

which to completely relieve the facial spasm, but rather to partially relieve with the preserve of facial weakness.

CONCLUSION

Prior to performing microvascular decompression of HFS, surgeons should be aware of a possibility for rare complex anatomy, such as compression by an artery passing through the facial nerve, which cannot be observed by modern imaging techniques.

References

1. Adler CH, Zimmerman RA, Savino PJ, Bernardi B, Bosley TM, Sergott RC : Hemifacial spasm : evaluation by magnetic resonance imaging and magnetic resonance tomographic angiography. *Ann Neurol* 32 : 502-506, 1992
2. Altinörs N, Kars Z, Cepoğlu C : Rare causes of hemifacial spasm. Report of two cases. *Clin Neurol Neurosurg* 93 : 155-158, 1991
3. Auger RG, Piepgras DG : Hemifacial spasm associated with epidermoid tumors of the cerebellopontine angle. *Neurology* 39 : 577-580, 1989
4. Barraquer-Bordas, Zamora S, Abello-Vila P : [Hemifacial spasm and bilateral central deafness in the clinical picture of possible syringobulbia]. *Rev Esp Otoneurooftalmol Neurocir* 14 : 17-22, 1955
5. Bhayani R, Goel A : Occipital falcine meningioma presenting with ipsilateral hemifacial spasm : a case report. *Br J Neurosurg* 10 : 603-605, 1996
6. Bills DC, Hanieh A : Hemifacial spasm in an infant due to fourth ventricular ganglioglioma. Case report. *J Neurosurg* 75 : 134-137, 1991
7. Coakham HB : The microvascular treatment of trigeminal neuralgia,

- hemifacial spasm and glossopharyngeal neuralgia in Robertson JT, Coakham HB, Robertson JH (eds) : **Cranial base surgery**. New York : Churchill Livingstone, 2000, pp543-564
8. Destee A, Bouchez B, Pellegrin P, Warot P : Hemifacial spasm associated with a mixed benign parotid tumour. **J Neurol Neurosurg Psychiatry** 48 : 189-190, 1985
 9. Digre K, Corbett JJ : Hemifacial spasm : differential diagnosis, mechanism, and treatment. **Adv Neurol** 49 : 151-176, 1988
 10. Ehni G, Woltman HW : Hemifacial spasm : review of one hundred and six cases. **Arch Neurol Psychiatry** 53 : 205-211, 1945
 11. Gálvez-Jiménez N, Hanson MR, Desai M : Unusual causes of hemifacial spasm. **Semin Neurol** 21 : 75-83, 2001
 12. Gandon J, Trotoux J, Peynègre R, André J, Brasnu D : [Study of a series of 158 parotidectomies and histological problems in mixed tumors of salivary glands (author's transl)]. **Ann Otolaryngol Chir Cervicofac** 96 : 261-280, 1979
 13. Genis D, Ricart W, Fernández-Real JM, Dávalos A, Molins A, Ferrándiz M : Hemifacial spasm and hypothyroidism. **Lancet** 342 : 1112, 1993
 14. Girard N, Poncet M, Caces F, Tallon Y, Chays A, Martin-Bouyer P, et al. : Three-dimensional MRI of hemifacial spasm with surgical correlation. **Neuroradiology** 39 : 46-51, 1997
 15. Han IB, Chang JH, Chang JW, Huh R, Chung SS : Unusual causes and presentations of hemifacial spasm. **Neurosurgery** 65 : 130-137; discussion 137, 2009
 16. Ing EB, Savino PJ, Bosley TM, Sergott RC, Kelepouris N : Hemifacial spasm and osteitis deformans. **Am J Ophthalmol** 119 : 376-377, 1995
 17. Inoue T, Maeyama R, Ogawa H : Hemifacial spasm resulting from cerebellopontine angle lipoma : case report. **Neurosurgery** 36 : 846-850, 1995
 18. Jannetta PJ : **Cranial Rhizopathies : Neurological Surgery**, ed 3. Philadelphia : W.B. Saunders, 1990, pp4169-4182
 19. Jannetta PJ : Microsurgical exploration and decompression of the facial nerve in hemifacial spasm. **Cuff Topics Surg Res** 2 : 217-220, 1970
 20. Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS : Etiology and definitive microsurgical treatment of hemifacial spasm. Operative techniques and results in 47 patients. **J Neurosurg** 47 : 321-328, 1977
 21. Kerber CW, Margolis MT, Newton TH : Tortuous vertebrbasilar system : a cause of cranial nerve signs. **Neuroradiology** 4 : 74-77, 1972
 22. Kier EL : Development of cerebral vessels in Newton TH, Potts DG (eds) : **Radiology of the skull and brain**. St. Louis : Mosby, 1974, pp1089-1141
 23. Kiley MA, Voyvodic F, Burns RJ : An unusual cause of hemifacial spasm. **J Clin Neurosci** 6 : 349-351, 1999
 24. Kim Y, Tanaka A, Kimura M, Yoshinaga S, Tomonaga M : Arteriovenous malformation in the cerebellopontine angle presenting as hemifacial spasm--case report. **Neurol Med Chir (Tokyo)** 31 : 109-112, 1991
 25. Linazasoro G, Martí Massó JF : Paget's disease and hemifacial spasm. **Neurology** 42 : 1643-1644, 1992
 26. Loeser JD, Chen J : Hemifacial spasm : treatment by microsurgical facial nerve decompression. **Neurosurgery** 13 : 141-146, 1983
 27. Maroon JC : Hemifacial spasm. A vascular cause. **Arch Neurol** 35 : 481-483, 1978
 28. Maroun FB, Jacob JC, Weir BK, Mangan MA : Hemifacial spasm and craniovertebral anomaly. **Can J Neurol Sci** 17 : 424-426, 1990
 29. Martinelli P, Giuliani S, Ippoliti M : Hemifacial spasm due to peripheral injury of facial nerve : a nuclear syndrome? **Mov Disord** 7 : 181-184, 1992
 30. Matsuura N, Kondo A : Trigeminal neuralgia and hemifacial spasm as false localizing signs in patients with a contralateral mass of the posterior cranial fossa. Report of three cases. **J Neurosurg** 84 : 1067-1071, 1996
 31. Nishi T, Matsukado Y, Nagahiro S, Fukushima M, Koga K : Hemifacial spasm due to contralateral acoustic neuroma : case report. **Neurology** 37 : 339-342, 1987
 32. Nussbaum M : Hemifacial spasm associated with benign parotid tumor. **Ann Otol Rhinol Laryngol** 86 (1 Pt 1) : 73-74, 1977
 33. Oliveira LD, Cardoso F, Vargas AP : Hemifacial spasm and arterial hypertension. **Mov Disord** 14 : 832-835, 1999
 34. Osborn AG : **Diagnostic cerebral angiography**, ed 2. Philadelphia : Lippincott Williams & Wilkins Publishers, 1999
 35. Rapanà A, Guida F, Conti C, Rizzo G, Trincia G : Ependymoma of the fourth ventricle presenting with hemifacial spasm. Report of a case. **Rev Neurol (Paris)** 155 : 309-311, 1999
 36. Sataloff RT : **Embryology and anomalies of the facial nerve and their surgical implications**. New York : Raven Press, 1991
 37. Selky AK, Purvin VA : Hemifacial spasm. An unusual manifestation of idiopathic intracranial hypertension. **J Neuroophthalmol** 14 : 196-198, 1994
 38. Singh AK, Jain VK, Chhabra DK, Hongo K, Kobayashi S : Hemifacial spasm and cerebellopontine angle epidermoid : case report and review. **Neurol Res** 16 : 321-323, 1994
 39. Tan EK, Chan LL, Lim SH, Lim WE, Khoo JB, Tan KP : Role of magnetic resonance imaging and magnetic resonance angiography in patients with hemifacial spasm. **Ann Acad Med Singapore** 28 : 169-173, 1999
 40. Tan EK, Jankovic J : Psychogenic hemifacial spasm. **J Neuropsychiatry Clin Neurosci** 13 : 380-384, 2001
 41. Tan NC, Chan LL, Tan EK : Hemifacial spasm and involuntary facial movements. **QJM** 95 : 493-500, 2002
 42. van de Biezenbos JB, Horstink MW, van de Vlasakker CJ, van Engelen BG, van Eikema Hommes OR, Barkhof F : A case of bilateral alternating hemifacial spasms. **Mov Disord** 7 : 68-70, 1992
 43. Vermersch P, Petit H, Marion MH, Montagne B : Hemifacial spasm due to pontine infarction. **J Neurol Neurosurg Psychiatry** 54 : 1018, 1991
 44. Wang A, Jankovic J : Hemifacial spasm : clinical findings and treatment. **Muscle Nerve** 21 : 1740-1747, 1998
 45. Westra I, Drummond GT : Occult pontine glioma in a patient with hemifacial spasm. **Can J Ophthalmol** 26 : 148-151, 1991