

Case Report

Resolution of Isolated Unilateral Hypoglossal Nerve Palsy Following Microvascular Decompression of the Intracranial Vertebral Artery

Jin Hwan Cheong, M.D., Ph.D., Jae Min Kim, M.D., Ph.D., Moon Sul Yang, M.D., Choong Hyun Kim, M.D., Ph.D.

Department of Neurosurgery, Hanyang University College of Medicine, Guri Hospital, Guri, Korea

Isolated hypoglossal nerve paresis due to mechanical compression from a vascular lesion is very rare. We present a case of a 32-year-old man who presented with spontaneous abrupt-onset dysarthria, swallowing difficulty and left-sided tongue atrophy. Brain computed tomographic angiography and magnetic resonance imaging of the brainstem demonstrated an abnormal course of the left vertebral artery compressing the medulla oblongata at the exit zone of the hypoglossal rootlets that was relieved by microvascular decompression of the offending intracranial vertebral artery. This case supports the hypothesis that hypoglossal nerve palsy can be due to nerve stretching and compression by a pulsating normal vertebral artery. Microvascular decompression of the intracranial nerve and careful evaluation of the imaging studies can resolve unexpected isolated hypoglossal nerve palsy.

Key Words : Hypoglossal nerve paresis · Microvascular decompression · Vertebral artery.

INTRODUCTION

Hypoglossal nerve palsy (HNP) is a fairly common finding in neurological disease. However, isolated HNP due to mechanical compression from a vascular lesion is very rare. We present a case of isolated unilateral HNP that occurred spontaneously and was relieved by microvascular decompression of the offending intracranial vertebral artery (VA).

CASE REPORT

A 32-year-old man presented with spontaneous abrupt onset of dysarthria as well as difficulty in moving his tongue and swallowing food. This was preceded by a month-long history of progressive swallowing discomfort and feeling generally unwell. There was no history of neck trauma, manipulation, or vigorous exercise. He had no significant past medical history. The results of the general physical examination of the patient were unremarkable. He showed normal pharyngeal sensation and a brisk bilateral gag reflex. His tongue deviated to the left side on protrusion

(Fig. 1). Neurological examination revealed only left hypoglossal nerve palsy with ipsilateral atrophy without signs of other cranial nerve palsies, pyramidal signs, or Horner syndrome. Cerebellar function was normal, and no symptoms and signs of brain stem compression or increased intracranial pressure were observed. Laboratory examination, including inflammatory markers and CSF studies, were performed. Normal full blood count, erythrocyte sedimentation rate, and C-reactive protein ruled out an infectious cause. Negative viral serology confirmed these findings. General immunology and autoimmunity tests excluded an autoimmune cause. Renal function tests, liver function tests, and random blood glucose were all normal. Cerebro-



Fig. 1. Preoperative photograph of the patient showing deviation of the tongue to the left before surgery.

• Received : May 31, 2010 • Revised : July 13, 2010

• Accepted : February 6, 2011

• Address for reprints : Jae Min Kim, M.D., Ph.D.

Department of Neurosurgery, Hanyang University College of Medicine,

Guri Hospital, 249-1 Gyomun-dong, Guri 471-701, Korea

Tel : +82-31-560-2323, Fax : +82-31-560-2327

E-mail : kjm2323@hanyang.ac.kr

spinal fluid (CSF) analysis and culture were performed to rule out intracranial pathology. These were unremarkable. Computed tomography (CT) of the brain and the base of the skull was unremarkable. Magnetic resonance imaging (MRI) scans demonstrated the left VA to be more prominent than the right VA and closely approximated to the lateral aspect of the medulla (Fig. 2). There was no evidence of brainstem infarction and spaceoccupying lesion on MRI. We believed that there was a high probability that the VA was responsible for the twelfth nerve palsy and surgical exploration was performed. We performed retromastoid craniotomy in the lateral position by a far-lateral approach. The left intradural vertebral artery and the twelfth nerve were exposed.

The VA was confirmed as normal and the twelfth nerve was seen to be stretched and compressed around the VA. The trunk of the hypoglossal nerve was stretched and extremely thin. A Teflon felt was inserted between the hypoglossal nerve and the left VA (Fig. 3). At follow up 3 month later, further improvement in tongue movement was observed (Fig. 4).

DISCUSSION

The hypoglossal nerve is a pure motor nerve that innervates intrinsic as well as extrinsic muscles of the tongue. It can be divided

into five segments : medullary (nuclear), cisternal (extramedullary intracranial), skull base (the segment which passes through the hypoglossal canal), nasopharyngeal/oropharyngeal carotid (in close vicinity to the glossopharyngeal and vagus nerves as well as to the internal carotid artery) and sublingual (where its branches terminate innervating lingual muscles)¹⁾.

The hypoglossal nerve can be damaged anywhere along its course. Supranuclear disease affecting the nerve results in paralysis of the tongue contralateral to the side of the lesion. Deviation of the tongue will occur away from the side of the lesion. Fasciculation and atrophy of the tongue are absent. When disease affects the hypoglossal nerve at the nuclear or infranuclear level, the clinical signs and symptoms are ipsilateral. There is deviation of the tongue toward the side of the lesion, with associated atrophy of the intrinsic and extrinsic tongue musculature and fasciculation of the tongue^{1,12)}.

The hypoglossal nucleus in medullary segment may be affected by a variety of disorders, including medullary infarction, hemorrhage, neoplasms, and multiple sclerosis. Neoplasms may be primary (most often glioma) or secondary. Less common diseases may be involved, such as syringobulbia, poliomyelitis, botulism, and amyotrophic lateral sclerosis^{1,12)}. Lying in the premedullary cistern, the rootlets of the hypoglossal nerve may be affected by a variety of pathological processes. The vertebral arteries close to the nerve rootlets, which may be compressed by a vertebral aneurysm or by dolichoectasia. Direct extension of neoplasm of the skull base may lead to nerve dysfunction^{1,12)}.

Tumors, both benign and malignant, and trauma may damage the hypoglossal nerve as it traverses the skull base. Basal skull fractures may extend to involve the hypoglossal canal or the occipital condyle, resulting in an isolated or complex palsy of the hypoglossal nerve. Tumors of the skull base may affect the hypoglossal canal by expansion or destruction. Metastatic tumors and prima-

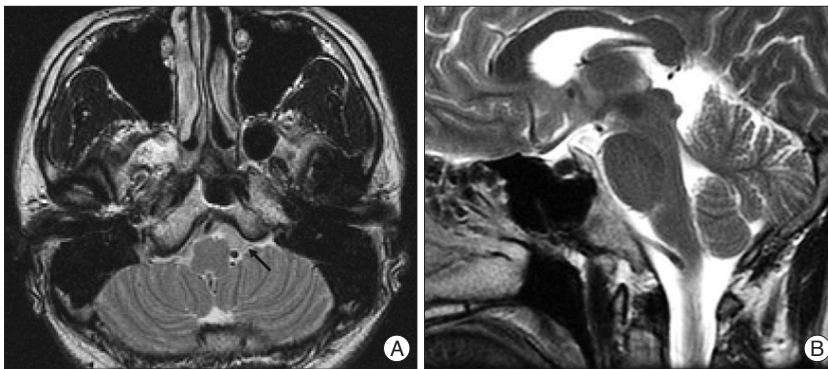


Fig. 2. Preoperative magnetic resonance (MR) images. Preoperative T2-weighted axial (A) and sagittal (B) MR images demonstrating medial deviation of the left vertebral artery, which attaches to the left lower lateral medulla and compresses of the root exit zone of the left hypoglossal nerve (arrow).

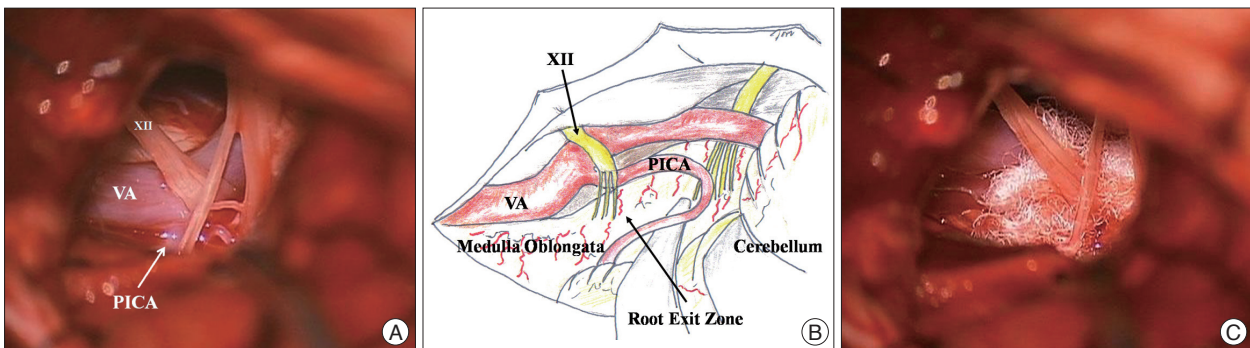


Fig. 3. Intraoperative photographs. A, B : Intraoperative photograph and schematic drawing showing compression of the root exit zone of the left hypoglossal nerve (long arrow) at the vertebral artery-posterior inferior cerebellar artery junction. C : Teflon felt was interposed between the pulsating vertebral artery and the hypoglossal nerve via a far lateral suboccipital approach.



Fig. 4. Three-month postoperative photograph of the patient demonstrating marked improvement of deviation.

ry nasopharyngeal squamous cell carcinoma may produce erosion of the skull base and dysfunction of the hypoglossal nerve^{1,12}. Hypoglossal nerve injury within the carotid space and sublingual space may result from a range of disorders, malignant disease being the most common^{1,12}. In a retrospective analysis of 100 patients with HNP, Keane found that tumors-predominantly malignant-were the most common cause of the HNP, and other causes included trauma, medullary infarction, hysteria, multiple sclerosis, Guillain-Barré neuropathy and infection⁵.

Impairment of function of cranial nerves in the form of either paresis or neuralgia following compression by tortuous and dilated intracranial vessels is a well established entity^{6,9}. Facial nerve palsy and glossopharyngeal neuralgia are the most common, with impairment of the fifth, eighth, and tenth nerves less frequently described. The close relationship to an adjacent cerebral vessel that compresses the nerve against the bone in the cerebellopontine angle explains this high rate of involvement. Moon et al.⁷ reported that downbeat nystagmus (DBN), occurring due to compression of the lower brainstem by a dolichoectatic vertebral artery, disappeared after microvascular decompression with Teflon.

The cranial nerve dysfunction syndrome occurs because the nerve is particularly vulnerable to continuous pulsatile pressure at the root exit zone, which is the transition area between the peripheral and central myelin in the brainstem.

Shiozawa reported a case of HNP due to venous compression of the hypoglossal nerve by an enlarged emissary vein inside the hypoglossal canal¹⁰. In this case the hypoglossal nerve compression was determined by a space occupying venous lesion but the enlarged vein does not apply a pulsatile pressure on the nerve.

The hypoglossal nerve is a closely adjacent structure to both the carotid and vertebral arteries and has been described in the setting of various vascular pathologies. Entrapment of peripheral nerve fibers has been associated with kinking of both the carotid and vertebral arteries^{3,9}. Morini et al.⁸ have reported two cases of isolated HNP due to an anomalous dolichovertbral artery course compressing the medulla oblongata at the emergence of the hypoglossal rootlets. Other authors have described

isolated unilateral HNP caused by the expanding hematoma of a dissecting carotid artery^{2,4,11}. However, in our case, the offending vessel was a normal artery, the site of contact was the root exit zone, the vertebral artery crossed the nerve perpendicularly, and the nerve was stretching and compressed by the pulsating normal vertebral artery similar to hemifacial spasm or trigeminal neuralgia. Isolated twelfth nerve palsy due to a vessel compression mechanism has rarely been demonstrated to our knowledge and this possibility is not usually included in the differential diagnosis of this condition.

MRI has a crucial role in the diagnostic approach. MR angiography, CT angiography, or digital subtraction angiography (DSA) could also be good additional tests. We suggest that MRI focusing on the hypoglossal nerve pathway should be obtained to localize the lesion in the event of suspected isolated HNP.

The symptoms of intracranial nerve paresis due to vessel compression usually improve or disappear spontaneously or after medical treatment, but unexpected isolated hypoglossal nerve palsy may require surgical treatment with microvascular decompression.

CONCLUSION

This case supports the notion that, in any case of unexplained isolated hypoglossal nerve palsy, nerve stretching and compression from a pulsating normal VA should be considered. Careful evaluation of imaging studies and microvascular decompression of the intracranial nerve can resolve the unexpected isolated hypoglossal nerve palsy.

Acknowledgements

The authors thank professor Jae-Hoon Kim, a neurosurgeon of Eulji University, Nowon Eulji Hospital for his exquisite drawing of intraoperative findings.

References

1. Boban M, Brinar VV, Habek M, Rados M : Isolated hypoglossal nerve palsy : a diagnostic challenge. *Eur Neurol* 58 : 177-181, 2007
2. Boukobza M, Ast G, Reizine D, Merland JJ : Internal carotid artery dissection causes hypoglossal nerve palsy : CT, MRI, and angiographic findings. *J Neuroimaging* 8 : 244-246, 1998
3. Carney AL, Anderson EM : Hypoglossal carotid entrapment syndrome. *Adv Neurol* 30 : 223-247, 1981
4. Goodman JM, Zink WL, Cooper DF : Hemilingual paralysis caused by spontaneous carotid artery dissection. *Arch Neurol* 40 : 653-654, 1983
5. Keane JR : Twelfth-nerve palsy. Analysis of 100 cases. *Arch Neurol* 53 : 561-566, 1996
6. Kerber CW, Margolis MT, Newton TH : Tortuous vertebrobasilar system : a cause of cranial nerve signs. *Neuroradiology* 4 : 74-77, 1972
7. Moon KH, Lee SA, Ahn JS, Kwun BD : Downbeat nystagmus associated with brainstem compression by vertebral artery. *J Korean Neurosurg Soc* 41 : 190-192, 2007
8. Morini A, Rozza L, Manera V, Buganza M, Tranquillini E, Orrico D : Isolated hypoglossal nerve palsy due to an anomalous vertebral artery course : report of two cases. *Ital J Neurol Sci* 19 : 379-382, 1998
9. Rollnik JD, Sindern E, Mosler F, im Spring B, Malin JP : Isolated peripheral hypoglossal palsy caused by a kinking of the left vertebral artery (hy-

- poglossal vertebral entrapment syndrome). *Eur Neurol* 36 : 324-325, 1996
10. Shiozawa Z, Koike G, Seguchi K, Shindo K, Sugita K : Unilateral tongue atrophy due to an enlarged emissary vein in the hypoglossal canal. *Surg Neurol* 45 : 477-479, 1996
 11. Sturzenegger M, Huber P : Cranial nerve palsies in spontaneous carotid artery dissection. *J Neurol Neurosurg Psychiatry* 56 : 1191-1199, 1993
 12. Thompson EO, Smoker WR : Hypoglossal nerve palsy : a segmental approach. *Radiographics* 14 : 939-958, 1994