Case Report

Isolated hypoglossal nerve neuropathy in vertebral dolichoectasia: Microvascular decompression by vessel transposition with Teflon cuff

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ABSTRACT

Background: A clinical case of isolated unilateral hypoglossal nerve (HN) neuropathy, which spontaneously occurred from vertebral artery dolichoectasia and was cured by a new method of microvascular decompression by transposition of the vertebral artery using the Teflon cuff.

Case Description: A young patient with an anamnesis of the disease for more than 4 years and complaints of a deviation of the tongue to the right and dysarthria was examined. MRI of the brain revealed compression of the medulla oblongata by an elongated, dilated, and deformed right vertebral artery. Compression of the medulla oblongata and HN was confirmed during surgery. A transposition of the vertebral artery was performed using a Teflon cuff in the ventral direction to the clivus. Three months after surgery, positive dynamics was noted in the form of regression of dysarthria and improvement of mobility and trophic language.

Conclusion: Thus, isolated HN neuropathy as a result of compression by an elongated, dilated, and deformed vertebral artery is a rare neurological disease that can be successfully treated by transposition using a Teflon cuff.

Keywords: Hypoglossal nerve, Microvascular decompression, Neuropathy, Transposition, Vertebral dolichoectasia

INTRODUCTION

The hypoglossal nerve (HN) provides motor innervation to the muscles of the tongue, as well as the genioglossus, styloglossus, and hypoglossus muscles. The nucleus of the HN is located in the lower parts of the medulla oblongata, near the floor of the fourth ventricle. Axons run ventrally to the anterolateral sulcus between the pyramid and the inferior olive and exit as part of a group of roots, connecting into 2 (80%) or 3 trunks (20%).

Nerve damage disrupts the balancing action of the genioglossus muscle, which deviates the tongue in the opposite direction. Therefore, the neuropathy of the HN (NHN) leads to the classic triad of chewing difficulties, dysphagia, and dysarthria.

HN has five segments: the medullary, cisternal, skull bases, carotid space, and sublingual segment. The intradural V3 segment of the vertebral artery lies in the premedullary cistern in close proximity to the HN. In addition to the intracranial region, there are two more areas of intersection of the sublingual nerve with large vessels, where vasoneural conflict is possible.
NHN is a rare disease[^8] and is mainly combined with palsy of other cranial nerves or damage to the brainstem. The differential diagnosis of NHN includes many different conditions of neoplastic, infectious, degenerative genesis, trauma, and other diseases.[^12] NHN is often the result of radiation therapy, surgical procedures, or anesthesia.[^14] The literature describes cases of nerve damage at the medullary segment, when the vertebral artery causes compression of the medulla oblongata and, accordingly, the motor nuclei of the XII nerve.[^14,20,26,28]

In the cisternal segment, HN damage can be caused by contact with the V4 segment of the vertebral artery[^1,17,23,24] and its dissection,[^13,15,16] thrombosis, kinking[^21] and abnormal course,[^18] as well as a consequence of an infectious process.[^10]

Tumors are the most common cause of NHN and can affect every segment of the nerve, but most commonly the one passing through the canal of the HN at the base of the skull. In the segment of the skull base, NHN can occur due to craniocebral trauma with a fracture of the condyle of the occipital bone. Isolated NHN due to mechanical compression by vascular structures may occur due to compression in the carotid space, aneurysms[^4] or dissections[^6] of the internal carotid artery. NHN can also be caused by such a rare neoplasm as schwannoma of the HN.[^3]
Table 1 contains works describing neurovascular conflict of the HN in the medullary or cisternal segment. In most cases, the tactics chosen were dynamic monitoring and/or medical treatment. There are a few cases describing microvascular decompression of the HN by interposition of the vertebral artery using a Teflon⁵ and one work provides details of performing vertebral artery transposition in five patients.⁶⁷

**CASE DESCRIPTION**

A 28-year-old man in full health condition spontaneously developed weakness of the right side of the tongue with its deviation to the right, 4 years later dysarthria appeared. At the debut of the disease, the patient noted fascial twitching (fasciation?) of half of his tongue. The patient had no history of neck trauma, infectious, and autoimmune diseases. He was treated as an outpatient under the care of a dentist. Results of physical examination of the patient were unremarkable. Neurological examination revealed only peripheral neuropathy of the right side of the tongue with significant atrophy with no signs of deficits on the part of other cranial nerves. A brisk bilateral gag reflex was detected. Cerebellar function was normal, with no symptoms or signs of brainstem compression or increased intracranial pressure. General blood count and C-reactive protein levels excluded an infectious cause. MRI revealed a dolichoectasia of the right VA, which severely compressed the medulla oblongata and roots (XII cranial nerve?) exiting the trunk at the level of the deformity [Figure 1]. The depth of the premedullary cistern was 9 mm according to MRI data, which was considered to be enough distance to move the artery. We believed that there was a high probability that it was the vertebral artery that was the “causal factor” for the neuropathy of the HN at the medullary segment. As the patient showed negative dynamics with the development of dysarthria, surgical intervention was suggested to prevent the progression of neurological deficit.

We performed a right-sided “enough” lateral retrosigmoid craniotomy in the position of the patient on the left side [Figure 2]. The V3 segment of the vertebral artery, caudal cranial nerves, cerebellum and medulla oblongata are visualized. The vertebral artery was characterized by stiffness in the segment of interest, formed an elastic nondisplaced C-shaped deformity, and compressed the brain stem. At attempts to displace it from the ventral surface of the medulla oblongata, the artery returned to its previous position, while no significant compression of the HN roots by the artery was detected; therefore, it was concluded that NHN was caused by nuclear compression.

Given the rigidity of the causal vessel in the area of conflict and the predicted low efficiency of interposition, we performed transposition of the vertebral artery by forming a cuff of Teflon material and moving the artery anteriorly by fixing the cuff in the anteromedial direction to the dura mater of the posterior surface of the clivus with single sutures. During transposition, to reduce the stiffness of the main artery, temporary clipping of the vertebral artery was used for 7 min [Figures 3 and 4, Video 1]. After arterial transposition, control intraoperative Doppler and intraoperative fluorescence angiography using ICG were performed: no signs of blood flow abnormalities were detected. In addition, interposition was performed by placing a Teflon fiber gasket between the medulla oblongata and the right VA.

In the 1st day after the surgery, the patient underwent MRI of the brain, visualized the displaced vertebral artery without signs of brainstem compression, the right vertebral artery and other arteries of the posterior cranial fossa contrasted homogeneously, no arterial narrowing or deformities were
observed [Figure 5]. In the 1st week after the operation, the patient noted improvement in the neurological status in the form of dysarthria regression. He was discharged from the hospital on the 7th day after the operation.

At the follow-up examination after 3 months, the atrophy of the tongue muscles had not regressed, but the patient noted an improvement in the mobility of the tongue [Figure 6].

**CONCLUSION**

Classical methods of interposition using Teflon material in microvascular decompression are mainly used in cases of neurovascular conflicts with small caliber arteries. By definition, this method is of poor efficacy in cases of conflicts caused by large arteries such as the vertebral and basilar arteries, especially in their dolichoectasia and sclerotization. The large diameter of the arteries, thicker, rigid or elastic walls, and high blood pressure in the lumen of these vessels make them less mobile, difficult to shift, and prone to recoiling.

The Teflon gaskets are sometimes not reliable enough to hold and fix such vessels in the optimal position. Therefore, microvascular decompression in cases caused by large caliber arteries is always a difficult technical problem. There are various techniques of using a combination of Teflon (and other synthetic materials) strips and fibrin-thrombin glue,[19] transposition due to hemostatic material,[11] and many modifications of this technique using other materials. There is also developed technique of arterial transposition with the use of vascular microclips.[6]

The technique of large artery transposition used in this clinical case does not require additional instrumental and material costs and does not disturb the normal anatomy. The advantages of the technique include reliable arterial fixation, and its universality allows its use for neurovascular conflicts on different floors of the posterior cranial fossa.

This clinical case demonstrates the possibility of diagnostic isolated neuropathy of the HN through a detailed clinical and instrumental examination. Compression of the HN at the medullary segment, a rare neurological disease that can be successfully treated by transposition of the vertebral artery using a Teflon cuff.
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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ahmed SN, Aladdin Y, Siddiqi ZA, Khan K. Hypoglossal-vertebral entrapment syndrome. Neurology 2008;71:461.
2. Bademci G, Batay F, Yaşargil MG. “Triple cross” of the hypoglossal nerve and its microsurgical impact to entrapment disorders. Minim Invasive Neurosurg 2006;49:234-7.
3. Bindal S, El Ahmadieh T, Plitt A, Aoun SG, Neeley OJ, El Tecle NE, et al. Hypoglossal schwannomas: A systematic review of the literature. J Clin Neurosci 2019;62:162-73.
4. Cano-Duran AJ, Sanchez Reyes JM, Sevilla MT, Yucumá D. Carotid petrous segment aneurysm presenting as hypoglossal nerve palsy. Neuroradiology 2021;63:447-50.
5. Cheong JH, Kim JM, Yang MS, Kim GH. Resolution of isolated unilateral hypoglossal nerve palsy following microvascular decompression of the intracranial vertebral artery. J Korean Neurosurg Soc 2011;49:167-70.
6. Choudhri O, Connolly ID, Lawton MT. Macrovascular decompression of the brainstem and cranial nerves: Evolution of an anteromedial vertebobasilar artery transposition technique. Neurosurgery 2017;81:367-76.
7. Gibo H, Marinkovic S, Nikodijevz I, Stimec B, Erden A. The blood supply of the hypoglossal nerve: The microsurgical anatomy of ots cisternal segment. Surg Neurol 1997;48:85-91.
8. Graham RM, Thomson EF, Baldwin AJ. Isolated hypoglossal nerve palsy due to a vascular anomaly. Int J Oral Maxillofac Surg 2007;36:759-61.
9. Hafkamp HC, Manni JJ, van der Goten A. Unilateral spontaneous dissection of the internal carotid artery presenting as hypoglossal nerve palsy. Eur Arch Otorhinolaryngol 2004;261:405-8.
10. He J, Chun J, Lam L, Adlan T. Clival osseousitis and hypoglossal nerve palsy-rare complications of Lemierre’s syndrome. BMJ Case Rep 2015;2015:bcr2015209777.
11. Ichikawa T, Agari T, Kurozumi K, Maruo T, Satoh T, Date I. “Double-stick tape” technique for transposition of an offending vessel in microvascular decompression: Technical case report. Neurosurgery 2011;68 Suppl 2:377-82.
12. Keane JR. Twelfth-nerve palsy analysis of 100 cases. Arch Neurol 1996;53:561-6.
13. Kesserwani H. Isolated palsy of the cisternal segment of the hypoglossal nerve due to arterial dissection of the V4 segment of the vertebral artery: A case report with a side note on nerve trunk ischemia. Cureus 2020;12:e9930.
14. Kollmann P, Rauq C, Fransen P. Isolated hypoglossal nerve paralysis and hypoglossal vertebral entrapment syndrome. Acta Neurol Belg 2017;117:377-80.
15. Mahadevappa K, Chacko T, Nair AK. Isolated hypoglossal nerve palsy due to vertebral artery dissection. Clin Med Res 2012;10:127-30.
16. McKeon A, Murphy S, McNamara B, Ryder DO, Galvin RJ. Isolated hypoglossal nerve palsy due to compression by a dissecting vertebral artery. Eur Neurol 2005;53:162-4.
17. Meila D, Wetter A, Brassel F, Nacimiento W. Intermittent hypoglossal nerve palsy caused by a calcified persistent hypoglossal artery: An uncommon neurovascular compression syndrome. J Neurol Sci 2012;323:248-9.
18. 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. 1998;19:379-82.
19. Nonaka Y, Hayashi N, Matsumae M, Fukushima T. Wedge-technique for transposition of the vertebral artery in microvascular decompression for hemifacial spasm: Technical nuances and surgical outcomes. Acta Neurochirurgirum 2019;161:1435-42.
20. Ren J, Sun H, Diao Y, Niu X, Wang H, Wei Z, Yuan F. Successful treatment with microvascular decompression surgery of a patient with hemiparesis caused by vertebral artery compression of the medulla oblongata: Case report and review of the literature. World Neurosurg 2017;108:994.e11-9.
21. Rollnik JD, Sindern E, Mosler F, Im Spring B, Malin JP. Isolated peripheral hypoglossal palsy caused by a kinking of the left vertebral artery (hypoglossal vertebral entrapment syndrome). Eur Neurol 1996;36:324-5.
22. Sai Kiran NA, Sivaraju L, Furtado SV, Vidyasagar K, Raj V, Aryan S, et al. Far lateral approach without occipital condylar resection for intradural ventral/ventrolateral foramen magnum tumors and aneurysms of V4 segment of vertebral artery: Review of surgical results. Clin Neurol Neurosurg 2020;197:106163.
23. Salvi F, Mascalchi M, Plasmati R, Tognoli V, De Grandis D. Hypoglossal vertebral entrapment syndrome. Muscle Nerve 1999;22:288-9.
24. Straube A, Linn J. Unilateral headache attacks and ipsilateral atrophy of the tongue due to neurovascular compression of the hypoglossal nerve. Cephalalgia 2008;28:996-9.
25. Thompson EO, Smoker WR. Hypoglossal nerve palsy: A segmental approach. Radiographics 1994;14:939-58.
26. Toldo I, Manara R, Sartori S, Suppiej A, Drigo P. Unilateral hypoglossal nerve palsy due to neurovascular conflict in a child. Brain Dev 2009;31:461-4.
27. Tomasello F, Alafaci C, Salpietro FM, Longo M. Bulbar compression by an ectatic vertebral artery: A novel neurovascular construct relieved by microsurgical decompression. Neurosurgery 2005;56 Suppl 1:117-24.
28. Yamamoto M, Suzuki K, Takekawa H, Hirata K. Isolated hypoglossal nerve palsy caused by neurovascular compression. Intern Med 2011;50:2701-2.