Case Report

Medullary Compression Due to Ectatic Vertebral Artery—Case Report and Review of Literature

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Medullary compression syndrome due to anomalous course of blood vessels is a rare disease most commonly seen in the adult population. The offending vessels causing this syndrome are mostly posterior inferior cerebellar artery or the vertebral artery. The symptoms of this syndrome vary from most common hypertension to various other neurologic deficits like hemiplegia, dysesthesia, and dysarthria. Intractable dizziness is a rare symptom of this disease. The definite management plan for this disease is microvascular decompression. We present our case of medullary compression syndrome which manifested as intractable dizziness. We describe our experience in the management of this patient as well as present a review of literature of this rare disease.

Key Words: Vertebral artery—ectasia—intractable dizziness—medullary compression syndrome

Introduction

Medullary compression syndrome is a rare manifestation of tumors arising in the region of skull base[1,2] as well as many congenital and acquired bony abnormalities of the craniocervical region.[3,4] However, this condition presenting due to abnormal ectatic blood vessels is rare. The symptoms of this condition vary depending upon the nature of compression. We describe a case of medullary compression due to dolichoectatic vertebral artery which manifested as intracrable dizziness.

Case Report

A 57-year-old man presented with history of intractable dizziness for the last 3 years. He was seen by the otolaryngologist several times in these years. He was given labyrinthine sedatives for the management of his symptoms which gave him only temporary relief. During the course of the disease the symptoms progressed to the extent that he began to have multiple falls. On examination, his higher mental fuctions were normal. He was found to have both downbeat and upbeat nystagmus. He did not complain of oscillopsia. There were no lower cranial nerve palsies. Hearing was normal in both ears. Motor and sensory systems were found to be within normal limits. There were no exaggerated deep tendon reflexes nor any sensory loss over the body. There were no cerebellar signs. Skull and spine were normal. He was investigated with magnetic resonance imaging during which it was revealed that he had an anomalous ectatic vertebral artery which was distorting and grooving the left side of the medulla Figure 1 (A, B, and C). The management plan was to relieve the compression of medulla by macrovascular decompression surgery (Fig 2).

Surgery

He was taken up for surgery in prone position. A far lateral suboccipital craniectomy was done on the left side.

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Received July 10, 2019; revision received September 17, 2019; accepted October 2, 2019.

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1052-3057/$ - see front matter

https://doi.org/10.1016/j.jstrokecerebrovasdis.2019.104460

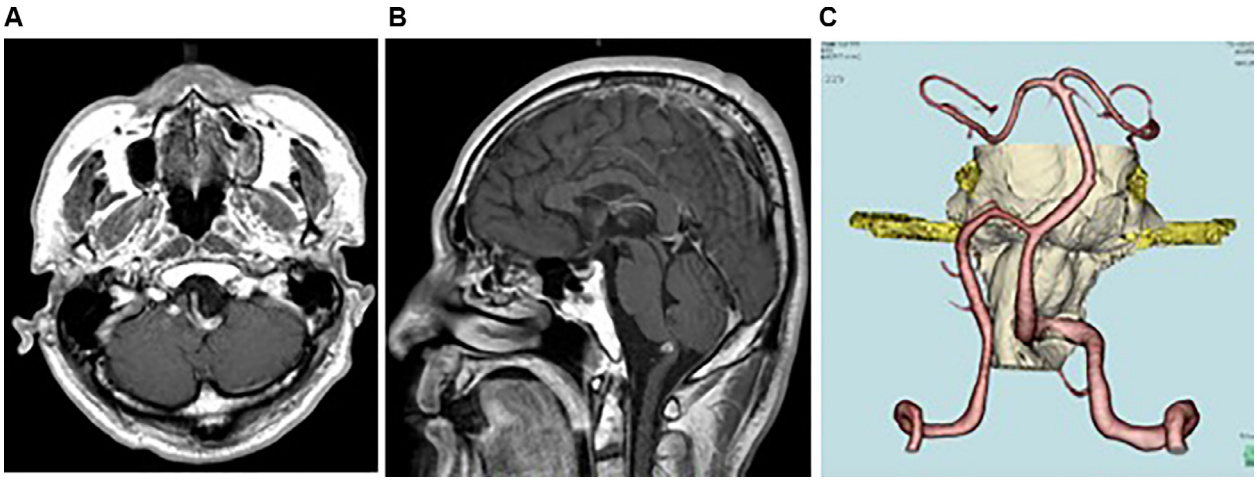

Figure 1. *MRI brain. (A) MRI T1W image axial with contrast showing the deep indentation of the vertebral artery into the medulla. (B) MRI brain T1W sagittal image with contrast showing the kinked medulla by the abnormal course of the vertebral artery. (C) Fusion image of CT angiogram with MRI brain—demonstrating the ectatic vertebral artery grooving the left side of the medulla.*

The tubercle of the condyle was drilled to get a clear view of the lower cranial nerves and vertebral artery. During the dissection it was found that the vertebral artery had an anomalous course after entering the foramen transversarium of first cervical vertebrae as it ascended anteriorly without arching over the groove of atlas and pierced the dura to enter the intracranial subarachnoid space. The dura was opened and the tortuous vertebral artery was found in a groove over the medulla. The artery was elevated off the medulla and transposed to the bony part of the posterior fossa with fibrin glue. The vertebral artery and the medulla were separated by insertion of a Teflon sponge in between them (Fig 2 and Fig 3). The dura was then closed and bone flap was reinserted. The wound was closed in layers. MRI Brain was taken during the Postoperative period which showed thin CSF filled space between the ectatic VA and the brain stem suggestive of good decompression (Fig 4). Symptomatically his dizziness slowly began to improve and he began to walk without difficulty. The downbeat nystagmus which was present preoperatively disappeared following surgery but upbeat nystagmus persisted albeit at a lower amplitude.

Discussion

Medullary compression syndrome (MCS) due to skull base abnormalities or tumors have been long recognized. However, the occurrence of this syndrome owing to the compression by ectatic arteries near the brainstem is uncommon. Dolichoectasia of the intracranial arteries most commonly involve the vertebrobasilar system. The prevalence of this disease is about 4 % and more common in women[5] and elderly population. Ventrolateral medulla is the most common part to be involved in the compression by a dolichoectatic vertebral artery with a propensity to the left side. The etiology of dolichoectasia of the intracranial artery was thought to be due to hypertension but recent evidence suggests that there may be

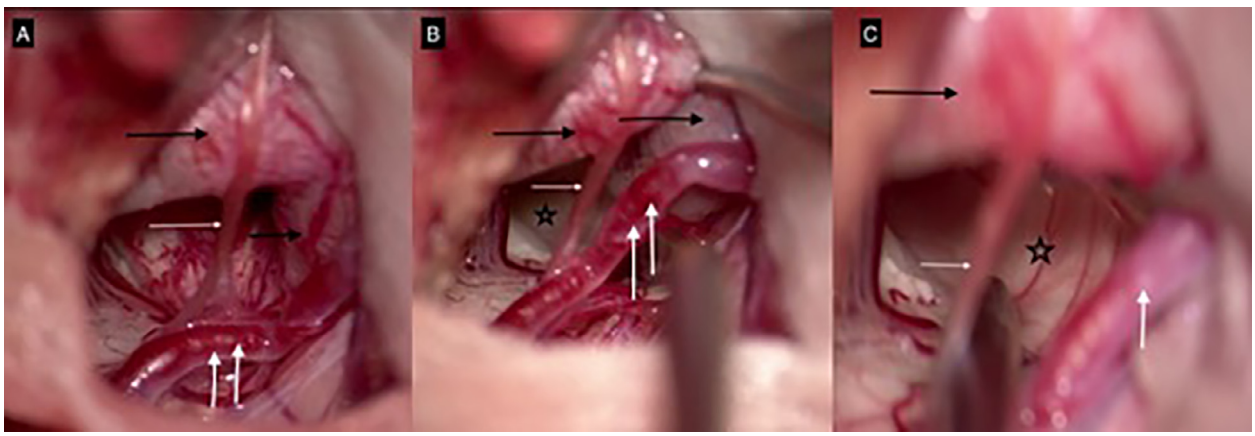

Figure 2. *Intra operative pictures—vertebral artery (VA) in the region of cerebellomedullary fissure. (A) Horizontal black arrows—VA, horizontal white arrow—11th nerve, vertical white arrow—Posterior inferior cerebellar artery (PICA). (B) The VA is lifted off the medulla demonstrating the groove created by it in the medulla (black star). (C) Zoom in view of the groove (black star) of the VA on the medulla.*

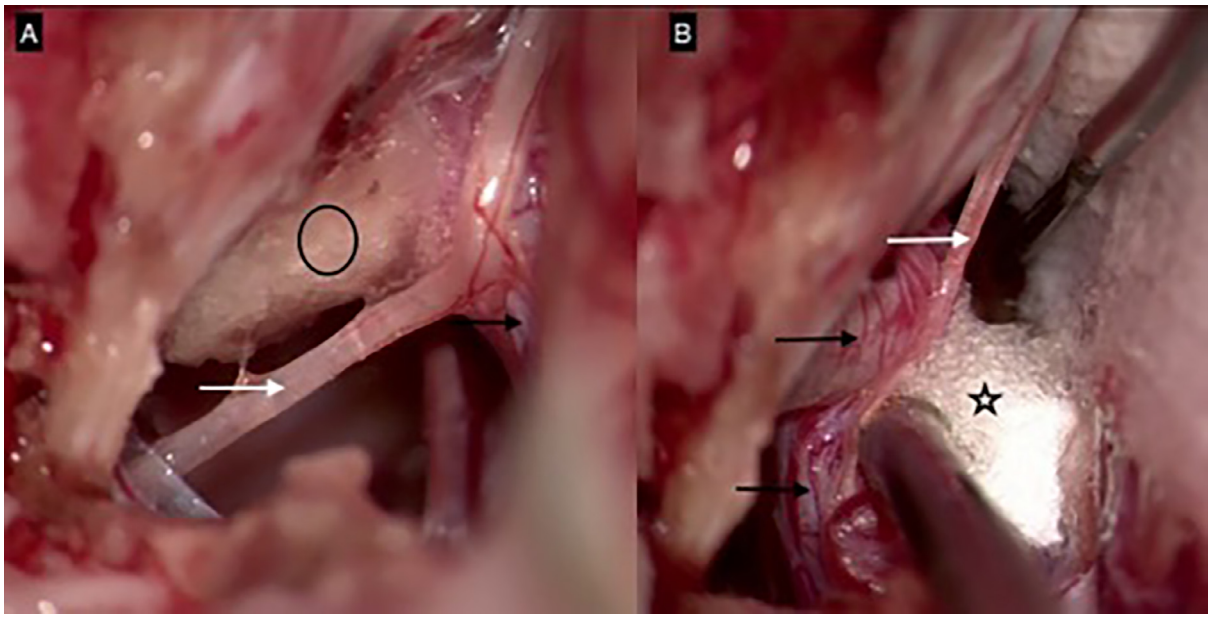

Figure 3. *Intraoperative picture. (A) Transposed vertebral artery to the petrous dura with gelfoam soaked in fibrin glue (black circle). (B) Teflon sponge (black star) being inserted between the Vertebral artery (horizontal arrow) and the medulla.*

an interaction of interlacing factors like congenital and autoimmune mechanisms.[6] Some authors have also postulated vertebrobasilar dissection to be the cause of dolichoectasia.[7] MCS was earliest recognized to be a cause of neurovascular hypertension in 1980 by Fein et al, when they successfully managed 2 cases of intractable hypertension by macrovascular decompression of the root entry zone of vagus nerve compressed by the vertebral artery.[8] In another report, Kim et al, demonstrated resolution of symptoms in a patient with hemiparesis cause by dolichoectatic vertebral artery by transposition and interposition of Teflon in 1985.[9] Following these 2 early descriptions of Vertebral Artery Medullary Compression Syndromes (VAMCS), there have been about 33 reports of MCS reported till now with varying symptomatology (Table 1). The atypical symptoms other than hemiparesis and hypertension ascribed to VAMCS are Cranial neuropathies,[10] palatal myoclonus,[11] diplopia,[12] quadriplegia,[13] episodic dysarthria,[14] respiratory failure, [15] hypoesthesia,[16] and rare variant syndromes like Opalski[17] and Rabbit syndromes.[18] Our patient had long standing dizziness and nystagmus most likely due to the compression of the vestibular nuclei and its connections. The substrates of symptoms responsible for most of described cases lie in the medulla and its connections with the vital brainstem nuclei. In our patient the symptoms could be mostly attributed to the involvement of the vestibular nucleus and its connections with the cerebellum, or central otolithic organs. Nystagmus usually occurs in patients with vestibular disturbances. Many authors have in the past had described patient with downbeat nystagmus with medullary compression syndrome due to aberrant vertebral artery.[19-21] In our patient, inspite of severe compression the symptoms were restricted to involvement of vestibular nucleus without any pyramidal or cerebellar signs. In a largest series of patients with VAMCS, Savitz et al, noted that there was poor correlation between the imaging and clinical findings.[22] The external compression of the brainstem coupled with the deep perforator ischemia has been postulated to be pathogenesis of symptoms in VAMCS. In a study by Passera et al, it was shown that in compression syndromes related to the vertebrobasilar system the symptoms can be subtle and may not be detected clinically. However, they emphasized that signs of early brainstem malfunction may be detected early through electrophysiological studies. Prolongation of blink reflex, abnormalities in motor evoked potentials, and brainstem auditory evoked potentials have been shown to act as a guide in deciding for both intervention as well as follow-up in relatively asymptomatic patients.[23] One Of most interesting symptom that has been extensively described and studied in VAMCS is hypertension and its substrates. Janetta et al, in their baboon model described inducible hypertension by passing a balloon system connected from aorta to left ventrolateral medulla in the region of olive and inflating it in a pulsatile fashion.[24] Further, more evidence surfaced in the postmortem studies of patients with essential hypertension in which Naraghi et al, observed a higher incidence of vascular conflicts in the region of the root entry zone of vagus nerve.[25] Later on, Janetta et al, in their series went on to successfully manage 12 patients with essential hypertension with microvascular decompression. In these series, only 3 had a demonstrable

Table 1. *A summary of articles published till now about vertebral artery compression syndrome and the patient symptoms*

Fein[8] et al (1980)	Hypertension
Kim[9] et.al., (1985)	Hemiparesis
Maruyama[39] et al (1989)	Unsteadiness of gait, numbness of limbs
Kobayashi[40] et al (1992)	Hemiparesis
Himi[19] et.al., (1995)	Downbeat nystagmus
Murata[42] et al (1995)	Progressive tetraparesis, Sensory disturbances in all 4 limbs
Janetta[26] et al (1998)	Hypertension
Levy[30] et al (1998)	Hypertension
Hongo[10] et al (1999)	Hemiparesis, cranial nerve dysfunction
Meyer[11] et al (2000)	Palatal myoclonus
Salvi[31] et al (2000)	Hypertension, hemiparesis
Lee[20] et al (2001)	Downbeat nystagmus
Takano[43] et al (2001)	Mild hemiparesis, headache, and facial hypoesthesia
Ubogu[12] et al (2002)	Diplopia, quadriparesis
Koyama[44] et al (2002)	Quadriparesis, unconsciousness
Ishikawa[32] et al (2004)	Hypertension, diziness
Akimura[33] et al (2005)	Hypertension
Tomasella[45] et al (2005)	Dysphagia, paraparesis
Chan[34] et al (2005)	Hypertension
Haangi[35] et al (2009)	Hypertension, hypoesthesia
Hongo[41] et al (2009)	Hemiparesis and gait ataxia
Haubrich[14] et al (2010)	Episodic dysarthria
Koguchi[36] et al (2011)	Respiratory failure
Dembo[17] et al (2013)	Opalski syndrome
Kamada[46] et al (2013)	Paraparesis
Nakahara[15] et al (2014)	Respiratory failure, dysphagia
Betgeri [13] et al (2015)	Spastic quadriparesis
Park[18] et al (2016)	Rabbit syndrome
Moon[21] et al (2016)	Downbeat nystagmus
Sadashiva[37] et al (2016)	Quadriparesis
Savitz[22] et al (2016)	Hypertension, tinnitus, hoarseness, weakness
Ren[38] et al (2017)	Hemiparesis
Lombarski[16] et al (2018)	Hypoesthesis, hemiparesis

neurovascular conflict on preoperative MRI. Hence, thereafter it was demonstrated that not only large arteries but also smaller arteries in the ventrolateral medulla can induce hypertension.[26] The management of VAMCS can be both surgical as well as medical. In the largest case series by Savitz et al, demonstrated resolution of symptoms with anticoagulants. Only 2 patients in their series underwent surgery. In our patient, labyrinthine sedatives were prescribed for a long time before he came to us. But since his symptoms were progressive, we decided to do a macrovascular decompression. In a study by Grasso et al, it was suggested that during dealing with MCS the offending vessels needed to be

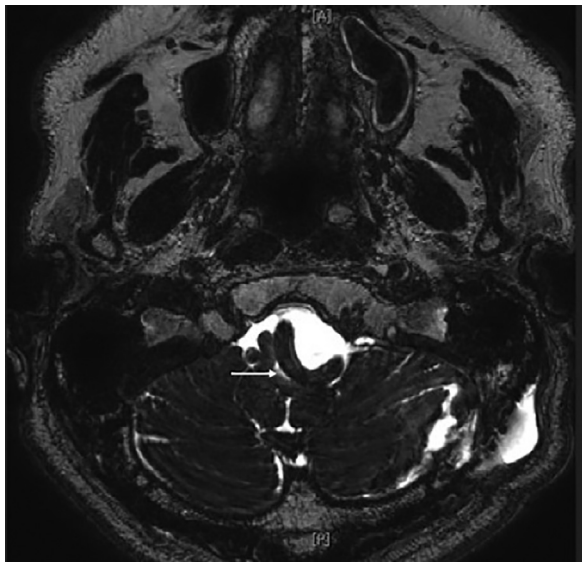

Figure 4. *Post operative MRI brain plain T2W image. Thin CSF filled space (white arrow) between the ecstatic vertebral artery and the brainstem.*

identified accurately. The membranes around the neural and vascular structures should be dissected meticulously. The vessels to be mobilized should be inspected for perforators before attempting mobilization. A separation of at least 5 mm was recommended distance between VA and brainstem as the objective of the surgery in VAMCS.[27] In our patient there were several deep perforators were found on the VA precluding a wide transposition. The decompression was attempted by marginal transposition of the VA to the basal dura with the help of fibrin glue as well as interposition of Teflon graft between the medulla and the vertebral artery. Transposition of vessels with sling surgeries are also a viable option in VAMCS.[28,29] The outcome of macrovascular decompression in cases of VAMCS can be dramatic with appropriate surgery. Marked resolution of symptoms has been reported within a matter of months. This can be well appreciated in the reports of MCS causing respiratory paralysis where patients were weaned off the ventilator and breath normally after medullary decompression.[15] In our review of literature, we noted that among 33 reports nearly 50% managed conservatively had either no change or worsening in their symptom. Whereas those managed surgically, nearly 80% has good resolution of symptoms.

Conclusion

VAMCS is a rare syndrome with varied presentations related to either vestibular, cerebellar or pyramidal systems. Most often clinic-radiological findings may vary. The definitive management is by macrovascular decompression.

Conflict of Interest

Nil.

Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

References

1. Zakaria J, Wemhoff M, Anderson D. Microvascular decompression and arachnoid cyst fenestration for treatment of medulla oblongata compression caused by arachnoid cyst-vascular loop complex. World Neurosurg 2018;113:293-295.
2. Gelabert-Gonzalez M, Reyes-Santias RM, Garcia-Pravos A. Dorsal medullary compression due to a primary vertebral chondrosarcoma. Rev Neurol 2000;30:322-324.
3. Gladi M, Iacoangeli M, Specchia N, et al. Endoscopic transnasal odontoid resection to decompress the bulbomedullary junction: A reliable anterior minimally invasive technique without posterior fusion. Eur Spine J. 2012;21(SUPPL. 1):555-560.
4. Kyoshima K, Kakizawa Y, Tokushige K, et al. Odontoid compression of the brainstem without basilar impression—"Odontoid invagination." J Clin Neurosci 2005;12:565-569.
5. Ubogu EE, Zaidat OO. Vertebrobasilar dolichoectasia diagnosed by magnetic resonance angiography and risk of stroke and death: a cohort study. J Neurol Neurosurg Psychiatry 2004;75:22-26.
6. Kansal R, Mahore A, Dange N, et al. Dolichoectasia of vertebrobasilar arteries as a cause of hydrocephalus. J Neurosci Rural Pract 2011;2:62.
7. Matsukawa H, Shinoda M, Fujii M, et al. Basilar dolichoectasia and the spontaneous intradural vertebral artery dissection. Brain Inj 2016;30:90-94.
8. Fein JM, Frishman W. Neurogenic Hypertension Related to Vacular Compression of the Lateral Medulla .pdf. Neurosurgery.1980(6):615-22.
9. Kim P, Ishijima B, Takahashi H, et al. Hemiparesis caused by vertebral artery compression of the medulla oblongata. J Neurosurg 1985;62:425-429.
10. Hongo K, Nakagawa H, Morota N, et al. Vascular compression of the medulla oblongata by the vertebral artery: report of two cases. Neurosurgery 1999;45:907-910.
11. Meyer MA, David CE, Chahin NS. Palatal myoclonus secondary to verterbal artery complresion of the inferior olive. J Neuroimaging 2000;10:221-223.
12. Ubogu EE, Chase CM, Verrees MA, et al. Cervicomedullary junction compression caused by vertebral artery dolichoectasia and requiring surgical treatment. J Neurosurg 2009;96:140-143.
13. Betgeri SS, Rajesh S, Adkatalwar V, et al. Spastic quadriparesis caused by anomalous vertebral artery compression of spinal cord at the cervico-medullary junction. Neuroradiol J 2015;28:67-71.
14. Haubrich C, Kleiser N, Kosinski C, et al. Episodic dysarthria related to vascular medullary compression. J Neurol 2010;257:296-299.
15. Nakahara Y, Kawashima M, Matsushima T, et al. Microvascular decompression surgery for vertebral artery compression of the medulla oblongata: 3 cases with respiratory failure and/or dysphagia. World Neurosurg 2014;82. 535.e11-535.E16.
16. Lombarski L, Kunert P, Skawiński M, et al. Symptomatic medulla compression by vertebral artery. Neurol Neurochir Pol 2018;52:519-527.
17. Dembo T, Tanahashi N. Opalski syndrome caused by vertebral artery compression of the lateral surface of the medulla oblongata. Intern Med 2013;52:1115-1120.
18. Park H-E, Lee J-E, Son B, et al. Rabbit syndrome associated with medullary compression by vertebral artery. Neurol Sci 2016;37:1167-1168.
19. Himi T, Kataura A, Tokuda S, et al. Downbeat nystagmus with compression of the medulla oblongata by the dolichoectatic vertebral arteries. Am J Otol 1995;16:377-381.
20. Lee A.G.Downbeat nystagmus associated with caudal brainstem compression by the vertebral artery. 2001;21:219-220.
21. Moon KH, Lee SA, Ahn JS, Kwun BD. Downbeat nystagmus associated with brainstem compression by vertebral artery. J Korean Neurosurg Soc 2007;41:190-192.
22. Savitz SI, Ronthal M, Caplan LR. Vertebral artery compression of the medulla. Arch Neurol 2006;63:234-241.
23. Passero S, Rossi S, Giannini F, et al. Brain-stem compression in vertebrobasilar dolichoectasia. Multimodal Electrophysiol Study 2001;112:1531-1539.
24. Jannetta PJ, Segal R, Wolfson SK. Neurogenic hypertension: etiology and surgical treatment. I. observations in 53 patients. Ann Surg 1985;201:391-398.
25. Naraghi R, Huk W, Fahlbusch R, et al. Posterior fossa neurovascular anomalies in essential hypertension. Lancet 1994;344:1466-1470.
26. Jannetta PJ, Levy EI, Clyde B, et al. Medullary compression and hypertension. J Neurosurg 1994;89:169-170.
27. Grasso G, Alafaci C, Passalacqua M, et al. Landmarks for vertebral artery repositioning in bulbar compression syndrome: anatomic and microsurgical nuances. Neurosurgery 2005;56(1 SUPPL.):160-164.
28. Raabe A, Jaiimsin A, Seifert V, et al. Use of a strip-clip technique to maintain transposition of a vertebral artery in microvascular decompression surgery. Acta Neurochir (Wien) 2011;153:2393-2395.
29. Shigeno T, Kumai J, Horikawa H, et al. Transposition technique of microvascular decompression for hemifacial spasm without using a brain retractor. Neurol Surg 2009;37:35-42.
30. Levy EI, Clyde B, McLaughlin MR, et al. Microvascular decompression of the left lateral medulla oblongata for severe refractory neurogenic hypertension. Neurosurgery 1998;43:1-9.
31. Salvi F, Mascalchi M, Bortolotti C, et al. Hypertension, hyperekplexia, and pyramidal paresis due to vascular compression of the medulla. Neurology 2012;55:1381-1385.
32. Ishikawa T, Nagayama M, Iida M, et al. Bulbovascular compression by megadolichobasilar artery manifested as neurogenic and refractory hypertension. Rinsho Shinkeigaku 2004;44:359-364.
33. Akimura T, Furutani Y, Jimi Y, et al. Essential hypertension and neurovascular compression at the ventrolateral medulla oblongata: MR evaluation. Am J Neuroradiol 1995;16:401-405.
34. Chan LL, Lo YL, Lee E, et al. Ventrolateral medullary compression in hypertensive patients with hemifacial spasm. Neurology 2005;65:1467-1470.
35. Hänggi D, Steiger HJ. Symptomatic vertebral artery conflicts to the medulla oblongata and microsurgical treatment options: review of the literature. Neurosurg Rev 2009;32:143-149.

36. Koguchi M, Nakahara Y, Kawashima M, et al. Successful microvascular decompression of the medulla oblongata for a case with respiratory failure: case report. Neurol Surg 2011;39:1085-1089.
37. Sadashiva N, Shukla D, Bhat DI, et al. Vertebral artery dolicoectasia with brainstem compression: role of microvascular decompression in relieving pyramidal weakness. Acta Neurochir (Wien) 2016;158:797-801.
38. Ren J, Sun H, Diao Y, et al. 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.e11-994.e19.
39. Maruyama K, Tanaka M, Ikeda S, et al. A case report of quadriparesis due to compression of the medulla oblongata by the elongated left vertebral artery. Rinshō shinkeigaku = Clin Neurol. 1989;29:108-111.
40. Kobayashi T, Ogawa A, Kameyama M, et al. Chiari malformation with compression of the medulla oblongata by the vertebral arteries. J Neurosurg 2009;77: 307-309.
41. Hongo K, Kobayashi S, Hokama M, et al. Vertebral artery section for treating arterial compression of the medulla oblongata. J Neurosurg 2009;79:116-118.
42. Murata H, Waga S, Kojima T, et al. Medulla oblongata compression by tortuous vertebral artery: case report. Neurol Surg 1995;23:349-353.
43. Takano S, Saitoh M, Miyaseka Y, et al. A case report of hemiparesis due to compression of the medulla oblongata by an elongated vertebral artery. No Shinkei Geka 2001;29:247-251.
44. Koyama S, Maeda T. Medulla and upper cervical cord compression by bilateral vertebral artery presented with myelopathy and drop attack: case report. Brain Nerve 2002;54:435-439.
45. Tomasello F, Alafaci C, Salpietro FM, et al. Bulbar compression by an ectatic vertebral artery: A novel neurovascular construct relieved by microsurgical decompression. Neurosurgery 2005;56(1 SUPPL.):117-124.
46. Kamada T, Tateishi T, Yamashita T, et al. A case of medulla oblongata compression by tortuous vertebral arteries presenting with spastic quadriplegia. Rinsho Shinkeigaku 2013;53:356-361.