

## Case Report

## Anomalous vertebral artery compression of the spinal cord at the cervicomedullary junction

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**Abstract****Background:** Myelopathy from ectatic vertebral artery compression of the spinal cord at the cervicomedullary junction is a rare condition.**Case Description:** A 63-year-old female was originally diagnosed with occult hydrocephalus syndrome after presenting with symptoms of ataxia and urinary incontinence. Ventriculoperitoneal shunting induced an acute worsening of the patient's symptoms as she immediately developed a sensory myelopathy. An MR scan demonstrated multiple congenital abnormalities including cervicomedullary stenosis with anomalous vertebral artery compression of the dorsal spinal cord at the cervicomedullary junction. The patient was taken to surgery for a suboccipital craniectomy, C1–2 laminectomy, vertebral artery decompression, duraplasty, and shunt ligation. Intraoperative findings confirmed preoperative radiography with ectatic vertebral arteries deforming the dorsal aspect of the spinal cord. There were no procedural complications and at a 6-month follow-up appointment, the patient had experienced a marked improvement in her preoperative signs and symptoms.**Conclusion:** Myelopathy from ectatic vertebral artery compression at the cervicomedullary junction is a rare disorder amenable to operative neurovascular decompression.**Key Words:** Cervicomedullary stenosis, microvascular decompression, myelopathy, vertebral artery**Access this article online****Website:**[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)**DOI:**

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**Quick Response Code:****INTRODUCTION**

The vertebral arteries are notoriously variable in their course. This can be problematic in the surgical management of many conditions of the cervical spine. Occasionally, anomalies of the vertebral arteries themselves become symptomatic. Vertebral artery anomalies have been reported to cause a variety of symptoms including neck and arm pain,<sup>[2,8,11,12,15,18,19]</sup> occipital neuralgia,<sup>[4,9,10]</sup> torticollis,<sup>[18]</sup> myelopathy,<sup>[3,6,9,13,14,15,17]</sup> drop attack,<sup>[6]</sup>

radiculopathy,<sup>[2]</sup> pyramidal tract signs,<sup>[6,10,16]</sup> and cranial nerve deficits.<sup>[5,16,17]</sup>

One mechanism by which anomalous vertebral arteries may become symptomatic is the direct compression of adjacent neural structures. The type and severity of the symptoms therefore arise from the location and degree of compression. Both surgical and endovascular techniques have been used to treat vertebral artery anomalies. One reported case was successfully treated by endovascular occlusion of the vertebral artery.<sup>[8]</sup> Most

patients, however, have been treated by neurovascular decompression. Follow-up of one patient 6 years after vascular decompression for myelopathy and pain revealed a persistent improvement in symptoms.<sup>[19]</sup>

Few cases of direct vertebral artery compression of the spinal cord resulting in myelopathy have been reported.<sup>[3,6,14,15,17]</sup> The authors present a novel case of a patient who developed a sensory myelopathy in association with vertebral artery compression of the dorsal spinal cord which improved after decompression.

## CASE REPORT

### History

A 63-year-old woman employed as a sign language interpreter presented to an outside hospital in 2006 with a 3- to 4-year history of gait disequilibrium and occasional bladder incontinence. Other medical history was only significant for essential tremor for which she was taking propranolol. She was initially diagnosed with “normal pressure hydrocephalus” for which she was treated with a ventriculoperitoneal shunt. However, immediately following the procedure, she awakened with symptoms of whole-body numbness, particularly in the extremities. For the next 3 years, her condition waxed and waned with a trend toward deterioration. According to the patient, her arms and legs were numb as if she had been “sleeping on them all of the time.” She occasionally noticed painless burns and cuts on her hands and fingers. She was intermittently treated with steroids and intravenous immunoglobulin (IVIG) for a possible inflammatory radiculoneuropathy but without lasting benefit. An MR scan of the head obtained at that time reported small bilateral subdural hematomas without mass effect. No other abnormalities were reported. She did not notice weakness, but she was increasingly clumsy. Her gait became progressively more unsteady, particularly in the dark, and she had fallen at least twice. More troubling, she was losing her ability to gesture language with her hands – a skill critical for her employment. She was referred to our institution with a working diagnosis of “atypical chronic inflammatory demyelinating polyneuropathy” (CIDP).

### Examination

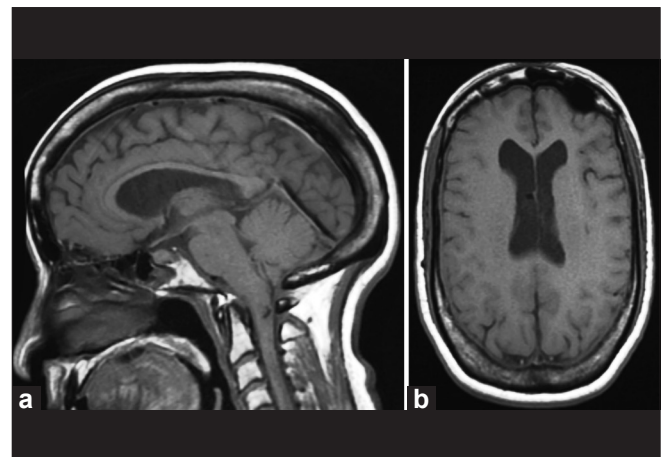
On examination, she had a dolichocephalic head. Alertness and orientation were normal. Cranial nerves were intact. Strength was normal in all extremities, but her alternating motion rates were reduced. She had a marked impairment in vibration and joint position sense in both upper extremities, slightly worse on the right than the left. Lower extremity sensation was also impaired, but less so. She had minimal ataxia but tandem gait was impaired and her Romberg test was moderately positive. Knee and ankle reflexes were mildly increased; plantar responses were flexor and there was no clonus.

A thermoregulatory sweat test was obtained and revealed mild hypohydrosis of the lower extremities, which was of uncertain significance. Quantitative sensory testing of the left upper extremity demonstrated hyposensitivity to vibration distally which normalized proximally. The heat/pain thresholds were reduced. Somatosensory potentials of the right median and tibial nerves were also obtained. For the tibial nerve, spinal and cortical potentials were normal both in amplitudes and latencies. The right median potential, however, demonstrated prolongation of both the N13–N20 and N9–N20 interpeak latencies.

### Radiology

Imaging included an MR scan of the head which demonstrated dolichocephaly as noted on the clinical examination as well as the presence of the known ventriculoperitoneal shunt [Figure 1]. The previously noted small subdural hematomas were no longer present, but there was diffuse enhancement of the dura overlying the convexities consistent with intracranial hypotension. On an MR scan of the cervical spine, the vertebral arteries were identified as entering the spinal canal at C1–2 where they passed posterior to the cord until they nearly touched at the midline before turning cranially and back laterally. The anomalous vertebral arteries severely compressed the posterior aspect of the cord with a resultant intramedullary T2 signal change. Ectopic cerebellar tonsils caused further crowding at the foramen magnum. There was no syrinx in the spinal cord [Figure 2].

The patient was diagnosed with a sensory myelopathy related to a Chiari malformation and posterior cervical cord compression at the C1–2 levels secondary to anomalous vertebral arteries.



**Figure 1:** Sagittal (a) and axial (b) T1-weighted MR scans demonstrate an elongated, narrow head with relative flattening of the basal angle and suboccipital bone. Note the crowding of the cervicomedullary junction consistent with a mild Chiari malformation. The subdural hematomas seen on a prior study have resolved

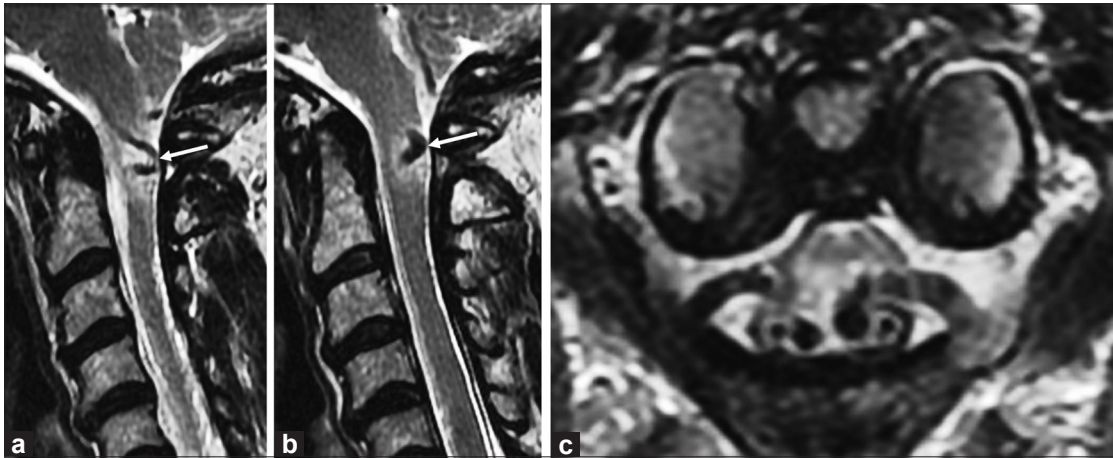
## Intervention

The patient was taken to surgery for a suboccipital craniectomy, C1–2 laminectomy, and duraplasty. After exposure of the suboccipital skull and upper cervical spine, the vertebral arteries were identified lateral to the facets at the C2–3 interspace by manual palpation and Doppler ultrasonography and then carefully protected during subsequent dissection. After completion of the suboccipital craniectomy and C1–2 laminectomies, a Y-shaped incision in the dura was made over the inferior cerebellum and upper cervical spinal cord. The cerebellar tonsils were observed to extend down to the level of C1 consistent with a mild Chiari malformation. The most striking finding, however, was the course of the vertebral arteries [Figure 3] which entered the cervical canal at the C1–2 level laterally and immediately compressed the dorsal and lateral aspects of the spinal cord on both sides

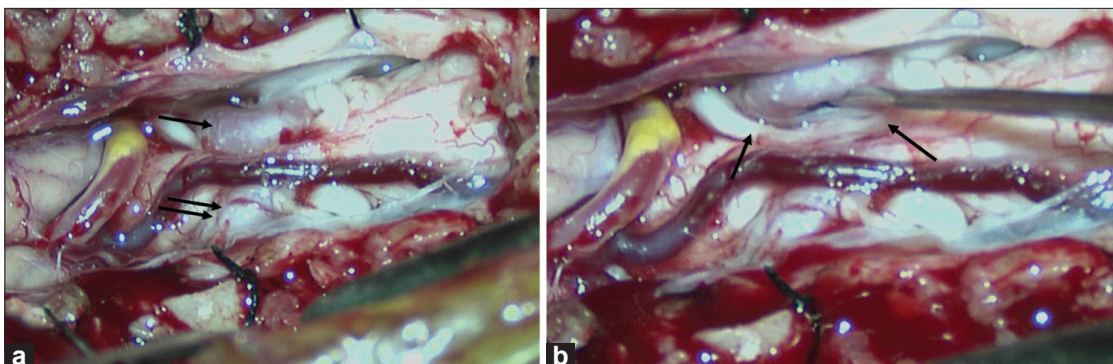
as they converged toward the midline. They then turned superiorly and back laterally passing anterior to the cord at C1. The vertebral arteries were gently dissected from the dorsal surface of the cord but their turgor and short length precluded a complete decompression. Their impression into the cord substance could be clearly observed. The cerebellar tonsils were partially resected to assure satisfactory cervicomedullary decompression. The dura was closed with a bovine pericardial patch graft. The ventriculoperitoneal shunt was ligated through a small infraclavicular incision.

## Postoperative course

There were no perioperative complications. The patient was kept under observation in the hospital for 3 days. Physical medicine and rehabilitation services assisted with mobilization and in activities of daily living.



**Figure 2:** Sagittal T2-weighted MR scans of the cervical spine demonstrate anomalous right (a) and left (b) vertebral arteries (arrows) dorsal to and impressing into the cord substance. Note the T2 signal change in the adjacent cord. Again seen is the crowding and tonsillar herniation at the cervicomedullary junction consistent with a mild Chiari malformation. No syrinx was present. (c) Axial T2-weighted image of the C1–2 interspace demonstrating anomalous vertebral arteries deforming the dorsal spinal cord. T2-signaling abnormality in the dorsal cord is evident



**Figure 3:** Intraoperative photographs of the dorsal cervicomedullary junction. The cerebellar tonsils and an anomalous posterior inferior cerebellar artery can be seen on the left (rostral). (a) The right (single arrow) and left (double arrows) vertebral arteries can be seen penetrating the dura at C1–2 and passing dorsal to the spinal cord until they nearly touch at midline. They then turn rostrally and anteriorly. (b) After the mobilization of the right vertebral artery, the impression left by the artery in the cord substance can be readily observed (arrows)

Shortly following operation, she began to experience improvement in her symptoms and her ability to gesture sign language was improving by the time of discharge. At her 3-month follow-up visit, her position sense in the left hand was normal and in the right hand nearly normal. Vibration sense, which had been essentially absent in the right hand, was near normal. Her gait was stable.

## DISCUSSION

Although anomalies of the vertebral arteries may not be uncommon, reports of deficits due to extracranial neural compression are. Most affected patients present with radiculopathic pain or cranial nerve deficits.

The patient presented here experienced a prolonged course of progressive neurologic deficits refractory to various treatments including ventriculoperitoneal shunting, high-dose steroids, and IVIG before appropriate diagnosis and treatment were instituted. Her original symptoms were gait instability and bladder incontinence. Although such symptoms can suggest normal pressure hydrocephalus, we postulate that her gait unsteadiness was due to posterior column dysfunction and shunting was a misguided intervention that potentially worsened her symptoms. We suspect that the shunt induced or aggravated brain sag as evidenced by the subsequent subdural hematomas, exacerbating the compression already present at the foramen magnum and upper cervical cord.

The patient exhibited predominantly sensory symptoms and signs. Although sensory findings may occur in patients with Chiari malformations, they are usually related to secondary syrinx development. The imaging of the cervical spine in this patient did not reveal any evidence of a syrinx; rather, flow voids corresponding to the vertebral arteries were seen pursuing an anomalous course dorsal to the spinal cord. The impression of the arteries into the cord substance and the associated T2 signal change strongly implicated this as the etiology of the patient's symptoms.

The significance of the other coexisting developmental anomalies, including platybasia, dolichocephaly, and a mild Chiari malformation is uncertain. Clearly, the patient had lived with these anomalies her whole life but only developed symptoms in recent years, symptoms likely exacerbated by the ventriculoperitoneal shunt. It is possible that the chronic pulsatile compression resulted in local demyelination as has been observed in other vascular compression syndromes.<sup>[1,7]</sup> For other disorders, vascular decompression has been proven to be a reliable treatment strategy.

In some cases, techniques to mobilize and restrain the vertebral arteries have been described to maintain

neural decompression. In this case, however, the turgor and short length of the impinging vessels precluded a satisfying decompression of the involved cervical cord. Nevertheless, the patient's neurologic deficits improved which we attribute to the completion of a wide decompressive laminectomy and duraplasty, reduction of the cerebellar tonsils at the foramen magnum, and arrest of the exacerbating CSF drainage by shunt ligation.

## CONCLUSION

Based upon this case and a few others in the literature, we emphasize the role of anomalous vertebral arteries as a potential cause of compressive myelopathy and the importance of its recognition so as to direct appropriate treatment.

## REFERENCES

- Cheshire WP, Odell JA, Woodward TA, Wharen RE Jr. Cervical sympathetic neuralgia arising from a schwannoma. *Headache* 2007;47:444-6.
- Detwiler PW, Porter RW, Harrington TR, Sonntag VK, Spetzler RF. Vascular decompression of a vertebral artery loop producing cervical radiculopathy. *Case report. J Neurosurg* 1998;89:485-8.
- Furumoto T, Nagase J, Takahashi K, Itabashi T, Iai H, Ishige N. Cervical myelopathy caused by the anomalous vertebral artery. *A case report. Spine (Phila Pa 1976)* 1996;21:2280-3.
- Kim K, Mizunari T, Kobayashi S, Ishii S, Teramoto A. [Occipital neuralgia caused by the compression of the fenestrated vertebral artery: a case report]. *No Shinkei Geka* 1999;27:645-650.
- Kitagawa M, Nakagawa Y, Kitaoka K, Kobayashi N, Ishikawa T, Nagashima M. [Accessory nerve paralysis due to compression of the fenestrated vertebral artery]. *No Shinkei Geka* 1998;16:1173-7.
- Koyama S, Maeda T, Komine A. [Medulla and upper cervical cord compression by bilateral vertebral artery presented with myelopathy and drop attack: Case report]. *No To Shinkei* 2002;54:435-9.
- Love S, Coakham HB. Trigeminal neuralgia: Pathology and pathogenesis. *Brain* 2001;124:2347-60.
- Maiuri F, Iaconetta G, Gallicchio B, Briganti F. Coiling of the vertebral artery presenting with neuralgic pain. *Clin Neurol Neurosurg* 1997;99:56-9.
- Morikawa K, Ohkawa N, Yamashita S. [Surgical decompression for the C-1 and C-2 sensory roots and upper cervical cord in a case with cervical myelopathy and occipital neuralgia due to bilateral fenestration of vertebral artery: A case report]. *No Shinkei Geka* 1993;21:1035-8.
- Nishiura T, Fujiwara K, Handa A, Gotoh M, Tsuno K, Ishimitsu H. [Cervical myelopathy caused by bilateral vertebral artery compression]. *No Shinkei Geka* 1998;26:45-50.
- Paksoy Y, Levendoglu FD, Ogun CO, Ustun ME, Ogun TC. Vertebral artery loop formation: a frequent cause of cervicobrachial pain. *Spine (Phila Pa 1976)* 2003;28:1183-8.
- Satoh S, Yamamoto N, Kitagawa Y, Umemori T, Sasaki T, Iida T. Cervical cord compression by the anomalous vertebral artery presenting with neuralgic pain. *Case report. J Neurosurg* 1993;79:283-5.
- Takahashi T, Tominaga T, Hassan T, Yoshimoto T. Cervical cord compression with myelopathy caused by bilateral persistence of the first intersegmental arteries: Case report. *Neurosurgery* 2003;53:234-7; discussion 237.
- Takahashi Y, Sugita S, Uchikado H, Miyagi T, Tokutomi T, Shigemori M. Cervical myelopathy due to compression by bilateral vertebral arteries--case report. *Neurol Med Chir (Tokyo)* 2001;41:322-4.
- Takei H, Sagae M, Chiba K, Ogino T. The long-term follow-up of surgical treatment for cervical myelopathy with severe nape and upper arm pain caused by the anomalous vertebral artery: case report. *Spine (Phila Pa 1976)* 2008;33:E611-3.

16. 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;56:117-24.
17. Vincentelli F, Caruso G, Rabehanta PB, Rey M. Surgical treatment of a rare congenital anomaly of the vertebral artery: Case report and review of the literature. *Neurosurgery* 1991;28:416-20.
18. Watanabe K, Hasegawa K, Takano K. Anomalous vertebral artery-induced cervical cord compression causing severe nape pain. Case Report. *J Neurosurg* 2001;95:146-9.
19. Yano K, Murase S, Kuroda T, Noguchi K, Tanabe Y, Yamada H. Cervical cord compression by the vertebral artery causing a severe cervical pain: Case report. *Surg Neurol* 1993;40:43-6.