

Cervicomedullary junction compression caused by vertebral artery dolichoectasia and requiring surgical treatment

Case report

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✓ A case of progressive brainstem syndrome secondary to vertebral artery (VA) dolichoectasia is reported. The patient presented with partial bilateral abduction paralysis, which progressed to quadriplegia, ataxia, and areflexia. The initial diagnosis was stroke, but because of the patient's deterioration, a diagnosis of Miller–Fisher syndrome was made. Neuroimages obtained at that time revealed an ectatic left VA with minimal cervicomedullary compression. The patient continued to deteriorate despite medical management. Follow-up imaging demonstrated worsened cervicomedullary compression. An emergency posterior fossa neurovascular decompression was performed using a Gore-Tex sling and resulted in mild neurological improvement. This case emphasizes that early recognition and surgical intervention to prevent progressive neurological sequelae are crucial in symptomatic VA dolichoectasia.

KEY WORDS • cervicomedullary compression • dolichoectasia • neurovascular decompression • vertebrobasilar artery

CERVICOMEDULLARY compression is a complication of Type I Arnold–Chiari malformations,⁷ achondroplasia,^{1,21} and other congenital craniovertebral and skull base anomalies. Dolichoectasia is a rare vasculopathy that causes arterial elongation and enlargement.² Several case reports indicate that VA dolichoectasia is associated with cervicomedullary compression.^{8–12,14,15,20,21} There are few case reports in which the benefit of neurovascular decompression in the management of cervicomedullary compression is discussed,^{8,9,11,18,20} and none describes surgical intervention late in the course of the disease. We report a case of cervicomedullary compression caused by a dolichoectatic left VA, in which progressive deterioration occurred that required surgical decompression.

Case Report

History and Examination. This 70-year-old right-handed man with recently diagnosed hypertension presented with horizontal diplopia. Examination showed skew deviation and partial bilateral abduction paralysis. A brain MR image revealed small lacunar pontine infarcts on fluid-attenuat-

ed inversion-recovery images. The patient was started on an aspirin regimen for a presumed small-vessel ischemic stroke and then discharged.

He subsequently developed worsening of his symptoms, with neck stiffness and upper-extremity weakness and numbness. A motor examination revealed mild bilateral upper- and lower-extremity paresis, with a Medical Research Council¹⁷ grade of 4. Dysmetria and appendicular gait ataxia were also present. A sensory examination showed impaired proprioception and pinprick sensation bilaterally, worse on the left side. Reflexes were normal, with absent Babinski sign. A computerized tomography scan of the brain yielded normal results. A lumbar puncture showed an elevated protein level of 111 mg/dl; otherwise routine laboratory tests were unrevealing. Repeated MR imaging and MR angiography demonstrated a dolichoectatic left VA with minimal cervicomedullary compression, which was not thought to be significant (Fig. 1).

A needle electromyographic study revealed mild to severe bilateral upper-extremity denervation with cervical paraspinal involvement, with no demyelination. Results of a nerve conduction study were normal. A provisional diagnosis of Miller–Fisher syndrome was made. The patient remained clinically unchanged despite plasmapheresis and intravenous administration of immunoglobulin. Results of repeated electromyographic and nerve conduction studies confirmed the previous findings. Tests for an antiglycolipid

Abbreviations used in this paper: MR = magnetic resonance; SPECT = single-photon emission computerized tomography; VA = vertebral artery.

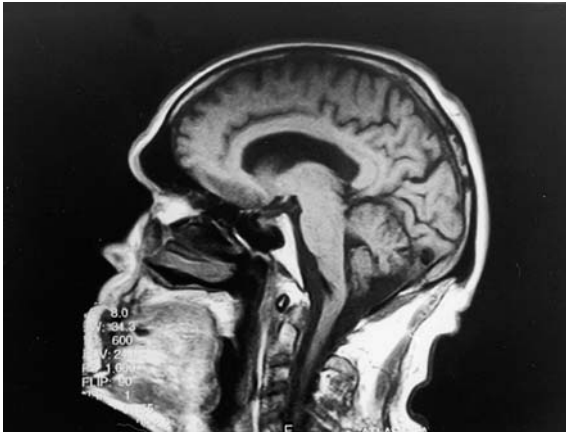


FIG. 1. Admission sagittal T₁-weighted MR image demonstrating mild cervicomedullary junction and upper cervical spinal cord compression.

(GQ1b) and paraneoplastic antibodies (anti-Hu and anti-Yo) were negative.

After a 4-month period of stabilization, the patient experienced incoordination, slurred speech, difficulty hearing, and dysphagia to solids and liquids. A cranial nerve examination revealed complete bilateral abduction gaze palsies with limitation of the upward gaze on pursuit evaluation, mild left facial droop, bilateral hearing loss, an absent gag reflex, dysarthria, and right tongue deviation. The patient also had titubation, hypertonia, and quadriparesis, with complete paraplegia of the legs and paresis of the upper extremities. He experienced episodes of right autonomic hyperactivity accompanied by flushing and hyperhidrosis, along with hyperesthesias and right tonic neck deviation. Dystonic orofacial movements and jerky left adduction nystagmus were also observed. He became progressively emotionally labile and unresponsive to commands and developed respiratory distress.

An emergency MR image demonstrated worsening cervicomedullary compression caused by the ectatic left VA (Fig. 2). The angiogram confirmed a tortuous distal left VA (Fig. 3). A trial 15-minute occlusive balloon angioplasty was performed to ascertain the feasibility of intraoperative arterial ligation. A ^{99m}Tc SPECT scan of the brain demonstrated a diminution in the size and volume of the left cerebellum and the medulla could not be visualized below the level of the cerebellar hemispheres. The patient underwent emergency neurovascular decompression.

Operation. A left suboccipital craniectomy and C-1 laminectomy were performed using a U-shaped myocutaneous scalp flap based inferiorly. The posterior third of the left occipital condyle was removed to facilitate lateral exposure. The dura was then opened using a hockey stick incision based laterally. The left VA was larger than normal and included a tortuous segment that compressed on and deformed the left medulla and upper cervical spinal cord (Fig. 4 left). The VA was gently mobilized and an approximately 5 × 40-mm strip of Gore-Tex (W.L. Gore & Associates, Inc., Flagstaff, AZ) was passed around the vessel to form a sling. A short right-angled aneurysm clip was used to secure the ends of the sling, which was then sutured to the lateral posterior fossa petrous dura with three interrupted 4-0 nylon stitches (Fig. 4 right).



FIG. 2. Emergency preoperative sagittal T₁-weighted MR image demonstrating angulation and posterior displacement of the lower medulla and upper cervical spinal cord at the level of the cervicomedullary junction (arrow), which is more severe than in Fig. 1. No osseous deformities or cerebellar tonsillar herniation are visualized.

Postoperative Course. A postoperative MR image revealed significant improvement in the cervicomedullary compression (Fig. 5). The patient demonstrated some mild improvement in his neurological deficits. The orofacial and neck dystonia, upgaze palsy, inability to follow simple commands, and respiratory distress resolved. His quadriparesis was mildly improved, more significantly on the right side and in the upper extremities. He was then transferred to a spinal cord rehabilitation unit. An evaluation performed 6 months postoperatively showed complete resolution of the abduction paralysis, facial paresis, and hearing deficits, although his quadriparesis persisted.



FIG. 3. Preoperative vertebral arteriogram, frontal view, demonstrating a normal right VA and a dolichoectatic left VA, with deviation of the distal segment medially before the vertebralbasilar junction (arrow).

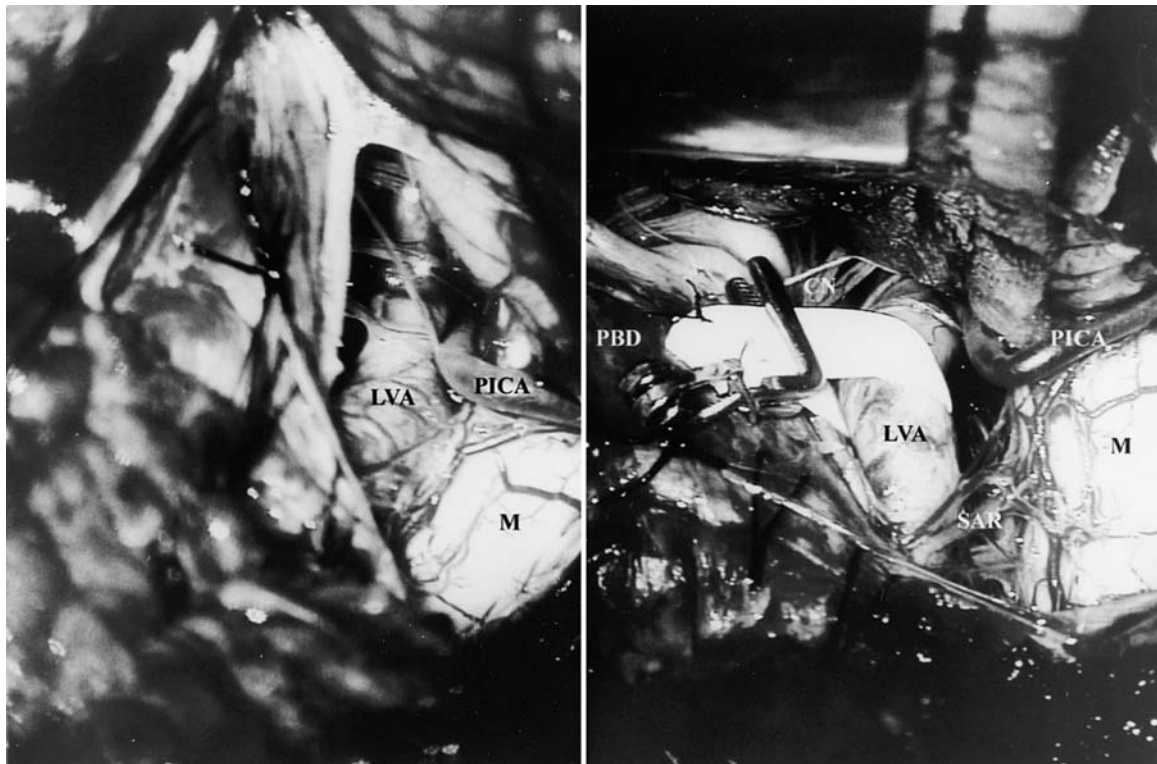


FIG. 4. *Left:* Intraoperative photograph showing an enlarged left VA (LVA) with a tortuous segment compressing the medulla (M) and the upper cervical spinal cord. The posterior inferior cerebellar artery (PICA) is also demonstrated from its origin. *Right:* Intraoperative photograph showing neurovascular decompression of the medulla by using a Gore-Tex belt fixed to the dolichoectatic left VA by a curved aneurysm clip, with suturing to the petrous bone dura (PBD). The ninth and 10th cranial nerves (CN) and the spinal accessory nerve rootlets (SAR) are shown rostral and caudal, respectively, to the posterior inferior cerebellar artery.

Discussion

This is a case of a complex, near-fatal brainstem syndrome related to VA dolichoectasia. A variety of clinical syndromes have been associated with ectatic vertebrobasilar arteries: these include, most commonly, an assortment of cranial nerve syndromes,^{3-5,9,16,22} transient or permanent motor deficit,^{9,10,13,15,22} cerebellar dysfunction,¹⁰ central sleep

apnea,¹⁴ hydrocephalus, and ischemic stroke.^{6,12,22} Ischemia to the lower pons, which includes the retinal slip (sensory stimulus for vertical pursuit eye movements), can be attributed to direct compression of the pontomedullary junction by the dolichoectatic VA or to distal microemboli induced by the arterial malformation. The dynamic nature of the presentation can possibly be explained by hypertension-induced arterial dissection or worsening dolichoectasia leading to worsening brainstem compression.

The evaluation of these patients includes the use of conventional angiography, MR angiography, computerized tomography scanning, or computerized tomography angiography.^{5,6,19} The diagnosis is best reached by combining the clinical presentation with adjunctive neuroimaging modalities. Symptomatic VA dolichoectasia may be associated with significant morbidity and a high mortality rate. In a small study on clinical outcomes in patients with VA dolichoectasia it was suggested that the 3-year survival rate may be 60% or less depending on the clinical features at the time of presentation.¹²

The disease in our patient was diagnosed using MR imaging and MR angiography, which were further supported by cerebral angiography. The use of SPECT scanning in our case was helpful in confirming brainstem hypoperfusion, attesting to the clinical significance of the left-sided VA dolichoectasia and the need for surgical intervention. Surgical decompression was performed late in the course of the disease. The patient experienced a mild recovery, and



FIG. 5. Postoperative sagittal T₁-weighted MR image demonstrating reduced angulation and posterior displacement of the lower medulla and upper cervical spinal cord.

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the procedure halted further progression of his symptoms. The outcome of the surgical intervention may have been affected by a delay in diagnosis, which led to irreversible brainstem ischemia.

Conclusions

Earlier surgical intervention is of paramount importance and might have resulted in fewer sequelae from this vascular anomaly. The mild improvements noted in this case indicate that early recognition and surgical intervention in the setting of symptomatic VA dolichoectasia may be beneficial in reversing neurological deficits and improving outcomes. This diagnosis should be considered early in patients with isolated cranial nerve findings and progressive brainstem syndromes of unclear origin. This case illustrates that functional neuroimaging, including SPECT brain scanning, aids in the clinicopathological correlation of VA dolichoectasia and can be used in equivocal cases, which may help in directing therapeutic options early in cases of this disease.

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