Type 2 persistent primitive proatlantal intersegmental artery, a rare variant of persistent carotid-vertebrobasilar anastomoses

Gagandeep Choudhary, MD^a , Narendra Adhikari, MD^a , Jad Chokr, MD^b , and Nishant Gupta, MD^c

^aDivision of Neuroradiology, Department of Radiology, University of Alabama at Birmingham, Birmingham, Alabama; ^bDepartment of Radiology, Clemenceau Medical Center, Affiliated with Johns Hopkins International, Beirut, Lebanon; ^cDepartment of Radiology, Columbia University Medical Center, New York, New York

ABSTRACT

We describe a 48-year-old man with type 2 persistent primitive proatlantal intersegmental artery found incidentally on imaging. It is one of the rare persistent carotid-vertebrobasilar anastomoses in which the anomalous vessel arises from the external carotid artery and enters the skull through the foramen magnum.

KEYWORDS Anomalous vertebral artery; persistent carotid-vertebrobasilar anastomoses; primitive proatlantal intersegmental artery

During the early embryonic phase, four vessels originate from the primitive internal carotid artery (ICA) and dorsal aorta: the trigeminal, otic, hypoglossal, and proatlantal intersegmental (ProA) arteries. They are the primitive carotid-vertebrobasilar anastomoses that will supply the future vertebrobasilar system. Within a week, these anastomoses regress with development of the vertebrobasilar system. In rare instances, they are incidentally discovered in adults and considered remnants of the embryonic circulation. We present a patient with a persistent ProA, with emphasis on the segmental property of the primitive carotid-vertebrobasilar anastomoses, followed by a review of cases presented in the literature and their associated anomalies.

CASE REPORT

A 48-year-old man presented to the emergency room with complaints of mild abdominal and back pain after a motor vehicle crash. No neurological deficit, neck pain, or other abnormality was present on initial assessment. All vitals were within normal limits. As part of a routine protocol at our institute, the patient underwent total-body computed tomography (CT) scanning, including CT angiography of the neck. No significant imaging abnormality was seen, apart

from a mild amount of fluid in the pelvis, which was attributed to physiological state in this age group. CT angiography showed no evidence of fracture, traumatic vessel injury, vascular occlusion, or flow-limiting stenosis.

There was a three-vessel aortic arch. The right vertebral artery (VA) was hypoplastic arising from the subclavian artery and ended as a posterior inferior cerebellar artery. The left VA was absent. Both common carotid arteries, carotid bulbs, ICAs, and external carotid arteries were patent. An anomalous artery equal to the caliber of the ICA was seen arising from the proximal left external carotid artery at the origin of the lingual artery at the C2 vertebral level (*Figure 1*). The anomalous vessel coursed cranially and entered the foramen magnum coursing between the C1 arch and the occiput, without passing through the transverse foramina of the cervical vertebrae. Intracranially, it had a tortuous course and continued as a basilar artery, which terminated in posterior cerebral arteries. The right-sided posterior communicating artery was present but small in caliber, and the left posterior communicating artery was not seen. Both intracranial ICAs and proximal middle cerebral arteries, the anterior cerebral arteries, and the anterior communicating artery were present and unremarkable. The patient did not have any symptoms attributable to this vascular variant.

Corresponding author: Jad Chokr, MD, Department of Radiology, Clemenceau Medical Center, Clemenceau Street, Beirut 11-2555, Lebanon (e-mail: jad_1986@hotmail.com)

Color versions of one or more of the figures in the article can be found online at www.tandfonline.com/ubmc.

Received August 22, 2018; Revised September 24, 2018; Accepted October 4, 2018.

Figure 1. (a) Diagram demonstrating a three-vessel aortic arch (AA), which is the most common variation. The first branch is the brachiocephalic trunk (BCT), which divides into the right subclavian artery (Rt-SCA) and common carotid artery (CCA). The second branch is the left CCA, and the third branch is the left subclavian artery (Lt-SCA). (b) Various carotid-basilar anastomoses from caudal to cranial: proatlantal artery type 1 (ProA-1), proatlantal artery type 2 (ProA-2), hypoglossal artery (HA), otic artery (OA), trigeminal artery (TA), and posterior communicating artery (PCOM). The blue circle shows the internal auditory canal; black circle, hypoglossal canal; and green circle, foramen magnum. BA indicates basilar artery; ECA, external carotid artery; ICA, internal carotid artery; VA, vertebral artery.

He was discharged in stable condition with a short course of pain medication.

DISCUSSION

In the early phase of embryonic development, the blood supply to the posterior fossa is entirely dependent on the anterior circulation through carotid-vertebrobasilar anastomoses. At the 4- to 5-mm embryonic stage, the hindbrain is supplied by two parallel neural arches. These neural arches get their blood supply from the anterior circulation through the trigeminal artery, the otic artery, the hypoglossal artery, and the ProA. The first three arteries are also known as pre-segmental arteries and are named after the cranial nerves they follow.^{1,2} The life span of these vessels is approximately a week. As the hindbrain structure develops, the blood vessels undergo extensive modification and the posterior circulation becomes progressively independent from the anterior circulation, with obliteration of these carotid-vertebrobasilar anastomoses. The neural arches consolidate to form the basilar artery, with formation of posterior communicating arteries and VA. The otic artery is the first vessel to obliterate, followed by the hypoglossal, trigeminal, and ProA arteries.¹ The ProA persists until the VAs are fully developed and, in fact, a short segment of ProA is incorporated into the distal cervical VA.¹

Persistent carotid-vertebrobasilar anastomoses are rare, with an overall prevalence of 0.1% to 1.0%.³ The incidence rate of persistent carotid-vertebrobasilar anastomoses is inversely related to the order of regression. The trigeminal is the most common (0.1%–0.6%), followed by the hypoglossal (0.02%–0.10%), otic, and ProA arteries. *Table 1* lists previously reported cases of persistent ProA type 2 and their associated anomalies.

The ProA is an important blood supply to the posterior fossa structures until the seventh and eighth weeks before VA development. Persistent ProAs can be divided into two types based on their origin. Type 1 (~57%) corresponds to the first segmental artery, arises from the ICA, and joins the V4 segment of VA, and type 2 (43%) corresponds to the second segmental artery, arises from the external carotid artery or rarely from the common carotid artery, and joins the V3 segment of VA.^{4,5} Irrespective of their origin, the vessels enter the skull through the foramen magnum. The hypoglossal and ProA are similar but can be differentiated by their course and skull entry site. The hypoglossal course can have a higher origin at the C1 vertebra or the C1 to C2 interspace, is vertically oriented, and enters the skull through the hypoglossal canal. In contrast, the ProA typically arises from the C2 or C3 vertebral level, has a suboccipital horizontal course similar to the V3 segment, and enters the skull through the foramen magnum.⁵ Proximal VAs are absent or hypoplastic in half of the individual with persistent ProA.⁶

In our case, the incidental anomalous artery arose from the proximal part of the left external carotid artery. It did not follow the expected course along the transverse foramen but ran parallel to the cervical vertebra and continued as the V3 segment of the VA, entering the skull through the foramen magnum, which makes it consistent with ProA type 2. In addition, the hypoplastic right VA and right posterior communicating artery with the absence of the left VA and posterior communicating artery makes this left persistent ProA the only major vessel supplying the posterior fossa structures (*Figure 2*).

Most persistent ProAs are detected incidentally on imaging done for unrelated reasons, as in our case. There is no evidence suggesting that the variant arterial anatomy predisposes to cerebrovascular diseases. However, understanding anomalous vascular anatomy, persistent carotid-vertebrobasilar anastomoses, and their association with other anatomical variations is very

Table 1. Previously reported cases of persistent proatlantal artery type 2 with associated anomalies

No.	Author	Age/sex	Side	Additional anomaly
1	Purkayastha et al ⁶	6 years/F	Bilateral	Vein of Galen malformation
2	Purkayastha et al ⁶	1 year/F	Left	Vein of Galen malformation
3	Purkayastha et al ⁶	3 days/M	Left	Vein of Galen malformation
4	Zarghouni and Marichal ⁸	62 years/M	Bilateral	Absence of both VAs
5	Kurose et al ⁹	74 years/F	Right	Aneurysm at the MCA trifurcation
6	Rao and Sethi ¹⁰	23 years/M	Left	Sellar mass
7	Hackett and Wilson ¹¹	63 years/M	Right	Right MCA occlusion
8	Pinstein and Gerald ¹²	52 years/F	Left	Seizures
9	Flynn ¹³	N/A	Left	Saccular aneurysm of the left ICA
10	Berger et al ¹⁴	60 years/F	Left	Tight stenosis at the origin
11	Akay et al ¹⁵	58 years/M	Left	None
12	Arráez-Aybar et al ¹⁶	64 years/F	Left	AVM fed by PCA
13	Lode ¹⁷	57 years/M	Bilateral	Bilateral ICA occlusion
14	Liechty et al ¹⁸	63 years/F	Left	Left ICA stenosis
15	Pelz et al ¹⁹	64 years/M	Bilateral	Right internal capsule infarct
16	Uchino et al ⁷	83 years/M	Left	Left persistent trigeminal artery
17	Obayashi and Furuse ²⁰	50 years/M	Left	Absent left VA, hypoplastic right VA
18	Lui et al ²¹	48 years/F	Bilateral	Absence of both VAs
19	Sato et al ²²	58 years/M	Right	Absent right VA
20	Berman and Stuckey ²³	29 years/F	Left	AVM fed by left PCA
21	Li et al ²⁴	52 years/F	Right	Pulsatile tinnitus

AVM indicates arteriovenous malformation; ECA, external carotid artery; ICA, internal carotid artery; MCA, middle cerebral artery; PCA, posterior cerebral artery; VA, vertebral artery.

Figure 2. Multiple volume-rendered images of the computed tomography angiogram of the neck. (a) Left common carotid artery is bifurcating into the external carotid artery and internal carotid artery. A large persistent proatlantal artery type 2 is seen arising from the external carotid artery and entering the skull via the foramen magnum, (b) continuing as the vertebral artery. (c) Right vertebral artery is present but hypoplastic, and the left vertebral artery is absent.

important in head and neck surgical planning. Recognition of the ProA is also important if surgical ligation or embolization of the external carotid artery is planned.⁷

ORCID

Gagandeep Choudhary <http://orcid.org/0000-0002-3261-1979>

Narendra Adhikari <http://orcid.org/0000-0002-4136-8114>

Jad Chokr <http://orcid.org/0000-0002-2478-9195>

Nishant Gupta <http://orcid.org/0000-0003-0611-5733>

1. Raybaud C. Normal and abnormal embryology and development of the intracranial vascular system. *Neurosurg Clin N Am.* 2010;21:399–426.
2. Choudhary G, Ahuja K, Khan R, Kubal W. Bilateral persistent primitive hypoglossal artery presenting with hemiplegia. *J Radiol Case Rep.* 2018;13(5):1072–1075. doi:10.1016/j.radcr.2018.04.022.
3. Yilmaz E, Ilgit E, Taner D. Primitive persistent carotid-basilar and carotid-vertebral anastomoses: a report of seven cases and a review of the literature. *Clin Anat.* 1995;8(1):36–43. doi:10.1002/ca.980080107.
4. Kolbinger R, Heindel W, Pawlik G, Erasmi-Korber H. Right proatlantal artery type I, right internal carotid occlusion, and left internal carotid stenosis: case report and review of the literature. *J Neurol Sci.* 1993;117(1-2):232–239. doi:10.1016/0022-510X(93)90178-2.
5. Menshawi K, Mohr JP, Gutierrez J. A functional perspective on the embryology and anatomy of the cerebral blood supply. *J Stroke.* 2015;17(2):144–158. doi:10.5853/jos.2015.17.2.144.

6. Purkayastha S, Gupta AK, Varma R, Kapilamoorthy TR. Proatlantal intersegmental arteries of external carotid artery origin associated with Galen's vein malformation. *Am J Neuroradiol*. 2005;26(9):2378–2383.
7. Uchino A, Saito N, Inoue K. Type 2 proatlantal intersegmental artery associated with persistent trigeminal artery diagnosed by MR angiography. *Surg Radiol Anat*. 2012;34(8):773–776. doi:10.1007/s00276-011-0839-1.
8. Zarghouni M, Marichal D. Persistent bilateral proatlantal type II artery. *Proc (Bayl Univ Med Cent)*. 2013;26(1):50–51.
9. Kurose K, Kishi H, Nishijima Y. Type 2 proatlantal artery associated with a ruptured aneurysm—case report. *Neurol Med Chir (Tokyo)*. 1990;30(3):191–193.
10. Rao TS, Sethi PK. Persistent proatlantal artery with carotid-vertebral anastomosis. Case report. *J Neurosurg*. 1975;43(4):499–501. doi:10.3171/jns.1975.43.4.0499.
11. Hackett ER, Wilson CB. Congenital external carotid-vertebral anastomosis. A case report. *Am J Roentgenol Radium Ther Nucl Med*. 1968;104(1):86–89.
12. Pinstein ML, Gerald B. Anomalous communication of the external carotid and vertebral arteries. Persistence of the proatlantal artery. *Radiology*. 1976;118(3):626. doi:10.1148/118.3.626.
13. Flynn RE. External carotid origin of the dominant vertebral artery. Case report. *J Neurosurg*. 1968; 29(3):300–301. doi:10.3171/jns.1968.29.3.0300.
14. Berger L, Gouicem D, Lebied el D, Felisaz A, Coffin O, Maiza D. Internal carotid and ipsilateral type II proatlantal artery stenoses causing simultaneous hemispheric and vertebrobasilar transient ischemia. *J Vasc Surg*. 2011;53(2):475–477. doi:10.1016/j.jvs.2010.08.017.
15. Akay H, Ozturk A, Oguz KK, Cekirge S. Type 2 persistent proatlantal intersegmental artery: demonstration by multislice computed tomography angiography. *Eur J Radiol*. 2005;56(3):65–67. doi:10.1016/j.ejrex.2005.10.009.
16. Arráez-Aybar LA, Navia-Alvarez P, Mendez-Cendon JC. A case of a type II proatlantal artery with arteriovenous malformation. *Surg Radiol Anat*. 2011;33(1):85–89. doi:10.1007/s00276-010-0685-6.
17. Lode I. Persistence of both proatlantal arteries with occlusion of the internal carotid arteries. A case report. *Acta Radiol*. 2000;41(5):410–411.
18. Liechty JM, Weddle RJ, Shutze WP, Smith BL. Occurrence of a type 2 proatlantal intersegmental artery during carotid endarterectomy for symptomatic stenosis. *J Vasc Surg*. 2016;64(3):807–808. doi:10.1016/j.jvs.2014.10.100.
19. Pelz D, Menon RK, Swartz RH, Aviv RI, Symons SP. Bilateral type II persistent proatlantal intersegmental arteries. *Can J Neurol Sci*. 2013;40(06):873–874. doi:10.1017/S031716710001605X.
20. Obayashi T, Furuse M. The proatlantal intersegmental artery: a case report and review of the literature. *Arch Neurol*. 1980;37(6):387–389.
21. Lui CC, Liu YH, Wai YY, Tsai CC. Persistence of both proatlantal arteries with absence of vertebral arteries. *Neuroradiology*. 1987;29(3):304–305.
22. Sato H, Ogawa A, Kitahara M, Kayama T, Sakurai Y, Suzuki J. Persistent primitive first cervical intersegmental artery (proatlantal artery II) with occlusion of the basilar artery—a case report. *No to Shinkei [Brain and Nerve]*. 1988; 40(3):219–224.
23. Berman IG, Stuckey SL. Anomalous communication of external carotid and vertebral arteries with associated intracranial parenchymal arteriovenous malformation: Magnetic resonance angiography and angiographic findings. *Australas Radiol*. 2003;47(4):472–474.
24. Li TH, Lan MY, Liu JS, Tseng YL, Wu HS, Chang YY. Type II proatlantal intersegmental artery associated with objective pulsatile tinnitus. *Neurology*. 2008;71(4):295–296. doi:10.1212/01.wnl.0000318276.07527.70.