Type I Proatlantal Artery with Bilateral Absence of the External Carotid Arteries

C. Cinar Basekim, Emir Silit, Hakan Mutlu, M. Zekai Pekkafali, Ersin Ozturk and Esref Kizilkaya

AJNR Am J Neuroradiol 2004, 25 (9) 1619-1621
http://www.ajnr.org/content/25/9/1619

AJNR Am J Neuroradiol 25:1619–1621, October 2004

Case Report

Type I Proatlantal Artery with Bilateral Absence of the External Carotid Arteries

C. Cinar Basekim, Emir Silit, Hakan Mutlu, M. Zekai Pekkafali, Ersin Ozturk, and Esref Kizilkaya

Summary: During color Doppler examination of a 41-year-old man who presented with vertigo, a right vertebral artery could not be found. Both MR angiography and digital subtraction angiography revealed a large anastomotic vessel between the right internal carotid and vertebral artery. It was thought to be type I proatlantal artery. Furthermore, the external carotid arteries were bilaterally absent. Although each vascular anomaly mentioned above is rare, it even more rare for these variations to occur simultaneously.

Persistence of the proatlantal artery is a rare anomalous communication between the carotid and vertebrobasilar systems. Since it was first described in the 19th century, more than 40 cases have been reported in the literature. Although some vascular variations have been described coexisting with the persistent proatlantal artery, we have not encountered a case of bilateral absence of the external carotid arteries with a persistent proatlantal artery. We present a case of bilateral absence of the external carotid arteries, aplasia of the right vertebral artery, and hypoplasia of the left vertebral artery in combination with right type I persistent proatlantal artery.

Case Report

A 41-year-old man was admitted to the hospital because of vertigo. Vertebral artery Doppler examination was performed to exclude vertebrobasilar insufficiency. A hypoplastic left vertebral artery was seen on color Doppler examination. The right vertebral artery, however, could not be seen. Although consultation with the otorhinolaryngology department revealed the cause of vertigo as vestibular neuritis, angiographic examinations were performed to evaluate the vertebrobasilar system. An arch aortogram revealed a variation of the aortic arch in which a left brachiocephalic trunk gave off two terminal branches (the left common carotid artery and the left subclavian artery) after a short segment from its origin (Fig 1). Also, the right vertebral artery was not seen. Both MR angiography and digital subtraction angiography of the supraaortic vessels revealed a large anastomotic vessel connecting the right internal carotid artery and the right vertebral artery (Figs 2 and 3). It branched off from the right internal carotid artery at the C2 level, coursed dorsally at the skull base, and entered the skull via the foramen magnum. This large anastomotic vessel was thought to be a persistent proatlantal artery. The right vertebral artery was absent, and the left vertebral artery was hypoplastic. Furthermore, the external carotid arteries were absent bilaterally and the arterial branches that usually emanate from the external carotid artery arose from the common carotid artery directly (Fig 3). The right common carotid artery was larger than that on the left.

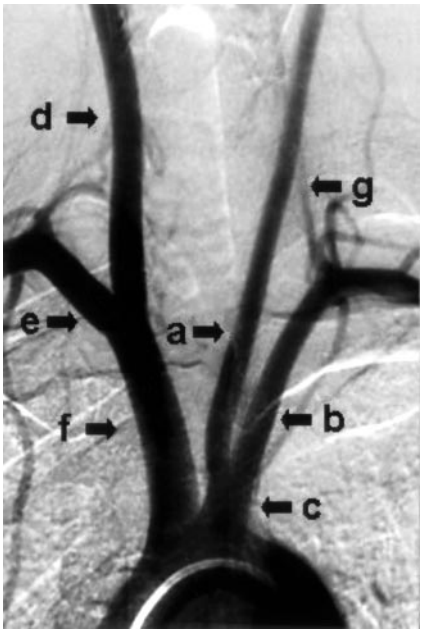

FIG 1. Arch aortogram in left anterior oblique projection shows both the left common carotid artery (*A*) and the left subclavian artery (*B*) originating from a left brachiocephalic trunk (*C*). Normal origin of both the right common carotid artery (*D*) and the subclavian artery (*E*) from the right brachiocephalic trunk (*F*) can be seen. Note the smaller size of the left hypoplastic vertebral artery (*G*).

Discussion

Primitive embryonic anastomotic vessels between the carotid and basilar arterial systems occasionally persist into adult life. There are four transient anastomoses between the posterior vascular plexus and the anterior carotid artery in early fetal life. By the 7–12-mm embryonic stage, these connections have involuted, with the proatlantal artery being the last to do so. Failure of involution of one of these connections causes four different types of anomalous arteries known as the persistent fetal anastomoses. These arteries are the trigeminal, otic, hypoglossal, and proatlantal arteries, from cephalic to caudal (1). They are named for the cranial nerves with which they run, except the proatlantal artery. The most commonly

Received January 22, 2004; accepted after revision February 16.

From the Department of Radiology, GATA Haydarpasa Training Hospital (C.C.B., E.S., H.M., M.Z.P., E.K.), Istanbul, Turkey; and the Department of Radiology, Aksaz Naval Hospital (E.O.), Mugla, Turkey.

Address correspondence to Ersin Ozturk, MD, Department of Radiology, Aksaz Naval Hospital, Marmaris 48700, Mugla, Turkey.

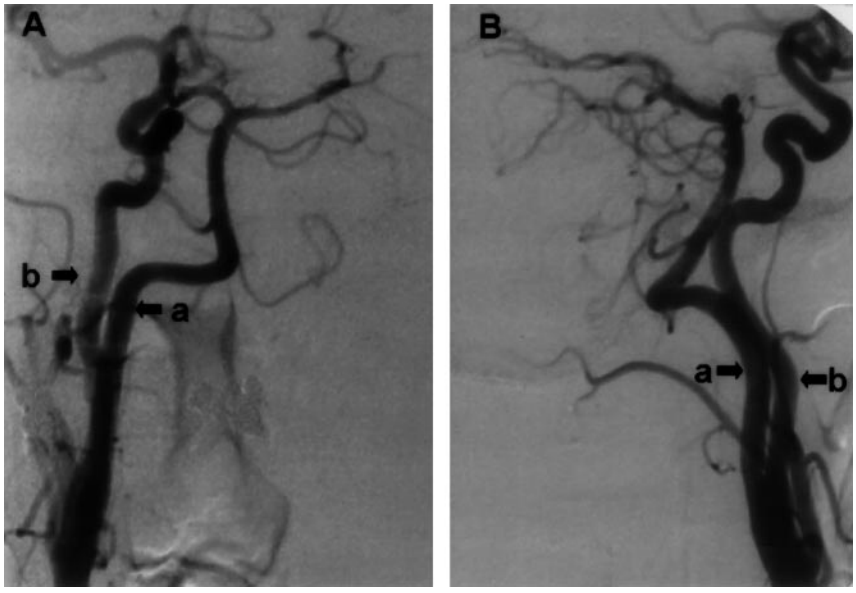

FIG 2. Selective right common carotid arteriograms show the type I proatlantal artery (*A*) originating from the right internal carotid artery (*B*) at the C2 level.
A, Anteroposterior projection.
B, Lateral projection.

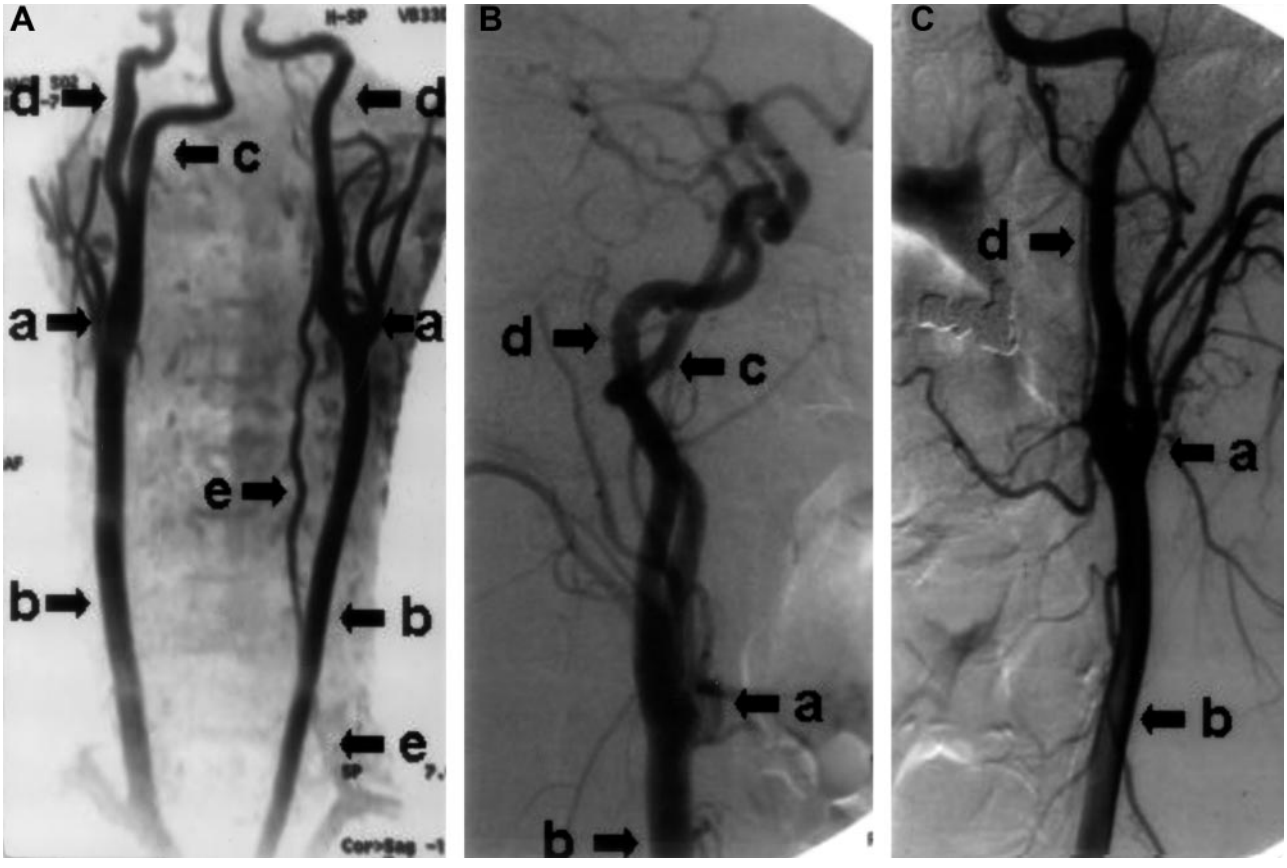

FIG 3. Both the contrast-enhanced 3D MR angiograms of the neck and the selective common carotid arteriograms show the branches of the external carotid arteries (*A*) originating directly from the common carotid arteries (*B*) at the C4 level on both sides. The type I proatlantal artery (*C*), the internal carotid arteries (*D*), and the left hypoplastic vertebral artery (*E*) can be seen.
A, Contrast-enhanced 3D MR angiogram.
B, Right selective common carotid arteriogram in lateral projection.
C, Left selective common carotid arteriogram in anteroposterior projection.

found persistent fetal anastomotic vessel is the trigeminal artery, followed by the hypoglossal, proatlantal, and otic artery.

Two types of proatlantal arteries have been described. The type I proatlantal artery (corresponding to the first segmental artery) arises from the internal carotid artery, as in the case herein presented, whereas the type II proatlantal artery (corresponding

to the second segmental artery) arises from the external carotid artery (2). Fifty-seven percent of the described proatlantal arteries are of the type I variety, 38% are type II, and 5% arise from the common carotid artery (3).

The persistent fetal anastomoses are usually large and are associated with hypoplasia or aplasia of the vertebral arteries. When the proatlantal artery is large, the vertebral arteries usually are hypoplastic and the ipsilateral vertebral artery may be absent (4). These findings are consistent with the MR angiography and digital subtraction angiography findings in our case.

Differentiation between the type I proatlantal artery and the more common hypoglossal artery requires careful analysis because of their similar origin from the internal carotid artery (5). The typical origin of the proatlantal artery is at the level of C2 or C3, whereas the hypoglossal artery originates from the internal carotid artery at the level of C1–C3. The persistent proatlantal artery in our case may be distinguished from the hypoglossal artery by its passage through the foramen magnum rather than the hypoglossal canal or by its dorsal course at the skull base, as opposed to the hypoglossal artery's relatively vertical course (5).

The common carotid artery, as a rare occurrence, may ascend in the neck without a bifurcation into either the external or the internal carotid artery. The absence of the external carotid artery has been explained by the fact that the development of the external carotid artery arising from the ventral aorta is disturbed by unknown factors in the early embryonic stage. In the process of embryonic development, the internal carotid artery and branchial arteries (that are to be components of the external carotid artery) have remained without connection and without development of the external carotid artery trunk (6). The branches usually derived from the external carotid artery arose directly from the upward continuation of the common trunk.

The external carotid artery may be absent bilaterally, as in our case, or unilaterally. We were not able to find any case of bilateral absence of the external carotid artery in our review of the literature, although we encountered seven cases of unilateral absence of the external carotid artery (6–12).

Many variations occur in the number and position of vessels arising from the aortic arch. There may be as few as one or as many as six branches. The branching order was the right subclavian artery and the right common carotid artery from the right brachiocephalic artery, and the left common carotid artery and the left subclavian artery from a common trunk in our case. The common trunk on the left side is named as left brachiocephalic trunk. This anomaly has been described as biinnominate artery variation, and it represents fewer than 3% of all the aortic arch variations (13).

Conclusion

Our case had triple developmental anomalies, bilateral absence of the external carotid arteries, left brachiocephalic trunk, and type I proatlantal artery. Although these anomalies are usually asymptomatic, and found incidentally, their importance may be important in diagnosis and therapy.

References

1. Patel AB, Gandhi CD, Bederson JB. **Angiographic documentation of a persistent otic artery.** *AJNR Am J Neuroradiol* 2003;24:124–126
2. Lasjaunias P, Theron J, Moret J. **The occipital artery: anatomy: normal arteriographic aspects: embryological significance.** *Neuroradiology* 1978;15:31–37
3. 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;117:232–239
4. Fujita N, Shimada N, Takimoto H, Satou T. **MR appearance of the persistent hypoglossal artery.** *AJNR Am J Neuroradiol* 1995;16: 990–992
5. Anderson RA, Sondheimer FK. **Rare carotid-vertebrobasilar anastomoses with notes on the differentiation between proatlantal and hypoglossal arteries.** *Neuroradiology* 1976;11:113–118
6. Nakaoka T, Matsuura H. **Unilateral absence of the external carotid artery: three case reports [in Japanese].** *No Shinkei Geka* 2002;30:1337–1342
7. Thomas RL, Mitchell DP. **Facial paralysis with absence of internal and external carotid arteries.** *J Otolaryngol* 1980;9:353–356
8. Kaneko K, Akita M, Murata E, et al. **Unilateral anomalous left common carotid artery: a case report.** *Anat Anz* 1996;178:477–480
9. Franklin PD, Lee RG, Allard JC, et al. **Unilateral absence of the external carotid artery.** *Can Assoc Radiol J* 1988;39:293–294
10. Kunishio K, Yamamoto Y, Sunami N, Asari S. **Agenesis of the left internal carotid artery, common carotid artery, and main trunk of the external carotid artery associated with multiple cerebral aneurysms.** *Surg Neurol* 1987;27:177–181
11. Teal JS, Rumbaugh CL, Bergeron RT, Segall HD. **Congenital absence of the internal carotid artery associated with cerebral hemiatrophy, absence of the external carotid artery, and persistence of the stapedial artery.** *Am J Roentgenol Radium Ther Nucl Med* 1973;118:534–545
12. Rodriguez HE, Ziauddin MF, Podbielski FJ, et al. **Congenital absence of the external carotid artery: atherosclerosis without a bifurcation.** *J Vasc Surg* 2002;35:573–575
13. Kadir S. **Regional anatomy of the thoracic aorta.** In: Kadir S, ed. *Atlas of normal and variant angiographic anatomy.* 1st ed. Philadelphia: WB Saunders;1991:19–54