A Five-Vessel Aortic Arch with an Anomalous Origin of Both Vertebral Arteries and an Aberrant Right Subclavian Artery

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Summary

In this case presentation we describe a patient with an anomalous origin of the right vertebral artery arising from the right common carotid artery in combination with an aberrant right subclavian artery and a left vertebral artery originating from the arch between the left common carotid artery and left subclavian artery. Hence there were five vessels originating from the aortic arch. The possible embryological mechanism as well as a postulation on the importance of the level of entrance of the vertebral artery in the cervical transverse foramen is discussed.

Introduction

An anomalous origin of the vertebral artery on either side is a rare finding as pointed out in a review of the literature by Lemke et Al in 1999¹. The most common vertebral artery variation is the origin of the left vertebral artery (LVA) directly from the aortic arch, between the left common carotid artery (LCCA) and left subclavian artery (LSCA). The second most common variation is the origin of the right vertebral artery (RVA) from the right common carotid artery (RCCA) combined with an aberrant right subclavian artery (ARSCA). The first variation has been described in 15 angiographic case reports until 1999. The second variation

was documented in only ten case reports¹. In this paper we describe a patient with a combination of the two variations. A five-vessel arch is reported to have a frequency of 0.1% but the combination with an anomalous origin of the right vertebral artery is even less common and has not been previously reported².

History

A 68-year-old female presented to the clinic with headaches and symptoms of transient ischemic attacks. The initial CT and MRI showed no abnormalities. A second evaluation by MRI showed signs of iron (hemosiderin) deposition or pial siderosis along the sulci of the right cerebral hemisphere. An MRA was subsequently performed and showed neither signs of intracranial vascular abnormalities that would explain the clinical symptoms nor the cause of the pial siderosis. An incidental finding, however, was the anomalous origin of the right and left vertebral arteries. The RVA originated from the common carotid artery whereas the LVA originated from the aortic arch. Furthermore, the right subclavian artery had an aberrant origin distal to the left subclavian artery. (figures 1,2) Angiography was performed to exclude a source of previous SAH.

The source images of the MRA as well as the unsubtracted images of the angiography showed

Figure 1 Angiography of the aortic arch showing the five-vessel origin.

Figure 2 3D MRA reconstruction showing the five-vessel aortic arch.

that both vertebral arteries entered the transverse foramen at the level of C4. Other findings included a fetal PCA on the left with a caudal fusion of the basilar artery ending in the superior cerebellar artery on the left, and a normal P1-segment of the PCA on the right. The right vertebral artery segment distal to the PICA origin, was hypoplastic and showed to be the unilateral supply to the anterior spinal artery.

Normal Development

The development of the vertebral arteries is well described in literature^{3,4}. The vertebral arteries are the result of persisting longitudinal anastomoses between the cervical intersegmental arteries. The cervical intersegmental arteries are present on both sides of the longitudinal anastomoses as shown in figure 3. The intersegmental cervical arteries will regress with an exception for the seventh, which will eventually form the subclavian artery. The longitudinal anastomoses are continuous and connect to the seventh intersegmental artery; hence the vertebral artery will be connected to the subclavian artery in normal development (figure 4).

In order to facilitate the discussion at the end of this paper we propose to make a distinction between the two channels on either side of the longitudinal anastomoses. We suggest naming them the 'medial-' and the 'lateral' intersegmental cervical arteries. In this definition the medial cervical intersegmental artery is located between the longitudinal anastomoses and the IVth aortic arch. The lateral intersegmental cervical artery is located on the opposite side of the longitudinal anastomoses as pointed out in figure 3.

Variants in the Literature

Lemke et al gave an overview of the published variants until 1999. He presented a case of a patient with a right vertebral artery being the last artery to arise from the aortic arch, distal to the left subclavian artery, and postulated that this was the result of a persisting eighth intersegmental artery, which is caudal from the obliteration zone (region of involution in figure 3) and results in a connection to the left descending aorta¹.

In the literature of the last five years several cases on the anomalous origin of unilateral or bilateral vertebral arteries have been published:

Figure 3
Schematic overview
of embryo at 14 days
of gestation.

Figure 4
Normal development
of right vertebral
artery after regression
of the intersegmental
arteries (grey area).

Figure 5
Development of the right vertebral artery originating from the C-IV lateral cervical intersegmental artery after regression of the other intersegmental arteries. This origin is considered metameric and would originate from the ascending cervical artery.

Figure 6
Development of the right vertebral artery originating from the common carotid artery due to persistence of the medial C-V cervical intersegmental artery after regression of the other intersegmental arteries.

Glunic et Al described an anomalous origin of both vertebral arteries in one patient⁵. The left vertebral artery originated from a common trunk with the left subclavian artery and the right vertebral artery originated from the right common carotid artery associated with an aberrant right subclavian artery. Both carotid arteries shared a common trunk. Unfortunately it is not absolutely clear from the paper at what level the vertebral arteries entered the transverse foramina since the description is conflicting in the text and the table. The right vertebral artery is described to enter the third transverse foramen twice in the text but it is said to enter the second transverse foramen in the accompanying table. Because of a lacking C7 vertebral body in the figure in the article, this is also not helpful in deciding on the actual level.

Albayram et Al described a case with bilateral arch origin of the vertebral arteries between the left common carotid and left subclavian artery⁶. They hypothesized that this was related to a persisting proximal dorsal aorta on the right with regression of the right dorsal aorta between the sixth and seventh intersegmental arteries. The left vertebral artery origin is from the arch because of the persistence of the sixth rather than the seventh intersegmental artery.

Best et Al portrayed a case of an anomalous right vertebral artery origin from the right common carotid artery in combination with an aberrant origin of the right subclavian artery². This aberrant right subclavian had its origin from the arch between the left common carotid and the left subclavian. No mechanism for this anomaly was suggested. The left vertebral artery originated from the left subclavian artery.

Koenigsberg et Al showed five cases of which two had an anomalous origin of the right vertebral artery⁷. One of those arteries originated from the thyrocervical trunk whilst the other originated from the common carotid artery. The latter case did not have an anomalous origin of the left vertebral artery, like in our case. Only limited explanation for the anomalies are given in the report.

In 1968 Lie described the carotid-vertebral anastomoses in an overview of the congenital anomalies of the carotid arteries⁸. One of his drawings showed that the persistence of a cervical intersegmental artery might lead to a vertebral artery originating from the common carotid artery.

Theoretical Background for the Discussion

From the literature it is suggested that in the case of an anomalous origin of the vertebral artery the vessel responsible for taking over this origin is likely to be one of the metameric arteries³. Which metameric artery is involved can be difficult to assess but we postulate that it might be deduced from the level of entrance in the transverse foramina.

To support this postulation, some definitions and theories will have to be addressed first:

Phylogenetically the occipital bone is to be considered as the first cervical vertebral body. Given this assumption, the atlas (C1) will be the second cervical vertebral body and will henceforth be named C-II. Correspondingly the remaining vertebral bodies will be named C-III for C2, C-IV for C3 through C-VIII for C7. This is in keeping with the nerve root exiting below the vertebral body (i.e. nerve root C8 at the level C-VIII) and hence compliant with the metameric distribution.

In the proatlantal I variant, which is believed to be the persisting first intersegmental cervical artery, the artery will enter the foramen of C-I which is the foramen magnum. The proatlantal II variant, being the persisting second intersegmental artery, is entering the transverse foramen of the atlas (C-II).

Considering the metameric distribution, the ascending cervical artery is believed to be the metameric artery of C-III and C-IV and the costocervical (deep cervical) artery is regarded as the metameric artery of C-V and C-VI as proposed by Lasjaunias et Al³. The evolution of this metameric origin is depicted in figure 5. Lasjaunias et Al also recognized that the C-VII metamere corresponds to the proximal course of the vertebral artery. In normal development the vertebral artery is therefore expected to enter the transverse foramen at C-VII (i.e. C6).

From this, one can postulate that the level of entrance in the transverse foramen is related to the intersegmental cervical artery or metamere of which the "anomalous vertebral artery" originates.

Correlation with our Case and Discussion

In our case both vertebral arteries enter the transverse foramen at the C4 level, which is phylogenetically the C-V level. This would suggest that the anomalous vertebral artery origi-

nates from a persisting fifth intersegmental artery, corresponding to the deep cervical artery from the costocervical trunk. Lasjaunias described in his 'variations of the vertebral arteries' that: "the vertebral artery entering the spinal canal at C5, C4 or C3 corresponds to the ascending cervical artery, which is not found as a separate vessel in these situations"⁹. This corresponds to the case Koenigsberg described with the right vertebral artery originating from the thyrocervical trunk⁷.

In our case however, contrast injection of the left subclavian artery showed a thyrocervical trunk with the normal ascending cervical artery arising. The costocervical trunk is also visible and suggests presence of the deep cervical artery. If the postulation, that we can predict the metameric artery involved in the anomalous origin, were true, we would expect absence of either the deep cervical or the ascending cervical artery in our case.

A possible explanation for this discrepancy is the fact that it is not the lateral intersegmental cervical artery that persists in this case, but rather the medial intersegmental cervical artery. This would also explain the origin of the right common carotid artery since the persisting medial intersegmental artery is connected to the IVth aortic arch (figure 6) that eventual-

ly will become part of the common carotid artery or right brachiocephalic trunk. This change in origin from lateral to medial is supported by the Glunic case where the origin was also of the common carotid with an associated entrance at the C2 or C3 transverse foramen.

Conclusions

In this case presentation the abnormal origin of both vertebral arteries is used to describe the mechanism involved in the development of these anomalies.

It is postulated that in the case of 'medial C-IV to C-VI intersegmental cervical artery' persistence, there will be an origin from the common carotid artery. If, on the other hand, the 'lateral C-IV to C-VI intersegmental cervical artery' persists, this will lead to an origin from the metameric artery, being either the deep cervical artery or ascending cervical artery. It is therefore suggested to make a distinction between the medial- and lateral intersegmental cervical arteries in the description of embryology and developmental variants of the vertebral arteries. Furthermore the level of entrance in the transverse foramen is likely to be associated to the actual level of the persisting intersegmental cervical artery.

References

- 1 Lemke AJ, Benndorf G et Al: Anomalous origin of the right vertebral artery: review of the literature and case report of right vertebral artery origin distal to the left subclavian artery. *Am J Neuroradiol* 20: 1318-1321, 1999.
- 2 Best IM, Bumpers HL: Anomalous origin of the right vertebral, subclavian and common carotid arteries in a patient with a four-vessel aortic arch. *Ann Vasc Surg* 16: 231-234, 2002.
- 3 Lasjaunias P, Berenstein A, Ter Brugge KG: Surgical neuro-angiography I, clinical vascular anatomy and variations. 2nd Edition. Springer Verlag, New York, 2001.
- 4 Osborn AG: Diagnostic Cerebral Angiography, 2nd Edition. Lippincott Williams & Wilkins, Philadelphia, 1999.
- 5 Glunic V, Ivkic G et Al: Anomalous origin of both vertebral arteries. *Clin Anat* 12: 281-284, 1999.
- 6 Albayram S, Gailloud P, Wasserman BA: Bilateral arch origin of the vertebral arteries. *Am J Neuroradiol* 23: 455-458, 2002.
- 7 Koenigsberg RA, Pereira L et Al: Unusual vertebral artery origins: examples and related pathology. *Cath Card Vasc Int* 59: 244-250, 2003.
- 8 Lie TA: Congenital anomalies of the carotid artery. Excerpta Medica Foundation, Amsterdam, The Netherlands 1968.
- 9 Lasjaunias P, Berenstein A: Surgical neuro-angiography III, functional vascular anatomy of the brain, spinal cord and spine. Springer Verlag, Berlin, 1990.

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