

Rare Case of Absent Common Carotid Artery Likely Associated with Intracranial Aneurysm Development

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Congenital absence of the common carotid artery without separate origin of internal carotid artery and external carotid artery from the aortic arch is a very rare anomaly. A persistent proatlantal intersegmental artery into adult life is also a rare condition. We report a patient with the coexistence of such rare conditions who presented with cerebral infarctions and intracranial aneurysms.

Key Words: Common carotid artery—Agenesis—Fetal intracranial artery—Aneurysm
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Introduction

Absence of the common carotid artery (CCA) is a rare condition. Most cases of absent CCA are associated with separation of the internal carotid artery (ICA) and external carotid artery (ECA) from the aortic arch.¹ Absence of the CCA without separate origins of the ICA and ECA is very rare. A persistent proatlantal intersegmental artery serving as a main supplier to the ECA is also rare. We present a case involving such rare vascular anomalies combined in a patient with cerebral infarctions and unruptured intracranial aneurysms. These findings are extremely rare. Our report is the first case of these anomalies with intracranial aneurysms and cerebral infarctions.

Case Presentation

A 57-year-old woman with an unremarkable medical history except hypertension presented with a 2-day history of blurred vision. Her physical and neurological examinations were normal. Diffusion weighted magnetic resonance

imaging showed multiple acute cerebral infarctions in the territory of the right posterior cerebral artery (Fig 1, A). Computed tomography angiography revealed a hypoplastic left ICA and 2 aneurysms, which were located at the top of the basilar artery (BA) and the proximal right A1 segment of the anterior cerebral artery (Fig 1, B), respectively. Axial computed tomography on the skull base showed a significantly narrow left carotid canal (Fig 1, C). Digital subtraction angiography was performed, and it demonstrated an absence of the left CCA from the aortic arch (Fig 1, D). There was an abnormal anastomosis between the left vertebral artery and left proximal ECA, considered to be a persistent proatlantal intersegmental artery (Fig 1, E). Through this artery, the left vertebral artery supplied blood flow to the left ECA. The left anterior intracranial circulation, except territory of the anterior cerebral artery, was mainly supplied from the posterior circulation via the left posterior communicating artery. The contralateral ICA partly contributed to the left middle cerebral artery territory through the anterior communicating artery.

The BA aneurysm was treated with stent-assisted coiling and the right anterior cerebral artery aneurysm was treated with simple coiling. Follow-up digital subtraction angiography after 1 year revealed recanalization of the BA aneurysm. Further endovascular coiling for this aneurysm was done without any complication.

Discussion

Agenesis is failure of an organ to develop during embryonic growth because of an absence of primordial tissue. The exact definition of CCA agenesis has not been

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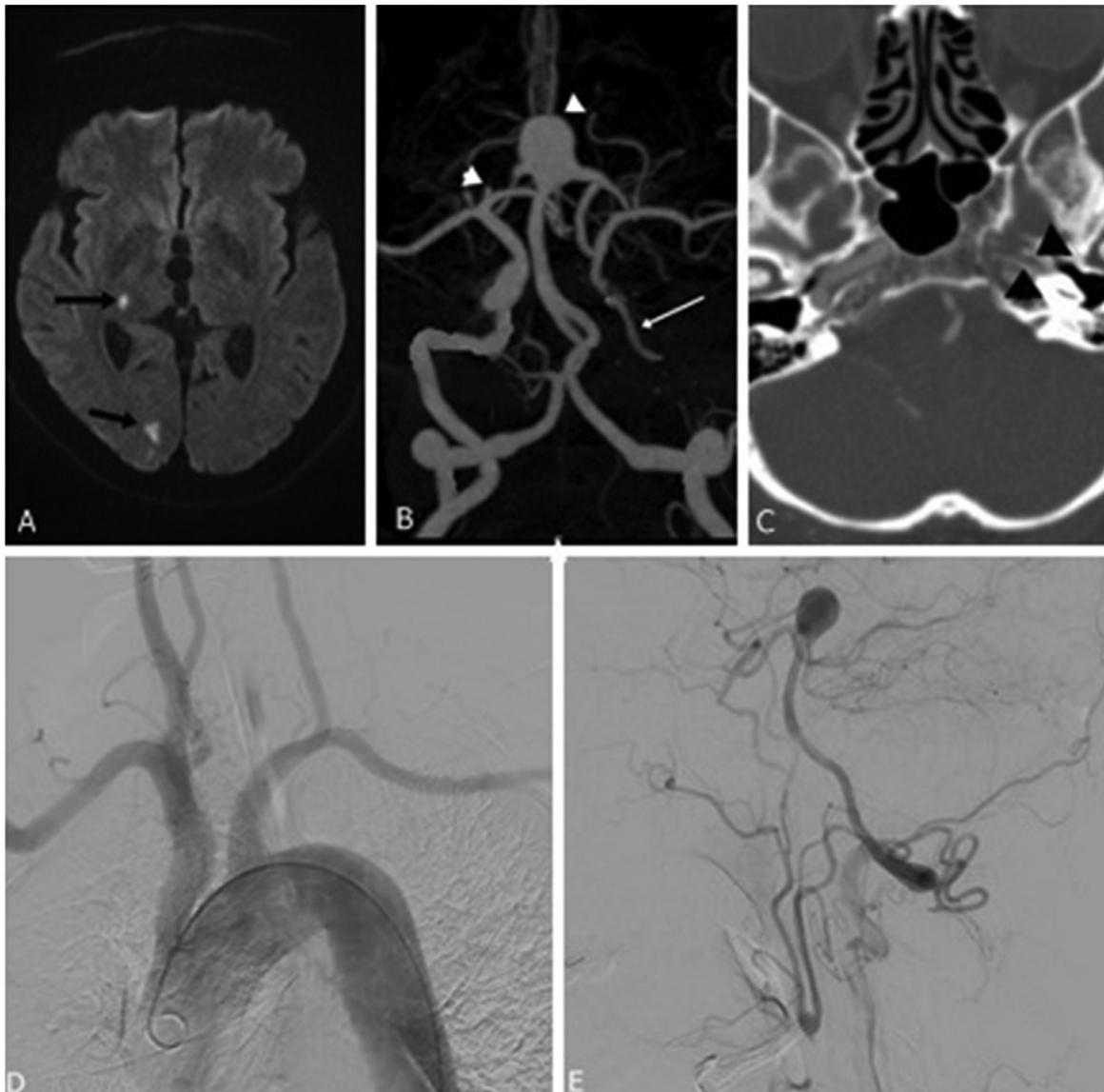


Figure 1. (A) Diffusion-weighted magnetic resonance imaging demonstrates acute infarction in the right occipital lobe and right thalamus (black arrows). (B) Computed tomography angiography shows asymmetric decreased caliber of the left internal carotid artery (white arrow) and 2 aneurysms (white arrowheads). (C) Axial computed tomography shows hypoplastic left carotid canal (black arrowheads). (D) Aortogram shows absence of the left common carotid artery. (E) Left vertebral artery angiography reveals persistent proatlantal intersegmental artery which supplies blood to the external carotid artery.

established, but the lexical meaning will refer to a congenital complete absence of CCA that is not acquired. With respect to the acquired or congenital nature of a missing carotid artery, hypoplasia or aplasia of the bony carotid canal indicates congenital abnormalities.² In addition, persistent primitive carotid-basilar anastomoses are often related to congenital conditions.^{3,4} Therefore, our case is likely to have occurred congenitally. However, we cannot completely exclude the existence of a fibrous remnant of the artery or primordial tissue. That could only be confirmed by surgical exploration. Therefore, although the terms “congenital” and “complete absence” are correct in our case, the use of the term “agenesis” may be controversial in terms of using the correct definition.

The most common finding of absent CCA is separate origins of the ECA and ICA from the brachiocephalic trunk or the aortic arch. However, in our case, absent CCA did not arise from separation of the ECA or ICA, and there was a lack of connection between the aorta and carotid system. This difference can be explained in terms of embryologic aortic arch development. The CCA derives from the third aortic arch and ventral aorta, and it develops as a result of regression of the first 2 aortic arches and dorsal aorta between the third and fourth arches (the ductus caroticus). Abnormal regression of the third arch before obliteration of the ductus caroticus can result in absent CCA with separate origins of the ECA and ICA. In this case, the ductus caroticus becomes the proximal ICA

and supplies blood to the intracranial region. Whereas, when both the ductus caroticus and proximal carotid system are regressed, there is no connection between the aorta and carotid system, as in our case.

The type of anomaly of the carotid system may also be determined depending on the site where the abnormal regression occurred. In our case, because carotid bifurcation and the ICA were retained, although they were hypoplastic, abnormal regression is presumed to have occurred in the ventral aorta or proximal part of the third aortic arch. Chan et al⁵ reported a case of a lack of communication between the CCA and cervical ICA with a persistent proatlantal intersegmental artery, but the ECA was normally from the CCA. In this case, unlike in our case, it seems that the disruption had occurred in a more distal part of the third aortic arch.

The primitive intracranial anastomoses (trigeminal, otic, hypoglossal, proatlantal intersegmental artery) normally regress after formation of the posterior communicating artery in the early embryonic stage. These primitive anastomoses could persist into adult life, when the carotid system is incompletely developed.³ Cali et al hypothesized that the anastomoses that persist may be a function of the time when disruption to the embryonic carotid system occurs. Disruptions that occur before the 24-mm embryonic stage may result in persistent primitive anastomoses, whereas disruptions that occur after this stage may result in persistent anastomoses via the circle of Willis.⁶ In our case, the left anterior intracranial circulation is supplied through the circle of Willis, but the proatlantal intersegmental artery is also persistent. We speculate that after regression of the ductus caroticus and proximal carotid system, the proatlantal intersegmental artery mainly supplied blood to the affected side at an early stage, and later the role of supplying blood through ICA was replaced by the posterior communicating artery. These unusual vascular connections have reduced the role of the ICA and presumably resulted in a hypoplastic ICA.

The combined vascular anomalies reported in this case have been reported only twice before. Cao et al⁷ reported that a 42-year-old male presented with absent CCA and a proatlantal intersegmental artery. The patient had dizziness, but the history was otherwise uneventful. In the second case, Quinones et al¹ reported absence of the CCA with a proatlantal intersegmental artery in a 28-year-old male. The patient presented with a post-traumatic headache. In both cases, the vascular anomalies had no symptoms and were an incidental finding. Our report is the first case of these anomalies with intracranial aneurysms and cerebral infarctions.

There is an increase in the rate of aneurysm development associated with the congenital absence or hypoplasia of one of carotid systems.⁸ The aneurysm is often on the pathway providing collateral circulation because there is the additional hemodynamic stress due to increased blood flow and altered flow direction. In our case, there

was an aneurysm in the right A1 and an aneurysm at the top of BA. The BA aneurysm in our case recurred. This is significant because it may be the case that the additional hemodynamic stress potentially resulting from absent CCA with a hypoplastic ICA may have increased the likelihood of the aneurysm's recurrence.

Acute cerebral ischemia is common in patients with ICA aplasia or hypoplasia.⁸ Altered hemodynamics that result from abnormalities of the carotid system and cerebral vessels could be potential causes of cerebral ischemia. Further, the consequences of an ischemic stroke could be affected by such factors that include morphological or hemodynamic changes, failures of mechanisms to compensate for such changes, and a potential decrease in the efficacy of the intracranial collateral circulation. However, in our case, the territory of cerebral infarction was in the territory of the right posterior cerebral artery, which is a different area from the vascular abnormalities. This may be caused by blood flow reduction in the right posterior cerebral artery due to a compensation mechanism for supplying the left anterior circulation. The large BA aneurysm may also have caused a cerebral infarction. There have been several reports of ischemic strokes associated with large cerebral aneurysms.^{9,10} Therefore, the exact cause of the cerebral infarctions in our case is unclear.

Conclusion

This report describes an extremely rare anomaly of the carotid system with a persistent proatlantal intersegmental artery. In congenital absence of the CCA or ICA in the carotid system, primitive anastomoses may persist into adulthood, and care should be taken when intervention or neck surgery is performed. It should also be noted that this condition may cause aneurysms or cerebral infarctions due to altered hemodynamics.

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