



Coexistence of the Absence of the Left Common Carotid Artery, a Common Origin of the Left External Carotid Artery and the Right Common Carotid Artery, and an Aberrant Right Subclavian Artery: A Case Report

좌경동맥 결손, 좌외경동맥과 우경동맥 공통줄기 기형 및 이상 우쇄골하동맥의 동반 발생: 증례 보고

Soo Jeong Lee, MD , Hye Jeong Choi, MD* , Sang Heum Kim, MD

Department of Radiology, CHA Bundang Medical Center, CHA University, Seongnam, Korea

The absence of the common carotid artery (CCA) and the common origin of the left external carotid artery (ECA) and the right CCA are rare anomalies of the cervical vascular system. We report here a case involving the coexistence of these vascular anomalies with an aberrant right subclavian artery, which is a common congenital anomaly in the aortic arch, and review the embryologic mechanism and clinical importance of this case.

Index terms Common Carotid Artery; External Carotid artery; Internal Carotid Artery; Aberrant Right Subclavian Artery; Computed Tomography Angiography; Magnetic Resonance Angiography

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*Corresponding author
Hye Jeong Choi, MD
Department of Radiology,
CHA Bundang Medical Center,
CHA University, 59 Yatap-ro,
Bundang-gu, Seongnam 13496,
Korea.

Tel 82-31-780-6138
Fax 82-31-780-5381
E-mail isahj99@gmail.com

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ORCID iDs

Soo Jeong Lee
<https://orcid.org/0000-0003-3712-4077>
Hye Jeong Choi
<https://orcid.org/0000-0002-4103-769X>
Sang Heum Kim
<https://orcid.org/0000-0001-7504-5148>

INTRODUCTION

The absence of the common carotid artery (CCA) is a rare anomaly of the vascular system. The incidence of agenesis of the CCA is unknown, and only 87 cases have been reported up to 2019 (1, 2). This anomaly can occur at both sides and generally coexist with independent origins of the internal carotid artery (ICA) and external carotid artery (ECA) (2), and the common origin of the ECA and the contralateral CCA is even rare. We present a case of coexistence of these vascular anomalies with an aberrant right subclavian artery, which is a common congenital anomaly in the aortic arch (3), and reviewed the embryologic mechanism and clinical importance.

CASE REPORT

A 39-year-old-female was admitted to undergo total hip arthroplasty for secondary osteoarthritis of the right hip. She had mental retardation and congenital hearing loss. During her pre-operative evaluations, she underwent brain MRI and MR angiography (MRA) for these neurological problems. MRA incidentally showed the absence of the left CCA, a common origin of the left ECA, and the right CCA in a low bifurcation of the right CCA. Additionally, there was a retroesophageal aberrant right subclavian artery arising from the left descending thoracic aorta and an aberrant origin of the left vertebral artery from the aortic arch. After the hip surgery, she had no neurological complications except for mild headache. For further evaluation, the patient underwent CT angiography (CTA) (Fig. 1A-E), and there was no significant difference from the MRA. She was managed conservatively for headache and was discharged after post-operative care.

DISCUSSION

CCAs are paired structures that normally originate from the aorta for the left CCA and brachiocephalic trunk for the right CCA and ends at the carotid bifurcation forming the ICA and ECA. The absence of CCA is a rare vascular anomaly. The incidence of the absent CCA is unknown, but only 87 cases have been reported up to 2019 (2). It can occur at both sides and bilaterally (right:left:bilateral:unknown laterality = 47.1%:39.1%:6.9%:6.9%) (2). Several cases accompanied by these vascular anomalies have been reported. Goyal and Sinha (1) reported the case of an absent left CCA with the bovine origin of the left ECA, and Yang and Lee (4) reported the coexistence of an aberrant right subclavian artery and the common carotid trunk. To our knowledge, there has been no report of the coexistence of the absence of the left CCA, common origin of left ECA and right CCA, and aberrant right subclavian artery.

The absence of CCA has been explained to be related to abnormal embryonic developmental processes including the aortic arch (5). At first, a pair of longitudinally directed channels arises in the paramedian location to form dorsal aortae on the 19th day of an embryo. From days 21 to 25th days, the ventral aorta sac connects to the dorsal aortae via the paired first aortic arches. From days 21 to 32nd days, six paired aortic arches have formed with five branchial arches and regress during embryonal development. This results in the development of

Fig. 1. Absence of the CCA and coexistent vascular anomalies in a 36-year-old female.

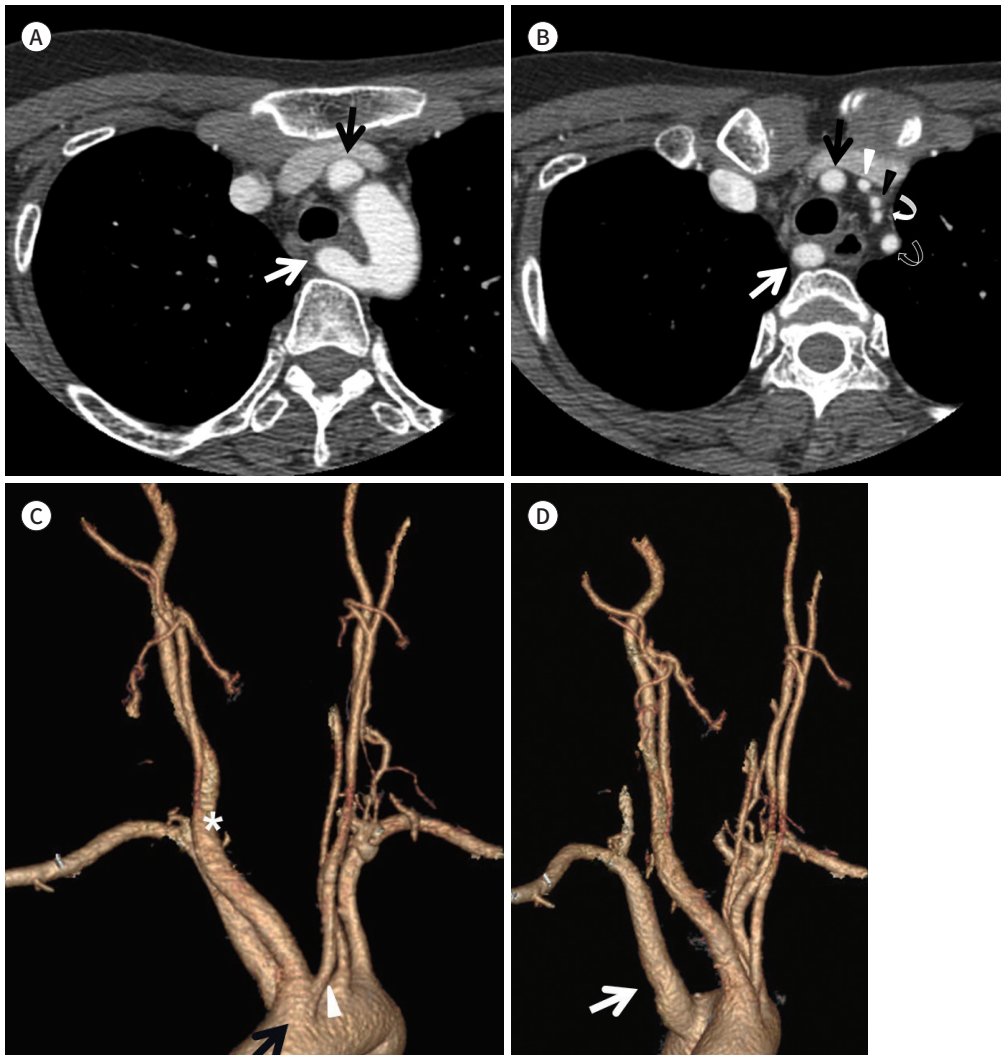
A. CT angiography axial image shows the aberrant right subclavian artery (white arrow) and the right CCA (black arrow) in the aortic arch level.

B. The right CCA (black arrow), the left ECA (white arrowhead), the left ICA (black arrowhead), the left vertebral artery origin (white curved arrow), the left subclavian artery (black curved arrow) and the aberrant right subclavian artery (white arrow) are shown.

C. CT angiography volume rendering image shows the common origin of the right CCA (black arrow) and the left bifurcation of the right CCA (white asterisk).

D. The aberrant right subclavian artery (white arrow) directly originated from the aortic arch.

CCA = common carotid artery, ECA = external carotid artery, ICA = internal carotid artery



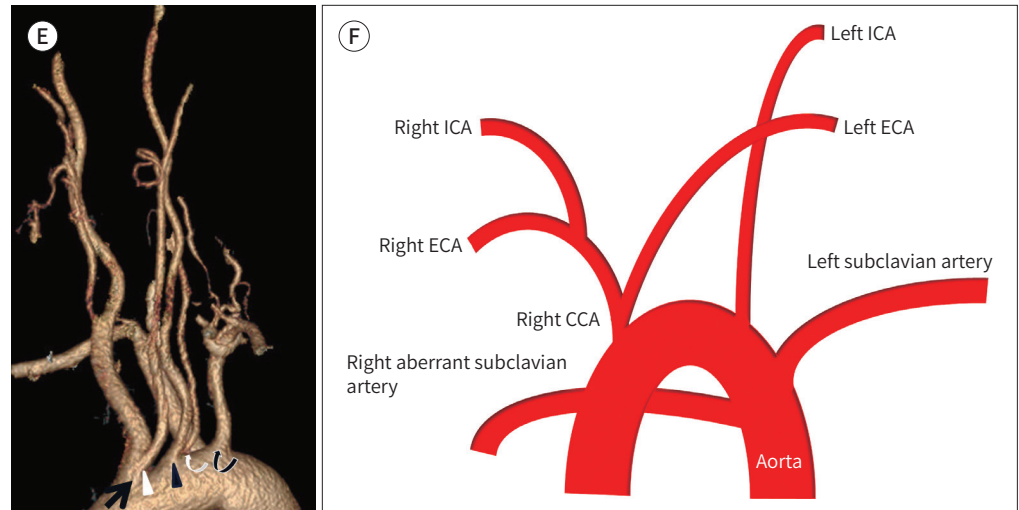
the carotid system. In a normal developmental process, the ductus caroticus, which is a dorsal aorta between the third and fourth arches, regress by 6 weeks, and the combination of remaining structures such as the ventral aortic sac, the third aortic arch, and the dorsal aorta rostral to the arch forms the CCAs and ICAs. The third arches are precursors to the carotid system, and the CCA normally develops from the root of the paired ventral aortae between the third and fourth arches. However, if the ductus caroticus persists with regression of the third arch or the fourth arch regress, the CCA was not formed (5, 6). The absence of CCA can coexist with other vascular anomalies such as double aortic arches, cervical aortic arch,

Fig. 1. Absence of the CCA and coexistent vascular anomalies in a 36-year-old female.

E. The common origin of the right CCA (black arrow) and the left ECA (white arrowhead), the left ICA (black arrowhead), the left vertebral artery origin (white curved arrow), and the left subclavian artery (black curved arrow) that originated from the aortic arch are also demonstrated.

F. A schematic illustration of the present case is shown.

CCA = common carotid artery, ECA = external carotid artery, ICA = internal carotid artery



nal artery, or cerebral venous sinus thrombosis (1).

Additionally, the coexisting aberrant right subclavian artery is one of the variant branching patterns of the aortic arch presented as the last branch of the aortic arch. Its prevalence is approximately 1% (range between 0.5% and 1.5% in the general population) (3, 7). Its development is explained by the abnormal regression of the right aortic arch. Embryologically, the fourth pair of aortic arches appears shortly after the third pair of arches at the end of the fourth week, and they develop differently between the right and left sides. On the right side, the fourth arch forms the proximal portion of the right subclavian artery and is continuous with the seventh segmental artery (8). The normal right subclavian artery developed from the distal fusion of a persistent right proximal dorsal aorta with the right seventh intersegmental artery. Therefore, involution of the right fourth vascular arch and proximal right dorsal aorta with the persistence of the seventh intersegmental artery results in the aberrant origin of the right subclavian artery (9). Although most cases with aberrant right subclavian artery are asymptomatic, the retroesophageal course of this artery might result in dysphagia (7). It can coexist with the truncus bicaroticus, Kommerell's diverticulum, and aneurysmal formation with a relevant risk of rupture (3).

Most carotid system anomalies are typically asymptomatic unless associated with a concomitant arterial lesion that results in any neurological deficits to be investigated (6). The absence of CCA is a usually incidental finding because there are no definite symptoms or signs caused by this anomaly. Therefore, it can be difficult to interpret when we face it. In our case, the patient had congenital hearing loss and mental retardation as neurological problems.

In conclusion, we report a case of absence of the left CCA with a common origin of the left ECA and the right CCA coexisting with the aberrant right subclavian artery. It can help understand the embryology and evaluate the vascular anomalies in CTA or MRA.

Author Contributions

Conceptualization, C.H.J., K.S.H.; investigation, all authors; supervision, C.H.J.; visualization, C.H.J., L.S.J.; writing—original draft, C.H.J., L.S.J.; and writing—review & editing, C.H.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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좌경동맥 결손, 좌외경동맥과 우경동맥 공통줄기 기형 및 이상 우쇄골하동맥의 동반 발생: 증례 보고

이수정 · 최혜정* · 김상흠

경동맥 무발생 및 좌외경동맥과 우경동맥 공통줄기기형의 동반 발생은 매우 드문 선천적 목혈관기형이다. 저자들은 위의 드문 기형들과 함께 흔한 대동맥궁 기형인 이상 우쇄골하동맥이 발생한 증례를 경험하였고 발생학적 기전과 임상적 중요성을 문헌 고찰을 통해 보고하고자 한다.

차의과학대학교 분당차병원 영상의학과