



# Combined Anatomical Anomalies of Direct Aortic Arch Origins of the Left Internal Carotid, Left External Carotid, and Left Vertebral Arteries: A Case Report

대동맥궁에서 독립적으로 기시하는 왼쪽 속목동맥, 왼쪽 바깥목동맥 및 왼쪽 척추동맥의 복합변이: 증례 보고

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Various branch anomalies of the aortic arch have been reported, but cases with separate origins of the internal and external carotid arteries with combined direct aortic arch origin of the left vertebral artery are extremely rare. Herein, we present a rare case of aplasia of the left common carotid artery with separate origins of the ipsilateral internal and external carotid arteries and vertebral artery from the aortic arch in a 10-year-old girl. In addition, we review the embryological development and clinical implications of these anatomical variations.

**Index terms** Internal Carotid Artery; External Carotid Artery; Common Carotid Artery; Magnetic Resonance Angiography; Computed Tomography Angiography

## INTRODUCTION

The aplasia of the common carotid artery (CCA) is a rare congenital vascular anomaly that is often associated with separate origins of the internal carotid artery (ICA) and external carotid artery (ECA) (1). In particular, separate origins of the ICA and ECA combined with the

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aortic arch origin of left vertebral artery (VA) are extremely rare (2, 3). Although these vascular anomalies are mostly asymptomatic, the identification of these anatomical variations can be a significant diagnostic and therapeutic challenge in clinical practice (4). Herein, we present a rare case of co-existence of direct aortic arch origins of the left ICA, ECA, and VA in a 10-year-old girl that was incidentally detected on head and neck CT angiography (CTA). Moreover, we discuss the embryological mechanisms and clinical implications of these anomalies.

## CASE REPORT

A 10-year-old girl visited our hospital's outpatient department with a 7-day history of conjunctival injection. Her medical history was unremarkable and her vital signs were stable. Ophthalmological examination raised concerns of bilateral papilledema. She underwent contrast enhanced brain MRI and intracranial time-of-flight MR angiography (MRA) to evaluate the cause of bilateral papilledema. On intracranial MRA, the left ICA showed diffuse narrowing up to the fetal type of the posterior communicating artery (Fig. 1A). Subsequent head and neck CTA revealed the agenesis of the left CCA with separate origins of the left ICA and ECA from the aortic arch (Fig. 1B). On axial CT images, the diameters of the left ICA and ipsilateral carotid canal were smaller than those of the contralateral ICA, which was a consistent finding to that of intracranial MRA (Fig. 1C, D). The left VA was also directly originated from the aortic arch with anomalous entering into the transverse foramen at the 5th cervical vertebra, which was at a lower level than the contralateral side (Fig. 1E). There was no significant abnormality in the brain parenchyma.

This study was approved by the local Institutional Review Board, and patient's informed consent was waived (IRB No. ISPAIK 2022-04-019).

## DISCUSSION

The aplasia of the CCA with separate origins of the ICA and ECA is a rare congenital vascular anomaly. A previous study reported 87 cases of CCA aplasia of which 34 had left CCA aplasia (5). Combined anatomical anomalies of direct aortic arch origins of the left ICA, ECA, and VA are extremely rare, and to our best knowledge, only two cases have presented these anomalies to date (2, 3). Braun et al. (2) reported a case of vascular anomalies similar to those reported in the present study. However, unlike our case, the left ICA first originated before the left ECA in their case. Bhat et al. (3) also reported a case with the similar anomalies as described in our case; however, that case had no available images of the intracranial vessels.

During embryonic development, the third primitive aortic arch develops into the CCA and proximal ICA (5). The ductus caroticus connects the third and fourth primitive aortic arches and normally regress during development. However, if the ductus caroticus does not regress and persists with regression of the third primitive aortic arch, it forms a separate origin for the ICA (5).

The VA usually arises at the ipsilateral subclavian artery and the variant origin of left VA from the aortic arch has been reported in 2.9% cases (6). The VA is formed through the development of a postcostal longitudinal anastomosis of the cervical intersegmental arteries (7). If the

**Fig. 1.** Coexistence of separate origins of the left ICA and ECA from the aortic arch and the anomalous origin of the left VA from the aortic arch in a 10-year-old girl.

**A.** A time-of-flight MR angiography maximum intensity projection image is presented. The anteroposterior view of the intracranial arteries depicts the diffuse narrowing of the left distal ICA. The diameter of the left distal ICA became is to that of the right distal ICA (arrow) at distal locations beyond the origin of the left posterior communicating artery (arrowhead).

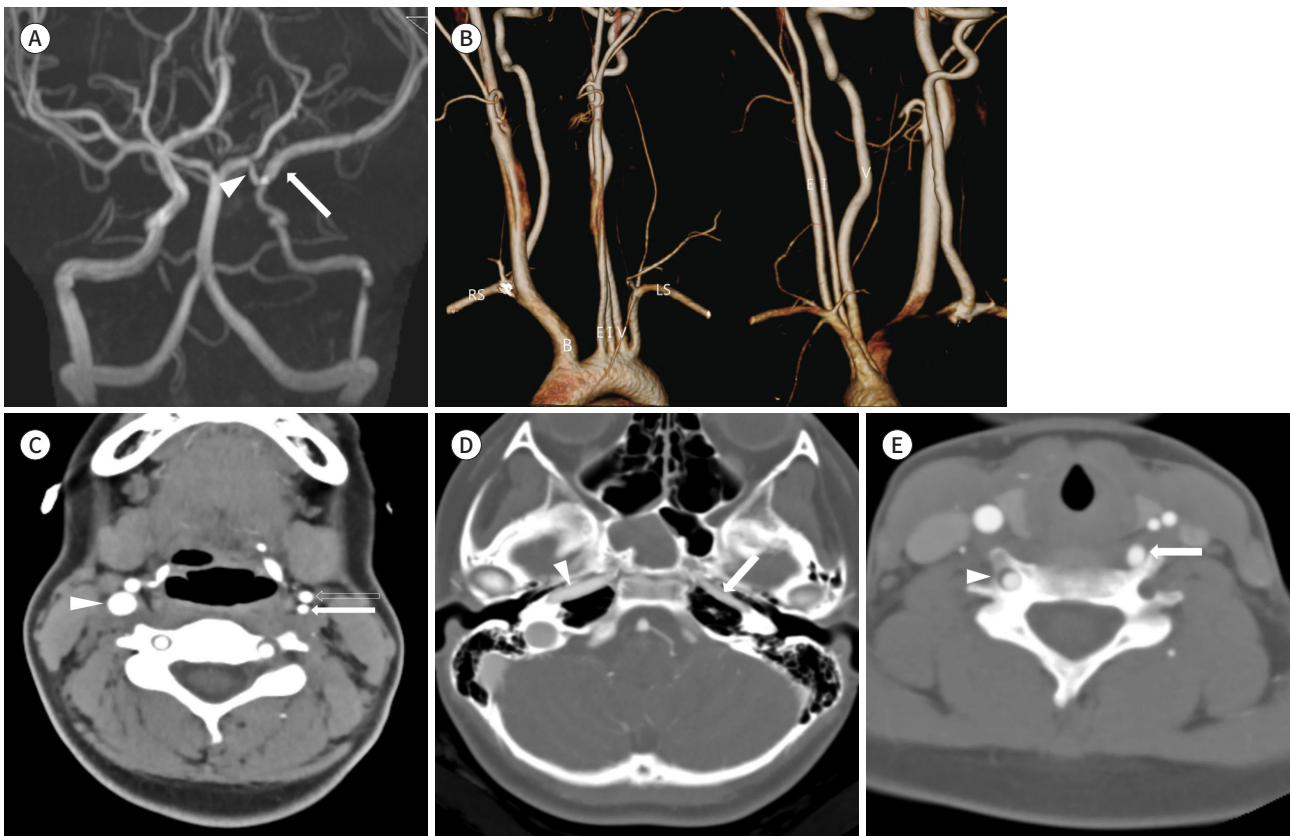
**B.** Left anterior oblique and lateral projections of three-dimensional volume-rendered CT angiography reveal five vessels that originate directly from the aortic arch: right brachiocephalic trunk (B), left ECA (E), left ICA (I), left VA (V), and LS.

**C.** The axial CT image shows that the diameter of left ICA (arrow) is smaller than those of the right ICA (arrowhead) and left ECA (blank arrow).

**D.** The axial CT image shows that the left carotid canal (arrow) is narrower than the right carotid canal (arrowhead).

**E.** The axial CT image acquired at the 6th cervical vertebra level shows the right VA (arrowhead) located in the right transverse foramen. The left VA (arrow) is not visible in the left transverse foramen in this figure, but it is located anteriorly.

B = brachiocephalic artery, E = left ECA, ECA = external carotid artery, ICA = internal carotid artery, I = left ICA, LS = left subclavian artery, RS = right subclavian artery, V = left VA, VA = vertebral artery



anastomosis does not develop between the left 6th and 7th intersegmental arteries and the left 6th intersegmental artery does not regress, the left VA originates from the aortic arch (7). In addition, a previous study suggested that the VA's origin is related to the entry level of transverse foramen, and the left VA originating from the aortic arch most commonly enters the transverse foramen of the 5th cervical vertebra as seen in our case (7).

Some cases of hypoplasia of the ICA accompanied by CCA aplasia have been reported similar to our case (5). Although, most cases of this anomaly are asymptomatic, symptoms related to ICA hypoplasia have been reported previously (8-10). Bryan et al. (8) reported a case of transient ischemic attack that may have occurred due to a hypoplastic left ICA. Hiratsuka et al. (9) reported a patient who presented with intracerebral hemorrhage in the right basal

ganglia and left ICA hypoplasia. Warschewske and Benndorf (10) reported a case of a contralateral giant ICA aneurysm. The authors suggested the possibility of hemodynamic stress from left ICA hypoplasia (9). In our case, the diameter of the left distal ICA became similar to that of the right distal ICA after the origin of the posterior communicating artery, and the left A1 segment of the anterior cerebral artery was hypoplastic. This may indicate that collateral circulation through the circle of Willis provides left cerebral blood supply and may cause hemodynamic stress on the right side and vertebrobasilar system. Thus, the careful attention and follow-up is needed, although no aneurysms were observed in our case.

In conclusion, we present a rare case of combined anatomical variations of direct aortic arch origins of the left ICA, ECA, and VA. Although these anomalies are usually asymptomatic, they can confound the diagnosis and treatment process. The accompanying ICA hypoplasia also can cause hemodynamic stress on the contralateral side. Thus, recognizing the congenital vascular anomalies of the aortic arch is important for diagnosis and treatment, and follow-up is needed.

#### Author Contributions

Conceptualization, L.B.; data curation, H.Y.J.; investigation, H.Y.J.; methodology, H.Y.J.; project administration, L.B.; supervision, L.B.; validation, H.Y.J.; visualization, P.D.Y.; writing—original draft, P.D.Y.; and writing—review & editing, L.B.

#### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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박대윤 · 이병훈\* · 황윤준

대동맥궁 줄기의 다양한 기시변이는 잘 알려져 있으나 속목동맥과 바깥목동맥의 독립적인 기시와 동반된 척추동맥의 대동맥궁 직접 기시에 대한 보고는 매우 드물다. 저자들은 10세 여아에서 왼쪽 온목동맥의 무형성과 동측 속목동맥, 바깥목동맥 및 척추동맥이 대동맥궁에서 직접 기시하는 매우 드문 복합 변이를 증례로 보고하고자 한다. 또한, 이 증례 보고를 통해 해당 변이의 발생학적 기전과 임상적 의의를 살펴보았다.

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