pISSN 2384-1095
eISSN 2384-1109

iMRI 2021;25:193-196

https://doi.org/10.13104/imri.2021.25.3.193

iMRI
Investigative Magnetic Resonance Imaging

Congenital Absence of the Bilateral Internal Carotid Arteries: a Case Report

Jihoon Noh, Hyunkoo Kang

Department of Radiology, Seoul Veterans Hospital, Seoul, Korea

Congenital absence of the bilateral internal carotid arteries (ICA) is a very rare occurrence. Recognition of this rare anomaly is important, when considering intracranial endovascular interventions in the event of thromboembolic events with revascularization, transsphenoidal surgery, and the surveillance and detection of associated cerebral aneurysms. We report a case of a 25-year-old man who presented with headache since 2 years ago, and was incidentally discovered to have a congenital bilateral absence of ICAs.

Keywords: Congenital absence of the bilateral internal carotid artery; Computed tomography; Magnetic resonance imaging

Case Report

Received: March 29, 2021
Revised: May 20, 2021
Accepted: June 9, 2021

Correspondence to:
Hyunkoo Kang, M.D.
Department of Radiology, Seoul Veterans Hospital, 6-2 Dunchon-dong, Gangdong-gu, Seoul 05368, Korea.
Tel. +82-2-2225-3969
Fax. +82-2-2225-1433
E-mail: knroo@hanmail.net

INTRODUCTION

Congenital absence of the bilateral internal carotid arteries (ICA) is an extremely unusual case (1, 2). This natural absence may be consequent of agenesis, aplasia, or hypoplasia. Congenital absence of the ICA is often associated with carotid canal atresia and other abnormalities, such as anomalous origin of the ophthalmic artery, pituitary hypoplasia, sympathetic nerve dysplasia, and abnormal development of collateral circulation pathways (3). Absence of the bilateral ICAs is associated with collateral blood flow, typically from the circle of Willis, for this reason, this abnormality is frequently asymptomatic and detected incidentally via imaging such as computed tomography (CT), or magnetic resonance imaging (MRI) (3). If there is a lack of adequate collateral flow, cerebrovascular accidents could occur and various symptoms could be present (4). The presented case involves a 25-year-old male patient with an incidental finding of bilateral absence of the ICAs.

CASE REPORT

A 25-year-old man presented with headache, which occurred two years ago while in military service. There was no history of other symptoms. Recently, the headache recurred.

A brain MRI was performed, which revealed an absence of the expected flow voids along the course of the petrous and cavernous segments of the both intracranial ICAs. There was no space occupying lesion or no signs of chronic ischemic changes in the brain parenchyma. A brain MR angiography was subsequently performed, which

revealed non-visualization of bilateral ICAs, prominent bilateral vertebral arteries, and prominent basilar and posterior communicating arteries (PCOMs). The PCOMs were supplying both middle cerebral arteries (MCAs) and the anterior cerebral arteries (ACAs) via the circle of Willis. The A1 segment of the right ACA was absent with the A2 segment being reformed via the anterior communicating artery. There was no evidence of an aneurysm or other intracranial collateral circulation (Fig. 1a). Anomalous origin of the both ophthalmic arteries derived from the ipsilateral middle meningeal arteries (Fig. 1b, c). Carotid MR angiography revealed markedly enlarged vertebrobasilar arteries (vertebrobasilar dolichoectasia) with normal external carotid arteries (ECAs) and non-visualization of the ICAs on both sides (Fig. 1d). Brain CT showed the absence of the both carotid canals which confirmed the agenesis of both ICAs (Fig. 1e). Brain 99mTc-ethylcysteinate dimer single-photon emission computed tomography (99mTc-ECD SPECT) showed mild reduced resting brain perfusion in the bilateral MCA territories (Fig. 1f). Brain mean transit time (Fig. 1g) perfusion MRI showed mild delayed perfusion in the bilateral MCA territories. These findings were representative of bilateral agenesis of the ICAs.

The patient's symptom resolved spontaneously during admission and was attributed to either transient ischemic attacks or migraine headaches.

DISCUSSION

The absence of the internal carotid artery is not a common congenital anomaly. There are only a few number of reported cases (2, 3). Among ICA congenital anomaly, bilateral ICA agenesis is extremely rare and the reported cases are limited. The first documented case of carotid agenesis, discovered on postmortem examination, was reported by Tode in 1787 (5). In 1954, the first case of ICA agenesis at cerebral angiography was reported by Verbiest (6).

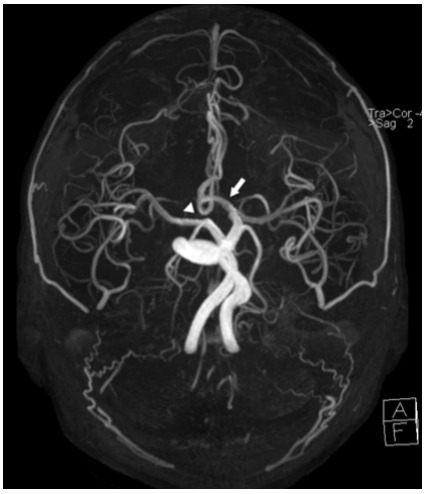
Although an exact cause of ICA agenesis has not been established, these variations are thought to represent the sequela from an insult to the developing embryo. Postulated causes of unilateral absence have centered on mechanical and hemodynamic stresses placed on the embryo. To date, an explanation for bilateral absence of ICAs has not been rendered (2).

Development of the carotid canals at the skull base occurs in the presence of the embryonic ICA during early gestation. A small or absent carotid canal therefore indicates a congenital internal carotid abnormality, differentiating it from the acquired causes of ICA narrowing (7). Agenesis, aplasia, and hypoplasia should be differentiated. Agenesis is the complete developmental failure of an organ and its primordium, whereas aplasia and hypoplasia are the incomplete development of a structure (8). Therefore, in agenesis the bony carotid canal is absent, whereas in aplasia and hypoplasia the bony carotid canal is present.

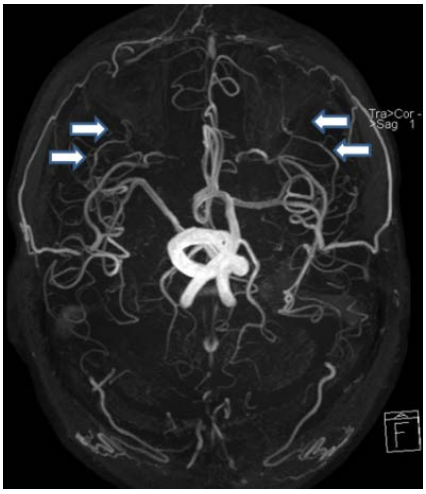
Three major collateral pathways to the anterior cerebral circulation have been described. The most common collateral pathway is through the enlarged PCOM, as was the case for this patient. The other types of collateral pathways area through anastomosis between the ECA and the ICA at the skull base, or via persistent fetal circulation (2). MR-based perfusion is widely used in the assessment of collaterals. This study is most commonly performed using a bolus of contrast and assessing flow dynamics by monitoring the passage of the bolus through the vessels and parenchyma (dynamic susceptibility contrast MRI, dynamic contrast enhanced MR perfusion). A more recent technique known as arterial spin labeling (ASL) magnetically labels arterial blood water using radiofrequency pulses that then decay with T1 relaxation and allows for quantitative assessments of regional cerebral blood flow without the need for a tracer. Other methods of measuring cerebral perfusion include positron emission tomography (PET) and SPECT. These techniques, in addition to the ones mentioned above, can be combined with a vasodilatory stimulus to determine the resilience of the cerebral circulation to ischemic insults, termed the cerebrovacular reserve (CVR).

Most of the patients are asymptomatic due to the collateral circulation, but the delayed blood flow can lead to low flow transient ischemic attacks, causing re-occurring headaches lasting a few minutes. Intermittent headaches are caused by the transient delayed hypo-perfusion in the watershed zone, mainly in the bilateral ACA and MCA territories. Other clinical symptoms, such as pulsatile tinnitus, migraine, and Horner's syndrome (9), have been reported.

Absence of the ICA may be associated with aneurysms of 24%-34%, compared to a 2%-4% prevalence observed in the general population (2). In many reported cases, the anterior communicating artery (ACOM) is the most common location of aneurysms (2). This implies there is an association between the increased flow through collateral vessels, such as ACOM, due to the agenesis of ICA and the risk of aneurysms (2). Rare syndromes, such as

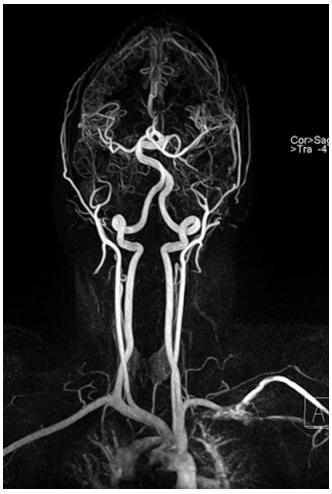


a

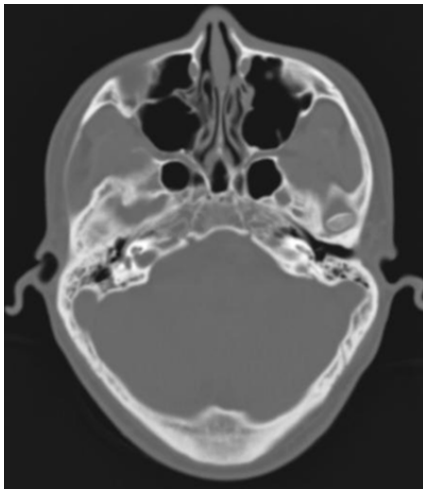


b

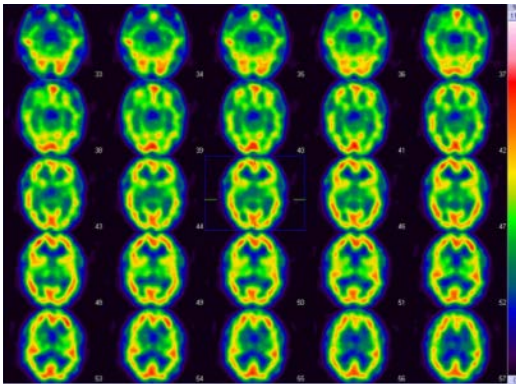
c



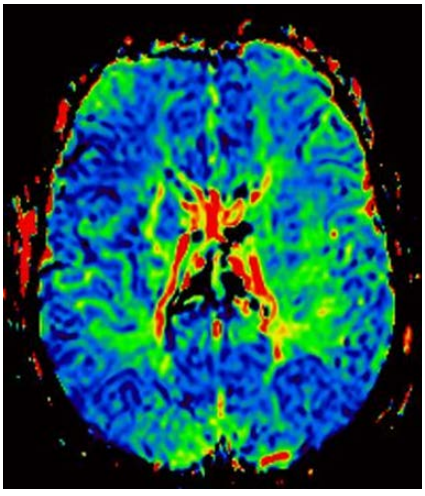
d



e



f



g

Fig. 1. (a) Brain magnetic resonance angiography (MRA) reveals total non-visualization of the both ICAs, and prominent bilateral vertebral arteries, and prominent basilar and posterior communicating arteries (PCOMs). The basilar artery is tortuous to the right side. The PCOMs are supplying both middle cerebral arteries (MCAs) and the anterior cerebral arteries (ACAs) via the circle of Willis. The A1 segment of the right ACA is absent (arrowhead) with the A2 segment being reformed via the anterior communicating artery (arrow). (b) Brain MRA reveals anomalous origin of the both ophthalmic arteries derived from the ipsilateral middle meningeal arteries (arrows). (c) Tranfemoral cerebral angiography reveals anomalous origin of the right ophthalmic arteries derived from the ipsilateral middle meningeal artery (arrow). (d) Carotid MRA reveals markedly enlarged vertebrobasilar arteries (vertebrobasilar dolichoectasia) with normal external carotid arteries and non-visualization of the ICAs on both sides. (e) Brain computed tomography (CT) in bone window at the skull base shows a lack of bilateral bony carotid canals. (f) Brain 99mTc-ECD SPECT shows mild reduced resting brain perfusion in the bilateral MCA territories. (g) Brain mean transit time perfusion MRI shows mild delayed perfusion in the bilateral MCA territories.

cardiac abnormalities/aortic coarctation and eye (PHACE) abnormalities, Goldenhar syndrome, Klippel-Feil syndrome, have also been reported in the setting of carotid agenesis (3).

Recognition of the absence of bilateral ICAs is important when planning intracranial vascular intervention, as both cerebral hemispheres may be dependent on a basilar artery. It is also important to recognize the intercavernous collateral pathway when planning transphenoidal hypophyseal surgery (2). Although most patients remain asymptomatic, the association with a high prevalence of aneurysm is an indication for clinical and radiological surveillance for these patients.

REFERENCES

1. Furruqh F, Biswas A, Thirunavukarasu S, Vivekandan R. Congenital absence of bilateral ICA: an unusual incidental finding in an adult male. BMJ Case Rep 2016;2016
2. Given CA 2nd, Huang-Hellinger F, Baker MD, Chepuri NB, Morris PP. Congenital absence of the internal carotid artery: case reports and review of the collateral circulation. AJNR Am J Neuroradiol 2001;22:1953-1959
3. Li S, Hooda K, Gupta N, Kumar Y. Internal carotid artery agenesis: a case report and review of literature. Neuroradiol J 2017;30:186-191
4. Chaudhry SR, Barreto S, Ezhapilli SR. Bilateral congenital absence of the internal carotid arteries: a case report. Radiol Case Rep 2018;13:1146-1149
5. Midkiff RB, Boykin MW, McFarland DR, Bauman JA. Agenesis of the internal carotid artery with intercavernous anastomosis. AJNR Am J Neuroradiol 1995;16:1356-1359
6. Verbiest H. Radiological findings in a case with absence of the left internal carotid artery and compression of several cranial nerve roots in the posterior fossa by the basilar artery. Med Contemp 1954;72:601-609
7. Anderson DW. Bilateral absence of the internal carotid artery: MR angiography and ultrasound findings. Br J Radiol 2005;78:569-572
8. Lie TA, Hage J. Congenital anomalies of the carotid arteries. Plast Reconstr Surg 1968;42:283
9. Cohen JE, Gomori JM, Leker RR. Internal carotid artery agenesis: diagnosis, clinical spectrum, associated conditions and its importance in the era of stroke interventions. Neurol Res 2010;32:1027-1032