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Case Report

Unilateral agenesis of internal carotid artery associated with superior cerebellar artery aneurysm and anomalous origin of ophthalmic artery arising from the ipsilateral posterior communicating artery ☆,☆☆,★,★★,‡

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ABSTRACT

Congenital agenesis of the unilateral internal carotid artery is a rare anomaly. The agenesis of the internal carotid artery is associated with a higher incidence of intracranial aneurysms. This report describes a rare case of internal carotid artery agenesis associated with an aneurysm of the left superior cerebellar artery in a 52-year-old woman. The left middle cerebral artery was supplied from the vertebrobasilar system through the dilated left posterior communicating artery associated with anomalous origin of the left ophthalmic artery from the left posterior communicating artery. This report demonstrates a rare combination of left internal carotid artery agenesis with associated intracranial aneurysm and anomalous origin of the ophthalmic artery, discussion of embryogenesis, clinical significance, and imaging findings.

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Introduction

Agenesis, aplasia, or hypoplasia of the internal carotid artery (ICA) are rare congenital vascular anomalies, seen in less than 0.01% of the population [1,2]. Most cases of ICA agenesis are clinically silent due to well-developed sufficient collateral circulations; therefore, it is usually detected incidentally on head and neck imaging [3]. The most common type of collateral flow is through the circle of Willis and less commonly provided via persistent embryologic vessels or transcranial collaterals through the external carotid artery system [2]. To the best of my knowledge, this is the extremely rare case describing unilateral agenesis of the ICA associated with an unruptured aneurysm of the left superior cerebellar artery (SCA) and anomalous origin of the ophthalmic artery (OA) from the posterior communicating artery (PCOM). Recognition of this rare anomaly is essential during direct head and neck surgery, endovascular interventions, and the surveillance and detection of cerebral aneurysms.

Case report

A 52-year-old woman presented with a mild headache for 1 year. She had no hypertension, diabetes mellitus, or smoking history. Cranial and cervical magnetic resonance angiography (Fig. 1) revealed the absence of flow-related signal intensity within the left ICA with the collateral flow to the left middle cerebral artery (MCA) territory through the posterior communicating artery via the vertebrobasilar system. Aneurysmal dilatation was detected at the origin of the left SCA.

Distal subtraction angiography (Fig. 2) revealed a left common carotid artery of the reduced caliber that terminated in the external carotid artery with no identifiable remnant of the ICA. The left anterior cerebral artery (ACA) was supplied from the right ICA through a patent anterior communicating artery

(ACOM) (Fig. 2B). The left MCA was directly supplied from the vertebrobasilar system through a hypertrophied left PCOM accompanied by an aneurysm at the origin of the left SCA on vertebral angiography (Fig. 2C). An anomalous origin of the left OA derived from the left PCOM was also noted. The aortic arch and major branches showed a typical pattern. Axial computed tomography (CT) at the bone window revealed absent left bony carotid canal (Fig. 3A). Coronal contrast-enhanced CT showed bilateral OA passing through the optic canal (Fig. 3B).

Since the maximum dimension of the aneurysm was less than 3 mm, we believed that imaging follow-up would be more appropriate than endovascular coil embolization or surgical clipping. Follow-up CT angiography performed 7 years after the initial diagnosis did not show growth of the SCA aneurysm or new development of intracranial aneurysms.

Discussion

ICA agenesis is a rare congenital anomaly [3]. The congenital absence of ICA may be unilateral or bilateral, with a higher proportion of cases being unilateral, and there is a left-sided predominance of ICA agenesis with a ratio of 3:1 [2]. The present case also presented with agenesis of the ICA on the left side. As agenesis of the ICA usually leads to sufficient collateral circulation to the affected side of the brain parenchyma, most cases are asymptomatic and are therefore frequently identified incidentally [1,4]. However, some patients can also present with subarachnoid hemorrhage from aneurysm rupture or symptoms related to associated vascular insufficiency due to changes in the collateral flow [3,5].

Embryologically, the primitive ICA originates from the fetal third aortic arches and cranial extensions of the embryonic dorsal aorta around the 3-mm fetal stage [3,6]. ICA development is completed at 6 gestational weeks [1]. Although the exact mechanism of unilateral agenesis of the ICA has not been established, postulated causes of this developmental anomaly

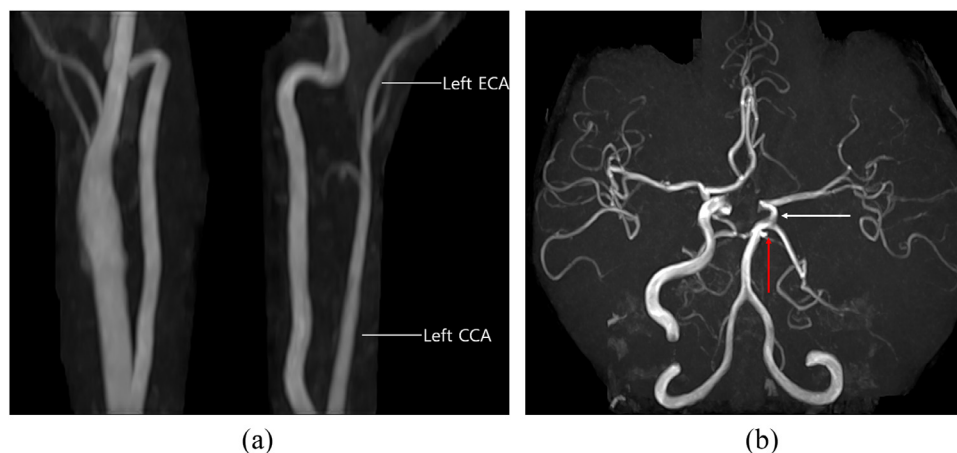


Fig. 1 – Time-of-flight magnetic resonance angiography (MRA) of the neck (A) revealed the absence of flow related signal intensity within the left internal carotid artery. MRA of the circle of Willis (B) revealed collateral flow to the left middle cerebral artery via the hypertrophied left posterior communicating artery (white arrow). Note aneurysmal dilatation at the origin of the left superior cerebellar artery (red arrow). CCA, common carotid artery; ECA, external carotid artery

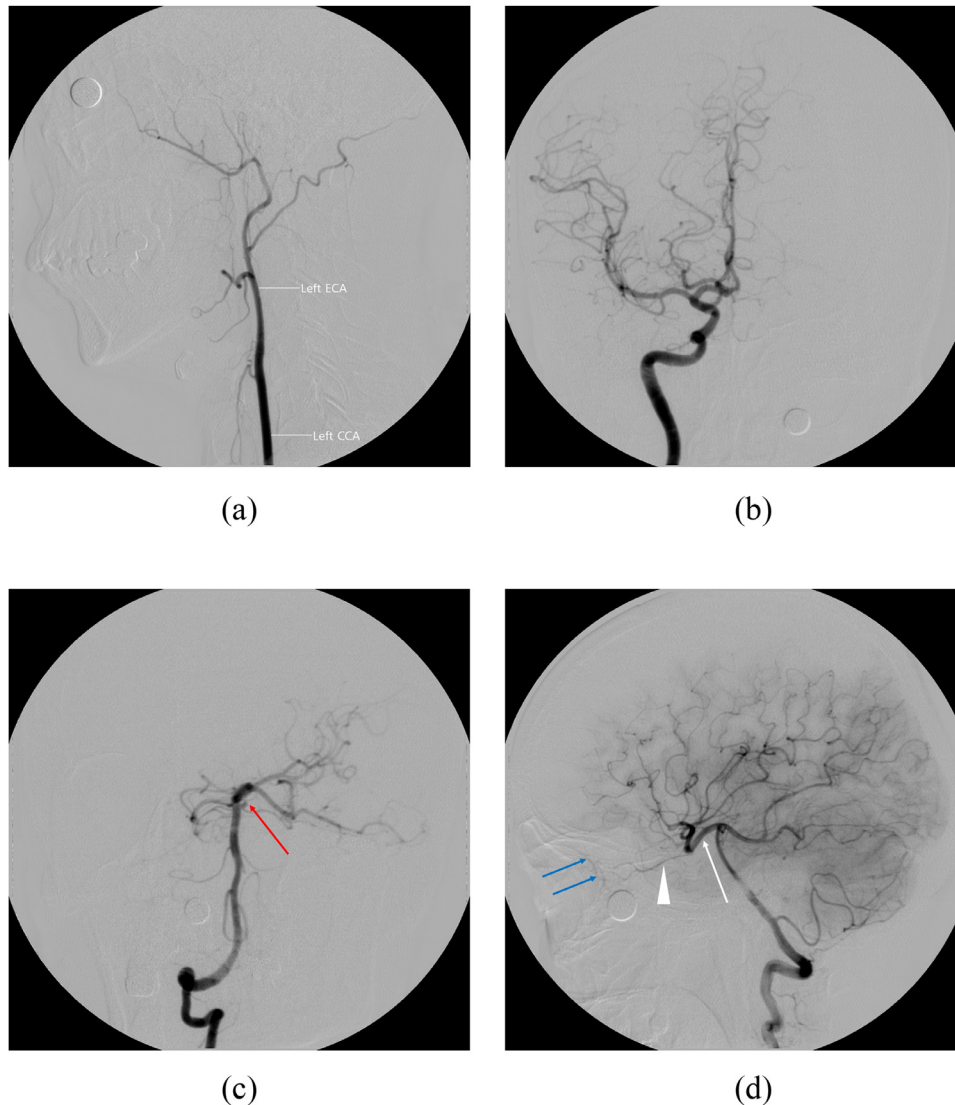


Fig. 2 – The left common carotid angiography (A) demonstrates that the small caliber of the left CCA and the complete absence of the left internal carotid artery. Right carotid angiography (B) showed the left anterior cerebral artery supplied by the right side via the anterior communicating artery. The vertebral angiography, anteroposterior (C), and lateral views (D) demonstrate that the left middle cerebral artery supplied from the basilar artery via the dilated posterior communicating artery (white arrow). Note aneurysm arising from the left superior cerebellar artery (red arrow). The left ophthalmic artery (arrowhead) originates from the posterior communicating artery, a choroidal blush (blue arrows) in the capillary phase of lateral angiography. CCA, common carotid artery; ECA, external carotid artery

have been thought to present the sequela from an insult to the developing embryo [2].

The carotid canal develops in association with the ICA [7]. ICA forms by the 4th embryologic week, while the skull base forms at 5-6 weeks [4]. If the embryonic ICA does not develop or fails to develop before the fifth embryonic week, the ICA and carotid canal cannot develop [5]. Therefore, the absence of the bony carotid canal is essential to differentiate congenital agenesis from the acquired type of ICA occlusion, such as atherosclerotic occlusion or dissection [3]. The present case showed no bony carotid canal on the left side.

In the congenital absence of ICA, collateral circulation develops through the circle of Willis, persistent embryonic

arteries, or transcranial collaterals of the external carotid artery [1]. Lie described 6 pathways of collateral circulation in association with the absence of the ICA [8]. In the absence of ICA, the most common collateral circulation develops through the circle of Willis [9]. Lie's type A, the most common type of unilateral ICA agenesis, represents the unilateral absence of the ICA associated with collateral circulation to the ipsilateral ACA through a patent ACOM and to the ipsilateral MCA from the posterior circulation through a hypertrophied PCOM [9]. The present case belongs to Lie's type A.

Other less common collateral flows are through persistent embryonic arteries or transcranial collaterals of the external carotid artery system [1,5]. It is associated with the embryonic

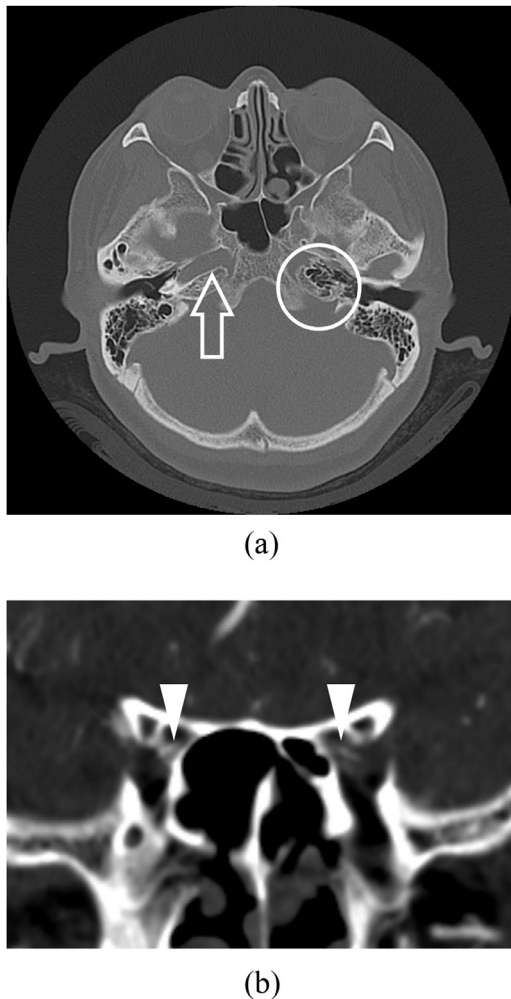


Fig. 3 – An axial computed tomography (A) image in the bone window at the skull base reveals the absence of a carotid canal on the left (circle). Note the patent carotid canal on the right (open arrow). Coronal contrast enhanced computed tomography (B) showed ophthalmic arteries passing through the optic canal bilaterally (arrowheads)

period in which there is a disruption of ICA development [1]. The circle of Willis forms during the 7-24 mm stage of the embryonic period [1]. The basilar artery is formed at the 7-12 mm stage of embryonal development, and the ACOM is formed at the 24-mm stage [10]. If the embryologic insult occurred after the 24-mm stage of development, collateral flow through the circle of Willis would dominate [11]. If the disruption occurred before the completion of the circle of Willis, primitive pathways of collateral circulation would prevail [4]. In this patient, the collateral pathway to the left MCA was maintained through the basilar artery via the ipsilateral posterior communicating artery. The left ACA was supplied from the contralateral ICA through the ACOM. Also, no primitive pathway was identified. Therefore, the disruption of normal ICA development possibly occurred after the 24-mm stage in the present case.

When the ICA is congenitally absent or hypoplastic, collateral circulation develops through the circle of Willis from

the contralateral ICA or the vertebrobasilar system, or both, to supply the involved cerebral hemisphere [4]. The reported incidence of aneurysms associated with the absence of the ICA is 24%-34%. This percentage is much higher than the 2%-4% incidence of aneurysms in the general population [2]. The high hemodynamic stress, such as increased flow and altered flow through collateral arteries, may be plausible explanations for this increased prevalence [2]. Therefore, these anomalies are frequently associated with cerebral aneurysms [4]. ACOM is the most frequent site of aneurysm formation in agenesis of the ICA [2]. Aneurysms were also located in the PCOM, MCA, basilar artery, or basilar tip in this anomaly [9,10,12]. However, aneurysm at the SCA's origin associated with the unilateral ICA's agenesis has not been reported before in the literature.

Because OA arises from the ICA, if the ICA is not formed, the OA will be abnormal [9]. The OA mostly originates from the intradural portion of the ICA just distal to the dural ring; however, it can also arise from the middle meningeal artery, anterior cerebral artery, accessory meningeal artery, basilar artery, middle cerebral artery, anterior temporal artery, and external carotid artery [9,13]. There are a limited number of cases of anomalous origins of the OA from the posterior communicating artery in the absence of ICA [13].

OA has complex embryogenesis, which is closely related to the development of the ICA [3]. The ventral OA arising from the ACA reaches the orbit through the optic canal and forms an anastomosis with the ICA, and its proximal segment regresses. Then, the primitive OA reaches its exact origin by considerable caudal migration [13,14]. The present case showed left OA arising from the ipsilateral PCOM and passed through the optic canal. Therefore, one of the possible embryologic explanations of the OA's PCOM origin in the present case is that the ventral OA had stopped migrating caudally at the segment where the PCOM joins the terminal ICA segment [13].

Conclusion

Association of unilateral ICA agenesis, superior cerebellar artery aneurysm, and anomalous origin of the OA from the ipsilateral posterior communicating artery are extremely rare. Recognition of this rare variant may help prevent the erroneous diagnosis of carotid dissection, severe carotid stenosis, or ICA occlusion in ischemic stroke. Therefore, careful investigation of the collateral circulation is important in surgical approaches such as trans-sphenoidal hypophyseal surgery, carotid endarterectomy, or endovascular treatment in the thromboembolic event. In addition, because the frequency of aneurysm formation is increased in agenesis of the ICA, close follow-up is also recommended in these patients.

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