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Case Report

Rare Segmental Agenesis of Internal Carotid Artery without Rete-Like Collaterals: A Case Report

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Objective: Internal carotid artery (ICA) agenesis has been classified into six types: A–F. Type F demonstrates distal reconstitution of the ICA via anastomosis with distal branches of the external carotid artery. Herein, we report the ICA agenesis of type F without rete-like collaterals, which has not been previously reported.

Case Presentation: An 80-year-old woman presented with segmental agenesis of the right ICA accompanied by an unruptured intracranial aneurysm. Stent-assisted coil embolization was successfully performed. Digital subtraction angiography showed segmental agenesis of the right ICA from the cervical to the ascending foramen lacerum segment, which was preoperatively supplied with collateral blood flow by a dilated right accessory meningeal artery (AMA) anastomosed with the inferolateral trunk (ILT)-posteromedial branch. Based on the segmental concept, the case was diagnosed with segment 7 (horizontal intracavernous portion until ICA branches off the ILT) agenesis, which may have resulted in secondary regression of the ICA proximal to segment 7. According to the ICA agenesis classification, this was of type F because the case showed collateral flow to the distal ICA via transcranial anastomoses from the AMA without carotid rete-like collaterals.

Conclusion: These findings suggest that the carotid rete-like collaterals did not form because the AMA was first developed during embryonic development.

Keywords segmental agenesis, internal carotid artery, rete-like collaterals, accessory meningeal artery, aneurysm

Introduction

The concept of segmental identity and vulnerability, first proposed by Lasjaunias, posits seven segments of the internal carotid artery (ICA) between the cervical carotid bifurcation and the posterior communicating artery.[1)] Komiyama[2)] further developed the segment identity concept by dividing the ICA into 10 segments using the new segmental concept. These concepts were crucial when considering the pathogenesis of ICA agenesis.

The frequency of ICA agenesis is estimated to be <0.01%.[3)] Six types of circulation to the anterior cerebral artery (ACA) and middle cerebral artery (MCA) on the ipsilateral side of carotid agenesis have been previously described by Lie and Hage.[4)] Among these, type F demonstrates distal reconstitution of the ICA via anastomosis with distal branches of the external carotid artery (ECA). These branches may present a meshwork morphology formed by multiple arteries and arterioles originating from the internal maxillary artery (IMA) and ascending pharyngeal artery. This vascular network is defined as the carotid rete-like collaterals.[5)] In ICA agenesis of type F without the carotid rete-like collaterals, how the collateral cerebral circulation is formed should be considered based on the segmental concept and the origin of the branches of the ICA and ECA.

Herein, we report the rare segmental agenesis of the ICA without rete-like collaterals, which has not been previously reported.

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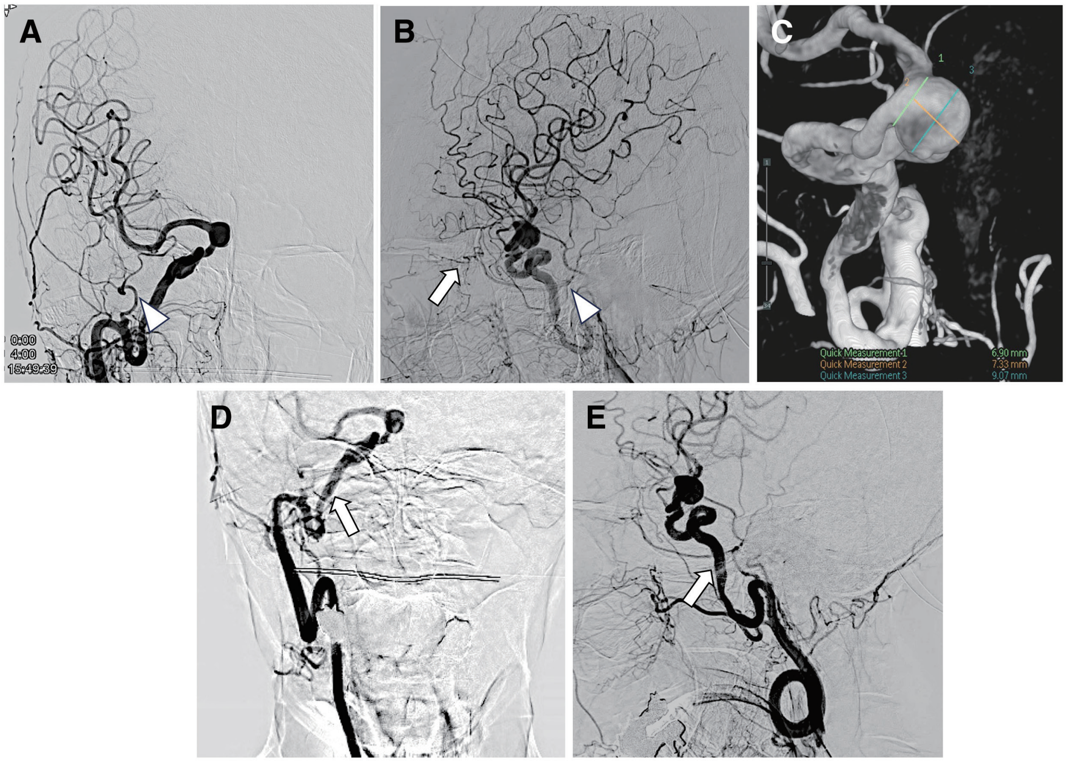

Fig. 1 (**A–C**) Right common carotid angiography (anterior and 3D view) shows a saccular aneurysm with a wide neck in the C1 portion of the right ICA. The ophthalmic artery (white arrow) originates from the MMA (white arrowhead). (**D** and **E**) Right external carotid angiography (lateral and anterior view) shows a dilated AMA (white arrow). AMA, accessory meningeal artery; ICA, internal carotid artery; MMA, middle meningeal artery

Case Presentation

An 80-year-old woman was being followed up for an asymptomatic unruptured cerebral aneurysm with a dome diameter of 7.3 mm, a neck diameter of 6.7 mm, and a height of 6.6 mm in the C1 portion of the ICA (by Fischer's segmental classification), which had been discovered incidentally 6 years prior. She had aortic stenosis, diabetes, and hypertension and had undergone surgical treatment for aortic stenosis. The patient had no family history of cerebral aneurysm. The size of the aneurysm increased over time. Follow-up angiography revealed a saccular aneurysm with a dome diameter of 9.1 mm, a neck diameter of 6.9 mm, and a height of 7.3 mm. The ophthalmic artery originates from the middle meningeal artery (MMA) (**Fig. 1A–1C**). Angiography incidentally revealed right ICA agenesis with collateral blood flow via the dilated accessory meningeal artery (AMA) (**Fig. 1D** and **1E**). Thin-slice CTA showed the absence of the right carotid canal. The dilated AMA was anastomosed with the posteromedial branch of the inferolateral trunk (ILT) at the cavernous portion to provide intracranial ICA blood flow without rete-like collaterals (**Fig. 2**).

Clipping surgery was challenging due to the patient's anatomy and age. As an alternative, stent-assisted coil embolization was performed. We attempted coil embolization via the right transbrachial approach because we failed to navigate the guiding sheath into the common carotid artery (CCA) via the transfemoral approach owing to the tortuosity of the CCA. A 90 cm 6 Fr FUBUKI XF guiding sheath (ASAHI INTECC, Aichi, Japan) was placed in the proximal CCA as a guiding catheter (**Fig. 3A**). Then, a 115 cm 6 Fr SOFIA (MicroVention Terumo, Aliso Viejo, CA, USA) was advanced into the ECA as a distal access catheter (DAC) (**Fig. 3B**). Using a Synchro2 soft 0.014 inch 200 cm micro guidewire (Stryker, Kalamazoo, MI, USA), a 150 cm 1.7 Fr Excelsior SL-10 (Stryker) was navigated into the M1 distal portion of the MCA. At this point, only one microcatheter (MC) could be inserted into the DAC due to kinking; therefore, the Neuroform Atlas stent (4.5 × 30 mm; Stryker) was placed first. After stenting, the MC was changed to a 156 cm 1.6 Fr HeadwayDuo (MicroVention Terumo), which was navigated into the aneurysm via the trans-cell approach to perform coil embolization.

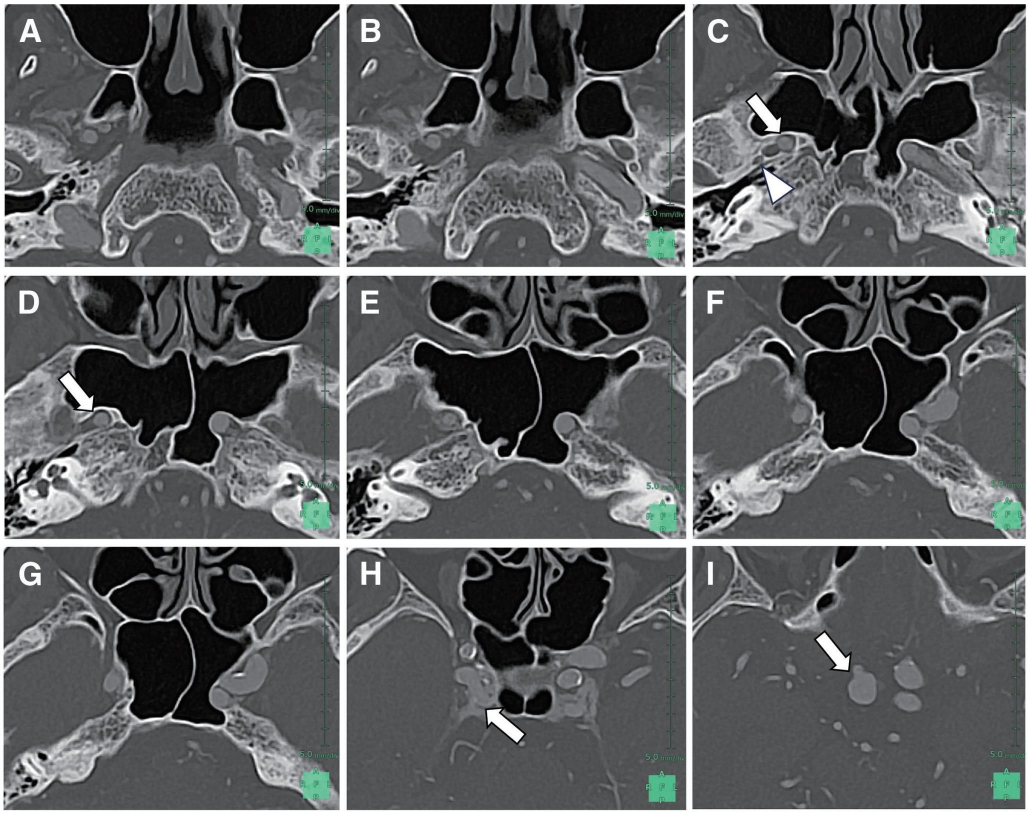

Fig. 2 (**A** and **B**) Computed tomography angiography shows no right carotid canal. (**C** and **D**) A dilated AMA (white arrow) runs through the foramen ovale. MMA runs through the foramen spinosum (white arrowhead). (**E–H**) AMA runs intracranially and anastomoses with the inferolateral trunk (white arrow) to transition to the ICA. (**I**) Aneurysm in ICA C1 portion (white arrow). AMA, accessory meningeal artery; ICA, internal carotid artery; MMA, middle meningeal artery

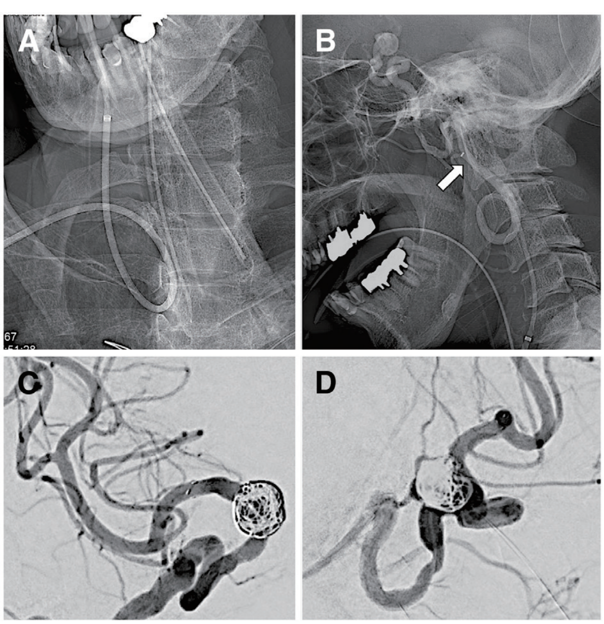

Fig. 3 (**A**) Right common carotid angiography (anterior view) shows the guiding of the guiding sheath. (**B**) Right common carotid angiography (lateral view) shows the guiding of the distal access catheter (white arrow). (**C** and **D**) Right common carotid angiography (anterior and lateral view) after coil embolization.

The following five coils were used for the procedure:

- The i-ED COIL Complex Infini SilkySoft 4–8 mm × 30 cm (KANEKA, Osaka, Japan);
- two Barricade complex finish coils 5 mm × 13 cm (Balt USA, Irvine, CA, USA);
- i-ED COIL Complex Infini SilkySoft 3–5 mm × 15 cm (KANEKA);
- Barricade complex finish coil 3 mm × 8 cm (Balt USA); and
- Barricade complex finish coil 3 mm × 10 cm (Balt USA).

Then complete occlusion of the aneurysm was confirmed (**Fig. 3C** and **3D**).

The volume embolization rate of the treated aneurysm was calculated to be 32.3%. Magnetic resonance imaging 6 months after the operation confirmed complete occlusion of the aneurysm.

Discussion

In this case, the AMA was developed as a collateral channel and was anastomosed to the posteromedial branch of the ILT.

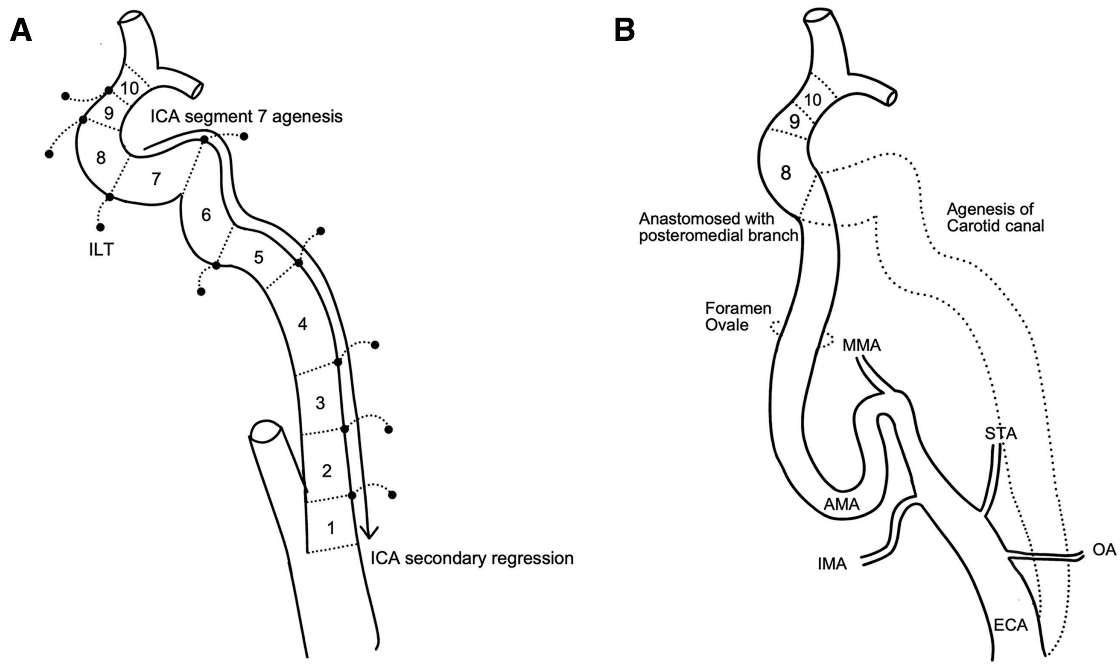

Fig. 4 (**A**) Based on the segmental concept, there are 10 segments between the cervical bifurcation and the origin of the posterior communicating artery. Between the segments, there are primitive arteries or their remnants. ICA segment 7 agenesis resulted in the secondary regression of the ICA proximal to segment 7. (**B**) The AMA had developed and anastomosed with the ILT-posteromedial branch to supply blood flow for intracranial ICA. AMA, accessory meningeal artery; ECA, external carotid artery; ICA, internal carotid artery; ILT, inferolateral trunk; IMA, internal maxillary artery; MMA, middle meningeal artery; OA, occipital artery; STA, superficial temporal artery

According to the ICA agenesis classification, type F demonstrates distal reconstitution of the ICA via anastomosis with distal branches of the ECA.[4] This case can be classified as type F because the AMA showed collateral flow to the distal ICA via transcranial anastomoses from the branches of the ECA system without carotid rete-like collaterals. There have been no previous reports of a case with this vascular anomaly.[4] Based on the segmental concept, segment 7 refers to the horizontal intracavernous portion until the ICA branches off the ILT. This case demonstrated segment 7 agenesis, which might have resulted from secondary regression of the ICA proximal to segment 7 (**Fig. 4A**).[2]

In the present case, the right carotid canal was absent, supporting the diagnosis of right ICA agenesis. The carotid canal is formed at 5–6 weeks of gestation. Therefore, the absence of the carotid canal indicates that secondary regression of the ICA occurred before this gestation stage. In ontogenetic processes, connecting the first and second arches to the dorsal aorta regresses and contributes to ECA formation. Non-regression of communication with the dorsal aorta may persist as a transcranial ECA-ICA anastomosis. An AMA develops from the stapedial artery and forms a branch of the IMA.[6] In the present case, the AMA and MMA originated separately from the IMA. The AMA can supply blood flow to the distal ICA via the posteromedial branch of the ILT.[7] Ikezawa et al.[8] reported that the anastomosis between the AMA and ILT is rarely dilated to supply adequate blood flow to the ICA. However, in this case, the AMA was developed from the embryonic period as a collateral blood vessel after ICA regression. The developed AMA, with a diameter similar to that of the ICA, supplied the ICA with sufficient blood flow (**Fig. 4B**). Mahadevan et al.[9] reported that segmental agenesis of the ICA occurred by regression after its development and that the carotid rete-like collaterals were formed in early development as a secondary collateral pathway. However, the carotid rete-like collaterals in the present case were not formed due to the previously and sufficiently developed AMA. This difference in etiology may be due to segmental agenesis, with rete-like collaterals forming in a shorter, more distal segment and at a later time, thus inducing the collaterals via the MMA, ILT, artery of the foramen rotundum, and other parasellar branches, unlike the present case without carotid rete-like collaterals.[10]

An incidence rate of 25%–43% has been reported for intracranial aneurysms in association with ICA agenesis, which is much higher than that found in the general population (2%–4%).[11] Two mechanisms have been postulated to explain this association between an intracranial aneurysm and ICA agenesis: (1) both conditions can occur independently during embryonic life as a result of developmental error[12] and (2) the aneurysm may develop secondary to hemodynamic stress.[13] Since the hemodynamic stress of intracranial arteries by collateral blood flow is unlikely to have increased in the present case, the aneurysm most likely developed via the first mechanism.

Stent-assisted coil embolization using a DAC was performed on the patient. In the present case, the ophthalmic artery originated from the MMA due to ICA agenesis. When we perform clipping surgery, we should avoid injury to the orbital branch of the MMA by the high-speed drill or during the dissection of the dura mater.[14] Patients with ICA agenesis often develop aneurysms. When aneurysms occur, the complex vascular anatomy makes the choice of surgical approach difficult. Therefore, careful imaging assessment and appropriate surgical intervention are necessary.

Conclusion

Herein, we report for the first time on rare segmental agenesis of ICA without rete-like collaterals. The etiology was the agenesis of segment 7, with the development of AMA as collateral.

Declarations

Ethics statement

Ethics approval was not required for this study. Written informed consent was obtained from the patient.

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The authors declare no potential conflicts of interest with respect to the research, authorship, or publication of this article.

Data availability

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

Disclosure statement

The authors declare that there is no conflict of interest.

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