

Segmental Aplasia of Bilateral Internal Carotid Arteries Accompanied by Intracranial Aneurysms: A Case Report

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A 68-year-old woman presented with segmental aplasia of bilateral internal carotid arteries accompanied by unruptured intracranial aneurysms. The abnormality was discovered incidentally at the age of 44 years. Cerebral angiography showed occlusion of bilateral internal carotid arteries, and the carotid territory was supplied by each posterior communicating artery with small intracranial aneurysms. Endovascular treatment for the intracranial aneurysms was planned. However, the patient did not want to undergo the endovascular procedure because of the increased risk due to the associated bilateral carotid abnormalities. Cerebral angiography was performed again at the age of 66 years, and the size of the aneurysms had not changed. Based on their segmental identity, aplasia of segment 6 of the internal carotid artery (ICA) including the first portion of the ophthalmic artery was observed bilaterally.

Keywords: carotid arteries, intracranial aneurysm, abnormalities

Introduction

Aplasia/hypoplasia of the internal carotid artery (ICA) is rare, with an estimated prevalence of 0.01%.^{1,2} This abnormality is associated with a high occurrence of intracranial aneurysms, ranging from 25% to 43%,^{3–6} compared to 2% to 4% in normal individuals. This abnormality may be harmless, but associated conditions such as intracranial aneurysms should alert clinicians to the possibility of deterioration to a life-threatening condition, such as subarachnoid hemorrhage.^{1,7,8} Endovascular treatment or surgical clipping is performed for treatment,^{8–10} but the natural history and the average annual risks of rupture associated with unruptured intracranial aneurysms in this abnormality are unknown. A case of segmental aplasia of the bilateral ICAs with small saccular aneurysms, which was managed conservatively for over 20 years, is presented.

Case Presentation

The patient was a 68-year-old woman with a history of diabetes mellitus and uterine myoma. She was referred to

our medical center for headache at 44 years of age. Cerebral angiography incidentally revealed occlusion of bilateral ICAs, developed bilateral posterior communicating arteries, and collateral flow to the carotid territory supplied by each posterior communicating artery (Fig. 1). The circulation pathway associated with the absence of the ICA was Type C based on Lie's classification.¹¹ Digital subtraction angiography (DSA) at 46 years of age showed saccular aneurysms of about 4 mm at the basilar artery and left superior cerebellar artery (SCA) region and on the right posterior communicating artery (PCoA) (Fig. 2). Since both intracranial aneurysms were small and unruptured, the aneurysms were managed conservatively. Ten years ago, coil embolization was planned for the intracranial aneurysms, but the treatment was cancelled at the request of the patient and family because of the increased risk due to the associated bilateral carotid abnormalities. At 66 years of age, DSA was repeated for the purpose of evaluating the ICAs and intracranial aneurysms. DSA showed that the size of the basilar-left SCA aneurysm and the right PCoA aneurysm had not changed (Fig. 2). The bilateral ICAs were occluded at each inferolateral trunk (ILT) as terminal branches, the origins of the bilateral ophthalmic arteries were also occluded, and the distal portion of the ophthalmic artery was supplied by the deep recurrent ophthalmic artery of the ILT and several branches from the external carotid artery (Fig. 3). Based on segmental identity,^{12,13} these findings indicated that the bilateral ICAs might be hypoplastic from segment 1 to segment 5, with aplasia of segment 6, including the first portion of the ophthalmic artery. Computed tomography (CT) and time-of-flight (TOF) were readily fused using a 3D-imaging workstation (Synapse Vincent, Fujifilm Medical, Tokyo, Japan). CT and TOF fused images showed the bilateral hypoplastic ICAs in each hypoplastic carotid canal, the origin of the ILT, and absence of segment 6 of the ICA bilaterally (Fig. 4). The aneurysms have been followed conservatively for over 20 years.

Discussion

Aplasia/hypoplasia of the ICA is a rare abnormality and associated with a high rate of intracranial aneurysms; however, there have been no case reports of long-term follow-up of unruptured intracranial aneurysms associated with this carotid abnormality. To the best of our knowledge, this is the first case of long-term follow-up of unruptured intracranial aneurysms associated with aplasia/hypoplasia

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of the ICAs. Knowledge of the clinical course of this case is valuable for many neurosurgeons and neuroradiologists.

Lie described six pathways of collateral circulation in association with absence of the ICA. Type A cases involve the unilateral absence of the ICA and the supply of blood to the ipsilateral anterior cerebral artery (ACA) through the anterior communicating artery (ACoA) and to the middle

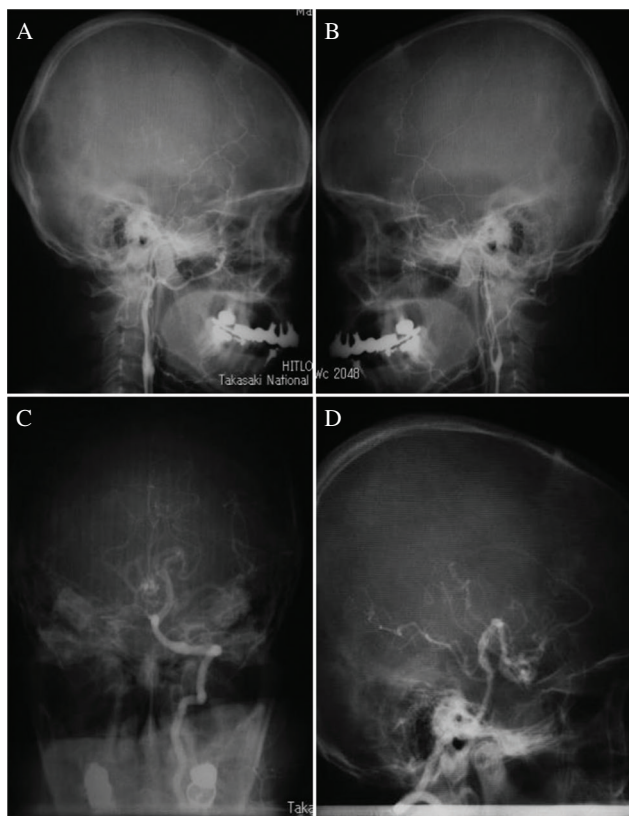


Fig. 1 Conventional angiogram at 44 years of age. Lateral view of the bilateral carotid angiogram showing occlusion of the bilateral internal carotid arteries (A, B). Angiograms of the left vertebral artery showing bilateral posterior communicating arteries, with collateral flow to the carotid territories (C, D).

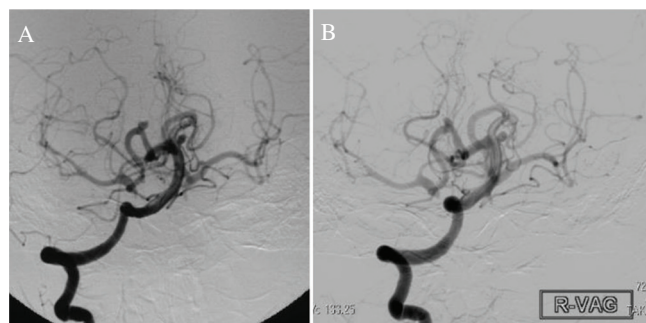


Fig. 2 Digital subtraction angiography of the right vertebral artery at 46 years of age showing small unruptured cerebral aneurysms in the basilar-left superior cerebellar artery and right posterior communicating artery (A). Digital subtraction angiography of the right vertebral artery at 66 years of age shows that the size of the basilar-left superior cerebellar artery aneurysm and the right posterior communicating artery aneurysm have not changed (B).

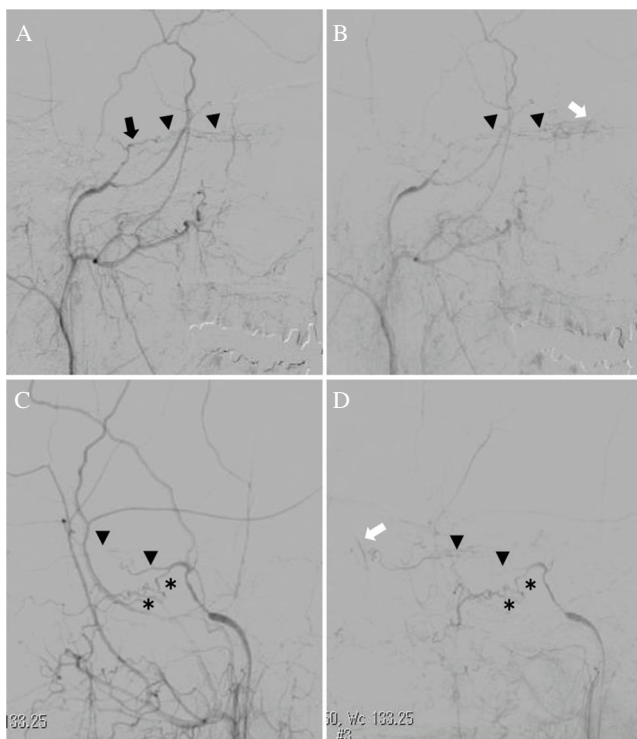


Fig. 3 Lateral view of the right carotid angiogram showing the deep recurrent ophthalmic artery (arrowheads) of the inferolateral trunk (arrow) (A) and choroid blush (white arrow) supplied mainly by the deep recurrent ophthalmic artery (arrowheads) (B). Lateral view of the left carotid angiogram showing the deep recurrent ophthalmic artery (arrowheads) and recurrent artery of the foramen rotundum (asterisks) (C, D), and choroid blush (white arrow) (D).

cerebral artery (MCA) through a hypertrophied PCoA. Type B cases involve the ipsilateral ACA and MCA, with the supply of blood through the ACoA. Type C cases show bilateral agenesis of the ICA, with the supply to the anterior circulation through carotid-vertebrobasilar anastomoses; a review of the literature found that this is generally accomplished through hypertrophy of the PCoA,^{9,14} and the present case is thought to correspond to type C. Type D cases show unilateral agenesis of the cervical portions of the ICA with an intercavernous communication to the ipsilateral carotid siphon from the contralateral cavernous ICA. In type E, small ACAs are supplied by bilateral hypoplastic ICAs, while the MCAs are supplied by enlarged PCoAs. Type F cases show collateral flow to the distal ICA via transcranial anastomoses from the internal maxillary branches of the external carotid artery system, the so-called rete mirabile.

Aplasia/hypoplasia of the ICA is associated with a high rate of intracranial aneurysms, ranging from 25% to 43%,³⁻⁶ compared to 2% to 4% in normal individuals. The annual rate of rupture for unruptured intracranial aneurysms has been estimated by previous studies,¹⁵⁻¹⁸ whereas the annual rate of rupture of unruptured intracranial aneurysms associated with aplasia/hypoplasia of the ICAs has been unknown because of the inadequate number of cases and the short follow-up duration in the literature. However, intracranial aneurysms associated with aplasia/hypoplasia of the ICAs

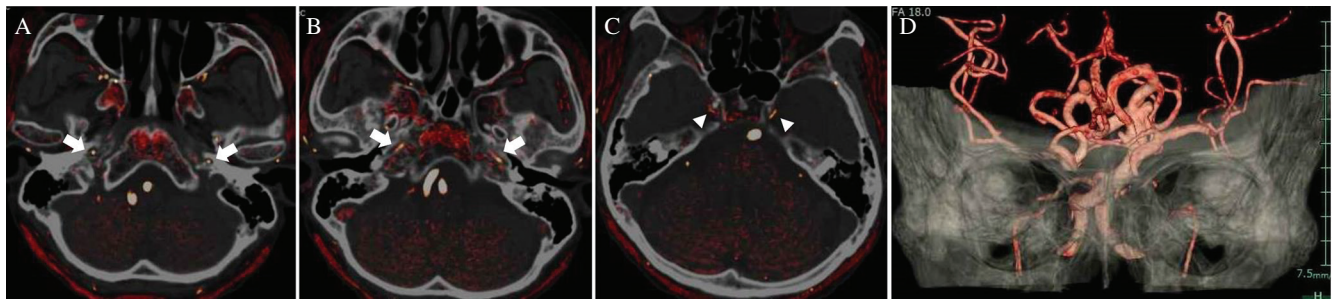


Fig. 4 Computed tomography (CT) and time-of-flight (TOF) fused images showing bilateral hypoplastic ICAs in each hypoplastic carotid canal (white arrows) (A, B), and the origin of the ICA (white arrowheads) (C). 3D CT and 3D TOF MR angiography fused image showing developed bilateral posterior communicating arteries and the small aneurysms (D).

may have a high tendency to bleed because of the increased blood flow in the parent arterial system,^{1,7,8)} and this condition may be life-threatening. Clipping or coil embolization for the prevention of this cerebral aneurysm rupture may be appropriate, even if the aneurysms have not ruptured or are asymptomatic.^{8–10)}

The patient and family might hesitate when faced with an endovascular procedure because of the increased risk, as in the present case. The reasons for the increased risk are that serious neurologic sequelae are more likely to develop because of the anatomical characteristics of the bilateral aplasia/hypoplasia of the ICAs if thromboembolic complications or cerebrovascular dissection were to occur as an adverse event. When we follow-up the aneurysms out of necessity, meticulous image follow-up of the aneurysms is desirable because of the risk of rupture of the aneurysms by hemodynamic stress. If the size of the aneurysms has changed, treatment to prevent rupture of the aneurysms should be strongly considered.

Conclusion

A case of long-term follow-up of unruptured intracranial aneurysms associated with segmental aplasia of bilateral ICAs was described.

Conflicts of Interest Disclosure

The authors declare that there are no conflicts of interest.

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