Ruptured Saccular Aneurysm of a Dolichoectatic Internal Carotid Artery in a Patient With Agenesis of the Contralateral Internal Carotid Artery
—Case Report—

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Abstract
A 42-year-old woman presented with very rare cases of ruptured saccular aneurysm of a dolichoectatic internal carotid artery (ICA) associated with agenesis of the contralateral ICA manifesting as sudden onset of severe headache and nausea without neurological deficits. Angiography and three-dimensional computed tomography demonstrated intraventricular hemorrhage with slight subarachnoid hemorrhage and dolichoectasia of the right ICA with agenesis of the contralateral ICA, as well as a saccular aneurysm of the ectatic right ICA. The aneurysm neck was clipped successfully. The patient remained ambulatory with no neurological deficits at discharge 15 days after the surgery. The saccular aneurysm in our case was formed in the dolichoectatic ICA, presumably due to both abnormal hemodynamics and abnormal arterial wall.

Key words: dolichoectasia, agenesis, internal carotid artery, saccular aneurysm

Introduction
Dolichoectasia and agenesis of the internal carotid artery (ICA) are both rare anomalies that can cause clinical stroke. The prevalence of dolichoectasia has been estimated as 0.05%3) to 0.06%,22) and that of agenesis of the ICA is thought to be less than 0.01%.1) We treated a patient with a ruptured saccular aneurysm of a dolichoectatic ICA associated with agenesis of the contralateral ICA.

Case Report
A 42-year-old woman suddenly developed severe headache and nausea, and was admitted to our hospital by ambulance. On admission she had no neurological deficits. Skull radiography showed calcification of the ICA. Computed tomography (CT) demonstrated calcification of the right ICA and intraventricular hemorrhage with slight subarachnoid hemorrhage (Fig. 1). CT using bone windows demonstrated absence of the left carotid canal (Fig. 2).

Cerebral angiography confirmed the absence of the left ICA and demonstrated a greatly enlarged and tortuous right ICA (Fig. 3A). The A1 segment of the left anterior cerebral artery (ACA) was hypoplastic, and both ACAs as well as the right middle cerebral artery (MCA) were supplied by the right ICA. The P1 segment of the right posterior cerebral artery (PCA) was absent. The right PCA was supplied by the right ICA via the right posterior communicating artery (PComA). The left MCA was supplied mainly by the basilar artery via the left PComA (Fig. 3B). The left ophthalmic artery was supplied by the left external carotid artery. CT with three-dimensional reconstruction also demonstrated the enlarged, irregularly contoured right ICA, as well as a saccular aneurysm on the right ICA (Fig. 4).

Surgery was performed via the right pterional...
Fig. 1 Computed tomography scans showing calcification of the right internal carotid artery and intraventricular hemorrhage with slight subarachnoid hemorrhage.

Fig. 2 Computed tomography scan using bone windows showing absence of the left carotid canal (arrows).

Fig. 3 A: Right internal carotid arteriograms, anteroposterior and lateral views, showing the enlarged and tortuous right internal carotid artery (ICA). B: Right vertebral arteriograms, Towne and lateral views, showing both anterior cerebral arteries and the right middle cerebral artery (MCA) are supplied by the right ICA. The P1 segment of the right posterior cerebral artery (PCA) is absent, and the right PCA is supplied by the right ICA via the dilated right posterior communicating artery (PComA). The left MCA is supplied mainly by the basilar artery via the left PComA.

Fig. 4 Computed tomography scan with three-dimensional reconstruction demonstrating the enlarged, tortuous right internal carotid artery with a saccular aneurysm (arrow).

approach. The dome of the aneurysm including the rupture point was buried in the inferior choroidal fissure. The right ICA was tortuous and calcified. However, the neck of the aneurysm was not calcified and was successfully clipped. Mild transient euphoria occurred during the postoperative course. The patient was discharged in ambulatory condition with no neurological deficits 15 days after the operation.

Discussion

Our patient presented with two extremely rare conditions, dolichoectatic left ICA and agenesis of the right ICA.

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Intracranial arterial dolichoectasia is characterized by enlargement, tortuosity, or elongation of the major arteries at the base of the brain, most often the distal vertebral arteries, basilar artery, or the distal ICA segments.\(^{6,9,22}\) Congenital factors have been implicated in the pathogenesis of dolichoectasia, including a defect or destruction of the internal elastic lamina.\(^{15}\) Several diseases also have been implicated, including Ehlers-Danlos syndrome,\(^6\) Marfan’s syndrome,\(^4\) tuberous sclerosis,\(^2\) and moyamoya disease.\(^{21}\) Hypertension and arteriosclerosis, both acquired factors, are also suspected to be involved.\(^{8,15–17,22}\) Dolichoectasia occurs particularly often in the vertebrobasilar system.\(^{5,8,18,22}\) Dolichoectasia is frequently associated with ischemia,\(^5,11,32\) and can also progress to arterial rupture with subarachnoid hemorrhage via dissection of the arterial wall.\(^{3,8,22}\)

In our case, a saccular aneurysm had formed on the dolichoectatic ICA, and then ruptured to cause intraventricular hemorrhage with slight subarachnoid hemorrhage. The saccular aneurysm was confirmed at operation and clipped successfully.

Agenesis of the ICA is thought to occur before the 5th and 6th weeks of gestation.\(^{10}\) The diagnostic radiologic findings are absence of the ICA by angiography and absence of the bony carotid canal by CT, as seen in this case.\(^7^,13^\) Cases of agenesis of the ICA can be classified as either fetal or adult type according to the supply of the ipsilateral MCA.\(^{10}\) In the fetal type, both ACAs are supplied by the contralateral ICA, whereas the ipsilateral MCA is supplied by the basilar artery via the PComA. In the adult type, both the ACAs and MCAs are supplied by the contralateral ICA. The fetal pattern suggests that ICA agenesis occurred before formation of the circle of Willis, whereas the adult pattern reflects agenesis occurring after formation of the circle. Most cases of ICA agenesis are the fetal type, which can cause enlargement or tortuosity of the PComA and/or vertebrobasilar system.\(^{20}\)

In our case, the ipsilateral MCA was supplied mainly by the posterior circulation, typical of the fetal type of ICA agenesis, whereas the contralateral ICA including the PComA was enlarged and tortuous. This rare condition may have resulted from compensative changes associated with the absence of the right P1 segment in addition to the ICA agenesis. Therefore, not only congenital factors but also hemodynamic stress may have caused the contralateral dolichoectatic ICA.

Up to one-third of cases of agenesis of the ICA are associated with subarachnoid hemorrhage from an aneurysm in the collateral circulation that has presumably arisen as a result of ongoing hemodynamic stress caused by the anomaly of the circle of Willis.\(^{1,4,15}\) The aneurysm in our case was formed in the contralateral dolichoectatic ICA. A saccular aneurysm in the dolichoectatic artery is very rare, excluding fusiform aneurysm. In addition to the congenital factor, hemodynamic stress resulting from the unique collateral circulation associated with agenesis of the ICA may have caused saccular aneurysm formation.

References


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Dolichoectasia and Agenesis of the ICA


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