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Agenesis of the Left Internal Carotid Artery Associated with Anterior Communicating Artery Aneurysm: Case Report

Anterior Kominikan Arter Anevrizması ile Sol İnternal Karotid Arter Agenezisinin Birlikteliği: Olgu Sunumu

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ABSTRACT

We present a rare case of agenesis of the left internal carotid artery in a 43-year-old woman, associated with an aneurysm of the anterior communicating artery and presenting with subarachnoid hemorrhage. The left internal carotid artery was not visualized on the left carotid angiogram. The left middle cerebral artery was perfused from the basilar artery via the dilated posterior communicating artery on vertebral angiogram. Absence of the left carotid canal was proven on temporal bone computed tomography. Absence of the left internal carotid artery was verified at operation. Absence of internal carotid artery is discussed in relation to aneurysm formation.

KEY WORDS: Agenesis, Internal carotid artery, Subarachnoid hemorrhage

ÖZ

43 yaşında, subaraknoid kanama ile başvuran ve anterior komunikan arter anevrizması ile birlikte sol karotis interna agenezi olan nadir bir kadın olgu sunduk. Sol karotis anjiyografisinde; sol karotis interna görüntülenemedi. Vertebral anjiyografide, sol orta serebral arter,dilate posterior komunikan arter yoluyla baziler arterden besleniyordu. Sol karotid kanal yokluğu, temporal kemik bilgisayarlı tomografide görüntülendi. Sol karotis interna yokluğu ameliyatta doğrulandı. Karotis interna yokluğu ve anevrizma oluşumu arasındaki ilişki tartışıldı.

ANAHTAR SÖZCÜKLER: Agenez, Karotis interna, Subaraknoid kanama

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INTRODUCTION

Agenesis, aplasia, or hypoplasia of the internal carotid artery are rare congenital developmental anomalies, occurring in less than 0.01% of the population (3,7,11).

Agenesis of internal carotid artery (ICA) is commonly asymptomatic under normal conditions. It is often detected as an incidental finding or after a cerebrovascular event, such as subarachnoid hemorrhage after rupture of a coincidental aneurysm or cerebral infarct (2,3,5,7,10,11).

In this paper, we reported a case of ICA aplasia associated with an aneurysm of the anterior communicating artery aneurysm and discuss its embryological etiologies, and neuroradiological and clinical features. The importance of altered hemodynamic forces on aneurysm formation produced by the agenesis of the internal carotid artery is emphasized.

CASE REPORT

A 43-year-old woman presented with acute onset of headache and vomiting. Neurological examination disclosed a diminished level of consciousness and nuchal rigidity.

A nonenhanced plain computed tomography scanning revealed subarachnoid hemorrhage and hydrocephalus.

The right carotid angiography demonstrated an anterior communicating aneurysm. The left anterior cerebral artery (ACA) system was supplied by the right side via the anterior communicating artery (ACoA).

The left common carotid angiography demonstrated the left external carotid system and complete absence of the cervical, petrous and cavernous left ICA (Figure 1).

Posterior circulation filled normally with selective vertebral injection and also the left middle cerebral artery (MCA) was supplied from the basilar artery via the dilated posterior communicating artery (PoCoA). An arch aortogram revealed normal origin and course of right common carotid, left external carotid and both vertebral arteries. The left common carotid artery showed no bifurcation in the neck, and terminated in the left external carotid artery. No internal carotid artery was visualised (Figure 2).

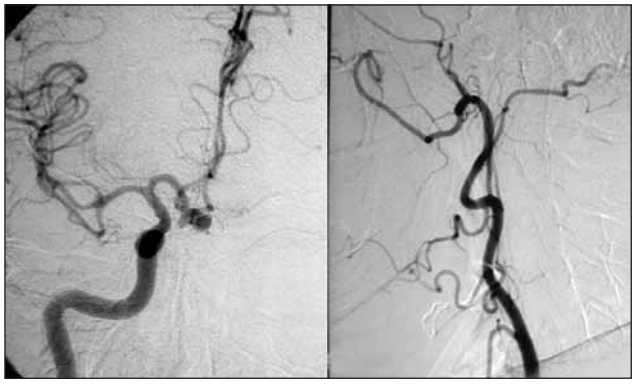

Figure 1: The right carotid angiography demonstrates an anterior communicating aneurysm. The left ACA system is supplied by the right side via the ACoA. The left CC angiography demonstrates the left external carotid system and complete absence of the cervical, petrous and cavernous left ICA.

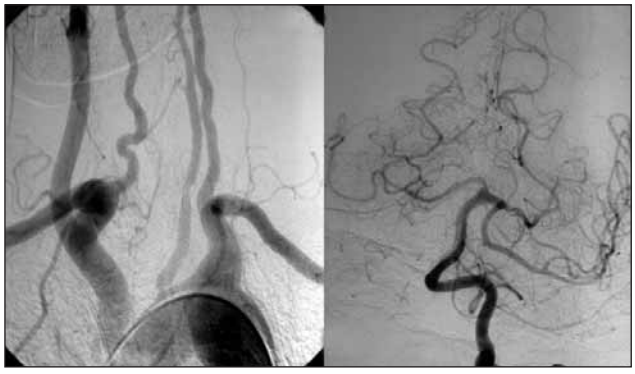

Figure 2: The left middle cerebral artery is supplied from the basilar artery via the dilated posterior communicating artery in selective vertebral injection. On the arch aortogram, the left common carotid artery shows no bifurcation in the neck, and terminates in the left external carotid artery.

A high-resolution computed tomography (CT) scanning of the base of the skull demonstrated a normal bony carotid canal on the right side, and complete absence on the left (Figure 3).

No surgical or interventional correction was attempted for ICA aplasia. Aneurysmal neck clipping by the pterional transsylvian approach on the right side was performed for the ACoA aneurysm. Complete absence of the left internal carotid artery was verified at operation. The postoperative course was uneventful and the patient was discharged without neurological deficits.

DISCUSSION

The first description of an absent internal carotid artery was made by Tode in 1787 (12). In 1954, the first

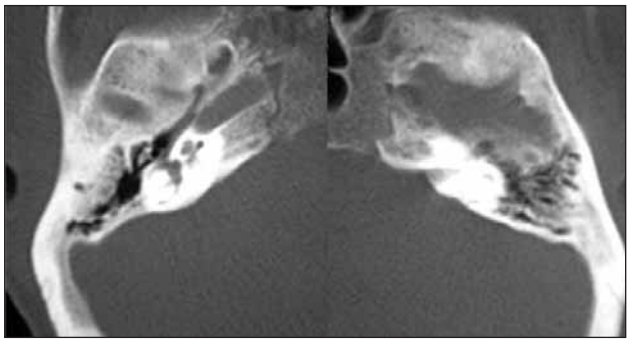

Figure 3: A high-resolution CT scan of the base of the skull demonstrates a normal bony carotid canal on the right side, and complete absence on the left.

case of ICA agenesis at cerebral angiography was reported by Verbiest (14). In 1980, Handa et al first reported the absence of the bony carotid canal (4).

The definitions of agenesis, aplasia, and hypoplasia of the ICA seem to be unclear and are often used as interchangeable. The diagnosis of aplasia or agenesis of the ICA was based on angiographic findings and the presence of an absent or hypoplastic bony carotid canal by temporal bone CT (2,3,5,11,15).

At the 4 to 5 mm embryonic stage, the primitive ICA arises from the terminal segments of the paired dorsal aorta and the third aortic arch. The distal parts of the ICAs originate from the dorsal ends of the first and third aortic arches. It is completely formed by about 6 weeks after fertilization (9). Agenesis or aplasia of the internal carotid artery is therefore most likely based on atresia or involution of the third aortic arches and the distal portion of the dorsal aortas at this stage (6).

The carotid channel develops in association with the ICA. The skull base does not begin to form until the 5th to 6th weeks of fetal life. If the embryonic primordium of the ICA fails to develop before the 3rd to 5th embryonic weeks, the ICA and the carotid channel cannot develop (5,10).

The diagnosis of congenital absence of an internal carotid artery requires no visualization of an internal carotid artery on angiography and absent carotid canals in the base of the skull on CT. Otherwise, it may be confused with acquired stenosis or occlusion of the ICA (5,6,15).

Unilateral absence or hypoplasia of ICA is more frequent on the left side. In reported cases, the ratio of right to left to bilateral agenesis of the internal carotid artery has been found to be 1:3:1 (1,2,3,7,15).

The absence of one or both internal carotid arteries may be entirely asymptomatic under normal conditions, due to the presence of the collateral vessels. These are transcranial anastomoses from the external carotid artery, persistent embryonic vessels and normal anastomotic pathways through the circle of Willis (1,3,7,11).

Tsuruta and Miyazaki classified three collateral circulatory configurations in patients with agenesis of one internal carotid artery (13). Lie summarized six basic patterns of collateral circulation in association with absence of the ICA (8).

In our case, the ACA territory was supplied from the contralateral ICA via the ACoA, and the MCA territory was supplied from the vertebrobasilar system via the PoCoA. This is a pattern type I of Tsuruta's or type A of Lie's classification.

As seen in our case, the anomaly is generally associated with additional vascular anomalies and especially intracranial intracranial aneurysms. (1,2,3,5,7,10,11).

The incidence of intracranial aneurysm in association with agenesis or aplasia has been reported as 24-67%, which is much higher than that found in the general population, 2-4% (1,2,6,7,10).

The increased occurrence of intracranial aneurysm formation may be due to the increased hemodynamic load on the normal side and the result of a developmental defect. Increased regional blood flow along with congenital defects of the vessel wall and systemic hypertension are important factors in the development of cerebral aneurysms (2,7).

CONCLUSION

Agenesis of ICA is an uncommon developmental vascular anomaly. It may be misdiagnosed as stenosis or occlusion of ICA.

Patients with agenesis of ICA present with subarachnoid hemorrhage because of the increased frequency of intracranial aneurysms. They should therefore be closely followed clinically and radiologically.

On the other hand, abnormal vascular anatomy should be carefully investigated prior to direct or endovascular surgery for a ruptured aneurysm in patients with agenesis of ICA.

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