

Congenital Hypoplasia of Internal Carotid Artery Accompanying with Cerebral Aneurysms

Geum-Seong Baek, M.D., Eun-Jeong Koh, M.D., Woo-Jong Lee, M.D., Ha-Young Choi, M.D.

*Department of Neurosurgery, Research Institute of Clinical Medicine, Chonbuk National University,
Medical School / Hospital, Jeonju, Korea*

Hypoplasia of the internal carotid artery is a rare congenital anomaly. Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are frequently associated with cerebral aneurysms in the circle of Willis. Authors report two cases with congenital hypoplasia of the ICA accompanying with the aneurysms. Transfemoral cerebral angiography (TFCA) in one patient identified nonvisualization of the left ICA. Bilateral anterior cerebral artery (ACA) and middle cerebral artery (MCA) were supplied from the right ICA accompanying with two aneurysms at anterior communicating artery (AcoA) and A1 portion of the left ACA. TFCA in another patient demonstrated hypoplastic left ICA and left ACA filled from the right ICA accompanying with AcoA aneurysm. Left MCA was filled from basilar artery via posterior communicating artery (PcoA). Skull base computed tomography (CT) in two patients showed hypoplastic carotid canal. Authors performed direct aneurysmal neck clipping. Follow up CT angiography (CTA) at one year after surgery did not show regrowth or new development of the aneurysm. In patients with hypoplastic ICA, neurosurgeons should be aware of the possibility of development of the aneurysms, presumably because of hemodynamic process. Direct aneurysmal neck clipping is a good treatment modality. After operation, regular CTA, magnetic resonance angiography (MRA) or TFCA is needed to find progressive lesion and to prevent cerebrovascular attack (CVA).

KEY WORDS : Hypoplasia · Internal carotid artery · Aneurysm · Hemodynamic process.

Introduction

Hypoplasia of the internal carotid artery are rare congenital anomalies. Together with agenesis and aplasia, this anomaly is seen in less than 0.01% of the population^{1,3)}. Agenesis, aplasia, and hypoplasia of the ICA are frequently associated with cerebral aneurysms of the circle of Willis, presumably because of increased circulatory stress that derives from altered hemodynamics. Incidence of the cerebral aneurysms in patients with hypoplasia of the internal carotid artery has been reported to be 25-43%, which is much higher than that found in the general population, 2 to 4%^{1,3,16)}. In contrast to findings in patients with acquired ICA occlusion, ipsilateral cerebral infarctions are not frequently observed in patients with congenital hypoplasia of the ICA, because collateral circulation from the contralateral ICA or vertebrobasilar system, or both, is usually developed.

Authors report two patients with hypoplasia of the left ICA.

One patient accompanied with unruptured multiple aneurysms in AcoA and A1 of the left ACA. Another patient accompanied with ruptured solitary aneurysm in the AcoA.

Case Report

Case 1

A 58-year-old man presented with dysarthria and general weakness that developed abruptly days prior to admission and resolved spontaneously at seventh hospital day. He had no hypertension, diabetes mellitus, smoking history, and family history of CVA.

In computed tomography, subarachnoid hemorrhage or infarction were not shown. Brain MRI showed only small old infarctions in the bilateral basal ganglia (Fig. 1A). Three dimensional CTA (3D-CTA) and TFCA showed abnormal angiographic findings with nonvisualization of the left ICA distal to the bifurcation of the left common carotid artery

• Received : December 18, 2006 • Accepted : March 6, 2007

• Address for reprints : Eun-Jeong Koh, M.D., Department of Neurosurgery, Research Institute of Clinical Medicine, Chonbuk National University, Medical School / Hospital, 634-18 Geumam-dong, Deokjin-gu, Jeonju 561-712, Korea Tel : +82-63-250-1870, Fax : +82-63-277-3273, E-mail : dmsqfns@paran.com

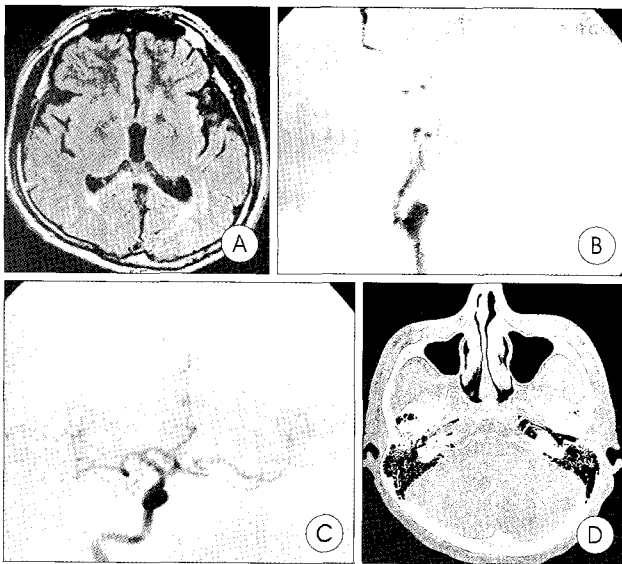


Fig. 1. A : Brain magnetic resonance imaging showing small old infarctions in the bilateral basal ganglia. B : Left common carotid angiogram demonstrating abrupt discontinuation of left internal carotid artery (ICA). C : Transfemoral cerebral angiography shows bilateral anterior cerebral artery (ACA) and middle cerebral artery (MCA) supplying from the right ICA accompanying with two aneurysms at anterior communicating artery (white arrow) and A1 portion of the left ACA (arrow head). D : Skull base computed tomography revealing hypoplastic left carotid canal (white arrows).

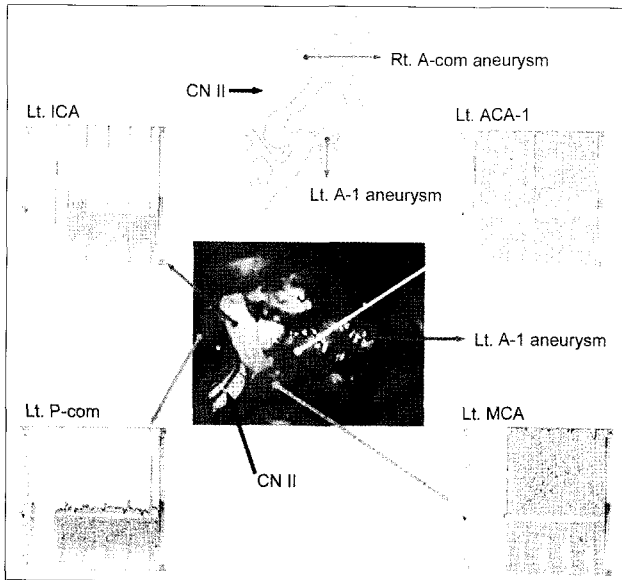


Fig. 2. Intraoperative doppler shows left anterior cerebral artery A1 portion (ACA-1) and middle cerebral artery (MCA) with normal direction of blood flow, and left internal carotid artery (ICA) and posterior communicating artery (PcoA) with a scanty of retrograde blood flow.

(Fig. 1B). Bilateral ACA and MCA were supplied from the right ICA accompanying with two aneurysms at AcoA and A1 portion of the left ACA (Fig. 1C). Left cerebral hemisphere was mainly supplied from right ICA via AcoA and extracranial-intracranial (EC-IC) anastomosis via ophthalmic artery was identified. Left PcoA was not visualized and, whereby, there



Fig. 3. Three dimensional computed tomographic angiogram reveals two aneurysm clips at anterior communicating artery (large arrow) and left A1 portion (arrow head) after twelve months. There is no remnant aneurysm or newly developed aneurysm. Small arrow is right A1 portion.

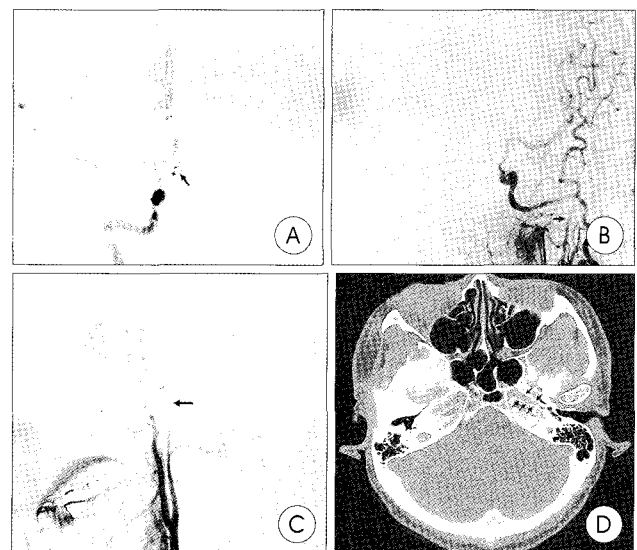


Fig. 4. A : Transfemoral cerebral angiography (TFCA) shows right filling anterior communicating artery (AcoA) aneurysm (black arrow) and left anterior cerebral (ACA) filled from right internal carotid artery (ICA). B, C : TFCA demonstrates hypoplastic left ICA and left middle cerebral artery (MCA) filled from hypoplastic ICA (black arrow). D : Skull base computed tomography reveals hypoplastic left carotid canal (black arrows).

was no connection between the anterior and posterior circulation.

Skull base CT, which is able to make a distinction between congenital absence and acquired occlusion of the ICA, revealed hypoplastic left carotid canal (Fig. 1D). Findings from this study was sufficient to make a diagnosis of the congenital hypoplasia of ICA.

He had two stage direct aneurysmal neck clipping, the first for right feeding AcoA aneurysm via right pterional approach and the second for left A1 aneurysm via left pterional approach at intervals of two weeks. It was difficult to clip the left A1 aneurysm via right side approach because the aneurysmal sac was projecting to right side. During surgery of the left A1 aneurysm, rudimentary left ICA and PcoA were observed. Intracranial doppler indicated retrograde blood flow in the left ICA and scanty of blood flow in the left PcoA (Fig. 2).

Tortuous, thin and weak arterial wall near the aneurysmal sac was reinforced with cotton sheet and fibrin glue. After surgery, he recovered without any neurologic deficit.

Follow up 3D-CTA at one year after surgery did not show regrowth or new development of the aneurysm (Fig. 3).

Case 2

A 38-year-old woman was admitted with severe headache. CT showed subarachnoid hemorrhage in all cisterns. 3D-CTA showed nonvisualization of the left ICA accompanying with AcoA aneurysm. The left ACA was filled from the right ICA. Ruptured AcoA aneurysm was fed from the right ICA (Fig. 4A). TFCA, however, demonstrated hypoplastic left ICA which was unable to look out in 3D-CTA and left MCA filled from basilar artery via PcoA. TFCA also showed left MCA supplied from hypoplastic left ICA and basilar artery via enlarged PcoA (Fig. 4B, C). EC-IC anastomosis was not identified. Skull base CT revealed hypoplastic left carotid canal (Fig. 4D), which is an evidence of congenital hypoplasia of ICA.

She had a direct aneurysmal neck clipping. After clipping, authors strengthen the tortuous, thin arterial wall with cotton sheet and fibrin glue. She recovered without any neurologic deficit.

Discussion

The definitions of agenesis, aplasia, and hypoplasia of the ICA seem to be unclear and are often used interchangeably. Lie¹³ defined 'agenesis' as the total absence of the entire artery due to an embryological arterial developmental failure, and used the terms 'hypoplasia' and 'aplasia' to describe the situation when a portion, or remnant, of the artery was present and when the initial segment of the artery is normal in size or even slightly enlarged proximal to its abrupt narrowing.

When the ICA is congenitally absent or hypoplastic, collateral circulation develops through the circle of Willis from the opposite ICA and/or the basilar artery to supply the involved hemisphere. Therefore, neurologic deficits are few; however, these anomalies are frequently associated with cerebral aneurysm^{1,7,9}. Previous studies have reported aneurysms on Willis circle at PcoA or AcoA or basilar tip in relation to development of ICA obstruction¹². First case of our patients with unruptured multiple aneurysms had complained only transient dysarthria and general weakness. On admission, neurological examinations implied cerebral infarction. Brain MRI showed only small lacunar infarction. Brain CTA and TFCA visualized complete obstruction of the left ICA accompanying multiple aneurysms in AcoA and left distal A1 portion which was very unusually located. Second case presented headache due to subarachnoid

hemorrhage by AcoA aneurysmal rupture.

In case of absence of the ICA, the pattern of collateral blood flow to the distal ICA and intracranial vasculature is dependent upon the state at which the disruption occurred. Cali et al.² postulated that primitive pathways of collateral circulation would prevail if the disruption occurred before completion of the circle of Willis. Likewise, collateral flow through the circle of Willis would dominate if the disruption occurs after 24-mm stage of development. Lie¹³ described six pathways of collateral circulation in association with absence of ICA. In type A, unilateral absence of the ICA is associated with collateral circulation to the ipsilateral ACA through a patent AcoA and the ipsilateral MCA from the posterior circulation through a hypertrophied PcoA. In type B, the ipsilateral ACA and MCA are supplied across a patent AcoA. Type C represents bilateral agenesis of ICA with supply to the anterior circulation via carotid-vertebrobasilar anastomosis through hypertrophied PcoA. Type D represents unilateral agenesis of the cervical portion of the ICA with an intercavernous communication to the ipsilateral carotid siphon from the contralateral cavernous ICA. In type E, diminutive ACAs are supplied by bilateral hypoplastic ICAs, and the MCAs are supplied by enlarged PcoAs. The type F provides collateral flow to the distal ICA via transcranial anastomoses from the internal maxillary branches of the extracranial artery system, the so-called rete mirabile. Our first case belongs to Lie's type B and second case comes under Lie's type A.

ICA dysgenesis is known to occur more frequently in the left side^{6,16}. Our two cases also presented ICA hypoplasia in the left side.

Brain MRA, CTA, and TFCA may not enable one to clearly distinguish between hypoplasia and agenesis, and may suggest acquired stenosis or occlusion of the ICA. Distinctions have clinical implications, as the most common pattern of collateral flows in cases of acquired occlusion of the ICA is similar to the type B pattern seen with unilateral absence⁸. In such cases, skull base CT may be obtained because ICA forms by the 4th embryonic week, while skull base forms at 5-6 weeks, agenesis or hypoplasia of the ICA can be differentiated by identification of the carotid canal^{4-6,8,15}. If the carotid canal is absent, it is agenesis of ICA. If the carotid canal is small, it is aplasia or hypoplasia of ICA. If the canal is of normal size, an acquired lesion is likely. In our cases, skull base CT revealed clearly hypoplastic carotid canal in the left side. This finding is sufficient to make a distinction of congenital hypoplasia of ICA in both cases.

Most patients with congenital ICA agenesis, aplasia, and hypoplasia live long time without any neurologic symptom. However, some patients showed symptoms related with cerebral ischemia because of decreased blood flow, and enlargement

of collateral vessels may lead to associated intracranial aneurysm with obvious problem^{1,10,11,14}. Therefore, regular follow up brain CTA, MRA or TFCA should be done to avoid life threatening CVA.

When aneurysm is detected, direct aneurysmal neck clipping is a good treatment modality. Under direct vision, aneurysm can be resolved and weak surrounding vascular wall can be supported by wrapping with thin cotton sheet and fibrin glue.

Conclusion

Authors report two patients with obstructed hypoplastic ICA accompanying with development of aneurysm. Skull base CT is considered of importance in differentiation congenital type from acquired type of ICA occlusion.

Authors like to recommend to do a direct aneurysmal neck clipping and a reinforcement of weak vascular wall surrounding aneurysm in case associated with ICA hypoplasia. And regular follow up with CTA or MRA or TFCA are also recommended to monitor progressive vascular lesion and to prevent against CVA.

References

1. Afifi AK, Goderski JC, Menezes A : Cerebral hemiatrophy, hypoplasia of internal carotid artery and intracranial aneurysms : A rare association occurring in an infant. *Arch Neurol* 44 : 232-235, 1987
2. Cali RL, Berg R, Rama K : Bilateral internal carotid agenesis : a case study and review of the literature. *Surgery* 113 : 227-233, 1993
3. Chen CJ, Chen ST, Hsieh FY, Wang LJ, Wong YC : Hypoplasia of the internal carotid artery with intercavernous anastomosis. *Neuroradiology* 40 : 252-254, 1998
4. Czarnecki EJ, Silbergleit R, Mehta BA, Sanders WP : Absence of the supraclinoid internal carotid artery in association with intracranial aneurysms. *Neuroradiology* Jan 40 : 11-14, 1998
5. Dinc H, Gumele HR, Kuzeyli K, Baykal S : Unilateral Agensis of Internal Carotid Artery with Subarachnoid Hemorrhage : Report of Two Cases. *Int J Angiol* 8 : 157-160, 1999
6. Florio F, Balzano S, Nardella M, Strizzi V, Cammisa M, Bozzini V et al : Congenital absence of the internal carotid artery. *Cardiovasc Intervent Radiol* 22 : 74-78, 1999
7. Goldstein SJ, Lee C, Young AB, Guidry GJ : Aplasia of the cervical internal carotid artery and malformation of the circle of Willis associated with Klippel-Trenaunay syndrome. Case report. *J Neurosurg* 61 : 786-789, 1984
8. Hattori T, Kobayashi H, Inoue S, Sakai N : Persistent primitive trigeminal artery associated with absence of internal carotid artery. *Surg Neurol* 50 : 352-355, 1998
9. Ide M, Jimbo M, Yamamoto M, Hagiwara S : Posterior cerebral artery aneurysm associated with unilateral internal carotid artery agenesis-Case report. *Neurol Med Chir (Tokyo)* 35 : 825-829, 1995
10. Jung BO, Lee JI, Choi JH, Kim SK, Jo YJ : Congenital Hypoplasia of the Bilateral Internal Carotid Artery-A Case Report-. *J Korean Neurosurg Soc* 31 : 282-284, 2002
11. Jung H, Ahn YB, Lee SK, Park MS : Bilateral Hypoplasia of Interna; Carotid Arteries Associated with Intracranial Aneurysm. *J Korean Neurosurg Soc* 25 : 211-216, 1996
12. Lee JH, Oh CW, Han DH : Aplasia of the internal carotid artery. *Acta Neurochir* 145 : 117-125, 2003
13. Lie TA : Congenital anomalies of the carotid arteries : An angiographic study and a review of the literature. *Amsterdam : Excerpta Medica Foundation* : 30-35, 1968
14. Midkiff RB, Boykin MW, McFarland DR : Agenesis of the internal carotid artery with intercavernous anastomosis. *Am J Neuroradiol* 16 : 1356-1359, 1995
15. Pilleul F, Guibaud L, Badinand N, Rouviere O, Pracros JP : Bilateral internal carotid agenesis : value of CT angiography and correlation to embryogenesis. *Eur Radiol* 11 : 858-860, 2001
16. Teal JS, Naheedy MH, Hasso AN : Total agenesis of internal carotid artery. *AJR* 1 : 435-442, 1980