Imaging Classification of Internal Carotid Artery Hypoplasia Based on Distal Ophthalmic Segment Occlusion

Zhiyong Zhang, MD,* Zhiqin Liu, MD,† and Zunjing Liu, MD*

Background: Internal carotid artery (ICA) hypoplasia (ICAH) is rare. The classification of ICAH is largely unclear. The aim of the study is to propose a new imaging classification for ICAH based on the occlusion of the distal ophthalmic segment and discuss the clinical and radiological differences between the different types. *Materials and Methods:* This was a retrospective study of patients with congenital ICAH diagnosed at the Department of Neurology of the China-Japan Friendship Hospital between June 2011 and June 2016. The patients underwent temporal bone computed tomography (CT), brain CT, cranial magnetic resonance imaging, transcranial Doppler, and head and neck CT angiography. *Results:* A total of 20 ICAH patients were divided into the distal occlusion (12 cases; 60%) and nondistal occlusion (8 cases; 40%) types based on whether the distal ophthalmic segment was occluded. The frequencies of collateral circulation from the circle of Willis ($P = .01$) and dilated cerebrovascular lesions ($P = .001$) in the distal occlusion type was higher than in the nondistal occlusion type. Five (25%) patients developed adverse cerebrovascular events during followup: 3 ischemic cases were of the nondistal occlusion type, and 2 cases with subarachnoid hemorrhage were of the distal occlusion type. *Conclusions:* A novel classification of ICAH was revealed based on the occlusion of the distal ophthalmic segment. The 2 types may show differences in collateral circulation patterns, coexisting cerebrovascular abnormalities, and potential clinical outcomes.

Key Words: Internal carotid artery hypoplasia—bony carotid canal—collateral circulation—posterior communicating artery—intracranial aneurysm

Introduction

Internal carotid artery hypoplasia (ICAH) is a rare congenital vascular anomaly that occurs in less than .01% of the world population.[1-3] Agenesis or aplasia of the internal carotid artery (ICA) signifies the complete absence of the ICA, whereas in ICAH, development is incomplete and is accompanied by hypoplasia of the ipsilateral bony carotid canal (BCC). Most patients with ICAH are asymptomatic owing to sufficient collateral circulations and they are usually diagnosed incidentally during angiographic examinations. Nevertheless, some ICAH patients are often misdiagnosed with other diseases such as carotid dissection, fibromuscular dysplasia, and severe atherosclerotic stenosis owing to similarities in radiological findings[4,5] and improper management is performed. So the imaging features of ICAH should be clearly distinguished for an accurate differential diagnosis.

ICAH is usually associated with other abnormalities, and the aneurysm is the most common associated condition.[6-9] Previous studies have confirmed that the frequency of intracranial aneurysm in patients with ICAH is significantly higher than in the general population.[6-8] Nevertheless, the association between ICAH and ischemic

From the *Department of Neurology, China-Japan Friendship Hospital, Beijing, China; and †Department of Neurology, Xi'an Central Hospital, Xi'an Jiao tong University School of Medicine, Xi'an, China.

Received May 2, 2019; revision received July 7, 2019; accepted July 21, 2019.

Financial Disclosure: This work was supported by the National Natural Science Foundation of China [grant number 81173595], and Research Fund of the China-Japan Friendship Hospital [grant number 2015-2-QN-39].

Address correspondence to Zhiyong Zhang, MD and Zunjing Liu, MD, Department of Neurology, China-Japan Friendship Hospital, No. 2 East Street, Sakura garden, Chaoyang District, Beijing 100029, China. E-mails: zhiyong_zhang888@126.com, liuzunjing@163.com.

1052-3057/$ - see front matter

https://doi.org/10.1016/j.jstrokecerebrovasdis.2019.104304

stroke has not yet been clarified.[4] Previous studies, which are mainly clinical case reports, display the variously associated anomalies and complex cerebrovascular collateral circulation patterns.[6-15] The imaging classification of ICAH remains unclear because of the low rate of detection. The early diagnosis and precise classification of ICAH may bring potential benefits in preventing the occurrence of cerebrovascular adverse events.

The aim of this study was to explore an imaging classification of ICAH based on the radiological characteristics, and investigate the collateral circulation patterns, coexisting cerebrovascular abnormalities and potential clinical outcomes from different imaging types.

Materials and Methods

Study Design and Patients

This was a retrospective study of patients with congenital ICAH from a registry of consecutively enrolled patients with ICA lesions diagnosed at the Department of Neurology of the China-Japan Friendship Hospital between June 2011 and June 2016. The patients had to have undergone the following imaging: temporal bone computed tomography (CT), brain CT, cranial magnetic resonance imaging (MRI), transcranial Doppler, and head and neck CT angiography (CTA).

Imaging and Analysis

The temporal bone CT was performed to observe the structure and morphology of the BCC. Brain CT and cranial MRI were performed to observe whether theses patient had stroke. Transcranial Doppler, head and neck CTA were performed to observe the intracranial and extracranial vascular conditions and evaluate the collateral circulation. Some patients further received digital subtraction angiography to determine the types of collateral circulation more clearly.

Temporal bone CT, brain CT, and head and neck CTA were performed using a 256-slice (Brilliance iCT, Philips, Best, the Netherlands) or 320-slice CT scanner (Aquilion ONE, Toshiba, Tokyo, Japan). Cranial MRI was performed using a 1.5-T (Signa 1.5T Excite HD, GE Healthcare, Waukesha, WI) or a 3.0-T MRI scanner (Ingenia 3.0T, Philips, Best, Netherlands). Transcranial Doppler was performed using an EMS-9U instrument (Delicasz, Shenzhen, China). Digital subtraction angiography was conducted using an angiography system (Allura Xper FD20, Philips, Best, Netherlands). Imaging data stored in the Picture Archiving and Communication Systems (Neurosoft 3.0, Shenyang, China) were reviewed and analyzed by 2 experienced neuroradiologists together.

The diagnosis of ICAH was primarily based on the presence of ICA narrowing on angiographic imaging and ipsilateral small BCC on temporal bone CT [3]. The imaging findings were evaluated as follows: (1) morphology and diameter of hypoplastic ICA and structure of intracranial segment of the hypoplastic ICA; (2) morphology and diameter of diminutive BCC; (3) collateral circulation patterns of the ipsilateral anterior circulation (AC); (4) abnormalities of other cerebral vessels; and (5) ischemia or infarction in the ipsilateral AC. The distal ophthalmic segment was observed for occlusion, as well as the presence of collaterals and the circle of Willis. Patients with enlargement of cerebrovascular beyond the average diameter cerebrovascular vessels or intracranial aneurysm were considered as dilated cerebrovascular lesions.

Follow-up

All enrolled patients underwent regular outpatient follow-up for at least 1 year. Adverse clinical events (cerebrovascular events including transient ischemic attack, cerebral infarction, cerebral hemorrhage, subarachnoid hemorrhage (SAH), myocardial infarction, and all-cause mortality) were recorded.

Statistical Analysis

All statistical analyses were performed using SPSS 17.0 (IBM, Armonk, NY). All continuous variables were expressed as means ± standard deviation and tested using the independent sample's t test after Kolmogorov-Smirnov tests. Categorical variables were presented as count and percentage, and tested using the chi-square or Fisher's exact test, if appropriate. A 2-sided P value less than .05 was considered statistically significant.

Results

Imaging Features and Classification of ICAH

Twenty patients were included (51.6 ± 14.2 years of age; 60% male). A total of 24 hypoplastic ICAs were discovered including 4 patients with bilateral ICAH. The mean diameter was 1.6 mm (range, 1.4-1.9 mm). The diminutive ICAs in all patients were accompanied by evenly narrowed ipsilateral BCCs, with an average diameter of 1.8 mm (range, 1.5-2.0 mm). The multiplanar reformation of CTA clearly demonstrated the presence of hypoplastic ICA and BCC.

The angiographic images showed that all hypoplastic ICAs presented luminal narrowing from the distal end of the carotid sinus to the ophthalmic artery level, with a normal carotid sinus, followed by either continual narrowing or occlusion distally. ICAH was divided into 2 types based on the occlusion of the distal ophthalmic segment: distal occlusion type (12 cases; 60%) and nondistal occlusion type (8 cases; 40%) (Fig 1), and there was no other transitional pattern. All 4 patients with bilateral ICAH were with occlusion in the distal ophthalmic segment. No significant difference was observed between the 2 types regarding gender, age, symptoms, ICA diameter, BCC diameter, other abnormalities, ischemia, and adverse clinical events (Table 1).

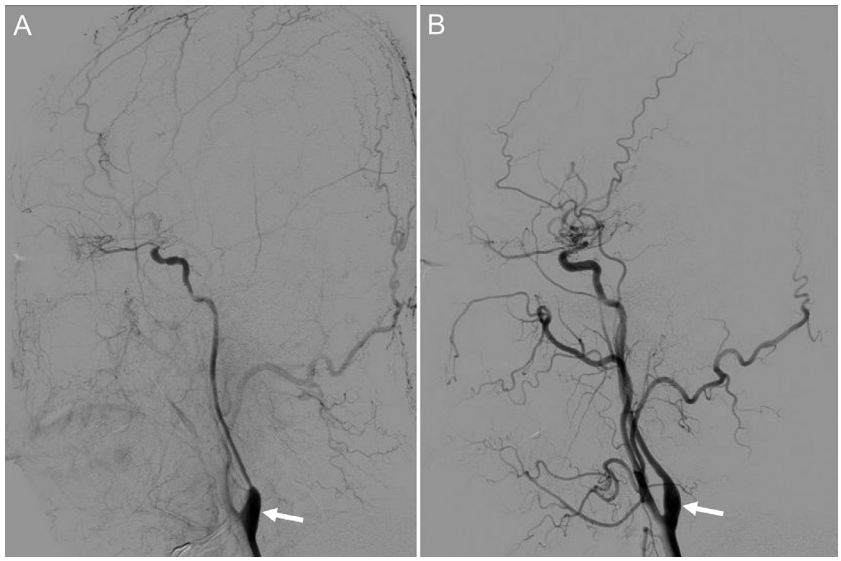

Figure 1. *Two types of internal carotid artery (ICA) hypoplasia based on the distal ophthalmic occlusion. (A) A lateral digital subtraction angiography (DSA) shows a hypoplastic ICA terminating to the ophthalmic artery level, followed by complete occlusion distally, which is the distal occlusion type. (B) A lateral DSA shows a hypoplastic ICA from the distal carotid sinus to the terminal segment, which is the nondistal occlusion type. Lateral DSA also shows the carotid sinus was not involved (white arrows).*

Table 1. *Characteristics of the patients according to the presence of occlusion in the distal ophthalmic segment*

	All (n = 20)	Distal occlusion (n = 12)	Nondistal occlusion (n = 8)	*P* value
Age (mean ± SD years)	51.6 ± 14.2	55.3 ± 12.4	49.5 ± 15.2	.944
Gender (male)	12 (60%)	7 (58.3%)	5 (62.5%)	>.99
Asymptomatic	19 (95%)	12 (100%)	7 (87.5%)	.834
ICA diameter (mean ± SD, mm)	1.6 ± .2	1.6 ± .3	1.6 ± .2	.54
BCC diameter (mean ± SD, mm)	1.8 ± .2	1.7 ± .2	1.8 ± .2	.318
Collateral circulation from the circle of Willis	13 (65%)	11 (91.7%)	2 (25%)	.01
Dilated cerebrovascular lesions	13 (65%)	12 (100%)	1 (12.5%)	.001
Ischemia or infarction in ipsilateral AC	7 (35%)	4 (33.3%)	3 (37.5%)	>.99
Other associated abnormalities	10 (50%)	6 (50%)	4 (50%)	>.99
Adverse clinical events	5 (25%)	3 (25%)	2 (25%)	>.99

Abbreviations: AC, anterior circulation; BCC, bony carotid canal; ICA, internal carotid artery; SD, standard deviation.

Collateral Circulation Patterns

In the patients with the distal occlusion type, 11 patients (91.7%) showed the collateral contribution to the affected AC via the ipsilateral posterior communicating artery (PcomA). Of these, 9 patients also showed anastomoses of the distal leptomeningeal arteries between the ipsilateral posterior cerebral artery (PCA) and the middle cerebral artery (MCA) (Fig 2). One patient presented with intercavernous anastomosis. The collateral circulation patterns in the nondistal occlusion type were multiform (Table 2). The frequency of collateral circulation from the circle of Willis was observed less frequently than that in the distal occlusion type (25% versus 91.7%, $P = .01$) (Table 1).

Coexisting Cerebrovascular Abnormalities

Thirteen (65%) patients presented with PcomA structural changes, mainly arterial tortuosity, extension, and dilation (Fig 3). Among them, 3 (15%) patients developed PcomA aneurysm, 2 (10%) showed dilation of the ipsilateral PCA, and 4 (20%) showed dilation of the bilateral vertebral arteries (these patients had bilateral ICAH). The frequency of dilated cerebrovascular lesions in the distal occlusion type was significantly higher than that in the nondistal occlusion type (100% versus 12.5%, $P = .001$) (Table 1).

Furthermore, other cerebrovascular abnormalities were found in 10 (50%) patients: 4 (20%) with hypoplasia of the bilateral MCA and anterior cerebral artery (ACA), 2 (10%) with vertebrobasilar hypoplasia, 2 (10%) with contralateral fetal PCA, 1 (5%) with vertebrobasilar dolichoectasia, and 1 (5%) with an aberrant vertebral artery.

Ischemia or Infarction in Ipsilateral AC

Cranial MRI showed that 7 (35%) patients had ischemic lesions in the ipsilateral AC. These lesions manifested as

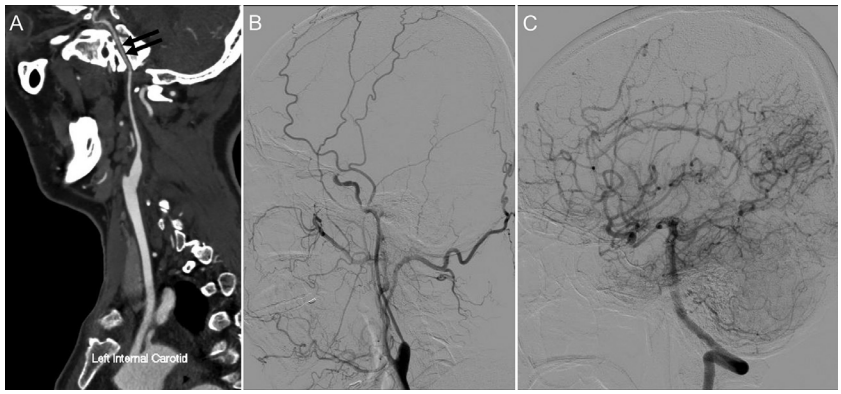

Figure 2. *Collateral circulation patterns in the distal occlusion type. (A) The multiplanar reformation of computed tomography angiography (CTA) clearly shows the hypoplastic internal carotid artery hypoplasia (ICA) and bony carotid canal (double black arrows). (B) Lateral digital subtraction angiography (DSA) reveals the occlusion of the distal ophthalmic segment, (C-D) collateral channels from the opening of the ipsilateral posterior communicating artery (PcomA), and anastomoses of the distal leptomeningeal arteries between the ipsilateral posterior cerebral artery and middle cerebral artery.*

Table 2. *Collateral circulation patterns and prognosis in patients*

Number	Sex/Age	Distal ophthalmic occlusion	Right/Left	Collateral circulation patterns	Prognosis	Follow-up time (months)
1	M/75	No	Right	Pcom A + Acom A	Well	15
2	F/33	No	Left	PCA-MCA + ACA-MCA + ICA-ECA	Well	12
3	M/57	No	Right	Acom A + ACA-MCA + Moya	Well	14
4	M/60	No	Left	PCA-MCA + Moya	CI	16
5	F/52	No	Left	ACA-MCA + PCA-MCA	TIA	26
6	M/48	No	Right	ACA-MCA + PCA-MCA	Well	12
7	M/62	No	Left	PCA-MCA + retemirable	CI	13
8	F/50	No	Left	ACA-MCA + PCA-MCA + ICA-ECA	Well	14
9	F/73	Yes	Bilateral	Pcom A+ PCA-MCA	Well	14
10	M/36	Yes	Bilateral	Pcom A + PCA-MCA	Well	12
11	M/30	Yes	Bilateral	Pcom A + PCA-MCA	Well	15
12	M/60	Yes	Bilateral	Pcom A + PCA-MCA	SAH	18
13	F/28	Yes	Left	Pcom A + PCA-MCA	Well	16
14	F/67	Yes	Left	Pcom A + PCA-MCA	SAH	14
15	M/55	Yes	Left	Pcom A + PCA-MCA	Well	20
16	M/48	Yes	Right	Pcom A + PCA-MCA	Well	21
17	F/52	Yes	Right	Pcom A + PCA-MCA	Well	14
18	F/62	Yes	Right	Pcom A	Well	14
19	M/39	Yes	Left	Pcom A	Well	16
20	M/42	Yes	Left	Intercavernous anastomosis	Well	12

Abbreviations: ACA-MCA, anastomosis between ACA and MCA; AcomA, anterior communicating artery; CI, cerebral infarction;ICA-ECA, anastomosis between ICA and external carotid artery; Moya, moyamoya-like collateral vessels; PCA-MCA, anastomosis between PCA and MCA; PcomA, posterior communicating artery; TIA, transient ischemic attack; SAH, subarachnoid hemorrhage; Well, no adverse clinical events.

old lacunar infarctions (n = 3, 15%) or ischemic lesions (n = 4, 20%) sporadically distributed in the cerebral cortex or subcortical white matter. DWI did not detect any abnormal fresh infarction.

Follow-up

The median follow-up was 15 months (range, 12-26 months). Five of 20 (25%) patients developed adverse cerebrovascular events during follow-up: 3 (15%) with cerebral ischemic events in the ipsilateral AC, including 1 case of TIA and 2 cases of new cerebral infarction; and 2 (10%) with SAH. Based on the imaging data, ischemic patients were of the nondistal occlusion type, while 2 patients with SAH secondary to rupture of PcomA aneurysm were of the distal occlusion type. Moreover, these ischemic patients had no collateral circulation via the circle of Willis (Table 2), and CT perfusion showed deficient cerebral blood flow in the affected AC.

Discussion

ICAH is rare and its clinical and imaging characteristics and classification system have not yet been clarified.

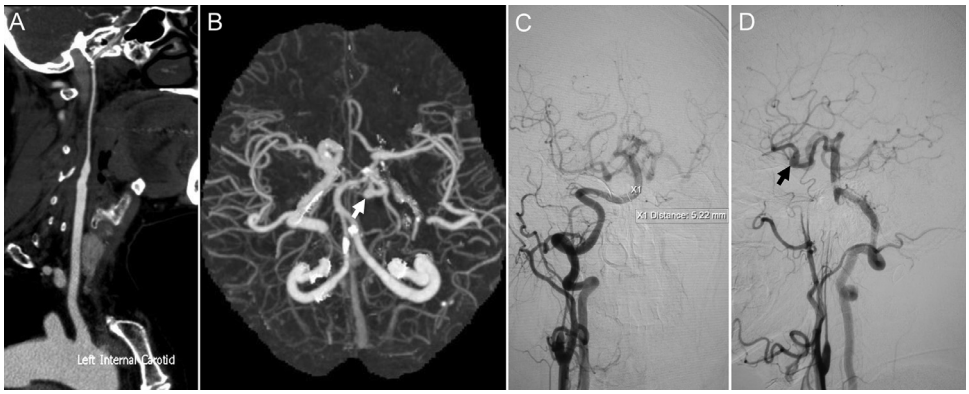

Figure 3. *Two cases of hemodynamic posterior communicating artery (PcomA) structural changes. Case-1, 55-year-old man (A-B). (A) The head and neck computed tomography angiography (CTA) shows internal carotid artery hypoplasia (ICAH) on the left side and ipsilateral bony carotid canal (BCC) hypoplasia, (B) collateral channel to left anterior circulation (AC) through ipsilateral PcomA, and structural changes of left PcomA including tortuosity, extension, and dilation (white arrows). Case-2, 60-year-old man (C-D). (C) The posteroanterior digital subtraction angiography (DSA) reveals right ICAH, collateral channels to bilateral ACs via bilateral posterior communicating arteries, and tortuosity with expansion of vertebrobasilar artery diagnosed as vertebrobasilar dolichoectasia. (D) The lateral DSA further identifies aneurysmal formation of PcomA (black arrow).*

Therefore, the present study aimed to propose a new imaging classification for ICAH based on the occlusion of the distal ophthalmic segment and discuss the difference in collateral circulation patterns, coexisting cerebrovascular abnormalities and potential clinical outcomes, which will have to be confirmed.

ICAH is commonly seen unilaterally, and the bilateral occurrence is even rarer.[5,6,16,17] The primordial ICA forms gradually during the first 3-4 weeks of embryonic development and the BCC develops from the embryonic primordium of the ICA during 5-6 weeks.[7] Therefore, the imagine diagnosis of ICAH primarily focuses on 2 aspects: ICA narrowing revealed by angiographic imaging and ipsilateral BCC hypoplasia confirmed by temporal bone CT.[3] Compared with the other examinations, our study showed that head and neck CTA not only revealed the structure of the entire ICA and abnormalities of other cerebral arteries, but also allows visualization of the morphology and diameter of the BCC through the multiplanar reformation of CTA.

In our study, the featured angiographic imaging of ICAH showed luminal narrowing of the affected ICA from the distal end of the carotid sinus. This phenomenon can also be observed in distal hypoperfusion owing to dissection or severe atherosclerotic stenosis of the initial segment of the ICA or carotid fibromuscular dysplasia.[4,5] In addition to the specific imaging of these diseases, such as the double-lumen sign or string-of-beads sign, hypoplasia of the ipsilateral BCC is a key factor for differential diagnosis.[7] In this study, CTA confirmed that all hypoplastic ICAs were accompanied by the narrowing of BCCs (within 2 mm). Interestingly, we found that the diameter of carotid sinus in patients with ICAH was normal. In 2006, Yasaka et al reported that this sign can occur in moyamoya disease, and which was firstly called "champagne bottle neck sign."[18] Although this sign has been as a characteristic feature of Moyamoya disease,[18,19] this imaging finding is not unique to moyamoya disease, and can also be seen in patients with moyamoya syndrome.[20] Due to the rare and special morphological feature, we think that it also should be as a relatively specific radiologic feature for ICAH. The underlying pathophysiologic mechanisms of this sign are not clear, but for ICAH, it may be related to embryonic development and the difference of anatomical structures between the carotid sinus and the distal ICA.

Furthermore, the imaging data in this series revealed 2 courses of hypoplastic ICAs: either continuing intracranially or becoming occluded in the distal ophthalmic segment, no other transitional pattern. Consequently, ICAH patients were categorized as the distal occlusion type and the nondistal occlusion type. In terms of embryology, the carotid artery develops from different embryological arterial segment branches and abnormalities in any branch may lead to structural changes in the ICA.[21] Thus, we speculate that patients in the distal occlusion type might suffer from developmental arrest of embryonic branches forming the terminal segment of ICA due to either endogenous or exogenous factors.

Theoretically, hypoperfusion should be inevitable in the ipsilateral AC for patients with ICAH, but most patients have no symptoms due to effective collateral flows.[7,10] Lie[22] reported 6 typical pathways of collateral circulation in association with ICA absence. Nonetheless, ICAH was only mentioned as follows: "Type E: Bilateral hypoplastic ICAs supplying ACAs, and the MCAs are supplied by enlarged PcomAs." In the present study, we found that each patient had a certain degree of collateral circulation, and the frequency of collateral supply via the circle of Willis in the distal occlusion type was higher than that in the nondistal occlusion type. The circle of Willis is formed when the embryo size is 7-24 mm. If the blood flow from ICA is blocked before complete development of the circle of Willis, anastomotic channels develop mainly through primitive embryonal

anastomoses such as supracavernous anastomosis, intercavernous anastomosis, or supraclinoid-supraclinoid anastomosis.[11-14] Conversely, if the blood flow is blocked after the formation of the circle of Willis, collateral circulation develops mainly from the circle of Willis.[23,24] In this study, the collateral circulation in the distal occlusion type was established via the PcomA from the circle of Willis. Thus, we inferred that these patients might experience abnormal blood supply of the ICA after the formation of the circle of Willis. Furthermore, the reason for opening the PcomA rather than the anterior communicating artery may be partially associated with the timing of the development of the anterior communicating artery and PcomA during embryonic growth.[25]

Interestingly, this study also found that patients in the nondistal occlusion type showed multiform collateral channels, and overlapping occurred at all levels of collateral circulations. Such phenomena might be due to both congenital and acquired factors. Moreover, secondary and tertiary collateral circulation patterns were frequently observed, which might be attributed to the partial blood supply from the ipsilateral ICA in the nondistal occlusion type compared with the distal occlusion type. Although this collateral blood supply may be sufficient during the embryonic stage, brain development and oxygen consumption increase occur after birth, and the carotid blood supply becomes insufficient and other collateral circulation pathways are needed to establish an adequate blood supply.[23]

ICAH is often associated with dilated abnormalities of other cerebral vessels.[6-9] Accordingly, the present study showed a significantly different frequency of arterial dilated lesions between the 2 types after the imaging classification, which might be associated with different collateral circulation patterns. Compared with the nondistal occlusion type, the blood supply of the ipsilateral AC was primarily from the posterior circulation via the PcomA. This high-flow collateral pathway inevitably affects the hemodynamics of the posterior circulation, thereby leading to abnormal enlargement or tortuosity of the vertebrobasilar system, and causing aneurysmal formation as well. Dilated lesions of the PcomA are the most common abnormalities. The PcomA is a crucial structure for bridging the AC and posterior circulation. Nevertheless, the PcomA varies in diameter from the ICA to the PCA. Thus, the passage of blood through the PcomA gives rise to a pressure gradient. After the ipsilateral ICA is occluded, the local blood flow and flow velocity of the PcomA increase significantly, and the stress placed on the PcomA increases dramatically. Vascular wall elasticity decreases continuously with an increase in stress, leading to enlargement of the PcomA.[7,8] Other studies suggested that the malformed remodeling of congenital embryonic vessels might also play a role in arterial dilation.[11,13,21]

The present study found that 4 patients with bilateral ICAH also displayed dilated bilateral vertebral arteries, in addition to the dilated lesions of the PcomA. Since their collateral compensatory supply in bilateral ACs is mainly supplied by the posterior circulation, only opening or enlarging the PcomA alone may be insufficient for improving the anterior blood supply. The lumen of the bilateral vertebral arteries undergoes gradual remodeling under the impact of blood flow, which leads to adequate compensation by the dilatation of the vertebral artery lumen.

Intriguingly, in addition to dilated lesions, ICAH is also accompanied by other anomalies of cerebral vessels such as hypoplasia or dysplasia of ACA, MCA, or vertebrobasilar artery,[4,6,26,27] primitive trigeminal artery,[27] "rete mirabile" aneurysm,[28] and abnormality or deformity of the aorta and aortic arch.[15,27] In this study, 50% of patients presented with other abnormal cerebrovascular structures, and hypoplasia was frequently observed.

In the early phase of ICAH, most patients are asymptomatic or present only nonspecific symptoms, and ICAH is often detected only incidentally.[7,11,29] Only a few patients show definite symptoms that are caused by aneurysmal SAH or compression of adjacent structures by abnormally enlarged vessels.[8,11,30] Furthermore, only a few studies have reported the natural history of ICAH and its relationship with ischemic stroke has yet to be elucidated.[4,31] In this study, 25% of patients developed endpoint cerebrovascular events during follow-up and patients from different types experienced different events. For 3 patients displayed ICAH of the nondistal occlusion type and suffered from ischemic cerebrovascular events, no collateral circulation via the circle of Willis was found. Previous studies have suggested a significant correlation between an incomplete circle of Willis and ischemic stroke.[32,33] Those 3 patients showed cerebral blood flow perfusion defects in the ipsilateral AC. Thus, the etiology of ischemic events was thought to be due to the insufficient collateral circulation resulting in distal hypoperfusion of the ipsilateral AC.

No ischemic events were encountered during follow-up in the distal occlusion type, which might be associated with a relatively sufficient blood supply from the PcomA. In addition, expansion of arteries from the posterior circulation was observed in some patients, which further improved the perfusion of the AC. Nevertheless, these collateral circulations caused hemodynamic changes, which led to tortuosity and dilation of collateral arteries, thereby resulting in aneurysms.[8,10] In this study, 2 patients with SAH suffered from hemodynamics-related rupture of the PcomA aneurysm.

The present study has limitations. The number of patients is small owing to the rarity of ICAH, and we only divide into 2 types according to the characteristics of our own cases. We do not rule out that as the gradual expansion of number of cases collected, there may be other types or more accurate classification or update one based on our classification. Secondly, no treatment data are available. Finally, much study is still necessary to determine the natural history of ICAH, but this might be a difficult point to examine since ICAH development occurs during fetal development.

Our study suggested a novel classification of ICAH based on the occlusion of the distal ophthalmic segment (distal occlusion type and nondistal occlusion type). The 2 types may show different collateral circulation patterns, coexisting cerebrovascular abnormalities and potential clinical outcomes, which requires to be confirmed.

Declaration of Competing Interest

All authors declare that they have no competing interests.

References

1. Luk YS, Man EM, Sy AN. Bilateral hypoplasia of the internal carotid arteries. Singapore Med J 2010;51:e163-e165.
2. Nicoletti G, Sanguigni S, Bruno F, et al. Hypoplasia of the internal carotid artery: collateral circulation and ultrasonographic findings. A case report. J Ultrasound 2009;12:41-44.
3. Chen CJ, Chen ST, Hsieh FY, et al. Hypoplasia of the internal carotid artery with intercavernous anastomosis. Neuroradiology 1998;40:252-254.
4. Nardone R, Venturi A, Ausserer H, et al. Transient ischaemic attacks in two cases of internal carotid artery hypoplasia. Neurol Sci 2005;26:282-284.
5. Ide C, De Coene B, Mailleux P, et al. Hypoplasia of the internal carotid artery: a noninvasive diagnosis. Eur Radiol 2000;10:1865-1870.
6. Mujagic S, Kozic D, Huseinagic H, et al. Symmetry, asymmetry and hypoplasia of the intracranial internal carotid artery on magnetic resonance angiography. Acta Med Acad 2016;45:1-9.
7. Bhat DI, Somanna S, Kovoor JM. Bilateral hypoplasia of the internal carotid artery. Indian J Radiol Imaging 2011;21:257-260.
8. Zink WE, Komotar RJ, Meyers PM. Internal carotid aplasia/hypoplasia and intracranial saccular aneurysms: series of three new cases and systematic review of the literature. J Neuroimaging 2007;17:141-147.
9. Tasar M, Yetiser S, Tasar A, et al. Congenital absence or hypoplasia of the carotid artery: radioclinical issues. Am J Otolaryngol 2004;25:339-349.
10. Siddiqui AA, Sobani ZA. Bilateral hypoplasia of the internal carotid artery, presenting as a subarachnoid hemorrhage secondary to intracranial aneurysmal formation: a case report. J Med Case Rep 2012;6:45.
11. Erdem Y, Yilmaz A, Ergun E, et al. Bilateral internal carotid artery hypoplasia and multiple posterior circulation aneurysms. Importance of 3DCTA for the diagnosis. Turk Neurosurg 2009;19:168-171.
12. Hong JH, Chang HW, Kim JM, et al. Internal carotid artery agenesis associated with a rare collateral pathway: supraclinoid-supraclinoid anastomosis. A case report. Surg Radiol Anat 2016;38:261-263.
13. Kumaresh A, Vasanthraj PK, Chandrasekharan A. Unilateral agenesis of internal carotid artery with intercavernous anastomosis: a rare case report. J Clin Imaging Sci 2015;5:7.
14. Sinha R, Gupta R, Abbey P, et al. Carotid agenesis with intercavernous anastomosis. Turk Neurosurg 2012;22:371-373.
15. Okawa M, Higashi T, Komiyama M, et al. Left internal carotid artery agenesis with trans-sellar collateral and a right aortic arch: case report. Interv Neuroradiol 2015;21:759-764.
16. Lath N, Taneja M. Bilateral congenital hypoplasia of the internal carotid arteries. J HK Coll Radiol 2008;11:129-131.
17. Briganti F, Maiuri F, Tortora F, et al. Bilateral hypoplasia of the internal carotid arteries with basilar aneurysm. Neuroradiology 2004;46:838-841.
18. Yasaka M, Ogata T, Yasumori K, et al. Bottle neck sign of the proximal portion of the internal carotid artery in moyamoya disease. J Ultrasound Med 2006;25:1547-1552.
19. Yasuda C, Arakawa S, Shimogawa T, et al. Clinical significance of the champagne bottle neck sign in the extracranial carotid arteries of patients with Moyamoya disease. AJNR Am J Neuroradiol 2016;37:1898-1902.
20. Shimogawa T, Morioka T, Sayama T, et al. Champagne bottle neck sign in a patient with Moyamoya syndrome. World J Clin Cases 2014;2:474-477.
21. Lasjaunias P, Santoyo-Vazquez A. Segmental agenesis of the internal carotid artery: angiographic aspects with embryological discussion. Anat Clin 1984;6:133-141.
22. Lie TA. Congenital anomalies of the carotid arteries 1968.
23. Cali RL, Berg R, Rama K. Bilateral internal carotid artery agenesis: a case study and review of the literature. Surgery 1993;113:227-233.
24. Midkiff RB, Boykin MW, McFarland DR, et al. Agenesis of the internal carotid artery with intercavernous anastomosis. AJNR Am J Neuroradiol 1995;16:1356-1359.
25. Menshawi K, Mohr JP, Gutierrez J. A functional perspective on the embryology and anatomy of the cerebral blood supply. J Stroke 2015;17:144-158.
26. Kim DS, Kang SG, Yoo DS, et al. Ruptured right posterior inferior cerebellar artery aneurysm associated with hypoplasia of the right vertebral artery and both internal carotid arteries. Br J Neurosurg 2009;23:551-553.
27. Jaeger HJ, Mehring UM, Gissler HM, et al. Congenital absence of the internal carotid artery and the basilar artery with persistent trigeminal artery associated with coarctation of the aorta. Eur Radiol 2000;10:1805-1809.
28. Aburto-Murrieta Y, Dulce BD. Asymptomatic carotid rete mirabile and contralateral carotid agenesis: a case report. Vasc Endovascular Surg 2011;45:361-364.
29. Akfirat M, Cihangiroglu M, Ozdemir H, et al. Internal carotid artery hypoplasia in two cases. Clin Radiol 2002;57:951-954.
30. Kikuchi K, Kowada M, Kojima H. Hypoplasia of the internal carotid artery associated with spasmodic torticollis: the possible role of altered vertebrobasilar haemodynamics. Neuroradiology 1995;37:362-364.
31. Sfaihi L, Boukedi A, Aloulou H, et al. [Hypoplasia of the internal carotid artery: a rare cause of cerebral ischemic stroke in children]. Arch Pediatr 2010;17:1657-1660.
32. Hoksbergen AW, Legemate DA, Csiba L, et al. Absent collateral function of the circle of Willis as risk factor for ischemic stroke. Cerebrovasc Dis 2003;16:191-198.
33. Zhou H, Sun J, Ji X, et al. Correlation between the integrity of the circle of Willis and the severity of initial noncardiac cerebral infarction and clinical prognosis. Medicine 2016;95:e2892.