Clinical characteristics of internal carotid artery aplasia

Jian-Jun Tang, Yong-Bo Zhao, You-Yu Lu, Guo-Dong Wang, Qing Zhang, Xiao-Lin Zhou, Xiang Shen, Xiao-Yan Song, Hai-Yan Lv, Qiao-Shu Wang, Yun-Cheng Wu

Department of Neurology, Shanghai General Hospital, Shanghai Jiao Tong University School of Medicine, No.100, Haining Road, Shanghai 200080, P.R. China

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Abstract: The rarity of internal carotid artery aplasia (ICA) limits the research of this condition. As the endovascular treatment of cerebrovascular diseases is applied more and more widely, further understanding of ICA aplasia is important. We reviewed 4 consecutive internal carotid artery (ICA) aplasia patients admitted in our department between June 1, 2015 and December 31, 2016. The presentations were: ipsilateral hemisphere ischemic symptoms in 2 patients, headache in 1 patient, unfixed numbness in 1 patient and ischemic stroke in the contralateral middle cerebral artery (MCA) territory in 1 patient. Collateral channels to the ipsilateral hemisphere include the ophthalmic artery from the middle meningeal artery (MMA), the muscle branch from the ipsilateral vertebral artery (VA), anterior communicating arteries (ACOM), posterior communicating arteries (PCOM) and leptomeningeal arteries. Associated anomalies were: varied origins of common carotid artery (CCA) or VA, ipsilateral CCA hypoplasia, agenesis or aplasia of bone carotid canals, hypoplasia of posterior cranial fossa, the extremely hypertrophic VA. MCA or anterior cerebral artery (ACA) aplasia was confirmed in 2 of 4 cases. Fusiform aneurysms were found in 2 cases. Distinguishing ICA aplasia, associated anomalies and collateral channels are important for preparing the endovascular treatment and surgery of head and neck.

Keywords: Internal carotid artery, aplasia, cerebrovascular disease, collateral channel, angiography

Introduction

Internal carotid artery (ICA) aplasia, alternatively used with ICA hypoplasia or ICA agenesis, refers to the failure development of ICA. The rarity of this entity limited our understanding of this condition. Lie defined ICA agenesis as complete failure in ICA development, ICA aplasia as lack of development of ICA with its precursor once existed, and hypoplasia as incomplete development of the organ [1]. Case reports suggested intracranial arteriovenous malformations (AVMs), intracranial aneurysms as well as several types of collateral channels were associated with ICA aplasia [2-9]. We analyzed the medical records of 4 cases of ICA aplasia in order to prompt the understanding of this condition and improve the safety of endovascular treatment of cerebrovascular disease as well as head and neck surgery.

Materials and methods

Medical records of 4 cases of ICA aplasia consecutively admitted into our department between June 1, 2015 and December 31, 2016 were reviewed and analyzed. This study was approved by the Institutional Review Board of our hospital and informed consents were obtained from all the research subjects.

Results

There were 2 males and 2 females summarized (Table 1). The age ranged from 30 to 53 years with the mean age of 43 years. Among the 4 cases, there was 1 case of ICA agenesis, 1 case of ICA aplasia and 2 cases of ICA hypoplasia according to Lie’s theory [1]. Case 1 (Figure 1) complained of mild dizziness. Then computed tomography angiography (CTA) confirmed the absence of the whole left ICA. Case 2 (Figure 2) was admitted for ischemic stroke then the CTA occasionally found the hypoplastic ICA contralateral to the stroke foci, which was confirmed with angiography. Case 3 (Figure 3) was admitted for recurrent serious left headache and right weakness. Then digital subtraction angiography (DSA) revealed the left ICA aplasia. Case 4 (Figure 4) complained of unfixed nu-
### Table 1. Clinical characteristics of 4 cases of ICA aplasia

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td>Male</td>
<td>Female</td>
<td>Female</td>
<td>Male</td>
</tr>
<tr>
<td><strong>Age</strong></td>
<td>52</td>
<td>30</td>
<td>53</td>
<td>37</td>
</tr>
<tr>
<td><strong>ICA status</strong></td>
<td>Lt agenesis</td>
<td>Rt hypoplasia</td>
<td>Lt aplasia</td>
<td>Lt hypoplasia</td>
</tr>
<tr>
<td><strong>Chief complain</strong></td>
<td>Dizzy</td>
<td>Rt side weakness, seizure</td>
<td>Rt side weakness, slurred speaking, Lt side headache</td>
<td>Unfixed numbness</td>
</tr>
<tr>
<td><strong>Aortic arch</strong></td>
<td>Normal</td>
<td>Lt CCA origins from brachiocephalic trunk</td>
<td>Normal</td>
<td>Lt VA origins from aortic arch</td>
</tr>
<tr>
<td><strong>Ipsilateral CCA</strong></td>
<td>Hypoplasia</td>
<td>Normal</td>
<td>Hypoplasia</td>
<td>Hypoplasia</td>
</tr>
<tr>
<td><strong>Bone carotid canal</strong></td>
<td>Ipsilateral agenesis, contralateral intact</td>
<td>Bilateral intact</td>
<td>Ipsilateral hypoplasia, contralateral incomplete bone wall</td>
<td>Bilateral intact</td>
</tr>
<tr>
<td><strong>Collateral channel</strong></td>
<td>ACOM, PCOM</td>
<td>ACOM, PCOM</td>
<td>Ophthalmic artery from MMA, leptomeningeal artery from Lt PCA, internal carotid artery remnant from vertebral artery branch</td>
<td>ACOM, leptomeningeal artery from Lt ACA and Lt PCA</td>
</tr>
<tr>
<td><strong>Perfusion</strong></td>
<td>Mild delayed MMT and TTP</td>
<td>NA</td>
<td>NA</td>
<td>Mild delayed MMT and TTP</td>
</tr>
<tr>
<td><strong>Aneurysm</strong></td>
<td>No</td>
<td>No</td>
<td>Ipsilateral MCA</td>
<td>Ipsilateral ACA</td>
</tr>
<tr>
<td><strong>Other associated anomaly</strong></td>
<td>No</td>
<td>No</td>
<td>Hypertrophic VA, hypoplastic posterior cranial fossa and Lt cerebellum</td>
<td>Hypoplastic ipsilateral MCA, hypertrophic ipsilateral ACA and PCA</td>
</tr>
<tr>
<td><strong>Atherosclerosis</strong></td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td><strong>Ipsilateral stroke/TIA</strong></td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Contralateral stroke</strong></td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td><strong>Risk factors of stroke</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tobacco use</td>
<td>No</td>
<td>Yes</td>
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<tr>
<td>Alcohol degestion</td>
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<tr>
<td>Hypertension</td>
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<td>Hyperlipidemia</td>
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<td>No</td>
<td>No</td>
<td>No</td>
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<tr>
<td>Diabetes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
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<tr>
<td>Hyperhomocysteinemia</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td><strong>Follow up duration</strong></td>
<td>18 months</td>
<td>8 months</td>
<td>6 months</td>
<td>6 months</td>
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<tr>
<td><strong>Outcome</strong></td>
<td>Stable</td>
<td>Stable</td>
<td>Stable</td>
<td>Stable</td>
</tr>
</tbody>
</table>

**Notes:** Lt: left; Rt: right; ICA: internal carotid artery; CCA: common carotid artery; VA: vertebral artery; ACOM: anterior communicating artery; PCOM: posterior communicating artery; MMA: middle meningeal artery; ACA: anterior cerebral artery; PCA: posterior cerebral artery; MTT: mean transit time; TTP: time to peak; NA: not available. TIA: transient ischemic attack.
mbness and magnetic resonance angiograph (MRA) revealed a fainted ICA, and the ICA hypoplasia was confirmed with DSA. In the 4 cases, lacunar infarction foci were found in 1 territory of 4 hypoplastic ICAs, in which case the MCA distal to the affected ICA was also aplastic. During the follow-up, no stroke happened in all 4 cases.

Collateral channels

In these 4 cases, collateral channels through anterior communicating arteries (ACOMs) were found in 3 cases (Figures 1, 2 and 4). Collateral channels through posterior communicating arteries (PCOMs) were found in 2 cases (Figures 1 and 2). An ophthalmic artery from the MMA...
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(middle meningeal artery) as a collateral channel was confirmed in 1 case (Figure 3). Leptomeningeal arteries as collateral channels were documented in 2 cases (Figures 3 and 4). A rare collateral channel originating from the muscular branch of vertebral artery (VA) to the remnant of ICA was confirmed in 1 case with angiography (Figure 3). CT perfusion (CTP) completed in 2 cases and revealed almost normal cerebral blood flow (CBF) and cerebral blood volume (CBV) but slightly delayed mean transit time (MTT) and time to peak (TTP).

Carotid bone canals

In the patient with ICA agenesis, ipsilateral carotid bone canal (CBC) agenesis was confirmed with skull base CT (SBCT) scan. In the patient with ICA aplasia, ipsilateral CBC hypoplasia and incompleteness of the contralateral CBC were confirmed with SBCT. In the 2 cases of ICA hypoplasia, the ipsilateral CBCs were both intact.

Associated abnormalities

Fusiform intracranial aneurysms were seen in 2 patients. No saccular aneurysms were found. Aortic variations were seen in 2 cases. Posterior cranial fossa hypoplasia was found in one patient with ICA aplasia. MCA hypoplasia and ACA hypoplasia were respectively confirmed in two patients. An extremely hypertrophic VA was confirmed in one patient with ICA aplasia.

Discussion

The incidence of ICA aplasia is less than 0.01% with the ratio between right:left:both being 1:3:1 [10]. Tode discovered carotid agenesis on post-mortem examination in 1787 [11]. ICA aplasia can be associated with congenital combined pituitary hormone deficiency [12], neurofibromatosis [13, 14], intracranial aneurysms [2]. The mechanism of ICA aplasia is still to be determined. ICA develops at 4- to 5-mm embryonic stage and completes by 6 weeks of fetal life [15, 16]. The basilar artery is formed at the 7-12 mm stage of embryonic development and the circle of Willis is complete when the anterior communicating artery is formed at the 24 mm stage [17]. The skull base begins to form since the fifth or sixth week of fetal life [18]. Lie illustrated six types of collateral channels in ICA aplasia. Type A - Unilateral ICA aplasia with collateral circulation to the ipsilateral ACA through an ACOM and the collateral channel to the ipsilateral MCA through a PCOM, Type B - Unilateral ICA absence with the collateral channel to the ipsilateral hemisphere through an ACOM, Type C - Bilateral ICA agenesis with collateral channels to anterior circulation.
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Figure 3. Case 3: A 53 years old female patient with recurrent left headache, intermittent right side weakness and slurred speech. A. Frontal projection of angiography showed the small caliber of the right CCA and the enlarged left VA. B. Lateral projection of left CCA angiography revealed one collateral channel through the ophthalmic artery from the MMA (hollow arrow). C. Lateral projection of left VA angiography revealed the remnant of ICA supported with a muscular branch of VA (black triangles) and a fusiform aneurysm of left MCA (star). D. CTA showed the absence of A1 segment of right ACA and a fusiform aneurysm of left MCA (star). E. CT of scull base revealed ipsilateral CBC aplasia, incomplemetn of bone wall (white arrow) of the contralateral CBC and hypoplasia of posterior fossa and left cerebellum (curved white arrow).

through carotid-vertebrobasilar anastomoses and PCOMs, Type D - Unilateral cervical portion ICA agenesis with collateral flow through intercavernous communication from the contralateral ICA, Type E - Bilateral ICA hypoplasia with collateral channels through bilateral PCOMs, Type F - ICA hypoplasia with collateral flow through rete mirabile [1, 7]. Associated collateral channels may reflect the time and nature of ICA occlusion. If ICA absence happens before embryonic arteries regression, the embryonic vessels will become the collateral channels. On the contrary, if the ICA regresses after the regression of embryonic arteries, they can’t be the collateral channel [19]. Supposed mechanism of unilateral ICA aplasia may be due to the mechanical stress such as exaggerated embryo folding and amniotic bands constriction [20]. In our case 3, the patient had a remnant of distal ICA while the initial segment was absent. Additionally, hypoplasia of the patient’s posterior fossa and bilateral CBCs was documented. These appearances were consistent with the mechanical pressure theory.

In case 4, the collateral channels mainly through leptomeningeal arteries suggested the blockage of ICA happened at a late embryonic stage. The coincidence of ICA hypoplasia and MCA hypoplasia on 1 patient is difficult to be explained with mechanical pressure. We proposed that the mechanism of thromboembolism at the terminal of the developed ICA at the fetal stage should be taken into account. CBCs develop depending upon the presence of ICAs at 5-6 weeks gestation. Absence of CBC indi-
cates ICA agenesis [21]. If ICAs occlude after the skull base development, the CBCs will be normal. Otherwise the CBCs will be hypoplastic or absent. 2 of our 4 cases have normal CBCs, which suggests that their ICAs were once fully developed. Whether the reduced blood flow may induce the atrophy of ICA is to be determined.

In our case 3, we identified collateral channels through the remnant of ICA from the muscular branch of VA and the ophthalmic artery from the MMA which was documented in some case reports of intracranial aneurysms [22]. Two patients received CTP scan and the results suggested normal CBV and CBF, while MTT and TTP slightly delayed. Since ICA aplasia happened at the early stage of life, compensatory blood flow can be established well and special treatment is not necessary for ICA aplasia. However, the collateral channels may be more vulnerable as ageing and non-specific neurologic symptoms such as dizzy, headache or weakness may appear after middle age.

Figure 4. Case 4: A 37 years old female patient with unfixd numbness. A. Aortic arch angiography showed the left VA originated from the arch and small caliber of left CCA. B. Left CCA angiography showed the hypoplastic ICA as well as the hypoplastic left MCA (long black arrow). C. MR revealed 2 lacunar infarction fuci in the ipsilateral hemisphere. D. CTP suggested slightly delayed TTP and MTT with CBF and CBV normal. E. A fusiform aneurysm was found at the ipsilateral ACA (curved black arrow).
Prevalence of aneurysms reported in ICA aplasia patients was 24-67%, which is significantly higher than in general population, 2-4% [2, 21, 23]. Paschoal et al. reviewed patients of ICA hypoplasia and concomitant rete mirabile, and found the initial symptom was most frequently subarachnoid hemorrhage (SAH) (34.2%) [6]. Among the Lee et al. reported series of ICA aplasia, 3 out of 9 patients presented with SAH [2]. In our 4 cases, 2 unruptured fusiform aneurysms were found in 2 cases. No saccular aneurysms were found. The aneurysms seem to be related to increased blood flow and changed hemodynamics. No SAH was presented.

Similar to the cases reported before, in 3 of our 4 cases, calibers of ipsilateral CCAs were significantly smaller than the contralateral ones. The diameter difference between bilateral CCAs may be used as an indication of ICA aplasia in future [3], which worth further research. Some cases of hypoplastic ICA should be differentiated from MOYAMOYA disease, which presented as stenosis or occlusion at the terminal of ICA with abnormal vascular network nearby [24]. It is important to distinguish the ICA aplasia from ICA acute occlusion when the guide wire goes through the blockage to treat ischemic stroke. In preparing for head and neck surgery, documenting the ICA aplasia and related collateral channels is of great significance. Since this is a small scale retrospective study, the mechanism of ICA aplasia and the risk of associated anomalies including intracranial artery aplasia and scull base hypoplasia need further research.

Patients with ICA aplasia may have risks of hypoplasia of intracranial arteries, scull bases and cerebellums as well as intracranial aneurysms. In addition to mechanical oppression, ICA system thromboembolism in the early life stage should be taken into account when the mechanism is studied. It’s important to detect the collateral channels and accompanied anomalies for surgery or endovascular therapy of head and neck in these patients.

Disclosure of conflict of interest

None.

Address correspondence to: Drs. Yun-Cheng Wu and Qiao-Shu Wang, Department of Neurology, Shanghai General Hospital, Shanghai Jiao Tong University School of Medicine, No.100, Haining Road, Shanghai 200080, P.R. China. Tel: 86-21-37798584; Fax: 86-21-63240825; E-mail:yunchw@medmail.com.cn (YCW); qiaoshuwang@gmail.com (QSW)

References


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