Acta Neurochir (2003) 145: 117–125
DOI 10.1007/s00701-002-1046-y

Acta Neurochirurgica
Printed in Austria

Clinical Article

Aplasia of the internal carotid artery

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Published online February 10, 2003

Summary

Background. The majority of previous reports on this rare agenesis of the internal carotid artery (ICA) have been limited to reporting upon its association with other congenital anomalies case by case. In order to collectively summarize this congenital anomaly of ICA, we have reviewed nine cases of ICA aplasia and their associated abnormalities.

Method. Nine cases of ICA aplasia were reviewed. 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 computed tomography (TBCT). Their presumable embryological aetiologies, initial presenting symptoms, unusual collateral circulations, as demonstrated by angiographies, and various associated anomalies are reviewed.

Findings. The initial presentations were; subarachnoid haemorrhage in three patients, headache in one patient and ischemic symptoms and signs in three patients. The remaining two cases were found incidentally during angiography for other diseases. Collateral circulations to the middle cerebral artery ipsilateral to the ICA aplasia were via posterior communicating artery (P-com) or anterior communicating artery (A-com). On TBCT, all cases but one demonstrated agenesis of the bony carotid canal and the remaining case showed a hypoplastic canal. Cerebral aneurysms were found in six patients, four with A-com aneurysm, one with a basilar bifurcation aneurysm, and one with both a right P-com and a left cavernous ICA aneurysm; two incidentally found cases had no aneurysm. Other associated abnormalities were found in four cases; one case of hypoplasia of the common carotid artery (CCA) with an arachnoid cyst at the temporal pole, one case of abnormal origin of the right CCA from the aorta and the right subclavian artery from the descending aorta, one case of congenital temporomandibular joint (TMJ) ankylosis, and one case of nasopharyngeal angiofibroma with atresia of the upper basilar artery. Except for the atresia of the upper basilar artery, all such abnormalities were found on the same side as the ICA aplasia.

Interpretation. Agenesis or aplasia of ICA may be entirely harmless. However, associated conditions such as cerebral aneurysm or abnormal collateral channels should alert clinicians to the possibility of deterioration to life-threatening conditions, such as subarachnoid haemorrhage or irreversible ischemia. Other associated anomalies are commonly depicted on the same side as the ICA aplasia and may also give rise to issues of clinical importance.

Keywords: Aplasia; aneurysm; carotid canal; internal carotid artery.

Introduction

Cerebrovascular insufficiency is a clinical condition due in most cases to acquired diseases, such as atherosclerosis. Aplasia, hypoplasia and agenesis of the ICA are rare congenital anomalies, with less than 100 cases reported worldwide [12] since Tode first described a case with absence of the ICA in 1787 [38] and Verbiest demonstrated the condition by angiography for the first time in 1954 [41].

The definitions of agenesis, aplasia, and hypoplasia of the internal carotid artery seem to be unclear and are often used interchangeably, but Lie made an effort to distinguish between the term agenesis of the internal carotid artery and aplasia. He referred to '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 [21].

Patients with narrow or absent ICA, which we believe to have resulted from developmental hypoplasia, rarely present with cerebrovascular insufficiency, because of their rarity and the frequent development of abnormal collateral channels, which has caused some hesitancy in the diagnosis of this anomaly [27].

Most of the previous reports on this abnormality of the ICA have been limited to descriptions of its association with various other congenital anomalies case by case. In this report, we collectively reviewed nine cases of ICA aplasia with their presumably embryological aetiologies, and describe the initial presenting symptoms and signs (including the presence of ischemic symptoms), unusual collateral circulations (as demonstrated by angiography) and various associated anomalies. The knowledge of this rare pathology and its frequently associated conditions would prompt further clinical evaluation to rule out potentially life-threatening conditions, such as cerebral aneurysms, and eventually facilitate management of the case.

Material and methods

Subject

The medical records and radiological studies of nine patients with agenesis, aplasia or hypoplasia of ICA, registered between 1988 and 2000, were reviewed retrospectively. There were five male and four female patients, the age ranged from 15 to 57 years with a mean age of 37.5 years.

Radiologic studies

In all nine patients of our series, angiography was performed for causes other than a suspected ICA aplasia. Once the aplasia or agenesis of the ICA was detected by angiography, TBCT and brain magnetic resonance image (MRI) were routinely performed. The diagnosis of aplasia or agenesis of ICA was based on the angiographic findings and the absence or a hypoplastic bony carotid canal on TBCT. The possibility of an association between the ICA aplasia and cerebral aneurysm or abnormal collateral channels feeding the ipsilateral cerebral hemisphere was also investigated. MRI was routinely performed on each patient to examine the possibility of cerebral hemiatrophy or an associated ischemic or infarct lesion, regardless of the presentation of ischemic symptoms and signs.

Treatment

No surgical or interventional correction was attempted for ICA aplasia in any of the cases in our series. For associated cerebral aneurysms, direct aneurysmal neck clipping by the pterional approach was performed in all four patients with A-com aneurysm associated with ICA aplasia. In one patient with a basilar bifurcation aneurysm, the aneurysmal sac was embolized with a Guglielmi detachable coil (GDC). In the remaining patient with a right P-com and a left cavernous ICA aneurysm, no further therapy was performed because of a faint blood supply to right MCA territory through the right P-com and the location of aneurysm in the left cavernous sinus. Surgical correction by craniofacial reconstruction was performed for a TMJ ankylosis patient and conventional radiotherapy was given for a patient with nasopharyngeal angiofibroma. An arachnoid cyst of the left temporal lobe in a patient with left ICA aplasia combined with hypoplasia of the CCA was left for observation because of its small size and the lack of neurological deficit.

Results

The results of radiological studies are summarized in Table 1.

Table 1. *Summary of presenting symptoms and radiological studies*

	Age/sex	Presenting Sx	ICA	CCA	Collateral to MCA	Carotid canal	Associated aneurysm	Other associated anomalies
Case 1	55/M	SAH	Rt agenesis	intact	P-Com	agenesis	A-Com	–
Case 2	19/F	SAH	Lt agenesis	hypoplasia	P-Com	agenesis	A-Com	Lt temporal arachnoid cyst
Case 3	51/M	SAH	Rt agenesis	intact	P-Com	agenesis	A-Com	–
Case 4	57/F	headache & general weakness	Lt agenesis	intact	P-Com & A-Com	agenesis	A-Com	–
Case 5	50/F	loss of vision, Rt (amaurosis fugax)[a]	Rt agenesis	intact	A-Com	agenesis	P-Com Lt cavernous sinus	Rt CCA from aorta Rt subclavian artery from descending aorta
Case 6	22/F	incidentally found	Rt agenesis	intact	P-Com & A-Com	agenesis	–	Rt congenital TMJ ankylosis
Case 7	15/M	incidentally found	Rt agenesis	intact	A-Com	agenesis	–	Rt nasopharyngeal angiofibroma basilar artery tip atresia
Case 8	31/M	headache	both hypoplasia	intact	P-Com	hypoplasia	basilar bifurcation	–
Case 9	34/M	recurrent TIA[b]	Lt agenesis	intact	P-Com	agenesis	–	–

Rt Right, *Lt* left, *ICA* internal carotid artery, *MCA* middle cerebral artery, *CCA* common carotid artery, *SAH* subarachnoid haemorrhage, *A-com* anterior communicating artery, *P-com* posterior communicating artery, *TIA* transient ischemic attack, *TMJ* temporomandibular joint.

[a] Right thalamic infarction in MRI.

[b] Left frontal lobe infarction in MRI.

Presenting symptoms

Three patients presented with severe headache and altered mentality due to subarachnoid haemorrhage, and three patients presented with ischemic symptoms; one with headache and general weakness, another with amaurosis fugax, and the third with repeated attack of transient ischemia (TIA). The patient with amaurosis fugax and with TIA revealed an infarct in the right thalamus (Fig. 1) and left frontal lobe on MRI, respectively. The remaining three patients were admitted for other causes; one patient with repeated history of headache, one patient with right congenital TMJ ankylosis, who also was found to have an absence of carotid canal on TBCT, and one patient with a recurrent history of epistaxis who showed highly vascular mass in the nasopharynx on MRI (Fig. 3).

Lateralization

Five patients presented with right side ICA agenesis, while three patients presented with left side agenesis. The remaining patient showed both ICA hypoplasia to ophthalmic artery level and then distal agenesis (Fig. 2). Hypoplasia of the ipsilateral common carotid

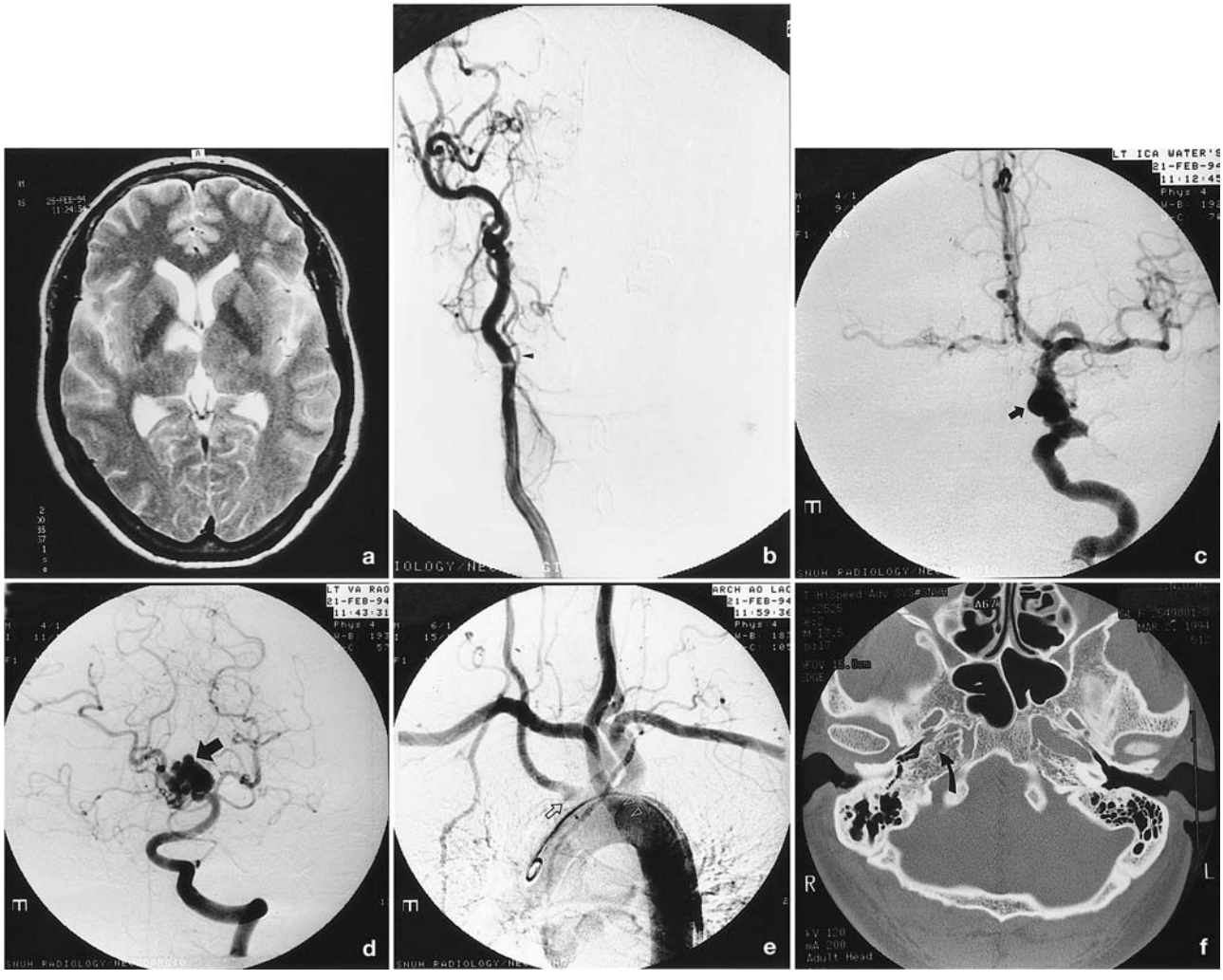

Fig. 1. A 50 years old female patient with the symptom of amaurosis fugax (case 5) shows a small infarct in the right thalamus on the T2-weighted axial view of brain MRI (a). Angiography of the right common carotid artery shows faint staining of internal carotid artery (arrowhead) (b). Selection of the left internal carotid artery shows a saccular aneurysm (small arrow) in a cavernous portion of ICA and a collateral channel to the right MCA territory through the left A-com (c). Vertebral angiography reveals a huge fusiform aneurysm (large arrow) in the P1 to P2 portions of the right posterior cerebral artery (d). Selection from the aortic arch showing the anomalous origin of the right common carotid artery from the aortic arch (open arrow) and the right subclavian artery from the descending aorta (open arrowhead) (e). The TBCT shows absence of the petrous carotid canal on the right side (curved arrow) (f)

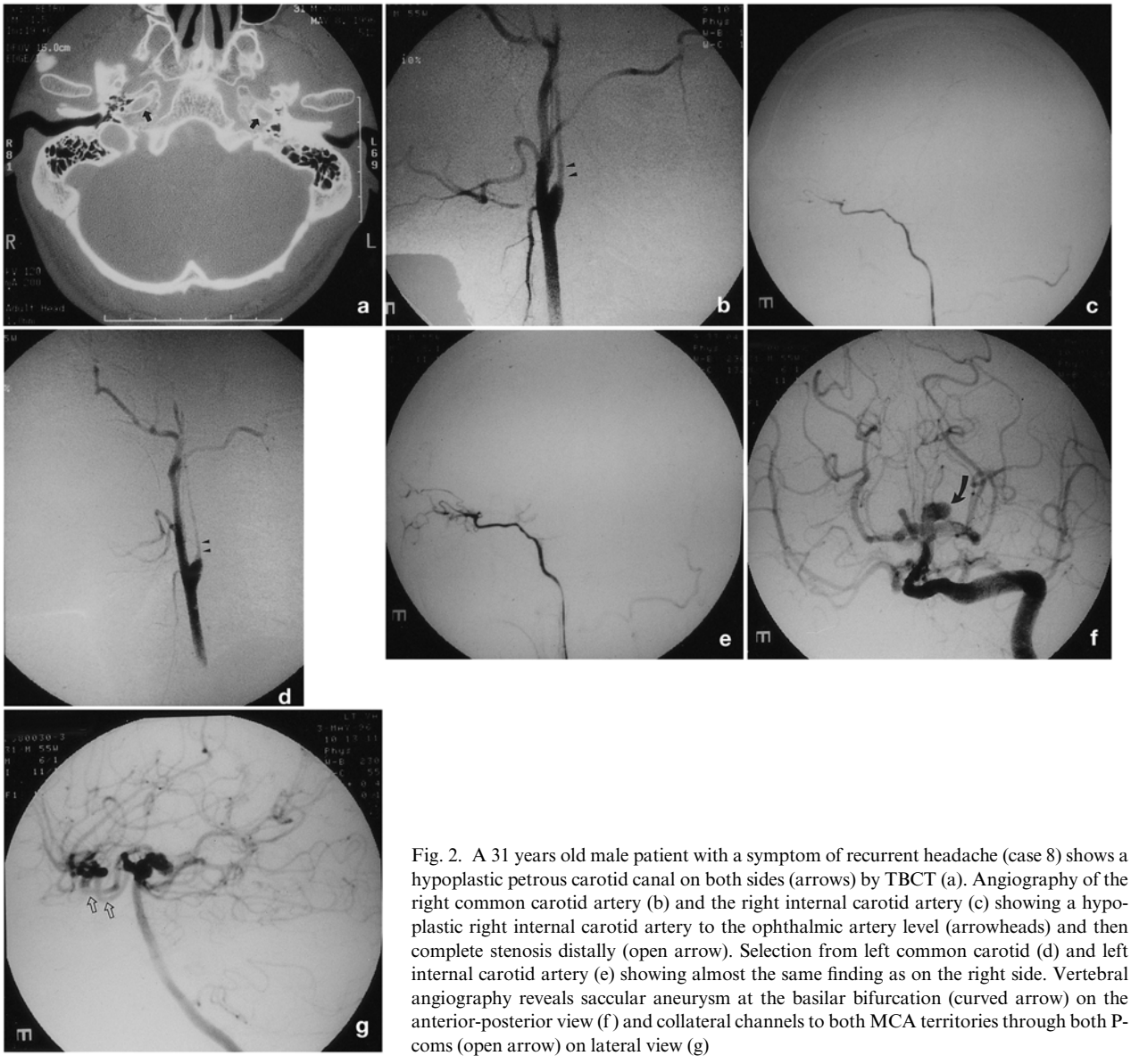

Fig. 2. A 31 years old male patient with a symptom of recurrent headache (case 8) shows a hypoplastic petrous carotid canal on both sides (arrows) by TBCT (a). Angiography of the right common carotid artery (b) and the right internal carotid artery (c) showing a hypoplastic right internal carotid artery to the ophthalmic artery level (arrowheads) and then complete stenosis distally (open arrow). Selection from left common carotid (d) and left internal carotid artery (e) showing almost the same finding as on the right side. Vertebral angiography reveals saccular aneurysm at the basilar bifurcation (curved arrow) on the anterior-posterior view (f) and collateral channels to both MCA territories through both P-coms (open arrow) on lateral view (g)

artery was present in only one case with left ICA agenesis (case 2).

Carotid canal

The decreased size or absence of the lumen of the carotid canal compared to the contralateral normal side was demonstrated in all nine cases by TBCT. All cases but one demonstrated agenesis of the bony carotid canal and the remaining one (case 8), with both ICA hypoplasia, showed hypoplastic canal in its petrous portion over the foramen lacerum on both sides (Fig. 2).

Collateral channel

Collateral circulations to the MCA territory ipsilateral to the ICA aplasia were: via the P-com in five cases, including three cases of subarachnoid haemorrhage and one case of both ICA aplasia (Fig. 2), via the A-com in two cases, including the nasopharyngeal angiofibroma case (Fig. 3), and via both P-com and

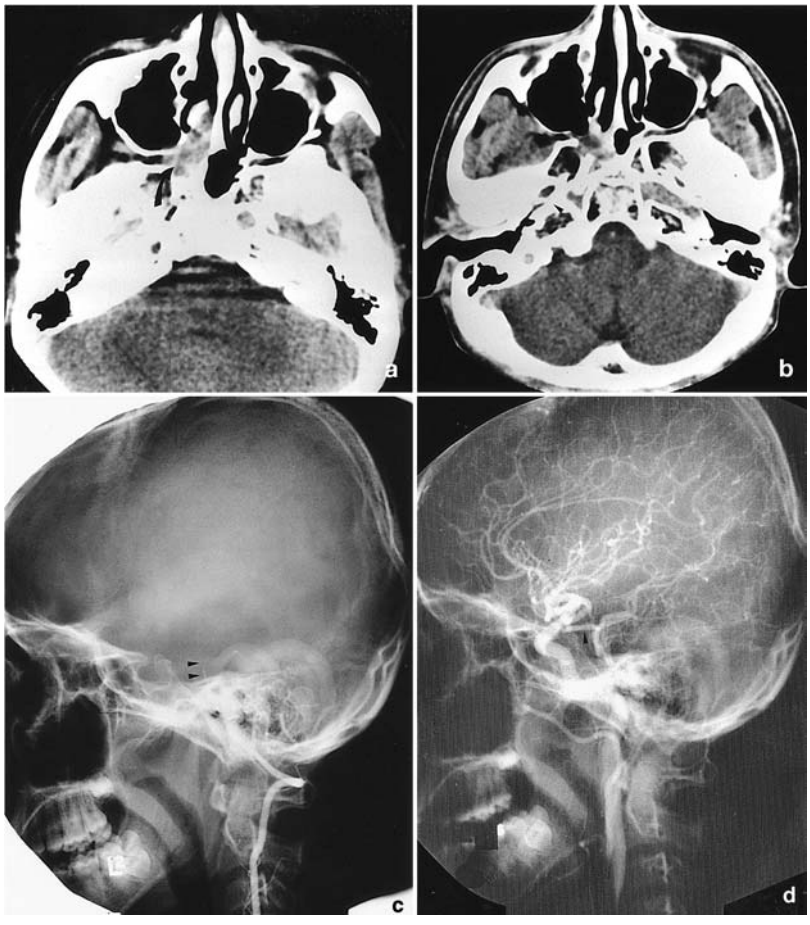

Fig. 3. A 15 years old male patient with symptom of recurrent epistaxis (case 7) had a mass lesion with slightly high attenuation in the right side of the nasopharynx (curved arrow) (a) and complete absence of the petrous carotid canal in the right side on the TBCT (arrowhead) (b). He was diagnosed with nasopharyngeal angiofibroma later. Vertebral angiography in the lateral view showing upper basilar artery atresia (arrowheads) (c). Carotid angiography reveals the collateral channel to both posterior cerebral arteries through both P-coms from the anterior circulation (arrowhead) (d)

A-com in two cases including the case with TMJ ankylosis. No abnormal collaterals, such as transsellar intercarotid connections or from the external carotid artery ('rete mirabile') were found.

Associated cerebral aneurysm

Cerebral aneurysms were found in six patients, four with anterior communicating artery (A-com) aneurysm, one with a basilar bifurcation aneurysm (Fig. 2), one with both right posterior communicating artery (P-com) aneurysm and left cavernous ICA aneurysm (Fig. 1). The three incidentally found cases had no aneurysm. All such aneurysms were located ipsilateral to the side of ICA aplasia. Three of the four A-com aneurysms and the basilar bifurcation aneurysm presented as ruptured cases with subarachnoid haemorrhage, while the other two cases, presented as ischemic events, were incidentally found on angiography performed to examine the possibility of occlusion of the ICA from an acquired cause, such as atherosclerosis.

Other associated congenital anomalies

Other associated abnormalities were found in four cases, these included: one case of hypoplasia of the common carotid artery (CCA) with an arachnoid cyst at the temporal pole (case 2), one case of abnormal origin of the right CCA (from aorta) and the right subclavian artery (from descending aorta) (case 5) (Fig. 1), one case of congenital temporomandibular joint (TMJ) ankylosis (case 6) and finally one case of nasopharyngeal angiofibroma with atresia of the upper basilar artery (case 7) (Fig. 3). Except for the atresia of the upper basilar artery, all such abnormalities were found on the same side as the ICA aplasia.

Discussion

Incidence & lateralization

The incidence of internal carotid artery agenesis is estimated to be less than 0.01% [30], with an usual

symptom onset in young adulthood rather than in childhood or adolescence. This condition is known to occur more frequently in left sided ICA's (according to the previously reported data, the ratio between right : left : both is 1 : 3 : 1) [11]. In our series, the right side of the ICA was involved almost twice as often as the left side (5 : 3).

Associated intracranial vascular anomalies

Three types of intracranial vascular anomalies are known to be associated with hypoplasia of the internal carotid artery: 1) aneurysms related to the circle of Willis, 2) arterial anomalies with abnormal collateral channel around the circle of Willis and 3) dilated vascular channels [20].

Association with Cerebral Aneurysm. The incidence of intracranial aneurysm in association with agenesis or aplasia has been reported as 25–43%, which is much higher than that found in the general population, 2 to 4% [1, 19, 28, 36]. In our series, six of the nine patients (67%) had associated aneurysms, which is twice as high a rate than previously reported. Two mechanisms have been postulated to explain this strong association between intracranial aneurysm and agenesis or aplasia of the internal carotid artery; 1) both conditions could occur independently during embryonic life as a result of developmental error [34], or 2); the aneurysm might develop secondary to the hemodynamic derangement [1, 8, 28]. More aneurysms or kinks and coils of the contralateral ICA or extracranial carotid arteries have been observed in cases with unilateral absence than in bilateral cases according to the previously reported data. This might be attributed to the increased hemodynamic load on the normal side [3, 19]. Furthermore, Tasker presented four observations that support the latter concept; 1) aneurysm formation in the distribution of the posterior cerebral and basilar arteries in moyamoya disease, 2) the association between anomalies of the cerebral vascular tree and aneurysm formation, 3) that acquired carotid occlusion favours aneurysmal formation, enlargement, or rupture and 4) the aneurysmal development contralateral to a iatrogenic carotid artery occlusion. He concluded that increased regional blood flow is an important factor in the development of cerebral aneurysms, along with congenital defects of the vessel wall, systemic hypertension, and certain other factors [35]. However, in our series, all six patients presented with an aneurysm on the ipsilateral side to the aplastic ICA, supporting rather their congenital origin than any hemodynamic factors.

Abnormal collateral channels. Collateral circulation accompanying agenesis or aplasia of the internal carotid artery can be classified into three forms; 1) transcranial anastomosis from the external carotid artery (so called 'rete mirabile'), 2) persistent embryonic vessels, such as a trigeminal artery, and 3) normal anastomotic pathways through the circle of Willis [33, 43]. The collateral blood supply is needed to compensate the affected carotid distribution [33], but it may contribute to form other anomalies associated with internal carotid artery anomaly. Several authors have reported upon these abnormal collateral channels; Osborn *et al.* reported an internal carotid artery hypoplasia with extensive collateral vessels from maxillary arteries on both sides and the ophthalmic artery [24]. Beresini reported a collateral circulation via the external carotid artery – vidian, accessory meningeal, and foramen rotundum arteries – in a case involving the absence of the internal carotid artery bilaterally with sellar enlargement by eroded sella turcica [5]. Moreover, Elefente *et al.* reported unusual transsellar intracavernous intercarotid connections, which Lie proposed to have resulted from fusion of two primitive trigeminal arteries [12]. He further suggested that the primitive maxillary artery might have persisted and made a collateral channel in this case [9]. In addition, some have reported anastomotic channels via the circle of Willis in agenesis or aplasia involving the internal carotid artery. Tsuruta *et al.* proposed 'Tsuruta and Miyazaki's classification' for agenesis of the internal carotid artery. He distinguished three types for unilateral agenesis, according to the collateral channels via circle of Willis [39]. In type I, the anterior cerebral artery on the side with arterial absence is filled from the contralateral side ICA and the middle cerebral artery from the basilar artery via the P-com artery. In type II, in which the anterior cerebral artery is filled from the middle cerebral artery on the same side. And in type III, in which the blood flow on the side with arterial absence is supplied from the contralateral ICA via anastomotic vessels. Although the absence of one or both ICAs may be entirely asymptomatic under usual condition, care should be taken when performing an operation in and around the sella turcica. This might be due to the risk of interrupting associated cerebral aneurysms or abnormal collateral channels, such as intercarotid communicating artery [19, 39]. In our series, no such abnormal collateral channels were detected.

Embryological origin

The above mentioned collateral circulations associated with the anomalous internal carotid artery are not only a compensatory mechanism, but are also a finding suggestive of the congenital nature of the internal carotid artery anomaly [31, 33]. Normally, the internal carotid arteries are derived from portions of the first and third aortic arches and the paired dorsal aorta, when the embryo has attained the 3-mm stage. The root portions of the internal carotid arteries are formed from the third aortic arches. The dorsal aorta between the third and first arches form the intermediate portions, and the distal parts of the internal carotid arteries originate from the dorsal ends of the first aortic arches [8]. In the 4-mm embryo, the anterior and posterior divisions of the internal carotid arteries begin to form distally. These divisions subsequently form the anterior, middle, and posterior cerebral vessels. At the 5- to 8-mm stage, the basilar artery arises from the posterior extensions of the posterior communicating arteries bilaterally. In the 14-mm embryo, the vertebral arteries are virtually completed, having formed from anastomoses between the cervical segmental branches of the dorsal aorta. The upper cervical segmental branches now unite the vertebral and basilar arteries. All aortic connections are subsequently obliterated except for the seventh intersegmental branches that become the subclavian arteries. The common carotid arteries begin to form in the 12- to 14-mm embryo, following the involution of portions of the paired vertebral aortic roots between the third and fourth aortic arches. The external carotid arteries arise from the aortic sacs and migrate up to the third arches. The portions of the third arches proximal to the external carotid arteries become the common carotid arteries [26]. Agenesis or aplasia of internal carotid artery depends on the abnormal regression of the first and third aortic arches [16, 27]. Kunishio *et al.* also mentioned that the anomaly is based on the atresia or involution of the third aortic arches and the distal potion of the dorsal aortas in the 20-to-24-mm stage [19, 37]. As the basilar artery is formed at the 7–12-mm stage of embryonal development and the circle of Willis is complete when the anterior communicating artery is formed at the 24-mm stage, it seems that the collateral circulation via the circle of Willis reflects agenesis or aplasia of the internal carotid artery during early embryonic development i.e., after the development of the basilar artery, but prior to the completion of the circle of Willis [43]. Also, it has been hypothesized that anomalous transsellar communications represent the primitive circulation which persisted when the ICA involuted, that is before the development of the circle of Willis [6, 22, 32].

Clinical presentations and other associated anomalies

Agenesis or hypoplasia of internal carotid artery may be asymtomatic [5, 30, 40]. Despite the markedly altered vascular anomaly, MRI or SPECT did not frequently demonstrate evidence of brain lesions or perfusion defects in previous reports [17]. However, there are various reported clinical presentations, such as, headache [24], symptomatic epilepsy [42], cerebral ischemia [4, 37], hemiplegia [10], or intracranial haemorrhage [10, 20, 37]. Osborn *et al.* found that these symptoms did not lend themselves to correct diagnosis [24]. Also, sporadic cases of absent ICA have been reported to be associated with cerebral hemiatrophy [1], Klippel-Trenaunay syndrome [13], cardiac anomaly such as ventriculoseptal defect [2, 14], arachnoid cyst [18], neurofibromatosis [7] and even with a large haemangioma of the tongue [23]. In our series, nasopharyngeal angiofibroma or temporal tip arachnoid cyst located on the same side as ICA aplasia was demonstrated. These multiple maldevelopments seem to have occurred almost simultaneously on the same side during the formative stage of the vasculature at 4–5 weeks of embryogenesis.

Radiological findings

The diagnosis of agenesis of internal carotid artery has two aspects; 1) the absence of the internal carotid artery by angiography, and 2) the absence of the bony carotid canal to exclude acquired carotid occlusion [9, 15, 24]. The latter can be shown by autopsy, operative findings, or by tomography of the bony carotid canal, but more practically, CT scanning offers an accurate and non-invasive in vivo method of examining the temporal base of the skull [12, 15, 25]. Recently, colour Doppler imaging is also known to give important clues for the correct diagnosis of ICA aplasia by demonstrating the significantly reduced lumen diameter of the affected side ICA, and by detecting the large communicating arteries, because the lumen reduction is not visible in cases with ICA occlusion of atherosclerotic origin or after dissection [29]. Absence of the carotid canal in agenesis of ICA can be also interpreted em-

bryologically, as the primordial ICA is well defined by the fourth embryonic week, while the skull base does not begin to form until the fifth or sixth week of fetal life. Therefore, if the embryonic primordium of the ICA fails to develop or involutes early during embryonic development, no carotid canal develops [25]. Worthington *et al.* mentioned that the above two criteria might be necessary but are in themselves insufficient to prove true carotid agenesis. So he proposed that a collateral circulation consistent with an early developmental stage as another condition that could be used to diagnose the anomaly [43]. So the agenesis or aplasia of the ICA is supported in our series by a faint or a lack of visualization of the ICA along its entire course, a small sized or an absent bony carotid canal, and by a collateral circulation consistent with an early developmental stage.

Conclusion

Agenesis or aplasia of internal carotid artery may be entirely harmless. However, associated conditions may be of clinical importance. In particular, the asymmetry or absence of one or both of the carotid canals should prompt further evaluation to rule out the presence of potentially life-threatening intracranial vascular abnormalities such as cerebral aneurysm, even in an asymptomatic patient. In addition, we must both recognize the anomaly and the significance of its associated collateral system and should be alert whilst performing manipulation around the sellar portion, when using temporary clipping, or when inducing hypotension, because such an iatrogenic manipulation may disturb compensatory collateral circulation and induce irreversible neurological deficit.

Acknowledgment

This work is supported in part by a grant from the Seoul National University Hospital Research Fund.

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Comments

The authors present a series of patients with internal carotid hypoplasia. True aplasia appears to be a very rare anomaly, and the review of the entity and the associated anomalies which usually lead to the diagnosis is interesting. As stated also by the authors, internal carotid hypoplasia is not caused by a single development aberration but by a group of potential mechanisms. Some of the cases presented may be explained by internal carotid occlusion during childhood, for example by moyamoya like mechanisms.

H.-J. Steiger

The authors present nine new cases of aplasia of the internal carotid artery (ICA) along with an extensive revision of this subject. It is a good original series collected from 12 years of clinical practice and represents about one tenth of all published cases on this pathology. Some prominent features of these new cases are worth noting: eight cases had the defect located in the bony carotid canal; all cases were symptomatic, three out of them due to SAH and three to cerebral ischemia; and cerebral aneurysms coexisted in six patients, always ipilateral to the aplasia. These features are somewhat different to what is reported in the majority of previous publications. Moreover, they show the potential clinical danger that this entity involves, as well as the probable congenital role (rather than the hemodynamic secondary effect) of the associated vascular anomalies, namely the cerebral aneurysms. A great deal of attention is drawn to the embryological and pathophysiological aspects of these malformations, that are reviewed and discussed in detail. Although no surgical intervention was carried out on these patients, their outcome would be worth mentioning, so the clinical consequences of such rare cerebral vascular anomalies could be better appreciated.

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