Congenital Absence of the Internal Carotid Artery

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Abstract

Absence of the internal carotid artery is thought to result from developmental failure, which occurs before the complete development of the circle of Willis. Even though most patients are asymptomatic, this anomaly may be associated with other potentially severe malformations and disorders. An absent or hypoplastic carotid canal found on Computed Tomography (CT) scanning, suggests the presence of a congenital vascular anomaly with or without a potentially life-threatening intracranial aneurysm; further evaluation with CT angiography, or Magnetic Resonance (MR) angiography should be considered in such cases. We present the clinical features and radiological findings of five cases of congenital absence of the internal carotid artery.

Key words: Congenital, Internal carotid artery

Introduction

Congenital absence of one or both internal carotid arteries (ICA) is a rare abnormality, which occurs in less than 0.01% of the population. Unilateral agenesis is more common than bilateral ICA agenesis. There are three types of developmental anomalies of the ICA. Agenesis is defined as the total failure of the development of an organ, aplasia as a lack of development in spite of precursors and hypoplasia as incomplete development of an organ. Absence of the carotid canal is associated with agenesis of the ICA. However, if there is a carotid canal, aplasia or hypoplasia might be present. Intrasellar intercarotid communicating arteries are common if one of the ICAs is absent. In cases with bilateral ICA agenesis, the anterior communications are usually supplied by enlarged posterior communicating arteries and the basilar artery through the circle of Willis. There is also a high incidence of intracranial aneurysms associated with ICA agenesis due to the development of collateral vessels. This increases the incidence of intracranial hemorrhaging. However, there have been some cases reported of intracranial hemorrhage without evidence of an aneurysm.

Here we report five cases of aplasia or hypoplasia of unilateral or bilateral ICAs in two males and three females the age ranged from 30 to 58 years. The diagnosis of hypoplasia or agenesis of the ICA was based on a hypoplastic or absent bony carotid canal by CT scan of the skull base and conventional angiography, CTA or MR angiographic findings.
Methods and Results

Case 1

A 35-year-old woman presented with generalized tonic seizures with sudden onset four hours prior to presenting to the hospital, and lasting for two to three minutes. The patient felt a tingling sensation in the left arm for several months. The brain MRI showed focal small infarcts in the left basal ganglia. The transfemoral cerebral angiography revealed bilateral hypoplastic ICAs (Fig. 1A, B). The branches of the ICA distal to the ophthalmic artery were not visualized (Fig. 1C). Both vertebral arteries were prominent and the posterior communicating arteries were enlarged and supplied the anterior circulation (Fig. 1D, E). This patient was diagnosed with bilateral hypoplasia of the ICAs.

![Fig. 1. A thirty-five year-old woman presented with generalized tonic seizures.](image)

(A) Right ICA angiogram shows hypoplastic right ICA (arrows). (B) Left ICA angiogram shows a hypoplastic left ICA (arrows). (C) The branches distal to the ophthalmic artery of the ICA are not visualized. (D, E) Both vertebral arteries are prominent and the posterior communicating arteries that supply the anterior circulation are enlarged. There was no evidence of a vascular anomaly such as an aneurysm.

Case 2

A 58-year-old man presented with a history of intermittent headaches for several months. The brain MRI and CT angiography of the neck revealed almost a completely non visualized right ICA and a partially visualized right supraclinoid ICA. The supraclinoid ICA was supplied from the right posterior communicating artery (Fig 2A, B). Conventional angiography was not performed. On the CT angiography, the lumen of the right
carotid canal was not well delineated in contrast to the left carotid canal the left ICA was normally visualized (Fig. 2C). At that time, the patient was diagnosed with cancer of the right tonsil with metastasis to the tongue and salivary glands. Wide excision of the right tonsil with a modified radical neck dissection was performed.

Case 3

A 51-year-old woman who was diagnosed with stable angina due to significant stenosis of the proximal left anterior descending artery had a PCI stent inserted. The patient complained of paresthesias of both feet and hands at night for 1 year. Nonvisualization of the left ICA with a small left CCA was identified, and the left carotid canal was absent on the CT angiography of the neck. The left ACA and MCA were visualized through the anterior communicating and posterior communicating arteries.

Case 4

A 30-year-old man complained of a headache. The brain MRI showed no abnormal signal intensities in the brain parenchyma. The right ICA was not seen after the bifurcation and the left vertebral artery was prominent on the MRA. Conventional angiography revealed that the right CCA could be visualized from the aortic arch directly; however, the ICA was not seen (Fig. 3A). The right MCA originated from the basilar artery through the right PCA (Fig. 3B, C). On the left internal carotid arteriogram, the right ACA was visualized through the anterior communicating artery from the left ACA; the right MCA was visualized through a small connecting artery (Fig. 3D). There was no evidence of an aneurysm or AVM. The right subclavian artery originated from the descending aorta; the right vertebral artery was the first branch of the subclavian artery (Fig. 3E).
Case 5

A 58-year-old woman was transferred to our hospital due to a subarachnoid hemorrhage. The patient had a severe headache for two days. There was a history of mitral valve stenosis that was treated by surgery 35 years ago. The brain CT scan revealed a subarachnoid hemorrhage mainly in the right Sylvian fissure. The left carotid canal was significantly narrowed compared to the right side (Fig. 4A). The CT angiography showed a markedly narrowed left ICA. On the cerebral angiography, the left ICA was diffusely and markedly narrowed from the proximal portion, and the right MCA was obstructed at the M1 portion with delayed visualization of the MCA branches through irregular collaterals from the
Fig. 4. A fifty-eight years-old woman presented with a subarachnoid hemorrhage. (A) The brain CT revealed a left carotid canal (arrowheads) that was significantly narrowed compared to the right side (arrows). (B) On cerebral angiography, the left ICA was diffusely and markedly narrowed from the proximal portion.

ACA and proximal M1 branches (Fig. 4B). Although there was a subarachnoid hemorrhage noted on the brain CT, the brain CTA and angiography did not show evidence of an intracranial aneurysm. The clinical and imaging findings of the reported cases are summarized in Table 1.

### Discussion

Patients with ICA agenesis or hypoplasia are usually asymptomatic because cerebral perfusion is usually adequate due to the development of collaterals. Common symptoms and signs include headaches, blurred vision, hearing loss, hemiparesis with or without cranial nerve palsy, epilepsy, cerebrovascular insufficiency, and in the most severe cases, intracranial hemorrhaging due to a ruptured aneurysm. The patients in this report presented with headache, seizure and paresthesias of both the feet and hands.

The exact mechanisms leading to agenesis of the ICA remain unknown, but some investigators have suggested the occurrence of various mechanical disturbances during early development, such as pressure effects and excessive bending of the cephalic portion of the embryo to one side or the other. Lee et al. reported six cases with an
aneurysm that had agenesis or aplasia of the ICAs, and in all six cases, the aneurysms were detected ipsilateral to the aplastic ICA, supporting a congenital developmental abnormality rather than hemodynamic factors.⁵

ICAs develop from portions of the first and third aortic arches and paired dorsal aorta, at the 3-mm embryo stage. The root and distal portions of the ICAs each originate from the first and third aortic arches. The dorsal aorta, between the first and third arches, forms the intermediate segment.⁶

The external carotid arteries arise from the aortic sacs and migrate up to the third arches. The CCAs develop from the third arches proximal to the external carotid arteries in the 12- to 14-mm embryo.⁷ Therefore, the developmental abnormalities of the first and third aortic arches result in agenesis or aplasia of the ICA. The carotid canal develops in association with the ICA. The primordial ICA is well defined by the fourth embryonic week. However, the skull base does not begin to form until the fifth to sixth weeks of fetal life. If the embryonic primordium of the ICA fails to develop before the third to fifth embryonic weeks or involutes early in embryonic life, the ICA and carotid canal cannot develop. However, if an insult is either relatively mild or occurs relatively late during the course of development of the skull base portion of the ICA, a hypoplastic ICA within a hypoplastic carotid canal can result.⁸

In three of the cases reported here, the carotid canal of the skull base was not delineated ipsilateral to the nonvisualized ICA. Therefore, these cases illustrate agenesis of a unilateral ICA. For the other two cases bilateral or unilateral hypoplastic bony carotid canals were identified, and hypoplasia was diagnosed. A reduced diameter of the lumen of an ICA on the affected side, on conventional angiograms, provides a clue to the correct diagnosis, because a reduced lumen is not visible in cases with ICA occlusion of atherosclerotic origin or after a dissection.⁹

Aneurysms and abnormal collateral channels around the circle of Willis are commonly associated vascular anomalies such as agenesis or aplasia of ICAs. The incidence of intracranial aneurysms associated with agenesis or aplasia has been reported in about 25-67% of cases, which is much higher than that found in the general population, which is 2-4% without this anomaly presenting with an intracranial hemorrhage as the initial symptom.¹⁰ However, none of our cases had an aneurysm. In one case, an aberrant subclavian artery developed from the descending aorta. This was the case of a 58 year old man diagnosed with right tonsillar cancer and metastasis to the right side of the tongue.

In cases of unilateral absence of the ICA, three types of collateral circulation were suggested by Lie: the fetal type is the most common of the three types of collateral circulation the ipsilateral anterior cerebral artery, to the side of the agenesis or aplasia of the ICA, is supplied by the normal contralateral ICA via the anterior communicating artery, whereas the middle cerebral artery is supplied by an enlarged posterior communicating artery. This type of collateral circulation develops during early embryonic stages. If both the anterior and middle cerebral arteries are fed by the anterior communication artery, this is referred to as the adult type, because it resembles what usually occurs in cases of thrombosis of the ICA. Intercavernous, supraclinoid anastomosis was suggested by Lie to be a fusion of two primitive trigeminal arteries that develop from the contralateral ICA in the absence of the cervical and petrous portion of the ICA, this is the third type.⁵

The lumen of the carotid canal was not de-
lineated in all cases on the CT scans of the skull base. In this report, there are four cases with the fetal type of collateral circulation that the MCA supplies through the posterior communicating artery. There is one case with an ipsilateral MCA associated with ICA hypoplasia that was supplied through the ACA. There were no cases with transsellar intercarotid or external carotid artery connections.

**Conclusion**

We report five cases of agenesis or hypoplasia of the ICA. This anomaly might be associated with other intracranial aneurysms or the development of collateral vessels. Some patients present with clinical findings due to intracranial hemorrhaging or malformations of other vessels. An absent carotid canal on CT scanning and angiography should raise suspicion for agenesis or hypoplasia of the ICA and further evaluation for the detection of collaterals and/or an associated aneurysm should follow.

**References**
