Bilateral hypoplasia of the internal carotid artery

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Abstract

Agenesis and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies, occurring in less than 0.01% of the population. We report a rare case of bilateral hypoplasia of the ICA in a patient with post-traumatic subarachnoid hemorrhage. We describe the embryological development of the cerebral vasculature and present a review of literature.

Key words: Agenesis; aortic arches; developmental anomaly; hypoplasia; internal carotid artery

Introduction

Agenesis and hypoplasia of the internal carotid artery (ICA) are very rare congenital anomalies, occurring in less than 0.01% of the population. They are more often unilateral. Bilateral ICA agenesis or hypoplasia is even rarer. Of the slightly more than 100 cases of ICA agenesis or hypoplasia reported, only around 20 were bilateral. Many of these patients are asymptomatic due to good collateral flow, and the anomaly may be detected only incidentally. We report a case of asymptomatic bilateral ICA hypoplasia in a young adult, which was detected incidentally while the patient was being investigated for a head injury. The pertinent literature and the embryological development of the cerebral vasculature are reviewed.

Case Report

A 36-year-old man was brought to the emergency department with a history of fall from a height of 8 feet. The cause of his fall was not known as he had amnesia for the event. He had had transient loss of consciousness, but there were no significant external injuries. Neurologically he was drowsy but there were no other deficits. CT scan of the head revealed subarachnoid blood in the right sylvian fissure and right frontoparietal convexity, along with a small temporal hematoma [Figure 1]. He was further investigated with digital subtraction angiography to rule out a distal middle cerebral artery (MCA) aneurysm. The angiogram revealed bilateral hypoplastic ICAs. The common carotid artery (CCA), the proximal 1 cm of the ICA, and the external carotid artery (ECA) were normal bilaterally. The cervical and petrous carotids on both sides were visualized as thin streaks, while the cavernous and the supraclinoid part of the ICAs up to the origin of the posterior communicating artery (PCoA) were not visualized [Figure 2]. The anterior circulation was supplied by the basilar artery through bilateral dilated and tortuous PCoAs. The right ophthalmic artery was probably arising from the anterior carotid artery–anterior communicating artery (ACA-ACoA) complex. The left ophthalmic artery was not visualized [Figure 3]. There were no associated aneurysms or abnormal transcranial ECA–ICA anastomosis or any primitive persistent arteries. CT scan showed bilateral diminutive carotid canals in the petrous bone [Figure 4]. The hypoplastic ICAs were considered to be incidental and congenital in nature. As there was no evidence of aneurysms or vessel wall pathology, the subarachnoid hemorrhage was attributed to trauma and the patient was managed conservatively. He was discharged 9 days after admission at which time he was conscious and alert with no neurologic deficits.

Discussion

Agenesis and hypoplasia of the ICA are rare developmental
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Figure 1: Axial noncontrast CT scan shows a right Sylvian fissure bleed (arrow)

Figure 2 (A,B): Frontal digital subtraction angiograms show bilateral ICA hypoplasia, with only the proximal ICA being normal (arrows)

Figure 3 (A,B): Lateral (A) and AP (B) digital subtraction angiograms show the ACA and MCA on both sides being filled by the basilar artery via dilated PCoAs (arrowhead in A and curved arrow in B). The right ophthalmic artery (arrows) is seen arising from the ACA–ACoA region.

Figure 4 (A,B): Axial CT scans (bone windows) show bilateral small carotid canals (arrows)

The development of the aortic arch, great vessels, and the craniocerebral vasculature starts around the 24th day of embryonic life, with the appearance of the first aortic arch. The first two aortic arches involute by day 28 of embryonic life. The third aortic arch, which is well formed by then, forms the CCA and the proximal portion of the ICA. The rest of the distal ICA arises from the cranial part of the dorsal aorta [Figure 5]. The ECA arises as a sprout from the CCA (i.e., the third aortic arch) and also receives contribution from the first and second aortic arches. The stapedial artery from the second aortic arch contributes to the formation of the internal maxillary branches and the middle meningeal branch of the ECA. Initially, during the 4th week of embryonic life, the aortic arches are connected to the dorsal aorta. As development progresses, the connection of the first and second arches to the dorsal aorta regresses and they contribute to the formation of the ECA. Nonregression of the communication with the dorsal aorta may persist to some insult sustained by the growing embryo during the 4th–8th weeks of gestational life. The cause of the unilateral hypoplasia of the ICA has been postulated to be exaggerated folding of the embryo to one side or constriction by amniotic bands. However, the cause of bilateral hypoplasia has not been identified.[2]

The terms agenesis, aplasia, and hypoplasia of the ICA are often used erroneously interchangeably.[3] Agenesis refers to the complete failure of development of the ICA; hypoplasia indicates that the development of the ICA has taken place, but the artery is of a very small caliber; and the term aplasia is used when only vestiges of the ICA are present.[5] The cause of incomplete development of the ICA is probably due
as a transcranial ECA–ICA anastomosis. Through this anastomosis the internal maxillary artery and the middle and accessory middle meningeal arteries can supply the distal ICA in cases of hypoplasia of the ICA (known as rete mirabile in the region of the cavernous sinus).

The basilar artery develops as two longitudinal vascular plexuses dorsal to the third and fourth arches during the 5th week of intrauterine development. A number of primitive vessels connect the developing basilar artery with the ICA, all of which involute and disappear except for the cranial-most one, which persists as the PCoA [Figure 6]. In cases of unilateral or bilateral ICA agenesis or hypoplasia, the PCoA most commonly enlarges and supplies the ACA and the MCA territories. Lie classified possible collaterals in cases of ICA agenesis into six types. Bilateral absence of the ICA, with dilated bilateral PCoAs supplying the anterior circulation (our case), comes under type C. In types E and F also there is bilateral ICA hypoplasia with bilateral PCoAs supplying the MCA, the hypoplastic ICA supplying the ACA in type E and bilateral rete mirabile in type F. In cases of unilateral hypoplasia, the distal ICA can also receive collaterals from other persistent primitive arteries or through intercavernous anastomoses.

The origin of the ophthalmic artery in cases of ICA agenesis has not been commonly reported. Embryologically, there is a ventral and a dorsal ophthalmic artery supplying the orbit. The former arises from the ACA and the latter from the carotid siphon region. The ventral artery anastomoses with the ICA in the supracavernous portion (the region where the artery arises in adults) and loses its proximal attachment to the ACA. Another anastomosis takes place in the orbit between the ventral and dorsal ophthalmic arteries, and the distal portion of the dorsal artery loses its proximal connection. The proximal part of the dorsal ophthalmic artery persists as the inferolateral trunk of the cavernous ICA. Anomalous origin of the ophthalmic artery in cases of ICA agenesis is most commonly from the ACA. Other sites of origin include the PCoA and basilar artery. In our case, the exact site of origin of the right ophthalmic artery could not be identified. It probably arose from the ACA–ACoA region. On the left side, the ophthalmic artery was not visualized.

Hypoplasia of the ICA must be differentiated from acquired stenosis (especially when unilateral). Unilateral narrowing of the ICA is more likely to be due to chronic dissection, severe atherosclerosis, fibromuscular dysplasias or, sometimes, moyamoya disease. It is well known that the carotid canal gives the clue to the presence of hypoplasia of the ICA. The carotid canal is closely linked to the development of the ICA during embryonic life. The skull base develops between the 5th and 6th weeks of embryonic life and is temporally associated with ICA development. The presence of a developing ICA is essential for the formation of the carotid canal. Thus the carotid canal is absent in cases of agenesis and is small and not well developed in cases of hypoplasia, as in our case. In acquired stenosis of the ICA, the carotid canal will be of normal size as the ICAs are developmentally normal.
Many patients are asymptomatic due to good collateral flow and the condition is usually diagnosed incidentally. The presence of a well-perfused brain in such cases has been shown using single-photon emission computed tomography (SPECT) studies. However, patients can present with transient ischemic attacks, infarct, seizures, aneurysmal subarachnoid hemorrhage, or parenchymal bleeds. In our case, the CT scan showed a right sylvian fissure and a cortical surface subarachnoid bleed, which prompted us to get an angiogram done to rule out a ruptured distal MCA aneurysm. However, as the angiogram did not reveal any aneurysm or vascular pathology to account for the cortical bleed, the patient was managed conservatively.

There is an increased risk of aneurysm formation in patients with aplastic or hypoplastic ICAs (23%–45% as compared to 2%–4% in normal individuals). These aneurysms are more common along the basilar artery and the posterior cerebral artery (PCA). This could be due to increased hemodynamic stress as a result of increased flow through the collaterals and altered direction of blood flow and/or may be due to a genetic disorder that has led to both hypoplasia of the ICA and vessel wall weakness, predisposing to aneurysm formation. Our patient had tortuous and dilated posterior communicating and basilar arteries but no aneurysms.

**Conclusion**

Bilateral ICA hypoplasia is a very rare condition. Many cases are probably asymptomatic, being identified only incidentally. The exact cause of this developmental anomaly is not known. A small or an absent carotid canal indicates the congenital nature of the pathology. ICA hypoplasia has to be distinguished from acquired stenosis as the management of the two conditions is different.

**References**


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