

Case report: Bilateral agenesis of internal carotid arteries associated with aneurysmally dilated plexus of vessels in the posterior fossa

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Bilateral agenesis of the internal carotid arteries (ICAs) is a rare congenital anomaly and is associated with aneurysms of the circle of Willis^[1] and dolichoectasia of the vertebrobasilar system.^[2] The cerebral perfusion is maintained by collateral flow from the posterior circulation through the posterior communicating arteries, or by a persistent fetal circulation or, transcranially, through branches of the internal maxillary artery.^[3] We would like to describe an unusual posterior fossa malformation, consisting of a tangle of dilated arteries, associated with bilateral ICA agenesis—an association that we believe has not been reported before.

Case Report

A 10-years-old boy presented with a history of headache and vomiting for one year and medial deviation of both eyeballs for six months. Clinical examination revealed no significant abnormality except for bilateral VIth nerve paresis, which was more on the right side. There was no history of any surgical or vascular intervention.

An MRI was performed. It showed multiple flow voids in the perimedullary and prepontine cisterns, with tortuous and dilated left vertebral and basilar arteries [Figure 1]. Flow voids of the petrous portions of both ICAs were not seen. There were multiple small flow voids in both cavernous sinus regions. MRI angiography (MRA) of the cerebral arteries showed absence of both ICAs. The supraclinoid ICAs were reformed by multiple collaterals across the skull base. CT scan showed absent carotid canals on both sides [Figure 2] and bilateral enlarged foramina ovale due to dilated accessory meningeal arteries. There were multiple, round and oval, hyperdense foci with irregular calcifications in the perimedullary and prepontine cisterns due to a dilated tangle of vessels.

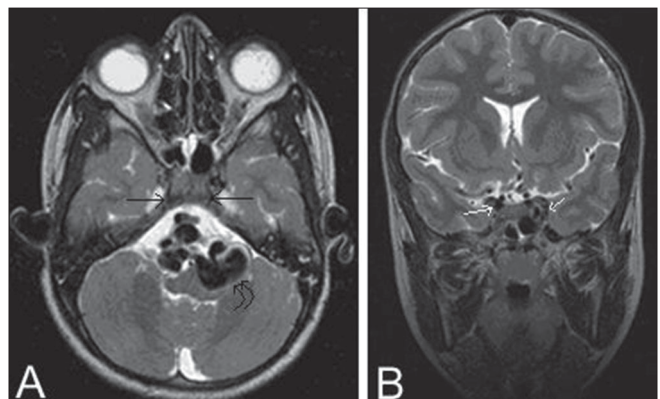


Figure 1 (A and B): T2W axial (A) and T2W coronal (B) images showing bilateral absent ICA flow voids (black arrows) with flow voids of collaterals in the cavernous sinus region (white arrows) and multiple dilated vessels in the posterior fossa (curved arrows).

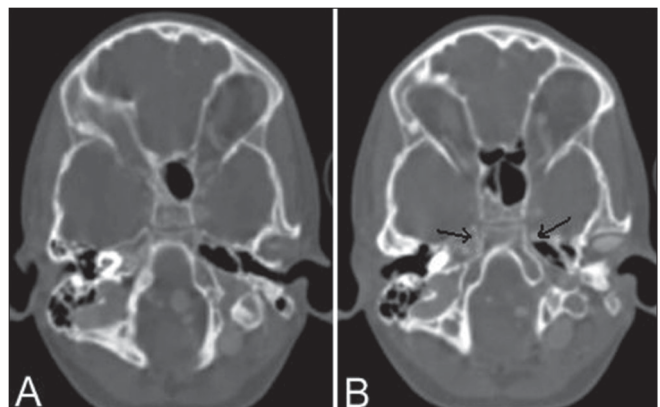


Figure 2 (A and B): Axial CT images of the skull base demonstrating the absence of carotid canals on both sides (arrows).

Digital subtraction angiography (DSA) [Figure 3] confirmed the absence of both ICAs. The common carotid arteries (CCAs) continued as the external carotid arteries (ECAs).

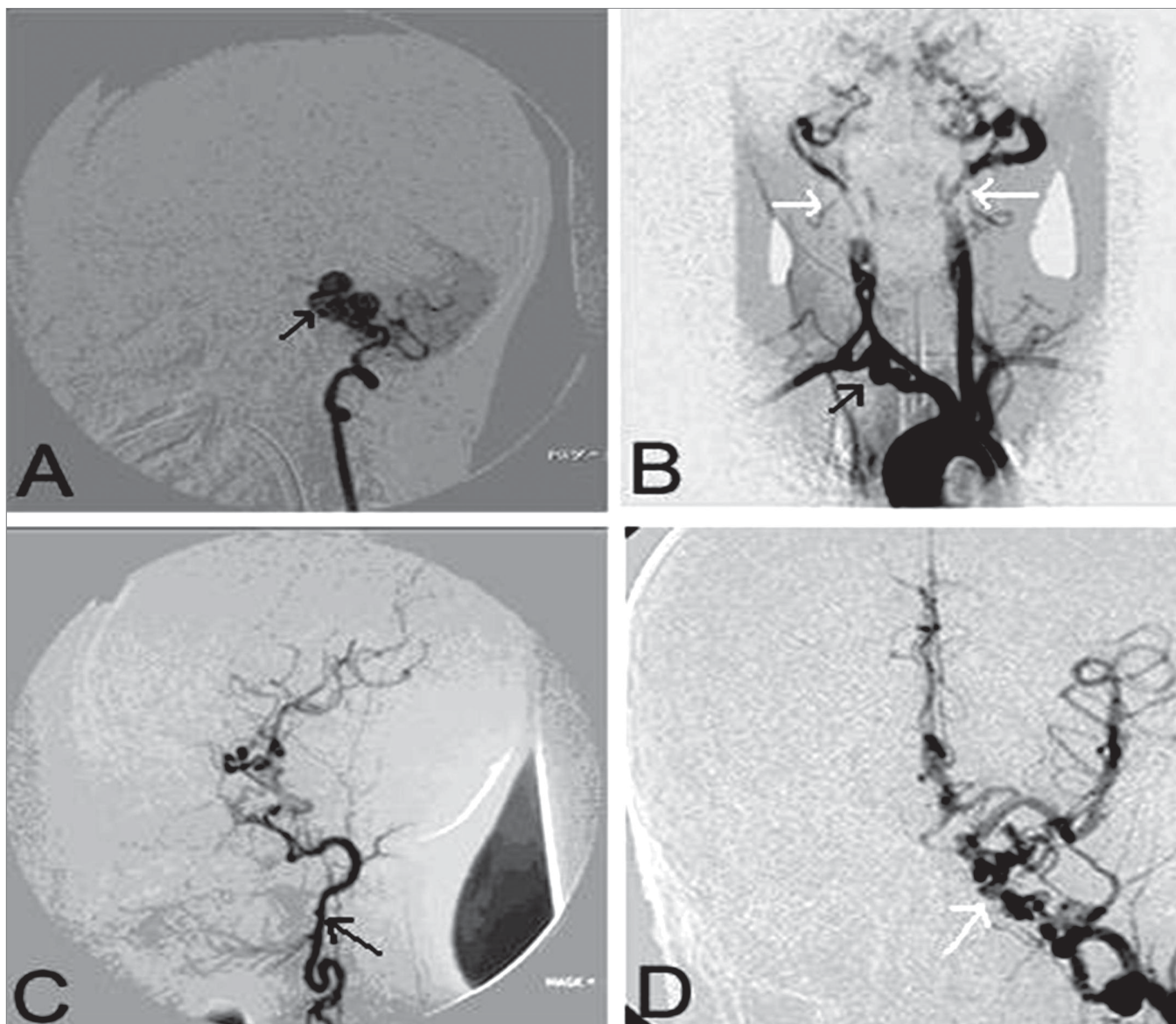


Figure 3 (A-D): Left vertebral arteriogram (A) showing a dilated tortuous tangle of vessels (black arrow). Arch aortogram (B) showing tortuosity of the right subclavian artery (black arrow) near its origin, with bilaterally absent ICAs (white arrow). Right CCA lateral arteriogram (C) and left CCA AP arteriogram (D), showing both CCAs continuing as ECAs (black arrow), without visualization of the ICAs. Transcranial collateral circulation is seen (white arrow).

The ECAs and their internal maxillary branches were dilated on both sides, especially so on the left side. Multiple dilated, tortuous, collateral branches from the left internal maxillary artery were seen to reconstitute the supraclinoid left ICA and the right anterior cerebral artery (ACA). The right middle cerebral artery (MCA) was reformed by tortuous collaterals from the right internal maxillary artery and the right posterior communicating artery [Figure 3C]. The right vertebral artery could not be cannulated, as the first part of the right subclavian artery was very tortuous [Figure 3B]. A plexus of vessels with multiple aneurysmally dilated sacs was seen arising from both vertebral arteries, mainly from the left, reconstituting a tortuous ectatic basilar artery [Figure 3A]. No early venous filling in the posterior fossa

was noted on left vertebral injection, thus excluding the presence of an arteriovenous malformation in the posterior fossa.

Discussion

Absence of the ICA is a rare congenital anomaly with an incidence of less than 0.01%.^[1] Bilateral absence of ICAs is rarer still, occurring in less than 10% of cases of ICA agenesis.^[1] Tode was the first person to document ICA agenesis in 1787 in a cadaver.^[3] The first case of ICA agenesis, diagnosed at angiography, was documented by Verbiest in 1954.^[4] Dysgenesis of the ICA is classified into agenesis, aplasia, and hypoplasia. The term agenesis is used

when there is total failure of ICA formation in the embryo; aplasia, when a precursor is present in the embryo and is represented by a remnant such as a fibrous band; and hypoplasia, when a small caliber ICA is present.^[1]

The exact etiology of aplasia, agenesis, and hypoplasia is not known. Embryologically, the ICA forms from the 3rd aortic arch and the CCA from the ventral aortic root between the 3rd and 4th arches. Lasjaunias *et al.*^[5] have suggested that the ICAs can be divided into six segments: cervical, petrous, vertical cavernous, horizontal cavernous, clinoid, and cisternal segments, which develop from the embryonic arteries: ventral pharyngeal, hyoid, mandibular, primitive maxillary, trigeminal, dorsal ophthalmic, and ventral ophthalmic, respectively. Each of these segments is independent and displays a specific course and limit. Each segment being independent, an anomaly in that segment may result in segmental agenesis of the ICA. Formation of the ICA is completed at the 6th week of gestation (4th embryonic week), while the carotid canals form between the 5th and 6th embryonic weeks.^[1,6] Since the presence of the ICAs is a prerequisite for the formation of the carotid canals, ICA agenesis results in absence of the carotid canals. In case of aplasia or hypoplasia, the size of the carotid canals is reduced.^[7]

Collateral supply in ICA agenesis is either from the posterior circulation, through the posterior communicating arteries; via a persistent fetal circulation, such as a persistent primitive trigeminal artery; or through transcranial collaterals from branches of the internal maxillary artery or inter-cavernous collaterals.^[1,3,7] Dilatation of the vertebrobasilar system, anterior circulation, or the external carotid system results in adequate collateral blood supply in the agenetic segment. In unilateral agenesis, the predominant supply is through the contralateral ICA^[4] and in bilateral agenesis, through the vertebrobasilar system.^[1] In our patient, the supply was predominantly through the external carotid system and partly from the posterior circulation through the right posterior communicating artery. The hemodynamic stress placed on vessels due to increased flow through the collateral channels may result in the development of flow-related aneurysms. The prevalence of cerebral aneurysms in the general population is 2–4%, whereas it is 24–34% in patients with ICA agenesis.^[1,4]

Patients with ICA agenesis are usually asymptomatic since cerebral perfusion is usually adequate due to collateral supply. This anomaly is, therefore, rarely seen in children. These patients may present with symptoms of cerebrovascular insufficiency, e.g., transient ischemic attacks, hemiparesis, or hypotensive episodes due to atheromatous changes in the vessels from which the collaterals arise or they may present with signs of a mass effect due the collateral channels, e.g., cranial nerve palsies

(as seen in our patient), Horner's syndrome,^[6] or seizures, etc.) They may also present with complications (e.g., subarachnoid or intracerebral hemorrhage) arising from associated conditions such as aneurysms or arteriovenous malformations.

ICA agenesis is associated with many conditions, but most commonly with aneurysms. Though associated aneurysms of the anterior communicating artery are most commonly described in literature,^[3,8] aneurysms of the basilar artery,^[9] posterior communicating artery, posterior cerebral artery, and the first part of the subclavian artery have also been described. Other associated conditions reported are cerebral arteriovenous malformations, hypopituitarism, neurofibromas, interruption of the aortic arch, aberrant origin of the ophthalmic artery, CCA agenesis/hypoplasia, basilar artery aplasia with nasopharyngeal angiofibroma, corpus callosum agenesis,^[10] and congenital temporomandibular joint ankylosis. ICA agenesis may also occur as part of the PHACE (posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of aorta, cardiac defects, and eye abnormalities) anomaly.^[11]

A plexus of vessels arising from the vertebral arteries, with multiple aneurysmal sacs and calcifications, reformation of the basilar artery from the plexus of vessels, along with a tortuous first part of the right subclavian artery, has not been reported so far in association with ICA agenesis. Disregulated angiogenesis during the formation of the vertebral arteries might have resulted in this plexus of vessels in the posterior fossa. The aneurysmal dilatation may have been due to weakened vessel walls.

The ICA can be severely narrowed or occluded in various conditions such as atherosclerosis, arteritis, arterial dissection, fibromuscular dysplasia, Moya-moya disease, and sickle cell disease. In these conditions, ICA agenesis may be misdiagnosed. In such cases, the ICA vessel stump, the thickened arterial wall, and the offending plaque or thrombus are usually well seen and the distal run-off can usually be visualized. In the case of dissection of the arterial wall, the intimal flap is usually seen on contrast-enhanced CT or MRI. Aggenesis can be distinguished from acquired stenosis by looking for the absence of the carotid canals, which is best demonstrated on a CT scan of the skull base. CT of the skull base is also needed to distinguish aplasia from agenesis. In our patient, both the carotid canals were absent and the foramina ovale were enlarged.

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Source of Support: Nil, **Conflict of Interest:** None declared.

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